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The surgical treatment of cloaca

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ABSTRACT

Similar to other anorectal malformations, cloaca also represents a spectrum of defects that goes from "benign" cloaca with a good functional prognosis that can be repaired with a relatively simple surgical technique, to very complex malformations with many anatomic variations that require different surgical maneuvers to be able to successfully reconstruct those patients.

The group of patients born with a "benign" type of cloaca will have bowel and urinary control, will become sexually active and may get pregnant and deliver by cesarean section. All this is possible, provided the malformation is repaired with a meticulous and delicate technique. Fortunately this represents more than 50% of all cloacas. Our belief is that the surgical technique to repair this group of defects is reproducible and can be taught to pediatric surgical trainees.

On the other hand, complex cloaca with a common channel longer than 3 cm should be repaired by surgeons fully dedicated to repair these malformations.

The experience reported in this paper is based on 570 patients with cloaca operated by Dr. Alberto Peña and the author in the last 8 years.

Newborn management

At birth, the diagnosis of a cloaca is made by identifying a single perineal orifice, usually located where the urethra would normally be observed.[1]

The genitalia appearance in cloaca patients varies according to the complexity of the cloacal malformation, ranging from almost normal looking genitalia with well-formed buttocks, good midline groove, and good delineation of the sphincter mechanism by a well-defined anal impression; to hypertrophic folds of skin in the area of a single tiny perineal orifice with a "flat bottom" and a not well-defined anal impression. The first group tends to correlate with more benign type of cloaca (< 3 cm common channel) and with better future functional prognosis. In the second group, the hypertrophic folds of skin are frequently misdiagnosed as disorders of the sexual development. In our series of 570 cloacas, 66 were misdiagnosed as "intersex," before coming to our institution, and received unnecessary studies to confirm that they were female patients. More seriously, three patients were started on steroids that in retrospect, were not indicated.

During the first 24 h of life, before taking the patient to the operating room, the clinician should rule out important associated anomalies (Table). The one that is important to emphasize is the presence of a dilated vagina, known as hydrocolpos,[2] which happens in 38% (217/570) of the patients born with a cloaca. The hydrocolpos may compress the trigone of the bladder causing extrinsic uretero-vesical obstruction, bilateral megaureters, and hydronephrosis; it can also get infected and therefore it should be properly drained with an indwelling transabdominal catheter placed during the colostomy opening. This tube should remain in place until the main repair, in order to avoid re-accumulation of fluid. Since 57% (325/570) of the patients with cloaca have duplication of the Mullerian structures (two hemi-vaginas and two hemi-uteri) it is important to assure that both vaginas are properly drained. A window can be created in the vaginal septum allowing for a single tube to drain both hemi-vaginas. During the operation, the uterus and the cervices should be identified by visualization and palpation and the drainage tube should be placed away from the Fallopian tubes, uterus, and cervices. Trauma to these structures could impair future fertility. The drainage tube can be a pigtail or a Foley catheter, the surgical technique is the same as a suprapubic or gastrostomy tube placement. When the vagina is very dilated a tubeless vaginostomy is also an option. Depending on the size of the hydrocolpos, an infra-umbilical midline incision or an oblique left side incision can be used for the colostomy opening and hydrocolpos drainage. Laparoscopy has also been described as an option for hydrocolpos drainage.[3]

The colostomy recommended for cloaca patients is the same as that recommended for all patients with anorectal malformation[4]: a descending divided colostomy with a tiny distal mucous fistula located far enough from the main stoma to allow the placement of

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Table
Associated anomalies that should be ruled out during the first 24 h after birth.

Associated anomalies	Diagnostic method
Cardiac anomalies	Echocardiogram
Esophageal atresia	NG tube placement
Duodenal atresia and vertebral anomalies	Babygram
Hydronephrosis and hydocolpos	Kidney and pelvic ultrasound
Tethered cord	Spinal ultrasound
Sacral anomalies	Sacral radiograph AP and lateral

the stoma bag covering only the proximal stoma. During colostomy opening the distal segment should be cleared of all meconium with copious normal saline irrigation.

Before the main repair the patient should have radiological studies (distal colostogram and injection of contrast through common channel/bladder/vagina) as well as endoscopy of the common channel for the surgeon to plan its reconstruction. The main information that the surgeon should try to obtain preoperatively include: location of the rectum in relationship to the sacrum (is it reachable from a posterior sagittal approach?), length of distal bowel available for the pullthrough (is there enough length of bowel to reach the perineum?), length of the common channel (less or more than 3 cm?), and presence, size, and location of the vagina. Even with very sophisticated studies (3D cloacogram[5] and pelvic MRI), sometimes the entire anatomy can only be figured out during the operation and that is when experience with dealing with this defects helps to choose an appropriate operative maneuver to solve different anatomic problems.

After the colostomy has been opened, the hydrocolpos is drained; the main repair can be done when the patient is stable, growing, and developing normally. The age also depends on the experience of the surgeon and infrastructure of the institution (anesthesia and intensive care unit).

