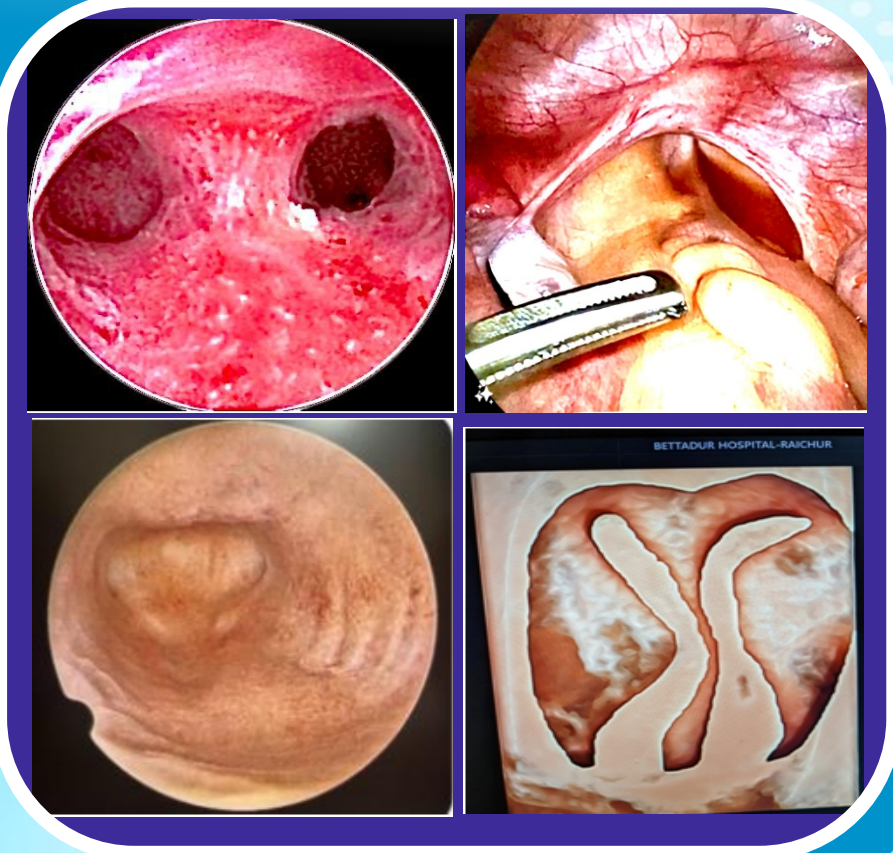
KSOGA UPDATE

ENDOSCOPIC COMMITTEE

April - 2024



INSIGHT AND INTERVENTION NAVIGATING FEMALE REPRODUCTIVE TRACT ANOMALIES

3rd Edition

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Chairperson, Endoscopic Committee

Dr. Sonia Batra
Hony. Secretary, Endoscopic Committee

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Dr. Vidya Thobbi


Dr. Rajashree Paladi


Dr. Sandesh Kade


Dr. Jayaprakash Patil


Dr. Subhash Mallya

PATRON ADDRESS

Dr. M G Hiremath
Chief Patron
KSOGA & KCOG

Dear Esteemed Members of the Endoscopic Committee,

I am writing to you today not just as the Patron of the Karnataka State Obstetrics and Gynaecology Association (KSOGA) but also as an ardent supporter of the continuous progress in medical science, especially within the field of obstetrics and gynaecology. It is with immense pride and satisfaction that I extend my heartfelt congratulations on the successful publication of the third edition of the Endoscopic Committee News Bulletin.

Under the exemplary leadership of Dr. Ravikanth G O as Chairperson and Dr. Sonia Batra as Secretary, and with the guidance of our Chief Adviser, Dr. Ramesh B, and Master Trainer, Dr. Vidya Bhat, this edition has emerged as a pivotal resource. Covering the intricate topics of "Female Reproductive Tract Abnormalities: From Embryonic Development to Classifying, Diagnoses, Hysteroscopy, and Laparoscopic Management," it offers profound insights and knowledge indispensable to our fraternity.

The collaborative effort of our twelve distinguished authors has not just contributed to the expansion of our understanding but has also significantly uplifted the quality of care we can provide to our patients.

I would also like to express my gratitude to Dr. Vidya Thobbi, our KSOGA President, Dr. Rajashree Paladi, our Secretary, and Dr. Suman Gaddi, our Treasurer, for their unwavering support towards the endeavors of the Endoscopic Committee.

The dedication, expertise, and tireless work you all have displayed in bringing this edition to life exemplify the spirit of KSOGA—commitment to excellence and leadership in medical education and patient care.

As we share this invaluable resource with our members and the broader medical community, let us celebrate this achievement and look forward to more such innovative and educational initiatives led by the Endoscopic Committee.

Once again, congratulations on your remarkable work, and thank you for setting a sterling example of professional dedication and excellence.

Warm regards,
Dr. M G Hiremath
Patron, Karnataka State Obstetrics and Gynaecology Association (KSOGA)

PRESIDENTIAL ADDRESS

Dr. Vidya Thobbi
President,
Karnataka State Obstetrics and Gynaecologists Association

Dear Members,

As the President of the KSOGA, I am thrilled to share some remarkable achievements and our steadfast progress in the realm of gynecological endoscopy. Our journey, marked by unwavering dedication and an insatiable quest for excellence, continues to enrich our field with invaluable insights and advancements.

Under the esteemed guidance of our Chief Advisor, Dr. Ramesh B, we have navigated through the complexities of endoscopic surgery with unparalleled expertise and foresight. His vision has been the cornerstone of our committee's accomplishments.

I am especially proud of the leadership showcased by Dr. Vidya Bhat, our Master Trainer, and Dr. Ravikanth, the Chairperson of our committee, during the successful conduct of the Hysteroscopy workshop at the state conference in Vijayapura in association with FOGSI Endoscopic Committee Chairperson Dr Subhash Mallya . Their collaborative efforts have set a new benchmark for skillful execution and knowledge dissemination in our community.

Our Secretary, Dr. Sonia Batra, has been instrumental in bridging communication within our committee. Her commitment ensures we remain informed and cohesive in our endeavours.

The Endoscopic Committee has been active in extending our knowledge beyond the operating room. Our webinars, led by experts like Dr. Sandesh Kade, have covered crucial topics such as Urinary Tract Endometriosis, Cesarean Scar Pregnancy, and innovative approaches to Safe portal Entry,Myomectomy and Cystectomy. These sessions have played a pivotal role in enhancing our understanding and skill set in treating complex conditions.

Today, we stand on the precipice of another milestone — the release of the third edition of our News Bulletin. This edition focuses on Female Reproductive Tract Anomalies, encompassing embryology, classification, diagnosis, imaging, systematic approaches, and the surgical management via Hysteroscopy and Laparoscopy. Furthermore, it emphasizes the importance of psychological counselling for patients and their families, underlining our commitment to holistic care.

This bulletin is a testament to our collective wisdom, experience, and the relentless pursuit of perfection. It is a resource that promises to enlighten, guide, and inspire. Let it serve as a beacon, illuminating our path towards excellence and empathy in patient care.

Your engagement, feedback, and contributions are vital to our growth and success. Together, let us continue to elevate the standards of gynecological endoscopy and make a significant impact on the lives we touch.

Dr. Vidya Thobbi
President,
Karnataka State Obstetrics and Gynaecologists Association

GENERAL SECRETARY ADDRESS

Dr. Rajshree Paladi
Honorary Secretary
KSOGA 2023-2024

Greeting's to Everyone!

Dear Endoscopic Committee of KSOGA,

I am writing to extend my heartfelt congratulations and commendation to the entire Endoscopic Committee of KSOGA for the remarkable evolution in endoscopic techniques witnessed under your guidance and expertise.

The strides you've made in advancing endoscopic procedures have not gone unnoticed.

The dedication to refining techniques, enhancing patient outcomes, and pushing the boundaries of innovation in the field of endoscopy is truly commendable.

The commitment to staying at the forefront of technological advancements and unwavering pursuit of excellence are exemplary.

The impact of the efforts resonates not only within the medical community but also in the lives of countless patients who benefit from the expertise and dedication.

As a Hon Secretary of Ksoga , I am inspired by achievements and the standard of excellence the Endoscopic committee upholds .

A continuous pursuit of improvement sets a benchmark for others to strive towards.

Do accept my sincerest appreciation for the tireless work and dedication to advancing endoscopic techniques.

The contributions to the field are invaluable, and I eagerly anticipate witnessing further breakthroughs .

With utmost wishes and admiration,

Best regards,

Dr. Rajshree Paladi
Honorary Secretary, KSOGA 2023-2025

EDITORIAL

Dr Vidya Bhat
Secretary

Hard work and perseverance is the key to success. Change is constant and if you want to see change, you be the change.

Endoscopy committee KSOGA is working hard to bring in awareness on Endoscopy pan Karnataka, this is our 3rd bulletin and I thank all the committee members for their dedication and laud them for all the good work.

We have always thought of challenging and confronting topics in Endoscopy which is difficult in decision making for practitioners. Hysteroscopy, Electrosurgery and now mullerian anomalies.

Mullerian Anomalies are seen in 4 percent of the female population. Apart from the diagnostic dilemmas. There is confusion in the management of mullarian anomalies. In this booklet, we are bringing the nuances in the management and enlightening everyone to correct treatment of it

Mullerian Anomalies is a vast topic, in this bulletin we have tried our level best to include all topics which need endoscopic corrections and in turn give the best outcome to our patients

I thank all the faculty who have contributed to this bulletin for all their efforts.

My sincere thanks to Dr. Ravikanth for his dedication and perseverance. My heartfelt thanks to M. G Hiremath Sir, Dr. Vidya Thobbi, Dr Rajashree Palladi, Dr. Hema Divakar, Dr Muralidhar Pai for their encouragement and constant support.

"Harmony in teamwork fulfills dreams."

Regards,

Dr. Vidya V Bhat

" Team work makes the dream work "

Bang Gae

CHAIRPERSON'S ADDRESS

Dr. Ravikanth G. O
Chairperson, Endoscopic Committee,
Karnataka State Obstetrics and
Gynaecologists Association

Dear Members and Esteemed Colleagues,

I seek blessings from our patron of KSOGA Dr M G Hiremath sir and Dr Nagaraj sir who always encouraged me in all aspects, Dr Vidya Thobbi madam president of KSOGA, Dr Rajashree Paladi madam Secretary of KSOGA and Dr Suman Gaddi madam treasurer of KSOGA have always supported in all our academic activities, Dr Ramesh B Sir the Chef Adviser of Endoscopic committee and Dr Vidya Bhat the Master trainer of our committee has guided me in all aspects

It gives me immense pleasure and pride to announce the release of our latest news bulletin titled "Insights and Interventions: Navigating through Female Reproductive Tract". This comprehensive bulletin is a testament to the dedication and expert knowledge within our community, particularly the Endoscopic Committee of KSOGA, which I have the honor of chairing.

Over the past few months, our committee has been dynamically engaged in enriching the field through various initiatives, including a hysteroscopy workshop led by Dr. Vidya Bhat, and Dr. Subhash Mallya, and a series of thought-provoking webinars. These webinars covered critical topics such as urinary tract endometriosis presented by Dr. Sandesh Kade, cesarean scar pregnancy in association with the Safemotherhood Committee, which was very exhaustive and had eight speakers and was very successful and a mixed bag session on safe port entry, myomectomy, and laparoscopy which was well spoken by Former Chairperson of Endoscopic Dr Jay Prakash Patil ,Secretary of Endoscopic Committee Dr Sonia Batra and our active member Dr Santhosh Rathod

Our forthcoming news bulletin is a culmination of these efforts, showcasing articles penned by distinguished contributors. These articles delve into pivotal aspects of female reproductive health, from the embryological origins and classification of anomalies to diagnosis, management, and psychological support for affected individuals and their families.

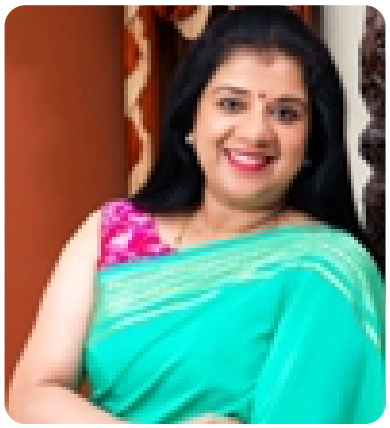
I am proud to have contributed an article on "Decoding the Origin: Embryology of Female Reproductive Tract". The bulletin also features invaluable insights from renowned specialists such as Dr. B M Keerthi on classification of anomalies, Dr. Raju Giradi on diagnostic imaging, Dr. Sunil Eshwar on differential diagnosis, and Dr. Santhosh Rathod on systemic diagnostic approaches. Dr. Prema D Cunha has eloquently addressed the psychological implications and support mechanisms, while Dr. Ramesh B sir, the backbone of our committee, has offered strategic management insights for Müllerian agenesis. Furthermore, Dr. Vidya Bhat ,The Master trainer of our Endoscopic committee madam has written on guidance on managing Mülerian anomalies through hysteroscopy, Dr Rajesh Bhakta the main trouble shooter of our Committee has written on optimal management strategies. Dr. Beeresh C S discusses optimizing fertility outcomes in reproductive tract anomalies, Dr Prabha Desai, always go to person in need has written on Understanding endometriosis in within the spectrum of Mullerian anomalies, Dr Jay Prakash Patil former chairperson of Endoscopic committee has written on Decisive dilemma: Optimizing management of operable and inoperable reproductive tract anomalies

This bulletin stands as a cornerstone of our collective wisdom and commitment to advancing gynecological endoscopy and women's health. We believe it will serve as an invaluable resource for practitioners, researchers, and academicians alike.

I extend my heartfelt gratitude to all contributors and supporters who made this achievement possible. Let us continue to collaborate, innovate, and lead with compassion and expertise in our journey towards bettering women's health care.

With warm regards,
Dr. Ravikanth G.O
Chairperson, Endoscopic Committee, Karnataka State Obstetrics and Gynaecologists Association

EDITORIAL TEAM



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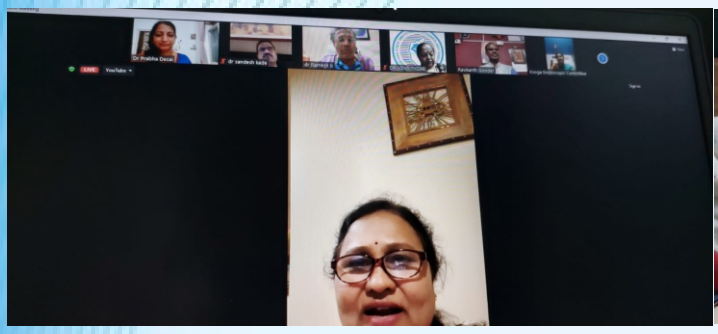
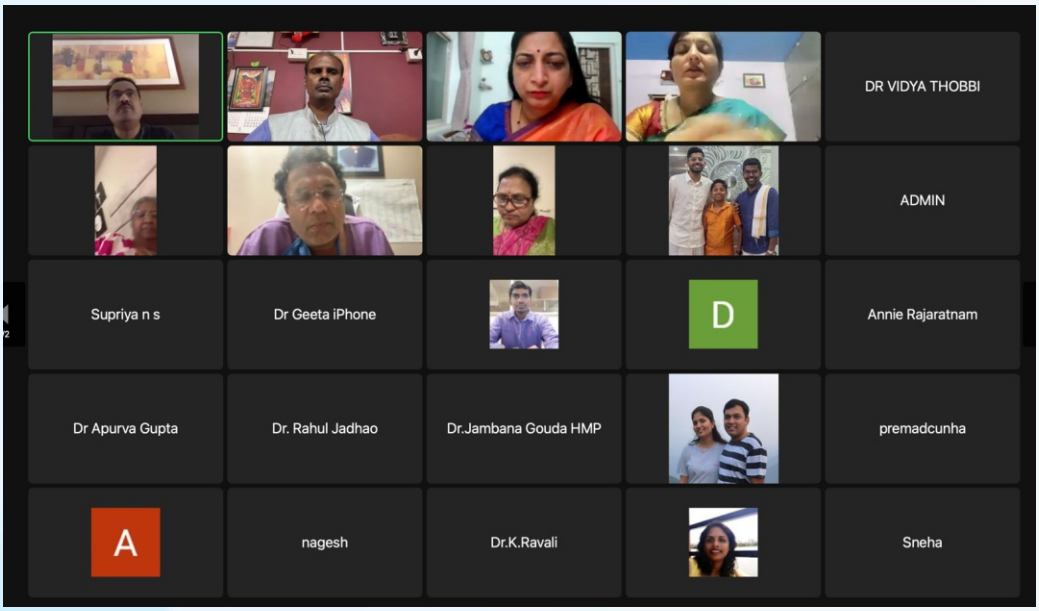
KSOGA

Webinar on Hidden battle:

In Association with unravelling the Enigma of Urinary tract Endometriosis was done by Dr Sandesh Kade on 4th November 2023It was very well appreciated by Dr M G Hiremath sir and DrRamesh B sir





Take home message of 'Hidden Battle Unravelling the enigma urinary tract endometriosis'

1) 10% women are affected by endometriosis

2) 180 million through out the world are suffering from endometriosis

3) 20-40% of pelvic endometriosis are suffering from urinary tract endometriosis

4) No specific symptoms of urinary tract involvement

5) Bladder is commonest site

6) Isolated urinary tract involvement is rare but possible

7) Nodular endometriosis at uterosacral ligament-think of ureteral involvement in 90% of cases

7) Treatment should be multidisciplinary aspect- involve urologist in these cases

8) Rectum is resistant to invasion by endometriosis when compared to bladder

9) Trigonal endometriosis should be careful

10) MRI helps in pre operative diagnosis and planning

11) Hydronephrosis means 30% of kidney is damaged

12) Most of the time ueretaral involvement is external -Fortunately

13) Uterosactral involvement- Hygpgastrict nerves are involved- urinary retention in the postoperative- Counsel for self catheterisation if there is bilateral involvement

14) Urodynamic studies is necessary if bilateral uterosacral involvement

15) TVS Bladder involvement

16) MRI more specific for smaller leasions

17) CT Urogrogram MRI Urogram useful in severe endometriosis

17) DMSA- Dexa scintigraphy shows Kidney function

18) Minimum 10% function should be there to retain the kidney

19) Bladder lesions - excisions

20) URETEROLYSISI as it is involved 60% of cases it's extrinsic

21) RARELY REIMPLEMENTATION but if it's intrinsic then re implantation

22) Barbed sutures give uniform tension and better results

23) Trigonal involvement is challenging-Rotational flap to prevent VVF(Dome of the bladder is brought down & omentum between the bladder and vagaina)

24) URTEROLYSIS very careful to avoid damaging adventia of ureter

25) MRI Enzian scores are helpful in planning the surgery

26) Sheaths of endometriosis encroaching the ureter should be released

27) Sometimes the ureter is compressed distal to uterine artery in the Ureteric tunnel here maximum chance of fistula formation Rap the omentum to support it after careful dissection

28) Uretero uretoretic anastomosis is easy to perform but more chances of fistula so reimplementation is preferred with boari flap

29) Nephrectomy if kidney function is less than 10% function is present by DMSA

30) PRIMARY COLOSTOMY rarely necessary unless low rectum is involved

31) Recurrence after surgery is possible some form ovarian suppression (continuous with Dienogest or ocp till menopause)unless patient wants to have pregnancy

32) Regular fallow up is necessary for these cases of conservative surgery

33) Calcium supplements and bone scan for those who are on long standing suppression to prevent osteoporosis

34) LnG IUS is only for mild endometriosis and adenomyosis

35) No serum markers are reliable

36).Salivary diagnostic test are under study

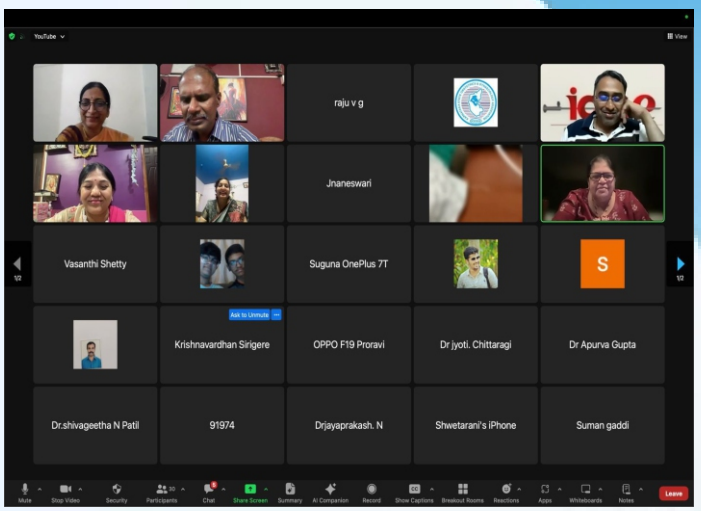
37) Till Specific treatment is available suppression of ovaries and excision of endometriosis is only available treatment

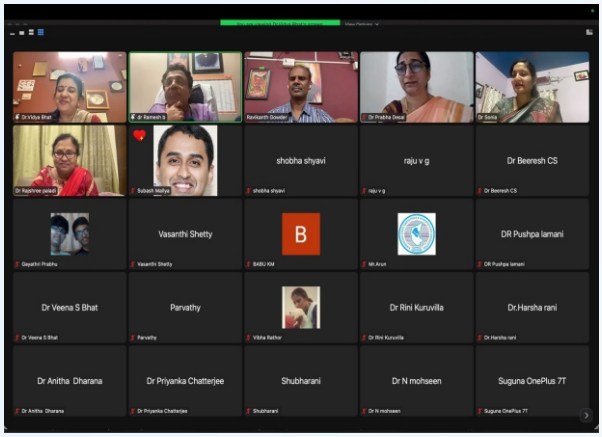
38) 50-60 % of all these patients conceive naturally

39) IVF is needed in 30% of cases

SKILL TRANFER PROGRAM with Endotrainer to the postgraduate students was organised at KSOGA Vijayapura 2023

Webinar on Cesarian scar pregnancy with safe motherhood committee KSOGA Multifaceted approach to Diagnosis and management with Eight experts on 24/01/2024It was attended by highest number of audience and very well appreciated





Salient points of the webinar on CAESAREAN SCAR PREGNANCY.

Conducted by Endoscopic Committee in association

with

Safe Motherhood committee KSOGA.

It was discussed on these headlines.

- PATHOPHYSIOLOGY
- CLASSIFICATION
- DIAGNOSIS
- Differential diagnosis
- Hysteroscopic management
- Laparoscopic management
- PRIMARY PREVENTION

PATHOPHYSIOLOGY

- ✓ Incidence 1 in 1800 to 2226 of overall pregnancy
- ✓ Not an ectopic since within boundaries of uterus
- ✓ CSP is the pregnancy seen On scar defect or dehiscence, may result in live newborn
- ✓ May continue to Presents in 2^{nd}/3^{rd} trimester pregnancy .
- ✓ Most probable mechanism is Blastocyst drops down into the lower part of the uterus and implantation of the blastocyst on the microtubular tract of the caesarean scar defect.
- ✓ In the scar defect area, there is deficiency of the nitabuch layer which prevent the deeper invasion of the trophoblast into myometrium which Leeds to placenta acreta syndrome disorder
- ✓ Such a defect can develop through any trauma caused to uterus.
- ✓ Risk of scar implantation might be proportional to the size of anterior uterine wall defect possibly due to larger surface area induced by the scar.
- ✓ Iatrogenic – due to scarred uterus after myomectomy/caesarean section/curettage/manual removal of placenta/hysteroscopy/metroplasty.
- ✓ Cesarean section done for Prolonged labour / dialated cervix --more chance
- ✓ An incision on the upper segment of the uterus and Single layer uterine closure
- ✓ Impaired woung healing
- ✓ Post operative adhesions
- ✓ Retroverted uterus can cause scar defect
- ✓ Some additional factors :-

  Nulliparity
  Diabetes
  Emergency surgery
  Infection

- ✓ Terminologies

  RMT (Residual Myometrial Thickness)- thickness of the anterior myometrium at the site of the scar.
  AMT (Adjacent myometrial thickness)- Thickness of anterior myometrium adjacent to c- section site.

  NICHE (ISTHAMOCELE) Defect in the scar area >2 mm but RMT is >5mm.
  CESEAREAN SCAR DEFECT – Deeper gutter formation in the scar area but the RMT is 50 -80% less than the AMT.

- ✓ Timor-Tritsch et al told all patients with Placenta Accreta Syndrome (PAS) had hysterectomy ?
- ✓ Zoosmer, Noel, Poomski told patients with PASD, 50-100% patients underwent hysterectomy ?

CROSS OVER SIGN (COS) :-

- ✓ Relationship between the gestational sac of the CSP, analyzed by new sonographic sign, the COS.
- ✓ In a sagittal view of the uterus, a straight line was drawn connecting the internal cervical os and the uterine fundus through the endometrium (endometrial line)
- ✓ The gestational sac was identified and its superior–inferior (S–I) diameter, perpendicular to the endometrial line, was traced.
- ✓ **COS 1** the gestational sac was implanted within the Cesarean scar, gestational sac was above the endometrial line, towards the anterior uterine wall
- ✓ **COS 2** Gestational sac was BELOW the endometrial line, further divided in two different categories (COS-2+) if the less than 2/3 of gestation sac is below the endometrial line or (COS-2-) more than 2/3 of the G sac is below the endometrial ine.

CLASSIFICATION

- Type 1 – Endogenic CSP, pregnancy grows inside uterus
- Type 2 - Endogenic CSP, pregnancy grows outwards, i.e anterior surface of the uterus but there is RMT>3mm.
- Type 3 - Endogenic CSP, pregnancy grows outwards with no identifiable RMT.

2017 Classification

- Type I / Endogenic variety – placenta attatched at c-scar and good RMT, gsac grows towards cervico-isthmic or uterine cavity.
- Type II / Exogenic variety – Blastocyst embedded in niche, RMT reduced, grows towards bladder.

January 2024, Rohangui et.al.classification

- Type 1- Closure type
  1a- pregnancy grows towards the uterus
  1b- grows towards cervix.
- Type 2- Implantation type
  2a RMT >0.2cm
  2b RMT <o.2 cm >0.1cm
- Type 3 – Infiltration type
  3a RMT <1mm
  3b no RMT

Common CLINICAL FEATURES

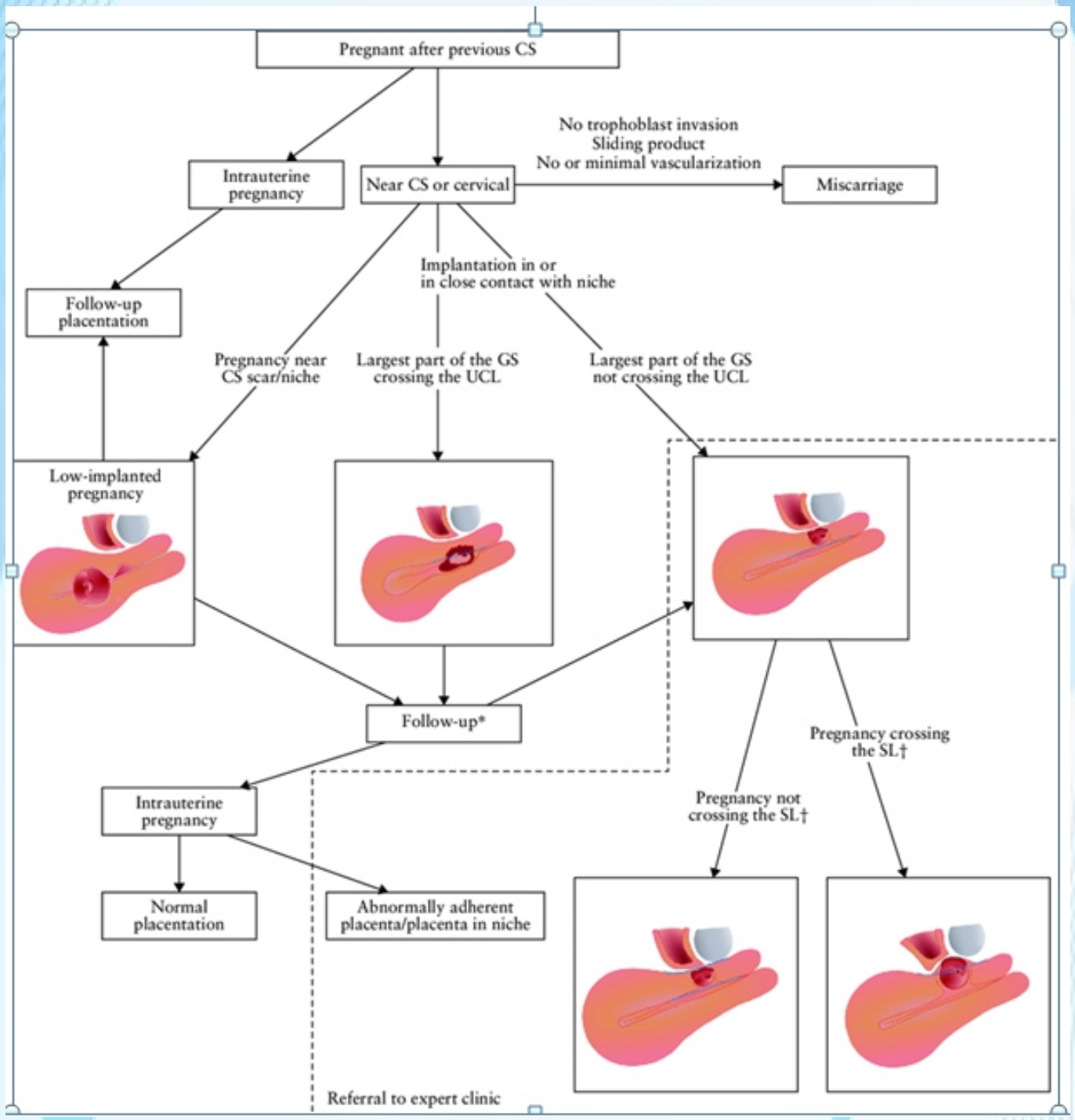
✓ CSP may present as early as 5-6 weeks to as late as 16 weeks
✓ Painless vaginal bleeding +/- abdominal pain
✓ Asymptomatic or only abdominal pain
✓ Severe acute pain with profuse bleeding implies an impending rupture
✓ Collapse or haemodynamic instability strongly indicates a Ruptures CSP.

DIAGNOSIS

✓ UPT – Positive
✓ **SONOGRAPHIC CRITERIA –**
Empty uterine cavity and endocervix.
Gsac and placenta embedded in LUS c-scar.
Sac appears oblong or triangular in shape.
Thin(1-3mm) or absent myometrium.
Colour doppler shows increased vascularity
Presence of placental lacunae predicts Placenta Accreta Syndrome
Circular peritrophoblastic perfusion surrounding gsac tells relation of placenta to scar and bladder.
Sliding sign- On putting gentle pressure over niche with probe, there is no sliding of the pregnancy
✓ TVS is better for seeing scar thickness.

MANAGEMENT

- Expectant
- Medical with injection methotrexate
- Surgical
- Curettage(obsolete)
- Laparotomy
- Vaginal approach
- Laparoscopy
- Hysteroscopy



	CSP	CERVICAL ECTOPIC	FAILED PREGNANCY
GSAC	Anterior LUS	Within cervical canal	Within cervical canal
Overlying anterior myometrium	Thin	Normal	Normal
Sliding organ sign	Negative	Positive	Positive
Doppler	Marked peritrophoblastic colour doppler around gsac	Vascular flow around & within gsac	Lack colour flow
Short follow up	Growing	Growing	Not fixed in location, not growing

SUCTION CURETTAGE

- ✓ Done under TAS/TVS guidance
- ✓ Plan with possible interventions to control bleeding:-
  Intracervical balloon tamponade
  Angiographic embolization
  Cervical encirclage
  Ligation of uterine arteries
  Local hysteroscopic endocervical resection of the gsac
- ✓ C/I unruptured

COMBINATION OF SYSTEMIC AND LOCAL INJECTION OF METHOTREXATE

- ✓ Timor-Tritsch Recommendation of combined local and systemic injection
  a) 25mg methotrexate intragestational sac
  b) 25mg methotrexate placental site as the needle is being withdrawn
  c) 25mg methotrexate intramuscular prior to patient discharge from hospital.
- ✓ University of Illinois at Chicago protocol
  a) Half dose of 50mg/m2 BSA injected into the intragestational sac
  b) Remaining half dose intramuscular.

HYSTEROSCOPY

- ✓ 5-10ml vasopressin can be given while doing hysteroscopic removal
- ✓ Use energy only if it is necessary
- ✓ Reverse cutting can also be done
- ✓ Using resectoscope can release the sac in bits & pieces.
- ✓ If red flag sign seen- wait for clearance
- ✓ If RMT is 3-5mm, can do hysteroscopically
- ✓ If ventrally uterus is suspended, attatched to anterior abdominal wall, do Cystoscopy too !
- ✓ Short operative time
- ✓ Rapid reduction in b-hcg levels to baseline ~ 30 days
- ✓ Less hospital stay.

LAPAROSCOPY

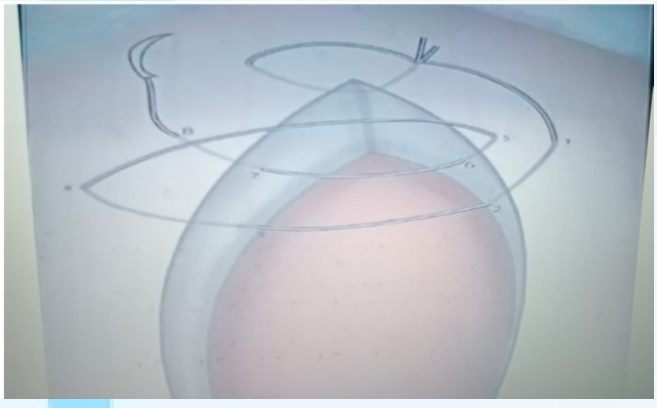
- ✓ CONSIDERATIONS
  Patient should be haemodynamically stable
  Desire for future fertility
  Compliance
- ✓ CSP Location
  Gestational age
- ✓ Surgical expertise
- ✓ Best suited in type 2 CSP
- ✓ STEPS:-
  Laparoscopic hysterotomy with wedge resection of CSP and previous scar
  Temporarily occluding blood supply to uterus- decreases blood loss
  -Enables complete resection.
  CSP mass incised and products removed in endobag
  Vasopressin locally (1 unit/ml,5-10ml); bipolar diathermy; endoscopic suturing can be used to control bleeding.
- ✓ RMT decides the management approach
  >3mm – 1st optimal surgical approach
  <3mm – use of mesh, undertrial
  Better to put stent/foleys for 2-3 weeks, to prevent adhesions/stenosis.
- ✓ Freshen the margins and close uterus in 2 layers, can put intercede to prevent adhesions.
- ✓ Good to combine with methotrexate.

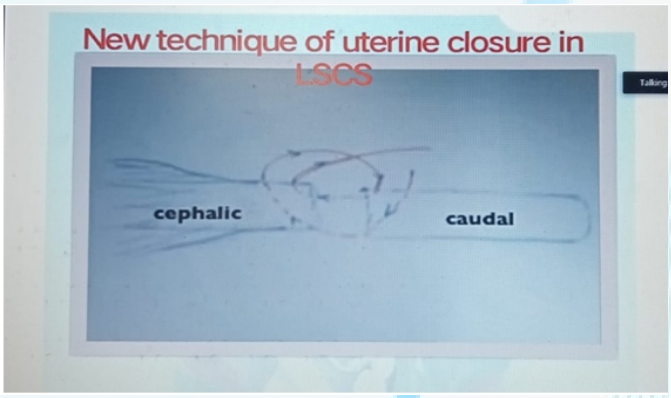
ADVANTAGES :-

- ✓ Minimally invasive
- ✓ Direct visualization
- ✓ Removal of the scar
- ✓ Success rate 97%
- ✓ Faster resolution of b-hcg
- ✓ Long term outcomes

SUTURE TECHNIQUES to prevent caesarean scar defect

- Things to remember during lscs, to prevent CSP.
  - ✓ Modified Mattress Suture
  - ✓ 1st at junction of decidua and myometrium
  - ✓ 2nd serosa and subserosa(without including the decidua)
  - ✓ Tighten the deep loop and give traction to the deep bite.
  - ✓ Double breasting technique – for thinned out LUS.





- Things to do while operating for CSP
  - ✓ Insert a dilator inside the cervical canal to better approximation of both upper and lower part of the anterior wall
  - ✓ Round ligament plication to prevent retroversion
  - ✓ Usually diverticula will be seen over right side of uterine closure of LSCS
  - ✓ Counsel patient not to conceive after lscs till gets her regular menstrual cycle.
  - ✓ If pt. planning for pregnancy

    Yes - go for laparoscopy so that we can repair the scar defect

    No – go for hysteroscopy where scar defect may remain but pregnancy will be evacuated .

"Advancements in Laparoscopy: Excelling in Cystectomy, Myomectomy, and Safe Abdominal Entry"

Dr. Sonia Batra, Laprascopic and Pelvic surgeon.

Importance of safe entry techniques in laparoscopy to minimize complications.

- Approximately 50% of all complications during laparoscopy are entry-related,
- To diminish entry-related complications, surgeons should follow safe abdominal entry guidelines, which include proper positioning of the patient, safe placement of veeris needles and trocars, and identification of high-risk patients.
- The operating table should be horizontal, not tilted, to avoid complications related to blood vessels while placing the primary trocar or veeres needle.
- The abdomen should be palpated to check for any masses and the position of the lota before inserting needles or trocars.
- Sharp and right-angled needle inserted vertically from the base of the umbilicus.
- The needle should be inserted with two audible clicks and minimal lateral movement to avoid complications.
- Left upper quadrant placement of the needle is associated with fewer attempts and conversions to alternative sites.
- The initial intra-peritoneal pressure should be less than 10 mmHg.
- The weight of the patient is considered the most reliable indicator for determining the direction of the various needles.
- If the weight is less than 75 kg, the angle should be 45° towards the pelvis to avoid injuring blood vessels.
- If the weight is between 75 and 90 kg, the angle should be between 45° and 90° to ensure proper placement.
- For patients weighing more than 90 kg, the angle should be 90° to ensure entry into the abdominal cavity.
- According to guidelines, a pressure of 20 to 25 mmHg should be used for gas insufflation before inserting the trocar. This helps maintain a safe distance between the abdominal wall and the organs to prevent injury.
- The primary trocar should be inserted at the thinnest part of the abdominal wall, known as the Palmer point.
- It is located in the mid-clavicular line, three finger widths off the midline, and three finger widths below the left costal margin.
- Specific measures are required for laparoscopic surgery in obese and thin women.
- For obese women, an open technique or entry at the Palmer point is recommended.
- For thin women, the Hasson technique or insertion at the Palmer point is recommended for beginners.
- Clear optical entry, minimize entry wound size, reduce insertion force, and maintain the integrity of the facial layers. However, there is insufficient evidence to conclude that visual entry systems are superior to blind methods of trocar insertion.
- The direct trocar insertion technique, disposable visual entry systems, and shielded trocars are other safe entry technique. The choice of technique depends on the surgeon's skills and experience. Dr. J Prakash Patil . laparoscopist and pelvic surgeon

Laparoscopic myomectomy,
Dr. Jayprakash Patil

- The importance of preoperative planning and mapping of the fibroids.
- This allows for a better understanding of the location and number of fibroids, which helps in determining the type of incision and suturing technique to be used.
- He prefers a vertical incision for his myomectomies, as it allows for easier movement of his hands and transverse suturing.
- Choice of incision depends on the surgeon's preference and the location of the fibroids.
- Reducing dead space at the end of suturing to minimize the risk of hematoma formation and poor healing.
- Removal of multiple fibroids through a single incision or separate incisions, depending on their proximity.
- Using ultrasound during surgery to ensure that all fibroids are removed and to avoid leaving any behind.
- Follow the principles laid down by Dr. Victor Bon, who emphasized reducing the exposure of suture material and minimizing raw areas to reduce adhesions.
- Proper approximation of the myometrium to avoid creating defective scars.
- Iuse of barbed sutures has made the procedure easier and faster, while also ensuring uniform tension across the suture line. This results in minimal or no dead space.
- Baseball technique for suturing, where the bites are taken inside out until the suturing is complete.
- large fibroid that required cutting more to identify the good plane for enucleation.
- The overlap technique, where one flap of the fibroid's myometrium goes below and obliterates the dead space, while the other flap comes over the first flap to form a three-layered suture. This technique decreases dead space, reinforces the myometrial tissue, and creates a strong scar.
- I transverse incision is faster and allows for immediate hemostasis, but there is a risk of entering the broad ligament.
- On the other hand, a vertical incision does not have this risk, but the incision may be smaller than the uterus. The speaker emphasizes the importance of adequate incision size and proper suturing techniques.
- 30 cm length suture for beginners and suggest that a longer length may be used once the surgeon becomes more experienced.

