Alternative Methods for Nasotracheal Intubation and Extubation in a Patient With Apert Syndrome

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Apert syndrome is a rare autosomal dominant disorder characterized by craniofacial abnormalities, craniosynostosis and syndactyly. Nasotracheal intubation for a patient with Apert syndrome can be a challenge because of abnormal facial anatomy. We experienced the anesthetic management of a patient with Apert syndrome who underwent partial resection of mandible and cleft palate repair with nasotracheal intubation. Nasotracheal intubation using a gastric tube and extubation using an airway exchange catheter proved useful in this case of airway compromise.

Key Words: Apert syndrome; General anesthesia; Nasotracheal intubation.

In cases with congenital anomaly in the oral and maxillofacial region, procedures for corrective surgery frequently require nasotracheal intubation for optimal surgical access and to secure the airway. Reports in patients with Apert syndrome revealed airway concerns including stiff or vertically fused tracheal rings, macroglossia, and tonsillar and adenoid hypertrophy.¹ In addition, it has also been reported that the nasotracheal tube, when it is advanced through narrow nasal passages, may injure the nasal tissue and/or be inserted into inappropriate tissue planes. There are reports that surgery has been canceled because of the bleeding from the posterior wall of the nasopharynx.^{2,3} Since severe craniofacial anomalies often make it difficult to perform nasotracheal intubation and extubation, there have been various techniques attempted to ensure safe airway management.^{3,4}

We describe a potentially useful and convenient method for nasotracheal intubation and extubation in a patient with Apert syndrome undergoing biopsy and resection of the tumor in the mandible.

CASE REPORT

A 14-year-old girl with Apert syndrome received general anesthesia twice for biopsy of a tumor in the left mandible and for its resection. She was diagnosed with Apert syndrome by genetic testing when she was an infant having had the typical clinical features of craniofacial abnormalities, craniosynostosis, and syndactyly of all limbs. She displayed mental retardation, and her intellectual development corresponded to a 5-year-old child. She had undergone syndactyly repair at the age of 3 months and Le Fort II osteotomy, cleft palate repair, and decompressive craniectomy at the age of 3 years. On physical examination, she was 54 kg in weight and 164 cm in height. She had craniofacial abnormalities, syndactyly, exophthalmos, macroglossia, maxillary dental arch stenosis, and tonsillar hypertrophy. Mallampati grade was class 3.

At the first biopsy surgery, anesthesia was induced with inhalation of sevoflurane in oxygen after the start of noninvasive monitoring for blood pressure, electrocardiogram, oxygen saturation (SpO₂), and temperature. Concentration of sevoflurane was initially 1% and was gradually increased to 8%. Mask ventilation was easy. Midazolam 2 mg and fentanyl 100 µg were given after peripheral intravenous access was obtained. Rocuronium 30 mg was administered intravenously to facilitate tracheal intubation. We evaluated both sides of the nasal

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Figure 1. The distal end of the gastric tube by a Magill forceps was placed through the tracheal tube to outside.

cavity with a swab of 2% lidocaine containing 1:200,000 epinephrine and 7.0 mm nasopharyngeal airway. A 14 Fr gastric tube (Covidien, Tokyo, Japan) was used as a guide to allow a 7.0-mm nasotracheal tube to pass easily through the nasal cavity. The tip of the gastric tube was pulled out from the mouth using a Magill forceps (Figure 1), and the nasotracheal tube was advanced into the pharynx via the gastric tube. We confirmed that there was no bleeding of the nasal cavity. After removing the gastric tube, manual ventilation was possible through the nasotracheal tube. We were able to intubate smoothly by using a laryngoscope. A fiber-optic bronchoscope was available if difficulty in conventional laryngoscopy occurred. Anesthesia was maintained with sevoflurane, air, and oxygen in addition to intravenous remifentanyl. The surgery proceeded uneventfully. She was extubated when fully awake from the anesthesia and did well, with stable vital signs. A few weeks later, the tumor in the left mandible was diagnosed as mandible odontogenic myxoma.

Two months after the initial operation, the patient underwent resection of the right mandibular odontogenic myxoma from the left second premolar to the right second molar region with immediate iliac bone grafting using the greater auricular nerve and reconstruction plate. Anesthesia was induced and maintained similarly as was done previously. The duration of surgery was more than 8 hours. Blood loss volume was about 1000 mL, urine volume was about 3000 mL, and total fluid resuscitation was with normal saline 200 mL, Ringer's solution 3900 mL, and 700 mL of colloid solution. At the end of the operation, we evaluated external facial swelling as well as upper airway edema using a fiber-optic scope. Suspecting possible airway obstruction after extubation, we used an 11 Fr airway exchange catheter (AEC; Cook Critical Care, Bloomington, Ind; Figure 2).

Figure 2. Nasotracheal tube with the Cook airway exchange catheter.

After inserting the AEC into the tube, the nasotracheal tube was extubated out of the trachea, and we observed spontaneous respiration. After we confirmed upper airway patency, we removed the AEC. After extubation, her respiratory and hemodynamic conditions were stable.

DISCUSSION

Apert syndrome is an inherited, autosomal dominant disorder with an incidence rate of 1/100,000.^{5,6} It is defined by the triad of syndactyly, craniofacial abnormalities, and craniosynostosis. Other conditions include developmental delay, bulging and downward-slanting eyes at the lateral canthus, cleft palate, abnormal occlusion, cervical vertebral fusion, and hydrocephalus.⁷ Procedures for surgery in the oral region usually require nasotracheal intubation for surgical access and a secure airway. Of critical concern is the possibility of difficult airway due to facial abnormal abnormalities and the patency of postoperative upper airway secondary to postoperative edema.

We performed nasotracheal intubation for this procedure although alternative airway management options such as submental intubation with use of a flexible or wire-reinforced endotracheal tube or tracheotomy could have been considered. Both were felt to be more invasive than the traditional nasotracheal approach.

In the first biopsy surgery, we were able to overcome the difficult nasotracheal intubation by guiding the nasotracheal tube via a gastric tube. We pulled out the tip of the gastric tube by a Magill forceps and advanced the tracheal tube from the nasal cavity to the pharynx without bleeding or damage to the nasopharynx.⁸

In the second surgery, in which we were also concerned with postoperative airway edema, we used the AEC to allow rapid reintubation should compromised

upper airway patency due to postoperative edema around the epiglottis be encountered, as has been previously reported.⁴ In addition, the AEC can be used to temporarily ventilate and provide oxygen until the airway can be secured. The AEC may result in a decrease in the incidence of complications when difficult tracheal reintubation may be required. The postoperative follow-ups during the next 24 hours did not show a need for sedatives or analgesics. No complications related to anesthesia were encountered.

We report a case of successful anesthetic management with airway-exchanging techniques in a patient with Apert syndrome who had craniofacial anatomic abnormalities.

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