

Cloaca, The Most Severe Degree of Imperforate Anus

Experience With 195 Cases

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Objective

To provide a follow-up of 195 patients with cloacal malformations seen by the author from 1959 to 1998.

Summary Background Data

Cloaca, which occurs in approximately 1 of 50,000 births, is the most complex type of imperforate anus with confluence of the rectum, vagina, and bladder in a urogenital sinus. Functional results for the bowel, the genital tract, and the urinary tract were formerly poor. Cloacal exstrophy, which is an even more complex spectrum of malformations, was uniformly fatal until 1960. In addition to imperforate anus, these babies have an omphalocele, two exstrophic bladders, between which there is an open cecum, and a blindly ending colon hanging down in the pelvis from the cecum.

Although both of these diagnoses contain the word "cloaca," which is Latin for sewer, they are really two separate entities in terms of surgical management. Cloaca and cloacal exstrophy in most cases are very different anatomic problems. However, there are variants that are like a hybrid, which is the rationale for reporting together an experience with both entities.

Methods

Records were reviewed of 154 patients with cloaca and 41 patients with cloacal exstrophy to assess anorectal function, urinary continence, and sexual function where available.

Results

Follow-up was available in 141 cloaca patients: 82 have spontaneous bowel movements and satisfactory control, 38 use enemas to evacuate, 9 have a colostomy, 7 have fecal soiling, and 5 are too recently operated to evaluate. Regarding urinary control, 83 void spontaneously, 40 catheterize to empty, 4 have urinary diversion, 1 has a continent diversion, 5 patients are wet, and 8 are too recently operated to judge. Twenty-four patients are now adults, 17 of who have experienced coitus and 7 have not. Seven have had babies, all except one by cesarean section.

Results of surgery for cloacal exstrophy are not as good, but are encouraging nonetheless for an anomaly that was uniformly fatal before 1960. Of the 41 cloacal exstrophy patients being followed, 7 have not undergone surgery. Fifteen have a colostomy; 19 had pull-through of the colon, but 3 were subsequently reversed for fecal incontinence. Most depend on enemas to evacuate. Urinary dryness was attained in 30 patients, usually by intermittent catheterization of the bladder, which was augmented with small bowel or stomach or both. Only three void voluntarily. Fifteen of the completed long-term patients wear no bag. Only three of the completed patients wear two bags. The rest have one bag.

Conclusions

Imperforate anus and associated malformations in cloaca and cloacal exstrophy are not hopeless problems. A reasonable lifestyle can be achieved for most of these children with comprehensive surgical planning.

Cloaca is the Latin word for sewer. In the human embryo at the 7.5-mm stage, it is a common chamber into

which converge the primitive urinary tract, genital tract, and bowel.¹ At the 22-mm stage a urorectal septum descends, separating the rectum from the urogenital sinus. The urogenital sinus then differentiates into the urinary and genital tracts. Persistent cloaca is normal in birds, reptiles, and some fish. It can be disastrous for a human infant if not managed well. Cloaca is the most severe type of imperforate anus in girls. Persistent cloaca

Presented at the 118th Annual Meeting of the American Surgical Association, Palm Beach, FL, April 1998.

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Accepted for publication April 1998.

RECTUM VARIATIONS

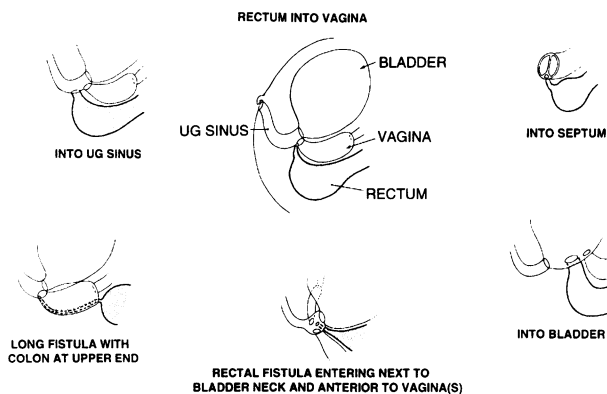


Figure 1. The most common anatomy seen in persistent cloaca is shown at top center. There is a urogenital sinus. The vagina enters just below the bladder neck. The rectum enters just below the vagina. The level of confluence of the structures can be high, low, or intermediate; this can be determined by radiographic contrast studies and endoscopy. Variations in the rectal anatomy are shown. Proceeding clockwise, at top right the rectal fistula enters at the base of a septum dividing two vaginas. Double vagina is often present. At bottom right, the rectum enters the bladder (rare). At bottom center, the rectum enters the urogenital sinus close to the bladder neck and slightly above the entry of two separate vaginas, which enter laterally and diverge away from the midline. At bottom left is shown a high rectum with a long fistula, which runs the full length of the vagina but enters in the most common location. At top left, the rectum enters the urogenital sinus separate from the vagina (rare). It is essential to be aware of the many variations in anatomy of the rectum, and urogenital sinus.

occurs once in 50,000 births. The diagnosis includes a wide spectrum of pelvic malformations. Figure 1 depicts the most common anatomy of a cloaca and shows some of the many variations in the rectal anatomy. Similarly, Figure 2 shows the perineum of four patients with cloacal malformations, each quite different from the others. It is a wide spectrum. At the mild end is a persisting urogenital sinus opening, which drains urine, and an anteriorly displaced anus adjacent to it. This is the *forme fruste* of a cloaca. Progressing in severity, all three tracts converge inside the pelvis. This can be low, just beneath the skin with a short common channel, or the structures can join high near the bladder neck, an entirely different surgical challenge. Rarely the structures converge in the bladder with a cleft where the bladder and vagina failed to separate. At birth these infants may be very distended because not only is the colon obstructed, but also the lower urinary tract. The mechanism for this is shown in Figure 3. Urine flows from the bladder into the long urogenital sinus and backs up in the vagina, which can become very distended and displace the bladder forward. This causes functional bladder outlet obstruction as well as compression of the ureters. Hydronephrosis is common.

Cloacal exstrophy is an even more difficult problem to manage than "ordinary" cloaca. Fortunately it occurs only once in every 250,000 births. Antenatal diagnosis by ultra-

sound followed by pregnancy termination may reduce the already low incidence of this and other malformations even further in the future. Rickham reported the first successful salvage of a baby with cloacal exstrophy in 1960.² Features of cloacal exstrophy are shown in Figure 4. They include omphalocele, two hemibladders with everted cecum between them, a rudimentary hemipenis on each side in boys, and a blindly ending microcolon, which hangs down in the pelvis from the everted cecum. Commonly the ileum prolapses as the infant cries. Figure 5 shows the appearance at birth of a boy with cloacal exstrophy. Cloacal exstrophy is believed to be caused by premature rupture of the cloacal membrane before complete descent of the urorectal septum. Spinal malformations are common. Many have a myelomeningocele or lipomeningocele with poor or absent perineal muscles.

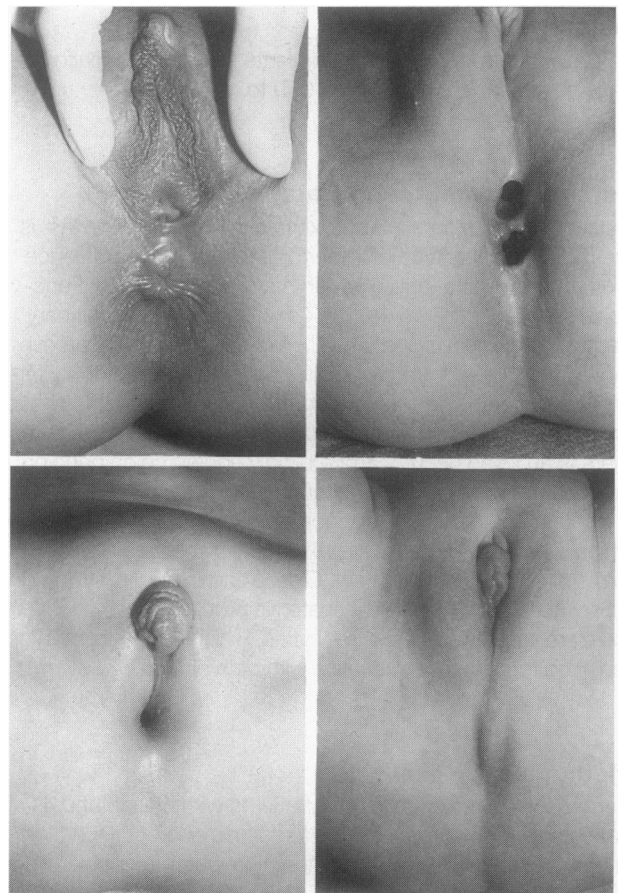


Figure 2. Four patients with cloacal malformations to illustrate the wide spectrum of perineal anatomy. (Top left) Minimal anomaly. Note the incompletely formed vaginal introitus with a web of perineum covering its posterior half and the anterior displacement of the anus from a normal position, which would be centered in the perianal skin pigmented. (Top right) Large urogenital sinus opening and anal opening just behind it. The two passages are separated by a thin urorectal septum. (Bottom left) A single perineal opening into which opened the colon, vagina, and urethra at a low level. (Bottom right) Typical cloacal malformation with high confluence of all systems at the upper end of a long urogenital sinus. The urogenital sinus ended in a small opening at the base of the slightly enlarged clitoris.

