Pierre Robin Syndrome

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Abstract
A single live female baby was born with retroglossalia, micrognathia, cleft palate and glossoptosis was followed up till eight months of age, with healthy growth and development on orogastric tube feeds, thus preventing repeated choking episodes and aspiration pneumonia proving to be an excellent alternative in care of children despite other methods available in children with cleft palate and glossoptosis till the age for surgical closure of cleft palate.

Keywords
Pierre Robin Syndrome; Feeding Practice with Orogastric Tube; Feeds for Healthy Growth

Introduction
Pierre Robins syndrome has three main features of micrognathia, macroglossia and cleft palate first described by Pierre Robin a French dental surgeon in 1923 [1] is a chain of deformation sequence with a basic cause being failure of lower jaw to develop normally at about 7-10 weeks of pregnancy thus positioning the tongue between the two halves of the palate, preventing the palate from closing result in U-shaped soft palate cleft or V-shaped, soft and hard palate cleft. The small oral cavity positions the tongue at the back of the mouth causing breathing difficulty by obstructing the airway passage at birth [2]. Gastroesophageal reflux is more prevalent and reflux of acidic contents in the posterior pharynx worsens airway obstruction [3], hence upright prone or side lying position with frequent small feeds minimizes vomiting and use of pharmacotherapy such as proton pump inhibitors.

Case Report
A single life girl baby was born by elective caesarean section at 36+3 weeks gestation to 35 year old, fifth gravida mother with three living children all males and one previous abortion with pregnancy complicated by gestational diabetes mellitus and polyhydraminos. Baby established spontaneous respiration at birth. Apgar score was 8 at 1min. and 9 at 5 min. Birthweight was 2800g. On examination was found to have retroglossalia, micrognathia, glossoptosis and cleft palate as seen in Figures 1, 2 and 3 showing lateral, anterior and antero-lateral views and Figure 4 showing cleft palate. Baby was shifted to NICU and kept in prone position with no episodes of apnea or cyanosis. Parenteral fluids and oxygen by hood at 5 liters/min was instituted for first 24 hours. Nasopharyngeal tube feeds was then started.

The baby developed gastro-oesophageal reflux with arching and was treated in side lying position, with proton pump inhibitors and small frequent feeds to prevent vomiting as seen in Figure 5. Subsequently oral feeds with long spout cup (pallada) feeds were initiated, however after bronchopneumonia. The baby was put back on orogastric tube feeds and sent home, the mother taught how to give tube feed baby with monthly follow up to replace orogastric tube and to monitor growth and
development, thus preventing repetitive broncopneumonia in the first months of life related to swallowing impairment with cleft palate. The baby thrived, gained weight and at eight months of age was healthy with adequate weight gain with normal development was playful, alert and active as seen in Figures 6 and 7.

**Figure 1**: Anterior View

![Figure 1: Anterior View](image1)

**Figure 2**: Lateral View

![Figure 2: Lateral View](image2)

**Figure 3**: Antero-Lateral View

![Figure 3: Antero-Lateral View](image3)

**Figure 4**: Cleft Palate

![Figure 4: Cleft Palate](image4)

**Figure 5**: Gastro-Oesophageal Reflux with Arching

![Figure 5: Gastro-Oesophageal Reflux with Arching](image5)

**Figure 6**: Follow-Up at 7 Months

![Figure 6: Follow-Up at 7 Months](image6)

**Figure 7**: Follow-Up at 8 Months

![Figure 7: Follow-Up at 8 Months](image7)

**Discussion**

Pierre Robin (pronounced Roban) syndrome (PRS) is also known as Pierre Robin sequence, Pierre Robin triad or Pierre Robin anomaly. In some patients “Robin sequence or Robin Complex” may be more appropriate. The pathogenesis of the syndrome is attributed to mechanical compression of the mandible, genetic growth disturbance, teratogen exposure, and growth arrest due to an in utero insult.

