**Título:** ANESTHETIC CONCERNS IN A PATIENT WITH CONGENITAL MYASTHENIA GRAVIS AND SEVERE SCOLIOSIS

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**Resumo:**

Background:

Congenital Myasthenia Gravis (CMG) is a rare heterogeneous disease caused by genetic mutations affecting the neuromuscular junction. Transient or permanent weakness of voluntary muscles frequently begin at birth or early childhood with a wide range of phenotypes and severities. Medical treatment for alleviating symptoms include 3,4-diaminopyridine, albuterol and ephedrine.

Case Report:

A 25-year-old male, 40 Kg, diagnosed with testicular torsion with 4-day evolution was admitted for urgent orchidectomy. Medical history included CMG with permanent weakness of bulbar, axial and limb muscles, severe scoliosis and chronic respiratory failure managed with overnight BiPAP.

After preoperative fasting, he presented in the operating room with severe dyspnea, unable to tolerate dorsal decubitus. A neurologist medical observation was requested and a supplemental oral dose of salbutamol and ephedrine was given. Referral to the Intensive Care Unit was made in case of need of invasive ventilatory support.

Induction of general anesthesia was obtained with a low dose of fentanyl and titrated doses of propofol and maintenance achieved with sevoflurane. I-Gel® Laryngeal Mask Airway (LMA) was inserted, and respiratory support was provided with a pressure-controlled volume-guaranteed mode. Emergence from anesthesia and removal of LMA in a semi-seated position was uneventful. Non-invasive ventilation with BiPAP was initiated at the Post Anesthetic Care Unit and he had medical discharge at the second day.

Discussion:

Management of patients with CMG requires multidisciplinary approach and preparation for surgery. The choice of the anesthetic technique was conditioned by medical history, the risk of postoperative complications and the need for post-operative ventilation support.

Preoperative fasting, careful titration of drugs, avoidance of neuromuscular blockers, advantageous use of the LMA and non-invasive positive pressure ventilation during recovery proved to be a safe strategy to avoid cardiopulmonary complications, minimize respiratory fatigue, allow early return of preoperative muscle function and early discharge.

References:

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