GUIDELINE

Choanal Atresia

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 standardized management and follow up to infants and families of infants with Choanal Atresia's.

Risk

Infants with Choanal atresia may not receive appropriate inpatient management and follow-up.

Background

The nasal choanae are paired openings that connect the nasal cavity with the nasopharynx. Choanal atresia is a congenital condition in which these openings are occluded by membranous soft tissue, bone, or a combination of both due to failed recanalization of the nasal fossae during fetal development. The blockage can be either completely bony (30 percent of cases) or be composed of both bone and membranes (70 percent of cases). There is no known specific cause of choanal atresia. Most believe that choanal atresia occurs when the tissue that separates the nose and mouth area during fetal development remains after birth. It occurs in 1:5000 to 1:8000 births. It is more often unilateral than bilateral (60% vs. 40%) and occurs more frequently in females than in males (ratio 2:1).

Clinical presentation of choanal atresia varies from acute life-threatening airway obstruction to chronic recurrent nasal discharge on the affected side, depending on a unilateral or bilateral nature of the abnormality.

- Unilateral choanal atresia is often asymptomatic in neonatal period.
- Bilateral choanal atresia is an acute otolaryngologic emergency. The interprofessional team should promptly recognize this condition to avoid severe morbidity and mortality.

Bilateral choanal atresia, presents with episodes of acute respiratory distress with cyanosis that is relieved with crying and with the return of cyanosis with rest as newborns are obligatory nose breathers (paradoxical cyanosis).

Feeding difficulty can be the initial alerting event in which the infants can present with progressive airway obstruction and choke episodes during feeding because of their inability to breathe and feed at the same time.

Unilateral choanal atresia rarely presents with respiratory distress. The most common presentation is a purulent nasal discharge and obstruction on the affected side and/or a history of chronic sinusitis.

In some cases, the correct diagnosis is not reached until adulthood due to the non-specific symptoms of unilateral nasal obstruction.

The clinical suspicion of choanal atresia can be confirmed by examination with a flexible nasal endoscope and CT scan which further delineates the characteristics of malformation and useful in differentiating from other causes of nasal obstruction.

- Differential diagnoses include pyriform aperture stenosis, nasolacrimal duct cysts, turbinate hypertrophy, septal dislocation and deviation, antrochoanal polyp, or nasal neoplasm.
- Choanal atresia may be associated with various other anomalies (in 50-70% cases), CHARGE syndrome is the most common of these and consists of coloboma, heart disease, atresia choanae, growth, and mental retardation, genital hypoplasia, and ear anomalies.

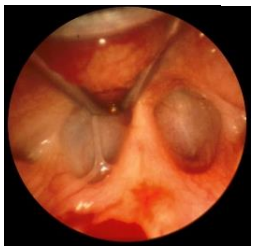
Clinical Description

Bilateral choanal atresia

- It is a medical emergency as neonates are obligatory nose breather
- Cyclic respiratory distress relieved with crying
- Cyanosis
- Noisy breathing
- Feeding difficulties
- Inability to pass a size 6Fg nasogastric tube into the nasopharynx through both nostril
- Nasal drainage

Unilateral choanal atresia

- Often asymptomatic in neonatal period
- Nasal discharge
- Mouth breathing
- Inability to move air on one side
- Noisy breathing
- Fewer feeding difficulties
- Inability to pass a size 6Fg nasogastric tube into the nasopharynx through one nostril



Choanal atresia or stenosis

Common associated genetic conditions (in 50-70 % cases)

- Charge syndrome
- Treacher Collins syndrome
- Crouzon syndrome
- DiGeorge syndrome
- Acrophalyngosyndactyly
- Amniotic band syndrome
- Malrotation of the bowel
- Antley-Bixter syndrome
- de Lange syndrome
- Chromosome 18, 12, and XO anomalies
- Polydactyly
- Colobomas
- Clefts of the face, nose, and palate
- Congenital heart disease
- Tracheoesophageal fistula
- Craniosynostosis

