Pediatr Surg Int (2005) 21: 264–269
DOI 10.1007/s00383-005-1380-2

ORIGINAL ARTICLE

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Pitfalls in the management of newborn cloacas

Accepted: 28 December 2004 / Published online: 22 February 2005

Abstract Clinicians caring for newborns with persistent cloaca face significant challenges in the newborn period. Avoiding key pitfalls during this time can have dramatic implications. We reviewed the medical records of 361 patients with cloaca operated on at our institution and analyzed sequelae that resulted from incorrect management in the newborn period. Of 361 patients, 282 underwent primary operations at our institution, and 79 patients were referred to us after a failed repair at other institutions. Pitfalls in management during the newborn period included the following: (1) Failure to recognize and manage hydrocolpos, which occurred in 46 patients. Of these, three patients developed pyocolpos (two progressed to vaginal perforation), and 43 suffered from persistent bilateral hydronephrosis, megaureters, recurrent urinary tract infections, persistent acidosis, or failure to thrive due to undrained hydrocolpos. They underwent unnecessary urinary drainage procedures (nephrostomy, ureterostomy, cystostomy, or vesicostomy) in the newborn period. When the vagina was finally decompressed, all of these symptoms disappeared. (2) Colostomy or vesicostomy problems, which occurred in 50 patients. These included incorrect placement of the colostomy (too distal, which interfered with the pull-through) in 24 and colostomy prolapse in 23. Incompletely diverting loop colostomies led to urinary tract infections in 49 patients. Vesicostomy prolapse occurred in three patients. (3) Clinical misdiagnosis, which occurred in 42 patients. Six were incorrectly diagnosed as ''intersex'' and 36 as ''rectovaginal fistula.'' In this group only the rectum was repaired, and the patients were left with a urogenital sinus that required reoperation. Proper management of a newborn with cloaca includes drainage of a hydrocolpos, which avoids unnecessary urinary diversions and pyocolpos. Our preferred colostomy is one with separated stomas, adequate distal bowel for the pull-through, and use of a proper technique to avoid prolapse. Correct clinical diagnosis of cloaca avoids problems during the definitive repair.

Keywords Cloaca · Anorectal malformation · Hydrocolpos · Review

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Introduction

Clinicians caring for patients with persistent cloaca face significant challenges in the newborn period [1–5]. These infants need urgent clinical and radiologic evaluation by an experienced pediatric surgeon and radiologist. An analysis of a large series of patients with cloaca allowed us to identify sequelae that resulted from incorrect newborn management. These included failure to recognize and manage hydrocolpos, pitfalls in colostomy and vesicostomy creation, and clinical misdiagnoses. We propose that in order to achieve the best clinical results, newborn management must include detection and early treatment of hydrocolpos, a correctly performed colostomy, and an accurate evaluation of the perineum.

Material and methods

We reviewed the medical records of 361 patients with cloaca operated on at our institution and analyzed sequelae that resulted from incorrect management in the newborn period. The manuscript was prepared following the rules and guidelines of the hospital's institutional review board.

Results

Of 361 patients in the series, 282 underwent primary operations at our institution, and 79 were referred to us after a failed repair at other institutions. Because our institution is a referral center for pediatric colorectal problems, the vast majority of patients with cloaca whom we saw were managed as newborns at other institutions. They were subsequently referred for evaluation and definitive surgical treatment.

We identified three categories of pitfalls in the management of cloacas during the newborn period: (1) Failure to recognize and manage hydrocolpos, (2) colostomy and vesicostomy problems, and (3) clinical misdiagnoses.

Hydrocolpos

Seventy-seven patients (25%) had hydrocolpos (Figs. 1, 2). In 31 of the 77 cases (40%), the condition was successfully detected and treated during the neonatal period. In 46 patients, the hydrocolpos was either not identified or was mismanaged. In three of the 46 cases, the patients suffered from serious vaginal infection (pyocolpos). In two of these patients, the vagina perforated and produced an acute abdomen with severe peritonitis.

