



Anesthetic considerations in a child with Sotos syndrome –A case report–

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Sotos syndrome is a rare sporadic genetic disorder characterized by pathognomonic facial features, motor developmental delay induced by hypotonia, learning difficulties, and cardiac and renal anomalies. This report describes the case of a 4-year-old child with Sotos syndrome who underwent a right hydrocelectomy under general anesthesia. We report our experience with airway management, choice of anesthetic drugs, and other anesthetic implications in Sotos syndrome. (*Anesth Pain Med* 2017; 12: 240–242)

Key Words: General anesthesia, Muscle Hypotonia, Neuromuscular blockade, Sotos syndrome.

Sotos syndrome, also known as cerebral gigantism, is a rare genetic disorder characterized by excessive growth during the first years of life, characteristic facial appearance, and learning difficulties [1]. This overgrowth stabilizes between the ages of 2 and 6 years with head circumference and height above the 97th percentile. Subsequently, the height tends to normalize by adulthood [1,2]. Facial appearance typically includes a high anterior hairline, macrocephaly, frontal bossing, a long thin face, frontotemporal hair scarcity, and downward slanting palpebral fissure. The other cardinal features that have been described are non-progressive hypotonia, delayed language and motor development, and mild mental retardation. In addition,

cardiac defects, genitourinary anomalies, seizures, scoliosis, and increased risk of tumorigenesis have been reported [1,3]. There are few reports that deal with the anesthetic implications in characteristic craniofacial anomalies, hypotonia, and other coexisting conditions associated with Sotos syndrome [4–6]. Herein, we report the perioperative management of a child with Sotos syndrome who underwent a right hydrocelectomy under general anesthesia.

CASE REPORT

A 4-year-old boy with Sotos syndrome was scheduled for right hydrocelectomy. He was born normally at a gestational age of 38⁺⁶ weeks with a birth weight of 3,600 g. Ventriculomegaly was identified at 6 months of age on cranial imaging, but it did not result in any long-term problems. Motor developmental delay was initially detected when he was not able to creep at 9 months of age. Additionally, he suffered from clumsiness, truncal muscle weakness, lack of social interaction, mild mental retardation, and severely delayed language development (ability to speak only one or two simple words). The diagnosis of Sotos syndrome was confirmed by genetic testing at 18 months of age. He had received regular occupational, speech, and adaptive physical therapy and had no surgical history. A recent transthoracic echocardiogram revealed that the ventricular septal defect that was identified in the newborn period had closed spontaneously and there were no significant cardiac abnormalities except for mild aortic regurgitation.

On physical examination, macrocephaly with head circumference above the 97th percentile (54.2 cm) was noted while height and weight were at the 75th percentile (108.1 cm and 19.4 kg, respectively). Other clinical features included mild scoliosis, long thin face, frontal bossing, dolichocephaly, fron-

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totemporal hair scarcity, high anterior hairline, and large hands and feet. No other abnormality was noted during the preoperative evaluation except mild cardiomegaly (cardiothoracic ratio = 0.58). Airway evaluation was difficult to perform because of poor cooperation. As premedication, 100 µg of glycopyrrolate was administered by intramuscular injection.

On arrival in the operating room, Ringer's lactate solution was infused at 60 ml/h via a 22-gauge intravenous cannula, which was inserted in the ward. Even though he was calm and cooperative with his mother present in the operating room, we took precautions to prevent any accidental fall due to unexpected agitation. Anesthesia was induced with 5 mg/kg of thiopental sodium and sevoflurane. Since preoperative evaluation of the cervical spine was not conducted, mask ventilation was performed with a head neutral position. However, the large occiput caused difficulty in head positioning and this problem was easily overcome by removing a pillow and fixing the head with the help of an assistant. We decided to give 0.3 mg/kg of rocuronium to facilitate endotracheal intubation after initiating ulnar nerve neuromuscular monitoring. Train-of-four count (TOF) 0 was obtained at 4 minutes following the rocuronium injection. Endotracheal intubation was easily performed with the help of a Macintosh size 2 blade at the first attempt, after visualizing the vocal cords with a grade 1 on Cormack-Lehane classification. Internal diameter of the tube used was 4.5 mm and it was fixed at 17 cm at the right corner of his mouth. Correct tube positioning was confirmed by the method of intentional right endo-bronchial intubation followed by withdrawal of the tube by 2 cm, and capnography. The cuff pressure was set at 20 cmH₂O. Anesthesia was maintained with 2–3% sevoflurane-medical air-oxygen (fraction of inspired oxygen being 50%). It took about 10 min for the first twitch of TOF to return. Fentanyl citrate (about 1 µg/kg) was administered to relieve postoperative pain and to prevent emergent agitation at the start of the operation. Duration of the surgical operation was about an hour. At that time, the TOF ratio had recovered to 44% and he received pyridostigmine 5 mg (0.25 mg/kg) with glycopyrrolate 0.2 mg. He was extubated when awake. However, we could not determine the TOF ratio because he had detached the peripheral nerve stimulator leads during emergence. He did not complain of any problems such as severe emergent agitation, respiratory depression, severe pain, or nausea during his time at the post-anesthesia care unit and he was transferred to the general ward.

