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Palliative Care in Amyotrophic Lateral Sclerosis

FROM DIAGNOSIS TO BEREAVEMENT

THIRD EDITION

EDITED BY
DAVID OLIVER | GIAN DOMENICO BORASIO | WENDY JOHNSTON

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Preface

Although there is increasing understanding of the genetic basis of amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND) and the basic science of the disease, currently available treatments offer only a modest increase in life expectancy. Therefore, a palliative approach to caring for the patient and their family is at the centre of good clinical practice in ALS. This book considers the various aspects of palliative care for patients with ALS, based on the most recent research in the field. This edition has been updated to include research and publications from the past 7 years, as well as new chapters on complementary medicine and family experience.

All contributors are clinicians involved in the care of people with ALS who provide clear guidelines of the care involved throughout the disease process, from imparting the diagnosis to bereavement. The international authorship allows a comparison of the differences across cultures, social circumstances, healthcare systems, and nations. The palliative approach to the care in ALS is outlined and the role of the multidisciplinary team emphasized. This book adopts an evidence-based approach, grounded in the day to day care of patients. A short case history starts all chapters, focusing the reader on the clinical application of the research to be discussed.

The authors aim to provide a reference for all involved in the care of patients with ALS, including neurologists, primary-care physicians, rehabilitation specialists, allied health professionals, and particularly multidisciplinary palliative care teams. The care of a person with ALS, and their family, is often a challenge, but one that all should take up so that patients can live their lives as fully as possible.

Throughout the book the term 'amyotrophic lateral sclerosis' (ALS) has been used rather than 'motor neuron disease', as the name ALS is recognized internationally.

We would like to thank all involved in the production of this volume, especially Caroline Smith from Oxford University Press, Professor Declan Walsh, who was a co-editor of the first and second editions, and all our colleagues who have helped us with comments and suggestions throughout the publication process.

DJO, Rochester, UK
GDB, Lausanne, Switzerland
WJ, Edmonton, Canada
June 2013

Contents

List of abbreviations

AAC	augmentative and alternative communication
AAN	American Academy of Neurology
AD	advance directive
AFO	ankle–foot orthosis
ALS	amyotrophic lateral sclerosis
ALS-bi	ALS behavioural impairment
ALSFRS	ALS Functional Rating Scale
BDNF	brain-derived neurotrophic factor
BMI	body mass index
bp	base pair
bvFTD	behavioural variant of FTD
CAM	complementary and alternative medicine
C9orf72	chromosome 9 open reading frame 72
CCTR	Cochrane Controlled Trials Register
CNS	central nervous system
CNTF	ciliary neurotrophic factor
CSF	cerebrospinal fluid
DPS	diaphragmatic pacing
DVT	deep venous thrombosis
EAAT	excitatory amino acid transporter
EAPC	European Association for Palliative Care
EEG	electroencephalogram
EFNS	European Federation of Neurological Societies
EMG	electromyography
EOL	end of life
ETRAN	eye transfer communication board
FEES	fibreoptic endoscopic evaluation of swallowing
FTD	frontotemporal dementia
FUS	fused in sarcoma
FVC	forced vital capacity
GABA	gamma-amino-butyric acid
GERD	gastro-oesophageal reflux disease
IGF	insulin-like growth factor
IPPB	Intermittent positive pressure breathing
IV	intravenous
LMN	lower motor neuron
LTMV	long-term mechanical ventilation
MBS	modified barium swallow
MEP	maximum expiratory pressure
MI-E	mechanical insufflator–exsufflator
MIP	maximum inspiratory pressure
MMN	multifocal motor neuropathy
MNDA	[UK] Motor Neurone Disease Association
MRI	magnetic resonance imaging
MS	multiple sclerosis
NF	neurofilament
NGF	nerve growth factor
NICE	National Institute for Health and Care Excellence
NIPPV	non-invasive positive-pressure ventilation
NIV	non-invasive ventilation
NSAID	non-steroidal anti-inflammatory drug
ODWDA	Oregon Death with Dignity Act
OT	occupational therapy
PAD	physician-assisted death
PAS	physician-assisted suicide
PCF	peak cough flow
PCVGV	pressure-controlled volume-guaranteed ventilation

PE	pulmonary embolism
PEG	percutaneous endoscopic gastrostomy
PLS	primary lateral sclerosis
PMA	progressive muscular atrophy
PRG	percutaneous radiological gastrostomy
QOL	quality of life
REM	rapid eye movement [sleep]
ROM	range of movement
SBMA	spinobulbar muscular atrophy
SDB	sleep-disordered breathing
SLT	speech and language therapist
SMA	spinal muscular atrophy
SNIP	sniff nasal inspiratory pressure
SOD1	superoxide dismutase 1
SSRI	selective serotonin re-uptake inhibitor
SVC	slow vital capacity
TENS	transcutaneous electrical nerve stimulation
TV	tracheostomy ventilation
UMN	upper motor neuron
VFSS	videofluoroscopic swallowing study
WMA	World Medical Association

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Chapter 1

Amyotrophic lateral sclerosis/motor neuron disease

Christopher E. Shaw, Annika Quinn, and Emma Daniel

Summary

The aim of this chapter is to answer the question 'What is amyotrophic lateral sclerosis (motor neuron disease)?'. We will start with an account of the typical clinical presentation and diagnostic work-up of a patient with amyotrophic lateral sclerosis (ALS) and a brief description of the conditions that cause or mimic motor neuron degeneration. Then we will consider the pathological features of ALS and what molecular genetic and cell biology research can tell us about its pathogenesis. Lastly, therapies designed to alter the course of the disease will be discussed.

What do we mean by motor neuron disease?

The term 'motor neuron disorders' covers a range of conditions in which the motor neuron cell bears the brunt of the disease process. Clinical, pathological, and, more recently, molecular genetic studies have helped to distinguish many of these disorders from typical motor neuron disease. Amyotrophic lateral sclerosis (ALS) is the name used for typical motor neuron disease in most parts of the world—in the UK and other Commonwealth countries it is known as motor neuron disease and in the United States it is often known as Lou Gherig's disease, after the famous baseball player who died of ALS.

It was originally thought to be a muscular condition, until Charcot in 1869 published clinico-pathological studies which correctly identified the degeneration of motor neurons as the cause of muscle wasting.[1] The feature that distinguishes this disorder from others affecting the motor system is the combination of degeneration of both upper and lower motor neurons (the neurological pathways are shown in Fig. 1.1).

Lower motor neurons (LMNs) reside in the spinal cord and brainstem and project out in peripheral nerves to make direct contact with and activate muscle

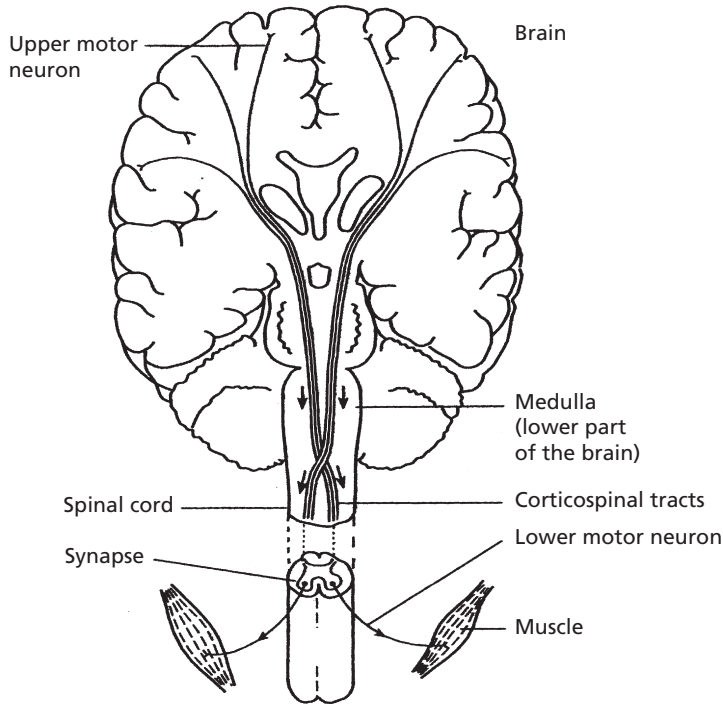

Fig. 1.1 The neurological pathways within the spinal cord and brain.

fibres. When LMNs degenerate, the muscles they activate become weak and wasted and fasciculate (twitch). Upper motor neurons (UMNs) reside in the motor cortex in the frontal lobes of the brain and they project down to and activate the LMNs. When UMNs degenerate, muscles become spastic and the deep tendon reflexes are exaggerated. The plantar responses may be extensor rather than flexor (Babinski's sign—the big toe goes up rather than curling down when the sole of the foot is scratched). Most patients with ALS have a mixture of UMN and LMN signs; however, when only LMN features are present the condition may be called progressive muscular atrophy (PMA) while a pure UMN syndrome is known as primary lateral sclerosis (PLS). Pathological studies suggest that these syndromes are all forms of ALS but lie at the ends of a clinical spectrum predominantly affecting the lower or upper motor neurons respectively. Interestingly, both variants are associated with a slower disease progression and longer survival.

The diagnosis of ALS is essentially a clinical one, and supportive evidence is obtained by a range of investigations. Typically ALS presents with progressive weakness and wasting of the muscles controlling limb movement, speech, or

- **Must be** progressive, have fasciculation and near normal motor nerve conduction
- **Must not** have cognitive, parkinsonism, sensory, visual, autonomic, sphincter involvement.

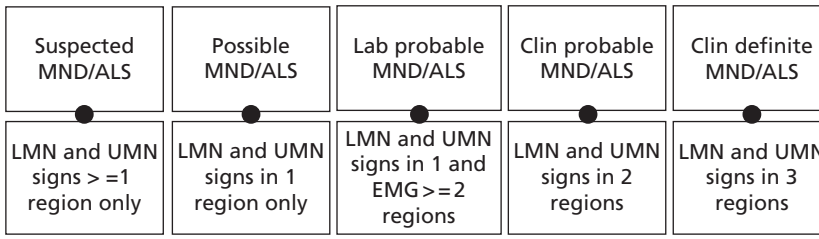

Fig. 1.2 Revised El Escorial criteria for the diagnosis of motor neuron disease (MND)/amyotrophic lateral sclerosis (ALS) (LMN, lower motor neuron; UMN, upper motor neuron; EMG, electromyography).

swallowing. Motor neurons controlling eye movement and sphincters of the bladder and bowel are usually spared, as are neurons in the sensory and autonomic nervous systems. Diagnostic criteria for ALS were established at a conference in the Spanish castle El Escorial and were subsequently updated in 2000[2] (Fig. 1.2). They are principally used as inclusion criteria in therapeutic trials but emphasize the importance of detecting UMN and LMN signs in multiple regions to establish a more definite diagnosis of ALS.

Motor neuron disorders that mimic ALS

There are many different motor neuron disorders that may be mistaken for ALS. Given the very poor prognosis for most patients it is vital that the diagnosis be correct and all possible alternatives excluded. The most common mimic is spinal cord and/or nerve root compression due to degeneration of the vertebrae and discs in the spine. This can present with painless muscle wasting, weakness, and fasciculation in one or more limbs. Detailed views of the spinal cord and nerve roots by magnetic resonance imaging (MRI) scanning are essential in every patient suspected to have ALS. MRI may also reveal rare spinal cord tumours or syrinx (cystic expansion within the cord). Another condition that requires careful consideration is multifocal motor neuropathy (MMN). This is an autoimmune disorder where antibodies selectively attack the motor nerves, and it usually presents as an asymmetrical weakness of the upper limbs. MMN is often associated with patches of reduced electrical conduction in motor nerves, demonstrated by careful nerve conduction studies, and anti-GM1 ganglioside antibodies in the serum.[3] This mimic of ALS is not lethal and can improve following immunosuppressive treatment, such as intravenous or subcutaneous human immunoglobulin.

Kennedy disease, or spinobulbar muscular atrophy (SBMA), is a rare condition but a significant cause of misdiagnosis in ALS. It is a pure lower motor neuron syndrome that solely affects adult men. It causes slowly progressive muscle wasting with prominent involvement of the tongue and tremor of the outstretched hands. Other features include testicular atrophy and gynaecomastia (breast enlargement), due to low androgen levels.[4] Nerve conduction studies commonly show a mild sensory neuropathy even though there may be no evidence of sensory loss on examination. The mutation is an expanded CAG nucleotide repeat sequence in the androgen receptor gene on the X chromosome.[5] The CAG triplet encodes for the amino acid glutamine and the expansion mutation makes the androgen receptor protein unfold and accumulate, which is toxic to motor neurons.

Even rarer is the autosomal recessive condition spinal muscular atrophy (SMA). Although SMA usually presents in infancy or childhood, a small minority of cases present in adulthood with SMA type IV, a slowly progressive, pure LMN syndrome with wasting, weakness, and absent reflexes.[6] The survival motor neuron gene (*SMN*) is found to be deleted or disrupted in more than 95% of all cases of SMA.[7]

Many other conditions need to be considered in the differential diagnosis, such as multiple sclerosis and hereditary spastic paraplegias which can cause UMN signs. Muscle twitching without focal wasting is a feature of the benign fasciculation syndrome, as well as of disturbed thyroid and parathormone disorders, so a metabolic screen should be undertaken (Table 1.1).

The clinical course of typical ALS

In the majority of people with ALS, muscle weakness begins insidiously in one limb (85%), causing weakness of grip or the foot to catch on the pavement. Affected muscles become progressively more wasted and fasciculate as LMNs in the spinal cord degenerate and die. Sometimes muscle cramps or spasms precede wasting and weakness, which may reflect early UMN involvement. Spasticity may accompany weakness, and eventually all of the limbs become affected. In a minority of people symptoms begin in the throat (15%) with slurred speech or difficulty swallowing.[8] Bulbar symptoms arise when LMNs in the brainstem (previously known as the bulb), degenerate causing wasting and weakness of the tongue and pharyngeal muscles, often referred to as 'progressive bulbar palsy' (Fig. 1.3). Sometimes UMN signs affect the throat, resulting in nasal and slurred speech, due to poor elevation of the soft palate, and spasticity of the tongue accompanied by a brisk jaw jerk, together often referred to as 'pseudobulbar palsy'. These two syndromes are also variants of ALS and are

Table 1.1 Conditions mimicking amyotrophic lateral sclerosis

Condition	Diagnostic screening tests
Spinal disease causing cord and/or nerve root compression (myelo-radiculopathy)	Magnetic resonance imaging scan
Autoimmune neuropathies including multifocal motor neuropathy	Nerve conduction studies, anti-ganglioside antibodies, protein electrophoresis
Spinobulbar muscular atrophy (also known as Kennedy disease)	Genetic test for androgen gene CAG expansion mutation
Multiple sclerosis	Magnetic resonance imaging of the brain and cerebrospinal fluid, immunoglobulin electrophoresis
Hereditary spastic paraplegia	Electromyography (no evidence of lower motor neuron disease), gene testing
Benign fasciculation syndrome	Electromyography (no evidence of lower motor neuron disease)
Myopathies (e.g. inclusion body myositis)	Muscle biopsy
Spinal muscular atrophy	Gene testing
Diabetic amyotrophy	Glycosylated haemoglobin
Thyrotoxicosis	Thyroid hormone assay
Hyperparathyroidism	Calcium, phosphate
Gangliosidoses	White cell enzyme hexosaminidase levels

usually accompanied by motor neuron symptoms and signs. Approximately 90% of patients with limb-onset disease will eventually develop bulbar symptoms, and only a small minority, more commonly men, have solely spinal disease.

ALS causes significant disability early and relentlessly progresses so that most patients ultimately lose the ability to walk, feed and toilet themselves, speak, and swallow. A particular cruelty is that intellectual function is largely spared so that patients are fully aware of their circumstances but are trapped in bodies that no longer work and are isolated by an inability to interact or communicate. In around 10% of patients, abnormal personality, behavioural, and language problems develop that typify frontotemporal dementia, but up to 30–40% of people with ALS have similar but more subtle cognitive deficits (see Chapter 8). Respiratory weakness due to thoracic and high cervical spinal cord involvement is almost universal prior to death. Respiratory weakness is often underestimated because the patient's mobility is compromised and a drop in the ventilatory capacity to 60% of predicted may be asymptomatic. Death is usually due to

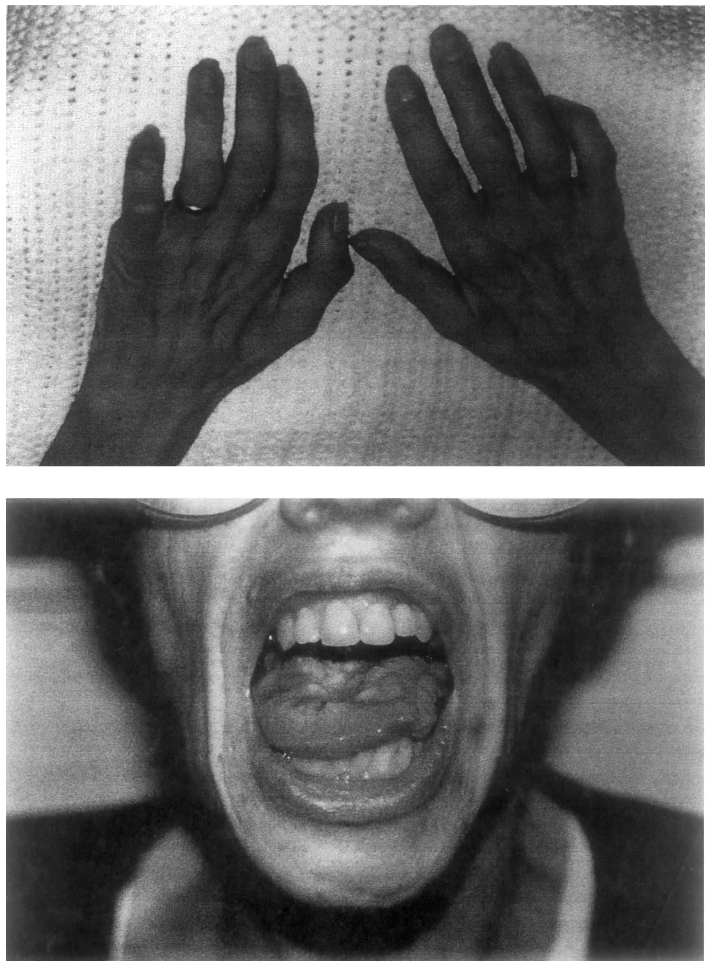

Fig. 1.3 Clinical appearance of amyotrophic lateral sclerosis. Note the pronounced muscle wasting of the patient's hands and tongue.

respiratory failure and mean survival from symptomatic onset is on average 3 years. Only 25% of patients survive 5 years and 10% are alive at 10 years.[9] Elderly female patients who have a bulbar onset of symptoms have a significantly poorer prognosis, while more slowly progressive disease is seen in younger men who present with weakness in the limbs.

How can the diagnosis of ALS be made?

ALS is relatively uncommon, and the diagnosis can be difficult. Most general practitioners will encounter only one case in their working lifetime. For this

reason they may not recognize the earliest symptoms or signs so that patients may be referred to specialists in rheumatology, orthopaedics, or otolaryngology before they see a neurologist. While the symptoms and signs on examination may be suggestive of ALS there is no simple 'test for ALS'. Furthermore, as the implications are so profound, neurologists may delay discussing the possibility of ALS until they are absolutely certain of the diagnosis. These factors can contribute to a considerable delay from the time of first symptom to diagnosis.

The investigations that are most useful are those that exclude other conditions that mimic ALS (Table 1.1). The most important is MRI of the spine or head to exclude an extrinsic or intrinsic lesion. Nerve conduction studies are essential to exclude a generalized or multifocal neuropathy and search for nerve entrapment or conduction block. Electromyography (EMG) is necessary to confirm evidence of acute and/or chronic lower motor neuron loss, particularly in regions not symptomatically affected, and to exclude myopathy. Typical features of ALS on EMG are spontaneous fibrillations and slow-frequency fasciculations (0.3 Hz) with unstable and complex motor unit potentials on voluntary activation. Muscle biopsy and spinal fluid studies are usually unnecessary unless the presentation is very atypical and an alternative diagnosis, such as a degenerative muscle condition or an inflammatory disease such as multiple sclerosis is suspected.

Who gets ALS?

For reasons that are not known men develop ALS more frequently than women with a ratio of approximately 1.7:1.[1] The mean annual incidence of ALS is approximately 1–2 per 100 000 people per year, which is roughly half that of multiple sclerosis in the UK. Because of the relatively short survival of patients with ALS, the prevalence rate is only 3–7 per 100 000. The prevalence appears to be fairly consistent despite geographical, socio-economic, and racial differences. High-risk foci exist in the Kii Peninsula in Japan and the Pacific island of Guam where it is often associated with Parkinsonism and dementia. The incidence in Guam has been has been falling over the last three decades, which suggests that exposure to an environmental agent may be responsible: however, many case–control studies have failed to identify the particular toxin or infection that might be a risk factor for ALS. Epidemiological studies outside Guam suggest that a history of previous musculoskeletal injury[11] and occupations with a high degree of electrical exposure or electric shock[12] appear to increase the risk of ALS two- to three-fold, but the effect is relatively small.[9] The association with increased age is clear, however, as there is a steep rise in incidence after the age of 50 which continues to increase in age-adjusted analyses.

What happens to motor neurons in ALS?

Although muscular and anterior spinal root atrophy is striking at post-mortem, the brain and spinal cord usually look reasonably normal. The minor external changes are in contrast to the dramatic changes that occur at a microscopic level. Charcot[1] was one of the first to describe the abnormal appearance of neurons within the spinal cord. The characteristic features are of often severe loss of motor neurons with proliferation and hypertrophy of neighbouring astrocytes (supporting brain cells). The few surviving motor neurons are either shrunken or swollen and have a variety of protein aggregates (cytoplasmic inclusions) and axonal degeneration (Fig. 1.4). Similar changes are seen in the large UMNs in the cerebral cortex (Betz cells), although there is increasing evidence that neuronal loss is common in other, non-motor, regions of the cortex.[13] Cytoplasmic aggregates of the TAR DNA-binding protein (TDP-43) of motor neurons are the pathological hallmark of ALS and are present in more than 90% of cases.[14]

The genetics of ALS

To date the most important clues as to the pathogenesis of ALS have come from molecular genetics. Although most cases of ALS occur sporadically, approximately 5–10% of the time other members of the family are also affected. Most commonly, familial ALS is a dominant disorder, being passed down through the generations, and in most cases it is clinically indistinguishable from sporadic disease.

The first genetic factor linked to ALS was reported in 1991 to lie on chromosome 21,[15] and 2 years later that factor was identified as the copper/zinc superoxide dismutase gene (*SOD1*), in which mutations were discovered.[16] SOD1 is a powerful antioxidant catalytic enzyme responsible for neutralizing potentially harmful free radicals produced as a result of normal cellular activity. To date more than 150 different mutations have been described, mostly due to a single base change causing a single amino acid substitution.[17,18] Interestingly, not all toxic ALS-associated SOD1 mutations impair its antioxidant function, and the toxicity of mutants appears to be due rather to the accumulation of SOD1 protein.[19] Patients with *SOD1* mutation show accumulation of SOD1 protein but not of TDP-43, a pathology distinct from that of most other ALS.[20,21] Mutations in *SOD1* are found in approximately 20% of familial and 3% of apparently sporadic cases.[22]

The most common gene associated with ALS in European populations is a massive expansion in the chromosome 9 open reading frame 72 (*C9orf72*) gene.[23,24] *C9orf72* mutations account for around 35% of familial and 5% of sporadic ALS.[25] Most people have only two to eight repeats of a 6 base-pair (bp)

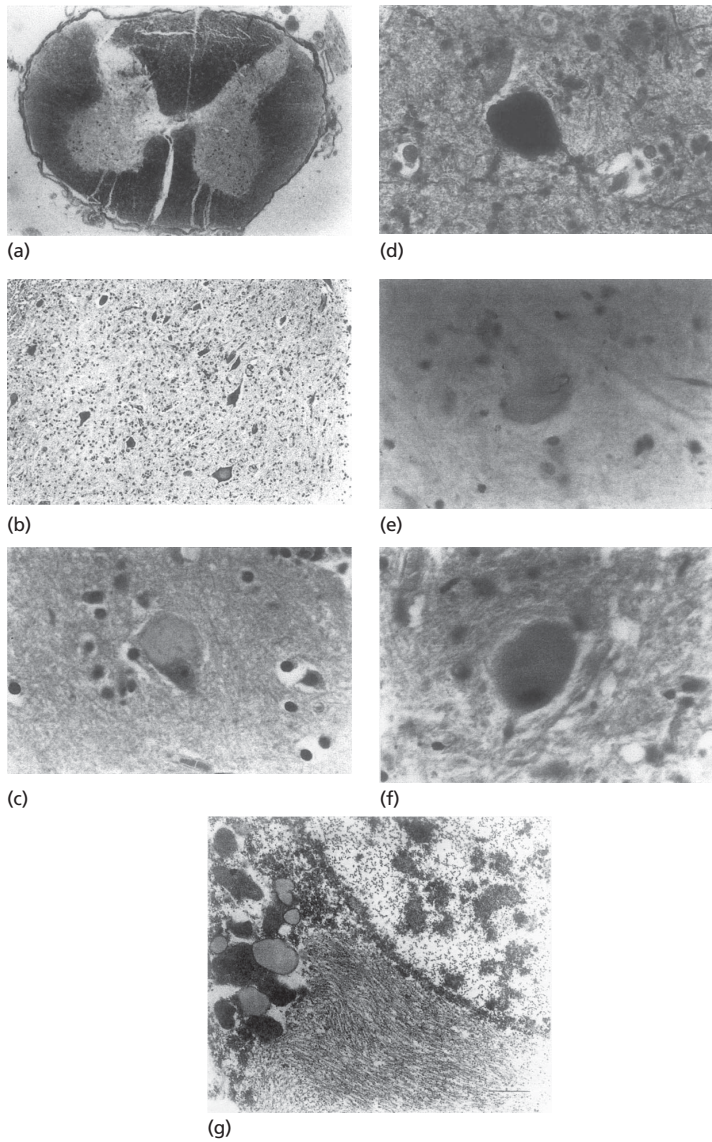

Fig. 1.4 Pathological features of amyotrophic lateral sclerosis by light and electron microscopy. (a) Spinal cord in cross section showing a low number of motor neurons in the anterior horn. (b) At low-power magnification motor neurons in the cord appear swollen or shrunken. At higher power a variety of aggregates can be seen: including (c) hyaline inclusions, (d) those that immunolabel with antibodies to neurofilaments and with ubiquitin showing (e) the characteristic thread-like skeins, and (f) Lewy body-like and proximal axonal inclusions. (g) Transmission electron microscopy reveals that the cell body is packed with neurofilaments, lipofuscin, and other protein aggregates (scale bar = 1 μm).

sequence (GGGGCC), but those carrying the mutation have 600–2000 repeats. The function of the C9orf72 protein is currently unknown. Loss of C9orf72 protein has been proposed as a mechanism of disease in these patients, as has a toxic effect of the long repeat RNA or the production of aggregate-prone proteins from the repeat RNA. Cases with *C9orf72* mutation are characterized pathologically by an accumulation of TDP-43 protein, although aggregates devoid of TDP-43 are also present.[26]

Mutations in the gene encoding TDP-43 are rare, accounting for only about 1% of familial and sporadic ALS, but accumulation of TDP-43 is a feature of 95% of ALS cases, whether familial or sporadic. This connection between genetics and pathology confirms that the accumulation of TDP-43 is toxic.[13,27] TDP-43 normally resides in the nucleus where it plays a major role in the regulation and editing of gene transcription. In ALS, and 60% of cases of frontotemporal dementia, TDP-43 is lost from the nucleus and it forms granular, globular, and skein-like inclusions in the cell body and axons, that are decorated with ubiquitin and p62.[13] TDP-43 is more likely to accumulate in the cytoplasm and aggregate leading to toxicity when it is mutated, but the overexpression or loss of healthy 'wild-type' TDP-43 is also potently toxic. It is clear that levels of TDP-43 protein must be controlled within a narrow window for motor neurons to survive. Protein levels can be regulated by targeting damaged or mutated proteins for degradation through the attachment of ubiquitin and p62 which facilitate the delivery of protein to the recycling machinery. For reasons that are still unknown, this degradation process breaks down and the accumulation of toxic proteins such as TDP-43 may directly contribute to the death of motor neurons.

Fused in sarcoma (FUS) is another nuclear protein involved in regulating gene expression that has been implicated in ALS by the discovery of FUS mutations in 1–4% of families.[28] Like TDP-43, accumulation of FUS protein in the cytoplasm of motor neurons is a feature of patients carrying pathogenic mutations.

Other genes implicated in ALS include those that are involved in degradation of proteins such as valosin containing protein (*VCP*), ubiquilin 2 (*UBQLN2*), and sequestosome 1 (*SQSTM1*). The VCP, UBQLN2, and SQSTM1 proteins all help to deliver damaged proteins to the cellular recycling system. Failure to clear TDP-43 leads to its accumulation, which subsequently causes neurodegeneration.

Hypotheses about the pathogenesis of ALS

The genetic mutations outlined in the previous section (The genetics of ALS) show a strong causal relationship with ALS, leading to a clear pattern of inheritance in families. Genetic factors with smaller effects on the risk of developing

ALS are also beginning to be identified. It is likely that sporadic ALS may involve interactions between several genetic risk factors and/or between genetic and environmental factors.[29]

There are a great number of environmental insults postulated to play a role in ALS. Although viral infection, exposure to toxins, and autoimmunity have been implicated, an extensive body of research has failed to support these hypotheses. Although there is some evidence that head or spinal injury or exposure to electrical shocks increases risk, the effect is relatively modest at around two- to three-fold. An association with physically demanding occupations has also been proposed, based upon incidences in sportspeople and those who exercise frequently, but robust epidemiological evidence is awaited.

This section discusses what the genetic and environmental factors associated with disease can tell us about the pathogenesis of ALS. The focus will be on mechanisms for which there is the greatest experimental evidence: protein aggregation, altered regulation of gene expression, excitotoxicity, oxidative injury, cytoskeletal disruption, and loss of neurotrophic support (summarized in Fig. 1.5). Each theory has its proponents and can be argued for by observation and experimental research, but none are mutually exclusive. Given the diverse genetic factors associated with risk of developing ALS, it may be that primary dysfunctions in any one of several biochemical systems converge to a common pathway and that each of these mechanisms may contribute to the death of motor neuron cells.

Protein aggregation

The abnormal clumping of proteins and their deposition as aggregates in neurons is a common feature in many neurodegenerative diseases, including ALS. Cases with *SOD1* mutation are characterized by SOD1 protein aggregates, and cases with *FUS* mutation by FUS protein aggregates, while almost all other cases are characterized by TDP-43 protein aggregates. However, the role of protein aggregates themselves in initiating disease is controversial. Evidence from research on Alzheimer's disease suggests that many small clusters of abnormal protein stuck together can do more harm than one large aggregate.[30] This harm may include clogging the protein degradation machinery in the neuron or recruiting bystander proteins into the cluster. Protein aggregation may therefore contribute to loss of the normal activity of the aggregating protein as well as bystander proteins. Current efforts to find therapeutics for ALS involve screening for drugs which reduce protein aggregation.

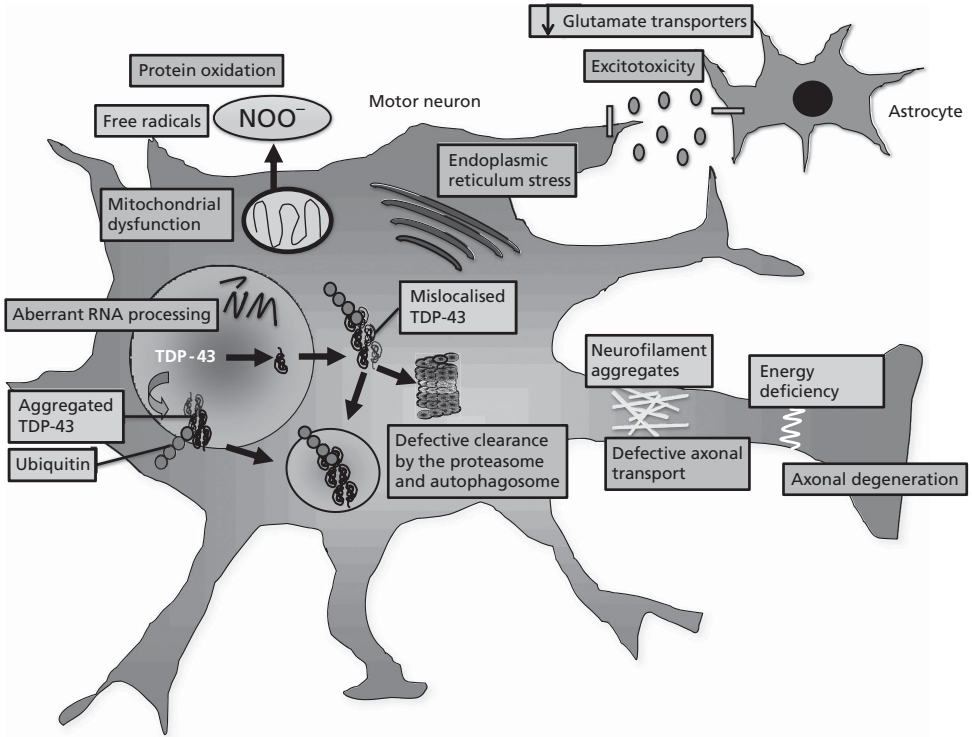

Fig. 1.5 Pathogenesis of amyotrophic lateral sclerosis (ALS): summary of hypotheses.

The molecular mechanisms leading to motor neuron degeneration in ALS. By various mechanisms including aberrant RNA processing in the nucleus and overproduction of free radicals secondary to mitochondrial dysfunction, the nuclear protein TDP-43 folds aberrantly, aggregates, and mislocalizes to the cytoplasm. The aberrantly folded TDP-43 is tagged for degradation with the molecule ubiquitin; however, neither the ubiquitin–proteasome system nor the autophagosome degradation system are able to clear the protein, and aggregates of TDP-43 remain in the cell. These aggregates lead to stress at the endoplasmic reticulum. Astrocytes contribute to motor neuron damage in many ways, including the reduced expression of glutamate receptors, leading to excitotoxicity. Aggregated neurofilaments in the axon may act to block axonal transport and lead to distal energy deficiency, which may contribute to axonal degeneration.

Altered regulation of gene expression

Two proteins linked to ALS are involved in modulating gene expression (TDP-43 and FUS). Gene expression, or the production of proteins from genes, is a highly dynamic process which allows neurons to adjust to stress, starvation, and differing activity levels. Mutations in the master proteins controlling gene expression affect the production of a huge number of proteins under their control, and therefore influence the ability of neurons to cope with insults. Recently, it was found that TDP-43 regulates the expression of over 600 different proteins.[31] C9orf72 mutations may also alter gene expression. The long repeat RNA becomes tangled, so that in addition to not delivering the message to produce C9orf72 protein, this RNA may tie up proteins which play similar roles to TDP-43 and FUS in regulating gene expression.[32]

Glutamate and neuronal excitotoxicity

The finding that neurotransmitters and their chemical analogues which activate neurons can also harm them prompted a great deal of research into their role in the pathogenesis of many neurodegenerative diseases. The excitotoxic hypothesis in ALS has centred around the handling of the amino acid glutamate. When a neuron is sufficiently 'excited', an electrical impulse is propagated down its axon to the terminal. This causes the release of a chemical messenger or neurotransmitter into the synapse, the tiny space between the communicating neuron and its target neuron. Glutamate is the principal neurotransmitter used by neurons to communicate with the motor neurons in the cortex and spinal cord. Glutamate is potentially toxic, and the brain contains more than 1000 times the amount of glutamate necessary to kill every neuron. Therefore the strict regulation of glutamate levels is of vital importance and is principally the task of neighbouring astrocytes. Only a small proportion of the glutamate released into the synapse binds to receptors on the target neuron; the remainder is taken up and removed from the synaptic cleft by excitatory amino acid transporters (EAATs) (see Fig. 1.5).

Given that glutamate is so potently toxic to neurons, what evidence is there that its regulation is disturbed in ALS? In one study, levels of glutamate in the cerebrospinal fluid (CSF) were found to be increased up to three-fold in 80% of ALS patients compared with controls.[33] One of the transporters responsible for glutamate reuptake from the synapse, EAAT2, is reported to be selectively reduced in the motor cortex and spinal cord of up to 70% of patients with sporadic ALS.[34] Reduced reuptake of glutamate may therefore be responsible for the increased levels seen in the CSF. However, other laboratories report that

glutamate is increased in the CSF in only a minority of patients (30%)[35] and that decreased levels of EAAT2 protein are only found in the spinal cord.[36] Thus, glutamate-induced toxicity may contribute to, but is unlikely to be solely responsible for, motor neuron degeneration.

Free radicals, oxidative stress, and ALS

During normal oxidative metabolism, several species of free radicals containing reduced oxygen ions are produced (O^-, NO^-, and OH^-). Free radicals are capable of damaging proteins, lipids, and nucleic acids and are thought to contribute to the normal ageing of cells. All cells have a range of self-protective antioxidant molecules to mop up free radicals as well as mechanisms to identify and remove oxidized cellular components. Protein oxidation has been detected in the motor cortex and spinal cord in patients with ALS[37] and mutant *SOD1* transgenic mice.[38]

Another potential oxidative toxin to cells is peroxynitrite (NOO^-). This free radical can bind to the amino acid tyrosine, a major component of many proteins, to form 3-nitrotyrosine. Evidence of NOO^- injury and 3-nitrotyrosine has been described in tissue of people with sporadic and familial ALS[39] and in mutant *SOD1* transgenic mice.[40] Moreover, dramatically increased levels of 3-nitrotyrosine have been detected in the CSF of people with sporadic ALS.[41] While these increases are non-specific and occur in other neurodegenerative diseases, they do suggest that the nitrosylation of proteins is occurring at an increased rate in ALS, supporting the case that oxidative stress contributes to the degeneration of motor neurons.

The ALS-linked proteins TDP-43 and FUS reveal another potential role for oxidative stress in ALS. Both are involved in the regulation of gene expression under conditions of cellular stress, such as oxidative injury. In order to remain healthy in the face of stress, the cellular stress response must focus on producing proteins that are critical to cell health while pausing production of non-essential proteins. TDP-43 and FUS are part of the process which draws non-essential RNAs together as granules in the cytoplasm and stalls their protein production.[42,43] It has been proposed that these granules, containing high levels of TDP-43 and/or FUS protein, may initiate the formation of the cytoplasmic aggregates seen in ALS patient tissue.

Cytoskeletal disruption and motor neuron degeneration

Motor neurons are the biggest cells in the body and in the majority of people they function perfectly throughout a normal lifespan. Following the identification of

neurofilament (NF) aggregates within motor neurons, a role for neurofilament proteins in the pathogenesis of ALS has been postulated. Neurofilaments are linked together to form the internal scaffolding for neurons, in particular maintaining their axonal diameter. They are aligned with microtubules which are the highways along which mitochondria, vesicles, and proteins, for example, are transported. Mice transgenic for a point mutation (a single 'spelling mistake') in the mouse NF light chain[44] or over-expressing normal human NF heavy chain[45] develop motor neuron degeneration with some pathological similarities to ALS. When mice deficient in neurofilaments were crossed with mutant *SOD1* mice the onset of motor neuron degeneration and death was significantly delayed.[46] While mutations in the NF genes have not been found in familial MND/ALS cases, deletions in the tail domain of the NF heavy chain gene were detected in 10 sporadic cases.[47,48] In addition, the stability of NF light chain RNA is controlled by TDP-43, meaning that TDP-43 mislocalization or aggregation may precede NF disruption.[49] Collectively this work suggests that NF aggregation may have a contributory role in motor neuron injury but it is not a primary or essential mechanism in the pathogenesis of ALS.

Neurotrophic factors and neurodegeneration

The discovery that selected populations of neurons are dependent on specific neurotrophic factors, both during early development and for long-term survival, has prompted research into their role in the pathogenesis and treatment of neurodegenerative diseases. While embryonic dorsal root ganglion neurons have an absolute requirement for a range of neurotrophic factors, this is not the case for motor neurons. Ciliary neurotrophic factor (CNTF) and insulin-like growth factor (IGF) do enhance the survival of embryonic motor neurons grown in culture, and both CNTF and brain-derived neurotrophic factor (BDNF) will rescue facial motor neurons following axotomy.[50] Furthermore, CNTF can retard the degeneration of motor neurons in the progressive motor neuronopathy (pmn) mouse.[51] A relative decrease in CNTF expression in the anterior spinal cord and nerve growth factor (NGF) in the motor cortex in ALS have been described in ALS tissue at post-mortem but it is difficult to distinguish cause and effect. While it is an attractive hypothesis that defective generation of neurotrophic factors by target cells or their retrograde transport in axons will result in neurodegeneration, clear evidence in human disease states is lacking. However, neurotrophic factors may still have a role in increasing the survival of ailing neurons and a great deal of experimental work in this field is currently under way together with several phase III clinical trials.

Drugs that alter survival in ALS

In a survey of published therapeutic trials in ALS recorded in the Cochrane Controlled Trials Register (CCTR) 57 studies were identified using 31 different treatments.[52] Of these only the glutamate release inhibitor riluzole has shown a statistically significant effect on survival in a multicentred, double-blind, controlled trial of 957 patients.[53] Analysis of this trial, and three subsequent randomized controlled trials, showed a 3-month delay in the time to death or tracheostomy in the active treatment group compared with controls during an 18-month trial period.[54] Clinical trials have not shown other agents that influence the glutaminergic pathways to modify function or survival in ALS.[55–57] Of the neurotrophic agents, IGF, CNTF, and BDNF have all been trialled using a subcutaneous route of administration without a statistically significant effect on survival.[58,59] Another important tool in screening drug efficacy is the range of transgenic mouse models of ALS. Mutant *SOD1* mice have been the predominant drug screening model for ALS; however, *SOD1* mutation is a rare cause of ALS. More recently, mice which show accumulation of TDP-43, as seen in 95% of ALS patients, have also begun to be used for drug screening. Questions still remain about how valid transgenic mice are as a model of ALS, but the obvious advantage is that hundreds of drugs can be screened with no risk to patients providing considerable savings in expenditure and time.

Conclusion

Charcot was the first to attempt to answer the question 'What is motor neuron disease?'. His observations helped to define the essential pathological clinical and morphological features of motor neuron degeneration. The molecular basis of these abnormalities has been characterized, and neurofilamentous, ubiquitinated, and other neuronal inclusions provide a pathological hallmark for the diagnosis of ALS. Recent advances in molecular genetics and cell biology have transformed hypotheses of causation based on speculation to those based on fact. Now, 135 years after Charcot, we have identified a number of genetic causes of ALS (mutant *SOD1*, *TDP-43*, *FUS*, *C9orf72*, *SQSTM1*, *VCP*, *UBQLN2*, and others), found a unifying pathological feature (TDP-43 accumulation) and have a modestly effective treatment (riluzole). As we fit together more pieces of the jigsaw puzzle we may soon be able to translate insights into the pathogenesis of ALS into effective treatments. The very fact that the therapeutic door has been prised open gives researchers, clinicians, and those living with ALS hope that drugs capable of slowing down or halting disease progression will soon be developed.

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Chapter 2

Palliative care

David Oliver

Summary

Palliative care aims to improve the quality of life of both the person with ALS and their family by the careful, multidisciplinary assessment of the problems that are experienced—physical, psychosocial, and spiritual. This care should be provided by all health and social care professionals but there may be a need for more specialist services to be involved with the more difficult issues.

Case history

Mr S was a 45-year-old man cared for by his wife, who had two children from her earlier marriage and a 4-year-old daughter from this relationship. He had been a very active man, working as a hairdresser until he had developed weakness of his hands and then legs as a result of ALS. As he deteriorated he became restricted to a wheelchair and communication was very difficult. His wife found it increasingly hard to care for him as well as the family, and he was admitted to a hospice for a period of assessment. During this time his wife expressed her concerns that she could not cope if he came home again. After much discussion and among the family and staff it was agreed that he would return home, with increased support and regular readmission for respite care. Communication was helped by new communication aids, following assessment by the speech and language therapist, and his drooling was eased by regular anticholinergic medication. His wife and children received regular support from the social work and nursing teams. The children were involved and supported as he deteriorated, and after his death at home there was ongoing support for the funeral and in bereavement.

Palliative care is defined as:

> An approach that improves the quality of life of patients and their families facing problems associated with life-threatening illness, through the prevention and relief of suffering, early identification and impeccable assessment, and treatment of pain and other problems, physical, psychosocial, and spiritual.[1]

The only treatment that can currently be offered for people with ALS will, at best, delay the progression of the disease process and is not curative, so it can be argued that the mainstay of the care of these patients is palliative from the time of diagnosis.[2]

The aim of palliative care is to look at the 'whole patient' in the context of their social support system, which is usually their family. This holistic approach is crucial in the care of someone with ALS and is relevant from the time of investigation, before the diagnosis has been confirmed, and throughout the progression of the disease.

It is important to stress that palliative care:

- affirms life and regards dying as a normal process
- neither hastens or postpones death
- provides relief from pain and other distressing symptoms
- integrates the psychological and spiritual aspects of patient care
- offers a support system to help the family cope during their relative's illness and in their bereavement
- uses a team approach to address the needs of patients and their families, including bereavement counselling, if indicated
- enhances quality of life, and may also positively influence the course of the illness.[1]

In the past there have been many debates, especially in the care of cancer patients, about the differing roles of curative and palliative treatment. It has been suggested that curative treatment should be continued until no further benefit can be obtained and at this point palliative care should be instituted. The timing of this sudden switch in the care of the patient can be very variable and may occur very late in the disease process, when death is imminent; this may deny the patient supportive care, such as the control of symptoms or psychosocial care. There is now a greater awareness of the need for an integrated approach to the care of a patient with a potentially incurable disease, particularly when the trajectory of disease progression may be very variable and uncertain. Research in the United States has shown that early involvement of palliative care support not only improved the quality of life of patients with lung cancer but also extended their prognosis.[3]

Within neurology there is an increasing appreciation that palliative care is appropriate for patients with certain neurological disorders. The Ethics and Humanities Subcommittee of the American Academy of Neurology (AAN) has stated that 'neurologists understand and apply the principles of palliative care'.[4] ALS is seen as one of the progressive and incurable neuromuscular diseases for which this approach may be appropriate and 'optimal medical care depends on determining the most appropriate means of achieving those goals for each patient'.[4] Studies have shown that there has been an improvement in the care of people with ALS, and over 90% are able to die peacefully.[5,6]

However, there is often a reluctance to accept the progressive nature of the disease, and patients and their families receive over-optimistic ideas concerning its nature and progression. Doctors have little training in the breaking of bad news and may find it difficult to provide patients with this information.[7,8] As a result the care received by patient and family may be less than optimal, with symptoms remaining unrelieved and psychosocial concerns never addressed or only partly recognized until near to death, when the possibilities of resolution are limited. As ALS is a disease of progressive loss and increasing disability it is essential that all aspects of care are addressed as early as possible:

- physical aspects, such as the control of symptoms
- psychological aspects, such as the fears and concerns about the disease
- social aspects, involving families and those close to the patient
- spiritual aspects, the areas concerning the meaning of life and the fears for the future.

It is very difficult to address the more profound concerns of the patient if communication is limited due to the loss of speech. Earlier intervention, when the patient is more easily understood and communication aids are not necessary, and when cognitive change is less likely to be present, allows better interaction and communication.

There may be barriers to the provision of palliative care—and these may be from the healthcare professionals and/or the patient and family.[2] Many professionals still see their role as to cure and alleviate disease, whereas in reality the majority of patient care involves the palliation of symptoms and of disability. All healthcare professionals need to accept the limitations of the care that can be given and approach patients and their families in a positive, but realistic, way. This particularly applies to the care of people with ALS, who face continual losses as the disease progresses and may require a great deal of help in coping with these losses and the ensuing changes in their lifestyle.

The negative attitudes of some healthcare professionals may profoundly influence the patient and family, and these attitudes may be difficult to change—as they are often related to discomfort in dealing with dying patients, lack of acceptance of the inevitability of symptoms, disengagement when 'active' treatment is not possible, and ignorance about palliative care.[2] There is a need for greater education in the principles of palliative care for all health professionals and awareness of the symptomatic treatments that can be offered to people with ALS.[9] Within Europe, the European Association for Palliative Care (EAPC) and the European Federation of Neurological Societies (EFNS) have been looking at a consensus paper on the palliative care of neurological patients and a core

curriculum for both neurologists and palliative care professionals, so as to achieve a greater awareness and understanding.[10] All too often palliative care is seen as applying only to the final stages of the disease, whereas end-of-life discussions and care will start at an earlier stage and early intervention may alleviate physical and psychological distress throughout the progress of the disease.[2]

Many people with ALS will require more specialized help. This may be provided by a specialist neurological team, a disability or rehabilitation team, or specialist palliative care services. Whichever model is used, a multidisciplinary team approach is essential, with the many different disciplines working collaboratively to maximize the care offered to the patient and family. There may be limitations as to team involvement in the care; for instance in the United States specialist palliative care services only receive Medicare funding for the last 6 months of life, although this can be extended. However, the need for specialized multidisciplinary care extends throughout the disease process, maybe for many years. It is essential that the same palliative care principles are provided by all the services throughout the patient's care pathway.

Specialist palliative care

In the UK many specialist palliative care providers are involved in the care of people with ALS. A survey in 2003 showed that at least 90% of inpatient units were involved in the care of this patient group.[11] The involvement varied, with units providing care:

- throughout the disease, from the time of diagnosis
- respite care as the disability progressed
- end-of-life care, in the final weeks or days of life.

However, the units that responded appeared to be involved later in the disease process and involvement from the time of diagnosis was unusual—only 8% of the responding units. The majority of units were involved later—for the palliation of symptoms (88%) or in the terminal phase (32%).[12] This can hinder the provision of palliative care because the team only meet a person for the first time when there are more severe symptoms and often restricted communication.

The specialist palliative care services may provide care and support in different ways:

- at home, with the support of the multidisciplinary team, in collaboration with the general practitioner (family physician) and community nurses;
- in an inpatient palliative care unit or hospice, for symptom control, respite care, or end-of-life care;

- in a day hospice, providing care for the day, allowing respite for carers and the opportunity for multidisciplinary assessment, involvement in rehabilitation and other activities, complementary therapies, and socialization;
- in hospital, with a palliative care team providing advice and support to the patient and family and healthcare professionals;
- specialist psychosocial care from social workers, counsellors, and psychologists;
- care after the patient's death, with bereavement support and counselling for families.

This specialist palliative care follows the patient, and the aim should be to provide a seamless service, wherever the patient is at that particular time. The specialist nature of the care is ensured by the close involvement of the multidisciplinary team, including:

- medical practitioner, in particular a consultant in palliative medicine
- nursing specialist
- social worker
- speech and language therapist
- occupational therapist
- physiotherapist
- chaplain and others trained in providing spiritual care and support
- clinical psychologist
- dietician
- pharmacist
- complementary therapists, e.g. providing aromatherapy or massage.

All members of the team should be working primarily within specialist palliative care, have been given training, and be receiving ongoing support and training in this area of expertise. There is a need for a collaborative approach with other teams involved in the care of a person with ALS—this can be rehabilitation, gastroenterology, or respiratory services, for example. There can be confusion and misunderstanding if this collaboration is not encouraged. A 'key worker' or 'key team' approach, where one team or person takes on the role of ensuring a coordinated approach, may be helpful.[13] It is always important to ensure that the patient and family, and all teams involved, are clear as to who is leading and taking responsibility for the care provided and ensuring that decisions are clearly made and communicated to everyone involved.[14]

Specialist palliative care aims to look at the positive aspects of a person's life and abilities and to enable patients and their families to remain as active as possible. For many people, including healthcare professionals, a hospice may be viewed merely in terms of end-of-life care and is sometimes perceived very negatively.

Several studies have shown that the needs of patients with ALS can be met by hospices and specialist palliative care providers.[6,12,15,16] O'Brien et al.[17] found that only 15% of patients had been referred for symptom control, although many had uncontrolled symptoms which could benefit from the multidisciplinary assessment of the palliative care team. It has been suggested that the involvement of specialist palliative care services may be involved in a more episodic way—helping and supporting the patient, family, and other healthcare professionals at certain points during disease progression when there are increased needs for specialist help—for instance at the time of diagnosis, consideration for gastrostomy placement, discussions about ventilator support—in the later stages of the disease and at the end of life.[18]

End-of-life care

Many patients and families, and often even health and social care professionals, fear the final stages of disease progression. There are fears of choking, breathlessness, and pain but although many symptoms may be experienced by the person with ALS during disease progression they can be managed effectively, and choking to death and severe distress at the end of life are very rare.[6,17]

The recognition of the final months or weeks of life can be difficult in progressive neurological diseases such as ALS, as there is great individual variation and the transition to end-of-life care may be gradual and insidious and less well recognized. However, it can be very important to recognize that a patient is reaching the end of their life, so that the emphasis of care is towards ensuring their comfort and the support of patient, family, and carers, i.e. health and social care. Triggers have been suggested that may allow the end-of-life phase to be considered for all neurological diseases:

- patient request
- family request
- dysphagia
- cognitive decline
- dyspnoea

- repeated infection—in particular aspiration pneumonia
- weight loss
- marked decline in condition
- significant complex symptoms—such as pain, spasticity, nausea, or psychosocial or spiritual issues.[13]

Specific triggers have also been suggested for ALS:

- respiratory failure or increased breathlessness
- reduced mobility
- dysphagia.[13,19]

These triggers may allow teams involved in the support of a person with ALS to recognize that a deterioration has occurred and that the emphasis of care may need to more specifically address the end-of-life issues—such as advance care planning, the provision and availability of medication to cope with any distressing symptoms, and discussion amongst the patient, family and team about the goals of care. Initial studies have suggested that these triggers have validity and that the number of triggers increases as death approaches.[20]

There are challenges with this approach, as patients and families may not wish to discuss issues about the end of life; sometimes as they do not wish to face the reality of this deterioration or health and social care professionals involved in their care may lack the experience or willingness to recognize the triggers and discuss the issues—maybe due to their previous close involvement with the patient or family or their own personal views of care. However, the recognition of end of life, and the possibility of ensuing discussion, may allow patients and families the opportunities to discuss their wishes, make plans, and undertake the activities they wish.

Ethical dilemmas

In the care of a person with ALS there may be many ethical dilemmas to be faced and difficult decisions to be made.[21] There are decisions to be made about treatment that can alter the life of the patient. As treatments such as riluzole are developed to slow the progression of the disease process, there will be increasing ethical debates on the effectiveness and appropriate use of these new, and usually expensive, drugs. There are other decisions about treatment interventions, such as gastrostomy feeding or ventilatory support, that affect the prognosis and survival of the patient. The appropriateness of these treatment options will need careful discussion, and it may be very difficult to discuss such issues with

patients and families. Both the benefits and risks must be discussed. This may include discussion about the possibility of becoming 'locked in' and unable to move or communicate for someone contemplating ventilator support (see Chapter 6). In the UK, the guidelines produced by the National Institute for Health and Care Excellence (NICE) has suggested that these discussions should be undertaken when non-invasive ventilation is initially discussed, and particularly when it is instituted.[22] At the same time there needs to be a full assessment of the palliative care needs of the patient and family so that palliative care can continue alongside more active care. This often occurs in the care of patients with cancer, as there are needs for symptom control and psychosocial support while oncological treatments such as chemotherapy are continued. There is a need for close collaboration between neurological services and palliative care providers to ensure the patient is offered the maximum number of treatment possibilities.

Palliative care services can be involved in these processes and the need for accurate and responsible communication is very much part of the role of palliative care. Close collaboration is helpful, and some neurological centres are suggesting the involvement of palliative care services earlier in the disease process so that this collaboration can be fostered. It is suggested in some centres that all patients being considered for non-invasive ventilation should have contact with a specialist palliative care team so that the discussions about the future, and how to cope when ventilatory function deteriorates further, can be instituted by a local service who can support the patient at this time. This aims to overcome the risk of invasive ventilation being instituted in an emergency situation by a hospital team with little knowledge of the patient, when the patient and family have already discussed this issue earlier and decided not to be considered for ventilation[23] (see Chapter 6).

Patients with ALS face many of these issues and fears, particularly of pain and choking. Many influential textbooks and books on diseases still talk of death from ALS being distressing and due to choking, even though there is much evidence that with good symptom control this is unlikely.[6,17] In the UK there have been two court cases brought by people with ALS, including Annie Lindsell, who asked the courts to allow administration of medication when their distress was such that they no longer wished to live. After several court hearings it was felt that there was no case and the action was dropped. The arguments were based on fears of choking and respiratory distress, and as the cases had a high profile within the media many other patients with ALS have come to fear their own deaths. These fears continue to be reinforced in the press during the debates on assisted dying; the negative aspects of dying always seem to be emphasized and the possible supportive role of palliative care minimized.

Palliative care aims to help and support the patient and family and does not intend to shorten life. With the control of distress many patients feel more positive and life may even be extended. There may be rare occasions when the shortening of life may be a foreseeable consequence of treatment, but the intention is never to shorten life. On occasions patients may decide against procedures or treatments that could prolong their lives, for instance a feeding gastrostomy or ventilatory support. As long as the patient is able to make this decision clearly the healthcare team should provide support to the patient, and family, in making this decision and help to minimize distress by the provision of good palliative care.[3]

There may be increasing challenges with the discussion of these issues, because the prevalence of cognitive change appears to be greater than once was thought—with evidence of frontal lobe changes in up to 65% of people with ALS and dementia in up to 15% (see Chapter 8). The decisions regarding treatment options will need to be made earlier in the disease process while the person is competent, although at this time the shock of the diagnosis of ALS and coming to terms with all the losses resulting from the disease may make it difficult for a person with ALS to contemplate and discuss these issues. It may, however, be possible to discuss the person's preferences soon after diagnosis in a more general way, and in this way raise the issues. The use of the 'Ethics Questionnaire' or 'Patient Preference Tool' has been suggested[24,25] although some patients may decline these discussions—up to 30% in one study.[26] Speaking about these issues earlier may allow further discussion later and the development of an advance directive (in the United States and Europe) or an advance decision to refuse treatment (in the UK) which will clarify their wishes (see Chapters 5 and 18).

A person with ALS faces many challenges and many losses as the disease progresses. These affect the family and close carers as well, and the healthcare professionals involved with the care may also be affected. Palliative care provided by a multidisciplinary team, working collaboratively with all the other services involved in the care, can help the patient and family to continue to function as effectively as possible, with the aim of ensuring that the quality of life can be as good as possible.

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Chapter 3

Communication: breaking the news

Richard Sloan and Gian Domenico Borasio

Summary

Telling a patient that they have an incurable disease such as ALS is a highly-sensitive task with many potential pitfalls. Through the use of research-based communication skills, the burden on the messenger and distress to the patient can be minimized.

Case history

During a hospital admission, Michael had just been diagnosed with ALS. He noticed that his neurologist seemed in a rush to get away after telling him on the open ward, and the nurses seemed embarrassed. However, Michael didn't care much as he was relieved to be told that he could go home.

Not having heard of ALS before, he and his wife looked it up together on the Internet. The first phrase they saw was '. . . an inevitably fatal disease with an average life expectancy of 2–3 years from diagnosis'. It was a Friday evening and Michael's family physician wasn't around to talk about it until the Monday. Michael and his wife recall that weekend as the worst of their lives.

On the Monday morning, they booked an appointment to see their family physician. He hadn't yet received any information from the hospital and, having not seen a case of ALS since medical school, wasn't able to answer the many questions that Michael and his wife had. At a follow-up appointment, after several weeks, the neurologist painted a gloomy picture of increasing disability, breathing, and swallowing problems, leading to death. One phrase particularly stuck in Michael's mind—that, apart from a drug to slow the process down, the doctor said 'I am afraid there is nothing else we can do'. This completely devastated him. He felt as if he had been cast adrift to face an awful death full of suffering for him and his family.

Introduction

Giving the diagnosis to patients with ALS is a daunting task for any neurologist. Obviously, breaking the news about ALS is not a procedure that can be standardized. However, proven techniques exist to reduce the trauma to the patient and ease the burden on the doctor, thus reducing the risk of burnout and the

tendency to 'pull away' from the patient. Such communication skills are of fundamental importance to clinical practice and should be more prominent in medical teaching. The way the patient is told the diagnosis is now recognized to be the first and one of the most delicate steps in palliative care.

The task of telling a patient that they have ALS generally falls to the neurologist diagnosing it. Whilst the messenger can get blamed for bad news, there is anecdotal and research-based evidence that it is not always performed optimally.[1,2] Because of the poor prognosis there has been reluctance in the past to tell the patient the whole truth. Often, the patient was given 'reassuring' statements while the relatives were informed more fully, with the addendum 'there is nothing more that can be done for him'. This practice still exists, although most clinicians now respect the ethical principle of patient autonomy.

