Case Report

Anesthetic Management in an Unanticipated Difficult Airway Because of Oropharyngeal Stenosis in a Patient With Pierre Robin Syndrome Undergoing Pediatric Congenital Heart Surgery: A Case Report

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ABSTRACT

Background: Difficult intubation in pediatric patients with a history of relatively common respiratory and cardiac problems and syndromic cases should always be considered. Pierre Robin syndrome is a rare birth defect characterized by a small jaw, tongue retraction, and upper airway obstruction, also known as Pierre Robin syndrome.

Case Report: We describe a 5-month-old boy, 3500 g in weight, suffering from Pierre Robin syndrome. The patient was a candidate for the surgical closure of ventricular septal defect (VSD) and patent ductus arteriosus (PDA). After the induction of anesthesia in the operating room, we encountered problems with intubation in that it was not possible to secure an airway with a tracheal tube via different methods. The patient woke up, and the surgery was postponed. His airway was then thoroughly evaluated before VSD and PDA surgical closure was performed successfully.

Conclusions: In elective surgical cases with unexpectedly challenging intubation, the surgery should be postponed until the airway is fully checked and a safe airway is secured. ***(Iranian Heart Journal 2023; 24(3): 94-99)***

KEYWORDS: Airway management, Cardiac surgery, Anesthesia, Pierre Robin syndrome, Congenital disease

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One of the formidable challenges encountered by anesthesiologists is patient intubation. Most patients referred for congenital heart surgery are children who have other syndromic problems as well. [1] Pierre Robin syndrome is a congenital problem of unknown origin. Patients with this syndrome have a large tongue and a small jaw and catch colds repeatedly. Often, cleft palate and tracheal narrowing appear. Pierre Robin syndrome is generally diagnosed at birth, but sometimes it is determined by chance and based on symptoms. [2] While the prevalence of this disease is equal in boys and girls, this complication is more common among twins.

Teamwork of specialists is required to improve the affected functions, including breathing, hearing, nutrition, and sleep. This disease is known as a sequence because of a series of events occurring during the development of the fetus. While the exact cause of this disease is unknown, the sequence of abnormalities that occur in the womb begins with a lack of normal jaw growth, causing a tongue shift and the formation of a U-shaped cleft palate. [3] Whereas some studies have pointed to genetic links, others have considered uterine pressure to be involved in the occurrence of this problem.

Congenital heart diseases are associated with Pierre Robin syndrome in 20% of sufferers. Ventricular septal defect (VSD), patent ductus arteriosus (PDA), and atrial septal defect (ASD) are among the most frequent associated cardiac disorders. [4]

Airway surgery for patients with Pierre Robin syndrome is preferred to all elective surgeries. If the patient is anesthetized for cardiac surgery, and intubation is not possible, the patient should be woken up, and the surgery should be postponed. If it is not feasible to postpone the surgery, the operation should be performed after an airway examination and, if necessary, after an elective tracheostomy. [5]

Our patient was electively scheduled for the surgical closure of VSD and PDA. Nonetheless, we unexpectedly found intubation impossible. We referred him to another center for a detailed airway evaluation. Finally, he underwent a successful surgical operation for VSD and PDA closure in our center.

CASE REPORT

The patient was a 5-month-old boy, 3500 g in weight. He was Asian and had a history of frequent colds and snoring at night. His medical records showed that the boy suffered from VSD and PDA based on an echocardiographic examination. Standard electrocardiography (heart rate: 145 bpm), noninvasive blood pressure measurement (97/55 mm Hg), pulse oximetry, and capnography were set. A physical examination revealed a smaller-than-usual jaw and irregular baby teeth (fig 1).