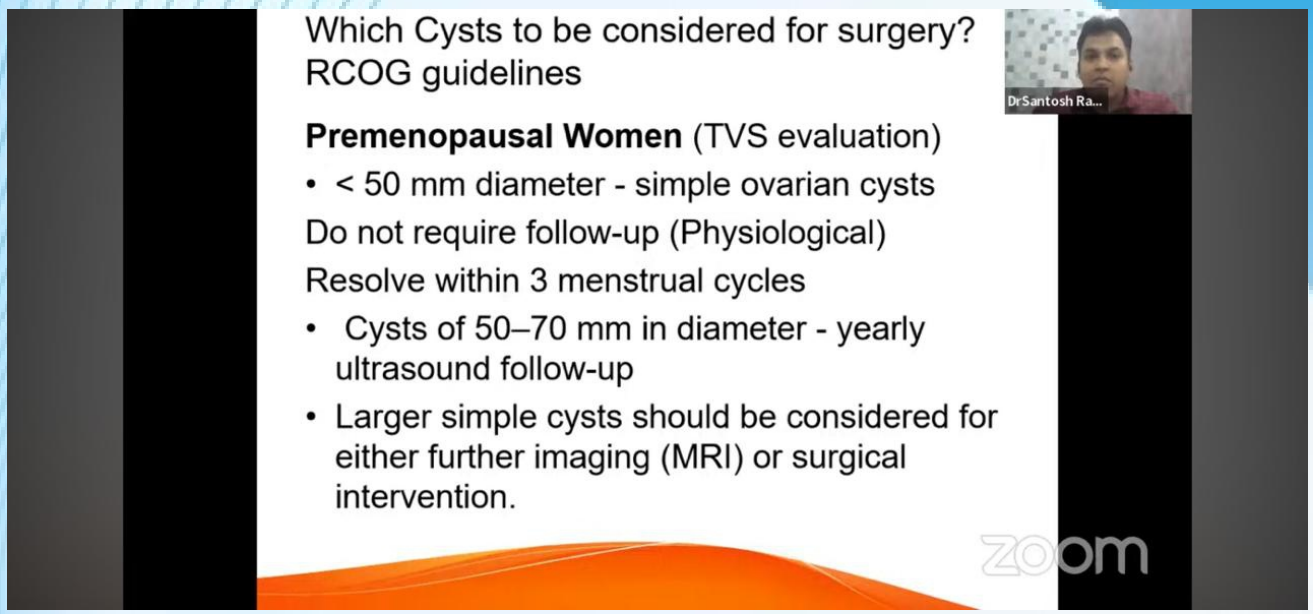
Laparoacopic cystectomy
Dr Santosh Rathod. Laparoscopic surgeon

- If there is suspicion of malignancy or persistent complex evaluation using MRI or CT and surgical intervention may be necessary
- Laparoscopy has become a preferred modality of treatment, citing benefits such as reduced pain, shorter hospital stay, and faster recovery, which ultimately leads to lower costs and less postoperative morbidity.
- informed consent and discussing all available treatment options with the patient.
- Mention the possibility of excessive bleeding, non-separation of the cyst, and the risk of malignancy, which should be explained to the patient in detail.
- Routine pre-operative bowel preparation is not recommended for gynecological procedures, according to recent guidelines.

- Instruments are essential for performing a cystectomy.Plain grasper, tooth grasper or needle holder, suction canula with a wide bore, and a good quality endobag.
- Tailoring the first entry based on surgical requirements and tumor size.
- Thorough inspection of the cavity upon entry and looking for the other ovary and tubes.
- Adequate adhesiolysis is also mentioned as it allows for better evaluation of the cyst surface.
- The goals of the surgery include complete resection of the tumor with minimal harm to normal ovarian tissue and correct dissection in the cleavage plane.
- The success of the surgery depends on how well the cleavage plane between the ovarian tissue and cyst wall is identified.
- Puncher and aspiration cystectomy,

- Gentle pulling during cystectomy procedures to avoid tearing the cyst and causing further damage to the ovarian tissue.

- Traction and countertraction to separate the cyst wall from the ovarian tissue.
- Endometriotic cyst -Carefully separate the ovary from the ovarian fossa where it is adherent, and due to the thin wall nature of the cyst, it gets ruptured.
- Take a nick above and drain the entire endometriotic cyst.
- anti-Mullerian hormone (AMH) levels. Studies have shown that there is a significant decline in AMH levels immediately after benign surgeries, but recovery of AMH levels can occur within 3 to 6 months of follow-up.
- However, in cases of endometriosis, there is a permanent loss of ovarian tissue, leading to a permanent decrease in AMH levels.
- Salpingo-oophorectomy, which is reserved for patients with highly suspicious masses or solid tumors, particularly in postmenopausal women.
- Proper exposure, medialization of the adnexa, and careful dissection and coagulation of the pelvic ligaments to safeguard the ureter.
- Smaller cysts can be extracted directly through the primary port using a reducer and a 5mm scope,
- while larger cysts can be divided into halves and taken out through the umbilical port or placed in an endobag and extracted through an ancillary or umbilical port.
- Dermoid cysts are placed in an endobag, punctured, and the contents are aspirated and removed using forceps.
- If Endo bag is not available, a Euro bag can be used instead.
- The importance of using warm saline during the procedure is emphasized, as it helps to make the sebaceous material more watery and easier to suction out.
- ovarian torsion, which is the twisting of the ovarian structure is more common on the right side
- laparoscopic management is ideal for diagnosing and treating torsion.
- Cystic teratomas and functional cysts are more prone to torsion due to their voluminous nature.
- Cysts in pregnancy is only if the cyst is symptomatic or shows malignant features.
- Careful intraoperative monitoring and coordination with the anesthetic team due to the physiological changes that occur during pregnancy. The speaker also mentions the need for DVT prophylaxis in pregnant patients.
- the use of diluted vasopressin in cases of dermoids and endometriomas to minimize damage to normal tissue during cyst wall excision.


Which Cysts to be considered for surgery?
RCOG guidelines
Premenopausal Women (TVS evaluation)
• < 50 mm diameter - simple ovarian cysts
Do not require follow-up (Physiological)
Resolve within 3 menstrual cycles
• Cysts of 50–70 mm in diameter - yearly ultrasound follow-up
• Larger simple cysts should be considered for either further imaging (MRI) or surgical intervention.
Dr Santosh Ra...
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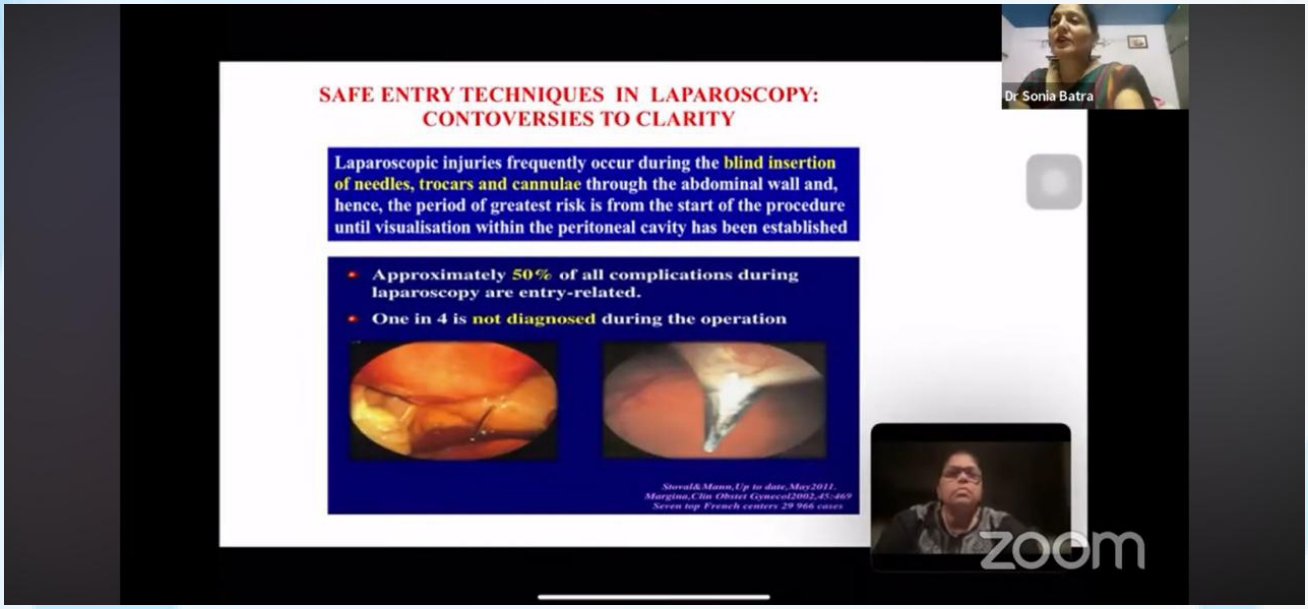
SAFE ENTRY TECHNIQUES IN LAPAROSCOPY:
CONTOVERSIES TO CLARITY
Laparoscopic injuries frequently occur during the blind insertion of needles, trocars and cannulae through the abdominal wall and, hence, the period of greatest risk is from the start of the procedure until visualisation within the peritoneal cavity has been established
Approximately 50% of all complications during laparoscopy are entry-related.
One in 4 is not diagnosed during the operation
Dr Sonia Batra
zoom


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In association with
ENDOSCOPIC COMMITTEE, KSOGA
Webinar
Welcomes you all
12TH MAR 2024
TUESDAY
7.00pm to 8.30pm
Guest of Honor
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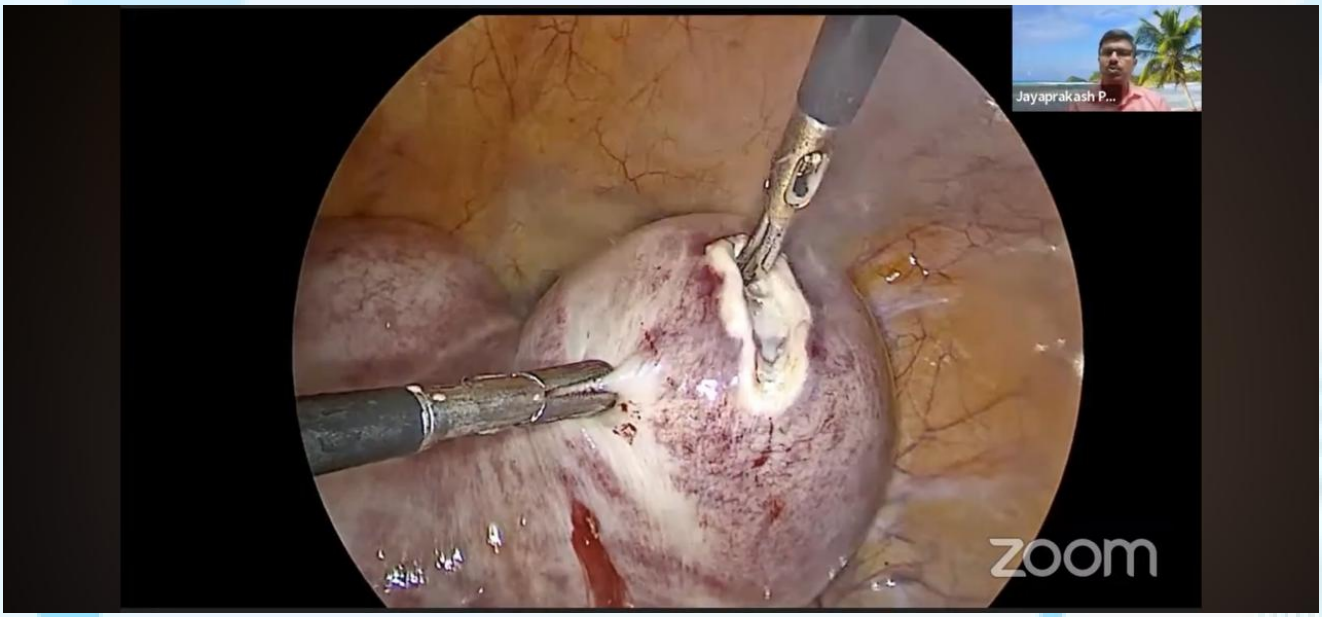
Dr. Vidya Thobbi
Dr.Bharathi Rajshekar
Dr.B.Ramesh
Dr Vidya Bhat
Topic:
"Advancements in Laparoscopy:
Excelling in Cystectomy, Myomectomy, and Safe Abdominal Entry"
Topic:
"From Controversies to Clarity:
Mastering Safe Entry Techniques in Laparoscopy"
07:05 TO 07:20 pm (Duration 10 minutes)
Speaker
Dr. Sonia Batra
Secretary,
Endoscopic Committee,
KSOGA
Topic:
"Laparoscopic Myomectomy:
Mastering Incisions and Sutures"
07:20 TO 07:40 pm (Duration 20 minutes)
Speaker
Dr Jayaprakash Patil
Director,
Bettadur Advanced 3D Laparscopic center
Raichur,Imm Past Chairman Endoscopic Commit
KSOGA
Dr Ravikanth G O
Chairperson,
Endoscopic Committee,KSOGA
Topic:
"Laparoscopic Cystectomy Essentials:
Pre-Op Prep and Safe Execution"
07:40 TO 07:55 pm (Duration 15 minutes)
Speaker


Jayaprakash P...
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Hysteroscopy Workshop:
At KSOGA Gadag 2023, where we performed six cases led by Dr. Vidya Bhat and Dr. Subhash Mallya, was very well appreciated by the audience.

Insight and intervention: Navigation Female Reproductive tract Anomalies"

Decoding the origins: The Embryology of Female Reproductive Tract.

Dr Ravikanth G O
Chairperson, Endoscopic Committee, Karnataka State Obstetrics and Gynaecologists Association

The development of female reproductive tract is interrelated with the development of urinary system and lower gastrointestinal tract.so the malformations are also interrelated. Understanding the development will be helpful to understand the problem and timing of pediatric gynecological problems.

The development of the female reproductive tract occurs in three phases.

I). Gonadal development.

II). Development of the Female Reproductive tract

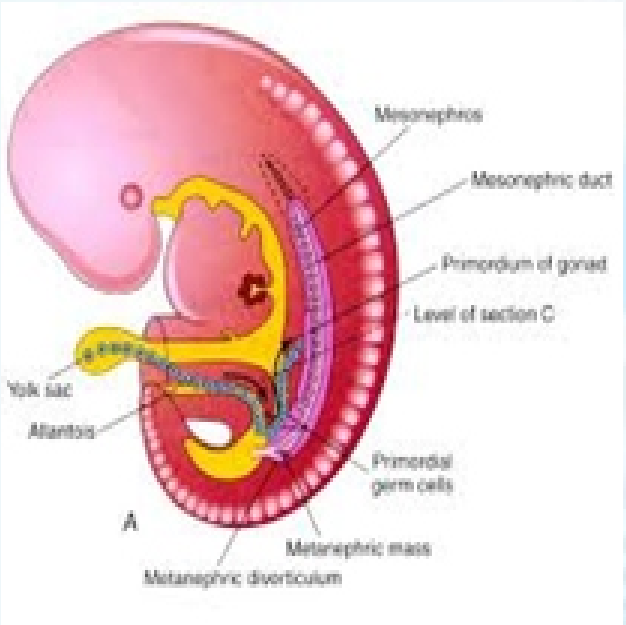
III). Development of External genitalia.

The gonadal development is the first thing to happen which occurs before completion of the Embryonic period. The Gonads decent into the pelvis inutero. The external genitalia development completes by the end of first trimester and start of second trimester. The development of the reproductive tract is continuous throughout intrauterine life. It continuous to matures early in childhood till Puberty.

I). Gonadal development.

It is better understood in four different but interrelated phases.

1). Indifferent Gonadal Phase.
2). Sexual Differentiation of Gonads
3). Development of Ovaries.
4). Pelvic Decent of Ovaries.



1). Indifferent Gonadal Phase.

The Gonadal development occurs from three sources. They are primitive germ cells, Mesothelial cells of the Posterior abdominal wall and the adjacent Mesenchyme.

The genotype of the gonad is decided at the time of conception. It is called indifferent because it has the capabilities of differentiation into either male or female gonad.

The primitive germ cells are developed in the yolk sac by fourth intrauterine week. They migrate through the dorsal mesentery of the Hindgut into Gonadal Ridge. The finger like projection from mesothelial cells is developed medial to developing Mesonephros to form Gonadal Ridge.

The primitive germ cells in the gonadal ridge along with mesenchyme are incorporated into finger like projection of the gonadal ridge to form Primary sex cord cells. The primary sex cord cells are short lived in females and disintegrate into the medulla as rete Ovaries. The cortex of the indifferent gonad as Secondary sex cord cells which incorporate the growing Primordial germ cells. The germ cells are most important component of the Gonads. Without the germ cells the gonads form streak gonads.

2). Sexual Differentiation of Gonads

The fetus is Bipotential in first three months of life. The Harmons and the biochemicals decide it either male or female. The embryo is destined to develop into Female by default. It is the nature rule. The absence of Y chromosome develops the embryo into Female. Presence of two XX chromosomes is necessary in the early fetal life for the complete development of the Female Embryo. In females it is the secondary sex cord cells that incorporate into cortex of the ovaries and preserve the primordial germ cells.

The presence of Y chromosome produces SRY protein from the gonads. This protein stimulates the primary sex cord cells in the medulla and help in development of the Testis. The mesenchymal cells produce anti-Mullerian Harmons. This inhibits the growth of Female reproductive tract. The interstitial cells also produce Testosterone which stimulates the development of Male External Genitalia.

The development of immature female embryo is influenced by the absence of three factors i.e. Y Chromosome,Anti-Mullerian Harmone and Testosterone. Furhter development is influenced by the Estrogen.

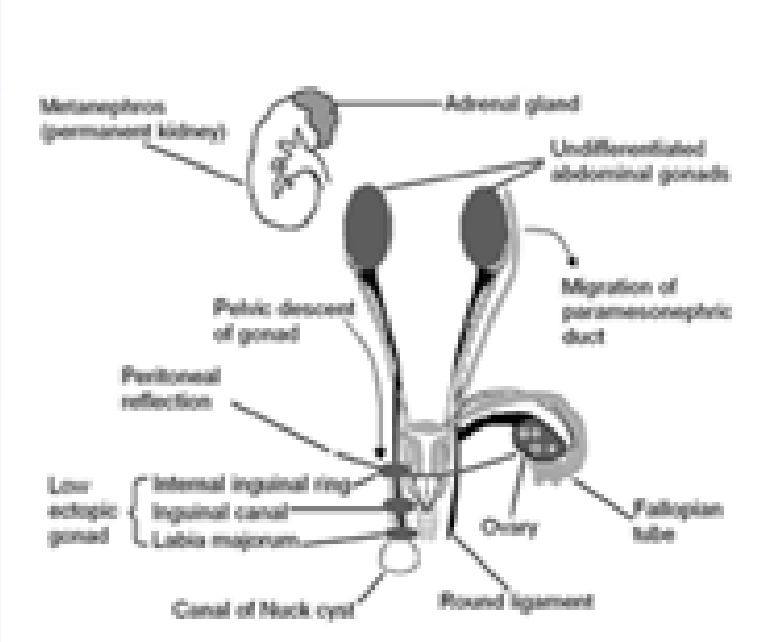
3). Development of Ovaries.

The secondary sex cord cells in the cortex develop as primitive germ cells. The germ cells migrated from the yolk sac into urogenital ridge undergo mitotic division from fourth week under the influence of placental hormones. By the end Fourth or Fifth-month intrauterine life there are 6-7 million germ cells in the gonads. The germ cells covered by the epithelial cells are called primordial germ cells. No further mitotic division is possible. Most primordial germ cells disintegrate by apoptosis. By the end of full term only 2- 4 million primordial germ cells remain. By puberty only 4 lakh follicles remain. They can remain in dormant stage for decades. The primordial follicles undergo arrest in first meiotic stage. The further completion firs

4). Pelvic Decent of Ovaries.

Gonadal ridge was developed in the lumbar region. The decent of the ovaries into pelvis happens due to marked development of the abdomen relative to the pelvis. It is guided by the Gubernaculum. It is the peritoneal fold from the caudal part of the ovaries to the uterus and uterus to the mons pubis which later forms ovarian and Round ligament respectively.

Maldescent of the ovaries, though rare, yet possible along this tract. Imaging has shown that the ovaries are located below the iliac crest, anterior superior iliac spine, umbilicus and above the pubis till the puberty.



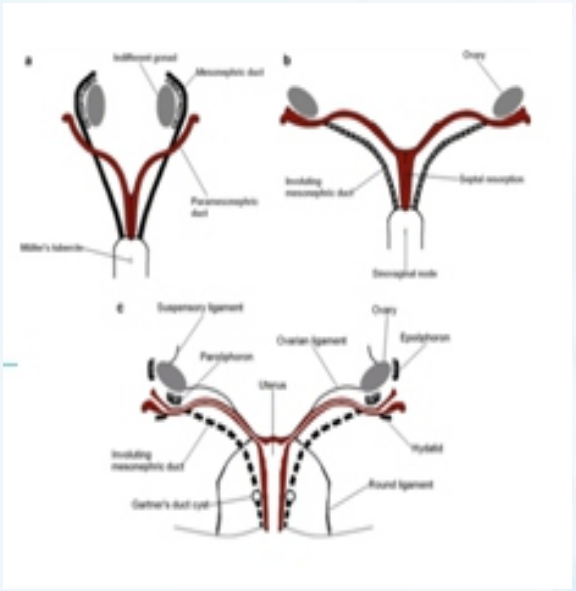
II). Development of the Female Reproductive tract

Basic knowledge of the urinary system development is necessary to learn about Female reproductive tract. The development of the urinary system occurs in three stages. They are pronephros in cervical region are paired temporary organs but important in the development of future kidneys. The Mesonephrons in thorasic act as temporary kidney. It is the Metanephrones in lumbar region develop into future Kidney.
The mesonphric duct drains mesophrones into cloaca till third month of life in females which later disintegrate and present as garnter duct, epoophoron,and parooophoron.

The most of Female reproductive tract is developed from.
1). Paramesonephric duct – Most of Female reproductive tract
2). Sinovagainal bulb of urogenital sinus - only lower one third to half of vagina is developed.

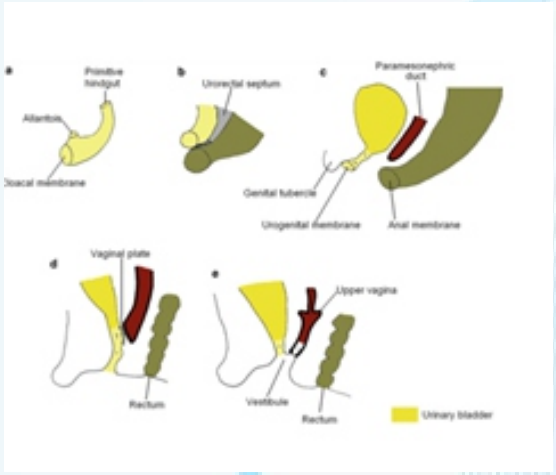
1). The paramesonphric duct.

It is the invagination of the mesothelium lateral to mesonephric duct which is the precussor of fallopian tube uterus,and upper half to two third of vagina.
The paired paramesonephric duct lateral to the mesonephric duct in the early part of life
grow caudally and course medial to mesonephric duct to join. This joining is called Mullerian organogenesis. The unfused cranial end forms fallopian tube and remains open into abdominal cavity. The fused cranial end forms uterus and cervix. The fused caudal end forms upper one half to two third vagaina.
Lateral fusion occurs by seventh to eight weeks. Complete epithelialization and disappearance of septum occurs by 20th week. Caudal end of the paramesonphric duct fuses with the sinovagainal bulb of urogenital sinus by eight weeks (Vertical Fusion).



2) Sinovagainal bulb of urogenital sinus.

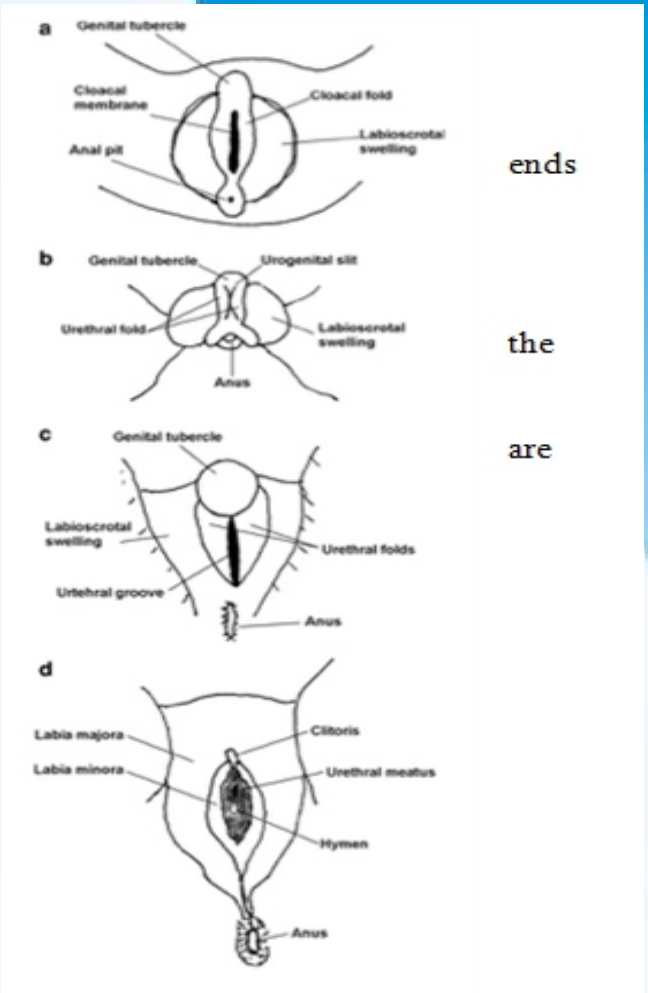
Cloaca is unified opening of the urinary system and gastrointestinal tract in the early part of life.
The abdominal wall below the umbilicus is formed by cloaca. Caudally the lateral folds are called cloacal folds and anteriorly they are fused to form Genital tubercle.
The urorectal septum divides cloaca into anterior urogenital membrane and posterior anal membrane. These membrane ruptures and has free communication with amnitic cavity.
The folds surrounding the urinary system form urethral folds which gives rise to urethra and surrounding the anal membrane as anal fold which gives rise to anus.
The urogenital system has caudal phallic portion and cranial pelvic portion.



The urethral groove and caudal phallic portion of urogenital sinus form virginal introitus. The narrow pelvic portion of urogenital sinus form urethra and distal vagina. The Sinovaginal bulb of Urogenital sinus meets Mullerian tubercle to make vertical fusion.

III). Development of External genitalia.

Cloacal membrane gives rise to genital ridge which form clitoris and lateral ends of the
cloacal membrane forms cloacal folds form labioscrotal swellings. The unfused Labioscrotal swelling form Labium majus. The unfused medial ends of urethral folds swelling form Labium minus.



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are

Conclusion: The female reproductive tract development is decided by the conception. The absence of Y chromosome is necessary to develop the embryo into female. The development is interrelated with the development of the urinary system and lower genital tract. So the anomalies are also present in combination.

Spectrum of variation: Classifying the female reproductive tract abnormalities

Dr. B MALLIKARJUNA KIRTI
MBBS DGO FRCOG CCT (UK)

Mullerian duct anomalies (MDA) are uncommon but can be a treatable form of infertility . Patients with MDA are known to have higher incidences of infertility, repeated first trimester spontaneous abortions, fetal intra-uterine growth retardation, fetal malposition, pre- term labour and retained placenta . The role of imaging is to detect and classify these MDA so that appropriate treatment is undertaken.

Embryology

The female reproductive tract develops from a pair of Mullerian ducts that form the following structures: fallopian tube, uterus, cervix and the upper two-thirds of the vagina. The ovaries and lower third of the vagina have different embryological origins derived from germ cells that migrate from the primitive yolk sac and the sinovaginal bulb, respectively.

Normal development of the Mullerian ducts depends on the completion of three phases: organogenesis, fusion and septal resorption. Organogenesis is characterised by the formation of both Mullerian ducts. Failure of this results in uterine agenesis/hypoplasia or a unicornuate uterus. Fusion is characterised by fusion of the ducts to form the uterus. Failure of this results in a bicornuate or didelphys uterus. Septal resorption involves subsequent resorption of the central septum once the ducts have fused. Defects in this stage result in a septate or arcuate uterus.

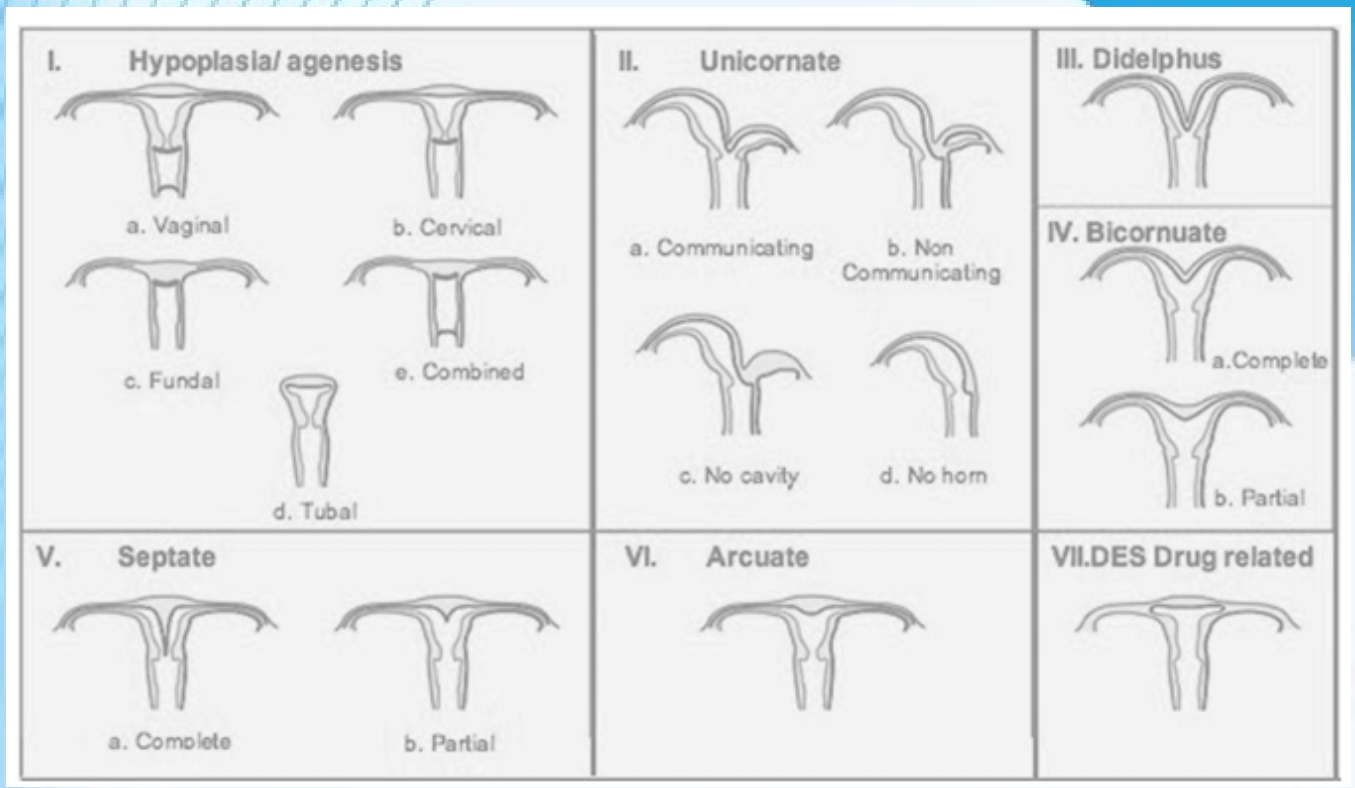
Classification

Until now, three systems have been proposed for the classification of female genital tract anomalies, although historically attempts for their categorization started quite earlier : the American Fertility Society's (AFS) currently American Society of Reproductive Medicine system , the embryological-clinical classification system of genito-urinary malformations and the Vagina, Cervix, Uterus, Adnexae and associated Malformations system based on the tumor nodes metastases (TNM) principle in oncology .

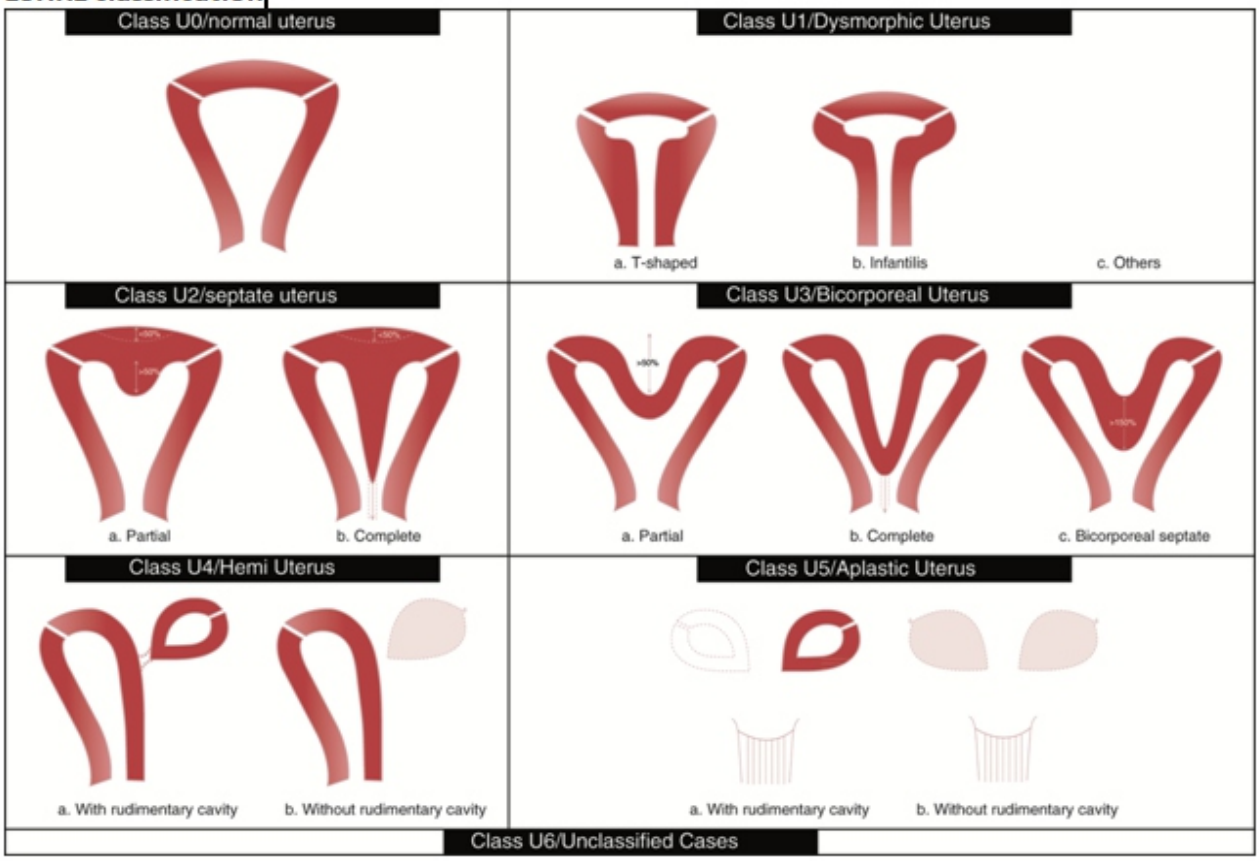
Although each proposal does not have the same acceptance, with that of the AFS classification system to be higher than the others, all of them seem to be associated with serious limitations in terms of effective categorization of the anomalies, clinical usefulness, simplicity and friendliness . It is noteworthy to mention that these limitations also gave place to further subdivisions for certain categories of anomalies . A systematic re-evaluation of the current proposals, within a project of the European Academy for Gynecological Surgery (EAGS), has been already published underlying the need for a new and updated clinical classification system .

The European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE), recognizing the clinical significance of female genital anomalies, have established a common working group under the name CONUTA (CONgenital UTerine Anomalies), with the goal of developing a new updated classification system. For this purpose, a scientific committee (SC) has been appointed to run the project, looking also for consensus within the scientists working in the field through the use of DELPHI procedure .

ASRM classification



ESHRE classification



ESHRE/ESGE classification
Female genital tract anomalies

	Uterine anomaly		Cervical/vaginal anomaly
	Main class	*Sub-class*	*Co-existent class*
U0	Normal uterus		
U1	Dysmorphic uterus	**a.** T-shaped **b.** Infantilis **c.** Others	
U2	Septate uterus	**a.** Partial **b.** Complete	
U3	Bicorporeal uterus	**a.** Partial **b.** Complete **c.** Bicorporeal septate	
U4	Hemi-uterus	**a.** With rudimentary cavity (communicating or not horn) **b.** Without rudimentary cavity (horn without cavity/no horn)	
U5	Aplastic	**a.** With rudimentary cavity (bi- or unilateral horn) **b.** Without rudimentary cavity (bi- or unilateral uterine remnants/aplasia)	
U6	Unclassified malformations		

C0	*Normal cervix*
C1	*Septate cervix*
C2	*Double 'normal' cervix*
C3	*Unilateral cervical aplasia*
C4	*Cervical aplasia*
V0	*Normal vagina*
V1	*Longitudinal non-obstructing vaginal septum*
V2	*Longitudinal obstructing vaginal septum*
V3	*Transverse vaginal septum and/or imperforate hymen*
V4	*Vaginal aplasia*

U	*C*	*V*

Associated anomalies of non-Müllerian origin:

Drawing of the anomaly

The ESHRE/ESGE classification system is presented here . It has the following general characteristics:

1. Anatomy is the basis for the systematic categorization of anomalies.
2. Deviations of uterine anatomy deriving from the same embryo- logical origin are the basis for the design of the main classes.
3. 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.
4. Cervical and vaginal anomalies are classified in independent sup- plementary sub-classes.

Definitions: uterine main classes and sub-classes

Class U0 -Incorporates all cases with normal uterus. A normal uterus is any uterus having either straight or curved interostial line but with an internal indentation at the fundal midline not exceeding 50% of the uterine wall thickness. The use of absolute numbers (e.g. indentation of 5 mm) is avoided in definitions as uterine dimensions as well as uterine wall thickness could normally vary from one patient to another. Thus, it was decided to define uterine deformity as proportions of uterine anatomical landmarks (e.g. uterine wall thickness). The addition of normal uterus gives the opportunity to independently classify congenital malformations of the cervix and vagina

Class U1 or Dysmorphic uterus incorporates all cases with normal uterine outline but with an abnormal shape of the uterine cavity excluding septa. Class I is further subdivided into three categories:

Class U1a or T-shaped uterus characterized by a narrow uterine cavity due to thickened lateral walls with a correlation 2/3 uterine corpus and 1/3 cervix.

Class U1b or uterus infantilis characterized also by a narrow uterine cavity without lateral wall thickening and an inverse correlation of 1/3 uterine body and 2/3 cervix.

