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Case Report Open Access

Target Control Infusion (TCI) Anesthesia in Morquio Syndrome During Cranio Spinal Surgery

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Abstract

Mucopolysaccharidosis (MPS) are a group of metabolic disease caused by lysosome enzyme anomalies linked to glycosaminoglycan metabolism. MPS are classified in 7 different types and multiple sub-types, depending on gene involved. In this particular case, our patient was affected by Morquio syndrome, also known as mucopolysaccharidosis IV A. This specific type of mucopolysaccharidosis is characterized by N-Acetilgalactosamine-6-solphate deficiency and it is primarily associated with multiple skeletal disorders. Patients with MPS have multiple comorbidities such as joint laxity and atlanto-axial instability that require surgical interventions through their life. In particular, Morquio syndrome represent an anaesthetic challenge, due to respiratory problems such as restrictive pulmonary defect due to thoracic cage alteration. In these syndromes, it is frequent to observe upper airway obstruction during head flexion and compression of the cervical spinal cord due to hypoplasia of axis dens causing difficult intubation. Usually show difficult airway management because glycosaminoglycan can accumulate in the upper airway resulting in hypertrophy of adenoids, tonsils, tongue, and larynx. These patients usually undergo multiple surgeries, including spinal surgery in these particular surgery is very important to avoid muscle relaxant drugs and inhalational agents because they affect evoked potential. This case report show how Morquio syndrome can be managed during general anesthesia and in particular, the feasibility of TCI anesthesia.

TCI anesthesia is becoming widely popular for its safety and broad range of indication, in particular during spinal surgery this technique minimize any interference with evoked potential. Mucopolysaccharidosis may be quite challenging considering pharmacokinetics because patients are in a smaller size than their real age, this may affect drugs distribution volume and its metabolism. Our case show how to manage these pathology safely and in particular, we focused on using TCI anesthesia during spinal surgery. In this particular case we chose to use Kataria model for Propofol and Minto model for Remifentanil, patient remained stable throughout surgery. She did not show delay in emergence after general anesthesia and the post-operative period was uneventful. We can therefore suggest that TCI anesthesia can be used safely in patient affected by Morquio syndrome during spinal surgery.

Keywords: Morquio syndrome; Spinal surgery

Case Report

Morquio syndrome is a rare autosomal recessive disorder caused by N-Acetilgalactosamine-6-solphate deficiency, it that is estimated to occur in one of every 200,000 births. This syndrome is part of a wider spectrum of metabolic disease called muchopolysaccharidoses, a congenital metabolic disease caused by deficit of lysosomal enzymes. MPS are classified in 7 types and many sub-types depending on enzyme deficiency. In this particular type, diagnosis usually does not occur at birth because; symptoms may appear between 1 and 3 years old. This disorder, also known as mucopolysaccharidosis IV A, causes multiple deformities to soft tissues and skeletal system. Furthermore, Morquio syndrome may be classified in two sub-categories: A type N-acetilgalactosiamine 6 sulphate deficiency and B type beta-galactosidase deficiency.

Regardless type A or B, children affected, usually have common signs and symptoms such as: spine deformities, abnormal heart and skeleton development, dwarfism, pectus carinatum, odontoid hypoplasia, atlanto-axial instability, joint laxity. Mucopolysaccharidosis may present multiple comorbidities that may affect anesthetic management. Patients usually have difficult airways due to glycosaminoglycan deposition in upper airways this cause lymphoid hypertrophy and macroglossy, atlanto-axial instability is also present, this precludes sniffing position during ventilation or intubation. Other relevant features that may affect anesthesia are: possible aortic insufficiency and rib cage alteration.

We present a case of total intravenous anesthesia in a 15 year old girl with Morquio syndrome during spinal surgery for atlanto-axial instability. She was born from non consanguineous parents with physiologic delivery at 37 gestational weeks, pregnancy was reported as uneventful. She was diagnosed at 3 year old for facial dimorphisms and multiple episodes of respiratory infections, other physical features suggesting mucopolysaccharidosis were presented from birth such as pectus carinatum and kyphoscoliosis. A definitive diagnosis was made thanks to increased level of heparin sulfate, and β-galactosidase level. She also had PLU10X and PC507L gene modification.

The patient was 15 year old, her length was 110 cm and weight 25 kg and she was affected by dwarfism a typical feature of this MPS type. Other relevant features to highlight were: large head, saddle nose, short neck, macroglossy. She showed also slight intellectual disability an hyperactive trait. Due to spine anomalies and joint anomalies, she also showed duck wadding gait. Before surgery, she underwent MRI,

showing epistropheus hypoplasia and cranio-vertebral stenosis requiring surgery and she also performed pulmonary spirometry showed mild restrictive deficit. EKG was normal except sinusal tachycardia, while echocardiography showed mild tricuspid and mitralic insufficiency.

Morquio Syndrome requires a careful anesthesia management, if possible, regional anesthesia when indicated will be the first choice; general anesthesia may be a risk for these patients due to lung, thorax and cardiovascular anomalies such as heart defects or rib cage alteration as mentioned before [1].

Considering the possibility of difficult intubation and ventilation, we prepared different devices in order to manage difficult airways such as: laryngeal masks, video laryngoscope, and emergency coniotomy kit [2]. Perioperative monitoring included electrocardiogram, noninvasive blood pressure, oxygen saturation (SpO_2) and end-tidal carbon dioxide. We chose to perform arterial cannulation after general anesthesia induction, to avoid further discomfort.

