Mullerian Duct Anomalies:  
A Clinical Review

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Embryology of the Female Reproductive tract

The Development of the genital system begins at the 5-6th week of gestation
- Female development is determined by the presence of XX  
- As ~9th week of gestation the Mullerian and Wolffian ducts coexist
- The absence of testosterone leads to regression of the Wolffian duct
- The absence of Anti-Mullerian Hormone allows for the development of the Mullerian Ducts

Mullerian Ducts (paramesonephric)
- The Mullerian Ducts develop Cephalocaudally from the embryonal mesoderm forming the fallopian tubes
- The Mullerian Ducts then fuse distally creating the uterus, cervix and upper 1/3 of the vagina.
- By the 12th week of gestation, the Uterus assumes its mature morphological shape.
- By the 22nd week of gestation, the entire process is complete resulting in uterus, cervix and uterine cavity.
- Mullerian Duct Anomalies are a result of interruptions at different stages of development of the female genitourinary system.
- Renal tract anomalies are associated with MDA in up to 10% of cases due to the close embryologic relationship between the paramesonephric and mesonephric ducts.

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Embryology of the Female Reproductive tract

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ASRM Classification and Estimated Prevalence of MDAs

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<td>VII</td>
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<td>DES related uterine anomalies</td>
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<td>VII-A</td>
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<td>T-shaped uterus</td>
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<td>VII-B</td>
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<td>T-shaped uterus with dilated horns</td>
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<td>VII-C</td>
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<td>Uterine hypoplasia</td>
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Class I: Mullerian Agenesis or Hypoplasia

- A 15-year-old girl presented to her pediatrician with primary amenorrhea.
- Her pediatrician saw an abnormal vaginal opening and referred the patient to a gynecologist.
- The patient reported thelarche at age 11 and adrenarche at age 12.
- She denied vaginal spotting, unusual drainage, and either cyclical or noncyclical abdominal pain.
- She was otherwise healthy and on no medications.
- She denied any sexual activity.
- FH: Her family history was negative for any gynecological issues.
- PE:
  - Tanner stage 4 breasts and a normal abdominal examination without any discomfort or palpable masses.
  - The patient was noted to be thin, and there was no evidence of a perineal mass.
  - She refused a pelvic examination because the last time it was very uncomfortable.
  - Imaging:
    - Abdominal ultrasound examination revealed a small uterus and normal ovaries.
    - Lab Studies:
      - Follicle-stimulating hormone and estradiol levels were in the normal postpubertal range.
- Plan:
  - A computed tomography revealed a normal appendix;
  - Ultrasonography revealed left ovary measuring 3 x 2 cm with multiple 0.5-mm simple cysts.
  - A diagnostic laparoscopy revealed the following:
    - Left ovary was visualized and fallopian tube was noted to be twisted
    - Right ovary and tube were not visualized in the appropriate location
    - Right adnexal structure was buried in the right sidewall.
  - Chromosomes were 46,XX, and her hormonal evaluation was normal.

Class I: Mullerian Agenesis or Hypoplasia

- An 11-year-old prepubertal girl presented with severe left lower quadrant abdominal pain and mild rebound.
- A computed tomography revealed a normal appendix;
- Ultrasonography revealed left ovary measuring 3 x 2 cm with multiple 0.5-mm simple cysts.
- A diagnostic laparoscopy revealed the following:
  - Left ovary was visualized and fallopian tube was noted to be twisted
  - Right ovary and tube were not visualized in the appropriate location
  - Right adnexal structure was buried in the right sidewall.
  - Chromosomes were 46,XX, and her hormonal evaluation was normal.

Class I: Mullerian Agenesis or Hypoplasia

- Most Severe type of MDA
- Segmental agenesis and variable degrees of uterovaginal hypoplasia
- Mayer-Rokitansky-Kuster-Hauser Syndrome
  - consists of complete vaginal agenesis
  - 90% of patients presenting with associated cervical and uterine agenesis
  - 10% of cases a rudimentary mullerian structure is identified which can be functional or nonfunctional (with or without endometrial layer)
- Failure of the distal ends of the mullerian ducts to progress beyond the 8th week of gestation results in complete absence of the uterus and vaginal. If both mullerian ducts fail to reach the urogenital sinus, only the vagina will be absent.

