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EDITORIAL

The yin and yang of gastrostomy in the management of ALS

Friend or foe?

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Though we do not yet have a cure for amyotrophic lateral sclerosis (ALS), we can provide treatment, and the host of medical and other interventions provided by ALS specialists and multidisciplinary care teams increases survival and substantially improves quality of life for patients and their families. Dysphagia is one of the most consequential symptoms in ALS, and ultimately affects the majority of patients. It causes dehydration, weight loss, choking, and chronic aspiration, which substantially increase the risk of potentially fatal aspiration pneumonia. Weight loss alone worsens progression and survival in ALS,[1] presumably because caloric deficit due to dysphagia promotes muscle catabolism, adding to underlying muscle loss from motor neuron death, thereby accelerating weakness and decline. In attempts to circumvent this cascade, enteral tube feeding has been used in the management of ALS for many years, principally via percutaneous endoscopic gastrostomy (PEG).[2] However, gastrostomy tube placement can be hazardous in advanced cases, as impaired respiratory function may increase the risk of respiratory arrest during sedation, as well as ventilator dependence postoperatively. Consequently, gastrostomy tubes are typically only recommended for those patients having forced vital capacity (FVC) >50% of the predicted normal value.[3,4]

Despite widespread adoption, however, there are no randomized controlled trials proving the superiority of PEG over other enteral feeding techniques, or over oral feeding with blended foods. Numerous observational studies suggest a survival benefit with PEG, but this has not been definitively proven.[5–8] This putative survival benefit is constrained by the severity of the disease,[9] weight loss before PEG,[6,7,10] age,[10,11] and, most importantly, poor respiratory function (FVC <50%) at the time of PEG placement.[9,10,12] Increased caloric intake after PEG was associated with improved survival in several studies,[5,7] and quality of life (QOL) scores improved following PEG insertion in one study,[13,14] though not in another.[15] Gastrostomy insertion is relatively safe in patients with ALS overall[6,10] (with severe complications occurring in about 2% of cases, independent of the specific method used)[10] and, after tube placement, 75% of patients live 6 months or longer.[16] In recent years, a less invasive method, the radiologically inserted gastrostomy, has been increasingly employed; it is less invasive than PEG and appears otherwise equivalent, but could be safer (theoretically) in patients with respiratory dysfunction, as it employs a smaller tube and usually requires less sedation.[2]

However, not all studies have shown benefit, and one study of 331 patients enrolled in other ALS clinical trials reported a more rapid decline and an increased mortality risk associated with gastrostomy.[17] In this issue of *Neurology*®, McDonnell and coauthors[18] explored whether gastrostomy affects survival and QOL in a database of 481 patients with ALS enrolled at multiple sites in a nationwide trial of ceftriaxone for the treatment of ALS. Gastrostomy was implemented in 47% of these patients. No evidence of improved QOL was seen in this cohort and, furthermore, survival was worse for this group than for those not undergoing gastrostomy. Importantly, approximately one-third of those receiving gastrostomy had a FVC ≤50% at the time of the procedure, which may explain the adverse association with survival.[18] Some limitations of the study include its retrospective, nonrandomized design, incorporating data from multiple sites collected during the course of another study (i.e., not population-based), and the lack of reports/analysis of the types of gastrostomy employed or the frequency of ventilation and type of ventilation needed during the procedure (noninvasive vs invasive), among others.

Notwithstanding these results and the lack of Level I evidence, gastrostomy appears safe and likely beneficial for appropriately selected patients with ALS, based upon a preponderance of the available data, as well as the clinical experience of many prominent ALS centers over many years. However, its benefits are greatly dependent upon judicious patient selection, appropriate timing of the procedure, and management during and after the procedure. An

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overly aggressive policy for gastrostomy placement in patients with ALS may shorten their survival and increase the risk of death, particularly in patients already having respiratory compromise, while excessively early placement exposes them to some procedural risk,[19] and may decrease QOL during what should be their best remaining months. Ideally, patients should have dysphagia, or substantial weight loss or aspiration, prior to gastrostomy, but should also have preserved respiratory function with FVC >50%. Some patients with early or rapid respiratory decline and FVC >50% may also be considered candidates even in the absence of dysphagia, as they might be disqualified by FVC criteria by the time dysphagia becomes apparent, and patients with severe chronic aspiration alone might also qualify in the absence of dysphagia, to decrease the risk of aspiration pneumonia. During the procedure itself, light sedation is often employed, but in persons with respiratory function approaching the critical threshold, local anesthesia alone may be considered. If ventilatory support is required intraoperatively, noninvasive ventilation is preferred, with intubation used only if absolutely necessary. Without careful attention to these critical details, the procedure can be deleterious and may even hasten the patient's death. Future studies should focus on better methods to ensure appropriate nutrition and to control and circumvent dysphagia and aspiration in patients with ALS.

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DISCLOSURE

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