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Case Report Open Access

Airway Management in Neonates with Pierre Robin Syndrome along with Mobius Syndrome and Platoglossal Fusion

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Abstract

Pierre Robin Syndrome is a rare congenital syndrome presenting challenges of airway management not only during anesthesia, but also in the Neonatal Intensive Care Unit (NICU). We present a case of Pierre Robin Syndrome which was unusual because it had Mobius Syndrome and palatoglossal fusion as comorbidies, further complicating airway management. We successfully intubated the patient using a fiberoptic laryngoscope via the nasal route.

Keywords: Pierre robin syndrome; Mobius syndrome; Palatoglossal fusion; Fiberoptic intubation

Introduction

Pierre Robin Syndrome (PRS) is a congenital syndrome with multiple anomalies, including micrognathia, receding and short mandible, and cleft lip and/or palate [1-5]. The major challenge for the anesthesiologist in these patients is airway management due to glosssoptosis causing obstructive sleep apnea (OSA). This airway obstruction can exist at multiple levels and result in loss airway patency and death [2,4,6,7]. Micrognathia and cleft palate add further complications to airway management, both in mask ventilation and in intubation [1,3] and limited oral opening can further intensify the challenges of airway management.

Moebius syndrome, also known as congenital oculo-facial paralysis is a very rare neurological disorder with varying severity, typically characterized by 6th and 7th cranial nerve palsies leading to congenital facial weakness and abnormal ocular abduction. These patients require somewhat specialized anesthetic care due to their abnormal airway kinetics, frequently associated congenital cardiac anomalies and positioning issues [8-11].

We present here the case of a 2 week old child, who has PRS, Moebius syndrome and palatoglossal fusion, and we will discuss our efforts to secure his airway in the face of these congenital problems.

Case Report

The patient is a male born two weeks earlier by C-Section in an outlying hospital at 38 weeks gestation. At that time he weighed 2.42 Kg, and he was referred to our institution for further management of his breathing and feeding problems. Antenatal history was essentially unremarkable except for family history of cerebral palsy and prematurity in a sibling. Physical exam by the otolaryngologist demonstrated palatoglossal fusion in the midline with possible underlying bilateral cleft palate, retrognathia and microcanthia with a patent right nasal cavity, but narrowed left nasal cavity (Figure 1) [12-14]. An echocardiogram showed him to have a small patent ductus arterioles (PDA) with a small right to left shunt and bilateral talipes equinovarus. Cranial nerve evaluation suggested Moebius syndrome with sixth and seventh cranial nerve palsies. Upon admission the patient was stable with no attacks of apnea and no periods of desaturation, and routine blood work, chest X-ray and blood gasses were all within normal limits. He was scheduled for surgery to release the palatoglossal fusion to facilitate both airway patency and feeding. Following a preanesthetic evaluation, the patient was assigned an ASA physical status III due to his multiple anomalies. The child already had a PIC line in his right arm, and was receiving formula via a nasogastric tube [15-17].

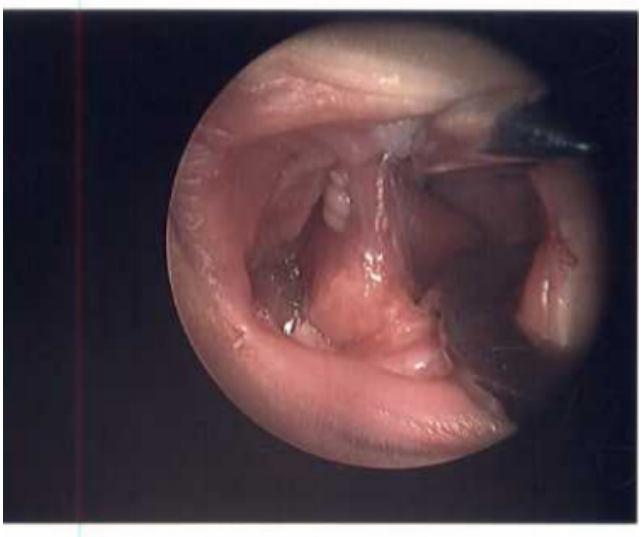

Figure 1: Platoglossal fusion in the midline.