Class I: Mullerian Agenesis or Hypoplasia

- Symptoms
  - Complete agenesis
    - Primary Amenorrhea
    - Normal Ovarian Function → Normal secondary sex characteristics
  - Functional Remnant
    - Primary Amenorrhea
    - Normal Ovarian Function → Normal secondary sex characteristics
    - Severe cyclic abdominal pain secondary to Cryptomenorrhea and Hematometra

Class I: Mullerian Agenesis and Androgen Insensitivity

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<td>Ovaries</td>
<td>Testes</td>
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<tr>
<td>Pubic/Axillary Hair</td>
<td>Normal female</td>
<td>Absent/Sparse</td>
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<tr>
<td>Breast Development</td>
<td>Normal</td>
<td>Normal</td>
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<tr>
<td>Testosterone Level</td>
<td>Female Range</td>
<td>Male Range</td>
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<tr>
<td>Heredity</td>
<td>Unknown</td>
<td>Maternal X-linked recessive; 25% risk of affected child, 25% risk of cancer</td>
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<tr>
<td>Other Anomalies</td>
<td>Frequent</td>
<td>Rare</td>
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<tr>
<td>Gonadal neoplasia</td>
<td>Normal Incidence</td>
<td>5% incidence of malignant tumor</td>
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Class I: Mullerian Agenesis or Hypoplasia

- Imaging
  - Ultrasound
    - Initial evaluation; however evaluation of uterine remnant may be difficult due to limited acoustic window of US
  - MRI
    - Use to complement US
    - Important for differentiating between Uterine agenesis and hypoplasia
    - MRI Criteria
      - Uterine body and fundus- No uterine tissue (agenesis) or uterine tissue but no complete uterus (remnant)
      - Endometrium-Endometrial tissue may be present
      - Cervix- Absent, distorted, or length < of uterine body; absent or distorted endocervical canal
      - Vagina- Absent or replaced by a thin band of fibrous tissue
      - Other- Obstruction may be present
Figure 13a. Müllerian agenesis in a 17-year-old girl with primary amenorrhea and normal secondary sexual features.

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Class I: Mullerian Agenesis or Hypoplasia

Management

Correct Anatomic Abnormalities
- Phenotypic girls with Mayer-Rokitansky-Kuster-Hauser syndrome pt may opt for creation of neovagina
- Vaginal Dilators
  - Pt applies pressure to vaginal dimple twice a day for 20-30 minutes
  - First the length of the vagina is established and then the desired width using graduated dilators
  - Average amount of time needed to create neovagina is 12 months
  - Success rate is 93%
- McIndoe Procedure
  - Reconstructs the vagina between the bladder and rectum using a split thickness graft
  - Success rate is 89-92%
- Davydove Procedure
  - Reconstructs the vagina between the bladder and rectum using peritoneum
  - May be done Laparoscopically
- Vecchietti Procedure
  - A plastic "cone" is placed on the vaginal dimple
  - Traction is placed on the device ad subperitoneal sutures (placed via laparotomy or laparoscopy) are then used to pull the dimple upwards

Functional Mullerian remnants is present
- Surgery is recommended because of potential for pregnancy
- Evaluation of Kidneys and Spine. If either abnormal then evaluate Cardiac and Auditory function.

Fertility

In Vitro Surrogacy
- 50% live birth rate
- No increase in congenital anomalies

Adoption

Class II: Unicornuate Uterus

Class II: Unicornuate Uterus

22yo female G1P0 was brought to the hospital by her parents. She was amenorrheic x 140 days. She presents with the complaint of severe upper abdominal pain and right shoulder pain, weakness, and diaphoresis.

