Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology 27 (2015) 665–669

Contents lists available at ScienceDirect

Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: www.elsevier.com/locate/jomsmp

Clinical Observation

# A new protocol for the management of Robin sequence

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ARTICLE INFO

*Article history:*
Received 16 January 2014
Received in revised form
11 December 2014
Accepted 19 January 2015
Available online 10 March 2015

*Keywords:*
Pierre Robin sequence
Mandibular hypoplasia
Tongue lip adhesion
Neonatal respiratory distress

ABSTRACT

*Aim:* The aim of this paper is to put-forth a protocol for the successful management of patients with Robin sequence reducing the morbidity.
*Subjects and methods:* Three patients with Grade 3 PRS reported to us in a failure to thrive scenario. All of them presented within 3–4 weeks of birth with low birth weight. These patients were successfully managed NG tube feeding initially followed by endoscopic airway assessment with simultaneous tongue lip adhesion and mandibular distraction at a later date.
*Results:* All the patients managed by our protocol showed a significant improvement in the airway status minimizing the respiratory efforts, definite weight gain and good quality of life.
*Conclusion:* Judicious timely intervention is essential for management of Robin sequence to achieve successful results. The protocol put forward in this paper helps achieve good results with minimal morbidity.



## 1. Introduction

Congenital mandibular hypoplasia with retroglossal airway obstruction is one of the main causes of respiratory distress in neonates with high mortality rate [1]. Mandibular hypoplasia may be associated with various craniofacial anomalies the most common one being Pierre Robin sequence (PRS). This anoumalad as described by Randall et al. includes mandibular hypoplasia with glosoptosis and cleft palate [2] with or without airway obstruction. Increased respiratory efforts to maintain a patent airway, feeding difficulties predispose the patient to a catabolic status that progresses to a state of failure to thrive. Neonatal tracheostomy was considered a primary treatment modality to maintain airway patency and circumvent the issues of respiratory distress. However this could be extremely morbid. Alternative measures including glossopexy and mandibular distraction have been tried in patients with mandibular hypoplasia and have shown to have predictable results. The aim of this paper is to describe a series of patients of Pierre Robin sequence who have been successfully managed at our centre by glossopexy and mandibular distraction without the need of tracheostomy and put forward a protocol for the management of patients with PRS.

## 2. Subjects and procedure

Three new born infants of 1–2-week old of non-consanguineous parentage had presented to us with mandibular hypoplasia, cleft palate, low birth weight and significant respiratory distress associated with tracheal tug and intercostal in-drawing (Fig. 1a and b). The infants could maintain a patent airway only in lateral position. The presence of cleft palate associated with nutritional deficiency with increased respiratory efforts predisposed the patient to a failure to thrive scenario. The initial antero-posterior discrepancy between the maxilla and the mandibular arches ranged between 12 and 14 mm. A working diagnosis of PRS Grade 3 [3] was arrived at and naso-gastric feeding was initiated. Airway assessment using bronchoscopy with simultaneous glossopexy (TLA – tongue lip adhesion) was planned with informed consent of the parents. A preoperative echocardiogram was performed to rule out congenital heart disease. Fibreoptic-guided endoscopic airway evaluation was performed which showed retroglossal airway obstruction. Douglas TLA [4] was performed immediately under endotracheal intubation

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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http://dx.doi.org/10.1016/j.ajoms.2015.01.005
2212-5558/