The majority of patients *do* want to have information on their condition, even if it is life-threatening.[3,4] Without this, the stress they suffer through uncertainty can often be worse than knowing they will die. One in five patients with cancer develop full-blown psychiatric disorders.[5] The main predictive factor for this is the way in which bad news is broken.[6] The feeling of hopelessness present on both sides when the diagnosis of ALS is disclosed should not lead to the withholding of information that might be essential for the patient's life-planning.[7] Breaking the news is an ongoing process throughout the course of the disease: it is not just limited to the communication of the diagnosis (which in itself is a multistep procedure), but encompasses all aspects of the flow of information from the physician to the patient regarding the disease process, thus including good as well as bad news. This chapter offers some suggestions on how to achieve this.

Background

Johnston and co-workers[2] interviewed 50 patients about their experiences of being told they had ALS. Most patients saw the providing of a label for their condition as a positive thing—'at least I now know what I have to contend with'. Negative aspects included being told too late—'I could have used the past year better if I had known it might have been my last'; being told the diagnosis without the option of having a relative present; lack of privacy (e.g. in an open ward); being told in vague or confusing terms; and too much detail all at once. Although there was no evidence that poor communication at the time of diagnosis was responsible for prolonged mood disturbance, patients may well have undergone unnecessary psychological trauma at the time they were told. A summary of the findings is shown in Table 3.1.

Lack of adequate communication skills also leads to stress in doctors.[8] Burnout is more likely amongst consultants who feel insufficiently trained in communication

Table 3.1 Communicating the diagnosis of ALS[2]

Most people see positive aspects on being told the diagnosis, especially as it provides a label for the condition
People prefer the diagnosis to be communicated in a direct, empathetic style
Being able to ask questions is important
Doctors should guard against giving too pessimistic a message all at once
Information on where to get further help (e.g. from a patients' association) is important at the time of diagnosis
People generally prefer to be told with someone else present

Source: Johnston M. et al. Communicating the diagnosis of motor neurone disease, *Palliative Medicine*, Volume 10, Issue 1, pp. 23–34, Copyright © 1996 by SAGE Publications, doi:10.1177/026921639601000105. Reprinted by Permission of SAGE.

Table 3.2 Why breaking bad news is difficult for doctors[11]

Fear of the messenger getting blamed for bad news
Perceived lack of time
Lack of training
Fear of causing distress
Fear of being asked difficult questions
Fear of not having all the answers
Invoking fears of one's own mortality

Source: data from Buckman R., *How to Break Bad News,* Papermac, London, UK, Copyright © 1996.

skills.[9] As the Health Service Commissioner for the UK[10] pointed out, improvement in communications between healthcare workers, patients, and relatives would result in a marked reduction in complaints. Table 3.2 outlines the reasons why breaking bad news is difficult.

What to tell

Minimum information

After the diagnosis is firmly established, the patient should be informed that they have a progressive disease of the motor nerves, for which no curative therapy is available. The name of the disease must be stated and explained to avoid confusion, for example, with multiple sclerosis. If the family history is negative, it is reassuring for patient and family to know that their children are unlikely to be at risk. Positive aspects (e.g. no pain, no disturbances in sensation, continence,

etc.) should be stressed, as well as the availability of effective palliative measures for practically all symptoms of ALS.[12] Current research efforts and, where available, the possibility of taking part in clinical studies of new drugs should be pointed out as a means to foster hope.

Beginning of the disease

Patients often suspect a connection between the onset of the disease and a specific event (e.g. an accident, operation, private or professional crisis, etc.). This question should be addressed, and patients should be informed that the disease starts many years if not decades before the first symptoms appear. Since at least 50% of the spinal motor neurons have to degenerate before clinical symptoms appear, it is impossible to establish when the disease began in any single patient, even with a large degree of approximation.

Prognosis

Many ALS patients ask directly about their remaining lifespan since, at the time of diagnosis, they have usually experienced the progression of the disease for several months if not years. The answer to this question should include the information that there are no sudden relapses, that periods of relative stabilization may occur, and that rare remissions have been observed. The patients should be told that the course of ALS may vary between months and decades (e.g. Stephen Hawking), making a firm statement on prognosis all but impossible for any single patient. In our experience, many patients appreciate a rough approximation, for example, 'I think you have probably got several months rather than several years left'. This allows them to plan the rest of their lives and is more helpful than an outright refusal to speculate. However, if a patient explicitly asks for available statistics, they have the right to know. Sensitive questioning about suicidal thoughts may be advisable if the patient appears depressed. Such acknowledgement reduces rather than increases suicidal attempts.

Available therapies

Riluzole, the first substance with a modest life-prolonging effect in ALS, has been on the market since 1996.[13] Subject to the availability of reimbursement for this drug, an open discussion about its pros and cons should take place. Patients must be told that whilst the drug may slow down future progression, they will not notice any improvements in function due to riluzole. The possible side-effects (nausea, asthenia, dizziness) should be mentioned. This discussion avoids raising false hopes, which might otherwise eventually lead to disillusionment and frustration.

Unconventional treatments

It is all too understandable that patients with ALS start looking for help outside the boundaries of 'classical' medicine. This topic should therefore be addressed the first time the diagnosis is given. Patients should be reassured that their physician does not object to their trying homeopathic medication, acupuncture, etc., if they so wish. However, patients and families must be warned that some alternative methods (e.g. so-called 'stem cell therapy' outside rigorous clinical studies, snake toxins, etc.) may be dangerous to their health, while others may entail serious financial consequences to the benefit of charlatans and common criminals (including, sadly, some physicians). A reliable website for patients and professionals which gives objective descriptions of results of past clinical trials is <http://www.alsuntangled.com>.

Patients' associations

Patients can obtain invaluable help (indeed, more than their doctors can usually offer) from the ALS patients' associations, which are present in almost every major country in the world and are grouped together in the Alliance of ALS/MND associations (a list with addresses and contact details can be found at <http://www.alsmndalliance.org>). It is therefore essential to inform patients and families from the beginning about the existence of these associations, provide them with the relevant addresses and telephone numbers, and explicitly encourage them to get in touch.

Second opinion

Many patients, when confronted with a diagnosis like ALS, will want a second opinion. Again, this is more than understandable and should be accommodated by the physician who is breaking the news. Possible tertiary referral centres should be discussed with the patient. This can strengthen the doctor–patient relationship and help to avoid the patient self-referring from one doctor to another.

If the diagnosis is only suspected

What has been said so far presupposes an unequivocal clinical diagnosis of ALS, i.e. 'probable' or 'definite' ALS according to the El Escorial criteria of the World Federation of Neurology.[14] If these criteria are not met, the physician will obviously take a more careful approach, depending on the degree of clinical certainty. The offer of information on possible differential diagnoses and the diagnostic plan can reduce the stress of not being told anything. However, if the El Escorial criteria for 'possible' ALS are fulfilled, information on appropriate

palliative measures, available drug options, and therapeutic studies should be considered as these patients rarely turn out to have something else.

Discussing mechanical ventilation and the terminal phase

For many people with ALS, the onset of dyspnoeic symptoms marks a turning point during the course of their disease. Often, the first dyspnoeic bouts happen after choking on food, on exertion, or during sleep. At the beginning of clinically manifest involvement of the respiratory muscles, anxiety is usually the most prominent symptom during dyspnoeic attacks (which therefore respond very well to short-acting benzodiazepines, e.g. lorazepam 0.5–1 mg sublingually). If the subject has not been discussed previously, the following factors should be the trigger for the offer to talk to the patient about the terminal phase of the disease: (1) when the first dyspnoeic symptoms appear; (2) symptoms of chronic nocturnal hypoventilation develop; (3) a rapid drop in forced vital capacity (FVC) below 50% occurs.[15] The reason for this recommendation is that almost all patients, when questioned at this stage, report fears of 'choking to death'. In our experience, describing the physiological mechanism leading to terminal hypercapnic coma and the resulting peaceful death in sleep will relieve these fears in most cases. At this point, assisted ventilation, although not wanted or tolerated by everyone, should be described (see Chapter 6). Patients and relatives should also be informed that the array of medications available for breathlessness, if correctly applied, do prevent suffering.[16] This information needs to be reiterated in subsequent visits.

At this stage, patients should be asked whether they would wish to be intubated and ventilated in the event of terminal respiratory insufficiency. Patients who have been informed about the possible subsequent clinical course, which may in the end manifest in 'locked-in' syndrome on an intensive care unit,[17] will usually decline such a procedure. This refusal must be documented by the physician and is often best incorporated into an advance directive (see Chapter 5). The consequences of such a decision must be discussed with the patient, their family, and the family physician (e.g. concerning the use of medications in the terminal phase for relieving distressing physical or psychological symptoms).[18]

How to break the news

The factors to be considered when breaking bad news were given by Maguire et al.[19] (outlined in Table 3.3) and will now be discussed in detail. They defined three underlying principles in the process:

1 Do not withhold information if the patient wants it.
2 Do not impose information if the patient does *not* want it.
3 Gauge and respond to the patient's reaction to the news.

The setting

In some countries, for example the United States, diagnostic procedures for ALS are usually performed in an outpatient setting, while in others (e.g. Germany and the UK), patients are usually referred to a neurology department for inpatient work-up once ALS is suspected. The latter procedure gives the physician the possibility of a stepwise offering of information over several days, thereby allowing the patient to reflect overnight and ask additional questions. If the diagnosis is told in an outpatient setting, a short-term follow-up visit should be scheduled.

Privacy is essential, so that both doctor and patient can feel free to discuss difficult issues without feeling self-conscious that others are listening in. Disclosure of personal medical details on the open ward is a potential breach of confidentiality and is unacceptable. Even if the patient is immobile, with planning, it should be possible to transfer them to a quiet room. There should be no interruptions. The phone should be off the hook and bleeps handed to someone else for taking messages. The manner in which the news is broken is just as important as the words used.[20] Warmth, eye contact, empathy, lack of medical jargon, and above all sufficient time for expressing their own thoughts, anxieties, and questions are considered important by patients.

Patients often say that they would have liked to have had the option of having someone else with them when the news was broken, usually their closest

Table 3.3 The stages in breaking bad news[19]

Setting
Finding out what the patient already knows or suspects
Finding out how much more they want to know
Firing the warning shot
Providing information in stages (the hierarchy of euphemisms)
Acknowledging and responding to the patient's reaction to the news
Contract for the future
Reinforcement of information

Source: data from Maguire P. and Falkner A., Communicating with cancer patients: 2 – handling uncertainty, collusion and denial, *British Medical Journal*, Volume 297, Issue 6654, pp. 907–9, Copyright © 1988 BMJ Publishing Group Ltd, doi: http://dx.doi.org/10.1136/bmj.297.6654.972

relative or a nurse. This may be helpful for picking up the pieces afterwards. The patient can also check with the other person details they may not have taken in fully the first time. When it is known that results of investigations are likely to be available by a certain day, the patient should be asked in advance if they would like someone else there with them.

Roger Carus, a patient who wrote about his experience of being diagnosed with ALS in the mid-1970s, found out inadvertently whilst in hospital for investigations. He was not sure whether or not his wife had been told the diagnosis by his neurologist and recalls 'I spent two of the most agonising weeks of my life trying to find out whether she knew or not'.[1]

Cross-cultural differences should be recognized and taken into account when delivering the diagnosis.[21] In some countries, such as Japan, it is customary for the patient to delegate any decision concerning, for example, end-of-life issues to the family and the physician, although a trend towards more open discussion has taken place in recent years.[22]

Finding out what the patient knows or suspects

To launch straight into an explanation of the diagnosis can be disastrous. Finding out what the patient understands so far helps gauge the pace of the consultation and where the patient is starting from. Some patients may genuinely have no idea that there is anything seriously wrong. Others may be terrified that they are going to die the following week. 'What have you made of things so far?' is a good opening question.

Finding out how much more they want to know

Although the majority will want more information, a small minority would be devastated if told bad news at any stage. Say something like: 'Some people wish to know everything about their health and others just what the doctor's treatment plan is. Which group do you fall into?'. Those who habitually cope by using denial will relate to the latter and be spared unnecessary distress.

Firing a warning shot

Where the patient is asking for more information, it is still important to signal that difficult news is coming before disclosing it. This lessens the blow for those who are still relatively unsuspecting. 'I'm afraid that the results of your investigations are not what I was hoping for'. There should then be a pause during which the doctor gauges the patient's reaction. If they look shocked, ask what is going through their mind. If in doubt as to whether to continue, ask whether they want more information or more time to think about it. If the patient says 'What do you mean, not what you were hoping for?', this is a clear request for more information and it is right to proceed.

Providing information in stages

It is better to give the patient small pieces of information at a time and check whether they have understood before going on to the next bit of detail—'Does this make sense to you?' or 'Do you want me to go over that again?'. To conduct a monologue only to find out that the patient froze after the first sentence is a waste of everyone's time. Again, if the patient looks shocked, ask what is on their mind and check out whether they are ready to go on or not.

A good technique for delivering information stepwise is known as the 'hierarchy of euphemisms': small pieces of information of increasing seriousness are told to the patient, with pauses in between for the patient to digest them and the physician to judge by the patient's verbal and non-verbal reactions if they want to continue or leave it there for the time being. This avoids extreme distress and the patient 'switching off' to any further explanation through shock. For example: 'The tests we have performed show you have a problem with the nerves which transmit instructions to the muscles'; 'The nerves are gradually being destroyed so that your muscles can't work properly'; 'The condition you have got is called amyotrophic lateral sclerosis, or motor neurone disease'; 'Unfortunately, there is no known cure at the moment, although there is a drug (riluzole) which modestly slows the deterioration down'; 'Yes, the disease is eventually fatal'.

Acknowledging and responding to the patient's reaction to the news

In order not to impose information that the patient is not yet ready for, the doctor pauses at each step to gauge whether they are ready for further information. If the patient looks shocked or cries, the messenger's response is crucial. Carrying on through embarrassment will appear uncaring to the patient who will be unable to take anything more in anyway. 'This must be shocking news' acknowledges that this is an understandable reaction in the circumstances. Try to find out exactly what the source of their distress is—'Can you bear to tell me what's going through your mind right now?'. This allows them to decline if it is too difficult to talk about. The answer may seem obvious, but will be different for different people, based on previous concepts and experiences. It is an opportunity to address misunderstandings. If the patient is expressing undue pessimism, such as thinking they are going to die next week, appropriate reassurance can be given.

Contract for the future

It is important at the end of the consultation to make specific arrangements for the patient to be seen again and have the opportunity for clarification or to ask more. Where there is no definitive medical treatment, it is tempting to tell the

patient they don't need to come back again, but this can be very frightening. Offering to see the patient again yourself conveys to them the feeling that they are being supported, not abandoned. A pilot study suggested it is possible for experienced clinicians to identify patients who may struggle to cope with a diagnosis of ALS early by conducting a patient-led interview.[23]

Phrases like 'I'm afraid there's nothing more we can do' leave patients devastated, presuming falsely that they are condemned to intractable suffering. We have found it helpful to demonstrate concern for the patient by informing them of sources of help and support as already mentioned (see Patients' associations), and arranging a short-term follow-up meeting. Further exploration of the emotional impact of the diagnosis can be provided by a specialist member of staff, such as a social worker, either straight away or a few days later in the patient's own home.[24] Often, patients tend to forget information, even if they appear to have fully understood it when they were first told. It is therefore important, on each clinic visit, to check again where the patient stands and to move on jointly from there.

Reinforcement of information

Evidence from the cancer literature suggests that patients might profit from being handed out an audiotape of the consultation or receiving a letter outlining the consultation in lay terms, with a preference for the former.[25] This approach has not yet been tested in patients with ALS. Data from a small sample[26] show that a large proportion of patients and relatives felt that they received no or insufficient information from their physician at the time of diagnosis. A brochure containing general information on ALS in lay terms was regarded to be informative by almost all patients and relatives. Therefore, handing out written information after the consultation will be helpful in most cases. However, it might be advisable to ask patient and relatives whether they feel that they are ready for it or whether they would prefer to receive the material at a later stage.

Collusion

Collusion is defined as a secret agreement to conspire to deceive. In health care, it usually takes the form of a relative asking the doctor or nurse not to tell the patient they have a serious illness or a poor prognosis. Ethically, this is unjustifiable as patients have the right to know information about themselves. Alternatively, patients may ask professionals not to tell relatives or children of their illness. Although this too may eventually cause problems, by contrast it is legally imperative, as patients have the sole right to decide who has access to confidential medical information about themselves.[27]

The colluder's motive is usually the protection of loved ones from distressing news. This instinctive action, well meaning though it is, usually results in more distress in the long run—both for the patient who may become isolated and anxious on becoming less well, and for the colluder, as secrets become harder to maintain in the face of that deterioration.[26] If not recognized and addressed, the situation also becomes more stressful for the professionals involved. They become torn between the duty to be truthful to the patient and incurring the wrath of the colluder. It is easy for an unsuspecting doctor or nurse, unaware of the collusion, to innocently disclose information and be blamed by everyone.

Often, because of lack of training, professionals don't confront the collusion and everyone ultimately suffers the consequences. There are, however, guidelines to prevent the situation escalating.[19] They are based on two fundamental ethical premises:

- patients have the right to medical information about themselves if they want it and
- patients have the right to decline medical information about themselves if they do not want it.

The guidelines aid negotiation with the colluder to find out which category the patient is in. Because the protective instincts of the colluder are very strong, it is crucial to first understand and openly acknowledge why they are doing it. This prevents a confrontation developing and wins them over so that you can then negotiate to check with the patient if there is anything else they wish to know. The colluders can then be helped to see the potential downsides of their well-meaning actions.

Guidelines for dealing with collusion

1. Ascertain from the colluder why they have chosen to do it and acknowledge their reasons—if they fear the patient going to pieces, do they have any evidence from the past that this might indeed happen? Where there is justified concern, the doctor would rightly be more cautious in his or her subsequent dialogue with the patient.
2. Find out the cost to the colluder of the collusion—colluders may not acknowledge the stress on themselves or the strain on their relationship with the patient: 'How have things changed between you since he got his illness?'; 'How are you sleeping?'.
3. Discuss the downside to collusion for patient and colluder—although relatively easy to maintain at first, collusion usually becomes more and more stressful with time.

4 Negotiate with the colluder to see the patient on your own to find out how they are feeling about their situation and whether there is anything else they wish to know—promise that you won't force information on them if they don't ask for it. However, most patients do want to know in time and, if asked, you would give honest answers.

5 Interview the patient alone. If they are in denial and do not want any more information, allow them to maintain this coping mechanism, saying that they can always ask anything in the future if they have any concerns. More often, they will appreciate the opportunity to find out more, but may have been keeping quiet because of the imposed block to communication—colluding with the collusion. Many patients will have worked out for themselves that their illness is a serious one by the awkwardness of family and professionals. Patients often express relief when things are finally out in the open. Offer to talk with them and their family to share what has just been discussed.

6 Talk with the patient and family together. This models open communication within the family and gets over the fear of how to break the ice.

The concept of hope

When faced with adversity, people often avoid going to pieces by holding out hope for better days. In the case of serious illness, hope for a cure may persist, even when this is highly unlikely. Professionals should be wary of killing all hope—this may remove the patient's main coping mechanism and risks rejection of the messenger. Nor should professionals be over-optimistic either as this jeopardizes trust when reassurances turn out to be false.

The task for the healthcare professional is to help the patient adjust their goals realistically to their situation at a pace which is acceptable to them. This may mean consoling the person who is finally realizing that they are never going to regain full functional capacity whilst giving them realistic hope that mobility can still be maintained to a lesser degree in a wheelchair.

For those coming to realize their mortality from a neurological disease, the pledging of continuing support whatever happens is important in maintaining hope for a comfortable, peaceful death.

Conclusion

Whilst breaking the news in ALS will never be easy, proven techniques exist to reduce the trauma to the patient and ease the burden on the doctor. Such communication skills are of fundamental importance to clinical practice, and should be given as much prominence as clinical skills in both undergraduate

and postgraduate teaching.[28] The way the patient is told the diagnosis is now recognized to be the first and one of the most important steps in palliative care.[29] There is increasing research evidence about optimal communication skills, how to acquire and retain them, and the beneficial effects they have on the psychological well-being of patients.[30–32] To quote from Caplan:[33]

> The increased number of diagnostic and treatment options makes even more crucial the physician's skill in managing illness and the art of communicating with patients and their loved ones. There but for the grace of God go we all, for all of us and our families are, or will eventually become, patients.

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Chapter 4

Decision-making

Wendy Johnston

Introduction

Prognosis and quality of life have steadily improved for people with ALS. Still, the lack of a cure and the anticipated progression of the disease mean that decisions about the tenor of care, including its intensity and location, and accepting or forgoing life-sustaining therapies are required throughout its course[1] Management of ALS requires the development of individualized advance care plans, particularly to address specific decisions about respiratory and nutritional support. Requests to hasten death are common. The medical, psychosocial, and spiritual context for each individual and family as well as medical, community, and financial resources will influence both the decisions and their implementation.

Case history

Maud, an 86-year-old widow, was diagnosed with bulbar ALS after experiencing a rapid loss of speech and swallowing. She accepted a percutaneous feeding tube with the aim of continuing to reside in her own home. None of her family lived in the same city, but given the choice of relocating early or seeking support to continue in her own home, Maud decided to stay. She remained independent with minimal home care for a year, accepting nocturnal non-invasive ventilation (NIV), when progressive weakness of her limbs compromised her independence. With family and home care support she remained at home until she could no longer perform her self-care. She discontinued NIV in favour of opioids and benzodiazepines for management of dyspnoea. Now bed bound, with little appetite, she requested to stop nutrition and hydration. Comfortable, and at peace, she became impatient that death did not come. When asked specifically about a desire to have physician aid in dying, she said wished to die naturally and was 'simply venting her frustration'. As her care became more than home health and family could support, she accepted admission to an inpatient hospice where she died peacefully a week later.

What are 'decisions for the end of life'?

Any decision to limit or forgo a life-sustaining intervention, to accept symptom relief over life-prolonging therapy, or to hasten death is a decision for the

end of life (EOL). Specific goals may include completion of tasks, reconciliation of conflicts, or determining a legacy. The preferred location of death and the presence of loved ones are important considerations. Planning for EOL necessitates an understanding at some level that death, while perhaps not imminent, is the likely outcome of disease progression. Decisions may be disease specific, for example whether to forgo mechanical ventilation via tracheostomy in ALS, or value driven, such as the desire to die at home. Each decision will be subject to variables particular to the individual and their milieu.[1] A recent structured review of the literature on decision-making in chronic neurological diseases concluded that there is evidence that involving patients in treatment considerations throughout the disease course can positively influence the quality of EOL.[2]

Clinicians treating ALS must also make decisions. They must acknowledge that their own values and traditions may influence the presentation of information for decision-making. Interventions recommended in published guidelines are subject to local limitations and pressures. Conflict may arise when values clash. Clinicians wish to do no harm, and fear censure and legal difficulties related to the provision of EOL care. While death and dying are not ethical issues per se, the ethics of withdrawing and withholding care, and the use of medications that putatively hasten death, are unclear to many practitioners.

What are advance directives, and when should advance directives be prepared?

An advance directive (AD) is the patient's documented preferences for the type and timing of medical interventions (discussed in further detail in Chapter 5). Generic ADs would appear to be of limited utility, and major studies have documented that ADs are frequently overridden or ignored. However, disease-specific, detailed ADs help guide physicians, especially in conjunction with ongoing discussions with the patient and designated proxy decision-maker.[3]

The timing of the discussion of EOL issues and advance care planning with patients and their families should balance their receptivity and the need to make timely decisions about life-sustaining therapies. The timing and manner of disclosure of the diagnosis can have a lasting impact on the patient and family.[4] Some studies suggest that early discussion of mechanical ventilation increases satisfaction and decreases remorse later on.[2] A survey of German neurologists and ALS patients found that both groups adopted a 'wait and see' approach to living wills, waiting for symptoms of respiratory failure to emerge

before completing a generic, not an ALS-specific, form.[5] Both neurologists and patients in this study perceived an AD as a harbinger of imminent death. The authors concluded that the goals of preparing an AD were not met in this cohort. However, both the American[6] and European[7] practice guidelines advocate the preparation of an AD as well as the designation of a proxy decision-maker. The content of the AD should be reviewed with the patient and proxy by the patient's medical practitioner at least every 6 months. In practice, while the ALS CARE database, largely reflecting major multidisciplinary clinics in the United States and Canada, showed that the majority utilized ADs and that ADs were respected,[8] a recent study from France showed that despite the majority requesting specific heath information, only 20% of ALS patients in a multidisciplinary clinic actually completed an AD.[9]

The ALS Peer Workgroup identified six triggers for discussion of EOL issues:[10]

1 request from the patient or family
2 severe psychological, social, or spiritual distress or suffering
3 pain requiring high doses of analgesic medication
4 dysphagia requiring a feeding tube
5 dyspnoea or symptoms of hypoventilation, or FVC of 50% or less
6 loss of function in two bodily regions.

EOL care should be addressed routinely as part of discussions about prognosis, or when considering interventions that could have a low probability of success. More urgent discussions should be prompted by concerns that death is imminent (as perceived by the physician, patient, or family) or that disease progression has been severe. An expressed desire to die, queries about assisted suicide, and interest in hospice or palliative care must be dealt with immediately. Severe suffering is an emergency that demands medical intervention.[11]

The failure to address advance care planning leads to unplanned interventions, particularly mechanical ventilation.[12,13] Overall, the use of tracheostomy and mechanical ventilation is low in the United States at 4% or less.[13–15] Despite NIV being recommended for symptomatic management of respiratory failure, its use remains low[14] (see Chapter 6). This could reflect a lag in knowledge, but could also reflect the values and attitudes of the medical practitioners providing the care. Variability in rates of mechanical ventilation in ALS patients in Illinois, USA was related to attitudes of physicians in the medical centre where the patient was managed.[13] Physicians 'frame' the discussion of mechanical ventilation in chronic lung diseases in either a positive or a negative way[16] depending on their perception of the patient's quality of life and the potential reversibility of respiratory failure. This has not been evaluated for ALS patients,

but physicians' own attitudes to NIV and long-term ventilation may be the main determinants of the likelihood of their use. Variability in the use of riluzole in different centres as reported in the ALS CARE database probably reflects framing of the information presented for ostensibly patient-centred decision-making.[14]

Patient autonomy in decision-making can only be assured when the available information is presented fully, but neutrally. Funding and medical support, as well as family and community support, must be understood. Advance care planning should be firmly grounded in the values of the individual, who in turn should understand the consequences of the decisions, both for themselves and their family. The individual's values may not reflect the mainstream; exploring the spiritual and cultural values of a person with ALS and their family should be integral to the decision-making process, and should be established before crises occur. When cultural differences appear to preclude patient-centred decision-making, or appear at odds with the values of the clinicians, consultation with an institutional ethics committee, community leaders, or spiritual counsellors of the individual may resolve potential conflict.

How do cognitive dysfunction and depression affect the decision-making capacity of ALS patients?

Competence, also called decision-making capacity, reflects the ability to make autonomous choices. The central abilities required are that the patient must be able to:

- communicate a choice
- understand the relevant information
- appreciate the situation and its consequences
- manipulate the information rationally.

Communication itself is a challenge in ALS. Any necessary alternative and augmented communication techniques can be time-consuming. In addition, communication about preferences for treatment is not an event but a process, and preferences may change as the patient experiences progression of disease.[17] Patients must understand the relevant information, i.e. the risks, benefits, and burdens of alternative courses of treatments available, including the risk of death. Patients 'appreciate' their situation when they can apply information to their personal circumstances. For example, depressed patients may not be able to appreciate the benefits of treatment if they are too hopeless to imagine that an intervention such as a gastrostomy tube might improve the quality of their life.[18] The reasoning behind the decision must be logical and understandable, even

when the clinician does not agree with it. Impairments in appreciation and rationality are most often found in patients with major psychiatric disorders such as psychosis, mania, or depression.[19]

Unlike patients with other terminal illnesses, ALS patients often retain decision-making capacity up until the final hours of life. However, intact decision-making abilities cannot be assumed. Delirium, depression, and cognitive impairments are the most common reasons for a lack in decision-making capacity. Delirium is found in up to 90% of cancer patients in hospices during the final weeks of life and universally interferes with decision-making capacity. Family caregivers of 50 ALS patients who had died reported that 26% were confused in the final month of life.[35] Among ALS patients, dehydration, hypercarbia, infections, and other organ system dysfunctions may all contribute to delirium. Medications commonly used at the EOL that may cause delirium include anticholinergics, benzodiazepines, and opioids. Ironically, at times, patients whose wishes are not known will require ventilation or hydration in order to recover capacity to make these decisions.

Cognitive impairment and even frank dementia may occur in ALS (see Chapter 8). In some studies, impairment in executive function and other frontal lobe-mediated domains was found in half of the study participants.[20] The frontal lobe is especially important in the ability to flexibly consider and weigh alternatives, apply one's values and goals, and appreciate relevant information. In ALS frontal dysfunction appears progressive, underscoring the importance of beginning discussions about values and goals early in the course of the illness.

Estimates of the prevalence of depression in ALS vary,[21–23] and did not increase in a prospectively followed hospice group in the last months of life.[22] Symptoms of depression that have an impact on decision-making ability include hopelessness, pessimism, low self-esteem, and suicidal thoughts. While mild to moderate depression did not influence decisions, elderly, severely depressed patients, after effective treatment, were more likely to prefer life-sustaining treatments when presented with hypothetical scenarios of illness. Patients with severe depression who would decline treatment expressed less hope, overestimated the risks and burdens of treatment, and underestimated the benefits.[18]

Patients accept or decline treatment through the process of informed consent. Choices must be voluntary and be made without undue influence from others. Clinicians must ensure that patients have adequate and balanced information. Informed consent is not required should patients waive their right to make an informed decision and ask that their decision be made by their family, or even their physician. Second, informed consent is not required for emergencies.[24] Failure to determine and clearly document goals of care in advance can

result in inappropriate use of emergency interventions. Even when patients have advance directives, these are often overridden, ignored, or never communicated in emergency settings.

What is the best response to requests for physician-assisted suicide or euthanasia?

The re-emergence of the 'right to die' social movement in the mid-twentieth century, and its rise to prominence, parallels the development of successful medical interventions to extend life, as well as the legal milestones sanctioning the right of individuals (or their proxy decision-makers) to refuse or withdraw life-sustaining measures.[25] Patients with ALS have featured prominently in legal challenges for the right for physician-assisted death (PAD).

Patients with ALS seem to be more likely to request and complete physician-assisted suicide (PAS) or euthanasia than those with other terminal diseases such as cancer.[1] Interest in assisted suicide is high and sustained.[21,26,27] There remains a high level of interest in hastening death even in those patients enrolled in a hospice, or under the care of physicians with palliative care training.[28,29]

The debate about the ethics of PAS and euthanasia centres around the interpretation of basic principles of medical practice that superficially do not conflict: the imperatives to relieve suffering, respect patient autonomy, and to do no harm. Medical ethicists have written in support of PAS and euthanasia for patients with ALS[29] and in opposition to it.[30] It is therefore not surprising that a survey of neurologists reveals a range of attitudes, as well as persistent concerns about the morality and legality of withdrawal of life support and the use of medications that sedate or potentially depress respiratory function.[31]

The AAN and other professional organizations specifically condemn PAS and euthanasia.[32] Yet 44% of neurologists surveyed indicated willingness to perform PAS if it were legalized, and 13% would do so under current conditions.[33] The most recent EFNS guidelines have no recommendations as to how to respond to requests for assisted death, despite its availability in several European nations; however, studies on the prevalence of, and determinants for, wishes for a hastened death were recommended.[7]

Studies of the apparent motivation of those requesting or completing PAS in Oregon, USA, suggest that loss of autonomy, control, and independence, and the inability to pursue pleasurable activities play a greater role than physical symptoms.[33,34] In addition, fear of future suffering and higher levels of hopelessness (but not depression) as well as fixed characteristics of the individual, including male gender, higher educational and socio-economic status, and potentially modifiable factors including religiosity were also significantly associated with

interest in PAS.[21] In contrast, a request for PAS in the last month of life did correlate with higher pain scores and insomnia.[35]

Nearly 20% of ALS patients in a hospice setting[28] revealed a significant interest in hastening death. The decision to hasten dying was expressed consistently before death. Those who hastened dying reported poorer mood and less religiosity; they were more likely to have depressive symptoms of clinical significance, feel less in control, and more hopeless.

Studies in the Netherlands of patients dying of ALS in two periods, 1994–9 and 2000–5, showed the rate of physician assistance in dying (PAS and euthanasia combined) was constant at about 20% of ALS deaths.[36,37] The choice of PAD was positively associated with dying at home, higher education, anxiety and hopelessness, and lesser importance of religion but not demographic features or physical symptoms. Fear of choking, no chance of improvement, dependence on others, and fatigue were significantly more likely to be reported by caregivers as the reason for PAD[37] Compared with those with cancer or cardiac failure, the causes of unbearable suffering in ALS patients using PAD were fear of suffocation, dependency, loss of dignity, dyspnoea/poor communication, and fear of dependency, not somatic symptoms or depression.[38]

The earlier study yielded other interesting information about death and ALS. No EOL decision was made in 27%, and in 18% 'such decisions could not be made because the patients died suddenly'. Thus 45% of patients studied died without a decision about respiratory interventions. Although tracheostomy was present in 3% and NIV used in 16%, no comment was made about withdrawal of respiratory support or how such cases were classified. Guidelines for withdrawal of respiratory support from ventilator-dependent patients provide specific recommendations that include both sedation and analgesia;[6,7,39] however, it is possible that euthanasia may have been used instead.

Requests for hastened death are challenging for physicians, even where it is legal. Interviews with physicians in Oregon who received these requests demonstrated that they are emotionally difficult both for physicians who might participate in PAS as well as those who feel they cannot.[40,41] The physician should be ready to listen thoroughly and assure the patient that no matter what the final decision, the physician is available to the patient through the illness, even if he or she cannot—or will not—prescribe a lethal medication.[41–43] Some physicians reported a sense of hopelessness and failure after receiving a request. At other times, too much empathy and identification with the patient will lead to failure to look thoroughly for alternatives.

Interest in, or requests for, PAS may reflect a number of concerns. These queries should be approached as the opening to discuss EOL issues in general. In our experience, patients who persist in wanting PAS have strong needs

for control, negative views of the future, and a strong dislike of being dependent on others—all areas in which ALS particularly affects people. There is the risk that too much medical intervention may result in the patient feeling more dependent. Every effort to improve the patient's independence and avoid institutionalization should be made, even if safety in the home is not optimal.[40]

Summary and conclusions

The course of ALS is marked by frequent and important decisions, particularly about life-sustaining therapies and EOL care. Early involvement of the patient and loved ones in the process of informed decision-making can have a positive impact on quality of life as well as ensuring that medical interventions are consistent with the values and goals of the individual with ALS.

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Chapter 5

Advance directives

Gian Domenico Borasio and Raymond Voltz

Summary

The use of advance directives (ADs) is increasing in many parts of the world, allowing the wishes of patients to be clear even when they are not able to express them due to the progression of their disease. Discussing and formulating an AD in ALS can be difficult and time-consuming, and physicians and healthcare professionals may feel inadequately prepared for this task. This chapter includes a checklist designed to help professionals feel more comfortable with these discussions.

Advance directives in ALS

ADs are increasingly recognized as an important tool for safeguarding patient autonomy at a stage when patients cannot be asked directly about their preferences, as is often the case in the terminal phase of many diseases, including ALS. However, there is no clear evidence that ADs improve quality of life in ALS.[1] Specifically, ADs did not improve patient–physician communication or decision-making in two studies involving cancer patients.[2,3] On the other hand, educating physicians about ADs significantly increased the frequency of their use and improved physicians' attitudes towards ADs.[4] Importantly, physicians are more likely to follow detailed therapy- and disease-specific ADs that are supported by a discussion with the patient, and a proxy designation rather than a generalized AD.[5]

It is clear from several studies that both physicians and patients do want to use ADs.[6] Interestingly, although patients want to start the discussion about ADs earlier than their physicians, both patients and physicians believe that it should be the doctor who initiates discussion.[7] Unfortunately, such discussion often does not take place.[8] In addition, a cross-cultural study found large variations in the use of ADs between the United States, Europe, and Japan,[9] despite a uniformly positive attitude towards ADs amongst healthcare professionals.[10] A qualitative study underscored the importance of early patient–physician communication on ADs.[11]

In a sample of ALS patients receiving mechanical ventilation, 96% had completed or wished to complete an AD.[12] The same study showed that patients with ALS who had completed an AD were significantly more likely to have communicated their preference to stop ventilation to their family and physician than patients without an AD. A separate study in a general ALS population showed that the great majority (81%) of ALS patients want 'as much information as possible'. Importantly, a significant proportion of surveyed patients (8 of 26) changed their preferences for life-sustaining measures (e.g. ventilators) over a 6-month period.[13] In a study on the last month of life in ALS patients,[14] caregivers reported that 86% of patients had a living will, 76% had nominated a healthcare proxy, and 88% had at least one of these ADs. Half of the caregivers reported that the AD was helpful, and half reported that it had 'no effect on care'. Four patients received cardiopulmonary resuscitation, including two for whom the primary goal of care was to relieve pain and discomfort even if it meant shortening life.

Based on the available evidence and our clinical experience, we propose the following guidelines for AD in ALS:

1 The physician should initiate discussion of AD in ALS. Such a discussion should be attempted with all patients.
2 The discussion must be the result of intensive and long-standing communication between the healthcare professionals involved, patients, and relatives.
3 The discussion needs to be initiated well in advance of the terminal phase (at the latest when the first dyspnoeic symptoms appear; see Chapter 3).
4 The AD should be as detailed and therapy-specific as possible. It should include clear statements about life-sustaining and invasive treatments (e.g. percutaneous endoscopic gastrostomy, tracheostomy).
5 The family, particularly the primary caregiver, should be closely involved in the process.
6 Cultural differences need to be recognized and taken into account when discussing and formulating the AD.
7 A healthcare proxy should be appointed according to local laws and regulations. He or she should also sign the AD.
8 A copy of the AD should be forwarded to the home physician and the emergency room and intensive care unit of the local hospital.
9 All professionals involved in the care of the patient, including nurses, counsellors, hospice staff, etc., should be informed about the AD.
10 Once established, the AD should be periodically re-evaluated and re-signed by patient and proxy at 6-month intervals.

With the advent of new therapies and new methods for prolongation of life, the importance of ADs will undoubtedly increase. Care must be taken to comply with local laws and regulations, which may vary greatly between countries and states. With patients from some cultures, it may be appropriate to initiate the discussion with the family first.[15] However, the final decision resides with patients unless they prefer otherwise.

Discussing and formulating an AD in ALS can be a difficult and time-consuming effort. Physicians often feel inadequately prepared for this task, and fear legal consequences, especially with regard to decisions involving refusal or withdrawal of life-sustaining treatments. Collaboration with hospice institutions can be invaluable here.[16] A checklist which may be helpful for the planning of end-of-life decisions follows. Suggestions for ADs specific to ALS have been published,[17] but are still a matter of controversy.[18] A pilot study has shown the feasibility of an interactive, computer-based decision aid for advance care planning in ALS.[19] More education and research is needed in this area to develop accepted guidelines and increase the awareness of patients and physicians of the importance of ADs for preservation of patient autonomy at the end of life.[20]

Checklist for planning of end-of-life decisions and ADs in ALS

Contents

1 Medical therapy decisions:
   - disease-modifying treatments (e.g. riluzole): when to stop them?
   - non-invasive ventilation—when to stop?
   - percutaneous enterogastrostomy
   - cardiopulmonary resuscitation
   - invasive ventilation
   - antibiotics
   - nutrition/hydration
   - specific emergency treatments: dyspnoea, pain.

2 Locus of care:
   - stay at home?
   - admission to hospital?—which hospital?
   - hospice care?

3 Network of care and support:
   - for example family, friends, emergency phone numbers, physicians, nurses, social worker, other professionals, technical help, hospice group, proxy.

4 Personal matters:
   - financial situation/constraints?
   - testament?
   - specific spiritual assistance required?
   - funeral details?

5 Advance directives:
   - proactive for specific AD
   - will the patient give an oral AD?
   - does patient want information on a written AD?

6 Appointing a proxy:
   - informal or implicit appointment?
   - wants information on formal appointment?

How to proceed

1 Advance directives:
   - Must be result of an intensive and long-standing communication between healthcare professionals, patients, and relatives.
   - Note oral ADs in the medical record.
   - Help with a written AD: patient must be fully informed about diagnosis, prognosis, options; patient must have decision-making capacity (statement of treating physician included, if in doubt: neuropsychological testing required); Must be as specific and individualized as possible—regular revision required.
   - A witness should be present and the AD must be signed by the patient. If this is not possible, assistance by a lawyer/notary or video documentation of the patient's agreement are recommended.

2 Appointing a proxy:
   - Must be the result of intensive and long-standing communication between healthcare professionals, patients, and relatives.
   - Has the patient informally or implicitly named a proxy already?

- Help in formally appointing a proxy according to national laws: proxy must be trustworthy, fully informed, present; check conflict of interests; regular revision and discussions.

3 Accompanying measures:
- Check compliance with national laws.
- Signature of witness, caregivers, relatives, physicians.
- Statement of treating physician: patient has signed AD in my presence; patient is fully informed about disease and the meaning of an AD; patient has discussed the AD with other professionals and relatives; patient does not show signs of cognitive dysfunction, depressive syndrome, or other mental impairment; patient is informed about the possibility of withdrawal of the AD at any time.
- Statement of patient: fully informed about the disease and meaning of an AD; has discussed the AD with other professionals and relatives; is informed about the possibility of withdrawal of the AD at any time; 'I want my physician/my proxy to have no/slight/moderate/much leeway in the actual decision'.
- Statement of relative/proxy: fully informed about contents of the AD and willing to comply with it.

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Chapter 6

Respiratory complications

Deborah Gelinas

Summary

Progressive loss of function of the respiratory muscles ultimately results in respiratory failure, the cause of death in the majority of ALS patients. However, before the strength of the respiratory muscle declines to a critical level a number of symptoms can arise, generally insidiously. Recommendations for respiratory management, based on the best evidence and expert opinion, have been published in detail.[1–4] This chapter will describe the symptoms that arise from progressive weakness of the respiratory muscles and outline available management strategies.

Case history

Mr DS, a 64-year-old supplier of car parts, developed left foot drop, and within 5 months noted wasting of the muscles of his left hand, at which time a diagnosis of ALS was made. Within a year of his first symptom he complained of a 'muzzy' head in the morning, excessive daytime sleepiness, and was often found sleeping at his desk at work. A reduced appetite and resulting weight loss worried his wife. He complained of shortness of breath, and by this time his left leg weakness had progressed such that he was unable to walk and used a wheelchair to get around. His sniff nasal inspiratory pressure (SNIP) was reduced (31 cm H_2O versus the predicted 100 cm H_2O). Ear lobe blood gases showed a Pco_2 of 6 kPa, Po_2 of 9.4 kPa, and a bicarbonate of 30 mmol/L. A polysomnogram showed repeated arousals due to hypoventilation. He started to use NIV overnight. He had increased energy levels in the day and returned to a full day's work. His appetite improved and he gained weight. In January 2000 he experienced dyspnoea after exertion and breathlessness after meals and began using NIV after eating and after exertion such as taking a shower. He relied on NIV almost continuously for the 3 months prior to his death. In that period, he still liked to shower, using sublingual lorazepam for relief of dyspnoea. He died peacefully at home, while on NIV. Although he had difficulty clearing his secretions in the weeks prior to his death, he declined invasive ventilation. His secretions were managed with hyoscine patches and suction that his wife applied. During the last few months of his illness, he was unable to attend the hospital clinic but was visited at home by community palliative care team. His wife was his sole carer.

Symptoms of respiratory muscle weakness

Overt breathlessness, or shortness of breath, is not usually the first symptom of respiratory muscle weakness in ALS. The insidious onset of exercise intolerance or dyspnoea on exertion may be overlooked, especially if motor decline limits physical exertion. The first manifestation of significant respiratory muscle weakness may not be dyspnoea but symptoms of sleep-disordered breathing (SDB). Hypoventilation may occur during sleep when overall respiratory muscle strength still remains sufficient for daytime ventilation.[5,6] Nocturnal hypoventilation causes complaints of non-refreshing sleep, daytime sleepiness, increased frequency of and particularly vivid nightmares (as arousal in rapid-eye-movement (REM) sleep causes waking mid-dream), reduced ability to concentrate, mood disturbance, and loss of appetite. Some patients report that a dry mouth wakes them as the increased respiratory effort can result in 'panting' at night. Eventually hypoventilation leads to significant nocturnal hypercapnia which gives rise to morning headache. Excessive daytime sleepiness, lack of appetite, and personality change can occur. Orthopnoea (breathlessness when lying flat) is a symptom of weakness of the diaphragm as assuming a recumbent position displaces the contents of the abdomen, normally held in place by the diaphragm, into the thorax, thereby reducing lung volume. By the same mechanism dyspnoea can occur when standing in deep water or bending over. However, orthopnoea is not a universal finding and may be absent in the presence of severe generalized respiratory muscle weakness, when weakness of the abdominal wall prevents supine upward displacement of the diaphragm. As weakness of the respiratory muscles becomes more generalized, patients may notice exertional dyspnoea; however, this may be under-recognized if mobility is curtailed by limb weakness. Patients describe dyspnoea on talking and a decline in speech volume, with their cough and sneeze becoming audibly weaker. Dyspnoea at rest occurs when global inspiratory muscle weakness is profound; by this stage patients are close to hypercapnic ventilatory failure.

The signs of inspiratory muscle weakness are subtle, starting with increased respiratory rate (tachypnoea), orthopnoea, difficulty managing secretions, weak cough, weak voice, and weight loss. Later, use of auxiliary respiratory muscles, decreased chest wall movement, and paradoxical movement of the abdomen (caused by diaphragm weakness) can be observed. In late stages only straining of the neck muscles and absent chest wall motion may be seen.

Assessment of respiratory function

The respiratory muscles are traditionally subdivided into groups, according to their predominant function.

Inspiratory muscle function

The diaphragm is the most important *inspiratory muscle*, which, at rest contributes over 70% of minute volume. However, increasing ventilatory requirement, for example during exercise, involves activation of accessory muscles (the intercostal muscles, scalenes, trapezii, and sternocleido-mastoid). Use of these muscles at rest is a sign of respiratory insufficiency.

Initially, nocturnal hypoventilation occurs in rapid eye REM or 'dreaming' sleep as the accessory muscles, particularly the intercostal muscles, are less active in this sleep stage.[7] Ventilation then becomes exclusively dependent on the diaphragm, which, if weak, is further disadvantaged by the supine position. As weakness progresses, episodes of hypoventilation occur throughout sleep, causing repeated arousals and sleep fragmentation. Other patients ascribe their arousal to urinary problems; however, complaints of nocturia frequently resolve with treatment of hypoventilation, being a symptom rather than a cause of the sleep disturbance. In other patients it is their bed-partners who are first aware of restless and disturbed sleep, and may report periods of apnoea or notice change of personality or mood.

Expiratory muscle function

The abdominal muscles are predominantly *muscles of expiration*. Working in coordination with the muscles of the upper airway, their most important contribution is the generation of an efficient cough. ALS patients with significant abdominal muscle weakness may complain that they are unable to clear secretions, or may develop a recurrent cough.

There may be few signs of expiratory muscle weakness; however, poor performance of a voluntary cough can be revealing, with an audible abnormality. As the generation of an efficient cough requires coordination of inspiratory, expiratory, and upper airway muscles, an ineffective cough may arise in a number of ways and will be discussed in Measures of Respiratory Muscle Strength.

Upper airway function

The many muscle of the upper airway and vocal cords are affected progressively in ALS patients, causing episodes of choking and severe dyspnoea, especially when swallowing liquids.

Abnormalities in the flow-volume loop of pulmonary function tests are associated with narrowing of the airway due to weakness or paralysis of the vocal cords and may even anticipate the appearance of bulbar signs.[8] In ALS the cricopharyngeal sphincter muscle becomes hyper-reflexive and hypertonic, and the normal coordination of laryngeal closure with voluntary swallowing is lost.[9]

Patients with UMN bulbar signs particularly report the distressing symptom of choking,[10,11] perhaps because of a lower threshold for triggering the closure reflex, enhancing the risk of upper airway obstruction.

Investigations of respiratory function in ALS patients

Measures of respiratory muscle strength

Symptoms and signs are unreliable markers of respiratory muscle weakness, but there is no definitive test that detects early respiratory failure.[1] Detection of respiratory muscle weakness has prognostic significance[3] and timely introduction of respiratory aids including NIV can improve quality of life and improve survival. Therefore pre-symptomatic testing of respiratory function is considered the standard of care.[1–4] The most frequently performed tests are of vital capacity measured as FVC or slow vital capacity (SVC). These can be performed with the patient sitting or supine, the latter method being more sensitive to diaphragm weakness, which can correlate with respiratory symptoms in ALS.[12–15] The results are reported in litres, and as a 'per cent of predicted' based on normative data for gender, height, and weight. Maximum inspiratory pressure (MIP) and maximum expiratory pressure (MEP) are measured by inhaling and exhaling through a mouthpiece connected to a pressure transducer and are considered to be measures of inspiratory and expiratory muscle force, respectively. Patients with weakness of the bulbar muscle have difficulty with these tests, and they can be less accurate in this group.

The SNIP is a useful screening investigation, performed by inserting a nasal plug and measuring the pressure generated by serial sniffing. This can be more accurate in patients with bulbar weakness.[16,17] A result within the normal range means significant weakness is unlikely.

A cough is generated by inspiration of a volume of air, closure of the vocal cords, contraction of the expiratory muscles and generation of increased pressure in the thorax, and finally sudden opening of the cords, with subsequent explosive exhalation. Weakness in any muscle group can reduce the effectiveness of a cough. Peak cough flow (PCF) can be measured. Patients cough into a peak flow meter—more commonly used to measure peak flow in asthma. Using measurements of PCF, Bach[18] has suggested a plan of 'cough management' for patients with ALS. An adequate cough produces a PCF greater than 160 L/min. All the above tests require effort and coordination with the patient, which may be compromised by fatigue, secretions, coughing, or by cognitive impairment.

Blood gas analysis

Most ALS patients have normal lungs, and by the time the result of an arterial blood gas test (the gold standard of respiratory failure) shows hypoxia, in addition to hypercapnia, respiratory muscle weakness is profound and, without assisted ventilation, the prognosis is likely to be limited to months or weeks. Analysis of arterial blood gas tension is at times a useful adjunct to other measures of respiratory function. Progressive respiratory muscle weakness results in hypercapnic ventilatory failure and chronic respiratory acidosis. Renal excretion of hydrogen ions in exchange for bicarbonate ions compensates for this by raising plasma bicarbonate, which increases chloride excretion. Therefore, chronic hypercapnia will result in reduced venous chloride and raised venous bicarbonate. Two studies in ALS[19,20] have shown that venous bicarbonate rises and chloride falls to abnormal levels close to death. In the latter study, the mean (range) time from detection of abnormality to death was 2.2 (0.5–6) months, suggesting that the development of hypercapnia is a pre-terminal event.

In ALS, it is unlikely that weakness of the inspiratory muscles is the cause of severe hypoxia in the absence of hypercapnia, and in this case a lung parenchymal cause, such as pneumonia or pulmonary embolism (PE), should be sought.

Overnight oximetry and polysomnography

Measurement of nocturnal oxygenation saturation at home (where available) or laboratory-based polysomnography can provide additional information about respiratory function at night, when respiratory drive may be reduced, diaphragm weakness may be compounded by position, and other nocturnal symptoms may disrupt restorative sleep. During polysomnography, measurements are made of oxygen and carbon dioxide levels, abdominal and thoracic movement, as well as the electroencephalogram (EEG) from which sleep can be staged. Indications for a polysomnogram may include symptoms of sleep disturbance without evidence of daytime respiratory failure, in order to gather evidence of hypoventilation-related arousal and exclude causes of sleep disturbance not due to the respiratory muscles. If arousal due to hypoventilation is present, the sleep disturbance may be effectively treated with nocturnal ventilation; if not then other causes of sleep disturbance should be sought. However, polysomnography is not always feasible. Nocturnal hypoventilation can be identified by recording nocturnal oxygen saturations using pulse oximetry. Many oximeters can store at least 8 h of data and patients can use the machines at home. A number of studies[21–23] have shown that they are useful for the management of ALS patients and can identify hypoventilation due to sleep-disordered breathing. However, nocturnal studies are not required to initiate NIV.[4]

Treatments of the symptoms of respiratory muscle weakness

Symptomatic treatment of respiratory symptoms consists of airway and secretion management, medical (mostly pharmacological) interventions for control of dyspnoea, and ventilator support, with NIV or invasive ventilation.

Recognition of the impact of uncoordinated swallowing, vocal cord dysfunction, and pooling of secretions on the perception of choking and breathlessness is important for the control of symptoms of breathlessness and the success of NIV. (Chapters 7 and 9 give recommendations for the management of symptoms related to dysphagia and secretions.)

Maintaining an unencumbered airway includes developing an effective cough strategy when muscle weakness, whether of the abdominal muscles, diaphragm, or upper airway, compromises effective airway clearance and effective cough. Peak cough expiratory flow can be measured with a peak flow meter or during spirometry. Values >160 L/min are needed to clear the airways of food or secretions, but values < 225 L/min were associated with poorer outcome during respiratory infections.[1] Techniques include assisted cough with abdominal thrusts, usually administered by a caregiver, or lung-volume recruitment followed by assisted cough techniques; mechanical cough devices are available.

Breath stacking is done by voluntarily closing the glottis between breaths to prevent loss of the inspired volume of air. With a weak glottis, as in ALS, expiration can be occluded with a one-way valve. In lung-volume recruitment, inspired volumes can be provided with a resuscitation bag connected by a piece of short tubing to the one-way valve that is in turn connected to a mouthpiece or mask. With bulbar weakness, occlusion of the nose and use of a mask is required. Several pumps of the resuscitation bag through the closed circuit allow air to be retained in the lungs; this can be used to treat or prevent atelectasis and the inspired air used for an assisted cough.[4]

The mechanical insufflator–exsufflator (MI-E) delivers rapidly alternating positive and negative pressures via a facial mask. Studies have quantified the magnitude of enhancement of PCF afforded by various techniques [24,25] and the latter study found that MI-E produced the greatest improvement, but the majority of patients studied had minimal bulbar dysfunction. Sancho et al.[26] described collapse of the upper airway when the device was used in ALS patients with significant bulbar weakness. Bach[18] suggests a plan of 'cough management', involving cough enhancement techniques including MI-E. Comparison of assisted cough techniques in 28 patients with advanced ALS found that all instrumental methods (including manually assisted breath stacking) were significantly superior to manual methods. Patients preferred to use their NIV

machines but judged the MI-E mist effective. While a variety of instrumental techniques were used initially, all surviving patients were using MI-E eventually.[27] MI-E is not widely available in all countries. At the very least patients and carers should be taught the assisted cough technique, breath-stacking, or lung-volume recruitment, and, where indicated and available, MI-E should be supplied. Although several surveys in the United States[28] and the UK[29] found it used in only 5% of neuromuscular clinics, the revised AAN Practice Parameter recommends the use of cough assist machines for patients who cannot clear secretions.[1]

Thick, tenacious secretions may be exacerbated by dehydration. Together with adequate hydration, mucolytic agents such as acetylcysteine 200–400 mg three times a day, or beta-receptor antagonists (e.g. metoprolol or propranolol) in a saline nebulizer with ipratropium or theophylline may allow mobilization with cough assist techniques.[2] A portable suction device and humidification are also necessary.

Breathlessness and other symptoms of dyspnoea can be relieved by upright positioning, airflow on the face, and other manoeuvres. Pharmacological management consists of opiates, morphine being the best established, and benzodiazepines for anxiety. The risk that morphine will precipitate or worsen respiratory failure is over-emphasized. In a small study six ALS patients with symptomatic dyspnoea were given morphine in doses ranging from 2.0 to 20 mg; this improved tachypnoea and was associated with a rise in blood oxygen without affecting carbon dioxide levels. Morphine was associated with a significant drop in dyspnoea scores whereas oxygen administration was not.[30] Judicious use of morphine following established palliative care protocols for breathlessness is therefore justified and recommended, since there are no studies on protocols specifically for ALS. Dyspnoea may be alleviated by use of NIV. However, this may not be tolerated in all patients and may not be indicated in the terminal stages of ALS. Patients may require management of anxiety and dyspnoea while acclimatizing to NIV and as the benefits of NIV decrease with advancing respiratory failure.

Treatment of respiratory muscle weakness with non-invasive positive-pressure ventilation

Non-invasive positive-pressure ventilation (NIPPV) is a treatment modality whereby ventilation is delivered via a nasal or facial mask.[31] NIPPV can improve the polysomnographic and blood gas abnormalities associated with SDB,[32] relieve symptoms of sleep disturbance and breathlessness, improve cognitive function,[33] and reduce the morbidity of gastrostomy placement.[34–36]

NIPPV can be used when respiratory muscle weakness becomes profound and the patient is incapable of ventilator-free respiration (Bach[18] reported patients using 24-h NIV). Survival is increased with NIPPV.[37] The majority of studies compared survival of patients using ventilation with those who either did not accept or 'tolerated' NIPPV, and demonstrated prolonged survival in those using NIPPV. A single well-designed randomized trial of 41 patients to receive NIPPV or 'standard care' demonstrated both prolonged survival and improved quality of life in those tolerating NIV.[38]

The practice of offering NIPPV to ALS patients had a slow uptake. Despite recommendations, many patients with ALS are not offered NIPPV.4,[39] Although the prevalence of NIV is growing, only 20% of patients with respiratory dysfunction received mechanical support in a province of Italy[40] and in a survey of ALS tertiary care centres in Canada.[41] Barriers cited in the latter study included patient intolerance of NIPPV and lack of availability of appropriate specialist respiratory care. This disparity of management may also be due to the expense of NIV, as well as decreased adherence of ALS patients with bulbar and cognitive impairment.[41] ALS patients with normal to moderately impaired bulbar function benefit most from NIV, with both improved survival and quality of life (QOL).[37]

Initial data on QOL in ALS patients receiving NIPPV showed an improvement in QOL scores after initiation.[42,45] Bourke et al.[38] demonstrated that patients randomized to receive NIPPV had significantly improved QOL compared with those receiving standard care.

Areas of controversy in NIPPV for ALS

The optimal time for initiation of ventilation is not precisely known.[1–4] Discussion of disease progression and respiratory muscle weakness may provoke anxiety in patients and carers. However, failure to recognize impending ventilatory failure may result in patients being intubated and ventilated before their views on this treatment can be established. Acute ventilation has a poor outcome; in one study 29% of patients died in the intensive care unit and 54% required permanent tracheostomy.[46] Current recommendations are to discuss respiratory support with patients and families before symptoms occur, in part to educate them about subtle symptoms, including those related to SDB. NIPPV should be offered when symptoms occur, particularly orthopnoea, or when evidence of progression on respiratory testing reaches the recommended thresholds for asymptomatic initiation of NIPPV. Hypercarbia ($P_aco_2 > 45$), FVC < 50% predicted, or MIP < 60 mm H_2O should prompt consideration of NIPPV.

The benefit to patients with bulbar symptoms, particularly severe ones, is controversial. The factors that influence the magnitude of the effect of ventilation

on subsequent survival and QOL suggest that the presence of significant bulbar weakness decreases the survival benefit of NIV while still benefitting QOL. There is a theoretical risk of increased aspiration with NIPPV, as the positive pressure can potentially blow upper airway secretions through an inadequately protected airway. Studies report differing effects of bulbar weakness on the response to NIPPV, with some suggesting poor tolerance of NIPPV[45,47] in the presence of severe bulbar weakness and others finding bulbar symptoms did not influence acceptance.[48,49] Secretion management is extremely important for successful use of NIPPV by patients with significant bulbar weakness.[50] Bourke et al.[52] found that patients with bulbar weakness using NIPPV noted improved sleep and reduced breathlessness, but QOL was not improved to the same extent as in patients with lesser bulbar weakness. Survival in patients with bulbar weakness using NIPPV was not significantly prolonged. Volanti et al.[51] found that with careful preparation and education, most patients offered NIPPV, including those with severe bulbar involvement, were able to tolerate it and use it long term.

Abnormal vocal cord movement seen at bronchoscopy as well as polysomnographic evidence of upper airway obstruction, presumably from the adducted vocal cords, could explain intolerance in some patients with episodic choking symptoms. Ventilators capable of delivering expiratory pressure can be used to try and overcome this obstruction, but NIPPV did not improve symptoms in these patients and in some cases precipitated episodes of stridor. Other patients with bulbar involvement and profound hypercapnia had minimal symptoms. Although NIPPV improved nocturnal hypercapnia, patients did not feel better. It may be that such patients have a reduced drive to breathe due to brainstem involvement and although hypercapnic do not recognize this, hence the lack of symptoms. Even patients with milder degrees of bulbar dysfunction may have nocturnal patient–ventilator asynchrony,[52] and therefore periodic monitoring of NIPPV with polysomnography or nocturnal oximetry is advised to provide optimal support. Some centres offer NIPPV to all patients with bulbar dysfunction having symptoms of sleep-disordered breathing or respiratory failure, vigorously treating secretions in order to improve acceptance. Such patients are advised that the risk of aspiration may be increased with NIPPV and early gastrostomy may be offered. It is important to emphasize to patients and carers that the treatment may not be helpful to everyone and that patients should make their own choice to use NIPPV or not. As patients and carers have become more aware of the potential improvement in survival with NIPPV, patients who do not feel that it improves either symptoms or QOL significantly have occasionally felt pressured to use it to prolong life.

