



Medical Academy named after S. I. Georgievsky

Chair of obstetrics, gynecology and perinatology # 1

# Abnormalities of development and position of the internal female genitalia

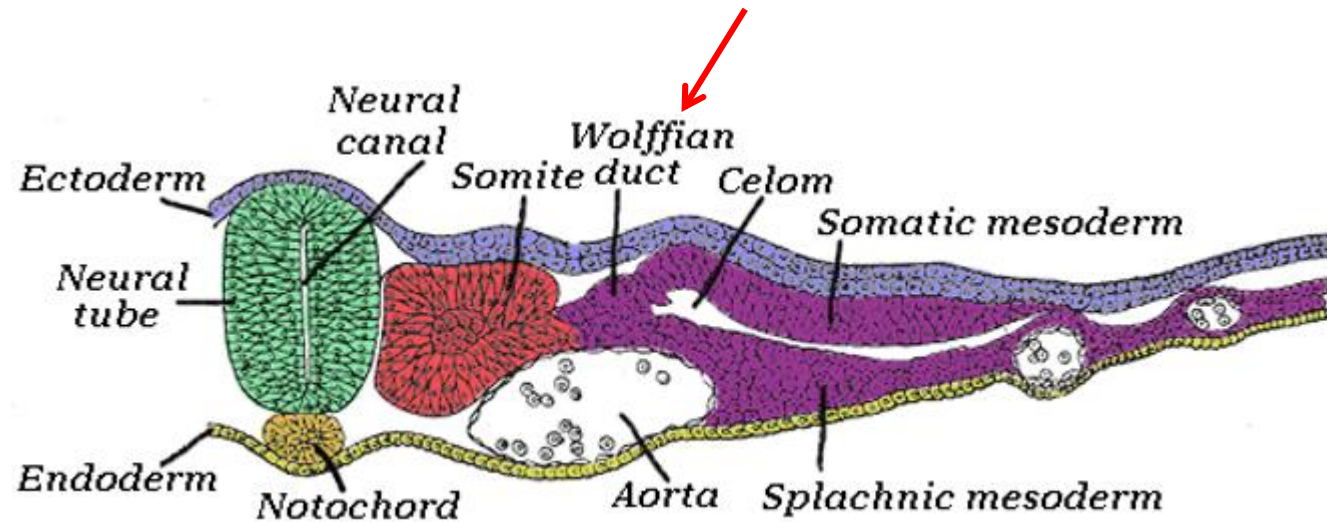
*Livshyts I. V.*

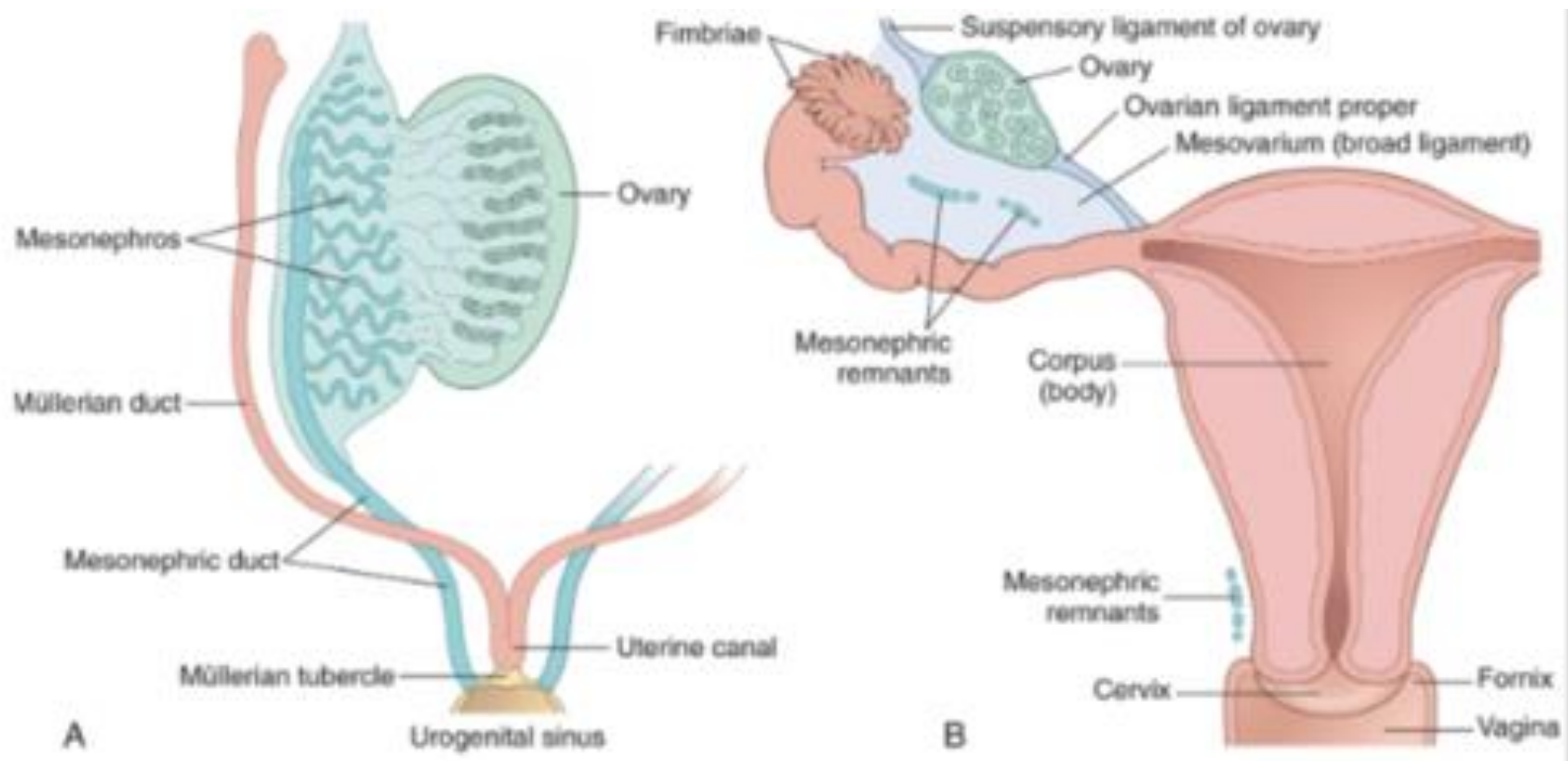
*PhD, Associate Professor*

- Congenital malformations of the female genital tract are defined as deviations from normal anatomy resulting from embryological maldevelopment of the Mullerian or paramesonephric ducts.
- They represent a rather common benign condition with a prevalence of 4–7%
- Müllerian malformations are frequently associated with abnormalities of the renal and axial skeletal systems

