Saliva, Swallowing, and Breathing: The Ultimate Challenge of Amyotrophic Lateral Sclerosis

We have known for many decades that patients with amyotrophic lateral sclerosis (ALS) with weakness that initially affects the muscles of speech and swallowing (bulbar onset) have a worse prognosis than those patients with limb-onset weakness.[1] A higher likelihood of aspiration, greater difficulty clearing secretions, and, ultimately, a markedly elevated risk of respiratory failure and death are the result of early bulbar involvement in individuals with ALS. Unfortunately, developing medical treatments for ALS has been an extraordinarily difficult scientific challenge, largely because the cause of most cases of ALS is not known.[2]

Currently available medications for ALS treatment have a relatively small effect on survival in ALS.[3] Because respiratory failure is the leading cause of death and medications for the treatment of ALS have a limited effect at this time, it is not surprising that ventilation and noninvasive ventilation (NIV), in particular, have been extensively used. NIV has been shown to result in a significant prolongation of survival and quality of life for individuals with ALS, in fact, greater than medications at the current time.[4-6] However, NIV, whether using a mask or a mouthpiece, requires open access to gas-exchange units in the lungs through the "gates" of the upper airway, the bulbar muscles. For NIV to be effective, the bulbar muscles must be functional enough to allow a safe pathway for air flow to occur without increasing aspiration of secretions or inducing blockage of air flow into the lungs entirely. Unfortunately, with bulbar ALS, this is not always the case.

In this issue of RESPIRATORY CARE, Cazzolli et al[7] presented the use of a straightforward measurement tool, the oral secretion scale, to assess bulbar muscle function. They used the scale to assess the ability of subjects with ALS to use NIV and to prognosticate survival. The oral secretion scale measures both the amount of salivary secretions coming out of the mouth as well as the ability of the individual to swallow secretions. The measurement was previously validated for inter- and intra-observer reliability in two different countries with a variety of health-care providers. On a first read, the scale seems somewhat wordy; however, the principle of the scale is very straightforward and becomes easier with use over time. In essence, if saliva is not being handled automatically through reflex swallowing and instead frequent oral suctioning and use of paper or cloth absorbent material in or near the mouth is required, then the individual has impaired bulbar function that is likely to prevent effective use of NIV. I trialed the use of the oral secretion scale in my own clinic (with a printed copy in hand) and found it relatively easy to use. Unlike the commonly used ALS Function Rating Scale Revised, the oral secretion scale combines secretion and swallowing criteria and thus gives an overall functional assessment of bulbar muscle impairment in one number.[8]

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Cazzolli et al[7] applied the oral secretion scale measurement to a group of individuals with ALS over an extended period of time and showed that a higher (better) score on the oral secretion scale was predictive of the ability of the individual with ALS to use NIV effectively and that a lower score was predictive of the inability to use NIV and a higher mortality. This fits with previously published data[4,5] in this area that looked at shorter follow-up periods. The article describes a measurable value of bulbar function that should be a useful tool to the clinician caring for patients with ALS.[7] My one caveat to the use of the oral secretion scale is that, even if an individual with ALS has a low score, which suggests an inability to effectively use NIV, then an "*n* of 1 trial" of an NIV device in the clinic or in the home is still warranted. Some patients with ALS (albeit a small number) and with advanced bulbar dysfunction are able to use NIV effectively despite difficulty with secretions and swallowing. In this group, NIV can sometimes result in a greater survival time to make advanced-care planning decisions and spend time in a meaningful way with loved ones. In a disease with no cure, this is a real blessing.

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