Bach and colleagues have emphasized the importance of adequate cough capacity both in the choice of mode of ventilation and the effect of NIPPV on survival.[18,22,28] They point out that NIPPV compensates for inspiratory muscle weakness but suggest that prolonged survival also requires attention to expiratory and bulbar muscle dysfunction.

Practical aspects of NIPPV in ALS

When discussing the use of NIPPV with patients and their carers, there are a number of points that need to be considered. First, it is important to be sure that the symptoms of sleep disturbance are due to respiratory muscle weakness and might therefore be ameliorated by NIPPV. Second, it is important to make an assessment of the patient's home situation. Efficient ventilation requires careful mask application, and patients with significant upper limb and hand weakness are unlikely to be able to achieve this independently. ALS patients living alone may need to consider accepting a night-time carer. NIPPV is very likely to prolong survival, and therefore patients are likely to experience increasing disability whilst using a ventilator. Additionally, ventilation does not prevent progressive weakness of the respiratory muscles; therefore, patients who accept nocturnal ventilation gradually begin to use the machine in the daytime to alleviate dyspnoea as the strength of the respiratory muscles declines. This leads to increasing ventilator use, and most patients will eventually use 24-h ventilation. Clearly this has implications for care provision. Although long-term 24-h NIPPV has been described,[18] NIPPV is less efficient than tracheostomy ventilation (TV), and at this stage TV may be considered; however, both patients and caregivers report NIPPV to be preferable to TV.[53] Tracheostomy care involves nursing input; domiciliary use is thus expensive and may be burdensome to families,[54] and in some health systems rarely accomplished. Cazzolli and Oppenheimer[55] found that no ALS patients cared for at home with NIPPV required additional nursing input compared with 80% of TV patients. However, the increased care burden of all forms of mechanical ventilation should not be underestimated.[56]

The ventilator may be used for dyspnoea while awake, but this does not preclude the use of pharmacological means of symptom control. It is also stressed that patients may cease using ventilation whenever they wish, and that drug treatments can be used to alleviate distressing symptoms during discontinuation. Agreed end points should be discussed at or just after initiation of NIPPV.

Choice of ventilator

Ventilators suitable for home NIPPV use are programmed to deliver a set positive inspiratory pressure or volume, differing in magnitude during inspiration and

expiration, via a nasal or full facial mask or mouthpiece. It is essential that the ventilator can be programmed with a back-up rate capable of achieving adequate ventilation during apnoea or hypoventilation. Most can be triggered by the patient to deliver extra breaths if required. Some ventilators have an integral battery, so can be used by ventilator-dependent patients who wish to go out for short periods of time.

A newer option in mechanical ventilation which combines the benefits of both pressure ventilators and volume ventilators is pressure-controlled volume-guaranteed ventilation (PCVGV) in which a specific tidal volume is chosen which is appropriate for the patient and the ventilator delivers that volume using a decelerating flow and a constant pressure. The inspiratory pressure which is needed to deliver the tidal volume is adjusted breath by breath so that the lowest pressure is used. PCVGV compensates for changes in lung compliance, mask leakage, and positioning of the patient. Potential benefits are the use of a lower inspiratory pressure and reduced lung injury from over distension.[57,58]

A full discussion is beyond the scope of this chapter, but PCVGV is reviewed in detail by Gruis and Lechtzin.[4]

Choice of interface

The interface, that is the method of delivering ventilation, can vary from a nasal mask, a full-face mask, nasal cushions, or a mouthpiece. All of these come in a wide variety of sizes and designs. It is important to choose a system that is comfortable and can be easily applied for each individual, taking into consideration the degree of upper limb weakness. A number of patients will require a customized mask for an adequate fit. The nasal mask is usually the simplest to fit but pressure sores can occur on the bridge of the nose. This occurs more frequently when the mask is too tight or when it is used for longer periods of time. Once a sore has developed this can be difficult to heal so prevention is the key. First, a well-fitting mask is essential as patients and carers frequently over-tighten the straps in response to leaks, which can be uncomfortable, particularly if air goes into the eyes. Second, patients can use protective dressing on the bridge of the nose. If patients begin to use ventilation both in the daytime and nocturnally it is helpful to use several different interfaces such as nasal mask, nasal cushions, or mouthpiece. To reduce the risk of mouth leak, which may decrease the efficiency of ventilation, patients can use a chin strap or full face mask.

Ventilator dependency and end-of-life issues

The fear of ventilator dependency and of patients reaching a locked-in state in which they are unable to communicate, may have limited the widespread use of mechanical ventilation in the past. It had been suggested that the use of NIPPV

(in contrast to TV) would avoid this situation as it was thought that the technique was not capable of sustaining ventilation when respiratory muscle weakness was such that 24-h ventilation was required . However, there is now considerable evidence that patients can survive for many years with 24-h NIPPV, particularly when techniques are used to achieve adequate clearance of secretions. Bach[18] reported a series of patients whose mean survival with 24-h NIV was 3.9 years, with a range of 2 months to 26 years. Some patients used NIV continuously. This requires the use of different interfaces and considerable skill from carers to prevent skin abrasions. Others may prefer to alternate NIPPV and pharmacological means to treat dyspnoea. Not all patients will use NIPPV continuously and some may die of pneumonia before they need 24-h ventilatory support.

Although patients and carers should be fully aware of all the options available to them prior to initiation of NIPPV, discussions need to be continued, particularly with increasing use of the ventilator. Coping with the changing situation is made easier when patients and carers have built up good relationships with both a respiratory and a palliative care team. As the disease advances, patients are likely to be using the ventilator to treat breathlessness by day, as well as to improve sleep. Other treatments of dyspnoea such as opiates, benzodiazepines (e.g. lorazepam), or buspirone hydrochloride can be used to treat breathlessness and allow time off the ventilator. There are a number of practical difficulties in patients who are ventilator-dependent—in whom discontinuation of NIPPV will result in severe dyspnoea, if not respiratory arrest. A back-up ventilator must be available in case of malfunction and interruption of the electricity supply should be planned for. Patients and carers should be made aware that when the respiratory muscles are so weak that they cannot sustain spontaneous ventilation, sudden deterioration and death from ventilatory failure may occur. At this stage, symptomatic treatment of increasing breathlessness with pharmacological treatment in addition to NIPPV may be the most appropriate course of action. This eventuality should be anticipated with patients and carers, and an acceptable management plan discussed and agreed. It may be appropriate for patients to be managed in the home or a hospice setting.

Patients should be aware that they can discontinue NIPPV at any time, and severe breathlessness and anxiety can be avoided in this event by pre-medication with opiates and anxiolytics.[59] This approach should not be regarded as unethical, nor does it amount to assisted suicide or euthanasia.

Common complaints from ALS patients using NIPPV

Nasal discomfort

Patients frequently complain of nasal discomfort, particularly in the first few weeks of using NIPPV. Excessive nasal secretions (rhinorrhoea) can be treated

by short-term use of an anticholinergic nasal spray, although prolonged use may result in excessive drying. Occasionally, particularly if the patient sleeps in a cool bedroom, warming the inspired air by entraining the ventilator tubing under the bedclothes may reduce rhinorrhoea. Nasal congestion responds to a steroid nasal spray, but frequently requires the addition of humidification. Humidifiers that can be inserted into the ventilator circuit are available, and room humidification may be helpful. Alternatively nebulized saline can be used.

Abdominal bloating

Patients may swallow excessive air during ventilation. This can be a particular problem if the patient is using a full-face mask, and may be solved by changing to a nasal interface. Drugs which increase gastric emptying, such as metoclopramide, may be useful and relief of constipation is helpful. Patients who have a gastrostomy tube can relieve distension due to gastric air by opening the tube and compressing the gastric air 'bubble'.

Leaks

Air leaks can cause discomfort and may also reduce the efficiency of ventilation. Air leaks into the eye are particularly uncomfortable, and can cause conjunctivitis. A well-fitting mask is essential and needs to be changed regularly, as masks can become stretched and less well-fitting with use.

Increasing dyspnoea

Patients may notice that they are particularly dyspnoeic immediately after coming off the ventilator. This is probably due to the sudden increase in the work of breathing by the respiratory muscles. Patients should be advised to anticipate this, and reduce the load on the muscles by sitting up before removing the mask. The sensation will then generally abate. Patients may report dyspnoea while using the ventilator. In some cases, adjustment of the settings may be required to compensate for increasing respiratory muscle weakness. However, it is possible to over-ventilate patients and the resulting hypocapnia produces the sensation of dyspnoea. If increased ventilation does not reduce dyspnoea, then repeated increases should be avoided until ventilation-induced hypocapnia can be excluded by blood gas measurement.

Other aspects of respiratory care

Oxygen therapy

Oxygen can provide symptomatic relief of breathlessness in patients with hypoxia. However, it may decrease respiratory drive and worsen the symptoms of hypoventilation, such as headache. It may also lead to severe and unpleasant dryness of the mouth. The use of oxygen in ALS should be restricted to patients

with a concomitant lung disorder such as chronic obstructive pulmonary disease or those who cannot tolerate NIV and are suffering from terminal dyspnoea.

Prevention of infection

It is advised that patients obtain influenza and pneumonia vaccinations.

Diaphragmatic pacing for patients with chronic respiratory failure

On the basis of a prospective multicentre trial, diaphragmatic pacing (DPS) has been approved in the United States for compassionate use in patients with ALS suffering from respiratory failure. Eighty-eight patients (38 of whom had ALS) underwent laparoscopic diaphragm motor point mapping with intramuscular electrode implantation, with subsequent stimulation of the electrodes to condition and strengthen the diaphragm. No perioperative mortality was reported in the ALS patients and the need for mechanical ventilation was delayed compared with historical controls. After conditioning the diaphragm with DPS, the average rate of decline in FVC was 0.9% per month compared to the pre-implantation decline of 2.4% per month, which extrapolated to an additional 24 months of ventilator-free survival. When compared with historical controls with the same degree of respiratory compromise as those included in the study (FVC < 50% or MIP < 60 or hypercapnia or nocturnal hypoxia of <88% for five consecutive minutes) diaphragm-paced ALS patients had an additional 16 months survival compared with ALS patients who did not use NIV and an additional 9 months survival compared with ALS patients who used NIV.[60] Patients who may be candidates for DPS are those with stimulatable diaphragms as determined by phrenic nerve stimulation and chest fluoroscopy. Although DPS appears to be reasonably safe and may improve both quantity and quality of life in carefully selected ALS patients, there remains much scepticism due to the absence of blinding in reported outcomes.[61]

Mechanical ventilation via tracheostomy

Case history

KL was a 55-year-old retired army sergeant major, a 7 year-survivor of ALS. A leader in the ALS community, he was charismatic and optimistic about his role as a patient advocate, adviser, and leader. After 5 years his use of NIV had reached 24 h a day and he was experiencing difficulty handling oral secretions. He had previously decided to accept mechanical ventilation via TV when NIV was no longer adequate. After a prolonged winter of weight loss, increasing fatigue,

and eventually aspiration pneumonia, he underwent tracheostomy and began TV. He remained in hospital for several weeks while his wife learned how to do endotracheal suction and valve changing in order to permit vocalization through his tracheostomy as well as the other nursing tasks required to safely bring him home. As he was already wheelchair-dependent, very few additional adaptations were needed to his home living situation. His wife did not work outside the home and thus was able to provide full time caregiving, with help from his two daughters. Within 3 months of TV, KL was again a major participant at ALS Association fundraisers. Throughout his convalescence from TV, he continued to answer more than 50 e-mails a day from ALS patients all over the country. His message was consistent: 'Life can be good. Make it good'. He continued to hold family meetings, although his wife joked that when the family was tired of his 'pontificating' she disconnected his speaking valve. They joked about the shared control they each maintained. Two years later he was actively mentoring young juvenile offenders at a local state penitentiary and getting involved in local government.

Treatment of ventilatory failure with invasive ventilation (indications, timing, and outcomes)

When patients opt for life-sustaining therapy such as long-term mechanical ventilation by TV there would appear to be no role for palliative care. The decision for TV may be made as an emergency due to acute respiratory failure (usually preceded by endotracheal intubation), or when NIV is no longer effective or not tolerated. Persistent oxygen desaturation (<95% for more than 24 h) despite the use of NIV and airway clearance strategies, signals the advent of acute respiratory failure and the need for TV to sustain life.[22] TV without mechanical ventilation may be used in patients who develop paralysis of the vocal cords or whose airways become severely encumbered with secretions.

What TV offers in terms of long-term prognosis varies significantly depending on the study. In a study of 101 patients with TV cared for at home, 87% were alive 1 year later, 69% at 2 years, 58% at 3 years, 50% at 4 years, and 33% at 5 years.[64] Of 52 ventilator-dependent patients, survival ranged from 11 months to 14.5 years with a mean survival of 4.4 ± 3.9 years.[65] In a 10-year, population-based study involving 1260 ALS patients, 10.6% underwent tracheostomy.[66] Overall 20.6% died in hospital, roughly half were discharged home, and 30% went to long-term care. The median survival was less than 1 year post-tracheostomy. In a single tertiary-care ALS centre, 31.1% of patients underwent TV.[63] Patients on TV survived a median time of 12 months longer than the NIPPV group. Survival for more than 1 year occurred in 36%, yet only 4% survived for 4 years post-tracheostomy and none past 5; 13/52 died in the first

month. Low FVC and age over 60 years correlated with poorer prognosis; 85% of those who died in the first month had undergone emergency TV.

Prolonged survival for patients suffering from ALS is possible using TV, yet the prevalence of TV for ALS patients is variable, ranging from 30% or higher in Japan,[4] Italy,[63] and Denmark[67] to 1.5% in a survey of Canadian ALS centres, despite similar publicly funded health systems.[41] In the United States, where many different medical coverage plans exist and TV may or may not be feasible, the use of TV varies greatly. The total cost of TV, including home nursing needs, is so great that it routinely exceeds all insurance plan coverage and the out-of-pocket expenses to the individual may be considerable.[68]

Physician attitudes may play a role. The use of TV for ALS in various Muscular Dystrophy Association centres, varying from 1.6 to 14.3%, correlated most closely with the personal attitudes of the treating physicians. Where the use of TV was greatest, the physicians indicated they would use TV themselves for ALS; where it was rarely used, physicians stated they would never use TV themselves and actively discouraged ALS patients from using it. Where use of TV was intermediate, physicians presented informed consent in a neutral fashion.[68] In both the United States and Japan, 70% of physicians state that they would not choose TV for themselves if they suffered from ALS.[69]

The role of the palliative care team, therefore, is as justified in the patient seeking TV as at any other point of the disease course. Patients choosing TV are still at significant risk of death, with high needs for social and spiritual support, as are their caregivers. On-going evaluation of QOL, as well as expert symptom control and support for decision-making, provide ample scope for the practice of palliative care in this setting.

Patient autonomy and competency

Ideally, additional information and assistance provided by other ALS patients using TV and their caregivers would provide prospective users of TV and their families with a more balanced informed consent process.[70]

According to ALS patients themselves, the decision regarding TV should be theirs. In an interview with 16 ALS patients contemplating the use of mechanical ventilation, nine discussed the decision in advance with family and friends, five with a physician, and two reported that they did not discuss it with anybody.[71] The factors considered to be most important in choice of a ventilator were: quality of life, severity of disability, ability to return home, ability to discontinue ventilation in the future if desired, and concern for the well-being of the family. In many cases patients choose TV knowing that it may entail a sacrifice of families' emotional, financial, and physical well-being.

The question of cognitive impairment in patients with ALS has added further complexity to decision-making with respect to TV. Frontal temporal dysfunction, reported in as many as 60% of patients with ALS[72] (see Chapter 8) affects the ability to plan for the future, make informed decisions, and appreciate the consequences of those decisions (especially how those consequences affect others).[73] Frontal temporal dementia is higher in ALS patients with bulbar-onset disease and is associated with decreased adherence to medical regimens and increased death rates.[74] Such bulbar-onset ALS might be expected to present in respiratory crisis, as did the majority of ALS patients in a survey of ALS patients on mechanical ventilation in six US states. Although 79% said they had been informed by their physician about TV prior to their respiratory failure, only 21% of them chose it in advance. Of those who were intubated and ventilated in an emergency situation, another 21% erroneously thought it would be only temporary.[68] Yet, awareness of the option of TV and a willingness to consider it does appear to play a role in choosing TV: in a prospective trial evaluating stated preferences regarding TV and actual choice of TV, 20% of ALS patients who stated that they favoured TV had one in the 12-month follow-up period compared with 3.4% of those who did not previously favour TV.[75] Hands-on information sessions may have an impact; 76% accurately predicted their subsequent choice of respiratory care after completion of such a workshop.[76] Those whose ultimate decision was not predicted all chose palliative care over respiratory interventions.

Patient and family satisfaction

Whether TV is chosen in advance or not, 90% of home-ventilated ALS patients were glad of their choice and would make the same choice again,[54] whereas 72% of those cared for in institutions were satisfied.[55] Interestingly, when their spouses were questioned regarding satisfaction with TV they answered enigmatically that they were glad the ALS patient had made the choice for TV but that they themselves would never choose TV because the decision made life too hard on the family. A study comparing the QOL of ALS patients with TV or NIPPV and their caregivers showed a comparable QOL for the two patient groups, but a much higher burden for the TV caregivers.[77]

Although the majority of ALS patients with TV rate their QOL as good, a review of the daily activities of TV users reveals severe restrictions. Days were spent in simple activities: 21% never left their homes, 95% spent their days watching television or talking with family and friends.[54] Increasing sophistication of communication strategies, interface with assistive devices including computers, and access to the Internet have had a positive impact, including burgeoning online communities such as PatientsLikeMe (<http://www.patientslikeme.com>). However, the ability to communicate in any fashion

eventually becomes threatened as patients become more limited, progressing from verbal communication to functional communication via an augmentative device to eye gaze systems, alphabet boards, and ultimately to a complete locked-in state.[78] The QOL for people with ALS at this stage, although uncertain and untestable[46] is regarded as not desirable by virtually all patients. In such advanced cases, it becomes impossible to determine the desires or even state of cognitive awareness of the TV user.[62]

As TV users in such a locked-in state are unable to indicate pain, depression, or the desire to terminate TV, an agreement should be reached before communication is lost as to an AD specifying the limits of TV. The physician and other caregivers involved in the management of ALS patients on TV must perform a role in the informed consent process at this juncture as most TV users will not specify limits of TV unless strongly advised to do so by their physicans.[64]

TV poses major challenges to family members: 58% of caregivers when interviewed felt TV was a major burden and 47% reported that their own health had suffered as a result of caring for their family members affected with ALS.[54] In addition, caregivers reported a loss of privacy in their homes and decreased time for outside friendships, as well as increased tension, depression, and anxiety.[79]

The medical relationships of ALS patients often change once TV is instituted as access to ALS care centres may be increasingly limited, especially if reliable transport and caregivers are not readily available. ALS patients may feel abandoned by the very centres that initially advised and encouraged TV. Patient care is often provided by home-based ventilatory teams headed by pulmonologists trained in invasive ventilatory management rather than the clinic-based multidisciplinary ALS team. This decreased access to ALS specialists should be discussed in advance of the decision to use TMV so that the new relationship may be anticipated and the impression of medical abandonment may be dealt with proactively.

Medical complications of TV are frequent and can be serious. In a prospective study of 354 consecutive TV patients, a total of 400 complications occurred.[80] These included problems related to tracheostomy—pneumothorax, bleeding at the stoma site, subcutaneous emphysema, nosocomial infections, tracheomalacia, tracheoarterial fistula, and tracheoesophageal fistula—as well as complications related to mechanical ventilation—pulmonary emphysema, pneumomediastinum, pneumopericardium, acute respiratory distress syndrome, venous air embolism, oxygen toxicity, and systemic haemodynamic instability.[81]

Yet TV is a viable and desirable option for some ALS patients who have a desire to continue living. The majority of such patients report contentment and happiness with their choice of TV.[46]

Factors predicting success

When assessing patients for home-based TV, characteristics that most favour a positive outcome include:

1 A highly motivated patient who is empathic to the needs of the family and engaged with the family and the community.
2 A reliable means of communication between the patient, the family, and medical caregivers.
3 A slowly progressive course of muscular paralysis.
4 A thorough understanding of the alternatives to TV.
5 A thorough understanding of the progression of ALS and the possible cognitive involvement.
6 A well-informed family that is willing and able to take on the burdens of TV.
7 Financial resources for equipment, caregiving needs, and multidisciplinary support.
8 An AD for discontinuing TV[70]
9 Capacity for both patient and the family to remain flexible and adaptable to constantly changing caregivers, equipment, and physical limitations.[82]

The length of hospitalization after TV varies, but in the absence of critical illness, most patients will be moved from intensive care to intermediate-level care within 21 days of tracheostomy.[83] The duration of hospitalization can be long, averaging 52 days in one study.[66] A multidisciplinary approach includes input from pulmonology, neurology, gastroenterology, nursing, respiratory, occupational, physical and speech therapy, nutrition, social work, and chaplaincy services. Discharge to home requires coordination between hospital and home care services, including respiratory care providers, as well as the outpatient clinics that will follow the patient. Assessment for home management of TV includes an assessment of the physical environment, financial, technical, nursing, and community support, and resources,[84] and the American Association for Respiratory Care clinical practice guidelines[85] offer excellent checklists and guidance for the management of the patient on TV in the home.

End of life

Death on TV is predominantly due respiratory disorders such as pneumonia or to medical complications including ALS-associated circulatory collapse and sudden death.[4,86] Some TV users choose to discontinue life support.[59,67] In most countries the decision to discontinue life support is viewed as ethically and legally equivalent to not initiating life support and is therefore permitted upon

the patient's request. Guidelines for the discontinuation of TV, including a step-by-step algorithm and medications to maintain the comfort of the patient, have been published in the United States.[87] Even where the ethical course is clear with regard to discontinuing TV, the practical course of how to do it is ambiguous. The family, although perhaps in principle in agreement with the decision, is always to some degree ambivalent about the role they play and certainly unable and unwilling to simply 'pull the plug', the patient cannot discontinue him or herself, and the clinician is not routinely accustomed to purposefully ending life. In some areas, hospice units may perform this service for the patient. The primary goal in withdrawing TV is to maintain patient comfort. In patients who are unable to communicate their comfort, tachycardia, tachypnoea, or restlessness may be indications of patient distress. Analgesia and sedation should be offered before, during, and after ventilator withdrawal.[88,89] Medications ideally should be given intravenously to bypass erratic gastrointestinal absorption due to poor tissue perfusion. Morphine is a preferred drug due to its wide therapeutic window, tendency to promote euphoria, dry airway secretions, and dilate pulmonary vasculature. An initial intravenous bolus of 5–10 mg may be followed by a continuous infusion of 50% of the bolus dose per hour. When the bolus dose needs to be repeated, the continuous infusion should be increased accordingly.[88] In addition, agitation may be managed by the addition of benzodiazepines. An initial bolus of 2–10 mg of midazolam, 2–4 mg of lorazepam, or 5–10 mg of diazepam should be followed by the same dose as continuous hourly infusion. In this way, pain, fear, and anxiety can be effectively relieved.[89] Supplemental oxygen and positive end expiratory pressure may be discontinued followed by conversion to a T-piece and spontaneous breathing.[90] Bedside nursing measures such as eliminating unnecessary procedures, removing unnecessary tubes, clearing secretions, and administering antipyretics when indicated are also extremely helpful, especially to family members.

Additional support is sometimes needed for the medical team involved in ventilator withdrawal and can be provided by reviewing the decision to discontinue TV in advance with an ethics panel comprising other colleagues, clergy, and lay people in the community.

Conclusion

Respiratory management is a key component in ALS care. Whether patients with ALS choose respiratory aids or opt for symptomatic management alone, the care of ALS patients is best managed collaboratively between disciplines. In addition to expert symptom management, palliative care professionals can offer support for decision-making as well as guidance for spiritual and psychosocial

care. Even patients opting for apparently life-sustaining therapy with mechanical ventilation need the services of palliative care to ensure best continuity.

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Chapter 7

Control of symptoms: dysphagia

Edith Wagner-Sonntag

Summary

Dysphagia is very common in people with ALS. The assessment comprises clinical examination and, for specific questions, instrumented methods such as videofluoroscopy and/or endoscopic evaluation of swallowing. Treatment of dysphagia in ALS patients includes an array of measures such as positioning, compensatory techniques, dietary modifications, and safety strategies. In certain patients with severe dysphagia, augmented feeding techniques, i.e. nasogastric tube feeding or feeding via a percutaneous endoscopic gastrostomy (PEG) or a jejunostomy, may be necessary. Many dysphagia-related symptoms like sialorrhoea, thick mucous secretions, and gastro-oesophageal reflux can be successfully treated pharmacologically. Palliative surgery, for example cricopharyngeal myotomy, is rarely indicated. The efficacy of palliative therapy can be assessed by measuring nutritional status, e.g. body mass index (BMI), and by use of specific dysphagia-related scales as well as quality-of-life scales. Management of dysphagia in ALS requires a multidisciplinary team approach and ongoing assessment: physicians, nursing staff, speech and language therapists, dieticians, the patients and their family should thereby closely work together.

Case history

Mr L, a 62-year-old active farmer, was admitted to hospital with speech and swallowing problems of unknown aetiology. Clinically he presented with rather unintelligible speech and a monotonous low voice. He reported difficulties with eating meat and bread, whereas drinking was quite normal. He had a weight loss of 5 kg within the last 2 months. The examination revealed a reduced tongue motility and tongue force with bilateral atrophy and fibrillations. Sometimes choking occurred on saliva with a markedly reduced reflexive cough. Mealtimes were very long and Mr L. frequently stopped eating because of fatigue and loss of appetite. The diagnosis of ALS had been confirmed. Videofluoroscopy and fibreoptic endoscopy revealed a considerable risk of aspiration with reduced and ineffective cough on food as well as on thin liquids. Therefore, soft-textured food with a caloric supplement and sipping of thickened liquids was recommended. The patient was informed about the advantages of early insertion of

a PEG tube and the use of this type of nutrition as a supplement, especially when working. He decided to begin with dietary modification. Three months later, after losing another 7 kg in weight, he asked for a PEG to be inserted. Because his FVC was at that time below 50% of predicted, a percutaneous radiological gastrostomy (PRG) was inserted and some weeks later he reported a good experience with the new additional feeding.

Introduction

Eating and drinking are two of the most basic functions of life, so progressive loss of the ability to eat and drink has a huge impact on quality of life, affecting both the patient and their family. The assessment of these swallowing problems and their management is crucial in the patient's care and requires a multidisciplinary approach.

Poor nutrition and weight loss are common in people with ALS and have functional consequences which may add to the disability caused by the underlying disease, leading to both mental and physical deterioration, increased complications, and impaired quality of life. Malnourished patients have a seven-fold higher risk of death.[1] The prevalence of malnutrition varies between 16 and 55% in patients with ALS, 21% being moderately to severely malnourished, including those with no apparent swallowing problems.[2,3]

Frequency of dysphagia in ALS

The site of significant neuronal loss and the duration of the disease will affect the development of dysphagia. The reported prevalence of dysphagia in ALS is highly variable, varying between 48 and 100%.[4]

As far as disease duration is concerned, dysphagia appears on average 4 months after onset of the disease,[5] but during the course of the disease nearly 100% of ALS patients have problems due to oral or pharyngeal involvement.[6] In the later stages of ALS dysphagia is a very common symptom, and aspiration pneumonia, either on its own or in conjunction with respiratory insufficiency, may lead to death in ALS patients.[7]

A report from St Christopher's Hospice on 124 patients with ALS found that only 21% of patients were able to swallow all foods, 42% semi-solids only, and 27% liquidized or puréed foods; only 8% of this series of patients required tube feeding. Moreover there were problems with patients' feeding: only 19% of their patients were able to feed unaided and a further 24% were only able to do so with specially prepared food of modified texture, with the remaining patients all needing help with eating.[7] Another hospice study found that dysphagia was a problem for over 90% of patients with ALS.[8]

Pathophysiology

The three anatomical levels which may be affected in ALS are associated with the development of dysphagic symptoms:

1 UMN involvement causes supranuclear symptoms, also termed pseudobulbar palsy.
2 LMN involvement of the cranial nerve nuclei in the medulla oblongata and pons innervating the muscles of the jaw, face, tongue, pharynx, and larynx affects chewing, swallowing, speech, and voice with subsequent bulbar symptoms.
3 The motor neurons of the spinal cord can be affected and this may lead to dysphagia as the result of progressive respiratory dysfunction. Regardless of the onset being bulbar or non-bulbar, dysphagia progresses as respiratory function declines.[9,10]

Dysphagic symptoms may develop as a result of neuronal loss at any of the three levels—supranuclear, bulbar, or spinal—which are often affected in combination.

Since the swallowing process deals with two components—saliva and nutrition—patients have to manage both in order to avoid complications. At the onset of the disease there may be problems in coping with liquids or saliva. Many patients report that they produce too much saliva, but it seems that there is no measurable increase in secretion rather a difficulty in handling the saliva, which creates the impression of too much secretion being produced.[11] Therefore, the term sialorrhoea should be preferred to hypersalivation. With increasing loss of muscle tone and muscle strength in lip closure patients tend to drool. The inability to keep the lips closed leads to increased breathing through the mouth and this can tend to thicken the oral secretions, making them difficult to swallow.

As the disease progresses, a feeling of fatigue when eating arises as the motility and strength of the facial and oral/lingual musculature decrease. This leads to difficulties in the oral preparation, mastication, and oral transport of food. Weak muscles make it difficult to manage thin liquids and dry and crumbly food. The swallowing reflex becomes difficult, causing the subsequent problem of aspiration,[12] and food spills downward before laryngeal closure can be initiated. With increasing weakness of laryngeal adduction, the mechanism of compensating the late triggering of the swallowing reflex is no longer sufficient and laryngeal decompensation occurs with the ensuing risk of aspiration. Aspiration is defined as passage of material into the larynx below the level of the vocal cords, whereas penetration means passage of material into the larynx above the

glottic level. At this stage it is crucial that the patient has a forceful cough reflex, as the less effective the cough, the more dangerous oral intake becomes. Reduced tongue force, delayed triggering of the swallowing reflex, and weak elevation of the hyoid and larynx result in reduced opening of the upper oesophageal sphincter. This leads to retention of saliva, food, and liquids in the valleculae and piriform sinuses, also aggravating the risk of aspiration. A recent study showed that pressure profiles of the oesophageal sphincter are not disturbed in ALS patients.[13]

Laryngeal abduction and/or adduction may also be affected and worsen dyspnoea. In some cases, therefore, it will be necessary to discuss whether a patient could benefit from a tracheostomy (see the section on Surgery).

Assessment of dysphagia

A multidisciplinary approach is necessary for the assessment of dysphagia, including cooperation between neurologist, ear, nose, and throat specialist, speech and language therapist, dietician, physiotherapist, and occupational therapist. The clinical examination includes history taking, the examination of swallowing ability, the evaluation of caloric intake and of the BMI, and respiratory function (e.g. vital capacity). During the assessment the speech and language therapist will examine the structures involved in swallowing, observe the patient's ability to manage food and drinks, define aspiration risks, plan therapeutic strategies to maintain a sufficient and secure oral intake, and establish a current diet which the patient is able to manage. This assessment should be ongoing throughout the progression of the disease at defined intervals.

The following aspects of the oral phase of the swallowing process can be directly observed:

- Lip function: the ability to maintain a lip seal, which is important for the oral retention of food and fluid. A good lip seal also permits a positive pressure within the oral cavity, allowing initiation of the swallowing and facilitating laryngeal elevation. The ability to take food from a utensil and to use a straw should be assessed.
- Tongue function: the strength, rate, coordination, and range of tongue movement, from which inferences regarding the ability to collect and control a bolus within the mouth and to effectively push the bolus into the pharynx can be made. The efficacy of tongue function can also be observed by noting the presence of oral residue after swallowing. Articulatory abilities are also affected by tongue movements, so the presence of dysarthria is a sign of reduced speed or range of movement of the tongue.

- Pharyngeal function: while the pharyngeal phase cannot be directly observed, information can be inferred through observation of swallowing. Coughing before, during, and after swallowing may indicate that penetration or aspiration is occurring. It must, however, be remembered that some people do cough insufficiently or stop coughing because of weakness of the laryngeal closure and respiratory musculature. Penetration or aspiration without any subsequent cough is termed silent. Since there is usually no sensory pharyngolaryngeal deficit in ALS patients, silent penetration or aspiration is caused by the weakness of the laryngeal or respiratory muscles, as described in the section Pathophysiology. The most effective way of ascertaining that aspirated material is cleared effectively through a cough is by videofluoroscopy or a fibreoptic endoscopic evaluation of swallowing. The timing and degree of laryngeal elevation during swallowing can be grossly estimated by placing one's fingers on the suprahyoid muscles, the hyoid bone, and the larynx and noting the approximation of these structures during a swallow.

Videofluoroscopic and endoscopic swallowing evaluation

Videofluoroscopic swallowing study (VFSS) is an important technique in the evaluation of swallowing disorders for its ability to reveal insights into oral, pharyngeal, laryngeal, and oesophageal abnormalities. The coordination of the swallowing events, the presence of penetration and aspiration, as well as the efficiency of the cough reflex can be detected during the investigation. Based on these results, a treatment plan for swallowing therapy can be established and the decision made as to whether to provide dietary modification or alternative feeding methods.[14,15]

Because of the frequent finding of respiratory dysfunction and the high risk of aspiration in ALS patients with dysphagia, a modified barium swallow (MBS) may be dangerous with respect to aspiration pneumonia[16] and should, if at all, only be performed in patients with minor dysphagic symptoms. As hyperosmolar contrast media such as Gastrografin entail the risk of provoking life-threatening lung oedema if aspiration occurs,[17] a water-soluble, isosmolar contrast agent with no significant side-effects even in the case of aspiration may be preferred.[18]

Transnasal fibreoptic endoscopic evaluation of swallowing (FEES)[19] allows direct visualization of the pharynx and larynx and can also be helpful in the assessment of swallowing. In some patients with hyper-reflexive reactions the method cannot be performed without difficulty. It may, for example, produce excessive saliva or bring the patients to constant sneezing.

VFSS and FEES are complementary methods and both have advantages and disadvantages; real food can be used in both examinations.[20] FEES is normally well tolerated by the patient, it is more portable than VFSS, and can even be performed in bedridden patients. VFSS provides an image of the entire duration of the swallow and allows viewing of the complete oral and pharyngeal tract including the upper oesophageal sphincter. VFSS and FEES recordings are useful in the education of patients and carers as they reveal the process of swallowing very clearly and elucidate the risks involved in oral nutrition. Compensatory strategies and the benefits of modifications to the feeding process can be tested and demonstrated.[21]

Management of dysphagia

Positioning

The patient should be in a comfortable position while eating and drinking, usually in an upright position. Patients with ALS may require extra support as the effort may result in an unwanted increase in tone or fatigue. The involvement of a physiotherapist can be very helpful.

Compensatory techniques

There are several compensatory techniques that can aid patients' swallowing and reduce the risk of aspiration:

- Supraglottic swallowing is a technique which helps to close the vocal cords during swallowing. The patient holds his or her breath while swallowing and exhales at full force immediately afterwards.[22] Penetrated food or secretion can be expelled from the laryngeal vestibulum by this technique in order to avoid aspiration. This technique is recommended when laryngeal closure becomes weak and triggering of the swallowing reflex is delayed, and it is appropriate for patients with minor oral, laryngeal, and respiratory dysfunction.
- The Mendelsohn manoeuvre is a technique which helps to open the upper oesophageal sphincter and prolong its opening time. The patient has to hold the upward movement of the larynx during swallowing for about 2 s. This technique is especially appropriate for patients with deficient opening of the upper oesophageal sphincter. In ALS patients where forceful movements become a problem, the Mendelsohn technique may be performed more moderately, since it should not distress the patient.
- Postural changes can be helpful. Patients with pseudobulbar symptoms or impaired tongue movements (and resulting difficulty initiating a swallow)

but with intact pharyngeal phase of swallowing can tilt the head backwards to guide the bolus into the pharynx. Patients with bulbar symptoms, with difficulty in triggering the reflex and premature spilling of liquids can tilt the head forward, known as the 'chin tuck'. In order not to drool food or liquids patients should be encouraged to seal their lips with their hand.

Although neuropsychological disturbances may occur in ALS, they are in many cases only subtle and undetected without comprehensive neuropsychological testing[23] (see Chapter 8). Therefore, for many ALS patients the cognitive prerequisites for adopting these strategies are good, even in the late stages of the disease.

Exercises

There are no specific studies showing that the use of exercises to aid swallowing is of benefit to patients with ALS. It has not been shown that exercises improve swallowing function and it has been suggested that exercises may merely exhaust the weakened muscles. Anecdotal reports show that mild to moderate exercise may preserve range of motion for a while. For a small proportion of patients with slowly progressing disease exercises may be of limited benefit in maintaining function. Exercises may be helpful in lip closure and tongue movements.

Patients may be encouraged to improve their swallowing rate by learning to swallow before trying to open the mouth or to speak. A simple timer can be used, which reminds them at an individually determined interval to swallow (for example every minute) following a 'beep' signal.

Since fatigue is a very common symptom which occurs frequently while performing exercises,[24] short therapy sessions several times a day with resting periods are preferred.

Dietary modifications

As eating becomes more of a problem the patient should also be seen by a dietician so that an assessment of their intake can be made and advice given on how to enrich meals with calories, proteins, and vitamins. Energy intake should correspond with patients' changing requirements during the progression of the disease.[25] If a patient's eating is slowed it may be that they could adapt by taking longer over meals. In practice, people do not do this, possibly because slow eating induces earlier satiety but also because they are embarrassed to take longer than others and curtail their meals accordingly. Increasing messiness with eating is also a source of embarrassment, and may cause people to avoid eating in company, or miss out items which cause them problems.

Dietary modification may be necessary with the aim of maintaining nutrition when chewing becomes more and more difficult and frequent episodes of aspiration or choking occur. It can also prevent extensively long mealtimes, fatigue, and dread of meals. Soft textures or puréed food can compensate for a poor oral preparation phase and ease oral and pharyngeal transport. Liquid supplements may be helpful, but beyond a certain stage there tends to be choking, especially with thin liquids, and patients find thickened drinks easier to manage. Food can be pureed or liquidized and mixed with commercial preparations such as Thick & Easy™ or Nutilis, which make a semi-solid consistency that is easier to swallow. Puréed food can also be made much more attractive by reconstituting it into its original shape and appearance by using thickeners and moulds. For example, meat, vegetables, and potatoes can be given an almost normal appearance by liquidizing the different components, mixing them with thickener, and then moulding them on the plate in their original form, which retains the visual and pleasurable aspect of the food. Experience has shown that semi-solid food presented in this way is much more likely to be eaten than if it is presented in the form of a dull-coloured purée, like the weaning foods used for babies.

Patients and relatives need instruction on how to prepare appropriate meals, like high-caloric food or specific textures that are easy to chew and swallow (including thickening of liquids to avoid dehydration). Another problem can be abdominal weakness and failure of glottic closure, which can lead to constipation. In these cases dietary fibre has to be added. Triggering of the swallowing reflex can be enhanced by emphasizing taste or temperature. Cooled drinks are often easier to swallow.

A recent study showed that dietary modification is efficient in more than 90% of patients with mild to moderate dysphagia.[26]

Rate of intake

Time must be allowed for the weakened muscles to coordinate their movements to complete swallowing and recover during meals. There is often a restriction on the size of bolus that can be tolerated by the patient. The task of feeding the patient can be tedious or cause trepidation, and the speech and language therapist may need to spend time training carers, both family members and healthcare professionals, in the skills required to safely feed someone with dysphagia. It is also essential to ensure that food remains hot and palatable and retains the correct consistency during the entire meal.

Safety strategies

During mealtimes patients should avoid distractions such as conversation, television, radio, or other noisy and stress-inducing situations. When patients show

a significant level of fatigue, they are advised to eat several small meals a day. If episodes of choking occur, the Heimlich manoeuvre can be applied by the carers.

Augmented feeding techniques

As dysphagia becomes more severe other feeding techniques may need to be considered. These techniques can also being used as a supplement while the patient is still able to eat and drink.[27] They can enhance quality of life by avoiding long mealtimes and preserve time to perform other activities (see Case history section). These decisions will need careful discussion with the patient and family and each patient will need an individual assessment and resulting management plan.

Nasogastric tube feeding

Fine-bore nasogastric tubes can be used for feeding, at least in the short term. There are problems of the tube becoming displaced and the tube is very obvious and may be uncomfortable for the patient and family. Nasogastric feeding also appears to be associated with increased risks of ulceration and aspiration pneumonia[28] and there may be an increase in the amount of oropharyngeal secretions with the presence of a nasogastric tube.

Gastrostomy and jejunostomy

The fine-bore PEG technique may be offered to patients with ALS quite early in their disease, since the risks associated with insertion are much less at this stage and the gastrostomy can be used just for supplementation. When respiratory muscle weakness has supervened the procedure is much more hazardous, as mild sedation is necessary.[29] It is important to try and avoid putting the high-risk patient to unnecessary hazard, and also to be certain that the patient and their family really want it and understand its full implications. The decision to consider a PEG should only be made after careful discussion with the patient and family. It has been suggested that the discussion should be undertaken soon after the onset of dysphagia and when the patient's FVC is still over 50% of predicted to reduce the risks of the procedure.[30]

The procedure is not without risk in patients earlier in the disease. The mortality of this procedure is less than 1 in 100, particularly from problems such as pneumonia. Clinical contraindications to this technique are abnormal coagulation, portal hypertension or ascites, gastric outflow obstruction, previous gastric surgery, major cardiorespiratory problems, and *Clostridium difficile*-associated diarrhoea.

Although major complications such as peritonitis and cellulitis have been described, complications are usually minor and related to local skin infections, reflux, or mechanical problems with the tube, e.g. blocking or fracture of the hub. The technique of inserting a PEG has been described widely.

Jejunostomies have been used successfully in patients with neurological dysphagia. The advantage of this technique is that food is put directly into the jejunum, overcoming the problems associated with reflux of food into the pharynx. Unfortunately the insertion of a needle-catheter jejunostomy necessitates a general anaesthetic with a mini-laparotomy for its insertion. Alternatively, where reflux has been a problem, the PEG technique can be used, threading a fine-bore jejunal tube through the gastrostomy, round into the jejunum, under endoscopic guidance.

The insertion of a PRG does not require sedation.[31] It may be considered when the respiratory reserve of the patient is such that the insertion of a PEG may be considered to be very difficult and with increased risk of morbidity or mortality. NIPPV with oxygen support can be administered during insertion of a PEG or PRG in patients with a FVC of less than 50%.[32]

Nourishment via a PEG has been shown to improve both the nutritional state of the patient and survival.[33] A study of 35 ALS patients showed that there was an improvement of BMI in patients with a PEG, whereas there was a reduction for controls. After 6 months the mortality of PEG patients was lower than in the control group.[34] Other studies have shown a probable improvement in the quality of life, and the majority of patients maintained or improved their weight.[33]

Ethical concerns have been raised about the use of augmented feeding techniques, as there is the risk of extending life when the quality of the life is deteriorating and the patient does not wish to continue.[35] The patient and family need to be carefully involved in the decision about the insertion of a PEG, and it is essential that there is adequate explanation of the implications of the procedure as well as the risks of not having the procedure, both on the quality and length of life. It is also important to ensure that the patient and family can cope with the procedures involved in feeding through the tube and that there are facilities and support for them in the community.[27]

Management of sialorrhoea and mucous secretions

Patients may experience problems with their oropharyngeal secretions. Although the total amount of saliva produced remains the same, sialorrhoea develops as swallowing is reduced. The patient should be reminded to swallow before trying to open their mouth or speak and to be aware of frequent swallowing to keep the mouth free of saliva.

Patients may complain of dryness, particularly on waking, but drooling may occur later in the day as fatigue increases or after meals. Depending on the nature of the problem different management approaches may be used. If the mucous secretions are thick and ropy, sipping fruit juices, especially dark grape juice, can be beneficial. Sipping water may be sufficient for dryness, but later in the disease artificial saliva may be required. Special, enzyme-containing dental-care products like mouthwash and tooth paste, without alcohol and mint, or oral moistening gel may also be of benefit.

Pharmacological interventions

Reduction of salivary flow in ALS patients with drooling is possible with transdermal scopolamine lasting 24–72 h[36] and other anticholinergic medication, such as atropine, or antidepressants, such as amitriptyline. Injection of botulinum toxin into the parotid and submandibular glands is also well tolerated and may be helpful for sialorrhoea or hypersecretion and reduce these socially disabling symptoms[37] (see Chapter 9).

If swallowing is disturbed because of thick mucous secretions, *N*-acetylcysteine may be helpful,[38] although it is of limited value for many ALS patients with a weak cough pressure (in such cases a mechanical insufflator–exsufflator like Cough Assist® is described as being very helpful to get rid of the pulmonal secretions).

Gastro-oesophageal reflux disease is frequent and may aggravate or even cause swallowing and respiratory problems.[39] Therefore, in ALS patients suffering from reflux, proton pump inhibitors such as omeprazole should be administered.

Surgery

Palliative surgery in ALS patients with dysphagia should be considered only in certain cases, and then with caution, since surgical trauma may contribute to neuronal death and a more progressive course of ALS.[40] Furthermore, in general anaesthesia the rate of perioperative mortality has been reported to be high.[41] Among surgical procedures, the efficacy of cricopharyngeal myotomy remains controversial. There are several prerequisites for a cricopharyngeal myotomy:[42]

- cricopharyngeal dysfunction
- normal elevation of hyoid and larynx
- swallowing therapy (e.g. by use of the Mendelsohn manoeuvre) not successful in achieving the opening of the upper oesophageal sphincter
- pharyngeal pressure sufficient to propel a bolus through the open sphincter
- there should be no gastroesophageal reflux.

All these prerequisites occur rarely in ALS patients, and a recent study showed that pressure profiles of the upper oesophageal sphincter are not disturbed in ALS.[13] Therefore, for the majority of ALS patients treatment of dysphagia by cricopharyngeal myotomy is inappropriate.

In order to avoid irreversible myotomy, injections of botulinum toxin into the upper oesophageal sphincter are an alternative,[43] but this procedure shares the same prerequisites as cricopharyngeal myotomy. Since a general weakness has been described in a patient with ALS after focal botulinum toxin injection,[44] this procedure cannot be generally recommended.

For ALS patients who cannot swallow their own secretions safely, the possibility of tracheostomy may be discussed with the patients and relatives[45] (see Chapter 6).

Assessment of efficacy

Measurement of nutritional status plays a pivotal role. For this purpose, measures such as dietary histories and BMI can be recommended, in order to avoid malnutrition, i.e. BMI <18.5 kg/m^2. Dysphagia-specific scales such as the bulbar section of the Norris Scale, the ALS Functional Rating Scale-R (ALS FRS-R) bulbar sections and the Dysphagia Outcome Severity Scale (DOSS) can also be used.[46] For assessment of dysphagia-specific quality of life, the SWAL-QOL and SWAL-CARE are also suitable outcome tools.[47]

Conclusion

Attention to eating and drinking is part of the optimal care of all patients. When normal mechanisms begin to fail, the help of a speech and language therapist, occupational therapist, physiotherapist, and dietician should be sought to help the patient overcome mild to moderate degrees of activity limitation. The adoption of special techniques may aid swallowing, as may the provision of attractive food of semi-solid consistency. Adaptive strategies like positioning and using equipment that can be handled easily by the patient also support oral intake. As dysphagia becomes more advanced, consideration should be given to the use of augmented means of feeding by gastrostomy, jejunostomy, or nasogastric tube, but always with the full consent of the patient and the cooperation of family and carers. Nutrition is only one part of the overall management of patients with this distressing condition, which requires a multidisciplinary team approach.

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Chapter 8

Control of symptoms: cognitive dysfunction

Laura H. Goldstein

Summary

In addition to the relatively small percentage of ALS patients in whom a frontotemporal-type dementia may occur, a significant percentage of non-demented patents may show mild to moderate cognitive change, predominantly of a dysexecutive nature. Memory deficits may occur and, increasingly, language involvement has been noted. Disease-related factors, mood disorder, and medication may compound cognitive impairment and need to be accounted for when assessing cognition. There has been no research into the effectiveness of psychological interventions for cognitive dysfunction in ALS, although cognitive neuropsychological rehabilitation techniques may be applicable. The development of interventions for behavioural changes that may occur in ALS dementia may be guided by approaches used in other neurodegenerative diseases, especially frontotemporal dementias (FTDs). People with ALS should have access to clinical neuropsychologists who can assess the presence of cognitive dysfunction, undertake behavioural assessments, and work alongside other members of the clinical team to ensure the best provision of care for the person with ALS and their carers.

Case history

Mr MK is a 55-year-old man who had been diagnosed with ALS 2 years earlier. He is married with three children. He had recently become severely dysarthric but was now using a Lightwriter, although not particularly efficiently. His verbal output using this, and also through writing, was becoming increasingly limited, although he was physically able to write in a legible manner. His family reported that he appeared not to be remembering what they were telling him about recent events, seemed rather impulsive in his attempts to swallow food, and was not able to follow the speech and language therapist's advice regarding safe feeding.

A clinical neuropsychology assessment suggested that Mr MK was experiencing impairments in a number of cognitive domains, which had implications for his everyday functioning. Thus his family's impression of his poor memory was borne out on testing, although assessment also highlighted his difficulty in understanding more complex grammar, which

may have also led them to think that he was not remembering what they said to him when he may simply not have understood them in the first place. He may also have had trouble trying to divide his attention between several people telling him different things which they expected him to remember. The perseveration observed during testing was also felt to contribute to his difficulties using his Lightwriter and, particularly importantly, also eating safely. His difficulties in naming and word generation during testing also substantiated the everyday impression of his reduced verbal output when writing/using his Lightwriter.

The cognitive deficits observed during testing were explained to Mr MK and his family so that they could improve how best to communicate with him, and were also relayed to the other members of the team working with Mr MK so that they could modify the complexity of the instructions given to him, reduce how much they expected him to remember without writing things down for him, and address, in particular, the guidelines given to him regarding eating.

Introduction

Despite the traditional view of ALS as a neurodegenerative disease that does not impact on cognitive functions, convincing evidence has now accumulated that, in at least some people, a detectable degree of cognitive involvement is present.[1] This can vary in magnitude and, at worst, there may be a frank dementia. This chapter will document the nature of this cognitive involvement with comments on some of the practical implications of such compromised cognitive functions.

It is also important to note that many people with ALS and their carers are not aware of the potential for cognitive impairment. How professionals communicate such possible consequences of ALS may be particularly important, as people's own anticipation of being cognitively impaired has the potential to influence their future treatment choices.[2]

ALS and dementia

Early suggestions were that in approximately 3% of sporadic cases and 15% of familial cases of ALS a FTD might occur.[3,4] However, more recent estimates suggest a higher prevalence of dementia in sporadic ALS[5,6] due to improved diagnostic criteria for FTD. In such cases a clear breakdown in behaviour, personality, and cognition occur, reflecting frontal dysfunction. Cognitive, personality, and behavioural difficulties usually, but not always, precede the physical signs of ALS and approximately 10% of people with FTD then develop ALS.

Where neuropsychological assessment is possible, impaired abstract reasoning, rigid thinking, reduced verbal fluency, poor response inhibition, impaired attention, reduced naming, and a non-fluent dysphasia have been reported:

memory is variably affected but more posterior functions are thought to remain unaffected (see, for example, work by Neary et al.[3], Peavy et al.[7], and Vercelletto et al.[8]).

Although the practical implications of dementia for people with ALS have received little attention, the development of socially inappropriate and other problem behaviours, characterized in some cases as disinhibited, jocular, impatient, gluttonous, and with stereotypical gestures,[9] may produce disruption to everyday life and to the care provided. Schulz et al.[10] reported different subtypes of FTD in their patients with sporadic ALS: the majority were characterized by dysexecutive behaviour typical of lateral frontal involvement, whilst small numbers demonstrated disinhibited behaviour, primary progressive aphasia, or semantic dementia.

ALS aphasia

Bak et al.[11] described six patients with ALS in whom communication problems developed at an early stage. They all developed a progressive non-fluent aphasia and five patients were also found to have deficits in syntactic comprehension. Interestingly, comprehension and production of verbs were consistently more affected than for nouns, and this relative deficit persisted. The classical physical signs of ALS developed over the following 6 to 12 months. The behavioural symptoms ranged from mild anosognosia to personality change implicating frontal-lobe dementia, confirmed in three cases post mortem. Bak et al. support other views[12] that whilst the existence of ALS dementia has been well recognized, the incidence of a possible ALS aphasia is possibly underestimated and can occur in the absence of the 'frontal' features characteristic of ALS dementia.

Behavioural impairment without dementia

A number of studies suggest that a significant proportion of people with ALS may demonstrate evidence of behavioural change, representing a subclinical form of the behavioural variant of FTD. This has been referred to as ALS behavioural impairment (ALS-bi)[13] and has more recently been defined[14] as being present when the person has two or more non-overlapping supportive diagnostic features derived from established criteria for behavioural variant FTD.[15,16] In terms of the range of behavioural changes that might occur in ALS cases, a carer-based interview[17] indicated that complaints about the person with ALS from caregivers included:

- self-centredness/selfishness (69%)
- irritability (63%)

- altered sensory behaviour (exaggerated sensory/heat/cold response and/or reduced pain awareness) (50%)
- apathy/reduced interest in activities (38%)
- blunting of emotion (25%)
- loss of embarrassment (19%)
- loss of concern for hygiene (19%)
- repetitive behaviours, compulsions, rituals (19%)
- socially disinhibited behaviour (13%)
- lack of awareness of/concern for own illness (13%)
- increased aggression (13%).

In addition, altered eating habits were very occasionally observed. Some changes were more characteristic of the semantic dementia subtype of frontotemporal lobar degeneration, but the most prominent features were more characteristic of the behavioural variant subtype.

When trying to classify patients as demonstrating ALS-bi there is a need to ensure that the behavioural change is new and disabling but cannot be better accounted for by physical limitations imposed by the disease. Information must be sought from at least two sources, including interviews with carers, observations of the person with ALS, and/or the completion of standardized questionnaires (which have been reviewed elsewhere[1,18]).

Cognitive impairment without dementia

The emphasis over the past 25 years or so on the potential compromise of cognitive functions in ALS has arisen out of, and in turn encouraged, a range of formal neuropsychological studies of people with ALS.[1,19] There has been some concern that clinic-based samples that have recruited highly selected patients may yield results that are not representative of the disease as a whole and that population-based samples give more reliable estimates of the prevalence and nature of impairment associated with the disease.[1] However, these approaches seem to give rise to similar findings.

It is possible that for many people the cognitive deficits are sufficiently subtle and only revealed on formal neuropsychological testing, but in other cases aspects of changed everyday behaviour may suggest compromised cognitive ability. Whether such deficits are detected may be influenced by whether a clinic routinely refers all people with ALS for neuropsychological assessment (resources allowing) or does so only if cognitive deficits are suspected. From a practical viewpoint, the physical care required by someone

with ALS may detract from more subtle cognitive changes, which might otherwise be ascribed to mood changes or to other psychological reactions to the illness.

Executive functions

The most consistently reported cognitive difficulties for people with ALS have been in what are referred to as executive functions. These are functions typically defined as being involved in the organization and planning of behaviour, mental flexibly, the ability to switch between tasks and divide attention, to follow rules and inhibit responses when appropriate, think in abstract rather than only in concrete terms, and also to be able to generate responses fluently.

A range of neuropsychological tests assess these abilities. Different studies have yielded a consistent pattern of results whereby people with ALS, as a group, appear impaired in their verbal fluency, i.e. their ability to generate rapidly written or spoken words. The most rigorous approach here[1] has been to control for the writing or speaking speed involved in the test so that the impairment is not simply due to the physical limitations imposed by the disease (e.g. Abrahams et al.[20,21]). Although the most consistent findings have been for tasks requiring the generation of words beginning with predetermined letters,[20–23] the findings apply more variably to semantic categories.[21,24,25] Difficulties with fluent generation of responses may not just be limited to verbal material (e.g. Abrahams et al.[21]). Taken together these findings indicate potential difficulties associated with the rapid generation of responses, and that these are associated with the thinking time involved in the generation of such responses and cannot simply be explained on the basis of slowed speech or impaired writing skills.

Deficits have also been found on a range of other executive function tests. Thus impairments have been found on a task of reasoning involving the logical sequencing of pictures to tell a story,[26] the ability to determine a rule in a task, follow it, and then flexibly find new rules when the task changes,[20,27–30] and also to think sufficiently flexibly in order to generate random sequences.[20,30] In addition, evidence for rather poor planning and impulsive responding has been reported.[20]

Tests of verbal and visual attention may elicit deficits, and it is possible for people with ALS to have difficulties with focused and/or divided attention.[6,28,30–32] Deficits in reasoning, mental flexibility, and attention, together with possible memory deficits (see Memory) may influence patients' ability to deal with changes to their lifestyle and adhere to new treatment options, which may cause potential difficulties for care provision and uptake.

Memory

Deficits in memory in people with ALS have been reported, but generally these difficulties are found less consistently than difficulties with executive function, and the very wide range of memory tests employed in the different studies also limits between-study comparisons. Where deficits have been reported, they have included difficulties largely reflecting weak encoding rather than poor subsequent storage.[33] In addition experimental studies have suggested impairments in working memory.[25,33–36] In terms of remembering verbal material in a situation that is perhaps more similar to that in everyday life, some deficits have occasionally been reported in the recall of short stories (e.g. Ringholzet al.[6]). The potential attribution of memory difficulties in people with ALS to weak encoding of the material to be remembered may imply that care needs to be taken to ensure that the person has encoded the material sufficiently well in the first case to be able to recall it later on. Clearly this has implications for the assessment of people's understanding of their treatment options and ultimately for determining their capacity to consent to treatment.

Language

The potential for detecting an impairment in spoken language in people with ALS is clearly more difficult where the person has pronounced bulbar involvement, but nonetheless a routine comprehensive language assessment for the person with ALS has been recommended.[37] This would seem particularly advisable if there is any doubt as to the person's ability to comprehend what is said to them. Accumulating evidence adds support for language impairment in ALS, although early studies suggested that language impairments were found in only a relatively small number of cases.[26,38–40] In studies with limited assessments of language, some evidence of deficits in confrontation naming (i.e. naming line drawings of objects) has been reported.[25,26,28,38,39] In our own recent studies, whilst object naming scores were lower than those for healthy controls, the patients' scores were not clinically impaired[22,41] and we have not always detected naming deficits.[21,23]

More recently, however, our study of 51 patients with ALS has indicated that language impairments may be detected on a range of measures not limited to naming.[42] While no difference between patients and healthy controls was found on The Pyramids and Palm Trees Test, the patient group made more errors on The Kissing and Dancing Test, suggesting a pattern of impaired verb as opposed to noun processing, as was previously reported for patients with ALS aphasia.[11] The ALS group also showed impairments, relative to the controls, on measures of synonym judgement, single word and sentence comprehension, lexical access

(i.e. detecting real words as opposed to non-words), and spelling. Forty-nine per cent of the ALS group showed impaired performance on single-word comprehension, and 35% on sentence comprehension.

Visuoperceptual and visuospatial functions

Although less emphasis has been placed on examining visuoperceptual functions, it would generally appear that a wide range of measures has elicited intact performance[23,25,26] but inconsistent findings have been reported for The Judgement of Line Orientation Test[22,25,28] and deficits have been found on the Motor Free Visual Perception Test.[40] The reported visuoconstructional deficits have not taken into account possibly confounding motor deficits.[25]

Emotional processing, theory of mind, and social cognition

A relatively new line of enquiry, relevant to the detection of memory changes in ALS, has begun to examine whether people with ALS show the typical profile of being better able to remember material that is emotional rather than neutral in valence. Papps et al.[43] demonstrated that a group of people with ALS did not show this typical pattern when asked to recognize previously seen words with either emotional or neutral valence. Instead they recognized a similar number of emotional words and more neutral words than control participants. However, a more recent study did not find similar results.[44] In addition, patients with ALS have been shown to demonstrate impaired performance on a test of affective decision-making by failing to adjust their responses on the basis of reward schedules on the Iowa Gambling Task.[45] Therefore it may be important to consider whether an alteration in the processing of emotionally salient material might influence people's reaction to their illness and what they remember of the information they are given about it, since other research also suggests that people with ALS may respond differently (more positively) from healthy controls to socio-emotional stimuli.[44,46]

Impairments have also been detected on tests requiring participants to infer the mental states of other people (i.e. theory of mind) using a variety of test materials that include cartoons, stories, and accounts of potential *faux pas* about which judgements have to be made,[47] judgements about social contexts,[48] and also of preferences based on the direction of eye gaze.[45]

In terms of socially salient material, people with ALS have also been shown to have difficulty recognizing emotions depicted in facial expressions,[45] in judging the 'approachability' of people from their facial expressions,[49] in inferring mental states from eye expressions,[45] and in judging emotions from sound clips (i.e. tests of prosody).[50] This may also be evidence that patients with ALS have

reduced empathy for others, which can have a significant effect in increasing the stress for carers, both family and professional.

