Gynecologic Concerns in the Treatment of Teenagers With Cloaca

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Background/Purpose: Gynecologic anomalies are common in patients with persistent cloaca, but, except for hydrocolpos, these patients remain asymptomatic in the neonatal period. Anatomic abnormalities may become manifest in puberty when menses occurs. The authors sought to describe the sequelae of these anatomic defects in the teenage cloaca patient and to determine recommendations for prevention and treatment in the neonatal period.

Methods: From a series of 198 patients operated on for persistent cloaca, the authors report a group of 22 patients who have reached puberty.

Results: Seven patients are menstruating normally. Six patients have primary amenorrhea because of absent or atretic uteri. Nine patients presented with abdominal pain and cystic abdominal masses and required surgical resection of inflamed collections of old blood in uteri, hemiuteri, tubes, blind vaginas, hemivaginas, or in the peritoneum. The common denominator of this last group was an obstruction of one or more Mullerian structure that interfered with the drainage of menstrual blood. Asymmetric gynecologic anatomy or a vaginal atresia in the neonatal period seemed to correlate with future obstruction to menstrual flow.

Conclusions: To prevent future problems, the management of the neonatal cloaca must include the early clarification of the gynecologic anatomy, specifically the patency of Mullerian structures. Unilateral atretic structures and tubes connected to atretic uteri should be resected. Also, clinicians must be suspicious of menstrual problems in teenagers operated on early in life for persistent cloaca.

INDEX WORDS: Persistent cloaca, menstruation, puberty, hematometra, hematocolpos, hematosalpinx, hemiuterus, vaginal replacement.

A PERSISTENT CLOACA is the conjunction of rectum, vagina, and urethra into a single common channel. It is associated with a wide spectrum of complex anatomic abnormalities involving the urinary, spinal, genital, and gastrointestinal systems. Gynecologic anomalies other than hydrocolpos tend to remain asymptomatic in the neonatal period, but may cause serious problems at the onset of menses. Of these patients, 77% have associated genital tract anomalies,[1] with a septated system in 33% to 50%.[1-3]

The surgical treatment of cloacas has evolved rapidly during the last 15 years.[1-9] With new and sophisticated surgical techniques, most cases can be anatomically repaired. We are still learning about the management of the functional urinary and fecal sequelae. As time goes by, more of these patients are reaching puberty and adolescence, and we have seen a significant proportion of them suffer from serious unpredicted abdominal problems that require surgery. These have included the need for resection of hematocolpos, hematometra, hematosalpinx, and peritoneal pseudocysts. Very little has been written about the gynecologic concerns of these patients when reaching puberty; we found mention of only six such patients in the literature.[4,10] The senior author has operated on 198 cloaca patients from 1982 until 1996. From these, 22 have already reached adolescence and were available to be studied. The purpose of this study is to alert the pediatric surgical community to these problems. From our limited experience, we discuss the best way to suspect, diagnose, prevent, and treat them. As more patients with cloacas survive and undergo repair, we should expect an emerging group of young girls with previously unrecognized problems.

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MATERIALS AND METHODS

The senior author's series of 198 patients operated on for persistent cloaca was reviewed. There were 22 patients 14 years old or older for whom clinical data were available. Seven of these patients were menstruating normally, six patients had primary amenorrhea, and nine had specific gynecologic problems that required surgical intervention.

Clinical features, gynecologic anatomy, presence of hydrocolpos at birth, sacral development, cloacal common channel length, operative repair, and associated gastrointestinal, spinal, and urologic anomalies were investigated for each patient.

RESULTS

Group A, Normal Menses

There were seven patients (32%) with normal menses who were free of abdominal symptoms. All had a symmetric gynecologic system with two hemiuteri and a vaginal septum. Four patients had an absent kidney: three were missing the right kidney, and one the left kidney.

Group B, Amenorrhea Group

Six of the 22 patients (27%) had primary amenorrhea. Three of these patients were determined to have a congenitally absent uterus that was demonstrated at the original operative repair. One patient was found to have atretic remnants of uteri at the time of the original operative repair, and these remnants were resected. Two patients had two atretic hemiuteri found at the original repair; these were not resected, and these patients have never menstruated. At the time of original repair, three patients were found to have small, blind vaginas; one patient had no vagina; and two had normal vaginas. One patient had an absent right kidney, and one had an absent left kidney.

