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Cloacal exstrophy—pull-through or permanent stoma? A review of 53 patients

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Abstract

Purpose: Patients with cloacal exstrophy have complex anomalies of the genitourinary and gastrointestinal tract with a spectrum of colonic length. Often, colon is lost during the initial management by use of ileostomies and for urologic and genital reconstruction. It is a common belief that these patients require permanent stomas, which we hypothesized is inaccurate, and therefore reviewed our experience with exstrophy, focusing specifically on a patient's potential to undergo a colonic pull-through.

Methods: All patients with exstrophy or exstrophy variant treated by the authors were retrospectively reviewed. Their ability to form solid stool was assessed via bowel management involving a constipating diet, antidiarrheals, bulking agents, and a daily enema through the stoma. Patients who underwent successful bowel management through the stoma were offered a pull-through.

Results: Fifty-three patients were treated over a 26-year period, including typical cloacal exstrophy (27), or a covered variant (16), and complex anorectal malformations with short colon (10). Newborn operations (48 done at other institutions, 5 by us) involved ileostomy in 11 or end colostomy in 42. Eight patients with ileostomies suffered acidosis and failure to thrive and underwent "rescue" operations to incorporate all defunctionalized colon into the fecal stream. Four had colon used for their urologic reconstruction and 6 for their genital reconstruction, leaving them borderline or unable to form solid stool. Twenty-three are undergoing bowel management or being observed for growth of the colonic pouch to determine if they are pull-through candidates. Of the others, 90% (27/30) underwent colonic pull-through. Ten percent (3/30) had a permanent stoma. Of 20 available for follow-up after pull-through, 17 are clean with bowel management (85%), 2 (10%) have voluntary bowel movements with occasional soiling, and 1 is incontinent but noncompliant.

Conclusions: Indication for pull-through depends on successful bowel management through the stoma, which depends on the ability to form solid stool. To maximize this potential, it is crucial to use all available hindgut for the initial colostomy and avoid use of colon for urologic or genital reconstruction. Most patients have poor prognosis for bowel control but can remain clean with bowel management. Our experience indicates that a permanent stoma is not required for the most of these patients and that bowel management can keep them clean, which we believe provides them with a better quality of life. Using these criteria, most exstrophy patients, contrary to popular belief, are candidates for a pull-through.

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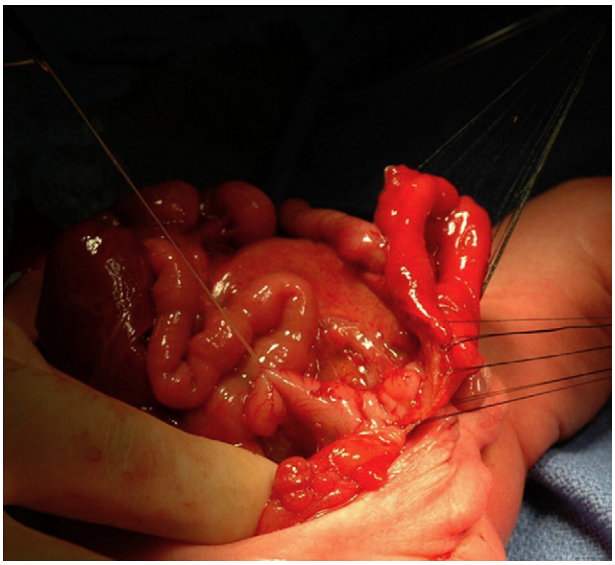

Fig. 1 Cloacal exstrophy patient with cecal plate mobilized and hindgut rescued from the pelvis. The cecum will be tubularized and the hindgut fashioned into an end colostomy.

The spectrum of cloacal exstrophy extends from its mildest defect of epispadias occurring in 30% of patients, to bladder exstrophy in 60%, and finally to its most severe form of cloacal exstrophy occurring in 10% [1-3]. Approximately 15 new cases of cloacal exstrophy, the most complex of all anorectal and genitourinariy malformations, are diagnosed in the United States each year.

