

DEFINITION

“ Congenital anomaly of the genitalia is a medical term referring to any physical abnormality of the male and female internal or external genitalia present at birth”

CAUSES

- Exact cause is unknown
- Hormone deficiency and excess
- Teratogenic effects
- Genetic determination

DEVELOPMENTAL ANOMALIES OF EXTERNAL GENITALIA

❖ PERINEAL OR VESTIBULAR ANUS-

- Detected at time of birth.
- The anal opening is situated either close to posterior end vestibule or in vestibule.
- Rarely it is situated in vagina (congenital recto vaginal fistulla)
- Future reproduction not a problem, generally cesarean section advised.

❖ ECTOPIC URETER-

VAGINAL ABNORMALITIES

❖ NARROW INTROITUS-

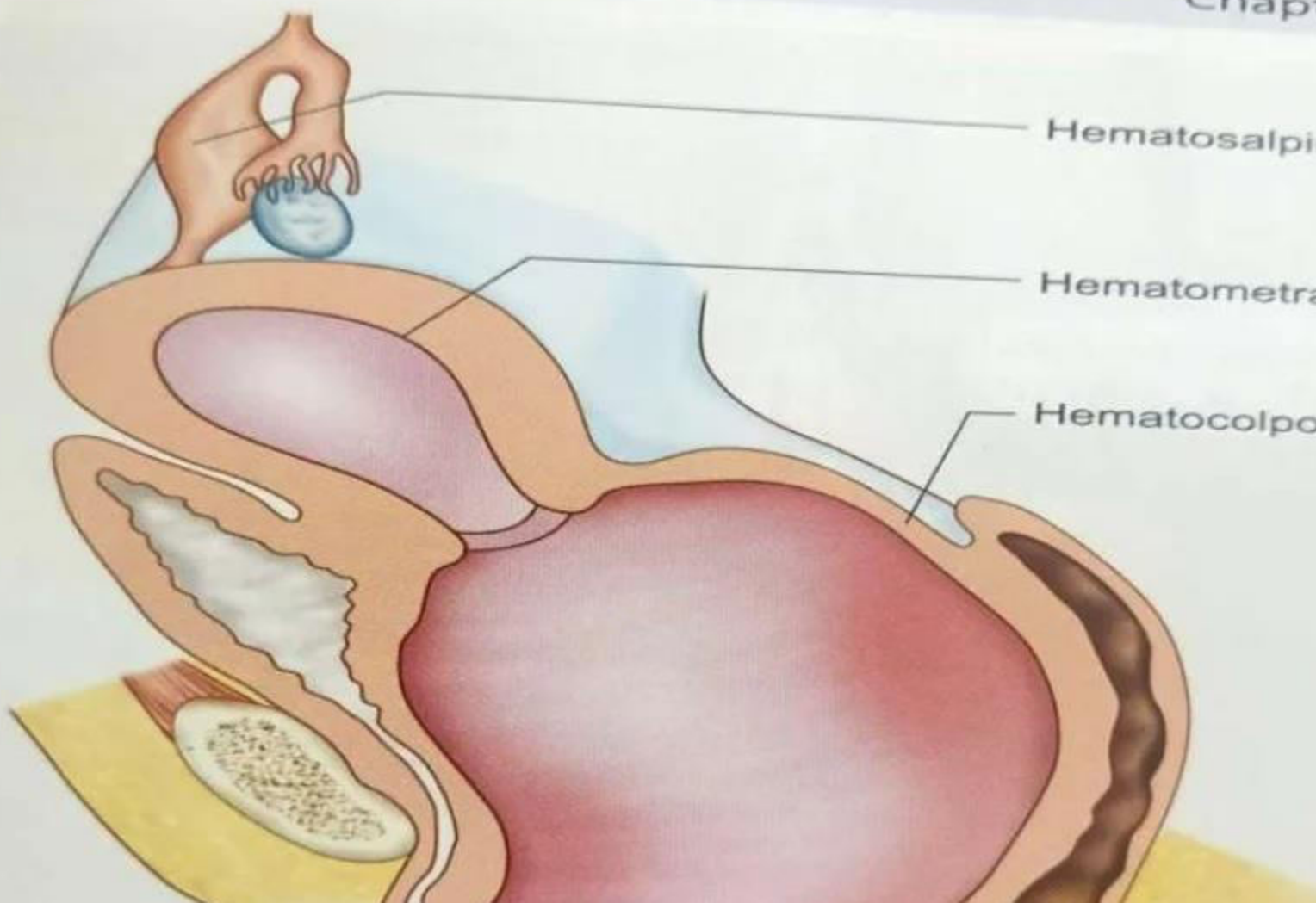
- Revealed after marriage because “dyspareunia” may be the first complaint.

