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CASE REPORTS

Anesthetic management of scoliosis operation in a pediatric patient with Frank-ter Haar syndrome: a case report

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Abstract Frank-ter Haar syndrome is a rare disorder characterized by multiple skeletal, cardiovascular abnormalities, and facial features. Some of these characteristic facial features are important for anesthesiologists to predict the difficult airway. We present the anesthesia management of an 8-year-old boy with Frank-ter Haar syndrome who underwent posterior spinal instrumentation operation for scoliosis. In these patients, it is vital to anticipate possible difficult intubation before surgery and make all necessary preparations.

Introduction

Frank-ter Haar syndrome (FTHS) is a rare autosomal-recessive disease and characterized by skeletal, cardiovascular, and ocular abnormalities. Mutations in the *SH3PXD2B* gene on chromosome 5q35.1 are the most common underlying genetic defect in FTHS.[1] The syndrome is characterized by multiple skeletal abnormalities, developmental delay, and characteristic facial features (unusually large cornea, flattened back of the head, wide fontanels, prominent forehead, widely spaced eyes, prominent eyes, full cheeks, and small chin). Very few cases have been reported worldwide.[1] Some of these characteristic features are important for anesthesiologists to predict the possible difficulties during induction of anesthesia and intubation. As far as we know, there are few reports about anesthetic management of a patient with FTHS.

Case report

An 8-year-old boy, weighing 21 kg, was admitted to the hospital with scoliosis. After written informed parental consent was obtained, posterior spinal instrumentation was planned for the patient. He was born by cesarean-section delivery at term. The patient had the diagnosis of FTHS. Atrial septal defect, ventricular septal defect, and patent ductus arte-

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riosus had been detected in the neonatal period. He had cleft lip, cleft palate, and pes equinovarus. He had surgery for closure of the ventricular septal defect and cleft lip and palate. There were no past medical reports available. He was diagnosed with heart failure. He was under pediatric cardiology follow-up and on tablet enalapril 5 mg once a day. He had a history of frequent respiratory tract infections, and his last infection was 15 days before with cough and purulent sputum which was treated by 7 days of antibiotic medication.

The patient's general physical examination revealed typical facial dysmorphism, gingival hypertrophy, pectus carinatum, and thoracolumbar kyphoscoliosis. Abnormal blood gas analyses and decreased lung volumes in pulmonary function tests were detected because of the restrictive lung capacity. Examination of the cardiovascular system revealed a pan systolic murmur on the apex. Echocardiogram showed a dilated left atrium, severe mitral valve regurgitation, and mild aortic valve regurgitation. The pediatric cardiology unit suggested prophylaxis for infective endocarditis with ampicillin 50 $mg.kg^{-1}$ and gentamicin 2 $mg.kg^{-1}$. Routine preoperative laboratory tests were normal. The child had retrognathia and micrognathia and a mild restriction in his neck movements. He had a grade 3 Mallampati score. According to these findings, arrangements were made for the patient with a difficult airway such as laryngeal mask airways (LMAs), a Storz DCI (Karl Storz, Tuttlingen, Germany) C-MAC video laryngoscope, fiberoptic bronchoscope, and variable sizes of endotracheal tubes.

In the operation room, standard monitoring was installed, including SpO_2, noninvasive blood pressure, and an electrocardiogram. Subsequently, anesthesia induction was performed with 8% sevoflurane carried by 6 $L.min^{-1}$ oxygen flow. The mask ventilation was successful, and anesthesia was induced with fentanyl 1 $mcg.kg^{-1}$ and rocuronium 0.6 $mg.kg^{-1}$ intravenously. At first, a C-MAC videolaryngoscope was used to intubate the patient, but there was no view of the epiglottis or vocal cords. The Cormack-Lehane grade was 4. Then, a Fastrach LMA (size 3) was placed, and it provided adequate ventilation. The child had a low body mass index, but his upper airway was suitable for the Fastrach LMA (size 3). As the patient would be operated in the prone position, a secure airway with tracheal intubation was needed. The endotracheal tube (size 5.0) was inserted through the Fastrach LMA. The first attempt failed with esophageal intubation. Afterwards, the patient was ventilated through the LMA after oxygenation. This time, endotracheal intubation was attempted with a smaller-sized 4.5 cuffed endotracheal tube. Confirmation of endotracheal intubation was made with a capnograph and auscultation. The anesthesia was maintained with oxygen/air and infusions of remifentanil and propofol. The left femoral artery was cannulated, and the right femoral vein was catheterized. The patient was turned prone for posterior spinal instrumentation. Somatosensory-evoked potentials and motor-evoked potentials were monitored during the operation. A loading dose of 10 $mg.kg^{-1}$ followed by a continuous infusion of 1 $mg.kg^{-1}.h^{-1}$ tranexamic acid was given to the child until skin closure. Throughout the surgery, an Hb threshold of 8 $g.dL^{-1}$ was aimed to be kept with blood-gas monitoring. The patient was transfused with pediatric erythrocyte suspensions of 25 $mL.kg^{-1}$ (~525 mL) and fresh frozen plasmas of 15 $mL.kg^{-1}$. The urine output was 230 mL. To prevent airway edema, the child received 20 mg of methylprednisolone. The duration of the operation was 6 hours. Intravenous paracetamol of 15 $mg.kg^{-1}$, tramadol of 1 $mg.kg^{-1}$, and morphine of 0.1 $mg.kg^{-1}$ were administered before the end of the surgery. The patient was sent to the intensive care unit in intubated state for mechanical ventilator support. Within 5 days of admission to the intensive care unit, he was extubated without any complications. Morphine-based intravenous patient-controlled analgesia was used to maintain postoperative analgesia. Self-reported pain intensity was assessed by using the Numerical Rating Scale 0–10. The postoperative period was uneventful with no changes worthy of note. He was transferred to the service of orthopedics and traumatology clinic. The patient was discharged home on the tenth postoperative day.

