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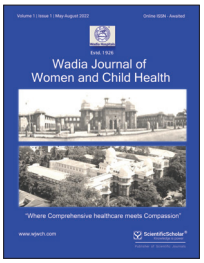

Case Series

Nutritional rehabilitation in children with Pierre Robin syndrome

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ABSTRACT

Pierre Robin Syndrome (PRS) or sequence is a common craniofacial abnormality characterised by micrognathia, which causes glossoptosis and upper airway obstruction. This causes feeding difficulty resulting in severe malnourishment. Cleft plate is also commonly seen in this condition. Here, we present three cases less than 6 months of age with PRS and concurrent nutritional compromise. Common presenting symptoms included respiratory distress, stridor, cyanosis, and poor growth. In addition, caregivers had difficulty in feeding these children, resulting in severe acute malnutrition requiring hospitalization. These children had to undergo early surgical intervention in the form of tongue lip adhesion and jaw advancement procedures to ensure better airway resolution and nutritional outcomes. They were, then, started on F-75 formula with improved methods of feeding as per severe acute malnourishment under 6 months protocol, which resulted in adequate weight gain and good overall outcome.

Keywords: Pierre Robin sequence, Severe acute malnutrition, Cleft palate, Mandibular distraction osteogenesis, Tongue-lip adhesion

INTRODUCTION

Pierre Robin Sequence (PRS) is a common craniofacial abnormality characterized by micrognathia, glossoptosis, and airway obstruction. This commonly leads to feeding difficulty (FD), which leads to children becoming severely malnourished. A cleft palate (CP) may be associated with this condition, further hindering nutritional outcome. The management of PRS has been revolutionized by the use of tongue-lip adhesion (TLA) and mandibular advancement surgery. This helps not only in maintaining the airway until definitive correction but also helps in the nutrition of the child and leads to early correction of the CP if present.

CASE SERIES

Case 1

A 4-month-old baby girl, was admitted with recurrent episodes of respiratory distress, stridor and occasional cyanosis. She was a term baby with a good birth weight of 2.7 kg. However, she had a history of admission to the neonatal intensive care unit (NICU) with similar complaints

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in the neonatal period, requiring mechanical ventilation. Clinical findings included a CP and micrognathia with suprasternal retractions on examination, pointing to a possibility of anatomical airway compromise. Subsequently, she was admitted for similar complaints twice over 2 months. The child was never breastfed and was given inadequate formula feeds of improper dilution.

In the current admission, the child was diagnosed with aspiration pneumonia and required non-invasive positive pressure ventilation along with broad-spectrum injectable antibiotics. Bronchoscopy revealed type 3 laryngomalacia. She was planned for TLA for the significant retrognathia, significant airway compromise, and failure to gain weight. Anti-reflux medication was started as an adjunctive therapy. The FD was tackled using an infant feeding tube. Medical nutrition therapy, that is, F-75 formula feeds, was initiated according to severe acute malnutrition (SAM) protocol at 130 mL/kg/day. The feed volume was increased gradually to 200 mL/kg/day. However, the child continued to have regurgitation with prolonged oxygen dependency, for which a mandibular jaw advancement procedure was done. There was significant improvement post-surgery with gradual reduction in oxygen requirement to omission. The child also displayed weight gain of 7.5 g/kg/day for the duration of their stay in the nutritional rehabilitation center.

Case 2

A 5-month-old baby boy was admitted with complaints of noisy breathing, FD, and failure to thrive. On examination, the baby had SAM with weight for length < - 4 SD. He was born at term with a birth weight of 3 kg with subsequent FD, for which he was under medical attention for 15 days. However, he was then started on bottle feeds, which he was unable to tolerate because of an underlying cleft palate.

Airway assessment revealed type 3 laryngomalacia through direct laryngoscopy. Anti-reflux medications were started and TLA was performed as retrognathia was a contributory factor for the FD. Post-operatively the child was nutritionally rehabilitated by starting F-75 feeds as per the SAM protocol at 130 mL/kg/day. As per tolerance, the volume of the feeds was increased. There was a weight gain of 9.5 g/kg/day, and the stridor and suprasternal retractions decreased.

