Hydrocolpos, Uterus Didelphys and Septate Vagina in Association with Ascites: Antenatal Sonographic Detection

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Hydrometrocolpos is a rare congenital disorder consisting of cystic dilation of the vagina and uterus and accumulation of mucus secretions resulting from genital tract obstruction.1 The obstruction can be produced by imperforate hymen, transverse vaginal membrane, or some degree of vaginal atresia.2 Excessive intrauterine stimulation of the cervical mucus glands by circulating maternal estrogens produces the accumulated fluid.1,3 This was first described by Godfroy in 1856.1,4,5 Only sporadic cases were reported until 1940, when Mahoney and Chamberlain presented their definitive descriptions of the condition. Since then 200 cases have been presented or reviewed in the literature.1,5 Hydrocolpos or hydrometrocolpos commonly first appears either in the neonatal period or at menarche.5 This report describes the antenatal sonographic detection of a cystic pelvic mass subsequently proved to be hydrocolpos in a fetus with ambiguous genitalia and uterovaginal duplication.

Few reports on the prenatal diagnosis of hydrometrocolpos by ultrasonography have been published. Davis and coworkers were the first to describe the antenatal sonographic diagnosis of hydrometrocolpos caused by an imperforate hymen.5 Hill and Hirsch reported an additional case of fetal hydrocolpos,3 also caused by imperforate hymen, detected by sonography,1 and Russ and colleagues described a case of hydrometrocolpos occurring with uterus didelphys and septate vagina detected in utero.4 Our case is notable for the association of fetal ascites and uterovaginal duplication in a patient with persistent urogenital sinus.

CASE REPORT

A 25 year old gravida 1 para 0 woman was referred for ultrasonographic evaluation at 31 weeks’ gestation for polyhydramnios. The first sonographic examination identified a moderate polyhydramnios, and fetal biometry results were in agreement with a mean menstrual age of 30 weeks. Real-time sonography of the fetus revealed a homogeneous midline cystic, septate mass in the fetal pelvis. The mass measured 5.5 cm in its widest transverse dimension, and in a sagittal plane it appeared tubular and elongated, with smooth, well-defined margins (Fig. 1). On transverse section, another fluid-filled image was observed that we thought was the fetal bladder (Fig. 1). We found no indication of bowel obstruction or other abnormality at this first examination. The fetus appeared to be female.
A subsequent ultrasonographic study 3 weeks later demonstrated a slight increase in size (7.2 cm in its widest diameter), no changes in texture, fetal ascites, and mild hydronephrosis (Fig. 2). We could still see another cystic structure (the fetal bladder) that appeared to be compressed by the mass. Follow-up sonographic examination showed increase in size of the mass. At the last ultrasonographic study (at 37 weeks of gestation) the mass measured 9.6 cm in its widest diameter and ascites could still be seen. At this time, the mother underwent cesarean section, and a 3070 g infant was delivered with Apgar scores of 6 and 8 at 1 and 5 min, respectively.

Physical examination of the neonate demonstrated a huge abdominal mass, ambiguous genitalia with a single perineal outlet for the urethra and vagina (a urogenital sinus), and an enlarged phallus-like clitoris (Fig. 3). The anus was patent.

A neonatal ultrasonogram was obtained, confirming the prenatal findings. The abdominal radiograph demonstrated a large mass that displaced the bowel loops to the left and superiorly (Fig. 4).

The infant underwent operation. At surgery the diagnosis of hydrocolpos was confirmed, and a uterus didelphys and septate vagina were visualized. No other abnormalities were detected after birth.

The infant died 1 month later of septicemia.

**DISCUSSION**

The prevalence of congenital dilation of the uterus and vagina is less than 1 in 30,000 births, although it has been reported to account for 15% of abdominal masses in newborn girls, surpassed in frequency only by hydronephrosis.1,6

Hydrocolpos (cystic dilation of the vagina) and hydrometrocolpos (cystic dilation of the vagina and uterus) result from congenital vaginal obstruction that is manifest in the newborn period.

Two major types of vaginal obstruction occur. In the first type, the presence of an imperforate hymen, a transverse vaginal septum, or segmental vaginal atresia results in cystic dilation of the vagina or uterus, or of both, from the accumulation of mucous secretions.³ The second type of hydrometrocolpos is associated with persistent urogenital sinus and cloacal anomalies. Normally, when the embryo is 7 weeks old, the urorectal septum arises between the allantois and the hindgut and gradually grows caudally, dividing the cloaca into an anterior portion (urogenital sinus) and a posterior portion (anorectal canal). Failure or maldevelopment of the urorectal septum results in cloacal anomalies.⁸

Neonates with a urogenital sinus have a single vestibule for the bladder and vagina; with a cloacal malformation there is a single perineal orifice for the bladder, vagina, and rectum.⁷ This can be the consequence of virilization of the fetus from congenital adrenocortical hyperplasia or exogenous or endogenous maternal androgens. An enlarged phallus from clitoral hypertrophy is present. In other cases, no obvious hormonal basis for the ambiguous genitalia and no chromosomal abnormality is evident.⁹

Hydrometrocolpos may occur because the communication of the distal vagina with the urogenital sinus frequently is stenotic and results in distention of the urine-filled vagina and sometimes the uterus. Alternatively, the urogenital sinus itself may be stenotic, which results in obstruction and distention of the bladder or vagina, or both.⁹

