Surgical Conditions Affecting Neonates in Scotland Managed Clinical Network

POSTNATAL STABILISATION AND MANAGEMENT OF BABIES WITH EXOMPHALOS

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.

Contents

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 exomphalos.

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.

The guidelines are underpinned by the following key principles:

1. Babies with exomphalos 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) – see *Appendix 1* for contact details.

2. Network pathways and processes should be in place to ensure that
   - Where there is an antenatal diagnosis of exomphalos, the parents receive multi-disciplinary team (MDT) counselling involving fetal medicine, neonatology and paediatric surgery, ideally as a joint session.*
   - Where there is an antenatal diagnosis of exomphalos, birth is planned in a maternity hospital co-located with a surgical NICU.
   - Where 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.
   - Repatriation from the surgical NICU to the local hospital is facilitated as soon as possible when clinically appropriate (2).

*Note MDT counselling may not be required if the parents opt to not continue the pregnancy.

Clinical Background

Exomphalos is a defect of the anterior abdominal wall which results in bowel contents herniating into the umbilical cord. In exomphalos major the defect is >5 cm and usually contains bowel and liver, whilst in exomphalos minor the defect is <5 cm and contains only bowel. Most commonly the amniotic membrane sac covering the contents is intact at delivery although occasionally sac rupture can occur. Around half of babies will have associated anomalies, including underlying genetic / chromosomal disorders.

Management at birth

- Prior to birth a team brief involving obstetric, anaesthetic and neonatal staff should be held to facilitate effective communication and establish a stabilisation plan. This plan

should be informed by the antenatal findings, including size of the exomphalos and presence of any associated anomalies.

- 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] Exomphalos itself is not considered a contra-indication to DCC. When cut, the umbilical cord should be left at least 15cm long.

- Neonatal staff with appropriate training and competencies should attend the delivery and initial stabilisation should be in accordance with NLS guidance.

- Intubation is not routinely recommended and any decision to intubate should be based on NLS guidance rather than the presence of exomphalos. However if the defect is large and the predicted lung volumes low the baby may require ongoing respiratory support / intubation.

- Routine monitoring should be applied (oxygen saturations on the right hand, heart rate and temperature), and a large bore naso/orogastric tube inserted to drain the stomach.

- The exompahlos sac should be inspected for evidence of damage (if any is identified the surgeons should be informed immediately) and wrapped carefully in occlusive (cling film) covering or placed in a plastic bag. This will provide protection and minimise evaporative heat and fluid losses.

- Once stabilised, position the baby on their right side. This avoids traction on the hepatic vessels and minimises the risk of cardiovascular instability.

- If clinical condition allows parents should have the opportunity to cuddle their baby prior to transfer to NICU, taking care to support and protect the exompahlos sac.

Management following admission to NICU

1. Monitoring and vascular access

- Standard NICU monitoring should be applied.
- Peripheral venous access should be established and baseline bloods (including genetic and blood bank samples – see below) and a blood gas obtained. Many babies with exomphalos, especially those with exomphalos major need several weeks of parenteral nutrition before establishing enteral nutrition and consideration should be given to the early insertion of a per cutaneous long line (PICC).

2. Referral to/discussion with specialist surgical 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.

3. Intravenous fluid management/fluid balance

Intravenous fluid management should be guided by local practice. Babies with exomphalos may initially have high gastric losses, and consideration should be given to replacing these if volumes in excess of 20ml/kg are aspirated, especially if accompanied by signs of hypovolaemia. Normal (0.9%) saline with additional potassium chloride (10mmol/500ml) is recommended as replacement fluid.

4. 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.

5. Antibiotics

Antibiotics are not routinely recommended in the absence of other risk factors for, or clinical signs of sepsis.

6. Enteral feeding

Enteral feeding is usually delayed until a decision regarding management of the defect has been made.

7. Specialist review / investigations

Exomphalos is commonly associated with underlying syndromic/genetic diagnoses and all babies should have the following investigations performed in addition to a thorough clinical examination and plotting on an appropriate growth chart:

- Genetic testing (karyotype, microarray and mutation analysis for Beckwith-Wiedemann syndrome)
- Cardiology review and echocardiogram
- Cranial/renal ultrasound scans

8. Surgical review and management

Surgical review of a patient with exomphalos is generally not time critical unless the exomphalos sac has been damaged, in which case the surgical team should be informed immediately. A decision on the most appropriate method of closure is based on assessment of the size, content and sac integrity of the defect, as well as the presence of co-morbidities e.g. congenital heart disease. Management options include early, primary closure or delayed closure with escharisation.

9. Early surgical closure

Small defects are usually amenable to early, sutured closure in theatre. The choice of repair will be determined by the operating surgeon who will also make an assessment of the rotation of the bowel during surgery if possible.

Post-operative care after surgical closure should include:

- Gastric decompression with appropriate nasogastric drainage.
- Monitoring for signs of abdominal compartment syndrome (which is a risk after return of bowel into a small abdominal cavity) for at least the first 48 hours after surgery. This should include assessment of urine output, abdominal distension and peripheral perfusion including lactate levels.
- Analgesia as indicated by pain scoring/nursing assessment.
- Peri-operative antibiotics for 24 hours after closure unless culture positive or evidence of ongoing sepsis.
- Early initiation of trophic enteral feeding can be considered any time after closure but should have commenced within 48 hours of gastric losses becoming non-bilious. Maternal breast milk should be recommended

10. Delayed closure with escharisation

There are various methods of delayed closure. All include the same basic process of covering the sac in a compound which promotes escharisation, and covering this with a dressing to contain liver growth. A detailed description with images is available in the accompanying SCANS guideline.

