

Endometriosis in Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome: A case report and literature review on uterine-conserving approaches

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ABSTRACT

Introduction: Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a congenital disorder characterized by agenesis or aplasia of the uterus and upper part of the vagina in females with a normal female karyotype (46, XX). In rare cases of MRKH with functioning rudimentary uterine remnants, endometriosis is common. **Case Description:** A 38-year-old, single woman known to have Type II MRKH syndrome presented with recurrent severe pelvic pain. She was first diagnosed at the age of 16 after complaining of primary amenorrhea associated with cyclical pelvic pain. Karyotyping, a diagnostic laparoscopy, and findings of conductive hearing loss and unilateral renal agenesis confirmed Type II MRKH syndrome. Magnetic resonance imaging and serial pelvic ultrasounds revealed a functioning right rudimentary uterine remnant with hematometra. Over a period of 22 years, the patient suffered from mild-to-moderate dysmenorrhea and was treated with medical therapy unsuccessfully. Finally, the patient required admission for severe pelvic pain not responding to hormonal suppression. Despite absolute uterine factor infertility and the prohibition of gestational surrogacy, the patient was still keen to conserve her functioning uterus. Diagnostic laparoscopy revealed the presence of endometriosis and a right ovarian endometrioma. **Discussion:** The mainstay of treatment for chronic pelvic pain and endometriosis associated with obstructed functioning rudimentary uterine remnants in women with MRKH is surgical resection. However, the psychological impact of having MRKH syndrome should not be underestimated. Alternatives to surgical resection must be discussed with thorough counselling, support and careful dialogue with the patient is necessary.

Haematocolpos due to imperforate hymen: Seek and you shall find

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ABSTRACT

Introduction: Imperforate hymen is a condition with an incidence of 0.05-0.1%, where the hymen obstructs the vaginal opening completely. This may lead to haematocolpos, where menstrual blood is trapped in the vagina as opposed to being expelled. Diagnosis and treatment are crucial to prevent potential sequelae including infection, urinary retention, hydronephrosis, renal failure, and subfertility. **Case Description:** A 10-year-old girl, presented to the emergency department with a three-day history of suprapubic pain. She had 2 prior visits to the emergency department within the same week and was treated for urinary tract infection and constipation colic. She was referred to us on her third visit for a suspected twisted ovarian cyst. Abdominal palpation revealed a tender mass palpable at the umbilicus level. Perineal examination showed an imperforate bulging hymen. Ultrasound showed a fluid-filled mass measuring 13 x 6 cm posterior to the bladder, inseparable from and inferior to the uterus, consistent with haematocolpos due to imperforate hymen. A cruciate hymenotomy was performed and she made a good recovery post-operatively and subsequently had normal menses. **Discussion:** Often there is a missed or delayed diagnosis of this condition attributed to its low incidence, non-specific symptoms, and infrequent genital examination. This case depicts the importance of gynaecological assessment in adolescent girls presenting with abdominal pain.