OBSTRUCTION BY URINE FILLED VAGINA

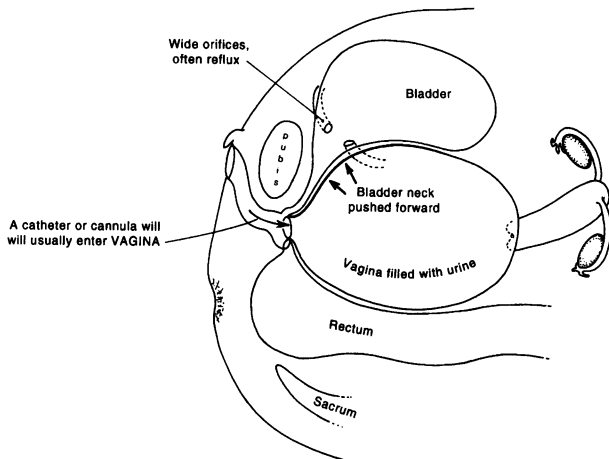


Figure 3. Typical anatomy of a high cloacal malformation. As urine enters the long urogenital sinus, the vagina becomes distended, displacing the bladder and ureters forward, which can cause severe hydronephrosis. In many such cases intermittent catheterization will empty the urine-filled vagina and relieve hydronephrosis. Vesicostomy or vaginostomy are not usually necessary. When there is low confluence of the pelvic structures this sequence is not usually present—the child can empty the bladder, and the vagina does not become distended. However, more than half of these patients have important vesicoureteral reflux.

METHODS

A retrospective review was made of all the patients I have seen with cloaca and cloacal exstrophy since 1959. It included 154 patients with cloaca and 41 patients with cloacal exstrophy.

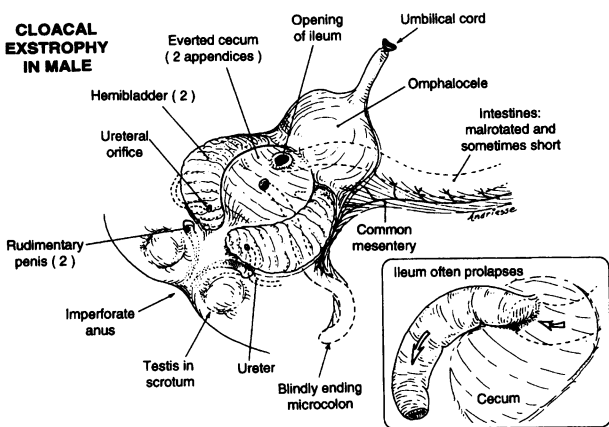


Figure 4. Typical anatomy of a baby boy with cloacal exstrophy. Testes and scrotum are present. There is usually a rudimentary hemipenis on each side, widely separated. Cloacal exstrophy occurs in both boys and girls. Girls usually have two vaginal openings, widely separated, entering the lower aspect of each hemibladder. Note the cecum between two hemiblasters. The blindly ending microcolon hangs down from the cecum in the pelvis. It is common for terminal ileum to prolapse from the cecum. Rarely the colon has more length and its distal end can terminate in the exstrophied bladders.

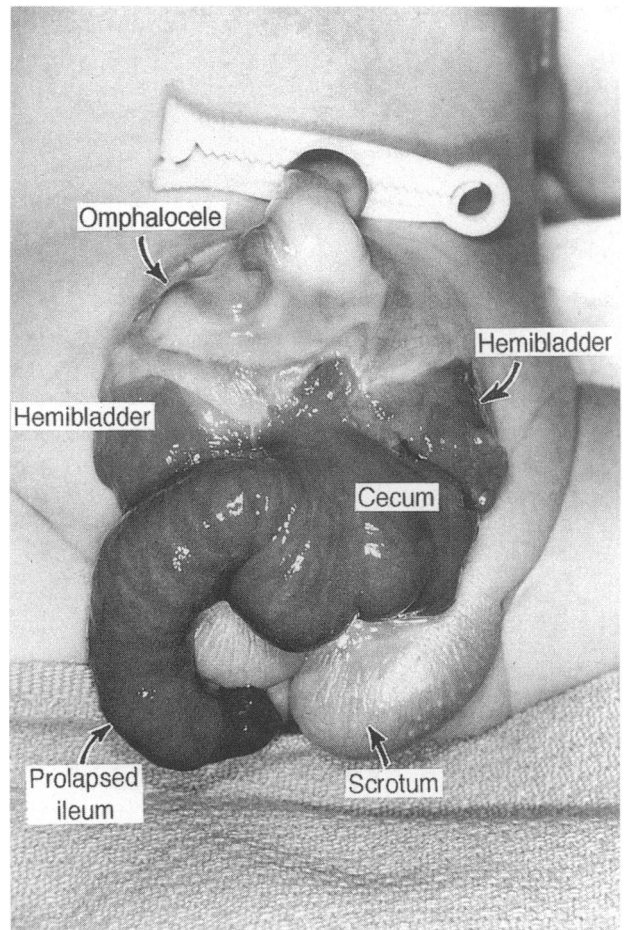
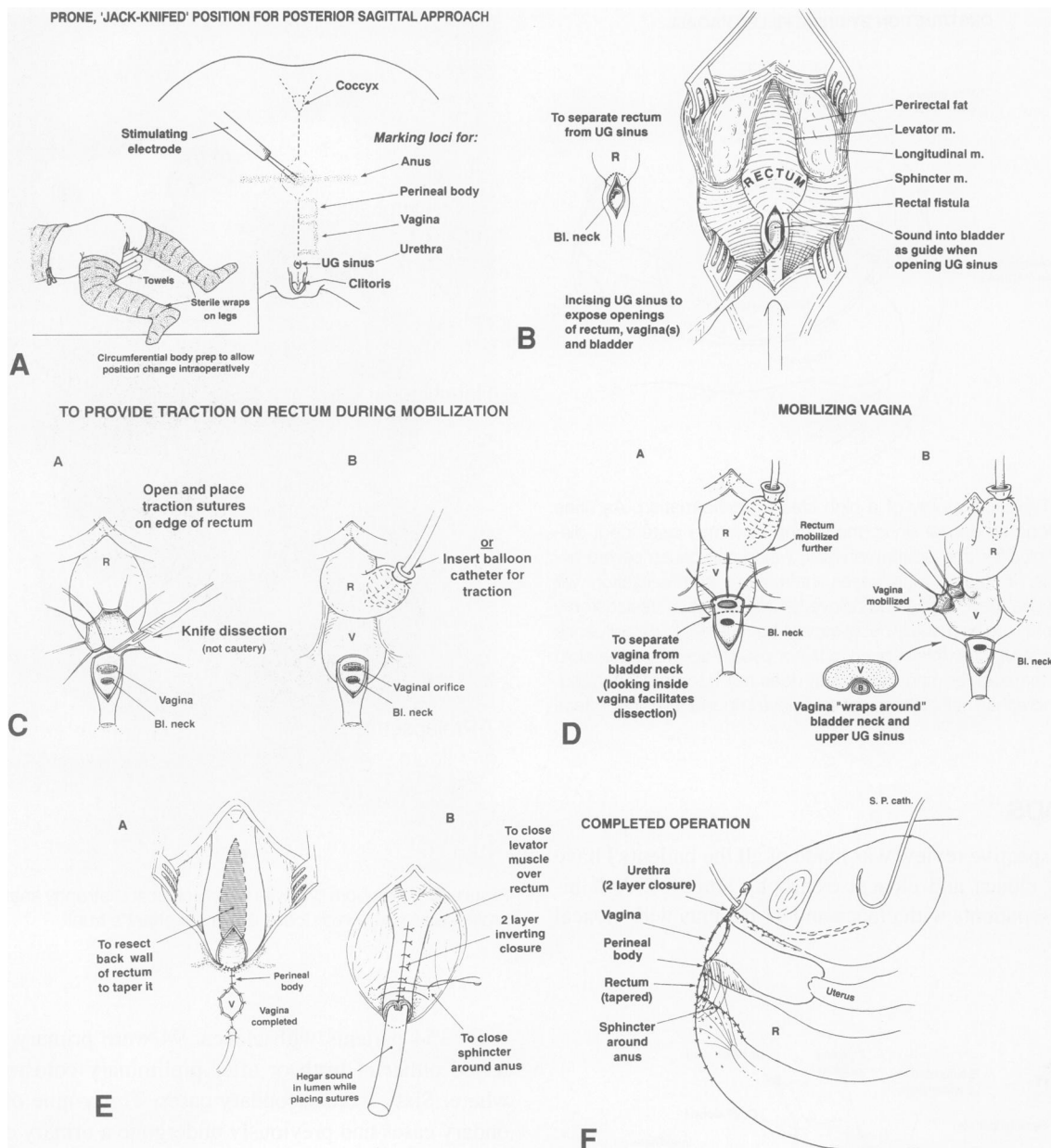


Figure 5. A newborn boy with typical cloacal exstrophy anatomy. Note prolapsed ileum, which looks like an elephant's trunk.

