Distraction Osteogenesis for Management of Severe OSA in Pierre Robin Sequence: An Approach to Elude Tracheostomy in Infants

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Abstract

Background Severe obstructive sleep apnoea (OSA) is a life threatening condition associated with Pierre Robin sequence (PRS) due to mandibular micrognathia and glossoptosis. Often these patients require tracheostomy at an early age which has high morbidity. Distraction osteogenesis (DO) is an accepted method of treatment for patients with hypoplastic mandible to achieve mandibular lengthening without need for a bone graft. It has also been used in respiratory distressed neonates and infants to avoid tracheostomy.

Case report An eight month old baby, a diagnosed case of PRS with severe OSA and recurrent episodes of aspiration pneumonia and on nasogastric tube feeding since birth was referred to us for evaluation and possibility of therapeutic augmentation of the mandible by DO. After a thorough clinico-radiological assessment the child was

operated for bilateral extraoral placement of horizontal corpus distractor. A total distraction of 12 mm was carried out and consolidation of callus was monitored by USG. Postoperatively the patient was followed up for 12 months. Presently she has normal respiratory and feeding function without any episode of aspiration pneumonia.

Conclusion Mandibular corpus DO is a safe and effective technique that can be applied to predictably relieve severe upper airway obstruction in selected PRS cases. In order to avoid the limitations of alternative surgical procedures and the tracheostomy-associated morbidity, DO should be considered among the routine treatment modalities.

Keywords Pierre Robin sequence (PRS) · Micrognathia · Obstructive sleep apnoea (OSA) · Distraction osteogenesis (DO)

Introduction

Pierre Robin sequence (PRS) is a series of events arising from the poor development of the mandible with micrognathia causing the tongue to be displaced back in the mouth (glossoptosis). The tongue may interfere with the approximation of palatal shelves resulting in a horse-shoe shaped cleft palate. Concomitant reduction of the oropharyngeal airway leads to upper airway obstruction. The patients may present with symptoms of obstructive sleep apnoea (OSA) that in severe cases needs tracheostomy [1–3].

Computed tomography (CT) and 3-dimensional (3D) reconstructive imaging techniques are widely used in the diagnosis, treatment planning, surgical guidance, evaluation of results, and follow-up studies of maxillofacial deformities. Though considered very useful, the undesirable exposure to radiations cannot be justified.

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The management of PRS varies according to the degree of severity of the individual case. Though conservative management includes prone positioning and placement of nasopharyngeal airway stents, significant airway obstruction requires more aggressive therapy, including tongue lip adhesion and hyomandibulopexy. Tracheostomy is considered the conventional alternative for management of upper airway obstruction in PRS; however, it is associated with high morbidity and mortality rates. The average age of decannulation for children with PRS is 3.1 years [4, 5].

Distraction osteogenesis (DO) is an accepted method of treatment for patients with hypoplastic mandible to achieve mandibular lengthening without need for a bone graft and in cases of ramus condyle unit reconstruction. It is shown to be effective in resolving upper airway obstruction and tracheostomy decanulation. It has also been used in respiratory distressed neonates and infants to avoid tracheostomy [6, 7]. Troullis et al. [8] described ultrasonography (USG) as an accurate non-invasive technique to assess mandibular DO. We report a case of PRS with dysphagia and recurrent aspiration pneumonia treated at our centre by DO and followed up for a period of 18 months with successful improvement of OSA symptoms and weaning of nasogastric feeding tube and improvement in oral feeding.

Case Report

An eight month old infant, diagnosed case of PRS with retromicrognathia, glossoptosis, OSA and recurrent aspiration pneumonia was referred from the department of Paediatrics. She was managed conservatively since birth with prone positioning and nasogastric tube feeds. Infant suffered severe airway obstruction, and could barely breathe with extended neck and feeding was only possible through the nasogastric tube. At birth her weight was 2.1 kg. She had retromicrognathia, glossoptosis, and symptoms of OSAS as noisy breathing, frothing and drooling of saliva, fragmented sleep, pauses in respiration and excessive daytime somnolence. She was unable to control her airway during oral feeding, as evidenced by repeated episodes of choking and obstruction. She developed seven episodes of aspiration pneumonia since her birth and was admitted twice to paediatric intensive care unit (PICU).

