CASE REPORT

Total intravenous anesthesia for aortic aneurysm replacement surgery in a patient with limb-girdle dystrophy

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Abstract We report the anesthetic management with total intravenous anesthesia of a 61-year-old male diagnosed with limb-girdle muscular dystrophy admitted for replacement of ascending aorta due to an aortic aneurysm. Limb-girdle muscular dystrophy belongs to a genetically heterogeneous group of muscular dystrophies involving shoulder and hip girdles. Although the risk of malignant hyperthermia does not seem to be increased in these patients compared with the general population, the exposure to inhaled anesthetics and succinylcholine should probably be avoided because these patients have a predisposition to hyperkalemia and rhabdomyolysis. We chose to use total intravenous anesthesia with propofol, remifentanil and muscle relaxants to reduce oxygen consumption, and later to reduce the doses of propofol and remifentanil. The combination of a carefully planned anesthetic strategy, anesthetic depth, and neuromuscular blockade monitoring is explained.

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KEYWORDS
Limb-girdle muscle dystrophy; Aortic aneurysm repair surgery; Ascending aorta replacement surgery; Total intravenous anesthesia

PALABRAS CLAVE
Distrofia muscular de cinturas; Reparación de aneurisma aórtico; Cirugía de sustitución de aorta ascendente; Anestesia total intravenosa

Anesthesia total intravenosa para sustitución de aorta ascendente por aneurisma en paciente con distrofia muscular de cinturas

Resumen Presentamos el tratamiento anestésico con anestesia total intravenosa de un varón de 61 años diagnosticado de distrofia muscular de cinturas para sustitución de aorta ascendente por aneurisma aórtico. La distrofia muscular de cinturas es un grupo genéticamente heterogéneo de distrofias musculares que afecta predominantemente la cintura escapular y pélvica. Aunque el riesgo de hipertermia maligna no parece estar aumentado en estos pacientes en comparación con la población general, la exposición a anestésicos inhalatorios y succinilcolina probablemente deba evitarse ya que existe una predisposición a la hiperpotasemia y rhabdomiolisis.

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Utilizamos anestesia intravenosa total con propofol y remifentanilo, además de bloqueantes musculares durante el procedimiento quirúrgico, para reducir el consumo de oxígeno y minimizar las dosis de propofol y remifentanilo. La combinación de una estrategia anestésica cuidadosa, monitorización de bloqueo neuromuscular y profundidad anestésica se describen a continuación.
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Introduction

Limb girdle muscular dystrophy (LGMD) belongs to a genetically heterogeneous group of muscular dystrophies involving shoulder and hip girdles. They include a large group of diseases that are generally clinically indistinguishable. The diseases are characterized by proximal muscle dystrophy, a wide range of age onset, and sometimes include respiratory insufficiency, cardiac conduction abnormalities and cardiomyopathy. They are autosomal dominant or recessive forms, but genetic testing is limited to a research setting. We present the case of a patient with LGMD who required ascending aortic aneurysm repair surgery. The anesthetic management and special features are reported.

Case report

A 61-year-old male diagnosed with LGMD was admitted for surgical replacement of ascending aorta due to an aneurysm. His previous symptoms consisted of dysarthria, dysphagia, and hemiatrophy of the right side of the tongue due to paralysis of the right hypoglossal nerve caused by a spontaneous idiopathic dissection of right carotid internal artery. He was receiving enalaprilate and clopidogrel treatment. The patient suffered from limb weakness for four years, especially of the lower limbs, fatigue, and shortness of breath. Preoperative blood tests showed a mild persistent elevation of CK, between 250 and 400 U/L. Neurological examination revealed bilateral calf hypertrophy, mild weakness in cervical muscles, and limitation of dorsal bilateral flexion of ankle articulations. The patient did not have relatives affected by muscle disorders.

The preoperative testing showed a preserved ventricular systolic function, moderate tricuspid and mitral regurgitation, and moderate aortic stenosis and regurgitation, and an ascending aortic aneurysm of 54 mm diameter.

The patient gave informed consent for open surgery and anesthetics; he also gave his consent to publish this report. He was premedicated with 300 mg of iv ranitidine. Intraoperative monitoring consisted of invasive arterial pressure, central venous pressure, 5 lead electrocardiogram, pulse oximetry, end tidal capnography, bispectral index (BIS), central temperature, hourly diuresis, and neuromuscular blockade monitoring (NMB) by means of accelerometry (TOF-WATCH monitor, Organon Ireland Limited, Dublin, Ireland) in the left posterior tibial nerve. We used a TCI (Target Controlled Infusion) device with a Schindler model for the propofol infusion, and a Minto model for remifentanil infusion. Induction of anesthesia consisted of a target dose 3-5 μg/ml of propofol and 2-3 ng/ml of remifentanil, and 1.2 mg/Kg bolus of rocuronium, due to a suspected increased risk of aspiration and airway collapse. Anesthesia was maintained with infusions rates of 3-5 μg/ml for propofol, and 1-3 ng/ml for remifentanil, to keep BIS between 40 and 60, and rocuronium boluses to keep TOF count below 2; during the cardiopulmonary bypass, infusion rates were decreased to 2 - 2.5 μg/ml of propofol, and 0.5 - 1.2 ng/ml for remifentanil.

