

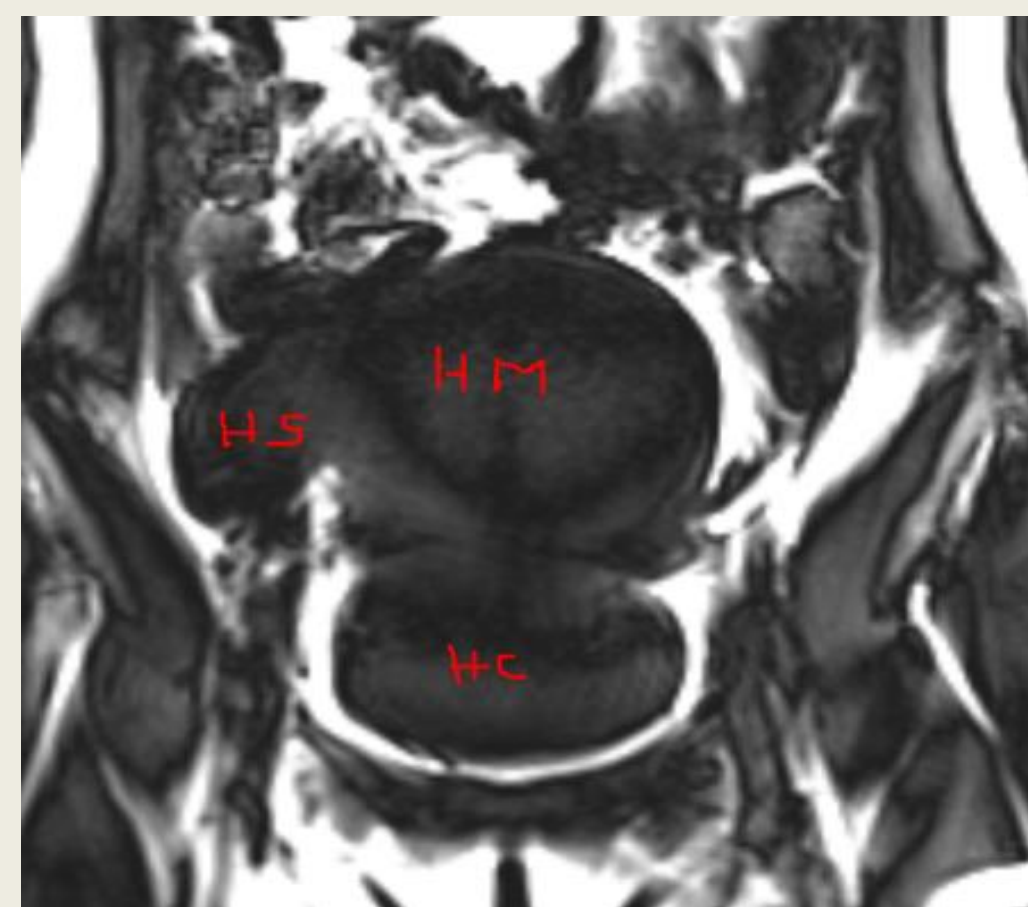


Case report

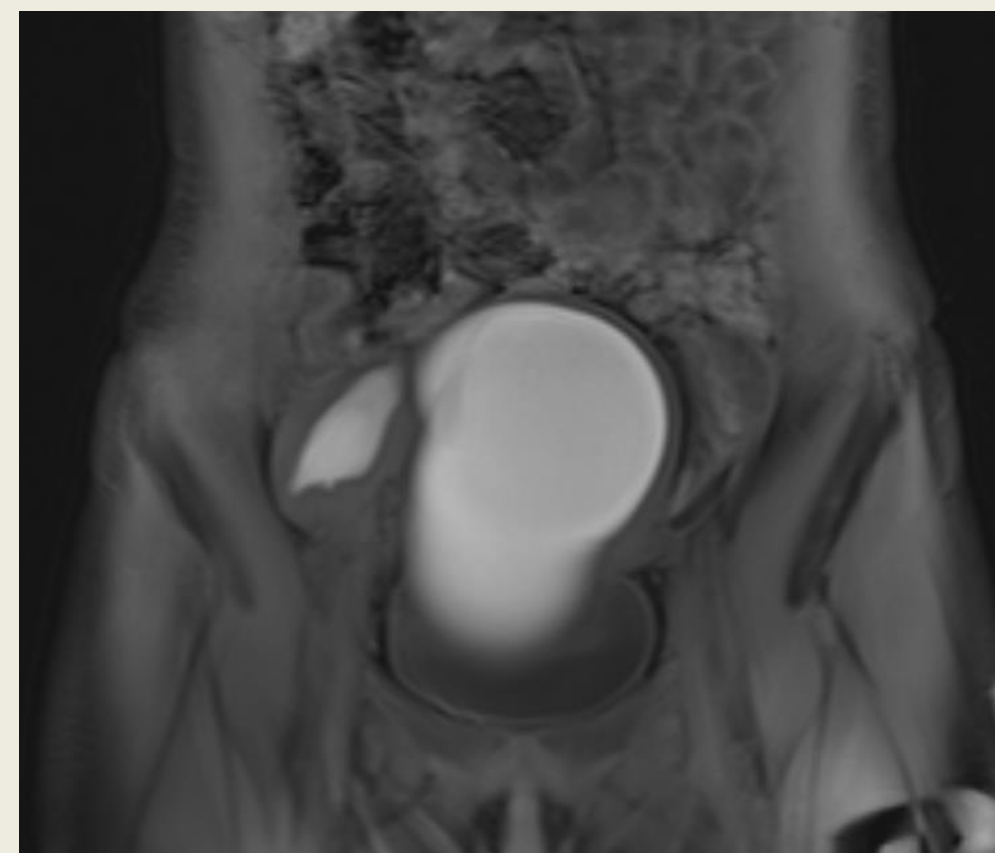
Our patient is a 15- year -old girl who was under the care of a gastroenterologist for her complaints of chronic constipation. She has a h/o anal fissure that was managed conservatively and had healed. She is also known to have a congenitally absent right kidney; renal functions are normal. She presented this time with cyclical pain around her back passage for the last one year, approximately starting with her menarche. She had regular monthly periods, but the bleeds were light.

Examination was unremarkable , no palpable abdominal mass, - no imperforate hymen, no vaginal bulge.

MRI pelvis – uterus didelphys with right sided vaginal septum, right sided obstructed hemi vagina with blood in the right side of the vagina, right horn of the uterus and right fallopian tube. Normal left uterine horn and tube, normal ovaries. Confirmed absence of the right kidney, with compensatory hypertrophy of the left kidney