PE
- Pulse 140/65 BP 70/30
- Gen- pale, clammy, cold
- Abdominal exam- distended abdomen, tender to palpation with both rebound and guarding
- Bimanual exam- Cervical motion tenderness present, forniceal fullness appreciated

Imaging
- Ultrasonography revealed fluid in Morrison's pouch, paracolic gutters and pelvis. Fetus with biparietal diameter of 27mm was visualized in the anterior uterovesical pouch.

Management
- Fluid Resuscitation
- Laparotomy
  - Fetus was visualized attached to placenta by intact umbilical cord
  - Placenta was located in the ruptured right rudimentary horn
  - The right rudimentary horn was excised along with the pregnancy


[PDF]
Class II: Unicornuate Uterus

- Asymmetric anomaly in which a single-horned uterus opens into normal vagina.
- During approximately the 9th week of gestation, one Mullerian duct fails to elongate or reach the urogenital sinus with the contralateral duct.
- Most cases have a contralateral rudimentary horn which can be cavitary or noncavitary.

Symptoms
- Dysmenorrhea
- Hematometra
- Endometriosis
- Infertility
- Ectopic pregnancy
- Due to transperitoneal sperm migration
- Rupture of Rudimentary Horn
- Incidence of Rudimentary Horn Pregnancy: 1:76,000-140,000
- 70-90% rupture in second trimester
- Results in life-threatening hemorrhage with 90% of deaths occurring within 10-15 minutes of rupture
- Physical Exam Findings
  - Baart de la Balle’s sign: bimanual exam finding of palpable mass extending outward from uterine angle
  - Ruge Simon Syndrome: displacement of fundus to contralateral side with rotation of uterus and elevation of the affected horn
Class II: Unicornuate Uterus

- Obstetric Complications
  - Abnormal Fetal Lie
  - Intrauterine Growth Restriction
  - Live birth may be achieved in 29.2%
  - Preterm Delivery Rate is 44%
  - Miscarriage Rate is 29%
  - Ectopic pregnancies 4%.

Imaging
- Ultrasound
  - Suggested if a small, rounded uterus is identified in the lateral aspect of the pelvis.
  - Cannot accurately detect this MDA type.

- MRI
  - MRI Criteria for classification:
    - Uterine body and fundus: Elongated, banana-shaped, eccentric uterus.
    - Dilated normal horn with normal endometrial—myometrial width ratio.
    - A rudimentary horn may be present, may have endometrial tissue, and may communicate with main cavity.
    - Classified according to a rudimentary horn as follows:
      - Absent rudimentary horn
      - Rudimentary horn present with no endometrial tissue (nonfunctioning).
      - Rudimentary horn present with endometrial tissue that communicates with main cavity.
      - Rudimentary horn present with endometrial tissue that does not communicate with main cavity and may obstruct. The latter may present with abdominal pain and require surgical intervention.
    - Cervix: Normal.
  - If nonfunctional uterine horn is present, it typically has a low signal intensity on T2-weighted MRI with loss of normal zonal anatomy.
  - If functional uterine horn is present after puberty, it appears as a cavity deformed by the enlarged rudimentary horn which has high-signal-intensity center on both T1/T2 weighted images (finding compatible with hematometra).

Class III- Unicornuate Uterus
Class III: Unicornuate Uterus

- **Management**
  - Nonfunctional rudimentary horn
    - No surgical intervention required
  - Functional noncommunicating horn
    - Pregnancy can occur regardless of if horn is communicating or noncommunicating
    - If pregnancy is identified before mid-second trimester then abortion is recommended
    - If pregnancy achieves mid-second trimester, then attempt tocolysis
      - Do not use oxytocin, prostaglandin, or uterine stimulants
      - Hysterotomy should be completed if uterine wall thickness is <5mm or fetal lung maturity obtained.

