Pediatric and Adolescent Gynecology
UNIVERSITY OF COLORADO **ANSCHUTZ MEDICAL CAMPUS**

Children's Hospital Colorado

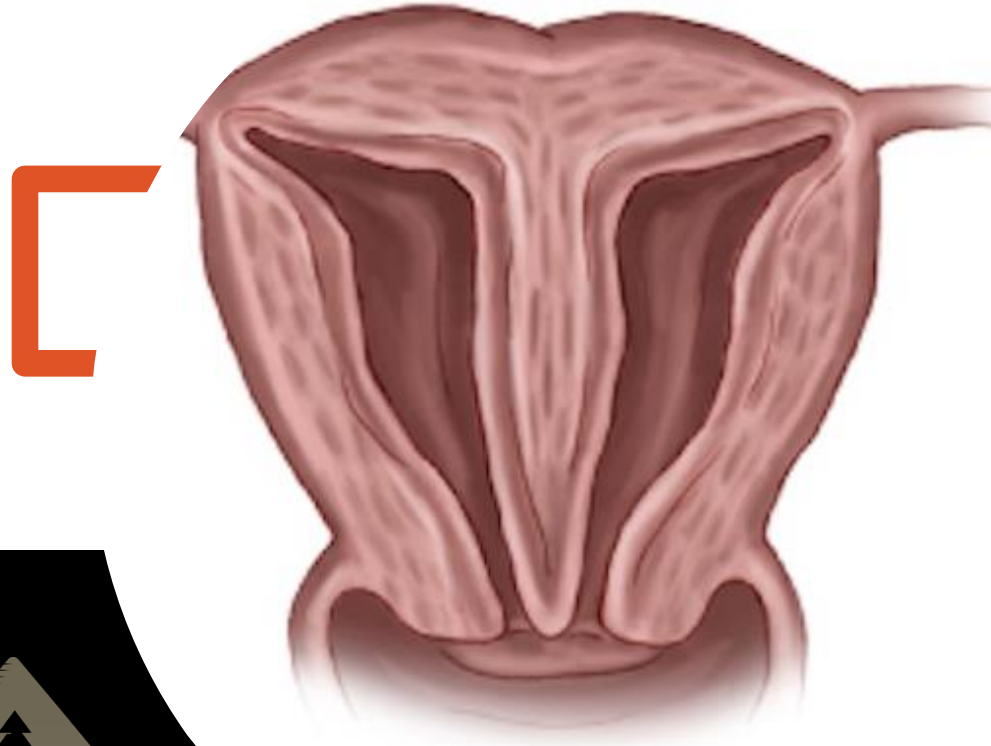
Mullerian Anomalies

Objectives (Vail)

- Use ASRM MAC 2021 to characterize mullerian anomalies
- Identify obstructive and non obstructive uterine and vaginal anomalies
- Recommend treatment options for vaginal and uterine anomalies

Background

- Mullerian duct anomalies are present in up to 7% of women.
- More common with other congenital anomalies.
- US is the first line imaging modality but MRI is the gold standard for evaluating Mullerian duct anomalies.
- Mullerian anomalies are now classified by descriptive terminology
  - ASRM MAC 2021.



Saravelos SH, Cocksedge KA, Li TC: Prevalence and diagnosis of congenital uterine anomalies in women with reproductive failure: a critical appraisal. *Hum Reprod Update* 2008, 14(5):415-429.

Embryology Review

1. Organogenesis (7 wk GA)

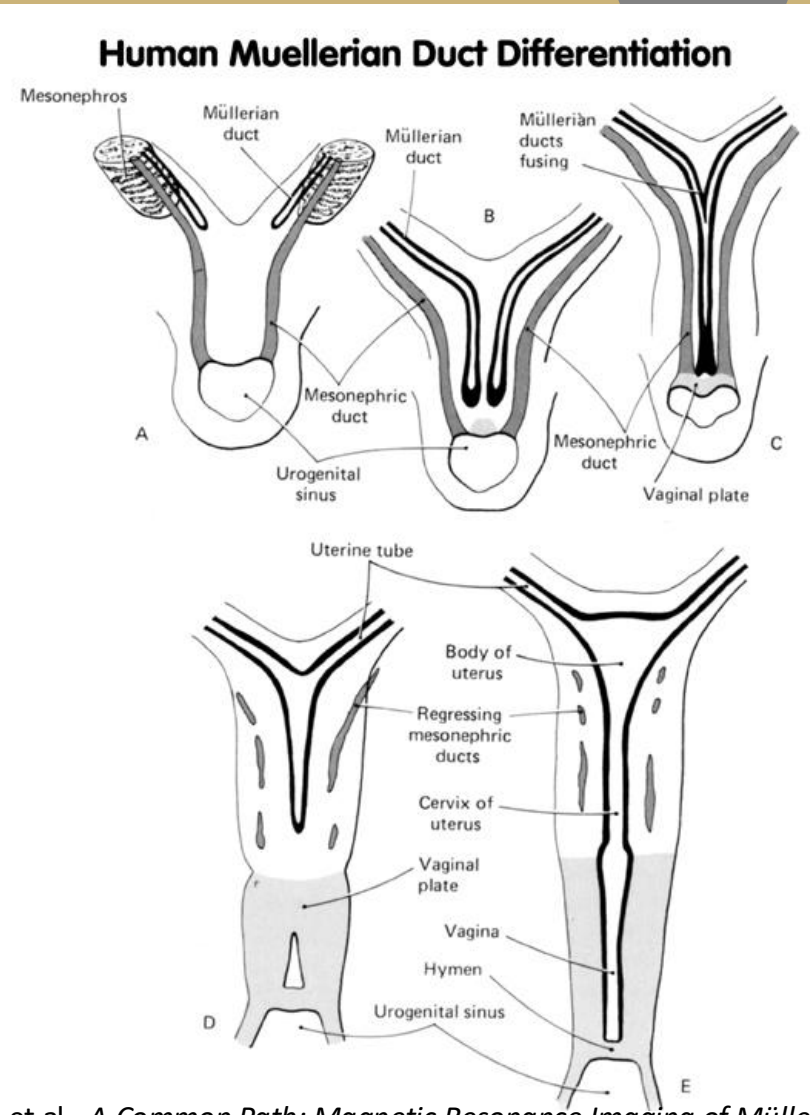
- Bilateral Mullerian (paramesonephric) ducts elongate

2. Fusion (8-12 wk GA)

- Cephalic portions of the Müllerian ducts develop into the fallopian tubes
- Caudal portions fuse to form the uterus and upper vagina
- Mullerian ducts fuse with urogenital sinus

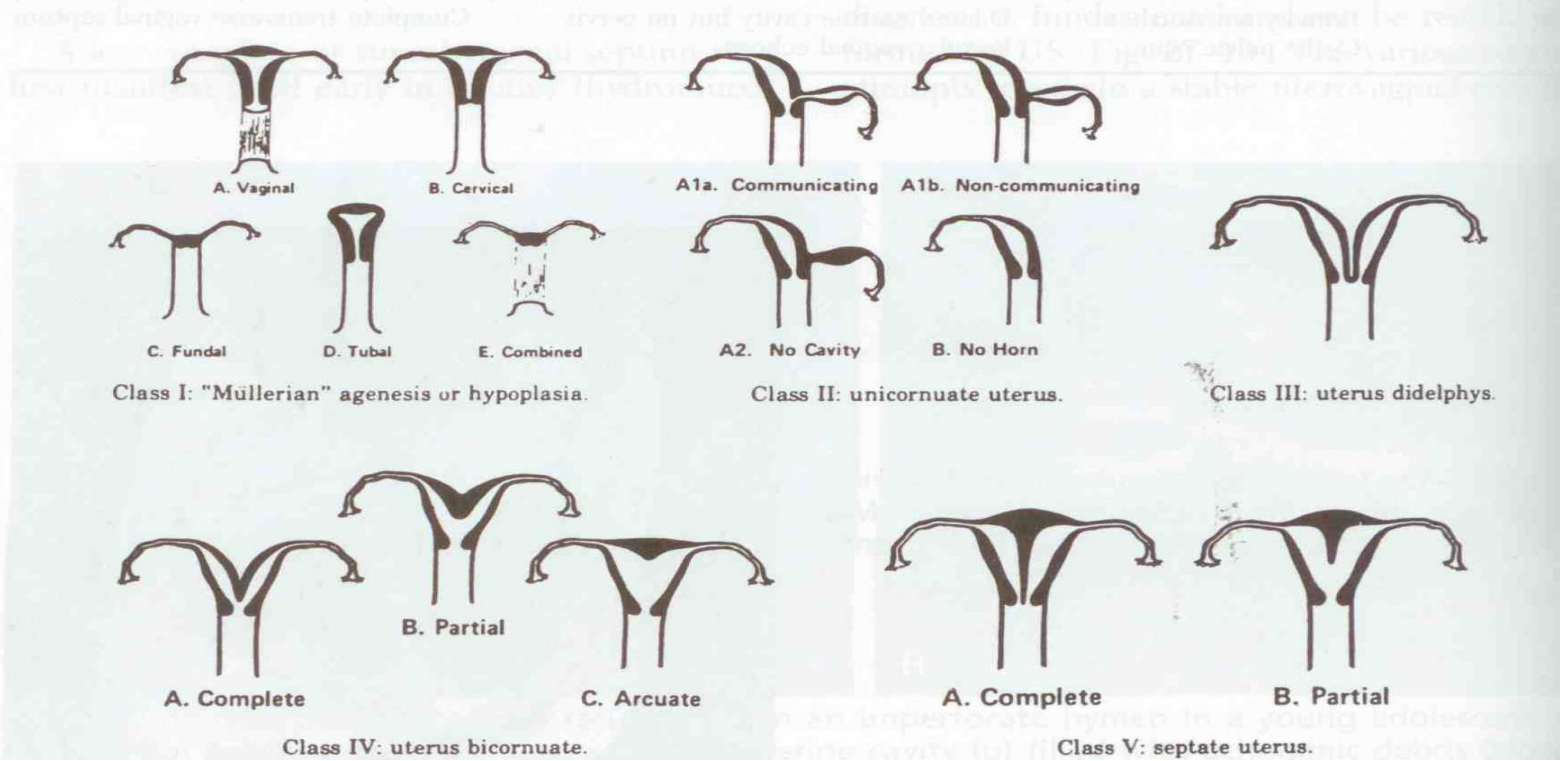
3. Canalization (20 wk GA)

- Midline septum of the developing uterus is resorbed by cellular apoptosis
- Uterovaginal plate canalizes to form the lumen of the vagina



Udayakumar, N., et al., *A Common Path: Magnetic Resonance Imaging of Müllerian and Wolffian Duct Anomalies.* Curr Urol Rep, 2023. **24**(1): p. 1-9.

Old Classification



Buttram, V.C., Jr. and W.E. Gibbons, *Müllerian anomalies: a proposed classification. (An analysis of 144 cases).* Fertil Steril, 1979. **32**(1): p. 40-6.

American Society for Reproductive Medicine: Mullerian Anomaly Classification 2021

- Mullerian anomalies are identified by descriptive terminology

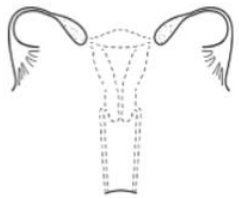
  - Mullerian agenesis
  - Cervical agenesis
  - Unicornuate uterus
  - Uterus didelphys
  - Bicornuate uterus
  - Septate uterus
  - Longitudinal vaginal septum
  - Transverse vaginal septum
  - Complex anomalies

Pfeifer, S.M., et al., *ASRM müllerian anomalies classification 2021.* Fertil Steril, 2021. **116**(5): p. 1238-1252.

ASRM MÜLLERIAN ANOMALIES CLASSIFICATION 2021

Scan QR code to view the ASRM MAC 2021 tool (page 1 of 2)

MÜLLERIAN AGENESIS

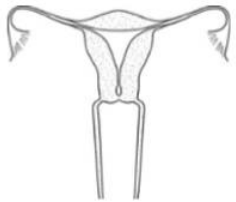


MÜLLERIAN AGENESIS

MÜLLERIAN AGENESIS WITH R/L ATROPHIC UTERINE REMNANT WITH FUNCTIONAL ENDOMETRIUM

CERVICAL AGENESIS

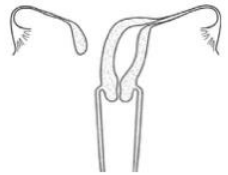
CERVICAL AGENESIS



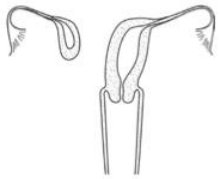
DISTAL CERVICAL AGENESIS

UNICORNUATE UTERUS

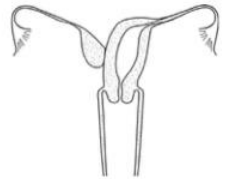
R/L UNICORNUATE UTERUS



R/L UNICORNUATE WITH R/L DISTAL ATROPHIC UTERINE REMNANT



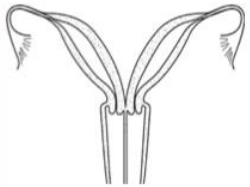
R/L UNICORNUATE WITH R/L DISTAL UTERINE REMNANT WITH FUNCTIONAL ENDOMETRIUM



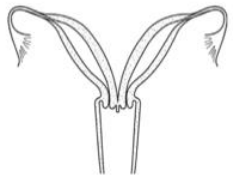
R/L UNICORNUATE WITH R/L ASSOCIATED ATROPHIC UTERINE REMNANT

R/L UNICORNUATE WITH R/L UTERINE HORN COMMUNICATING AT LEVEL OF CERVIX

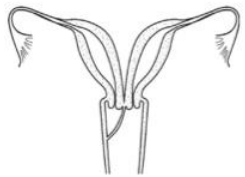
UTERUS DIDELPHYS



UTERUS DIDEPHYS AND LONGITUDINAL SEPTUM

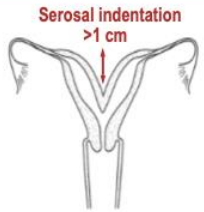


UTERUS DIDELPHYS AND +/- LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH

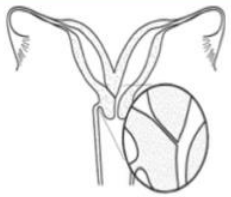


UTERUS DIDELPHYS AND OBSTRUCTED R/L HEMIVAGINA

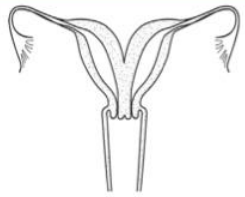
BICORNUATE UTERUS



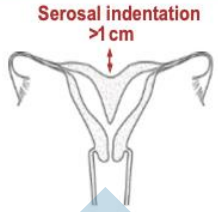
BICORNUATE UTERUS



BICORNUATE UTERUS WITH R/L COMMUNICATING TRACT

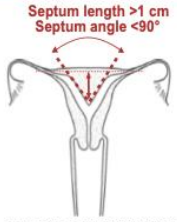


UTERUS BICORNUATE BICOLLIS

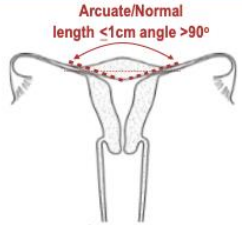


COMBINED BICORNUATE SEPTATE UTERUS

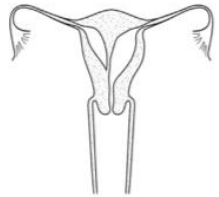
SEPTATE UTERUS



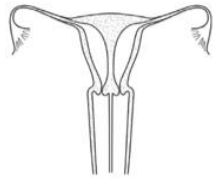
PARTIAL SEPTATE UTERUS



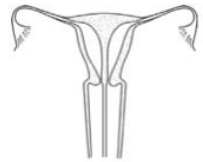
NORMAL/ARCUATE UTERUS



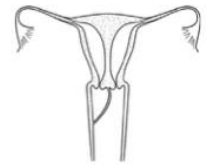
ROBERT'S UTERUS



COMPLETE SEPTATE UTERUS WITH DUPLICATED CERVICES AND LONGITUDINAL VAGINAL SEPTUM



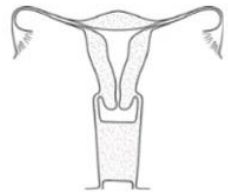
COMPLETE SEPTATE UTERUS WITH SEPTATE CERVIX AND LONGITUDINAL VAGINAL SEPTUM



COMPLETE SEPTATE UTERUS, DUPLICATED CERVICES, AND OBSTRUCTED R/L HEMIVAGINA

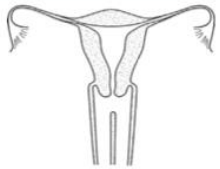
TRANSVERSE VAGINAL SEPTUM

MID VAGINAL SEPTUM

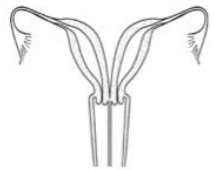


DISTAL VAGINAL AGENESIS

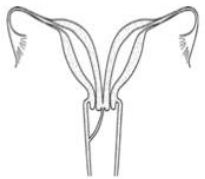
LONGITUDINAL VAGINAL SEPTUM



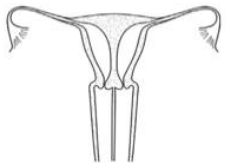
LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH



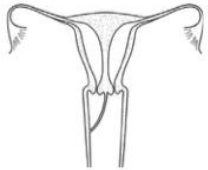
LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH AND UTERUS DIDELPHYS



OBSTRUCTED R/L HEMIVAGINA AND UTERUS DIDELPHYS

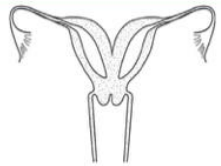


LONGITUDINAL VAGINAL SEPTUM OF VARIABLE LENGTH AND COMPLETE SEPTATE UTERUS WITH DUPLICATED CERVIX

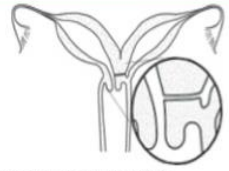


OBSTRUCTED R/L HEMIVAGINA AND COMPLETE SEPTATE UTERUS WITH DUPLICATED CERVICES

COMPLEX ANOMALIES

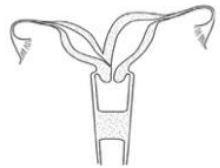


BICORNUATE UTERUS WITH BILATERAL OBSTRUCTED ENDOMETRIAL CAVITIES

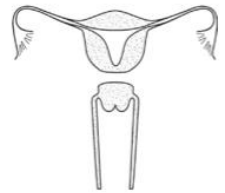


UTERUS DIDELPHYS WITH COMMUNICATING HEMIUTERI AND UNILATERAL R/L CERVICO-VAGINAL ATRESIA

OBSTRUCTED R/L HEMIVAGINA, HEMIUTERUS AND SINGLE CERVIX WITH SEPARATE CONTRALATERAL R/L PATENT HEMIUTERUS, CERVIX AND VAGINA