Feeding was withheld for 6 h previous to surgery, but replacement fluids were provided via the PIC line. The operating room was

prepared with an unusually wide assortment of airway management devices including, in addition to routine devices, a pediatric fiberoptic bronchoscope, a rigid bronchoscope, a crycothyroidotomy set, a tracheostomy set and an otolaryngologist was present [18]. No premedication was administered, but after applying the appropriate monitors, the patient was preoxygenated and given Glycopyrrolate 5 mg to reduce secretions and minimize bradycardia. The patient was given Sevoflurane and oxygen via facemask, starting at 2% Sevoflurane, and gradually increasing to 5% Sevoflurane. Once the patient had reached a deep plane of anesthesia, a 2.8 MM pediatric fiberoptic laryngoscope was passed into the patient's nose and to the vocal cords, but it would not fit into the narrow laryngeal inlet. During this time several episodes of desaturation occurred, but responded well to assisted face mask ventilation with little difficulty [19,20]. After two failed attempts, a 2.5 mm pediatric fiberoptic laryngoscope was passed through one nostril while a 2.5 mm endotracheal tube was passed through the other nostril and passed into the trachea by focusing on respiratory sounds while manipulating the larynx externally. Passage of the tube into the trachea was observed using the 2.5 mm fiberoptic laryngoscope. Proper placement was further confirmed by waveform capnography, bilaterally equal chest movement, equality of air entry and negative gastric auscultation (Figures 2 and 3).

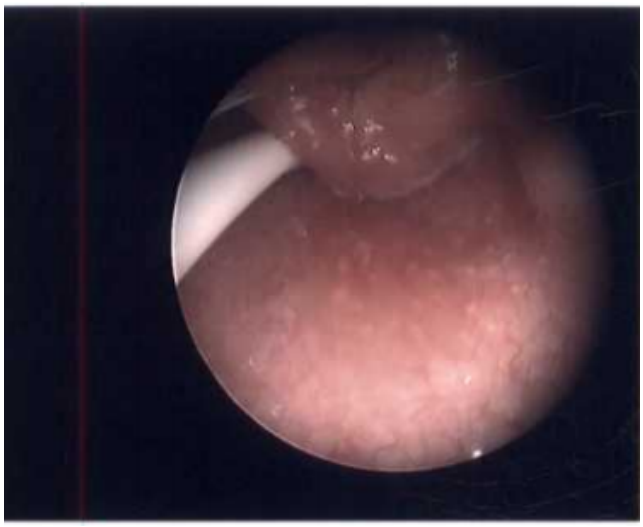

Figure 2: Fibreoptic views before nasal intubation through the right patent nostril (in the view the nasogastric tube).

Once endotracheal tube placement was confirmed, the patient was given fentanyl 10 mcg. Anesthesia was maintained with Sevoflurane 2-3% and the inspired oxygen concentration was reduced to 50% by the addition of nitrous oxide. Mechanical ventilation was initiated in PVC mode at a rate of 25-30 respirations/minute, and I:E ratio of 1:2, and peak pressures less than 25 cm H_2O.

The surgical release of the palatoglossal adhesion was uneventful, but the cleft palate and lip were left for repair at an older age. The patient was returned to the NICU in stable condition and remained on mechanical ventilation for the first 24 h due to the possibility of edema of the tongue, a risk for which he was receiving steroids. He was extubated on the second post-operative day in the prone position without any respiratory difficulties or complications, and he was discharged home on the fifth post-operative day.

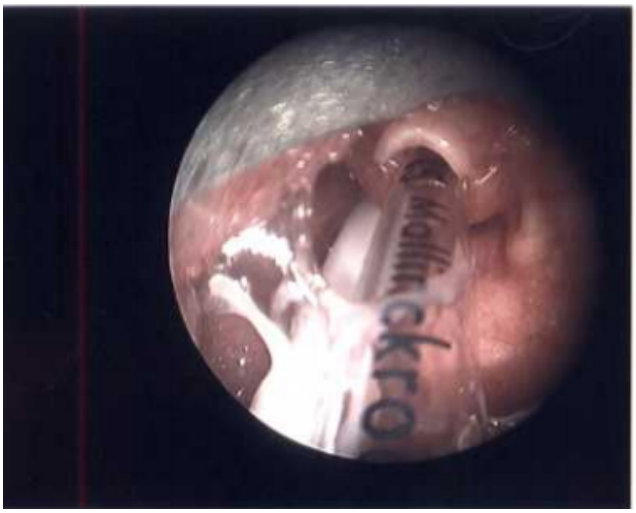

Figure 3: Manipulation of the endotracheal tube into laryngeal inlet after external laryngeal manipulation using breathing as a guide.

