For Use in:	NICU, CAU, Blakeney, Buxton, Paediatric Medicine and Paediatric Surgery Departments, Radiology and Anaesthetic Departments, NNUHFT_
By:	Medical and Nursing Staff
For:	Babies born with Oesophageal Atresia (OA)/ Tracheo-Oesophageal fistula (TOF)
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Clinical Guideline for the Management of Oesophageal Atresia and Tracheo-Oesophageal Fistula in Neonates

This guideline has been approved by the Trust's Clinical Guidelines Assessment Panel as an aid to the diagnosis and management of relevant patients and clinical circumstances. Not every patient or situation fits neatly into a standard guideline scenario and the guideline must be interpreted and applied in practice in the light of prevailing clinical circumstances, the diagnostic and treatment options available and the professional judgement, knowledge and expertise of relevant clinicians. It is advised that the rationale for any departure from relevant guidance should be documented in the patient's case notes. The Trust's guidelines are made publicly available as part of the collective endeavour to continuously improve the quality of healthcare through sharing medical experience and knowledge. The Trust accepts no responsibility for any misunderstanding or misapplication of this document.

Clinical Guideline for the Management of Oesophageal Atresia and Tracheo-Oesophageal Fistula in Neonates

Version and Document Control:

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1.2	17/06/2024	Minor changes made	Khizer Mansoor, Ashok Daya Ram Richard England Ashish Minocha

This is a Controlled Document

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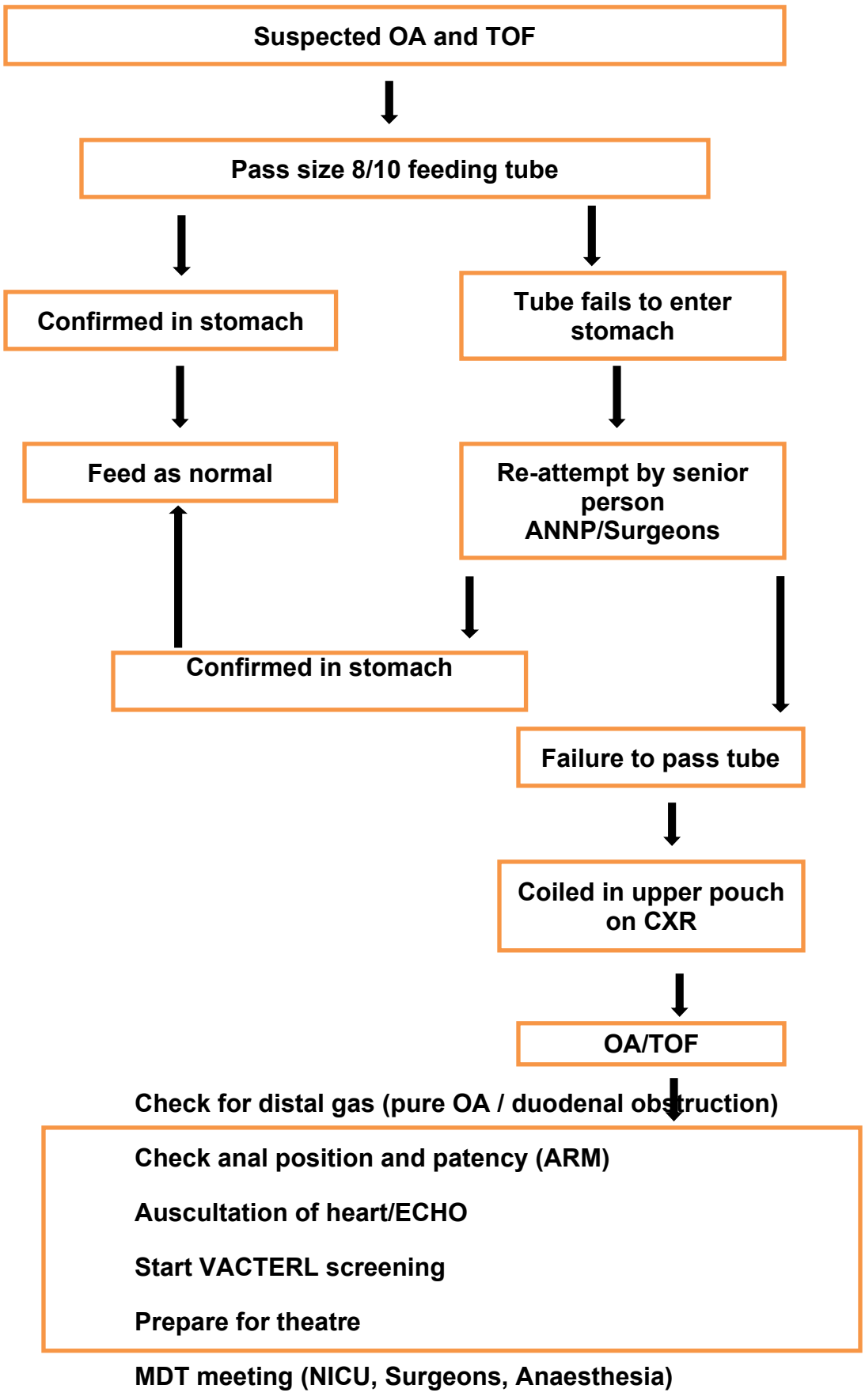