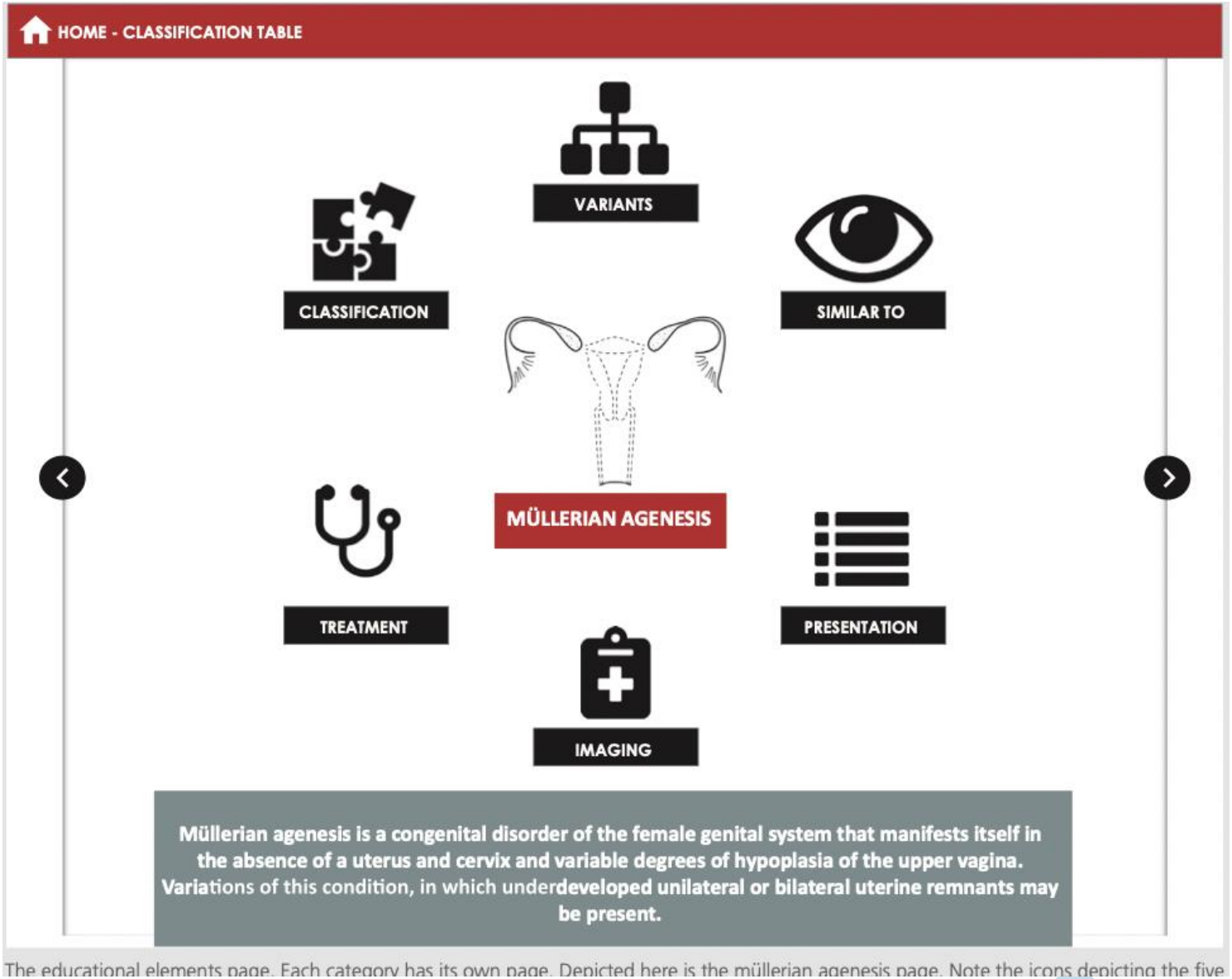


BICORNUATE UTERUS WITH R/L COMMUNICATING TRACT AND TRANSVERSE VAGINAL SEPTUM



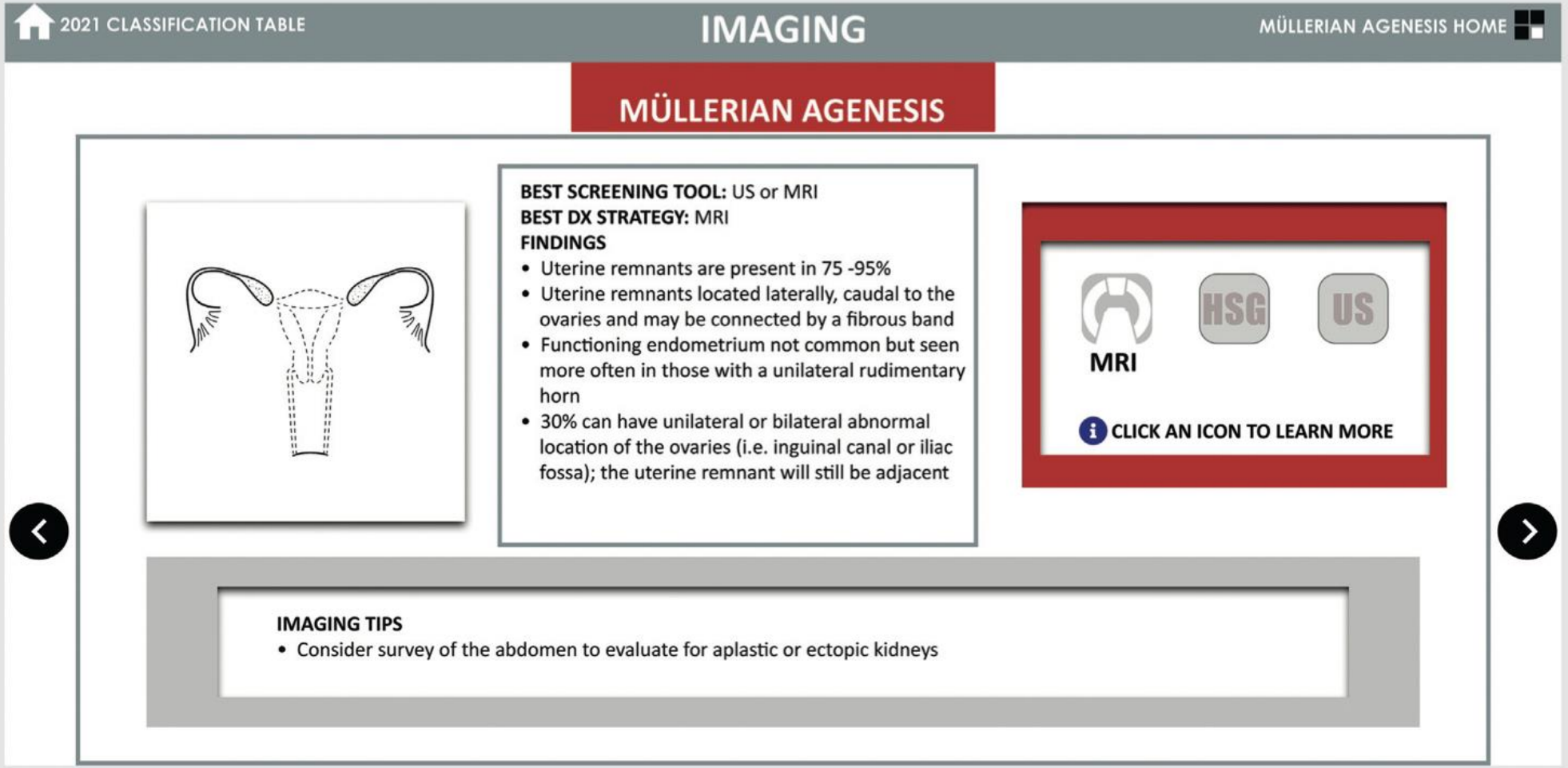
UTERUS ISTHMUS AGENESIS

Scan QR code to view the ASRM MAC 2021 tool (page 2 of 2)



The educational elements page. Each category has its own page. Depicted here is the müllerian agenesis page. Note the icons depicting the five

SYMPTOMS	EXAMINATION	DIFFERENTIAL
• Pubertal female with primary amenorrhea • Primary amenorrhea with cyclic pelvic - Pain suggests presence of - Rudimentary uterine horn(s) with - Functional endometrium • Primary amenorrhea with chronic - Pelvic pain from retrograde - Menstruation	• Vaginal canal shortened (dimple) or several centimeters long • No palpable uterus or cervix on vaginal or rectal exam • Rudimentary uterine horn(s) may be present on imaging	• Must differentiate from: - Androgen insensitivity syndrome - Distal vaginal agenesis - Transverse vaginal septum - Imperforate hymen - Cervical agenesis • Evaluate for renal anomalies.



IMAGING section shows best screening tool, diagnostic strategy, and typical findings. Clicking on the MRI icon opens pictures of pertinent images and comparative normal images for educational purposes and to help comparative diagnosis.

Pfeifer. Müllerian anomalies classification tool. Fertil Steril 2021.

Vaginal Anomalies

Obstructive Anomalies

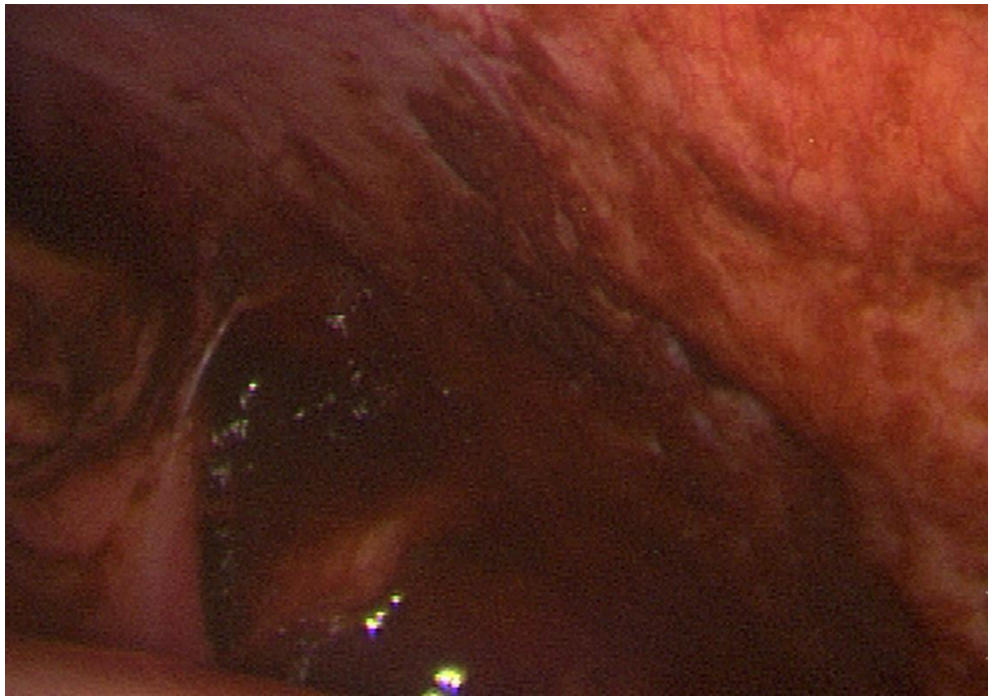
- Imperforate hymen
- Vagina
  - Distal vaginal agenesis
  - Septum
    - Duplication with obstruction
    - Transverse

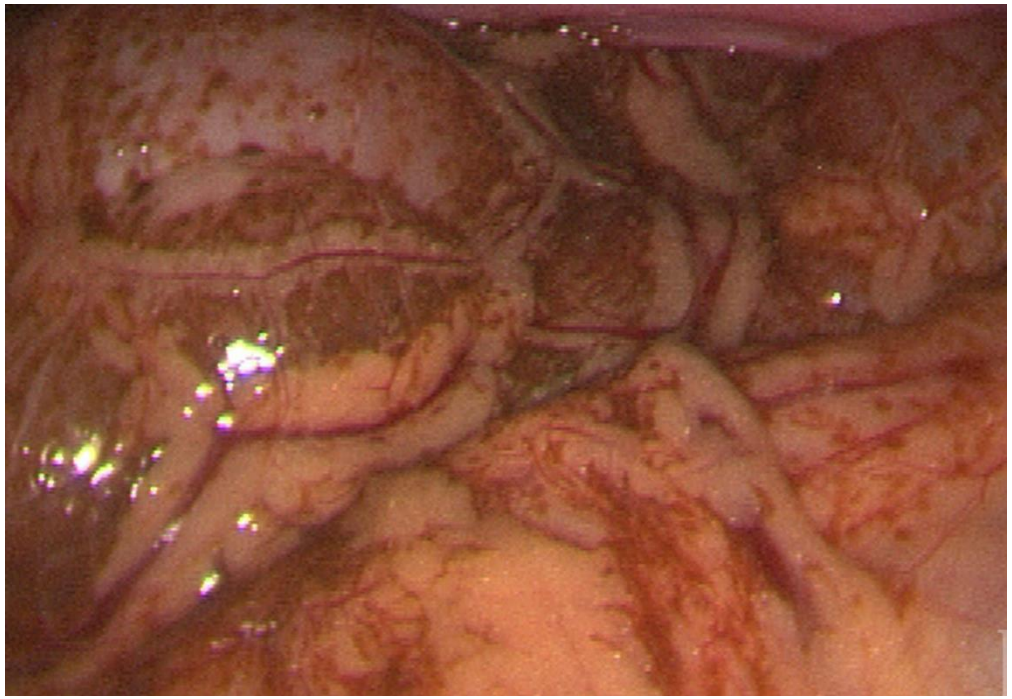
Non Obstructive Anomalies

- Hymenal anomalies
  - Vagina
    - Mullerian agenesis - MRKH
    - Septum
      - Longitudinal
      - Perforated transverse

Obstructive Anomalies

- Cyclic abdominal pain
- Abdominal mass
- Endometriosis



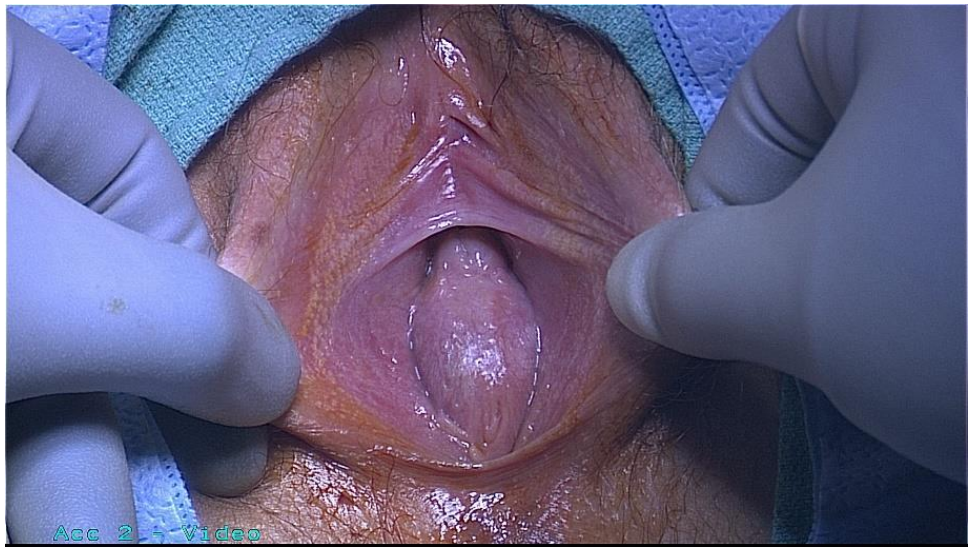


Imperforate hymen

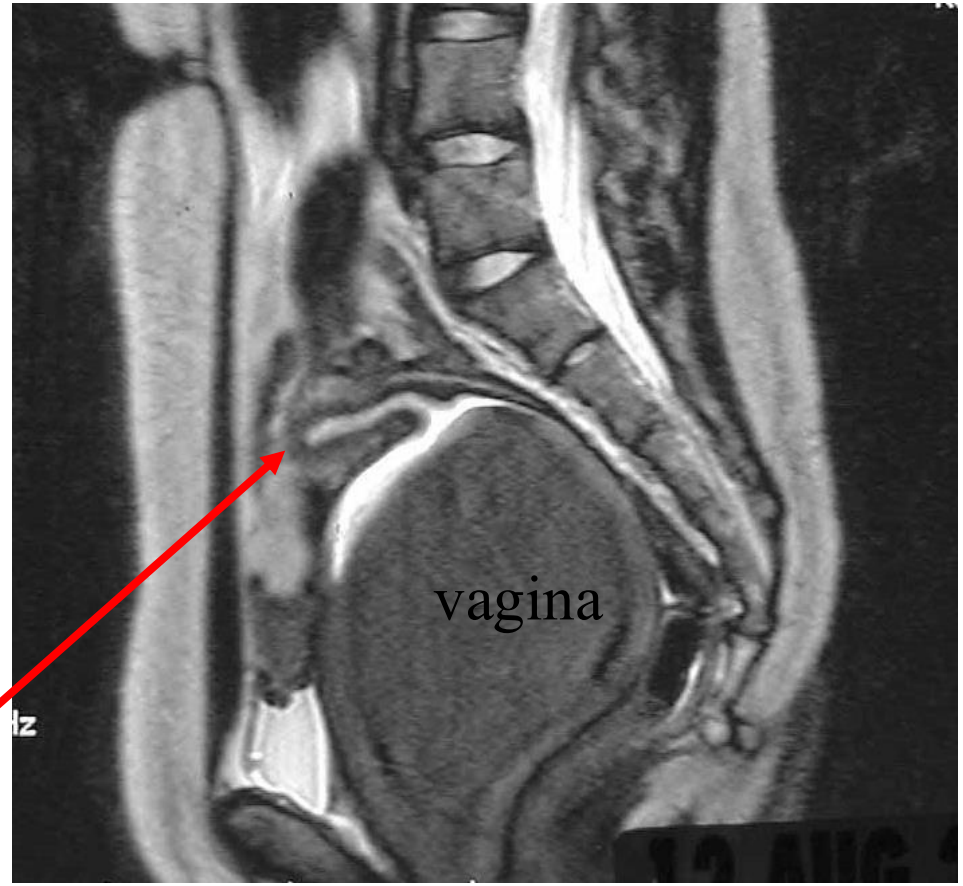
- Presentation: primary amenorrhea, pelvic pain, back pain, urinary retention

