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Caring for children with colorectal disease in the context of limited resources

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The management of patients with colorectal disease in the pediatric population is challenging. Such management is all the more challenging when facing the constraints imposed by an environment with limited clinical resources. Three types of colorectal problems are highlighted in this article: anorectal malformations, Hirschsprung's disease, and acquired rectovaginal fistula in the human immunodeficiency virus–positive population. Through the use of illustrative cases, the authors discuss the pitfalls and challenges encountered in the diagnosis, treatment, and appropriate disposition of these patients. The bulk of the experience used to write this article was acquired in low- and middle-income countries in Africa. The authors hope that the lessons learned will help others manage such patients in the context of limited resources, but recognize that challenges will vary from place to place. There is no substitute for local, contextual expertise.

In environments with limited resources, the management of patients with colorectal disease is particularly challenging. Three pathologic conditions (anorectal malformations [ARM], Hirschsprung's disease [HD], and acquired rectovaginal fistula in human immunodeficiency virus [HIV]–positive patients) have been selected and several cases described to highlight some of the "pearls" and "pitfalls" of patient management in such environments. The authors recognize that every environment will pose its own challenges. By illustrating and discussing these cases, the hope is that treating surgeons will not only realize that they are not alone in facing such challenges but that they may take away practical points for diagnosis, treatment, and ongoing management.

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Management of anorectal anomalies in resource-limited settings

Case 1

A 1-day-old male child was brought to a district hospital with the complaint of an absent anal opening. The child was generally well, had well-formed buttocks and a bucket-handle skin tag on the perineum. There was no obvious meconium draining. An invertogram was ordered but the x-ray machine was not working. A decision for surgery was made and a divided sigmoid colostomy fashioned. The child

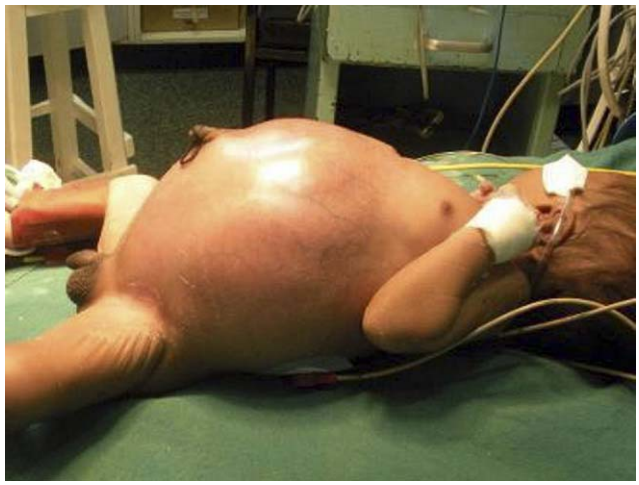

Figure 1 Delayed presentation (5 days) of male ARM. (Color version of figure is available online.)

was discharged with referral to the national hospital for definitive care, but remained untreated for 3 years due to lack of funds for transportation to the capital city. He was finally assessed in another suitable unit where a diagnosis of low imperforate anus was made by distal colostogram and perineal ultrasound. An anoplasty was fashioned and the colostomy closed immediately thereafter. On follow-up, the child was found to be well but suffering from constipation. Dietary measures were suggested.

Points of discussion

- *One day old*: This is a surprisingly early presentation for resource-limited settings. The more usual scenario is that of delayed presentation (Figure 1), but generally male defects are brought in earlier because of the inability to pass stool. Many infants with ARM are missed because a small amount of meconium is noted in the diapers.
- *Bucket-handle skin tag*: In resource-limited settings, clinical signs are particularly important. A majority of bucket-handle tags are associated with a low anorectal malformation, and could be treated by immediate anoplasty if a surgeon comfortable with such a procedure were available. Careful probing with a lacrimal probe (or equivalent) at the base of the bucket-handle may have revealed a tiny opening draining meconium.
- *Invertogram*: A simple investigation that is generally (but not uniformly) available. Proper timing and technique are crucial. It may be done as a cross-table lateral radiograph, which is easier on the patient. If performed at least 8 to 24 hours after birth, the distance from the distal end of the air column to a radiopaque marker on the skin in the anal dimple will always be equal to or greater than the true distance to the rectal pouch. Low forms (<1 cm) may be treated by immediate anoplasty in the hands of an experienced surgeon.
- *Colostomy*: The ability to perform an urgent or emergent colostomy in an appropriate hospital setting (often anesthesia being the limiting factor) is an essential skill in the management of patients with anorectal anomalies. It is preferable to divert the fecal stream whenever in doubt regarding the level of the anomaly, or if the operator is unsure of his/her ability to perform an immediate unprotected anoplasty. It must be emphasized that the initial anoplasty affords the best chance at function: putting this long-term function at risk is not acceptable. (While colostomy in adults is seldom well accepted, it is more acceptable in small children. Availability of colostomy supplies varies according to the environment. When not available, simple stoma coverage with a washable wrap-around cloth seems quite effective and culturally acceptable.)
- *Untreated for 3 years*: Ironically, the life-saving ability to perform colostomies in neonates may result in a backlog of non–life-threatening congenital conditions. Having transformed the acute situation from an emergent to an elective one, these congenital problems become long-term chronic disabilities. It is crucial for pediatric surgeons to be advocates for comprehensive approaches to health care provision, which focus not only on treating emergency conditions but also on addressing the chronic surgical disabilities and their related challenges. Although some of these "long-term" problems can be treated through "short-term" surgical missions, the long-term solution involves training more local specialists and retaining them in state/provincial and other affordable specialized hospitals.
- *Perineal ultrasound*: A simple investigation, often available in low- and middle-income settings. It is important though to ensure that the technology comes with the skill—the distance from skin to rectal pouch can be erroneously shortened if excessive pressure is applied with the probe. A good invertogram or its equivalent is sufficient and much less operator-dependent.
- *Anoplasty*: The definitive procedure. The first attempt is the best opportunity for long-term near-normal function. In the context of limited follow-up, we recommend a wide anoplasty done by a well-trained surgeon to lessen or avoid the need for dilatations.
- *Mobile clinic*: The need for follow-up in patients with anorectal anomalies is so great that every effort should be made to review the patients on a regular basis. This may include going to where the patients are located, providing funds for their transportation, and/or mobilizing social workers or community health care workers and involving them in the follow-up process.
- *Constipation*: The reality in the follow-up of patients with ARM is that the lower the defect, the higher the chance of constipation. This problem must be foreseen by the surgeon and explained to the family. Before labeling the postoperative patient as being "constipated," a careful rectal examination must be done to rule out the possibility of anal stenosis (which would mandate dilatation) or associated presacral mass (which would involve exci-

