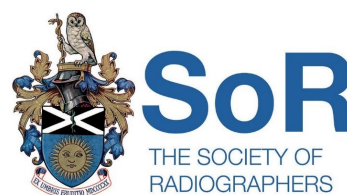


National care principles for the management of Congenital Diaphragmatic Hernia (CDH)

Endorsed by:



Bliss

As a patient organisation, Bliss strongly supports the aim of this document to improve the quality and consistency of care for women with affected pregnancies and their babies, and, in particular, the importance the document places on providing clear and compassionate communication and support to parents which empowers them to be fully involved in all decisions regarding their pregnancy.

ECMO

The UK ECMO centres have discussed the document and are supportive of its recommendations.

Background

In December 2014 MBRRACE-UK launched a report on the management of congenital diaphragmatic hernia (CDH) based on the findings of a confidential enquiry (1). The cases selected for the enquiry were derived from an existing study (2) and had been drawn from across the UK and represented all types of CDH from the most severe to the most benign. Although many examples of good practice were identified a key finding from the enquiry was the enormous variation that existed in every aspect of care provided from the moment the diagnosis was first suspected through to either discharge / follow up or death. As a consequence of these findings it was recommended that the care of women with affected pregnancies and their babies should become more consistent. To achieve this aim the British Association of Perinatal Medicine (BAPM) convened a group comprised of representatives from all of the relevant UK specialty organisations and other stakeholders (see Appendix) with the aim of:

1) Identifying the principles of care that should be delivered in managing this condition from the time it is first suspected.

2) Highlighting the organisational changes necessary to meet the aspirations of parents in terms of the quality and consistency of care they receive wherever this is delivered across the UK.

This document represents the agreed view of that group and highlights the approach that centres managing this condition should deliver.

Diagnosis

Around 60% of pregnancies in which the baby is affected by CDH are diagnosed antenatally by mid trimester ultrasound. CDH is one of the 11 auditable conditions that are included in the fetal anomaly screening programme (FASP). This programme is based on the 18⁺⁰ to 20⁺⁶ week fetal anomaly screening scan in England and other similar programmes in the devolved nations¹(see examples below). Existing FASP standards require that where an anomaly is suspected at the 18⁺⁰ to 20⁺⁶ week scan the woman should be seen by a specialist obstetrician or fetal medicine consultant for discussion and confirmation and that this second opinion should take place within 3 or 5 working days of the initial scan depending on whether the confirmation occurs in house or requires referral to a tertiary centre. CDH may also be diagnosed on scan for the first time after the 18⁺⁰ to 20⁺⁶ fetal anomaly scan as part of a growth monitoring scan, or scan for some other clinical reason, and these cases do not come under the FASP remit. These present routine screening arrangements permit confirmation of the diagnosis and discussions regarding whether to continue the pregnancy to take place locally with involvement of an obstetrician experienced in ultrasound diagnosis working in a centre that does not provide ongoing care for babies affected by CDH. However, the group reviewing the care pathway took the view that once a diagnosis of CDH was confirmed all women should:

1) receive relevant literature such as that produced by ARC and CDH-UK;

2) have the opportunity of a multi-disciplinary discussion about the management options in a centre that regularly cares for pregnant women and their babies affected by CDH.

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https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/421650/FASP_Standards_April_2015_final_2_.pdf

<http://www.nsd.scot.nhs.uk/services/nmcn/cdh.html>

It was agreed that the same time limit of five working days from the initial screening scan, as applies currently under the FASP programme, should apply to discussions at the specialist centre (generally termed by clinicians as counselling). These specialist discussions should encompass all aspects of CDH including its management, complications and prognosis and involve as a minimum:

1. A fetal medicine specialist;
2. A neonatologist;
3. A neonatal surgeon.

However, access to a psychologist should be readily available in addition. Access to a genetic specialist should also be routinely offered in order that parents can discuss the risk of associated problems in the current (affected pregnancy) and the risk of recurrence in future pregnancies.