In the remaining 43 patients, the hydrocolpos led to ureteral obstruction, producing persistent bilateral hydronephrosis and megaureters. These patients frequently underwent unnecessary urinary drainage procedures such as nephrostomy, ureterostomy, cystostomy, or vesicostomy. In addition, if the hydrocolpos remained undrained, the patients suffered from recurrent urinary tract infections, persistent acidosis, and failure to thrive. All of these symptoms disappeared when the vagina was finally decompressed with a vaginostomy.

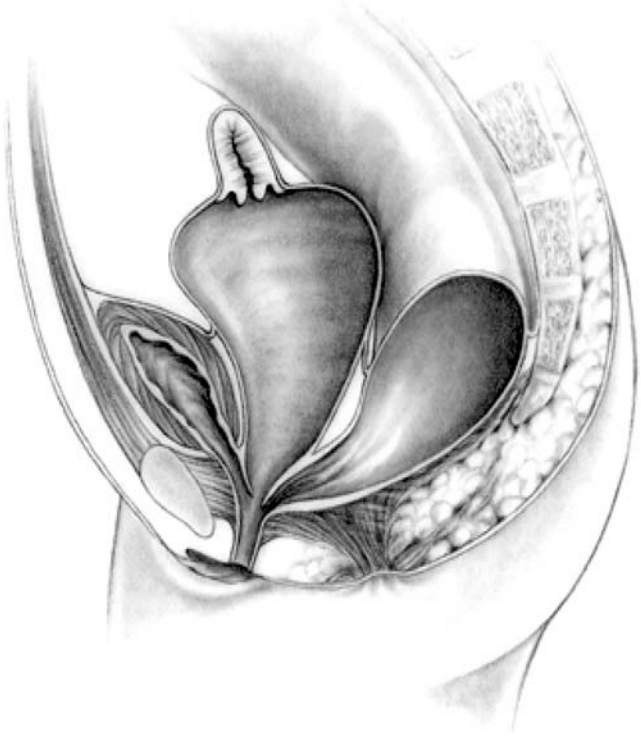

Fig. 1 Artist's drawing of a cloaca with hydrocolpos

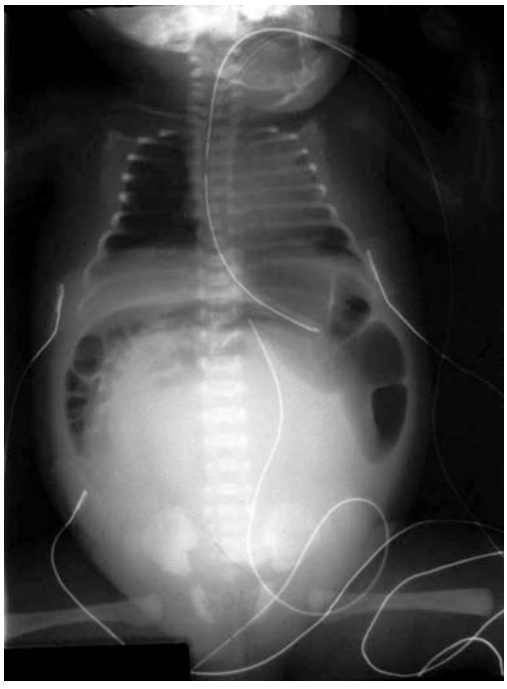

Fig. 2 Plain radiograph of a patient with hydrocolpos manifesting as a large abdominal mass

Colostomy and vesicostomy

Colostomy problems occurred in 47 patients. These included incorrect placement of the colostomy (too distal, which interfered with the pull-through) in 24 patients (Fig. 3a, b), and 23 suffered from colostomy prolapse, particularly when the colostomy was created at a mobile portion of the colon (40%) (Fig. 4) or when a loop-type of colostomy was used (60%). All of these patients required a colostomy revision prior to the main repair.

There were 49 loop colostomies in the series, which led to urinary tract infections from incomplete diversion of the fecal stream. Three patients suffered from vesicostomy prolapse, which required revision prior to the main repair.