The patient was transferred to the operating room for the surgical closure of his VSD and PDA. Anesthesia was induced with 8% sevoflurane, and a venous line (No. 22) was placed in the right leg. Next, 2 μ/kg of fentanyl, 0.25 mg of midazolam, and 0.2 mg/kg of cisatracurium were injected. Hemodynamic changes were minimized with the induction of anesthesia. Several attempts were made to intubate the patient, all of which proved unsuccessful. Consequently, intubation was attempted with the help of a bogie, which was unsuccessful again. Afterward, a fibrotic device was employed, but it was still impossible to examine and visualize the epiglottis and trachea because of the patient's large tongue. It should be mentioned that a suitably-sized laryngeal mask airway was not available in the operating room. Considering that the surgery was elective, the patient was properly ventilated until proper awakening and full spontaneous breathing in the operating room. Subsequently, he was transferred to the pediatric intensive care unit (PICU).

The following day, in the PICU, a computed tomography scan with airway reconstruction (virtual bronchoscopy) was performed for a detailed assessment of the patient's airway. The results demonstrated the narrowing of the oropharynx area and upper tracheal stenosis (the glottic area) (fig 2).

Figure 1: The pictures present the facial features of the patient (the small jaw).

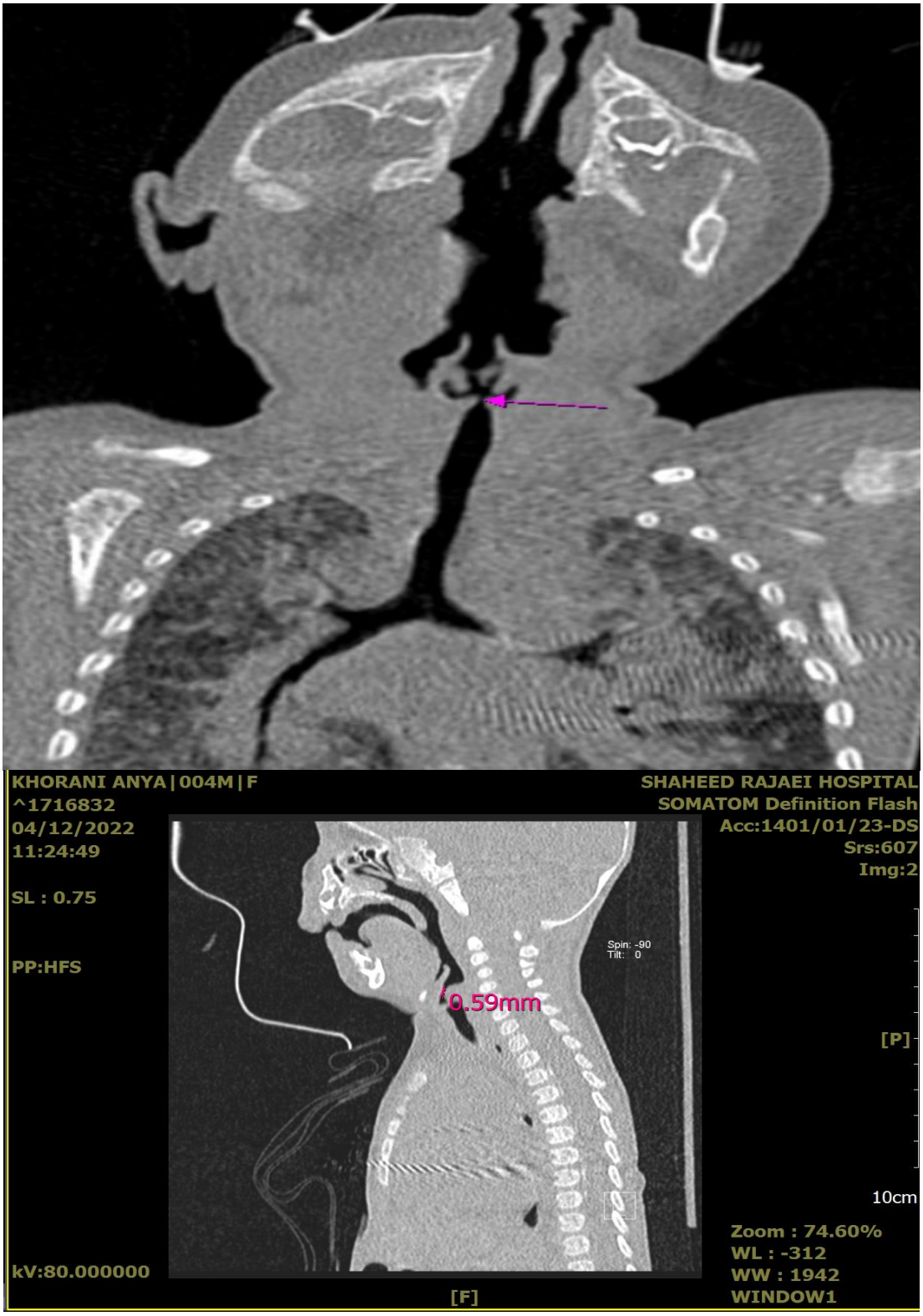

Figure 2: The computed tomography scans depict the narrowing of the oropharynx area and upper tracheal stenosis (the glottic area).

Two days later, the patient was admitted to the pediatric ward and then discharged from the hospital. Two days afterward, he was taken to an otolaryngology center because of his Pierre Robin syndrome symptoms. The boy was examined by an otorhinolaryngology specialist via a rigid bronchoscopy under anesthesia with spontaneous breathing. The specialist reported the narrowing of the oropharynx area and recommended cardiac surgery under anesthesia with intubation and without the need for elective tracheostomy. Additionally, the use of a soft bronchoscopy was advised.