It may be detected during investigation of infertility.

- Surgical enlargement is treatment.

❖ HYMEN ABNORMALITY-

- Imperforate hymen is important abnormality .
- Always unnoticed until 14-16 years of age.
- Uterine functioning is normal menstrual blood is pent up inside vagina behind hymen. (**Cryptomenorrhoea**)
- Depending upon amount of blood accumulated finally descend into vagina (**Hematocolpos**)
- The uterus is next involved and cavity dilated (**Hematometra**)



➤ Symptomms-

- Periodic lower abdominal pain, continuous nature.
- Primary amenorrhoea.
- Dysuria and retention of urine due to elongation of urethra.
- On examination-
 - Abdominal exam. : Suprapubic swelling

➤ **TEATMENT –**

- Incision made in hymen, quadrants of the hymen partially excised, not too close to vaginal mucosa.
- Spontaneous escape of dark tarry colored blood following incision.
- Antibiotic should be given

❖ VAGINAL MALDEVELOPMENTS-

➤ **Agenesis of vagina –**

- It is a birth defect when vagina does not develop fully. (1:5000)

➤ **Septate vagina-**

- Complete or incomplete longitudinal septum.
- Transverse vaginal septum.
- May be asymptomatic or may represent

➤ **Partial atresia of vagina-**

- Segment of vagina may be atretic , it may be associated with cervical atresia.
- Primary amenorrhoea, periodic pain in lower abdomen .
- Excision of atretic portion and suturing the margins of vaginal wall.

➤ **Complete atresia of vagina-**

ASSOCIATED ABNORMALITIES

- Vesico-vaginal fistula- Abnormal opening between vagina and bladder.
- Recto-vaginal fistula- Abnormal opening between rectum and vagina.

UTERINE ANOMALIES

- **American Fertility Society (AFS) classification-**

Class I: Mullerian agenesis/ Hypoplasia

Class II: Unicornuate uterus with or without rudimentary horn.

Class III: Uterus didelphys

Class IV: Bicornuate uterus

Class V: Septate uterus

MULLERIAN AGENESIS

Failure of development of one or both Mullerian ducts:

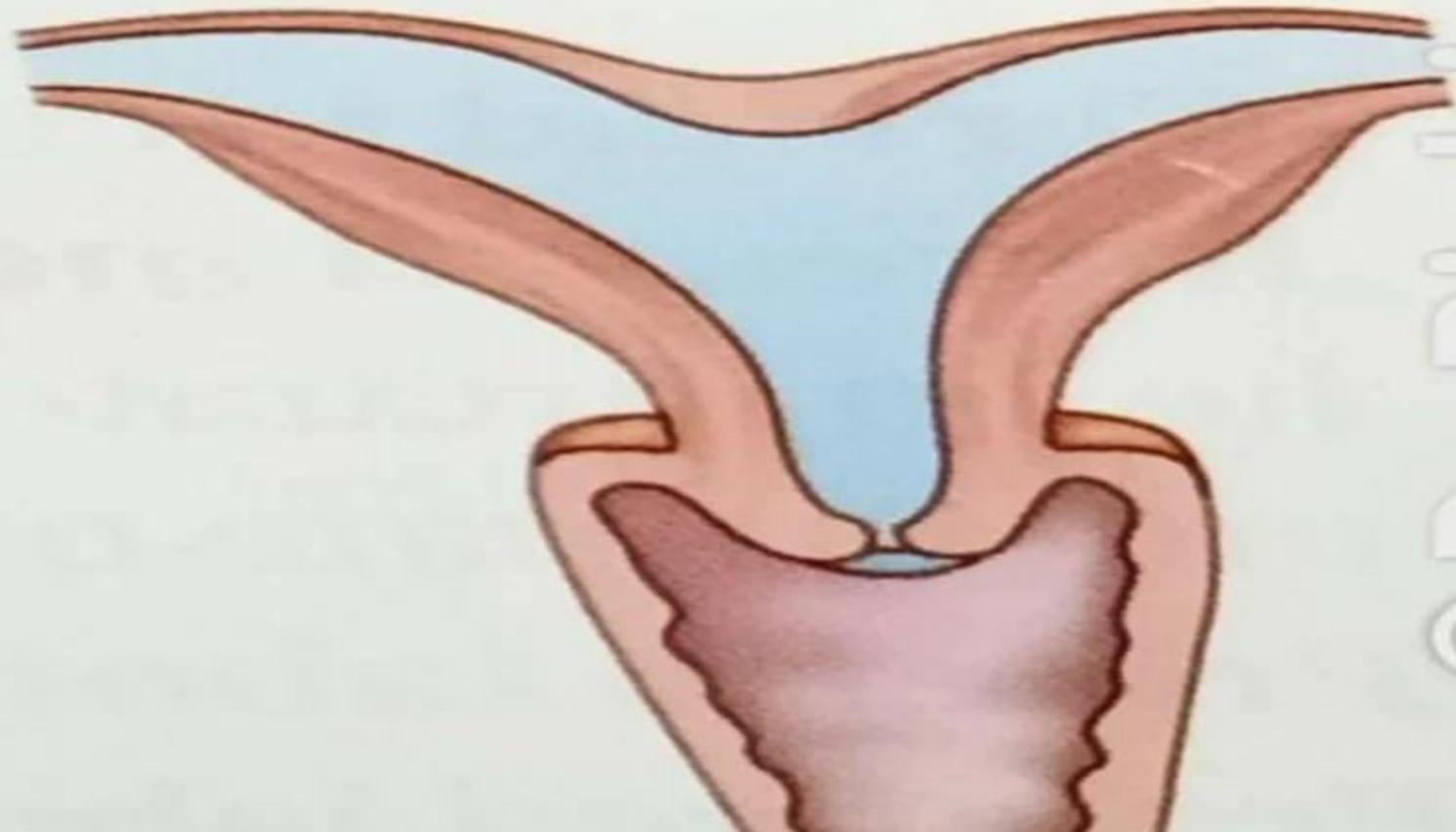
- Absence of both ducts leads to absence of uterus including oviduct and absence or vagina as well.

Primary amenorrhoea is chief complaint.

- Absence of one duct leads to unicornuate

ARCUATE UTERUS (18%)

