Postural management of airway obstruction secondary to Pierre Robin Sequence

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ABSTRACT

Pierre Robin Sequence commonly presents with upper airway obstruction secondary to craniofacial anomalies and can be managed with several treatment modalities. In mild cases, non-invasive postural management is a good treatment option. Prone position has been established as the posture of choice to reduce potential upper airway obstruction. We report a case of Pierre Robin Sequence with airway obstruction that did not respond to prone position but responded to supine position with slight tilting of the head.

Keywords: Pierre Robin Sequence, respiratory distress syndrome, supine position

INTRODUCTION

Pierre Robin Sequence (PRS) is a congenital condition with anomalies that include micrognathia, glossoptosis and cleft palate. [1] As a result of the small chin, the tongue tends to fall backward and gets trapped between the cleft palate. This can lead to significant upper airway obstruction when the child is lying in the supine position. Therefore, management includes placement in a prone position. Nevertheless, invasive procedures such as distraction osteogenesis or tracheostomy may be required in more severe cases that fail to respond to postural management. We report a case of PRS with airway obstruction that was successfully managed in a modified supine position after failing to respond favourably in prone position.

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CASE REPORT

A newborn male infant without any remarkable antenatal history was delivered at full term via spontaneous vaginal delivery to a young couple (mother, 27 years old and father, 30 years old). Upon delivery, the child was active, vigorous and had a good cry. His body weight was 2.9kg and the Apgar score was nine at one minute. However, two minutes later, he developed grunting and tachypnoea. Physical examination revealed a baby boy with prominent eyeballs, small chin and a U-shaped cleft at posterior two-third of the

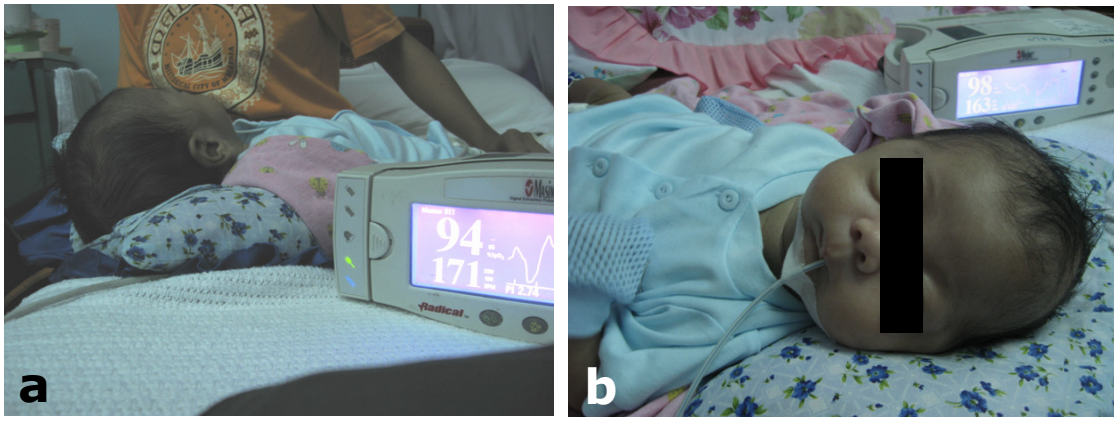

Fig. 1: a) showing desaturation with SpO_2 of less than 95% when placed in supine position, b) tilting of the head laterally improved the saturation to 98%.

palate. He was intubated for respiratory distress with 3.5mm endotracheal tube (ETT). He was breathing spontaneously and was maintaining good oxygen saturation with low ventilatory setting in the Neonatal Intensive Care Unit. During this period, there were two incidents of ETT dislodgements. On both occasions, the baby had stridor, tachypnoea and desaturation when nursed in both supine and prone positions, requiring reintubation. He was later referred to the Otorhinolaryngologist for further assessment.

Direct laryngoscopy performed on the 26th day of life without assisted ventilation showed that the posterior part of the tongue was in contact with the posterior pharyngeal wall. The epiglottis could not be visualised due to the glossoptosis. Post endoscopy, the infant exhibited respiratory distress when he was placed on his back (Figure 1). However, this was relieved when his head was titled laterally (Figure 2). He was managed conservatively using this position as the oxygen saturation could be maintained at greater than 95%.

The condition of the child was explained to the parents and the mother was educated on the position maneuver to maintain airway patency. The child was discharged well at one week and a reassessment three months later revealed a healthy and well thriving child.

