

# TECHNICAL CONSIDERATIONS IN THE REPAIR OF CLOACAL VAGINAL DEFORMITIES

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## ABSTRACT

Various techniques of vaginal repair used in the reconstruction of cloacal deformities are discussed. In 4 years we performed primary reconstruction of cloacal anomalies in 4 children, 3 of whom had hydrocolpos and 1 vaginal agenesis. Vaginal reconstruction was done using a variety of techniques tailored to the primary anomaly. Distal vaginal occlusion related to ischemia occurred in 2 patients and in both the vagina was successfully reconstructed at a second stage procedure. Overall cosmetic and functional results were good. All patients are dry but require clean intermittent catheterization and 3 currently require a bowel regimen at followup of 18 months to 4 years.

KEY WORDS: cloaca; anus, imperforate; vagina; urogenital system

Cloacal anomalies are rare, occurring in an estimated 1 per 50,000 births annually, and represent one of the more complex forms of anorectal atresia in female subjects. They result from a mesenchymal defect that leads to failure in caudal growth of the sino-vaginal plate and the urogenital ridge that normally separate the allantois, vagina and hindgut, respectively, in the developing embryo. Patients typically present with a blank perineum and a sub-clitoral urogenital sinus that communicates with the bladder, vagina and colon. The plain external appearance gives little indication of the complexity of the reconstructive procedure. A central aspect of reconstruction is vaginal repair, which is best done in conjunction with an anorectal pull-through. Successful vaginal repair requires an appreciation of the frequent variations in cloacal pelvic anatomy as well as the variety of techniques available.

## CASE HISTORIES

*Case 1.* A white female neonate born at 32 weeks of gestation, weighing 1,600 gm., was noted at birth to have a blank perineum without introitus or anus and a sinus at the base of the clitoris (fig. 1). Initial physical examination also revealed a pelvic mass with abdominal distention. Ultrasound of the abdomen demonstrated 2 normal kidneys without hydronephrosis and a cystic pelvic mass, which did not evacuate with catheterization of the sinus tract. At laparotomy hydrocolpos was noted, and tube vaginostomy and splenic flexure colostomy with mucous fistula were performed. Postoperative sonogram revealed a long urogenital sinus leading to a large bladder, and communicating at the level of the bladder neck with a distended vagina and colon. The patient was treated with clean intermittent catheterization for 9 months before definitive reconstruction.

The reconstructive procedure was performed through a posterior sagittal approach with the patient in the prone jackknife position. The urogenital sinus was opened to the level of the bladder neck, where the vagina and colon were disconnected from the sinus and separated from each other (fig. 2). The sinus was tubularized to form a neourethra from the bladder neck to the perineum. A rectangular vaginal flap was created on the posterior aspect of the vagina and tubularized to facilitate mobilization of the vagina to the perineal skin (fig. 3). The perineal body was reconstructed and the

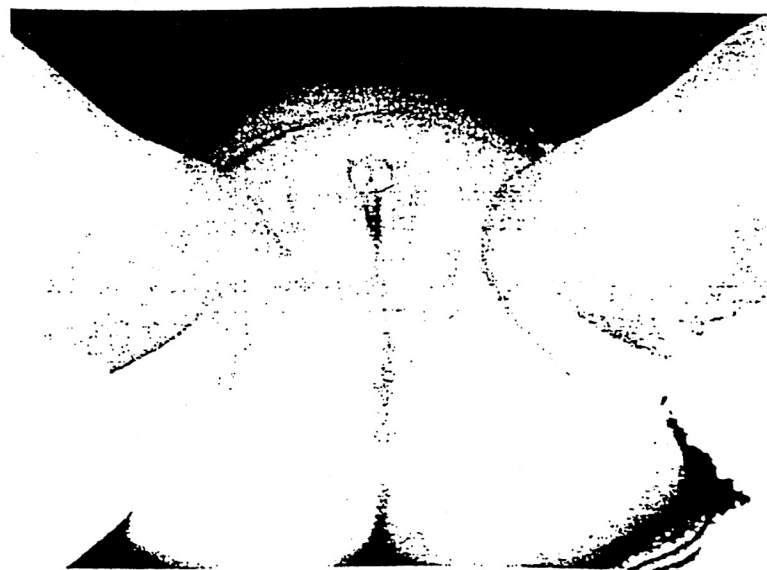


FIG. 1. Typical appearance of cloacal anomaly

vaginal flap was anastomosed to the skin. The sphincter complex was then reconstructed with the rectum in the anatomically correct position.

