# Airway Management for Cleft Palate Repair Surgery in an Pediatric Patient with Pierre Robin Syndrome: A Case Report

## Pierre Robin Sendromlu Pediatrik Bir Hastada Yarık Damak Onarımı Cerrahisi İçin Havayolu Yönetimi: Bir Olgu Sunumu

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## ABSTRACT

The children with Pierre Robin Sequence (PRS) have anatomical and physiological challenges of airway management. The children with PRS have difficult airway caused by micrognathia, glossoptosis, and cleft palate. The patients' airway management who have PRS is dificult and requires attention. Here we present a 5-year-old girl with PRS, admitted to our hospital for cleft palate repair surgery. She also had a history of tracheostomy. Otholaryngologists had to perform trachestomy for this operation. The patient had an uneventful operative and postoperative period. We wanted to discuss the difficulties of airway management of the children with PRS with this case report. Anaesthesiologists must be ready for the challenges and otholaryngologists, must also be ready to manage these difficult airways. In addition, bronchospasm and airway obstruction after extubation are very common complications, so careful postoperative monitoring should be performed.

**Keywords:** Airway management, Anesthesia, Cleft palate, Pierre Robin syndrome, Tracheostomy, Tracheal stenosis

## ÖZ

Pierre Robin Sendromlu (PRS) çocukların hava yolu yönetiminde anatomik ve fizyolojik zorluklar vardır. PRS'lu çocuklarda, zor havayoluna sebep olan, mikrognati, glossopitozis vardır. PRS'lu hastalarda havayolu yönetimi zordur ve dikkat gerektirir. Burada PRS'lu 5 yaşında bir kız çocuğu, yarık damak onarımı için hastanemize başvuran bir hastayı sunuyoruz. Ayrıca hastanın trakeostomi öyküsü de vardı. Bu operasyon için, kulak-burun-boğaz uzmanlarının trakeostomi açması gerekti. Hastanın operasyon ve operasyon sonrası dönemi sorunsuz geçti. Bu olgu sunumu ile PRS'lu hastaların havayolu zorluklarını gözden geçirmek istedik. Anestezistler, zorluklar için hazır olmalıdır ve kulak burun boğaz uzmanları da bu zorlukların üstesinden gelinmesi için hazır bulunmalıdırlar. Ek olarak, bronkospazm ve havayolu tıkanıklıkları ekstübasyon sonrası sık görülen komplikasyonlardır, bu nedenle dikkatli postoperatif monitörizasyon uygulanmalıdır.

**Anahtar Sözcükler:** Havayolu yönetimi, Anestezi, Yarık damak, Pierre Robin sendromu, Trakeostomi, Trakeal stenoz

## BACKGROUND

The children with Robin sequence (formerly Pierre Robin Sequence) have a clinical triad of glossoptosis, micrognathia and cleft palate which is often associated with difficult airway. Because of their small jaw, these babies may have airway obstruction, even when they are awake. These children rarely may need tracheostomy after birth and sometimes they are needed to be nursed prone to displace the tonque from the back of the pharynx. In time, the jaw and oral cavity of the patient grows and allows the patient breathe normally. Chronic airway obstruction or associated syndromes may affect other organ systems. But the main problem is airway difficulty during endotracheal intubation when these patients

face anesthesia during early ages (1). We wanted to discuss the difficulties of airway management of the children with PRS with this case report.

## CASE PRESENTATION

A 5-year-old girl patient with height 98 cm, and weight 16 kg was scheduled for surgery under general anesthesia for cleft palate repair. The operation was planned 7 months ago, but patient had persistent upper respiratory tract infection and the operation was postponed. The patient had Pierre Robin sequence (PRS), retrognatia and pectus carinatus. She had tracheostomy procedure, in the Intensive Care Unit (ICU), when she was newborn because of respiratory failure. Tracheostomy was decanulated 2,5 years ago. The patient did not had a history of difficult intubation, but we were prepared for tracheostomy, considering a possible difficult intubation. It is difficult to evaluate Mallampati in patient with PRS, but her thyromental distance was within the normal range, and head-neck extension was comfortable (Figure 1).

