KANSAS JOURNAL of MEDICINE

Case Report

Airway Management in a Pediatric Patient Presenting with Pierre-Robin Sequence

Logan Foster, D.O.[1], Declan Foster[2], Harsh Nathani, M.D.[1,3], William Krogman, M.S.[1]
[1]University of Kansas School of Medicine-Wichita, Wichita, KS
Department of Anesthesiology
[2]Burrell College of Osteopathic Medicine-Las Cruces, NM
Osteopathic Medical Student
[3]Wesley Medical Center, Wichita, KS
Wichita Anesthesiology Chartered

Received Dec. 11, 2023; Accepted for publication April 1, 2024; Published online April 26, 2024
https://doi.org/10.17161/kjm.vol17.21529

INTRODUCTION

Pierre-Robin sequence (PRS) was first described in 1923 by Pierre Robin, who noted a combination of three clinical signs often occurring together: micrognathia, glossoptosis, and partial upper airway obstruction.[1] Subsequently, the association of cleft palate with this presentation became part of the diagnostic criteria,[2] resulting in the current understanding of PRS, which has an incidence of approximately 1 in 8,500 births.[3] The exact cause of PRS is not yet fully understood, but it is believed to result from a combination of genetic factors.[2,4] One challenge in identifying the exact cause is that PRS can occur either as an isolated abnormality or in conjunction with other genetic malformations, known as syndromic PRS.[5] Both forms of PRS can complicate airway management, especially during endotracheal intubation in pediatric patients. This is due to the retrognathic mandible characteristic of PRS, which causes the tongue to be positioned anteriorly, leading to glossoptosis and potential airway obstruction, making feeding difficult. Additionally, during anesthesia induction, relaxation of the upper airway tissues can exacerbate the obstruction caused by the tongue.[6]

This report discusses the challenges encountered in a pediatric patient undergoing surgical correction of micrognathia through mandibular osteotomy and distraction. The approach involved using an awake laryngeal mask airway (LMA) placement in the lateral position, combined with fiberoptic visualization for endotracheal tube placement with the LMA. This technique offers a novel and effective method of managing the airway, particularly in cases with severe micrognathia and glossoptosis associated with PRS. Prior to presenting this report, written informed consent was obtained from the patient's legal guardian.

CASE REPORT

A two-month-old neonate underwent a planned direct laryngoscopy, rigid bronchoscopy, and mandibular osteotomy with distraction. The patient's mother had an uneventful pregnancy and delivery. Preoperatively, the patient did not show signs of respiratory distress but was found to have significant airway obstruction due to micrognathia and glossoptosis while lying flat. Airway examination revealed severe micrognathia and glossoptosis, making it difficult to see the back of the mouth. The patient was given a peripheral intravenous access in the left forearm and was taken to the operating room while awake.

During transport, the patient's heart rate was 170 bpm, blood pressure was 90/50 mmHg, and oxygen saturation was 94% on 12 L of 64% oxygen via mask. To prevent airway obstruction, the patient was positioned on their side. Intravenous atropine 0.05 mg was given to reduce the parasympathetic response and dry secretions. Topical lidocaine 2% jelly was applied to numb the mouth, and nasal decongestant oxymetazoline 0.05% was applied to the nostrils to help with nasotracheal tube placement.

With the tongue pulled forward, an LMA was placed while the patient was still on their side. Once the LMA was in the correct position, the patient was placed on their back. Sevoflurane was used for inhalation induction via the LMA. After achieving adequate anesthesia, a rigid bronchoscopy was performed, but an endotracheal tube could not be passed orally afterward. The LMA was replaced, and a fiberoptic scope with a 3.0 mm cuffed endotracheal tube was inserted through the left nostril, and the vocal cords were visualized after retracting the LMA. The endotracheal tube was then passed through the vocal cords under fiberoptic guidance. The LMA was removed, and ventilation continued through the endotracheal tube. Tube placement was confirmed, and fentanyl was administered. The patient continued to breathe spontaneously while on the ventilator with sevoflurane for maintenance. Dexamethasone was given to prevent post-operative airway swelling. The mandibular osteotomy and distraction were completed successfully, and the patient was transferred to the neonatal intensive care unit with the nasotracheal tube in place. The patient was successfully extubated on post-operative day five without complications.

A second surgical procedure performed 30 days later showed significant improvement in the patient's micrognathia, and a Grade 1 view was achieved using a GlideScope during subsequent endotracheal intubation.

DISCUSSION

Pierre-Robin sequence presents significant challenges in managing the airway, especially in pediatric patients. The pediatric airway anatomy, characterized by a large occiput, short neck, large tongue, and short and narrow hypopharynx, makes it challenging to align the oral and tracheal axes and to place supraglottic airways.[7]

Various techniques have been described for managing PRS patients. Historically, direct laryngoscopy was used, with fiberoptic endotracheal intubation as a backup. A study from 2012 found a 63% failure rate with direct laryngoscopy, necessitating endotracheal intubation with a fiberoptic bronchoscope.[5] The advent of video laryngoscopy has improved airway management, but best practices for PRS patients remain unclear.

