

Mandibular Distraction Osteogenesis in the Management of Upper Airway Obstruction

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3/30/20

Objectives

- Describe strategies to **evaluate airway obstruction** in a micrognathic child
- Discuss an algorithm for the **management of airway obstruction** in a child with Robin Sequence
- Present **evidence-based risks and benefits of mandibular distraction osteogenesis (MDO)** in the management of symptomatic glossoptosis and upper airway obstruction

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Evaluation of airway obstruction of 3d in NICU

- NICU team has observed
 - Stertor and obstruction when supine associated with desats to low 80's
 - Relief of obstructive symptoms in lateral and prone position



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Pierre Robin Sequence

- Micrognathia with clinically significant glossoptosis
 - Airway obstruction
 - Wide, U-shaped cleft palate



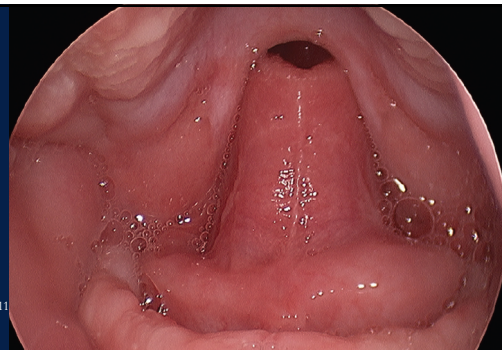
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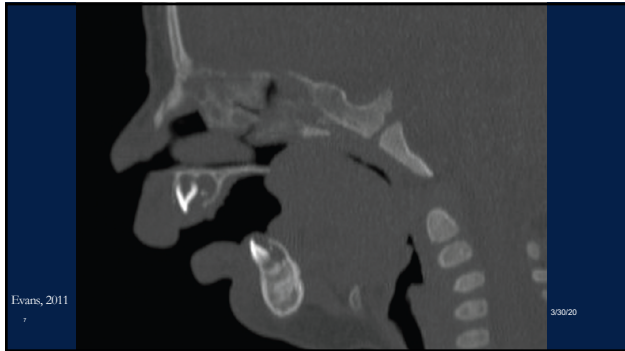
Evans, 2011

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Evans, 2011

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Pierre Robin Sequence

Epidemiology

- Approximately 1 in 8500 live births
- Association with Stickler, 22q11.2 deletion, Treacher–Collins, Nager
- Most commonly seen in isolation

Evans et al. 2011

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Pierre Robin Sequence

Other clinical signs and symptoms

- Feeding difficulties, FTT
- CO2 retention
- Heart failure
- Delayed neurocognitive development
- mortality of 1.7% to 65%

Evans et al. 2011

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Pierre Robin Sequence

Evaluation of airway obstruction

- Flexible laryngoscopy
- Empirical titration of respiratory support with prone/lateral positioning
- Direct laryngoscopy, bronchoscopy
- Capillary blood gas (PCO2)
- +/- polysomnography

Evans et al. 2011

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Airway Evaluation: Flexible Laryngoscopy

Credit: Skarke

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Airway Evaluation: Direct Laryngoscopy, Bronchoscopy

- Retrospective review of 22 patients with failed MDO (Breik, 2016)
 - 15/22 undiagnosed concomitant airway anomalies
 - Laryngomalacia, tracheal stenosis, tracheomalacia, or subglottic stenosis
- Retrospective review of 55 MDO patients (Andrews, 2013)
 - laryngomalacia (53.3 percent)
 - tracheal web (20.0 percent)
 - vocal cord paralysis (13.3 percent)
 - epiglottal collapse (6.7 percent)
 - infraglottal narrowing (6.7 percent)
- Rigid bronchoscopy may not be possible

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Pierre Robin Sequence

Other targets of evaluation

- pH probe (GERD)
- Echocardiogram (pulm HTN, shunting)
- Genetic evaluation (COL mutations, FISH)
- Brain imaging (IVH, holoprosencephaly)



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Pierre Robin Sequence

Conservative management of airway obstruction (70-90% effective)

- prone positioning (49% effective)
 - Parent education
 - GERD precautions



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Pierre Robin Sequence

Conservative management of airway obstruction (70-90% effective)

- prone positioning (49% effective)
- nasal continuous positive airway pressure (CPAP)



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Pierre Robin Sequence

Conservative management of airway obstruction (70-90% effective)

- prone positioning (49% effective)
- nasal continuous positive airway pressure (CPAP)
- nasopharyngeal airway (effective 48.8-100%)



