Case in Point

Amyotrophic Lateral Sclerosis, Nutrition, and Feeding Tube Placement

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This case report details the challenges of sedation and feeding tube placement in addressing the risk of malnutrition in patients with amyotrophic lateral sclerosis.

Amyotrophic lateral sclerosis (ALS) is a degenerative motor neuron disease, characterized by progressive muscle wasting, leading to paralysis of all voluntary muscles, including those used for swallowing and respiration.[1-3] Malnutrition is estimated to develop in 25% to 50% of patients.[4] Malnutrition can lead to further muscle weakness and immunodeficiency as well as a shortened lifespan.[1,5-7] The risk of death may be increased by as much as 3.5 times for malnourished patients compared with those receiving adequate nutrition.[6] To combat malnutrition risk, the American Academy of Neurology (AAN) recommends an evaluation of the nutritional status of ALS patients every 3 months.[8] The AAN also advises consideration of feeding tube placement when patients demonstrate swallowing difficulty or alterations in nutritional status.[8] Feeding tubes can provide a safer, more dependable route for nutrition.

Two types of feeding tube placements are typically discussed with ALS patients: percutaneous endoscopic gastrostomy (PEG) and radiological inserted gastrostomy ([RIG] sometimes referred to as percutaneous radiological gastrostomy [PRG]).[3] These tubes differ in the method of placement. During a PEG procedure, an endoscope is passed through the mouth into the stomach to visualize the site of tube placement. The RIG procedure uses x-ray imaging to locate the tube insertion site.

Typically, at our facility as well as most others, both procedures use moderate sedation and local anesthesia with good success. A small number of other facilities note using RIG without sedation in this population to prevent difficulties associated with the passing of the endoscope and sedation use.[9-11] These issues will be addressed later in greater detail.

CASE PRESENTATIONS

Case 1: Mr. N

Mr. N was diagnosed with ALS in 2007 after noticing progressive weakness in his left upper extremity as well as muscle spasms and lower extremity stiffness contributing to a change in gait. Just 6 months after disease onset, Mr. N presented to his neurology appointment in a wheelchair due to fatigue with ambulation. He reported a change in his speech as well as recent choking on foods and saliva.

During this time, Mr. N's forced vital capacity (FVC) was measured at 59% of predicted. A dysphagia evaluation revealed mild oropharyngeal dysphagia. A mechanical diet (ground texture) with nectar thickened liquids was advised.

PEG tube placement was discussed with Mr. N's physician in December 2007, with Mr. N deciding not to consent to the procedure at that time. He stated that he wished to give the procedure more consideration and requested follow-up. The gastroenterologist examining Mr. N noted that he was "a good candidate for a PEG tube attempt."

About 2 months later, after having increased difficulty swallowing and poor oral intake resulting in a 21-pound weight loss in 6 weeks (12% of previous body weight), Mr. N decided to undergo the PEG procedure. PEG was attempted in February 2008. Per one physician report, Mr. N did not tolerate conscious sedation, becoming agitated, and further sedation was not administered due to Mr. N's poor pulmonary reserve. Another report also noted that sedation was not tolerated due to a drop in oxygen saturation (OS) accompanied with elevations in heart rate and blood pressure. An additional report noted that he did not tolerate the supine position.

Mr. N was admitted to the hospital for observation. Tube placement was performed the next day under general

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anesthesia. During the procedure, Mr. N was intubated and spontaneous ventilation was used. No complications were noted. Per the anesthesiology report, Mr. N's OS remained at 100%.

Mr. N was discharged home shortly after successful placement. Three days postdischarge, he presented to the emergency department. Symptoms consisted of weakness, including limited control of his neck muscles, shortness of breath, fever, and poor oral intake. Mr. N was unable to speak on arrival and was only able to move his upper extremities against gravity with no movement in his lower extremities.

He was found to be in respiratory failure. Noninvasive ventilation was first attempted with subsequent intubation. He was admitted to the medical intensive care unit. The physician noted that Mr. N was suffering from septic shock secondary to aspiration pneumonia. Ten days later, due to progressively worsening respiratory function, Mr. N received a tracheostomy for continued invasive ventilation.

Soon after, erythema and discharge were noticed around the PEG site, indicating an infection. This did not improve quickly, and nasogastric feedings were initiated due to continued site infection not allowing use of the tube.

