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Tongue-tied: Management in Pierre Robin Sequence

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Abstract
An 18-month-old male with Pierre Robin Sequence (PRS) presented to A&E with airway obstruction and hypoxia due to retroglossoptosis. The patient was resuscitated immediately and intubated. Gold standard treatment was surgical management by mandibular distraction osteogenesis. However, as the patient was unable to afford the surgery, a simpler and cheaper surgical technique had to be employed. The procedure involved pulling the base of the tongue anteriorly and tying to the hyoid bone. This maintained airway patency and patient was extubated. Mother was given feeding and positioning advice for the child. It is expected that the mandibular growth will eventually catch up with the tongue growth.

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1. Background

Pierre Robin Sequence (PRS) is a congenital condition of facial abnormalities in human defined by a classic triad: (a) Micrognathia (b) Retroglossoptosis (c) Airway obstruction. It has a prevalence of 1-in-8500 live-births, with a male to female ratio of 1:1.

Airway obstruction and respiratory distress are clinical hallmarks and this is due to retropositioning of the tongue base [1]. Patients may present with stridor, retractions, cyanosis, feeding difficulties, reflux, otitis media or even failure to thrive [2]. The average age of presentation varies between few days post-natal to 2-years-old [2].

Management depends on the airway obstruction severity. This includes prone-positioning, nasopharyngeal airways, tongue-lip adhesion (TLA), mandibular distraction osteogenesis (MDO) and tracheostomy.

Treatment options have not been widely described before. This case was of particular interest as it highlights that a less common procedure that is simpler and cheaper, can be employed instead of the gold-standard surgery, in resource-scarce settings.

2. Case Presentation

The patient was an 18-month-old boy, brought into the Paediatric A&E with sudden decorticate posturing of limbs, seizures, peripheral cyanosis, pooling of secretions, fever and falling oxygen saturation. He was acutely treated with airway management and administration of oxygen, intravenous fluids, and phenytoin. Hypoxia subsided on starting oxygen and seizures resolved in 30 minutes following phenytoin administration. However, oral and nasal secretions persisted.

According to the mother, the child had had a fever, noisy breathing and cough for 2 days before this admission. Fever was described as low-grade, intermittent, with no rigors or diurnal variation. He also had a history of recurrent upper/lower respiratory tract infections, sepsis and hypoxic seizures.

The patient was clinically diagnosed with Global Development Delay and PRS, a few weeks after birth. When he was 8-months-old, a ventriculoperitoneal shunting had been done for aqueductal stenosis and hydrocephalus.

Child's mother and father were of South-Indian origin, aged 19 and 24 respectively. There was no evidence of consanguinity. There was also no significant family or antenatal history.

The child was delivered at term following a caesarian section. Birth weight was 2.45kg (<0.1 percentile) [3] and he was given phototherapy for 24-hours due to neonatal jaundice.

On general examination, patient appeared irritable, but there was no drowsiness or altered sensation. His length was 80cm (25^{th} percentile) and weight was 6.5kg ($<3^{rd}$ percentile) [3]. He was tachypnoeic and tachycardic. Coarse bilateral crepitations were also present. Extra-oral examination showed mild dysmorphic features, with retrognathia and a characteristic bird face appearance (*Figure 1&2*).

3. Treatment

Initial management was conservative, to resolve symptoms such as breathing and feeding difficulties. Patient

