GUIDELINE

Oesophageal Atresia / Tracheoesophageal Fistula

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

The aim of this guideline is to outline the general principles in the diagnosis and pre and post -operative management of neonates with oesophageal atresia (OA) +/- trachea-oesophageal fistula (TOF). Special attention has been paid to H-shaped trachea-oesophageal fistula whose diagnosis may be missed or delayed if clinical index of suspicion is not high.

Risk

Inappropriate management of an unrepaired oesophageal atresia could severely compromise the safety of the neonate, potentially resulting in, but not limited to, gastric rupture or aspiration pneumonitis.

Delayed diagnosis of H-Shaped TOF can lead to prolonged morbidities and potentially mortality due to acute aspiration.

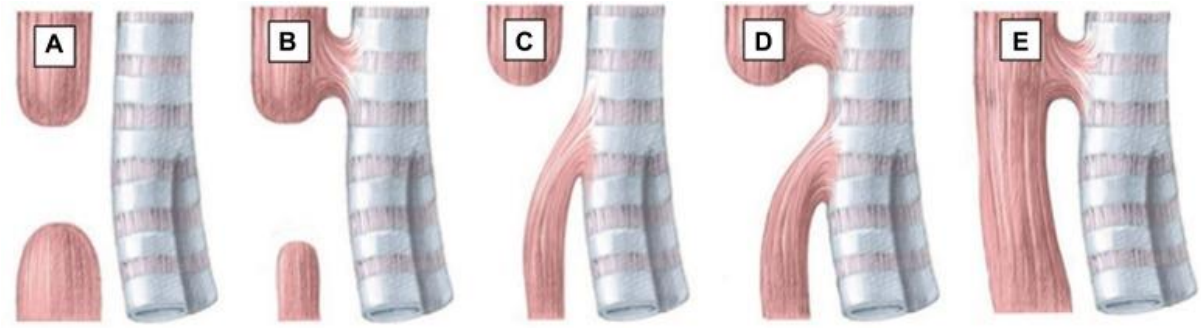
The non-adherence to guidelines post-operatively could lead to severe complications such as anastomotic leak, oesophageal rupture, and delayed healing of the repair site.

Definitions

- **Oesophageal atresia (OA):** a congenital anomaly in which the oesophagus ends in a blind upper pouch.
- **Tracheo-oesophageal fistula**: An abnormal connection between the trachea and oesophagus
- **Long-gap oesophageal atresia:** Either Pure OA or with a TOF where the gap between the proximal and distal blind ends is **>2 cm or >2 vertebral bodies** and where a primary oesophageal anastomosis is unable to be performed. Neonates with long-gap OA are often managed with the creation of a feeding gastrostomy and delayed oesophageal repair.
- **Replogle tube:** a double lumen tube used for the drainage of saliva. The tube is placed in the upper oesophageal pouch and connected to continuous low-pressure suction (-15cm to -20 cmH20), which allows drainage of saliva thereby preventing aspiration.

Types of Oesophageal Atresia / Tracheoesophageal Fistula

- Failure of the normal development of the oesophagus and separation of the trachea from the oesophagus results in a spectrum of anomalies that can result in one of the following:
    - Isolated (pure) oesophageal atresia (8% of cases) - Type A
    - Oesophageal atresia with proximal tracheoesophageal fistula (1%) - Type B
    - Oesophageal atresia and distal tracheoesophageal fistula (most common – 84% cases) - Type C
    - Oesophageal atresia with double tracheoesophageal fistula (3%) - Type D
    - H-shaped trachea-oesophageal fistula without oesophageal atresia (4%) - Type E.



Presentation

Neonates with OA may present with:

- Copious oral secretions post-delivery
- Coughing and choking with feeds or inability to swallow feeds
- Aspiration of feeds / secretions.
- Inability to pass an oro/nasogastric tube

There may also be a history of maternal polyhydramnios.

Clinical presentation of H-shaped TOF: A very high index of suspicion is needed for the diagnosis of H-TOF. Many times, its diagnosis is delayed until late in the first year of life. These babies will be able to swallow milk feeds because there is no oesophageal atresia. However, they have issues such as coughing after feeds, silent wet coughs, becoming slightly dusky after feeds, unexplained tachypnoea and recurrent aspiration pneumonias. Some of them will have abdominal distension due to the entry of air through the H-TOF into the stomach. If an excess of gas in the stomach is aspirated from the feeding tube, one needs to be alert to the possibility of H-TOF.