# Early embryo development

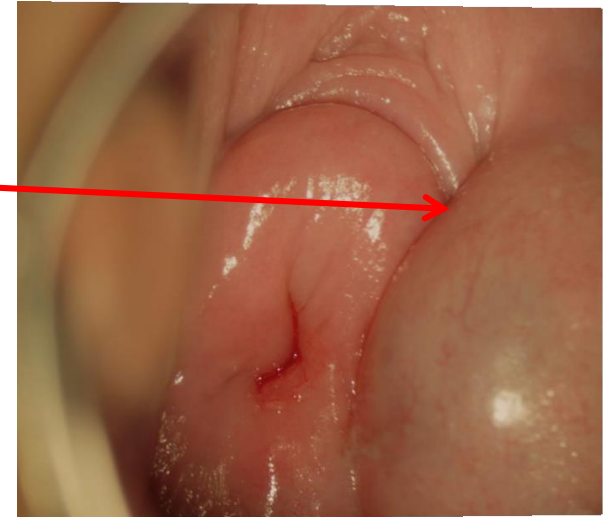
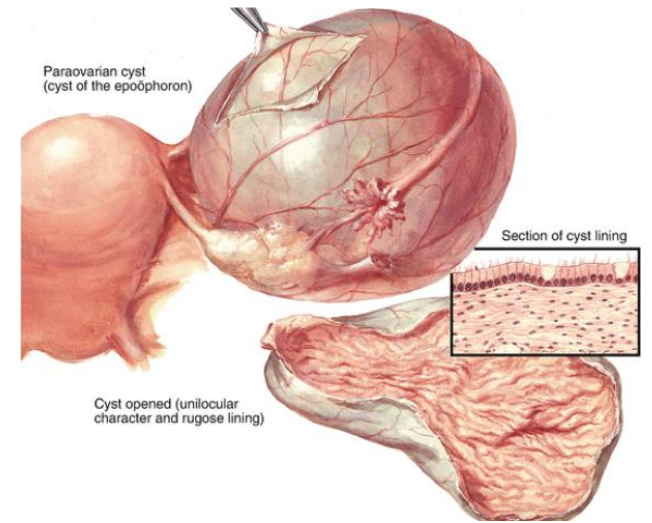
- Until 8 weeks' gestation, the human fetus is undifferentiated sexually and contains both male (wolffian) and female (müllerian) genital ducts.
- Wolffian structures (in the male fetus) differentiate into the vas deferens, epididymis, and seminal vesicles.





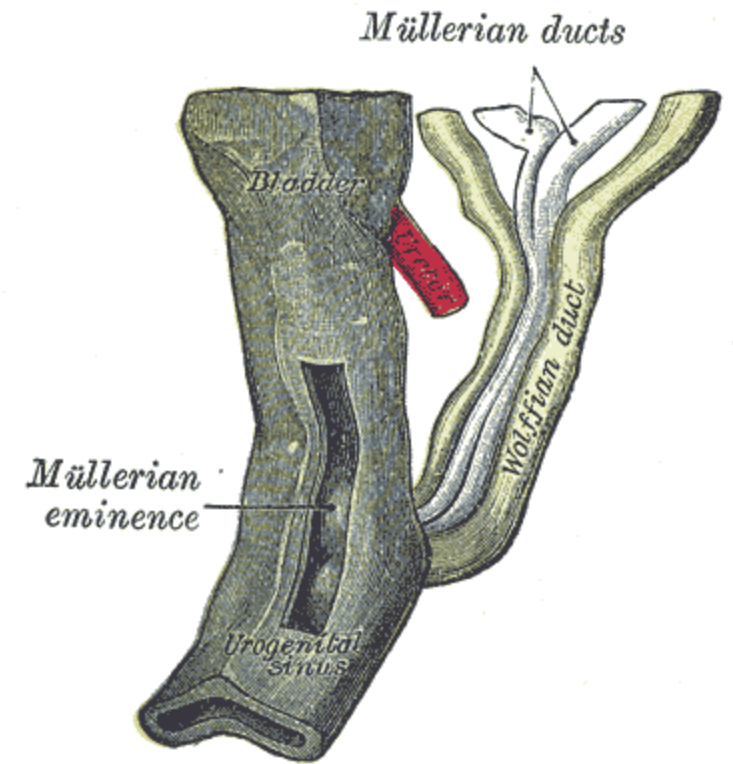
# Wolffian (mesonephric) duct

- In the female fetus, with the absence of testosterone secretion, the Wolffian duct regresses, but inclusions may persist.
- The epoophoron and Skene's glands may be present.
- Also, lateral to the wall of the vagina a Gartner's duct or cyst could develop as a remnant.



