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ORIGINAL ARTICLE

The Persistent Cloaca

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Abstract

Normally, at birth, females have separate perineal orifices for the urinary, reproductive and digestive tracts, while males have individual openings for the urogenital and digestive tracts. However, during fetal development, both sexes pass through a stage when all the systems empty into a common chamber, the cloaca. The cloaca opens to the exterior through a solitary opening until it divides into the various systems. Occasionally, the tracts do not separate and the baby is born with a persistent cloaca. It is a complex anomaly that is often associated with abnormalities in other systems that have to be taken into consideration to manage the patient appropriately.

Keywords: Persistent cloaca, anorectal anomalies, cloacal anomalies

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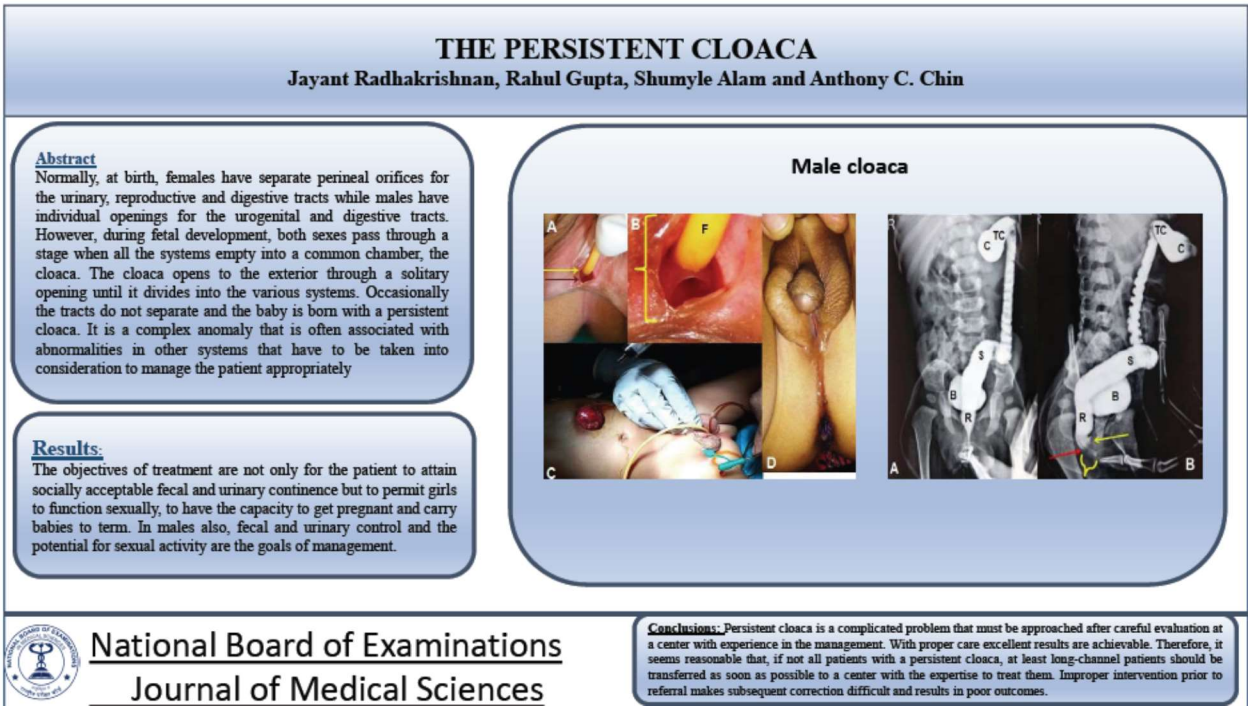
Graphical Abstract



Introduction

The overwhelming majority of placental mammals have separate orifices from which they evacuate urinary, genital and intestinal contents. In contrast, amphibians, birds, and reptiles have a single cavity, the cloaca (from Latin: sewer). Urine, stool and products of reproduction are stored in the cloaca and expelled through its solitary orifice [1].

Embryology

In the human fetus, the cloaca is a temporary widening of the hindgut at 5 weeks of gestation. It is lined with endoderm and it divides into the posterior hindgut and the anterior urogenital section by the 7th week. The original hypothesis was that a wedge of mesenchyme, known as the urorectal septum (URS), develops between the allantois and the hindgut and it grows in a craniocaudal direction until it fuses with the cloacal membrane, thereby dividing the cloaca into the urogenital sinus (UGS) anteriorly and the rectum posteriorly. It was further believed that the failure of the URS to extend caudally and to fuse with lateral infoldings of the cloaca, Rathke's folds, resulted in anorectal anomalies, including the persistent cloaca. This theory was upended in 1986 when van der Putte demonstrated in the pig embryo that the shift of the dorsal part of the cloaca and the adjacent gut to the body surface of the tail groove is the predominant event in the development of the anorectum. In other words, the caudal extension of the URS is a secondary, passive process that results from differential growth in the cloacal region which, in turn, causes the caudal body axis

of the embryo to unfold. The dorsal part of the cloacal membrane must regress for this step to transpire. The genesis of this part of the cloacal membrane prevents the normal shift of the anorectum to the surface of the body, resulting in the development of anorectal malformations. According to van der Putte the size and nature of this defect dictate the type of resultant anorectal anomaly. Van der Putte further postulated that deformities of the cloacal membrane could be the cause of cloaca-derived malformations [2]. In 1995, Kluth et al. compared the cloacal region of normal rats to that of semi-dominant gene mice (SD mice), as the latter often have abnormal cloacae. They found that in the embryos of abnormal mice, the cloacal membrane is too short and the site of the future anal opening is not identifiable, while in normal rats, this site can be identified soon after the cloacal membrane forms. They found a variety of cloacal malformations in SD mice that conform to the types of anorectal malformations found in humans. Their findings also confirmed that a normal URS forms passively when cloacal development is normal and that the URS is not the primary agent in normal cloacal development [3]. In 2018, passive growth of the URS was demonstrated in human embryos between 4 and 10 weeks of development. The authors found that the ventral and central areas of the cloaca grew rapidly, while there was almost no growth in the cranial and dorsal areas. This differential growth resulted in separate urogenital and anorectal compartments. Later, differential growth straightened the curved caudal body axis of the embryo and in the process, the URS advanced caudally. Dysregulation of dorsal cloacal growth could be the cause of anorectal malformations [4].

Although we now believe that we understand cloacal development, we still do not know why normal differentiation of the cloaca is thwarted in some instances [5].

Incidence

Persistent cloaca is estimated to occur in 1:20,000–50,000 births. It is reported almost exclusively in females with a 46 XX chromosomal structure. However, it has been identified in human [6,7] and canine [8] males on rare occasions. Whether it is truly rare in males or has been misdiagnosed as an anorectal malformation with a fistula to the urethra is an unanswered question.

Pathological anatomy

In Females, The basic abnormality in female patients is a confluence between the bladder-urethral complex, the Müllerian structures and the hindgut. All the structures empty into one cavity, which opens to the exterior through a solitary perineal orifice under a hooded clitoris. However, there is considerable internal variation amongst the components that must be delineated carefully in each patient before settling on a plan for surgical correction [9,10].

To manage these girls appropriately, the following issues must be defined clearly:

1. Whether the components fuse at the same location or at different sites. The site of communication between the components must be precisely localized.

