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Radiologic diagnosis of a newborn with cloaca

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ABSTRACT

When a female is born and has only a single perineal orifice on the newborn clinical examination, a diagnosis of cloaca type of anorectal malformation is made. Along with associated malformations which may initiate the ordering of radiologic imaging, there are a finite number of radiologic tests that are performed to help in the immediate management of the patient with cloaca. The following discussion will outline the most important radiologic tests and demonstrate examples of images from newborn females with cloaca.

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When a newborn female is examined for the first time and is found to have just a single perineal orifice, the clinical diagnosis of anorectal malformation (ARM) is made, specifically, cloaca. At our hospital, all females with a clinical diagnosis of cloaca undergo a finite number of radiologic tests which are performed to help in the immediate management of the patient, and to provide prognostic information that can help in the initial discussions with the parents, concerning the infants' malformation and what the parents can expect in terms of the possibility of fecal, urinary continence, and sexual function. The infant with cloaca will invariably be booked for an operation, usually a colostomy with separated stomas, to be done within the first 24–48 h of life. That time, represents a window of opportunity to perform a series of studies aimed to rule out associated conditions that represent an immediate risk for the baby's life. These include cardiac defects, esophageal atresia, duodenal atresia, and obstructive uropathy. Decisions related with the possibility of performing a colostomy or performing a primary repair of an anorectal malformation, should not be taken during the first 24 h of life. In other words, the baby can only be taken to the operating room after all the major associated anomalies have been ruled out.

In the specific cases of cloacas, the surgeon should not take a baby girl to the operating room without having ruled out the presence of a dilated vagina (hydrocolpos) with or without a septum. Why is this important? Aside from diagnosing hydrocolpos and to plan for its operative management (drainage) at the time of colostomy, its presence is almost always associated with bilateral obstructive uropathy at the level of the distal ureters, causing megaureters, and hydronephrosis, which may provoke renal damage and even early acute renal insufficiency. Undiagnosed, the hydrocolpos may become infected (pyocolpos), and provoke sepsis with its clinical ramifications. We have seen cases of vaginal perforation, with dissemination of pus in the peritoneal cavity. Hydrocolpos cannot be diagnosed based on clinical exam only, since obstruction of the colon will invariably cause a distended abdomen. Therefore, each infant born with cloaca requires an immediate postnatal pelvic and renal ultrasound (Figure 1). Hydrocolpos can involve a single vagina or 2 of them (hemi-vaginas) (Figure 2), and the degree of hydrocolpos can be variable from marked hydrocolpos as seen in Figure 1 to moderate or mild degrees of hydrocolpos (Figure 3). During the ultrasound, which can be performed at the bedside in the neonatal intensive care unit, it is possible to attempt decompression of a dilated vagina by placing a small feeding tube, retrograde through the single perineal orifice, fortuitously catheterizing it, and draining it. Since it is the dilated vagina which obstructs the distal ureters at the level of the bladder trigone, simply decompressing the vagina(s) can temporarily relieve the ureteral obstruction (Figure 4) until definitive vaginal drainage can be performed, usually at the time of colostomy. Other more invasive maneuvers such as percutaneous nephrostomies, diverting ureterostomies or vesicostomies, may not be necessary after the hydrocolpos has been drained. Those operations may be more harmful than helpful and can lead to unnecessary complications. If it is not possible to catheterize the vagina via the perineal orifice (also known as the common channel) one can drain the vagina percutaneously by ultrasound guidance or preferently by surgical drainage and catheter placement at the time of diverting colostomy.

Associated congenital renal abnormalities are common in all patients with anorectal malformations, especially cloaca, and are

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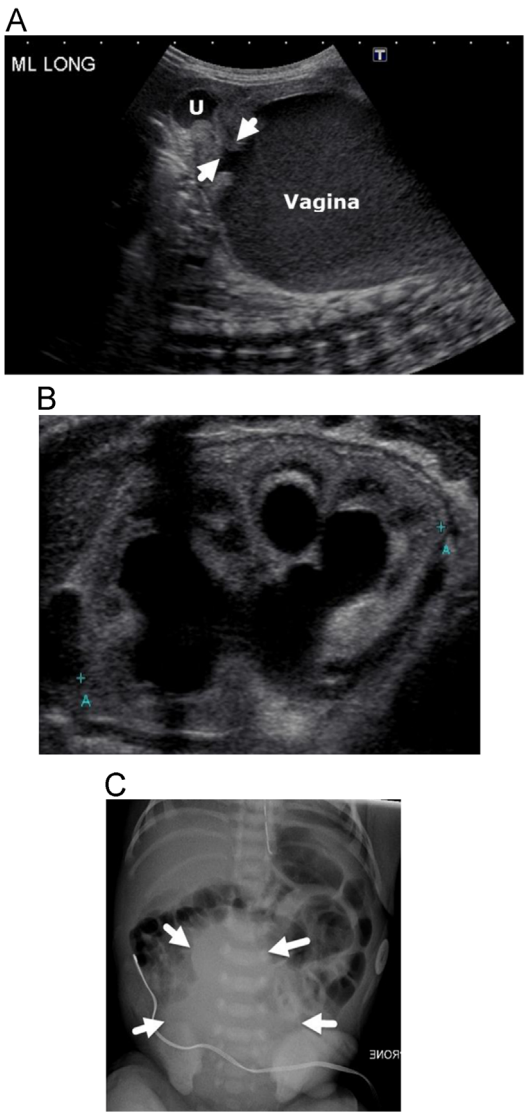