Figure 2 Fecal passage via urethra in child with ARM. (Color version of figure is available online.)

sion). True postoperative constipation (unrelated to stenosis) may in great part be managed with long-term dietary measures or bowel management (such as bowel washouts), or a combination thereof. In low-income countries where food choices and alternatives are limited, a region-specific list of constipating foods to be avoided and non–constipating foods to be suggested may be put together.

Case 2

A 2-year-old boy was brought to a specialized pediatric surgery unit for "closure of colostomy": the latter often entails the "definitive surgery" as well. According to the history, he was born without an anus, and liquid feces began passing through the penis, albeit with great difficulty. A colostomy was done at 1 month of age in a district hospital. He returned to that hospital on several occasions during the first year of life with gastroenteritis and dehydration. The parents were not able to bring him for the definitive surgery until now. On examination the child appeared well: he had a supraumbilical loop colostomy, a large palpable infraumbilical mass, midshaft hypospadias, and no obvious anal dimple. A distal colostogram revealed that the sigmoid colon was tethered in the right upper quadrant: the colostomy had been unknowingly created in the sigmoid colon, but matured in the upper abdomen. A colostogram was performed with pressure injection of contrast: the anomaly appeared to be high but no fistula was seen. An abdominal ultrasound confirmed that the large mass was a fecaloma, but failed to locate the left kidney. A Posterior Sagittal AnoRectoplasty (PSARP) was performed with the guidance of an anesthesia nerve stimulator to help define the muscle complex. A rectoprostatic urethral fistula was demonstrated and ligated. The distal bowel was tethered by the colostomy and, to obtain the length necessary to carry out the anorectoplasty, the colostomy was taken down and resited to the left lower quadrant. Removal of the fecaloma and copious irrigation of the distal bowel was also done on-table. The anoplasty was intentionally made longer than usual to accommodate the larger rectal circumference. Postoperative dilatations were started at 10 days using a homemade dilator. Four weeks later, dilatations were done using the mother's finger. The child did well but a year later still had not gained fecal continence. A constipating diet of plantains and other local foods was started, but the child was then lost to follow-up.

Points of discussion

- *Two-year-old boy*: Fairly typical age for re-presentation after creation of colostomy in low-income settings.
- *Passing feces through penis*: As unlikely as this seems, some boys with imperforate anus and no colostomy will be able to survive for months by stooling through the urethra, without contracting urinary tract infections. The fact that they are exclusively breastfed may help by softening the stool (Figure 2).
- *Gastroenteritis and dehydration*: In the infant, a proximal colostomy or ileostomy can carry a significant morbidity (and even mortality) due to uncompensated fluid losses. Because of difficult access to care in low-income countries, time to reversal of stoma should be minimized in these patients. Infants are particularly vulnerable to fluid and electrolyte imbalance: the parents of these patients with stomas must be urged to come for evaluation and care in all cases of diarrhea.
- *Transverse colostomy*: Unfortunately, transverse colostomies are still frequently done for patients with ARM. These stomas are too far removed from the rectum to allow proper investigation and irrigation: significant fecal impaction may occur if there is spill-over into the distal limb.
- *Divided* vs. *loop colostomy with large infraumbilical mass*: This fecaloma is likely because the loop stoma was not completely diverting (Figure 3). A properly created

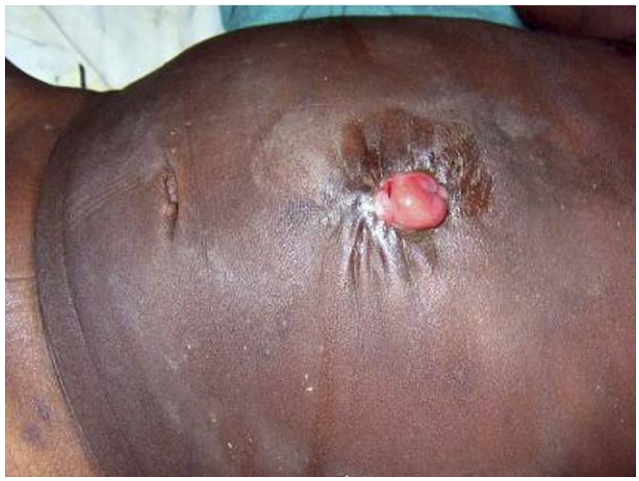

Figure 3 Stenosed loop colostomy. (Color version of figure is available online.)