Class U1c or others which is added to include all minor deformities of the uterine cavity including those with an inner indentation at the fundal midline level of ,50% of the uterine wall thickness. This aims to facilitate groups who want to study patients with minor deformities and to clearly differentiate them from patients with septate uterus. Usually, dysmorphic uteri are smaller in size.

Class U2 or septate uterus incorporates all cases with normal fusion and abnormal absorption of the midline septum. Septate is defined as the uterus with normal outline and an internal in- dentation at the fundal midline exceeding 50% of the uterine wall thickness. This indentation is characterized as septum and it could divide partly or completely the uterine cavity including in some cases cervix and/or vagina (see cervical and vaginal anomalies).

Class U2 is further divided into two sub-classes according to the degree of the uterine corpus deformity:

Class U2a or partial septate uterus characterized by the existence of a septum dividing partly the uterine cavity above the level of the internal cervical os

Class U2b or complete septate uterus characterized by the existence of a septum fully dividing the uterine cavity up to the level of the internal cervical os. Patients with complete septate uterus (Class U2b) could have or not cervical (e.g. bicervical septate uterus) and/or vaginal defects (see cervical/vaginal anomalies)

Class U3 or bicorporeal uterus incorporates all cases of fusion defects. As bicorporeal is defined the uterus with an abnormal fundal outline; it is characterized by the presence of an external indentation at the fundal midline exceeding 50% of the uterine wall thick- ness. This indentation could divide partly or completely the uterine corpus including in some cases the cervix and/or

vagina (see cervical and vaginal anomalies). As it could easily be imagined, it is also associated with an inner indentation at the midline level that divides the cavity as happens also in the case of septate uterus

Class U3 is further divided into three sub-classes according to the degree of the uterine corpus deformity:

Class U3a or partial bicorporeal uterus characterized by an exter- nal fundal indentation partly dividing the uterine corpus above the level of the cervix.

Class U3b or complete bicorporeal uterus characterized by an external fundal indentation completely dividing the uterine corpus up to the level of the cervix.

Class U3c or bicorporeal septate uterus characterized by the pres- ence of an absorption defect in addition to the main fusion defect. In patients with bicorporeal septate uterus (Class U3c) the width of the midline fundal indentation exceeds by 150% the uterine wall thickness; these patients could be partially treated by hysteroscopic cross section of the septate element of the defect. It should be noted, also, that patients with complete bicorporeal uterus (Class U3b) could have or not co-existent cervical (e.g. double cervix/formerly didelphys uterus) and/or vaginal defects (e.g. obstructing or not vaginal septum).

Class U4 or hemi-uterus incorporates all cases of unilateral formed uterus. Hemi-uterus is defined as the unilateral uterine development; the contralateral part could be either incompletely formed or absent. It is a formation defect; the necessity to classify it in a different class than that of aplastic uterus (formation defect) is due to the existence of a fully developed functional uterine hemi-cavity.

Class U4 is further divided into two sub-classes depending on the presence or not of a functional rudimentary cavity

Class U4a or hemi-uterus with a rudimentary (functional) cavity characterized by the presence of a communicating or non- communicating functional contralateral horn.

Class U4b or hemi-uterus without rudimentary (functional) cavity characterized either by the presence of non-functional contralat- eral uterine horn or by aplasia of the contralateral part. The presence of a functional cavity in the contralateral part is the only clinically important factor for complications, such as hemato-cavity or ectopic pregnancy in the rudimentary horn or hemato-cavity and treatment (laparoscopic removal) is always recommended even if the horn is communicating

Class U5 or aplastic uterus incorporates all cases of uterine aplasia

It is a formation defect characterized by the absence of any fully or unilaterally developed uterine cavity. However, in some cases there could be bi- or unilat- eral rudimentary horns with cavity, while in others there could be uterine remnants without cavity . Treatment options in patients having rudimentary horn with cavity are not yet clear . Furthermore, it should be noted that patients with aplastic uterus could usually have co-existent defects (e.g. vaginal aplasia/Mayer-Rokitansky-Ku sterHauser syndrome).

Class U5 is further divided into two sub-classes depending on the presence or not of a functional cavity in an existent rudimentary horn:

Class U5a or aplastic uterus with rudimentary (functional) cavity characterized by the presence of bi- or unilateral functional horn

Class U5b or aplastic uterus without rudimentary (functional) cavity characterized either by the presence of uterine remnants or by full uterine aplasia. The presence of a horn with cavity is clinically important and it is used as a criterion for sub-classification because it is combined with health problems (cyclic pain and/ or hemato-cavity) necessitating treatment.

Class U6 is kept for still unclassified cases. Modern imaging technology (ultrasound and/or magnetic resonance imaging) could provide objective estimations of uterine anatomy for the

needs of differential diagnosis among the six groups. However, infrequent anomalies, subtle changes or combined pathologies could not be allocated correctly to one of the six groups. A sixth class was created for these cases in order to keep the other groups 'clear'. Furthermore, the system is designed to include, hopefully, all cases resulting from formation, fusion or absorption defects of normal embryological development. Duplication defects or ectopic Mu llerian tissue anomalies, if existing, could not be described; these anomalies could be put in this class.

Definitions: co-existent cervical anomalies

Sub-class C0 or normal cervix incorporates all cases of normal cervical development.

Sub-class C1 or septate cervix incorporates all cases of cervical ab- sorption defects. It is characterized by the presence of a normal exter- nally rounded cervix with the presence of a septum.

Sub-class C2 or double cervix incorporates all cases of cervical fusion defects. It is characterized by the presence of two distinct externally rounded cervices; these two cervices could be either fully divided or partially fused. It could be combined with a complete bicorporeal uterus as a Class U3b/C2 in the formerly didelphys uterus.

Sub-class C3 or unilateral cervical aplasia incorporates all cases of unilateral cervical formation. It is characterized by the unilateral, only, cervical development; the contralateral part could be either incompletely formed or absent. Obviously, this has happened in Class U4 patients; however, this is not necessary to be mentioned in the final classification report (Class U4 instead of Class U4/C3) as being apparent. On the other hand, this sub-class gives the opportunity to classify other seldom anomalies such as complete bicorporeal uterus with unilateral cervical aplasia as Class U3b/C3, which is a severe obstructing anomaly. Anomalies and they could also draw the scheme of the malformation. cervical aplasia but, also, those of severe cervical formation

Sub-class C4 or cervical aplasia incorporates all cases of complete defects. It is characterized either by the absolute absence of any cervical tissue or by the presence of severely defected cervical tissue such as cervical cord, cervical obstruction and cervical fragmentation. The decision to include all variants of cervical dysgenesis in sub-class C4 was made in order to avoid an extremely extensive sub-classification, which does not seem to be user friendly. This sub-class could be combined with a normal or a defected uterine body and gives the opportunity to classify all obstructing anomalies due to cervical defects. It is characterized either by the absolute absence of any cervical tissue or by the presence of severely defected cervical tissue such as cer- vical cord, cervical obstruction and cervical fragmentation. The decision to include all variants of cervical dysgenesis in sub-class C4 was made in order to avoid an extremely extensive sub-classification, which does not seem to be user friendly. This sub-class could be combined with a normal or a defected uterine body and gives the opportunity to classify all obstructing anomalies due to cervical defects.

Definitions: co-existent vaginal anomalies

- Sub-class V0 or normal vagina incorporates all cases of normal vaginal development.
- Sub-class V1 or longitudinal non-obstructing vaginal septum. The incorporated anomaly in this sub-class is clear; it gives the opportunity to classify variants of septate or bicorporeal uteri together with septate or double cervices.
- Sub-class V2 or longitudinal obstructing vaginal septum. The incorporated anomaly in this sub-class is also clear and, its utility for the effective classification of obstructing anomalies due to vaginal defects is obvious.

- Sub-class V3 or transverse vaginal septum and/or imperforate hymen. This sub-class incorporates obviously different vaginal anomalies and their variants (mainly those of transverse vaginal septa); this was decided in order to avoid an extremely extensive sub-classification for the classification system's simplicity. The decision to put together those vaginal anomalies in this sub-class is due to the fact that they are usually present as isolated vaginal defects and they have the same clinical presentation (obstructing anomalies).
- Sub-class V4 or vaginal aplasia incorporates all cases of complete or partial vaginal aplasia.

Association with renal anomalies

Renal anomalies occur in 29% of MDA and are more commonly associated with unicornuate uteri than with other MDA . They are reported in roughly 40% of unicornuate patients and are ipsilateral to the rudimentary horn . Renal agenesis is the most commonly reported anomaly, occurring in 67% of cases . Other anomalies include ectopic kidney, horseshoe kidney, renal dysplasia and duplicated collecting systems

Conclusion

Acknowledging the important steps that have already been taken in the classification of congenital uterine anomalies (ASRM MAC2021, ASRM 2016, ESHRE/ESGE 2013and 2016 and CUME), the next step should be to achieve international consensus on a universally accepted classification system, which takes into account the strengths and limitations of all available classifications and is supported by high-quality research evidence.

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2. Human Reproduction, Vol.28, No.8 pp. 2032–2044, 2013
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Insightful Imaging: Diagnosisng Female Reproductive Tract Abnormalities

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The literature related to embryology and classifications has been discussed in previous sections.
Here in this section, we shall go through salient features of imaging mullerian anomalies.
A significant proportion of anomalies are diagnosed during workup in subfertility issues. Accurate diagnosis and correct classification help in the appropriate counselling of women about their potential reproductive prognosis and risks and for planning any intervention with a view to improve the reproductive outcome. Evaluation of the internal and external fundal contours of the uterus are the key in making a diagnosis and correctly classifying a uterine anomaly.
Imaging modalities such as ultrasonography (USG), hysterosalpingogram (HSG), sonohysterogram and magnetic resonance imaging (MRI)are less invasive modes of screening and classifying various uterine anomalies[1]. While conventional 2D transvaginal ultrasound (TVS) and HSG are considered as good screening modalities, 3D TVS and MRI can accurately diagnose and classify the types of CUAs [4,5], as they can define both external and internal uterine contours.

In this section the importance of USG is emphasised as we majority gynaecologists do our scans and also, we are familiar with knowledge of USG picks which define the abnormalities.

2D Transvaginal Ultrasound Conventional transvaginal ultrasound is minimally invasive and is less expensive way of assessing uterine morphology and ruling out uterine anomalies. Ultrasound evaluation can be timed in the secretory (luteal) phase of the menstrual cycle as the endometrium, being bright and echogenic, is easy to visualise; but still with good resolution we can analyse the defect in all phases of cycle in majority of cases. If needed we can subject for review scan in secretory phase. The visualisation of a double endometrial complex on a transverse plane point towards a uterine anomaly (Figure 1) and the differential diagnosis would be a bicornuate, septate, or subseptate. However, 3D ultrasound facilitates simultaneous visualization of both the external (serosal surface) and internal (endometrial) contours of the uterine fundus through its unique feature of providing the coronal plane of the uterus and can correctly classify the uterineanomaly into a bicornuate, septate or subseptate.

Systematic scanning through the longitudinal (sagittal) plane of the uterus may reveal a uterine complex that then disappears while moving to the opposite side, followed by the appearance of a second uterine complex, suggesting that the uterus may be a partial or complete bicorporeal uterus (bicornuate or didelphys). The transverse plane provides more information and widely placed double endometrial echoes, especially at the upper portion of the uterus towards the fundus (Figure 1C), and an indentation at the fundus on an oblique plane (if obtainable) is typical of a bicornuate uterus. The double endometrial echoes will be closer or wide apart in a septate uterus provided the serosal lining is maintaining its integrity without an indentation (Figure 1B) in contrast to that in a bicornuate uterus the serosal lining shows a prominent indentation with appearance of different echos (could be of bowel, peritoneum or omentum) which differentiates from the normal myometrial echo.

In uterus didelphys, the two whole uterine body with endometrial echoes will be separate to each other and placed apart (Figure 1D), and the clinical demonstration of two cervices confirm the diagnosis. Two uterine horns may be symmetrical or asymmetrical and two separate vaginas may be seen on speculum examination (as the vaginal septal identification is poor in TVS).

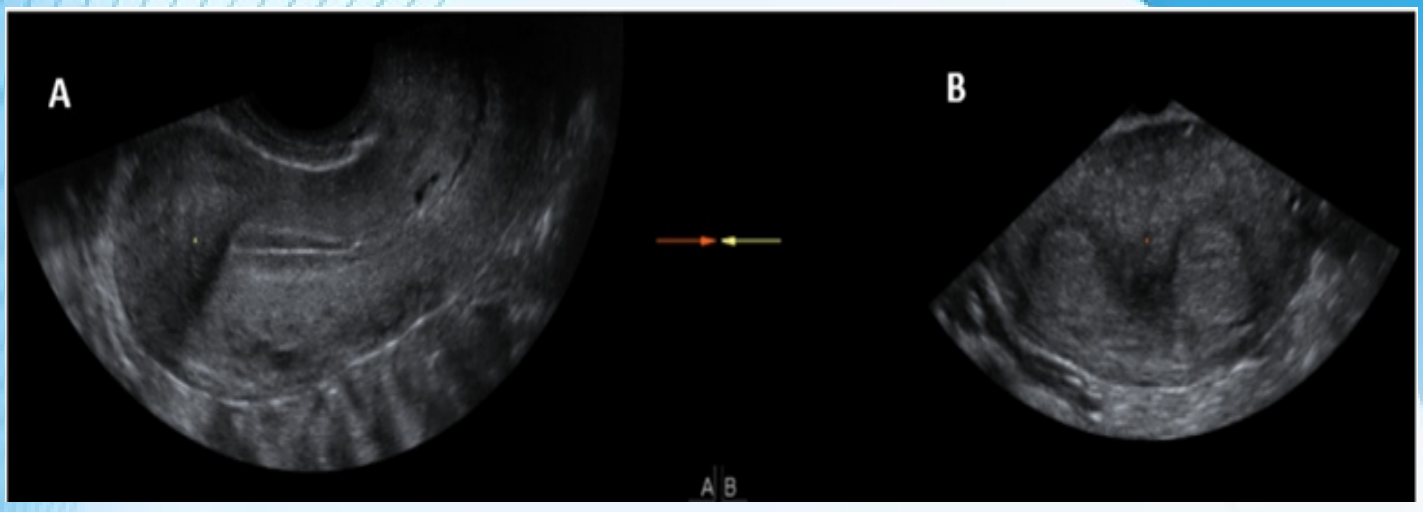


Figure 1: Longitudinal section of subseptate uterus in midsagittal plane (A); transverse plane of a subseptate uterus showing two endometrial echoes (B).

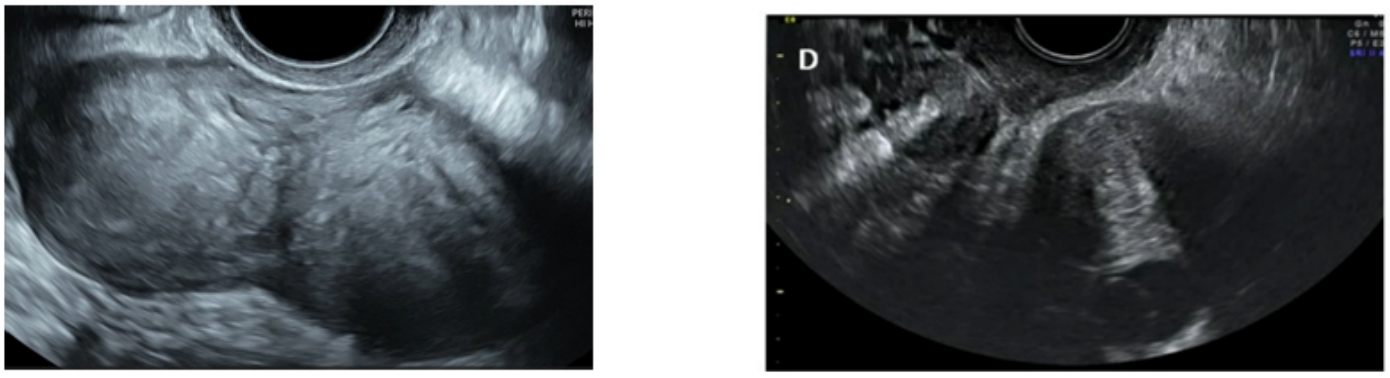


Figure 1: transverse plane of a bicornuate uterus showing two endometrial echoes (C); transverse plane of uterus didelphys showing two uterine bodies (D).

A unicornuate uterus may be difficult to diagnose on a conventional 2D scan. On a longitudinal scan, a normal looking sagittal axis of the uterus is seen on one side (we manoeuvre to get into long axis of the uterus as it is usually deviated to one side rather being in midline) in the pelvis with no or a rudimentary uterine shadow on the other side. On the transverse plane, the uterus is tapered to one side and at the level of the fundus, a beak like projection from the endometrial shadow (uterine angle or shoulder from where the interstitial portion of the fallopian tube starts) is seen only on one side (Figure 2 A). A rudimentary or severely hypoplastic uterine horn is seen as an isoechoic pear-shaped structure with or without a central thin echogenic endometrial line (Figure 2 B). A 3D ultrasound, again, is confirmatory, demonstrating a banana shaped uterine cavity with a single interstitial portion of the fallopian tube seen in the coronal plane (figure).

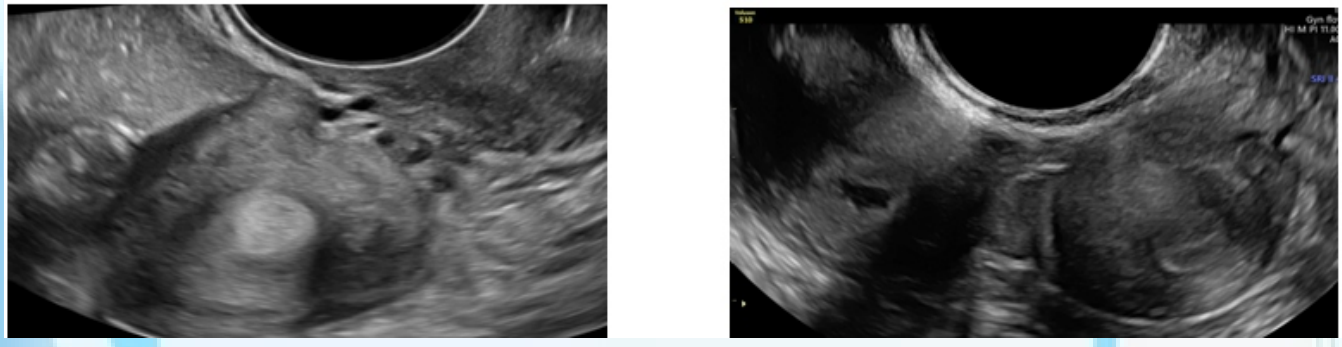


Figure 2: Unicornuate uterus: 2D transverse view showing only one uterine angle (shoulder)(A); a small right rudimentary uterine horn (B)

Three-Dimensional (3D) Transvaginal Ultrasound: The 3D transvaginal ultrasound is considered the gold standard tool for the assessment of uterine anomalies as it is less invasive; it facilitates simultaneous visualization of both the external (serosal surface) and internal (endometrial) contours of the uterine fundus through its unique feature of providing the coronal plane of the uterus and, therefore, can correctly classify the types of uterine anomalies. While the method for diagnosis is agreed, there are different international ultrasound criteria reported. The criteria for the classification of uterine anomalies based on 3D ultrasound have been well described by various international bodies like the ASRM, ESHRE and CUME (as it is been elaborated in previous chapters) although these have been criticized by some experts[2,3,6,7].

Uterine Morphology	Internal Contour	External Contour
Normal	Straight or convex	Uniformly convex or with indentation < 10 mm
Arcuate	Concave fundal indentation with central point of indentation at obtuse angle (>90°)	Uniformly convex or with indentation < 10 mm
Subseptate	Presence of septum, which does not extend to cervix, with central point of septum at an acute angle (<90°)	Uniformly convex or with indentation < 10 mm
Septate	Presence of uterine septum that completely divides cavity from fundus to cervix	Uniformly convex or with indentation < 10 mm
Unicornuate	Single well-formed uterine cavity with a single interstitial portion of fallopian tube and concave fundal contour	Fundal indentation > 10 mm dividing the two cornua if rudimentary horn present
Bicornuate	Two well-formed uterine cornua	Fundal indentation > 10 mm dividing the two cornua
T-Shaped	T-shaped uterine cavity	

Classification of uterine anomalies based on 3D ultrasound assessment Note: Arcuate is now recognised as normal.

Technical Considerations A suitable 3D US machine with a high frequency 3D transvaginal probe is used. The ultrasound setting is optimised to obtain a good quality image. The sweep angle is kept maximum (typically 120◦) and the acquisition speed of maximum quality is selected.The uterus is identified in the sagittal/longitudinal plane (unless it is a complete Bicorporeal /didelphis uterus in which case it can be identified in the transverse plane to obtain the uterine complex in the same sweep). The depth of the window is such that the uterine body occupies at least three-quarters of the screen. The focus is placed at the level of the endometrial cavity. The reference plane is kept as the midsagittal plane of the uterus. The 3D function is activated and both the operator and the patient should remain completely still while the acquisition takes place. The acquisition should incorporate the entire uterine body(ies) 8. Once the 3D acquisition is completed, the uterus is displayed on the screen in three orthogonal planes (A, B, C), which can be viewed in various different modes according to the US machine used (Figure 3). The default display is in the sectional mode and the operator can choose the render mode, which enhances the contrast between the two areas by recreating an impression of depth and improves visual perception. The operators should familiarise themselves with manipulation of the uterus in the three planes using the X, Y and Z functions on the US console.

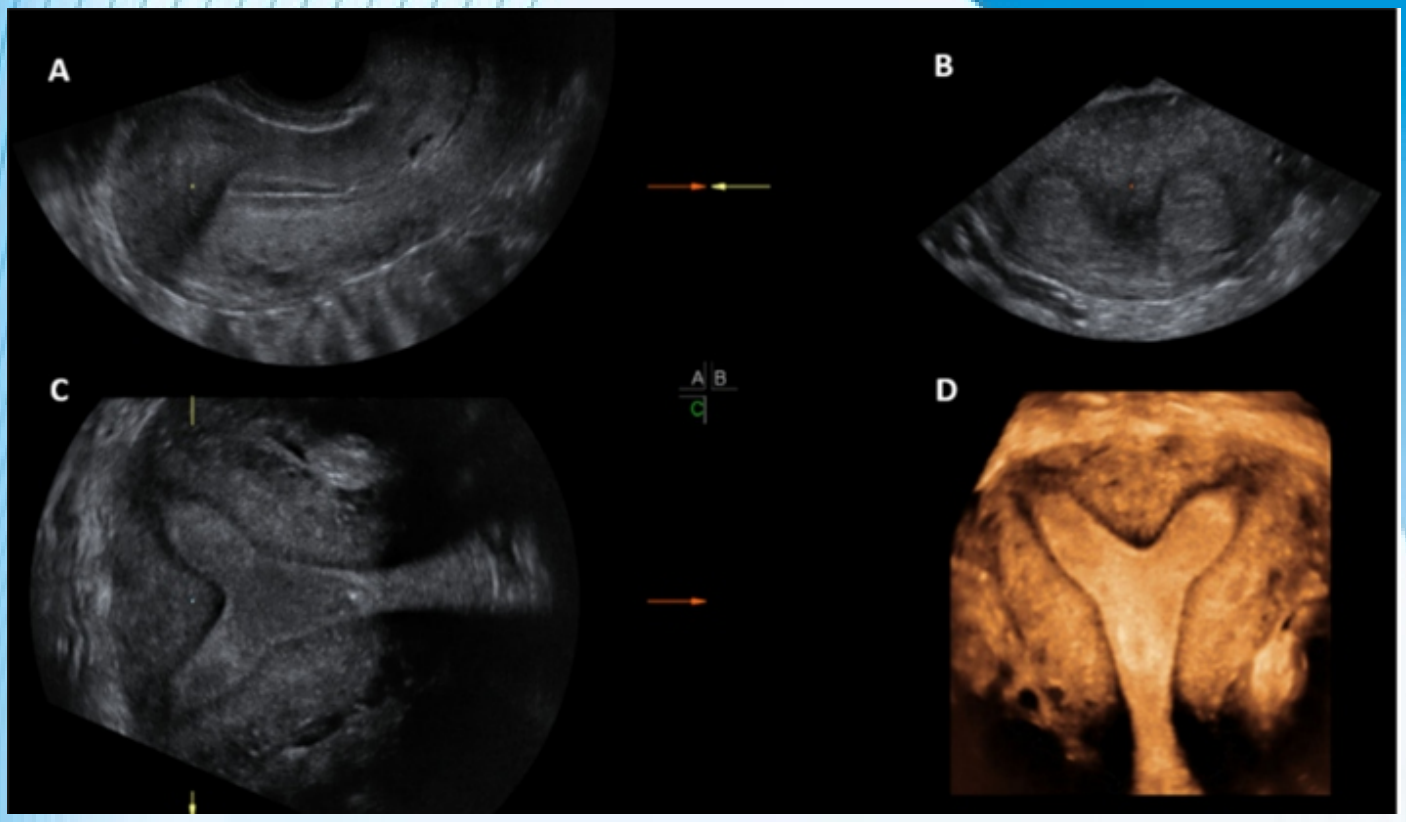
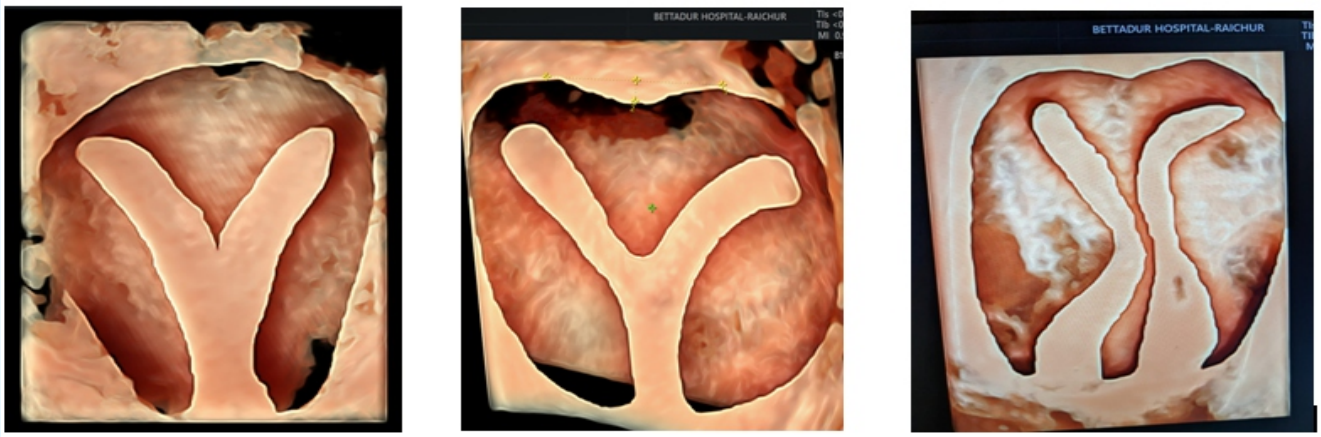


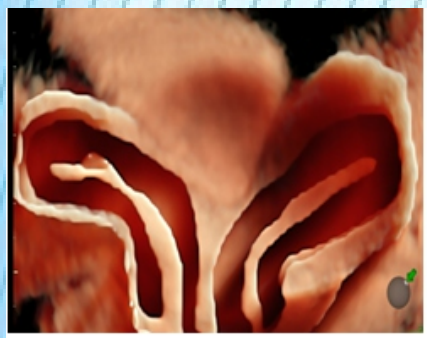
Figure 3: 3D ultrasound scan of a subseptate uterus showing simultaneous display of longitudinal plane (A), transverse plane showing two endometrial echoes (B), coronal plane (C), unique for 3D ultrasound and Rendered view of coronal plane demonstrating subseptate uterus (D).

When the render mode is selected, a region of interest (ROI) box will appear on the screen. The ROI box should be manipulated so that it covers the endometrial cavity in its entirety. The green line of the ROI box indicates the direction of rendering and this should ideally be placed on top. Final adjustments can be made, such as curving the green line, is placed along the curvature of the endometrial cavity and applying different render options, including Omniview, Volume Contrast Imaging (VCI), HD live and Magic cut, which produces a more realistic image. Once the operator is satisfied with the 3D image, this can be enlarged and can correctly classify the type of CUA. The entire volume and individual image can be saved for later analysis 9. The post-processing of the stored image can be performed as described above on the ultrasound machine itself (commonly done).

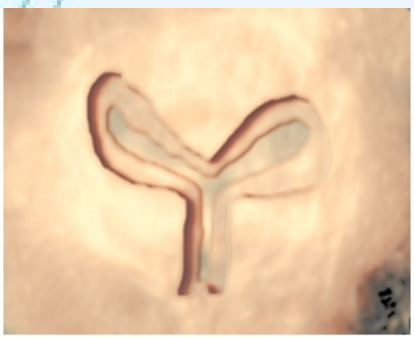
Below are common mullerian anomalies on 3D rendering.



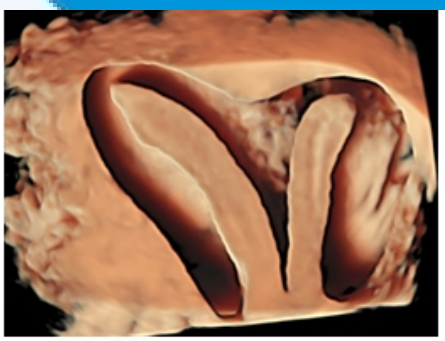
Subseptate | Broad based subseptate | Complete septate



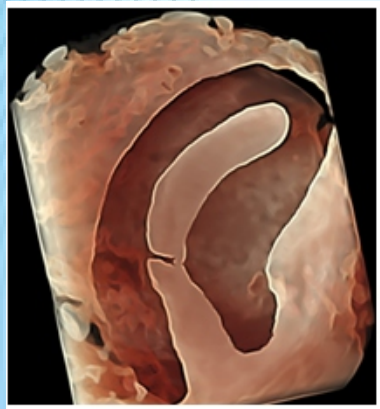
Bicorporeal bicollis/ Didelphys



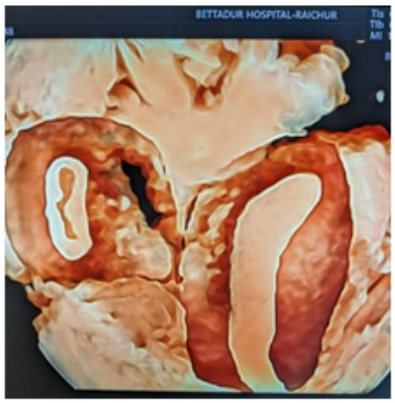
Bicorporeal unicollis



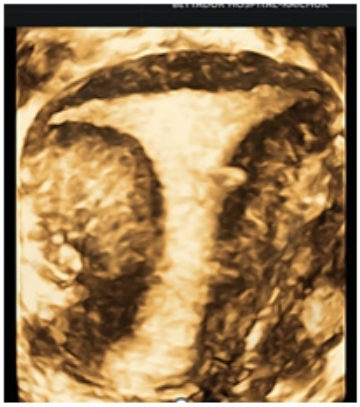
Bicorporeal septate



Unicornuate



Unicornuate with rudimentary horn



T shaped

Diagnostic Criteria Based on 3D Ultrasound The diagnostic accuracy of 3D ultrasound compared with laparoscopy ± hysteroscopy in diagnosing CUA is highest among other imaging modalities including MRI. The diagnostic accuracy of 3D ultrasound is reported as 97.6% with sensitivity and specificity of 98.3% and 99.4%, respectively 9.

The morphology of the uterus is best examined in the coronal plane (Figure 4) using the interstitial portions of the fallopian tubes as reference points. A line joining the tubal ostia (interostial line) is the reference line (Figure 9). A parallel line on top of the fundus can be drawn and the vertical distance between this line and the interostial line is the uterine wall thickness. In cases of septate uterus, a parallel line along the apex of the internal midline indentation is drawn and its vertical distance from the interostial line is the septal length. The vertical distance between the parallel linealong the bottom (apex) of the external indentation and interostial line is the depth of the external cleft in cases of partial bicorporeal (bicornuate) uterus.

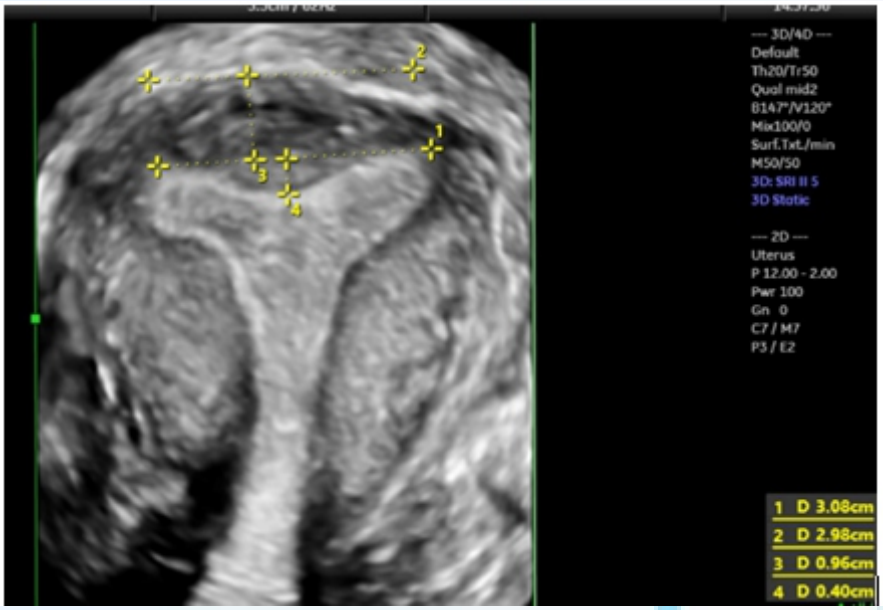


Figure 4: 3D coronal plane of uterus with assessments: interostial line (measurement 1); a parallel line along the serosal surface (measurement 2); uterine wall thickness (measurement 3) and septal indentation length. This uterus is not septate, but may be classified as arcuate uterus, which has no clinical relevance based on the recent ESHRE and ASRM guidelines.

There are differences in diagnostic criteria of these mullerian anomalies, especially in defining the septate variety. The ESHRE/ESGE classification of uterine anomalies based on the 3D scan assessment is based on using uterine wall thickness as the reference. An internal indentation at the fundal midline of more than 50% of the uterine wall thickness is used to diagnose a septate uterus. On the other hand, a bicorporeal (bicornuate) uterus is diagnosed when the external indentation is more than 50% of the uterine wall thickness. The ASRM in the publication 'Uterine septum: a guideline'

has suggested diagnosis of normal or arcuate uterus when the distance between the interostial line to the apex of indentation is less than 1 cm and the angle of indentation is more than 90 [10]. It also reported an arcuate uterus as a normal variant and clinically irrelevant. A septate uterus is diagnosed when the indentation depth is more than 1.0 cm and the angle of indentation less than 90 . A bicornuate uterus is diagnosed when the external fundal indentation is

more than 1 cm.

The Congenital Uterine Malformation Experts (CUME) group has been critical of the ESHRE/ESGE criteria as overestimating and the ASRM criteria as underestimating the prevalence of septate uterus[3]. In a diagnostic accuracy study using 3D ultrasound, the proportion of septate uteri using the ESHRE/ESGE classification was demonstrated to be

higher than using the ASRM criteria (RR 13.9; 95% CI 5.9–32.7, p ≤ 0.01)[11]. The concern about overdiagnosis is that it may lead to increased surgical intervention. The CUME group proposed a definition to diagnose septate uterus as an internal indentation of more than 10 mm, an angle of septal indentation of <140 and an indentation to the uterine wall

thickness (I–WT) ratio of >110%. They proposed to use the septal indentation of >10 mm alone as the simplest and most reproducible criteria, if used alone.

Comparison of measurements by 3 major guidelines to define septate variety is depicted in below Figure 5.

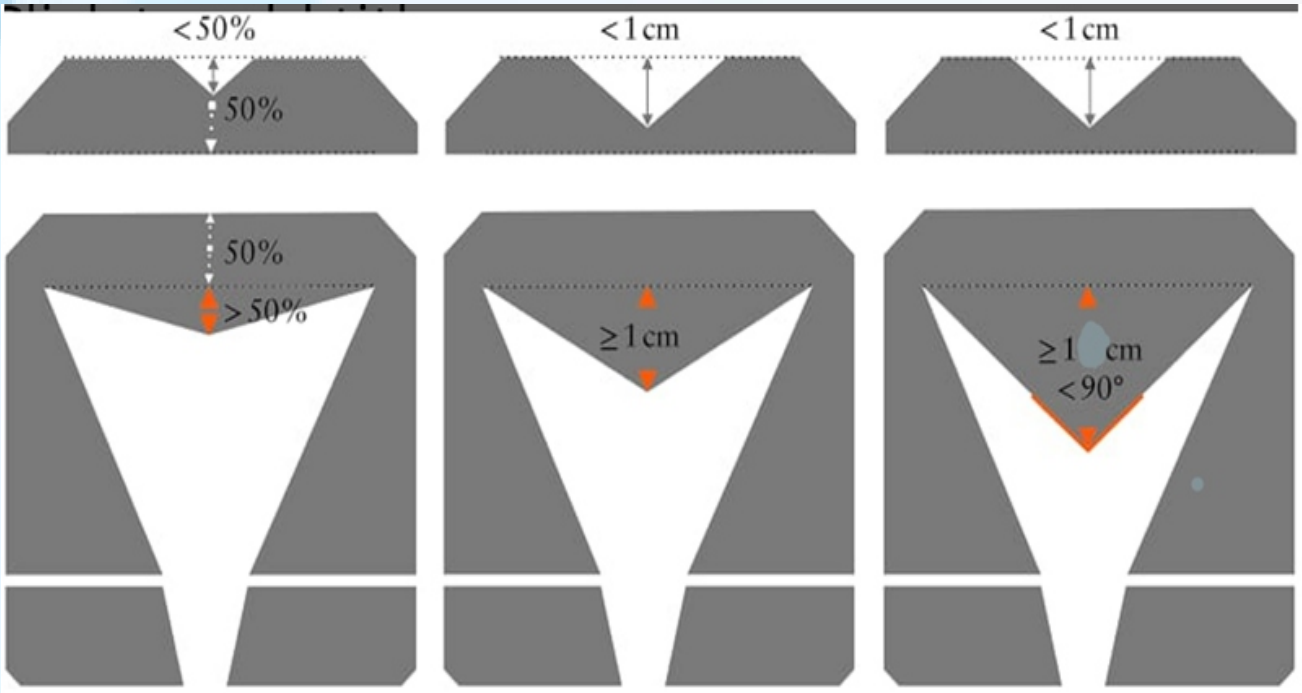


Figure 5: ESHRE/ ESGE 2016 CUME 2018 ASRM 2021

Congenital Uterine Malformations by Experts (CUME) is a better criterion to distinguish between Normal/ Arcuate and septate uterus. Following figure 6, explains the CUME criteria for diagnosis.