When the patient was 18 months old the perineal vaginal orifice was noted to be occluded. Transabdominal exploration was performed through an extended Pfannenstiel incision. A full thickness vaginal cuff was circumferentially dissected away from surrounding structures down to a common wall between the vagina and bladder neck, where it was detached, preserving the ovarian pedicle. The distal vaginal mucosa was completely excised using submucosal vaginal dissection and a persistent vesicovaginal fistula was repaired. The vaginal cuff was divided longitudinally to resect a vaginal septum and the vagina was then tapered before being pulled through to the perineal skin. At age 4 years the patient is dry on a clean intermittent catheterization program and has no fecal soiling on a daily bowel program.

*Case 2.* A full-term 2,000 gm. white female newborn was noted to have a blank perineum and a urogenital sinus at the base of the clitoris, in addition to abdominal distention and a palpable midline pelvic mass. Ultrasound of the abdomen demonstrated a hydronephrotic solitary left kidney and hydrocolpos. Double-barreled left transverse colostomy and cystoscopy were performed via the sinus, which revealed communication with the vagina and rectum above the level of the

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FIG. 2. Posterior sagittal exploration of cloacal anomaly. A, urogenital sinus. B, confluence of vagina and bladder neck. C, rectum disconnected from cloaca.

exploration was performed but the urogenital sinus was not opened, and the vagina and colon could not be visualized. Using total body preparation laparotomy was done via an extended Pfannenstiel incision that incorporated the vesicostomy. Bilateral ureteral catheters were placed through the open bladder, and the colon, vagina and bladder were separated by meticulous dissection. A full thickness vaginal cuff was developed circumferentially down to the common wall between the bladder and the vagina, where it was disconnected from the bladder, and the distal vaginal mucosa was excised using submucosal vaginal dissection (fig. 4). The urogenital sinus was closed at the level of the bladder neck. A vaginal septum was excised, and the vagina was reconstructed and mobilized on an ovarian pedicle. The vesicostomy was closed around a suprapubic cystostomy tube, and the vagina and colon were pulled through to the perineum. The patient was then returned to the prone jackknife position. The vaginal cuff was anastomosed to the perineal skin and the sphincter complex was reconstructed with the rectum in the correct anatomical configuration. The colostomy was closed 3 months later without incident. At age 3½ years the patient is infection-free and continent of urine and stool on a clean intermittent catheterization program.

*Case 3.* A black female neonate born at 37 weeks of gestation, weighing 2,360 gm., was noted to have a blank perineum and a urogenital sinus at the base of the clitoris. Physical examination revealed a distended abdomen with a palpable mass. A sonogram of the abdomen demonstrated grade II to III bilateral hydronephrosis. Tube vaginostomy and end-sigmoid colostomy with mucous fistula were performed. Postoperatively the vaginostomy tube drained little fluid and was removed. Subsequently urinary sepsis developed, which was treated with clean intermittent catheterization and prophylactic antibiotic therapy without success. A Blocksom vesicostomy was performed with dramatic reduction in the degree of vaginal dilatation and grade of hydronephrosis. Simultaneously the patient improved clinically without any further episodes of urinary sepsis.

At age 12 months posterior sagittal exploration was done with the patient in the prone jackknife position without opening the urogenital sinus. Because the vagina and colon could not be adequately mobilized from below, the patient was returned to the supine position using total body preparation and transabdominal exploration was performed. Bilateral ureteral stents were placed through the open bladder to facilitate dissection between the bladder, vagina and colon. A full thickness vaginal cuff was developed circumferentially



FIG. 3. Rectangular vaginal flap is used to mobilize vagina to level of perineum.

bladder neck. Subsequently multiple episodes of urinary sepsis developed, which were unresponsive to clean intermittent catheterization and urinary prophylaxis. When the patient was 5 months old a Blocksom vesicostomy was performed, following which the vagina was reduced and there were no further episodes of urinary sepsis.

