NOTE:
- Different Institutions and countries have varying methods of making or providing a nasopharyngeal airway. Literature has been included in this evidence table if the artificial airway maintains the basic principle of bypassing the level of airway obstruction at the base of the tongue, but ending before the epiglottis.
- Conditions in infants causing Upper airway obstruction (UAO) at the level of the base of the tongue are relatively rare, and therefore large systematic reviews and randomised controlled trials are not available for this group.

<table>
<thead>
<tr>
<th>Reference (include title, author, journal title, year of publication, volume and issue, pages)</th>
<th>Evidence level (I-IV)</th>
<th>Summary of recommendation from this reference</th>
</tr>
</thead>
</table>
• International consensus was achieved regarding the three distinguishing features (micrognathia, glossoptosis, and upper airway obstruction)
• Cleft palate is not considered a prerequisite for the diagnosis
• Upper airway management plays a central role in treatment
• Non-surgical interventions include positional change, management of the nasopharyngeal airway |
<table>
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<th>Page</th>
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| VI   | - Nasopharyngeal airways are used in urgent situations to alleviate airway obstruction  
- NPA patency is critically dependent on both the internal diameter of the airway and the position of the distal tip  
- Ideal position of NPA distal tip should protrude beyond pharyngeal end of the soft palate, but not pass the epiglottis  
- Vital that an NPA is sized correctly  
- If too short it will fail to separate the soft palate and pharynx, and if too long it can pass into the larynx and aggravate cough and gag reflex  
- Ideal position is 10 mm above epiglottis  
- Allows for movement with flexion and extension of the head without aggravating gag  
- Awareness of respiratory obstruction even with an airway should always be maintained |
| VII  | - Robin sequence is a congenital condition characterised by micrognathia, glossoptosis, and upper airway obstruction  
- Glossoptosis is defined as displacement of the tongue base into the oropharynx and hypopharynx  
- Degree of glossoptosis and resultant upper airway obstruction can vary  
- In severe cases, there will be inspiratory stridor with increased respiratory effort and possible apnoea and cyanotic episodes  
- Mild glossoptosis may permit the child to maintain an adequate airway when awake, but have heightened risk for sleep disorder breathing  
- Glossoptosis is the primary cause of respiratory problems in Robin sequence  
- Delay in diagnosis may be due to intermittent nature of upper airway obstruction  
- Clinical signs include stridor, laboured breathing, diaphoresis, apnoea, restlessness. Can be assessed when asleep, awake and feeding  
- Jaw thrust manoeuvre can pull tongue forward and help illustrate tongue based obstruction  
- Chronic airway obstruction may be indicated by poor growth due to increased energy expenditure caused by greater work of breathing and feeding challenges |


| --- | --- |

- Management of airway obstruction in infants with PRS are controversial and varied
- Some management providers exhaust nonsurgical management strategies whereas others choose an early operative approach
- For initial treatment, prone and lateral positioning as accepted and can resolve obstruction in 40–70% of cases
- Other non-operative treatment options include oxygen supplementation, NPA, CPAP, and intubation
- CPAP has been shown to relieve moderate to severe airway obstruction in patients with PRS
- Surgical options include tongue lip adhesion, mandibular distraction osteogenesis, and tracheostomy
- Mandibular distraction has replaced tracheostomy as the first line operation
- Tracheostomy now reserved for patients with multilevel obstruction and comorbidities