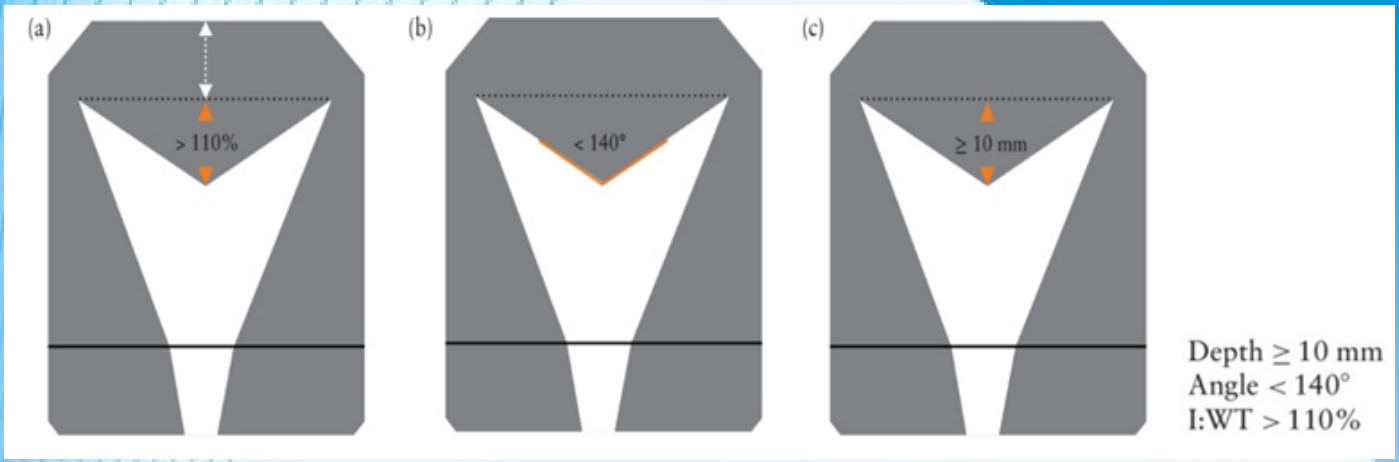
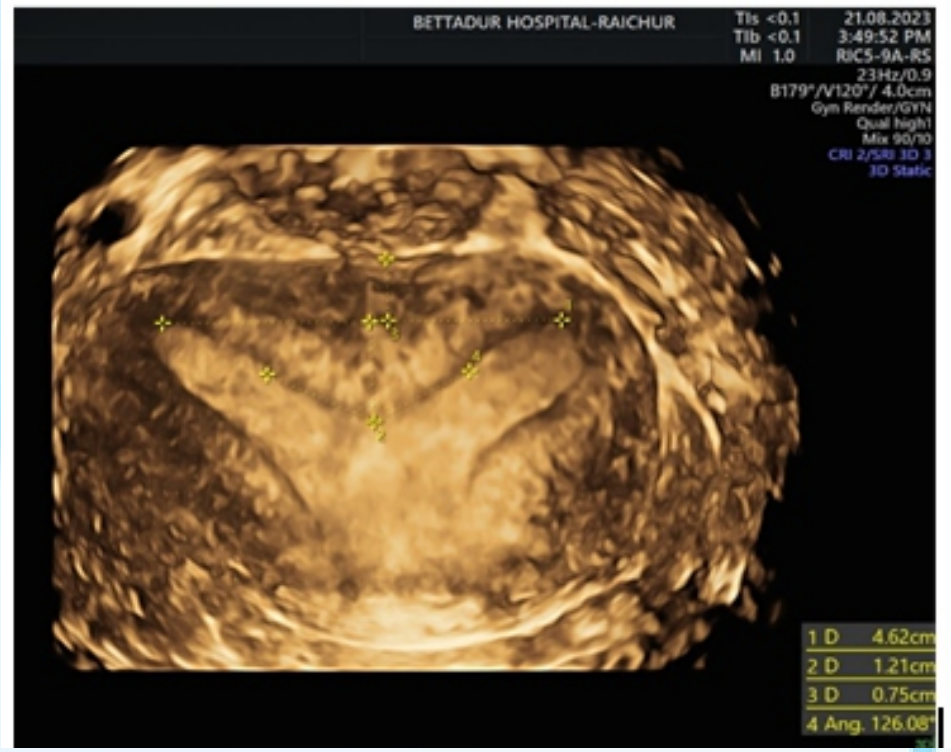


Figure 6: CUME Criteria and best cut-offs for distinguishing between normal/arcuate and septate uterus. (a) indentation-to-wall-thickness ratio >110 %; (b) indentation Angle <140

A summary of the criteria for diagnosing a septate uterus is detailed in Table below.

Guideline	Diagnostic Criteria
AFS (1988) [5]	Subjective impression and clinically relevant
ESHRE/ESGE (2013) [6]	Indentation (I) to wall thickness (I–WT) ratio > 50%
ASRM (2016) [19]	Septal angle < 90° and Septal length > 15 mm
CUME (2018) [9]	Septal length > 10 mm, Septal angle < 140° and I–WT > 110%
ASRM (2021) [7]	Septal angle < 90° and Septal length > 10 mm

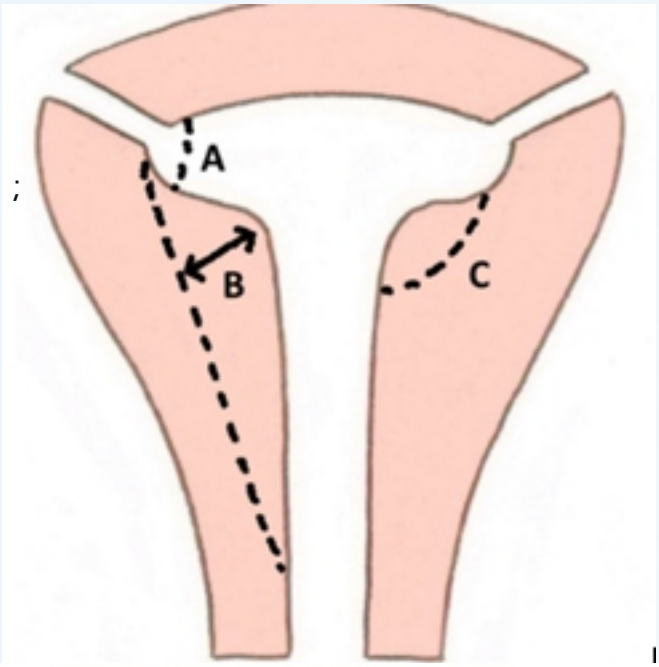
A case for analysis: Here is a case of Subseptate uterus

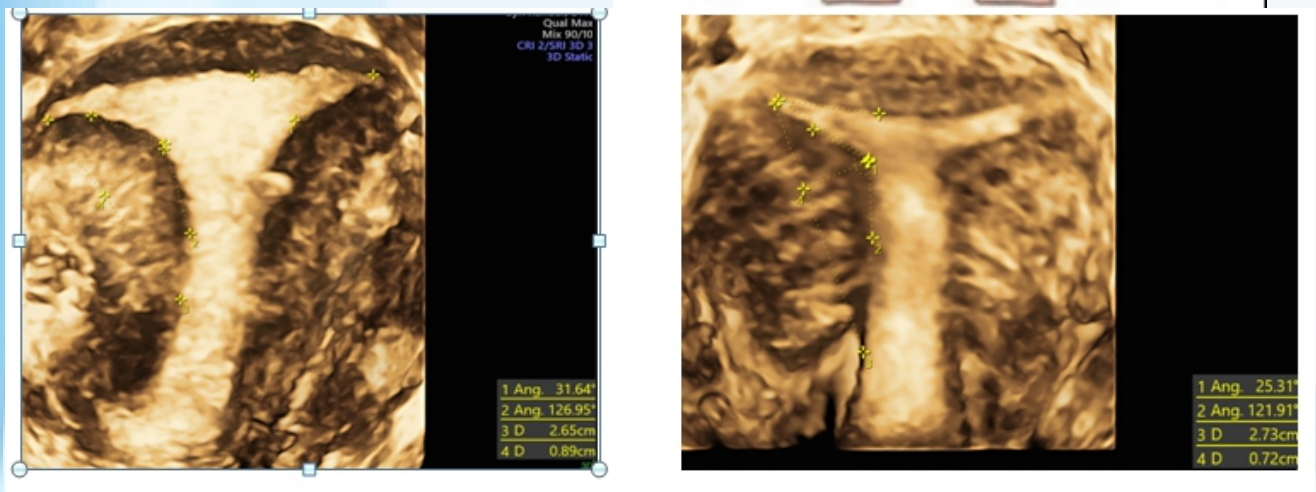


In the figure above 3D shows the regular convexity of serosal surface with no indentation. The indentation depth (measurement 2 is more than 1cm). The bifurcation angle is within 140deg. The indentation to wall thickness ratio is >110%. It fulfils the CUME criteria to define this as septate variety.

T shaped uterus:
While the T-shaped uterine cavity is diagnosed subjectively by some authors, the ESHRE/ESGE has suggested diagnosis based on a narrow uterine cavity due to thickened lateral walls without specifying a definition and cut-off for thickened lateral walls and narrow uterine cavity[12]. The CUME group proposed a lateral indentation angle ≤ 130^0 lateral indentation depth ≥ 7 mm and T-angle ≤ 40 as the criteria for diagnosing a
T-shaped uterus.

Criteria for diagnosing T-shaped uterus according to CUME ([22]). A—T-angle ≤ 40 ;
B—lateral indentation depth ≥ 7 mm;
C—lateral indentation angle ≤ 130
Below are examples which fulfil the CUME criteria for T shaped uterus





T shaped uterus is difficult to diagnose or have suspicion on 2D TVS. 3D rendering is must to diagnose and also quantify the defect.

Magnetic Resonance Imaging (MRI)

MRI has the multiplanar capability, allows better soft-tissue characterisation and permits a wider field of assessment at once than other imaging modalities including ultrasound . MRI, due to its ability to demonstrate both the external and internal contours of the uterus, is sensitive and specific for diagnosing nearly all uterine anomalies.

MRI is helpful in delineating the endometrium and detecting uterine horns regardless of the uterine position and anatomical variation. Furthermore, it is accurate in defining aberrant gonadal location or renal anatomy and is less invasive compared to laparo-hysteroscopy. In addition, it does have the ability to examine the details of the nearby structures including the cervix and vagina. While MRI is not routinely recommended in all women suspected

to have a uterine anomaly, it proves useful for those patients with suspected complex anomalies, particularly if there is a combination of cervical and vaginal anomalies and for those at higher risk for associated anomalies. MRI is also useful if there is any diagnostic dilemma with other imaging modalities.

As mentioned prior, MRI in mullerian analysis is beyond the scope of discussion in this section so not discussed in detail.

Conclusions

Uterine anomalies are commonly seen in women presenting with a history of reproductive problems. While 2D ultrasound and HSG are adequate for screening for uterine anomalies, 3D ultrasound, MRI and combined laparoscopy and hysteroscopy can correctly classify the type of uterine anomaly due to their ability to show both external and internal contours of the uterus. While 3D ultrasound is now considered as the gold standard diagnostic tool for uterine anomalies due to its high degree of diagnostic accuracy, less invasive nature and it being comparatively less expensive, MRI is reserved for diagnosing complex Mullerian anomalies or if there is a diagnostic dilemma. Laparoscopy and hysteroscopy are an invasive modality for diagnostic purposes and should be offered only in the context of concomitant surgical treatment after a thorough non-invasive evaluation of a Mullerian anomaly. Imaging for renal anomalies is recommended if a uterine anomaly is diagnosed.

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Decoding distinctions- Navigating deferential diagnosis of Female reproductive Abnormalities.

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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%. The clinical presentation varies over a wide range and is related to both the complexity and spectrum of underlying and associated conditions. Knowledge of genitourinary embryology is essential for the understanding, diagnosis and subsequent treatment of genital malformations. Differential diagnosis of Müllerian tract abnormalities typically involves considering various congenital anomalies and acquired conditions that may present with similar symptoms or findings.

External genital abnormality can be diagnosed at the time of birth but most of the mullerian abnormality are not diagnosed until puberty and many times extremely late in life due to reproductive outcome. Therefore, it becomes convenient to discuss the differential diagnosis depending on the presentation and symptom. Mullerian abnormality can broadly be classified into prepubertal and post pubertal.

Prepubertal

External genital abnormalities - external genital abnormalities are classified separately from Müllerian abnormalities because they involve different structures and embryological processes. However, both types of abnormalities can sometimes occur together in the context of broader genitourinary or reproductive developmental disorders. In the presence of external genital abnormality patient should be assessed.

Non obstructive mullerian abnormalities are usually asymptomatic. Routine physical examination would not reveal an isolated Müllerian anomaly and therefore when diagnosis is made in the young girl it is usually in association with other malformations or as an incidental radiologic or surgical finding.

Primary amenorrhea without pelvic pain

Müllerian agenesis is the second most common cause of primary amenorrhea and is the most common structural defect resulting in primary amenorrhea without pelvic or abdominal pain. Mayer Rokitansky- Küster-Hauser (MRKH) syndrome, occurs in 1 of 4–5000 live female birth. Individuals with MRKH have a 46 XX karyotype. Because ovarian function is unaffected in girls with MRKH, pubertal onset and development of secondary sexual characteristics is unaffected. On physical examination, normal breast development and pubic hair is evident. Other conditions associated with Müllerian duct aplasia include Müllerian-renal-cervical syndrome (MURCS), Klippel-Feil, Fraser syndrome, and some anorectal malformation.

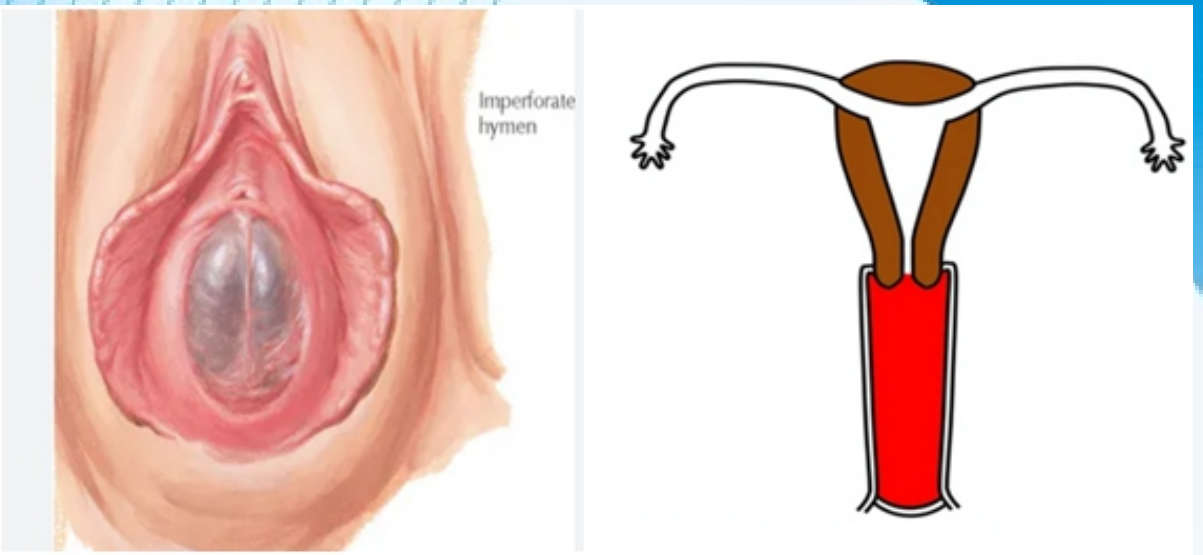
Disorders of sexual differentiation are associated with varying degrees of Müllerian or vaginal agenesis including mixed gonadal dysgenesis, complete androgen insensitivity syndrome (CAIS), or partial androgen sensitivity syndrome (PAIS). Chromosomal studies and testosterone levels are important to help differentiate between these complex diagnoses. Individuals with CAIS have a 46 XY karyotype but appear phenotypically as normal females.

Other causes of primary amenorrhea without pain would include hypothalamic amenorrhea, ovulatory dysfunction, constitutional delay, ovarian failure, and other endocrine imbalances or aberrant pharmacologic or environmental exposures including chemotherapeutic agents or radiation and Tuberculosis.

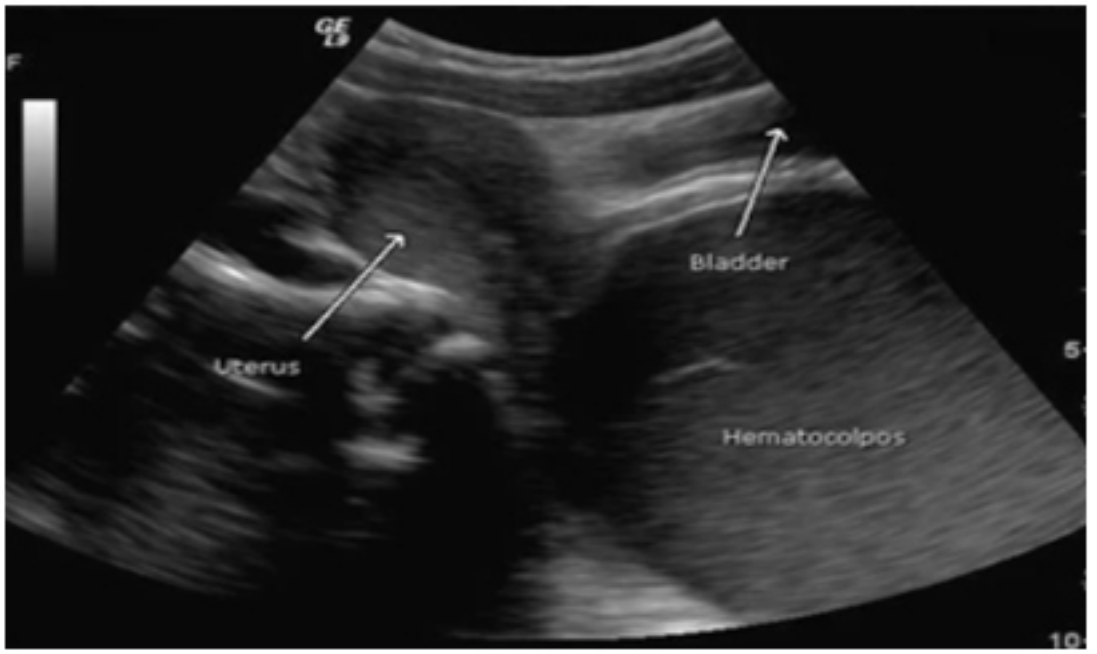
Amenorrhea with Pelvic Pain.

Mullerian defects with obstruction to the outflow of menstrual blood leads to progressive pelvic pain with varying intensity over the period. Initially starts as episodic going on to become episodic with continues pelvic pain with time. The obstruction can involve the hymen, vagina, cervix, or uterine horn.

Imperforate hymen - vagina is not affected structurally. accumulated menstrual blood can cause significant vaginal distention leading to a large pelvic/abdominal mass. visual inspection of the vulva reveals a hymenal membrane that is bulging. Valsalva manoeuvre or gentle pressure on the suprapubic mass intensifies the hymenal bulge of an imperforate hymen when the hymenal examination is equivocal. Imaging by ultrasound, CT scan, or MRI is necessary to rule out pelvic tumour and assess the extent of hematocolpus. Hematocolups can be sometime associated with hematometra and pelvic collection.



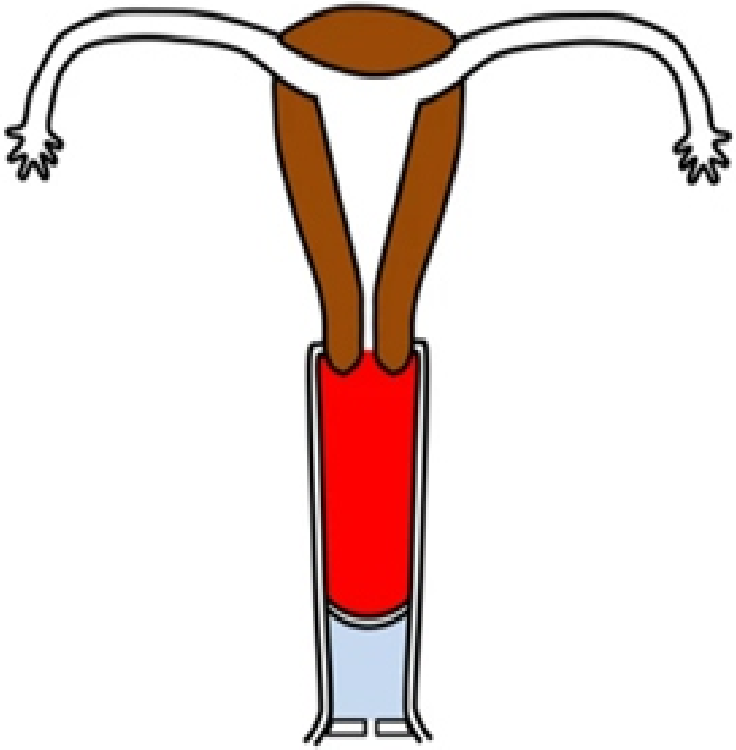
Imperforate Hymen bulging as blue membrane.



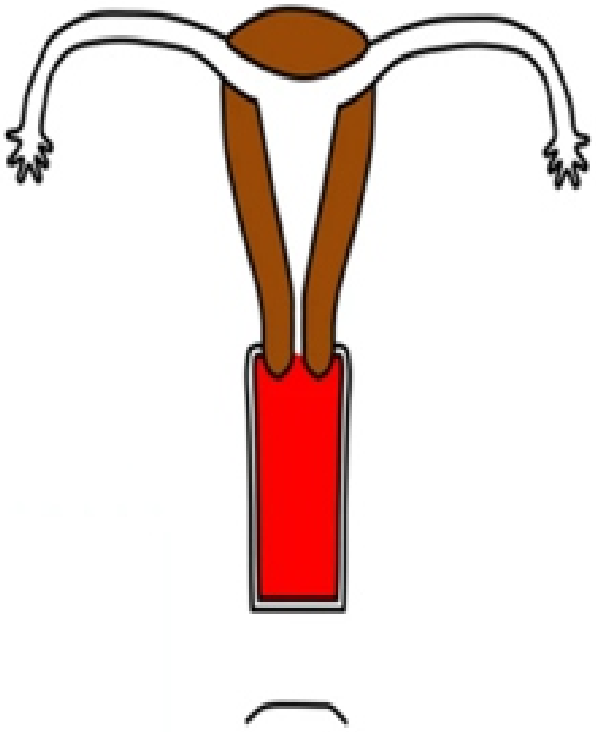
Transabdominal Ultrasound with Hematocolpos.

Transverse vaginal septum – It can present with amenorrhea and cyclic pelvic pain associated with a pelvic mass. Girls with a transverse vaginal septum have a normal hymenal opening but a short vagina. Vaginal depth varies with the location of the septum.

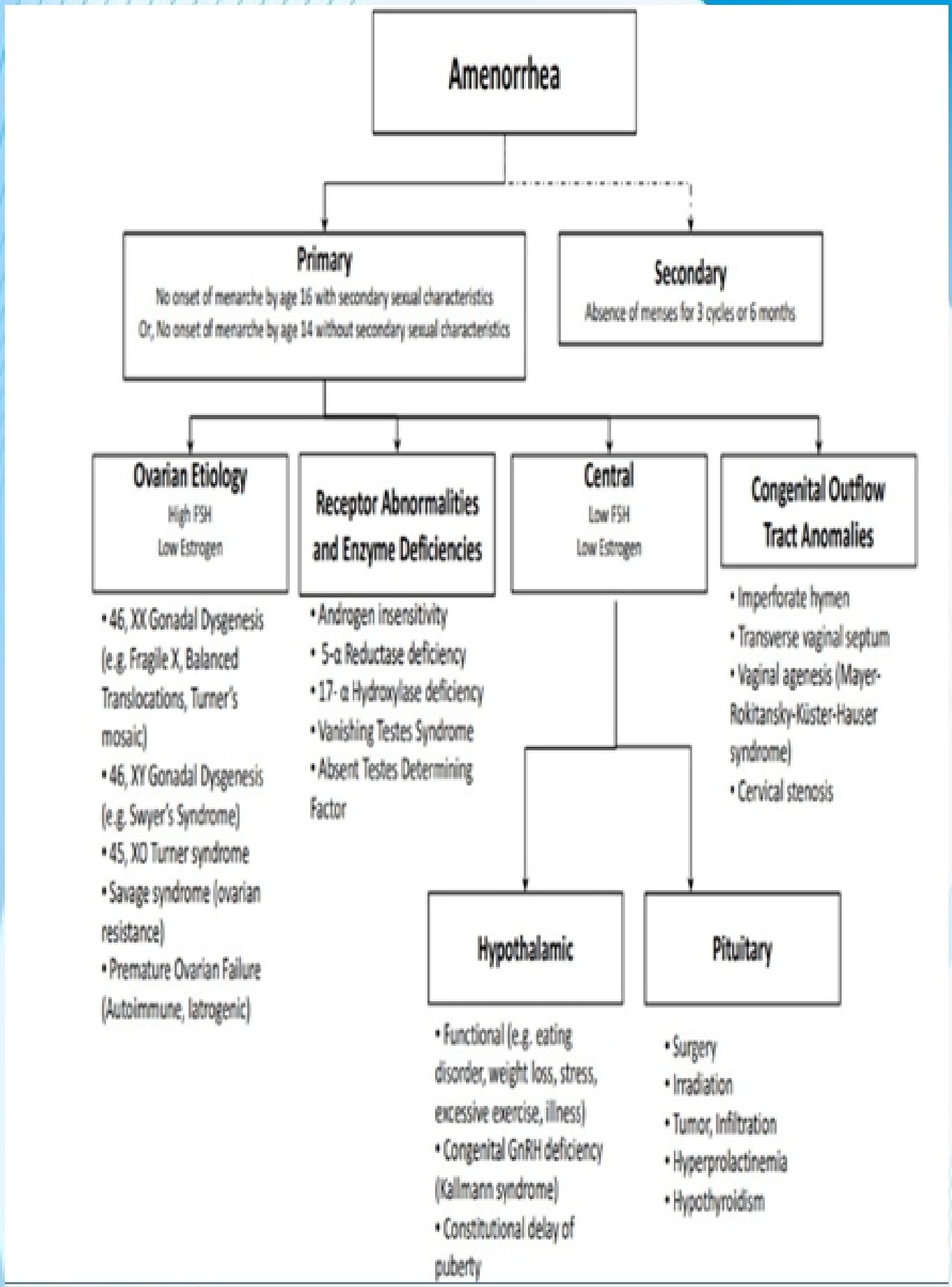
Cervical agenesis -presents in a similar fashion, although the vagina is usually present, but blind ended and non-communicating with the uterus and there is no vaginal or hymenal bulging.
Pelvic ultrasound may be sufficient to assess uncomplicated Müllerian anomalies. In contrast, complex anomalies must be further evaluated by pelvic MRI to assess the degree and exact location of the defect to allow for adequate surgical planning.



Transverse vaginal Septum

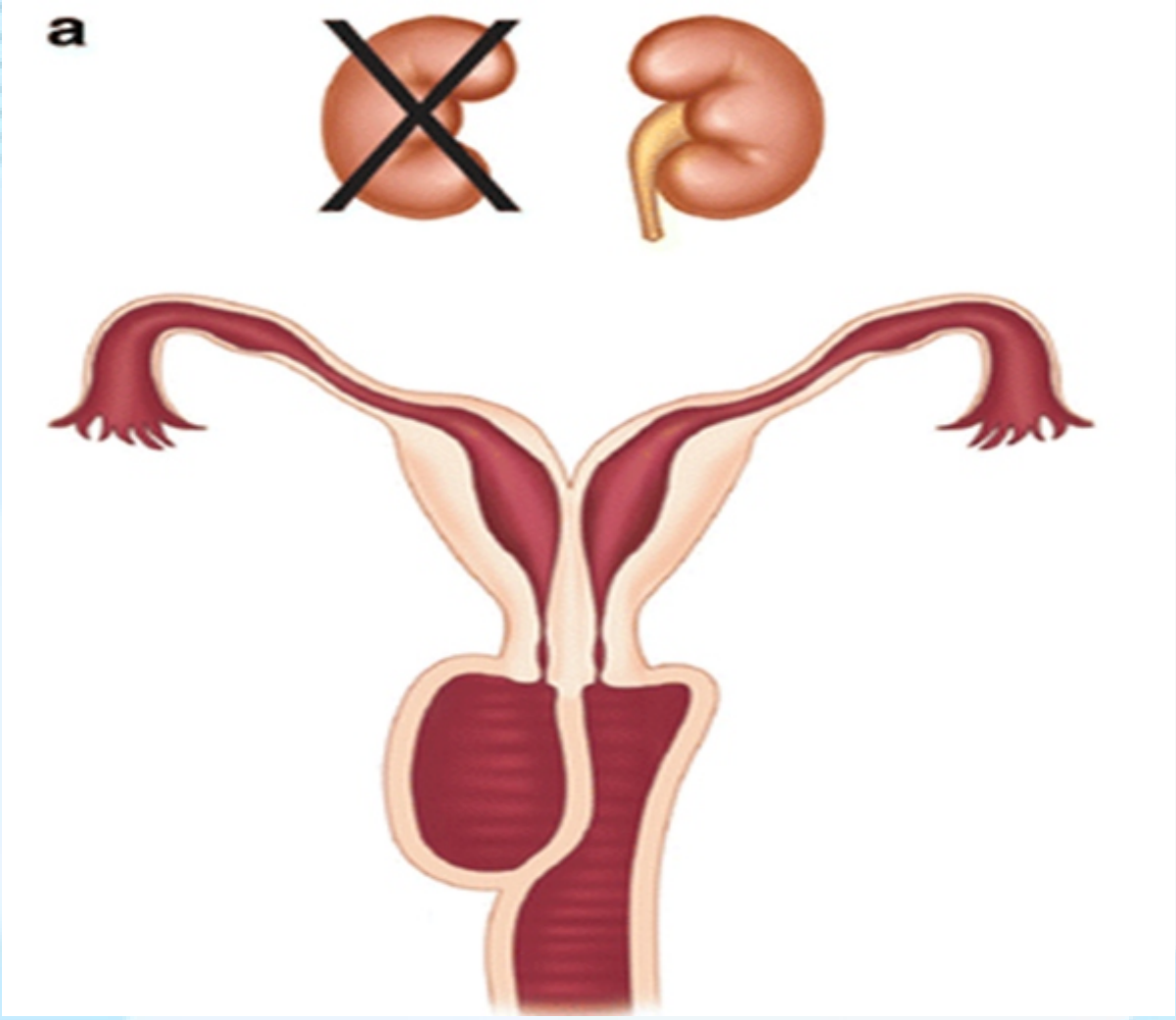


Partial vaginal Agenesis

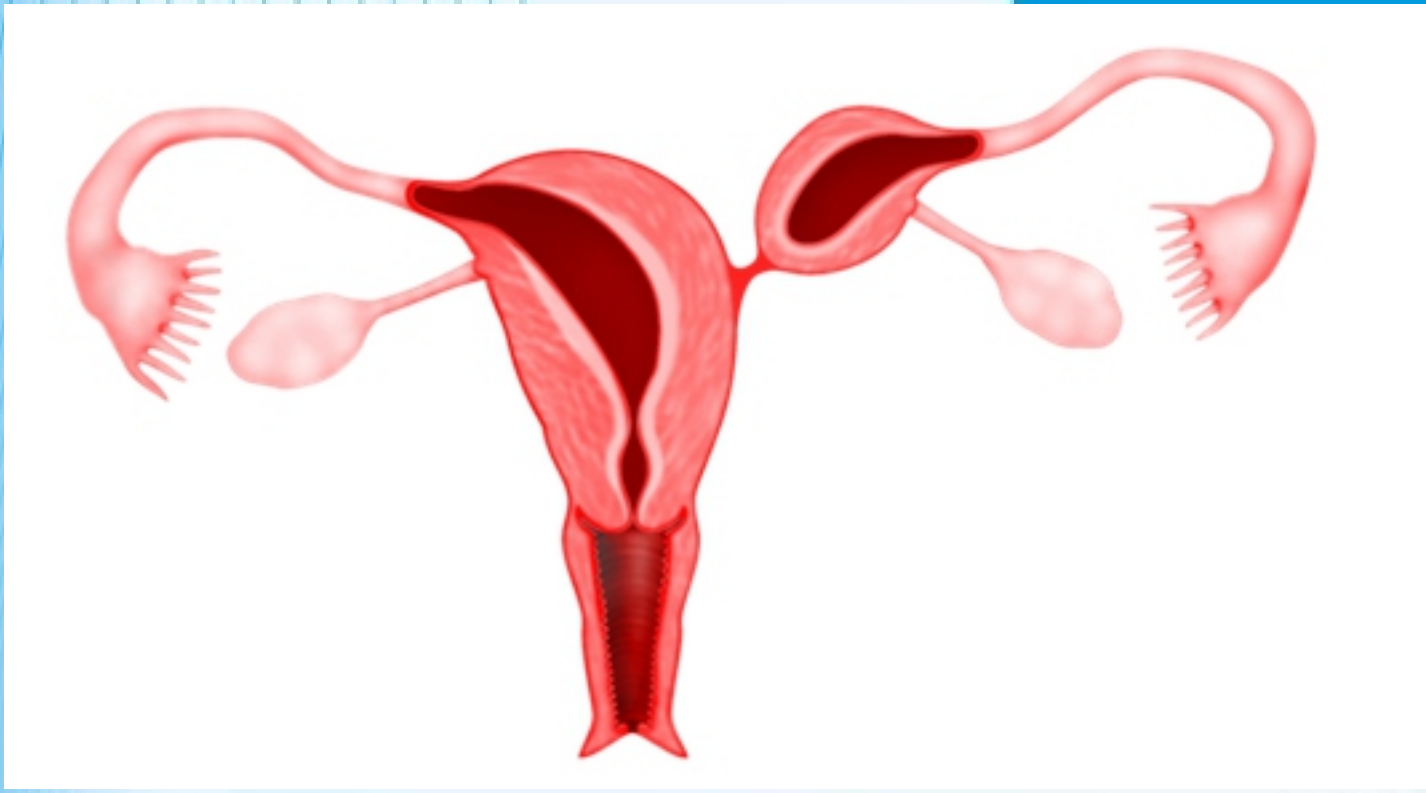

Amenorrhea
Primary
No onset of menarche by age 16 with secondary sexual characteristics
Or, No onset of menarche by age 14 without secondary sexual characteristics
Secondary
Absence of menses for 3 cycles or 6 months
Ovarian Etiology
High FSH
Low Estrogen
Receptor Abnormalities and Enzyme Deficiencies
Central
Low FSH
Low Estrogen
Congenital Outflow Tract Anomalies
• 46, XX Gonadal Dysgenesis (e.g. Fragile X, Balanced Translocations, Turner's mosaic)
• 46, XY Gonadal Dysgenesis (e.g. Swyer's Syndrome)
• 45, XO Turner syndrome
• Savage syndrome (ovarian resistance)
• Premature Ovarian Failure (Autoimmune, Iatrogenic)
• Androgen insensitivity
• 5-α Reductase deficiency
• 17- α Hydroxylase deficiency
• Vanishing Testes Syndrome
• Absent Testes Determining Factor
• Imperforate hymen
• Transverse vaginal septum
• Vaginal agenesis (Mayer-Rokitansky-Küster-Hauser syndrome)
• Cervical stenosis
Hypothalamic
Pituitary
• Functional (e.g. eating disorder, weight loss, stress, excessive exercise, illness)
• Congenital GnRH deficiency (Kallmann syndrome)
• Constitutional delay of puberty
• Surgery
• Irradiation
• Tumor, Infiltration
• Hyperprolactinemia
• Hypothyroidism

Pelvic Pain Associated with Menstruation.

Patients with a duplicated Müllerian system may have a complex configuration where one Müllerian duct is patent while the contralateral side is non-communicating. Menstruation occurs through the patent side. One of the more common conditions associated with this scenario is double uterus with obstructed hemi-vagina. This anomaly is usually seen in combination with ipsilateral renal agenesis and is known as "obstructed hemi-vagina and ipsilateral renal agenesis" (OHVIRA) or Herlyn-Werner-Wunderlich Syndrome (HWW). These girls menstruate from the patent tract but simultaneously develop hematocolpos with or without hematometra on the obstructed side.



Unicornuate uteri and contralateral functional rudimentary uterine horns can develop hematometra in the non-communicating uterine horn. This complex defect can present with unilateral cyclic abdominal pain associated with menstruation that is refractory to pain medications or hormonal treatments. With this condition, there is not usually a large pelvic mass, and the uterine horn does not distend significantly. With this condition, there is not usually a large pelvic mass, and the uterine horn does not distend significantly. Occasionally, a pelvic collection, pelvic infection, or even hemodynamic instability from a ruptured ectopic pregnancy may be the first evidence of a functional non communicating uterine horn.

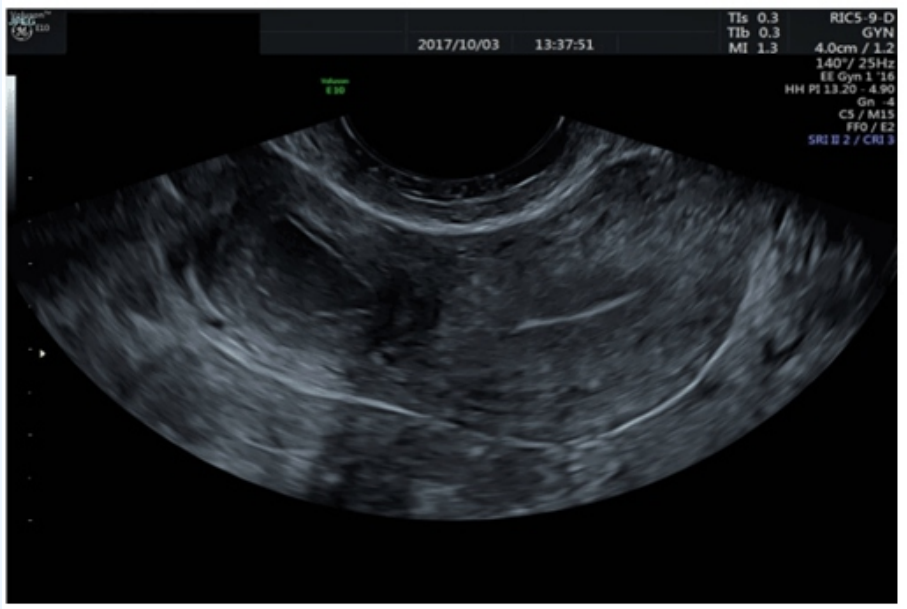


non communicating rudimentary horn

Unicornuate uterus with non-communicating horn should be differentiated from pedunculated subserous fibroid with degeneration. Typical history of cyclical pain is usually absent in fibroid cases.

ACUM (Accessory and cavitating uterine mass) represent a variant of Mullerian anomalies that are generally located at the level of the insertion of the round ligament and are possibly related to dysfunction of the female gubernaculum. It has significant clinical manifestations, particularly severe dysmenorrhea and recurrent pelvic pain. differential diagnosis, which includes rudimentary and cavitated uterine horns such as those found in other uterine malformations (unicornuate and bicornuate uterus), adenomyosis with cystic or degenerated areas, degenerated leiomyomas.

Ultrasound image of ACUM



Diagnosis- There are many options for imaging the female pelvis. Ultrasound is recommended for initial evaluation. However, MRI is an excellent non-invasive imaging technique that allows for greater tissue detail in different imaging planes. As 3D ultrasound is more accessible and accurate the use of MRI for evaluating uterine anomalies is limited to evaluation of complex Mullerian abnormalities and in adolescent girls.

Conclusion

Diagnosis of Müllerian anomalies should be based on symptoms, history, and physical examination. Family history of congenital anomalies or infertility should be obtained, as family associations with Müllerian anomalies have been described.

General physical examination and assessing sexual development with Tanner staging is very essential reaching diagnosis.

Precise radiologic diagnosis is necessary to guide surgical intervention and is imperative for appropriate fertility counselling. Transabdominal, transvaginal, transrectal, trans perineal, and 3D ultrasound have all been used to clarify the anatomy when an anomaly is suspected. Pelvic ultrasound is the preferred initial imaging modality. MRI is considered the gold standard for diagnosis of Müllerian anomalies and is used to assess the length of the vagina, the thickness of a septum, and to delineate the configuration and number of the cervix or cervices and uterus or uteri.

Mullerian abnormalities are associated with other system abnormalities especially renal, cardiac and skeletal system. Through assessment of these should also be the part of management

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Development of the Female Reproductive Tract and Associated Anomalies Janeen L. Arbuckle, MD, PhD* Kimberly H. Hoover, M , Curr Treat Options Peds (2016) 2:131–142 DOI 10.1007/s40746-016-0064-6

Assessing Anomalies: A Methodical Approach To Female Reproductive Tract Evaluation

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The prevalence of female genital tract anomalies is estimated to be around 4-7% in general population, with an even higher prevalence in the infertile women. However, specific anomalies may have different prevalence rates. Advances in imaging techniques has made diagnosis more easy and precise.

