Journal of Anesthesiology
2013; 1(3): 21-23
Published online November 10, 2013 (http://www.sciencepublishinggroup.com/j/ja)
doi: 10.11648/j.ja.20130103.11

Anesthetic management of a child with family history of malignant hyperthermia: Case report

Tiago Freire da Fonte, Lucas Gianni Menegaz, Marcos Lopes de Miranda, Rodrigo Montebello de Araújo, Carlos Darcy Alves Bersot

Department of Anesthesia, Hospital Federal da Lagoa, Rio de Janeiro, Brazil

Email address:
cet.lagoa@gmail.com(C. D. A. Bersot)

To cite this article:
Tiago Freire da Fonte, Lucas Gianni Menegaz, Marcos Lopes de Miranda, Rodrigo Montebello de Araújo, Carlos Darcy Alves Bersot. Anesthetic Management of a Child with Family History of Malignant Hyperthermia: Case Report. *Journal of Anesthesiology.* Vol. 1, No. 3, 2013, pp. 21-23. doi: 10.11648/j.ja.20130103.11

Abstract: Malignant hyperthermia (MH) is a rare and autosomal dominant myopathy triggered by volatile anesthetics and depolarizing neuromuscular blocking agents such as succinylcholine and is characterized by an acute hypermetabolic clinical state. This report presents the importance of preoperative preparation and perioperative management of patients with a family history of MH, in which there is no possibility of a diagnostic confirmation. Attention must be directed to the preparation of the anesthetic machine because modern workstations need longer cleansing times than their predecessors.Case Report: A two-year-old male child, weighing 16 kg, with a family history of MH, confirmed by muscular biopsy, underwent an elective umbilical and unilateral inguinal hernioplasty, and postectomy under intravenous general anesthesia associated with a caudal block. The preanesthetic care involves the preparation of the surgical environment and assessment of possible perioperative events. The patient's exhaled CO_2 fraction and body temperature were continuously monitored throughout the surgery and immediate postoperative period. The patient recovered without further events and was discharged from the hospital after two days.In a patient with family history of MH the administration of intravenous general anesthesia with the adequate preparation of the surgical environment allowed safe anesthetic management for the proposed surgical procedure.

Keywords: Malignant Hyperthermia, Anesthetic Management, Anesthetic Machine

1. Introduction

Malignant hyperthermia (MH) is a rare myopathy with autosomal dominant genetic inheritance characterised by an acute hypermetabolic state of the muscular tissue following the use of volatile anesthetics or depolarising neuromuscular blockers. The largest prevalence is seen in pediatric patients, where it occurs at a rate of 1:15,000, whereas in adults its prevalence is 1:40,000[1]. The clinical signs include hypercarbia caused by increased CO_2 production, masseter muscle rigidity, tachycardia, and tachypnea.[2] This case report presents the case of a child with a confirmed family history of MH scheduled for an elective surgery. Due to the presence of restrictions on age and weight it was not possible to perform a MH diagnostic muscular biopsy in the child. Assuming a high possibility of a positive diagnosis the pediatric surgery was successfully performed using a combined technique of intravenous general anesthesia and caudal epidural block.

2. Case Report

A two-year-old male child patient, weighing 16 kg, with a confirmed by muscular biopsy family history of MH, was admitted for elective umbilical and right inguinal hernioplasty and postectomy. The preanesthetic care involved the preparation of the anesthesia machine (Datex-Ohmeda AS/3 Anesthesia Delivery Unit, GE Healthcare, United Kingdom) by replacement of the CO_2 canister and airway circuits, removal of the volatile anesthetic vaporisers, and cleanup of the gas system with a steady flow of oxygen at 10 l/min for 30 min. There was immediate availability of sodium dantrolene and iced solutions. The preparation also included the reservation of a bed in the pediatric intensive care unit and the presence of highly trained nursing staff.