Discussion of the colonic issues involved with exstrophy may include patients with classic cloacal exstrophy, covered cloacal exstrophy, and certain complex anorectal malformations with short colon. Although not technically exstrophies, their treatment is similar and, therefore, they are appropriate to include in this discussion.

With recent advances in neonatal intensive care and nutrition, survival with cloacal exstrophy has improved from 0% at the turn of the century to 50% in 1960, when Rickham first described successful reconstruction, to currently greater than 80% [2,4]. Patients with cloacal exstrophy are usually assumed to have fecal incontinence, with need for permanent fecal stomas [1,5-7], and urinary incontinence, which usually requires urinary diversion. Thus, the definitive reconstruction and postoperative management of the gastrointestinal and genitourinary tracts and their impact on long-term quality of life are still in need of improvements [1,6,8].

When one of these children is born, a multidisciplinary approach must be used, in which collaboration between pediatric surgeons, urologists, orthopedic surgeons, neurosurgeons, gynecologists, and neonatologists is paramount [1,6,9]. Initial management first consists of identification of associated anomalies including renal anatomy, hydronephrosis, hydroureter, tethered cord, myelodysplasia, and a separated pubic symphysis. During the initial surgical management of the newborn, the omphalocele must be repaired, bladder exstrophy closed (either primarily or in stages) with or without osteotomies, and the fecal stream diverted. In the past, most children underwent the creation of end or loop ileostomies [7,10]. This left the distal hindgut not only out of the fecal stream but remaining in continuity with the genitourinary tract. In their later repair, many children further lost colonic length with the use of colon for the urologic or genital reconstruction. This strategy was originally used given the assumption that these children were born with little to no colon [1,6]. However, this is a misconception, as these children are born with a spectrum of colonic length, from normal to none [1], and thus, the treatment plan needs to be individualized for each patient. We advocate a newborn operation with tubularization of the cecal plate, rescue of the hindgut, and creation of an end colostomy, thereby incorporating all available intestine into the fecal stream (Fig. 1) [1,6].

Nearly normal pelvic and sacral anatomy, as well as the presence of normal sphincter mechanisms [1,7,11,12], were thought to be requirements to perform a pull-through operation. It is clear that most of these patients have little to no sphincter and can be assumed to be fecally incontinent [1,7,11,12]. It is for this reason we believe that the concept that they will require permanent stomas is prevalent. However, as demonstrated for other patients with anorectal malformations who have little or no sphincter and are fecally incontinent, bowel management is an effective artificial means to keep such children clean [5,11]. We believe that a child's ability to remain fecally clean should be based upon successful bowel management, which must be compared to the quality of life with a permanent stoma [6,11], and have used the criteria of colonic length and the child's ability to form solid stool as the key determinants for successful bowel management and, thereafter, a pull-through.

The goal of this study was to review our experience with cloacal exstrophy as well as exstrophy variants and assess the

Table 1 Constipating diet

Foods to avoid	Foods to encourage
Milk or milk products	Apple sauce
Fats	Apple with skin
Fried foods	Rice
Fruits	White bread
Vegetables	Bagels
Spices	Jelly (No jam)
Fruit juices	Soft drinks
French fries	Banana
Chocolate	Pasta
	Pretzels
	Tea
	Potato
	Boiled, broiled, baked meat, chicken, or fish
	Pectin-based gelatins

ability of these patients to undergo a successful pull-through and remain clean.

1. Materials and methods

All patients with exstrophy or exstrophy variants treated by the authors were retrospectively reviewed. Data were collected regarding the patient's demographics, exstrophy classification, associated anomalies, initial newborn surgery, subsequent surgeries, colonic length, and ability to form solid stool. The patient's ability to form solid stool was assessed with a bowel management program which involved a constipating diet (Table 1), antidiarrheal medications, bulking agents, and a daily enema via the stoma. The enema volume and content was determined by trial and error over a period of 1 week with daily visits and plain abdominal radiographs [11]. Patients who underwent successful bowel management with the ability to have an empty stoma with no stool at all for 24 hours, with evacuation once daily with an enema, were offered pull-through operations. Follow-up regarding the patient's ability to remain fecally clean was then ascertained. The review was performed in accordance with institutional review board requirements.