# Müllerian (paramesonephric) ducts

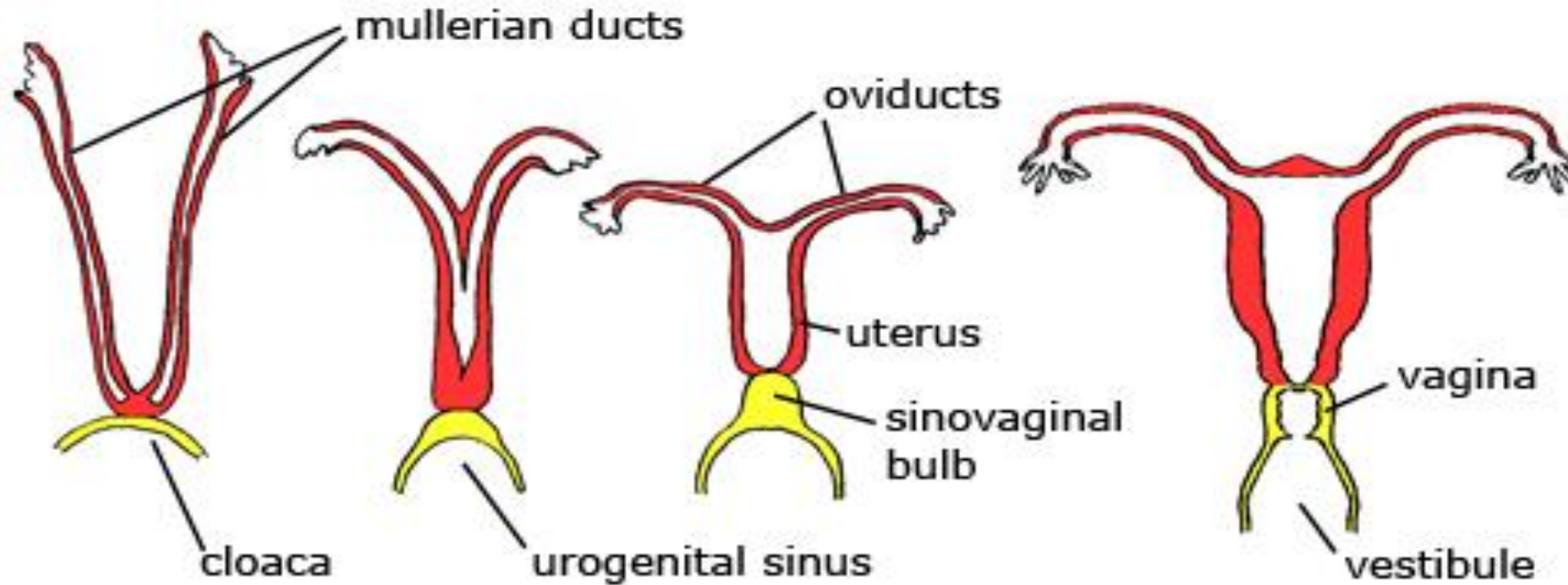
- Paired ducts of the embryo that run down the lateral sides of the urogenital ridge and terminate at the sinus tubercle in the primitive urogenital sinus.
- In the female, without the influence of AMH, they will develop to form the fallopian tubes, uterus, cervix, and the upper one-third of the vagina.
- In the absence of testosterone and dihydrotestosterone, the genital tubercle develops into the clitoris, and the labioscrotal folds do not fuse, leaving labia minora and majora.





The müllerian ducts (mesoderm) are the primordial anlage of the internal female reproductive organs and differentiate to form the fallopian tubes, uterus, uterine cervix and superior aspect of the vagina. The urogenital sinus (endoderm) is thought to give rise to the inferior and mid vagina.

## Embryonic formation of female reproductive system



- A reliable classification system seems to be extremely useful for their management
- Effective categorization enables more effective diagnosis and treatment as well as a better understanding of their pathogenesis



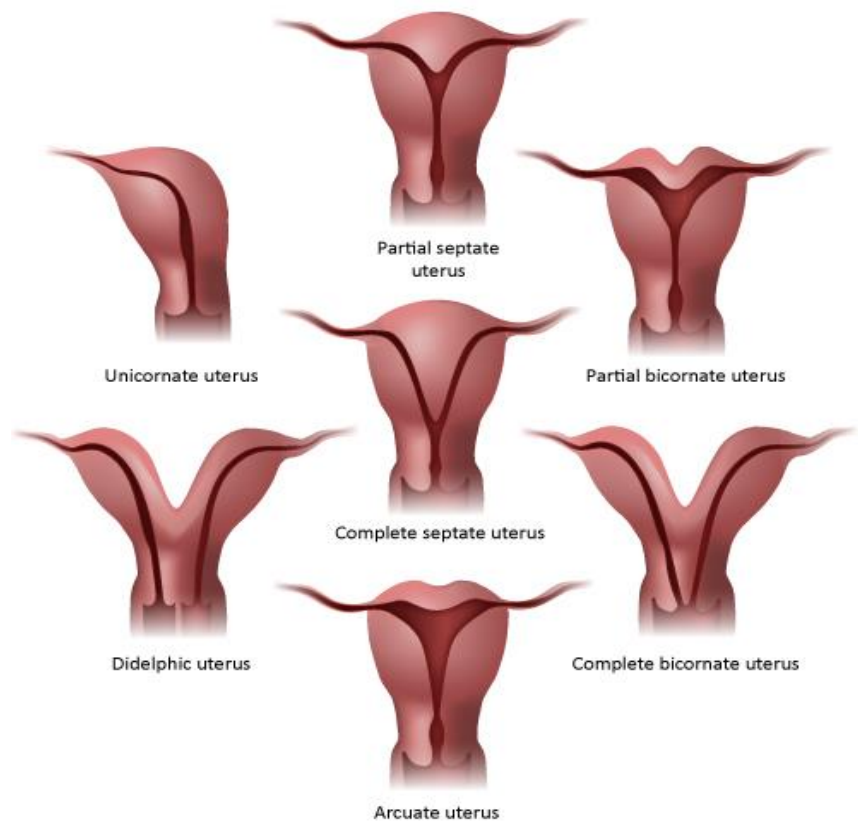
- Refinements in surgical techniques, such as the Vecchietti and McIndoe procedures, have enabled many women with müllerian duct anomalies to have normal sexual relations.
- Other surgical advances have resulted in improved fertility and obstetric outcomes.
- Developments in assisted reproductive technology allow some women with müllerian duct anomalies to conceive and deliver healthy babies.

