GUIDELINE

Cleft Lip and Palate

Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this disclaimer

Contents

Aim

To provide standardised management and follow up to infants and families of infants with cleft lip and/or palate (CL/P).

Risk

Infants with cleft lip and/or palate may not receive appropriate inpatient management and follow-up in the absence of standardised care.

Background

Cleft lip and/or palate are openings or splits in the upper lip, the roof of the mouth (palate) or both.

- **Cleft lip**: The failure of fusion of the frontonasal and maxillary processes, resulting in a cleft of varying extent through the lip, alveolus, and nasal floor (an incomplete cleft does not extend through the nasal floor, while a complete cleft implies lack of connection between the alar base and the medial labial element).
- **Cleft palate**: The failure of fusion of the palatal shelves of the maxillary processes, resulting in a cleft of the hard and/or soft palates. Clefts arise late in the first trimester. Exactly where they appear is determined by locations at which fusion of various facial processes failed to occur, this in turn is influenced by the time in embryologic life when some interference with development occurred
- There are a range of subclinical features: craniofacial measures, dental anomalies (tooth agenesis, microdontia, and supernumerary teeth), brain structural differences, and dermatoglyphic lip print whorls. Subclinical phenotypes of the lip and palate include microform clefts (also known as congenital healed cleft lip or forme fruste cleft lip), defects of the *orbicularis oris* muscle, bifid uvula, submucous cleft palate. VPI can be a result of a cleft but is not a cleft per se.

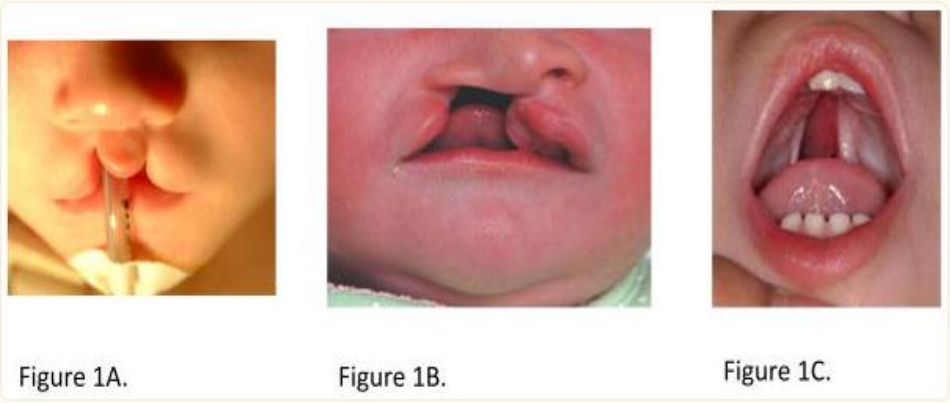


Figure 1A. Figure 1B. Figure 1C.

Incidence and prevalence

- Overall incidence of orofacial cleft is around 1.5 per 1000 live birth (about 220,000 new cases per year). WA birth incidence is approximately 2 to 2.2 per 1000.
- Wide variation across geographic areas, ethnic group (higher in Caucasians and Asians) and nature of cleft itself. Higher incidence is also seen in Aboriginal West Australians.
- CL/P occurs more frequent and more severe in boys than in girls.
- The typical distribution of cleft types is: Cleft lip alone (15%), Cleft lip and palate (45%), Isolated cleft palate (40%).