Figure 4 Anesthesia nerve stimulators. (Color version of figure is available online.)

divided stoma has the greatest probability of achieving complete diversion of the fecal stream. The fecaloma may also be attributable in part to not having carried out distal washouts at the time of colostomy creation. Assessing the need for repeated, diligent distal bowel washouts can be a key factor in improving the results of the anorectoplasty. Though rare, a hydrocolpos should figure in the differential diagnosis of such female patients.

- *Hypospadias*: genitourinary (GU) abnormalities are the most common associated anomalies in patients with ARM. The presence of 1 GU anomaly should alert the surgeon to the possibility of others.
- *No obvious dimple*: This may hint to a higher ARM, in keeping with the associated GU anomalies.
- *Distal colostogram*: An essential investigation for patients with ARM. It is available in many low- and middle-income settings. The limiting factors include availability of fluoroscopy and the appropriate contrast agent. With regard to lack of fluoroscopy, a series of plain abdominal x-rays can be obtained with injection of the contrast agent. Judicious amounts of pressure should be used, ideally via a Foley catheter with the balloon inflated in the distal loop of bowel. This is best carried out by the treating surgeon as significant judgment must be exercised regarding inflation pressures. The treating surgeon should inject contrast agent until appropriate resistance is met: multiple films are then taken. Regarding the contrast agent, one should try to use water-soluble contrast and to avoid barium. Even if the latter is universally available and cheaper, barium has a strong tendency to inspissate and contribute to fecaloma formation, even when diluted. Moreover, if there is any fistula, barium it will pass into the urinary tract. Most forms of water-soluble contrast material (as long as they are not the older hyperosmolar solutions) are preferable. If only barium is available, intensive washouts are required immediately after the study.
- *Tethered sigmoid colon*: A classic surgical error. Accidentally pulling a large redundant sigmoid loop instead of the transverse colon is a common mistake. This can be avoided by using generous incisions, which allow clear identification of the bowel being brought out. The attachment of the sigmoid to the left retroperitoneum is a key anatomic identifier that the surgeon should look for. Transverse colostomies should be avoided in patients with ARM. The ideal colostomy remains a completely divided proximal sigmoid colostomy (at the junction of the descending and sigmoid segments of colon). The distal limb, which consists of sigmoid and rectum, provides more than enough length to obviate any concerns about "tethering."
- *No fistula is seen*: This is a common occurrence in low-income settings, primarily due to the long intervals between creation of colostomy and colostogram. Even with properly performed distal colostograms, the fistula may be plugged with inspissated stool and/or debris. As mentioned earlier, not exerting enough pressure during injection of contrast may also contribute to the inability to demonstrate the fistula.
- *Absent kidney*: The GU tract should be imaged whenever possible, but particularly in patients with higher ARM. Ultrasound examination is accessible in resource-limited settings and should be used as part of the initial assessment.
- *Anal mapping*: An essential component in the management of patients with ARM, especially in higher forms where the sphincter location is not obvious on inspection. Lack of availability of such instrumentation is a significant limiting factor in resource-limited settings. Typical replacements for the popular Peña muscle stimulator are the inexpensive anesthesia nerve stimulators that can be attached via alligator clamps to needles or to a bipolar forceps (Figure 4). There are also different versions of the Peña stimulators made in other countries. A homemade, very low-cost version of the Peña stimulator also exists (using a BIC pen—Figure 5). The regular electrocautery device may also be used if the sphincter complex is sufficiently well developed.
- *Resiting of the colostomy*: Though having to resite or refashion a colostomy is unfortunate, cutting corners

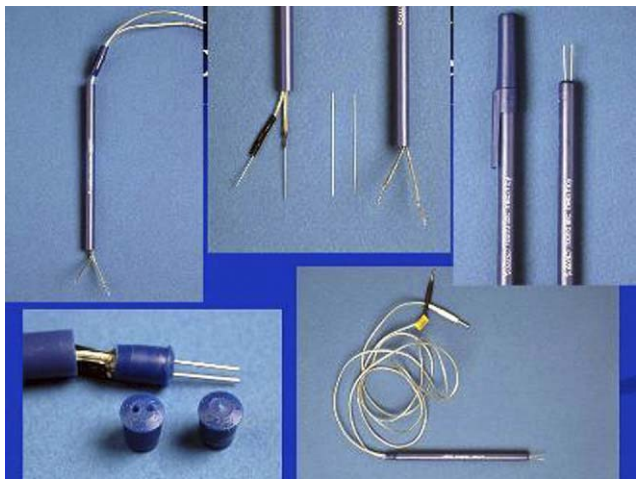

Figure 5 Homemade "Pena" stimulator (courtesy of Prof J-M Laberge, Montreal). (Color version of figure is available online.)

should not be entertained as the margin for error is much smaller in these environments.