Of 154 patients with cloaca, 94 were primary cases referred either at birth or after preliminary colostomy elsewhere. Sixty were secondary cases. Thirty-nine of the secondary cases had previously undergone a urinary diversion: vesicostomy in 22, ureterostomy in 8, ileal loop in 8, and continent diversion in 1. This underscores the fact that the lethal part of cloacal malformation is the urinary tract. Ninety-six of the 154 patients had significant vesicoureteral reflux.

The 41 cloacal exstrophy patients included 11 primary cases and 30 secondary cases. Twenty-four were genetic males, but only three were raised as boys because the phallic structures are usually rudimentary and not well suited for constructing a good penis. Seventeen were genetic females. Seven of the patients have not yet undergone surgery. Three are infants who will undergo reconstruction in the near future. Four are older children who are living with a colostomy and complete urinary incontinence, in diapers, whose families have not been able to face the substantial uncertainty of the outcome of major reconstructive surgery. This is understandable.



Management of Cloaca

The neonate with a cloaca must have a decompressive colostomy, except for those rare instances where there is an actual perineal opening of the colon. A right transverse colostomy is preferred over use of a left lower quadrant colostomy, which can compromise a subsequent pull-through procedure, especially if located too low, or can make it difficult to use a segment of bowel to extend a vagina that will not reach the perineum. In patients with hydronephrosis and filling of the vagina by urine and mucus, this must be drained. That can usually be accomplished by intermittently catheterizing the urogenital sinus. The catheter invariably enters the vagina and not the bladder, which is angulated forward. It is not usually necessary to perform a vesicostomy or a vaginostomy, both of which we

have seen in secondary cases. It is sometimes helpful to cut back the urogenital sinus a few millimeters at the time of colostomy to facilitate catheterization of the baby's vagina. The temptation to do a formal abdominal exploration at the time of colostomy should be resisted because it will create abdominal adhesions. The information about pelvic structures present can be obtained by appropriate radiographic and endoscopic studies in the next few weeks of life.³ A magnetic resonance imaging examination of the lumbosacral spine is indicated because a third of these infants have tethering of the spinal cord.⁴ Nearly all cloacal exstrophy patients have a tethered cord. Neurosurgical release can prevent nerve deficit with growth but will not improve already established neurologic changes. If the infant has severe vesicoureteral reflux or another life-threatening urop-

Figure 6. Cloacal repair using the posterior sagittal approach. (A) Sterile preparation of the entire lower half of the patient allows working from behind or turning the patient into the supine position or then elevating the legs for the lithotomy position. Frequently, more than one position is used during surgery. The stimulating electrode determines the center of the rectal sphincter. It is useful to mark that point, as well as the perineal body, the vagina, and the urethra. (B) A midline incision is made from the coccyx through the muscle complex and down to the rectum and urogenital sinus. A metal sound should be placed in the urogenital sinus to ensure staying in the exact midline. If the urogenital sinus is wide, it should be opened completely to its end to enable tapering its caliber to a size suitable for a urethra. If the distal urogenital sinus is not dilated, opening it completely is not necessary. (C) The rectal communication has been opened and is circumscribed to mobilize the rectum away from the vagina. Traction sutures can be applied to its edges, or a balloon catheter can be introduced for gentle traction. The bowel is always thoroughly prepared before surgery, irrigating from below through the rectal fistula at endoscopy or from above through the distal limb of the colostomy. Inspissated mucus should be washed away, sometimes using McGill forceps to extract solid pieces of mucus, or an Ellik evacuator applied to the end of an endoscope. (D) Mobilizing the vagina. This is the most difficult part of a cloacal repair because the vagina is closely adherent to the urogenital sinus and bladder neck and often wraps around the bladder outlet. This is a tedious dissection by any means. The tissues are thin and the blood supply is tenuous after this dissection. Therefore, I prefer to do this separation by sharp knife dissection, often with a finger in the vagina if the patient is large enough. Frequently after this dissection there are holes in the anterior wall of the vagina. This will not cause a problem if the vagina is rotated 90° when pulled through, to have an intact lateral vaginal wall overlying the closure of the bladder neck and urogenital sinus. It is important to avoid inadvertent injury to the bladder, which is apt to cause a fistula to form after surgery. I find that using cautery for this separation can be convenient, and saves some time for the dissection, but carries a greater risk of postsurgical fistula formation from thermal injury to these delicate tissues. (E) The urogenital sinus has been closed. A second layer of periurethral tissue can usually be closed over it. It is important to have intact vaginal wall cover this, by rotating the vagina 90° to have intact lateral vaginal wall covering the urethral closure. If the vagina is slightly short, local flaps of perineum can be inserted into it to avoid tension. The perineal body is then closed. The rectum is brought down to the perineum. If it is dilated, its caliber should be tapered appropriately by resecting a wedge of back wall and closing it in two layers. Alternatively, if it is only slightly dilated, it can be plicated to reduce its caliber. The perineal muscle complex and levators are closed over the rectum with a sound of appropriate size to avoid excessive narrowing of the canal. (F) The completed closure. A straight catheter is used rather than a Foley catheter to avoid the complications that could occur if a Foley were pulled through the closure with the balloon intact; this could cause serious damage to the repair.

athy, it should be corrected straightaway. It is a serious error to concentrate on the rectal pull-through, leaving the urogenital reconstruction until later. I have seen many secondary cases where that was done, and I strongly caution against it.

Definitive correction of a cloaca can be done between 6 and 24 months. Preliminary endoscopy to review the anatomy is useful, and the colon should be cleansed thoroughly at that time. This can be time-consuming. Irrigating through the colon fistula from below and out the colostomy can help. This can be done through a small endoscope or by inserting a Foley catheter under endoscopic guidance into the fistula to irrigate large volumes of saline, not water, from below and out the distal limb of the colostomy. The urologic Ellik evacuator attached to an endoscope either above through the colostomy or below can be helpful. Blunt McGill anesthesia forceps can be helpful for extracting lumps of inspissated puttylike mucus from the defunctioned colon. These global operations should not be attempted without thorough cleansing of the bowel first.

Figure 6 illustrates surgical correction of a cloaca using the posterior sagittal approach. We have used this for cloaca cases and for other high imperforate anus cases since Peña and deVries described it in 1982.^{5,6} Abdominal perineal exposure was used previously in the majority of our cases before 1982 and is still used in some where the entire

operation cannot be performed from behind. GoLYTELY (Braintree Laboratories, Braintree, MA) is used to prepare the functional bowel.

Complete body preparation allows starting in one position and turning to another. We sometimes start in the posterior sagittal position and then turn for laparotomy if structures cannot be fully mobilized from below, alternatively picking up the legs to work in the lithotomy position. The belly can be closed temporarily with a few sutures, turning back to the posterior sagittal position if necessary. It is much easier than some other approaches used for abdominal perineal exposure. Turning back and forth is much easier in a small child than in an adult. Two anesthesiologists are mandatory for turning, one to manage the endotracheal tube and the other the infusion and monitoring lines, which are in the upper extremities. Occasionally the operation is started from the belly if the rectum is very high or if structures are tethered upward by a previous vesicostomy, ureterostomy, or low colostomy.

There should be a sound in the urogenital sinus when it is opened. This ensures staying in the midline. If the urogenital sinus is wide, it can later be tapered to form a suitable urethra. Mobilizing the rectum is the easy part of a cloacal repair. The coccyx can be split or removed to get up to the peritoneal cul-de-sac. Distending the rectum with air during

its mobilization can be helpful. In an older patient, inserting a finger into its lumen can prove helpful.

The most difficult part of these reconstructions is separating the vagina, or vaginas, from the bladder neck and urogenital sinus. I prefer to do this by sharp dissection, not cautery. There was one vagina in 66 patients, two in 68 patients, and none in 20. It was necessary to use bowel to reconstruct the vagina in 46 patients. In some this was as an extension of a small vagina, and in others it was a substitute for the absent vagina. When the vagina is mobilized from the bladder neck and urethra, every effort should be made to stay out of the bladder. It is common to make one or more holes in the anterior vaginal wall during this dissection, however carefully it is done. It is helpful to have a finger inside the vagina during its mobilization if possible. The lateral walls of the vagina should be intact after mobilization. By rotating 90° an intact lateral wall of vagina over the urogenital sinus, fistula will be prevented. Creating local flaps of perineum can help accomplish sewing the vagina to the perineum without tension. It is a formidable procedure to close a vesicovaginal fistula after cloacal repair, so it is highly desirable to avoid that complication.

Management of double vagina is variable. Most commonly the two are side by side and can be converted to one by simply incising the septum endoscopically with a cutting electrode during infancy. When there are two separate vaginas, it may be better to pull through only one if it is larger than the other. In some cases the two can be sewn together and pulled through or extended downward with bowel. *Willingness to improvise is essential in this type of surgery.*

When the rectum is very dilated, its size should be reduced. This can be done by wedge resection and two-layer closure or by simple plication, over a sound of appropriate size if the rectum is only mildly dilated. Enfolded tissue after plication disappears in time, just as it does with a plicated ureter.