Suspected OA and TOF
Pass size 8/10 feeding tube
Confirmed in stomach
Tube fails to enter stomach
Feed as normal
Re-attempt by senior person ANNP/Surgeons
Confirmed in stomach
Failure to pass tube
Coiled in upper pouch on CXR
OA/TOF
Check for distal gas (pure OA / duodenal obstruction)
Check anal position and patency (ARM)
Auscultation of heart/ECHO
Start VACTERL screening
Prepare for theatre
MDT meeting (NICU, Surgeons, Anaesthesia)

Clinical Guideline for the Management of Oesophageal Atresia and Tracheo-Oesophageal Fistula in Neonates

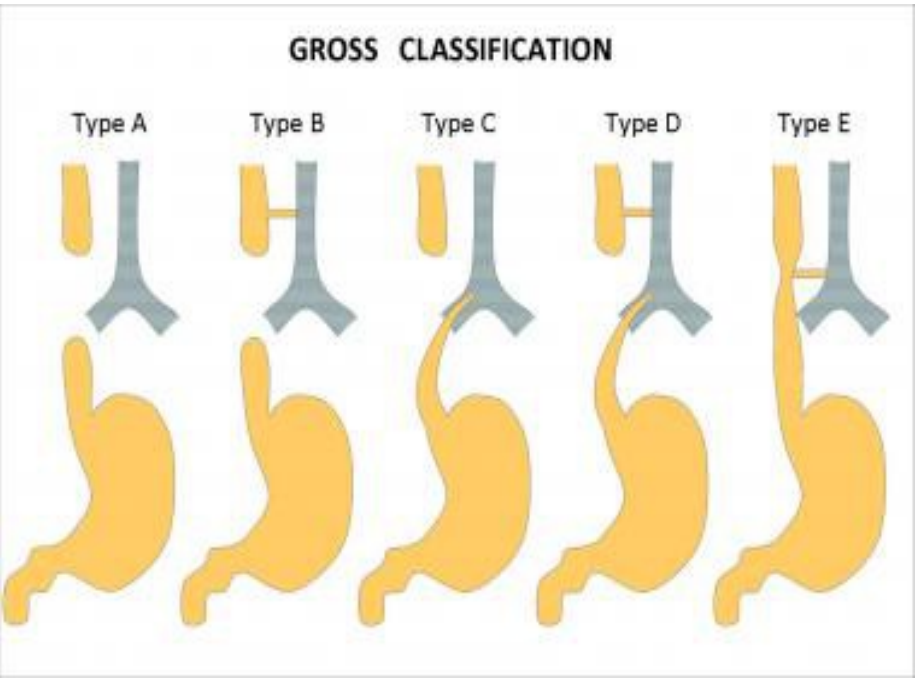
Background

Oesophageal atresia (OA) is a congenital abnormality in which there is a blind ending oesophagus. Its incidence is approximately 1:2500 live births. There is no sex preponderance. It can occur in isolation or there may be one or more fistulae communicating between the abnormal oesophagus and the trachea, known as a tracheo-oesophageal fistula (TOF). The exact aetiology is unknown.