# Systems for the classification of female genital tract anomalies

- American Society of Reproductive Medicine system (formerly AFS)
- The embryological-clinical classification system of genito-urinary malformations
- the Vagina, Cervix, Uterus, Adnexae and associated Malformations system based on the tumor nodes metastases (TNM) principle in oncology
- The ESHRE/ESGE classification system(European) of female genital anomalies

# The ESHRE/ESGE classification system

- (i) Anatomy is the basis for the systematic categorization of anomalies.
- (ii) Deviations of uterine anatomy deriving from the same embryological origin are the basis for the design of the main classes.
- (iii) Anatomical variations of the main classes expressing different degrees of uterine deformity and being clinically significant are the basis for the design of the main sub-classes.
- (iv) Cervical and vaginal anomalies are classified in independent supplementary sub-classes.



<p><b>Class U0/normal uterus</b></p>	<p><b>Class U1/Dysmorphic Uterus</b></p> <p>a. T-shaped      b. Infantilis      c. Others</p>	
<p><b>Class U2/septate uterus</b></p> <p>a. Partial      b. Complete</p>		<p><b>Class U3/Bicorporeal Uterus</b></p> <p>a. Partial      b. Complete      c. Bicorporeal septate</p>
<p><b>Class U4/Hemi Uterus</b></p> <p>a. With rudimentary cavity      b. Without rudimentary cavity</p>		<p><b>Class U5/Aplastic Uterus</b></p> <p>a. With rudimentary cavity      b. Without rudimentary cavity</p>
<p><b>Class U6/Unclassified Cases</b></p>		

# Gynecological examination

- Some vaginal and some cervical malformations (aplasia, double cervixes, longitudinal septa reaching to the external cervical os) can be diagnosed objectively by inspection.
- Palpation (through the vagina and/or the rectum in cases of vaginal aplasia) cannot provide information for the uterine cavity and uterine wall and it could provide only some useful, but highly subjective, information for the uterine body.
- It is always the starting point and an essential part of any woman's clinical evaluation.
- Careful inspection of the vulva, vagina and vaginal vault with a speculum to establish the presence of one or more cervical body(ies) or one cervical body with one or two external cervical opening(s).

# Imperforate hymen (and hematocolpos)

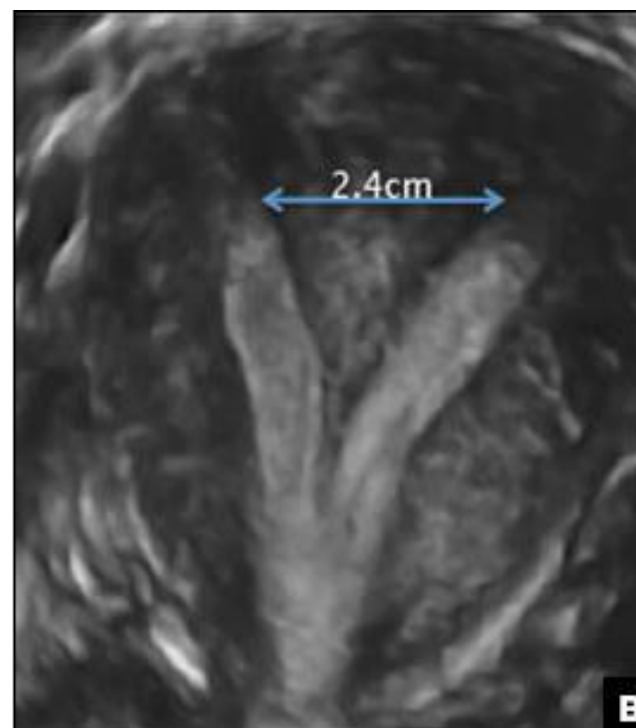


# Two-dimensional ultrasound

- It is non-invasive, simple, low cost, available in almost every setting.
- It could provide reliable, objective and, most importantly, measurable information for the anatomy of the cervix, the uterine cavity, the uterine wall, the external contour of the uterus.
- It could provide useful information of associated pelvic pathology, e.g. ovarian pathology (benign and malignant tumors, endometriosis), hydrosalpinges, renal anomalies
- It may provide information on the vaginal cavity, especially in the presence of imperforate hemivagina.



