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Posterior cloaca—further experience and guidelines for the treatment of an unusual anorectal malformation

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Anorectal malformation

Abstract

Introduction: The term *posterior cloaca* refers to a malformation in which the urethra and vagina are fused, forming a urogenital sinus that deviates posteriorly to open in the anterior rectal wall or immediately anterior to the anus.

Methods: A retrospective review of 411 patients diagnosed with cloaca was performed to identify the ones with a posterior cloaca. Special emphasis was placed on anatomy, diagnosis, associated anomalies, and outcome in terms of urinary and fecal continence. Surgical treatment was a total urogenital mobilization with a transrectal approach.

Results: Twenty-nine patients were diagnosed with a posterior cloaca. Of these, 15 had a single orifice at the normal location of the anus with the urogenital sinus opening in the anterior rectal wall. Fourteen had the urogenital sinus opening immediately anterior to the normally located anal opening (2 orifices), which we considered a posterior cloaca variant. Nineteen patients (65%) had hydrocolpos. Twenty-seven patients (93%) had associated urologic anomalies, 12 patients (41%) had gynecologic anomalies, and vertebral malformations occurred in 41% of cases. Other anomalies included gastrointestinal (7 patients), cardiac (5), and tethered cord (2). Late diagnosis occurred in 2 patients. Twenty patients were available for long-term follow-up: 17 are fecally continent, 3 are fecally incontinent, 11 are urinary continent, 5 are dry with intermittent catheterization, and 4 have dribble urine.

Conclusion: The most important characteristic of the posterior cloaca is the high frequency of a normal anus, which differentiates this malformation from the classic cloaca. Often, many associated malformations are present and therefore should be suspected and diagnosed. The main goal during the operation should be to not mobilize the anus and thereby preserve the anal canal. A total urogenital mobilization, transperineally or with a transanorectal approach, is ideal for the repair.

In a typical cloaca, the urethra, vagina, and rectum are fused, forming a common channel that opens as a single orifice in the perineum at the site where the urethra would normally be located [1]. We use the term *posterior cloaca* to refer to a malformation in which the urethra and vagina are fused, forming a urogenital sinus that is posteriorly deviated and opens in or immediately anterior to the rectal wall (Figs. 1-4) [2,3].

The most relevant difference between a typical cloaca and the posterior cloaca is that, in the latter, the anus is normally located and has a pectinate line. This means that the anal canal

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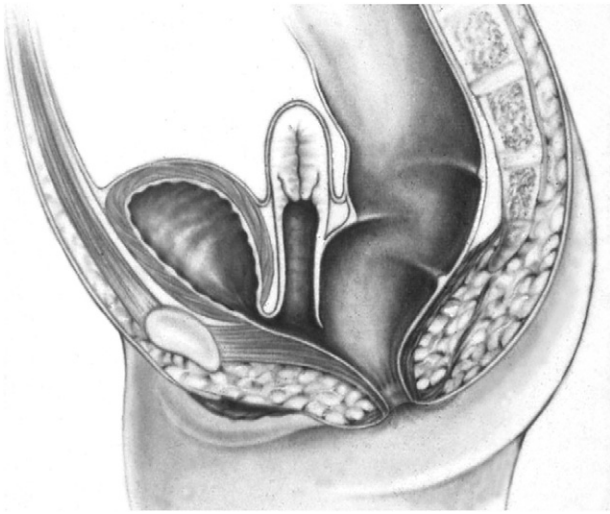

Fig. 1 Artistic diagram of a posterior cloaca.

is present; and therefore, the patient has normal sensation, a normal sphincter mechanism, and potential for full bowel control. It is vital that the surgeon recognizes this characteristic before the definitive repair, and it is for this reason that the anus should not be mobilized during the repair.

In 1998, we published our experience with 9 cases [2] of posterior cloaca; and we now present a series of 29 cases with new lessons that we have learned.

