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Mandibular distraction osteogenesis for the management of upper airway obstruction in children with micrognathia: a systematic review

O. Breik[1,2], D. Tivey[2], K. Umapathysivam[2], P. Anderson[3,4]
[1]Oral and Maxillofacial Surgery, Royal Melbourne Hospital, Melbourne, Victoria, Australia; [2]Joanna Briggs Institute, School of Translational Science, University of Adelaide, Adelaide, South Australia, Australia; [3]Australian Craniofacial Unit, Women's and Children's Hospital, Adelaide, South Australia, Australia; [4]Oral and Maxillofacial Surgery Unit, Universiti Sains Malaysia, Kota Bharu, Malaysia

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Abstract. Mandibular distraction osteogenesis (MDO) is increasingly used for neonates and infants with upper airway obstruction secondary to micrognathia. This systematic review was conducted to determine the effectiveness of MDO in the treatment of airway obstruction. The databases searched included PubMed, Embase, Scopus, and grey literature sources. The inclusion criteria were applied to identify studies in children with clinical evidence of micrognathia/Pierre Robin sequence (PRS) who had failed conservative treatments, including both syndromic and non-syndromic patients. Overall 66 studies were included in this review. Primary MDO for the relief of upper airway obstruction was found to be successful at preventing tracheostomy in 95% of cases. Syndromic patients were found to have a four times greater odds of failure compared to those with isolated PRS. The most common causes of failure were previously undiagnosed lower airway obstruction, central apnoea, undiagnosed neurological abnormalities, and the presence of additional cardiovascular co-morbidities. MDO was less effective (81% success rate) at facilitating decannulation of tracheostomy-dependent children ($P < 0.0001$). Failure in these patients was most commonly due to severe preoperative gastro-oesophageal reflux disease, swallowing dysfunction, and tracheostomy-related complications. The failure rate was higher when MDO was performed at an age of $\geq$24 months. More studies are needed to evaluate the long-term implications of MDO on facial development and long-term complications.

Key words: mandibular distraction tracheostomy; decannulation; Pierre Robin sequence; upper airway obstruction.

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Micrognathia is a congenital condition involving an abnormally small mandible. This condition tends to occur in conjunction with posterior tongue displacement (glossoptosis), which can lead to physical obstruction of the oropharyngeal and hypopharyngeal regions on inspiration. This upper airway obstruction may be life-threatening and may require urgent medical intervention.

In 1923, a French stomatologist was the first to describe the constellation of symptoms associated with upper airway obstruction in neonates now known as Pierre Robin sequence (PRS).[1] This sequence is a craniofacial anomaly characterized by mandibular micrognathia (mandibular hypoplasia), glossoptosis, and in some cases a 'U-shaped' cleft palate. There is only limited epidemiological data, but the incidence has been reported to range from approximately 1 in every 8500 live births in Liverpool, UK[2] to 1 in 14,000 live births in Denmark.[3] The most recent study from Germany reports an incidence of approximately 1 in 8000 births.[4] This variation in incidence is related in part to the inconsistent definition of PRS in the literature.

The diagnosis of patients with PRS is challenging due to the wide spectrum of PRS phenotypes, variation in degree of airway obstruction, feeding difficulties, and the need for treatment. This has led to some authors only characterizing those with airway obstruction needing treatment as having PRS[5]; others will include all patients with micrognathia and glossoptosis, or limit the PRS diagnosis to those with associated cleft palates.[3] Although these clinical features are most commonly seen in isolation,[6] they can also occur in association with other syndromes of the craniofacial skeleton; for example, Treacher Collins syndrome, Stickler syndrome, and Nager syndrome. Such co-occurrences further complicate the diagnosis. These syndromes differ in pathogenesis from isolated PRS, but all can have micrognathia with glossoptosis and hence airway obstruction. For simplicity, those without an associated syndrome are referred to in this study as having 'isolated PRS' and those with an associated syndrome are referred to as having 'syndromic micrognathia'. The varying phenotypes and presumed causes of this anomaly make comparison of the myriad of protocols advocated for management difficult.[7]

