Anorectal Malformations

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Current terminology in anorectal malformations (ARMs) is based on the location of the distal rectum and rectal fistula, which has both prognostic and therapeutic implications (Table 67.1). Inaccurate terms such as *high*, *intermediate*, and *low* are confusing and should no longer be used.

Over half of male patients with imperforate anus will have a fistulous connection between the rectum and the urinary tract (*rectobladderneck fistula*, *rectoprostatic urethral fistula*, or *rectobulbar urethral fistula*) and about one-third have a *rectoperineal fistula*, in which the rectal opening is on the perineal skin anterior to the anal dimple. Rarer malformations include *imperforate anus without fistula* and *rectal atresia*. *Imperforate anus without fistula* is more commonly found in patients with trisomy 21 and the rectum ends blindly in the pelvis, almost always at the level of the bulbar urethra. In patients with *rectal atresia*, there is a normal appearing anus with the rectum ending blindly at 2–3 cm. Many cases of rectal atresia are also associated with a presacral mass.

In females, there are three main types of malformations: *rectoperineal fistula, rectovestibular fistula*, and *cloaca*. A *perineal fistula* opens on the perineal skin, anterior to the sphincter complex and anal dimple, similar to males. In the case of a *vestibular fistula*, the opening lies within the introitus, but distal to the hymen (Fig. 67.1). A *cloaca* is a malformation in which the rectum, vagina, and urethra all open into a single channel, which subsequently opens onto the perineum usually just below the clitoris (Fig. 67.2). True *rectovaginal fistulas* are extremely rare.

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Prenatal Imaging

Parental prenatal counseling may be requested after fetal imaging suggests an ARM. The muscular components of the anal sphincter can be confidently visualized using ultrasound after approximately 23 weeks gestation. Ultrasound findings that increase suspicion of an ARM include dilation of the distal bowel, intraluminal calcified meconium or enterolithiasis. Presence of a cystic pelvic mass in a female fetus may suggest a cloacal malformation. Absence of the perianal muscular complex on 3D ultrasonography has also been shown to have a high sensitivity and specificity for identifying anorectal atresia. MRI is useful after 20 weeks gestation in fetuses suspected of having an ARM and can routinely delineate the musculature of the levator ani and external anal sphincter complex. Additionally, MRI can characterize abnormalities of the urinary tract, Mullerian abnormalities, limb anomalies, absent sacrum, and a presacral mass if present.

Diagnosis

The initial diagnosis of imperforate anus is almost always made during the first newborn physical examination, although a rectoperineal fistula is sometimes missed. Two important questions must be answered in the first 24 h of life: (1) should a colostomy be opened, deferring the repair of the defect until later in life, and (2) does the patient need urgent treatment for an associated defect (Figs. 67.3 and 67.4).

Males

The presence of a well-developed midline groove between the buttocks, a prominent anal dimple and meconium exiting through a small orifice located anterior to the sphincter in the midline of the perineum is evidence that the patient has a rectoperineal fistula. Occasionally one may see a prominent skin bridge over a tiny opening, giving the appearance of a

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Table 67.1 Classification of anorectal malformations and prognostic factors of bowel control

a. Anatomic classification of anorectal malformations	
Male	Female
Rectoperineal fistula	Rectoperineal fistula
Rectourethral fistula	Rectovestibular fistula
Bulbar	Imperforate anus without fistula
Prostatic	Cloaca
Rectobladder neck fistula	Rectal atresia
Imperforate anus without fistula	Rectal stenosis
Rectal atresia	Complex malformations
Rectal stenosis	
b. Prognosis for bowel control	
Good	Poor
Normal sacrum	Abnormal sacrum
Normal appearing bottom (good muscle)	Flat perineum (poor muscle)
Obvious anal dimple	*Types of anorectal malformations*
Types of anorectal malformations	Bladder neck fistula
Rectal atresia	Prostatic fistula
Perineal fistula	Cloaca with >3 cm common channel
Imperforate anus without fistula	Complex malformations
Bulbar fistula	
Vestibular fistula	
Cloaca with <3 cm common channel	

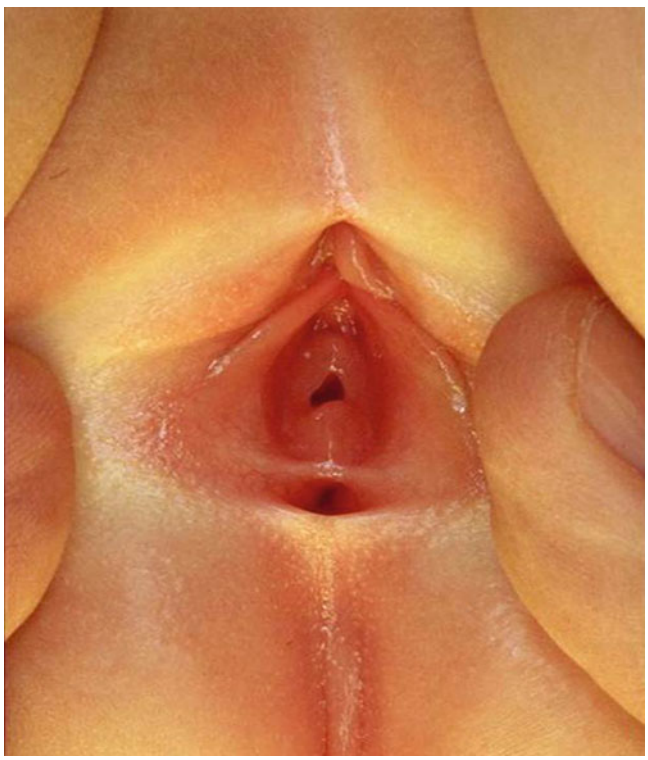

Fig. 67.1 Classic appearance of imperforate anus with rectovestibular fistula (supine position). Note that the fistula is located within the introitus but outside of the hymenal ring

bucket handle (Fig. 67.5), or a midline raphe, which can appear as a white or black ribbon of subepithelial meconium. These malformations can all be repaired via a perineal approach without a diverting colostomy. If a rectoperineal fistula is present, it may be technically advantageous to dilate the fistula to allow passage of stool until he is 3–4 months of age then perform a limited posterior sagittal anorectoplasty (PSARP). On the other hand, a flat bottom, with no evidence of a perineal opening and the presence of meconium in the urine are indications of a rectourethral fistula. A colostomy should be opened in these patients and the repair deferred.

If meconium is definitely seen on the perineum or in the urine, a conclusion as to the presence or absence of a fistula can be made with certainty. However, it may take some time for the intraluminal pressure to force the meconium past the pelvic musculature and out through a perineal or urinary fistula. This usually does not occur until after the first 24 h of life. A cross-table lateral radiograph of the abdomen and pelvis with the infant in prone position can be obtained to allow one to estimate the distance between the end of the dilated bowel and the skin. If this distance is <1 cm, a primary repair can be considered if the surgeon is experienced. If the distance is >1 cm, a colostomy should be performed.

Rarely, the anus may have a funnel-shaped appearance with a long skin-lined channel and a narrow opening (Fig. 67.6). This should prompt one to consider the diagnosis of Currarino syndrome with rectal atresia or stenosis and an associated hemisacrum and presacral mass.

