CHAPTER 77
Anorectal Malformations

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Introduction

Anorectal malformations (ARMs) occur commonly throughout the African continent.[1] Even under the best of circumstances, children undergoing operative treatment for ARMs may have lifelong bowel management problems of constipation, incontinence, and encopresis. In some areas of Africa with severely limited resources, few surgeons have expertise in the definitive management of these malformations. Operations are often performed out of desperation by doctors with minimal experience using inadequate equipment. Many of the affected children, not unexpectedly, suffer a lifetime of misery from bowel management problems. Other children undergo a colostomy in the newborn period, and definitive treatment is delayed until a surgeon with appropriate expertise is available. Due to the paucity of trained paediatric surgeons in most of Africa, many children throughout Africa may be living with a colostomy and hoping that someone with expertise will come along to help them achieve a more normal life. In some instances, particularly in rural areas, neonates identified with ARMs at birth are abandoned or euthanized because they are considered "nonviable" due to their abnormality. Some females with rectovestibular fistulas large enough for adequate defecation are not brought for operative correction until it is time for marriage (Figure 77.1).

Pathophysiology

A description of the embryological events resulting in ARMs is beyond the scope of this chapter. It is much more important that the African paediatric surgeon concentrate on understanding the various types of ARMs and how to best manage them. ARMs occur in an estimated 1 in 4,000 births worldwide, but this estimate is based on data from more developed countries where accurate birth records are available. To the paediatric surgeon sitting in a grossly overcrowded surgery clinic in a major African teaching hospital, the incidence of ARMs appears much higher. This, however, may be an artifactual observation because most children with ARMs are referred to these tertiary referral centres, whereas children with other less complicated operative problems are managed locally without referral. An impressive multicentre African study[1] demonstrated that ARMs are at least as common in Africa as in other parts of the world. ARMs in children are among the most common and most complex problems that the African paediatric surgeon will confront.

The term "imperforate anus" has traditionally been used to describe all anorectal abnormalities in females and males. Although imperforate anus implies that the anus never opened anywhere, a purely blind anal pouch is actually rare. Usually the rectum has opened either onto the perineum or into the genitourinary tract. The spectrum of abnormalities is quite broad, and therefore this chapter uses the term "anorectal malformations" for all of these abnormalities with further clarification to describe the specific malformation. "Imperforate anus" then refers to the specific portion of any anomaly where the rectum does not open properly through the anal musculature.

The most common ARM defect in males is imperforate anus with a rectourinary tract (usually urethral) fistula (Figure 77.2(A)). The next most common ARM defect for males is a fistula into the perineum, a bucket-handle abnormality, or the presence of meconium in a midline perineal, scrotal, or penile raphe.

In females, the most common defect is imperforate anus with a rectal fistula into the vestibule of the vagina (Figure 77.2(B)). Perineal fistulas and bucket-handle deformities also occur occasionally in females. An uncommon but very complicated female defect is persistent cloaca, in which the urethra, vagina, and rectum empty into a single common perineal opening (Figure 77.2(C)).

Diagnosis

The diagnosis of ARM should be made during the newborn physical examination. Many children in Africa are not born in health care facilities, however, and the absence of an anus may not be appreciated by the family until hours or days after birth,[2,3] when it is noted that the child's abdomen is distending and the infant has not passed meconium (Figure 77.3).

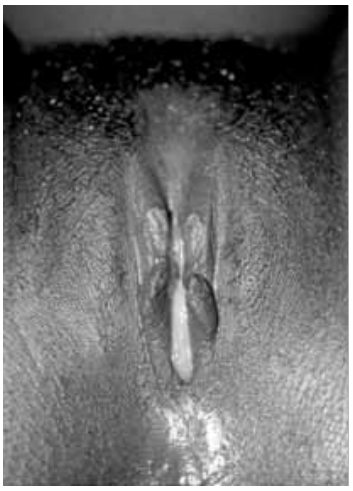

Figure 77.1: Previously untreated congenital ARM (rectovestibular fistula) in a 22-year-old female. The patient was brought for operative correction by her father, who hoped to increase her bride price. Note previous female circumcision.