Discussion

Pierre Robin Syndrome usually presents with three cardinal features - retrognathia (backward placed or receding mandible), cleft palate, and retroglossoptosis (an abnormally large tongue usually causing airway obstruction and is the hallmark of this syndrome [1-4]. The presence of this large and superiorly positioned tongue prevents the normal closure of the palatal shelves. In our patient, the tongue was fused with the palatal shelves on both sides, removing oral intubation from a reasonable list of options for this patient's intubation. Interestingly, the fusion of the tongue to the palatine tissue prevented the tongue from falling into the hypo pharynx when the patient was placed in the supine position for nasal intubation.

Apart from feeding difficulties, airway obstruction and respiratory distress are the primary issues in patients with PRS and palatoglossal fusion. Panendoscopy is invaluable in determining the site of airway obstruction, and patients should also be evaluated by turning them in different positions to see how gravity can ameliorate of exacerbate their airway obstruction. Prone position can relieve airway obstruction in as many as 70% of these patients, contrary to the American Academy of Pediatrics "Back to Sleep" program to minimize sudden infant death syndrome [21,22]. As much as 60% of patients with PRS may have associated congenital anomalies such as Stickler, Velocardiofacial and Treacher-Collins syndromes. If these syndromes are suspected, especially if a murmur is present, a preoperative echocardiogram should be performed [2,5].

Ventilation can be difficult in patients with PRS because of the retrognathia and retroglossoptosis. To relieve this obstruction, two handed jaw thrust along with one of more airway adjuncts may be required. In the unfortunate event of inability to ventilate after induction of anesthesia, emergent bronchoscopy with rigid bronchoscope may be the only option to ventilate a patient if direct laryngoscopy and intubation failed [2,3,5]. Emergency tracheostomy and extracorporeal membrane oxygenation (in patients >2 kg and older than 34 weeks gestational age) can be the last resorts in a pediatric "cannot ventilate, cannot intubate" situation [2,3,5,7].

While the hallmark of managing the pediatric difficult airway in PRS is maintaining spontaneous ventilation, many devices can be used to assist in this management including the fiberoptic bronchoscope, a retrograde wire, the Glidescope, Sikhani scope, Air-Q scope, Airtraq, Story C Mac video laryngoscope and the laryngeal mask airway (LMA), among others.

In a case series of 5 neonates with PRS requiring general anesthesia, they report LMA insertion in all five cases without any anesthesia and report that all the neonates became calm once the LMA was passed beyond the obstruction. A related procedure is to place an LMA first, then pass a fiberscope through the LMA, after which an endotracheal tube, which had been previously place over the fiberscope, is passed over the fiberscope into the trachea.

Although we used Sevoflurane as our main anesthetic agent, Isoflurane could have also been used as well, although desflurane , despite its ultra-short time of action, would not have been a good option due to its propensity to increase airway reactivity, secretions and laryngospasm [2,21]. Ultra-short acting opioids like Remifentanil or alpha two agonists like Dexmedetomidine or low dose ketamine could also be used to provide post-operative analgesia while avoiding the respiratory depression of a long-acting opioid [19,20].

Postoperative airway obstruction can be caused by edema due to airway manipulation, surgical manipulation of the tissue or muscular hypotonia, and can result in hypoxia and negative pressure pulmonary edema. This can sometimes be prevented by use of the prone position, specific airway adjuncts, and/or tying the tongue to the chin. Occasionally a tracheostomy may be necessary [2,22].

Moebius syndrome is characterized by congenital palsy of the 6th and 7th cranial nerves, but may also be associated with palsies of other cranial nerves, including the 9th and 10th, creating dysfunction of the pharynx, dysphagia, feeding difficulties, retention of oral secretions and recurrent bouts of aspiration pneumonia [8-11,25], and can also be associated with central alveolar hypoventilation. Postoperative use of opioid, sedative and anesthetic agents in th presence of central hypoventilation can predispose to grave respiratory consequences. In addition, postoperative pain assessment is difficult due to absence of facial expression because of the facial palsy [9,10,25,26].

Conclusion

The coexistence of PRS, Moebius syndrome and palatoglossal fusion is extremely rare, and poses multiple anesthetic challenges. Maintaining spontaneous ventilation and avoiding opioids and muscle relaxants is crucial to maintaining a patent airway until the airway is secured. Meticulous preparation and team coordination is required in the management of such a patient if a good outcome is to be expected.