1. Methods

A retrospective review of 411 medical records of patients diagnosed with cloacal malformations was performed to identify the ones with a posterior cloaca. Special emphasis was placed on anatomy, diagnosis, associated anomalies, and outcome in terms of urinary and fecal continence. Surgical treatment consisted of a total urogenital mobilization [4] with a transanorectal approach [5] (splitting the anterior and posterior rectal wall in the midline) (Fig. 5), followed by a meticulous reconstruction of the sphincter mechanism as well as both anterior and posterior rectal walls. Based on this review, we have formulated a series of recommendations for the treatment of this condition.

We obtained institutional review board approval (2008-1317) for this study.

2. Results

Twenty-nine patients (7%) were diagnosed with a posterior cloaca. Of those, 15 had a single orifice at the normal location of the anus with the urogenital sinus opening in the anterior rectal wall (Figs. 1 and 2); and 14 had the urogenital sinus opening immediately anterior to a normally located anal opening (2 orifices) (Figs. 3 and 4), which we considered a posterior cloaca variant. Nineteen patients (65%) had hydrocolpos. Twenty-seven patients (93%) had associated urologic anomalies, including an accessory stenotic urethra opening at the tip of the clitoris (12 patients) (Fig. 6), a single/nonfunctional kidney (7), bilateral vesicoureteral reflux (7), bilateral hydronephrosis (7), unilateral vesicoureteral reflux (3), unilateral hydronephrosis (2), bladder diverticulum (2), urachal cyst (1), crossed renal ectopia (1), bilateral dyplastic kidneys (1), right ureterocele, and kidney duplication (1).

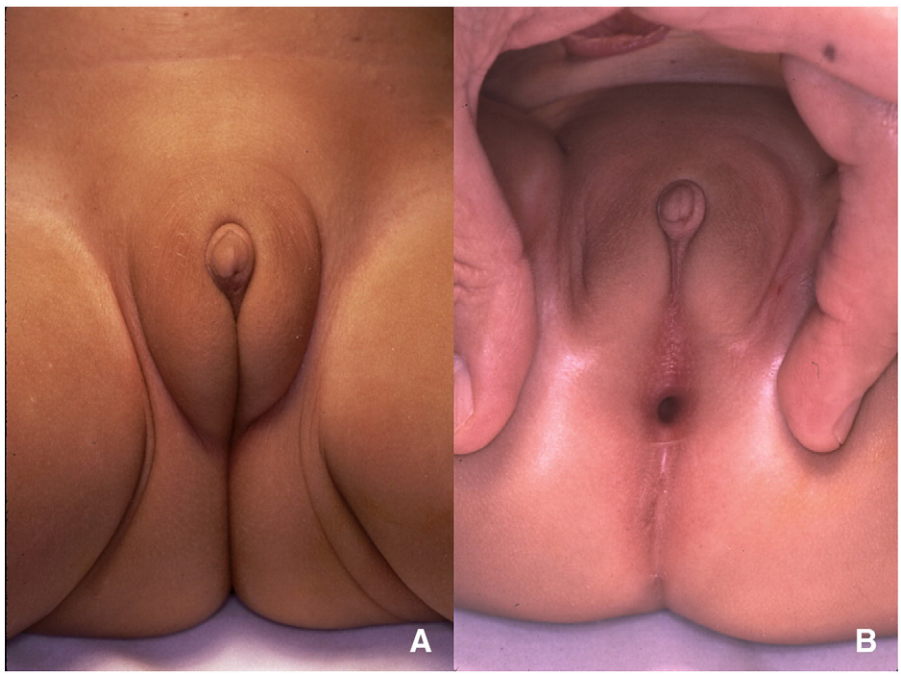

Fig. 2 Perineum of a posterior cloaca without spreading the labia (A) and after spreading the labia (B) showing the single orifice.

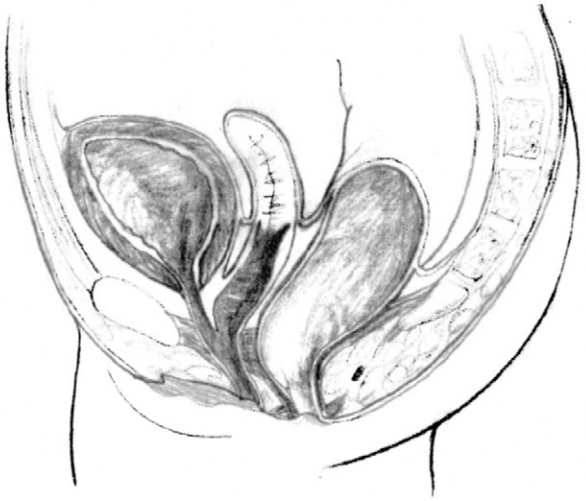

Fig. 3 Artistic diagram of a posterior cloaca variant with urogenital sinus posteriorly deviated, opening immediately anterior to the anus.