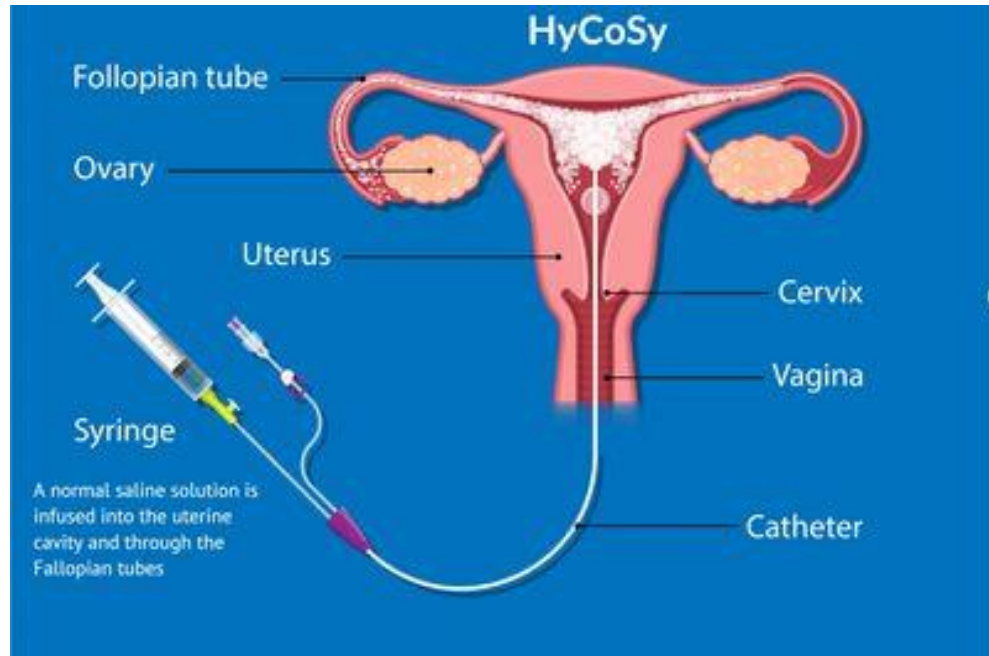
# Bicornual uterus and uterine septum (2D-US)



# Hysterosalpingo-contrast-sonography






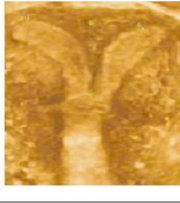




- It can provide reliable, objective and, most importantly, measurable information for the anatomy of the cervix, the uterine cavity, the uterine wall, the external contour of the uterus and for other peritoneal structures (e.g. ovaries) with the exception of tubes.
- The imaging of the uterine cavity is better owing to the use of the contrast medium or saline, enhancing the accuracy in identifying uterine cavity defects.
- Hysterosalpingo-contrast-sonography (HyCoSy) also could be used as a tubal patency test

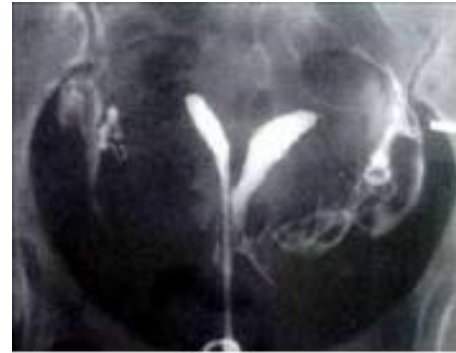
# HyCoSy



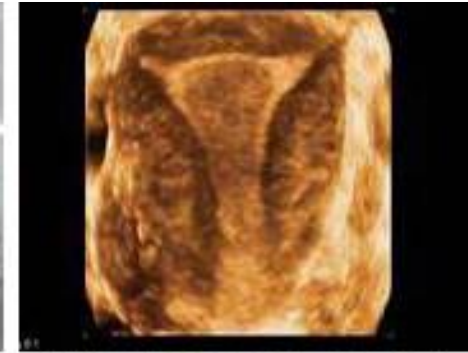
# Three-dimensional ultrasound (3D)

- It can provide highly reliable, objective and, most importantly, measurable information for the anatomy of the cervix, the uterine cavity, the uterine wall, the external contour of the uterus and for associated pelvic pathology.
- Diagnosis of associated vaginal anomalies can be achieved by transperineal acquisition of the pelvic floor volume after filling the vagina with gel or saline
- Not so widely available as 2D US (up to now). Need for experienced sonographers.