- Diagnosis: physical exam and inspection is usually all you need

Imperforate Hymen

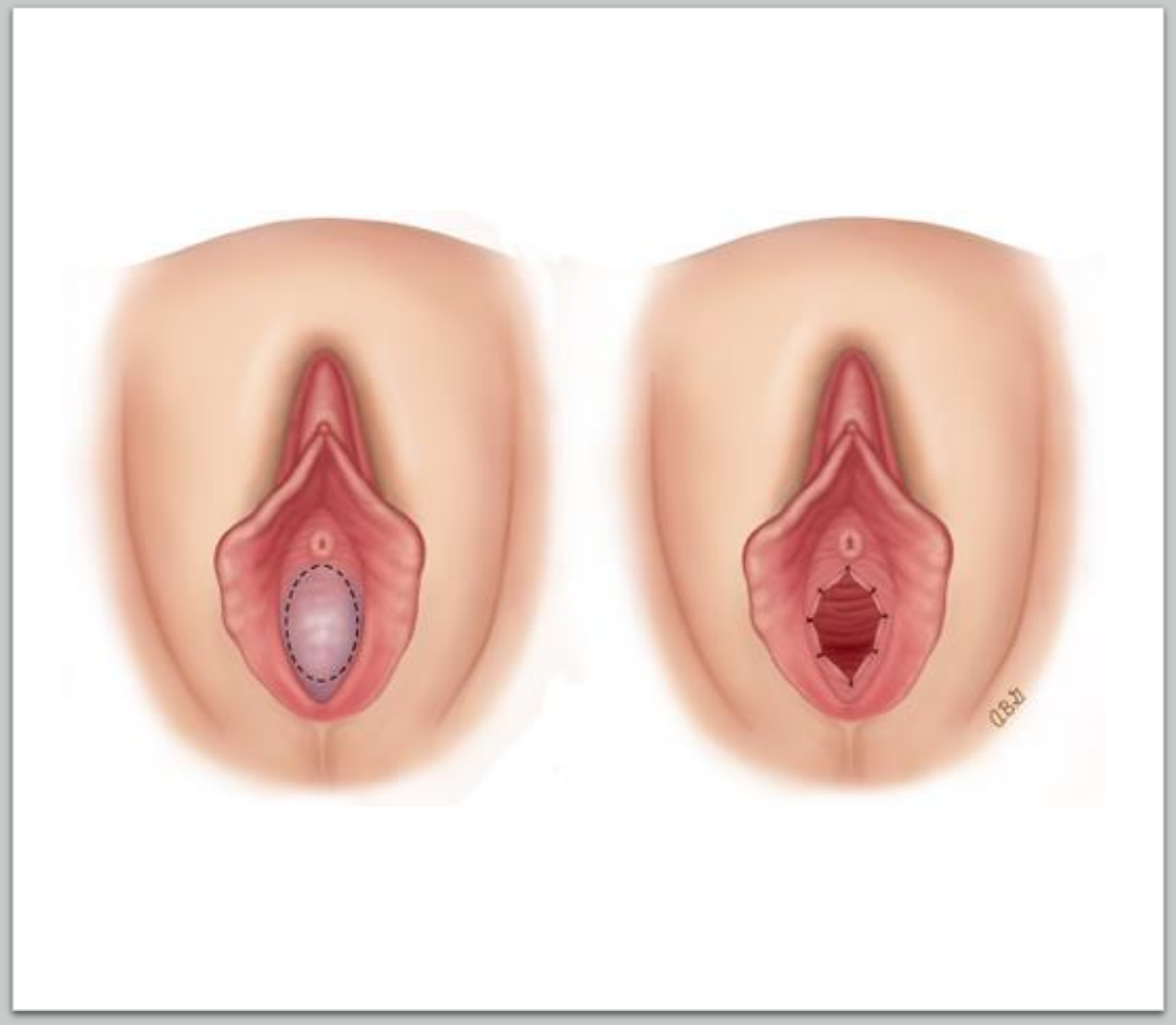


MRI



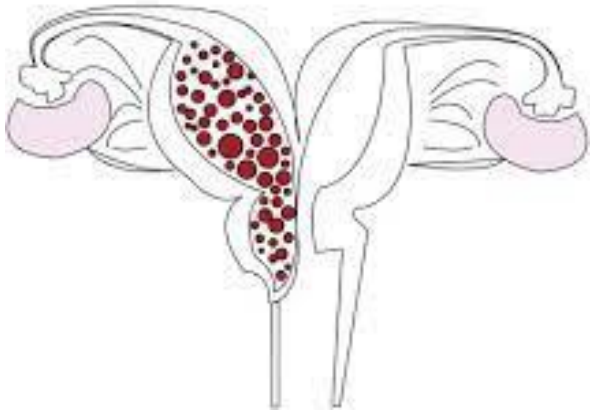
Treatment of Imperforate Hymen

- Suppression if OR is not available
- Hymenectomy
  - Resect fibrous tissue if present



Obstructed Hemivagina and Ipsilateral Renal Anomalies (OHVIRA)

- Uterine Duplication with Oblique transverse vaginal septum
- Over 90% have solitary /dysplastic kidney at the obstructed side



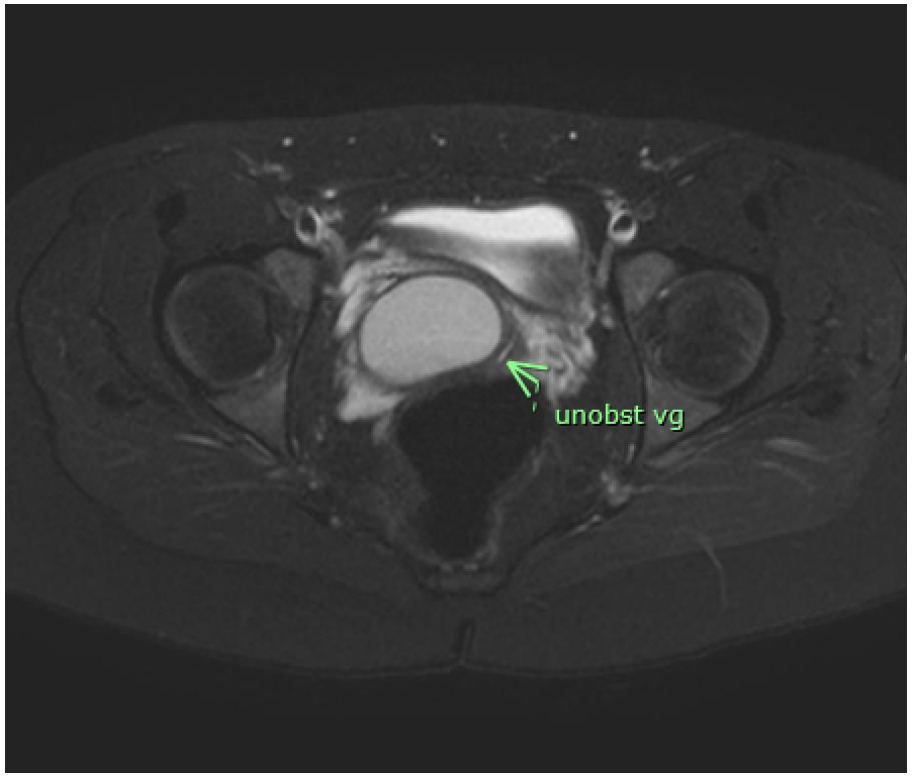
Obstructed Hemi Vagina

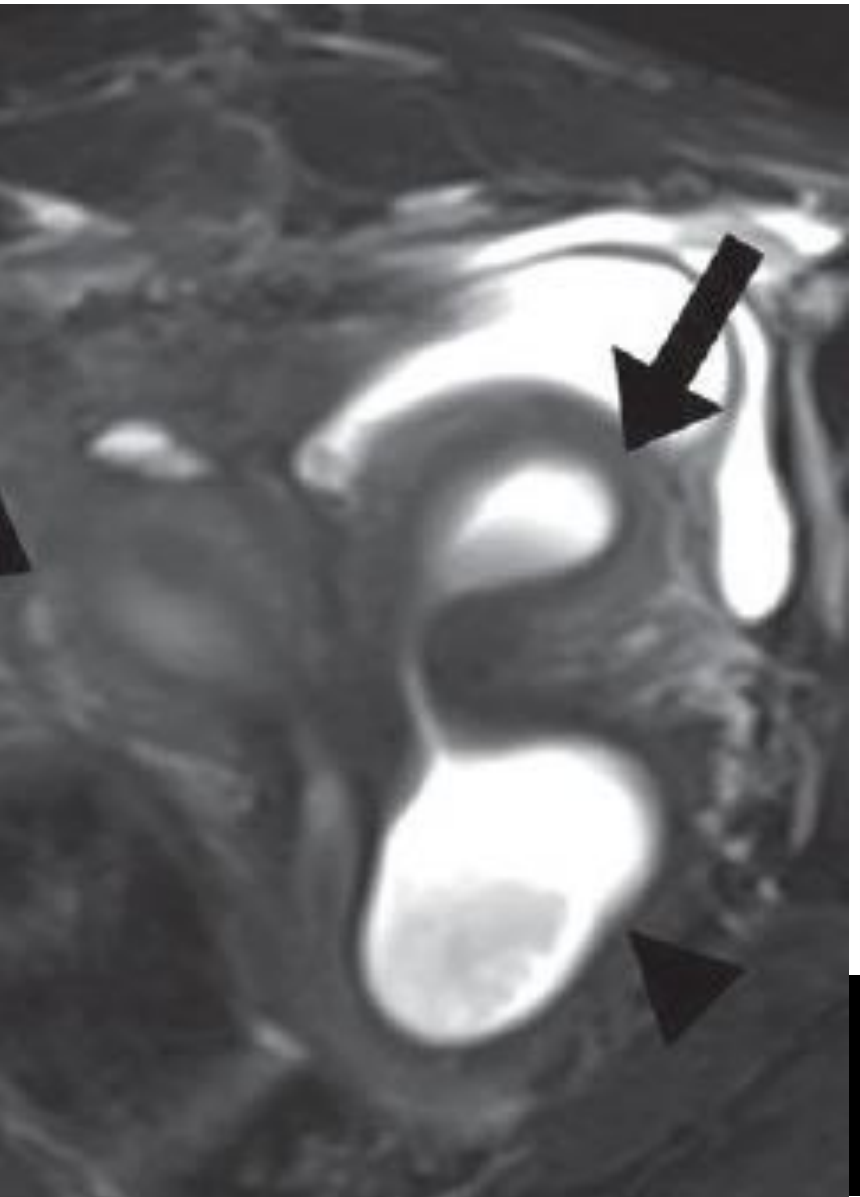
Key MRI Findings

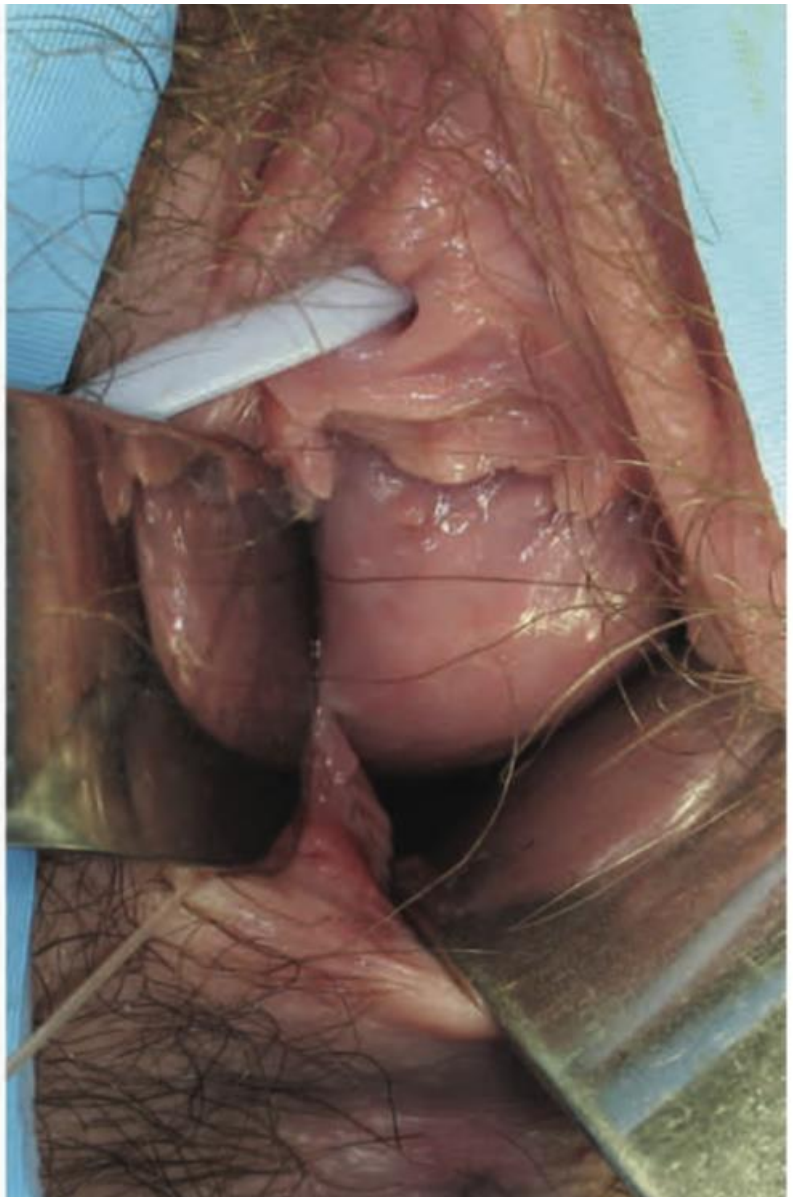
- Distended hemi-vagina with T1 hyper-intense blood products
- Cervix may or may not be dilated
- Length of the upper vagina can be measured on sagittal view
- Ipsilateral Renal Anomaly

Rivas, A.G., et al., *Magnetic resonance imaging of Müllerian anomalies in girls:*

OHVIRA MRI







Treatment for Obstructed Hemi Vagina

- Suppression
- Surgery:
  - Incise the septum at a low point
  - Drain of old blood (can be very thick)
  - Excise the whole septum (usually in pieces) up to the second cervix

High OHVIRA versus Cervicovaginal Dysgenesis

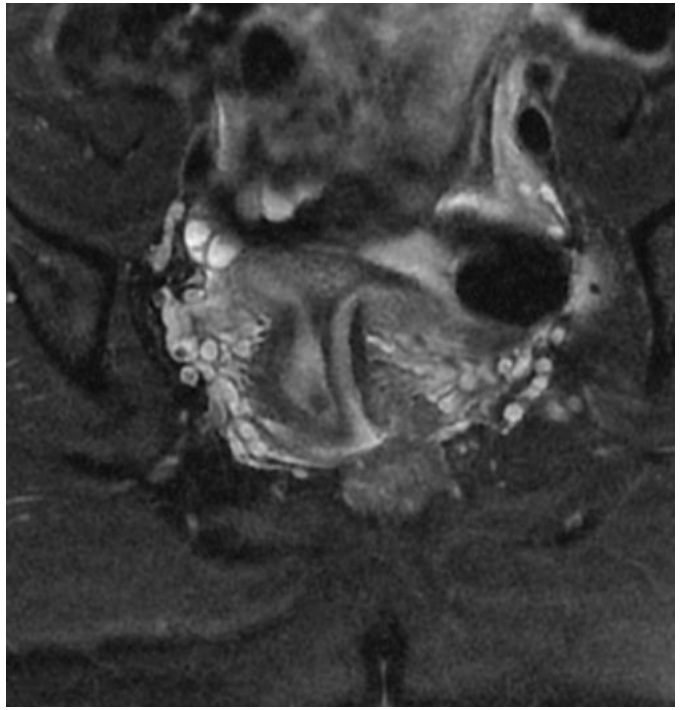
- Be cautious/suspicious of a “High OHVIRA”
- Resection of a high obstruction may be associated with increased risk of vaginal stenosis
  - Consider placing stent or post op vaginal dilation
- Consider diagnosis of Uterine didelphys with unilateral CVAD (cervicovaginal agenesis/dysgenesis)

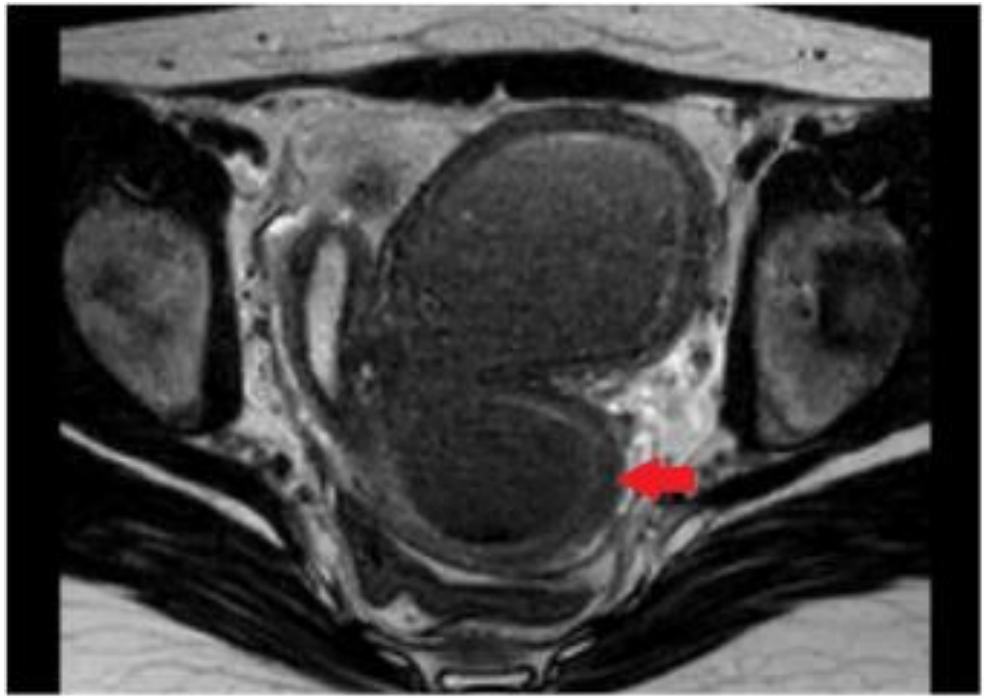
Uterine Didelphys with Unilateral CVAD

- Incidence unknown
- Unilateral cervix is dysplastic or hypoplastic.
- Different from uterine remnant which is a smaller underdeveloped piece of uterus
- Can mimic an OHVIRA
- Hemi hysterectomy is recommended for management

Moon et al. Presentation and Management of Uterine Didelphys with Unilateral Cervicovaginal Agenesis/Dysgenesis (CVAD): A Multicenter Case Series. JPAG 37 (10): Feb 2024.