Gynecologic anomalies were present in 12 patients (41%) including 2 hemivaginas and hemiuteri (10 patients) and a bicornuate uterus (2 patients). Vertebral malformations occurred in 41% of cases: 10 had hemivertebrae, 1 had sacral agenesis, and 1 had fused vertebrae. The sacral ratio was available in 21 patients; of those, 12 had a sacral ratio greater than 0.7; 7 patients had a sacral ratio between 0.4 and 0.7; and 2 patients had a sacral ratio of less than 0.4. Two patients had a tethered cord. Gastrointestinal anomalies were found in 21% of the cases and included: esophageal atresia with tracheoesophageal fistula (4), malrotation (2), and duodenal atresia (1). Five patients had cardiac malformations: ventricular septal defect (4), patent ductus arteriosus (2), and coarctation of the aorta (1). Late diagnosis of the posterior cloaca occurred in 2 patients, at 2 years and 8 years of age.

Twenty patients were available for long term follow-up: 17 are fecally continent, and 3 are fecally incontinent. Of the 3 fecally incontinent patients (patients 11, 15, and 21; Table 1), one had sacral agenesis and one had tethered cord and a very hypodeveloped sacrum (sacral ratio = 0.2). For the last one, we had no information about the sacrum. Eleven are continent of urine, 5 are dry with intermittent catheterization, and 4 have dribble urine. Three patients are menstruating and are sexually active.

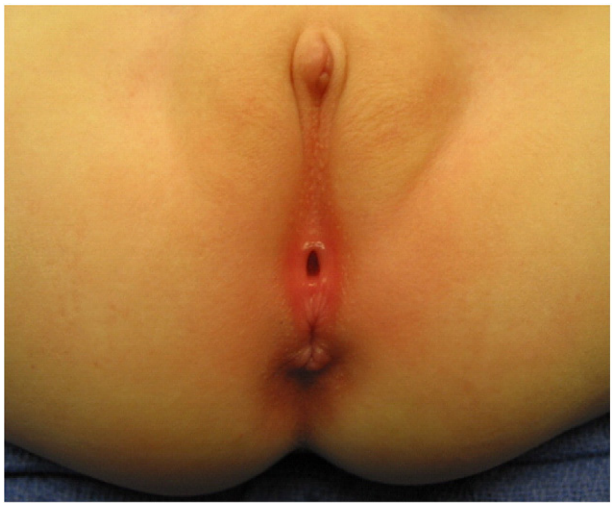

Fig. 4 Perineum of a posterior cloaca variant with 2 orifices—posteriorly deviated urogenital sinus and normal anus.

3. Discussion

Strictly speaking, the term *cloaca* should only be used when a single orifice is found in the perineum. However, we found impressive similarities between the group of patients with 1 orifice (anus only) and 2 orifices (anus and urogenital sinus). The most important features of both groups are the unique characteristic of the posterior deviation of the urogenital sinus and the normal anus. We therefore considered both defects as variants of the same type of anomaly.

We are aware of the existence of a urogenital sinus with normal rectum and anus, with or without adrenal hyperplasia (virilization). In all those cases, the urogenital sinus opens much more anteriorly, at the location of a normal urethra. In the "posterior cloaca spectrum," the urogenital sinus is located much more posteriorly; and one can see only skin between the clitoris and the urogenital orifice (Figs. 3 and 4).

Our main concern in describing this unique malformation goes beyond semantics. From the practical and therapeutic points of view, surgeons must correctly identify this defect to treat it adequately. Specifically, we want to emphasize that a circumferential dissection of the rectum, like in all other types of anorectal malformations, is contraindicated. In posterior cloacas, the anus is normally or almost normally located, has a pectinate line and an anal canal, and is surrounded by a normal sphincter. The innervation of the anal canal comes laterally, from both sides; and a perirectal dissection would denervate it, as has been experimentally demonstrated [6]. It is for this reason that we advocate the transrectal approach in this specific group of patients.