HC - haematocolpos HM – Haematometra
HS- Haematosalpinx



Discussion- Embryology

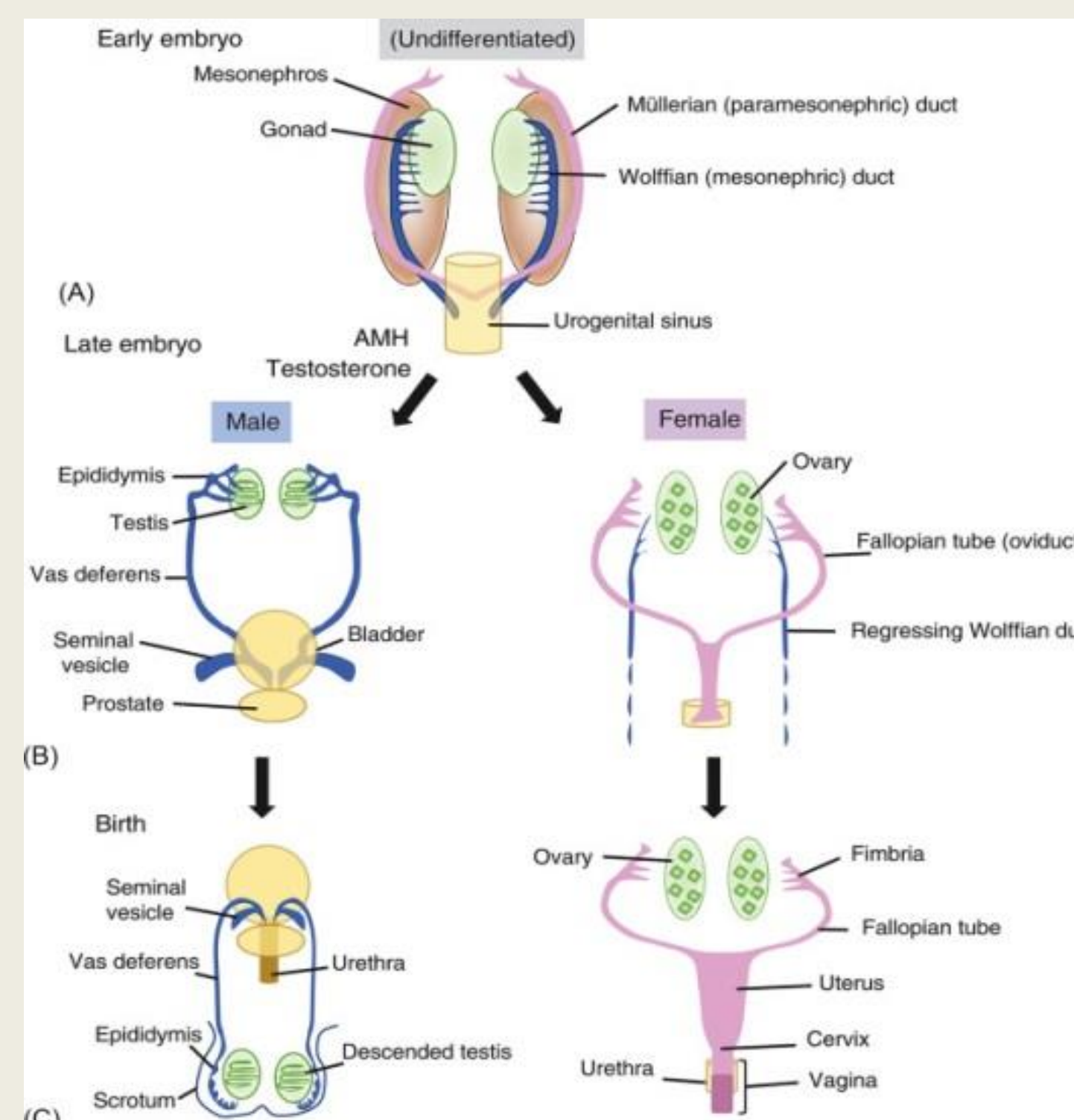
Classic theory

According to the classic theory, the uterus, fallopian tubes and upper third of the vagina develop from the fusion of the paired paramesonephric (Mullerian) ducts and the lower 2/3rd of vagina from the urogenital sinus. Aberration in the vertical or the horizontal fusion or arrest of paramesonephric ducts during course of development results in various uterine anomalies. These could be associated with renal, cardiac, axial skeletal and hearing defects in up to 30% of women.

