

Surgical Management of Cloacal Malformations: A Review of 339 Patients

By Alberto Peña, Marc A. Levitt, Andrew Hong, and Peter Midulla
New Hyde Park, New York

Background/Purpose: The aim of this study was to describe lessons learned from the authors' series of patients with cloaca and convey the improved understanding and surgical treatment of the condition's wide spectrum of complexity.

Methods: The medical records of 339 patients with cloaca operated on by the authors were retrospectively reviewed.

Results: A total of 265 patients underwent primary operations, and 74 were secondary. All patients were approached posterior sagittally; 111 of them also required a laparotomy. The average length of the common channel was 4.7 cm for patients that required a laparotomy and 2.3 cm for those that did not. Vaginal reconstruction involved a vaginal pull-through in 196 patients, a vaginal flap in 38, vaginal switch in 30, and vaginal replacement in 75 (36 with rectum, 31 with ileum, and 8 with colon). One hundred twenty-two patients underwent a total urogenital mobilization. Complications included vaginal stricture or atresia in 17, urethral strictures in 6, and urethro-vaginal fistula in 19, all of which occurred before the introduction of the total urogenital mobilization. A total of 54% of all evaluated patients were continent of urine and 24% remain dry with intermittent catheterization through their native urethra and 22% through a Mitrofanoff-type of conduit. Seventy-eight percent of the patients with a common channel longer than 3 cm require intermittent catheterization compared with 28% when their common channel was shorter than 3 cm. Sixty percent of all cases have voluntary bowel movements (28% of them never soiled, and 72% soiled occasionally). Forty percent are fecally incontinent but remain clean when subjected to a bowel management program. Forty-eight patients born at other institutions with hydrocolpos were not treated correctly during the neonatal period. The surgeons failed to drain the dilated vaginas, which interfered with the drainage of the ureters and provoked urinary tract infections, pyocolpos, and/or vaginal perforation. In 24 patients, the colostomy was created too distally, and it interfered with the pull-through. Twenty-three patients suffered from colostomy prolapse. All of these pa-

tients required a colostomy, revision before the main repair. Thirty-six patients underwent reoperation because they had a persistent urogenital sinus after an operation done at another institution, and 38 patients underwent reoperation because they suffered from atresia or stenosis of the vagina or urethra. The series was divided into 2 distinct groups of patients: group A were those with a common channel shorter than 3 cm (62%) and group B had a common channel longer than 3 cm (38%).

Conclusions: The separation of these groups has important therapeutic and prognostic implications. Group A patients can be repaired posterior sagittally with a reproducible, relatively short operation. Because they represent the majority of patients, we believe that most well-trained pediatric surgeons can repair these type of malformations, and the prognosis is good. Group B patients (those with a common channel longer than 3 cm), usually require a laparotomy and have a much higher incidence of associated urologic problems. The surgeons who repair these malformations require special training in urology, and the operations are prolonged, technically demanding, and the functional results are not as good as in group A. It is extremely important to establish an accurate neonatal diagnosis, drain the hydrocolpos when present, and create an adequate, totally diverting colostomy, leaving enough distal colon available for the pull-through and fixing the colon to avoid prolapse. A correct diagnosis will allow the surgeon to repair the entire defect and avoid a persistent urogenital sinus. Cloacas comprise a spectrum of defects requiring a complex array of surgical decisions. The length of the common channel is an important determinant of the potential for urinary control, and predicts the extent of surgical repair.

J Pediatr Surg 39:470-479. © 2004 Elsevier Inc. All rights reserved.

INDEX WORDS: Cloaca, anorectal malformation, cloacal malformation, hydrocolpos, vaginal replacement, total urogenital mobilization.

BEFORE 1982, the available information related to the surgical repair of persistent cloaca was scant. The few published articles in the English-language liter-

ature¹⁻⁷ contained very limited information about the wide spectrum of anatomic characteristics of this malformation.

The first posterior sagittal repair of a cloacal malformation was performed in 1982. A preliminary report of our first 54 cases was published in 1989.⁸ We have now operated on 339 patients for this complex anorectal malformation. The lessons learned from this large experience have been significant. Our experience represents a great opportunity to learn about the anatomy of these malformations and to correlate the anatomic characteris-

From Schneider Children's Hospital, New Hyde Park, NY.

Presented at the 34th Annual Meeting of the American Pediatric Surgical Association, Fort Lauderdale, Florida, May 25-28, 2003.

Address reprint requests to Alberto Peña, MD, Schneider Children's Hospital, 269-01 76th Ave, New Hyde Park, NY 11040.

© 2004 Elsevier Inc. All rights reserved.

0022-3468/04/3903-0031\$30.00/0

doi:10.1016/j.jpedsurg.2003.11.033

tics with the clinical manifestations and the final functional prognosis.

The most important lesson that we have learned is that we are dealing with a very wide spectrum of defects. It is vital to understand the anatomic complexity and variations as well as the associated urologic problems that must be addressed in the majority of these patients. Dr Hardy Hendren's publications⁹⁻¹⁷ represent a major contribution and the most comprehensive reading material available. Long-term follow-up of these patients has allowed us to learn about long-term gynecologic concerns.¹⁸ We also have come to understand important therapeutic dilemmas that the pediatric surgeon faces when dealing with these patients in the neonatal period and during the surgical repair. Seventy-four of the patients in our series were referred to us for revisional surgery after failed primary repairs. An analysis of these cases has brought to light common errors that occur in the management of these children.

Our overall experience has allowed us to identify 2 major subgroups of cloaca patients. On the more benign side of the spectrum of these defects (group A), we found that 62% of the patients operated on by us primarily had a common channel shorter than 3 cm as well as a significantly lower incidence of associated defects. The repair of this group of cases is considerably less difficult than its counterpart. On the complex side of the spectrum (group B), we found that 38% of the patients had common channels longer than 3 cm, with a conspicuously high incidence of associated defects and complex anatomy. The repair of this latter group required long, meticulous operations, and the functional results were clearly inferior to the first group. We believe that early identification of these 2 categories is important for surgical planning. This report represents an effort to communicate all of these experiences for the benefit of babies born with cloaca and the pediatric surgeons dealing with these serious technical challenges.

