

Abdominal Distention and Acute Urinary Retention secondary to Congenital Distal Vaginal Obstruction in a Newborn Female

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Abstract

Congenital distal vaginal obstruction is usually asymptomatic in a newborn female. On rare occasions, it may present as an acute emergency with life threatening complications.

This paper is reporting the rare condition of two newborn females presenting urgently with abdominal distension and acute urinary retention as a result of congenital distal vaginal Obstruction. The case history and urgent management shall be presented and both conditions shall be discussed.

Keywords: Imperforate Hymen; Distal Vaginal Atresia; Hydrometrocolpos; Acute Urinary Retention; Mullerian Duct Anomalies.

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Introduction

In a newborn female, the impact of common congenital anomalies of the genital tract is highly variable; some are asymptomatic chance findings during routine peri-natal examination requiring no immediate intervention, others may present early and urgently with life threatening complications requiring immediate management ⁽¹⁾.

Many congenital anomalies manifest as vaginal obstruction; Imperforate Hymen, Transverse and Longitudinal Vaginal Septum, Distal Vaginal Atresia and Agenesis of the vagina are the most frequently encountered anomalies ⁽²⁾.

Distal vaginal obstruction may manifest at variable periods during early childhood, adolescence and puberty; only rarely it may present in the neonatal period with life threatening complications ⁽³⁾. These congenital anomalies may occur as an isolated lesions or part of more complex malformations that need further management ⁽³⁾.

In the present paper, we are reporting the rare case of two newborn females with distal vaginal obstruction due to different pathologies, presenting the same clinical picture of abdominal distension and acute urinary retention. The case history and urgent management shall be presented, followed by a discussion of both conditions.

Case History

The first case was a 12-day-old newborn female, presented to the emergency department on the 6th of Feb. 2007 with history of progressive abdominal distention since birth. One day before presentation, she started vomiting, reluctance for oral feeding and inability to void. Bowel function was normal.

On examination, the abdomen was distended with a non-tender huge mass, cystic in consistency, arising out of the pelvis and extending far above the level of the umbilicus. Bowel sounds were normal.

On examination of the perineum, the external genitalia looks normal with a normal urethral opening but a grayish-white bulging hymen can be seen completely occluding the vagina. The anal verge

looks normal and the rest of infant examination was negative (Photo.1).

Photo.1: (Case I) Distended Abdomen and Bulging Imperforate Hymen



Urgent Ultra Sound (US) examination reported a large cystic mass filled with thick fluid and seen unrelated to the bowel. The cervical and vaginal canals were dilated and two normal looking small ovaries could be seen. A moderately distended bladder could be identified anteriorly.

A size 6⁰F Foley catheter was introduced and recovered 100 cc of clear urine. Urgently; the obstructed utero-vaginal canal was relieved by a longitudinal hymenotomy recovering more than 350 cc of thick-whitish odorless fluid (Photo.2). The hymenotomy margins were sutured to the vaginal mucosa at the vaginal ring to prevent early closure. The second day, the patient was discharged for follow-up. Two weeks later, the infant looked reliably well and the abdominal mass disappeared. Voiding and feeding were normal.

Photo.2: (Case I) Suction of the Thick Fluid after Hymenotomy



The second case was a 7-day-old newborn female referred to the emergency department because of abdominal distention, reluctance for feeding, frequent vomiting, dehydration and acute urinary retention. The abdomen was moderately distended; a cystic mass was palpable out of the pelvis, reaching above the umbilicus. On perineal examination, the external genitalia were under developed with a small vestibule and a normal looking urethral opening but no vaginal opening could be seen (Photo.3). The rest of the examination of the infant was normal.

Photo.3: (Case II) Catheter in the Urethra, no Vaginal Opening can be Identified



The US findings were identical to that in the first case. A size 6⁰F Foley catheter recovered 120 cc of clear urine. Urgent surgical decompression of the mass was decided. Because of the absence of a vaginal opening and the demonstration of a dilated utero-vaginal canal by US, a preliminary diagnosis of Distal Vaginal Atresia was made. The distended uterus was approached through a small infra-umbilical transverse incision. Drainage of more than 250 cc of thick whitish odorless fluid- through a small opening in the fundus of the uterus- was performed and a size 12⁰F Foley catheter was left in place for continuous drainage (Photo.4).