Peña has described an alternative technique for bringing down the urogenital sinus and vagina in a low cloaca.⁷ It involves *en bloc* mobilization of the urethra and vagina to pull them down together as a single unit. I have used this method in two favorable cases and agree that it should be part of the armamentarium in treating cloaca.

After a patient has been in the prone position for several hours, the face is very edematous. These children routinely spend a day or two in the intensive care unit after surgery, often intubated and with ventilatory support. After major pelvic operations, I prefer to keep the patient flat, not propped upright, to minimize pelvic venous pressure, which promotes venous stasis. Pneumatic leg boots are used on older patients to reduce the likelihood of pulmonary embolism. Postsurgical urinary tract drainage is maintained with a small straight plastic catheter. A Foley catheter is not used because its balloon can injure the bladder neck and urogenital sinus closure if it should be pulled out inadvertently.

These patients, and all other pediatric surgical patients who may need multiple operations, are at risk for develop-

ing latex allergy.⁸ Powdered latex gloves are no longer used in our operating room to protect both patients and operating personnel from this recently recognized common problem. Indeed, one of our operating room nurses recently experienced an almost fatal anaphylactic pulmonary disorder that was caused by chronic latex exposure.

Three weeks after surgery, endoscopy is performed to assess anatomic healing and to institute intermittent catheterization if the patient cannot void spontaneously. New vaginal and anorectal passages are routinely dilated by the parents using a Hegar dilator of appropriate size. This is done with decreasing frequency until postsurgical induration has disappeared. Colostomy closure is performed 4 to 6 weeks after cloacal reconstruction. A daily washout of the colon using saline may be needed if a child does not evacuate spontaneously. Saline is warm tap water combined with 2 teaspoons of table salt per quart of water. Most parents do not know how to give an enema and need appropriate instruction, stressing slow intermittent irrigation to avoid painful cramps.

Management of Cloacal Exstrophy

Surgical management of an infant with cloacal exstrophy is entirely different from the newborn with a cloaca. A major operation is done at birth, not just a colostomy. This entails omphalocele closure, separating the gastrointestinal tract from the bladders, sewing the two hemibladders together, and closing them. The widely divergent pubis is approximated. In some infants, iliac osteotomy is performed to facilitate pubic closure. Wire is not used to approximate the symphysis, because if it later drifts apart the wire can cut underlying soft tissue. Positioning the baby in modified Bryant's traction, with legs suspended at 90° from the body, will help hold together the two halves of the pelvis and the attached soft tissues.

Management of the colon varies. If there are severe spinal defects, with sacral agenesis and no perineal muscle (as determined by electrical stimulation), a pull-through immediately or later is futile. End colostomy is performed in that circumstance. A cecal closure, once separated from the bladder, is tenuous in a neonate with a new colostomy downstream from it. Therefore, it is often safest to perform a temporary decompressive loop ileostomy proximal to the cecal closure, which will be stressed from edema of an end colostomy or a pull-through distal to the cecal closure. The ileostomy can be closed several weeks later after a contrast study demonstrates the bowel is intact distally. If there is an excellent perineal muscle complex, the colon can be pulled through at birth or later, centering its end in the point of maximal contraction of the perineum.

The rudimentary microcolon should never be removed because it has the capacity to enlarge, lengthen, and function as colon. Commonly, as the child grows, a microcolon will attain 30 to 40 cm in length and will provide satisfactory water absorption. It should never be used for bladder

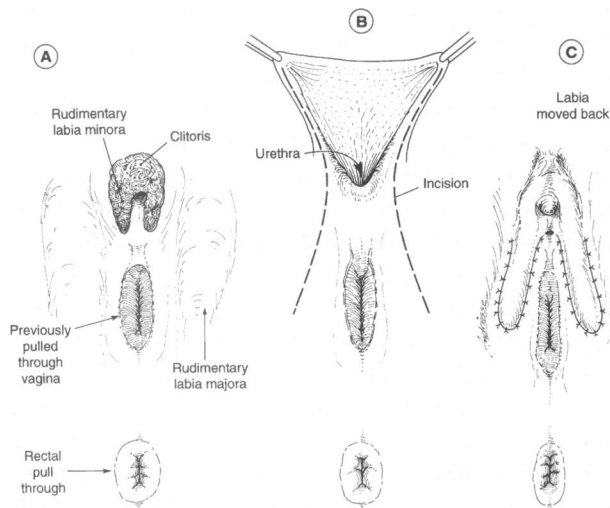


Figure 7. Labioplasty, which is usually deferred for several years. (A) The labia minora are often rudimentary and surround the clitoris. (B) The labia are opened as indicated. (C) They are displaced posteriorly to surround the vaginal opening, giving a much more normal appearance ultimately.

augmentation or for a bowel conduit, which was commonly done in the past, because its water absorptive function must be saved. Otherwise, the patient will be forced to go through life with an ileostomy with very loose stools. Bladder augmentation can be done using stomach or small bowel. If a catheterizable conduit is needed in a patient with a continent diversion, the conduit can be made from tapered small bowel or from a ureter, if a spare one is present. These patients seldom have an appendix that can be used in the manner described by Mitrofanoff, because most have two short, rudimentary appendices.

Most surgeons agree that it is preferred to raise a genetic male as a girl if a reasonable penis cannot be constructed. The family must be prepared, nevertheless, to accept a certain degree of male behavioral imprinting seen in some of these patients despite early gonadectomy and being raised in the female gender role since birth. The alternative of raising a child as a boy, with no possibility for making a satisfactory penis in the future in the majority of cases, can be socially disastrous when the patient becomes a young adult and faces the reality of being unable to perform as a man.

Patients with cloaca often have rudimentary labia surrounding the clitoris. Figure 7 shows a method for opening that tissue and advancing it posteriorly to improve greatly the cosmetic appearance of the perineum.

Later reconstruction in cloacal exstrophy offers a great challenge. It is begun by opening the patient widely. A sound is placed through the incontinent bladder outlet, up to the dome of the bladder; the surgeon opens down on it and transects the interpubic ligament, thus creating bladder exstrophy. The soft tissues are mobilized widely from the adjacent pubis and rolled into a tube to create a urethra and bladder neck and, one hopes, satisfactory outlet resistance.

The bladder requires augmentation. Ureters must be reimplanted. This is difficult in a bladder that has a scar down its posterior midline where two hemibladders were joined at birth. In some, the reimplants are best done in an augment. This is easy to do if the augment is stomach. When there is no reasonable soft tissue with which to make a bladder neck and urethra, a technique we have used successfully in six patients is to create a bowel nipple from a segment of ileum (Fig. 8). Alternatively, a continent diversion can be created, abandoning the concept of an outlet in the perineum through which the patient will empty by self-catheterization. In fact, I regard such a reconstruction as a continent diversion whether the patient catheterizes from below or above. Only rarely have we seen a patient able to void like other children after reconstruction for cloacal exstrophy, and this must be explained to parents before surgery.

The pelvic outlet in patients with cloacal exstrophy is much different from that in patients with imperforate anus or imperforate anus and cloaca. Therefore, when performing a pull-through, I have not used the posterior sagittal approach. The point of maximal muscle contraction is identified electrically, and the surgeon dissects in through the contractile muscle complex, reaches up in the midline with a clamp, and pulls through the rectum into the center of the muscle. The posterior sagittal approach is used today for

Bowel for nipple and augmentation

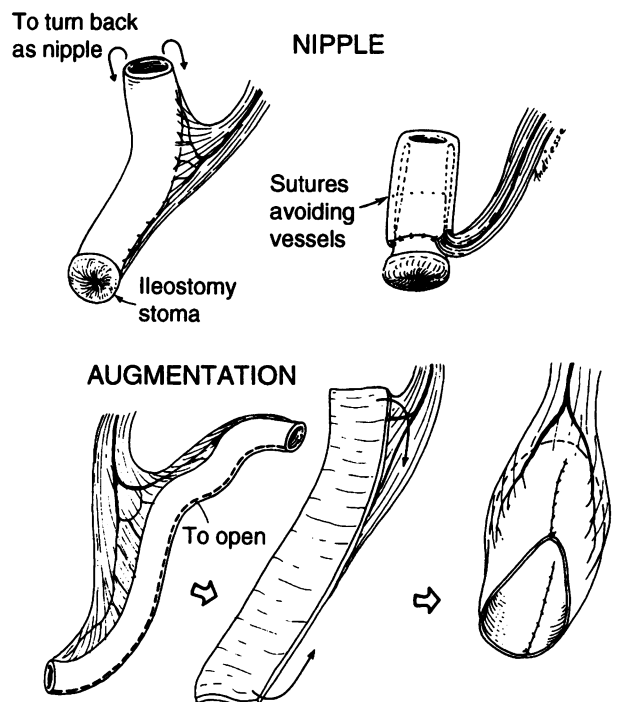


Figure 8. Creation of bowel nipple from the ileal segment is a method for attaining urinary continence when there is insufficient tissue to create a urethra and bladder neck for outlet resistance. Also shown is the technique of bladder augmentation with small bowel. Colon should never be used; it should be retained as gastrointestinal tract.

virtually all other high anorectal atresia cases except those with cloacal exstrophy.