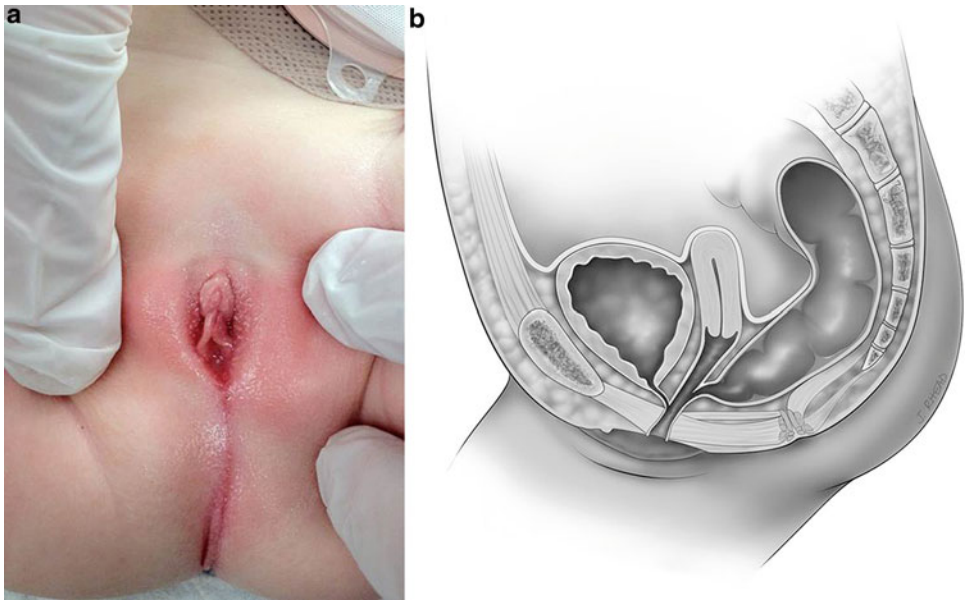

Fig. 67.2 Cloacal malformation. The urethra, vagina, and rectum meet to form a single common channel. (**a**) External appearance demonstrating underdeveloped labia and single orifice. (**b**) Illustration of short common channel

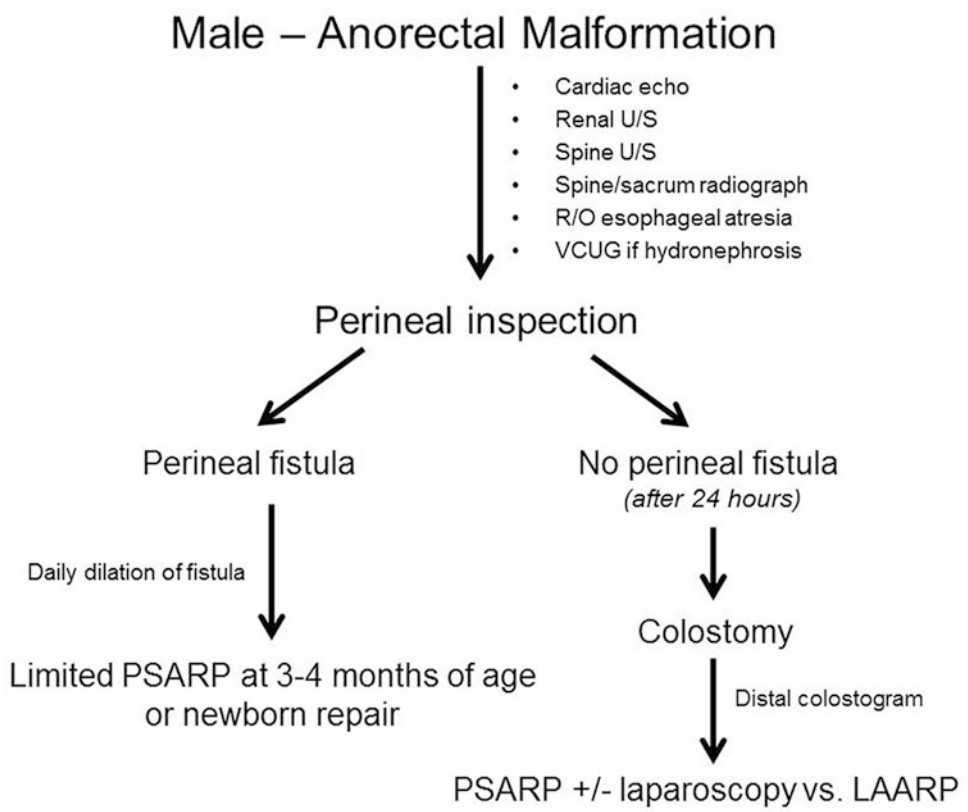

Fig. 67.3 Clinical algorithm for a male with anorectal malformation. *VCUG* voiding cystourethrogram, *PSARP* posterior sagittal anorectoplasty, *U/S* ultrasound, and *LAARP* laparoscopic assisted anorectoplasty

Females

The presence of a single perineal orifice in a newborn female establishes the diagnosis of a cloaca. All infants with cloaca require a colostomy and some also require a vaginostomy to drain a hydrocolpos.

In a female with a normal urethra, the presence of a rectal orifice located within the vestibule of the female genitalia but outside of the hymen confirms the diagnosis of a rectovestibular fistula. In these cases, some surgeons dilate the fistula to allow stool to pass and alleviate abdominal distention and defer the definitive repair until 3–4 months of age. Other surgeons prefer to open a colostomy and perform the repair at a later date. Surgeons who are experienced in the treatment of this abnormality may choose to do a primary repair in the newborn period with or without a protective colostomy.

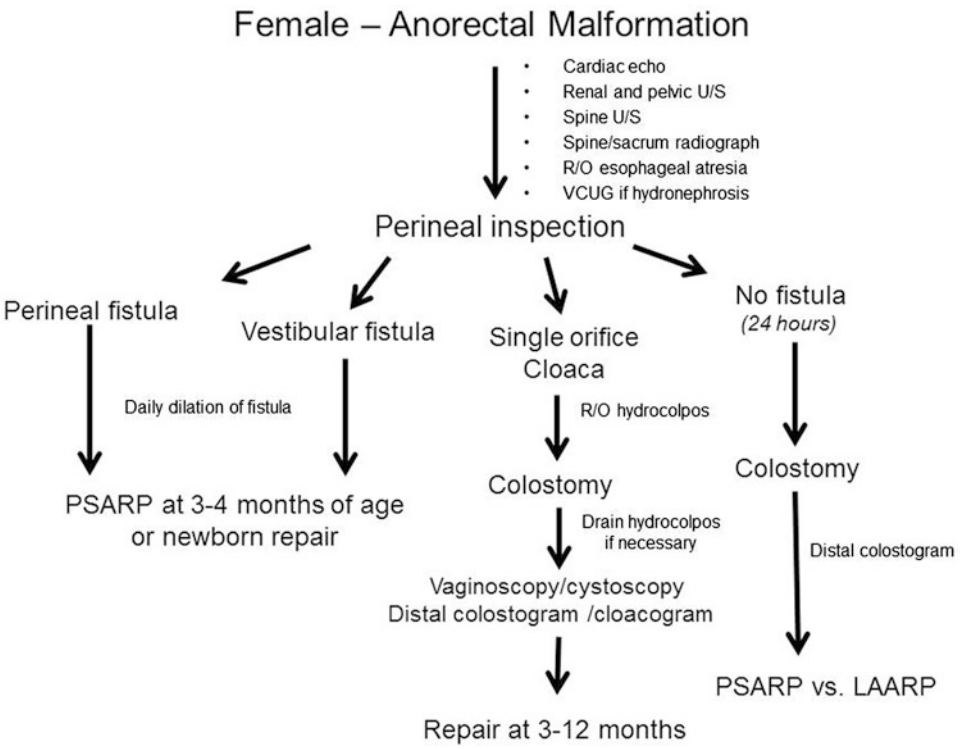

Fig. 67.4 Clinical algorithm for a female with anorectal malformation. *VCUG* voiding cystourethrogram, *PSARP* posterior sagittal anorectoplasty, *U/S* ultrasound, and *LAARP* laparoscopic assisted anorectoplasty