2. Results

Over a 26-year period, from 1980 to 2006, a total of 53 patients were treated by the authors with malformations consisting of classic cloacal exstrophy (27), covered exstrophy (16), and complex anorectal malformations with short colon (10).

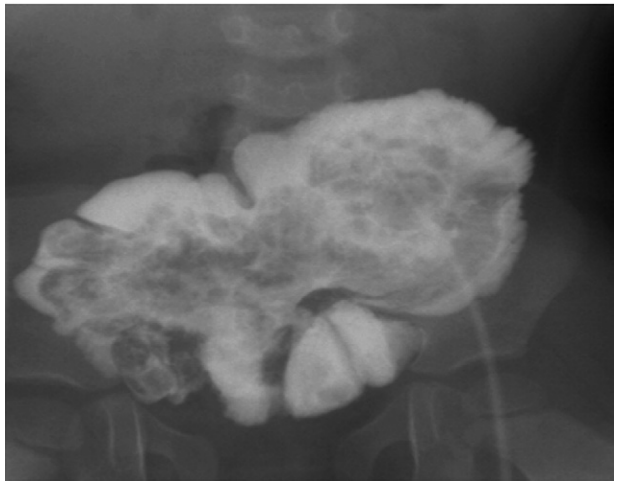

Fig. 3 Colostogram via the stoma of a cloacal exstrophy patient showing the colonic pouch. This patient was able to form solid stool.

Forty-eight newborn operations were performed at other institutions, and 5, at our Center. Initial newborn operations involved ileostomies in 11 patients and end colostomies in 42. Eight patients with ileostomies suffered acidosis and failure to thrive as a result of resorption of urine by bowel mucosa [5,8,9,12]. These patients subsequently underwent "rescue" or colon salvage operations at which all defunctionalized colon was incorporated into the fecal stream. Thereafter, they experienced resolution of these sequelae.

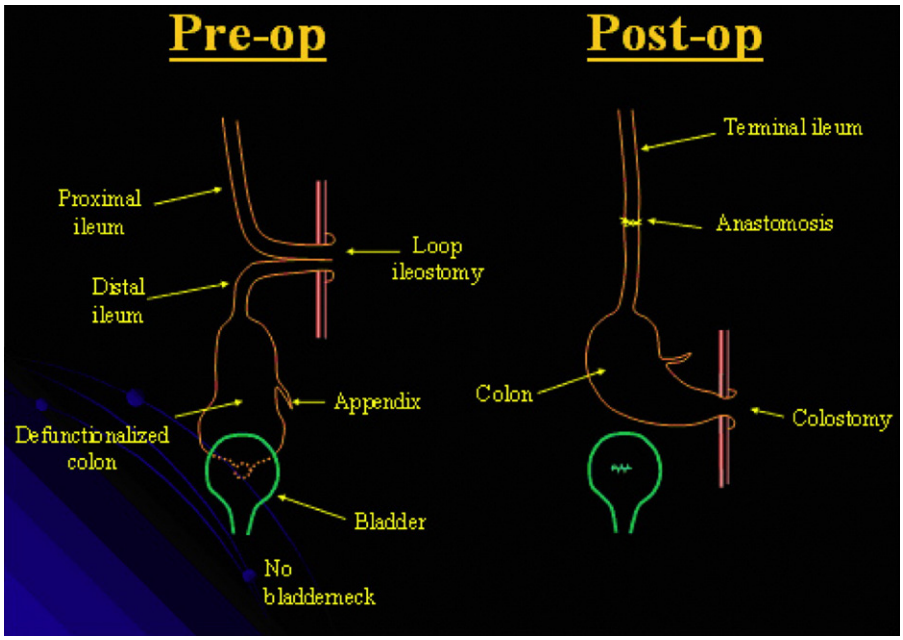

Fig. 2 Diagram of prerescue operation anatomy (end ileostomy with distal colon left out of fecal stream) and postrescue operation anatomy (end colostomy with entirety of available intestine incorporated into fecal stream).

