Outcomes from the correction of anorectal malformations

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Purpose of review

Anorectal malformations have been recognized and managed since antiquity, with surgical treatment evolving to maximize anatomic reconstruction, avoid complications, and understand mechanisms of incontinence, ultimately leading to improved quality of life for patients. This review describes recent advances in the management of anorectal malformations, including prenatal diagnosis, newborn treatment, surgical correction, and postoperative care.

Recent findings

Surgical treatment has improved with better understanding and exposure of anatomy and appreciation of the intimate relation between rectum and urinary tract. Repair of cloacal malformations has evolved to include the total urogenital mobilization and an appreciation of the complex associated Mullerian anomalies. The importance of associated urologic, gynecologic, neurologic, and orthopedic malformations has been recognized. Addition of a bowel management program to patients' postoperative care has increased dramatically the number of children who are clean and dry.

Summary

Management of anorectal malformations requires an accurate clinical diagnosis, proper newborn treatment, meticulous anatomic reconstruction, and comprehensive postoperative care with the goal of having a child who is clean and dry, with an excellent quality of life, because they either have the capacity for continence or can be kept artificially clean with a comprehensive bowel management program.

Keywords

anorectal malformation, bowel management, cloaca, cloacal malformation, imperforate anus

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Abbreviation

PSARP posterior sagittal anorectoplasty

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Introduction

Imperforate anus has been treated since antiquity by creating an orifice in the perineum and has evolved from the first anoplasty by Amussat, in 1835, to an abdominal perineal pull-through, which pulled the bowel close to the sacrum through the 'puborectalis sling' [1]. Then the posterosagittal approach, which was performed first in 1980 [2,3], for the first time allowed direct exposure of this complex anatomic area and demonstrated that the sphincter mechanism was a funnel-shaped confluence of muscles. The approach enabled correlation of the external appearance of the perineum with the operative findings and, subsequently, with the clinical results and clarified a terminology and classification.

Classification

Imperforate anus occurs in one of every 4000–5000 newborns (Table 1) most frequently with a rectourethral fistula in boys and a rectovestibular fistula in girls [4]. Persistent cloaca, when the urinary tract, vagina, and rectum form a common channel, was traditionally considered an unusual defect whereas rectovaginal fistula was common. In retrospect, many of these were in fact cloacas, and the unfortunate consequence of this misdiagnosis was surgical repair of the rectal component only, leaving the patient with an unrepaired urogenital sinus [5].

Recto-bladder neck fistula is the only real supralevator malformation and occurs in 10% of male patients [4]. It is the only defect in boys that requires an abdominal approach (either laparotomy or laparoscopy), in addition to the perineal incision because the rectum is unreachable from below. Anorectal malformations represent a wide spectrum. The former 'low,' 'intermediate,' and 'high' malformations are arbitrary designations without therapeutic or prognostic meaning. A more therapeutic and prognostically oriented classification is depicted in Table 1.

Male defects

Rectoperineal fistula, determined by perineal inspection, is what traditionally was known as a 'low defect.' The rectum is located within most of the sphincter mechanism, but the lowest part is anteriorly mislocated and narrow. The terms 'covered anus,' 'anal membrane,' and 'anteriorly-mislocated anus' refer to this anorectal defect. The most frequent defect in male patients is imperforate anus with rectourethral fistula. The fistula is at the lower (bulbar) or upper (prostatic) urethra. Immediately above the fistula site, the rectum and urethra share a common wall, an

Table 1. Classification of anorectal malformations

Boys
Cutaneous (perineal fistula)
Rectourethral fistula
Bulbar
Prostatic
Rector – bladderneck fistula
Imperforate anus without fistula
Rectal atresia
Girls
Cutaneous (perineal fistula)
Vestibular fistula
Imperforate anus without fistula
Rectal atresia
Cloaca
Complex malformation

important anatomic fact with significant technical implications.

The rectum is usually distended and surrounded laterally and posteriorly by the levator muscle. Between the rectum and the perineal skin are striated voluntary muscles called the muscle complex. At the level of the skin, a group of voluntary muscle fibers, parasagittal fibers, are located on both sides of the midline. Rectobulbar fistulas are usually associated with good-quality muscles, a well-developed sacrum, a prominent midline groove, and a prominent anal dimple. Rectoprostatic fistulas are more frequently associated with poor-quality muscles, an abnormally developed sacrum, and a flat perineum, with a poor midline groove, and a hardly visible anal dimple. Patients with a recto – bladderneck fistula have a poor prognosis because the levator muscle, muscle complex, external sphincter, and sacrum are poorly developed. Patients with imperforate anus without fistula, a common defect in Down syndrome, usually have a well-developed sacrum and good muscles with the rectum ending approximately 2 cm from the perineal skin.