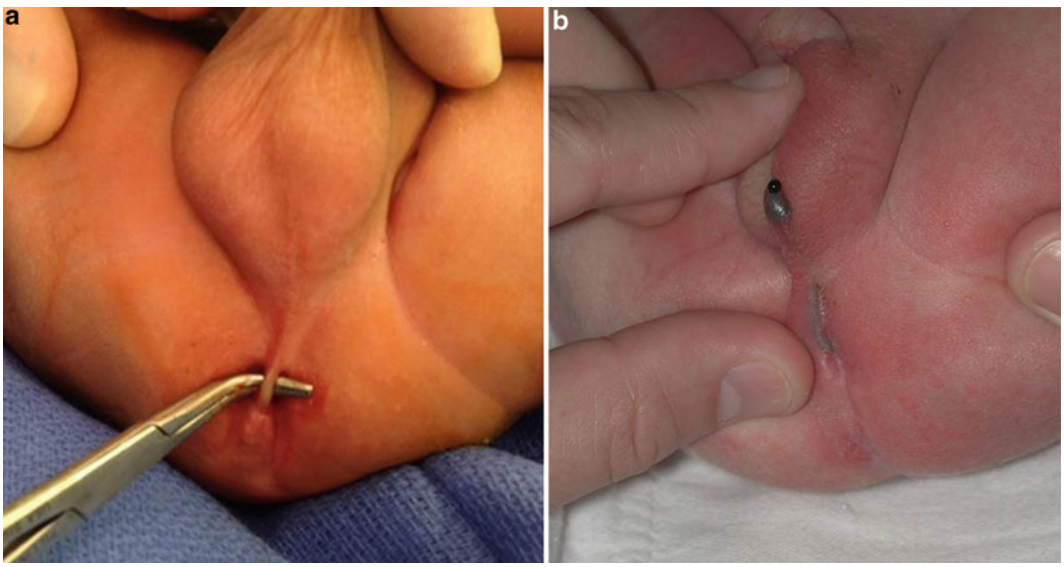

Fig. 67.5 Rectoperineal fistula in a boy. (**a**) Skin bridge referred to as a "bucket handle" deformity. (**b**) Subepithelial midline meconium

When the rectal orifice is located anterior to the center of the sphincter but posterior to the vestibule of the genitalia (in the perineal body), the diagnosis of rectoperineal fistula is established. These babies can undergo a primary anoplasty without a protective colostomy in the newborn period or after a period of dilating the fistula, similar to males. A common confusion is a female with a normal urethra and vagina and an anal opening that appears slightly anterior. If this opening is of normal caliber (Hagar 11 or 12 in the newborn), is within the sphincter, is supple (not fistulous tissue) and an adequate perineal body is present, then this is a normal variant and no surgery is indicated.

The absence of any of the above findings and the lack of meconium coming out through the genitalia after 24 h of life indicate that the patient most likely has imperforate anus without a fistula (more common in patients with trisomy 21).

Additional rare anomalies include: (1) *H-type rectovaginal* fistula where on inspection of the perineum, the patient may have a normal appearing anus (Fig. 67.7), (2) true *rectovaginal* fistula with the fistula located above the hymenal ring, a normal urethra and vaginal introitus, and no anal opening (Figs. 67.7), and *rectovestibular fistula with absent vagina*, in which there is a normal urethra, a rectal fistula in the vestibule, and no vaginal opening.

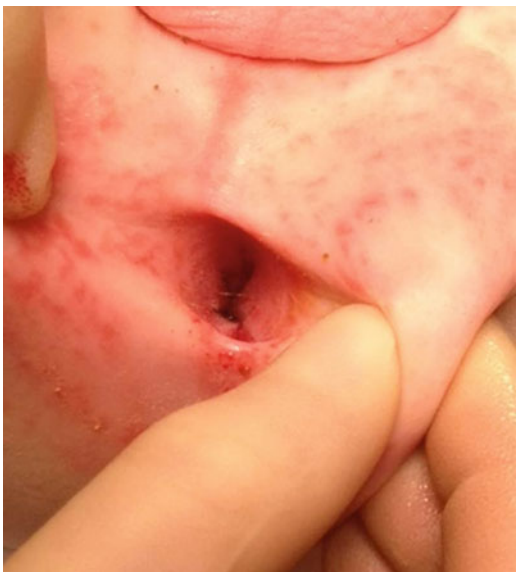

Fig. 67.6 Skin-lined funnel appearance of the anus in a patient with rectal stenosis and Currarino syndrome

Associated Anomalies

The waiting period of 16 to 24 h can be used to answer the second question concerning associated defects (Table 67.2) the majority of which are of the genitourinary system. In general, the more severe the anorectal anomaly, the more likely an associated defect will be present. However, even patients with the most benign malformation (rectoperineal fistula) should be completely evaluated, as up to one-third will have at least one associated malformation. In patients recognized to have a rectoperineal fistula outside the newborn period, a Cardiology consult to determine the need for an echocardiogram and Neurosurgery consult to determine the need for MRI of the spine is useful. The patient with a cloaca has a significant risk of an associated urologic defect as does the patient with a vestibular fistula, although the risk is less than cloacas.

The incidence of associated defects in males with imperforate anus varies based on the level of the defect, with a rectobladder neck fistula having the highest incidence and a rectoprostatic fistula or rectobulbar fistula being lower. All patients must have an ultrasound study of the kidneys to rule out hydronephrosis, the most common cause of which is vesicoureteral reflux. If hydronephrosis is identified, a voiding cystourethrogram should be obtained. In girls with a cloaca, the abdominal ultrasound must include the pelvis to rule out the presence of hydrocolpos or a distended bladder.

A nasogastric tube should be passed to rule out esophageal atresia and decompress the stomach while waiting for clinical evidence of a rectourethral or perineal fistula. All patients

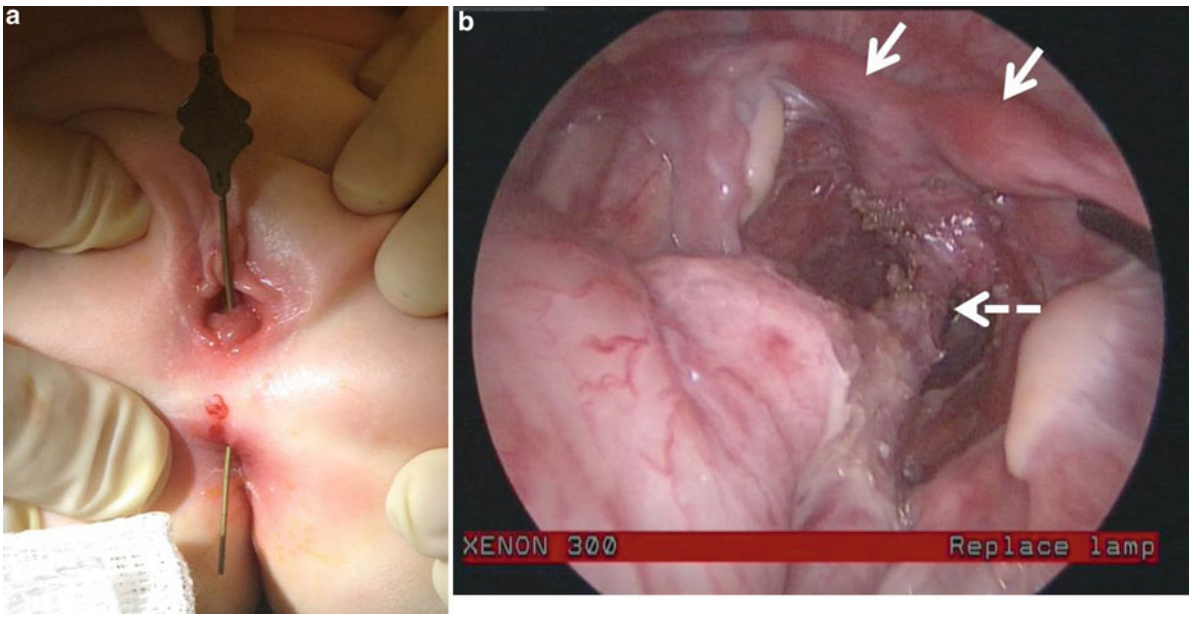

Fig. 67.7 Rectovaginal fistula. (**a**) H-type fistula. (**b**) Laparoscopic appearance of a rectal fistula entering into the posterior vagina. *Broken white arrow* showing rectovaginal fistula; *white arrows* identifying hemiuteri