Feeding and Growth (more common in bilateral atresia)

- Swallowing discoordination
- Poor suck swallow coordination in presence of cleft
- Aspirations due to upper airway weakness and stent in situ
- Poor weight gain
- Gastroesophageal reflux
- Hyponatremia due to losses in excessive secretion

Initial Neonatal Management

Management	Additional Information
1. Admit to the NICU following delivery, transfer 3B PCH when stabilised.	
2. Cardiorespiratory monitoring, nurse in supine position.	Note: Prone position carefully considered only if tongue-based obstruction suspected with syndromic association.
3. If clinically significant airway obstruction, consider inserting Guedels airway or LMA.	Secure adjunct airway such as Guedel airway or LMA using brown tape with central hole cut to diameter of airway. Appendix 1
4. Suction airway 2 to 4 hourly as required.	
5. Severe cases of upper airway obstruction may require Intubation. Consider alterative airway devices for difficult airways.	Refer to NETS WA Airway Management and Neonatology Difficult Airway and Intubation guidelines.
6. Insert orogastric tube for feeding and decreasing gastric air.	Enteral feeds can be commenced through OGT as soon as clinically stable.
7. Clinical examination of the baby by the Senior Registrar or Neonatal Consultant to evaluate for syndromic association.	If there are clinical concerns, refer to genetics for an inpatient review. Genetic Referral Pathway guideline
8. Complete microarray- parental consent required. (MR411.00 Neonatal Consent for Genetic Testing and Information Sharing)	Enter patient details in the neonatal follow-up book for chasing the results, located in 3B Level 2 nursery.
9. Test for charge syndrome in all infants with bilateral choanal atresia which requires specific genetic testing as cannot be excluded if initial microarray test normal.	Organise for CHD7 testing after discussion with geneticist on call.
10. Referral and assessment of suck feeding by the feeding team and experienced bedside nurse.	
11. All infants to be referred to ENT for a Flexible Nasopharyngeal Endoscopy (FNE) as soon as possible.	
12. MRI of head and ear in consultation with ENT team.	Note: Request for special semicircular canal views for ears.
13. CT scan of face additionally required for operative planning for choanal atresia	
14. Additional abdominal ultrasound organised to rule out associated anomalies.	

Definitive Management of Choanal Atresia

The treatment of choanal atresia is essentially surgical. The objectives are to restore choanal patency, not to interfere with the patient normal craniofacial development, to minimize invasiveness, and to avoid recurrences.

Mild cases (mostly unilateral choanal atresia): often no respiratory distress or significant feeding issues

- Close observation
- Supplemental oxygen as needed
- Nasal saline drops or spray to keep the nasal linings healthy and free of discharge
- Definitive surgery often planned for older age

Severe cases

- Treated as soon as possible
- Establish secure airway
- Surgery often required
    - Five different surgical approaches have been proposed, including transpalatal, transeptal, sublabial, transantral, and transnasal. The transpalatal and endoscopic endonasal approach is more commonly used.

Post-Operative Management

- Multidisciplinary approach/discussion relating to airway management at the fortnightly Complex Airway Team (CAT) Meeting
- Maintaining adequate weight gain is essential.
    - Consider 24 calorie per ounce feeds to supplement nutritional intake, to achieve adequate growth.
    - Monitor serum and urine sodium level (if concerns of excessive losses in secretions) and consider the need for sodium supplementation for adequate growth.
- Stent management: Nasal stents could be unilateral or bilateral. Stents are at high risk of blockage due to excessive secretion.
    - This is a significant safety concern with bilateral stent in situ (Appendix 2)

Discharge Planning

Discharge age is dependent on type of stent and other associated syndromic association. It is preferable to discharge infants without nasal stent or unilateral stent in situ. Discharge with bilateral nasal stent is carefully considered based on clinical stability of infant, social dynamic of family and parent comforts level.