Similarly, the vagina is a much different problem in cloacal exstrophy. It is almost always duplicated. The vaginas lie far apart and diverge laterally. In genetic females it may be easiest to mobilize only one and bring it to the perineum, using local flaps to enable it to reach, while removing the other. In genetic males, a segment of bowel is used. Because the rectum and the constructed urethra will be very close to each other, it may be better to defer making a bowel vagina during infancy. A bowel vagina made from a tube of small bowel will need to be enlarged at an older age.

RESULTS

Of 154 cloaca patients, 141 are available for current follow-up. The 13 excluded comprise 8 who have not undergone surgery, 4 who died before surgery from severe cardiac or renal disease, and 1 whom we saw in consultation only, but never for actual reconstruction. Regarding bowel control, 82 children have spontaneous bowel movements and claim satisfactory continence. Thirty-eight are on an enema program and are clean. In two of them, the enema is given through an appendicocostomy.⁹ Nine patients have a permanent colostomy. One patient who has recently developed ulcerative colitis will ultimately require an ileostomy because she is not a candidate for an endorectal pull-through. Seven young children soil and have not elected to do anything further at this point. Five patients are too recently operated to judge their bowel continence. It is not unusual for one of these patients to require enema washouts to empty the colon initially but to be able to discontinue that later.

Regarding urinary control, 83 patients void spontaneously, 40 catheterize to empty, 4 have urinary diversions, and 1 has a continent diversion. Five have wetting, which may be improved by subsequent surgery. Eight are deemed too recently operated to judge urinary control.

Twenty-four patients are now adults. Fourteen are married. Seventeen have coitus and six have had babies, five by cesarean section and one vaginally.¹⁰ There are seven patients who report no coitus yet; curiously, two of them are married. In one there is a psychological barrier to coitus; in the other couple the man has severe hypospadias but has not yet elected repair.

Results in cloacal exstrophy patients are not as good, as would be expected. Nineteen had colon pull-through, but three were later reversed because of continuing incontinence and inability to control soiling and excoriation of the perineum medically. The patients and their parents requested return to colostomy. Fifteen patients had a colostomy, which has been maintained. Seven patients have not yet had surgery. Four of the presurgical patients are older children who, together with their parents, have elected not to proceed with a major reconstructive operation, being content at present to be in diapers

for the urinary tract and to wear a bag for the colon. Urinary continence is also less well developed in these patients than in those with cloaca.

Patients who should be considered for colon pull-through are those with a good spine and sacrum, a gluteal cleft, and good perineal muscle when electrically stimulated under anesthesia. Patients with a lipomenigocele and a "rocker bottom," typical of those with severe neuromotor defect, should not be considered for a pull-through. The parents are always advised that a saline enema program will be needed to evacuate the colon and to provide freedom from uncontrolled fecal soiling. However, rarely a patient will learn to appreciate a full rectum and be able to evacuate without an enema. Rarely in cloacal exstrophy there is a normal length of colon that ends like an end colostomy between the two hemibladders; colon function may be much more normal in that instance. The three patients who elected to return to a colostomy with a bag on the abdomen had constant loose stools with perineal excoriation. They could not be medically controlled with drugs such as loperamide or enema washouts. The other successful pull-through patients are generally free from fecal soiling on an enema washout program. One child had biofeedback training and can evacuate and retain stool without enemas.

At best, the reconstructed bladder is a nondynamic pouch in patients with cloacal exstrophy, because augmentation has been necessary in all but two. Augmentation was done using stomach in 13, small bowel in 11, and both stomach and small bowel in 4. Four patients are diverted. Seven have not undergone surgery. The urinary continence mechanism was by narrowing the bladder outlet in 18; these patients rely on self-catheterization to empty. Periurethral injection of teflon has improved incontinence in some of them by increasing outlet resistance. Six patients are dry with a bowel nipple at the bladder outlet. All self-catheterize. In a seventh patient, the nipple sloughed from inadequate blood supply. That patient is currently one of six with a continent diversion. Coitus has been reported in only one of our cases to date, a genetic male who has a small bowel vagina.

The three male patients brought up as boys had that decision made elsewhere before we saw them. One is an adult who is angry and frustrated because his phallus is so rudimentary. Another was raised as a boy at the insistence of the father, who had three prior daughters and was fixated on having a son; this may well bode for social disaster in the future. The third patient has a variant of cloacal exstrophy and has a good midline phallus. In general, I favor raising the genetic male infant as a girl when there is inadequate phallic tissue with which to build a reasonable penis.

It is encouraging that 16 of these patients have no collection bag on the abdomen. Thirteen patients have one bag, a colostomy. Five patients have two bags. Seven patients have not undergone surgery.

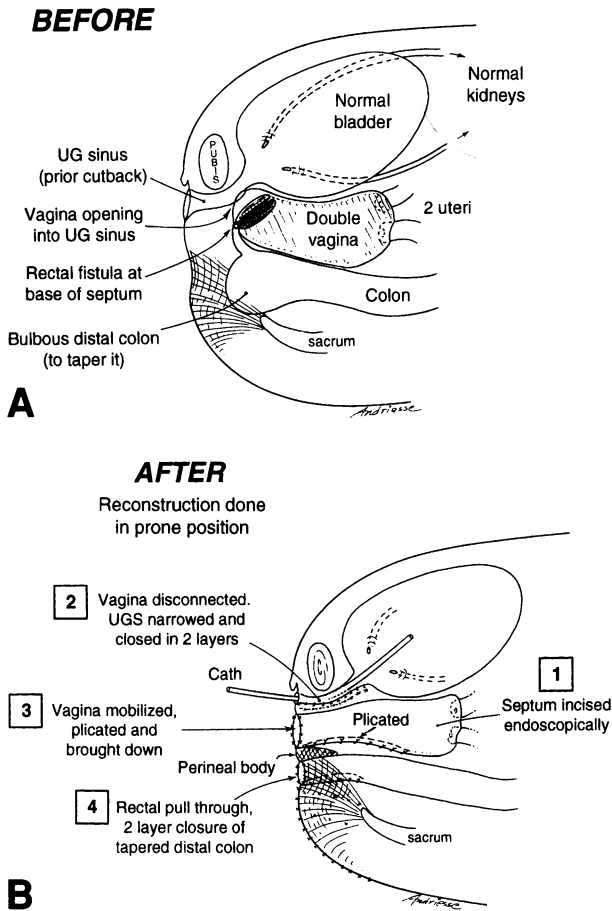


Figure 9. Case 1, a typical uncomplicated cloaca repairable through a posterior sagittal approach. (A) Before surgery. (B) After surgery.

Illustrative Cases

In previous publications we have described many details of cases.¹¹⁻²² The following six cases are selected to show typical examples of this reconstructive surgery.

Case 1 (Figs. 9 and 10) is a typical cloaca case. This infant was referred at age 16 months in 1986. Colostomy had been performed at birth. There had been severe hydronephrosis, which disappeared soon after intermittent catheterization of the vagina was instituted. The presurgical anatomy is shown in Figure 9A. At surgery the intervaginal septum was incised endoscopically, converting it to a single vagina. Through a posterior sagittal approach, reconstruction was accomplished as shown in Figure 9B. The colostomy was closed 3 months later. Figure 10 shows her appearance before surgery and 4 months after surgery. The patient is now age 14 years and has normal urinary control, bowel control, and menstrual periods.

Comment

This is a classic case with straightforward anatomy, which should produce an excellent functional result, especially when there is no neurologic problem secondary to a tethered cord or serious spinal malformations.

Case 2 (Fig. 11) shows a complex cloacal reconstruction. This patient was referred in 1984 at age 2 weeks after colostomy. There was severe bilateral vesicoureteral reflux. Hydronephrosis and pyelonephritis did not respond to intermittent catheterization, so ureteral reimplantation was performed at age 7 months. (In retrospect, I would perform it much earlier today.) Urinary infection was controlled and hydronephrosis subsided. Reconstruction was performed at age 16 months, starting in the prone position to disconnect the pelvic structures and pulling them through with a combination of exposure through the abdomen and perineum. The high rectum required mobilization from above. The small vaginas required extension to the perineum using the distal colon. This was facilitated by having intact blood supply of the left colon. The left colon was pulled through as rectum on the left colic vascular pedicle. The colostomy was closed 4 months later. Magnetic resonance imaging of the spine for tethering was normal. The patient, now age 14 years, leads an essentially normal life. Although intermittent catheterization was necessary initially, she presently voids to completion. Renal architecture is stable. She empties the colon with a warm saline enema each day and can remain clean from fecal soiling on that program. Menses are satisfactory. In the future, Z-plasty to the vaginal introitus will be performed to enlarge it.