Fig. 1. Longitudinal ultrasound image of the pelvis (A) in a newborn female born with a single perineal orifice (cloaca) and abdominal distention shows a markedly dilated vagina (hydrocolpos) with echogenic material within it (likely a combination of meconium and urine), a patulous cervix (white arrows), and a small amount of fluid refluxing into the uterus (U). Longitudinal ultrasound image of the right kidney in the same patient. (B) Moderate to severe hydronephrosis; bilateral hydronephrosis and hydroureter (not shown) results from compression of the distal ureters by the dilated vagina in newborns with cloaca and hydrocolpos. Frequently, the urinary bladder is difficult to see on ultrasound due to the marked distal ureteral compression and one might misinterpret the dilated vagina as a dilated urinary bladder. (C) Abdominal radiograph in this same patient showing mass effect (white arrows) from the dilated vagina, displacing bowel loops peripherally.

frequently noted on renal ultrasound (Figure 5) including unilateral renal agenesis, renal ectopia (crossed fused ectopia or horseshoe kidney). and multicystic dysplastic kidney to name a few. Occasionally when a patient with cloaca is found to have hydronephrosis and hydroureter either with or without hydrocolpos,

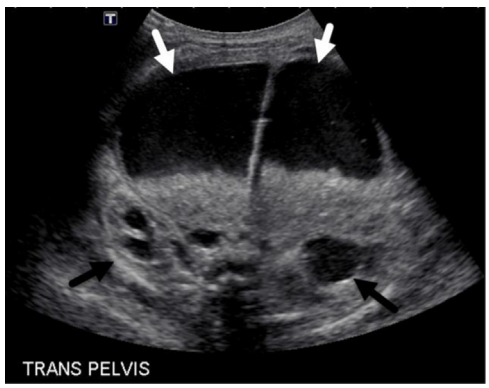

Fig. 2. Transverse ultrasound image of a newborn female diagnosed clinically with cloaca shows marked hydrocolpos of hemi-vaginas (white arrows) filled with a mixture of urine and meconium; the hydronephrotic kidneys due to distal ureteral obstruction by the enlarged hemi-vaginas are also seen posteriorly (black arrows).

one can pass a catheter through the single perineal orifice and with some luck, into the bladder and perform a VCUG which might show vesicoureteral reflux (Figure 6).

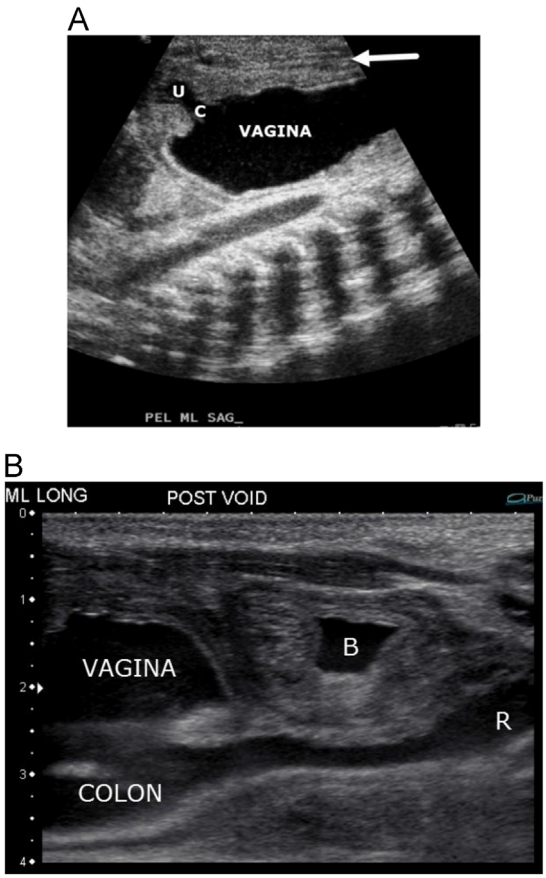

Fig. 3. Longitudinal ultrasound image of the pelvis in another newborn female with clinical diagnosis of cloaca shows only moderate hydrocolpos, an open cervix (C), and fluid filled uterus (U); the urinary bladder is empty (white arrow). (B) With less dilation of the vagina, the urinary bladder (B) may fill as it did in this same patient later in the study; the ultrasound also showed fluid in the colon and rectum (R).

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