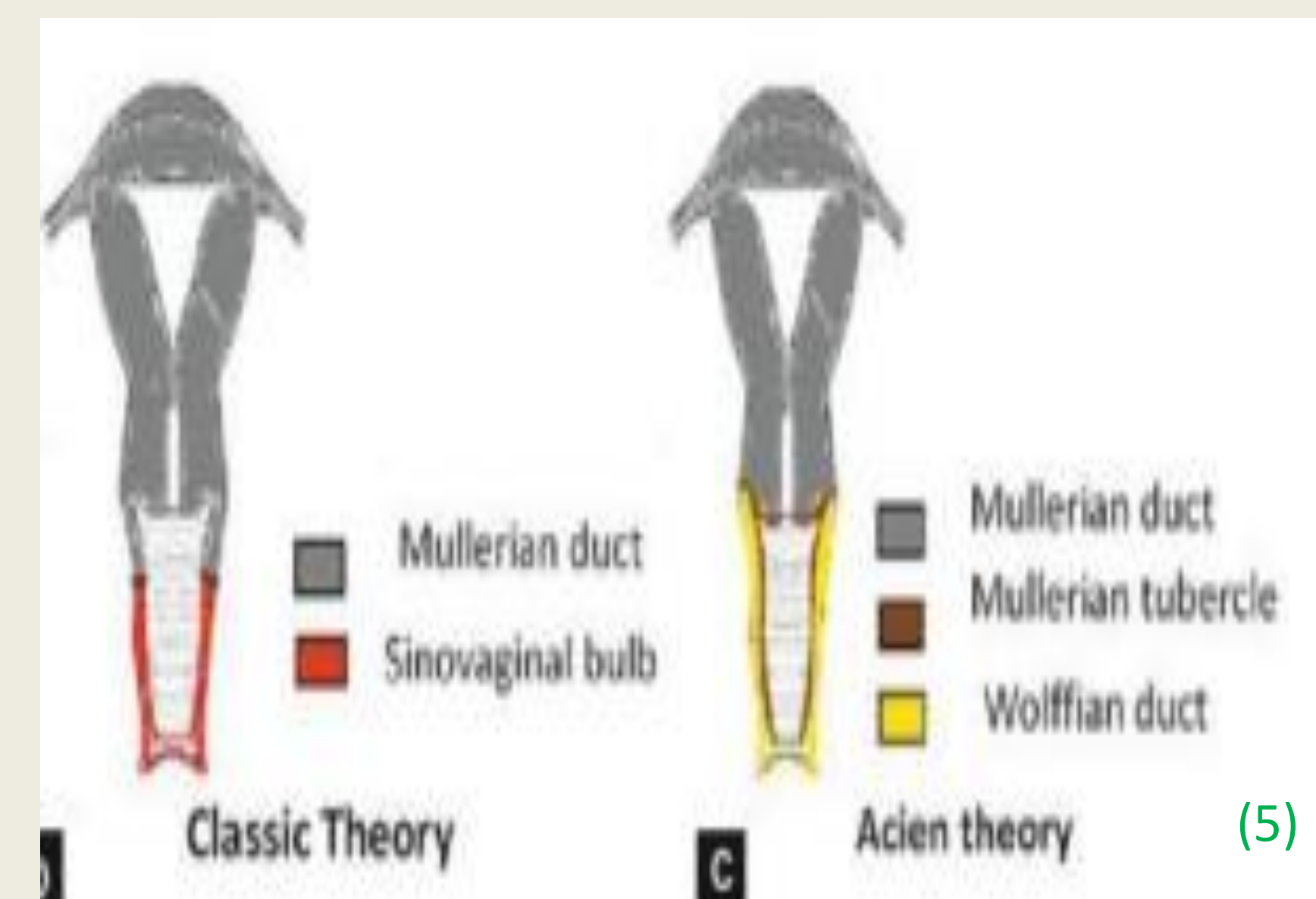
Acien's theory (2)

The classic theory was found inadequate in explaining complex Mullerian malformations like OHVIRA and their coexistence with renal anomalies. Acien proposed that the vagina was completely of mesonephric origin (Wolffian) (2) (4), explaining the OHVIRA syndrome. Hence, paramesonephric ducts do not contribute to formation of the vagina although Mullerian tubercle does, hence, the vagina is lined by cells from Mullerian tubercle.