Comment

This case illustrates the serious problem of vesicoureteral reflux seen in more than half of these patients. It underscores the need to correct that, because reflux is the life-threatening aspect of the malformation in the future if not corrected. The case shows a way to manage the colon when it is too high to reach through a posterior sagittal approach and a means for extending the vagina when it is too small to reach the perineum.

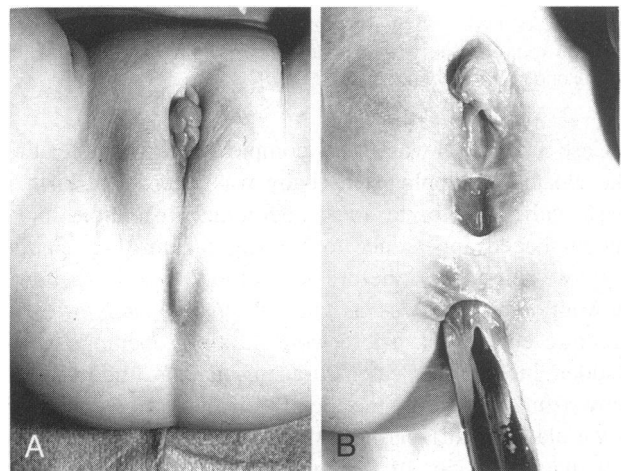


Figure 10. Case 1. (A) Presurgical view of the perineum. (B) Four months after surgery. There is a Hegar sound in the anus. The vagina is visible. There is a normal perineal body between the anus and the vagina. The rudimentary labia were later moved posteriorly at puberty to normalize the appearance of the perineum.

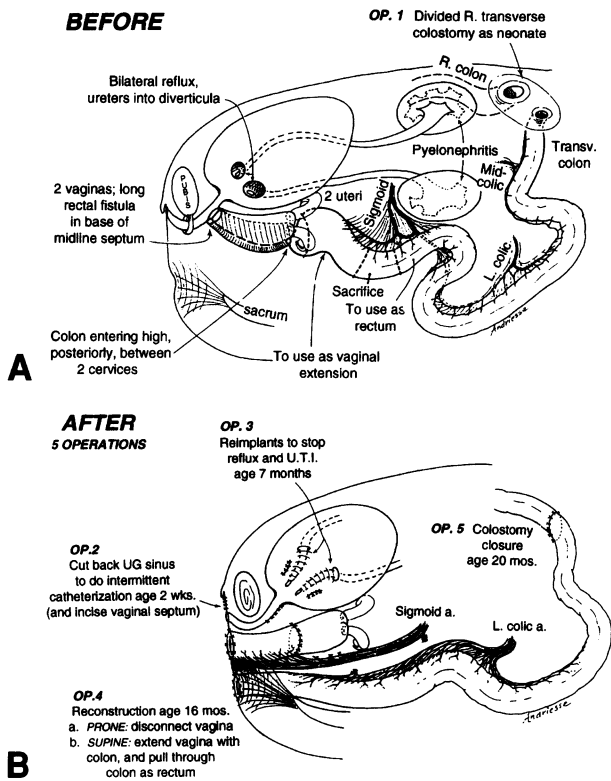


Figure 11. Case 2, a more complex cloaca. (A) Presurgical anatomy. There was severe bilateral reflux with recurrent pyelonephritis and hydronephrosis. Note two small vaginas and high rectum with long fistula. The rectum is too high to reach through a posterior sagittal operation. Note that distal rectum will be used to lengthen the vagina, performing pull-through of the next higher segment of rectum. A sigmoid colostomy would make that difficult. For this reason, a transverse colostomy is preferred at birth in these neonates. (B) After reconstructive surgery. Colostomy had been the first operation at birth. The second operation was a short cutback of the urogenital sinus to allow intermittent catheterization of the vagina, which collected urine and compressed the bladder forward (see Figure 3). Reflux and pyelonephritis were the indications to proceed next with ureteral reimplantation surgery. The major cloacal reconstruction was deferred until age 16 months. The procedure was started with the patient in the prone position, but she was turned supine to extend the vagina with colon and to pull through the colon as rectum. She was subsequently turned prone to finish the vaginal and rectal reconstruction.

Case 3 (Fig. 12) was a very complex cloaca with features like cloacal exstrophy. This baby was referred at birth in 1987 with imperforate anus, abdominal distention, and a funnel-shaped appearance to her vaginal introitus. Endoscopy was performed before laparotomy to relieve bowel obstruction. There was a wide urogenital sinus with no structure entering it. There were multiple openings in the bladder, including a patent urachus. It was impossible to know from endoscopy the identity of the various openings in the bladder. At laparotomy ileal atresia was seen. Below it there was a gap in the mesentery. There was a short segment of colon in the lower abdomen, one end of which was blind; the other end connected with the bladder. The urachus was closed. The atretic ileum was exteriorized. The blindly ending colon was also exteriorized, not knowing

whether the end was bowel that might have been in continuity with the ileal atresia or whether it was the end of the bowel, as usually seen in cloacal exstrophy. At age 7 months endoscopy was repeated, but the findings were difficult to decipher. At 15 months endoscopy was repeated with the radiologist in the operating room and with the help of a C-arm fluoroscopic unit. As each orifice was visualized and catheterized, contrast medium was injected, obtaining films. Only in that fashion was it possible to decipher the anatomy shown in Figure 12A. There were two hemibladders, four ureters, two vaginas entering the bladders, and a bowel opening in the septum between the two hemibladders. There was a diverticulum at the apex of the right hemibladder with communicating openings. Repair was performed as shown in Figure 12B at age 2 years. There were no teniae on the colon. The bladders were separated from the bladder, sewn together, and pulled through. The ureters were reimplanted. The midline septum between the

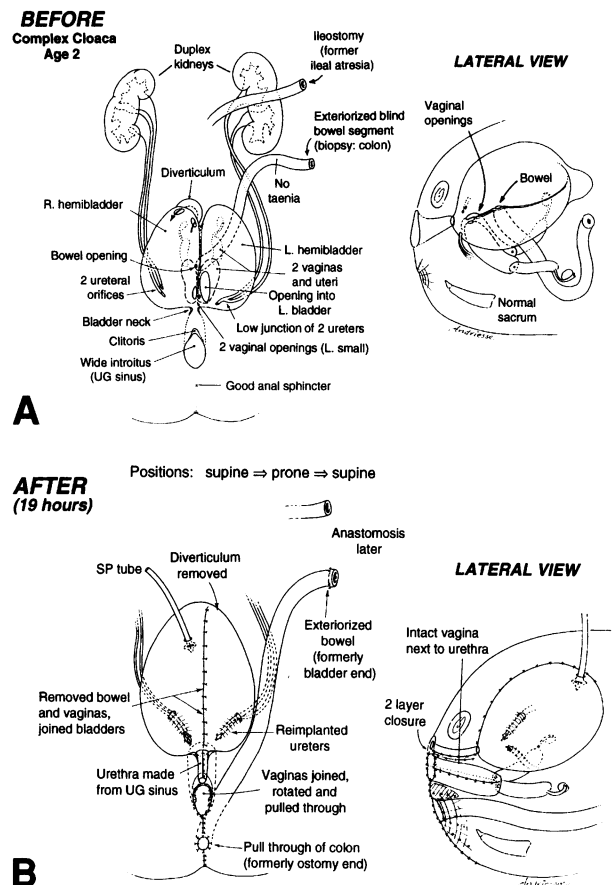
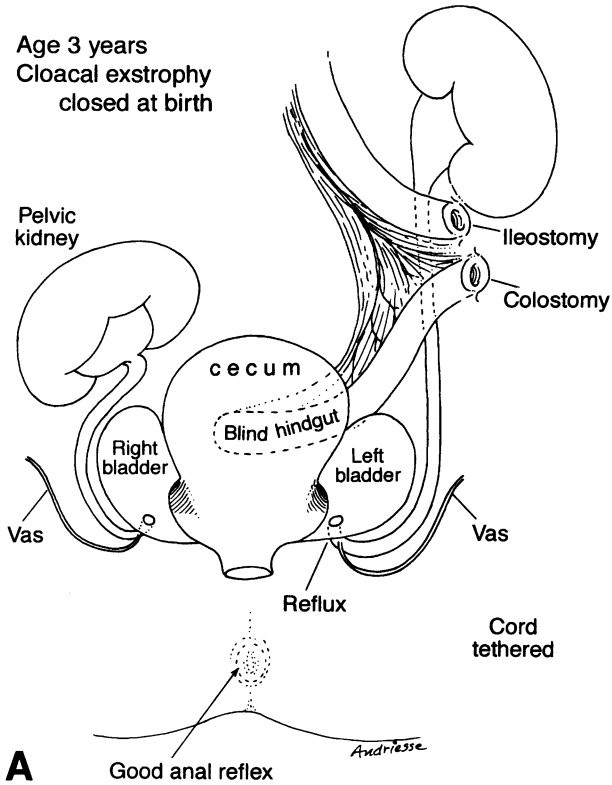


Figure 12. Case 3, a very complex cloacal malformation, illustrates the broad anatomic spectrum that can be encountered in these cases. (A) Presurgical anatomy. Note the wide urogenital sinus and the entry of small vaginas and bowel directly into bipartite bladder, a feature common in cloacal exstrophy but rare in "ordinary" cloacas. Note the blindly ending distal bowel, which had been previously exteriorized. (B) Anatomy after a long reconstructive procedure. Proper orientation of bowel to pull through was guided by intraoperative biopsy of the two ends, which had been reversed in their orientation—in other words, the ostomy end had originally been distal bowel, as seen in cloacal exstrophy.