We believe that this malformation is more common than previously thought. In our literature review, we found authors referring to this defect as *posterior cloaca* [2,3,7-9]; but we also found terms such as *partial urorectal septal defect* and others [10-14] in which the anatomical features, seen in photographs and diagrams, were consistent with the diagnosis of a posterior cloaca.

The association between this malformation and hydrocolpos (65% in our series) was a surprise to us, compared with typical cloacas and hydrocolpos (25%) [15]. Hydrocolpos is still a condition underdiagnosed in most cloaca patients [15]. Our findings reaffirm the importance of suspecting, diagnosing, and adequately draining the hydrocolpos [15], particularly in cases of posterior cloacas. Perhaps the anatomical configuration of this malformation makes drainage of the fluid from the vagina more difficult, leading to a hydrocolpos.

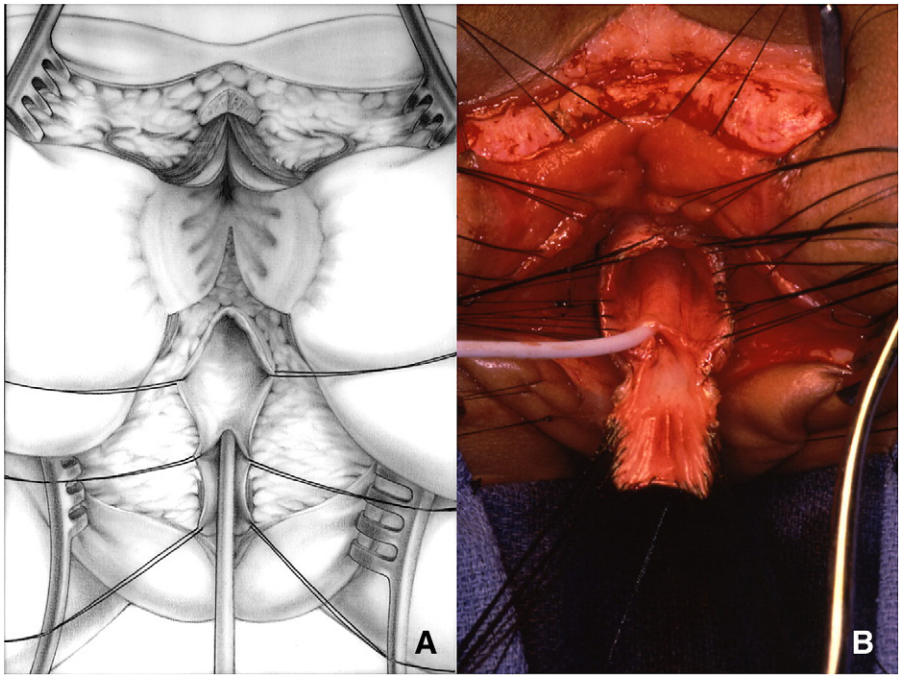

Fig. 5 A, Artistic drawing of a transanorectal approach ready for a total urogenital mobilization. B, Photograph of a transanorectal approach with the urogenital sinus already mobilized.

The association between urologic anomalies and anorectal malformations is well known; and in cloacas, it varies from 59% (if the common channel is <3 cm) to 91% (common channel >3 cm) [16]. In this series, the associated incidence for posterior cloacas was 93%. The most frequent urologic malformation found in posterior cloacas was an accessory urethra that has also been described by other authors [8,9,13]. As already mentioned, it is a very stenotic urethra; and its reconstruction should not be attempted because the patient has a functioning urethra that is posteriorly deviated. It is very easy to just resect the accessory urethra.

Looking at this malformation (Fig. 6) makes us wonder if the posterior cloaca is the female version of what we call the *absent penis spectrum* [17]. Interestingly, in both groups of patients (males and females), there seems to be a very prominent overgrowth of the pubic bone. Whether this is cause, consequence, or coincidence remains unknown. Also in both groups, there is a posteriorly deviated urogenital sinus and a spectrum of hypodeveloped orthotopic urethra that ranges from a complete absence (with absent penis in the male) to different degrees of urethral stenosis, which are generally very difficult to dilate.