Summary of cognitive deficits in ALS

In terms of everyday manifestations of cognitive impairment, then, it is possible that the following may be apparent:

- difficulty with generating thoughts/words—which may be manifest in limited conversation and poor word-finding—not to be confused with dysarthric speech
- difficulty planning activities
- difficulty switching from one idea to another and perseveration (i.e. continuing to do something even when it is no longer appropriate to the situation)
- attentional/concentration difficulties, possibly leading to distractibility
- impulsivity
- forgetfulness/problems with learning new things
- language difficulties, especially single word and sentence comprehension and naming/word-finding
- difficulty judging emotions and mental states in other people, potentially leading to misunderstandings or misinterpretations of social situations.

Not all of these changes, if any, will necessarily be present in the same person, and they may be obscured by speech and other motor difficulties. It is also important to note that certain changes in cognition occur as a result of the normal ageing process and in relation to anxiety and depression, so a careful clinical evaluation will be necessary in order to determine the significance of any observed changes. In addition, changes relating to behavioural difficulties may be more prominent than performance on specific cognitive tests. The everyday implications of identifying and characterizing cognitive dysfunction in someone with ALS are illustrated in the case history presented at the start of the chapter.

How common is cognitive impairment in ALS?

Ringholz et al.[6] noted that the prevalence of cognitive impairment in people with ALS has been estimated at between 1 and 75%. However, the samples studied have varied in size, and a range of cognitive measures has been employed. An earlier study of 146 patients from the same centre found that 35.6% of patients were cognitively impaired (with impairment defined on the basis of

performance at or below the fifth percentile on at least two out of eight neuropsychological tests administered).[28]

Since differing classifications of cognitive impairment may clearly lead, even within the same sample, to differing prevalence estimates, consensus criteria[14] have attempted to define cognitive impairment in patients with ALS as occurring when patients show a deficit (at or below the fifth percentile for age- and education-matched controls) on at least two different measures of executive function. However, difficulties with this definition have been recognized in terms of the likely differential sensitivity of tests of executive function to impairment in ALS,[1] but have also been highlighted more recently by studies pointing to the greater than expected occurrence of language deficits that had previously not been studied in detail.[42,51] In particular, using those consensus criteria,[14] 25% of Taylor et al.'s[42] sample were cognitively impaired based on executive dysfunction. However, when the criteria were instead applied to language measures, 39% could be classified as cognitively impaired. Of note, as with tests of executive dysfunction, language measures were differentially sensitive to impairment.

It is likely that the consensus criteria[14] will need to be modified to take into account such findings concerning language impairment. Confirming the prevalence of cognitive impairment in the absence of dementia has clear resource implications and suggests that greater provision of neuropsychology services is needed for ALS patients.

What factors might be associated with cognitive dysfunction in ALS?

It is important to be aware of a number of factors that may be associated with the presence of cognitive impairment in ALS because some, but not all, are amenable to intervention.

Bulbar function

Early work[27] suggested that cognitive impairment was more prominent in patients with bulbar symptoms and advanced disease; however, as most of that sample had bulbar impairment, clarification of this issue has required further investigation. Massman et al.[28] showed that cognitive impairment was present in 48.5% of their patients with dysarthria—presumably reflecting bulbar dysfunction—although 27.4% of their non-dysarthric patients were also cognitively impaired. It has been shown[20] that patients with evidence of pseudobulbar palsy (i.e. UMN involvement in the bulbar region) were more impaired than patients without pseudobulbar palsy on several neuropsychological tests of executive function.

Inconsistent findings exist concerning the association between limb- or bulbar-onset forms of ALS and the extent of cognitive impairment detected in studies. Certainly bulbar-onset ALS is not always associated with cognitive impairment[52] just as limb-onset disease may be associated with cognitive dysfunction.[40]

Respiratory weakness and nocturnal hypoventilation

Respiratory muscle weakness, producing sleep disruption (as a result of oxygen desaturation) accompanied by daytime sleepiness, headaches, and loss of appetite may occur in as many as 44% of ALS patients with bulbar symptoms.[53] ALS patients with evidence of hypoventilation, sleep disturbance, and respiratory muscle weakness performed significantly worse on measures of memory and verbal fluency than did ALS patients without respiratory or sleep difficulties; improvements were seen on two of the memory measures in the former group following 6 weeks of nocturnal NIPPV, with a further trend towards improved verbal fluency.[54] Whilst it is unlikely that bulbar-associated respiratory weakness accounts for all the cognitive deficits reported to be associated with bulbar involvement, the role of potentially compromised respiratory muscle function should be considered when interpreting the cognitive assessment of an individual along with whether any of the cognitive impairment is potentially reversible.

Mood and medication

Many of the cognitive studies of ALS patients have excluded the possibility that cognitive deficits might be accounted for by raised depression scores. Measures of mood, however, may vary over time independently of cognitive function[55] and the severity of assessed depression may vary depending on the measure used.[56]

Most studies do not report the medical regimens of their patients. However, a range of psychotropic medications as well as other substances may affect cognitive functions[57] and in different ways. For example, benzodiazepines may affect new learning of material and tricyclic antidepressants may cause sedation and impair memory.[57] In addition, medication prescribed for ALS itself, such as riluzole, may have psychotropic effects and could potentially mask an otherwise measurable depression.[58]

Are cognitive deficits in ALS progressive?

Longitudinal studies of ALS are inevitably subject to considerable attrition due to disease factors. There is a general impression from the literature that patients

with bulbar- as opposed to limb-onset disease show greater deterioration over time.[40,59–62] However, cognitive impairment may be present relatively early in the disease and may not increase significantly on follow-up, suggesting that cognitive decline does not necessarily occur in tandem with motor deterioration.[55,62] Abrahams et al.'s[55] finding suggested that executive dysfunction (deficits in verbal fluency) may occur relatively early in the disease, with signs of language dysfunction developing as the disease progresses. However, that study did not differentiate between those with bulbar- or limb-onset disease.

Potential interventions

Very little has been written about the potential for alleviating the cognitive and behavioural changes that may occur in people with ALS, especially in cases of full-blown dementia.[18] The literature on managing cognitive and behavioural change in neurodegenerative diseases has largely focused on patients with Alzheimer's disease, and a number of techniques used in such patients have much in common with those who have suffered severe traumatic brain injuries.

Patients, especially those meeting the criteria for ALS FTD, may have limited insight into their own cognitive/behavioural changes.[63] When caregivers report changes to the clinician, it is important for them to receive basic information about the types of changes that can occur in ALS, and for a full assessment to be arranged.[64] To date relatively little easily accessible information is available to patients, carers, and professionals, although material is available from charities.[65–67]

When considering patients with FTD, but of clear relevance to ALS, Lough and Garfoot[68] recommend educating caregivers about the biological basis for behavioural changes, since caregivers may find it easier to attribute, for example, changes in interpersonal behaviour to brain pathology than to a change in their relationship. Thus it may be helpful to explain that there may be an association between frontal lobe pathology and symptoms such as aggression, impulsivity, and irritability and that there is also a biological basis for emotional lability, which predicts carer distress.[69] Lomen-Hoerth and Murphy[64] also recommend informing caregivers that some reductions in certain behaviours (e.g. affection) may not be amenable to change whereas the development of disturbing behaviour (e.g. aggression, inappropriate language) may be addressed with, they suggest, a combination of psychotropic medication, and behavioural interventions (see also Anneser et al.[70]). It has been suggested that emotional lability may be addressed using selective serotonin re-uptake inhibitors (SSRIs)[71] or dextromethorphan/quinidine.[72]

Interventions for cognitive and behavioural problems

Memory

Evans[73] has summarized a range of compensatory approaches that may be used when memory processes are failing. Since encoding difficulties may be particularly relevant in ALS-related memory weakness, approaches that enhance learning may be particularly useful. For example, encouraging and enabling the person to pay more attention to the material to be remembered, whilst also reducing possible distractions, may be helpful. Repetition of the material to be remembered, both initially and after increasing amounts of time, may help the person recall the material better.

More elaborate techniques, such as that known as PQRST, may be helpful for people with mild to moderate memory difficulties who need to recall the contents of articles/documents, since this technique increases the meaningfulness and memorability of information.[73] Thus when someone wishes to recall the content of a letter detailing important changes, for example, to financial arrangements/state benefits, they might be advised to:

- **P**review the letter (i.e. read it through quickly first of all); then set themselves some
- **Q**uestions about what is contained in the letter (e.g. what are the main changes in my benefits? when do they come into force?); they would then
- **R**ead the letter through again and would then
- **S**tate to themselves what the important information was.

Finally they would

- **T**est themselves again by asking the relevant questions, and possibly repeating this testing after increasing amounts of time.

Evans[73] suggests that for many memory-impaired people the use of mnemonics is too difficult, although many people do find 'mental retracing' (mentally retracing one's route to think about where one may have left something) may be helpful; he suggests that where the person is demonstrating a degree of impulsivity (reflective of executive dysfunction) a 'stop–think–retrace your steps' approach may be helpful. He notes, however, that the most commonly used memory aids tend to be 'external' aids, such as notebooks, diaries, wall calendars, memo boards, alarms, pill reminders/dosette boxes, as well as signposts and labels on doors of cupboards etc.[55]

Executive dysfunction

A 'stop–think' approach may be of use when impulsivity is apparent in a person's behaviour. This may be of use, for example, when considering poor compliance

with advice over swallowing food. Thus a person with bulbar dysfunction who is tending to swallow food too quickly and without having chewed it sufficiently could be encouraged, when eating, to count the number of times they have chewed their food, and then to stop before swallowing to ensure they have chewed it sufficiently, before concentrating on the advice regarding head position etc. that has been given to them by their speech and language therapist to enable them to swallow without choking. On the basis of approaches described by Lough and Garfoot,[68] flashcards reminding the person how many times to chew, or to stop chewing before attempting to swallow, might be usefully be incorporated into such a programme.

Executive dysfunction is likely to result in a person having difficulty doing several things at once (multi-tasking) or dealing with a large amount of information in one go. Reducing the 'cognitive load' placed on someone may help. Rather than asking the person complicated questions requiring the consideration of multiple sources of information, it is helpful to keep things simple and to try to get the person to restrict themselves to dealing with one problem at a time. Keeping questions phrased in simple terms may also be important should the person have difficulties with language comprehension. Spontaneous problem-solving skills may be reduced with executive dysfunction, so helping someone to talk through a problem, and providing the framework for them to consider their options, may overcome their own limitations. This may be of particular value when the person needs to consider lifestyle choices and future treatment options.

If perseverative behaviour is present, then helping the person to self-monitor how often they are doing/saying the same thing may be more effective in helping them to change their behaviour than simply telling them they are repeating themselves. Eliminating sources of distraction may also help reduce potential distractibility, and may be important during eating if swallowing behaviour is the focus of an intervention.

Behavioural change

Gregory and Lough[74] provide some helpful illustrations of how challenging behaviours occurring in FTD may be dealt with using psychological interventions, and also indicate the value of adopting interventions designed for people who have had traumatic brain injuries when working with people with FTD. In addition, Lough and Garfoot[68] have recommended a combination of approaches derived from learning theory, cognitive behavioural therapy, and neuropsychological rehabilitation. This can include environmental manipulation (for example removing items from the environment that might provoke perseverative behaviour such as questioning; removing sources of distraction), careful

structuring of the person's day to overcome apathy, and the use of self-monitoring to overcome perseverative behaviour. Gregory and Lough[74] have illustrated the potential benefits of encouraging someone to use self-control statements when trying to reduce disinhibited behaviour. The presence of executive dysfunction might render it necessary for the person to have external prompts, for example flashcards with instructions, to encourage the person not to engage, for example, in verbal aggression or abuse in response to the presence of certain individuals, since spontaneously generating the self-control statements might be more difficult for the person, particularly when in a highly aroused state.

Conclusions

The potential for cognitive involvement in ALS, ranging from mild impairment to dementia, cannot be ignored, even though not all patients will be affected. Specialist neuropsychological, and if necessary behavioural, assessments should be available for people with ALS, irrespective of whether or not they have developed a full dementia. A clinical neuropsychologist should be available to work with other members of the clinical team to ensure the best provision of care for the person with ALS as well as their carers. Since cognitive involvement may occur very early in the disease process, their involvement should be an integral part of the service offered to people with ALS and their families.

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Chapter 9

Pain, psychological distress, and other symptoms

David Oliver, Gian Domenico Borasio, and Wendy Johnston

Introduction

Clinicians caring for patients with ALS are faced with the increasing complexity of symptom management as many assumptions about ALS are now shown to be untrue and unhelpful.[1] Studies are emerging on symptom prevalence, severity, and risk factors, but with some encouraging exceptions there have been few trials of symptom management demonstrating efficacy. Understanding the prevalence of symptoms may assist in the development of appropriate screening and detection, but clinical care must still be tailored to the individual. Even if a minority suffer a given symptom, its recognition and need for management must be incorporated into ALS care.

Since the publication of the first AAN Practice Parameter,[2] more guidelines and evidence-based reviews have been published for ALS in general as well as palliative care in ALS.[3–7] Despite some advances in clinical studies demonstrating the efficacy of some therapies, these publications still rely heavily on expert opinion and clinical consensus. However, the guidelines also attempt to provide a framework for future studies, generating hypotheses even when relying on non-ALS-based studies for practice recommendations.

It may be helpful to explain to patients and their caregivers that there are some symptoms directly due to ALS and others that are provoked by or indirectly caused by ALS (see Table 9.1). Adding to the challenge of the expanding spectrum of symptoms due to or provoked by ALS is the relative lack of evidence-based practice for symptom management in general, and specifically in ALS.

The sections in this chapter are organized into symptom groups as might be elicited in a symptom review during clinical evaluation.

Table 9.1 Symptoms directly and indirectly attributable to ALS

Symptoms directly attributed to ALS	Motor	Weakness, fasciculations, cramps, spasticity (including laryngospasm, trismus, and tongue biting)
	Cognitive	Apathy, behavioural disturbances, cognitive impairment, dementia, impaired decision-making
	Pseudobulbar affect	Pathological laughing and crying
Symptoms indirectly caused by ALS	Pain	Immobility, injury, weak unsupported joints, skin pressure and breakdown, headache, limb pain including dependent oedema
	Secretions	Sialorrhoea (drooling), thick tenacious secretion, nasal congestion
	Urinary	Urinary frequency, incontinence, retention
	Gastrointestinal	GERD, bowel management (including constipation)
	Psychological	Fatigue, insomnia, depression, and anxiety

ALS, amyotrophic lateral sclerosis; GERD, gastro-oesophageal reflux disease.

Case history

A 58-year-old man was diagnosed with ALS based on progressive leg weakness and spasticity. Mechanical back pain, which had previously responded to physical therapy and non-steroidal anti-inflammatory drugs (NSAIDs), now returned and persisted despite implementation of the same measures. Initially he declined the use of a cane, but following a fall in which he broke his right wrist he agreed to an ambulation aid. The back pain persisted despite muscle relaxants, a change in NSAID, and ongoing physiotherapy.

At the first follow-up visit in the ALS clinic after the wrist fracture he reported pain in his wrist and back as well as painful cramps in his legs, with frequent nocturnal awakenings due to pain and cramps. Reluctant to use stronger pain medications, he accepted baclofen at bedtime, increasing to 10 mg twice a day, then 20 mg. The cramps and nocturnal pain improved, and he managed his back pain by resting more frequently. The wrist fracture healed well, and he reported his pain as predictable but manageable. He relied on a light-weight portable self-propelled wheelchair for distances and walked in the house with a cane.

One year after diagnosis he was increasingly wheelchair dependent and a power chair was recommended, anticipating future arm weakness. Arm, bulbar, and pulmonary function remained normal. He continued to use baclofen at 10 mg three times a day as well as riluzole and NSAIDs as needed. He had more problems with constipation and he was provided with dietary advice regarding fibre and hydration, and a laxative as needed. While final adjustments to his power chair were under way he once again reported back and now hip pain. He declined narcotic analgesics because of the risk of worsening constipation; increased physical therapy was helpful. Symptoms remained confined to his legs and back, with back pain and leg cramps a recurring problem. Clinical examination also revealed mild weakness and

atrophy of his hand muscles and severe spastic weakness of his legs. Tramadol was prescribed, and further counselling regarding bowel management provided. An intrathecal pump for administration of baclofen was discussed but declined due to the risk that the intervention would lead to decreased tone and increased leg weakness.

Following a transatlantic flight, he was hospitalized for a deep venous thrombosis (DVT), and discharged on oral anticoagulants. He enjoyed 6 months of stability, but following a prostate biopsy he developed respiratory failure, and was found to have a pulmonary embolus (PE), his anticoagulants having been stopped for the biopsy. Following his recovery he continued on anticoagulation. At the last clinic visit he was using a wheelchair for all ambulation, retained good bulbar and arm function, and felt his symptoms were controlled.

Pain

The characterization of ALS as a motor neuron disease with relative sparing of sensory pathways led to the erroneous assertion that pain was not a feature of ALS. However, sensory disturbance in patients in ALS was not unknown; as early as 1953, 11% of ALS patients were reported to have sensory symptoms.[8] In patients who otherwise met the criteria for ALS, almost 25% had abnormal sensory nerve abnormalities on electrophysiological testing, with 12.5% meeting the criteria for polyneuropathy.[9]

The prevalence of severe pain in ALS patients on admission to a hospice ranged from 50 to 57%;[10] 64% of patients seen in a neurological service were found to have pain;[11] a survey of dying patients found a pain prevalence of 70% for patients dying at home and 70% for patients dying in the hospice;[12] 48% of patients had pain and 72% complained of 'discomfort other than pain' in the last month of life.[13] A study of ALS patients attending a neurological clinic found that for men pain had been present for an average of 18 months,[11] and at the hospice although 57% of the patients complained of pain only 12% had received a strong analgesic.[10] In the ALS Care Database, of 1000 patients nearly 24% reported pain and 75% had received medication to control pain.[14]

In a population-based study, pain was found to be more frequent and severe in patients with ALS (57%) compared with age- and sex-matched controls (39%).[15] Reports of pain were correlated with longer disease duration and lower score on the ALS Functional Rating Scale (ALSFRS). Patients were more likely to be treated for pain (69% compared with 39%), with NSAIDs most frequently used in both groups and opioids only in the ALS group. Pain more often interfered with more activities in ALS patients than in controls. The pain was more often located in the extremities, notably the shoulders and hips.[15] The localization of pain to the extremities was confirmed in a clinic-based study, which demonstrated that about half of the patients surveyed reported pain.[16] Using

FVC respiratory testing to stage ALS, the prevalence of pain was equal at all stages, but in patients with pain there was a correlation with functional status—those with poorer status had higher pain scores.

There are no randomized or quasi-randomized treatment trials of pain in ALS.[17] The treatment of pain will depend on the cause.

Cramps and fasciculations

Fasciculations (painless twitches of small numbers of muscle fibres, visible to the eye but with no movement effect on the joint), thought to be due to the degeneration of the intramuscular motor axons, are an early symptom, often seen before weakness develops. They can lead to painful muscle cramps. Cramps occur more commonly in the earlier stages of ALS, may appear in unusual locations, such as the abdominal and paraspinal muscles, and may be the cause of significant discomfort.

Fasciculations rarely require treatment, but anecdotally both baclofen and gabapentin have been helpful. Levitiracetam 1500 mg twice a day decreased the severity and frequency of cramps in an open-label trial.[18] Quinine sulphate was used by 58% of European centres,[6] but is specifically proscribed by the United States Food and Drug Administration. To date studies have been negative, but underpowered.[19] Low-dose nabilone, a synthetic cannabinoid, reduced spasticity-related pain in a double-blind placebo-controlled crossover study.[20] Tetrohydrocannabinol is reported in one double-blind placebo-controlled study to have no benefit; however, the study may have been underpowered.[21]

Musculoskeletal and joint pain

Joint pain develops because progressive wasting and weakness offer little support. Shoulder pain has been described as common, affecting 28% of ALS patients in a retrospective chart review.[22] Although 25% of those affected had prior shoulder pain, the mean time to occurrence was 31 months; the only clinical correlate was proximal arm weakness, not site of ALS onset or predominantly UMN versus LMN involvement. Patients with shoulder pain were more likely to report pain elsewhere, however. Aetiology was not reported in 67%, and only 18% were diagnosed with adhesive capsulitis, or frozen shoulder. Just over half received therapy (physical therapy or pain medication). Intra-articular injection of pain medication or steroids was not described. Careful positioning and regular gentle movement may be helpful, and support of flaccid arms, particularly in a car or wheelchair, should be considered.[23] Intra-articular injections of local anaesthetic and/or steroids can be helpful if there is particular inflammatory pain in a single joint.[24] For musculoskeletal pain, NSAIDs such as ibuprofen or naproxen can be useful.

Generalized and regional pain, particularly back and hip pain, may occur as a patient becomes less mobile.[25] Regular range-of-movement exercises supervised by physical therapists are indicated. Although caregivers may be taught to assist, a regular review of technique by the healthcare team ensures that good practices are maintained.

Skin pressure, due to immobility and the preservation of the sensory nerves may be perceived as pain, and painful pressure ulcers may occur.[26]

Patients with ALS will experience pain related to pre-ALS and concomitant conditions. In fact the immobility, wasting, and weakness may predispose patients to the recurrence of prior conditions, for example cervical or lumbar radicular pain. Risk factors for recurrence are unknown, as is whether preventative measures may reduce the risk. Although Rivera et al.[16] used the neuropathy pain scale in their survey of pain in ALS patients attending an ALS multidisciplinary clinic, whether ALS patients are more likely to experience neuropathic pain is unknown.

The treatment of pain not controlled by repositioning, splints, or braces, or by physical therapy, will need regular administration of analgesics, using the World Health Organization three-step analgesic ladder developed for the relief of cancer-related pain.[27] The aim of the 'ladder' is to allow progression from step to step until pain control is achieved.

Initially patients may benefit from simple analgesics or weaker opioids, given on a regular basis and titrated to the patient's response. However, many patients may require opioid analgesics to control the pain and discomfort. Many patients and their families fear the use of opioids, as do some medical and nursing practitioners, but the response to opioids can be dramatic and the quality of life can be greatly improved by their use. Experience in the care of cancer patients has shown that if analgesics are used correctly and carefully, titrated to the individual patient's pain, and given orally and regularly, pain can be controlled without appreciable side-effects, with patients remaining lucid and alert.[28] The fear of shortening a patient's life through opioid medication is unfounded.[29] In a pilot study of six ALS patients taking opioid medication to treat dyspnoea, significant improvement in objective and subjective symptoms occurred without evidence of hypercarbia or respiratory depression.[30]

Morphine is the most widely used opioid—as morphine elixir or tablets every 4 h, modified-release tablets, or capsules every 12 h. Morphine can be used effectively and safely. A hospice study showed in the last 72 h of life the mean dose of morphine was 80 mg/24 h, and did not appreciably increase in that period.[31] Other strong analgesics may be considered—such as oxycodone elixir, capsules, or tablets, or a fentanyl transdermal patch, which can be particularly useful if swallowing is difficult and the pain syndrome is stable.

If swallowing becomes difficult a liquid formulation may be necessary so that the medication can be given through a PEG. In the later stages of the disease subcutaneous infusions using a syringe driver may be helpful, as this technique allows the administration of other medication, for example midazolam for stiffness or agitation and an anticholinergic such as hyoscine for secretions (see also Chapter 7).

The AAN Practice Parameters and EFNS Guidelines recommend that opioids be used and the WHO pain management recommendations should be the standard for the administration of analgesics.[2–7] However, there has been little progress in the development of ALS-specific protocols or further research into ALS-related pain.[5,17] These are areas for further development.

Pain can come to dominate a patient's life, and that of the family and carers. The assessment and treatment of pain can have an enormous impact on the patient's quality of life and allow them to enjoy the company of their family and friends.

Weakness, mobility, and spasticity

Weakness is the cardinal symptom of ALS. The *timely* discussion and provision of adequate aids (from a cane, to an ankle–foot orthosis, to a wheelchair, etc.), is the best intervention. Physiotherapy is very important to prevent contractures and maintain mobility, and is discussed in detail in Chapter 12. The initial discussion may not be welcome, or heeded by the patient and family, but most families in retrospect feel that the process for obtaining durable equipment should have been started sooner. Patients and families tend to take their cues from medical professionals regarding bad news, whether it is the need for a wheelchair, for a feeding tube or respiratory support, or for advance care planning in general (for example Chapters 5–7).

Pharmacologically, there is little to offer. Nascent neuromuscular junctions, as seen in early re-innervation of denervated muscle, may have a high incidence of transmission failure. Therefore, anti-acetylcholinesterase medications such as pyridostigmine may lead to transient improvement in muscle strength, especially early during the disease. This seems more pronounced in patients with bulbar disease. Some patients have reported a period of benefit when it is taken prior to meals. However, improvement is not seen in all, and it usually only lasts for days to a few weeks. Acetylcholinesterase inhibitors do not alter the course of ALS. The short-term use of pyridostigmine (up to 40 mg three times a day) can only be recommended for special situations, like a plane trip or a holiday. There is no rationale for long-term therapy with pyridostigmine in ALS, and more often than not patients will have to be taken off this medication to avoid

unnecessary side-effects. At best it is a temporizing manoeuvre that can only be offered with appropriate caveats and only for short-term, targeted goals. A clinical trial would be ideal.

Creatine monohydrate was evaluated in three clinical trials but has not influenced survival or change in the ALSFRS.[32]

Spasticity of the extremities, which is due to degeneration of the UMNs, can be clinically severe. It may be effectively relieved by appropriate medication. Undesirable side-effects of the commonly used drugs include weakness and fatigue. For these reasons, and since a moderate degree of spasticity may assist mobility, careful titration of dose is required. Baclofen is the most widely used drug, although no specific study for use in ALS is available, as shown by a recent Cochrane Review.[33] Tizanadine, gabapentin, and benzodiazepines have also been used. The only quasi-randomized trial reported for ALS spasticity showed that moderate exercise of the affected limb improved outcome at 3 months.[34] Clinical efficacy in the reduction of multiple sclerosis (MS)-related spasticity was demonstrated in placebo-controlled trials of a cannabinoid, nabiximols (Sativex®).[35]

Physiotherapy with passive movements, with the patient positioned carefully and comfortably, continued very regularly several times a day by family or other careers, may be very helpful. The physiotherapist may be able to assess the role of spasticity in the causation of pain, and assess the response to treatment.[22]

For severe spasticity, baclofen intrathecally via an implanted pump is reported to benefit selected patients.[36,37] The most recent study selected only ALS patients with severe lower extremity spasticity and having UMN-predominant disease. A good outcome was reported, with reduction in pain and use of medication for those who chose to proceed to pump implantation compared with a comparable and eligible group who declined the intervention. At follow-up there was similar loss of ambulation in both groups.

An alternative for such refractory cases, uncommon in ALS, may be intramuscular injections of botulinum toxin, which have been shown to be effective in MS.[37–40] Dantrolene, which acts directly on the muscle, should not generally be used as first-line medication because it enhances weakness. However, extreme spasticity in the terminal phase which could only be relieved by high doses of intravenous dantrolene has been reported.[41]

Secretions, breathing, and airway concerns

The impact of secretions on overall quality of life, on the success of NIV, and on the development of life-threatening complications is greater in ALS than in any other neuromuscular condition, and is closer to the experience of patients with

other neurodegenerative disorders such as Parkinson's disease. The combination of progressive restrictive respiratory failure with secretion and airway difficulties is uniquely challenging, particularly in rapidly progressive disease with bulbar onset.

Sialorrhoea

Excessive saliva (sialorrhoea), also called drooling, is a frequent complaint in ALS. It is due to a combination of weakness of the facial muscles and reduced swallowing ability. It is important to recognize and discuss the social impact of drooling, which can cause social isolation and significant discomfort.[13]

1 The most widely used drugs are glycopyrrolate, with class I evidence in Parkinson's disease (but not ALS), and amitriptyline, although no controlled trials are available for the latter. Sublingual atropine drops provide short-acting relief. Transdermal hyoscine patches applied every 3 days offer prolonged relief, and have the advantage of bypassing oral delivery, which may be very helpful in bulbar cases.
2 Botulinum toxin, injected into the salivary glands, is one the few evidence-based treatments that can be recommended, but it requires a skilled practitioner as it is associated with the risk of worsening swallowing.[3,6] Botulinum toxin injected into the upper oesophageal sphincter showed benefit in ALS patients with UMN-type swallowing disorders.[42]
3 Radiation therapy is another option for treating sialorrhoea. It has been shown to be effective for patients with different neurological diseases, including ALS, when directed at the parotid glands or the submandibular and sublingual glands.[3,6] Side-effects include pain in the parotid area, dryness of the mouth, burning of the skin, sore throat, and nausea; these are usually transient, but on occasions they are more severe.
4 Surgery to sever the nerves to the parotid glands has been reported, but the risks generally outweigh the benefits in typical ALS.

Thick secretions

1 Mucolytics: a mucolytic including *N*-acetylcysteine, 200–400 mg three times daily, may be beneficial.
2 Beta-receptor antagonists and a nebulizer with saline and/or an anticholinergic bronchodilator and/or a mucolytic and/or furosemide may be used in combination.
3 Suction used in combination with the above, in presence of a sufficiently forceful cough, or assisted cough.

4 Patients and caregivers need to be taught to perform assisted cough, with lung volume recruitment or mechanical cough assist.
5 Other treatments which have shown benefit in an uncontrolled survey include red grape juice, papaya enzymes, sugar-free citrus lozenges, grape seed oil, and betablockers (see Chapter 14). Reduction of alcohol and caffeine, substitution of dairy products, increased fluid intake, moistening of the air, and steam inhalation may provide some benefit. A satisfactory therapy for thick mucous secretions in ALS is still not available and more research is needed in this area.

Nasal congestion

Chronic or seasonal rhinitis may become more symptomatic, especially with bulbar involvement with a weakening of the nasopharyngeal muscles. Attention to precipitants and the use of topical decongestants (nasal sprays) may alleviate congestion and post-nasal drip. Anecdotally, symptoms may be helped, especially at night, by elevating the nasal bridge with nasal tape.

Laryngospasm

This sudden reflexive closure of the vocal chords can cause panic due to a sensation of choking. Several types of stimuli (e.g. emotions, strong flavours or smells, cold air, fluid aspiration, gastro-oesophageal reflux) may provoke this symptom, which usually resolves spontaneously within a few seconds. Repeated swallowing while breathing through the nose can accelerate resolution; sublingual lorazepam has been anecdotally reported to assist in management.

Trismus, jaw spasms, and clenching may develop in patients with pseudobulbar involvement in response to noxious stimuli such as cold, anxiety, or pain, and may be relieved by benzodiazepines (e.g. sublingual lorazepam or clonazepam).[43] There is a case report of recurrent trismus and stridor being successfully treated with botulinum toxin injection into the bilateral masseter and pterygoid muscles.[44]

Bowel, bladder, and sexual function

Although sexual function is not affected by the disease process, sexuality is affected by changing body image, fatigue, and changing family dynamics, with a significant loss of interest and moderate loss of sexual activity compared with the situation before the diagnosis of ALS.[45] Problems with sexuality went from 20% pre-diagnosis to 65% and 75% for patients and partners, respectively, post-diagnosis.

Patients with ALS do not typically suffer from direct involvement of the autonomic nervous system or smooth muscle, but weakness of striated muscle can affect both the lower oesophageal sphincter (from weakness of the diaphragm), leading to gastro-oesophageal reflux disease (GERD), and the perineal muscles, affecting socially appropriate bladder control.

The treatment of GERD includes appropriate positioning during and after meals, avoidance of meals for 2–3 h before lying down, and medication. Proton pump inhibitors can be given empirically if typical symptoms of heartburn and regurgitation (which could manifest as recurrent cough) are present.[45]

Constipation is a significant issue in the general population, with the incidence increasing with age, nursing home placement, and in many neurological illnesses. Immobility, dietary change, and decreased fluid intake can contribute to a potentially major problem in advanced disease, further compounded by medications used for symptom management such as opioids for pain and dyspnoea and anticholinergic drugs for control of secretions.

Patients with ALS should be screened for pre-existing constipation and provided with dietary advice, including adequate fibre and hydration. Stool softeners are of questionable benefit. Stimulant laxatives such as senna and bisacodyl are useful for intermittent or short-term constipation, whereas chronic constipation is best treated with osmotic laxatives such as lactulose or polyethylene glycol, the latter being judged superior in clinical trials because it does not affect electrolytes. Medication-related constipation can be difficult to manage, particularly that associated with opiates. A bowel regimen should be instituted when opiates are initiated. Opiate rotation, particularly to transdermal fentanyl (associated with lower constipation risk), may relieve symptoms. Specific treatment with opiate antagonist agents such as naloxone, that can cause severe withdrawal symptoms, methylnatrexone, which has poor penetration to the central nervous system (CNS) and thus a low risk of withdrawal symptoms, or lubiprostone, which has not been studied in palliative care, would be best undertaken by those experienced in their use. Enemas are also used. Obstipation should be suspected if abdominal pain emerges and appropriate investigations initiated.

In the absence of studies specifically for ALS, patients are best managed with palliative and chronic care protocols.

Cognitive and behavioural changes

The incidence of cognitive and behavioural changes associated with ALS is now recognized to be significant (see Chapter 8). Overlap with FTD, including the behavioural variant (bvFTD), as well as a spectrum of dysexecutive disorders

have been described. The impact can be greater on the family and other caregivers than on the patient. Patients and families need to be informed of the risk of these changes as much as they need to be informed and prepared for the potential for respiratory and bulbar changes.[3,6] In an online survey, the majority of patients (62%) and carers (71%) indicated a desire to be informed that cognitive change or dementia might occur.[48] There are implications for legal and advance care planning (see Chapters 4, 5, and 18).

Management of behavioural changes is based on recommendations for other frontal lobe syndromes, as there are no ALS-specific studies. As described in Chapter 8, there are sound strategies for behavioural management. Little evidence-based research for pharmacological management is available, even for FTD patients.

Pharmacological treatment with acetylcholinesterase inhibitors, *N*-methyl-D-aspartate (NMDA) antagonists, antidepressants, and antipsychotic medications have been reported in the management of FTD and bvFTD. The best evidence, with only one trial so far, supports the use of trazodone at up to 300 mg per day to improve behavioural symptoms in FTD. Other antidepressants are well tolerated and may improve behaviour, but the evidence is Class IV. Paroxetine, galantamine, and memantine have had clinical trials showing no benefit. Antipsychotic medications, which cause increased risk of death in the elderly, have no convincing evidence of efficacy in this population at this time.[49] Decisions for medication-based management in ALS-FTD and the other cognitive–behavioural variants in ALS will need to be made on a case-by-case basis, as needed for behaviour change, but no medication is indicated at this time for improving cognitive outcome.

Mood, sleep, and affect

Depression is prevalent in ALS, although it is most prominent in early stages pre- and post-diagnosis and at end of life. ALS patients are at higher risk of suicide than the general population, particularly in the first year after diagnosis.[50] The prevalence of depression is debated, and relates in part to screening methods and instruments used.[51] Depression has a high impact on quality of life, and on the quality of life of the caregivers. In turn, stress and depression in caregivers adversely affect ALS patients (see Pagnini[52] for a detailed review).

Anxiety, not necessarily associated with depression, is also more common in ALS patients than in other terminally ill people. The anxiety, often related to fear of the future, of future suffering, specifically about symptoms of choking or respiratory failure, is predictive of desire to hasten death (see Chapter 4).

Hopelessness distinct from depression is highly associated with interest in hastening death, as is loss of meaning in life. These symptoms may reflect spiritual concerns, as discussed in Chapter 11.

Using a more general measure of distress, the Brief Symptom Inventory, ALS patients were found to be generally more distressed than the general population and showed distress equivalent to two-thirds of that measured for a cohort of distressed psychiatric outpatients.[53]

The considerable amount of research undertaken to quantify and categorize psychological distress has not given rise to a consensus on the best screening methods, or treatment trials specific to ALS. Distressed patients and their loved ones should be offered diagnostic interviews to assess the contribution of psychosocial and spiritual factors as well as physical symptoms.[54] Appropriate choice of antidepressants, with follow-up to assess efficacy, should be coupled with psychological counselling and family support.

Pseudobulbar affect

The development of uncontrollable bouts of laughter and/or tearfulness (the latter occurring more often) is referred to as 'pseudobulbar affect'. The pathophysiology is poorly understood. It is not specific for ALS, but also occurs in other diseases of the CNS such as MS.

Pathological laughing/crying occurs in up to 50% of ALS patients in the course of their disease. It is an abnormal display of affect due to dysregulation of the motor components of emotional experience and may be related to frontal cognitive dysfunction, but is not correlated with cognitive impairment. The symptom is socially disturbing.

Patients should be asked about the symptom and counselled that it is a reflex related to ALS, not a sign of underlying psychiatric disease. The most widely used drug is amitriptyline,[55] but SSRIs are also used, with Class IV evidence. The combination of 30 mg dextromethorphan hydrobromide and 30 mg quinidine sulphate twice daily in the palliation of pathological laughing/crying has been demonstrated in two randomized, double-blind placebo-controlled studies and is approved in the United States[6] and Europe.

Fatigue

Patients with ALS report fatigue in excess of normal controls, and fatigue does not correlate with weakness alone, suggesting a central and peripheral component. Depression, SDB, and respiratory insufficiency are potentially treatable causes of or contributors to fatigue.[56] In ALS patients in whom these factors have been ruled out or appropriately treated, modafinil may improve

subjective fatigue and excessive daytime somnolence. An open label study and a small, randomized, placebo-controlled trial showed that modafinil 300 mg per day significantly reduced fatigue during the 4-week double-blind phase and the 8-week open label follow-up period, with no serious adverse effects.[57]

Insomnia

Sleep disturbance is common in ALS, particularly in advanced stages.[13] Prior to treating with hypnotic medication, other causes need to be rigorously explored. The most common are:

- respiratory insufficiency with oxygen desaturation and dyspnoea
- psychological disturbances, anxiety, depression, nightmares
- inability to change position during sleep due to weakness
- fasciculations and muscle cramps
- dysphagia with aspiration of saliva
- anxiety and/or depression.

Once other causes have been identified and treated, or treatment offered, attention to sleep and comfort at night should be ongoing. Sleep hygiene and a gentle hypnotic may be indicated as first-line treatment, using medication such as zopiclone, amitriptyline, or mirtazapine. Adequate sleep is required for the patient and family, and the quality of life for all will suffer if sleep disturbance is not minimized. Many state the desire to 'die in my sleep'. To achieve this goal, they have to sleep.

Other symptoms and complications

Skin disorders and pressure ulcers are said to occur with less frequency in ALS than in other neurological diseases such as MS, but this assertion has been disputed. In fact, pressure ulcers may occur at any stage of ALS due to patient-related factors.[26] Family and professional caregivers should monitor the skin integrity of ALS patients with the same vigilance as in any other disabling neurological condition.

Dependent oedema of the hands and feet occurs in weak limbs because of reduced muscle pump activity, and may progress to involve much of the affected extremity. Limb elevation, physiotherapy, and compression hose are helpful. Diuretics may also be helpful, but urinary frequency, dehydration, and renal insufficiency may limit use. If pain develops or swelling persists despite prolonged elevation, a DVT should be ruled out.

ALS patients are at higher risk of DVT (2.7–3.0%), than the general population (0.1%) or hospitalized patients (1.3%).[58,59] According to the reporting on adverse events from a trials database comprising 501 patients, risk factors include lower FVC, ALSFRS, and leg strength at the time of enrolment. In the same population PE occurred in eight patients, three of whom did not have a DVT documented. Based on personal experience, once prophylaxis for DVT is initiated it should be continued for life, as withdrawal appears to be associated with a high risk of recurrence with life-threatening PE. The latter recommendation would be best subject to an objective study; however, until data are available ALS patients should be considered at high risk of recurrence due to immutable risk factors.

Conclusion

In addition to the symptom burden directly and indirectly due to the disease, ALS can exacerbate pre-existing symptoms. Strategies for managing previous and emerging symptoms must be tailored to the individual patient, and symptoms unique to ALS must be recognized, explained, and strategically managed. Knowledge of the incidence, risk factors, and implications of these symptoms is improving, and treatment trials have established efficacy of some interventions. More research into ALS-specific care is clearly needed.

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Chapter 10

Psychosocial care

Sue Smith and Maria Wasner

Summary

Psychosocial care is the responsibility of everyone working with those affected by ALS. It includes the giving of bad news, respecting and developing coping strategies, acknowledging fears, and maintaining hope. The assessment and continuing review of psychosocial concerns must recognize the particular issues faced by carers and children living with ALS. Good psychosocial care also recognizes the challenges for the professionals involved. ALS has an impact on the patient's role and relationships with family and friends. For all concerned the experience is not just physical; the 'total pain' concept[1] which encompasses physical, emotional, social, and spiritual elements, acknowledges the link between symptoms and psychological distress. A patient's experience of illness is a direct product of the interaction between all aspects of life.[2] Competent psychosocial care can significantly improve the quality of life of the patient and the family caregivers.[3]

Case history

Paul and his family were keen sailors, and when he first attended the clinic he was sailing his yacht single-handed and running his business from his home. Six months later, on attendance, he explained that he still goes sailing as often as possible but is now helped onto the boat and sits and watches whilst others are doing the physical tasks, and he is struggling to use his computer in order to continue to work.

Introduction

Psychosocial care is a core component of good palliative care, and underpins the practice of all professionals and volunteers. At its heart is the principle of working with the patient, family, and friends as the unit of care:

> Psychosocial care is concerned with the psychological and emotional well-being of the patient and their family/carers, including issues of self-esteem, insight into and adaptation to the illness and its consequences, communication, social functioning and relationships.[4]

Working with the emotional, social, and spiritual elements of an individual or family should not be apportioned to one professional.[5] A patient will choose who they talk to about these issues—so for example it may be with a speech and language therapist that they voice concerns about choking to death. A specialism of psychosocial care does exist and is most commonly provided by social workers, counsellors, and psychologists. Each brings their own expertise and way of working, such as counselling (individuals, couples), family therapy, group work, direct interventions with children, bereavement care, and knowledge of the support and benefit systems. Such professionals are a fundamental element of any palliative care team: where a team does not exist, access to a specialist in psychosocial care is important. Evidence supports the benefit of psychosocial interventions in cancer.[5–7] Whilst a diagnosis of ALS does not necessarily lead to depression, for those who do experience this Rabkin et al.[8] have shown that these patients are more interested in life ending earlier, and whilst this does not manifest itself as increased interest in assisted suicide, depression predicts shorter survival time in ALS.[9] Responding to the psychosocial needs of patients, their families, and carers influences their quality of life and may affect the quantity of life.

The practice of psychosocial care is founded on a secure base of knowledge, skills, and values. These help in the initial assessment of the patient and family, which takes into account their experiences and capacity for change. Oliviere et al.[10] list the skills essential for carers, health professionals, and volunteers. Workers should respect the individuality of the patient and those connected with them: this is recognized by placing them at the centre of the care, offering realistic choices, and being non-judgemental. Sheldon[11] discusses four key concepts in psychosocial palliative care: attachment, loss, meaning, and equity. An understanding of these is essential to the practice of any palliative care professional: they are inextricably linked to much of the discussion in this chapter. Some of the issues facing people affected by ALS will also be encountered in cancer or a chronic illness such as Parkinson's disease, and distinguishing areas of emphasis specific to psychosocial care is important. Naturally, the extent to which individuals and their families experience these issues will vary. Their inclusion here is intended to raise the awareness of professionals and volunteers and consequently improve the quality of psychosocial care.

Living with ALS

Breaking bad news

The difficulties in giving the diagnosis of ALS or other bad news should not be minimized. Doctors may feel helpless when the role of healer is missing and

feelings of sadness can be evoked in both the recipient and the giver of the news. Dealing with other reactions, such as anger and despair, also causes concern. Faced with this there may be a tendency to give false reassurance, likened to a 'conspiracy of speech',[13] or use closed instead of open questions (see Chapter 3).

Assessing and responding to behaviour and feelings keeps the situation patient-focused and reinforces the importance of emotional care at an early stage. It gives a clear message that having and talking about feelings is normal and that all staff share the responsibility for this. This in turn has implications for professional training in communication skills.

Assessment

One of the challenges of psychosocial care is to recognize the interplay of the experience for the patient and those around them whilst also seeing each individual in the situation as just that—an individual. Each person brings their own past, values, choices, and feelings. No assumptions should be made about how each person will perceive or be affected by the illness based on visual clues, such as skin colour, age, or gender. Comparisons should not be made between patients or families in apparently similar circumstances. As Earll et al.[18] conclude: 'it is important that professionals do not assume either that they can anticipate the patient's view of their condition, or that the medical perspective is the one adopted by patients'. The lack of clarity in attempts to determine a psychological profile of the ALS patient supports the individualized approach which is sensitive to the culture of the patient, family, and community.[19–23] Consideration of the patient's competency is also required during assessment, given the prevalence of cognitive change in ALS,[24] and this may affect the speed with which information is given.

During assessment the practitioner creates a partnership with the patient and other individuals involved. Assessment can touch on sensitive issues, which raise ethical concerns requiring careful moral scrutiny.[25] This demands that professionals explain the reasons for their inquiries, establish consent, and check on it at intervals as the assessment progresses. Saying: 'If there is something you would rather not discuss, please let me know', is a clear way of 'handing back as much control as possible to the person whose body is out of control';[11] this is equally applicable to others being assessed. Establishing a partnership is also helped by discussing confidentiality and with whom information can be shared. This is crucial when working as part of a team.

Assessments vary in their formality. Whatever format is used, an initial assessment, which may not be completed in one visit, will identify those individuals and families at particular risk who will benefit from more support. Such an assessment will need to explore:

- the understanding of the individual patient
- the effect of the illness on family roles and relationships
- personal histories of family members
- family life-cycle issues, e.g. births, children leaving home, marriages, retirement
- previous crises and losses, how they were handled, and additional concurrent crises
- other vulnerable individuals in the family, for example someone with learning difficulties
- the family's physical and social resources.

Numerous studies,[26–29] stress the importance of not accepting patients or family members as proxies for one another's opinions and experiences. Agreeing which problems can be changed and what the plan is to enable change keeps control firmly with the patient. Any plan must be based on realistic resources to support it and needs to be re-evaluated as the condition and circumstances alter. For example, Albert and colleagues[30] note the capacity of patients to express preferences for life-extending technologies, but caution that these may change over time and that education is required throughout the illness.

For some people with ALS who lose the ability to communicate verbally, alternative ways of gaining their thoughts and feelings must be considered. First, the patient and family should work with a system or aid with which they feel familiar and comfortable (e.g. a Lightwriter or iPad). Discussing whether tiredness may affect the length of visits determines how focused assessments need to be and whether future visits should be planned. In these circumstances, a predominance of closed questions which draw on an awareness of the patient's situation can be helpful: 'Other people I have met with ALS have worried about becoming more dependent upon their family. Is this something you have thought about?'. Whilst this type of question appears leading, the physical needs and limitations of the patient must be considered. If the assessor shows sensitivity to this issue, the patient is more likely to feel like a partner in the process. If the patient wants to give a fuller answer, they must be allowed the time they need, rather than the professional anticipating what the patient is trying to communicate after a few words on the communication aid.

Assessment is not just a means to an end. In work where time and contacts may be limited, the assessment is an intervention in its own right: 'building up a therapeutic relationship with the ill person and family, engaging them in a helping process, allowing them to feel safe and comfortable, communicating interest and respect, carving out time to allow their anxieties to surface'.[9]

Hope

Case history

Jim and his wife Carol were devastated by his diagnosis and felt that their pleasant life in early retirement was over. They had enjoyed cruising holidays, and the social worker suggested that they plan another cruise as soon as they could. When seen in clinic 4 months later Jim told the social worker that she was 'bankrupting' him because Carol had followed the advice and had three further cruises planned over the next 8 months. Jim and Carol were much more relaxed, both with each other and with the team.

Attempts have been made to define hope: Buckley and Herth[31] summarize it as 'an inner power or strength that can enrich lives and enable individuals to look beyond their pain, suffering and turmoil'. What is the relevance of such a concept to someone given a diagnosis of ALS with a limited life expectancy? The presence and experience of hope will depend in part upon the way in which information about the diagnosis, prognosis, and treatment is given. If it is understood that the future will be dramatically different from that envisaged before the diagnosis, then the way hope is viewed will also be affected. For many with ALS the shock of the diagnosis is compounded by the relentless progression of the condition, with one loss following another: this can easily leave them feeling helpless and hopeless. Maintaining hope even under these conditions is evident, but it may be different from, for example, the case of someone undergoing chemotherapy. Hope is often attached to treatment, whether curative or palliative. Even now, with treatment regimes for MND in their infancy, individuals will readily agree to new medication or take part in trials, some in the hope of a better future for themselves but others selflessly to promote a better future for those diagnosed later.

Kim[32] suggests that 'an individual's level of hope determines whether human beings live or die'. Identifying what hope means to someone, what helps maintain their hope, what causes them to lose hope, and how it changes during an illness is important in psychosocial care. Palliative care professionals and volunteers can play their part alongside the patient's family and friends in fostering hope through categories identified by Buckley and Herth.[31] Maintaining relationships with children, considering spiritual issues (see Chapter 11), and supporting independence for as long as possible are some areas to focus on, even as death approaches. Working on a life review with the patient[33] not only gives the opportunity to recall the highlights of their life but may also be done with fun and humour; the benefit of which others have commented on.[34] Others may describe their experience in terms of suffering: there are parallels between this multidimensional concept[35] and the loss of hope for people with ALS.[36] Attempts to understand how terminally ill people suffer presents challenges and

opportunities. Whilst suffering is more complex than purely physical pain, good symptom management is essential in combating suffering.[36,37] It is equally clear that the wider understanding of and engagement with suffering lies beyond medicalization.[38] This is essential if the extent and language of suffering and hope are to be fully comprehended. Only then will professionals develop the competencies to work with them effectively.[39,40]

Loss

Psychosocial interventions may assist the person with ALS to deal with the many losses they encounter. Such losses occur at different levels of visibility and different stages of the disease. At diagnosis, the patient and family may grieve for the future they had planned and expected. It will be difficult to grieve for subsequent losses, as they overlap. We often have the human capacity to adapt to loss and cope with the concomitant stress. This usual adaptive model, however, does not allow for a spiralling scenario in which a period of adjustment and equilibrium is interrupted by the next loss. ALS seems set apart from other chronic illnesses in this respect.[41] The re-formation of self for a burns survivor[42] following enormous losses is possible during rehabilitation. Those with ALS, who experience irreversible and unremitting physical losses, including breathing difficulties, will not necessarily view this as affecting their quality of life,[43,44] though there is an apparent association with depression.[45] The meaning of an apparent loss may have other underlying and devaluing implications; losing a job can affect status, role in the family, and self-esteem. The full implications will probably emerge only later, by which time the patient may have had to face loss of mobility or communication. If willing, they may find it helpful to talk about the numerous losses with an outsider, given permission to mourn the changes and express powerful emotions.

Control and choice

Issues of loss and control are linked. Feelings of being out of control may predate the diagnosis: testing, misdiagnosis, and increasing problems may produce a lack of faith in medical professionals. Hearing about the history of the illness at the initial assessment acknowledges that the way a patient feels today may stem from an unresolved or unspoken past event. Unobtrusive questioning and precise summarizing along with genuine interest in the person's feelings about the events demonstrate tangible concern. Returning a sense of control to the patient and family is a hallmark of good psychosocial care. Though ALS patients are no more or less likely to feel an internal sense of control,[20] those who do believe in chance or the power of others over their health are more likely to feel hopeless.[46] The extent to which they feel in control of their life will vary from

person to person and probably from day to day. ALS does not deprive patients of their ability to convert their early treatment preferences into real choices.[30] Similarly, professional involvement need not leave the patient, family, and carers feeling disempowered if it includes those willing to participate in decision-making. Working with issues of the patient's and family's choosing maximizes their strengths, and shifts the locus of control from external to internal. A patient-held record is a positive and visible way of creating this shift and enhancing communication between professionals.[47]

Providing adequate and timely information, suggesting separate means for patients and their families to become knowledgeable (e.g. helplines, books/leaflets, self-help groups), means that realistic and informed choices can be made. Making a decision to enter a hospice or another establishment either for respite care, symptom control, or terminal care will often evoke powerful conflicting feelings in an individual whilst simultaneously creating differences of opinion between the patient and family. Planning ahead wherever possible and giving families and individuals the support (alone and together) and time they need to make a mutually agreeable decision is vital. A visit before admission may help dispel myths or fantasies about an institution and the care it offers and access to the hospice's day therapy unit will enhance this and open up opportunities for the family to access support. Utilizing those parts of the home care regime that can be transferred whilst acknowledging the distinct difference between the two locations (e.g. ward routines, health and safety requirements) will establish a care contract after admission based on mutual knowledge and agreement. Individuality is preserved when patients can continue to inform and review their care plan.

Use of a patient passport such as described in the MND Association's 'Understanding my needs' leaflet[48] enables staff to begin to understand the individual and their wishes from the moment of admission. Staff must make efforts to preserve a patient's dignity and be flexible in their practice so that individual identity and needs are not submerged in standardized protocols. Those family members who want to be involved in care during the stay should be included in discussions; this will facilitate a transfer of skills and ways of working between the two locations and help alleviate any feelings of guilt that the relative or carer may have.

Fears of death and dying

Case history

Audrey, aged 76, asked to speak to the team in clinic about her future. Her daughter who was present with her became distressed and it was agreed that the social worker would take her to another room to give her the opportunity to express her concerns. Whilst she was away Audrey asked how and when she was likely to die and she was relieved to be told that death usually followed the weakening of the respiratory system and that she would probably

gradually lapse into unconsciousness and peacefully stop breathing. She was also told what medication would help if things should be more difficult. Whilst appearing sad she was very pleased and relieved to have been given this information.

At all stages, but especially as death approaches, the person with ALS may wish to discuss both the process of dying and death itself. It is important to indicate a willingness to discuss difficult topics, but not to press the point with those who are reluctant. Appropriate questions may both offer prompts and legitimize discussion: 'Do you ever find yourself thinking about the future and how things will go?'. Anticipation of future losses can be as worrying for patients and carers as current ones.[49,50] Breathlessness, pain, incontinence, and dying by choking may be some concerns. Many will welcome information about how deterioration will be recognized and what symptoms mean in terms of progression of the illness and survival. They will value information about physical aids and adaptations, help in the home, and medications for symptoms. Written information[51,52] may be helpful, but reassurance is vital as fears are often greater than reality: for example, only one patient out of over 200 at St Christopher's Hospice died in a choking attack.[53] Fears about the future may lead some to conclude their new world is unbearable and judge an early death to be preferable.[54] Cues can be provided: 'Does your situation ever make you feel like it's not worth going on?'. A positive response requires serious exploration of the issue and any fear, such as dependency.[56] Drawing up an advance directive (whether legally enforceable or not) may help with a sense of control. Whatever the legal position about euthanasia, good palliative care will inform and expand patients' options. However, as Sheldon reminds us, it is important [that] assurances are given that despite any disagreements about this issue, the team will continue to offer their skill and experience and will strive to respect the patient's wish to remain in control in all other areas, provided these do not infringe the autonomy of others.[11]

Careful risk assessment, incorporating multidisciplinary working, must differentiate between those whose wish to die is rational and those suffering from mental ill health. The extent to which clinical depression exists in ALS is disputed (see Goldstein and Leigh[45] for a summary). Talking about the future is a sensitive matter, but a brave professional can provide patients and their families with the opportunity to discuss issues such as preparing a will, future care of children, and options about the desired place of death.

Coping strategies

The complexity of losses, including plans, privacy, skills, and changed relationships, all place great stress on someone with ALS and those close to them. It is

more surprising that 'in the face of an incurable disabling disease . . . the majority of people' are 'neither depressed nor even in serious psychological difficulty'.[18] To cope with the demands, patients, family members, and carers adopt, often unconsciously, strategies to make the situation bearable. These strategies will change depending on the difficulties encountered and their current perception of the illness and disability.[57] A patient may use different coping strategies from his or her family or carers.[58] The way in which individuals deal with stressful events will partly be determined by their past and the extent to which threats have been successfully combated before. Horta[59] remarks that 'neither the initial nor the ongoing emotional reaction of patients to ALS is merely a "here and now"' response' and professionals can use the assessment, in part at least, to discover an individual's past coping abilities. This process may help them and the professionals involved to recognize their strengths or alternatively their need for support.

The use of denial is common in situations in which a person's known world and integrity are threatened. It replaces the reality of the diagnosis and its implications with an alternative reality. It is a valid and perhaps necessary coping mechanism when adjusting to loss (see Task 1 in Worden[60]). Horta summarizes that 'denial is a protective and primarily unconscious method of retaining psychological equilibrium'.[61] One daughter of a person with ALS remarked that 'denial . . . is a rather convenient and tidy way to control the amount of reality to be dealt with at any one time'.[61] Putting pressure on someone who is resistant to a different perception of the illness may reinforce the denial, although Hogg et al.[62] showed it to be predictive of psychological ill health. Having regard for the patient's family and carers is essential in detecting clashes of coping styles and needs.[63] Scenarios in which a patient talks of getting better and going back to work whilst his or her partner wants to plan for a different future can cause tremendous tension and resentment. There may be limits to an outsider being able to establish consensus between the two; equal respect for each individual's position is the starting point. Thereafter, providing opportunities for the individuals, alone and together, to discuss the other's views may help maintain a relationship when they need one another most.

People who use denial present dilemmas for everyone (non-professional and professional alike), particularly as those who rely upon it may be more physically impaired.[64] Many use it as a short- or medium-term measure, along with a need to take one day at a time to reframe their world and future. This allows a patient to control their life and emotions by no longer thinking in terms of pre-ALS life but rather finding meaning and purpose in the present.[34] One patient talked of feeling more positive as a result of a support group for ALS patients. He realized that he was not 'as bad off' as other patients. Peer group support may also afford exchange of helpful ideas between patients and families. People

with ALS can be helped by active problem-solving for themselves, for instance by alternative or complementary therapies.[18] 'Intellectual stimulation'[34] can be positively affected by professionals who enable patients to access information. As a patient's condition deteriorates, page-turners or access to the Internet can maintain a knowledge base and contact with other ALS patients through online discussion groups. Some people with ALS can be horrified at the prospect of being together with other people with ALS whose greater degree of disability and dependence would cruelly predict their future. This different perception of the usefulness of a particular activity underlines the point that individuals will select, often unwittingly, ways that help them to carry on living with stability. O'Brien[64] revealed how this control over information can be sabotaged by unsolicited sources such as the media. Psychosocial care can help to expand an individual's coping repertoire[65] by suggesting ways that have helped others cope. Recognizing the value of normality, and taking time off from the disease, has parallels with Stroebe's dual-process model of coping.[66] It can be reflected in a practitioner's intervention by not just concentrating on the disease and its consequences but by also showing genuine interest in whatever else the person wants to talk about. The sensitive use of humour with patients not only conveys warmth and humanity but mirrors how some people with ALS maintain personal relationships and reframe terrifying prospects.[34]

Family, carers, and friends

All patients have a family, whether they live alone, are a sole survivor, or part of a large intergenerational group, and family and friends are essential for a patient's quality of life.[66] The family is a complex system that changes over time, and has a past and a future that exerts pressures on the present. Patients will also connect to other networks containing significant relationships, some of which may be more important than biological links. The entire patient network exists within a social and cultural context that helps define the possibilities for the individual and their family.[67,68] Cultural expectations about the roles, rights, and responsibilities of individual family members also have an impact.[11,40,69]

Help for the family can get lost in anxiety for the patient. Family life will deteriorate along with that of the patient, but the patient and family members have different needs at different times, require different types of support, and may have conflicting agendas.[44,57] Care for the family has an important preventive health component. In the literature depression rates in ALS family caregivers lie between 23[70] and 61%.[71] Neurobehavioural symptoms[72] and emotional lability of the patient[73] are extremely distressing for caregivers, seem to have a profound negative impact on caregivers' psychological status, and were related to caregiver burden. Lillo et al. found that abnormal behaviour was the strongest predictor of

high caregiver burden—rather than physical disability.[74] In addition, symptoms of respiratory insufficiency are distressing for caregivers[44,75] as well as missing or insufficient social support[76] and the lack of time for oneself.[77]

Family members will live on into a future shaped by their experience of the patient's illness and death. With help they can emerge strengthened into a changed but safe family future. Kissane and Bloch's[78] research on families affected by cancer emphasizes the importance of family functioning during the illness for adaptive grief outcome and the pivotal importance of psychosocial care.

