

Bulbar ALS: Unique Management Issues (Part II)

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“Bulbar ALS” is a term commonly used by physicians and patients to refer to those patients in whom ALS affects muscles of speech and swallowing. It may be just one part of generalized ALS in those individuals with weakness affecting other areas. Or, bulbar involvement may be the only manifestation of ALS, or the predominant one. For example, there are individuals whose bulbar involvement is so severe that they have no ability to speak or swallow, yet have normal strength in their arms and legs, along with normal respiratory function. In contrast, other individuals have severe arm, leg, and respiratory muscle weakness, yet minimal bulbar involvement. We do not know why some individuals develop bulbar involvement as the first symptom of ALS, while others develop bulbar involvement only as a late manifestation, or not at all.

Bulbar involvement presents its own unique set of problems. These are most commonly the following: 1) difficulty controlling saliva; 2) extreme, uncontrollable laughter or crying, also known as emotional lability; 3) impaired swallowing; 4) impaired speech. In the last newsletter, I discussed difficulty controlling saliva. In today’s column, I would like to discuss control of emotions.

Treatment of emotional lability is an important of issue. The tendency to laugh or cry easily which occurs in individuals with bulbar ALS is known as pseudobulbar affect, and is also seen in patients with other neurological problems, including strokes and head injuries. When it produces a tendency to cry, it must be distinguished from depression, which is also common in ALS. Patients with emotional lability often laugh or cry uncontrollably when things are only mildly funny or sad. This is not something that these individuals can control, and it may occur in individuals who have not been particularly emotional in the past. This can be a social embarrassment, and can lead to other people misinterpreting how the individual with ALS really feels. It can also interfere with normal social interaction, because the individual will burst out laughing or burst into tears with little or no warning, and often is unable to stop laughing or crying for quite some time.

The usual treatment is with antidepressants. Tricyclic antidepressants are the most established treatment, and have been used for many years. The strongest is amitriptyline (Elavil), although nortriptyline and imipramine are also used. These medications are available in 10, 25, and 50 milligram (mg) tablets. They may have a number of side effects, including sedation, confusion, an excessively dry mouth, constipation, urinary retention, a rapid heart rate, heart palpitations, and blurred vision. Because of the potential for sedation, they are commonly taken at bedtime, beginning at a very low dose such as 10-25 mg, and increasing as tolerated, up to 75, 100, or even 150 mg. Their effectiveness at producing sedation and drying up the mouth makes them particularly useful for individuals with bulbar ALS who are having difficulty sleeping and have and who sialorrhea (excessive, uncontrolled saliva production). However, sedation and confusion can be severe in some individuals, particularly in the elderly. Other antidepressants which are often used include sertraline (Zoloft) 50-200 mg/day, paroxetine (Paxil) 20-60 mg per day, venlafaxine (Effexor) 75-225 mg per day, citalopram hydrobromide (Celexa) 20-40 mg per day and bupropion hydrochloride (Wellbutrin) 100-300 mg per day. These other antidepressants are less sedating and have less anticholinergic side effects than the tricyclic antidepressants, so are often better tolerated. They can be combined with another medication to dry up saliva or to help with sleep if needed.