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Anaesthetic Management in Pierre Robin Sequence with Severe Upper Airway Obstruction: A Case Report

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Abstract: Introduction: A rare congenital birth abnormality known as Pierre Robin sequence (PRS) is characterised by an underdeveloped jaw, tongue displacement backward, and obstruction of the upper airway. Early breathing and feeding difficulties are typical in babies with PRS because of the placement of the tongue, the smaller jaw, and the development of a cleft palate.
Case presentation: In this report, we will discuss a case of PRS, who had history of recurrent respiratory tract infections present. Also detailing the anaesthetic management of a 2year old child weighing 9.2 kg diagnosed with Pierre Robin sequence (PRS) with severe upper airway obstruction undergoing complete cleft palate posted for tongue lip adhesion release with palatoplasty.
Discussion: The procedures mentioned above call for general anaesthesia as an anaesthetic intervention. For patients undergoing computed tomography (CT) and magnetic resonance imaging (MRI), anesthesiology-assisted sedation may be necessary.
Conclusion: This study concluded that a PRS patient with a complete cleft palate poses challenges on airway management.

Keywords: Pierre robin syndrome, micrognathia, Cleft palate, intubation, Anaesthesia

1. Introduction:

Micrognathia, posterior-inferior displacement of the tongue base (glossoptosis), and airway obstruction comprise the Pierre Robin sequence (PRS).[1] Every year, up to 1 in 14,000 babies are affected by PRS. While PPRS can happen on its own, it is more frequently linked to other disorders such Treacher-Collins syndrome, velocardiofacial syndrome, Stickler syndrome, and foetal alcohol syndrome. [2,3] Neonatals primarily show respiratory distress symptoms (stridor, retractions, and cyanosis) at birth; some also show feeding issues, reflux, aspiration, and failure to thrive. [4,5]
We will present a case of PRS, who had history of recurrent respiratory tract infections present. This study presents a case report detailing the anaesthetic management of a patient in Pierre Robin sequence with severe upper airway obstruction undergoing complete cleft palate posted for tongue lip adhesion release with palatoplasty.

Case Presentation:
In this report, we will discuss the approach of anaesthetic management of a 2year old child weighing 9.2 kg diagnosed with PRS and an associated complete cleft palate posted for tongue lip adhesion release with palatoplasty. A term girl baby with birth weight of 1.6 kg, born by LSCS. No family history of congenital disorders was present but mother was diagnosed with rheumatic heart disease, mild MS/MR at 5th month of gestation – as an incidental finding in echo. Baby developed severe dyspnoea immediately after birth and had micrognathia with cleft palate. Baby was intubated in view of severe airway obstruction and was on mechanical ventilation, on day nine of life tongue lip adhesion surgery was done. Post 14days of ventilation extubation was done. Has a history of NICU admission for two months since birth and on nasogastric feed for 6months. Had delayed milestones and history of recurrent respiratory tract infections present. Blood investigations –Hb 7.9, other tests were normal. Echo shows –EF 70% with normal biventricular function and contraction. On general physical examination- baby was alert and active, height -85cm, weight-9.2kg. Oral examination – micrognathia and u-shaped cleft palate.

Airway examination – COPUR score:15, with anticipated difficult intubation. History revealed deferred surgery -1year back in view of difficult intubation. On day of surgery – difficult intubation arrangements were done. Video laryngoscopy, paediatric fiber optic intubation kept ready. Before the operation, breathing circuits and anaesthesia workstation checkup done. Baby was premedicated with glycopyrrolate 0.1 mg and midazolam 0.5 mg in the preoperative room and shifted to OR. In OR ECG, NIBP, axillary temperature probe and pulse oximetry were connected. Child was pre oxygenated,and premedicated with inj. Glycopyrollate. Inj. Fentanyl 20 mcgs was given and induction done with sevoflurane. Mask ventilation was attempted with 2 handed jaw thrust technique, once mask ventilation was possible inj. atracurium 4.5 mg was given. Child was intubated with 3.5 mm cuffed flexometallic tube with the aid of bougie using videolaryngoscopy with hyperacute blade. POGO score 80 % and intubation difficulty score >5. Throat packing was done and ett secured at 14 cms along the right angle of the mouth. Paracetamol suppository 250 mg placed per rectal prior to surgery. Anaesthesia maintained with Sevoflurane (0.5-1.5%) + Oxygen and air (2:2), Intermittent doses of atracurium and pressure-controlled ventilation. Child was given anti emetics and dexamethasone 1 mg. Intraoperative period was uneventful, tongue adhesion release done followed by palatoplasty. Intra-op blood loss was approximately 50 ml. Post procedure child was given reversal and was extubated after adequate spontaneous respiratory efforts without any complications. Child was shifted PICU for observation.