Key components of the multidisciplinary discussions should be:

- 1) That the overall rate of survival for babies diagnosed with CDH at around 20 weeks of gestation is 50% (local figures based on recent cases should not be used).
- 2) That in babies who survive there are commonly long-term problems which are most frequently neurodevelopmental, respiratory or gastrointestinal in nature.
- 3) That the presence of additional genetic problems is associated with a worse prognosis especially in terms of long-term problems.
- 4) The presence of major cardiac abnormalities is associated with a worse prognosis.
- 5) The use of imaging to estimate the severity of the CDH should be offered as part of the assessment process. However, the optimum approach to severity prediction requires further research.
- 6) That in-utero interventions exist but that at present all of these remain experimental and none have been shown to reliably improve outcome in human studies. Involvement in these types of intervention should only occur where parents are fully informed of existing evidence and, after appropriate explanation, they have signed a consent form.

In all cases a written summary of the discussions should be made available to the mother / parents and all relevant primary and secondary healthcare providers within 5 working days of meeting with the MDT.

Care should be organised in a way that allows the parent(s) the maximum amount of time possible to consider the treatment options available to them.

Patient choice not to continue the pregnancy

Where, after a full discussion of the overall prognosis of the baby as understood by the clinical team, the parent(s) decide not to continue the pregnancy examination of the fetus by a perinatal pathologist and genetic counselling should be offered. In addition a follow up visit should be offered with a fetal medicine specialist in the tertiary centre previously involved in the management. Where all decisions and counselling occur locally a follow up appointment at a tertiary centre should still be offered.

Although the law in much of the UK largely excludes termination of a pregnancy after 24 weeks of gestation there will be circumstances where the evidence emerging about a fetus affected by CDH is such that the criteria for late termination (i.e. after 24 weeks) are fulfilled. Termination is not permitted in Northern Ireland.

Patient choice to continue the pregnancy but with palliative care after birth

Parent(s) should be made aware during discussions following diagnosis that where additional information makes the baby's overall prognosis particularly poor (e.g. when diaphragmatic hernia is part of an underlying diagnosis of trisomy 13 or other major structural anomaly) they have the option to continue the pregnancy but with palliative care provided after birth. Where this option is chosen a careful plan covering all aspects of the delivery and care to be provided to the baby after birth should be developed with the family in a timely fashion and well in advance of the expected date of delivery. A copy of this plan should be placed in the maternal notes as well as the mother's hand-held record and all members of the MDT involved in the mother's care should be made aware of it. This applies to the mother's local hospital as well as any additional tertiary referral centre.

Care during pregnancy

Arrangements for care of the mother during the pregnancy need to reflect family's wishes, personal circumstances, geography and the needs of each individual case for regular specialist fetal medicine involvement. However, the pregnancy should proceed under the supervision of the relevant tertiary centre.

Parent(s) should be aware that the routine use of MRI to monitor the baby:

- may be recommended on a case by case basis
- is not of proven benefit and centres that use fetal MRI should undertake formal evaluation in comparison to detailed ultrasound to determine if additional information is obtained and more accurate prediction characteristics are provided
- does not offer additional certainty regarding the long-term prognosis.

Similarly parent(s) should be aware that the practice of administering steroids to the mother prior to labour in order to improve the baby's lung function after birth is of no proven benefit in cases of CDH. However, their use may be offered on the basis of theoretical benefit and the unlikely risk of harm.

Delivery

The need for postnatal transfer should be minimised. This requires, where practicable, delivery to take place in a centre with facilities to carry out surgical care, and capable of providing on site all aspects of neonatal intensive care in the pre- and post-operative period.

Where possible the pregnancy should continue until at least 39 weeks of gestation. There is no evidence that delivery by caesarean section offers improved outcome in terms of the baby's condition at birth. However, the risks of caesarean delivery to the mother should be discussed and weighed against the advantages for the baby of being able to control the timing of delivery and hence ensuring an appropriate resuscitation team is immediately available.

Transport

The management plan for babies identified antenatally as affected by CDH should aim to minimise the need for transfer after delivery. However around 30% of babies affected by CDH will only present after delivery.

