

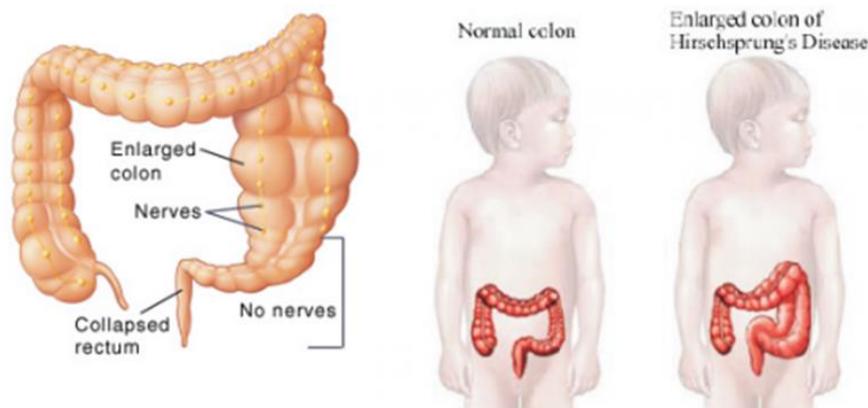
Ward 1C Neonatal Surgical Unit

## Hirschsprungs Disease

Information for Parents &amp; Carers

### What is Hirschsprungs Disease (HD)?

Healthy bowel action is important for us all. Hirschsprung's Disease is a condition in which poo (faeces / stool) is held up in the bowel and causes a blockage. In the UK one in 5000 suffer from this condition, which is diagnosed in babies, or in some cases older children. It is more common in boys than girls.



Picture 1

[https://www.networks.nhs.uk/nhs-networks/staffordshire-shropshire-and-black-country-newborn/documents/Hirschsprungs\\_Disease.pdf](https://www.networks.nhs.uk/nhs-networks/staffordshire-shropshire-and-black-country-newborn/documents/Hirschsprungs_Disease.pdf)

### Why does poo become stuck in the Bowels?

Like most of our body parts, nerves will be present in our bowels too. They lie between the layers of muscle tissue in the bowel. When these nerve cells (ganglionic) are present, the muscles in the bowel squeeze and relax. This action known as Peristalsis pushes the poo forward and along. When the nerves are absent (aganglionic) in some or the entire bowel, the poo becomes stuck. When this happens, your baby will show varying signs of discomfort. It is important to understand that HD is not caused by anything you may or may not have done during pregnancy.

### Can I Inherit HD?

It is very important for parents to understand that HD did not happen because of what they did or did not do in pregnancy. It is scientifically proven that HD happens in the womb as early as 12 weeks. We do not have a clear reason as to why the nerve cells are absent during development of your baby. Some studies suggest an alteration in one of the genes. There is also evidence that HD can run in families. This means that if you have had a child with HD before, you are more likely to have another child with it. HD can sometime be linked to other conditions such as Downs Syndrome and heart conditions. If you wish, you can have the opportunity to discuss future pregnancies with genetic specialists.

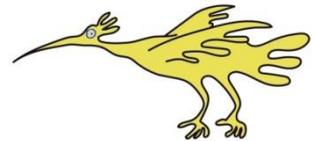
## Is Hirschprung's Serious?

Early identification of the problem is important in treating HD. It is not a serious condition if recognised and treated early on. Very rarely and if left undetected, it can cause a serious infection of the bowel known as enterocolitis.

## What are the Early Signs of HD?

Because poo is held up in the bowel, your baby can experience some or all of the following problems.

- Your baby may not be able to pass meconium (first black poo) within 24-48hrs of life or it may be less in amount and infrequent.
- Your baby may vomit which can be milky or be a green like fluid known as Bile.
- Your baby will be reluctant or slow to feed.
- Your Baby's tummy can become swollen/ distended (abdominal distension) and feel hard as he or she is unable to have his or her bowels opened.
- Your baby may show signs of pain and discomfort.



## How to Confirm HD?

HD can be suggested on x rays. Either an x ray of the abdomen or a contrast enema, which is fluid, introduced through the rectum via a thin tube, so the large colon (bowel) can be seen more clearly. Some sections of the bowel will look dilated and the section after this part will look narrower than usual (Picture1).

However, a more definitive test to diagnose HD will be a Suction Rectal Biopsy. This procedure takes place on the unit and using a lubricated instrument, which is passed into your baby's rectum (last part of the bowel) and three samples of tissue (biopsies) are taken. The samples are then sent to the laboratory and examined under the microscope, to see if ganglion cells are present. If there are no cells (aganglionic) then HD is present. It can take a couple of days or longer before the results are known.

## How can HD be Treated?

First line of treatment will be to decompress the tummy. Milk will usually be stopped and fluids will be given to keep your baby hydrated through a drip into a vein.

- A tube will be passed through your baby's nose or mouth down the esophagus (food pipe) (Picture 3). Through this tube, the stomach contents can be allowed to drain into a collecting bag attached to the end of the tube. The bag will be emptied using a syringe every 3-4hrs, thus helping to prevent any further vomiting. This tube will stay in place until no longer needed.