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References

1. Breugem C, Courtemanche D (2010) Robin Sequence: Clearing Nosologic Confusion. Cleft Palate Craniofac J 47: 197-200.
2. Cladis F, Kumar A, Grunwaldt L, Otteson T, Ford M (2014) Pierre Robin Sequence: A Perioperative Review. Anesthesia and Analgesia.; 119: 400-412.
3. Bissonnette B, Luginbuehl I, Marciniak B, Dalens BJ (2006) Rapid recognition and Perioperative Implication; Pierre Robin Syndrome; McGraw-Hill Education.
4. Richard E, Kirschner AK (2009) Pierre Robin Sequence. 1st ed New York, NY McGraw Hill.
5. Kaufman MG, Cassady CI, Hyman CH, Lee W, Watcha M, et al. (2016) Prenatal Identification of Pierre Robin Sequence: A Review of the Literature and Look towards the Future. Fetal Diagn Ther 39: 81-89.
6. Mukhopadhyay P, Mukherjee P, Adhikari M (1992) Problems in Anesthetic Management of Pierre Robin and Treacher Collin Syndromes. Indian Pediatr J 29: 1170-1172.
7. Marston AP, Lander TA, Tibesar RJ, Sidman JD (2012) Airway management for intubation in newborns with Pierre Robin Sequence. Laryngoscope 122: 1401-1404.
8. Verzijl HT, Van Der Zwaag B, Cruysberg JR, Padberg GW (2003) Möbius syndrome redefined: a syndrome of rhombencephalic maldevelopment. Neurology 61: 327-333.
9. Gondipalli P, Tobias JD (2006) Anesthetic implications of Moebius syndrome. J Clin Anesth 18: 55-59.
10. Matsui K, Kataoka A, Yamamoto A, Tanoue K, Kurosawa K, et al. (2014) Clinical characteristics and outcomes of Möbius syndromein a Children'sHospital. Pediatr Neurol 51: 781-789.
11. Budic I, Surdilovic D, Slavkovic A, Marjanovic V, Stevic M, et al. (2016) Moebius Syndrome: Challenges of Airway Management. Acta Clin Croat 55: 94-97.
12. Khan TH, Ghayas A, Ayesha, Bakht SK, Khan N (2013) CASE REPORT-Successful intubation with air-Q in Pierre Robin Syndrome. Anesthesia Pain and Intensive care. 17:71-74.
13. Stricker P, Budac S, Fiadjoe J, Rehman MA (2008) Awake laryngeal mask insertion followed by induction of anesthesia in infants with the Pierre Robin sequence. Acta Anaesthesiol Scand 52: 1307-1308.
14. Vlatten A, Aucoin S, Gray A, Soder C (2009) Difficult airway management with the STORZ video laryngoscope in a child with Robin Sequence. Paediatr Anaesth 19: 700-701.
15. Cladis AA, Kumar A, Grunwaldt L, Otteson T, Ford M (2014) Robin Sequence: A Perioperative Review Anesth Analg 119:400-412.
16. Cohen SR, Simms C, Burstein FD, Thomsen J (1999)Alternatives to tracheostomy in infants and children with obstructive sleep apnea. J Pediatr Surg 34: 182-186.
17. Templeton TW, Bryan YF (2016) A two-stage approach to induction and intubation of two infants with Pierre Robin Sequence using a LMA ClassicTM and Air-Q®: two cases report. Korean J Anesthesiol 69: 390-394.
18. Asai T, Nagata A, Shingu K (2008) Awake tracheal intubation through the laryngeal mask airway in neonates with upper airway obstruction. Pediatr Anesth 18: 77-80.
19. Brown KA (2011) Outcome, risk, and error and the child with obstructive sleep apnea. Paediatr Anaesth 21:771-778.
20. Dsouza S, Barad D, Bharne S (2012) Intubation using dexmedetomedine and ketamine in a child with Pierre Robin sequence. J Anaesthesiol Clin Pharmacol 28: 545-546.
21. Von Ungern-Sternberg BS, Saudan S, Petak F, Hantos Z, Habre W (2008) Desflurane but not sevoflurane impairs airway and respiratory tissue mechanics in children with susceptible airways. Anesthesiology 108: 216-224.
22. Mackay DR (2011) Controversies in the diagnosis and management of the Robin sequence. J Craniofac Surg 22: 415-420.
23. Jarrahy R (2012) Controversies in the management of neonatal micrognathia: to distract or not to distract, that is the question. J Craniofac Surg 23: 243-249.

24. Brown KA, Laferrière A, Moss IR (2004) Recurrent hypoxemia in young children with obstructive sleep apnea is associated with reduced opioid requirement for analgesia. Anesthesiology 100: 806-10.

25. Ferguson S (1996) Moebius syndrome: a review of the anaesthetic implications. Paediatr Anaesth 6: 51-56.

26. Igarashi M, Rose DF, Storgion SA (1997) Moebius syndrome and central respiratory dysfunction. Pediatr Neurol 16: 237-240.