Mandibular distraction osteogenesis for infants with Pierre Robin Sequence and airway obstruction

Yuk-Ping TSANG 曾玉萍,* Wa-Keung CHIU 趙華強,1 Genevieve Po-Gee FUNG 馮寶姿,1 Lilian Po-Yee LEE 李寶儀,1 Siu-Chung FUNG 冯少聰,2 Victor ABDULLAH 石偉棠,3 Hin-Biu CHAN 陳衍標1
1Department of Paediatrics and Adolescent Medicine; 2Department of Dentistry & Maxillofacial Surgery; 3Department of Otorhinolaryngology, Head & Neck Surgery, United Christian Hospital, Hong Kong

Abstract
Pierre Robin Sequence in newborns usually presents with respiratory distress soon after birth. Various treatment modalities exist all aiming to relieve the underlying upper airway obstruction. We describe a newborn with Pierre Robin Sequence initially treated with tracheostomy and successfully decannulated shortly after mandibular distraction osteogenesis. Good outcome was able to achieve in both respiratory and feeding aspects.

Keywords: Mandibular distraction osteogenesis, Pierre Robin Sequence

Introduction
Pierre Robin Sequence (PRS) comprises of a triad of micrognathia, glossoptosis and the resultant airway obstruction. Infants with PRS present with signs of respiratory distress after birth and are usually first treated with non-invasive measures like prone-positioning and nasopharyngeal airway with or without continuous positive airway pressure. However, about one third of these children failed conservative measures and required surgical treatment. Tracheostomy has long been used as the most definitive method to secure the airway but is related to a long list of complications. We describe a male infant having PRS with severe upper airway obstruction initially treated by tracheostomy. Bilateral mandibular distraction osteogenesis (MDO) was subsequently performed and he was successfully decannulated shortly afterwards. We will further discuss about the potential role of MDO as an alternative to conventional surgical methods in managing infants with severe airway obstruction resulting from PRS.

Case presentation
A male neonate, weighing 1710 g, was born at gestation of 34 4/7 weeks by spontaneous vaginal delivery because of premature rupture of membrane and severe maternal pre-eclampsia. The Apgar score was 6 and 7 at one and five minutes, respectively. Antenatal ultrasound revealed severe micrognathia as well as one artery and one vein in umbilical cord but baby's mother refused amniocentesis. Baby was found to be flaccid and bradycardiac with poor respiratory efforts at birth. Heart rate and colour improved upon bagging via face mask but respiratory distress persisted. Intubation failed at the labour ward as the vocal cords could not be visualised on direct laryngoscopy. The baby was escorted to neonatal intensive care unit with continuation of bagging via face mask. Physical examination revealed low set ears, U-shaped central cleft palate, large bulky tongue with glossoptosis and severe micrognathia. Examination of other systems was normal. The clinical picture was compatible with Pierre Robin sequence with upper airway obstruction caused by glossoptosis. Baby was put in prone position and started on nasal continuous positive airway pressure (CPAP) support. He responded well initially but developed worsening of respiratory distress on day four of life with carbon dioxide retention. Laryngeal mask was applied immediately. Attempted intubation by anaesthetist was unsuccessful and fibreoptic assisted intubation by Ear, Nose and Throat (ENT) team was performed. Baby remained stable on low setting of ventilatory support. Subsequent ENT assessment revealed severe glossoptosis with no view of epiglottis by Benjamin slotted laryngoscope and suspension scope. Fibreoptic scope showed normal larynx. Difficult airway was anticipated. Screening for associated anomalies including echocardiogram, cranial
Further growth of maxilla restored the facial profile. He remained well on subsequent follow up.

The major problem encountered by these children is upper airway obstruction caused by displacement of the tongue into the hypopharynx, causing occlusion of the airway at the level of epiglottis. Other mechanisms leading to upper airway obstruction include tongue prolapsing into the cleft palate, disproportionate tongue growth, lack of voluntary control of tongue musculature, negative pressure pull of the tongue into the hypopharynx as well as midface hypoplasia. Feeding difficulties were also common in PRS. Glossoptosis and upper airway obstruction prevent normal swallowing and these children often require tube feeding or even gastrostomy feeding to maintain adequate nutritional intake. Without proper treatment, mortality will be high from respiratory failure and malnutrition. Although the mandible has significant growth potential in postnatal period and airway obstruction usually improves with time, most of the PRS infants require some sort of treatment after birth. Approach to management comprises of widening of pharyngeal space (by prone positioning, tongue lip adhesion and mandibular distraction), bridging or stenting the obstructed airway (by nasopharyngeal airway, continuous positive airway pressure and tracheostomy) as well as correction of glossoptosis by palatal plate or pre-epiglottic Batson plate. Conservative measures like prone positioning, placement of nasopharyngeal airway with or without the use of CPAP are usually tried first. However about 35% of infants with isolated PRS will subsequently require surgical treatment with tongue-lip adhesion and tracheostomy being the two most commonly used surgical procedures. The reported successful rate is 33-100% for tongue lip adhesion and it usually requires further interventions. Tracheostomy is long considered as the definitive method to secure a stable airway in infant with severe upper airway obstruction. However, long term tracheostomy is associated with a long list of complications which include sudden airway obstruction from mucous plugging and accidental decannulation, formation of suprastormal granuloma with airway bleeding, airway infection and inhibition of normal swallowing and speech development. It also requires meticulous nursing care and may delay progression to home care. Although being a valid life-saving procedure, it interferes with child care, parental bonding as well as daily life and should be considered as the last treatment resort because of its high morbidity and mortality rate. The latter was up to 5%. Even in optimal cases, it could take two to four years to safely decannulate the child.