Class III: Uterine Didelphys

- A 37yo G2P3, with a documented 2-horned bicornuate uterus, was diagnosed at 9 weeks with a twin pregnancy with one embryo in each horn.
- **HPI**
  - At 27 weeks, the patient presented with sudden abdominal pain radiating to the back. Patient reported no vaginal bleeding, no loss of fluid, irregular contractions, fetal movement present.
- **PE**
  - Abdomen was soft non-tender to palpation, no rebound/guarding
  - Mild irregular uterine contractions with no changes of the 2 cervices at digital examination.
  - Fetal tracing and ultrasonography were normal for both fetuses.
- **Day #1**
  - Labs - Hemoglobin - 10.4 g/dL; platelet count and liver enzymes were normal. No proteinuria was detected.
  - Diagnosis of PTL was considered and Tocolytic given (nicardipine 2mg/h). Betamethasone was also administered for fetal lung maturation.
  - Uterine contractions ceased rapidly, but the patient still complained of abdominal pain, which intensified the following day.
- **Day #2**
  - Labs - Hemoglobin - 8.5 g/dL. Platelet count, prothrombin time, and activated clotting time were normal. Liver function tests showed no hepatic cytolysis, but total hyperbilirubinemia, mostly the free form, and hemolysis were present. The urinary dipstick became protein positive.
  - Despite tocolysis, irregular uterine contractions reappeared and vaginal bleeding occurred.
  - Abruptio placentae in the right horn was suspected, and an elective cesarean was determined to be necessary.
  - Both twins were delivered through transverse hysterotomies, performed on both hemiuteri, but the incision in the right horn proved to be transplacental.
  - The placentas were removed, and no evidence of abruptio placentae was found.
  - Retrospective review of all ultrasound reports, performed at weekly intervals, revealed no evidence of abruptio placentae in the right horn.
  - Both neonates had uneventful postnatal outcome and were discharged at 6 and 8 weeks.
Class III: Uterine Didelphys

- Uterine didelphys is a symmetric anomaly
- Two completely separate uterine cavities are identified each with normal zonal anatomy, endometrial cavity and cervix
- Occurs at the 9th week of gestation when both paramesonephric ducts develop but the ducts fail to fuse at resulting in a duplicated system. The presence or absence of vaginal septum is defined by the degree of fusion failure
- No communication between the two cavities is present
- Complete or partial longitudinal vaginal septum is associated with the anomaly in 75% of cases
- Some patients with uterine didelphys present with unilateral hemivaginal septum → hematometrocolpos

Symptoms
- Symptoms are related to presence of obstruction
- Nonobstructive uterus didelphys
  - Asymptomatic
  - Diagnosed incidentally during pelvic exam when two cervices are identified
  - Diagnosed incidentally during imaging for another indication
- Obstructive uterus didelphys
  - Obstructed hemivagina typically becomes symptomatic with menarche → cyclic pelvic pain
  - Endometriosis
  - Pelvic adhesive disease
Class III - Uterine Didelphys

- Obstetric Complications
  - Fetal survival occurs in 41-64% of pregnancies
  - Premature births occur 20-45% of the time
  - Miscarriages occur in 32-52% of women

- Imaging
  - Ultrasound
    - Two completely separate uterine horns, each with its own endometrial cavity can be visualized
    - No communication between the two horns can be seen
    - Large fundal cleft may be noted, as well as, cervical duplication
  - MRI
    - MRI Criteria for Classification
      - Uterine body and fundus: Two separate uteri, which can be joined at body, deep fundal cleft
      - Endometrium: No communication between the two endometrial cavities, normal endometrial—myometrial width ratio in each uterus
      - Cervix: Double
      - Vaginos: Longitudinal or oblique vaginal septum always present
  - Hematometrocolpos with varying degrees of distention may also be visualized within an obstructed hemivagina
Class III- Uterine Didelphys

- Management
  - Dependent on presence or absence of obstruction
  - Surgical removal of vaginal septum if obstruction or at patient request
  - Typically Uterine Didelphys does not require surgical intervention

