Cloacal Exstrophy: A Unified Management Plan

By Samuel Z. Soffer, Nelson G. Rosen, Andrew R. Hong, Mihai Alexianu, and Alberto Peña
New Hyde Park, New York and Bronx, New York

Background/Purpose: The belief that patients with cloacal exstrophy have a short and therefore useless colon is all too common. Frequently, the colon is used for urinary or vaginal reconstruction, and the possibility of a pull-through is lost. In the authors' experience, the use of a unified management plan allowed most patients to undergo pull-through and avoid a permanent stoma.

Methods: Twenty-five patients were treated for cloacal exstrophy in the authors' institution from 1985 through 1999. In all patients, bladder closure, omphalocele repair, and creation of a colostomy were performed at birth. All available colon, no matter how small, was incorporated into the fecal stream. After at least 1 year, patients were assessed for the ability to form solid stool through their stoma. Normal colonic length, capacity to form solid stool, or success with a bowel management regimen through the stoma were considered indications for pull-through. Genitourinary reconstruction was contingent on the colorectal plan.

Results: Colonic length ranged from normal in 12 patients, 40 to 70 cm in 3 patients, 10 to 30 cm in 4 patients, and less than 10 cm in 2 patients. All 25 patients underwent pull-through. Three are totally continent, 4 are continent with occasional soiling, 11 remain clean with a bowel management regimen, and 4 are too young to assess. One patient was clean, but now refuses bowel management. Two early patients, both with less than 10 cm of colon, now have ileostomies.

Conclusions: During neonatal repair, a colostomy should be formed incorporating all pieces of colon, no matter how small. With time, most patients will be able to form solid stool, and a pull-through should be undertaken if that ability exists. Decisions regarding genitourinary reconstruction should be made only after the gastrointestinal plan is established to achieve the optimal use of available bowel.

INDEX WORDS: Cloaca, exstrophy, colonic pull-through, bowel management.

CLOACAL EXSTROPHY is a devastating malformation that occurs with an incidence ranging from 1 in 200,000 to 400,000 births.[1,2] As of 1976, there were under 200 reported cases in the literature.[3] From 1960 when Rickham[4] reported the first successful repair until 1976 the mortality rate in surgically treated patients was still about 50% with only 1 report of an untreated survivor.[5] These discouraging results improved dramatically in the 1980s with survival rates of 83% to 100% reported in several series.[1,6] Despite this recent improvement in survival rate, the challenge remains to improve the poor quality of life of these unfortunate children.

Primary fecal or urinary continence after repair is uncommon.[7,8] The majority of treated patients live with a permanent fecal stoma, and most require a continent diversion and intermittent catheterization to manage their urine. It is clear that there remains substantial room for improvement in the management of these defects.

Because this defect affects the gastrointestinal tract as well as the genitourinary tract, these patients frequently are treated by at least 2 groups of pediatric surgical specialists, general pediatric surgeons, and pediatric urologists. Both groups frequently work independently without a specific master plan in mind. A common misconception when describing cloacal exstrophy patients is the assumption that they are born with minimal or no colon.[9] Although this assertion often is true, we have found that a wide spectrum of defects is represented in these patients, from those with minimal or no colon to those with normal colon. Therefore, the treatment plan must be individualized.

Most investigators agree that the initial approach should include repair of the omphalocele, closure of the bladder with or without osteotomy, and creation of a fecal diversion.[1,6] Almost all of these patients will need some form of urinary and vaginal reconstruction at a later date, often requiring the use of intestine. However, the length of colon and the capacity to form solid stool are critical factors in deciding whether a patient is a candidate for a pull-through. When the decision to use bowel to recon-

From the Division of Pediatric Surgery, Department of Surgery, Schneider Children's Hospital–Long Island Jewish Medical Center, New Hyde Park, NY, and The Clinical Campus of the Albert Einstein College of Medicine, Bronx, NY.

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Address reprint requests to Alberto Peña, MD, Chief, Division of Pediatric Surgery, CH-158, Schneider Children's Hospital, 269-01 76th Ave, New Hyde Park, NY 11004.

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Table 1. Patient Data

Diagnosis	No. of Patients		
	Male	Female	Total
Classic exstrophy	5	6	11
Covered exstrophy	0	9	9
Variant exstrophy	1	4	5
Total	6	19	25

struct a component of the genitourinary system is made independently by a pediatric urologist without the participation of a pediatric surgeon the possibility of undergoing a colonic pull-through frequently is lost. We present a unified management plan that first attempts to determine whether the patient is a candidate for a pull-through before the genitourinary reconstruction is considered. This decision is made by the pediatric surgeon and is based on the potential the patient has for fecal continence or successful bowel management, either of which will leave the patient clean after the repair.

