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Respiratory distress in Pierre Robin: successful use of pharyngeal tube

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Index words:
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Abstract

Background/Purpose: The study describes a safe and least aggressive method to resolve airway obstruction in children born with a Pierre Robin sequence (PRS).

Methods: In a retrospective study, we analyzed the assessment of airway obstruction at birth and for the following months. The definition of PRS was based on the anatomical anomaly triad cleft palate, micro/retrognathia, and glossoptosis with some degree of airway obstruction. We defined 3 categories of children depending on their difficulties of breathing or eating at birth.

Results: From 1984 to 2004, 48 children were born in our hospital with a diagnosis of PRS. There were 32 children with nonsyndromic PRS (nsPRS) and 16 with syndromic PRS (sPRS): respectively, 40% (13) and 32% (5) had slight respiratory and/or feeding problems; 26% (8) and 56% (9), isolated feeding difficulties; 34% (11) and 12% (2), severe respiratory and feeding problems. Pharyngeal tube was used in 8 children with nsPRS and in 2 with sPRS. Neonatal surgery was not necessary. Primary palatoplasty was performed at almost the same time as for the patients with isolated cleft palate.

Conclusions: Children born with PRS have a good prognosis at birth provided that adequate respiratory support is given using either positive airway pressure mask or pharyngeal tube.

Pierre Robin sequence (PRS) is characterized by cleft palate, micro/retrognathia, and glossoptosis. Glossoptosis is generally responsible for pharyngeal obstruction [1]. Because of these anatomical anomalies of the upper airway and/or a degree of brainstem immaturity that promotes apnea and hypoxia, these children are at high risk of severe respiratory insufficiency and prolonged hypoxia [2]. These abnormalities must be anticipated at birth to avoid chronic and unrecognized respiratory obstruction that leads to hypoxia and impaired mental status. Close follow-up is mandatory to prevent neurologic sequelae after prolonged or chronic hypoxia [2,3].

The severity of airway obstruction varies considerably in children with PRS. When upper airway obstruction is mild, children can be handled conservatively, being placed in a prone or lateral position. In severe upper airway obstruction, babies may have respiratory distress and hypoxemia [2,3]. Tracheal intubation can be very difficult and complicated with pneumothorax, subcutaneous emphysema, or death [3,4]. Conservative techniques include the use of a continuous positive airway pressure (CPAP) mask and the use of a laryngeal mask airway [5]. Surgical methods proposed are glossopexy [6,7], tongue-lip adhesion [8,9], tracheotomy, or mandibular traction [10].

Physiopathologic characteristic of airway obstruction because of anatomical anomalies involves posterior

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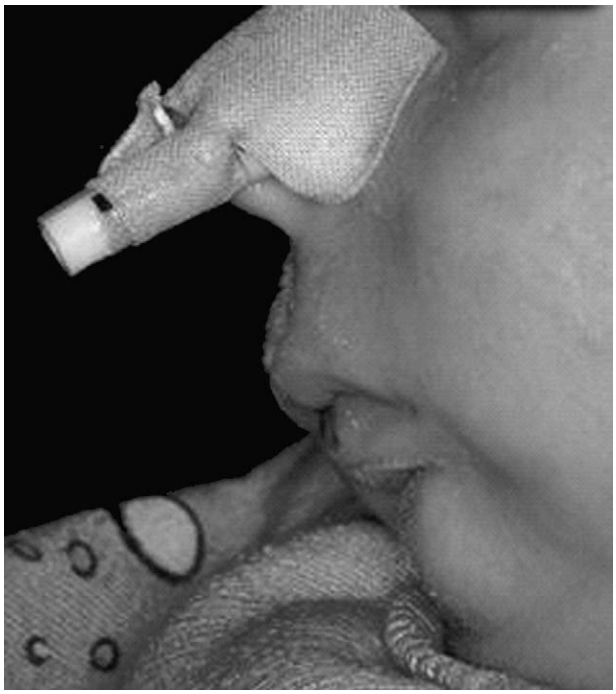

Fig. 1 Nasopharyngeal tube for the relief of airway obstruction.

retraction of a normal-sized tongue with interposition of the velum, neuromuscular impairment of the genioglossus muscle and other parapharyngeal muscles, prolapse of the posterior wall of the pharynx, and medial movement of the lateral pharyngeal walls [11-14]. During growth, airway obstruction will improve as the mandible grows and the coordination of the parapharyngeal muscles improves in conjunction with voluntary tongue control. After a few months, mandibular catch-up growth improves the prognosis of these children.