To solve these problems, a new surgical technique called mandibular distraction osteogenesis was introduced. Distraction osteogenesis was first introduced in Bologna, Italy by Codivilla in 1905. Clinical application of distraction osteogenesis in human mandible was first performed in America by McCarthy in 1992, involving children between 2 to 11 years old with craniofacial abnormalities. The first local case was
performed in a girl with Treacher Collins syndrome in 1997 by Prof LK Cheung. MDO is a technique which aims to lengthen the mandible gradually and hence the tongue and the suprahypophysis muscles were brought forward, thereby increasing the size of the pharyngeal airway and relieving the upper airway obstruction. It involves five phases of treatment. The first phase comprises of osteotomy and insertion of distractors which is followed by a short latency period. Then distraction is carried out at a slow pace, allowing concomitant expansion of the soft tissue with related muscles, blood vessels, nerve fibers so that the lengthened bone can be accommodated. Consolidation phase subsequently begins, allowing the bone segments to be secured in the newly advanced position. The final phase consists of the removal of the distractors. A meta-analysis published in 2008 reviewed a total of 646 patients with bilateral MDO demonstrated a successful rate of 91.3% in preventing tracheostomy in neonates and infant with severe upper airway obstruction. 78.4% of patients with existing tracheostomies were successfully decannulated after MDO.

Further reports studying the effect of MDO as an alternative to tracheostomy in neonates with severe upper respiratory obstruction were published since the previous meta-analysis. They all showed favourable outcomes in term of increasing airway cross-sectional area, decreasing in desaturations with improved apnoea hypoxia index, facilitating oral feeds and hospital discharge. A study by Tibesar et al demonstrated that the airway improvement was sustained in long term with 88% of patients remained free of tracheostomy at long term follow up.

Timing to execute MDO is a debatable subject. There are concerns about if MDO needed to be revised and repeated later on if the procedure was performed on the very young neonate in case the mandible did not grow with the child. The study by Tiebesar et al showed that the younger group of patient (MDO at age less than 3 months) actually performed better than the group of children distracted at an older age. Other studies also demonstrated significant airway improvement with MDO done in neonatal period. Early MDO also may avoid tracheostomy and its related complications and allow early discharge to home. Young age alone should not be the reason declining MDO in this age group provided that there is adequate expertise. However, our case is one of the first few patients that had MDO performed in our hospital, we adopted a conservative approach of first doing a tracheostomy then followed by MDO.

Figure. Clinical photographs (from left to right) showing the stereo-model of mandibular distraction osteogenesis (MDO), facial profile and CT neck of the patient, respectively. Upper row: before MDO; lower row: after MDO.
Although the impact is less dramatic than that of airway obstruction, MDO also has positive effect on overcoming feeding difficulties in children with micrognathia. Relieving airway obstruction allows easier coordination between breathing and swallowing. Distracted children showed improved oral feeding and less dependence on tube feeding in many studies.22,26

Long term complications of MDO include anterior open bite deformity, permanent dental loss/malformation/dentigerous cyst formation, long term facial nerve injury, temporomandibular joint ankylosis and hypertrophic scarring at the pin sites.22,26,27 The incidence of these complications was relatively low and would be prevented by careful preoperative planning and modeling22 and MDO can be considered as a safe procedure for mandibular lengthening in patient with craniofacial abnormalities.26

To conclude, MDO was shown to have significant and sustained airway improvement on children with upper airway obstruction caused by micrognathia. There was also improved outcome on oral feeding. Results of early MDO in neonatal period were encouraging. Together with the relatively low complications and relapse rate, MDO provides an alternative to conventional surgical methods in managing infant with moderate to severe upper airway obstruction secondary to micrognathia as in the case of Pierre Robin Sequence.

References