

# Pierre Robin Sequence as Birth Asphyxia in a New Born

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## ABSTRACT

A new born baby boy presented with birth asphyxia and respiratory distress. He went into cardiac arrest twice but was resuscitated. On detailed evaluation, he had low set ears and micrognathia with glossoptosis consistent with features of Pierre Robin sequence. Episodes of apnoea disappeared, on nursing, the baby in prone position and later on tongue-lip retention suture were applied. Prompt diagnosis and efficient airway management by following the principles of airway resuscitation can save the lives of such babies without specialized care.

**Key words:** *Micrognathia. Glossoptosis. Birth asphyxia. Respiratory distress.*

## INTRODUCTION

Pierre Robin syndrome was first described on 2 patients with micrognathia, cleft palate, and retroglossoptosis by Lannelongue and Menard in 1891. Pierre Robin published the case of an infant with complete syndrome in 1926. Until 1974, the triad was known as Pierre Robin syndrome but it is a sequence of events arising from the poor development of mandible. Small lower jaw (micrognathia) causes the tongue to ball-up at the back of mouth (glossoptosis) which interferes with the closure of the palate resulting in horse shoe-shaped cleft palate. The above sequence of anomalies causes breathing problems with a clinical presentation of asphyxia and respiratory distress at birth.<sup>1</sup> One such case is hereby reported.

## CASE REPORT

A 32-year-old unbooked multigravida presented at term in labour. She had unremarkable previous obstetric history with 5 healthy live issues delivered at home. She did not have any past history of uterine fibroids, infectious disease or rash during pregnancy. A male baby was delivered through spontaneous vertex delivery that went into respiratory arrest immediately after birth. The baby had spontaneous respiration at the time of delivery but went into apnoea and became cyanosed after being nursed in supine position. He had an Apgar score of 2/10 and was promptly resuscitated. Shortly, thereafter, he developed another episode of respiratory

distress, cyanosis and cardiac arrest. Active resuscitation revived the cardio-respiratory activity. However, his recurrent respiratory distress necessitated the use of oropharyngeal airway (Guedel's airway).

Subsequent detailed clinical examination revealed a poorly developed lower jaw and retracted chin. Examination of the oral cavity revealed posteriorly placed tongue and high arched palate. Infact, the rolled - up tongue was creating difficulty in maintaining a patent airway and placement of the oropharyngeal airway (Figure 1). Due to micrognathia, glossoptosis and high arched palate in a neonate, a provisional diagnosis of Pierre Robin sequence was made and the baby was nursed in prone position with Guedel's airway for patency of airway. Glossopexy with tongue lip retention suture was applied the next day, using 2/0 silk (Figure 2).



Figure 1: Micrognathia with oropharyngeal airway in situ.



Figure 2: Tongue-lip retention suture (antero-posterior and lateral view) .

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The baby started feeding normally without any further episode of respiratory arrest or distress. The mother was trained to nurse the baby in prone position and parents were counselled about the benign nature of the condition.

Further detailed investigations including 2D-echo, chest radiograph and plasma glucose levels of the baby failed to reveal any other congenital defect or anomaly. Anti-rubella antibodies were found positive in high titres in the mother. The child was followed-up after 4 weeks, which showed adequate growth of mandible and retention suture was removed. The child made uneventful recovery and continued normal feeding without any respiratory disturbances.

## DISCUSSION

Pierre Robin Sequence (PRS) is a rare condition with an incidence of 1 in 8,000 to 30,000 live births and a male to female ratio of 1:1.<sup>1</sup> Prompt diagnosis and efficient airway management by following the principles of airway resuscitation can save the lives of these babies without specialized care, which is also the purpose to highlight this case.

PRS has been referred to with various names like "Pierre Robin Malformation Sequence", "Robin Anomalad", and "Cleft Palate, Micrognathia and Glossoptosis." The etiology of PRS is not clear. Apart from hereditary factors, maternal virus infection in the early stages of pregnancy and folic acid deficiency are other areas that have been researched and nothing conclusive has been determined.<sup>1</sup>

Retraction of the inferior dental arch 10-12 mm behind the superior arch known as micrognathia is reported in the majority of cases (91.7%). The growth of the mandible catches-up during the first year and the child attains a normal profile by approximately 5-6 years of age. Glossoptosis is noted in 70-85% of cases. Macroglossia and ankyloglossia are uncommon findings seen in 10-15% cases. The prevalence of cleft palate varies from 14-91% and affects the soft and hard palate. These features of micrognathia and glossoptosis can also be detected on ultrasound during pregnancy.<sup>2</sup>