Diagnosis and Management

- In a neonate with suspected OA, an attempt should be made to **gently** pass a size 10 FG feeding tube **(a smaller bore tube may curl in the pouch).**
    - If this is met with resistance, an X-ray should be ordered with the feeding tube in place. If the upper pouch ends at thoracic vertebra 2 - 4 this is **indicative of OA**. Lower than this requires further investigation, such as a contrast study.
    - If the X-ray is indicative of an OA, a **replogle tube** should be inserted to prevent aspiration of secretions (size 8 for babies <2.5kgs, size 10 for babies >2.5kgs).
    - Insert the replogle tube orally until resistance is felt (usually around 8-10 cm from lips), then withdraw 0.5cm and secure.
        - Note the length that the replogle is taped at and record in the patient notes. Nurse the neonate with the head of the bed elevated.
    - The Replogle tube should be placed on continuous suction – See Set-up of Replogle Tube and Care of the Neonate with a Replogle Tube
- **Diagnosis of H-Shaped TOF**: If clinically suspected, ENT surgeons should be contacted, and an urgent Barium swallow requested. It will usually demonstrate the presence of fistula. If barium swallow is normal, but clinically H-shaped TOF is suspected, ENT team will do a direct laryngo-tracheo-bronchoscopy (LTB), which is the definitive test for this condition.
- Oesophageal atresia/TOF is often associated with vertebral, cardiac, gastrointestinal, genitourinary and limb abnormalities.
    - A neonate diagnosed with OA or TOF should be examined carefully to exclude further anomalies such as:

        1. **VACTERLS Association** (V = vertebral abnormalities; A = anal atresia; C = cardiac defects; TE = tracheal-oesophageal abnormalities; R = renal and radial abnormalities; L = limb abnormalities; S = single umbilical artery).

        2. **CHARGE Syndrome** (C = coloboma and cranial nerve defects, H = heart defects; A = atresia of the choanae; R = retardation of growth and development; G = genital underdevelopment; E = ear abnormalities and sensorineural hearing loss.
    - Renal, and head ultrasounds and X-Ray of spine should be performed in all infants with OA/TOF. Spinal ultrasounds are requested where indicated after discussion with the neonatologist.

- The need for genetic studies such as microarray should be discussed with the Neonatologist.

Pre-operative Management

- An **echocardiogram** must be performed pre-operatively to note the position of the aortic arch and detect any cardiac anomalies. A right sided aortic arch may necessitate a left sided thoracotomy instead of the conventional right sided thoracotomy approach.
  - Cardiology team should be contacted as soon as OA/TOF is diagnosed.
- Routine pre-operative management is per the pre-operative care guideline.

Ventilation in the pre-operative period

- Intubation and ventilation may be required for various reasons – for example, aspiration, infection, HMD.
- Positive-pressure ventilation can result in air entering the stomach via a distal TOF. This can result in catastrophic stomach rupture, especially if high pressures are used.
  - The early use of Surfactant has reduced the requirement for high pressure ventilation in those neonates with HMD.
- If an **emergency intubation** is required, the least possible ventilator pressures should be used.
  - Neonatal consultant and surgeon should be notified immediately.
  - Urgent chest & abdominal x-ray should be performed (AP and lateral)
  - In the case of a **pneumoperitoneum** or a **rapid deterioration** in the neonates condition, consider **emergency needle paracentesis of the abdomen**.
    - An urgent surgical consult will facilitate emergency ligation of the fistula +/- gastrostomy.
- Whilst CPAP is not strictly contra-indicated, it should be used in caution and after discussion with surgeons.

Post-Operative Management

- All babies post oesophageal atresia repair will come back ventilated.
  - Care should be taken to ensure correct ETT suction depth to avoid damage of the repaired fistula site. **Do not suction beyond the end of the ETT.**
- On return from theatre, the infant will have a **TRANSANASTOMOTIC TUBE (TAT)** in place.