Clinical misdiagnoses

Six patients were misdiagnosed as having "intersex" and underwent expensive and unnecessary endocrinologic

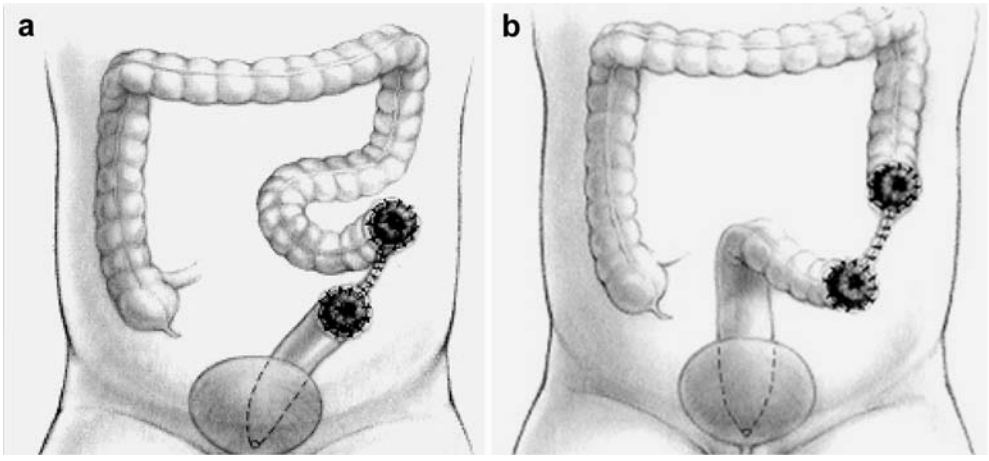

Fig. 3 **a** Colostomy placed too distal, which would interfere with the pull-through. **b** Ideal colostomy with separated stomas

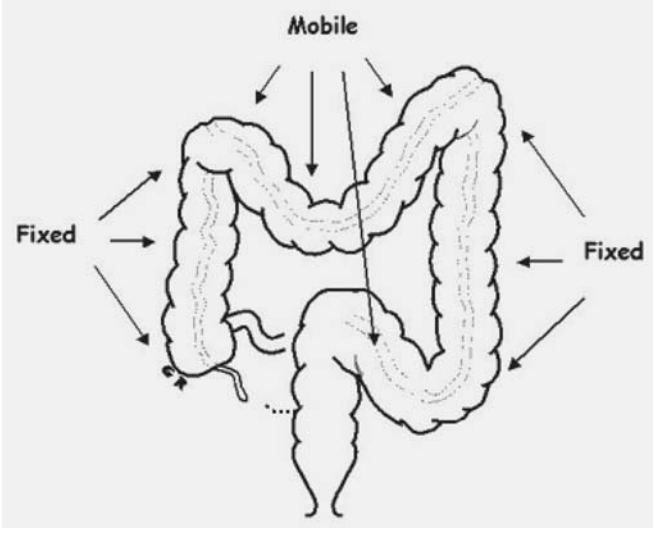

Fig. 4 Areas of mobile colon prone to prolapse

evaluation. The hypertrophied clitoris that is sometime visible in cloacas was perhaps confounding to the clinicians (Fig. 5). All 361 patients in the series had normal ovaries.

Thirty-six patients required reoperation because they had a persistent urogenital sinus after the original operation done at another institution. All of these patients were originally misdiagnosed as having ''rectovaginal fistula'' rather than cloaca, and the urogenital component was not recognized. Essentially, only their rectum was mobilized, and they were left with a urogenital sinus (Fig. 6). Reoperation was performed using a posterior sagittal approach with rectal mobilization and correction of the urogenital sinus.

Discussion

The vast majority of cloacas referred to our center were managed as newborns at other institutions. We were therefore able to analyze a variety of ways in which newborn cloacas were cared for and could draw conclusions from these data. We identified three categories of pitfalls in the management of cloacas during the newborn period: (1) Failure to recognize and manage hydrocolpos, (2) colostomy and vesicostomy problems, and (3) clinical misdiagnoses (Table 1).