MATERIALS AND METHODS

The medical records of 339 patients with persistent cloaca operated on by us were retrospectively reviewed. The follow-up was performed, by phone, by letter, or by personal visit at our clinic. Special emphasis was placed on the characteristics of the malformation, length of the common channel, complexity of the operation, and the final functional result, including voluntary bowel movements, soiling of underwear, urinary control, intermittent catheterization, either through their native urethra or through a Mitrofanoff-type of conduit in cases that required a continent diversion. The report was prepared following the rules and guidelines of the hospital Institutional Review Board.

RESULTS

A total of 265 patients underwent primary operations. Seventy-four were secondarily operated on by us after the patients underwent an attempted but failed repair at

Table 1. Vaginal Reconstruction (n = 339)

Vaginal pull-through	196
Total urogenital mob. only	84
Total urogenital mob. plus*	38
Vagino-urethoplasty	74
Vaginal replacement	75
Rectum	36
Small bowel	31
Colon	8
Flaps	38
Vaginal flap	34
Skin/labial flap	4
Vaginal switch	30

*Additional maneuvers to complete the repair.

other institutions. A total of 111 patients required a laparotomy in addition to the posterior approach. The average common channel of the malformation was 4.7 cm for patients that required a laparotomy and 2.3 cm for those that did not. The vaginal reconstruction of the malformation involved a vaginal pull-through in 196 patients, creation of a vaginal flap in 38, a vaginal switch maneuver in 30, and a vaginal replacement in 75 cases (utilizing rectum in 36, terminal ileum in 31, or colon in 8 cases; Table 1). One hundred twenty-two patients, during the last 11 years, underwent a total urogenital mobilization¹⁹ as a part of the surgical repair of the malformation. Complications included vaginal stricture or atresia in 17 patients, urethral stricture or atresia in 6 cases, and urethral vaginal fistula in 19. All of these complications occurred before the introduction of the total urogenital mobilization maneuver.

A total of 193 patients could be evaluated for urinary continence and 156 for fecal continence. The remaining group was not included either because they were less than 3 years of age, were lost to follow-up, or we lacked reliable information.

Fifty-four percent of all evaluated patients are continent of urine. Twenty-four percent remain dry with intermittent catheterization through their native urethra, and 22% use a Mitrofanoff-type of continent diversion (Table 2). Seventy-eight percent of the patients with a common channel longer than 3 cm require intermittent catheterization to empty their bladders (through a native urethra, or a Mitrofanoff type of conduit). A total of 28% of the patients that had a common channel shorter than 3 cm require intermittent catheterization to empty their bladder (Table 2). Sixty percent of all cases have voluntary bowel movements (28% of them never soil, and 72% of them soil occasionally). Forty percent are fecally incontinent, but all remain clean when subjected to our bowel management program (Table 3).²⁰

A small group of patients (19 cases) have a malformation with a very short common channel (1 cm), an

Table 2. Urinary Continence (n = 193)

	No. of Patients	%
Continent	104	54
Dry with intermittent catheterization	89	46
Through native urethra (24%)		
Continent diversion or Mitrofanoff (22%)		

NOTE. Intermittent catheterization and length of common channel: long (>3 cm), 78%; short (<3 cm), 28%.

excellent functional prognosis for urinary function, and a relatively good prognosis for bowel control (Table 4).

Several clinically important issues arise during the neonatal period for these patients. One of the most significant we found was that 77 patients were born with hydrocolpos. Twenty-nine of these babies had hydrocolpos successfully detected and were treated during the neonatal stage. In the remaining 48 patients, the hydrocolpos was not identified or was mismanaged. In these cases, the surgeons were prompt to open a colostomy but did not drain the dilated vagina. Three of these patients suffered from vaginal infection (pyocolpos), and 2 suffered from vaginal perforation, developing an acute abdomen with severe peritonitis. In the remaining 43 patients, the vaginas remained distended and interfered with the drainage of the ureters, producing persistent bilateral hydronephrosis and megaureters. The patients suffered from urinary tract infections, persistent acidosis, and lack of growth or underwent potentially unnecessary urinary drainage procedures such as nephrostomy, ureterostomy, cystostomy, or vesicostomy. All of these symptoms disappeared when the vagina was finally decompressed by performing a vaginostomy.

In 24 patients, the colostomy was created too distally and interfered with the pull-through of the rectum at the time of the main repair. Twenty-three patients suffered from colostomy prolapse, particularly when the colostomy was created at a mobile portion of the colon (9 cases) and when a loop colostomy was used (14 cases). All of these patients required a colostomy revision before the main repair.

Thirty-six patients required a reoperation because they had a persistent urogenital sinus after the original operation done at another institution. All of these patients were operated on with the misdiagnosis of "rectovaginal fistula," and thus the rectum was moved, but the urogenital sinus was not repaired. The reoperation consisted of

Table 3. Fecal Continence (n = 156)

	No. of Patients	%
Voluntary bowel movements	94	60
72% occasional soiling		
28% totally continent		
Fecally incontinent*	62	40

*Clean with bowel management program.²⁰

Table 4. Cloaca Type 1—Clinical Results (1 cm Common Channel; n = 13)

	No. of Patients	%
Urinary continent	13	100
Voluntary bowel movement	10	77
Totally continent*	7	54

*Voluntary bowel movement with no soiling.

a complete redo, posterior sagittal approach, rectal mobilization, and dealing with the urogenital sinus in the same way that we treat a cloaca. Thirty-eight patients underwent reoperation by us because they suffered from atresia or stenosis of the vagina or urethra after an attempted failed operation done at another institution.

Surgical Technique

During the first 14 years of this experience, the standard approach used by us consisted of a posterior sagittal operation, with or without a laparotomy depending on the complexity of the defect. During the posterior sagittal approach, the rectum was separated from the urogenital sinus; the vagina was then separated from the urethra in an attempt to bring both structures (urethra and vagina) down to be placed in a normal location. What used to be the common channel was then reconstructed as a neourethra.