Carers' needs

Carers face conflicting demands, for example juggling employment and children with the demands of physical caregiving. They also receive bewildering advice from friends, family, and professionals and may pay a price for support received in invasion of privacy and potential criticism.[79] Changes in family structure may also add to the burden, for example divorce, split and reconstituted families, geographical distance. Practical support is a priority.[80] Many families struggle with rapid, frightening changes in the physical needs and capacities of the ill person and the physical caring tasks. They require knowledge of what is available, how to obtain it, and advocacy to obtain it in time. Sykes et al.'s[81] study demonstrated how often practical supports arrived too late.

Carers require:

- value given to their experiences
- provision of adequate information about diagnosis, health status, prognosis, trajectory, and treatment options
- confident, committed family doctors
- good symptom control
- adequate nursing support
- coordinated care that is individual and flexibly delivered
- access to specialist care
- guidance with practical aspects of care, personal care needs, and household tasks
- respite care within an inpatient or a home sitting service
- training in skills to enhance patient comfort
- financial support
- advice and information on the services available and help to secure them
- counselling
- emotional support directed specifically at the carer.[79,82–86]

Individual time for carers needs to be negotiated at the start of the relationship so that it is part of the contract rather than only being offered when difficulties arise, when it may be harder for the carer or patient to accept separate meetings without suspicion and guilt. Rabkin and colleagues[8] demonstrated high concordance between patient and caregiver distress and suggested that attention to the mental health needs of caregivers may alleviate patient distress. As Payne[87] notes, it is important to balance accounts of carer burden[45] with the positive aspects and rewards of caring. Whilst carer assessments should be provided when the patient is assessed for care, they are often not; statutory organizations should ensure that carer assessments happen.

Caregivers can be taught how to do what they can, while letting go of the need to control the outcome. Caregivers may need instruction in setting and maintaining clear limits and boundaries. Many caregivers have never learned either the legitimacy of setting limits or the means to do so.

What helps patients and their families?

Specific psychosocial interventions for family carers of ALS patients are still lacking, but some common principles of psychosocial palliative care can be mentioned:

- ensuring clear adequate information and an opportunity for questions
- acknowledging emotional pain and anxieties and facilitating their expression
- permission to grieve the person who was
- reassurance about powerful unfamiliar feelings
- timely interventions that anticipate fears and problems (generalization can help here: 'Many families tell us . . . What is it like for you?')
- offering acceptable frameworks for why people are behaving as they are
- confirming coping and acknowledging positive aspects of caring
- helping the family decide what is important and giving them the confidence and/or resource to act upon it
- addressing uncertainty and the distress it causes—simple open questions that acknowledge feelings can help: 'What is the worst thing at the moment? Who are you most worried about?'
- affirming individual needs and offering help in ways that are acceptable to the individual.

Social problem solving and religiosity/spirituality were strong predictors of caregivers' quality of life.[88,89] Therefore, developing interventions to teach ALS caregivers effective methods of problem solving and interventions to find meaning in this distressing time would be of benefit to them.

The UK MND Association's Carer's Pack is full of helpful practical information. Their Carer's Strategy recognizes that acknowledgement of and support for the carer are equally as important as support for the patient.[90]

Special pressures occur in families where the patient has lived with ALS for over 5 years.[28] The preliminary findings by the Northern Regional Care Advisers of the UK MND Association UK in 1999 indicate that most carers had been told the prognosis was relatively short. This led to them give up everything. When the illness stretched into 5 or more years they felt enormous guilt about being unable to cope. Equally the patient felt in some cases that they had wasted the life of their partner.

Family meetings can anticipate and address difficult decisions and conflict.[91] Some guidelines are:

- Effective preparation—who should attend, where it should be held, potential objectives.
- Decide how to begin—explain the rules, for example the right to speak for yourself, and give some idea of the time frame.
- Speak in language that everyone can understand and check this frequently.
- Be neutral and find out how everyone defines the problem—all need to feel that the professional understands their viewpoint.
- Define problems in a positive form, e.g. 'You love your children and you want to protect them'.
- Anticipate and acknowledge differences and conflicts—try to find similarities: 'You are both feeling lonely and resentful' . Help people negotiate and compromise, which may mean helping them retreat from fixed positions.
- Be realistic and encourage focus on concrete, specific, achievable goals.
- Check agreements, rehearse potential difficulties, and give clear summaries.
- End the interview in a safe place; refer to a lighter topic, use humour etc.
- Do not do all the work—remember the aim is to help the family solve the problem in a way they feel comfortable with, not to sort it all out for them.

Marital relationship and sexuality

Individuals within a relationship experience ALS very differently. In McDonald's survey,[28] spouses were lonelier than patients and often experienced psychological or spiritual distress at different stages of the illness. Ginsberg[92] discusses how anger and frustration at the loss of independence causes demanding and regressed behaviour on the part of the patient, provoking resentment in family members. Marital boundaries and roles blur and change. Old issues of power and control re-emerge in the anger and frustration engendered by the illness.[83,93] Wasner and

colleagues[94] found that, compared with the time before disease onset, there was a decrease in sexual interest and sexual activity and an increase in reported sexual problems. The problems reported were mainly decreased libido, passivity of the partner, and the patient's own passivity. Around half of the respondents in that study reported an improvement in their overall relationship through ALS, and in some cases even an improvement in the sexual relationship. For more than one-third of the couples it was still an important issue and the current marital relationship seems to be influenced by patients' psychosocial functional impairment and by feelings of burden. In addition, the quality of the pre-illness marital relationship is a significant predictor of ongoing marital relationship in both ALS patients and their spouse carers.[95]

It is important that professionals help couples retain a sense of intimacy and enjoyment in previous shared activities, even if this requires paid carers to take over some physical provider roles. Carers will need encouragement to take a break from caring and pursue individual activities. Intimate relationships and body image may be deeply affected by ALS, yet are largely neglected in the literature.[96] Issues of intimacy and sexuality are about much more than intercourse. They concern fundamental needs to communicate and receive love, to feel at ease with their bodies, and physical closeness. The combination of sex, disability, and death is a powerful inhibitor. The anxieties of professionals about their own sexuality may lead to defensive designation of the whole area as one that requires a particular specialism. Yet if professionals remove sex from the agenda they can isolate people further from the help they need to gain the love and acceptance that will support them in losses in so many other areas. In reality, the communication skills required are the same as for any other sensitive topic. Indeed a recent palliative care study[97] found that patients with all types of cancer were willing to discuss the impact on their sexual lives. The research of Kaub-Wittemer et al.[44] confirms that sexuality is important for many ventilated ALS patients, although less so for their caregivers.

Case history

Jo was 33 when he was given a probable diagnosis of ALS/MND, and was rapidly convinced that he must take very early retirement with a very poor pension package from his employer, having been told it was the best possible in view of his not having a 'definite' diagnosis. To visitors he presented a cheerful accepting front, but when alone with his wife Jackie he was depressed, tearful, and frequently angry.

He declined counselling support for himself but requested it for her. On seeing the social worker she expressed her anger and distress both that he would not accept his emotional difficulties and that his family were constantly thanking her for 'sticking with him following his diagnosis'. They had been married for 3 years and she felt there was no question of her not being with him.

Intimacy and sexuality

Although most professionals cannot be expected to become specialist psychosexual counsellors, all have a responsibility to become comfortable about offering and responding to cues about sexuality, offering first-line help, and referring for specialist support.[98,99] In Vincent et al.'s[100] study of women with cervical cancer, 80% of those receiving treatment wanted more information about the impact on their sexuality, but 75% said they would not raise the question first. Vulnerable individuals often give the professional a cue to test out whether he or she is safe enough to receive their confidences. An example would be the man who said of his wife: 'She just doesn't seem to love me anymore'. Bland reassurances may confirm that this subject is indeed too difficult and painful to discuss. Graded, open questions may help: 'How has your illness changed your work life/home life/life as a couple/ability to get close to one another physically?'. It also helps to generalize and to give permission: 'People often have questions they'd like to ask about the sexual side of life'. Monroe offers a useful set of history-taking questions.[10] Annon's widely referenced PLISSIT model (which stands for four levels of treatment: permission, limited information, specific suggestions, and intensive therapy) also provides an incremental approach.[101] This model has been recommended as a useful template for the assessment of sexuality and sexual health in palliative care settings. The model provides a general framework on how to initiate a dialogue about sexual issues and how to further the discussion if warranted. More recently, Taylor and Davis modified Annon's model by developing the extended PLISSIT model (or Ex-PLISSIT).[102] The Ex-PLISSIT model suggests that the permission level should involve requesting permission to discuss sexual issues as well as permission to be a sexual being, and this should be integrated into each step of the model.

Concerns may be expressed about body image, sexual function, or both. In some relationships the physical dependency created by the illness can disturb a well-established pattern of lack of intimacy. Some partners find that their change of role to carer alters their feelings and sexual desires, or that the physical deterioration in a partner creates similar changes. Many patients feel anxious about the changes in the way their body looks and behaves. Anxiety about rejection can lead to retreat and a wall of silence which may be eased by a facilitated discussion.[94] Some couples appreciate advice about alternative positions or alternative methods of love-making such as mutual caressing. Professionals should avoid assumptions based on age, gender, culture, marital status, apparent relationship, or experience. Those without a partner may discuss the relationships they had in the past or those they now fear they will never have.

Children and their needs

When someone in the family is ill, everyone is affected, including the children. However, the desire of adults to protect children may leave children alone and confused, at the mercy of their fears and fantasies, which may be worse than reality. Children are always aware when something is happening in the family. They sense adult anxiety, overhear conversations, are aware of practical changes, and often hear adult gossip from school friends. Many research studies show the cost of inadequate support and involvement for children facing bereavement.[103–106]

Children need:

- Respect and acknowledgement.
- Information about what is happening, and why, and what might happen next—information needs to be clear, simple, and truthful, and must be repeated as children struggle to come to terms with what is happening.
- Reassurance—children become frightened as they watch a parent or relative becoming dependent or emotionally labile or irrationally angry; they need an explanation, to know that they did not cause the illness and cannot catch it.
- Children will want reassurance about practical issues such as what is going to happen to the family and their own care after the person has died.
- Appropriate involvement in helping the patient.
- A chance to talk about feelings and know facts with adults who are prepared to share theirs.
- A variety of media for self expression, e.g. drawing, writing, playing games.
- Opportunities to reflect and remember, to know that life will go on and it is all right to have fun.
- To be consulted along with the other family members regarding all major decisions.

Many studies confirm the importance of pre-death experiences in mediating and influencing the course and outcome of bereavement.[102,103] Significant factors include the relationship of the child with the ill person before the death, the openness of family communication, the availability of community support, and the extent to which the child's parenting needs have continued to be met.[107]

Caring adults are often struggling with seemingly impossible and conflicting demands and may need help to negotiate compromises. For example, a parent struggling with the impending death of a partner may lose sight of their child's fear that they are losing not one parent but two.

Helping parents

Parents know their children best, and children need their families—who will be around long after the professionals. The professional task is to support parents to help their children. Professionals should work with what parents can manage and help them to develop a sense of confidence and competence.[108,109] What, and how, children are told about the situation is the responsibility of their parent(s) or guardian. Parents will need encouragement and support to understand how best to communicate with and involve their children. They may have good reasons for their reluctance to share information about illness and death with their children. They are often struggling to maintain control in the midst of uncertainty and may feel both physically and emotionally overwhelmed. They may be avoiding the truth and wonder if they can cope with the child's grief when they are anxious about managing their own. They may under-estimate what a child understands and worry about saying the wrong thing or making matters worse. Research indicates that children as young as 3 years begin to understand what the word death means.[110,111] Children informed about a parent's impending death are less anxious (Rosenheim and Richter 1985 cited in[99]) It is important to support parents with their own emotional needs before they can contemplate those of their children.

Parents often welcome the opportunity to think about what their children might ask and how they might respond. Christ's[112] meticulous study confirms the value of helping parents to understand and respond to their children's developmental needs. Parents will need advice on how to anticipate and understand altered behaviour in their children; why they are more clingy, do not want friends to come home, or seem to be frightened or embarrassed by the person who is ill. For many parents, rehearsal of the issues with a professional will be sufficient and they will want to speak to their children alone, perhaps with a later opportunity to review the conversation and concerns raised. Others will welcome sharing the task with a professional and may gain confidence from the presence of a doctor, nurse, or social worker as they try to answer their children's questions. Parents can be helped by being offered appropriate resources, such as suggestions of books to read to their children, or for their children to read. An example, produced by the British MND Association is a work book for parents to use with their younger children.[113] This is in a loose-leaf format to enable the parent to remove and add in sections as they wish.

Parents may need encouragement to widen their children's support network by involving other adults close to them: a relative, another adult friend, a youth club leader, or, most importantly, the school. They may need support with strategies to manage other friends or family members who are resistant to involving

the children. Everyone will need reassurance about children's resilience. The aim should be to help parents talk to their own children themselves. In the rare cases when this is impossible and the professional undertakes direct work with the child, care must be taken to work within family values and culture. The professional must discover what knowledge is permissible in the family and what words have already been used, and should negotiate an agreement with the child and parent about how and what to tell the parent about the content of the session.

Young people

Adolescents face a particular struggle between balancing independence and attempts to find a new identity and an event that draws them back into a changed family and additional unwelcome demands.[114] They often value a separate opportunity to talk to a professional about the illness, and may also want to discuss difficulties in relationships with their friends. A serious illness is frightening and embarrassing for their peers, making them different just when they most want to be the same. In separate work with adolescents it is important to have clear agreements about confidentiality with them and their parents. Parents need help to understand that a young person's withdrawal is not just careless or selfish. Information about feelings can help young people feel more in control, especially in written form so that it can be read whenever the person chooses. The UK MND Association has a good example: 'So what is MND anyway?',[115] It is important to remember that talking is not the only solution for helping young people;[116] keeping a diary, watching films, reading books, playing sport as a release, or practising relaxation techniques all help adolescents express their feelings. Above all young people need affirmation wherever possible. Professionals must also be aware of the needs of young people and children who are acting as carers, often in single-parent families. Frank[117] offers useful principles and guidelines for this.

Working with ALS

Many professionals and volunteers have commented on the emotionally taxing nature of working with people with ALS[28,92,118] but they also report high levels of job satisfaction.[119] ALS may confront professionals with fears about their own mortality[120] and their own death. Carroll-Thomas[121] reminds us that how we intervene in and react to a family will be based upon our own values, psychological make-up, and professional culture. She asserts: 'The more challenging the problem, including feelings of helplessness, the more likely that cultural values will emerge in clinician and patient interactions'. Professional attitudes and

values can affect the allocation of resources, and choice of language may affect the quality of information given. Multiprofessional teams extend the services and options for the patient and family, optimizing coordinated care and an effective response to complex needs,[45,122] and often reducing the number of separate visits required and the need to constantly re-tell their story. Research demonstrates the impact of teamwork in better care outcomes.[124,124] Working in teams also offers the chance to share dilemmas, ethical questions, and stresses of caring for people living with ALS. Even someone who works alone can create a team by thinking flexibly and imaginatively about those involved. Teams must respect the autonomy of the individual and the family's own style. Professional views on safety may need to be balanced against the wishes of the patient and family and the importance of allowing families to do things in the way they choose. A multiprofessional team should include someone specializing in psychosocial care, not only for the needs of the patient and family and sometimes to advocate for them in team discussions but also to take a lead in the care of the professionals; for example, encouraging appropriate personal disclosure and team review of complex pieces of work. Papadatou[125] explores the grief experiences of healthcare professionals and emphasizes the importance of team-based meaning-making.

Individual professionals have a duty of care for themselves. They need to acknowledge their feelings, and when necessary find support for them.[126] Stress reduction techniques may be one possible way to cope with this challenging job.[119] Carmack[127] has some helpful insights on balancing engagement and detachment in caregiving. She comments that the longer and more intense the caregiver's involvement, the more important it is to learn this balance. A booklet from the UK National Council for Palliative Care 'Difficult Conversations. Making It Easier to Talk about the End of Life with People Affected by Motor Neurone Disease'[128] may prove very useful.

Conclusion

For now ALS remains incurable. Psychosocial care plays a particularly important role in determining how patients and their families respond to the impact of illness. It can help them cope with experiences of loss and change and expand their sense of what is possible. McDonald states:

> In many cases quality of life had little to do with physical disability. Many patients and families maintain high quality lives at all stages of physical disability and all lengths of illness. The key lies in their psychosocial and spiritual well-being.[28]

The psychosocial care described is not a luxury, but essential in the kind of effective palliative care that can help patients and those close to them move into a changed future with a sufficient sense of confidence.

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Chapter 11

Spiritual care

Robert Lambert

Summary

Spiritual care is of great importance in the treatment of ALS. It is essential to consider the importance of spiritual issues, within both the illness and its treatment, to the whole person, rather than the physical aspects alone. Spirituality is defined to include both religious and non-religious elements. Spirituality can be considered as a resource for coping, as a potential source of conflict and suffering, and as an arena to explore in finding meaning. There are many different universal spiritual themes which may be expressed in ALS: faith, hope, a sense of the sacred, meaning, gratitude, forgiveness, vocation, and acceptance of death. We need clinically based, non-denominational spiritual care and it is important for all clinicians to be aware of their patients' spiritual values.

Case history

A woman of 45 was diagnosed with ALS. Raised in a strict Christian family, she had a mostly negative experience of it as a severe religion and no longer practised. But in her mid-30s a friend invited her to a lecture on Buddhism, which piqued her interest. She was drawn to the accepting environment and to the ordered thinking of Buddhist teachings. She began practising meditation and attending lectures, until eventually she identified herself as a Buddhist. Here is how she makes sense of her illness:

> I am fortunate that I was already practising when I got ALS. Where I grew up they would have told me that this was a punishment from God. But it's not. This is not from God. In Buddhism, life itself is suffering. And this is my suffering. It will transform me. Everyone has to live with suffering but this is just life. My life won't end with this suffering, only this life. I must finish this life with the right attitude and in peace so my soul will continue in a higher life later. I am suffering now, but I'm not attached to this suffering. I can see beyond it. That is why I meditate, to detach and be at peace.

Paradox

People with ALS live with paradox. Paul the Apostle wrote some 2000 years ago that 'In the midst of life, we are in death', expressing this universal human dilemma. Knowing that we shall die, how do we make sense of life? We manage to keep the thoughts of our own death out of conscious awareness most of the time. We may be reminded of it while watching a film, attending a relative's funeral, or waiting outside the surgery on a visit to the doctor, but we manage to push it aside and carry on with life. A diagnosis of ALS forces this sudden awareness through the compression of time, the daily reminders of physical decline, consecutive losses, and the reactions of loved ones. A second paradox is expressed in ALS. Not only are we more than our bodies, it is sometimes in the absence of physical capacities that people are most profoundly alive. This was put simply by a friend a few years ago as he approached his own death due to a brain tumour: 'It's odd, but I feel just as alive now that I am near death as I was years ago when it was still far off . . . maybe even more so'. Here we are in the realm of the spiritual—that aspect of life that has little to do with physical quality or capacities—that which is at once most dynamic and enduring about a person.

What do we mean by spiritual?

In an earlier age the terms spiritual and religious were synonymous, and everyone knew what they meant. It is no longer so simple. The advance of the sciences, secularism, and critical scholarship in religious studies have separated these terms in the West. Now religion is more often associated with public practices, such as membership of an institution, participation in rituals, and adherence to the teachings of a tradition. Spirituality has become associated with the private realm of one's personal experiences, beliefs, and values.[1] Both refer to the inner life of a person, but religion is more likely to be shared with others and to bind one to a culturally defined group. Religion can be described as organized, institutionalized spirituality.[2]

Spirituality seeks meaning in lived experience. It uncovers and tries to understand truth, not as fact but as meaning. Spirituality encompasses religious beliefs and practices but is not limited by them. We might say then that all people are spiritual; some people are also religious. Robert Fuller notes that we are spiritual whenever we wonder why we are here or what happens when we die. We are spiritual when we are 'moved by values of beauty, love, or creativity that seem to reveal a meaning or power beyond our visible world'.[1]

The crisis of terminal illness heightens one's awareness of the spiritual. With patients, I find that the spiritual surfaces in three different ways. First, spirituality

may be a resource for coping with illness. People turn within to find spiritual resources: beliefs, values, trusted experiences or images, which give them strength, encouragement, or comfort. Some may seek strength in the practice of their faith, through the support of their community, and in the guidance of religious leaders.

Second, spiritual conflicts may emerge. One person's spiritual life can be a source of strength and joy: another's can be a source of pain and suffering. The spiritual may even be a dark place filled with punitive gods, failure to live up to ideals, memories of rejection or judgement by religious figures, or frightening images of the afterlife. Some associate illness with judgement, punishment, or the injustice of a capricious god.

Third, the spiritual emerges in the searching questions which arise from within. Reflecting on their past and future, people may engage in a search for meaning. Why am I here? Why is this happening to me? What should I do with my remaining time? The questions are not only about the meaning of this event but about life itself. Illness becomes an occasion for going deeper, for reinterpreting the meaning of one's life and for understanding how this new chapter connects to all the rest.

Our spiritual self differs from our philosophical self in that it seeks to discover meanings, not to create them; and it differs from our scientific self in that it seeks to discover meanings, not causal forces. Seen from the standpoint of psychology, religion refers to those overt and covert aspects of personality in which there is a questioning drive, a quest, to relate meaningfully the particular to the whole.[3]

As one finds meaning in the present, it is possible for life to be experienced as deeper, richer, and more rewarding even while one is living with physical decline.

Spiritual themes

People with ALS do not face different spiritual issues from everyone else, but a terminal illness intensifies spiritual needs and questions. The spiritual issues I will discuss in this chapter are universal ones. All religions have their ways of addressing these issues and each person, religious or not, seems to confront them sooner or later. The field of pastoral care, in its evolution into a clinical discipline, recognized the advantage of seeing these themes through a clinical lens rather than a partisan one. Hiltner's suggestion of a pastoral diagnosis,[4] and Pruyser's proposal of 'diagnostic variables',[5] were great steps forward toward viewing spiritual themes through clinical eyes. This list of spiritual themes is neither unique nor exhaustive, but it describes the themes that I have heard repeated while accompanying patients with ALS.

Does life make sense? The existential question

Why is this happening to me? What will become of me? The lack of a scientific explanation for the cause of ALS is not at the root of these questions—people with other illnesses ask the same questions. The question is one of meaning and not of causality. When someone hears the diagnosis of a terminal illness there is a sense of shock, of being knocked off balance. What one had expected from life has been taken away and replaced by another scenario, difficult to believe and even more difficult to accept. As the news sinks in, questions about meaning emerge, and behind these questions are the greater questions about life itself: What is life all about? If this thing that makes no sense is happening to me now, does anything else make sense?

Religious people may speak of God's will, God's plan, or a divine purpose. The monotheistic religions speak about the goodness of God, the created order of the universe, and natural laws. Although it is mysterious, somehow illness can and must be accounted for within those thoughts. Ancient writings like the book of Job are attempts to make sense of illness within this frame. Other religions have their way of describing the order of things. Buddhism's four-fold noble truth begins by stating that to live is to suffer. The nature religions fit illness and death into the natural cycles of the earth. Religions speak of a world in which there is a meaningful connection between people and events.

The spiritually inclined may look for a benevolent goodness, an intelligent design, or at least a natural order. Inherent in all of these approaches is the idea of providence, of the goodness of the created order, despite all possible appearances to the contrary. Fleischman refers to this as the need for lawful order.[3] If there is an order to things, then even our crises and hardships fit into this purpose in some way that we can discover.

But without some sense of goodness and order what can we trust, what is worth doing, what is good, and what is not? The seemingly random nature of ALS is difficult to fit into such an order. The age-old questions are raised, as they are when people are hit by a tsunami or a hurricane. But ALS does not happen to a geographical region, it happens to one person. How could God allow this to happen to me? Some find a way to make sense and others do not. A man who was diagnosed with ALS at age 66 said this:

> I am sad to be nearing the end. I love life. It's good and it has been good to me. But it has to be my turn sometime, and my turn is now. That is the way life is made. It's my turn to go, and someone else will be next. I look around and think, what can I expect? Sixty years seems young, but when you look around the world, you know I've lived longer than I would have in most places! So this is the way things are designed and I have to accept it.

Is anything sacred?

The wife of an ALS patient told me that her husband's illness was so distressing for her that she could find comfort only when she attended the service at the Anglican church. She and her husband had been going there for more than 30 years but he could no longer come with her and she missed him beside her there. She described how she would lose herself in the music of the organ and the choir and find a refuge in the familiar sights and symbols of the liturgy. At the moment of receiving communion she felt God was present and she would be all right. I asked her husband if he missed the services. He said he had gone mostly to support his wife, but it was not there that he felt the divine presence. What he missed were the late summer evenings at their cottage on the lake. There surrounded by his family and watching the sunset he felt a sense of awe and reverence that gave him peace inside. 'The church is fine', he said, 'but it's there at that lake that I know we are not alone'.

These two halves of a couple demonstrate very different ways of experiencing something unifying and similar: the sense of the sacred. Pruyser[5] noted that nearly everyone has some sense of the sacred, the awareness that there is something holy, beyond the human realm, if asked the question: 'Does anything give you a sense of awe and reverence?', some will describe overtly religious experience, while others speak of nature, music, art, or of some aspect of human relationships such as love, fidelity, or trust. A patient described a new interest in listening to birds on his deck in the mornings. Before he had ALS he had neither the time nor the interest to notice the natural world around him; now each morning he felt accompanied by all of life.

In experiences of awe, reverence, and transcendence people touch what is sacred to them and feel a connection beyond that of the body. Such experiences are not merely a resource to help someone to live with illness, but a way of living more deeply with more satisfaction than before.

Faith

The word faith is often used to describe something static, an intellectual idea, a doctrine, or an allegiance to a particular set of beliefs. We say, 'I have faith (believe) in God' or 'She is of the Muslim faith'. But faith also describes something much more dynamic. Faith is a person's stance toward life. It refers to the motivating force within a person, to what drives them and nourishes them to continue. The theologian Paul Tillich spoke of faith as 'the courage to be'.[7] Faith is also related to trust and confidence. I asked a patient about her faith:

> I have faith in God that he will take care of me. God always takes care of us and it's always through others and their kindness. In my life I have tried to do the same for others because I know that God works through me for their sake. So now it's important to trust. I have to trust that my husband and daughter will continue to be there for me. The aides who come to help us each morning and night, I have to trust them. There is a lesson here for me somewhere and I will have to learn it. My faith gets stronger every time I see someone reaching out to me.

Another patient described how he looked within himself for the courage to continue. His children and his wife wanted him to live as long as possible. Despite his discouragement and fatigue he tried to continue for them. He said he was not religious but felt perfectly natural praying for courage. I asked to whom he prayed:

> To whatever force there is beyond us. I don't think we have to know all about it in order to pray. After all, we got here without asking to be born, so there's a force somewhere that carries us along.

Thankfulness and grace

Religions describe life as something received which points to a transcendent other from which we come. There is an unmerited quality to life, reflected in celebrations of thanksgiving, prayers of gratitude, requests for divine help, or rituals, which enact the receiving of goodness from beyond ourselves. Religions cultivate gratitude and underpin the experience of grace. Grace is the experience of receiving what one did not earn or could not merit only because of the generosity of another. One may experience grace when receiving a gift, when granted a reprieve, or even when waking up, taking the next breath, or moving forward to the next moment.

Illness can make it difficult for one to experience thankfulness or grace. ALS appears to strike in a random and unmerited way: there is nothing one could have done to have caused it and one may feel unfairly singled out for mistreatment. Grace is challenged by such an illness and can easily be replaced by resentment. Yet some patients maintain and express a sense of thankfulness about life in spite of their illness. A dignified orthodox Jewish man demonstrates how gratitude can be a spiritual strength:

> When I come here I really am reminded that life is a gift. I have had 70 very good years. I am not happy to be sick but I am 70 and I look around me and see others of 30 or 40 with this disease, and I cannot help but think I blessed I am to have had 70 years of health and happiness! Some people have said to me that this is not fair. You know, in fact, it's more than fair to me. I did nothing to deserve a longer life than some of these others around me, and I can only be grateful that I have lived so well.

Guilt, forgiveness, and change

When one is suddenly faced with a foreshortened future, time becomes precious and the past looms large. Coming to terms with the past sometimes feels urgent. In the literature about dying it is common to speak about 'life review'. As people review what was good about their lives they inevitably uncover regrets. It is important that these regrets find some expression and relief, but people may also wish to make changes and live differently in the time that remains.

Repentance, literally to turn again, is a basic spiritual need of human beings. This capacity to regret is fundamental to making sense of one's life. Without it there is no sense of agency, of responsibility for one's actions and choices. Viktor Frankl wrote in his book[8] about life in a concentration camp how important it is to choose one's actions, even when the range of one's possible choices is extremely limited. The ultimate choice even if facing death, he wrote, is to choose how to think about it.

As one nears the end of life with a diminished capacity for independent action, this issue often surfaces as a powerful spiritual need. One patient had spent his entire life at the office and made a mountain of money. It is a classic story. When his two daughters were young he was rarely home. As young adults they were angry and distant, coming to him only to ask for money. Now at 55 he had been diagnosed with ALS. A moment of repentance arrived, not because of a revelation from above nor out of religious conviction, but because his 27-year-old daughter, who had just given birth to her second child, confronted him in anger:

> You have another chance. You were never there for us; you can be there for your grandchildren. They need to know you and you have no more excuse because you're home all the time! Don't blow it!

The patient told me this story with tears in his eyes and a shy smile on his face as his daughter sat beaming beside him. She visits him three times a week with both of her children. 'You know what?' he said, 'My grandchildren don't even care that I can't walk!'. Facing the past led to peace in the present and three generations were healed in this one moment. Most stories are less dramatic and many do not end with such resolution, but it is important that people have a chance to express their regrets in words or in actions.

Belonging

Belonging is less about which religious group someone is part of it than it is about the sense of connectedness one feels to others, to the world, to life itself.

Fleischman[3] suggests that to understand a person one must ask the question, 'Of what whole is this person a part?'. Though the answer will vary widely, the importance of that question is the same. Human beings need 'a place inside of, and an orientation to, history. This need to overcome one's individual skin, one's isolation and fragmentariness, to have a group, an affiliation, a community, can be called a need for membership'. This need may be met through participation in a religion, membership of a political party, identification with a culture, language, or ethnic group, or it may be through bonds of family, neighbourhood, or colleagues.

One of the most disturbing aspects of ALS is that it separates the patient from the community, the group, the whole to which he or she belongs. Isolation as a result of progressive paralysis and immobility, loss of communicative abilities, and increasing dependence lead to isolation from former groups. When one loses the sense of belonging and 'membership' one's inner integrity and wholeness is challenged. Who am I when stripped of my relational and role identities, my participation with others and the mutuality of giving and receiving? An elderly patient described the inability to eat and the loss of speech as the two worst losses he had endured because of ALS. They were more difficult than the loss of his mobility and more upsetting than the thought of dying. He had been an intensely social man, involved in business, community groups, volunteering in several associations, and as a leader in his congregation. His extended family was accustomed to long, happy hours around the table each week. Though adequately nourished through a PEG, he now found it too painful to be present while others ate and drank. He found it unbearable to be present in his former community groups since he could no longer respond to people verbally. The loss of belonging was an attack on his self-esteem and identity. He maintained his sense of belonging as well as he could by seeing a few of his former colleagues individually in his home, but it was a sad compromise.

It is important to notice what aspects of belonging are lost for a patient and to look for ways to maintain or replace them when possible. One ventilated patient developed complications and was hospitalized for an extended period of time while awaiting placement for long-term care. Though able to communicate only through eye blinking, small hand movements, and a letter board, she developed a remarkable circle around her, of nurses, volunteers, housekeeping staff, and others. The staff became so attached to her that they held a party when she was finally transferred some months later. Every staff member who had cared for her signed the goodbye gift, hand-made by one of the nurses. One wonders if it was only her own needs that were being met, or if she was also touching the need for belonging in the staff.

Hope

In medicine hope is often linked to prognosis, which is to say, hope for survival and a longer life. By contrast, religions speak of hope beyond this life: salvation, heaven, or nirvana. What do patients hope for when they know that they will eventually die of ALS? Some continue to hope for a cure or a miracle, but most let go of this fairly soon. Others hope for salvation beyond this life. Surprisingly, even religious people are less focused on this hope than one might expect. As the disease progresses and people begin to adjust to a diminished physical state, we hear them hope more in the immediate sense: for comfort, dignity, intimacy, or for daily satisfactions. Some hope for the completion of projects or to live long enough to participate in a particular event.

One of our patients was 33 at the time of diagnosis. Warm and personable, he talked openly with the staff about his losses and his hopes. His life's hope was to be a father and raise children. He had been married only a year at the time he entered the clinic and his wife was pregnant with their first child. Shattered by the diagnosis, it was especially difficult when he realized that the disease was progressing rapidly. But within a few months he regained his determination to live. His sights were focused entirely on the pregnancy and the coming birth of their first child. 'I will be there for my wife when this baby is born' he told me. 'I must be there. Pray that I will live until this child knows me'. He did live for nearly 9 months after his child's birth. Though he could no longer speak, his whole face lit up as his wife showed photos to the clinic staff and talked about their baby.

Some hopes look further into the future. The Buddhist patient mentioned in the case history at the beginning of this chapter found hope in the teachings about reincarnation and the practice of meditation. They helped her to adapt to illness and to be less afraid of dying. She believes she will be reincarnated and continue her development in another form:

> At the beginning you just hope that they are wrong about the diagnosis and that you will discover that you don't have ALS after all. Gradually, after enough symptoms come and they don't go away, you start to realize that its true and you will have to deal with it. Then you start hoping they will find a cure. In time you realize that it will be too late, and that this is going to be your life until the end. Then you can hope in something more after this life and that is the real hope.

She explained how meditation, cultivating peace, showing compassion, and living without inner conflict would help her be peaceful at the time of her death. This peacefulness will allow her next life to be at a higher plane. She meditates several times a day to achieve this peace. In spite of her losses, the difficulties of daily life with ALS, and a shortened future, there is a genuine joyfulness about

her. She faces physical changes with a certain calm acceptance. Hope based on her spiritual life is a strength for living in the here and now.

Near the end hope can take a different form. It is sometimes heard in the longing for death as release from dependency, discomfort, or from the trials of life in a body that no longer moves or breathes on its own. People express hopefulness related to how they will die. 'I don't want to be alone at the end', one says. 'I want to go before my family is exhausted from caring for me', another. Those who have religious or spiritual ideas about an afterlife often turn to them near the end. A Roman Catholic patient hospitalized with pneumonia expressed this simply: 'I'm tired and I've had enough. Can you ask the priest to come?' In his way he was looking forward with hope.

Vocation

When a person becomes a patient and is faced with the loss of work, social role, and parental responsibilities, there is a deep crisis of identity. It is a spiritual crisis as well as a psychological one, touching not only identity but the very meaning of life. People ask, 'Why am I here?', 'Who needs me?', and 'What must I do?'. In traditional religious terms these questions defined one's vocation or calling. There is something one is meant to do. Doing so fulfils both an inward need and an outward one: the needs of someone else. Without this sense of vocation there is an emptiness and a loss of connection (belonging) with others.

A physician who was diagnosed with ALS continued to practice until the last moment possible. Near the end of his practice he could no longer use his hands. Attached to a teaching hospital, he counted on the residents in his charge to conduct physical exams, to take blood pressures, and to carry out treatments, while he looked on as supervisor. His patients appreciated his continued presence, trusting in his experience and knowledge of their cases and feeling that he cared about them. The crowd at his funeral was a sea of former patients, their families, former students, and colleagues who spoke proudly of how he had touched their lives.

Another patient, a college instructor for 20 years, found the most difficult loss to be ending his career due to disability. He could no longer function at the university because of his lack of strength but he found the days at home interminable. He was depressed and withdrawn until a neighbour, a primary school teacher, convinced him to become a tutor to two of her students. He met those two, then more until his schedule was full, meeting one or two students every afternoon to help with their reading, maths, and history lessons. His depression lifted and he spoke with satisfaction about his renewed vocation.

Acceptance of death

One of our basic needs is to make sense of our own end. Related to the need for belonging, one strives to see how one's arrival and passing fits into the order of things. Related to vocation, one looks to know if one's mission is complete. I have saved this issue for the end, not because of the obvious chronology of things but because there is a tendency to think of religion and spirituality as concerned primarily with death. While religions do have ways of addressing death, neither religion nor spirituality is focused there. Religion seeks to structure common meanings for the life of a people. Within religion the reality of death is lifted up to underscore the precious and sacred nature of life itself. Because we die, life is limited and must be valued. Spirituality, whether separate from or part of religion, is involved in deepening one's awareness and connection to daily life. It is about living more fully, more deeply until the end. Realizing one's finitude, the present time becomes precious.

Many people have observed that, paradoxically, in coming to terms with death, they are then free to live more fully. The wise old professor with ALS says to his student in the book *Tuesdays with Morrie*: 'Learn how to die, and you learn how to live'.[9] He explains how knowing that he might die any day leads him to pay attention to the spiritual: loving relationships, the universe, the things we take for granted. He continues to explain how, being confined to his room, he appreciates the window more than his students do, who can get up and go outside at will.

Another remarkable example of spiritual growth in the wake of ALS is the life of Philip Simmons. His book *Learning to Fall* should be read by all ALS patients, their families, and caregivers. Phil struggles through his disease and manages, thanks also to meditation techniques, to reach an incredible serenity and acceptance, which pervades the whole book.[10]

Not all patients come to an acceptance of death and not all learn from its approach, but many do. In the process some need to discuss how the end will arrive for them. Some are fearful of suffocation, of choking to death, or of dying alone. They need to be reassured of how their care will be provided and of their power to make choices at the end. In my experience, concerns about assisted suicide or about having life artificially prolonged often point to a need to communicate about how death will come and what it will mean. Patients need to know that their rights and wishes will be respected and they may need to find some reassurance that they are still valuable as human beings.

Some simply need to talk about dying with someone who does not turn away from the subject. To listen patiently and without reaction communicates to the patient that we are not fearful about what is happening to them and that we

believe they are capable of living all the way into that mystery which is death. Listening demonstrates that we believe in the strength that lies within them to face their own end. 'In the end, to accept the spiritual dimension of the other is to have confidence in what the other is becoming'.[6] Conversations about dying nearly always open a door to the spiritual. When facing death, Fleischman writes:[3]

> The least religious or superstitious persons will still feel themselves to be in the presence of an unknown. A door opens. Both the dying and those with them, who do not close off in fear, feel deepening love, and life itself never feels more ephemeral and indomitable.

Indeed, as witnesses to the final days, we are put in touch with just how 'ephemeral and indomitable' life is. I was particularly moved and instructed by one patient whom I had followed for several years. A few weeks before his death he related to me what had happened at his synagogue the previous week. The rabbi had invited him to come forward to read from the Torah in honour of his and his wife's fiftieth wedding anniversary. He was surprised and embarrassed at first, knowing that he could no longer speak and could barely walk with assistance. But he dutifully went to the front with the help of his son and his grandson. He was surprised again as his grandson lifted the Torah and announced, 'Today I will read from the Torah in honour of my grandfather who taught me his faith'. His grandson had completed his bar mitzvah just a few weeks earlier, symbolizing his entrance into the faith as an adult.

In these few moments were captured the fulfilment of vocation, belonging, an encounter with the sacred, and a sense of order. It was also a moment of preparation for his death. As he repeated the story he wrote at the bottom of his pad, 'Something is finished. I can go in peace'. A few weeks later he died.

Spiritual care as accompaniment

Spirituality is to 'take one more step'. Take one more step in accepting my fatigue, in accepting my limits, the limit of my intelligence, of my lack of understanding in the face of suffering . . . to take one more step with or without belonging to a religion. . . . To be spiritual is simply, wherever one is, to take another step. To accompany then is to help the other person to do just that, in the midst of their suffering, at the heart of who they are. . . . One who offers spiritual accompaniment is therefore someone who can accompany this 'restarting' and encourage that opening, while helping the other to avoid stopping at their symptoms and identifying with them.[6]

I met the patients mentioned in this chapter in the ALS outpatient clinic of a neurological hospital in Canada. My role there is of pastoral counsellor, chaplain

(the traditional term), or spiritual care provider. The field of pastoral care is evolving as the population continues to diversify and become more secular and as needs are increasingly identified as spiritual rather than religious. In our context the spiritual care provider is a non-denominational, clinical member of the multidisciplinary healthcare team.

This necessitates a clinical approach by those providing spiritual care. A clinical approach means that contacts with patients are interventions whose goals are therapeutic and supportive. It assumes that these interventions are not partisan or biased toward the religious or spiritual point of view of the clinician. It assumes that one has adequate clinical training in addition to a background in theological or religious studies. To take a clinical approach means to respect the patient's beliefs and values and to view them with a certain objectivity. The clinician must consider how a particular aspect of the patient's spiritual life might contribute to health or to illness and how it might be a factor in treatment decisions.

Participation on the team also encourages all members of the multidisciplinary team to be aware of the patient's spiritual values. In the same way that the contributions of the social worker encourage the team's awareness of social needs, conversations about the spiritual beliefs and practices of the patient brings them into focus. The whole team is invited to view the patient somewhat differently. Marie de Hennezel writes about the importance of healthcare professionals 'welcoming' the spiritual needs of the patient:[6]

> The 'spiritual request' is rarely formulated as such, but it is nearly always present, because it is actually the request to be recognized as a person, with all his mystery and his depth. This request is not addressed to 'spiritual specialists'; it is addressed to every human encountered: You who cares for me or who accompanies me, how do you see me? Am I reduced to a broken body wasting away? What value or what meaning do you place on the time that remains for me to live?

The patient examples discussed in this chapter illustrate some of the ways that spiritual issues emerge during treatment of ALS and the important impact that they can have on the quality of life. We noted that spiritual issues can emerge in three ways: as a resource to the patient; as an area of conflict or suffering; or in the search for meaning while nearing the end of life. These suggest the kinds of intervention to be offered by the spiritual care provider in the relationship of accompaniment.

When spirituality is a resource, the clinician's goal is to strengthen and support what is already present. The Buddhist patient described in our case history finds support and encouragement in discussing her practices of meditation and how they offer her inner peace. In addition, it is important that the team understand this as a core value for her, which impacts on both her decisions and how she feels about dying.

When spiritual beliefs create conflict for the patient, the spiritual care specialist seeks to play a therapeutic role. The goal of the intervention is the resolution of conflict, to the degree that it is possible, so the patient will not have the burden of spiritual pain added to his or her physical problems. The orthodox Jewish man mentioned in the section 'Acceptance of death' had previously never missed a day of saying the traditional prayers aloud each morning and evening. This gave him peace, comfort, purpose, and a sense of belonging. But when his voice became too weak to pray aloud he was in a spiritual crisis. His family, his rabbi, and the clinic staff all minimized the importance of this (it seemed obvious he could simply pray in silence), but he remained troubled. As we talked (through writing) about how this affected him I discovered that it was a deep source of shame. His own father had always said the prayers aloud, even on the day of his death. Not only did this connect him to his father and ancestors, but it also fulfilled a duty, praying aloud as a daily witness to his children and grandchildren. The importance of this loss had to be fully understood and recognized before he could move beyond this particular grief and be at peace with silent prayers.

In other situations the spiritual care specialist is engaged in an exploration of the meaning of what is happening to the patient. This is not to find the cause, but to deepen the experience of life, even in the midst of serious illness, by finding meaning and connecting this moment to the rest of life. The man who talked of listening to the birds on his deck each morning also observed that he had a new interest in his children. 'I'd rather have come to this without getting sick' he said, 'but maybe it took getting sick for me to become a real father. It sounds strange but actually I'm happier than I was a few years ago'. This was a spiritual discovery. However long or short his life, each day has more meaning than before.

Scientific evidence

Since the beginning of the twenty-first century, the importance of religiousness and spirituality for the quality of life of ALS patients and their caregivers has been repeatedly highlighted in the scientific literature. Several studies have shown that the individual quality of life in ALS patients is not dependent on physical function, but rather on psychosocial and spiritual factors.[11,12] Spirituality and religiosity play a major role in treatment and end-of-life decisions in ALS.[13] In addition, a study looking at personal values in patients in palliative care (half of whom had ALS) found that all patients showed a shift towards self-transcendent (as opposed to self-enhancement) values. The highest-scoring values were benevolence and universalism.[14] Given the fact that a patient's quality

of life is closely related to that of their caregiver(s),[15] it comes as no surprise that the existential well-being and spirituality of individuals with ALS are also related to the psychological well-being of their caregivers.[16] In turn, religiousness is positively associated with the quality of life of ALS caregivers.[17] A study looking at meaning in life in ALS patients also found a significantly higher impact of spiritual and existential issues on their perceived meaning in life as compared with the general population.[18] More research is needed to better clarify the relationship between religiosity, spirituality, and wishes for hastened death, which are known to be present in several ALS patients, especially in advanced stages (see Chapters 4 and 17).

Conclusion

This brief review of the spiritual issues present in patients with ALS illustrates the importance of spiritual care as an integrated part of multidisciplinary team care. Of course there is a role for clinically trained spiritual care specialists. More importantly, there is a need for all who are involved with treatment to be attentive to the spiritual meanings expressed by our patients: to be present, to listen, to respect, and recognize the inner importance of what each one is living until the last moment.

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Chapter 12

Multidisciplinary care: physiotherapy

Ulrike Hammerbeck and Emily Jay

Summary

The physiotherapist plays an important role in the coordinated multidisciplinary management of treatable symptoms in a progressive, untreatable disease.[1–3] The assessment of physical and respiratory symptoms and provision of advice, exercises, management strategies, and aids allow adjustment to altered demands of activities of daily living.[1] An improvement in the quality of life is the principal aim of intervention in ALS and can be achieved by person-centred care, shaped to the goals of each individual.[1,4–6]

Introduction

The main presenting sign of ALS is asymmetrical muscle weakness[1,2] which can be rapidly progressive, leading to functional limitations and reduced exercise tolerance.[7] Presentation and progression can vary significantly according to the site and extent of motor neuron damage, and therefore management has to be tailored to specific needs identified during assessment.[8]

The initial physiotherapy assessment provides a baseline and establishes current needs by identifying impairments as well as limitations in activity and participation.[9] It focuses on the musculoskeletal system with regard to muscle length, power, and tone, joint range of movement (ROM), as well as respiratory function, activity levels, and fatigue.[10] Difficulties with mobility and activities of daily living are established, as well as perceptions of quality of life.[4] Treatable symptoms of the disease are identified and interventions planned. The findings provide information to assist the multidisciplinary team in coordinating care and are used for monitoring and predicting progression throughout the course of the disease. This allows timely advice regarding anticipated changes in disease presentation.[5]

The findings from the initial assessment enable the therapist to assist the patient in setting treatment goals, allowing the individual affected by ALS to

maintain autonomy of care.[8,11–14] Treatment goals enable the physiotherapist to plan an appropriate, comprehensive, and meaningful rehabilitation programme and to review achievements and change.[1] In the initial stages of the disease, therapy goals may be aimed at optimizing function and independence, and with disease progression the goals change to focus on maintaining functional mobility by provision of aids and maximization of quality of life.[8]

Physiotherapeutic interventions in ALS are aimed at:[10]

- advice on activity levels and provision of an exercise programme to increase exercise tolerance and fitness[15]
- maintenance of the musculoskeletal system in terms of ROM, tone, and muscle strength[15,16]
- maintaining mobility[1,17]
- posture management[10]
- prevention and treatment of respiratory complications
- fatigue management
- pain relief
- maintaining functional independence
- optimizing quality of life[4]
- educating and supporting carers.[18]

With disease progression, the optimal setting of physiotherapy provision changes. Ease of access to services, treatment goals, and available resources determine where care should be delivered. The patient's intervention path often leads from review and treatment by specialist ALS multidisciplinary teams in outpatient departments, to treatments provided at home by community services, to therapy provision in a hospice setting. Care should be taken not to overwhelm people affected by ALS with services. Early identification and involvement of local agencies is essential for continuity of care.[5,11]

The UK Motor Neurone Disease Association (MNDA) has provided online care standards, which can be used to monitor that the needs of individuals affected by ALS are being met.[6]

Muscle weakness and exercise in ALS

Muscle weakness is the main presenting symptom in ALS.[1,2] Reduced activity levels due to this weakness result in atrophy, muscle imbalance, and functional difficulties as well as cardiovascular deconditioning.[19] Muscle imbalance leads to shortening of connective tissue and muscle with resultant contractures and discomfort.[20]

The effectiveness of exercise provision for strengthening of weakened muscles has been established in non-progressive neurological conditions such as stroke.[21] Exercise regimes are provided to reduce impairment and improve functional independence. The benefits of exercise in both health and disease are well recognized, including increased efficiency of muscle activation with better aerobic capacity and greater endurance, reduced prevalence of cardiovascular disease, and psychological benefits resulting in an increased feeling of well-being.[21] Additional benefits of training for individuals with neurological disease may include a reduction in spasticity and a reduction of pain from musculoskeletal origin.[16] However, the efficacy of exercise in reducing impairment for individuals with ALS has not been equivocally established.[19]

Exercising neurologically weakened muscles, particularly in progressive neuromuscular disease, should be approached with care, due to the fear that muscles that are already working close to their maximum are more susceptible to overuse weakness and injury.[22] This finding has been investigated in a variety of animal studies. Studies showed that in the case of moderate exercise this fear was unfounded, and exercise actually led to improvements in function and even prolongation of life.[23,24] If, however, the training was delivered at high intensity the findings were not uniform, with some studies reporting an improved aerobic capacity[25] and others a quicker deterioration of physical condition and hastened death, especially in male animals.[26] When the moderate and high-intensity treatments were directly compared in slowly progressive neuromuscular disease, moderate training yielded greater benefit with significant improvements in strength after 12 weeks of training.[27]

In humans with ALS, findings are not that uniform. Some studies found no significant improvement due to training,[28] while others found that moderate strengthening and aerobic exercise[8,16] increase muscle strength, cardiovascular fitness, and endurance.[15]

Recent animal studies have investigated the neuroprotective abilities of exercise in ALS. In line with findings in other diseases, a study in a mouse model has found positive effects of exercise when given in addition to an insulin-like growth factor, resulting in improved motor function and increased survival.[23]

Combination of these findings from animal and human studies has led to the current opinion that exercises appear to be of some benefit for individuals with ALS but that high-intensity exercises should be avoided. The exercise programme should be tailored to the specific needs of each individual and comprise stretching and flexibility exercises, specific moderate strengthening exercises of weakened muscles, and a moderate aerobic fitness regime.

Tone management

ALS is an unusual neurological condition in which patients may present with increased or decreased muscle tone due to the progressive loss of both UMNs and LMNs.[29] LMN damage in ALS is due to loss of anterior horn cells causing hyporeflexia and muscle atrophy, the main cause for the observed muscle weakness.[29] Some individuals will also develop spasticity at some point during the course of the disease as a result of UMN damage.[30]

Spasticity

There is limited evidence to inform the management of spasticity in ALS.[30] In a Cochrane Review of the management of spasticity in ALS,[30] only one study investigating the effectiveness of moderate exercise fulfilled the criteria for inclusion.[16] This study found that 3 months of active ROM exercises significantly reduced spasticity in ALS, as has been demonstrated in other neurological conditions.[31] However, this effect was not maintained at 6 months.

The medical management of spasticity consists predominantly of the use of centrally acting antispasmodic agents (see Chapter 9). The evidence for the use of these medications in ALS is very limited, and treatment is extrapolated from treatment of spasticity in other neurological conditions.[32] These systemic agents, although efficacious in reducing spasticity, have a known side-effect of causing muscle weakness and therefore need to be administered with care.

To avoid systemic side-effects from centrally acting antispasmodic medications, focal spasticity has been successfully treated in recent years with botulinum toxin. It may be useful for treating isolated spastic muscles which may otherwise shorten and lead to joint contractures.[33] In clinically wasted muscles, caution needs to be exercised as it can lead to further weakness.

The physiotherapeutic management of spasticity also consists of positioning and posture management, splinting, exercise, use of orthotics, and maintenance of muscle length and ROM.[34]

Lower motor neuron signs

Features of LMN dysfunction include: muscle weakness, muscle atrophy, fasciculations, hypotonia, and cramps. The extremities are usually the first areas to be affected by LMN loss, and maintenance of normal joint alignment and ROM might require the provision of orthotic devices.[34]

Orthotics, splinting, and collars

In the upper limbs, weakness may cause joint hypermobility and secondary subluxation, particularly at the shoulder girdle.[35] Severe proximal weakness

around the shoulder girdle can result in flail arms. Shoulder slings, collars and cuffs, strapping techniques, and other supports are available to help to support the shoulder and prevent traction injuries.[34] The effectiveness of these devices needs to be assessed individually, based on fit and support. Muscle weakness of wrist and finger muscles leads to muscle imbalance and altered hand posture, most typically evident as a flat hand with limited thumb opposition (Fig. 12.1). Splints can support the medial and longitudinal arches of the metacarpal bones in order to assist function and grip strength by improving thumb opposition.[1] This support can either be provided by off-the-shelf neoprene thumb splints (Fig. 12.2) or can be fabricated with Softcast® by an experienced physiotherapist (Fig. 12.3).

Orthotics may assist in maintaining mobility for people suffering from foot drop due to weakness of ankle dorsiflexors. An ankle–foot orthosis (AFO), either light-weight off-the-shelf or custom made, is commonly prescribed to help correct this problem. It can hold the ankle in a neutral position when mobilizing and prevent the person from tripping during gait.[1,10]

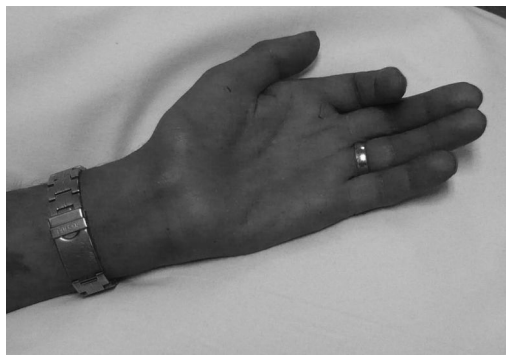

Fig. 12.1 Hand with muscle wasting.

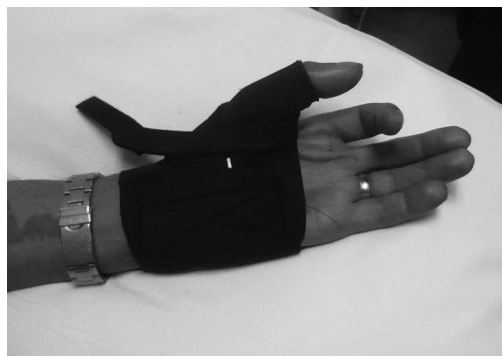

Fig. 12.2 Neoprene splint to correct thumb alignment.

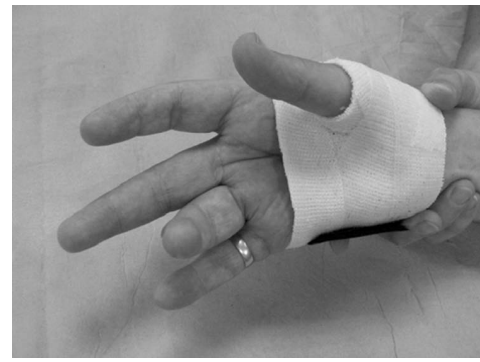

Fig. 12.3 Soft-cast splint to correct thumb alignment.

Progressive weakness can also affect the neck muscles, and this can be a challenge to manage. This becomes most evident after prolonged periods of being upright, resulting most commonly in an anterior head drop due to an inability to hold the head up against gravity. Different collars can be useful to maintain head position and prevent muscle fatigue. A simple soft collar can be sufficient, but pressure on the pharynx can increase the shortness of breath caused by inspiratory muscle weakness and can restrict swallowing efficiency. Liaison with the speech and language therapist is required to determine whether the person with ALS is best advised to eat with or without the collar in place. The two most frequently used collars are the Headmaster Collar™ and the Salts MND collar (Fig. 12.4). The UK MNDA also provides information on a wide variety of collars (<http://www.mndassosciation.org>) but for more complex lateral instabilities, collars that have been custom made by an orthotic department may be more appropriate. Collars can be used for the largest part of the waking day or for shorter periods when excessive neck movement is experienced, for example when travelling in a car.[35]

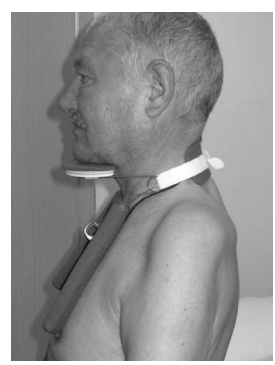

Fig. 12.4 Example of a collar.

Mobility

The maintenance of independent mobility is a marker of quality of life and perceived well-being.[35,36] As mobility becomes more difficult and the risk of falls increases, sticks, crutches, frames, and rollators can aid the individual and maintain safety.[1] There is no definitive timepoint in the progression of the disease when increased support or aids are required. This decision has to be a clinical one in collaboration with the patient. Early consideration and discussion of walking aids will help with acceptance of these when provision becomes necessary.[7] Environmental adaptations might be needed to enable independence at home, for example rails, ramps, or a stair lift, and the involvement of an occupational therapist is helpful.

Transfers between surfaces can become difficult and the carer needs to be educated with regards to manual handling techniques.[18] Transfers can be aided by the use of sliding boards; if more assistance is needed, a hoist may be indicated.[35] There are a variety of hoists available and an occupational therapist will be able to advise on the most appropriate type (see Chapter 13). Standing hoists may be useful, and they have the added benefit of standing a patient.[37] Regular review of transfer equipment is essential.

Many individuals affected by ALS require a wheelchair. The provision of a wheelchair can be perceived as a negative milestone, thus sensitivity needs to be exercised when broaching the subject. Good posture influences pressure management, respiratory function, saliva management, and the general feeling of well-being.[35] Wheelchair users report an increased ability to interact in the community[17] and wheelchair use can aid energy conservation.[1,38] Wheelchairs can be used intermittently for community access or as the main seating and positioning option during the day. Because of the sometimes rapid progression of ALS, frequent reassessments will aid the provision of the most appropriate seating option.[1] The chair can vary from a lightweight self-propelling chair to, in the later stages of the disease, an attendant-propelled, reclining chair with a head rest, pressure-relieving cushion, lateral trunk supports, and arm rests. Electric wheelchairs with various environmental controls are also available and provide users with greater independence and an improved sense of well-being.[17]

Respiratory management

Most people with ALS will experience weakness of the respiratory muscles at some stage during disease progression.[39] To compensate for muscle weakness, the breathing pattern is altered and accessory muscles are used to maintain oxygenation levels.[40] During the night, when the respiratory drive is lower, the

changed breathing patterns are more pronounced. This leads to retention of carbon dioxide resulting in morning headaches—a sign of type 2 respiratory failure.[1] NIPPV is an adjunct used to assist ventilation, relieving shortness of breath, improving the quality of life, and possibly leading to prolongation of life.[41,42] Respiratory failure is the major cause of morbidity and mortality in people with ALS. Guidelines for respiratory function testing encourage an initial assessment with 3-monthly reviews to establish impairments, the rate of progression, and the patient's needs and wishes.[43]

Patients often report the feeling of choking on their secretions or saliva due to an impaired swallowing mechanism.[44] Aspiration risk can be minimized by close collaboration with a speech and language therapist.[35] The cough is an important defence mechanism, and an ineffective cough predisposes to recurrent chest infections. Weak respiratory muscles and poor glottis function result in ineffective clearance of secretions due to a reduced ability to create sufficient pressure prior to, and sufficient force during, a cough.[44] Chest infections are the leading cause of hospital admissions in ALS patients.[45]

It is important to reassure the patient and their carers and have strategies to assist with these episodes. Strategies may include teaching an active cycle of breathing, manually assisted cough, use of portable suction, and mechanical insufflator–exsufflator (MI-E) techniques. The UK MNDA recommends a Just In Case Kit (JIC Kit), which provides advice sheets for doctors, nurses, and carers and suggests a range of medications that can be kept in the box to alleviate symptoms.

A weak cough can be optimized and made more effective by using an assisted cough manoeuvre.[41] Contraindications to this manoeuvre are a paralytic ileus, internal abdominal damage, a bleeding gastric ulcer, and rib fractures.[46] Peak expiratory flow rate is a useful simple tool that can aid with the decision of when to instigate cough assist techniques. The strength of an individual's cough can be measured using a peak flow meter, with the patient forcibly coughing into a face mask attached to the peak flow meter. Cough effectiveness is suboptimal when peak cough flow (PCF) is <270 L/min (requiring an assisted cough; see Table 12.1).[47]

Self-assisted cough

The patient uses his or her upper limbs to assist or replace the weak abdominal muscles. One forearm is placed flat against the stomach below the ribs, and the other hand is crossed over it. As the individual initiates the cough he or she presses with the forearm in and up, to push the diaphragm up and create the necessary pressures for an effective cough. If the upper limbs are not strong enough for this manoeuvre a carer-assisted cough is required.

