

CASE 4

A. 16 Y/O F

B. 19 Y/O F

Brief history-A

- 16 year old female
- Chief complain:
 - Amenorrhea for consultation
- PHx:
 - None

Imaging studies-A

- Pelvic MRI (NoC + C)

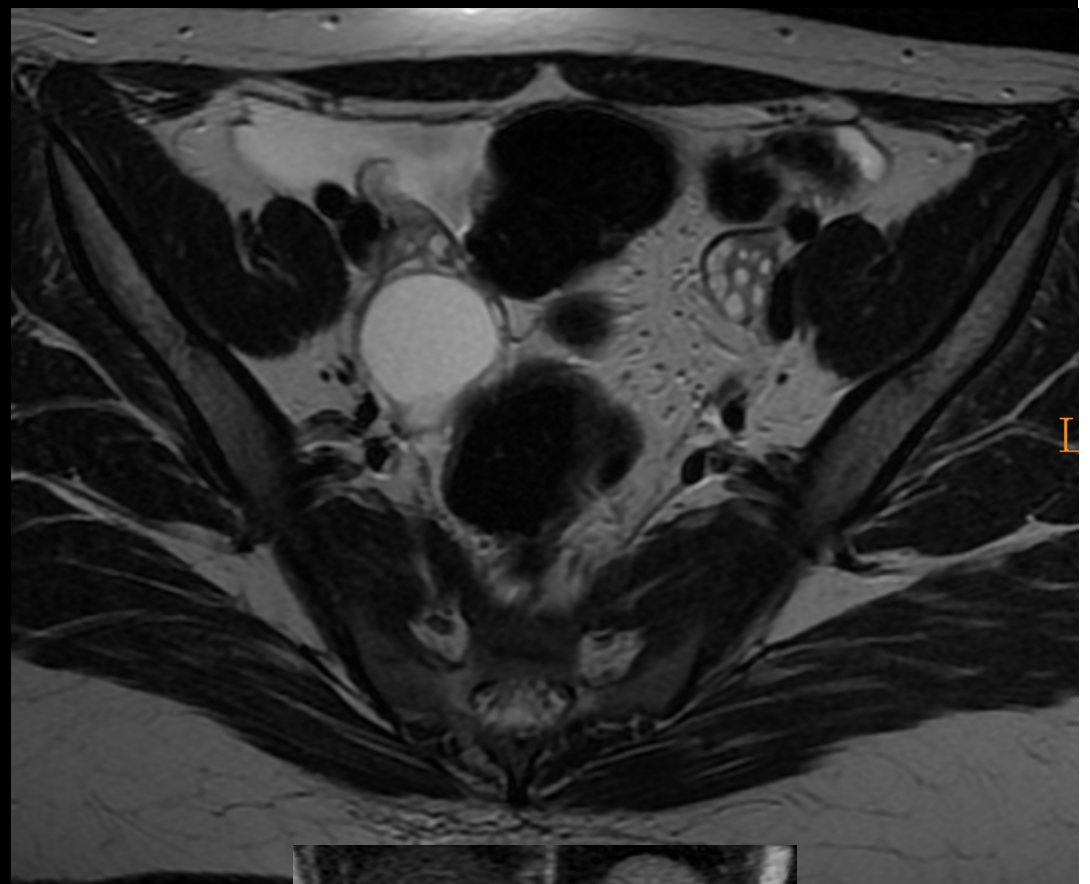
Brief history-B

- 19 years old female.
- PH: Development delay, amenorrhea
 - poor impulse control. Short attention span.

