Management of Upper Airway Obstruction in Pierre Robin Sequence

South Wales Cleft Team
Pierre Robin Sequence

- Triad of cleft palate, micrognathia and airway obstruction was described by St Hilaire in 1822, by Fairbain in 1846, and by Shukowsky in 1911

- 1920s Pierre Robin described glossoptosis and its relationship with micrognathia and airway complications

- Now PRS is usually defined as a variable combination of
  - Micrognathia
  - (Cleft palate)
  - Glossoptosis
  - & resultant airway obstruction

- The literature describes wide variability in management of babies with PRS
- Care must be customized for each patient, as PRS is quite heterogenous:
  - some have isolated PRS
  - some have associated anomalies
  - and some are syndromic
Degrees of Micrognathia in PRS

The skeletal, soft tissue, and airway relationships in PRS: Mandibular hypoplasia, glossoptosis and a narrowed airway
Cleft, glossoptosis & UAO in PRS

Mechanisms of UAO in PRS

- Displacement of the tongue into the hypopharynx → so occluding the airway at the level of the epiglottis
- Disproportionate tongue growth
- Tongue prolapse into the cleft palate
- Lack of voluntary control of tongue musculature & abnormal maxillary morphology causing midface hypoplasia, especially in syndromic PRS

- But there is agreement that **UAO at the tongue base caused by glossoptosis is a defining feature of the PRS phenotype**
  - There are 4 endoscopically classified types of airway obstruction in children with craniofacial anomalies
  - Type 1 obstruction is posterior movement of the dorsal tongue against the pharyngeal wall, i.e. glossoptosis
  - In 62 children with probable isolated PRS evaluated by nasopharyngoscopy, 90.9% were classified as having type 1 obstruction, 75% of whom responded to nonsurgical management. (Marques et al. 2001)
Current Practice in South Wales

• Any baby with cleft palate detected at birth is assessed for respiratory distress and ability to feed, prior to discharge from the post natal ward (PNW)

• This assessment is undertaken by
  – the local Paediatric team
  – & the Clinical Nurse Specialists (CNS) from the Cleft Team who visit baby & mum on PNW
Management thereafter is variable

• Management of airway and feeding is very variable and may result in
  – unnecessarily prolonged hospital stays
  – or early discharge with concerns that respiratory distress, obstructive apnoea or poor feeding may be inadequately diagnosed & addressed
So how can this be improved?

• Internationally there is paucity of good evidence

• But we have been advocating early use of NPA

• Also this is now becoming accepted as UK wide practice and is believed to be safer

• And a recent review from GOS supports our view  (Abel, F. et al. Arch Dis Child 2012;97:331–334)

• So we propose the following step-by-step guidance
@ initial assessment

- Meet with family
- Confirm diagnosis – i.e. fits criteria for PRS
- Counsel the family that breathing +/- feeding difficulties will worsen over the first 4 – 6 weeks of life
- Reassure them that by 2 – 3 months improvements start to appear
- Talk through the proposed management plan & discharge pathway
NB – This guidance is NOT intended for feeding advice

• This guidance is for care of upper airway obstruction (UAO), so does not specifically address feeding

• Feeding issues should be con-currently addressed by the local paediatric team & Cleft team CNS on a case-by-case basis, re timing / necessity of NGT
Clinical assessment

• Baby is observed in the lying supine position and signs of respiratory distress are noted:
  any of these: raised respiratory rate, recessions, tracheal tug, nasal flaring, grunting, cyanosis

• Baby is observed taking a feed in the position that parents would normally feed baby

• This guidance is for care of upper airway obstruction (UAO), not intended as guidance for feeding management – which should of course be addressed on a case-by-case basis
@ initial assessment

- Baby may have no respiratory distress at rest or on feeding

- Baby may have no distress while upright and feeding – but distress while lying supine

- Baby may have distress at rest and on feeding
Summarised Care Pathway for Management of UAO in PRS