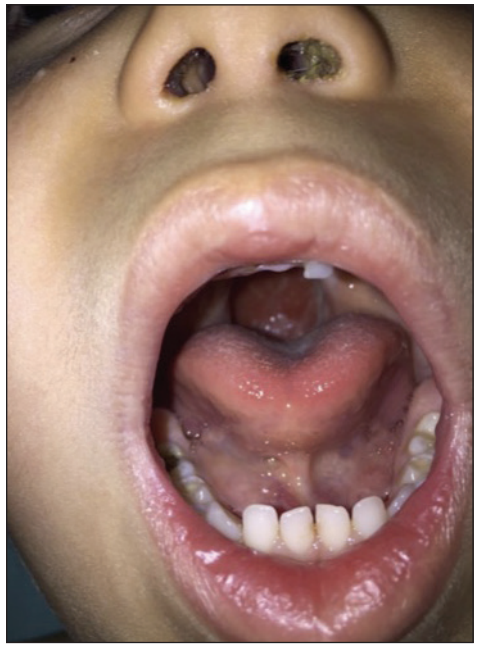

**Figure 1:** Inability to evaluate Mallampati but his head-neck extension was comfortable, and his thyromental distance was within the normal range.

She also had mental retardation, and patent ductus arteriosus. Patent ductus arteriosus had been treated and closed before. Also she had history of tracheostomy, inspite of decanulation almost 2,5 years ago, it hasn't been fully closed yet (Figure 2). Besides of tracheostomy preparations, we prepared a variety of equipments, like laryngeal mask airway (LMA), fiberoptic bronchoscope and video laringoscope. We predicted that she might have tracheal stenosis as complication of previous tracheostomy. Her preoperative laboratory and radiology tests were within the normal range. She had no electrocardiogram or chest X-ray abnormality. On the operating day, no premadication was administered to the patient, and she was monitored by electrocardiography, Peripheral Oxygen Saturation ($SpO_2$), noninvasive blood pressure and end-tidal carbon dioxide pressure ($ETCO_2$) after intubation. 1mcg.kg$^{-1}$ fentanyl, 3 mg.kg$^{-1}$ sodium thiopental and 0,6 mg.kg$^{-1}$ rocuronium was given for anesthesia induction. Tracheal intubation was established with videolaryngoscope (Figure 3). We tried to intubate with 4 mm internal diameter (ID) endotracheal tube (ETT), but the ETT's diameter was larger than the patient's vocal cords. Then we tried with 3,5 mm ID ETT, but it was also large. After second attempt of intubation we administered 1 mg.kg$^{-1}$ prednisolone, to prevent laryngeal oedema. She was succesfully intubated with 3.0 mm ID ETT. However, ETT was too short fort he patient, because it was small size, and patient's ventilation was not adequate. The operation was in the oral cavity and short ETT had a risk of self extubation. Due to the risk of self extubation, we decided to perform tracheostomy procedure. Tracheostomy was performed by otoloringologists, as the patient had a tracheostomy when she was new-born. After intubation the ventilator settings were; tidal volume 110 ml,

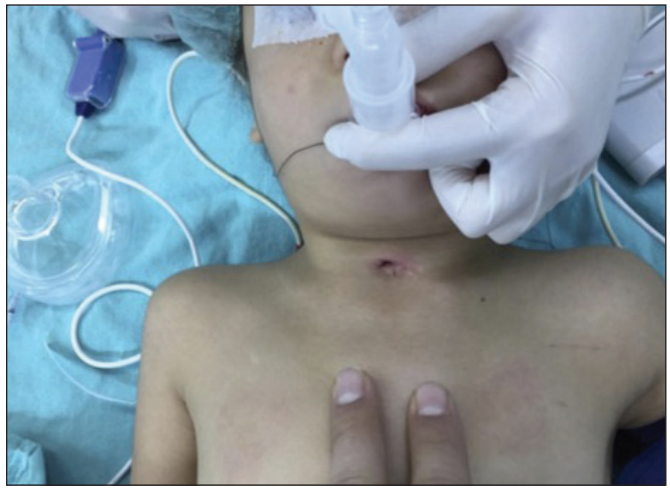

**Figure 2:** Patient had history of tracheostomy which hasn't fully closed (Picture 2) inspite of decanulation almost 2 years ago.

