ARM

Anorectal Malformations *Family Guidebook*

A Guide For A Lifetime of Care

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Mariam Hobeldin: Designer, Medical Illustrator and Linguistic Editor

Nationwide Children's®

When your child needs a hospital, everything matters.

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This book is dedicated to the life and passion of

Brenda Ruth, BSN, CWON.

5/26/1960- 7/18/2021

Brenda Ruth started her nursing career at Nationwide Children's Hospital in 1981 in the Surgical ICU as a floor nurse. She quickly became a nursing care coordinator and for a brief time from 1988-1992 even acted as a Nurse Manager. She soon missed direct patient care and went back to hands on, bedside care.

In 1995 she shifted her focus and became board certified as an Enterostomal Therapist. Today, this is known as a Wound, Ostomy and Continence Nurse. Brenda was the only clinician for the entire hospital for many years. She independently coordinated the hospital's first Pressure Ulcer Prevalence survey in 2002 and completed ostomy education to new ostomates and their families. She also coordinated wound care for patients throughout the hospital. She was instrumental in creating protocols during the early stages of Nationwide Children's Colorectal Program development and continued to work side by side with the team to ensure families and staff received proper education. She was a content expert for national organizations such as Solutions for Patient Safety and wrote numerous articles on best practices for wound and ostomy care.

Brenda essentially grew the program into what it is today. We have a certified, 5-person team dedicated to both the inpatient and outpatient care of this unique population. Brenda instilled into our team what it means to be a hard-working, inquisitive, patient advocate and to not be afraid to stand up and do the right thing. She was a strong leader, a direct communicator and enjoyed teaching others, while continuously learning herself. She had a kind and gentle approach when teaching and inspired so many staff and families throughout her career. She loved her patients and coworkers as if they were her own family and had an infectious laugh that could be heard throughout the hospital.

Brenda passed away from Glioblastoma July of 2021 while we were working on this special Anorectal Malformations Family Guide. She was a key contributor to several of the chapters.

Brenda was one of the pillars of Nationwide Children's Hospital, but she would have told you more importantly she was a wife, mother, Mimi, daughter, sister, friend, and coworker. The world was a better place because of her and it will not be the same without her.

Thank you Brenda for inspiring us all to selflessly give and be the best we can be each day.

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1. Letter to Parents

Dear Parents,

Congratulations on the birth of your child! This excitement is lost, and you are shocked when you are told that your child has an Anorectal Malformation.

Anorectal Malformations are often not picked up on the scans you had while pregnant. So, you probably were told about this condition after your child's birth. It is likely you never knew that a child could be born with an abnormal anus. This shock could be more intense because of how you were given the news.

Panic and the overwhelming feeling of things being out of control replace feelings of joy and excitement about your new baby. Often, news of this diagnosis is followed by further information about other abnormalities. Sometimes this information results in you becoming confused and very concerned. Not knowing the reasons for anorectal malformations does not make it any easier. These feelings are understandable and expected, and we are here to help you through this.

We realize that there are very few resources written for you, your child, and other caregivers. This book was created with the guidance of parents and patients who have been through this journey. Our goal is to give you back control by increasing your knowledge and understanding of the condition, the surgeries, and the short- and long-term care associated with this condition.

We hope that this book will be your guide on this new journey and would love to hear from you. Please feel free to reach out to us with any comments you may have so that we may address them.

Kind regards,

Ihab & Richard

Ihab Halaweish, MD,

is a pediatric surgeon at Nationwide Children's Hospital in the Division of Pediatric Surgery and Center for Colorectal & Pelvic Reconstructive Surgery. He is also an assistant professor of Pediatric Surgery at The Ohio State University College of Medicine. Dr. Halaweish's practice is focused on the care of neonates, infants and adolescents with Hirschsprung disease and anorectal malformations including cloacal malformations.

Dr. Halaweish has a special interest in creating family-centered surgical literature to help families navigate complex medical diagnoses.

Richard J. Wood, MD,

serves as the Chief of Colorectal and Pelvic Reconstructive Surgery at Nationwide Children's Hospital. He is a Professor of Surgery at The Ohio State University College of Medicine. Dr. Wood trained in South Africa, England and the United States and is Fellowship trained in General Pediatric Surgery as well as Pediatric Colorectal Surgery. He has a special interest in the reconstructive surgery for complex anorectal malformations, cloacal malformation, cloacal exstrophy, and Hirschsprung disease.

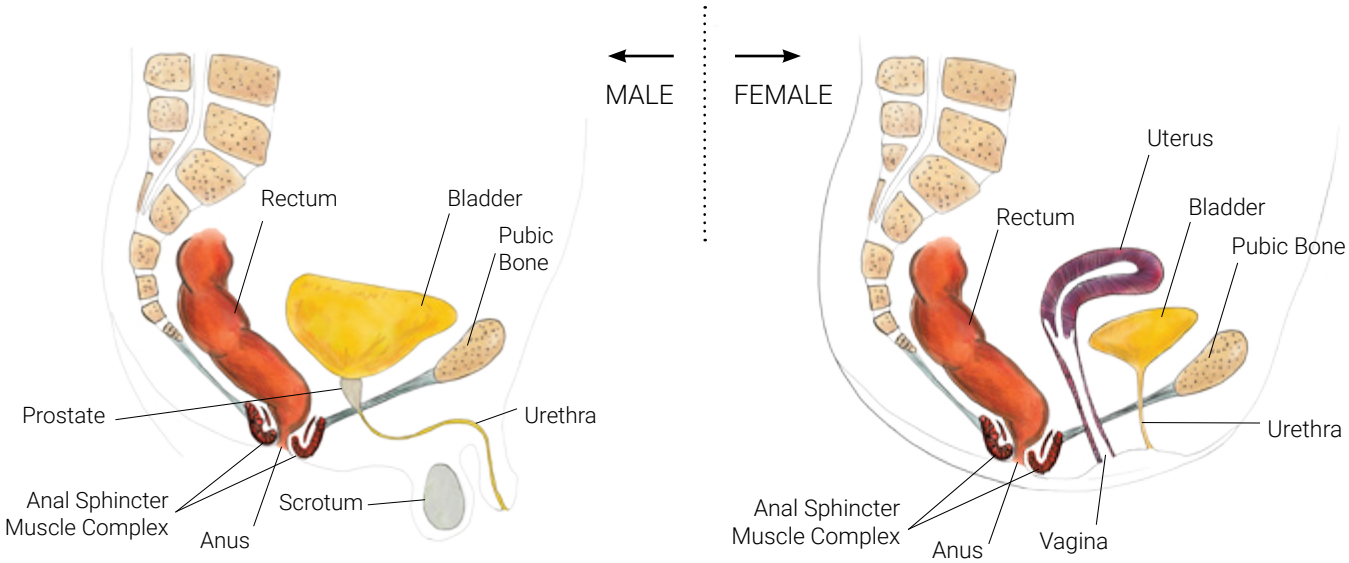
Dr. Wood's research work in anorectal and cloacal malformations focuses on complex reconstruction techniques, outcomes, quality of life and advocacy. He serves as a medical advisor for Pull-Thru Network and on the Board of REACH.

2. What are ARMS

What are anorectal malformations?

Anorectal malformations (ARMs) are a group of defects that happen early in pregnancy while a baby is still developing. The last part of the colon, called the rectum, is just above the opening, the anus, through which the poop passes. Both parts of the bowel did not develop correctly before birth.

The terms used to describe ARMs can be confusing because not all providers and resources use the same names for these conditions. ARMs is an umbrella term that describes different conditions. We will cover these conditions in the following chapters.



How common are ARMs?

About 1 in 5,000 babies will be born with an ARM. It is slightly more common in males than in females.

Why do babies get ARMs?

No one knows why some babies are born this way, but there is a family history in some cases. The genetics of ARM are discussed in Chapter 6.

What are the different types of ARMs?

First, let's discuss some medical terms to make this easier.
Please see the diagrams in this chapter.

Fistula:	This is an abnormal connection between two different organs that should not be connected.

Urethra:	This is the tube that carries urine from the bladder to the outside of the body.

The next few pages have an overview on the different types of ARMs

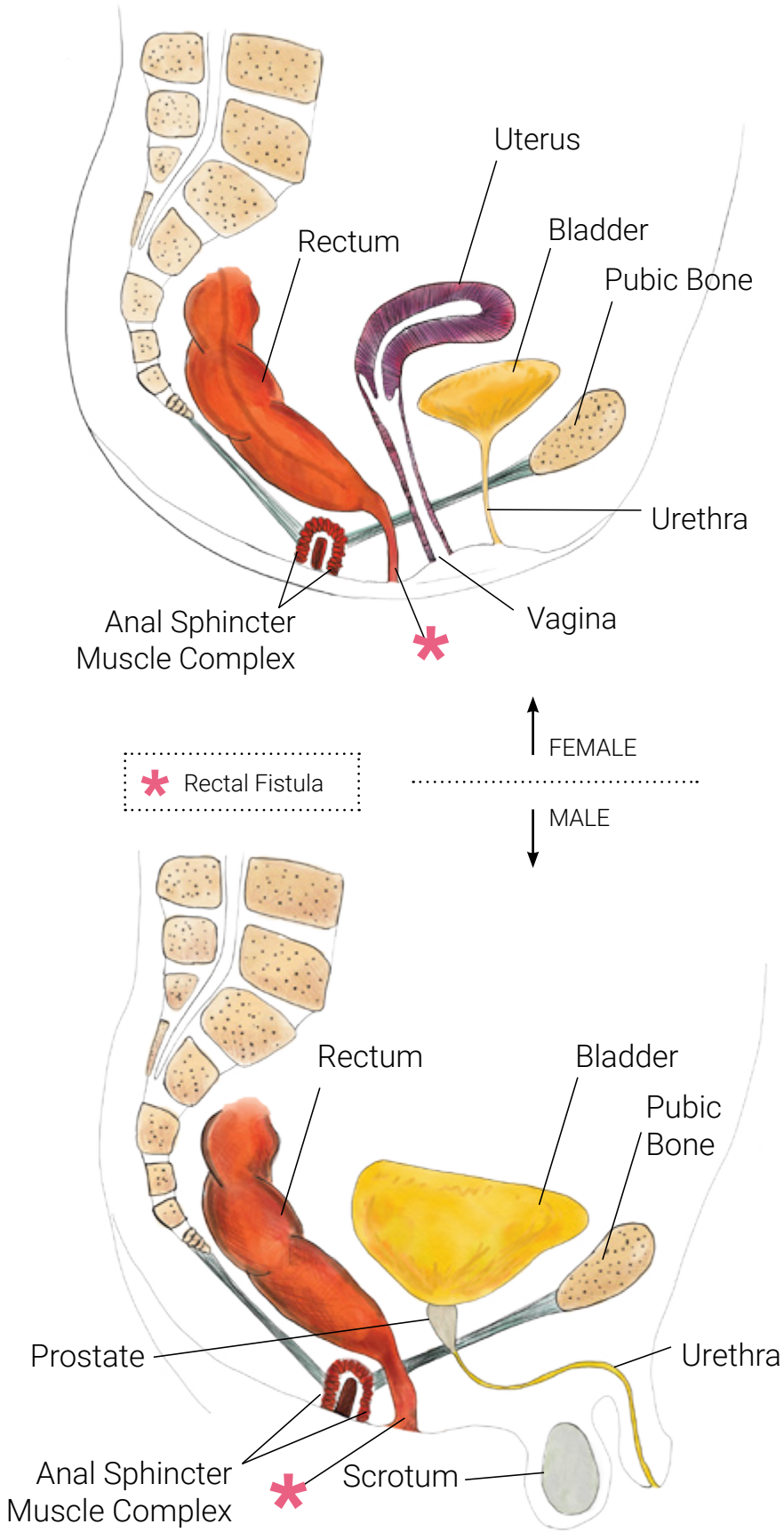
BOYS and GIRLS | Perineal Fistula

In a perineal fistula, the opening of the anus is not in the right place. It is outside the ring of muscle or sphincter that keeps the anus closed. Usually, the opening is too small.

A perineal fistula may be diagnosed at birth. Poop will come out from this abnormal location. It can sometimes take a day or two for your baby's poop to drain out of this opening. The fistula opening is usually too small and needs to be opened or dilated (see Chapter 8) over time until your child has surgery (see Chapter 10 & Chapter 11).

In some cases, the perineal fistula is not found at birth because the abnormal opening is big enough to allow poop to pass. Your child can empty their bowels, especially if they take breast milk which keeps the poop liquid and soft. However, as your child grows, they may begin to have constipation and problems passing poop.

Sometimes your pediatrician will notice an opening that is not in the right place and is too small during your baby's checkups, which leads to a diagnosis.



BOYS ONLY

In ARMs in boys, the rectum may not connect with the anal opening and instead, the rectum connects in the bladder or in the urethra, which is the tube that carries urine from the bladder through the penis. In rare cases, the rectum has no opening and ends in a blind pouch.

Usually, ARMs with no anal opening are discovered at the time of birth in boys. In some cases, there may be signs that the baby will develop an opening outside the normal anus (perineal fistula) and your doctors may chose to wait until your baby is one or two days old for this to progress.

If there is truly no opening after one to two days, your child will need a colostomy to allow for stool to come out. (See Chapter 7).

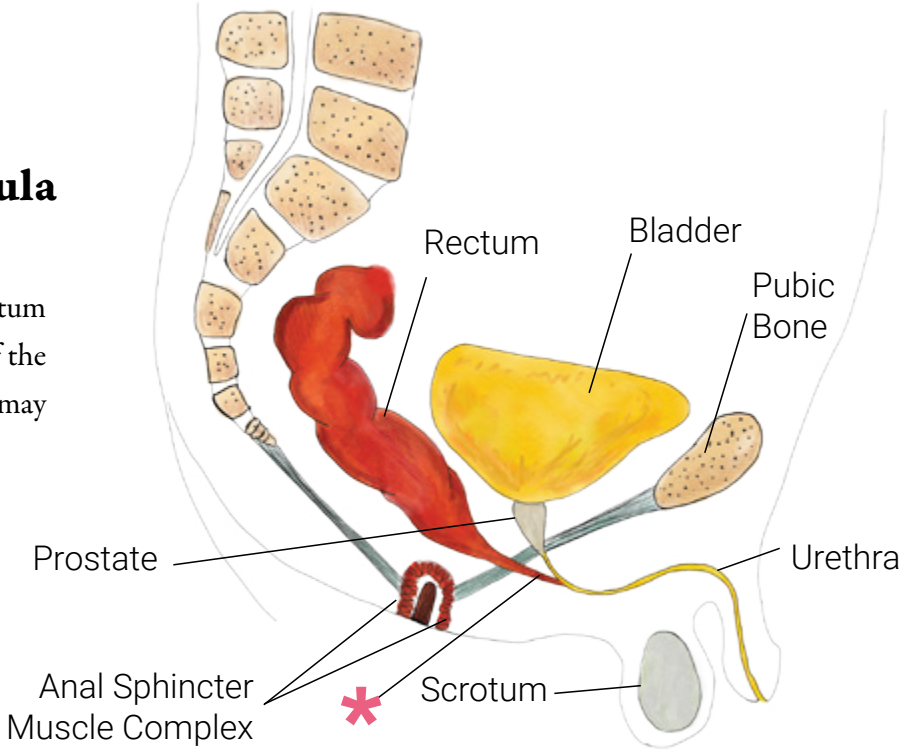
ARM types are named depending on what part of the urinary system the rectum connects to.

Surgery to repair these different types of ARMs is covered in Chapter 10.

Sometimes a sign that the child has an ARM, where the rectum connects to the urinary system is the presence of meconium, or newborn stool, in the urine.

Rectobulbar Fistula

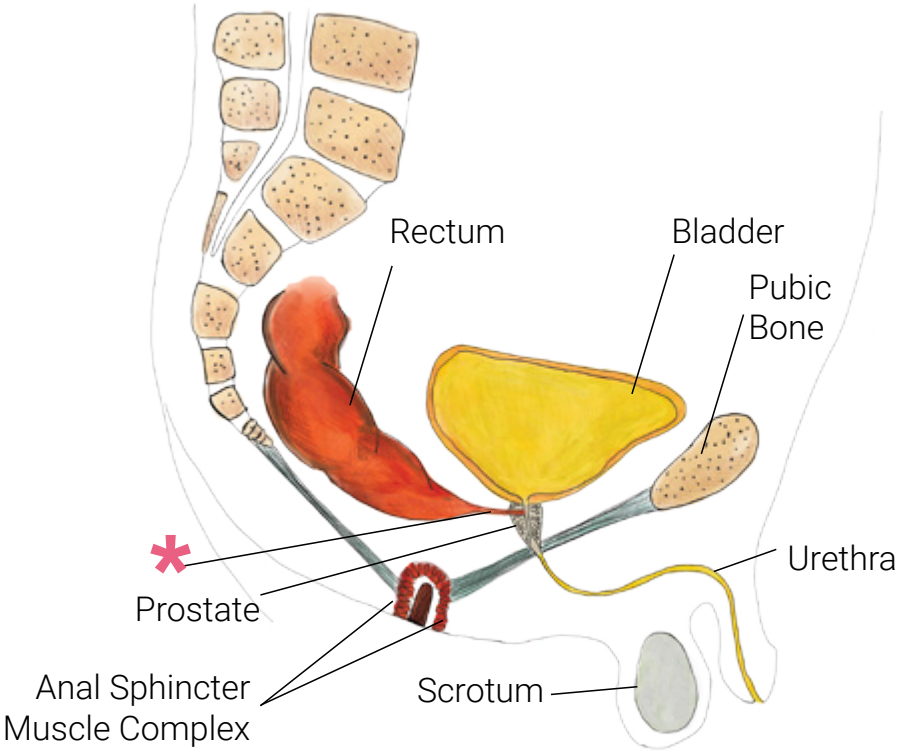
In this type of ARM, the rectum connects to the lower part of the urethra. The medical team may refer to it as a "low" ARM.



Rectoprostatic Fistula

The rectum connects to the upper part of the urethra near the prostate in this type of ARM.

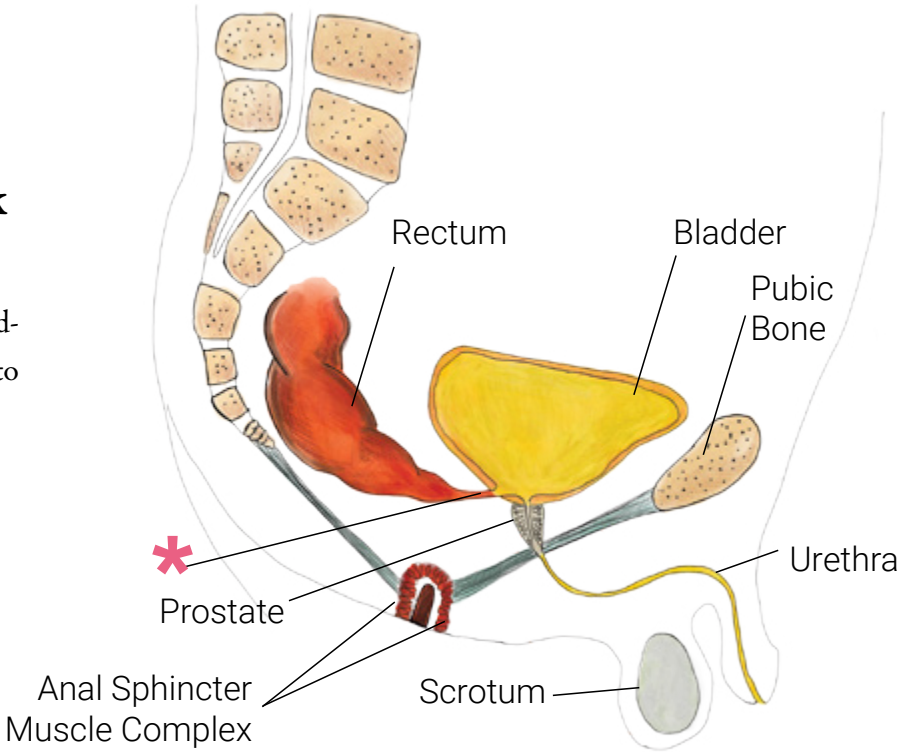
The medical team may refer to it as a "high" ARM.



* Rectal Fistula

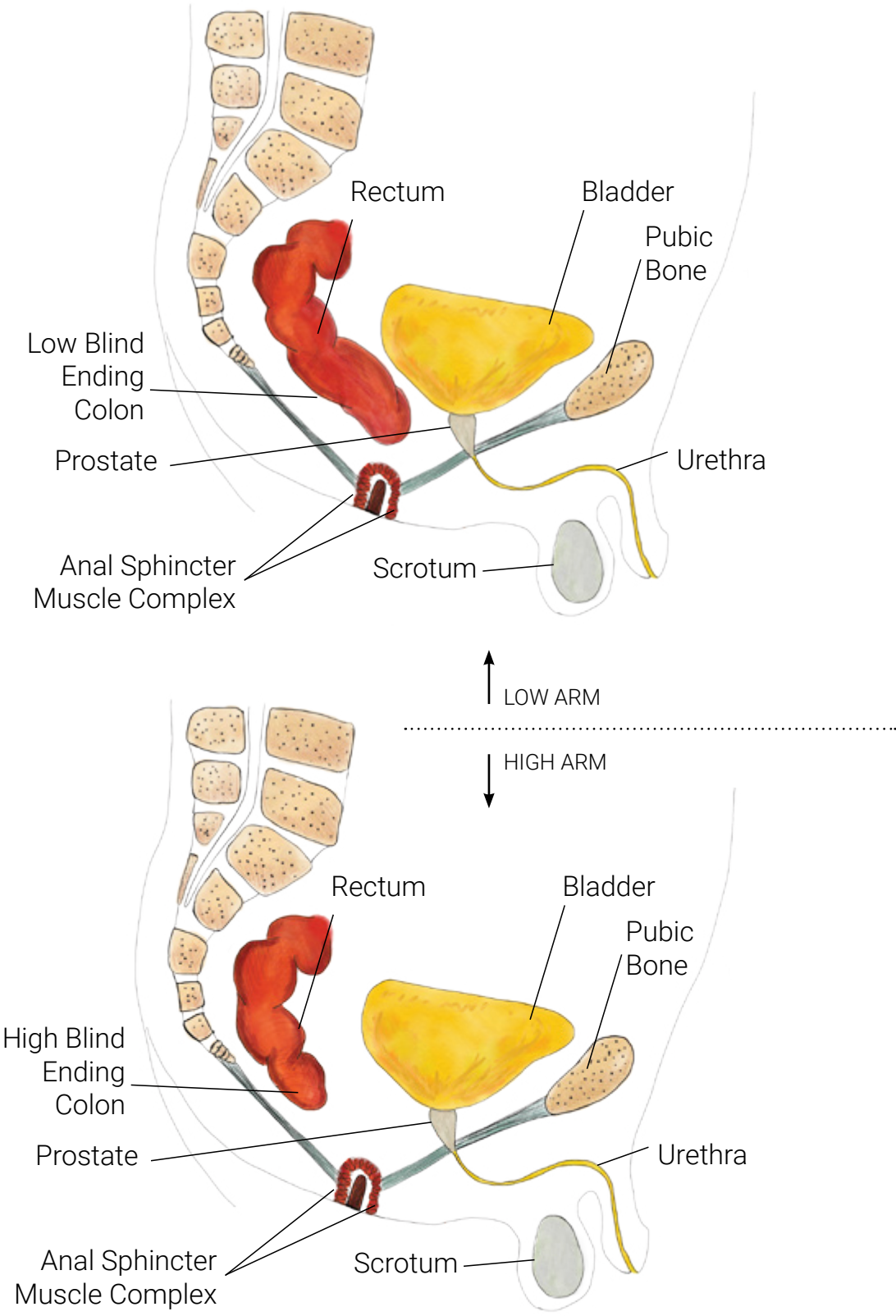
Rectobladder Neck Fistula

The rectum connects to the bladder itself and may be referred to as "high" ARM.



Anorectal Malformation without Fistula

In this type of ARM, the rectum does not connect with any other organ and ends in a pouch. Depending on the location of the pouch, the medical team may refer to it as "low" or "high" ARM. This type of ARM without a fistula is seen more often in babies with Down Syndrome.

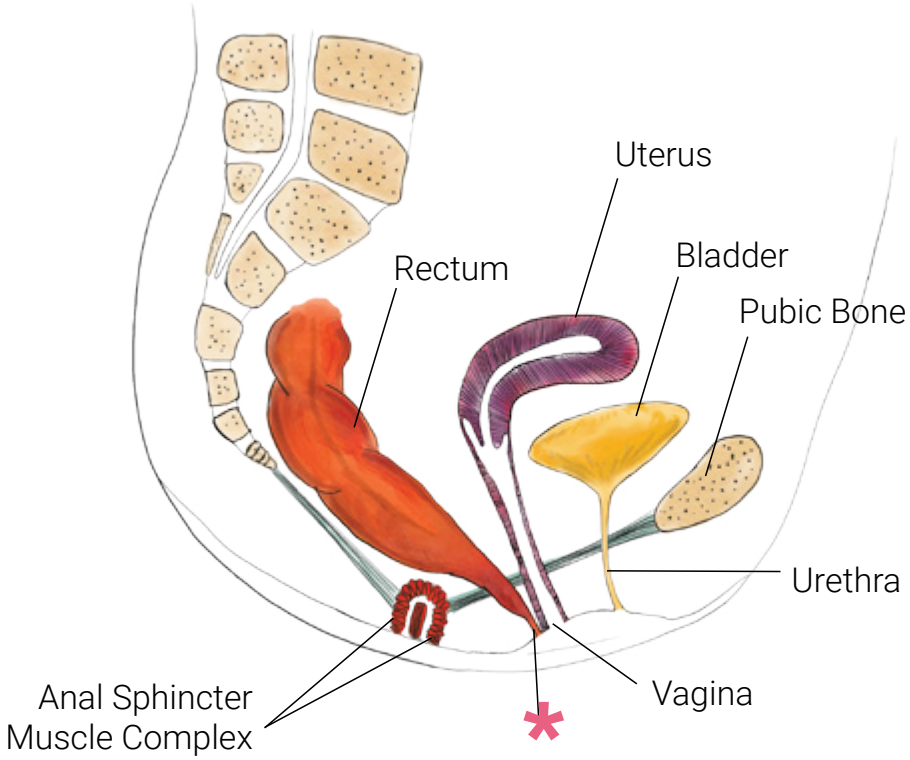


GIRLS ONLY

Rectovestibular Fistula

The lower part of the vaginal opening is called the **vestibule**. Rectovestibular (also referred to as: Vestibular) fistula occurs when the fistula opening is in the vestibule, instead of the usual location at the anus. There should be normal openings for the urethra and vagina that are separate and easily seen.

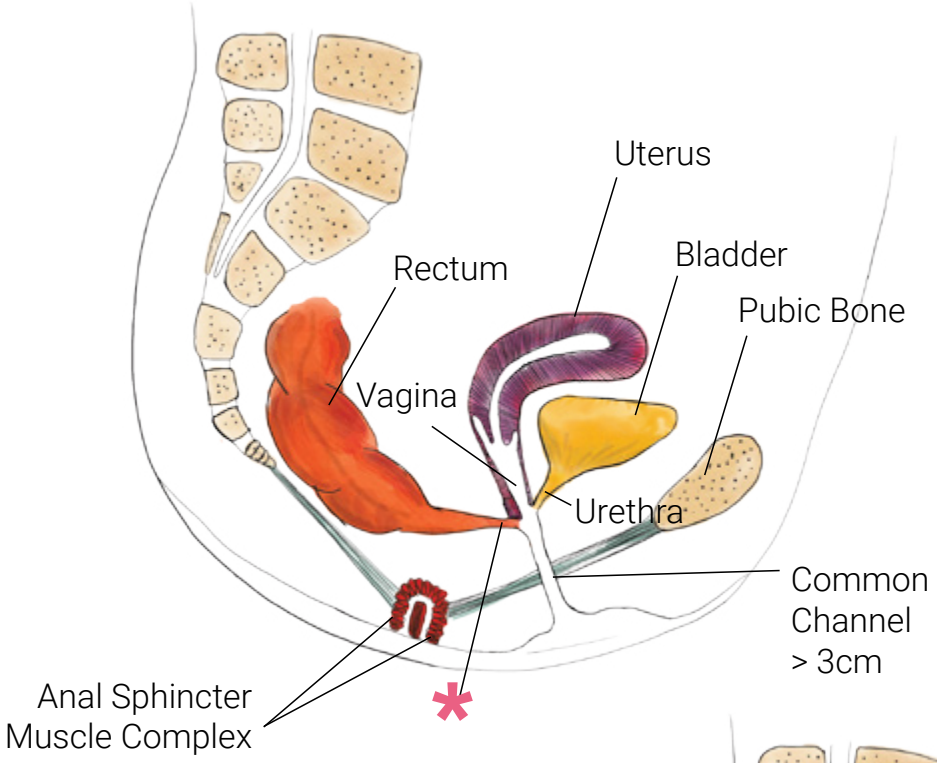
In most cases of rectovestibular fistula, the rectal opening (fistula) will be dilated (see Chapter 8) until it reaches a size that allows for poop to pass. Surgery is usually delayed until the child is a few months old (see Chapter 11).



Cloacal Malformation

Instead of having three separate openings (urethra, vagina, and anus), some girls can be born with a single opening called a cloacal malformation, where both urine and poop pass.

A child with a cloacal malformation will need a colostomy (See Chapter 7) until they are several months old and can have the repair. Cloacal malformation surgery is covered in Chapter 13.

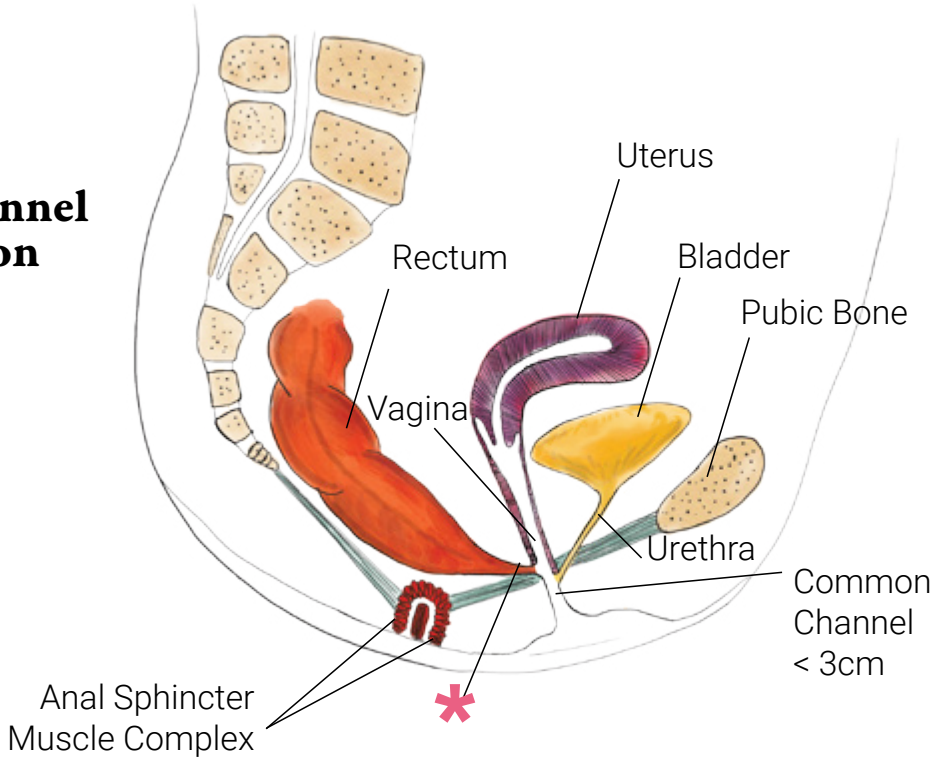


Long Common Channel Cloacal Malformation

Long Common Channel Cloacal Malformation is when the common channel is greater than 1 inch (3 cm) long and the urethra is usually shorter than 0.5 inches (1.5 cm).

Short Common Channel Cloacal Malformation

Short Common Channel Cloacal Malformation is when the common channel is less than 1 inch (3 cm) long and the urethra is usually longer than 0.5 inch (1.5 cm).

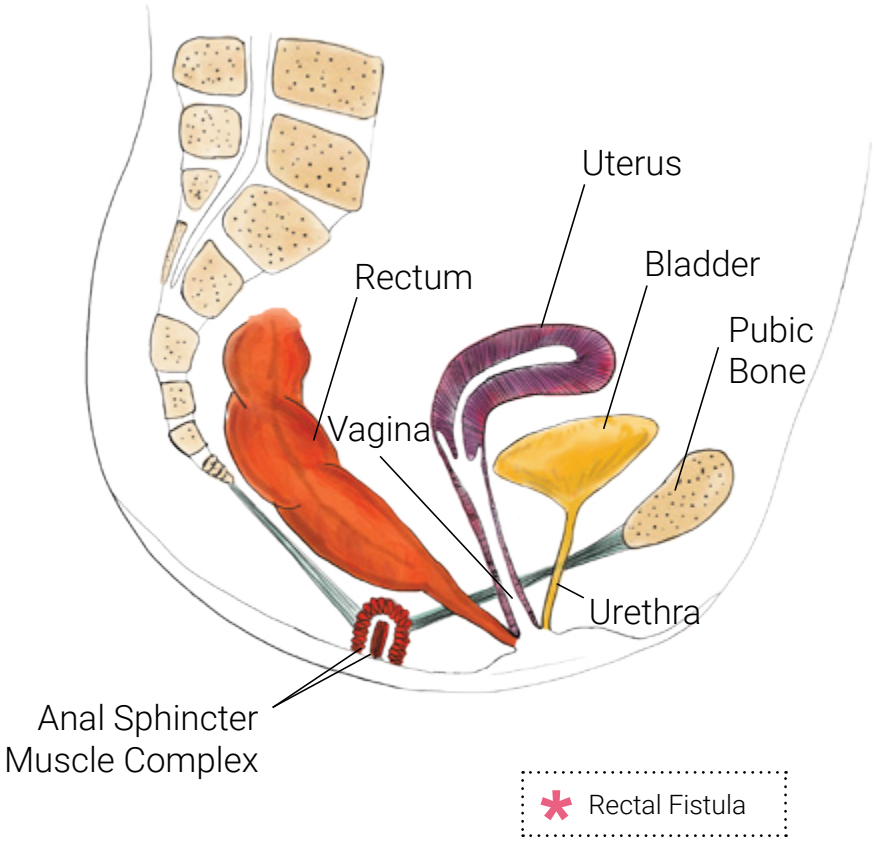


Type 1 Cloacal Malformation

Type 1 cloacal Malformation is when the common channel is 0.4 inches (1 cm or less in length.

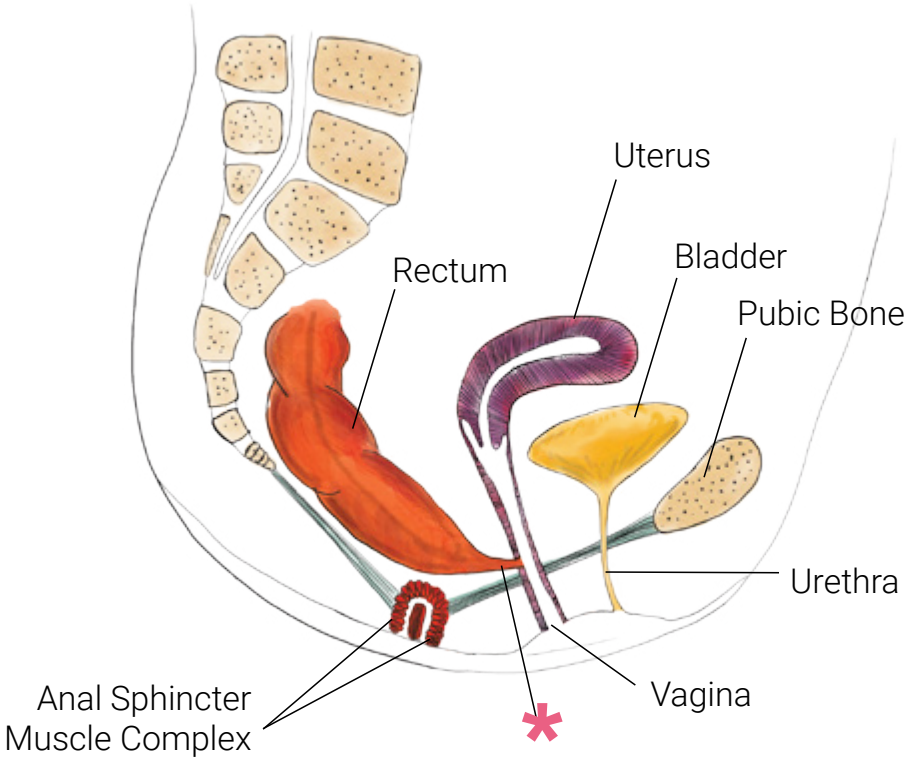
The term "common channel" refers to the single tube where the urethra, vagina, and rectum join together.

The distance of this tube can be "short" (less than 3 cm), or "long", (more than 3 cm)



Rectovaginal Fistula

In very rare cases in girls, the rectum instead of connecting to the anus goes into the vagina, higher than the vaginal opening. In these cases, the child will have a normal opening for urine (urethra) and normal vaginal opening. This is different than a cloacal malformation where all three structures connect to one opening. Surgery to repair a rectovaginal fistula is covered in Chapter 11.



3. Common Tests for ARMs

Once your child is diagnosed with an ARM, the doctors will need to evaluate if other conditions exist.

There is a group of conditions called **VACTERL** that are sometimes seen in ARM babies (see Chapter 5). Some of the tests that might be done to look for these conditions are:

X-rays, Echocardiogram, Magnetic Resonance Imaging (MRI) and Ultrasound.

X-rays can sometimes help your team see where the rectum ends. This is based on where they see gas on the X-ray. X-rays will also help find any problems with the bones of the spine and tailbone or sacrum.

Echocardiogram is an ultrasound of the heart to help find any heart defects.

MRI is sometimes done when the baby is older, around 6 months. It helps look at the spinal cord and organs in the pelvis.

Ultrasound

Kidney (renal) ultrasound show if there is a problem with your baby's kidneys, bladder, or the tubes that drain the urine from the kidney into the bladder, called the ureters.

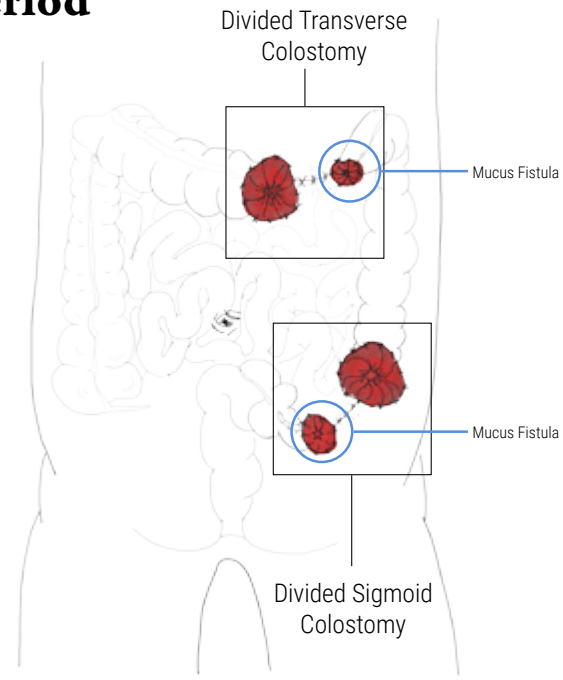
Pelvic ultrasound (for girls only) show if there is a problem with your baby's reproductive organs.

Spinal ultrasound will show if there is a problem with your child's spinal cord.

Special Tests Beyond the Newborn Period

Distal Colostogram

The repair of an ARM often needs more than one procedure. Once your child has recovered from their colostomy procedure (see Chapter 7), surgeons will do a study called a distal colostogram. A small tube is put into the mucus fistula. The mucus fistula (circled in blue on the right) is the smaller hole of your baby's colostomy. It does not drain poop but sometimes drains mucus or even urine. The illustration shows three different colostomy types and examples. Your child's will look similar to one of them. After putting in the tube, the radiologist will give a fluid called "contrast," which helps them see inside the fistula to better understand the anatomy.



Ostomy Types

3D Cloacagram
(for girls only)

A 3D Cloacagram is a type of "3D" x-ray done after tubes are put into the rectum, bladder, and vagina. It is done for children born with a cloacal malformation, to help plan for surgery.

Contrast Enema

A contrast enema is an X-ray test that helps your team see inside the colon. The term enema refers to fluid given through the anus. In this case, the liquid is a "contrast" that helps the radiologist see the rectum and colon to better understand the anatomy.

Anorectal Manometry

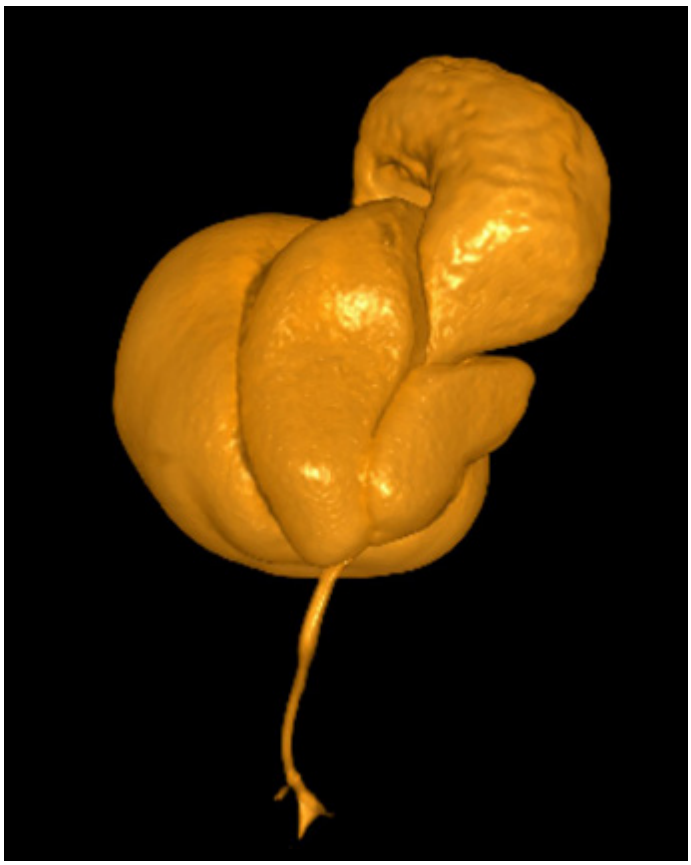
Anal manometry is a test to see how well the anus, internal and external muscles around the anus or sphincters, and the muscles in the pelvis are working. Your child may have this test if they are older and can help with the test. A small tube with a balloon at the end (called a Foley catheter) is put into your child's rectum. The balloon is gradually inflated to test how your child's nerves and muscles in the rectum work together. Your child may be asked to squeeze, cough, or try to push the balloon out of their rectum. The tube is connected to a computer which will provide information about the health of these muscles. These results will help your team decide what treatments are needed, including physical therapy.