Female defects

Girls with rectoperineal fistula have similar anatomy and prognosis to boys with this defect. Rectum and vagina are well separated. The reconstruction places the rectum fully within the sphincter mechanism and reconstructs a perineal body. It is not uncommon for these conditions to escape detection in the newborn period and to be diagnosed several months later when the child has severe constipation. The most common defect in girls is a rectovestibular fistula, which can also be misdiagnosed as 'rectovaginal fistula.' This defect has an excellent functional prognosis and is diagnosed with a careful inspection of the newborn genitalia. The clinician will observe a normal urethral meatus, a normal vagina, and a third hole in the vestibule, which is the rectovestibular fistula. About 10% of these patients have two hemivaginas.

A number of pediatric surgeons repair this defect without a protective colostomy. This is a well-recognized trend in the management of anorectal malformations [6,7] that avoids the potential morbidity of a colostomy and reduces the number of operations to one from three (colostomy, main repair, and colostomy closure). A perineal infection can provoke severe fibrosis, however, which may interfere with the sphincteric mechanism [4]. Secondary operations do not render the same good prognosis as a successful primary operation [4]. The decision to open a colostomy or to repair the defect primarily must take into consideration the surgeon's experience and the patient's clinical condition. Imperforate anus without fistula in girls has the same therapeutic and prognostic implications as described for male patients.

Persistent cloaca represents the extreme in complexity of female malformations in which rectum, vagina, and urinary tract form a confluence. The diagnosis is a clinical one and is based on finding a single perineal orifice. The length of the common channel varies from 1–7 cm, which has technical and prognostic implications. Common channels longer than 3 cm are usually associated with complex defects. In these patients some form of vaginal replacement is often used during the definitive repair. A common channel of less than 3 cm usually means that the defect can be repaired with a posterior sagittal incision alone, without opening the abdomen [8••].

Frequently, the vagina is abnormally distended and full of secretions (hydrocolpos) that can compress the trigone and interfere with the drainage of the ureters, leading to megaureters. The dilated vagina can also become infected, which is called 'pyocolpos' and may lead to perforation and peritonitis [9].

A frequent finding in cloacal malformations is the presence of different degrees of vaginal and uterine septation, duplication, or atresia. During puberty, a patient's menstrual blood cannot drain through the vagina and may accumulate in the peritoneal cavity. These malformations have important obstetric implications [10].

Associated defects

Sacral deformity is a frequently associated defect with one or several sacral vertebrae missing. A hemisacrum is usually associated with a presacral mass and poor bowel control. Other vertebral abnormalities such as spinal hemivertebra have a negative implication for bowel control. Children with anorectal malformation have different degrees of sacral hypodevelopment, which can be objectively quantified by calculating a sacral ratio [4].

Emphasis has been placed on the diagnosis and treatment of tethered cord, which is a defect frequently associated with anorectal malformations [11,12]. It has been assumed that the presence of tethered cord is associated with poor functional prognosis in these children with

regard to fecal and urinary continence. The presence of tethered cord by itself coincides with the presence of a very high defect, a very abnormal sacrum, or spina bifida. Therefore, it is difficult to know whether the tethered cord itself is responsible for the bad prognosis. It is also unclear whether releasing the tethered cord changes the functional prognosis for the patient.