When the patient was 10 months old posterior sagittal

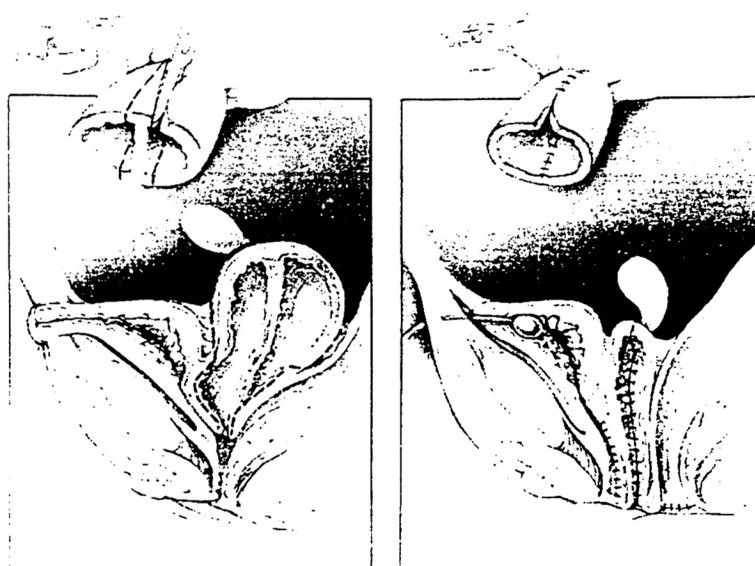


FIG. 4. Proximal vagina is mobilized on ovarian pedicle and reconstructed longitudinally before being pulled through to perineum. Dotted line indicates plane of dissection.

down to the level of the common wall between the bladder and vagina, where the vagina was disconnected from the urogenital sinus that was closed. The distal vaginal mucosa was excised using submucosal vaginal resection. A vaginal septum was excised, and the vagina was reconstructed and mobilized on an ovarian pedicle. The vesicostomy was closed around a suprapubic cystostomy tube, and the rectum and vagina were pulled through to the perineum. The patient was returned to the prone jackknife position. The vaginal cuff was anastomosed to the perineal skin and the sphincter complex was reconstructed with the rectum in the correct anatomical position.

When the patient was 16 months old the vaginal orifice was noted to be occluded. Limited perineal exploration revealed the distal vagina approximately 2 cm. from the perineal skin. The distal vaginal canal was reconstructed using a split thickness skin tube 1/16,000-inch thick (fig. 5). The colostomy was closed several months later without incident. At age 3 years the introitus is widely patent, and the patient is on a clean intermittent catheterization and bowel program without any further episodes of urinary sepsis.

*Case 4.* A black female neonate born at 31 weeks of gestation, weighing 1,500 gm., was noted to have a blank perineum and a urogenital sinus beneath the clitoris. When she was 1 week old end-sigmoid colostomy was performed. She was referred to our institution at age 3 months and a urogenital sonogram revealed no clear communication between the urogenital sinus, vagina or colon. An excretory urogram demonstrated bilateral ectopic ureters with left hydronephrosis.

When the patient was 16 months old posterior sagittal exploration, leaving the urogenital sinus intact, did not reveal the vagina or distal colon. Using total body preparation the patient was turned to the supine position. Exploration performed through an extended Pfannenstiel incision demonstrated complete vaginal agenesis with a bicornuate uterus and bilateral ectopic ureters. Trans-trigonal bilateral ureteral reimplantation was done. Subsequently an artificial vagina was constructed using a vascularized segment of distal colon that was tapered and pulled through to the perineum (fig. 6). The distal colon was then pulled through to the perineum in a dorsal position behind the neovagina. The patient was turned to the prone jackknife position and the neovagina was anastomosed to the perineal skin dorsal to the urogenital sinus. The sphincter complex was reconstructed with the rectum in the correct anatomical position. At age 2½ years the patient is continent of urine on a clean intermittent catheterization program.

#### DISCUSSION

Surgical correction of cloacal anomalies is complicated by variable anatomy of the primary defect, frequent occurrence

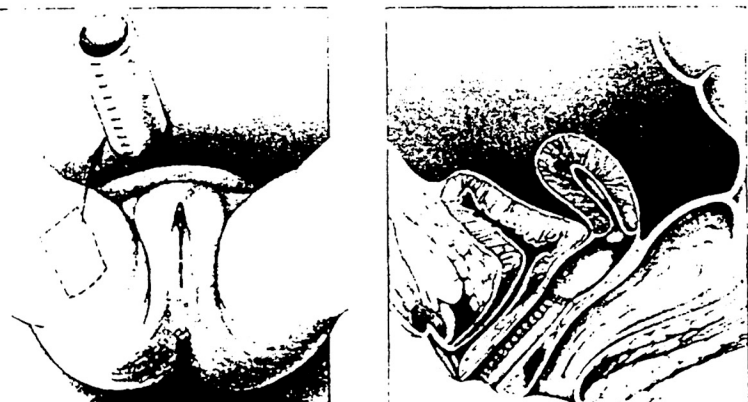


FIG. 5. Second stage procedure. Tubular skin graft is used to connect vagina and perineal skin.