CVAD MRIs

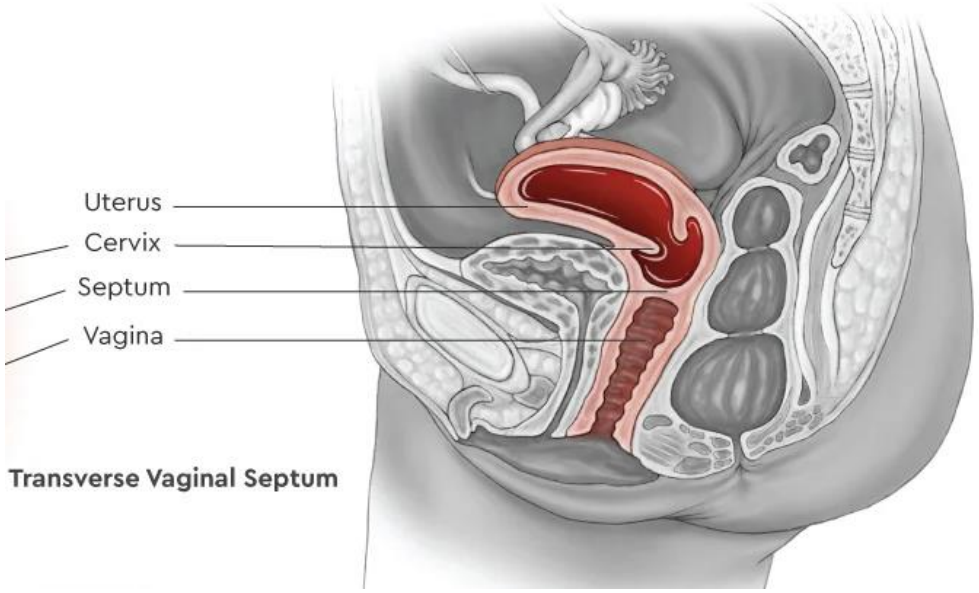




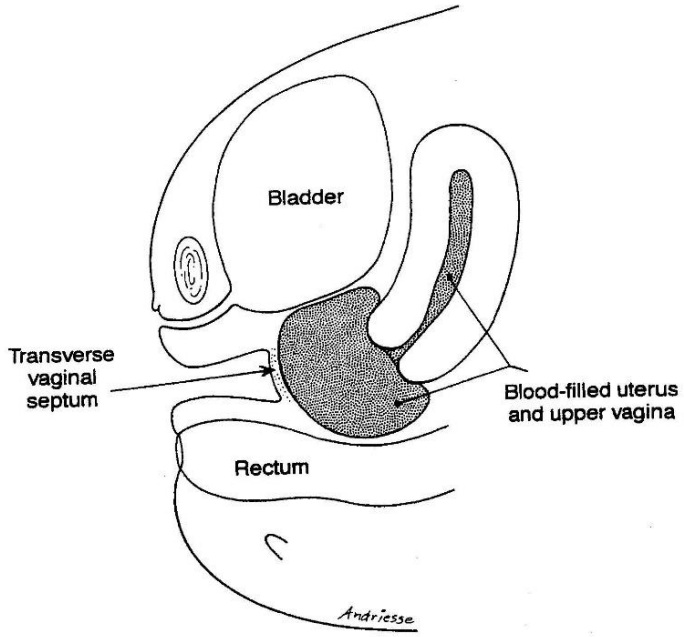
Transverse Vaginal Septum

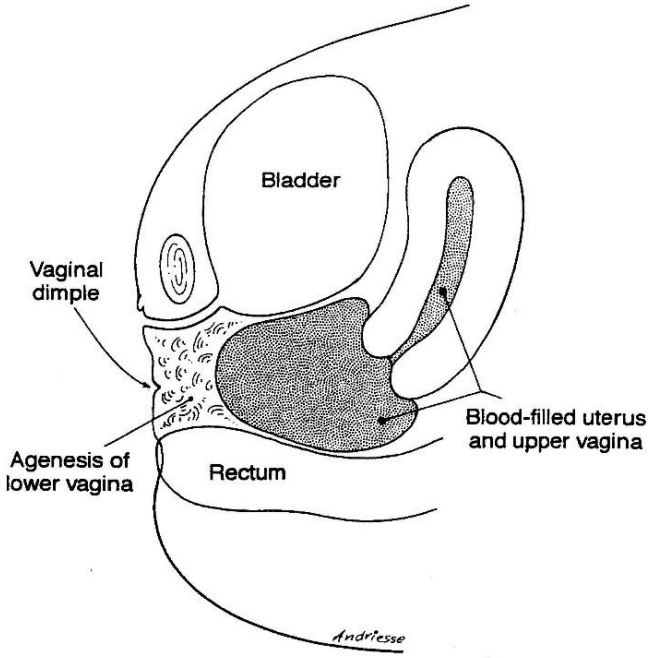
- Results from failed fusion or canalization of the vaginal plate and caudal end of the mullerian ducts
  - Vaginal plate is squamous epithelium. When it proliferates caudally, it replaces the mullerian epithelium

- Characteristics
  - Position: low, mid, or high
  - Thickness: considered thin if < 1 cm
  - Presence of perforations
    - Most commonly imperforate



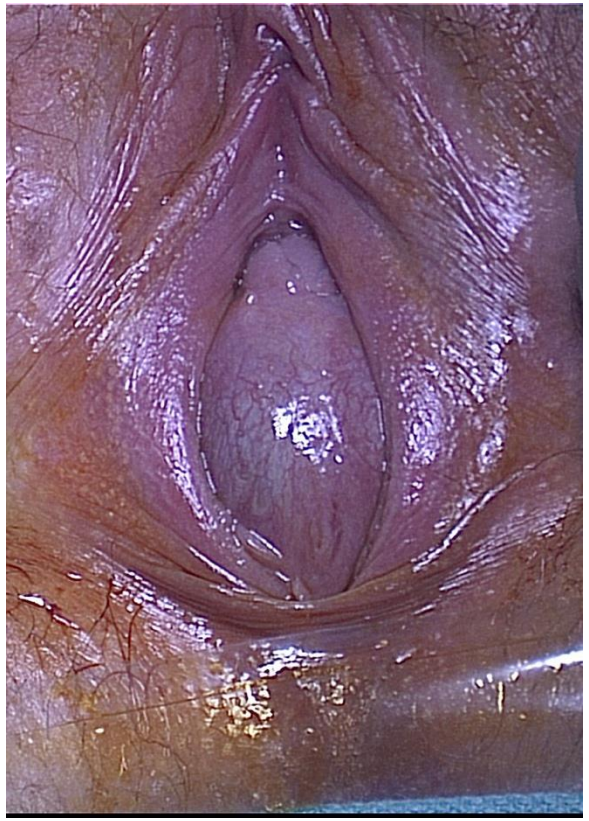
Transverse Vaginal Septum vs Vaginal Agenesis

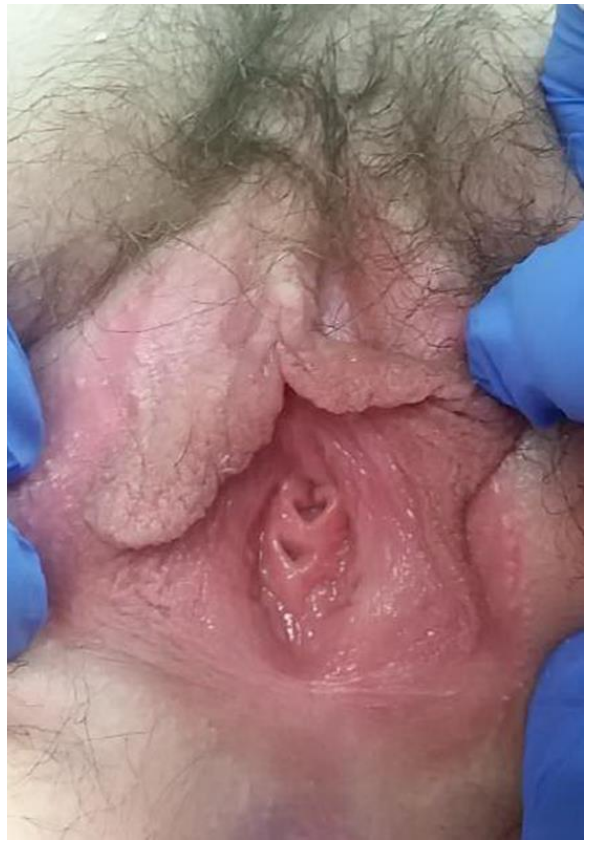




Obstructive Anomalies: Imaging

Obstructive Anomalies: Exam





Transverse Vaginal Septum

- Management options
  - Primary resection and anastamosis
  - Drainage and suppression of menstruation with delayed surgery
- With surgical management:
  - Resect the whole septum and use interrupted sutures
  - Assess vaginal caliber at the end
  - If very thick septum, may need mobilization of upper vagina
  - Always postop dilatation
    - Consideration for age of adolescent

Ultrasound Guided Drainage

- Typically performed by Interventional Radiology
- Percutaneous approach preferred
  - Vagina versus uterus
- Tissue plasminogen activator (TPA) to break up clot

Childress KJ, Williams AK, Somasundaram A, Alaniz VI, Chan SH, Gill AE. Management of Hematometra and Hematocolpos in Obstructive Mullerian Anomalies by Image-Guided Drainage with Interventional Radiology: A Multi-Institutional Case Series. J Pediatr Adolesc Gynecol. 2023 Aug;36(4):372-382. doi: 10.1016/j.jpag.2023.02.006. Epub 2023 Mar 4. PMID: 36878355.

Y-plasty

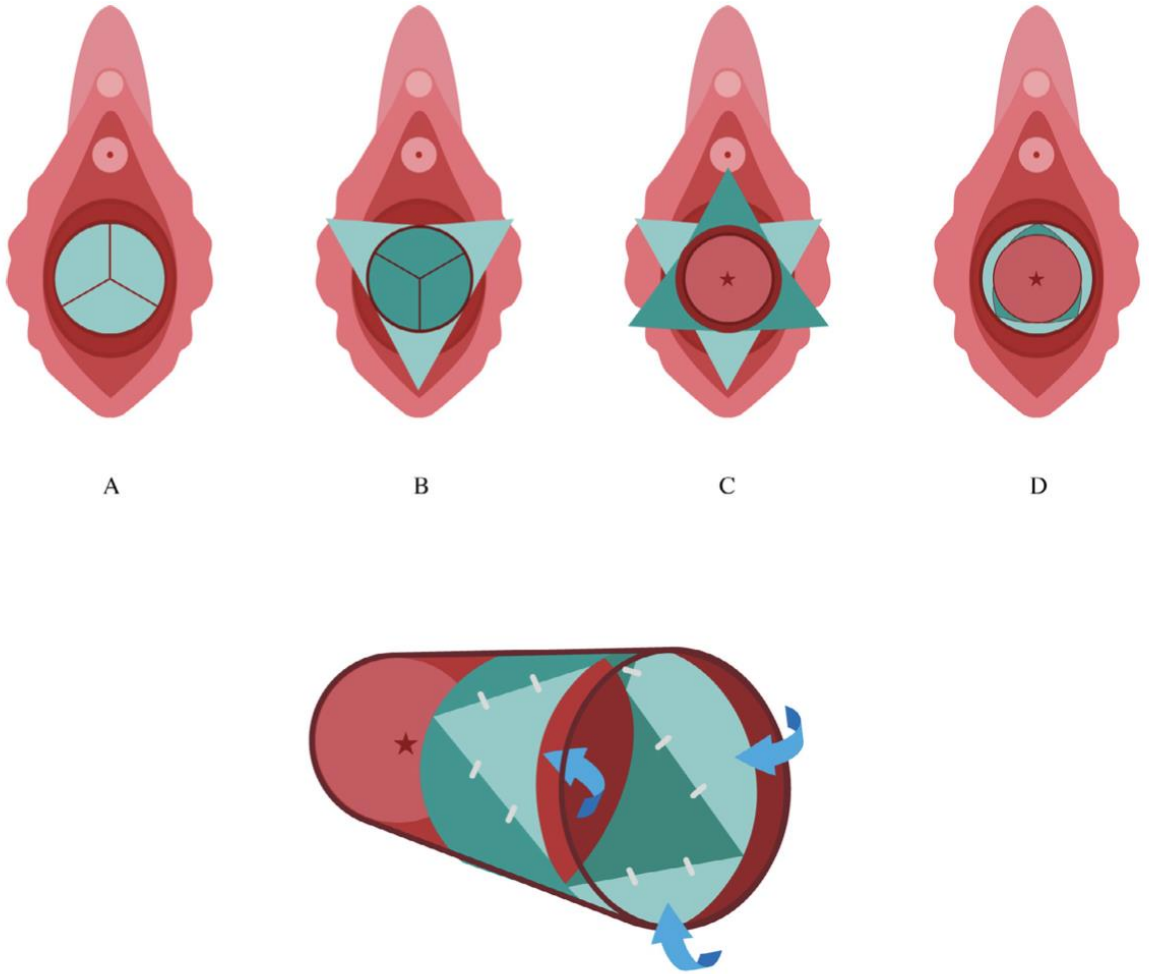
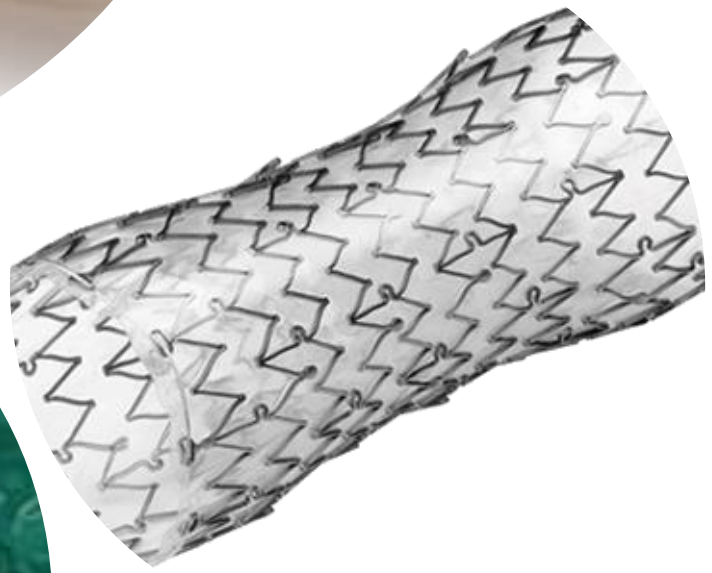
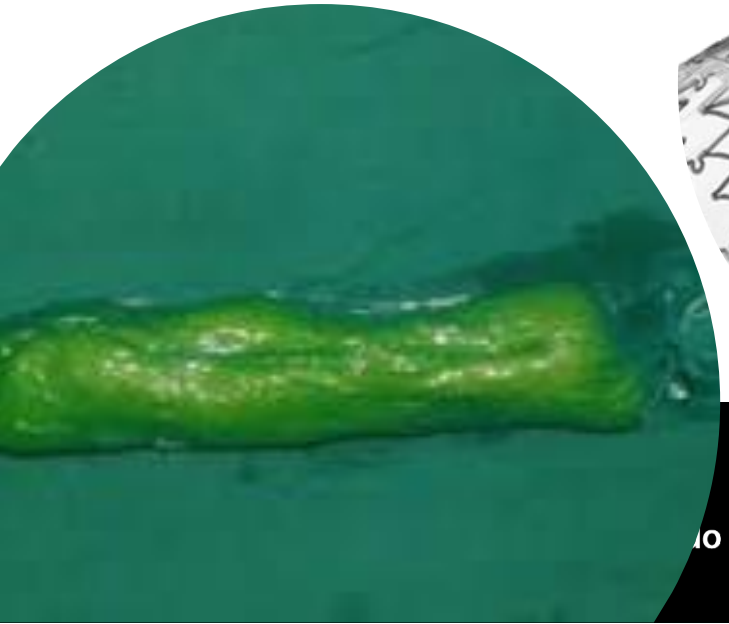


Fig. 2. Illustration of vaginal Y-plasty. Transverse vaginal septae are made up of 2 leaflets of epithelial tissue with fibroareolar tissue in between. The distal/lower epithelial layer is incised in the shape of a "Y" to the level of the fibroareolar tissue creating 3 triangular shaped flaps (*A*). These are held outward via stay sutures (*B*). The fibrous tissue between the layers is carefully dissected off the proximal/upper epithelial layer. This inner layer is incised similarly to the first, but the "Y" is rotated 180°. This will release the hematocolpos. These flaps are also held outward with stay sutures (*C*). The 6 triangular flaps are interposed and sutured with interrupted, absorbable suture, such that the apex of each triangular flap from 1 layer lies between the bases of the flaps from the other layer (*D*, *E*).

Transverse vaginal septum

- Surgical Complications
  - Stenosis and reobstruction
  - Injury to bowel/bladder
- Impact on Fertility and Reproduction
  - Endometriosis
  - Tubal dysfunction if hematosalpinx present
  - Dyspareunia
  - Limited outcome data on pregnancy





Post op management

- There are no clear guidelines using stents or dilators after vaginal surgery
- A variety of medical devices are used and described in the literature for the prevention of acquired vaginal stenosis.
- Custom mold or stents are not readily available and have to be individually created for each case.

Non Obstructive Anomalies

- Much more variable presentation
  - Incidental finding
  - Amenorrhea
  - Dyspareunia
  - Difficulty using tampons
  - Fertility and pregnancy complications

Mullerian agenesis

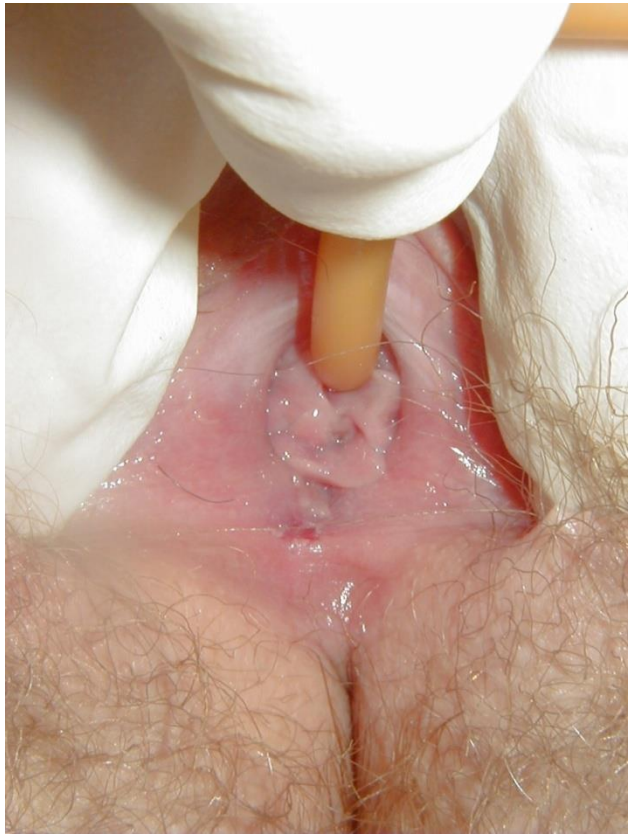
- Mayer-Rokitansky-Kuster-Hauser/ vaginal agenesis
- Incidence 1:4000-5,000
- Presentation: primary amenorrhea with normal puberty
- Associated anomalies
  - Kidney anomalies (overall 30%, renal agenesis most common)
  - Spine anomalies (scoliosis most common)

ACOG CO 562: Obstet Gynecol. 2013 May;121(5):1134-7

Physical Exam

- Normal pubic hair
- Normal hymen
- Rectal: no masses
- Q tip: minimal vaginal depth

