FAQ

ALS-FTD

Frequently asked questions about ALS-FTD

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1. What are ALS and FTD, and what is the ALS-FTD spectrum?[1-4]

Amyotrophic lateral sclerosis (ALS) and frontotemporal dementia (FTD) are progressive neurodegenerative disorders that primarily affect different parts of the nervous system. ALS predominantly affects upper and lower motor neurons, leading to progressive muscle weakness and wasting, spasticity, fasciculations, and difficulties with speech, swallowing and, later in the disease, breathing. FTD comprises a diverse group of neurodegenerative syndromes involving the frontal and anterior temporal regions of the brain, resulting primarily in progressive changes in behaviour, personality, executive function and/or language.

Although ALS and FTD have historically been considered distinct disorders, substantial overlap in their clinical features, genetics, and underlying pathology has led to them being understood as two ends of a shared ALS-FTD spectrum. Between these two ends are a range of intermediate and mixed presentations involving different combinations of motor, cognitive, behavioural, and language impairments.

Cognitive and behavioural symptoms can occur in people with ALS – up to approximately 50% develop some degree of cognitive or behavioural impairment, with around 15% meeting diagnostic criteria for FTD – while motor-neuron dysfunction can also occur in people with FTD. The spectrum concept does not mean that ALS inevitably develops into FTD, or vice versa; rather, it reflects the biological and clinical overlap between the disorders and the considerable variation in how the disease spectrum presents between individuals.

2. What are the causes of ALS and FTD?[1,2,5]

The causes of ALS and FTD are complex and are not fully understood. Across the ALS–FTD spectrum, disease risk arises from a combination of genetic susceptibility and non-genetic factors that accumulate over a person's lifetime. Ageing is an important risk factor, and differences related to

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biological sex have also been observed. Environmental and lifestyle exposures may contribute to risk, although the strength of evidence varies between individual factors.

Genetics can have a particularly strong influence. Important genes associated with ALS include C9orf72, SOD1, TARDBP and FUS, while common genetic causes of familial FTD include C9orf72, GRN and MAPT. The C9orf72 repeat expansion is especially important because it can cause ALS, FTD or a combined ALS–FTD presentation.

At the cellular level, ALS and FTD also share several disease mechanisms. Abnormal TDP-43 protein accumulation is present in most ALS and in a substantial proportion of FTD, and shared pathways include disruption of RNA processing, protein homeostasis and autophagy, mitochondrial function and cellular transport, as well as neuroinflammation.

3. How common are ALS and FTD?[6-9]

Estimates of prevalence rates for ALS and FTD vary between regions and studies. For ALS, around 2 people per 100,000 are diagnosed each year, and about 4–5 people per 100,000 are living with the condition at any given time.

For FTD, around 2 people per 100,000 are diagnosed each year, while approximately 9 people per 100,000 are living with FTD. Estimates vary, partly because FTD can be difficult to diagnose and its symptoms can overlap with those of other neurological and psychiatric conditions.

Both ALS and FTD become more common with age, although they can also affect younger people. Both conditions are reported more often in males than females. As populations age, and as diagnosis and reporting improve, the number of people living with disorders across the ALS–FTD spectrum is expected to increase.

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4. What are the symptoms and signs of ALS and FTD?[3,10,11]

Symptoms vary widely across the ALS–FTD spectrum. ALS mainly affects movement and muscle control. Common symptoms include progressive muscle weakness and wasting, muscle stiffness or twitching, and difficulties with speaking or swallowing. Symptoms often begin in the arms or legs, although for some people they first affect speech and swallowing, and more rarely breathing.

FTD is more likely to cause changes in behaviour, personality, thinking or language. People with behavioural-variant FTD may become less motivated, more impulsive, less empathetic, develop repetitive behaviours or changes in eating habits, or have difficulty with planning and problem-solving. Some forms of FTD mainly affect language, making it harder to produce speech or understand the meaning of words.

There can also be overlap between these symptoms. People with ALS may develop changes in thinking, language or behaviour, while some people with FTD develop problems with movement and muscle control. The symptoms, their severity and the order in which they appear can therefore vary considerably from person to person.