During the neonatal period, a baby with cloaca should not be taken to the operating room until the urinary tract has been adequately evaluated and the presence or absence of hydrocolpos established [2–5]. An abdominal ultrasound must include both the upper abdomen, looking for hydronephrosis, and the lower abdomen, looking for hydrocolpos. Occasionally the abdominal radiograph suggests the presence of hydrocolpos (Fig. 2). If the baby has a hydrocolpos, it is mandatory for the surgeon not only to open a colostomy

Fig. 5 Hypertrophied clitoris in a patient with cloaca

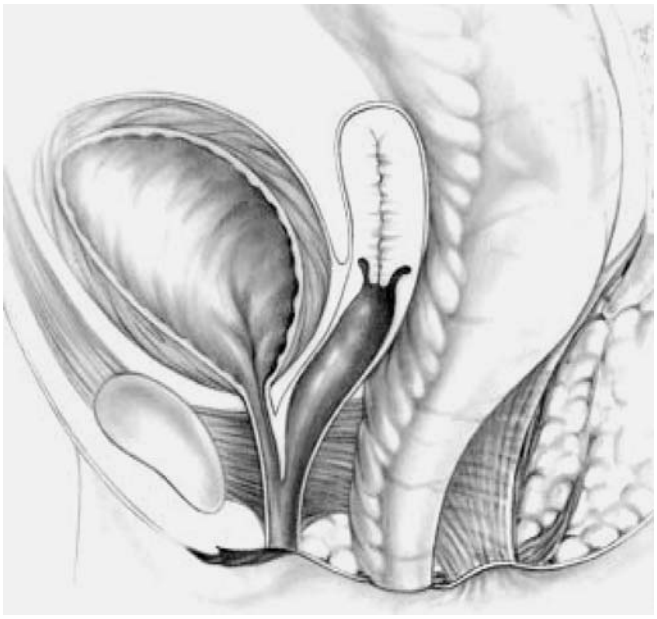

Fig. 6 Artist's drawing of a urogenital sinus left after pull-through of the rectum when the cloaca was misdiagnosed

Table 1 Pitfalls in the management of newborn cloacas

Pitfall	Patients
Failure to manage hydrocolpos	46
Urinary obstruction	43
Pyocolpos	3
Colostomy problems	
Too distal	24
Prolapse	23
Incompletely diverting loop	49
Vesicostomy problems	
Prolapse	3
Clinical misdiagnosis	42
Intersex	6
"Rectovaginal" fistula	36

but also to drain the dilated vagina, or vaginas, and thus prevent complications [3–5].

If hydronephrosis exists, the presence of hydrocolpos must be ruled out before making a decision about creating a ureterostomy or a nephrostomy, as the hydrocolpos may be interfering with the drainage of the ureters [3, 4]. The dilated vagina compresses the trigone of the bladder and thus interferes with ureteral drainage. Vaginal drainage may be all that the patient needs to decompress the urinary tract [4].

Rarely, some patients suffer from an incapacity to empty the bladder due to an almost atretic urethra, and they require some sort of bladder drainage [4]. This rare situation becomes clear if, once the hydrocolpos is decompressed, a dilated pelvic structure remains, which may be the bladder. In such a case, bladder drainage is appropriate [6]. A vesicostomy should be used when the drainage is expected to last over 3 months, and a cystostomy tube should be used if bladder drainage is needed for less than 3 months. The vesicostomy should be performed in the dome of the bladder, sutured to the midline of the abdominal wall below the umbilicus. This puts some stretch on the bladder and prevents vesicostomy prolapse.

Several techniques to drain a hydrocolpos can lead to difficulties. Catheterization of the common channel is unreliable because it may inadequately drain the vagina(s) if the catheter passes into the bladder [7]. Some surgeons employ a plasty of the single perineal orifice, which may provide inadequate drainage particularly in cases of a long common channel. Dilatation of the common channel likewise may inadequately drain the dilated structures.

We recommend a tube vaginostomy in most cases. If the vagina is large enough to reach the umbilicus, it can be sutured to the abdominal skin much like a colostomy. It is vital to perform this drainage procedure during the colostomy opening. The surgeon must also investigate whether there are two dilated hemivaginas, and drain both of them, either with separate tubes or with one tube and creation of a window in the septum between the two vaginas.