Picture 3: Baby with nasogastric tube in place and tube attached to drainage bag.

- To help move the poo along your baby's bowel, rectal/bowel washouts will be needed. These involve, passing a tube into your baby's bottom and warm saline fluid is pushed through the tube to help soften the poo/stools. When the stool is softened, it will pass down the tube. The tube is then removed. The frequency of these washouts will be decided by the surgeon. If this is an option, Nursing Staff will help teach you this procedure.
- Antibiotics will be required to treat and to prevent the possibility of severe infection.

Once a diagnosis of HD has been made, your surgeon will discuss with you the steps for surgery, depending if rectal washouts are successful or unsuccessful.

If the rectal washouts are successful, your baby will be discharged home, continuing with these until the date of surgery for a 'Pull Through Operation'. The 'Pull Through Operation' is performed via keyhole (laparoscopically) or 'open surgery' via an incision on the abdomen. The surgeon will discuss with you which method is appropriate. During this operation, the surgeon will identify the part of the bowel missing the ganglion cells, using a series of biopsies and remove the affected portion. The healthy part of the bowel will be attached to the anus to create a working bowel with enough nerve cells to control the muscles so that your baby/child can have a bowel action.



Baby with stoma and bag attached over stoma

If rectal washouts are not successful in removing the poo from obstructing the bowel, or your baby is too unwell at the time, then a temporary stoma will be created.

### At Home what to watch out for?

A complication of HD is enterocolitis which is severe inflammation and often infection of the bowel. Important signs of enterocolitis are:

- Explosive diarrhoea,
- Foul smelling stool (poo),
- Vomiting,
- Abdominal distension (bloated tummy) and
- Fever.

If these signs are seen, please seek urgent medical attention or call 999.

### What does the Future hold for my baby with HD?

It does depend on how much bowel has been affected/removed during surgery. In most cases of HD, the bowel will start to work properly after treatment. Initial management involves expert advice on diet and bowel training. The studies suggest that most children will enjoy a very good quality of life following surgery. Very few may need repeat surgery.

## Some of the Long-term Problems Include:

Constipation: This can be due to many reasons. One of the reasons for constipation could be due to the tightness around the anal opening. A procedure called anal dilatation (widening of the anal opening) can help with this situation. Doctors can discuss this with you if necessary. Your baby/child might need laxatives and a structured bowel programme.

- Some may have problems controlling their bowel movements until they are older. Support from the specialists for bowel training can be given in these circumstances.
- Poor growth and development: When large portions of the bowel is removed, children can have long-term problems with growth and development. This is because part of the lost bowel was where most of the digested food and nutrition gets into the blood. Poor Weight gain is a common occurrence in such situation. Specialists like dieticians will assist you with advice and support if this is the case with your child. There are support groups that can help following the diagnosis of your baby/child having HD. some of them mentioned below.

## Contact:

If you have any concerns or worries about your baby once you are home you can call the Neonatal Surgical Unit and one of our Nurses will be happy to give you advice and reassurance.

### Neonatal Unit 1C Yellow Pod

Tel:0151 252 5378

## Other Useful Contacts

There are support groups that can help following the diagnosis of your baby/child having HD. Some of them mentioned below:

**Bladder and Bowel UK:** Part of the charity Disabled Living, provides qualified impartial advice and information regarding continence issues, products and services for children and adults with bowel and/or bladder problems.

Website: [www.bladderandboweluk.co.uk](http://www.bladderandboweluk.co.uk)

Phone: 0161 214 4591

**Breakaway Foundation:** Breakaway are a UK wide charity offering support, holidays and activities to children from birth to eighteen, with bladder and/or bowel diversions/dysfunction and their families.

Website: <http://www.breakawayfoundation.org.uk/>

**Champs Appeal:** Raising Hirschsprung's Disease awareness CHAMPS is an appeal set up to raise awareness of children with Bowel and Bladder disorders, dysfunctions and diversions. They raise funds that will directly benefit children by supporting research, awareness campaigns and support groups.

Website: <https://www.raredisease.org.uk>

Email: [info@raredisease.org.uk](mailto:info@raredisease.org.uk)

Phone: 0300 124 0441

**Contact a Family:** Contact is a UK charity for families with disabled children. They offer information and support on specific conditions and rare disorders as well as advice regarding related issues such as benefits and finance.

Website: <http://www.contact.org.uk>

Contact a Family: National freephone helpline: 0808 808 3555 Monday-Friday, 9.30am-5pm

Manchester Office: St James House, Salford: 0161 743 0700

e-mail: [info@contact.org.uk](mailto:info@contact.org.uk)

## Space for the Parent/Carers notes

This leaflet only gives general information. You must always discuss the individual treatment of your child with the appropriate member of staff. Do not rely on this leaflet alone for information about your child's treatment.

This information can be made available in other languages and formats if requested.

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