MULLEIAN ANOMALIES
By

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Uterus

- Pear-shaped muscular organ
- 8 cm long
- 5 cm wide
- 3 cm thick
- Non-pregnant state
- Pelvic organ
MULLERIAN ANOMALIES

- Normal
- Complete Septate
- Partial Septate
- Partial Bicornuate
- Complete Bicornuate
- Didelphic
- Unicorneate
INTRODUCTION

- MD forms tubes, uterus, cervix and upper part of vagina
- Ranges from agenesis to duplication.
- Associated with renal and axial skeletal systems anomalies
- Has varying presentation ranging from primary amenorrhea to menstrual disorders, infertility and pregnancy complications like PTL, Ectopic, etc
- MDA has varying treatment from ability to have coitus to conceive and deliver normal babies.
INCIDENCE

- Dates back to 16th century a case utero vaginal agenesis
  - Columbo et al (1600)
- General population – 0.1-3.5% - Byrene et al
- Fertile women – 4.3%
- Infertile women – 3.6%
- Sterile group - 2.4%
- Recurrent Aborters 5 - 13% - Grimbizis et al
ETIOLOGY

- Dysregulation occurring in differentiation, migration, fusion and canalisation
- Associated with renal anomalies, axial skeletal anomalies and rarely cardiac and auditory anomalies
- Probable causes: Intrauterine infection, genetic aberration, Teratogens like DES and Thalidomide.
GENETICS OF MDA

- Sporadic
- Familial
- Multifactorial
- Autosomal dominant
- Autosomal recessive
- X linked
- Variants of GALT (Galactose 1 phosphate uridyl transferase enzyme defect)

Genes Associated :- HOXA 9, 13 & WNT 4
Embryogenesis of the Reproductive Tract
CLASSIFICATION OF MDA

- 1979 – Buttram and Gibbons classification
  Modified

- 1988 – American Fertility Society classification
American Fertility Society Classification of Mullerian Anomalies

<table>
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<th>I. Segmental müllerian hypoplasia or agenesis</th>
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<td>B. Cervical</td>
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<th>II. Unicornuate uterus</th>
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<td>A. Communicating rudimentary horn</td>
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<td>B. Noncommunicating horn</td>
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<td>C. No endometrial cavity</td>
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<td>D. No rudimentary horn</td>
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<th>III. Uterine didelphys</th>
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<th>IV. Bicornuate uterus</th>
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<td>A. Complete (division to internal os)</td>
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<tr>
<th>V. Septate uterus</th>
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<tbody>
<tr>
<td>A. Complete (septum to internal os)</td>
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<td>B. Partial</td>
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<th>VI. Arcuate</th>
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| VII. Diethylstilbestrol related              |

Source: Semin Reprod Med © 2006 Thieme Medical Publishers
INCIDENCE OF MDA ACCORDING TO AFS

- Arcuate uterus: 32.8%
- Septate uterus: 33.6%
- Bicornuate uterus: 20.0%
- DES exposed uterus: 0.8%
- Unicornuate
- Uterine didelphys: 33%
EFFECT OF MDA UPON REPRODUCTION

- Infertility
- Endometriosis
- Ectopic pregnancy
- Recurrent Pregnancy Loss
- Prematurity, IUGR, fetal malposition
- Uterine dysfunction
- Uterine rupture
- Increased perinatal morbidity and mortality
DIAGNOSIS OF MDA

- Clinical
- Hystero salphingogram
- Sonosalphingogram
- MRI – 100% accuracy
- Hystero laparoscopy
- Laparotomy
Vulvar Abnormalities

Vulval and lower 1/3rd vagina atresia

Labial Fusion
- Most commonly due to congenital adrenal hyperplasia.

Imperforate hymen
- Persistence of the fusion between the sinovaginal bulbs at the vestibule
- Associated with primary amenorrhea and hematocolpos
Vaginal Abnormalities

Developmental abnormalities of the normal single vagina include:

- Vaginal agenesis
- Vaginal atresia
- Double vagina
- Longitudinal vaginal septum
- Transverse vaginal septum
Obstetrical significance of vaginal abnormalities

- Complete mullerian agenesis – pregnancy is impossible because uterus and vagina is absent

- About one third of women with vaginal atresia have associated urological abnormalities

- Complete vaginal atresia – precludes intercourse and then pregnancy

- In most cases of partial atresia, because of pregnancy-induced tissue softening, obstruction during labor is gradually overcome. interferes with descent
Obstetrical significance of vaginal abnormalities

- **Complete longitudinal vaginal septum** usually does not cause dystocia because half of the vagina through which the fetus descends dilates satisfactorily.

- **Incomplete septum**, however, occasionally interferes with descent.
Cervical Abnormalities

**Atresia.**
- This may be combined with incomplete development of the upper vagina or lower uterus

**Double cervix.**
- Each distinct cervix results from separate müllerian duct maturation.
- Both septate and true double cervices are frequently associated with a longitudinal vaginal septum.
- Many septate cervices are erroneously classified as double.

