



# Anesthetic considerations for a pediatric patient with Wolf-Hirschhorn syndrome: a case report

Masanori Tsukamoto<sup>1</sup>, Hitoshi Yamanaka<sup>2</sup>, Takeshi Yokoyama<sup>2</sup>

<sup>1</sup>Department of Dental Anesthesiology, Kyushu University Hospital, Fukuoka, Japan

<sup>2</sup>Department of Dental Anesthesiology, Faculty of Dental Science, Kyushu University, Fukuoka, Japan

Wolf-Hirschhorn syndrome is a rare hereditary disease that results from a 4p chromosome deletion. Patients with this syndrome are characterized by craniofacial dysgenesis, seizures, growth delay, intellectual disability, and congenital heart disease. Although several cases have been reported, very little information is available on anesthetic management for patients with Wolf-Hirschhorn syndrome. We encountered a case requiring anesthetic management for a 2-year-old girl with Wolf-Hirschhorn syndrome. The selection of an appropriately sized tracheal tube and maintaining intraoperatively stable hemodynamics might be critical problems for anesthetic management. In patients with short stature, the tracheal tube size may differ from what may be predicted based on age. The appropriate size ( internal diameter ) of tracheal tubes for children has been investigated. Congenital heart disease is frequently associated with Wolf-Hirschhorn syndrome. Depending on the degree and type of heart disease, careful monitoring of hemodynamics is important.

**Keywords:** Anesthesia, General; Intubation, Intratracheal; Wolf-Hirschhorn Syndrome.



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Wolf-Hirschhorn syndrome is a rare hereditary disease that results from 4p chromosome deletion and was initially described by Cooper and Hirschhorn in 1961 [1-3]. The frequency is estimated as one per 50,000-20,000 [2,3]. Patients with this syndrome are characterized by craniofacial dysgenesis, seizures, growth delay, intellectual disability, and congenital heart disease [1-3]. Although several cases have been reported, very little information is available on anesthetic management for Wolf-Hirschhorn syndrome [1]. We encountered a case requiring anesthetic management for a 2-year-old girl with Wolf-Hirschhorn syndrome. She had postoperative atrial septal defect (ASD) closure, short stature, and low weight. In this case, the selection of an appropriately sized tracheal tube and maintaining intraoperatively stable

hemodynamics might be critical problems for anesthetic management.

## CASE REPORT

The patient was a 2-year-old girl, 72.9 cm in height and 6.2 kg in weight. She was born through a normal delivery and weighed 2010 g at birth. Using gene analysis, she was diagnosed with Wolf-Hirschhorn syndrome. She had short stature, congenital heart disease, including ASD and ventricular septal defect (VSD), mental retardation, and bilateral cleft palate, but no micrognathia or seizures. She was scheduled for cleft palate repair under general anesthesia. Her past surgical

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Corresponding Author: Masanori Tsukamoto, Department of Dental Anesthesiology, Kyushu University Hospital, 3-1-1 Maidashi, Higashi-ku, Fukuoka 812-8582, Japan  
Tel: +81-92-642-6480 Fax: +81-92-642-6481 E-mail: [tsukamoto@dent.kyushu-u.ac.jp](mailto:tsukamoto@dent.kyushu-u.ac.jp)

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history included an ASD and VSD closure.

On preoperative anesthesia evaluation, echocardiogram and electrocardiogram (ECG) were normal. In addition, chest radiographic findings were normal and laboratory test results were also within normal limits. The tracheal diameter at the level of the sixth cervical (C6) vertebra on her chest radiograph was 5.4 mm. She could speak only a few words, but we could not communicate with her. She could take a sitting position, but could not stand by herself. The clinical assessment of her airway was difficult.