Types and Classification

There are five types described as per Gross classification as below.

Anatomic Characteristics	Frequency
Type A: OA with no fistula	8.0%
Type B: OA with proximal fistula	0.8%
Type C: OA with distal fistula	89%
Type D: OA with proximal and distal fistula	1.2%
Type E: TOF with no atresia	1%



Objectives

The objective of this guideline is to ensure best practice in the diagnosis, investigation and management of a baby born with oesophageal atresia and trachea oesophageal fistula: in the perinatal period and up to the time of discharge.

Rationale

This guideline has been written to ensure that all babies born with the condition are diagnosed and managed promptly and appropriately. It also aims to help medical and nursing staff explain the management clearly to the parents. These are derived from standard practice among paediatric surgeons and published literature.

Definitions

OA:	Oesophageal atresia is a condition which occurs when the upper part of the oesophagus does not connect with the lower oesophagus and stomach.
TOF:	Tracheo-oesophageal fistula is an abnormal connection between the upper part of the oesophagus and the trachea.
Replogle Tube:	This is a double-lumen tube which is inserted through the baby's nostril or mouth into the blind-ending oesophageal pouch and used to drain saliva to keep the upper pouch empty and thus prevent aspiration (Appendix 1).
Tracheomalacia:	Is the floppiness of the windpipe which occurs when the cartilage in the trachea has not developed properly. It may be part of the OA/TOF disease process.
GOR:	Gastro-oesophageal Reflux is effortless passage of stomach secretions or milk from the stomach up the oesophagus. Sometimes this presents as vomiting or an uncomfortable sensation of heartburn. It is common in babies but can be worse in oesophageal atresia.
TAT:	Trans-Anastomotic-Tube. This is a Nasogastric feeding tube which passes through the oesophageal anastomosis to allow early feeding. Repassing this in the early postoperative stages can cause damage to the anastomosis.
VACTERL:	This is the association of Vertebral, Anorectal, Cardiac, Tracheo-Esophageal (American spelling), Renal and Limb anomalies. This can all occur in OA and need to be searched for with a careful screening process (Appendix 2).

Antenatal diagnosis

- OA/TOF cannot be diagnosed with certainty in the antenatal period.
- The combination of polyhydramnios and a small or undetectable stomach on a prenatal ultrasound scan is suggestive for OA. Sometimes a dilated upper oesophageal pouch might be visible.
- Associated abnormalities may be seen on ultrasound imaging, such as the presence of cardiac defects.
- The fetus is usually small for gestational age.
- In highly selected cases fetal MRI may be used for further evaluation to aid appropriate counselling.

Antenatal counselling with a paediatric surgeon should be arranged in all cases suspected of OA after their fetal medicine appointment. Information on neonatal care can also be provided by the neonatologists. In complex cases of multiple abnormalities and likely syndromic cases it may be appropriate to arrange a joint MDT meeting to discuss the case.

Postnatal diagnosis

Oesophageal atresia

- Inability to feed.
- Coughing / Choking.
- Excessive frothing at mouth.
- Inability to pass a nasogastric tube.
- Cyanotic episodes.
- Signs of aspiration.

H-type fistulae

- Usually detected later in infancy or childhood.
- Able to feed.
- Unexplained respiratory symptoms.
- Coughing with feeds.
- Recurrent chest infections.
- Excessive abdominal gaseous distension.

Delivery and postnatal care

If antenatally suspected, delivery should be planned at a centre with NICU and neonatal surgery facilities. Mode of delivery is dictated by obstetric indications only. A Paediatric alert in cases of antenatal suspicion should prompt attendance of the NICU team at delivery and the baby be brought to a neonatal cot where a nasogastric tube can be passed. Easy passage of a tube and aspiration of acid is reassuring such that in the absence of other obvious abnormalities and further concerns the baby can be returned to the mother and taken to the postnatal ward where feeding should be observed closely.

If acid cannot be aspirated from the NGT then the baby should not be fed but taken to NICU for further evaluation. The NGT should not be removed and a chest x-ray performed to confirm if it is in the stomach via a normal oesophagus before the diagnosis is ruled out. If this rules out oesophageal atresia then the baby can be returned to the postnatal ward and fed.