We induced general anesthesia using Propofol and Remifentanil in TCI mode, we chose Kataria model for Propofol considering age and weight of our patient. Whereas for Remifentanil infusion, we chose Minto model. In order to avoid complication during endotracheal intubation, we decide not to administer muscle relaxant drugs [3]. We obtained optimal intubation condition when Propofol plasmatic concentration was 4.5 μg/mL We chose not to use BIS monitoring because prone positioning was used and it is sometimes associated to pressure ulcers during lengthy surgery [4,5].

Laryngoscopy was performed with size 3 Macintosh blade by an experienced anaesthetist using a neutral head positioning, sniffing position or head hyperextension was not performed considering spine instability. Epiglottis showed imbibition caused by mucopolisaccarid storage, patient was easily intubated using wire reinforced cuffed endotracheal tube no. 6. During laryngoscopy, anaesthetist achieved Cormack 1 without particular issue.

To perform surgery, carefully patient was pronated, anaesthetist and neurosurgeon stabilized the head during rotation and this delicate moment was easier thanks to TCI anesthesia.

MEP (Motor Evoked Potential) were easily assessed after induction in order to have a baseline before starting surgery and throughout surgical time in order to monitor or highlight any relevant modification.

Anesthesia was maintained with Propofol 3.3 μg/mL, Remifentanil concentration varied with surgical stimuli. Patient showed great haemodynamic stability throughout surgery, without any episode of hypotension or bradycardia considering the delicate surgical site. Emergence from anaesthesia was slow and gentle; she opened her eyes when Propofol reached 1.5 μg/mL concentrations at plasmatic site. Extubation was performed when patient reached adequate tidal volume for its weight. She was then moved into recovery room when she reached a satisfactory respiratory and haemodynamic stability.

As intraoperative analgesia, we administer Paracetamol 15 mg/kg and a single shot of Morphine 0.1 mg/kg, we also provided post-operative nausea and vomiting prophylaxis at the end of surgery, administering 0.1 mg/kg Ondansetron. Post-operative 24 h analgesia was maintained with Paracetamol 15 mg/kg each 8 h, no rescue analgesia was required (VAS<4). Patient was discharged 5 days after surgery in well-being state, no surgical complication was reported.

General Anesthesia was the unique choice for this patient, moreover intraoperative neurophysiological monitoring required a Propofol-based anaesthesia [6], so we planned to use TCI anesthesia using Kataria Model (Propofol) and Minto Model in order to avoid interference with MEP (Motor Evoked Potential) and SSEP (Somatosensory evoked potential).

Intravenous anesthesia was a mandatory choice during general Anesthesia with MEP. Sekimoto et al. and Kalmat et al. demonstrated that evoked potential are affected by inhaled gas, in lesser extent by halothane, specifically a reduction in evoked potential amplitude and a latency increased was reported [7,8]. Propofol, has minimal or no effect on SSEP, so it is advisable to use it when possible [9,10].

Conclusion

TCI may represent a valuable choice in patient with Morquio syndrome undergoing spinal surgery, in order to have a "tailor-made" anesthesia when performing MEP during surgery. TCI also shows benefit during prone positioning because drugs are safely administered during patient rotation, lower risk of PONV and post-operative shivering.

References

1. Tobias JD (1999) Anesthetic care for the child with morquio syndrome: general versus regional anesthesia. J Clin Anesth 11: 242-246.
2. Gan TJ, Meyer TA, Apfel CC, Chung F, Davis PJ, et al. (2007) Society for Ambulatory Anesthesia guidelines for the management of postoperative nausea and vomiting. Anesth Analg 105: 1615-1628.
3. Minardi C, Astuto M, Spinello CM, Murabito P, Pellegrino S, et al. (2012) Dose-Response of Propofol for Tracheal Intubation in Children Correlated to Intubation Condition Score and Cerebral State Index. Randomized, Double-Blinded Trial. J Anesthe Clinic Res 3:202.
4. Inoda A, Nagata H, Miura H, Doe A, Suzuki K (2015) Bispectral Index Electrodes May Induce Skin Lesions. Masui 64: 318-320.
5. Yamashita S, Mizutani T, Shimizu T, Tanaka M (2011) Skin lesions associated with BIS monitoring in pediatric patients undergoing cardiac surgery. J Anesth 25: 477-478.
6. Pajewski TN, Arlet V, Phillips LH (2007) Current approach on spinal cord monitoring: the point of view of the neurologist, the anesthesiologist and the spine surgeon. Eur Spine J 16: 115-129.
7. Sekimoto K, Nishikawa K, Ishizeki J, Kubo K, Saito S, et al. (2006) The effects of volatile anesthetics on intraoperative monitoring of myogenic motor-evoked potentials to transcranial electrical stimulation and on partial neuromuscular blockade during propofol/fentanyl/nitrous oxide anesthesia in humans. J Neurosurg Anesthesiol 18: 106-111.
8. Kalkman CJ, Drummond JC, Ribberink AA (1991) Low concentrations of isoflurane abolish motor evoked responses to transcranial electrical stimulation during nitrous oxide/opioid anesthesia in humans. Anesth Analg 73: 410-415.
9. Boisseau N, Madany M, Staccini P, Armando G, Martin F, et al. (2002) Comparison of the effects of sevoflurane and propofol on cortical somatosensory evoked potentials. Br J Anaesth 88: 785-789.
10. Deletis V, Sala F (2008) Intraoperative neurophysiological monitoring of the spinal cord during spinal cord and spine surgery: A review focus on the corticospinal tracts. Clin Neurophysiol 119: 248-264.