Group C, Surgical Group, Abnormal Menses

Nine patients (41%) had severe abdominal symptoms and cystic abdominal masses requiring surgical intervention. Figure 1 shows their gynecologic anatomy and the surgical treatment that was necessary. All of them came for consultation or were seen in the emergency department complaining of severe abdominal pain. Two underwent emergency surgery. Six patients never menstruated, and three menstruated irregularly. Seven patients had cyclic abdominal pain with monthly exacerbations, one patient had chronic abdominal pain, and one patient was pain free. All nine patients had palpable abdominal masses. Diagnostic study findings (sonography, computed tomography, magnetic resonance imaging) demonstrated very large cystic abdominal masses (Fig 2). At the time of exploration, the cystic masses were found to be large, inflamed collections of old blood in uteri or hemiuteri (hematometra, n = 6), in the tubes (hematosalpinx, n = 8), in a blind vagina or hemivagina (hematocolpos, n = 2), and/or in the peritoneum (hemoperitoneum with pseudocyst, n = 7), which represented retrograde menstruation. The large cystic and inflamed masses involved any or all of the gynecologic structures (Figs 3 and 4).

The common finding in this group was a partial or complete obstruction to menstrual flow. A septated system was found in seven of the nine patients. Six patients had asymmetric systems. Three patients had one atretic side and one patent side and menstruated through the patent side. One patient had a blind left hemivagina with a patent right hemivagina, and one patient had a right atretic hemiuterus (resected as a neonate) with a normal-appearing left side. Four patients had congenitally absent vagina. Four patients had an absent right kidney, one had a horseshoe kidney, and one had bilateral ectopic kidneys.

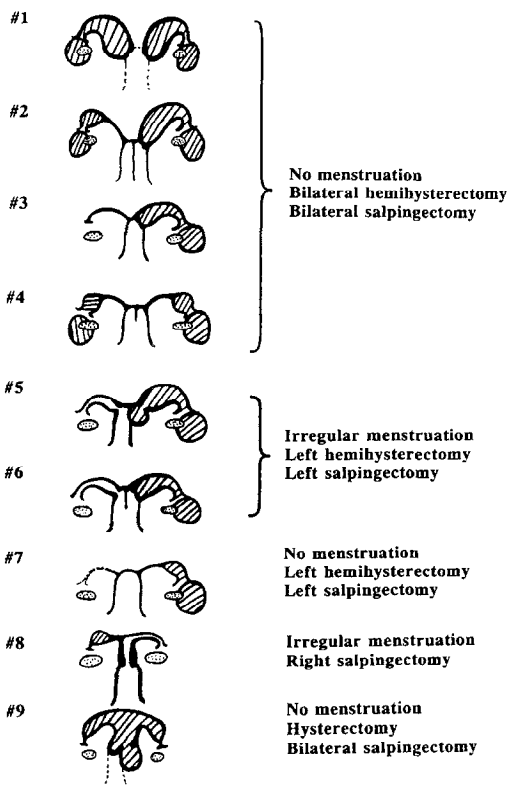

Fig 1. Surgical group, gynecologic pathology, symptoms, and treatment.

We compared the characteristics between each group looking for differences between them that could be predictive of future difficulties. The patients with normal menses all had hemiuteri, and had similarly divided vaginas with vaginal septums. Their systems were divided symmetrically. Of the patients with menstrual problems requiring surgery, six of nine had asymmetric systems, with three having one functionally normal side. Four of the patients in the surgical group had an absent vagina whereas all patients in the normal menses group had vaginas. All 22 patients had normal ovaries. There were no significant differences between the groups in their sacral development, in the cloacal common channel length, in whether hydrocolpos was present at birth, or in their associated anomalies. Renal anomalies were common in all groups, and we could discern no pattern relating the side of the renal anomalies to abnormal gynecologic anatomy.