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Figure 77.2: Three types of anorectal malformations: (A) male rectourethral fistula, (B) female rectovestibular fistula, (C) female persistent cloaca.

When consulted for a newborn with ARM, the African surgeon should initially note the child's gender and examine the perineum carefully to see whether there is any evidence of meconium from a perineal fistula (Figure 77.4) or along a midline raphe (Figure 77.5) or whether the child has a true bucket-handle deformity (Figure 77.6). If the child is male and there is no initial evidence of visible meconium, the child should be observed for 12 hours or more to see whether meconium appears in the perineum. During this time, gauze is placed over the penis so that the urine can be examined for evidence of meconium. If there is meconium in the urine, the diagnosis of a rectourinary tract fistula is made, and no further diagnostic procedures are indicated.

If, however, there is no meconium identifiable in the urine or on the perineum, an invertogram is performed because a very small percentage of children (more common in children with Down syndrome) will have a blind pouch without a fistula. The child is placed in a prone jackknife position with the buttocks higher than the rest of the body for at least 30 minutes. After that time, a radio-opaque object (drop of barium or coin) is placed on the anal dimple, a cross-table lateral x-ray is performed, and the distance between the rectal air bubble and the perineal skin is measured (Figure 77.7). If the distance is less than 1 cm, the lesion can be treated as a low lesion (see next section), but any distance greater than 1 cm should be managed as a high lesion. In females, the labia should be grasped for traction and the posterior portion of the vestibule (just external to the hymen) examined for a rectovestibular fistula (Figure 77.8). In persistent cloaca, there is only one perineal opening, and therefore the separate openings for urethra, vagina, and rectum are not visible.

When the diagnosis of ARM is made, the child should be examined for other components of the VACTERL (**V**ertebral and spinal cord, **A**norectal, **C**ardiac, **T**racheo**E**sophageal, **R**enal and other urinary tract, **L**imb) complex of anomalies. Diagnostic modalities must be appropriate for the particular locale. Therefore, in some African locales an echocardiogram may be utilized, whereas in others a stethoscope may be the only cardiac diagnostic modality available. An orogastric tube is used to test for patency of the esophagus. An ultrasound, if available, is the best way to initially assess the urinary tract for abnormalities.

Treatment

Initial determination of the particular type of ARM will determine the proper initial treatment for the newborn. For many years, defects were classified as high, mid, or low. For the purpose of simplifying the treatment protocol for African paediatric surgeons, lesions in this chapter will be classified only as high or low. A colostomy is recommended as the initial treatment for high lesions, whereas low lesions can be treated primarily with an anoplasty. Low lesions that can be treated without a colostomy include those with evidence of meconium in the perineum or a bucket-handle lesion and those with a blind pouch less than 1 cm from the anal dimple, as demonstrated on invertogram. The most common lesions, including rectourinary tract fistulas in males and rectovestibular fistulas in females, should be treated as high lesions with an initial colostomy.

Creation of a proper colostomy is a difficult operation; it should *not* be assigned by default to the lowest-ranking physician who happens to be taking call after a long day in the operating theatre. The most common colostomy performed in African hospitals is a right transverse loop colostomy because it can be done quickly by simply pulling the colon out through a small incision and keeping it from returning to the abdomen by placing a rubber catheter beneath the protruding colon. Without adequate fixation, however, prolapse often occurs[4] (Figure 77.9), which can reach gigantic proportions. Death has even been reported from complications of a colostomy.[5] Children in Africa often have to live with their colostomies for months or years, and mistakes made at the initial colostomy creation result in long-term misery for the patient and the family (Figure 77.10).

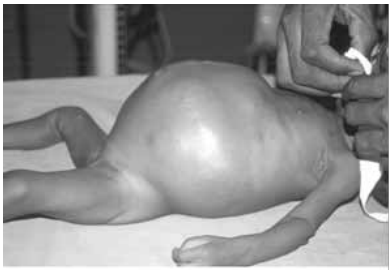

Figure 77.3: Four-day-old male with ARM brought to hospital moribund with a history of abdominal distention and no meconium since birth.