Safe Discharge Criteria

Infant

- No clinically significant bedside desaturations.
- Discharge home in supine position.
    - Prone position carefully considered if tongue-based obstruction suspected with syndromic association.
- Commenced on suck feeds and deemed safe by the feeding team.
- Adequate weight gain; 150-200 grams/week.
- Reviewed by ENT Clinical Nurse Specialist, ENT specialist and Feeding Team prior to discharge.
- All infants discharged with a home monitor, supplied from home monitoring unit both for unilateral and bilateral choanal atresia.

Parent / Caregiver

- Confident in feeding the infant.
- Confident with the basic care/positioning of their infant.
- Completion of the Gastric Tube Feeding – Going Home if gastric tube feeds are required.
- Meet with the 3B CNS to assist with discharge planning and safe transition home. The CNS will liaise with caregiver to provide stent care training, monitoring and equipment training and supply ordering.
    - The ENT CNS (ext: 65507) is also available for stent management and training.
- Home suction unit (hired through the PCH ECS).
- Supply of suction catheters, saline and syringes and referral to PCH ECS for ongoing supplies.
- Education for Safe Infant Sleeping
- Attendance at Infant CPR training.
- Home monitoring supplied and parent/s competent with the use of the monitor
- Car seat testing - 30-60 minutes: no desaturation.
- Rural/regional families – travel advice: adult to travel in the back seat with the infant during long car trips. Ensure suitable breaks if journey is a significant distance.
- Provide information about available resources.

Outpatient Referrals

Referral Service	Additional Information
Home Monitoring Clinic	**e-referral and phone call handover:** 1 week prior to discharge to arrange home monitor.
Feeding Team	**e-referral:** Advise of plans to discharge home with expected feeding methods
ENT CNS	Fortnightly phone consult with parents/caregiver post discharge until ENT clinic appointment
ENT Clinic	**e-referral:** Please state “Choanal atresia infant for review with ENT Consultant 6 weeks post discharge”
Hospital in the Home (if in catchment area)	**e-referral and phone call handover**: for infants going home with gastric tube feeds or with nasopharyngeal stents
Ophthalmology services	**e-referral:** Please state “Choanal atresia infant for review in first year of life due to high rate of associated anomalies”
Audiology	**e-referral:** All infants with choanal atresia will be referred to Audiology for further assessment in the first year of life.
Genetic Services	**e-referral or Central Referral Service** Genetics Referral Pathway
Neonatal Follow-up Clinic	SR clinic in 2 weeks. No further follow-up organised with neonatal services
General paediatrics Future: “Charge syndrome clinic’	Infant with bilateral choanal atresia is preferably referred to general paediatric follow up for long-term management.

Related CAHS internal policies, procedures and guidelines

Neonatology Clinical Guidelines

- Difficult Airway (Neonatal)
- Gastric Tube Feeding – Going Home
- Genetics Referral Pathway
- Intubation

NETS WA Guidelines

- Airway Management

References

1. Murray S, Luo L, Quimby A, Barrowman N, Vaccani JP, Caulley L. Immediate versus delayed surgery in congenital choanal atresia: A systematic review. Int J Pediatr Otorhinolaryngol. 2019 Apr; 119:47-53.
2. Meleca JB, Anne S, Hopkins B. Reducing the need for general anesthesia in the repair of choanal atresia with steroid-eluting stents: A case series. Int J Pediatr Otorhinolaryngol. 2019 Mar; 118:185-187.
3. Chan J, Ullas G, Narapa Reddy NVS. Completely Endoscopic Approach Using a Skeeter Drill to Treat Bilateral Congenital Choanal Atresia in a 33 Week Born Pre-term Baby. Indian J Otolaryngol Head Neck Surg. 2018 Dec;70(4):608-610.
4. Kurosaka H. Choanal atresia and stenosis: Development and diseases of the nasal cavity. Wiley Interdiscip Rev Dev Biol. 2019 Jan;8(1):e336.
5. Smith MM, Ishman SL. Pediatric Nasal Obstruction. Otolaryngol Clin North Am. 2018 Oct;51(5):971-985
6. Zawawi F, McVey MJ, Campisi P. The Pathogenesis of Choanal Atresia. JAMA Otolaryngol Head Neck Surg. 2018 Aug 01;144(8):758-759.
7. Mohan S, Fuller JC, Ford SF, Lindsay RW. Diagnostic and Therapeutic Management of Nasal Airway Obstruction: Advances in Diagnosis and Treatment. JAMA Facial Plast Surg. 2018 Sep 01;20(5):409-418.
8. Baumann I, Sommerburg O, Amrhein P, Plinkert PK, Koitschev A. [Diagnostics and management of choanal atresia]. HNO. 2018 Apr;66(4):329-338.
9. Moreddu E, Rossi ME, Nicollas R, Triglia JM. Prognostic Factors and Management of Patients with Choanal Atresia. J Pediatr. 2019 Jan; 204:234-239.e1
10. Rajan R, Tunkel DE. Choanal Atresia and Other Neonatal Nasal Anomalies. Clin Perinatol. 2018 Dec;45(4):751-767.