Normal male and female genital development- classic theory

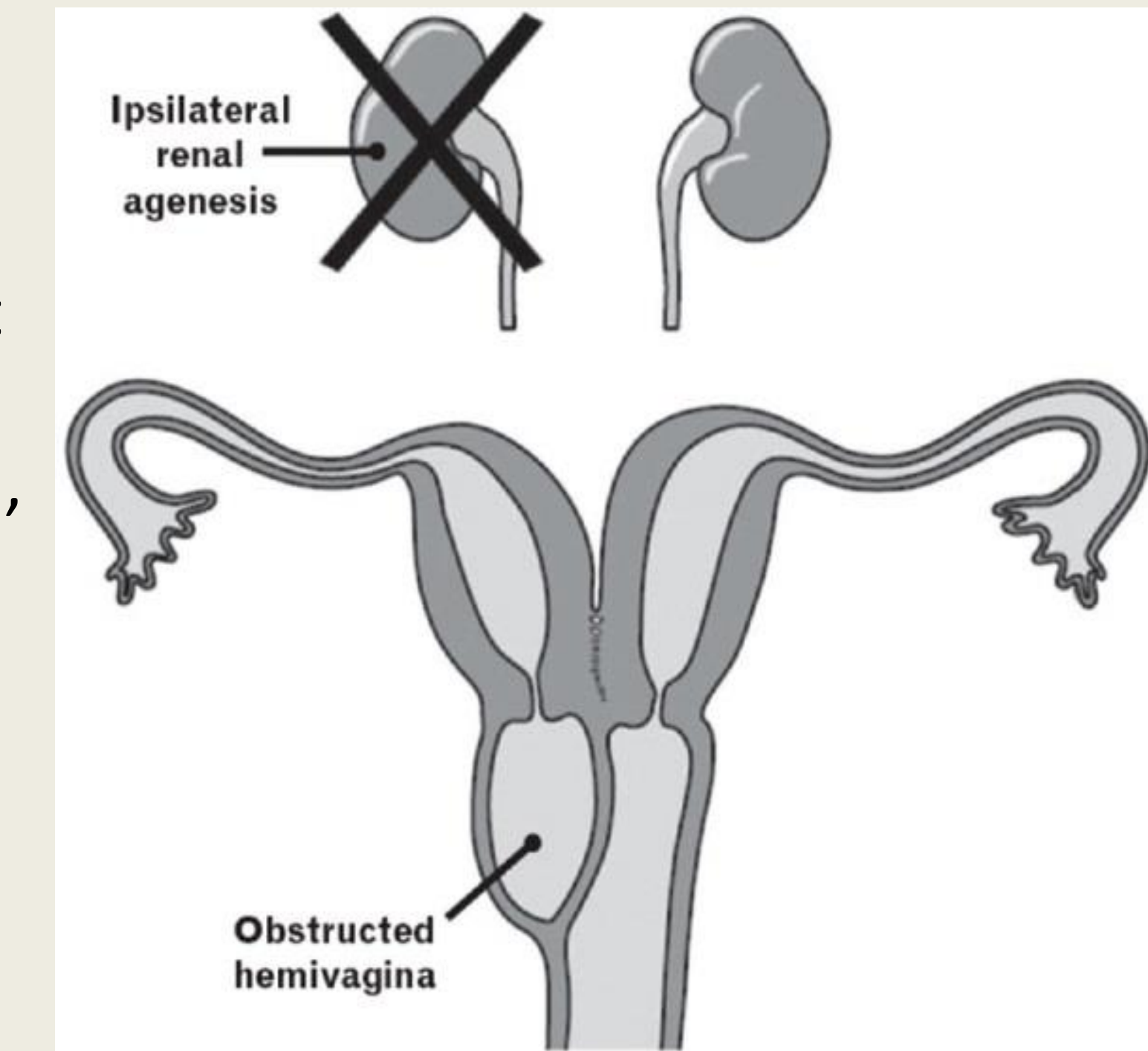


Acien's theory of vaginal development



OHVIRA -obstructed hemi-vagina with ipsilateral renal agenesis, also known as Herlyn-Werner-Wunderlich syndrome

It is a type of didelphic uterus in which a girl is born with a double uterus, a double vagina, with one side obstructed by a wall of tissue that may block menstrual blood from flowing out of the body and a missing kidney on the same side. Symptoms may include Pelvic pain, dysmenorrhoea, pelvic mass , Irregular bleeding, unusual vaginal discharge, vomiting, and constipation. Treatment is surgical, involving septal resection. Good counselling is an important aspect of management



Conclusion

The incidence of Mullerian defects is about 1.1 - 3.5% [1] and much higher (around 25%) in women with recurrent miscarriages and subfertility.[2] Review of literature revealed the mean incidence of uterus didelphys to be 11.1%. [2] with a strong association of renal agenesis with uterus didelphys (81%) [3] Only isolated case reports of OHVIRA have been published. Many of these present in an early infancy because of collection of secretions in the obstructed vagina under influence of maternal hormones; others are picked up within 1 - 2 years of onset of menarche like our patient, due to development of haematocolpos, hematometra, or even haematosalpinx. These patients present with cyclical dysmenorrhea, which can later evolve into persistent pelvic pain and endometriosis. To understand the pathogenesis of Mullerian anomalies and their association with renal anomalies, the embryology of utero-vaginal development needs to be reviewed. The new theory by Acien throws some light on this and is a topic of debate and more work.(4)

References

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Diagnosis-

OHVIRA- OBSTRUCTED HEMIVAGINA WITH IPSILATERAL RENAL AGENESIS

Patient and her parents were counselled. Suppression of menstruation by down regulation with the GNRH analogue-leuporelin 3.75mg monthly, for 6m now. Planned for vaginal surgery- vaginal septal resection at higher centre. Meanwhile she has developed menopausal symptoms and has been started on HRT with tibolone 2.5mg daily. Currently awaiting date for surgery