2. The length of the common channel helps determine the surgical approach and the likelihood of eventual urinary continence. It has generally been accepted that a common channel shorter than 3cm. can be repaired entirely from a sacral approach, while in those with a common channel longer than 3 cm an abdominal exposure is also required [11-13].
3. The length of the urethra also matters. A urethral length of 1.5 cm with a short common channel requires a simpler perineal operation and results in better continence. On the other hand, if the urethra is short and the common channel is long, not only is abdominal exposure required, but ultimately, continence is not as satisfactory [14].
4. An anatomically normal bladder could be distended if the urethra is obstructed, or it may fail to empty if associated spinal lesions render it neurogenic. It could also be distended with mucus if the Müllerian structures and/or the hindgut drain directly into it.
5. The greatest variations occur in the Müllerian structures and they must be carefully identified. Any of the following variations are possible: a normal uterus and vagina opening at a common confluence with the bladder and hindgut: a normal uterus perched atop a vagina filled with mucus and/or urine: a didelphic uterus in which one or both sides are distended. Furthermore, the septum between the halves may be partial or complete and the vagina and uteri may be symmetric or asymmetric if one side is atretic. Finally, the two Müllerian ducts may enter the bladder separately [13].
6. The hindgut typically enters the confluence, but rarely, it may end blindly without any communication with the other structures. In addition, when the two Müllerian ducts do not fuse and enter the bladder individually, the hindgut may also drain between the two of them directly into the bladder.
7. It is also essential to determine whether the hindgut has descended far enough towards the perineum to be brought down to its desired location using only a perineal approach or whether the abdomen would have to be entered to free up an adequate length of the rectum.

Cloacal anomalies may be classified as follows [9]:

1. Persistent cloaca. It is the most common lesion and has a solitary perineal opening.
2. Cloaca variant.
3. Posterior cloaca.
4. Posterior cloaca variant.
5. Urogenital sinus (UGS).
6. Cloacal dysgenesis. It has no perineal opening and is the least common and most severe of all these anomalies.

In Males. In males with a persistent cloaca, the urinary tract and the rectum drain into a short common channel, which constitutes the only external outlet for both structures. On occasion, atrophic Müllerian

structures may also communicate with the common channel [6,7].

Associated abnormalities

To facilitate diagnosis and therapy, other abnormalities seen in these patients could be divided into two broad groups: associated developmental lesions and coexistent complications.

Associated developmental lesions. Persistent cloaca is the most severe form of anorectal anomaly; hence, vertebral, cardiac, tracheo-esophageal, renal and limb (VACTERL) abnormalities are to be expected. It is particularly important to look for spinal and renal/urinary abnormalities [15].

In patients with persistent cloaca, vertebral anomalies affect the lower vertebrae and the sacrum. Some vertebrae may be missing or deformed, the sacrum could be deformed or absent and a tethered spinal cord may create a neurogenic bladder and also cause problems with fecal control and evacuation after reconstruction.

Renal and urinary abnormalities are also common. Renal agenesis, horseshoe kidney, hydronephrosis, ureteric duplication, vesicoureteric reflux and obstructive uropathy have all been reported. It is important to note that sometimes these problems may not be identified initially.

Numerous limb abnormalities of varying severities may also coexist. Examples are polydactyly, syndactyly, radial aplasia and hypoplasia or displacement of the thumb.

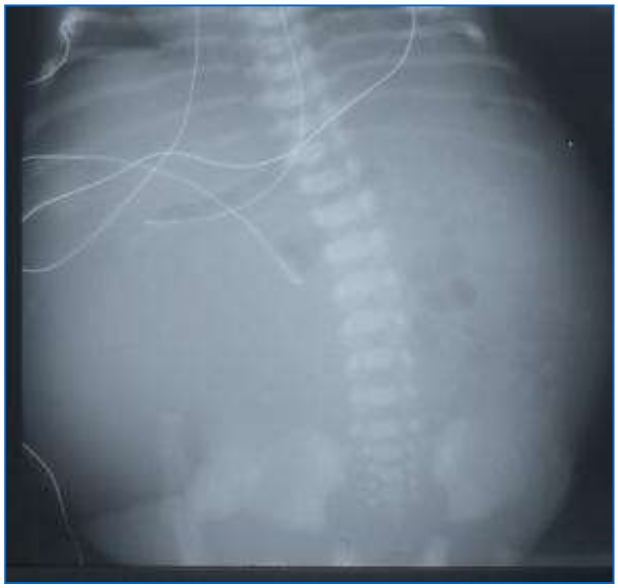
Coexistent complications. Coexistent complications involving the bladder, Müllerian structures and hindgut mentioned previously are much more of a problem and require careful evaluation and management either before or when the cloaca is being corrected.

Antenatal diagnosis

Antenatal ultrasonography is the obvious screening test since it is noninvasive, easy to carry out, avoids radiation and is inexpensive. Suspicious findings consist of cystic lesions in the pelvis of a female fetus, a highly placed rectum, fetal ascites, renal abnormalities, an irregular appearance of the bladder, oligohydramnios and ambiguous genitalia. However, all these findings could be due to other pathologies. It is claimed that a fluid-filled, dilated colon with intraluminal enteroliths and meconium in the form of debris in the urinary tract constitutes the most specific combination of findings to diagnose a persistent cloaca [9]. Additionally, the anal sphincter can also be evaluated by ultrasonography.

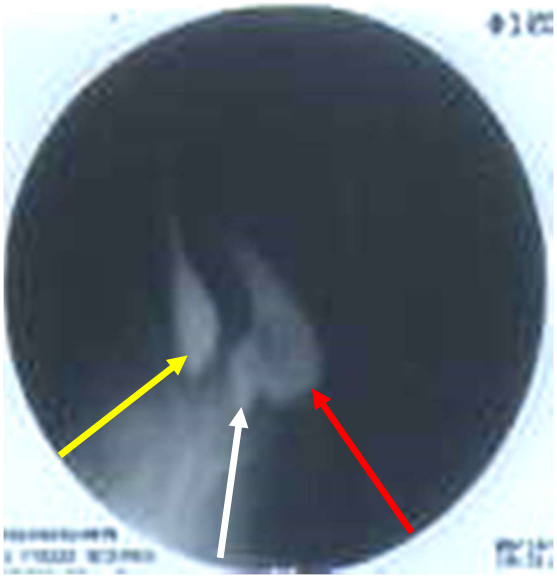
Magnetic Resonance Imaging (MRI) of the fetus is the definitive test to confirm the diagnosis and also to evaluate each entity in detail. MRIs are carried out under carefully defined criteria to help differentiate the six variants of cloacal abnormalities mentioned above [9] (Figures 1 to 5).

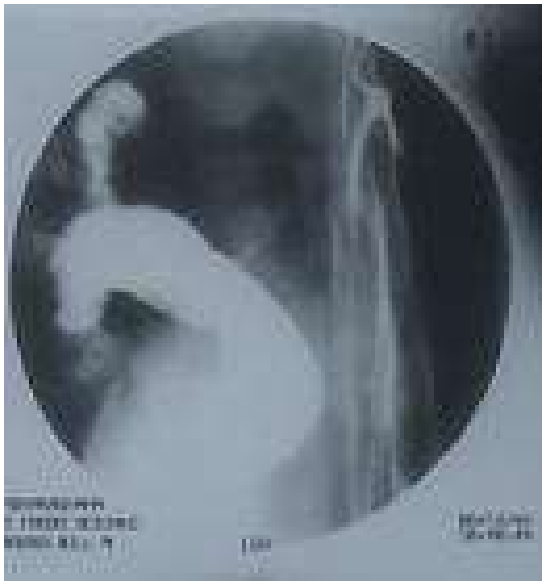
1A.



1B.