If an infant with a cloaca is not growing well in the first weeks or months of life after the colostomy, it is most likely because a complicating problem has been missed. Most commonly, the hydrocolpos has not been drained, which can lead to urinary tract infections and acidosis (Fig. 7a, b). Failure to identify associated urologic problems such as vesicoureteral reflux may also be the cause [8–12]. Prenatal diagnosis of hydrocolpos has been described [13, 14], and fetal intervention has been employed [7]. Therefore, it is likely that decisions about cloaca and management of hydrocolpos during pregnancy will become more common.

An undrained vagina can become infected (pyocolpos), as happened in three of our patients. Two of these patients' vaginas perforated, leading to peritonitis. The most serious sequela of a pyocolpos is the development of severe fibrosis of the vagina. Despite the fact that these patients were born with a large vagina that would have been the ideal structure to use for vaginoplasty, they subsequently required a vaginal replacement because the fibrotic vaginal remnant was not suitable for mobilization.

In addition to managing a hydrocolpos, the surgeon must be thinking about the patient's gynecologic structures in order to prevent future problems [15, 16]. During the colostomy creation, during the definitive repair if a laparotomy is required, or during the colostomy closure, identifying and inspecting the gynecologic structures is vital. Patients may have many variations of Mullerian structure atresias or stenoses that can lead to future amenorrhea or obstruction of menstrual flow and that may affect their ability to conceive and carry a baby to term [10, 15, 16]. Failure to understand the gynecologic anatomy is another potential error in managing cloaca patients.

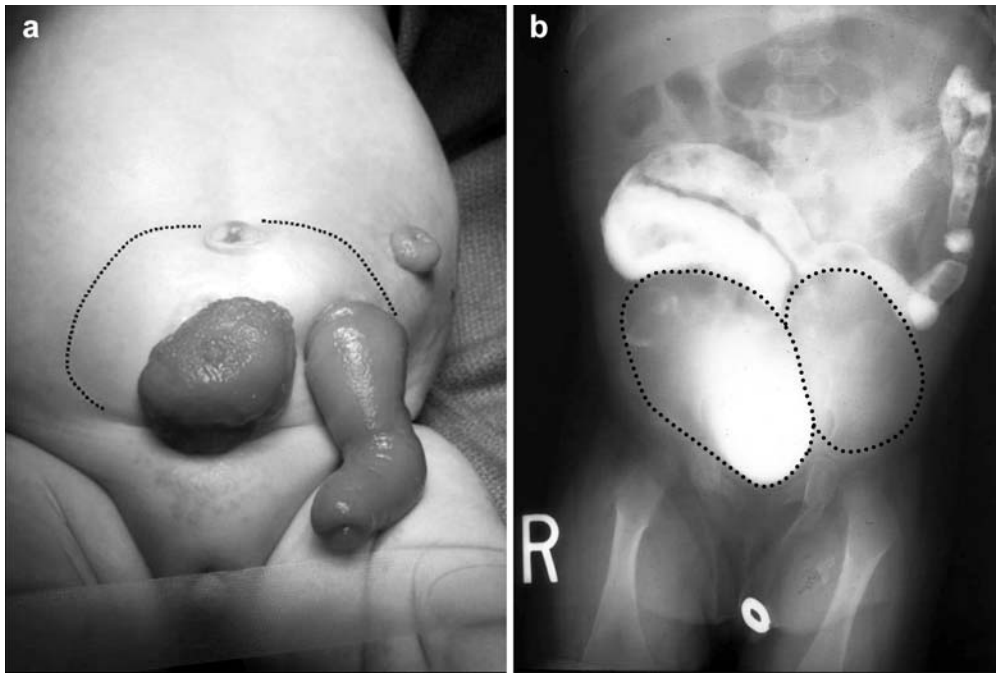

Fig. 7 **a** Vesicostomy prolapse, colostomy prolapse, and hydrocolpos. **b** Contrast study showing two undrained hemivaginas appearing as mass effects in the right and left lower quadrants

Our preferred colostomy is one with separated stomas [5, 17]. The stomas should be separated enough to allow for the placement of a colostomy bag on the proximal stoma (Fig. 3b). In addition, there needs to be adequate distal bowel for the future pull-through. A colostomy without adequate length might lead to an unnecessary laparotomy to gain length for the pull-through. During the main repair, the technique to gain adequate length on the distal piece of bowel is challenging even with a correctly placed colostomy and requires meticulous dissection of the mesenteric vessels. The main trunk is preserved, and the most peripheral branches are ligated so that the distal rectum survives on its intramural blood supply [4, 18] (Fig. 8).