The most important consequences of micrognathia and PRS are the inability to effectively breathe or feed due to airway obstruction. The majority of children born with micrognathia or PRS have no respiratory distress. Those with mild symptoms of respiratory distress can often be treated conservatively with prone positioning or non-invasive techniques, such as a nasopharyngeal airway or the application of nasal continuous positive airway pressure (CPAP). The rate of success with the use of nasopharyngeal airways varies in the literature, ranging from 48%[8] to 100%.[9] A large case series study focusing on children with non-syndromic PRS demonstrated that less than 10% required a surgical intervention.[10]

For neonates with severe respiratory distress, or those who fail initial conservative treatment, the airway dysfunction can be a life-threatening emergency. The nasopharyngeal airway and CPAP can only be tolerated for a limited period of time, and in some cases, children need to be intubated and ventilated to maintain adequate oxygenation.[11] Children who require prolonged treatment with these measures may require a more definitive surgical intervention.

Several surgical treatments have been described for the treatment of the child with micrognathia. In 1946 Douglas described the use of tongue–lip adhesion (TLA) for the treatment of upper airway obstruction associated with micrognathia.[12] This procedure involves surgically fusing the tongue to the anterior lower lip to hold the tongue in an anterior position. The adhesion is usually reversed with another surgical procedure at 9–12 months of age. However, the underlying cause of the obstruction is not fully addressed by TLA, and wound dehiscence and feeding difficulties are common,[6,13] thus many centres have abandoned it as a viable treatment option.[11]

Other surgical options described include mandibular traction and advancement appliances,[14,15] and sub-periosteal release of the floor of the mouth musculature.[16,17] However these procedures have not met with widespread success and have largely been abandoned by the larger centres.

Mandibular distraction osteogenesis (MDO)

Since the introduction of distraction osteogenesis for the craniofacial skeleton in the mid-1980s to early 1990s, it has been used to deal with various types of reconstructive dilemma.[18,19] MDO for infants with micrognathia has been used for unilateral mandibular lengthening by distraction for cases of hemifacial microsomia[20] and bilateral MDO for cases of Treacher Collins syndrome.[21] Initially MDO was used to resolve upper airway obstruction and to facilitate the removal of a tracheostomy. Since then, it has been used increasingly as the primary surgical option for the management of neonates and infants with micrognathia or PRS with upper airway obstruction.[22]

MDO relieves the airway obstruction by lengthening the mandible. This stretches the tongue attachments to the mandible (genioglossus muscle), which positions the tongue more anteriorly, relieving the glossoptosis. Most children with upper airway obstruction have demonstrated an improvement in their respiratory status within a few days of distraction. For those children who are intubated and mechanically ventilated, this may mean extubation and transfer to a regular hospital ward.

Several case series have demonstrated the effectiveness of MDO in alleviating upper airway obstruction in neonates, infants, and older children with PRS.[23] Most patients were able to avoid tracheostomies, and those who already had tracheostomies could be decannulated. A systematic review performed in 2008 evaluated the effectiveness of MDO in several clinical applications.[23] The review evaluated 178 studies including 1185 patients. Success in preventing tracheostomies was achieved in 91.3% of patients. However, the authors of that review searched only the PubMed database on the applications of unilateral and bilateral mandibular distraction in both children and adults. Limiting the search to a single database is a significant methodology limitation of that review. In addition the study also included all possible causes of micrognathia, including temporomandibular joint (TMJ) ankylosis, hemifacial microsomia, and syndromic micrognathia, which have different aetiologies to isolated PRS. No comparative subgroup analyses were performed to differentiate between these groups. Furthermore, the authors did not evaluate any long-term outcomes in children and did not discuss reasons for failure of distraction.

This current review was performed with the aim of extending the search across multiple databases to include the current available evidence for the effectiveness of mandibular distraction for the treatment of upper airway obstruction in children with micrognathia. This review also reports reasons for failure and compares outcomes between isolated PRS/micrognathia patients and syndromic micrognathia patients. A further aim is to determine the effects of mandibular distraction on the other complications of micrognathia, including feeding and weight gain, gastro-oesophageal reflux,