PIERRE ROBIN SEQUENCE (PRS)

First described by a French physician, is characterised by a small jaw known as micrognathia and glossoptosis, where the tongue base is displaced in the oropharynx. In addition, the infant usually has a cleft palate. PRS can occur as an isolated finding, or it can be a combination with a genetic syndrome. Examples include Stickler syndrome, hemifacial macrosomia, velocardiofacial syndrome and Treacher Collins syndrome. They may also have poor weight gain and gastroesophageal reflux.

If the infant is suspected of having, or is diagnosed with Pierre Robin Sequence, position the infant prone immediately. Prone positioning encourages the tongue to move anteriorly out of the pharynx, thereby relieving the anatomical obstruction. Full observation of the infant during all types of activity is needed to assess (feeding, sleeping, crying and awake periods).

Perform a thorough physical assessment on the infant. If the infant is diagnosed with a cleft palate in addition to PRS on admission, please inform the Cleft Clinic Coordinator. Infants with a cleft are also reviewed by a dental surgeon as they may require an orthodontic appliance (OA).

Infants with PRS are often unable to coordinate suck, swallow and breathing because of the anatomical displacement of the tongue and may exhibit airway obstruction episodes which includes symptoms of stridor, intercostal retraction, and oxygen desaturation OR infants may be pink and still have marked respiratory compromise. Listen at the nares with a stethoscope for air movement, it is an effective way to discover an airway obstruction during respiration.

Some Infants with PRS may require the insertion of a nasopharyngeal tube to assist with the management of obstructive episodes. Attend all nursing cares in the prone position (nappy changing, bathing and feeding).

The initial oral feeds should be by a nurse experienced in feeding infants with difficulties in coordination. Infants with mild PRS who have few obstructive episodes may be nursed in a side lying position, following discussion with the medical staff. Firstly, view the video entitled; “Feeding an Infant with the Pierre Robin Sequence” 2001.

FEEDING AN INFANT WITH PRS

- 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
Nurse the infant on your lap / thigh 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.

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 infants mouth for them to swallow without choking. Coordinate squeeze, sucking and swallowing. Allow short breathing spells without removing teat from the mouth.

Pull the jaw with your middle and other fingers (May enable the airway to remain more open during feeding).

Ensure constant monitoring (cardiac, oxygen saturation and visualisation of infant) occurs throughout the feed.

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.

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 infants jaw to fall forward, thus preventing airway obstruction during the burping process).

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

Fortified feeds should be considered, as the infants often have insufficient weight gain due to feeding difficulties.
DISCHARGE PREPARATION AND INFORMATION

Ensure that the parents/carers of the infant have watched the video entitled; “Feeding an Infant with the Pierre Robin Sequence” 2001; and are competent with feeding their infant using either the Haberman Teat or the Squeezy Bottle.

All parents/carers require education on CPR for infants with PRS and education on appropriate positioning for their infant to minimise airway obstruction (sideling and prone positions).

Prior to discharge, the infant will be place supine to assess their level of obstruction. This will be done by an experienced Nurse or Medical staff member with caring for infants with PRS.

All infants with PRS will be referred for a “sleep study” to assess the occurrence of obstructive episodes during sleep, prior to discharge and will require constant home monitoring if they are at risk of obstructive airway episodes. Approximately 1 week prior to discharge the infant must be referred to the Monitoring Clinic Consultant to make provisions for monitoring (Corometrics) within the home.

HOME FEEDING EQUIPMENT AND SUPPLIES

Parents/carers must be competent in using any monitoring and suctioning equipment they will be using 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 will also need to consider the type of car seat they will purchase. Parents are able to bring their car seat up to the ward to trial the infant’s condition when placed into the car seat, prior to discharge.

REFERENCES