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ROYAL COLLEGE OF SPEECH & LANGUAGE THERAPISTS

Supporting people living with motor neurone disease

Motor neurone disease may affect speech, communication and swallowing. It may also affect thinking and behaviour. Speech and language therapists (SLTs) are a key part of the multi-disciplinary health and care team supporting people living with motor neurone disease and their families and carers. They promote better quality of life both for people with the disease and their loved ones. They support people with changes in speech and communication, including through the use of a range of technologies to support or replace spoken communication. They support people to eat, drink and swallow safely. SLTs can also support the determination of mental capacity and contribute to supported decision-making for those people living with motor neurone disease who experience cognitive and behavioural changes.

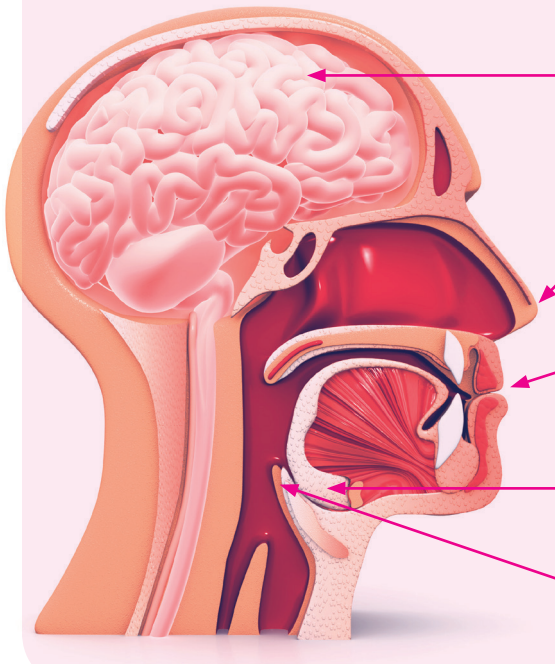
How many people does motor neurone disease affect?

- The annual incidence of MND is **1.5 - 2.5 per 100,000**, with **6,000** individuals affected in the UK at any one time.[1]
- MND kills a **third** of people within a year of diagnosis, and more than half within two years.[2]
- **Six** people are diagnosed with MND every day[3] and six people die from it every day.[4]
- A person's lifetime risk of developing MND is **1 in 300**.[5]

What is motor neurone disease?

Motor neurone disease (MND) is a progressive neurological condition. It describes a group of diseases that affect the nerves (motor neurones) in the brain and spinal cord that tell muscles what to do. In someone with MND, messages from these nerves gradually stop reaching the muscles, leading them to weaken, stiffen and waste.[6] MND can affect how a person walks, talks, eats, drinks and breathes. Some people also experience changes to their thinking and behaviour. However, everyone is affected differently by MND[7] so individualised assessments and regular reviews of the support needed are required.

What will listeners notice?



LANGUAGE word selection and sentence formation	If affected, can impact on the meaning of what is said
BODY LANGUAGE Reduced head movements and hand and arm gestures	Can impact on conversational turn talking
SPEECH articulation, resonance and pronunciation	Indistinct, slurred words with a possible nasal quality
INTONATION Melody of speech	One level of loudness with low pitch
VOICE	Harsh or 'strangled' voice quality, sometimes 'wet'

Impact on speech, communication, swallowing, thinking and behaviour

Speech and communication

- Speech problems (dysarthria) occur in more than 80% of people living with MND as the disease progresses.[8]
- Approximately one third of people with MND experience bulbar motor neurone disease, a particularly aggressive form affecting speech and swallowing muscles early in the disease process.[9]
- Speech can deteriorate over a period of a few months and is cited by people living with MND as one of the most problematic symptoms.[10]
- There is an association between early augmentative and alternative communication (AAC) intervention for people living with the disease and higher quality of life for patients and carers.[11]
- A deterioration in intelligibility does not correlate with a perceived deterioration in social interaction.[12]
- Respiratory weakness sometimes means that a non-invasive ventilation (NIV) face mask is worn, and this can compromise intelligibility.
- Fatigue is a significant symptom of MND and this can cause an individual's speech to deteriorate as the day progresses.