If the colostomy is performed in a mobile portion of the colon (Fig. 4), it is important to fix the colon to the abdominal wall to prevent prolapse, which can be a serious complication in an infant. We recommend placing the colostomy in the descending colon just after the colon takes off from its left retroperitoneal attachments [17]. Another important technical maneuver is to make the mucous fistula tiny, which also prevents prolapse. Prolapse leading to resection of bowel is potentially disastrous in a patient with anorectal malformations because it might convert a patient with a tendency to form solid stool to one who has only loose stool, a condition much harder to manage if the patient is fecally incontinent.

Misdiagnosis of a ''rectovaginal'' fistula may lead to a repair whereby the surgeon only mobilizes the rectum and thus leaves a urogenital sinus untouched [19]. This situation requires a complete redo operation with

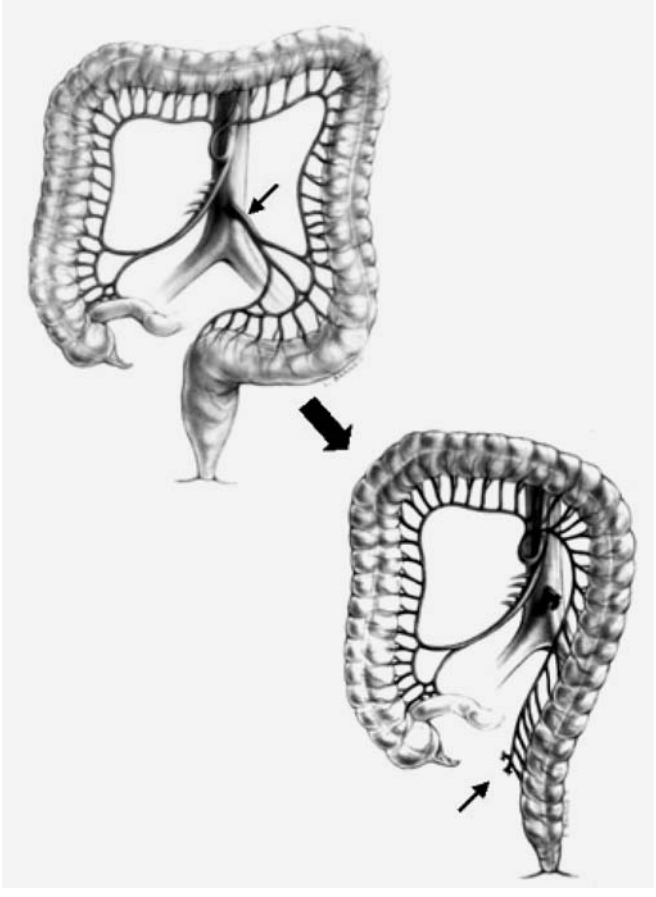

Fig. 8 Technique to gain length on the distal piece of bowel by ligating peripheral branches

mobilization of the rectum out of the way, and then mobilization of the urogenital sinus [19]. The misdiagnosis comes from an inadequate inspection of the perineum and lack of suspicion. In a newborn with no anus, a single perineal orifice defines a cloacal malformation and is a clinical diagnosis. Use of the term ''rectovaginal fistula,'' which is common in the literature [20–29], is confusing, and it is likely that many patients so labeled actually had a rectovestibular fistula or a persistent cloaca. We have found imperforate anus with rectovaginal fistula to be an almost nonexistent condition [4, 19].

Patients with cloaca often have a hypertrophied clitoris, and thus a clinician might suspect an intersex anomaly (Fig. 5). However, all patient in our series of cloacas had two normal ovaries and were chromosomally XX.

Newborn management of cloaca, including drainage of a hydrocolpos, a correctly placed and performed colostomy and vesicostomy, and a precise clinical diagnosis, are vital in preventing serious sequelae.

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