Where transport of an antenatally diagnosed baby is required for any reason this should be carried out by a team that includes a senior transport clinician with experience of managing complex surgical patients. Transport teams should arrive at the baby's cot side within 3.5 hours of the transfer request being made. This should include, where necessary, the use of air transport to achieve these timings. Arrangements for the transfer of the mother and father in order to be with the baby should be set in place at the earliest opportunity. However, staff should aim to take account of the financial and practical difficulties for the family associated with transfer away from home.

Resuscitation

The resuscitation of the baby should be led by a neonatal consultant physically present at the delivery who will be responsible for:

- a. Decisions regarding specific aspects of management such as immediate intubation, the early use of muscle relaxants, and the passage of a nasogastric tube as well as the approach to respiratory and cardiovascular support.
- b. Any decision to re-orientate care based on the baby's response to initial resuscitation.

The consultant should also ensure that parents are kept informed of the baby's progress in a timely manner and that whenever possible parents have the opportunity to be actively involved in decision making.

Neonatal care

Any neonatal unit providing care for babies affected by CDH should have immediate access to cardiac scanning (structural and functional) and on site specialist support from paediatric respiratory medicine specialists, paediatric radiologists, paediatric anaesthetists and neonatal surgeons with experience in the surgical management of CDH. They should also be cared for by appropriately trained and experienced neonatal nurses.

In terms of initial management units should:

- a. Ensure that the initial establishment of monitoring, vascular access and support is performed efficiently and in a timely fashion under the direct supervision of a consultant.
- b. Have a clearly established approach to such babies in terms of escalating support that recognises the need to:

- i) minimise potential ventilator-induced lung damage,
 - ii) reduce pulmonary hypertension
 - iii) optimise systemic blood pressure.
- c. Be capable of making decisions based on multi-disciplinary discussions with parents about the timing of surgery and/or re-orientation of care.
- d. Have established care pathways in relation to the use of ECMO.
- e. Ensure there is clear and regular communication of progress to parents.
- f. Ensure arrangements are in place to meet the wider needs of parents including accommodation, breast feeding support and clear policies for the delivery of family-centred care. Information from Bliss is available [here](#) and should be made available to parents.

Anaesthesia

Arrangements for anaesthesia should be directly led by a consultant paediatric anaesthetist. No particular anaesthetic technique is recommended however due consideration should be given to the general issues that relate to the pathophysiology of the condition and their treatment such as problems of reduced gas transfer capacity, compliance and lung volumes, increased pulmonary resistance, right ventricular strain and failure.

Surgery

Timing of Surgery - Repair of the CDH should be undertaken when there is clinical and echocardiographic evidence of resolving pulmonary hypertension and at a time of haemodynamic stability. However the scheduling should also take account of the availability of appropriate staff and hence should occur during normal working hours.

Theatre Team - Teams should include a consultant anaesthetist and consultant neonatal surgeon who have experience of managing newborns with CDH. Where transfer to an operating theatre may compromise haemodynamic stability, the team should have the ability to relocate to the NICU to carry out the repair in that setting.

Surgical technique/principles:

- The benefits and risks for approach (open or minimal access), technique (patch or flap) or material (synthetic or biological) and other reasonable alternatives should follow consent guidance and should be documented as for any other procedure;
- Routine use of intercostal drains in the immediate post-operative period is not recommended;
- Surgical documentation should include:
 - a) Herniated contents
 - b) Size (grade) of defect as proposed by the International CDH Study Group (3)
 - c) Type of repair, including sutures and prosthetic material (if used)
 - d) Other structural anomalies identified

Post-operative management. The neonatal surgical team should be included in decision-making in the post-operative period including regular discussion with parents. Surgeons should provide clear post-operative instructions, including points of contact. Post-operative management decisions should be clearly documented.

Follow up

Following discharge children born with CDH should have on going multi-disciplinary follow up that continues throughout childhood. This should include access to neonatal medicine / paediatrics (depending on the child's age), paediatric surgeons as well as specialists in gastroenterology, respiratory medicine, cardiology, dietetics and neurodevelopment.

In many cases it will be appropriate for children to be given open access to the children's ward.

