

CASE SERIES

ANESTHESIA & CONCURRENT DISEASE

Accidental extubation in a patient with Pierre-Robin syndrome: a case report

Karolina Czajkowska¹, Liuba Germanova², João Bettencourt³

Author affiliations:

1. R. Prof. Mota Pinto, 3004 561, Coimbra Hospital and University Centre, Coimbra, Portugal.
2. R. Prof. Mota Pinto, 3004 561, Coimbra Hospital and University Centre, Coimbra, Portugal; E-mail: liubagermanova28@gmail.com
3. Av. Lu s de Cam es 6180, 9000 177, Hospital Dr. Nélio Mendonç, Funchal. Portugal; E-mail: jbettencourt92@gmail.com

Correspondence: Karolina Czajkowska; Phone: (+351) 926095762; E-mail: karolinamariacz@hotmail.com

Abstract

The Pierre Robin syndrome (PRS) is characterized by a clinical triad of micrognathia, glossoptosis and airway obstruction. These patients frequently require anesthesia for diagnostic and therapeutic procedures and may present a significant challenge to the anesthesiologist in charge. The principal difficulties when approaching these patients are related to the airway management.

We present a case of a 4 years old boy with a PRS, scheduled to undergo elective tympanoplasty. We performed an awake intubation with a fiberscope without complications. At the beginning of the surgery we verified an accidental extubation of the patient and we initiated attempts of manual ventilation, but without success. Due to hypoxemia and severe bradycardia, the patient suffered a cardiac arrest. We initiated advanced life support maneuvers and at this point the manual ventilation with a face mask was established. The patient recovered the sinus rhythm and we managed to intubate him again with a fiberscope. In conclusion, in case of patients with PRS it is essential to plan in advance the airway management approach and make sure that after establishing an intubation, the tube is firmly secured, as an accidental extubation might lead to catastrophic events.

Key words: Pediatric anesthesia; Accidental extubation; Perioperative complications; Pierre Robin syndrome; Perioperative cardiac arrest

Citation: Czajkowska K, Germanova L, João Bettencourt J. Accidental extubation in a patient with Pierre-Robin syndrome: a case report. *Anaesth. pain intensive care* 2022;26(3):416-418. DOI: [10.35975/apic.v26i3.1903](https://doi.org/10.35975/apic.v26i3.1903)

Received: Nov 17, 2019; **Reviewed:** Dec 10, 2019; **Accepted:** Dec 28, 2019

1. Introduction

The Pierre Robin syndrome (PRS) consists of a triad of anomalies characterized by micrognathia, glossoptosis and palatal defect. Patients with the PRS can be very challenging in the perioperative period due to: airway obstruction, difficult manual ventilation and tracheal intubation, common accidental extubation, post-operative pulmonary complications, concurrent presence of other syndromes and congenital diseases.

In pediatric patients, especially in case of patients with the head and neck abnormalities, an accidental extubation can rapidly progress with severe hypoxemia, and if a manual ventilation cannot be not achieved cannot be not achieved promptly, it can lead to a cardiac arrest.

We present a case report of an accidental extubation of a patient with PRS, when the patient suffered a cardiac arrest due to difficulty in establishing a successful ventilation.

2. Case report

A 4-year-old child with a PRS was admitted to our hospital in order to be undergo an elective tympanoplasty. His previous history was positive for an incident of difficult intubation, succeeded only with the use of a fiberscope, when the patient underwent a palatoplasty 2 years back. On the day of the surgery he received premedication according to our protocol (0.2 ml/kg of a mixture of midazolam 30 mg/6 ml + droperidol 2.5 mg/ml + syrup 13 ml). When the patient

arrived in the operating room, we proceeded with an awake nasotracheal intubation with a fiberscope. The correct position of the tube was confirmed by pulmonary auscultation, presence of capnography and indirect visualization with the fiberscope. General anesthesia was induced with propofol and mechanical ventilation was initiated. For the maintenance of anesthesia we used sevoflurane. The nasotracheal tube was secured with a tape. At the beginning of the surgery, while moving the head to the side to facilitate the surgical approach, we observed a sudden capnography loss. We immediately checked the position of the tube and verified that it had accidentally exteriorized and was not inside the trachea any more. We asked to prepare the fiberscope to intubate but it was not immediately available. We attempted to ventilate the patient manually through a face mask, but with no success. The child suffered from a rapid desaturation and bradycardia. While administering atropine to rise the heart rate, asystole was observed on the monitor. Advanced live support maneuvers were initiated. At that moment, the manual ventilation of the patient was possible and we managed to recover the oxygenation. After two cycles of advanced live support maneuvers the sinus rhythm was restored. When the fiberscope was available, we proceeded with the second tracheal intubation. The tube was secured with a tape and with a special band obtained from the Intensive Care Unit (ICU). The surgery was cancelled and the patient was transferred to the ICU. He remained hemodynamically stable and after verifying all the safety conditions, the patient was extubated with no complications. He was discharged the next day.