Colonic Manometry

Colonic manometry shows how the colon works by putting a tube into your child's colon and attaching it to a computer. The computer records the strength and coordination of how the muscles in the colon contract.

Urodynamic Study

A urodynamic study provides information about the function of the bladder. In this test, a flexible tube with a balloon on the end is put into your child's bladder. The bladder is slowly filled with warm saline. The flow of urine is measured as your child passes urine into a special, private toilet. During this time, the bladder pressures are recorded along with any feelings your child is having.



3D Cloacal Malformation Imaging

4. Diagnosing ARMs Before Birth

In the past, ARMs were hard to diagnose before birth and were usually not found until birth or after. However, advances in using ultrasound and MRIs have allowed for better diagnosis of ARMs before birth.

Ultrasound Scans

Sometimes an ultrasound during pregnancy, at around 20-24 weeks, suggest your child has an ARM. Such findings include:

• Over-stretching of the rectum and the last part of the colon before the rectum or **sigmoid colon** is seen in the third trimester.

• **Meconium pearls** are seen when urine and the black sticky first poop of a baby or meconium mix. This is due to the abnormal connection or a fistula between the bladder or urethra with the bowel.

• If your baby does not have an anus or the muscle that allows the anus to open and close when making poop (sphincter), it might suggest an **ARM**.

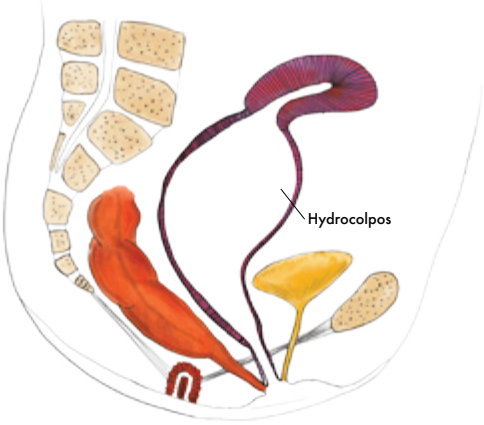
• Small anus or a narrow canal in the anus.

• If the genitalia is located close to the anus, and there is not a normal amount of tissue between the anus and vagina or scrotum (**perineal body**).

FOR GIRLS ONLY:

There are findings your ultrasound specialist may see on the ultrasound that suggests your child may have a cloacal malformation:

• Defects of the urinary system
• An enlarged vaginal cavity with fluid or hydrometrocolpos.



VACTERL

About 50% of newborns with ARMs have more than one abnormal problem that happened during development. These problems often include several clinical conditions called VACTERL (see Chapter 5).

If there are abnormal findings on the ultrasound that occur in these areas of the body, it might suggest that your child has an ARM.

V	Vertebral
A	Anorectal
C	Cardiac
T E	Tracheo - Esophageal
R	Renal
L	Limb

(See Chapter 5)

MRI Scans

MRI scans of your baby before birth can also help diagnose ARM. MRI scans are not usually needed in routine cases. However, if there are several abnormal findings seen during your ultrasound screening, an MRI might be suggested. The MRI gives more detail about whether there is an ARM and any other abnormalities. These abnormalities may include a horseshoe kidney, tethered cord, or absent sacrum that may make a difference on how the ARM is treated. In girls, an MRI is better than ultrasound when it comes to diagnosing a cloacal malformation.

What comes after diagnosing an ARM before birth?

In general, there are no other tests needed after an ARM is diagnosed before birth. There are no treatments needed during pregnancy. You may be asked to see a pediatric surgeon to learn more about an ARM diagnosis. Rarely will you need any specific additional exams during your pregnancy.

If a complex ARM is diagnosed before birth, such as a cloacal malformation or rectobladder neck fistula, delivering your baby close to a medical center that specializes in treating ARM is beneficial. The baby will likely need surgery within the first few days of life and will need testing to see if there are other abnormalities.

5. Common Conditions Seen with ARMs

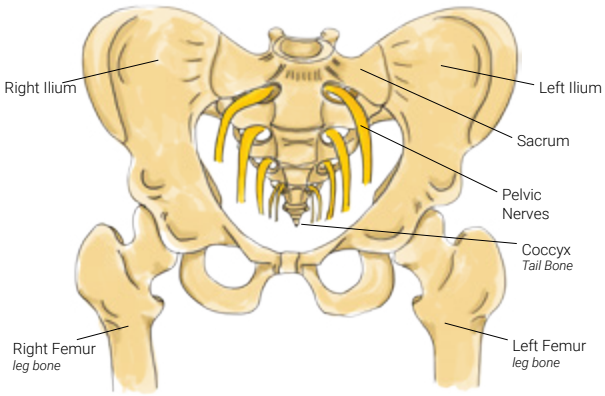
About 50% of babies with ARMs have other abnormal findings. There is a group of these findings that are often seen together in babies with ARMs. The acronym VACTERL describes these problems. Your baby may have one or more of these problems. If your child has three or more of these conditions, they are considered to have a **VACTERL Association**. Below is a list of these findings:

V – Vertebral Problems

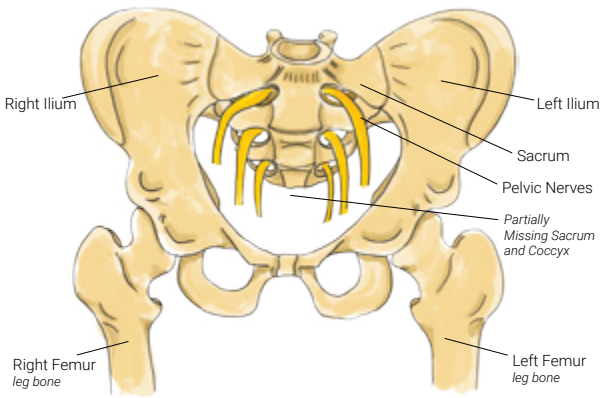
The bones of the spinal column, or the vertebrae, can be abnormally formed in children with ARMs. The vertebrae are small (**hypoplastic**) or sometimes only half-formed (**hemivertebrae**). Some babies will also have missing ribs, an increased number of ribs (**supernumerary ribs**), **rib fusions**, or **splitting of ribs**.

Sacral Agenesis

The tailbone (**coccyx**) can also be missing, or the lowest part of the spinal column is not there (**sacral agenesis**), as illustrated to the right. These differences in anatomy are not life-threatening and, most of the time, do not need to be fixed. However, sometimes, your child can be at risk of developing an abnormal curvature of the spine, or scoliosis, later in childhood. An x-ray of the spine is used to diagnose most of these differences in anatomy.



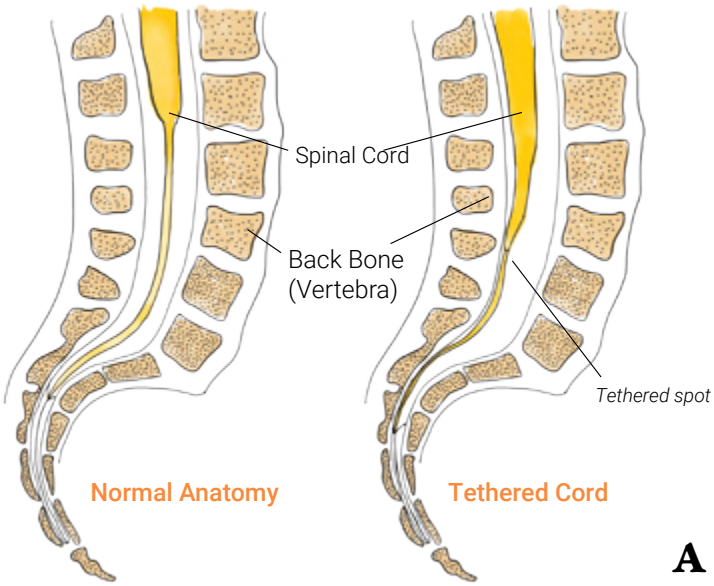
Normal Anatomy Of The Pelvis (Hip)



Example Of Sacral Agenesis

Tethered Spinal Cord

There can also be other problems with the spinal cord. Usually, the spinal cord floats free inside the spinal canal. However, in some babies with ARM, the **spinal cord** is pulled down and stuck or tethered to the spinal canal by abnormal



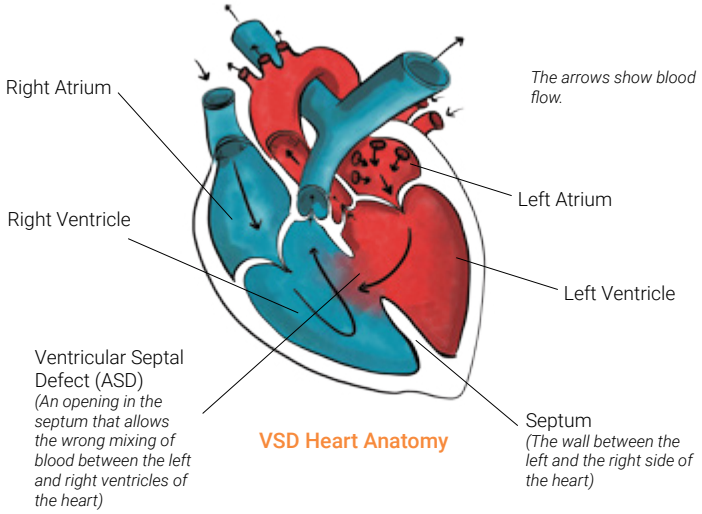
attachments. This is known as **Tethered Spinal Cord**. If your child's spinal cord is stuck, the cord will stretch as they grow like a rubber band which can cause lasting damage to the spinal nerves. Therefore, it is vital to get an ultrasound of the spine when your baby is less than three months old to look for a tethered cord. Beyond the age of 3 months, your child will need an MRI since the ultrasound is not helpful because the bones become denser. Not all children with a tethered spinal cord need treatment, but your child should be seen and followed by a neurosurgeon.

A - Anorectal Malformation

Please read those sections on ARMS that are written throughout this book.

C - Cardiac (Heart) Problems

There are heart conditions seen with ARMs, ranging from those that do not need surgery and resolve over time to life-threatening conditions that need surgery while your child is a newborn.



Ventricular Septal Defect (VSD)

The most common heart defect seen with VACTERL is a **ventricular septal defect** (VSD). This is where there is a hole in the wall (septum) between the lower chambers (ventricles) of the heart. Depending on the size and location of the hole, it may just need to be watched over time and will not need surgery. However, if the hole is large and your child develops signs that the heart cannot pump or fill correctly (congestive heart failure), surgery might be needed.

Atrial Septal Defect

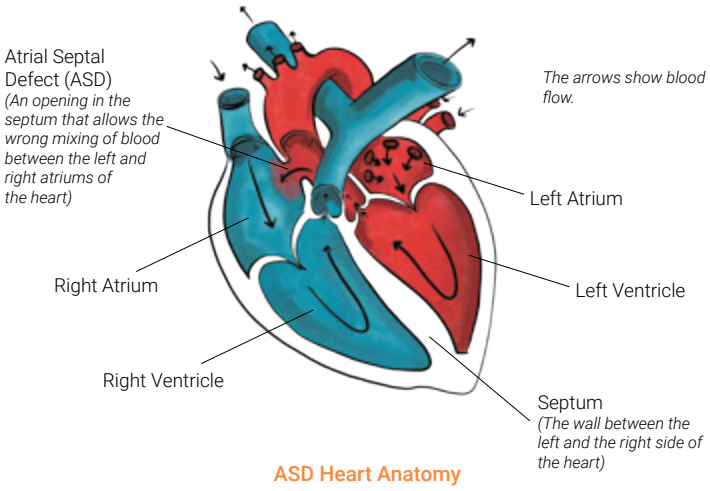
Additional heart problems seen with VACTERL include an Atrial Septal Defect, or a hole in the wall between the heart's upper chambers or atria. Many of these do not need to be fixed.

Tetralogy of Fallot

There can also be Tetralogy of Fallot, a condition where the passage between the blood vessel that leads to the lungs (pulmonary arteries) and the body's major artery (aorta) fails to close after birth.

The heart ultrasound (echocardiogram) done at birth will show any of these problems. Babies with these conditions are usually seen and followed by a cardiologist or heart doctor that treats children.

If your doctor discovers that your child has an ARM when they are older, typically a perineal fistula (see Chapter 2), a lot of cardiac testing is not done, if your child is healthy and does not have heart murmurs.

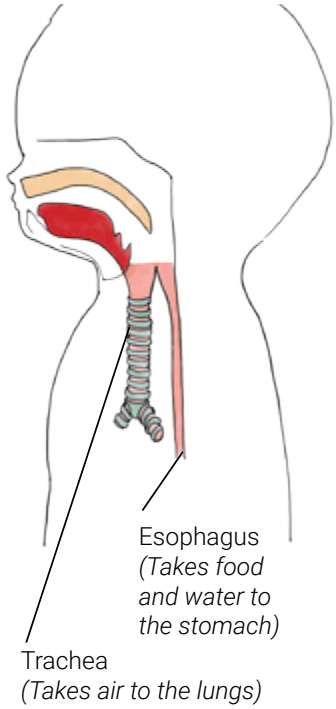


ASD Heart Anatomy

TE - Tracheoesophageal (Problems with the Esophagus)

The tube that connects the mouth with the stomach, or esophagus, can have problems. When the esophagus does not form normally, several things can occur:

Most of these conditions may need immediate repair while your child is a newborn.

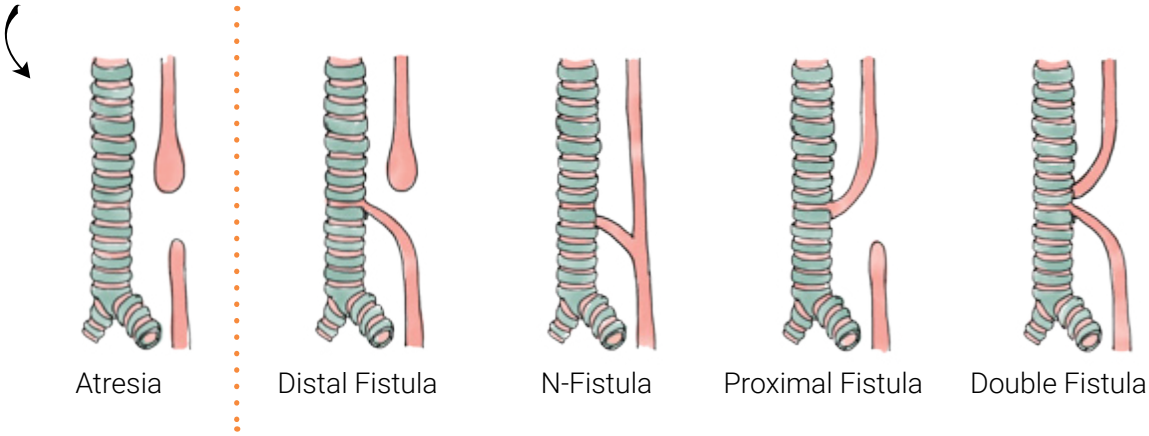


Esophageal Atresia

Esophageal Atresia is when an esophagus ends in a blind pouch. This condition is diagnosed if your baby does not swallow after the first feedings after birth or if a tube cannot be passed down into their esophagus.

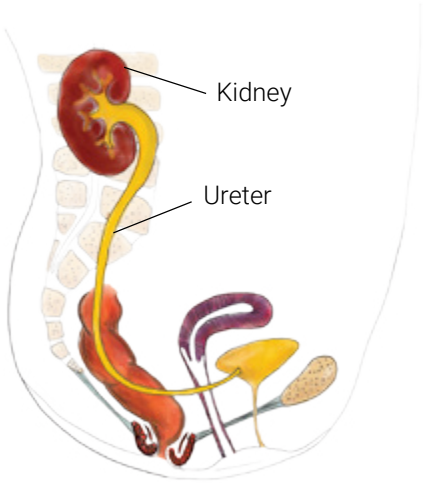
Tracheoesophageal Fistula

Tracheoesophageal Fistula is when the esophagus with a blind-ending pouch has an abnormal connection to the windpipe (trachea).



R - Renal (Kidney) Conditions

Some children have abnormalities in the urinary system. The urinary system includes the kidneys, ureters, bladder, and urethra.



Normal Anatomy

Vesicoureteral Reflux

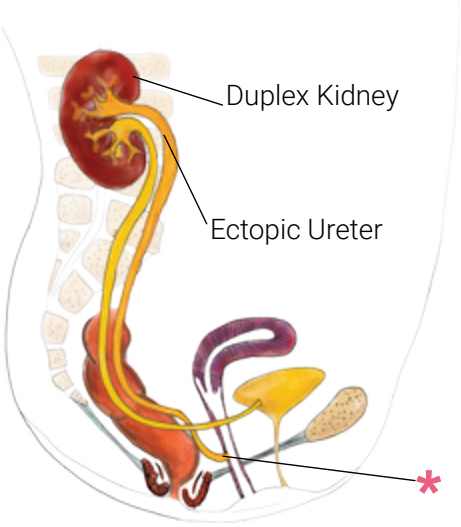
Vesicoureteral Reflux, the most common condition, is an abnormal backflow, or reflux, of urine into the ureter. The ureter is the tube that carries urine to the bladder. **Hydronephrosis**, an enlargement of the ureters, may be caused by **vesicoureteral reflux**. This reflux causes the ureters to enlarge and, in the long term, can cause kidney damage.

Renal Aplasia

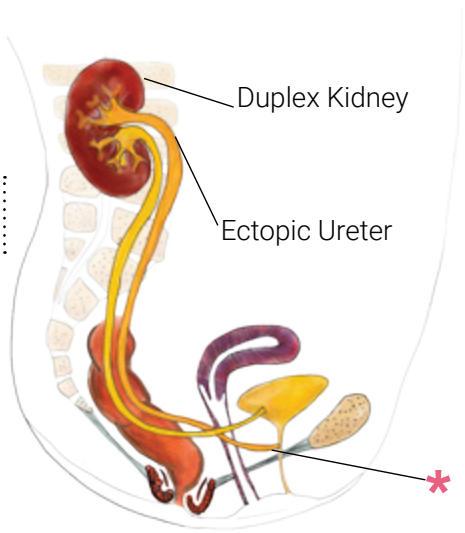
Renal Aplasia is when one or both kidneys are missing.

Renal Dysplasia

Renal Dysplasia is when one or both kidneys do not develop normally.



Duplex Kidney And Ectopic Ureter (Vagina)



Duplex Kidney And Ectopic Ureter (Urethra)

Fistula

Renal Ectopia

Renal Ectopia is when the kidney is not in the normal position within the body.

Duplex Ureter

Duplex Ureter is when there is an extra tube that carries urine from the kidney to the bladder.

Most of these conditions are diagnosed on the kidney ultrasound while your child is a newborn and will not need to be fixed.

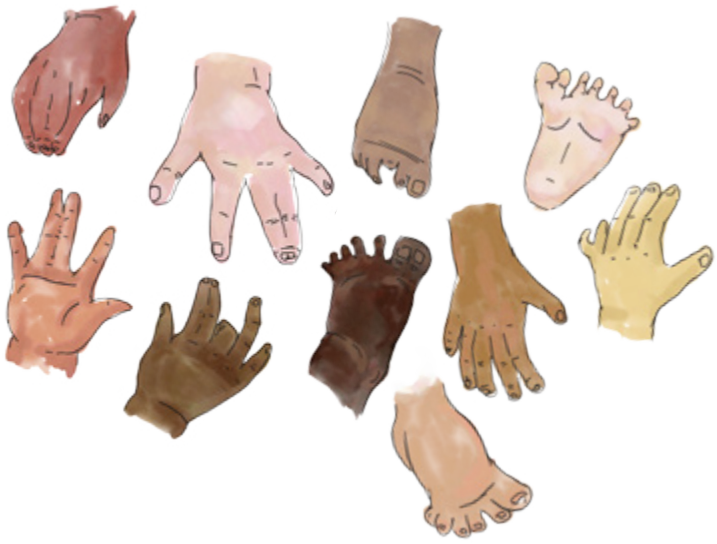
However, your child should be followed by a urologist.

L - Limb (Arms or Legs)

Some children have abnormally developed bones of the arms or legs, such as:

- Missing or abnormally shaped thumbs
- Extra fingers, known as **polydactyly**
- Fusion of the fingers known as **syndactyly**
- Polydactyly and syndactyly together are known as **polysyndactyly**
- Abnormal fusion of the forearm bones known as **radioulnar synostosis**
- Abnormal lower limbs, such as **clubfoot**.

Some of these conditions may need surgery or casts when your child is older. If your child is diagnosed with any problems with their arms or legs, they should be seen and followed by an orthopedic surgeon.



Polysyndactyly, Polydactyly And Syndactyly

Other less common birth defects that are not part of VACTERL have been reported in babies with ARMs:

Hemifacial Microsomia

The abnormal shape and size of the ears.

Laryngeal Stenosis

Narrowing of the upper part of the airway.

Choanal Atresia

Narrowing of the passages from the back of the nose to the throat

Omphalocele

Some of the intestines, liver, or other organs bulge out or protrude through the belly button with a thin, nearly clear covering.

Malrotation

A blockage or twist in the intestine. Malrotation occurs when the intestine does not form properly inside the womb. This leads to the intestine sitting in an abnormal place within the belly, making the child at risk for a blockage or twist.

Currarino Triad

Currarino triad is a rare condition, seen in 1 out of 100,000 people, that can be seen with ARMs. It is a genetic condition that has 3 abnormal problems (triad).

1. Sacral agenesis:

An abnormally developed lower spine (see page16).

2. ARM:

This can be any of the following:

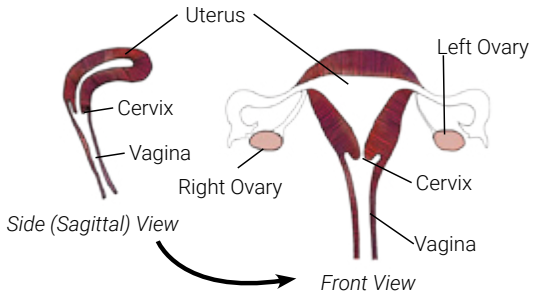
- An abnormally narrow anus (**anal stenosis**)
- The rectum is missing but the anus looks normal (**rectal atresia**)
- A misplaced anus (**perineal fistula**). (see Chapter 2).

3. Presacral mass:

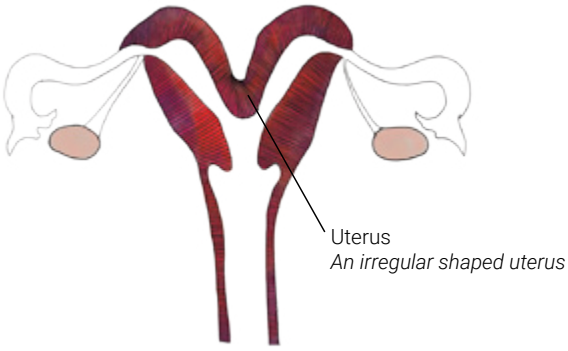
An abnormal growth in front of the sacrum or the lower part of the spine. This mass can sometimes be a type of childhood tumor (a **teratoma**), most of these tumors are benign. Very rarely however patients can develop cancer in this **teratoma**. In addition, patients may have an abnormal lower spinal cord and spinal canal (**anterior sacral meningocele**).

Not every child with **Currarino Triad** will have all three problems. Only 1 out of 5 children with this condition have all three problems. Children with this condition can have varying degrees of severity. Almost a third of children have no symptoms and get diagnosed later in childhood, usually from having severe constipation and a narrow or abnormal anus.

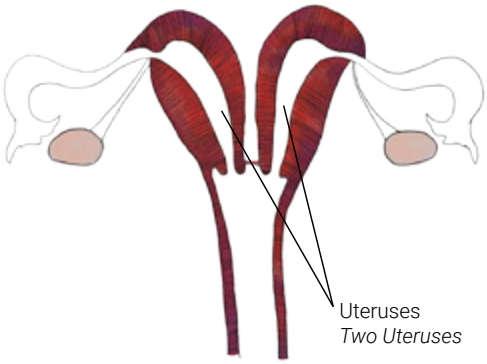
Gynecological Anomalies



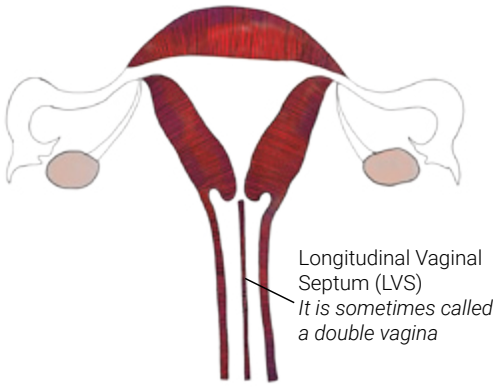
Normal Anatomy



Bicornuate Uterus



Uterus Didelphys



Normal Uterus W/ Longitudinal Vaginal Septum

About a third of girls with ARMs may have some changes in the anatomy of their vagina or uterus.

The simplest of these changes is termed a "**vaginal septum**." It is a wall of tissue running up and down the length of the vagina, dividing it into two separate cavities. It is sometimes also called a "**double vagina**." Surgery removes the wall or septum and keeps the vagina as one cavity. It is best to delay this procedure until the child is older.

The most common problems of the uterus are **bicornuate uterus** and **uterus didelphys**. A **bicornuate uterus** is where the uterus is shaped like a heart instead of the usual pear shape.

A **uterus didelphys** is where the uterus is double.

Sometimes, a child may also have two vaginas and two cervices. These changes in the uterus are usually noticed during surgery for ARM or picked up by ultrasound or MRI. Some of these conditions may not need any treatment right away and are dealt with when your child is older and goes through puberty. However, it is important to have follow-ups as your child grows since some vaginal and uterus issues can cause problems with menstruation. Your colorectal team, especially your pediatric gynecologist (GYN), can help you through this process and explain any care and follow-up your child may need over time.

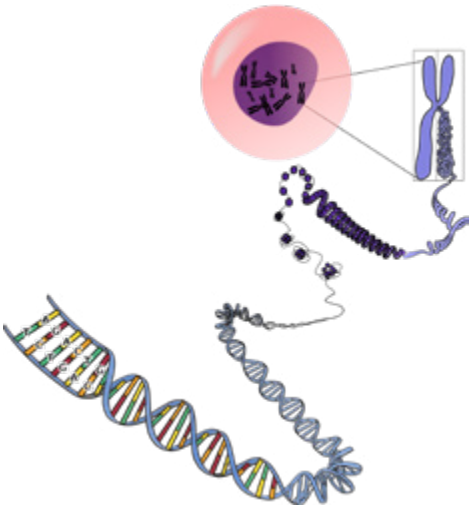
6. Genetic Counseling

Anorectal malformations or ARMs are present from birth and develop during pregnancy. Most of the time, the cause for ARMs is unknown. As a parent, you may have feelings of guilt, anger, and shock. These feelings are understandable.

You can discuss any of your concerns with your child's medical team at any time.

How Genetics Impacts Your Baby's Development

As your baby grows in your womb, the organs develop at different times. The lower end of the intestines or bowels forms early in pregnancy. For the rectum and anus to be normal, there are specific ways the intestines must form during the 7th to 10th weeks of pregnancy. Sometimes this does not happen as it should, and the intestines may not develop properly.



Our body has billions of cells and inside each cell are chromosomes. Chromosomes are structures that have thousands of genes. Genes hold information about features, such as our eye color and hair color. Genes also carry instructions that tell your cells how to work and grow.

A person can have changes or mutations in one or many genes. Sometimes, those changes do not cause any issues. However, some gene changes can cause conditions like ARMs.

ARMs occur when both genes and environment interact.

In other words, most of the time, no single gene causes an ARM. Instead, it is believed that genetic and environmental factors combined cause ARMs.

Sometimes ARMs occur as an isolated defect, meaning the child is otherwise healthy. However, ARMs can also occur as part of a set of abnormal findings that occurred during the baby's development, such as **VACTERL** changes or **Currarino Triad.** A set of abnormal findings are known as a **syndrome.** (See Chapter 5)

ARMs have also been seen with changes in **chromosomes**, such as **Trisomy 21 (Down Syndrome), Trisomy 18, and Trisomy 13.**

Your child may be the first in your family to have an ARM. Sometimes, however, there are patterns that exist, meaning another family member also has the condition. In most children with ARMs, genetic testing is unnecessary.

Depending on the type of ARM your child has, there may be chances of future children having this condition. The risk of **rectovestibular fistula** and **perineal fistula** may be as high as 3 to 4% for brothers or sisters from the same parents.

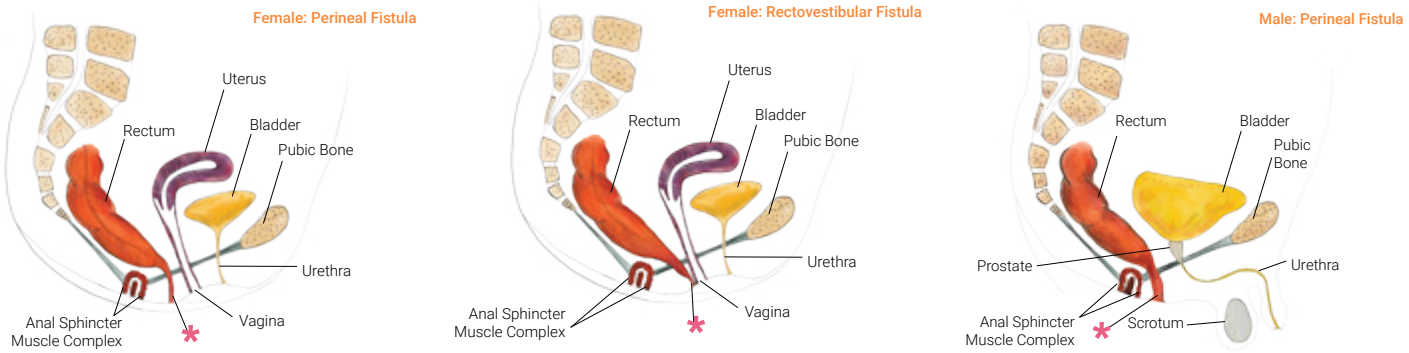
If your child has an ARM, you may decide to meet with a **genetics healthcare provider** or a **genetic counselor**. They can have a more in-depth discussion of your situation and any risks with future pregnancies.

7. Surgery for Your Newborn Baby

Perineal Fistula and Rectovestibular Fistula

Your newborn can sometimes avoid surgery right away if your baby has either:

- A small anus opening in the wrong spot (**perineal fistula**).
- A small anus opening next to the vagina (**rectovestibular fistula**).



Hegar Dilator

A medical instrument, called a **Hegar Dilator**, helps open or dilate the narrow opening. This allows poop to leave the body, which avoids surgery while the baby is a newborn. Using a Hegar Dilator regularly for the first few months after birth, can help delay surgery (PSARP) to fix the ARM until your child is a few months old. See Chapter 8 to read about Hegars and dilation.

Sometimes, surgeons may decide to do **PSARP Surgery** (See Chapters 10 & 11) for your newborn with a perineal fistula or rectovestibular fistula. Surgery is generally safe in a newborn child if the surgeon has done this surgery many times.

Surgery

Most babies with perineal fistula and rectovestibular fistula will not need a colostomy as these can be managed initially with dilations. However, in some cases your provider may opt to create a colostomy before the PSARP surgery needed to repair the condition. In all other types of ARMs your baby will need a colostomy right away.

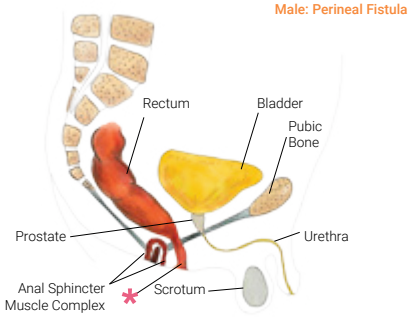
A colostomy is a surgery that allows poop to come out of your baby through an opening that the surgeon makes. A colostomy helps the body function, delaying the PSARP surgery until your child is a few months older.

Male

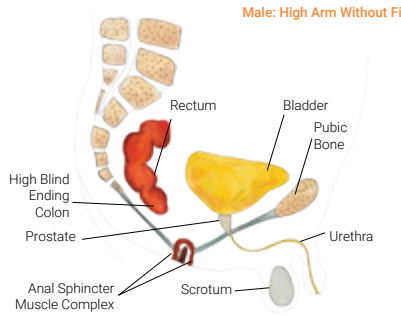
Below are the different types of surgeries that your newborn might need depending on the type of ARM they have.

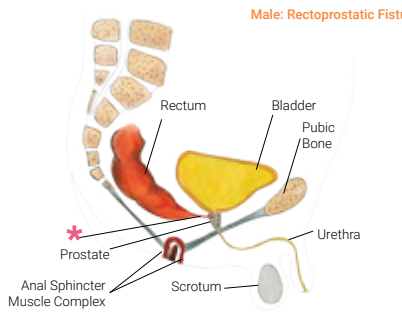
Type of ARM	Bowel drainage (colostomy)	Bladder drainage (suprapubic tube [SPT], or vesicostomy tube)
Perineal fistula	Rare	Very Rare
All other ARMs	Always	Rare

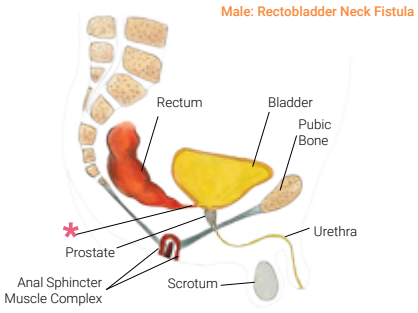
Perineal Fistula

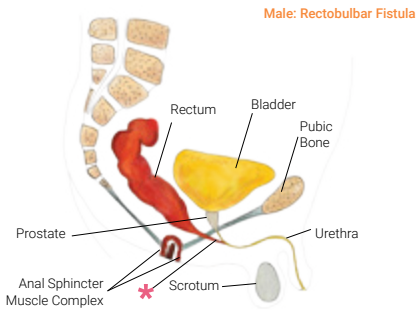


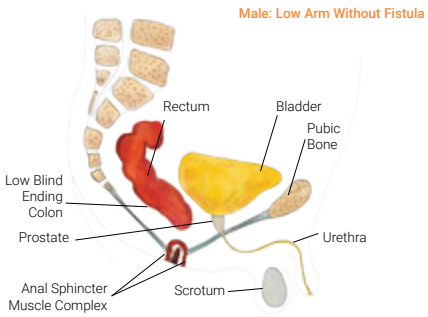
All other ARMs







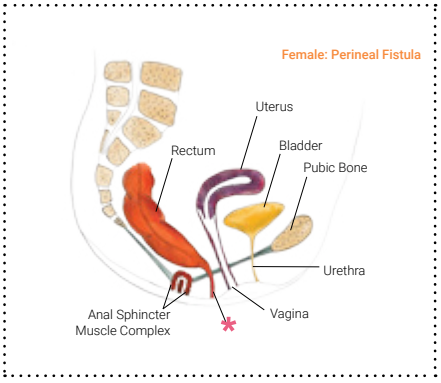




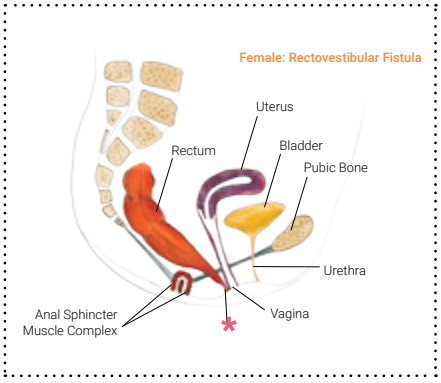
Female

Type of ARM	Bowel drainage (colostomy)	Bladder drainage (suprapubic tube [SPT], or vesicostomy tube)	Vaginal drainage (vaginostomy)
Perineal fistula	Rare	Very Rare	Very Rare
Rectovestibular fistula	Sometimes	Very Rare	Very Rare
Rectovaginal fistula	Always	Very Rare	Very Rare
Cloacal malformation	Always	Sometimes	Sometimes

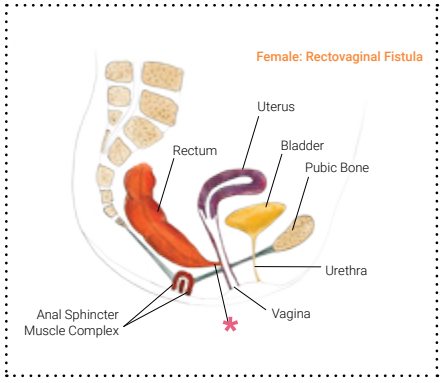
Perineal Fistula



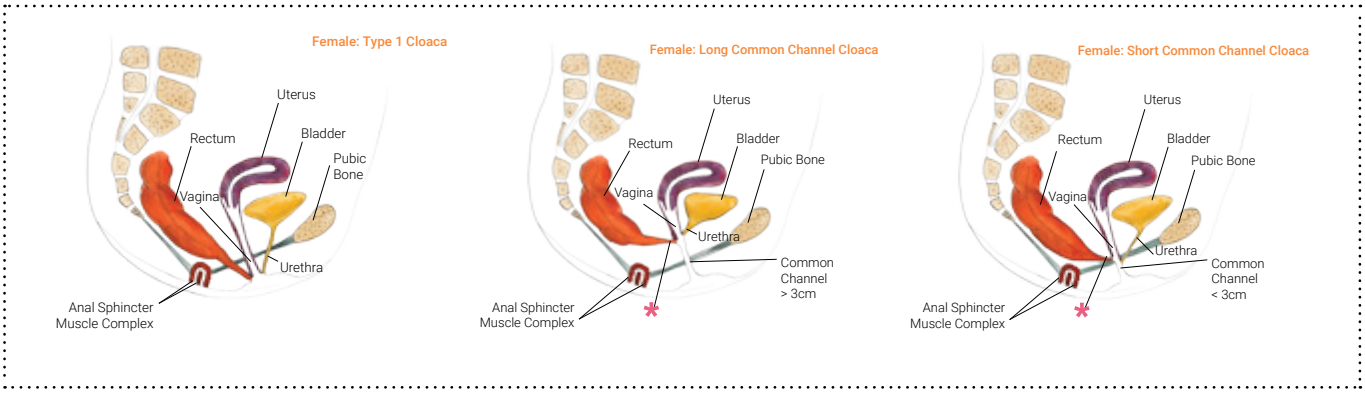
Rectovestibular fistula



Rectovaginal fistula



Cloacal Malformation Types



Colostomy

If your child needs a way to get their poop out, they will need a **colostomy.** After the colostomy, your baby will have two openings on the skin. The ostomy is an opening (**stoma**) from the colon inside the body to the outside. It allows poop to come out of your baby's body until it is time to do the surgery for the new anus. The other end of the bowel that goes down to the rectum is brought out of the body separately. This is called a **mucus fistula**. A **mucus fistula** only drains mucus and sometimes small amounts of urine, depending on the type of ARM your baby has.

There are two ways for colostomy surgery to be done: open or laparoscopic.

Open Colostomy Surgery

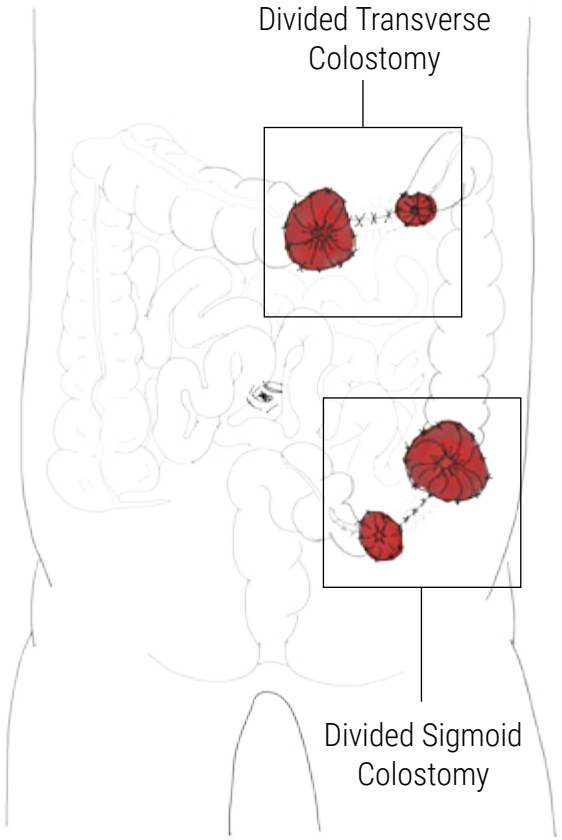
In an open colostomy surgery, a cut is made in the left lower part of the belly. The colostomy and mucus fistula are brought out separately, and the cut is then closed with stitches (top right drawing).

Laparoscopic Colostomy Surgery

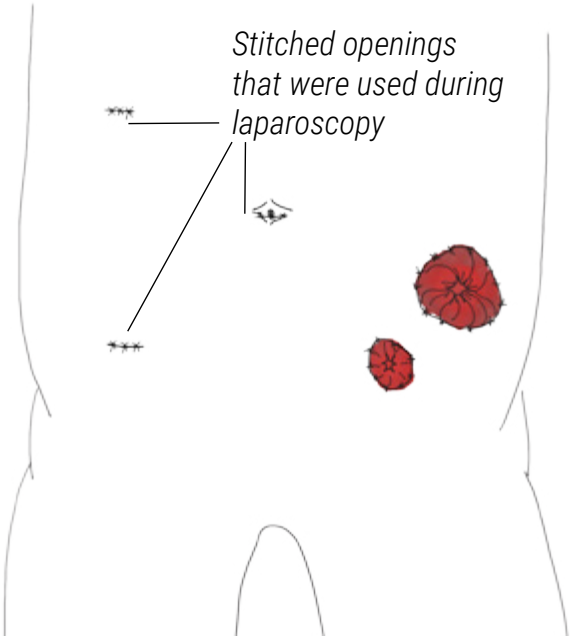
With the laparoscopic type of surgery, only a few tiny cuts or holes to enter the belly are needed. The colostomy and mucus fistula are brought out through smaller cuts. (bottom right drawing).

After Colostomy Surgery

Once your child has recovered from having the colostomy done, the surgeons will do an X-ray test called a **distal colostogram**. With this test, a small tube is put into the mucus fistula and fluid, called **contrast**, is pushed through the tube. This allows the radiologist to see where the liquid is going on an X-ray and help to identify the anatomy.