Photo.4: (Case II) Catheter Drainage of the Distended Uterus and Vagina



Two days later, the infant's general condition remarkably improved, oral feeding resumed and the patient discharged for follow-up. Two weeks later, the infant was seen with no abdominal distention, normal voiding and feeding with minimal drainage from the catheter. Six weeks later, the catheter had been removed. Because of the present situation in Iraq, the patient was lost for follow up.

Discussion

The conditions presented demonstrate a similar urgent clinical picture- but different pathologies- for congenital lower vaginal obstruction in newborn females. Although the US findings were identical in both cases, careful local clinical examination of the perineum was helpful in identifying the possible cause; an Imperforate Hymen in the first case and a Distal Vaginal Atresia in the second.

In both female infants, the mass represented a distended uterus filled with secretions; a Hydrometrocolpos ⁽⁴⁾. The secretions are believed to result from the maternal estrogen effects on the vaginal and/or cervical glands. The fluid collected can be watery, milky, or mucoid in nature; giving the hymen a characteristic whitish-gray dome appearance, in contrast with the blue dome appearance of an Imperforate Hymen in hematocolpos ⁽⁴⁾.

The pressure of accumulated fluid in the vaginal canal above the obstruction results in opening of the cervical canal with progressive accumulation of fluid inside the uterus and the consequent appearance of a palpable mass and abdominal distension ⁽⁴⁾. The pressure exerted by the distended vagina and uterus can cause urinary retention and even acute renal failure ⁽⁵⁾.

Imperforate Hymen and Vaginal Atresia are considered as varieties of Mullerian Duct Anomalies, which also include a wide range of other malformations ⁽⁶⁾. The general incidence of Mullerian Duct anomalies is between 0.1% - 0.3% ⁽⁶⁾.

Imperforate Hymen is uncommon (incidence of 0.1%) and in most of the cases its diagnosis is mainly clinical ⁽⁷⁾. It is usually sporadic, but rare familial cases do occur and both the recessive and the

dominant (sex-linked and autosomal) modes of transmission had been reported⁽⁸⁾. However, no genetic markers or mutations have been proven as etiological factors⁽⁸⁾⁽⁹⁾.

It usually presents as an isolated abnormality; the internal and external genital organs are normally developed, however, associated obstructive Mullerian anomalies like vaginal septi and cervical atresia, although rare, has to be excluded⁽¹⁰⁾⁽¹¹⁾.

In normal development, the Mullerian ducts elongate and reach the urogenital sinus by 9 weeks of gestation⁽¹²⁾. The uterovaginal canal forms and inserts in to the urogenital sinus at Muller's tubercle, the ducts fuse and internal canalization and septum resorption occur by 20 week's gestation⁽¹²⁾. By the time the child is born, the typical hymen is annular or circumferential; completely surrounds the vaginal orifice and has a central hole⁽¹³⁾. If canalization of the urogenital sinus with the Mullerian system is incomplete, the resulting hymen can be imperforate or microperforate⁽¹³⁾.

Although spontaneous opening of an Imperforate Hymen had been reported⁽¹⁴⁾, it usually does not; yet it remains asymptomatic and undetected until puberty where it may present as an obstructive anomaly causing lower abdominal pain, back pain and even a lower abdominal mass (Hematometrocolpos)⁽¹⁵⁾⁽¹⁶⁾⁽¹⁷⁾. Serious complications, in the form of secondary infection and pelvic abscess collection, had also been reported⁽¹⁸⁾.

Rarely, symptoms manifest antenatally as well as in the neonatal period⁽¹⁹⁾. The accumulated Hydrometrocolpos may exert pressure symptoms causing urinary retention and even acute renal failure⁽²⁰⁾⁽²¹⁾.

In spite of the fact that the diagnosis of this emergency is possible on clinical grounds; US is necessary. It is the imaging modality of choice for evaluating the pediatric female pelvis⁽²²⁾. The proper knowledge of the normal US appearance of the pelvic organs is the basis for the recognition of these pathological findings; US will confirm the diagnosis, demonstrate internal anatomy and reveal whether there is any extrinsic urethral compression and hydronephrosis⁽²²⁾. Computerized Tomography scanning (CT Scan) and Magnetic Resonance Imaging (MRI) should be reserved for more complex cases⁽²²⁾⁽²³⁾. Combined modalities may be necessary to exclude the rare presence of concurrent pathologies, especially when incision of an Imperforate Hymen fails to drain a Hydrometrocolpos⁽²³⁾⁽²⁴⁾.

