

# Hirschsprung's disease

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

## What is Hirschsprung's disease and why does your 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. This means that the poo cannot be pushed through the bowel as normal, so it is trapped in the large bowel causing it to be blocked and enlarged. This can lead to a bowel infection called enterocolitis. The amount of bowel that is affected can vary, but 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 are:

- big tummy
- repeated green vomit
- constipation
- baby may be reluctant to feed.

## Does your child need any tests to confirm the diagnosis?

Yes. A physical examination of your baby's abdomen (tummy) is needed, as well as a rectal examination which requires the doctor/nurse to put a finger into your baby's bottom to feel for any abnormalities. Your baby may also have other tests, such as an X-ray of their tummy, and some blood tests to check for any signs of infection (enterocolitis).

A rectal suction biopsy is also required which is necessary to test reliably for HD. This test takes place on the Neonatal Unit and involves taking a biopsy (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 examined under a microscope to confirm if there is an absence of ganglion cells, which would confirm the diagnosis of HD. The results can take several days.

## What treatments are available?

All children with HD will eventually need surgery, however the first 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. This involves passing a catheter (small tube) into your baby's bottom and up into their bowel. More information is available in our 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 one
- have antibiotics if they have enterocolitis.

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

## What happens if the rectal washouts are not successful?

Sometimes the rectal washouts do not fully clear the bowel of poo, meaning the baby will need more than 2 washouts a day. In this situation the baby may need to have an operation to have a stoma formed. A stoma is an opening in the abdomen which will allow the poo to drain out into a special bag.

A stoma may also be needed if the baby is very unwell with an infection, if there is a blockage in the bowel, or if there are other serious conditions. More information is available in the our leaflet, **Your child and their stoma**.

## What happens if your child does not receive treatment?

### Are there any alternatives?

**No.** Surgery is needed, but the type of surgery needed will depend upon the age of the child and how well they are at the time of diagnosis. There are a range of surgical treatments for HD depending on a number of factors such as how much 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 any plans for surgery.

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.

## What you can do to help your child

We will teach you how to do the bowel washouts and, once you are confident with the procedure, you can take your baby home. We will organise the provision of the equipment needed to be delivered to your home, and we will try to give you some nursing support at home. The same will happen if your baby has a stoma. Once home, if your baby develops any of the symptoms, of a big tummy, fever, green vomits or is reluctant to feed, you should contact your GP or go to your nearest 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, **tel:** : 07935 787776, **email:** info@hirschsprungs.info, **web:** www.hirschsprungs.info

### Contact us

If you have any questions or concerns about your baby and Hirschsprung's disease, please contact the Neonatal Unit, **tel:** 020 7188 4045 or the surgical CNS nurses, **tel:** 07598 552485.

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit **web:** 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.

**tel:** 020 7188 3003, Monday to Friday, 10am-5pm, **email:** letstalkmedicines@gstt.nhs.uk

### Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS), **tel:** 020 7188 8801, **email:** pals@gstt.nhs.uk. To make a complaint, contact the complaints department, **tel:** 020 7188 3514, **email:** complaints2@gstt.nhs.uk

### Language and accessible support services

If you need an interpreter or information about your care in a different language or format, please get in touch, **tel:** 020 7188 8815, **email:** languagesupport@gstt.nhs.uk

### NHS 111

This service offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day, **tel:** 111, **web:** www.111.nhs.uk

### NHS website

This website gives information and guidance on all aspects of health and healthcare, to help you take control of your health and wellbeing, **web:** 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, please get in touch, **tel:** 0800 731 0319, **email:** members@gstt.nhs.uk, **web:** www.guysandstthomas.nhs.uk/membership



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