Most pediatric surgeons believe that this decision should be based on the quality of the sphincter mechanism and the integrity of the spine and sacrum. We wish to convey the message that the decision should rather be based on the capacity to form solid stool, which depends primarily on the length of the patient's colon. The presence of a good anal sphincter and a normal sacrum, which is very unusual, should not necessarily be a universal indication for pull-through because a patient with these findings and liquid stools will still be fecally incontinent.

Conversely, a patient with very poor sphincters and a dysplastic sacrum, with or without a meningocele, may be a candidate for a pull-through provided they have enough colon to form solid stool. This last characteristic will allow them to remain clean with a bowel management regimen in the likely event that they do not achieve primary fecal continence. If it is decided that a patient with marginal colonic length will undergo a pull-through, the pediatric urologist should plan the genitourinary reconstruction with the assumption that colon should not be used. If it is decided that a patient will require a permanent fecal stoma, the pediatric urologist then is free to use colon or small bowel for the reconstruction.

MATERIALS AND METHODS

The records of 25 patients with cloacal exstrophy born between 1976 and 1999 were reviewed retrospectively. Twenty-three were born at other centers, underwent their primary operations during the neonatal period elsewhere, and were referred to our institution for further reconstruction at ages ranging from 1 to 15 years. Two patients were born in our institution between 1985 and 1999. Eleven patients were born with classic cloacal exstrophy, 9 with covered cloacal exstrophy, and 5 with variant forms (Table 1). All children appear to possess normal intelligence.

Initial Management

Twenty-one patients had colostomies formed at birth, and 4 patients had ileostomies. An omphalocele was present in 19 patients and was closed successfully at the initial procedure in all but 1 who required reoperation to close a large abdominal wall defect. Colonic length was measured in 21 of 25 patients (Table 2) and ranged from only 7 cm in one patient to near-normal length in 12 patients. Normal length of colon was defined as a colon containing all segments including cecum, ascending, transverse, descending, and rectosigmoid colon. Near-normal colon was defined as one that has all of these segments but missing one portion.

The extrophied bladder or hemibladders were closed successfully at the first operation in all but 3 patients who required additional procedures to close the bladder. Nineteen patients were genetic girls, and 6 were genetic boys. Five of 6 genetic boys had bilateral orchiectomies at birth at which time they were assigned the female gender.

Gastrointestinal Management

All 25 patients underwent colonic pull-through procedures. Twenty-two of the 25 children were between the ages of 1 and 3 at the time of the reconstruction. The first 8 patients were all operated on via a posterior sagittal approach or a combined posterior and anterior sagittal approach. The remaining 17 patients underwent surgery using an anterior sagittal approach only. Before any procedure, a colostogram was performed to ascertain colonic length. In addition, the family was questioned about the child's ability to form solid stool.

Six of the 25 patients underwent a bowel management program preoperatively through the stoma to see if they could form solid stool and remain completely free of stool for 24 hours. Four children were found to be able to form solid stool when receiving a constipating diet and antidiarrheal medication; subsequently, enemas delivered through the stoma kept these patients completely clean for over 24 hours. The other 2 children had ileostomies and defunctionalized portions of colon after the initial operation at birth. Both underwent laparotomy and a colon salvage procedure to incorporate all available colon into the fecal stream with the aim of allowing the colon to grow and develop its maximal water absorption capacity. Both of these children subsequently had the capacity to form solid stool and have undergone successful pull-through. Twenty of 25 patients had their stomas closed during the pull-through procedure, and 5 patients had them closed at a later date. Two of our earliest children required reoperation and formation of a permanent ileostomy because of continuous soiling.

Urinary Management

Nine bladder augmentations were performed using sigmoid colon in 4 cases, small bowel in 3, and stomach in 2. Mitrofanoff procedures, to create a continent catheterizable conduit to the bladder, were performed

Table 2. Colonic Length

Colonic Length	Number
Normal	12
60-69 cm	1
50-59 cm	1
40-49 cm	1
30-39 cm	0
20-29 cm	1
10-19 cm	3
Less than 10 cm	2
Total	21

NOTE. Accurate colonic length information only available for 21 patients.

on 12 patients utilizing a piece of ureter in 4 cases, tunneled bladder tissue in 3, the appendix in 3, and small bowel in 2 cases. Nine children are waiting to reach the age of urinary reconstruction.

Genital Management

Eighteen patients underwent genital reconstructive procedures. Four of the assigned girls have undergone creation of a vagina. Another patient, who recently underwent pull-through, is awaiting further genitourinary reconstruction. Our one genetic boy who has maintained the male gender was born with a normal penis and did not require any phallic reconstructive surgery.

