

“Mandibular distraction osteogenesis for severe airway obstruction in Robin Sequence. Case report”

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SUMMARY. Mandibular distraction osteogenesis (MDO) has been increasingly gaining interest over the last decade as a treatment alternative for the challenging airway management in infants with the Robin Sequence (RS). This paper is a case report of a male child diagnosed with RS, suffering from life-threatening airway obstruction and feeding difficulties, treated with tracheostomy and gastrostomy since infancy. After evaluation of the patient by a multidisciplinary team of specialists, MDO performed as soon as possible, was considered the optimal treatment, not only to address the severe micrognathia but also to allow early tracheal decannulation. As the lack of space intraorally contraindicated the use of internal distractors, they were placed externally bilaterally. The patient was successfully decannulated 3 weeks postoperatively and the gastrostomy was removed 1 month postoperatively. The mandibular expansion exceeded 20 mm bilaterally and the maxilla-mandible discrepancy was fully corrected. There were no complications related to device placement, activation or removal. Follow-up clinical and radiographic examinations of the patient 1 year after the removal of the distractors revealed improved mandibular projection and continued mandibular growth. No significant scarring occurred at the surgical site and the patient has normal respiratory and feeding function. © 2009 European Association for Cranio-Maxillo-Facial Surgery

Keywords: Robin sequence, mandibular distraction osteogenesis, airway obstruction

INTRODUCTION

The birth of a baby with Robin Sequence (RS) often constitutes a medical emergency in the delivery room. 44–70% of the affected neonates may experience varying degrees of respiratory distress ranging from positional respiratory obstruction, episodic apnoea or cyanosis and carbon dioxide retention due to complete obstruction (Marques et al., 2001; Monasterio et al., 2002). Feeding difficulties mainly due to oro-oesophageal dysmotility and swallowing disorders are common, resulting in malnutrition and deterioration of the patient’s general condition (Monasterio et al., 2004). Additionally frequent airway and pulmonary infections or aspiration pneumonia attributed to the gastro-oesopharyngeal reflux may occur (Evans et al., 2006). To prevent the above mentioned consequences as well as long-term problems such as pulmonary hypertension and cor pulmonale, inability to thrive and developmental delay, immediate measures are required in most RS cases (Li et al., 2002).

Both non-operative and surgical techniques have been proposed and applied in order to manage severe airway obstruction and improve airway and nutrition permeability in RS cases. Prone positioning of the infant, laryngeal mask, oral appliances such as mandibular repositioning appliances (Hoekama et al., 2004), prolonged nasopharyngeal intubation (Wagener et al., 2003), tongue-lip

adhesion or glossopexy (Kirschner et al., 2003), release of the musculature of the floor of the mouth, mandibular distraction osteogenesis (MDO) (Denny and Amm, 2005) and tracheostomy have been used (Marques et al., 2001).

MDO has been gaining interest in the last decade for the treatment of micrognathia in RS cases (Daskalogiannakis et al., 2001; Monasterio et al., 2002; Mandell et al., 2004; Burstein and Williams, 2005; Burstein, 2008; Dauria and Marsh, 2008). Its contribution to early decannulation of tracheostomy-dependent RS patients has been well documented (Denny et al., 2001; Figueroa and Polley, 2002; Monasterio et al., 2002; Steinbacher et al., 2005). The procedure in the neonatal period is not without risks and complications requiring careful patient selection (Schaefer et al., 2004; Marques et al., 2005).

The aim of this paper is to report a case of a tracheostomy and gastrostomy-dependent 9-month-old male with RS, treated with bilateral MDO.

CASE REPORT

The patient, a monozygotic twin brother of a phenotypically healthy male, was diagnosed with the RS at birth, as he presented with the triad of mandibular micrognathia, glossoptosis and cleft palate.