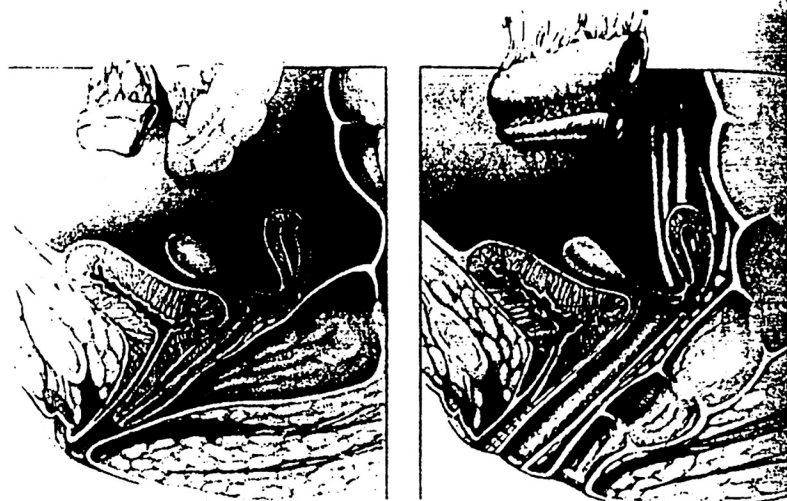


FIG. 6. Sigmoid is mobilized on mesenteric pedicle and tapered to form sigmoid vagina.

of associated urinary malformations and relative inaccessibility of pelvic structures. Our understanding of cloacal anomalies has been greatly enhanced by the work of Peña and Hendren, among others. de Vries and Peña developed the technique of posterior sagittal reconstruction, which allows precise visualization and reconstruction of the urogenital sinus and sphincter complex.<sup>1-3</sup> Hendren emphasized the importance of combining reconstruction of the urinary tract with the cloacal anomaly to prevent chronic urinary sepsis and renal deterioration.<sup>4-6</sup> Others have delineated the variable anatomical relationships commonly observed in cloacal malformations.<sup>7,8</sup>

Cloacal anomalies are readily diagnosed in female subjects by the characteristic findings of a blank peritoneum and sub-clitoral sinus tract. Embryologically they result from a mesenchymal defect that leads to a urogenital sinus malformation in conjunction with imperforate anus. Anatomically they are extremely variable relative to the configuration of the urogenital sinus and its relationship to the vagina and rectum. Approximately 60% of children with cloacal anomalies have associated upper urinary tract malformations and a similar percentage have associated hydrocolpos caused by partial or complete obstruction of the vaginal outlet.<sup>5</sup> Frequently hydrocolpos may contribute to bladder outlet and/or ureteral obstruction, urinary stasis and sepsis. When this occurs intermittent catheterization, tube vaginotomy or vesicostomy may be required.<sup>9</sup>

Because of the inherent diversity of cloacal anomalies, it is extremely important to define the precise anatomy before attempting repair. Sonography and pyelography may be used to define upper urinary tract malformations as well as the possibility of urinary obstruction caused by hydrocolpos. Sinography and sinoscopy are essential to delineate the course of the urogenital sinus, and its relationship to the vagina and rectum. If the urethra and vagina join distal to the external urethral sphincter, then flap vaginoplasty may be performed via a perineal approach.<sup>6</sup> However, in most cases the vagina enters the urogenital sinus at a higher level, necessitating a more complex pull-through vaginoplasty. In these cases urine often follows the path of least resistance into the vagina, where it becomes entrapped, leading to hydrocolpos. Hydrocolpos occurred in 3 of our 4 patients and in all 3 the vagina was septated, hindering efforts to provide drainage before definitive reconstruction. Our patient without a dilated vagina was found to have vaginal agenesis. Vaginal dilatation may not occur when the urogenital sinus is relatively short or wide but this is unusual in our experience. More likely the absence of vaginal dilatation on sonography or sinography may indicate vaginal agenesis.