Vaginal reconstruction for these genetically male patients was handled individually. In 3 patients the colon used for the pull-through was bisected in half, and the anterior portion was used to create a neovagina. In one child a vulva and vagina were fashioned from scrotal tissue. This patient currently is living with an ileostomy and will require creation of a functional vagina when she gets older.

Thirteen of the 19 genetic girls have undergone operations to create a functional vagina. Six of these patients required the creation of a neovagina with colon utilized in 3 cases, small bowel in 2, and the remnants of a large extrophied sac of native tissue in 1 case. In one patient, two hemivaginas were anastomosed to create one larger vagina. This patient required vaginoplasty 2 years later to improve her cosmetic appearance. The remaining 6 patients required smaller procedures including removal of vaginal septum, clitoroplasty, and vaginoplasty.

RESULTS

All 25 patients are alive and continue to undergo follow-up. Gastrointestinal results are summarized in Table 3. Three children are primarily fecally continent and do not soil. Two of these children were born with covered exstrophy, and 1 has a variant form in which no omphalocele was present. Four children are fecally continent but occasionally soil. Two children now live with permanent ileostomies. Both of these children were born with very short colons of 7 cm and 10 cm, respectively, and neither was ever able to form solid stool before their pull-throughs. Eleven patients are clean with the aid of a bowel management regimen. Four additional patients are candidates for bowel management, which has not yet been instituted because of their age. Another patient, who is now 18 years of age, remains incontinent of urine and feces and refuses to administer the necessary rectal enemas or creation of a stoma.

With respect to urinary continence (Table 4), only 1 child is primarily continent of urine. This child is our 1 genetic boy who maintained the male gender and represents one of our variant patients because he was born with bladder exstrophy and imperforate anus but with an intact pubic symphysis and a normal penis. The 12 patients who underwent Mitrofanoff procedures remain dry with intermittent catheterization. Two other children have undergone urinary reconstruction but are too young to assess.

Overall, our 25 patients have undergone 134 surgical procedures averaging more than 5 operations per child. We anticipate that most of these children will require additional procedures in the future.

Table 3. Gastrointestinal Results

Status	Number
Primarily continent	3
Continent with occasional soiling	4
Clean with bowel management	11
Incontinent	1
Permanent ileostomy	2
Too young to assess	4
Total	25

Table 4. Urinary Results

Status	Number
Continent	1
Incontinent	1
Dry with intermittent catheterization	12
Awaiting further reconstruction	9
Too young to assess	2
Total	25

DISCUSSION

Cloacal exstrophy represents a wide spectrum of disease that includes in the unfavorable side of the spectrum classic exstrophy of bladder, imperforate anus, prolapsed terminal ileum connected to the bladder, different lengths of colon, omphalocele, and a very abnormal spine. On the benign side of the spectrum is what has been termed *covered cloacal exstrophy* in which a patient has the bony and visceral features of cloacal exstrophy covered by dermis or an intact abdominal wall.[10,11] Although thought to be exceptionally rare, covered exstrophy was seen in nine of our 25 patients. Most people consider only the most severe type when discussing cloacal exstrophy. We included the covered exstrophy cases because they represent a similar technical challenge in terms of dealing with bowel control, urinary control, and sexual function.

With respect to fecal continence, most of these patients fall into the poor-prognosis group. Most of these children will have either poor sacrums or significant vertebral defects. In addition, most will have suboptimal anal sphincter and perineal muscle function. It is the rare cloacal exstrophy patient who will achieve primary fecal continence after repair.

However, despite the poor prognosis in most patients for primary continence, the capacity to form solid stool permits the use of a bowel management program whereby children should be able to remain fecally clean 24 hours a day and to become socially acceptable. The success of this program depends on the capacity to form solid stool, which is most closely related to the length of the colonic segment. The incapacity to form solid stool uniformly results in failure of bowel management. Therefore, every centimeter of colon is crucial, and we echo the prevailing wisdom that no colon should be sacrificed at the initial

operation.[12-15] In addition, utilizing as much colon as possible in creating the initial stoma helps greatly in managing these patients because of fewer electrolyte abnormalities.[13,16] Therefore, it is vital that every piece of hindgut, no matter how small, be incorporated into the fecal stream because it has been noted that these apparently useless small segments of colon will grow considerably with usage and time.[17] If a piece of colon remains defunctionalized and is not incorporated into the fecal stream it will atrophy, often rendering it useless for the purpose of reconstruction several years later.