DISCUSSION

Sotos syndrome is a rare genetic disorder characterized by typical facial abnormalities, developmental delay, and macrocephaly [2]. Majority of the cases are sporadic and associated with NSD1¹⁾ abnormalities [3]. Pathognomonic facial features of Sotos syndrome include frontal bossing, macrodolichocephaly, high anterior hairline, frontotemporal hair sparsity, and downslanting palpebral fissures; these are distinctive in children aged 1 to 6 years [1–3]. Non-progressive hypotonia, clumsiness, mental retardation, and delayed motor and language development are also present in Sotos syndrome [2]. Other clinical manifestations include cardiac and renal anomalies that are variable in type and severity, ventricular dilatation on cranial imaging, seizures, scoliosis, and tumorigenesis [1–3].

Endotracheal intubation has been shown to be easy [4,6,7] despite the oral findings such as high arched palate, pointed chin, and premature erupted teeth [8]. However, preoperative airway assessment was hard to perform since the patient was very uncooperative in this case. We omitted the use of preoperative anxiolytic agents such as midazolam and ketamine to secure the airway. Instead of using sedatives, we allowed parental presence during the induction of anesthesia and stay in the post-anesthesia care unit to allay the fear and anxiety of the child and to increase his compliance [9]. He seems satisfied with the overall anesthetic care. Glycopyrrolate 5 µg/kg was administered intramuscularly as an anticholinergic premedication. In our case, depth of insertion of the orotracheal tube was 3 cm deeper than that obtained through the standard formula ($12 + \text{age}/2$). A previous case report had also demonstrated deeper endotracheal tube insertion in a child with Sotos syndrome [4]. In our case, we assumed head rotation to the right side withdrew the endotracheal tube tip from the carina and his large occipital size influenced the tube placement [10].

We were concerned about the use of muscle relaxants in patients with Sotos syndrome because of hypotonia. However, no complications associated with muscle relaxant use have been reported in cases of this syndrome [6,11]. We surmise that patients with Sotos syndrome do not have any significant aberrant clinical pharmacologic effects with the use of muscle relaxants. In addition, the use of muscle relaxants has been found to diminish pharyngolaryngeal symptoms after extubation and provide better conditions for intubation than muscle

1) nuclear receptor binding SET domain protein 1

relaxant-free induction [12]. Therefore, we administered low-dose rocuronium [13] for tracheal intubation under neuromuscular monitoring. No additional muscle relaxants were administered during the operation. The recovery trend of TOF in our case showed a normal response with no postoperative muscular hypotonia.

Fentanyl has been reported to prevent emergence agitation under sevoflurane anesthesia in children and decrease postoperative pain [14]. We focused on the effect of fentanyl to prevent emergence agitation rather than postoperative respiratory depression as we had anticipated low compliance due to mental retardation [14]. Severe emergence agitation might result in physical trauma to the child, eventually requiring supplemental sedatives or analgesics. Additionally, we knew that maintaining a patent airway was easy in this case and the neuromuscular function was found to recover without any significant delay.

We were concerned about the patient's poor cooperation and hence did not consider regional anesthesia. There has been a report of a case of Sotos syndrome which regional anesthesia was administered for neurological flat foot surgery [5]. However, in that case, the authors also sedated the 7 year-old child with sevoflurane [5]. Hence, we decided that general anesthesia provided safer and comfortable conditions for the operation.

It is important to identify coexisting clinical features associated with Sotos syndrome preoperatively. Preoperative cardiac evaluation should be performed due to high incidence of congenital heart defects in Sotos syndrome. Antibiotic prophylaxis for infective endocarditis is necessary in unrepaired cyanotic congenital heart disease (CHD), completely repaired congenital heart defect with prosthetic material during the first 6 months after the procedure, and repaired CHD with a residual defect at the site or adjacent to the site of a prosthetic device [15]. Checking preoperatively for premature eruption of teeth might be helpful to avoid trauma to the teeth during the induction and emergence period, and to obtain appropriate informed consent from the parents. It may be better to choose anesthetics that increase seizure threshold because of the high prevalence of seizures in Sotos syndrome. Laboratory investigations should be performed appropriately to manage any other medical problems associated with this syndrome.

In conclusion, we state that airway management in a patient with Sotos syndrome might not be difficult despite the presence of craniofacial abnormalities. Muscle relaxants and opioids are also safe despite the presence of hypotonia in this

syndrome. In addition, understanding the comorbidities of patients with Sotos syndrome is essential for anesthetic management.

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