Table 2 Summary of patients undergoing bowel management, pull-through, or permanent stoma creation

	No. of patients	Completed bowel management (n = 30)	Still undergoing bowel management (n = 23)	
		Pull-through	Permanent stoma	
Classic cloacal exstrophy	27	89% (16/18)	11% (2/18)	9 (33%)
Covered exstrophy	16	86% (6/7)	14% (1/7)	9 (56%)
Complex anorectal malformation with short colon	10	100% (5/5)	0% (0/0)	5 (50%)
Total	53	90% (27/30)	10% (3/30)	23 (43%)

This procedure involves taking the end or loop ileostomy down and allowing the entirety of the patient's small and large intestine to be in continuity as part of the fecal stream (Fig. 2). An end colostomy is then created. This then allows for growth of the colonic pouch [1,6,7,13]. Ten (19%) of 53 patients were assumed to have minimal to no colon with little hope of ultimately undergoing successful pull-through operations. Thus, the colon in these patients was used for urologic (in 4) or genital reconstruction (in 6), leaving them borderline or unable to form solid stool.

Patients were observed for growth of the colon. Colonic length and growth was assessed by colostogram (Fig. 3).

Twenty-three patients are currently undergoing bowel management or being observed for growth of the colonic pouch to determine if they are pull-through candidates (Table 2). Of the remaining 30 patients, 27 (90%) underwent colonic pull-through. Of the 30, 3 (10%) were unable to form solid stool and, thus, were left with a permanent stoma. Of the 20 patients available for follow-up after the pull-through operation, 17 (85%) patients are clean, 2 (10%) patients have voluntary bowel movements with occasional soiling, and 1 patient is incontinent but noncompliant (Table 3).

3. Discussion

With improvements in ultrasound and the advent of fetal magnetic resonance imaging, prenatal diagnosis of cloacal exstrophy has improved dramatically and allows accurate diagnosis as early as 16 to 20 weeks of gestation [14,15]. This has prompted many of these patients to be born at tertiary care centers providing for their complex needs. It has also likely increased the rate of pregnancy termination [15].

Upon initial evaluation, the medical team must first identify associated anomalies, evaluate for life-threatening issues that must be dealt with immediately, and determine gender assignment. The team must then coordinate a surgical plan taking all factors and malformations into account.

The initial newborn surgical management must not only stabilize the patient but also minimize long-term consequences. This requires closure of the omphalocele, reapproximation of at least the posterior bladder walls, and fecal diversion. End or loop ileostomies for fecal diversion should be avoided [5,8,9,16]. They can lead to fluid, electrolyte, and nutritional abnormalities, dehydration, metabolic acidosis owing to urine absorption via the distal hindgut, and urinary tract infections [5,8,9]. Perhaps most importantly, with respect to the possibility of a future pull-through, the diverted distal hindgut becomes defunctionalized and later atrophic, negating its future use in definitive reconstruction [8,9]. We rather advocate the creation of an end colostomy [1], preservation of all pieces of colon, so that the entirety of the patient's intestinal tract is exposed to intestinal contents and allowed to grow [7,13,16]. This requires mobilization of the cecal plate from between the 2 hemibladders, and its tubularization. The hindgut, which can be duplicated, needs to be rescued from the pelvis and fashioned as an end stoma (Fig. 1).

In cases in which an initial ileostomy was created, there sometimes exists an opportunity to "rescue" the hindgut and incorporate it into the fecal stream (Fig. 2). This allows the patient's colon to grow and maximizes the potential for that colon to form solid stool.