Table 12.1 Guidelines for assessing assistance required for an effective cough

Peak cough flow	Assistance required for cough
>270 L/min	Assisted cough may only be required during chest infections
>245 L/min	Stable patients may be able to clear secretions effectively with manual assisted cough
<245 but >160 L/min	Add breath stacking to manual assisted cough
<160 L/min	Mechanical insufflator–exsufflator may be required

Carer-assisted cough while sitting

This follows the same principles as self-assisted cough. The carer positions one forearm on the stomach, just underneath the ribs, and the other behind to create counter pressure. It can be helpful to lock the fingers on the opposite side. The flat aspect of the forearm is used and as the cough is initiated the carer pushes in and up to assist the cough. Timing between the initiating of the cough by the patient and the pressure of the carer is crucial to achieve an effective cough (Fig. 12.5).

Carer-assisted cough while supine

This uses the same principles as those for carer-assisted cough while sitting. The carer now uses their forearm to mimic the action of the abdominal muscles in one of two ways, as demonstrated in Fig. 12.6 using both forearms to push up and in or by placing both hands under the ribs and pushing up and in when the cough is initiated. The same procedure can be performed with two therapists (Figs 12.7 and 12.8).

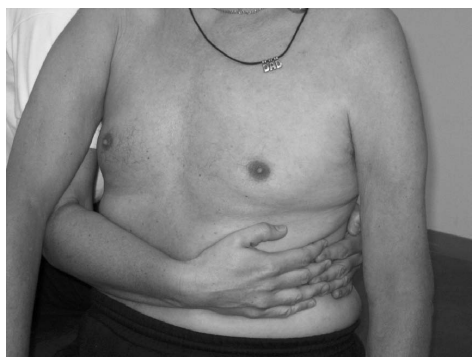

Fig. 12.5 Carer-assisted cough while sitting.

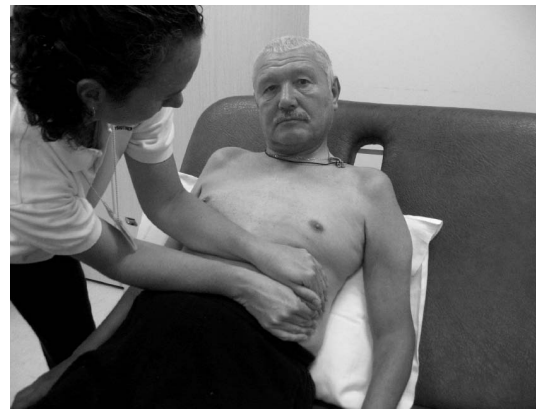

Fig. 12.6 Carer-assisted cough while supine.

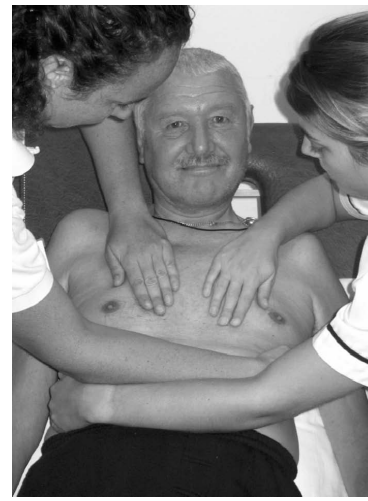

Fig. 12.7 Assisted cough with two therapists.

Physiotherapeutic intervention in the case of respiratory failure due to an opportunist infection should be aimed at maximizing ventilation and aiding the removal of secretions; for example breath stacking, active breathing cycle, and forced expiratory techniques.[48] However, if muscle weakness has advanced and an effective cough or huff is not achievable, an assisted cough will be more appropriate to aid the removal of secretions.[38] Intermittent positive pressure breathing (IPPB) can be used to help prevent atelectasis and achieve a maximal inspiration when the primary problem is inspiratory muscle weakness. An

Fig. 12.8 Assisted cough with two therapists.

assisted cough can also be used in conjunction with IPPB. Modified postural drainage can be used for the clearance of secretions,[35] but toleration of the positions can be difficult[38] and the intervention often proves to be too tiring for patients.[49] Suction to clear airway secretions is invasive and distressing and can increase shortness of breath, but is sometimes required in an acute exacerbation. There is no evidence to show a significant effect on lung function following respiratory muscle training in ALS.[50] Positioning can optimize ventilation–perfusion matching during a chest infection and a reclined or tilted position in a wheelchair can reduce shortness of breath by changing the effect of gravity on the diaphragm and abdominal mass.[35]

If the manually assisted cough is not effective or PCF is below 160 L/min, a MI-E (e.g. CoughAssist™, Emerson, Cambridge, MA, USA) can potentially aid the clearance of secretions from the central airway.[45,51] Positive pressure is used to achieve maximal lung inflation followed by a sudden negative pressure to the upper airways. This procedure mimics a cough by producing more effective PCFs and assisting with the clearance of secretions.[45,49] To achieve adequate pressure build-up in the production of a cough some bulbar function needs to be preserved.[45]

Pain management

Pain is a frequent symptom in ALS with a negative impact on functional status.[52] The pathological process of ALS does not cause pain, but in the later stages of the disease progressive muscle wasting and reduced mobility can cause pain due

to adverse stresses on the musculoskeletal system by poor alignment of joints and the inability to regularly change position.[7,52] Muscle cramps and spasms might also precipitate discomfort.

Pain management is complex and may require a multidisciplinary team approach (see Chapter 9).[52] Physiotherapeutic management is aimed at the prevention of malalignment and contracture formation, and this can be achieved by providing stretching exercises for the patient and educating the carer to perform regular passive movements. Various methods of pain relief including transcutaneous electrical nerve stimulation (TENS), hydrotherapy, acupuncture, local heat, ice, and complementary therapies such as massage have been reported anecdotally to offer relief. Shoulder supports may help reduce pain in flail arm and neck supports may help reduce the pain associated with head drop. Advice on joint support, provision of the correct wheelchair, mobile arm supports, and hospital beds may help to prevent complications.[1,35]

Conclusion

Physiotherapeutic intervention encompasses a variety of aspects and is aimed at prevention of complications as well as symptomatic relief.[1] The treatment plan needs to be flexible in considering fatigue levels to allow activities of daily living to take precedence over exercises provided by the therapist. The patient must be involved and actively participate in all decision-making processes.[7,12] Well-coordinated multidisciplinary care can enhance the health of the individual, increase lifespan, and improve quality of life.[1,5,11,39] Although research into the care of the patient with ALS has increased over recent years, further research is needed to consolidate our knowledge about the interventions we can offer and support we can provide.

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Chapter 13

Multidisciplinary care: occupational therapy

Chris Kingsnorth and Sarah Lavender

Summary

Occupational therapeutic interventions in a patient with ALS cover a broad range of the person's life—for example activities of daily living, such as self-care, and work and leisure activities—throughout the changing demands put on the patient and their carers by the rapid progress of the disease. This chapter aims to give an overview of the domains of concern, and ideas and examples of how occupational therapy can help individual patients function in the activities that are important to them. Topics covered include environment, mobility (including wheelchairs), transfers, hand function, posture, social communication, and suggestions for some of the assistive technology that is available to assist patients and their carers.

Case history

John was 47 years old when he was diagnosed with ALS. He was first assessed when he had reduced finger dexterity, making domestic chores and activities more difficult. He was regularly assessed throughout the course of his disease and the occupational therapist later provided him with a manual wheelchair and a riser recliner armchair when his walking became difficult. The therapist also provided training to use a bathlift and ordered ramps as soon as he was psychologically ready to accept them. He eventually required an electric wheelchair and an overhead gantry hoist for transfers in the house which the therapist demonstrated for his wife to use when the professional carers were not present. The occupational therapist continued to provide support for John's wife by providing one-way straws, allowing him to continue to drink a little even when he was in bed and very weak, until John died at home. Advice on moving and handling him in bed.

Introduction

Occupational therapy (OT) intervention for a person with ALS involves consideration of the domains of occupational performance as described by Rennie

and Thornton.[1] Occupational performance is 'an individual's ability to perform activities of daily living, influenced by their environments, within a satisfactory time frame and at an age appropriate level, to fulfil life roles'. Activities of daily living are 'the range of everyday tasks an individual needs to perform to fulfil their occupational roles in self-care, work and leisure'. In the OT model, the performance components comprise the skills necessary for personal activities. Disruption of occupational performance may be the first indication to an individual that there is something wrong.[2]

In the management of ALS, aspects of the domain of concern have an impact on the person's ability to fulfil desired roles. It is important that the occupational therapist recognizes the uniqueness of the disease experience to each person and incorporates this into their assessment of that person's abilities and needs.[3] However, there are some common strategies: some of the equipment which enables an individual to achieve a desired occupational role is expensive, and it can be argued that due to the short duration of the condition, rapid changes, and the small number of patients, a selected collection of equipment could be recycled economically. An equipment system such as that of the Irish Motor Neurone Disease Association[4] is effective.

Domains of concern

Age

Age at onset affects the extent of the patient's life roles that are influenced by impairments. For example, patients under 45 years referred to OT may have issues such as paid employment, parenting, and outdoor activities. They are more likely to have a partner who is able to assist them in their roles and in self-care. These younger patients may not accept their impairments and seek to 'keep ahead' of the disease by strategies, techniques, and equipment to compensate for the changes. This makes for a challenging and satisfying partnership with the occupational therapist. It can also be frustrating when the required equipment is unavailable, expensive, or not yet invented! Younger patients may also have a greater tolerance of assistive technology. Older patients are often more accepting of their fate, and because of this, or a reluctance to place demands, require encouragement to accept assistance. The older person is more likely to have an aged carer who may themselves have a disability and their needs are greater. These patients, however, do not usually have the same range of life roles.

Environment

Intervention with OT will be influenced by both the social and the physical environment. Where the patient is socially isolated there is greater need for

access to community services, immediate response to equipment needs, and the possibility of being unable to remain at home in the terminal stages. The cultural/social milieu may be one in which the 'sick' role requires that the person with ALS be assisted in all aspects of daily living even though independence could be maintained. In this situation, intervention will be aimed towards assisting the carer to carry out their role, for example training the carer in practical skills such as manual handling and the use of assistive equipment.[5,6]

The physical environment requires regular evaluation as abilities and needs change. It is difficult to plan ahead while being sensitive to the patient's and carer's need to be positive and hopeful. It is also important to assess the patient's and family's ability to cope with these changes and challenges,[7] as the introduction of equipment before the patient is prepared may lead to rejection because it is 'symbolic of disability and reminds him of his deteriorating condition'.[3] Deterioration in the ability to walk suggests future need for a wheelchair, but it may also be necessary to suggest measuring for ramps or stairlift when the patient is still walking. The ability to remain independently mobile at home, where lower limb weakness is the major symptom, is affected by access (steps/stairs), width of doorways, and circulation space. When the person lives alone there are the added issues of ability to open doors and use a key.

The problem of a suitable bed may be as simple as providing blocks to raise the existing bed to a height that it is easier to transfer from, or to be level with the wheelchair, or a board between the mattress and base for firmer support. The more complex needs of adjustable height and postural control can be met with a hospital bed or by the patient purchasing their own electronically controlled bed. It is important that this has height and both head and leg adjustment and minimum underclearance of 150 mm to accommodate a bed table or mobile hoist. A knee break reduces the risk of pressure areas due to skin shear. There are control options to accommodate profound loss of motor function. In one case where a patient's mother was the primary carer she was exhausted through having to adjust her son's posture throughout the night. The purchase of an electric bed meant he could adjust his posture independently and his mother was then better able to cope with the demands of the day. Community nurses and carers were also more able to adhere to occupational health and safety guidelines in providing ongoing assistance.

Time

The progressive nature of ALS means that needs change rapidly. Complex systems of funding and purchasing equipment mean that equipment or a service is

often not provided in a timely fashion. It is often necessary to develop an equipment resource, such as electric wheelchairs, which can fill a need until other sources are available. Voluntary support groups, such as the UK MNDA, can help.[8]

Performance components

Those performance components most commonly related to OT intervention are motor and social. Complete loss of motor ability is linked with all aspects of daily living. Sophisticated environmental controls allow a patient to continue social interaction and make changes in his or her environment with the subtlest input.

Motor

Muscle strength and active range of movement

These components are associated with wasting and dysfunctional joint positions. Commercially available or custom-made thermoplastic resting splints for the wrist and hands maintain optimum muscle length and functional joint positions. These are usually worn at night or for 2 h during the day, for example when watching television. ALS patients have a low tolerance for static splints with poor compliance for wearing them. A compromise may be to wear them on alternate hands on alternate nights.

Shoulder supports alleviate joint stress during ambulation where there is loss of strength in antigravity muscles. They have also been useful in relieving pain.[9]

Compensatory devices which allow optimum use of residual muscle power may be needed. This may be to provide distal support in the upper limb, i.e. the wrist and fingers, thus allowing effort to be channelled into controlling the shoulder, for example when eating or using a keyboard. A finger splint (oval 8) can be used on individual finger joints to enable finger function to be maintained. Toe-ups can also be provided to prevent the toes from curling up when shoes are worn. Alternatively, the shoulder and elbow may be assisted with a gravity-eliminating device allowing the use of residual wrist and finger function. These perform a dual role of enhancing independence while maintaining joint ROM. Another simple example is providing raised seating for the toilet and sitting room to allow larger proximal muscles to be utilized for standing up.

Loss of endurance

It may be necessary for the patient to make choices about which activities they continue to expend energy on and which they will accept help with. It has been

noted that 'there is no correlation between independence in Activities of Daily Living (ADL) and quality of life'.[10] Patients may be counselled to accept help with self-care, allowing more time and energy for quality time with their family and leisure or work, in this way 'lowering their expectations regarding their physical disabilities and by focusing on life areas that are independent of physical capabilities'.[10] Education and advice on energy conservation and work simplification principles can be given to the patient and carers and applied early in the disease, for example sitting for tasks such as showering and washing dishes.

Hand function

Specific examples will be given in the section on Activities of daily living, but there are some general principles. In the early stages when grip strength and precision are affected by loss of intrinsic function, large handles and lighter tools may be useful. Devices for holding items allowing a less precise grip or movement may be useful, such as a palmar cuff to hold cutlery or a pen holder for writing. There are more complex assistive devices such as the Neater arm support which allow movement with minimal muscle power in the later stages and can be used for eating and other activities.

Mobility

Consideration needs to be given to both patient and carer when deciding on a wheelchair. Initially a manual wheelchair may suffice, but the long waiting period for an electric wheelchair means forward planning is required. Change in status from walking to wheelchair is devastating to some, and preparing patients for this requires sensitive handling by all. Where patients are able to walk short distances at home, a successful interim solution is a mobility scooter. They are more acceptable as they are considered less 'disabled-looking' than a wheelchair and soften the blow of not being able to continue driving a car.

Manual wheelchairs need to be lightweight, with wheels that are easily removed so it is easier for a carer to lift. Armrests that are removable and height-adjustable reduce the weight while allowing for transition from standing transfer to side transfer. Friction grip on the push-rims allows longer-term independent wheeling as grip strength deteriorates. Pressure points must be addressed with cushioning, together with considerations such as cushion covers. For example, shiny Lycra cushion covers facilitate slide-board transfers and removal of clothing for toileting.

If an electric wheelchair is inappropriate or unavailable, an attendant-controlled manual chair is useful. When trunk and head control deteriorate,

these provide the option of 'tilt in space'. This is preferable to reclining from a postural viewpoint as it allows a change in seating pressure while supporting the trunk and head and maintaining a stable pelvis position. Subtle changes of tilt in space can negate the need for external head fixation such as neck collars. Consultation with the speech and language therapist regarding head position is vital where swallowing is compromised. When the patient is unable to self-propel, a pushchair is easier for carers.

Most patients with lower limb involvement require an electric wheelchair. There are many options. Choice will be governed by availability, cost, portability, postural requirements, and additional attachments, for example a lightweight folding demountable chair or a fixed chair with customized seating, electric tilt and recline, and additional head support. The type of controller will depend on motor ability and range from a standard hand control, hand control with arm support, to head control, knee, or foot control, with different switching options. The optimum chair allows the patient to have some control over their life, independent mobility, ability to change position, and, with current models, to access other areas such as communication and environmental control equipment.

Posture

While patients are still ambulatory, a riser recliner electric armchair enables them to independently adjust their sitting position to assist breathing and with transfers. Maintenance of posture in a wheelchair has implications for the control of pain, access to equipment, and pressure care. Pelvic stability is central to a functional sitting posture. As muscle strength deteriorates it is vital that cushioning supports the pelvis in a stable position. Lateral trunk support may be required through customized cushioning or choice of wheelchair. In the later stages the position of the trunk most conducive to breathing or to allow head control is with the body tilted back, as with tilt in space. This makes it difficult to see a communication device or computer screen, so modifications are required to position these items and posture needs to be changed for eating. A simple, adjustable neck brace has been used as a short-term measure to support the head forward for eating or while travelling in wheelchair transport. A wheelchair with electrically controlled postural adjustment increases the ability to independently adjust posture and reduces the anxiety associated with breathing difficulties.

Bed mobility

In the early stages, movement in bed may be assisted by using simple measures such as creating a firm base with the use of a bed board, a simple bed rail which

slides under the mattress, or by wearing satin nightwear. As abilities decrease, a suitable pressure mattress may preclude the need to move at night, when there is manual assistance with posture during the day. Other options include an electronic bed (see section on Environment): an electronic double bed is available with separate mattresses to allow couples to sleep together and maintain their personal relationship. Some find it more comfortable to sleep in a riser–recliner arm chair.

Transfers

The critical factor in facilitating independent transfer is the height of surfaces. When there is lower limb weakness, seating needs to be raised (see section Performance components, Motor). When a wheelchair is used, positioning of items to allow safe transfers is important. For example, the bed may be moved to allow the chair to be positioned appropriately. Chairs and beds which have been raised to assist standing transfers may need to be lowered to the height of the wheelchair if side or slide-board transfers are introduced. Slide-mats and transfer belts are useful for assisted slide-board transfers. Standing and side transfers to a car may be assisted by a handybar or a wedge cushion on the car seat to level it out, with a Lycra cover or a fabric turntable cushion to assist slide and swivel. Where dependent standing transfers are used a swivel board or turntable may assist the carer. As complete dependence in transfers occurs, a hoist, either electric or hydraulic, may mean the difference between staying at home or being cared for in an institution. There are many hoists and slings; therefore careful choice must reflect the requirements of patient, carer, and environment. When the patient can bear weight with assisted support, standing hoists have been used. They allow easy access for all hygiene activities and the patient maintains an upright posture. As the disease progresses a more dependent hoist is required, whether mobile, overhead gantry, or ceiling hoist. This transition may be difficult to accept by the patient or the carer, but continuing to use a standing hoist puts both at risk of injury.

Driving

The patient must be advised with regard to driving ability and be given an assessment if necessary. Where the disease process has been slowly affecting lower limbs only, hand controls may allow continued driving; aids such as hand grips for the steering wheel, for example a spinner knob, may help.

Social

Holidays are an important part of a family's life and the occupational therapist can advise on what equipment to take and provide information on accessible

accommodation. For example, a woman booked a cruise and the occupational therapist liaised with the cruise company to provide an electronic bed and mobile shower chair.

A two-seater riser–recliner armchair may enable a patient to maintain physical contact with loved ones whether reading stories to grandchildren or having a 'cuddle'. Skills to maintain communication and social contacts are also areas on which the therapist can advise.

Writing

Writing will involve assessment of hand function as described in the section Motor function.

Using a keyboard or computer

There are many alternatives to a standard computer keyboard to assist with typing, for example an on-screen keyboard, key protector, voice-activated screen, and infrared sensors. The mouse can be customized with a single-click ball device or by an on-screen scanner with the controller spot positioned individually for each patient. An over-chair table with an adjustable angle can minimize the physical activity required to access the patient's laptop, Kindle, or iPad with word prediction, single switch access, and speech apps. There is the need to coordinate the assessment with the speech and language therapist (see Chapter 14).

Telephone, text, e-mail

Modifications can be made to hold a standard telephone, and modified telephones are available, for example with large pressure-sensitive keys. There are hands-free telephones which allow communication to continue by telephone. Communication by text or e-mail and an answer-phone is helpful as speech deteriorates, as it allows a message to be taken without embarrassment or confusion for the caller or the patient.

Activities of daily living

Self-care

There has been a great deal written on solutions for problems with self-care. This section will be limited to ideas to solve individual problems. Self-care deficits vary according to the type and onset of ALS. Those with bulbar involvement remain ambulatory for longer; initial motor problems are usually in the upper limbs. This may result in difficulty with eating, toileting, dressing, and using a keyboard or communication device. Those with involvement of the lower limbs have problems with transfers and mobility. By the time their upper limbs are

sufficiently involved to affect eating and using a keyboard they require assistance in all areas of self-care. The ideas that follow include those for the primary upper limb involvement first.

Personal hygiene: toileting

Where grip strength is diminished, moist towelettes instead of toilet paper can help. Simplified clothing without fastenings, and loops on the waist or the use of braces assist with clothing. More expensive options include a bidet with sensor which both washes and dries after toileting.[11]

The most commonly used equipment includes an over-toilet frame (to raise the seat), hand rails (fixed or suction), and later a wheeled shower/commode chair. The latter can be wheeled by patients or the carer over the toilet and then into the shower, limiting the transfers required. It can be used with standing, slide-board (side) transfers, or hoist transfers.

Bathing

Liquid soap in a pump pack, a thermostat for water, a tap-turner or lever taps, and a soap mitt may be useful. There are shower and bath seats, fixed or adjustable, to allow the patient to be raised and lowered, and these contribute to safety and energy conservation. The wheeled commode/shower chair is invaluable where the shower is accessible or there is drainage in the bathroom floor. A tilt and recline shower commode, with a hoist, allows a patient who is dependent in posture and head control to shower while being safely supported. Electric tilt recline shower commodes are also available.

Dressing

Simplified clothing, and easy fittings with minimal fastenings are advisable. Loops on trousers and socks and shoes with hook and loop fastenings or slip-ons aid dressing. Some women find wearing a skirt proves easier for toileting.

Feeding

When hand function deteriorates large-handled cutlery and a splayed fork may enable independent eating to continue. Other equipment includes plate surrounds, thermal and two-handled mugs, and a non-slip mat. These can be easily taken out to enable patients to maintain their social contacts. The Neater Eater (<http://www.neater.co.uk/eating-aid>) has been used successfully by some. It is clamped to the table and there are individual models and adjustments which allow the user to bring food to the mouth with little physical input. Some are reluctant to try it because of the 'robotic' appearance, perhaps reinforcing their loss of function, while others are pleased to maintain their ability to share mealtimes with their family.

Work

The length of time for which the person with ALS can remain working is determined by the nature of the work, the standing of the employee, and willingness of the employer to adapt to the person's changing ability. Where the work requires keyboard skills and is sedentary, adaptations are easier. For example, one patient worked as a merchant banker for many months with an electric wheelchair for independent mobility. He was delivered to the building goods entrance by wheelchair taxi so that he could access his office through a door using a modified key, as the main doors were too heavy. Circulation space, desk orientation, and height were altered for easier access. Initially he transferred to a suitable office chair and later worked from his wheelchair. Provision was made for an Ergo Rest to assist with keyboard use, and typing splints were made.

A voice-activated computer and head-piece for the phone enabled a young man using an electric wheelchair to continue working with an understanding employer. His care package was adjusted to facilitate working commitments.

Often the OT intervention requires partnership with the patient to problem-solve, identify, and adapt to the rapid changes and individual needs. Optimal service is dependent on recognition by funding bodies of their unique need for services and equipment in a flexible and timely manner.

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Chapter 14

Multidisciplinary care: speech and language therapy

Amanda Scott and Maryanne McPhee

Summary

The speech and language therapist has an important role in the multidisciplinary assessment and care of a person with ALS, particularly with regard to the management of changes to speech and communication. Within a holistic person-centred approach information is focused on the individual needs of the person and their family and encourages choice and autonomy. This will include assessment of both speech and swallowing (see Chapter 7). Speech impairment, or dysarthria, is a cardinal feature in ALS. The speech and language therapist has a vital role in the assessment and management of this problem. The progressive nature of ALS and the wider implications of speech impairment necessitate adaptation to the interventions. Augmentative and alternative communication (AAC) is an important part of clinical intervention. The successful use of AAC is derived from the match between people's abilities, preferences, and needs and technologies and services available to support them and their families. The provision and support of AAC systems requires specialist knowledge and skills. This chapter provides an overview of the motor speech changes in ALS, management options, and a conceptual framework for introducing for AAC.

Case history

Mary was a 58-year-old woman with a supportive husband and three adult daughters. She was working as a school teacher and noticed very early voice changes. Initially she was sent to an ear, nose, and throat specialist and was found to have normal vocal function. Her GP then thought that her voice changes may be due to 'little strokes'. He then referred her to a neurologist, and a diagnosis of probable ALS was made. This diagnosis was confirmed within 6 months. Her speech deteriorated rapidly and she ceased working.

At her initial assessment her speech was characterized by hypernasality, imprecise articulation, and reduced rate. She was spontaneously slowing her rate of speech and over-articulating to enhance intelligibility. At this stage she was approximately 80% intelligible to familiar listeners. To supplement dysarthric speech she and her adult daughter, who was IT savvy, looked at options on the Internet.

They chose an iPad to support her declining speech function. She was using a type and speak program which had limited ability to store phrases and had no word prediction function. This was reviewed to ascertain how this was working. Mary and her husband reported that they were finding it slow. The speech and language therapist (SLT) demonstrated other app options that could speed up the process. At this stage the SLT observed early signs of upper limb weakness. A word prediction app was introduced straight away which resulted in an immediate increase in the speed and efficiency of communication. This established the family's trust that the SLT had some specialized skills which could supplement their approach and resulted in a positive outcome for all involved.

In between the clinic visits the SLT and the occupational therapist began to investigate switching options for the iPad. At that time only one was available. At a review 3 months later Mary's hand function had deteriorated. Her dysarthria was more marked and she was augmenting her communication with facial expression and gesture. The switching system for the iPad was demonstrated, and both Mary and her husband perceived this as too slow. Other options, such as a laser pointer, eye-gaze systems, and low-tech boards were discussed and the possibility of coming back to try these out was offered.

At the next visit, a joint session with the occupational therapist, Mary's communication goals were discussed. These included: maintaining quality communication with family and friends, easy access to SMS, e-mail, and the Internet. Mary's other options to support iPad use were examined and a range of different switches were trialled. Mary decided to continue using direct access to the iPad even though this was now becoming very slow. Subsequently an ETRAN (eye transfer communication board) and an eye-gaze phrase board were introduced, and later a head mouse and software to access the Internet and e-mails were added.

As the disease progressed Mary became more physically disabled and less able to use the iPad, becoming more reliant on the ETRAN and phrase board. This led to increased responsibility for all communication partners. To support this, the SLT provided ongoing education and support to Mary's family. These methods enabled communication until the end of Mary's life.

Throughout this process the SLT worked in partnership with Mary and her family, allowing Mary's wishes to be the basis of decision-making even when the SLT may have thought other options to be more efficient and effective.

Background information

The ability to communicate is a fundamental human activity, enabling us to share our thoughts covering the full range of life experiences. In ALS, speech may become unintelligible and sometimes language and cognitive functions become impaired. These problems can lead to difficulties interacting with family and loved ones, carers, and health professionals. Those caring for people with ALS have a responsibility to provide the most efficient and effective means of communication possible.

This chapter outlines the role of the SLT in ALS, and focuses on aspects of speech impairment and communication as the disease progresses. In early bulbar impairment, focus on speech production for short periods may help, but alternative means of communication are more important later in the disease.

Because communication always includes other people, optimizing communication should include family, friends, health professionals, and carers.

The ability to communicate declines gradually, which allows time for adjustment to loss and the impact this has on relationships. Early recognition of declining intelligibility of speech and communication makes it easier for the person to convey decisions regarding the disease and the changes it imposes. It also provides the opportunity for timely strategies to facilitate communication.

Dysarthria

The motor speech system requires the integrated functioning of the respiratory system, phonation, and articulation, which involves the lips, tongue, soft palate, and pharynx. Impairment of the motor speech system is one of the most important clinical problems in ALS. Dysarthria is the initial or early predominant sign in about 25–30% of people with ALS[1] and over 90% will develop bulbar symptoms.[2]

Over the course of ALS, speech function may range from mild dysarthria, presenting as a slight slurring of speech, to anarthria, or no speech. Dysarthria in ALS is usually a mixed pattern involving both upper motor neurons (UMNs) and lower motor neurons (LMNs). The loss of UMNs leads to spastic-type muscle weakness, and the loss of LMNs causes weak flaccid muscles. The mixed dysarthria of ALS is characterized by reduced rate, range, strength, and accuracy of muscle movement.

Respiration

Weakness of the respiratory muscles frequently occurs with bulbar symptoms. Respiration is the driver of speech. Impaired respiratory support for speech decreases volume and leads to a reduction in the prosodic features of communication, such as stress, rhythm, intonation, and rate. As the number of words produced per breath decreases, speech becomes more effortful. People experience fatigue and difficulty communicating against background noise when volume is decreased. This restricts their ability to contribute to group conversations and to socialize. The reduction in the prosodic features limits the ability to express the subtleties and emotions in speech such as irony or excitement.

Phonation

The larynx is the primary organ for phonation, or voice. The vocal cords, which are located within the larynx, are open during respiration and come together into the midline; they vibrate as the airstream passes through, to produce a

clear, strong voice. Changes in vocal pitch depend on extrinsic muscles of the larynx and the flexibility of the vocal cords. As the larynx raises and tilts forward the vocal cords lengthen to produce higher-pitched sounds. When the larynx is lowered the vocal cords shorten and produce lower-pitched sounds.

Impairment of LMNs results in a soft, weak voice with a breathy quality because the reduced tone of the flaccid vocal cords allows greater air flow. The voice becomes lower pitched and monotonous.[3] UMN impairment causes tightening of the vocal cords which leads to a harsh and strained voice. This harsh vocal quality can give the impression that the person is angry, leading to misinterpretations of communication. People with voice impairment commonly have problems communicating against background noise and experience vocal fatigue, which further restricts conversation.

Articulation

Soft palate and pharyngeal function

Most speech sounds are produced with airflow directed through the oral cavity. This is achieved by closure of the nasopharyngeal opening by elevating the soft palate and contracting the pharynx. However, during production of 'n', 'm', and 'ng' (as at the end of so*ng*) the soft palate is lowered against the elevated back of the tongue to allow air to flow through the nose. This results in the nasal quality of these sounds. Impaired soft palate/pharyngeal closure results in nasalization of all speech sounds and the loss of clarity and intelligibility. Nasal emission reduces oral airflow, and inefficient use of air leads to fading at the end of words and phrases, shorter phrases, and fewer words per breath. This condition is called velopharyngeal incompetence and the resulting hypernasality and nasal emission are common.[3]

The tongue

The tongue is the principal articulator of speech. A range of subtle movements produce the diversity of speech sounds. During the production of the 'k', 'g', and 'ng' the back of the tongue elevates and then releases the airflow. The sounds 't' and 'd' require elevation of the tip of the tongue and a sudden release of the airflow; 's' 'sh', and 'z' are produced using turbulence between the tongue and palate; and oral resonance is altered by the tongue position for the sounds 'l', 'r', and 'n'. To produce 'th' air turbulence is produced between the tongue and the upper teeth.

The generalized tongue weakness typically found in ALS is the major cause of reduced speech intelligibility. Mild tongue impairment is associated with slurring. As tongue function deteriorates, fewer sounds can be accurately produced and eventually speech becomes unintelligible.

The lips

Lip closure immediately before the sudden release of air is required for the production of 'p' and 'b', while lip posture is important for other sounds. Rounding is required to produce 'w' and lip closure for 'm'. During the production of 'f' and 'v' the lower lip rests on the upper teeth to produce turbulence.

Spastic lips are tight and have a retracted appearance, often with the teeth visible. This causes difficulty obtaining lip closure and making subtle movements. Flaccid lips result in weak sound production and drooping at rest. Poor oral closure is associated with drooling, which can cause embarrassment and reluctance to speak. People with ALS have commented on the loss experienced when impaired lip movement makes it impossible to purse the lips to kiss their loved ones. The facial muscles are usually affected when lip impairment is present and result in alteration in sound quality and reduced facial expression.

Assessment of dysarthria

Clinical assessment of dysarthria principally relies on the perceptual judgements of the SLT. The Frenchay Dysarthria Scale is a useful means of standardizing judgements and quantifying progress across a range of functions.[4] This assessment is thorough, but the SLT needs to consider the impact of monitoring decline on the person with ALS and use assessments sparingly. The ALS Functional Rating Scale[5] includes a brief five-point subscale for speech function based entirely on the judgement of the clinician. This may be a non-confrontational means of measuring the progression of speech decline.

Other problems which affect communication

Positioning and general comfort

The effectiveness of communication may depend on physical comfort. Appropriate positioning decreases abnormal tone, reduces the effort required to maintain a sitting position, minimizes reflexive responses and clonus, facilitates access to communication devices, and optimizes respiratory function. Liaison with physiotherapists and occupational therapists about seating, head, neck, and trunk support, and upper limb function is essential.

General comfort also relies on basic needs being met often by others, e.g. drinking, toileting, blowing the patient's nose, etc. A quick and efficient method of communicating attention to these tasks must be established.

Pathological laughing and crying

Pathological laughing and crying, also referred to as emotional lability or pseudobulbar affect, are frequent problems in ALS but are poorly understood.[6]

Pathological laughing and crying are considered to be disturbances of motor affective behaviour, and are probably related to frontal lobe dysfunction. Pathological laughing and crying are usually associated with spastic bulbar impairment. The laughing or crying presents as an unconstrained triggering of an emotional response and can be conceptualized as perseveration of the motor responses for emotion. It is usually appropriate in type but excessive in magnitude, and once initiated the response is difficult to stop.

Clinical experience suggests that the discussion of emotive issues is likely to trigger pathological crying, but in severe cases, pathological laughing or crying can be triggered with mildly emotional topics. Pathological laughing or crying can be an impediment to verbal communication and social activity. Changing the pattern of breathing may assist people to gain some control over their motor affective behaviour. This may be achieved by concentrating on inspiration during uncontrolled laughter and expiration during uncontrolled crying. In mild cases, simple strategies include a reassuring but not excessive response and a change of topic to help break the pattern. Patients have reported that thinking positively has assisted them in breaking the pattern.[6] Various medications have been found to be useful in managing this problem (see Chapter 9).

Impairment of cognition and language

Many people with ALS have changes in cognitive skills and processes and/or behavioural change. There is considerable evidence supporting the existence of cognitive and behavioural dysfunction in ALS, including a spectrum of frontotemporal syndromes and more classically defined dementias[7] (see Chapter 8). The impairments reported range from florid personality changes with severe impairment of frontal lobe executive functions to mild cognitive deficits. Frontal and temporal lobe involvement have been identified post mortem[8–10] and through neuropsychological testing.[9–11] The problems associated with language function are deficits of verbal fluency, spelling, the ability to generate concepts or shift mind sets, and problem-solving. A small number of people with ALS have been found to have aphasia in association with FTD.[10] These people have been identified as having a progressive non-fluent aphasia and have demonstrated difficulties with the comprehension and production of verbs.[12]

Impairments in cognition and language are thought to be under-reported because they may be masked by dysarthria or behavioural abnormalities, leading to poor recognition of the extent of the deficits by communication partners. However, the implications for both verbal communication and use of augmentative

and alternative communication can be profound. Cognitive impairment needs to be considered when communication breaks down. Signs of cognitive impairment may include:

- a deterioration in spelling
- inappropriate responses to questions
- introduction of seemingly unrelated topics in conversation
- poor eye contact, attention directed to peripheral aspects, e.g. fidgeting in a handbag during conversation
- difficulty switching topic in conversations
- loss of organizational skills, e.g. losing things, difficulty managing finances, missing appointments
- continuing to write a message even though the content has been accurately predicted
- an over-formalized, pedantic communication style.

As cognitive and linguistic functions deteriorate the communication partner must assume more responsibility. Inaccurate responses and inability to either write concise messages or to write key words of a verbal message hinder the flow of an interaction. Where these problems occur, the role of the communication partner changes to include interpretation of incomplete messages or to take responsibility for maintaining the topic.

Problems with oropharyngeal secretions

Individuals swallow about 600 ml of saliva per day.[13] This is a largely subconscious activity and problems with oropharyngeal secretions often co-occur with speech difficulties and dysphagia (see Chapter 7). An oral lubricant or a light oil, such as grape-seed oil, can be beneficial for lubricating the oropharyngeal structures and facilitating oral movements during speech.

Problems managing saliva are worse following meals (including enteral feeds) when there is a post-prandial increase in digestive secretions, and towards the end of the day when the person is fatigued. At these times, individuals may be reluctant to engage in communication for fear of spitting or dribbling.

In mild cases increased saliva can be managed by regular, volitional swallowing. Sipping a drink during interactions to swallow pooled saliva or deliberately swallowing before speaking can help. As swallowing problems progress, drooling may become so troublesome that medication is necessary to reduce saliva (see Chapter 9). Medication is often most effective when used in certain situations for short periods, e.g. before a social event.

Intervention

Environmental considerations

Environmental factors include any external influences on the communication function. These can be positive or negative influences.[14] An example of this, with regard to people with dysarthria, is background noise or a group setting. This can result in reduced intelligibility or the person straining to be heard. Simple listener strategies such as turning off the television and radio, shutting doors, or moving to a quieter location make speaking and listening easier. When conversing, seating arrangements should facilitate communication by ensuring people are face to face and that lighting is adequate. Having the light source, such as a lamp or window, behind the person with dysarthric speech should be avoided, as this makes it difficult for the listener to see facial movements and expressions to supplement intelligibility. Both the person with ALS and their family/carers need to be open to environmental adaptation, and this can be challenging, especially with rapid deterioration. Timely introduction of change is important for the person with ALS and their and family.

The role of direct speech therapy in ALS

Speech is much more than the encoded linguistic message. Speech also conveys aspects of personality, such as sense of humour or feelings towards the subject of the conversation. With progressive loss of speech, there is a corresponding loss of the ability to converse. This encompasses loss of control of the environment, loss of identity, change in self-image and self-esteem, and a loss of purpose. These changes have an impact on relationships within the family and broader community. Discussing these changes as the person with ALS is experiencing deterioration in speech can assist both the person with ALS and their family, friends, and carers to make adjustments to reduce the impact on relationships and everyday life.

Degenerative conditions require special consideration when choosing therapeutic interventions. Four out of five people with ALS will require intervention regarding their communication.[3] There is limited research into the effectiveness of intervention for dysarthria in ALS. Several compensatory strategies have been suggested for mild dysarthria.[3] These include slowing the speech rate, substituting articulation manoeuvres such as using alternative words which have fewer syllables or easier phonemes, over-articulation, writing or spelling out topic words, and syllabification of speech.[14,15] Gentle onset speech to reduce respiratory effort, assistance with initiation problems,[16] and reducing vocal tension have been recommended.[17] Rate reduction strategies may improve

intelligibility and the timing of periods of vocal rest may also be important when fatigue is an issue.[16]

Careful consideration of the rate of disease progression in relation to the timing of therapy and the duration of intervention is important. The desire to ameliorate speech problems by setting up therapy programmes, which include repetitive speech and muscle exercises, has the negative effect of providing a means by which the person can monitor their own deterioration. Resistance exercises, oral motility and strengthening activities, isometric exercises, and loudness activities may cause decreased voice quality and a more rapid rate of decline in speech intelligibility.[18] However, all the participants in a study by Murphy[19] expressed a strong desire to use their own speech for as long as possible, so functional speech strategies need to be encouraged.

SLTs also need to be mindful of the needs and abilities of the communication partners. Deciphering dysarthric speech takes concentration and results in listener fatigue. Providing communication partners with strategies to maximize listening effectiveness should be part of the intervention process. Strategies include having quarantined time for communication, use of yes/no questions to establish immediate needs such as during positioning or toileting, and allowing the communication partner to speak for the patient in some situations, such as with strangers. The SLT can support the communication partner by providing a safe environment to express feelings of frustration and grief at the changes in communication that occur in ALS.

Introducing augmentative and alternative communication (AAC)

For most people with ALS, speech deteriorates over time, and with support many are able to make adjustments. Open discussion concerning the loss of speech, the meaning of this to the person, the impact on their relationships, and the changes in role and loss of identity, may assist the person to accept the need for AAC and improve their uptake of a new and different way to communicate.

An interview to establish the person's wants, needs, and motivations should occur. Honest discussion with the person regarding their expectations of AAC and what AAC can realistically provide is essential. Open acknowledgement that an AAC option will never replace speech, nor be as efficient or as easy to use, is also helpful. The goal of the AAC strategy is to provide the person with a means of communicating with their family, friends, health professionals, and carers. Murphy[19] found that the ability to maintain social closeness with another person was seen as the most important purpose of communication—above the

person's desire to indicate wants or needs or pass on information. Fried-Oken et al.[20] also provide evidence that AAC provides people with ALS with a means for participation, engagement, conversation, and message transfer. Strategies to encourage acceptance and use must be tailored to the individual to ensure uptake and effective use.

The authors recommend that therapists are involved from diagnosis to ensure an ongoing relationship which will facilitate the measurement of changes in speaking rate and anticipate communication needs.

Communication, whether via gesture, verbal, written, or using a communication device, conveys information at various levels of complexity and there is a wide range of communication requirements using AAC.

The AAC framework

AAC options need to reflect the wishes and needs of the person and their family and carers. Early and timely referral for AAC assessment and management is critical and should be based on knowledge of the disease, disease phenotype, and rate of progression. For example, patients with bulbar onset should be referred early, around the time of diagnosis, to ensure an ongoing relationship which will facilitate appropriate monitoring and review. This may include the monitoring of changes in the person's speaking rate or changes in their ability to access and effectively use their current AAC system.

The appropriateness and timing of AAC options play a significant role in acceptance. Timely referral enables the person to learn how to use the device before it becomes an absolute necessity. As well as the rate of disease progression, the person's psychological and emotional well-being and cognitive capacity must be considered. Individual coping styles need to be respected. Some people like to plan and find the early introduction reassuring. Others prefer a 'wait and see' approach, and for these people early introduction can be confronting and conflicts with their coping strategies. It may also cause anxiety and stress by indicating they will lose their speech.[19] However, the AAC system must be available when the person is ready.

Assessment for AAC is a multifaceted process and limited practice guidelines exist.[22] Physical disability, linguistic functioning, and cognitive status all need to be considered when determining the type of AAC option to be recommended. Researchers highlight the difference in approach between AAC specialists and general clinicians. Advice and input from an AAC specialist may also be required for complex cases. Working within a neuropalliative rehabilitation model[23] for AAC provision ensures that the team draws on principles of rehabilitation within a palliative framework—so that goals are

realistic and are focused on maintaining independence and maximizing quality of life.

As well as ensuring AAC options are provided in a timely manner, it is essential that they are reliable, portable, have good-quality voice output, and are available for an appropriate loan period. Murphy[19] highlights the importance of spending time discussing vocabulary and the types of messages people want. This requires regular monitoring and review as needs and circumstances change. Appropriate modifications or new strategies may need to be found. More than one mode of AAC may be used for different environments or situations. Occasionally, a person may not perceive their loss of speech to be significant and will therefore be less likely to embrace AAC.

Efficient methods of communicating the needs of day-to-day living should be well established early in the disease and modified as required. These should be clearly documented to ensure that others are aware of them, particularly if the person moves from their home to facility-based care. This ensures communication boards and electronic devices can be updated and relevant yes/no questions asked, and avoids anxiety and frustration when the person has to repeat directions to meet changing needs.

Figure 14.1 provides a general framework and summary of practice guidelines to guide clinicians with their AAC decision-making.

Involvement of communication partners

Communication does not occur in isolation, and any intervention should include the communication partners. The inclusion of communication partners should begin early when the AAC option is first being discussed. Adjustment to AAC has an impact on everyone with whom the person interacts. For this reason, family, friends, carers, and health professionals should be included in training, education, and therapeutic interventions. Spouses have reported adapting social activities in order to maintain their partner's involvement in the community as well as with family and friends.[22] When an AAC strategy or device is used, effective communication is dependent upon the active participation of all. The communication partner may be required to assist with positioning, setting up, or maintaining the AAC technology. For some people, this role can be an added burden and the SLT must be sensitive to the needs of whole family and provide ongoing monitoring and support. This support should be provided by the most suitable mode of communication—face to face, e-mail, Skype—whatever works best for the family.

For people with ALS and FTD or significant cognitive impairment, the use of AAC can be particularly challenging and places an extra burden on the

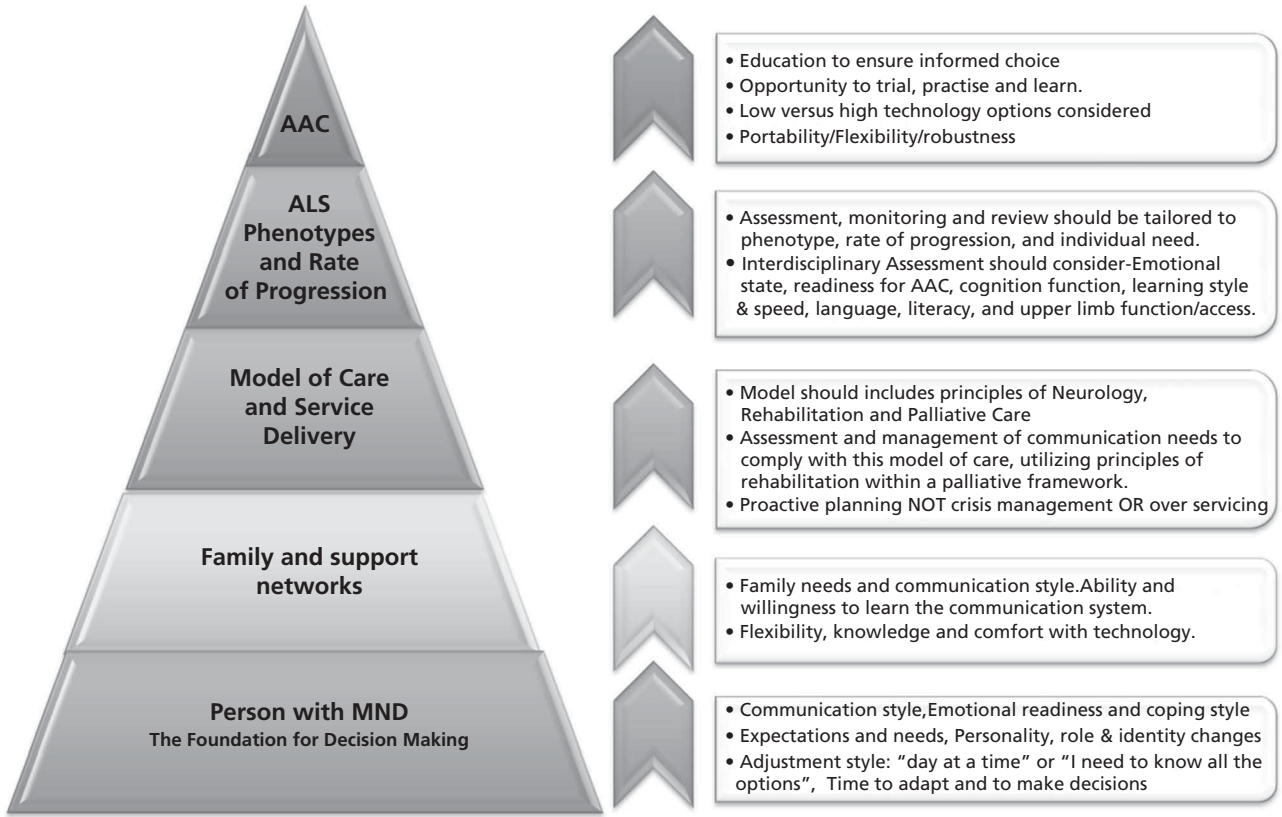

Fig. 14.1 Framework for AAC decision-making.

communication partner. Low-tech options have been most successful along with specific training in partner-dependent strategies.

AAC approaches

There are two general approaches to AAC available—low-tech and high-tech.

Low-tech options

Low-tech options include voice amplifiers, pen and paper, magnetic writing boards, and alphabet charts. These options are quick, convenient, and portable and require little training. Many find these low-tech AAC methods suitable if upper limb function is adequate. With more advanced physical deterioration, and when fatigue and/or cognitive impairment become an issue, pre-recorded message boards, alphabet boards, and picture boards can be more effective.

Eye-gaze boards such as ETRAN boards™, or Eye link, which only require eye movements to indicate target letters, are useful. Since these depend on appropriate skills for both the person and their communication partners, intensive training and practice for all concerned is important to facilitate effective communication.

A GEWA Laser Pointer™ fixed to a pair of glasses, a headband, or a hat and used with individually designed alphabet/phrase boards can be effective for some in the later stages of the disease.

High-tech—text to speech options

High-tech AAC options include electronic communication devices such as the Lightwriter™ text to speech software, computer packages, and computer-based communication devices. The Lightwriter™ has the advantage of voice output, which enables telephone use and communication with young children and in groups. Prediction functions can assist people with poor keyboard skills or impaired dexterity. SMS messaging is also a feature of this device. Scanner options maybe appropriate as they can be added at a later stage if upper limb function deteriorates, although they are slow and other more efficient options should be considered. It is important to liaise with the occupational therapist when setting up scanning systems to find the best switch access. The role and needs of the communication partner must also be considered.

Mainstream technology for AAC

There has been rapid growth in mainstream technology and this is now offering people with ALS some new AAC opportunities.

The SLT must objectively evaluate the benefits and limitations of these new technologies—and not 'get caught up in the hype and excitement'. Patients and families need information and support to make informed decisions regarding the most appropriate AAC options that will meet their needs now and in the future.

iPads and Android tablets have many advantages—and one of the most important being that they look mainstream and 'normal'. They are portable, lightweight, and are reasonably robust if placed in a protective case. For people with good upper limb function, such as those with bulbar onset ALS, they are easy to use and provide the person with easy access to the Internet via WiFi or 3G.

Currently, there are several apps available that are able to do 'text to speech' in varying ways. In some apps you are able to type in a phrase or sentence, then press 'speak' or similar, and the software will read aloud what you have written. Some apps allow you to copy and paste text from somewhere else (e.g. the Internet, an electronic book, a saved document, etc.) and it will then read the text aloud. Other apps are able to read straight from a website, without the need to copy and paste (e.g. visit a newspaper's website to have the app read the paper aloud to you).

There are a range of websites that provide up-to-date information: for text to speech apps for the iPad see <http://appadvice.com/appguides/show/text-to-speech-apps-for-ipad> and for more specific information on text to speech communication apps see <http://www.spectronicsinoz.com/article/iphoneipad-apps-for-aac>. At present there is limited access to tablet technology for those people with reduced upper limb function. Switching is an option with the iPad via the 'Predictable' app. This is fairly slow and other AAC options may need to be considered, such as a laser pointer or head mouse.

Computer options for AAC

A number of assistive technology solutions are available to assist people to access technology.

TrackerPro is a mouse replacement device that enables people with little or no hand movement to perform all mouse functions by moving their head. It is attached to the computer via a USB port and a high-resolution camera is mounted to the top of the computer screen, laptop, or communication device. A tiny reflective dot is worn on the forehead or glasses and TrackerPro's camera tracks the person's head movements. It can be combined with some AAC software such as the Grid and for some people can be an effective AAC option. Clicking can be accomplished by using ability switches or Dwell Clicker™ software. Dwell technology built into many AAC devices also works.

Eye-gaze computer systems, such as the MyTobii™ or the system by LC Technologies, are operated entirely with the eyes. Eye movements are calibrated and by looking at control keys on a screen, a person can synthesize speech, control the environment (lights, call bells, etc.), type, run computer software, operate a computer mouse, and access the Internet and e-mail. These systems are costly but effective, and can significantly improve a person's quality of life. They do require set up and some level of knowledge and comfort with IT. It is also beneficial to have a low-tech option for fast efficient communication that is portable and easy to use in other environments such as outdoors or in the bathroom.

High-tech AAC options may never be suitable for some, while for others they can enhance communication significantly. In the authors' experience, young tech-savvy patients with a disease that is moving relatively slowly are usually suitable candidates for a high-tech system. It is important to remember that high-tech communication systems cannot give words intonation, spontaneity, humour, or wit. An investigation of why people choose not to use an electronic device found a variety of reasons.[19] Some felt that by not using speech they were admitting defeat, others would rather say it than write. The shared understanding of daily events in close and long-term relationships was considered to reduce the need for things to be said by some people. It was reported that as the disease progresses communication becomes restricted to familiar people and communicating with unfamiliar people is too tiring. Because a significant amount of communication is non-verbal, some felt the AAC device would remove closeness. The slowness of the speech output and the voice quality of electronic devices were deterrents. Others complained the learning requirements were too complex and time-consuming.

Communication during the later stages of the disease

The SLT, along with other members of the multidisciplinary team, must facilitate and support the person with ALS and their family to ensure good-quality communication throughout the disease. This will support the person to remain active in the decision-making process.

Ideally, early in the disease process, people with ALS and their families should be asked to make decisions about what types of treatment they would like and what issues are important to the maintenance of their quality of life. This information is often recorded in an advance care plan. These 'conversations' occur over a number of sessions and allow time for the person and their family to make informed decisions about the kind of medical interventions or care the person with ALS would like to receive in the later stages of the disease.

Efficient methods of communicating the needs of day-to-day living should be well established and modified as required. These should be clearly documented to ensure that others are aware, particularly as the person moves from their home to facility-based care. Communication boards and electronic devices can be updated and relevant yes/no questions asked, to avoid anxiety and frustration when the person has to repeat directions to meet changing needs. Communication of wants and needs is particularly important during this later stage.

Strategies found to be helpful during this later stage are:

- lists of daily needs, likes/dislikes
- common phrases, sayings often used
- outline of the daily routine—this is particularly useful if there are multiple carers
- digital photos of comfortable positions (the person can then indicate with yes/no responses if they want to vary their position, with the photo used as the starting position); this is helpful for people who have difficulty finding a comfortable position.

At night, the following strategies may be employed:

- photos of various sleeping positions (helpful if the person is waking at night and needs to adjust their position)
- list of possible reasons the person may wake at night—this list is put together with the patient and family/staff with yes/no responses used to confirm the current issue. This increases the speed of communication for carers/family/nurses at night.

At this stage, it is imperative to focus on the persons' communication style and their needs and allow them to stop using high-tech devices and consider other options without feeling they are letting someone down. Some will persist with options that are no longer suitable and may need gentle encouragement to try other strategies.

In formal care settings, such as a hospital, a nursing home, or a hospice, the provision of staff who are familiar with the patient can become a point of contention. When communication is difficult, either because speech is difficult or communication via an alternative means requires skill and cooperation from the communication partner, individuals often express a preference for specific staff. This is usually those people who are best able to communicate with that person. If only one or two members of the team are able to effectively use the communication system this can create tensions amongst the team—with 'avoiders' not knowing how to communicate with the patient other staff may develop a tendency to be 'over involved'. Professional boundaries must be maintained to

ensure appropriate patient-centred care. The SLT must provide training in AAC to all those who care for people with ALS. Staff should learn the AAC system and have opportunities to practice. Photos, video footage or step-by-step guides, and e-learning/online packages can be helpful resources for staff.

Conclusion

The SLT, as a member of the multidisciplinary team, has the expertise and knowledge to provide effective communication solutions and ensure the patient's wishes and needs are respected.

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Chapter 15

Multidisciplinary care: nursing care

Dallas A. Forshew

Summary

A nurse is a generalist who needs the ability to be all things to all people at an ALS centre. She or he must have the qualities of flexibility, creativity, warmth, empathy, and more, acting as a specialist practitioner, manager, counsellor, coordinator, educator, supporter, researcher, mentor, and innovator. The nurse has an essential role in the support of the person with ALS, their family, and the other members of the multidisciplinary team.

Introduction

The role of the nurse in ALS is as varied and complex as the needs of patients and their families. The nurse cares for the patient and gives guidance from the time of the initial diagnosis through to the terminal stage of the illness. The nursing role is concerned with a wide spectrum of needs and responsibilities—emotional and psychological, education regarding the disease process, referrals to community agencies, sorting through insurance and governmental regulations, the rationale for and practical aspects of interventions such as feeding tubes and non-invasive ventilation (NIV), and symptom management.

In addition, the nurse in a multidisciplinary team is often responsible for the general coordination and management of the team members and clinic organization.

At the time of diagnosis

The period of investigation, while the diagnosis is unclear, may well be the most emotionally difficult time for all concerned.[1] While the diagnostic work-up is in process, there is a sense of 'hanging in the air'. Lives may be put on hold while the reasons for the patient's symptoms are sorted out. Most patients know that there is something seriously wrong: when the diagnosis is given, the reaction may be tinged with some relief because 'at least now we know what we are dealing with'.[1,2]

However, the overwhelming feelings of the patient and family revolve around the knowledge that their lives are changed forever. Their hopes and expectations for the future are shattered. It is the responsibility of the multidisciplinary team to help them find ways to readjust their expectations to meet their new reality. In fact, this readjustment must happen over and over again throughout the disease process as every loss of function is grieved for.[3,4] ALS is not a static disorder—it is dynamic and it seems that every time an adjustment is made to a new loss another loss arises and adjustment is needed again.

The nurse has two main goals at the time of diagnosis. First, the patient needs to leave the clinic with a knowledge of the disease; second, the patient needs to feel supported and secure in the belief that he or she will be well cared for.[4–6]

The physician gives the diagnosis and initial information to the patient, but no matter how sensitive and skilled the physician is, the patient may not hear much after the diagnosis is initially discussed. The patient will immediately benefit from a session with a nurse if one is available. The nurse can augment the physician's discussion by quietly listening to the patient's interpretation of what was said and then talking in plain language about what ALS is and what it is not. The role of dispelling myths about the disease is just as important as giving new information. Written information should be given and the patient told that they may telephone with questions or concerns. Many clinics have a folder of material that is given to every new patient. Referral to the local support agencies, including the local ALS/MND associations, is critical.

Attention to the family

ALS affects not only the patient but also their family and close friends:[4] many patients will admit that they believe the disease is harder on their family than on them. It will give patients a great deal of comfort to know that their family will be given attention. This can be accomplished in many ways. It is important to consider the family members and simply ask how they are doing, such as enquiring if they are getting enough sleep, because the nurse can make suggestions to ease many sleep issues. Do they have someone to talk to? Do they need counselling? Are they attending to their own medical care? Many caregivers let their own health decline because they are so focused on caring for their loved one. Do they get out of the home and get breaks from the caregiving role frequently enough? They must remain rejuvenated in order to provide the best care. Are they keeping close ties with their friends? They will need these friends throughout the disease but even more after their loved one has died.

Who attends clinic appointments with the patient is, of course, at the patient's discretion. Encouraging family and even friends to attend clinic appointments will contribute to better follow-through on recommendations because the others

will reinforce the recommendations at home. Everyone will have a better understanding of the rationale for the recommendations if several people have heard the discussion.[7]

Of course, the patient is always addressed directly during the interview, even if they have difficulty with their speech. A professional would never direct questions to the family if the questions should be directed to the patient. However, the family will often contribute to the clinic interview by supplementing what the patient says, and it may be appropriate to direct some questions to the family. This can bring important information to light.[8]

Patients and family members will often admit that they protect each other from certain information or feelings. They will all feel better supported if they are each given some private time during the interview. This can be extraordinarily empowering to each individual.

Coordination with support agencies

The role of support agencies is integral to the well-being of the patient and family. These agencies attempt to reach every community. They include such organizations as the ALS/MND associations throughout the world. The Internet has made it possible to reach out to those areas that are too remote for a local group (see the Appendix, which contains a list of useful websites).

The strength and range of services of the support agency will vary in each community. The basic services usually include education materials, support groups, and equipment loan. Those agencies that are better funded may offer special programmes for children, transportation and funding for accommodation to attend clinic appointments, help in the home for bathing and dressing of the patient, a nurse or other professional to visit the home, or a visiting programme to provide a companion while the caregiver goes out.

It is the responsibility of the nurse to know which services are available in the community and to coordinate with the agencies. The nurse can seek out information from the agency about their interactions with the patient and family that would be useful in their clinical care. Likewise, the nurse can coordinate with the agency after the patient's clinic visit or telephone contact. The nurse can request a home visit or recommend a piece of equipment and may ask the agency to reinforce a clinic recommendation.

Referral to and coordination with home care and palliative care/hospice

Home care services may be available to support someone at home—depending on local facilities and funding. It is most often the nurse who will carry out the referral and then keep in close communication with the agency. The services are

provided to the patient in their home and may include a home nurse, physiotherapist, occupational therapist, social worker, or home health aid. The clinic nurse should clearly state the needs of the patient and then keep in contact with the agency to make sure those needs are being met. Likewise, the agency may identify additional needs and seek approval from the clinic nurse to provide extra services. The nurse coordinates with the physician and team members to keep them informed and to seek their input. The patient is best served if the nurse and agency keep in close contact and coordinate care.[9]

Like home care services, the initiation of palliative care and hospice services requires further discussion and involvement of the wider multidisciplinary team. The referral should be made as early as feasible to best serve the patient and family.[4,10] It is often the nurse who will make the referral and give details to the hospice of the patient's and family's needs. In turn, the hospice will discover additional needs and keep in close contact with the nurse. The nurse will then communicate this information to the other team members, enabling the palliative care team and the hospice and clinic to collaborate and inform each other of the care they are providing.