During the last 11 years, we have used a new maneuver called "total urogenital mobilization."¹⁹ With this technique, the rectum was separated as previously done, but then the vagina and the urethra were mobilized together as a unit down to the perineum (Fig 1). We found that this maneuver shortened the operating time approximately 50% to 70%. The complications of urethral and vaginal strictures were eliminated because the blood supply of these 2 structures, when kept together, was excellent, and the cosmetic result was better. The functional results obtained in the group of patients subjected to this last maneuver were similar to the results obtained with our previous technique (Table 5).

We have learned that when a patient has a common channel longer than 3 cm, the likelihood of repairing the entire defect via a posterior sagittal approach alone decreases significantly. Under these circumstances, it is usually mandatory to open the abdomen to complete the repair of the malformation. Once the surgeon opens the abdomen, he or she becomes involved in a complex decision-making algorithm to repair these complex defects. The bladder must be opened, and ureteral catheters must be introduced in each ureter to avoid damage to them because the common wall that separates the bladder from the vagina involves a significant portion of both ureters. The vagina must be separated from the urinary tract, which is one of the most tedious and delicate maneuvers in the repair of these defects, and requires a

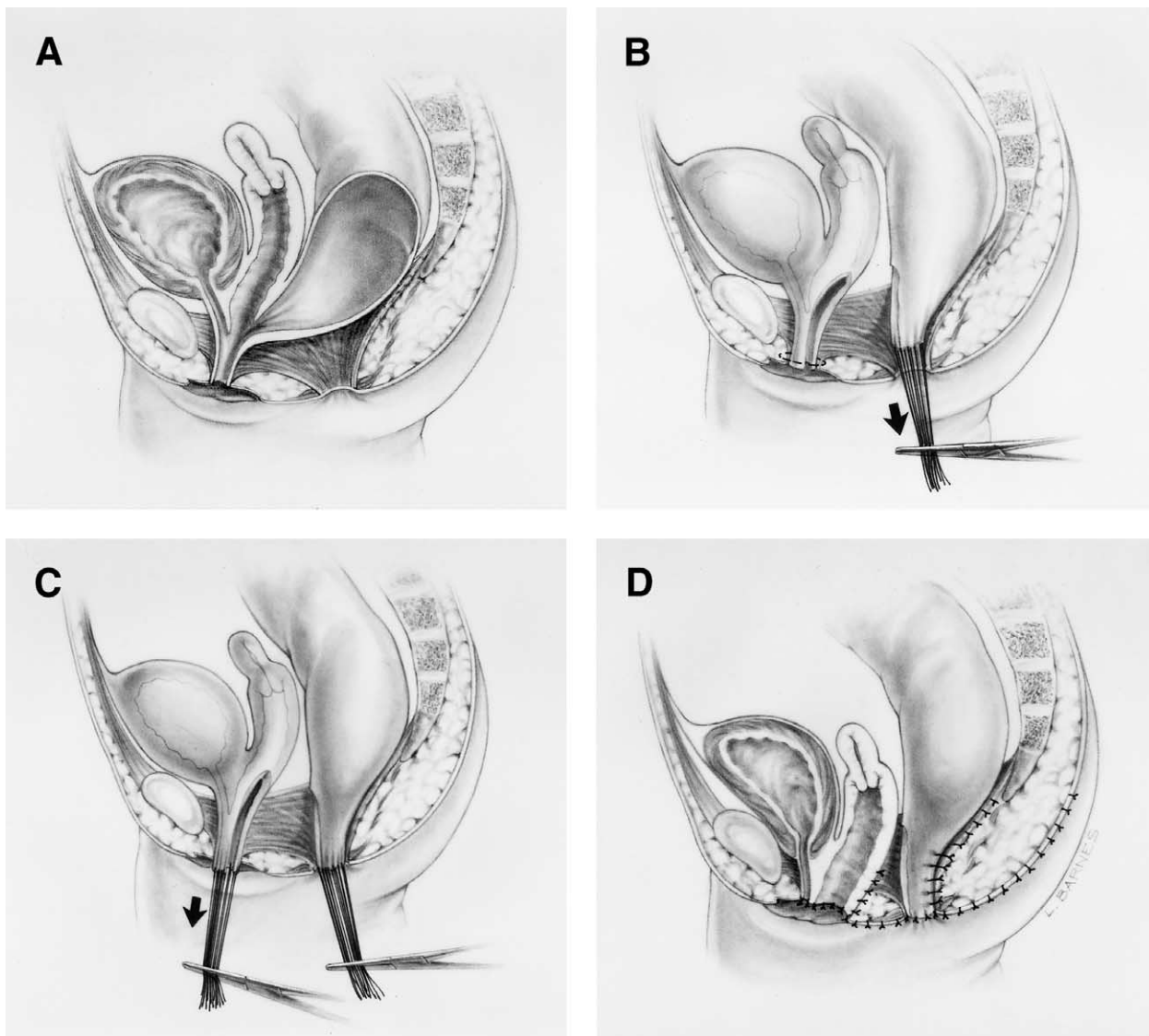


Fig 1. Total urogenital mobilization. (A) Sagittal view of a cloaca, (B) rectal separation and mobilization, (C) urogenital sinus was completely divided and is being mobilized, (D) operation completed. Reprinted with permission.¹⁹

meticulous technique, finesse, patience, and dedication. At that point, the surgeon must look at the characteristics of the vagina, as well as its size, to determine the best way to repair the genital component of the malformation.

When both vaginas are very dilated (bilateral hydro-

colpos), and the patient has a very long common channel, a technical maneuver called “vaginal switch” seems to be ideal to repair this specific defect (Fig 2).²¹ In other cases, the vagina, once separated from the urinary tract, can be mobilized down to the perineum without the vaginal switch maneuver. It is important for the surgeon to have enough experience to determine which maneuver is appropriate for which vaginal anatomy.

