Case Report:

Anesthesia for Mowat-Wilson Syndrome

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CASE REPORT

Past Medical History

- 15 year-old (25 kg), Mowat-Wilson Syndrome, Hirschsprung disease, epilepsy and mental retardation (ASA III)
- Severe upper gastrointestinal bleeding associated with pyloric stenosis and marked gastric distension
- Proposed for a laparoscopic gastric sleeve and gastrojejunostomy

Physical examination

- Deep-set eyes, broad nasal bridge, prominent and pointed chin, heavy eyebrows and macroglossia
- No further criteria for difficult airway (DA) - Mallampati I, mouth opening >3 cm, adequate cervical mobility
- Laboratory results and chest x-ray were normal

INTRAOPERATIVE COURSE

Airway

Rapid sequence induction

Plan A succeeded: Cormack I laryngoscopy, easy intubation

Breathing

Volume-controlled ventilation, PEEP

Blood gas analysis (pO₂ 162-222 mmHg, pCO₂ 34-45 mmHg)

Circulation

Invasive blood pressure, ultrasound-guided central venous catheter

Goal-directed fluid therapy (pulse pressure variation)

Hemodynamic stability without vasopressors, Lactate 0.5-1 mmol/L

Drugs and Maintenance

Sevoflurane (MAC and BIS guided), Oxygen/Air and Remifentanyl

Bilateral ultrasound-guided quadratus lumborum block (Ropivacaine 0.375% 10 mL each side), using a 120 mm needle

Post-operative analgesia (Paracetamol, Ketorolac, Tramadol SOS)

Exposure

Forced air warming

DISCUSSION

Mowat-Wilson syndrome is a genetic disease caused by heterozygous mutations of the ZEB2 gene. It is characterized by typical face, mental retardation, epilepsy and variable congenital malformations, including Hirschsprung disease, congenital heart disease, agenesis of the corpus callosum and eye defects. Pyloric stenosis (PS) affects 4.7% of the patients.

Management of the airway represented our biggest challenge. On one hand, craniofacial malformations could be associated with DA. On the other hand, PS posed a considerable risk of aspiration.

As it would be difficult for the patient to cooperate with an awake intubation we decided for a RSI. However, the DA car, including a fibroscope, was readily available.

Since it was first described by Blanco, QLB has been extensively studied in abdominal and laparoscopic surgery. The lateral approach may generate analgesia from T7 to L1, effectively covering all the laparoscopic ports needed for this surgery.

Compared with the TAP block, QLB has proved to result in a widespread and long-lasting effect in laparoscopic surgery.

The clear opioid-sparing effect guaranteed by regional anesthesia is not to be forgotten, considering the systemic nature of the disease.

The surgery went on without mishappenings, NB was reversed with Sugammadex 2 mg/kg and the patient safely extubated.