Table 67.2 Associated anomalies

Cardiovascular	*Genitourinary*
Atrial septal defect	Vesicoureteral reflux
Patent ductus arteriosus	Renal agenesis and dysplasia
Tetralogy of Fallot	Cryptorchidism
Ventricular septal defect	Hypospadias
Transposition of great vessels	*Gastrointestinal*
Hypoplastic left heart	Tracheoesophageal fistula
Spine, sacrum, vertebrae	Duodenal atresia
Lumbosacral: hemivertebrae, scoliosis, hemisacrum, butterfly vertebrae	Malrotation
Tethered cord	*Gynecologic*
Spinal lipomas	Vaginal septum
Syringomyelia	Vaginal atresia
Myelomeningocele	Bicornuate uterus
	Uterine didelphys

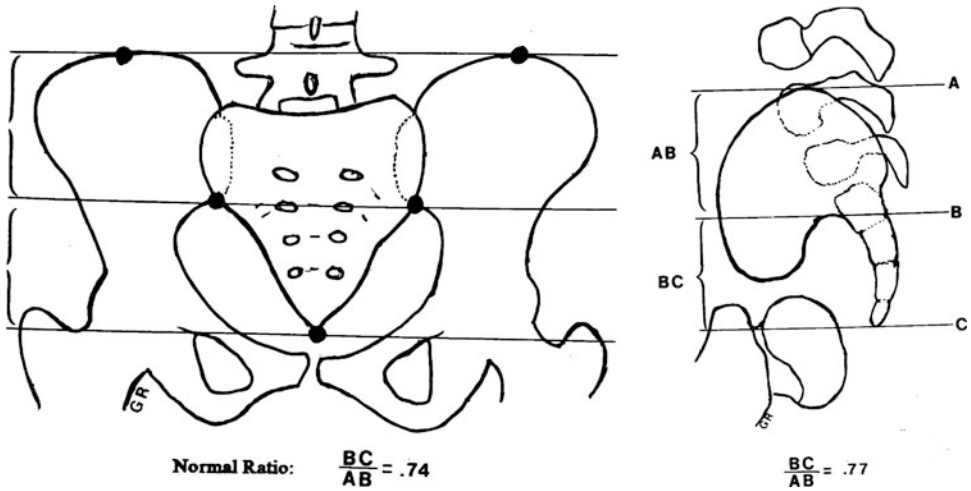

Fig. 67.8 Sacral ratio. *Line A* is drawn across the uppermost aspect of the iliac crests. *Line B* is drawn across the lowest point of sacro-iliac joints. *Line C* is drawn parallel to *line B* at the tip of the coccyx. The ratio is calculated by dividing the distance between *lines B* and *C* by the distance between *lines A* and *B*. (Pena A. Anorectal malformations. Semin Pediatr Surg. 1995;41(1):35–47. Reprinted with permission from Elsevier)

with imperforate anus should have an echocardiogram, as about 10 % of patients are found to have patent ductus arteriosus or a more serious structural cardiac defect such as tetralogy of Fallot or ventricular septal defect.

An ultrasound of the spine is also important to rule out the presence of a tethered cord, which occurs in about 25 % of all patients with ARMs. Although US remains the recommended screening modality to evaluate the spinal cord, recent studies have questioned its utility due to its poor sensitivity in detecting occult spinal dysraphism; it also can only be performed in the first 3 months of life, before sacral ossification. A radiograph of the spine must also be obtained. The presence of hemivertebra of the spine or sacrum indicates an increased risk of associated urologic defects and has negative prognostic implications in terms of bowel control. AP and lateral radiographs of the sacrum allow calculation of the *sacral ratio* (Fig. 67.8). Sacral ratios obtained during the newborn period may be misleading due to incomplete ossification. However, the accuracy of this ratio is improved at 5–6 months of age. A poorly formed sacrum (ratio <0.4) is associated with more severe malformations and carries with it a poor prognosis for bowel control. Evidence suggests that a lumbosacral MRI should be performed in patients with a sacral ratio <0.6 due to the tests ability to detect clinically significant spinal lesions. The presence of a hemisacrum might indicate the presence of a presacral mass, such as a teratoma, lipoma, or anterior meningocele, which can be confirmed with spinal ultrasound or pelvic MRI.

Initial Surgical Management

The ideal colostomy is created at the junction of the descending and sigmoid colon. Making the stoma more proximal ensures that the distal colon will be long enough to allow a tension-free pull-through later. In addition, stomas fashioned from the proximal sigmoid colon are less likely to prolapse because the colon is tethered by retroperitoneal attachments. To prevent prolapse of the more mobile distal stoma, the mucous fistula should be made with a very small external opening. The stomas should be far enough apart on the abdomen that the ostomy appliance can be placed comfortably over the functional stoma without covering the mucous fistula.

Loop colostomies have been discouraged in patients with imperforate anus because in theory they are incompletely diverting, which can allow overflow of stool into the distal

limb exposing the patient to fecal contamination of the genitourinary tract. While loop colostomies do have a higher rate of prolapse, the risk of urinary tract infection, development of megarectum, and need for stoma revision does not seem to be significantly different than in patients with a divided stoma provided the repair is performed early in life.

Under no circumstances should the distal stoma be closed as a Hartmann's pouch, as this will result in a mucocele and make it impossible to perform a contrast study of the distal rectum. A transverse colostomy is not recommended for several reasons: it makes it very difficult to clear inspissated meconium from the distal colon, it does not allow an adequate distal colostogram to be performed, and, in the presence of a fistula to the urinary tract, it can result in the resorption of urine, which can cause significant acidosis. At the time of the colostomy, use a soft rubber catheter to gently irrigate the distal limb with warm saline to remove all of the meconium from the lumen; this will prevent significant problems with inspissated meconium later on.

If the baby has *hydrocolpos*, it must be drained during the initial operation. Usually the hydrocolpos can be drained by tube vaginostomy. If there is bilateral hydrocolpos, two vaginostomy tubes can be placed or the vaginal dome can be opened and a window created between the hemivaginas. An undrained hydrocolpos will often cause hydronephrosis due to ureteral compression at the trigone. No treatment for the hydronephrosis should be considered until the hydrocolpos has been addressed. Once the vagina has been drained, the hydronephrosis should disappear. Even in the absence of hydronephrosis, it is important to decompress a hydrocolpos as the undrained vagina can become infected (pyocolpos), which can then lead to perforation or sepsis and may result in loss of the vagina.

