Bulbar ALS: Unique Management Issues (Part III)

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"Bulbar ALS" is a term commonly used to refer to those patients in whom ALS affects muscles of speech and swallowing. 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 previous newsletters, I discussed items 1 and 2. In today's column, I would like to discuss impaired swallowing.

Difficulty swallowing, or dysphagia, is often first characterized by episodes of coughing and choking when attempting to swallow thin liquids such as water, juices, or coffee. Individuals with ALS may also notice that they sometimes choke on their own saliva. As the disease progresses, there may be progressively more difficulty swallowing solids as well. Eventually, the ability to swallow may be lost altogether. Why is dysphagia a matter of concern? There are a number of reasons: Dehydration may result from decreased intake of water. Weight loss is common, and may be severe, resulting in malnutrition. Dehydration and malnutrition make individuals with ALS feel even weaker than they would from the disease itself, leading to exhaustion and a decrease in overall quality of life. Individuals with dysphagia are at high risk for food and fluids going down the trachea (windpipe) rather than the esophagus (the pathway to the stomach). This process, known as aspiration, may lead to pneumonia.

We use several steps to assess an individual's difficulty swallowing. If individuals report difficulty swallowing, episodes of choking, mealtimes greater than 30 minutes, or a loss of 10-20% or more of their usual body weight, then evaluations by a speech pathologist and nutritionist are called for. A nutritionist can assess whether the intake of fluids and calories is adequate, which the speech pathologist can test the ability to swallow. Based on their assessments, initial recommendations usually involve changing food consistencies and changing swallowing techniques. For example, the avoidance of thin liquids, the use of thickeners, moistening foods (using gravy and sauces), and a switch to a chopped or pureed diet are often recommended. Other recommendations commonly are smaller but more frequent meals, an

increase in fat intake, high calorie supplements (for example, Sustacal, Boost, or Ensure), and the use of a chin tuck maneuver when swallowing.

Eventually, such measures may not be successful. In that case, individuals should consider having a feeding tube placed. Most commonly, this is a percutaneous endoscopic gastrostomy (PEG). To put in such a device, the patient is partially sedated, a flexible scope is put down the throat and into the stomach, and a small hole is put through the abdominal wall into the stomach, through which the tube is inserted. Feedings can be started within hours after the placement of the tube. We commonly keep patients in the hospital one night, and then send them home the next day. Feedings can be done by the patient him/herself if they have adequate arm and hand function, or by a caregiver. Weights can be monitored, and the number, volume, and timing of feedings can be adjusted by a nutritionist to maintain or even increase weight. The placement of a PEG does not prevent the individual from taking in some food and fluid by mouth, as long as the speech pathologist believes that this is still safe. Thus, individuals often continue to take small amounts of nutrition by mouth for enjoyment, such as ice cream and applesauce, while obtaining most of their calories through their PEG.

There are some risks associated with the placement of a PEG. If breathing muscles have been markedly weakened by ALS, there is the risk that the individual could stop breathing during the placement of the PEG, requiring some emergency measures. Thus, it is best to have a PEG placed before the breathing muscles have become severely weak. Bleeding, infection, spasm of the larynx (muscles in the back of the throat), and pain at the PEG site are potential complications as well. However, for most individuals, the procedure goes smoothly, particularly if done by an experienced specialist. While no treatment will be ideal for everyone, I have found that almost all individuals who have had a PEG placed have told us later that they thought this was an excellent idea, and would make the same decision if given the choice again.