Given the improved survival in patients with cloacal exstrophy, we must now look toward helping them achieve as normal a quality of life as possible. True fecal continence is rare; however, social continence is an attainable goal [1,5,17]. This review demonstrates that such patients can undergo an effective pull-through rather than being left with a permanent stoma. It is our impression that, like other patients with anorectal malformations and fecal incontinence, bowel management and a clean child is better in terms of quality of life than a permanent stoma [11]. With the help of a constipating diet, antidiarrheal medications, and bulking agents, these children can usually form solid stool that is then evacuated once daily with the administration of an enema via the stoma, a new concept we introduce here. In this review, 17 (85%) of our 20 pull-through patients available for

Table 3 Results after pull-through operation—20 patients available for follow-up

Patient condition	No. of patients
Clean	17 (85%)
Voluntary bowel movement with occasional soiling	2 (10%)
Incontinent	1 (5%)
Total	20

follow-up achieved this. We anticipate a similar group, currently undergoing bowel management, will achieve success as well.

After a period of adequate growth, we propose that all patients be given the opportunity to prove whether they will be able to remain fecally clean with a pull-through operation. Rather than relegating these children to a lifetime with a stoma, each child can have an individualized treatment plan, based on their colonic length, ability to form solid stool, and capacity to remain fecally clean. Thus, in our treatment algorithm, patients with short colonic length who are unable to form solid stool, once tested, should remain with a permanent stoma. However, patients with enough colon, having the ability to form solid stool, who remain fecally clean for 24 hours with bowel management through the stoma, are considered candidates for a pull-through operation. Our series demonstrates that a successful pull-through is possible in most of such children.

As part of the pull-through operation, we also advocate the creation of a Malone appendicostomy, allowing the catheterization of the cecum from the anterior abdominal wall often via the umbilicus. The status of the colon can usually be determined before the proper timing of definitive urologic reconstruction, and thus, the pull-through and the appendicostomy can be coordinated to be done at the same setting. If the appendix is too small or nonexistent, a neoappendix can be created from the colonic wall with a mesenteric pedicle and used as a conduit [17,18]. This procedure enables the child to self-catheterize and administer an enema via the appendicostomy while sitting on the toilet and evacuating once daily. Patients then remain on the bowel management protocol via an antegrade enema after the pull-through operation to remain clean [18].

Patients with short colons should not have any of their colon used for urologic or genital reconstruction. It is thus imperative that collaboration exist between the pediatric surgery, urologic, and gynecologic teams before any intestine is used. However, those patients that have failed bowel management, who were unable to remain fecally clean for 24 hours, and were thus not considered candidates for a pull-through operation and will remain with permanent stomas, can have portions of their colons used for genitourinary reconstruction.

Using this protocol, most exstrophy patients, and those with complex malformations with short colon, contrary to popular belief, are candidates for a pull-through operation and have the capacity to remain fecally clean. This, hopefully, can provide a better quality of life for these complex children.

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Discussion

W. Hardy Hendren, MD (Duxbury, Mass): I would like to emphatically agree with the message that you have just given. I have been very distressed by some of the same things that you have talked about.

Six years ago, Dennis Lund and I reported 50 cloacal exstrophy patients in the *Journal of Pediatric Surgery* in 2001, and half of those patients had a pull-through, and in only 4 cases was it necessary to go back and reverse them because they simply could not hack it in terms of continence.

I would like to credit Dr John Raffensberger with giving me this thought 25 years ago because he referred a youngster for secondary urologic reconstruction, and she had a pull-through procedure. John had done that as a newborn, and so, we have even stretched this concept to doing a few newborn pull-throughs in order to never give them a colostomy at all except a complementary one at the time that we were doing the initial pull-through. I think that some very favorable cases can even be stretched to that point.

In the 50 cases that we reported, there was only one mortality, and that was a preoperative case that we had seen in consultation who was waiting to be admitted and had one of these acidotic episodes. I would say that it is our experience that most of them need to depend on washouts for managing their colon and on self-catheterization of their bladder, which is certainly not a normal bladder, but it suffices and they can do perfectly well.