Aetiology

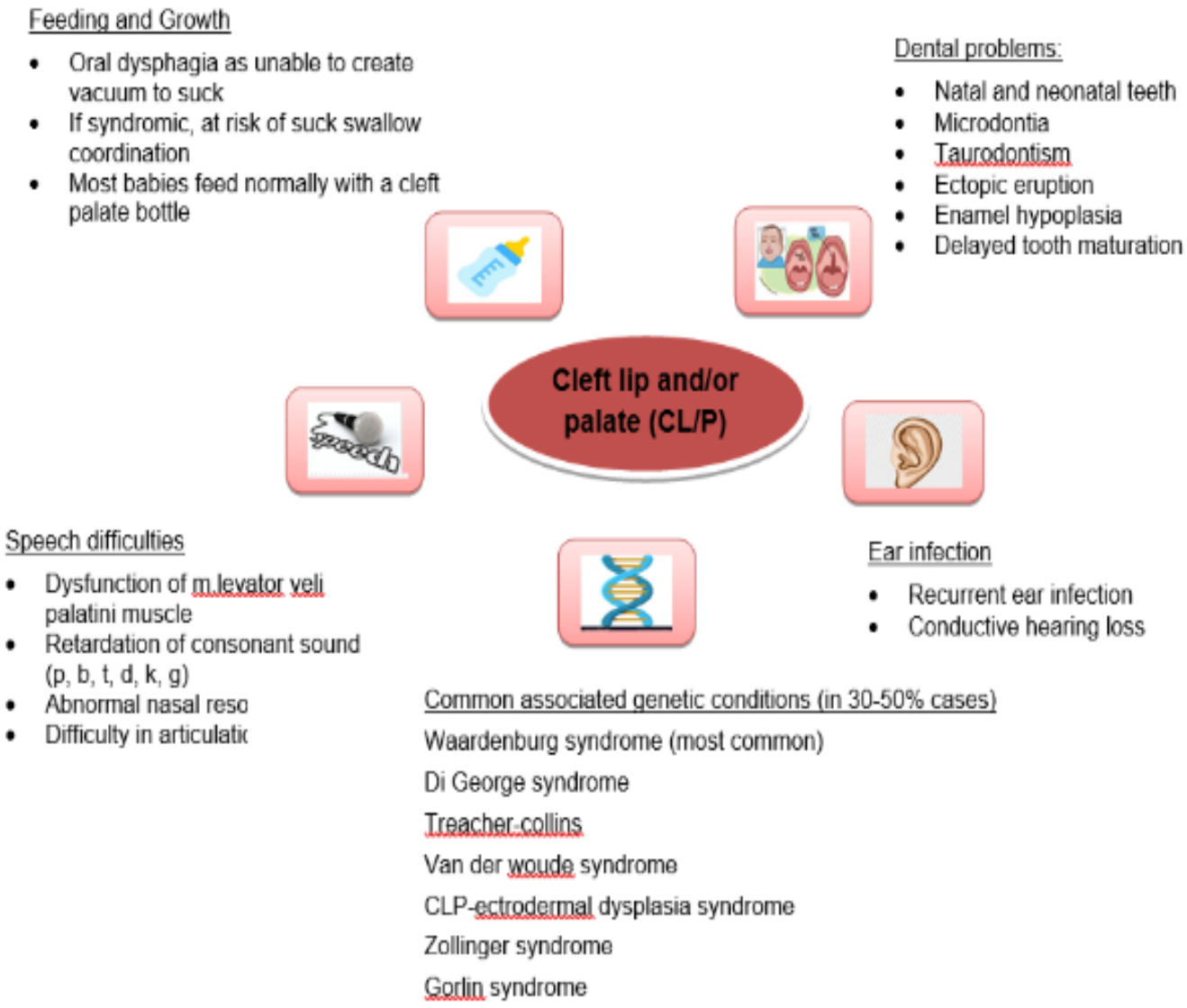
The aetiology of cleft lip and palate is complex and thought to involve genetic influences with variable interactions from environmental factors.

