

JAW/ MANDIBULAR DISTRACTION OSTEOGENESIS MANAGEMENT IN NEONATES

PRACTICE GUIDELINE[®]

DOCUMENT SUMMARY/KEY POINTS

- Mandibular Distraction Osteogenesis is an operative technique that involves bilateral osteotomies of the mandible followed by daily distraction or lengthening of the mandible.
- This surgical procedure can be used in the management of neonates with Pierre Robin Sequence and severe obstructive airway disease to prevent a tracheostomy.
- The goals of MDO are to increase the size of the mandible and improve airway obstruction, relieving any breathing difficulties.
- Post-operatively extra care must be taken with the airway as the neonate is muscle relaxed/sedated.
- Wound care is essential to prevent a post-operative infection.
- Key Performance Indicators (KPI):
 - No post-operative infection
 - Adequate pain management during distraction of drive screw procedure
 - Improve airway obstruction by reducing level of respiratory support required

This document reflects what is currently regarded as safe practice. However, as in any clinical situation, there may be factors which cannot be covered by a single set of guidelines. This document does not replace the need for the application of clinical judgement to each individual presentation.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1 st February 2017	Review Period: 3 years
Team Leader:	Nurse Educator	Area/Dept: GCNC

CHANGE SUMMARY

- Complications Following a Mandibular Distraction
- Addition of Appendix 1 Jaw Distraction Flow Sheet.
- Addition of Appendix 2 to provide Record of Pin Distraction.

READ ACKNOWLEDGEMENT

- To be read and acknowledged by all nursing and medical staff working in Grace Centre for Newborn Care.

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Introduction

Jaw/Mandibular Distraction Osteogenesis (MDO) is an operative technique used to improve upper airway obstruction in neonates with Pierre Robin Sequence (PRS). The procedure (Mandibular Distraction Osteogenesis) involves a bilateral osteotomy of the mandible followed by daily distraction or lengthening of the mandible in the NICU.^{3,7} Lengthening of the mandible brings the tongue forward, preventing it from occluding the airway.⁹ Jaw distraction is most commonly required in infants with Pierre Robin Sequence, which is identified as a combination of micrognathia and glossoptosis with evidence of mild to severe respiratory distress.^{1,5} Most neonates with PRS will also have a cleft palate,³ as a result of the tongue blocking the fusion of the palate and forming a U-shaped palate^{2,5} as well as micrognathia and glossoptosis. Micrognathia is a very small or recessed mandible.^{3,4,8} Glossoptosis is a retro position of the tongue which obstructs the upper airway.^{3,8}

In the past neonates with severe airway obstruction have been treated with a tracheostomy^{3,4,5} or a nasopharyngeal airway necessitating a prolonged hospital stay. A tracheostomy is associated with swallowing difficulties, speech and language problems,^{3,7} a risk of decannulation and/or death. There is also significant financial and psychological impact on the family.³ Mandibular Distraction Osteogenesis is used as an alternative to tracheostomy, thereby reducing long term complications for families caring for an infant with a tracheostomy.^{4,7}

Background

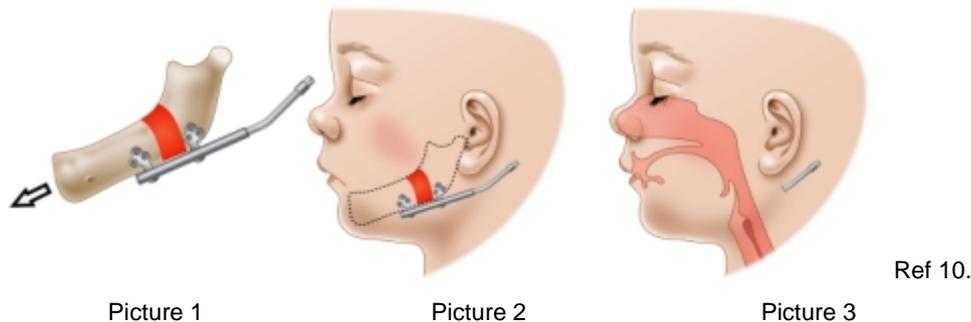
- Pierre Robin Sequence (PRS) can be isolated or associated with other disorders such as Treacher Collins syndrome, Stickler syndrome, velocardiofacial syndrome, fetal alcohol syndrome and Nager syndrome. Isolated PRS is caused by intrauterine forces on the mandible.
- The degree of respiratory obstruction depends on the severity of the micrognathia and glossoptosis.⁴
- Breathing difficulties caused by airway obstruction may lead to repeated periods of oxygen desaturations resulting in cyanotic episodes,¹ and obstructive sleep apnoea with requirement for home CPAP.
- Breathing and feeding difficulties are caused by the position of the tongue.¹⁰ This often results in failure to thrive of the neonate.⁸
- Glossoptosis prevents a normal swallow and infants will require feeding through an intragastric tube.⁸