1C





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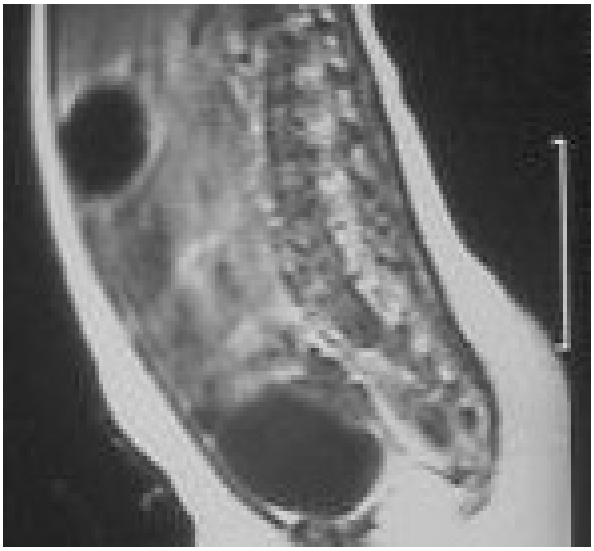

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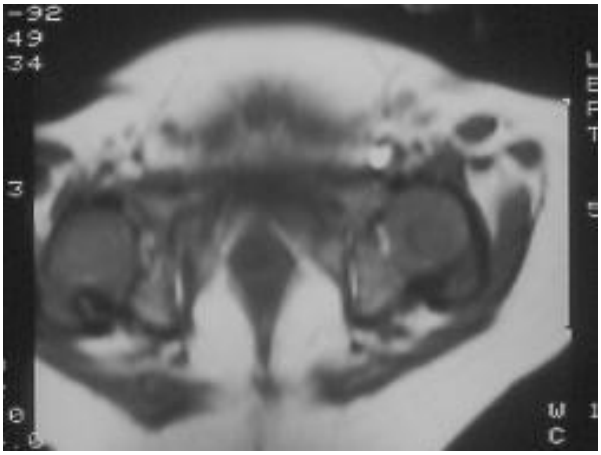
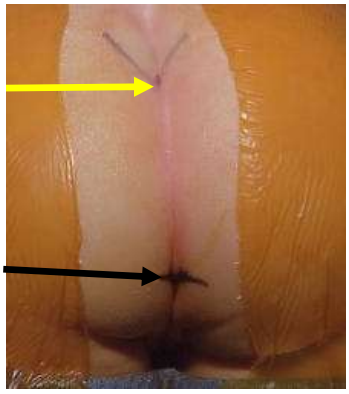
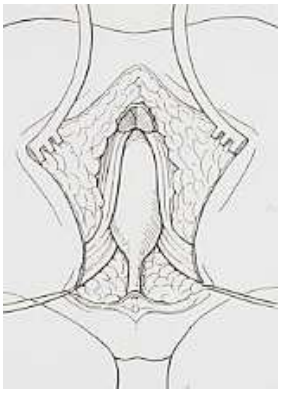


Figure 1. Female persistent cloaca case #1. This patient was referred for evaluation as her abdomen was distended, and no perineal orifices were visible to the examiner.
1A. Patient is supine. She has mild clitoral hypertrophy and excess of preputial skin. The solitary orifice at the base of the clitoris is hidden by the skin fold. The midline raphe is well-defined and she had a well-developed anal sphincter.
1B. Plain anteroposterior abdominal radiograph at birth demonstrating massive abdominal distension with a ground glass appearance and a few loops of intestine displaced upwards by a very large pelvic mass, the hydrocolpos. The hydrocolpos was catheterized and drained through the urogenital sinus (UGS) and a diverting colostomy was created in the descending colon.
The patient was studied in detail once she was thriving.
1C. Lateral radiograph of sinogram. The yellow arrow points to the bladder, and the white arrow indicates the UGS and vagina. The red arrow demonstrates the hindgut.
1D. Lateral radiograph of the distal colostogram through the colostomy. There is an adequate length of the distal colon for a pull-through procedure.
1E. MRI: Normal spinal cord with no tethering.
1F. MRI: Normal anal sphincter mechanism on transverse section through the pelvis.

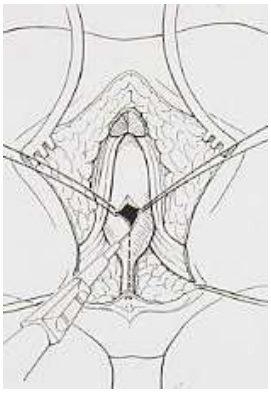
1G.



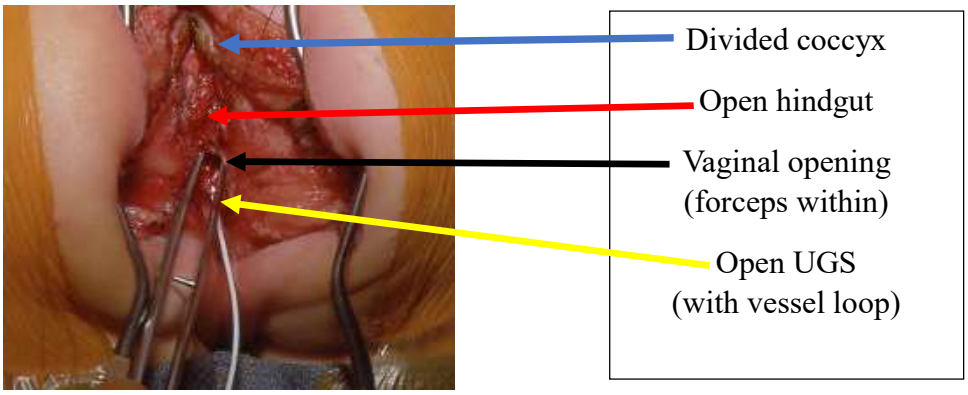
1H.



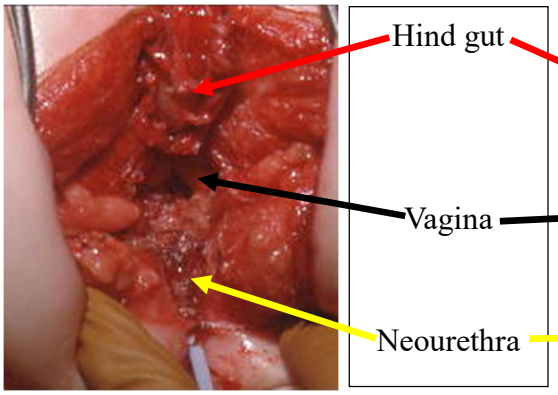
1J.



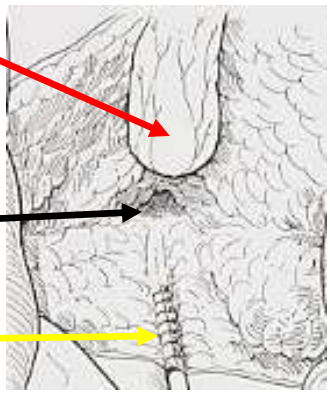
1K.



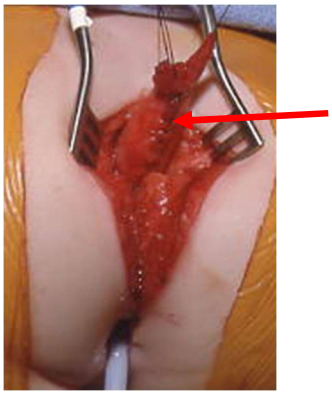
1L.



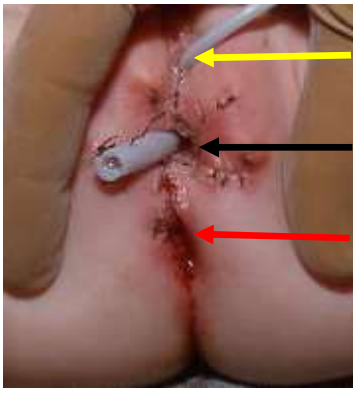
1M.



1N.



1O.