- 30-50% of Cleft lip and or palate (CL/P) are associated with syndromes.
- Non syndromic clefts are found to have some recognisable genetic mutations such as TGFA, TGF 133, MTHF3, RARA, ET1 etc.
- Family history of cleft lip and/or palate:
  - One parent affected: 3.2% chance of child having cleft lip and palate
  - One parent and one sibling affected: 15.8% chance of second child with cleft lip and palate
  - One child affected: 4.4% chance of next child to have cleft lip and palate

Clinical Description

Cleft of lip and palate are most common serial congenital anomalies to affect the orofacial region. The potential problems of the condition include social handicaps such as impaired suckling and resultant failure to thrive, speech impediment, deafness, malocclusion, gross facial deformity and severe psychological problems

Note: Look carefully for associated micrognathia and glossoptosis in cases of isolated soft cleft palate for Pierre Robin sequence (refer to CAHS PRS guideline). Robin sequence is more often associated with a wide U-shaped cleft of soft and hard palate "Veau II"



Initial Neonatal Management

- Clinical examination of the baby by the Senior Registrar or Neonatal Consultant to evaluate and consider chromosomic association.
- Cardiorespiratory monitoring and positioned in the supine position.

Transfer/Admission to PCH NICU if:

1. **Cleft lip and palate:** All patients with complete clefts of the lip and palate require transfer and admission to PCH NICU to commence the presurgical moulding in first week of life which is associated with improved outcomes. KEMH NICU to liaise with ward 3B for maternal bed availability when the mother can be transferred with her baby.

2. **Cleft palate only:** All patients with isolated cleft palate require an initial assessment of the upper airway for any obstruction or associated retrognathia. Transfer to PCH NICU for assessment and management by Plastics/Dental/Speech Pathology.
3. **Cleft lip only:** Not routinely required to be transferred to PCH NICU. If feeding well and gaining weight, KEMH NICU to discuss with plastic surgeons on call and follow up with the Plastics as an outpatient.

Admissions to PCH NICU

- O/A - Inpatient referral to Plastic surgery/Cleft team with a brief medical history and phone call to Plastics on call registrar and on call dentist. Plastic surgery team will review as an in-patient and discuss the timing of definitive surgery for the lip and/or palate. If there are clinical concerns, refer to genetics for an inpatient review.
- Inpatient referral to Dental team/orthodontists. Dental will assess the need for an orthodontic plate, bonnet and upper lip strapping or any other immediate procedures. See Appendix 2
- **Feeding assessment is the most important of care of infants** by an experienced bedside nurse (Appendix 1)
- Consider microarray (parental consent required) and/or Head ultrasound in bilateral cleft lip and palate or if clinical concerns of dysmorphism.
- Contact Clinical Photography (extension 60357) for 2D/3D facial photos. Parental consent needed.

Definitive management of cleft lip and palate

- Balance between the maxillary growth and speech development is taken into consideration to achieve functional and aesthetic well-being.
- Dental plate is considered in complete cleft of lip and palate (u/l or b/l). Aim is to fit the plate by 3-7 days of age
- The timing of cleft repair is usually at 3 months for cleft lip repair and 9 months for cleft palate repair.

Goals for discharge

- No clinically significant oxygen desaturations
- Sucking feeds and adequate weight gain; 150-200 grams/week or 10-20 gm/kg/day
- Consider tube feeds at home is suitable infants
- Hearing screening completed
- Referrals and follow-up arranged as below

Parent / Caregiver

- Confident in infant feeding, mouth and plate care and basic care/positioning of their infant. Education for Safe Infant Sleeping
- If dental plate in situ, appropriate equipment provided at discharge (2 bonnets of appropriate size, small torch, 'Bonnet, and Strapping Kit', Parrafin).
- Squeezy bottles can be purchased from the Dental department or from CleftPALS Association of WA
- Completion of the Gastric Tube Feeding Learning Package if gastric tube feeds are required.

Outpatient Referrals and Follow up

Referral Service	Additional Information
Speech pathologist Cleft palate	Follow up service provided by speech pathologist for feeding support as below: • Phone review in 4 weeks • Outpatient review in 6 weeks • Then review at 4-6 months, 9-12 months and post cleft repair follow up.
Plastic surgery clinic **e-referral**	Cleft lip and palate, out-patient review planned 6 weeks after discharge
Dental clinic **e-referral**	Cleft palate, out-patient review planned 1 week after discharge if dental plate in situ
ENT referral **e-referral**	If cleft palate: review in clinic at 6 months with audiogram
Genetic Services **e-referral or Central Referral Service**	If suspected syndrome on clinical examination
Hearing Assessment	Refer to PCH Audiology/ENT if failed newborn hearing screen. All infants with cleft lip and palate will be referred in the first year of life
Neonatology	No routine follow-up organised with Neonatal team, at discretion of discharging clinician based on other associated medical issues. Routine follow up with GP