Ostomy Types



Laparoscopy For Sigmoid Colostomy

Bladder Drainage

If your baby girl has a **cloacal malformation**, the urine drains through a common opening. The common opening is the channel where the urethra, vagina, and rectum meet in the wrong place. (See Chapter 2).

In most cases, putting a tube into the common opening a few times each day (called **clean intermittent catheterization (CIC)**) is sufficient to help drain the urine. In some cases, your baby may have problems emptying her urine through this single opening. Your baby's surgeon might create another way for urine to leave the body during their colostomy surgery.

The two common ways for urine to leave the body are to either:

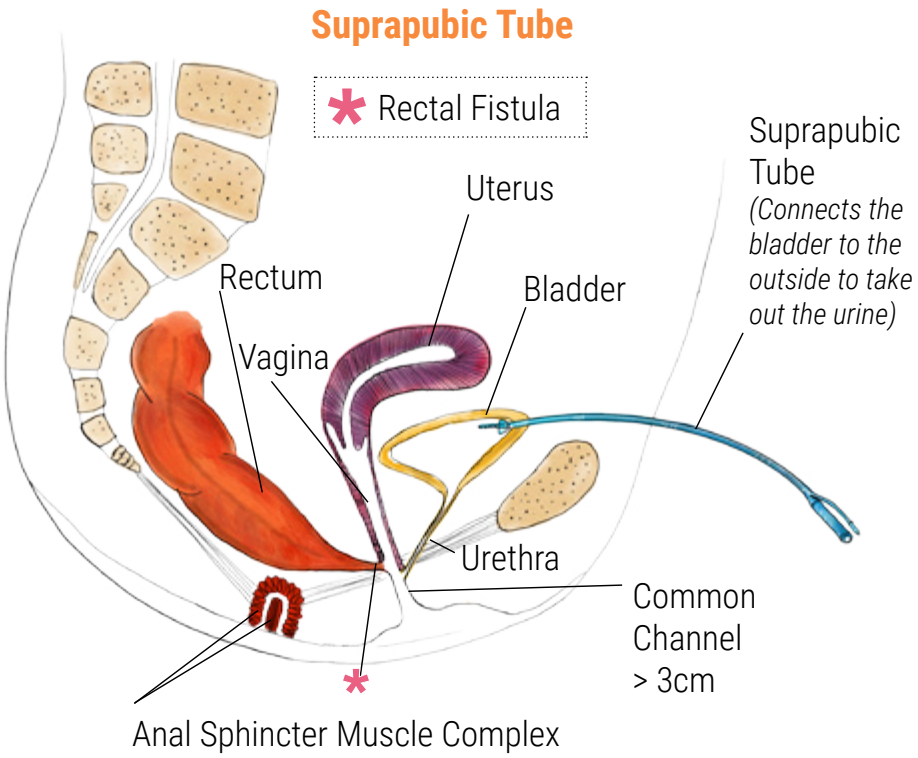
- Place a **suprapubic tube**
- Do a **vesicostomy**

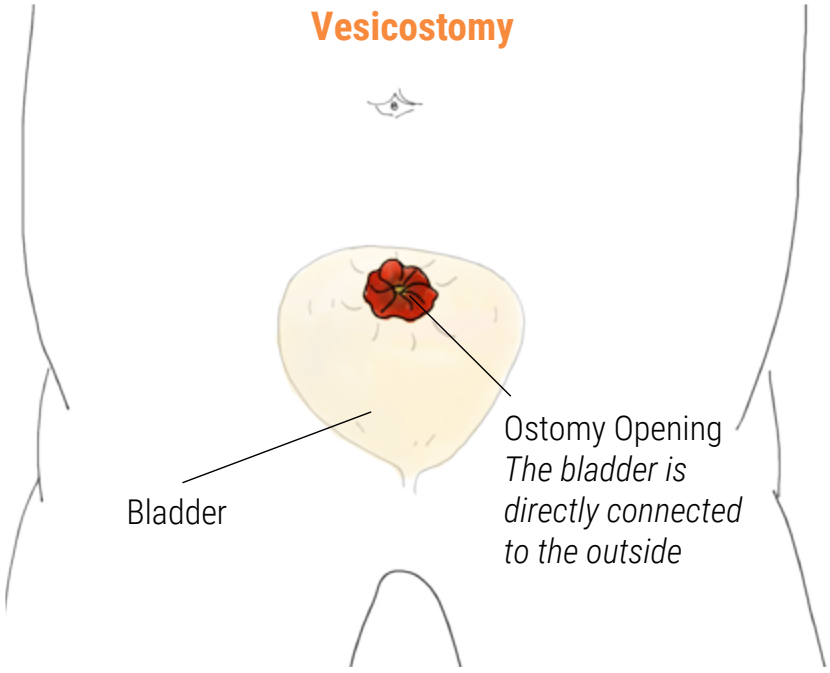
Suprapubic Tube

A **suprapubic tube**, also called an **SP tube**, is directly put into the bladder through the skin above the pubic area. The tube is then attached to a bag to collect the urine outside the body.

Vesicostomy

A **vesicostomy** is where the surgeon brings a part of the bladder to the outside of the belly as a **stoma**.



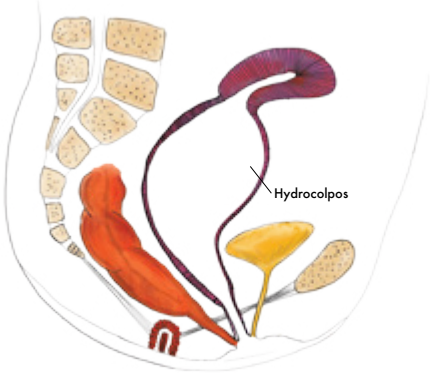


Vaginal Drainage

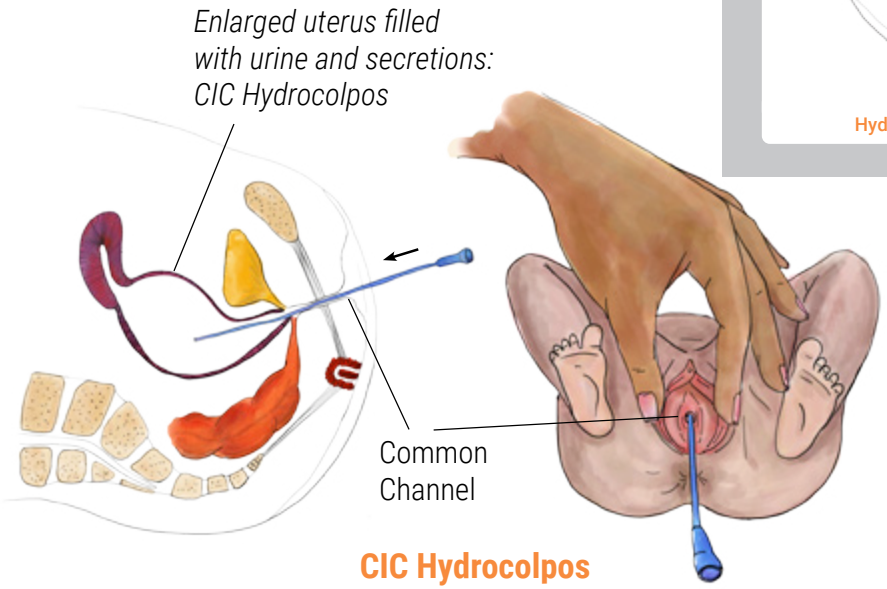
If your baby girl is born with a cloacal malformation, she may have a sizable urine-filled vagina, called **hydrocolpos**.

Hydrocolpos is drained by putting a tube into the common cloacal malformation opening a few times each day. This is called **clean intermittent catheterization (CIC)** (see drawing below).

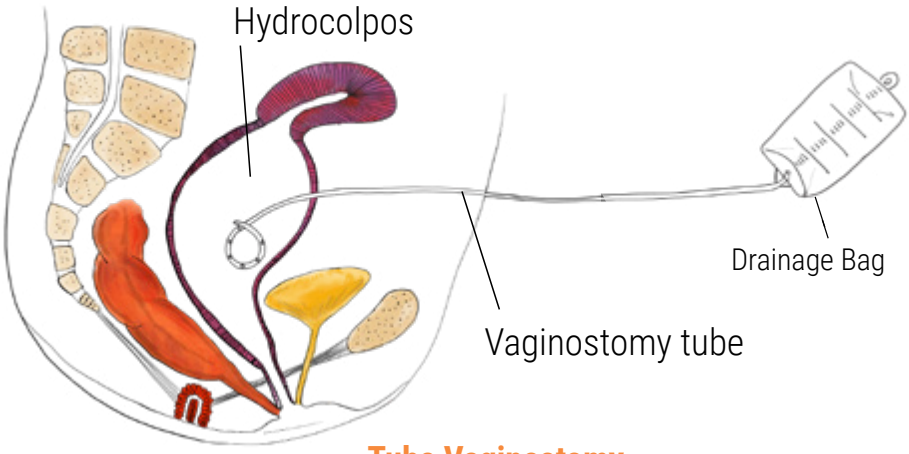
A **hydrocolpos** forms when urine flows into the vagina, since the bladder, vagina, and rectum share a common opening. Sometimes, the hydrocolpos is so large that it causes breathing and feeding problems for your baby.



Hydrocolpos (Hydrometrocolpos), Cloaca



CIC Hydrocolpos



Tube Vaginostomy

Tube Vaginostomy Vs Vaginostomy

However, sometimes your baby needs the hydrocolpos drained at the time of her colostomy surgery. Surgeons either insert a tube directly into the vagina and bring it to the outside (**tube vaginotomy**) (illustrator below), or they bring part of the vagina to the outside as a **stoma** (**vaginostomy**), like a colostomy stoma.

In the majority of children, bladder and vagina drainage procedures are temporary. After the final repair of the ARM, these will be reversed.

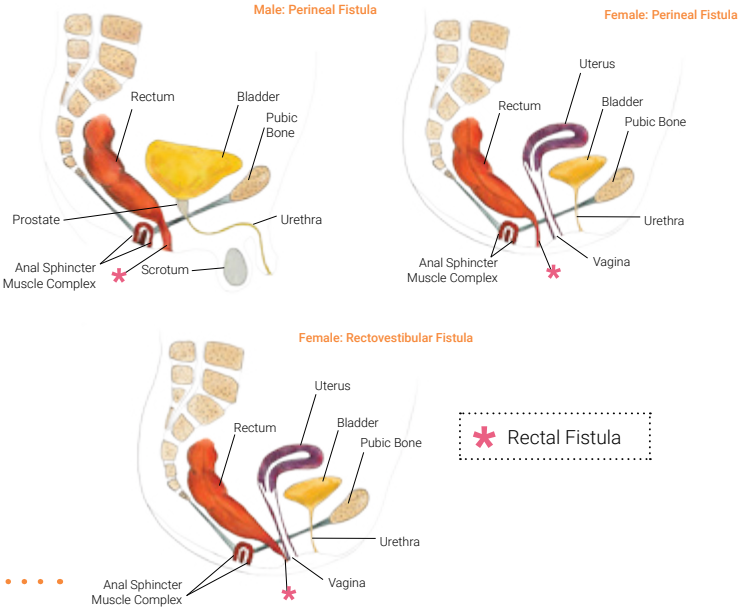
8. Dilation in the Newborn Period

All types of ARMs will need surgical repair. Certain ARMs may be treated with dilation when your baby is a newborn. Dilation is the first way to help your baby get rid of their poop.

The conditions in which dilations are used include:

- **Perineal fistula (male and female)**
- **Rectovestibular (Vestibular) fistula (female only)**

In these two conditions, the anal opening, or fistula, is too small and not in the right place.



Dilations

A dilation is when a metal or plastic rod called a **Hegar Dilator** is passed into your baby's fistula **one or two times a day**. This stretches the opening to allow the poop to come out more easily. The goal of dilation is to delay surgery while your baby is a newborn. The surgeon will decide the best time for your baby to have a surgical repair.

Your surgeon decides on the size of the fistula before dilations begin. The dilator's size will be slowly increased based on how well your baby does with the dilation. Your surgeon will have a plan for the final size dilator.

While your baby is in the hospital, you will be taught how to dilate your baby's fistula.

After Surgery

Sometimes dilations still need to be done after surgery. These dilations keep scar tissue from forming over the new opening and reducing its size. Your surgeon will let you know if you need to start dilations after surgery. They will also let you know when to start dilations and the Hegar size to use. You will be told when you can slowly stop doing the dilations. This is done by doing it one time a day for a while and then every other day until you are dilating only one time a month.

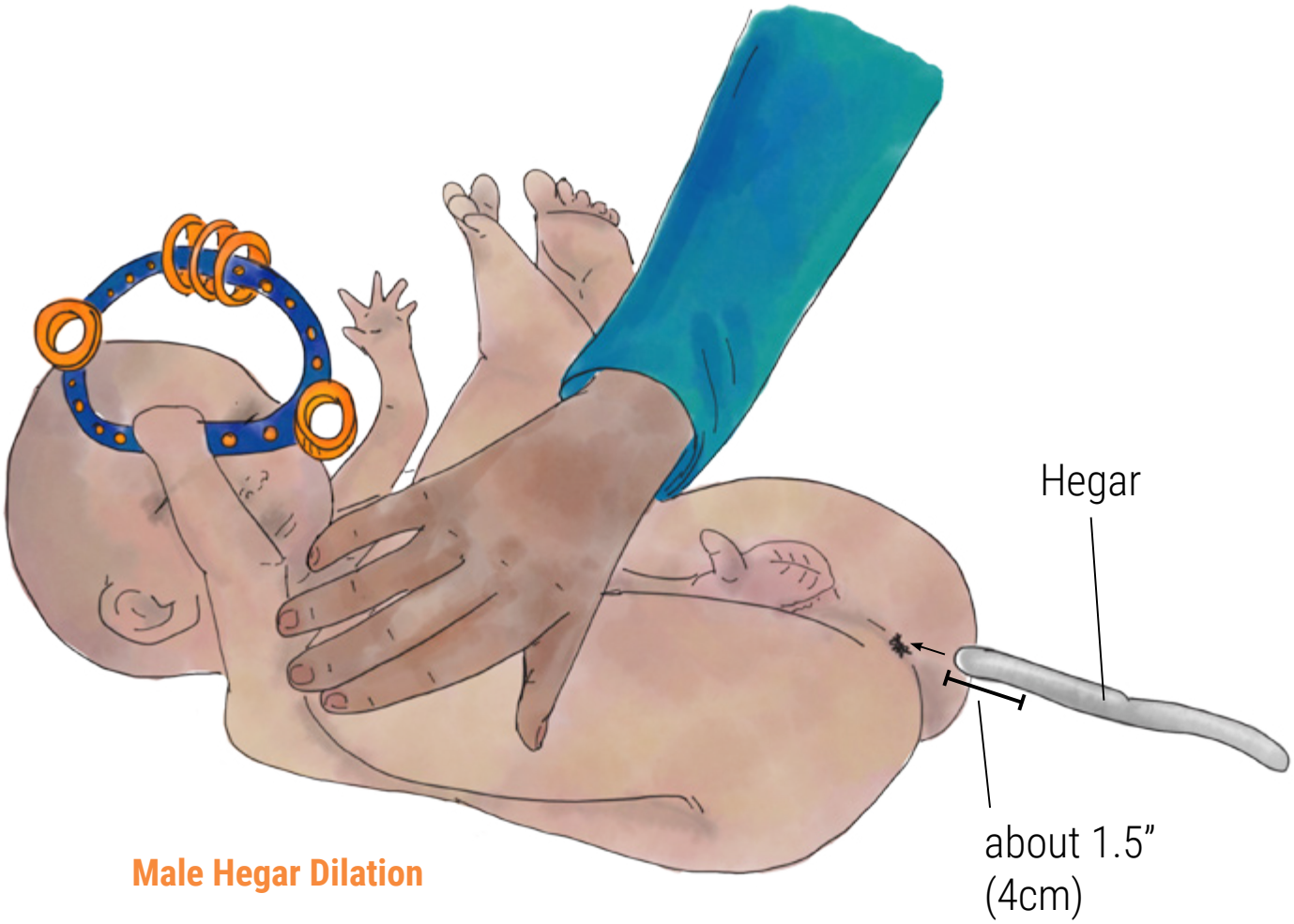
Preparing For The Dilation:

- The dilation will typically need to be done two times a day. One time in the morning and one time in the evening.
- Place your baby on their back on a flat surface. It is helpful to have a second person to help hold the legs up and over.
- Choose the right size dilator that your medical team said to use. The size is marked on each dilator.
- Apply water-soluble gel to the end of the dilator.
- The dilator is held in the hand that you write with like a pencil.

To Do Dilation:

- Use the hand that you do not write with to separate your baby's butt cheeks.
- Gently put the dilator into the fistula about 2 inches (50.8 millimeters).
- The dilator should go in smoothly with gentle pressure.
- Once the dilator is in place, spin the dilator between your fingers for 5 seconds.
- Remove the dilator and wash it with warm soapy water.

Below is an illustration of a male baby. The next page has a detailed illustration for female babies and it is important to make sure dilations are performed properly.

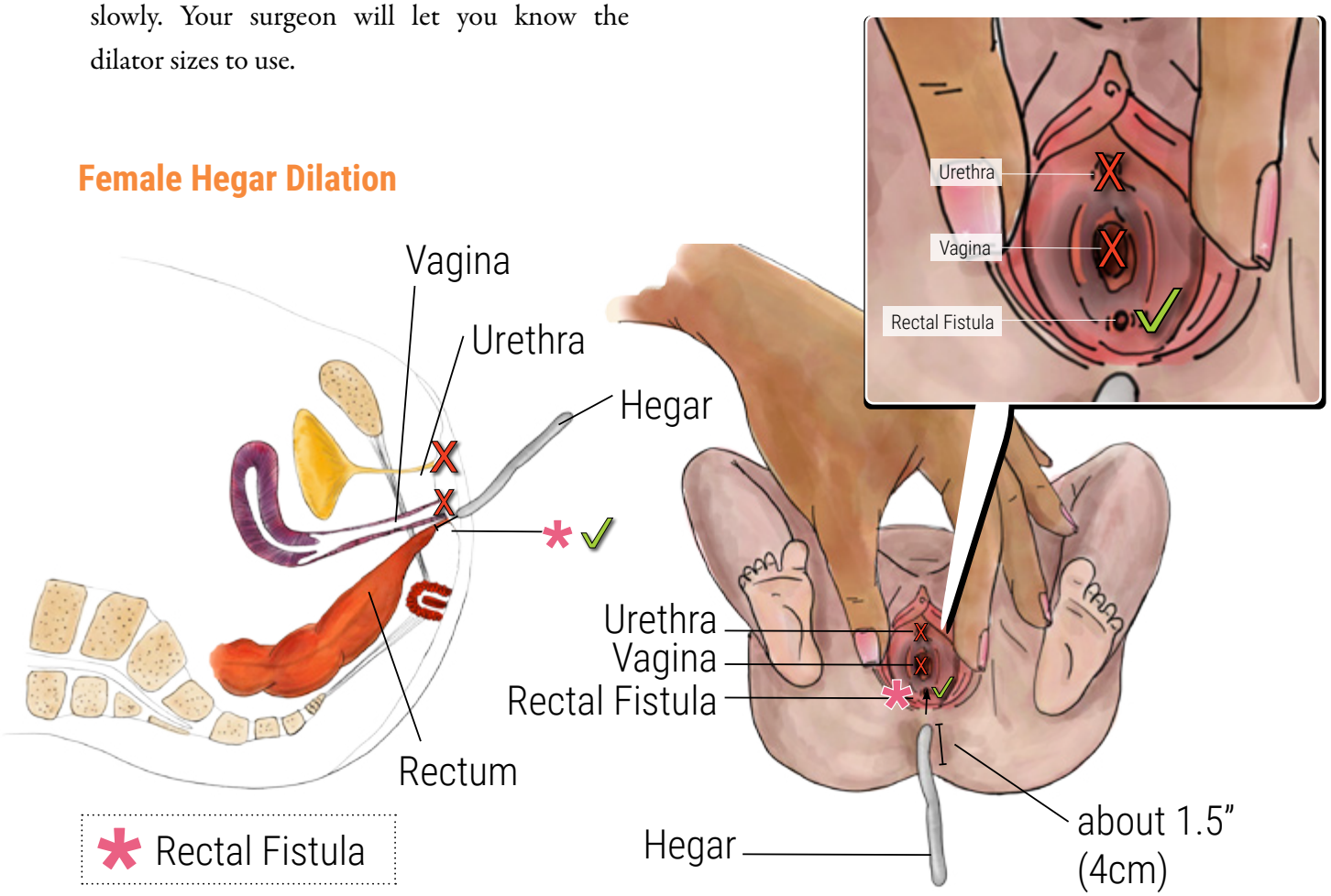


Male Hegar Dilation

Precautions:

- **If the dilator does not go in gently, do not force it.**
- If your baby is crying and bearing down, hold the dilator against the fistula with gentle pressure.
- When your baby takes a deep breath, put the dilator in. It should go in with ease.
- Often the dilator size needs to be increased slowly. Your surgeon will let you know the dilator sizes to use.
- When increasing the dilator size, use the smaller size first, then the larger size. Keep using both for the first day or two.
- When the larger size is easy to put in, you do not need to do the smaller size first.
- When you reach the final size dilator, decided by your surgeon, you will use that size until surgery.

Female Hegar Dilation



DO NOT place the Hegar into the urethra or vagina openings.
This could be very dangerous!

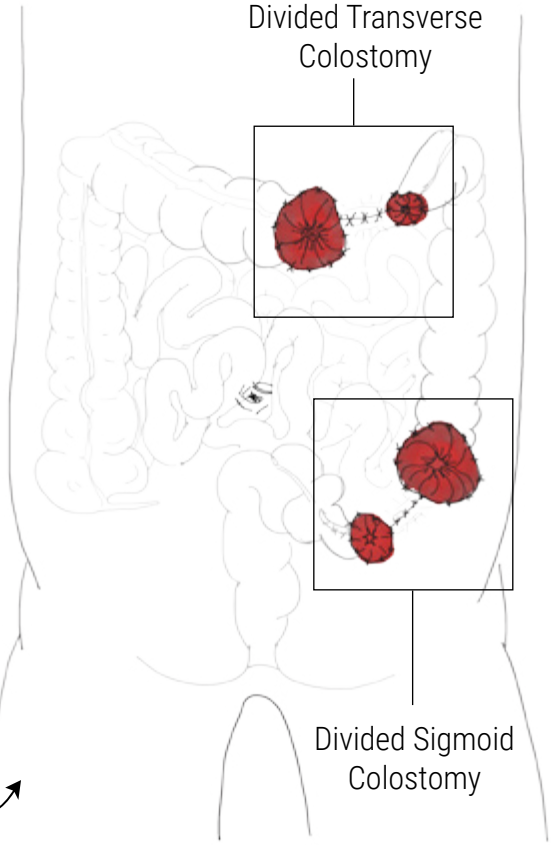
MAKE SURE to put the Hegar in the rectal fistula opening where poop comes out

9. Ostomy Care

Having the Right Team

Anorectal Malformation can be a complex diagnosis requiring specially trained health care professionals to help care for your baby. A team approach helps to coordinate the best care for your baby. **Pediatric Colorectal Surgeons, Urologists, Gynecologists, and Radiologists** are only a few of the team members that may be involved in your baby's care.

In addition to the doctors, **a Certified Wound and Ostomy Nurse (WOC)** provides support and teaches you about ostomy care, while you are in the hospital. The WOC also provides ongoing support to you and your family after discharge.



These are different ostomy types, where, and what they may look like.

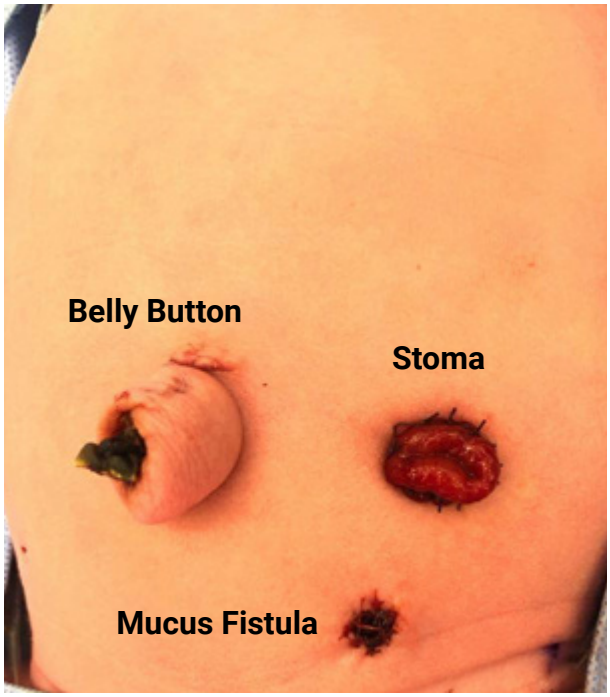
The Value of Having Wound and Ostomy Nursing Care

A certified WOC nurse holds a Baccalaureate degree or higher and has completed more education at an accredited wound/ostomy school. Wound and ostomy care is their specialty. Your WOC nurse knows how to solve any problems with the pouch and skin issues. A WOC nurse knows about different ostomy products and their use. Another benefit of having a WOC nurse is that they will help you choose products that will work the best for your baby over time. The WOC nurse can also act as a go between to help you communicate with the surgical team and supply company while your baby is in the hospital and after discharge.

You will be taught how to care for your baby's ostomy from a Certified Wound\Ostomy Nurse before going home from the hospital. This will prepare you to care for your baby's ostomy by yourself once you are home.

What to Expect Right After Colostomy Surgery:

As covered in Chapter 7, some children born with ARMs without an anal opening will need surgery soon after birth. An **ostomy**, which is an opening (**stoma**) from the colon inside the body to the outside, is done to allow poop to pass until the time comes for a new anus to be created.



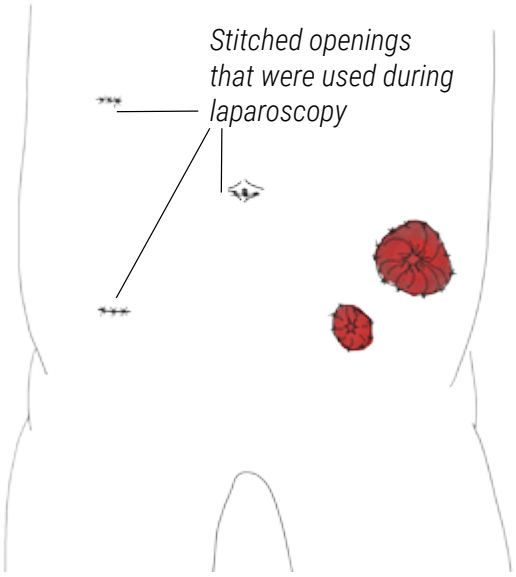
The stoma is the red budded end of the bowel or intestine that sits just above the skin's surface. This is where your baby's poop will now come out of the body and be collected in an ostomy pouch.

The other end of the bowel that goes down to the rectum is brought out to the skin separately and is called a **mucus fistula**. A mucus fistula only drains mucus and sometimes small amounts of urine depending on the type of ARM.

In general the stoma is larger than the mucus fistula.

When seeing your baby's belly for the first time, you may be nervous or scared. It can be stressful, so your nurse or other caregivers can be with you to help. Remember, most ostomies in babies are temporary.

- The stoma will be a beefy red color.
- The stoma may be swollen, which is expected.
- The size and shape of the stoma may change for up to 8 weeks.
- There may be stitches around the stoma. These will dissolve on their own.
- The stoma itself does not have feeling. Your baby may have some pain from the surgery, but the actual stoma does not have pain nerves.



Laparoscopy For Sigmoid Colostomy

Ostomy Care

Below is a guide to help you care for your baby with an ostomy. **Seek medical advice if you are unsure from your WOC nurse or medical team.**

There are two main parts of an ostomy device

- The **wafer or barrier** that sticks to the skin
- The **bag or pouch** that collects the poop

The **one-piece type** has the pouch attached to the wafer. A t**wo-piece device** has a wafer or sticker that attaches to the skin and a pouch that then connects to the wafer.

Your WOC nurse will help decide if your baby should use a one-piece or two-piece device. One of your WOC Nurse's goals is to find a device that is strong and long-lasting.

We like to use the simplest device that is the best for your baby to keep the poop in the pouch and protect the skin around the stoma.

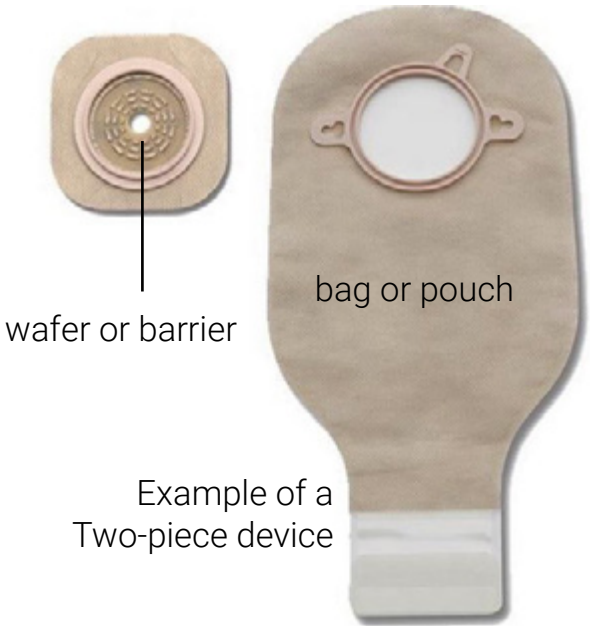
Applying the Pouch and Wafer

You will need to change the pouch and wafer every 2 to 3 days. However, there may be times when the pouch leaks and needs to be changed sooner.

If the device needs to be changed more than one time a day, call your WOC nurse for help.

Pouch and Wafer Care:

An ostomy pouch or device holds your baby's poop. There are a few types of devices, both one-piece and two-piece sets.



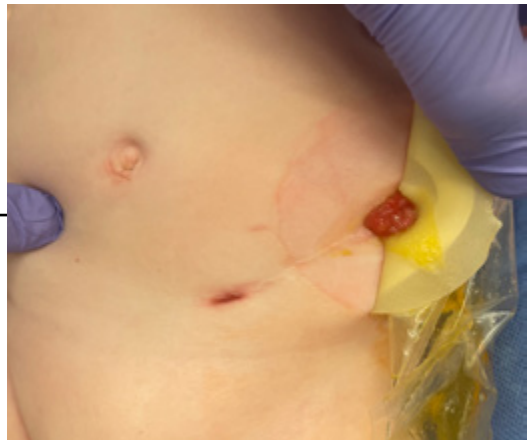
Example of a
Two-piece device

Example of a
One-piece device

Steps to Change the Ostomy Device

1. Gather all supplies and then empty the ostomy pouch.

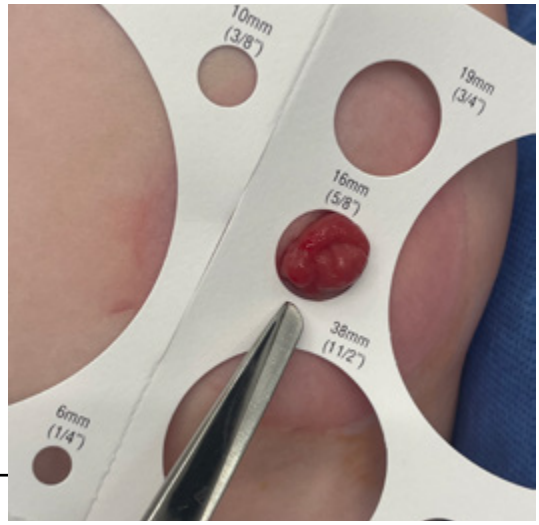
2. Remove the wafer or barrier by gently peeling up the wafer's edge while pushing down on the skin. This helps to separate the sticky side from the skin.



3. Wash the skin with water and dry it using a washcloth or paper towel. **Do not use soaps, baby wipes, and lotions on the skin.** They can create a film that may keep the wafer from sticking to the skin.

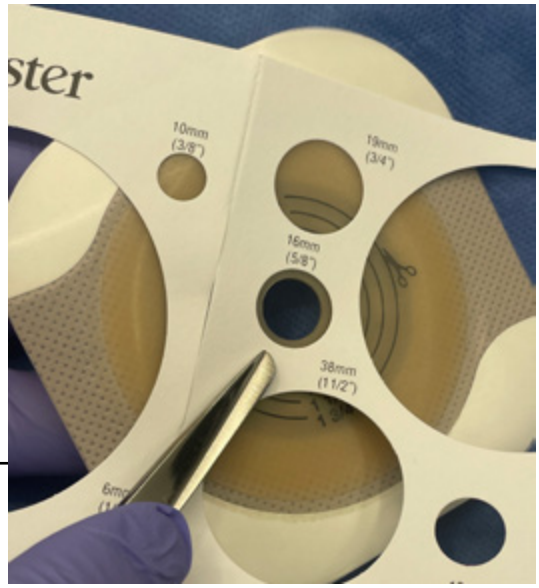
4. Use an alcohol-free skin wipe with a barrier to protect the skin. Allow the area to dry well before putting on the wafer.

5. The stoma should be measured using the measuring guide that is in with the supplies. Stoma size may change over the next 6 to 8 weeks as the swelling goes down. **Place the measuring guide over the stoma to choose the correct size of the opening.** **Allow an additional 1/8 inch (0.03cm) around the stoma.**



6. Place the measuring guide on the back of the wafer to trace the correct size for the opening.

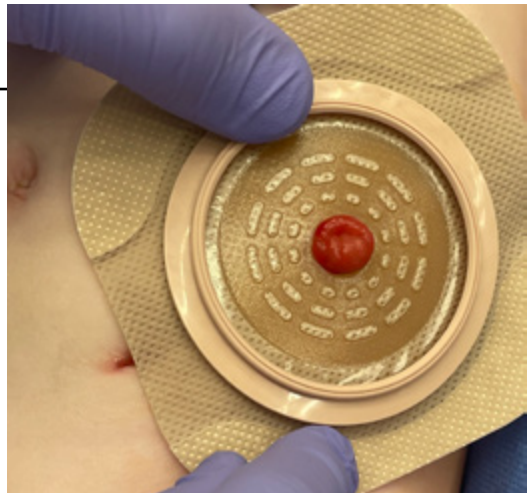
7. Cut the traced hole in the wafer following your template. **This should match the stoma's size plus the additional 1/8 inch (0.03cm).** Smooth any rough edges with your finger that you have created by cutting the opening.

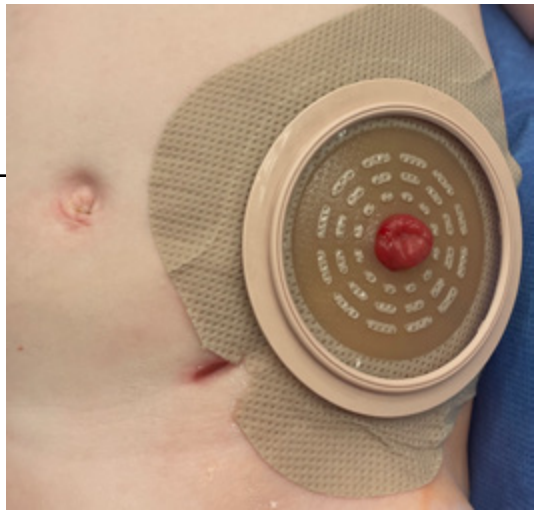


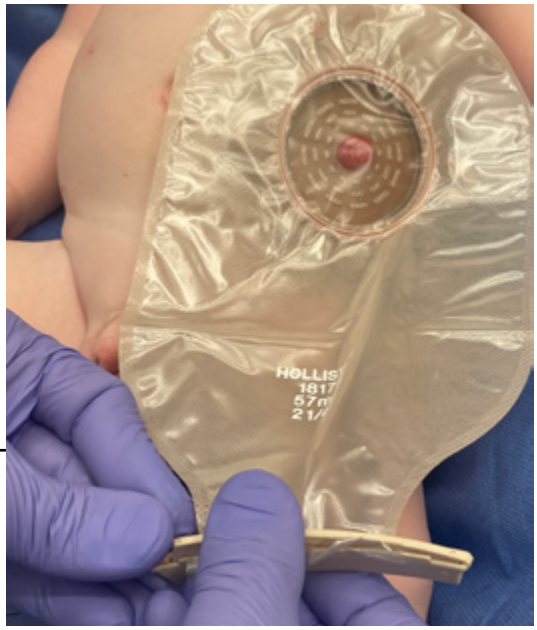
8. Check the fit of the hole you cut against the stoma to ensure proper fit. Peel the paper backing off and apply the wafer and pouch to your baby's skin.

9. If the ostomy wafer overlaps with your child's mucus fistula, cut out a small part of the wafer so it doesn't cover the fistula.

10. The ostomy bag should have a good seal all around. Make sure the bag is closed so no poop leaks out.







Helpful Tips

A WOC nurse will decide if any special products will need to be used, based on your baby's stoma and belly.

- You should empty the ostomy pouch when it is half full of poop or gas. You may want to check the pouch at each diaper change.
- A flat surface allows for the pouch to stick better to the belly. You can use **barrier rings** or **strips** to smooth the surface of your baby's belly by filling these creases.
- If the stoma becomes **weepy and red,** you can use a powder on the skin to create a barrier around the stoma. The powder provides a dry surface for the wafer to stick to. Apply powder on your hand then on the baby. **Do not shake the can of powder on your baby to keep them from breathing in powder.**
- **The stoma does not have a muscle to stop the poop from coming out while the pouch is off.** The stoma may need to be covered with a cloth to help keep the poop in while you cut the new wafer.

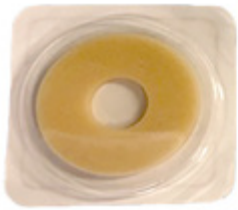
- There may be less poop if you change your baby before you feed them.
- Although a wafer and pouch change is not painful, your baby may cry while you are changing the pouch. They often cry because they are being held still or don't like to have their clothes off. Until you feel comfortable with changing the pouch, it may help to have someone comfort your baby and help them to stay still.
- It is important to carry extra ostomy supplies in the diaper bag.

Diet

Babies with a stoma can generally have a regular diet. Your team will talk with you about the diet and will check your baby's weight.

Bathing/clothing

- Your baby should be sponge washed until their umbilical cord has fallen off. Once the umbilical cord is off, you can bathe them in a tub with the ostomy device on or off.
- Soap and water will not hurt the stoma. However, be sure to rinse off all soap and water if you are bathing your baby without the pouch.
- Avoid using lotions or oils. These may interfere with how well the wafer sticks to your baby's skin. If you leave the pouch on during bath time, make sure to dry the wafer's edges to prevent moisture against the skin. Moisture left on the skin increases the risk of rashes.
- One-piece sleepers or "onesies" help prevent your baby from finding the pouch and pulling it off.



Activity

Your baby can do all the normal activities a baby should do at that age of development.

You should use an infant car seat when traveling.

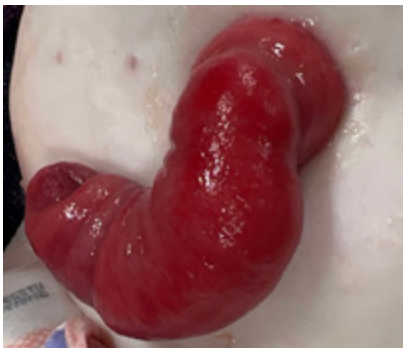
Tummy time is allowed at the age when it is normal to do this. Empty the pouch before tummy time.

Problems that can occur

There are some problems that can occur when your baby has a colostomy. Call your surgical team if any of these occur.

Prolapsed stoma

This is when the stoma becomes larger or longer than it has been. If the stoma stays beefy red and brings out poop as usual, it is not an emergency. Call your surgeon or WOC nurse to let them know this occurred and if you need help with putting on the pouch. **If the stoma is purple or black and there is no poop, or your infant is not acting right, call your surgeon and go to the Emergency Room right away.**



Retraction

Stoma retraction is when the stoma is at skin level or below. It may look like the stoma is shrinking in. Retraction can cause a problem with keeping the pouch on, as the poop gets under the wafer. If the stoma keeps putting out poop, it is not an emergency. Call your WOC nurse to help adjust the device. If there is no poop coming from the stoma, call your surgeon.

Yeast or Rash

A red rash under the ostomy wafer is not normal. Irritation or skin breakdown can affect the seal of the wafer. This leads to changing the pouch often. If there is redness and small red dots under the wafer, this could mean there is a fungal rash. If there is redness and the skin is raw looking or open directly around the stoma, it may be from poop on the skin. Use your measuring guide to ensure the proper size and fit of the wafer. Call your WOC nurse for help with treating the rash and skincare right away.

Bleeding

Your baby's stoma has a good blood supply. It is normal to see small spots of blood when cleaning or changing the pouch. **It is not normal to have a large amount of bleeding that does not stop from the stoma into the pouch. If this happens, go to the Emergency Room.**

Family Resources

Your WOC nurse is your first resource to answer your questions and help solve any problems you are having. Other resources you can use include:

Pediatric Ostomy Home skills kit, American College of Surgeons:
www.facs.org/patienteducation

Wound, Ostomy and Continence Nurse Society (WOCN): www.wocn.org

United Ostomy Associations of America:
www.ostomy.org

Medical Emergencies

Some problems are Medical Emergencies. If your baby has any of the following, call your surgeon and go to the emergency room:

- The stoma color is black or purple
- Vomiting and no poop for 12 to 24 hours
- Signs of diarrhea and dehydration, which include emptying the pouch more often of liquid poop and having fewer wet diapers a day.

10. Male PSARP Surgery

Posterior sagittal anorectoplasty (PSARP) is a surgery used to correct ARMs. It is the standard way to do ARM surgery.

This surgery creates an anus placed within the muscles that allow for the opening and closing of the anus, called the **sphincter muscles**.

