



COLORECTAL AND PELVIC
RECONSTRUCTION SERVICE

Anorectal Malformations

Information for families

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Colorectal and Pelvic Reconstruction Service (CPRS)
Information for families

Design, photography and medical illustrations by
The Royal Children's Hospital Melbourne

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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.

Anorectal malformations

An Anorectal malformation (ARM) is a congenital (born with) condition that affects the development of the rectum and anus early in pregnancy while the child is still developing. It may also be referred to as an imperforate anus (IA). ARM occurs in approximately one in 5000 live births, and is slightly more common in males than females. The cause of ARM is currently unknown.

Some children diagnosed with an ARM will have related anomalies. This is known as VACTERL association. Each letter of the word VACTERL represents an area where the anomalies may be. This means that VACTERL is not so much a disorder, but a collection of potential issues that may be associated with one another.

V—Vertebral anomalies

A—Anorectal malformation

C—Cardiac anomalies

T—Tracheo-oesophageal fistula and oesophageal disorders

R—Renal anomalies

L—Limb anomalies

Children with an ARM will range from mild to complex cases. In some of the more difficult cases, there may be a fistula between the rectum and the urinary (renal) system.

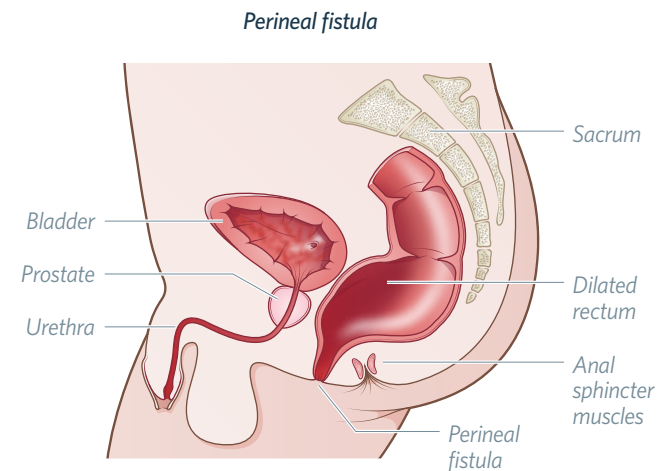
A fistula is an abnormal connection or passageway that joins two structures that do not usually connect. A fistula may develop anywhere in the body, including between the bowel and the skin, or between the vagina and the rectum.

**Please refer to the Typical Bowel Anatomy booklet for more information*

Anorectal malformations — male

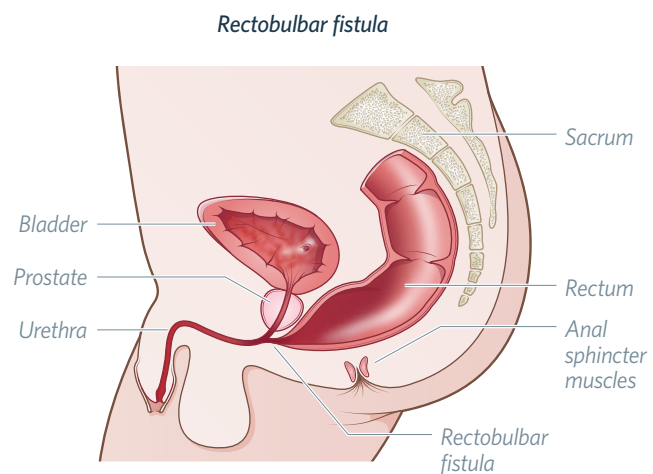
Perineal fistula

In this type of ARM, the anal opening is slightly closer to the scrotum than it should be. This may be obvious at birth, or it may only be diagnosed when the child is older and develops constipation.



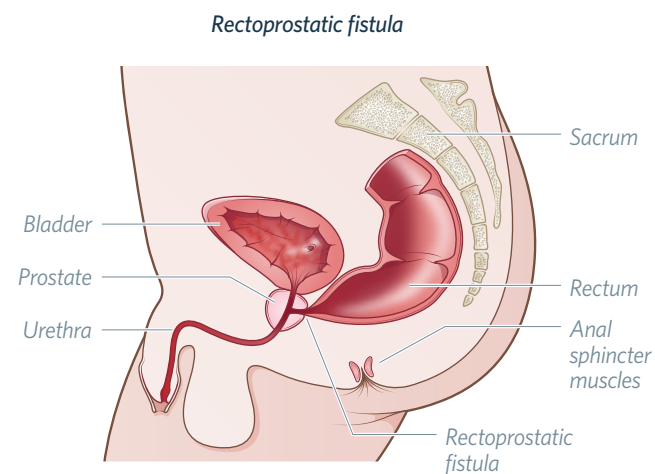
Rectobulbar fistula

This is the most common type of ARM in males. The rectum ends in a fistula that joins the urethra (urinary tube). There is no anal opening in the normal position on the skin. It is usually obvious at birth that the anus is not present.



