Imperforate Anus

A guide for parents

What is imperforate anus?

In most individuals, the anus opens to the skin through the muscles at the bottom of the pelvis, known as the anal sphincter. These muscles allow control of the evacuation of the stool from the intestine. Imperforate anus is a birth defect in which the anal opening is absent, or not in the correct location. If the anus is absent, the intestine may end blindly or may be connected to the urinary system or vagina by an opening called a fistula. These abnormalities may prevent a newborn from passing bowel movements after birth (if there is no opening to the skin) or cause bowel movements to be passed through the fistula into the urinary system or vagina. This birth defect occurs in approximately 1 of every 5,000 newborns and is about three times more common in boys than girls. Some children present with a more complex birth defect called a persistent cloaca. This is an abnormality in which the urethra that drains the bladder, the vagina that drains the uterus and the rectum, or very end of the large intestine, join into a single common opening to the skin. There is a spectrum or range of abnormalities in children with persistent cloacas. This is one of the more common examples, however the specific anatomy will vary from newborn to newborn. Your child’s surgeon will explain your child’s specific anatomy and surgical plan.

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**IMPERFORATE ANUS - FRONTAL VIEW**

- **Normal Anatomy**
  - Large intestine
  - Rectum
  - Anus

- **Imperforate Anus**
  - anal opening is missing or closed off
How is imperforate anus diagnosed?

If the infant does not pass stool or passes stool through an abnormal opening, this is considered imperforate anus. This will be determined during a physical examination. If the stool cannot pass, the infant will develop an enlarged or swollen abdomen and may begin vomiting.

What causes imperforate anus?

There is no known cause. Most cases of imperforate anus are isolated and do not run in families. The mother of a child with imperforate anus has about a 1 percent chance of having future children with this defect.

What other health problems are associated with imperforate anus?

About half of newborns with imperforate anus can have other health problems. Of these problems, defects (abnormalities) in the genitourinary system are the most common. Other defects include vertebral or spinal cord problems (such as tethered cord), bladder and kidney problems, heart problems, and esophageal or limb abnormalities.

What is tethered cord?

Tethered spinal cord syndrome is a neurological disorder caused by abnormal tissue attachments that restrict the movement of the spinal cord within the spinal column. Normally, in the absence of a tethered cord, the end of the spinal cord floats freely, and is without attachment. Abnormal

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**Instructions:**

Inflate the foley catheter balloon with 30 cc of air to test it prior to insertion, then deflate the balloon prior to insertion.

Pour the prescribed amount of warmed saline into the bag. Let it flow through the tubing all the way to the end, to empty the air from the tubing. The volume recommended will be ordered by the surgeon and surgical nurse.

Have your child position him or herself on the knees with the buttocks up in the air. Have a pillow or towel available to rest his or her head on.

Lubricate the tip of the catheter and place the catheter into the rectum 4–5 inches. Inflate the balloon to no less than 15 cc and no more than 30 cc and pull back gently until it stops. Keep some tension on the tube so the saline does not leak out. If the saline leaks out, more tension is needed on the catheter to seal the anal opening.

Connect the enema bag to the catheter and let the solution infuse over 5–10 minutes. This rate should be comfortable, and not cause pain.

Let the solution stay in the intestine for another 10 minutes, in the buttock up position, with tension on the tubing to prevent leaking.

Move the child to the toilet, holding tension on the catheter, then deflate the balloon and the catheter will fall out. The child may need to sit on the toilet for 45 minutes to finish evacuating stool. Toys, books, music or other entertainment will help pass the time.

Wash and reuse the catheter and drain the enema bag for the next treatment. Keep a diary ever day to monitor the progress of the enema program. Include the amount of enema solution used, the time needed to empty the intestine and if the child had accidents during the day. This will help the surgeon and surgical nurse determine if any changes are needed to the program.

**Support groups**

Pull-thru Network, an organization devoted to children born with anorectal malformation, colon disease and associated diagnoses.

