The Royal Children's Hospital Melbourne

COLORECTAL AND PELVIC RECONSTRUCTION SERVICE

Hirschsprung Disease

Information for families

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Colorectal and Pelvic Reconstruction Service (CPRS)
Information for families

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Acknowledgements

We are indebted to the contributions of the many families that are cared for by the CPRS team. This resource is for all families affected by colorectal and pelvic conditions.

About this booklet

The Colorectal and Pelvic Reconstruction Service (CPRS) at The Royal Children's Hospital Melbourne (RCH) is leading the way in colorectal and pelvic care in Australia.

We aim to deliver the highest quality clinical care to children and families with colorectal and pelvic conditions. We play a vital role in increasing the awareness, understanding and knowledge of these conditions in the community, and work collaboratively to educate health care professionals.

This booklet has been developed to support parents, carers and children who have colorectal and pelvic conditions. The CPRS seeks to establish a healthy relationship with all families, as we believe this enables the best care possible. The content of this booklet has been developed based on the extensive clinical experience of the authors and the most recently published evidence for this clinical condition.

This CPRS booklet has been categorised into different stages of your child's journey, which allows you to read the information that is important to you at the time. Some parts may appear repetitive. This is because some of the information is relevant throughout different periods of your child's care.

Everyone learns differently. Some people like to read instructions, some like to learn by having information explained to them, and many like to do both. Make sure you tell the members of the CPRS team if you are finding any information in this booklet difficult to understand.

Hirschsprung disease

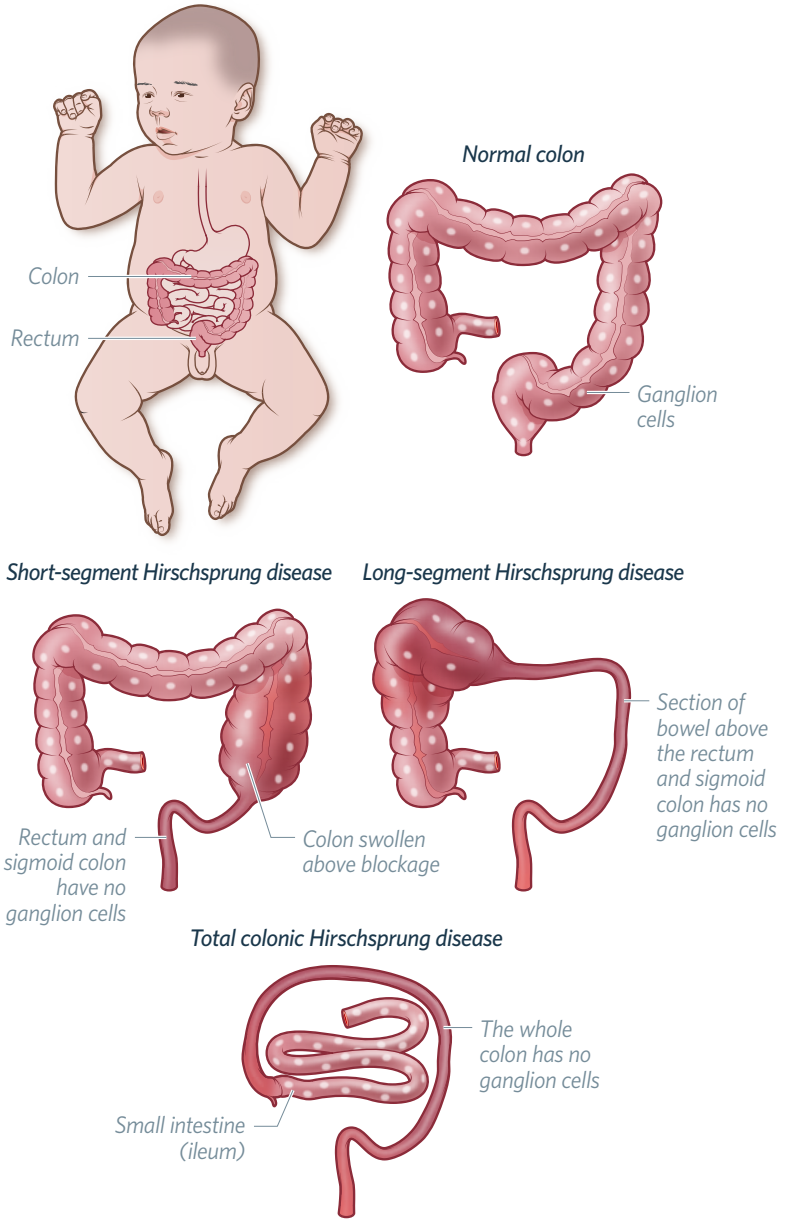
Hirschsprung disease is a congenital (born with) condition that occurs when the nerve cells in the lower part of the bowel do not form properly. These nerves are responsible for the wave-like motion that helps push food through the bowel, so that it may be digested and passed out of the body as stool (poo). If these nerves (ganglionic cells) are missing, it may cause partial or total blockage in the lower part of the bowel.