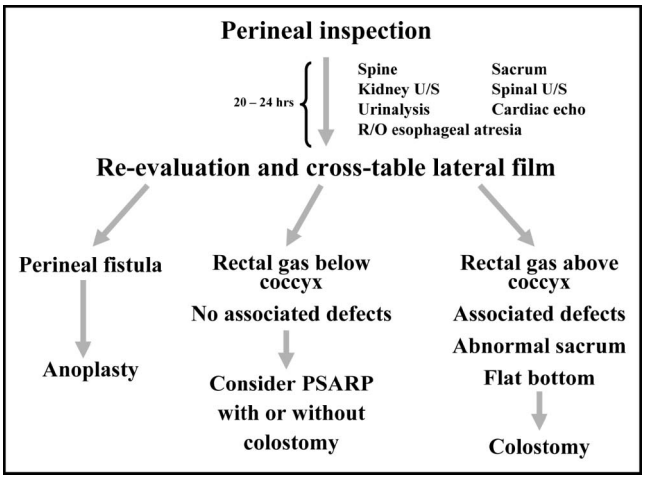
Associated urologic defects are common [13–16], particularly with higher malformations. Hydronephrosis, urosepsis, and metabolic acidosis from poor renal function represent the main sources of morbidity and mortality in newborns with anorectal malformations. Thus, a thorough urologic investigation is mandatory including an ultrasound to evaluate for a urologic obstructive process, and specific to cloacas, to look for hydrocolpos.

Neonatal management

The management of anorectal malformations begins with the prenatal diagnosis [17,18]. Findings such as hydrocolpos, hydronephrosis, hemisacrum, a pelvic mass, hemivaginas, absent radius, or absent kidney all may suggest an anorectal malformation. Fetal intervention has been employed (Fig. 1) [19]. Prenatal management decisions will become increasingly more common.

Once the child is born, a thorough perineal inspection of the newborn boy usually gives the most important clues about the patient's type of malformation [20]. It is important not to make a decision about colostomy or primary operation before 24 hours of life because significant intraluminal pressure is needed for the meconium to be forced through a fistula orifice. If meconium is present on the perineum, that is evidence of a perineal fistula. If meconium is in the urine, there is a rectourinary fistula. Radiologic evaluations do not show the true anatomy before 24 hours, because the rectum is collapsed, and intraluminal pressure has not overcome the muscle tone of the sphincters that surround the lower rectum. During the first 24 hours, the baby should receive intravenous fluids and antibiotics and should be evaluated for cardiac malformations, esophageal atresia, and urologic and spinal problems. Radiography of the lumbar spine and the sacrum and a spinal ultrasound help with this evaluation. An ultrasound of the abdomen will rule out the presence of hydronephrosis and hydrocolpos.

Figure 1. Newborn management for male patients



Algorithm for the management of the newborn male with an anorectal malformation. PSARP, posterior sagittal anorectoplasty; R/O, rule out; U/S, ultrasound.

If the baby has signs of a perineal fistula, an anoplasty without a protective colostomy can be performed. This can be done in the newborn period, or if mitigating circumstances exist can be delayed, with dilations of the fistula until the date of the reconstruction. If on a cross-table lateral film the rectal gas is seen located 2 cm or more above the coccyx, or if the patient has meconium in the urine, with important associated defects, an abnormal sacrum and particularly in the presence of a flat bottom, a colostomy is indicated, and the main repair deferred for a subsequent operation that can be done 4–8 weeks later provided the baby is gaining weight normally.

A divided descending colostomy is ideal for the management of anorectal malformations because it allows for bowel decompression, protection for the final repair, and performance of the distal colostogram [21]. A descending colostomy has advantages over a right or transverse colostomy [22], as it leaves a relatively short segment of defunctionalized distal colon and avoids atrophy of the distal bowel and megarectosigmoid, which occurs with a more proximal colostomy. Mechanical cleansing of the distal colon prior to the definitive repair is much less difficult when the colostomy is located in the descending colon. In the case of a large rectourethral fistula, the patient can pass urine into the colon. A more distal colostomy allows urine to escape through the distal stoma without significant absorption, as opposed to a more proximal colostomy (transverse), where the urine remains in the colon and is absorbed, which can lead to metabolic acidosis. Loop colostomies permit the passage of stool from proximal stoma into the distal bowel, which produces urinary tract infection, distal rectal pouch dilation, and fecal impaction. Prolonged distention of the rectal pouch may produce irreversible dilatation, leading to bowel hypomotility, and severe constipation later in life. Colostomy prolapse is more frequent with loop colostomies. Another problem is a colostomy that is created too distal in the area of rectosigmoid, which may interfere with the mobilization of the rectum during the pull-through.

