CASE SERIES

# Airway challenges in Pierre Robin Syndrome children with cleft conditions in a surgical mission

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**Abstract**

Surgical missions are often conducted in outreach areas where facilities are often not as advanced as in an institution. Excellent team coordination, detailed planning, and good communication are necessary for a successful outcome. Unfamiliar work environment, new acquaintances within the team, and language barriers at the mission site are some of the challenges often faced. Careful patient selection is crucial in a mission setup as most complications can be traced back to the preanaesthetic check and preoperative preparedness. We describe airway management strategies (airway assessment, mask ventilation, intubation options, and the postoperative airway plan) contrived by our anaesthesia team at one such surgical mission where four children with Pierre Robin Syndrome and incomplete cleft palate (involving posterior hard palate and complete soft palate)were successfully operated.

**Keywords:** Cleft Lip, Cleft Palate, Paediatric Anaesthesia, Pierre Robin Syndrome, Surgical Mission

## Introduction

Surgical missions are often conducted in outreach areas where facilities are often not as advanced as in an institution [1]. Careful patient selection is crucial as most complications can be traced back to the Preanaesthetic Check (PAC) and preoperative preparedness [2]. Most missions do not schedule syndromic children and children with anticipated difficult airways. However, as the expertise and facilities available were good at this particular mission site, four children with Pierre Robin Syndrome (PRS) were accepted for surgery and successfully operated. The stigma associated with being a cleft child is immense. The parents of these children strive hard to get functional/cosmetic correction done and they face rejection at various stages in the system. The fear of handling small babies who are syndromic and the lack of medical facilities at smaller hospitals add to their burden. It is not uncommon to encounter parents who hide important information (such as cardiac condition/ respiratory infection) during PAC, with the hope of being posted for surgery despite all the risks.

This mission site was shortlisted after a thorough fact-find was undertaken by the anaesthesia team to ensure that the infrastructure and ancillary facilities were good. The presence of a functioning

blood bank, a well-equipped Post-anaesthesia Care Unit (PACU) and a Paediatric Intensive Care Unit (PICU) were additional strengths of this mission site. Necessary airway equipment was procured on the day prior to surgery. A difficult airway cart [including airway adjuncts such as Oropharyngeal Airway (OPA), Nasopharyngeal Airway (NPA), stylette, bougie and appropriate-sized laryngeal mask airways] was kept ready.Craniofacial anomalies can lead to difficult mask ventilation. Alternate techniques of mask ventilation (such as a two-handed two-person technique, or the use of a jaw thrust manoeuvre) with OPA or NPA in place can alleviate this problem. Alternatively, mask ventilation can be attempted in the lateral position to tackle the problem of the large tongue falling backward causing airway obstruction. While retrognathia and glossoptosis lead to airway obstruction in children with PRS, cleft palate adds to the problem, posing a difficulty in laryngoscopy. The practical problem of the laryngoscope blade slipping into a left-sided palatal cleft is common and can be overcome by placing a small gauze roll to fill the alveolar cleft during laryngoscopy. Reporting of these cases reiterates that patient safety is foremost in such scenarios and that a difficult airway can be safely dealt with at outreach areas with a meticulous approach at each stage and good team coordination.

**Case series**

Fifty-one children were screened on this mission and 32 children were scheduled to be operated. After a multidisciplinary discussion between anaesthesiologist, surgeon and paediatrician, four children with PRS and incomplete cleft palates (involving posterior hard palate and complete soft palate) were successfully operated at this camp. None of these children had an active upper respiratory tract infection, other comorbid conditions, or existing lip abnormalities and they had not been operated upon previously for their cleft condition.

Parents of these children were informed about the risks of a difficult airway that safe and sincere attempts would be made by an experienced team to secure the airway. In the event of failure to secure the airway. In the event of failure to secure the airway, the child would be awakened and subsequently referred to a centre equipped with a Video Laryngoscope (VL) and Fibreoptic Bronchoscope (FOB). The need for prolonged observation in the PACU/PICU was also explained. As per protocol, all children underwent a thorough PAC. All the four children were underweight. However, as delay in primary surgery is the most common reason for poor nourishment, we did not reject them on the basis of being underweight [3].

As per mission protocol, inhalational induction with sevoflurane was performed and an intravenous line was secured. The plane of anaesthesia was deepened with 1-2 mg/kg of propofol and 2-3 mcg/kg of fentanyl and intubation was attempted. The initial two attempts were performed without muscle relaxation. The third attempt (the final attempt after which the child would be woken up if the intubation was unsuccessful) was performed by the senior anaesthesiology team leader 1 minute after administering 2 mg/kg of succinylcholine. Mask ventilation was done with 100% oxygen between laryngoscopic attempts. There was no fall in saturation below 90%. Relevant clinical information and technique of intubation are detailed in Table 1.