The child was referred at the Department of Oral and Maxillofacial Surgery of the Children’s Hospital “A. &

P. Kyriakou", at the age of 8 months, being severely under weighed (4.8 kg). After experiencing multiple episodes of severe obstruction and choking (sleep apnoea episodes were more than 20 per hour), the patient had undergone tracheostomy at the age of 15 days. As he failed to gain weight and thrive, he was nourished through a gastrostomy applied at the age of 2 months (Fig. 1). The whole physical development of the child, who had been hospitalized several times for pulmonary infections, was far behind the normal for his age and the parents were desperately seeking for ways to improve the child's life and comfort him.

After thorough examination by a multidisciplinary team and taking into account that the child's main problem was the micro- and retrognathia, (which led to the placement of the floor of the mouth and tongue posteriorly), a therapeutic augmentation of the mandible via distraction osteogenesis was planned. Radiological evaluation to help plan the operation, included lateral and anterior-posterior cephalograms, as well as CT and three-dimensional CT scans (Fig. 2).

It was decided that the distractors would be placed bilaterally in the posterior mandibular body close to the angle, through an extraoral approach due to the lack of space intraorally. The capacity of the chosen devices would enable 2 cm of lengthening. At the age of 9 months, with a body weight of 5.1 kg the child was operated on, under general anaesthesia through the tracheostomy. The operative site was approached through a submandibular incision, the device was positioned, screws were inserted prior to osteotomy and the device was removed. The osteotomy was performed and the distractor was fixed and its function was checked, the device was kept closed and careful suturing followed. The same procedure was performed bilaterally (Figs. 3–5).

Active distraction at a rate of 1 mm/day at each side began the 4th post-operative day and lasted for 20 days. The parents were instructed in how to activate the devices and the patient was discharged after 1 week. As the mandible increased in size and the tongue was allowed more space anteriorly, breathing, (checked daily), became possible and the tracheostomy tube was removed 3 weeks postoperatively. Gastrostomy was retained for the first post-operative month although the child was already tolerating liquid diet. After a consolidation period of 4 weeks the patient was examined clinically and radiologically to confirm ossification of the distraction sites and a bilateral length gain of 2 cm. The distractors were removed without difficulty 2 months postoperatively under general anaesthesia (Figs. 6 and 7).

Follow-up continued monthly (Fig. 8) and after 1 year the symptoms of airway obstruction had resolved and glossoptosis and the consequent obstruction had subsided. The child has been slowly learning to chew and swallow and the improved maxillomandibular relationship has been maintained (Fig. 9). A noteworthy growth catch-up occurred in the meantime and his body weight although not yet normal for his age, was 13.5 kg at the age of 22 months. There was an obvious improvement in the child's communication and social attitude and currently no additional surgery is anticipated. Though it is



Fig. 1 — The child immediately pre-operatively, at the age of 9 months.

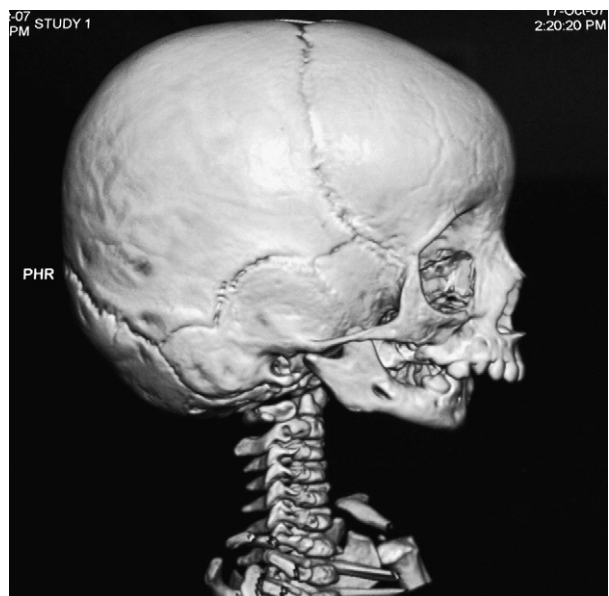


Fig. 2 — Preoperative three-dimensional CT of the patient's craniofacial complex. Note the severe micrognathia.

difficult to accurately assess the inferior alveolar nerve function at that age, no special response to painful stimuli at the chin, no drooling or oral motor defects were detected at follow-up. Although scarring was not significant, revision may be required in the future. Three months follow-up will continue for the next 3 years, and the closure of the rather small cleft palate is planned.