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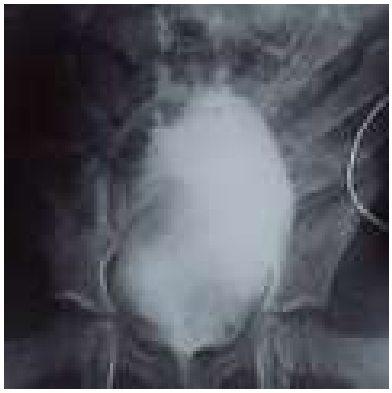


Case #1 cont'd. Steps of surgical reconstruction by posterior sagittal ano-recto-vagino-urethroplasty (PSARVUP)

1G. With the patient in the knee-chest position the tip of the coccyx (yellow arrow) and the center of the anal sphincter (black arrow) delineate the extent of the incision.

1H. The midline incision is deepened until the posterior wall of the cloaca and urogenital sinus are clearly visualized.

1J. The posterior wall of the cloaca and UGS are incised down to the perineal opening.

1K. Photograph of the open cloaca demonstrating the open hind gut (red arrow), the vaginal opening (black arrow) and the open UGS (yellow arrow).

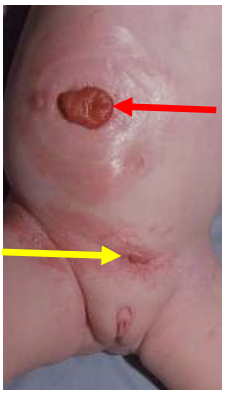
1L and 1M. Photograph and illustration of the structures after dissection is complete. Hind gut (red arrow), vaginal orifice (black arrow) and urethra reconstructed from the UGS (yellow arrow).

1N. The urethra has been reconstructed and the vagina mobilized and sutured to the perineum. The hind gut has been mobilized and tapered in preparation for it to be sutured at the perineum (red arrow).

1O. Appearance of the perineum 1 week after surgery with the patient supine. The foley catheter (yellow arrow) is in the neourethra, the rubber tube (black arrow) is in the vagina and the neoanus is visible posteriorly (red arrow).

1P. Antero-posterior radiograph of the postoperative voiding cystourethrogram (VCUG) demonstrating good capacity, shape and retaining ability of the bladder. She was able to void spontaneously.

2A.



2B.

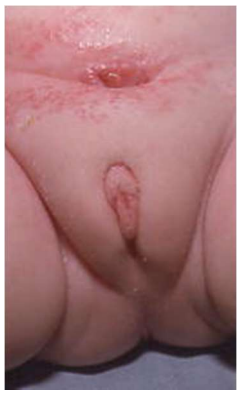


Figure 2. Female persistent cloaca case #2. This patient was referred to us at 4 months old as she was not thriving. Her birth weight was 7 lbs. and she had not gained any weight in the 4 months. She was also septic. At birth a diverting ileostomy had been carried out elsewhere. Later they also carried out a

vesicostomy believing that her recurrent hydrocolpos was due to retrograde drainage of urine into the vaginae. The vesicostomy did not correct the problem.
2A. Photograph demonstrating a loop ileostomy (red arrow) and vesicostomy (yellow arrow).
2B. Close up of the vesicostomy showing severe candidiasis.

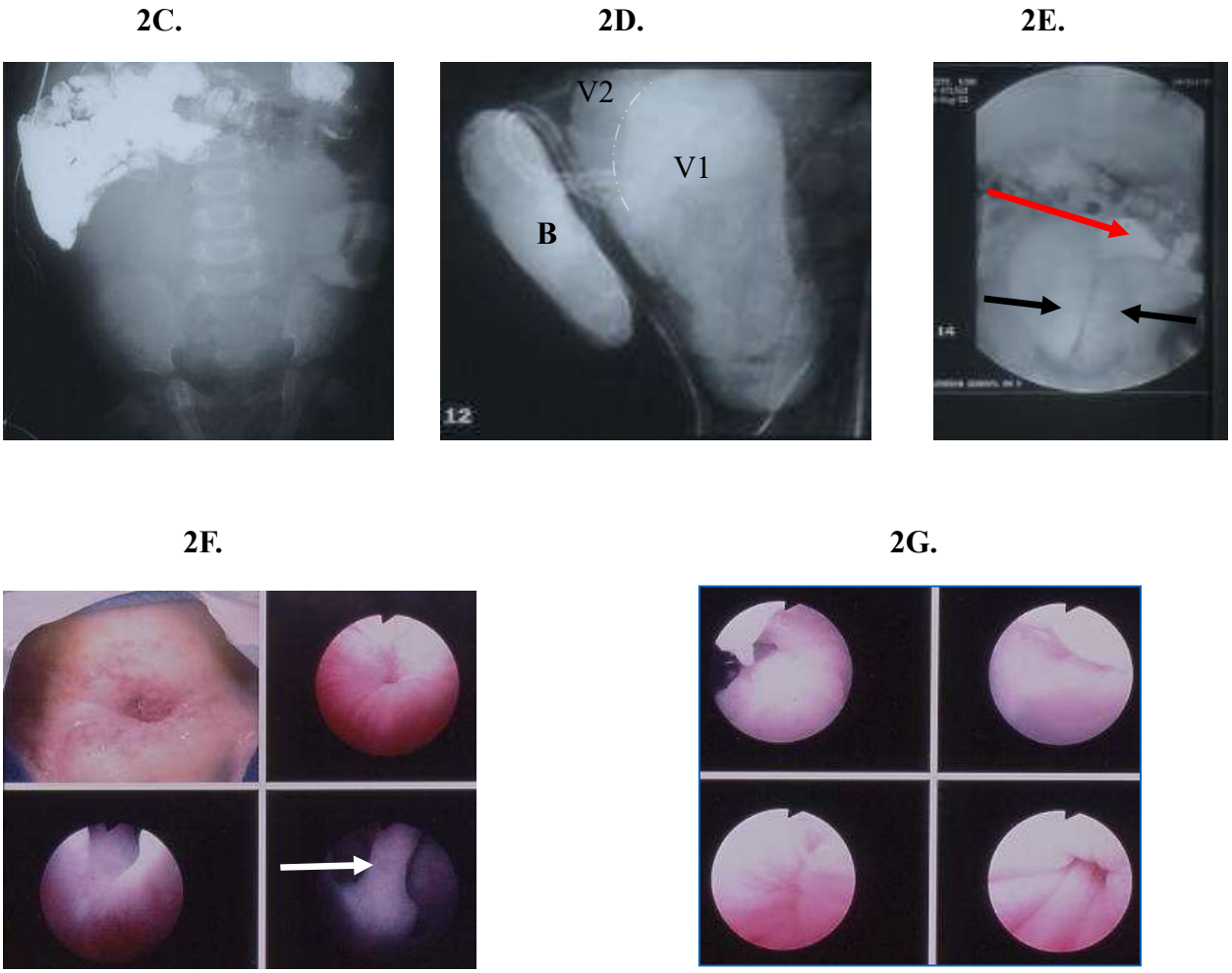
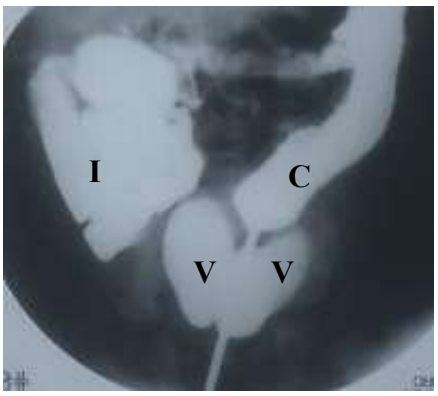


Figure 2. Female cloaca case #2 (cont'd.). Preoperative studies

2C. Upper gastrointestinal contrast study revealing upward displacement of intestines by a large mass (hydrocolpos) arising from the pelvis.
2D. Sinogram carried out through the UGS after plugging the vesicostomy. The bladder (B) is seen to the left and the large double vagina to the right. The two halves of the vagina are V1 and V2. The dashed white line in an arc demarcates the upper extent of the left vagina with the right vagina visible above it.
2E. Antero-posterior view of the sinogram after emptying the bladder. The two hemivaginae (black arrows) are visualized and contrast also enters the colon (red arrow).
2F. Cystoscopy through the vesicostomy. Top left: Vesicostomy. Top right: Bladder neck visualized from within the bladder. Bottom left: Septum between the hemivaginae visualized upon entering the UGS from the bladder. Bottom right: Orifice visible on the septum between the vaginae (white arrow).