Eating, drinking and swallowing

- Swallowing problems (dysphagia) occur in more than 80% of people living with MND as the disease progresses.[13]
- Swallowing problems put people at risk of choking (asphyxiation), chest infections (aspiration pneumonia), malnutrition and dehydration.
- Pleasure from eating and drinking may be reduced, which can impact on patients' enjoyment of social occasions, which often involve meals and drinks.[14]
- Drooling is common in people with MND due to difficulties in managing saliva.[15]
- Adequate adjustments to diet and changes to posture to preserve oral feeding can postpone the need for being fed by tube.[16] Adjustments can include changes to the consistency of food, fluids and medication.

Thinking and behaviour

- Around 35% of people with MND experience mild cognitive change – in other words, changes in thinking and behaviour. A further 15% show signs of frontotemporal dementia.[17] These changes can affect planning and problem-solving or lead to impulsive behaviour.

The role of speech and language therapists

SLTs play a key role in supporting people living with MND and their families and carers, including in preparing for changes in speech, communication and swallowing. They can also contribute to supported decision-making.

Communication: SLTs support all aspects of communication and changes to communication in partnership with people living with MND. They also help people prepare for, and support the potential transition to, augmentative and alternative communication (AAC). This may include anticipatory voice or message banking.

AAC involves a range of technologies to support or replace spoken communication, including: writing, gestures, signing, symbols and pictures, communication boards, tablets and computers. How people access these systems physically is an important consideration (for example, via switches or using eye-gaze technology).

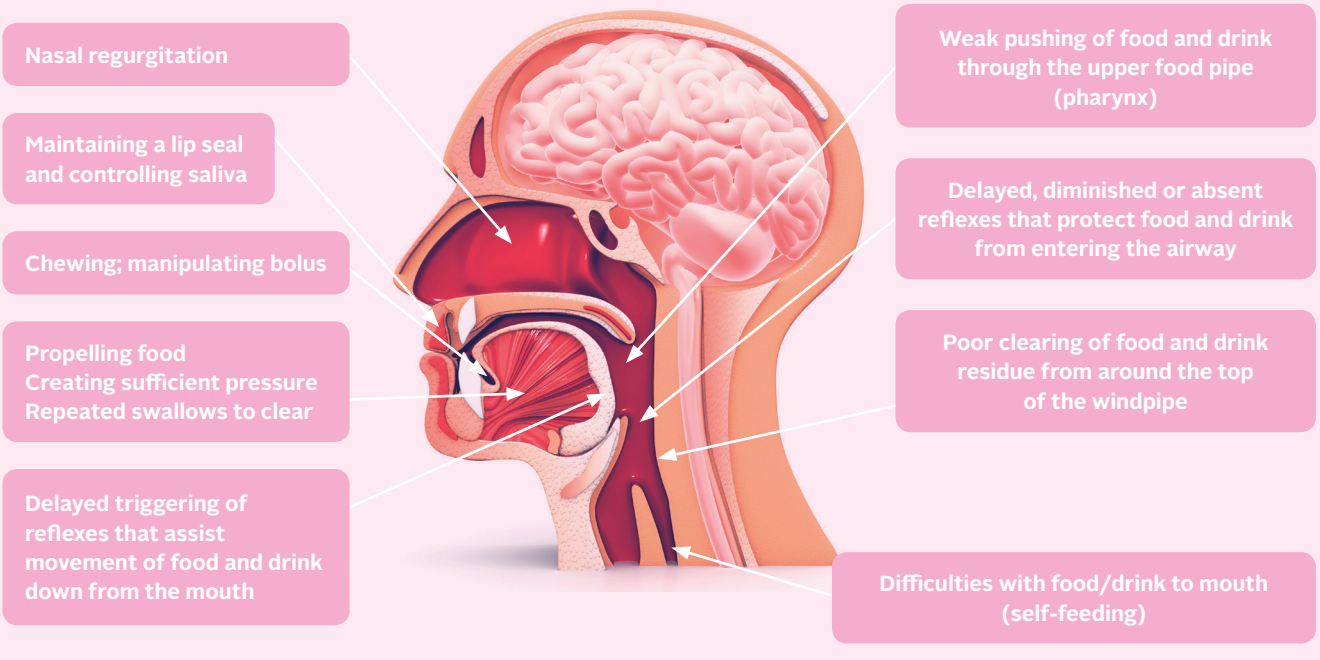
Regular reviews of AAC are required because as an individual's condition progresses, their physical ability to access a device may change.

Swallowing: SLTs help to minimise the physical, emotional and psychological effects of dysphagia. They promote quality of life and safety in eating, drinking and swallowing and saliva management.