Polyhydramnios should alert the neonatal team to other causes of impaired fetal swallowing such as syndromic or dysmorphic features.

If the CXR demonstrates a curled up NGT in the upper oesophagus this should be replaced with an 8 or 10 Fr Replogle tube (Appendix 1). Slight pressure should be applied on the tube and the CXR retaken.

If there is any doubt about the diagnosis then the baby should stay on NICU for further assessment. It should be noted that rarely a NGT can find its way into the stomach via the trachea and fistula in the presence of oesophageal atresia. If the passage of the tube appears abnormal it is wise to obtain a lateral view of the neck and thorax with the tube in place.

If a baby is referred from another centre for suspected TOF/OA, it should be discussed between the surgical and neonatal teams before accepting transfer. If a baby weighs less than 1.5kg and/or the baby has major cardiac abnormalities, the baby should be discussed with Great Ormond Street Hospital for further management. If the baby is born in the unit, an MDT meeting with the anaesthetic team must be arranged ASAP.

Clinical examination

A careful clinical examination of the baby should be performed noting any dysmorphism and likely syndromic conditions. A cardiovascular examination is performed and the patency of the anus is checked.

Investigations post diagnosis

CXR/AXR:

A combined CXR and AXR must be performed. An isolated CXR is inadequate. The CXR may show the NG tube coiled in the upper pouch. This should be repeated with a Replogle tube in situ to give an assessment of gap. It can also demonstrate the heart

size, any vertebral and rib abnormalities and lung fields. If gas is present in the stomach and bowel a TOF must exist whereas if the abdomen is gasless the diagnosis is likely to be pure oesophageal atresia. A double bubble shadow suggests a co-existing duodenal atresia. Assessment of vertebral bodies from thoracic to sacral spine should be requested.

Echocardiography:

Echocardiography should be performed ideally prior to surgery in all cases to rule out a right-sided arch (2.5% cases) as well as other major associated cardiac abnormalities. Babies who have congenital cardiac conditions may need to be transferred to another centre for surgery.

VACTERL Screening:

Please refer to the VACTERL investigation and referral proforma (Appendix 3). This should be started pre-operatively and completed in all babies once surgical repair is performed and the baby is stable enough to be transported to the radiology department. The complete screening process must be completed prior to discharge.

Preparation of a neonate for surgery

A 10F Replogle tube is placed nasally or orally well into the upper pouch. It is connected to continuous low flow suction (25 mm Hg, 3.5-4 Kpa, 35-42cms water). It is also flushed intermittently with 0.5 mLs of 0.9% sodium chloride every 15 minutes to ensure the patency of the tube. See Appendix 1.

- Continuous oxygen saturation and cardiac monitoring.
- IV fluids as per protocol.
- Broad-spectrum intravenous antibiotics as per local guidelines.
- Nurse supine with the head elevated by approximately 45°.
- All babies should have a long line placed as soon as is practical and PN initiated until full enteral feeds established postoperatively.
- Pre-operative investigations should also include FBC, U&E, Clotting and X-Match 1 unit.
- After discussing with the neonatologist and the geneticist, mum may be counselled for saving the baby's blood for genetic testing and sent for microarray.

The parents must be briefed about the nature of the congenital anomaly explaining the pathologic anatomy, intended repair and the possible complications after which consent is obtained for the operation.

If the child is less than 1.5 kg and/or has major cardiac abnormality, multi-disciplinary team meeting should determine the need to transfer to other centres eg. GOSH.

Pre-operatively, an MDT meeting between surgical, anaesthetic and neonatal teams is arranged and all pre-operative investigations reviewed.

Surgical repair:

Open right sided thoracotomy, closure of the fistula and oesophageal anastomosis is the standard method. This is usually performed in the extrapleural space.
Thoracoscopic approaches are also available in some other centres.

In OA without fistula, an assessment of gap and formation of gastrostomy will be required. A decision on surgical strategy must be made regarding whether a delayed primary repair can be achieved and the length of time to wait for the gap to reduce. If one of the numerous oesophageal lengthening procedures or oesophageal substitution is more appropriate then discussion with a high volume oesophageal centre may be appropriate.