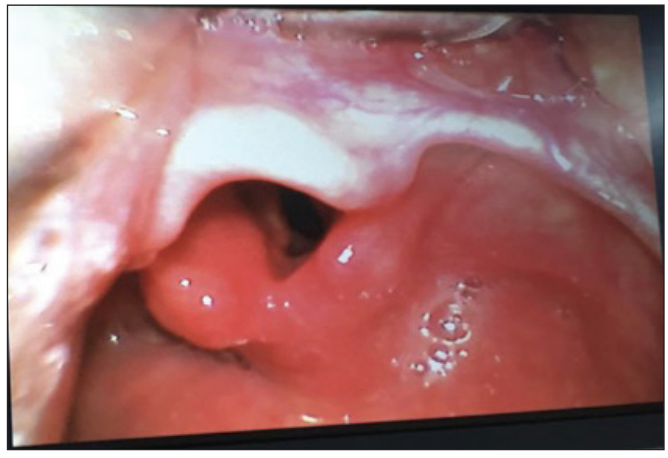

**Figure 3:** Vocal cords and epiglottis.

respiratory rate 24/minute, inspiration and expiration ratio was 1:1 at volume control mode. Airway pressure was kept below 15 cm $H_2O$ and $ETCO_2$ was kept between 31-35 mmHg. Intravenous 2–3 $mcg.mL^{-1}$ remifentanyl infusion, sevoflurane 2-2,5% and 0,1 $mg.kg^{-1}$ rocuronium (totally 10 mg) was intermittently injected intravenously for anesthesia maintance, and remifentanyl dosage was adjusted according to noninvasive blood pressure monitoring. Operation lasted 120 minutes, and hemodynamic parameters were stable during the operation. $SpO_2$ levels remained 100% during entire operation. After the operation, we transported the patient to the pediatric ICU with tracheostomy cannula. She spent her first postoperative day in the pediatric ICU. Her vitals were within normal range during the perioperative period and all parameters were normal at first postoperative day. 24 hours later, the patient was transfered from the ICU to regular ward with her tracheostomy cannula. Her respiratory and hemodynamic parameters remained stable until she was discharged from hospital. She had no sign of respiratory failure. The patient was discharged from hospital on the postoperative fifth day with her tracheostomy cannula. She is under otolaryngologists follow up for tracheostomy decanulation.

## DISCUSSION

In literature rewiew, PRS may have difficult airway during intubation for general anesthesia. A variety of techniques can be used, such as; LMA, intubating LMA, fiberoptic bronchoscope, retrograde intubation, and awake fiberoptic intubation (1-4). Other studies also showed that, neonates with PRS were successfully intubated with awake fiberoptic bronchoscope through a laryngeal mask (8).

Asai et al. reported that, awake fiberoptic intubation can cause severe hypoxia in neonates. But this problem can be solved by insertion of a laryngeal mask while the patient is still awake. They had a patient; one-month-old neonate with PRS, who had constant upper airway obstruction, and the patient's tongue was fixated to mandible surgically. They planned awake fiberoptic intubation but failed, because the patient's $SpO_2$ levels rapidly decreased during the attempts. They were able to insert laryngeal mask while the patient was still awake and oxygenation improved as upper airway obstruction was revealed by laryngeal mask. And after that they were able to intubate the patient by fiberoptic endoscopy via laryngeal mask (2).

We were prepared for difficult airway challenge including fiberoptic bronchoscope, but we decided to try endotracheal intubation with video laryngoscope first. Inspite of patient's retrognatia and small jaw, we did not have any problem during mask ventilation. But, we had some problems with ETT diameter size during endotracheal intubation.