In consideration of all of these factors, our approach to the repair differs from others in terms of the timing of the reconstruction (Fig 1). After the neonatal omphalocele repair, initial bladder closure, and creation of an end colostomy with all available colon we observe the patients for 1 or more years to allow for colon maturation and to determine the quality of stool they are able to produce. If a patient forms solid stool they are a candidate for a pull-through procedure even if they have a poor sacrum, vertebral defects, or suboptimal sphincter or perineal muscle function. This procedure is offered because a bowel management program can be instituted to keep these patients clean in the likely event they do not achieve primary fecal continence. We have found in our anorectal malformation population that the majority of children prefer this way of life as opposed to living with a stoma.[18]

In those children who did not form solid stool and had shortened colonic length on colostogram, a constipating diet was begun in an effort to see whether these children can be induced to form solid stool. Our bowel management protocol was then instituted, and enemas were administered directly through the stoma to ascertain if the child could remain clean for 24 hours. We learned that a patient should be considered a candidate for a colon

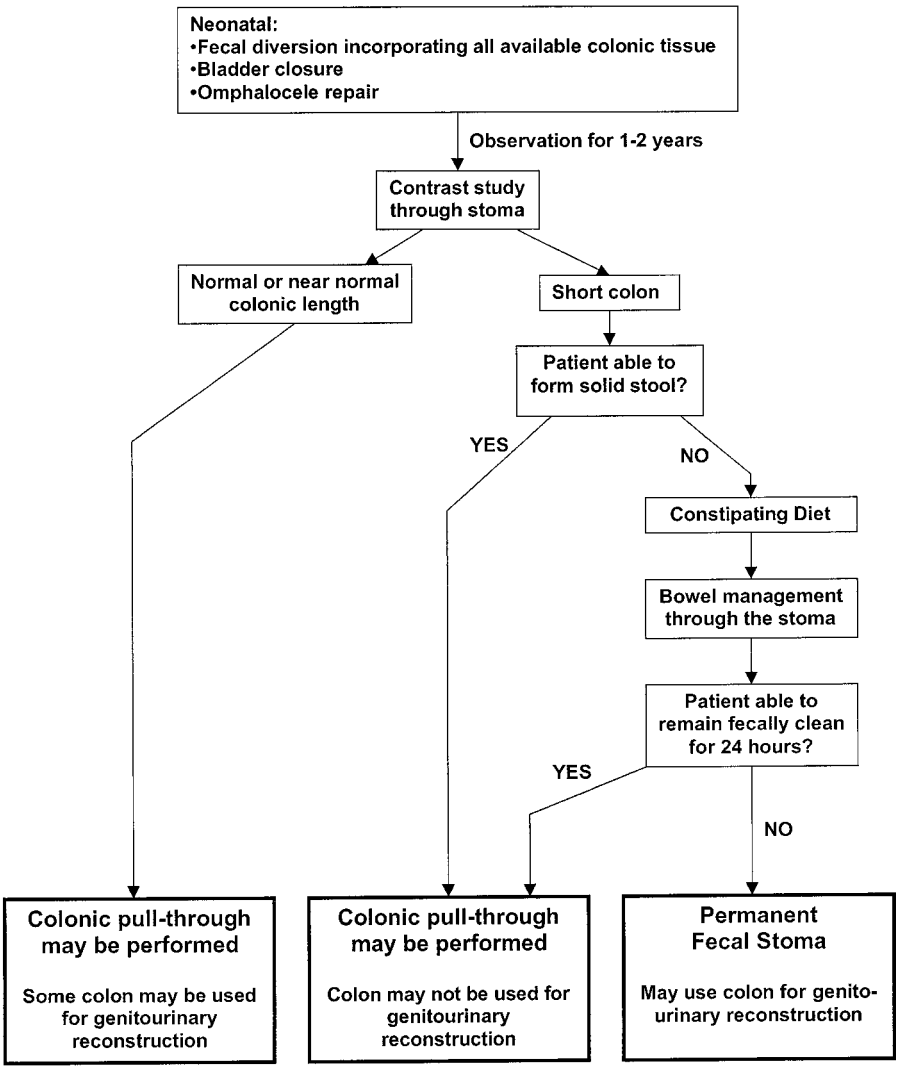

Fig 1. Algorithm for the management of cloacal exstrophy.

pull-through only when bowel management applied through the stoma was successful. Lack of response to this bowel management regimen now is considered a contraindication for a pull-through even with the presence of a normal sacrum and good sphincters since, in our experience, it is destined to fail.