5. How are ALS and FTD diagnosed?[3,10-12]

ALS and FTD are diagnosed primarily through clinical assessment - there is no single routine test that can diagnose every case. With ALS, clinicians look for progressive motor impairment together with evidence of upper and lower motor neuron dysfunction and assess whether another condition could better explain the symptoms. Electromyography (EMG) can provide supportive evidence of lower motor neuron involvement, while imaging and laboratory tests are often used to exclude alternative diagnoses.

FTD diagnosis is also primarily clinical and is based on the pattern and progression of behavioural,

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cognitive or language symptoms. Established criteria are used for behavioural-variant FTD and the primary progressive aphasia variants. Brain imaging can aid with diagnosis by revealing typical patterns of atrophy or altered metabolism, and genetic testing can support the diagnosis in certain cases.

Because cognitive and behavioural changes are common in ALS, ALS-specific screening tools such as the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) or ALS Cognitive Behavioural Screen (ALS-CBS) may be used. Mixed ALS–FTD presentations are classified using criteria that recognise cognitive impairment, behavioural impairment and overt FTD in people with ALS.

6. What treatment options are available for ALS and FTD?[13-16]

While there is currently no cure for ALS or FTD, treatment aims to slow disease progression, manage symptoms, maintain function and quality of life, support caregivers, and plan for future care needs.

For ALS, disease-modifying medicines can provide modest benefits in survival or disease progression. Symptom-directed treatment is also important for problems such as spasticity, cramps, mood symptoms and pseudobulbar affect (uncontrollable episodes of laughing or crying that do not match how a person actually feels).

There are currently no approved treatments that can slow or stop the progression of FTD. Instead, medicines may be used to help manage specific symptoms, such as changes in behaviour or mood. For example, antidepressants may help with some behavioural symptoms, while antipsychotic medicines may be used cautiously in more severe cases. Alongside medication, care from a range of healthcare professionals is important to help manage symptoms and maintain quality of life. This may include support with breathing, nutrition, movement, communication, thinking, and behaviour.

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7. What strategies can support management and quality of life in people living with ALS and FTD?[13,17-19]

Care is usually most effective when it is coordinated by a multidisciplinary team and adapted as symptoms change. Depending on the individual, this may include physiotherapy and appropriate exercise, occupational therapy and mobility aids, respiratory monitoring as well as non-invasive ventilation, nutritional support, and speech and language therapy. Communication aids can help maintain communication as speech becomes more difficult.

Cognitive and behavioural symptoms should be recognised early because they can affect treatment adherence, decision-making, and caregiver burden. Education, psychological or psychiatric support, practical environmental strategies, and regular reassessment can help patients and families manage these changes.

Shared decision-making and early planning for future care are also important. Caregivers should receive information, training, psychosocial support and access to respite, or peer support where available. Palliative care can be introduced early alongside other treatments to help manage physical and psychological symptoms, support families and align care with the person's preferences and values.

Lexicon[20,21,22]

ALS (amyotrophic lateral sclerosis): A progressive neurodegenerative disorder characterised mainly by degeneration of upper and lower motor neurons, leading to muscle weakness, wasting and loss of motor function. ALS is the most common form of motor neurone disease (MND).

FTD (frontotemporal dementia): A group of clinical syndromes caused by degeneration of frontal and/or temporal brain regions, typically affecting behaviour, personality, executive function and/or language.

ALS–FTD spectrum: The concept that ALS and FTD represent overlapping neurodegenerative disorders with shared clinical, genetic and pathological features, including intermediate and mixed presentations.

Behavioural-variant FTD (bvFTD): A form of FTD in which progressive changes in behaviour, personality and executive function are prominent. Features can include apathy, disinhibition, reduced empathy, repetitive behaviour and altered eating.

Primary progressive aphasia (PPA): A group of progressive disorders in which language impairment is the main early feature. The semantic and nonfluent variants are commonly associated with the FTD spectrum.

Motor neuron: A nerve cell that controls voluntary muscle movement. ALS involves degeneration of both upper motor neurons in the brain and lower motor neurons in the brainstem and spinal cord.

MND (motor neurone disease): A group of conditions affecting the motor neurones, the nerve cells in the brain and spinal cord that control movements such as walking, speaking, swallowing and breathing. There are several forms of MND, which differ in which motor neurones are affected first and how the condition develops. ALS is the most common form.

Fasciculation: A small, involuntary muscle twitch that can occur when lower motor neurons and the muscle fibres they supply are affected.

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