Useful resources
Infant Monitoring Clinic – Home Use Guide Safe Infant Sleeping Choanal Atresia | Children's Hospital of Philadelphia (chop.edu) Choanal_atresia_F0564_A5_col_FINAL_Sep17.pdf (gosh.nhs.uk) Nasal_stents_F0176_FINAL_Nov15.pdf (gosh.nhs.uk)

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

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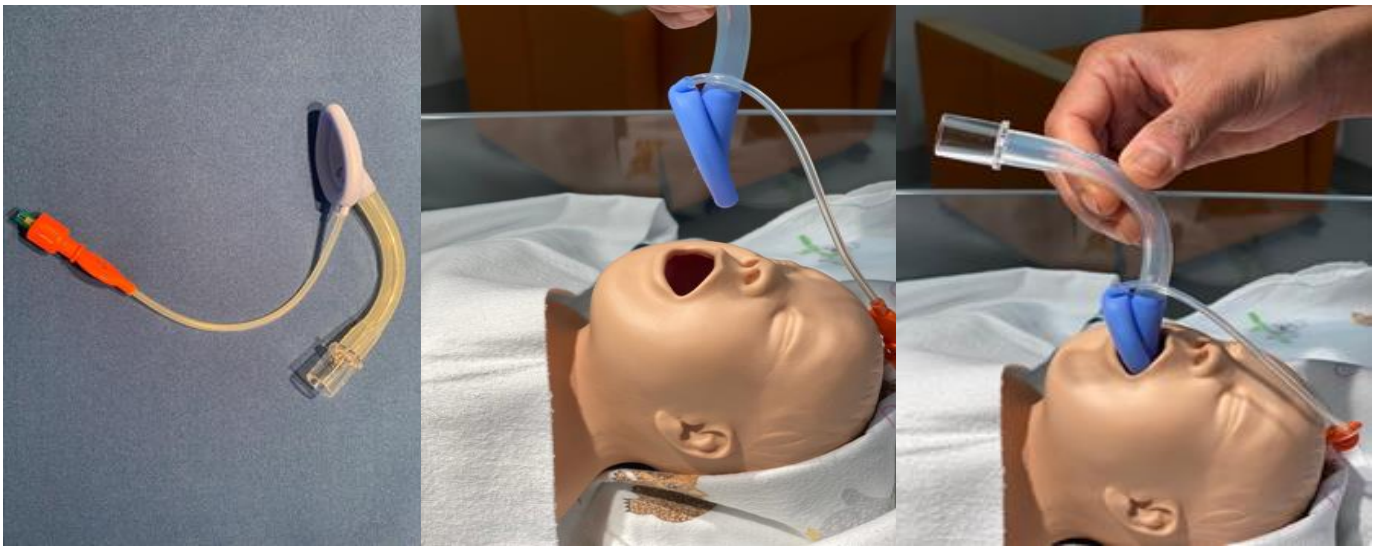
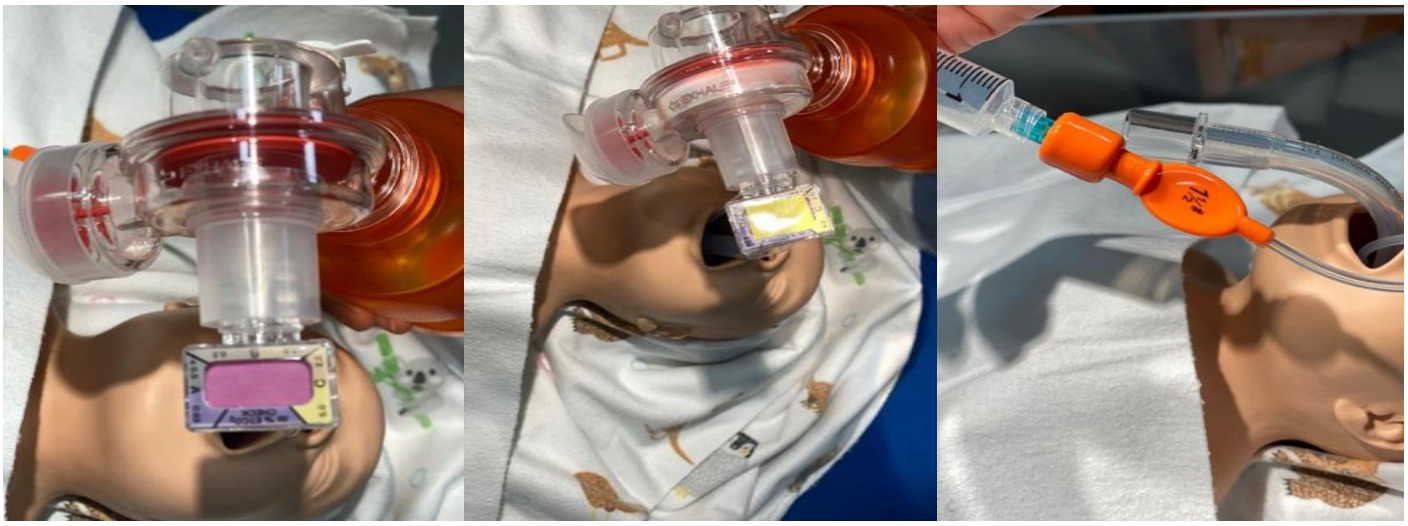
Appendix 1: Laryngeal Mask Airway (LMA) Insertion

