

Recommendations of Practice

Background

• A cleft lip and/or palate is a craniofacial condition which is caused by the atypical development of the embryo in around Week 4-10

Te Whatu Ora

Health New Zealand

- The incidence of clefts in New Zealand is (1:565); 1.79 for every 1000 live births, compared to the general worldwide population which is approximately (1:700). The difference in New Zealand statistics is attributed to the higher incidence of cleft palate in the Maori population which is 2.37 per 1000 live births
- An isolated cleft palate is commonly only able to be diagnosed postnatally.

Objective

• To optimise guidance and ensure a standardised approach is followed for the accurate detection and management of fetuses presenting with a cleft lip and/or palate.

Definition

- A facial cleft is a separation or gap in the baby's lip which may extend into the soft and/or hard palate and nasal cavity. Clefts range in severity from isolated cleft lip to cleft lip and palate. Clefts can be unilateral, bilateral, complete or non-complete (see Bluebook for illustrations).
- Classifications:
 - Incomplete unilateral cleft lip
 - Complete unilateral cleft lip and palate
 - o Bilateral cleft lip and palate
 - Cleft of hard and soft palate.

Differential Diagnosis

- Normal philtrum
- Amniotic band sequence
- Chromosomal i.e. Trisomy (13/18)
- Syndromic vs non-syndromic aetiology
- Micrognathia (Pierre Robin Sequence)
- Facial mass
- Facial cleft (Tessier cleft).

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Important History

• Family history of cleft lip or palate, or genetic syndrome.

Ultrasound

- Nose/lips view coronal is most suitable including clips and 3D imaging
- Facial profile to exclude micrognathia
- Superior alveolar ridge views/hard palate views
- Uvula views
- Detailed anatomy review to exclude other abnormalities (microngathia, midline defects, spinal anomalies)
- 24+ week echocardiogram if adequate views not obtained at anatomy scan
- Growth surveillance
- Amniotic fluid volume
 - Polyhydramnios may be an indicator of potential airway issues at birth.
- Cleft location important for predicting associated abnormalities:
 - Unilateral cleft lip/palate 10%
 - Bilateral cleft lip/palate 25%
 - Midline cleft lip/palate 100%.

Investigation

- Offer invasive testing for microarray
- Syndrome
 - Approximately 300 syndromes are associated with cleft lip and palate
 - Around 30% of cases will have a syndromic cause
 - The exact syndrome may not be able to be identified antenatally.

Prognosis

- Isolated cleft lip and or cleft lip/palates have an excellent prognosis
- Fetuses with associated trisomies, syndromes and or other related anomalies have a poorer prognosis, making overall outcome variable.
- Associated craniofacial problems (defect dependant):
 - o feeding difficulties
 - hearing and speech impairment.

Management /Treatment

- The management of an isolated cleft lip will differ from the management of cleft lip and palate (CLP)
- CLP requires a long-term, multidisciplinary treatment strategy that will be evaluated and tailored to individual cases depending on complexity
- Antenatal referral to a Lactation Consultant and CLP team may be helpful for the whānau
- Specialities involved:
 - o Maternal Fetal Medicine Specialist
 - o Plastic Surgeons
 - o Cleft lip/palate nurse/co-ordinator (primary link for integrating services)
 - \circ Orthodontics
 - o Paediatric Dentistry
 - o Oral/maxillofacial surgery
 - Speech language therapy

- \circ Genetics
- Otolaryngology (Ear, Nose and Throat Specialist)
- Audiology
- Paediatrics
- Lactation consultants
- Babies with bilateral CLP and/or polyhydramnios may require delivery at a tertiary centre depending on local services' availability for potential airway support at birth.

Surgical Management

- Cleft lip repair around 3-6 months
- Cleft palate repair around 9-12 months
- Additional cleft-related surgeries may be required and are case by case dependent: ie cleft revisions for obtaining facial symmetry, ongoing orthodontic review to assess missing or displaced teeth, braces for realignment, bone grafting to repair affected gum line, and in some instances jaw surgery.

This Recommendation of Practice was created by Nicola Harrison with input from members of Wāhi Rua NZMFM Network. Endorsed in March 2023

The most up to date version of this Recommendation of Practice can be found on Healthpoint Wāhi Rua: New Zealand Maternal Fetal Medicine Network (NZMFM) webpages: <u>https://www.healthpoint.co.nz/public/wahi-rua-new-zealand-maternal-fetal-medicine/</u>

References

- Bartlett, G., de Chalain, T. *Surgery*. Cleft New Zealand. Retrieved December 4, 2022 from https://www.cleft.org.nz/healthinfo/surgery/
- Cleft New Zealand: Te Manatopu Ngutu Riwha o Aotearoa: *The Blue Book: A handbook for parents of children born with cleft lip/palate- Blue Book 1 Antenatal to age one*. 5th edition
- <u>Health Research Council of New Zealand (n.d). Retrieved December 4, 2022 from</u> <u>www.hrc.govt.nz/resources/research-repository/environmental-and-genetic-risk-factors-cleft-</u> <u>lip-and-palate</u>
- Mink van der Molen *et. al.* Clinical Practice Guidelines on the Treatment of Patients with Cleft Lip, Alveolus, and Palate: An Executive Summary. *Journal of Clinical Medicine*. 2010(21):4813.
- Smit R, Fowler PV. Non-syndromal orofacial clefts in the Canterbury/West Coast region during the 2000-2009 period. *New Zealand Dental Journal*. 2010: 106(4), 129-31.
- Thompson JM, Stone PR, Sanders M, van der Zee H, Borman B, Fowler PV. The incidence of Orofacial Cleft in live births in New Zealand. *New Zealand Medical Journal* 2016:19(129), 64-71.
- Vyas T, Gupta P, Kumar S, Gupta R, Gupta T, Singh HP. Cleft of lip and palate: A Review. *Journal Family Medical Primary Care*. 2020 Jun 30;9(6):2621-2625.
- Woodward, et al. (2005). Diagnostic Imaging, Obstetrics. Amirsys.