They also collaborate with other members of the multidisciplinary team such as dietitians, doctors and nurses to help advise patients when feeding tube placement would be appropriate and timely. Due to fatigue, an individual's swallow may deteriorate as the day progresses.

Decision-making: SLTs can contribute to supported decision-making and the determination of mental capacity where changes to cognition and behaviour impact on thinking or give the impression of impacting on thinking. This promotes better planning and decision-making and reduces the risk of people being perceived as lacking capacity due to their communication needs not being appropriately supported.[18]

How are eating and drinking affected in motor neurone disease?



NICE guideline

The 2016 National Institute for Health and Care Excellence (NICE) guideline on motor neurone disease: assessment and management[19] highlights that:

- the disease has an impact on speech and swallowing;
- the multidisciplinary team caring for people living with MND should assess, manage and review a range of areas including swallowing, speech and communication;
- the multidisciplinary team should include SLTs;
- when assessing speech and communication needs during multidisciplinary team assessments and other appointments, face to face and remote communication (for example, using the telephone, email, the internet and social media) should be discussed;
- the assessment and review should be carried out by an SLT without delay;
- AAC equipment that meets the needs of the person should be provided without delay to maximise participation in activities of daily living and maintain quality of life; and
- all discussions should be tailored to the person's needs, taking into account their communication ability, cognitive status and mental capacity.

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Mary's story

When Mary's speech became slurred and she started coughing when drinking tea she was referred to a neurologist by her GP. After a series of tests she was diagnosed with motor neurone disease. This diagnosis left her and her family devastated. She explained, 'I was so scared that I wouldn't be able to communicate if I lost my speech'. Mary met her speech and language therapist, Tahsin, within a week. Tahsin spent time with Mary, listening to her concerns and exploring her needs and priorities. As Mary's eating and drinking became harder, Tahsin worked closely with a dietician and reassured Mary that she would always be able to enjoy something to eat even if she needed to be fed non-orally at a later point. As her speech deteriorated, Mary was reluctant to use a computer to communicate. But with Tahsin's reassurance and the expert advice of the regional AAC team, she began to use a tablet with synthesised speech output. Mary says, 'Tahsin and her colleagues have made such a difference to my life. MND is a horrible disease but the speech therapists have given me time to express myself as well as the motivation to carry on'.

REFERENCES AND RESOURCES

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[2] Motor Neurone Disease Association https://bit.ly/2qYVWSz

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[5] See Johnston CA, and Stanton BR, et al (2006) Amyotrophic lateral sclerosis in an urban setting: a population based study of inner city London. J Neurol 253: 1642-1643. Cronin S, Hardiman O, and Traynor B, (2007) Ethnic variation in the incidence of ALS: a systematic review. Neurology 68: 1002-7. Alonso A, et al (2009) Incidence and lifetime risk of motor neuron disease in the United Kingdom: a population-based study. Eur J Neurol June 16(6): 745-751. Al-Chalabi A, et al (2012) The genetics and neuropathology of amyotrophic lateral sclerosis. Acta Neuropathol 124: 339-352. Factor-Litvak P, et al (2013) Current pathways for epidemiological research in amyotrophic lateral sclerosis. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration 14(Suppl. 1) 33-43.

[6] Motor Neurone Disease Association https://bit.ly/2lXKaub

[7] Ibid.

[8] Tomik, B. and R.J. Guiloff, *Dysarthria in amyotrophic lateral sclerosis: A review.* Amyotrophic Lateral Sclerosis, 2010. 11(1-2): p. 4-15.

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[17] Goldstein L. H. and Abrahams S. Changes in cognition and behaviour in amyotrophic lateral sclerosis: nature of impairment and implications for assessment. Lancet Neurol. 2013; 12(4): 368-80.

[18] For more on the role of speech and language therapists in supported decision-making and the determination of mental capacity, see the RCSLT factsheet on mental capacity. https://bit.ly/2hJopMn

[19] NICE (2016), Motor neurone disease: assessment and management www.nice.org.uk/guidance/ng42

Resources

For more information about the support available to people living with MND and professionals working with them in England, Wales and Northern Ireland, visit the Motor Neurone Disease Association website:

www.mndassociation.org/

For information about Scotland, visit the MND Scotland website:
www.mndscotland.org.uk/

Acknowledgements

Photographs courtesy of Motor Neurone Disease Association.