Anoplasty

In infants with a perineal fistula, a formal anoplasty can be performed during the first several days of life without the need for a protective colostomy if the surgeon is experienced. The fistula can be dilated until the child is several months of age, at which time an anoplasty can be performed. This is technically easier in an older infant but requires a full preoperative bowel prep. The operation utilizes a minimal posterior sagittal incision from the fistula to the anal dimple. Multiple fine silk sutures are placed circumferentially around the rectal opening for uniform traction and to facilitate dissection. Circumferential full-thickness dissection of the rectum is performed until enough length is gained for the rectum to be placed accurately within the limits of the sphincter. In boys, the rectum and urethra are closely associated and special care is needed to avoid urethral injury. A Foley catheter should always be placed prior to anoplasty in a boy. In a girl, the posterior vaginal wall is adherent to the rectal fistula but there is a plane of separation. One should mobilize the rectum from the vagina to a point where the fistula can be placed within the limits of the sphincter without tension.

Almost all infants with a perineal fistula eventually have excellent bowel control although constipation is common and requires early management to achieve a good outcome. In addition to giving the child the best chance for normal bowel function, girls with perineal fistulas should be repaired to increase the size of the perineal body for future obstetric reasons.

Definitive Operative Repair

The PSARP remains the operation to which all technical modifications are compared. Laparoscopic assisted anorectoplasty has become increasingly popular for the management of ARMs. Proponents of this technique argue that functional outcomes will be better because a large posterior sagittal incision may be avoided and that the rectum is more accurately placed within the center of the sphincter muscle complex. However, studies examining long-term function after PSARP versus laparoscopic repair of ARMs have been inconclusive. We advocate the use of laparoscopy when it can replace laparotomy or an extensive posterior sagittal dissection. Thus it seems ideal for use in the treatment of rectobladder neck fistulas, in which the fistula is above the peritoneal reflection, and in repair of high rectoprostatic fistulas. In females, it might be useful to visualize the internal gynecologic anatomy and divide the fistula when the rectum inserts high on the posterior vagina in patients with or without a cloaca (a rare occurrence). Laparoscopy is not recommended for patients with rectobulbar fistula or no fistula as the fistula is well below the peritoneal reflection and shares a long common wall with the urethra.

Distal Colostogram

It is extremely important that the surgeon know the location of the distal rectum prior to the definitive repair. An augmented-pressure distal colostogram should be performed in all male patients who undergo a colostomy and all females with a cloaca. Information obtained from the study includes the location of the fistula between the rectum and the genitourinary tract, the length of available colon from the colostomy to the fistula site, the distance between the rectum and the anal dimple, and the relationship of the rectum to the sacrum. It may also demonstrate the characteristics of the vagina in females.

The study should be performed in the fluoroscopy suite using a balloon-tipped catheter to create a seal with the fascia

occluding the lumen of the colon. Hand controlled injection of contrast material is performed under pressure. It is begun with the patient in the supine position. Water-soluble contrast material should be used as barium is contraindicated in the presence of a recto-urinary tract fistula. A radiopaque marker is placed at the anal dimple. With the patient in the supine position, the surgeon can see the length of bowel available for the pull-through. The patient is then turned onto his or her side. As the injection is performed, contrast material will usually stop progressing at the pubo-coccygeal line. This line represents the upper limit of the levator muscles. It requires a significant increase in hydrostatic pressure for the contrast material to progress beyond this point.

In a male, the injection continues until the contrast material passes into the urethra. Usually the contrast goes up into the bladder rather than toward the penis and the injection should continue until the bladder is full and the baby starts to void. Films are taken during the entire sequence, particularly during voiding. The surgeon should be able to see the location of the fistula and its relationship to the bladder, bladder neck, and urethra (Fig. 67.9). If the study shows a bladder neck fistula, the surgeon knows that the rectum cannot be reached through a posterior sagittal incision and must be approached by laparoscopy or laparotomy. With a prostatic fistula on distal colostogram, the surgeon can anticipate locating the rectum in the soft tissue 1–2 cm deep to the coccyx during the posterior sagittal approach or just below the peritoneal reflection at laparoscopy. The presence of a bulbar urethral fistula indicates that the rectum will easily be found just deep to the levator muscles within 3–4 cm of the perineal skin. In the case of an imperforate anus with no fistula, the surgeon will not need to spend time looking for the fistula site and knows that the distal rectum is located adjacent to the bulbar urethra.

A distal colostogram is not necessary in patients who have a vestibular fistula. In patients with cloaca, one can complement the study by inserting a catheter into the perineal orifice and injecting contrast to delineate the bladder and vagina. A cystoscopy with catheter insertion into the bladder may also be done. Injection via a vesicostomy or vaginostomy, if present, delineates the anatomy. The goal of the study in patients with cloaca is to have images of all three crucial structures, bladder, vagina, and rectum, in AP and lateral projections and, if possible, in three dimensions. A three-dimensional cloacogram using a rotating c-arm in the fluoroscopy suite may provide additional useful information for operative planning.

Males

Patients with a rectobulbar or rectoprostatic fistula can be repaired using the posterior sagittal approach. It is mandatory that a Foley catheter be placed prior to commencing the operation. The patient is placed in the prone position with the pelvis elevated. The incision is made in the midline from above the coccyx to below the anal dimple. One must stay exactly in the midline, leaving an equal amount of muscle on each side of the incision. The posterior wall of the rectum is located and then opened in the midline. The incision in the rectal wall is continued distally up to the fistula site. The rectum is then separated from the urinary tract. A submucosal dissection is performed for about 3–5 mm above the fistula until a more obvious plane of separation is reached. This is facilitated by first delineating the lateral planes of the rectum. Once the rectum is fully mobilized, the urinary fistula is closed with long-term absorbable sutures.

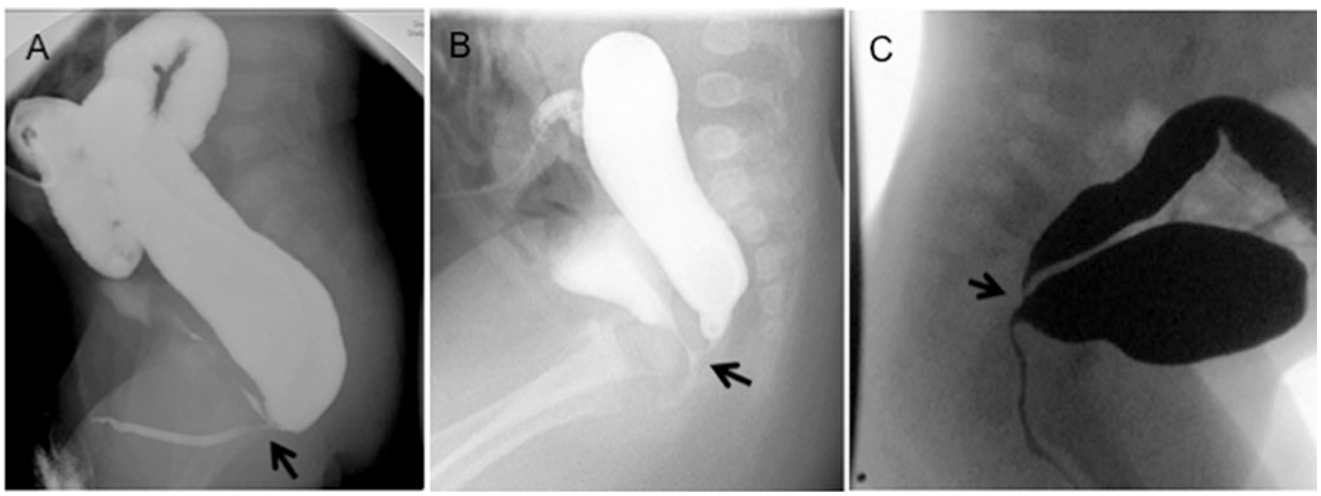

Fig. 67.9 Distal colostogram in the male with imperforate anus. (**a**) Rectobulbar urethral fistula. (**b**) Rectoprostatic fistula. (**c**) Rectobladder neck fistula. *Black arrow* indicates fistula location