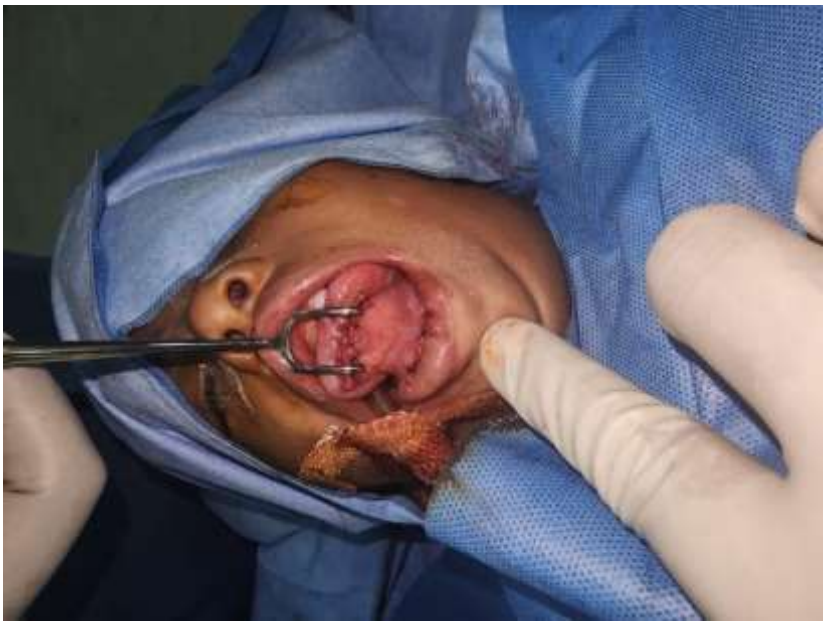

Fig:1

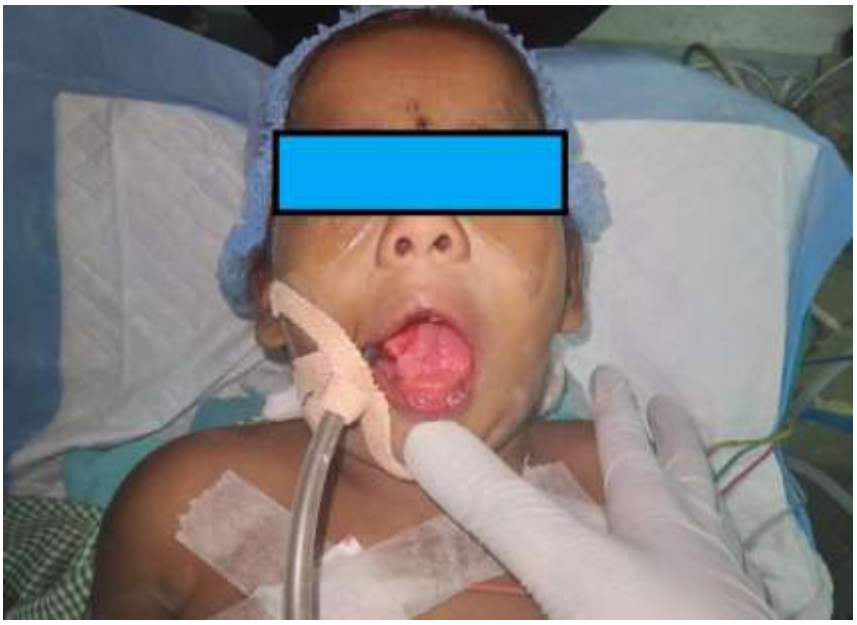

Fig:2

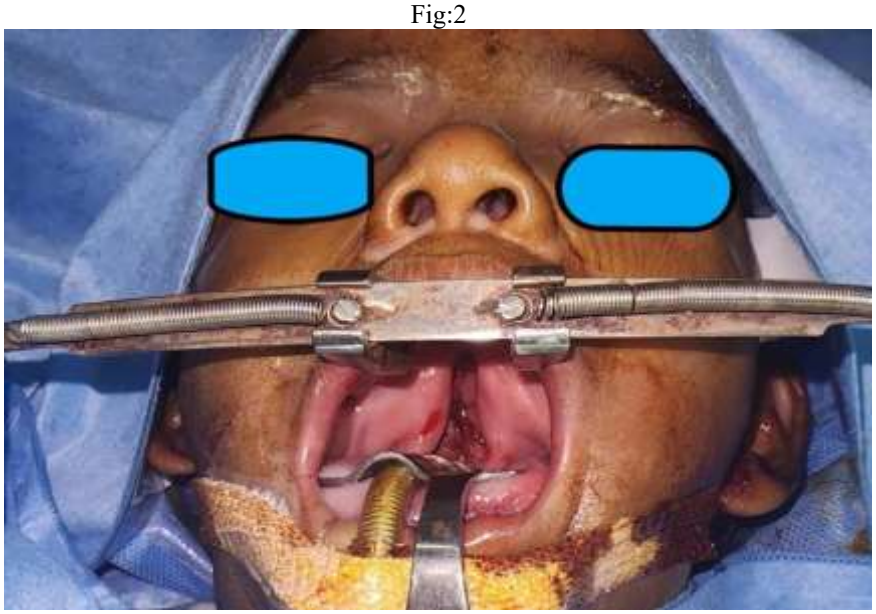

Fig: 3

2. Discussion:

French stomatologist Dr. Robin discovered the parameters that define PRS in 1923. Dr. Robin named the condition glossoptosis, which is the tongue obstructing the oral cavity with a short mandible. [6,7] The small mandible (micrognathia), the backward and downward displacement of the tongue base (glossoptosis), and airway obstruction are the clinical characteristics that characterise Parkinson's disease (PRS). PRS is defined in certain literatures as involving micrognathia, glossoptosis, and cleft palate. [8] Though it is not always present, PRS is frequently linked to cleft palates. Nonetheless, all PRS patients have airway obstruction, which supports the clinical diagnosis.[9] In our case cleft palates present in child with severe upper airway obstruction. The severity of PRS symptoms varies, and minor cases may go undiagnosed. This could account for the fluctuating incidence of 1:5,000–1:85,000. Patients experiencing breathing and feeding difficulties are among the signs and symptoms of PRS. A newborn or infant with respiratory issues may exhibit stridor, retractions, and cyanosis, as well as respiratory distress and airway obstruction. In addition, reflux, trouble feeding, and underdevelopment could result from airway blockage.[9] Many operations, such as tongue-lip adhesion, mandibular osteogenesis, Nissen fundoplication, gastrostomy, or tracheostomy in cases of refractory airway obstruction or failure to thrive, require anaesthesia for patients with PRS. [8] The anaesthetist may find it extremely difficult to manage a PRS patient's

