

Anesthesia and Airway Management in Two Cases of Apert Syndrome: Case Reports

Apert Sendromlu İki Olguda Hava Yolu Yönetimi ve Anestezi: Olgu Sunumu

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Abstract

Apert syndrome is a type of acrocephalosyndactylia that consists of craniofacial synostosis, midface hypoplasia and syndactyly, with an autosomal dominant inheritance pattern. During anesthesia, difficult intubation and ventilation may be observed because of abnormal airways.

In one of our patients, visceral anomalies, such as esophageal stricture and post-strictural dilatation, may cause respiratory problems because of aspiration. The second case was a Mallampati grade 2 with craniofacial synostoses, midface hypoplasia and syndactyly.

In the case of apert syndrome, anesthetists must be prepared for intubation difficulties, airway and ventilation problems and even visceral anomalies.

Keywords: Airway and ventilation problems, Anesthesia, Apert syndrome

Özet

Apert Sendromu otozomal dominant geçişli, sindaktili, midface hipoplazisi ve kraniofasiyal sinostosisin eşlik ettiği entübasyon ve ventilasyon güçlüğü olabilen bir yenidoğan anomalisidir.

4 yaşında mallampati grade 2, kraniostosis, midface hipoplazi ve İnkomplet nasal obstruksiyonlu ve 2,5 yaşında doğumdan itibaren solunum problemleri, beslenme zorluğu ve post strüktürel dilatasyon nedeniyle aspirasyon problemleri olan Apert sendromlu 2 olguya operasyon planlandı.

Apert sendromlu hastalarda hava yolu dismorfizmine ve visceral anomalilere karşı uyanık olunmalıdır. Alt ve üst solunum yolu enfeksiyonları, sekresyon artışı gibi solunum yolu problemleri ve zor entübasyon ihtimaline karşı, spontan solunum korunur şekilde entübasyon denemelidir.

Anahtar Kelimeler: Anestezi, Apert sendromu, Ventilasyon ve hava yolu problemleri

Introduction

Children with craniofacial anomalies often have abnormal airways. Apert syndrome is associated with the loss of lower and upper airway patency as well as respiratory complications during general anesthesia [1,2]. Apert syndrome is a type of acrocephalosyndactylia that consists of craniofacial synostosis, midface hypoplasia and syndactyly [1,2]. The inheritance is autosomal dominant, with a frequency of 1 in 100,000-160,000 [2,3]. Intubation and ventilation may be difficult during general anesthesia [1,4]. There may also be visceral and other anomalies [5].

Case Reports

Case I:

The first patient, a 4-year-old male in whom Apert syndrome was previously diagnosed, was scheduled for reconstructive surgery of the hands and feet, which demonstrated syndactyly. He had craniosynostosis and midface hypoplasia (Figure 1a,b) but no difficulty in feeding. Complete airway obstruction, snoring or sleep apnea problems were not been reported. There was, however, incomplete nasal obstruction. Preoperative airway assessment revealed a Mallampati grade 2 with a maximum interdental gap. There were no visceral or other systemic anomalies. The laboratory findings were normal. Lateral craniography showed midface hypoplasia and a shortened anterior cranial base (Figure 1c). The thyrohyoid distance and the atlanto-occipital joint movement were normal. Premedication consisted of intramuscular (im) midazolam (0.1 mg·kg⁻¹) and atropine (0.015 mg·kg⁻¹). Non-invasive blood pressure, electrocardiogram (ECG) and SpO₂ were monitored. Since we anticipated a difficult intubation, the induction of anesthesia was carried out with inhalation anesthetics. The anesthetic induction occurred with sevoflurane in nitrous oxide/oxygen (65%-35%) via a facemask followed by laryngoscopic oral intubation without neuromuscular blockers. A 4-mm



Fig. 1 — First case, demonstrating face, hands and lateral craniography

internal diameter (ID) endotracheal tube with an inserted curved guide wire was passed at the second laryngoscopy. Anesthesia was maintained with 2%sevoflurane in nitrous oxide/oxygen. The patient was extubated after surgery with no anesthetic complications.

Case II:

The second patient, a male aged 2.5 years, had been diagnosed with Apert syndrome at birth. He had a maximum mouth opening (Figure 2a) and a grade 3 intubation score. There was coronal synostosis, midface hypoplasia, syndactyly, esophageal stricture and post-strictural dilatation (Figure 2b, c). He had difficulty with feeding, malnutrition and respiratory problems since birth, for which he had been treated several times in the pediatric intensive care unit (PICU). Several general anesthetics with endotracheal intubation had previously been given for esophageal dilatation.