The technique of cloacal reconstruction is guided by preoperative evaluation and is best performed in patients 9 to 12 months old. It is advantageous to perform posterior sagittal

exploration initially since the entire reconstructive procedure may be done through this approach in a number of patients. A critical step in the procedure is the disconnection of the vagina from the urogenital sinus, which is then tubularized to create a neourethra. The decision to open the entire sinus is based on its length and width. When the urogenital sinus is short and wide, it may be longitudinally opened and tapered to enhance urinary continence. However, when it is longer than 3 cm. and/or less than 10F in diameter this technique is not necessary and increases the risk of stenosis or urethral-vaginal fistula.

The major considerations in performing a combined abdominal-posterior sagittal procedure are the length and viability of the vaginal flap as well as associated upper urinary tract malformations. Mobilization of the vagina to the perineal skin may be accomplished from below by circumferential dissection and is facilitated by the use of a rectangular vaginal flap<sup>3</sup> or perineal skin flaps.<sup>3, 6</sup> When insufficient length or vascular compromise prevents vaginal-perineal anastomosis, transabdominal mobilization of the vagina preserving the ovarian pedicle may be required. This approach also permits excision of vaginal septations.

Another important consideration is the occurrence of associated congenital urinary malformations. Certain malformations, such as ureterovesical junction obstruction, ectopic ureter or ureteral duplication, are more efficiently managed via a transabdominal approach. In addition, cystotomy and placement of ureteral stents may facilitate dissection of the vaginal cuff.

Vaginal reconstruction in patients with cloacal anomalies and vaginal agenesis usually requires a combined abdominal-posterior sagittal procedure.<sup>10</sup> The diagnosis is confirmed by finding normal ovaries and salpinx along with absent or dysplastic vagina and uterus. Construction of an artificial vagina in this instance may be accomplished with a split thickness skin tube<sup>8</sup> or a tapered segment of sigmoid colon based on its vascular pedicle.<sup>11</sup> The sigmoid vagina offers several advantages in that it is self-lubricated and does not appear to require repeat dilation. Like the skin tube, the sigmoid vagina may be tapered to any width but it does not require a postoperative stent or dilation.

Complications of cloacal repair are usually related to the vaginal cuff. The most difficult complication is vesicovaginal or urethrovaginal fistula, which has been reported in 11% of patients<sup>3</sup> but occurred in none of our patients. This complication may present with urinary sepsis and/or incontinence, and is best repaired by colostomy and subsequent perineal exploration with the patient in the Kraske position.

More commonly, distal vaginal stricture or occlusion may occur due to ischemia of the vaginal flap. This complication may be prevented by gentle handling and adequate mobilization of the vaginal flap. Repair of distal vaginal occlusion depends on the type of reconstruction previously used. For example, after occlusion of a rectangular vaginal flap transabdominal vaginal reconstruction may be performed by mobilizing the vagina on its ovarian pedicle (case 1). Vaginal occlusion after transabdominal vaginoplasty may be sal-

vaged by the use of a tubular split thickness skin graft. In the event of complete or near complete loss of the vaginal cuff construction of an artificial vagina using a vascularized segment of sigmoid colon may be performed.

The results of total pelvic reconstruction for cloacal anomalies are generally good but clean intermittent catheterization and bowel programs are frequently required to ensure that patients remain clean and dry. All of our patients are dry but they require clean intermittent catheterization. Peña showed in a much larger series that urinary and bowel continence partially depends on the integrity of the lumbosacral spine.<sup>3</sup> In 1989 he reported that 74% of patients with a normal sacrum were continent versus 28% with an abnormal sacrum. In our series only 1 patient had a normal sacrum and she was continent of stool at age 3½ years. Our other patients have an abnormal sacrum and require a bowel program or are not yet old enough to be toilet trained.

In conclusion, posterior sagittal exploration may be combined with laparotomy when indicated for the treatment of cloacal anomalies. The technique of vaginal reconstruction used is variable and depends on the configuration of the urogenital sinus in relation to the vagina as well as the presence of vaginal dilatation. Associated urinary tract malformations are frequent and should be repaired at the time of definitive reconstruction. Urinary and fecal incontinence is often related to associated lumbosacral spinal anomalies, and may be treated by clean intermittent catheterization and a bowel program. Complications most frequently involve the vaginal cuff and may be repaired using a variety of techniques.

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