This study shows that children born with PRS and severe respiratory problems have a good prognosis provided that adequate support is given by a multidisciplinary team. The least invasive treatment should be considered to minimize morbidity and mortality of these children at birth and for the following months. In our study, treatment concentrating on the relief of airway obstruction using either CPAP or nasopharyngeal tube (PT) was successful in dealing with neonatal respiratory difficulties and avoiding chronic obstruction (Fig. 1).

1. Materials and methods

We identified all children born or admitted in our hospital with PRS between 1984 and 2004. Pierre Robin sequence was defined by a triad cleft palate, micro/retrognathia, and glossoptosis. We divided our patients in 2 groups: patients with nonsyndromic PRS (nsPRS) and those with syndromic PRS (sPRS). The definition of a syndromic patient was based on the presence of other anomalies. In these 2 groups, we identified those with slight respiratory and/or feeding problems (category 1), those with adequate respiration in prone position but feeding difficulties (category 2), and those with severe respiratory and feeding problems requiring either CPAP or PT (category 3) [3]. Accurate assessment was a combination of clinical observation, monitoring, and control of blood gases. Normal venous blood gases values were as follows: pH, 7.4; $Paco_2$, 40 mm Hg; Pao_2, 100 mm Hg; HCO_3^-, 24 mmol/L; base excess, 0.

Children were assessed with oxygen saturation monitored continuously with pulse oximetry. Clinical criteria for respiratory distress were agitation, dyspnea, tachypnea, intercostal recession, tracheal tug, stridor, and position-dependent airway obstruction. Other criteria included continuous pulse oximetry with desaturation at less than 90% and blood gas analysis with respiratory acidosis performed every 2 days. The clinical assessment was carried out by cradling the sleeping baby upright and slowly lowering the child to a supine position while looking for signs of respiratory distress. This was repeated while bottle-feeding. The children were assessed while asleep, feeding, or awake. Indications for PT were either persistent desaturation at less than 90% with clinical evidence of respiratory distress or chronic carbon dioxide retention as evidenced by a base excess of more than 6.5.

A PT was introduced after a lateral x-ray to cut the tube at the right length and was taped to the face. The diameter of the tube was measured to avoid nostril stenosis. The treatment was based on the relief of airway obstruction by CPAP or PT. The CPAP mask was always tried first, and if unsuccessful, PT was used. Nasoendoscopy and bronchoscopy under sedation were performed consecutively by an otolaryngol-

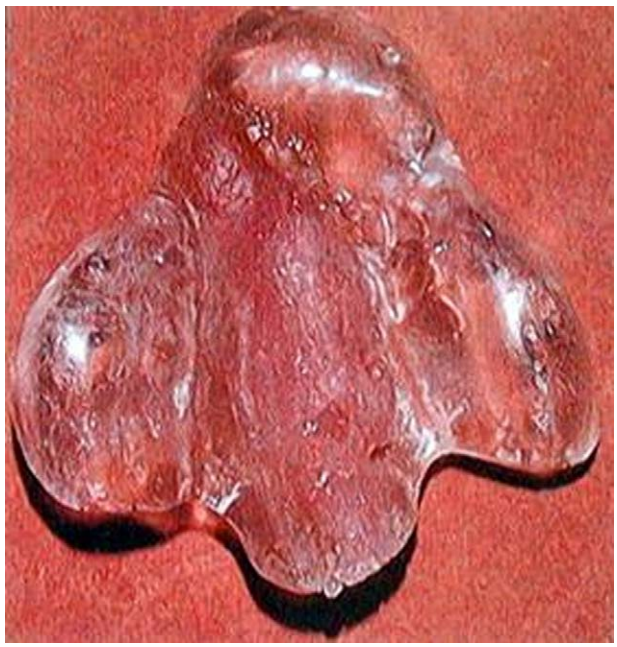

Fig. 2 Palatal appliance held in place by daily application of denture paste.