3. Discussion

Patients with the PRS present with a triad of anomalies characterized by micrognathia, glossoptosis and palatal defect. Clinically the PRS manifests itself with an airway obstruction, difficulty in alimentation and in severe cases, if not treated, it can lead to death still in the neonatal period. These patients frequently need anesthesia care to undergo diagnostic and therapeutic procedures and can present many challenges in the perioperative period. Principal perioperative problems in this population are airway obstruction, difficult tracheal intubation and pulmonary complications that may occur.¹ Anesthesiologist responsible for a patient with PRS should have an experience with a difficult airway management, be familiar with current life support algorithms and plan how to approach these patients in advance.²

The occurrence of accidental extubation, though well studied in the intensive care area, is still not well investigated in the context of perioperative care. It is a rare event (< 0.3%),³ still it can have catastrophic consequences anytime in one's clinical life. In patients

with craniofacial anomalies, due to anatomic differences, the risk of an accidental extubation may be higher than in the general population. In this particular group, a sudden loss of a secured airway can lead to dramatic events and even to a "Can't intubate, can't ventilate" situation. In these scenarios, the pediatric patients can rapidly develop severe hypoxemia and if adequate ventilation is not established fast, a cardiac arrest may occur. There are many factors that put pediatric population at a risk of rapid desaturation: reduced lung functional capacity that limits oxygen reserve during a period of apnea and predisposes to atelectasis development and an oxygen consumption two times higher than in the adults. In pediatric patients, cardiac arrest can occur as a consequence of bradycardia that can be triggered by hypoxemia or the laryngeal reflex if the airway is stimulated without sufficient anesthesia depth.⁴

It is essential to be familiar with the difficult airway algorithms. Those can vary between the countries or even between hospitals in the same country, but every algorithm must have in common, anticipation and preparation for the difficulties that an anesthesiologist might experience when approaching the patient's airway. In patients with craniofacial anomalies, when the airway is predicted to be difficult, it is necessary to establish a plan preoperatively, frequently proceeding with an awake intubation with a fiberscope. There is a great diversity of equipment available for the pediatric difficult airway but experience in using it is the most important issue. To approach the airway in case of an accidental extubation or if the fiberscope is not immediately available, a videolaryngoscope or a supraglottic airway device may be used. In a 'Can't intubate, can't ventilate' situation, an emergent tracheostomy is the last recourse. Extubation of a patient with difficult airway can be performed with the patient awake, after excluding systemic and airway factors that might affect maintaining a secured airway.

Managing critical events makes part of anesthesia practice. Once a critical event occurs, fast recognition and adequate strategy are essential to prevent patient's deterioration and possible mortality. We emphasize here the importance of case discussion in the preoperative period in order to facilitate identification of potential problems and complications, as well as in the postoperative period to discuss every critical event that may have occurred.⁵ Team training is extremely important and there is a special interest in performing simulation of critical events that can help anticipate and manage most challenging situations of the perioperative period. In this regard manikins can be of great help and allow multiple training sessions to every member of the staff.^{6,7}

4. Conclusion

Pediatric patients with craniofacial anomalies can be very challenging. The Pierre Robin syndrome can result in difficult ventilation and intubation, which can rapidly lead to severe hypoxemia and to cardiac arrest, if not managed promptly. It is extremely important that the tracheal tube, once well positioned, is also well secured in order to avoid accidental extubation, and its position checked after every maneuver of the head and neck of the child to confirm the proper position. Vigilance is the price of safety.

5. Conflict of interest

Nil declared by the authors

6. Consent to publish

The consent to publish this report was obtained from the parents of the child.

7. Authors' contribution

KC: main author responsible for the case, manuscript editor.

LG: manuscript editing

JB: literature research

8. References

1. Cladis F, Kumar A, Grunwaldt L, Otteson T, Ford M, Losee JE. Pierre Robin Sequence: a perioperative review. *Anesth Analg.* 2014 Aug;119(2):400-412. [PubMed] DOI: [10.1213/ANE.0000000000000301](https://doi.org/10.1213/ANE.0000000000000301)
2. Sonwane RB, Patil TS, Jewalikar S, Makhija S. Airway management for Pierre Robin sequence: an anesthetic challenge. *Ann Int Med Den Res.* 2017;3(2):AN05-AN08. [FreeFullText]
3. Wagner KM, Raskin JS, Carling NP, Felberg MA, Kanjia MK, Pan IW, et al. Unplanned intraoperative extubations in pediatric neurosurgery: analysis of case series to increase patient safety. *World Neurosurg.* 2018 Jul;115:e1-e6. [PubMed] DOI: [10.1016/j.wneu.2017.10.149](https://doi.org/10.1016/j.wneu.2017.10.149)
4. Jones P, Dager S, Peters MJ. Bradycardia during critical care intubation: mechanisms, significance and atropine. *Arch Dis Child.* 2012 Feb;97(2):139-44. [PubMed] DOI: [10.1136/adc.2010.210518](https://doi.org/10.1136/adc.2010.210518)
5. Spond M, Burns T, Rosenbaum T, Lienhart K. Crisis management of accidental extubation in a prone-positioned patient with Klippel-Feil Syndrome. *A A Case Rep.* 2016 Jun 15;6(12):383-6. [PubMed] DOI: [10.1213/XAA.0000000000000324](https://doi.org/10.1213/XAA.0000000000000324)
6. Butt MN, Hoda MQ. Knowledge, training, and practice of cricoid pressure application by the anesthesiologists of teaching hospitals of a developing country: A national survey. *Anaesth Pain Intensive Care.* 2019;23(2):151-156. DOI: [10.35975/apic.v23i2.1058](https://doi.org/10.35975/apic.v23i2.1058)
7. Akbar TM. Role of effective communication in management of medical emergencies. *Anaesth Pain Intensive Care.* 2015;18(4):229-31. [FreeFullText]