LMA are only suitable for use in neonates over 1.5kg (Size 1.5 up to 5kg)



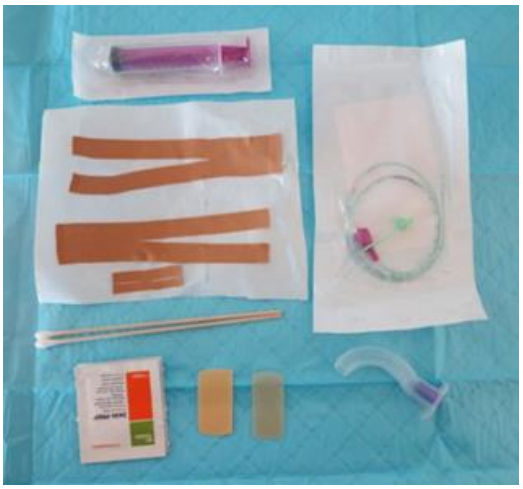
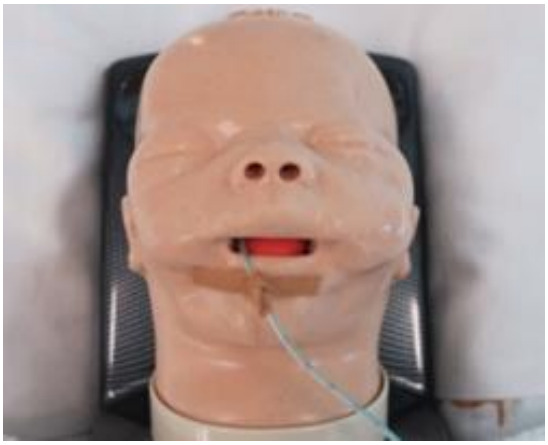
Size 1.5 LMA. Deflate cuff prior to use. Lubricate the LMA. Head in neutral position, flatten the LMA tip against the hard palate, guiding the LMA tip against the hard palate, guiding the LMA in a cranial direction following contour of hard and soft palate until resistance felt at the base of hypopharynx.

