

OHVIRA Syndrome: A Case Report

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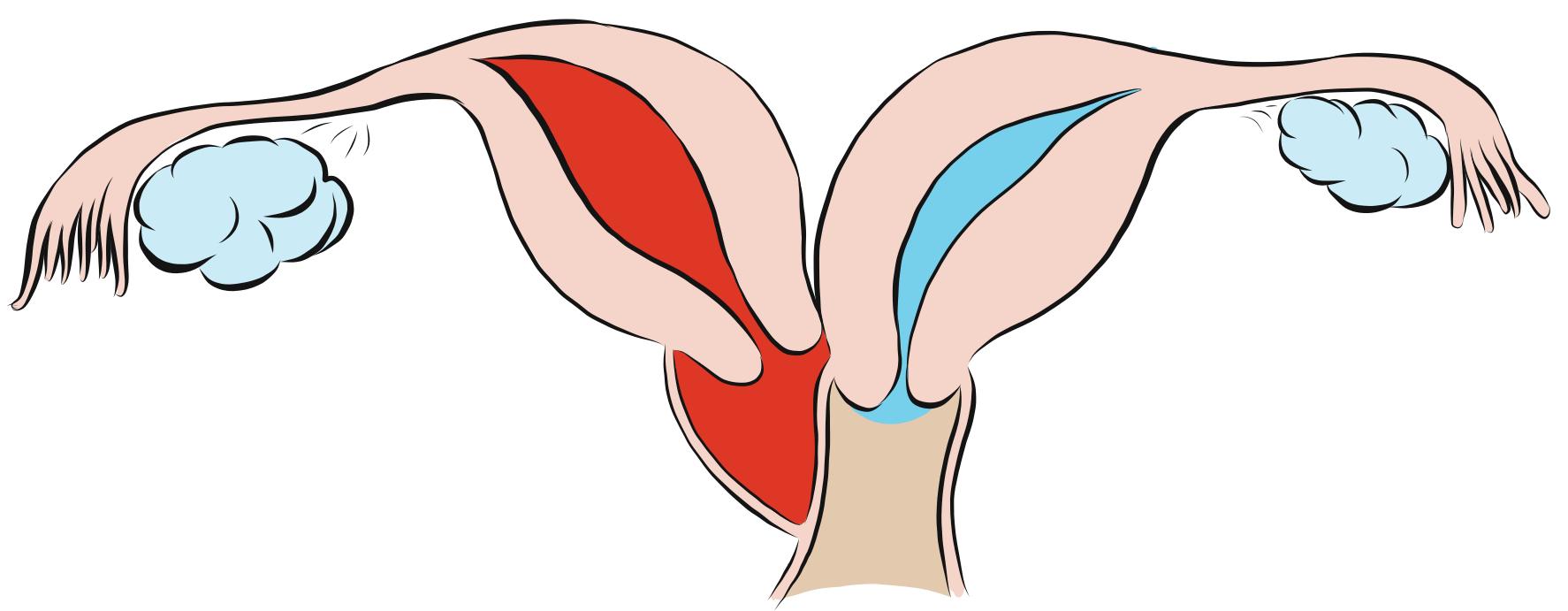
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Background

OHVIRA syndrome or Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly of female urogenital tract represented by the triad of uterine didelphys, obstructed hemivagina and ipsilateral renal agenesis. This rare variant of Müllerian duct anomalies represent failure of vertical and the lateral fusion of Müllerian ducts around 9 weeks of gestation. OHVIRA syndrome is about 2-3% of Müllerian abnormalities.