When the patient has very short vaginas, located very high in the pelvis, then it is necessary to perform some sort of vaginal replacement that can be done with the rectum (Fig 3), colon (Fig 4), or a small bowel (Fig 5) depending on the specific anatomic situation. If the patient has a wide rectum, one can replace a vagina with a portion of it, longitudinally separating the neovagina

Table 5. Urinary Continence and Total Urogenital Mobilization (TUM)

	TUM (n = 49)		Without TUM (n = 72)	
	No.	%	No.	%
Urinary continent	28	58	43	60
Dry with intermittent catheterization	21	42	29	40
Through native urethra		22		22
Diverted or Mitrofanoff		20		18

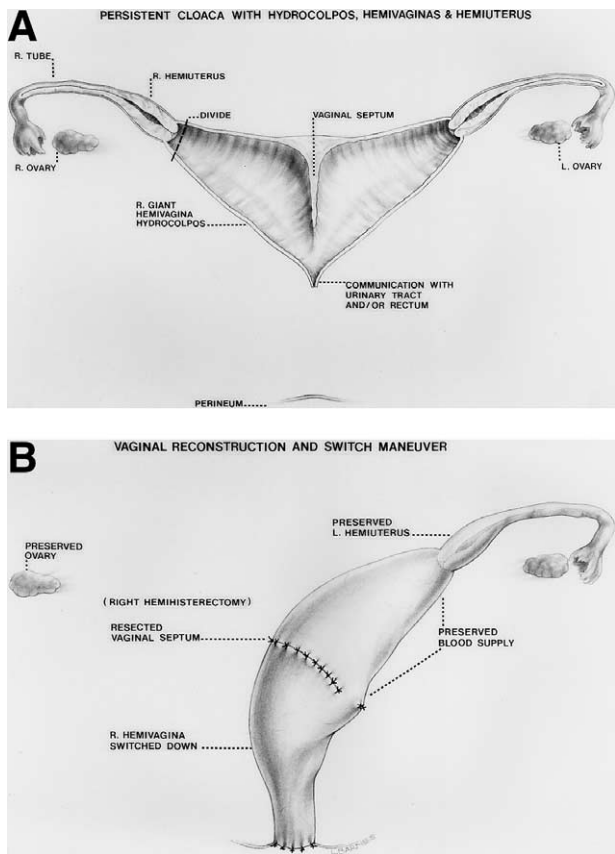


Fig 2. Vaginal switch. (A) Two large dilated hemivaginas, 2 hemiuterus, long common channel. (B) The right müllerian structures were resected, the ovary was preserved, the right hemivagina was switched down, the blood supply of the left hemivagina was preserved.

and preserving its blood supply (Fig 3). When the patient has a colostomy that interferes with the use of the colon or if the patient has a tendency to suffer from diarrhea, then one has to use the small bowel to replace the vagina.

On the most extreme end of this spectrum of malformations, one can find patients that have 2 hemivaginas attached to the bladder neck (Fig 6). In addition, the rectum may also open into the bladder neck. When one separates these structures, the patient is left with no bladder neck. At that point, the pediatric surgeon or urologist must decide whether that bladder neck is worth reconstructing or if the patient would do better with a permanent closure of the bladder neck and opening of a temporary vesicostomy. In the latter situation, the patient will eventually require some sort of continent diversion that usually includes a bladder augmentation, reimplantation of the ureters, and a creation of a Mitrofanoff type of conduit.

It is vital to know in advance whether the patient suffers from vesicoureteral reflux. If the patient belongs to the group that requires a laparotomy, the main oper-

ation may represent the ideal time to reimplant the ureters. On the other hand, if the patient belongs to the short common channel group, the malformation can be repaired posterior sagittally, without opening the abdo-

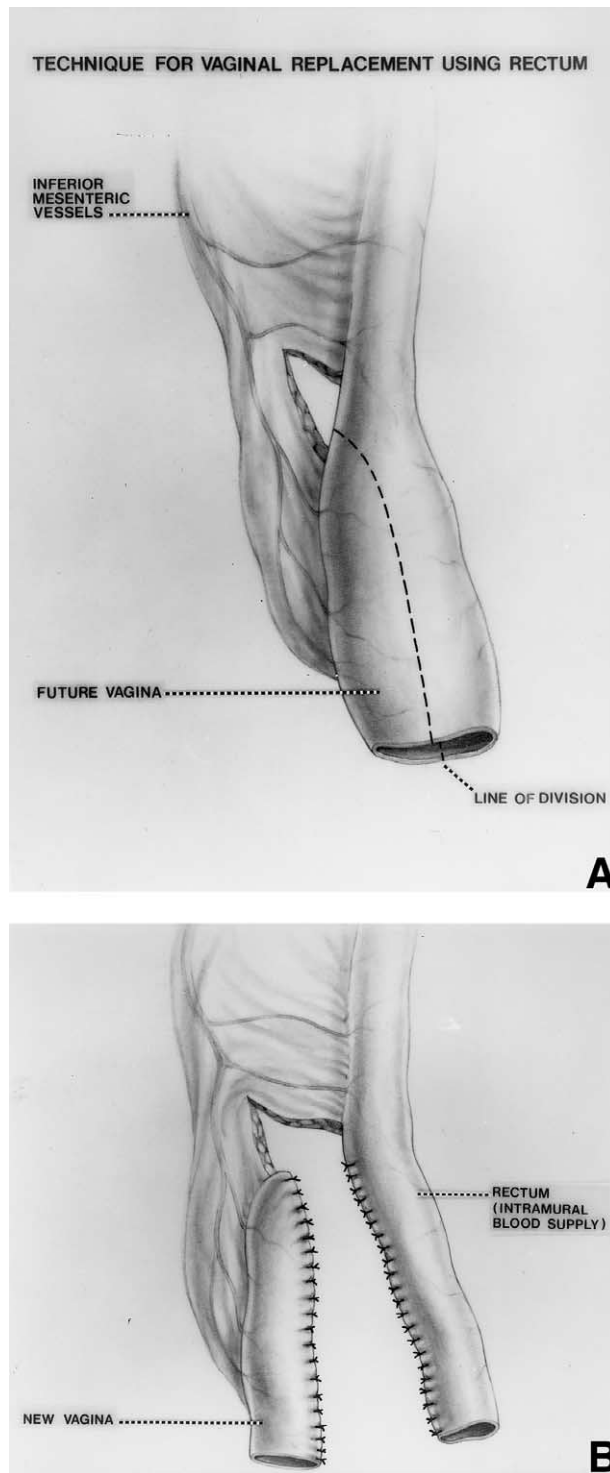


Fig 3. Vaginal replacement with rectum. (A) Longitudinal incision of a dilated rectum. (B) Neovagina and rectum created.

