CLINICAL GUIDELINE	
Pierre Robin Sequence (PRS) and Nasopharyngeal Airway Insertion	
Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

This document should be read in conjunction with this DISCLAIMER

Contents

First described by a French physician, PRS is characterised by a small jaw (*micrognathia*) which displaces the tongue into the back of the throat (*glossoptosis*) causing obstructed breathing. In addition, the infant usually has a central cleft palate. PRS can occur as an isolated anomaly (35%) or associated with other anomalies/syndromes (65%). Stickler syndrome is the most common associated condition occurring in 15-20% of cases of PRS: this is an autosomal dominant condition and usually asymptomatic in the newborn and often undiagnosed an adult. Other much less common conditions are: hemifacial macrosomia, velo-cardio-facial syndrome and Treacher-Collins syndrome.

Upper Airway Obstruction

The main problem with PRS is upper airway obstruction; this can be mild to severe and can vary from moment to moment (symptoms include stridor, intercostal retraction and oxygen desaturation). Infants with PRS should be nursed in the **prone position,** which encourages the tongue to move anteriorly out of the pharynx, thereby relieving the upper airway obstruction. Over time a baby may spend more time in the supine position e.g. with nappy changes but always with full cardio-respiratory monitoring and direct observation. Infants with severe PRS may require the insertion of a **nasopharyngeal tube** placed just above the larynx to relieve respiratory obstruction. Refer to Nasopharyngeal Tube Insertion.

Infants with PRS and a cleft may require a 'feeding plate' (orthodontic appliance) although this is becoming much less frequent. The feeding plate is fitted by an orthodontist **but should never be fitted in the 1st week or 2 of life when there can be significant respiratory obstruction.**

Cleft Palate Coordinator

If the infant has a cleft palate, please inform the Cleft Clinic Coordinator during office hours who will then contact the plastic surgeons and orthodontists.

Feeding and Infant with PRS

Feeding and weight gain in infants with PRS can be very challenging because of their difficulty to coordinate suck, swallow and breathing. The initial oral feeds should be by a nurse experienced in feeding infants with PRS. Frequently, 24 calorie per ounce feeds are used to supplement nutritional intake. Infants with mild PRS who have few obstructive episodes may be nursed in a side lying position, following discussion with the medical staff. Please view the video entitled **"Feeding an Infant with the Pierre Robin Sequence" 2001.** They may also have poor weight gain and gastroesophageal reflux.

Feeding Equipment

- Haberman Feeder or for borderline PRS pigeon bottles/squeezy bottle may be used.
- Needs cardio respiratory and pulse oximetry monitoring.
- Laryngoscope and resuscitation equipment at the bedside.

Procedure

1. Nurse the infant on your lap in an extreme upright position. Place your hand between the shoulders and the neck to support the baby during feeding. Place your feet on a foot stool. Your body should be at a 90 degree angle.
2. Use the Haberman Teat or Squeezy Bottle, place the nipple in the centre of the infants mouth and as the infant sucks gently squeeze the bottle or Haberman teat (with your thumb and fore finger) to allow sufficient amount of milk to flow in the infant's mouth for them to swallow without choking. Co-ordinate squeeze, sucking and swallowing. Allow short breathing spells without removing teat from the mouth.
3. Pull the jaw forward with your middle and other fingers (May enable the airway to remain more open during feeding).
4. Ensure constant monitoring (cardiac, SaO_2 and direct vision) throughout the feed.
5. Excessive drooling/ frothing of milk are usually an indication of the teat sitting under the tongue. Remove the teat and place on top of the tongue.
6. Frequently burp the infant as they tend to swallow excessive amounts of air (Keep the infant in an upright forward leaning position to allow the infant's jaw to fall forward, thus preventing airway obstruction during the burping process).

7. If the feed is prolonged (30 minutes or more) consider finishing the feed via NGT. (Thus preventing use of excessive amounts of energy to feed).

Sleep Studies

Sleep studies are a relatively new tool for measuring the degree of upper airway obstruction. **A sleep study is not used as the main determinant of discharge.** A PRS infant without cot side oxygen desaturations may still have significant obstruction and require a nasopharyngeal tube. On average the 1st sleep study is performed in the 3rd week of life. Sleep studies in PRS are rarely normal over the first year of life.

Discharge Preparation and Information

Agreeing a safe discharge time is a very challenging task. Many factors need to be taken into consideration and these factors will vary from baby to baby and family to family. The average length of stay for babies with PRS is 1-2 months. Baby factors include:

- Minimal/no cot side oxygen desaturations/bradycardias.
- At least 50% suck feeds.
- Adequate weight gain; around 150 grams/week.
- Infant able to cope with short period in supine position.*
- Parents are confident feeding their infant.
- Parents are confident with basic care/positioning of their infant.*
- Sleep study reassuring.
- Parents completed ‘Tube feeding package” if required.
- Parent/s complete ‘Infant Resuscitation’ training.
- Home monitoring supplied and parent/s competent with monitor.**
- There is adequate home support in place.

