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What is a Congenital Diaphragmatic Hernia (CDH)?

CDH occurs when the baby's diaphragm does not form properly during devleopment. With CDH, some of the baby's abdominal organs, such as the bowel and liver, go through a hole in the diaphragm at about 10 to 12 weeks into the pregnancy and take up space in the chest where the lungs should be growing. CDH occurs on either side, but much more commonly on the left.

What happens in CDH?

The main problem in CDH is that the lung on the side of the hernia (and even the lung on the opposite side) is squashed by the abdominal organs cannot grow to its full size. This is known as lung hypoplasia. Many babies with CDH also have pulmonary hypertension. This is due to a failure of the blood vessels in the lungs to relax after birth and let blood flow through them so that they can pick up oxygen from the lung air sacs

What causes CDH and how common is it?

There is no known cause of CDH, although it may be genetically linked. CDH occurs in about 1 in every 3,000 babies and accounts for 8% of all major birth defects. Up to 20% of infants also have a chromosomal abnormality such as Down's or Edward's syndrome.

The chance of having another baby with isolated CDH (no other genetic condition) is 2%.

CDH during pregnancy

Many babies with CDH are detected at the 18-20 week fetal anomaly ultrasound scan (FAS) that is routinely offered to pregnant women (around 25-30% are not identified on antenatal scans for a variety of reasons). Mothers who have had a fetal diagnosis of DH made will have access to up-to-date counselling advice together with written information and details of further resources including detailed scans by fetal medicine experts.

Perinatal

For all identified antenatal diagnosis of DH, delivery will be planned by a multidisciplinary perinatal medicine team at one of the three Maternity Units in Scotland which are allied to Neonatal Surgery Units. These are in Aberdeen, Edinburgh and Glasgow. This will be in accordance with the SDHCN Antenatal pathway.

If not diagnosed antenatally, and delivered in a more peripheral unit, the affected neonate should be transferred to one of the above units according to the agreed SDHCN Postnatal pathway. This transfer would be undertaken by the most appropriate Neonatal Transfer Team with the destination unit depending on the clinical condition of the patient.

Surgery and Long –Term Follow-up

Surgery will be timed according to the response of the infant to medical management along agreed guidelines. Post-operative management will be multidisciplinary with similar guidelines to pre-operative management. Average length of stay is difficult to predict and is dependant on the complexity of the case, but is rarely less than 3 weeks.

It is important to recognise that these babies require long term follow-up by many different health professionals. Many hospitals provide this in a multidisciplinary clinic. This ensures that their surgical, nutritional, respiratory, neurodevelopmental and emotional needs are addressed in a coordinated way allowing children to achieve their maximum potential.

Scottish Diaphragmatic Hernia Clinical Network (SDHCN)

The SDHCN is a nationally supported network, designated in 2008, that includes doctors, midwives, nurses and parents. It was set up to produce and deliver best standards of care for CDH, so that mothers and babies affected by CDH receive the best treatment.

The SDHCN encompasses the diagnosis (antenatal if appropriate), medical and surgical treatment and long-term follow up of children born with congenital diaphragmatic hernia. This condition requires the input of a tertiary centre but many aspects of follow-up care can be managed locally. The network facilitates consistency to ensure that all health professionals, parents/carers and patients themselves have equitable and evidence based treatment plans.

To date the NMCN has undertaken work to create pathways and guidelines for the management of CDH at antenatal, inpatient and follow up stages. Parent information leaflets have been developed to ensure all parents get appropriate information at antenatal and postnatal stages. The network has developed a website to share information about CDH and all materials developed by the network are available here to professionals and families. The network has also developed and published 8 evidence based standards of best practice, aligned to different stages of the patient pathway. Each of these standards has a measurable quality indicator.

The main objectives of the SDHCN are as follows:-

1. To ensure equity of access across Scotland to a standardised high quality care pathway for mother and baby across antenatal, perinatal, surgery and long-term follow-up.
2. To collect and evaluate service delivery model against agreed standards and quality indicators.
3. To introduce a formalised management structure that will support the coordinated strategic long term planning for future service delivery of diaphragmatic hernia.
4. To identify education and training needs and facilitate the design and delivery of education and training.