The presence of 2 hemivaginas and hemiuteri (2 cervices) that occurred in 10 of our patients should be diagnosed and followed because of possible future menstrual problems (obstruction) and obstetric implications [18,19].

The late diagnosis of the malformation that occurred in 2 patients reinforced our idea that this is the type of anomaly that must be suspected to be diagnosed. Because patients have a normal anus, the clinician can only diagnose it by spreading the labia and observing the absence of a urethral and vaginal opening (Fig. 2A, B).

We believe that the treatment of choice for a posterior cloaca, so that the potential for bowel function is preserved, is preservation of the anus and a total urogenital mobilization [4], which can be done through the perineum in cases with a short common channel or with 2 separated orifices (Figs. 3 and 4). However, in cases with longer common channels or when the urogenital sinus opens in the anterior rectal wall (Figs. 1 and 2), the transanorectal approach [5] seems to be ideal. The transanorectal approach has been experimentally and clinically proven to not compromise bowel control [20] because the anal canal is preserved.

In the long-term follow-up of the 20 patients that were available for evaluation (Table 2), 17 (85%) had voluntary

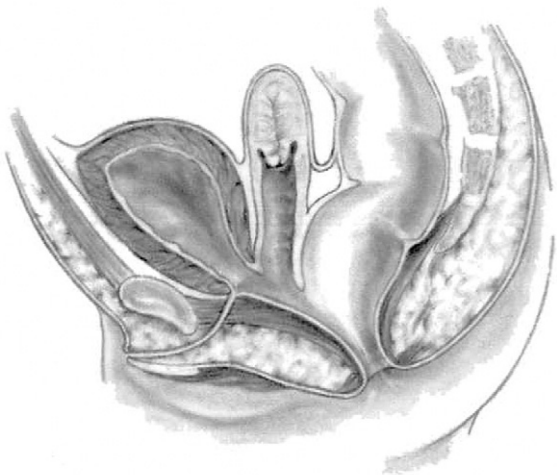

Fig. 6 Artistic diagram of a posterior cloaca with an accessory urethra.

Table 1 Patients with posterior cloaca and their associated anomalies

Patient no.	Urologic anomalies (93%)	Gynecologic anomalies (41%)	Vertebral anomalies (41%)	Gastrointestinal anomalies (21%)	Cardiac anomalies (17%)	Neurologic (6.8%)
1	Single/nonfunctional kidney	None	None	None	None	None
2	None	None	Hemivertebrae	None	None	None
3	Accessory urethra	None	None	Esophageal atresia with TEF	None	None
4	None	2 Hemivaginas and hemiuterus	None	None	None	None
5	Bilateral hydronephrosis, urachal cyst	None	Hemivertebrae	Malrotation	None	None
6	Bilateral hydronephrosis, right reflux	2 Hemivaginas and hemiuterus	Hemivertebrae	None	None	None
7	Bilateral hydronephrosis, bilateral reflux	Bicornuate uterus	None	None	None	None
8	Single/nonfunctional kidney, bladder diverticulum	None	None	None	None	None
9	Accessory urethra	None	None	None	None	None
10	Accessory urethra, bilateral reflux	None	Hemivertebrae	None	None	None
11	Dysgenetic kidneys	2 Hemivaginas and hemiuterus	Absent sacrum	Esophageal atresia with TEF, malrotation	VSD	None
12	Accessory urethra, bilateral reflux	None	None	None	None	None
13	Bilateral hydronephrosis	2 Hemivaginas and hemiuterus	None	None	None	None
14	Single/nonfunctional kidney, right hydronephrosis	2 Hemivaginas and hemiuterus	Hemivertebrae	None	None	None
15	Bilateral reflux	None	None	None	None	Tethered cord
16	Accessory urethra	None	Hemivertebrae	None	None	None
17	Single/nonfunctional kidney, right reflux	2 Hemivaginas and hemiuterus	None	None	None	None
18	Accessory urethra, right reflux, left crossed renal ectopia	None	Hemivertebrae	None	None	None
19	Accessory urethra	None	Hemivertebrae	None	None	None
20	Accessory urethra, left hydronephrosis	None	None	None	None	None
21	Accessory urethra, single/nonfunctional kidney, bilateral hydronephrosis, bilateral reflux	Bicornuate uterus	None	None	None	None
22	Bilateral reflux, bladder diverticulum	None	Fused vertebrae	Duodenal atresia	VSD	None
23	Accessory urethra, single/nonfunctional kidney	2 Hemivaginas and hemiuterus	Hemivertebrae	None	VSD	None
24	Bilateral hydronephrosis	None	None	None	VSD, PDA, aorta coarctation	None
25	Single/nonfunctional kidney, bilateral reflux	None	None	None	None	None
26	Single/nonfunctional kidney	2 Hemivaginas and hemiuterus	None	None	None	None
27	Accessory urethra, bilateral hydronephrosis	2 Hemivaginas and hemiuterus	None	None	None	None
28	Right ureterocele and kidney duplication	None	Hemivertebrae	Esophageal atresia with TEF	PDA	None
29	Accessory urethra	2 Hemivaginas and hemiuterus	None	Esophageal atresia with TEF	None	Tethered cord