A CT and MRI scan depicted normal brain parenchyma with no focal lesion. She was observed in PICU for two consecutive days with continuous monitoring for oxygen saturation (SpO₂) using a pulse oximeter. The SpO₂ ranged between 75 and 85 % (mean of 81 %). The child was evaluated by a multidisciplinary team of specialists with recommendations of tracheostomy or DO for airway

management and gastrostomy for feeding purposes as possible management modalities.

The baby was referred to our centre for consideration of therapeutic augmentation of the mandible by DO. Oral examination revealed adequate mouth opening and presence of single deciduous tooth (tooth no 71). The mandibular arch was contained in the maxillary arch and the horizontal discrepancy (over jet) was 10 mm (Fig. 1). Radiological evaluation to help plan the operation, included PA and lateral skull radiograph. Bilateral mandibular corpus distraction osteogenesis (MDO) of 12 mm was considered the optimal treatment, performed as soon as possible, not only to address the severe micrognathia but also to allow early weaning of the nasogastric feeding tube.

Preanaesthetic assessment was carried out and high risk consent was obtained from the parents. The surgical procedure was performed under general anaesthesia with orotracheal intubation. Small mandible and lack of space intraorally indicated the use of semi buried distractors extra orally. A submandibular incision was given and dissection carried out to expose the upper and lower border of body of mandible at its transition to ramus. Position of a unidirectional distractor (Mandibular horizontal distractor, KLS Martin, Tuttlingen, Germany) of 15 mm size was predetermined by engaging one screw in each foot plate in the adjoining holes at the most proximal part of the corpus at lower border. The vector of distraction was parallel to the lower border of mandible. The device was removed and the corticotomy and low intensity fracture completed. The device was returned to its predetermined position and secured with titanium mini screws of 2 mm diameter and 7 mm length. A total of six mini screws were used, three in each foot plate (Fig. 2). The distractor was activated to verify the completeness of the osteotomy. Similar procedure was carried out on the other side. Soft tissues closure was done, dressing applied and the activation port was left

Fig. 1 Mandibular arch contained in the maxillary arch

Fig. 2 Distractor fixation at lower border of mandible

Fig. 3 Open activation port

Fig. 4 Negative over jet

Fig. 5 X ray mandible lateral view confirming distracted segments

Fig. 6 USG evaluation of distracted callus at 9 weeks

uncovered (Fig. 3). After latency period of 48 h both the devices were activated simultaneously at a rate of 1 mm/day and a rhythm of 0.5 mm 12 hourly. A total distraction of 12 mm was done to achieve negative over jet (Fig. 4). A radiograph was taken to confirm the distraction (Fig. 5). Patient was monitored in PICU during the period of distraction. The patient was successfully weaned off nasogastric tube 1 week post distraction and the symptoms of OSA improved with absence of noisy breathing and drooling of saliva. Oxygen saturation improved to 95–99 % (mean 97 %). The regenerated callus was evaluated at an interval of 3, 6 and 9 weeks by USG to assess the consolidation (Fig. 6). The distractors were removed under general anaesthesia after 10 weeks of consolidation. The deposited bone evaluated subjectively at the time of distractor removal had intact cortices with no mobility.

The patient is on regular follow up at 1, 3, 6 and 12 months. Eruption of all deciduous teeth is completed

and mandibular projection is maintained. No significant scarring is observed at the surgical site and the patient has normal respiratory and feeding function. Weight gain is satisfactory and there is no episode of aspiration pneumonia.

Discussion

The features of PRS are micrognathia, glossoptosis and cleft palate. Our case had presentation of micrognathia and glossoptosis leading to OSA. There was no cleft defect. Imaging evaluation of the morphological airway changes has been performed in patients with OSA using techniques such as cephalometric radiography, CT, and magnetic resonance imaging [9–11]. CT scans though can create 3D images, exposes the child to ionizing radiation naturally experienced during approximately 8 months. It is mandatory to discuss the benefits and risks with the parents [12]. CT scan was advised by paediatrician to rule out any focal lesion of brain. In our case, lateral cephalogram was difficult due to unstable head position. Monitoring of distracted callus maturation was carried out by USG as it involves no radiation exposure.