If your child had a **colostomy** done before the PSARP, the surgeon will usually leave it in place after the PSARP. The colostomy protects the area where the surgery was done from poop and allows for the best healing. The colostomy can be closed 6 to 8 weeks after surgery if the area has healed well. (See Chapter 15)

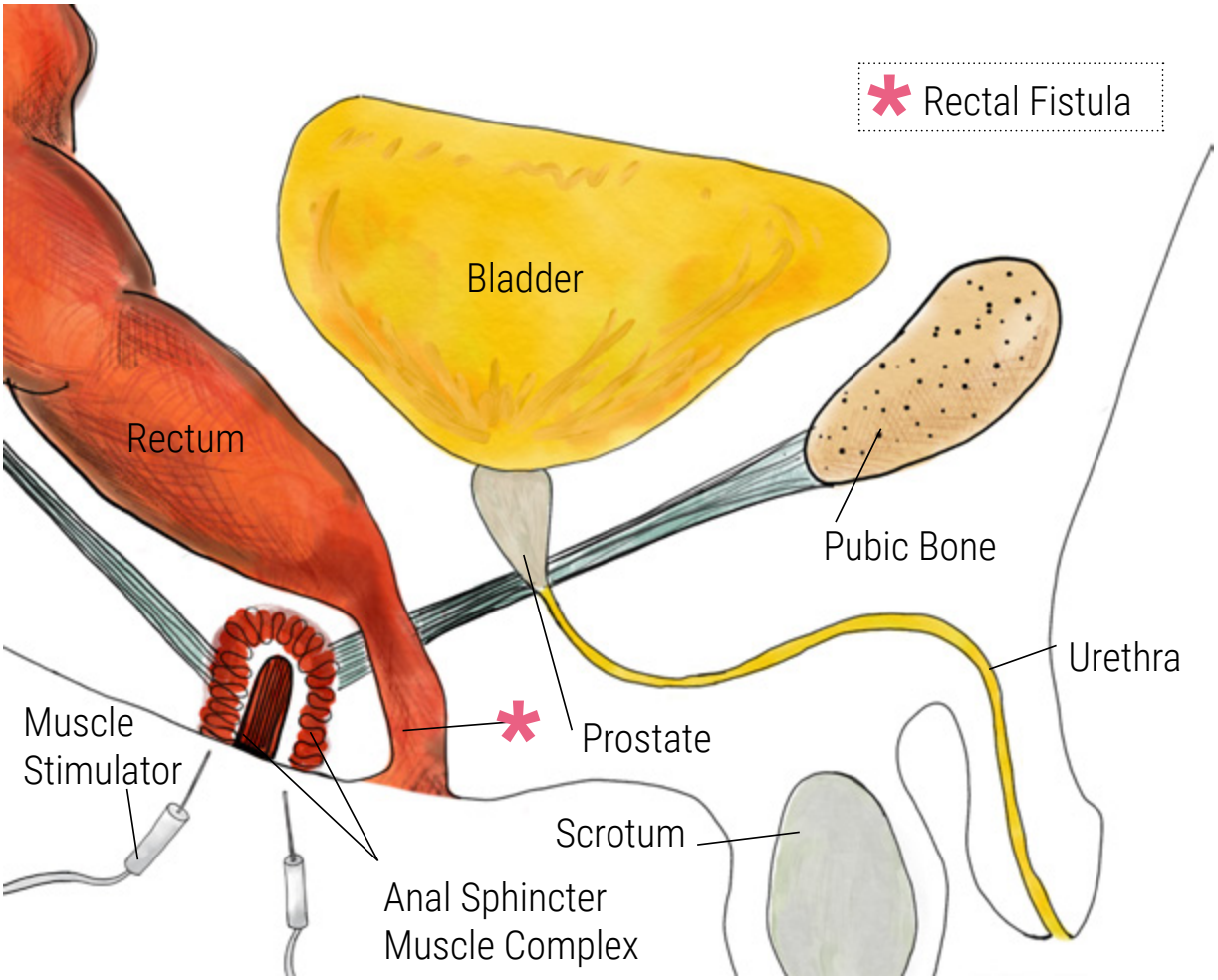
This section focuses on male PSARP surgery for perineal fistula, rectobulbar, recto-prostatic, and rectovesical fistula).

Chapter 11 is on female PSARP surgery, and Chapter 13 is on cloacal malformation surgery.

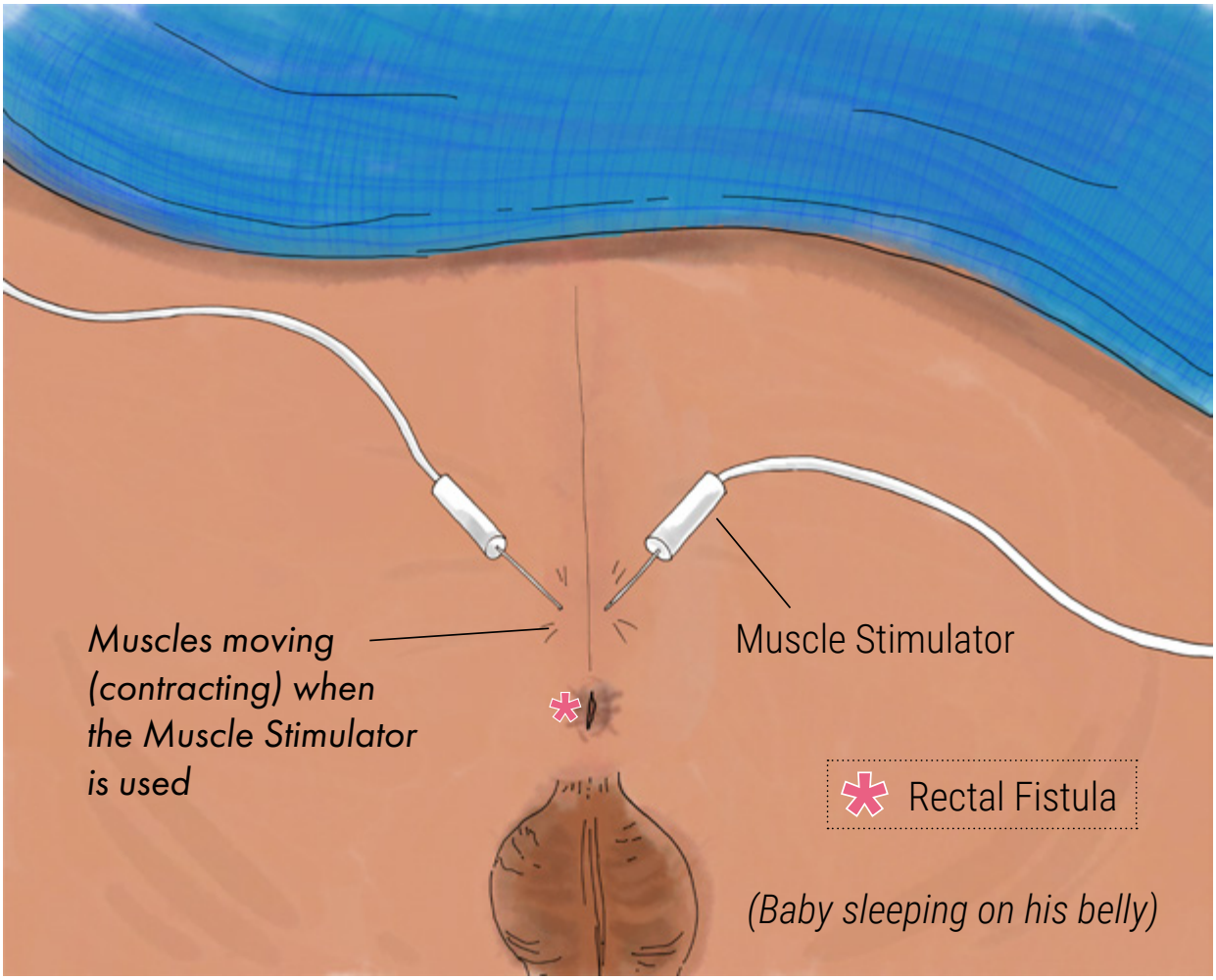
Remember, this book is a guide, building a trusted relationship with your child's surgical team is always recommended.

PSARP for Perineal Fistula

As you read in Chapter 2, a perineal fistula is where the anus opening is in the wrong place and often too small to allow poop to pass easily. Dilations help enlarge the opening until surgery is done. (See Chapter 7). During PSARP surgery for perineal fistula, your baby's muscles that control poop, or the external sphincters, are found using a stimulator. The anal opening is moved to this site to place the muscle around it. This helps control how poop comes out. The opening is made large enough to allow poop to pass. Sometimes, dilations are still needed to avoid scarring that can narrow the anal opening after surgery. Usually, after a few months, the dilations are stopped.



In this picture, we can see the perineal fistula in front of the sphincter muscle complex. This is a sideways view of your baby's anatomy. Your surgeon cuts the fistula from the skin so that it can be put in the middle of the sphincter muscles.

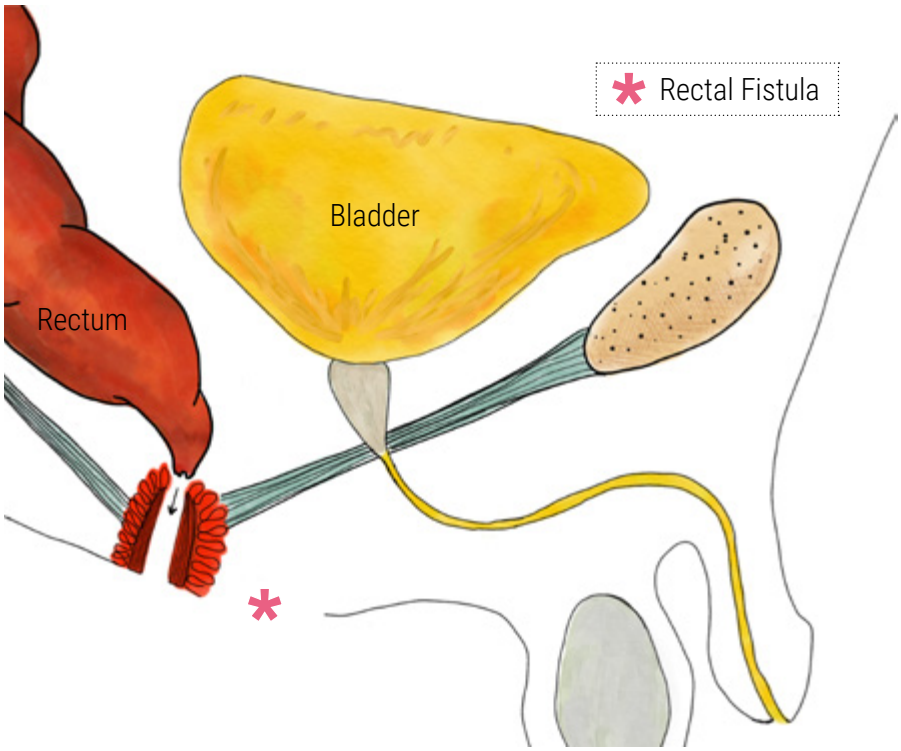


This is the outside view of your baby's bottom while laying on their belly. To find the right place for the anus, a special device, called a muscle stimulator, is used on the outside of the skin.

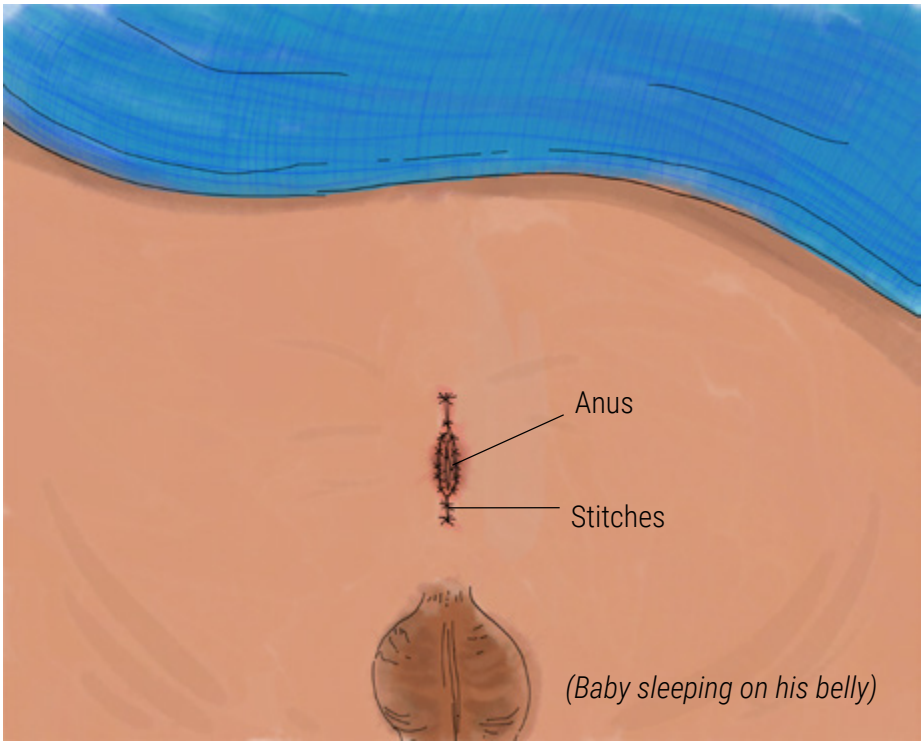
The stimulator makes the sphincter muscle move and contract. This allows the surgeon to know where the anus is supposed to be and to make the right cut in the middle of the muscle. This is very important to make sure that the anus works correctly, so that your baby has a better chance to control their poop when it comes to potty training in the future.

After the cut is made in the middle of the sphincter muscle, the end of the colon is relocated here and stitched to the skin to recreate the new anus.

The original fistula opening is stitched closed.



In this outside view, you can see the stitches and the new anus.



PSARP for ARM with Rectourethral Fistula

As noted in Chapter 2, where there is a missing anus in boys (**anorectal malformation with rectourethral fistula**), the rectum connects to the urinary tract instead of connecting with the anus.

This connection, or **fistula**, can go to the tube that carries urine from the bladder through the penis (urethra), or to the bladder neck itself.

When the connection is to the lower part of the urethra (**rectobulbar fistula**), it is called a **"low", or "less complex" ARM.**

When the connection is to the upper urethra (**rectoprostatic fistula**) or to the bladder neck itself (**rectobladder neck fistula**), they are called **"high" or "complex" ARMs.**

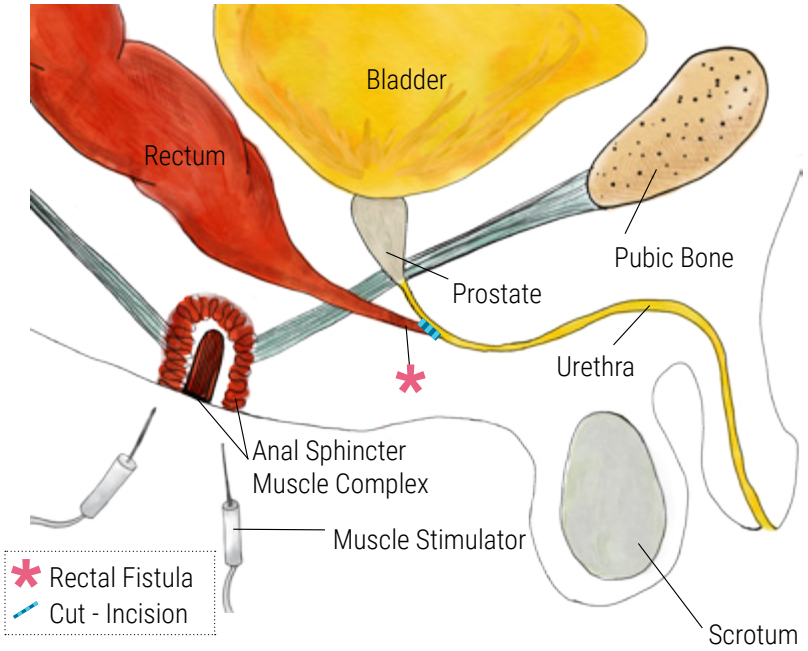
For a "low" ARM, the PSARP surgery is often done from your baby's bottom without the need to enter the belly.

In the next few pages, you will see two examples of PSARP surgery for rectourethral fistula.

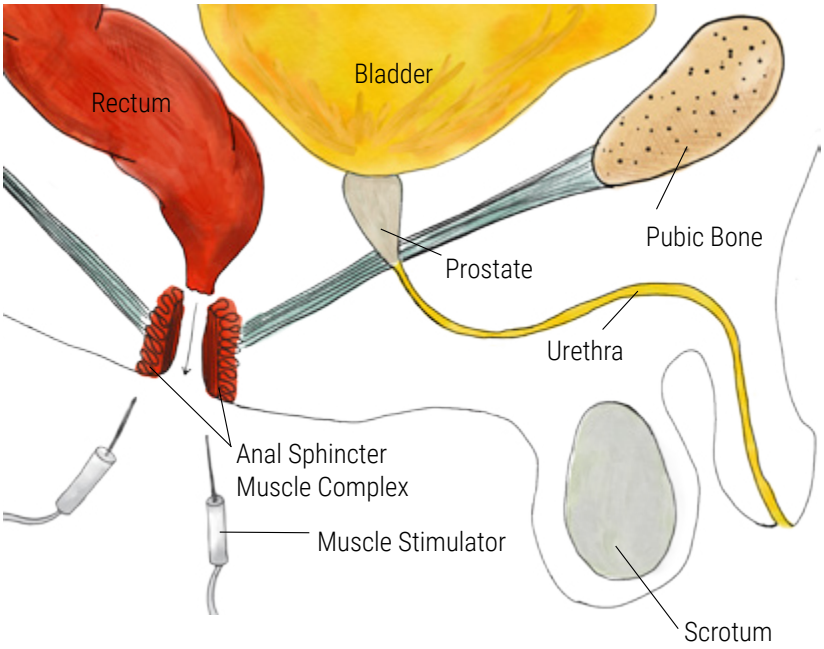
PSARP for Rectobulbar Fistula

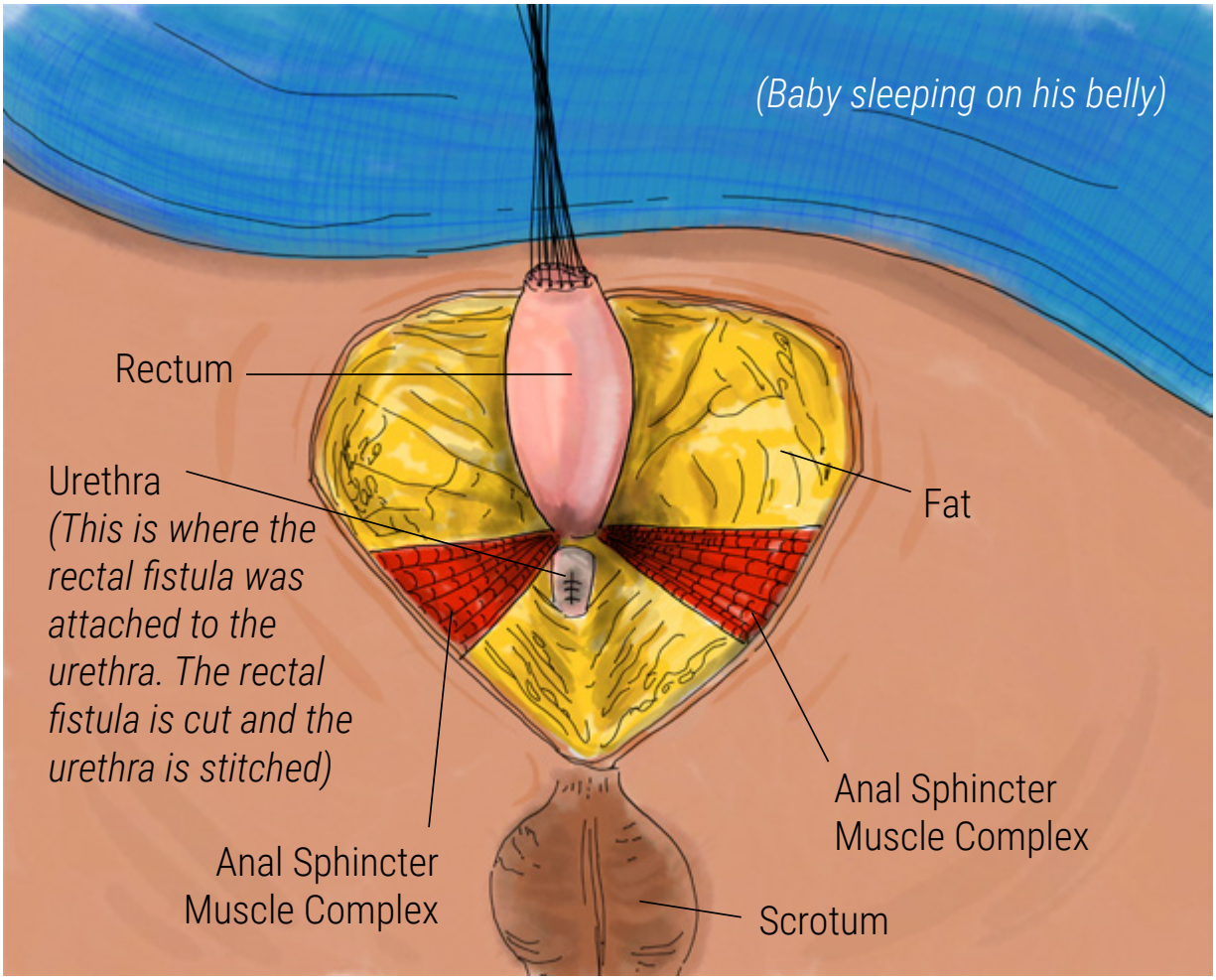
First, your baby's muscles that control poop, called the external sphincter muscle complex, are found using a stimulator.

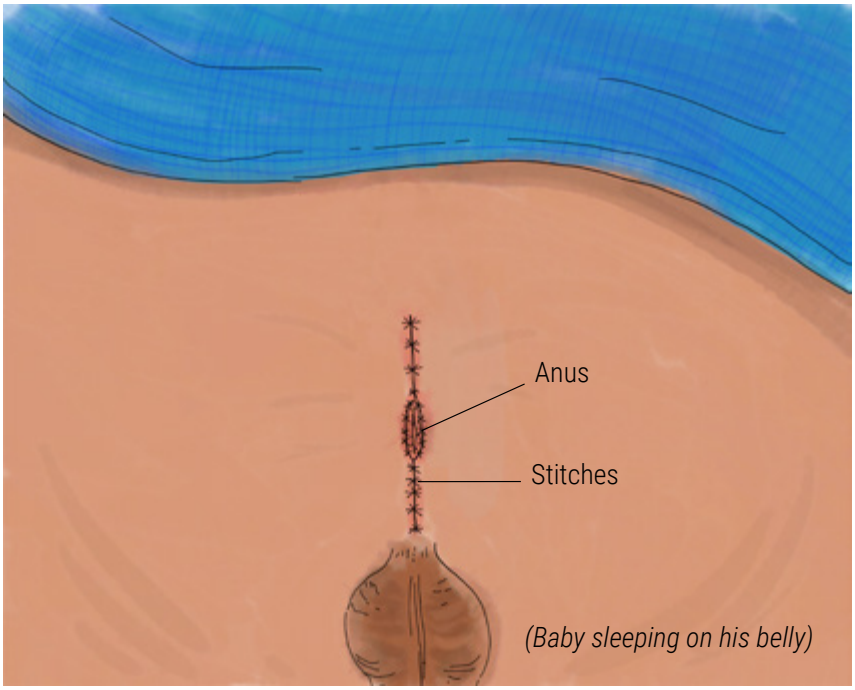
A cut is made from below the scrotum to near the tailbone. The rectum is found and the fistula that enters the urethra is dis-connected. The hole in the urethra is repaired and the rectum is then brought down to the skin level.



The rectum then will be surrounded by the external sphincter muscles in the correct location.

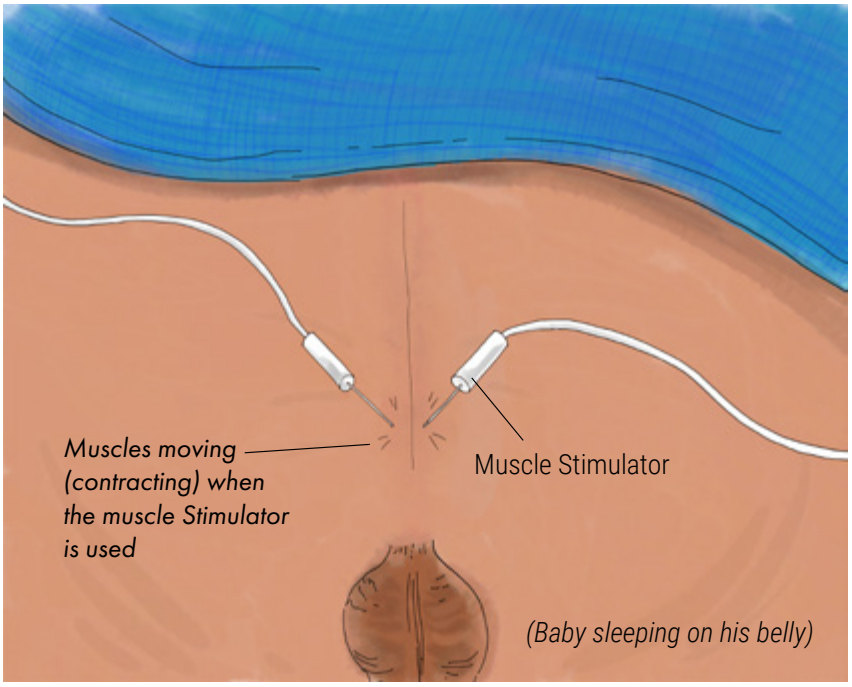




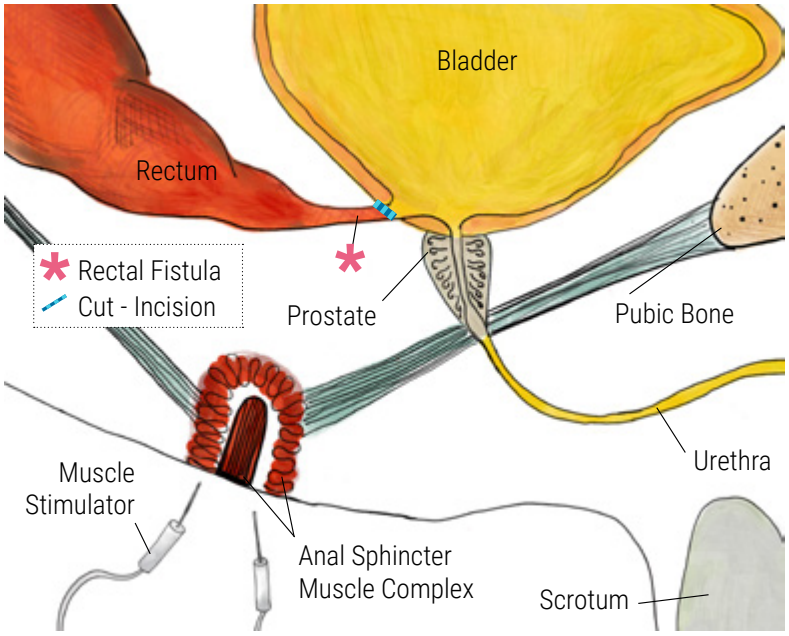


The opening is stitched up and a new anus is formed.

For a "high" ARM where there is a connection to the upper part of the urethra or bladder neck, your surgeon will likely do the surgery through the belly and from the bottom. Your surgeon may decide to do the surgery with one larger cut in the belly or will do it laparoscopically. Laparoscopic surgery is where small tubes with a camera are placed through tiny cuts in your baby's belly. This avoids a larger cut at the bottom.

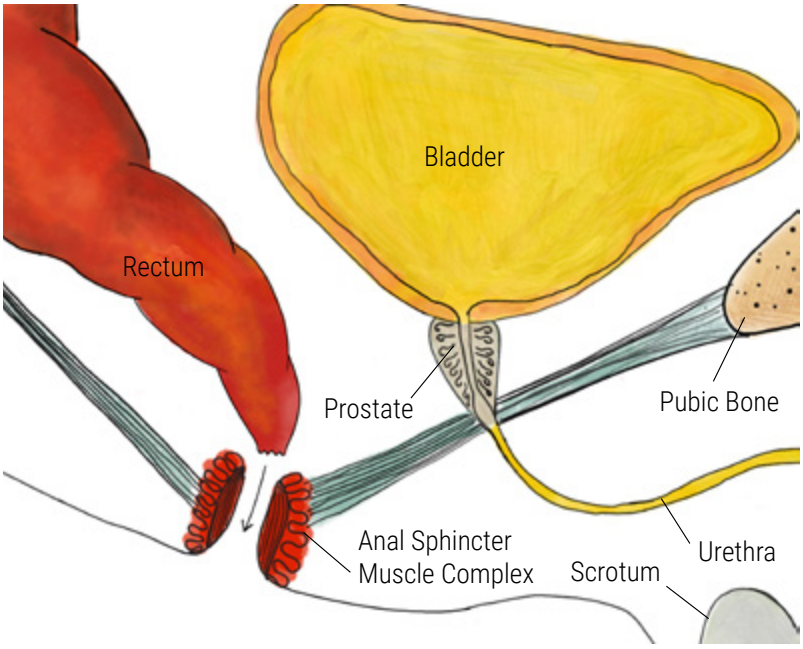


The first step is to use a stimulator to find the muscles that control poop from leaving the body (external sphincter).



Next, the surgeons will locate where the rectum connects to the urinary system and disconnect it.

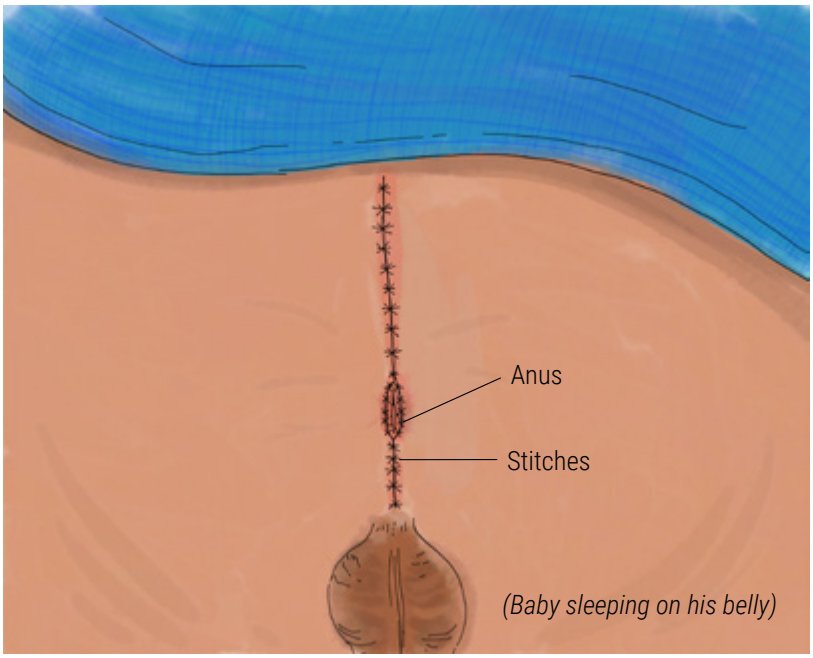
This is done either through a cut in the belly or by using small tools that are put into the belly through small cuts (laparoscopy).



The hole in the urinary system is repaired, and the rectum is then brought down to the skin level.

The rectum then will be surrounded by the external sphincter muscles in the correct location.

If laparoscopy is used, the cut at the bottom can be kept small since most of the repair was already done in the belly.



This is what the new anus looks like after surgery. The stitches and wound will heal overtime.

PSARP for ARM without Fistula

Sometimes, your baby has an ARM where the rectum does not have a connection to the urinary tract. This is called an **anorectal malformation without fistula**. As you learned in Chapter 7, your baby's first surgery will be a colostomy to divert the poop. This surgery is done until they are a few months older for a bigger surgery. The second surgery is the PSARP to create the anus. Like the PSARP surgery noted above, the first step is to use a stimulator to find the muscles that control poop from leaving the body (**external sphincter**).

Next, the rectum is brought down, to form the anal opening at the skin level. It will be surrounded by the external sphincter muscles in the correct location.

After PSARP Surgery

Your baby will be in the hospital after surgery so your team can check healing from the surgery. Your baby can usually start eating right after surgery if a colostomy was done. If your baby has a perineal fistula or rectovestibular fistula and does not have a colostomy, they will need to wait to eat. This allows for the best healing to take place. Some surgeons will allow your baby to have clear liquids, such as **Pedialyte**® or breast milk, for 5 days while the repair heals. Other surgeons may want your baby to not have anything by mouth during this time. Some surgeons may want a thin tube to be put into a vein in the arm that goes to larger veins near the heart. This is called a **PICC line** and is used to give nutrition into the vein. During this time, your team will check on the repair to make sure it is healing well. Swelling does occur and it can take a week or more to go down.

Problems with PSARP Surgery

Problems are not common with PSARP surgery.

After your baby recovers from the PSARP "surgery, the surgeons will check that the anal opening is a right size. The anal opening may get smaller in about a quarter of patients as the body heals and creates a scar. It is easily treated with dilations that can be used less often as the anus reaches the right size. (See Chapter 8 on dilations).

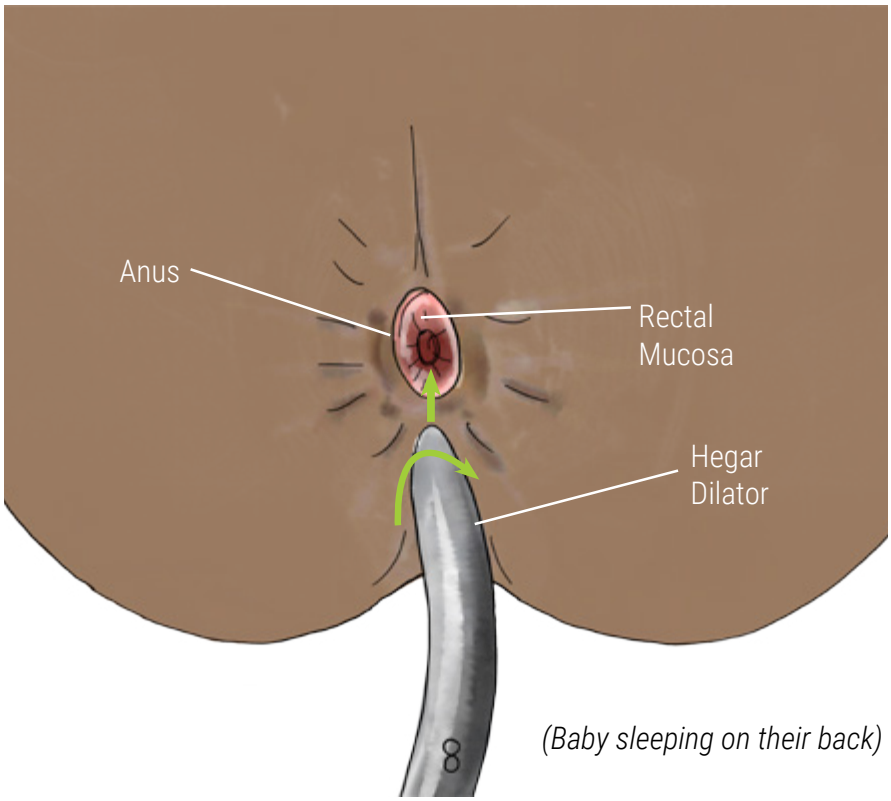
There is a small risk of infection of the repair. If this happens, sometimes **antibiotics** are all that are needed to treat the infection.

However, your baby may need another surgery if the infection leads to the skin and repair separating. If your baby did not have a colostomy before the PSARP and has a lot of problems with the repair, they may need a colostomy. **A colostomy will allow the repair to heal properly.**

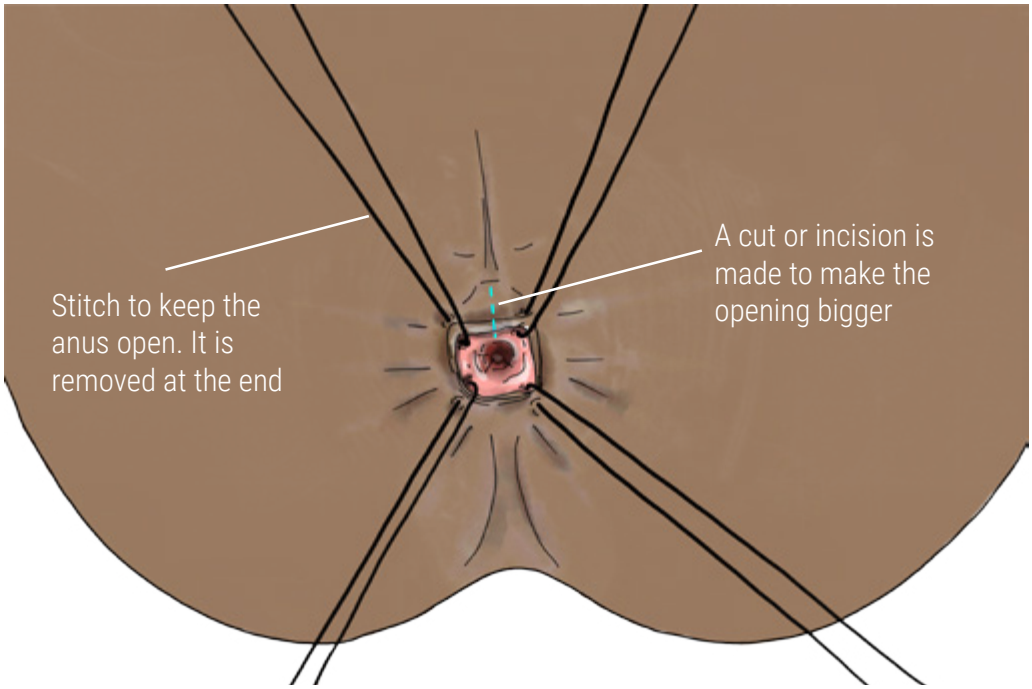
Heineke Mikulicz Anoplasty

Some surgeons do not use dilations. If the anus becomes narrow, a minor outpatient surgery (**Heineke Mikulicz Anoplasty**) can be done to release the scar so there is a larger opening.

First, the surgeon checks the size of the anus using a device called a Hegar dilator.

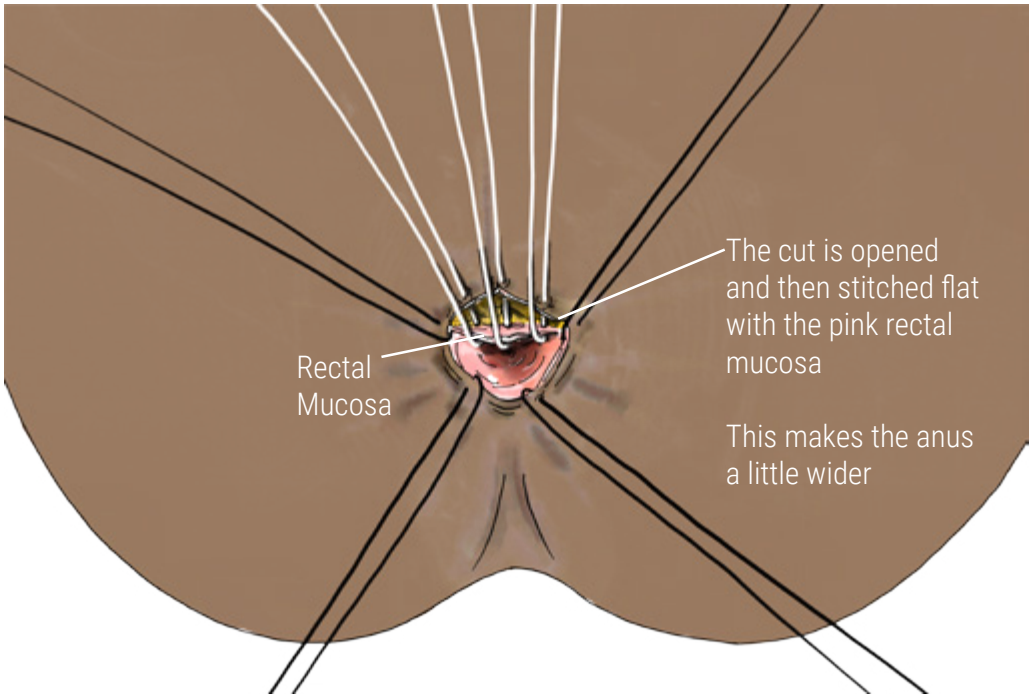


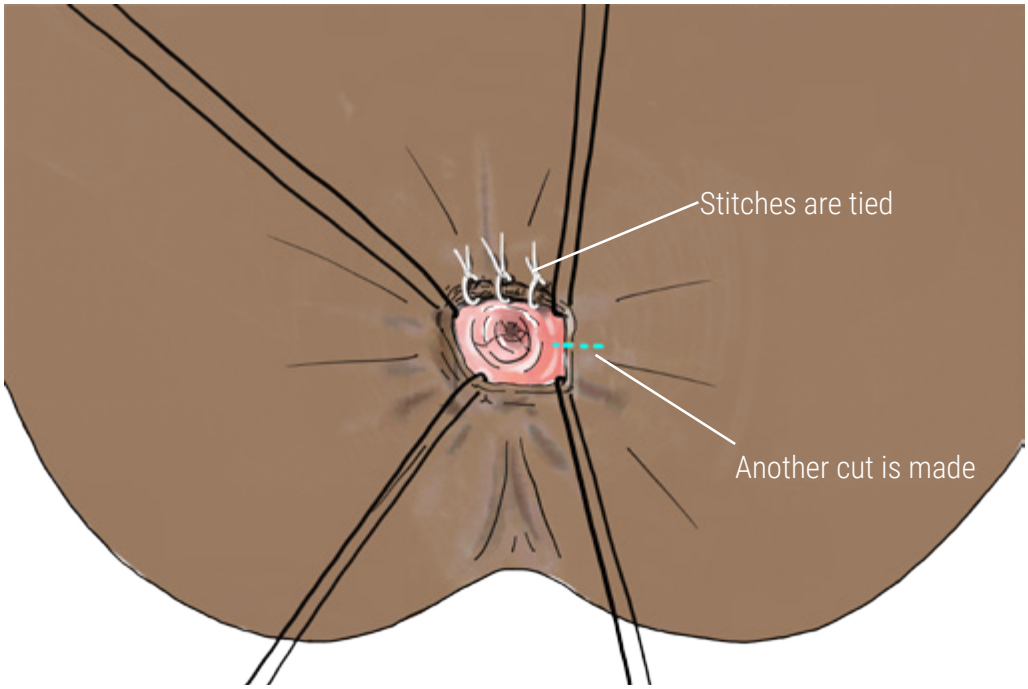
(Baby sleeping on their back)



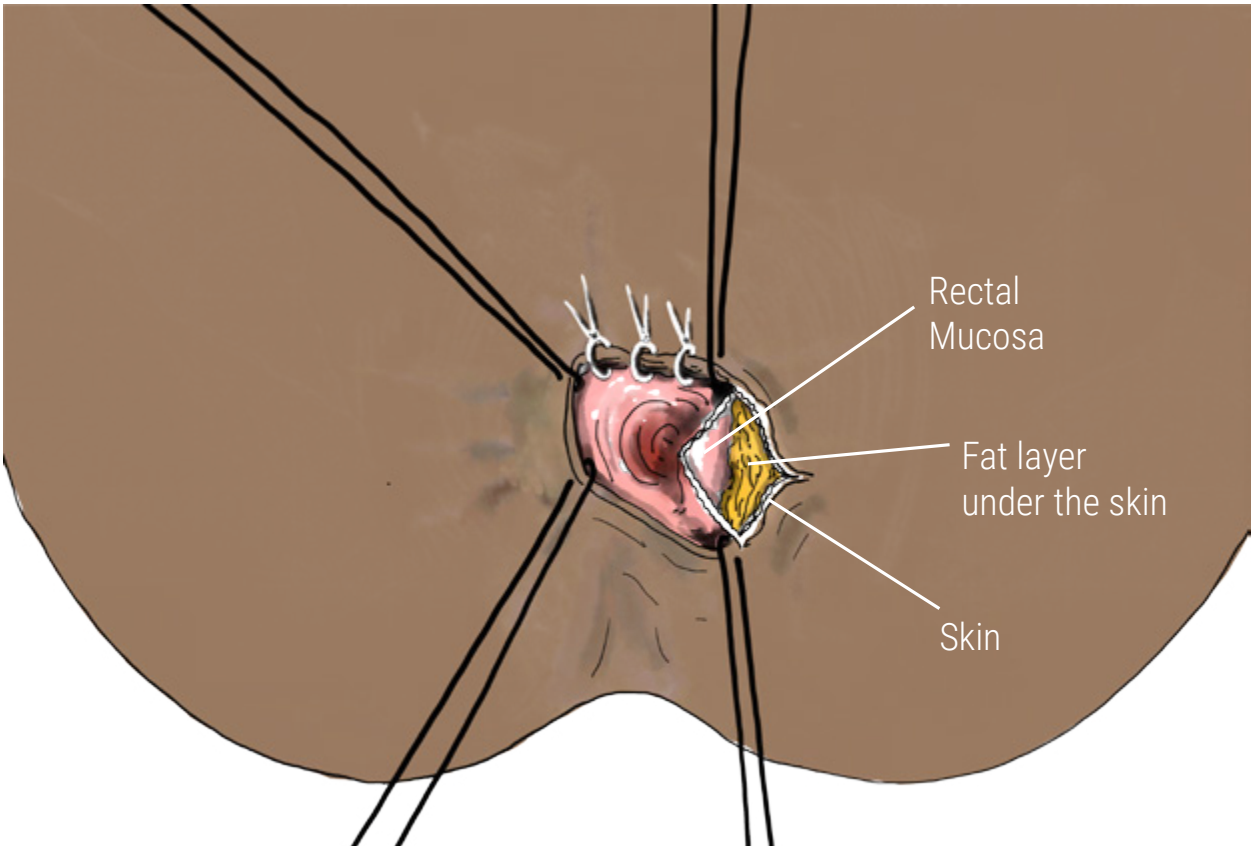
A small cut is made at the top of the anus creating an upside-down V shape. This makes the anus wider from the top.