The dissection should be performed as close to the rectal wall as possible while keeping the wall intact to preserve the intramural blood supply. The dissection continues until enough rectal length has been gained that an anastomosis between the rectum and the skin of the perineum can be created without tension. The electrical stimulator is used to determine the anterior and posterior limits of the sphincter mechanism. The anterior portion of the incision is closed, and the rectum is placed within the sphincter. The posterior edges of the levators are reapproximated behind the rectum. The posterior aspect of the muscle complex is also sutured together in the midline, incorporating the seromuscular layer of the posterior rectal wall in order to anchor the rectum. Finally, the anoplasty is performed by creating an anastomosis between the rectum and skin with interrupted long-term absorbable sutures. The rectum is occasionally very dilated, in which case the posterior wall should be excised and the rectum tapered in order to allow it to fit more easily within the sphincter mechanism.

Laparoscopic Assisted Anorectoplasty

Approximately 25 % of boys with an ARM have a rectoprostatic fistula and approximately 10 % have a bladder neck fistula. These patients are well suited for a laparoscopic approach to repair. A total body prep from the costal margins inferiorly is performed so that the entire lower half of the child may be included in the surgical field allowing access to both the perineum and abdomen. The operation begins with a laparoscopic approach, in which the distal rectum is dissected and the fistula is ligated. We typically use an absorbable endoloop for ligating the fistula. Further dissection is then performed to allow for the rectum to comfortably reach the perineum. The dissection is performed as close as possible to the rectal wall to avoid injury to nerves, reproductive structures, and the ureters. If getting the distal rectum to reach is difficult because the colostomy has been placed too distal in the sigmoid, it may be necessary to take down the mucous fistula. It is sometimes necessary to ligate one or more distal branches of the inferior mesenteric vessels in order to allow adequate mobilization of the rectum. The vascular arcades might have been disrupted when creating the colostomy, therefore one must be careful to ligate these distal inferior mesenteric artery branches close to the rectal wall, relying on its excellent intramural blood supply.

Once adequate length has been achieved, the rectum is ready to be pulled through. This can be done with the child in the lithotomy position after identifying the limits of the sphincter complex. The anorectoplasty may be performed using the technique of sequential placement of trocars or using a limited posterior sagittal incision. The latter technique allows for tacking of the rectum to the posterior edge of the muscle complex in order to prevent prolapse.

Females

All girls should undergo visual inspection of the introitus to identify a vaginal septum, which should be resected at the time of repair and to be certain there is not a distal vaginal atresia. Vestibular fistulae are repaired using a posterior sagittal incision with the patient in prone position. These defects may be repaired in a single stage using a preoperative bowel prep and a period of NPO and antibiotics postoperatively. Multiple silk sutures are placed circumferentially in the external orifice of the fistula, as uniform traction on the rectum facilitates the creation of a dissection plane. The posterior rectal wall is identified and the dissection continued between the rectal fascia and the rectal wall, staying as close as possible to the rectal wall.

The dissection progresses laterally on both sides until a point is reached where the distal rectum and fistula must be separated from the vagina. These two structures have a very thin common wall with no true plane of separation. A submucosal plane on the rectal side must be developed for the first 3–5 mm or so, until eventually a full-thickness plane can be developed. Circumferential dissection of the rectum is performed until adequate length is achieved for the anastomosis to be performed under mild tension preventing rectal prolapse. If defects were made in the posterior vagina or anterior rectum during the dissection, the rectum is mobilized further until it can be positioned such that the two suture lines are not adjacent to each other in order to avoid a rectovaginal fistula. The perineal body is reconstructed using interrupted long-term absorbable sutures, bringing together the anterior limits of the sphincter, and the rectum is placed within the limits of the sphincter. The remainder of the operation is as previously described.

Rectovaginal fistulas are uncommon. Repair may be performed using a posterior sagittal approach or laparoscopically if the fistula connects to the proximal vagina. In patients with an H-type rectovaginal or rectovestibular fistula where the anus is normally positioned within the sphincter and a fistula is present between the vestibule, vagina, or labia and the rectum at the level of the dentate line, a transanal Swenson-like repair similar to Hirschsprung disease may be performed.

Posterior Sagittal Anorectovaginourethroplasty (PSARVUP) for the Repair of Cloaca

Cloacal malformations represent a wide spectrum of defects. A major determinate of the technical approach to repair and prognosis is the length of the common channel. Cloacas with a common channel <3 cm may be repaired by general pediatric surgeons who have had adequate training in this procedure with acceptable outcomes expected. In defects with a

common channel >3 cm in length or more complex defects, the repair is often challenging and better outcomes may be achieved when the repair is performed by an experienced surgeon. In these more complex patients, urinary and fecal continence is often poor and requires artificial methods of keeping patients dry (intermittent catheterization) and clean (bowel management program).

Vaginoscopy and cystoscopy are performed as a separate procedure *after* the newborn period. This will provide useful information for surgical planning and prognosis. This should not be performed at the time of colostomy as visualization is difficult and it will often result in significant distension of the bladder or vagina, making the colostomy more difficult. With the scope positioned at the confluence of the urethra and vagina, the length of the common channel, also known as the *urogenital sinus*, is measured from the tip of the endoscope to the perineal skin. Also, the distance from the urethral take off and the bladder neck is determined. When the common channel is <3 cm, it should be possible to repair the entire malformation from a posterior sagittal approach. If the common channel is >3 cm, it is better to prepare the patient with a total body prep in order to be able to turn the patient and open the abdomen if necessary.

There are several key steps in repairing a cloaca with a common channel <3 cm. The posterior sagittal incision is continued anteriorly until the surgeon reaches the single perineal orifice and the common channel is opened in the midline. Total urogenital mobilization can be done in the majority of cases and significantly reduces the future risk of a urethro-vaginal fistula, vaginal stricture, or acquired vaginal atresia. The rectum needs to be dissected off the posterior aspect of the vagina. If a vaginal septum is present, the rectal fistula is usually located within the proximal aspect of the septum. The vagina and urethra are then mobilized as a single unit. The common channel is then split in the midline and secured to the skin becoming the edges of the labia. The urethral orifice is sutured approximately 5 mm behind the clitoris and the vaginal edges are sutured to the introitus. If the patient has two hemivaginas, the vaginal septum should be excised. The perineal body is reconstructed and the rectal component of the malformation is repaired as in a typical PSARP.

When dealing with a longer common channel, total urogenital mobilization may still be utilized but additional length is frequently needed requiring a laparotomy. If the length is still not adequate, then the urinary tract and vagina must be separated. Alternatively, the decision to separate the vagina from the common channel is made at the beginning of the case. Creation of a neovagina with colon or small bowel, or a vaginal switch maneuver should be in the armamentarium of a surgeon repairing these defects if the vagina does not easily reach the perineum.

If a laparotomy is needed during the main repair or at the time of colostomy closure, the Müllerian structures should be inspected and the presence of normal structures and patent fallopian tubes documented. In patients with the more complex malformations described above, a suprapubic cystostomy tube or vesicostomy is needed until intermittent catheterization can be started. The reconstructed urethra may need long-term stenting with a Foley catheter or circle stent.