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Nasopharyngeal Airway

- Effective non-surgical means of bypassing tongue base obstruction
- 85.7% require feeding tube (Parhizkar et al, 2011)
- Technically simple
 - Verify position via fiberoptic laryngoscopy or lateral neck film
 - Change weekly, can be used for months
 - Dispo, not simple



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| First author and year | Design | N | Mean age of the start of treatment | Indication | Primary outcome (1 = 50/50, 2 = AHI, 3 = mortality) | Complication rate | MINORS |
|-----------------------------------|---|----|--|---|--|---|--------|
| Chang et al. (2000) | Retrospective study design | 6 | N/A | N/A | 1. 100% (all) through 4 respiratory supplemental oxygen. Mean follow-up duration not reported. 2. N/A | During the initial period 2 children had respiration of feeding | 4 |
| Wilson et al. (2000) | Retrospective study design (date obstruction) | 7 | N/A | Diffused, but in general poor weight gain and desaturations to 80% | 1. Not reported 1. 67% (6/7) in follow-up duration (not reported) 2. N/A | None reported | 3 |
| Wagner et al. (2003) | Case series | 20 | N/A | Un satisfactory oxygen saturation and weight gain (moderate obstruction) or in case of severe airway obstruction at first | 1. Not reported 1. 100% (n=low-up duration) (not reported) 2. N/A | 25% | 7 |
| Anderson et al. (2007) | Retrospective study design | 13 | N/A but reported to be close to the median age of admission (0-122 days) | Un satisfactory oxygen saturation and weight gain (moderate obstruction) or in case of severe airway obstructive at rest. | 1. 100% (6/6) in follow-up duration (not reported). 2. N/A 3. None reported | None reported | 5 |
| Parhizkar et al. (2011) | Retrospective case series | 18 | N/A (no specific data for RS available) | N/A (no specific data for RS available) | 1. N/A (no specific data for RS available) 2. N/A | None reported | 5 |
| (link in milk) Abel et al. (2012) | Retrospective study design | 63 | N/A (Varied from 1 to 330 days) | Sleep study showing moderate to severe UAO (According to Nooni's criteria), moderate UAO for a set of at least three clusters of desaturations with at least 3 dips below 85% (but not below 80%) and severe UAO for a set of at least three clusters of desaturations with atleast 3 dips below 80%. | 1. UAO (up) for RS population 1. 81.8% (n=12) in a median follow-up period of 12 months (range 2-30 months) 2. N/A 3. None | None reported | 6 |

VanLeishout et al., 2016

Surgical Management of Airway Obstruction

- Tongue Lip Adhesion
- Tracheotomy
- Mandibular distraction osteogenesis



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Tongue Lip Adhesion (TLA)

- Surgically tethering ventral surface of the tongue to the oral surface of the lower lip.
- May be combined with subperiosteal release of the floor of mouth
- Typically reversed at age 9-12 months or at the time of palate closure



Morrow et al., 2015

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A Systematic Review of the Effectiveness of Tongue Lip Adhesion in Improving Airway Obstruction in Children With Pierre Robin Sequence

Alex Vitez-Mathieu, MD, BSc,* Tyler Safran,[†] and Mirko S. Gilardino, MD, MSc*
(*J Craniofac Surg* 2016;27: 1453–1456)

- Retrospective review of 13 studies, 268 patients
- Relief of airway obstruction in 81.3%
 - Improved O2 sat or AHI
 - Avoidance of further airway intervention
 - Decannulation
- 12.3% required revision procedure, almost all trach



Morrow et al., 2015

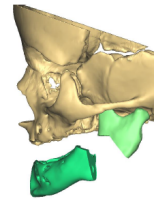
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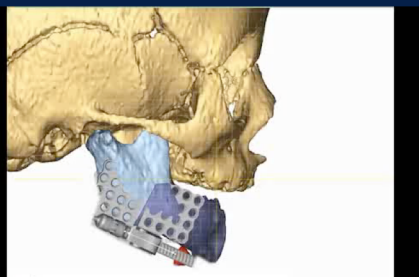
Mandibular Distraction Osteogenesis (MDO)

- Mandible is divided and distraction device placed across the osteotomies
- Unlike conventional orthognathic surgery
 - Incremental traction on bony callus
 - Potential for more skeletal elongation
 - Allows for adaptive changes to soft tissue envelope



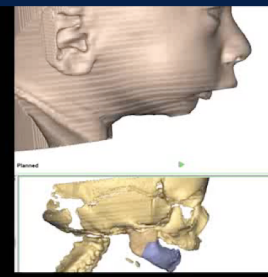
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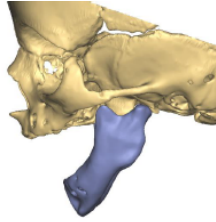
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Preoperative Evaluation