A 2023 study compared time to intubate and time to ventilate on a Pierre-Robin Manikin using different types of laryngoscopes including a GlideScope LoPro blade, a straight GlideScope blade, a Spectrum Miller blade, a conventional Macintosh blade, and a conventional Miller blade. The Spectrum Miller blade showed the highest intubation success rate, though no significant differences were demonstrated in time to intubate and time to ventilate among blades.[8] A 2022 case report

advocates for the use of hyperextension and cricoid pressure in combination with video laryngoscopy as a means of improving vocal cord visualization; however, this was performed in a four-year-old patient and not in a neonate.[9] Another recent case report even described the use of a video ureteroscope for endoscopic airway visualization to facilitate the placement of an oral endotracheal tube in a patient undergoing mandibular distraction.[10]

The use of LMA as an adjunct for intubation is gaining popularity. Studies have shown a higher success rate of intubation on the first attempt with LMA techniques.[7,11,12] Awake fiberoptic intubation through an LMA has been successful in some cases. A staged approach using an awake LMA followed by placement of an air-Q® and fiberoptic intubation through the air-Q® has also been successful.[13] In our case, the placement of an awake LMA in the lateral decubitus position followed by nasotracheal exchange under fiberoptic visualization minimized airway obstruction and facilitated quick induction, reducing trauma to the pediatric airway. This underscores the importance of early comprehensive airway management planning and a multidisciplinary approach in PRS cases. While no consensus exists on the best practice for intubating PRS patients, the approach used in this case was effective.

REFERENCES

1 Robin P. A fall of the base of the tongue considered as a new cause of nasopharyngeal respiratory impairment: Pierre Robin sequence, a translation. 1923. Plast Reconstr Surg 1994; 93(6):1301-1303. PMID: 8171154.
2 Giudice A, Barone S, Belhous K, et al. Pierre Robin sequence: A comprehensive narrative review of the literature over time. J Stomatol Oral Maxillofac Surg 2018; 119(5):419-428. PMID: 29777780.
3 Bush PG, Williams AJ. Incidence of the Robin Anomalad (Pierre Robin syndrome). Br J Plast Surg 1983; 36(4):434-437. PMID: 6626822.
4 Carroll DB, Peterson RA, Worton EW, Birnbaum LM. Hereditary factors in the Pierre Robin syndrome. Br J Plast Surg 1971; 24(1):43-47. PMID: 5547958.
5 Marston AP, Lander TA, Tibesar RJ, Sidman JD. Airway management for intubation in newborns with Pierre Robin sequence. Laryngoscope 2012; 122(6):1401-1404. PMID: 22460229.
6 Gangopadhyay N, Mendonca DA, Woo AS. Pierre Robin sequence. Semin Plast Surg 2012; 26(2):76-82. PMID: 23633934.
7 Harless J, Ramaiah R, Bhananker SM. Pediatric airway management. Int J Crit Illn Inj Sci 2014; 4(1):65-70. PMID: 24741500.
8 Irouschek A, Moritz A, Kremer S, et al. An approach to difficult airway in infants: Comparison of GlideScope® Spectrum LoPro, GlideScope® Spectrum Miller and conventional Macintosh and Miller blades in a simulated Pierre Robin sequence performed by 90 anesthesiologists. PLoS One 2023; 18(8):e0288816. PMID: 37535590.
9 Mehrpisheh S, Farhadi R. Effective intubation technique to manage the airway obstruction in a case of Pierre Robin sequence: A case report. Clin Case Rep 2022; 10(3):e05547. PMID: 35280079.
10 Bhardwaj A, Grewal R, Trivedi S, Singh S. A novel approach to airway management in Pierre Robin syndrome-a case report. Oxf Med Case Reports 2022; 2022(12):omac132. PMID: 36579079.
11 O'Neill B, Templeton JJ, Caramico L, Schreiner MS. The laryngeal mask airway in pediatric patients: Factors affecting ease of use during insertion and emergence. Anesth Analg 1994; 78(4):659-662. PMID: 8135383.
12 Asai T, Nagata A, Shingu K. Awake tracheal intubation through the laryngeal mask in neonates with upper airway obstruction. Paediatr Anaesth 2008; 18(1):77-80. PMID: 18095971.
13 Templeton TW, Goenaga-Díaz EJ, Runyan CM, Kiell EP, Lee AJ, Templeton LB. A generalized multistage approach to oral and nasal intubation in infants with Pierre Robin sequence: A retrospective review. Paediatr Anaesth 2018; 28(11):1029-1034. PMID: 30284747.

Keywords: case report, pediatrics, Pierre Robin syndrome, laryngeal mask, mandibular osteotomy