

CASE REPORT

Neonatal Hydrometrocolpos Secondary to Imperforate Hymen in a Two-Month Old Girl

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ABSTRACT

Congenital hydrometrocolpos is a rare disorder in infants characterized by an expansion of vaginal and uterine cavities with fluid accumulation. This case report describes a rare condition, hydrometrocolpos, caused by an imperforate hymen in a two-month-old girl. The patient presented with excessive crying and abdominal distension. The imaging studies demonstrated a large fluid-filled cystic structure conforming to the shape of the uterus involving the upper part of vagina and the whole of the uterine cavity to the level of umbilicus. Careful clinical examination of the perineum and inspection of the introitus revealed a bulging hymenal membrane; thus, the diagnosis was established, and surgical intervention was planned promptly. A hymenectomy was performed to open the hymen and relieve the obstruction. The patient remained hemodynamically stable and was discharged after three days. Follow-up ultrasound showed normal morphology of the uterus.

Keywords: Congenital hydrometrocolpos, imperforate hymen, hymenectomy, cystic structure, hydronephrosis.

INTRODUCTION

Congenital hydrometrocolpos in infants is a rare disorder characterized by the expansion of vaginal and uterine cavities with fluid accumulation. It is often resulting from distal vaginal obstruction owing to increased production of secretory glands of the reproductive tract [1]. Hydrometrocolpos is exceptionally uncommon with an incidence rate of 0.006% [2]. This disorder is mainly caused by underlying congenital pathologies such as imperforate hymen, lower vaginal atresia, uterovaginal anomalies, and cervical stenosis [3]. The most common cause is imperforate hymen, in which the hymen completely obstructs the vaginal opening and it happens in 0.05-0.1% of the infant female population [4].

Congenital hydrometrocolpos can present as an intraabdominal cystic mass with or without associated dilatation of the pelvicalyceal system on ultrasound examination. Antenatal or early postnatal diagnosis is essential, as it allows prompt management of hydrometrocolpos and reduces the risk of complications like dilatation of the pelvicalyceal system and bowel compression. We present multi-planar computed tomography images illustrating hydrometrocolpos and associated right-sided hydronephrosis in a two-month-old girl.

CASE PRESENTATION

This is a case of a month girl, who presented to the pediatric emergency department with complaints of excessive crying and abdominal distention. An abdominal ultrasound was advised to rule out the underlying cause of abdominal distention. There was no

history of any anomalies in prenatal sonographic examinations. Ultrasound examination revealed a large, well-defined anechoic cyst in the pelvis extending to the umbilical region, and an initial impression was made of a fetal enteric duplication cyst or mesenteric cyst. This patient was further evaluated with the contrast-enhanced CT scan abdomen and pelvis, which showed a large fluid-filled structure conforming to the shape of the uterus involving the upper two third of vagina and the whole of the uterine cavity to the level of umbilicus. The distal vaginal canal was not appreciated. It measured 6.6 x 5.1 x 10.6 cm with a volume of 178 ml and its mean attenuation value is 178 ml. It was anterior to the compressed rectum and was in close approximation to a minimally filled urinary bladder (**Fig. 1**). Mild right-sided hydronephrosis due to compression by a large distended uterine cavity (**Fig. 2**). CECT abdomen and pelvis suggested congenital hydrometrocolpos secondary to distal vaginal obstruction, further clinical examination and MRI may consider for better evaluation of lower one-third of the vagina. Clinical examination of the perineum and introitus, revealed a bulging

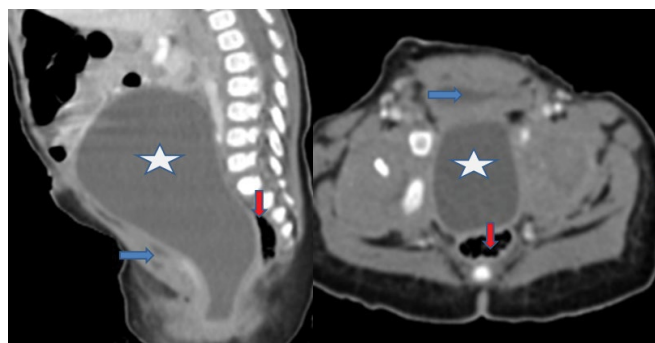


Fig. (1): Sagittal and axial CT images of abdomen and pelvis show a large fluid-filled structure (star) conforming to the shape of a uterine cavity between the minimally filled urinary bladder (blue arrow) and compressed rectum (red arrow).