Hirschsprung disease occurs in one in 5000 live births. The disease occurs more often in males than in females, and sometimes may be associated with inherited conditions, including Down syndrome.

It is not a result of anything that the child's mother has done during pregnancy.

The levels of Hirschsprung disease include:

- **Short-segment** — ganglion cells are missing in the rectum and sigmoid colon (the last segment of the colon). The majority of children (80 – 85%) with Hirschsprung disease will have short-segment disease
- **Long-segment** — ganglion cells are missing in the section of the colon that is before the sigmoid colon and rectum
- **Total colonic** — ganglion cells are missing in the entire colon
- **Small bowel aganglionosis** — ganglion cells are missing in the entire colon, as well as a variable length of the small bowel



Hirschsprung disease — signs and symptoms

The initial signs and symptoms of Hirschsprung disease are most often seen within the first 24 to 48 hours of life.

The most common signs and symptoms in newborn babies include:

- Failure to pass meconium (a newborn's first stool) in the first 24–48 hours of life
- Swollen tummy
- Vomiting
- Temperature

Older children may have these signs and symptoms:

- Severe constipation
- Delayed growth (failure to thrive)
- Poor appetite
- Sepsis (a severe infection)
- Watery stools

Hirschsprung disease — diagnosis

The majority of children with Hirschsprung disease are diagnosed during the newborn stage, although some children are diagnosed later in life. Doctors perform several tests in order to confirm the diagnosis, and different tests may be used depending on your child's age.

These tests include:

- **Abdominal x-ray** — this test may show a bowel obstruction and is the first step. However, an abdominal x-ray will not provide an exact diagnosis
- **Suction rectal biopsy** — a small part of your child's bowel is biopsied, via the anus (bottom), in three sections to identify whether the nerve cells are present. The biopsies usually occur at your child's bedside
- **Full thickness rectal biopsy** — this is a surgical procedure and will occur in theatre if the initial biopsy is inconclusive or your child is too old to have a suction rectal biopsy (> three months)
- **Contrast enema** — this is a special type of x-ray, where contrast (dye) is inserted via the anus into the rectum to show the features of the bowel more clearly
- **Manometry** — these tests involve the placement of a catheter (tube) into the colon, rectum and anus. The pressure changes in the bowel are recorded to determine if the bowel is functioning normally. This is only performed in older children

**Please refer to Manometry booklet for more information*

- **Urine sodium** — this test measures the level of sodium (salt) in the body. It is important to monitor this regularly

Hirschsprung disease — treatment

Each child with Hirschsprung disease has unique needs. The CPRS team will make a treatment plan suited to your child's condition and their type of Hirschsprung disease.

The majority of children with Hirschsprung disease will go on to have healthy bowel function.

Bowel washouts (rectal)

Bowel washouts are the first step to remove trapped stool and gas. Nursing and medical staff will initially perform this procedure by inserting a tube into your child's anus and flushing small amounts of warm salty water.
This aims to clear the rectum of stool and gas and provide relief to your child. These washouts are done twice daily, or as per your surgeon's request, until surgery.

Bowel washouts are able to be performed at home, and you will be taught how to perform them prior to discharge home from the ward.