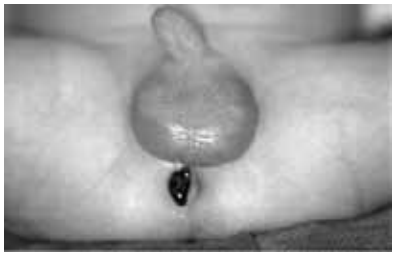

Figure 77.4: Perineal fistula in male newborn.

Figure 77.5: Newborn male with peno-scrotal midline raphe meconium.

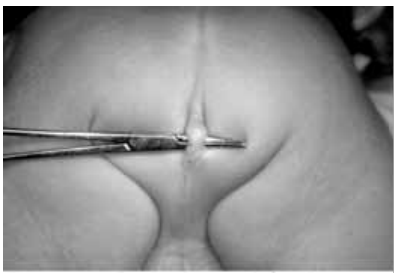

Figure 77.6: Bucket-handle deformity.

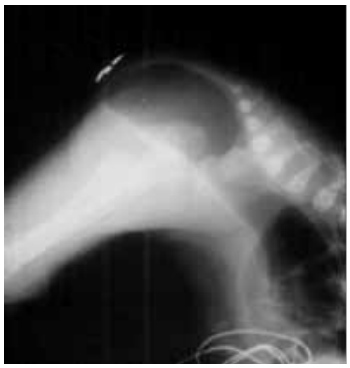

Figure 77.7: Invertogram demonstrating a low (<1 cm) blind pouch.

Alberto Peña has repeatedly emphasized the importance of a double-barrel colostomy to achieve total diversion of faeces.[6] This is particularly important in males because there is usually a fistula between the distal colon and the urinary tract, and undiverted stool in the distal colon may cause repeated urinary tract infections (UTIs). It is best to place the colostomy in the distal descending or proximal sigmoid colon to make the distal limb shorter than it would be in a transverse colostomy. Many male children with ARMs have urine flow from the urethra into the distal colon and then out the mucous fistula (distal stoma), and if urine stays in the colon for long periods of time, it can cause a significant metabolic acidosis. However, if the distal sigmoid is mistakenly selected for the colostomy, the distal colon may be too tethered to properly come down to the perineum and may require taking down the colostomy at the time of anorectoplasty.

A recommended method for creation of a colostomy is to make a transverse muscle transecting incision in the left abdomen just below the level of the umbilicus. The sigmoid colon is identified and traced proximally and distally to be sure that it is truly the sigmoid and not the transverse colon. A point is selected in the *proximal* sigmoid area, and the colon is transected at this point. The proximal sigmoid is less likely to prolapse because the distal descending colon is fixed to the left lateral peritoneal reflection. After transecting the colon, it is quite important to irrigate the distal colon with warm normal saline to remove all meconium, taking care that the effluent does not get into the peritoneal cavity. If this meconium is not properly removed at the time of colostomy, the meconium will desiccate and form large impacted faecal rocks, which complicate subsequent anorectoplasty. In order to prevent prolapse, the two stomas are secured to the peritoneum and fascia at opposite ends of the wound by using a minimum of six small absorbable sutures (3/0-5/0 vicryl) for each stoma. Approximately 2 cm of each stoma should protrude past the skin level. There is no need to mature these small stomas because they will spontaneously mature, and attempts at operative maturation may occlude the lumen. One or two sutures (3-0 vicryl, polydiaxanone, nylon, silk) are used to approximate the peritoneum, fascia, and muscle between the stomas to minimize parastomal herniation.