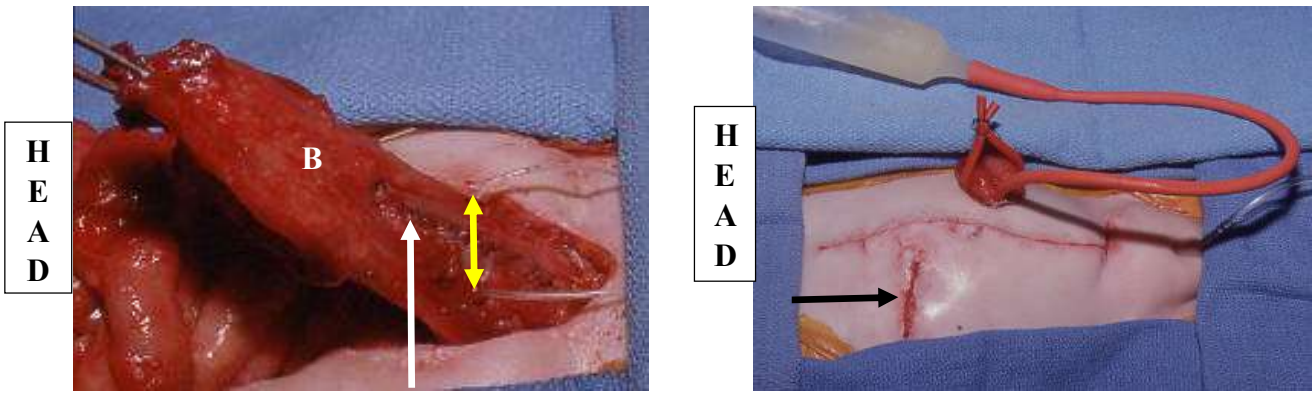
2G. Top left: Cystoscope passed from the perineum through the UGS into the orifice on the intervaginal septum. Top right, Bottom left and Bottom right: Normal colonic mucosa.

2H.



2H. Radiographic contrast instilled through the cystoscope demonstrating the two vaginae (V), the cystoscope entering the colon through the septum and contrast filling the colon (C) and distal ileum (I).

2J. **2K.**



Female cloaca case #2 (cont'd.) First operation

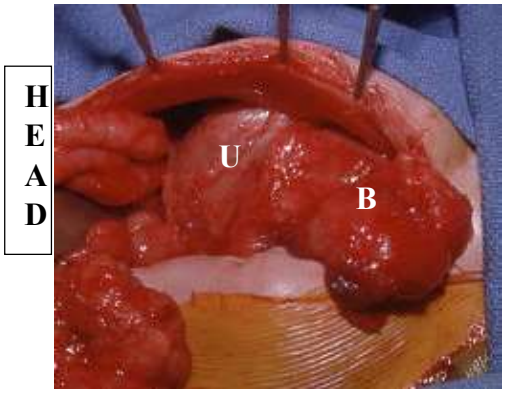
Case #2 cont'd. First operation to correct the problems created by the previous management.

Both photographs were taken from the patient's right side. Her head is to the viewer's left in the photographs.

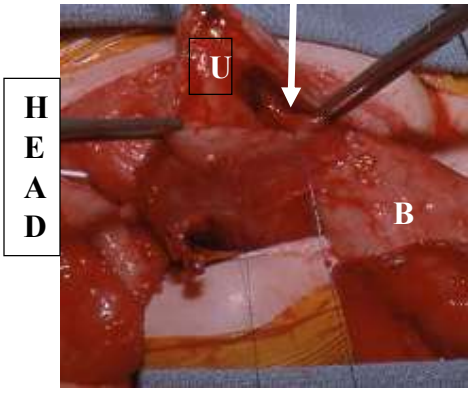
2J. An extremely large bladder (B) is visible. The vesicostomy has been closed (white arrow) after cannulating both ureters (yellow double headed arrow).

2K. After closure of the ileostomy (black arrow) a left colon diverting colostomy has been created. The red rubber catheter is introduced into the proximal limb of the colostomy and a copious amount of mucous has been aspirated from the colon and distal ileum. The recurrent hydrocolpos was due to intestinal mucus filling the vaginae.

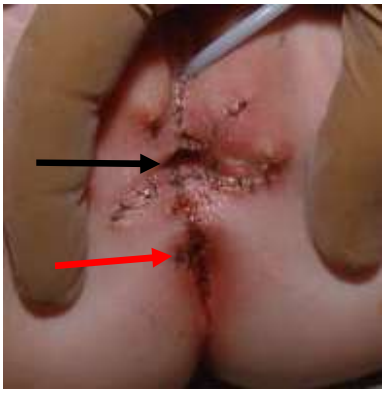
2L.



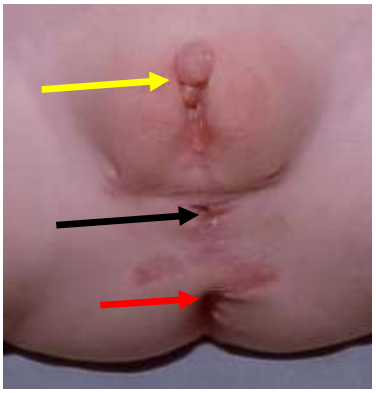
2M.



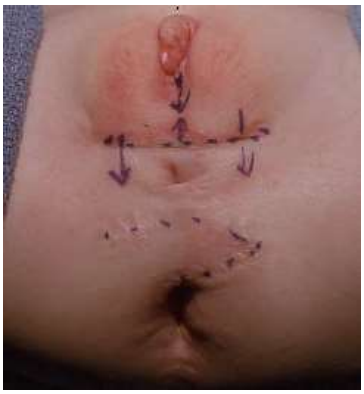
2N.



2O.



2P.



2Q.

Case #2 cont'd. Second and definitive operation.

Both photographs were taken from the patient's right side. Her head is to the viewer's left in the photographs.

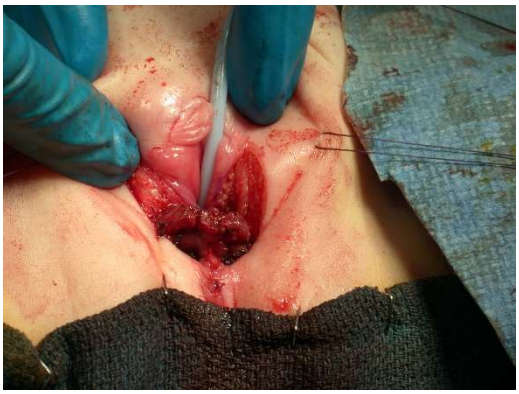
2L. A thick-walled uterus and vagina (U) and an empty bladder (B) are seen.

2M. The uterus and vagina are opened (U). The white arrow points to the septum between the vaginae which is being held up with an Allis clamp. The septum was resected. The bladder (B) has been displaced out of the pelvis.

