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Clinical Paper
Craniofacial Anomalies

Outcomes after tongue–lip adhesion or mandibular distraction osteogenesis in infants with Pierre Robin sequence and severe airway obstruction

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Abstract. The objective was to review and compare outcomes after tongue–lip adhesion (TLA) and mandibular distraction osteogenesis (MDO) in infants with severe breathing difficulties related to Pierre Robin sequence (PRS). A single-centre retrospective (2002–2012) study was carried out; 18 infants with severe breathing difficulties related to PRS resistant to conservative treatment, who underwent TLA or MDO to correct airway obstruction, were enrolled. The primary outcome measures were successful weaning from respiratory support and resumption of full oral feeding. Nine underwent TLA and nine MDO. Eight of the nine infants who underwent MDO and all those treated with TLA were successfully weaned from respiratory support. After discharge, residual respiratory distress was diagnosed more commonly after TLA than after MDO (6/9 vs 1/9, $P = 0.050$). Infants resumed oral feeding sooner after MDO than after TLA (mean days after surgery to full oral feeds 44 ± 24 vs 217 ± 134, $P < 0.003$). The length of hospital stay was longer for infants treated with MDO than for those treated with TLA. The rate of complications was similar. Infants with severe airway obstruction related to PRS can benefit safely from either TLA or MDO. Although MDO lengthens the time to discharge, this option stabilizes airway patency of infants with PRS more efficiently and achieves full oral feeding more rapidly than TLA.

Keywords: Airway obstruction; Mandibular distraction; Pierre Robin sequence; Tongue–lip adhesion.

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Introduction

Pierre Robin sequence (PRS) is a rare (1:8500) congenital anomaly characterized by the triad micrognathia, glossoptosis, and cleft palate, a feature present in more than 90% of cases. The major physiological sequelae after PRS are breathing and feeding problems due to the abnormally small jaw that encourages the tongue to fall back into the pharynx and obstruct the airway.[1] In most neonates with PRS, airway obstruction responds to non-invasive treatments such as prone positioning and nasopharyngeal continuous positive airway pressure.[2] Up to 23% of patients with micrognathia may, however, require interventions beyond these supportive measures, including intubation or tracheostomy.[3] Although tracheostomy in neonatal airway obstruction may be life-saving, it lengthens hospital stays, increases health care costs, and raises problems with care outside the hospital.[4]

The tongue–lip adhesion (TLA) procedure, or glossopexy, was introduced as a surgical option to avoid tracheostomy.[5] In TLA the tongue is anchored to the lower lip and mandible ensuring an anterior lingual position to alleviate the upper airway obstruction. This procedure is typically done in the first months of life and is reversed at around 12 months. Although it is frequently effective in relieving a tongue-based airway obstruction, several investigators consider TLA a temporary procedure because it often requires multiple secondary interventions to achieve airway patency and adequate feeding.[6] TLA may also lead to several complications, including dehiscence, tongue lacerations, injuries to Wharton's ducts, wound infections, scar deformation, and aspiration pneumonia.[7]

Another surgical procedure for the management of airway problems in infants with micrognathia is jaw advancement by mandibular distraction osteogenesis (MDO).[8] Distraction osteogenesis is a technique that was first introduced for lengthening the long bones in the body. As the mandible is lengthened, the anterior mandibular muscles pull the tongue forward, increasing the airway space and relieving airway obstruction. By acting on bone and soft tissue, MDO can definitively correct micrognathia, eliminating the need for a tracheostomy in 90–95% of cases.[9] Although MDO has advanced craniofacial surgery remarkably, it requires time to complete and may lead to complications resulting from injury to the marginal mandibular branch of the facial nerve and the inferior alveolar nerve and damage to the premolar germs.[10]

No studies published to date have compared the effectiveness of TLA and MDO in managing breathing and feeding in infants with PRS treated in a single institution. Having this information would help in the selection of the surgical procedure most likely to provide the best outcome for infants with PRS.

In this single-centre retrospective study we compared the outcomes of infants with severe breathing difficulties related to PRS resistant to conservative treatment, who underwent TLA or MDO procedures to correct severe airway obstruction. The primary outcome measures were successful weaning from respiratory support and resumption of full oral feeding.

Patients and methods

All patients admitted consecutively from 2002 to 2012 to the neonatal and paediatric intensive care unit (ICU), with a diagnosis of moderate to severe respiratory distress related to PRS, who were treated with either TLA or MDO, were identified and included in this retrospective study. Until 2005, all infants admitted with PRS and severe airway obstruction underwent TLA, whereas from 2006 onwards, when an experienced paediatric maxillofacial surgeon joined the multidisciplinary paediatric airway team, infants with the same indications underwent MDO. Institutional board approval and parental consent were obtained.

Infants with clinical features of PRS, defined according to the presence of mandibular deficiency (retrognathia or micrognathia) and tongue malpositioning (glossoptosis) were evaluated within 24–48 h for signs of respiratory distress. Cleft palate was not a required feature. The severity of PRS was defined retrospectively in accordance with the new grading proposed by Cole et al. in 2008, who classified infants with PRS into three grades based on the severity of airway obstruction, oral feeding difficulties, and tongue positioning on finger touch.[11] Infants with PRS and associated physical anomalies related to known syndromes were considered to have syndromic PRS, otherwise PRS was considered isolated. Other malformations unrelated to a specific syndrome were considered comorbidities.

Infants were managed in accordance with the institutional protocol. After the initial evaluation, infants with signs of airway obstruction were assessed by paediatric intensivists and a paediatric pulmonologist to establish the site and degree of airway obstruction, and by a paediatric neurologist and by a geneticist to seek an associated syndrome. To complete the preoperative physical examination, infants underwent airway endoscopy to distinguish the tongue-based obstruction from other causes, such as tracheomalacia, subglottic anomalies, hypotonia, and tongue anomalies. After airway assessment, infants were placed in the lateral position and those with more severe apnoeas had a Guedel airway inserted. If conservative airway management failed to resolve the breathing difficulties, a nasopharyngeal tube was placed for non-invasive ventilation. Infants who still had obstructive apnoea underwent endotracheal intubation. Infants were referred for surgical treatment (paediatric surgeon for TLA and maxillofacial surgeon for MDO), if non-invasive ventilation failed or physical signs of respiratory distress (chest retractions, tracheal tug, stridor) associated with obstructive apnoeas and prolonged feeding difficulties developed.

To compare patients treated with TLA to those treated with MDO we retrieved data from the hospital charts: demographic features, associated syndromes or comorbidities, perioperative clinical data, age at TLA or MDO, surgical complications, gastrostomy tube, time to full oral feeding, need for subsequent surgery, length of hospital stay, residual respiratory distress, and survival.

The primary outcome measures were successful weaning from respiratory support and resumption of full oral feeding. We decided to consider only short-term and medium-term outcomes; long-term outcomes were excluded because TLA and MDO differ in their effects on mandibular growth and the types of complication they carry. We considered the following time points to evaluate these outcomes: time at successful extubation, time at discharge from hospital, and time at full oral feeding. In addition, because respiratory distress, in the form of sleep-disordered breathing, might have developed after hospital discharge, we prolonged the assessment until the infants underwent palatoplasty. We chose this time point because this is when breathing has to be carefully evaluated to avoid possible residual obstructive apnoea secondary to palatoplasty, which may arise in infants with PRS.

For TLA, surgeons used a modified Routledge procedure that induced mucosal and muscular adhesions.[12] Tongue–lip detachment surgery took place at around 1 year of age under general anaesthesia.

The surgical protocol for MDO comprised jaw osteotomy and device