We do not attempt extensive urologic reconstructions until the gastrointestinal plan is clarified. Thus, if a child has borderline colonic length we will not commit the use of any colon to reconstruct the genitourinary tract until bowel management has been instituted through the stoma. If this child can form solid stool, either primarily or with the aid of a constipating diet and loperamide, and can remain clean with bowel management, then it is vital that no colon be diverted for other uses. If any colon is utilized for other systems the child may lose the ability to form solid stool and the opportunity to undergo pull-through.

Alternatively, if the child's bowel management plan fails because of the inability to form solid stool, a life-long fecal stoma will be necessary anyway. In these cases, some colon may be used for urogenital purposes. In those children with normal or near-normal colonic lengths, the use of colon in the reconstruction of other systems becomes less of an issue. In some cases, the patient not only has normal colonic length, but also megasigmoid with hypomotility and constipation. In such cases we have used colon successfully for both bladder augmentation and vaginal reconstruction without detrimental effect on the child's postoperative bowel control.

Currently, all patients are approached anterior sagittally entering the abdomen and pelvis through the same incision between the pubic rami. All children born with cloacal exstrophy require extensive urologic reconstruction after the initial bladder closure.[19,20] Our goal for these patients is that they be able to achieve social continence through a program of intermittent catheterization. This requires a bladder of adequate size and compliance to serve as an effective reservoir as well as a conduit with a continence mechanism to prevent leakage.[21] Currently, for bladder augmentation, we prefer colon, small bowel, and stomach in that order.

Reconstruction of the genitalia is another challenging enterprise. One alternative in the treatment of genetically male patients is to leave the testes and raise the patient as a boy, which will require an orchiopexy and phallic reconstruction. Another option is to perform orchiectomy and raise the child as a girl with eventual creation of a vagina for sexual but not reproductive function and a lifetime of hormone therapy. In all cases in which the male gender is maintained the patients have phallic inadequacy as adults. Although in some cultures this phallic inadequacy is insignificant when compared with losing the male's societal role,[22] the reverse is more often true. In our series, 5 of 6 genetic boys were converted to the female gender with no attempts at phallic reconstruction. Some investigators have reported dismal results with attempted phallic reconstructions with 2 reports of suicide in reconstructed males thought to be owing to phallic inadequacy.[1,2] Most pediatric surgeons and pediatric urologists support the idea of raising genetically male patients as girls, but a trend of enthusiastic surgeons is emerging in favor of phallic reconstruction motivated by the unhappy experience of observing masculine behavior in patients raised as girls.

Our genetically female patients also require extensive vaginal reconstruction. In only 1 case were we able to form a single vagina from two hemivaginas, and this patient still required further vaginal reconstruction later. Usually the vaginas are too widely separated, and only 1 can be used.[6] Most of our patients will require further revision and reconstruction of their genitalia as they approach menarche and adulthood. None of our patients have reached the age to become sexually active.

Children who are born with cloacal exstrophy should be given every chance for the best quality of life possible. The opportunity to become an independent, stoma-free adult who is able to remain dry at all times with intermittent catheterization and fecally clean at all times with bowel management is within reach for many patients. We emphasize remaining clean with bowel management and not continence as a primary goal. Although continence may be achieved in a few patients, it currently is the exception and not the rule.

Cloacal exstrophy continues to test the limits of how much we as surgeons can do to repair what at first sight may seem hopeless. It is absolutely vital that a thorough plan be instituted from birth that takes into consideration all of the pertinent issues.[23] Often, circumstances will dictate necessary changes to the master plan. However, it would be unfortunate if a child loses reconstructive options because an earlier stage of the repair was planned poorly.

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Discussion

D. Farmer (San Francisco, CA): You did not make any comment about your approach to vaginal reconstruction in these patients.

S. Soffer (response): In the manuscript there is a lot more on the urinary reconstruction and on the vaginal reconstruction and how we would approach it. Given the time limits here, we were unable to talk about that.

In some cases in which there is normal colonic length, we were able to make a neovagina out of colon. And we did that in 3 cases. In one patient we were able to form a rudimentary vagina. And that patient is going to need further work later on. Because that was one of the patients that had marginal colonic length, we were afraid that if we took any piece of colon, that patient will no longer be manageable with bowel management. So that patient may require the use of small bowel and vaginal reconstruction later on.

C. Paidas (Baltimore, MD): I wonder if I could ask you about the status of the sphincters when you do these pull-throughs and the relationship of the sacral ratios in those patients with cloacal extrophy.

S. Soffer (response): The majority of these patients have poor sphincters and poor muscles and poor sacral ratios. What we are trying to say is that that is not a contraindication for pull-through, given the ability to form solid stool, because they can be managed with bowel management.