Surgical Conditions Affecting Neonates in Scotland Managed Clinical Network

POSTNATAL MANAGEMENT GUIDELINES FOR BABIES WITH SUSPECTED OESOPHAGEAL ATRESIA WITH OR WITHOUT TRACHEO-OESOPHAGEAL FISTULA

NOTE

This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined based on all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

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Management after birth of babies with suspected Oesophageal atresia with or without Tracheo-oesophageal Fistula

These guidelines have been developed by the Steering Group of the Surgical Conditions Affecting Neonates in Scotland (SCANS) Managed Clinical Network. These recommendations are applicable to all healthcare professionals caring for babies with Oesophageal Atresia (OA) with or without Tracheo-oesophageal Fistula (TOF).

The guidelines are underpinned by the following key principles:

1. Babies with OA TOF should be managed on a surgical neonatal intensive care unit (NICU) with joint care provided by neonatologists and paediatric surgeons (1). For the purposes of these guidelines, the surgical NICUs are located in Glasgow (Royal Hospital for Children), Edinburgh (Simpson's Centre for Reproductive Health) and Aberdeen (Royal Aberdeen Maternity Hospital)
2. Network pathways and processes should be in place to ensure that;
    1. Where there is an antenatal suspicion of OA TOF, the parent(s) receive multi-disciplinary counselling involving fetal medicine, neonatology and paediatric surgery, ideally as a joint session
    2. Where there is an antenatal suspicion of OA TOF, delivery is planned in the surgical NICU to negate the need for postnatal transfer and the potential separation of mother and baby
    3. Where the diagnosis is confirmed postnatally or the baby is born in a non-surgical centre, referral to the surgical NICU is performed in a timely manner, ideally within 2 hours of birth or sooner. Referral should be via the **ScotSTAR emergency line 03333 990 222**, which will initiate a conference call
    4. Repatriation from the surgical NICU to the local hospital is facilitated as soon as possible when clinically appropriate (2).
3. Throughout this document we use the term parent to mean all parents, carers and legal guardians, and the term mother to mean all women and people who have given birth.

Contacts details for the three surgical NICUs can be found in Appendix 1 at the end of this guideline

OA TOF: Background

OA is a congenital abnormality where the oesophagus ends in a blind upper pouch. It can occur in isolation or there may be one or more fistulae between the abnormal oesophagus and the trachea (tracheo-oesophageal fistula). OA TOF is rarely diagnosed definitively before birth, but it may be suspected due to maternal polyhydramnios and a small/absent stomach bubble.

Babies born with OA TOF need to have intensive neonatal care prior to corrective surgery, normally within days of birth.

Postnatal presentation includes:

- Coughing/choking/colour change during feeds

- Respiratory distress
- Frothing/ dribbling of saliva
- Inability to pass a nasogastric tube (NGT) into the stomach
    - May later be confirmed on CXR as coiled (classically) or short

Delivery room (DR) management in suspected OA TOF

Routine delay of umbilical cord clamping (DCC) for at least 60 seconds is recommended in babies who are not compromised at the time of birth[2-4]. Suspected OA is not considered a contra-indication to DCC.

- Neonatal staff with appropriate training and competencies should attend the delivery
- Stabilise the baby in accordance with NLS guidance
    - However, care should be taken where possible to avoid positive pressure ventilation due to the risk of oesophageal pouch distension, gastric distension (if there is a fistula) and rarely gastric rupture.
    - If the baby needs ongoing respiratory support, use the lowest effective pressures possible
    - If the baby is preterm, a trial of heated, humidified, high flow nasal cannula therapy may be preferable to conventional management with CPAP
- There may be copious oral secretions – regular suction is likely to be required in the BR
- Apply routine BR monitoring (oxygen saturations on the right hand, heart rate and temperature)
- Transfer the baby to NICU for ongoing care when stable

Ongoing neonatal unit management

1. The infant should be nil enterally and will receive all nutrition parenterally.

2. Monitoring and vascular access

- NICU monitoring
- Establish peripheral venous access. Once the diagnosis is confirmed, thought should be given to insertion of a percutaneous long line (PICC line).
- If an NGT has proven difficult to pass, a CXR should be performed to demonstrate coiling in upper oesophagus
- If OA-ToF is suspected for any other reason, a wide-bore NGT should be passed and a CXR performed
- Once diagnosis confirmed, site a replogle tube – see WoS guideline
- The replogle should be pushed on by an appropriate member of staff and a CXR performed to confirm diagnosis, this should be documented
- Echocardiography should be performed prior to surgery

3. Analgesia and sedation

Monitoring of pain/sedation using an appropriate assessment tool should be part of routine care.

If analgesia/sedation is required this should be administered according to local guidance.

4. Refer/discuss with specialist centre

Once stabilised babies born out with one of the three regional surgical NICU should be discussed with the nearest specialist team as soon as possible after birth. A conference call involving ScotSTAR Neonatal Transport Service should be initiated to facilitate timely transfer.

5. Fluid balance

Intravenous fluid management should be guided by local practice, as should correcting any associated electrolyte imbalances.