Class IV- Bicornuate Uterus

- 26yo G2P2 presented to her physician for contraceptive counseling.
  - An IUD was placed at 6 wks postpartum
  - After Several Months the patient presented to her physician with a complaint of 2 wks delay in menses
  - Pregnancy Test – Positive
  - Ultrasound Exam Revealed a bicornuate uterus with pregnancy in one cornua and IUD in the second cornua
Class IV Bicornuate Uterus

- Two symmetric cornua are fused caudally with communication of the endometrial cavities at the level of the uterine isthmus.
- At the 9th week of gestation, the mullerian ducts incomplete fuse at the level of the uterine fundus.
- It is similar to uterine didelphys except some degree of fusion does take place.
Class IV- Bicornuate Uterus

- Symptoms
  - Typically Asymptomatic
  - May be discovered incidentally at imaging for other indications
  - May be discovered incidentally during cesarean delivery

- Obstetric Complications
  - Fetal Survival Rate is 57-63%
  - Higher prevalence of pregnancy loss
    - Spontaneous abortion rate 28-35%
    - Higher Incidence of preterm delivery
      - Premature Birth Rate 14-23%
  - The length of the septum is directly related to the incidence of abortion and preterm delivery with complete bicornuate uterus having the highest rate
  - Typically have fewer reproductive problems than Classes I-V

- Imaging
  - Ultrasound
    - Large indentation in the uterine fundus
    - Divergent uterine horns
    - Echogenic endometrial complexes
  - MRI
    - MRI Criteria for Classification
      - Fundus Indented; cleft, 1 cm or more deep
      - Septum- Present, muscular or combined muscular and fibrous
      - Bicornis: septum to external os
        - unicollis: septum does not reach external os
      - Cavitas- Single or divided by a septum
      - Vagina- Vaginal septum may be present in some cases
    - Normal zonal and endometrial-myometrial ratio are seen in both horns
    - Bicornuate Uterus is differentiated from the septate uterus by the shape of the external contour
    - The septum may consist of fibrous tissue demonstrating a low signal intensity on T2 weighted imaging
Class IV - Bicornuate Uterus

- Management
  - Typically does not require surgical intervention
  - Historically treated with metroplasty
    - Fertility outcomes were not improved

Class V - Septate Uterus

- Most common Mullerian Duct Anomaly
- After complete fusion of the mullerian ducts, the septum between them must be resorbed. Typically this resorption should occur by the 12th week of gestation.
- Studies have linked presence of a uterine septum with genetic absence of bcl-2 gene which is responsible for apoptosis.
- Septum is located in the midline fundal region
- Septum is composed of poorly vascularized fibromuscular tissue
- Complete septum extends from the fundal zone to the internal or external os and divides the endometrial cavity
- A partial septum does not reach the internal os
Class V - Septate Uterus

**Symptoms**
- Typically Asymptomatic
- Complete Septum
  - unilateral obstruction
  - dysmenorrhea
  - endometriosis

**Obstetrical Complications**
- Fetal Survival Rate 6-28%
- Premature Birth Rates 9-33%
- Spontaneous abortion rates 26-94%
- Length of septum does not correlate with observed obstetrical outcome
Class V: Septate Uterus

- Imaging
  - Ultrasound
    - Partial septa may be visualized in the intermediate aspect of the uterine cavity.
    - Complete septum demonstrates myometrial echogenicity in the fundal segment and hypoechoic echogenicity in the inferior segment in the fibrous caudal component.
    - External uterine contour may be convex, flat, or mildly concave (<1 cm).
    - 3D Ultrasound may be used to visualize the size and shape of the uterine cavity and the serosa.
  - MRI
    - MRI criteria for classification:
      - Fundus: Convex, flat, or minimally indented (depth <1 cm deep) Morphology of outer fundal contour is key to diagnosis.
      - Septum: Muscular, fibrous, or combined muscular and fibrous.
      - Complete septum to external os.
      - Partial incomplete septum that does not reach external os.
      - A short septum can be difficult to differentiate from arcuate uterus on MRI. In fact, there may be a continuum between these two entities.
      - Cervix divided by a septum.
      - Vagina: Vaginal septum may be present in some cases.
      - Partial or complete division of the endometrial canals by a solid mass.
      - Inferior segment of the complete septate uterus is composed of fibrous tissue so it appears as low-signal intensity linear band on T2 weighted images.