On admission, her heart rate (HR) was 135 beats/min, and her oxygen saturation (SpO<sub>2</sub>) was 98% in room air. No premedication was given before she was transferred to the operating room. Anesthesia was induced with inhalation of sevoflurane 1-8% in oxygen after the start of noninvasive monitoring for SpO<sub>2</sub> (100%). After loss of consciousness, standard non-invasive monitoring was conducted, including ECG, HR (96 beats/min), blood pressure (BP) (85/40 mmHg), and bispectral index (BIS value: 54; spectral edge frequency [SEF]: 14-20 Hz, signal quality index [SQI]: 95%). Rocuronium (4 mg) was also administered intravenously to facilitate tracheal intubation. We could smoothly intubate by using a laryngoscope with a tracheal tube (inner diameter, 4.0 mm; Mallinckrodt Medical, Athlone, Ireland). The air leak around the tube was 20 cm H<sub>2</sub>O pressure. The correct position of the tube was confirmed using 5-point auscultation and capnography. Anesthesia was maintained with desflurane 8.5-9.0%, remifentanyl 0.1-0.15 µg/kg/min in air, and oxygen (FiO<sub>2</sub>: 0.4). BP was maintained at 75-95/45-57 mmHg, HR was 122-140 beats/min, end-tidal carbon dioxide was 35-40 mmHg, and BIS value was maintained between 40 and 58 with an SEF of 10-15 Hz and SQI of 95%. In addition, intermittent boluses of fentanyl were administered with reference to the BIS and hemodynamics. In addition, the patient's cardiac status was monitored using Aesculon<sup>®</sup> with a noninvasive cardiac function monitor. During anesthesia, BP was maintained at 80-110/50-70 mmHg, cardiac output was 3.1-3.9 L/min, cardiac index was 2.6-3.0 L/min/m<sup>2</sup>, stroke

volume was 10-14 ml, and stroke volume variation was 6-11%. The surgical procedure was completed uneventfully. The patient's body temperature was maintained at 36.4-37.5°C with a warming blanket during surgery. The operation was completed in 236 minutes without any surgical or anesthetic problems. There was minimal blood loss during the operation, and she subsequently received a total of 225 ml of lactated Ringer's solution, and her urine volume was 27 ml. She emerged from general anesthesia in 4 minutes after stopping desflurane inhalation. She was extubated after confirming sufficient spontaneous respiration. After extubation, her respiratory and hemodynamic conditions were stable.

## DISCUSSION

Wolf-Hirschhorn syndrome is a very rare hereditary disease resulting from partial loss of the distal short arm of chromosome 4. It is associated with a high mortality rate, approximately 30% within the first 2 years of life due to congenital heart disease [3]. Patients with Wolf-Hirschhorn syndrome have characteristic features including growth delay followed by short stature and slow height gain, variable degrees of intellectual disability, epilepsy, and heart disease. The selection of an appropriately sized tracheal tube and maintaining intraoperatively stable hemodynamics might be critical problems for anesthetic management.

It is known that an unexpected tracheal tube size might be needed for patients with short stature [4]. The appropriate size (internal diameter) of tracheal tubes for children have been investigated [5,6]. Simple formulas have been proposed using weight and height to predict the optimal tracheal tube size, and the age of the child (in years) has been used in formulas to select the size of the tracheal tube. However, it was reported that age-based formulas [(age/4) + 4] have been inaccurate in up to 60% of children [4]. In this patient, tracheal intubation was needed to perform cleft palate repair. The tracheal tube was selected according to her tracheal

internal diameter at C6, her age, and her height. Patients with short stature might have a narrowed subglottic or tracheal airway compared with healthy patients. Thus, it may be necessary to consider using a smaller tracheal tube than would be predicted according to age.

Congenital heart disease is frequently associated with Wolf-Hirschhorn syndrome. Depending on the degree and type of heart disease, careful monitoring of hemodynamics is important. In this patient, the ASD, VSD, and PS would be considered mild. Anesthetic agents and hemodynamic responses to them might cause significant circulatory disturbances. We carefully administered anesthetics with titration in reference to hemodynamics change such using Aesculon<sup>®</sup> [7].

Malignant hyperthermia (MH) in Wolf-Hirschhorn syndrome has been previously reported, although our patient did not develop MH. The relationship between Wolf-Hirschhorn syndrome and MH might require further investigation [1]. It was reported that MH could occur during general anesthesia and after a surgical procedure. The symptoms of MH are tachypnea, tachycardia, and hyperthermia [1]. If such symptoms develop, anesthesiologists must consider other possible causes such as overheating and infection and to administer dantrolen.

#### AUTHOR ORCIDs

Masanori Tsukamoto: <https://orcid.org/0000-0001-6848-0870>

Hitoshi Yamanaka: <https://orcid.org/0000-0003-1303-9660>

Takeshi Yokoyama: <https://orcid.org/0000-0003-3017-8865>

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