The process of escharisation can take many weeks or months and therefore where possible normalisation of neonatal care should be encouraged. Families can be supported to learn how to perform the dressing themselves, which may facilitate earlier discharge home or repatriation to their local hospital. By definition, a delayed closure will require surgical control of the ventral hernia in later life – but this is beyond the scope of this document.

11. Importance of allied healthcare professionals

Babies with exomphalos major benefit hugely from the expertise of allied healthcare professionals. They often have poor weight gain due to a combination of a high calorie requirement and co-existing severe gastro-oesophageal reflux, and early input from specialist dieticians is essential. Their long, often complex inpatient stays and poor truncal musculature can impact negatively on their development and regular review by developmental physio and speech and language therapists is a fundamental component of their care. Additionally specialist orthotic services are required to fit and custom make the corsets required to control the ventral hernia as they grow.

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 DCC
4. Newborn Resuscitation and support of transition of infants at birth Guidelines, Resuscitation Council UK, 2021

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 Exomphalos
Infants with exomphalos 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 exomphalos 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 presentation to their local centre for prompt transfer after birth.
- 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. The call should be made before birth wherever possible, to allow confirmation of cot destination and immediate stabilisation plans to be made without delaying transfer.
- Referrals of these infants should be should be triaged by the on-call neonatal transport consultant and the team dispatched accordingly.

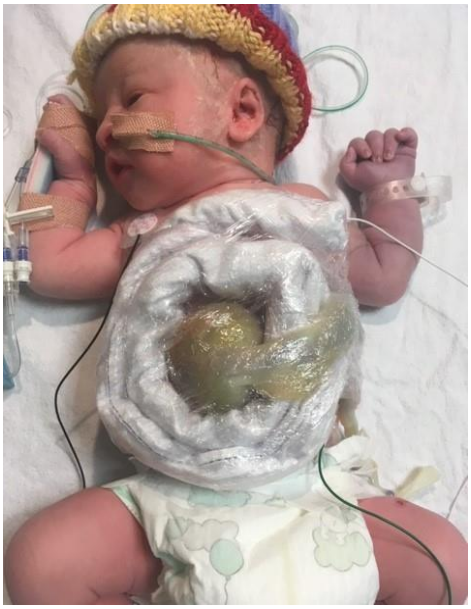
Transfer Considerations

- Follow the Delivery room management on page two of this guideline
- Leave the NG tube on free drainage and aspirate regularly. Close monitoring/documentation of output is necessary as these can be large and persistent in these infants. Losses should be replaced when >20mls/kg/day in a well perfused infant. If replacing losses please use 0.9% sodium chloride with 10 millimoles of potassium per 500ml bag.
- Secure 2 points of IV access. Avoid the umbilical route in babies with exomphalos.
- IV fluids/ medications should be in 50ml syringes in preparation for transfer.
- Analgesia or sedation needs to be considered prior to transfer and handling of the baby. This is not usually required with this condition unless ventilated for a respiratory reason.
- Infants should be nursed on right side to reduce tension on hepatic vessels, particularly in exomphalos major (see below).
- Temperature monitoring on at all times
- Ensure maximal interventions in place to minimise heat and fluid loss.
- Ensure the guidance of the occlusive plastic wrap has been followed (see below).
- Keep incubator doors closed and utilise heat shields where possible when doors are open
- Place the baby with the wrapped defect into a plastic bag when transferring into the transport incubator
- Weigh nappies to allow adequate measurement of fluid balance.

- Ongoing frequent assessment for signs of hypovolaemia and hypothermia and address promptly.
- If persistent acidosis, be sure to check the lactate, chloride and bicarbonate with each blood gas. In these infants it is common for an acidosis to be driven by high chloride following fluid resuscitation rather than a metabolic acidosis from hypo-perfusion.
- Prepare fluid boluses prior to departing from referral unit to deliver during the journey.

Positioning
Positioning of the infant will be key to optimise visualisation of the defect and protect the integrity sac during the transfer. **Baby should be nursed on right side** to reduce traction on the hepatic vessels. 'Kinking' of the hepatic vessels can lead to haemodynamic instability. Lights should be on in ambulance/incubator for journey to allow good visualisation and assessment throughout. Particular attention should be paid during loading and unloading as this is when there is considerable risk from movement. It is imperative to clearly document the appearance, size and any leakage of the defect before during and on completion of the transfer.

This picture shows a supportive doughnut around the defect which is then all wrapped in a clear occlusive dressing to protect it all. The baby should still be nursed on right side with or without the doughnut support for transfer.



Parents
Parents need to be aware that there may be the need to expedite surgical review / surgical treatment, and depending on the time-frames there is a chance that they may not be present when this happens.

Sac Rupture
The rupture of the sac makes transfer more emergent. On these occasions, priority must be given to rapidly facilitating transfer of the infant to the transport incubator

followed by rapid transfer to the destination hospital. Communication with the surgical team receiving the baby is imperative for any significant clinical change.