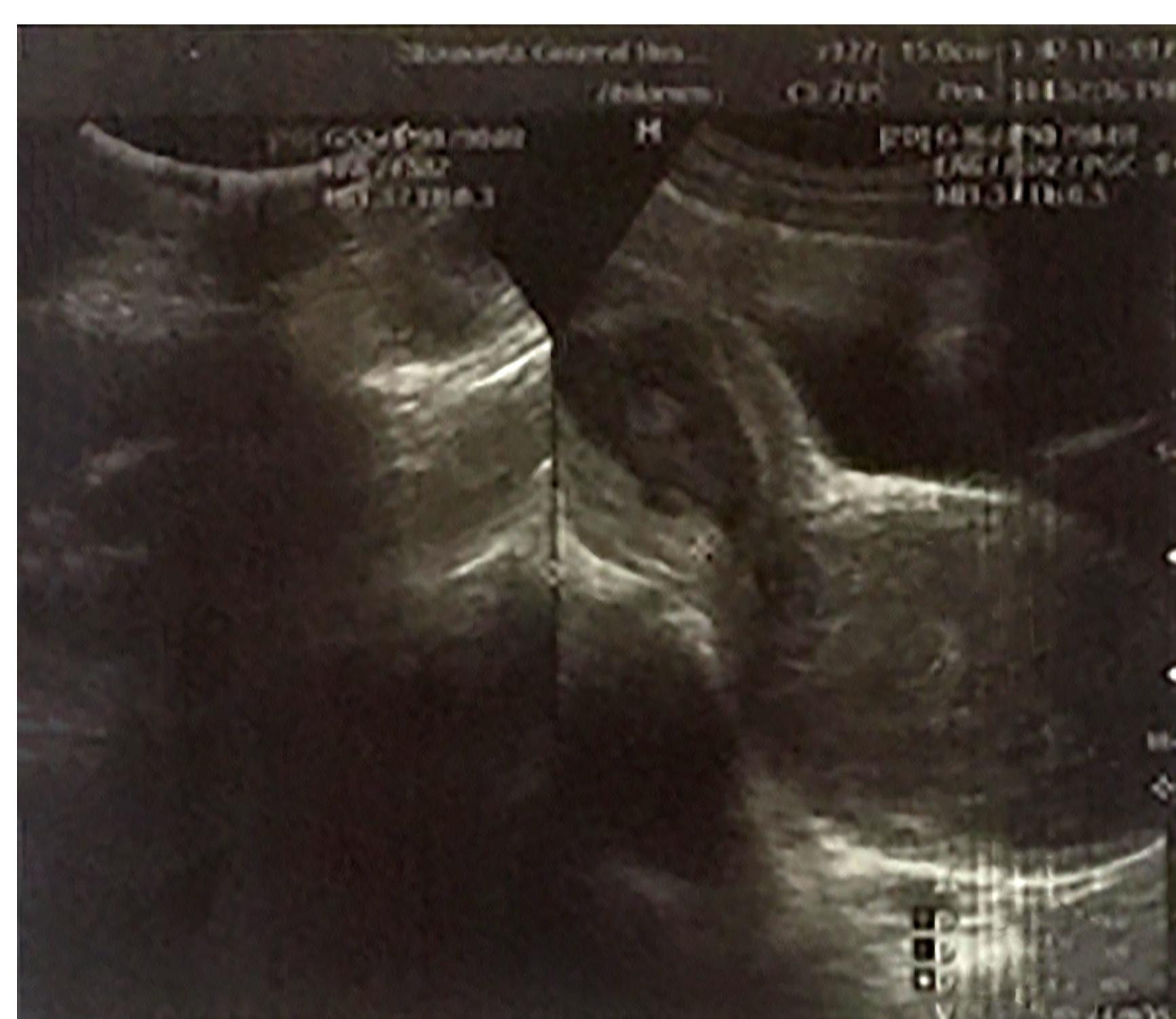
Paramesonephric duct abnormalities are often associated with mesonephric duct abnormalities (43%) and ipsilateral renal agenesis is the most common association.



Case Report

A 13 years old unmarried girl admitted in Gynae department of Faridpur Medical College Hospital, Bangladesh, on 15th March 2018 with acute retention of urine and lower abdominal pain. It was not related to fever, vomiting, dysuria or loin pain. She had no history of any urinary complaints before this incidence. Her menstruation was established 6 months back with regular cycle, scanty flow and associated with dysmenorrhea. After catheterization, a tender mass (about 8x6 cm) was palpable in lower abdomen with restricted mobility and get below of the swelling was not possible. Transabdominal ultrasonography of the whole abdomen showed uterus was bicornuate with hugely dilated cervix with hematocolpos. Right kidney was absent. Our clinical diagnosis was uterine didelphys where left half was functioning well with patent outflow tract, for which reason the patient was regularly menstruating. But in right half there was obstruction in some level producing hematocolpos. As she was a regularly menstruating girl, it was unusual to think about hematometra due to outflow tract obstruction. On the other hand, such a large chocolate cyst is not common in a girl with a history of menarche 6 months back.

EVA revealed a mass in right side of lower abdomen bulging through vagina. Cervix could not be identified. A cruciate incision was made over vaginal wall. Huge amount (about 1.5 liter) of inspiced blood was escaped out. Part of vaginal wall was excised. A Folley's catheter was introduced through cervix to keep patency of the outflow tract. In surgical exploration, a patent vaginal cavity and cervix was felt in left side.



Patient was discharged on third postoperative day with antibiotics and intrauterine catheter. She came for follow up after 14 days when catheter was removed. Subsequent follow ups were scheduled after 3 months and 6 months. Her menstruation was regular with normal flow and duration.

Discussion

OHVIRA is a rare syndrome of Müllerian and Wolffian duct abnormalities. HWW is commonly diagnosed at the time of menarche. The most common presentation is pelvic pain (90%), abdominal mass (40%) and pressure symptoms like urinary retention. Due to its rarity and lack of awareness, it is often misdiagnosed or diagnosis is delayed. A regular menstrual history always makes a confusion in diagnosis of outflow tract obstruction. A simple excision of vaginal septum is enough to relieve the symptoms and fertility preservation.

Reference

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