Mullerian Agenesis versus Imperforate Hymen



Mullerian Agenesis

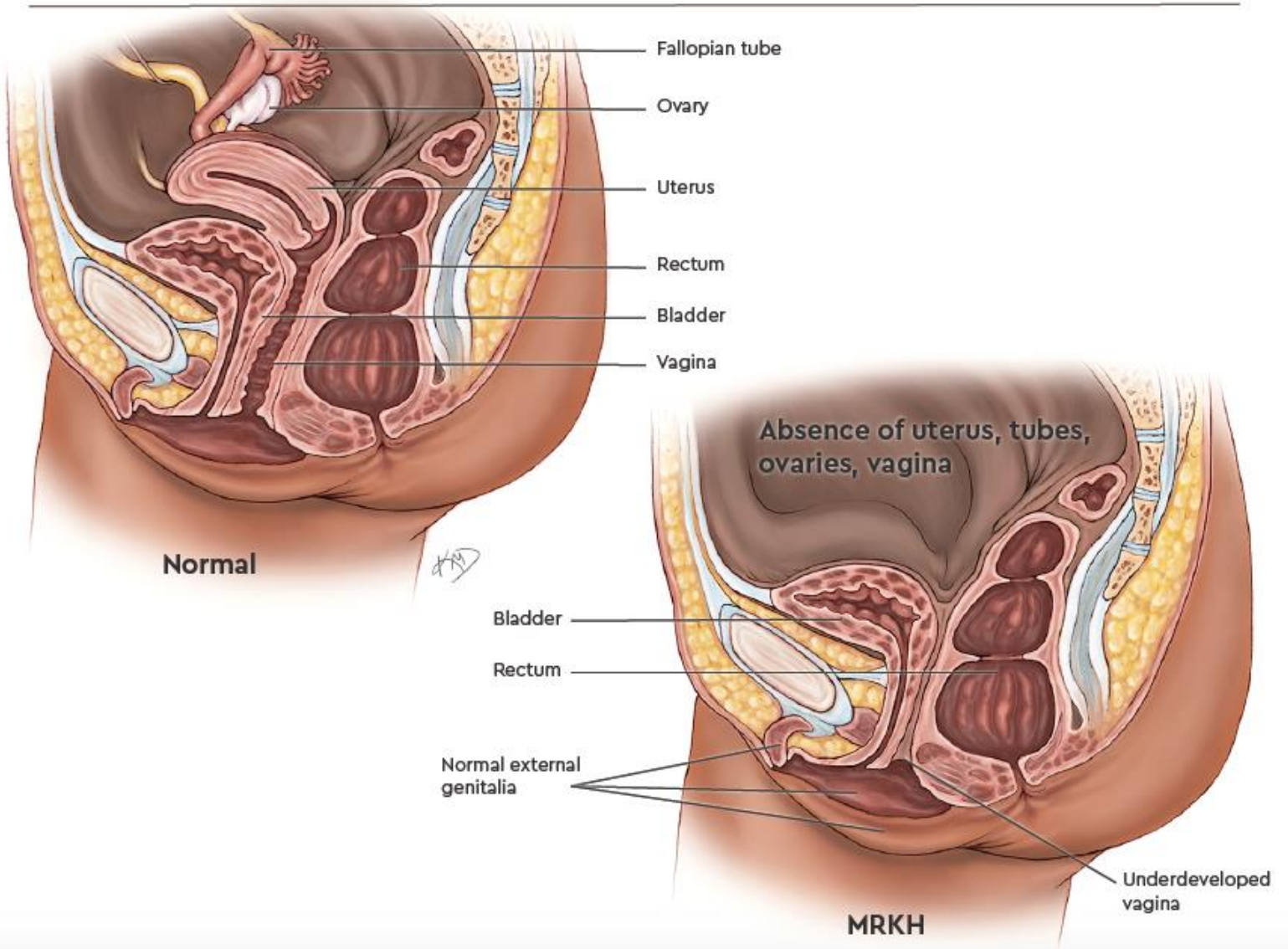
- Work up
  - Primary amenorrhea labs
  - Pelvic imaging
    - Screening ultrasound
    - Confirmation MRI
  - Testosterone OR Chromosomes

Mullerian Agenesis vs CAIS

	Mullerian Agenesis	CAIS
Vaginal agenesis	+	+
Pubic hair	Normal	scant
Testosterone	female	male
Chromosomes	46 XX	46 XY
Uterine remnants	possible	No

MRKH Syndrome

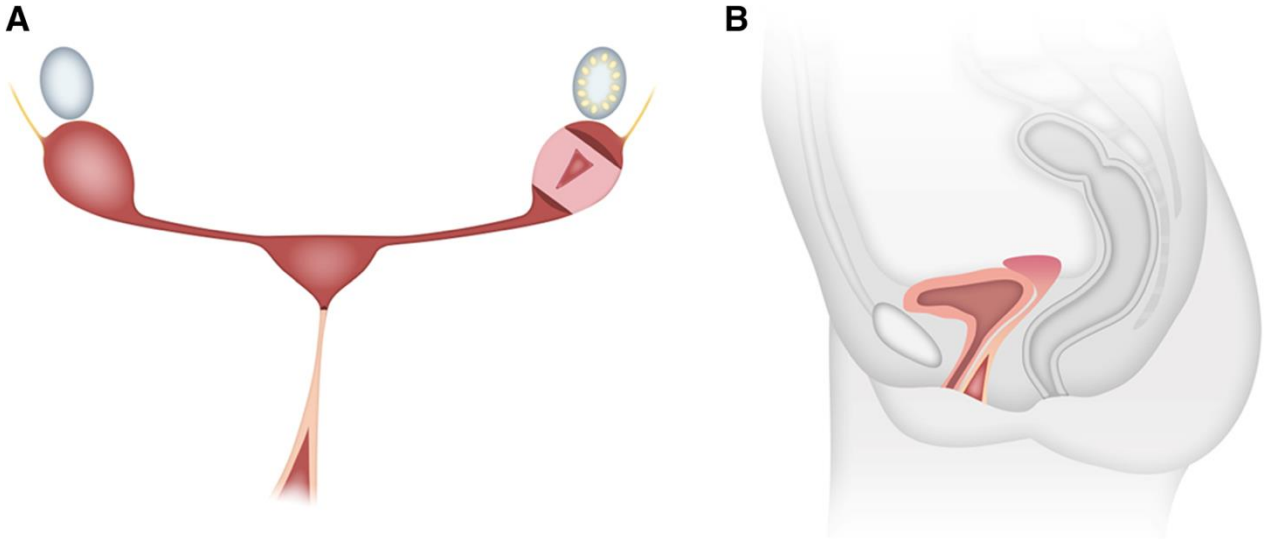
Children's Hospital Colorado



Mullerian Agenesis

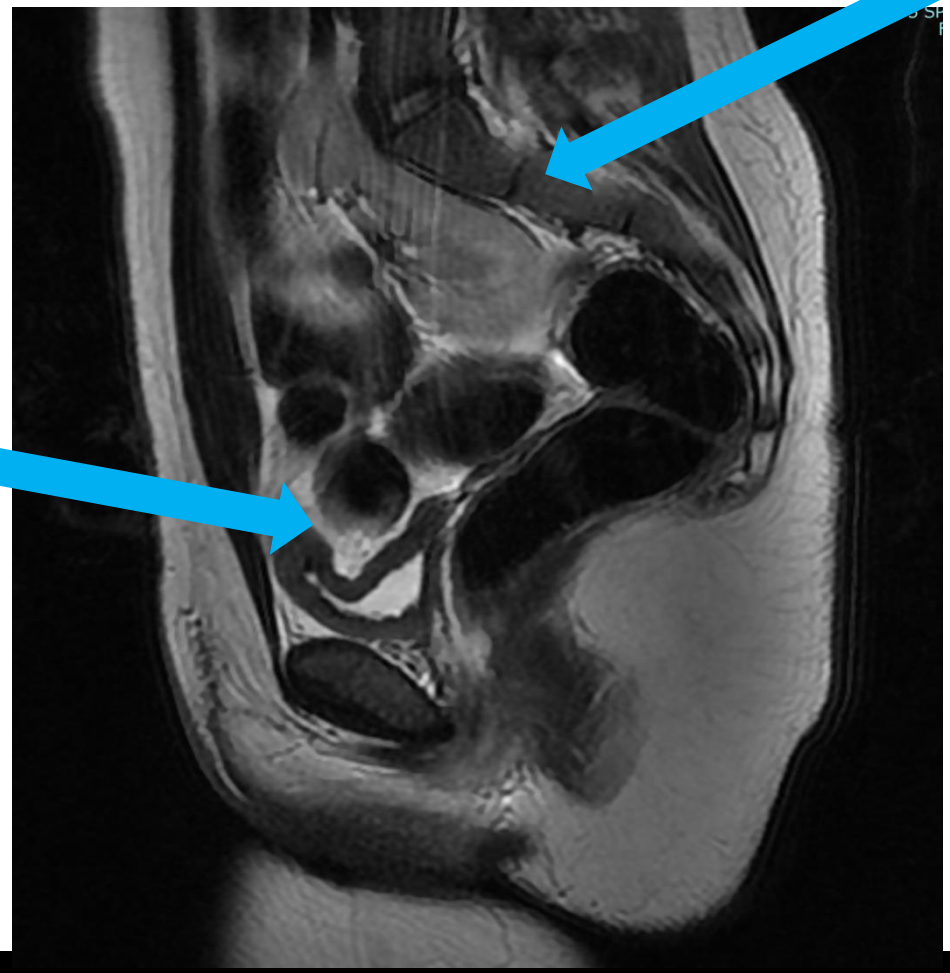
Key MRI findings

- Pelvic structures include bilateral uterine buds, fibrous band, and midline triangular soft tissue.
- Midline soft tissue is continuous with the atrophic vagina
- Normal ovaries near uterine buds

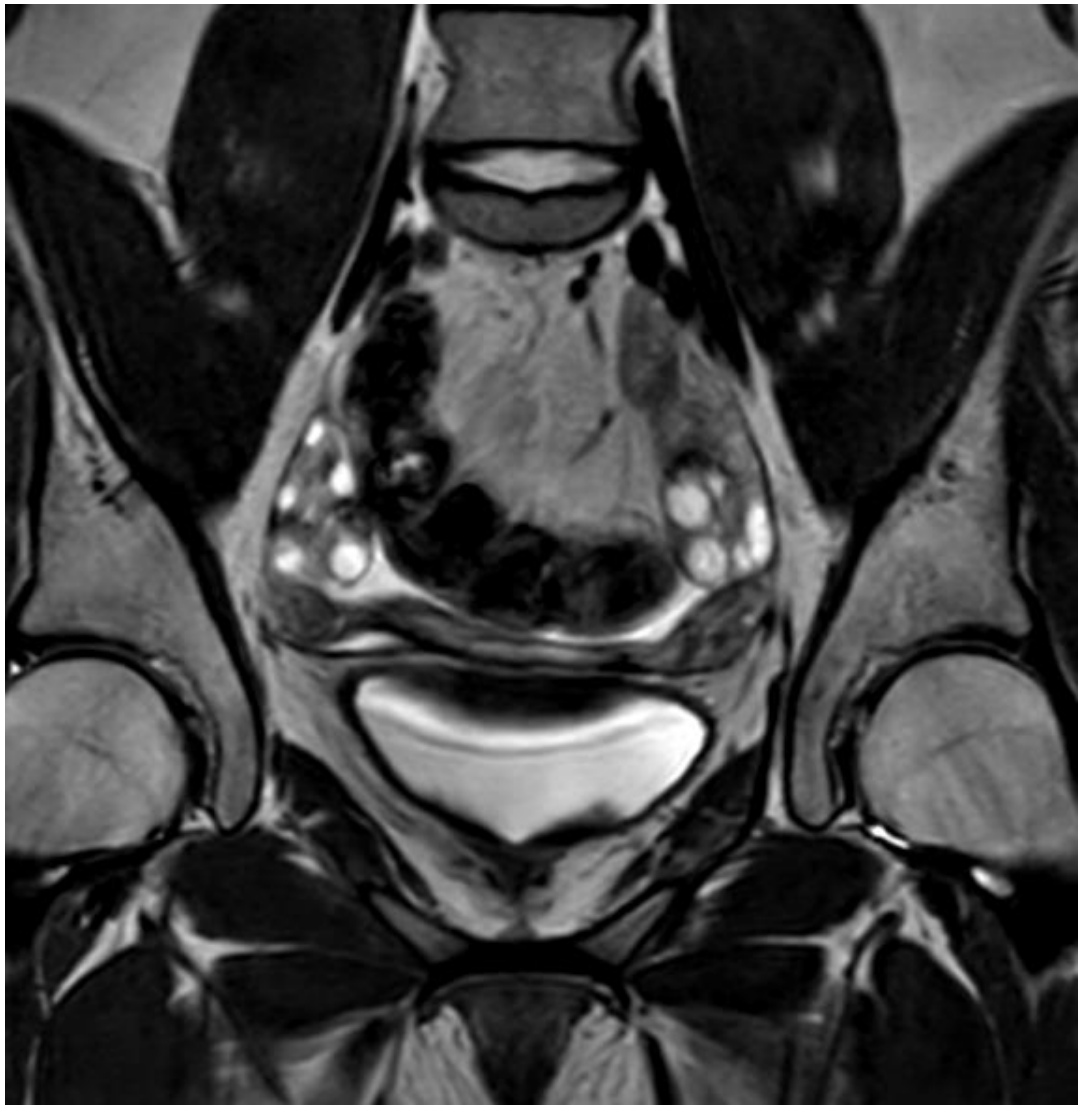


Pelvic MRI

Absent uterus without remnants



Mullerian Agenesis MRI



Types

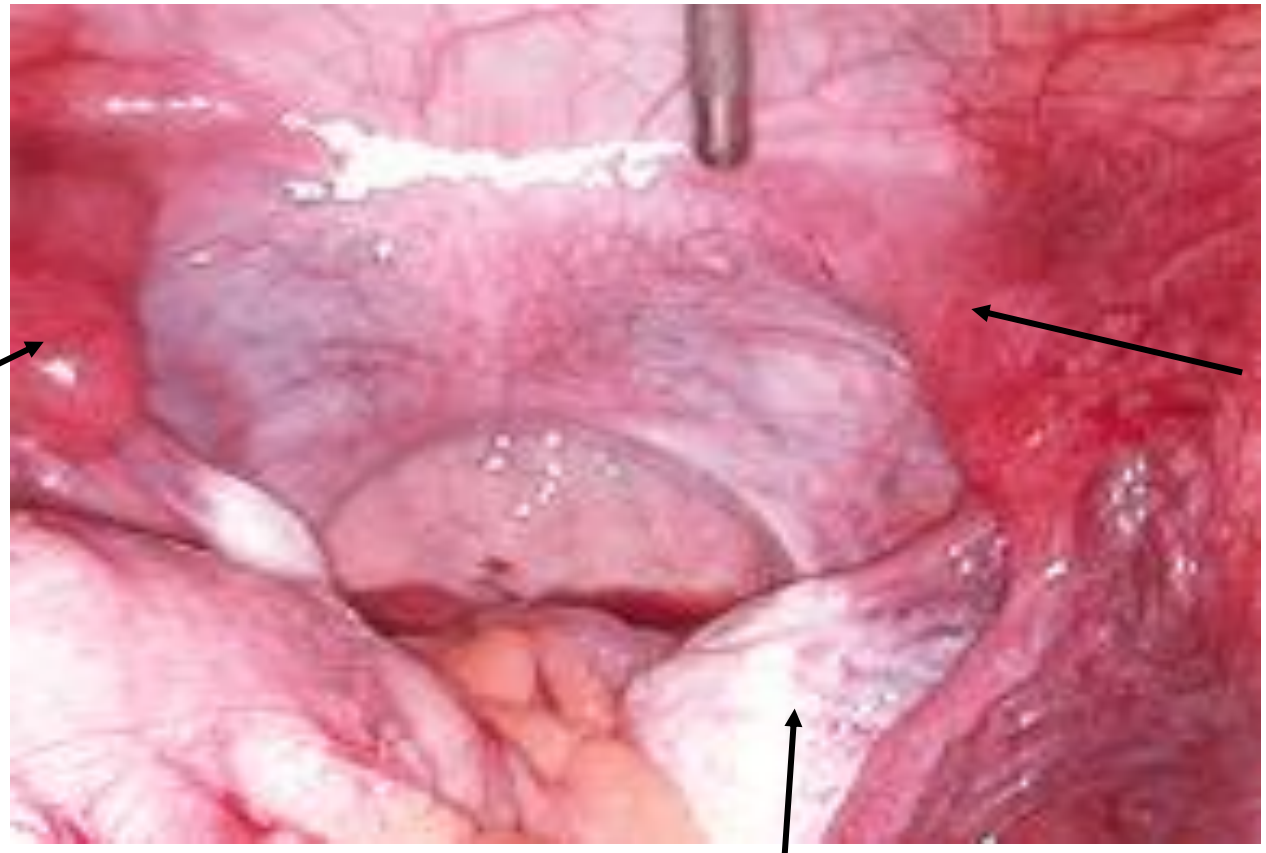
Type 1

- Isolated Mullerian agenesis

Type 2

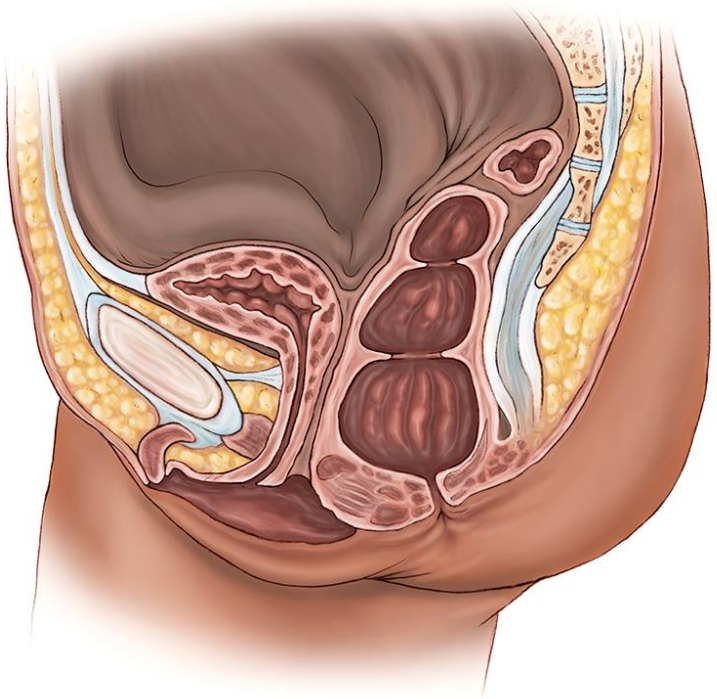
- Mullerian agenesis with another associated congenital anomaly

Laparoscopy



Considerations and Treatment

- Psychological and peer support
- Screening for Anomalies
- Discussion about fertility
- Vaginal creation
- Evaluation and treatment of any pelvic pain

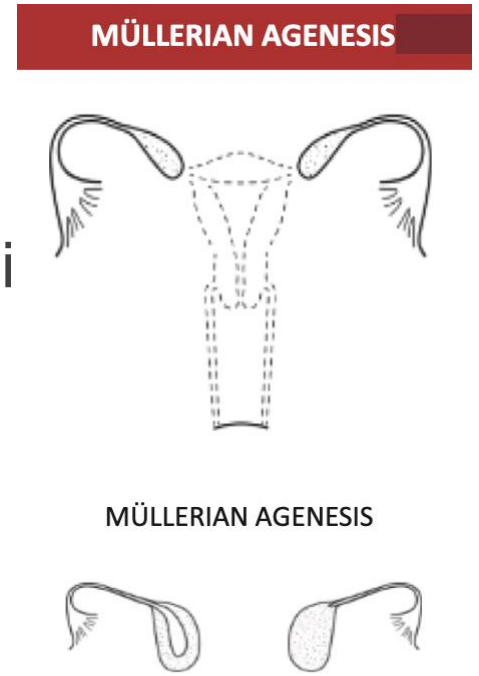


Pelvic Pain

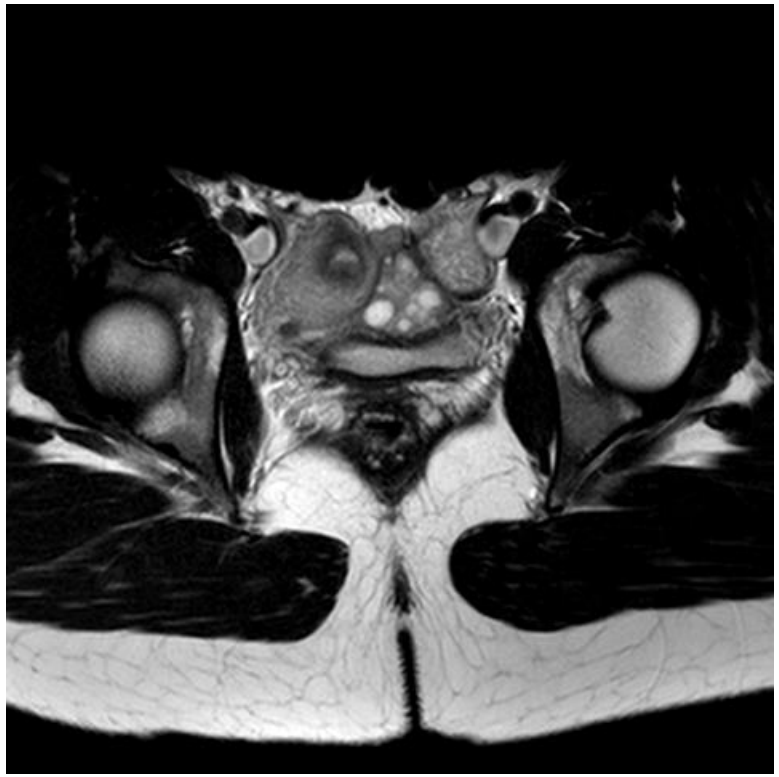
- Why do patients with Mullerian Agenesis present with pai