Related CAHS internal policies, procedures and guidelines

Neonatology Guidelines

Pierre Robin Sequence

Gastric Tube Feeding – Going Home

Genetics Referral Pathway

References and related external legislation, policies, and guidelines *(if required)*

1. Eom, J. S. & Lee, T. J. (2011). Changing Trends in Cleft Lip and Palate Management Taught in Training Programs in Korea. *Journal of Craniofacial Surgery, 22* (2), 430-433. doi: 10.1097/SCS.0b013e31820745e1.
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9. Sharma RK, Nanda V. Problems of middle ear and hearing in cleft children. Indian J Plast Surg 2009;42:144-8.
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11. Leslie EJ, Marazita ML. Genetics of cleft lip and cleft palate. Am J Med Genet C Semin Med Genet. 2013 Nov;163C(4):246-58. doi: 10.1002/ajmg.c.31381. Epub 2013 Oct 4. PMID: 24124047; PMCID: PMC3925974.

Useful resources

https://www.cleftpalswa.org.au/

Infant Monitoring Clinic – Home Use Guide

Safe Infant Sleeping

This document can be made available in alternative formats on request.

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Appendix 1: Care and Feeding of an Infant with Cleft Lip and Palate

Equipment - Admission Pack Found in Cleft Trolley

- Pigeon (squeezy) bottle - ordered from milk room
- Laryngoscope
- Paper cups
- Chlorhexidine gel
- Soft white paraffin gel
- Plastic medicine spoon
- Boiled water (kept by patient bedside, replenished every 24 hours)
- Shortened buffered swab sticks

Infants with cleft lip and or palate are routinely fed with a pigeon (squeezy) bottle with a short, fast teat as they are generally unable to create an adequate seal to feed with a normal bottle or “strip” the breast with the tongue against the hard palate whilst breast feeding.

- When feeding a baby with a pigeon (squeezy) bottle, the bottle is gently squeezed as the baby sucks. Squeezing the bottle ceases when the infant is not sucking.
- Infants should be fed in a sitting up position to prevent the risk of aspiration. If a baby is fed in the more usual cradled position, milk may pool in the oral cavity and aspiration may occur.
- Milk needs to be warmed to body temperature to reduce the incidence of vasovagal episodes from cold milk entering the nasal cavity. Warming the milk also softens the bottle making it easier to squeeze.
- As infants with clefts do not maintain a good seal, they tend to swallow a lot of air with the feed and therefore require winding/burping at the middle and end of the feed.
- Feeds should take approximately 30 minutes; extended feeds limit the baby’s ability to rest. Feeds should be given 3 to 4 hourly to demand.
- Breast feeding, although difficult, may be possible for infants with minor clefts, especially after a good milk supply has been established. Mothers should be encouraged to express breast milk. Expressed milk can be given via a pigeon bottle. Please discuss with the Lactation consultant on Ward 3B after admission.
- Mouth care should be given after each feed for the first 48 hours then twice daily.
- Nasogastric tube (NGT) feeding is not generally required; infants should be encouraged to suck all their feeds. If NGT feeding is required, the NGT should

be inserted into the unaffected nostril. If placed through the clefted side, the NGT is more difficult to stabilize and can migrate between the nasal and oral cavity, causing possible erosion to the mucosa.

- Dummies are discouraged as they can cause erosions around the cleft and cannot be used in the later post-operative phase as it puts strain on the suture and may interfere with the healing process.

Suggestions for Positioning the Infant for Breastfeeds

Mothers should be encouraged to provide the protective benefits of breast milk, in preference to formula milk.