Reproductive tract anomolies can profoundly impact fertility, pregnancy and child birth, menstrual patterns, urinary and bowel functions and over all quality of life. Accurate diagnosis and correct classification helps in the appropriate counselling of women about their potential reproductive prognosis and to tailor the optimal treatment strategies and surgical approaches.

A methodical approach to narrow down available information from symptom and physical signs, followed by confirmation and classification of anamoly using specific investigative method will help in planning suitable treatment option.

Clinical presentation

Asymptomatic Cases

A notable characteristic of congenital anomalies of the female genital tract is that a significant proportion of affected individuals may remain asymptomatic. In these cases, the anomalies are often incidentally discovered during routine pelvic examinations, imaging studies or investigations for unrelated health concerns.

Reproductive Issues - Infertility

Female genital tract anomalies exert a significant impact on fertility as they directly affect the intricate processes of conception and implantation. Anomalies associated with complete agenesis of cervix, vagina and uterus do not allow fertilization and structural variations of normal anatomy like septate, unicornuate or bicornuate uterus can impede the optimal conditions required for the implantation of a fertilized egg, thereby affecting the chances of achieving a viable pregnancy.

Recurrent pregnancy loss

Certain congenital anomalies elevate the risk of recurrent pregnancy loss, creating additional hurdles for fertile couples. Anomalies like septate uterus, bicornuate uterus, unicornuate uterus can disrupt the regular vascular supply to the uterine lining and reduce intrauterine space, thus contributing to recurrent miscarriages.

Menstrual Disorders

Congenital anomalies of the female genital tract can influence menstrual patterns and lead to abnormal uterine bleeding, dysmenorrhea, or irregular menstrual cycles. The presence of a transverse vaginal septum can lead to obstructive symptoms, causing difficulties in the passage of menstrual flow. This collection causing hematocolpos, hematometra and hematosalphynx as a sequele can cause mass effect leading to intractable pain and pressure symptoms.

Urinary Tract and Bowel Symptoms Urinary symptoms

Female genital tract anomalies, particularly those involving the cervix and vagina, can manifest in urinary symptoms due to their same embryologic origin as urinary tract. In more complex cases, such as cloacal anomalies which involves the fusion of the urinary and reproductive tracts result in intricate urinary manifestations.

Bowel symptoms: Cloacal anomalies, characterized by the fusion of the rectum, vagina, and urinary tract, may result in shared channels and complex interactions affecting bowel function.

Medical History and Physical Examination Medical history

Initiating the diagnostic process for congenital anomalies of the female genital tract begins with a comprehensive medical history. This essential step involves gathering pertinent information such as menstrual history, reproductive experiences, any instances of infertility or recurrent pregnancy loss, and details regarding urinary or bowel symptoms. Patients presenting with severe dysmenhorrea around age of menarche hints on obstructive pathologies like imperforate hymen, transverse vaginal septum, cervical agenesis, active non-communicating rudimentary horn. Those with early pregnancy failure or recurrent miscarriages hint on structural defects causing uterine space obliteration like septum, unicornuate or bicornuate uterus. Furthermore, thoroughly examining family history is crucial to identify genetic factors contributing to these anomalies, allowing for a more holistic understanding of the patient's condition.

Physical examination

A detailed physical examination, including a meticulous pelvic examination, is indispensable in diagnosing congenital anomalies of the female genital tract. This examination serves to assess both the external and internal genital anatomy. Specific physical findings, such as the presence of a bulging bluish structure at introital level indicates Imperforate hymen / Transverse vaginal septum, the identification of a vaginal dimple or presence of band like thickening on rectal examination indicates abnormal cervical configuration hinting at cervical or vaginal agenesis. Specific examination techniques like per-rectal examination to assess cervical tissue and uterus in case of lower tract agenesis, two finger test to assess cloacal defects are helpful.

The insights gained from the history and physical examination contribute to developing a targeted diagnostic plan, guiding subsequent imaging and laboratory investigations for a precise and tailored diagnosis.

Diagnosis

A significant proportion of anomalies are diagnosed during fertility investigations. The gold standard test has been the combined laparoscopy and hysteroscopy in the past. Imaging modalities such as ultrasonography, hysterosalpingogram (HSG), sonohysterogram and magnetic resonance imaging (MRI) are less invasive modes of screening and classifying various uterine anomalies. While conventional 2D transvaginal ultrasound (TVS) and HSG are considered as good screening modalities, 3D TVS and MRI can accurately diagnose and classify the types of anomalies, as they can define both external and internal uterine contours.

Hysterosalpingogram

HSG helps in evaluating the uterine cavity but a definitive diagnosis of uterine anamoly requires evaluation of the external uterine contour, which is poorly defined by HSG. However, HSG, a commonly employed test to assess tubal patency as a part of fertility investigation, is a good screening test for congenital uterine anomalies. In addition, HSG can visualise the uterine cavity only if it communicates with the cervix and cases of non-communicating rudimentary uterine horn may be missed.

2D Transvaginal Ultrasound

Conventional transvaginal ultrasound is minimally invasive and a less expensive way of assessing uterine morphology and ruling out uterine anomalies. Ultrasound evaluation can be timed in the secretory (luteal) phase of the menstrual cycle as the endometrium, being bright and echogenic, is easy to visualise. The visualisation of a double endometrial complex on a transverse plane points towards a uterine anomaly and the differential diagnosis would be a bicornuate, septate, subseptate or arcuate uterus. However, 2D ultrasound fails to provide visualization of internal, external and cornual uterine features all in same plane thus making it difficult to accurately identify anomalies.

Saline infusion sonography may be helpful in diagnosing communicating rudimentary horns as saline can be clearly seen in the unicornuate uterus, with passage into the rudimentary horn.

Three-Dimensional (3D) Transvaginal Ultrasound

The 3D transvaginal ultrasound is considered the gold standard tool for the assessment of uterine anomalies as it is less invasive; it facilitates simultaneous visualization of both the external (serosal surface) and internal (endometrial) contours of the uterine fundus through its unique feature of providing the coronal plane of the uterus and, therefore, can correctly classify the types of uterine anomalies. The criteria for the classification of uterine anomalies based on 3D ultrasound have been well described in the literature for the first time by Salim et al. in 2003. The ESHRE/ESGE have subsequently described the criteria in the Thessaloniki consensus on the diagnosis of female genital anomalies.

The diagnostic accuracy of 3D ultrasound compared with laparoscopy ± hysteroscopy in diagnosing congenital uterine anomalies is highest among other imaging modalities including MRI. The diagnostic accuracy of 3D ultrasound is reported as 97.6% with sensitivity and specificity of 98.3% and 99.4%, respectively. The morphology of the uterus is best examined in the coronal plane using the interstitial portions of the fallopian tubes as reference points. A line joining the tubal ostia (interostial line) is the reference line from where measurements like uterine wall thickness, septal length, the extent of cleft in bicornuate uterus can be measured.

Magnetic Resonance Imaging (MRI)

MRI has the multiplanar capability, allows better soft-tissue characterisation and permits a wider field of assessment at once, than other imaging modalities including ultrasound. MRI, due to its ability to demonstrate both the external and internal contours of the uterus, is sensitive and specific for diagnosing nearly all uterine anomalies. MRI is helpful in delineating the endometrium and detecting uterine horns regardless of the uterine position and anatomical variation. Furthermore, it is accurate in defining aberrant gonadal location or renal anatomy and is less invasive compared to laparo-hysteroscopy. In addition, it does have the ability to examine the details of the nearby structures including the cervix and vagina. While MRI is not routinely recommended in all women suspected to have a uterine anomaly, it proves useful for those patients with suspected complex anomalies, particularly if there is a combination of cervical and vaginal anomalies and for those at higher risk for associated anomalies. MRI is also useful if there is any diagnostic dilemma with other imaging modalities.

Laparoscopy and Hysteroscopy

The role of Hystero-laparoscopy in diagnosing and also if possible treating the anomalous condition in same sitting was considered gold standard earlier. With the invent of 3D ultrasonography Hystero- Laparoscopic approach being invasive modality is mainly used as a treatment option than for diagnostic purpose.

Conclusion

Uterine anomalies are commonly seen in women presenting with a history of reproductive problems. While 2D ultrasound and HSG are adequate for screening for uterine anomalies, 3D ultrasound, MRI and combined laparoscopy and hysteroscopy can correctly classify the type of uterine anomaly due to their ability to show both external and internal contours of the uterus. While 3D ultrasound is now considered as the gold standard diagnostic tool for uterine anomalies due to its high degree of diagnostic accuracy, less invasive nature and it being comparatively less expensive, MRI is reserved for diagnosing complex Mullerian anomalies or if there is a diagnostic dilemma. Laparoscopy and hysteroscopy are an invasive modality for diagnostic purposes and should be offered only in the context of concomitant surgical treatment after a thorough non-invasive evaluation of a Mullerian anomaly. Imaging for renal anomalies is recommended if a uterine anomaly is diagnosed.

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Compassionate Care: The Essentials Of Psychosocial Counselling And Support For Women With Reproductive Tract Abnormalities

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FMMC Mangaluru

INTRODUCTION

Congenital malformations of the genital tract do not occur frequently. Hence the management of this uncommon condition is not freely available. What is however known is that the management both in terms of the psychological and the physical welfare of the patient needs a sensitive approach to consider it being optimal care.

CHALLENGES

The patients and relatives may experience various psychosocial problems that impact their lives. Psychological counselling was most frequently given to parents (74%), followed by children (39%), adolescents (37%) and adults (11%).

PARENTS AND FAMILIES

Diagnosing a child with a rare disorder, such as disorder of sex development (DSD), is often stressful for parents and families. The levels of stress in parents depends on various factors such as the diagnosis, type of disease, severity, cause, and presence of non-invasive, invasive, surgical, mental, and behavioral changes and treatment or therapy. Information is often lacking and if available is conflicted, hence increasing the anxiety levels.

Parents often experience extreme stress about the disclosure of the problem to family and friends and find uncertainty very unsettling. This is worsened if they were told the sex of the baby in antenatal period and then in the neonatal period this uncertainty develops. Parents worry about the stigma associated with the diagnosis. Four major domains parents struggle to make sense are : (1) the gender assignment process, (2) decisions regarding genital surgery, (3) disclosing information about their child's DSD and (4) interacting with healthcare providers.

PATIENTS

The problem is sometimes identified in the antenatal period but definitely known in the neonatal period. The child is unaware at this time and decisions affecting the child are handled by the parents. Psychosocial and social-environmental problems and management of DSD that are identified in earlier times can re-emerge during childhood and adolescence. There are crucial times when psychology has a role to play in relation to both medical management and individual development. Decisions about sex of rearing, surgical intervention, introduction of sex hormones, information about diagnosis and decisions about sex re-assignment, as well as routine support and monitoring of social and emotional well-being need this consideration. In addition, psychological input should be available for unforeseen issues, including, for example, parental or individual concerns about gender identity development, school-related problems and bullying. Women diagnosed with MRKH have a lasting negative psychological impact, but as yet little is known about how to manage this. Patient feedback and outcome research that has shown the long-term

psychological distress is associated with – among other things – surgery without consent, medical photography, repeated genital examinations, widespread disclosure or unhelpful phrasing of sensitive information. Women diagnosed with CAH as a child will continue to have a high incidence of anxiety and depression. Adults diagnosed with DSD are dissatisfied with binary gender, dissatisfied with the DSD terminology, fear of devaluation, negative body image, social isolation, non-entitlement to relationships, preoccupation with heterosexual intercourse, functional sexual difficulties, barriers to communication with significant others and experiencing normalizing surgery as dilemmatic. Overarching all of these is the anxiety, sadness and depression.

MANAGEMENT

PSYCHOSOCIAL COUNSELLING is a service provided by a skilled professional counselor to an individual, family, or group for the purpose of improving well-being, alleviating distress, and enhancing coping skills.

1. COUNSELLING ACTIVITIES:

In regards to the care of the individual with a DSD, important decisions are often taken on their behalf early in life by their family in collaboration with the multi disciplinary team and, therefore, psychological care needs to encompass the family unit. Psychological input is best considered as a process and not an event and is both routine, or emergency as issues arise. In order to impact the psychological health of the patients, psychological support and treatment for parents and patients includes quality of life assessment and identification of sociocultural factors affecting them.

COUNSELLORS:

Should be well trained and easily available. The attitude should be respectful and sensitive, open and understanding. Choices should be coercion-free and informed; neutral, understandable, and on evidence-based information. Shared decision making is a collaborative and confidential decision-making process ensuring quality, respectful, and timely care, and dignity.

2. INFORMATION

This should be given to the family in a consistent manner that is appropriate for their level of understanding, with awareness of religious and cultural beliefs and preconceptions. Optimally, contact with psychology is initiated at the time of first contact with the team and, as with all members of the team, a collaborative, trusting relationship should be actively fostered

Give information about the available psychosocial care services (for instance, counselling, psychotherapy, support groups or online support options). Make sure that the instructions to access these options are easy to follow and the process is confidential. Communication skills, respect, competence, involvement, and information provision and continuity of care are associated with better patient well-being. There are specific tasks at different stages of development, which should be planned with families from an early stage. In particular, families may require considerable support from the multi-disciplinary team to feel confident to give information to their child about their diagnosis and personal history in relation to this condition. The pacing and timing of giving information and making decisions is ideally based on individual circumstances, together with information about the DSD. Whilst decisions should be informed by evidence when possible, most often there is no 'right 'course of action that applies in all circumstances. Rather there are broad goals, which are best approached with flexibility on a case-by- case basis. With this in mind, families and individuals require clear information about DSD and, sometimes, intensive input to empower them to

successfully address challenges as they arise. Parents have to make decisions on gender assignment and genital surgery and information management.

In childhood and adolescence these decisions will be needed to be revisited. Diagnosis and management of mental health needs of child; formal evaluations of child's cognitive status to assess child's ability to participate in decision-making; assessment of parent/child relationship and facilitation of healthy parent/child relationship need to be undertaken.

This is the real-world phase and so gender reassignment, sexuality and information management need to be dealt with the patient itself.

3. COPING STRATEGIES

Most importantly, however, the family should feel supported, and strategies for coping with the uncertainty of gender assignment will be discussed with them. In particular, advice as to how best to impart information to family and friends will be given, which helps to alleviate some of the stress that the family inevitably feels that they are under.

4. PSYCHOTHERAPY OPTIONS

Cognitive behavioral and problem-solving psychosocial interventions have demonstrable efficacy in improving the psychosocial functioning of patients and families in other conditions. Examples of this can be found in group work drawing on cognitive and narrative approaches improving self-evaluation of women with Turner syndrome, cognitive behavioral therapy reducing specific stresses, and psychological interventions improving well-being for women with Mayer–Rokitansky–Küster– Hauser Syndrome.

5. SUPPORT OPTIONS

Peer support is a key component. This relieves patients from isolation and provides a unique source of identity support, anticipatory guidance and medical information accessible to individuals of all levels.

Group psychological interventions can improve well- being for women with MRKH syndrome, and group cognitive behavioral therapy can reduce specific stresses. Group work drawing on cognitive and narrative approaches appears to improve self-evaluation of women with Turner syndrome.

Online support options: should be provided so that counselling is available in an emergency with no delays.

6. MEDICATIONS:

There may be need for medications to treat anxiety and depression to sometimes need for anti-psychotic drugs.

CONCLUSION

In conclusion, psychosocial care for patients and relatives affected by a disorder of sex development is currently limited. Families struggle with challenges such as genetic information, medical complexity, anatomical differences (whether or not surgery is done), surgical complications, lack of clarity in some gonadal tumors risks, doubt about the stability of the child's gender identity, and fertility potential. There is a clear role for a psychologist or similar professional in the multi-disciplinary team. This dynamic integration of medical and psychological support that provides the best model of

Over Comming Absence Strategic Management of Mullerian Agenesis Patients

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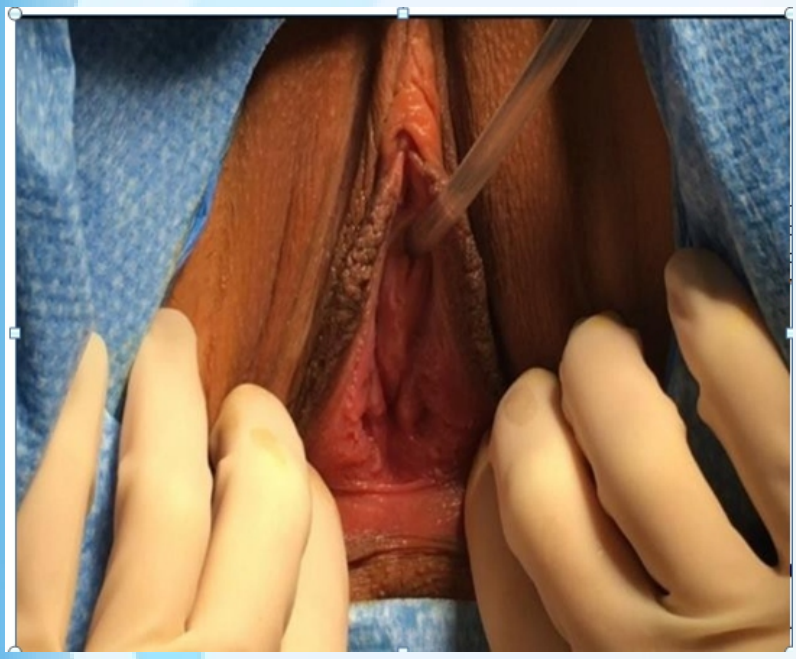
INTRODUCTION

Mullerian agenesis is also referred to as ***Mullerian aplasia*** or ***Mayer-Rokitansky-Kuster- Hauser syndrome*** or ***Vaginal agenesis***. As per the ACOG 2018, the incidence is said to be around 1 per 4500-5000 females(1).

Mullerian agenesis occurs due to the under development of the mullerian duct during the embryonic period – leading to the absence or atresia of the vagina or the uterus or both. In MRKH syndrome, the vaginal canal is usually shortened and may appear as a dimple below the urethra. Uterine remnants, including a single midline remnant or uterine horns, may be present. The ovaries, which develop from a different embryonic origin, are typically normal in structure and function, but may be found in unusual locations(2).

Patients with MRKH syndrome have normally developed secondary sexual characters, normal external genitalia, 46 XX karyotype, functional ovaries with a rudimentary or absent uterus and an absent vagina.

Figure 1- Patient with primary amenorrhoea. O/E patient had normal secondary sexual characters, normal external genitalia with absent vagina (vaginal dimple+).



Two different forms of mullerian agenesis are present –

Type A – the typical form – congenital absence of uterus and upper 2/3rd of vagina with normal ovaries and tubes.

Type B – the atypical form – with associated abnormalities of the ovaries, fallopian tubes, and renal abnormalities.

DIAGNOSIS

The diagnosis of MRKH is usually made during adolescence on evaluation of primary amenorrhoea with otherwise normal growth and secondary sexual characteristics. Occasionally patients may have cyclical pain or chronic abdominal pain due to the presence of endometrium in the rudimentary uterus.

On physical examination, individuals with mullerian agenesis exhibit normal height, breast development, body hair distribution and external genitalia. The vagina may be seen as a small dimple below the urethra or may be present as a longer structure but may lack a cervix at the vaginal apex(2).

A small rudimentary uterus or uterine horns with or without an endometrial cavity may be seen in the midline. The ovaries are usually normal and functional as they arise from a different embryological structure. Sometimes fibroids may be seen in the absence of uterus as well – the management of which is the excision of fibroid followed by vaginoplasty(3).

The 2 main imaging options are ultrasound and magnetic resonance imaging (MRI). Ultrasound may not always effectively identify underdeveloped müllerian structures and ovaries, which are typically located high in the pelvis, often at the level of the pelvic brim(2). For surgical planning, MRI is the preferred method, despite being more expensive than ultrasound – showing rudimentary mullerian structures in 90% of cases. These structures are challenging to interpret in ultrasonography – which may be misleading before puberty(2).

Laparoscopy may be necessary in some cases, especially when pelvic symptoms are present due to uterine horns and müllerian remnants containing functional endometrium. However, it is not the preferred diagnostic tool because it is invasive and requires general anesthesia(4).

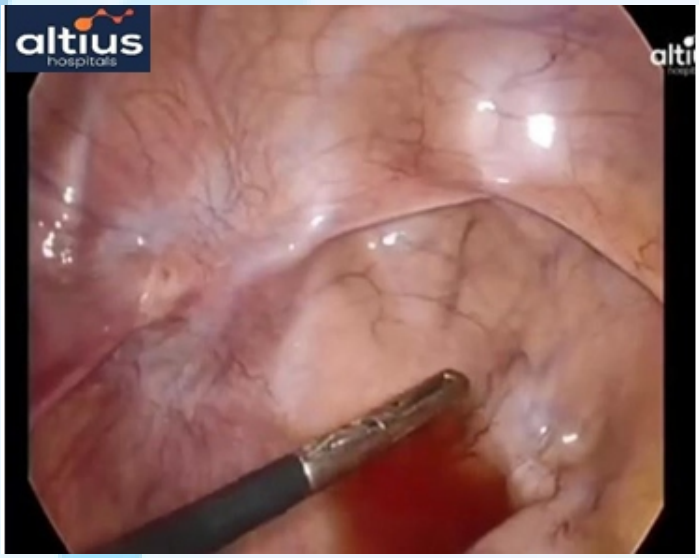


Figure 2- Laparoscopic image of a patient with MRKH syndrome showing absent uterus

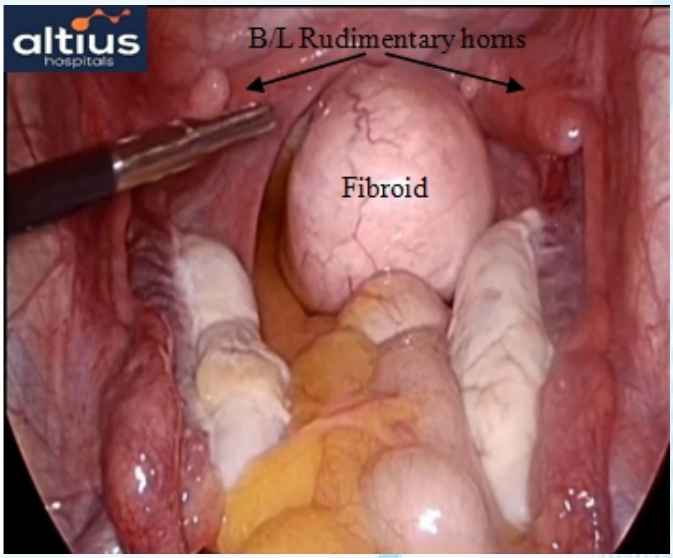


Figure 3- Laparoscopic image showing a large fibroid in the absence of uterus.
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EVALUATION OF MULLERIAN AGENESIS

The ***initial evaluation*** of a patient without a uterus may include the following laboratory tests:

* Testosterone level,
* Follicle-stimulating hormone (FSH) level,
* Karyotype analysis, and
* Imaging – to assess for the presence of a midline uterus.

* Ultrasonography
* MRI – Rudimentary uterus and presence of endometrial cavity within the rudimentary structures may be better visualized by an MRI scan(2).

Assessing for associated congenital anomalies is crucial in müllerian agenesis as up to 53% of patients may have additional congenital malformations, particularly affecting the urinary tract and skeleton(5).

* ***Renal anomalies*** in patients with mullerian agenesis have been around 27-29%(2).
* ***Skeletal anomalies*** have been reported in approximately 8–32% of patients like(2)

* scoliosis,
* vertebral arch disturbances,
* hypoplasia of the wrist.

Therefore, an ultrasound scan as well as an XRAY may be crucial even in asymptomatic patients. Mullerian agenesis may also be associated with VACTER/ VACTERL association(2).

Karyotype evaluation of patients with müllerian agenesis will be 46XX in most individuals

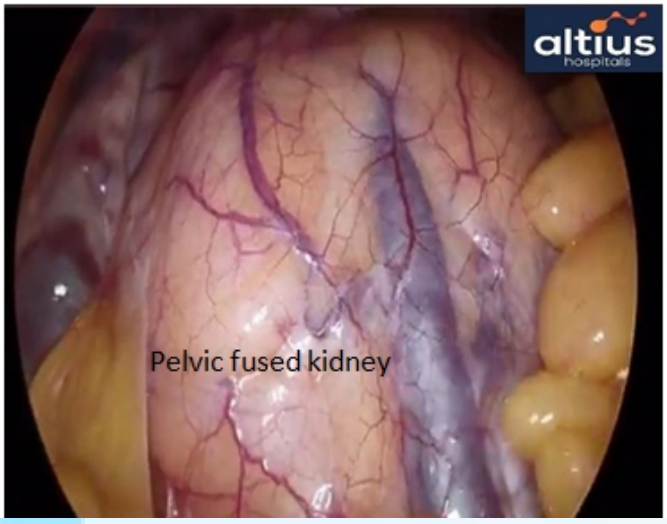


Figure 4- Laparoscopic image of pelvic fused kidney with dilated renal vessels.

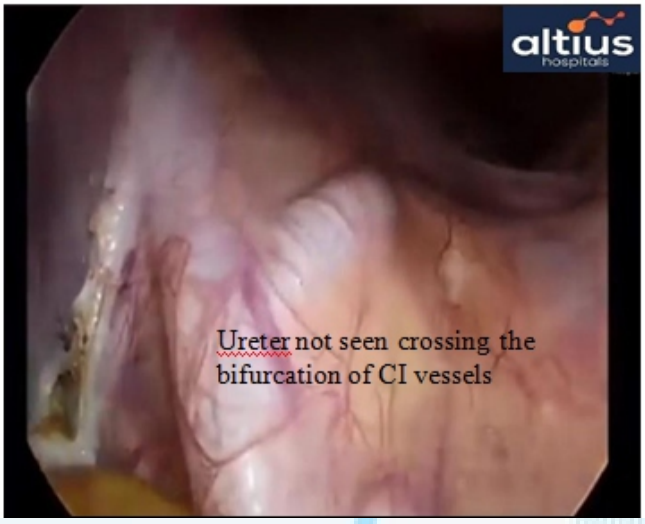


Figure 5- Laparoscopic image of absent kidney and ureter on left side – ureter not seen crossing the bifurcation of the common iliac vessels.

PSYCHOSOCIAL COUNSELLING AND SUPPORT

After receiving a diagnosis of MRKH syndrome, many young women often experience anxiety and significant psychological distress. Hence, it is crucial for physicians to provide thorough and sensitive counseling to these patients before discussing treatment options. The initial approach and manner in which these patients are treated will greatly influence their emotional well-being and long-term outlook.

It is recommended to use brief assessments of psychological distress regularly, both after the initial diagnosis and following treatment, to monitor the patient's emotional well-being over time(4).

The successful creation of a functional neovagina can help address what often manifests as "sex role insecurity" in MRKH patients. The absence of a uterus and vagina can raise questions about their ability to fulfill the female sex role, both personally and socially. It is crucial to emphasize to patients, once the diagnosis is made, that with treatment, they can have sexual intercourse and build healthy sexual relationships.

While successful treatment of the anatomical abnormality is central to achieving sexual well-being, the addition of psychological support and adequate information is equally important for fulfilling the complexities of female sexual response.**FERTILITY OPTIONS**

Patients with MRKH have the option of building a genetically related family through the use of **in vitro fertilization (IVF)** and a **gestational carrier**. This allows them to have biological children even if they cannot carry a pregnancy themselves due to the absence of a uterus. Ovaries are typically normal in MRKH patients, allowing for oocyte retrieval and embryo transfer to a gestational carrier for pregnancy(4).

Ovulation monitoring is used to determine menstrual cycle timing and enable the use of standard protocols for ovarian hyperstimulation in MRKH patients. Egg retrieval may be challenging and can be done either vaginally or abdominally – using transabdominal scan or laparoscopically.

However, the response to treatment, including the number of oocytes retrieved, fertilization rate, and embryo quality, is often slightly lower than average. Consequently, pregnancy rates among MRKH patients undergoing assisted reproduction techniques have been reported to be below average for infertile patients.

In MRKH patients, the unique pelvic anatomy may necessitate oocyte retrieval via a transabdominal route rather than the typical transvaginal approach. Despite the small number of reported pregnancies after IVF in MRKH patients, this method has become an appealing option for women who previously felt hopeless about having children(4).

Uterus transplantation (UTx) represents a groundbreaking advancement in infertility treatment for women with MRKH syndrome, offering them the opportunity for complete

motherhood, including gestation, genetic connection, and legal recognition from the start(6). During UTx surgery for MRKH women, a sub-umbilical midline incision is typically made. The vaginal vault is then dissected free from the bladder and rectum, often by cleaving the uterine remnant in the midline. The external iliac arteries and veins are dissected and cleaned to prepare for anastomosis. Fixation sutures may be applied to the sacro-uterine ligaments, round ligaments, and cardinal ligaments(6).

The chilled and flushed uterus is then brought into the pelvis from the back-table. End-to-side anastomoses are performed, connecting the internal iliac segments on the uterine vessels of the graft to the external iliac vessels of the recipient. After reperfusion, the vagina is opened for end- to-end vaginal anastomosis, followed by fixation of the uterus to the ligaments(6).

Unlike other solid organ transplants, UTx is ephemeral, meaning that immunosuppression is only required for a limited time. Immunosuppressive medication can be withdrawn when the graft is removed, typically within 5 years, once the transplanted woman has given birth to the desired number of children. This approach helps avoid the negative long-term side effects of calcineurin inhibitors, such as nephrotoxicity, which is particularly important in the context of MRKH syndrome, where several patients have single kidneys(6).

TREATMENT OPTIONS

The key components of effectively managing mullerian agenesis include,

* Accurately diagnosing the condition,
* Evaluation for any associated congenital anomalies,
* Psychosocial counselling,
* Implementing treatments or interventions to address the functional effects of genital anomalies.
* Future options for having children.

Nonsurgical vaginal elongation by dilation should be the first-line approach(2).

NON-SURGICAL METHODS	SURGICAL METHODS	
	Traction-based methods	Graft-based methods
Frank's method	*Vecchietti vaginoplasty*	*Abbe- McIndoe vaginoplasty*
Ingram's method		*Baldwin vaginoplasty*
D'Alberton's method		*Davydov's vaginoplasty*

NON-SURGICAL CREATION OF A NEO-VAGINA

In most patients, the initial treatment approach for müllerian agenesis involves the non-surgical creation of a neo-vagina, which is preferred due to its lower morbidity and as primary vaginal dilation is successful for more than 90–96% of patients(2).

The most commonly used non-surgical method includes Frank's dilator method and the Ingram
method(4).

Dilatation (Frank's technique) is a common method used, and more than 90% of patients achieve functional success with this approach(2). It was introduced in 1938 by Frank, and involves the use of a vaginal mold as a dilator device(4).

Method – The dilation process involves placing successive dilators on the vaginal dimple for 30 minutes to 2 hours each day. The patient gradually increases the length and width of the dilator over time. This requires the presence of a short vaginal dimple to start with and typically takes around 6 months to attain a functional depth and width of the vagina. The challenges to this procedure include cramping and fatigue, lack of comfort, privacy issues, and difficulty in finding time to dilate daily(4).

Ingram aimed to address these challenges by utilizing the patient's body weight and gravity to aid in dilation. They described a method that involved attaching progressively larger dilators to a bicycle seat. Patients would sit on the seat, slightly leaning forward to provide perineal pressure. This method required patients to engage in 15–30-minute intervals of dilation for at least 2 hours per day. Similar to the Frank method, the dilators would increase in size gradually(4),(2).

This method requires highly motivated and emotionally mature patients, as achieving the desired outcome may take several months. Support from other women who have successfully undergone dilation can be beneficial in motivating these patients(2).

The advantages of any of these methods include no hospitalization, patient control, cost-effectiveness, and minimal morbidity and complications. Although there are clear advantages with the non-surgical methods, the disadvantages include the length of time required to achieve a functional vagina, discomfort, and increased risk of vaginal prolapse(4).

An alternative non-invasive option is dilation by intercourse, also known as ***d'Alberton's method.*** This approach has been reported to have good anatomical and functional outcomes compared to self-dilation and surgery(6). However, this requires regular coital activity with a partner.

The three methods based on dilation of the vaginal dimple will provide the vagina with a normal mucosal lining. This can be advantageous in a uterus transplantation situation, as it ensures a normal vaginal microbiota. This may be important for the success of embryo transfer and for accurately grading rejection by cervical biopsy(6).**SURGICAL CREATION OF A NEO-VAGINA**

Surgery becomes an option for patients who are unsuccessful with dilators or for those who prefer a surgical approach. The goal of the surgery is to create a vagina that is in the correct axis, of sufficient size, and with secretory capacity to enable intercourse without the need for ongoing postoperative dilation.

Typically, surgery is considered in late adolescence when the patient is more mature and better able to comply with postoperative dilation and instructions. Various surgical techniques have been employed for müllerian agenesis. Regardless of the approach, it is crucial for the surgeon to be experienced with the procedure, as initial surgeries are more likely to succeed than follow-up procedures. Repeat surgeries carry increased risks of operative injury to surrounding tissues and the possibility of a poor functional outcome. The decision of which surgical method to offer is often based on the surgeon's personal experience and preference. Currently, there is no consensus regarding the best surgical correction method(2).

The surgical treatment could be -

- **Traction based techniques** –
  - Vecchietti method (open or laparoscopic) and

- **Graft based techniques** –
  - Abbe-McIndoe Vaginoplasty (Vaginal approach),
  - Baldwin Vaginoplasty/ Intestinal vaginoplasty (open and laparoscopic)
  - Davydov's Vaginoplasty (Combined vaginal and laparoscopic approach using peritoneum)(4).

ABBE-MCINDOE OPERATION

The Abbe McIndoe procedure is the most commonly used procedure for creating a neovagina in müllerian agenesis.

Procedure

- This technique involves ***dissecting a space between the rectum and the bladder***, followed by ***placing a mold covered with a split-thickness skin graft*** into the space. The graft may be taken from the anterior thigh or buttock, or it could be a segment of the bowel, or a bio-engineered tissue.
- The ***mold with the split skin graft*** stays in place ***for the first 7 postoperative days***. After the initial healing from the surgery, the patient has a functional vagina.

Postoperatively, diligent vaginal dilation or frequent intercourse is essential to prevent skin graft contracture.

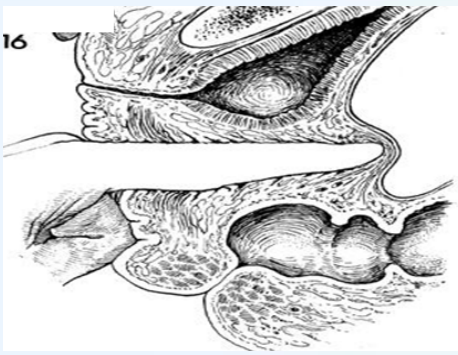


Figure 6-

Blunt dissection to open the space between the bladder and rectum – the correct plane being – below the pubo-vesical cervical fascia and superior to perirectal fascia.

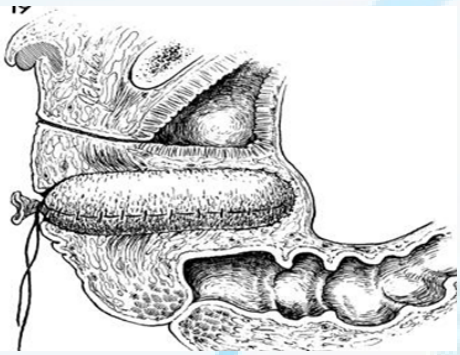


Figure 7-

Vaginal mold with split skin graft placed inside the newly created vagina.

Failure to perform postoperative dilation can result in a non-functional vagina (2). A modification to the original McIndoe procedure is the use of autologous in-vitro-cultured vaginal tissue as the graft material(7).

Baldwin's vaginoplasty involves using a segment of the sigmoid colon, ileum, or jejunum to create a neo-vagina.

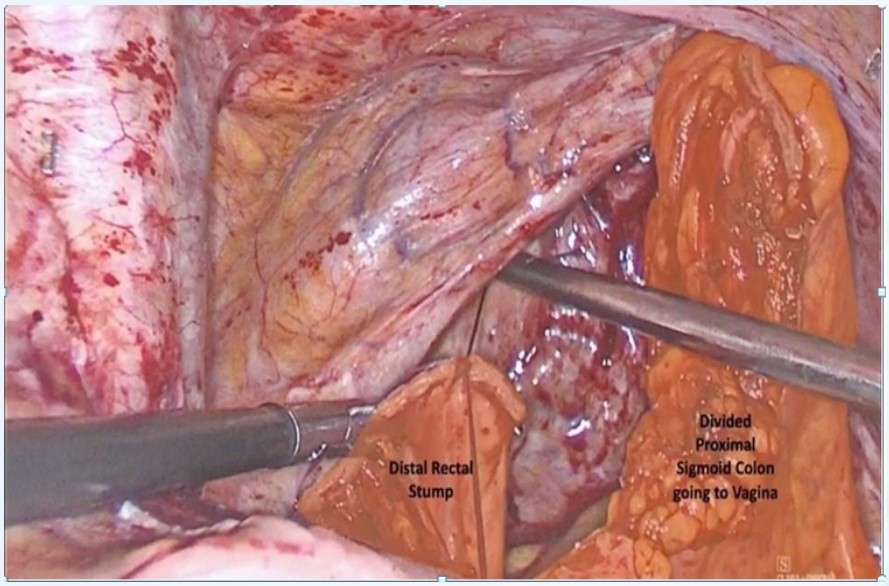
Advantages of the procedure

- Low risk of tissue shrinkage
- Has its own blood supply unlike skin grafts – chances of necrosis lesser.
- It offers distensibility and compliance which are not available with skin grafts – thus reducing the need for long term vaginal dilatation(8).

Limitations of the procedure

- The neovagina produced by this method can secrete a lubricating fluid; however, at times, the discharge can be excessive.
- Requires bowel resection and anastomosis.
- Prolapse of the neovagina(8)

Figure 8-
Sigmoid vaginoplasty Harvesting the intestinal segment typically requires a laparotomy, although some cases have been reported using laparoscopic approaches with bowel resection and anastomosis. The laparoscopic approach may be associated with increased morbidity compared to traditional laparotomy(4),(9).



(10) **Figure 8-** Sigmoid vaginoplasty

VECCHIETTI PROCEDURE

The Vecchietti procedure involves using a traction device attached to the abdomen to gradually create a vagina through traction. The procedure was first described in 1965 by Giuseppe Vecchietti(11).

It was initially developed as an open technique and has later been modified using a minimally invasive laparoscopic approach. This involves placing sutures through the pre-vesical space to a traction device on the ventral abdominal wall, establishing traction on a bead positioned in the perineum(11).

Advantages of the procedure

- Systematic dissection of the vesico-rectal space is not indispensable.
- Short duration of anesthesia
- Shorter postoperative stay
- No laparotomy scar in laparoscopic Vecchietti procedure

Limitations of the procedure

- Pain with continued tightening of the traction device
- Potential abdominal or pelvic complications due to traction thread placement
- Discomfort and inconvenience in wearing an indwelling vaginal form for months to prevent vaginal contraction and stenosis(11).

Mechanism

This approach uses surgical methods to create a vagina through tissue stretch. The continuous upward pressure from the traction bead – stretches and elongates the vaginal agenesis dimple, creating a mucosal cavity(11).

Procedure

Diagnostic laparoscopy is conducted in order to assess the pelvic peritoneum, pelvic structures – including presence of absence/ hypoplastic uterus and tubes, the ovary status, presence of endometriosis, other pathology like pelvic kidney/ other renal problems as well as the state of the recto-vesical space.

A **small *transverse incision*** is made ***over the vaginal vestibule***. The ***rectovaginal space is dissected*** until the peritoneum is reached. Care should be taken to identify the correct plane to prevent injury to the bladder and rectum. Doing it under laparoscopy guidance gives advantage to prevent this injury.

Figure 19- 23yr old with primary amenorrhoea with absent cervix and vagina with hypoplastic uterus with 46XX karyotype and female secondary sexual characteristics.