- Microlaryngoscopy, bronchoscopy
- CT scan without contrast
 - 3D reconstruction
 - Virtual surgical planning



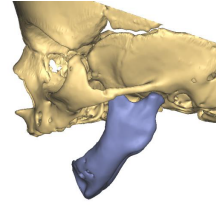
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Preoperative Evaluation

- Microlaryngoscopy, bronchoscopy
- CT scan without contrast
 - 3D reconstruction
 - Virtual surgical planning



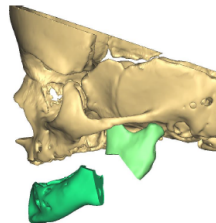
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Preoperative Evaluation

- Microlaryngoscopy, bronchoscopy
- CT scan without contrast
 - 3D reconstruction
 - Virtual surgical planning



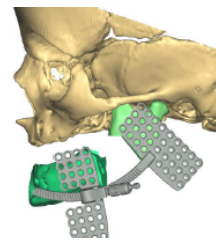
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Preoperative Evaluation

- Microlaryngoscopy, bronchoscopy
- CT scan without contrast
 - 3D reconstruction
 - Virtual surgical planning



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Osteotomy and Placement of Plates

- 2 cm incision beginning inferior and anterior to the angle, parallel to the mandible
- Dissect deep to the fascia investing the submandibular gland and reflect it up, follow it to the mandible.
- Incise the pterygomasseteric sling to expose the mandible
- L shaped osteotomies, protect IAN



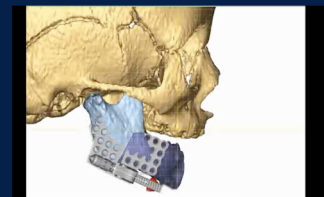
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Distraction Protocol

- Latency period (1-5 days, shorter in infants)
- Active distraction (1-2 mm/day)
- Consolidation (4-8 weeks)



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Relief of Airway Obstruction in MDO

- Poor standardization of outcomes across studies
 - Polysomnography
 - Relief of obstructions
 - Avoidance of trach



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Systematic Review and Meta-Analysis **Medicine** OPEN

Distraction osteogenesis as a treatment of obstructive sleep apnea syndrome
A systematic review

Wai Kin Tsui, BDS, MDS(OMS), MOS RCSEd, FRCRCS, FCDShK(OMS), FHKAM(Dental Surgery)[®],

- 12 retrospective case series, 9 with children, 8 included syndromic patients
- In adult studies, PSG data was available
- In children without PSG data, success rates based on absence of obstructive symptoms, avoidance of tracheotomy, or decannulation

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Systematic Review and Meta-Analysis **Medicine** OPEN

Distraction osteogenesis as a treatment of obstructive sleep apnea syndrome

- Overall success rate ranged from 90% to 100%.
- Success rate for the adult group was 100%, while the cure rate was from 82% to 100%
- Adult group, the mean AHI/RDI changed from 51.7/h (43.0–58.0/h) preoperatively to 2.9/h (2.1–3.4/h) postoperatively
- Child/infant group, the mean preoperative AHI/RDI ranged from 10 to 50/h and was reduced to 1.1 to 5/h

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Airway and Feeding Outcomes of Mandibular Distraction, Tongue-Lip Adhesion, and Conservative Management in Pierre Robin Sequence: A Prospective Study

Khansa I, 2017.

| | Initial AHI | Post-intervention AHI |
|---------------------|-------------|-----------------------|
| Observation | 6.1 | 5.5 |
| Tongue Lip Adhesion | 15.2 | 2.8 |
| MDO | 27.7 | 1.5 |

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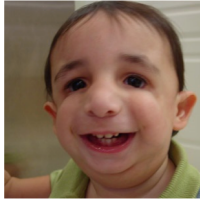
Airway and Feeding Outcomes of Mandibular Distraction, Tongue-Lip Adhesion, and Conservative Management in Pierre Robin Sequence: A Prospective Study

- In MDO group
 - 0% had residual moderate OSA
 - Palate repaired at 11.7 mo
- In TLA group
 - 12.5% had residual moderate OSA
 - Palate repaired at 14.9 mo

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Syndromic vs. Non Syndromic

- Meta-analysis of success rates in 711 MDO patients (Tahiri et al, 2014)
 - Isolated PRS (97.6%)
 - Syndromic PRS (94.0%)
- Meta-analysis of 490 MDO patients (Breik et al, 2016)
 - Isolated PRS (97.6)
 - Syndromic PRS (90.7) [p=0.007]