**Single hemicervix.**
- This arises from unilateral müllerian maturation.

**Septate cervix.**
- This consists of a single muscular ring partitioned by a septum.
- The septum may be confined to the cervix, or more often, it may be the downward continuation of a uterine septum or the upward extension of a vaginal septum.
Uterine anomalies
CLASS I- ROKITANSTY SYNDROME

- Primary amenorrhea
- Feminine patients
- Short vagina

DD: Testicular feminization syndrome
Class I mullerian anomaly: Segmental mullerian agenesis or hypoplasia with subdivisions.
INVESTIGATIONS

- Karyotyping
- USG/MRI
- Hormone assay
- IVP (associated vertebral anomalies can be detected) and renal sonography
- Diagnostic Laparoscopy is not routinely done.
TREATMENT

- Vaginal Reconstruction
  - Vagino plasty: Mac Indoes Vaginoplasty; Williams vulvovaginoplasty, Vecchietti procedure
- Fertility – by surrogacy
- Psychological support
Women with a unicornuate uterus have an increased incidence of infertility, endometriosis, and dysmenorrhea.

Implantation in the normal-sized hemiuterus is associated with increased incidence of:

- spontaneous abortion
- preterm delivery
- intrauterine fetal demise
UNICORNUATE UTERUS

- Unilateral failure of development of MDA
  Incidence: 2.5-13%

Types: Unicornuate

  Unicornuate with rudimentary horn
    - Communicating
    - Non communicating
      - with endometrium
      - without endometrium

Associated Renal anomalies like renal agenesis, Horseshoe kidney and pelvic kidney 44% (In the presence of obstructed horn)
Class II

A-1-a. Communicating

A-1-b. Noncommunicating

A-2. No cavity

B. No horn
CLINICAL FEATURES

- Haematometra
- Endometriosis
- Preterm labour – 43%
- IUGR
- Mal presentation
- Ectopic -4.3%
- Pregnancy in accessory horn -2%
- Rupture uterus
IMAGING MODALITIES IN UNICORNUATE UTERUS

HSG  3D USG  MRI
DIAGNOSIS AND SURGICAL MANAGEMENT

- HSG – non communicating horn cannot be diagnosed
- USG – 3D or High Resolution
- MRI – banana shaped uterus
- Laparoscopy – indicated for excision of rudimentary horn which has endometrium
- IVU or renal sonography

Cervical encirclage is mandatory if patient conceives
REPRODUCTIVE OUTCOME IN UNICORNUATE UTERUS

- Live birthrate: 43.7%
- Abortion rate: 35-43%
- Preterm delivery: 27%
- Term delivery: 31%
NONCOMMUNICATING RUDIMENTARY UTERINE HORN
* attached fallopian tube (arrow) was patent*
UNICORNUATE UTERUS WITH RUDIMENTARY HORN
Uterine Didelphys (Class III)

- This anomaly is distinguished from bicornuate and septate uteri by the presence of complete nonfusion of the cervix and hemiuterine cavity.
- Except for ectopic and rudimentary horn pregnancies, problems associated with uterine didelphys are similar but less frequent than those seen with unicornuate uterus.
- Complications may include:
  - preterm delivery (20%)
  - fetal growth restriction (10%)
  - breech presentation (43%)
  - cesarean delivery rate (82%)
DIDELPHYS

- Failure of midline fusion of MD either completely or partially
- Incidence: 11%
- Types: Total Septum
  - Partial Septum
  - Transverse Septum
Class III müllerian anomaly: Uterine didelphys.
CLINICAL FEATURES

- Asymptomatic – Failure of tampons to obstruct menstrual flow
- Hematometrocolpos if there is
- Hematometra obstruction
- Hematosalpinx 20% renal anomalies
- Endometriosis
- Other associated anomalies: bladder exstrophy, cervical agenesis
IMAGING MODALITIES IN DIDELPHYS UTERUS

HSG | 3DUSG | MRI
DIAGNOSIS & SURGICAL MANAGEMENT

- Clinical
- USG
- MRI- 2 widely separated uterine horns, 2 cervices are typical identified. Intercornual angle > 60 degree
- Laparoscopy
- IVP
UTERUS DIDELPHYS
SURGICAL MANAGEMENT

- With obstruction
  - Excision of the horn
- Non obstruction
  - Strassmann metroplasty only in selected cases

Cervical encirclage is mandatory if patient conceives
REPRODUCTIVE OUTCOME IN DI DELPHYS

- Term delivery: 20%
- Ectopic: 2.3%
- Abortion: 20%
- Live birth: 68%
- Preterm delivery: 24%
Bicornuate and Septate Uteri (Classes IV and V)

- Marked increase in miscarriages that is likely due to the abundant muscle tissue in the septum
- Pregnancy losses in the first 20 weeks were reported by Buttram and Gibbons
  - 70 percent for bicornuate
  - 88 percent for septate uteri
- There also is an increased incidence of preterm delivery, abnormal fetal lie, and cesarean delivery.
BICORNUATE UTERUS