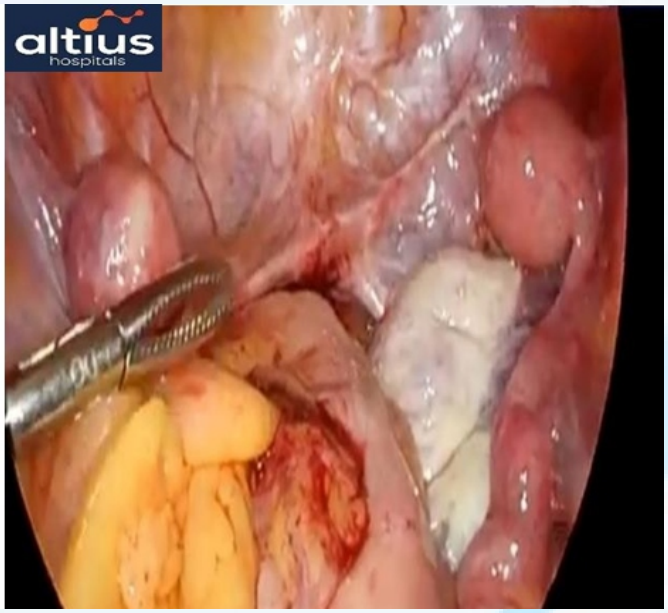


Figure 20- Laparoscopic view of hypoplastic uterus with B/L rudimentary horns with normal ovaries – rudimentary horns excised.

Laparoscopically, the peritoneal strand connecting the rudimentary uterus is lifted and incised transversely and ***peritoneum is mobilized***. *Rudimentary horns* of uterus *if present, are excised* so as to remove any endometrial tissue which may be a cause for cyclical pain.

The ***recto-vesical space is created*** laparoscopically to expose the vaginal plate. The correct plane has to be identified and dissection proceeded in between the bladder and rectum at the obliterated vaginal plate.

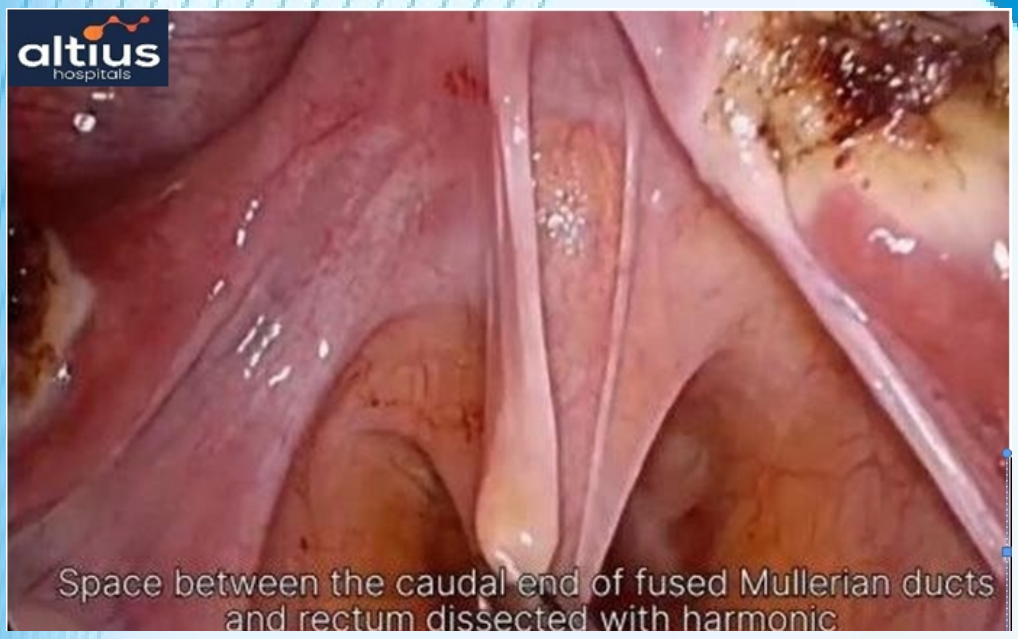


Figure 21-

Recto-vesical space created laparoscopically between the rectum and bladder – care is taken to avoid injury to bladder and rectum.

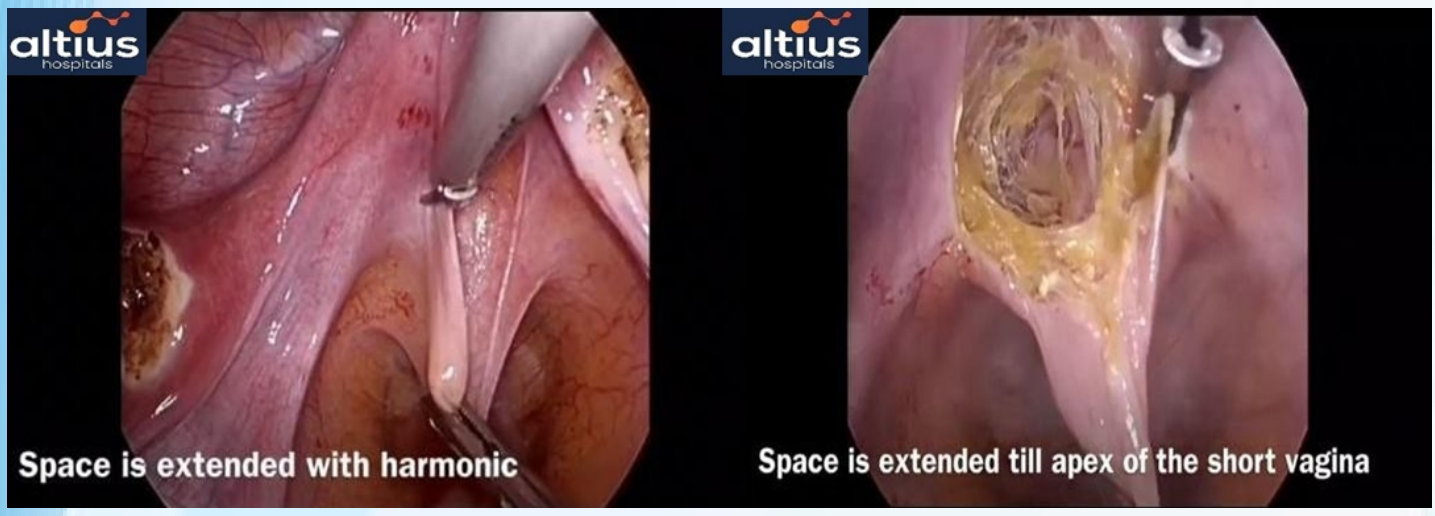


Figure 22- (left) Space created between the rectum and bladder using harmonic; **(right)** Incision extended below and dissection done till the apex of blind vagina.

A gauze is pushed vaginally for easy identification of the vaginal plate. Using the cutting blade of the harmonic, a ***transverse incision made over the vaginal plate*** pushed by the vaginal gauze from 1 uterosacral to the other. The medial fibers of the levator ani may be cut to reduce the risk of constriction of the neovagina later on. Dissection is continued till the perianal skin is visible.

Figure 23- Gauze pushed vaginally with a help of a sponge and holder from the vaginal end.

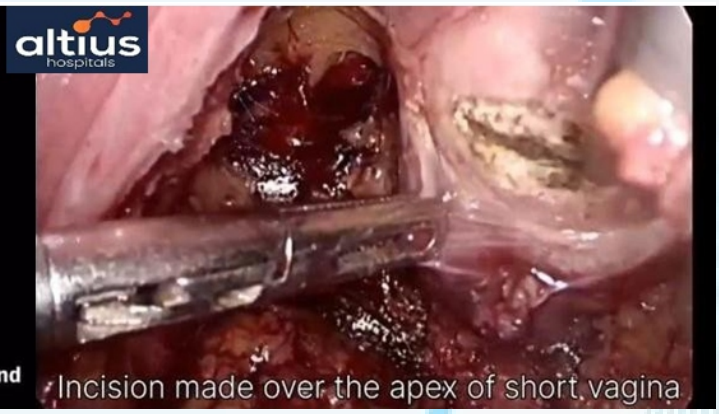


Figure 24- Once the apex of the short vagina is reached after dissection – incision is made over the vaginal plate assisted by a vaginal gauze.

Anteriorly the ***bladder peritoneum is mobilized*** which becomes the anterior wall of the vagina and posteriorly the ***rectal peritoneum mobilized*** becoming the posterior wall of the vagina. The peritoneum is then separated from the underlying bladder and rectum. The ***incision is extended laterally*** along the uterosacrals on both sides – to have a larger vaginal plate and so that the vagina does not shrink easily.

The success of the surgery is determined by how well the peritoneum is mobilized.

During mobilization of the peritoneum, the ***ureter is dissected and freed from the peritoneum*** to prevent injury to the ureter during the peritoneal pull through to the vagina.

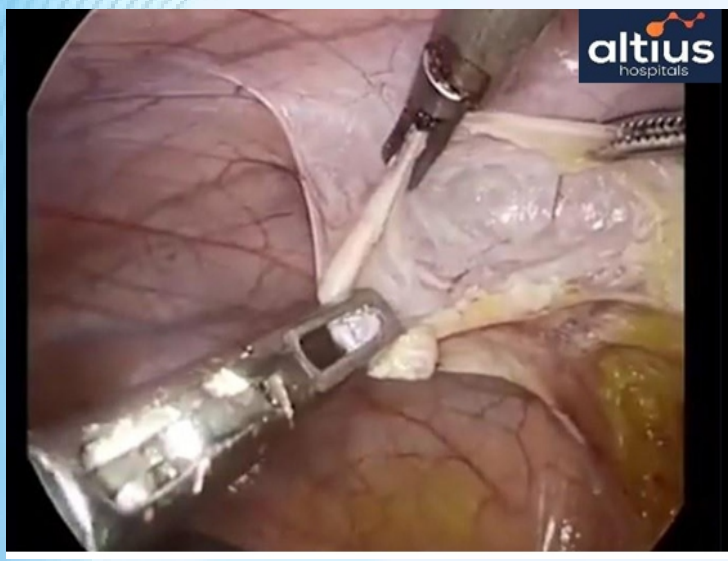


Figure 25- Anterior bladder peritoneum dissectedposterior to the bladder fat – to create the anterior wall ofthe neovagina.

Figure 26- Incision extended laterally along theuterosacrals to the pararectal space to mobilize theperitoneum – to create the AL walls of the neovagina.

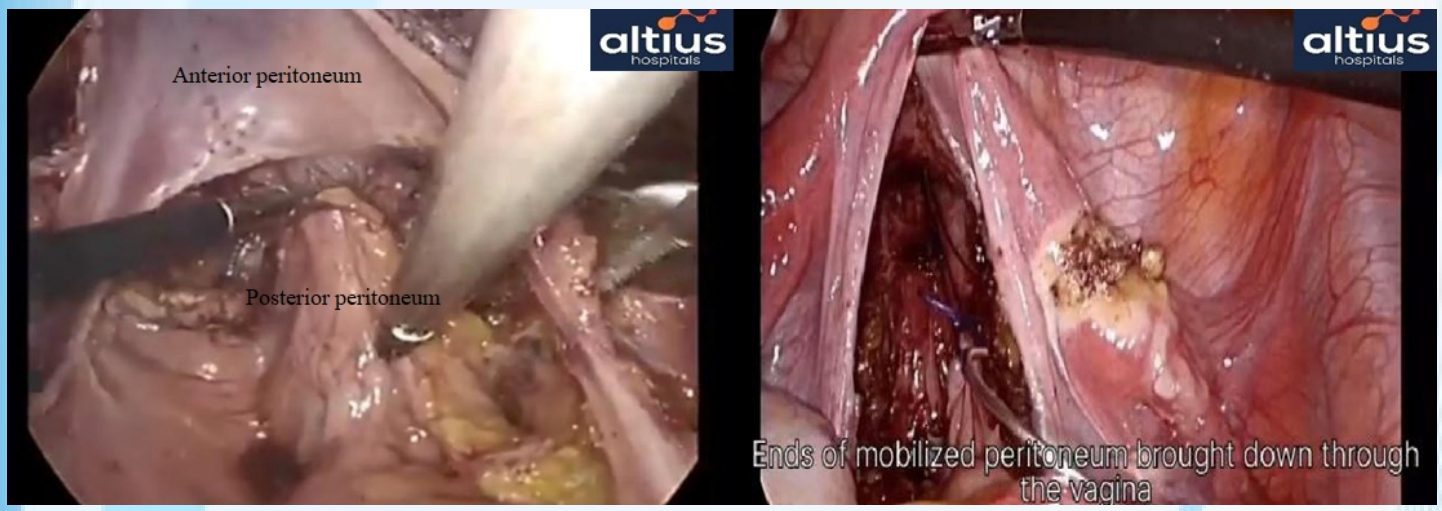


Figure 27- (left) Anterior and posterior peritoneum mobilized;
(Right) the mobilized peritoneum brought down through the vagina.

The ***mobilized peritoneal strand*** is opened and ***pulled anteriorly and posteriorly downwards***. Anterior and posterior peritoneal sutures are taken at the upper cut edges and brought out separately to the vaginal apex. The ***peritoneum*** is then ***sutured to the edges of the vaginal mucosa***.

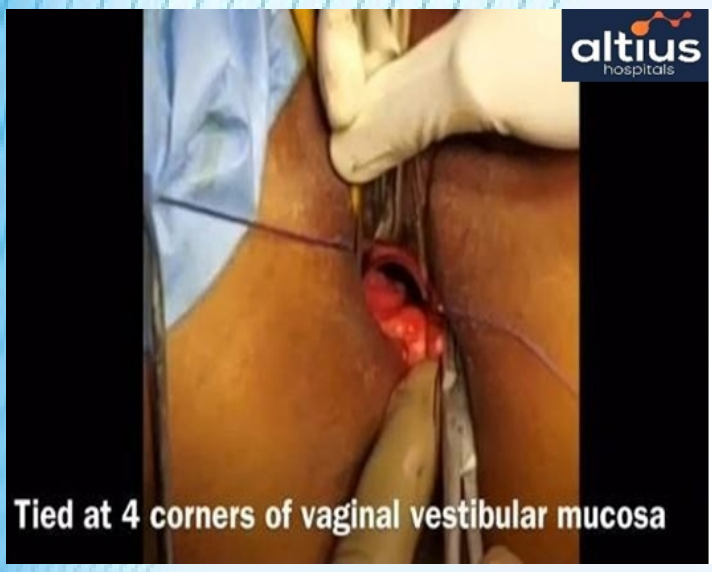


Figure 28- The peritoneum is then sutured to the vaginal mucosa on all 4 corners.

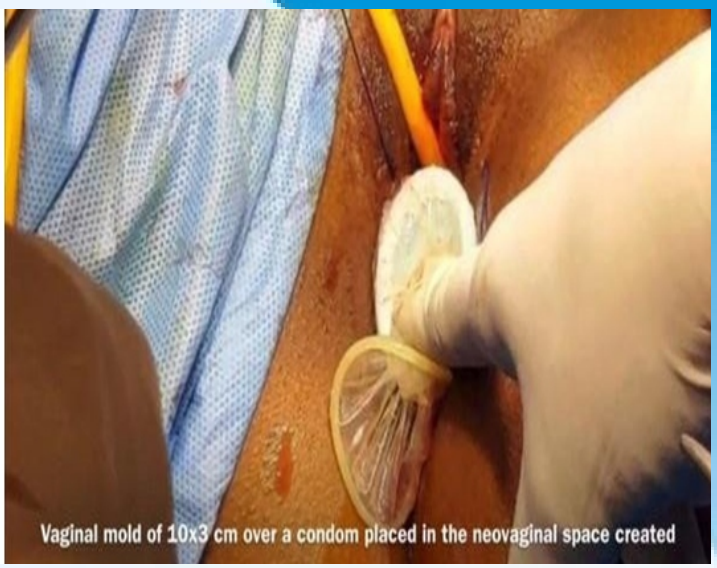


Figure 29- Vaginal mold covered with a condom is inserted into the neovagina.

A ***vaginal mold is then placed*** inside the newly created vaginal space. Purse string sutures are then taken to transfix the round ligament, tubal isthmus, ovarian ligament, lateral peritoneal leaflet, and rectal mucosa – to reinforce the vaginal vault. The ***peritoneum is closed abdominally over the vaginal mold***. The integrity of the newly created neovagina is confirmed at the end of the surgery. Indwelling foley's catheter is left in situ for bladder drainage.

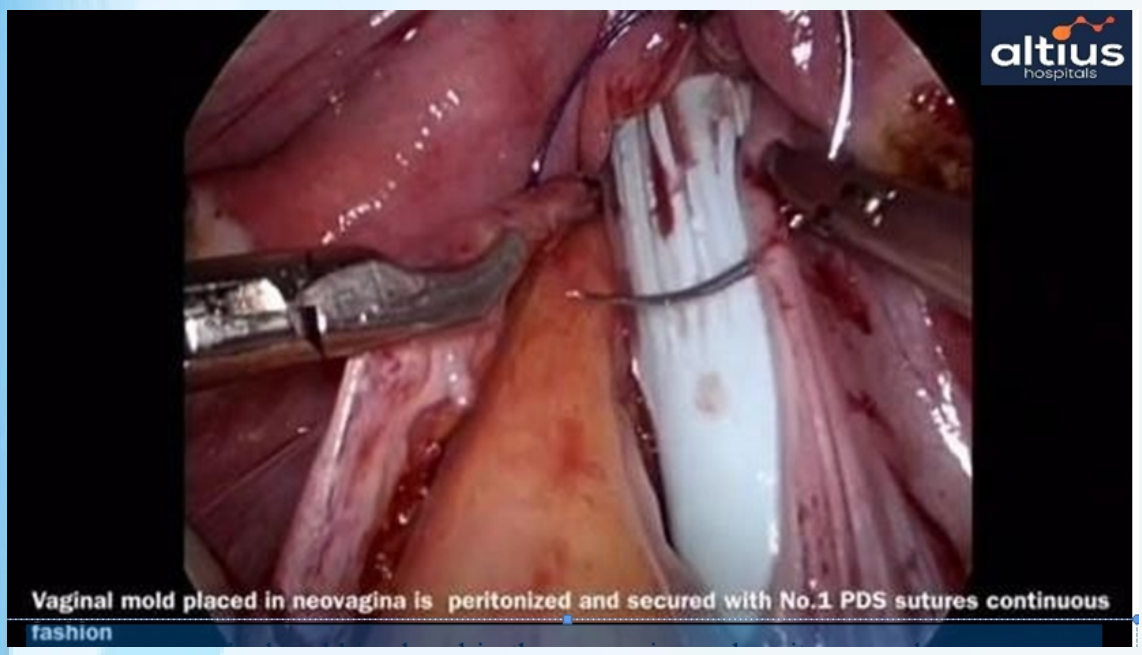


Figure 30- Vaginal mold is placed in the neovagina and peritoneum closed around it with delayed absorbable sutures in continuous manner.

The vaginal mold is inserted for 6 weeks to 6 months postoperatively and regular vaginal dilators are used until commencement of sexual activity. Regular vaginal examinations and cervical dilatation are necessary in the follow-up.

Anatomic success of vaginoplasty is defined as a neovagina width allowing the insertion of 2 fingers and a neovaginal length of at least 6 cm. ***Functional success*** was defined as the presence of regular sexual intercourse.

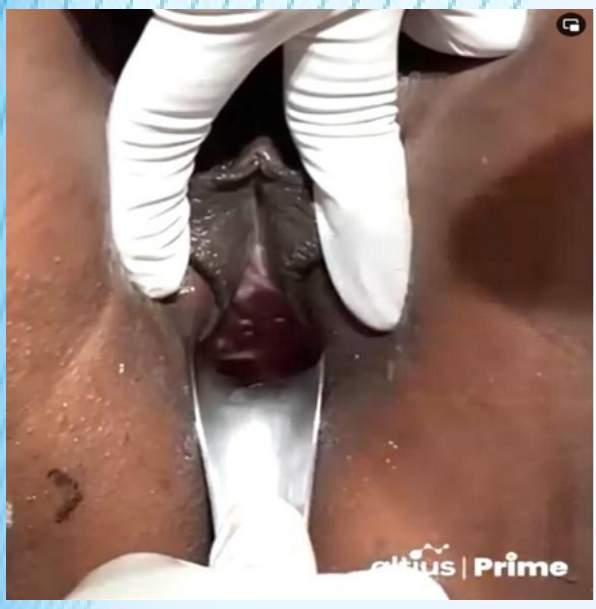


Figure 31- Post-op day 10, vaginal speculum shows a healthy, patent vaginal canal.

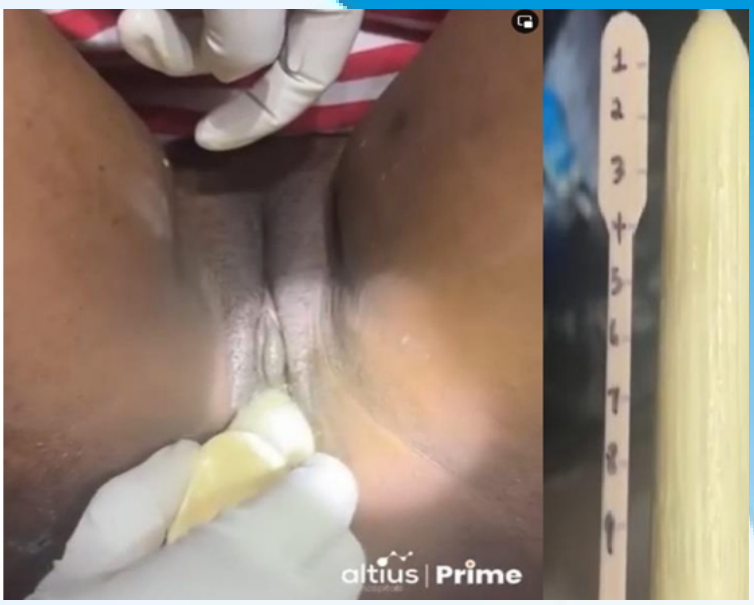


Figure 32- Vaginal mold removed and measured – showing a length of 8 cm.

Fertility is mostly attained by in-vitro fertilization. Some surgeons prefer to reposition the ovaries at the vaginal vault for ease of ovum pick up – although it is controversial owing to the difficulty in moving the ovaries and hampering of the blood supply to the ovaries in the process. Ovum pick up may also be done at the time of vaginoplasty.

More recently, vaginoplasties using cultured autologous vulvar tissue and tissue-engineered biomaterial have been suggested(6).

Compared with primary vaginal dilation, vaginoplasty complications are much more common and include bladder or rectal perforation, graft necrosis, hair-bearing vaginal skin, fistulae, diversion colitis, inflammatory bowel disease, and adenocarcinoma – the most common being rectal injury and obliteration of the neo-vagina(2).

ROUTINE GYNAECOLOGICAL PRACTISE

The patient should be asked about any vaginal discharge, bleeding, pelvic pain, or dyspareunia. Care should be taken to avoid questions related to menstrual cycle. Pelvic examinations should be performed if there are concerns about complications, vaginal stricture, or stenosis. If a patient is symptomatic, vaginal speculum examination and inspection should be performed to check for possible malignancy, colitis, ulceration, or other problems. If an abnormal lesion is identified, biopsy is warranted(2).

Sexually active women with mullerian agenesis should be informed about the risks of STI and advised to wear condoms during unprotected intercourse. Screening for STI should be screened according to the guidelines for women without mullerian agenesis. Human papillomavirus vaccination is recommended to decrease the risk of vulvar and vaginal neoplasia and genital warts(2).REFERENCES

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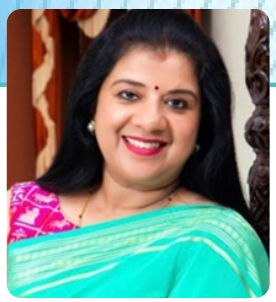
Shaping Solution The Role of Hysteroscopy in Mullerian abnormalities Mangement

Authors: Dr. Vidya.V. Bhat[1] Dr. Mythreyi Kadambi[2]



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Introduction

Congenital uterine anomalies have long been heralded as the pinnacle of infertility surgery. The sheer magnitude of importance that the uterine cavity commands befits the status it holds as the temple of fertility. It is the only part of the female reproductive system which is absolutely crucial for conception and progressing to a favourable pregnancy outcome.

The ASRM Müllerian Anomalies classification 2021 (MAC2021) classifies müllerian anomalies into the nine categories below. Because müllerian anomalies represent a continuum of development and many have combined elements, some anomalies appear in more than one category. This is particularly true for the vaginal anomalies, since they are often seen in combination with uterine and cervical anomalies[1]

1. Müllerian agenesis
2. Cervical agenesis
3. Unicornuate uterus
4. Uterus didelphys
5. Bicornuate uterus
6. Septate uterus
7. Longitudinal vaginal septum
8. Transverse vaginal septum
9. Complex anomalies

Septate uterus is an uterine anomaly where an intrauterine septum divides the uterine cavity either partially or completely. This septum may extend from the fundus to the cervix, resulting in a structural abnormality that can impact fertility and pregnancy outcomes.

The septate uterus is the most commonly encountered Müllerian anomaly. ASRM defines a septate uterus with an indentation depth of >15 mm and an indentation angle of <90°. In contrast, The European Society of Human Reproduction and Embryology (ESHRE) defines a septate uterus using indentation-to-wall-thickness (I:WT) >50%, and the Congenital Uterine Malformation by Experts (CUME) suggest using an indentation depth of ≥1 cm [2,3]

ASRM MÜLLERIAN ANOMALIES CLASSIFICATION 2021

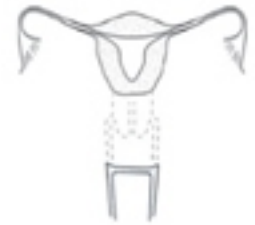
Scan QR code to view the ASRM MAC 2021 tool (page 1 of 2)

MÜLLERIAN AGENESIS

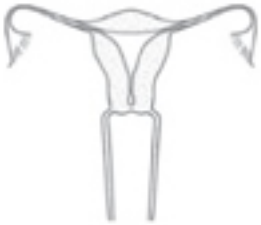
MÜLLERIAN AGENESIS

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

UTERUS DIDELPHYS

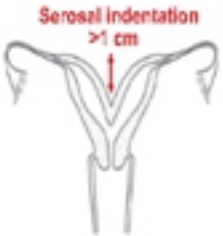
UTERUS DIDEPHYS AND LONGITUDINAL SEPTUM

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

UTERUS DIDELPHYS AND OBSTRUCTED R/L HEMIVAGINA

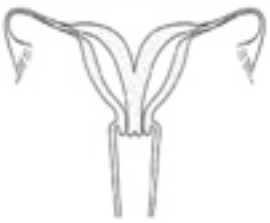
R/L UNICORNUATE WITH R/L UTERINE HORN COMMUNICATING AT LEVEL OF CERVIX

BICORNUATE UTERUS

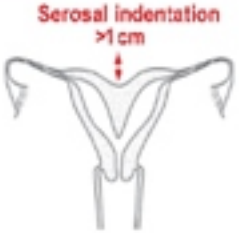


BICORNUATE UTERUS

BICORNUATE UTERUS WITH R/L COMMUNICATING TRACT



UTERUS BICORNUATE BICOLLIS



COMBINED BICORNUATE SEPTATE UTERUS

Etiology: [4]

The etiology of a septate uterus is primarily congenital, meaning it arises during fetal development. It occurs due to incomplete fusion or resorption of the Müllerian ducts during embryogenesis, typically between the 8th and 12th weeks of gestation. Normally, the Müllerian ducts fuse to form the uterus, fallopian tubes, and upper part of the vagina. However, if this fusion process is incomplete or interrupted, it can result in various uterine anomalies, including a septate uterus.

Several factors may contribute to the development of a septate uterus, including genetic predisposition, environmental factors, and hormonal influences during fetal development. However, in many cases, the exact cause of septate uterus remains unknown. It's considered a sporadic congenital anomaly rather than a hereditary condition, although there may be some familial clustering or genetic predisposition in some cases.

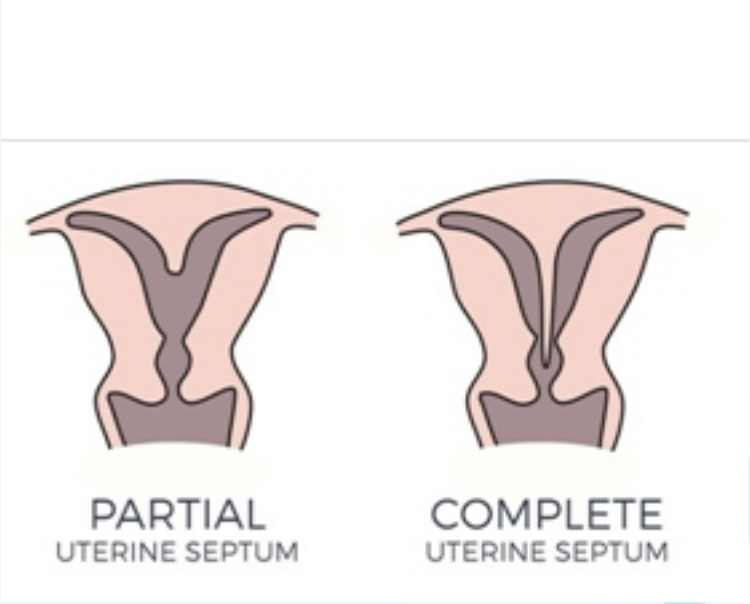
Epidemiology:

Uterine septum accounts for 35% of the uterine anomalies and is associated with the highest rate of adverse reproductive outcomes

The prevalence is estimated to be up to 1.5% in reproductive-age females. However, it is difficult to account for the prevalence of septate uteruses accurately; there are multiple accepted definitions, and many patients with the anomaly are asymptomatic.[5]

Types of septate uterus:

Complete Septate Uterus



In a complete septate uterus, the septum extends fully from the top of the uterus (fundus) to the cervix, dividing the uterine cavity into two separate compartments.

The septum is typically thick and well-defined, completely separating the uterine cavity into two distinct cavities of equal or near-equal size.

This type of septate uterus is associated with a higher risk of fertility issues, recurrent miscarriages, and pregnancy complications due to the complete division of the uterine cavity.

Partial Septate Uterus:

In a partial septate uterus, the septum only partially divides the uterine cavity, resulting in a larger, dominant cavity and a smaller, less developed cavity.
The septum may extend partially from the fundus towards the cervix, but it does not reach all the way down to completely separate the cavity.

This type of septate uterus may vary in severity, with the septum ranging from thin and membranous to thicker and more fibrous.

While the partial septate uterus still poses risks for fertility and pregnancy complications, the degree of septal involvement may influence the severity of associated issues

Due to the timing of embryonic development, Müllerian anomalies are strongly associated with abnormalities of the upper and lower urinary tract; coexistence occurs in up to 40% of patients. Although the concurrent incidence of uterine septa and renal anomalies lacks specific data, it is believed that renal anomalies are less frequently observed in cases involving impaired resorption rather than impaired fusion of the paramesonephric ducts. It is important to note that patients with isolated uterine septa do not require renal imaging unless otherwise indicated.[6,7]

Clinical Presentation:

Septum tissues have fewer blood vessels and a relatively high fiber content, and the endometrium covering the septum shows a relatively poor response to hormones, affecting fertilized egg implantation as well as normal growth and development of the placenta, which may lead to infertility, miscarriage, premature birth, abnormal fetal position, and so on. Among all uterine malformations, uterine septum is the only one that can be treated and corrected by hysteroscopic surgery[8]

Recurrent Miscarriages: Women with a septate uterus may experience recurrent miscarriages, typically in the first trimester. The presence of the septum can interfere with embryo implantation or proper placental development, leading to pregnancy loss.
Infertility: Septate uterus can contribute to infertility by affecting embryo implantation or disrupting the normal uterine environment necessary for successful pregnancy. Women with a septate uterus may have difficulty conceiving or maintaining a pregnancy.
Preterm Labor: Septate uterus is associated with an increased risk of preterm labor and delivery. The abnormal uterine structure can lead to reduced uterine capacity, abnormal fetal positioning, or placental abnormalities, predisposing women to preterm birth.

Abnormal Uterine Bleeding: Some women with a septate uterus may experience abnormal uterine bleeding, including heavy or prolonged menstrual periods, irregular menstrual cycles, or intermenstrual bleeding. The presence of the septum can disrupt normal uterine contractions and blood flow, leading to menstrual irregularities.
Pelvic Pain: In some cases, women with a septate uterus may experience pelvic pain, discomfort, or pressure. This may be related to the abnormal uterine structure causing mechanical issues or inflammation within the uterine cavity.

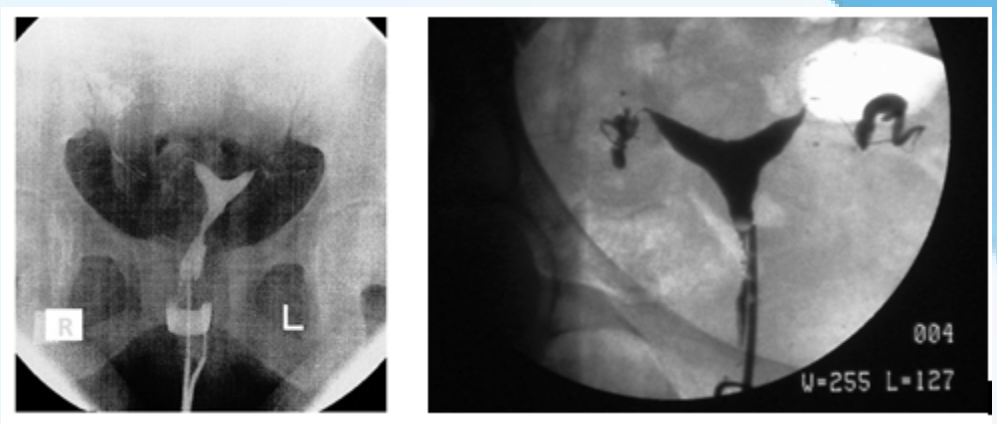
It's important to note that not all women with a septate uterus will experience symptoms, and the clinical presentation can vary widely among individuals. Additionally, the severity of symptoms may be influenced by factors such as the size and thickness of the septum, as well as any associated reproductive or medical conditions

Evaluation:

The septate uterus may be visualized either directly during a diagnostic or therapeutic hysteroscopy or laparoscopy or indirectly via imaging studies such as ultrasonography (USG), hysterosalpingography (HSG), or magnetic resonance imaging (MRI).

Each option has its own advantages and should be addressed separately:

Hysterosalpingography (HSG)

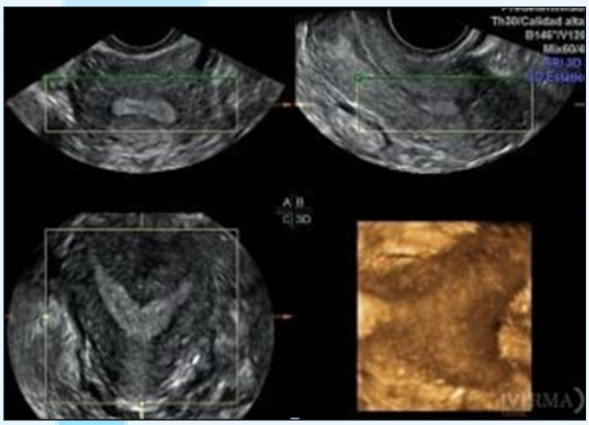


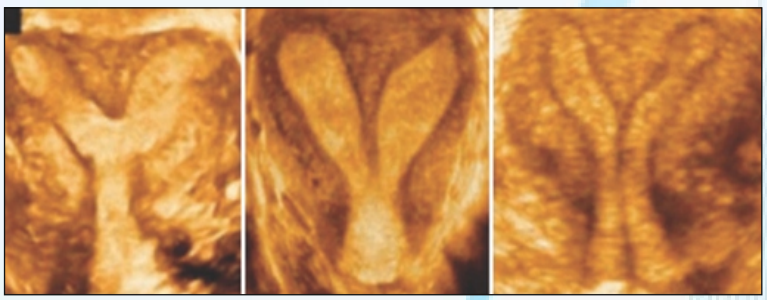
HSG images showing uterine septum

HSG is the radiologic imaging technique that facilitates structural evaluation of the endometrial cavity and fallopian tubes. Either oil- or water-based radiopaque contrast is delivered into the endometrial cavity under fluoroscopic guidance, and radiographs are taken. A uterine septum may appear as an inverted triangular piece of tissue dividing the uterine cavity. However, the diagnostic accuracy of HSG in a septate uterus is low due to the difficulty distinguishing this finding from a bicornuate uterus. Additionally, if the HSG reveals findings consistent with a septate uterus, this must be confirmed with 3D ultrasound, saline-infused sonogram, MRI, or laparoscopy to visualize the fundal contour and rule out a bicornuate uterus.[9]

Ultrasonography

2D and 3D ultrasound have seen revolutionary advances recently and have become the gold standard in diagnosis uterine anomalies. Delineation of outer uterine contour as well as cavitary contour can accurately differentiate between all uterine anomalies.

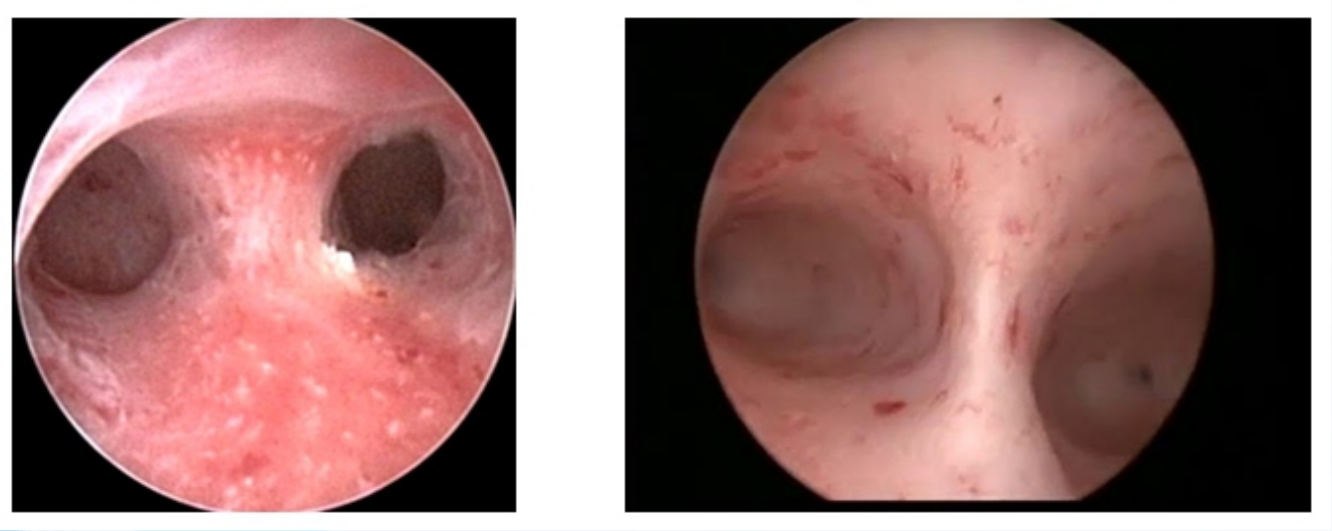




3D ultrasound images, showing partial uterine septum, complete uterine septum and a combine cervico-uterine septum

A saline-infusion sonohystogram combines real-time uterine with the injection of sterile saline into the endometrial cavity to visualize its contour. To measure a uterine septum during sonography, one line is drawn from cornu to cornu (line 1), and a second perpendicular line is drawn from line 1 to the tip of the septum (line 2). The length of line 2 is the septum measurement. In one study, sonohysterography was more sensitive and specific for a septate uterus than HSG and diagnostic hysteroscopy. There was no significant difference between HSG and diagnostic hysteroscopy.[10]

Hysteroscopy



Office hysteroscopy clearly visualising duplication of the uterine cavity, divided by a septum

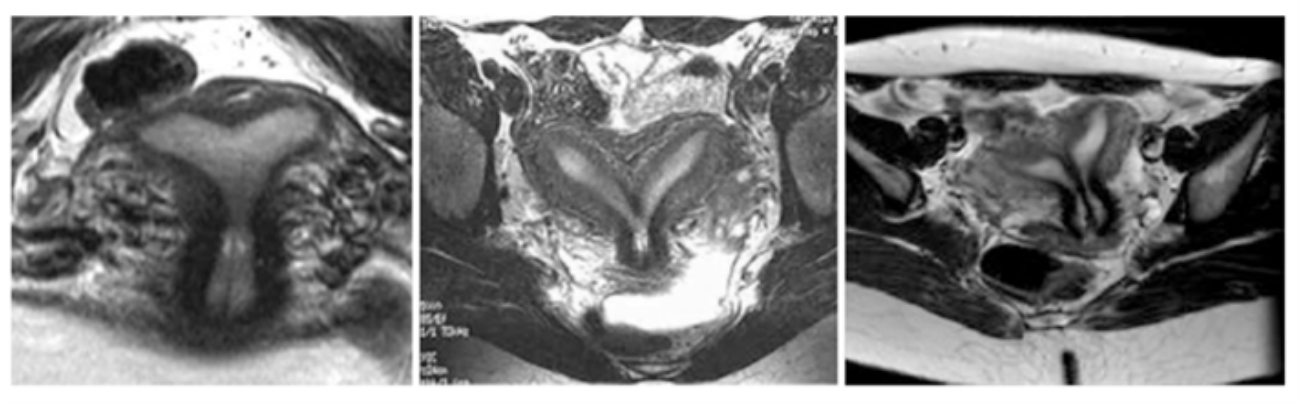
Hysteroscopy is the insertion of a rigid or flexible hysteroscope through the cervical canal into the endometrial cavity. Visualization of the entire endometrial cavity is accomplished using distention media. Hysteroscopy provides a complete view of the contour of the endometrial cavity, both tubal ostia; uterine septa are directly visualized if present. However, hysteroscopy does not allow visualization of the contour of the serosal surface of the uterine fundus; a second imaging modality, such as concurrent ultrasound or laparoscopy, is needed to rule out a more significant uterine anomaly.