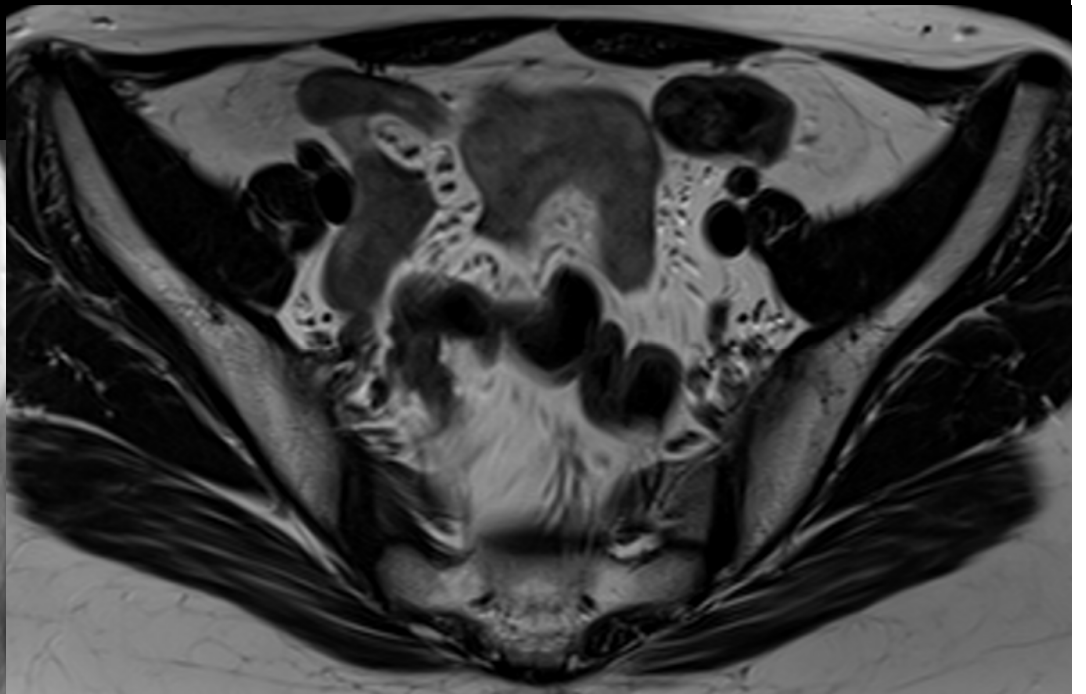
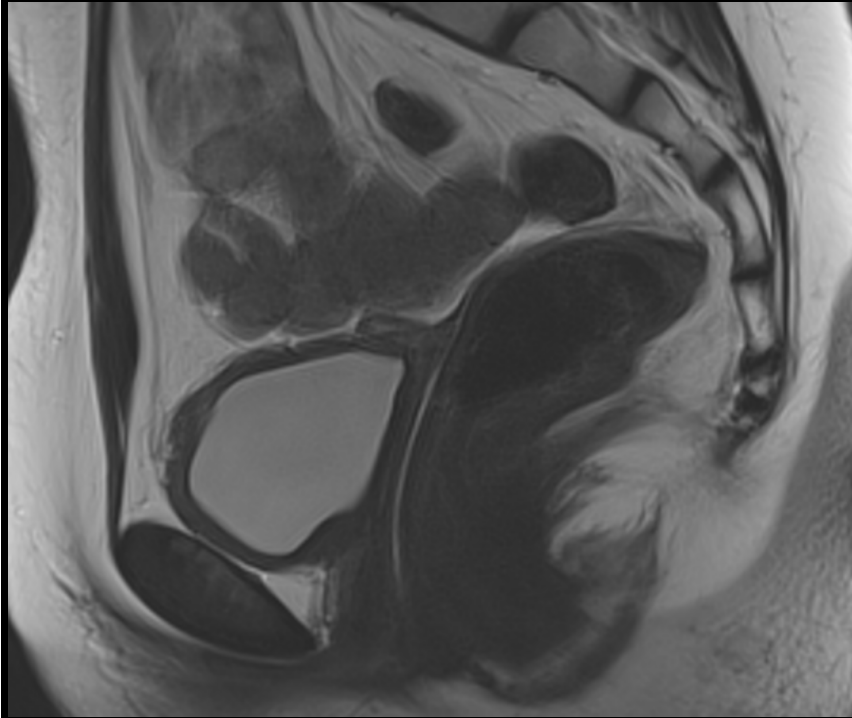
Imaging studies-B

- Pelvic MRI (NoC)

A.



B.



- DDX:
 - A. **Mayer-Rokitansky-Küster-Hauser syndrome**
(Extreme type of Müllerian Duct Anomalies)
 - B. **Gonadal dysgenesis + Müllerian Duct Anomalies**
 - This patient is a 46XY female

Classification of Müllerian Duct Anomalies

- Müllerian agenesis or hypoplasia (class I in American Fertility Society classification system)
 - Complete or segmental agenesis or variable degrees of uterovaginal hypoplasia
 - **Mayer-Rokitansky-Küster-Hauser syndrome**
 - Extreme presentation of müllerian agenesis with absence of proximal vagina, cervix, and uterus
- Unicornuate uterus (class II)
 - Partial or complete unilateral hypoplasia
- Uterus didelphys (class III)
 - Duplication of uterus
- Bicornuate uterus (class IV)
 - Incomplete fusion of superior uterovaginal canal
- Septate uterus (class V)
 - Incomplete resorption of uterine septum
- Arcuate uterus (class VI)
 - Near complete resorption of uterine septum

END