Hydrometrocolpos caused by vaginal or cervical atresia or by vaginal membrane is frequently accompanied by other anomalies.¹ The most common associations are imperforate anus, often with a rectouterine or rectovaginal fistula: urinary anomalies, including persistent urogenital sinus, unilateral renal agenesis, or hypoplasia; polycystic

**Figure 1** Sonogram at 31 weeks of gestation. Transverse scan (A) and longitudinal scan (B) show a double cystic mass measuring 5.5 × 4.3 cm in fetal pelvis.
kidneys; urethrovaginal or urethrouterine fistula; bicornuate uterus; septate vagina; polydactyly; congenital hip dislocation; esophageal atresia; and sacral hypoplasia.\textsuperscript{1,6,7}

A duplicated genital tract may be present, as in this case.\textsuperscript{9–11} The arrested descent of the urorectal septum, which normally separates the genitourinary tract from the gastrointestinal tract, interferes with fusion of the müllerian ducts. This results in duplication of the uterus and vagina (or a bicornuate or septate uterus or septate vagina).\textsuperscript{9}

Hydrocolpos occurs prenatally, as seen in seven of Blask and coworkers’ cases\textsuperscript{9} and in three others reported in literature by Davis and colleagues,\textsuperscript{4} Hill and Hirsch,\textsuperscript{1} and Russ and associates.\textsuperscript{3} Blask and coauthors reported a retrospective study of nine neonates with obstructed genital tract evaluated with sonography, five of whom had a urogenital sinus. Of these, four had prenatal ultrasonographic examinations and only one had uterovaginal duplication and intersex phenotype. Seven of the nine patients had a large spherical or ovoid cystic pelvic-abdominal mass that represented a markedly distended fluid-filled vagina.

In the case described by Davis and coworkers, the hydrometrocolpos was caused by imperforate hymen and appeared as a hypoechoic mass in the fetal pelvis, extending superiorly into the fetal abdomen and caudally into the perineum.\textsuperscript{4}

The fetal hydrocolpos reported by Hill and Hirsch also was caused by imperforate hymen and appeared as a homogeneous midline mass of medium echogenicity posterior to the bladder, which demonstrated a slight increase in size on a subsequent sonographic study, and mild right hydrenephrosis.\textsuperscript{1}

Russ and coworkers reported a case similar to ours of hydrometrocolpos occurring with uterus didelphys and septate vagina detected in utero.\textsuperscript{3} The mass was described as a cystic structure arising from the fetal pelvis and extending into the fetal abdomen, dividing it into four compartments. Moderate hydrenephrosis of both fetal kidneys was demonstrated. The infant had a persistent urogenital sinus.

None of these cases of hydrocolpos or hydrometrocolpos reported in literature had ascites as a presenting feature. Fetal ascites as seen in our case was described in three cases of cloacal anomalies reported by Petrikovsky and colleagues, but none of them had uterus didelphys.\textsuperscript{8} All three patients in this series were referred for evaluation of fetal ascites documented on the initial ultrasonogram and had a large cystic structure arising from the fetal pelvis.

Two patients had vaginal stenosis, one in association with hydrocolpos and the other with hydrometrocolpos.\textsuperscript{7} Fetal ascites was transient in the cases reported by Petrikovsky and colleagues, and it was also the referring cause for the ultrasonographic evaluation. In our case the double cystic structure arising from the fetal pelvis was the first abnormality detected; ascites was noted only on the second sonogram and was present on all follow-up studies and confirmed postnatally. No other anomalies and no other causes of ascites, such as hydrops fetalis, meconium peritonitis, ovarian cysts or bowel obstruction, were noted. Although bifid uterus, hydrocolpos, and urogenital sinus can be found among the various conditions associated with nonimmune hydrops fetalis, the only finding in this case was ascites. No pleural or pericardial effusions, subcutaneous edema, or placental thickening was observed.

On the basis of the sonographic findings on serial

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**Figure 2** Static image scan. Ascites (arrows) and polyhydramnios are evident.

**Figure 3** Newborn infant with abdominal distention (mass) and ambiguous genitalia. Note the hypertrophied clitoris and single orifice with urethral catheter.
ultrasonography in their study, Petrikovsky and coauthors suggested that in the early stage of the formation of a cloacal anomaly, urine is allowed to escape via the fallopian tubes into the abdominal cavity, causing ascites, and that later occlusion of the tubal mucosa occurs, probably because of chronic irritation by urine and meconium, leading to blocking of abdominal leakage of urine.8

In our case, however, the ascites was not transient and the hydrocolpos was not associated with a cloacal anomaly but with a persistent urogenital sinus. The bladder frequently cannot be identified on prenatal or postnatal sonograms, presumably because of compression by the distended vagina or because the vagina is more distensible than the bladder and fills preferentially.3 This was not the case in our patient. The bladder was identified in the first examination and was compressed by the distended vagina on serial sonograms.

Hydrocolpos and hydrometrocolpos should always be a differential diagnostic consideration when a cystic midline mass or multiple cystic masses with internal echoes or a fluid-debris level are identified arising from the fetal pelvis. Ovarian cyst, meconial cyst, duplication cyst, bladder or ureteral abnormality, and a rectovaginal fistula in a male infant with anal atresia are other differential diagnostic possibilities.3,12,13

REFERENCES


Figure 4 Plain film of the abdomen. A large mass displacing the bowel loops to the left and superiorly (arrows) can be observed.