Class U0, or Normal Uterus	
	
Class U1, or Dysmorphic Uterus	
	
Class U2, or Septate Uterus	
	
Class U3, or Bicornuate Uterus	
	
Class U4, or Hemim Uterus	
	



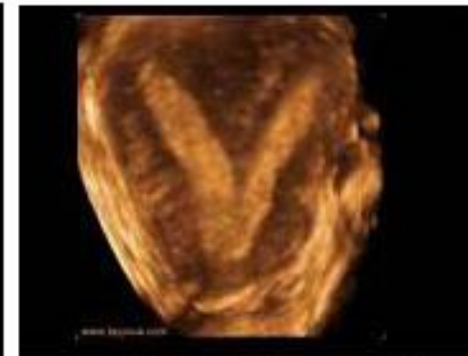
HSG



3D ultrasound - normal



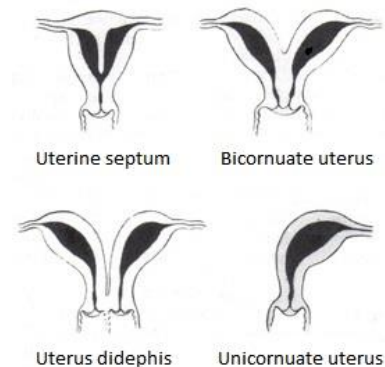
3D ultrasound - arcuate



3D ultrasound - septum

# X-ray hysterosalpingography

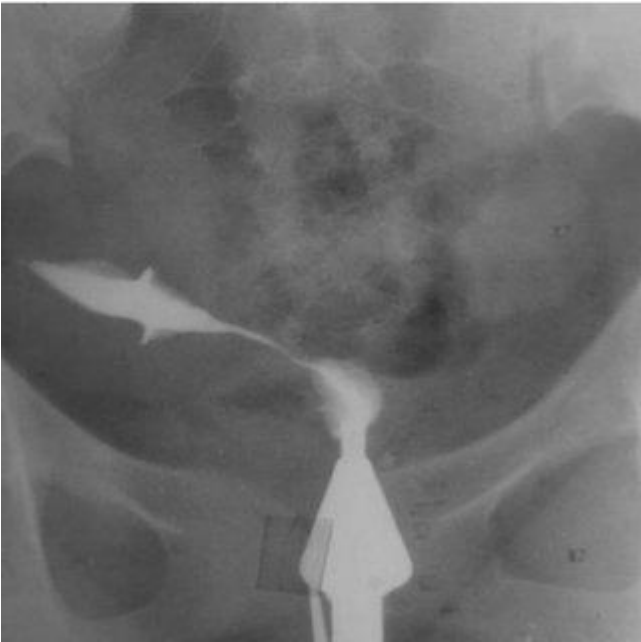
- It provides some reliable information for the anatomy of the uterine cavity in the absence of cervical obstruction.
- It offers additional useful information in cases of infertile women for potential intra-cavitary pathology and tubal morphology.
- It cannot be used for the diagnosis of obstructing anomalies.



Bicornuate uterus

Hemicornuate uterus

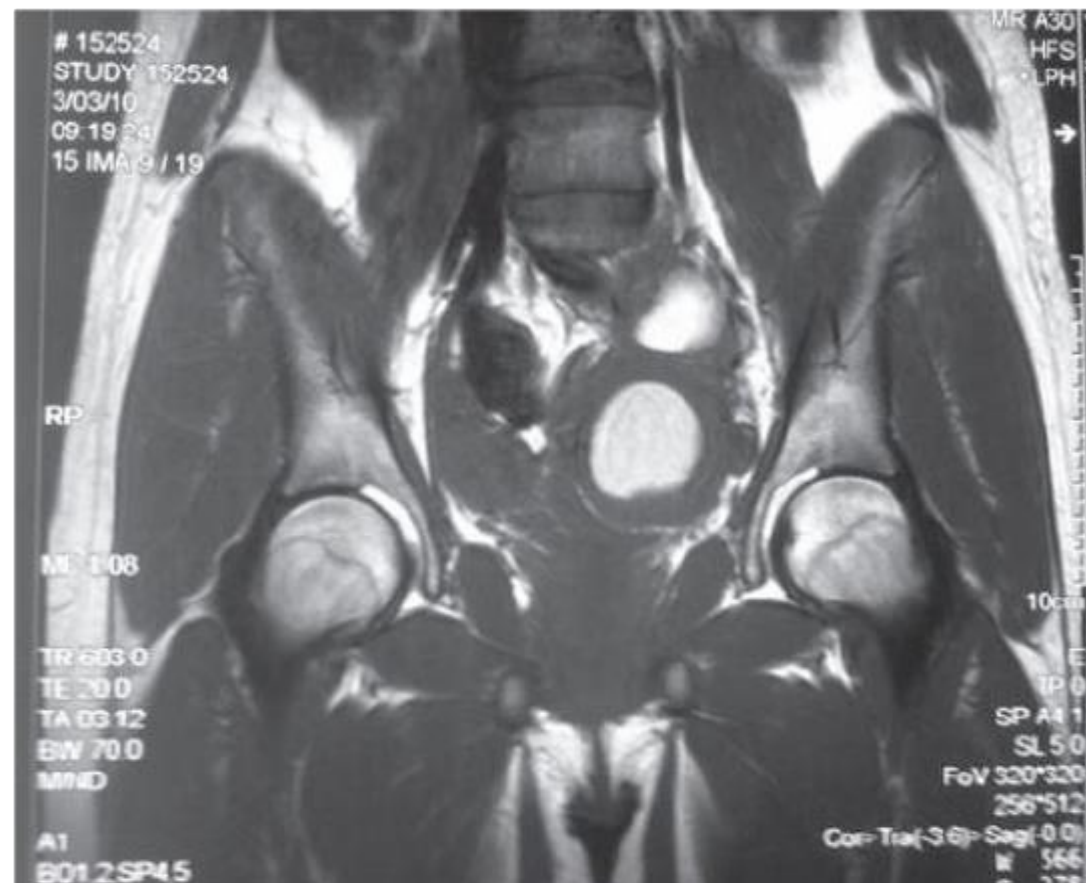
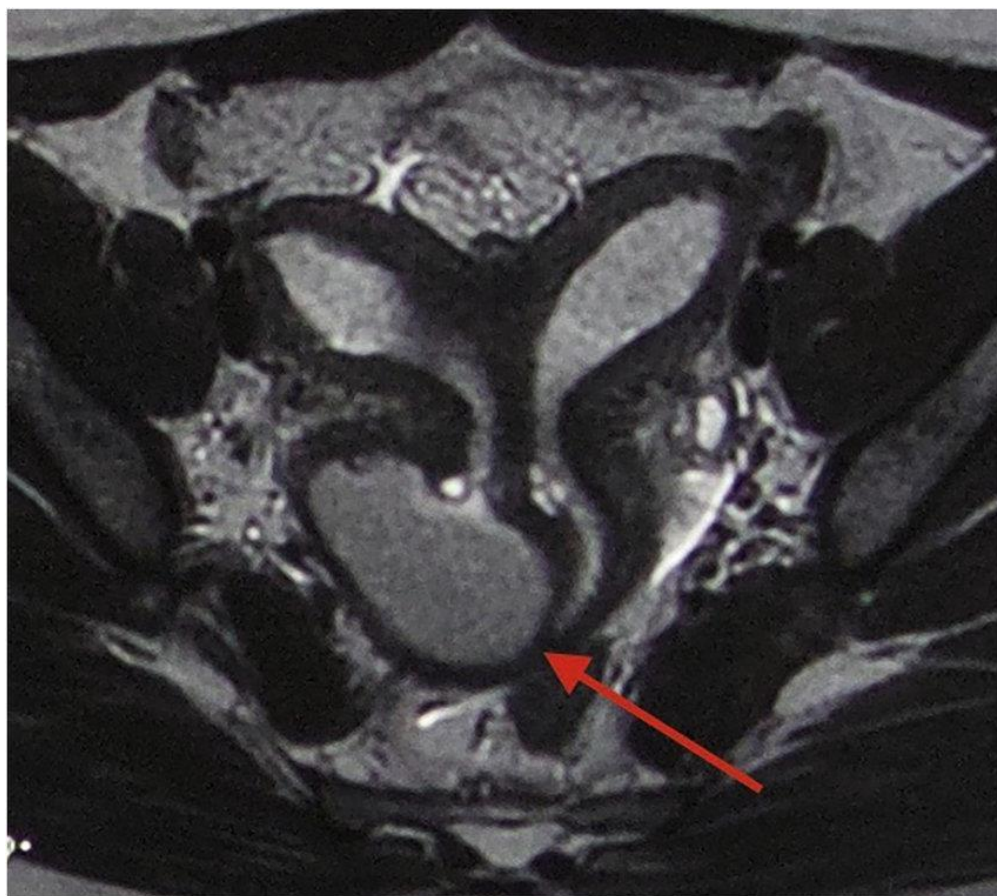
(X-ray HSG)