Following oral premedication with midazolam 8 mg, achiving mild sedation, and 30 minutes prior to venous

catheter insertion, the patient's was monitored by continuous electrocardiographic monitoring (DII, V5), pulse oximetry, non-invasive blood pressure measurement and chest auscultation. Anesthetic induction was performed with lidocaine 20 mg, fentanyl 80 µg, propofol PFS (target-controlled infusion with a target of 4µg.ml-1), and cisatracurium 2 mg. After the recommended time for optimal drug action orotracheal intubation was carried out and mechanical ventilation with FiO2 of 50% in air was started. Continuous sidestreamcapnography was performed while the face mask or the tracheal tube was in place. Shortly thereafter the orotracheal intubation an oropharyngeal thermometer was placed in position. Epidural block was performed with a 25 mm × 7 mm hypodermic needle positioned on the sacral hiatus and confirmed by positive Dogliotti. Six millilitres of 0.5% ropivacaine and 20 µg of clonidine were administered. Anesthesia was maintained by continuous infusion of propofol PFS. The patient remained clinically stable throughout the surgery, with a heart rate around 110 beats per minute, a pulse oximetry of 98%, a systolic blood pressure between 70 and 80 mmHg, an end-tidal carbon dioxide pressure of 34 mmHg, and a body temperature of 36.8 °C. Surgery was performed without complications and lasted for 150 minutes. Ten minutes after discontinuation of the propofol PFS infusion, the patient was spontaneously ventilating, and reversion of the neuromuscular block was performed with atropine 0.4 mg and neostigmine 0.8 mg. The patient was then taken to the post-anaesthetic recovery room where he stayed closely monitored for four hours. After that, he was sent to the pediatric ward where he remained without further events being discharged from the hospital after 48 hours.

3. Discussion

Since the description of the first case of MH by Denborough and Lovell in 1960, its prevention, diagnosis, and treatment have improved.[3] Knowledge of the patient's and/or the patient's relatives' clinical history helps the anaesthesiologist to perform a more rigorous assessment.

MH is a hypermetabolic disorder of the skeletal muscle and has different presentations depending on the species, race, and triggering agents. An important physiopathological process in this condition is intracellular hyperkalaemia, which may lead to cellular death.[4]

MH may manifest itself in several ways. In its classical form, the initial signs are hypercarbia, tachypnea, and tachycardia following hypermetabolism. Other symptoms may include masseter muscle rigidity, generalised muscle rigidity, respiratory acidosis, metabolic acidosis, myoglobinuria (rhabdomyolysis), arrhythmia, cyanosis, poor skin perfusion, diaphoresis, elevation of body temperature, hemodynamic instability, and bleeding (alterations in coagulation).[5] In most cases, the clinical presentation starts in the operating room, with a 90.5% prevalence of tachycardia, 82.8% prevalence of hyperventilation, 79.2% prevalence of skeletal muscle rigidity, 77.6% prevalence of alteration in blood pressure, and 69.2% prevalence of cyanosis.[6] All patients showing suspected episodes of MH, even if atypical, are candidates for muscular biopsy for a definitive diagnosis of MH. Since it is a hereditary disease, diagnostic confirmation is extremely important not only to the patient but also to the patient's relatives. On the basis of one confirmed case, pre-symptomatic diagnoses and genetic studies may be performed in selected families.

If susceptibility to MH is known, the use of alternative anaesthetics and non-polarising neuromuscular blockers may prevent the onset of an MH crisis. Confirmation of MH diagnosis is possible by using the halothane-caffeine contracture test on a skeletal muscle fragment.

The halothane-caffeine test is performed using a biopsied fragment (1–3 g) of the vastus lateralis muscle, on the outer part of the thigh. The minimum size of the fragment necessary for the test (1-3 g) makes it impossible to perform this test on patients weighing less than 25 kg.[7]

In MH-susceptible patients or in those with a family history of MH, in whom halothane-caffeine diagnostic test cannot be performed, measures should be taken to avoid clinical presentation of MH. The necessary prophylactic measures and therapies must be explained to the patient and their relatives by the anaesthesiologist. Adequate preparation of the surgical environment and anaesthetic equipment, as per the manufacturer`s recommendations, is indispensable for the safety of these patients. Currently, the simplest and most effective method for the removal of halogenated residues is the maintenance of high flows of fresh gases.8The goal is to decrease the residual anesthetic vapor concentration within the breathing circuit. These precautions represent the standard of care for the management of MH-susceptible patients.