  - The purpose of this tube is to act as a stent for the repair site, as well as for commencing nasogastric feeds.
  - It is a normal nasogastric tube being used as a stent. It is marked with green and yellow striped tape.
  - Aspirates should be measured, described, and recorded as for any nasogastric tube
  - The tape and position of the TAT must be checked regularly to ensure it isn't accidently dislodged.

UNDER NO CIRCUMSTANCES SHOULD THE TUBE BE REMOVED WITHOUT THE SURGEONS' PERMISSION.

If the TAT is accidentally removed, do not attempt to reinsert it - contact the surgeons immediately.

- Surgeons may ask for the neonate to be nursed in a supine position with **their chin tucked onto their chest** to reduce tension on the anastomotic site. This is done when there is a tight closure between the oesophageal structures. Ensure the post-operative surgical orders are checked.
- A chest x-ray (AP and lateral) should be done ASAP on return from theatre to check ETT position. Ensure Senior registrar or Consultant have reviewed the x-ray.
- Ensure the neonate is adequately treated with Morphine or Fentanyl, to provide pain relief and sedation. The need for muscle relaxant should be discussed with the Surgeon and Neonatal Consultant – considering the risk / benefit of long-term muscle paralysis.
- In infants with long-gap OA, the gap is often too large to perform a primary repair. In this case, a gastrostomy tube will be inserted to facilitate feeding until anastomosis can take place. (Note: Gastrostomy tubes are NOT aspirated pre-feed).
  - The replogle tube will remain in situ, and flushing will proceed as per guidelines.
- Most neonates will return from theatre with a chest (mediastinal) drain. This is usually a wound drain and is **not** placed on suction unless specifically ordered by the surgeon. Even low-pressure suction may cause damage to the newly repaired oesophagus.
- Neonates who require post-extubation support require approval from the Consultant and Surgeon before commencing CPAP or High-flow nasal prongs.

- Oropharyngeal suctioning must avoid the anastomotic suture line - the surgeon will clarify the length of the anastomosis from the mouth.
- Occasionally a **Foker procedure** is done for long-gap OA when the oesophageal ends remain too far apart for anastomosis
    - This process involves gradual stretching of the oesophageal ends – which thereby stimulates growth.
    - Surgeons perform daily tightening of externally placed ‘wires’ to help approximate the 2 ends of the oesophagus. Consider pain relief prior to the daily tightening of wires. The dressing should not be handled without the presence of surgeons.

Considerations for Feeding Neonates post OA Repair

- Any feeds commenced will be via the **TAT ONLY**.
- Neonates will require TPN and Lipids in the immediate post-operative period.
- An Oesophageal contrast study will be performed approximately 7 days postoperatively to detect any anastomotic leak prior to commencing oral suck feeds.

No oral feeds (bottle/breast) are to be given until ordered by the surgeon.

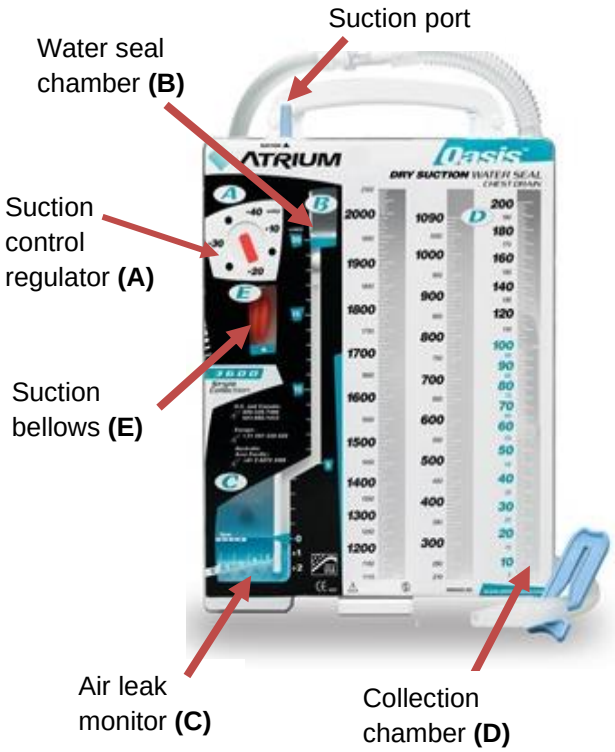
- Gastro-oesophageal reflux can be a significant problem in neonates post OA repair due to oesophageal dysmotility
    - The early use of omeprazole is often used to protect the anastomosis site and reduce reflux associated pain. It is usually continued for few months. Please check with surgeons regarding their preference for omeprazole duration.
    - The head of the bed should be elevated 30-45 degrees when the infant commences feeds.
- Post-op laryngomalacia or tracheomalacia is relatively common and can make feeding difficult and in worst case scenarios require CPAP. It presents as a barking cough or respiratory distress.