- **Pull-thru Network, Inc.**
  http://www.pullthrunetowrk.org
  2312 Savoy St.
  Hoover, AL 35226-1528
  (205) 978-2930
  PTNMail@chartner.net

There is a Bay area chapter of the pull through network:

- **PTN Support Bay Area**
  P.O. Box 40342
  Berkeley, CA 94704-4342
  PTN.BayArea.Owner@gmail.com
defects often have more difficulty controlling bowel movements and may benefit from daily enemas to keep the colon clean. Enemas can begin when the child is between ages 3 and 4.

To get started, it is helpful to have children sit on the toilet after each meal. This should be a relaxing time. It is helpful to praise or reward children when passing urine or stool in the toilet. If children are having soiling accidents in between sitting on the toilet, contact your surgeon’s office for advice.

In general, positive reinforcement, such as praising/rewarding as described above, is very helpful, while negative reinforcement, such as expressing disappointment or becoming angry, can be very harmful.

**Why does my child need a constipating diet?**

Some children, in particular those with high defects, may have problems with permanent fecal incontinence, or lack of control of bowel movements. These children are best managed by diet and medications that constipate combined with daily enemas to clean the colon of stool (described below). This diet begins with constipating foods and Loperamide (Imodium), a medication to decrease stooling frequency. This diet is very strict and specific instructions will be provided to you by the nutritionist. Once the diet is underway and the child stays clean between bowel movements in the toilet, new foods may be introduced one at a time. The foods that increase colon motility or cause diarrhea should be eliminated and the foods that do not may be continued. It is recommended to keep track of these additions/deletions in a diary of the food consumed and the resulting stooling patterns.

**When are enemas necessary?**

For select children, particularly those with high or complex defects, enemas control bowel movements and keep the large intestine clean between enemas. An enema given each day should provoke a bowel movement. Ideally, this is followed by no stool or accidents for 24 hours. This is achieved through trial and error.

Enema solutions are usually made of saline. Large volumes are usually required for children with high defects. Warming the enema solution to body temperature may be more comfortable. Monitoring bowel movements after enemas is necessary to determine effectiveness. If saline alone is ineffective, additives such as liquid glycerin or liquid castile soap may be helpful. Your surgical nurse will give you instructions on using these products.

It is recommended to give the enema following a meal at the most convenient time of day for your family allowing about one hour to deliver the enema and evacuate the intestine. Do this at the same time each day.

**Saline solution:**

- 1000 ml tap water and 1 1/2 teaspoon salt (do not adjust the amount of salt).

**Equipment:**

- Enema bag (or feeding bag) and hook to hang to allow the saline to flow by gravity. The rate of the flow of saline can be controlled by using the roller clamp on the feeding bag,
- 22 or 24 Silicone Catheter with 30 ml balloon
- 30 ml syringe
- Water soluble lubricant

**What is the treatment for imperforate anus?**

Imperforate anus requires one or more operations. The operation(s) vary with the specific abnormality. Some children with high or complex defects will have a colostomy first, followed by an operation to bring the lower intestine to the skin through the anal sphincter, and a third operation to close the colostomy. Children, with lower defects may have one operation to move the intestine to the skin within the center of anal sphincter. The type of operation(s) needed will be determined by your child's surgeon according to your child's condition and the specific type of defect. In general, it is best to complete these surgeries by the time the infant is about six months to nine months old, but this may depend on the presence and treatment of other associated defects.

**Why are anal dilations, following repair of the imperforate anus, necessary?**

Anal dilations are necessary because the size of the new anus after surgery may be smaller than normal or may not enlarge normally as the child grows. Dilations slowly stretch the new anus to the normal size for the child. When dilations are not done, the anus may heal closed.

Your child will receive a set of anal dilators before leaving the hospital. Anal dilators are plastic or metal instruments that are available in several sizes. The sizes are graduated from small to large and are numbered. Please bring these to your child's first surgical post-operative appointment. The sizes needed to dilate your child's anus are determined by the surgeon's exam and your child's age. The surgeon and surgical nurse will teach you to perform the dilations in the office, starting with the size that fits most comfortably and increasing the dilator by about one size per week until the goal size is reached. Dilations are usually performed twice each day until the goal size is reached. The dilations are then tapered or stopped.