Investigations and Management after admission to the NICU

- On admission to the NICU the neonate's airway is assessed for signs of obstruction and increased work of breathing. If the obstruction is mild, repositioning the infant or a nasopharyngeal airway is inserted to relieve symptoms. If the obstruction is moderate to severe further tests such as a sleep study and laryngoscopy, bronchoscopy & oesophagoscopy (LBO) will be undertaken.
- Polysomnography (sleep study) is performed to determine if the breathing pattern has central (neurological) or obstructive (functional) episodes.^{1,3,8,10}
- Facemask CPAP can be trialled to overcome obstructive airway disease.^{1,3,4} If the trial of CPAP is unsuccessful and /or symptoms of airway obstruction persist, then discussions are held with the speciality teams about proceeding to an MDO.
- An LBO is performed prior to making the final decision to undertake an MDO to rule out associated airway anomalies.
- A CT scan or MRI may be performed to provide exact measurements of the mandible. This allows accurate planning for the osteotomies and placement of the screws. Tooth buds are also identified to avoid future complications.^{8,10}
- Neonates are referred for genetics review. Chromosomal testing maybe performed.
- Further investigations and consultations are performed as required, to investigate other anomalies. These investigations include ophthalmology review, head, cardiac and renal ultrasounds and skeletal surveys.
- A multidisciplinary team, including Sleep, Ear, Nose and Throat Surgeons, Plastics, Cleft/Speech, Neonatology & Nursing, will assess whether the neonate requires the MDO procedure.¹⁰ (See Appendix 1, Jaw distraction flow sheet).
- Results of the investigations are discussed in a multidisciplinary meeting with the parents, where the potential surgical options are discussed i.e. MDO or a tracheostomy.

Procedure

- Mandibular Distraction Osteogenesis is a surgical procedure where bilateral osteotomies are created on the mandible, followed by daily distraction (turning) of the implanted screws to lengthen the size of the mandible.^{3,4,6,7,9,11}
- Usually resorbable plates and screws are used which reduce the need for further surgery and are resorbed within 12 months.¹ However there may be occasions where devices used are not resorbable and require removal in an operating theatre.
- Glossopexy (lip-tongue adhesion) may also be performed. During this procedure the tongue is secured to the lower lip to prevent the tongue from obstructing the palate and occluding the airway.^{1,2}



Picture 1: Bilateral osteotomies are created. **Picture 2:** Resorbable plate and screws applied. **Picture 3:** Drive screws are brought through the skin superior or inferior to the ears.

Goal of Mandibular Distraction Osteogenesis

- To lengthen the size of the mandible and prevent airway obstruction and relieve any breathing and feeding difficulties.^{4,6,7}

Pre-Operative Preparation

- The neonate is prepared for surgery in accordance to the CHW Guideline No: 0/C/08:0000-01:01 [Transfer of a neonate to operating theatre and other hospital investigative departments](#)
- Perform two washes of the jaw and behind the ears prior to surgery using triclosan 1%.

Post-Operative Management

- On return to the NICU a full head-to-toe assessment and examination of the neonate is conducted by both medical and nursing staff, including a pain assessment and temperature.
- A chest x-ray is undertaken to confirm the position of the ETT.
- Arterial blood gases and blood sugar level are taken within one hour and repeated as requested by the medical officer.

- FBC & EUC are analysed on return.
- Head of the bed is elevated 30 degrees to minimise facial swelling.