- *On-table lavage*: Essential step in successfully managing patients with ARM in these environments. The fecalomas forming after months or years of poor fecal diversion are refractory to less aggressive measures and often require repeated attempts at evacuation under general anesthesia. The dilated bowel containing the fecaloma will return to normal or near-normal size if properly decompressed over a 6- to 12-month period. A further option at operation is to leave the lavage catheter in situ when the patient is turned prone (especially if a colostogram is not available). The catheter can then be manipulated during the PSARP to help the surgeon identify the rectum.
- *Large anoplasty*: Not only does it accommodate the larger rectum of older, poorly diverted children, but it has the added advantages of not requiring as aggressive postoperative dilatation and being less prone to stenosis. Stenosis is a major risk in settings where follow-up is notoriously challenging.
- *Homemade dilator*: Hegar dilators are the "gold standard" for anal dilatation, but as with gold they may be unaffordable, especially for giving to patients to take home. Alternatives include appropriately sized candles, spigots from Foley catheters, rounded ball pen ends, and, most importantly, the guardian's fingers. Any of the former can be calibrated and used until the child can be "graduated" to the guardian's fingers. Proper instruction with direct observation is critical to ensure that the correct orifice is being dilated adequately and deeply enough (width and depth).
- *Fecal incontinence*: A child with a high ARM has a significant chance of incontinence in the best of circumstances. Soiling is socially unacceptable in all societies, and the stigma in school-age children is great regardless of the environment. Unfortunately, Malone-type enema procedures are not readily accepted and the necessary time and equipment make them a poor option. Often, dietary methods are the only alternative. It may even be suggested that a permanent colostomy is preferable to a "perineal stoma" (though this may not be the most appropriate choice from a cultural standpoint).
- *Lost to follow-up*: A very common situation, significantly affecting our ability to care for children with ARM, no matter what the environment (low-, middle-, or high-income countries). Some of the major issues regarding lack of follow-up, such as transport costs, can and should be addressed through advocacy by the treating health care professionals.

Case 3

An 8-year-old girl presented to the pediatric surgical unit with the complaint of stooling through the vagina. The problem had been noticed by the mother soon after birth, but funds were not available to allow the child to be taken to a health care facility. The family neglected the problem. The girl seemed fairly continent of stool. She did however develop a strong tendency for constipation and overflow diarrhea. Examination revealed a healthy girl with a standard fourchette fistula. A divided sigmoid colostomy was performed, note being made of a very dilated stool-filled sigmoid. Tap water irrigations via the mucous fistula were performed, followed by a standard PSARP. Postoperatively, some liquid stool was noted to seep from the anoplasty, likely from retained inspissated stool in the distal limb. By postoperative day 5, perineal dehiscence was observed. This was managed conservatively with sitz baths, and eventual discharge home after teaching digital anal dilatation. The patient missed the designated follow-up appointment and returned only 2 months later. At this point the anus had returned to the posterior fourchette position: evidently, the mother had been dilating the vagina (not the anus) on the few occasions when she managed to overpower her unwilling daughter. The child was scheduled for re-do PSARP after a week-long course of colostomy irrigations with dilute gastrografin in water. A wide anoplasty was created to avoid the need for dilatations, and the postoperative course was uneventful. The child was well at 1-month follow-up. She failed to return for further appointments.

Points of discussion

- *Eight years old*: In low-income settings, late presentations (up into the teens and even twenties!) are not uncommon, especially in female patients. Genital areas are extremely private, and if the problem is not addressed by the mother during the child's infancy, it often remains hidden until adolescence.
- *Fairly continent*: Surprisingly, most girls with rectovestibular fistulas are fecally continent. However, the small diameter of the fistula predisposes to constipation, with its associated symptom of overflow diarrhea.
- *Colostomy was performed*: In high-income countries, selected female patients with ARM may undergo definitive repairs without colostomy: this is unwise in low-income settings. The children are almost never seen in the newborn period, only much later—by which time the sigmoid has become grossly distended with fecal matter. Moreover, the children are often malnourished when first seen, and proceeding directly with a PSARP may lead to catastrophic results. Finally, keeping the patient *nil per os* (NPO) for a week to 10 days in environments where total parenteral nutrition is not available (to minimize fecal contamination of the fresh anoplasty) is impractical.
- *Stool-filled sigmoid*: This constitutes one of the most difficult problems when managing children with ARM in low-income settings. The problem must be addressed aggressively: a few postoperative enemas are unlikely to be effective. Even when the effluent is clear, there can remain an inspissated fecaloma, sometimes to the point of calcification, which must be manually broken down, dissolved, or removed. When available, dilute gastrografin may be administered between disimpactions.
- *Perineal dehiscence*: This may be a common occurrence, often due to early fecal contamination of the wound. If

persistent contamination and deep sepsis can be controlled, the wound slowly closes—but often with anterior migration of the anoplasty back toward its original position. HIV infection should be considered if recurrent wound breakdown takes place or if repeated wound sepsis is encountered.
- *Dilating the vagina*: Much attention must be devoted to good training of the parents, particularly when there is a language barrier.
- *Child's strong opposition*: School-age children are much more difficult to dilate, especially in single-parent homes or when no other help is available. Performing a wider anoplasty, which is less prone to stenosis, even without regular dilatations, is a good option in this setting.

Management of HD in resource-limited settings

Case 1

A 3-day-old boy presented to the pediatric ward in the teaching hospital with a history of not having passed meconium since birth. Two days later he was referred to the pediatric surgical team with signs of peritonitis. He had a normal-looking anus and rectal examination revealed only a small amount of meconium. At laparotomy he was found to have meconium peritonitis with a cecal perforation. There was a transition zone in the descending colon and the diagnosis of HD was made. The cecal perforation was oversewn, the abdominal cavity was washed out, and the child was given an end colostomy on the proximal transverse colon. The distal end was closed.