Babies presenting for the first time after birth

Babies presenting in this way tend to have a better overall prognosis both in terms of long term survival and morbidity. Presentation may occur immediately after birth or be delayed by days, weeks or even years. The same general principles of management should apply but in those babies that are broadly stable the approach to transfer and initial management can be tailored to their needs.

The role of extracorporeal life support (ecls)

The potential role of Extracorporeal Life Support (ECLS) should be mentioned as part of antenatal discussions although parents should be informed that it may well not be needed or be felt to be inappropriate. Babies considered for Extracorporeal Membrane Oxygenation (ECMO) referral should meet the general eligibility criteria. If parents agree to ECMO referral, one of the 6 UK neonatal ECLS centres should be approached using the established neonatal ECLS referral pathway. Early discussion with the ECMO centre is encouraged.

In cases where there is uncertainty about the suitability of ECMO, a further opinion from at least one other ECLS centre should be sought. This further opinion should be organised by the ECLS centre. If the referral ECLS centre cannot offer support for a patient with CDH, they should take responsibility to find an alternative ECMO centre able to offer care as a matter of urgency.

Formal written consent is not mandatory but parents/carers should be made aware of the principles, and risks, of ECLS.

Bereavement care

Of those women identified during pregnancy as carrying a baby affected by CDH around 50% of their babies will ultimately not survive. As a result psychological support and or bereavement care

should be readily available. In particular at specific times of high stress where the outcome is uncertain for example:

- a) During initial discussions regarding whether to continue the pregnancy or opt for a termination;
- b) Around the time of birth / resuscitation;
- c) The period of initial stabilisation on the neonatal unit;
- d) Discussions regarding the appropriateness of carrying out surgery;
- e) Discussions / decisions regarding the re-orientation of care to a palliative approach.

Where a baby dies or care is re-orientated to a palliative approach the lead clinicians should ensure there is excellent communication between all relevant providers.

Parents should be signposted to external sources of emotional support. These sources may include CDH-UK, ARC and SANDS.

Organisation of care

It is estimated that between 200 and 300 pregnancies are affected by CDH in the UK each year (4,5). The initial diagnosis and management of these pregnancies is shared amongst a large number of centres many of whom will deal with very few cases annually. Of those affected pregnancies continuing to delivery (estimated to be 150 to 200 annually (4,5)) not all will be sufficiently stable to undergo surgery. However the remaining cases that do have surgery are cared for in some 27 units. Since most surgical centres will have four or more surgeons individual experience of this condition will often be limited to a single case per year. The same limited experience will also affect neonatal teams as well as anaesthetic and theatre teams.

It is our view that the current organisational arrangements hinder attempts to improve both the patient (family) experience and outcomes in this condition. The features of the care pathway outlined in this document will be difficult to achieve under the present organisational arrangements. Focussing of the care of CDH patients in a smaller number of highly specialised centres would facilitate the achievement of these principles and allow those centres providing the care to develop a genuine body of experience in dealing with this condition. In deciding on the number of centres providing care for babies affected by CDH it will be important to recognise that in certain parts of the country, because of geography, decisions over where care should be delivered will also need to take account of access to services and potential transport difficulties. Similarly, and affecting many more families, the increased focussing of care will require the provision of facilities for families to be resident in hospital for a prolonged period. This will include not only appropriate accommodation but access to support services such as access to a psychologist, access to social services and financial support where appropriate.

Research and Development (R&D)

As well as improving patient care the focussing of CDH cases in a smaller set of specialised centres would provide additional opportunities including research and development. For example centres providing care for children affected by CDH should have a lead consultant for R&D who should:

- a. Facilitate the reporting of all CDH cases to a national register. The group agreed that such a register was a key step to improving both the management and outcomes in this condition. The data submitted to a national register could also be used to:
 - i. Focus incrementally on particular aspects of care (e.g. the role of fetal MRI) with the aim of improving outcomes at a national level.
 - ii. Supply data to the national congenital anomalies register.
- b. Facilitate involvement in both local and national audits. This will include ensuring the outcome of affected babies is communicated to the relevant teams within the local network.
- c. Facilitate the formation of a research network focussed on the management of CDH.

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