Repair of these defects in the newborn period without a protective colostomy has been suggested [23]. Such a

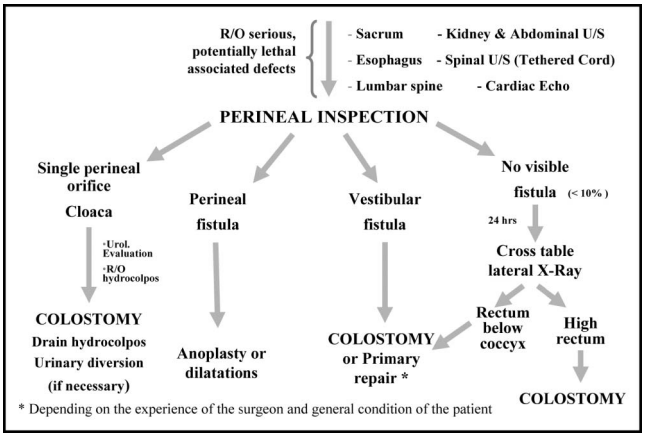
repair is performed without a preoperative distal colostogram, and thus the precise anatomy is not known. During the search for and dissection of the rectum, the urethra, the bladder neck, an ectopic ureter, the vas deferens, or seminal vesicles are at risk of injury [24].

In girls the perineal inspection usually makes the anatomic diagnosis (Fig. 2) [20]. A single perineal orifice establishes the diagnosis of a cloaca. Such babies need a colostomy, which should be performed proximally enough to allow for enough redundant, distal rectosigmoid for the pull-through and vaginal reconstruction if necessary [9]. A hydrocolpos if present must be drained. An endoscopy is recommended in babies with cloaca to define the anatomy. The perineal inspection may show the presence of a perineal fistula, for which a primary anoplasty without a colostomy can be performed. The repair can be delayed with preceding dilations in certain circumstances. A rectovestibular fistula can be repaired during the neonatal period without a protective colostomy if the surgeon has experience and uses a meticulous technique. A colostomy is still the most effective way to avoid perineal infections or dehiscence, which can affect the final functional prognosis.

Surgical techniques

Most pediatric surgeons worldwide perform the posterior sagittal anorectoplasty (PSARP) [2–4] to repair these malformations with or without laparotomy or laparoscopy. Other surgical techniques to repair anorectal malformations have been described including endorectal dissections, an anterior perineal approach [25–27], and many different types of anoplasties [28]. Recent contributions have included performance of these operations primarily [23] and the use of laparoscopy with a minimized perineal incision [29–31].

Figure 2. Newborn management for female patients



Algorithm for the management of the newborn female with an anorectal malformation. R/O, rule out; U/S, ultrasound.

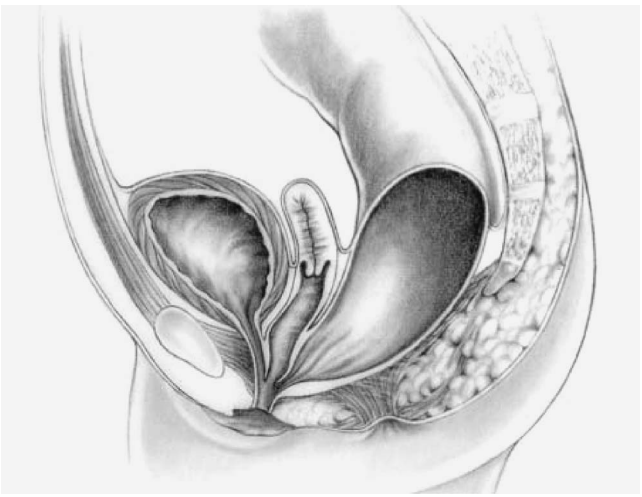
The posterior sagittal approach involves a prone position with the pelvis elevated. An electrical stimulator elicits muscle contraction during the operation, which guides a midsagittal incision, leaving equal amount of muscle on both sides. The incision exposes the subcutaneous tissue, parasagittal fibers, muscle complex, and levator muscles. The sphincter mechanism is divided and the rectum is then separated from the urogenital structures. This delicate maneuver is well visualized with the posterior sagittal approach and injuries are more likely avoided. About 90% of defects in boys can be repaired via posterior sagittal approach alone without entering the abdomen.