Class V- Septate Uterus

- Imaging
  - Ultrasound

Class V- Septate Uterus

- Imaging
  - MRI
Class V: Septate Uterus

- Management
  - May be treated surgically via hysteroscopic resection of the septum to improve obstetric outcome
  - Full-term delivery rate has been demonstrated to improve from 3% prior to hysteroscopic resection to 80% after procedure

Class VI: Arcuate Uterus

- Small Indentation of the fundal endometrial canal
- Normal external contour and no division of the uterine horns
- Some consider this to be a normal uterine variant
- Results from the near complete resorption of the uterovaginal septum
Symptoms

- Typically Asymptomatic

Obstetric Complications

- No Known impact on reproductive or obstetric outcomes

Imaging

- Ultrasound
  - Smooth indentation of the fundal endometrial canal that can best be appreciated in the transverse plane
  - No division of the uterine horns is seen
  - Normal external fundal contour is noted
- MRI
  - MRI Criteria for Classification
    - Fundus- Convex
    - Endometrial cavity- Short muscular saddlelike thickening of fundal myometrium that indents endometrial cavity
    - Cervix- Single
    - Myometrial fundal groove is broad and isointense relative to normal myometrium
Class VI- Arcuate Uterus

Management
- Hysteroscopic correction is rarely indicated

Class VII- Diethylstilbestrol-related Anomalies

- A 26-year-old woman was evaluated because of 3 years of primary infertility.
- HPI-
  - Her mother had been given 1.5 mg of DES daily for the first 6 months of pregnancy.
  - Menarche and secondary sexual characteristics occurred appropriately, but she experienced oligomenorrhea.
- PE
  - A hypoplastic cervix and corpus were noted.
- Imaging
  - Hysterosalpingogram highlighted a normal endocervical canal, abnormal uterine cavity (T-shaped cornual constriction bands, linear filling defect in lower segment), and bilateral tubal patency.
Class VII- Diethylstilbestrol-related Anomalies

- DES is a synthetic nonsteroidal estrogen that was given until 1971 to prevent miscarriage in women with prior miscarriage
- DES is associated with Vaginal Clear Cell Carcinoma in females with intrauterine exposure to this drug
- Structural anomalies of the uterine corpus, cervix and vagina have been described

Class VII- Diethylstilbestrol-related Anomalies

- Obstetrical Complications
  - spontaneous abortion
  - ectopic pregnancy
  - premature labor
  - increased risk for incompetent cervix during pregnancy also has been proposed

Class VII- Diethylstilbestrol-related Anomalies

- Imaging
  - MRI
    - MRI Criteria for Classification
      - Fundus- Convex
      - Endometrial cavity- Single T-shaped or hypoplastic with irregular margins
      - Cervix- Single
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<td>V</td>
<td>55</td>
<td>Septate uterus</td>
</tr>
<tr>
<td>V-A</td>
<td></td>
<td>Complete septate uterus (septum extends to the internal os)</td>
</tr>
<tr>
<td>V-B</td>
<td></td>
<td>Partial septate uterus (Septum does not reach the internal os)</td>
</tr>
<tr>
<td>VI</td>
<td>?</td>
<td>Arcuate uterus</td>
</tr>
<tr>
<td>VII</td>
<td>DES related uterine anomalies</td>
<td></td>
</tr>
<tr>
<td>VII-A</td>
<td></td>
<td>Iatrogenic cervicoisthmic stenosis</td>
</tr>
<tr>
<td>VII-B</td>
<td></td>
<td>Trigonal cervicoisthmic stenosis</td>
</tr>
<tr>
<td>VII-C</td>
<td></td>
<td>Uterine hyperplasia</td>
</tr>
</tbody>
</table>

References

- Dockery, Keith, M.D. Imaging Interesting Cases- Case263. [Link]