An early report studying how gas solubility and fresh gas flow rates influence clearance of anesthetic agent in an older anesthesia machine was conducted by Tarq *et al.*2 They examined the effect of anesthetic gas solubility on residual anesthetic concentration by measuring the solubility of halothane, desflurane, isoflurane, and sevoflurane in various plastic and rubber machine components obtained from a conventional anesthesia circuit. From these data, plastic/gas and rubber/gas partition coefficients were determined,and the following order, from most soluble gas to least, was obtained: halothane>isoflurane>sevoflurane>desflurane. The washout times of these gases from an older generation Ohmeda anesthesia machine (Ohmeda, Madison, WI) with a conventional breathing circuit were then measured. At a 5 l/min flow rate for 20 min, the concentration of desflurane was reduced by 99.9%; sevoflurane, 99.7%; halothane, 99%. The significance of the fresh gas flow rate on the washout kinetics of volatile anesthetics was noted when desflurane, a relatively insoluble gas, required greater than 1 h to reach a reduction of 99% when the flow rate was decreased to 1 l/min.

One study looking at newer generation anesthesia machines was conducted by Schonell *et al.* examining five Datex-Ohmeda anesthesia workstations (AS/3 Anesthesia Delivery Unit, Bromma, Sweden). As was the case with past studies, it was found that the anesthesia machine could be purged of gases quickly in 10 min using an oxygen flow rate of 10 l/min resulting in a gas concentration of 2 ppm of isoflurane at the common gas outlet. However, inclusion of the patient breathing circuit and ventilator required 30 min of ventilating an artificial lung (1-l breathing bag) at 10 l/min to achieve concentrations less than 5 ppm. The tidal volume of 1 l was chosen to ensure adequate gas volume to flush the bellows, tubing, and patient circuit. The effects of replacing the soda lime, patient circle circuit, 1-l breathing bag and hose, ventilator hose, and the ventilator bellows were studied as well. Their findings suggested that changing only the breathing hoses, breathing bag, and soda lime cartridge was necessary. But recently it has been demonstrated that newer anesthesia work stations are more complex and can hold large amounts of inhaled anesthetics owing to the higher amounts of gas-absorbing materials used (plastic, rubber, and silicone components), which require more time for purging anesthetic gases.

For the Datex–Ohmeda AS/3 Anaesthesia Delivery Unit used, an oxygen flow of $10l \cdot min^{-1}$ for 30 min is recommended in order to remove halogenated residues from the gas system reducing the volatile agent concentration to less than 10 ppm following the removal of the vaporiser. The ventilator should be included in this purge, at a tidal volume of 1 litre. Replacement of the airway circuits, respiratory pouch, and CO_2 reabsorbent canister is also necessary.[8-9]

Despite the development of the anesthesia equipment, new guidelines on the preparation of a safe anesthesia for MH-susceptible patients are not available. New protocols are necessary for the safe handling of the new equipment in these patients.

This case suggests that in selected surgical procedures, the combination of intravenous and regional anesthesia associated with an adequate preparation of the surgical environment is particularly safe for the performance of anesthesia and surgery on patients with a family history of MH.

References

[1] Hopkins PM – Malignant hyperthermia: advances in clinical management and diagnosis. Br J Anaesth, 2000; 85:118.

[2] Larach MG, Gronert GA, Allen GC, Brandom BW, Lehman EB – Clinical Presentation, Treatment, and Complications of Malignant Hyperthermia in North America from 1987 to 2006. Anesth Analg, 2010; 110:498-507.

[3] Denborough MA, Lovell RRH – Anaesthetic deaths in a family. Lancet, 1960; 2:45.

[4] Allen GC, Brubaker CL – Human malignant hyperthermia associated with desflurane anesthesia. Anesth Analg, 1998; 86:1328-1331.

[5] Ranklev-Twetman E – Malignant hyperthermia: the clinical syndrome. Acta Anaesthesiol Belg, 1990; 41(2):79-82.

[6] Britt B, Orkin FK, Cooperman LH – Complications in Anesthesiology. Philadelphia: JB Lippingcott, 1993; 291-313.

[7] European Malignant Hyperpyrexia Group: A protocol for the investigation of malignant hyperpyrexia. Br J Anaesth, 1984; 56:1267

[8] Kim TW, Nemergut ME – Preparation of Modern Anesthesia Workstations for Malignant Hyperthermia-susceptible Patients. Anesthesiology, 2011; 114:205-212.

[9] Schonell LH, Sims C, Bulsara M – Preparing a new generation anaesthetic machine for patients susceptible to malignant hyperthermia. Anaesth Intensive Care, 2003; 31:58-62.