Points of discussion

- *Three days old*: Unusually early presentation as most cases of HD in resource-limited settings present in infancy or childhood. In retrospect, symptoms will nearly always have been present from birth.
- *Cecal perforation*: In the absence of a colonic or anorectal atresia, this is diagnostic for HD. The perforation site can be safely closed.
- *Colostomy*: In neonates with HD, the distal colon is empty and its proximal end can safely be closed. Whether to do a mucous fistula or not is left to the discretion of the operating surgeon and will depend on the specific context.
- *Minimal surgery*: These neonates are usually very sick and there is limited anesthetic and intensive care unit support; delays must be avoided and surgery restricted to the urgent problem.
- *Biopsies*: Biopsies may be taken during laparotomy for confirmation of HD. However, circumstances may be such that it is unwise to prolong the operation and it may be better to postpone diagnostics until later.

Case 2

A 9-year-old boy was referred from a nearby district hospital. He had been admitted there with abdominal distension and dyspnea. He was noted to be chronically ill-looking. The referring diagnosis was ascites of unknown origin. From his history it became clear that he had long-standing complaints of intermittent constipation and diarrhea. He had been seen at the same district hospital 2 years earlier with distension and abdominal pain and had been treated for schistosomiasis, worms, and abdominal tuberculosis. These treatments had not affected his symptoms. He was the sixth born of 7 children; his mother's third child had also required a colostomy for abdominal distension but had subsequently died.

On examination he was found to have a hugely distended abdomen (Figure 6). Plain abdominal x-ray confirmed massive distension of the colon with extensive fecal loading (Figure 7). A clinical diagnosis of HD was made and a divided colostomy created on the very distended transverse colon (Figure 8). He subsequently required repeated admissions for treatment of a prolapsed colostomy and for washout of the distal colon to remove a large fecaloma. Rectal biopsy and barium enema confirmed the diagnosis (Figure 9).

Eventually, 3 years after presentation, he underwent a transanal endorectal pull-through procedure. Three months later his colostomy was closed. He has been well since.

Points of discussion

- *Late diagnosis*: The diagnosis of HD is often not considered early on and patients may even be seen as adults at

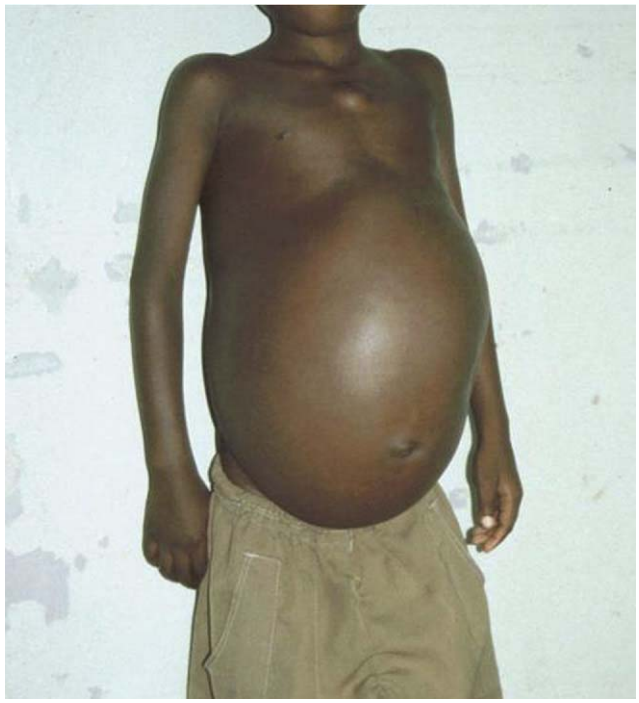

Figure 6 Massive abdominal distension with delayed diagnosis of Hirschsprung's disease. (Color version of figure is available online.)

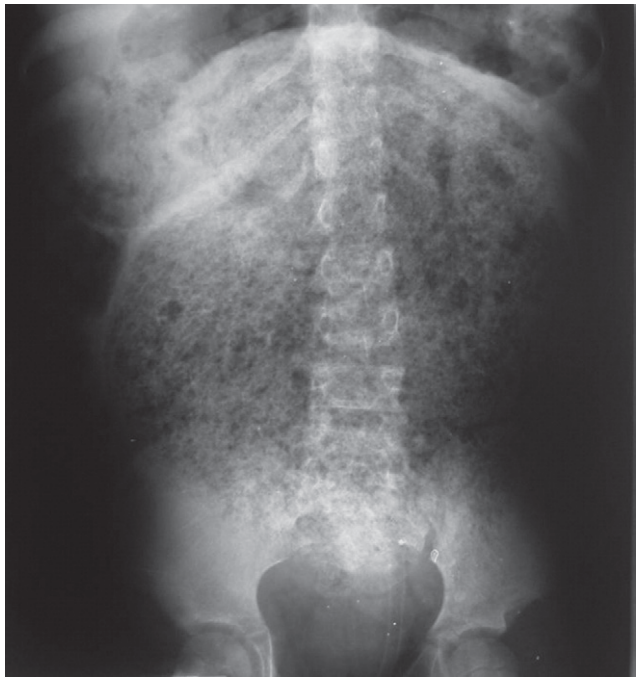

Figure 7 Plain abdominal radiograph showing massive fecal loading.

initial presentation. These patients are always malnourished, sometimes severely so, and this together with the extreme abdominal distension may result in any number of erroneous diagnoses.