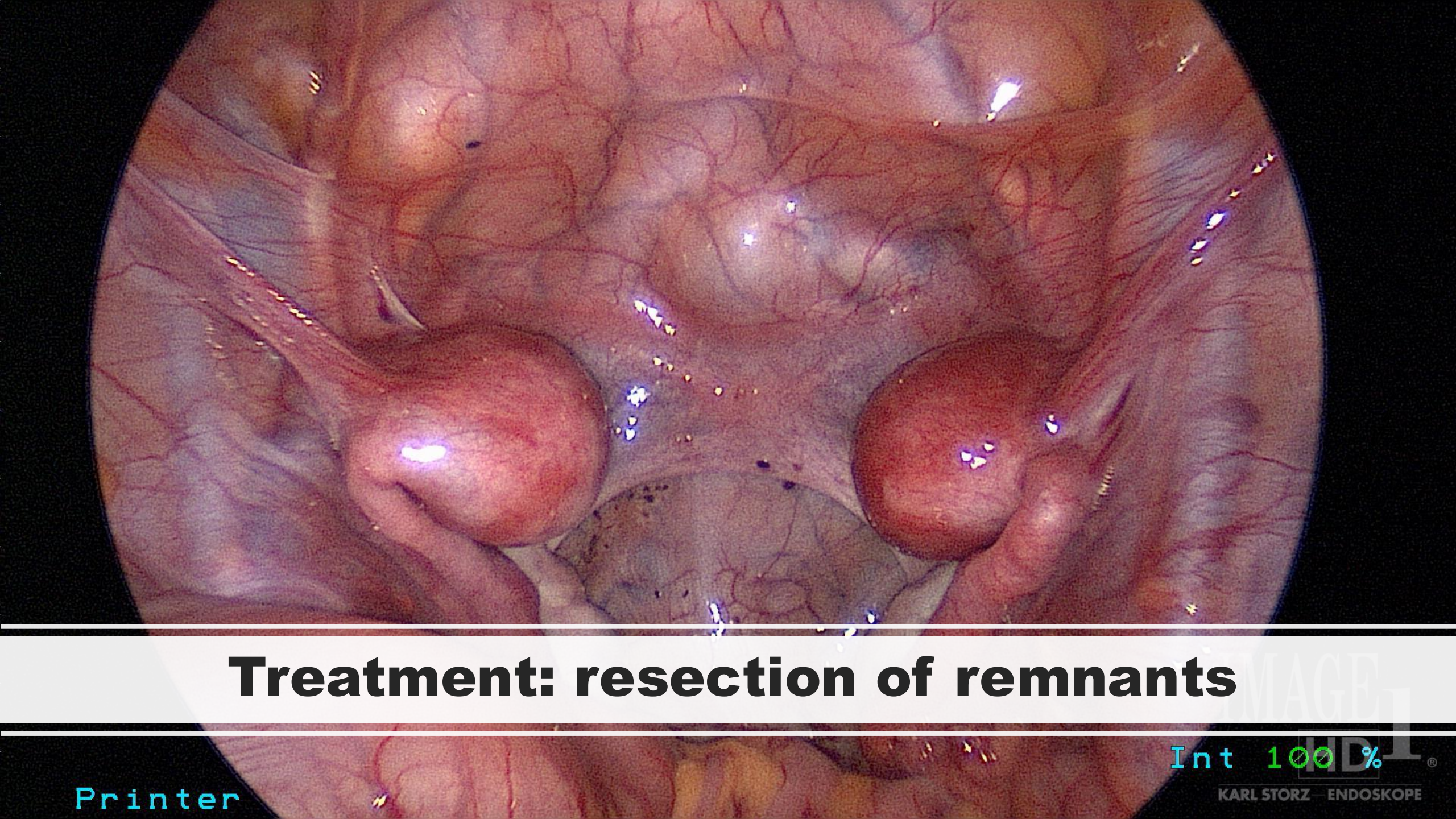
- Ovulation pain
- Uterine remnants
- Endometriosis

- All of the above



17 year old with Mullerian Agenesis and cyclical pain





Treatment: resection of remnants

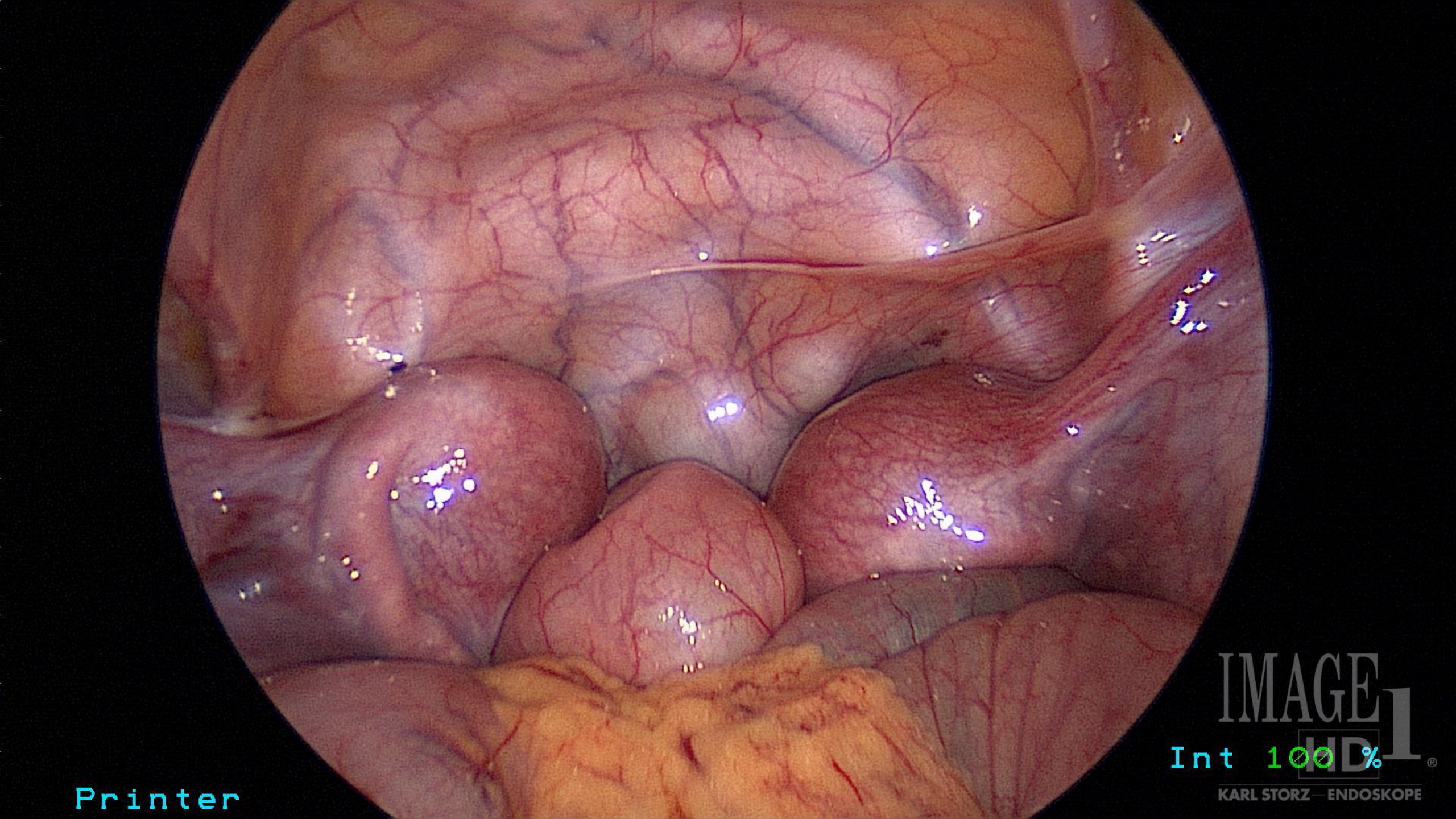
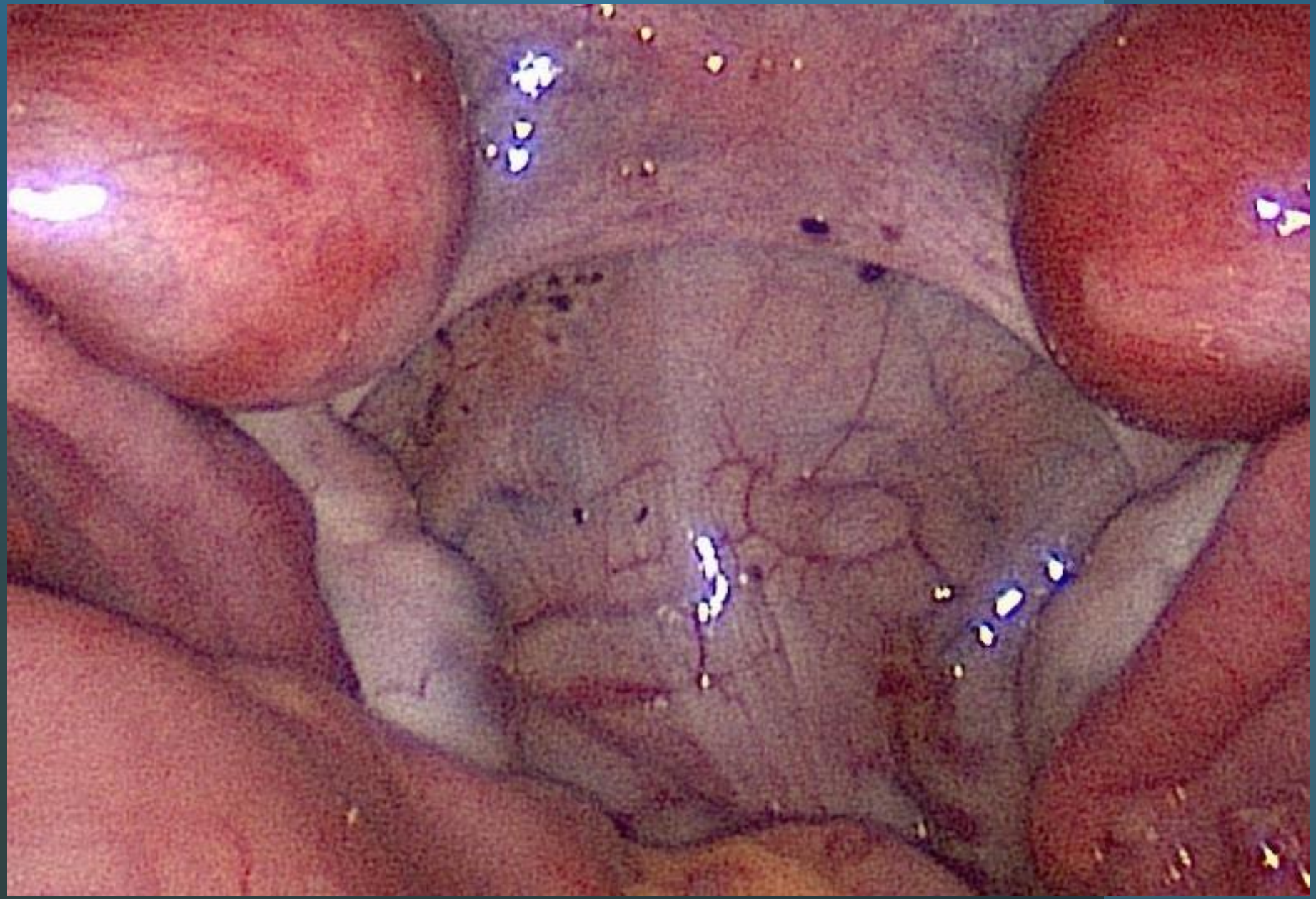


IMAGE1 HD
Int 100%
KARL STORZ — ENDOSKOPE
Printer



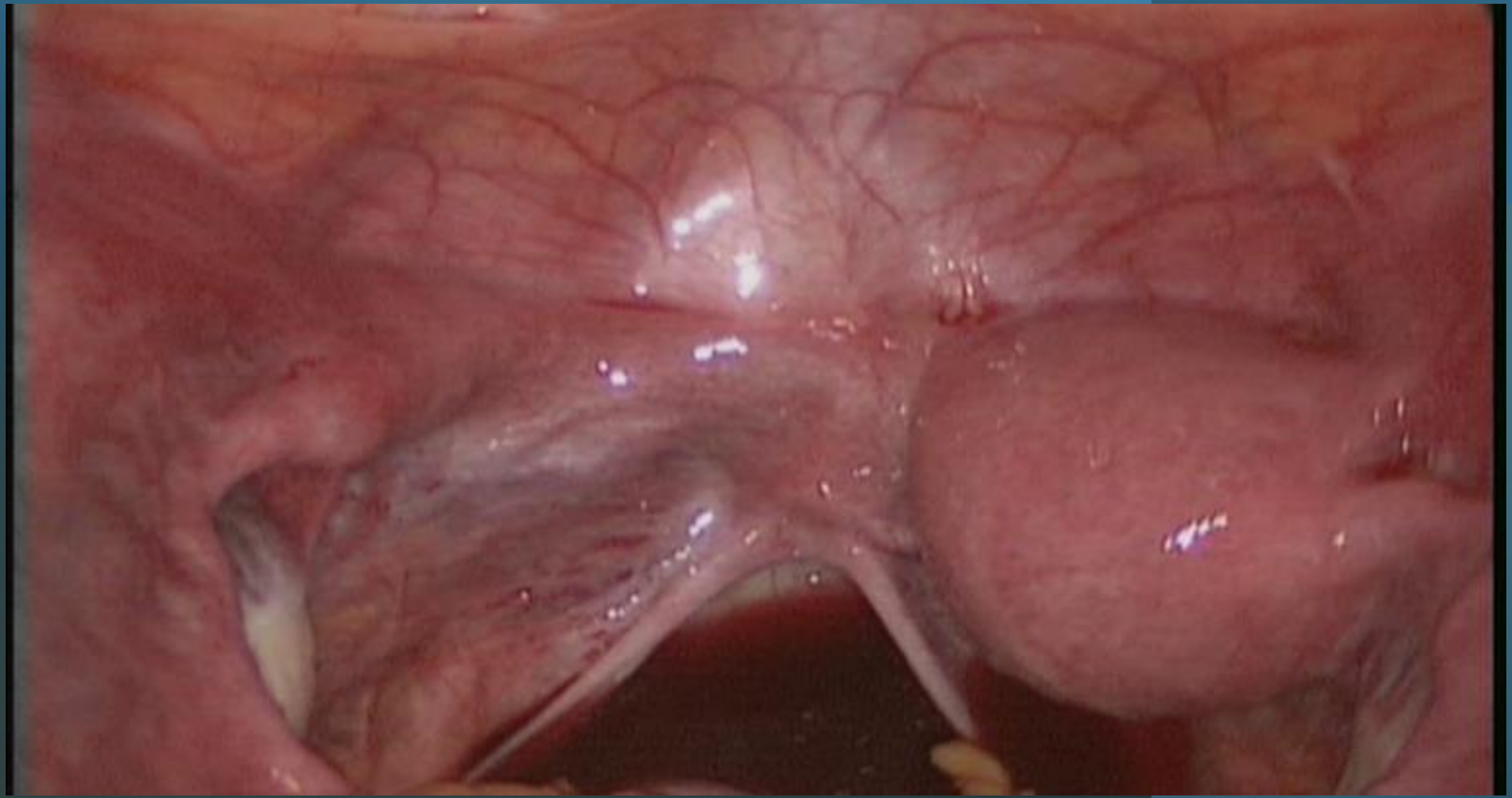



IMAGE1
HD
KARL STORZ - ENDOSKOPE

Vaginal Dilation

- First line option for vaginal creation
- Only when patient is ready
- Self dilation
  - Use of serial dilators
  - Length before width
  - 20-30 minutes per day
  - Privacy
  - Takes around 6 mo-1 year
- Coital dilation
- Physical therapy

Mayer-Rokitansky-Küster-Hauser syndrome: a review of 245 consecutive cases managed by a multidisciplinary approach with vaginal dilators

D. Keith Edmonds, F.R.C.O.G., Gillian L. Rose, F.R.C.O.G., Michelle G. Lipton, D.Clin.Psy., and Julie Quek, R.N.

Queen Charlotte's and Chelsea Hospital, Imperial College Healthcare NHS Trust, London, United Kingdom

- All patients admitted to the hospital x 3 days for instruction and monitored dilation
- Psychological support provided
- Seen Q 2 weeks after hospitalization
- 95% success rate

Sexual function and quality of life after the creation of a neovagina in women with Mayer-Rokitansky-Küster-Hauser syndrome: comparison of vaginal dilation and surgical procedures

Jia Kang, M.D., Na Chen, M.D., Shuang Song, M.D., Ye Zhang, M.D., Congcong Ma, M.D., Yidi Ma, M.D., and Lan Zhu, M.D.

Department of Obstetrics and Gynecology, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Sciences, Beijing, People's Republic of China

- Cross sectional study including women treated with vaginal dilation (n=88) and surgery (n=45)
- Vaginal length shorter in women who dilated
- No difference in sexual function and Quality of Life

Functional Vaginal Length

=

Success

If vaginal dilation fails…

Vaginoplasty Method	Advantages	Disadvantages	Anatomical success rate
Vecchietti	Avoid initial self-dilation; Fast and effective	Hospitalization; complications (2%), Need for stent and patient compliance	97-99%
McIndoe	Avoid initial self-dilation; Non abdominal access;	Hospitalization; complications (1-7%); Need for stent and patient compliance; hair growth	83%
Davydov	Avoid initial self-dilation; Faster; minimal risk of shrinkage	Hospitalization; complications (3-5%); Granulation (8%)	
Intestinal graft	Avoid initial self-dilation; highest vaginal length	Bigger operation. Mucous production and stricture risk (10.5%).	
Wharton Sheares George	No transplant needed; low complications	Stricture risk (9%); long term dilation needed	93%

Kölle A, Taran FA, Rall K, Schöller D, Wallwiener D, Brucker SY. Neovagina creation methods and their potential impact on subsequent uterus transplantation: a review. BJOG. 2019 Oct;126(11):1328-1335.