Pre-operative laboratory findings were normal. After 6 h of fasting, atropine 0.015 mg·kg⁻¹ and midazolam 0.1 mg·kg⁻¹ were given im for premedication. Non-invasive blood pressure, ECG and SpO₂ were monitored. Induction and maintenance were performed with sevoflurane in nitrous oxide/oxygen (65:35). Following intravenous atracurium (0.1 mg·kg⁻¹), tracheal intubation with a 3-mm ID uncuffed endotracheal tube was easily performed. Nissen modification surgery and esophageal dilatation were performed. During anesthesia, there was wheezing, and clear secretions were frequently aspirated from the endotracheal tube. After extubation, nasotracheal aspiration was frequently necessary in the recovery room. The patient was returned to the intensive care ward when swallowing returned to normal.

Discussion

Anesthetists should be aware that airway dysmorphism carries a risk of difficult intubation and respiration problems in patients with Apert syndrome [2]. Craniofacial anomalies are often associated with airway obstruction, especially during sleep, and can cause obstructive sleep apnea [6,7]. However, preoperative evaluation showed airway or other problems in our patients. Elwood et al. [1] found respiratory complications in 33% of patients with Apert syndrome. As in our cases, upper and lower respiratory infections were due to respiratory complications from the underlying condition, particularly increased secretions [1,3], as in our first patient.

The following classical features should be evaluated preoperatively:

- high Mallampati score
- reduced thyromental distance
- reduced sternomental distance
- restricted atlanto-occipital joint movement
- restricted temporo-mandibular joint function
- limited mouth opening

Intubation aids, such as Combitube, laryngeal masks and emergency tracheotomy sets must be available. Since the first patient had not been previously intubated, spontaneous breathing was preserved with inhalation agents, and neuromuscular

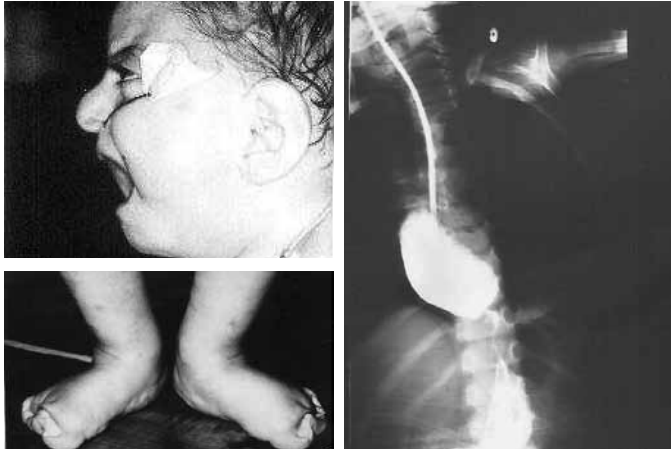


Fig. 2 — Second case, demonstrating maximum mouth opening, syndactyly and esophageal stricture with post-stricture dilatation

blockers were avoided. Intubation had been previously successful in the second patient; therefore, we felt confident in using neuromuscular blockers.

Cartilaginous abnormalities of the trachea, fusion of the cervical vertebrae, tracheal stenosis and angular deviation of the

trachea may contribute to respiratory morbidity or difficult intubation in Apert syndrome [3]. Tracheotomy is difficult in children and may lead to complications; percutaneous cricothyroidotomy is less likely to be required in children with craniofacial anomalies, such as Apert syndrome [6,7]. Fiberoptic and retrograde intubations, which are difficult in children, have been used in craniofacial anomalies [2,6]. Visceral anomalies may also cause complications [2]; for instance, the gastrointestinal anomalies in our second case required prolonged PICU treatment.

We prepared laryngeal masks and emergency tracheostomy sets for our cases, although neither was needed. However, in Apert syndrome, the anesthetist must be ready for airway problems, intubation difficulties and even visceral anomalies. In particular, Apert syndrome with visceral anomalies may cause significant problems for anesthesia and surgery. In addition, if difficult airway management is a concern, the induction of the anesthesia should be administered with inhalation agents and without neuromuscular blockers to maintain spontaneous ventilation. For these reasons, thorough pre-operative evaluation and planning is essential.

Conflict interest statement The authors declare that they have no conflict of interest to the publication of this article.

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