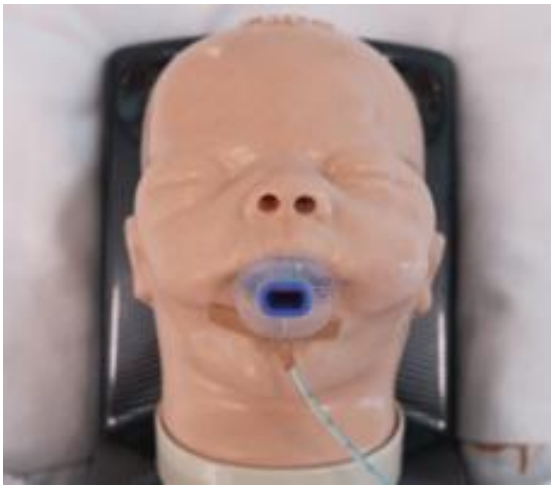
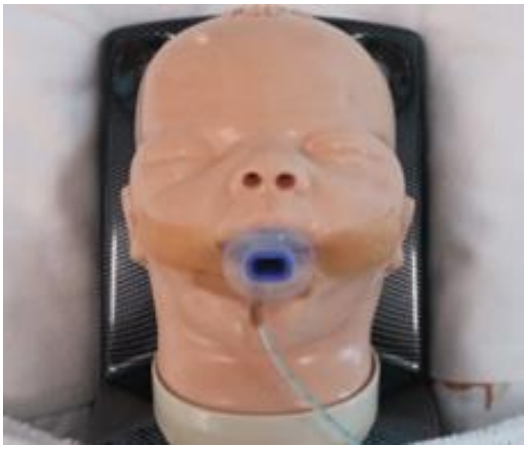
Attach CO_2 detector to see a colour change to confirm the correct location, a positive result is purple to gold. Inflate the cuff with recommended air volume. Ensure adequate chest wall movement.

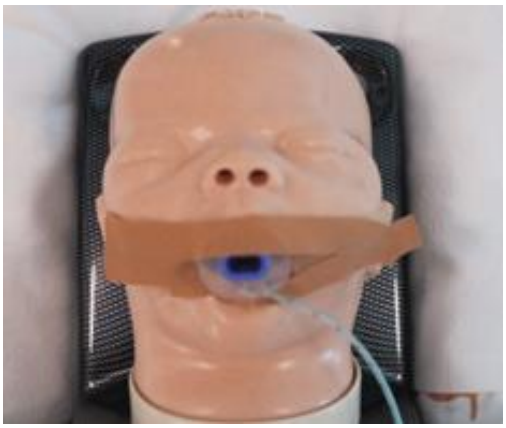
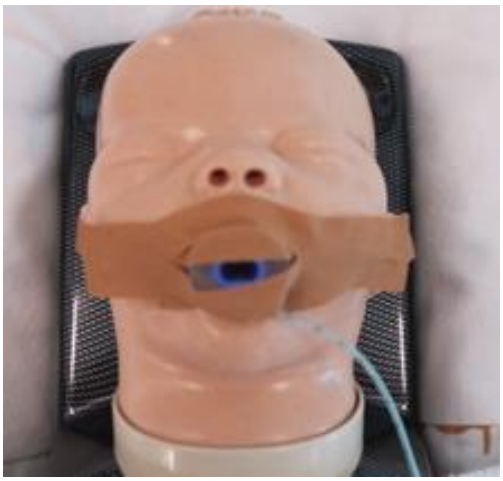


Tape the LMA in place as shown above. Cut two separate pieces of brown tape, cut as trouser legs. Place the first leg under the mouth and 2nd leg around the tube. With the 2nd piece of tape place one leg over the mouth and the second leg around the tube.