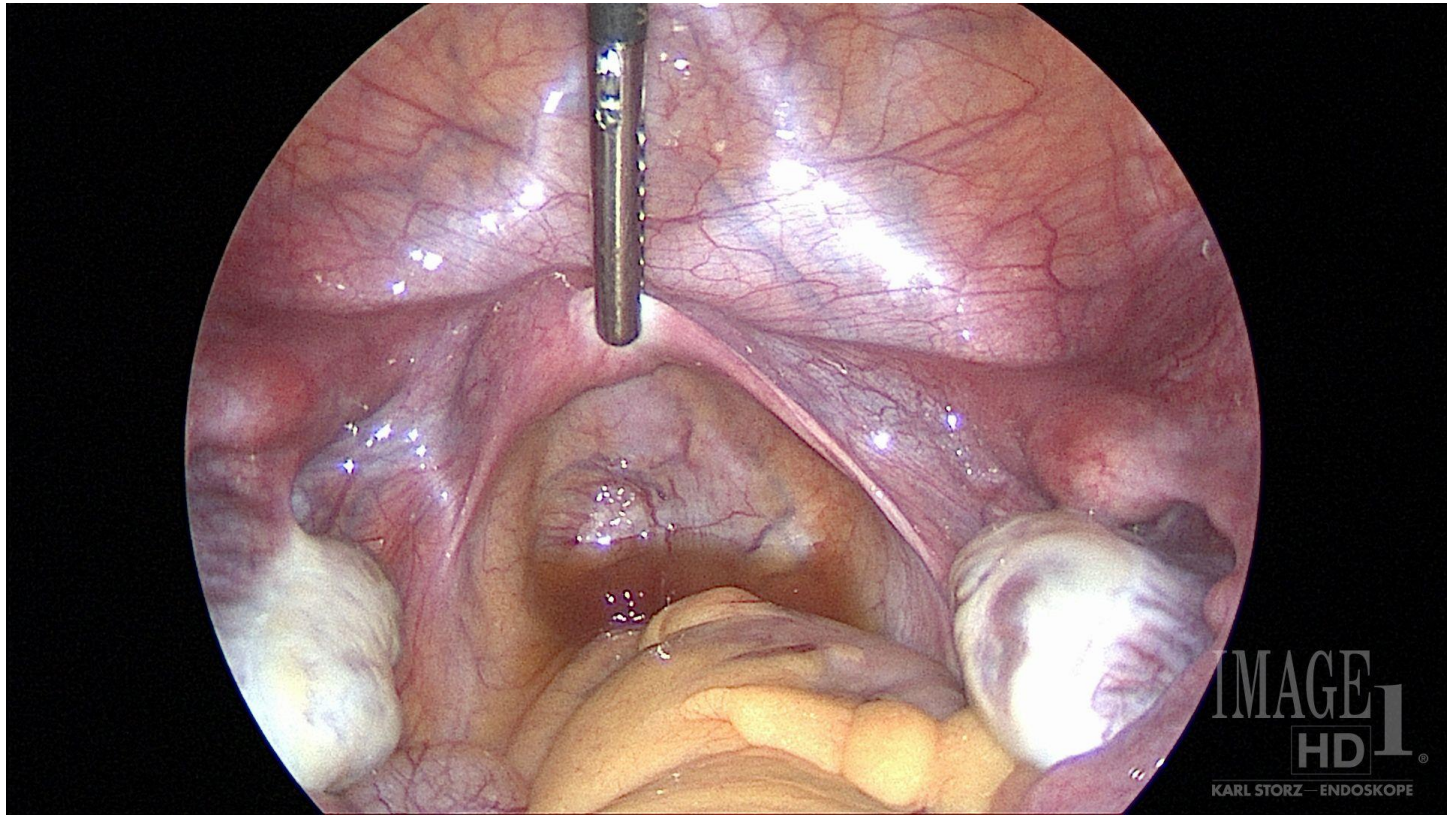
Other Graft Options

- AlloDerm
  - Acellular cadaver skin graft
  - Acts as a tissue matrix for regeneration

- Fish skin
  - Tilapia
  - Atlantic cod (Kerecis)
  - Similarly acts as a tissue matrix for regeneration

- Placed via modified Wharton Shears Vaginoplasty

Vecchietti



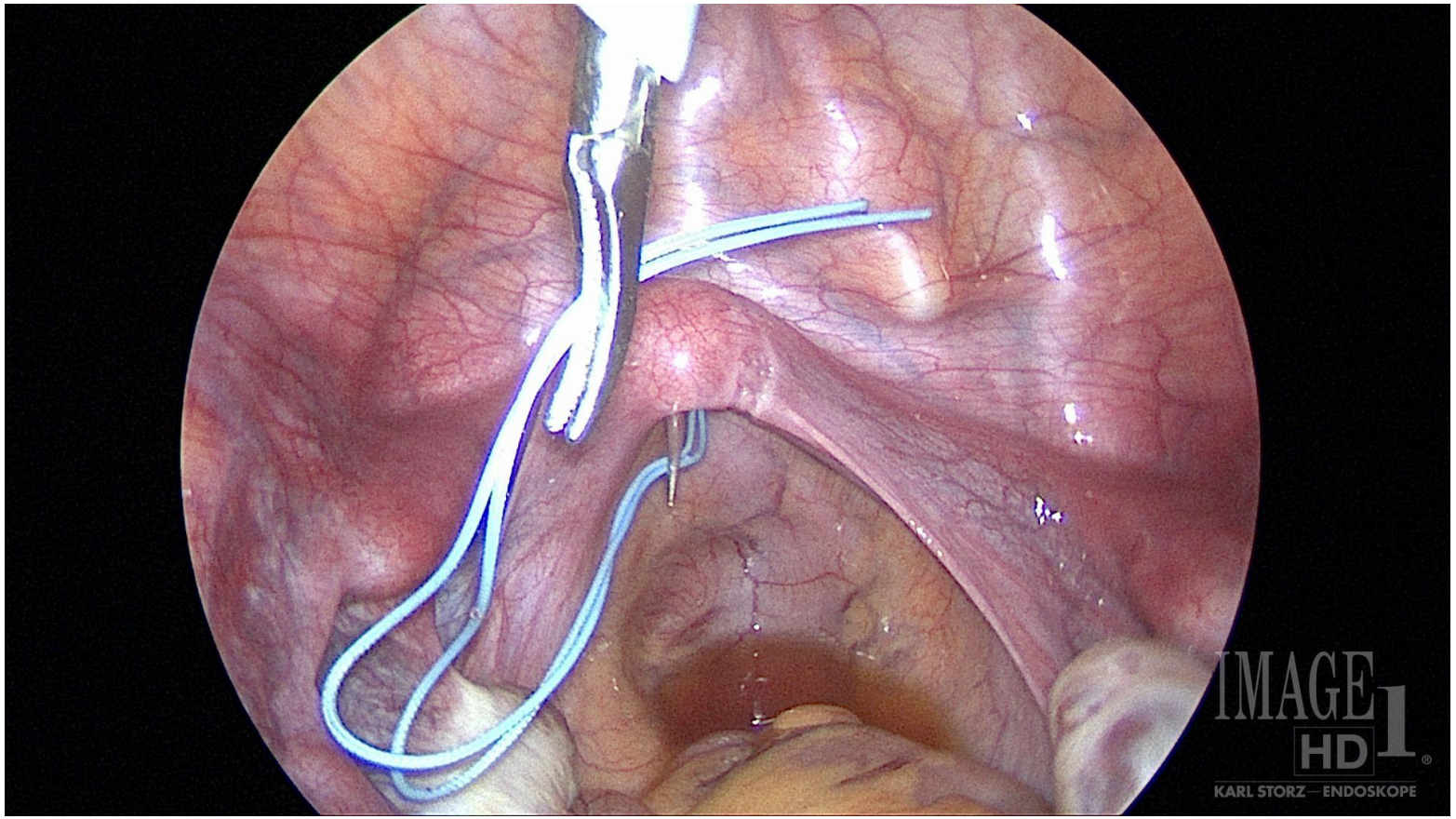


IMAGE 1 HD
KARL STORZ — ENDOSKOPE

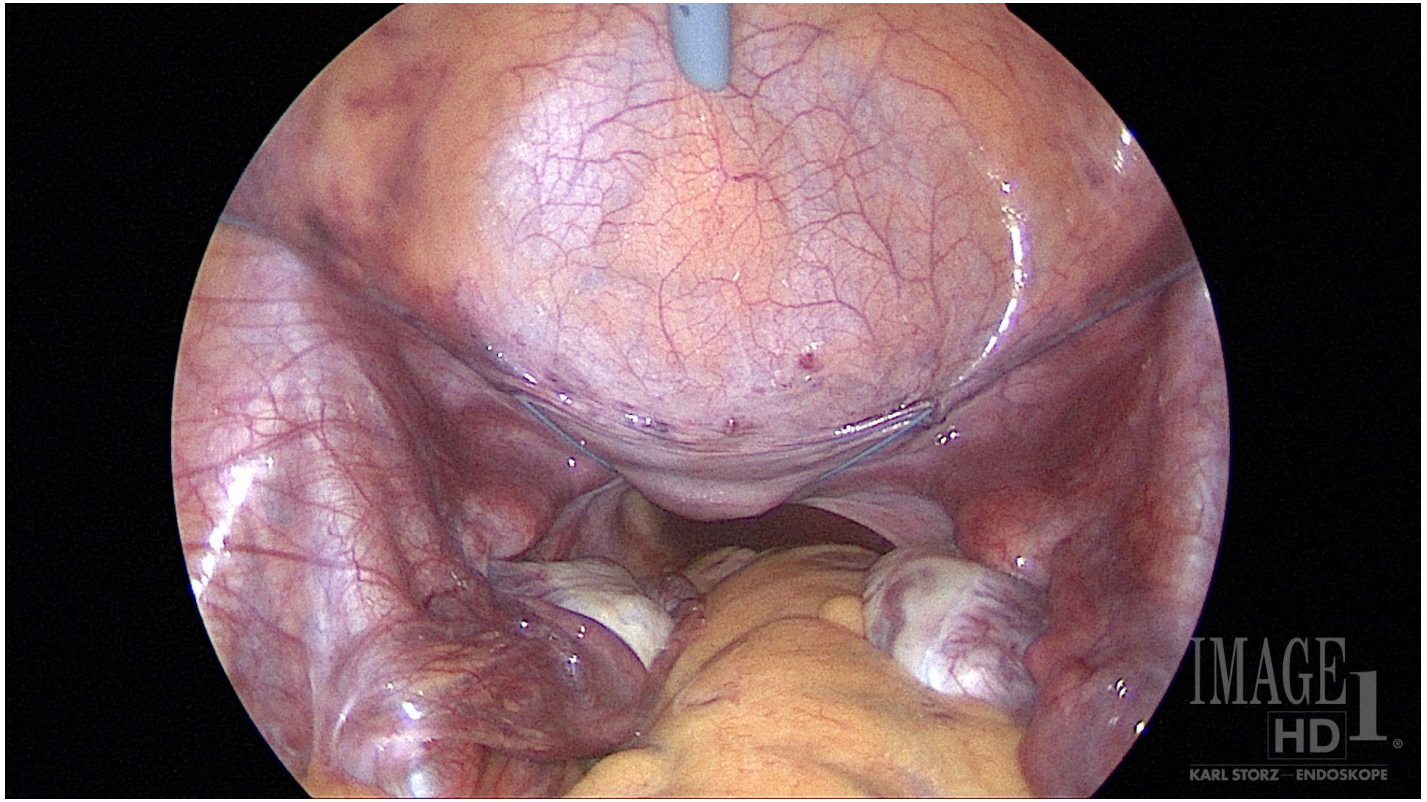
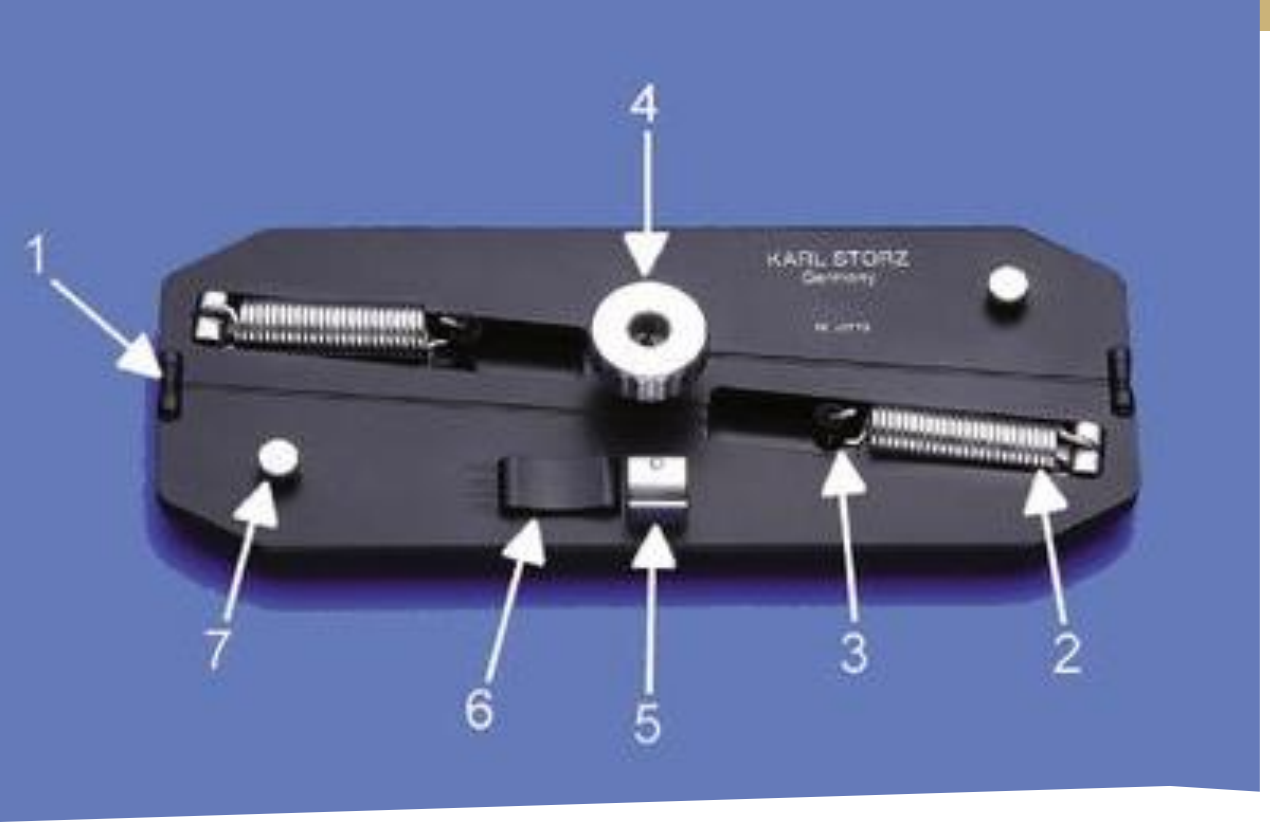


IMAGE 1 HD
KARL STORZ — ENDOSKOPE



Laparoscopic Vecchietti

Children's Hospital Colorado

Pediatric and Adolescent Gynecology
UNIVERSITY OF COLORADO **ANSCHUTZ MEDICAL CAMPUS**

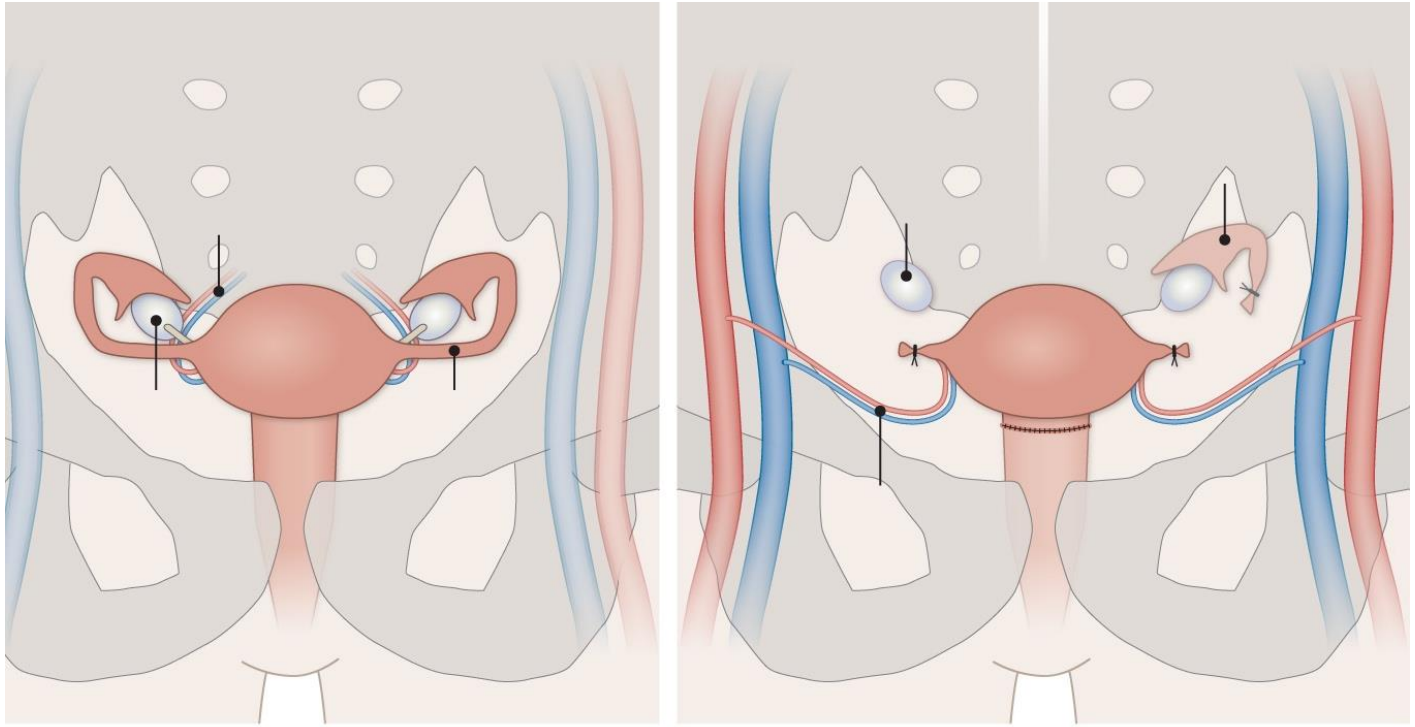
Uterine Transplant Considerations

- Few abdominal surgeries and minimal adhesive disease
- Elastic neovagina of sufficient length (> 8 cm) and width (> 2 cm)
- Natural epithelium
- Natural anatomic axis

- Prior recipients
  - Vechietti neovagina
  - Dilation
  - Native vagina

Uterine Transplant Outcomes

- First 5 years in the US
  - 33 recipients
  - 94% had Mullerian agenesis
  - 64% received from living donors
  - 74% one year graft survival
  - 59% (19/33) delivered 21 children