The use of mandibular distraction osteogenesis (MDO) in alleviating upper airway obstruction in infants with micrognathia is rapidly expanding. Series presented by various authors [13–15] confirm the utility of this option. New internal devices have been shown to have significant advantages over the bulky external devices [16, 17]. Unidirectional vector forces are adequate to achieve a good maxillomandibular relationship without any open or cross bite deformities [18]. Functional loading and bony remodelling in young age group are important factors in achieving this relationship.

Internal distractors allow effective application of the distraction force, reducing the surgical trauma and have more stable biomechanics, diminishing intra and post-distraction complications. Technically its application is more challenging and has limitations related to the size of the device and restricted intraoral access [19, 20]. Use of external distractor in infants with severe micrognathia and airway obstruction is well established. However, external devices may result in significant scarring and require care to maintain expander integrity and pin-site hygiene [21, 22]. Recently developed internal appliances are smaller and resorbable one stage devices favour application even in new born and infants [23, 24]. The devices used in this case are originally designed for intraoral application and were selected because of its small size and its convenience postoperatively for both the infant and parents. Small mandible and poor intra oral access mandated the extraoral application.

Nerve injuries, mandibular growth disturbance, tooth bud injury and transient feeding difficulties have been described as potential risks and complications. Malunion, malalignment and infection are infrequently observed in MDO due to the excellent vascularity of the craniofacial region [7, 10, 25]. In our case there was no noteworthy intra or post-operative complication. Surgical access and visibility was good due to extraoral approach. To avoid damage to the tooth buds the distractors were placed at the lower border of mandible and secured with 7 mm long mini screws. Continuity of oral epithelium was maintained thereby avoiding contamination of the corticotomy site with oral fluids.

Patient selection is critical to ensure appropriate application and successful outcome of MDO. Treatment protocol must be individualized for every PRS patient, depending on the underlying cause of obstruction [26]. In this case, tracheostomy and gastrostomy, though temporarily may have helped the infant to survive, would undoubtedly have more surgical morbidity.

MDO is a safe and effective technique that can be applied to predictably relieve severe upper airway obstruction in selected PRS cases. However, patient selection, preoperative evaluation, meticulous surgical technique 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 as a routine treatment modality for PRS patients with severe airway obstruction.

USG is an early indicator of callus mineralization and could be useful in ascertaining the timing of distractor removal [27]. We monitored callus maturation by USG and distractors were removed after 10 weeks of consolidation. Owing to the plasticity and malleability of the infant mandible, that allow good bone remodelling, unidirectional internal distractors can be used to achieve satisfactory maxillomandibular relationship.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Human and Animals Rights Statement This article does not contain any studies with human participants or animals performed by any of the authors.

Informed Consent Informed consent was obtained from the parents of the patient included in the study.

References

1. Cohen SR, Simms C, Burstein D (1998) Mandibular distraction osteogenesis in the treatment of upper airway obstruction in children with craniofacial deformities. *Plast Reconstr Surg* 101:312–318