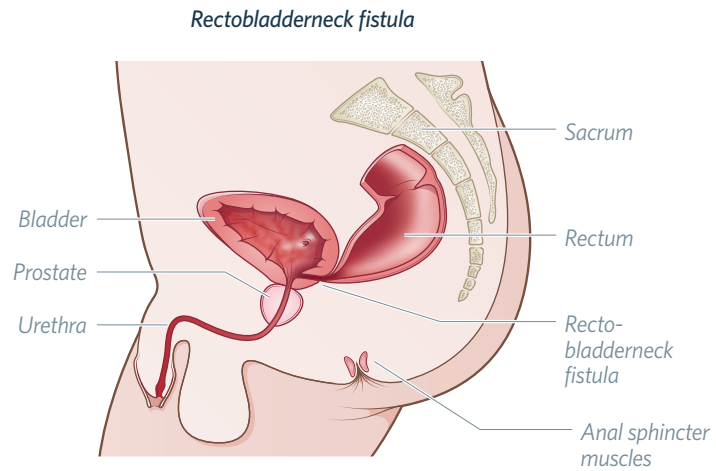
Rectoprostatic fistula

In this form of ARM, the rectum ends in a fistula that joins the urethra (urinary tube) as it passes through the prostate. There is no anal opening in the normal position on the skin. It is usually obvious at birth that the anus is not present.



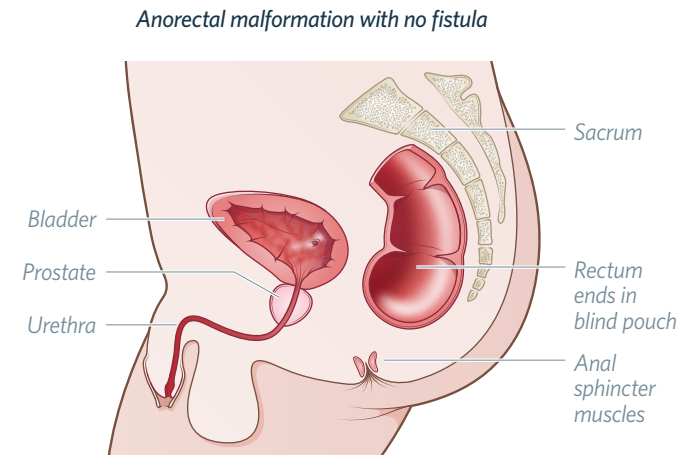
Rectobladderneck fistula

This is a rare and more complex form of ARM. The rectum ends in a fistula that joins the bladder (organ where urine is stored). There is no anal opening in the normal position on the skin. It is usually obvious at birth that the anus is not present.



Anorectal malformation with no fistula

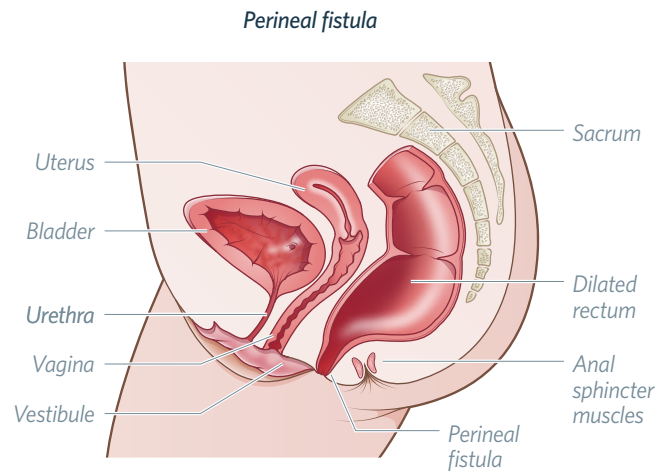
The rectum ends in a blind pouch and does not connect to anything. There is no anal opening in the normal position on the skin. It is usually obvious at birth that the anus is not present.