A laparoscopic approach, with a very minimal perineal incision, has been described and gives a beautiful view of the pelvis [29]. This approach must be contrasted to a purely posterior sagittal approach, applicable for the vast majority of patients, which is a minimally invasive option, with no abdominal component. For a higher rectum, laparoscopy can be used to avoid a laparotomy. For a rectobladderneck fistula, and possibly a high rectoprostic fistula, the dissection of the rectum from the urinary tract can be facilitated by laparoscopy. Technical limitations currently exist when the rectum is very high and does not reach the perineum. For such a case a meticulous dissection of the blood supply is needed to gain length, which often requires an open approach. Also, if tapering is necessary, this is challenging with only a laparoscopic approach. The step in the laparoscopic procedure of passage of the trocar through the perineum has the potential of injuring the urinary system. In addition, the incidence of postoperative prolapse is not yet known (it is only 3% following PSARP [32]) but may be a concern because of the avoidance of several key PSARP steps, most notably tacking of the rectum to the pelvic muscles [32]. This may be avoided with the recommended laparoscopic pelvic hitch [26]. Finally, long-term continence results are not yet available for this procedure.

The repair of rectoperineal fistula, rectovestibular fistula, and imperforate anus without fistula in girls similarly involves dissection of the rectum but obviously includes dissection of rectum off of the posterior vaginal wall. The most complex defect, seen only in girls, is a cloacal malformation, which is a significant technical challenge [8••] and involves separating the confluence of urinary tract, vagina, and rectum (Fig. 3). Prior to undertaking the main repair, the surgeon should perform an endoscopic study of the cloacal malformation, with the specific purpose of determining the length of the common channel.

There seem to be two distinct groups of cloacal malformations [8••]. There are infants born with a common channel shorter than 3 cm. These defects can be repaired with a posterior sagittal approach only, without a laparotomy. The second group has a longer common channel, and

Figure 3. Cloacal malformation



An artistic drawing of a cloacal malformation in which there is a confluence of urinary tract, vagina, and rectum.

usually needs a laparotomy and multiple complex maneuvers for urinary and vaginal reconstruction and require of the surgeon extensive experience and special training in urology.

The technique of total urogenital mobilization [33] was innovative in that the vagina no longer is separated from the urinary tract but rather the urogenital sinus is mobilized as a unit. With this maneuver, one can gain 2 and 3 cm of length in the repair. The maneuver has the additional advantage of preserving an excellent blood supply to both urethra and vagina, avoids urethrovaginal fistulae, and makes the urethra smooth and visible to facilitate intermittent catheterization when necessary.

Patients with cloaca have a significant incidence of Mullerian anomalies including absent uterus, hemiuterii, atretic cervices, atretic Fallopian tubes, and hemivaginas. The gynecologic anatomy must be investigated either during the main repair or at the time of colostomy closure so that proper reconstruction of internal female anatomy to vagina or neovagina can be performed, pathways for future menstrual flow can be ensured, and potential for pregnancy can be ascertained [10]. Much will be learned in the coming years about the obstetric potential in patients undergoing these types of reconstructive procedures.

Postoperative management

Two weeks after the repair, the patient is started on a protocol of anal dilatations (Table 2). Once the desired size is reached, the colostomy may be closed. Even before the age of toilet training, a bowel movement pattern emerges. A patient who has one to three bowel movements per day, remains clean in between bowel movements, and shows evidence of a feeling or pushing during bowel movements

Table 2. Dilatation protocol

Size of dilator according to age	
Age	Hegar dilator #
1–4 mo	12
4–8 mo	13
8–12 mo	14
1–3 y	15
3–12 y	16
≥12 y	17

usually has a good prognosis. This type of patient is trainable. A patient with multiple bowel movements or one who passes stool constantly without showing any signs of sensation or pushing usually has a poor functional prognosis. Urinary continence is usually only an issue in cloaca patients. Patients with cloacas with a common channel shorter than 3 cm require intermittent catheterization to empty the bladder 20% of the time. Patients with common channels longer than 3 cm will require intermittent catheterization in 70–80% of cases. The rest are successful in voiding spontaneously.