- *Abdominal x-ray*: This is acceptably inexpensive and available in many district hospitals. The combination of extreme gaseous distension, hugely dilated loops of large bowel, and massive fecal loading of the colon is diagnostic for HD. Occasionally, there is a clear transition zone below which there is no air in the distal colon. This may provide a clue to the level of the transition zone.
- *Differential diagnosis*: Colonic distension can be excessive and should be differentiated from conditions such as hollow visceral myopathy. The latter is diagnosed by the presence of ganglion cells and abnormal muscle histology (this is a difficult diagnosis and requires sophisticated diagnostic equipment).
- *Colostomy*: If a transverse upper abdominal incision is used, the distended transverse colon can be divided and the distal and proximal ends sutured into either end of the abdominal incision. It is imperative to have access to the distal, fecally affected, colon for subsequent colonic irrigation. This is necessary if the distended colon is to reassume near-normal proportions.
- *Fecaloma*: The retained feces in the distal colon can be very difficult to remove, especially after a barium enema (see later in the text). To do this successfully requires dedication and application from the nursing staff, often supplemented by separate bowel washouts in the operating room and, if necessary, on-table lavage. If this is not diligently and thoroughly done, the colon will remain distended and the pull-through operation will be unnecessarily difficult.
- *Rectal biopsy*: The resources to obtain and interpret suction rectal biopsy specimens are not usually available, and the only option is a full-thickness posterior rectal biopsy carried out under a general anesthetic. An adequate quantity of the bowel muscle wall must be included for evaluation.
- *Barium enema*: It is unfortunate that other, more expensive, water-soluble contrast materials are, as a rule, not available. Inspissated barium contributes significantly to a fecaloma. In many hospitals, fully trained radiologists are not available and the quality of the contrast study is

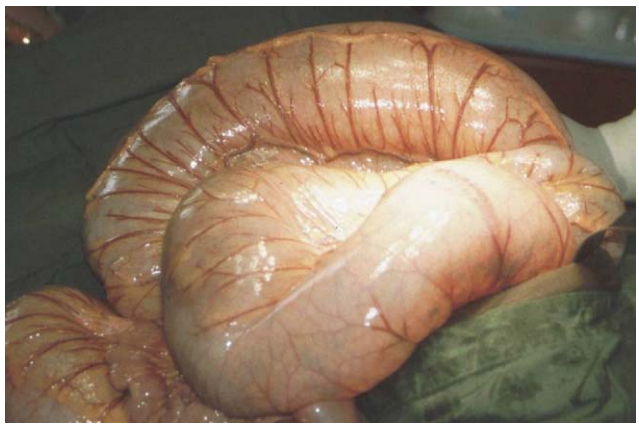

Figure 8 Delayed diagnosis of Hirschsprung's disease operative finding; massively distended proximal colon. (Color version of figure is available online.)

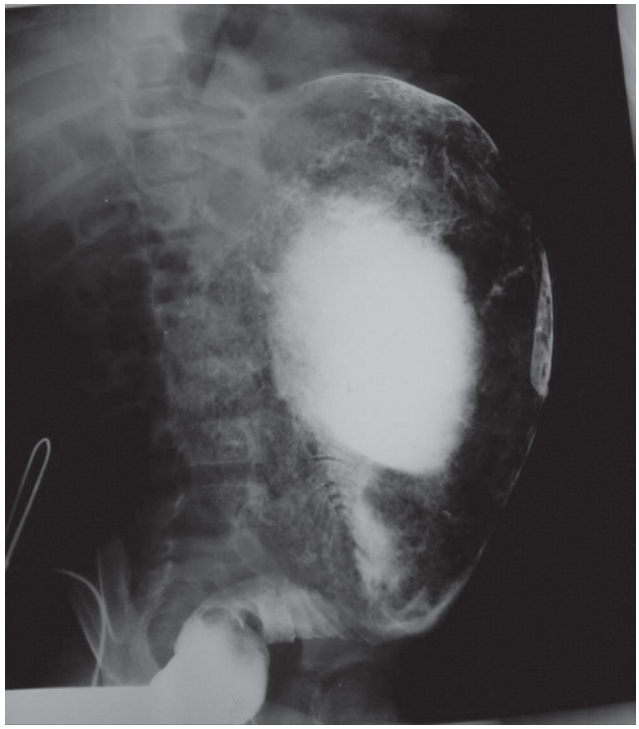

Figure 9 Massive proximal colon distension on contrast enema in a child with Hirschsprung's disease.

compromised. As a result it is not often possible to obtain a reliable indication of the level of the affected segment.

- *Transition zone*: Late presentation of long segment HD is rare. The transition zone in most patients who present late is in the rectosigmoid.
- *Primary pull-through*: The use of a primary procedure in the malnourished, late-presenting patient with an extremely dilated proximal colon is not advisable. Once the colostomy is performed, the distal colon requires 3 to 12 months to adequately reduce in size for a successful pull-through.
- *Transanal pull-through procedures*: In view of the usually limited length of the affected segment, a transanal pull-through is feasible if the local expertise exists. In the longer segment cases, a transanal pull-through may be combined with a laparotomy. Usually, the residual fecaloma in dilated bowel is a good clinical indication that the transition zone has been reached. When there is doubt about the level of the transition zone and histologic confirmation is not available, the bowel at the level of a well-functioning colostomy may be used for the pull-through. Histologic confirmation remains the gold standard, but may not always be available.