Set-Up of the Replogle Tube

Replogle tube with Atrium *Oasis* UWSD system (PCH)

Equipment

- *Argyle* Replogle tube size 8Fg (<2.5kg) or 10Fg (>2.5kg)
- Atrium *Oasis* Under Water Seal Drain (UWSD)
- Standard wall suction
- Suction tubing
- 2mL Leur-slip syringe and 0.9% Saline
- Sterile water

Set up of UWSD & Connection to Replogle	Additional Information
1. Remove the ampoule of sterile water from the back of the drain. Add water to the water seal chamber (B) through the suction port. Fill the water seal chamber to the 2cm fill line.	
2. Connect suction tubing to the suction port and wall suction. Turn on the wall suction pressure until the orange bellows (E) is expanded to the arrow – usually this is between -20mmhg and -70mmhg	This demonstrates that the suction is working. If the bellows is not at or past the delta mark, increase suction pressure at the wall. This is normal suction – NOT low flow.
3. Connect replogle tube to the suction tubing which is connected to the UWSD.	

Set up of UWSD & Connection to Replogle	Additional Information
4. Suction is usually dialled to commence at -15cmH_20 on the suction control regulator - indicated by (A) on Oasis UWSD.	Suction pressure is controlled through the *suction control regulator* on the UWSD – **not the wall suction.** Suction can be increased to -40cmH20 on the UWSD if required, after d/w Consultant and Surgeon and documented in patient notes.

Replogle tube management at KEMH

- Use the Atrium *Oasis* UWSD system if available. If unavailable place the replogle tube on continuous **low-pressure** suction of -20 to -30mmHg.

Care of the Neonate with a Replogle Tube

- Neonate with a replogle insitu is to be nursed with their head elevated.
- The replogle is flushed every 15 minutes to maintain patency.
- 0.5mL of 0.9% Saline followed by 0.5mL of air is flushed into the vent lumen of the replogle tube. The syringe is then removed which will further draw air in and allow for movement of oral secretions from the pouch. This fluid should be seen draining up the suction lumen of the replogle tube.
- Approximately every 4-6 hours the replogle tube needs a full flush. Keeping the tube attached to the face, flick the replogle tube out the neonate's mouth and flush 2mL of 0.9% Saline into a paper cup. Reinsert the replogle tube back into the neonate's mouth / pouch.
- Troubleshooting a blocked replogle tube - see Appendix 1
- Consider offering 'No Flow' teats as initial feeding experience prior to Sham feed commencement.

Nursing Checks and Documentation

- Ensure the level of sterile water in the suction control chamber on Atrium Oasis UWSD is maintained at 2cm. Top-up with sterile water if needed.
- Document the suction pressure hourly (reading from the suction control dial on UWSD)
- Document 15 minutely flushes on the observation chart
- Check replogle tube position hourly - ensuring it remains at the correct measurement
- Replogle tube and Atrium UWSD unit are changed weekly, or earlier if necessary. Label the UWSD with the time and date of change.
- Ensure spare Replogle tube is kept by bedside.

Sham Feeds

- Sham feeding is used to enable the neonate to learn how to feed orally, either by breast or bottle, prior to repair of the oesophageal atresia reducing the risk of developmental feeding delays including oral aversion.
- It is used for neonates with a long-gap oesophageal atresia who may not be able to feed orally for many months
- Sham feeding involves the neonate feeding via bottle or breast with a replogle tube connected to suction draining the milk from the upper oesophageal pouch to prevent aspiration.
    - After the feed is removed from the oesophageal pouch by suction it is then re-fed to the neonate via the gastrostomy tube to enable oral feeding to be associated with milk entering the stomach at the same time.

Indications for sham feeding:

- Neonates with long-gap OA awaiting a delayed repair by oesophageal anastomosis or oesophageal replacement surgery
- Infants should be tolerating >100 mL/kg/day of bolus enteral feeds via gastrostomy prior to commencing sham feeds
- The decision to sham feed is made in consultation with the surgical team, the Neonatologist, and the primary caregiver.