Laparoscopy

Laparoscopy is a surgical procedure performed under general anesthesia that permits direct visualization of intraperitoneal abdominopelvic structures. Laparoscopy can be used in conjunction with hysteroscopy and HSG to visualize the fundal contour and identify the class of Mullerian anomaly. The method of using hysteroscopy and laparoscopy together has been shown to have the same diagnostic yield for a uterine septum as 3D ultrasound. Thus, 3D ultrasound is the preferable modality given the less invasive approach.

Magnetic Resonance Imaging (MRI)

MRI of the pelvis using T2 images in three planes can measure a uterine septum. Long- and short-axis views are prescribed relative to the endometrium in the sagittal plane. The long-axis view allows visualization of the uterine fundal contour, while the short-axis view allows measurement of the uterine septum. [11]



MRI images showing a partial uterine septum,
complete uterine septum and a combine cervico-uterine septum

Management:

Up until now, no consensus has been reached on the management of septate uteri, and it is still a great debate among the scientific community

The management of a septate uterus depends on various factors including the severity of symptoms, reproductive goals, and patient preferences. Here's an overview of the management options:

Observation:

Asymptomatic patients may opt for conservative management with regular monitoring and no intervention, particularly if they have no immediate fertility concerns or pregnancy complications.

Hormonal Therapy:

Hormonal medications, such as oral contraceptives or gonadotropin-releasing hormone (GnRH) agonists, may be prescribed to regulate menstrual cycles and alleviate symptoms such as abnormal uterine bleeding or pelvic pain.

Assisted Reproductive Technologies (ART):

For women with fertility issues associated with a septate uterus, ART procedures such as in vitro fertilization (IVF) may be recommended. IVF bypasses the uterine cavity, allowing for embryo transfer directly into the uterus, potentially overcoming implantation difficulties associated with the septum.

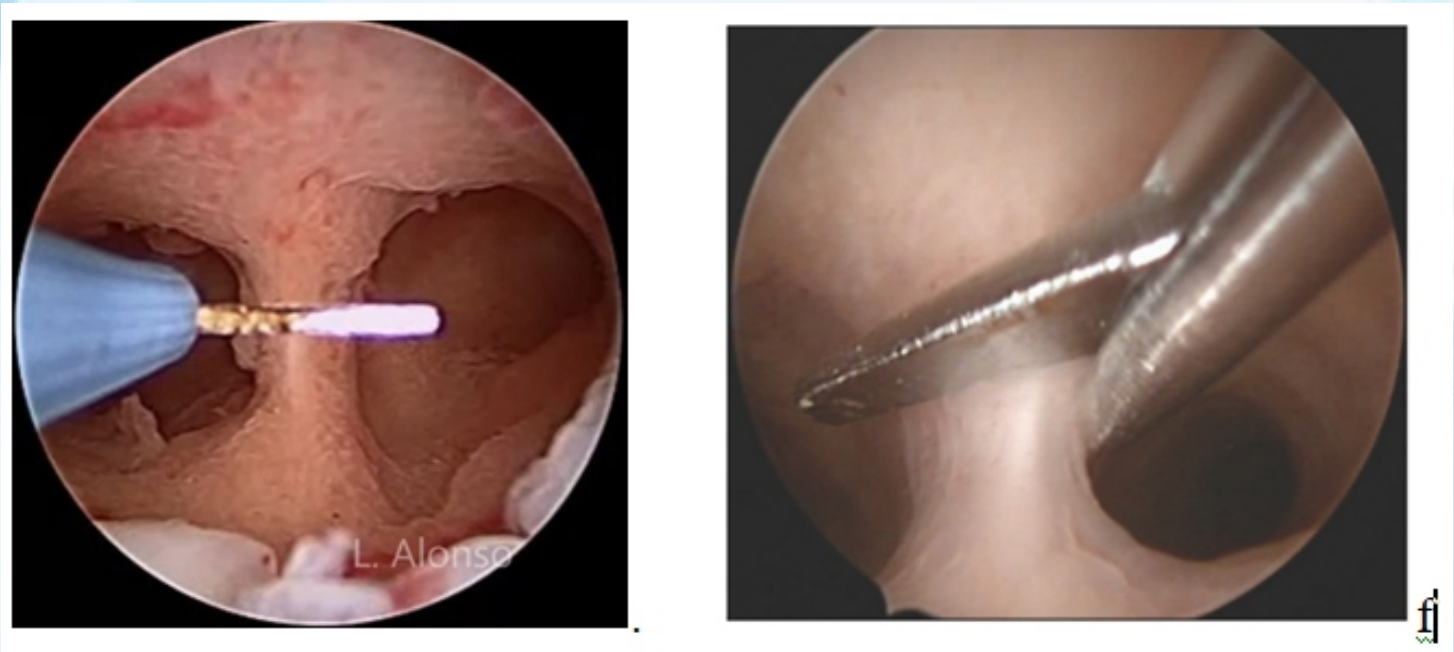
Hysteroscopic Septal Resection:

Surgical correction through hysteroscopic septum resection is the preferred treatment for symptomatic septate uterus, particularly in cases of recurrent miscarriages, infertility, or significant uterine anomalies. During this minimally invasive procedure, a hysteroscope is inserted through the cervix to visualize the uterine cavity, and the septum is excised using specialized instruments. This procedure aims to restore the normal uterine anatomy and improve reproductive outcomes.[12,13]

The new consensus is that septal tissue is incised and released rather than excised. The division leads to resumption of the septal tissue and incorporation into the anterior and posterior myometrium. Dividing the septum exactly in the mid-coronal plane will lead to tissue retraction and sub- sequently perfect healing.

Cold hysteroscopic scissors and electrocautery on a Collin's knife electrode are some of the most commonly used in septal surgery although other options exist: laser and morcellator. [14] There is no distinct advantage in terms of results between both options and the surgeon's preference and judgement is important in each specific case. The scissors are advantageous in an office setting where the septum can be corrected without anesthesia and utilizing a smaller diameter hysteroscope.

The electrocautery is superior in its haemostatic ability which makes it better suited in cases of very vascular septa. It is to be noted that the larger diameter of the resectoscope will usually require cervical dilatation till a corresponding size of Hegar dilators. It is very important to avoid traumatising the septum during dilatation, this can be ensured by passing the dilator

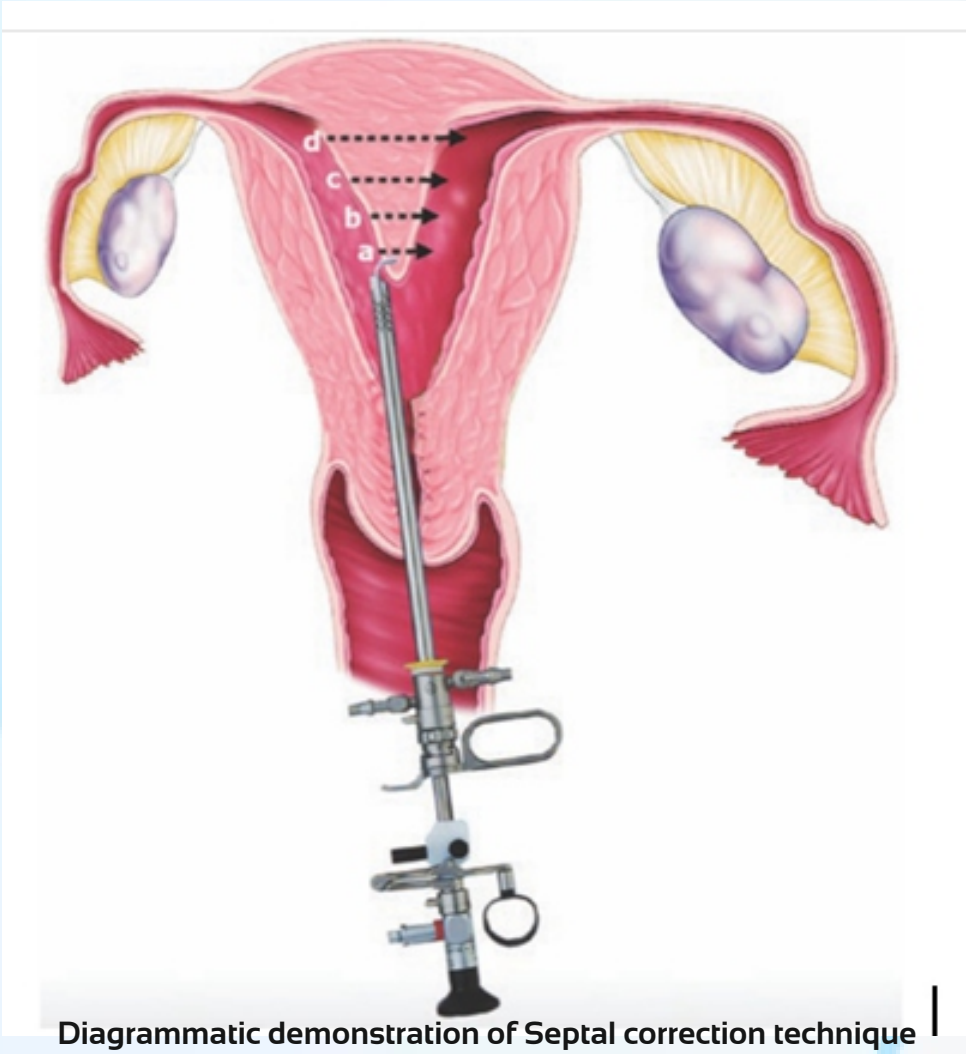


Septal resection using Collin's knife and cold scissors

Once the procedure has begun, division begins caudally and extends cephalic towards the uterine fundus. It is always important to use the tubal ostia as markers of the mid-septal plane to avoid losing your orientation.

The correct plane of incision using hysteroscopic scissors is shown in the figure.
During resectoscopy, The Collin's knife electrode should be utilised from a side to side position rather than the traditional pushing technique usually employed. Beginning at 3 o' clock to 9 o' clock position or vice versa.

The side-to-side movement minimises any change in anteroposterior level and allows more control of cutting for the surgeon. It also carries the benefit of lowering the risk of perforation associated with the more commonly used pushing technique.



Diagrammatic demonstration of Septal correction technique

To judge when the procedure is complete, both tubal ostea must lie on one plane. Some authors suggest that the appearance of excessive vascularity is the end point as this denotes the myometrium; however, this claim is refuted after the notion of an avascular septum was proven to be false.

.

Postoperative Monitoring:[15]

A variety of options exist to prevent the formation of adhesions following hysteroscopic metroplasty. A postoperative regimen for hormonal treatment is the use of 2 mg estrogen per day for 25 days, then Provera is added in the last 5 days of the treatment. This acts by stimulating endometrial growth and is probably the commonest adjuvant therapy used following metroplasty despite the fact that different studies concluded that hormonal therapy is unnecessary

The same has been proven regarding IUCD insertion following metroplasty .

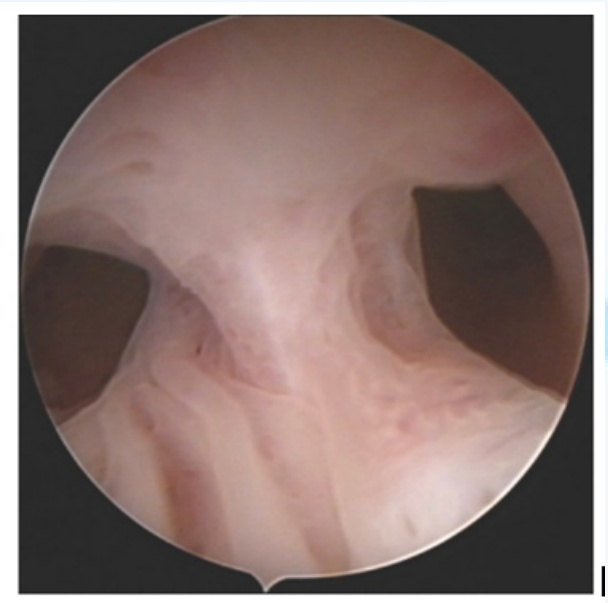
Second Look Office Hysteroscopy:

We recommend a second look hysteroscopy in an office setting after three cycles to ensure complete septum incision, check healing of the raw surface area and to release any possible adhesions or reformation of part of the sep- tum, which usually occurs at the fundus.[16] 2D/3D ultrasound and hysterosalpingography may be used as an alternative showever, the use of hysteroscopy is more preferable as it allows management of any adhesions or residual septum.

Special Considerations

Cervico-uterineSeptum

Special attention must be given to combined cervical uterine septa . Due to the caudal extension of the septum, the procedure is started with the entirety of the hysteroscopy located in the cervix. This makes manoeuvrability very difficult and will limit visualisation. It may be impossible to dilate the cervix in this situation for a large diameter resectoscopy and alternatively, hysteroscopic scissors may be used.[17]



Cervical Septum, identifiable by its endocervical mucosa

Vaginal Septum

A longitudinal vaginal septum may be non-obstructing or obstructing if it is fused to the lateral vaginal wall. The former usually results in dyspareunia whilst the latter causes cryptomenorrhoea and pelvic pain which usually presents after menarche.

The obstructed semi vagina is commonly associate with a bicornuate bicollis uterus, termed Herlyn – Werner – Wunderlich syndrome (also associated with ipsilateral renal agenesis). The correction of vaginal septum used to entail excision and suturing of the vagina. In the obstructive group, this usually meant performing a hymenotomy as well.

Hysteroscopy provides a unique alternative, where the hymen can be preserved and the correction does not require suturing as the tissue retraction leads to no excessive tissue as seen in the second look hysteroscopy, showing the healing of the vaginal septum along the anterior and posterior vaginal walls [18]

Overall, the management of septate uterus involves a personalized approach tailored to the individual patient's symptoms, reproductive goals, and medical history. Collaboration between patients and healthcare providers is essential in making informed decisions regarding treatment options and optimizing reproductive outcomes.

Complications of the Procedure

Hysteroscopic metroplasty is generally considered a safe procedure of short duration that rarely can be associated with complications as compared to the previous more morbid surgical approaches which required several extra days hospital stay, an abdominal incision and resultant scar, development of pelvic adhesions which faltered future fertility chances, Intrauterine adhesions as a result of the reduction metro- plasty which can affect implantation and progression to a term pregnancy and also the detrimental side effects of hav- ing a uterine incision which could lead to uterine rupture during pregnancy and increase the need for c-section delivery. As for any hysteroscopic procedure, general complications of hysteroscopy can occur. This includes cervical laceration from vigorous cervical dilatation or trauma by the hysteroscope, haemorrhage and spread of infection in cases of active vaginal and cervical infection.

Role of cervical cerclge after septal resection:

Ultrasound cervical assessment is useful tool to select patients for therapeutical cerclage.Cervical cerclage is warranted only in cases of cervical changes diagnosed by transvaginal sonography.

Conclusion

Uterine septum is a relatively common anomaly; it is the most common of all female genital anomalies. Surgeons must be prepared to diagnose it and provide the patient with a safe and efficient means of treatment. Hysteroscopic metroplasty is without doubt the gold standard for treatment and should be employed without hesitation.The complication rate is thankfully low, and therefore, it is advisable for any patient diagnosed with a uterine septum to be counseled for correction regardless of previous obstetric histo

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'Distinct Paths: Optimal Management of Unicornuate , Bicornuate, and Didelphys Uterine Anomalies'

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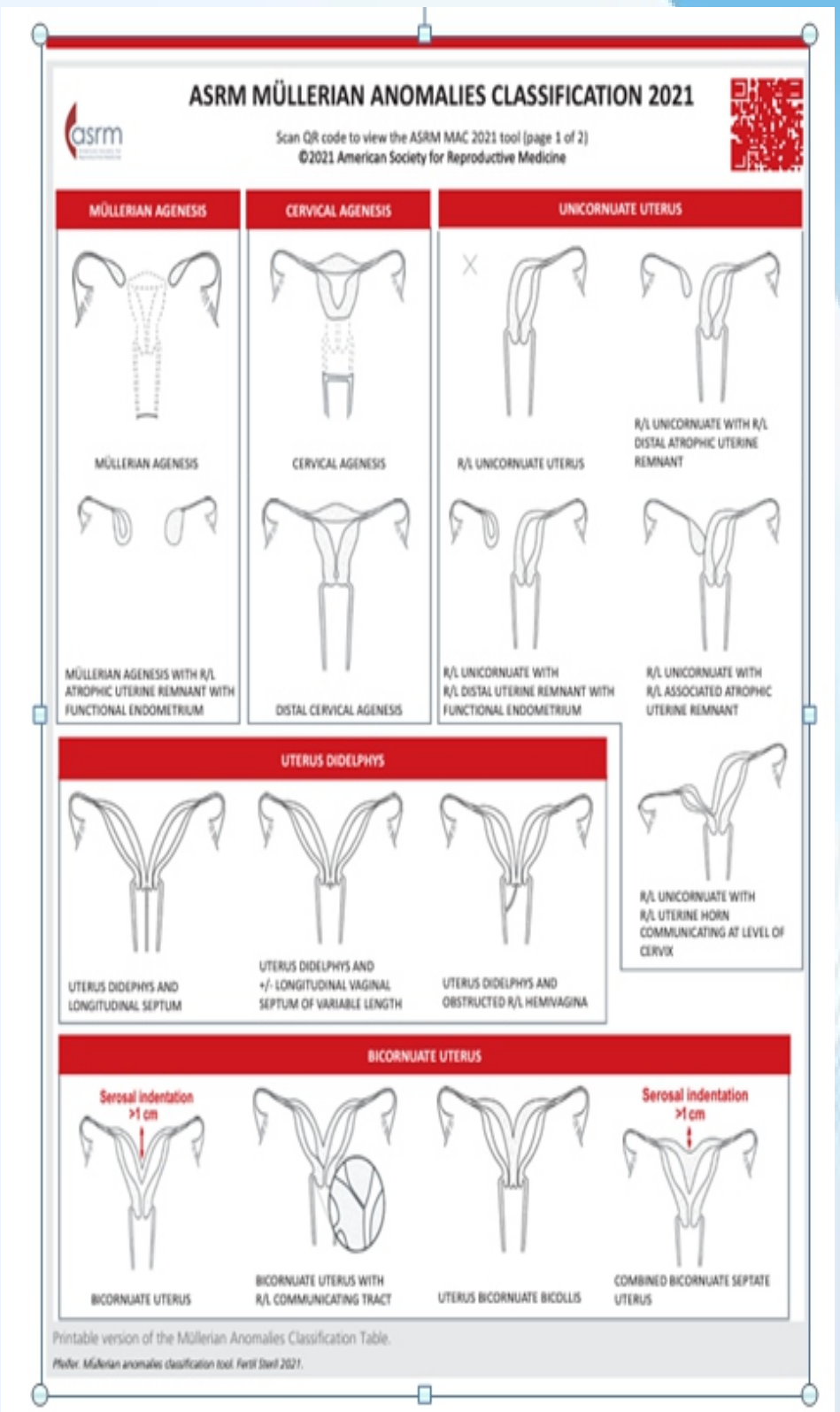
Introduction:

Mullerian anomalies are rare developmental anomalies of the female reproductive tract. The ASRM Mullerian Anomalies Classification 2021 (MAC2021) classifies Mullerian anomalies into the nine categories below. Because Mullerian anomalies represent a continuum of development and many have combined elements, some anomalies appear in more than one category. This is particularly true for vaginal anomalies, since they are often seen in combination with uterine and cervical anomalies. Depending on how the anomaly presents, the provider may perceive the anomaly as primarily vaginal or uterine; this classification allows for this flexibility.



Management of Uniconuate, Bicornuate and Didelphys uterine anomalies.

Unicornuate uterus:

Unicornuate uterus is not a unique anomaly, and its prevalence is estimated close to 0.1%, 0.5%, and 2% in unselected, and infertile women, and those with a history of miscarriage, respectively. Rarely (1 of 35 cases of unicornuate uterus), it may be a case of horn obstruction with hematometra, which may require the decision about surgical intervention or temporary expectant management. The recognition of obstructive Müllerian anomaly is made in adolescence due to the blood retention and pain. However, if the pain is mild and associated with menstruation the diagnosis may be delayed and associated with the long-term impact of retrograde menstruation. Independent of the intensified pain during the menses, it may cause the development of endometriosis and pelvic adhesions with its subsequent consequences. The choice of treatment options in all cases of suspicion of a rudimentary uterine horn with obstruction needs attention considering the proper diagnosis. It may not be a unicornuate uterus, but morphologically and symptomatically similar anomalies such as Robert's uterus or bicornuate uterus with obstruction of the hemicavity. The treatment of these anomalies may be different. The application of two-dimensional ultrasound and magnetic resonance imaging in the diagnosis of a rudimentary horn is a basic tool. This is true for physicians with limited access to three-dimensional ultrasound. Also, laparoscopic amputation of a noncommunicating functional rudimentary horn with hematometra is a typical approach to this type of anomaly.

Main goal in the management of obstructive anomalies of the female genital tract is to provide 1. Pain relief

2. Avoid sequelae
3. Select the best option(s) for reproductive function.

With the goal to minimize damage of the uterus and genital tract, modern management provides a few options including a hysteroscopic approach to obstructive anomalies with the creation of a communication between previously noncommunicating parts or laparoscopic /robotic/laparotomic connection of divided parts of the uterus. Amputation of obstructed horns is irreversible and fails to achieve the goal of reproductive function, but may be the only option for some women.

In cases of a unicornute uterus with hematometra in the rudimentary horn, a laparoscopic horn resection is fast, efficient, and technically easy in the hands of low-volume surgeons where the rudimentary horn is flimsy in connection with the main body of the unicornuate uterus. Resection needs only simple coagulation and incision of the tissue between the horn and the uterine body and coagulation of small blood vessels, which supply the horn. A unilateral salpingectomy is suggested to prevent a potential of an ectopic pregnancy.

In cases where the rudimentary horn may be solidly attached to the unicornuate uterus a laparoscopic technique can be more challenging. For safety reasons, this requires identification of the ureter. With concomitant endometriosis and adhesions, transperitoneal identification of the ureter can be a challenge, making a retroperitoneal approach essential. Suturing of the uterine wall may be necessary to restore integrity; bleeding may be more extensive and identification and ligation of the uterine artery may be necessary. In cases with the solid and wide attachment of rudimentary horn to the unicornuate uterus, we suggest considering the use of temporary occlusion of the uterine artery to minimize blood loss during the laparoscopic horn resection similar to that done during a myomectomy.

A very promising nonextirpative, yet restorative option for selected cases of unicornuate uterus with a functional noncommunicating rudimentary horn is metroplasty with unification into a single uterine body with an enlarged single uterine cavity. This approach is technically similar to the Strassman procedure for the bicornuate uterus, which is feasible by traditional or robotic-assisted laparoscopy. Although reproductive and safety outcome data are lacking, an uncomplicated twin pregnancy after such treatment has been described.

Bicornuate uterus:

A bicornuate uterus is a uterine malformation that is produced due to impairment in the fusion of Mullerian ducts. The bicornuate uterus is a rare anomaly, but it is associated with worse reproductive outcomes; recurrent pregnancy loss and preterm labor are most common. To avoid the obstetric complications related to this anomaly, a high suspicion should be maintained for adolescents presenting with menstrual complaints. Diagnosis should be followed by aggressive prenatal monitoring or surgical unification depending on an individual basis. This activity reviews the evaluation and management of bicornuate uterus and highlights the role of the interprofessional team in evaluating and improving the care of patients with this condition.

If a woman presents for a routine evaluation during her pregnancy gets diagnosed with a bicornuate uterus, then aggressive prenatal monitoring is indicated to prevent obstetric complications. Pay attention to the signs of preterm labor, malpresentation.

A patient can additionally present with a history of recurrent abortions or preterm labor in preceding pregnancies. The presentation mentioned above is an indication for the surgical unification of uterus, Strassman metroplasty. The procedure was first illustrated in 1907 by Strassman in 4 stages. The procedure initiates by making a transverse incision over the fundus of the uterus, staying away from uterotubal junctions to avoid injury. Subsequently, the uterine cavity is opened, and the septum is removed after splitting the partition. Thus the procedure transforms the double cavity into a single cavity. Ultimately the cavity is closed by vertical suturing to prevent endometrial adhesions.

The laparoscopic approach is being preferred in the present in place of abdominal metroplasty. The laparoscopic approach provides leverage in terms of less bleeding and decreased rate of infections. It also reports significantly reduced postoperative adhesion formation, which can be credited to a decrease in tissue handling and drying of tissues.

Didelphys Uterine anomaly:

Uterus didelphys occurs as a result of abnormal fusion of the paramesonephric ducts and is characterized by complete duplication of uterine horns, cervix, and very often also the vagina or presence of longitudinal vaginal septum. Most women with a uterus didelphys are asymptomatic; some cases may coincide with dyspareunia or dysmenorrhea. The anomaly is associated with a higher risk of miscarriage, preterm labor, breech delivery, and decreased live births. Diagnosis of uterine malformations is usually problematic and lengthy, due to the asymptomatic course of uterine anomalies. For diagnosis, the following are used: ultrasound, HSG, and MRI. However, the detection of uterine malformations has increased in the last decade thanks to better diagnostic methods. If there are no coexisting symptoms, treatment is waived. On the other hand, in case of obstetric failure, surgical treatment is possible, involving the fusion of two duplicated uteruses. The most widely used classification of uterine malformations is the American Society for Reproductive Medicine (ASRM) classification (updated and expanded American Fertility Society (AFS) classification), which outlines nine main types of uterine anomalies. According to AFS classification, uterus didelphys belongs to class III. ASRM, in their classification, divided the group of uterus didelphys into three: 1. Uterus didelphys and longitudinal septum, 2. uterus didelphys and +/- longitudinal vaginal septum of variable length, and 3. uterus didelphys and obstructed R/L hemivagina.

Fertility and uterine Anomaly

Uterine malformations are most often detected in patients of reproductive age due to fertility problems (previously no symptoms), as many as 2–8% of infertile women have a uterine malformation and 5–30% have a history of miscarriage. Bicornuate, unicornuate, and didelphic uterus are generally

not a direct cause of infertility. However, it might be associated with aberrant outcomes throughout the course of pregnancy. Uterus didelphys as an isolated anomaly is not an indication for either surgical treatment or termination of pregnancy by cesarean section. The patient's reproductive organ defect consists of a double uterus, but also two cervixes and one vagina, however partitioned by an elongated membrane, which was of particular interest for natural childbirth. The restriction of the size of the vagina and its division into two parts may have represented a risk of complications for the newborn and a prolongation of the second phase of labor.

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"Optimising Fertility Outcomes: Management Aproach For Mullerian Anomaly"

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Introduction

Congenital anomalies of the female reproductive tract may involve the uterus, cervix, fallopian tubes, or vagina. Depending on the specific defect, a woman's obstetric and gynecologic health may be adversely affected. Uterine anomalies are the most common of the Mullerian anomalies, but the true incidence is not known since many women are asymptomatic, and sensitive imaging modalities have only recently become available.

Uterine anomalies are associated with both normal and adverse reproductive outcomes; they occur in approximately 3–4% of fertile and infertile women, 5–10% of women with recurrent early pregnancy loss, and up to 25% of women with late first or second-trimester pregnancy loss or preterm delivery.

Overall, uterine anomalies are associated with difficulty maintaining a pregnancy, and not an impaired ability to conceive .The proper management of infertile women with uterine anomalies is controversial.

Clinical presentation related to fertility in Mullerian anomalies.

Of all Mullerian anomalies, those involving the uterus are most commonly implicated in poor obstetric outcomes. Uterine anomalies are associated with diminished cavity size, insufficient musculature, impaired ability to distend, abnormal myometrial and cervical function, inadequate vascularity, and abnormal endometrial development. These abnormalities of space, vascular supply, and associated local defects contribute to increased rates of recurrent pregnancy loss (RPL; 21–33%), preterm delivery, and malpresentation associated with uterine anomalies. Intrauterine growth restriction is similarly attributed to abnormal vascularization and a smaller uterine cavity. An increased cesarean delivery rate is associated with higher rates of malpresentation and vaginal anomalies such as a longitudinal vaginal septum. Furthermore, pregnancy may occur in an obstructed or rudimentary uterine horn; these conceptions rarely last until term, and mostly end in rupture. Other associated obstetrical complications include cervical incompetence, pregnancy induced hypertension (due to renal abnormalities), and antepartum and postpartum bleeding.

Indications for surgical intervention

Surgical correction of uterine anomalies in asymptomatic women or women with primary infertility is controversial. In general, uterine anomalies do not prevent conception or implantation, and these women can have normal reproductive outcomes. The mean prevalence of mullerian anomalies in infertile women is 3.4% (range 1.0–26.2%), similar to the prevalence in the fertile population, which suggests that these abnormalities have little effect on fecundity. In comparison, the prevalence of uterine anomalies in women with RPL is substantially higher at 12.6%. When women with uterine anomalies undergo IVF, they have similar clinical pregnancy rates compared with women with normal uteri, but experience higher rates of miscarriage and preterm delivery.

Currently, surgery for mullerian anomalies is indicated for women with pelvic pain, endometriosis, obstructive anomalies, and poor obstetric outcomes such as RPL and preterm delivery. Prior to performing surgery, it is important to exclude extra uterine factors which may cause pregnancy loss. The goals of surgery include treatment of pelvic pain, restoration of pelvic anatomy and uterine architecture, and preservation of fertility. Inherent developmental abnormalities, however, such as abnormal myometrium or altered vascularization, may permanently impair uterine function

Unicornuate uterus

During embryogenesis, the failure of one mullerian duct to develop and elongate results in a unicornuate uterus. This asymmetric lateral fusion defect usually results in a functional uterus with a normal cervix and fallopian tube, and varying configurations of abnormal mullerian development on the contralateral side: agenesis, or a rudimentary uterine horn (74%). This rudimentary horn may be noncommunicating (70–90%) or communicating with the unicornuate uterus, and may have no endometrial cavity or some functional endometrium.

Although rudimentary horns are commonly asymptomatic, an obstructed horn with active endometrium can result in cyclic or chronic pelvic pain, endometriosis, or a horn gestation. Furthermore, this anomaly is associated with a high incidence of renal abnormalities (40%), usually ipsilateral to the anomalous side. The unicornuate uterus is associated with higher rates of endometriosis, premature labor and delivery, and malpresentation.

Women with unicornuate uteri have impaired pregnancy outcomes; a compilation of studies reveals a spontaneous abortion rate of 36.5%, a preterm delivery rate of 16.2%, a term delivery rate of 44.6%, and a live birth rate of 54.2%.

Surgical reconstructive procedures (augmentation Metroplasty) have not been shown to improve pregnancy outcomes. Although prophylactic cervical Cerclage has been recommended to improve pregnancy outcomes, women with mullerian anomalies should be managed expectantly with adherence to standard indications for Cerclage placement. Additionally, removal of a functional rudimentary horn is recommended as treatment for pelvic pain and endometriosis, and to prevent conception in an obstructed horn.

Uterine didelphys

The failure of fusion of the two mullerian ducts results in duplication of mullerian structures; a didelphic uterus has two uteri, two endometrial cavities, and two cervices. A longitudinal vaginal septum is present in 75% of cases. An obstructed hemivagina can occur with uterine didelphys, and this constellation of findings is associated with ipsilateral renal agenesis (OHVIRA)

This uterine anomaly is associated with modest reproductive outcomes: spontaneous abortion rate of 32.2%, a preterm birth rate of 28.3%, a term delivery rate of 36.2%, and a live birth rate of 55.9%. In select women with RPL or preterm delivery, uterine reconstruction with the Strassman Metroplasty(open or lap) can be considered however, existing data do not support repair of a didelphic uterus to improve pregnancy outcome as first option and should be used as last resort. The Strassman Metroplasty achieves unification of two endometrial cavities in a divided uterus (bicornuate or didelphys), and is associated with a live birth rate greater than 80%. In contrast, incision of the longitudinal vaginal septum is indicated for an obstructed hemivagina with hematocolpos, dyspareunia, or difficulty with tampon placement.

Bicornuate uterus

The bicornuate uterus occurs due to incomplete fusion of the two mullerian ducts at the level of the fundus, resulting in a single cervix and two endometrial cavities. The degree of separation between the two endometrial cavities can be variable, extending as far as the internal cervical os depending on the length of incomplete mullerian duct fusion. The external uterine contour has an indented fundus, arbitrarily defined as more than 1 cm, and the vagina is generally normal

This anomaly is associated with obstetrical complications including pregnancy loss, preterm labor, and malpresentation. An overall spontaneous abortion rate of 36%, a preterm birth rate of 23%, a term delivery rate of 40.6%, and a live birth rate of 55.2% .Furthermore, the incidence of preterm delivery varies with the degree of cavity separation in partial (29%) and complete (66%) bicornuate uteri.

In the setting of RPL or preterm delivery and a bicornuate uterus, all other etiologies of pregnancy loss must be excluded prior to considering a uterine reunification procedure. The Strassman Metroplasty should be reserved for select women based on poor reproductive outcomes. Furthermore, the bicornuate uterus is associated with a high incidence of cervical incompetence (38%).Although studies have identified improvements in fetal survival rates and decreased pre- term delivery rates with a cervical Cerclage .expectant management and appropriate adherence to standard indications for Cerclage placement are warranted.

Septate uterus

A defect in resorption of the midline septum between the two mullerian ducts results in a fibromuscular uterine septum. The degree of septation is variable; a complete septum extends from the uterine fundus through the cervix, and a partial septum demonstrates resorption of a portion of the caudal aspect of the septum. Despite the endometrial cavity abnormality, the external uterine contour appears normal. A longitudinal vaginal septum is found most frequently with a septate uterus. Endometriosis has been identified in up to 30% of fertile and infertile women with septate uterus.

The septate uterus is considered the most common of the uterine anomalies, occurring in approximately 1% of the fertile population and is associated with the poorest reproductive outcomes. A compilation of studies of partial and complete septate uteri identified a pregnancy loss rate of 44.3%, a preterm delivery rate of 22.4%, a term delivery rate of 33.1%, and a live birth rate of 50.1% ,the timing of pregnancy loss into early (25.5% before 13 weeks) and late (6.2% between 14 and 22 weeks). Compared with the rest of the uterus, septal tissue has been shown to have decreased vascular supply and abnormal overlying endometrium, these findings help to explain the aetiology of pregnancy loss with the septate uterus.

Hysteroscopic Metroplasty has been demonstrated to significantly improve the live birth and reduce miscarriage rates to approximately 80 and 15%, respectively and is recommended when the uterine septum is implicated in RPL, second trimester loss, malpresentation, or preterm delivery .The Hysteroscopic approach is preferred due to its safety, simplicity, and excellent post treatment results .Concomitant laparoscopy enables evaluation of the pelvis and external uterine contour, and guides the extent of septum resection. Traditionally the cervical portion of a complete septum is left intact due to the risk of cervical incompetence, but a recent small randomized study demonstrated that resection of the cervical septum is associated with a less complicated surgical procedure and equivalent reproductive out- comes. Postoperative formation of intrauterine synechiae is rare, and routine use of an intrauterine balloon catheter, estradiol supplementation, or antibiotics have not been shown to be necessary .A follow-up examination should be performed 2 months after the procedure preferably by hysteroscopy.

Prophylactic Hysteroscopic Metroplasty in infertile women or women without a history of adverse reproductive outcomes is a controversial procedure since many women with a septate uterus

can have reasonable pregnancy outcomes, and there is no established causal relationship between a septate uterus and infertility. After Hysteroscopic Metroplasty in women with unexplained infertility, a modest improvement in pregnancy and live birth rates is demonstrated in nonrandomized trials, these rates are significantly higher after Metroplasty in women with RPL, which highlights the difference in fertility between these two populations. Prophylactic Metroplasty, however, may prevent miscarriage or other obstetrical complications, and is recommended in order to optimize pregnancy outcomes in women with prolonged infertility, in women older than 35 years, and in women planning to pursue assisted reproductive technologies.

Arcuate uterus

The arcuate uterus has been variably classified as a normal, bicornuate, or septate uterus. Anatomic hallmarks a slight midline septum with a broad fundus, sometimes with minimal fundal cavity indentation. Of all uterine anomalies obstetrical complications are least common in women with an arcuate uterus. Women with an arcuate uterus have an overall term delivery rate of 78% and a live birth rate of 83%, Compared with women with a normal uterus, women with an arcuate uterus have a higher proportion of second trimester losses and preterm labor .Reconstructive procedures on an arcuate uterus, however, do not improve pregnancy outcomes and should be reserved for those with unexplained infertility, miscarriage & preterm labour.

Type of Anomaly	*Presentation*	*Treatment which can be Offered*
Unicornuate uterus	-Miscarriage -Preterm delivery	-Augmentation Metroplasty -Cervical Encerclage -Excision of rudimentary/Non communicating horn
Bicornuate Uterus	-Miscarriage -Preterm delivery	-Strassman Metroplasty -Cervical Encerclage
Uterine didelphys	Miscarriage -Preterm delivery	-Strassman Metroplasty -Cervical Encerclage

Cervical anomalies

Most cervical abnormalities accompany vaginal or uterine anomalies; they are rarely isolated, Cervical anomalies include agenesis, atresia, abnormal length or width, obstruction, and hypertrophy. Cervical atresia is a rare anomaly; these women can present with primary amenorrhea and cyclic pelvic pain due to hematomata and retrograde menstruation. Successful pregnancies have occurred after utero-vaginal anastomosis for cervical atresia .Surgical correction of obstructive cervical anomalies, however, rarely results in a patent passage and is associated with a high risk of ascending infection; a hysterectomy is often necessary. The ovaries should be preserved, hence pregnancy can be achieved with IVF and a gestational carrier.