2N. Appearance of the perineum 1 week post-operatively. The patient is supine with a Foley catheter in the urethra. The black arrow points to the vagina and the red arrow to the neoanus. A Z-plasty in the

perineal skin and subcutaneous tissues was required to move the anus backwards into the center of the sphincter mechanism.
2O. Perineum of the patient 6 months later with excessive skin at the clitoris hiding the urethra (yellow arrow). The black arrow points to the vagina while the red arrow demonstrates the anus.
2P. Skin incisions used in the operation to improve the appearance of the perineum. The urethra was moved posteriorly to the point between the two arrows marked with ink in the anterior midline and the two labio-scrotal folds were displaced posteriorly along the two inked arrows on the sides. The inked dotted lines point to the previous incision scars which were not crossed to avoid damaging the blood supply of the skin flaps.
2Q. Post operative appearance of the perineum after 6 months with the patient supine.

3A.



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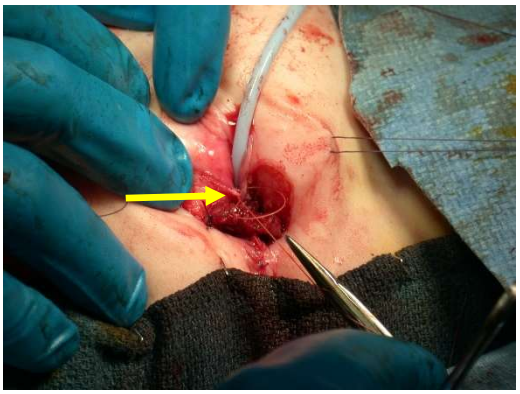

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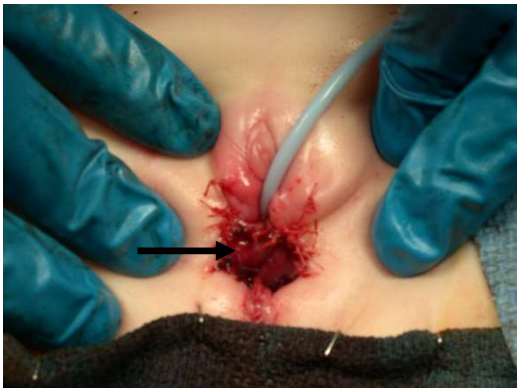

Figure 3. Perineum after Total Urogenital Mobilization.
Perineum of another patient in the supine position.
3A. The entire urogenital sinus is freed and brought down to the perineum.
3B. The posterior wall of the urethra is being constructed (yellow arrow).
3C. The completed procedure with a Foley catheter in the urethra. The vagina is demonstrated by the black arrow.

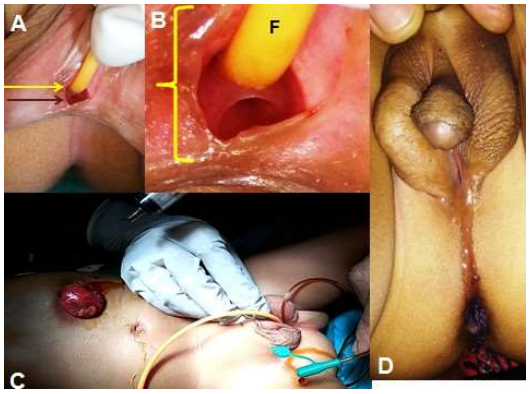

Figure 4. Male persistent cloaca.

Photographs showing single midline perineal opening at the proximal part of the scrotum with small cavity underneath it (**A**); two smaller openings, one anterior (with Foley's catheter *in situ* shown by yellow arrow) and other posteriorly placed (black arrow), are seen in the posterior wall of the cavity (**B**); infant feeding tube placed in posteriorly placed rectal opening and irrigation of the distal colonic loop done to confirm the rectal opening in the common channel (**C**); bilaterally descended testes, penoscrotal transposition, severe chordee, and perineal hypospadias along with neoanus are seen (**D**).

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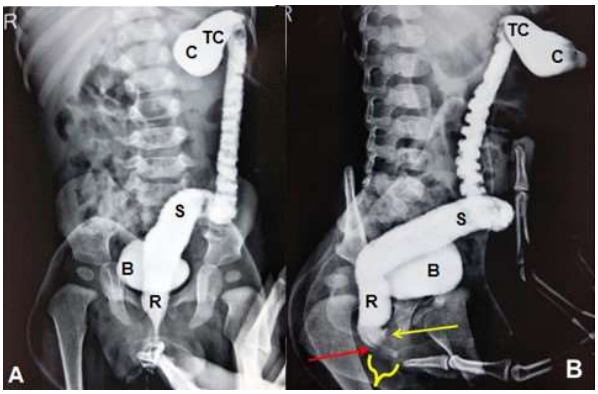

Figure 5. Male persistent cloaca (case cont'd.)

Nonfluoroscopic well-tempered pressure-augmented distal colostogram: Anteroposterior view (**A**) showing rectum tapering before entering into the cavity along with complete bladder filling; lateral view (**B**) showing rectum descending below the "I" line; rectum (red arrow) and urethra (yellow arrow) communicating with a common channel (yellow bracket). TC: Transverse colon, C: Colostomy, S: Sigmoid colon, R: Rectum, B: Bladder

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The primary purpose of antenatal evaluation is to alert the family and physicians that the baby has a major congenital anomaly that will require planned delivery at a center where the baby could be cared for appropriately. Early termination of pregnancy is not indicated.

Clinical presentation in females

Externally, there is an enlarged clitoris with an excess of preputial skin, even though they do not have disorders of sex development. Variations in the numbers and locations of perineal openings depend upon the type of anomaly. A persistent cloaca has a solitary opening close to the base of the clitoris, while the solitary opening of the posterior cloaca is where a normal anus would be located. The two openings in a cloaca variant are for the UGS, where the urethra would normally be located, and an anteriorly displaced anus. The posterior cloaca variant also has two openings, which are the anal opening in its normal position and a posteriorly displaced UGS. Patients with cloacal dysgenesis do not have an external opening [9].

A lower abdominal mass due to hydrocolpos is found in about 30% of patients [13]. Since the common channel appears to be open in them, it is assumed that a one-way valve mechanism results in the vagina or vaginae being distended with fluid.

On occasion, outflow from the bladder is obstructed and this, in turn, leads to obstructive megaureters and hydronephrosis.

Clinical presentation in males

They usually have a penoscrotal hypospadias with a bifid scrotum and possibly a penoscrotal transposition. The urinary and digestive systems open parallel to each other at the cranial end of a short and wide common channel within the pelvis, with a single orifice to the exterior. The lesion has to be differentiated from penoscrotal hypospadias and bifid scrotum in a patient with an anorectal anomaly and a rectobulbar or rectoperineal fistula. The latter lesions do not have a common channel and the fistula meets the urethra at an acute (Y-shaped) or right angle (T-shaped) [6].

Evaluation

The initial diagnosis of a persistent cloaca is relatively straightforward as the patient has a solitary orifice. At this time, it is worthwhile to also evaluate the appearance of the perineum. If it is “characterless” or flat, it indicates a high lesion with a poor or nonexistent anal sphincter mechanism. On the other hand, a well-defined anal dimple and midline raphe suggest the presence of an underlying sphincter mechanism and one should expect a good functional result after reconstruction.

The next priority is to identify any associated problems that may be incompatible with life. After a careful local and general physical examination, a series of studies are indicated. The first test should be an ultrasonographic examination of the pelvis, abdomen and spine to evaluate the pelvic organs, kidneys and spinal cord. Next, antero-posterior and lateral radiographs of the pelvis, abdomen, chest, spine and sacrum are indicated. Further

studies to determine the cardiac and renal status may be required and it is essential to rule out esophageal atresia by passing a nasogastric tube. The above studies should suffice to prepare the patient for the required immediate interventions.