Assessment for suitability of sham feeding

- The first Sham feed should be conducted with the feeding team present – see appendix 2.
- Neonates should be >35/40 corrected age, requiring no respiratory support and able to coordinate sucking, swallowing and breathing
- Neonates >2kg can have their replogle in their nose instead of mouth to assist with feeding

See Appendix 2 for Sham Feeding Assessment and Procedure

Discharge Planning

Safe discharge criteria:

Multidisciplinary Team:

Appropriate referrals made for follow up and outpatient clinics. See Appendix 3.

Neonate:

- Nil concerns for secretion management

- Commenced on suck feeds and deemed safe on feeding assessment by the feeding team (Exception for long-gap OA)
    - Some patients may still require enteral feed top-ups at time of discharge
- Adequate weight gain; 170-280g grams/week
- Reviewed by General Surgical team, Gastroenterology, Respiratory and feeding team prior to discharge (inpatient eReferrals required).

Parent / Caregiver:

- Confident in feeding their infant (oral and enteral feeding).
- Confident with the basic care of their infant
- Completion of the Gastric Tube Feeding Learning Package if gastric tube feeds are required
- Met with General Surgical CNS (please contact via vocera)
- Education for Safe Infant Sleeping
- Attendance at Infant Resuscitation training
- Rural/regional families – travel advice: adult to travel in back seat with the infant during long car trips. Ensure suitable breaks if journey is a significant distance.
- Provide information on resources:
    - Oesophageal Atresia Research Association (OARA): https://www.oara.org.au/
    - TOF UK (Charity Group): https://tofs.org.uk/
    - Esophageal Atresia Global Support Groups (EAT): https://www.we-are-eat.org/

Related CAHS internal policies, procedures and guidelines

Neonatology Clinical Guideline

- Pre-Operative Care

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Useful resources

http://oara.org.au/

www.tofs.org.uk

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

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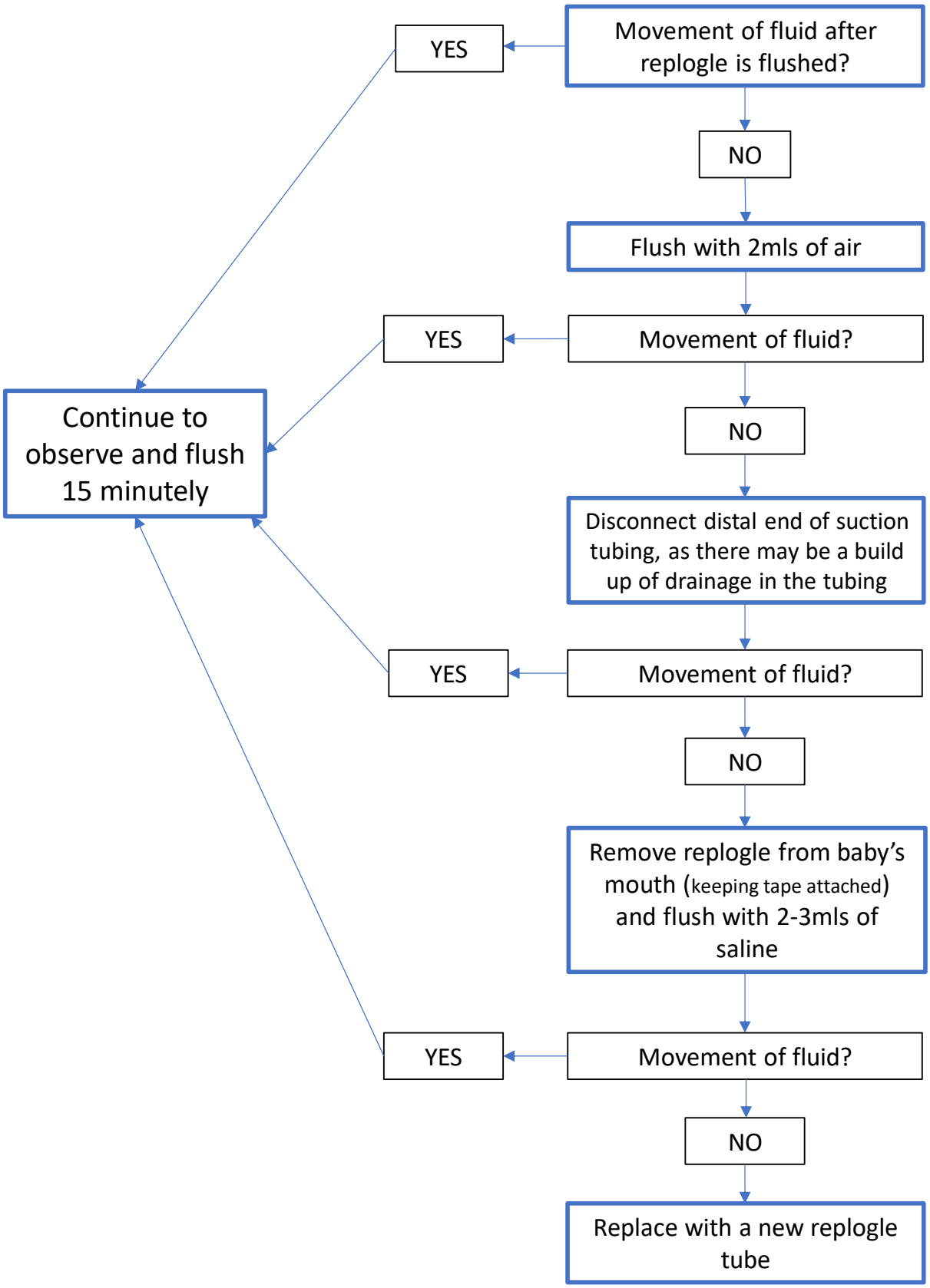
Appendix 1: Troubleshooting a blocked replogle tube