*****Prior to discharge, the infant will be placed supine to assess their level of obstruction. This will be done by a neonatal nurse or medical staff experienced in caring for infants with PRS.

******Approximately 1 week prior to discharge the infant must be referred to the Monitoring Clinic to make provisions for home monitoring (Corometrics).

Home Feeding Equipment and Supplies

Parents/carers must be competent in using any monitoring and suctioning equipment that may be required to care for their infant at home.

The Cleft Palate coordinator/nurse will refer the parents/carers to the Cleft Pals association of Western Australia. Cleft Pals provide Haberman Feeders to purchase and also offer support groups for infants with PRS.

Parents should test their infant in their car seat on the ward with an oxygen saturation monitor fitted over approximately 30-60 minutes to ensure their infant's safety in the car seat prior to discharge.

Nasopharyngeal Tube Insertion (NPT) for Pierre Robin Sequence

Parent Education Record - Nasopharyngeal Tube

Key Points

- Medical staff to insert initial NPT. Nursing staff deemed competent in the procedure can insert subsequent NPT.
- Use table above as a guide for tube size selection.
- X-ray to be performed after initial insertion for confirmation of tube position.
- Alternate nares when changing NPT.
- NPT to be changed every 48 hours for the first 10 days and then weekly.
- A pre-cut ETT is to be kept at the bedside in case of accidental / unplanned removal of tube.

Indications for Use

- Obstructive episodes.
- Respiratory distress.
- Episodes of desaturation.
- Sleep study indicating obstructive episodes.
- Poor feeding and weight gain.

Equipment

- Ivory endotracheal tube - see table below.
- Saline.
- Fixomull tape.
- Skin preparation wipe.
- Sterile scissors.
- Sucrose

Guide for Nasal Tracheal Tube Size

Weight	Tube Size
< 1500 g	2.5 mm
1500-2500 g	3.0 mm
2500-3500 g	3.5 mm
> 3500 g	4.0 mm

Measurement

Measure the length of the tube by measuring from the tip of the nose to the tragus of the ear and add 0.5cm. Cut the tube at the measured distance.

Procedure

1. Select appropriate size tube and measure as above. Review tube size and measurement, with every tube change.
2. Cut ETT as per Fig. 1 and 2 below.
3. Place infant in supine position.
4. Consider use of oral sucrose.
5. Moisten tip of tube with saline if required.
6. Insert tube into nare and gently advance to the measured length. The tip of the tube should end 1cm from the epiglottis.
7. Secure cut lengths of the tube to either side of the nose along cheeks with fixomull as per Fig. 3 below.
8. Document the following
    - ETT size and length at nare.
    - Date for next tube change.
    - Condition of infant during procedure.

Nursing Care of NPT

- Suction tube 2-3 hourly for the first 48-96 hours, then PRN.
- Observe and document at least each shift, condition of skin around nares and under taping.
- Change tape if soiled with milk or secretions.
- Remove tape with adhesive remover.

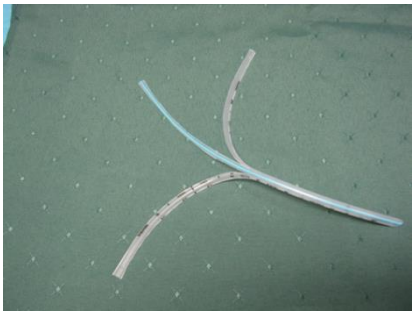

Fig. 1

Cut ETT into 3 strands

Cut a 5cm length along either side of blue line, then down the middle of remainder of tube.

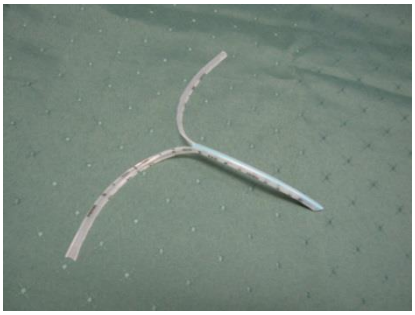

Fig. 2

Trim off strand with blue line, leaving 2 strands.

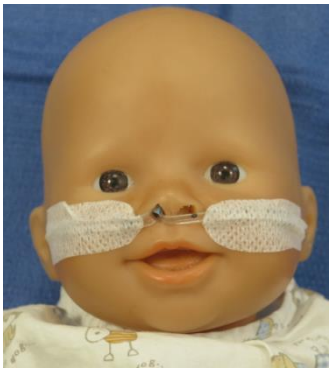

Fig. 3

Tape the NPT strands either side of the nose along the cheeks.

Useful resources (including related forms)
Pierre Robin Sequence (PRS) – Quick Reference Guide
Nasopharyngeal Tube – Parent Education Record

References

1. Annie Cole, Patricia Lynch, Rona Slator (2008) A New Grading of Pierre Robin Sequence. The Cleft Palate Craniofacial Journal, vol 45, no 6, pp. 603-606.
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Quick Reference Guide for the Management of the newborn with Pierre Robin Sequence