

An unusual presentation of a patient with OHVIRA syndrome: A case report

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Introduction

OHVIRA syndrome is referred to as uterine didelphys with obstructed hemivagina and ipsilateral renal agenesis.

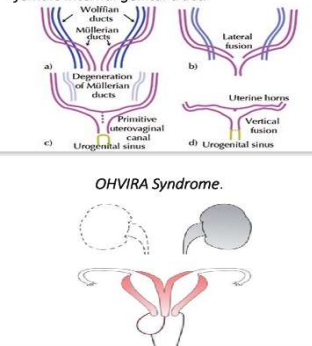
The reported incidence in the general population is 0.1%–3.8%. However, often this is incorrectly identified, incorrectly reported and, inappropriately treated.

The paired mesonephric and paramesonephric ducts represent the indifferent stage of the fetal internal genital system.

The paramesonephric ducts of either side fuse to form the female genital tract except the lower portion of the vagina. This process is referred to as Müllerian organogenesis.

Defects in this Mullerian organogenesis lead to complex malformations of the female genital tract such as OHVIRA syndrome.

Normal embryological development of the female internal genital tract.



Discussion

- Presentation of OHVIRA syndrome can be delayed especially when there is a small communication between two vaginal cavities.
- When sexually active they get secondary infections and present with pyocolpos and hence purulent vaginal discharge
- Early accurate diagnosis after menarche followed by excision and marsupialization of the blind hemivagina offers complete relief of symptoms and preserves reproductive potential.

Case history

- A 24-year-old patient was presented with purulent vaginal discharge for 18 months. Examination revealed a bulge on the R/S vaginal wall along with discharge.
- Ultrasound scan (USS) revealed a uterine didelphys, a fluid-filled oval shape structure along the right side of the vagina; suspected obstructed hemivagina, and an absent right kidney. That confirmed the diagnosis of OHVIRA syndrome.
- Examination under anaesthesia revealed a normal vagina with a bulge right vaginal wall and a purulent discharge from a small dimple on the top of the bulge. A transverse incision was made over the right-sided bulging area and approximately 80 ml of pus drained suggestive of pyocolpos.
- After complete drainage, a normal right-side cervix was visualized inside the vaginal opening.
- Bilateral tubal patency was demonstrated simultaneously by doing a laparoscopic dye test.

Intraoperative findings



Purulent discharge from the from the R/S Vaginal wall after making an incision



Laparoscopic view of uterine didelphys