# Magnetic resonance imaging (MRI)

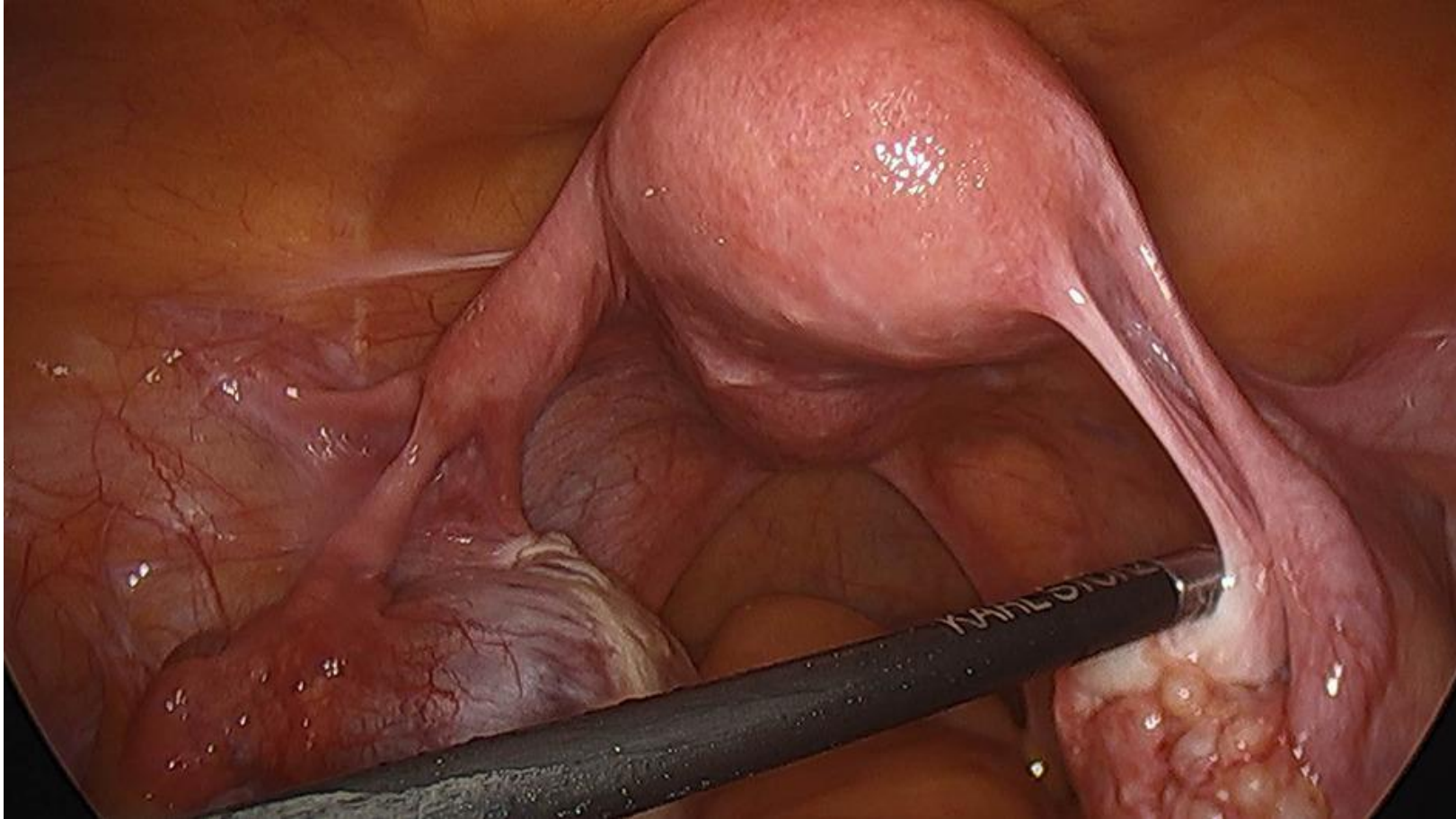
- Advantages. It is non-invasive and it has no radiation.
- Disadvantages. More expensive and less available than ultrasound.
- It can provide highly reliable and objective information for the anatomical status of the vagina, cervix, the uterine cavity, the uterine wall, the external contour of the uterus and for other peritoneal structures with the exception of tubes



# Endoscopy: laparoscopy and hysteroscopy

- It provides highly reliable information for the anatomical status of the vagina (vaginoscopic approach), the cervical canal, the uterine cavity, the tubal ostia, the external contour of the uterus and the intraperitoneal structures.
- Direct visualization of the cervical canal, endometrial cavity and the external contour of the uterus representing, until now, the 'gold standard' in the diagnosis and differential diagnosis.
- Disadvantages. It is invasive. No objective estimation of the uterine wall thickness.
- The invasiveness of the laparoscopic approach makes it not acceptable as a first line screening procedure; it complements indirect imaging in the diagnosis of more complex anomalies in combination with possible surgical actions. It offers supplementary information about partial or total absence of Fallopian tubes and abnormal localization of ovaries.

