Prenatal Diagnosis of Persistent Cloaca With Hydrometrocolpos and Ascites by Magnetic Resonance Imaging in One Fetus of a Dizygotic Twin Pregnancy

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A 34-year-old primigravid woman presented at 29 weeks of gestation for evaluation of fetal ascites and an intra-abdominal echogenic cystic mass (Figure 1) in one twin of a twin pregnancy. The woman had not undergone any assisted reproductive technology. Aspiration of the ascites and the cystic mass revealed multiple epithelial cells and cytogenetic analysis demonstrated a 46,XX karyotype in the affected co-twin. Ultrasound following aspiration showed a distended vagina connecting to the uterine cavity and compressing the urinary bladder (Figure 2). Ultrafast magnetic resonance imaging (MRI) of the affected co-twin revealed massive ascites, a compressed urinary bladder, a distended vagina, a dilated uterus, and a dilated distal colon, consistent with a diagnosis of persistent cloaca with hydrometrocolpos and ascites (Figure 3). The unaffected co-twin (1,306 g) and affected co-twin (2,108 g) were delivered uneventfully by cesarean section at 31 weeks of gestation. Both twins had a karyotype of 46,XX. A zygosity test determined dizygosity. The affected co-twin had meconium peritonitis, urinary ascites, and a persistent cloaca. The urinary, genital, and intestinal tracts converged into a cloacal canal with a single opening at the perineum. The ascites was caused by drainage of the urine into the abdominal cavity via the vagina, the uterus and the Fallopian tubes, as well as by irritation of the peritoneum by urine and meconium. Hydrometrocolpos was caused by fluid accumulation resulting from distal vaginal obstruction through backward pressure from the cloacal canal. The hydrometrocolpos compressed the bladder causing partial bladder outlet obstruction. Dilation of the distal colon was caused by direct compression from the hydrometrocolpos and narrowing of the rectal communication. The affected infant was doing well at 1 year and 6 months of age, after corrective reconstructive surgery.

A persistent cloaca results from failure or maldevelopment of the urorectal septum that divides the urogenital sinus and anorectal canal [1]. Hydrometrocolpos

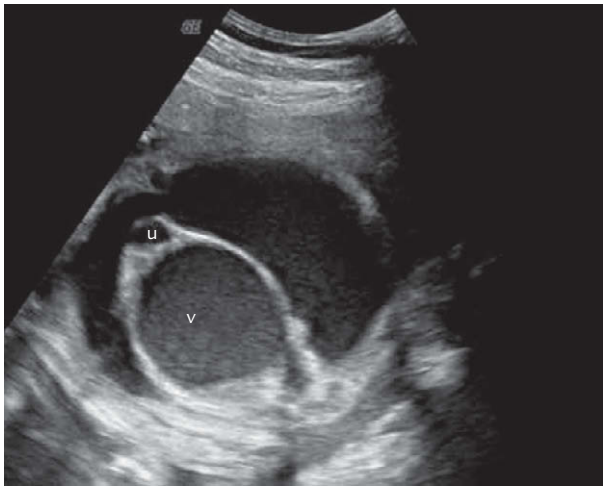

Figure 1. Prenatal ultrasound at 29 weeks of gestation showing fetal ascites with an intraabdominal echogenic cystic mass. u = uterus; v = vagina.

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Accepted: October 9, 2009

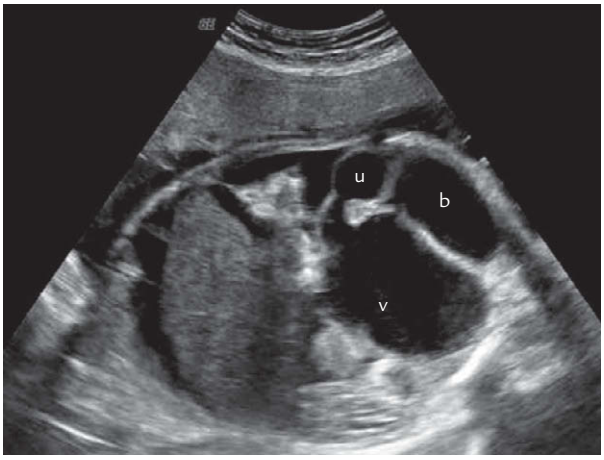

Figure 2. Ultrasound following aspiration showing a distended vagina (v) connecting to the uterine cavity (u) and compressing the urinary bladder (b).

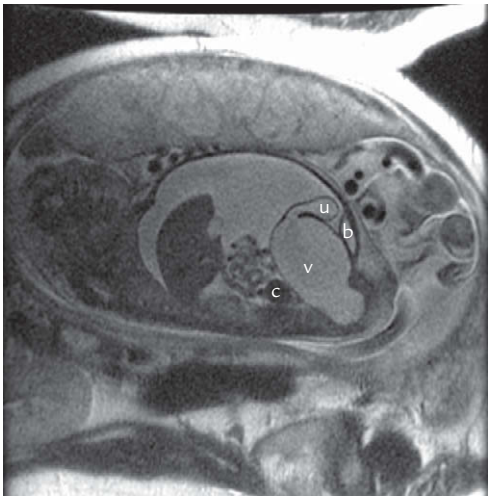

Figure 3. Magnetic resonance imaging showing a compressed urinary bladder (b), a distended vagina (v), a dilated uterus (u) and a dilated distal colon (c).

is caused by accumulated secretions from the reproductive glands resulting from obstruction of the vagina by an intact hymen, a midplane transverse vaginal septum or vaginal atresia, and/or from accumulation of urine caused by a stenotic urogenital sinus, associated vesicovaginal fistulas, vesicouterine fistulas and urethrovaginal fistulas, or cloacal anomalies [2]. Hydrometrocolpos may present in association with a variety of malformations and syndromes, such as cloacal dysgenesis sequence, McKusick-Kaufman syndrome, Ellis-van Creveld syndrome, and Bardet-Biedl syndrome [3–5]. Fetal hydrometrocolpos secondary to a cloacal anomaly can be identified by complementary MRI [6–8]. A dizygotic twin pregnancy with a persistent cloaca in one fetus is very rare. Subramanian [9] suggested that the rectal signal caused by retained meconium on fetal MRI is an important imaging marker of cloacal anomaly in association with hydrometrocolpos, as clearly demonstrated in the current case. The MRI findings in this case were in accordance with previous observations suggesting that MRI provides useful information in patients with a dilated distal colon in addition to hydrometrocolpos and ascites, thus facilitating the prenatal diagnosis of cloacal anomalies.

Acknowledgments

This work was supported by research grants NSC-96-2314-B-195-008-MY3 and NSC-97-2314-B-195-006-MY3 from the National Science Council, and MMH-E-98004 from Mackay Memorial Hospital, Taipei, Taiwan.

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