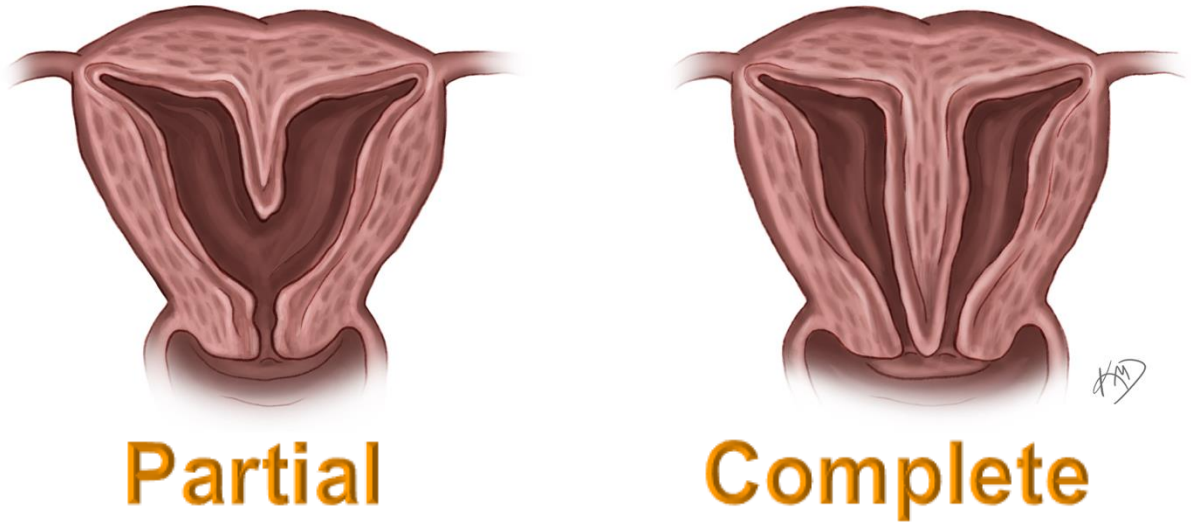


Johannesson L, Richards E, Reddy V, Walter J, Olthoff K, Quintini C, Tzakis A, Latif N, Porrett P, O'Neill K, Testa G. The First 5 Years of Uterus Transplant in the US: A Report From the United States Uterus Transplant Consortium. JAMA Surg. 2022 Sep 1;157(9):790-797.

www.beautifulyouMRKH.org

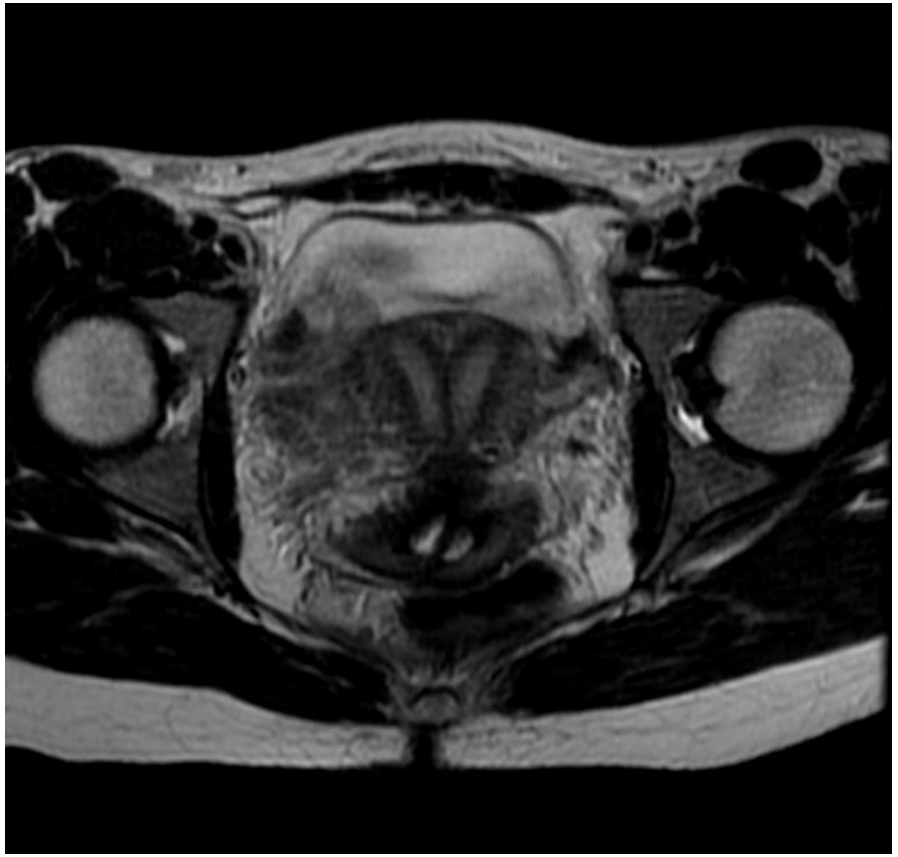
Septate Uterus

- May be associated with adverse pregnancy outcomes
  - Miscarriage
  - Preterm delivery
  - Intrauterine growth restriction
  - Breech presentation
  - C-section
  - Placental abruption



Septate Uterus

- Hysteroscopic septum resection can be considered to to optimize the uterine cavity
  - Evidence is limited
  - May have benefit in patients with history of poor pregnancy outcome.

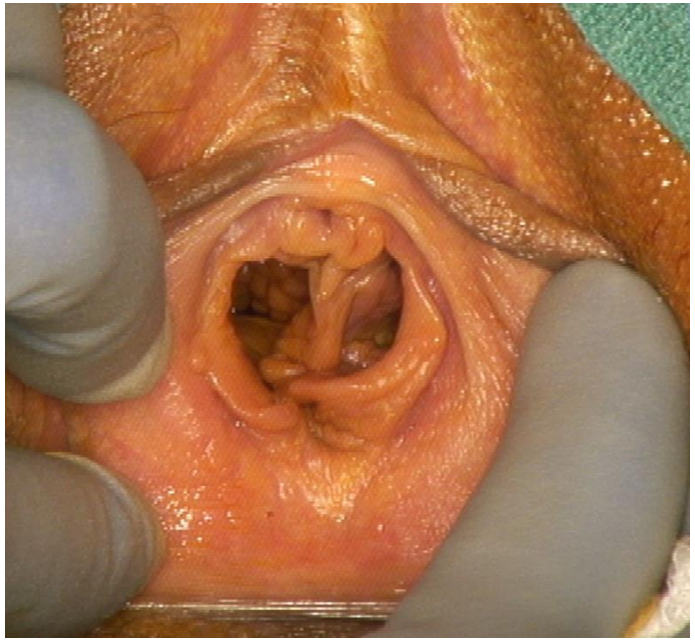


Septate Uterus

- Rikken et al. Septum resection vs expectant management in women with a septate uterus: an international multicentre open label RCT
  - Included 80 women with a septate uterus and history of subfertility, pregnancy loss, or PTB
  - Majority had a partial septum (92% in septum resection group, 90% in expectant management group)
  - No difference in live birth, pregnancy loss, PTB
- Carrera et al. Effect of hysteroscopic metroplasty on reproductive outcomes in women with a septate uterus: systematic review and meta analysis
  - 11 studies included in the analysis (1 RCT and 10 observational studies)
  - Hysteroscopic metroplasty reduces risk of miscarriage in complete and partial uterine septum
  - No difference in clinical pregnancy, term live birth or risk of c-section
- Kowalik et al. Septum resection for women of reproductive age with septate uterus (Cochrane review)
  - No RCTs at the time of review
  - No evidence to support surgical intervention

Didelphys Uterus and Longitudinal Vaginal Septum

- **Childhood:** No implications
- **Adolescence and adulthood:** Dyspareunia and difficulty with menstrual hygiene
- **Pregnancy:** Didelphys uterus is associated with growth restriction, preterm labor/birth, fetal malpresentation and need for C-section



Uterus Didelphys Pregnancy Outcomes

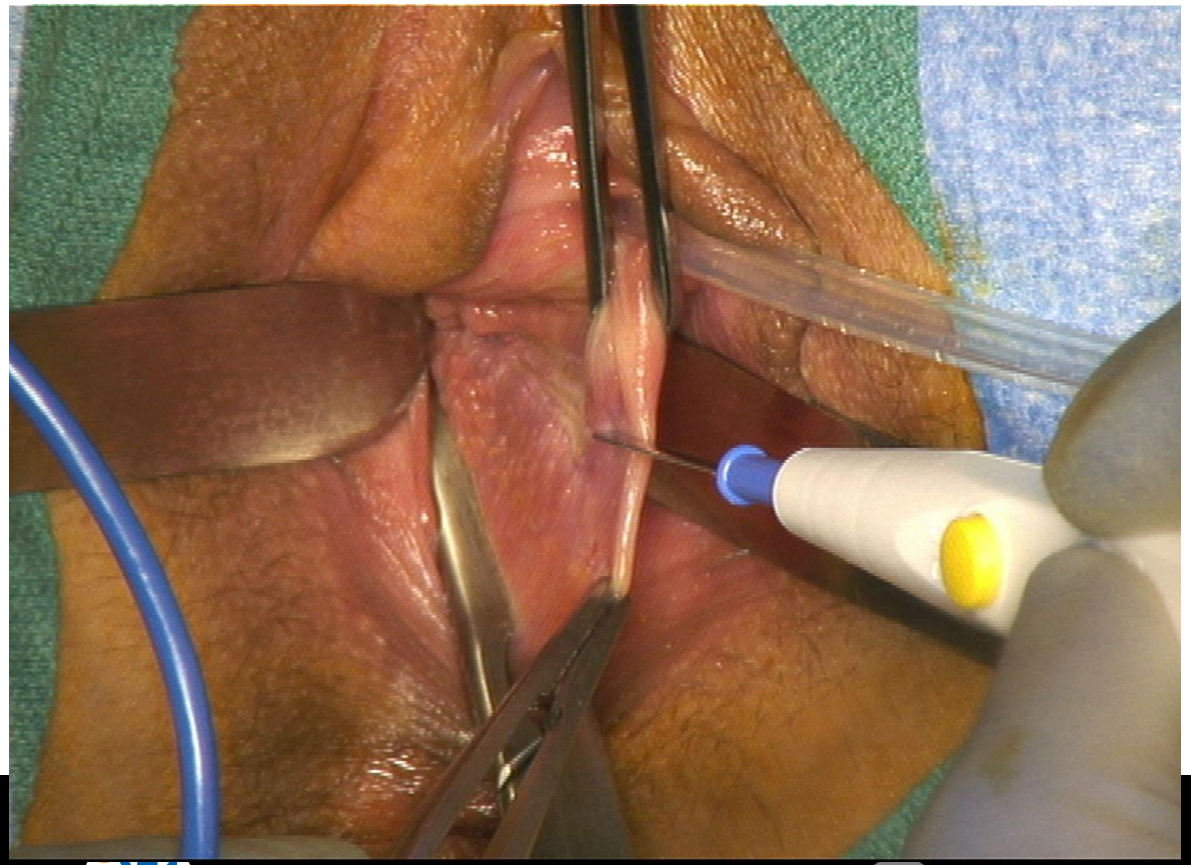
Table VI. Pregnancy outcome in patients with untreated didelphys uterus

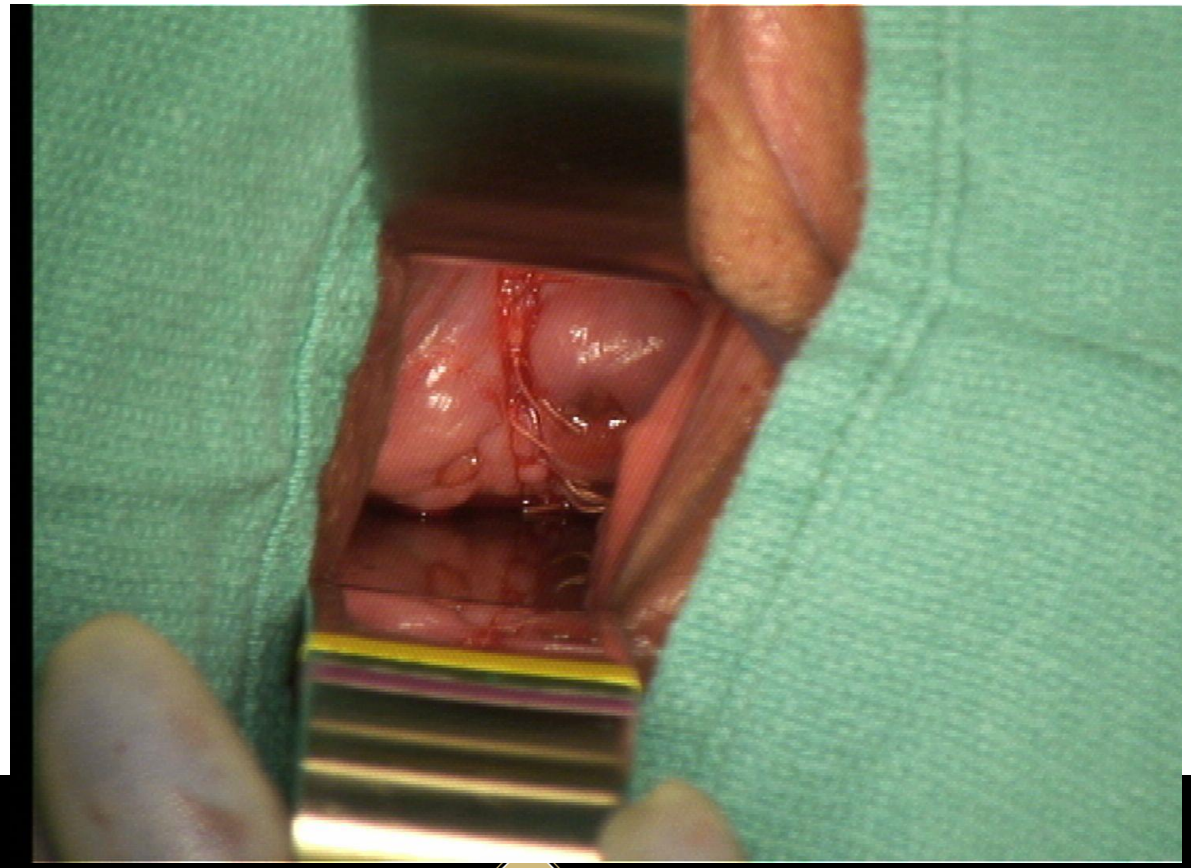
Study	Patients *n*	Conceiving *n*	Pregnancies *n*	Ectopics *n* (%)	Abortions *n* (%)	Preterm deliveries *n* (%)	Term deliveries *n* (%)	Live births *n* (%)
Michalas *et al.* (1976)	3	3	5	0	3 (60.0)	2 (40.0)	0	2 (40.0)
Heinonen *et al.* (1982)	21	13	25	0	8 (32.0)	6 (24.0)	11 (44.0)	16 (64.0)
Buttram (1983)	4	3	5	0	3 (60.0)	1 (20.0)	1 (20.0)	2 (40.0)
Fedele *et al.* (1988)	13	11	29	0	20 (69.0)	7 (24.1)	2 (6.9)	7 (24.1)
Stein and March (1990)	25	25	27	0	0	7 (25.9)	20 (74.1)	22 (81.5)
Moutos *et al.* (1992)	25	13	28[a]	1 (4.1)	6 (25.0)	9 (37.5)	8 (33.3)	17 (70.8)
Acien (1993)	15	10	18	0	5 (27.8)	3 (16.7)	10 (55.5)	13 (72.2)
Raga *et al.* (1997)	8	?	15	1 (6.7)	4 (26.7)	8 (53.3)	3 (20.0)	6 (40.0)
Total	114	78/106[b]	152	2 (1.3)	49 (32.2)	43 (28.3)	55 (36.2)	85 (55.9)

[a]Four pregnancies ended in elective abortion.
[b]Total number of patients from series with data on conception.

Grimbizis GF, Camus M, Tarlatzis BC, Bontis JN, Devroey P. Clinical implications of uterine malformations and hysteroscopic treatment results. Hum Reprod Update. 2001 Mar-Apr;7(2):161-74.

Longitudinal Vaginal Septum





Resection Using Bipolar (Ligasure)

Case Reports > J Pediatr Adolesc Gynecol. 2016 Dec;29(6):e95-e96.
doi: 10.1016/j.jpag.2016.06.002. Epub 2016 Jun 16.

Longitudinal Vaginal Septum Resection Using the Ligasure Device

Gisselle Perez-Milicua [1], Julie Hakim [1], Oluyemisi Adeyemi [1], Jennifer E Dietrich [2]

Affiliations + expand

PMID: 27321900 DOI: 10.1016/j.jpag.2016.06.002

Case Reports > J Pediatr Adolesc Gynecol. 2023 Dec;36(6):563-565.
doi: 10.1016/j.jpag.2023.08.002. Epub 2023 Aug 7.

Postoperative Bleeding Two Weeks After Longitudinal Vaginal Septum Resection with the LigaSure Device: A Case Report

Hana G Murphy [1], Melina L Dendrinos [2], Monica W Rosen [3]

Affiliations + expand

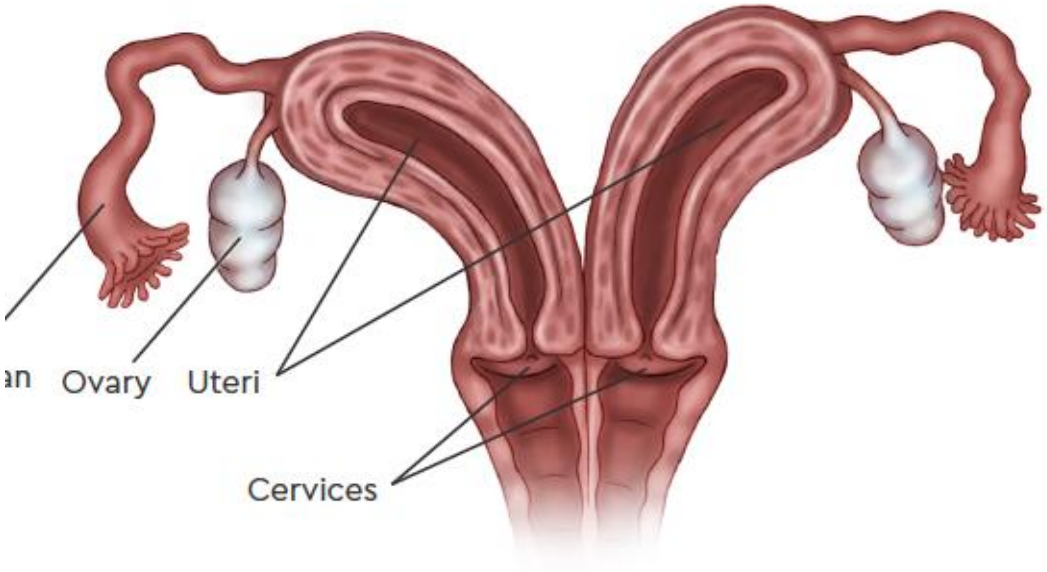
PMID: 37558158 DOI: 10.1016/j.jpag.2023.08.002

Cool Seal

SPECIFICATIONS	
Shaft diameter	5 mm
Shaft length	10 cm
Jaw type	Maryland
Jaw length	12 mm
Seal length	11 mm
Cut length	10 mm

CoolSeal generally offers a lower thermal profile, leading to less thermal damage and a smaller heat footprint compared to LigaSure

Pregnancy Surveillance



- Cervical length Q 2 weeks from 16-24 weeks gestation
- Fetal growth scans every 4 weeks

Congenital Anomalies Overall Summary

- Consider normal sequence of pubertal development when considering the diagnosis of anomalies
  - An obstructive anomaly usually presents earlier after thelarche and with pain
  - An anomaly without obstruction can go unnoticed for years
- Preop:
  - Make sure the diagnosis is correct
  - MRI is gold standard for anomalies
  - Phone a friend with any uncertainty

Pediatric and Adolescent Gynecology
UNIVERSITY OF COLORADO **ANSCHUTZ MEDICAL CAMPUS**

Children's Hospital Colorado

THANK YOU