The patient was transferred to our cardiac surgery center. Premedication was done with 0.15 mg/kg of midazolam and 1 µg/kg of fentanyl, and anesthesia was induced with 8% sevoflurane. With the aid of a soft bronchoscopy and spontaneous breathing, the boy was intubated (No. 4.5) without a cuff, and the intubation site was confirmed. Next, 0.2 mg/kg of cisatracurium was injected, and arterial and central venous lines were inserted. After a heparin injection of 400 units per body weight and the confirmation of an activated clotting time above 480, the patient was placed on the pump. The surgical operation was successfully accomplished. The duration of the surgery was 5 hours, and the cardiopulmonary pump time was 55 minutes. While still intubated, he was transferred to the PICU without incident. On the first postoperative day, given his stable hemodynamics, acceptable ABGs, and normal chest X-rays, the boy was returned to the operating room. He was extubated by a team of skilled anesthesiologists and under full monitoring. He was kept under observation for an hour before being transferred to the PICU with full consciousness and spontaneous breathing. In the next 3 days, the patient received special care and was transferred to the ward in stable cardiac and respiratory condition. Two days later, he was discharged in good general condition with no complications.

DISCUSSION

Our patient was susceptible to frequent colds according to his parents. The shape of the patient's jaw, which was smaller than normal, and his large and retracted tongue were also among the points that were noticed after the initial intubation failure. A preoperative virtual bronchoscopy determined the degree of airway narrowing and directed the otolaryngologist to only check tracheal narrowing with a rigid bronchoscopy, obviating the need for an elective tracheostomy before surgery. We woke the patient up after ensuring ventilation with an Ambu bag. Following a complete evaluation by an otolaryngologist and reassurances regarding the airway, intubation with a soft bronchoscopy and spontaneous breathing was recommended.