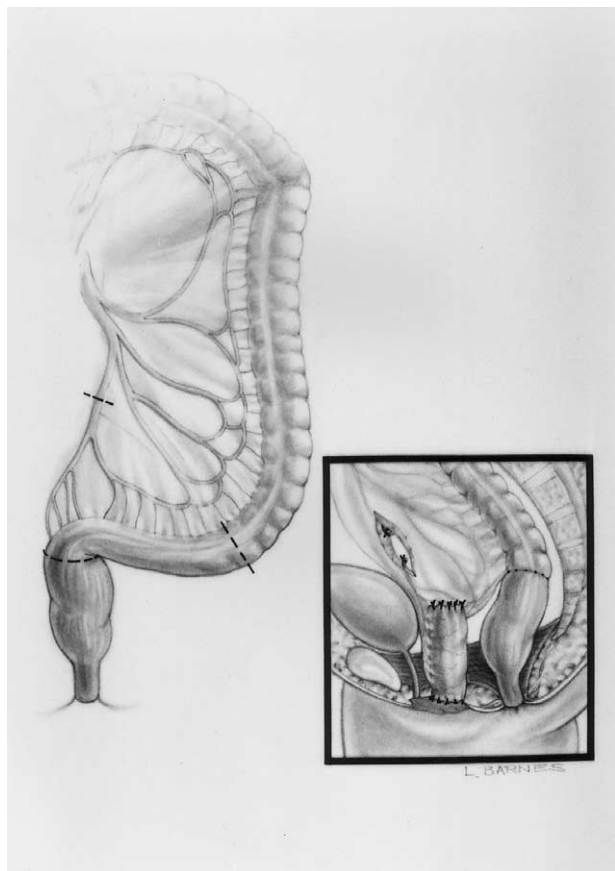


Fig 4. Vaginal replacement with colon.

men; and in such a case, the ureteral reimplantation can be performed at a later date.

The incidence of vesicoureteral reflux in patients with cloaca is extremely high (51%). The combination of the incapacity to empty the bladder and vesicoureteral reflux is a dangerous one. These patients suffer from frequent episodes of urinary tract infections and kidney damage. Five of our patients suffered from chronic renal failure, all of whom belong to the complex type of defects, and they were born with significant renal damage.

In the group of 19 patients with a 1-cm common channel, it is better to repair the rectal component and to perform only a vaginoplasty or introitoplasty without separating the vagina from the urinary tract and without total urogenital mobilization. The patient is left with a slight female hypospadias, which is clinically irrelevant.

The duration of the operation of the main repair in cloaca patients varies depending on the length of the common channel and the complexity of the malformation. In patients with a common channel shorter than 3 cm, the operation may last approximately 3 hours, the patient stays in the hospital for 2 days, and the functional results are very good. When the common channel is longer than 3 cm, the patient has a complex malforma-

tion with associated defects such as bilateral hydrocolpos or vaginas implanted in the bladder neck, megaureters, vesico-ureteral reflux, hydronephrosis, ectopic ureters, or atretic mullerian structures. These operations usually last from 6 to 12 hours (Table 6).

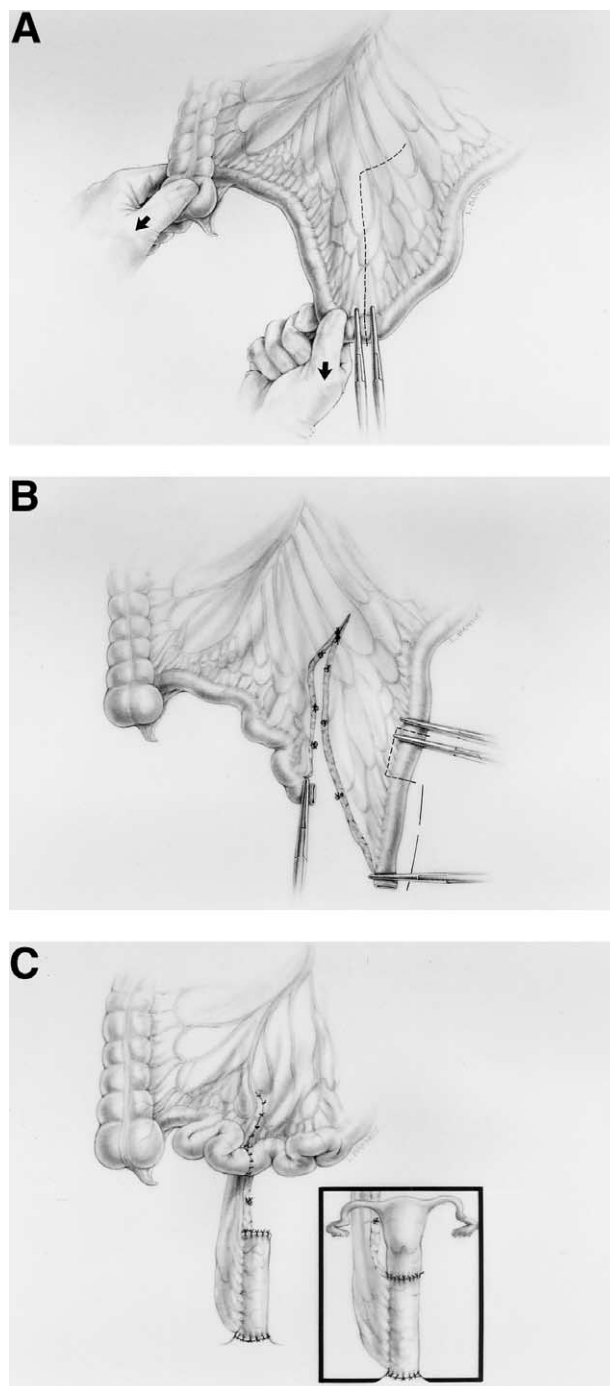


Fig 5. Vaginal replacement with small bowel. (A) Determine point of longest mesentery to divide the bowel, (B) divide vessels to gain length preserving the blood supply, (C) neovagina anastomosed to vaginal remnant or closed blind.