Palliative care will provide a wide variety of services, including assessment and recommendations from professionals such as the hospice nurse, physiotherapy, occupational therapists, dietician, speech and language therapist, social worker, pastoral care, and hospice physician.

Most ALS centres in the United States will maintain their close relationship with the patient for all disease-related issues such as nutrition, breathing, and symptom management. Hospice agencies have particular expertise in such areas as pain management, constipation, urinary tract infections, prevention of decubitus ulcers, and the social issues that surround this final and very natural stage of life.[10]

Patient education

Education of the patient and family is fundamental to the role of the nurse in every disease state, and ALS is no exception. Armed with accurate information, the patient and family are better equipped to cope with every stage of the disease. Education starts on the day of diagnosis and occurs with every interaction. Patients are more likely to accept a recommendation if they understand the reasoning behind it and it is explained in language the patient understands.[11]

The nurse needs to make sure that the patient and family have a good understanding of the disease and its progression, the rationale for all interventions and all aspects of medication use.

Compliance with interventions is enhanced if the subject is brought up early within a broad framework and then, as the time for needing that intervention comes closer, more direct information is given. For example, high compliance with feeding tubes can be achieved through a stepwise approach. Near the time of diagnosis, the patient can be empowered with the knowledge that nutrition is an important part of care for ALS, that they have control over this area, and that the clinic will give them the information they need along the way. As the patient progresses, they are given information about how to enhance their diet to prevent weight loss. When it is difficult for the patient to keep up with their caloric needs and it is deemed that they will eventually need a feeding tube, mention of an alternative to oral intake can be made well before a feeding tube is needed. Small pieces of information given over time will help the patient become aware that the eventual need for a feeding tube is natural and thus acceptance is more likely to be easy.

A basic part of the nurse's own education revolves around techniques used to teach the adult learner. Attention must be paid to the patient's readiness to learn. The day of diagnosis may not be a good time to discuss more than the basics of the disease, but further information can be given at another visit in a few weeks. Some ALS centres in the United States offer an orientation class for patients with a new diagnosis. This might be given monthly by the nurse or through the local support agency.

The level of education and understanding is an important consideration in the discussion with patients and families. Not all will be literate, and even a person with several university degrees may have trouble with concepts that are far from their field. The nurse must use language, free of all jargon, that the patient can understand. Educational materials should be written at a reasonable level, using simple words and short sentences but without 'dumbing down' the material.

Awareness of cultural differences needs to inform the communication approach that the members of the healthcare team use with the patient. In some cultures, difficult medical information is given to the family and not the patient. In others, it is not polite to look the patient straight in the face while talking. Although these examples might present difficulties in today's age of openness and full information, cultural differences must be respected. There are ways to get the information to the patient that are still respectful of their heritage.

Written material is needed to reinforce what is taught in the clinic. Patients will often forget the details of what was said because they are overwhelmed. It is recommended that information be given verbally with time to ask questions and then reinforced with written literature. The nurse may write or provide multiple information handouts that cover topics such as medication rationale,

potential side-effects and dose escalation schedules, rationale for interventions such as NIV, instructions for use of the feeding tube, a summary of recommended Internet sites, books, agencies, a summary of any ongoing research that the patient may be able to participate in, and many more. The nurse may also mentor team members in writing handouts pertinent to their speciality.

It is incumbent upon the nurse and all team members to remember that patients and their families will need further reinforcement of all teaching. It is the nature of any serious disease that the patient can only remember so much at any given time.

Outreach education to community agencies

ALS remains a rare disease, but with the advent of speciality clinics there are now centres of excellence for patient care. This is not true for home care or hospice agencies, where a patient with ALS is infrequent. It is in the best interest of our patients that we reach out to community agencies and provide specialized education about the disease and the varied needs of ALS patients. This can be accomplished by talking with counterparts at the agency, whether nurse-to-nurse or therapist-to-therapist. The nurse has an excellent opportunity to do this when the referral is initially telephoned to the agency and the nurse sets up expectations for further communication. Such opportunities continue as a cooperative relationship develops between the clinic and the agency.

The nurse can also offer to provide educational activities for the agency staff whenever a patient is referred, especially if it is an agency that has not been used in the past. This is an excellent way to build relationships and educate at the same time.

Coordination and management of the ALS centre

The multidisciplinary team approach has become the standard of care for people with ALS.[12] The organization, coordination, and management of such a team are often the responsibility of the centre's nurse.[6,7]

Attention is given to the entire patient experience from the first phone call through to the last clinic visit. Every aspect of the centre should be welcoming. Patients do not look forward to coming to the centre, because they know that they will be confronted with more news of weakness and loss. Everything possible must be done to make patients *want* to come again. Building strong personal relationships and always finding something positive to say to the patient will help in this regard.

An ideal centre will give a high priority to having the telephone answered by a person rather than a machine. The reception area should be quiet and the

receptionist approachable. The clinic area should afford privacy within a professional atmosphere. It is often the role of the nurse to set these as required expectations for the staff.

Staff education is important for the patient's sake but also for the professional satisfaction of each team member. Team members are more likely to work well for the centre if they feel that the centre is invested in them. It is recommended that there are regular opportunities for a team meeting that involves a particular clinic issue. Articles can be distributed every week or so. There can be a journal club for the staff. The nurse can organize these activities and act as an advocate for the staff. The director can make time available for such activities and provide funds for attendance at conferences. The director can show their interest in the staff by participating in these efforts. The director can also support the nurses as the nurses support the staff. Nurses should attend ALS conferences on a regular basis and have the opportunity to develop collegial relationships with nurses in similar roles.

A yearly retreat provides time for team building, education, staff support, and addressing administrative issues. It is advisable to hold the retreat away from the ALS centre. Sections might be devoted to discussion of difficult clinical issues, ethical problems, communication and coordination amongst staff, research, stress reduction, or others. Staff will feel more involved in the process if they are asked to contribute to forming the agenda.

Burnout among staff can be devastating for patients, the team, and the individual team member. The nurse manager can take measures to prevent this. The provision of staff education is important. Everyone in medicine is stretched and works very hard. The nurse can be watchful and make sure that the work is distributed in a reasonable fashion. A regular or occasional support group for staff indicates the high value that the centre places on their well-being. The support group might be led by the nurse, social worker, psychologist, or someone from outside the centre. In general, a nurse needs to be available to the staff for any concerns that they might have.

The flow of patients and staff through the clinic should be organized such that patients are taken into their rooms quickly and have little waiting time between seeing team members. Team members will appreciate a simple but well-organized system by which they know where they go next so that their time is used efficiently. Every clinic handles these issues differently, but the nurse can ensure that they are managed well.

In terms of management of the centre, it is the nurse's responsibly to coordinate with the director and carry out the director's wishes. It is the director's responsibly to use the nurse as a partner in the management of the clinic, for the nurse will have the ear of the team members and an overall sense of the centre's coordination.

Research

Many ALS centres in the United States are in academic institutions. These centres will be actively involved in drug studies or clinical research aimed at improving care. Patients appreciate being able to obtain care at a centre that is involved in research: it gives them a sense that they are being cared for by professionals who are very knowledgeable and who are doing important work. This *is* important work, and it should be acknowledged as such by every team member.

The benefit of drug studies is obvious: the benefit of sound clinical research is just as important and can carry even more benefit. When a question is asked about clinical management of ALS which then leads to a research project, it has the potential to radically change the patient outcome: changing management practices can lead to increased quality of life and greater longevity in ALS patients.

The nurse and medical director of the centre can inspire the staff to become interested in clinical research. Staff can be taught how to critically read a research article. Staff can also be integrated into clinical research at the centre. Nurses are often already involved in this research and can be a strong resource. A nurse might help the team member think of a clinical question and assist, along with the medical director, with the design of the project. With nurturing, clinical research projects can become a routine aspect of the centre's activities.

Patient care during and between clinic visits

It is advisable that the nurse telephones each patient a week or two before the scheduled clinic visit. This will provide invaluable information that will enhance the patient's care.

The nurse can perform a variety of duties during the clinic visit. The nurse might be the organizer and only see patients who have specific issues that are not addressed by the other team members. More often, the nurse has individual time with the patient and family and may perform a general assessment of the patient at the beginning of the visit and determine which team members the patient will see at that visit. The nurse may focus on patient and family education regarding new or ongoing issues. This might prepare the patient for a more thorough discussion with an individual team member. An example might be noting the patient's weight loss and stressing the importance of nutrition to the patient in a general way. This may help the patient to be more receptive to the specific recommendations of the dietician later in the visit.

Nurses will often review medications with the patient and obtain the patient's perception of their efficacy and side-effects.

Each team member will make recommendations throughout the patient's visit. The nurse needs to make sure that the patient is given written recommendations and should discuss these recommendations with the patient at the end of the visit. This gives the patient an opportunity to ask questions and the nurse an opportunity to be sure that the patient leaves with all the necessary paperwork and understands all of the instructions. The nurse can develop forms for this and a variety of other purposes.

If a team member is absent from clinic, the nurse should have enough knowledge of that team member's speciality to be able to deputize. In most centres, the entire team is only available on a particular day every week or two but the nurse is in the centre full time. The nurse must be able to assess and give recommendations in every area. This often includes performing bedside pulmonary functions tests, demonstrating the use of equipment such as a walker, and assessing the patient's nutritional status.

It is good clinical care for the entire team to meet after the clinic to discuss each patient. The nurse might be designated to organize this meeting. The physician usually leads such meetings with ample time allotted for each team member to contribute. The purpose of the meeting is to coordinate patient care and to be clear on what needs to be done by who to follow-up on the patient's needs. It is often the nurse who is responsible for seeing that every team member completes their task. An offshoot of the meeting is that staff education can be a prominent part of the discussion. The rationale for a recommendation might be explained or the difference between one kind of motor neurone disease and another such as ALS and PLS.

The nurse may telephone the patient about 2 weeks after their clinic visit, allowing a review of the recommendations and assessment of follow-through. This is a suitable time for problem-solving and further education.

The nurse has many responsibilities between the patient's clinic visits and is often the person who handles most of the patient phone calls and gives guidance or arranges referrals. Again, education is a mainstay of the nurse's role. The nurse can refill prescriptions under the direction of the physician and complete many of the forms sent by equipment companies or agencies. Patients and families take great comfort in the knowledge that the nurse is available for support and counselling.

Impact of FTD

It is now recognized that frototemporal dementia (FTD) and related cognitive disorders are prevalent in ALS[4,5,10,13] (see Chapter 8). It is imperative that all professionals working with ALS patients are aware of this issue and are given

tools to better work with these patients and support their families. Patients with FTD will learn in an entirely different way from patients with only ALS, and may be unable to make sound decisions regarding acceptance of interventions.[14] However, techniques used to work with people with traumatic brain injury of the frontal lobe can bring about remarkable results. An invaluable resource for professionals and families dealing with this is the book *What if it's not Alzheimer's: a Caregiver's Guide to Dementia* edited by Lisa Radin.[15] This book defines clear strategies that can be successful when dealing with this issue.

It is important that the centre nurse and physician make sure that all team members are aware of this overlap syndrome and the techniques needed to work effectively with these patients. Extra support and insight are needed for the family. The family is able to cope with the behavioural and/or judgement changes much better if they understand the nature of the problem. The Family Caregiver Alliance (<http://www.caregiver.org>) provides downloadable education materials on FTD and behavioural changes. However, information should only be given to the family if they are ready to accept this additional problem. Some families will chose to ignore or deny the problem. These family members should not be confronted with information that they are not prepared to handle.

Bereavement rituals for families and staff

Nearly all patients with ALS will be referred for hospice care. Every hospice has a bereavement programme for families after the patient's death and the nurse can reinforce the utility of this programme to family members.[16] Family members may feel better more quickly if they take advantage of the bereavement programme, and this is important for all family members not just the spouse.

Members of the multidisciplinary team also need time and space to grieve. The work with the patient is intense and the staff will become attached and have feelings for the patient. Every time a patient dies, the staff feels a loss.

Many clinics have rituals that are helpful to both families and team members. One simple ritual is to send a condolence card to the family on which each team member has written a small personal note. This tells the patient's family how much the staff cares and it helps to bring closure for the staff. Some centres hold a yearly memorial for families of patients who have died under their care. This can be quite meaningful for the families and perhaps even more meaningful for the staff.

Taking care of the nurse

Just as burnout can be a problem for other team members, so it can be for nurses too. It may be that while the nurse is taking care of everyone else, she or he may

neglect to care for themselves. By nature, many nurses will do their best to work until all of the staff's and patients' needs are met. This is an impossible task, because the work is never finished and never will be. Nurses must heed the advice that they give to the staff; prioritize, pace yourself, take your holiday time, seek rejuvenation though outside activities, and use the medical director or other appropriate person for support.

Conclusion

The nurse has a crucial role in the activities of the ALS centre and the coordination of patient care and support. The nurse will hold the centre together and move it forward in partnership with the physician director. To a great extent, it is the nurse who is responsible for implementing the principles of excellence in ALS centres.

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Chapter 16

Complementary and alternative medicine

Gregory T. Carter, Sunil Kumar Aggarwal, Michael Weiss, and Richard S. Bedlack

Case history

Sarah, a 38-year-old woman, was diagnosed with ALS after an initial history of pain in her left shoulder and weakness and wasting of the arm. Gabapentin titrated up to 800 mg three times a day provided partial pain relief; the addition of nortriptyline was too sedating and caused a dry mouth. Venlaflaxine titrated to 75 mg twice daily improved comfort but pain management was suboptimal. Oral morphine adequate for pain relief induced somnolence and difficulty breathing. Both pain and the side-effects of the drugs detracted from her primary goal of parenting her two young children.

A friend suggested medical cannabis, but with no experience of 'recreational drugs', Sarah was initially reluctant to consider its use. Poor pain control prompted her to reconsider. After counselling from her physician regarding the medical uses of cannabis, including side-effects, she was given a formal authorization, per Washington state administrative code RCW 69.51A, allowing her to possess and use cannabis for medical purposes.

Under her physician's direction she used a glycerine extract of whole cannabis, which, after 3 days of dose adjustment, provided pain relief without undue sedation or drug interactions with gabapentin and venlafaxine.

The same medical regimen was followed until a more concentrated oil extraction of cannabis was substituted for use through her PEG tube. She required no other medications in the course of her ALS until the final days, when she accepted benzodiazepines. She died at home, peacefully, just after her 44th birthday.

Sarah's husband continues to volunteer at the medicinal cannabis cooperative and to advocate for the use of cannabis to treat symptoms of ALS.

Introduction

Use of complementary and alternative medicine (CAM) is increasing worldwide, particularly in industrialized countries. This is understandably common in patients with incurable, rapidly progressive, ultimately fatal diseases like ALS.[1–4] The use of CAM has been reported in 54% of 92 ALS patients.[3] Over 73 different methods or substances were listed, with some individuals using up to

11 different treatments—the most widely used were acupuncture (47%), homeopathy (40%), naturopathy (24%), and esoteric treatments (20%).[3] In most cases alternative treatments were performed by a physician. In our experience many more will ask about and use CAM, and some use many simultaneously.

Despite emerging studies showing the efficacy of some CAM, none report on effects on ALS symptoms. ALSUntangled, a group of 90 clinicians and scientists representing 10 countries, developed a standard method for reviewing CAM. While not specifically targeting palliative care, since 2008 comprehensive information on CAM treatments in ALS has been available on the ALSUntangled website (http://www.alsuntangled.com) (see Box 16.1 for reviews of treatments to date).

Reports of the use of CAM in neurological disorders consist predominantly of Level III/IV evidence.[1–4] Unfortunately, better evidence, via randomized controlled trials, is unlikely to be forthcoming any time soon. Even in studies of disease-modifying treatments or pain control ALS trial enrolment rates are low, highly variable, and not influenced by trial design factors.[5–7] Barriers to completing such studies include those common to other ALS trials, but also lack of financial support and difficulty interesting patients who can access the treatments outside the research protocols.

Benefits and costs

Patients with ALS can have a very high burden of symptoms. Thus the understandable use of CAM (and possibly other therapies with unproven claims) should be anticipated by healthcare providers. Strong beliefs and hopes of both patients and healthcare providers may compromise communication, but it is still a medical responsibility to discuss cost, potential adverse effects, and drug interactions as well as the putative benefits of CAM.

Patients with ALS have spent an average of US$4500 on CAM, generally without reimbursement.[1,3] Certain CAMs can cost thousands of dollars per month. Some costs, such as massage or acupuncture, may be reimbursed by insurers.[1,3] The duration of treatment must be considered as well. One study in the United States hospice system has shown that the length of stay and cost of care for ALS patients was over twice as much as that for lung cancer patients, particularly in those patients choosing aggressive care.[8,9] Thus the possible out-of-pocket costs for CAM therapies may be substantial. Nonetheless, the high symptom burden in ALS and the difficult decisions about the use of life-prolonging therapies, such as long-term mechanical ventilation, makes it is hard to view things from a purely financial perspective.[10] The impact of ALS is also felt by family caregivers who are left to deal with the often enormous physical, financial, and emotional

Box 16.1 ALSUntangled completed reviews

1 Lyme disease and Iplex
2 The Hickey Wellness Center
3 Stem cell transplants at the Hospital San Jose Tecnologico de Monterrey
4 The XCell-Center
5 The Stowe/Morales ALS protocol
6 Marty Murray's method
7 Hyperimmune goat serum for ALS
8 Low dose naltrexone for ALS
9 Spirulina (blue green alga) as a treatment for ALS
10 Luteolin and Lutimax
11 NuTech Mediworld
12 Dean Kraft, energy healer
13 Bee venom
14 Mototab
15 Coconut oil
16 Cannabis
17 'When ALS is Lyme'
18 Apoaequorin (Prevagen)
19 Sodium chlorite
20 Deanna protocol
21 Fecal transplants
22 Propofol
23 The Rife Machine and retroviruses

Source: data from ALS Untangled <http://www.alsuntangled.com/> (as on 25 September 2013).

demands associated with this disease. Expert multidisciplinary care may improve both the quality and length of life of people with ALS.[11,12] The recent trend to involve palliative care specialists as part of the ALS multidisciplinary team may ultimately lead to more cost-effective treatment of end-stage symptoms, including pain management, although this remains poorly studied. Regardless of the care model, the cost of CAM therapies for patients with ALS would still fall upon them and their families.

CAM in the palliative care of ALS

Cannabis

Intriguingly, the endocannabinoid system is implicated in the pathophysiology of ALS in animal studies, either directly as part of the underlying disease mechanism or indirectly inasmuch as this system plays a role in the homeostatic functioning of the neuromuscular system. Cannabinoids slow disease progression in mouse models of ALS, perhaps as an anti-oxidant, among other mechanisms involving endocannabinoid receptor activition.[13–15] Awareness of this literature increases the interest of patients in cannabis use, beyond the potential symptomatic benefits.

Cannabis has been reported by ALS patients to manage disease symptoms.[16–18] In a survey of 131 patients with ALS, those who were able to obtain cannabis found it moderately effective at reducing symptoms of pain, spasticity, drooling, appetite loss, and depression.[16] The main barrier to its use was availability, for either legal or financial reasons or lack of safe access.[16]

Pain increases in frequency and severity as disease progresses in ALS, and can have a pronounced influence on the quality of life and suffering.[11,12,19] Despite this, a Cochrane Review found no controlled trials specifically looking at the use of cannabis for pain relief in ALS.[19]

Cannabinoids are centrally and peripherally acting analgesics with a different mechanism of action from opioids, although the analgesic effect produced by cannabinoids and opioids may involve similar pathways at the brainstem level.[20–22] Multiple, well-controlled clinical studies have demonstrated the analgesic efficacy of cannabis in the treatment of pain.[18,20] Cannabis does not cause suppression of the respiratory drive.[20] A recent systematic review and meta-analysis of double-blind randomized controlled trials that compared any cannabis preparation with placebo among subjects with chronic pain showed a total of 18 completed trials.[20] The review concluded that cannabis is moderately effective for treatment of chronic pain.[20] Combining cannabis with narcotics may also be beneficial, since, as noted above, the opioid receptor system is distinct from the cannabinoid system.[21] The anti-emetic effect of cannabis diminishes the nausea associated with opioid medications.[21]

Spasticity, a cause of pain and dysfunction in ALS, is induced at the level of both the motor cortex and spinal cord through the loss of motor neuron inhibition.[22–24] Cannabis has an inhibitory effect via augmentation of gamma-aminobutyric acid (GABA) pathways in the CNS.[24] This produces inhibition of motor neurons at spinal levels in mice and clinical studies, mostly in MS patients, suggest that cannabinoid therapy reduces spasticity symptoms.[22–24]

Cannabis effectively dries the oral cavity and upper airway, potentially reducing the risk of aspiration pneumonia and increasing patient comfort. Other unproven but potential benefits include improved appetite, mood, and sleep.[18,25] Dosing is by titration to effect and dependent on route of delivery.[26] However, smoking cannabis is never recommended.

Entheogens (classic hallucinogens)

The use of classical hallucinogenic compounds is experiencing a renaissance in medicine and psychiatry whether as adjuncts to psychotherapy or to induce spiritual experiences.[27] Trials of these compounds in the treatment of existential or death anxiety have focused on patients living with cancer. Articles, such as one in the *New York Times* in April 2012 entitled 'How psychedelic drugs can help patients face death',[28] have been profiled by ALS patients in online forums addressing anxiety regarding death and dying.

The psychological rationale for this therapeutic approach in terminal illness may lie in the purported capacity of entheogens to facilitate powerful states of spiritual transcendence that exert in the patient a profound therapeutic impact with often dramatic improvements in psychological well-being.[29]

Enthnobotanical researchers coined the term entheogen (from the Greek, meaning 'generate god within'),[30] to generically describe substances, typically plants, that contain psychoactive substances that alter a person's perception of reality, used to bring a patient closer to God or the realm of the transcendent.[29] Entheogens have been used by in many cultures for millennia but primarily sacramentally to enhance a religious/spiritual ritual.[29] Many patients feel ill-prepared for death and that there is insufficient knowledge to help them make choices that will cover their medical needs (pain, sleep, etc.) yet honour and maintain their autonomy and dignity.[31,32]

Psilocybin is the most studied entheogen from a palliative/end-of-life perspective. It is the active compound found in mushrooms belonging to the genus *Psilocybe*, which contain the psychoactive tryptamines psilocybin and psilocin.[33,34] Other species also contain the weaker psychoactive alkaloid compounds baeocystin and norbaeocystin, which may potentiate the effects of psilocybin. Psilocybin has been used for centuries for religious purposes.[34] Its inclusion as a Schedule 1 drug by the US Drug Enforcement Administration (DEA) has limited research on it.

Psilocybin administered under supportive conditions produced a dose-related mystical experience in 72% of subjects, although 39% also reported feelings of anxiety and fear. 'Set and setting', or the emotional state of the user and the type of environment they are in, largely determine whether the experience is positive.[34] Follow-up over 14-months found nearly 90% of the

subjects considered it to be the most personally meaningful and spiritually significant event of their lives to date.[34] Supratherapeutic doses of dextromethorphan induced effects similar to psilocybin but with more acute cognitive impairment.[35]

The contribution of psilocybin or other entheogens to treatment of end-of-life distress in people with ALS remains to be determined. Palliative medicine is concerned with spiritual and psychological care, but the administration of medical therapies for psychospiritual purposes faces barriers in both law and medicine.

Ayurvedic medicine

Ayurvedic medicine (Ayurveda) preceded and evolved independently of Western medicine.[36] The Sanskrit word *Ayurveda* means 'the scripture for longevity'.[37] Developed in India approximately 3000 BCE the sacred texts were passed on through generations of oral tradition,[36] and Ayurveda continues to be practised in India and other South Asian countries. It seeks to treat and integrate body, mind, and spirit using a comprehensive, holistic approach that emphasizes diet, herbal remedies, meditation, breathing, and physical medicine (exercise, physical therapy, yoga, etc.).

Ayurvedic medicine (Ayurveda) defines health as a state where the mind and body are returned to their original, natural, optimally functioning condition.[38–42] In Ayurveda the three primary mind/body principles (*doshas*) create our unique specific mental and physical characteristics.[37] Typically there are one or two dominant doshas, with the others being recessive or less expressed. The three doshas, known as Vata, Pitta, and Kapha, also encompass human physiology.[37] Therefore, everything that is in nature is also within. This unique combination of the three doshas is called Prakriti, which is the blueprint or constitutional code for optimum health.

Ayurvedic medicines are mainly oral or topical herbal preparations, and are often classed as tonics or rejuvenators. Of the many plants utilized in Ayurveda the best known and most scientifically investigated is ashwagandha (*Withania somnifera*) or 'Indian ginseng'.[37,38] Its berries, leaves, and roots have been used as a diuretic, as a 'rasayana', an elixir that works non-specifically to increase human health and longevity, and as an adaptogen, a non-toxic medication that normalizes physiological functions, disturbed by chronic stress, through the correction of imbalances in the neuroendocrine and immune systems.[37]

Most neurodegenerative disease, including ALS, would be considered a disruption of the dosha 'Vata'.[40] 'Vata disorders' encompass a wide range of pathophysiological problems, including inflammatory, degenerative, obstructive, and

functional disorders. Most of the signs and symptoms of ALS have equivalent counterparts for Vata derangements as described in Ayurvedic texts. Thus the Ayurvedic treatment would be focused on bringing back equilibrium between the three doshas.[40]

Vata disorders are thought to result from (or are triggered by) a 'factor'.[41,42] This could be a toxin, a foreign protein, or antigen damaging the Vata dosha, which is responsible for functions of the central, autonomic, and peripheral nervous systems.[41] A typical ayurvedic treatment for ALS might consists of several procedures, starting with a samsodhan (cleansing), proceeded by kaya kalp (rejuvenation), and sattvavajaya (counselling/cognitive therapy). A strict diet and lifestyle regimen would also be followed.

Ayurveda treatments can complement standard Western medical therapy; a patient will not have to choose one or the other. Moreover, adherence to ayurvedic regimens may improve general health substantially. Yet, even in Ayurveda, ALS would be considered 'kashta saadhya roga' (incurable).[40]

Other CAM modalities

Oriental/Chinese medicine and acupuncture

Traditional Chinese medicine is a system of medicine with theories, diagnoses, and treatments such as herbal medicine, acupuncture, and massage.[43–46] These theories derive from many sources including the theory of yin–yang, the five phases of natural phenomena, the human body channel system, Zang Fu organ theory, and others. Often Qigong (pronounced 'chee-gong'), a practice that integrates meditation, physical movement, and mind–body duality, is also strongly affiliated with traditional Chinese medicine. Acupuncture is the Chinese practice of inserting fine needles through the skin at specific points with the intent of curing disease, relieving pain, or optimizing well-being. Support for acupuncture as a therapy for ALS is lacking. However, there is a published report of two ALS patients having improved symptoms following injections of a homeopathic mineral and botanical medicine into acupuncture points over 4 weeks.[46] Reiki is a Japanese technique for stress reduction and relaxation that is administered by 'laying on hands' and is based on the idea that an unseen 'life force energy' flows through us and negative alterations in this energy produce sickness.[47,48]

Manual medicine: osteopathic manipulation, chiropractic adjustment, and massage

Manual medicine involves various procedures by which a practitioner's hands make direct contact with the body to treat articulations and/or soft tissues

through the use of specifically directed manual force. These systems of therapy hold that disease results from a lack of normal nerve function or alignment and employ manipulation and specific adjustment of body structures, including the spinal column. Massage involves manipulation of tissues (by rubbing, stroking, kneading, or tapping) with the hand or an instrument for therapeutic purposes.

Nutritional counselling, herbal and botanical medicine

Herbal medicine products are dietary supplements taken with the intention of improving health. Many have long traditions of claimed health benefits.[45] They are sold as tablets, capsules, powders, teas, extracts, and fresh or dried plants. However, some can cause health problems, some are not effective, and some may interact with other drugs that patients may be taking.

Yoga

Yoga is a mind–body exercise which combines controlled breathing with postures that focus on strength and flexibility with the goal of increasing relaxation and stabilizing mood.

The use of manual medicine, nutrition, and yoga has not been studied in the palliative care of patients with ALS, thus there is little discussion or recommendations for their use guidelines such as the practice parameters of the AAN.[11,12] Nonetheless, in our experience, all of these treatments may provide relief of discomfort and distress in some patients with ALS and should be given some consideration.

Future directions

Virtually no clinical trials have been conducted to demonstrate a role for CAM modalities as adjunct therapy in ALS. However, given their widespread appeal, potential benefit, and perceived low risk of harm, more research is suggested, specifically the need for randomized, controlled trials of these treatments. Until then, keeping an open mind about the utility of CAM seems like a reasonable approach to addressing the concerns of ALS patients from a palliative standpoint.

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Chapter 17

End-of-life care: ethical issues

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Summary

Many ethical issues need to be faced when caring for patients with ALS and their families—in the making of decisions, especially regarding nutrition and respiratory support, and at the end of life. Increasingly there is discussion about and practice of assisting dying—by assisted suicide or euthanasia. All of these areas need to be considered carefully while caring for someone with ALS, and everyone involved in the care of patients and their families needs to be aware of the ethical issues involved.

Case history: an ALS patient with cognitive changes

Mr M is a 59-year-old man with progressive dysarthria. Highly educated, he has been married for 38 years, has four grown-up sons, and has worked for years as an engineer. Bizarre behaviour and dysarthria caused his company to suspect alcoholism, and he was let go. His wife is divorcing him because he has become aggressive and 'changed' over the last 3 years. Mr M's biggest complaint is that he is unable to focus on words while reading. Evaluation reveals bulbar-onset ALS with FTD as the presenting feature.

Initially he has no life-threatening complications, but lacks insight into his condition. He has not designated a durable power of attorney and is unwilling to do so. His wife has told their children that he is 'faking' a disease to avoid a large divorce settlement and they are currently estranged from the patient. He reluctantly agrees that his physician can contact his sons. Once educated about the disease and the dementia they become more involved.

Over the ensuing months, Mr M develops dysphagia with weight loss. He accepts gastrostomy tube placement but insists on continuing to live independently. He progresses to anarthria and communicates by writing. His language significantly deteriorates and his clinicians doubt his ability to make end-of-life decisions. His oldest son attends the next clinic visit so decisions about an

advance directive and durable power of attorney can be established while he is still capable of communicating his wishes. Shortly thereafter, progressive dementia requires admission to a nursing home with subsequent hospice care. His sons were responsible for all decisions during the end stages of his disease.

Introduction

Morality refers to a shared set of beliefs about norms of right and wrong that prevail in a particular culture or subculture. Various cultures exhibit great differences in their customary morality. The term ethics, used by philosophers, refers to a method of examining and understanding morality. No single ethical vision prevails in Western pluralistic societies. The two extremes of imposing the morality of the group on those who differ, and non-interference in the moral lives of others, may coexist. Clinical ethics is usually framed as a system by which one examines morality, conduct, and social practices. The aim is to arrive at an answer to the questions 'Why should I (or we) do X or Y?' and 'What reasons would justify such action and why?' rather than to 'What ought I (or we) to do in this and similar situations?'. Four broad principles form the basis of ethical obligations:

1 *Beneficence* refers to the duty to benefit and further the welfare of patients.
2 *Non-maleficence* refers to the obligation to prevent harm and minimize risks.
3 *Autonomy*, also referred to as *respect for persons*, refers to the duty to protect and defend informed choices of patients with decision-making capacity.
4 *Justice* refers to an obligation to protect healthcare access and distribution of healthcare benefits.

In addition to these principles the established clinician–patient relationship includes implicit moral obligations of respect for privacy and confidentiality, honest communication about diagnosis, treatment, and prognosis, determining the capacity of the patient to participate in shared decision-making, and providing ethically valid informed consent. Paternalism, imposition of the physician's personal view of the patient's best interests, undermines autonomy and should be avoided. A moral dilemma may arise when an ethical obligation exists on both sides of a choice to perform or not perform an action, and ethical analysis provides a reason to support both alternatives. Clinical ethics offers a framework to justify, interpret, analyse, and potentially resolve moral dilemmas that arise within the context of patient care. A complete discussion of clinical ethical analysis is beyond the scope of this chapter. We favour the method of clinical pragmatism, a case based method of problem-solving.[1–3]

Ensuring a good death is an essential aspect of ALS care but presents a multitude of ethical issues. Depending upon the disease trajectory, life-threatening features of bulbar and/or respiratory muscle dysfunction may occur early or follow months or years of progressive weakness. [4–7] Delay in diagnosis, common in ALS, may confound recognition of their life-threatening potential until bulbar and/or respiratory dysfunction are fully manifest.[8,9] Once ALS is confirmed, the patient is faced with the inevitability of death, though the time frame may range from weeks to years. This reality forces the patient to make choices about life extension via gastrostomy tube insertion and/or mechanical ventilation and comfort during the terminal phase.[4,10,11] The ability to make this choice may be compromised by frontotemporal cognitive dysfunction.[12–14] Ideally, the clinicians caring for these patients should recognize life-threatening symptoms, monitor decision-making capacity, manage potential end-of-life scenarios, and provide comfort and hope whilst aggressively managing terminal symptoms.

Decision-making

Autonomy

In societies that emphasize autonomy, an informed, adult patient who possesses capacity usually makes the decisions concerning their medical treatment. A patient with decision-making capacity understands their condition and potential treatment options and effects, demonstrates reasoning consistent with the medical facts and their own values, and communicates their choice and its potential ramifications. Individuals with capacity are free to refuse any medical intervention before (withholding) and after (withdrawing) it is initiated even, for example a gastrostomy tube or tracheostomy, if this leads to death.[1,2,15,16]

Delegated decision-making

In societies where individual autonomy is paramount, someone with ALS may appoint a surrogate decision-maker in the event that they become incapable of making or communicating medical decisions. This may be informal, but is most effective if it is legally binding by appointment of a durable medical power of attorney. The decisions of the delegated surrogate are usually honoured as if the incapacitated patient themselves had made them. Optimally the surrogate exercises substituted judgment by making decisions using the known and expressed values or expressed choices of the patient. This should apply even if this decision conflicts with the choice the surrogate would make for themselves in the same situation. If patient values or choices are unknown the surrogate can choose based upon their own opinion of the patient's best interests.[1,2,17]

In some cultures someone with ALS may delegate medical decisions to a spouse, adult child, adult children, a family elder, a family group, a friend, or other trusted individual even if the patient has decision-making capacity. The person with ALS may refuse to be informed of the diagnosis and prognosis. While this may be considered an abdication of individual autonomy, it can also be viewed as an autonomous choice to delegate decision-making based upon established social or cultural practices. Whether breaking the news of the diagnosis or informing the patient of progression of disease, the clinician should anticipate the possibility of social or cultural differences in handling medical information and respect the patient's wishes. This may require the clinician to communicate with the person or persons designated by the patient to make decisions for the patient.

Impaired capacity

FTD is now recognized in the clinical and neuropathological spectrum of ALS (see Chapter 8). This may present as a progressive change in personality with changes in social and personal conduct, often with disinhibition causing impulsive, inappropriate, and compulsive behaviours for which the patient has little insight, or less commonly as a language disorder characterized by word-finding difficulty and poor category naming that may progress to muteness. Subtle cognitive impairment may be evident only with formal testing for mental flexibility, verbal fluency, abstract reasoning, verbal memory, and visual memory. Early detection is best accomplished by caregiver interview.[12–14,18–25] It is difficult to predict to what extent FTD will compromise capacity. As in Alzheimer's disease, ALS patients with FTD may lose decision-making capacity as behavioural and language deficits progress.[26,27] It cannot be assumed that someone with ALS will be capable of a discussion of advance directives when life-threatening complications of dysphagia and respiratory muscle weakness develop. For this reason clinicians should discuss advance directives with willing ALS patients with capacity before the development of life-threatening complications and before FTD develops or progresses. Recognition of FTD should prompt ALS clinicians to encourage patients to legally appoint one or more willing and knowledgeable medical surrogates and discuss treatment preferences with them in advance of cognitive decline.

An ALS patient with decision-making capacity can communicate a choice and discuss the potential ramifications. This may be significantly compromised in individuals who are unable to speak because of bulbar dysfunction and are limited in their ability to use their hands for writing or typing on a computer keyboard. Some individuals can utilize AAC devices to overcome these obstacles, but community resources may limit access. Meanwhile, progressive motor

dysfunction will limit the time frame in which AAC devices can be used. Early impairment of communication should prompt clinicians to discuss advance decision-making with willing patients before effective communication is lost. Clinicians should also encourage the appointment of surrogate decision-makers from within the family.

End of life

Discussion triggers

Many aspects of ALS are inconvenient, demoralizing, and even painful without being life-threatening. Whereas some may present with life-threatening bulbar or respiratory dysfunction, others have a more chronic, progressive course. The timing of discussion of end-of-life issues varies by patient. It depends on the symptoms and signs present, but is also dictated by the patient's desire for such a discussion and willingness and ability to participate in it. FTD should be considered in the timing of discussion. The first discussion of end-of-life care may be at diagnosis, either at the request of the patient or prompted by life-threatening symptoms. More commonly, at diagnosis a brief discussion about the progressive nature of the disease is revealed without the details of decisions regarding end-of-life care. Unless necessary or requested by the patient, excessive focus on terminal disease at diagnosis is considered by many clinicians to be ill advised as this does not foster hope.[10]

Is it near?

Comprehensive care in ALS includes routine symptom screening for bulbar and respiratory dysfunction.[10] NIPPV is usually instituted when the FVC falls below 50% of predicted. Discussion of NIPPV should prompt a discussion of progressive respiratory failure and advance decision-making, including both tracheostomy (for long-term mechanical ventilation, LTMV) and palliative care, usually facilitated by the institution of hospice care.[10,11]

Life-threatening aspects of bulbar disease include malnutrition and dehydration and inability to handle secretions, predisposing to aspiration pneumonia. Weight, swallowing dynamics, and handling of secretions inform the physician about progressive bulbar disease. Dysphagia, with weight loss of more than 10% of body weight, should prompt a discussion of gastrostomy. If gastrostomy is declined, hospice and palliative care should be discussed.[10,11]

Uncertainty

When considering the end of life, clinicians must accommodate to uncertainty regarding the 6-month life expectancy of any person with ALS. Uncertainty is

commonly present when the FVC is below 50% of predicted in a patient without symptoms and abnormal respiratory signs. Confidence can be increased by reframing the question as 'Would it be a surprise if this patient were to die within 6 to 12 months?'. Truth-telling obliges the clinician to discuss uncertainty while initiating or continuing advance decision-making. It is better to have this discussion too early than too late.

Presenting the options

Informing the willing patient and/or their chosen or legally appointed surrogate(s) about end-of-life options fulfils ethical responsibilities regarding autonomy, beneficence, and truth-telling. Paternalism is avoided by discussing all options in a balanced, unbiased, and factual way. An either/or choice paradigm in which the pros and cons of each choice are explored is most practical. When the option of tracheostomy and LTMV is discussed, so is the alternative of palliative and hospice care. The alternative to feeding tube placement is discussed in parallel. Regardless of the decision regarding life-extending or palliative care options, the patient should not be abandoned, and a plan of care should be put in place—palliative care may be instituted early in disease progression, alongside other care (see Chapter 2). While these conversations may be difficult for both the clinician and the patient, they should take place in time to allow both the patient and the family to process the information and discuss the issues at home. Follow-up visits should be scheduled quickly to allow for questions and for information to be presented. The physician must make clear that the patient can change his or her mind about treatment, and should actively but gently re-solicit the patient's wishes at subsequent visits, as preferences for treatment options often change over time.[28–30]

Life extension—pros and cons

Gastrostomy may extend life for those with severe dysphagia at risk of dehydration and malnutrition. The device can be placed with minimal risk and discomfort and is easy to use and maintain. It allows patients to continue to eat by mouth for pleasure and avoid anxiety-provoking and socially compromising coughing and choking. It provides access for medications to manage symptoms of ALS.[31–36] Some patients express concerns about a negative body image and dependence upon medical technology. Non-invasive respiratory aids can extend life and delay tracheostomy.[37] Attitudes regarding this vary between and even within cultures.[38] Tracheostomy for LTMV can extend life after respiratory failure. With good nursing and supportive care patients can be maintained for many years beyond the predicted natural history.[39–42] Small, portable, rechargeable ventilators can be carried on a motorized wheelchair and afford

mobility options to optimize social interactions. Some patients may continue to work. Unfortunately tracheostomy often renders speech impossible, except for those who can use a Passy-Muir valve.[43–45] AAC technologies may allow patients to communicate for a time (see Chapter 14).[46–48] However, disease progression will eventually culminate in the locked-in state making communication near-impossible.[49–51] Tracheostomy may compromise swallowing, necessitating a gastrostomy tube and reduction or cessation of oral feeding. If it is not an issue at the time a tracheostomy is placed, FTD may eventually develop. Because communication may be limited, FTD may be difficult to fully detect. FTD may compromise use of AAC technologies. LTMV is possible at home only for those with willing caregivers or home access to medical professionals. Institutional LTMV is an option for those without these benefits. In societies without universal medical access LTMV has financial ramifications that often limits this to individuals with insurance coverage or significant means.[52–54] Despite these limitations, patients on LTMV who can communicate may report a satisfactory quality of life.[55–57] Caregivers are, however, heavily burdened.[56,57]

Case history: a locked-in patient

Mrs C, a 57-years-old woman, has had ALS for 8 years but is new to the ALS clinic. She is accompanied by her husband, who provides the history. She had a tracheostomy for LTMV 4 years ago. Currently she is anarthric, quadriplegic, and uses a feeding tube for all liquids and nutrition.

Her husband states that she communicates by facial and eye movements that only he can decipher. On examination she is sitting in a motorized wheelchair. She has a tracheostomy with a portable ventilator. She has spastic quadriplegia with facial diparesis. She has slowly roving disconjugate eye movements and frequent facial grimacing. She does not visually fixate. She does not appear to voluntarily move her eyes or any portion of her face to command. She fails to respond to yes/no questions with ocular or facial movements. To the clinic team, her facial and ocular movements appear random.

These concerns are conveyed to Mr C but he vehemently disagrees, stating that only he can decipher his wife's eye and facial movements. You ask whether Mrs C had discussed or considered termination of the ventilator if she became unable to communicate. He does not recall any discussion of this.

Follow-up over 6 months convinced the clinic team that she is locked-in. On each visit the same concerns are discussed with Mr C. He continues to insist his wife is able to communicate with him. Next visit he reports his wife was thrown from the wheel chair onto the floor of the van in a motor vehicle accident on the

way to the clinic. Although she appears neurologically unchanged, there is swelling of the right calf suggestive of a fracture, which is confirmed by X-ray. She is treated with casting and analgesics. The lack of any indication of pain from her fracture further supports a diagnosis of the locked-in state, but he insists she is able to communicate. On the next clinic visit Mrs C is neurologically unchanged. Ophthalmological examination demonstrates severe conjunctival infection and a weeping purulent exudate of the right eye. She does not appear uncomfortable and is not blinking excessively. Once again the clinic team discusses locked-in syndrome because she shows no response to a painful condition. Mr C considers the issue over the next 2 weeks. After consulting with his daughter he decides that his wife is indeed locked-in and it would be in her best interest to withdraw care. He arranges to admit his wife to a local hospital. Her ventilator is discontinued after administering morphine and lorazepam. She dies peacefully.

Hospice and palliative care: pros and cons

Education regarding the benefits of palliative care and hospice involvement should be the focus for patients with life-threatening respiratory muscle weakness electing to forego tracheostomy and life-threatening dysphagia electing to forego gastrostomy tube insertion. Palliative care may also be offered to patients on ventilators or those using feeding tubes, although in the United States Medicare-funded hospices usually cannot accept patients on ventilators. There is under-utilization of palliative care in some countries and a need to educate patients and families about the effectiveness of hospice or palliative care for advanced ALS. The variability of presentation and progression in ALS causes an array of end-of-life problems that are well suited to interdisciplinary, multimodality comprehensive hospice care.[57] The physician should educate willing patients about the dying process and any potential symptoms they may experience, while emphasizing the potential for treatment. These include treatment for dyspnoea, air hunger, fear, and anxiety, and less frequently pain and choking.[58–62] Physicians should emphasize that they will not abandon the patient, but continue to provide care to minimize suffering during the dying process. If feasible, patients should be encouraged to continue visiting their physician once hospice care is initiated to minimize feelings of abandonment. While surviving caregivers report that despite hospice and palliative care patients suffer significant symptoms at the end of life, hospice patients are more likely to die outside the hospital, in their preferred place, and receive morphine. Despite significant symptoms, caregivers report their loved one was at peace, prepared for, and accepting of death.[59,60]

Purposeful withdrawal of life-extending measures and the principle of double effect

In some societies autonomous individuals with decision-making capacity or the selected or legally appointed surrogate may discontinue previously embraced life-extending measures. While withdrawal of treatment may seem and feel different from withholding a medical intervention, most Western societies do not draw an ethical distinction between these choices. Under medical supervision, mechanical ventilation can be terminated or weaned over 30 to 60 min at home or in a hospice or hospital. The patient and caregivers should be advised that death may occur within minutes, or be delayed for hours or even days, and that medications and supportive care will relieve distress or suffering.[15,61] Patients may also choose to discontinue food and water by mouth or gastrostomy; this usually leads to death within 2 weeks.[61–70] In one study nearly all deaths occurred within 15 days of cessation of nutrition and hydration; nurses reported that the median quality of death was 8 on a scale of 0 (very bad death) to 10 (very good death). Reports from nurses and caregivers indicate that individuals who choose this route usually die a 'good death'.[67] Benzodiazepines, opiates, and other medications can control suffering that may occur after withdrawal of life-extending measures. Since death is expected, there is no medical, ethical, or legal justification for withholding the medications necessary to control distressing symptoms. Side-effects may have the unintended effect of hastening death. This is justifiable and referred to as the ethical principle of double effect.[1–3,71] Administration of medication in excess of doses to control symptoms with the intention of quickening death is euthanasia.[1–3]

Case history: withdrawal of food and water

Ms P is a 77-year-old woman with bulbar-onset ALS who is anarthric and unable to eat or drink. She has been using a feeding tube for all nutrition and hydration for the past 6 months. Recently she developed weakness of her arms and hands, limiting her ability to perform activities of daily living, and leg weakness, limiting her ability to walk. Increasing symptoms caused her to move from the independent to the assisted living area of her nursing facility. She is very distressed over her increasing symptoms and waning independence. However, she denies depression. She has no cognitive difficulties.

Seen just before Christmas she writes, 'I am tired of this. I want this to end. Can you help me?'. Asked for clarification, she makes it clear she is asking for assistance in dying. Since her FVC is greater than 70% of predicted and she is receiving food and water via a gastrostomy tube she is not eligible for hospice

care in the United States. Nonetheless all aspects of palliative care including a hospice are presented to her, emphasizing that end-of-life symptoms can be managed when the time comes. She listens attentively but at the end writes that she does not want to wait until she develops even more symptoms. She wishes to end her struggle with ALS quickly. Physician-assisted suicide and euthanasia are legally precluded, and anyway her doctor is morally opposed. Presented with the option of stopping all food and water, and the emphasis that it is unlikely that she will suffer, that symptoms can be managed with hospice care, and it is likely she will die within 2 weeks, she opts to discuss this with her two sons.

One week later her sons call to report that she would like to withdraw food and water, and she starts hospice care at her nursing facility. Daily contact with her neurologist confirms that she is comfortable without complaint. Ms P dies 8 days after stopping food and water. The hospice nurses report death was peaceful. Her sons call to thank her neurologist for honouring their mother's wishes and allowing her to exercise her independence.

Physician-assisted suicide and euthanasia

Physician-assisted suicide (PAS) refers to the provision of a requested lethal dose of medication that the patient uses to end their own life. In PAS, the physician provides the means for death and the patient administers it. PAS is not euthanasia, which refers to a physician both providing and administering a lethal dose of medication. Euthanasia can be voluntary, at the request of the patient, or involuntary, without the patient's consent. PAS should be distinguished from palliative sedation, which refers to sedating a terminally ill, suffering patient to reversible unconsciousness, maintaining sedation over the ensuing days, and allowing the patient to die of the underlying disease, starvation, or dehydration.[3,65,72–78]

Oregon

The Oregon Death with Dignity Act (ODWDA) was passed as a citizens' initiative in 1994 with 51% in favour. Implementation was delayed by legal proceedings, but PAS has been legal in Oregon since 1997.[79]

The ODWDA stipulates that ending one's life within the rules of the act does not constitute suicide, and physicians providing lethal medication to carry out the act are not guilty of a crime. The act defines PAS as the voluntary self-administration of lethal medications prescribed by a physician for that purpose. It specifically prohibits euthanasia. To be eligible to participate patients must have a terminal disease (have less than 6 months to live), be older than

18 years, have been a resident of Oregon for more than 6 months, and be deemed capable of understanding and making the decision. Patients can receive the lethal medication only after two oral requests to their physician within 15 or more days, followed by a written request signed by two witnesses. If the physician has any concern about decision-making capacity or mental illness that may be influencing the decision, then the patient must be referred for psychiatric evaluation. The physician is obliged to discuss feasible alternatives to PAS, with emphasis upon pain control and hospice and palliative care. The physician can request the patient notify his or her next of kin, but the patient is not obliged to do so.[79]

The number of prescriptions given and patients following through with PAS through ODWDA has more than doubled since 1998. However, the number has remained small compared with the total number of deaths in Oregon, with less than 0.1% dying by PAS. ALS patients have been over-represented since PAS was legalized. The 2012 report[80] indicates that five ALS patients ended their life with PAS (49 since 1997). Although the numbers are small, ALS patients were 67 times more likely to use PAS than those with heart disease, the group with the lowest use of PAS.[81]

Over the same period the prescribing physicians for PAS indicated that the three most common end-of-life concerns in patients choosing PAS were loss of autonomy, decreasing ability to participate in enjoyable activities, and loss of dignity. Of 100 ALS patients surveyed before the ODWDA, 56% indicated they would consider PAS: they were more likely to be men, highly educated, have higher hopelessness scores, be less religious, and rate their quality of life as lower.[57] Fifty caregivers of deceased ALS patients from Oregon and Washington indicated that one-third of patients discussed wanting PAS in their last month of life. Those expressing interest in PAS in their last month of life were more likely to have greater hopelessness and expressed interest in PAS earlier in their disease. Compared with patients who did not express interest, those who did had greater distress at being a burden to others, more insomnia, more pain, and more discomfort other than pain.[58] Although many express interest in PAS, few ultimately utilize it.

In 2001 the US Justice Department ruled that assisting suicide was not a legitimate use of drugs controlled by federal drug laws and that the US Food and Drug Administration could stop the prescribing rights of, and potentially jail, any physician who authorized drugs for this purpose. The 9th Circuit Court of Appeals upheld Oregon's law in 2004. The US Supreme Court upheld the 9th Circuit decision and rejected the US Justice Department's case, thereby upholding Oregon's law in 2006. Euthanasia is illegal in the United States. The US Supreme Court ruled there is no constitutional right to assistance in dying.

However, the court left the decision to legalize PAS to each state. Washington and Vermont have followed Oregon in legalizing PAS by statute and Montana has legalized PAS by court decree. The issue continues to be debated in other states.

The Netherlands

The Criminal Code of the Netherlands contains provisions prohibiting the intentional taking of human life. However, termination of life upon request and assistance with suicide has been permitted in the Netherlands by a non-prosecution agreement between the Ministry of Justice and the Royal Dutch Medical Association. In 2001, the Dutch Parliament passed legislation whereby the long-standing practice of terminating life on request, and assistance with suicide, would not be a criminal offence if carried out by a physician fulfilling certain 'criteria of due care'. These stipulate that the physician must:

1 be satisfied that the patient has made a voluntary and well-considered request
2 be satisfied that suffering is unbearable, without prospect of improvement
3 have informed the patient about his or her situation and prospects,
4 have come to the conclusion, with the patient, that there is no reasonable alternative given the patient's situation
5 consult at least one other physician, who must have seen the patient and given a written opinion on the due care criteria referred to above
6 have terminated the patient's life or provided assistance with suicide with due medical care and attention.

The physician is also required to report the assisted death to the municipal coroner.[82]

Treating physicians in the Netherlands reported that 35/231 (17%) ALS patients chose euthanasia and died by that method, 6 (3%) died from PAS, and 48 (24%) received 'palliative treatment that probably shortened their lives'. Those choosing assisted death were more likely to consider religion as unimportant. There was no association with education, income, level of care, or any disease characteristics. ALS patients in this study were 10 times more likely to choose assisted death than those in Oregon and preferred euthanasia over PAS.[83]

Other nations

Belgium legalized euthanasia limited to adults and emancipated minors in 2002.[84] Although under Swiss law PAS is not specifically legal, it does not

punish those who aid someone in committing suicide for unselfish reasons. The Council of Europe rejected a draft resolution supporting assistance in dying.[85,86]

Ethical arguments for and against assisted death

Those in favour of assisted death frequently use the following arguments:

- Justice: Some terminally ill patients hasten death by refusing or terminating treatment. For others, refusal of treatment will not hasten death. For these the only option for hastening death is suicide, and assisted death should be allowed.
- Autonomy: Decisions about the timing and the nature of death are personal. Competent, terminally ill patients should be allowed to choose the time and nature of their death.
- Beneficence: Physicians have a duty to relieve physical or psychological suffering, but this is not always possible. PAS should be allowed in order to relieve terminal unbearable suffering.
- Liberty: The state's interest in preserving life is reduced when a person has a terminal illness and a desire to end suffering by ending their life. Prohibiting PAS is an excessive restriction on personal liberty.
- Honesty: Assisted death is occurring, but is now secret. Legalization would permit an open and honest discussion.

Those opposing assisted death frequently make the following arguments:

- Sanctity of life: There is a long-standing, strong, religious, moral, and secular prohibition against taking human life.
- Killing versus letting die: There is an important distinction between letting patients die of their disease by withholding or terminating care, a passive act, and killing using PAS, an active act.
- Slippery slope: Once legalized, there is no way to contain PAS. People who lack access or means to obtain proper medical care and support could be pushed into PAS; this might be encouraged, even unconsciously, by overburdened family members, and possibly by physicians. Eventually PAS would become a remedy for the suffering of individuals who do not have a terminal disease.
- Harm to medicine/distrust: The ethical traditions of medicine oppose taking of life. If PAS were legalized, the image of medicine would be harmed and the trust implicit in the physician–patient relationship undermined.
- Doctors are fallible: There is inherent uncertainty in diagnosis and prognosis. Legalizing PAS would lead to the death of patients who are, in fact, not suffering from a terminal disorder.

American Academy of Neurology position on assisted death

In 1998 the AAN published a position statement 'vigorously opposing physician-assisted suicide, euthanasia, and any other actions by neurologists directly intended to cause death'. Further, the AAN states that legalization of PAS will not make physician participation 'morally or ethically acceptable ipso facto', and reaffirms its position that 'aggressive and effective palliative care should be provided to all patients who need and desire such care'.[87]

World Medical Association position on assisted death

The World Medical Association (WMA), with members representing medical associations (including the American Medical Association) from 82 countries, has adopted strong resolutions condemning both PAS and euthanasia and urging all national medical associations and physicians to refrain from participating in them even if national law allows or decriminalizes the practices.[88]

The request for assisted death

Any request for assisted death should be handled with respect and compassion. The appeal should be explored to understand the motivation behind it and determine if there are any treatable physical or psychological symptoms driving the request. It is essential to determine if there are misunderstandings about the likely course of ALS, and if the patient has particular fears or concerns. It is helpful to review potential end-of-life scenarios and plans of care, emphasizing that symptoms such as air hunger, anxiety, depression, and pain can be treated, and suffering minimized or eliminated. Most patients derive reassurance and some sense of control from these discussions and no longer seek assisted death.[89]

Despite these efforts, some will persist in their request for PAS. While the AAN and WMA positions make no exceptions for the participation of physicians in PAS, physicians can consider the issue carefully and decide on their own position. Physicians should be advised that there are potential legal and professional consequences to participation in PAS, depending in part on the jurisdiction.

The palliative care community and right to die advocates may appear to be adversarial. However, Bernheim and colleagues describe how palliative care and right to die advocates in Belgium not only coexisted but also felt synergy in shared values, notably of patient autonomy.[90] Prokopetz and Lehmann, acknowledging the increasing endorsement by legislators of PAS, envision legalized assisted suicide not administered by physicians.[91] Many palliative care practitioners feel morally opposed to PAS, however, and must face the ethical challenge of non-abandonment in the face of a patient's choice, where legal.

Careful reflection ahead of time can prepare one to openly discuss one's position with the patient, acknowledging and respecting difference of opinion when it occurs. No physician, however, should feel forced to supply assistance if they are morally opposed to PAS

Alternatives to PAS

ALS patients should be informed that there are legal, and some would argue more ethically justifiable, alternatives to PAS to hasten death. As already discussed, patients can elect to discontinue life-extending measures while medications are used to control suffering and distress. Intractable physical and or emotional suffering close to death can be managed with palliative sedation.[84–87]

Despite extensive ethical, scholarly, and public discussion, confusion about the administration of medication for comfort at end of life, and particularly palliative sedation and withdrawal from mechanical ventilation, persists. An online survey of members of a US palliative care organization found that 25 (4%) of 643 respondents had been formally investigated following the death of a patient in whom they had no intent to hasten death.[92] The principle of double effect has been used to justify many practices, prompting a call to anchor practice in the original precepts, i.e. recognizing as ethical the practice of pursuing relief of suffering despite foreseeable, unintended consequences, such as hasted death from palliative sedation for intractable pain.[93] Understanding the principle and applying it judiciously can mitigate a charge of hastening death, and even then actions may be misunderstood.

Conclusions

There are many ethical issues to be considered in the care of people with ALS. It is essential that these issues be discussed at the appropriate time and with sensitivity—allowing for the differing cultural values and attitudes in society, with respect for different social and cultural groupings and individual families. The wider multidisciplinary team will also need to consider and discuss these issues and in this way enable patients with ALS, their families, and professionals to come to decisions that are acceptable to all.

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Chapter 18

End-of-life care in ALS

Nigel Sykes

Summary

The care of a person with ALS at the end of life will need particular attention and careful management. As death approaches for a person with ALS it essential that the person with ALS, his or her family and the professional team are aware of the changes and that there is proactive planning for the final stages of the disease. This may include advance care planning, provision of medication, so that symptoms can be managed effectively, and good communication amongst all involved. It is also important to support not only the patient and family but the professional carers. Many people with ALS fear a distressing death, but most are able to die without distress with appropriate palliative care and support for all those involved in the patient's care.

Case history

Jack is a 75-year-old man with a 2-year history of limb-onset ALS. By the time he was admitted to the hospice to give his family a period of respite he needed assistance to stand and was beginning to have problems with swallowing. He was, however, disinclined to accept a gastrostomy.

The admission was extended because Jack and his family decided that continued care at home was now too difficult and so a nursing home placement was sought. Over this time his swallowing was deteriorating, although he remained able to take thickened fluids, without evidence of aspiration. On the day before his death he was noted to be generally less well on waking, complaining of widespread discomfort throughout his body, and of intermittent breathlessness. It was agreed with him that nursing home transfer, which had been imminent, should be deferred.

Jack accepted administration of medication subcutaneously via a syringe driver, and this was set up at midday with morphine sulphate 7.5 mg/24 h, combined with glycopyrronium bromide 100 μg/24 h. His speech became less distinct during the course of the day but he remained able to make himself understood by expression, if not always by words. That evening Jack settled to sleep at his usual time. At 4.30 the next morning he was noted by nursing staff to be peaceful but breathing shallowly. Five minutes later Jack's breathing ceased and he died.

Recognizing the terminal phase

Most people with ALS die as a result of respiratory failure.[1] Consequently, the trajectory of decline in respiratory function is the key to anticipating the final stage of the illness. However, the onset of this stage, when it comes, is often rapid and ALS is a cause of sudden death. In a series of 124 ALS patients cared for until death, 40% deteriorated suddenly and died within 12 h; a further 18% had died within 24 h of a change in their condition first being noticed.[2] This rapidity has implications for the preparations the caring team must make in order to respond quickly to symptom changes and, particularly, the guidance they must give patients' families in order to prepare them for what may appear to be a catastrophically sudden demise.

Alternatively, the character of end-of-life ALS care can be altered completely by the use of assisted ventilation in the presence of gastrostomy feeding. It has become clear that NIV can extend life significantly[3] and that tracheostomy ventilation can do so for considerable periods, to the extent that functional deterioration continues to the point of a locked-in state. The timing and nature of the terminal event are then the result of physician action in deciding when and how ventilator withdrawal should be achieved (see Chapter 6).

If death occurs suddenly in ALS it seems most often to happen at night, and this may be a result of an exacerbation of nocturnal hypoventilation. Where there are clinical changes which herald the terminal phase of the illness, these usually take the form of an increase in the sense of breathlessness, even in the face of NIV and despite adjustments to the ventilator settings, or the onset of a reduced level of consciousness associated with diminished respiratory effort. Changes in medication can ease the distress associated with breathlessness, but as respiration fails, transition is into loss of awareness and ultimately a state of unrousability. At this point the patient's condition is not significantly different from that seen in the end stage of cancer or other terminal diseases, but in ALS the duration of this period is liable to be shorter than that seen elsewhere. There may also be a sudden deterioration from probable cardiac causes, related to autonomic dysfunction.

