

EP 0288 : A case Series of Herlyn-Werner-Wunderlich Syndrome (HWWS) -A rare Type of Mullerian Abnormality

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Introduction

Herlyn-Werner-Wunderlich syndrome is a rare type of Mullerian abnormality characterised by Uterus Didelphys, Obstructed Hemivagina due to transverse septum and unilateral renal agenesis. Usual presentation with cyclical abdominal pain few months to a year after menarche. Early diagnosis and treatment is necessary to prevent long term complications.

Discussion

Gynaecologists should have a high suspicion of different Mullerian anomalies when patient presents with cyclical abdominal pain with or without vaginal bleeding. It can be easily confused with Imperforate hymen. Further imaging like MRI pelvis is warranted to confirm the diagnosis as proper surgery is necessary to prevent impact on fertility in the future. Multidisciplinary team involvement including a radiologist will help in estimating the distance of the transverse vaginal septum from the introitus, which is useful when planning the surgery.

Conclusion

Early correct diagnosis, Proper imaging and planned surgery are important in HWSS to prevent irreversible complications such as infertility.

Case Series

Our first case is 15-year-old girl presented after 1 year of menarche with cyclical bleeding with severe dysmenorrhoea. Initial cyclical pain later progressed to continuous pain even without menstruation. On examination she had a palpable 14-week size uterus with open vaginal canal. Further imaging with Ultrasound Scan and MRI diagnosed HWSS with a uterine didelphys, right sided hematocolpus and hematometra and absence right kidney. Diagnostic Laparoscopy and hysteroscopy performed and confirmed patent left cavity with obstructed right uterine cavity and upper vagina with a transverse vaginal septum. Transverse vaginal septum was incised and hematocolpus drained. A vaginal dilator inserted postoperatively to prevent stenosis. Her symptoms resolved after surgery. On follow-up well healed surgical site noted without stenosis.

Our second patient is a 16-year-old girl presented with a similar presentation 1 year later. Examination and imaging confirmed HWSS with Right sided transverse septum and absent right kidney. Diagnostic laparoscopy and hysteroscopy were done to demonstrate that cervixes were connected to their respective uterus and uterus has a patent ostium and normal fallopian tubes indicating the possibility of conception in the future. Transverse vaginal septum was excised as in previous case and vaginal dilators inserted for transient period. Patient completely recovered without stenosis or pain. Both the patients were referred to nephrologist for advice regarding single kidney.