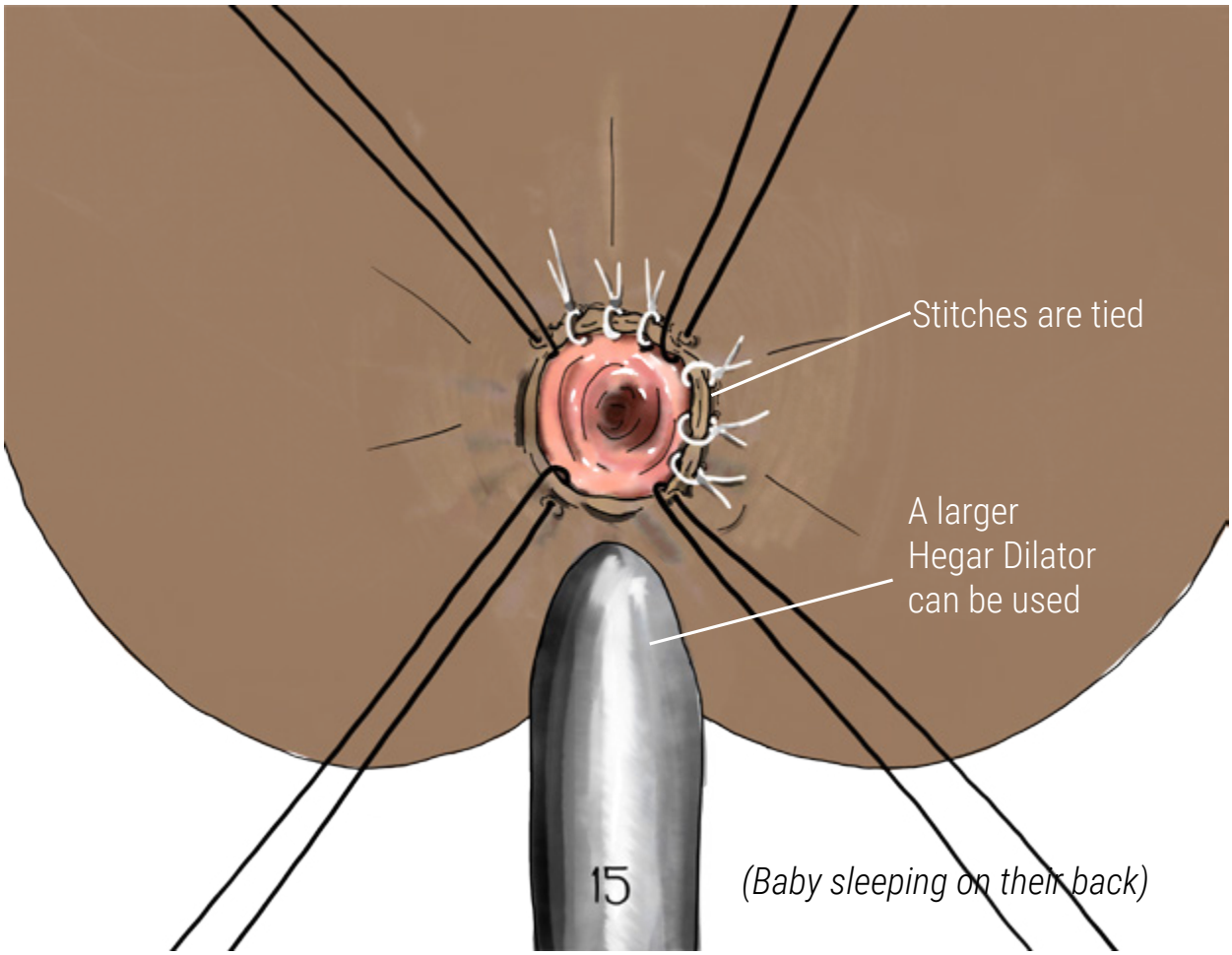
The V is stitched to make a flat line so that the skin and inside of the rectum are stitched together.





The same is done on the right side. The cut this time is horizontal, and a sideways V cut is made.





The skin and tissue are then stitched together in a flat line to make the opening of the anus wider.

A larger Hager is used to know the size of the repaired opening.

If the anus is very tight due to extra scar tissue or poor blood supply to the area, your baby may need another PSARP surgery.

11. Female PSARP Surgery

Posterior sagittal anorectoplasty (PSARP) is a surgery done to correct ARMs. It is the standard way to do ARM surgery. This surgery creates an anus that is the best size for your baby. It is placed within the muscles that allow for the opening and closing of the anus, called the **sphincter muscle complex.**

If your child has a **colostomy** done before the PSARP, the surgeon will leave it in place after the surgery. The colostomy protects the area where the surgery was done from poop and allows for the best healing. The colostomy can be closed 6 to 8 weeks after surgery if the area has healed well
(See Chapter 15).

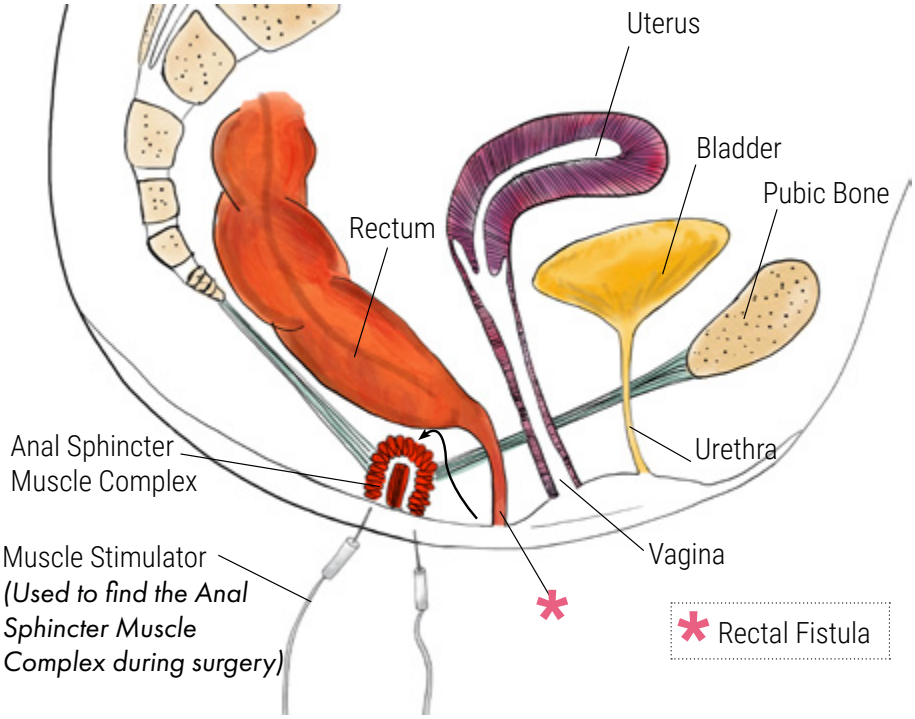
This section focuses on female PSARP surgery for Perineal Fistula, Rectovestibular Fistula, and Rectovaginal Fistula.

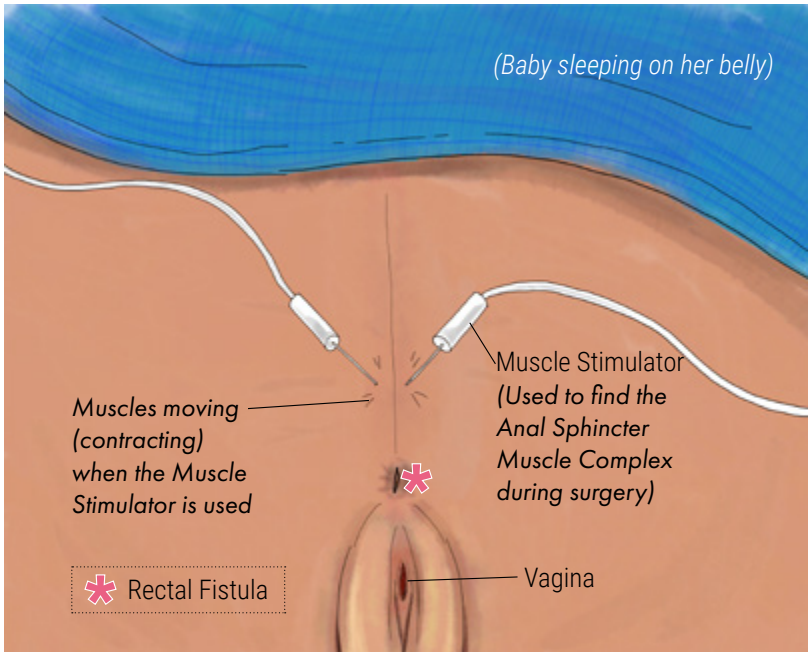
Chapter 10 is on male PSARP surgery, and Chapter 13 is on cloacal malformation surgery.

Remember, this book is a guide, building a trusted relationship with your child's surgical team is always recommended.

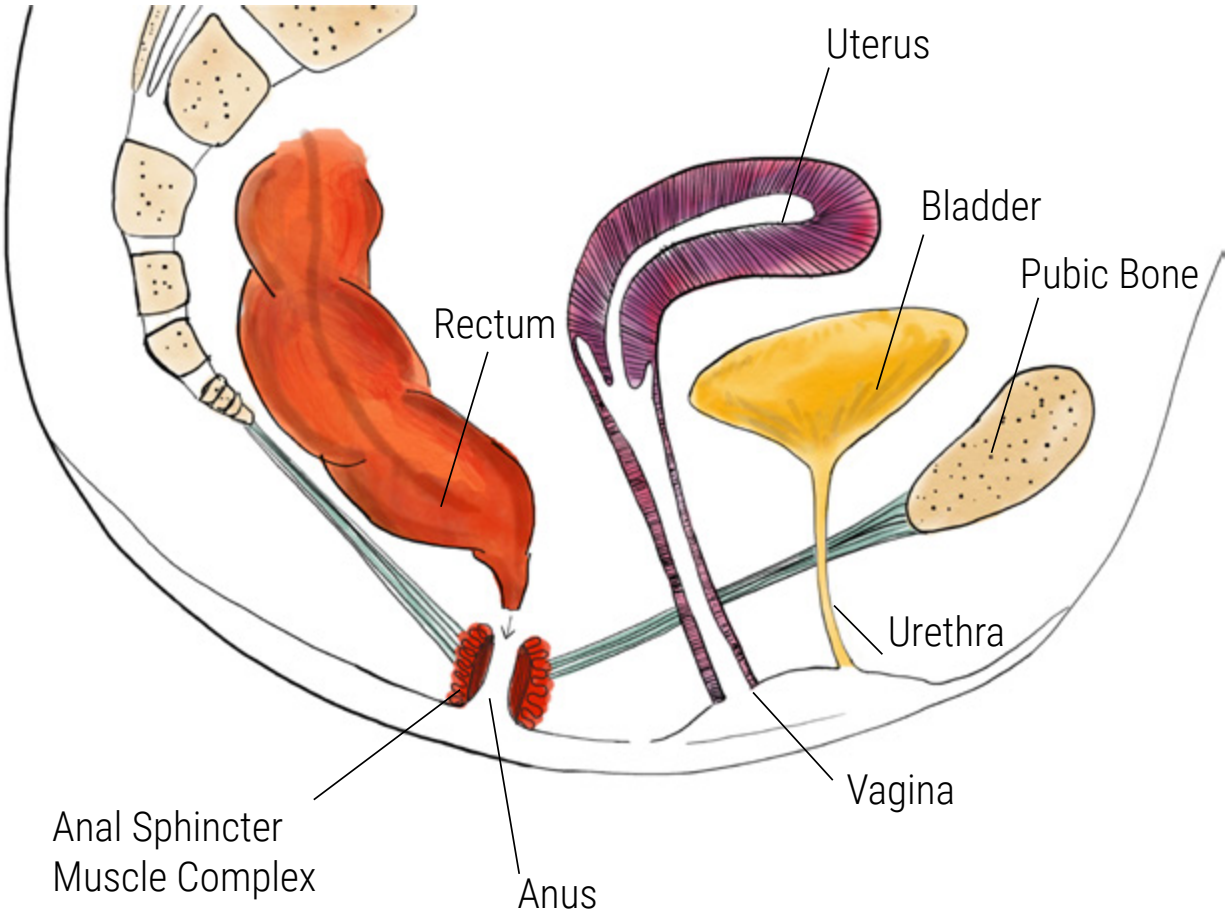
PSARP for Perineal Fistula

As you read in Chapter 2, a **Perineal Fistula** is where the anus opening is in the wrong place and often too small to allow poop to pass easily. Dilations help enlarge the opening until surgery is done. (See Chapter 8).





During PSARP surgery for perineal fistula, your baby's muscles that control poop, or external sphincters are found using a stimulator.



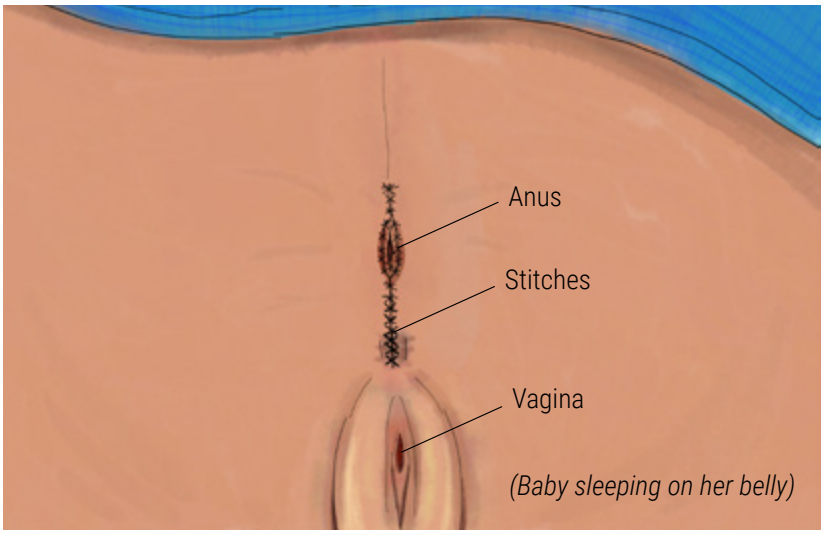
The anal opening is moved to this site to place the muscle around it.

This helps control how poop comes out. The opening is made large enough to allow poop to pass. The surgeons will make sure there is enough space between the vagina and the anus.

Sometimes, dilations are still needed to avoid scarring that can narrow the anal opening after surgery. Usually, after a few months, the dilations are done less often.

This is what the outside of the anus looks like after surgery.

The old opening or fistula is stitched closed, and the new anus is formed.

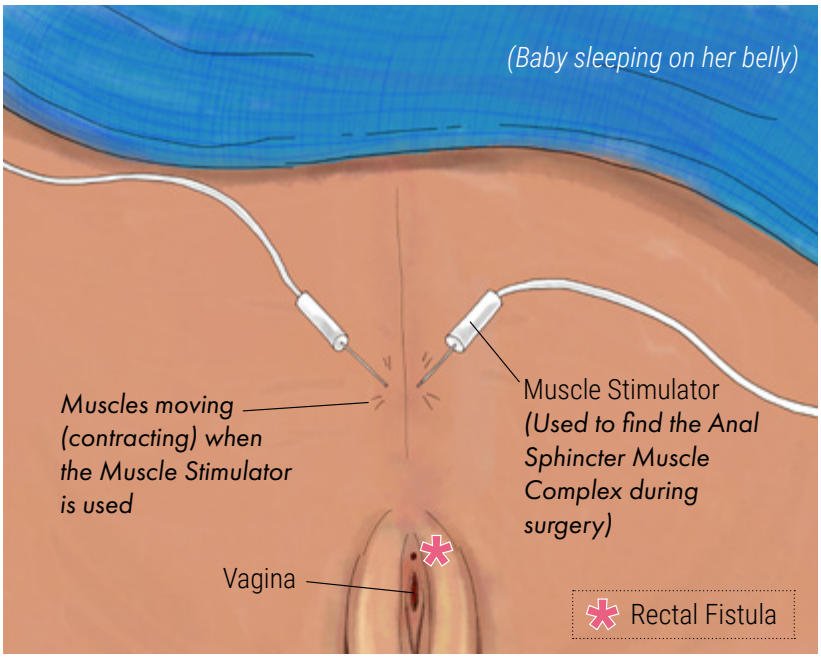


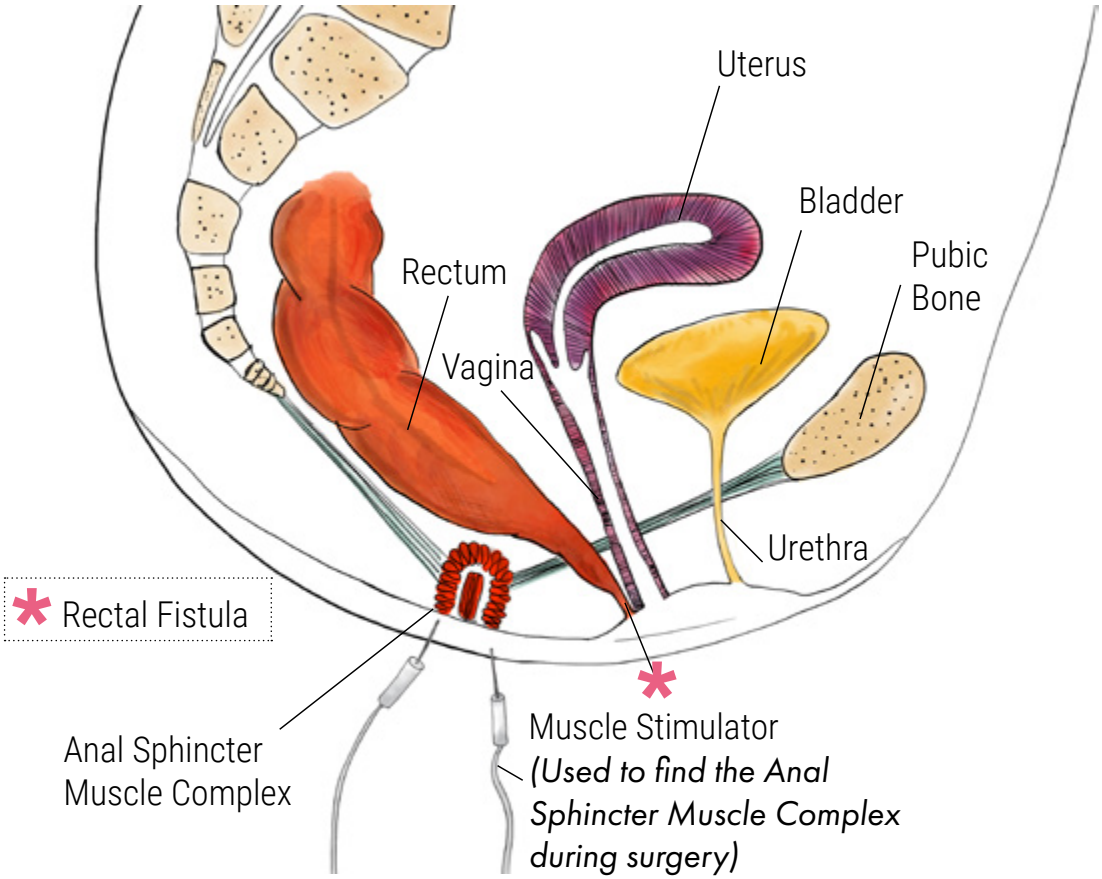
PSARP for Rectovestibular Fistula

As noted in Chapter 2, if the anal opening is close to the vaginal opening in girls, instead of in the normal place. This is termed a rectovestibular fistula. In most cases, dilations (See Chapter 8) help enlarge the opening until surgery is done.

Sometimes the surgeon might decide to do a colostomy early after birth if the dilations are hard to do. PSARP surgery to repair a rectovestibular fistula is like the surgery done for perineal fistula described above.

Your baby's muscles that control poop (external sphincters) are found using a stimulator.

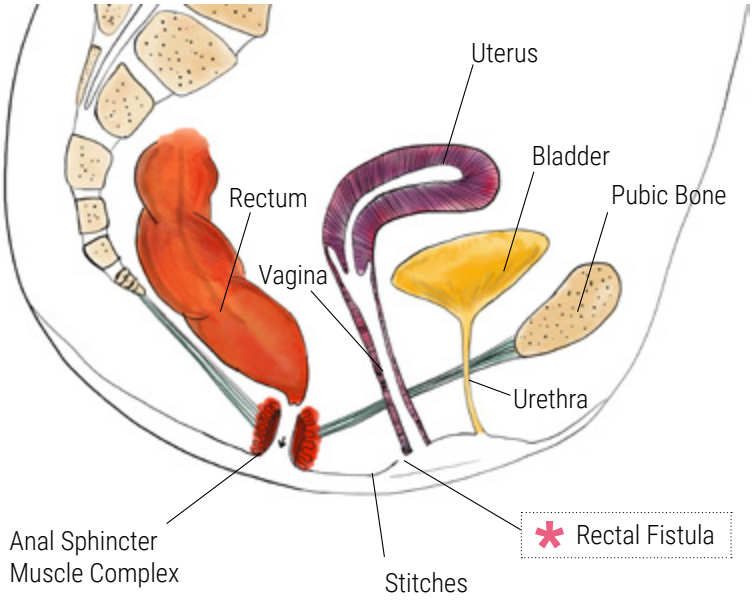




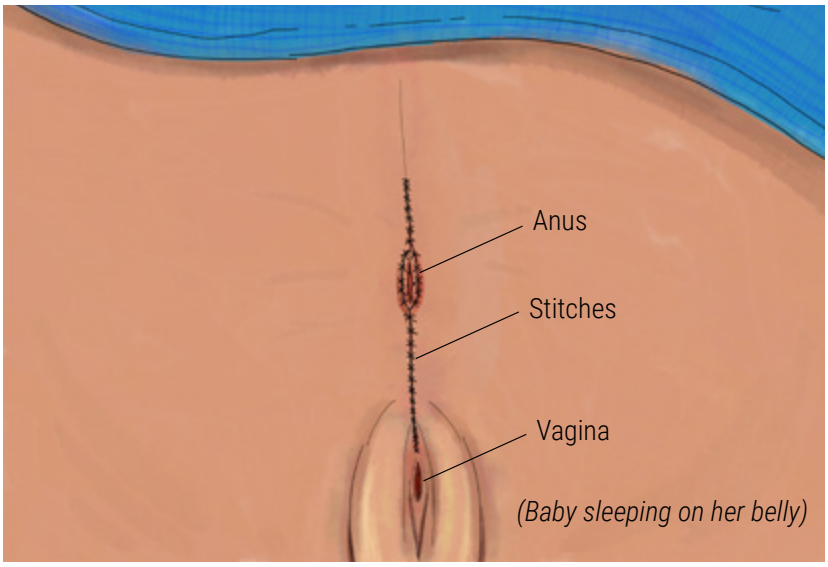
These are side internal views of the anatomy.

The anal opening is moved to the correct site where the muscles surround it. This helps control how poop comes out.

The opening is made large enough to allow poop to pass.



After surgery, the area will look similar to the illustration on the right.

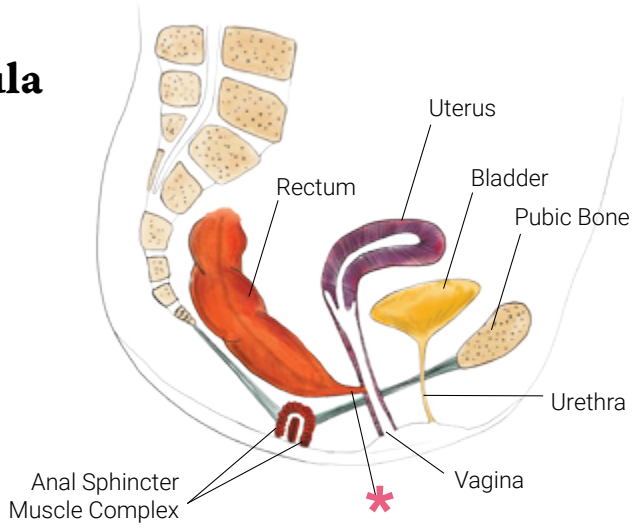


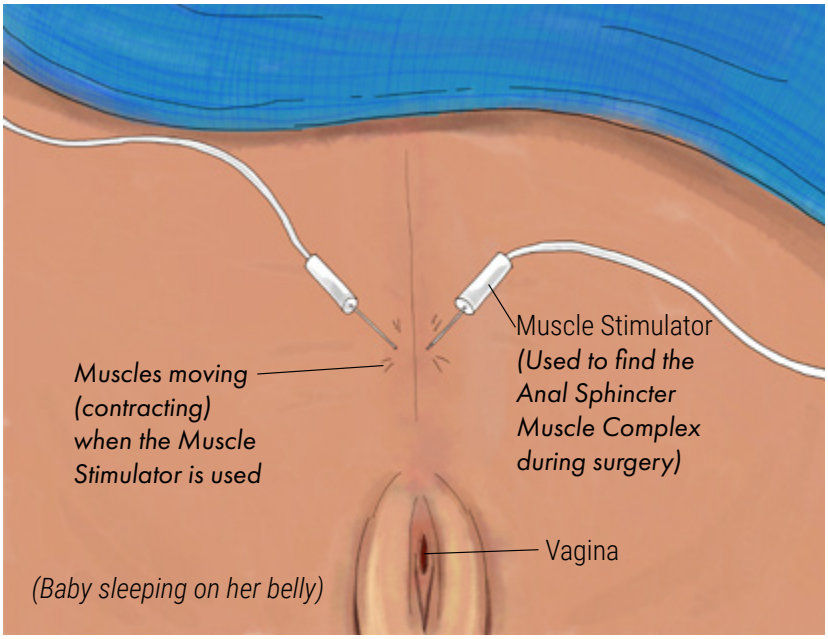
The tissue between the vagina and the anus (**perineal body**) is rebuilt. Sometimes, dilations are still needed to avoid scarring that can narrow the anal opening after surgery. Usually, after a few months, the dilations are done less often.

Creating a healthy perineal body is very important. If your daughter becomes pregnant in the future, delivering a baby through the vagina can cause damage to the anus. There needs to be enough healthy tissue between the anus and vagina to avoid damage of the anus during delivery. When the time comes in your daughter's future to deliver a baby, her obstetrician will discuss a safe way for the delivery.

PSARP for Rectovaginal Fistula

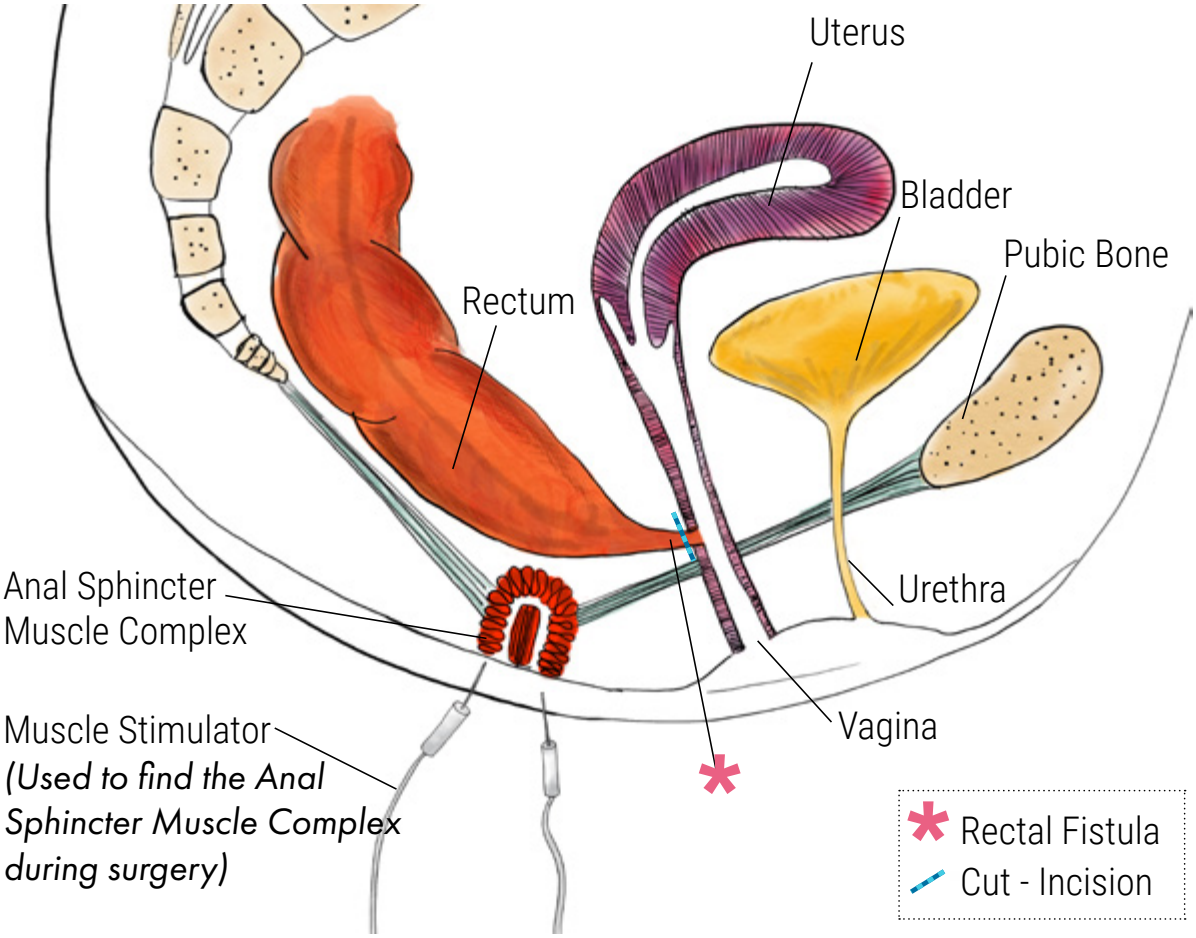
As described in Chapter 2, less than 1 percent of girls are born without an anal opening, where the rectum goes into the vagina instead of connecting to the anus. This type of ARM is called **Rectovaginal Fistula**. There is an opening for urine (**urethra**) and the **vagina**, but the anus is missing.

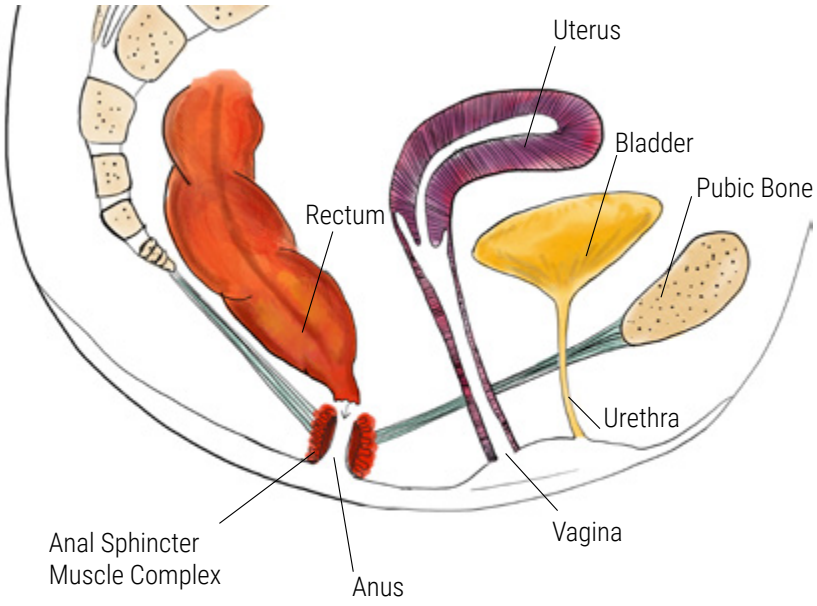




PSARP surgery to repair a rectovaginal fistula begins with finding the sphincter muscles using a stimulator.

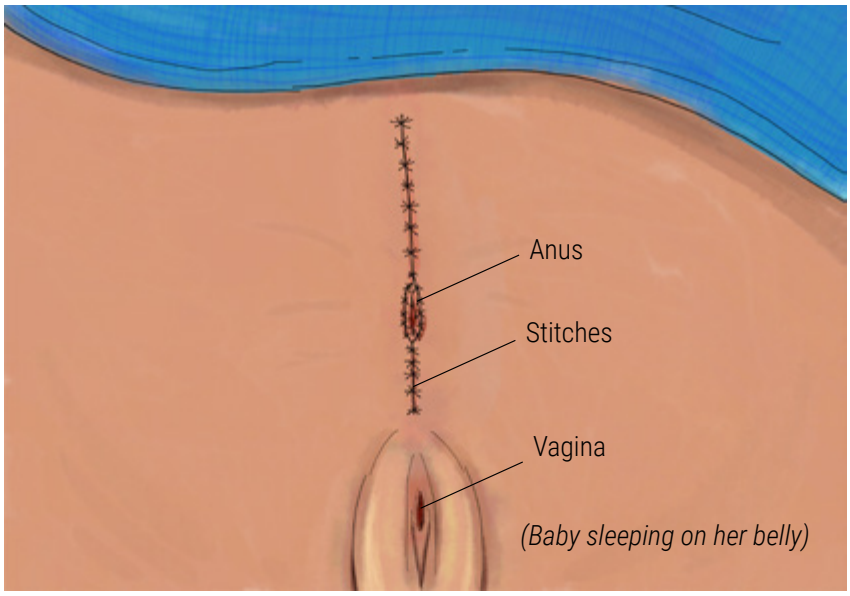
A cut is made through the muscles, and where the rectum connects to the vagina is detached.





The hole in the vagina is closed, and the rectum is then brought down to the skin level. The external sphincter muscles are then placed around the rectum in the correct place.

If the rectum's connection to the vagina is out of reach from below, a cut is made in the belly to locate it and free up the rectum. Laparoscopy can also be used for this surgery. The surgeons make small cuts in the belly and use a camera and small tools to help with the surgery.



This diagram shows how the cut may look like after surgery is complete and a new anus is made.

Problems with PSARP Surgery

Problems are not common with PSARP surgery.

After your baby recovers from the PSARP surgery, the surgeons will check that the anal opening is a right size. The anal opening may get smaller in about a quarter of patients as the body heals and creates a scar. It is easily treated with dilations that can be used less often as the anus reaches the right size (See Chapter 8 on dilations).

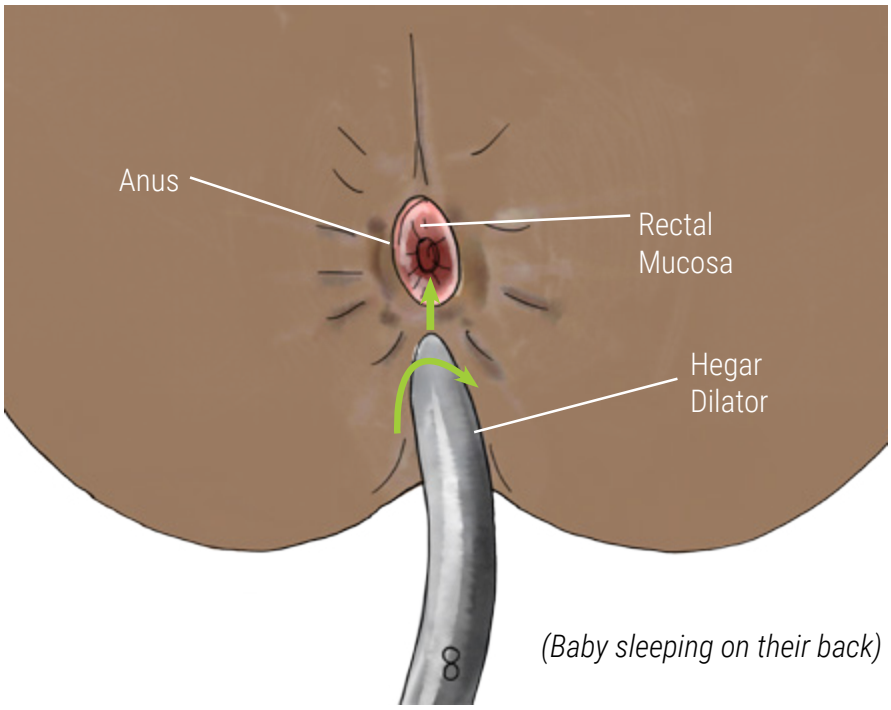
There is a small risk of infection of the repair. If this happens, sometimes **antibiotics** are all that is needed to treat the infection.

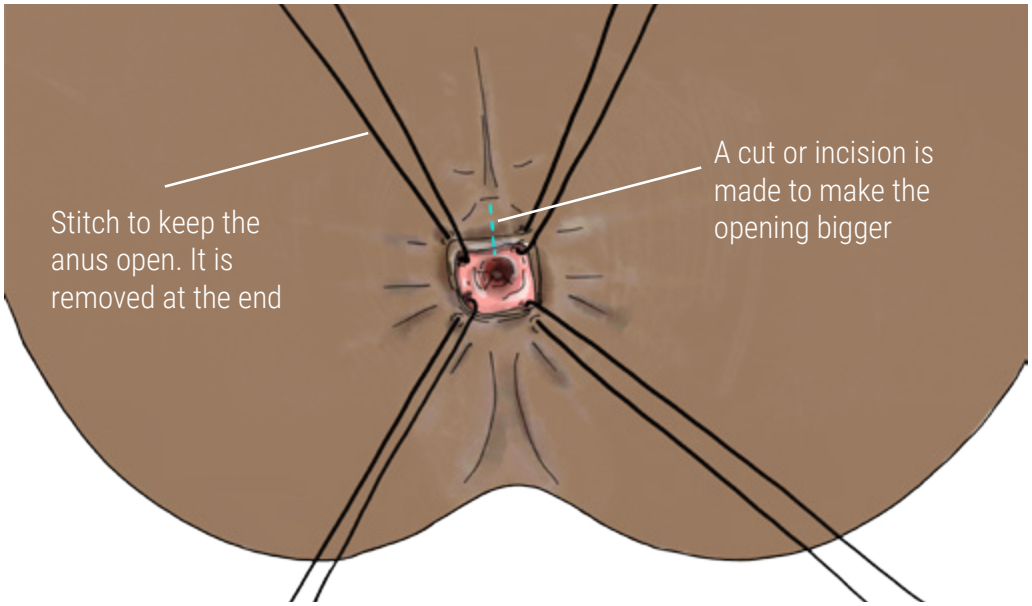
However, your baby may need another surgery if the infection leads to the skin and the repaired area separating. If your baby did not have a colostomy before the PSARP and has a lot of problems with the repair, they may need a colostomy. **A colostomy allows the repair to heal.**

Heineke Mikulicz Anoplasty

Some surgeons do not use dilations. If the anus becomes narrow, a minor outpatient surgery (Heineke Mikulicz Anoplasty) can be done to release the scar so there is a larger opening.