A skin bridge is created between the stomas by using interrupted, rapidly absorbable small sutures. The bridge must be wide enough to totally separate the stomas (Figure 77.11). When the child reaches approximately 2–3 months of age, and if the child appears in very good nutritional status as evidenced by a weight of 8–10 kg, a definitive posterior sagittal anorectoplasty (PSARP) can be considered. In males, a distal colostogram should be performed prior to operation to determine the site of the fistula. This is performed by inserting a Foley catheter into the mucous fistula and inflating the balloon enough to occlude the colon lumen. The child is placed in a lateral position on the x-ray table with the hips flexed. Under fluoroscopy, if available, a water-soluble contrast medium is injected to fill the distal rectum and adequately identify the place of entry into the urinary tract (Figure 77.12). If fluoroscopy is not available, proper timing of injection with a plain film x-ray can usually adequately define the fistula.

Posterior Sagittal Anorectoplasty Technique

The technique of PSARP, popularized by Alberto Peña in the 1980s,[7] is currently the most commonly utilized procedure for repairing ARMs. This is the procedure, with a few modifications for African practitioners, that is described in this chapter. PSARP in a male with a urethral fistula is described first because this is a commonly encountered lesion and is the most difficult to repair. A urinary catheter is inserted before definitively positioning the patient. Sometimes the catheter goes through the fistula and into the rectum instead of into the urinary tract. If no urine is obtained from the catheter, the balloon should not be inflated in case it has curled up in the urethra. If the catheter is in the rectum, it can later

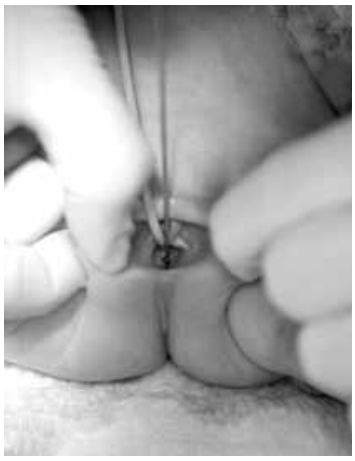

Figure 77.8: Most common female ARM. Catheter is in urethra; probe is in rectovestibular fistula.

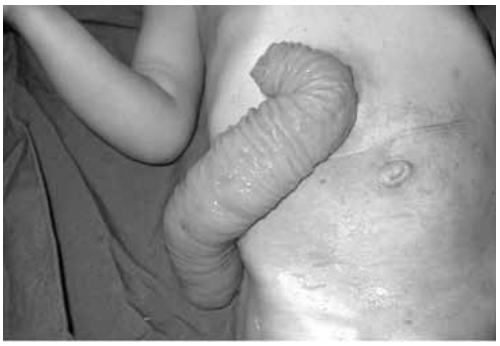

Figure 77.9: Prolapsed right transverse loop colostomy.

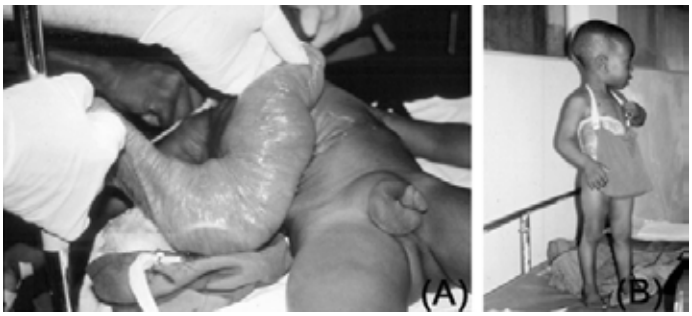

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Figure 77.10: This three-year-old child with Hirschsprung's disease underwent a colostomy at three months of age. Now he is awaiting someone to perform a definitive procedure. (A) Tremendous prolapse. (B) Colostomy bag improvised by the mother to keep child from stepping on his colostomy.

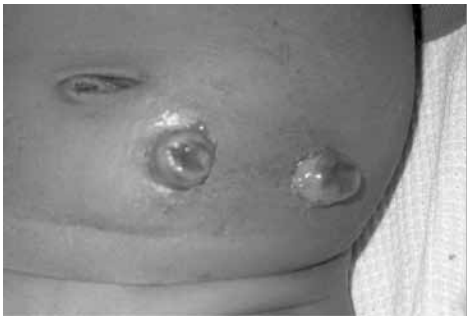


Figure 77.11: Double-barrel colostomy with intervening skin bridge.

be manipulated into the bladder. PSARP is performed with the child in a prone, jackknife position with all pressure points properly padded (Figure 77.13).