Surgery performed was replacement of the ascending aorta from the sinotubular ridge to the origin of the innominate artery with a dacron tube graft. By-pass time was 80 min, and the whole procedure lasted for 330 min.

Regarding the NMB management, 2 responses in the TOF-count were observed 135 minutes after the first dose, and two additional boluses (at 75 and 145 min) were needed. At the end of the surgical procedure, with a TOFr of 39%, we decided to reverse the blockade with 180 mg of sugammadex (2 mg/kg). The temperature of the patient was 35.8 °C at that time. Thirty minutes before the end of the procedure 300 μg of fentanyl and 1 g of paracetamol were administered for postoperative analgesia, and the patient was transferred to the Surgical Intensive Care Unit. Although NMB was reversed at the end of the procedure (TOFr of 92%), mechanical ventilation was maintained for 5 h after admittance, as per protocol. The patient was discharged to a medical ward 24 h afterwards.

Discussion

A decision was made not to perform genetic testing because of refusal of the patient to have children in the future, so genetic counseling seemed unnecessary.

Both the neurologist and the neurophysiologist suspected an LGMD because of the clinical findings, even in the absence of genetic testing, and the unspecific findings of the neurophysiological explorations.

As regards the anesthetic plan in a myopathy patient, the possibility of respiratory and cardiovascular disease should be kept in mind, as well as an anesthetic strategy should be developed in order to minimize the risk of triggering a muscular or metabolic complication. Patients with LGMD, are considered to have an increased sensitivity to some non-depolarizing relaxants. Even when respiratory muscle weakness with symptomatic hypventilation and respiratory failure is found in a few of the LGMD cases, respiratory function is a concern in the postoperative
setting, therefore anesthetic agents with rapid onset and metabolism should be chosen in order to prevent prolonged postoperative respiratory depression and excessive sedation.

Although the risk of malignant hyperthermia does not seem to be increased in these patients compared with the general population, the exposure to inhaled anesthetics and succinylcholine should probably be avoided because they have a predisposition to hyperkalemia and rhabdomyolysis. We chose to use muscle relaxants during the surgical procedure, in an attempt to reduce oxygen consumption, and reduce the doses of propofol and remifentanil.

TCI are based on population-based PK/PD models, and ignores inter-individual variability, limiting the accuracy of the estimated drug concentrations, and caution is needed whenever using the model in patients with cardiac disease, obesity, diabetes, nephropathy, alcoholism, and the elderly.

The pharmacodynamic behavior of the flexor hallucis brevis muscle (FHB), stimulated by posterior tibial nerve (PTN), is less thoroughly studied than the adductor pollicis brevis muscle (AP), stimulated by the ulnar nerve (UN), most studies showing similar peak effects, but longer onset times and shorter recovery from NMB. However, there is no clinically significant difference between the AP and UN in relatively intense or superficial NMB. We believed that PTN stimulation, and FHB monitoring, may be more appropriate in this case than AP UN stimulation, because the physical restraint imposed by the position of the patient and the surgical team would have prevented us from visually evaluating the eventual artifacts and clinical value of the numerical figure obtained from the TOF WATCH monitor, whereas the ankle may be more easily accessible, and lacks of the aforementioned imposed physical restraint.

In all post-junctional neuromuscular diseases, non-depolarizing drugs require careful dosing and meticulous monitoring. Since the development of sugammadex, a very effective agent to reverse the action of rocuronium, we believe that rocuronium might become the non-depolarizing neuromuscular blocking agent of choice in any kind of weak patients exposed to major surgery requiring muscle relaxation, even in cases when rapid sequence intubation is required. We believed that dysphagia and dysfunction of the tongue muscles suggested a change in the airway protective reflexes, and probably an increased risk of airway collapse with the induction of anesthesia, probably making mask ventilation difficult. Although there is scarce information on the use of sugammadex in neuromuscular disorders, we have found several reports that found it safe and effective.

The use of neuromuscular monitoring may have been useful in this case to rule out the need of reversal agents and to avoid excessive doses of muscle relaxants, and the use of BIS monitoring and short acting drugs probably helped to decrease the risk of intraoperative awareness, and to titrate the rates of propofol and remifentanil infusions. Although neuromuscular function was likely to spontaneously recover in a patient who otherwise was going to need mechanical ventilation for hours, we believe that complete reversal of the NMB in this patient was important because of two main reasons; the neuromuscular disease may predispose to an unpredictable response and duration of NMB, and, on the other hand, we should not solely rely on the data obtained from NMB monitoring, since a meta-analysis by Naguib et al., failed to demonstrate a reduction in post-operative recurarization despite the use of NMB monitoring in the intraoperative period. Recovery from NMB is not the only issue when deciding to withdraw mechanical ventilation after cardiac surgery; patients with neurological complications, hemodynamic instability, and surgical bleeding require a more conservative management in order to avoid the need of reinstallation of mechanical ventilation.

The combination of a carefully planned anesthetic strategy, anesthetic depth and neuromuscular blockade monitoring, short acting intravenous agents, and reversing agents helped in this case to perform an eventless and successful surgical and anesthetic procedure.

Conflict of interest

The authors declare no conflicts of interest.

References