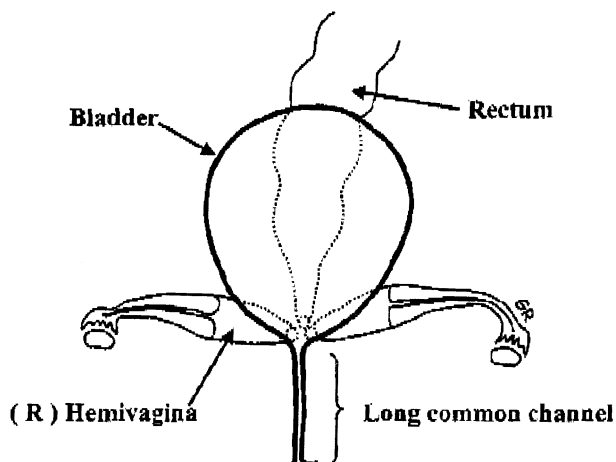


Fig 6. Complex cloaca that includes a very long common channel, 2 small hemivaginas, and a rectum connected to the bladder neck.

At the beginning of our experience, we repaired cloacas when the patients were 1 year of age. As we gained more experience in the management of these defects, we operated on them earlier and earlier. Currently, we operate on these babies when they are 1 month old, provided they have shown that they are growing and developing normally.

DISCUSSION

The retrospective analysis of this large experience allows us to detect 2 distinct groups of patients. We believe that the recognition of these 2 groups is important for the pediatric surgical community and, most importantly, for the patients. The first group is represented by patients with cloacas with a common channel shorter than 3 cm. The incidence of associated urologic defects in this group is 59%. The operating time, the type of operation required to repair these malformations, as well as the incidence of this subtype (more than 62%) make us believe that most pediatric surgeons can be trained to satisfactorily repair these types of defects. The

total urogenital mobilization is a reproducible maneuver that allows the successful repair of this group of malformations (Table 6).

The other group of cloacas represents 38% of our total and are those who were born with common channels longer than 3 cm and the series of anatomic features already described. Ninety-one percent of them suffer from associated urologic defects. The complexity of the operations required to repair these malformations requires not only a specialized pediatric surgeon with a large experience in these defects, but also someone with special training in pediatric urology to confront and solve all the challenges faced in the management of these cases. Table 6 shows the main differences between these two groups of patients.

We believe that a pediatric surgeon dealing with a patient with a cloaca should try to determine to which group the patient belongs. This is best done with a diagnostic cystoscopy and vaginoscopy. If he or she has a large experience in the management of these cases and also has special pediatric urology training, or works with a pediatric urologist, he or she can approach the complex group. Without this level of expertise, it is better to transfer the baby to someone with that kind of experience. On the other hand, we believe that in all training programs, special emphasis should be placed on the training of pediatric surgical trainees in the management of cloacas with a common channel shorter than 3 cm. The number of patients that are born in every country, plus the fact that the technique is reproducible, makes us believe that this is feasible.

It is extremely important to follow up with these patients on a long-term basis because of their many potential problems. The patients may suffer from constipation that must be treated aggressively, because we know that constipation produces overflow pseudoincontinence. Also, the patients always have the potential to suffer from urinary tract infections. The patients that received some sort of continent urinary diversion are

Table 6. Operation Data

	Group A	Group B
Common channel	Short, <3 cm	Long, >3 cm
Type of operation	Only posterior sagittal	Posterior sagittal and laparotomy
Length of procedure	3 h	6-12 h
Postoperative care	48 h	Several days
% Associated urologic defects	59%	91%
Incidence in our series	62%	38%
Voluntary bowel movements	68%	44%
Urinary continence	72%	28%
Average number of operations*	9	18
Intraoperative decision making	Relatively easy, reproducible operation	Complex, delicate and technically demanding†

*Including orthopedic, urologic, cardiac, and general.

†Bladder/vagina separation, ureteral catheter, ureteral re-implantation, vesicostomy, cystostomy, bladder neck reconstruction or closure, vaginal switch, vaginal replacement (with rectum, colon, small bowel).

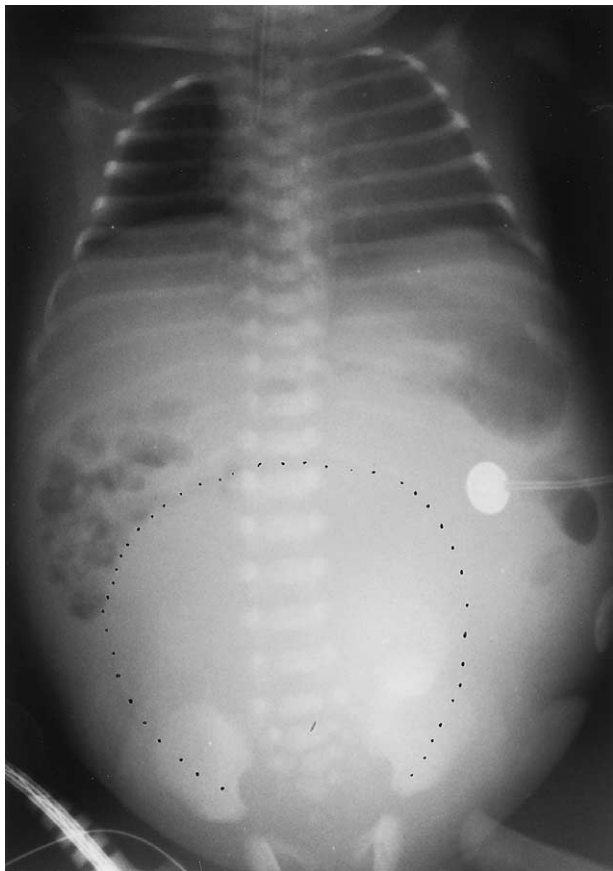


Fig 7. Plain abdominal film of a baby with hydrocolpos.

patients for life. They require supervision concerning the metabolic problems inherent to a continent diversion, evaluation of renal function, stone formation, potential development of vesicoureteral reflux, and urinary tract infections.