After urgent conditions have been dealt with, definitive studies are obtained in a controlled environment. The anatomy must be defined precisely by using all available modalities, such as cystography and sinography (instilling radiographic contrast into every perineal orifice), under fluoroscopic control and with still radiographs. This is followed by panendoscopy (endoscopic evaluation through every orifice) with photographic documentation to permit reevaluation. Colon contrast studies are used to define the level of anorectal obstruction. A cloacogram in 3-D is particularly valuable. Definitive information is obtained by MRI of the perineal area, spine, sacrum and upper abdominal organs. All other systems must also be evaluated to determine the general status and suitability of the child for major surgery.

Management

Management can be divided into initial urgent care and elective definitive treatment.

Initial care for females. Diversion of the hindgut and drainage of the hydrocolpos are typically required urgently. This is best carried out with a completely diverting high sigmoid or descending colon colostomy with a separate mucus fistula. It is essential to leave an adequate length of distal colon *in situ* for a bowel pull-through and, if necessary, for the construction of the vagina. Until the cloaca is definitively corrected, the hindgut must stay connected to it to function as an outflow channel for vaginal and bladder collections if they do not drain through the UGS. It is not recommended to divert the bowel at the level of the right transverse colon or proximal to it because this results in a large colonic reservoir for urine and mucus, which flows back and forth, causing urinary and vaginal infections. Hyperchloremic metabolic acidosis could develop if enough urine is absorbed from the colon.

The hydrocolpos must be drained immediately, preferably through the UGS, not only to prevent it from compressing the bladder outlet but also to keep it from getting infected. On occasion, a self-retaining catheter may have to be placed to drain the vagina or vaginae to one of the lower abdominal quadrants. If both segments of a duplicated Müllerian system are distended, a window created in the intervening septum will permit a single catheter to drain both sides. Occasionally, a vaginostomy may be necessary to decompress the vagina.

Next, the bladder may require catheterization. Rarely, catheterization is not feasible and a vesicostomy may be needed to alleviate a persistent urinary tract obstruction.

Definitive treatment for females. Treatment of these patients has to be individualized; therefore, no blanket rule is applicable to all of them. However, the following fundamental principles are worth bearing in mind:

1. Definitive treatment should be delayed, within limits, until the evaluation is complete and the patient is thriving.
2. The surgeon(s) must be familiar with all surgical options and all aspects of management.
3. When necessary, other specialists must be consulted prior to the repair and they should be available when the repair is carried out.
4. The team must be willing to modify operative plans as indicated by findings at surgery.
5. The above fact must be conveyed to the family prior to scheduling the child for surgery.
6. Management of the family's expectations is critical and they must be aware that multiple operations and procedures may be required over the patient's lifetime to correct the anomaly and deal with issues that arise in the future.
7. It is best to carry out the entire reconstruction in one operation, as it is difficult to reoperate in a pelvis that has been scarred by previous surgery.
8. It is essential to not perform a procedure that would make future operations more difficult or impossible. It is especially important not to excise any organs or structures without careful thought, as they cannot be replaced.
9. The entire body, from the nipples down to the knees, must be prepared circumferentially for surgery to allow for a change in the approach if so dictated by the findings.
10. The team must be prepared to operate through the posterior or anterior sagittal route, and the abdomen.

Surgical principles in females. In a rare condition such as persistent cloaca, it is essential to be familiar with the experience of two surgeons with the greatest experience in treating these patients and to be prepared for any eventuality. In 1959, Gough brought attention to a series of cloaca patients from the Great Ormond Street Hospital (GOSH) for children in London, UK [16]. However, Hendren was the first to carry out concerted, extensive and successful efforts to correct these problems [17-20]. Subsequently, Peña has collected a huge series of patients and he has made the most important advances in their care [11-13].

The following principles are now clear:

1. The operation should start along the posterior sagittal route.
2. If the hindgut reaches the level of the third sacral vertebra, it can be brought down to the perineum without an abdominal exploration and if the common channel is <3 cm, the entire procedure can be carried out through the posterior sagittal approach [10].
3. In 1997, Peña publicized a new surgical technique, entitled total urogenital mobilization, to deal with any UGS <3cm. in length [21]. It has numerous advantages over separating the vagina from the UGS. Total urogenital mobilization reduces operative time and complications greatly and the cosmetic result is better. The first part of the operation is unchanged, but once the cloaca is identified, the hindgut is

separated from the UGS. The suspensory ligament of the urethra is then divided and the entire UGS complex is mobilized and brought down to the perineum. The urethra is constructed at the perineum. Excess tissue of the UGS is sutured to the sides of the perineum to create the labia.

4. Patients with a common channel >3cm require the addition of an abdominal component to the procedure. Additional urologic procedures should be carried out at the same time. In these patients, special techniques, described below, may be necessary to make the vagina reach the perineum. In some patients, extended transabdominal urogenital mobilization is required [13].
5. Patients with an extra-long common channel of >5cm. may not be reparable even with the added abdominal approach for total urogenital mobilization. In this group, the posterior aspect of the pubis may be carved out to shorten the distance that the urethra and vagina traverse to reach the perineum. In patients with an ultralong channel, after carefully identifying and protecting the ureters with catheters, the bladder and urethra may have to be separated from the genitalia. The UGS forms the neourethra and is used for clean intermittent catheterization (CIC), which is required by the majority of these patients [13]. If a vesicostomy is carried out, it should be closed by 5–6 years of age and ureteric reimplantation can be carried out at the time. It is worthwhile to use anticholinergics to try to avoid bladder augmentation.
6. About half of these patients have Müllerian ductal anomalies that have to be addressed. As mentioned above, if the ducts have not fused, the septum between the two has to be resected partially or completely to permit adequate drainage of the system.

The first choice for repairing the vagina is a vaginal pull-through, which is possible in the majority of patients. If it is not possible, a variety of procedures are available, depending on the findings. In the case of a dilated vagina, a wide, inferiorly based vaginal flap can be created and rotated down on itself. The flap and native vagina are then tubularized to form a thinner vagina that extends to the perineum [11]. When the vagina ends just short of the perineal surface, vascularized, laterally based perineal skin flaps can be laid into the opening to augment the introitus of the vagina. However, one has to be careful to avoid being too aggressive in developing perineal skin flaps, as they might alter the shape of the perineal body and affect the urethral opening. If there are two large hemivaginae and hemiuteri, the septum and one hemiuterus could be excised, both hemivaginae tubularized, and the upper end of the hemivagina on the side from which the uterus was excised turned down to the perineum. Peña termed this procedure the vaginal switch [12]. If none of these options are available, the rectum, colon, or even ileum could be utilized to bridge the gap to the perineum or to create an entire vagina [11,22]. If a very dilated rectum is available, the layers of its mesentery could be separated and the bowel divided

longitudinally to develop a vascularized neovagina [12,23].

Initial care in males. It is best to initially divert the hindgut completely from the cloaca. No other procedure is urgently necessary.

Definitive treatment for males

1. Once the hindgut has been diverted and the patient is thriving, he is studied for associated anomalies.
2. The first step in definitive management is to correct the anorectal anomaly and reconstitute the cloacal channel to form the proximal urethra.
3. The final step in repair is the correction of the hypospadias, penile and scrotal anomalies.
4. If Müllerian remnants are found, they are best excised at the time of the rectal surgery.

Surgical principles in males. Management of the lesion in males is simpler, but there are a few important principles.

1. Regardless of the distance between the rectal pouch and the proposed site of the neoanus, the procedure can be carried out without an abdominal exploration [6,7].
2. The length of the cloacal channel is inconsequential since it does not have to be mobilized to reach the perineum.
3. After it is repaired, the cloacal channel must conform in circumference to the proximal and distal urethra to avoid developing a diverticulum or a stricture.
4. Genital reconstruction should be delayed until the cloaca and the anorectum are well healed and do not require further instrumentation or manipulation.
5. It is probably best to close the colostomy after all the entities are well healed.