Cloacas with a common channel shorter than 1 cm (46/570)

These patients are treated almost like those that are born with a recto-vestibular fistula. They do not require total urogenital mobilization as the urethra is perfectly visible in its natural location, even when it is a little hypospadic. The first step consists of approaching the patient through a posterior sagittal incision, identifying the posterior wall of the rectum, clearing the posterior and lateral walls of the rectum, and then performing a very meticulous dissection to separate the anterior rectal wall from the posterior vaginal wall. Once the separation is completed and the rectum reaches the center of the sphincter mechanism without tension, the posterior and lateral walls of the vagina are mobilized in order to create an adequate introitus (Figure 1). The limits of the sphincter are delineated with the use of an electrostimulator and the perineal body is created. An anoplasty is performed. Our series includes 46 cases with a short common channel.

Cloacas with a common channel length between 1 and 3 cm (184/570)

Patients with a common channel length shorter than 3 cm can be repaired through a posterior sagittal approach. After the separation of the rectum from the vagina, the surgeon should perform a maneuver called total urogenital mobilization.[6] To accomplish this, the posterior wall of the vagina and the entire common channel is divided to expose the urethral and vaginal openings. Multiple 5–0 silk stitches are placed taking the edges of the vagina and common channel. Another series of stitches are

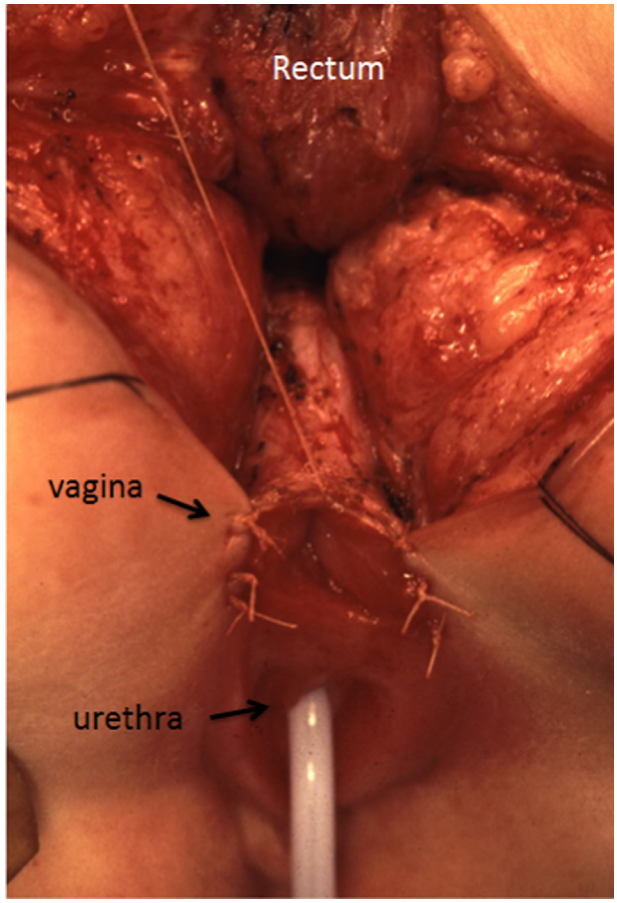

Fig. 1. The surgical repair in a cloaca with a 1 cm common channel length. The rectum is already separated from vagina. Lateral and posterior walls of the vagina are already mobilized. The urethra was left untouched.

placed in a horizontal manner, 5 mm proximal to the clitoris in order to provide uniform traction (Figure 2). With the use of a needle tip cautery, the urogenital sinus is divided between the clitoris and the traction sutures. Dissection is performed between the anterior wall of the urogenital sinus and the posterior aspect of the pubis. Between these two structures there is a natural plane,

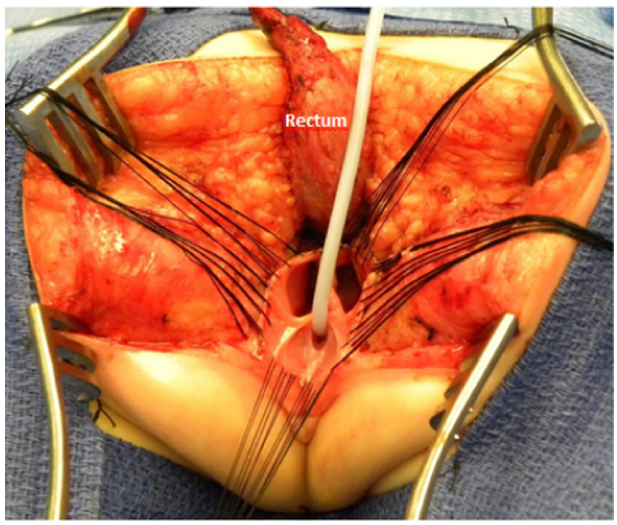

Fig. 2. The surgical repair in cloacas with a common channel length that measures less than 3 cm. After separation of the rectum from the vaginas, multiple 5–0 silk stitches are placed around the urogenital sinus; 5 mm from the clitoris and in the lateral and posterior wall of the vaginas.