

American Pediatric Surgical Association

## Prenatal Counseling Series

### Persistent Cloaca



**APSA**  
American Pediatric  
Surgical Association  
*Saving Lifetimes™*

from the  
**Fetal Diagnosis and Treatment Committee**

of the  
American Pediatric Surgical Association

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## **Persistent Cloaca**

- Persistence of the early embryological stage of a common opening for rectum, vagina and urinary tract
- Rare anomaly: 1:50,000 female births
- Differential diagnosis: Fetal Abdominal Cysts (Please refer to Fetal Abdominal Cysts Brochure)

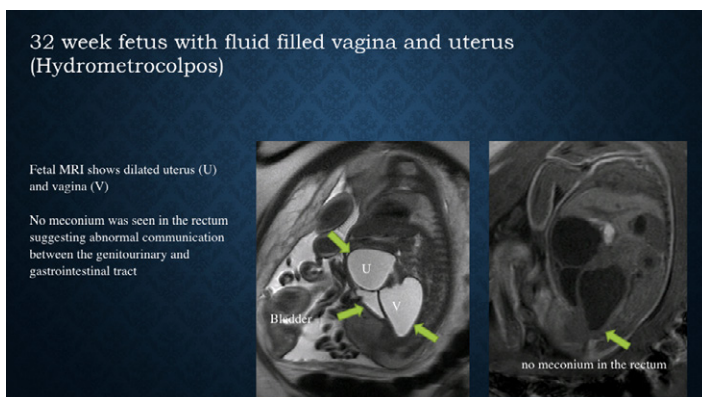
### **Embryological facts of cloaca:**

- Development starts at 3 weeks
- U-shaped endodermal cavity where hind gut, allantois, tail gut and later on the mesonephric ducts open
- Carried anteriorly upon folding of the embryo
- Cloaca is at first closed by the cloacal membrane which reaches up to the umbilical ring
- At 6th week: Urorectal septum descends down to fuse with the cloacal membrane and the lateral folds -> Urogenital cavity anteriorly and anorectum posteriorly
- Rapid growth of the genital tubercle displaces the cloacal membrane posteriorly
- At 7th week: cloacal membrane breaks down creating 2 openings
- Muscles surrounding the rectum develop at the same time (6th and 7th weeks)
- No sexual differentiation until 9th weeks' GA

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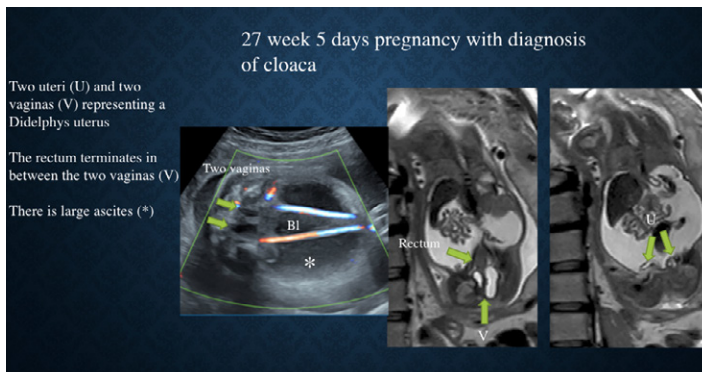
### Prenatal Considerations

- Most common prenatal presentation:  
**Fetal abdominal cystic mass**
- Cystic mass may be the hydrocolpos (50% of cases) or associated bladder outlet obstruction (BOO)



Images courtesy of Mariana Meyers, MD  
Colorado Fetal Care Center - Children's Hospital Colorado

- Duplicated vagina and uterus didelphys commonly seen



- Prenatal US findings are 2ry to underlying anomaly
  - Septate or bilateral cystic pelvic mass is the most common Mullerian tube anomaly associated with persistent cloaca
  - Hydrocolpos posterior to the bladder
  - Vesicomegaly and hydroureteronephrosis
- Associated with increased incidence of other anomalies: ascites, urogenital, cardiac, GI, and skeletal

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**Accurate Diagnosis:** Targeted US, fetal echocardiography, fetal MRI and amniocentesis

