Navigating Airway Obstacles: Effective Anesthesia Strategies for Severe Robinson Sequence in a 3 year old

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SUMMARY

Pierre Robin Sequence (PRS) is a congenital condition characterized by micrognathia, glossoptosis, and airway obstruction, often accompanied by a cleft palate. Severe cases pose additional challenges in surgical management due to anatomical abnormalities. A 3-year-old male with severe PRS underwent a cleft palate repair procedure, which was a multidisciplinary approach, and the successful repair of the cleft palate and uneventful postoperative recovery were attributed to the team's collaborative efforts. This case also highlights the importance of genetic evaluation and testing in managing PRS cases. This case highlights the importance of thorough preoperative assessment, advanced airway management skills, and genetic evaluation for better patient outcomes. Future research should explore innovative techniques and strategies to improve the outcomes of patients with PRS and other complex congenital conditions.

KEYWORDS:
FESS, Budesonide, Sinusitis, Steroid nasal spray, Nasal Polyp

INTRODUCTION

Pierre Robin Sequence (PRS) is a congenital condition characterized by a triad of anomalies [1]: micrognathia, glossoptosis, and airway obstruction, often accompanied by a cleft palate. This condition presents significant challenges in neonatal care, particularly in terms of maintaining a patent airway and ensuring adequate nutrition.[2] Severe cases of PRS, such as the one described in this report, pose additional difficulties in surgical management due to anatomical abnormalities.[3] Here, we present the case of a 3-year-old male with severe PRS, focusing on the clinical findings, the challenges encountered during surgical attempts to repair the cleft palate, and the multidisciplinary approach adopted to ensure a successful outcome. This case highlights the importance of thorough preoperative[1] planning and advanced airway management techniques to achieve favorable surgical results in patients with severe PRS.[4]

This report details the clinical findings, diagnostic and therapeutic challenges encountered, and coordinated efforts of a multidisciplinary team in managing his condition. This case highlights the critical role of interdisciplinary collaboration[5] and advanced airway management techniques in achieving favorable surgical outcomes in patients with severe PRS.[6]

CASE REPORT

The patient was a 3-year-old male weighing 10.5 kg with a medical history of severe Pierre Robin Sequence. Clinically, the patient exhibited a notable gap in the roof of his mouth since birth, indicative of cleft palate, and was scheduled for cleft palate repair surgery.

The antenatal history was uneventful, boen via expected vaginal delivery, with a birth weight of 2.3 kgs. Immediately after birth, the patient developed breathing difficulties, feeding difficulties, and nasal regurgitation, and was admitted to the NICU and treated conservatively.

At 1 year of age, patients with features such as micrognathia, microcephaly, mandibular hypoplasia, cleft palate, and no cleft lip were posted for elective cleft palate repair. After routine monitoring and preoperative assessment, the patient was shifted inside the OT, monitored were connected, and after adequate preoxygenation, trail laryngoscopy failed, video laryngoscopy-assisted intubation attempt failed followed by FOB-guided intubation was attempted, was failed due to severe mandibular hypoplasia, micrognathia, and complex airway. The patient was categorized as can ventilate, cannot intubate, and was shifted to the ICU for further management.

At 3 years of age, the patient was further referred for elective cleft palate repairsurgery. An appropriate preoperative assessment was performed, and patient under the difficult airway category was scheduled for the procedure.

Investigations revealed a hemoglobin level of 9.3 g/dL, bleeding time of 2 min, and clotting time of 9 min 10 seconds. The total leukocyte count was 16,970/mm³, and the platelet count was 430,000/µL. Renal and liver function tests were routinely performed. Serological results were negative for infectious diseases. Both the echocardiogram and chest radiograph were within normal limits.

Preoperative assessment revealed anticipated difficulties in ventilation and intubation. Anesthetic management included a thorough preoperative evaluation and preparation of the predicted difficult airway. To ensure comprehensive planning and coordination, discussions were conducted with a multidisciplinary team, including anesthesiologists, pediatric surgeons, and otolaryngologists.

Premedication was administered based on weight-based calculations, and an intravenous (IV) line was secured with a

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22G cannula. The medications given included Inj. Glycopyrrolate reduces secretion; Inj. Midazolam was used for anxiolysis, Inj. Fentanyl for analgesia, and Inj. Propofol for induction.

During induction, after ensuring that the patient was adequately ventilated. Succinylcholine was administered to facilitate muscle relaxation because it is a depolarizing muscle relaxant known for its rapid onset and short duration of action, making it a popular choice over other non-depolarizing muscle relaxants. The initial attempt at intubation using standard laryngoscopy failed to visualize the vocal cords because of severe micrognathia and glossoptosis. Consequently, video laryngoscopy (Mc GRATH MAC) was utilized to visualize the airway structures better, and a style-guided RAE tube was successfully inserted.

Intraoperative management included continuous monitoring of vital signs, such as heart rate, blood pressure, oxygen saturation, and end-tidal CO2. Anesthesia was maintained with inhalational agents, specifically sevoflurane, and intravenous medications, as needed.

Postoperative care involved extubation in the operating room after confirmation of adequate spontaneous breathing and muscle strength. The patient was then transferred to the ICU for close monitoring, the surgery was completed without complications, and postoperative recovery was uneventful. The patient demonstrated stable respiratory function, with no signs of airway obstruction. He was monitored in the ICU for 24 hours before being moved to the pediatric ward. He was discharged after adequate pain control and feeding assessment.

DISCUSSION

This case of a 3-year-old male with severe Pierre Robin Sequence (PRS) highlights several critical aspects of managing complex congenital conditions. PRS, which is characterized by micrognathia, glossoptosis, and airway obstruction, often poses significant medical and surgical management challenges. In this patient, the presence of additional anomalies, such as microcephaly and mandibular hypoplasia, further complicated the clinical picture.

