Anesthetic Management with Laryngeal Mask Airway during Percutaneous Gastrostomy in Duchenne Muscular Dystrophy

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Introduction: Duchenne muscular dystrophy (DMD) is the most common myopathy occurring in pediatric patients, with cardiac and pulmonary insufficiency eventually leading to death. While elective surgery is typically considered contraindicated in the face of decreased respiratory or cardiac function, these children are more regularly presenting for the elective placement of gastrostomy feeding tubes, as undernutrition occurs from progressive dysphagia, often late in the course of their disease. In the past we performed percutaneous gastrostomy tube (PEG) insertions in DMD patients utilizing a new technique of respiratory support with nasal or oral non-invasive positive pressure ventilation (NPPV), and now we progress to placement of a laryngeal mask airway (LMA), preventing hypoxic episodes and allowing for improved airway management. We have performed six such procedures, and here we provide two representative case reports.

Case Reports: The first patient is a 20 year old male with DMD chronic respiratory failure on nightly BiPAP and poor cardiac ventricular function. Airway was Malampati class IV with lingual hypertrophy. The second patient is a 21 year old male with DMD and chronic respiratory failure on 24-hour nasal BiPAP. His airway was Malampati class III. Each patient was scheduled for a PEG to improve poor nutrition secondary to progressive dysphagia. Standard monitors were placed, and the patients’ lungs were preoxygenated. Intravenous medications included midazolam, propofol by infusion and ketamine for analgesia. The patients’ remained spontaneously ventilating with NPPV in place, on usual settings, until eyelash reflex was abolished. The non-invasive device was then discontinued, and an appropriately sized LMA was placed. A 1 cm external diameter, well lubricated gastroscope was placed in the mouth behind the LMA and passed into the esophagus by the gastroenterologist. (Figure 1) Partial deflation of the LMA cuff may have facilitated passage of the endoscope. Ventilation was assisted as necessary to maintain PaCO$_2$ 35-40. SpO$_2$ was maintained above 90%. There was no displacement of the LMA during placement of the endoscope or the gastrostomy tube, even when multiple attempts were required. Upon conclusion of the procedure, the LMA was removed under deep sedation with spontaneous ventilation, and the NPPV device was replaced. Patients were monitored in the pediatric intensive care unit overnight, and all were discharged home in 24 hours.

Figure 1
Nasal BiPAP mask in place, with LMA positioned appropriately, and gastroscope passing easily behind.

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Discussion: PEG insertions under local anesthesia with sedation are a poor choice in DMD patients with severe weakness, because there is a high risk of hypoventilation during conscious or deep sedation. Intubation may be difficult secondary to lingual hypertrophy. These patients also have diminished pulmonary reserves and inability to clear secretions secondary to weak cough. Severe pulmonary dysfunction is further compromised during general endotracheal anesthesia by weakening or fatigue of inspiratory and expiratory muscles; atelectasis may lead to mucus plugging. These otherwise benign events may lead to acute respiratory failure, pneumonia, and prolonged hospitalization. We confirm that the efficacy of the LMA seal is increased with the gastroscope in situ. We did find, however, that manipulation of the LMA was occasionally necessary to facilitate passage of the endoscope, and the cuff was routinely deflated during passage of the scope past the oropharynx. Our cases presented here demonstrate that PEG tube insertion is feasible and safe without tracheal intubation by managing the airway with an LMA.