**Will my child have constipation?**

Constipation is common in children born with imperforate anus. This problem is reported to be worse in children with low defects. Children with high defects are more likely to have difficulty controlling bowel movements and are likely to have accidents. Preventing constipation is important to prevent swelling of the large intestine. This can lead to worsening constipation. Severe and persistent constipation can lead to loss of control of bowel movements and soiling or accidents.

Treatment varies with the type of the defect. Children with high defects may require daily enemas to control bowel movements Children with low defects have good potential for bowel control however they are more likely to have constipation and require laxatives to control bowel movements.

**Why are laxatives needed?**

Children with imperforate anus can have abnormal movement or contractions of the intestine, abnormal sensation or feeling as well as sphincters muscles that are not completely formed. All of
these affect the control of bowel movements. Laxatives are used to stimulate a bowel movement and keep the large intestine empty of stool every day. It is best to give the laxatives once a day, at the same time every day, to stimulate a predictable bowel movement. The amount to give is determined by trial and error and is often a large dose. In addition to laxatives, fiber is recommended to keep the stool soft and prevent diarrhea.

Once the laxative and fiber are started, it is important to monitor the bowel movements carefully. If stools are runny, decrease the dose of laxative and increase the dose of fiber. If bowel movements are hard to pass, increase the dose of laxative. If there is no bowel movement within 24 hours after administration of the laxatives, increase the laxative dose.

**Suggested laxatives include**

- **Little Tummys**
  - Sennosides 8.8 mg per 1.0 ml
- **Ex-Lax**
  - Chocolate Stimulant Laxative Sennosides 15 mg or 25 mg per square
- **Pedia-Lax**
  - Dissolve strips Sennosides 8.8 mg per strip
- **Senna containing tablets**
  - Multiple preparations and strengths available

**Types of fiber**

There are two types of fiber, soluble and insoluble.

**Soluble fibers** dissolve in water and slows the rate of digestion in the intestine while forming a gelatinous substance to keep stool soft. This fiber can contribute to intestinal gas formation.

If your child is taking laxatives, supplementing with soluble fiber will help bulk the stools. The amount of fiber recommended for children with imperforate anus is:

- 10 grams + one gram for each year of age.

Soluble fiber supplements include:

- **Benefiber**
  - Wheat dextrin, two rounded teaspoons provide three grams soluble fiber.
- **Fibersure**
  - Vegetable fiber inulin, one rounded teaspoon provides five grams soluble fiber.

- **Fruit Pectin**
  - Found in grocery stores as powder or liquid. Brand names include: Certo, Ball or Sure Jell.
  - Look for the unflavored variety as the taste may be unpleasant.

**Foods with soluble fiber include:**

- **Oatmeal**
- **Parsnips**
- **Oat bran**
- **Squash**
- **Corn meal**
- **Mushrooms**
- **Yams**
- **Apple sauce**
- **Sweet Potatoes**
- **Blackberries**
- **Artichokes**
- **Papayas**

**Insoluble fiber** moves stool through the intestine, stimulates regular bowel movements and prevents constipation. If your child is not on laxatives and wants to avoid constipation, provide foods with insoluble fiber. Most foods have both soluble and insoluble fiber.

Food high in insoluble fiber that have a laxative effect include:

- **Cauliflower**
- **Pinto beans**
- **Raw spinach/broccoli**
- **Lima beans**
- **Green cabbage**
- **Kidney beans**
- **Corn**
- **Strawberries**
- **Avocados**
- **Blueberries**
- **Carrots**
- **Cherries**
- **Cucumbers**
- **Dates**
- **Tomatoes**
- **Prunes and prune juice**
- **Green peas**
- **Raisins**
- **Potato with skin**
- **Apricots**
- **Green beans**
- **Pears**
- **Chick peas**
- **Guava**
- **Lentils**
- **Split peas**
- **Northern beans**

**When does toilet training begin?**

Toilet training usually begins between 2–3 years of age. Most children are toilet trained by age 3. Children with low defects need close monitoring to avoid constipation, and soiling or “accidents.” A laxative program and diet are often all that is needed to keep children clean. Children with high