airway. Respiratory distress, hypoxia, and respiratory failure are examples of perioperative problems that can result from airway blockage and challenging intubation. Patients with Parkinson's disease (PRS) is more susceptible to the negative effects of opioids due to their chronic airway obstruction and hypoxia. [9,10] A variety of methods and airway devices can be used to manage an airway in a patient with PRS. You have three options for doing the intubation: awake, sedation, or general anaesthesia. It is important to attempt to maintain spontaneous ventilation when managing a predicted paediatric difficult airway while under anaesthesia or sedation. A PRS patient's intubation and/or ventilation may be aided by the use of LMA, video laryngoscopes, fiber-optic scopes, and retrograde wires, among other airway devices and equipment. [11,12] One can insert an LMA without anaesthesia or sedation if there is acute respiratory distress and imminent failure. In 1992, Markakis et al. [13] provided a description of this. Later, in 2008, Asai et al. [14] reported on a group of five PRS infants in which the LMA was inserted without the need for sedation or local anaesthesia. After placing an LMA and restoring airway patency, all five of the newborns (weighing between 2.8 and 3.5 kg) became quiet, according to the authors. In order to assist the insertion of an endotracheal tube, Stricker et al. [15] reported putting an awake LMA in PRS patients prior to the induction of anaesthesia and then introducing a fiber-optic scope.

Sustaining spontaneous breathing is crucial if general anaesthesia is administered prior to airway closure. Because to airway blockage, maintaining ventilation—whether spontaneous or assisted—can be extremely challenging or perhaps impossible. As a result, it is necessary to have airway adjuncts available and to adhere to a complicated airway management protocol. The management of difficult ventilation and/or difficult intubation involves employing airways, whether oral pharyngeal, nasopharyngeal, or laryngeal, to aid in ventilation, calling for assistance, and performing a two-handed jaw push. When these manoeuvres fail to clear the patient's airway, endotracheal intubation trials should be carried out.

Adequate laryngoscopy proficiency for paediatric patients is a must for managing PRS cases under anaesthesia. Henderson [16] published a description of a paraglossal technique for PRS patient intubation in 1997. Semjen et al. [17] attempted to intubate six PRS patients with this method in 2008. Five out of the six PRS patients were successfully intubated. Using this approach, the tongue is moved to the left by applying pressure from the left and right corners of the mouth to advance the laryngoscope's blade. This method shortens the distance to the glottis's opening, making intubation easier. An LMA or nasal inhalation can be used to introduce a fiber-optic scope. In these situations, oxygen can be administered with or without anaesthetic gases using a nasopharyngeal airway, which is located in the opposite nares. The endotracheal tube can be inserted orally while the scope is entered nasally to view the glottic aperture and the tube is progressed under vision if the patient is too little to fit on the fiber-optic scope.[18]

Patients with PRS are more likely to experience postoperative respiratory problems because they already have prior airway obstruction, obstructive sleep apnea (OSA), persistent hypoxia, and higher opioid sensitivity.[19]By utilising nasopharyngeal/oropharyngeal airways or only neck extension and jaw thrust, the development of postoperative problems can be reduced. Nursing the infant in a lateral posture with their neck extended after surgery can also help to minimise potential issues. [20]

Advancements in video-
enabled instruments, such flexible bronchoscopes (also known as fiberoptic scopes or flexible intubating scopes) and videolaryngoscopes, have made it possible for anaesthetists to create novel methods for
treating patients who have difficult tracheal intubation.

Under some conditions, a hybrid technique
that makes use of both technologies at once can maximise their positive attributes while minimising their drawbacks.The videolaryngoscope lateralise the tongue and other surrounding tissues which allows the flexible bronchoscope to have a clear view and unhindered path to the glottis.

In situations where videolaryngoscopy provides a sufficient view but the tracheal tube is difficult to guide throu gh the glottis, the flexible bronchoscope can be utilised as a steerable stylet to aid in tracheal intubation.When th ere is resistance to tracheal tube advancement, the videolaryngoscopic view can also offer guidance for the flexi ble bronchoscope in advancing the tracheal tube.The technical challenges limit the use of the hybrid technique as it demands the need of two experienced clinicians to operate advanced airway equipment in a small, technically challenging space, simultaneously.[21]

3. Conclusion:

We concluded that a PRS patient with a complete cleft palate poses challenges on airway management. Use of video laryngoscopy in these patients (PRS) with anticipated difficult airway significantly improved laryngeal exposure thus facilitating endotracheal intubation.

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