2. Dauria D, Marsh JL (2008) Mandibular distraction osteogenesis for Pierre Robin sequence: What percentage of neonates need it? *J Craniofac Surg* 19(5):1237–1243
3. Marques IL, Barbieri MA, Bettiol H (1998) Etiopathogenesis of isolated Robin sequence. *Cleft Palate Craniofac J* 35:517–525
4. Denny A, Kalantarian B (2002) Mandibular distraction in neonates: a strategy to avoid tracheostomy. *Plast Reconstr Surg* 109:896–904
5. Mandell DL, Yellon RF, Bradley JP, Izadi K, Gordon CB (2004) Mandibular distraction for micrognathia and severe upper airway obstruction. *Arch Otolaryngol Head Neck Surg* 130(3):344–348
6. Looby JF, Schendel SA, Lorenz HP, Hopkins EM, Aizenbud D (2009) Airway analysis: with bilateral distraction of the infant mandible. *J Craniofac Surg* 20(5):1341–1346
7. Morovic CG, Monasterio L (2000) Distraction osteogenesis for obstructive apneas in patients with congenital craniofacial malformations. *Plast Reconstr Surg* 105:2324–2330
8. Troullis MJ, Coppe C, O'Neill MJ, Kaban LB (2003) Ultrasound: assessment of the distraction osteogenesis wound in patients undergoing mandibular lengthening. *J Oral Maxillofac Surg* 61(10):1144–1149
9. Pépin JLD, Veale D, Ferretti GR, Mayer P, Lévy PA (1999) Obstructive sleep apnea syndrome: hooked appearance of the soft palate in awake patients cephalometric and CT findings. *Radiology* 210:163–170
10. Hoponik EF, Smith PL, Bohlman ME, Allen RP, Goldman SM, Bleecker ER (1983) Computerized tomography in obstructive sleep apnea. *Am Rev Respir Dis* 127:221–226
11. Ryan CF, Lowe AA, Li D, Fleetham JA (1991) Magnetic resonance imaging of the upper airway in obstructive sleep apnea before and after chronic nasal continuous positive airway pressure therapy. *Am Rev Respir Dis* 144:939–944
12. Zoetelief J, Geleijns J (1998) Patient doses in spiral CT. *Br J Radiol* 71:584–586
13. Denny AD, Talisman R, Hanson PR, Recinos RF (2001) Mandibular distraction osteogenesis in very young patients to correct airway obstruction. *Plast Reconstr Surg* 108(2):302–311
14. Sidman JD, Sampson D, Templeton B (2001) Distraction osteogenesis of the mandible for airway obstruction in children. *Laryngoscope* 111(7):1137–1146
15. Wittenborn W, Panchal J, Marsh JL, Sekar KC, Gurley J (2004) Neonatal distraction surgery for micrognathia reduces obstructive apnea and the need for tracheostomy. *J Craniofac Surg* 15(4):623–630
16. Lin SJ, Roy S, Patel PK (2007) Distraction osteogenesis in the pediatric population. *Otolaryngol Head Neck Surg* 137(2):233–238
17. Roy S, Munson PD, Zhao L, Hollinger LD, Patel PK (2009) CT analysis after distraction osteogenesis in Pierre Robin sequence. *Laryngoscope* 119(2):380–386
18. Mohamed AM, Al Bishri A, Mohamed AH (2011) Distraction osteogenesis as followed by CT scan in Pierre Robin sequence. *J Cranio-maxillofac Surg* 39(6):412–419
19. Cope JB, Samchukov ML, Cherkashin AM (1999) Mandibular distraction osteogenesis: a historic perspective and future directions. *Am J Orthod Dentofac Orthop* 115(4):448–460
20. Cademartiri F, Luccichenti G, Lagana F, Brevi B, Sesenna E, Pavone P (2004) Effective clinical outcome of a mandibular distraction device using three-dimensional CT with volume rendering in Pierre Robin sequence. *Acta Biomed* 75:122–125
21. Rhee ST, Buchman SR (2003) Pediatric mandibular distraction osteogenesis: the present and the future. *J Craniofac Surg* 14:803–808
22. Tibesar RJ, Price DL, Moore EJ (2006) Mandibular distraction osteogenesis to relieve Pierre Robin airway obstruction. *Am J Otolaryngol* 27:436–439
23. Burstein FD, Williams JK (2005) Mandibular distraction osteogenesis in Pierre Robin sequence: application of a new internal single-stage resorbable device. *Plast Reconstr Surg* 115:61–67
24. Burstein FD (2008) Resorbable distraction of the mandible: technical evolution and clinical experience. *J Craniofac Surg* 19(3):637–643
25. Parashar SY, Anderson PJ, David DJ (2006) An unusual complication of mandibular distraction. *Int J Pediatr Dent* 16:55–58
26. Schaefer RB, Stadler JA III, Gosain AK (2004) To distract or not to distract: an algorithm for airway management in isolated Pierre Robin sequence. *Plast Reconstr Surg* 113:1113–1125
27. Issar Y, Sahoo NK, Sinha R, Satija L, Chattopadhyay PK (2014) Comparative evaluation of the mandibular distraction zone using ultrasonography and conventional radiography. *Int J Oral Maxillofac Surg* 43(5):587–594