

Hirschsprung's disease

This leaflet explains about Hirschsprung's disease. If you have any further questions or concerns, please speak to a doctor or nurse caring for your baby.

What is Hirschsprung's disease and why does my child have it?

Hirschsprung's disease (HD) is a rare condition that affects the nerve cells of the bowel causing severe constipation or blockage of the bowel. This disease can affect 1 in 5,000 babies and is seen more in boys than in girls.

Normally, the muscles in the bowel squeeze rhythmically to push poo through the bowel; these movements are controlled by nerve cells known as the ganglion cells. In HD, these nerves are missing from part of the bowel meaning that the poo cannot be pushed through the bowel as normal. Therefore, it is trapped in the large bowel causing it to be blocked and enlarged. This can lead to a bowel infection called enterocolitis. The length of the affected part of the bowel can vary. It is usually the last part of the large bowel that is affected.

What are the signs and symptoms?

It is usually recognised shortly after birth. A delay in the baby passing the black sticky poo, known as meconium, within 48 hours after birth is a common symptom. Other symptoms and signs that can be seen are:

- Big tummy
- Repeated green vomit
- Constipation
- Baby may be reluctant to feed

Does my child need any tests to confirm the diagnosis?

Yes, a physical examination of your baby's abdomen is needed, along with a rectal examination which requires the doctor/nurse to insert a finger into the bottom to feel for any abnormalities.

If HD is suspected then an x-ray of the baby's stomach and gut is done.

A rectal suction biopsy is also required which is necessary to test reliably for HD. This examination takes place on the Neonatal Unit and involves taking a small piece of tissue, using a lubricated instrument, from your baby's rectum (last part of the bowel). As there are no sensory nerves at the site of the biopsy, this is not painful. The sample is then taken to the laboratory and looked at under a microscope to confirm the absence of ganglion cells. The results can take several days.

Other tests include a blood test to check for infection (enterocolitis).

What treatments are available?

All children with HD will eventually require surgery (usually by three months of age), however the initial treatment is to:

- Stop milk feeds and give fluids through a drip into a vein
- Pass a tube (nasogastric tube) through the nose and into the stomach to drain away any fluid and air that has collected
- Have rectal washouts, once or twice each day. A rectal washout involves passing a small tube (catheter) into your baby's bottom and up into their bowel. Further information is available in the Trust leaflet **Rectal washouts – a guide for parents of babies with Hirschsprung's Disease or bowel motility problems**. If you do not have a copy, please ask your nurse for a copy
- Have antibiotics if they have enterocolitis

Once the tummy is soft and your baby is passing poo regularly with the help of bowel wash outs, milk feeds can be restarted.

What happens if the rectal washouts are not successful?

Sometimes the rectal washouts are unsuccessful at fully clearing the bowel of poo, requiring the baby to need more than two washouts a day. In this situation the baby may need to have an additional operation to have a colostomy formed. A colostomy is an opening from the body (the bowel) to allow the poo to drain out.

A colostomy will also be required if the baby is very unwell with an infection, if there is a blockage in the bowel, or if there are other serious conditions.

What happens if my child does not receive treatment?

Are there any alternatives?

No. Surgery is needed, however the type of surgery undertaken will depend upon the age of the child and how well the child is at the time of diagnosis. 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. All of this will be discussed with you, and you will be given an outpatient appointment to come back to see the surgeon, who will then discuss future plans for surgery.

What happens if my baby does not receive treatment?

If HD is not treated, poo can fill the large intestines and block it causing problems, such as infection, bursting of the colon and in some cases can develop into life threatening enterocolitis.

Is there anything I can do to help my child?

We will teach you how to do the bowel washouts, and once you are confident with the procedure, you may take your baby home. We will organise the provision of the equipment needed to be delivered to your home, and we will attempt to provide you with some nursing support whilst at home. The same will happen if your baby has a colostomy. Once home if your baby has any of the symptoms of big tummy, fever, green vomits or your reluctance to feed, you should contact your GP or attend your local Emergency Department (A&E) immediately.

Useful sources of information

UK Hirschsprung & Motility Disorders Support Network

A support group of parents and adults supporting parents of kids with Hirschsprungs, IND and other motility bowel problems.

t: : 07935 787776 e: info@hirschsprungs.info / www.hirschsprungs.info

Contact us

If you have any questions or concerns about your baby and Hirschsprung's Disease, please contact the Neonatal Unit on 020 7188 4045

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.evelinalondon.nhs.uk/leaflets

Evelina London Medicines Helpline

If you have any questions or concerns about your child's medicines, please speak to the staff caring for them or contact our helpline.

t: 020 7188 3003 10am to 5pm, Monday to Friday e: letstalkmedicines@gstt.nhs.uk

Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.

t: 020 7188 8801 (PALS) e: pals@gstt.nhs.uk
t: 020 7188 3514 (complaints) e: complaints2@gstt.nhs.uk

Language Support Services

If you need an interpreter or information about your care in a different language or format, please get in touch:

t: 020 7188 8815 e: languagesupport@gstt.nhs.uk

NHS 111

Offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day.

t: 111

NHS Choices

Provides online information and guidance on all aspects of health and healthcare, to help you make choices about your health.

w: www.nhs.uk

Get involved and have your say: become a member of the Trust

Members of Guy's and St Thomas' NHS Foundation Trust contribute to the organisation on a voluntary basis. We count on them for feedback, local knowledge and support. Membership is free and it is up to you how much you get involved. To find out more, and to become a member:

t: 0800 731 0319 e: members@gstt.nhs.uk w: www.guysandstthomas.nhs.uk/membership



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