Continence

Most patients who undergo repair of an anorectal malformation have some degree of a functional defecating disorder. All of these patients have an abnormal fecal continence mechanism [34]. Fecal continence depends on three main factors: sphincters, sensation, and motility. Voluntary muscle structures, which are represented by a levator muscle, muscle complex, and external sphincter and are used only for brief periods, when the rectal fecal mass reaches the anorectal area, pushed by the involuntary peristaltic contraction of the rectosigmoid motility. This contraction occurs only in the minutes prior to defecation. These voluntary muscle structures are used only occasionally during the rest of the day and night. Patients with anorectal malformations have abnormal voluntary striated muscles with different degrees of hypodevelopment. Voluntary muscles are used only when the patient feels that it is necessary to use them. For that sensation, the patient needs information that can only be derived from an intact sensory mechanism, a mechanism that many patients with anorectal malformations lack. Sensation resides in the anal canal. Except for patients with rectal atresia, most patients with anorectal malformations are born without an anal canal; therefore, sensation does not exist or is rudimentary. Distention of the rectum seems to be felt by many of these patients, provided the rectum has been located accurately within the muscle structures. This sensation seems to be a consequence of stretching the voluntary muscle (proprioception). The most important clinical implication of this is that liquid stool or soft fecal material may not be felt by the patient with anorectal malformations, as the rectum is not distended. Thus, to achieve some degree of sensation and bowel control, the patient must have the capacity to form solid stool. Bowel motility is perhaps the most important factor in fecal continence.

In a normal individual, the rectosigmoid remains quiet for variable periods (one to several days). During that time, sensation and voluntary muscle structures are almost not necessary because the stool remains in the colon if it is solid. The peristaltic contraction of the rectosigmoid that occurs prior to defecation is normally felt by the patient. Voluntarily, the normal individual can relax the striated muscles, which allows the rectal contents to migrate down into the highly sensitive area of the anal canal. There, accurate information is provided concerning the consistency and quality of the stool. The voluntary muscles are used to push the rectal contents back up into the rectosigmoid and to hold them if desired, until the appropriate time for evacuation. At the time of defecation, the voluntary muscle structures relax. The main factor that provokes the emptying of the rectosigmoid is an involuntary peristaltic contraction, helped sometimes by a Valsalva maneuver. Most patients with anorectal malformation have a disturbance of this sophisticated bowel motility mechanism. Patients who have undergone a PSARP or any other type of sacroperineal approach, in which the most distal part of the bowel was preserved, show evidence of an overefficient bowel reservoir (megarectum). The main clinical manifestation is constipation, which seems to be more severe in patients with lower defects. An ectatic distended colon, sometimes associated with a loop colostomy that allowed fecal impaction in the blind rectal pouch, eventually leads to severe constipation. The enormously dilated rectosigmoid with normal ganglion cells behaves like in a myopathic type of hypomotility disorder. The patients with fecal incontinence who have constipation can be treated with enemas. Patients treated with techniques in which the most distal part of the bowel was resected clinically behave as individuals without a rectal reservoir. This is a situation equivalent to a perineal colostomy. Depending on the amount of colon resected, the patient may have loose stools. In these cases, medical management consists of enemas plus a constipating diet and medications to slow down the colonic motility.

Evaluation of results

Each defect has a different prognosis. Categories such as 'high,' 'intermediate,' and 'low,' or even 'high' and 'low' frequently include individual malformations with different prognoses.

Table 3 shows the clinical results obtained in the largest reviewed series [35]. Patients with a very abnormal sacrum (<0.3 ratio) and flat perineum have fecal incontinence regardless of the type of malformation. Because cloacas represent another spectrum of defects themselves, they must be subclassified on the basis of potential for bowel and urinary control. The length of the common channel seems to be the most important prognostic factor.

Complications

Constipation is the most common functional disorder observed in patients who have undergone PSARP. Patients with the best prognosis have the highest incidence of constipation and patients with very poor prognosis, such as bladderneck fistula, have a low incidence of constipation. Constipation seems to be directly related to the degree of preoperative rectal ectasia. Colostomies that do not allow cleaning and irrigation of the distal colon lead to

Table 3. Clinical results

	Voluntary bowel movement		Soiling		Totally continent		Constipated	
	Patients	*n (%)*	Patients	*n (%)*	Patients	*n (%)*	Patients	*n (%)*
A. Bowel incontinence								
Perineal fistula	39/39	100	3/43	20.9	35/39	89.7	30/53	56.6
Rectal atresia or stenosis	8/8	100	2/8	25	6/8	75	4/8	50
Vestibular fistula	89/97	92	36/100	36	63/89	70.8	61/100	61
Imperforate anus without fistula	30/35	86	18/37	48.6	18/30	60	22/40	55
Bulbar–urethral fistula	68/83	82	48/89	53.9	34/68	50	52/81	64.2
Prostatic fistula	52/71	73	67/87	77.1	16/52	30.8	42/93	45.2
Cloaca (short common channel)	50/70	71	50/79	63.3	25/50	50	34/85	40
Cloaca (long common channel)	18/41	44	34/39	87.2	5/18	27.8	17/45	34.8
Vaginal fistula	3/4	75	4/5	80	1/3	33.3	1/5	20
Bladderneck fistula	8/29	28	39/43	90.7	1/8	12.5	7/45	15.6

