

Bulbar ALS: Unique Management Issues

Winter 2002

Zachary Simmons, MD

“Bulbar ALS” is a term commonly used by physicians and patients to refer to those individuals 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 today’s column, I would like to discuss the first of these. The others will be addressed in future columns.

Poor control of saliva, often associated with drooling, is termed sialorrhea. Individuals with ALS do not produce more saliva than other people. However, those with bulbar involvement have difficulty moving their mouth and tongue in such a way as to effectively prevent the saliva from coming out of their mouth, and they have difficulty swallowing their saliva. These problems are particularly severe for many when they are trying to eat or when they are sleeping at night. As a result, drooling often occurs, requiring the person to wipe their mouth frequently with a towel or tissue, or to sleep with a towel on their pillow. Sialorrhea is certainly very inconvenient, and often embarrassing. It is associated with an increased risk of aspiration (material going down the windpipe into the lungs, potentially causing pneumonia). A suction device may be helpful, but usually is used more for thick secretions which become caught in the mouth and cannot be brought out. Control of sialorrhea usually is aimed at decreasing the

amount of saliva produced. The medications used were not designed for ALS, but are used in ALS because they dry up secretions as a side effect. This is known as an anticholinergic property. We most commonly use glycopyrrolate (Robinul), which is available in 1 mg or 2 mg tablets. A tablet is taken 3 to 4 times a day, with a maximum dose of 2 mg, 4 times a day. Tricyclic antidepressants are also useful, and may serve double-duty by also helping with emotional lability. The strongest is amitriptyline (Elavil), although nortriptyline and imipramine are also used. These are available in 10, 25, and 50 mg tablets. Because they are somewhat sedating, they are commonly taken at bedtime, beginning at a very low dose, and increasing as tolerated, up to 75, 100, or even 150 mg. Other commonly used medications for sialorrhea are atropine (Sal-Tropine), diphenhydramine (Benadryl), scopolamine patch (Transderm Scop patch), propantheline bromine (Pro-Banthine), trihexyphenidyl (Artane), and benztropine (Cogentin). All anticholinergic agents may produce sedation, and some cause confusion. An excessively dry mouth, constipation, urinary retention, a rapid heart rate, heart palpitations, and blurred vision are other common side effects.

When these medications are ineffective or the side effects are intolerable, botulinum toxin injections into the parotid glands have been tried to decrease saliva production. The parotid glands are salivary glands located in front of the ears. This is an outpatient procedure, and takes only a few minutes. The effectiveness of the injections is temporary, usually a few weeks to a few months, and injections are repeated as needed. We are in the process of arranging an experimental protocol to allow us to do this for our ALS patients. As a last resort, radiation therapy to the parotid gland can be tried. While this may be very effective, some patients develop a sore throat, and in others the effectiveness is only temporary.

Some individuals with ALS prefer to avoid all these treatment, or have had intolerable side effects or a poor response to these therapies. As a result they prefer simply to carry around a towel or box of tissues. However, for many individuals with bulbar ALS and sialorrhea, there are effective options.