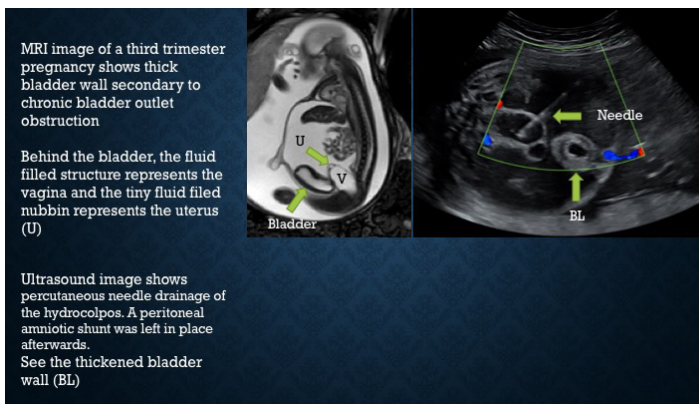
- It is important to make the distinction between hydrocolpos and distended urinary bladder
- Urinary ascites may be seen if there is drainage via the fallopian tubes
- MRI may be extremely helpful to delineate the anatomy and evaluate for associated anomalies

## Fetal Intervention

### Indications for prenatal decompression

- o Significant urinary obstruction leading to pulmonary hypoplasia - (This is a rare event)
- o Drainage of the hydrocolpos or peritoneal cavity may relieve obstruction

Very few reports of antenatal fetal intervention for persistent cloaca with hydrocolpos exist (Shimada et al., 2001)



## Postnatal Management

- Newborn screening for patients with anorectal malformation

### First 24 hours of life – rule out important associated malformations:

#### Cardiac anomalies (echocardiogram)

- 30% of patients have associated cardiac anomalies, 10% of them with hemodynamic repercussion. The most common are: patent ductus arteriosus, atrial septal defect and tetralogy of Fallot.

#### Gastrointestinal anomalies (nasogastric tube and babygram)

- 8% of patients have esophageal atresia, 3% have duodenal atresia.

#### Urological anomalies (kidney ultrasound)

- 50% of patients have an associated urological condition. The most common are: hydronephrosis, vesicoureteral reflux, absent kidney and megareter.

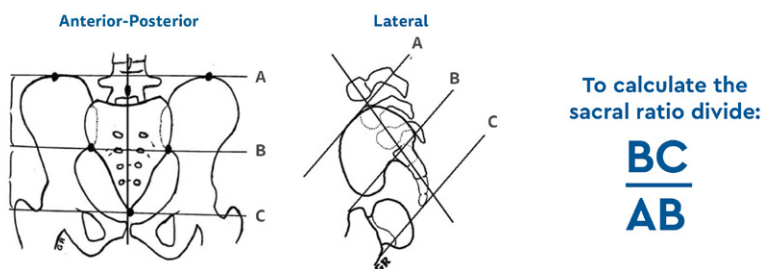
#### Spinal anomalies (sacral radiograph AP and lateral, spinal ultrasound)

- 25% of patients have tethered cord that can be detected with a spinal ultrasound. The sacral radiographs will rule out a hemi-sacrum (indication of a presacral mass) and would allow to calculate the sacral ratio (help to determine the prognosis for future bowel control).

#### Hydrocolpos in patients with cloaca (pelvic ultrasound)

- 30% of cloaca patients have a very distended vagina that should be permanently drained at the time of colostomy opening.

Source: Colorado Fetal Care Center and The International Center for Colorectal and Urological Care – Children’s Hospital Colorado



A value that is equal or more than 0.7 represents good prognosis for bowel control.

Values between 0.41 – 0.69 are considered undetermined.

A value that is equal or less than 0.4 represents poor prognosis for bowel control (fecal incontinence).

Source: Colorado Fetal Care Center and The International Center for Colorectal and Urological Care – Children’s Hospital Colorado

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- After ruling out important associated anomalies during the first 24 hours of life, the patient should be taken to the operating room for a diverting descending colostomy and hydrocolpos drainage, if indicated.
- The location of the proximal stoma should be in the center of a triangle formed by the umbilicus, the last rib and the top of the iliac crest (figure 1). The mucous fistula should be tapered to avoid prolapse and should be placed medially, and with enough distance from the proximal stoma, to allow for the stoma bag to only cover the proximal stoma.
- An important and time-consuming portion of the operation is the cleaning of the distal bowel. This can be performed by inserting a catheter into the center of a purse string suture (figure 2) and irrigate it with saline solution until all the meconium is removed, the bowel is collapsed and well perfused.
- When the hydrocolpos is small it can be drained through the same oblique incision done for the colostomy (figure 3). When there is a large bilateral hydrocolpos a midline infra-umbilical incision is preferred (figure 4).
- During hydrocolpos drainage the surgeon should palpate the cervix (cervices) to assure that the opening for the tube insertion is done in the vagina. A pigtail catheter or a Foley catheter can be used, a purse string suture is recommended and, when the anatomy allows for it, the vagina should be tacked to the abdominal wall. The tube should remain in place until the main reconstruction happens.

