Prolonged Neuromuscular Blockade following a bolus of Succinylcholine and Rett syndrome: a Case Report

Spencer, L.; Perry da Câmara, L., Salta, C.; Poeira, R.; Casteleira, M.

INTRODUCTION

Rett syndrome is a rare, genetic, neurodevelopment disorder that affects mainly women¹. It is caused by mutations in the gene that encodes methyl CpG binding protein 2 (MeCP2). This gene is located on the long arm (q) of the X chromosome, regulating the expression of several genes involved in neuronal physiology.²

This syndrome is characterized by the progressive loss of cognitive, motor and communication capacities; seizures and EEG changes. It may also be associated with electrolyte imbalance, cardiopulmonary and skeletal muscle abnormalities and airway deformities. These features lead this disease into a challenging anesthetic management.³

CASE REPORT

- 38 year old woman
- Surgery: dental extractions (tooth decays)
- Rett Syndrome (diagnosed in her childhood)
- No past history of anesthesia or surgery

PREOPERATIVE EVALUATION

- Rett Syndrome – stage 4: severe cognitive impairment, craniofacial dysmorphism, dorso lumbar scoliosis, hypotonia and limb spasticity
- Airway: limited evaluation due to patient condition
- Pulmonary Auscultation: normal
- Complementary Exams: normal
- Preoperative medication: midazolam *per os* (0,15mg/kg)

INTRA AND POSTOPERATIVE

- Induction: Rapid Sequence Induction: propofol (2mg/kg), fentanyl (1mcg/kg) e succinylcholine (1mg/kg)
- Maintenance: sevoflurane
- Standard Monitoring (ASA)+ NMB monitoring by acceleromyography
- No hemodynamic instability during the surgery
- TOF=0 one hour after the induction
- Complete reversal of the NMB (TOF>95%) 90 minutes after succinylcholine administration
- Postoperative recovery without complications
- Further investigation was proposed by the Genetics Department refused by the patient's family

CONCLUSION

In patients with Rett Syndrome succinylcholine should be carefully used due to the risk of arrhythmia. This case suggest the risk that the prolonged NMB is also present, may be due to an enzymatic deficit caused by a genetic mutation. Further investigation is necessary to test this hypothesis.

Image 1 – MECP 2 functions

DISCUSSION

- Rett Syndrome is a rare disease, that is why the optimal anesthetic management is not well defined. The choice of balanced general anesthesia for teeth extraction was due to the patient cognitive impairment.⁴
- Rett is associated with hyperkalemia with the underlying risk of arrhythmia. The use of succinylcholine in Rett patients, though controversial, is not contraindicated.
- In this case, in the absence of hyperkalemia in the preoperative laboratory values, succinylcholine was the neuromuscular drug chosen, provoking a prolonged neuromuscular block.
- Literature suggests the hypothesis of MeCP2 regulating, via microRNA (hsa-miR-212 e 132), the expression of BCHE and ACHE, that encode butyrylcholinesterase and acetylcholinesterase. A mutated MeCP2 would, therefore decrease butyrylcholinesterase and acetylcholinesterase transcription.⁵
- Non genetic factors causing prolonged NMB were excluded.

EEG – Electroencephalography, ASA – American Society of Anesthesiology, NMB – neuromuscular block, TOF – *train of four*

References:

- ¹Chahrour, M. et al; Science. May 2008; 320(5880): 1224–1229; ²Pierson, J, Mayhew, J.; AANA Journal/October 2001/Vol. 69, No. 5; ³Kako, H., Int J Clin Exp Med 2013;6(5):393-403; ⁴Guideline on management of dental patients with special health care needs. Pediatr Dent. 2008;30(7 Suppl):107-111; ⁵Acetylcholinesterase: old questions and new developments; Tsim, K., Soreq, H., Frontiers in molecular neuroscience. March 2013; ⁶Ananthanarayan C, Sigal M, Godlewski W. General anesthesia for the provision of dental treatment to adults with developmental disability. Anesth Prog. Winter 1998;45(1):12-17; ⁷Miller RD et al; Basics of Anesthesia, sixth edition, 2005, Elsevier Saunders; ⁸Morgan and Mikhail, fifth edition, 2013, McGraw hill