- Incomplete fusion of MD at uterine fundus level
- Incidence - 20%
- May be complete - bicornuate bicollis
- May be incomplete - bicornuate unicollis
Class IV

A. Complete

B. Partial

C. Arcuate
ULTRASOUND IMAGING OF SEPTATE AND BICORNUATE UTERUS

Anna Lev-Toaff, MD, Thomas Jefferson University, PA
Clinical features

- Asymptomatic
- Abortion 28%
- Preterm delivery 25%
- Live birth 63%
IMAGING MODALITIES IN BICORNUATE UTERUS

HSG  3D USG  MRI
DIAGNOSIS

- To be differentiated from septate uterus
- HSG
- USG during luteal phase shows 2 endometrial cavities with a deep dimple in the fundus.
- MRI – Ideal
- Intercornual distance is >105 degrees
- Myometrial tissue is seen in bicornuate uterus Vs septum in septate uterus with angle of <75 degree
- Laparoscopy
SURGICAL MANAGEMENT

- Metroplasty is reserved only in recurrent aborters
- Strassmann procedure either by Laparoscopy or Laparotomy
BICORNUATE UTERUS
BICORNUATE UTERUS WITH OBSTRUCTION IN ONE HORN
REPRODUCTIVE OUTCOME IN BICORNUATE UTERUS

- Increased incidence in infertile population.
- Term pregnancy rate 60%
- Live birth 65%
- Metroplasty is indicated only when other causes are ruled out.

Acien, 1993
SEPTATE UTERUS

- Incomplete resorption of medial septum
- Incidence: 33.6%
- Types: Complete, Incomplete

DD: Uterus didelphys

Renal tract anomalies are rare
Class V mullerian anomaly: Septate uterus with complete septum to the cervical os (A) or partial septum (B).
CLINICAL FEATURES

- Dyspareunia
- Dysmenorrhoea
- Primary or secondary infertility
- Poor reproductive performance
IMAGING MODALITIES IN SEPTATE UTERUS

HSG  USG  3DUSG  MRI
SURGICAL MANAGEMENT

- Hysteroscopic Septal Resection under Laparoscopic guidance using microscissors, electro cautery, laser, Versa point
- Stop dissecting
  - When both cornuae are seen in the same plane
  - Appearance of vascularity
  - Move the scope from one side to other
SEPTATE UTERUS
REPRODUCTIVE OUTCOME IN SEPTATE UTERUS

- Spontaneous abortion: 33-75%
- Live birth: 62%
- Term deliveries: 51%
- Preterm labour: 10%
- Ectopic: 2%

Metroplasty increases the incidence of live birth to 82%

Acien, 1993
POST OPERATIVE MANAGEMENT

- Estrogens may be used
COMPLICATION

- Uterine perforation
- Hemorrhage
- Cervical incompetence
- Residual septum
Class VI

Arcuate Uterus

- This malformation is only a mild deviation from the normally developed uterus.
ARCUATE UTERUS

- Near complete resorption of the uterovaginal septum.
- Small intrauterine indentation shorter than 1 cm and located in the fundal region diagnosed by HSG.
- Incidence: 32.8%
IMAGING MODALITIES IN ARCUATE UTERUS

HSG

3D USG

MRI
DIAGNOSIS & TREATMENT

- HSG
- MRI
- IVP and renal ultrasound
- Hysteroscopy
- Resection indicated in poor performers
REPRODUCTIVE OUTCOME IN ARCUATE UTERUS

- Preterm delivery: 05.1%
- Live birth: 66.2%
- Ectopics: 03.6%
- Spontaneous abortion: 20.0%
Diethylstilbestrol-Induced Reproductive Tract Abnormalities

- Development of rare vaginal clear cell adenocarcinoma.
- Increased risk of developing
  - cervical intraepithelial neoplasia
  - small-cell cervical carcinoma
  - vaginal adenosis,
  - non-neoplastic structural abnormalities
Diethylstilbestrol-Induced Reproductive Tract Abnormalities

Structural Abnormalities:
- transverse septa,
- circumferential ridges involving the vagina and cervix
- cervical collars
- smaller uterine cavities
- shortened upper uterine segments
- T-shaped and irregular
- oviduct abnormalities
Diethylstilbestrol-Induced Reproductive Tract Abnormalities

- Their incidences of miscarriage, ectopic pregnancy, and preterm delivery are also increased, especially in women with structural abnormalities.
T SHAPED UTERUS
MANAGEMENT OF T SHAPED UTERUS

- Lateral metroplasty
- Encerclage is mandatory in the event of pregnancy
CONCLUSION

- MDA are not so uncommon
- Presents at varying stages of life as primary amenorrhoea, infertility, Recurrent abortion, preterm labour,
- MRI helps in accurate diagnosis
- DHL is indicated only when intervention is needed.
- Corrective surgery improves pregnancy outcome