Anaesthesia and intubation:

- **Emergency intubation:**
  Intubation and ventilation should be avoided if at all possible due to the risks of gastric distension via a TOF.
  However, if this is unavoidable then the consultant surgeon should be informed and consideration given to informing the paediatric anaesthetist on call.
  Obvious airway abnormalities should prompt an early assessment by the paediatric ENT team.

- **Elective intubation and anaesthesia**
  There are a variety of techniques for the management of babies undergoing thoracotomy in the presence of a TOF and these will be determined by the preferences of the paediatric anaesthetist involved.

  In some babies there may be an indication for pre-operative bronchoscopy and the surgeon should discuss the need for this with the anaesthetist at the pre-operative MDT.

- **Emergency ligation of fistula**
  Babies who have been intubated and ventilated are at risk of developing severe gastric distention due to preferential ventilation of the TOF. Gross distension of the abdomen may then impede ventilation creating a ‘can’t ventilate situation’. Urgent gastric needle decompression may relieve this pressure effect at the expense of further preferential gastric ventilation via an open needle. This needs to be occluded intermittently.

The definitive resolution for this situation is ligation of the fistula.

- If the baby is peri-arrest this should be done at the cot-side.
- Inform paediatric anaesthetist.
- Inform Emergency theatre team.
- Arrange good lighting.
- Cut down set.

- Obtain Bipolar Diathermy and suction.

Once control of the fistula is obtained, transfer the baby immediately to theatre.

Post-operative care:

- The post-operative note will include particular postoperative care instructions for the baby as these may differ case to case depending on the gap length and security of the anastomosis. These instructions should be handed over to the NICU team.
- Most babies will require a period of postoperative ventilation and so will be transported back to NICU ventilated.
- Endotracheal tube suctioning should be mindful not to vigorously suction distal to the end of the ETT tube length as a fistula pit could be entered.
- **Premature extubation must be avoided** as subsequent intubation in the setting of a freshly closed tracheal fistula invites reopening of the fistula and neck hyperextension will place tension on the anastomosis.
- The chest draining tube if in place is placed in 2 cm of water only to seal it; it is not connected to a suction device.
- Watch for saliva exiting out the chest drain; this is a signal of anastomotic leakage with or without signs of sepsis.
- If a chest drain is not placed intra-operatively but there appears to be a pneumothorax on post-operative CXR then please discuss with the surgical team before placing a chest drain unless there is clinically a tension pneumothorax.
- Antibiotics are continued for at least 72 hours or as per post-operative instructions
- TAT tube feeding can be initiated as per surgeon's decision. The timing of feed initiation and rate of increase depends on the security (tightness) of the surgical repair.
- **If the TAT is removed accidentally, surgical team must be informed and no attempt to replace it should be made.**
- All babies have a high risk of gastro oesophageal reflux and thus anti reflux medication is prescribed till the child is at least 6 months of age.

Complications:

Early

- **Anastomotic leak**:

- Tends to occur 3-4 days after surgery.
- Has been reported in approximately 15% of cases.
- Pain and distress are often evident.

- Signs of sepsis may be present.
- A chest tube drains saliva.
- Treatment is supportive, waiting allows the leak to close.
- Appropriate IV antibiotics.
- Nil By mouth / Continue PN.
- An oesophagogram may be performed to assess the leak.

➢ **Anastomotic stricture**:

- Has been reported in as many as 50% of cases.
- Essentially, 100% of babies have a waist at the anastomotic site, but this may not be functionally significant.
- Contrast Swallow may be performed to assess the stricture.
- Oesophageal dilation is best and is most safely performed by means of a balloon technique under fluoroscopic control.
- Repeat dilations are often necessary.
- Anti-reflux medication should be maximised because acid reflux can be both an aggravating and a causative factor in stricture formation.

➢ **Chylothorax**

- Is an uncommon complication due to mediastinal/ thoracic dissection.
- Presents as persistent fluid drainage if a chest drain present.
- Collapse of lung and pleural effusion on x-ray.
- Management by chest drainage.
- Medium chain triglyceride (Monogen) enteral feeding.
- TPN may be required in persistent cases.

Late

- Gastroesophageal reflux.
- Oesophageal dysmotility.
- Tracheomalacia.