**Please refer to Bowel Washout (Rectal) booklet for more information*

Formation of stoma

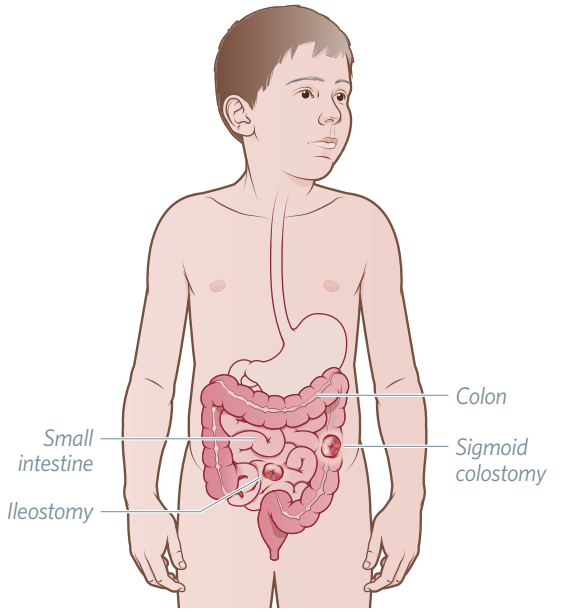
Colostomies and ileostomies are two different types of stomas. A stoma is an artificial opening which allows your child to pass stool to the outside of their body.

During an operation under general anaesthesia, the surgeon will bring the healthy end of the bowel to an artificial opening in your child's abdominal wall.
This means that stool will be pushed through the bowel to the stoma, where it is collected into a bag and disposed of. This gives the inflamed areas of the bowel time to heal and allows your child to grow prior to their definitive pull-through procedure (see information below on pull-through procedure).

The stoma is nearly always a temporary measure.

**Please refer to Colostomies and Ileostomies booklet for more information*

Colostomy and ileostomy



Pull-through procedure

The surgery for Hirschsprung disease is known as a pull-through procedure.

There are three main techniques for the pull-through procedure (Swenson, Soave, Duhamel). All three techniques may be performed either laparoscopically (keyhole) or via laparotomy (open surgery through a larger incision in the abdominal wall). The surgeon will discuss which method is most appropriate for your child.

During the pull-through procedure, your child's surgeon will remove small samples of tissue (biopsy) from your child's bowel to identify the area of the bowel where the ganglion cells are present.

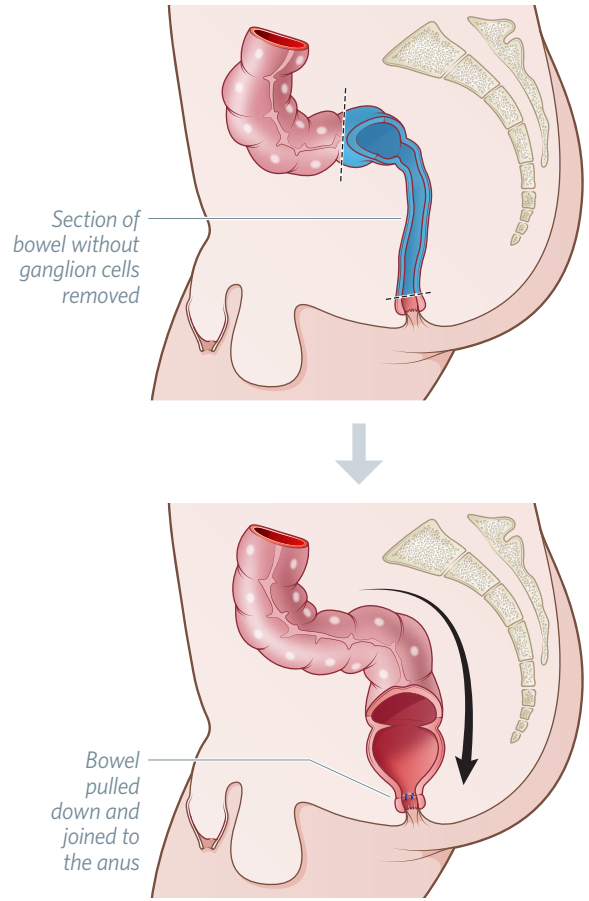
Once the correct area of bowel is identified, the pull-through procedure may begin. Your child's surgeon will remove the affected segment of bowel where no cells are present (the aganglionic section) and then connect the segment where cells are present (ganglionated section) to the anus.

Following the pull-through procedure your child will remain in hospital for five to seven days. This is to ensure that they recover from the surgery, their pain is well managed, their bowels are working, and they are able to eat and drink prior to going home.

