

Background	Case Details (cont'd)	Discussion (cont'd)	
Patients with Limb Girdle Muscular Dystrophy (LGMD) and other RYR1 variants present unique challenges to the anesthesiologist because of muscle weakness and potential for malignant hyperthermia (MH). We describe the anesthetic management of a patient with LGMD and a potentially difficult airway.	(0.7 mcg/kg/min), fentanyl (1-2 mcg/kg), and paralysis with rocuronium (0.6 mg/kg). The patient was managed conservatively in the postoperative period and extubated without incident the following day.	CK are also important in monitoring for development of MH. Dantrolene should be readily available in event of suspected MH. LGMD increases the risk of respiratory compromise under anesthesia because muscle relaxants can exacerbate existing respiratory weakness. Awake-fiberoptic intubation preserves spontaneous ventilation and mitigates the risk of respiratory decompensation associated with muscle relaxation, allowing for a controlled and gradual approach to securing the airway. Additionally, awake-fiberoptic intubation allows for thorough airway assessment and planning before induction. In this case, airway assessment revealed challenges (limited mouth opening, beak-shaped mouth and jaw) that could have complicated conventional intubation. Anticipation of airway challenges with advanced planning for awake-fiberoptic intubation ensured a smooth and safe intubation and successful anesthetic for this patient.	
	Discussion		
	Symptoms Characterized by progressive weakness in proximal limb-girdle muscles. Criteria include elevated CK and degenerative changes upon histological examination. Preoperative Testing Echocardiography, ECG, and pulmonary function testing are often performed. Serum CK is tested to establish a baseline.		
Anesthetic Concerns Volatile anesthetics and succinylcholine increase risk of malignant hyperthermia and rhabdomyolysis. For LGMD patients, cardiac arrhythmias, respiratory insufficiency, altered airway, and increased risk of aspiration can result in cardiopulmonary collapse. Anesthetic Management Local anesthesia is preferred. Alternatively, total intravenous anesthesia with trigger-free anesthetics is recommended. Muscle relaxants are used with caution. Laryngeal mask airway is recommended to avoid intubation when possible. Close temperature management, blood pressure monitoring, and lower thresholds for cardiopulmonary surveillance are recommended.			
Literature on safe anesthetic management of LGMD is limited. We highlight our successful use of TIVA in avoiding malignant hyperthermia (MH) triggers, and use of awake-fiberoptic intubation as an effective technique for securing a difficult airway in a patient with LGMD. Inhaled anesthetics and succinylcholine are contraindicated in LGMD because of an increased risk of MH and of developing rhabdomyolysis and hyperkalemia [1]. Using TIVA avoids the potential triggers of MH while maintaining appropriate anesthetic depth. Interval measurement of lactate and			
Case Details			
A 55-year-old male with LGMD and no prior exposure to anesthesia presented for resection of colon cancer. His LGMD predominantly affected his shoulders. Pre-anesthesia examination and routine blood tests were within normal limits. While LGMD is not associated with difficult airways, there was concern for one because of the patient’s small mouth opening and beak-shaped mouth and jaw. Thus, an awake fiberoptic intubation was performed and the airway was safely secured with a 7.0 mm endotracheal tube. The anesthesia team took precautions for Malignant Hyperthermia (MH) and used a new anesthesia machine to avoid triggers. In the operating room, the patient was positioned at 45 degrees and semireclined. Maintenance of anesthesia was achieved with total intravenous anesthesia (TIVA) with propofol (100 mcg/kg/min), dexmedetomidine			
References			
1. Murphy AP, Straub V. The Classification, Natural History and Treatment of the Limb Girdle Muscular Dystrophies. J Neuromuscul Dis. 2015 Jul 22;2(s2):S7-S19. doi: 10.3233/JND-150105. PMID: 27858764; PMCID: PMC5271430.			