



Anesthetic Management of a Patient with Limb-Girdle Muscular Dystrophy Receiving an Upper Endoscopy Exam

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Background

- **Limb-Girdle Muscular Dystrophy (LGMD)** is a group of inherited myopathy characterized by a mismatch between muscle breakdown and muscle repair. It typically presents as proximal muscle weakness involving the hip and shoulder girdles. LGMD has a frequency of 1 in 15,000 to 50,000.
 - Autosomal dominant (LGMD1) or autosomal recessive (LGMD2) with further subtypes
 - Severe cases may have cardiac involvement (dilated cardiomyopathy and arrhythmia) or respiratory problems.



Figure 1a): LGMD weakness with bilateral scapular winging, **b) and c)** Distal involvement of LGMD. By Xi et al used under CC BY 4.0 / Cropped from original

- Difficult airway features of LGMD
 - Tongue hypertrophy
 - Delayed gastric emptying due to hypomotility of gastrointestinal tract
 - Spinal rigidity, scoliosis, and limb contractures pose challenges in positioning for intubation
- When managing an anticipated difficult airway, combination techniques such as video laryngoscopy paired with a flexible intubation scope are recommended for greater first attempt success rate of endotracheal intubation.

Case Presentation

A 19-year-old male with a history of LGMD, dilated cardiomyopathy status post heart transplant, and chronic respiratory failure requiring continuous average volume-assured pressure support (AVAPS) presents with lower gastrointestinal bleeding requiring an esophagogastroduodenoscopy and sigmoidoscopy.

Past Medical History

- **LGMD:** patient uses wheelchair to ambulate around home, requires skilled nursing care for assistance for activities of daily living
- **Dilated cardiomyopathy:** rare manifestation of LGMD, underwent heart transplantation 7 years ago, currently on Tacrolimus and Mycophenolate
 - **Transthoracic echocardiogram:** decreased left ventricular (LV) cavity size, normal global LV systolic and diastolic function with normal LV filling, normal right ventricular systolic function, mild mitral regurgitation, EF 60%
- **Chronic respiratory failure:** requires AVAPS delivered via nasal cannula
- **Percutaneous endoscopic gastrostomy (PEG):** placed 2 years ago for nutritional supplementation but patient able to take some food orally

Physical Exam

- **Airway:** Large tongue, Mallampati IV, Interincisive distance >3 cm, hyomental distance >3 fingerbreadths, No loose teeth, Mobilizes neck to neutral position
- **Respiratory:** symmetrical, bilaterally diminished breath sounds, breathing comfortably with AVAPS
- **Musculoskeletal:** contracted upper and lower extremities

Anesthetic Plan and Airway Management

Anesthetic Course of Patient Case

- Given anticipated difficult airway (macroglossia, limb rigidity, aspiration risk), plan for general anesthesia with combined video laryngoscope and flexible intubation scope for endotracheal intubation under total intravenous anesthesia
- Avoid inhaled gases and succinylcholine to minimize risk of triggering hyperkalemia, rhabdomyolysis, and other muscular toxicities given patient's history of LGMD and immobility
- Maintain patient on AVAPS prior to induction to ensure adequate oxygenation
- Patient given 2 mg of midazolam and 50 mcg of fentanyl in divided doses with a propofol infusion
- With patient in head-up position, a size 3 GlideScope was inserted into the oral cavity to provide visualization of the vocal cords
- A 6.0 cuffed oral tracheal tube was placed over a BFP-190 Flexible Tracheal Intubation Fiberscope and directed through the vocal cords with double visualization from the fiberoptic scope itself as well as the GlideScope
- During endoscope placement for procedure, decrease in end tidal CO₂ was noted likely due to endotracheal tube displacement. Fiberoptic scope used to reposition endotracheal tube
- On emergence and extubation, oral suction with Yankauer suction tip performed. Neuromuscular blockade reversed with sugammadex, Adequate recovery of muscular function assessed by hand grasps. Patient extubated while spontaneously breathing and placed back on AVAPS.
- Patient transported to PACU on AVAPS and monitored closely, recovering without incidence.