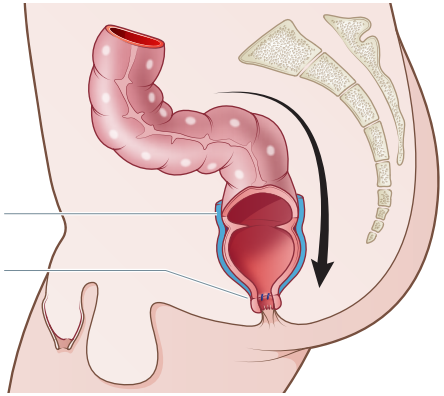
During your child's admission, members of the CPRS and inpatient team will support you in caring for your child.

**Please refer to Theatre booklet for more information*

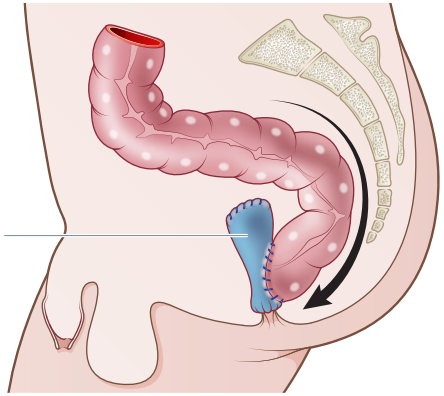
Swenson procedure



Soave procedure



Duhamel procedure



Sodium supplementation

Some children, particularly those whose entire colon is affected by Hirschsprung disease, may need an ileostomy for many months or, sometimes, years. The ileostomy may cause the body to lose sodium, which makes it difficult for your child to gain weight. For that reason, if your child has an ileostomy they may be prescribed sodium supplements by the CPRS team.

Anal dilatations

After surgery, the body has a natural tendency to close the surgical wound as part of the healing process. Care of your child after their pull-through procedure includes a treatment to prevent the bowel wound from closing. This is known as anal dilatation and may be required to keep the newly formed join in the bowel open to the right size. Dilatation involves placing a metal dilator into your child's anus once to twice per day. The dilator will slowly stretch the bowel wound to the correct size for your child's age.

**Please refer to Anal Dilatations booklet for more information*

Closure of stoma

The stoma is usually closed three to four months after the pull-through procedure. For the closure of stoma surgery, your surgeon will reconnect the two sections of bowel and close the hole in the abdominal wall. After the stoma is closed, your child won't be able to eat for 48 hours. This allows the new connection between the two parts of the bowel time to heal. Children usually start passing stool within two to three days, and most children may go home shortly after they begin passing stool. Your child will be in hospital five to seven days after their closure of stoma surgery.

**Please refer to Colostomies and Ileostomies booklet for more information*

Ongoing cares

Hirschsprung associated enterocolitis

Children diagnosed with Hirschsprung disease are at risk of developing an infection known as Hirschsprung associated enterocolitis (HAEC).

This infection leads to inflammation of the large bowel, and may cause your child to become unwell quickly. It is important to know the signs and symptoms of HAEC, and the care associated with this condition.

**Please refer to Hirschsprung Associated Enterocolitis booklet for more information*

Buttock care

Buttock care to prevent nappy rash is important. Nappy rash is a dermatitis confined to the area covered by the nappy. Nappy rash is not caused by one thing, but may be due to a range of problems. The most common cause of nappy rash (sometimes called diaper dermatitis) in children is irritation. The CPRS team will both educate and support you in the prevention and treatment of nappy rash.

**Please refer to Buttock Care booklet for more information*

Toilet training

A common question asked by families is when will be the right time to toilet train their child? Children with Hirschsprung disease are often delayed in their toilet training by six to 12 months. It is important to get the bowel in the best shape possible prior to commencing toilet training, and the CPRS team will work with you and your child to do that. It is important to remember that there is no rush to toilet train.

**Please refer to Toilet Training booklet for more information*

Bowel management

Children with Hirschsprung disease may suffer from different levels of constipation, diarrhoea and/or incontinence. Bowel management is a planned way to clean the large bowel of stool regularly. It helps your child stay clean, avoid accidents and wear regular underwear.

Bowel management may include the use of diet, medicines, laxatives and/or enemas to slow down or speed up the bowel.