A Peña muscle stimulator is the best method for visualizing the perineal musculature, but these stimulators are quite expensive (>US$3000), so they are economically inaccessible for most African paediatric surgeons. As a result, many African surgeons use an electrocautery to identify the musculature, but this may cause damage to precious tissues, even on a low setting. The risk is decreased if a needle-point diathermy is used for all incisions. A much less expensive (US$50) and more appropriate stimulator can be improvised by using an anaesthesia nerve stimulator and a piece of solid, double-pronged, insulated wire, available in hardware stores around the world (Figure 77.14).

The insulation is removed for a distance of 1 cm from both ends of both pieces of the wire. The wire itself can be sterilized by soaking in an appropriate sterilizing solution. One end of both wires is connected to the stimulator. The other ends of the wire are left close to, but not touching, each other. This then serves as the handpiece used to touch the patient. Nonsterile personnel push the "continuous tetany" button on the stimulator whenever the surgeon wants to stimulate the patient with the handpiece to assess muscle contraction.

It is important during PSARP operations that anaesthesia personnel not administer a muscle relaxant because this impairs the use of any muscle or nerve stimulator. If a muscle relaxant is absolutely necessary for intubation, it must be a very short-acting one because the muscle/nerve stimulator should be used early in the operation to define the musculature before any incision is made.

After mapping out the perineal musculature with a stimulator, temporary stay sutures are placed on either side of the midline at the anterior and posterior limits of the anal muscle complex (sphincter muscle) to identify the limits of the complex later in the operation if needed. A posterior sagittal incision is performed from the coccyx to the perineal body area, preferably with a needle-tip electrocautery placed on "cutting" current. After incising the skin, the deeper tissues can be incised by using the "coagulation" setting. The stimulator is used frequently to ensure that a true midline incision is being performed with equal musculature on either side. The incision is carried in the midline through the parasagittal fibres, the muscle complex, and the levator muscle (Figure 77.15(A)).

Deep to the levator, the white appearance of the rectum should be visualized. Attempts at dissecting around the rectum at this time should be avoided. Distally, the rectum is opened longitudinally to view the inside of the rectal lumen (Figure 77.15(B)). Anteriorly, the small fistula into the urinary tract is seen. Traction sutures are placed, and the anterior rectal wall is very carefully dissected from the posterior urethral wall (Figure 77.15(C)). This is the most difficult part of the operation.

After the initial dissection from the urethral wall itself, the dissection becomes easier above the prostate at the level of the bladder itself. The urinary fistula site should be carefully closed with an absorbable, fine suture. The rectum is then dissected circumferentially, staying close to the rectum itself so as to avoid damage to surrounding nerves, muscle, and urinary tract structures (Figure 77.15(D)). Fibrotic neurovascular bands are taken down to further free the rectum (Figure 77.15(E)). When the rectum is freed enough to come to the perianal skin without significant tension, the closure is performed. Significant tension will cause the neoanus to retract, resulting in lack of epithelial continuity with subsequent scarring and the need for another operation. The muscle anterior to the neoanus, including the perineal body, is closed with fine (4/0, 5/0 vicryl), absorbable suture (Figure 77.15(F)). The colon is placed deep to the levator muscle, and the levator approximated in the midline, taking a bite of the posterior colon wall also. The closure continues along the posterior portion of the muscle complex (sphincter muscle) out to the skin (Figure 77.15(G)). This closure should be close to the previously placed stay sutures marking the posterior limits

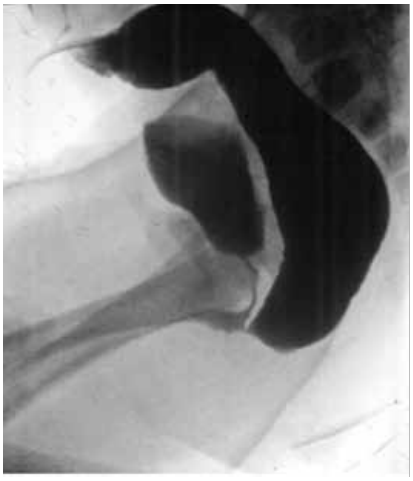


Figure 77.12: Colostogram showing fistula of rectum into bulbous urethra.