# Right unicornuate uterus (laparoscopic view)



# Doubled uterus (uterus didelphys)





# Uterine septum (hysteroscopic view)



## Techniques for müllerian aplasia (vaginal agenesis)

- Complete müllerian aplasia (Mayer-Rokitansky-Kuster-Hauser syndrome) is the most common variant encountered and it is characterized by congenital absence of the vagina and the uterus in 90-95% of cases. The fallopian tubes are normal, and the ovaries have normal endocrine and oocyte function
- The first-line approach in most patients should be primary vaginal elongation by dilation. Compared to surgery it is safer, patient-controlled, and more cost effective.
- The aim of surgical treatment is to create a neovagina. Skin grafting remains the most popular material used in vaginoplastie



# Surgical technique for rudimentary horn excision

- The indication for surgery is the presence of endometrium in the accessory horn.
- Laparoscopic hemihysterectomy of the rudimentary horn is the treatment of choice.
- Surgical treatment is not indicated when the rudimentary horn lacks an endometrium.

# Didelphys uterus

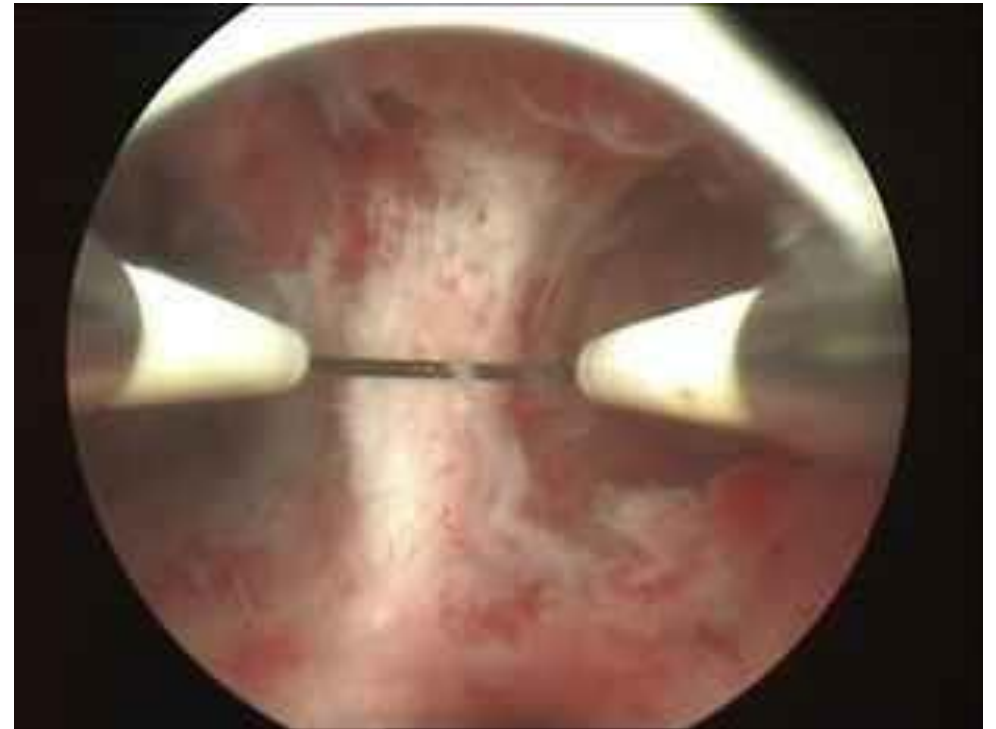
- Arises when midline fusion of the müllerian ducts is arrested, either completely or incompletely.
- Approximately 11% of uterine malformations are didelphys uterus
- Patients with a uterine didelphys are usually asymptomatic, unless an obstruction is present
- a. Uterine didelphys with obstructed unilateral vagina: Full excision and marsupialization of the vaginal septum is the preferred approach and is performed as a single procedure. After the septum has been excised, laparoscopy can be performed for potential treatment of associated endometriosis, adhesions, or both
- b. Uterus didelphys, nonobstructed: indications for septum resection in the nonobstructed didelphys uterus are limited.

# Bicornuate Uterus

- The bicornuate uterus is formed when the müllerian ducts incompletely fuse at the level of the uterine fundus
- Bicornuate uterus seldom requires surgical reconstruction
- The Strassmann procedure is the surgical treatment of choice for unifying the bicornuate and didelphys uteri

# Septate uterus

- The most common structural abnormality of all müllerian duct defects.
- It results from incomplete resorption of the medial septum after complete fusion of the müllerian ducts has occurred.
- The septum, located in the midline fundal region, is composed of poorly vascularized fibromuscular tissue.
- The surgical procedure of choice is hysteroscopic metroplasty with concurrent laparoscopy



# Conclusion

- Müllerian anomalies are a morphologically diverse group of developmental disorders that involve the internal female reproductive tract.
- Establishing an accurate diagnosis is essential for planning treatment and management strategies.
- The surgical approach for correction of müllerian duct anomalies is specific to the type of malformation and may vary in a specific group. For most surgical procedures, the critical test of the procedure's value is the patient's postoperative ability to have healthy sexual relations and achieve successful reproductive outcomes.

**THANK YOU!**