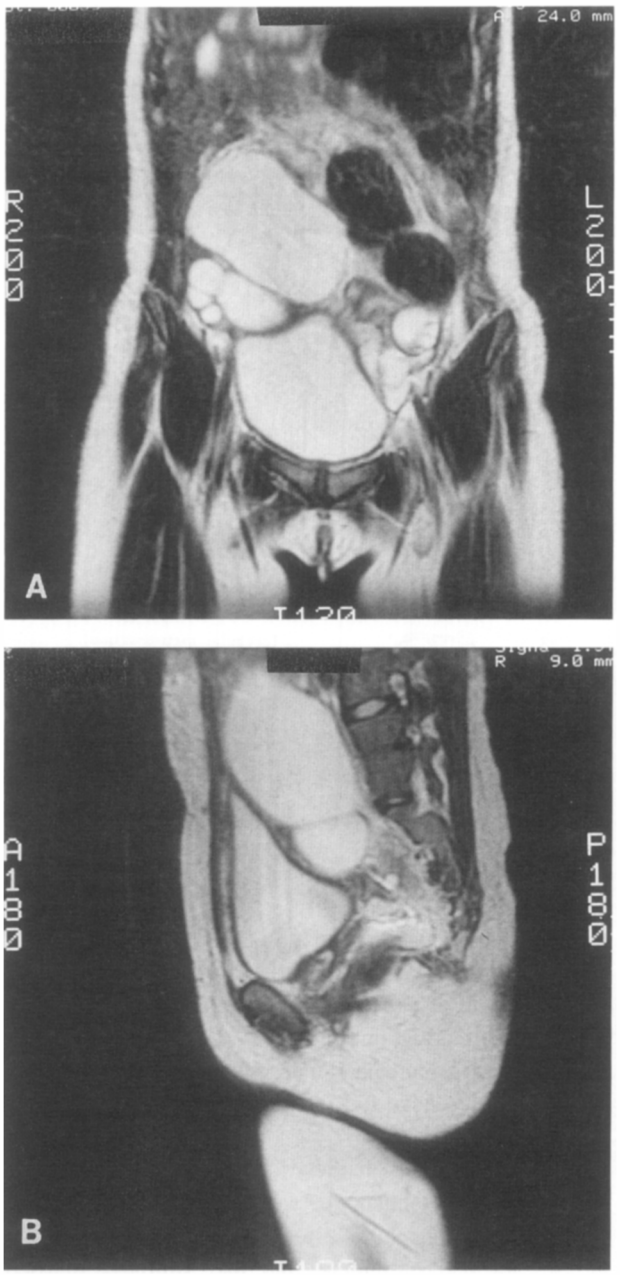

Fig 2. (A,B) Anterior-posterior and sagittal magnetic resonance images showing pelvic collections of menstrual blood.

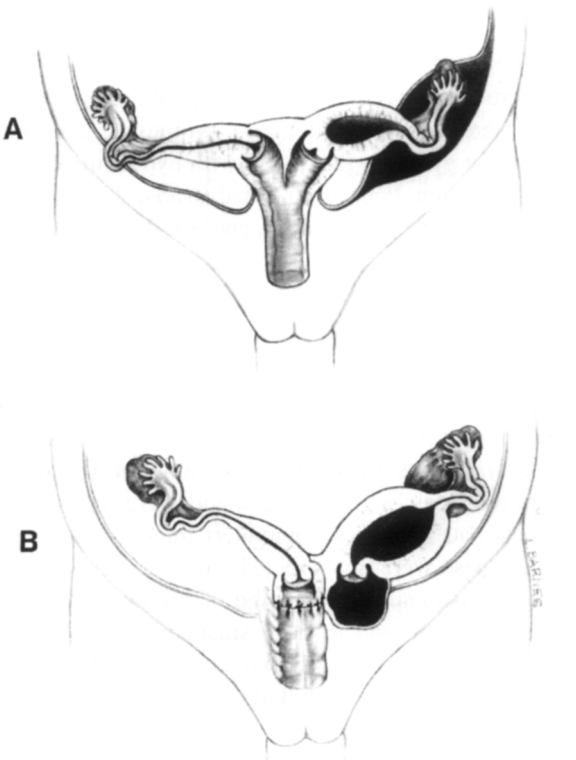

Fig 3. (A) Artistic rendition of a case of atresia of the cervix with ipsilateral hematometra and peritoneal pseudocyst. (B) Drawing of a case of an atretic left hemivagina. There is trapped menstrual blood in the hemiuterus and peritoneum. The right hemivagina is patent and drains through a neovagina made from colon.

DISCUSSION

At the time we performed the main operation for repair of the persistent cloaca, posterior sagittal anorectovaginourethroplasty (PSARVUP),[1] we did not fully appreciate that these patients would, when reaching puberty, suffer from sequelae of their anatomic anomalies. We should have been alerted to these problems in patients born with persistent cloaca, however, because Mollitt et al[10] in 1981 published related findings, presenting five cases of problems very similar to ours. The terminology in those years was different because they referred to their patients as having "urogenital sinus with imperforate anus," now known as persistent cloaca. We attempted in our study, in contrast to this previous report, to present specific recommendations on how to treat the gynecologic sequelae in the teenager and to discuss prevention of future problems when faced with the newborn cloaca.

