

Management of Perineal Fistulas

Perineal fistula is the simplest of all anorectal defects. Patients are born with an anal opening located more anterior (or towards the front of the body) than normal. While this has been called an "anterior anus" or an "anteriorly mislocated anus," this is a misnomer because a perineal fistula is not a real anus, but an abnormal anal orifice. A real anus has a normal caliber orifice that has an anal canal and is surrounded by a voluntary sphincter, the ring-like band of muscles that opens and closes the anus.

Perineal fistulas (or fistulae) are abnormal orifices and most of the time they are stenotic (narrowed or constricted). They do not have a normal anal canal. A small amount of voluntary muscle can be found in both sides of the orifice, but there is no voluntary muscle in its anterior aspect and the bulk of the voluntary sphincter is located posterior to (behind) the anal orifice.

Corrective Operation

When patients with perineal fistulas are born at Cincinnati Children's Hospital Medical Center, but are otherwise normal (normal weight, no associated defects, and in good condition), we perform an operation called minimal [posterior sagittal anoplasty](#). (Sagittal refers to the plane of the body that divides it into left and right sections). The procedure is done during the first few days of the baby's life. Basically, the operation consists of making a posterior sagittal incision, dividing the sphincter mechanism, and then dissecting the rectum in a circumferential manner, enough as to be able to move the orifice back to be placed within the limits of the sphincter. The incision in a newborn is not longer than 1 or 1.5 cm.

During this operation, proper precautions must be taken to avoid damaging the urethra (the tube that carries urine from the bladder and in the male serves also as a passageway for semen), as well as nerves responsible for erection and urinary control, and in girls, the vagina.

In male patients, even when the perineal fistula is not severe, the rectum is still intimately attached to the posterior urethra. In female patients, it is very unlikely that the surgeon would injure the vagina because the rectum and vagina in this particular malformation are significantly separated.

Performed by an experienced surgeon, the operation takes approximately 45 minutes. Newborn babies do not need any kind of preoperative bowel preparation. Postoperatively, we administer antibiotics intravenously over 48 hours. Patients can go home soon but must be subjected to a

standard protocol of [anal dilatations](#) postoperatively. In a series of 112 patients we operated on, 100% developed bowel control.

Special Considerations Concerning Surgery

A baby that is born with a perineal fistula and happens to be premature, suffers from respiratory distress or any other kind of serious systemic problems, such as a severe congenital cardiac defect, should not have a minimal posterior sagittal anoplasty to correct the perineal fistula. This is particularly true if the baby is born in a place where there is not a pediatric surgeon with experience in the management of this defect. In these cases, simple anal dilatations or a small procedure called a "cut-back" can be done under local anesthesia in the newborn unit. Dilatations or "cut-back" procedures leave a cosmetically unpleasant perineum but do not damage the capacity to have bowel control. Therefore, these procedures are considered acceptable under special circumstances.

Many times, patients brought to us for consultation are not newborns, but are 1, 2, 3 or 5 years old. They usually suffer from severe constipation and often have a stricture of the anal orifice. While the treatment we offer the family is a minimal posterior sagittal anoplasty, the same as for a newborn, we are dealing with a patient that usually has fecal impaction and a greatly enlarged colon.

To operate on these patients, we admit the child to the hospital the day before surgery, insert a nasogastric tube, and administer a solution (Go-lytely) to completely clean the entire intestines.

The day of surgery, we insert a central line, perform the surgery, and give the patient nothing by mouth for 7 to 10 days. We give intravenous antibiotics for the first 48 hours.

This may seem like elaborate preoperative and postoperative management for such a small malformation; but if a patient is subjected to this kind of surgery and suffers from dehiscence (parting of the sutured sections of the wound) retractions and infection, the patient may then suffer from more serious consequences, such as [fecal incontinence](#) the inability to voluntarily control bowel movements. In our experience, following this protocol, we never had a complication with these patients.

Most Patients Develop Severe Constipation

It is rather exceptional to see a baby born with a perineal fistula that does not develop severe [constipation](#). This constipation is incurable, but manageable. When it is not properly managed, constipation becomes worse. Untreated constipation produces a greatly enlarged rectum (megarectum) and this leads to more constipation, creating a vicious circle. Adequate, aggressive management, on the other hand, prevents this from happening.

Other problems arise when the parents are not adequately advised about the importance of preventing and treating constipation. As a consequence, the baby develops the vicious circle of constipation, megacolon, and more constipation, and the child is referred for a consultation for "fecal incontinence."

A [contrast enema](#) (a procedure that uses air contrasted with another substance to better visualize the colon) shows a very severe megacolon and chronic fecal impaction. The patient actually suffers from a condition known as overflow pseudo-incontinence. At that stage, the treatment is more complicated and requires a procedure to remove the fecal impaction, followed by a protocol to determine the amount of laxatives that the patient requires to stay clean, and eventually a sigmoid resection to make the constipation problem more manageable.

In other cases, the problem of constipation is confronted, but then improperly treated. In desperation to try to alleviate the problem of constipation, surgeons may try different kinds of surgery to enlarge the anal opening. While an anal stricture or narrowing may exacerbate the problem of constipation, the problem itself is a consequence of a hypomotility disorder, a very slow movement of food through the colon and is directly related to the degree of dilatation of the colon.

Some patients who suffer from constipation after an operation for perineal fistula are misdiagnosed as having [Hirschsprung's disease](#), a defect in which the large intestine is lacking in certain nerve cells and unable to move stool through. As a result of rectal biopsies that show nerve cells lacking (even though this can occur in cases without Hirschsprung's disease), surgeons perform an abdomino-perineal resection, and leave the patient completely cured from constipation, but 100% fecally incontinent, or unable to voluntarily control bowel movements. It would be rather unusual for a patient that suffers only from constipation and fecal impaction, and has never had an episode of [enterocolitis](#) (inflammation of the large and small intestines) to suffer from Hirschsprung's disease.

Continuing Problems If Not Treated

Unfortunately, we still see many patients who were born with perineal fistulas and developed serious problems. The most common problems occur in patients in whom the defect was completely missed at birth. The failure to detect the presence of a tiny orifice in the perineum can lead to an erroneous diagnosis and the incorrect surgery.

Some patients who were born with a perineal fistula were never operated on because the anal orifice was not strictured or narrowed and the patients initially did well. By the time they are school-age children, they are fecally continent. The location of the anal orifice anterior to the center of the sphincter does not make these patients incontinent. The rectum actually is passing through the entire funnel-like sphincter mechanism and is only deviated anteriorly in the lowest part. Such a small mislocation is not enough to provoke fecal incontinence.

The indication for surgery for these patients should not be based on the idea of making these patients continent, because they are naturally continent. An operation is indicated if parents are not willing to accept a cosmetic abnormality in the child's perineum. Our patients have included girls who born with this malformation; never operated on, but when they reach puberty, felt uncomfortable about having a mild abnormality in the perineum. Some of them request an operation. For adult females who were born with this defect and never had it corrected, problems may arise when giving birth and the obstetrician discovers the defect and must observe special precautions to avoid a rectal injury.

Contact the Colorectal Center at Cincinnati Children's

For more information or to request an appointment for the Colorectal Center at Cincinnati Children's Hospital Medical Center, please [contact us](#).