The basic cause appears to be the failure of the lower jaw to develop normally before birth. At about 7-10 weeks into a pregnancy, the lower jaw grows rapidly, allowing the tongue to descend from between the two halves of the palate. If, for some reason, the lower jaw does not grow properly, the tongue can prevent the palate from
closing, resulting in a cleft palate. The small or displaced lower jaw also causes the tongue to be positioned at the back of the mouth, possibly causing breathing difficulty at birth [2]. This “sequencing” of events is the reason why the condition has been classified as a deformation sequence. PRS is not generally diagnosed by ultrasound before birth as profile view of fetus is difficult to achieve [6]. This congenital anomaly with deformative sequence is autosomal recessive disorder with incidence of 1 in 80000 to 1 in 200000 live births [4]. Parents with one child having isolated Robin complex have a 1-5% chance of having another child with this condition. A greater incidence in girls is noted as palate takes approximately one week longer to fuse.

In some Robin Sequence/Complex is observed with Stickler syndrome, Velocardiofacial syndrome or Treacher Collins syndrome, thus in addition to genetic predisposition, environmentally induced tetraogenic syndromes such as Fetal alcohol syndrome and fetal hydantoin syndrome will in influence frequency of occurrences [4, 5]. Hence through evaluation should be done by geneticist. There is no known prevention during intrauterine period as radiological diagnosis is difficult. It is important to make a diagnosis soon after birth as in supine position with glossoptosis baby may develop breathing problems with respiratory distress and cyanosis requiring intubation with nasopharyngeal airway (NPA). Prone position prevents obstructive apnoea as tongue falls forwards aided by gravity. Cleft palate may cause aspiration with choking episodes while feeding with recurrent bronchopneumonia, may cause hypoxia with brain damage, congestive cardiac failure, pulmonary hypertension and death [6].

Airway obstruction soon after birth requiring a nasopharyngeal tube should alert the clinician to the diagnosis of Pierre Robin Syndrome and management is aimed at reducing breathing problems, choking aspiration pneumonia, by placing baby in prone or lateral position and adequate nutritional support of baby. Orogastric tube feeds prevent these complications in Pierre Robin sequence and adequate nutrition for normal neurodevelopmental outcome and physical growth. However other feeding practices are recommended such as bottle feeding and long cross cut nipple with slight pressure at the angle of mandible to compress nipple for better flow of milk with frequent burping advised.

Thus effective management without resort to surgical intervention during infancy as outlined in this case with orogastric tube feeds proved to be an excellent alternative in home care, the mother taught how to tube feed her child, with regular follow-up for replacement of orogastric tube [7]. This management proved to be effective in reducing breathing problems, choking and recurrent bronchopneumonia with hypoxia, cyanosis, and adequate nutritional support of babies in prone or side lying position. However caretakers should be aware of risks of tube feeding with knowledge of tube dislodgement, to come immediately to the hospital for proper insertion, also feeding tube needs to be changed regularly.

Paediatric ear, nose, throat specialist need monitor baby for ear disease as cleft palate predisposes fluid collection in the middle ear that may require drainage with ventilation tubes. Frequent ear infections may cause temporary loss of hearing that can affect speech and language development requires follow-up by an audiologist. Obstructive apnoea’s due to craniofacial anomalies with respiratory sleep symptoms and daytime sleepiness need evaluation to determine the sites and severity of obstruction, as well as systemic impairment [8-10]. Airway obstruction and related hypoxia, carries a high mortality risk in Pierre Robin sequence. In some cases jaw distraction is needed to aid breathing and feeding. Lip-tongue attachment is performed in some centres, though its efficacy has recently been questioned. However orogastric tube and lateral position circumvented early surgical intervention.

In most children with isolated PRS, the mandible grows rapidly during the first year and achieve near normal mandibular size by four to six years of age with almost normal profile. Cleft palate is generally repaired between ages by 1 to 2 years by a plastic or maxillofacial surgeon. Delayed or defective speech development is addressed by a speech and language therapist, an orthodontist and sometimes psychologist and nursing staff.

Conclusion

The primary management of Pierre Robin Syndrome for airway insufficiency due to glossoptosis with micrognathis is early diagnosis and placing infant in prone or lateral side lying position with orogastric tube feeds to prevent choking, aspiration and frequent bronchopneumonia due to cleft palate. This effective management resulted in good prognosis with healthy growth and development of infant till one year of age when surgical closure of cleft palate may be undertaken for normal feeding. Thus orogastric tube feeds avoided other
interventions resulting in good prognosis with healthy growth and development into normal adult life.

References


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