Counselling is required about the likely success of breastfeeding. There is moderate descriptive evidence that babies with CL can generate suction and Cleft Lip and Palate Page 4 of 7 Neonatal Guideline successfully breastfeed. Evidence suggests that direct breastfeeding is unlikely to be the sole method of feeding for babies with CP or CLP as they may have difficulty generating suction and have inefficient sucking patterns.

- Babies with CL/CLP should be assessed individually for their ability to breastfeed successfully, including type of cleft, mother's wishes and previous experience.
- Modification of breast-feeding positions may increase the efficiency and effectiveness of feeding. Infants with CL should be held so the CL is orientated towards the top of the breast.
- The mother may occlude the CL with her thumb or finger and/or support the infant's cheeks to decrease the width of the cleft and increase closure around the nipple.
- Positioning should be semi upright to reduce nasal regurgitation and reflux. This may be facilitated with a football hold with infant's shoulders higher than the body.
- Mothers may need to manually express breast milk into the baby's mouth to compensate for absent suction, and compression and to stimulate the let-down reflex.

Appendix 2: Orthodontic appliance for cleft palate

Orthodontic Appliance (For Cleft Palate)

The orthodontic appliance (OA), “dental plate”, facilitates moulding of oral structures. It acts as an obturator and assists with feeding by creating a false palate. It also prevents the tongue from migrating into the cleft and protects against erosions due to the activity of the tongue. The dentist will take an impression with alginate for an OA if considered necessary. The neonatal nurse assists ensuring oxygen and suction, are available.

Note: the mouth must be visualised after the impression and suctioned to ensure no alginate remains as this may be a choking hazard.

Creation of the plate takes 12 to 24 hours and then is fitted by the dentist. Dentists usually review the plate and palate each day to ensure a good fit and to check for areas of ulceration in the mouth caused by the OA. Using an OA creates less room in the oral cavity, which can make initial feeding more difficult. The OA remains in situ at all times apart from routine mouth care and cleaning.

Routine Mouth Care

1. Performed after each feed for the first 48 hours after the OA is fitted; then twice daily thereafter.
2. Remove the OA and wash in cooled boiled water.
3. Give the infant sufficient room temperature boiled water by teaspoon to effectively rinse the mouth and keep the mouth relatively curd free; usually 1 teaspoon. **Note: a vaso-vagal reaction can occur if cold water is used.**
4. Remove the OA from the water and apply a small amount of chlorhexidine gel to both sides of the plate; TDS for the first 48 hours and then twice daily thereafter.
5. The mouth should be inspected for any new areas of ulceration, bleeding and tooth eruption, using the laryngoscope. The OA may need to be adjusted by the Dentist.
6. Using a moist, swab sticks clean under the flattened nostril.
7. Carefully insert the OA slightly sideward for a unilateral cleft and straight for a bilateral cleft.
8. Apply soft white paraffin to all lip areas and the pre maxilla as needed and at each feed time.
9. Mouth care is not required for isolated clefts and in babies with Pierre Robin Sequence.

Bonnet & Strapping

- Commences **ONLY AFTER** the insertion of the OA. Strapping consists of a piece of soft elastic, fitted individually to the infant. It should sit on the protruding maxilla and cover the cleft lip, but it must rest on skin and never on oral mucosa.
- Pressure areas can occur if the strapping is too tight and be ineffective if the strapping is too loose.
- Bonnets should be tied snugly at the side, checked frequently and tightened when necessary.

- The cheeks should bulge over either side of the elastic. The position should be checked frequently by staff and parents.
- Each infant should be individually fitted for 2 bonnets. They are taken home and replaced in the dental clinic as required.
- Bonnets are washed at bath time and dried at the cot side.
- Strapping is always kept in place other than feeding and bathing.

Unilateral Cleft:

Start strapping from the non-cleft side and pull over the cleft; fix with Velcro tabs.

Bilateral Cleft:

The strapping is placed on the pre-maxilla and should cover the width of the cleft and the tabs are affixed to both sides of the bonnet in one motion.