The patients must also be followed up with closely looking for gynecologic and obstetric problems. We already detected a significant number of patients who reached adolescence and suffered from the incapacity to drain the menstrual flow. They developed large collections of menstrual blood trapped in the peritoneum owing to different types of atresias of the müllerian system.¹⁸ We now try to detect these atresias early in life. We intubate and irrigate the fallopian tubes to be sure that they are patent. We do this as soon as we have the opportunity to be inside the abdomen in one of these patients either during the main repair or during the colostomy closure.

Thus far, 2 of our patients have delivered babies vaginally, they belong to the group of the good prognosis type of cloaca. We do not yet have any of the complex cloaca patients that have become pregnant; therefore, we cannot give an opinion about this. In the coming years,

much will be learned as we continue to follow up with these patients.

It is imperative for pediatric surgeons dealing with cloacas to be aware of the fact that a very dilated vagina is a significant problem for these newborn babies. During the neonatal period, the baby should not be taken to the operating room until the urinary tract is adequately evaluated, and the presence of a hydrocolpos has been ruled out. If the baby has a hydrocolpos, it is mandatory for the surgeon not only to open a colostomy but to insert tubes into the dilated vagina or vaginas to decompress them and thus prevent complications, such as pyocolpos or ureteral obstruction (Figs 7 & 8). In a newborn baby with cloaca that suffers from bilateral or unilateral hydronephrosis, before making a decision about creating a ureterostomy or a nephrostomy, the pediatric surgeon and pediatric urologist must rule out the presence of hydrocolpos that is interfering with the drainage of the ureters. Vaginal drainage may be all that the patient needs to decompress the urinary tract.

If an infant with a cloaca is not growing well in the first weeks or months of life after a colostomy, it is most likely because a complicating problem has been missed. Most commonly, the hydrocolpos was not drained, which interferes with the ureteral drainage. There are some patients that suffer from an incapacity to empty the

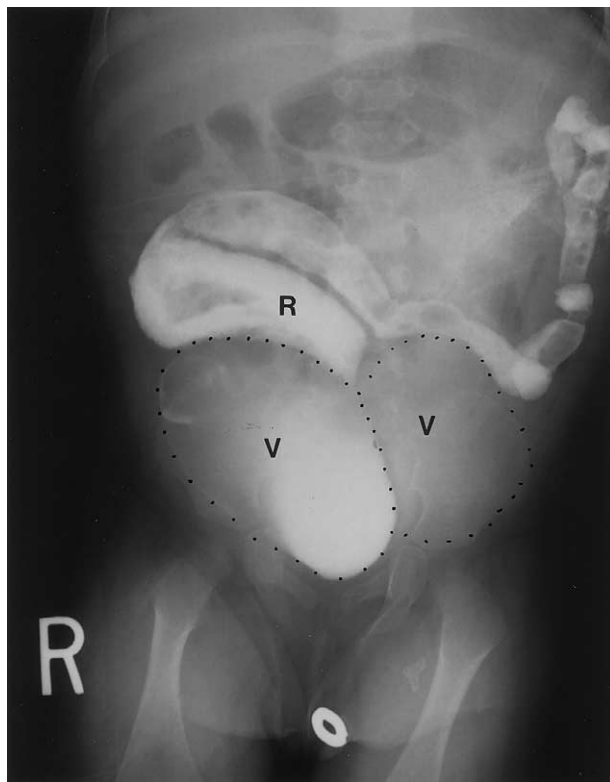


Fig 8. Distal colostogram performed in a baby with severe, bilateral hydrocolpos. R, rectum; V, vagina.

bladder owing to an almost atretic urethra and do require some sort of bladder drainage.

The preferred colostomy is one with separated stomas, being sure to leave enough distal bowel to not interfere with the pull-through. If the colostomy is performed in a mobile portion of the colon, it is very important to fix the colon to the abdominal wall to prevent prolapse, which can lead to serious complications.

The pediatric surgeon should try to endoscopically measure the length of the common channel as early as possible to try and establish the functional prognosis for

the patient, which will help the parents adjust their expectations. From a therapeutic point of view, this will help the surgeon determine whether the patient is going to be treated by him or her alone, in conjunction with a pediatric urologist, or whether the patient should go to a referral center that specializes in pediatric colorectal problems.

ACKNOWLEDGMENT

The authors thank George Rodriguez and Elyse Bellefond for their help in preparation of the manuscript.