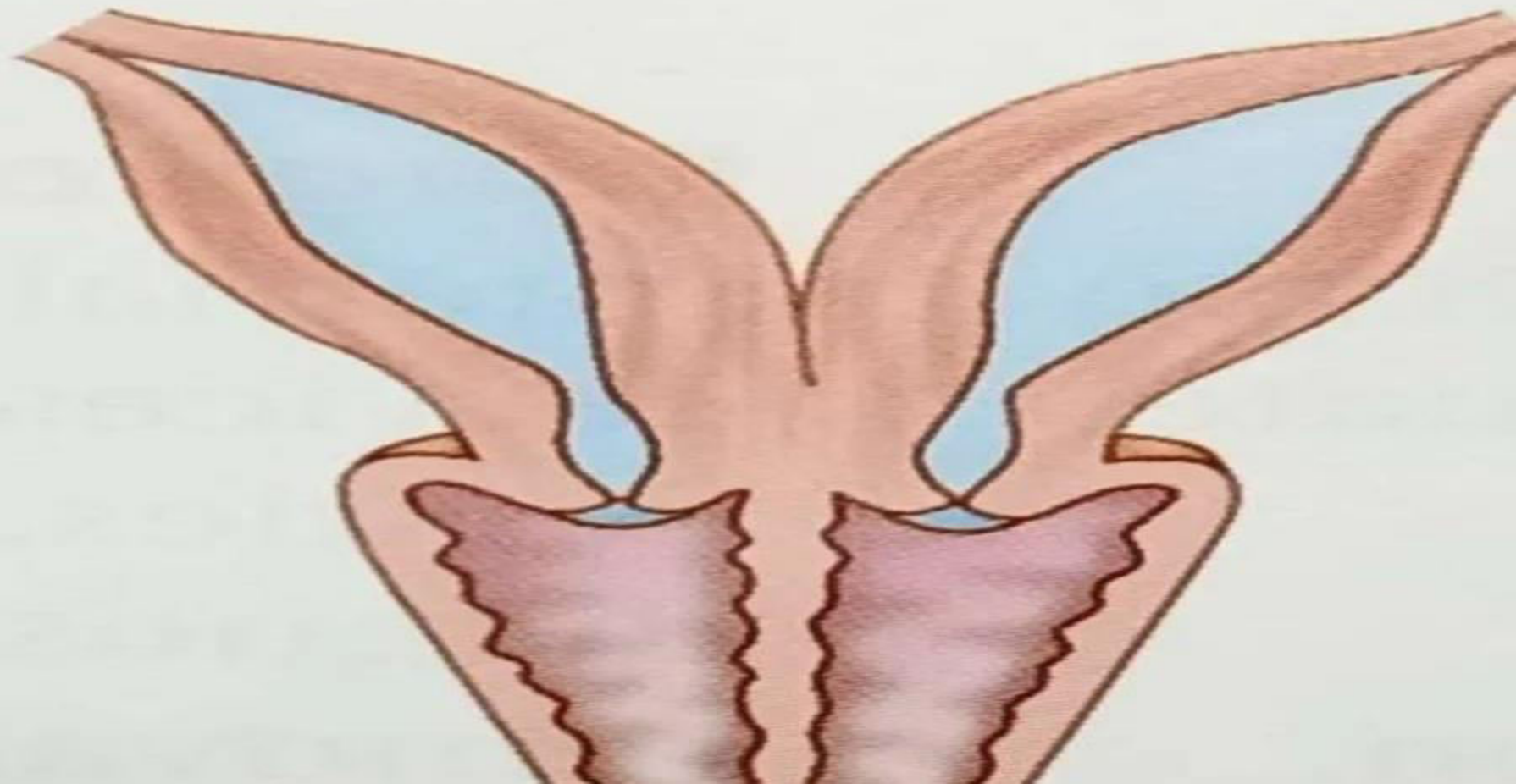
- Fundus looks concave with heart shaped cavity outline.
- This looks more like a normal uterus, except it has a deep or slight indentation at the top.
- It is a common abnormality, affecting 1:25 in general population



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UTERUS DIDELPHYS (8%)