- Mandibular distraction osteogenesis has shown to be successful in treating upper airway obstruction in patients with micrognathia
- Retro positioned hypoplastic mandible pushes the tongue base into the oropharynx causing upper airway obstruction
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<tr>
<td><strong>IV</strong></td>
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<tr>
<td>• The goal of initial treatment is to minimise any airway obstruction to prevent hypoxia and promote normal neurological development</td>
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<td>• A current treatment strategy is nasopharyngeal intubation to help the tongue move forward, freeing the airway and allowing the child to breathe through the nasopharyngeal tube</td>
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<tr>
<td>• Respiratory obstruction is classified as: Type one: the tongue is retro positioned and touches the posterior pharynx wall. Type 2: the tongue presses the palate against the pharynx wall type 3: there is a medial contraction of the pharynx and the pharynx is the cause of obstruction, while the tongue does not touch the pharynx wall; and type 4: the contraction of the pharynx is sphincteric.</td>
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<tr>
<td>• Type one is the most frequent</td>
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<td>• types 3 and 4 are not considered RS but Robin complex (as the tongue is not the cause of respiratory obstruction)</td>
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<td>• Since the obstruction occurs at the base of the tongue, the treatments aim to move the tongue base forward, away from the airway</td>
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<tr>
<td>• 107 infants in a 7 year period with Robin Sequence, severe respiratory symptoms and type 1 or 2 airway obstruction as classified with Sher, were treated with a nasopharyngeal airway (NPA).</td>
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<td>• Management with a NPA continues in hospital and then at home until a NPA was no longer required. Mean time with a NPA in situ was 57.4 +/- 37.6 days. There were no nasal injuries and no untoward incidences at home. NPA was safely managed at home by trained parents.</td>
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<tr>
<td>• In this group studied, NPA improved respiratory difficulties and improved feeding difficulties with 85% of patients fed orally.</td>
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<td>• There was low morbidity and zero mortality in this group in the first year of life</td>
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<td>• Previous studies have reported that use of NPA to relieve airway obstruction as successful</td>
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<td>• Reports suggest that natural growth may lead to resolution of airway obstruction and therefore no need for surgical intervention</td>
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<tr>
<td>• The main issues in patients with PRS are feeding difficulties and airway obstruction</td>
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<tr>
<td>• Because NPI improves airway obstruction, the infants ability to orally feed may also be improved</td>
</tr>
<tr>
<td>Reference</td>
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</tbody>
</table>
| Oh, M., Park, Y., Jung,Y., Choi, C., Kim,B., & Kwon, J.  **Three cases of Pierre robin sequence with upper airway obstruction relieved by nasopharyngeal airway insertion.** Neonatal Medicine 2019 26(3), 179-183 | IV | - There has been no consensus for the treatment of upper airway obstruction in infants with PRS however studies recommended attempting non-surgical interventions first  
- Nasopharyngeal insertion is highly effective in infants with PRS  
- This cases study follows 3 infants diagnosed with PRS. All 3 had nasopharyngeal tubes inserted and airway obstruction was relieved  
- Several studies have reported the effectiveness and safety of NPA insertion  
- NPA insertion can be considered as a primary treatment option for infants with PRS |
| Government of Western Australia North Metropolitan Health Service. Clinical Practice Guideline. **Pierre Robin Sequence (PRS) and Nasopharyngeal airway insertion** | VII | - Medical staff to insert initial NPT  
- Nursing staff deemed competent can insert subsequent  
- Alternate nares when changing NPT  
- Pre-cut ETT must be kept at bedside in case of accidental removal  
- Indications for use: Obstructive episodes, respiratory distress, episodes of desaturation, sleep studying indicative of obstruction, poor feeding/weight gain |
| Skirko, J., Pollard, S., Slager S., Hung, M, & Weir, C. **Family Experience With Pierre Robin Sequence: A Qualitative Study.** American Cleft Palate Craniofacial association. 2020;57(6):736-745. | VI | - Upper airway collapse causes significant breathing and feeding difficulties  
- Some infants can be managed with prone positioning, however others require a nasopharyngeal airway  
- Surgery may be required to stabilise the airway |

**Key abbreviations:**
- Gastro oesophageal Reflux (GOR)
- Mandibular Distraction Osteogenesis (MDO) – *also known as Jaw distraction*
- Nasopharyngeal Tube (NPT)
- Pierre Robin Sequence / Syndrome (PRS)
- Upper Airway Obstruction (UAO)
- Upper Respiratory Tract Infection (URTI)
- Feed facilitating techniques (FFT)
A guide to Levels of Evidence

Clinical Guidelines and parent information (Kids Health Info)
Royal Children's Hospital

The Hierarchy of Evidence

The Hierarchy of evidence is based on the National Health and Medical Research Council (2000) and Oxford Centre for Evidence-based Medicine Levels of Evidence (May 2001)

I  Evidence obtained from a systematic review of all relevant randomised control trials.
II  Evidence obtained from at least one properly designed randomised control trial.
III-1 Evidence obtained from well-designed pseudo-randomised controlled trials (alternative allocation or some other method).
III-2 Evidence obtained from comparative studies (including systematic reviews of such studies) with concurrent controls and allocation not randomised, cohort studies, case control studies, or interrupted time series with a control group.
III-3 Evidence obtained from comparative studies with historical control, two or more single–arm studies, or interrupted time series without a parallel control group.
IV  Evidence obtained from case-series, either post-test or pre-test and post test.
V  Expert opinion without critical appraisal, or based on physiology, bench research, or historically based clinical principles.

Kids' health info and clinical guidelines are based on reviews of the best available evidence. Level 1 evidence represents the gold standard for intervention studies; however it is not available for all areas of practice and for some factsheets it may be appropriate to utilise results from studies with lower levels of evidence. Many factsheets may also be informed by experts in the field, locally (RCH) and internationally – journal articles, expert opinion etc. This NHMRC hierarchy can be used to grade evidence. Please record details on the evidence table and return to Kids Health Info with your final draft. If you are basing parent info on a guideline, the Evidence Table will be the same.