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**BJAN\_2020\_60 - Case report**

**Combined spinal-epidural anesthesia for abdominoplasty and liposuction in Limb-girdle Muscular Dystrophy: case report<sup>a</sup>**

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<sup>a</sup> Study conducted at Hospital São Domingos, São Luís, MA, Brasil.

**Abstract**

We report the anesthetic management with combined spinal-epidural in a patient with limb-girdle muscular dystrophy type 2A, submitted to abdominoplasty and liposuction. The patient had onset of symptoms at 8 years old, diagnosed by muscular biopsy, presenting muscle weakness in the scapular and pelvic girdles, with reduced mobility. We performed monitorization with non-invasive blood pressure, oximeter, thermometer, and electrocardiogram. In the postoperative period, she showed no clinical signs of rhabdomyolysis, myotonia or adverse effects, maintaining hemodynamic stability. The anesthesia technique allowed spontaneous ventilation, monitoring of clinical parameters close to physiological conditions and used smaller doses of medication, reducing related risks.

**KEYWORDS**

Case Reports; Limb-Girdle Muscular Dystrophy; Anesthesia; Abdominoplasty; Perioperative Care

## Background

Limb-Girdle Muscular Dystrophy (LGMD), is a group of rare genetic syndromes involving muscular dystrophies, which preferably affect the muscles of the scapular and hip girdle, leading to progressive weakness and loss of function.[1] Classified as autosomal dominant, with 8 types (1A - 1H), or recessive with 26 types (2A - 2Z), have a prevalence of 1.68 cases per 100,000 inhabitants, the most common being type 2A, representing 15 to 40% of all cases.[1]

The symptoms can start at any age, and get worse over time.[1] It often occurs with high serum creatine kinase (CK) levels, but normal levels may be present.[1,2] The progression of the disease varies widely among individuals, who may have severe impairment of mobility, cardiac and respiratory function leading to decreased life expectancy,[1,2] due to this and the variation in the age range of onset of symptoms, the diagnosis can be difficult and may require genetic testing.[2]

The benefit of diagnosis arises when these patients require surgical interventions, usually orthopedic procedures, since the anesthetic management in these individuals presents some particularities, due to the higher risk of cardiac complications from malign arrhythmias, respiratory complications, malignant hyperthermia (HM) and rhabdomyolysis caused by drugs used in anesthesia.[2,3]

Few cases of operations performed in patients with LGMD were reported or included in reviews,[2,3] and there is no standardization of the anesthetic technique in these patients, only general recommendations, being necessary to individualize the perioperative care according to the degree of involvement to reduce risks and adverse pharmacological effects related to neuromuscular diseases.

Thus, this study aims to report, following the CARE guidelines,[4] the management with combined spinal-epidural (CSE) anesthesia in a patient diagnosed with LGMD type 2A. The patient was submitted to abdominoplasty and liposuction, presented important limb impairment and reduced mobility, requiring adequate monitoring to avoid complications and also ensure the patient's comfort during this period.

## Case report

We present a 35-year-old female patient, diagnosed with LGMD type 2A, who underwent abdominoplasty and liposuction in October 2019. The patient had onset of

symptoms at 8 years of age, with diagnosis at 30 years of age by muscle biopsy with the presence of two pathogenic mutations in heterozygosis in exons 4 and 22, respectively in the calpain gene, characteristic of type 2A. Reported familiar history of LGMD with diagnosis by muscle biopsy and surgical history of hysterectomy with no complications. In the preoperative evaluation, she presented flaccid tetraparesis with proximal muscle strength grade 2 and distal muscle strength grade 4, not being able to walk alone and requiring a wheelchair for movement. The preoperative examinations (electrocardiogram, echocardiogram, pulmonary function test, and laboratory tests) were normal, except for the values of CK and serum creatinine of  $315 \text{ U.L}^{-1}$  and value  $< 0.17 \text{ mg.dL}^{-1}$  respectively, which were following the pattern of LGMD type 2A. The proposed procedure was explained and the patient consented (Table 1).

It was decided to perform a combined spinal-epidural (CSE) neuraxial anesthesia, spinal anesthesia with bupivacaine 20 mg and morphine 0.03 mg, epidural (placement of a catheter for additional doses of anesthetic if necessary) and fentanyl (IV) 100 mcg in association with midazolam 5 mg for sedation. The patient's vital signs were monitored (non-invasive blood pressure, pulse oximetry, electrocardiogram, and axillary thermometer) according to ASA standards and with recommendations for monitoring in patients with myopathies[3] and spontaneous ventilation with a face mask was maintained during surgery. The temperature at the start of the procedure was  $36.5^{\circ}\text{C}$  and a thermal blanket was used to control the temperature; the patient presented a temperature between  $36^{\circ}\text{C}$  and  $36.5^{\circ}\text{C}$  during the procedure. The mean blood pressure (MBP) was between 78 and 97 mmHg, heart rate (HR) was between 80 and 98 bpm, respiratory rate (RR) of 18 breaths/min and oxygen saturation ( $\text{SatO}_2$ ) between 98 and 99% during the procedure.

The surgery lasted 240 minutes, without technical difficulties. Additional doses of local anesthetic were not necessary during the surgery. At the end of the procedure, parecoxib 40 mg, metamizole (dipyrone) 2 mg, ondansetron 4 mg, and dexamethasone 4 mg were administered.

In the postoperative period, hemodynamic stability was maintained without episodes of myotonia or adverse pharmacological effects. The effect of the anesthesia ended without complications. The patient remained in the post-anesthesia recovery unit for 2 h, requiring epidural administration of fentanyl 20  $\mu\text{g}$  and ropivacaine 2.5 mg using patient-controlled analgesia (PCA) dispositive to control postoperative pain.