6. Antibiotics

Antibiotics are not routinely required unless there are other risk factors for, or clinical signs of sepsis.

7. Enteral feeding

Breast milk is recommended after repair as there are lifelong benefits, and mother should receive lactation support to enable them to establish their milk supply whilst baby is nil by mouth. Early involvement of a specialist neonatal dietician is advised if there are issues with weight gain and growth.

8. Surgical review

For antenatally suspected OA TOF, surgical review should occur following admission to the NICU. Otherwise, surgical review should occur at the earliest opportunity following suspected diagnosis of OA TOF. It is anticipated that pending surgical review, all infants will be fasted, have a large bore nasogastric or Replogle tube in situ, and have x-ray imaging performed. The surgical team should assess the following:

- The length of the oesophageal pouch, this is done by performing a CXR while pushing on the replogle tube
- The presence (or absence) of gas in the stomach; gas in the stomach is indicative of a TOF
- The bowel gas pattern for evidence of other intestinal anomaly (e.g. duodenal atresia, anorectal malformation)
- The presence of any associated vertebral or rib anomalies
- The presence of absence of an anus, indicative of anorectal malformation

9. Specialist review / investigations

A clinical examination should be performed looking specifically for dysmorphism and associated anomalies, including cardiac, anorectal and limb. All babies should have their perineum examined as soon as possible, and the presence or absence of an anus confirmed. If necessary, a soft narrow bore nasogastric tube can be passed into the anus to confirm patency. In addition, the following investigations are required;

- Echocardiogram – this must be performed prior to surgery to determine the position of the aortic arch which will inform the surgical approach, as well as to exclude major cardiac anomalies
- Renal USS – this is not essential prior to surgery unless the baby has not passed urine or has biochemical renal impairment, in which an urgent scan should be considered prior to theatre
- Spinal ultrasound - can be done routinely after surgery
- Genetic investigations should be considered based on clinical findings and associated anomalies

Surgical management

1. Pre-operative work up

The Surgical management of these patients is complex and requires appropriate preparation. The following principles should apply:

- Informed consent should be taken by the surgeon performing the procedure in a quiet and comfortable area with support from nursing and neonatal staff.
- Patients should have appropriate venous access (umbilical venous or PICC)
- Patients should have appropriate arterial access before being transported to theatre (umbilical arterial line or peripheral arterial line)
- Patients should have documented passage of urine or a renal USS prior to theatre
- Patients should have documentation of any major cardiac abnormalities and the position of the aortic arch and the operating surgeon should be aware of this
- Samples should be sent for group and save according to local guidelines.
- The position of the anus should have been documented and the operating surgeon aware

2. Surgery

Surgical repair of oesophageal atresia has developed rapidly since it was first performed but remains a complex and challenging procedure. Where possible, the following key principles should be adhered to:

- Surgical management should be undertaken in normal working hours if possible as this ensures that the most appropriate staff are on site to manage what can be a complex patient
- Surgical units should consider having an oesophageal rota of specialist surgeons
- Anaesthetic care should be provided by paediatric anaesthetists with experience managing complex surgery in neonates, including thoracic and

thoracoscopic surgery who are comfortable managing complex thoracic surgery
- Anaesthetic and surgical teams should attempt to ensure that procedures happen promptly and are not routinely delayed by other cases
- The ability to perform a microlaryngobronchoscopy should be routinely available
- Both open and minimally invasive repair are considered acceptable forms of management for oesophageal atresia with fistula
- Minimally invasive repair should be performed by appropriately trained surgeons
- In patients with a long gap (type A) it is recognised that preserving the oesophagus is essential where possible, and these patients should have consideration of a delayed primary anastomosis. Consideration should be given to transferring these patients if that is not available in a unit when the patient is born
- Routine placement of a trans-anastomotic tube or a chest drain is dependent on surgical choice

3. Post-operative care

All patients should be managed in the neonatal intensive care unit after surgery. Most will remain intubated and ventilated. The following key principles apply:

- If the anastomosis was performed under tension, then consideration should be given to nursing principles that will minimise disruption (minimal movement of patient; chin to chest nursing etc)
- When feasible, sedation and ventilation should be weaned as soon as possible
- If a trans-anastomotic tube has been placed, it should be clearly labelled and secured in such a way that prevents inadvertent removal
- Chest drainage should be monitored and antibiotic cover should be provided as long as a drain remains in situ
- Enteral feeding can be commenced at the discretion of the surgical team
- Routine use of oesphageal contrast to assess leak is not required. If a leak is considered, then it is mandatory to document this using contrast.