Primary amenorrhea was predicted in a group of our

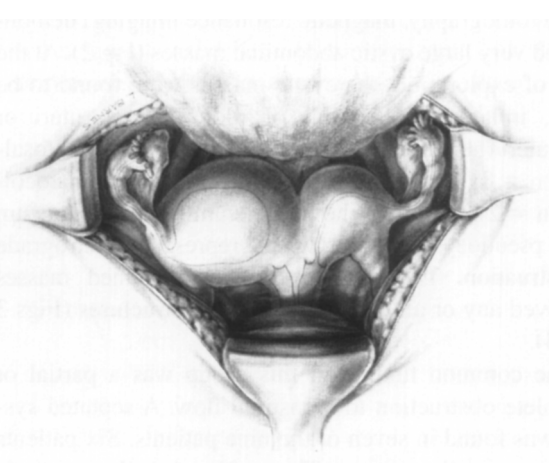

Fig 4. Drawing showing bilateral hematometra caused by bilateral atresia of the cervix.

patients from the finding of an absent uterus at the neonatal repair, or after resection of atretic uteri. In this scenario, it is vital to counsel a patient to expect amenorrhea in the teenage years. We had two patients in whom atretic hemiuteri were not resected. Neither of these patients has menstruated. Unfortunately, their gynecologic anatomy was described in retrospect when they were evaluated for primary amenorrhea, and they had not been forewarned of the probability of future amenorrhea.

In patients without cloaca, agenesis of the uterus and vagina is well described as part of the Mayer-Rokitansky syndrome.[11-18] As in our amenorrhea group, these patients have primary amenorrhea on the basis of a missing uterus.

The group of patients who presented as teenagers with abdominal pain and cystic abdominal masses required, as did those in Mollitt's group,[10] resection of inflamed cystic masses involving their tubes and/or uteri.

Similar problems are described in the gynecologic literature in patients without cloaca but with isolated reproductive tract congenital anomalies.[11,14-26] Mullerian anomalies such as rudimentary cornua, uterus didelphys, and vaginal atresia lead to inadequate outflow of menstrual blood,[11,15,20,27,28] which result in hematocolpos, hematometra, hematosalpinx, and endometriosis[11,14-16,19,20,22,27,28] from menstrual blood filling and dilating these structures.[14,17] The blood can also spill retrograde into the peritoneum forming cystic collections.[18,22] In these patients, as in ours, cystic and/or complex masses representing dilated Mullerian structures were found and resected,[14,15,20,23,26,29,30] and oopherectomy was performed when the inflammatory process involved the ovaries.[29,30] In addition to resection, hematocolpos and hematometra secondary to vaginal atresia has been treated with drainage,[17,26,29] anovulatory medication[22] (which may allow dilated structures to return to normal size), delayed repair involving vaginal pull-through,[22] and resection of vaginal septa.[14,17,31] Treatment of patients without cloaca but with cervical and uterine hypoplasia has been attempted with mixed results,[16,17] with reports of surgical reconstruction[32-34] and stenting.[35] We found case reports of five patients in whom treatment was attempted. Chronic pelvic pain, endometriosis, or infertility developed in four patients. In these case reports, we found only one report of an asymptomatic patient who was able to conceive and carry a pregnancy to term.[34]

After surgical resection, our patients were asymptomatic. The critical question, however, is whether it would have been possible to save their gynecologic structures. Our impression is that any attempt to create an effective uterine drainage would fail. We base this on the fact that all the structures had a severe inflammatory reaction. Also, assuming that we could repair these structures and create a communication, how functional would the hemiuteri be? We do not have an answer for this important question. Identifying anatomic anomalies before serious problems result could lead to their repair in an elective setting.

Additionally, we must ask whether there is a way to predict menstrual problems when a clinician evaluates a newborn with persistent cloaca. Should repair of abnormal structures be attempted, or should these abnormal structures be resected?

From our limited experience, we cannot fully answer these questions. However, we think that all pediatric surgeons dealing with cloacas should be aware that a significant number of these patients have some form of atresia that may some day obstruct the drainage of menstrual blood and provoke serious problems in adolescence. The suspicion should be stronger in cases of asymmetric gynecologic systems because our data suggest that this factor related to future problems with obstruction. Also, atresia of the vagina is an important predictor of future problems when found in association with an intact uterus.