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Received: April 06, 2025; Revised: June 30, 2025; Accepted: July 02, 2025
DOI: <https://doi.org/10.37184/nrjp.3007-5181.1.30>

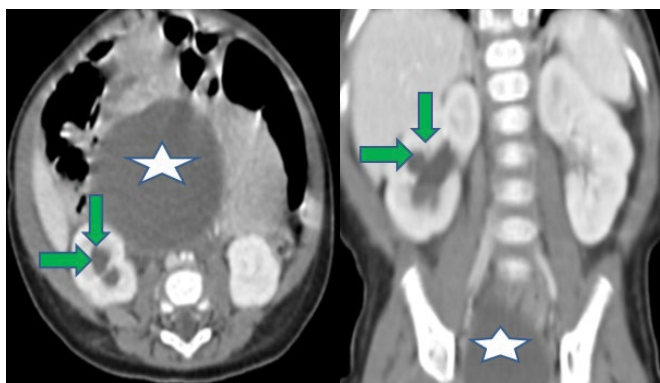


Fig. (2): Axial and coronal CT images of abdomen show mild right-sided hydronephrosis (green arrows) due to compression by a large distended uterine cavity.

hymenal membrane, completely covering the vaginal opening, thus the diagnosis was evident, and surgical intervention was undertaken promptly. A hymenectomy was performed to open the hymen, which relieved the obstruction and about 200 ml of milky fluid was drained. The patient remained hemodynamically stable and was discharged after three days and follow-up in outdoor patient department was advised. Follow-up ultrasound showed normal morphology of the uterus.

DISCUSSION

Congenital hydrometrocolpos is a rare disorder but an important diagnosis in infants presenting with abdominal distension. The fluid accumulation is attributed to the increased production of mucous glands of the cervix and endometrium which gradually causes the expansion of vaginal and uterine cavities due to lower vaginal obstruction, resulting in a cystic intraabdominal mass [1].

Many congenital urogenital malformations lead to this condition, most commonly caused by imperforate hymen. Vaginal anomalies are classified into five types, such as imperforate hymen (type 1), vaginal septum (type 2), lower vaginal atresia (type 3), vaginal atresia with urogenital sinus (type 4), and vaginal atresia with cloacal anomaly (type 5) [3]. Hydrometrocolpos typically occurs sporadically, but it can also be seen associated with other various conditions or syndromes [5]. This may also occur as a complication of steroid treatment for congenital adrenal hyperplasia [6]. It may present as an isolated finding or may also link to less common, genetic syndromes such as Bardet-Biedl, Pallister Hall, and McKusick-Kaufman syndrome [5].

Patients present with wide ranges of the clinical spectrum from asymptomatic masses revealed incidentally to severe symptoms such as marked abdominal distension, urinary retention, or even, intestinal obstruction. In this case, the patient presented

with excessive crying and abdominal distension, which is consistent with the literature describing postnatal presentation, when prenatal imaging is normal. The other differential diagnosis for intraabdominal cystic mass in a neonate and infant include a range of a dermoid cyst, ovarian cysts, sacrococcygeal teratoma, enteric duplication cysts, genital-urinary anomalies, anterior sacral meningocele and cystic lymphatic malformations [7].

Diagnosis is made primarily based on physical examination and imaging findings, sonography is the first line of modality, which may reveal hydrometrocolpos in the antenatal period as in many cases discussed in the literature [8]. However, in our case prenatal sonographic reports were unremarkable.

Imaging technologies like ultrasound, CT scan, and MRI scan are useful in diagnosing hydrometrocolpos. MRI offers excellent superior soft tissue contrast as compared to CT scans, making it an ideal modality for the evaluation of soft tissue body parts [9]. However, as demonstrated in this case, the patient underwent a contrast-enhanced CT scan due to initial misdiagnosis as an enteric duplication or mesenteric cysts, which underscores the importance of hydrometrocolpos in the differential diagnosis of pelvic masses and the importance of thorough perineal examination. Definitive diagnosis is confirmed through physical examination, revealing a bulging imperforate hymen; however, in the present case, physical examination was not performed initially.

Hydrometrocolpos may cause significant morbidity, so prompt identification and treatment is crucial. It is associated with various complications, the most common complication is bladder and ureteric compression, leading to dilatation of the pelvicalyceal system, which can ultimately cause kidney injury [10]. Additional adverse outcomes include ascites and pyometrocolpos which have been documented in the literature [11]. Infants with hydrocolpos and urogenital sinus are prone to sepsis, rupture, and peritonitis. Fatalities have been reported due to sepsis linked with hydrocolpos, particularly if diagnosis and intervention are delayed [12]. In our case, imaging studies demonstrate mild right-sided hydronephrosis due to compression by a large distended uterine cavity and there were no other complications.

The standard treatment is surgical intervention, timely hymenectomy or hymenotomy is required in cases of hydrometrocolpos to relieve the obstruction and drain accumulated fluid, thus preventing irreversible renal

injury and other complications [13]. Early diagnosis and treatment are crucial, as evidenced by this case where about 200 ml of milky fluid drained successfully. The outcome is excellent with timely intervention, as evidenced by the normalization of uterine morphology and resolution of hydronephrosis on follow-up imaging in this patient.

CONCLUSION

Hydrometrocolpos is a rare disorder in infants, usually detected in the immediate neonatal period but this case presented at two months of age. Radiologists should be vigilant and consider it as a possible differential while evaluating unexplained intraabdominal cystic masses in girls, even in the absence of prenatal findings. Prompt imaging and careful physical examination are essential for early diagnosis, while timely surgery helps prevent complications and ensure favorable outcomes.

CONSENT FOR PUBLICATION

Informed written consent was taken from the parents for publication of this case report.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

ACKNOWLEDGEMENTS

Declared none.

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