Before discharge

- VACTERL screening should be completed.
- Parents should have had opportunity to discuss and learn how to feed their child safely from a specialist paediatric Speech and Language Therapist.
- Parents must be informed of the nature of their child's condition, any associated

anomalies and potential complications.

- They should have received a copy of the patient information leaflet (Taking home our baby after a TOF repair, Trustdocs ID: 13980).
- Parents should be trained in CPR for babies and infants.
- They should have open access to CAU or the equivalent at their local hospital.
- TTO medication should include a PPI or H_2 receptor antagonist.

Follow Up:

- The GP should receive the Electronic Discharge Letter to ensure that he or she is briefed on the baby's history, condition, and expected outcome.
- The surgeon should follow up in one month to review progress with the parents and generally assess the child's condition, growth, swallowing and wound healing.
- The patient should return at 3 months for a similar assessment.
- At a 1-year follow-up and general assessment, swallowing function, respiratory issues, and other factors should be addressed.
- Radiologic assessment of the oesophagus is required only if a significant history of choking, cyanosis, regurgitation, dysphagia, growth failure, coughing, or wheezing is noted with a possible subsequent endoscopic evaluation.
- Patients with dysmorphism or more than one abnormality on VACTERL screening should be referred for clinical genetics review. Positive results on Microarray should be discussed with a clinical geneticist to assess relevance before relaying result to parents. Further counselling should be offered if required.
- Neonatology follow up is arranged for all pre term babies and those with multiple anomalies.
- If they do not live locally they should be referred to local Paediatrician for follow-up in addition to surgical follow-up here in Norwich.

Clinical audit standards / audit standards

To ensure that this document is compliant with the above standards, the following monitoring processes will be undertaken:

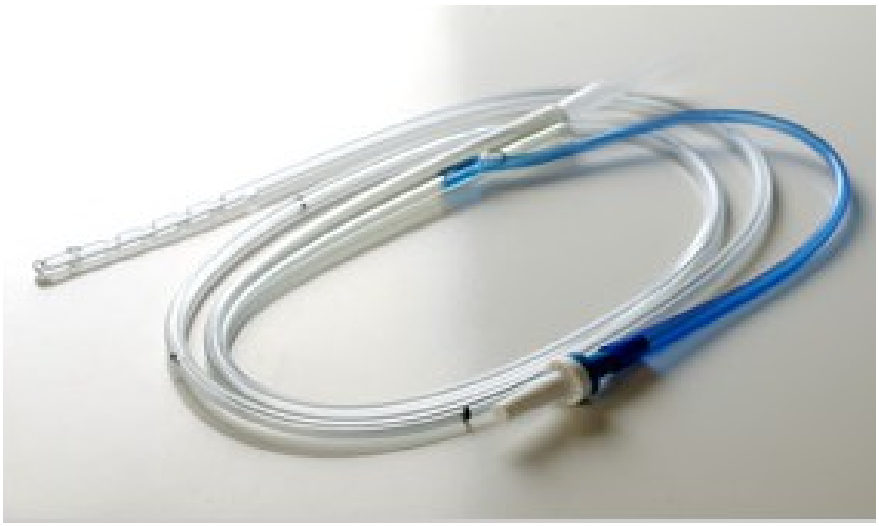
1. All babies with oesophageal atresia should be diagnosed and transferred to a tertiary centre with paediatric surgical services within 24 hours.
2. Upper pouch must be kept clear using the Replogle tube to avoid aspiration.
3. The surgical team must complete and document the examination of the baby in the case notes.

4. Use of VACTERL screening proforma and documentation of all the findings completed before discharge.
5. Pre-operative echocardiogram within 24 hours to exclude major congential cardiac anomalies precluding surgical repair at NNUH.
6. Documentation of the discussion with the parents in the relevant communication section of the notes.

