

Information leaflet for parents/carers

Hirschsprung's Disease

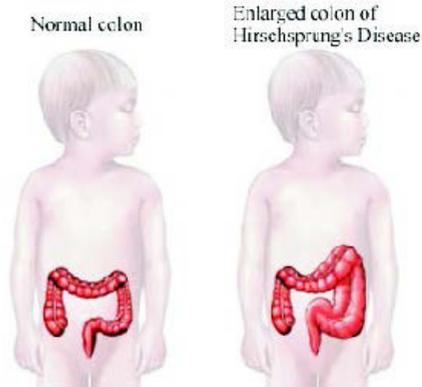


Hirschsprung's Disease (HD)

Hirschsprung's is a rare disease affecting about 1 in 5000 babies, and is much more common in boys than in girls.

Usually the muscles in the intestines push stools through to the bottom for the baby to pass them out.

The muscles work because there are nerve cells which make them push. In your baby these nerves are missing in the last part of the intestine (shown in the picture below on the left) and so your baby cannot pass stools out and they become trapped in the colon which becomes enlarged (shown in the pictures below)



Hirschsprung's Disease is a problem with your baby's intestines that mean your baby is unable to pass stools (poo). This will be explained in more detail below.

This leaflet will give you more information on the condition and what you can expect in the care and treatment your baby will receive.

How is it diagnosed?

The symptoms that suggest HD include:

- When a baby doesn't have their first bowel movement (poo) in the first 24 hours after birth
- When a baby vomits a green liquid called bile after they are fed
- Swollen tummy area

The problems a child experiences from HD will depend on how long a section of the intestine is affected. Most babies have only very a small section missing the nerves, but some may have a much larger section without nerves.

This problem is rarely diagnosed using ultrasound scans before birth

Treatment and Care After Delivery

Feeding

Breast milk is very important for every baby but especially sick babies and even more so for those with bowel/ stomach problems. Breast milk is far more than just food, it contains essential nutrients your baby needs to grow, develop, and recover from surgery or illness.

Although your baby cannot take milk at this time, in order to establish and/or maintain your breast milk supply for when they are well enough to feed, we encourage you to express your breast milk. It is important that you start expressing milk within 6 hours of birth and continue to do this at least 8-10 times a day, including at least once every night.

There is a quiet, private area on the ward that you can use to breast feed your baby or express breast milk. Expressing milk

can be done by hand, which is useful in the first few days after birth when you can expect to express small amounts of sticky clear or yellow fluid called 'colostrum'.

Electric breast pumps are available for use on the ward. Your expressed breast milk will be stored safely until your baby is well enough to feed. The nursing staff will be able to show you how to express and store your milk and arrange for you to have access to a breast pump.

Surgery

There are a range of surgical treatments for HD depending on a number of factors such as how small/ large a section of the intestine is affected. The surgeons will discuss this with you and explain the options available and what is best for your baby based on his/her specific case.

1. In many cases more than one operation will be needed. These include creating a stoma (a hole so that stools can pass from the healthy part of the intestine through a hole out of the tummy into a bag). This is known as a colostomy. This will be temporary. Another operation can be done to remove the affected part of the intestine and place healthy intestine near to the anus.
2. Another option is that surgery may not be performed until later and for a while before surgery happens your baby can return home. This is only an option in certain cases and would mean you would need to be taught by a nurse first how to provide special care for your baby. This would involve you giving your baby what is known as a 'washout' regularly. Because your baby would be unable to pass stools by itself, the stools need to be washed out by passing a tube into the baby's bottom and running fluids in then draining them out bringing the stools out with it.

3. As mentioned above, you would be given advice and support from nurses on how to do this to make sure you are happy and confident in providing this care.

The surgeon will explain about the options and operations in more detail, discuss any worries you may have and ask you to sign a form giving consent for your child to have the operation. An anaesthetist will also visit you to explain about the anaesthetic.

All the doctors who perform this operation have had lots of experience and will minimise the chance of problems occurring.

All operations carry a small risk of bleeding, during or afterwards. Every anaesthetic carries a risk of complications, but this is very small. Your child's anaesthetist is a very experienced doctor who is trained to deal with any complications.

After Surgery

Your child will come back to the ward to recover, and you will be able to visit as soon as he or she is settled back on the ward. All babies are closely monitored after the operation, and so your child will be connected to monitors to check his or her breathing, heart rate and oxygen levels. He or she will also be given pain relief through the 'drip'.

As your child recovers, you will be able to feed him or her from the breast or bottle, although this may take a few days. Over time and depending on how quickly your child is recovering, the drips and monitors will be removed one by one.

The nurses on the ward will encourage you to look after your child as much as you feel able while he or she is recovering. You may feel anxious, especially while your child is connected to drips and monitors, but it will become easier with time. If you are worried about caring for your child, please talk to the nurses.

At first after the operation your baby's may pass stools frequently and they can be very loose. This means special attention will need to be paid to keep the bottom area clean and dry to avoid the skin getting irritated.

If your baby has a stoma you will receive advice and support from the nurses on the unit and also from a specialist stoma care nurse.

What is the outlook for children with Hirschsprung's Disease?

Most children go on to have normal healthy bowels (are able to go to the toilet normally without problems), but some may have problems controlling their bowel movements until they are older, and in some cases not until they are into their teens. This may be difficult or embarrassing for your child and your local hospital will be able to offer ongoing support and help with these issues.

Alternatively your child may have problems passing stools as the anal opening can be tight. This can be helped by a procedure called anal dilation (widening of the opening). Doctors can discuss this with you if necessary.

The intestine is where nutrients and fluids are absorbed, so if a child has had a large section of intestine removed they may have long term problems with nutrition and fluids. If this is the case then you will receive advice and support from dieticians and doctors who will be able to advise you on how best to deal with these issue.

Is there a support group?

BLISS

BLISS is a support group which is able to offer support and advice to families with babies with a range of conditions.

68 South Lambeth Road
London SW8 1RL

Helpline: [0870 7700 337](tel:08707700337)

Email: Information@bliss.org.uk

Website: www.bliss.org.uk

A UK wide charity offering support, advice and information regardless of a child's condition or disability. Through their Making Contact serve they may also be able to put you in touch with other families who are affected by the same disability / medical condition as your child.

Contact a Family

209-211 City Road
London EC1 1JN

Freephone Helpline: [0808 808 3555](tel:08088083555)

Email: helpline@cafamily.org.uk

Website: www.cafamily.org.uk

Please use this space to write down any notes or questions you might have

Further Information

We hope this information leaflet has been useful and will help you to understand all about your child's condition. However some medical information can be difficult to understand. If you need more information or have any concerns please speak to a member of the medical team caring for you or your baby.

Looking after and sharing information about your child

We have a duty of care to help patients and families understand how information about them is kept and shared and we include the following information in all our patient leaflets:

Information is collected about patients relevant to their diagnosis, treatment and care. We store it in written records and electronically on computer. As a necessary part of that care and treatment we may have to share some patient information with other people and organisations who are either responsible or directly involved in the patient's care. This may involve taking the patient's information off site. We may also have to share some information for other purposes; such as research etc. Any information that is shared in this way will not identify the patient unless we have the patient's and parent's/carer's consent. If you have any questions and/or do not want us to share that information with others, please talk to the people looking after your child or contact PALS (Patient Advice and Liaison Service) on 0121 333 8403.

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