BEFORE

Age 3 years
Cloacal exstrophy
closed at birth



AFTER

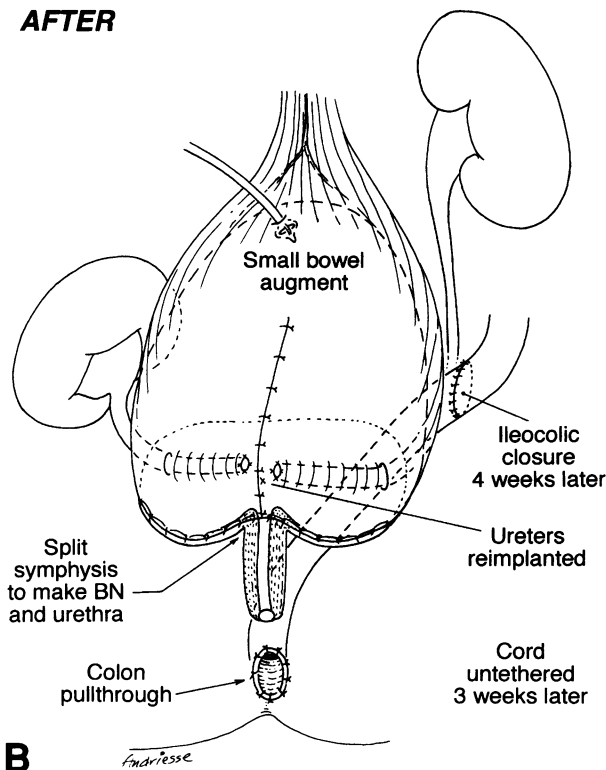


Figure 13. Case 4, cloacal exstrophy in a genetic male patient raised as a girl. (A) Presurgical anatomy. Note that cecum had been retained with bladders, but the hind gut had been detached and brought to the surface. Note the vasa entering ureters, which we have encountered in multiple cases. This patient had an excellent muscle reflex on stimulation of the perineum, making it feasible to pull through colon at the time of bladder reconstruction. (B) Anatomy after reconstruction. The bladder and urethra were laid wide open, dividing the interpubic ligament and mobilizing all soft tissue to roll into a bladder neck for continence. Ureters were reimplanted. Small bowel augmentation was performed. There was a satisfactory functional result from bladder and colon.

was pulled through as rectum. Intermittent catheterization was begun 2.5 weeks after surgery, and the suprapubic tube was soon removed. Bowel continuity was restored 4 months later. It was possible to discontinue intermittent catheterization a year later. The patient is now age 11 years. She self-catheterizes five times daily and is dry. She is clean of stool by saline washout each night, unless there is gastroenteritis with diarrhea.

Comment

This case is of special interest because the anatomy initially appeared so complex that there seemed little likelihood of achieving satisfactory reconstruction. That gloomy outlook proved incorrect. Thorough assessment of the anatomy, radiographically and endoscopically, was vital. The anatomy seemed to be a transition form between a cloaca and cloacal exstrophy because it included hemibladders, vesicointestinal fissure, and a blindly ending segment of bowel hanging in the pelvis. There was no omphalocele or bladder exstrophy.

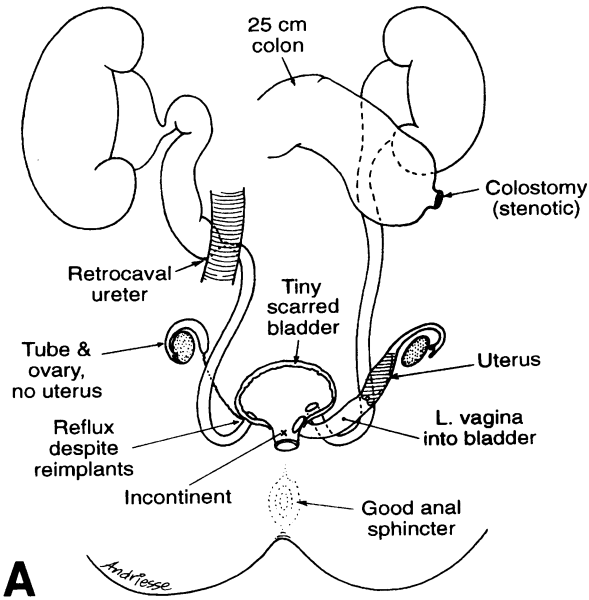
Case 4 (Fig. 13) shows cloacal exstrophy in a genetic male infant. This baby was referred at age 15 months in 1992 with the anatomy shown in Figure 13A. At birth, omphalocele closure had been performed, together with separation of the gastrointestinal tract from the bladders, although the cecum was still present between the two bladders. The reconstructive surgery was performed in 1994 at age 3 years (Fig. 13B). The spinal cord was untethered 3 weeks later by Dr. Michael Scott. One week after that, bowel continuity was reestablished. On two occasions bladder stones have formed and have been removed. The patient is dry. She is catheterized every 4 hours. She has good bowel control during the day. She receives loperamide and a fiber bulking agent for the stool. Saline enema washout is used to empty the colon.

Comment

This child copes well with life, raised in the female gender role. However, she is aggressive and at age 7 is always the leader of a group of peers. This undoubtedly represents male psychological imprinting, despite early gonadectomy and being raised in the female gender role. Long-term follow-up will be mandatory in all of these patients to note their psychosocial development. After hav-

bladders was removed. The urogenital sinus was tapered into a urethra and closed. A perineal body was constructed. The appropriate end of the bowel, identified histologically,

BEFORE
12 yr. old girl
Former cloacal exstrophy



AFTER
27 hours
1.2 L red cells
2 L albumin
9 L crystalloid

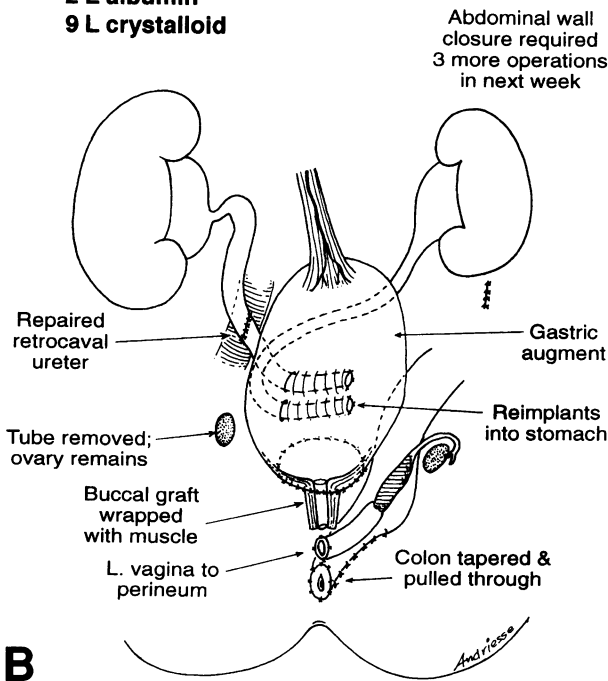


Figure 14. Case 5, a complex cloacal exstrophy case in a girl with total urinary incontinence and colostomy. (A) Presurgical anatomy showing colostomy, scarred bladder with left vagina entering bladder, absent right vagina and uterus, and retrocaval ureter with hydronephrosis. There was an excellent anal sphincter. (B) Anatomy after extensive reconstructive surgery. There was insufficient mucosa to tubularize for a urethra and bladder neck; therefore, the muscle that was present was closed around a buccal graft. The vagina was disconnected and pulled through to the perineum. The colostomy was taken down, tapered, and pulled through. The retrocaval ureter was repaired as shown. The bladder was augmented with stomach, reimplanting the ureters into the back wall of the stomach. Fluid collection in the abdominal viscera during surgery prevented primary closure of the abdominal wall. Prosthetic closure was effected, as would be performed for a large omphalocele in a neonate. The prosthetic material was removed during the next week in three stages as visceral edema subsided, creating an intact abdominal wall. Note the large amount of fluid volume replacement needed to maintain metabolic homeostasis during this global operation.

is preferred in most of these male infants who have little with which to construct a reasonable phallus.

Case 5 (Fig. 14) shows cloacal exstrophy in a girl with total urinary incontinence and a stenotic colostomy. This patient was referred in 1993 at age 12 years with the anatomy seen in Figure 14A. There was a tiny scarred bladder. The outlet was incontinent. There was reflux despite previous ureteral reimplantation surgery. There was one vagina, which entered the bladder. There was an excellent anal sphincter. In the lengthiest operation I can recall, reconstruction was performed as shown in Figure 14B. Fluid third-spacing in the abdominal viscera precluded primary closure. This was managed like a large omphalocele, closing with prosthetic material, which was removed in stages over the next week. Subsequently scoliosis surgery was performed by Dr. John Hall. Now age 15.5 years, she is dry using intermittent catheterization of the bladder and free from fecal soiling with daily enema washout. Despite orthopedic problems she is ambulatory with crutches and braces. She aspires to be a physician, and those who know her have little doubt that she will succeed in that ambition.

