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, SaO₂ 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.
References


Related WNHS policies, procedures and guidelines

Neonatology Clinical Guidelines – Nasopharyngeal Tube Insertion

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