Anesthesia in a child with Rett syndrome: A case report and literature review

Case summary

A 7-year-old girl presented for an open feeding gastrostomy because of progressive feeding difficulties. Her weight was 15.2 kg (below the 5th percentile). Her hemoglobin was 10 g/dL, and her electrolytes were within normal limits. The patient's medical history revealed that she was delivered at full term following an uncomplicated pregnancy. Her developmental milestones were normal until 18 months of age, when she stopped meeting developmental guidelines, and she was diagnosed with Rett syndrome at 2 years of age.

The patient was premedicated with 7.5 mg of oral midazolam. In the operating room, the electrocardiogram, noninvasive blood pressure, precordial stethoscope, and pulse oximetry monitors were placed. Anesthesia was induced with 70% nitrous oxide in 30% oxygen and 8% sevoflurane. A 20-gauge intravenous catheter was inserted. While the patient was under deep general anesthesia, direct laryngoscopy was accomplished without difficulty. The epiglottis and arytenoids were sprayed with 1 mL of 2% lidocaine, and the trachea was intubated using a 5.0 endotracheal tube. Anesthesia was maintained with 63.5% nitrous oxide, 35% oxygen, and 1.5% sevoflurane. Rocuronium was used for intraoperative muscle relaxation. At the conclusion of the 53-minute surgical procedure, the nitrous oxide and sevoflurane were discontinued, and the muscle relaxant was reversed with neostigmine and glycopyrrolate. The trachea was extubated when the patient opened her eyes on command. She was transported to the postanesthesia care unit, and her postoperative course was uneventful. The patient was discharged 3 days later.

Discussion

Rett syndrome is a progressive degeneration of the central nervous system that first appears in early childhood following a period of normal growth and development. Because of neurological problems associated with failure to thrive, these children present for neuroimaging studies as well as surgical procedures, such as gastrostomies for nutritional management and scoliosis correction. Scoliosis with a double-curved deformity is found in 80% of children with Rett syndrome and is considered a characteristic feature (Table 1). Among other features that may be of interest to anesthetists are air swallowing, screaming spells, and vasomotor hyper-reactivity of the feet (Table 2).

Although the first report describing the disease by Rett suggested elevated...
ammonia as a marker, this has not been found to be a consistent abnormality,\(^1\) and to date no test has been identified to help with the diagnosis. The diagnosis is based on progressive neurological deterioration characterized by a progressive loss of developmental milestones, with development of autism, automatism, convulsions, and mental retardation. Hand wringing, choreoathetosis, myoclonic movements, and stereotypic automatism also are characteristic of these patients.\(^1,2\) Because of decrease in nutritional intake, there is a marked loss of muscle mass.

Although during awake periods there may be episodes of irregular breathing and even periods of apnea,\(^3\) no such irregularities occur during sleep, where breathing is stable. This feature has not been adequately explained. The interaction between the respiratory system of patients with Rettsyndrome and anesthetic agents is unknown.\(^4\) Konarzewski\(^5\) reported a 3-year-old patient who experienced a prolonged recovery period following general anesthesia in which the patient was allowed to breathe spontaneously throughout the procedure. However, this particular patient had been premedicated with oral trimiprazine and ketamine followed by 10 minutes of nitrous oxide/oxygen/halothane anesthesia. One would expect that the type of premedication used would still have an effect into the postoperative period. The postoperative period also was complicated by seizures. It would seem inappropriate to state a causal relationship between Rettsyndrome and delayed recovery from anesthesia in this patient.

Most of the few cases reported in the literature,\(^4-6\) concerned patients who had undergone scoliosis surgery. Scoliosis is found in 50% of patients with Rettsyndrome, and in all of the reported cases the patients had significant anatomic and physiologic problems related to their scoliosis. Our patient exhibited the major features of Rettsyndrome, with developmental delay, mild scoliosis, hand wringing, loss of speech, and truncal ataxia. Although she did have mild scoliosis, it was not significant enough to warrant surgery. We were able to premedicate her with good results. She tolerated the anesthesia and surgical procedure well, with no intraoperative or postoperative problems and no signs of apnea or oxygen desaturation. She was admitted and observed for 3 days due to surgical concerns unrelated to the anesthetic technique.

### Summary

Rett syndrome is a devastating progressive deterioration of the central nervous system in the young child. Since the diagnosis is becoming more common,\(^1\) the anesthetist may encounter these children in clinical practice. This report demonstrates some of the features that may be of importance to the anesthetic management of these children (Table 3).

### Table 3. Anesthetic concerns of Rett syndrome

<table>
<thead>
<tr>
<th>Lack of cooperation</th>
<th>Muscle wasting</th>
<th>Abnormal continuous limb movement</th>
<th>Abnormal respiratory control</th>
<th>Difficult positioning secondary to scoliosis and chest deformity</th>
<th>Vasomotor instability</th>
<th>Metabolic abnormalities — increased lactic acid levels</th>
<th>Altered sensitivity to painful stimuli</th>
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### REFERENCES


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