

# Management of Pierre Robin Sequence (PRS) in a Direct Laryngoscopy and Cleft Palate Repair

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## Objectives:

The learner will be able to define the characteristics of PRS and their implications.

The learner will be able to develop an anesthetic plan for a patient with PRS.

The learner will understand how to safely induce a patient with PRS.

## Introduction:

Pierre Robin Sequence (PRS) consists of triad of micrognathia, glossoptosis, and airway obstruction, as shown in figure 1.

Cleft palate is present in the majority of PRS cases. Malnutrition is common due to the abnormal anatomy of the oropharynx and often requires NG tubes for nutrition.

PRS can be associated with other congenital disorders such as stickler syndrome, velocardiofacial syndrome, hemifacial microsomia, and fetal alcohol syndrome.<sup>1,2</sup>

Methods for relieving airway obstruction in PRS include: prone or lateral positioning, CPAP, Lip-tongue adhesion - usually up until a year old, mandibular distraction, nasopharyngeal intubation, and tracheostomy is severe.<sup>3</sup>

## Case Report:

A 15 month old female, weighing 10 kg, presenting for direct laryngoscopy and bronchoscopy (DLB), bilateral myringotomy tubes (BMT) and cleft palate repair. PMH of Pierre Robin Sequence with cleft palate and OSA. Has an NG tube for nutrition. Normal ECHO.

## Plan:

Surgeon wanted to place the BMT first, then perform a DLB to examine the airway. Afterwards, ENT would intubate and perform the cleft palate repair.

## Evidence/Practice Based Guidelines:

### Pre-op Assessment

Obtain a detailed airway and cardiac assessment, evaluate for aspiration risk and any recent URI's. If the patient has other congenital syndromes, cardiology should be consulted due to potential for underlying cardiac abnormalities.<sup>3,5</sup>

### Induction/Intubation

In preparation, emergency airway equipment must be ready with ENT surgeons ready for emergency surgical airway.

Patients with PRS are at high risk of losing the ability to mask ventilate and intubate. Awake intubations can done with precedex and ketamine, however, this can be difficult in uncooperative children.

It also accepted that mask induction can be done as long as spontaneous ventilation is maintained. To relieve airway obstructions, two handed jaw thrust and airway adjuncts, such as an oral airway or LMA, can be used. A right paraglossal approach has been a common DL method for intubating patients with PRS, as shown in figure 2.<sup>3,5</sup>

### Emergence/Post-op

To decrease possible airway obstruction and post-op respiratory complications, consider limiting opioids by using non-opioid analgesics and local/regional blocks. Examination for airway edema and an air leak test should be done due to the surgical airway manipulation. Complete NMB reversal and an awake extubation is also essential. If obstruction occurs after extubation, the patient should be placed in the lateral position. If obstruction still occurs, CPAP or a tongue stitch can be used in the PACU.<sup>3,5</sup>

## Discussion of the Case:

After mask induction with sevoflurane, an oral airway was placed and the patient was spontaneously breathing. An IV was placed and a propofol infusion was started. The BMT was completed and moved to DL by turning the bed 90 degrees for ENT. ENT had a grade 4 view on their videoscope by resident and surgeon.

## Evaluating the Risks of Preceding:

After discussion between the parents and the anesthesia and surgical teams, it was decided to cancel the cleft palate repair.

ENT surgeries have an increased chance of accidental extubation. Surgical site bleeding and swelling could also make an intra-op reintubation or post op intubation very difficult. This was also an elective procedure with no improvement to the underlying issue. These factors paired with difficulties associated with PRS and the grade iv view lead to the cancellation of the cleft palate repair.

The agreed safer course of action would be to perform the mandibular distraction to extend the jaw and help open up the airway to create better conditions for elective intubations.

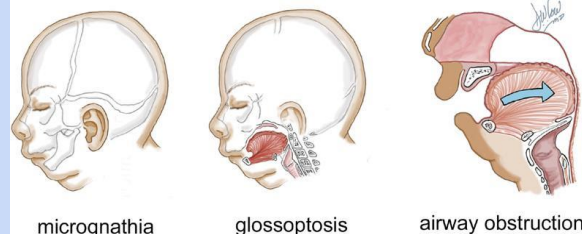


Fig 1: Pierre Robin Sequence triad: micrognathia, glossoptosis, upper airway obstruction<sup>6</sup>

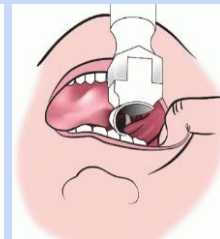


Fig 2: Paraglossal approach<sup>6</sup>

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