Anorectal malformations — female

Perineal fistula

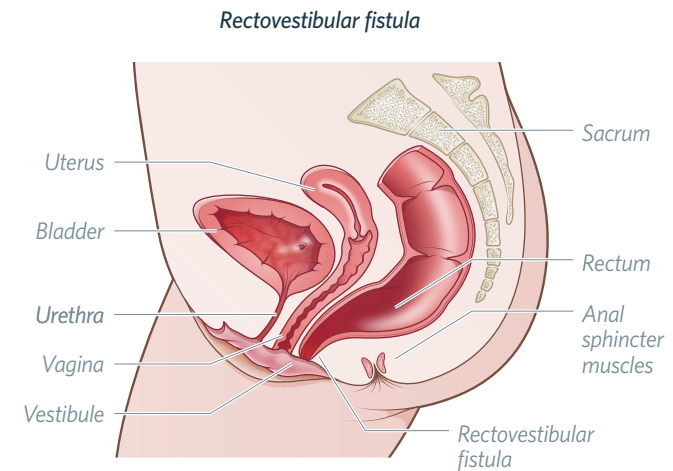
In this type of ARM, the anal opening is slightly closer to the vagina than it should be. This may be obvious at birth, or it may only be diagnosed when the child is older and develops constipation.



Rectovestibular fistula

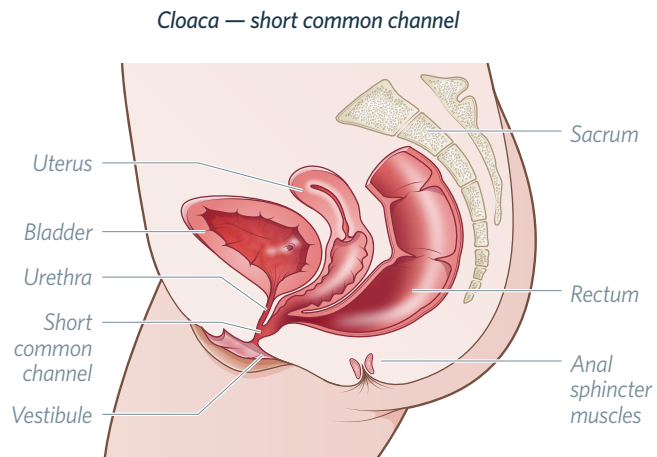
This is the most common type of ARM in girls. In this type of ARM, the anal opening is just behind the vagina, within the vestibule. The vestibule is the space in the vulva into which both the urethra and vagina open.

A rectovestibular fistula is usually diagnosed when the anus is not in the normal position or when stool (poo) is seen coming out of an area next to the vagina. This is normally diagnosed at birth but sometimes may not be diagnosed until the child is several months old. There may be associated anomalies in the genital tract.



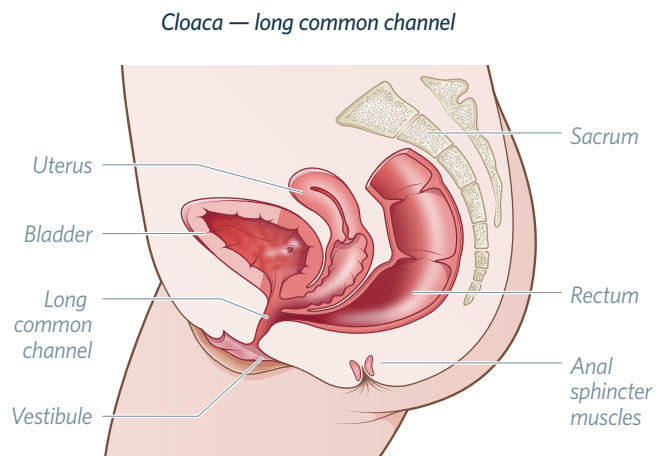
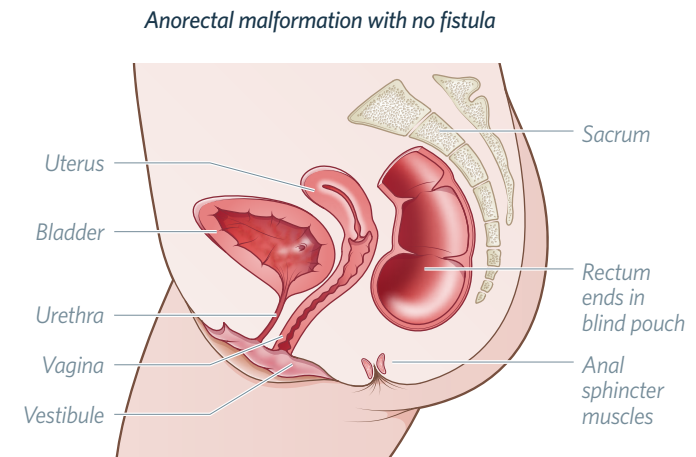
Cloaca

