

Congenital Diaphragmatic Hernia (CDH) Lay Report

In brief

In the UK today, almost one in every 100 babies is stillborn or dies soon after birth. Up to 100 women die every year during or just after pregnancy. These figures are unusually high for a developed country. A team of academics, clinicians and charity representatives called MBRRACE-UK has been asked by the NHS to look at why this is so, and how care for mothers and babies can be improved.

In the first of a series of Confidential Enquiries, MBRRACE-UK examined congenital diaphragmatic hernia (CDH), an uncommon and complex condition which is not well understood. They identified many instances of excellent care, with teams of specialists, led by a consultant, working well together and offering solid support to parents throughout the process, from diagnosis to follow-up care. But they also found examples of poor and unacceptable care, where parents received no or inadequate counselling, where documents were inaccurate or missing, or where life-saving surgery had to be delayed because not enough special cots were available.

As a result of the enquiries, MBRRACE-UK recommended that in future a package of care should be tailored to the mother and baby's medical and surgical needs, and offered at a small number of centres staffed by dedicated teams of specialists from obstetricians and neonatal nurses to psychologists and social workers.

Background

Every day in the UK 17 families are devastated by the death of their baby shortly before or soon after birth. Some of these deaths could be avoided. The NHS has recognised that improvements are needed in the health and care of women and their babies throughout the UK, and so it collects information about those who die or are born very sick every year to identify patterns in illness and disease in order to work out the best way to deliver care. This work is carried out by MBRRACE-UK, a team of academics and clinicians, a GP and representatives from the charity Sands.

MBRRACE-UK also conducts what are called Confidential Enquiries. These are an opportunity to gain a fuller picture of what happens when a particular illness occurs and how it might be treated or even prevented in the future. In a Confidential Enquiry, a panel of experts meets to look in detail at a sample of case notes (e.g. maternity records, referral letters and investigations such as scans, X-rays or a post-mortem examination) of mothers or babies who have died and to ask: did the mother or baby receive the right care, and how could it have been better?

For the 2015 report, MBRRACE decided to focus on the care of unborn and newborn babies with congenital diaphragmatic hernia (CDH). Relatively little is known about the best way to care for babies with CDH, and many either do not survive or survive with serious long-term health problems. At the moment they have to be monitored, cared for and treated by a large number of people.

What is congenital diaphragmatic hernia (CDH)?

CDH occurs when the baby's diaphragm (a thin sheet of muscle that separates the heart and lungs from the stomach and helps us to breathe) does not develop properly. This means that the stomach

generally there is no specific pathway of care that is followed each time a baby is diagnosed with CDH.

Also, there hasn't been much scientific evidence developed around what is the best care for babies with CDH. This evidence would form the basis of guidelines, policies and practices that could be used nationally or even internationally to make sure every baby got consistently good care.

What was considered to be good care?

The panel found many examples of good or excellent care, which included:

- A clear pathway from diagnosis to follow-up
- Teams of people from different specialties working well together, led by consultants who were available round-the-clock
- Excellent communication with and support for parents, e.g. encouraging mothers to breastfeed/express milk and giving families somewhere to stay during acute care, and
- Follow-up bereavement support at the parents' home, helping them to create memories and mementoes of their baby.

What was considered to be poor care?

- Sometimes counselling and support was only available at a specialist centre. At other times the wrong information was given. In a rare case, no psychological support was offered:
A woman diagnosed as carrying a child with CDH was transferred to her district hospital at 34 weeks, because she was bleeding and in pain. After being poorly looked after for 12 hours, she was sent home a day after delivering a stillborn girl. A plan of care was not followed and a lack of resources - "the unit was very busy" - was blamed.
- The care mothers and babies received wasn't consistent, with some receiving special treatment and others not. These include: surgery on the baby when still inside the womb (using a technique called FETO which encourages the baby's lungs to grow by putting a small balloon into the windpipe before birth); and ECMO (a machine which does the work of a baby's lungs and heart, breathing and delivering oxygen into the blood).
- There sometimes wasn't enough thought given to the time of day that the mother should give birth: delivering and stabilising a very sick baby is much more difficult overnight or at weekends when some important members of the team may not be available.
- There were not always enough special cots. This is particularly upsetting, when parents don't know from one day to the next when a cot will be available. It means that induction (when labour is brought on with the use of drugs) and even life-saving surgery may have to wait for several days:
A young girl, diagnosed as carrying a baby with CDH at 31 weeks, gave birth to a boy at 37 weeks in her local hospital. He spent six days in intensive care waiting for a bed at the local children's surgical unit. He then had to wait another five days for surgery.
- Follow-up care was patchy and regular follow-up care after the baby went home from hospital often depended on where parents lived and whether they had access to specialists such as dieticians who advise on feeding and nutrition or physiotherapists who help with the baby's development.

- Important documents were incomplete or missing. Some sets of notes contained very little detail, particularly concerning a decision to end a pregnancy. Others records referred to the wrong patient or provided information insensitively.

What do parents think?

This Confidential Review looked at cases from 2009 and 2010. To check whether or not the issues raised in the panel reviews were still relevant, CDH-UK asked its members about their experiences of care in 2013. From 115 responses, the charity found that little had changed in three to four years: parents felt that before birth, staff did not understand CDH and therefore did not support them or communicate well or sensitively about it; during birth, there were too few staff or resources such as cots; and after birth, there was a lack of knowledge in the non-specialist centres as well as lack of support and coordination of care after the baby was taken home or after bereavement.

What are the recommendations following the review?

The Confidential Enquiry recommended that the service be more patient-centred i.e. a package of care should be tailored according to the mother and baby's medical and surgical needs, rather than put together, sometimes hastily, depending on what resources are available.

This will involve changing the way care is delivered. This should be at a smaller number of centres which are run by clinical networks (teams of people with very specialist expertise e.g. obstetricians with particular experience of dealing with CDH) to manage the care of very ill babies.

While having a smaller number of centres with dedicated services will mean some parents will have to travel longer distances than they currently do, it will make it easier to:

- Include all important elements of care, such as counselling and psychological support (especially for decisions about ending a pregnancy so parents can make an informed choice).
- Agree on the best way to manage the care and treatment of babies diagnosed with CDH and how to handle late termination of pregnancy and set UK-wide standards.
- Produce a national 'information sheet' with clear and consistent information about CDH.
- Keep track of the correct and complete documentation of each case of CDH.
- Bring researchers together to work on scientific studies so that approaches to the care of babies with the condition can become more based on good-quality evidence.
- Make sure all cases of CDH are recorded and followed up properly on a proposed NHS register, which will give researchers a single, reliable source of information from which to learn more about trends in conditions where there is a problem about how the baby develops in the womb, such as CDH, across the UK.

CDH-UK also made some recommendations:

- there should be more information about CDH and its implications
- parents should have opportunities to talk to other parents
- parents should be given written information and details of support groups
- doctors and nurses other than those in specialist centres need better knowledge of CDH, and
- there should be better coordinated follow-up care.

Further reading

To read the full report, [MBRRACE-UK 2013/2014 Perinatal Confidential Enquiry – Congenital Diaphragmatic Hernia \(CDH\)](#).

To read more about the involvement of patients and the public in decisions about services, and to find out about the topics for confidential enquiries that MBRRACE will cover, see www.npeu.ox.ac.uk/MBRRACE-UK.