DISCUSSION

MDO allows molding of the bone into different shapes in order to address more adequately skeletal deformities and asymmetries, while also providing the advantage of larger skeletal movements without the inherent risk of relapse, associated with conservative treatment alternatives such as obturators (Cope et al., 1999). However, the ideal timing and technique for the procedure are still controversial (Denny and Kalantarian, 2002; Burstein and Williams, 2005; Lin et al., 2007).

Internal distractors allow more efficient application of the distraction force, reducing the surgical trauma and have more stable biomechanics, diminishing intra- and

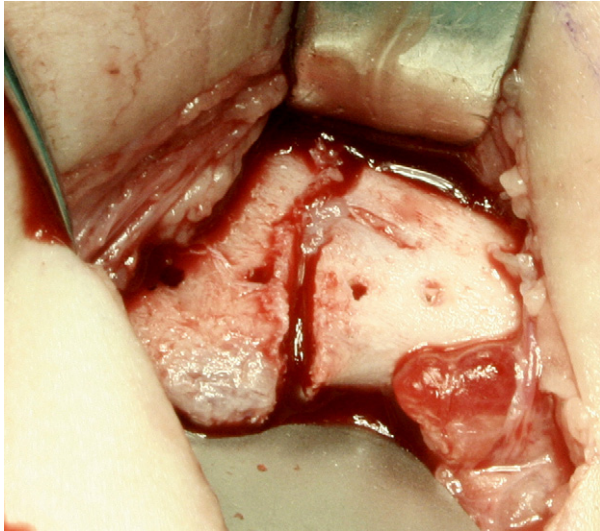


Fig. 3 – Osteotomy at the right side of the mandible.

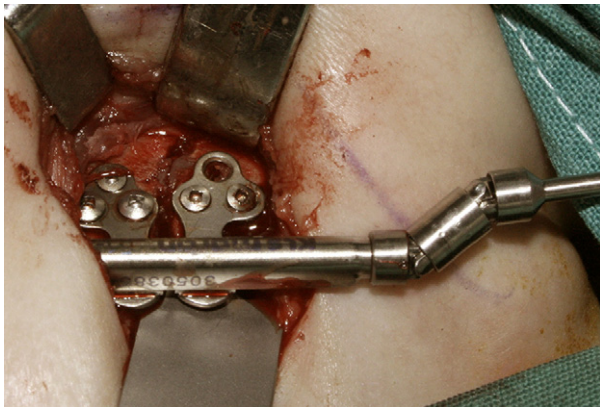


Fig. 4 – Final placement of the distractor.

post-distraction complications (such as device dislodgement during the consolidation period) but are technically more challenging to apply and have limitations related to the size of the device and the restricted access of the oral cavity (Cope et al., 1999; Cademartiri et al., 2004; Mandell et al., 2004; Lin et al., 2007).

Denny et al. (2001); Sidman et al. (2001); Denny and Kalantarian (2002) and Monasterio et al. (2002) have independently established the feasibility of using external distractors in infants with severe micrognathia and airway obstruction. However, external devices may result in significant scarring and require care to maintain expander integrity and pin-site hygiene (Rhee and Buchman, 2003; Tibesar et al., 2006). Recently developed internal appliances are smaller (Cademartiri et al., 2004; Tibesar et al., 2006) and resorbable one-stage devices (Burstein and Williams, 2005; Burstein, 2008) favour application even in the newborn and infants. In this case, the external approach was considered mandatory due to poor intra oral access and the small mandible. The devices used in this case were originally designed as internal distractors and were selected

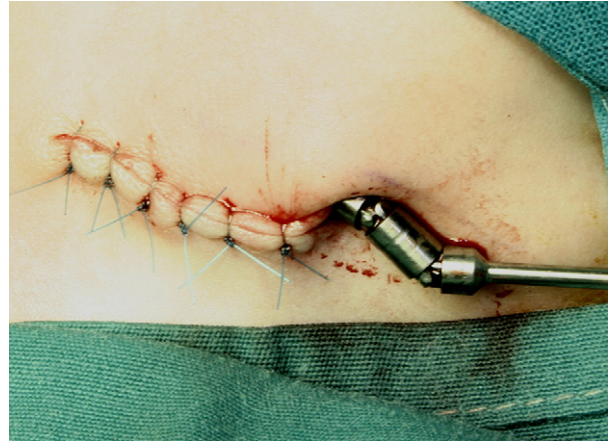


Fig. 5 – The surgical site after suturing.