Appendix 2: Sham Feeding Assessment and Procedure

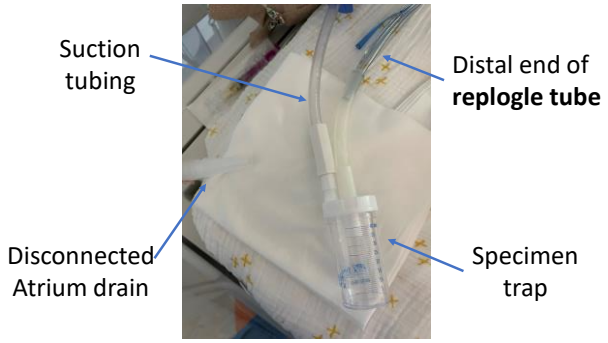
Assessment

Steps	Additional Information
Initial assessment • Ensure cardio-respiratory and oxygen saturations are within normal limits • Ensure patency of the replogle tube • Feeding Team must be present for the first sham feed	• Flush replogle with 3mL of saline and 2 mL of air, ensuring equal volume is removed from the pouch and clear for feeding. • Speech Pathologist to complete oro-motor and cranial nerve assessments, assessment of non-nutritive suck, swallow, breath coordination to determine suitability and progression towards sham feeds. • Occupational Therapist to support state regulation, postural stability and monitor infant stress cues during sham feed trials.
Ongoing assessment • Assessment of cardio-respiratory status during and post sham feeds is essential	Assess for signs of respiratory compromise: • Respiratory distress • Apnoea • Desaturation with Sp02 <90% • Bradycardia • Stridor • Use of accessory respiratory muscles
Assessment of feeding • Co-ordination of sucking, swallowing and breathing • Management of feed volume with no coughing, choking or aspiration episodes	If the neonate has any signs of respiratory distress/compromise or difficulty with the sham feed, then the sham feed should be ceased immediately.