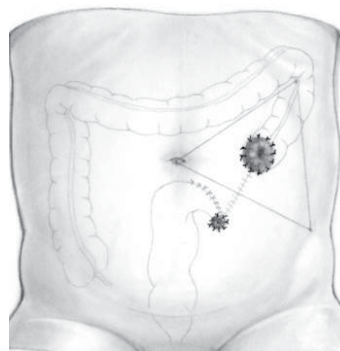


Figure 1: Descending colostomy and mucous fistula

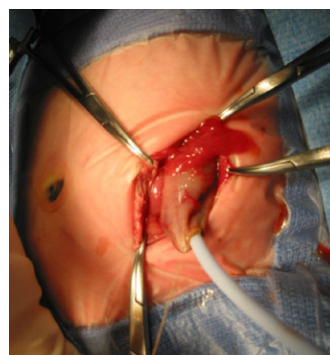


Figure 2: Cleaning of the distal bowel during colostomy opening



Figure 3: Oblique incision for colostomy opening and hydrocolpos drainage

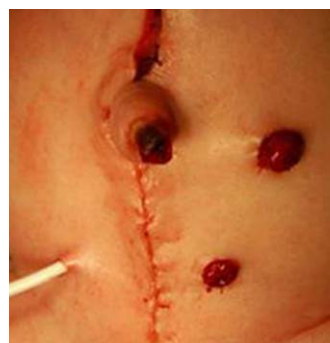
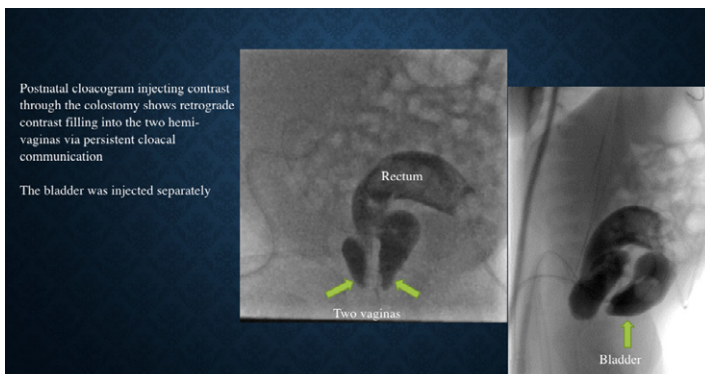


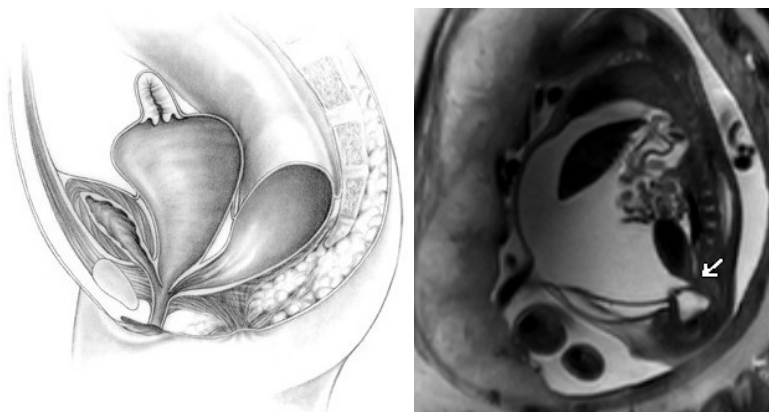
Figure 4: Midline incision for colostomy opening and hydrocolpos drainage

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- High pressure distal colostogram is indicated, once the baby is growing well, with simultaneous injection through the vaginostomy tube, if present, and/or common channel; for surgical planning.
- Panendoscopy to accurately measure the common channel



- Multidisciplinary approach
- Length of common channel varies from 1-12cm (Pena et al., 2004 - Rich et al., 1988)



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