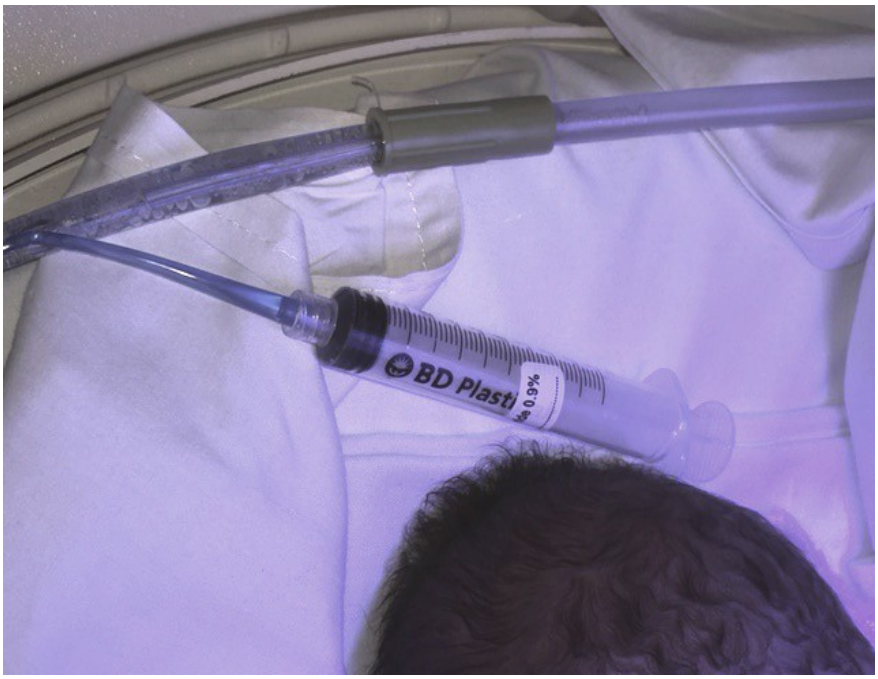
The audit will be performed every 3 years and the results will be sent to the *Paediatric surgery team / Clinical Director* who will ensure that these are discussed at relevant governance meetings to review the results and make recommendations for further action.

Appendices

Appendix 1: Picture of Replogle tube.



The tube has multiple holes at the end and a double lumen to allow flushes via the blue inner tubing.



Saliva can be seen in the large tube connected to the suction. A 0.5ml saline flush can be given with the syringe connected to the blue tubing every 15 mins.

Clinical Guideline for the Management of Oesophageal Atresia and Tracheo-Oesophageal Fistula in Neonates

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Appendix 2: VACTERL Screening Proforma.

Adapted specifically for oesophageal atresia patients. (next page)

Patient Identifier Label

VACTERL Component		Date *(ddmmyyyy)*: Requested	Date *(ddmmyyyy)*: Performed	Findings: Please also document in notes	Referral if abnormalities: *For all dates use dd/mm/yyyy*
Vertebral	**V** **Spinal X-Ray:** All AP Thoracic, lumbar, Sacral Consider sacral lateral/ AP cervical				To Paediatric Spinal Surgeons Referred to: …… Referred by: …… Date of referral: …… Date of Review: ……
	All spinal cord ultrasound				To Paediatric Neurosurgeon (including conus lower than L1/L2) Referred to: …… Referred by: …….. Date of referral: …… Date of review: ……..
Anorectal	**A** **Anal examination**	**Date performed** *(ddmmyyyy)*:		**Inspection of anus:** Consider measuring Sacral Ratio in ARM:	
Cardiac	**C** **All have pre-operative Echocardiogram**			**Heart sounds:** **Pulses:** **Echo:**	**Discharged:** Yes ☐ No ☐ If **no** follow- up arrangements:
Tracheo-Oseophageal	**T** **E**			**Feeding:** Yes ☐ No ☐ NG tube in stomach: Yes ☐ No ☐ Double bubble noted: Yes ☐ No ☐	
Renal	**R** **All renal Ultrasound**				To Paediatric Urology team Date of referral: …… Referred to: …… Referred by: ……..
Limbs	**L** **All limb examination**	**Inspection of limbs:**			To plastics (hands) or orthopaedics Referred to: …… Referred by: …….. Date: ……
Other	**Consider Genetics Referral** Dysmorphism or Multiple abnormalities	**Indication:** **Microarray to be sent in all OA patients.** **Date** *(ddmmyyyy)*:			Referred to: …… Referred by: …….. Date: …… Appointment: ……

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List of defects often identified in VACTERL patients[1]