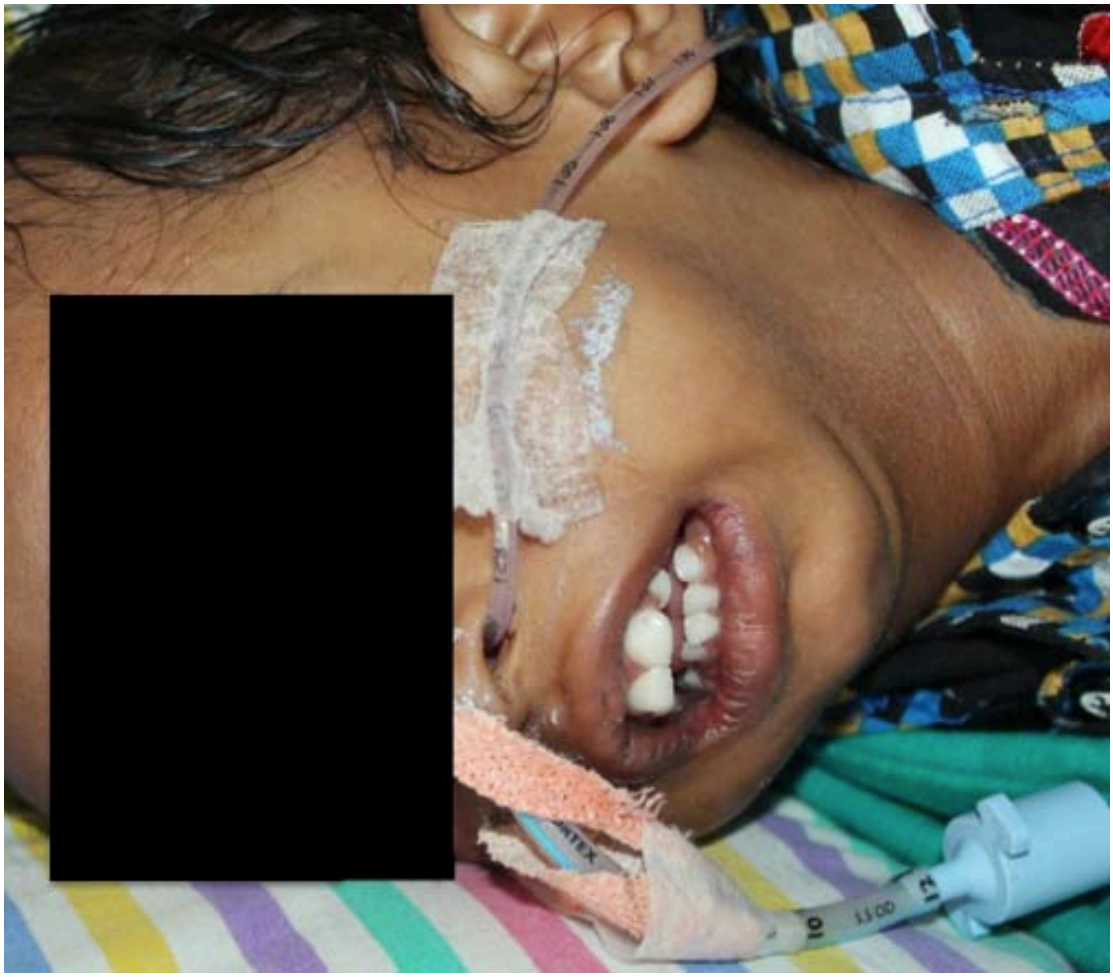

Figure 1. Retrognathia in the patient seen from a front view. A nasopharyngeal tube has been used to keep the airway patent.

was placed in prone position to allow gravity to pull the tongue base anteriorly to relieve airway obstruction. Prone-positioning was maintained 24-hours every day. Following insertion of nasopharyngeal airway prongs, patient was intubated by the paediatrician. Nasogastric tube was then inserted to facilitate feeding.

As the initial management was only symptomatic, a definitive surgical management was indicated. A technique described by Lapidot and Ben-Hur was performed during the surgery. A needle was inserted through the posterior midline portion of the tongue base, and directed anteriorly and caudally to emerge from the inferior border of the hyoid. The opposite end of suture was tunneled through a spinal needle to the foramen caecum and then directed inferiorly, to emerge on the superior aspect of the hyoid bone. A small skin incision was made opposite the body of the hyoid bone. The two free ends of the suture were then tied at the front of the hyoid body. This helped pull the base of the tongue anteriorly, away from hypopharynx to relieve the airway obstruction.

4. Outcome and follow-up

The patient was then extubated one week post-operatively. His vitals were constantly monitored. He was placed in the lateral position as per the post-operative advice. Regular chest physiotherapy and suctioning helped in clearing secretions. To reduce the risk of infections, the mother, was taught hand-washing techniques to be followed before child-handling. She was also given appropriate feeding and child-positioning advice.

The preferred definitive surgical management for this patient initially was MDO. However, the next best alternative (the above-mentioned technique) was agreed on due to inability to afford the implants used in the MDO.

5. Discussion

PRS is a heterogenous condition that may be associated with syndromes, resulting in varied presentations. Hence, the consensus in management remains elusive, with no definitive treatment protocols [4]. Management protocol