It seems that although NIV delays the onset of the terminal phase, in most cases it does not greatly alter its character or duration. On the other hand, a minority of patients appear to be sustained in a condition of virtual unrousability over several days. This situation can be a cause of much distress to both the family and staff, who perceive it as a pointless dragging out of the dying process and an affront to the dignity of the person concerned. After careful discussion between those involved, agreement may be reached that the most appropriate response is to terminate the ventilation, after which death ensues in a short while.

Supporting the patient

Quality of life may be seen as reflective of the degree of congruence between hopes and expectations and the reality of the person's achievement.[4] If there is a serious mismatch between activities a person regards as their principal source of satisfaction and their ability actually to pursue those activities, the results are frustration and a sense of meaninglessness. It is the role of carers to work to help the patient to adjust their horizons and focus to achieve enjoyment and a sense of worth out of activities which may be new or may previously have been discounted but which still lie within the patient's capabilities. This type of support is a major contribution of hospice day units and creative living facilities. Naturally the range of such activities becomes increasingly limited over time, but this aspect of care remains important right up to the onset of the terminal phase of the illness. It answers not only to the psychological needs of the individual but probably also the social and certainly the spiritual as well.

'Spiritual' signifies the need to find within one's present existence a sense of meaning. How this is defined is a matter for each person. It may, but often does not, involve a framework of religion and the notion of a God with whom it is possible to have a relationship. If religion is an important feature, the participation of clergy might be appropriate, but a patient might also wish to talk to a carer about 'spiritual' matters and carers should be prepared for this. It appears that certainty, whether in the existence or the non-existence of God, is associated with the lowest levels of anxiety in dying people.[5] For those who are uncertain there may be a need to talk through feelings of anger against God for a condition as dreadful as this, or apprehension about long-suppressed ideas of judgement.

If religion does not provide a context for meaning this must arise from a person's sense of relationship to others, the fulfilment they derive from favoured activities, and their perception of personal autonomy and control. All three of these factors can be severely impaired by ALS, and as the disease progresses it is a particular challenge for carers—professional, family, and friends—to demonstrate that the ill person still retains a valued place in a social setting, to allow them the maximum degree of personal control practicable, and to enable them to adjust their sources of satisfaction. Not everyone has the same ability to make these adjustments and the compromises necessary to coexist with a chronic, debilitating condition. From the lack of such reconciliations, from unresolved personal issues, and from the awareness of present losses with more to come, both in functions and relationships, comes the suffering which is inevitably a part of any terminal illness.

Only individuals themselves can achieve a resolution of their suffering. It is not amenable to medical therapeutics or nursing interventions. Nevertheless, the caring environment can be such as to either facilitate or hinder someone's progress in reaching a resolution of suffering. It is for this reason especially that the framework of care for patient and family must be a multidisciplinary team. No single discipline is adequate to meet the whole need, but the mutually complementary skills of professionals who respect and communicate with each other provide the best resource to enable the person with ALS to approach the end of their life with dignity and tranquillity.

ALS imposes huge burdens on those who have it, and on their carers, through the relentless increase in disability it brings about. The extent of non-respiratory disability is not necessarily a reliable guide to prognosis, especially in younger people, but it is claimed that the requirement for physical aids and adaptations marks approximately the half-way point of the disease course, with gastrostomy feeding at about 80% and ventilatory support at 80–90%.[6] While the ability of so many with ALS to adapt so remarkably to the ravages of their condition is a source of admiration to those close to them, the sense of physical disintegration, the loss of control, and of a great deal of what previously made life worth living mean that death is not necessarily seen as unwelcome by patients in the late stages of the disease.

It can be a consolation to someone greatly disabled by ALS, and whose chest function is deteriorating, to be told that their condition will not go on for much longer. An exact prognosis is impossible to judge, though, and if a time is asked for the answer has to be relatively imprecise. Simply to be told that time appears short is not enough without an assurance of continued support and symptom control, because there is also the issue of continued hope.

Some professionals find it very difficult to be asked by patients for an estimate of prognosis, not only because any attempt at accuracy is likely to be misplaced but also because they fear that the answer will cause the person to give up. Relatives are even more likely to take this view. There is evidence that the grounds for hope change as the illness progresses.[7] Because of its inexorable nature this transition is perhaps less pronounced in ALS than in other conditions encountered in palliative care, but there may still be a change from hopes that the disease may be slower in its progression or relatively limited in its scope or that it may be retarded by a therapeutic intervention, to the hope for relief of discomfort and for a peaceful end to life.

For any ill person to achieve a maximum degree of comfort they must feel esteemed and understood by their carers and confident in the ability of the professionals involved to respond appropriately to their needs for information

and physical care. Esteem implies a personal warmth and trustworthiness on the part of caring staff that is more than a professional façade yet not an inappropriate emotional closeness. True understanding of the patient's situation can often be particularly hard to gain in ALS because of the severe degree of disability and, especially, the impairment of communication. The fullest use of communication aids is required, together with all the skills of empathy and patience the carer possesses. Empathy is the imaginative entering into the other person's situation, but not the premature and superficial assurance that 'I understand how you feel', which distances and is the enemy of trust. Continuity of professional carers (without a burdensome reliance on a single 'favoured' individual) is often important in facilitating communication in advanced ALS, as much depends on knowledge of personality, facial expression, and patterns of speech.

Consciousness that death is getting nearer can induce a desire for resolution of outstanding personal conflicts, perhaps of long standing, or for the tidying of personal affairs. Even for someone with intact speech it can be difficult to broach these topics and request help. There needs to be sensitivity on the part of the caring team to the possibility that such issues might lie behind an appearance of restlessness or anxiety, and a readiness to probe gently for the existence of 'unfinished business'. Appropriate help can make a radical difference to the closing days or weeks of a patient's life. Conversely, not everyone can identify, share, or resolve issues like this, and the resulting anxiety can be relieved only by pharmacological means.

It should be remembered that any persistent discomfort makes any other discomfort worse and is also mentally wearing. Hence attention to detail is important in symptom control, so that by the time end-of-life care dawns a capacity for competent symptom control should have been demonstrated and maintained. There is much in ALS which cannot be fully alleviated, but patients and families usually accept that there are limits to what medicine can achieve as long as the doctor shows a commitment to stay alongside them and keep trying. Whatever the problems earlier on, it should be possible for the promise to be made that at the end distress can be controlled and that death need be neither painful nor frightening. Provided that the correct drugs are available, and are administered in the right doses and combinations, the promise can be fulfilled.

Despite this, some people with ALS still receive the information that they will experience increasingly severe pain leading to a death from choking. This is a terrifying prospect which has no basis in truth. Uncontrolled severe pain is exceedingly rare. Although choking sensations are indeed common in bulbar

ALS, choking is not the cause or the context of death. In O'Brien's series only one person was thought to have died in this way, and even then post-mortem examination showed that in fact the airways were clear.[2] A more recent multicentre study of ALS patients at the end of life found no instance of death through choking and 98% of the group were perceived to have had peaceful deaths.[8] Unfortunately in Britain the association between choking and ALS has become established by repeated misinformation in the media, requiring both an active approach to symptom control by doctors, nurses, and physiotherapists, and continual reassurance if fears are to be allayed. Accurate information provided at the right time can be a great help in a person's ability to cope with the advance of disease and, particularly, with the prospect of its end.

Advance care planning has increasingly become a key means of supporting both patients facing life-shortening illness and their family. This offers an opportunity, which may be spread over several meetings and quite a long period, for the person with ALS to learn what they wish to know about their condition and to express their preferences about how they wish to be looked after, where, and by whom. Advance care planning is meant to improve healthcare outcomes by making sure that decision-making is shared between patients and professionals. It also enables clinical care to continue to fit with a patient's informed decisions and preferences even after independent decisions are no longer possible, something that is particularly relevant to ALS in view of the communication and cognitive difficulties that often arise in its later stages.

This is important not only for patients, who can feel that their personal views are known and will be respected, and clinicians, but also for families, who can be relieved of the burden of feeling that they have to make potentially serious decisions about, for example, interventions such as ventilation or a gastrostomy placement, on behalf of the patient without guidance. At the same time, the process can uncover disparities in hopes and expectations between patient and family that then have a chance of being resolved before they cause disharmony or practical difficulty.

It is helpful if the results of the advance care planning process are set down in writing and made accessible to others who are involved, professionals and family or friends as appropriate. However, the dialogue may be recorded in sound or video instead, or its outcomes put down in the health record. In England this is known as a 'statement of wishes and preferences' that, although not legally binding, has been promoted as a valuable tool to guide and improve end-of-life care.[9] However, with recognition by legal frameworks such as the Mental Capacity Act 2005 in England and the Patient Self-Determination Act 1991 in the United States, advance care planning can result in more binding outcomes

in the form of either an instructional directive, otherwise known as a 'living will' or an 'advance decision to refuse treatment', or through the appointment of a person as proxy or attorney who is empowered to make decisions about medical treatment on behalf of the patient should the patient lose the capacity to decide these matters for themselves. Disease-specific advance decision templates have been produced for ALS.

Supporting the family

By family we mean those who are closest and most significant for the patient, whether they be relatives or friends. It is important, although not always easy, to identify who these are. This is not accomplished simply by obtaining details of the next of kin. It is a great help to construct a genogram, or family tree, which allows the professional team to see at a glance who constitutes the family, where they are, and what other difficulties the family might be enduring at the moment.[10] Around the edge of the genogram should be included particular friends and 'significant others' including—sometimes—pets. Ideally this should have been done well before the terminal phase of the illness.

Being close to someone with ALS is enormously stressful. The family may be at greater risk of depression and social isolation than the patient[11] and may score lower on non-physical components of quality of life measurement.[12] The progressive functional deterioration is apparent to all, but family members may not recognize how this is linked with shortening of the prognosis. In particular they may not realize that death from ALS can be sudden. When the end of life is considered, thoughts may be coloured by unfounded apprehension about symptoms such as suffocation, choking, and pain. A family's need for accurate information is at least as great as that of the patient.

The paralysis and loss of speech associated with ALS engender a feeling of impotence in all who care for those with it. This experience, which can be uncomfortable even for knowledgeable professionals, may be disabling and extremely distressing for lay carers. Without guidance as to what to expect as swallowing and respiration deteriorate, and how they can gain urgent help in a crisis, an episode of choking or breathlessness can be terrifying not only to the patient but also the family. In countries where tracheostomy ventilation is often used for ALS a significant proportion of people with ALS who receive it do so without prior discussion of the procedure, because it was initiated in the midst of a respiratory crisis to which the response had been to take the patient to the hospital emergency room. The same sequence of events occurs occasionally in Britain. This is not the way to begin such an invasive technique with so many grave implications. Prior involvement of a multiprofessional team with 24-h

availability can facilitate a process of advance care planning so that informed choices are made in anticipation of the event, and be a resource for advice and practical help in an emergency. Such access has been stipulated in the UK by NICE in their guidance on palliative care[13] and, complementary to this document, the wider availability of specialist palliative care to neurological patients has been recommended in the National Service Framework for Long-Term Conditions issued by the UK Department of Health.[14] A properly prepared written advance directive can also do much to ensure that appropriate action is taken in an emergency.

A multidisciplinary palliative care team can also facilitate the continuing care of the patient at home, where it has long been known that most terminally ill people wish to be.[15] As life becomes more difficult, the proportion of people wishing to stay at home diminishes but still remains at about 50%.[16] Despite assistance from community nursing services, the brunt of home care is borne by the family. Respite admissions to a hospice, hospital, or nursing home can help carers to recuperate and resume their task, or additional respite nursing help may be available for limited periods, but for a significant number of families there comes a point when they feel they can no longer look after the patient at home. The patient may agree with this, or there may be divergence.

Even if all are in agreement that admission is needed, family members may still be left with a sense of failure and of guilt that they have let their relative down. This may be especially marked if the person with ALS dies soon after they have been admitted, leading to feelings that 'if only we had kept going that little bit longer we could have looked after him/her to the end'. It is important for their response in bereavement that families receive reassurance about the quality of their caring efforts prior to the admission, and the appropriateness of seeking in-patient care at that point. It is also important that there are the facilities and encouragement to enable them to remain with the patient as death approaches, if that would be helpful to them.

For some families it is important that their relative does *not* die in their home, because of the memories that would leave for the future. Part of advance care planning is to have conversations with patient and family about the preferred place of death if at all possible, so that in the case of difference there can be discussion and understanding, if not always agreement, and plans made so that admission does not occur as an acute event in response to the distress of a family who have never been able to reveal their concerns about end-of-life care of their relative. Even when death does take place in a hospice or other inpatient unit, usually at least 80% of the patient's last year of life will have been spent at home, and afterwards the perception of families is that they have indeed cared for the person themselves.[16]

Most bereaved people do not need specific bereavement care: they will work through the feelings of loss, which are at some time part of every individual's experience, by themselves with the support of their own network of family and friends. For a minority, perhaps up to 25%, their adjustment can be helped by specialist bereavement support. This is increasingly widely available, and in the UK can be accessed through Cruse Bereavement Care (<http://www.cruse.org.uk/>) or the local hospice or social service department (see Chapter 20).

Supporting the professionals

Even for professionals committed to palliative care, ALS is perhaps particularly prone to engendering a pervasive sense of helplessness and failure. The difficulties in communication are a considerable contributory factor, as is the prominence of an irreversible process of functional deterioration which may be in contrast to the patient's continued mental vigour—inwardly they seem 'well' but their bodies inexorably weaken in a way the professionals can do little about. This leads to feelings of frustration and de-skilling that can result in avoidance of the patient by carers unless the problem is recognized.

The unpressured process of advance care planning required if end-of-life care is going to go as well as possible both for patient and family needs the clinicians involved to recognize well ahead of time the eventual outcome of the disease, before it becomes too late to uncover and allay wishes and fears about the dying process and about where and how it should be handled. While the conversation has to go at the patient's pace and be directed by his or her concerns it is vital that the professionals give the patient the opportunity to be aware of, and to explore, key options that may become available, such as gastrostomy insertion or NIV. These topics will arise out of conversations about the likely progression of the neurological symptoms, conversations that over time can lead naturally to a consideration of the end of life, what it will be like, and how it can be handled. People vary in how much they are able to participate in such conversations and how fast the dialogue can go (which also depends on the rate of deterioration of the particular condition). But the other important variable is the skill of the professional in terms of their ability to communicate clearly and empathetically, their knowledge of the illness, and their personal readiness to converse about issues of mortality that may resonate for them personally. Without these types of skill, the ability of staff to help patients prepare for the end of their life will be compromised.

To some extent these tensions are eased in the terminal phase, as the end comes into sight and the situation comes to resemble more closely that of people dying of cancer and other conditions. Afterwards, however, staff memories

of the case may be influenced by feelings which persist from earlier stages of care. It is likely that they will have known an ALS patient for longer than the average for palliative care patients as a whole, and so the sense of personal loss might be stronger. These factors risk leaving a residue of distress which might impair professionals' abilities to look after people with ALS in the future. The situation is worsened by the relative rarity of ALS, which means that staff can find it difficult to gain and maintain confidence in their expertise in caring for those who have this condition.

Therefore it can be helpful if there is a programme of staff education covering the management of ALS. Better integration and the extension of joint working between specialists in neurology, palliative care, and neuro-rehabilitation will also facilitate the exchange of expertise and ensure that the right skills are available for the changing needs as they arise. Afterwards it can be useful for staff to meet as a multiprofessional group for the sharing of views on how the care of a particular patient went. This session may well identify ways in which care could be improved in the future, but should in addition be a chance for congratulation on the ways in which things have gone well. It should be a managerial responsibility to identify staff members who have particular problems resulting from the experience of care and, without imparting a sense of inadequacy, enable them to talk through the important issues confidentially either on the ward or with an independent counsellor.

Symptom control

The crucial issue in symptom control at the end of life is preparedness. It is a wholly inadequate response to the onset of a distressing symptom if control has to wait on a doctor's order or the pharmacist's acquisition of the medication required. As respiratory capacity shows evidence of serious decline a stock of key drugs should be made available on the ward or in the home ready for use. If the MNDA Just in Case Kit is in use it is likely that some of the medication will already be available. The following categories of drug should be to hand:

- opioids
- relaxants
- anticholinergic agents.

If the drugs are to be used at home, it is sometimes possible for family members to administer them in a crisis via the gastrostomy, rectally, or as buccal preparations. Whether or not the family is willing to assume this responsibility following adequate guidance, locally agreed paperwork must be filled out to enable community nurses to give the drugs when needed.

Opioids

Morphine is effective in the management of pain, breathlessness, and nocturnal discomfort in ALS long before the terminal phase of the illness.[2] Doses are widely variable and not usually high, with a median of 60 mg/day being reported.[17] Even in its use for dyspnoea there is no evidence that morphine shortens life when it is used competently. Oliver et al.'s study found a median length of use of 51 days and a maximum of 970 days.[18] Dose stability has been noted over periods of years and it is not inevitable that dose escalation will be required in the terminal phase. However, the morphine dose may have to rise if new symptoms intervene or old symptoms worsen, in particular if breathlessness increases. In this case the dose should be titrated upward in the usual way, making increments of 20–50% of the preceding dose level in order to provide a noticeable therapeutic effect without causing excessive drowsiness. If morphine has not been used before it is appropriate to commence at 5–10 mg 4-hourly for pain, orally or by gastrostomy, or 2.5–5 mg 4-hourly for breathlessness.

If a gastrostomy is in situ the administration of medication can continue as before, but without it many patients will require a change of route from oral to parenteral. This can be accomplished by subcutaneous injections (made less uncomfortable if a plastic cannula is left in place to avoid repeated needle sticks) or more conveniently, if the length of the prognosis appears to justify it, by a subcutaneous infusion delivered by a portable syringe driver. Alternatively, morphine or oxycodone can be given rectally, fentanyl and buprenorphine (a partial opioid agonist) are available as transdermal patches, and fentanyl can also be administered sublingually.

An important consideration if there is a change of route of an existing opioid is to make an appropriate calculation of the equivalent dose, as shown in Table 18.1. All such ratios are subject to individual variation and resulting doses may require upward or downward adjustment according to response. Unless the opioid requirement is stable, transdermal fentanyl, although convenient to give, is a less satisfactory choice of parenteral opioid as dose titration is relatively difficult owing to the wide steps between patch sizes and the long delay (up to 23 h) in achieving and recovering from steady-state blood levels.[19]

A patient who has gained particular analgesic benefit from a NSAID can have it continued by suppository (e.g. in the form of naproxen or ketoprofen) or syringe driver. Ketorolac will mix in a syringe with diamorphine, but if, as is usually the case, other drugs are also required it is more reliable to use a separate syringe driver for it or to give an NSAID rectally.

Table 18.1 Conversion ratios from oral opioids to subcutaneous morphine

Oral opioid	To obtain dose of subcutaneous morphine divide by
Morphine	2
Tramadol	20
Codeine	20
Oxycodone	1
Hydromorphone	Multiply by 4

Whether in cancer or ALS, pain is not generally a problem at the end of life if it has been adequately controlled previously. In a patient who can no longer indicate their feelings, carers interpret non-verbal signs of distress, for instance groaning, grimacing, or restlessness. Before increasing medication, it should be checked whether there are remediable causes of discomfort, particularly a full bladder or rectum.

Relaxants

A generalized restlessness, which may be due to pain but also to anxiety, has to be distinguished from focal myoclonic jerks as these may be worsened by opioids, especially in the presence of phenothiazines. Rather than persist with opioid medication for restlessness it is more appropriate to use a benzodiazepine, either instead of or in addition, for its anxiolytic and muscle relaxant properties. This may take the form of diazepam suppositories rectally or liquid via a gastrostomy, given as necessary or two or three times a day, or midazolam subcutaneously. Midazolam combines satisfactorily with morphine or anticholinergic agents in a syringe driver. An initial midazolam dose is 2.5 mg immediately or 10 mg/24 h.

In ALS a principal role of benzodiazepines is in the control of breathlessness, where their action is complementary to that of opioids. For a sudden, and potentially distressing, episode of breathlessness lorazepam tablets may be given sublingually or midazolam buccally—these can be administered by family or carers if professional help is not readily available. However, in an acute deterioration in breathing associated with failing respiratory function it is appropriate to give a combination of morphine (or other opioid) and diazepam/midazolam initially and then to titrate the doses of each. A phenothiazine can be used instead of the benzodiazepine, e.g. chlorpromazine or the more sedating levomepromazine. Both have an antiemetic effect, if this is important, and there is limited evidence that chlorpromazine can palliate breathlessness.[20] Watch should be kept for myoclonic jerking, but the lowering of the seizure

threshold that these drugs induce is not a problem in end-of-life care in ALS. Levomepromazine can be given by subcutaneous infusion, but chlorpromazine causes too many skin reactions to be given by this route.

Contrary to popular impression, it is rare for good symptom control to shorten life. An exception can be the relief of frightening breathlessness in a patient with severe respiratory compromise, in whom any alleviation of their distressing anxiety may result in an inadequate respiratory effort. To anyone in the presence of such a disabled person the imperative to relieve their anguish is clear, even if this might result (and it is never definite that it will) in a shortening of their already extremely short prognosis. The ethical probity of this stance is endorsed by medical authorities in both the UK and the United States, and by the Roman Catholic Church.[21] The doses of drugs used must, of course, be proportionate to the patient's current regime.

Midazolam is also an appropriate choice of drug when it is agreed by the patient (if still conscious and capable of making such a decision), family, and professionals that life is now being prolonged by assisted ventilation to a degree that is intolerable. An immediate subcutaneous dose of 2.5–5 mg (1–2 mg IV), with 5–10 mg subcutaneous morphine (2–10 mg IV), should be given—or more if the patient is already receiving regular sedation or opioids. This should be supplemented by an anticholinergic agent if there have been difficulties with retention of secretions. A continuous infusion of about 50% of the bolus doses per hour should be set up and adjusted to achieve the required degree of comfort and sedation. The use of these drugs is also indicated if the patient appears comatose in order to guard against the onset of distress once ventilator settings are changed. Further bolus doses should be available throughout the procedure in case any distress occurs. This is not the place to discuss in detail the technique of ventilator withdrawal, but a process of weaning over 30 to 60 min allows prompt control of any distress that might appear after each adjustment of the ventilator without being unduly prolonged[22] (see Chapter 6).

Antisecretory drugs

Owing to bulbar weakness, retention of secretions in the upper airways is a frequent problem in ALS long before the terminal phase. However, any severely ill patient with reduced ability to cough can accumulate secretions in the upper airways, resulting in noisy breathing which if not always a distress to the patient may well be to attending relatives. At the end of life this problem is one that is best anticipated, as it is not easy to get rid of secretions which have already gathered. If respiratory failure is following on from unsuccessful treatment of a chest infection there are likely also to be purulent exudates present which

cannot be prevented, so that it is not possible to stop rattly breathing altogether. Hence the first step in management is to explain to the patient's family the mechanism of the noisy breathing, what is being done, and what its limitations are, and reassure them that by this stage of the illness the dying person is unlikely to be nearly as aware of the sounds as they are themselves.

If an anticholinergic agent is already being given by gastrostomy it can be continued and the dose increased if retention of secretions is worsening. Atropine tends to be arousing, though, and should dose titration be required it would at this stage be more appropriate to change to a more neutral drug, such as hyoscine butyl bromide, glycopyrronium bromide, or hysocine hydrobromide, which is normally sedating. Any of these can be given subcutaneously by syringe driver in combination with diamorphine and midazolam or levomepromazine. Dose ranges are:

- Hyoscine butylbromide: 20 mg subcutaneously immediately; 60–240 mg/24 h by subcutaneous infusion.*
- Glycopyrronium bromide: 0.2–0.4 mg subcutaneously immediately; 0.6–1.2 mg/24 h by subcutaneous infusion.
- Hysocine hydrobromide: 0.4–0.8 mg subcutaneously immediately; 1.2–2.4 mg/24 h by subcutaneous infusion.

Hyoscine butylbromide is very poorly absorbed enterally; the other drugs can be given in their injectable form via a gastrostomy. None has a licence for this pattern of use nor, in common with most drugs used in palliative care, for use in a syringe driver.

Conclusion

Care at the end of life is a crucial part of palliative care because it provides some very powerful memories for those who are left behind. The future attitudes of the patient's family and friends towards severe illness in themselves or others, and towards death itself, will be moulded by this experience, which in Western countries today is rare for anyone below advanced middle age. Good symptom control and good communication with the patient, for as long as this is possible, and with the family are crucial not only for the direct benefit they bring to the person with ALS who is dying but also as a public health measure for all those left behind who have been close to that person.

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Chapter 19

Bereavement

Joy Kelly

Summary

The bereavement journey of each individual is incomparable and unique. Through exploration of bereavement models there can be some understanding of this inevitable experience. This understanding will ensure that the appropriate support is offered to the bereaved person when the time is right. In bereavement from ALS, it may be that time spent in anticipatory grief impacts upon recovery time. It is therefore vital that support workers/counsellors are skilled in understanding and working with anticipatory loss. Adults and children need a wide and varying range of support in their loss, and time is needed to fully understand what this means for each individual.

Case history

Judith was a 47-year-old woman, married to John and with two daughters aged 14 and 16. She developed problems with walking, and after several months a diagnosis of ALS was made. Over the next year her walking deteriorated further and she became restricted to a wheelchair. There was a need to organize a bedroom downstairs and she needed carers to come into her home three times a day. Judith's daughters took on more responsibilities for household tasks to enable her husband to continue to work, which was important to him and enabled him to continue to support the family financially. He spent the evenings and weekends caring for his wife and helping to support his daughters. As Judith continued to deteriorate, the specialist social worker from the family and carers support team met regularly with the family, providing an opportunity to discuss the impact of the illness on them as individuals and as a family. The daughters were able to talk of the frustration that they were less able to go out in the evenings and shared their deep sadness about their mother's imminent death. After Judith's death, the social workers and a counsellor from the bereavement team continued to see both John and the daughters separately. This provided an opportunity to discuss their grief and make important realistic memories of life before and after the illness. As such they were supported through their bereavement experience and enabled to make new connections and access useful networks of support.

Introduction

A brave heart and a strong mind can triumph even in the frailest of stricken bodies[1]

Bereavements form ALS, as with other life-limiting conditions, often begins at diagnosis and continues into readjustment post-death. Over this time there will be many losses—some shared and others individual—for the whole family. Each person, to a varying degree, will face psychological, physical, emotional, social, and spiritual challenges. This can be a harrowing journey, as with each loss, resilience and resources will be called upon. Sadly, at present, there is only one outcome for the family and this knowledge also impacts upon the coping mechanisms of those caring for a loved one.

Various theories/models of grief that can be connected with the ALS carer's experience will be explored, and using these we shall look at appropriate ways to support adults and children in their bereavement.

Anticipatory grief

The concept of anticipatory grief was explored at a deep level in the early 1970s. The term is used to explain 'the premature emotional experience of people who face impending death and can include both the dying person and family members'.[2]

Emotional and psychological pain at the diagnosis of a life-limiting condition is to be expected, considering the impact upon the patient and family. This distress can lead to a re-evaluation of spiritual and religious beliefs and also uncertainty about the person's place in the world. Access to a spiritual counsellor or another worker sensitive to this aspect is important in this early stage of the journey (see Chapter 11).

For many, the awareness of coming loss often leads to anxiety and anger. Siegel[3] suggests a sequence of three responses: disbelief, dysphoria, and adaptation. The initial disbelief at diagnosis moves to general feelings of being unwell and sad, and finally through to adjusting to the reality of the condition. This adjustment and acceptance, as with bereavement, take time. The opportunity to explore concerns with an experienced practitioner will ease this stage and perhaps will allow early talk around death to begin. The patient may wish to consider unfinished business in his or her life and look at relationships with family and friends. For others involved with the patient, their needs may begin on a more practical level—perhaps their concerns may revolve around medical care, equipment, and financial concerns. Often family members will keep themselves busy to help cope with their fears. A watchful support worker will recognize this and encourage holistic explorations of needs.

There has been little research evaluating family distress, particularly that which includes children and parents. Adult children in particular may appear to experience little distress. The demands of their own families may shield them from the full impact of the situation. They may also have become adept at masking their worries from their parents and professionals so that their distress remains unseen.

Kissane and Bloch[4] conducted research involving comparative analysis of symptoms of distress between patients, spouses, and children. They demonstrated that a large number of adult children were experiencing a significant level of distress. The Brief Symptom Inventory was used to look at hostility levels in these adults, and it became apparent that adult children were revealing a high measure of hostility when compared with patients and spouses. This could be a manifestation of anticipatory grief as these family members work through a sense of injustice and considerations around the coming loss of a parent. Young children within the family will demonstrate their distress in different ways.

Caring for a dying loved one is exhausting and stressful. The prime carer may experience:

- Feelings of isolation—having to put their life on hold to care for another. Often the 'social circle' becomes smaller as friends drift away. There may be emotional withdrawal from the loved one which changes the relationship between them. It is not uncommon for the carer to wish the loved one dead.[5]
- Resentment toward the loved one for needing care, and perhaps anger and frustration at other family members for lack (or perceived lack) of support. These feelings may also be directed at professionals involved with care.
- Guilt—carers often question whether they are doing enough or whether they could be making better choices for their loved one. There may also be guilt around unexpressed feelings towards the person they are caring for.
- Helplessness in the face of many life losses and alongside the increasing demands as the loved one deteriorates.

As Ronnie, a grandson of an ALS patient said: 'With every clinic visit, the time we thought we had with her got shorter and shorter, like it was being stolen from us . . . '.

For some, receiving a diagnosis late in the disease course shortens the time for adjusting to loss. It can seem that everything moves too fast and feels out of control. This can catch, and keep, family members in the 'shock and disbelief' phase, which continues until the death of the loved one. Palliative care research offers some evidence that there may be some negative impact emotionally and physically on carers where anticipation of death exceeds 6 months.[6] Distress sends shockwaves throughout the family, and appropriate social support can help alleviate worries. This may be through professional teams or extended

family and friends. It should not be assumed that all families will want or need to become involved with 'the organization' initially. This may come later on as the disease progresses and the family feels they can no longer cope alone.

Younger children's experience in anticipatory grief is different from that of the adults in the family. Whilst adults are forward planning and carrying out daily activities, children are left dealing with all the changes they are experiencing right then.[7] Parents are possibly distracted and vulnerable and the mood in the household may be low and sad. The weight of coming loss may already be changing relationships within the family. Children may respond to this by withdrawing, demonstrating challenging behaviour/anger, or seeming to grow up overnight. All of these things show that the child is grieving. There needs to be some normality in the child's world. Parents should make schools aware so that their support network can grow as their needs increase.[7,8]

Children should not be offered false hope. Their questions should be answered in an age-appropriate way. Conversations need to end with reassurance that their needs will be met. Booklets, such as that from the UK MNDA, offer useful material to look at as preparation for parental loss.[9]

Emotional lability, with uncontrollable laughter or crying, is a particular problem for many ALS patients. Communication difficulties may become exacerbated[10] with patients and family members withdrawing from their usual forms of social network support: for example, as Lorraine, aged 50, said in a counselling session, 'I loved to read to my grandson and now he can't understand me'.

The UK MNDA issues a quarterly magazine 'Thumbprint' offering support, advice and contact for patients and carers,[11] and Build-UK.net (<http://www.build-uk.net/>) is a safe chat forum site that ALS patients and families can use for advice and social networking.

Following the death of the patient, support with family adjustment can often be through religious and cultural groups.[12] Such groups offer individual support as well as shared experience and will often continue this support long into bereavement when feelings of isolation can resurface. It is difficult to gauge when support for individual family members may be needed, therefore the 'umbrella' approach, which embraces the whole family, can be the best form of support for many families in their grief.

Models of grief

Theoretical models that help to explain the process of grief will now be explored. These models offer a way for each individual, in their grief, to begin to understand the process and meaning of the journey of loss and the varying responses.[13] Naturally, one model does not fit all, but such models can often help the bereaved

person to begin to understand their experience. Each model, whether traditional or modern, will consider social, economic factors, personality, and coping styles. This is useful for the professional planning an assessment for the bereaved to ensure that the support offered is holistic and appropriate to their needs.

Traditional models

These models evolved following the considerations of Freud[14] and the work of Bowlby on attachment some 60 years later.[15] It was suggested that individuals grieve because they have lost someone from a significant attachment. To allow them to cope with this loss and separation they would need to resolve the break in attachment to the deceased.

These traditional theories are more commonly recognized as 'phases/stages' models. Some more traditional practitioners will use these models as a way of tracking the progress of the bereaved through their grief journey, or perhaps as a way to demonstrate significant movement to a client.

The research of Kubler-Ross[16] moved the theories toward a five-stage model for understanding loss both before death and in bereavement. As Kubler-Ross and Kessler[17] suggest, in long-term diseases like ALS, MS, or Alzheimer's we may be losing our loved one so gradually that there is time to experience all five stages over a period of years. The five stages—denial, anger, bargaining, depression, and acceptance—are not meant to be taken as a linear progression, nor as a 'fit for all'.[17] They are rather to be considered as part of a framework that supports the bereaved in learning to live without a loved one.

Worden[18] suggested that grief is a normal process that could be resolved through 'tasks of mourning'. The tasks were originally as follows:

- Task 1. To accept the reality of the loss. This task involves the acceptance of the loss at an emotional and intellectual level.
- Task 2. To work through the pain of grief. Here the bereaved person is encouraged to explore and experience the pain of their loss. The use of rituals and creative artistic expression can be useful for expressing distress and emotion.
- Task 3. To adjust to an environment in which the deceased is missing. Here the bereaved will try out new roles and wonder how to fit into their new world. Some may try to avoid this task by further isolation and withdrawal in fear of leaving behind their loved one.
- Task 4. To emotionally relocate the deceased and move on with life. The bereaved needs to be able to think of their loved one in a safe space and reconnect at special times. Hopefully, they are now able to get on with life, yet still remember their loved one without being overwhelmed with grief. They can make new relationships and everyday life continues normally.

In 1982 Worden changed Task 4 to: 'withdrawing emotional energy from the deceased and reinvesting it in another relationship'. It could be considered that this was in step with Freud's theory that the bereaved need to 'decathect' from the dead.

Research was beginning to demonstrate that the bereaved continued to have conversations (out loud, or in their heads) with loved ones. So in 1991 Worden changed Task 4 to: 'To emotionally relocate the deceased and move on with life'.

As the continuing bonds theory continued to grow in popularity Worden changed Task 4 for the last time to: 'Find an enduring connection with the deceased in the midst of embarking on a new life'.

Whilst the more traditional 'phase/stage' models have been useful to understand the process of grief they appear to suggest that grief responses are rigid with systematic movement through the stages.[19] They do not allow for the unique experience of bereavement of those linked with ALS, when compared with other deaths,[19] and these limitations often encourage the bereaved to consider more modern models for support.

Modern models

Stroebe and Schut[20] offered a challenge to the traditional models of grief. Do people have to let go to be able to progress in their grief? Stroebe suggested that there is little research to support this, and the dual process model was introduced by them in 1995.[20] This was the first model to suggest that there were no defined stages of grief but instead they described two styles of coping processes:

- 'Loss-orientating' coping looks at the loss of the loved one.
- 'Resolution-orientated' coping deals with particular problems and considers new activities and pastimes.

People need to fluctuate between the two styles in their grieving, to allow full expression of feelings and distraction from grief.

Continuing bonds

Present-day thinking encompasses both letting go of bonds and keeping hold of the attachment.[21] Moving in and out of grief, paying attention to the present, appears to mirror the experience of grief.[22] The popularity of continuing bonds from around 1996 onwards[21] encouraged a wider thinking that maintaining connections with the deceased was universally beneficial. However, Bonanno et al.[23] assessed a group of bereaved people and found that those with strong continuing bonds had the most noticeable grief indicators 5 years following a death. Nevertheless, this model remains popular and offers hope and solace for many bereaved. Comfort is sought in memories and making meaning from history to

carry forward to the future. Often working with the bereaved in this way enables them to move forward in their grief by having permission to 'hold on'.[21]

Narrative

The dead help us to write their stories—ours as well. In a sense, every story has a ghost writer.[24] As humans, we understand our world by making meaning. When a loved one dies we need to understand what happened, and part of this process is to build a narrative around the loss.[25] This narrative or story will include what happened, the sequence of events up to and including the death, and how the bereaved felt then and now. Talking to others in the family network often helps the bereaved 'pad out' the narrative until it becomes a story of fullness. It appears that when meaning is found in a loss, the bereaved experience positive adaptation.[26]

Grief responses

Bereavement and loss are considered significant sources of stress for individuals and families. Using theoretical models of stress and coping we can begin to understand the impact of bereavement and how we respond physiologically, psychologically, and behaviourally.

Physical responses

Following a death, the bereaved may experience a range of physical responses. These are often not recognized as a response to grief, perhaps not until an assessment for grief counselling is offered.

Normal responses may include difficulty in getting to sleep or a broken sleep pattern. For an ALS carer, this broken sleep pattern may have become normal as the needs of their loved one increased throughout the night. They may experience weight loss or gain, headaches, chest pain, digestive problems, night sweats—the list goes on. There may be an increased susceptibility to stomach upsets and colds and coughs. Pre-existing conditions may become worse as the body's immune system is weakened. Irwin et al.[27] connected suppression of the immune system and disrupted sleep in the bereaved. Often, the bereaved person will lose interest in taking care of his or her physical well-being. In ALS-related bereavement the medical/physical needs of the carer have often been put on hold to care for the patient.

Psychological and emotional responses

The bereaved may experience a wide range of psychological responses in their grief. These could include: sadness, guilt, anger, numbness, shock, loneliness, fear, anxiety, and helplessness.

Bereaved people will often describe their experience as detachment—they are so shocked that they feel removed from what has happened. This is often the time when they will review the story from diagnosis to death and try and make sense of the loss—the beginning of the narrative process. Anger may follow here—perhaps expressed at professionals, family members, themselves, or the deceased. Some may feel overwhelmed by sadness and despair. Others struggle with their world being out of control. This anticipation and anxiety about new roles can tip the bereaved person into depression; this is a common occurrence. However, clinical depression needs to be recognized as being apart from normal feelings of sadness. If the bereaved person is unable to function normally, not caring for themselves or dependants and experiencing a continuing low mood, clinical depression may be present. They may need various forms of support at this time, including medication.

Loneliness and isolation are often particularly acute in grief. In ALS-related loss this may be highlighted owing to the length of time that has been spent caring for a loved one and the consequent disconnection from social networks.

Cognitive changes are often those that are least expected by the bereaved. Confusion, forgetfulness, and restlessness are very common, yet cause great anxiety for many. This experience can be made easier through normalization—either from professionals or through shared experience. Some find the processing of information hard, and other family members may express their concern at this. Dreams and hallucinations of seeing or feeling the dead are common and this can be experienced as comforting or promote further anxiety.

Behavioural responses

Crying is perhaps the most common behaviour and symbolizes the expression of loss. Other behaviours include outbursts of anger, social withdrawal, sleep disturbance, and, for some, eating too much or too little. Bronstein et al.[28] demonstrated in a study of widows that some 17% were still experiencing acute grief 13 months after the death of their husband. Sleep disturbance may reflect previous patterns of caring at night, and sleep takes a time to revert back to a more normal pattern.

Food issues often arise as the bereaved may be out of the routine of cooking, and even have forgotten what they like to eat. Loss of role and low self-esteem can make it a challenge for the bereaved to rediscover their appetite. Again, support from professionals or a person's social network can encourage a healthier interest in food.

Over time the bereaved person will notice that their grief responses are easing and that they are becoming more confident in their new environment. This may

be evident in their ability to reorganize themselves and make new friends. Slowly they will adjust to their experience and be able to recall happier memories linked with their loss, and recognize their resilience in coping and functioning.

Interventions

There is much written about resilience in the journey through bereavement. Bonanno[29] discovered that 45% of bereaved spouses showed no symptoms of grief (depression, anxiety, despair, or yearning) 6 months after their spouses had died. Their resilience had allowed them to maintain 'relatively stable, healthy levels of psychological and physical functioning'.[29] Bonanno's work on resilience has concluded that whereas emphasis on expressing anger and tears has played a large part in bereavement interventions, laughter and smiling can be more helpful in adjusting to loss.[30]

Many bereaved persons will experience the death of a loved one and move through the cycle of grief needing little if any intervention—making new relationships and facing the future with confidence. Others may benefit from professional input.[31] Following the death of a loved one from ALS, professionals will find it useful to be aware of a range of interventions that are available to assist the grieving family, should they require it.

Bereavement support

Individual counselling may be helpful for a person bereaved through ALS. For others, group therapy or a relatives' group may be more appropriate. Some may seek support through their family, church, or some other community group. Studies have shown the importance of religious or spiritual beliefs in bereavement and when adjusting to loss.[32] For someone bereaved through a life-limiting condition such as ALS, it may be difficult to come to terms with the fact that the illness journey has ended. The professional may need to make real the loss to allow the bereaved to engage in the grief process. The suggestions in the rest of this section may facilitate this.

Viewing the body

This can be the first step in facing the reality of the death and, for many, provide comfort. The bereaved may never have seen a dead body before. It is important to allow for expression of fears and expectations so that the bereaved can make an informed choice about whether viewing their loved one is right for them. Any religious or cultural differences need to be considered and outside experienced support sought if needed.

Providing information about the grief process and agencies that can offer additional support

For many bereaved people the grief process is unknown territory. This can add to the feelings of anxiety and confusion already being experienced. Offering information about the grieving process can be part of normalizing the experience. A list of available bereavement services allows the bereaved to consider support options when they feel able. Naturally, there are practical arrangements to take care of following a death. Organizing the funeral and dealing with a loved one's estate can be time-consuming and exhausting. The bereaved person can feel very alone and uncertain at this time. Information about available support can be invaluable.

Normalizing

In the grief reaction there can be a wide range of feelings and overwhelming emotions.[33] These emotions can throw the bereaved out of control and make them feel that they are tipping into chaos. This often leads many to ask: 'Is this normal' and 'Am I going mad?'. Normalizing and allowing for freedom of expression can offer the person a safe space to talk through their pain and understand that their experience is normal for them. Once this is understood the bereaved person will often then be able to move into a calmer phase of grief.

It may be that other family members have expressed that it is time to move on, or that they feel the individual needs a high level of support. Reassurance of a normal experience is often all the bereaved person needs to allow them to begin to make meaning of their loss.

Expression of emotions and feelings

It is vital that the bereaved person is allowed to identify and express their feelings around their loss,[34] and being encouraged to do so early after the death can set a positive beginning to their journey through the grief process. This process is exclusive to each individual, and is, for some, beyond words. Verbal skills may be limited; there may be a barrier to communication or maybe language skills are not fully developed to use words. The language of art and creative expression can speak in ways that words cannot.[35] The bereaved person can be encouraged through many media to explore the right way for them to work through their feelings related to the loss and to imagine the future where they are restored and adjusted to their new world.[36]

Research supports that writing about suffering and trauma has a positive impact on the growth and movement required to move through grief and loss.[36]

Resilience, special days, and memories

Birthdays, death anniversary days, wedding anniversaries, and any other times that mark milestones can be a challenge in grief. For many bereaved people it is hard to think about continuing to observe them. For some, the best way to approach the anniversaries is to view them as a time for remembrance and reflection. Others prefer a more proactive approach and will plan ceremonies and rituals that are appropriate for them and the family.

Social media, networking, and forums—making memories

Historically, the bereaved have always needed special places for rituals and memorials. These spaces can be linked with the continuing bonds theory. The bereaved will leave cards, flowers, and gifts and spend time there remembering their loved one. For many, the special place allows for expression of feeling that is otherwise locked away: 'In a democratized and mediatised society, it appears that every individual must count, and not have his or her existence pass unnoticed'.[37]

In a society that relies heavily on the Internet for shopping, leisure, information, etc., it is not surprising to discover that it is now used for memorialization and bereavement support.[33] Virtual memorials are created and maintained perhaps through a Facebook site[4] or a more specialized site such as Gonetoosoon.[4] This site had over 100,000 memorials in 2011. Such dedicated memorial sites have been described as 'web cemeteries'.[38]

These virtual memorials usually consist of photograph, a written narrative history of the deceased, and perhaps the cause of their death. This represents an opportunity for the bereaved to work on narrative—an 'enduring biography'[39] and to create a therapeutic zone.[40] Messages can be left 24 h a day and no one has to leave their home to express their feelings or find support when at a low ebb. Relatives across the world can be united in their grief and can share narrative, photos, and videos at the touch of a button. The internet site Much Loved[41] has memorial gardens and 'candle sanctuaries' which mirror real spaces, perhaps in a church or crematorium. Visitors here treat the virtual space as a creative space to form continuing bonds with the deceased.

Children in particular relate well to digital networking and can either work alone or with support from an adult. Access to computers for those without their own can be through schools, libraries, or community groups. Digital media can be a useful addition to the more 'physical' aspects of memorial making—such as memory boxes and photo albums. The child can be encouraged over time to collect relevant objects that have meaning and connection to their loved

one and the time they spent together. Items could include cards, music, a piece of jewellery, cinema tickets, perfume/aftershave, clothing, letters—in fact anything that will help the child create a narrative. This resource can be then used to promote storytelling within the family or to be browsed through alone during quiet times of remembrance.

For children within a family affected by ALS, young carers groups[42] can offer a good social network to help foster their resilience before a death. These groups are useful to reduce isolation and loneliness[43] and allow a shared/similar experience away from the family. They can offer the child a safe place to explore difficult feelings that the child feels unable to express at home. This free expression can change how children communicate with family members and increase their sense of worth and value.

Conclusion

There is still no adequate research on the impact of ALS in bereavement for families and caregivers. Practitioners will continue to support families following best practice and the needs of their clients as they understand them. As a collective, we will continue to search for support and direction at the time of bereavement. However, the dangers of becoming too prescriptive and offering blanket support for all are now becoming apparent. Outlining the normality and duration of normal patterns of grief, as distinct from a more severe reaction, will support the resilience of most bereaved people and focus additional support where it may be better aimed.

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Chapter 20

No time to waste: a family's journey from diagnosis to bereavement

Marika Warren, Michelle Warren, and Douglas Warren

Introduction

What does good (palliative) care look like for patients and families with ALS? We hope that the insights we gained from our journey will help to improve care for other patients and families.

Our family's experience with ALS can be summed up in two themes: 'seize the day' and 'be prepared'. These might seem to be in opposition, but being prepared enabled us to seize the day without wasting time worrying about what was to come next.

At the time of diagnosis our family consisted of Mary, 59, a school librarian and former nurse, who had ALS; Doug, 58, a mechanical engineer and Mary's husband of 35 years; Marika, 31, a bioethicist; and Michelle, 29, a theatrical seamstress. Mary, Doug, and Michelle were living in Edmonton, Canada and Marika was in Winnipeg with her partner, Alex.

In general, good ALS care helps the patient and family feel aware of and prepared for the challenges that arise as the disease progresses while helping them achieve important goals such as finishing projects, maximizing 'quality time' with loved ones, and leaving a legacy.

Experiences leading up to diagnosis

Mary first noticed some weakness in her arms at the age of 58. Although initially thought to be related to breast cancer or its treatments two decades earlier, after batteries of tests, ALS was diagnosed 8 months from her first symptoms and 2 months before her planned early retirement. She had been looking forward to spending more time on her hobbies, particularly quilting, walks with her two dogs, travelling, relaxing at the family cottage, and time with her family

and friends. The diagnosis of ALS changed all that, and Mary felt that she had been cheated of her well-earned retirement.

Diagnosis

The confirmed diagnosis was reached relatively quickly, partly due to Mary's self-advocacy for a timely referral to neurology. Given that Mary lived just a little more than 2 years after the first appearance of symptoms, we are grateful that the diagnosis came when it did so that we could prepare and start making the most of each day. Mary's father had died of ALS, so we probably had more information and a greater understanding of the disease than most others would at this stage.

Early stages (year 1)

Clinical condition and care situation

Mary stopped work and started NIV at night. As a result, she experienced an increase in energy. She was still climbing the stairs to get to the bedroom and walking around the house without assistance, but she did tire easily and most excursions involved a manual wheelchair.

Our needs and challenges

Our neurologist was very forthright and honest in answering our questions, and we focused on making the most of the time we had remaining. The ALS Society of Canada offered much information and support. The medical professionals who were the most helpful were those who anticipated future issues. Health system constraints, however, limited other providers to addressing immediate needs, not accommodating the progressive nature of ALS, which meant we were less prepared for the next challenges. A simple example was equipment for bathing. A bath stool provided after the first in-home needs assessment by occupational therapy quickly proved inadequate, but the more appropriate bench with back support was not funded by the health system because only one assessment/piece of equipment was supported in a set period.

We knew and accepted that all care was essentially palliative in terms of approach. We opted for time with family and friends, spending time at our cottage, and enjoying symphony concerts and live theatre. The therapeutic effect of having small dogs was also substantial for Doug and Mary; they helped to ease the pain by cuddling up and providing company. With the time left, we choose to make memories to help overcome Mary's fear of being forgotten and to leave

legacies for the future. We reproduced a family ring, created a citizenship award at the high school where Mary had worked, finished quilts, and made audio recordings of favourite children's stories. We also arranged regular gatherings of groups of friends, and a Danish exchange student who had lived with us in the late 1990s came back for a visit and to say goodbye.

We are a family of 'doers' and thus started to adapt the house in anticipation of the later stages of the disease, building wheelchair ramps into the living room, family room, and garage. Mary did not want to die in hospital, so we prepared our home to eventually become a hospital. We purchased a second NIV machine for backup in case one failed (which had already happened). The second machine made it easier for Mary to move around the house as symptoms worsened. We turned a main floor bathroom and adjoining closet into an accessible bathroom complete with a roll-in shower. Planning ahead and executing early meant that we could transition through the phases of the disease as seamlessly as possible and maintain caregiving by the family in the home to the very end. We also began to adapt Mary's wardrobe and equipment, drawing on Michelle's professional expertise as a seamstress and on the knowledge of a family friend who had muscular dystrophy.

Marika, in addition to frequent trips to Edmonton, visited every afternoon by Skype—although as soon as the quiz show *Jeopardy* came on, Mary summarily signed off. Such small rituals from pre-diagnosis life that were not disease-focused were important in our new routines.

Our greatest need at this stage was for information that would help us to prepare for what was to come, which was provided by both the ALS clinic team and the ALS Society of Canada. The relationships established with healthcare providers and medical equipment suppliers at this stage were of great therapeutic value as the disease progressed. Of particular value were relationships with the respiratory therapists who were extremely proactive in ensuring that Mary had the respiratory support she needed as the disease progressed. We were more capable of big-picture thinking, as was reflected in our work on the house. Most of the advance care planning was done as well, including the decision to insert a feeding tube. In retrospect it probably would have been a good time to transition to palliative home care.

Mid stages (second year, first 6 months)

Clinical condition and care situation

Mary began using the NIV for brief periods when awake to relieve increasing breathlessness. Communication was more effortful, and the computer became a central part of Mary's day, providing the ability to communicate and connect

with the outside world as venturing outside the house became increasingly challenging. As climbing stairs became excessively tiring we transformed the ground floor living room into Mary's bedroom, and thanks to the ramps built earlier she was still able to access the entire main floor of the house. Although a manual wheelchair was provided early on, it was obvious that a powered wheelchair would soon be needed.

Midway through this period it was necessary to have someone in the home at all times and therefore Michelle moved back into Mary and Doug's house to share in the work of caregiving.

Our needs and challenges

During this stage the acquisition of equipment and developing home-based care came to the forefront.

The process of obtaining the power wheelchair took far too long and caused unnecessary hardship. When it did arrive, it significantly improved Mary's quality of life. Other equipment was obtained in a more timely manner from a variety of sources including hospitals, medical suppliers, community programmes, and the ALS Society of Canada. Some maintenance of the equipment, modifications, and repairs were done by family members and this minimized the need for additional outside involvement. Internet searches provided some technical solutions. Building fit-for-purpose devices, such as a pressure-activated foot switch, allowed Mary to signal caregivers when only a very limited range of motion remained in her hands and feet. Devising solutions provided us with a sense of control and efficacy and constituted minor (yet important) victories over ALS. The ALS Society of Canada was exceedingly helpful with information, references, physical resources, and support for the caregivers.

Mary continued to attend ALS clinics, which were well organized with all of the medical disciplines in one location at one time, but onerous. Repeating information as each discipline came in for check-ups was one burden—some sort of 'real-time' information sharing would have minimized the effect on us. The tremendous amount of information delivered required multiple family members to ensure that all of the messages were heard, as often one or more of us became overloaded, distracted, or confused.

Building the relationship with the home care team was very helpful and therapeutic, but took time. Consistency of home care providers was crucial and made it easier for us to prepare for visits and maximize the time with these professionals. However, home care was not equipped to handle a rapidly progressive disability, nor for providing palliative care in the long term. Not all of the healthcare providers were versed in palliative approaches to care and some

of the advice we were given (e.g. regarding nutrition requirements) was inappropriate for Mary's situation.

One thing that was frustrating was being asked what we needed and then being told these requests could not be met given system constraints (e.g. when we requested additional respite care). It would have been useful for healthcare providers to describe what was available and let us choose those things that were the most applicable to our situation or stage of disease progression.

Respite workers were available periodically, but were of limited benefit due to lack of ALS-specific knowledge and skills. For example, they were often unfamiliar with the equipment, such as NIV, or were not permitted to administer medication on an 'as needed' basis. Turnover of personnel, in part due to the increased skill required as the disease progresses, was tiring as energy had to be expended in bringing new people up to speed.

Final months and weeks

Clinical condition and care situation

Eventually Mary found leaving the house to be too draining. To retain a critical semblance of normality, everyone, from the ALS clinic team to our family physician to Mary's friends and hairdresser, came to us.

Mary's daytime use of NIV increased until she was using it constantly, which further interfered with communication. She avoided using alternative communication, finding it slow and cumbersome, relying instead on family to interpret. The PEG tube was used for all nutrition and medication.

Marika took a leave of absence from work, returned home, and the entire family coordinated efforts to provide round-the-clock caregiving.

We understood that a typical palliative patient needs six to eight in-home caregivers; we did it with just the three of us. This isn't necessarily feasible for everyone, but we had done our research, were focused, and had done everything we could think of to be prepared. The ALS clinic team called us the 'model family', but we were just lucky; we had the right strengths in personality, skill, expertise, and financial resources. This is not to say that it wasn't difficult and exhausting at times or that we did not need physical and emotional support from healthcare providers.

Mary died only 20 months after the confirmed diagnosis and 28 months after the first symptoms appeared.

Our needs and challenges

Routine was important, particularly getting Mary out of bed. Mary strongly preferred being transferred by a caregiver and became anxious using a Hoyer lift.

Non-family caregivers would have probably been required to use the lift, which would have affected Mary's willingness and ability to get out of bed every day. In addition, the lift we were offered was better suited to a hospital environment and didn't roll smoothly on the thick carpet in the living room. This illustrates the challenge of transforming our home into, essentially, a small hospital.

Transitioning to the palliative home care team was a challenge, not because of fear of the implications but because we had to establish relationships with new providers. This was tiring, especially when the existing relationships were working well and were important to all of us. We were fortunate that some providers worked with both teams, and this provided some continuity. Being able to talk about other things like a healthcare provider's children or her upcoming vacation in addition to Mary's medical status made the interactions with healthcare providers much more positive and enjoyable.

We particularly appreciated honest and direct information from the palliative care teams (both home care and the ALS clinic). It was frustrating for us when people were talking in euphemisms and avoiding description of the end stages of the disease. It was particularly important to be prepared for a sudden crisis. We needed to know who to call and what to do. We wanted detailed information so that we could ensure that Mary would not suffer needlessly. When it wasn't clear to us how to manage such a situation it significantly increased our levels of anxiety. Palliative home care suggested preparing for the need for palliative sedation, but did not provide the specific details that would reassure us. We eventually got information regarding the experience of using palliative sedation from a family friend whose husband died of cancer. This was invaluable to us, but it would have been helpful to get that from the healthcare team.

As with many who find themselves in caregiving roles, self-care was challenging for us; while we recognized its importance, the respite resources available were often insufficient to provide a significant break (such as a full night's uninterrupted sleep). It was also challenging when all of us were doing all that we could for Mary, and we wanted recognition of that fact from each other, which wasn't always forthcoming. Affirmation from Mary and from the healthcare teams involved that we were doing a good job of caring for Mary was appreciated, but it was also helpful when they asked us how we were doing and didn't assume that because things seemed to be going well that there wasn't anything that we needed.

Individual thoughts and reflections

Michelle

The in-home clinics with both home care and ALS clinic staff were great for us. They generated some of the best discussions about Mom's condition not only because of sharing knowledge and brainstorming of solutions with all of the

healthcare providers but also because they strengthened the emotional relationship with the team. One discussion I remember well is the neurologist telling us that there might be a day where Mom wakes up and says, 'Shit, I woke up'. It was helpful to hear, at that point, about another person's experience. The reaction Mom had to the story, that she understood she might have that day, was painful but honest and I felt kind of gave her permission to feel that way.

Marika

It was important in this period that I was allowed to say goodbye; I needed to let Mom know that we would be okay and she didn't have to hang on for our sakes if her suffering became excessive. Some of the healthcare providers reacted to this by trying to emphasize positivity, which was not helpful for me.

Aftermath

After Mary died there was very little follow-up from the palliative home care team and some details were missed (a respite worker reported for a shift 2 days after Mary died). We would have appreciated more extensive follow-up with the whole family.

Individual thoughts and reflections

Marika

I feel that Mom had best possible end-of-life experience, which helped me to cope with her loss. As a bioethicist, I deal with end-of-life issues in my professional role, and this also provided a way for me to work through and reflect upon the whole experience.

Michelle

I do not remember any follow up from the ALS Society [of Canada] with me specifically after Mom's passing. Perhaps this was because of a lack of contact information but the impression it left was that I no longer needed 'caring for'. When I reached out, there was certainly someone there, even years later, but a check-in after Mom's death with myself and not just my Dad would have been appreciated. I believe that Mom was extremely well cared for by all involved but that our family was abandoned once she was gone.

Conclusions

Our experience was not necessarily typical; we had a lot of resources (financial, access to medical information, leave/time available, and professional expertise) that others might not. We also had knowledge of the ultimate outcome and could prepare for that, our goal being to prepare for the worst while hoping for

the best. Our preference, therefore, for early integration of palliative care into ALS management might not be universally shared.

For us, the most important aspects of the care we received, whether palliative or not, were the relationships that it created; it was critical that healthcare providers not just cared for, but also cared about, Mary and her family. These relationships thrive on consistency of care providers, and we appreciated the efforts that many healthcare providers made to maintain relationships once established.

The care that we received helped us to feel prepared and to capitalize on the time that we had left together as a family. As much as there were tears there was also laughter and a lot of love.

Appendix

List of useful websites for ALS patients

There are thousands of websites which claim to contain information relevant to ALS patients. However, in many cases the information has not been reviewed for accuracy and may sometimes be misleading or even directly aimed at obtaining financial profit by offering 'miracle drugs' or other devices with no proven benefit. Therefore, some degree of caution is advisable when surfing the net for information on ALS.

The following websites all have a very high standard and offer reliable information, addresses, and links to other websites of interest. This list, of course, is by no means exhaustive.

- World Federation of Neurology Research Group on Motor Neuron Diseases/Amyotrophic Lateral Sclerosis: <http://www.wfnals.org/>. An excellent site with regularly updated clinico-scientific news on ALS and a worldwide list of ALS specialists and centres.
- The International Alliance of ALS/MND Associations: <http://www.alsmndalliance.org/>. Includes a directory of ALS societies worldwide.
- The ALS Association [of America]: <http://www.alsa.org/> This site has a great deal of relevant information.
- The American Muscular Dystrophy Association: <http://mda.org/>. This also runs a strong ALS care programme.
- The Les Turner ALS Foundation (Chicago): <http://www.lesturnerals.org/>.
- The ALS Association [of Canada]: <http://www.als.ca/>.
- The [UK] Motor Neurone Disease Association: <http://www.mndassociation.org>.
- ScotMND (the Scottish Motor Neurone Disease Association): <http://www.scotmnd.org.uk/>
- The Irish Motor Neurone Disease Association: <http://www.imnda.ie>.
- Deutsche Gesellschaft für Muskelkranke eV [the German Neuromuscular Disease Association]: <http://www.dgm.org>.

- Internet portal for ALS.
- Award-winning website dedicated to providing information to ALS patients.
- A well-designed website for ALS patients, run by an ALS patient.
- The ALS Therapy Development Institute: <http://www.als-tdf.org/>.
- ALSUntangled, a group of 90 clinicians and scientists representing 10 countries, who have developed a standard method for reviewing complementary and alternative medicines: <http://www.alsuntangled.com>.
- *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration* [official journal of the World Federation of Neurology Research Group on Motor Neuron Diseases], abstracts and contents: <http://informahealthcare.com/loi/afd>.
- Family Caregiver Alliance [USA]: <http://www.caregiver.org>.
- Caring Connections [website of the National Hospice and Palliative Care Organization, USA]: <http://www.caringinfo.org/>.
- Website of the National Institute of Health National Institute of Neurological Disorders and Stroke: <http://www.ninds.nih.gov>.

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