	Patients	*n (%)*
B. Urinary incontinence		
Rectal atresia or stenosis	0/8	0
Perineal fistula	0/38	0
Bulbar – urethral fistula	2/85	2.4
Imperforate anus without fistula	1/37	2.7
Prostatic fistula	7/85	8.2
Bladderneck fistula	7/38	18.4
Cloaca (short common channel)	5/18	27.8
Vaginal fistula	1/5	20
Cloaca (long common channel)	37/48	77.1

megarectum, and transverse colostomies lead to a 'micro' left colon with dilatation of the rectosigmoid. Keeping the distal rectosigmoid empty and not distended from the time the colostomy is established results in better ultimate bowel function.

A large group of patients have severe complications after an attempted but failed repair of an anorectal malformation, particularly urologic injuries. The posterior sagittal approach when performed without a good previous distal colostogram was the most important source of these complications [24]. Neurogenic bladder in male patients with anorectal malformations is extremely unusual because it only happens in patients with a very abnormal sacrum, unless it reflects denervation of the bladder and bladderneck during the repair [24].

Cloaca patients have a deficient emptying mechanism of the bladder. These patients have a flaccid, smooth, large bladder that does not empty completely. Fortunately, most patients with cloacas have a very good bladderneck. The combination of a good bladderneck with a floppy, flaccid bladder is ideal for intermittent catheterization, which keeps the patient completely dry. Exceptions to this are patients without a bladderneck, such as those with a very long common channel, with hemivaginas attached to the bladderneck (in whom, after separation of these structures, there is essentially no bladder neck), and patients who are born with separated pubic who have no bladderneck. These patients eventually require a continent diversion. Urethrovaginal fistula was the most common complication in cases of persistent cloaca but has essentially been eliminated by the total urogenital mobilization maneuver [33].

Fecal incontinence

For patients with different degrees of incontinence, a program of bowel management exists for them to be kept completely dry of urine and clean of stool, either because they achieve urinary and bowel control or because the implemented program keeps them artificially dry and clean [36]. With the rational administration of bowel irrigations, diet, and drugs, most patients, including those with severe fecal incontinence, are able to remain clean for 24 hours. Only patients with severe diarrhea secondary to an absent or a short colon are candidates for a permanent colostomy.

Patients with fecal incontinence are evaluated and classified into those with constipation and those with increased motility (tendency to have diarrhea). In the first group, bowel irrigations must be aggressive. Because of the decreased bowel motility in constipated patients, they remain clean for the next 24 hours. No laxatives or special diet are required.

The second group (patients who have increased bowel motility due to loss of the rectal reservoir) requires a constipating diet, medication to decrease bowel motility, and a program of colonic irrigations. The program is adjusted by trial and error over a period of 1 week. The vast majority of patients remains clean and has an acceptable quality of life. Bowel management is started when the patient starts school, and all of his or her classmates are already wearing regular underwear. When the patient reaches the age of 7–12 years, or sometimes younger, he or she can gain more independence by self-administering the enema via a catheterizable stoma. A continent appendicostomy (Malone procedure) is an excellent option [37,38]. This operation creates a communication between the abdominal wall and the cecum through the appendix, creating a valve mechanism that allows the catheterization of the cecum but prevents leakage of stool. This allows the patient to administer his or her own enema while sitting on the toilet. The operation consists of plicating the cecum around the native appendix and exteriorizing the appendix in the deepest part of the umbilicus, making it inconspicuous. A button cecostomy has been advocated for a similar purpose [39,40]. A significant number of patients do not have an appendix. In such cases an appendix can be created with a tubularized flap of the cecum [38].

Conclusion

Management of anorectal malformations requires an accurate diagnosis, correct newborn management with either anoplasty or colostomy, and treatment of associated malformations. An anatomic reconstruction at the appropriate time is vital. Postoperatively, an understanding of continence and its comprehensive management lead to a child that is clean and dry with an excellent quality of life, because he or she either possesses the capacity for continence or can be kept artificially clean.

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- •• of outstanding interest

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