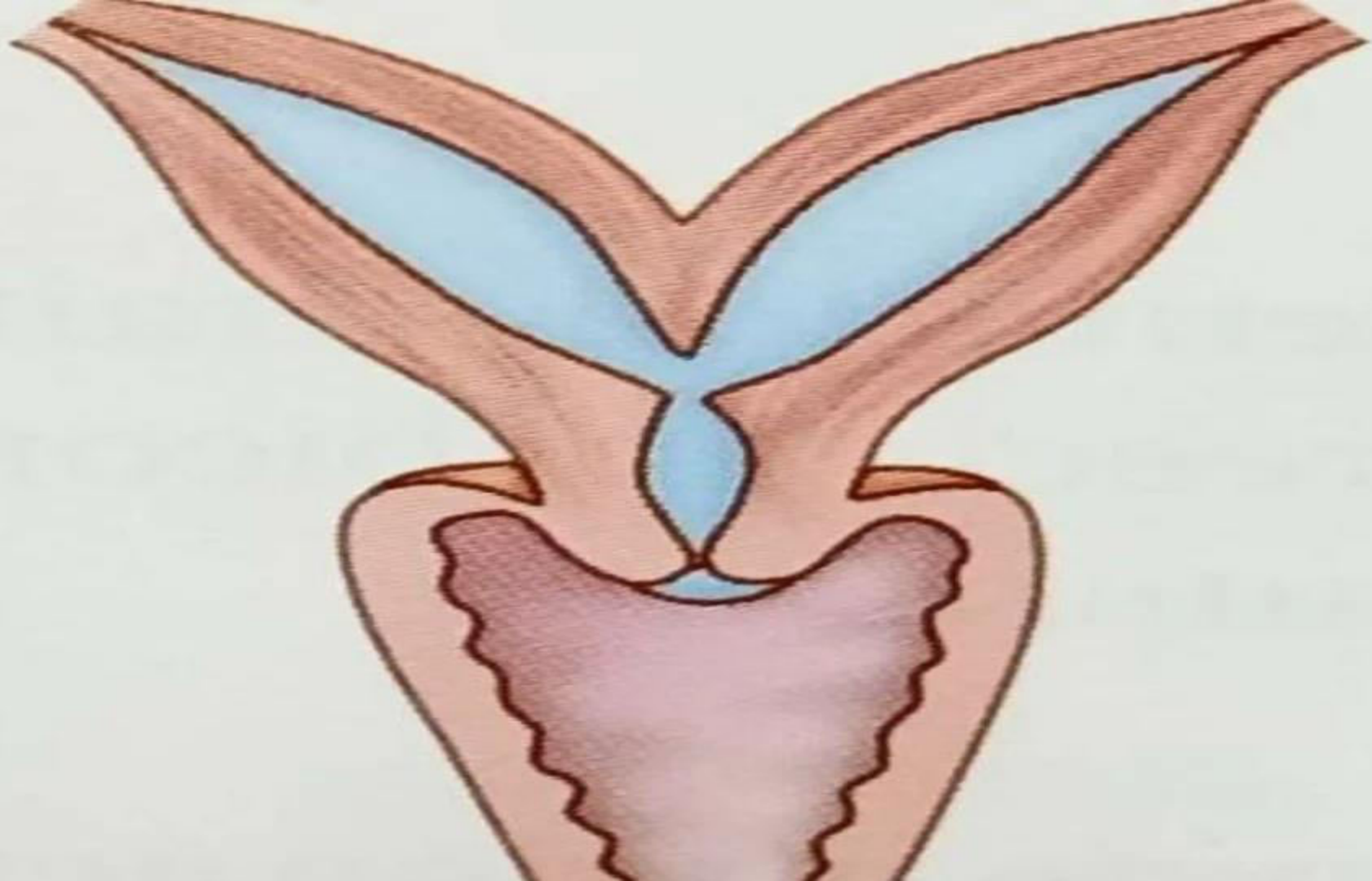
- Complete lack of fusion of the Mullerian ducts with a double uterus and double vagina.
- Uterus has two inner cavities, each cavity may leads to own cervix and vagina, so there are two cervix and two vaginas.
- It is uncommon affecting 1:350 women.



UTERUS BICORNIS (26%)