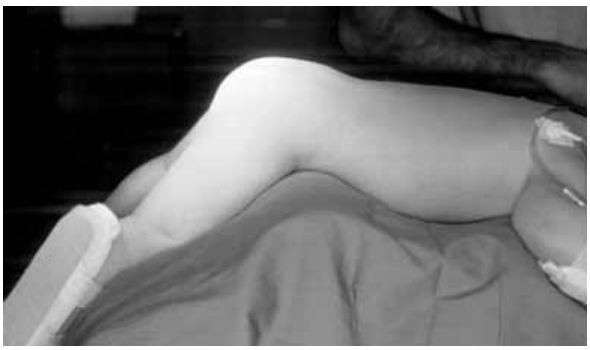
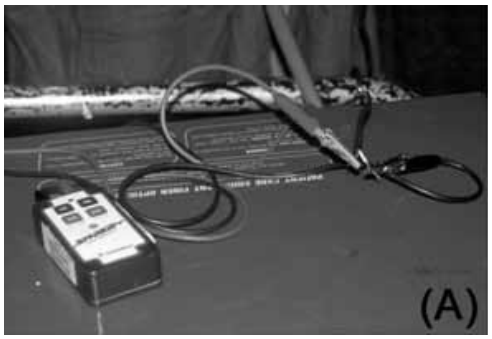


Figure 77.13: Positioning of patient for PSARP.



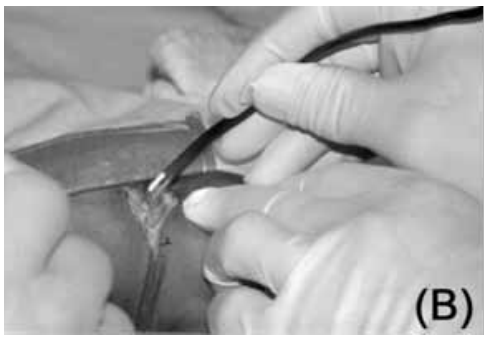


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Figure 77.14: Improvised anal stimulator: (A) anaesthesia nerve stimulator; (B) handpiece made from solid, double-pronged, insulated wire.

of the muscle complex. Any protruding rectum is debrided, but care is taken to avoid excess tension on the subsequent anastomosis. A 16-suture anoplasty is performed by suturing the full thickness of the colon (neoanus) to the full thickness of the skin (Figure 77.15(H)). A dilator is placed to be sure that the lumen has not been occluded. The rest of the closure is performed, including the parasagittal muscles and subcutaneous tissue. The skin is closed with interrupted full-thickness rapidly absorbing suture (gut) or with a removable running subcuticular suture. There is no good way to secure a dressing to the area, so it is either left open to the air or covered with an antibiotic ointment.

When the rectal fistula goes into the urinary bladder instead of the urethra, a combined abdominal and perineal approach is utilized. The urethral catheter is left in situ for 5–7 days after a male PSARP, and the urine is cultured on its removal. If the patient is nursed prone postoperatively, there is less likelihood of contamination of the wound.

The most common female lesion, a rectovestibular fistula, is also repaired by using PSARP, as described for male lesions. Stay sutures are placed around the vestibular fistula site, and the rectum is carefully dissected from the posterior vaginal wall. This part of the dissection is the most difficult part of the female operation. The rest of the female operation is almost identical with that described above for males.