This is the most complex type of ARM in girls, and occurs when the three tubes (urethra, vagina and rectum) are all joined together in a common channel. The length of the common channel may be short or long, and the treatment is different for each child.



Anorectal malformation with no fistula

The rectum ends in a blind pouch and does not connect to anything. There is no anal opening in the normal position on the skin. It is usually obvious at birth that the anus is not present.



Anorectal malformations — signs and symptoms

An ARM is most commonly diagnosed soon after birth. It is rare for children to be diagnosed with an ARM during the prenatal period.

Common signs and symptoms of an ARM include:

- Anus not present, not open, or in the incorrect position
- Failure to pass meconium (a newborn's first stool)
- Stool passing through the penis or vagina
- Swollen tummy
- Vomiting
- Born with associated anomalies (VACTERL)

Anorectal malformations — diagnosis

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

Tests that may be used to diagnose an ARM include:

- **Abdominal x-ray** — this will show how far the rectum reaches and if there are any problems with the way the lower part of the spine has developed
- **Renal ultrasound** — this will show if there are any anomalies in the kidney, bladder and urinary tract
- **Pelvic ultrasound** — this will show if there are any anomalies in the reproductive organs in girls
- **Spinal ultrasound** — this is used to assess the spine for a tethered cord, and to determine the number of segments in the sacrum
- **Magnetic resonance imaging (MRI)** — this is sometimes needed to make a definitive diagnosis of a tethered cord or other spinal anomalies. An MRI may also be used to view the pelvic muscles and reproductive organs
- **Echocardiogram (Echo)** — this is used to identify heart anomalies

The CPRS team will explain why each test is being done. The results will be discussed with you, and you will have an opportunity to view the images if you wish.

Anorectal malformations — treatment

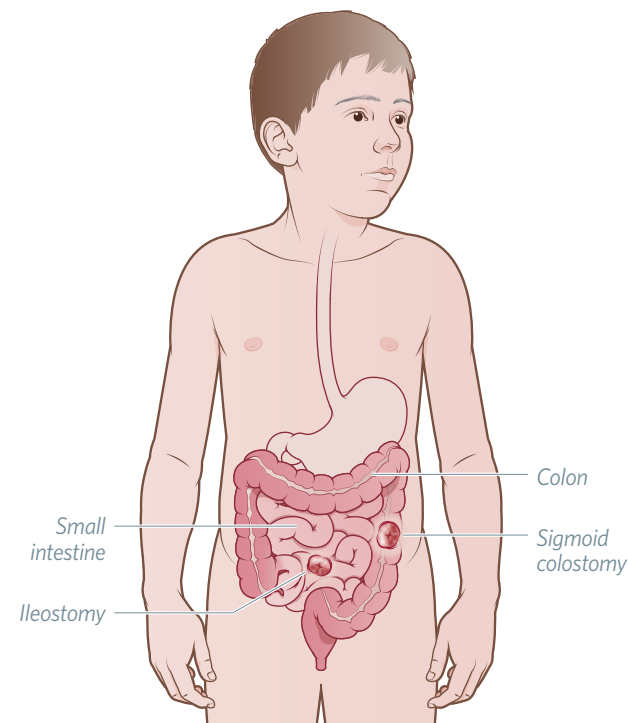
Each child with an ARM will require an individual treatment plan. The CPRS team will create a plan that is appropriate to your child's type of ARM, the presence of associated anomalies, and their overall health. The majority of children with an ARM will require surgery. The surgeries that are most common for ARM are listed below.

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 body a safe passage to pass stool and allows your child time to grow prior to their PSARP/LAARP surgery. The stoma is nearly always a temporary measure.