Ventilation

- Following MDO the neonate will remain intubated and ventilated for the first few days until an airway can be established and any swelling has decreased.⁷
- Neonates are given analgesia, sedated and/or muscle relaxed for 24-48 hours to establish adequate pain management and protect the surgical sites from excessive movement.
- Care for the ventilated neonates is in accordance to the CHW Practice Guideline No:0/C/07:0005-01:02 [Respiratory Support in Neonates](#)
- When post-operative swelling has reduced the neonate will return to the operating theatre for an airway assessment and trial extubation.^{3,7}

Nasal and Oral Pharyngeal Suction

- Care is taken when attending oral suctioning if a lip-tongue adhesion has been attended. Suctioning under the tongue should be avoided to prevent damage to the sutures around the tongue and lip.
- Suction is attended according to the technique in the CHW Practice Guideline 0/C/12:0002-01:00: Endo-Tracheal Suctioning in Neonates

Infection

- This is the most common complication of MDO⁸ and so meticulous hand-washing is required when attending the neonate and during the distraction procedure.
- Topical antibiotic ointment is prescribed by the surgeons and applied to the drive screw sites liberally to prevent infection.
- Topical antibiotic treatment will continue until the drive screws are removed approximately 6 weeks after distraction is complete.

Pressure Area Care

- Drive screws are to be stabilised and secured with dressings to prevent movement, leading to pressure areas or dislodgement.
- Protective padding (gauze) is to be placed behind the drive screws to prevent pressure areas on the scalp caused by the screws.
- Repositioning of the neonate is undertaken every 3-4 hours to avoid pressure areas with care being taken to avoid lying on distraction screws.
- The neonate should be nursed on a pressure reducing mattress.

- While the neonate is intubated and ventilated repositioning must be undertaken with the assistance of a second nurse.

Wound Care

- Steri-strips are applied to jaw incisions. These sites are regularly assessed for signs of bleeding or infection (swelling, redness, discharge, fever, discharge or pain).
- Drive screw sites are observed for redness, swelling or bleeding.¹¹
- Drive screw sites are cleaned at least daily with normal saline and topical antibiotic ointment applied.
- Medical officers are to be informed of any signs of infection.¹¹

Pain Management

- Pain is assessed on return to the NICU and at least every 4th hour. If the score is above 5, consideration is given to starting or increasing an opioid infusion. Results are recorded in CCIS.
- Pain is managed as outlined in the CHW Practice Guideline No: 0/C/06:0027-01:01 [Pain management in newborn infants](#)

Fluid and Nutrition

- Total Parenteral Nutrition is administered post operatively and continues until full feeds have been established.¹¹
- Infants are weighed three times a week (once extubated) to enable their fluids to be calculated accurately to encourage optimal growth.

Feeds

- Neonates are not fed enterally in the immediate post-operative period.
- Feeds are slowly commenced through an intragastric tube and are graded up as tolerated.⁷
- When full intragastric tube feeds are tolerated sucking feeds may be introduced cautiously. Most of these neonates have not fed orally prior to MDO and so will require a formal feeding assessment by the speech pathologist before commencing oral feeds.

Developmentally Supportive Care

- Neonates who have undergone the MDO with regular distractions require a supportive environment and need to be positioned and wrapped so that they are able to self-regulate their behaviours in response to the stressful interventions.

- Details of various strategies are found in the CHW Practice Guideline No: 0/C/06:0027-01:01 [Developmentally supportive care for newborn infants](#)
- Support the parents to undertake position changes and nappy cares. When the infant is stable a cuddle can be offered allowing parents the opportunity to engage with their baby.

Distraction

- Distraction of the drive screws is performed daily by the plastics team. A distraction driver is used to perform the distraction and this device is left at the neonates bedside and should remain with the neonate at all times.
- Distraction commences approximately two days after the initial procedure as directed by the operating surgeon.^{3,6,7,10}
- Distraction will be between 0.5-1mm each side per day until desired lengthening has been achieved as per ENT/Plastics team.^{3,6,10}
- Parents may be educated by the plastics team to perform daily distractions. Approval to go home during distraction phase may be given by the plastics team.
- Sucrose is administered prior to the distraction in accordance with the CHW Practice Guideline No: 1/C/06:8241-01:02 [Sucrose: Management of short duration procedural pain in infants](#)

Removal of Drive Screws

- The drive screws are removed by the plastics team in the NICU/Outpatient department or in some cases the neonate will be taken to the operating theatre for removal.¹⁰ This occurs approximately 6-10 weeks after distraction is complete to allow for bone consolidation.^{3,6,11}
- Steri-strips are applied when distraction screws are removed.
- If removed in the NICU sucrose is administered prior to removal in accordance with the CHW Practice Guideline No: 1/C/06:8241-01:02 [Sucrose: Management of short duration procedural pain in infants](#)

Discharge Planning

- A repeat sleep study is performed several weeks after the distraction is complete and prior to discharge to assess the success of the procedure. Some infants may still require home CPAP after having a MDO.
- Assessments are performed to gauge the success of the procedure. This includes rate of growth as studies have shown that successful distraction results in healthy growth and development^{3,4} through improvement in feeding.^{3,9}

- Parents will receive adequate education and training and be deemed competent in the care of distraction pins before patient is discharged.