Case 3

A 12-year-old girl was examined at a district hospital with a 2-week history of constipation and abdominal distension. She had previously passed some worms in her stools. On admission at the central hospital, she was noted to be dehydrated and to have massive abdominal distension. Distended bowel loops and fluid levels were seen on the abdominal x-ray. The pediatric working diagnosis was ascites with an underlying intra-abdominal tumor, possibly Burkitt's lymphoma. Pediatric surgical evaluation resulted in the new diagnosis of megacolon secondary to HD. At laparotomy, the patient was found to have a volvulus of the sigmoid colon (Figure 10), which was subsequently untwisted and deflated. A transition zone was noted in the lower sigmoid and a divided transverse colostomy was fashioned. A full-thickness rectal biopsy was carried out 3 months later but the results were never obtained. Six months after presentation, a repeat rectal biopsy finally confirmed the diagnosis of HD. One year after the original admission, a transanal endorectal pull-through procedure was carried out. The rectum and distal sigmoid colon were resected to a level well above the peritoneal reflection. Subsequent histologic examination suggested the presence of ganglion cells in both the distal and proximal ends of the second specimen. The patient subsequently required repeated dilatations of a stricture. This eventually resolved and the colostomy was closed approximately 2 years after the patient had originally presented. At the most recent review the patient still has some problems with constipation and remains undernourished and small for her age; at 14 she has not yet reached menarche.

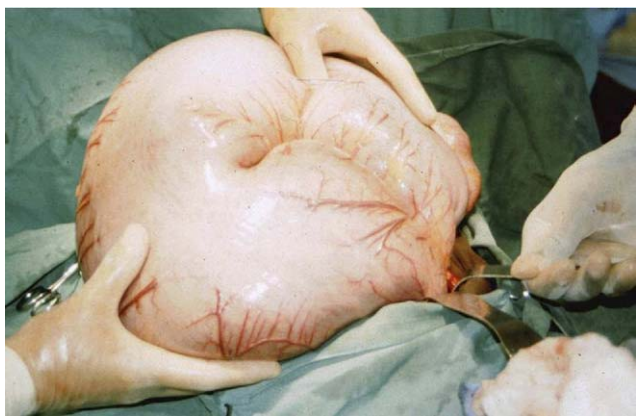

Figure 10 Sigmoid colon volvulus in a child with Hirschsprung's disease. (Color version of figure is available online.)

Points of discussion

- *Diagnosis*: Obtaining a good and reliable history can be a problem. In a setting with a high prevalence of infectious diseases, it is not surprising that the relatively uncommon presentation of HD is not considered.
- *Burkitt's lymphoma*: In some parts of Africa this is by far the most common cause of massive abdominal distension. However, with a good history and examination, this diagnosis should be easy to exclude.
- *Sigmoid volvulus*: In many parts of Africa, a redundant sigmoid colon predisposes to sigmoid volvulus, and this may be the most common cause of an acute abdomen in adults. In untreated HD, the dilated proximal colon may include the sigmoid and so predispose it to volvulus.
- *Biopsy*: A centralized pathology service, often at some distance from major hospitals, the lack of reliable patient registration systems and poor logistics all conspire against the successful reuniting of a patient and his/her biopsy result.
- *Histology*: Interpretation of biopsy specimens is very operator-dependent and in many low- and middle-income settings the necessary expertise may not be available.
- *Stricture*: An anastomotic stricture after endorectal pull-through procedures is a well-recognized complication. A rectal examination should be carried out at the first appropriate follow-up visit to diagnose/prevent postoperative stricture.
- *Malnutrition*: In countries where most children are malnourished to various degrees, the attendant malnutrition associated with HD is even more severe (particularly so in patients with a late diagnosis). It may take many years even after successful surgery before the growth indicators normalize.

Case 4

A 4-year-old boy is referred from a regional hospital. He had presented there with abdominal distension and constipation since birth. A clinical diagnosis of HD had been

made. With no further investigations, a subtotal colectomy with a low colorectal anastomosis was carried out as a primary procedure. Postoperatively, he developed renewed abdominal distension and, with a diagnosis of bowel obstruction, he was referred to the Pediatric Surgical unit of the nearby teaching hospital. Examination revealed a chronically ill and wasted boy with obvious bowel obstruction. At laparotomy a divided transverse colostomy was performed to relieve the obstruction. He was given nutritional support with some effect and was discharged home. He was readmitted 4 months later, still in a very poor general condition. On rectal examination, the colorectal anastomosis was found to be completely occluded. The stoma was working well. In view of his persistently poor nutritional status, he was tested for HIV and found to be positive. He was commenced on cotrimoxazole prophylaxis. A year and a half after presentation he finally underwent a CD4 count, which was 41.6%. At the age of 6 years he weighed 13 kg. Presently, he is still waiting to be commenced on antiretroviral (ARV) therapy. This is considered essential to improve his general condition so that he might eventually be considered for a pull-through operation.