No distress at rest or on feeding

Distress only when lying supine

Distress at rest and feeding

Saturation monitoring

Position side-to-side

NPA

Saturation monitoring

Home – on Discharge Pathway

Abnormal

Normal

Abnormal

Normal

Normal
In babies with no respiratory distress

- In absence of clinical signs of respiratory distress – baby should have continuous saturation monitoring to cover an overnight sleep and a daytime period of normal activity, handling and feeding
- If normal – baby is discharged and follows the Discharge Pathway
- If this shows significant desaturations – baby should be nursed side to side and repeat clinical assessment & saturations study to confirm normalises
- Then may discharge and follow the Discharge Pathway
- If not – insert NPA & repeat saturations study to confirm normalises
- Then may discharge and follow the Discharge Pathway
In babies who feed well and only have distress in supine positioning

- Nurse baby side to side
- Check continuous saturation monitoring to cover an overnight sleep and a daytime period of normal activity, handling and feeding
- If normal – baby is discharged, nursed side to side, and follows the Discharge Pathway
- If this shows significant desaturations – baby should have NPA and repeat saturations study to confirm normalises with NPA in situ – then can be discharged and follows the Discharge Pathway
In babies with respiratory distress in supine positioning & while feeding

- Place an NPA
- Check continuous saturation monitoring to cover an overnight sleep and a daytime period of normal activity, handling and feeding
- If normal – baby is prepared for discharge and follows the Discharge Pathway
- If this shows significant desaturations –
  - ensure NPA correctly positioned
  - exclude lower respiratory cause for distress
- If no improvement – refer to cleft team at Moriston for further assessment of UAO
Discharge Pathway

• Open access to local paediatric assessment unit
• Weekly home visit by Clinical Nurse Specialist
• Regular local paediatric reviews
• Review for need of input re feeding – as necessary

• If discharged home with NPA:
  – Liaison with local community paediatric nursing / health visiting team
  – Home suction equipment if available – but should not delay discharge
  – Does NOT require home saturation monitor / apnoea monitor

• If any deterioration: PTO ➔
If baby’s respiratory distress worsens

- Admit to local paediatric ward to assess clinically
- Treat any con-current illness as necessary
- If not already: reassess need for side to side positioning or NPA placement
- Following any intervention for UAO – baby should have continuous saturation monitoring to cover an overnight sleep and a daytime period of normal activity, handling and feeding
- If clinically improves & sats normalise – baby is discharged and follows the Discharge Pathway
- If not – refer to Cleft Team
When to wean off the NPA

• Natural history of the upper airway obstruction in PRS:
  – Least at birth & early neonatal period
  – Progressively worsens over first 4 – 6 weeks as tongue grows
  – Gradually improves over next 2 months as mandible grows

• So @ 10 – 12 weeks of age: admit for a trial off NPA – with clinical assessment & saturation monitoring again to cover an overnight sleep and a daytime period of normal activity, handling and feeding

• If unsuccessful – replace and repeat trial in a month
Significant desaturations

• Either go with existing local expertise

• Or guidance from literature *:
  

  – where moderate & severe = NPA was advocated in the GOS cohort (Abel, F et al. Arch Dis Child 2012;97:331–334)
  
  – **Mild** = at least 3 clusters of desats with at least 3 dips to 85-90%

  – Moderate = at least three clusters of desaturations with at least 3 dips below 85% but above 80%

Word of caution: oximetry is **not** the gold standard
Summarised Care Pathway for Management of UAO in PRS

No distress at rest or on feeding

Saturation monitoring

Abnormal

Distress only when lying supine

Position side-to-side

Saturation monitoring

Abnormal

Distress at rest and feeding

NPA

Saturation monitoring

Abnormal

Saturation monitoring

Normal

Normal

Normal

Home – on Discharge Pathway
# Measurement of NPA

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<th>WEIGHT OF BABY (KG)</th>
<th>LENGTH OF ETT (CM)</th>
<th>DIAMETER OF ETT (MM)</th>
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<td>&gt;3.5</td>
<td>8.0</td>
<td>4.0</td>
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NPA preparation
NPA insertion
References


• Evans, KN et al. Robin Sequence: From Diagnosis to Development of an Effective Management Plan. Pediatrics 2011;127;936-948.


• Clinical guidelines from the Royal Children’s Hospital Melbourne.