Marston et al. reviewed pediatric otolaryngology practice database of newborns before 3 months of age, with PRS who needed airway intervation. Indications for airway interventions, anesthetic managements, methods of endotracheal intubations, and comorbid diseases were recorded. There were thirty-five newborns with PRS; twenty of them had isolated PRS, and fifteen of them had PRS and a coexisting syndrome. Thirteen of these newborns (37%) were intubated with direct laryngoscopy, prior to mandibular distraction osteogenesis. The remaining twenty two of them (63%) were intubated by a flexible fiberoptic bronchoscope, since they were not be able to be intubated with direct laryngoscopy. None of the patients required rescue laryngeal mask airway or emergency tracheotomy, and all of them survived. These series demonstrate that, endotracheal intubation is safe and effective in newborns with PRS. And intubation over a fiberoptic bronchoscope is a reliable method for patients who could not be intubated with direct laryngoscopy. Airway management in newborns with PRS is a known challenge, but these difficult airway cases can be managed by experienced otolaryngologists and anesthesiologists (3).

Meyer et al. described some interventions used for successful airway management in patients with PRS. They reviewed pediatric otolaryngology clinic records of patients with cleft and craniofacial lessions, and children with PRS were identified. Data about; feeding interventions, airway interventions, and comorbid diseases were extracted. Seventy four cases of PRS were found, and thirty eight of them required airway intervention other than prone positioning. Fourteen of these thirty eight patients had nasogharyngeal airway and/or short-term endotracheal intubation and did not require surgical intervention. But the rest of these thirty eight patients (24 patients) needed surgical intervention. Also, eighteen of the twenty four patients, underwent distraction osteogenesis of the mandible, one of them underwent tracheostomy, and five of them underwent tracheostomy followed by eventual distraction. In these series, more than 50% of children with PRS, an airway intervention was needed. These interventions were either nonsurgical or surgical (4).

The airway management of PRS cases require special attention, and preoperative monitoring should be guaranteed. If the patient has a high risk for general anesthesia, regional anesthesia can be a good alternative (5). In fact, regional anesthesia has been shown to be a good alternative in neonates by previous studies (6).

Onal et al. evaluated a 8-year-old boy with PRS. The patient had restricted mouth openining, short extremities, mild airway obstruction, and respiratory distress at preoperative evaluation. In this case, 0.4 $mg.kg^{-1}$ hyperbaric bupivacaine was administered to L4-5 vertebral space for

spinal block. They suggested that, preperations for difficult airway management should be ready in the operating room, and spinal anesthesia may be a reliable technique for lower extremity surgeries (7).

In our case, patient's trachea was succesfully intubated with videolaryngoscope. But we had to perform a tracheostomy procedure because the surgery would be in the oral cavity and the oral intubation with a short tracheal tube wouldn't be safe. After the surgery the patient was transfered to the pediatric ICU, and she was discharged from hospital with her tracheostomy cannula. So she did not has any respiratory problems at postoperative period. She is under otolaringologists follow up for tracheostomy decannulation.

## KEY POINTS

- Patients with PRS has small chin, hight-arched palate, large tongue, and small openning at the roof of the mounth and all of these conditions can cause difficult airway management.
- All the preparations for managing the difficult airway, including fiberoptic broncoscope, should be ready in the operating room.
- LMA, intubating LMA, fiberoptic bronchoscope, retrograde intubation and rigid bronchoscopy can be used for airway management.
- If patient has a history of tracheostomy, it is necessary to clarify whether the patient has or has not tracheal stenosis.
- Bronchospasm and airway obstruction after extubation are very common complications, and careful postoperative monitoring should be performed.
- It is recommended that, the patient should spend the first post-operative day in the paediatric intensive care unit where the patient can be closely monitored.

## CONCLUSION

Airway management in patients with PRS is difficult and requires attention. Anaesthesiologists and otolaryngologists must be ready for the difficulties of the children with PRS. They must be ready for the interventions which are reguired to successfully manage these difficult airways. In addition, bronchospasm and airway obstruction after extubation are very common complications, and careful postoperative monitoring should be performed.

Literature review reveals that; in a patient with upper airway obstruction, intubation with videolaringoscopy or fiberoptic intubation can aggravate hypoxia, laryngeal mask can be used for facilitating oxygenation (by relieving upper airway obstruction) and in facilitating fiberoptic intubation (8).

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**Consent for publication:** Patient's parents gave written informed consent for publication of this case report.

**Conflict of interest:** The authors declare that they have no conflict of interest.

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