The initial surgical attempt to repair the cleft palate at age 2 years was unsuccessful because of the patient's complex airway anatomy. This outcome highlights the importance of thorough preoperative planning and potential complications in patients with severe PRS. The failure of standard laryngoscopy and subsequent successful use of video laryngoscopy for intubation during the second surgical attempt illustrate the necessity for advanced airway management techniques in such cases. These findings align with previous reports emphasizing the need for specialized equipment and skilled personnel to manage airway challenges in patients with PRS.

The multidisciplinary approach adopted in this case, involving anesthesiologists, pediatric surgeons, and otolaryngologists, was crucial for achieving a favorable outcome. This collaborative strategy ensured comprehensive care and addressed the multifaceted needs of the patients. Successful repair of the cleft palate and uneventful postoperative recovery highlight the effectiveness of this integrated approach. The literature supports the involvement of multidisciplinary teams in improving surgical outcomes and the overall management of patients with PRS and similar congenital anomalies.

Moreover, this case helps us to understand the importance of anticipating and preparing for difficult ventilation and intubation scenarios. The use of preoperative assessments and advanced planning, as demonstrated herein, can significantly mitigate the risks associated with airway management in patients with PRS. Studies have consistently shown that detailed preoperative evaluations and availability of advanced airway management tools are critical in reducing perioperative complications .

Case of Neonate with Severe Upper Airway Obstruction:
A 9-day-old male infant with PRS, weighing 3.5 kg, presented with severe upper airway obstruction due to a combination of cleft palate and a vallecular cyst. Multiple attempts at intubation using direct laryngoscopy, GlideScope, LMA, fiberoptic scope, and lighted wand were unsuccessful. Successful airway management was eventually achieved through digital intubation, highlighting the importance of a broad skill set in airway management techniques for such complex cases.[7]

1p36 Deletion Syndrome and PRS:
In a neonate diagnosed with 1p36 deletion syndrome complicated by PRS, severe airway obstruction was successfully managed using fiberoptic nasotracheal intubation immediately after birth. This case emphasizes the importance of preparedness with fiberoptic equipment and neonatologists' acquisition of relevant skills to manage unexpected airway compromises effectively.[8]

Stickler Syndrome with PRS:
A case involving A patient with Stickler syndrome type 1, identified through a novel COL2A1 gene mutation, presented with PRS. This case highlights that Stickler syndrome accounts for a significant proportion of PRS cases, stressing the need for genetic testing and family history evaluation in patients with PRS, mainly when other symptoms such as myopia or retinal detachment are present 7

Advanced Airway Management Techniques:
In a tertiary pediatric hospital, a case of PRS with unique anatomical challenges required old-fashioned digital intubation after multiple high-tech methods failed. This demonstrates the need for training in various airway management techniques, including traditional methods, to effectively handle challenging airway scenarios effectively.[9]
Severe Glossoptosis in PRS:

Another report described a severe case of glossoptosis in PRS, where the tongue was invaginated into the nasal cavity, necessitating immediate fiberoptic intubation for resuscitation. This case highlights the need for neonatologists to be adept at fiberoptic intubation to manage similar critical situations.[9]

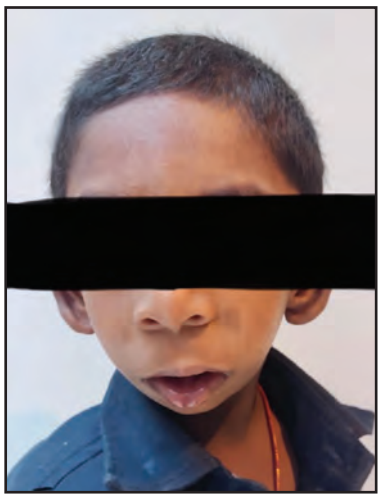

Fig. 1: Dysmorphic facial features

Fig. 2: Post intubation

Mandibular Distraction Osteogenesis (MDO) in PRS:
Several cases have reported the use of MDO as an effective intervention for severe airway obstruction in patients with PRS. The MDO gradually elongates the mandible, reduces glossoptosis, and improves airway patency. This surgical technique has shown promising results in the alleviation of airway obstruction in infants with severe PRS.[9]

These cases collectively illustrate the complex and varied nature of PRS management, prioritizing the importance of a multidisciplinary approach, advanced airway management skills, and the potential role of genetic evaluation in improving patient outcomes.

The successful management of this case resulted from meticulous preoperative planning, advanced airway management techniques, and coordinated efforts of a multidisciplinary team.

CONCLUSION

This case of a 3-year-old male with severe Pierre Robin Sequence (PRS) underscores the complexities and challenges associated with managing this congenital condition, particularly regarding airway management and surgical interventions. The initial failure to intubate due to difficult airway anatomy highlights the importance of comprehensive preoperative assessment and availability of advanced airway management techniques. The successful outcome achieved through a multidisciplinary approach and video laryngoscopy for intubation emphasizes the necessity for collaboration and specialized skills in treating patients with PRS.

This case also illustrates the critical role of tailored anesthetic management in achieving favorable surgical outcomes. Detailed preoperative planning and implementation of innovative techniques facilitated successful repair of the cleft palate and uneventful postoperative recovery. These findings align with the existing literature that supports advanced airway management tools and a multidisciplinary team approach for managing PRS.

This case contributes to the growing body of evidence advocating thorough preoperative assessments, advanced airway management training, and multidisciplinary collaboration for managing severe PRS. Future research should continue to explore and refine these approaches to improve the outcomes in patients with this challenging condition.

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Conflict of interest:
The author declare no conflict of interest.

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