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

Colostomy and ileostomy



Posterior Sagittal Anorectoplasty (PSARP)/Laparoscopically Assisted Anorectoplasty (LAARP)

The PSARP is performed with your child under general anaesthesia. During this surgery, your child will be placed face down on the operating table. The surgeon will make an incision down the midline of your child's bottom, between the cheeks.

The surgeon will then separate the layers of muscle, fat and connective tissue in the area until they locate the rectum. Your child's surgeon will lift the rectum up and away from the urinary tract (in boys) or vagina (in girls), and then surgically position it within the anal sphincter muscles to create an opening.

During the operation, the surgeon will also ensure there are no vaginal anomalies (in girls) or urological anatomical issues (in girls and boys).

The LAARP is performed with your child under general anaesthesia. During this surgery, your child will be placed on the operating table, and will undergo laparoscopic (keyhole) surgery. The rectum will be separated from the surrounding structures, and will then be surgically positioned within the anal sphincter muscles to create an opening. During the operation, the surgeon will also ensure there are no vaginal anomalies (in girls) or urological anatomical issues (in girls and boys).

After the PSARP/LAARP your child will be admitted to hospital for seven to 10 days. This is to ensure that they have recovered from the surgery, have their pain well managed and are able to drink prior to going home. During their admission the members of the CPRS and inpatient team will support you in caring for your child.

**Please refer to Theatre booklet for more information*

Anal dilatation

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 PSARP/LAARP includes a treatment to prevent the new anal wound 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 their anus to the correct size for their age.

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

Closure of stoma

The stoma is usually closed three to four months after the PSARP/LAARP. For the procedure, the 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 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 for five to seven days after their closure of stoma.

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

Ongoing cares

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

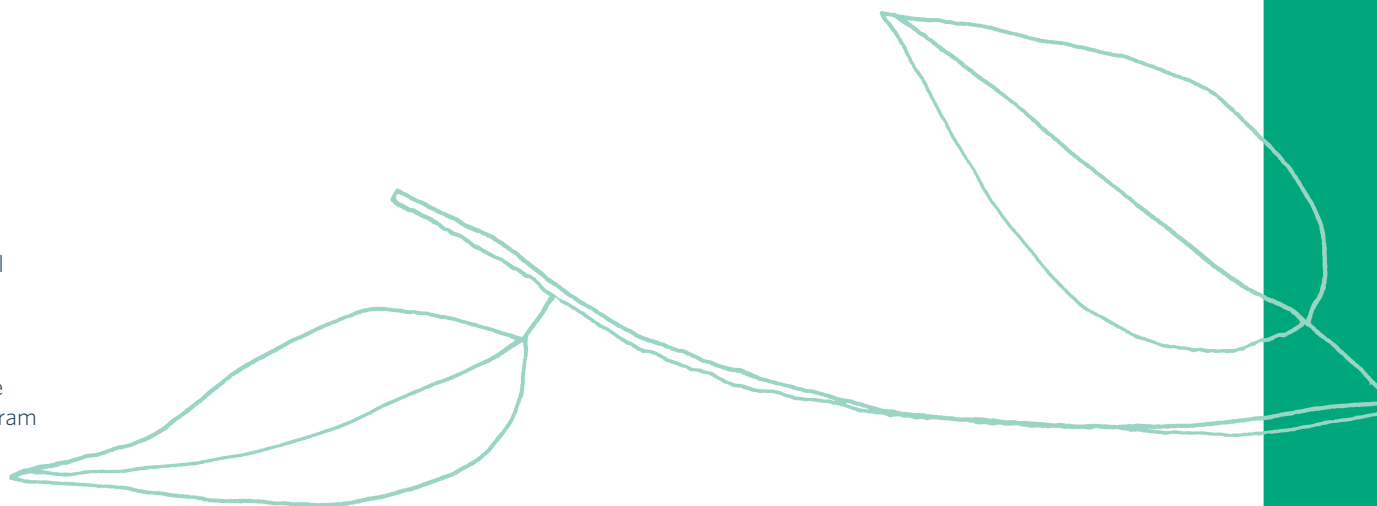
A common question asked by families is when will be the right time to toilet train their child? Children with an ARM are often delayed in their toilet training by six to 12 months. It is important to get the bowel in the best shape possible prior to commencing toilet training, and the CPRS team will 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 an ARM 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*



Anorectal malformations — pathway

The first year of a child's life with an ARM 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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