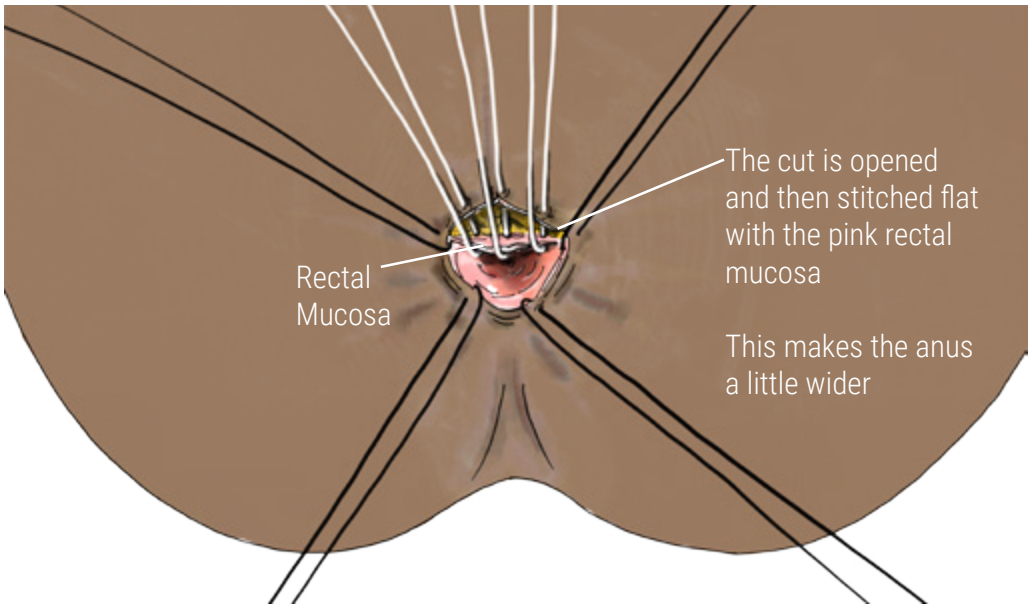
First, the surgeon checks the size of the anus using a device called a Hegar dilator.

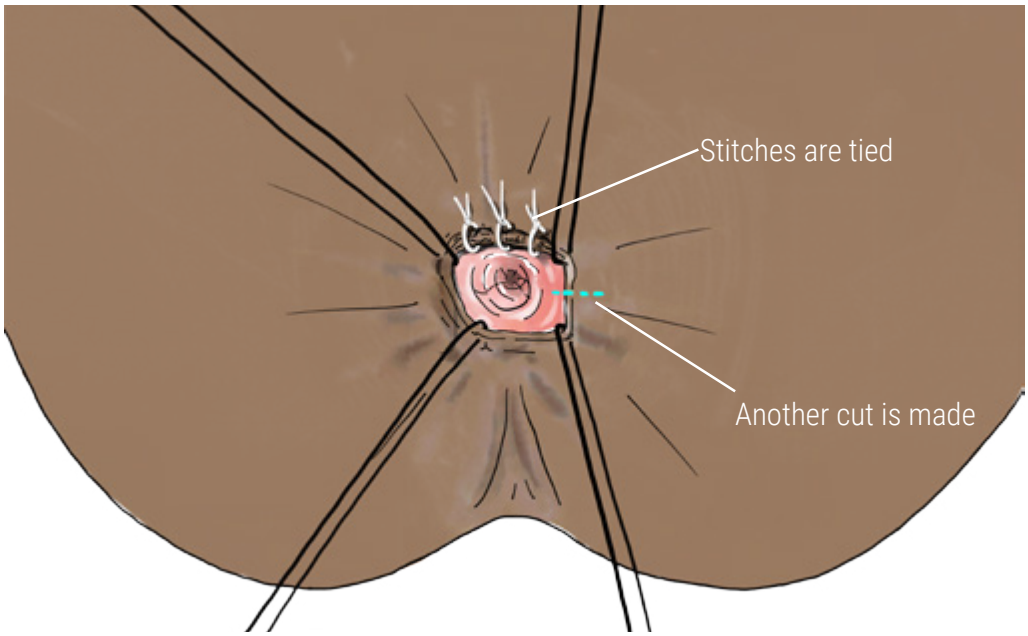




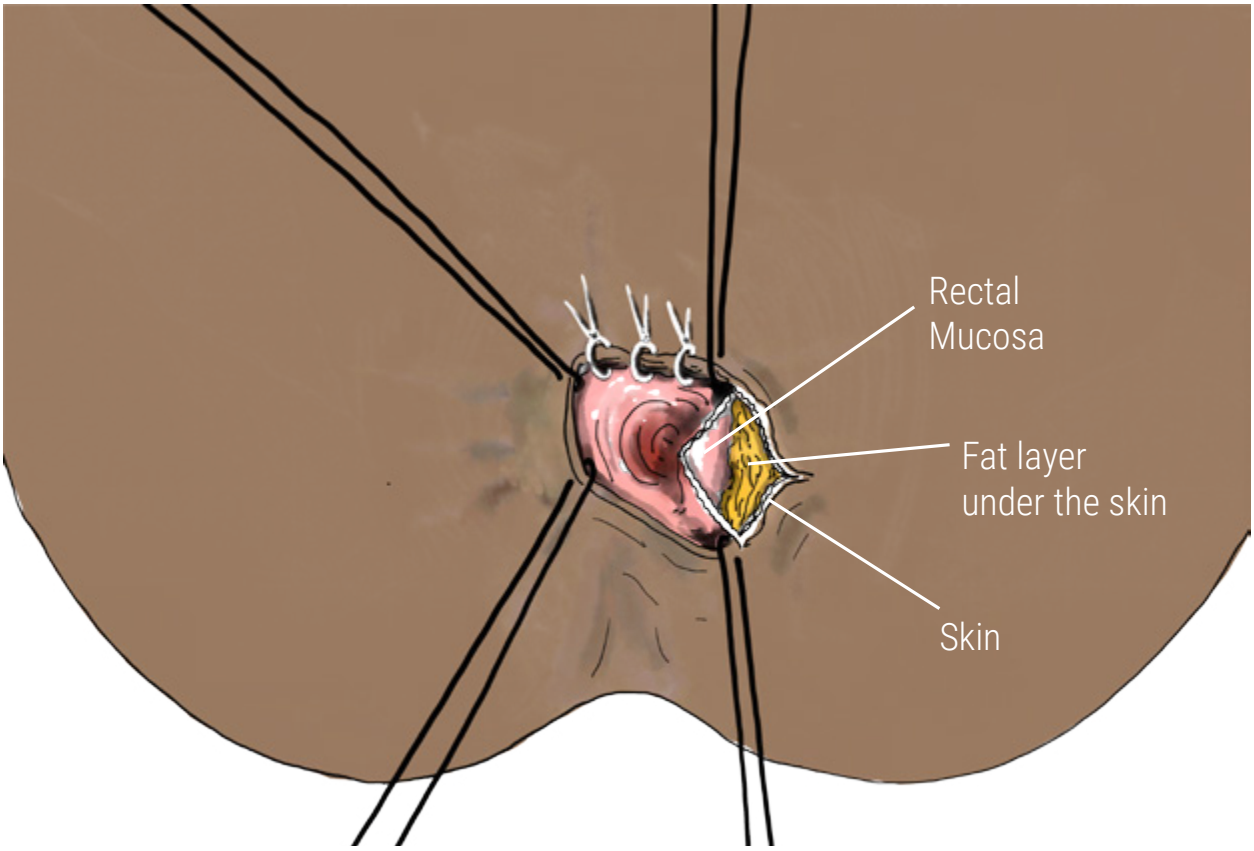
A small cut is made at the top of the anus creating an upside-down V shape. This makes the anus wider from the top.

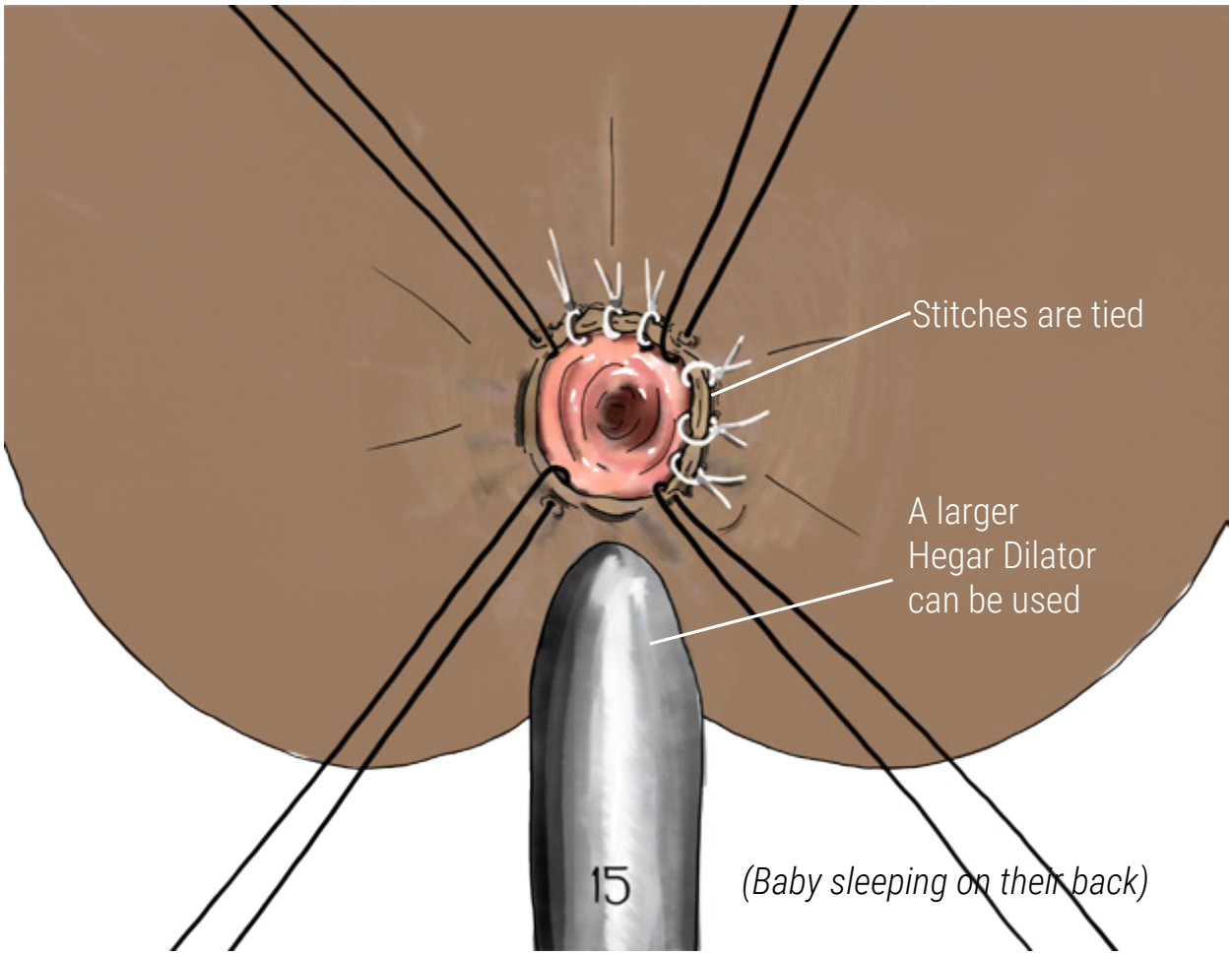
The V is stitched to make a flat line so that the skin and inside of the rectum are stitched together.





The same is done on the right side. The cut this time is horizontal and a sideways V cut is made.





The skin and tissue are then stitched together in a flat line to make the opening of the anus wider.

A larger Hager is used to know the size of the repaired opening.

If the anus is very tight due to extra scar tissue or poor blood supply to the area, your baby may need another PSARP surgery.

12. Care After PSARP Surgery

Recovery after PSARP surgery depends on how complex the ARM is and the extent of the surgery. In general, the longer and more complex the surgery is, the longer the recovery time and the amount of care needed after surgery. This chapter will describe the routine care after a PSARP surgery.

Eating and Drinking after PSARP Surgery

Children without a Colostomy

Babies with a perineal fistula or rectovestibular fistula may not have a colostomy. After the PSARP surgery with no colostomy, surgeons will usually wait for your baby to pass gas and poop before starting liquids. During this time, your baby will be given **IV Fluids of sugar and electrolytes.**

First 1-5 Days

Once poop returns, many surgeons will allow sugar-free clear liquids or breast milk for the first 5 days after surgery to give the wound time to heal.

If your baby passes poop that is hard through their anus before the wound heals, it may cause the stitches to come apart or the wound to get infected. The best way to limit the amount of poop is to control what your baby eats and drinks. Sugar-free liquids produce the least amount of poop.

Not eating or drinking much is probably going to be the hardest part of being in the hospital for you and your baby. Distraction is the best way to get through these days. Playing games, toys, painting, books, movies, and video games will help. In addition, you may bring items from home to help distract your baby during this time.

5+ Days

After 5 days of clear liquids, your baby's surgeon will check how well the wound is healing. If it is healing well, your child can start eating food.

Some surgeons don't want the baby to have anything by mouth during this time after surgery. Others may even use **IV nutrition** given through a longer tube than a regular IV.

Children with a Colostomy

If your baby has an ostomy, they will be able to start drinking clear liquids after surgery once gas and poop pass into the ostomy bag. If your baby can drink clear fluids without a stomach ache or vomiting, then they can start eating food.

Bowel Management

If your baby does not have an ostomy and is passing poop from their bottom, it is crucial to keep your baby's poop very soft for the first month after the PSARP surgery. The reason for this is to allow the wound to continue healing well. If hard poop passes through the anus or if your baby strains to poop, it may cause damage and delay the healing process.

To help keep your baby's poop soft, your doctor may want your baby to have a **stool softener (Miralax)** or **laxative (Senna)**:

- Breastfed babies usually only need Miralax, if anything.
- Formula-fed babies take Miralax and sometimes Senna.
- If your baby is old enough to eat solid food, your baby may need Miralax and Senna.
- All babies over one year of age will take both Miralax and Senna at home.

At the one-month after surgery follow-up visit, the Miralax is usually stopped, and your baby will be started on a different bowel program.

Anal Dilations

After your baby has PSARP surgery, the skin around the anus may start to close. This is a natural body's response to the healing process. **Anal dilations** can help prevent the skin around the anus from closing. You will be taught how to use dilators at your follow-up clinic visit if dilations are needed. **Dilations are usually done two times a day**. When the size of your baby's anus is what the doctor wants, the number of times a day you dilate will be slowly reduced.

Chapter 8 looks at anal dilations in more detail.

Heineke Mikulicz Anoplasty

Instead of dilations, sometimes the wound is allowed to heal on its own. In about a quarter of babies, scar tissue may narrow the opening after healing. If this occurs, a procedure to enlarge the size of the anus is done after PSARP. The procedure is called a **Heineke Mikulicz Anoplasty** and is usually done three months after your child's PSARP surgery.

In this procedure, the surgeon will make small cuts in the anus and rearrange the tissue to enlarge the anus. It is done as an outpatient, and the advantage is to avoid dilations, especially in older children who may not tolerate them. (see the end of chapters 10 and 11 for more information and illustrations).

Skin Care

Your baby will have softer and more poop during the first month after surgery, increasing the risk of diaper rash. Prevent diaper rash by cleaning your baby's bottom and applying a **barrier cream** with each diaper change. In addition, you should change the diaper as soon as possible anytime you notice poop in the diaper.

Essential rules to follow when cleaning your baby's bottom after a PSARP surgery:

• Only clean the skin with **water** using a squirt bottle or a **No-Rinse Foam Cleanser**.

• Pat dry with **dry wipes only**. The hospital should provide you with these wipes.

• Do NOT wipe where the stitches are because this may cause disruption.

• Avoid baby wipes bought from a store. **Use dry wipes only.**

• After your baby's bottom is clean and dry, apply a **barrier cream** to help prevent diaper rash or for the rash from becoming worse. Your nurses will teach you how to use all these barrier creams. You will get a bag of supplies before your baby goes home.

• Apply the barrier creams to all red areas, except the area close to the stitches.

Several practical steps to follow when using barrier creams:

• Apply **Cavilon No-Sting Wipe** that leave a clear liquid that coats the skin protecting it from poop.

• A barrier cream such as **Critic-Aid Skin Paste** acts as a barrier to keep poop from coming in contact with the skin. This paste provides relief from discomfort caused by a diaper rash.

• Avoid putting barrier creams on the stitches because it may cause them to dissolve or break down.

• Other products can be used if your baby's bottom has open or bleeding areas. These include **Stomahesive Powder, Ilex Skin Protectant Paste with Vasoline, and Marathon.**

• To further protect your baby's skin, the nurses may apply **Marathon** or **Cavilon**. These are liquids that form a strong layer to protect the skin. They will usually last for 3 days and should not be wiped off.

Pain Control after PSARP Surgery

Pain relief and comfort are essential for your baby's care after surgery. At first, **Tylenol** and **Ibuprofen** are given through an **IV** for pain relief, as your baby may not be allowed to take anything by mouth. **The IV name for Tylenol is Omefivir and the IV name for Ibuprofen is Toradol.**

If your baby has kidney disease or is at risk for having kidney disease, Toradol or Ibuprofen will not be used because it can affect the kidneys.

Your baby may also be given an **IV narcotic, such as Morphine or Dilaudid.** The narcotic is given by a nurse, or you will be taught how to give it by pushing a button on the IV. Once your baby is allowed to eat and if pain control is still needed, the IV narcotic may be switched to **Oxycodone,** which is given by mouth.

PSARP Urinary Catheter Care

If your baby has a recto-urethral fistula, a urinary tube or **catheter** is put in during surgery. This tube is put into the urethra during surgery to drain the urine from the bladder. There is a balloon at the end of the tube that inflates once inside the bladder. The balloon prevents it from coming out. To allow for complete healing, your baby may need to go home with this tube. It will stay in place until the follow-up clinic visit, from one and two weeks after surgery. The tube will be removed at that time.

Flushing the tube once a day, and as needed, prevents clogging. While your baby is in the hospital, your nurse will show you how to flush the tube once a day with 10 to 20 ml of sterile water.

When your baby has a urinary tube in place, they may not soak underwater. This includes taking a bath and swimming. Showers are fine for bathing.

13. Surgery for Cloacal Malformations

Newborn Period

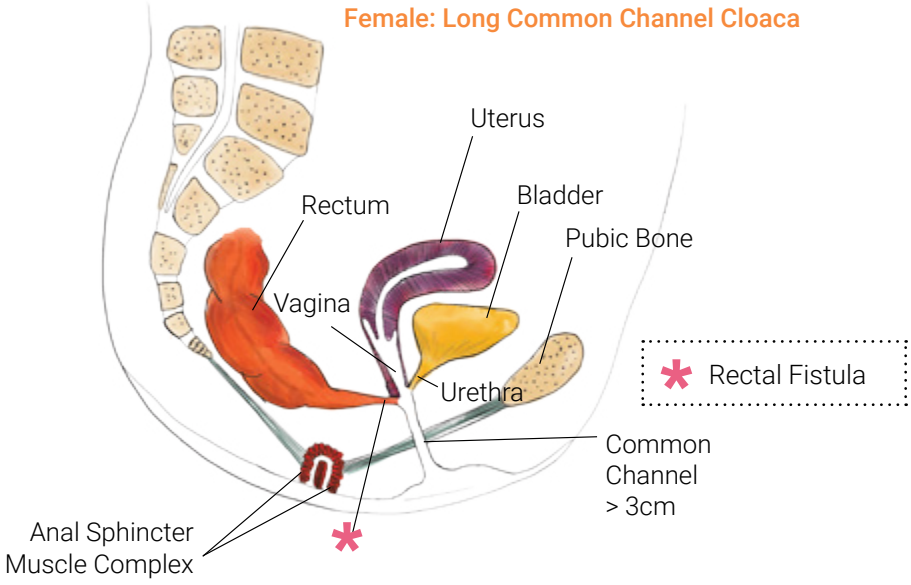
Cloacal malformations are complicated - some more complicated than others. A cloacal malformation happens before birth (congenital) and is a defect of the urinary, gynecological, and colorectal systems. It is not an abnormality of gender development. All children with cloacal malformations are little girls. These little girls need complex medical and surgical care to help control urine and poop, sexual function, and fertility as they grow up.

The focus for care is to prevent infection and kidney damage. In the first few days after birth, babies with cloacal malformations need to be at a medical center with surgeons who have skill in caring for these babies.

Once your baby is diagnosed, we will need to check if she has other issues like heart problems or a swollen vagina or vaginas (**hydrocolpos**), which may need treatment. By 48 hours after birth, a colostomy will be done where the colon is brought out to the skin to allow the poop to drain out (See chapter 7).

A **colostomy** prevents poop from mixing in the vagina and bladder, thus preventing severe infections called sepsis.

If hydrocolpos is present, it may need draining; otherwise, the kidneys may become blocked and damaged. In the past, while doing the colostomy, surgeons put a tube in the vagina from the belly (called a **vaginostomy** - See Chapter 7). More recently, surgeons treat most babies without a vaginostomy. They have been able to drain the vagina or vaginas by passing a small tube into a single hole on the outside. The vagina can drain in this way, and you can learn to do this at home. Your baby will have a few ultrasounds to see how the drainage flows and make sure the kidneys are healthy and draining well.



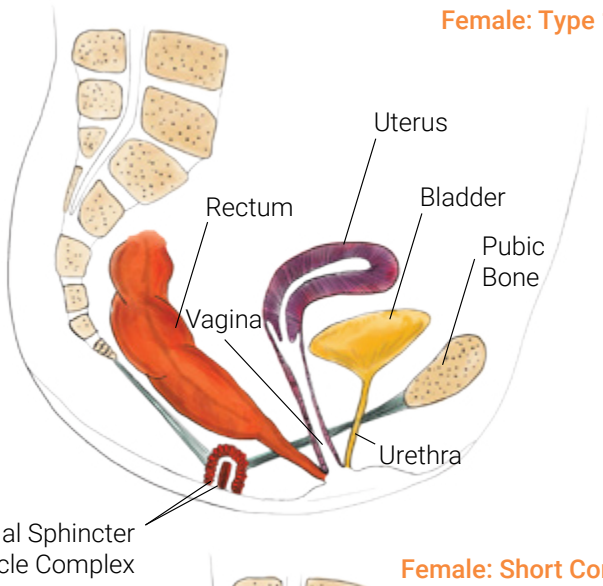
Planning for Surgery

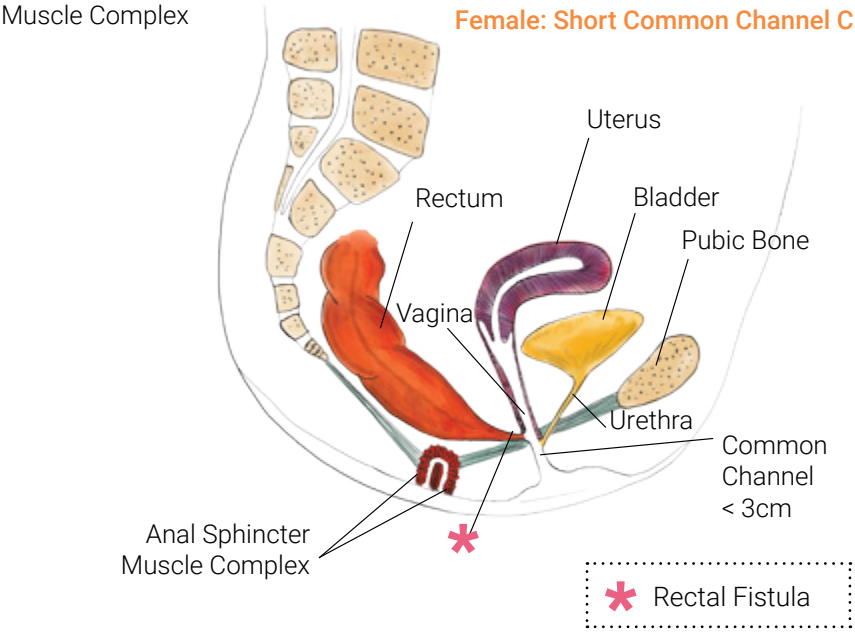
Once your doctors are certain urine is draining well, and you learn how to care for the colostomy, you and your baby can go home, to grow and bond with the rest of the family. Your baby will have follow-up visits and further testing for the next 6 months.

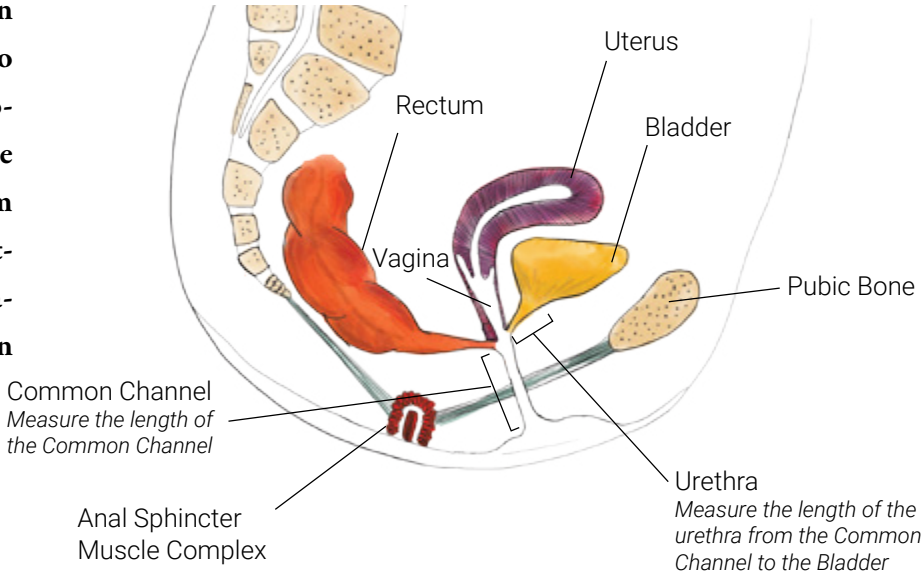
At that time, the surgeons caring for your daughter will do some testing to look at the anatomy of her cloacal malformation and plan surgery when she is ready.

Tests may involve passing small tubes with cameras into the belly in the operating room (OR), x-rays, ultrasound, MRI or a combination of these tests.

The tests help show the details of your baby's anatomy. They help your doctors predict how complicated the repair will be. **Your doctors will look at the length of the common channel, before it splits into the urethra, vagina(s), and rectum. The length of the urethra from the common channel to the bladder is also measured; and the anatomy of her vagina(s) and the position of her rectum to her pelvic floor is noted. With this information, your doctors can plan her surgery.**







Cloacal Malformation Repair (PSARVUP)

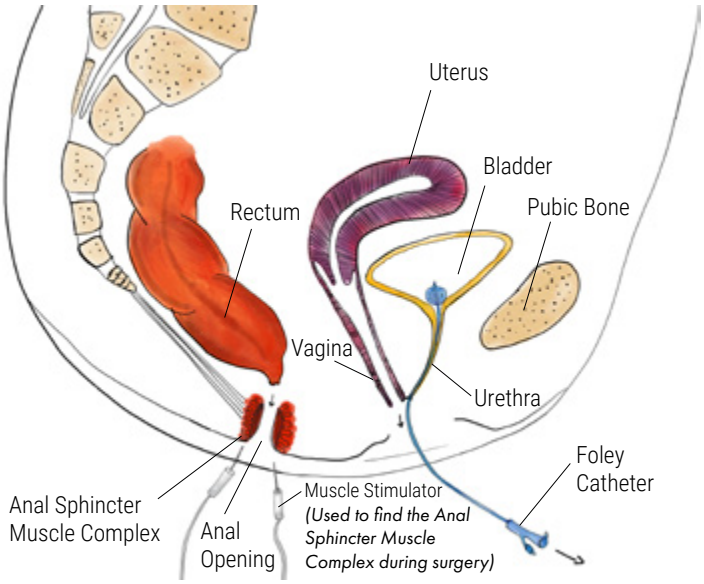
The surgery for cloacal malformation repair is generally termed: **Posterior Sagittal Anorectovaginourethroplasty (PSARVUP)**.

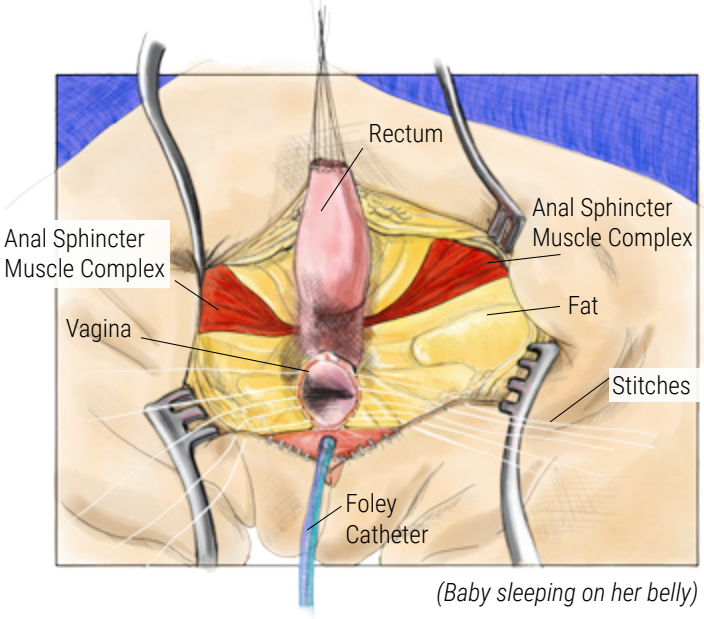
To repair your daughter's cloacal malformation, your doctors need to separate her rectum from the other parts and put it in the correct place for her **anal sphincter** (squeeze muscles). When it comes to repairing the urethra and vagina, there are two main ways to do this:

- **Total Urogenital Mobilization or TUM.**
- **Urogenital Separation**

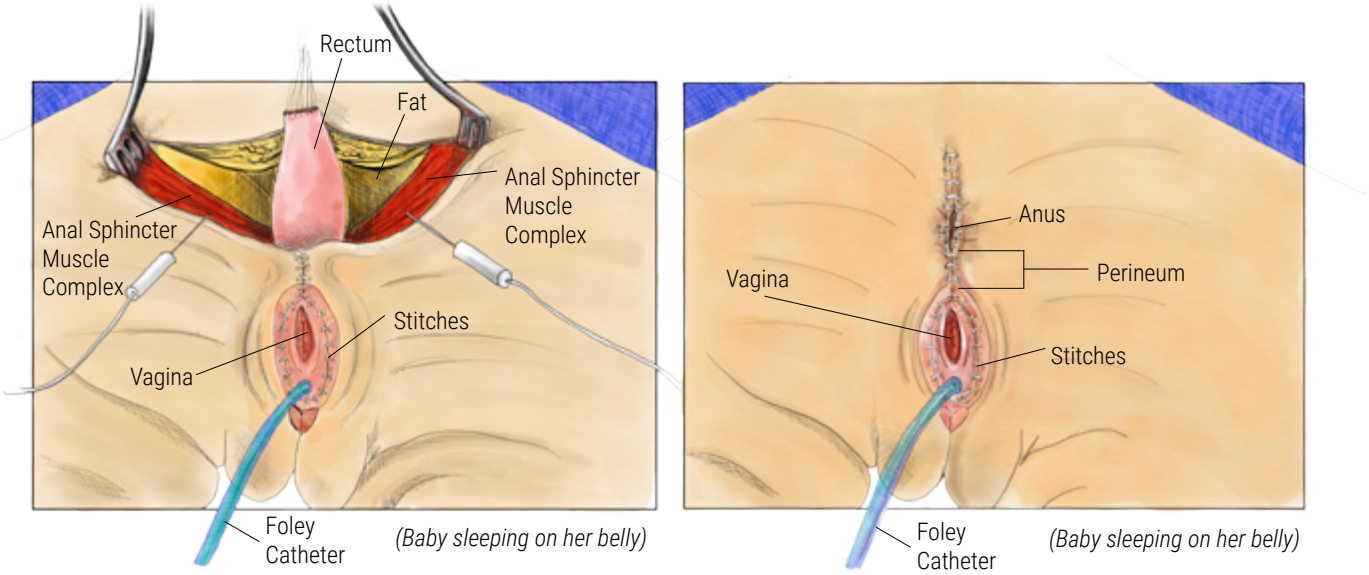
Total Urogenital Mobilization or TUM.

If the common channel is short (less than 3 cm), and the urethra is long (more than 1.5 cm), the urethra and vagina(s) can be freed up from the tissue around it. It is then brought down to the outside and stitched in place. This is called **Total Urogenital Mobilization or TUM**.



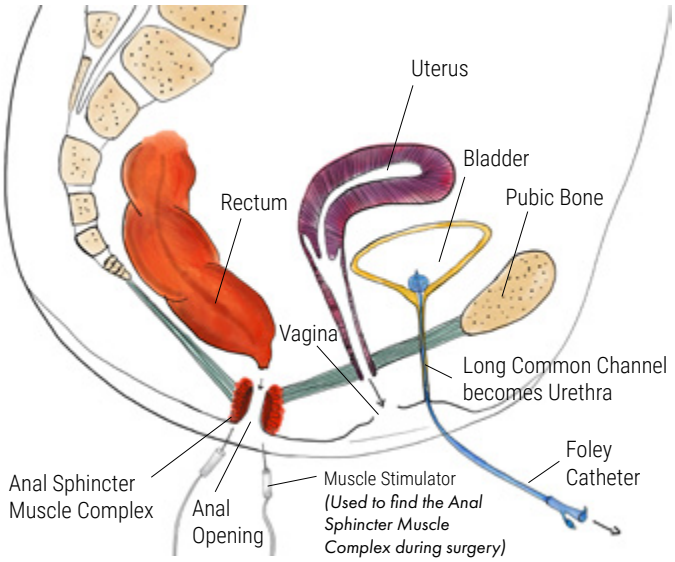


(Baby sleeping on her belly)



Urogenital Separation

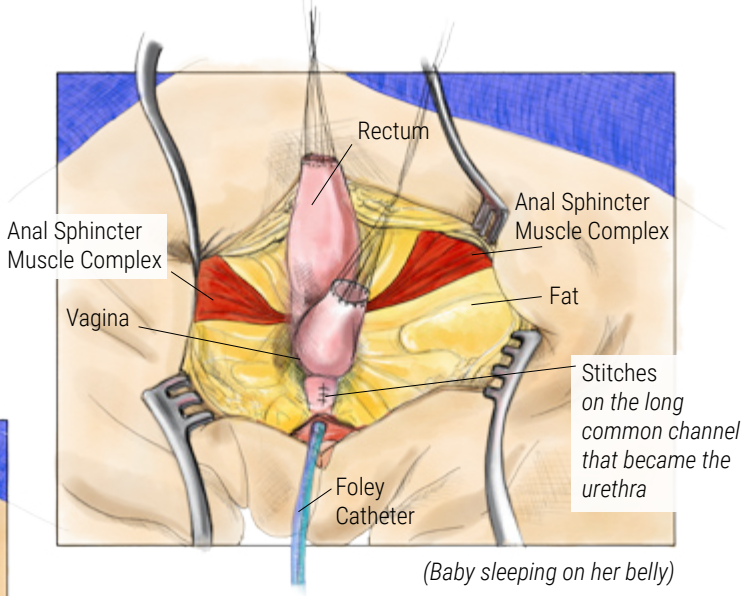
If the common channel is long (more than 3 cm), and the urethra is short (less than 1.5 cm), your doctors will separate her vagina(s) from her urethra and bladder and repair the connection. This procedure is called **Urogenital Separation**.



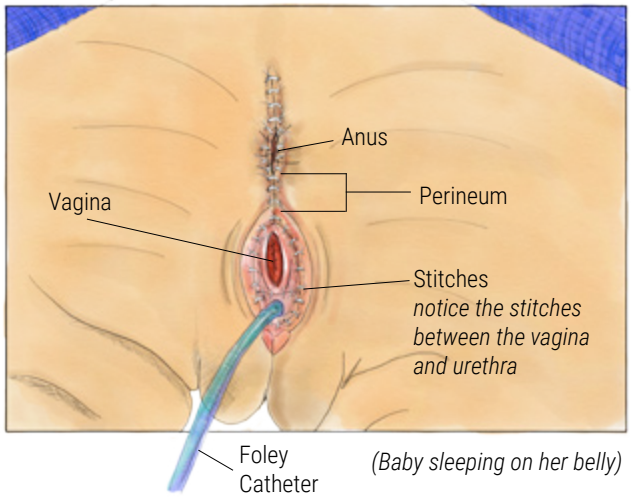
This surgery is much more complicated to do. Sometimes, even after the separation, her vagina may not reach all the way to the outside because she was born with it high and short.

Therefore, some babies need a tissue bridge made from a piece of bowel that goes from their vagina(s) to the skin on the outside. This is called a **Vaginal Replacement.**

A Urogenital Separation with Vaginal Replacement is a much more complicated surgery and needs to be done by surgeons that regularly care for babies with complex cloacal malformations.



(Baby sleeping on her belly)



(Baby sleeping on her belly)

After Surgery

After surgery, your daughter will have some tubes in place to drain the urine. These will stay in place for 2 to 6 weeks, depending on the type of surgery. Sometimes surgeons remove these tubes in the operating room so that a thin tube with a camera or scope is passed to check for healing.

After the repair has healed well, you may need to do anal dilations to keep the size of the anus where it should be. (See Chapter 8). **However, no vaginal dilations will be done.**

Once all has healed well, her colostomy can be closed. This will allow the poop to pass through her repaired anus.

Long-term Care

All girls with cloacal malformations need ongoing care to check their kidneys. They will also need follow-up for help with control of urine and poop.

When they are near puberty, follow-up is needed to check that menstrual blood can easily pass. Then, as your daughter becomes an adult, she will need to follow-up with urology, gynecology, and colorectal surgery to ensure everything is working correctly.

14. Care After Cloacal Malformation Surgery

As with any major surgery, the care after surgery for your baby with a cloacal malformation repair **Posterior Sagittal Anorectovaginourethroplasty (PSARVUP)**, can feel overwhelming. Given how complex the surgery is, recovery can feel slow at times.

Expected Length of Stay

The usual hospital stay after cloacal malformation surgery may vary but expect at least 5 days after surgery. Recovery can take longer, depending on how complex the surgery was.

PSARVUP Care

Eating and Drinking after a PSARVUP

The more work done on your baby's bowels, the longer it will take for the bowels to wake up. When your baby's bowels are still asleep from the surgery, it is called an **ileus.**

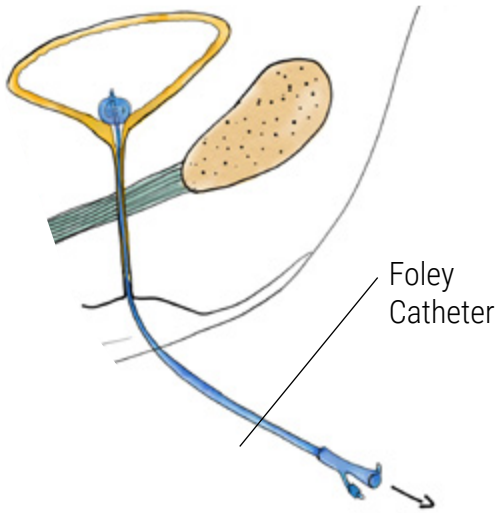
An **ileus** can last several days. If your baby can walk, walking will help the ileus resolve. Once your baby's ileus resolves, you will start to see gas and poop in her stoma bag. When this happens, she can begin drinking clear liquids. If your baby does well drinking, clear liquids without a stomach ache or vomiting, she can start eating food. Your baby needs to be eating and drinking enough so she has the right amount of fluid in her body to go home from the hospital. (See the section on **Complications After Surgery** in this chapter)

PSARVUP Urinary Catheter Care

Your baby may need to go home with a urinary catheter or tube to drain the bladder of urine and to help heal the urethra and tissue between the anus and vagina (perineum). Different types of tubes are placed during surgery. The types of tubes include:

- **Urethral Catheter**

A urethral catheter is put into the urethra. This tube has a balloon at the end that inflates once inside the bladder. The balloon keeps the tube from coming out.



Managing any Pain from the Tube

When a urinary tube is in place, it may cause **bladder spasms**. It may be difficult for your baby to tell you that they are having bladder spasms. If your baby is irritable or complaining of pain, your baby may be having bladder spasms. Bladder spasms can be very uncomfortable. While your baby is in the hospital, **Valium** may be given to help the bladder spasms.

Other drugs that can help with bladder spasms include **Oxybutynin** or **Ditropan.**

Water and Showers

When your baby has a urinary tube in place, they may not soak underwater. This includes taking a bath and swimming. Showers are fine however.

Premarin Cream

Premarin Cream is an **estrogen cream** used to increase blood flow to the genital area. The increase in blood flow helps prepare the area for surgery and helps with healing after surgery.

Premarin cream is only used for babies with cloacal malformation who have a normal vagina. If your surgeons decide to use this cream, you need to apply it 4 weeks before your baby's surgery.

Starting on day two after surgery, you will begin to use the Premarin cream and continue to do so for two more weeks. You will learn how to apply the Premarin cream before and after surgery.

Pain Control after PSARVUP Surgery

Pain relief and comfort are important for the care of your baby care after surgery.

Your child may be given **Tylenol** through an **IV** because they are not allowed to eat after surgery. The IV name for Tylenol is **Omefivir**.

Usually, babies with cloacal malformation are not allowed **Ibuprofen** or **Toradol**. This is because there is a higher risk for kidney damage since these drugs go through the kidneys.

Your baby may also be given an **IV narcotic**, such as **Morphine** or **Dilaudid**. This narcotic is given by a nurse, or you will be taught how to give it by pushing a button on the IV.

When your baby is allowed to eat, the IV narcotic will be switched to **Oxycodone**, which is given by mouth.

Your baby may also have an **epidura**l or a tube that provides local pain relief. The pain team manages these tubes and giving the pain medicine.

15. Colostomy Closure

A **colostomy** is done before the repair of some ARMs, such as rectourethral fistula in boys and cloacal malformation in girls.

Sometimes, it is done for a rectovestibular fistula and rarely for perineal fistula.

After PSARP surgery, the surgeons will wait at least 6 to 8 weeks to let the repair heal before doing surgery to close the colostomy. It is crucial to make sure the new anus has healed, and the opening is the right size before closing the colostomy.

(See Chapter 8 on anal dilations).

Before closing the colostomy, your baby's bowel might need to be prepared or cleaned out. This is usually done by having your baby drink a **bowel prep fluid** (**Go-LYTELYTM** or **MiraLAXTM**).

This can sometimes be done at home. After the poop from the colostomy becomes clear and free of solid poop, the bowel cleansing is stopped.

Surgery for closing the colostomy is done under general anesthesia. The cut to enter the belly may vary, but usually the cut used to make the colostomy is re-opened. The surgeon will free up the colostomy and the mucus fistula from the skin and muscles and then connect them to the colon. The connection is made with stitches or staples.

Sometimes a small plastic tube or drain will be left under the skin for a few days to allow for fluid to leave the body and reduce the risk of infection.

After Colostomy Closure

After the surgery, it may take a few days for your baby to poop from their anus, since the bowel slows down from the surgery and anesthesia. Sometimes surgeons will allow a small amount of clear liquids for comfort while waiting for the bowel to begin working again. You will know when it wakes up because your baby will start to pass gas and poop.

