Pediatric Anesthesia and Critical Care Journal 2013; 1(2):43-45
doi:10.14587/paccj.2013.10

PACCJ

Considerations about anesthesia in patients suffering from myopathy

G. de Francisci[1], M. La Sala[1], G. Addabbo[1] , P. De Santis[1], D. Galante[2] , M. Caruselli[2]

[1]Department of Anesthesia adn Intensive Care, Catholic University of Sacred Heart, Policlinic "A. Gemelli", Rome, Italy
[2]University Department of Anesthesia and Intensive Care, University Hospital Ospedali Riuniti, Foggia, Italy
[3]Department of Anesthesia and Intensive Care Salesi Children's Hospital, Ancona, Italy

Corresponding author: G. De Francisci, [1]Department of Anesthesia adn Intensive Care, Catholic University of Sacred Heart, Policlinic "A. Gemelli", Rome, Italy. Email: giovannigdf50@gmail.com

Key points

General anesthesia in myopathic patients poses several problems. The patient may be a child with initial symptoms, which must be subjected to muscle biopsy or a patient with overt myopathy, with all the events muscular, respiratory and cardiac related to it, to be submitted to any operation under general anesthesia.

Abstract

Dystrophies are generally classified into progressive, such as Duchenne, Becker, the Emery-Dreifuss, the Central Core, the Nemaline, the King Denborough and congenital and mitochondrial myopathies, which can be present at birth. General anesthesia in myopathic poses several problems. The patient may be a child with initial symptoms, which must be subjected to muscle biopsy or a patient with overt myopathy, with all the events muscular, respiratory and cardiac related to it, to be submitted to any operation under general anesthesia even if regional blocks are also possible. For this reason, it becomes crucial an initial framework preoperative of the myopathic patient.

Keywords: myopathy; general anesthesia.

Anesthetic assessment

Based on our clinical experience consider necessary the following assessment:

- in all myopathic patients should be carried out a cardiologic examination; in many myopathies, in fact, it can be observed a preclinical cardiopathy or an overt dilated cardiomyopathy, conduction disorders, shortening of the PQ, QT prolongation, especially in the advanced stages (1, 2).

- A respiratory evaluation should be performed because many myopathies in advanced stages are accompanied by a respiratory failure so it will be necessary to make an assessment of respiratory status and the patient's ability to mobilize secretions.

In relation to the risk of malignant hyperthermia with volatile anesthetics, which still represents a topic of discussion without unanimous agreement, we can say that in our opinion the risk is concrete with only two types of myopathy: the Central Core and the King-Denborough. In the other myopathies, in particular in the Duchenne, the volatile anesthetics can cause rhabdomyolysis-hyperkalaemia only for prolonged exposure. Short-term exposure, such as the inhalational induction, seems to be not a problem, even in our experience.

Intraoperative management

We believe, in the intraoperative management of the myopathic patient, it must be kept in mind a few simple concepts:

• the inhalational induction in children is not contraindicated

• the dose of neuromuscular blocking agents has to be reduced (3).

• in mitochondrial myopathies, prolonged exposure to propofol can cause PRIS ("Propofol Infusion Syndrome").

• succinylcholine is always controindicated, because of the risk of rhabdomyolysis-hyperkalemia, which can also be fatal (4).

• the prostigmine is not contraindicated, even in myotonia (5).

In our recent study we evaluated a total number of 14 patients, including 10 with overt Duchenne dystrophy, one affected by isolated hyper-CK-emia and 2 without defined diagnosis as described in Table 1.

Conclusions

For all patients, our anesthetic management has provided a monitoring system comprising: electrocardiogram, pulse oximetry, non-invasive measurement of blood pressure, capnometry and the use of thermal blankets to prevent hypothermia. Preoperatively we never gave premedication. In the intraoperative we always used an induction to Sevoflorane with high MAC (7-8%) in combination with Alfentanyl 12.5-25 µg / kg; as we have always associated a low-dose non-depolarizing muscle relaxant: the Rocuronium Bromide at dose of 0.6 mg / kg. For maintaining of anesthesia we never used halogenated anesthetic vapors but exclusively Propofol in intravenous infusion or in refracted bolus, depending on the duration of the operation, associated with an opioid (Alfentanyl, Fentanest).

At the end of anesthesia we used the reversal of muscle paralysis with neostigmine and patients were all awakened in the operating room.

With this approach, no case have shown postoperative complications, neither immediate nor after hours; vital signs were stable and the body temperature normal.

We finally point out that, based on these results, we believe "excessive" a number of precautionary measures taken in the past such as: the operation as first on the list, the caffeine-halothane test and the wash-out of the anesthesia machine with oxygen.

Table 1. Examined patients suffering from myopathies

PATIENT ID	AGE	DIAGNOSIS
ZF	6	DUCHENNE
UN	6	DUCHENNE
MA	10	DUCHENNE
CE	8	DUCHENNE
DA	2	DUCHENNE
SM	8	OTHER MYOPATHY NMD
PG	2	IPER-CK-EMIA
LD	4	UNDEFINED DIAGNOSIS
CE	9	DUCHENNE
PF	8	DUCHENNE
BA	7	DUCHENNE
MD	6	DUCHENNE
AO	9	UNDEFINED DIAGNOSIS
GM	8	DUCHENNE

References

1. Crean P, Hicks E. Essentials of neurology and neuromuscular disease. In Cotè, Leiman, Todres: a practice of anesthesia for infants and children, 2009
2. Jimenez N, Song K, Lynn A. Haemodinamic instability during prone spine surgery in a patient with muscular distrophy. Pediatric Anaesth. 2013; 23: 294-296
3. Muenster T et al. Rocuronium 0.3 mg/Kg induces a normal peak effect but an altered time course of neuromuscular block in patients with Duchenne's muscular dystrophy. Pediatric Anesthesia 2006; 16:840-845
4. Lavezzi W, Capocchione S, Muldoon S, et al.: Death in the emergency department: un unrecognized awake malignant hyperthermia-like reaction in a six year old. Anest.Analg. 2013;116:420-423
5. Veyckemans F. Myotonic dystrophies 1 and 2: anestetic care. Pediatric anaesthesia 2013 (in press)