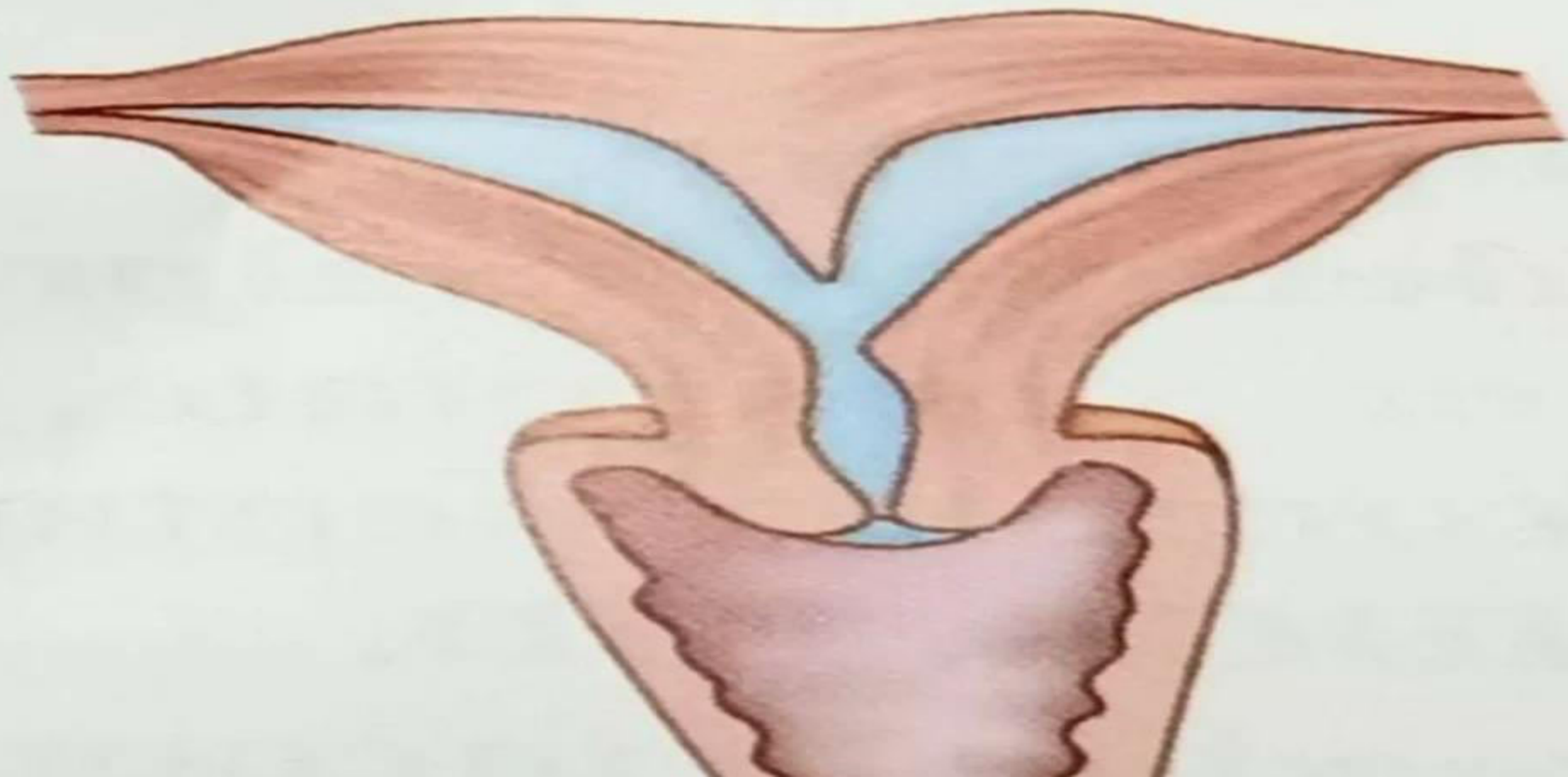
There are varying degrees of fusion of muscle walls of two ducts.

- **Uterus bicornis bicollis:** There are two uterine cavities with double cervix with or without vaginal septum.
- **Uterus bicornis unicollis:** There are two



SEPTATE UTERUS (35%)

- Two Mullerian ducts fused together but there is persistence of septum in between two either partially or completely



UNICORNUATE UTERUS (10%)