I would also like to emphasize that if there is any anomaly that should be regionalized, it is this one. There are only about 20 cloacal exstrophies in all of the United States in the course of a year, and for a pediatric surgeon or a pediatric urologist or anybody to tackle one of these cases without a lot of experience is simply crazy. Nobody will gain a lot of experience unless you refer them, and I think, ideally, we probably ought to have a place on the East Coast, in the middle of the country, on the West Coast and that's because that way, you can have several people in several locations who really have an interest in this and follow them through.

I would like to say that these patients must be followed forever. Tomorrow I am going to see a 35-year-old woman whose reconstruction was done many years ago. It took me 24 hours to reconstruct all of the things that she had, but she is leading a very good life. Let me say also that the patient that John Raffensberger sent who had the previous pull-through is now a very successful businesswoman worth several million dollars. She has a business that she started when she was in college and is now employing a whole bunch of people. I hope she is going to ultimately give the hospital some philanthropic help. (laughter)

Most sad to me is seeing a youngster where somebody who did not know the first thing about cloacal exstrophy has gone in and taken out that rudimentary colon which will come up to be about 30% of a normal colon length if you give it a chance as you just emphasized.

I would also say that we must prepare these families that they are going to need multiple operations, and I would just say that everybody in this room knows that kids do well despite having multiple operations if you fix what's ailing them, and we usually can do that.

Thank you for a great paper.

Grace Z. Mak, MD (response): Thank you for those comments.

Michael Helmrath, MD (Houston, Tex): Grace, very well presented. I wanted you to comment quickly about the timing of the pull-through with respect to the bladder. Are these done in conjunction with augmentations of the bladder? As the bladder matures, does that influence if you would pull some of these kids through? How long do you tend to wait with the typical patient?

Thank you.

Grace Z. Mak, MD (response): What we have done is oftentimes a collaborative effort, and so, the bladder reconstruction and the pull-through operation are often done simultaneously but the pull-through operation is done first.As far as the timing, I would have to refer to Dr Levitt for an answer

Marc A. Levitt, MD: Mike, thank you so much for that question. I think a collaborative plan from day 1 is really vital. We will wait until the child is 3 or 4 years of age, leaving them with their end colostomy all that time, allowing it to grow. We then will test the colon to see if it in fact has the capacity to form solid stool. If it does, then the patient is a candidate for a pull-through, and the best time to do that is at the time of the bladder reconstruction. You put the colon down posteriorly. It is a lot easier to do that than after an augmentation has been done. If the colon is not capable of forming solid stool, and you have really proven that, you can actually then use some colon to give to the urologist, but that has to be the last resort once you determine that that colon is not capable of forming solid stool.

Howard Snyder, MD (Philadelphia, Pa): All the commenters are people whose opinions I respect. We have been one of the centers for cloacal exstrophy because it was I think Peter Paul Rickham and C Everett Koop who realized that this was not a lethal anomaly which it really was still considered by many when I was a young surgeon.I want to rise just to caution you on 2 things. I think it is very important to remember that these patients never have totally normal colon or bowel function in general. I do not think we have a single patient who is fat who has cloacal exstrophy. This is a lifelong problem, and some of the most complicated metabolic patients we have are all cloacal exstrophy patients. They suffer from 2 further problems. They have a very high incidence of spinal anomalies and also a high instance of renal anomalies unlike classical exstrophy, so that they become very complicated patients to take care of. I think that what I have taken away from this elegant presentation is that you can manage a perineal colostomy pretty darn well, and I think that is really what the take-home message is. Our experience has been that these kids undergo their continent urinary reconstruction I think on an average 4

or 5 years later than the other kids that we take care of that require a reconstruction simply because they are nutritionally not ready for this. I think it is admirable if you have a subset where you can actually carry this out very early in life because that has not been our experience, but this is certainly an illustration of how regionalization can indeed benefit this most challenging of all our reconstructive patients.

Grace Z. Mak, MD (response): Thank you.