Discussion

FTHS is a congenital syndrome affecting mainly the skeletal system and has ocular and facial features. It was described by Frank and Ter Haar in 1973.[2] The most common abnormalities associated with this syndrome are brachydactyly, hypertelorism, wide nasal bridge, wide mouth, and depressed nasal bridge. Skeletal system anomalies like abnormality of the metacarpal bones, kyphosis, scoliosis, osteolysis, and beaking of vertebral bodies have been described. The most common ocular finding is macrocornea with or without glaucoma.[1] Cardiac problems like mitral valve prolapse or other congenital heart defects may be present. Maxillodental anomalies such as mandibular prognathy, gingival overgrowth, or premature loss of teeth may be observed.[3]

Even though the patient had multiple congenital heart defects, which were atrial septal defect, ventricular septal defect, and patent ductus arteriosus, his upper airway anatomy causing difficult endotracheal intubation was one of the most challenging features for us as anesthesiologists. Moreover, due to severe kyphoscoliosis, the patient had restrictive lung disease which made the postoperative intensive care unit follow-up obligatory.

In this case, predicting the difficult intubation at the preoperative examination was crucial. The first step was to prepare a wide range of equipment for intubation. According to the 2015 Difficult Airway Society Guideline, if tracheal intubation fails by laryngoscopy, the second step is placement of supraglottic airway devices. There are four options that may be applied after placement of a supraglottic airway device: (i) waking the patient up, (ii) intubating the trachea via the supraglottic airway devices, (iii) proceeding without intubating the trachea, or (iv) tracheostomy/cricothyroidotomy.[4] In the presented case, when we could not obtain an image with the C-MAC video laryngoscope, in accordance with the Difficult Airway Society Guideline, we switched to the second step and placed a Fastrach LMA to the patient. The patient was then intubated by placing an endotracheal tube through the LMA. In another case reported by Tommasino et al. (2018), difficult intubation was encountered in a child with the same syndrome.[3] In the paper they presented, the child was a 5-year-old male who underwent dental restoration under general anesthesia.

He had micrognathia, arched palate, and kyphosis. When he was 4 years old, difficult intubation was reported. In view of this, they prepared for a potentially difficult intubation. In our case, the patient was an 8-year-old male who underwent posterior spinal instrumentation operation for scoliosis. His neck movements were limited, while he also had retrognathia and micrognathia. The cardiac examination findings of both children were similar. In both cases, after induction with sevoflurane, the patients could be ventilated with a mask. Following this step, Cormack-Lehane grade 4 views were obtained in both of them by videolaryngoscopy. After that, they preferred to intubate the patient with a fiberoptic bronchoscope, whereas we intubated the patient by placing the LMA then delivering the tube through the LMA Fastrach.[3]

As widely known, major scoliosis surgeries generally cause significant blood loss. There are no definitive guidelines for patients who significantly bleed perioperatively. The American Society of Anesthesiologists guidelines offered restrictive 7.0 $g.dL^{-1}$ blood management protocols for adults, but they excluded infants, neonates and children under 35-kg.[5] The child was a 21-kg boy and there are no definitive suggestions for transfusion thresholds for this patient group. We preferred a little higher hemoglobin threshold of 8.0 $g.dL^{-1}$ because we were faced with active bleeding due to the operation, and we were managing with a little-known syndrome in a patient with cardiac anomalies.

There are few articles about the anesthetic management of children with FTHS, but he was one of the more challenging ones because we managed a major scoliosis surgery and a little-known syndrome at the same time. In these patients, it is vital to anticipate possible difficult intubation before surgery and make all necessary preparations. For patients undergoing scoliosis surgery, bleeding is also an important problem. Additionally, children with FTHS are challenging for anesthesiologists also due to potential cardiac problems and restrictive lung diseases. We think that the relevant medical specialists should be consulted before the surgery, and their recommendations about the patient should be taken into consideration.

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Zrhidri A, Jaouad IC, Lyahyai J, et al. Identification of two novel *SH3PXD2B* gene mutations in Frank-Ter Haar syndrome by exome sequencing: Case report and review of the literature. Gene. 2017;628:190–3.
2. Frank Y, Ziprkowski M, Romano A, et al. Megalocornea associated with multiple skeletal anomalies: a new genetic syndrome? J Genet Hum. 1973;21:67–72.
3. Tommasino C, Albicini M. Anaesthesia and orphan diseases: difficult tracheal intubation in a child with Frank-ter Haar syndrome. Eur J Anaesthesiol. 2018;35:542–4.
4. DAS guidelines for management of unanticipated difficult intubation; 2015 https://das.uk.com/guidelines/das_intubation_guidelines
5. American Society of Anesthesiologists Task Force on Perioperative Blood Management. Practice guidelines for perioperative blood management: an updated report by the American Society of Anesthesiologists Task Force on Perioperative Blood Management. Anesthesiology. 2015;122:241–75.