Equipment

- Specimen trap (40mL) – new one required for each feed
- 20mL or 50mL enteral feeding syringe.
- Sham feeding may be by breast or bottle

Procedure

Steps	Additional Information
1. Ensure $Sp0_2$ and ECG/respiratory monitoring	
2. Ensure replogle tube is in correct position and patent.	
3. Flush the replogle tube, ensure the pouch is clear of secretions then disconnect replogle from Atrium UWSD.	Flush replogle with 3mL normal saline followed by 2mL of air. Ensure a good return of secretions and fluid through suction lumen of replogle tube to clear tube and ensure patency.
4. Decrease wall suction to between 40 – 60mmHg	
5. Attach specimen trap to replogle tube and suction tubing. This is to collect the milk feeds. Suction is maintained during the feed	

Process Specific to Breastfeeding

- Midwife/Nursing staff and Speech Pathologist to be present for initial breastfeeding attempts in the first 1-2 weeks of sham feeding. Lactation Consultant to be present if available.
- Breastfeeding may start when suck/swallow/breathe is well coordinated.
- During breastfeed the neonate should be positioned head up with their trunk at least 45 degrees upright.
- Initial breastfeeds are to be on an empty breast, and only offering 1 breast.

Steps	Additional Information
• If breastfeeding well and volumes are tolerated over the first 2 days - the mother can breastfeed without expressing first; increasing as tolerated to offering both breasts at each feed (when neonate is on 150mLs/kg/day bolus gastrostomy feeds). • When either an appropriate volume is taken; the neonate is no longer interested in breastfeeding; or the specimen jar is nearly full, re-feed the breastmilk (from the specimen trap) into the neonate's gastrostomy. This breastmilk needs to be re-fed at the same time as the feed; it cannot be kept later for a feed. • Burp the neonate, as this gets the infant used to burping (which will be necessary post repair of oesophageal atresia). • Once tolerating sham feeds, aim to grade up over several weeks, to 4 sham feeds per day.	
Process specific to bottle feeding • Nursing staff and Speech Pathologist to be present for initial bottle-feeding attempts in the first 1-2 weeks of sham feeding. • Offer a bottle feed only if mother planning to bottle feed with EBM/formula and does not wish to breastfeed. • Initial bottle feed should be 10mL volume or less. • Warm entire volume of feed to be given, pouring amount for sham feed into the bottle & offer sham feed with extra-slow flow teat for first 2 days. Appropriate teat to be determined from Speech Pathologist assessment of suck, swallow, breath coordination. • Increase bottle feed by 5mL every 12 hours, as tolerated, up to full feed volume. • While holding infant in a comfortable upright position-place a few drops of EBM/formula on lips to initiate feed. Pace bottle feed as required by the neonate. • When either appropriate volume is taken; infant is no longer interested in feeding; or the specimen jar is nearly full, re-feed the EBM/formula (from the specimen trap) into the neonate's gastrostomy. This EBM/formula needs to be re-fed at the same time as the feed; it cannot be kept for a later feed. • Burp the neonate, as this gets the infant used to burping (which will be necessary post repair of oesophageal atresia). • Once tolerating sham feeds, aim to grade up over several weeks, to 4 sham feeds per	
6. At completion of the sham feed and re-feeding via gastrostomy tube, disconnect and discard specimen jar. 7. Reconnect the replogle to the Atrium UWSD**.** Flush replogle tube with 3mL normal saline and 2mL of air to ensure any residual milk is removed the pouch. 8. Record Sham feed on observation chart.	

Appendix 3: Follow-Up and Outpatient referrals

Referral Service	Additional Information
General Surgical; Complex Thoracic 'TOF' Clinic	**e-Referral:** 1 week prior to discharge to arrange review in multidisciplinary clinic led by General Surgery. Refer via General Surgery + select drop-down option 'Complex Thoracic Clinic' Please state "Newborn with TOF/OA. Follow-up in Complex Thoracic Clinic at 6 weeks of age."
General Surgical CNS	Contact via **Vocera** 1 week prior to discharge and advise patient for discharge. CNS available as contact for family for phone support until Complex Thoracic clinic review
Hospital in the Home	**e-Referral and phone call handover:** for infants going home with gastric tube feeds.
Gastroenterology	Will review at Complex Thoracic Clinic – no additional e-Referral required
Respiratory	Will review at Complex Thoracic Clinic – no additional e-Referral required
ENT	Will review at Complex Thoracic Clinic – no additional e-Referral required
Feeding Team (Speech Pathology and Occupational Therapy)	Will review at Complex Thoracic Clinic – no additional e-Referral required
Dietitian	**e-Referral:** 1 week prior to discharge for enteral feeding plan if going home with gastric tube feeds.
Neonatal Follow-up Clinic	4 and 8 months for monitoring of growth and development
Referral to local Paediatric services for regional families	