Background  Amyotrophic Lateral Sclerosis (ALS) is a progressive neurodegenerative disorder which can affect the muscles involved in swallowing, speaking, breathing, and ambulation by axonal degeneration of upper and lower motor neurons. Presentation is variable with two-thirds of patients experiencing weakness in the limbs first (1). The symptoms usually begin in one limb and then spread to contiguous limbs. One-third of patients develop bulbar weakness as the first symptom, usually presenting as dysarthria followed by dysphagia. Median survival after symptom onset in ALS is 30 months, but the range is anywhere from months to decades (1). Although the presentation and course are variable, ALS is an incurable and progressive illness that leads inexorably to death.

This will be the first of three Fast Facts addressing the management of common ALS symptoms. The intent of these Fast Facts is to inform readers of common symptoms clinicians may encounter when caring for ALS patients and to provide a brief overview on how to manage these symptoms. For more complex presentations, involvement of a neurologist, PMR clinician or ALS specialist is advised.

Sialorrhea  Bulbar weakness, spasticity, and loss of muscle control in ALS can lead to dysphagia which causes difficulty swallowing one’s saliva. In ALS, this condition is referred to as sialorrhea, although it is caused by difficulty clearing secretions rather than by an increase in saliva production as is the case in other conditions associated with the term sialorrhea. Sialorrhea in ALS often leads to drooling, which can be socially disabling as some patients are reluctant to engage in social interactions as a result of embarrassment. Difficulty with secretion management also increases the risk of perioral skin irritation and aspiration. There are various pharmacologic and non-pharmacologic management strategies for the treatment of sialorrhea in ALS:

• Experts often use medications that inhibit saliva production, such as atropine, glycopyrrrolate, tricyclic anti-depressants, and scopolamine patches, although there are no randomized trials specific to ALS which evaluate the effectiveness of these medications. These medications capitalize on the anti-cholinergic side effect of xerostomia; therefore, other less desirable anti-cholinergic side effects such as constipation, urinary retention, and cognitive dysfunction are a risk.
• The effectiveness of botulinum toxin injections into salivary glands has been demonstrated in randomized trials (2,3). The toxin reduces the production of saliva by inhibiting the release of acetylcholine at neurosecretory junctions within the salivary glands. Botulinum toxin A and B appear to have similar efficacy (4). The injections are typically performed by a neurologist in the office setting and generally must be repeated every 3 months. Inadvertent infiltration into adjacent muscles is a small risk. Rarely, xerostomia, dysphagia, or thickened secretions occur but the effects are most often temporary. Advantages of botulinum toxin treatment for sialorrhea are reduced systemic side effects and lack of drug-drug interactions.
• To minimize drooling, portable suction devices can be used to clear excess secretions.
• In refractory cases, unilateral salivary gland irradiation delivered over 1-5 fractions may improve sialorrhea within 24 hours (5). Maximum benefit usually occurs within a week. Although xerostomia is a risk, salivary function often returns after 3 months; hence retreatment may be indicated.
• In rare cases, laryngectomy is used for secretion management and prevention of aspiration in patients whose speech is already severely compromised (as it completely eliminates a patient’s ability to speak). This approach can be used regardless of whether a patient chooses long-term mechanical ventilation for ventilatory failure.

Summary  Although there is currently no cure for ALS, there are numerous meaningful interventions for the symptoms of the disease. Please see Fast Fact #301 for pharmacologic therapies and Fast Fact #300 for non-pharmacologic therapies of other common challenges faced by patients with ALS.
References

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