**Table 1: Case description and details of airway management**

| Age | Weight (kg) | Airway assessment | Initial CL grade and details of intubating procedure | Comments |
|---|---|---|---|---|
| 7 years | 12.3 | Mouth opening – adequate (more than 3 finger breadths). Large tongue and history of snoring present. Thyromental distance was decreased. Moderate retrognathia present. Upper lip bite test – Class 3. Room air saturation ⩾ 97% | Cook's modification of CL Grade 3B that improved to Grade 2B with OELM. Succinylcholine administered, intubation successful at 3rd attempt using oral RAE by senior anaesthesiologist | A two-handed jaw thrust technique used for mask ventilation. Mask ventilation was easy. NPA remained *in situ* for 24 hours postoperatively |
| 4 years | 12 | Mouth opening – adequate (more than 3 finger breadths). No history of snoring. Thyro-mental distance was decreased. Mild retrognathia present. Upper lip bite test – Class 2 room air saturation ⩾ 97% | Cook's modification of CL Grade 3A improved to Grade 2B with OELM. Intubated (oral RAE) in the 2nd attempt | E-C technique of mask ventilation with neck extension was performed. Mask ventilation was easy. |
| 1 year | 7 | Mouth opening appeared adequate.Large tongue and history of snoring present. Thyromental distance was decreased. Mild retrognathia present. Upper lip bite test – could not be assessed.Room air saturation ⩾ 97% | Cook's modification of CL Grade 4 improved to Grade 3B with OELM. Intubated on 3rd attempt by senior anaesthesiologist with size 3 mm ID conventional ETT with a J-shaped paediatric stylette after administering succinylcholine. Dingman's retractor caused an unacceptable kink of the ETT. A 7 Fr ureteric catheter was used as an "AEC" and the ETT was exchanged for a 3 mm ID flexometallic tube | E-C technique of mask ventilation with neck extension was performed. OPA used during mask ventilation. Paediatric stylette available could not be easily negotiated across the preformed bend of the oral RAE tube; hence conventional ETT was used initially. NPA remained *in situ* for 24 hours postoperatively |
| 2 years | 9.8 | Mouth opening – adequate (more than 3 finger breadths). No history of snoring. Thyromental distance was decreased. Mild retrognathia present. Upper lip bite test – Class 2. Room air saturation ⩾ 97% | Cook's modification of CL Grade 3B, improved to 3A with OELM. Intubated with oral RAE using a J shaped paediatric stylette | E-C technique of mask ventilation with neck extension was performed. OPA used during mask ventilation |

Anaesthesia was maintained with sevoflurane in an air-oxygen (1:1) mixture with spontaneous ventilation. Local infiltration with 1% lignocaine and 0.25% bupivacaine with 1 in 200,000 adrenaline was administered by the surgeon after placing the Dingman retractor. Rectal paracetamol (20 mg/kg) was given for supplemental analgesia. All children received dexamethasone 0.1 mg/kg intraoperatively. At the end of the procedure, a thorough oropharyngeal suctioning was performed and the children were extubated in the lateral position when fully awake.Fearing the development of airway oedema (oedema of tongue and posterior pharynx due to instrumentation) in the immediate postoperative period, all children received a tongue stitch and were nursed overnight in a lateral position [4-5]. The tongue stitch could be used to draw the tongue forward if there was airway obstruction. A silicon NPA was placed in one nostril prior to extubation, and it was retained in place for 24 hours postoperatively or as deemed necessary. The postoperative period was uneventful. Figure 1 shows the preoperative and postoperative images of a 7-year-old child with PRS operated at our mission.

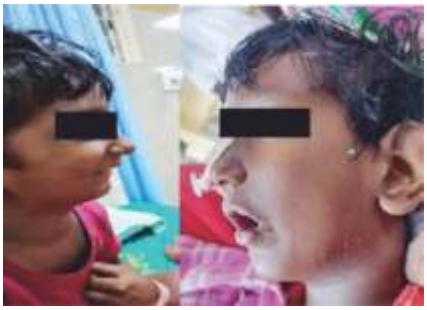

**Figure 1: Preoperative and postoperative images of a 7 year-old child with Pierre Robin Syndrome operated at our mission**

**Discussion**

Ventilation and intubation are challenging due to craniofacial dysmorphology in PRS. Establishing a good mask seal during face mask ventilation can be an arduous task due to facial deformities. Direct laryngoscopy and tracheal intubation tend to be difficult but become easier with age and mandibular growth. Alternatives such as the use of VL and FOB can enhance the intubation success. A tracheostomy may be rarely needed [6]. The availability of VL at surgical missions could prove a game changer in the decision to include children with PRS for surgery. VL is light, easy to transport and clean, and takes less time to set up. All team members can see the glottis aperture on the VL screen and provide constructive feedback. This is an added advantage. Hybrid techniques using VL and FOB can also be attempted. The technique of passing a guide wire through the working channel of the FOB and subsequently railroading the Endotracheal Tube (ETT) over it has been described in this context [6].

In our mission, we encountered a unique problem with the third case wherein, after a difficult experience of intubating the airway in the 3rd attempt using a conventional ETT, placement of the Dingman's retractor resulted in near-complete kinking of the ETT making ventilation next to impossible. To overcome this problem, a ureteric catheter of 7 French size was used as an Airway Exchange Catheter (AEC) and a flexometallic tube was successfully railroaded over this. This was possible due to the team effort, good communication and constructive feedback from all team members at all stages of the intubation process. The benefits of the use of NPA to alleviate airway obstruction cannot be adequately stressed.

Two out of 4 children had a NPA retained postoperatively for 24 hours. Whitaker *et al.,* (2003) had devised a novel technique of using a 5 mm Internal Diameter (ID) ETT to prevent airway obstruction in the absence of a paediatric NPA at

their hospital [7]. Despite being a mission set up, we could arrange for all the appropriate airway equipment including small-sized NPA on the day before surgery. Simulated PRS manikin-based training and the use of available airway equipment in various combinations on these manikins can help the anaesthesiologist understand the airway better and enhance the success of intubation [8-10].

## Conclusion

Asking leading questions, performing a thorough systemic examination, multidisciplinary teamwork, planning, airway preparedness, and parental counseling can put the whole team in the foreground to face the challenges of anaesthetising and successfully operating a syndromic child at surgical missions.

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