Females with a persistent cloaca defect present a very difficult challenge. These children should undergo initial decompression with a divided colostomy and a vesicostomy or vaginostomy, as needed to adequately provide urinary drainage. The definitive operation for repair of a persistent cloaca with an anorectoplasty and urogenital mobilization[8] should be delayed until a surgeon with proper expertise can undertake this difficult procedure in an appropriate referral centre.

Dilatation of the neoanus should begin 2–3 weeks postoperatively if the perineal wound has healed well. Ideally, plastic-graded dilators (commercially available for approximately US$10 each) can be provided for the mother to use at home. When such dilators are not available (the usual scenario), the mother can be taught to use her gloved fingers, progressing from the distal phalanx of the little finger to the proximal phalanx of the long finger over a few weeks. Another alternative is the use of an appropriately sized candle for the dilatations. The first dilatation should be performed by the surgeon in the clinic or ward. Starting with an 8-mm size, the surgeon sequentially passes dilators until there is slight resistance. The mother is sent home with this minimally snug dilators and the next larger one. She dilates the anus twice a day for a week and then moves to the next larger size for twice per day dilatations for the next week. After 2 weeks, she is seen in the clinic again, and the next two larger dilators are utilized for the next 2 weeks. When the target size (12 or 13 mm) is reached, and if the perineum has totally healed, the colostomy can be closed, but the dilatations must be continued for at least a year postoperatively.

Anorectoplasties performed by surgeons without proper expertise or equipment can result in a grossly misplaced anus (see Figure 77.16(A)) and a potential lifetime of misery for the child. A secondary operation by a surgeon with both expertise and equipment, although not as successful as a primary operation, is highly recommended *before* closure of the colostomy (Figure 77.16(B)).

Bowel Management Programs

Even under the best of circumstances, some children—particularly those with high ARMs—suffer from constipation or faecal incontinence caused by encopresis (escape of liquid stool around a large, hard faecal impaction). Bowel management programs have been initiated by hospitals in the United States to help children with these problems.[9]

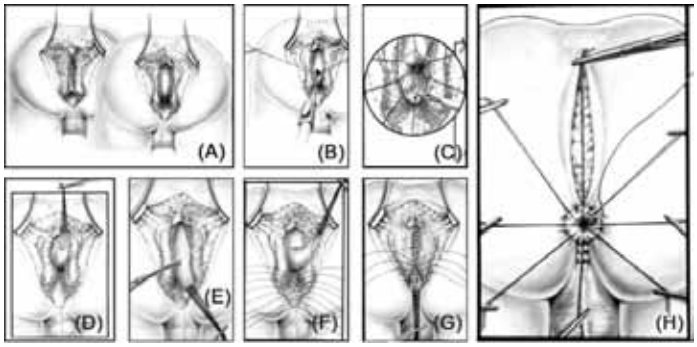

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Figure 77.15: The Peña technique for posterior sagittal anorectoplasty: (A) posterior sagittal incision through parasagittal fibres, muscle complex, and levator muscle; (B) distal rectum opened longitudinally; (C) dissection of rectum from fistula and posterior urethra; (D) circumferential dissection of rectum; (E) incision of neurovascular bands; (F) closure of muscle, including perineal body, anterior to rectum; (G) closure of levator muscle and posterior muscle complex; (H) 16-suture anoplasty.

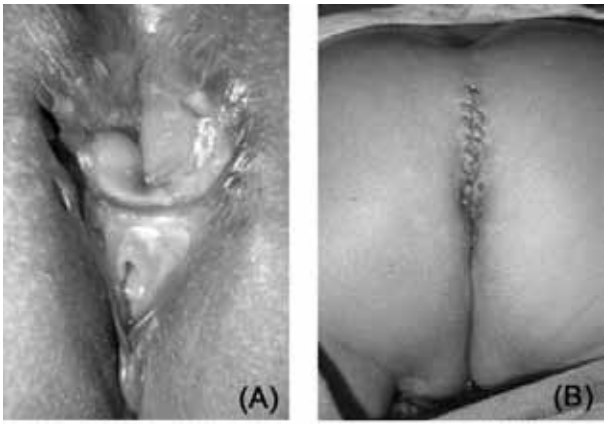

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Figure 77.16: (A) Misplaced anorectoplasty. (B) Better placement after a secondary salvage operation.