Possible Complications Following a Mandibular Distraction

Short Term	Long Term
Surgical site infection	Open bite deformity
Cellulitis	Tooth loss or malformation
Soft tissue dehiscence	Temporomandibular joint ankylosis
Facial nerve injury	Scarring
Device failure requiring replacement	Permanent facial nerve palsy
Pin loosening	
Incomplete osteotomy/premature ossification	

Ref 12,13,14

References

1. Cheng, A.T.L., Corke, M., Loughran-Fowlds, A., Birman, C., Hayward, P. & Waters, K. A. 2011 Distraction osteogenesis and glossopexy for robin sequence with airway obstruction. *ANZ Journal of Surgery* 81, 320-325.
2. Papoff, P., Guelfi, G., Cicchetti, R., Caresta, E., Cozzi, D.A., Moretti, C., Midulla, F., Miano, S., Cerasaro, C., Cascone, P. 2013 Outcomes after tongue-lip adhesion or mandibular distraction osteogenesis in infants with Pierre Robin sequence and severe airway obstruction. *International Journal of Oral and Maxillofacial Surgery* 42, 1418-1423.
3. Hong, P. 2011 A clinical narrative review of mandibular distraction osteogenesis in neonates with Pierre Robin sequence. *International Journal of Pediatric Otorhinolaryngology* 75, 985-991.
4. Scott, A.R., Tibesar, R.J., Lander, T.A., Sampson, D.E. & Sidman, J.D. 2011 Mandibular distraction osteogenesis in infants younger than 3 months. *ARCH Facial Plast Surg* 13, 3, 173-179.
5. Greathouse, S.T., Costa, M., Ferrera, A., Tahiri, Y., Tholpady, S.S., Havlik, R.J., Flores, R.L. 2015 The surgical treatment of robin sequence. *Annals of Plastic Surgery* 00: 00-00.
6. Breik, O., Tivey, D., Umaphysivam, K., Anderson, P. 2016 Does the rate of distraction or type of distractor affect the outcome of mandibular distraction in children with micrognathia? *Journal of Oral Maxillofacial Surgery* 1-13.
7. Tsang, C., Adil, E., Scott, A.R. 2015 Neonatal mandibular distraction osteogenesis. *Operative Techniques in Otolaryngology* 26, 131-135.
8. Meyers, A.B., Zei, M.G., Denny, A.D. 2015 Imaging neonates and children with pierre robin sequence before and after mandibular distraction osteogenesis: what the craniofacial surgeon wants to know. *Pediatric Radiology* 45, 1392-1402.
9. Seattle Children's Hospital. Pierre Robin Sequence. No date. Accessed via <http://www.seattlechildrens.org/medical-conditions/chromosomal-genetic-conditions/pierre-treatment/> <Accessed 2012, May 14>
10. The Royal Children's Hospital Melbourne. Kids Health Info. Jaw distraction (mandibular distraction osteogenesis). 2012. Accessed via http://www.rch.org.au/kidsinfo/factsheets.cfm?doc_id=15347 <Accessed 2012, May 14>
11. Pierre Robin Network. Jaw Distraction. No date. Accessed via <http://www.pierrerobin.org/jaw-distraction.htm> <Accessed 2012, May 14>
12. Paes, E.C., Mink van der Molen, A.B., Muradin, M.S.M., Speleman, L., Sloot, F., Kon, M., Breugem, C.C. 2013 A Systematic review on the outcome of mandibular distraction osteogenesis in infants suffering Robin sequence. *Clinical Oral Investigations* 17, 1807-1820.
13. Verlinden, C.R.A., van de Vijfeijken, S.E.C.M., Jansma, E.P., Becking, A.G., Swennen, G.R.J. 2015 Complications of mandibular distraction osteogenesis for congenital deformities: a systematic review of the literature and proposal of a new classification for complications. *International Journal of Oral and Maxillofacial Surgery* 44, 37-43.
14. Flores, R.L. 2014 Neonatal mandibular distraction osteogenesis. *Seminars in Plastic Surgery* 28, 199-206.

Appendix 1

Jaw Distraction Flow Sheet