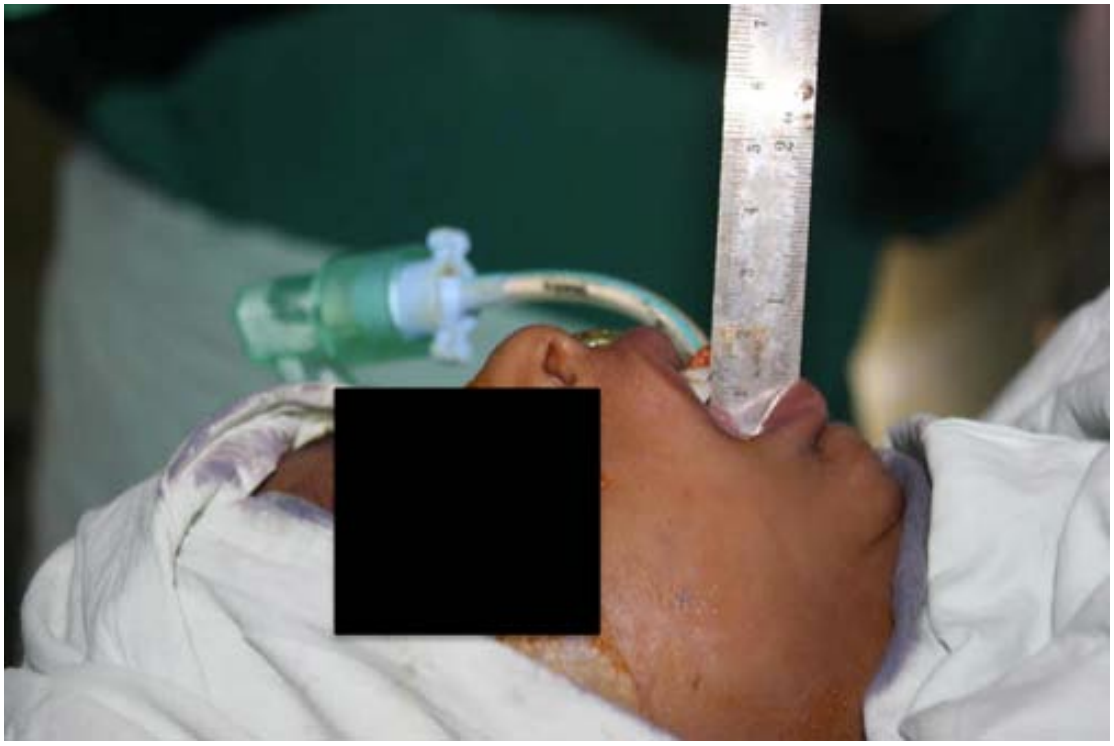

Figure 2. Retrognathia in the patient seen from a lateral view. The mandible was retropositioned 2cm relative to the maxillas.

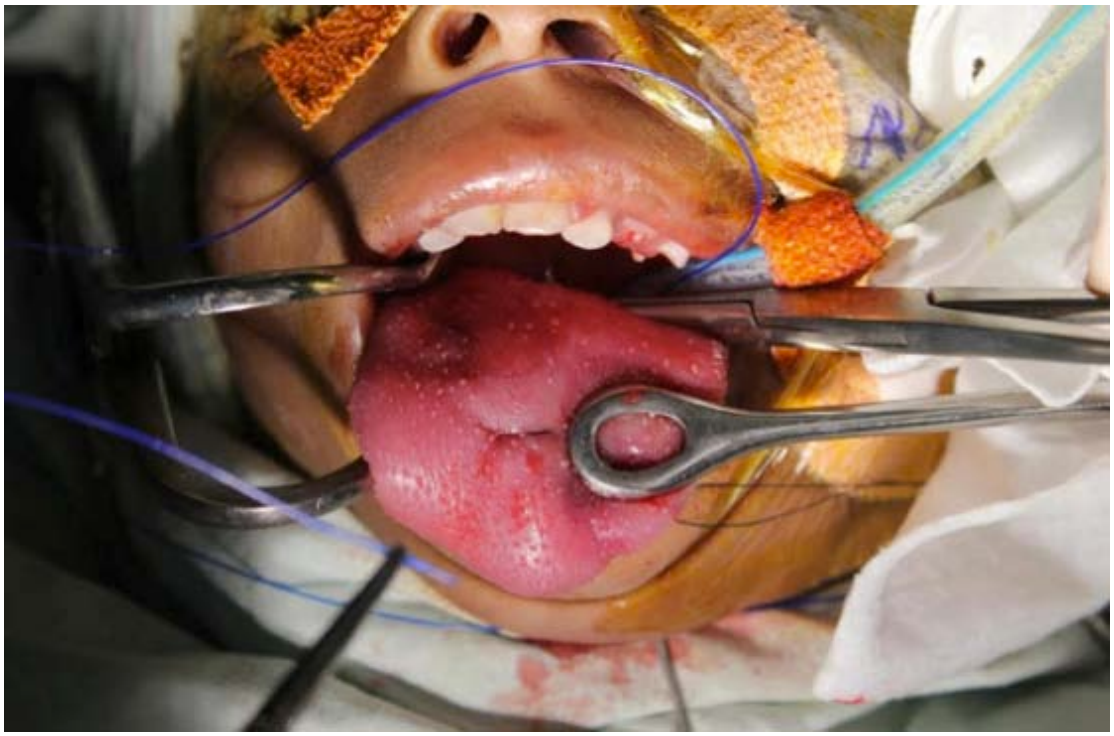

Figure 3. An intra- operative image showing the insertion of spinal needle at the level of hyoid bone for insertion of the suture from the foramen caecum to level of hyoid bone. Tongue is being held by the suture for accessibility.

should begin with the least invasive treatment and then progress to more invasive intervention until the patient is effectively managed.