Postoperative Care

Recovery after a PSARP is usually rapid and generally straightforward. The children seem to have surprisingly little pain. Patients with a colostomy can eat the same day of surgery. When a single-stage operation is performed in an infant or child who is passing stool, we withhold oral nutrition for 5–7 days after surgery, during which time they are maintained on parenteral nutrition. Broad-spectrum intravenous antibiotics are continued for 24 h. In boys who have had a rectourethral fistula repaired, the Foley catheter is left in place for 7 days. The catheter is removed and the parents are instructed to return if the child is unable to void within 6 h. If the Foley catheter is accidentally removed before the recommended time, it need not be replaced since the majority of patients will be able to void and the potential for urethral injury during recatheterization is significant.

Two weeks after surgery, the anus is calibrated in the clinic with Hegar cervical dilators and the parents are then taught how to dilate the neoanus. The dilator is lubricated and then passed through the anus into the rectum, held in place for 30 s, removed, and then passed again. The anus must be dilated twice per day. Every week the size is increased until the appropriate size for the patient's age is reached (Table 67.3). Once the desired size is reached, the colostomy can be closed and the frequency of dilations tapered.

After the colostomy is closed, patients often initially have very frequent bowel movements that can produce a severe perianal rash. It may take some time for this to heal. A variety of creams and ointments are available that attempt to create a barrier, usually with variable success. The best treatment for the severe diaper rash is to avoid prolonged contact between stool and the skin. We instruct parents to wash the perineum with mild soap and water every time stool appears. Soon the number of bowel movements will decrease and the patients typically develop constipation. Parents should be forewarned of this change so that they are ready to treat it. At this point, laxatives often need to be added. The family should then work toward a good bowel movement pattern, meaning 1–3 well formed stools per day. This is achieved with the right combination of diet, laxatives, and water-soluble fiber. This is the best scenario leading up to the time for the child to attempt toilet training.

Table 67.3 Anal dilation and tapering schedule following anorectoplasty

Dilation		Tapering	
Age of child	Dilator size	Frequency	Duration (months)
1–4 months	#12	Daily	1
4–8 months	#13	Every third day	1
8–12 months	#14	Twice per week	1
1–3 years	#15	Once per week	1
3–12 years	#16	Once per month	3
>12 years	#17		

Bowel Management

Constipation is the most common problem seen in patients with ARMs. It is more severe in less complex malformations. The more complex malformations have a poorer prognosis in terms of bowel control, but a lower incidence of constipation. It is extremely important that constipation be treated aggressively. Complications from prolonged constipation include soiling, overflow pseudoincontinence, and megarectum. Megarectum, in turn, provokes more constipation, which worsens the dilation, creating a vicious cycle. In order to avoid this, the child should empty the rectum every day, which is best achieved with a high-fiber diet and laxatives as needed. When properly treated, the majority of patients with ARMs will have voluntary bowel movements by the age of 3 years. However, about half still soil their underwear intermittently. This is usually due to constipation. If the soiling does not improve by the age of toilet training, a bowel management and enema program should be initiated. Parents should be informed that children born with ARMs will typically toilet train about a year later than normal but that the goal is to have the child clean and in normal underwear by the time they enter kindergarten.

Patients with ARMs may still suffer from fecal incontinence despite an adequate anatomic repair. However, these patients should be able to remain clean and completely free of accidents without having to wear diapers. An aggressive bowel management program that includes the use of enemas allows them to achieve this goal. Fecally incontinent patients may also have a tendency toward constipation. These patients typically still have their entire colon but have a megarectum. Most of these patients can be successfully managed with a large volume (500–1000 mL) saline enema with additives such as glycerin.

Once it is demonstrated that the bowel management has been successful, patients with severe constipation can be offered a Malone procedure, also known as a continent appendicostomy or antegrade continent enema (ACE) procedure (Fig. 67.10). An appendicostomy is created, in the umbilicus or right lower quadrant. This allows the enemas to be given in an antegrade fashion. The cecum is plicated around the base of the appendix to create a valve that prevents stool from leaking back through the umbilicus. Another option for antegrade enemas is a cecostomy tube. Procedures to create conduits for antegrade enemas are usually recommended when the patient wants to become more independent, as it allows the administration of the enema without parental assistance. It may also be indicated if the patient is becoming resistant to retrograde enemas. The Malone procedure is essentially just another way to administer an enema; therefore, it should in general only be performed in patients for whom bowel management has been successful. The urologic status of the patient should be known prior to surgery, as a Mitrofanoff, if needed, could be performed in conjunction with the Malone and the appendix potentially split for the two procedures. Also, any urologic reconstruction could be done at this time.

Outcomes and Quality of Life

In patients with perineal fistula, approximately 90 % experience fecal continence although around 20 % require laxative treatment for constipation (Table 67.4). Approximately 70 % of patients with vestibular and 50 % of patients with rectobulbar urethral fistulas are totally continent of stool. However, approximately 60 % of vestibular and rectobulbar urethral patients suffer from constipation.

Fecal incontinence has been shown to have significant negative impact on QoL in terms of social and emotional functioning, body image, and physical symptoms in children and adolescents as well as adults. However, QoL is significantly improved in about half of patients who follow a bowel management program. Patients with associated congenital anomalies and those with more severe ARMs have significantly poorer QoL.

With the exception of an occasional patient with myelomeningocele, a severe sacral abnormality or tethered cord, boys with imperforate anus usually have good urinary control. In the absence of a predisposing condition, we believe that most cases of urinary incontinence in male patients are due to injury during the peri-bladderneck dissection. This is also true of girls, with the exception of those with cloaca. Although the majority of patients with a common channel

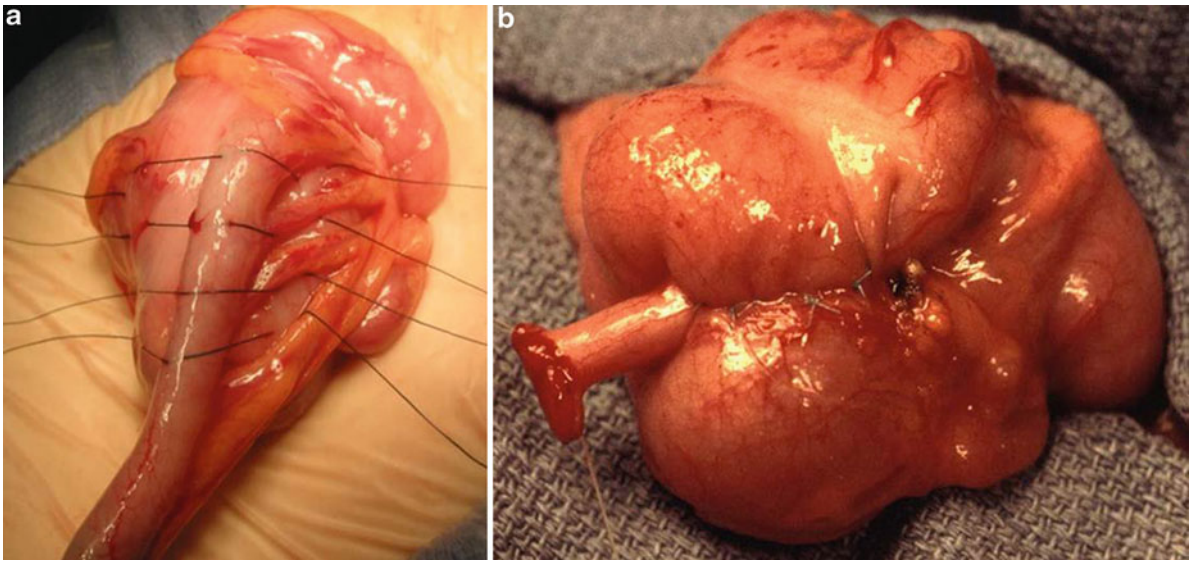

Fig. 67.10 Malone procedure. The cecum is plicated around the base of the appendix to create a valve, which prevents escape of gas or liquid through the appendix. (**a**) Non-absorbable suture placed in seromuscular layer of cecum and appendix. (**b**) Completed cecal plication