Fig. 6 – Anteroposterior cephalogram after consolidation showing the acquired bone.

because their small size and shape would be convenient postoperatively for both the infant and the parents; although improperly used they provided a successful outcome.

Potential risks and complications such as bony malunion, malalignment and infection are only infrequently observed in MDO, thanks to the excellent vascularity of the craniofacial region (Monasterio et al., 2002). Other potential complications of MDO include nerve injuries (Parashar et al., 2006; Tibesar et al., 2006; Lin et al., 2007), mandibular growth disturbance, tooth bud injury (Tibesar et al., 2006) and transient feeding difficulties (Howlett et al., 1999; Sidman et al., 2001). In this case there was not any noteworthy complication intra- or post-operatively.

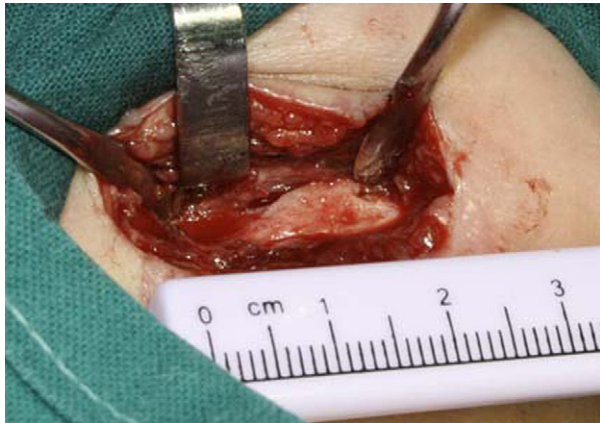


Fig. 7 — The osteotomy site after the removal of the distractor. The bilateral length gain of the mandible was 20 mm.



Fig. 8 — Post-operative photograph of the patient 1 month after the removal of the distractors.

Patient selection is critically important to ensure appropriate application and successful outcomes. Treatment protocols must be individualized for every RS patient, depending on the underlying cause of obstruction (Schaefer et al., 2004). According to Marques et al. (2005) a preoperative comprehensive airway endoscopy is essential to define the type of pathology and to determine which treatment modalities are indicated. Moreover, Tibesar et al. (2006) point out that the patient must fulfil certain criteria, before MDO is performed. In this severe case, the application of tracheostomy and gastrostomy, though temporarily helping the infant to survive, would undoubtedly have produced future difficulties. MDO offered spectacular and quick clinical change together with encouraging perspectives to the child's normal growth. Breathing became normal, having been previously traumatic for everyone due to



Fig. 9 — Post-operative photograph of the patient 1 year after the removal of the distractors.

tracheostomy. Swallowing has been continuously improving. One year after the removal of the distractors, MDO has proved to be a life-saving process for this child who, for 8 months, had suffered the consequences of a severe micrognathia. As breathing and mastication improve, the child's musculoskeletal system and mandible are expected to grow. Nevertheless, as the sequence has genetic background, multidisciplinary follow-up of the patient including registration of mandibular growth and possible changes of soft tissues in the pharyngeal region will continue and last as long as needed; further orthodontic or surgical interventions though, may be required.

CONCLUSION

MDO is a safe and effective technique that can be applied to predictably relieve severe upper airway obstruction in selected RS cases. However, preoperative evaluation, proper patient selection, meticulous surgical technique, as well as careful and appropriate post-operative care are mandatory to apply the procedure with minimal risk. In order to avoid the limitations of alternative surgical procedures and the tracheostomy-associated morbidity, MDO should be considered among the routine treatment modalities for RS patients with severe airway obstruction.

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