Table 2 Long-term follow-up available for 20 patients

Patient no.	Sacral ratio *	Urinary continence status	Bowel continence status
1	>0.7	Dribbling/leaking	Voluntary bowel movement
2	>0.7	Continent	Voluntary bowel movement
3	>0.7	Continent	Voluntary bowel movement
4	>0.7	Continent	Voluntary bowel movement
5	NA	Dribbling/leaking	Voluntary bowel movement
6	>0.7	Intermittent catheterization	Voluntary bowel movement
7	>0.7	Continent	Voluntary bowel movement
9	NA	Continent	Voluntary bowel movement
10	0.4-0.7	Continent	Voluntary bowel movement
11	<0.4	Dribbling/leaking	Permanent colostomy
12	>0.7	Continent	Voluntary bowel movement
13	0.4-0.7	Intermittent catheterization	Voluntary bowel movement
15	<0.4	Intermittent catheterization	Fecally incontinent
16	0.4-0.7	Continent	Voluntary bowel movement
17	0.4-0.7	Dribbling/leaking	Voluntary bowel movement
20	NA	Continent	Voluntary bowel movement
21	NA	Intermittent catheterization	Fecally incontinent
22	>0.7	Intermittent catheterization	Voluntary bowel movement, soils with diarrhea
23	NA	Continent	Voluntary bowel movement
24	NA	Continent	Voluntary bowel movement

* Sacral ratios greater than 0.7 are considered normal [21]. Between 0.4 and 0.7, we found that the sacral ratio does not allow prediction of the degree of bowel. In the authors' series, there has never been a patient with bowel control and a sacral ratio less than 0.4 (unpublished data).

bowel movements. This result is better when we compare the posterior cloaca group with the best type of cloaca, the ones with a 1-cm common channel, in terms of prognosis for bowel control (85% vs 77%) [21]; therefore, we cannot overemphasize the importance of preserving the normal anal canal and thereby the capacity for bowel control in patients with a posterior cloaca.

4. Conclusions

The most important characteristic of the posterior cloaca is the presence of a normal anus, which differentiates this malformation from the classic cloaca. Often, a significant number of associated malformations are present and therefore should be suspected and diagnosed. The main goal during the operation should be *to not* mobilize the anus, so that fecal continence is preserved. A total urogenital mobilization, transperineally or with a transanorectal approach, is ideal for the repair.

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Discussion

David Wesson, MD (Houston, TX): Why can't you repair this from anterior to the rectum by making a midline incision in the perineum?

Alberto Pena, MD (response): If you estimated that the common channel is short, you can try to repair everything. Make sure that the patient either has a colostomy or a completely clean bowel, so if necessary, you divide the entire, first the anterior rectal wall and then the posterior rectal wall, provided you make a meticulous reconstruction,so the patients don't lose bowel control. What is not valid is to mobilize the anus circumferentially because that denervates the patient and, you will have an incontinent patient for sure.