REFERENCES

1. Gough MH: Anorectal agenesis with persistence of cloaca. *Proc R Soc Med* 52:886-889, 1959
2. Bock JE, Madsen CM: Anorectal atresia with rectocloacal fistula. *Acta Chir Scand* 137:284-286, 1971
3. Palken M, Johnson RJ, Derrick W, et al: Clinical aspects of female patients with high anorectal agenesis. *Surg Gynecol Obstet* 135:411-416, 1972
4. Cheng GK, Fisher JH, O'Hare KH, et al: Anomaly of the persistent cloaca in the female infants. *AJR* 120:413-423, 1974
5. Kay R, Tank ES: Principles of management of the persistent cloaca in the female newborn. *J Urol* 117:102-104, 1977
6. Sieber EK, Klein R: Cloaca with non-adrenal female pseudo hermaphroditism. *Pediatrics* 22:472-477, 1958
7. Raffensperger JG, Ramenofsky ML: The management of cloaca. *J Pediatr Surg* 8:647-657, 1973
8. Peña A: The surgical management of persistent cloacas. Results in 54 patients treated with the posterior sagittal approach. *J Pediatr Surg* 24:590-598, 1989
9. Hendren WH: Surgical management of urogenital sinus abnormalities. *J Pediatr Surg* 12:339-357, 1977
10. Hendren WH: Urogenital sinus and anorectal malformation: Experience with 22 cases. *J Pediatr Surg* 15:628-641, 1980
11. Hendren WH: Further experience in reconstructive surgery for cloacal anomalies. *J Pediatr Surg* 17:695-717, 1982
12. Hendren WH: Repair of cloacal anomalies: Current techniques. *J Pediatr Surg* 21:1159-1176, 1986
13. Hendren WH: Urological aspects of cloacal malformations. *J Urol* 140:1207-1213, 1988
14. Hendren WH: Cloacal malformations: Experience with 105 cases. *J Pediatr Surg* 27:890-901, 1992
15. Hendren WH: Urogenital sinus and cloacal malformations. *Semin Pediatr Surg* 5:72-79, 1996
16. Hendren WH: Management of cloacal malformations. *Semin Pediatr Surg* 6:217-227, 1997
17. Hendren WH: Cloaca, The most severe degree of imperforate anus: Experience with 195 cases. *Ann Surg* 228:331-346, 1998
18. Levitt MA, Stein DM, Peña A: Gynecological concerns in the treatment of teenagers with cloaca. *J Pediatr Surg* 33:188-193, 1998
19. Peña A: Total urogenital mobilization: An easier way to repair cloacas. *J Pediatr Surg* 32:263-268, 1997
20. Peña A, Guardino K, Tovilla JM, et al: Bowel management for fecal incontinence in patients with anorectal malformations. *J Pediatr Surg* 33:133-137, 1998
21. Peña A: Anorectal anomalies, in Spitz L, Coran AG (eds): *Rob & Smith's, Operative Surgery*. ed 5. London, Chapman & Hall Medical, 1995, pp 423-452

Discussion

D. Lund (Madison, WI): Dr Hendren asked me to read some comments, and the challenge for me is going to be to read this without sounding too much like Dr Hendren.

"I have asked Dr Dennis Lund to read my discussion remarks because with apology I am en route to my granddaughter's school graduation in California.

Dr Peña has asked if I would discuss his paper and kindly presented it to me for preview yesterday on his laptop computer.

This is a huge experience by an exceptional surgeon. The conclusions are similar to my own experience with about 220 cases since 1962. The lethal problem with cloaca is the urinary tract. I would underscore what Dr Peña has said that the pull-through anorectoplasty should never be done first leaving the urogenital sinus malfor-

mation until later. A large number of my cases arrived after prior urinary diversions. Dr Peña is very facile dissecting the anatomy with fine cautery. I would caution others who would emulate this because it risks thermal trauma, which can cause strictures and fistulas. I have not experienced that problem so often as presented today preferring sharp dissection when cautery may seem risky.

Dr Peña has said that a short cloaca can be fixed by most surgeons. My own experience with a high percentage of redo cases referred by other surgeons would lead me to another conclusion. I believe that these are all difficult cases and that they should be sent to a surgeon with special interests and experience in fixing these relatively rare cases seen in only about 1 in 50,000 births.

A question for Dr Peña: What long-term advice do you offer regarding obstetric and gynecologic issues, which

will present when these patients become young women and have babies? Some of mine have reached that age. In fact, 1 nurse has had 5 babies, 1 delivered vaginally and 4 by C-section which included a set of triplets. None of the offspring of these women have had major malformations.”

I also have a question or 2 for Dr Peña. One is, what do you think is the optimal age at which to reconstruct these patients? And the other is, because in my practice I am inheriting a lot of these that have already been reconstructed, what do you think is the optimal follow-up plan for these patients?

A. *Peña (response)*: Thank you Dr Lund. We used to operate on these babies when they were 1 year old. Now, if they are born at our institution, we operate on them when they are 1 month old, but the majority of patients come to us between 6 and 12 months old.

Concerning the follow-up, I think these are patients for life. We try to follow up with them because we are learning so many new things from the long-term follow-up. We are learning mainly that we have been very naive, and these cases are more complicated and have more sequela than what we thought. We are entering into the gynecologic and obstetric concerns of these patients which is going to be a big chapter that has to be written sometime soon. That takes me to the question of Dr Hardy Hendren. I am concerned about these patients because we have some young adults that are having those problems, I called Dr Hendren about 2 years ago and said, “Hardy, I am very worried. Imagine a gynecologist without experience in these malformations going to do a cesarean section in a patient on whom you or me performed a pull-through, bladder augmentation, Malone

and Mitrofanoff. What is going to happen to those patients and what should we do?” And he said, “I agree with you Alberto. We must be there.” I said, “Hardy, when you say we must be there, how long are we going to be around?” He said, “I don’t know about you, but my insurance company tells me that I will be around until I am 100 years old.” (laughter)

A. *Coran (Ann Arbor, MI)*: Thank you for presenting this marvelous series. I think you epitomize what Pat Donahoe said this morning, taking a single problem and spending your life focusing on it brings great results. I have one question about the short channel ones, and that is how much of a female hypospadias would you tolerate in a short channel one and therefore not bother with doing the mobilization part of the procedure? It seems to me that a number of children can have the cloaca repaired when it is a channel of 3 cm or less with doing a perineal procedure on the urogenital sinus, doing the pull-through of the rectum but leaving the urethra hypospadiac, and it does not seem to make a lot of difference.

A. *Peña (response)*: Thank you for your comment. We have 19 patients that I did not have time to mention that were born with a cloaca of about 1 cm common channel, and those patients did not have a total urogenital mobilization. We only did something called introitoplasty and left the patient with a little female hypospadias because we know that a 100% of them have urinary control. On the other hand, if the patient has a very bad sacrum or some urologic condition that makes you believe that the patient will need intermittent catheterization, we are obligated to leave the urethral meatus in a perfectly visible position, but you are right—those patients don’t need all the procedure.