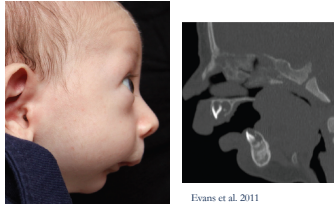


<http://www.health24.com>

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Feeding Difficulty in PRS

- 50 percent with FTT requiring non-oral feeding
 - NGT (51%)
 - GT(19%)
 - NGT then GT (30%) (Meyer et al., 2008)



Evans et al 2011

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The Laryngoscope
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CANDIDATE THESIS

Regional Variations in the Presentation and Surgical Management of Pierre Robin Sequence

Andrew R. Scott, MD, FACS; Nicholas S. Mader, PhD

Rate of Associated Diagnoses Comparison Among iPRS, sPRS, and All Newborns, 2006 and 2009.

| | All Newborns | iPRS | sPRS | P Value, Test of iPRS Mean = All Newborns Mean | P Value, Test of sPRS Mean = All Newborns Mean |
|----------------------|------------------|------------|------------|--|--|
| Total | 1,654,805 (100%) | 302 (100%) | 227 (100%) | | |
| Associated diagnoses | | | | | |
| Failure to thrive | 118,724 (7%) | 76 (25%) | 82 (36%) | <.001 | <.001 |
| Feeding problem | 91,061 (6%) | 72 (24%) | 73 (32%) | <.001 | <.001 |

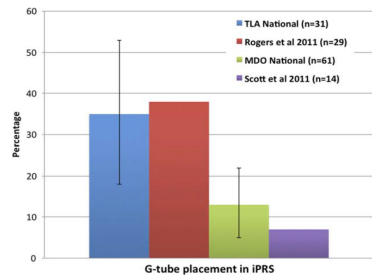
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Oral Feeding

- 25-30% of infants with PRS have feeding difficulty
- 35-38% who undergo TLA require GT
- 7-12% who undergo MDO require GT



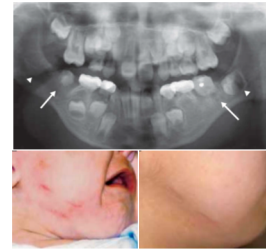
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Complications in MDO

- Local infection (6%)
- Injury to tooth buds (21-29%)
- Hypertrophic scar (2-21%)
- Device failure (2-7%)
- Repeat distraction (5-7%)
- Open bite deformity (2-5%)
- Permanent facial nerve injury (1-1.2%)
- TMJ ankylosis (0.5%)



Sidman, 2011

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Summary

- Nonsurgical airway management in PRS is effective as much as 90% of the time
- Mandibular distraction osteogenesis (MDO) is an effective means of improving the caliber of the oropharyngeal airway
- MDO is associated with a lower likelihood of requiring NG or GT feeds when compared to TLA or NP stents



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References

- Parhizkar N, Saltzman B, Grote K, et al. Nasopharyngeal airway for management of airway obstruction in infants with micrognathia. *Cleft Palate Craniofac J*. 2011 Jul;48(4):478-82.
- Manouk JS, van Lieshout MJ, Malthijssen IM, et al. Non-surgical and surgical interventions for airway obstruction in children with Robin Sequence. *J Craniomaxillofac Surg*. 2016 Dec;44(12):1871-1879.
- Ren XC, Gao ZW, Li YF, et al. The effects of clinical factors on airway outcomes of mandibular distraction osteogenesis in children with Pierre Robin sequence. *Int J Oral Maxillofac Surg*. 2017 Jul;46(7):805-810.
- Breik P, Tivey D, Umapathysivam K, et al. Mandibular distraction osteogenesis for the management of upper airway obstruction in children with micrognathia: a systematic review. *Int J Oral Maxillofac Surg*. 2016 Jun;45(6):769-82.
- Qiu AT, Cheung LK. Meta-Analysis of Mandibular Distraction Osteogenesis: Clinical Applications and Functional Outcomes. *Plast Reconstr Surg*. 2008 Mar;121(3):54e-69e.
- Tsui WK1, Yang Y, Cheung LK, et al. Distraction osteogenesis as a treatment of obstructive sleep apnea syndrome: A systematic review. *Medicine (Baltimore)*. 2016 Sep;95(36):e4674.
- Evans KN, Sie KC, Hopper RA, et al. Robin sequence: from diagnosis to development of an effective management plan. *Pediatrics*. 2011 May;127(5):936-48.

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