Once your baby begins to poop, their diet will become more normal. It is important that your baby doesn't develop a diaper rash since their skin has never had poop on it and it can easily become irritated. You can prevent skin problems by making sure you do good skin care and use creams that protect the skin (**barrier creams**).

Problems with Colostomy Closure

Colostomy closure is usually a straightforward and safe surgery. Of course, no surgery is without risk.

- A possible risk for this surgery is leaking from the colon's connection. Although rare, it can be a problem that requires another surgery and sometimes making an ostomy again.

- Another risk is a skin infection since this area has poop come in contact with it. Infections are treated with **antibiotics**. Sometimes, the skin may have to be opened to drain the infection.

- An infrequent problem is the narrowing of the connection in the colon. This is called an **anastomotic stricture** and may need a specific form of dilation using a colonoscopy and possibly surgery to fix it.

16. Urological Surgeries with ARM

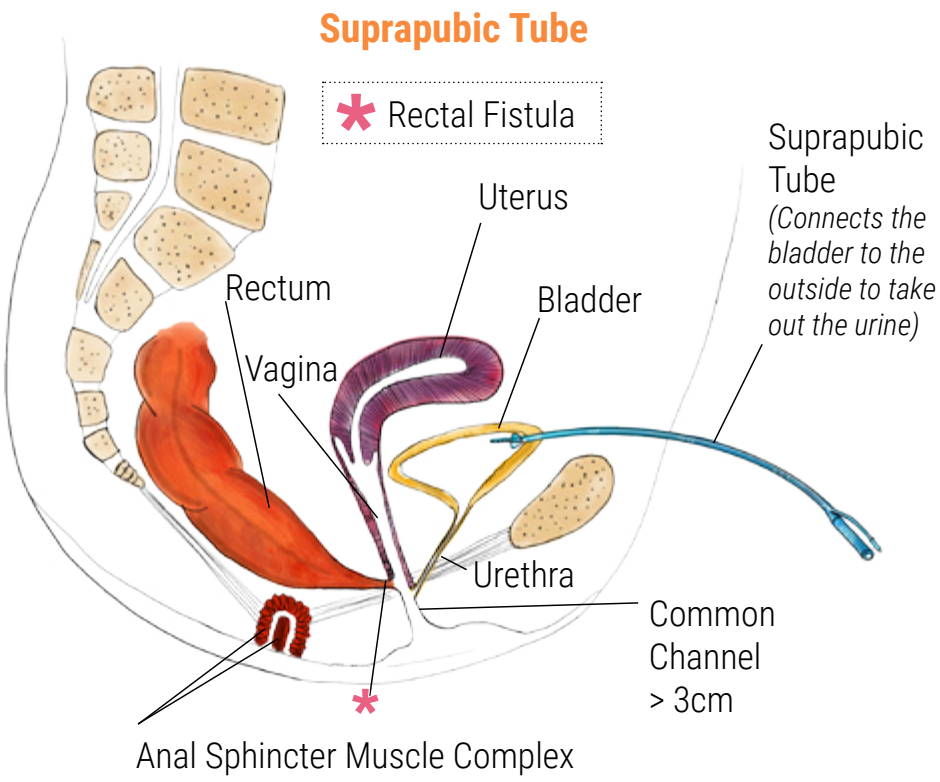
In the first few years of life in babies with ARM and cloacal malformations, the first concern is to prevent urinary tract infections and keep the kidneys healthy down the line as your child grows older. A team of doctors including pediatric surgeons, urologists, as well as kidney doctors (nephrologists) should be involved in the care of your child if they have any kidney problems. Sometimes, using a tube to drain urine from the bladder several times a day can stop infection and keep the kidneys healthy. However, sometimes surgery may be needed later as your child grows.

Different Ways to Drain Urine

Suprapubic Catheter

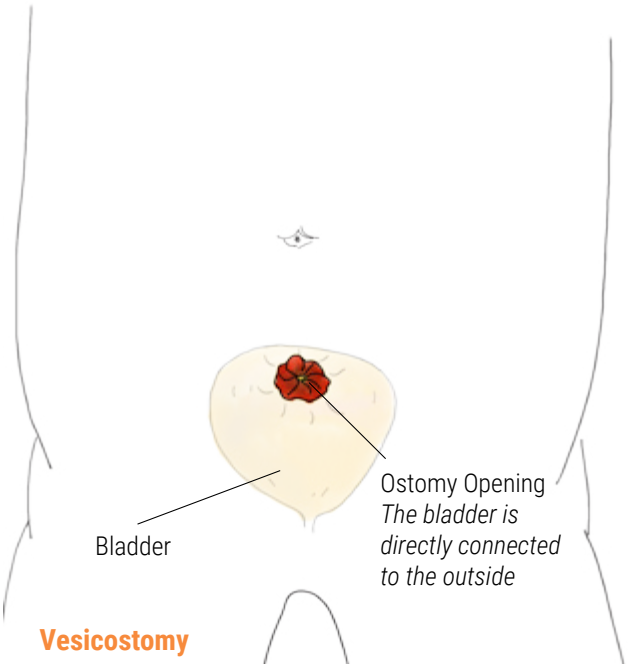
A suprapubic tube is a drain placed into the bladder during surgery that comes out of the skin under the belly button. This tube is used as a back-up in case the urethral or Mitrofanoff tubes get clogged. It is usually removed about a week after the Mitrofanoff tube is removed.

All these tubes drain urine all of the time, and your baby may have one or two of these tubes at one time.



Vesicostomy

A surgery called a vesicostomy may be needed if your baby has problems passing urine the normal way through the urethra. It also may be done if using the tube several times a day to drain urine does not work. A vesicostomy involves bringing the bladder up to drain through a hole in the belly. It allows the bladder and kidneys to drain freely and the pressure in the urinary tract to be kept as low as possible. This helps protect the kidneys and reduces the risk of infections. The vesicostomy is put low on the belly and drains into a diaper. Your baby just needs regular diaper changes. This surgery is used only for a short time for passing urine from the body. As your baby reaches the time for potty training, the urologist will talk with you about closing the vesicostomy and how to further take care of your baby's urine.



Cutaneous Ureterostomy

With ARM, the ureters that carry urine from the kidney to the bladder may connect to the wrong place in the bladder or urethra, or they can connect to the vagina, or other structures. This is called an ectopic ureter. These ureters are often blocked, which results in abnormal drainage and damage to the kidney. Surgery is needed to correct this problem. Sometimes, the baby is too young for this type of surgery that puts the ureter into the bladder to relieve the blockage. Therefore, it is necessary to bring the ureter up to the skin to drain the urine into a diaper. This surgery is like an **ileostomy** or **colostomy**. A **ureterostomy** is a short-term solution for draining the kidney. The ureter is usually put into the bladder when the baby is around 1 to 2 years of age, as described in the **urethral reimplantation** section below.

Ureteral Reimplantation

Urine traveling in the wrong direction from the bladder back up to the kidney (**vesicoureteral reflux**), is common in children with ARM and cloacal malformation. A small daily dose of antibiotics is given to keep the urine sterile and prevent infection. However, if infections occur while on antibiotics, surgery may be needed. This surgery tunnels the ureters across the bladder. In this way, the backflow of urine is prevented when the baby passes urine. This surgery is usually done around 1 to 2 years of age and helps prevent further damage to the kidneys from infection.

Nephrectomy

In rare cases, your baby may be born with the kidney not working well or a kidney blockage develops due to a dilated colon or hydrocolpos. If your baby's kidney is not working very well or not working at all, the doctor may talk to you about removing the kidney. This is called a **nephrectomy**. A kidney that is working poorly or not working at all risks infection and can lead to high blood pressure later in life. Kidney removal usually takes place when other surgeries, such as a colostomy closure, are done.

Babies with ARM and cloacal malformations may have a kidney that doesn't work, called a multicystic **dysplastic kidney (MCDK)**. This kidney never fully developed before birth. It often slowly decreases in size and with time disappears. Sometimes, a MCDK does not fully disappear and needs to be removed with surgery.

How to Prevent the Tube from Clogging

It is crucial to flush these tubes 1 to 2 times a day to prevent clogging.

If your baby did NOT have a **bladder augmentation** (see Chapter 22), you need to flush the tube with 10 to 20 ml sterile water one time every day.

If your baby had a **bladder augmentation**, you need to flush these tubes with at least 30 ml sterile water two times a day, and as needed. Since a bladder augmentation involves making the bladder bigger using some of the bowel's tissue, it is important to flush the tubes as directed. The bowel tissue connected to the bladder makes mucous all the time.

This mucous can easily clog the tubes and prevent them from draining. **If the tubes do not drain, urine cannot leave the bladder, which is an emergency.**

You will be taught how to flush the tube while your baby is in the hospital. If you have any worries, feel free to call your medical team.

17. Understanding Insurance & the Health Care System in the USA

Health insurance is very much a part of the total health care your baby receives. Healthcare today, especially complex surgery, is costly.

Often, families spend a lot of time searching to find the right hospital and health care team to trust with their baby's care. Sometimes, they are denied care due to insurance. **This chapter will help you learn how to meet the challenges facing you when seeking the best care for your baby. We will begin by reviewing the different types of insurances.**

Medicaid

If you want to travel to a hospital outside your state and you are insured by your state's Medicaid Program, you will need approval. This is because State Medicaid Agencies require you to look at all the options for care within the state before you seek care in another state. See the **Out of Network Approvals section** of this chapter for more.

State Medicaid Agencies often have many Medicaid plans from which families can choose. The plans, called **Medicaid Health Maintenance Organizations (HMOs)**, might be managed by large insurance companies like **Humana, Aetna, Molina**, etc. When choosing your Medicaid insurance plan, pay close attention to the plan's **Out-of-Network (OON) options.**

Some **Medicaid HMO** plans have no **OON option**. In those cases, your baby cannot seek care from an out-of-network hospital or surgical team. **If your baby has a complex anorectal, urologic, or gynecologic malformation, select a plan that has an OON option.**

If you're stuck in an existing plan, consider changing plans as soon as you can, or during the open enrollment period. Even if the new plan costs a little more, it will allow you to see the specialists your baby needs for the best possible care. It can also save you money in the long run, reducing the number hospital stays and surgeries your baby may need.

If your state Medicaid plan or Medicaid HMO has an OON option, you're in luck! You may be able to get approval for care from an out-of-state hospital. You'll need to understand your insurance plan's rules and ways to get approval. **How to get approval is described in the Out-of-Network Approvals section, which is used by most States. Before you begin, it's a good idea to call your insurer to make sure you understand this process. Then, reach out to the Hospital where you want to go. They will have staff members there ready to help you.**

Commercial (Employer-Provided) Insurance

Commercial insurance plans also want you to get approval before you get specialty care outside their network of physicians, surgeons, and hospitals. Therefore, you need to understand your insurer's network and out of network options. If you want to go to specialty care centers, you must have an insurance plan with an **OON option**.

Pay close attention to your insurance options during open season (November - December each year) when employers allow you to switch plans.

Even without an **OON option**, an insurer "may" approve you to seek care from an OON specialty service provider.

However, getting the insurance approval is often complex and takes a lot of time and you can't be sure you'll get approval. **An OON option gives you choices.**

Traveling for Medical Care

There are four steps to prepare you to visit an out-of-state hospital. Tips on how to get approval for out-of-network insurance will follow. Then we'll talk about health care provider relationships, insurance approvals, travel, and how to make your return home easier.

Step 1: Keep the Relationships with Your Local Doctors and Surgeons

Partnering with your doctors and surgeons is always the best way when seeking specialty care elsewhere. Your doctor wants the best results from the treatment given.

Support from your local, in-network doctors is often needed to get approval to have care from hospitals outside those your insurance covers. Also, your baby will need the ongoing care of your local doctors in the future.

Five best ways to partner with your local doctors to get support for your visit:

1. Make an appointment and talk with your local doctors.

2. Bring information to the appointment about the hospital you want to visit.

3. Share with your doctors why you want to use this particular hospital.

4. Share how this hospital could help and support you.

5. Ask your doctors what they think about the hospital and will it meet the needs for your baby's care.

Sometimes, talking to you doctors may be difficult, but it's important to be respectful and honest with them. This approach will strengthen your relationships with your local doctors, who have your best interests in mind.

Step 2: Check Your Insurance Plan's Network Coverage

Understanding your insurance coverage is essential when thinking about going to a distant hospital. Most commercial, employer-based plans allow you to go anywhere in the country for care. However, some of these plans are called **"small network plans."** These types of plans only allow you to get care from certain providers, only within your state or a small portion of your state.

The best way to check your insurance plan network coverage is to call your insurance carrier's member services phone number posted on the back of your card. You can also go to the insurance website noted on the back of your card. Ask if your chosen hospital and surgeons are in their network.

Now that you have this information, what do you do with it?

If you find that the doctors, surgeons, and hospital that you want to visit are in-network, most of your work is done. The doctor's office and the hospital where you want to go will do the rest. If however, your insurance plan is a small network plan, and the doctors and hospital you want to visit are out-of-network, don't get discouraged. A few more steps will be needed to get insurance approval from your small network insurance plan. We will discuss ways to get out-of-network insurance more in-depth later.

Knowing about your healthcare insurance network coverage can reduce your stress. It will give you a clear understanding of what you need to do to visit a specialty hospital. Being an active partner, will reduce any problems that often complicate the approval process.

Step 3: Never Travel Without Insurances Approval in Place

Make sure you have your insurance approvals completed before you travel for care. If you do not have these approvals in place, you run the risk of getting costly insurance bills that are difficult to get paid by your insurance. If the specialty hospital gives you appointments before talking about insurance, possible insurance problems may happen. **Always ask questions and partner with your new hospital to get the approvals before your visit.**

Step 4: Preparing for Your Visit – Medicines & Homecare

Now that you have your insurance approvals you are ready to begin planning your trip to the new hospital. Below are some things you need to get ready before your visit that will make your return home easier.

1. Make sure you bring your ID and insurance card, even though they should already be in your medical chart.

2. Have the name, address, and phone # of your local pharmacy. This information will make it easy for the staff to call your prescriptions in and for you to pick them up when you arrive home. Your local pharmacy will work on insurance approval for your drugs.

3. Have the name and phone # for the homecare supply service you want to use. This will help the nurses at the specialty hospital get your supplies and work with your insurance for approval.

If your insurance company denies paying for either your baby's drugs or homecare supplies, the team at your new hospital can help you.

Your local pharmacy or homecare supply company may not be able to get the information to work with insurance companies to get coverage for these items. Having this information will help your hospital care team support you.

Out-of-network Approvals

Getting out-of-network insurance approvals is often not easy. However, you can avoid costly charges or extra fees by taking a few simple steps. Insurance providers will direct you to in-network providers, which reduces their cost of sending you to an out-of-network provider. The in-state providers are usually more familiar and trustworthy to them. Approval for care from an out-of-network hospital depends on how well you, your insurance agent, and your doctors plan together

The following steps will help you get the out-of-network insurance approvals you need:

Step 1.
Get a complete medical plan for your baby's care from the specialty hospital you want to go to. This will give you an idea of what you will be asking insurance to approve.

Step 2.
Talk to your local doctor's office about partnering with you to get out-of-network insurance approval.

This step is essential because they can help provide materials on the medical care needed. Depending on what your insurance plan wants, additional materials of support, or referral to the insurance carrier may be required.

Step 3.
Collect letters from all your local doctors. These include information on the necessity of medical care, a brief medical history, and treatments, along with the need for you to visit the specialty hospital. An example of a letter is on the next page.

Why are letters of medical necessity important?

Remember, the insurance carrier's role is to find you a team of doctors in the network who can meet your baby's medical needs. These letters should show that you have contacted all in-network resources, and your doctors support your need to go out-of-network for care. Here are some examples of doctors from whom you need letters of medical necessity:

- Your local surgeon if you are going to the new hospital for surgery
- Other medical specialists involved in the care of your child
- Your child's primary care doctor.

If you do not have local doctors supporting your visit to the specialty hospital, you will have to find them, or your insurance will find them for you.

Ask the new hospital if they know pediatric colorectal surgeons in your state who can provide an evaluation of your child and make a recommendation to your insurance provider.

Step 4.
Get a letter of medical necessity from the hospital you plan to visit, along with their medical plan, and the reason why they are a good fit for the treatment your baby's needs.

Step 5.
You are ready to submit the forms to your insurance carrier. Depending on your plan, your local primary doctor's office might submit them for approval. Often, the hospital you are planning to go to will submit it on your behalf. Always talk with the health care team to ensure this step is completed.

Incomplete forms will be denied and getting approval on appeal is more difficult and less likely to succeed.

Slow, deliberate, complete work on the insurance materials is the best way forward. The insurance specialist at the new hospital you want to go to is the best resource to know when approval forms are ready to go forward.

Your insurance approval should include:

1. Letters of medical necessity from your local doctors.

2. A letter of medical necessity from the surgeon at the new specialty hospital.

3. A medical care plan from the doctors at the new hospital.

4. The most recent clinical information (imaging studies, lab results) with a short health history of your baby's care to date.

Following these steps can reduce the length of time it takes to get a response from your insurance carrier. These steps also increase your chances of getting good results. With the help of your local doctors and those at the specialty hospital, you need to show your insurance carrier the life-changing benefits your baby will get from being treated at the specialty hospital.

Sample Letter

Date: 4-12-2021
Re: Patient Name
DOB: 01/01/1111

To Insurance Carrier,
This patient is currently under my care and has been since birth. This pediatric patient has a complex past medical history, including cloacal malformation, patent foramen ovale, and abnormal spinal cord. In addition, past surgical history includes colostomy and mucous fistula creation, exploratory laparotomy, reopening of recent laparotomy, jejunal stricturoplasty, revision of colostomy and mucous fistula, cystoscopy, and vaginoscopy. Due to the complexity of this diagnosis and history, she requires a multidisciplinary team of pediatric surgeons who are experts in managing such children with rare conditions like this patient.

This multidisciplinary type of care will significantly improve this child's quality of life, which requires a care team devoted to the careful study of this special patient population and its long-term changes and health patterns. Therefore, I recommend her for a complete anatomical evaluation, future treatment, and continuation of care.

Please feel free to contact our office if you have any questions or concerns.
Thanks,
Local Provider
Address
Phone _____ Fax _____

Understanding Insurance Denials or Appeals

Insurance denials can be discouraging, but they don't have to be.

There are a few reasons why insurance requests get denied, but the main reason is the lack of "clearly explained" clinical information from the hospital where you want to go. This means the doctors there did not show how your baby's clinical need for care and their treatments will improve their quality-of-life. Insurance nurse reviewers and medical directors review hundreds of requests for approval. It is impossible for them to know every diagnosis and every type of treatment needed.

Level 1 Denial:
An insurance nurse that reviews the case doesn't approve the request. The request goes to the medical director, and a medical director denies it.

Level 1 Appeal "Peer-to-Peer":
Your health care team at the hospital you want to go to requests a conversation, called a "Peer-to-Peer," with the insurance medical director.

A Peer-to-Peer is an excellent time for your health care team to give a lot more detailed information to the insurance medical director. Most denials are overturned at this level of appeal.

Level 2 Denial:
The request was discussed during a "Peer-to-Peer," and the medical director still does not approve the request.

Level 2 Appeal or "Member Grievance"
is a more formal written request. This includes a member statement on a particular form explaining their appeal needs. There is also a written request from the health care team at the specialty hospital with new clinical information. It is usually a long process averaging 30 days.

Level 3 Denial:
The request was reviewed, and the additional clinical information was not enough for the medical director to approve the request.

Level 3 Appeal or "Third Party Appeal"
is the last and final type of appeal insurance providers offer. The request for approval is sent to an outside third party that reviews your requests and the insurance denials. Unfortunately, this also has a lengthy review process and can take up to 60 days, and sometimes longer.

This process can be stressful. Keeping in touch with your health care team at the new hospital should help keep everything on track and lead to a good result. Remember, this is not a sprint but a marathon.

Making Travel Plans and How to Reduce Out of Pocket Costs

Now that your insurance approvals are in place, it's time to start planning your travel to the specialty hospital. Here are a couple of cost-saving resources that may help to reduce your out-of-pocket cost when traveling.

- Many commercial insurance and state-funded medical plans have travel resources for members approved for care at a distant hospital. Call the member services number on the back of your insurance card or your state's Medicaid hotline on their website. Ask if there is a travel department or a team that helps with travel planning and payment. Both commercial and state-funded plans have **case managers**. These case managers do a great job connecting you with cost-saving travel options within your plan's benefits. If you don't have a case manager assigned to you, ask them to assign one to you, especially if you plan on traveling often for care.

- Ask the distant hospital if they have a **Social Work Service** within the hospital that can help with travel planning. A few non-profit organizations help with airfare. Many hospitals have discounts at local hotels or even free overnight places for travelers.

1. When traveling for medical care, the more connections you can make, the easier it will be to move things along faster and reduce the stress.

2. Keeping in contact with your doctors at the new hospital and your local health care team will help get your baby the long-term care they need.

3. Taking time to educate yourself on your insurance plans and benefit options will help you feel informed, confident, more relaxed, and in better control of getting care for your baby.

18. Adopting a Child with ARM

Caring for children with anorectal malformations (ARMs) is very challenging for any family. Managing the health care needs and balancing those carefully within your family, to allow all family members to thrive, is a very difficult task. A very special group of people choose to adopt children with ARMs, a noble undertaking.

Your new child, adopted from abroad with ARMs, may have some health care problems that need evaluation and care. However, most evaluations can be delayed for 3 to 6 months to allow your new baby or child to become an important part of your family. Sometimes treatment is needed right after adoption. For example, children with heart failure who have problems with exercise should seek help right away. Other conditions which may need to be checked right away include a baby or child who has a lot of bladder infections and cannot control urine, poop, or both. This condition leads to skin rashes and toileting problems.

Ideally, your child should be evaluated by an adoption clinic with experience caring for children with ARMs. Adoption clinics will do a full medical work-up, review vaccination records, and give you advice about your child's care. Outside of these issues, you can focus on bonding with your child and wait for a full workup and treatment.

Most parents who have adopted children from abroad know that the medical information can sometimes be incomplete. Usually, there are records, but these may not tell the complete story. In addition, babies or children with ARMs may have other problems such as VACTERL. (See Chapter 5). These problems should be evaluated, and in many cases, the results are either incomplete or lack the written information. So, we advise that a full evaluation on adopted babies or children should be done unless clear records exist. Over 60% of babies or children with ARMs seen after adoption from overseas may end up needing surgery. Therefore, we suggest an exam be done under anesthesia, including anal exam, cystoscopy of the bladder and vaginoscopy of the vagina. This exam allows your doctors to see if any surgery is needed. If you have a daughter, it will also help your doctors to see if there are concerns for future issues that may occur around the time of puberty.

The anatomy of your daughter may still need a further exam around puberty to make sure that menstrual blood can flow freely. Later, issues around sex and having children may need to be addressed. Therefore, a pelvic ultrasound should be done at the start of puberty. Also, if your daughter has other problems found during the screening or if she develops painful menstruation, more tests will be done.

Treatments that were done before will impact on which treatments should be done now. There is evidence that the use of anal dilations may affect children as they get older. Many adopted children will often have anal dilations done as part of their care in orphanages. Treatments done through the anus, such as enemas, may result in stress in these children and should be avoided if possible. Adoptive parents should be given ways for which anal procedures should be done on their baby or child with ARMs.

Having a specialist read your child's chart before adoption can give you a realistic understanding of the long-term care needed. In addition, this will also allow you to try to get more medical records, which will help with long-term care.

Adopting a child with complex medical needs, such as a history of ARMs, is not easy. However, hospitals who have a specialty in caring for these children have resources that can help support you and your new family member on this beautiful journey.

19. Toilet Training

Although toilet training may be years away from when you found out that your baby had an ARM, it is a common topic of discussion. You may be concerned about your child's ability to be independent as an adult and if they will be able to control their urine and bowels (**incontinence**).

Every child is unique, and the medical team may not be able to predict in infancy how their urine and bowel control will be in the future. However, here are helpful tips and information to help guide you through the toilet training years and beyond.

Being Ready for Toilet Training

Nerves that are more developed:

If your child has good feeling that they need to poop, they may show signs that they are ready to toilet train. In general, signs for being ready to toilet train include:

- Following instructions.
- Letting you know that they need to use the toilet.
- Showing an interest in toileting.
- Walking themselves to go to the toilet.
- Managing their clothes by themselves.
- Staying dry for several hours during the day.
- Waking up dry from naps or overnight.

Nerves that are less developed:

If your child has little or no feeling that they need to use the toilet, they will have more problems with toilet training. This does not mean your child will have to stay in diapers forever.

However, they may need more help with controlling urine and poop to avoid accidents in social settings. Many children who have little or no feeling for passing urine and poop can stay socially clean with help from their parents. Paying attention to your child's cues and seeking help from your health care team, will help you know when your child is ready to try toilet training.

When to start toilet training will vary and depend on your child's specific anatomy, where they are in development, and other medical conditions. Important information to consider is how well your child can feel when they need to pass urine and poop.

Nearly all children with ARMs have some loss of feeling in their anus. The nerves for feeling the need to pass gas and poop are often not developed enough or missing.

Predicting the Success of Toilet Training

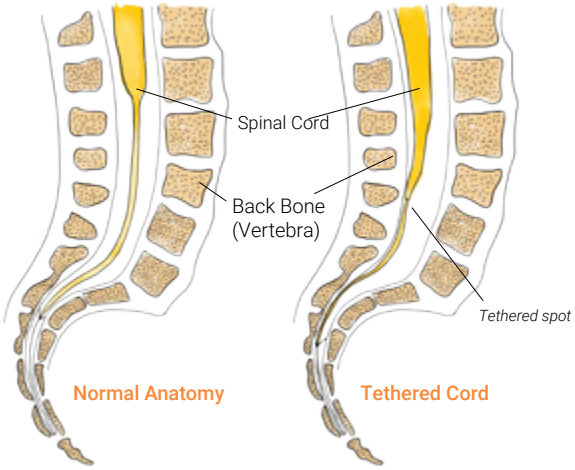
While it is impossible to predict how well a child can feel and control passing urine and poop when they are a baby, we can use information to help us know what to expect.

1. We look at the type and severity of the ARM.

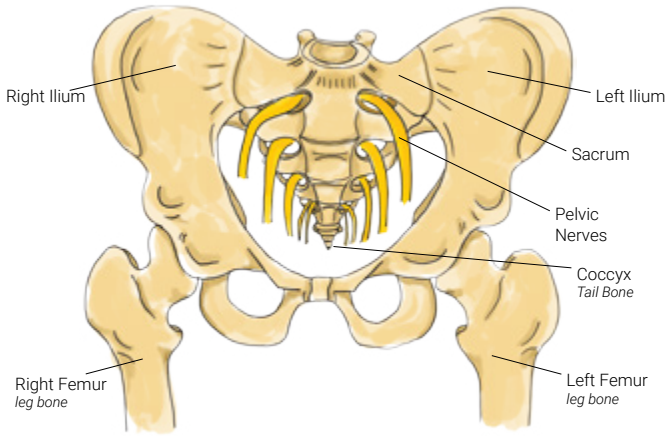
- Children with the "least complex" types of ARMs will usually do better with toilet training. This includes perineal fistulas, rectovestibular fistulas, and cases of anorectal malformation in boys where the rectal fistula is "low" (rectobulbar fistula).
- Children with more complex ARMs, such as "high" ARM with a high rectal fistula or some complex cloacal malformation cases, have more problems with lack of feeling and control of urine and poop.

2. We also consider whether a child has any type of spinal cord defect with their ARM. For example, conditions like **tethered cord** and **myelomeningocele** can impact a child's future ability to control urine and poop. (see below).

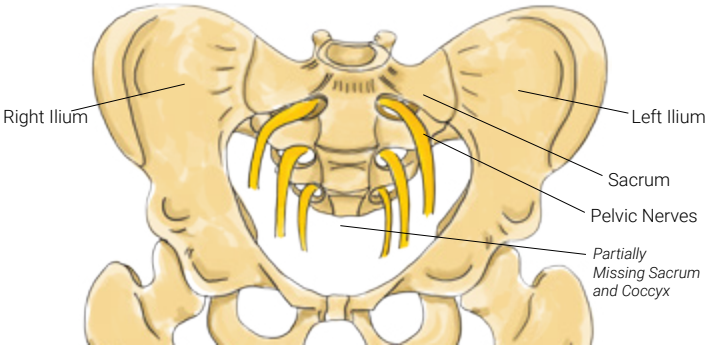
3. Finally, we look at how well your child's sacrum is formed. The sacrum is a triangular-shaped bone just above the tailbone. This bone may not be developed enough in children with ARMs. The sacrum has nerves passing through it. These nerves provide for the feeling and control of certain body parts in the pelvic area. If a sacrum is fully developed, we assume the nerves and muscles in that area are also developed. If a sacrum is only somewhat formed or absent, the child may have more problems with regular toilet training. (see illustrations below).



Normal Anatomy Of The Pelvis (Hip)



Example Of Sacral Agenesis



Nothing is ever for sure in predicting a child's ability to control urine and poop. However, it is helpful for the health care team and you to have an idea of what your child may need to stay clean and dry.

Setting the Right Expectations

As stated above, your expectations are a big part of toilet training. Knowing your child's anatomy and having an idea of how well they can feel and control their urine and poop, will make toilet training much less stressful.

Setting expectations that your child cannot meet will lead to a lot of stress for the entire family. It can also lead to your child having a sense of failure. Having the right expectations from the start can also help create a toilet training plan that is realistic.

If you are aware that your child is likely to struggle with lack of feeling and control of poop, you can work with your health care team to create a program for increasing control. Rather than using a usual toilet training plan, your health care team will develop a special training plan for your child.

This can lead to greater success with toileting and help your child stay socially clean.

Success results in more self-esteem and pride for your child.

Steps in Toilet Training

When first starting toilet training, the steps you will use for your child with an ARM are the same as for children without an ARM. **Your knowledge and readiness to begin toilet training is the most important first step.** You and others caring for your child should be prepared to spend time for toilet training daily for several months.

Beginning steps include:

- Decide what the family will call urine and poop and use those terms every time.
- Buy a toilet seat and make it special for your child.
- Encourage your child to sit on the toilet seat fully clothed to get used to it.
- Once your child is comfortable with the toilet, they can start sitting on it unclothed. Have your child do this at regular times to try to catch them passing urine or poop.
- Give your child praise when they try to pass urine or poop.
- Do not punish your child for having accidents. It can lead to more problems with toilet training and stress for your child.

If toilet training is not working after following the same toilet training plan every day, it is important to talk to your child's health care team. You need to know what to expect over time with your child's ability to control their urine and poop. Being realistic in what to expect is crucial to make toilet training as stress-free as possible.

For some children, their anatomy may not allow them to feel and control urine and poop in the usual way. In those cases, using large volume enemas (see Chapter 20) or taking medicines may be needed to help your child stay clean.

Children may develop challenging behaviors around toilet training. These may include holding

in urine and poop, hiding soiled underwear, or ignoring their body's cues to use the toilet. If your child has some of these behaviors, or are more prone to accidents, some of the following steps may help:

- Set a schedule for times to sit on the toilet and stick to it.
- Give rewards for using the toilet and getting good results.
- Withdraw things your child might want for behaviors that do not help with toilet training.

Reach out to your child's health care team for help if your child continues to have problems with toilet training.

Troubleshooting Accidents

Accidents will happen from time to time. These accidents occur whether your child can control their poop on their own, or if enemas are used to flush their colon each day.

Following are some common reasons for what may cause an increase in accidents:

- Changes in diet
- Changes in routine
- As children get older, they start to struggle for more independence. Sometimes this results in them not wanting to take their medicines that help manage their bowels.
- Constipation can also be a key issue when accidents start happening after a long period of being accident-free.

Your child's health care team can help you learn the cause of the accidents and find a solution that works. It is helpful to keep a record of your child's diet, medicines, bowel movements, when accidents begin, and how many accidents occur. This can help you, and your health care team learn what has caused the sudden change.

Toilet training for children with ARMs will be different for each child. Therefore, you must have realistic goals for your child before beginning toilet training. It is also important to reach out to your health care team early when issues arise.

While not all children with ARMs can feel and control poop and urine, most can stay clean and in regular underwear when in school and social settings.

With the help of your child's health care team, toilet training can be managed. This allows your child increased independence as they move into the school-age years.

20. Managing Constipation & Soiling

Constipation and soiling, or accidents with poop, are common problems for children with ARMs. Therefore, learning how to manage constipation is essential. Laxatives such as Senna or Miralax and using enemas are the first steps. It's critical to avoid constipation and to have your child empty their rectum of poop daily. This keeps the rectum from stretching. If the colon stretches too much, it is more difficult for your child to feel and control their poop, leading to accidents and soiling. If you manage constipation early after birth, toilet training becomes more successful when the time comes.

Medicines to Treat Constipation & Soiling

When we think about treating constipation with medicines, we usually consider stimulant laxatives like **Senna** or **Bisacodyl.**

The job of stimulant laxatives is to help push the poop out of the body. The starting dose depends on your child's size. The dose will often be changed until it reaches a level that helps your child have a poop every day, and an X-ray shows that poop is no longer in the colon or rectum.

While laxatives work well to help move poop through the colon, a side effect of using laxatives is that they often cause the poop to be liquid. To help bulk up the poop and keep it more solid, you may be asked to use a **water-soluble fiber.** Water-soluble fiber is important because, as your child gets to potty training age, a formed poop will be easier to control than if it is liquid. Some of the fibers used include **Nutrisource, Metamucil, Citrucel,** or **Pectin.**

Nutrisource has the advantage of being tasteless, odorless and does not change the texture of the food or liquid you put it in. For this reason, children seem to accept this best. However, Nutrisource does not work for every child, so other fibers can be used, such as **Metamucil** or **Citrucel.**

One of the problems with fibers like **Metamucil** or **Citrucel** is that they change the texture of the liquid you mix it with. This may cause small children to reject it. Some tips used by parents to help with this problem include:

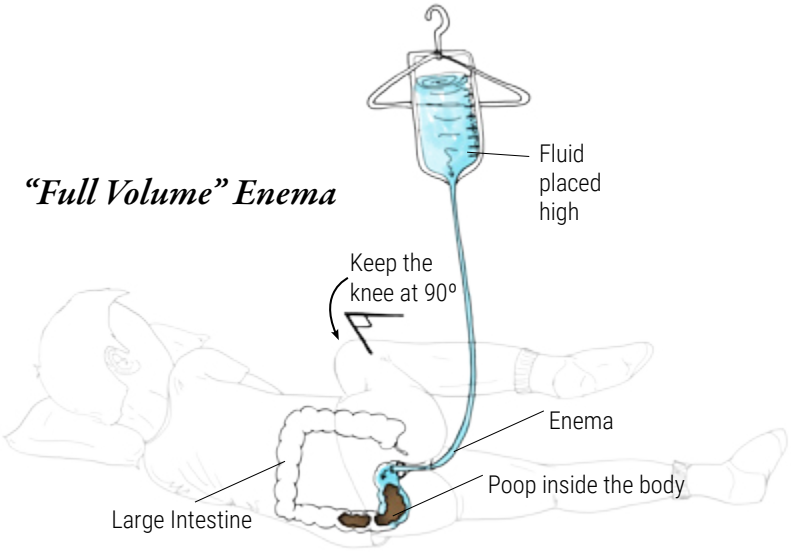
- Shaking the fiber and liquid to mix it together better
- Using a straw
- Providing rewards.

While it's best to limit sugary drinks, sometimes adding fiber to juice or Gatorade can help your child better accept drinking it.

Miralax®, which is a different type of a laxative, works by putting fluid into the colon to loosen the stool and making it easier to poop. **Miralax®** comes as a powder that is mixed into a drink. It is tasteless and children often like it better.

Rectal Enemas

You may be taught to do enemas to clean out your child's bowel to help with their constipation. Enemas can be used for a short time before medicines are used, or they can be used long-term, depending on your child's condition.



Enemas that are "full volume" use a bag that holds a solution that cleans out the bowel. Tubing with a balloon on the end of it goes into the rectum and is attached to the bag. The balloon blocks the solution from coming out while it is being given. The solution then goes into the rectum using gravity by hanging the bag up high. Your child will be asked to hold the solution in for about 10 minutes, and then sit on the toilet for 45 to 60 minutes to pass the solution out along with the poop.

You will typically mix the enema solution every day at home. The solution is made up of salt water The enema solution is usually made up of salt water or saline and irritants like **Glycerin**, **Castile Soap**, or **baby shampoo** that cause the bowel to empty. This solution might be changed until the right mixture is found that helps empty the bowel.

Bowel Management

As your child grows and begins to work on toilet training, they will sometimes need to have a bowel management program.

Some children with ARMs will toilet train without being in this formal program. Others will have problems controlling their poop and getting out of diapers.

Bowel management programs can help. They are typically a week-long program to help your child stay clean from poop and to be accident-free.

There are several office visits, X-rays, and enemas, along with reporting your child's patterns of pooping every day.

After reviewing all the tests and doing an in-depth evaluation, your healthcare team will talk with you and your child about goals for the week. Your child is then given either laxatives or enemas to help with toilet training. You and your team will work together to decide on any treatment and a schedule.

21. Surgery for Constipation & Soiling

If after all medical treatments for constipation and soiling have been tried but your child continues to have problems, your child may need a type of procedure called **antegrade enema**.

Antegrade enema procedures, including the malone procedure (also known as **appendicostomy**) or a cecosotmy procedure, allow the solution used to flush the poop out to be given into where the colon begins, instead of at the end, or through the rectum.

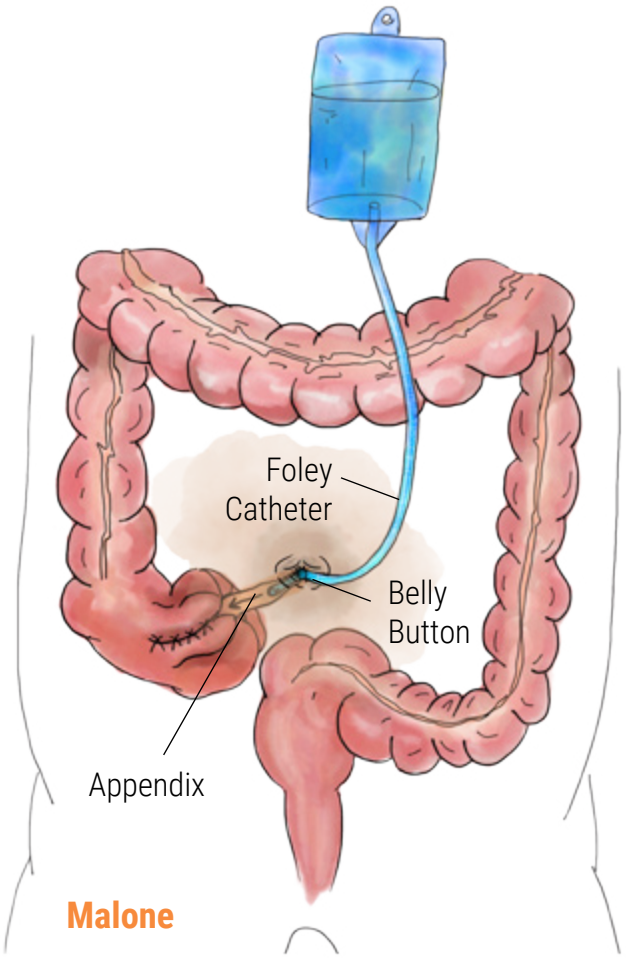
Malones and **Cecostomies** are also used for older children who can't stay clean and are ready to manage their bowels themselves. Most older children say that they like the independence and privacy that the procedure gives them.

Usually, rectal enemas will be tried for a while before having surgery to make sure your child will tolerate it. The trial period allows everyone to have an idea of what works best for your child.

Malone Appendicostomy

The Malone appendicostomy creates a channel from your child's bowel to the skin using the appendix or bowel. The tube from the flush solution is put into the channel so that the solution then goes directly into the channel and colon and not through the rectum.

Surgery to create the Malone is done through small cuts in the belly using a camera and small instruments (**laparoscopy**). However, sometimes a larger cut may be needed. Most of the time, the surgeons will make the opening of the appendix in the belly button (**umbilicus**) so that it's hidden and not as obvious. If your child has had other surgeries, the Malone is put in the right lower part of the belly. **A one-way valve is made using the colon.** This keeps poop and fluids from coming out from the Malone and allows your child the freedom to bathe, shower, and swim without concerns.



During surgery, your surgeon will likely leave a tube such as a **Foley** (urinary) type catheter, inside the **Malone channel** as it heals. This tube will usually stay in place until your follow-up visit 4 to 6 weeks after the surgery. After that, the tube is removed. Next, you will be taught how to put the tube into the channel, or to **catheterize** it, so that you can give the flush solution.

Cecostomy

A cecostomy is where a tube is put through the belly wall into the opening of the first part of the colon, or the cecum. The most common type of tubes used are **Mic-key**™, **MiniONE**™, or **chait tube**.

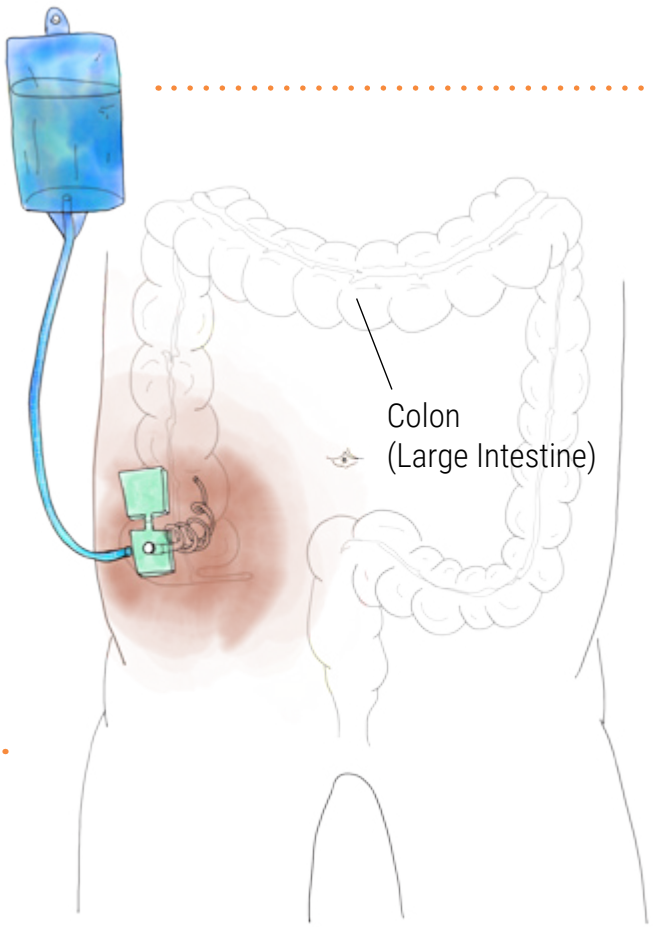
Surgery is typically done through small cuts (**laparoscopy**). Sometimes a larger cut may be needed. Unlike the Malone, this tube is usually left in place for several months and replaced by the surgeons or a radiologist, if needed.