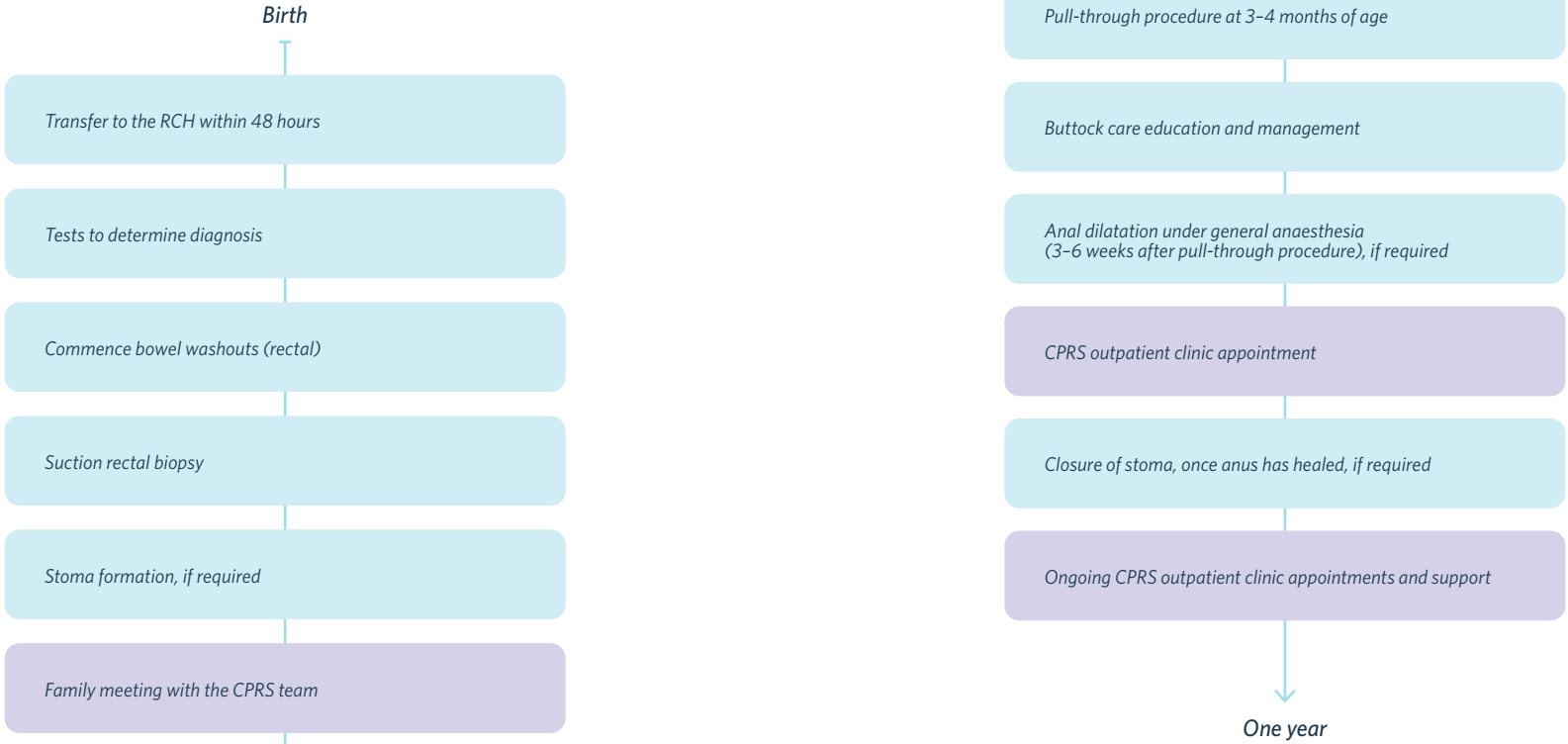
The goal of the CPRS Bowel Management Program is for your child to have a daily bowel movement and remain clean in-between.

**Please refer to Bowel Management booklet for more information*

Hirschsprung disease — pathway

The first year of a child's life with Hirschsprung disease may seem very overwhelming. The CPRS team is here to support your child and family.

Below is a pathway of what you may expect in the first year of your child's life, involving diagnosis, tests, imaging, surgeries, clinic appointments, phone calls and ongoing cares.



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