The aim of these programs is to return children to normal lives at school in unsoiled underwear. The overriding principle behind bowel management is that the child learns to evacuate in a socially acceptable place (home) at a socially acceptable time (before or after school). The primary technique used to achieve continence is an enema regimen. The child is given enemas each night or early in the morning at home to completely evacuate the colon. The child can then go to school with confidence, knowing that there will be no stool accidents. After a successful enema regimen has been established, an antegrade continent enema (ACE) operation (known also as an ACE procedure) can be considered.[10] This procedure uses an appendicostomy as a conduit for administering antegrade enemas for evacuating the colon. This technique has been utilized with great success in achieving continence in children who were previously social recluses due to their bowel management problems.[11]

Key Summary Points

1. Anorectal malformations (ARMs) occur commonly and are a cause of significant morbidity for African children.
2. The most common ARM in males is a fistula of the rectum into the urinary tract (usually urethra); in females, it is a fistula into the vestibule of the vagina.
3. ARMs are one component of the VACTERL complex, and diagnostic techniques appropriate for a particular locale should be used to look for the other components of this complex.
4. The specific type of ARM is diagnosed on the basis of the newborn physical examination and a few technologically easy techniques.
5. Low lesions, which unfortunately constitute the minority of lesions, can be treated by a primary anoplasty without a colostomy.
6. Most ARMs are high lesions and should be treated initially with a sigmoid colostomy.
7. Improperly constructed colostomies can be a source of great morbidity for children.
8. The recommended definitive treatment for ARMs is a posterior sagittal anorectoplasty.
9. Even after the best of operations, children with ARMs can suffer from bowel management problems. A proper bowel management program can make a significant positive change in the life of a previously miserable child with faecal incontinence.

References

1. Moore SW, Alexander A, Sidler D, Alves J, Hadley GP, Numanoglu A, Banieghbal B, Chitnis M, Birabwa-Male D, Mbuwayesango B, Hesse A, Lakhoo K. The spectrum of anorectal malformations in Africa. Pediatr Surg Int 2008; 24:677–683.
2. Uba AF, Chirdan LB, Ardill W, Edino ST. Anorectal anomaly: a review of 82 cases seen at JUTH, Nigeria. Niger Postgrad Med J 2006;13(1):61–65.
3. Adejuyigbe O, Abubakar AM, Sowande OA, Olayinka OS, Uba AF. Experience with anorectal malformations in Ile-Ife, Nigeria. Pediatr Surg Int 2004; 20:855–858.
4. Archibong AE, Idika IM. Results of treatment in children with anorectal malformations in Calabar, Nigeria. S Afr J Surg 2004; 42(3):88–90.
5. Johnson O, Ghidey Y, Habte D. Congenital malformation of anus and rectum in Ethiopian children. Ethiop Med J 1981; 19(1):9–15.
6. Peña A. Anorectal malformations. In: Ziegler M, Azizkhan R, Weber T, eds. Operative Pediatric Surgery. McGraw Hill Publishers, 2003, Pp 739–761.
7. Peña A. Atlas of Surgical Management of Anorectal Malformations. Springer-Verlag, 1989.
8. Peña A. Total urogenital mobilization: an easier way to repair cloacas. J Pediatr Surg 1997; 32:263–268.
9. Peña A, Guardino K, Tovilla JM. Bowel management for fecal incontinence in patients with anorectal malformations. J Pediatr Surg 1998; 33:133–137.
10. Malone PS, Ransley PG, Kiely EM. Preliminary report: the antegrade continence enema. Lancet 1990; 336:1217–1218.
11. Meier DE, Foster E, Guzzetta PC, Coln D. Antegrade continent enema management of chronic fecal incontinence in children. J Pediatr Surg 1998; 33(7):1149–1152.