- Failure of development of one Mullerian duct

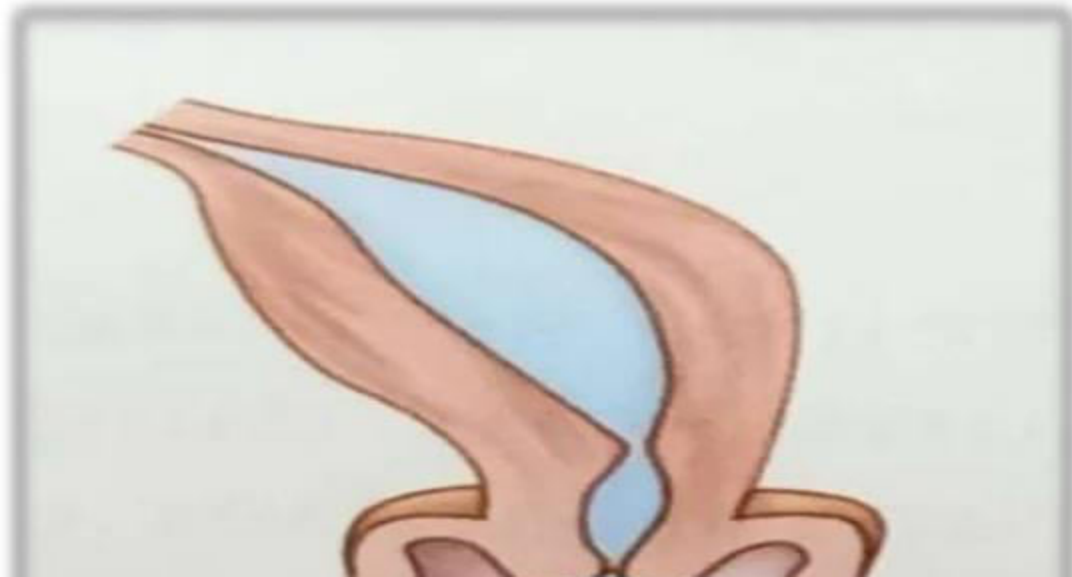




Fig. 4.6C: Hystero-graphic view of a septate uterus
(*Courtesy: Dr SS Murmu, RMO-clinical tutor, BSMCH*)



Fig. 4.6D: Hystero-graphic view of an arcuate uterus
laparoscopy



DES-RELATED ABNORMALITIES

It is due to DES exposure during intra uterine life, variety of mal formations are included-

- Vagina: Adenosis, adenocarcinoma
- Cervix : Cockscomb cervix, cervical collar.
- Uterus : Hypoplasia, T-shaped cavity, uterine synechiae.

CLINICAL FEATURES

❖ GYNAECOLOGICAL-

- Infertility and dyspareunia (vaginal septum)
- Dysmenorrhoea in bicornuate uterus or due to cryptomenorrhoea (pent up menstrual blood in rudimentary horn)
- Menstrual disorders like menorrhagia (due to

❖ **OBSTETRICAL-**

- Midtrimester miscarriage
- Rudimentary horn pregnancy.
- Cervical incompetence
- Increased risk of mal presentation.
- Pre-term labor, IUGR, IUD.
- Prolonged labor (d/t inco-ordinated uterine actions)
- Obstructed labor (obstruction due to non gravid

INVESTIGATIONS

- Hystero-graphy
- Hystero-scopy
- Laparo-scopy
- TVS USG.
- MRI

TREATMENT

Reproductive outcome:

- Septate uterus (86%)
- Bicornuate uterus (50%)
- Unicornuate uterus (40%) poor
- Uterus didelphys has best possibilities of successful pregnancy. (64%)

TREATMENT:

- Rudimentary horn excision to reduce ectopic pregnancy risk.
- Unification operation in bicornuate and septate uterus.
- Hysteroscopic metroplasty – resection of an intrauterine septum maintaining symmetry of uterine cavity.(80-90% success rate)

OVIDUCT ABNORMALITIES

- Elongated fallopian tube
- Absent of one side tube.

OVARY ABNORMALITIES

- Gonadal agenesis.
- Presence of accessory ovary.

OTHER ABNORMALITIES

- **LABIA MINORA-** Labia fusion due to developmental defect.
- **LABIA MAJORA-** Hyperplastic, hypoplastic, abnormal fusion
- **CLITORAL ABNORMALITY-** Clitoral duplication, clitoral megalia

THANK YOU