First-line management is non-surgical, involving prone

positioning, laryngeal mask, nasopharyngeal stenting, and intubation. For feeding difficulties, modifications to the nipple of the bottle, nasogastric feeding or gastrostomy may be performed. However, in some patients, nasopharyngeal airways, endotracheal intubation or NG tubings may not be sustainable or tolerated. Management of these devices may also be overwhelming for the caregivers.

The next line of treatment is surgical. The common surgical procedures used to relieve airway obstruction in PRS include TLA, MDO, and tracheostomy.

TLA corrects glossoptosis by pulling the base of the tongue forward to maintain a stable airway. It does not treat the mandibular hypoplasia but success depends on the concept of catch-up mandibular growth [5]. After adequate growth, the TLA must be released with a second procedure. The success rate at managing airway obstruction is as high as 89% in selected patients [6]. TLA lacks long-term scarring and is cheaper due to lack of specialised equipment.

The techniques described include the Douglas' Technique [7], Argamaso Technique [8] and the Lapidot and Ben-Hur Technique [9].

Douglas's technique involves a rectangular area denuded under the tongue, along with the floor of the mouth on the alveolus and on the lower lip. The tongue is then brought forward and the raw surfaces are coapted. Once healed, the mucosal attachment serves to tether the tongue anteriorly until infant develops a stable airway with growth. This technique is usually performed on infants who have not developed any lower teeth, as they could otherwise bite though the repair inadvertently.

Argamaso technique involves the detaching the genioglossus from the mandible. The tongue based flap is then sutured to the lower side of the opposing defect in the mucosa of the lower lip.

Lapidot and Ben-Hur technique is a modification of TLA and was the preferred technique for this patient. This involves fastening the base of the tongue to the hyoid bone. It was preferred to the Douglas' technique as the latter was thought to restrict mobile segments of the tongue, hindering the growth of the mandible. In addition, since the patient had growth of lower teeth, Douglas technique may have failed due to inadvertent nibbling or biting of the sutures. Given the age and the nutritional status of the patient, we predicted that the dissection involved in the Argamaso technique would not be tolerated. Complications in Douglas' and Argamaso also delayed speech development and feeding problems due to tongue movement restrictions.

MDO of the mandible is the process in which the mandible is distracted, lengthening the jaw anteriorly. This pulls the tongue base anteriorly through muscle attachment to the mandible. Distraction progresses slowly and the related muscles, blood vessels, nerves, skin, and mucosa also elongate simultaneously. This concomitant expansion of the soft tissue is the main advantage of MDO [10]. However, complications include damage to developing molars, external scarring from implantation, injury to the marginal mandibular branch of the facial nerve and inferior alveolar nerve damage, which run close to the site of operation. It also involves high cost.

TLA serves as a temporising measure rather than definitive, unlike MDO. Both interventions are proven and valid techniques for addressing airway obstruction in selected infants. The advantages and risks of either should be weighed in view of the patient's age, nutritional level, tolerance to the procedure and financial status. The cost was the primary reason for the patient in the above-mentioned case to be not treated with MDO.

When all the above-mentioned interventions fail in a patient, the definitive management will be tracheostomy. However, complications include airway infections, bleeding, tracheal stenosis, inhibition of speech and swallowing and compromised social interactions [10]. Monitoring, equipment maintenance, and tracheostomy wound management require skilled nursing care at home, which may not be managed by many families.

Micrognathia or retrognathia can be identified prenatally through ultrasound. Prognosis is better when reconstructive surgery of the mandible is performed in newborns [11]. Prognosis is usually good but depends on presence or absence of other syndromes and their complications.

Due to complications associated with any of these treatment choices, adequate monitoring and follow-up will be necessary. Follow-up will be required to ensure that child is able to tolerate an oral diet and that speech development is not significantly affected.

Overall, this case highlights a surgical technique not widely described in the literature for treatment in PRS. While gold-standard treatments may be the preferred options, it is important to realise that it is not possible to employ them in all situations (e.g. lack of resources, inability of patient to afford etc.). Hence, it is important for surgeons to be aware of other viable techniques that may be used under special circumstances.

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