Small-dose intrathecal morphine and perioperative alpha-2 agonists with lidocaine infusions in a patient with Rett Syndrome undergoing scoliosis surgery. A case report

O. Stiennon¹, L. Van Der Essen¹, G. Samouri¹, R. Rossillon², N. Parisi¹.

¹Saint Pierre, Anesthesiology, Ottignies, Belgium.

²Saint Pierre, Orthopedics Surgery, Ottignies, Belgium.

Rett syndrome is a rare genetic neurological disorder due to a mutation of MECP gene on chromosome X. Mains features that concerns anaesthesiologists are the obstructive sleep apnea, epilepsy, autonomic disturbance, long QT interval and high sensitivity to opioids and sedative agents.

We report the case of a girl aged 14 (1m51, 50kg), affected by Rett syndrome, complicated by a scoliosis (neuromuscular type) progressing quickly in spite of wearing a corset since 2013. Our case reports the analgesia results observed with our optimised management without complication or respiratory event. The surgery consisted in a posterior spinal instrumentation and fusion.

After inhalatory induction (sevoflurane and nitrogen oxide), anesthesia was maintained with propofol with TIVA Schnider's model, **lidocaine 2mg/kg/h, ketamine 0.25 mg/kg/h and dexmedetomidine 1µg/kg/h during paravertebral muscles dissection, then 0.7 µg/kg/h during the fusion, and stopped when reduction is complete.** The surgeon realised himself a spinal injection of morphine (1mcg/kg). The patient stayed hemodynamically stable during the 5 hours of surgery (figure 2). She was extubated in the operating room with a non complicated weaning from mechanical ventilation.

The patient was transferred extubated to intensive care with FiO2 50% and a noradrenalin infusion to treat transient vasoplegia, it was stopped on day 1.

On day 0, a moderate prolonged sedation was observed, with a positive Naloxone test due to spinal analgesia but she had neither respiratory nor haemodynamic complication.

The postoperative pain management consisted in paracetamol (60mg/kg/d), ketorolac (0,5mg/kg/ 8hrs) and **lidocaine infusion (1,33 mg/kg/h for 48 hours)**. As no verbal communication was possible, pain was evaluated with the FLACC scale (figure 3).

No non-invasive ventilation was necessary, nasal prongs delivered oxygen until day 2. After 2 days in intensive care, the patient returned to the pediatric unit. **No PCA with opioid was needed**, with very good control of analgesia and comfort.

We used smaller dose of intrathecal morphine than advised (5-10 µg/kg) but in this syndrome precaution is advisable with this technique. **Perioperative alpha 2 agonist and lidocaine seems to be a good alternative to opioids for scoliosis surgery.** To our knowledge, no literature is available for the moment on the association of these 3 drugs. Studies on bigger cohorts are needed.

Figure 1: Lumbar spine radio before (Cobb angle 53° between T8 and L4) and after surgery

Figure 2: Pain score via FLACC scale with the respiratory parameters of the patient

	Day 1	Day 2	Day 3	Day 4
FLACC Scale	1	1	2	1
SaO2 (%)	100	97	98	95
Lpm O2	4	2	0	0
Respiratory rate	13	15	17	18

Figure 3: Intra operative parameters