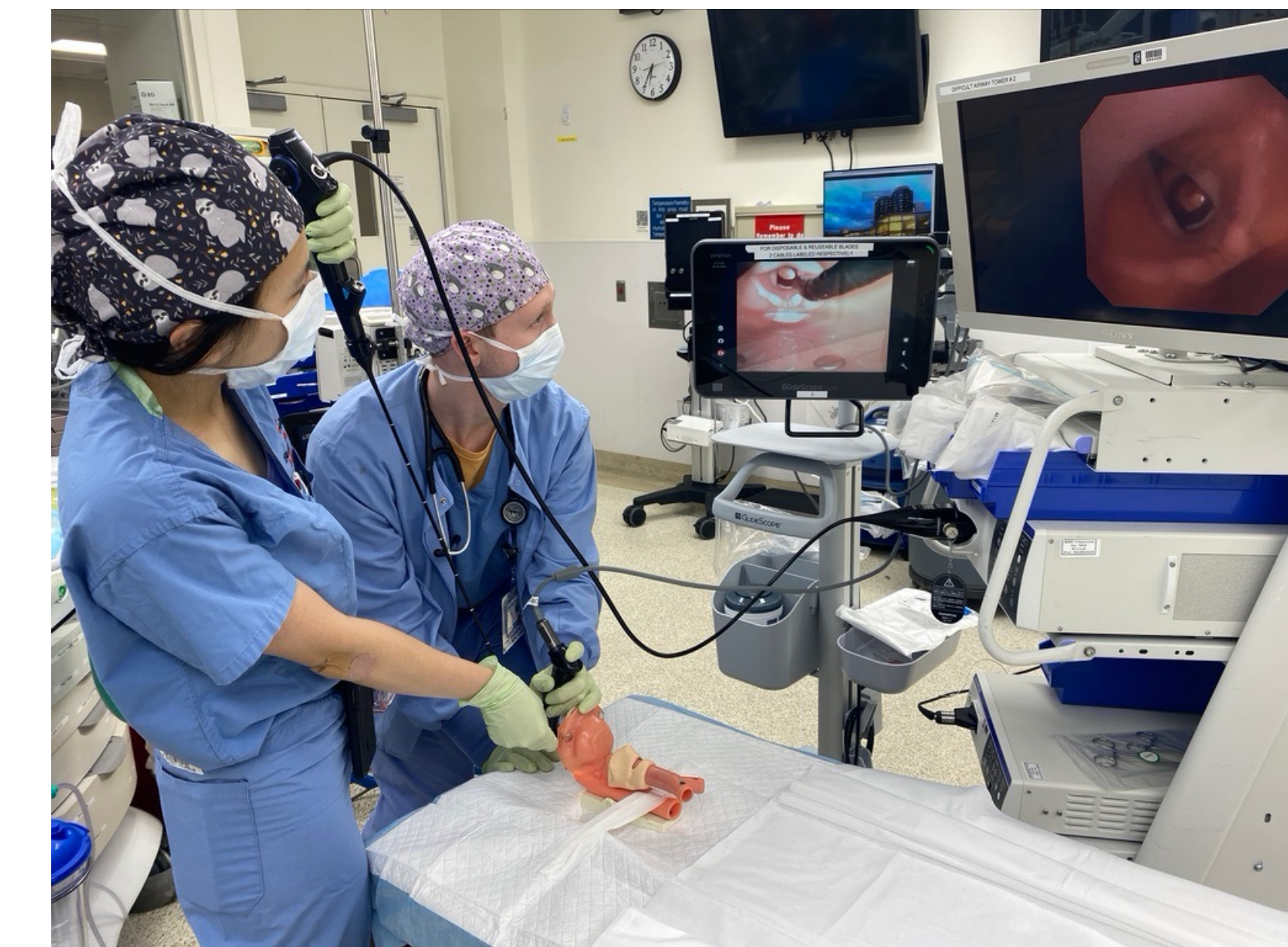
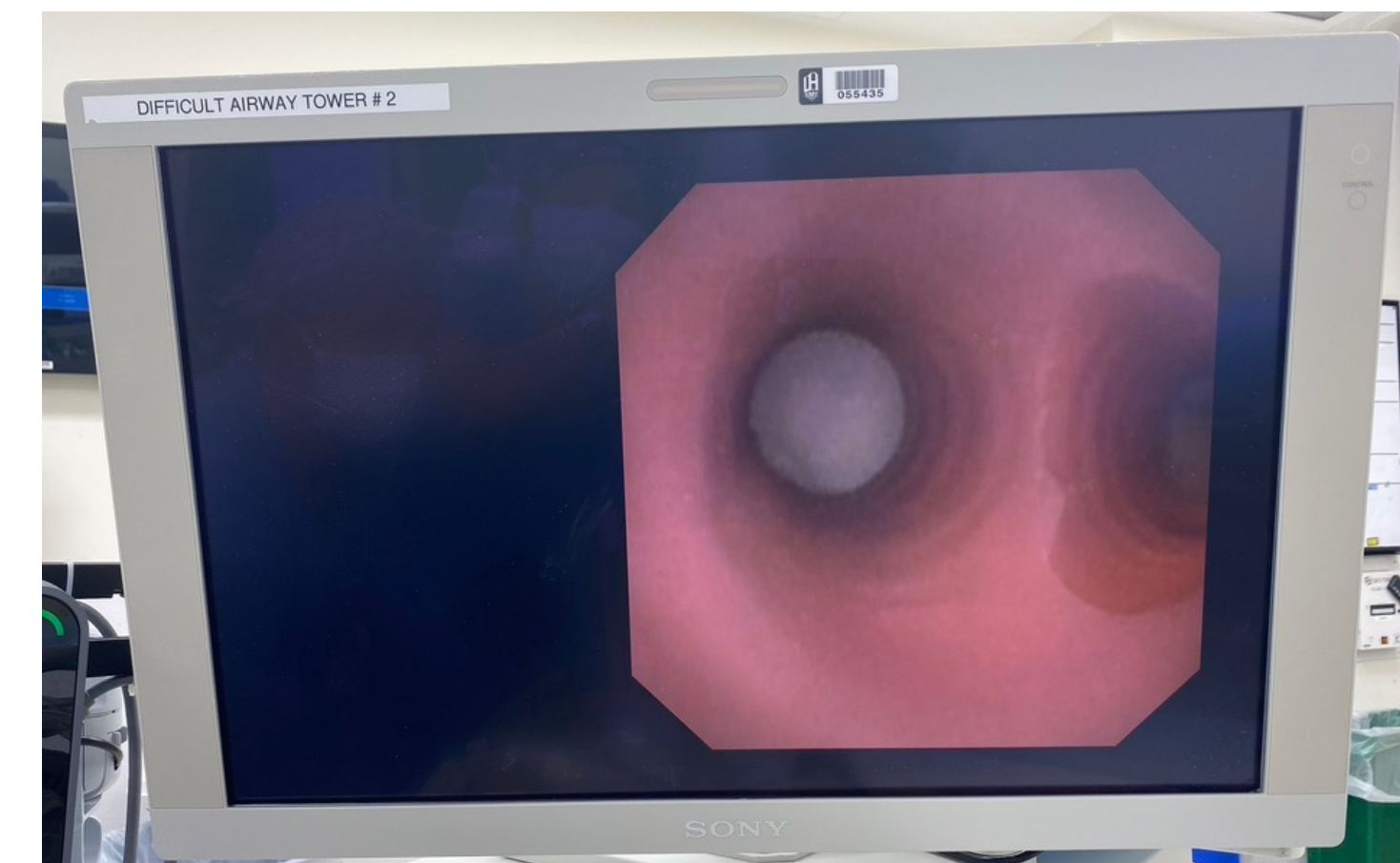


Figure 2: Two-provider combination technique with video laryngoscope and flexible intubation scope. One provider uses GlideScope to obtain visualization of vocal cords, while the other guides the flexible fiberscope through the glottic space into the trachea. Endotracheal tube placement is confirmed by visualization of the tip of tube above the carina.



Conclusion

- When assessing patients with rare diseases like LGMD, it is important to consider features that may lead to difficult airway and prepare accordingly. A thorough review of patient history, careful physical exam, and advanced airway equipment are crucial in these challenging cases
- A combination technique with video laryngoscope and flexible intubation scope allows for enhanced visualization of the larynx and precise maneuvering of the endotracheal tube as well as quick confirmation of tracheal placement.

References

References available on request, correspondence to Ashley.Wang@stonybrookmedicine.edu