Vaginal anomalies

Vaginal anomalies include a transverse vaginal septum (a vertical fusion defect), a longitudinal septum, and an imperforate hymen. Although the transverse septum and imperforate hymen are not associated with other mullerian anomalies, the longitudinal vaginal septum often occurs with uterine anomalies such as a septate or didelphic uterus. Regardless, the vaginal anomalies themselves should not interfere with reproductive outcomes

Vaginal agenesis is an uncommon condition, and most frequently occurs as congenital absence of the vagina with variable uterine development (Mayer-Rokitansky- Kuster-Hauser syndrome). This developmental anomaly occurs due to agenesis or hypoplasia of mullerian duct development. The incidence of this abnormality is one in 5000 female births. These women have a 46XX karyotype, and normal ovaries, ovarian function, female external genitalia, and secondary sexual characteristics, but experience primary amenorrhea This diagnosis must be differentiated from vaginal agenesis, androgen insensitivity, low-lying transverse vaginal septum, and imperforate hymen. Mullerian agenesis is also associated with extra genital anomalies: urologic (25 – 50%) and skeletal (10 – 15%) abnormalities, cardiac defects, auditory deficits, digital anomalies, and cleft palates. To enable sexual intercourse, a neovagina can be created with vaginal dilators or surgery; several successful approaches are available

Women with mullerian agenesis can achieve pregnancy with their own oocytes through IVF and the use of a surrogate gestational carrier. Although Uterine transplant could be considered the ideal solution for the management of infertility and the satisfaction of the reproductive and sexual needs of women with MRKH syndrome, it is still in early days to be considered as a safe mode of management due to the variable possible complications of the procedure itself and for the post operational period. It is still far from incorporating this method into therapeutic protocols targeted for infertility treatment for women with congenital uterus aplasia or agenesis. The future will only answer
if this intervention can be applied on a large scale.

Bicornuate Uterus	-Miscarriage -Preterm delivery	-Strassman Metroplasty -Cervical Encerclage
Uterine didelphys	Miscarriage -Preterm delivery	-Strassman Metroplasty -Cervical Encerclage
Septate uterus	-Miscarriage -Preterm delivery	-Septal Metroplasty
Longitudinal vaginal septum	-No reproductive outcome -Incidental diagnosis	-Septal incision / excision
Cervical /Vaginal Agenesis	-Primary Amenorrhoea -Cyclical abd pain	-Ureterocervical canalization -Vaginoplasty -Surrogacy -Uterine transplant
Mullerian Agenesis(MRKH)	-Primary Amenorrhea	-Vaginoplasty -Surrogacy -Uterine transplant

To Summarise

Conclusion

Although mullerian anomalies are relatively uncommon, they can significantly impact reproductive outcomes. A high index of suspicion is warranted in adolescents and reproductive-age women with pelvic pain, dysmenorrhea, abnormal bleeding, RPL, second trimester pregnancy loss, or preterm delivery. Detailed pelvic imaging with modalities such as HSG, ultrasonography, and MRI is necessary, as well as evaluation of the urinary tract as indicated. When a mullerian anomaly is identified, the woman should be counselled about reproductive prognosis, pregnancy outcomes, and evidence-based management. Many of these women can have normal reproductive outcomes, but intervention is recommended in the event of poor obstetric outcomes.

Reference

Concise Verbatim of Article
Beth W. Rackow, Aydin Arici Reproductive performance of women with mullerian anomalies Curr Opin Obstet Gynecol 2007 19:229 – 237

Understanding Endometriosis Within The Spectrum Of Mullerian Anomalies

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Indroduction

Endometriosis is defined as the presence of endometrial tissue exterior to the uterus such as in the ovary and the peritoneal cavity. One risk factor for endometriosis is obstruction of menstrual outflow therefore, it is possible that obstructive malformations of the female genital tract increase the risk of development of endometriosis. Most patients with endometriosis diagnosed by surgery are aged 25 years old and over. The nationwide cohort study reported that the prevalence of surgically verified endometriosis among young patients (10–19 years old) was 1.4% in 26,301 women . Systematic review showed that 648 out of 1011 (64%) symptomatic adolescents undergoing laparoscopy were found to have endometriosis . However, the etiology and mechanisms of the early onset of endometriosis, especially in adolescence, is not well known. Endometriosis and CUA are frequently detected in the exploration of infertility, because both of these are liable to impair fertility. Endometriosis is associated with obstructive anomalies and nonobstructive malformations, especially those concerning the septate uterus.

Embryology

Defect in organogenesis of the distal Mullerian ducts leads to cervico-vaginal agenesis and defect in lateral fusion of the paired Mullerian ducts lead to complete bicorporeal uterus . The lower uterine segment narrows to terminate in a peritoneal sleeve at a point well above the normal communication with the vaginal apex. Less than 200 cases of cervico-vaginal agenesis have been reported in the literature since 1942 ; out of which only 7 % of the cases had functional endometrium .

Symptomatology and diagnosis:

Patients typically present with hematometra, disabling pelvic pain in the presence of normal secondary sexual characteristics and normal karyotype . Obstruction of the menstrual flow results in development of pelvic endometriosis and adenomyosis .

These anomalies usually appear in adolescence and in girls, with or without menses

Presence of menses	Absence of Menses
• Longitudinal obstructing vaginal septum (obstructed hemivagina) • Unilateral cervical aplasia • Robert's uterus • Obstructed uterine horn • Accessory cavitated uterine mass (ACUM)	• Imperforate hymen • Transverse vaginal septum • Partial vaginal aplasia • Cervical aplasia

Basic good sonological evaluation with 3 D ultrasound plays major role in evaluating these cases. MRI is needed in complicated mullerian anomalies.

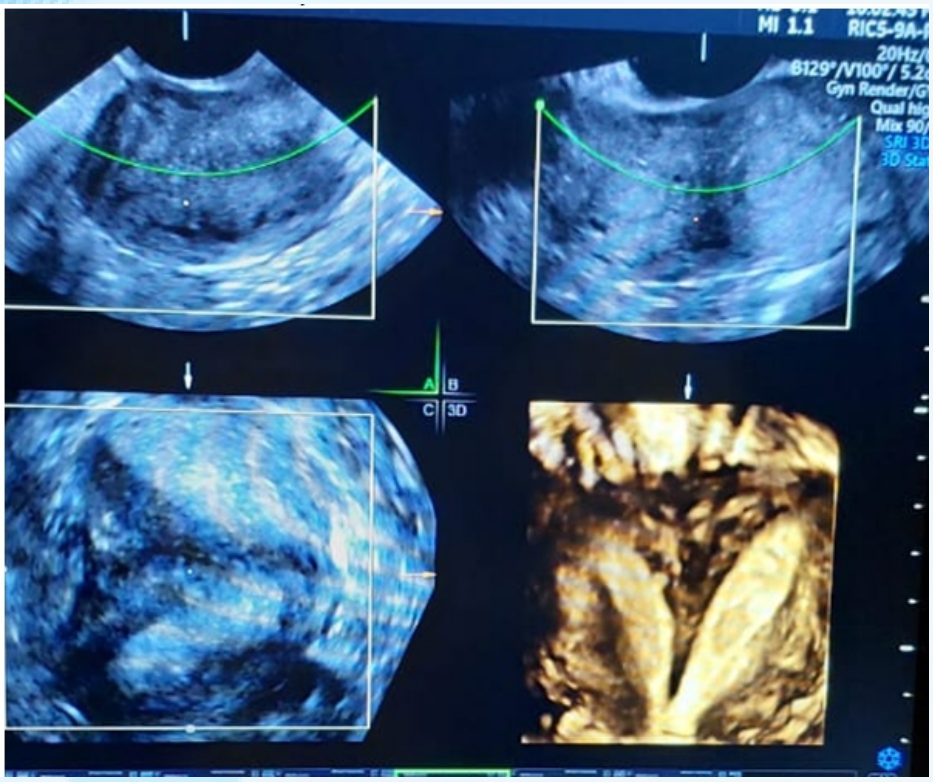

Fig -1 Complete septate uterus on 3D scan

These women may have a shortened blind vaginal pouch as the lower one third of the vagina arises from the urogenital sinus. Thus, blind vagina with abdominopelvic mass in the background of primary amenorrhoea with or without cyclical abdomino-pelvic pain should raise the possibility of either a transverse vaginal septum or vaginal/cervico-vaginal atresia; which cannot be distinguished only on clinical examination.

In these women accurate diagnosis is paramount for effective presurgical planning and preparation. Preoperative 3-D ultrasound or MRI aids in the diagnosis . Nevertheless, surgical exploration confirms the final diagnosis.

These anomalies are closely related to associated malformations of the genitourinary tract in 29 % of the times necessitating renal system evaluation .

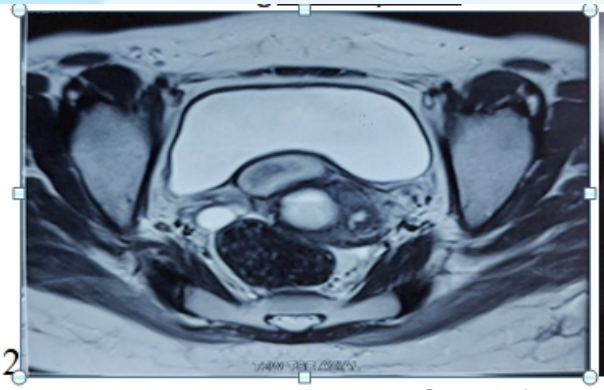

Fig 2

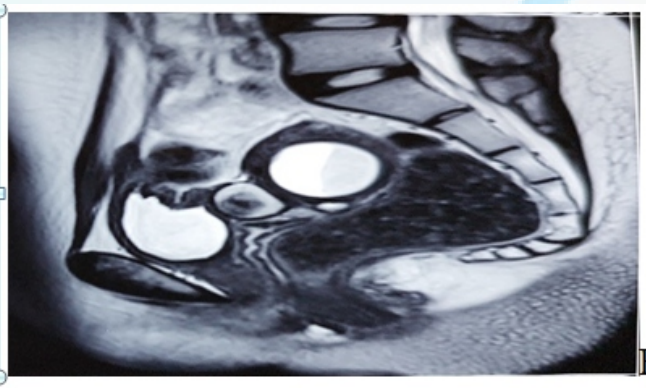

Fig 3

Fig 2 and 3are MRI films of ACUM with JCA in a single patient.

Management:

An effective psychological preparation of the patient is crucial as most of the times these are young adolescent girls who may face difficulty in having a normal sexual and reproductive life and may need corrective surgeries at different stages of their life.

Unlike most of the other mullerian anomalies, there is no well-established evidence-based surgical practice that gives the best outcome to patients with cervico-vaginal atresia. Co-existence of uterine anomalies such as bicorporeal uterus with functioning endometrium further adds to the surgical challenge of creating utero-vaginal anastomosis. Conservative surgical management have been proposed for restoring the continuity of the genital tract such as direct uterovaginal anastomosis/canalisation, creating neocervix using small intestinal submucosal/skin graft/peritoneal flap over Foley's catheter as a plastic stent . The aim of conservative surgeries is to restore menstruation, sexual activity, fertility and allow pregnancy till term. Small number of successful pregnancies following these procedures have been reported in the literature . Although reconstructive surgeries seem ideal, significant complications are known to occur such as sepsis, pelvic inflammatory disease, chronic pelvic pain, bowel and bladder injury, re-obstruction and stenosis . Successful laparoscopically assisted uterovaginal/vestibular anastomosis in patients with cervical atresia associated with partial or complete vaginal agenesis has been reported by Fedele et al. . However, anastomosis was done by the authors only when normal uterine morphology was found intra-operatively and hematometra was excluded. Kang et al. reported 4 cases of postoperative pelvic abscess developing after a mean interval of 67.7 months after primary canalisation among 54 patients who underwent fertility sparing surgeries for cervico-vaginal atresia . These women eventually required hysterectomy and pelvic abscess removal. Literature reports nearly 33 % of women undergoing conservative surgeries require to undergo hysterectomy owing to one or the other complications . Two cases of death following uterovaginal anastomosis due to peritonitis and septic shock at 6 days and 7 weeks after canalization have been reported .

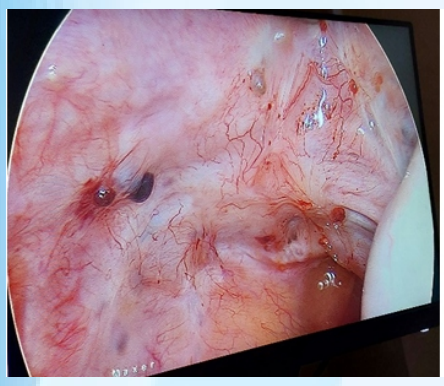

Fig 4Endometriosis in septate uterus

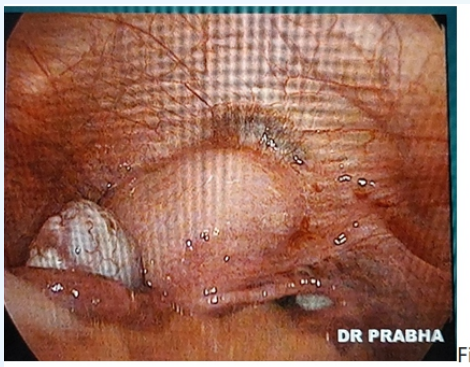

Fig5.

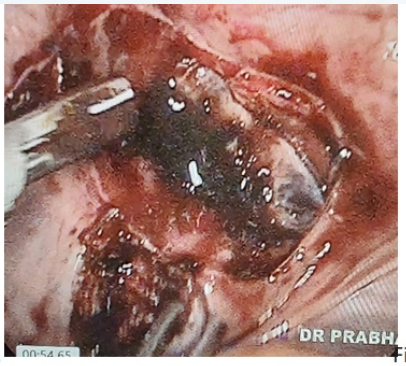

Fig 6

Fig 5 & 6 are lap view of ACUM and JCA

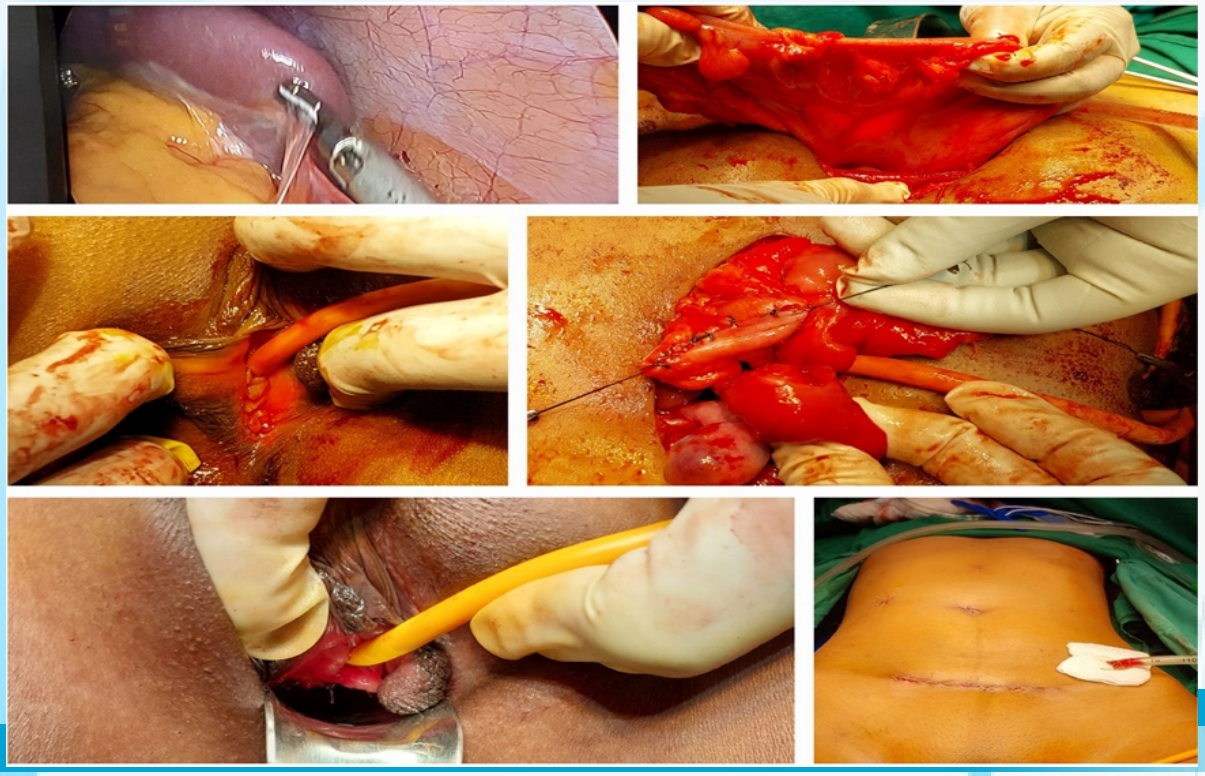

Fig 7 – Sigmoid cervico- vaginoplasty in cervicovaginal atresia withs single uterine horn at LYDM Sparsh Hospital

Conclusion

Early diagnosis of Mullerian anomalies can improve a woman's quality of life. Conservative surgeries should not increase future morbidity for a patient and should not let them undergo repeat surgeries for complications. Thus, it is vital to tailor the management for each patient according to the diagnosis, intraoperative findings, and associated pathologies. Discussion with other colleagues pre-operatively may help in taking a proper decision for patient's benefit. Assisted reproduction can help these women to have their own offspring where conservative surgery is not an option.

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"Decisive Dilemmas: Treating Operable And Inoperable Female Reproductive Tract Abnormalities"

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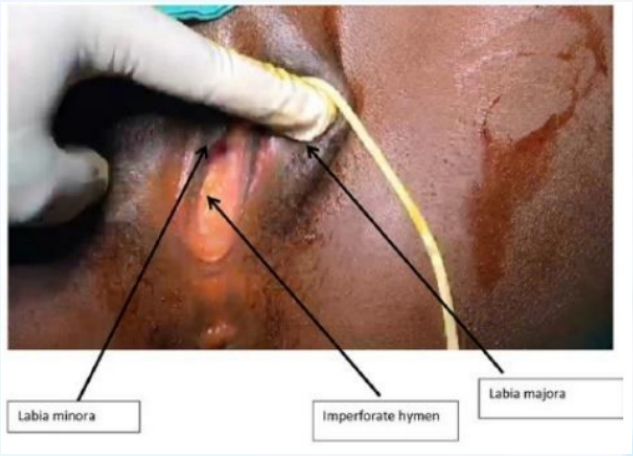
INTRODUCTION

Malformations of reproductive tract result from mal-development or developmental arrest at a critical stage of embryonic development but may also result from genetic mutations or environmental insults during the period of organogenesis.

Here we are going to discuss regarding treating operable and inoperable reproductive tract anomalies.

Surgical management depends on the type of anomaly, it's complexity, patient's symptoms and the proper embryological interpretation of anomaly. Most malformations can be resolved vaginally or by hysteroscopy, but laparoscopy or laparotomy is often needed. If there are fertility issues or breech or transverse foetal presentation, a uterine anomaly should always be excluded.



IMPERFORATE HYMEN

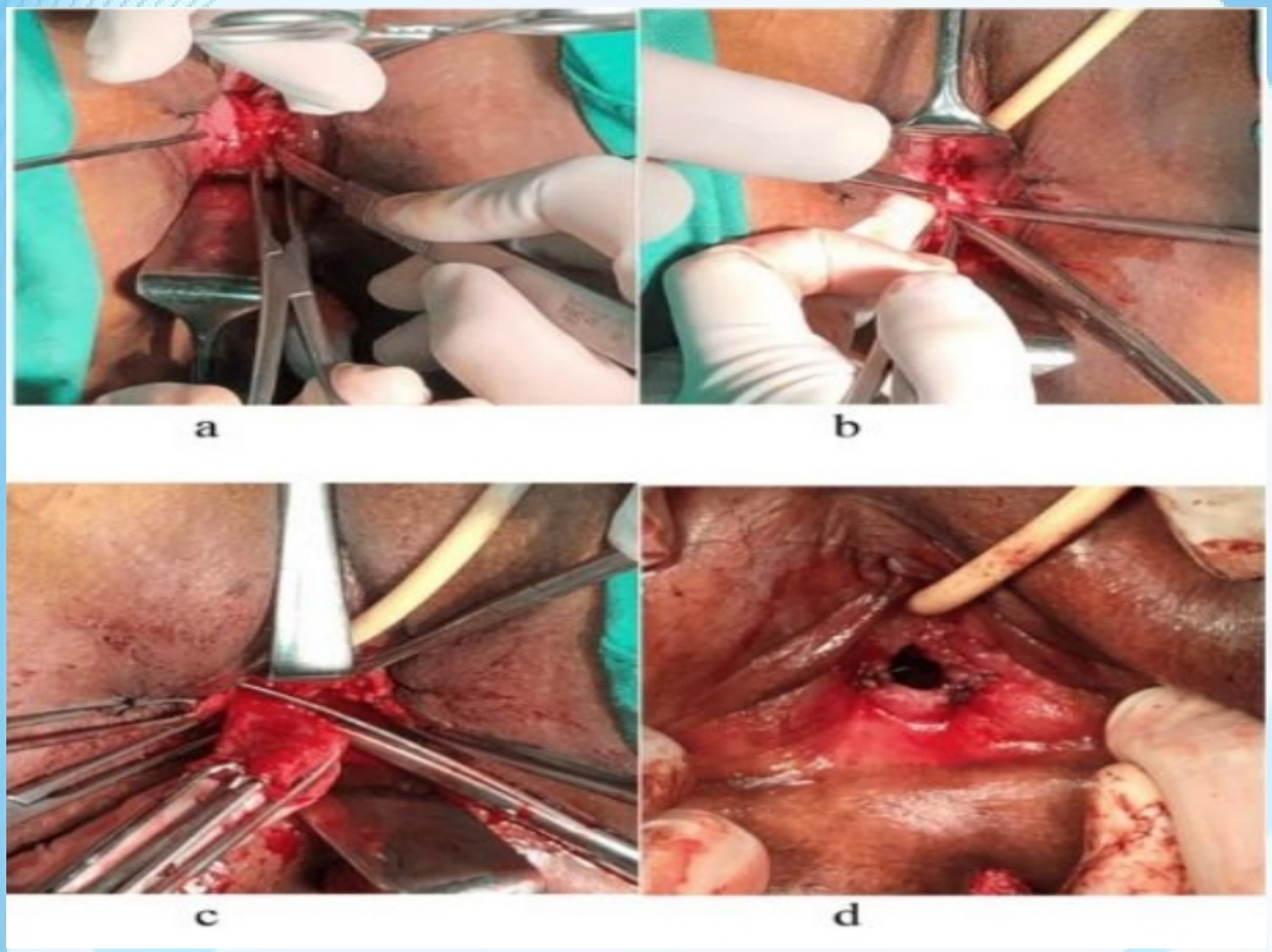
Most common obstructive lesion of the female genital tract. Occurs due to failure of inferior plate of vagina to canalise. Typically, adolescents present with cyclical pain as cryptomenorrhea. Definitive surgery should be done after appropriate evaluation of external genitalia that reveals bluish distended bulge at the introitus and digital rectal examination. Surgical repair, performed under anaesthesia, consists of an elliptical incision in the membrane close to the hymenal ring followed by evacuation of the obstructed material. The vaginal mucosa is sutured to the hymenal ring to prevent adhesions, recurrence and to prevent an incomplete egress of menstrual blood which can lead to ascending infection. Goal of

INCOMPLETE HYMENAL FENESTRATION

Partial obstruction of the hymen includes: micro perforate, septate, or cribriform. Mostly asymptomatic, may present with difficulty in inserting tampon or difficulty in coitus. Treatment involves resection of the excess hymenal tissue and interrupted sutures are placed to reapproximate the tissue.

TRANSVERSE VAGINAL SEPTUM

Results when there is failure of fusion and / or canalization of the urogenital sinus and mullerian ducts. Diagnosis is suspected when there is foreshortened vagina with inability to visualize cervix and presence of hematometra is encountered. MRI is useful in identifying whether cervix is present. Management of the vaginal septum with drainage of hematocolpos at an early age is necessary to preserve fertility and reduce the risk of endometriosis. Thin septa are treated with primary resection and end to end anastomosis. In case of thick vaginal septa, stenosis at the resection site remains the most common complication. Postoperative vaginal dilation may help to reduce scarring and stenosis.



LONGITUDINAL VAGINAL SEPTUM

The septum can involve the entire vagina or part of the vagina. Typically associated with uterine anomalies such as septate uterus and uterus didelphys. Thus, imaging should also involve evaluation of uterine and cervical anomalies.

Surgery is not required in asymptomatic patients. However, in symptomatic women, treatment involves complete resection of septum. Septal tissue should be excised in total as retained fragments of septum may cause dyspareunia. The septal tissue is resected by wedging out the fibrous septum and the normal mucosa from each vagina is sutured together over the defect created by resection.

OBSTRUCTED HEMIVAGINA

It may be associated with ipsilateral renal agenesis, renal anomalies, single uterus or two uteri, and
two cervices are typically present. Diagnosis made by history and physical examination, combined with appropriate imaging (typically MRI). Associated renal anomalies do not required further evaluation or referral to a urologist if patient is asymptomatic.
For patient with ectopic ureter,urologic evaluation and management are beneficial. During the procedure, the obstructed vagina is entered, the fluid is drained, and then the vaginal tissue between the two vaginas is resected.
The two cervixes may be at different levels; therefore, the tissue between them should be left in place so as not to compromise their blood supply. The surgery can be performed as a single procedure or by creation of window with second operation performed at a later time after the inflammation and distension have resolved.

VAGINAL AGENESIS (Mayer -Rokitansky-Kuster-Hauser syndrome)

Congenital absence of vagina with variable uterine development. 7-10 percent have functional endometrium within either a uterus that is obstructed, but otherwise structurally normal, or a rudimentary uterine horn. Patients often exhibit extragenital anomalies-Urologic, renal, skeletal abnormalities, other less common anomalies like congenital heart lesions and inguinal and femoral hernias. On examination vaginal dimple or small pouch is typically seen.

Nonsurgical treatment with self-dilation is recommended by ACOG. Surgery may be appropriate if nonsurgical methods fails or patient elects for surgery. Vaginal dilator is also needed pre or post operatively.

* NONSURGICAL PROCEDURE:

Frank and Ingram procedure- Using vaginal dilators progressively invaginate the vagina. Time
necessary to create vagina varies from 4 months to several years. Ingram modification involves the use of bicycle seat mounted on a stool to create pressure for
vaginal dilation.

* SURGICAL PROCEDURES:

McIndoe procedure: Utilizes split thickness skin graft. Skin graft is placed over a mould with dermal side out and sewn together to form a tube with one closed end. Transverse incision made at vaginal dimple and mould inserted and labia minora secured around the stent and later mould removed after 7 days.
William's vaginoplasty: Horseshoe shaped incision is made on perineum and full thickness skin flaps from labia majora are used to create kangaroo-like pouch. Vaginal dilator then used for 3 to 4 weeks postoperatively
Sigmoid vaginoplasty: Segment of sigmoid colon used for vaginoplasty
Modified vecchietti procedure: Olive is attached to a traction device that rests on the abdomen by sub peritoneal sutures placed laparoscopically. Sufficient traction is applied to olive to produce invasion, thereby creating neovagina.

Davydov procedure: Laparoscopic approach which includes dissection of recto vesical space, abdominal mobilisation of peritoneum to create the vaginal fornices, and attachment of the peritoneum to the introitus.

*** AGENESIS OF LOWER VAGINA:**

Results due to abnormal development of Sino vaginal bulbs and vaginal plate. Treatment is surgical, optimally when a large hematocolpos is present.

A transverse incision is made where the hymenal ring is to be located, dissection is carried until upper vagina is reached and obstruction is drained. Vaginal mucosa is brought down and sutured to hymenal ring.

*** UTERINE ANOMALIES**

Surgical correction is not warranted in asymptomatic patients and in patients with primary infertility as these typically do not prevent conception and implantation.

*** SEPTATE UTERUS**

Most common uterine anomaly. Evaluated in cases of infertility and dysmenorrhoea.

Management includes

1.Hysteroscopic metroplasty – most preferred. A partial septum may require only incision of the septum, after which it can be seen to spring away and open the upper uterine cavity. A large septum requires resection rather than incision. To recognise the base of the septum, one approach is to continue cutting until increased bleeding is noted, but this approach will not work if a coagulative instrument is used. Alternatively intraoperative USG or concurrent laparoscopy may be helpful.

2.Laparoscopic or open trans myometrial repair:

a) Jones metroplasty - Refers to wedge resection of portion of uterine fundus containing the septum. It is important to control the bleeding either using vasopressin or with tourniquet. The wedge is incised until the common uterine cavity is found and the septum can be totally removed.

Reconstruction of the uterus includes layered suturing. First layer includes endometrium and small amount of myometrium with knot placed into cavity.

b) Tompkins metroplasty – Similar to Jones procedure with no tissue resection. Uterus is incised in midline in antero-posterior plane until cavity is reached. Bilateral cavities are then unroofed, using sharp dissection, and the uterus is closed as in Jones procedure.

Attempts at pregnancy may begin two months post operatively if the procedure is deemed adequate.

ROBERTS UTERUS

Rare congenital anomaly, characterized as an asymmetric septate uterus that has a blind hemi cavity with unilateral menstrual fluid retention and a unicornuate hemi cavity connecting to the cervix . Surgery option is ultrasound guided hysteroscopic resection of septum.

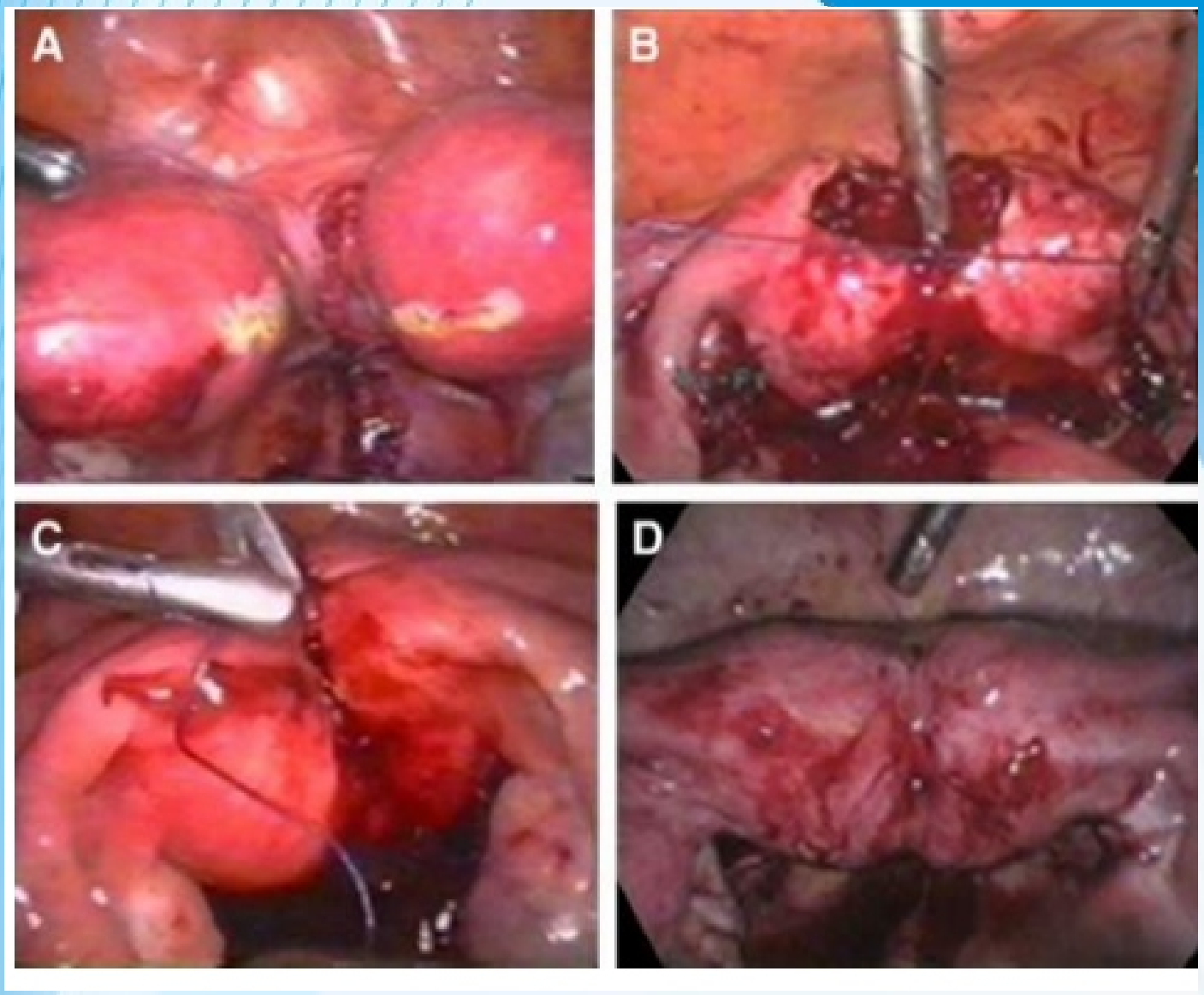
BICORNUATE UTERUS

Patients with partial bicornuate with indentation less than 1cm are not at increased risk of adverse

pregnancy outcomes Patients with severe bicornuate uteri may or may not benefit from surgery as the adverse pregnany outcome may or may not be related to the anomaly, so unification is only performed in patients with history of poor pregnancy outcome that is thought to be related to the anomaly after exclusion of other causes.

Strassman procedure: Transverse incision across the uterine fundus from one cornual end to the other, then two cavities are united into one by closing the incision in vertical direction with layers of interrupted suture. Attempts for conception can be initiated three to six months after surgery.

Patients who conceive should be delivered by caesarean.

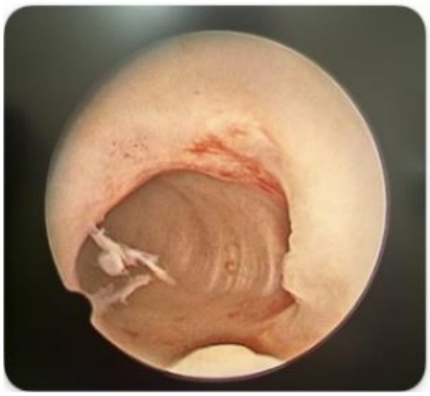


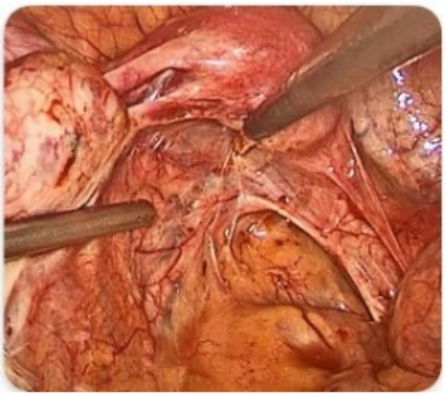
UNICORNUATE UTERUS

Itrasound will help to determine whether a hemi uterus with endometrium is present on the side

opposite the unicornuate uterus. Kidneys should be imaged.

Resection of an obstructed rudimentary horn: Obstruction is often associated with retrograde menses; the surgeon should be prepared to deal with endometriosis For excision of non-fused obstructed hemi uterus, begin hemi hysterectomy at the associated round ligament. The Ligament is transected and the retroperitoneal space is opened. Uterus is then retracted medially and the utero ovarian ligament is transected and then removed from the abdominal cavity. For excision of obstructed hemi uterus that is fused with the patent unicornuate system, the fused hemi uterus is excised from the non-obstructed side and then the non-obstructed side is closed in layers. Patients desiring fertility can attempt to conceive immediately after

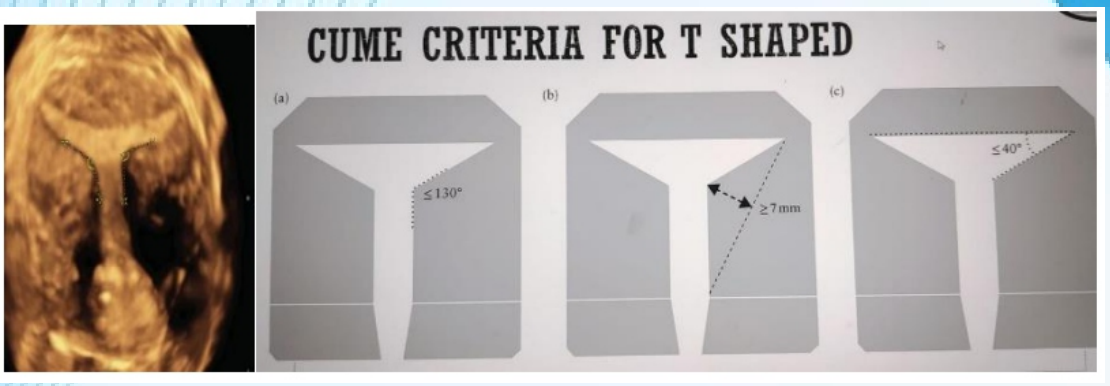




T SHAPED UTERUS

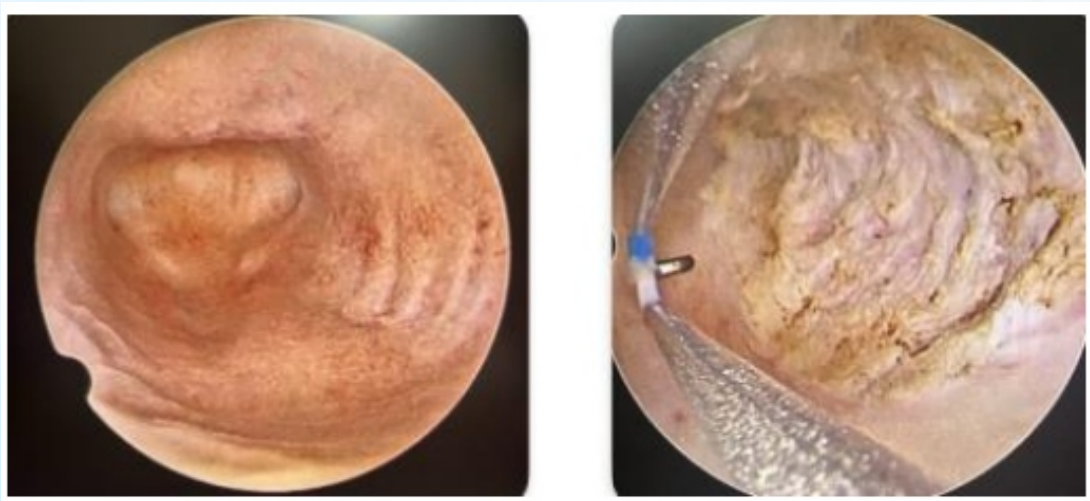
Diagnosis is by 3D ultrasound.

Hysteroscopic metroplasty using resectoscope is the treatment of choice.



I SHAPED UTERUS

Variant of T-Shaped uterus, mostly seen in patients with hyperandrogenism. Surgical anagement – lateral metroplasty is the treatment of choice.



ACUM – Accessory and Cavitated Uterine Mass

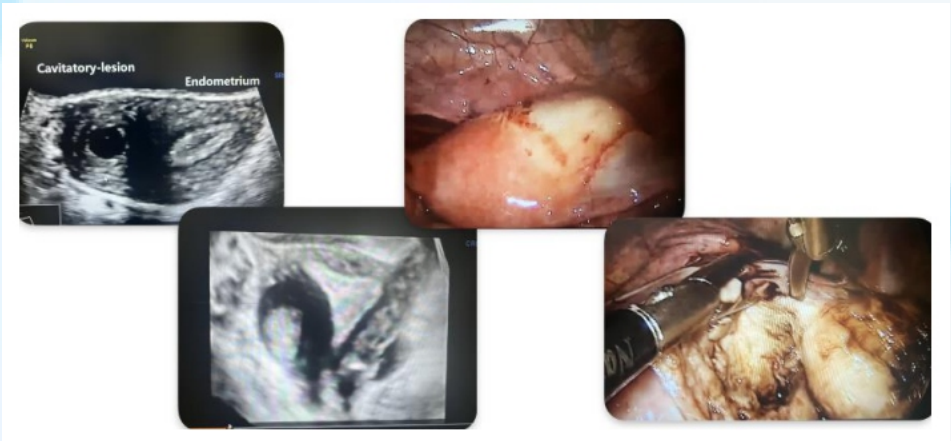
Diagnostic criteria:

> An isolated accessory cavitated lesion in lateral myometrium containing functional endometrium surrounded by myometrial mantle.

> Has concentric organisation of smooth muscle.

> Normal uterus, endometrial cavity, tubes and ovaries.

> Cavitated lesion filled with dark brown haemorrhagic content.



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