Every effort should be made in infancy to make an assessment of the gynecologic structures. It remains to be seen which diagnostic tool is more effective for this purpose. Endoscopic examination, also mentioned by Mollitt et al,[10] or direct inspection during the repair can show us the presence of a cervix but cannot tell us whether the entire uterine or tubal lumen is patent. Hysterosalpingography, magnetic resonance, or ultrasound scan could be helpful. The surgeon should inspect the gynecologic structures if laparotomy is required for the cloacal repair[1] or at the time of colostomy closure.

While managing a newborn cloaca, if an obstructed tract (vagina, uterus, cervix, tube) is identified, the surgeon can try to repair the obstructed systems, resect the uterus and the tube, or observe the patient until puberty.

We could find no reports in the literature on early repair of these atresias. From our experience, we predict that it would be very difficult and probably unsuccessful. The structures are extremely small and the atresias are frequently multiple.

It is possible for a patient to have an atresia on one side and a normal patent lumen on the other. That, we think, is an indication for resection of the affected side. Some patients have tubes that become thinner as one follows them toward the midline, becoming cordlike structures (remnant of uterus). In such cases, our specific recommendation is to resect both tubes because they frequently have multiple atresias and may have symptomatic cystic dilatations at puberty. In a case in which both sides are affected, one could be more conservative and consideration should be given to repair.

If the surgeon finds a single vagina, a meticulous search for a second vagina should ensue, because nearly 50% of the patients with cloaca have some form of septation or duplication of the gynecologic structures.[1] Furthermore, sometimes (as shown in our cases) one of the vaginas is patent and the other is not. If the surgeon finds the one that is open and does not look for the other, the patient may suffer serious problems in the future.

Observing the patient until puberty is an option. Perhaps hysterosalpingography before onset of symptoms could provide the clinician with anatomic information that would guide treatment.

Renal anomalies have been reported to occur on the same side as an atretic hemivagina, in association with uterus didelphys.[14,20-24,25,29,31,35-38] In our patients with similar gynecologic anomalies, renal anomalies were present but were not ipsilateral. There is likely a relationship between the side of a urologic anomaly and future menstrual problems, but in our group of patients, we could not discern a pattern.

One future question clinicians will face in these patients regards their obstetric potential. Hendren[4] reported on two patients who conceived and delivered infants by cesarean section after cloacal repair, but does not mention their original gynecologic anatomy. In patients born with isolated uterine anomalies without cloacas, conception is possible, but maintenance of pregnancy is difficult.[11] Bilateral hemiuterus (the most common finding in our patients) has been reported to have the best obstetrical outcome of any of the major congenital uterine malformations in patients without cloaca.[11,15,39,40]

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Discussion

A.G. Coran (Ann Arbor, MI): We just presented a review of our cloaca patients in whom very complex ovarian cysts developed, so complex that attempts at partially resecting the ovary did not relieve severe chronic abdominal pain. Have you seen any of these findings in your patients?

A. Pẽna (response): Yes, a couple of those patients in whom we resected those collections of blood also had ovarian cysts. I am aware of your paper, and I was very interested because we have seen something similar. However, your cases were cloacal exstrophies. This is different. This is a collection of blood in the peritoneum and in the tubes and the hemiuterus.

J. Kelly (East Melbourne, Australia): It was great to hear of your work with cloacas; the results are excellent. However, do you know if you resect the hemiuterus that the remaining uterus isn't going to rupture during later pregnancy?

A. Peña (response): We don't know. We just try to preserve as much as we can, assuming that it will work. The literature shows that there are women with a hemiuterus who have delivered babies. There are not many cases, but the gynecologic literature also indicates that those ladies are more prone to have abortions and premature labor. In the next 10 years we will be learning more about this population. This happens to be much more complicated than we thought.

W.H. Hendren (Boston, MA): I agree completely with the message of this paper, which is that some cloaca patients have late gynecologic complications. Four of the 152 cloaca patients whom I have cared for have, after puberty, acquired a blocked uterus and tube that required surgical removal. One of them presented late, with incipient spontaneous drainage by erosion through the abdominal wall in the right lower quadrant.

Six of the patients have become pregnant, one of them twice. All of the babies have been normal. One was delivered per vagina, and all the others by cesarean section. Many of these patients are prepubertal. I agree that they should all be carefully followed up into adult life, and I think we will learn some interesting things about them as they grow older. Of particular interest will be those girls in whom the vagina was too short to bring to the perineum, and so a bowel extension was performed. Some of those are old enough to be menstruating through the bowel extension, but I have not seen one of those who has yet become pregnant.