Management of the tethered cord

A tethered cord has to be released surgically, but the operation can be carried out after 1 year of age. A renal ultrasound and voiding cystourethrogram (VCUG) may be carried out 3 months later to determine the status of the kidneys and the bladder. Urodynamic studies are only reliable after the child is a year old. To follow-up, yearly ultrasound examinations are adequate unless abnormalities are found on ultrasonography.

Results

The objectives of treatment are not only for the patient to attain socially acceptable fecal and urinary continence but also to permit girls to function sexually and to have the capacity to get pregnant and carry babies to term. In males as well, fecal and urinary control and the potential for sexual activity are the goals of management.

If female patients are treated correctly by surgeons conversant with the various modalities necessary to manage them appropriately, they can have productive lives. Peña [13] has reported excellent results in a series of 490 patients, most of whom were treated through the posterior sagittal approach, with only 38% requiring a laparotomy. Furthermore, in 63% of patients, the genital reconstruction

consisted of a vaginal pullthrough. Fifty-three percent of his patients have spontaneous daily bowel movements, while the rest stay clean with a bowel management regime consisting of daily enemas.

Just over half of Peña's patients were naturally continent of urine after correction, while another quarter remained dry with intermittent catheterization *via* the native urethra. Twenty percent of patients required catheterizable urinary diversion. Almost 80% of patients with a common channel >3cm. required intermittent catheterization, while less than 30% of those with a common channel <3cm. did so. However, it is essential to remember that urinary control or continence does not imply that the urinary tract is normal. Furthermore, even in the absence of structural abnormalities, there is a risk of progressive renal injury in these patients. Therefore, their urinary tracts must be evaluated for life.

Complications are to be expected in any major, intricate reconstructive procedure and additional procedures and operations are inevitable over time. The more common complications pertaining to the urinary and genital structures are persistent UGS, vaginal strictures or atresia, urethrovaginal fistulae and urethral atresia. Complications affecting the hindgut are rectal prolapse, stricture, retraction, dehiscence, and atresia. In addition, on occasion, the rectum may be mislocated [13].

Information on urinary and fecal control and sexual activities in male patients is scant and there is no overview of the condition to date [6,7].

Future considerations

Future studies must standardize techniques and results. Some issues worth considering are discussed below.

First, based upon his extensive experience, Peña has created a valuable algorithm for the management of cloacae based upon the length of the common channel [13]. It would be easier to plan reconstructions, compare results and decide on management strategies if this algorithm were kept in mind.

Common channel <1 cm. A posterior sagittal anorecto-vaginoplasty in which the rectum is separated from the vagina. The UGS is left untouched. The lateral and posterior walls of the vagina are mobilized to suture to the future labia. The minimal female hypospadias is inconsequential since the urethra is clearly visible. These patients are continent of urine and stool.

Common channel 1-3 cm. The majority of patients fall into this category. They are operated through the posterior sagittal approach and do not require additional abdominal exposure. Through this approach, the rectum is separated from the vaginal or Müllerian structures. After total urogenital mobilization, approximately 2 cm. of length is obtained to bring the UGS down to the perineum, where it is split in the sagittal plane and the urethra and vagina are sutured to the neolabia.

Common channel 3-5 cm. In these patients, also the operation starts from the posterior sagittal approach and after the internal anatomy is defined, total urogenital

mobilization is carried out. If enough length is not obtained, the patient is placed supine and the abdomen is entered through an infraumbilical incision. The bladder dome is pulled upward and the lateral attachments of the bladder are divided. The UGS is brought up in the space between the bladder and the pubis and all avascular attachments of the bladder and the urethra are divided. If this maneuver gives the necessary, length the UGS is brought back down and the previously described repair is carried out.

If the UGS can still not be anastomosed properly to the perineum, the posterior half of the pubic cartilage is resected to create a more direct route for the UGS to reach the perineum.

In cases where, in spite of all the above maneuvers, the UGS cannot be anastomosed at the perineum, the urinary tract must be separated from the vagina/vaginae. To do, this the bladder is opened and both ureters are catheterized as they pass out in the wall between the vagina and the bladder. Reconstruction is carried out after the complete separation of the two structures. The choices are a vaginal switch, a vaginal flap, or the use of a bowel segment to bridge the gap or for the entire vagina.

Common channel >5 cm. These patients are best approached from the abdomen since the rectum and the vagina/vaginae open into the trigone of the bladder, where the ureters also open. Upon separation, the bladder neck may be damaged or even destroyed. If the bladder neck is competent, the UGS can be left as the channel for CIC. In some situations, the bladder neck can be closed and a vesicostomy fashioned. Heroic efforts to reconstruct the urethra or to pull through a short urethra are not justified since most, if not all, of these patients will have neurogenic bladders and the extensive efforts to mobilize structures can negatively impact the vaginal or rectal pullthrough.

Occasionally, an extremely high rectum cannot be reached from the posterior sagittal approach. In this situation, the rectum has to be dissected from the abdomen.

Secondly, since the length of the common channel is vital to the ultimate results, it may be worthwhile to measure it with relation to a fixed bony point rather than leave it to endoscopic measurements that are subject to operator error and can vary depending upon the age and size of the child when the measurements are made. In all other forms of anorectal malformations, we use fixed bony points as landmarks. It has been suggested that the pubic symphysis would make the ideal bony landmark for cloacal lesions. The common channel is short if the confluence is below the pubic symphysis, long when the confluence is at or above its upper border and intermediate when the components fuse behind the symphysis [24].

Thirdly, there are no true long-term follow-up studies yet. While the immediate and mid-term results of surgical repair seem to be acceptable in females, we must know what happens to these patients throughout their lives. It is also essential that detailed follow-up information be obtained for males.

Fourth, another modality that may be valuable for the abdominal part of the operation could be laparoscopy instead of

laparotomy. In the past, laparoscopy has only been used for repair of the rectum in patients with a high rectal pouch and a low UGS. Now, it is also being used to mobilize the vagina and there is some resurgence of interest in laparoscopic urogenital separation over total urogenital mobilization. With this modality too, studies are few and there are no long-term results [25].

Finally, one wonders if there is any place for the use of tissue expanders to obtain adequate perineal skin and tissue for the repair. If the vagina cannot be brought to the perineum, we have used broad-based, vascularized perineal skin flaps laid into the introitus to bridge the gap. The use of tissue expanders on either side of the perineal orifice would increase the amount of skin available and the flaps could be placed further into the introitus to complete the vaginal reconstruction. The skin may also be helpful in the repair of the urethra. Liu et al. used tissue expansion in persistent, cloaca but they placed the expanders within the UGS [26]. While they succeeded in widening the UGS to create a separate urethra and vagina, their technique is problematic because all their patients required a vesicostomy and hospitalization for the 3–4 weeks it took for sufficient tissue expansion. Thus, their technique is cumbersome, results in prolonged hospitalization, increases hospital costs tremendously, is uncomfortable for patients and is inconvenient for families. Tissue expanders placed on the perineum on either side may obviate these problems.

Conclusion

Persistent cloaca is a complicated problem that must be approached after careful evaluation at a center with experience in management. With proper care, excellent results are achievable. Therefore, it seems reasonable that, if not all patients with a persistent cloaca, at least long-channel patients should be transferred as soon as possible to a center with the expertise to treat them. Improper intervention prior to referral makes subsequent correction difficult and results in poor outcomes.

Statements and Declarations

Competing interests

None of the authors have any competing interests.

Conflict of interest

None of the authors have any conflict of interest.

Author contributions

All four authors reviewed the literature and contributed to the manuscript.

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