Insertion and Securement of Oropharyngeal Airway (OPA) / Guedels

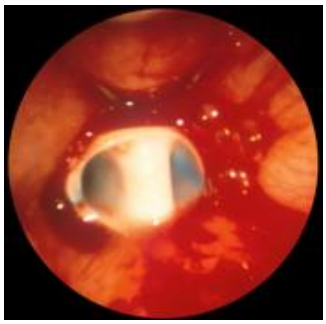
Steps	Additional Information
1. Gather the appropriate equipment to insert a feeding tube and an oropharyngeal airway.	
2. Insert the feeding tube orally and secure.	

Steps	Additional Information
3. Measure from the centre of the incisors to the angle of the mandible when laid on the face concave side up. Moisten the OPA using the patient's own saliva.	
4. Insert the OPA airway under direct vision, concave side down. If required use a tongue depressor to hold the tongue down.	
5. Once inserted place duoderm protection from the corner of the mouth to the outer middle cheek – use skin prep to help the duoderm secure to the skin.	

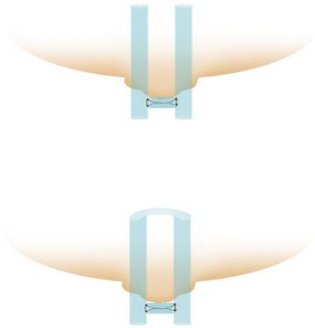
Steps	Additional Information
6. Using trouser leg taping secure the OPA across the top ensuring the opening of the airway is not occluded, then secure along the lower of the lip.	
7. Secure the second trouser leg taping in the opposite order securing the bottom of the OPA, then secure along the top of the lip. 8. OPA's should be removed every 8 hours. The mouth should be inspected for ulcers and mouth care attended. 9. OPA's are single use for 24 hours and should be replaced daily.	

Appendix 2: Nasopharyngeal Stent Management

Bilateral stents are often U-shaped stents with a suture running along the side of the stent



Posterior aspect of nasal stent



Stents are positioned through the new choanal openings using ET tubes. The tube size will depend on the size of the created opening.
The ET tube is cut and inserted via the mouth to stent the choanal. A smaller piece of tube is sutured across the front to prevent the tube sliding into the nose

Management of Stents	Additional Information
Patient RTW with stents insitu.	3B CNS or ENT CNS will create an airway profile for the stents to be placed at the bedside.
Stents require 1 to 2 hourly suction for the first 48 hours to maintain patency, then pre feed and prn.	The size of the suction catheter used will depend on the size of the stent.
Post-operative orders may contain instructions regarding stent care and suction. Each surgeon will have their own preference for stent care & management. Ensure these are checked.	The suction catheter should then be inserted to the correct length of the stents as documented by the surgical team. A tape measure cut to the correct length should be taped to the warmer or cot for visible confirmation of catheter length.
Only Suction Catheters are to be used to suction the stents.	0.9% Sodium chloride can be instilled into each stent prior to suction. Using a blunt cannula in 0.2 mL increments.
Gentle suction around the stents should also be applied regularly.	Use an appropriately sized suction catheter or small neotech little sucker.
Patency is checked by auscultation with stethoscope and or by placing a mirror near the nose and observe misting on expiration through the stents.	Stents can become blocked with secretions.
Suck feeds can commence when the infant is stable and able to tolerate feeds.	Ensure feeding team review prior to commencement of suck feeds. OGT feeds are given if not able to suck feed.
During hospitalisation the infant requires continuous saturation monitoring.	Home Monitoring Assessment will happen prior to discharge.
Stents are normally left in situ for 6 weeks then removed by the surgeon and patency observed via endoscope.	