Case 2: Mr. L

Mr. L was diagnosed with ALS in August 2009. Reports note that Mr. L was in his usual state of health until June 2009. The progression of his disease was rapid. His wife reported that by November 2009, Mr. L was totally dependent for daily activities, including self-care and mobility.

Mr. L consented to a feeding tube for comfort care but declined all measures for artificial life support. The pulmonary service evaluated Mr. L and recommended he begin noninvasive positive pressure ventilation (NIPPV) as well as a sleep study and full pulmonary function tests at a later date. Later, he presented to his gastrointestinal appointment with shortness of breath. He was later admitted to the hospital and found to be in respiratory acidosis. Neurology service noted that he was at an increased risk of developing respiratory failure due to the sedation used with the PEG procedure. A gastroenterologist discussed PEG with Mr. L and the use of minimal sedation to decrease the risk of respiratory failure. Additionally, a respiratory therapist was requested to be present during the procedure. A PEG tube was placed in December 2009, at about 11 AM.

On return to the spinal cord injury unit early in the afternoon, Mr. L's oxygen level desaturated to 88% and the respiratory service was called. Mr. L refused to use NIPPV. An hour later, his pulse oximetry showed an OS of 91%.

At 8:00 PM that evening, his OS was 93%. At approximately 11:30 PM, respiratory service placed Mr. L on NIPPV. Per the respiratory therapist report, Mr. L was compliant and wearing his device. On checking vitals at 4:30 AM, Mr. L was found to be unresponsive. He was pronounced dead at 5:40 AM with cause of death as respiratory failure secondary to ALS.

DISCUSSION

Although feeding tubes have the potential to provide a safer route for nutrition, the decision to receive a gastrostomy tube, whether endoscopically or radiologically placed, should not be taken lightly. Feeding tube placements, especially those that use sedation, are major nonsurgical procedures.[12,13] Multiple potential complications exist. Minor complications include wound infection, wound leakage, cutaneous or gastric ulceration (in those patients with a rigid bolster), pneumoperitoneum (secondary to stomach inflation and gastric needle puncture), temporary ileus, gastric outlet obstruction (due to tube migration), and persistent gastric fistula posttube removal.[14-17] Major complications include esophageal or gastric puncture, necrotizing fasciitis, buried bumper syndrome, aspiration, peritonitis, colocutaneous fistula, other tube misplacements, and death.[15-17]

Deaths related to feeding tube placement complications are not well studied. A portion of these deaths can be linked to the use of sedation. Using a sedative to ease patient anxiety and resistance to the procedure can be beneficial, but sedation comes with risks.[18,19] Sedatives naturally slow the central nervous system (CNS), depressing protective reflexes such as the gag reflex. A high mortality rate is associated with periprocedural aspiration that is often a result of sedation impairing this reflex.[12] In a study of mortality connected with PEG tube placement, 40% of deceased patients had postoperative aspiration pneumonia.[20] Patients with neurologic abnormalities are even more prone to this complication. Managing oral secretions that may precipitate aspiration can be particularly difficult with ALS patients.[21]

Sedation also depresses respiratory function and the cardiovascular system. Cardiopulmonary events related to sedation and analgesia are a frequent cause of endoscopy-related death. The severity of these complications range from transient, minor oxygen desaturation to life-threatening events such as apnea, hypotension, and myocardial infarction. Severe oxygen desaturation is rare, but some level of desaturation is estimated to occur in up to 70% of patients undergoing endoscopy procedures.[19] The desaturation may be related to the

variance in patient response to sedatives, creating issues with dosing that may lead to oversedation that further depresses the CNS.[18]

In the largest study of PEG procedure-related mortality, an analysis of the deaths of 719 patients listed sedation as a major risk factor.[20] Ten percent of studied patients received a sedation reversal medication, indicating oversedation. Three percent experienced hypoxemia (OS ≤ 90%) during the procedure.[20] The mortality analysis showed that 95% of the studied patients died of cardiovascular or respiratory disease with respiratory failure being the number 1 cause of death (51% of patients).[20] No patients diagnosed with ALS were studied.

To lower the rate of complications, the research suggested the use of perioperative antibiotics, meticulous postprocedure care, and appropriate patient selection. The researchers recommended that endoscopy units perform regular audits, reviewing all 30-day postprocedure mortalities. They also acknowledged that appropriate selection of patients remains a difficult and complex problem.[20] Staffordshire General Hospital in Stafford, United Kingdom, demonstrated that this difficultly in patient selection can be reduced by using a multidisciplinary nutrition team to assess potential PEG-placement patients.[13]

Literature regarding feeding tube placement complications, specific to ALS patients, is limited. A relatively small number of studies have attempted to address this research deficit, and most are focused on comparing the complication rates on the 2 most-used feeding tube placements, PEG and RIG.