Table 67.4 Functional outcomes

	Voluntary bowel movement (%)	Totally continent[a] (%)	Constipated (%)	Soiling (%)
Perineal fistula	97	83	58	16
Rectal atresia or stenosis	100	56	70	40
Vestibular fistula	90	64	55	36
Imperforate anus without fistula	78	50	49	51
Bulbar fistula	79	46	59	49
Prostatic fistula	65	18	42	78
Cloaca: <3 cm common channel	66	35	39	63
Cloaca: >3 cm common channel	35	12	28	84
Vaginal fistula	60	20	20	80
Bladder neck fistula	20	7	14	90

Source: Data from Coran AG, et al, eds. Pediatric Surgery, 7th ed., Levitt MA, Pena A. Chapter 103: Anorectal Malformations, p. 1309, Copyright (2012)

[a]Voluntary bowel movements and no soiling

<3 cm have normal urinary control, approximately 20 % may require intermittent catheterization for a flaccid bladder. With a common channel >3 cm, approximately 80 % will have difficulty with urinary continence requiring intermittent catheterization or some form of continent diversion. An ARM index, comprised of the (1) type of malformation, (2) quality of the sacrum, and (3) quality of the spine, takes into account anatomical aspects predictive of continence. These three things should be known for all ARM patients.

Complications

Posterior urethral diverticulum (PUD) is a common complication that may occur following repair of a rectourethral fistula. It has been speculated that this may be more common following laparoscopic assisted anorectoplasty (LAARP) but is also a known complication following PSARP. In patients with a rectoprostatic fistula who had an LAARP, MRI may identify PUD in a third of patients. In the majority of these patients the diverticulum is asymptomatic and requires no intervention. A technique described to reduce the risk of a PUD during LAARP is sharp division of the fistula flush with the urethra without the use of clips or ties. We make sure to dissect the distal rectum low enough so that a three-millimeter grasper fits across at the level of the fistula. Also, by only using laparoscopy for bladder neck and prostatic fistulas and not for lower fistulas where the rectum is below the peritoneal reflection with a long common wall, this complication can be avoided. A PUD should be suspected in patients who report daytime dribbling, recurrent UTI, or passage of mucus through the urethra. If these symptoms are present, an MRI performed with a

20–24 Fr Foley placed in the rectum without inflating the balloon is accurate in detecting a PUD. Whether or not a PUD may be observed in an asymptomatic patient remains an area of debate. However, we recommend surgical excision of the diverticulum as malignant degeneration into a mucinous adenocarcinoma has been reported.

Colostomy complications include prolapse, stricture, retraction, and peristomal wound infections and are reported in 15–30 % of patients with divided colostomies and 25–30 % in those with loop colostomies. Although a divided colostomy has been traditionally recommended in patients with ARMs to avoid these complications and the risk of UTI, this has not been clearly demonstrated in the literature. One should follow the principles of creating the colostomy close to the retroperitoneal attachments of the proximal sigmoid colon to reduce the risk of prolapse which is the most common stoma-related complication and to leave the mucous fistula as long as possible for the future pull-through.

Wound infections are often superficial, limited, and healed by secondary intention. *Anal strictures* may result from ischemia or excessive tension of the pull-through segment or noncompliance with the dilation schedule. Options for management of anal strictures include resuming dilations, redo anoplasty in which the rectum is mobilized circumferentially to bring down a healthy segment of bowel, or stricturoplasty using a V-Y or skin flap advancement technique.

The incidence of *rectal prolapse* ranges from 6 to 30 % following LAARP and approximately 4 % following PSARP. Low gluteal and sphincter muscle quality, vertebral anomalies, tethered cord, and LAARP are risk factors for developing rectal prolapse. Rectal prolapse may be mild and asymptomatic or may be significant enough to warrant redo anoplasty. It can interfere with a bowel control and can also cause bleeding and mucous production.

Future Considerations

The management of patients with ARMs is complex and requires an understanding of the multiple problems that these children can have. Although it is extremely important for the surgeon to be able perform the operations without causing injury, technical mastery alone is clearly not sufficient. A multidisciplinary team approach including the pediatric surgeon, pediatric urologist, gynecologist, neurosurgeon, gastroenterologist, radiologist, and others when associated anomalies are identified is critical to achieve optimal outcomes. As these patients become older, bowel management and urologic and gynecologic function become critical aspects to their care. If these patients are not followed closely, and bowel, renal, and bladder function issues are not addressed early, adverse health related issues will occur. Accurate long-term assessment of outcomes regarding overall function and quality of life as well as burden of treatment is lacking and represents an area for improvement in the care of patients with ARMs. It is essential that the surgeon be able to manage the significant postoperative functional sequelae that many of these patients have and recognize that these are patients for life.

Editor's Comment

Imperforate anus is one of the more common congenital anomalies requiring surgical correction and pediatric surgeons should be well versed in the correction of its more common varieties. One of these is the perineal fistula in girls, commonly referred to as the "anterior displaced anus." Some controversy exists over the best way to correct these seemingly benign lesions. Regardless of the technique used, the functional result is the same: moderate to severe constipation. In the past, the fistula was often simply dilated, which allows meconium to pass; however, patients were often incontinent and needed extraneous methods to evacuate. Similarly, for a time it was popular to use the "cutback anoplasty," in which the fistula is opened in the midline posteriorly to the center of the anal sphincter and the rectal mucosa is anastomosed to the perianal skin. The result is a keyhole-shaped anal opening that is only partially surrounded by sphincter musculature. A better anatomic and functional result is achieved when the rectum is brought up through the anal sphincter with either a "mini-PSARP" procedure or anal transposition.

Girls with cloaca are treated in many children's centers by a team of surgeons that includes a pediatric surgeon, a pediatric urologist, and sometimes an expert in pediatric gynecologic anomalies. Pediatric urologic consultation can also be useful when searching for a high rectum in boys, who are at risk for injury to the urethra, bladder neck, seminal vesicles, and vasa deferentia during the very difficult and tedious dissection that is often required in these cases. One can also be lulled into a false sense of security by the rare male infant with an apparent perineal fistula, classically thought to be associated with a low lesion and straightforward repair, but who instead has a rectum that ends at the level of the prostate and a long, narrow fistula that opens on the perineum. These infants often need to have a laparotomy for proper mobilization of the rectum.

In boys with no apparent fistula, it might be difficult to see evidence of urethral passage of meconium as it is diluted by urine and soaks into the diaper. It is useful in these cases to place a cotton gauze pad over the penis within the diaper. The urine passes through the gauze while the meconium is trapped in the interstices of the cotton mesh.

Finally, although it is certainly better to make a decision regarding repair or colostomy within 48 h of birth, there are rare situations in which an infant with a severe cardiac anomaly and no apparent fistula is felt to be too unstable even for a bedside colostomy or is thought to be unlikely to survive

for long with or without cardiac reconstruction. With adequate gastric decompression, these children can sometimes be maintained for a surprising length of time and, in very rare cases, will begin to pass meconium through the perineal dimple a few weeks after birth, at which point they are often amenable to dilation.

Suggested Reading

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