Vertebral Tethered cord Butterfly vertebra Vertebral fusion Hemivertebra Additional lumbar vertebra Additional or absent ribs	**Cardiac** Ventricular septal defect Atrial septal defect ASD and VSD Tetralogy of Fallot Dextrocardia Coarctation Double arch	**Urinary tract Anomaly** Reflux Horseshoe kidney Hypospadias Solitary kidney (agenesis) PUJ obstruction Cryptorchidism Dysplastic kidney
GI Atresia Imperforate anus Duodenal atresia **Other** Cleft palate / lip Abnormal chromosomes	**Oesophageal atresia** With distal fistula H type Pure atresia Double fistulas Proximal fistula	**Limb anomalies** Absent radius Digital anomalies Hip dysplasia

Consider overlapping syndromes[2]

Condition	Features in common with VACTERL	Features distinct from VACTERL
Alagille syndrome	Vertebral anomalies, cardiac anomalies; may have renal anomalies	Bile duct paucity and cholestasis, ophthalmologic anomalies (especially posterior embryotoxon), neurological anomalies, characteristic facial appearance
Baller-Gerold syndrome	Radial anomalies, may also include anal anomalies	Craniosynostosis, skin anomalies
CHARGE syndrome	Cardiac malformations, genitourinary anomalies; may also include TEF	Colobomata, choanal atresia, neurocognitive and growth impairment, ear anomalies, cranial nerve dysfunction, characteristic facial features
Currarino syndrome	Sacral malformations, ARM	Presacral mass
22q11.2 deletion syndrome (also known by other names, such as DiGeorge syndrome or velocardiofacial syndrome)	Cardiac malformations, renal anomalies, other VACTERL-type anomalies also reported	Hypocalcemia, palatal anomalies, learning difficulties, immune dysfunction, neuropsychiatric disturbances, characteristic facial features,
Fanconi anaemia	Virtually all features of VACTERL association may occur; radial anomalies are considered an especially key feature	Hematologic anomalies, pigmentation anomalies
Feingold syndrome	GI atresia, cardiac defects, renal anomalies	Brachymesophalangy, toe syndactyly, microcephaly, cognitive impairment, characteristic facial appearance,
Fryns syndrome	GI malformations, cardiac defects, GU anomalies	Diaphragmatic defects, neurocognitive impairment, characteristic facial appearance
Holt-Oram syndrome	Cardiac malformations, limb malformations	Cardiac conduction disease (also reported in VACTERL association)
Müllerian duct aplasia, renal aplasia, and cervico-thoracic somite dysplasia (MURCS association); also known as Mayer-Rokitansky- Küster-Hauser syndrome type II	Vertebral anomalies, renal anomalies, GU anomalies and anorectal malformations; may also have cardiac and limb anomalies	Syndactyly and hearing loss have been described
Oculo-auriculo-vertebral syndrome	Vertebral anomalies, cardiac abnormalities, limb abnormalities, urogenital anomalies	Ear anomalies (microtia), hemifacial microsomia, neurocognitive impairment, facial clefts (also described in patients with VACTERL association)
Opitz G/BBB syndrome	Anal anomalies, heart defects,TEF, hypospadias	Hypertelorism, syndactyly
Pallister-Hall syndrome	Imperforate anus, renal anomalies, limb anomalies (postaxial polydactyly should serve as a clue for the Pallister-Hall syndrome)	Hypothalamic hamartoma, bifid epiglottis (ranging to more severe types of clefts), nail hypoplasia
Townes-Brocks syndrome	Imperforate anus, thumb anomalies, renal anomalies, cardiac anomalies	Dysplastic ears, hearing loss

References for Appendix 2 - 1 Keckler SJ, St Peter SD, Palusek PA, Tsao K, et al. VACTERL anomalies in patients with esophageal atresia: an updated delineation of the spectrum and review of the literature. Pediatr Surg Int 2007;23:309-313 2Solomen BD. VACTERL/VATER

Association. Orphanet Journal of Rare Diseases 2011, 6:56

Useful Links

www.tofs.org.uk

www.gosh.nhs.uk/medical-information/oesophageal-atresia-tracheo-oesophageal-fistula

www.nhs.uk/conditions/oesophageal-atresia/