A prospective study of 50 ALS patients comparing outcomes of PEG and RIG procedures found that the frequency of all complications during placements and the first month of follow-up were not significantly different between the 2 procedures.[5] Survival rates were also shown as similar.[5] A retrospective study also examining 50 patients compared the 2 procedures and found improved survival in patients receiving the RIG.[22] Another retrospective study of 40 ALS patients concluded that successful and well-tolerated placements were more frequent with the RIG than the PEG.[23] In this study, all 3 patients who failed to successfully undergo the PEG procedure (2 of the 3 patients experienced respiratory decompensation) safely underwent RIG placement.[23]

According to the Oxford Motor Neuron Disease Care and Research Centre, the PEG procedure is considered standard and reliable when respiratory evaluation is adequate.[10] But the adequate evaluation of proper timing in the course of the disease to receive a PEG has not been shown.[7]

The AAN recommends PEG be performed before the patient's FVC falls below 50% of predicted.[8] The sedation, coupled with prolonged supine positioning, may be hazardous when FVC is low. A drop in OS may necessitate intubation, and the risk of mortality increases.

However, FVC is not a precise forecaster of early respiratory function decline. Some research suggests using FVC of < 65% to 70% of predicted as a better predictor.[24] Other literature notes that patients can develop respiratory failure with FVC of 70% of predicted.[25] Therefore, using FVC of at least 75% may be a more appropriate marker. A review of enteral feeding research by *The Cochrane Collaboration* noted that although more research is necessary, the earlier in the course of the disease a patient receives a PEG, the longer the survival period postprocedure with all other factors being equal.[6]

A PEG placement may still be performed successfully in ALS patients with declining respiratory function. Using NIPPV during the procedure may allow patients with a lower FVC to safely undergo a PEG.[26] Two retrospective studies reviewed PEG placements in ALS patients with FVCs from 7% to 52% of predicted where NIPPV was used. Few complications and no PEG-related mortality were reported.[6]

And the use of NIPPV with the RIG procedure, especially when used without sedation, may provide an even safer means of feeding tube placement. One study group successfully placed tubes using NIPPV and foregoing sedation in ALS patients whose mean FVC was < 30% with no major complications.[11] The study concluded that the RIG is the superior procedure due to its ability to manage the 2 major issues with ALS, aspiration risk secondary to bulbar weakness and declining respiratory function.[11]

Also, in patients with rapid disease progression, the risks and benefits of feeding tube placements should be even more carefully weighed. Alternate feeding routes such as nasogastric feedings or parenteral nutrition (PN) may be more beneficial. Nasogastric feedings may be uncomfortable for many patients, even with short-term

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use, and may not be conducive for quality of life. The use of home PN for this population has been scarcely studied, but may be a practical alternative. A prospective study of 30 patients receiving home PN concluded that this feeding method may be a valuable tool for quality of life for patients with severe respiratory failure unable to safely undergo a PEG procedure.[27]

SUMMARY

A variety of complications are possible with PEG feeding tube placement with some of these complications being fatal. In specific groups of patients, as exemplified by the complications in the patients with ALS reported in cases 1 and 2, there may be a higher rate of complications than has been commonly documented. Although we cannot make well-defined conclusions from 2 case studies, potential complications posttube placement are evident here, and the cases bring to light the need for additional research regarding feeding tube placements in this population.

Further research, especially those with larger sample sizes, is necessary to identify which method of feeding tube placement as well as which method for delivery of nutrition is best for patients with ALS. Better guidelines for indications and contraindications for each feeding tube placement technique are also needed. The focus should be on anticipating complications and providing safe nutrition support for this complex population.

As an integral part of the ALS multidisciplinary team, dietitians should be versed in the benefits as well as the potential risks of tube feeding placement. We should offer assistance in patient-procedure selection, helping to determine which route of nutrition delivery and feeding tube placement technique is optimal for our patients. It is our job to advocate for the best nutritional care of our patients, including decisions related to the nutrient delivery. ●

Author disclosures

The author reports no actual or potential conflicts of interest with regard to this article.

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