The prevalence of children with congenital heart diseases has been on the rise in recent years, and other symptoms that might be part of syndromic disorders should be taken into account in this group of patients. [6] Congenital heart diseases are associated with Pierre Robin syndrome in 20% of patients, with VSD, PDA, and ASD among the most common associated cardiac disorders. [4]

An integral component of the preoperative assessment of patients is airway assessment, especially in patients with congenital heart disorders and syndromic cases. In the operating room, anesthesiologists are likely to encounter airway-related challenges, which can be overcome with skill and experience. Diagnosing syndromes that affect the airway is helpful; nevertheless, some anomalies might be encountered by chance. [7] In patients with syndromic disorders or congenital heart diseases, it is essential to evaluate the airway accurately and meticulously. Points that lead us to a difficult airway include small jaws, large tongues, the improper position of teeth, the thyromental distance, the neck's length and diameter, the Mallampati score, the mouth

opening rate, and the lower-jaw movement rate. [8]

Adamczyk et al [9] maintained that a reliable and accurate way to examine the airway in cases with possible obstruction was to perform a computed tomography scan of the airway with reconstruction or a virtual bronchoscopy.

The crucial point in patients with Pierre Robin syndrome is to maintain spontaneous breathing with brief sedation until intubation and to ensure the location of the tube with capnography, followed by the injection of a muscle relaxant. It is recommended to ensure the amount and adequacy of ventilation with a mask at least once before injecting the relaxant. A laryngoscopy should be carried out to ensure the feasibility of intubation before the relaxing drug is injected. [10]

CONCLUSIONS

In patients with unexpected airway problems undergoing elective surgery, the operation should be postponed. Still, oxygenation should be continued in the operating room until the patient is fully awake. A thorough evaluation of the patient's airway is, then, necessary before scheduling the patient for surgery with appropriate airway management.

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REFERENCES

1. Ziyaeifard M, Azarfarin R, Ferasatkish R, Dashti M. Management of difficult airway with laryngeal mask in a child with mucopolysaccharidosis and mitral regurgitation: a case report. Res Cardiovasc Med. 2014; 3(2):e17456.
2. Baxter D, Shanks AL. Pierre Robin Syndrome. [Updated 2022 May 1]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK562213/
3. Giudice A, Barone S, Belhous K, Morice A, Soupre V, Bennardo F, Boddaert N, Vazquez MP, Abadie V, Picard A. Pierre Robin sequence: A comprehensive narrative review of the literature over time. J Stomatol Oral Maxillofac Surg. 2018 Nov; 119(5):419-428.
4. Pearl W. Congenital heart disease in the Pierre Robin syndrome. Pediatr Cardiol. 1982; 2(4):307-9
5. Krishna SG, Bryant JF, Tobias JD. Management of the Difficult Airway in the Pediatric Patient. J Pediatr Intensive Care. 2018 Sep; 7(3):115-125
6. Diana Raj, MB ChB MRCP FRCA, Igor Luginbuehl, MD FMH, Managing the difficult airway in the syndromic child, Continuing Education in Anaesthesia Critical Care & Pain, Volume 15, Issue 1, February 2015, Pages 7–13,
7. Abduljawad EM, AlHarthi A, AlMatrafi SA, Hussain M, Shawli A, Waggass R. The Prevalence of Congenital Heart Diseases in Syndromic Children at King Khalid National Guard Hospital from 2005 to 2016. Cureus. 2020; 12(4):e7891.
8. Krishna SG, Bryant JF, Tobias JD. Management of the Difficult Airway in the Pediatric Patient. J Pediatr Intensive Care. 2018; 7(3):115-125
9. Adamczyk M, Tomaszewski G, Naumczyk P, Kluczewska E, Walecki J. Usefulness of computed tomography virtual bronchoscopy in the evaluation of bronchi divisions. Pol J Radiol. 2013; 78(1):30-41
10. Rizk MS, Zeineldine SM, El-Khatib MF, Yazbeck-Karam VG, Ayoub SD, Bou-Khalil PK, et al. Relaxantes musculares não despolarizantes melhoram a visibilidade em laringoscopia direta sem efeito sobre a ventilação via máscara facial [Nondepolarizing muscle relaxant improves direct laryngoscopy view with no effect on face mask ventilation]. Rev Bras Anestesiol. 2017 Jul-Aug; 67(4):383-387