DISCUSSION

PRS was first reported by Pierre Robin, a French stomatologist who described the clinical triad of micrognathia, glossoptosis and a U-shaped cleft palate. [2] The most widely accepted incidence reported is 1:8500 live-births. [3] During embryogenesis, mandibular hypoplasia is believed to cause the high position of tongue leading to the failure of fusion of the palatal shelves resulting in cleft palate. [3] Due to the retrognathic mandible, the tongue is prone to fall backward and obstruct the upper airway causing stridor, respiratory distress, feeding difficulties, failure to thrive, aspiration pneumonia and even death in severe cases.

Upper airway obstruction in PRS can be classified based on endoscopic findings examination (Table 1). [3] Benjamin and Walker in 1991 proposed another classification that

Table 1: Classification of PRS upper airway obstruction based on endoscopic features. [3]

Types	Features
I	Posterior movement of the tongue contacting the posterior pharyngeal wall
II	The tongues moves posteriorly and compresses the soft palate or cleft palate posteriorly against the posterior pharyngeal wall so that there is a junction of the tongue, velum and posterior pharyngeal wall in the upper portion of the oropharynx. This junction obstructs the nasopharyngeal airway
III	The lateral pharyngeal walls move medially, opposing one another
IV	The pharynx constricts in a sphincteric manner

categorises PRS according to the manage–ment of the airway (Table 2). [4] In our case, the baby had all the typical features of PRS, micrognathia, glossoptosis and cleft palate. Our patient had type I upper airway obstruction according to Sher *et al*. and mild obstruction by the criteria of proposed by Benjamin and Walker.

In cases of mild upper airway obstruction, adopting a prone position to alleviate airway obstruction relies on the effect of gravity to bring the tongue forward in order to increase the pharyngeal space. [1] It is suggested that this position be maintained for 5-6 months to allow for spontaneous resolution of airway obstruction. This can occur with growth and development of the mandible, neuromuscular structures of the tongue and pharyngeal airway. [1, 4] Meyer *et al*. reported that only 38 out of 74 patients with PRS required airway interventions other than prone position. [2] However, others feel that this position may hinder the observation for signs of airway obstruction. [3] Importantly, the risk of 'Sudden Infant Death Syndrome' (SIDS) is increased in premature infants sleeping in the prone position. However 40% out of 224 neonatal units surveyed in the United Kingdom (with 81% response) still recommended non-supine sleeping position for infants with PRS or gastro-oesophageal reflux. [5] We also managed our patient initially in the prone position. However, he developed respiratory distress. We then tried the supine position, but with slight head tilting; the airway patency improved significantly.

Head rotation is reported to augment the anteroposterior distance and cross-sectional area in the retroglossal region significantly and promote upward displacement

Table 2: Classification of PRS upper airway obstruction based on management. [4]

Types of obstruction	Management
Mild	Airway obstruction resolves with posture
Moderate	Nasopharyngeal airway is used and prone position after removal of nasopharyngeal tube
Severe	Long-term management with nasopharyngeal or tracheostomy

of the airway in the neck. [6] With the head rotated, the pyriform sinus on the rotated side was compressed whereas the contra-lateral side was in a wide funnel shape. [6] Therefore, head rotation not only reduced the airway obstruction, it also reduced the incidence of aspiration. Interestingly, Logemann *et al.* also had reported that head rotation during swallowing could reduce the occurrence of aspiration in dysphagic patients. [7] Patients with PRS should be reviewed frequently to assess their growth and for symptoms of airway compromise. Our patient was nursed for six months in the supine position with regular follow up.

Apart from postural management, cases with more severe upper airway obstruction may require other intervention modalities. These include the use of nasopharygeal airway or endotracheal intubation to surgical intervention such as tongue-lip adhesion or glossopexy, mandibular distraction and tracheostomy. [2] However all these modalities are not without inherent complications.

In conclusion, non-invasive measures such as postural management should be tried in PRS with airway obstruction as success rate as high as 74% have been reported. [1, 2] In our case, the infant obviously benefited from the modified postural management. Tilting of the head helped to alter the dynamics of the upper aerodigestive tract resulting in the maintenance of airway patency. We suggest that a trial of postural management with or without modifications should be tried especially in those with mild airway compromise. However, close observation is required as these patients are at high risk of mortality. Surgical interventions should be reserved for more severe cases. To date, there has been no report of modified postural management for airway obstruction in PRS.

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