Points of discussion

- *Clinical diagnosis*: The difficulty with access to histologic diagnosis may result in surgery being undertaken based only on the clinical evaluation.
- *Inappropriate surgery*: Trained pediatric surgeons are often limited to the teaching hospitals. General surgeons in smaller hospitals may feel compelled to treat this condition. This may result in inappropriate operations and/or suboptimal outcomes. The resultant morbidity may be significant and require repeated surgical procedures. Colostomy however can be life-saving, and should be performed by general surgeons when pediatric surgeons are not available.
- *Immunosuppression*: In countries with high serum prevalence of HIV, it is inevitable that many patients with HD will be found to be immunocompromised. This, together with the already present nutritional depletion, may lead to considerable delays in treatment. The current recommendation is that all children found to be HIV-infected be started on ARV treatment as soon as possible.

Management of acquired rectovaginal fistula associated with HIV in resource-limited settings

Case 1

A 4-month-old infant was referred with the history of passing stool per vagina (Figure 11). She was born to an HIV-positive mother and has herself been confirmed to be HIV positive. A colostomy was fashioned before contemplating definitive surgical repair of the fistula. No ARV treatment was available. The colostomy was complicated by prolapse and required revision. The fistula was finally repaired at 1 year of age. Follow-up 3 months later demonstrated a recurrent fistula. During this time ARV treatment became available but could only be obtained with approval from the national Infectious Diseases Control Committee (due to the high cost). Because of her severe immunocompromise (CDC category III), the committee finally granted approval. Two months after beginning ARV therapy, the re-do fistula and sphincter repair was performed. The colostomy was closed 4 months later. Histology of the specimen showed squamous epithelium with mild chronic inflammation. There was no evidence of granulomas, organisms, or neoplasia. Examination under anesthesia during colostomy closure showed a well-healed perineum. She is currently 8 years old. She requires a bowel regimen to address fecal loading with associated soiling.

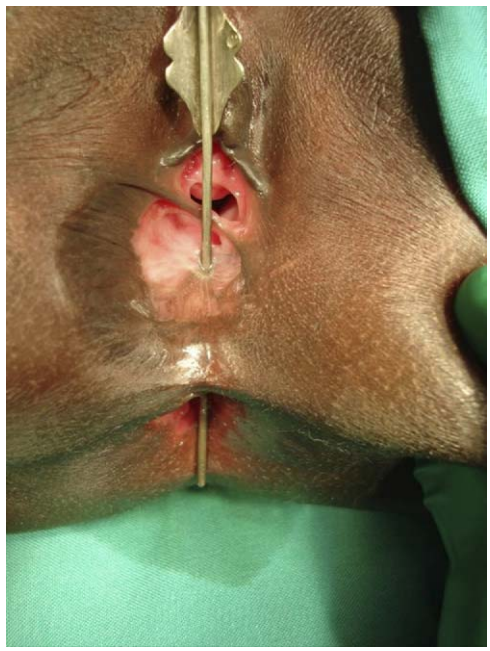

Figure 11 Acquired rectovaginal fistula in an HIV-positive child. (Color version of figure is available online.)

Points of discussion

- *Acquired fistula*: Acquired rectovaginal fistulae during the first year of life are well-recognized in HIV-positive children. In fact, an acquired rectovaginal fistula is usually diagnostic of HIV infection.[1] These patients usually present with a history of passing stool per vagina. They may also have multiple perineal fistulae.
- *Mother-to-child transmission*: Mother-to-child transmission can be seen in up to 30% of children born to HIV-positive mothers. Currently, 5.5 million people are living with HIV in South Africa alone. Almost 1 in 3 pregnant women were found to be HIV positive in 2004.
- *Availability of ARVs*: Access to ARV treatment may be difficult due to government policies and cost implica-

tions. Without ARV treatment death often occurs before the child is 1 year of age.

- In the past, treatment of this fistula was expectant. The current guidelines indicate that all HIV-positive children should be started on ARV therapy at the time that the HIV infection is diagnosed. Suggested treatment includes standard triple combination therapy (usually 2 nucleoside reverse transcriptase inhibitors plus either a protease inhibitor or a non-nucleoside reverse transcriptase inhibitor). Spontaneous closure of the fistula after initiation of ARV treatment is well described.[2]
- *Timing of surgery*: ARV medication should be given at least 4 to 6 weeks before the definitive treatment. Improved CD4 count, viral count as well as nutritional status should be seen preoperatively.
- *Surgical technique includes division of fistula and identification of the muscle complex*. The posterior wall of the vagina is reconstructed followed by creation of the perineal body. The muscle complex is identified and repaired before closure of rectal wall.
- *Covering colostomy*: This is required prior to, or at the time of, the fistula repair.

Conclusion

Caring for pediatric patients with colorectal pathology is a challenge even in the best equipped environment. These case presentations of patients with ARM, HD, and acquired rectovaginal fistulae (HIV-positive patients), managed in the context of limited resources, highlight some of the major issues surrounding diagnosis, management, and ultimate disposition. We recognize that when one combines the number of differing environments across low- and middle-income countries with the sheer variability of these disease entities, it is impossible to predict a single "best clinical practice" in the management of these patients. However, allowing for the fact that basic principles remain the same regardless of available resources, the questions then become: "how best to diagnose?", "how best to manage?", and "how best to follow?" in a given environment. Though the answers are specific to that environment, lessons learned in one context may be applicable in another.

The authors recognize the tremendous work done by those who look after these patients and hope to contribute in some small way to the clinical armamentarium available for their patient management. In environments with limited resources, children are the most vulnerable members of an already vulnerable society. We must therefore provide the highest standard of clinical care that the local environment permits.

References

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