Comment

The importance of superb anesthesia and postsurgical intensive care with ventilatory support cannot be overemphasized. This child demonstrates what can be accomplished if the family, the patient, and the physician team are committed to the task.

Case 6 (Fig. 15) shows cloacal exstrophy in a genetic male child requiring continent diversion. This patient was referred at age 7 years in 1994 with the anatomy shown in Figure 15A. At age 4 years a reconstructive procedure had been attempted elsewhere with breakdown of the bladder outlet. A segment of colon had been used to fashion a vagina, which was placed superior to the pubis. There were no tissues below the bladder from which an outlet could conceivably be reconstructed. Continent diversion using

ing seen some adult men, raised as boys with cloacal exstrophy, in despair over having a completely inadequate penis, I am currently convinced that the female gender role

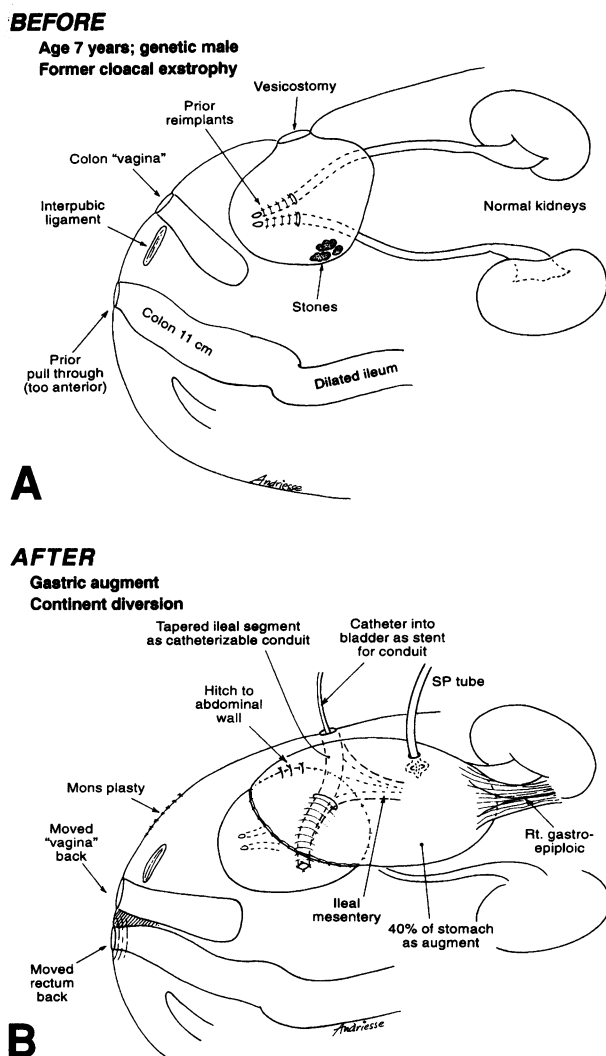


Figure 15. Former cloacal exstrophy with an exteriorized small bladder and no soft tissue from which a bladder outlet could be reconstructed. (A) Presurgical anatomy. (B) Anatomy after creating a continent urinary diversion. The appendix is not available as a catheterizable conduit in these patients because in cloacal exstrophy there are usually two rudimentary appendices attached to the cecum, which cannot be used for catheterization. Small bowel can be used as a substitute for the appendix, reimplanting one end like a ureter into the bladder so that it will not leak. The tube must be sufficiently snug to allow easy passage of a catheter, which will catch on mucosal folds if the conduit is too large. Stones seldom form in a bladder or continent diversion pouch where stomach has been used, unlike small bowel, which produces a type of mucus that often produces stones.

stomach was performed, fashioning a catheterizable conduit from tapered ileum (Fig. 15B). The patient is dry and stone-free on this regimen at age 10.5 years.

Comment

Continent diversion with self-catheterization through the abdominal wall is a reasonable goal in these complex cases. Stomach is an important part of the armamentarium for these cases for two reasons. First, its acid pH discourages infection and stone formation. It is particularly advanta-

geous if there is compromised renal function because the stomach excretes acid. Second, implantation of ureters or a catheterizable conduit can be accomplished easily in gastric wall. Neither of those can be implanted easily into a segment of ileum. Colon should be saved for the gastrointestinal tract in these cases and should not be incorporated into the urinary tract.

CONCLUSIONS

The late Willis J. Potts, former Surgeon-in-Chief at Children's Memorial Hospital in Chicago, wrote in 1959, "In general, atresia of the rectum is more poorly handled than any other congenital anomaly of the newborn. A properly functioning rectum is an unappreciated gift of greatest price. The child who is so unfortunate as to be born with imperforate anus may be saved a lifetime of misery and social seclusion by the surgeon who with skill, diligence and judgment performs the first operation on the malformed rectum."²³ These words, written four decades ago, are especially true for infants born with a cloaca or cloacal exstrophy. Results of surgery for cloaca in that era were poor.²⁴⁻²⁷ Today, a satisfactory outcome is possible for most of these children.²⁸⁻³² The best results will be attained in centers where surgeons have a special interest in this difficult type of reconstructive surgery. The same can be said about many rare and complex malformations, such as bladder exstrophy, craniofacial anomalies, and complex congenital heart defects. The problem of cloacal exstrophy is an even greater challenge.³³⁻³⁵ However, with persistence, a reasonable lifestyle can be achieved for the majority of these children.

Acknowledgments

The author thanks Dorothy M. Enos, RN, with great appreciation for her 36 years of tireless and expert assistance in the surgical correction of all the patients in this series, as well as in the continuing follow-up of these collaborative efforts; Linda Lapham, Barbara Davis, and Paula Cocoris, whose secretarial skills have been invaluable in the preparation of this paper; and Paul M. Andriese, medical artist, who has depicted clearly the pathology and surgical correction of these patients.

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Discussion

DR. JAMES A. O'NEILL, JR. (Nashville, Tennessee): Dr. Hendren is clearly the world expert on this subject of the persistent cloaca and its many variants. There are two entities that he dissected, if you get down to the basics. One is the cloaca and its multiple variations. Second is cloacal exstrophy. He stressed that there are principles of staging, while at the same time bringing in some new concepts, basically posterior sagittal anorectoplasty approaches in order to do almost complete primary repairs.

Now, it is very difficult to hear a ten-minute presentation on a subject that would take months to study. When you look at the manuscript, carefully, there is an enormous amount to be gained.

The evaluation of these anomalies is critical. And of course he uses endoscopy and a number of other things in order to make an evaluation of what needs to be reconstructed, in what order, what kind of staging, and the like. The principles are really the basics of both internal and external reconstruction. And the key here, whether he presented it strongly or not, the key is the urinary tract. Because that is the thing in the end which will kill these patients. You can do a pull-through for the imperforate anus and whether the patient is continent or not is one issue. But long-term, intelligent, well-done reconstruction of the urinary tract is key. And I think that is where Dr. Hendren and his results are clearly outstanding.

I have a couple of questions for you, Dr. Hendren. First of all, in the cloacal exstrophy group, about 50% of those patients have spinal dysraphism. A hundred percent will have tethered cord, as you pointed out. And the question comes up, really, whether any of those patients should have a pull-through. Now, you have indicated that perhaps half of them can have a pull-through. Should we use things such as MRIs to evaluate whether there are deficiencies in pelvic musculature in order to make a more considered decision about that issue? Secondly, what do you think is the ideal approach to vaginal reconstruction? Now, if you have sufficient material to pull through or to pull down the vagina from a posterior approach, well, that works pretty well. But in many of these instances it will be necessary to use some other tissue which is available. What do you think is the ideal?

DR. W. HARDY HENDREN (Boston, Massachusetts): Thank you, Dr. O'Neill. Those are two incisive questions. The first, regarding which cloacal exstrophy patients may be candidates for pull-through? They are the ones who have a good perineal muscle complex which contacts on electrical stimulation. Those babies with a rounded bottom and no gluteal cleft or contractile muscle should in most cases remain with a colostomy. Severe orthopedic disability with confinement to a wheel chair is another state which makes pull-through not feasible. Pull-through was reversed in three cases because loose stools were uncontrollable because colon length was very short. Each family is warned, therefore, that colostomy reversal may be needed in pull-through does not work out well. I have not relied on the pelvic MRI studies to make this decision.

Regarding vaginoplasty, if a vagina is present I try to free it and do a pull-through. If it is too short, the gap can be bridged by using perineal flaps or splicing in a segment of bowel to lengthen the vagina. The cardinal point is to not waste colon to make a vagina in cloacal exstrophy patients. They need every possible bit of colon mucosa for water absorption to give them a solid stool.