4. Outlook

In Scotland:

- Median time to full feeds is 12 days
- Median hospital stay is 23 days
- Survival to discharge is 95%
- Survival to 2 years is 95%
- At 2 years, 98% do not need any supplemental airway support
- At 2 years 79% do not require any nutritional support
- 58% will have required another surgical procedure in the first two years, most commonly dilatation of stricture

References

1. BAPM Service and Quality standards for provision of Neonatal Care in the UK, November 2022
2. Intrapartum Care for healthy women and babies. NICE guideline CG190, updated 2022, recommendation 1.14.14
3. RCOG Scientific Impact Paper 14, Clamping of the Umbilical Cord and Placental Transfusion, February 2015
4. Newborn Resuscitation and support of transition of infants at birth Guidelines, Resuscitation Council UK, 2021
5. Esophageal and trachea-oesophageal fistula; van der Zee et al; Seminars in Pediatric Surgery; 2017; 26: 67-71
6. MCN for Neonatology WoS Neonatal Guideline: Oesophageal Atresia and Tracheo-Oesophageal Fistula; February 2019
7. Contemporary management and outcomes for infants born with oesophageal atresia; Burge et al; BJS 2013; 100: 515-521

Appendix 1: Contact details for 3 Surgical Units in Scotland

- **Glasgow**

Royal Hospital for Children: Receiving consultant neonatologist 0141 452 2114 and / or on call consultant paediatric surgeon via switchboard 0141 201 0000

- **Edinburgh**

On call paediatric surgical registrar or consultant paediatric surgeon via switchboard 0131 536 0000 and on call consultant neonatologist via same number

- **Aberdeen**

On call consultant neonatologist and on call consultant paediatric surgeon via switchboard 0845 456 6000. Alternatively contact the neonatal unit directly on 01224 552602.

Appendix 2: Transport Recommendations

Transfer of the infant with Oesophageal Atresia +/- Tracheo-oesageal Fistula
Infants with Oesophageal atresia +/- Tracheo-oesophageal fistula born in a non-surgical centre will require an emergency transfer to a surgical unit for assessment and on-going management. These patients are at a high risk of deterioration and must be managed with caution.

Referral process
Infants presenting with Oesophageal atresia +/- Tracheo-oesophageal fistula require transfer to the neonatal surgical regional centres in either Aberdeen, Edinburgh or Glasgow.

- Every effort should be made to ensure that these infants deliver in the appropriate surgical centre. Where that is not possible, planning should begin on admission to their local centre to allow prompt transfer.
- Referral is made by calling the **ScotSTAR emergency number 03333 990 222** and the relevant surgical and neonatal teams should be brought into the call. If the baby has been delivered, dispatch of the transport team should not be delayed by difficulties in bringing others onto the conference call. This call should be made before birth wherever possible, to allow confirmation of cot destination and immediate stabilisation plans to be made without delaying transfer.
- If the baby is requiring invasive, positive-pressure ventilation, response to the referral should be an 'immediate dispatch' – team mobilising within 60 minutes of referral.
- For non-ventilated babies, the case should be triaged by the on-call neonatal transport consultant and the team dispatched accordingly.

Respiratory Status
Where possible, avoid positive pressure ventilation, this is due to the risk of oesophageal pouch distension, gastric distension (if there is a fistula), splinting of the diaphragam and rarely gastric rupture. If the baby needs ongoing respiratory support, use the lowest effective pressures possible

- **Self ventilating in air** – replogle tube as described, IV access, remain nil by mouth with timely transfer to one of the three surgical units
- **Non-invasive respiratory support** – If possible non-invasive support should be avoided, however if required to maintain respiratory stability there needs to be aware of the potential for gastric distension. If there is evidence of RDS then surfactant should be administered via ETT to ensure effective surfactant delivery into the lungs. Although timely transfer is advisable, stabilising and optimising the infant's clinical condition will be of most benefit.
- **Invasive respiratory support** – Pressure controlled ventilation is more effective in these patients with the PIP and PEEP kept as **low as possible** to maintain gas exchange, often a higher rate low pressure strategy is required to maintain gas exchange and reduce the gas in the stomach. Avoid volume-guarantee ventilation as this can worsen gastric distension. If there is evidence of RDS, administer surfactant to improve lung compliance and allow for reduced ventilator pressures.

Transfer Considerations

- Follow the Delivery room management on page two of this guideline
- The replogle tube should secured for transfer, Flush every **15 minutes** with 0.5ml of 0.9% sodium chloride. The vented (blue) end of the replogle tube must remain clear with nothing attached in between flushes
- Secure 2 points of IV access for fluids and drugs. Consideration should be given to obtaining arterial access either peripherally or umbilical prior to transfer, time and condition dependent
- IV fluids/ medications should be in 50ml syringes in preparation for transfer
- Ensure a chest X-Ray has been performed at the referring centre with replogle tube in-situ and that this has been seen by the team transferring the infant. See replogle tube guideline (linked).
- Analgesia/ sedation is not usually required prior to surgery but monitoring of pain and or sedation will be required particularly if requiring respiratory support
- Portable suction will be required on transfer therefore if an air transfer is needed a portable suction will be required to be taken as no suction unit is attached to the air kit. There should always be a spare portable suction unit in the neonatal ambulance. Do not delay transfer of an infant requiring respiratory support to gain arterial access. If the stomach distension has increased please call the surgical team to make them aware prior to arrival as these surgeries are not often done out of hours but may need expedited to prevent further clinical deterioration.