Case 3

A 3-month-old male child with a normal term delivery and a birth weight of 2.5 kg was admitted with PRS with significant retrognathia and associated CP. These complaints led to difficulty in feeding, resulting in inadequate weight gain since birth.

On examination, the child was cachectic and was found to have crepitations on the right side on chest auscultation. Chest x-ray showed features of aspiration pneumonia, for which the child was given high flow oxygen, intravenous antibiotics, and nebulization. After respiratory stabilization, tube feeds were started. TLA was done to improve the baby's breathing and feeding. The child was continued on tube feeds of F-75 formula, as per SAM protocol. A pH manometry, done for recurrent arching episodes and intermittent regurgitation, was suggestive of type 2 gastroesophageal reflux (GER) disease. Hence, anti-GER medicines and thickened feeds were started. He was weaned off oxygen support and had a weight gain of 8.6 g/kg/day observed during the ward stay [Figure 1].

DISCUSSION

FD and malnutrition is commonly seen in PRS. Feeding is an important step when it comes to handling severe malnutrition; however, it must be started in a small frequent amount as per tolerance. A starter diet or the "starter" formula is used for initial feeding and stabilizing the malnourished child. It contains 75 kcal and 0.9 g protein per 100 ml.[1] It is low in protein and sodium and high in carbohydrates, providing adequate glucose to prevent hypoglycemia. Stabilizing the child usually takes 2–7 days, after which a catch-up diet is initiated to rebuild wasted tissues. A catch-up diet has more calories and protein of 100 kcal and 2.9 g protein per 100 ml, as compared to a starter diet.[1]

FD is a common symptom in PRS, which leads to failure to thrive and developmental problems if not recognized and treated on time.[2] Up to 73% of infants with a CP have also been reported to suffer FD.[3] Furthermore, associated genetic abnormalities and growth factor defects might lead to growth retardation in PRS.[4] In addition, airway infections during 0–3 months of age can negatively affect growth.[5] Other origins of feeding disorders and growth retardation in patients with PRS also exist, such as primary brainstem dysfunction, or neuromotor disabilities, which are seen more in patients with PRS than those with only CP.[2] Furthermore, several other symptoms are seen which accentuate the problem such as dysphagia, vomiting, and/or GER.[2,6] If there is poor weight gain, nasogastric tube feeding should be started with ideal medical nutrition therapy according to the SAM protocol.[1] If one is suspecting GER, reflux therapy should be started, as the incidence of GER is observed to be higher in PRS.

TLA is a procedure where the tongue is attached anteriorly to the lower lip and opens the oropharyngeal airway space as the tongue base is pulled forward.[7] After the procedure, the patients start showing clinical improvement, with weight gain and decreased rate of respiratory infections.

The duration of nasogastric feeds has been found to reduce significantly after mandibular distraction osteogenesis (MDO) when compared to the other surgical

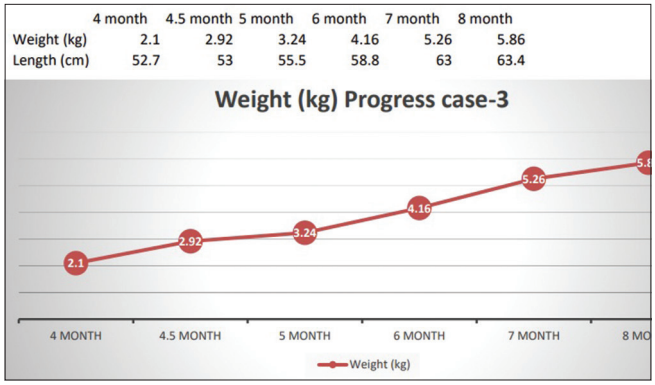

Figure 1: Case 3 nutritional recovery post tongue lip adhesion.

interventions.[8] However, MDO is indicated, only if TLA alone does not improve feeding and airway obstruction.

CONCLUSION

This case series highlights the surgical techniques used in PRS at our institute and the significant post-procedural benefits. FD is known to affect growth, maternal bonding, and social and cognitive development. PRS patients have significant GER and FD causing failure to thrive. Therefore, the right surgical technique and nutrition to overcome FD post-surgery, sensory stimulation, play therapy, speech therapy, and proper counseling of parents are required.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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