Pros and Cons of a Malone vs. Cecostomy

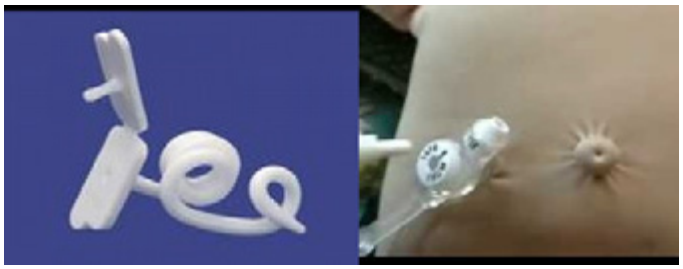
Deciding between these two options will be up to you and your team. One thing to consider is that typically with a malone, the child will need to have a tube placed into their every day. If your child will not be comfortable with this or has complex sensory issues, a cecostomy may be easier since the tube stays in all the time.

However, Cecostomy tubes do need to be replaced every 3 to 6 months. This is easily done in the hospital with or without a little sedation.

If your child's appendix has been removed, surgeons can create a special flap from the colon. This procedure is called a **continent neo-appendicostomy**. The length of surgery and recovery time is a little different, but it still works the same way as an appendicostomy.



Cecostomy



22. Surgery for Urine Control

Potty training with ARMs can be a challenging journey. Depending on your child's ARM, your doctors will decide the best way for your child to control their flow of urine.

If your child has a complex or high ARM, like a bladder neck fistula ARM or cloacal malformation, they may have problems toilet training.

Children with defects of the sacrum and spinal cord, like tethered cord (see chapter 5), have less chance of normally controlling their urine. Even when a child has a more normal anatomy of the spine and sacrum, only 75% of children with ARMs have **Customary Urinary Continence (CUC)**, which means less than one accident a week during the daytime.

Reaching CUC can vary for your child with an ARM. Therefore, urologists may advise you to give your child an extra year or so to try potty training. Also, it is not uncommon for your child to have had several surgeries or hospital stays that may delay their toilet training.

If your child cannot control their urine by a certain age, a **video urodynamic study (UDS)** is usually done to find out possible reasons why. The UDS tests show how well the bladder, muscles or sphincters, and urethra hold and release urine. These tests also can show how well the bladder works and why there could be leaks or blockages. This test is done while the child is awake and a tube with a balloon on the end of it (**Foley**) is put into the bladder before the test.

Sometimes, **Clean Intermittent Catheterization (CIC)** with or without medicine is all that is needed to stay dry. CIC involves putting a sterile tube into the urethra to drain the bladder of urine several times a day. This allows your child to stay dry between CIC times and protects the bladder from being overfilled with urine. **Overfilling the bladder harms the kidneys.**

However, your child may need surgery to be able to control their urine. There are different types of surgeries to safely stay dry. What surgery or combination of surgeries are needed will vary, depending on your child's condition.

Your urologist will always make decisions on what treatments are needed based on what needs to be done to protect your child's kidneys.

If there are problems with urine being drained or frequent infections because of the bladder, long-term damage to the kidneys can occur. Kidney failure and the need for cleaning the blood regularly since the kidneys are not working normally (**dialysis**), or **kidney transplantation** may be needed. Children with ARMs are already at higher risk for kidney damage if they are born with abnormal or missing kidneys.

Here are some surgeries that your urologist may discuss with you.

Bladder Neck Surgery

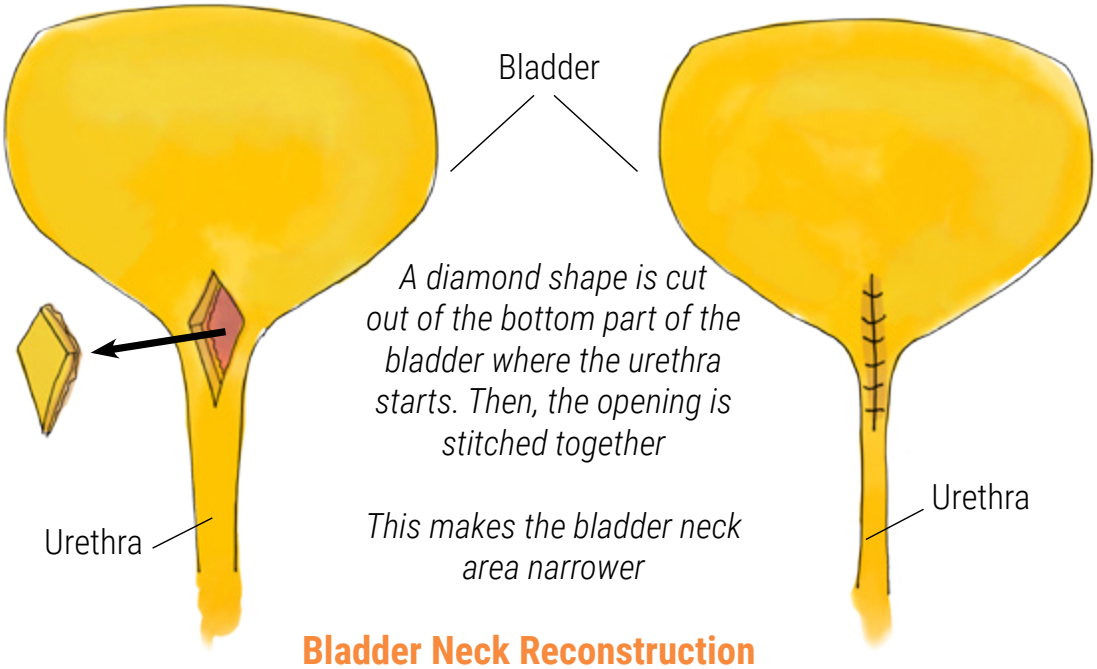
In ARM and cloacal malformations, the exit from the bladder, or bladder neck, may not function well. As a result, your child may leak urine when they cough or laugh, and sometimes they may constantly leak urine. If this occurs, your child may need surgery to tighten or close the bladder neck to prevent leakage.

Bladder neck "tightening" is done by narrowing down the size of the urethra and making it smaller. In addition to making the urethra smaller, a bladder neck sling is done to kink the urethra and suspend it to the pubic bone. Known as a **Bladder Neck Reconstruction with Sling**, this surgery prevents leakage from the urethra when the child is standing, running, or doing other activities.

The Sling is done rather than just the **Bladder Neck Closure** because it allows a small tube to be put into the urethra to drain urine if needed.

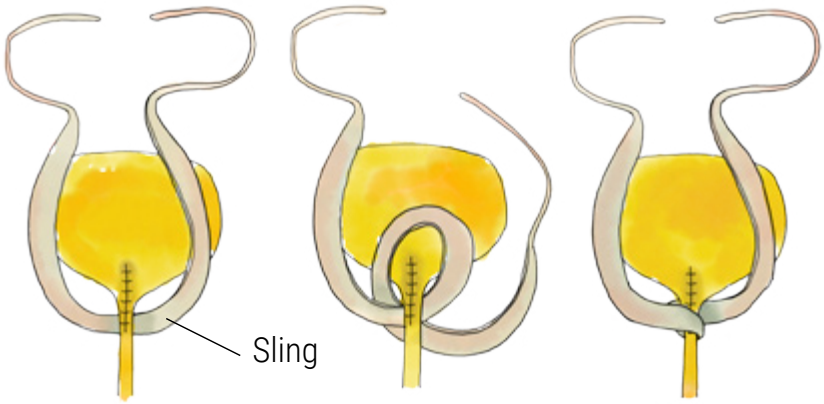
Sometimes, a child's urethra is too short, or the anatomy does not allow surgery to tighten the bladder neck. In this case, your child will have a **Surgical Bladder Neck Closure**. This surgery closes the natural exit of the bladder for good, so it is not often a first choice. However, when a major surgery of the perineum in girls is done, a **Surgical Bladder Neck Closure** may be the best choice for her to remain dry.

A **Bladder Neck Closure** needs a different path for the urine to drain. Usually, either a **Continent or Incontinent Abdominal Urinary Diversion** is done. Both are discussed later in this chapter.



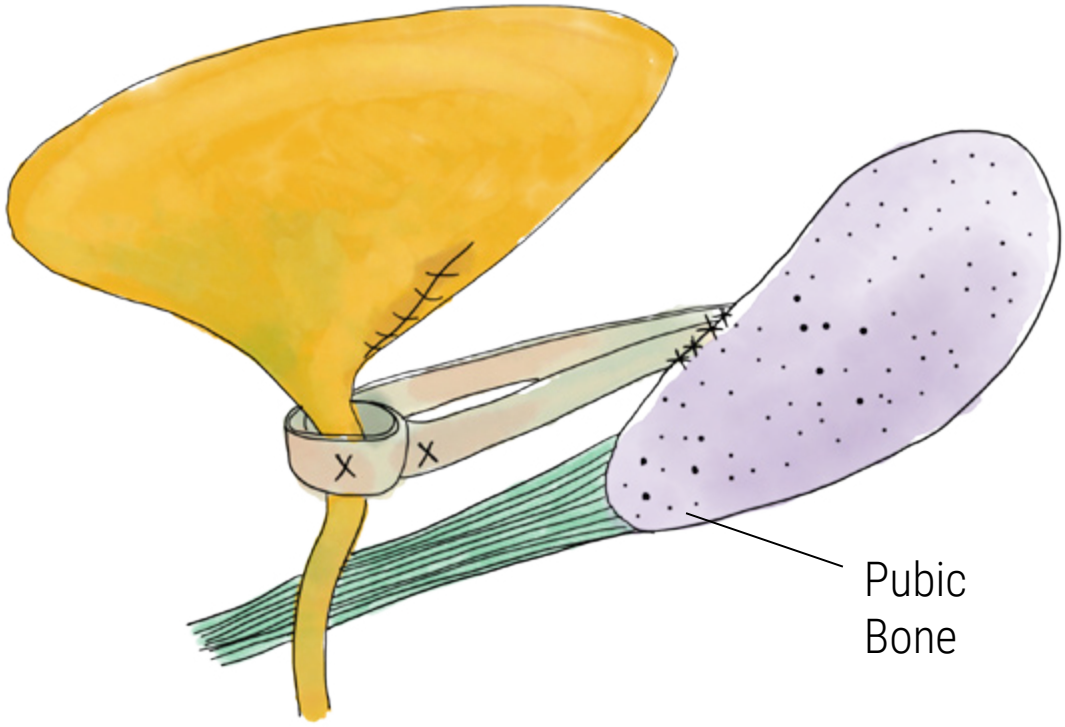
Bladder Neck Reconstruction

Another less common choice for bladder neck surgery is to use an **Artificial Urinary Sphincter** around the bladder neck. The **Artificial Urinary Sphincter** is a device that can act like the normal muscle, or **sphincter**, that opens and closes to allow the flow of urine. Usually, it stays in a closed position. The child activates the device to open it to empty the bladder, either by passing urine or by using the small tube to drain the urine. However, since this device fails 50% of the time over 15 years and needs replaced or fixed over time, doctors do not usually recommend it for children.



Bladder Neck Reconstruction With Sling

A sling is placed in the above way and stitched to the pubic bone to create a curve in the urethra tube. This helps the bladder control urine throw better



Abdominal Urinary Diversion

When your child has a **Bladder Neck Closure,** a different path is needed for their urine to drain. Therefore, either a **Continent or Incontinent Abdominal Urinary Diversion** is done.

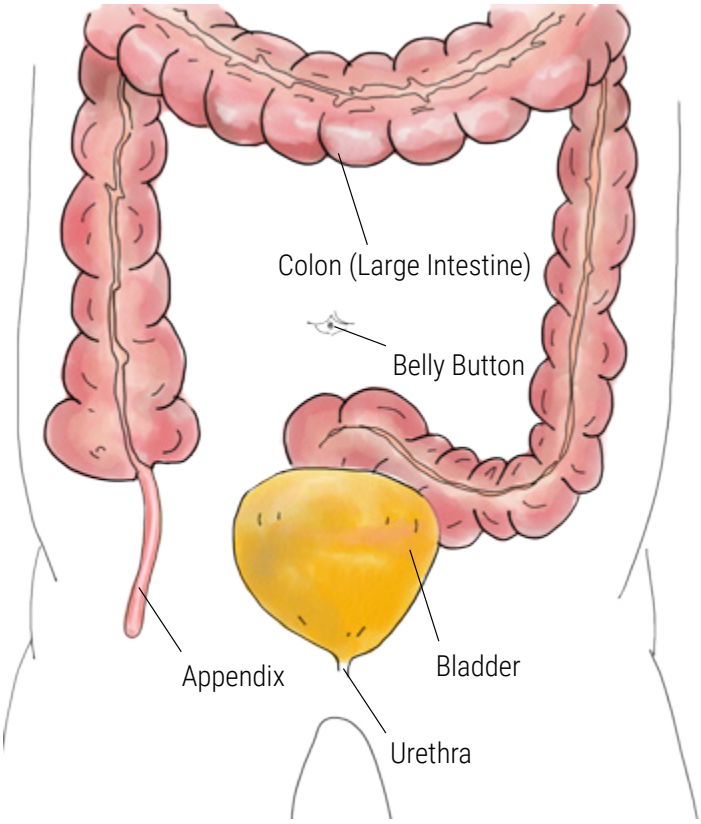
1. Continent Urinary Diversion

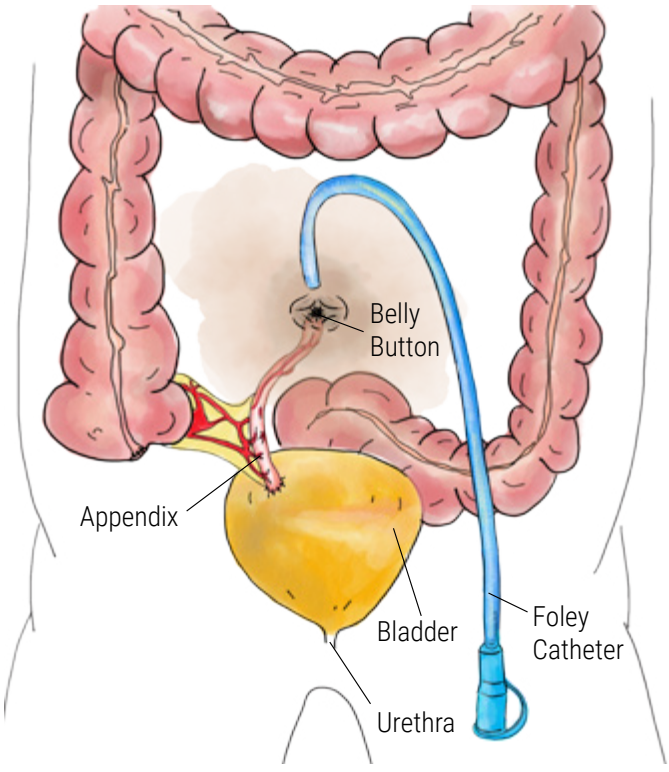
▪ Mitrofanoff Catheter

A Mitrofanoff is a channel made during surgery that goes from the bladder to the skin under the belly button. The appendix is used as the connection between the bladder and the outside of the belly. A small tube can then be passed into this connection to empty the bladder.

The tube is stitched in and will be kept in this channel for 3 to 4 weeks to allow healing.

At the follow-up visit, the stitches and tube will be removed. Then, you will be taught how to insert a tube that will only be used to drain urine from the skin opening every 3 to 4 hours.

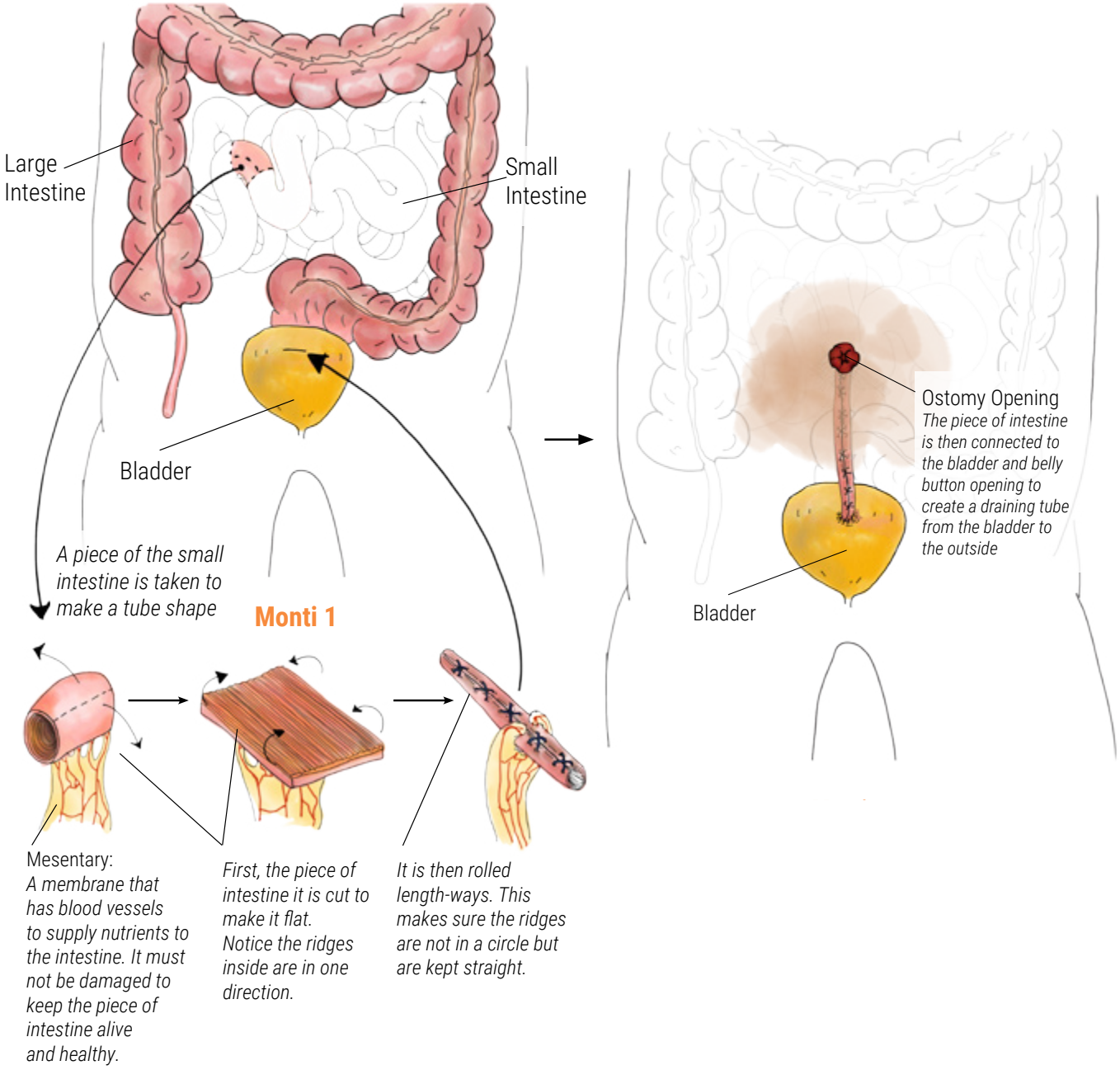




▪ Monti

Some children do not have an appendix because it was absent when they were born, it was surgically removed or used to create a **Malone**. In this case, a very short piece of the small bowel or intestine is used to make the channel the same way the appendix would have been used. This surgery is called a **Monti**.

Sometimes, this type of surgery has problems with narrowing of the channel at the skin and more surgery is needed. Opening the channel is usually a minor surgery. However, if there are problems with passing the tube or urine leaks it may need more than minor surgery. Fortunately, these problems are rare.



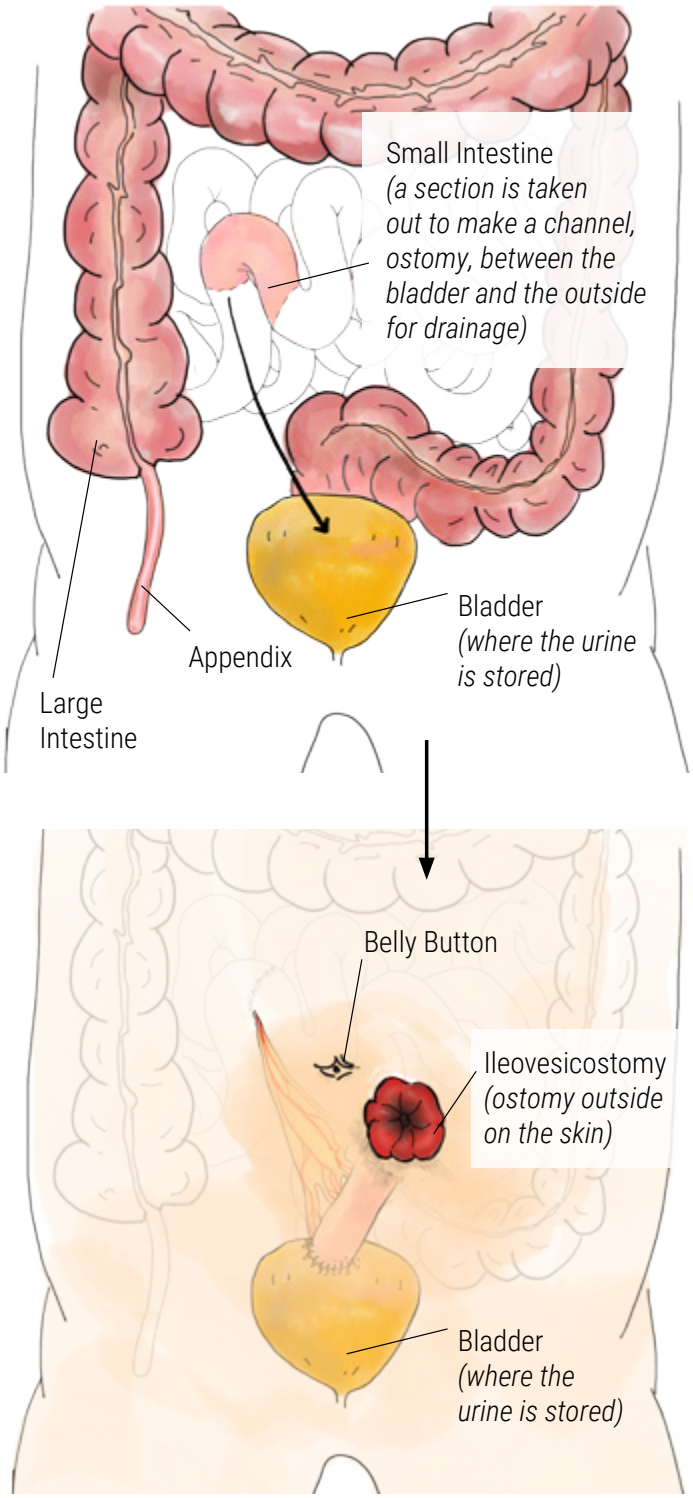
2. Incontinent Urinary Diversion

▪ Ileovesicostomy

Incontinent Urinary Diversion is a way of draining the bladder into a **stoma bag** rather than putting the small tube into a channel on the belly to drain the urine. Incontinent Urinary Diversion is like a colostomy or ileostomy.

Incontinent Urinary Diversion is sometimes used for only a short time, but most often it is done to always drain the urine in this way. This type of urinary diversion uses a stoma bag on the belly and allows the bladder and kidneys to drain at low pressure.

It is often a very safe way to, drain urine mainly in a child with poor kidney function or frequent infections. Some families decide that they would rather manage a stoma bag than use the tube every 3 hours to drain urine.



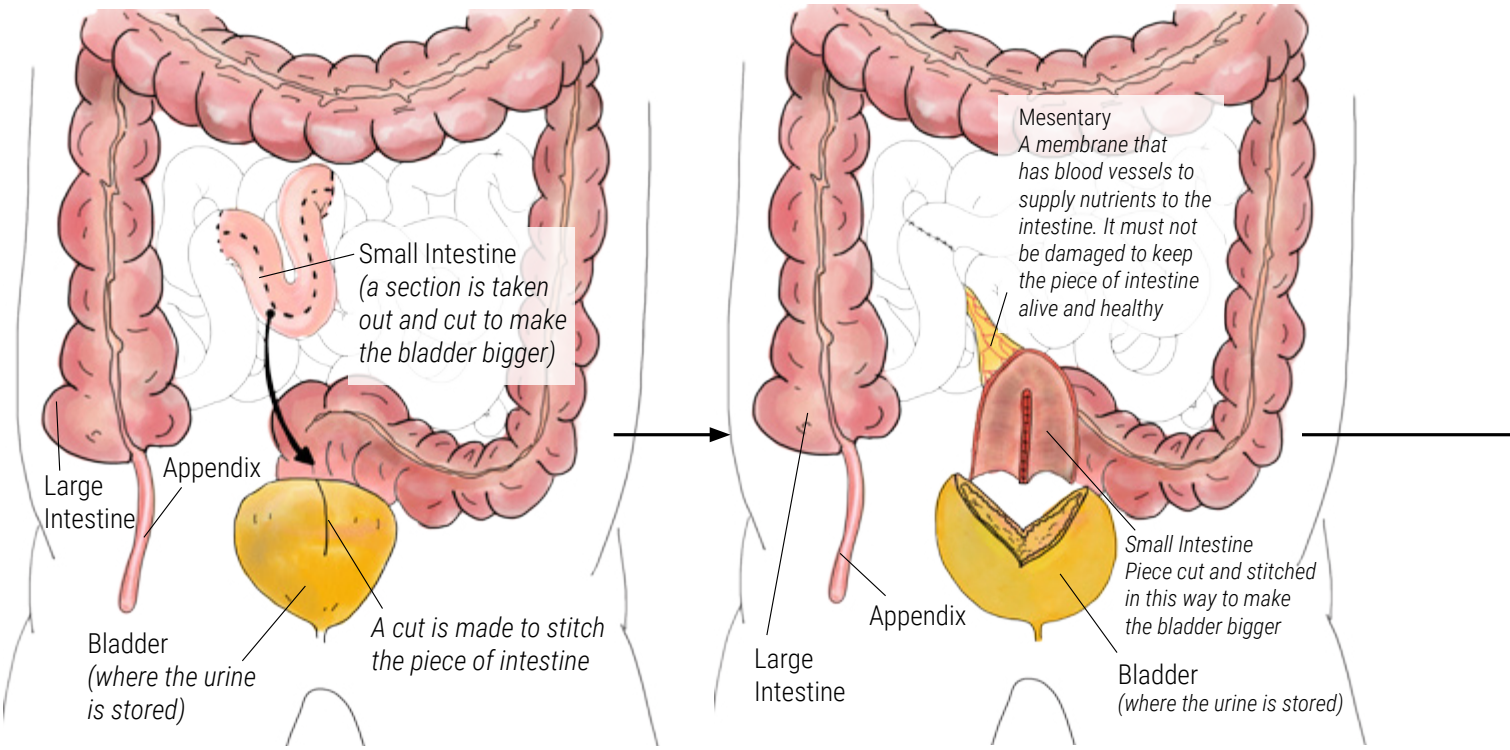
▪ Bladder Augmentation

In some children with ARM, the bladder may be small and overactive, which means it contracts often with low amounts of urine leading to leakage, or stores urine in the bladder at very high pressure. These conditions result in damage to the kidneys. In this situation, the bladder needs to be enlarged to hold more urine at low pressure, avoiding kidney damage.

The bladder can be made larger by taking a piece of the small intestine or colon and sewing it to the top of the bladder. This surgery is called **Bladder Augmentation** and allows the bladder to store larger amounts of urine at safe pressures.

There are two benefits of Bladder Augmentation:

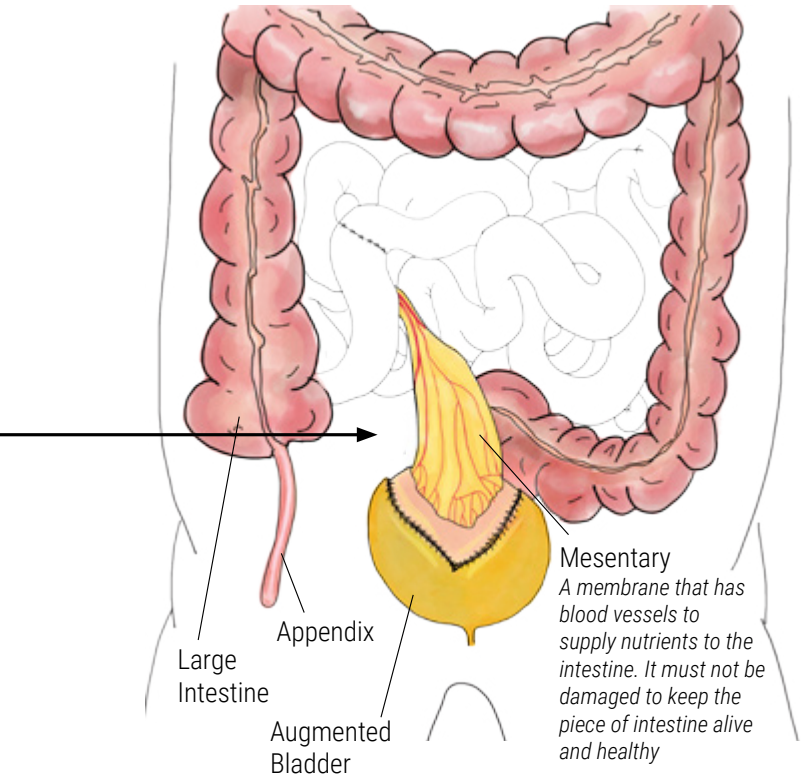
- The larger bladder allows for a longer time between the times when the bladder needs drained with the tube.
- The lower bladder pressures reduce pressure on the kidneys and improve overall kidney health.



There are, however, several disadvantages to bladder augmentation:

- The intestine used can produce mucus in the bladder leading to bladder stones and infections. Therefore, flushing the mucus out daily is vital.

- There is also a small risk of cancer within the bladder when you have intestine storing urine. The risk for developing this is very low and usually occurs after the age of 40. Because of this, it is advised that a yearly test that looks inside the bladder (**cystoscopy**) be done, beginning 10 years after the surgery.
- Augmented bladders can rupture, mainly after trauma such as a car accident. This risk is very low but the bladder needs to be checked right away if a trauma happens.



Due to these rare but high risks, urologists only do bladder augmentation when necessary. When the overall benefits outweigh the small risk of problems, this surgery usually protects the kidneys from the serious and more common risk of kidney failure. Long-term kidney failure could lead to dialysis and kidney transplantation.

In conclusion, some children with ARMs may need different ways to control their urine and become potty trained, depending on how complex their ARM is. Your child's healthcare team will help you during this time, so it is important to ask questions and discuss concerns to allow for smooth potty-training.

23. Moving from Pediatric to Adult Health Care

Health care transition means getting ready to move from child health care to health care as an adult.

The goals are:

- To improve the ability of youth and young adults to manage their own health care.
- To know how to use health care services to get the best health care results possible.

Health care during this transition should be a purposeful and planned. It needs to identify and address the medical, mental, social, educational, and vocational needs of teenagers and young adults with long-term physical and medical conditions. If teenagers do not successfully move from pediatric to adult health care, there can be major physical and mental health problems for them. **Poor long-term health outcomes, and misuse of health care resources can result if the transition is not done well.**

Youth with ARMs are especially at risk, because of ongoing challenges with surgeries, and physical, emotional, and social needs. Most youth with ARMs have a health care team working with them when they are a child that includes urology, gynecology, colorectal surgery, psychology, and social work. It is just as important to have those same resources and team members available into adulthood. Common issues that these youth may have during adulthood include soiling, ongoing constipation, reproductive (GYN) issues, sexual health concerns, and urologic problems, which may involve leaking urine.

The timeline for moving to adult health care begins at 12 years of age. Your child's medical team should prepare you for adult health care and help you understand the specific and safe plan used to make the move as seamless and easy as possible.

Age 12 — Youth and family become aware of the transition policy

Age 14 — Start the health care transition plan

Age 16 — Prepare youth and parents for how adult health care works and discuss transfer

Age 18 — Transition to care as an adult

Age 18-22 — Transfer care to adult medical home and specialists.

Age 23-26 — Integrate young adults into adult care

As your child nears the teenage years, it is vital for them to:

1. Know how to manage their health care:

- Understand and manage their medical condition and be able to explain it to others
- Know why they take a medicine and what it does
- Know their allergies and what medicines NOT to take
- Know their symptoms, including those that need them to see a doctor quickly
- Know what to do in case of an emergency
- Explain their customs and beliefs to others and how they affect health care treatments and decisions

2. Know how to work within the health care system:

- Learn how to find their doctors' contact information
- Make their own doctors' appointments
- Think of questions to ask before visiting a doctor
- Know where to get medical care when their doctor's office is closed
- Know how to fill out medical records
- Choose a pharmacy and learn how to get refills for their medicines

Recognizing the need for moving into adult health care is the first step for a successful transition. Your team should give you and your child general health knowledge tools each year to assess how ready you are for transition. This helps the team find out any issues that may need to be worked on together, so when your teenager reaches 18 to 22 years of age, they feel confident and ready to move into adult health care. Choose the adult health care team early so that everyone is comfortable with the transition of care. Additional health care team members may be needed in the adult world. These members may include pelvic floor physical therapists, fertility specialists, and high-risk maternal and fetal medicine doctors as sexual health and reproductive issues become more critical. Your health care team members are committed to working with you during this important time.

24. Gynecologic (GYN) Care for Baby Girls with ARMs

Baby girls born with ARMs have unique GYN needs. Many of these babies may have related GYN problems, including problems with their external genitalia, vaginal opening, vagina, and uterus. Because of this risk, it's important to have your baby's reproductive anatomy evaluated. This evaluation is done during office visits or while your child is having other tests or surgeries. Ideally, your child's GYN doctor or gynecologist works closely with your colorectal and urology doctors to help understand the anatomy, plan any necessary surgery, and make sure the future of your child's reproductive health is the best it can be.

Given the small size of the uterus and other reproductive parts in girls before the age of puberty, it may be difficult to learn the full anatomy of the uterus and if it will function normally. Therefore, more tests are done, such as ultrasound or MRI, as your child goes through puberty. These tests, and even surgery if it is done, helps the healthcare team learn more about your child's reproductive anatomy.

Even if your child has normal GYN anatomy, you may have concerns about their sexual or reproductive health because of the original diagnosis, surgeries, urinary or bowel function, or other VACTERL problems. **We advise that you follow-up with a GYN doctor as your child grows to discuss any concerns.**

Changes During Puberty and Managing Menstruation

The hormones that stimulate puberty are mainly made in the ovaries. Because ovaries are usually not a problem in a child with ARM, they normally progress through puberty at the usual age. As your child begins puberty, the ovaries will start making the hormone estrogen. This hormone develops the breasts and causes the uterus and lining of the uterus or **endometrium** to grow. Breast growth usually starts at age ten, and most girls have their first period (menarche) at age 12. Understanding the usual changes during puberty are important so that you can alert your healthcare team if you have any concerns.

Your GYN healthcare team may want an ultrasound done to continue evaluating the uterus and ovaries. They will check the anatomy of the uterus from the tests done earlier, to see if they match the anatomy after puberty. Also, a pelvic MRI might be needed to understand the shape of the uterus and the other reproductive parts.

It is important to know that the menstrual blood that comes from the lining of the uterus can get through the cervix and out the vagina. An ultrasound and sometimes a pelvic MRI, is helpful to make sure there is no blockage of this flow. If there is a blockage, these tests can helps your doctors learn where the blockage occurs. Sometimes, even if there is menstrual flow, a blockage may be found with ultrasound or MRI.

A GYN doctor will spend time asking questions about menses. Have periods started? How often do they occur? How long and how heavy is menstrual flow? Is there any cramping? Any other period-related symptoms? Do periods interfere with normal daily activities, such as school, sports, and hobbies? These questions help your doctors learn more about your child's menstrual health or any menstrual concerns. Your GYN healthcare team needs to know if school, sports, or family functions are affected by menstrual cramping. Also, report any use of large amounts of **NSAIDs** (non-steroidal anti-inflammatory drugs) needed for pain during menses. Some menstrual cramping may be normal, but if it is extreme, further evaluation for blockages or problems with the lining of the uterus (**endometriosis**) may be needed.

During puberty your GYN doctor should check your child's vaginal opening to make sure it is large enough for menstrual flow, tampon use, and vaginal sex. There are non-surgical and surgical treatments for an opening that is too narrow. Also, girls with ARM may have a vaginal septum or a wall of tissue that divides the vagina into two halves. Sometimes, this wall needs to be removed to comfortably use tampons and have vaginal sex.

Sexuality & Family Planning

Complete reproductive health care is important for all women.

A GYN doctor will provide birth control counseling, and will discuss safe sexual practices, including preventing and screening for STDs or sexually transmitted diseases. All women from age 9 to 26 years, and for some women ages 27 to 45, should get the HPV or human papillomavirus vaccine. This reduces the risk of HPV-related diseases, such as cervical cancer, anogenital warts, anal cancer, and some oropharyngeal cancers. Screening for cervical cancer and breast cancer should be done when the GYN doctor advises.

There are many ways to create a family. The ability to conceive and carry a pregnancy depends upon a person's reproductive anatomy (uterus, cervix, vagina, ovaries).

Knowing the anatomy and what surgeries have been done is important. This information helps your child's OB-GYN doctor to discuss any future pregnancy. Many women with ARM conceive, successfully carry a pregnancy, and deliver a healthy infant.

Ideally, every woman should visit with her OB-GYN doctor before getting pregnant.

A visit with your OB-GYN before becoming pregnant allows a discussion of:

- Maternal health
- The timing of pregnancy
- The safety of becoming pregnant
- Potential risks during pregnancy
- The anticipated way to deliver the baby.

Many factors impact whether a vaginal or cesarean section delivery for birth of the baby is needed. Also, the anatomy of the uterus may affect pregnancy risks, including preterm labor, preterm birth, and the position of the baby in the uterus.

Discuss family planning with your reproductive, colorectal, and urology healthcare teams. It is important to know how pregnancy and delivery may impact overall health, such as kidney health and control of bowels and urine. A healthcare team made up of many specialties including a high-risk OB-GYN doctor, colorectal surgeon, urologist, and anesthesiologist provides the best care for a safe pregnancy and delivery.

In conclusion, girls with ARMs can have unique GYN needs. Therefore, regular, and complete reproductive health care should be an essential part of long-term care.

25. How to Find a Quality Colorectal Team: Resources for Parents

When your child is born and you first hear the words "imperforate anus" (older term) or "anorectal malformation," you probably think to yourself, "what do you mean, my child doesn't have an anus? How is that even possible?" Your baby may have a birth defect that only occurs in one in 5,000 births, but hundreds of families are walking in the "same shoes" as you. Even though this may be a difficult journey you are beginning, you are not alone. There are many good resources for both you and your child.

After your baby's birth, they most likely went to a children's hospital and was seen by a pediatric surgeon. All pediatric surgeons have been trained in pediatric surgery. However, very few have the special training for pediatric colorectal surgery. Many doctors will tell parents that their child's ARM can be corrected with surgery and then the child will be fine. This is often true if the surgery is done correctly.

It is very important to find a pediatric colorectal surgeon to make sure you have the best outcome, including follow-up care. You can find skilled colorectal surgeons by looking for children's hospitals with colorectal centers. **The colorectal center should have many specialty doctors, including those who treat urinary tract problems (urology), gynecology (GYN), gastroenterology (GI), and nephrology (kidney), in addition to surgery.**

Each year, more of these colorectal centers with all the specialties are becoming available. You can find these centers by searching "pediatric colorectal center" on the internet.

Questions to Ask

Once you have found a colorectal center, you should ask the surgeon several questions to make sure they have the desired skill level.

Some questions to ask include:

- **How many ARM repairs have you done?**
- **How many ARM patients do you see in a year?**
- **What surgery will you do to correct my child's ARM?**
- **Will there be other doctors involved with the surgery, if yes, who are they?**
- **How long will my child be in the hospital after surgery?**
- **Will there be any "prep" before surgery? Are there follow-up office visits after surgery?**
- **Do you offer any program that will help my child control their bowels?**

The answers to these questions should help reassure you that you are in the right place. If you are not sure, please ask more questions or get a second opinion.

In addition to finding good, quality medical care for your child, it's also important to find emotional support for yourself and your child. Families often find it hard to talk about their child's ARM diagnosis outside of the family. It's also unlikely that you will find any support groups in your local area or even another family with an ARM diagnosis. It can seem very isolating and lonely at times. However, there are several groups on **Facebook** created by ARM families. There are also a few groups that offer support for families with ARM, including **Pull-thru Network, Inc** and **The ONE in 5000 Foundation**. Both these groups have websites with great information that can help families with a child who has ARMs connect with other families.

Pull-thru Network, Inc

www.pullthrunetwork.org

Pull-thru Network, Inc was formed in 1988 and is the longest running support group for families living with the challenges of ARMs and Hirschsprung Disease.

In addition to their website, Pull-thru Network publishes a magazine, "**PTN News**," for their members three times a year. Every other year, they host a national conference for families to meet in person and learn about the newest information about ARMs and Hirschsprung Disease. Most of their speakers are doctors and nurses from the major colorectal centers around the United States.

The ONE in 5000 Foundation

www.onein5000foundation.org

The ONE in 5000 Foundation began in 2018. It is an international group that brings greater awareness of ARMs around the world. **Their vision is based on their AIMS Program: Awareness, Information, Medical and Support.** Their hope is to make sure that every person affected by an ARM has the support and medical care they deserve.

There are several books written by or about colorectal experts, parents of children with ARM and ARM patients, themselves. All these books are available on **Amazon**.

- **"Monologues of a Pediatric Surgeon"** by Alberto Peña, MD, and **"The Work of Human Hands"** by G. Wayne Miller about Hardy Hendren, MD are excellent books about two of the pioneers in anorectal surgery.
- Devesh Dahale wrote about his son's first year of life with ARM in his book, **"The 5000th Baby."**
- J.T. Mestdagh writes about his life growing up with VACTERL in **"Untether."**
- After keeping his ARM diagnosis, a secret for over 50 years, Greg Ryan writes about his life in the book **"A Secret Life: Surviving a Rare Congenital Condition."** Greg created the One in 5000 Foundation, which has published personal stories of people with ARM titled **"Rare and Resilient: One in 5000 Anthology."**

As difficult as this diagnosis may be, today there are many medical centers and resources that will help you get the information you need to find and provide the best care possible for your child and your family.

Always remember, you are not alone!