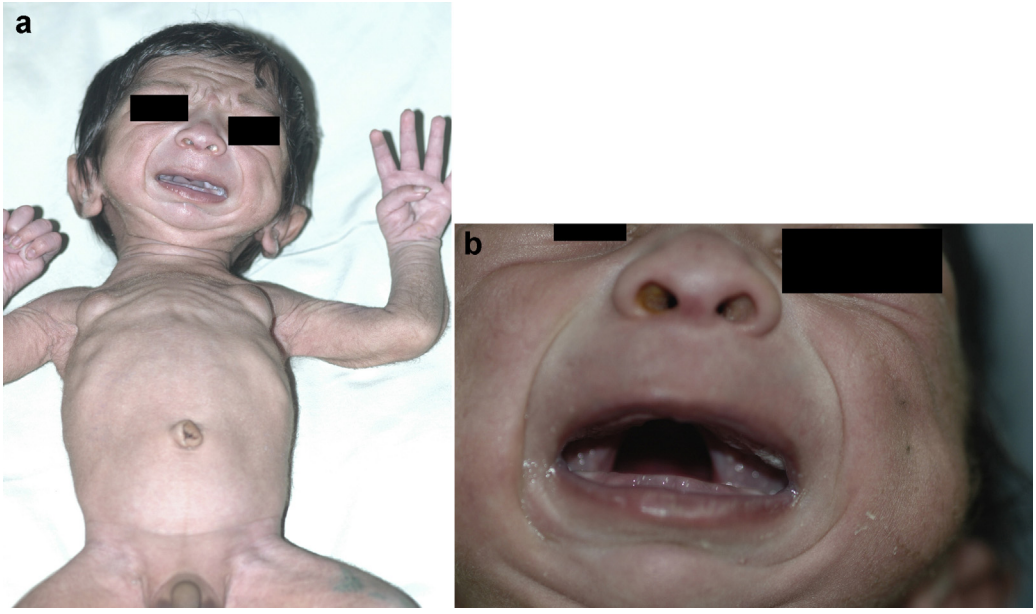


**Fig. 1.** (a) Frontal image of the patient with PRS showing mandibular micrognathia, intercostal in-drawing. (b) Intra oral image showing cleft palate.

to circumvent the issues of airway obstruction. The ventral surface of the tongue and the mucosal surface of the lower lip were deepithelialized and tongue was sutured to the lower lip in two layers – muscle and mucosa. The patient was extubated on the first post-operative day. The airway status improved significantly with glossopexy improving the general condition of the patient. The feeding was continued through the NG tube for the next two weeks after which oral feeding was initiated. There was no associated difficulty in breathing or feeding and the surgical site appeared healthy (Fig. 2). The patient was discharged at this point and reviewed every month. There was a significant decline in the respiratory effort with weight gain at the end of one month. There was some amount of increase in the mandibular length, however this was not significant as the patients continued to have episodes of obstructive sleep apnea (Fig. 3a and b).

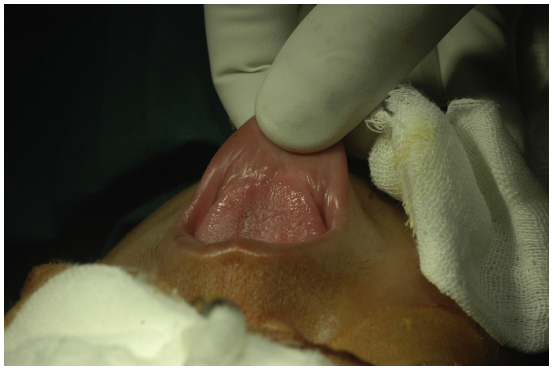

**Fig. 2.** Image of tongue lip adhesion.

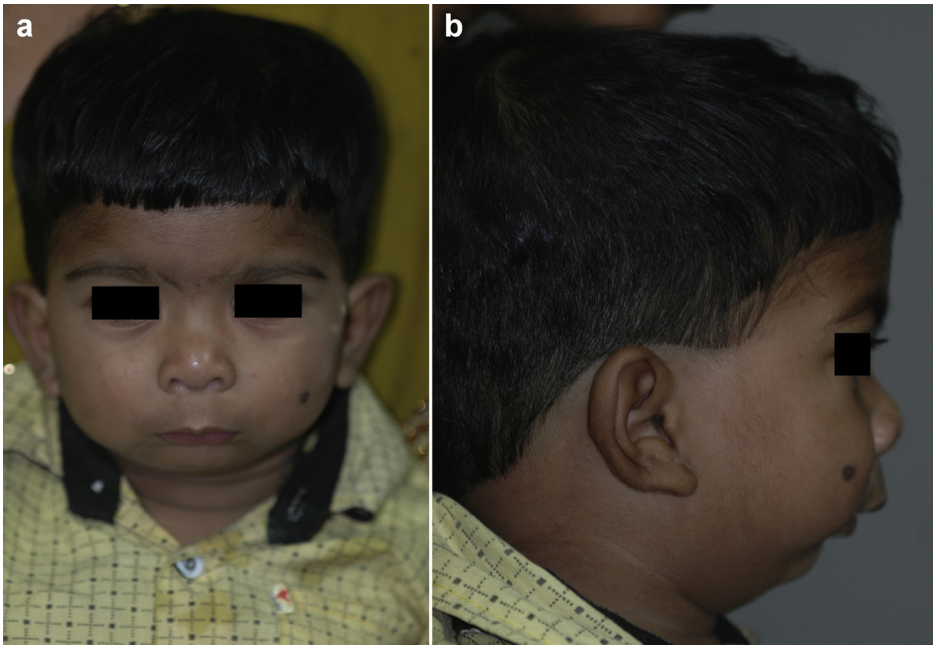


**Fig. 3.** (a) Frontal image of the patient at the age of 6 months. (b) Lateral profile of the patient showing mandibular retrognathism.