The urgent management of this condition is hymenotomy; it should be an adequate treatment for isolated lesions⁽²⁵⁾. Simple puncture must be avoided because the viscous fluid may not drain adequately

through a small hole and there will be an increased risk of ascending infection⁽²⁶⁾.

The second case demonstrates a different lower vaginal obstructive anomaly. Distal Vaginal Atresia may also present with a voluminous Hydrometrocolpos in a newborn female⁽²⁷⁾.

As in Imperforate Hymen, Distal Vaginal Atresia is attributed to failure of caudal development of the Mullerian ducts⁽²⁸⁾.

In the second case, the presence of underdeveloped external genitalia with the absence of vaginal opening- and the demonstration of a dilated uterovaginal canal by US- supported the clinical impression of isolated Distal Vaginal Atresia complicated by a Hydrometrocolpos. The rest of the negative general examination was helpful in the preliminary exclusion of two conditions; the "Bardet-Biedl" Syndrome (an autosomal recessive disorder where vaginal atresia is associated with postaxial polydactyly) and the "Mckusick-Kauffman" Syndrome (an autosomal recessive disorder where vaginal atresia and postaxial polydactyly are associated with imperforate anus and congenital heart defects)⁽²⁸⁾⁽²⁹⁾⁽³⁰⁾. More complex malformations may also present with an acute Hydrometrocolpos and demonstrate a challenging diagnostic dilemma⁽³¹⁾.

In this context it is also important to differentiate between vaginal atresia and vaginal agenesis; vaginal agenesis is usually associated with cervical and/or uterine agenesis and it is part of the "Mayer-Rokitansky-Kuster-Hausr" (MRKH) Syndrome⁽³²⁾. This syndrome is associated with renal and skeletal anomalies in about 34% and 12-50% of cases respectively⁽³²⁾.

A Transverse Vaginal Septum presenting with Hydrometrocolpos may occur coincidentally with an Imperforate Hymen⁽³³⁾. Associated coarctation of aorta, atrial septal defects and malformations of the lumbar spine may be found⁽³³⁾⁽³⁴⁾.

When suspected; the corner stone in the definition of more complex anomalies of the female genital tract is MRI, and especially; with the advent of the pelvic coil, MRI has proved to be the most accurate⁽³⁴⁾⁽³⁵⁾. It has become a necessary and an important tool when US reveals associated complex malformations or is inconclusive⁽³⁶⁾.

The urgent management of Isolated Distal Vaginal Atresia presenting with Hydrometrocolpos and pressure symptoms is a temporary drainage procedure⁽³⁷⁾. It allows local findings to subside and usually results in dramatic improvement in the condition of the patient⁽³⁷⁾.

The definitive surgery for distal vaginal atresia is either a "Cut-Back" vaginoplasty (for a short distal atretic segment), or some form of vaginal reconstruction, augmentation or replacement (for a

longer atretic segment) ⁽³⁸⁾. Surgery may be performed in the neonatal period, taking advantages of the effect of the remaining neonatal estrogen on the vaginal mucosa, but it is known to be associated with a higher rate of vaginal stenosis. Vaginaoplasty can be delayed until puberty, where patient can be usually motivated to help with dilatation to prevent stenosis ⁽³⁸⁾.

Conclusions

A Hydrometrocolpos is the result of vaginal obstruction and can become an emergency in the newborn period. Proper local examination is helpful in preliminary diagnosis of the cause. General physical examination is fundamental component of the workup; it may help in preliminary exclusion of associated congenital anomalies. US examination is crucial for diagnosis. The need for more sophisticated tools is primarily dependent on the result of clinical examination combined with the US results. Urgent management in these cases includes a form of local drainage of the Hydrometrocolpos. Local drainage is adequate for cases due to Imperforate Hymen. Distal Vaginal Atresia needs a definitive immediate or delayed vaginoplasty.

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