Patients with PRS may demonstrate various paediatric conditions like otic abnormalities, nasal deformities, laryngomalacia, speech defects, cardiovascular findings (valvular and septal abnormalities), gastroesophageal reflux, oesophagitis, musculoskeletal/central nervous system and genitourinary defects.<sup>3</sup>

PRS babies have posteriorly displaced chin which causes the tongue to fall backwards towards the posterior pharyngeal wall. This results in obstruction of the airway on inspiration. Crying or straining by these children can often keep the airway open. However, when the child relaxes, lies supine or falls asleep, airway

obstruction occurs. Due to these respiratory problems, feeding may become very difficult. The majority of these patients can be managed by placing the infant in the prone position until adequate growth of the jaw occurs. This causes the jaw and the tongue to fall forward, opening the airway. The use of nasopharyngeal tubes is recommended initially to alleviate the immediate consequences of hypoxia.<sup>4</sup> The use of simple oropharyngeal airway can also be a useful temporary measure to keep the airway patent. Surgical intervention is required in infants with severe respiratory distress.<sup>5</sup>

Infants may suffer significant complications, including failure to thrive, chronic hypoxemia, carbon dioxide retention and cor pulmonale, if left untreated. Deaths related to PRS are not as frequent as were reported in the early 20th century but presently reported mortality from the condition is at 20%.<sup>6</sup>

In a recent survey, 91% of paediatric otolaryngology fellowship programs cited tracheostomy as the safest and the most reliable surgical method for long-term airway management in infants with PRS.<sup>3</sup> According to Kirschner *et al.* the overwhelming majority of infants who fail to respond to positioning alone can be safely and effectively managed by a properly performed tongue-lip adhesion. Glossopexy is a useful procedure in the management of upper airway obstruction. It may vary from simple tongue-lip retention suture to formal tongue-lip adhesion techniques. The former is a temporary procedure, which may last for 2-3 weeks and the mother also learns how to nurse the child efficiently. Glossopexy may temporarily affect the development of pre speech skills and sound production. The onset of babbling and initiation of first words are often delayed in the subjects in comparison with syndrome-matched children who had not undergone glossopexy.<sup>8</sup> Subperiosteal release of floor of mouth is also an effective surgical procedure in infants with PRS in order to correct severe upper airway obstruction.<sup>9</sup> The age at surgery is preferably 2 to 13 weeks. Recently, early mandibular lengthening in infants with PRS by distraction osteogenesis has been advocated for infants with PRS, who demonstrate persistent obstructive apnea despite tongue-lip adhesion.<sup>10</sup> Considering the diversity of clinical features in these children, a multidisciplinary approach is essential in follow-up of these babies.

## REFERENCES

1. Marques IL, Barbieri MA, Bettio H. Etiopathogenesis of isolated Robin sequence. *Cleft Palate Craniofac J* 1998; **35**:517-25.
2. Bronshtein M, Blazer S, Zalel Y, Zimmer EZ. Ultrasonographic diagnosis of glossoptosis in fetuses with Pierre Robin sequence in early and mid pregnancy. *Am J Obstet Gynecol* 2005; **193**:1561-4.
3. Gruen PM, Carranza A, Karmody CS, Bachor E. Anomalies of the ear in the Pierre Robin triad. *Ann Otol Rhinol Laryngol* 2005; **114**:605-13.

4. Singer L, Sidoti EJ. Pediatric management of Robin sequence. *Cleft Palate Craniofac J* 1992; **29**: 220-3.
5. Myer CM, Reed JM, Cotton RT, Willging JP, Shott SR. Airway management in Pierre Robin sequence. *Otolaryngol Head Neck Surg* 1998; **118**: 630-5.
6. Benjamin B, Walker P. Management of airway obstruction in Pierre Robin sequence. *Int J Pediatr Otorhinolaryngol* 1991; **22**: 23-7.
7. Kirschner RE, Low DW, Randall P, Barlett SP, McDonald-McGinn DM, Shultz PJ, *et al.* Surgical airway management in Pierre Robin sequence: is there a role of tongue-lip adhesion? *Cleft Palate Craniofac J* 2003; **40**:13-8.
8. Argamaso RV. Glossopexy for upper airway obstruction in Robin sequence. *Cleft Palate Craniofac J* 1992; **29**: 232-8.
9. Caouette-Laberge L, Plamondon C, Larocque Y. Sub-periosteal release of the floor of the mouth in Pierre Robin sequence: experience with 12 cases. *Cleft Palate Craniofac J* 1996; **33**:468-72.
10. Denny AD, Talisman R, Hanson PR, Recinos RF. Mandibular distraction osteogenesis in very young patients to correct airway obstruction. *Plast Reconstr Surg* 2001; **108**: 302-11.

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