The patient was monitored in the postoperative period with non-invasive blood pressure, pulse oximetry, thermometer. She presented conditions for discharge on the following day, with MBP between 73 and 93 mmHg, HR between 80 and 90 bpm, RR 18 to 19 breaths/min, SatO<sub>2</sub> 98%, temperature of 36 to 36.7°C, without clinical signs of rhabdomyolysis, with cardiorespiratory stability, adequate pain control and without the necessity of additional doses of epidural anesthetic.

## Discussion

In a review of the literature, few reports of elective surgeries were found, the majority of the operations being performed under total intravenous anesthesia (TIVA), and few cases with neuraxial anesthesia supplemented with an intrathecal opioid, already included in other studies.[2,3] Two operations were performed under neuraxial anesthesia, included in the review by Cao et al.,[2] one laparoscopic cholecystectomy with spinal anesthesia technique and one cesarean section with the CSE technique, which included an intrathecal opioid. They performed neuraxial anesthesia to avoid respiratory complications and the need for intubation, invasive ventilation, and neuromuscular blockade, reducing the risks inherent to patients with myopathies, mainly the potential difficulty of weaning from ventilation.[2] In these cases, when neuraxial anesthesia was performed, a local anesthetic (bupivacaine) and intrathecal opioid (fentanyl) were also used, and adequate monitoring in the perioperative period was maintained[3] and in only one case a patient-controlled analgesia device was used in the postoperative period.[2]

Intrathecal morphine when added to a spinal anesthesia technique, even in low doses, promotes important analgesia, which can extend beyond the first 24 hours of postoperative, reducing the total consumption of opioids. However, it is associated with an increased risk of nausea, vomiting, itching and urinary retention, and, when in higher doses, with a higher risk of respiratory depression. Therefore, it is recommended prophylaxis for these effects and monitoring to detect complications early.[5]

In this case, we chose to perform CSE neuraxial anesthesia with an intrathecal solution containing local anesthetic and opioid in a low dose and treatment of postoperative pain with patient-controlled analgesia by an epidural catheter - ropivacaine (0.5 mg.mL<sup>-1</sup>) and fentanyl (4 µg.mL<sup>-1</sup>), bolus 5 mL, following the recommendations for local anesthesia in patients with neuromuscular diseases whenever possible, despite the possibility of acute toxicity of local anesthetics. CSE anesthesia is

indicated in patients with myopathies because it presents a lower risk of complications and allows spontaneous ventilation. Its advantages include a rapid onset of sensorial and motor blockade, reduction of the need for extra intrathecal doses, allows better control of the duration of anesthesia and holds a desired sensory level.[2,3] The option of associating morphine and local anesthetic aimed to promote an adequate pain control in the perioperative period, as well as reducing the consumption of opioids, following the recommendations of prophylaxis for related adverse effects.

The established anesthetic management contributed to the patient's comfort in relation to the pain control and return of functions, reducing doses by the combination of techniques and avoiding drugs that can be potential triggers for cardiorespiratory complications, HM and rhabdomyolysis. The study had as a limitation the non-performance of gasometry and specific laboratory tests in the postoperative period and because there were no clinical signs that justified it, besides the fact that the chosen anesthetic technique and the smaller surgical procedure represented reduced risks.

Considering the possibility of an unpredictable response to opioids, neuromuscular blockers and their antagonists, cardiorespiratory complications, myotonia, and adverse pharmacological effects can be avoided with the use of low concentrations of these anesthetics.[2,3] It is recommended to use neuromuscular blockers only when there is neuromuscular monitoring and to avoid the use of succinylcholine due to the high risk of rhabdomyolysis and HM.[2,3] It is worth noting that there is no direct contraindication of the use of inhalational agents due to the possible risk of HM, as there are no reports of cases of LGMD type 2A presenting this complication.

Despite the complexity of anesthetic management in patients with MDL, a surgical approach with careful preoperative planning is possible. A comprehensive preoperative evaluation, including a multidisciplinary team, minimizes surgical risks. Also, adequate perioperative monitoring is required to detect early cardiopulmonary complications of the adverse effects of anesthetics.

The reported case demonstrates a technique of CSE neuraxial anesthesia in a patient with LGMD submitted to abdominoplasty and liposuction without complications. The combination of local anesthetic and opioids in low doses facilitated postoperative pain control in combination with the PCA device, in addition to reducing the total doses of the drugs used.

**Ethical approval statement**

This case report was approved by the Research and Ethics Committee of Hospital São Domingos (Number 3.709.613).

**Informed consent statement**

The patient has provided written consent to present the case.

**Conflicts of interest**

The authors declare no conflicts of interest.

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**Table 1 - Clinical progression.**

1992	2008	2015	2016	September/ 2019	October/05/ 2019	October/06/ 2019
Onset of symptoms of muscle weakness in upper and lower limbs	Progressive loss of muscle strength, with need of wheelchair for movement	Muscle biopsy with results for LGMD type 2A	Hysterectomy, without complications, performed with neuraxial anesthesia	Preoperative evaluation and analysis of exams requested	Abdominoplasty and liposuction with CSE anesthesia, with local anesthetic + opioid	Postoperative with maintenance of monitoring, with discharge from hospital after confirmation of hemodynamic stability and no signs of rhabdomyolysis

LGMD, limb-girdle muscular dystrophy; CSE, combined spinal-epidural.