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Clinical Implications and ESHRE/ESGE Classification of Mullerian Anomalies: A Case Series

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ABSTRACT
Mullerian anomalies-developmental anomalies of the Mullerian system might involve the uterus, cervix, fallopian tubes and vagina of which anomalies of the uterus are the most common. Mullerian or paramesonephric ducts form the female genital tract and any deviation from the normal can lead to Mullerian anomalies.Müllerian duct abnormalities (MDAs) are a broad and complex spectrum of defects that are often associated with primary amenorrhea, infertility, endometriosis, and obstetric complications. Depending on the type of Mullerian anomaly; the presentations, required investigations and treatment options vary considerably and so does the counselling regarding the condition. Due to the highly varied profile of patients with Mullerian anomalies, a number of classification systems have been proposed to aid in their diagnosis and management. A thorough knowledge of the development of female genital tract and its associated anomalies is necessary to diagnose these disorders and an adequate workup is imperative before planning the treatment. This case series comprises of 12 cases of Mullerian anomalies, diagnosed through clinical history, physical examination, radiological scans or surgery. The case series is multiparametric in design and focuses on multiple parameters of an individual case. Through the present case series we aim to evaluate the various clinical presentations of the Mullerian duct anomalies, classify them as per ESHRE classification system, assess the other congenital anomalies associated with them and also study the optimum mode of management used for the respective cases.

I. INTRODUCTION

Reproductive tract anomalies can result from failure of the Mullerian duct to fuse in the midline, to connect with the urogenital sinus or to create the appropriate lumen in the upper vagina and uterus by resorption of the central vaginal cells and the septum between the fused Mullerian ducts. The prevalence is 7%.

The prevalence of various Mullerian anomalies is re-ported to be as follows**:** Arcuate- 20%; Septate- 35%; Bicornuate Uterus- 25%; Unicornuate- 10%; Didephic8% and others- 3%. .

Depending on the type of anomaly, it can either be obstructive requiring surgical intervention or nonobstructive. The time of presentation varies from early adolescence to adulthood.

Congenital anomalies of Mullerian duct are frequently accompanied by anomalies of renal tract such as renal agenesis presenting on the same side of Mullerian duct,severe renal hypoplasia,horseshoe and pelvic kidney.

Investigating modalities include ultrasound, MRI and HSG.

The European society of human reproduction and embryology (ESHRE) and the European society for gynaecological endoscopy (ESGE) have developed a system of classification in 2013 - Congenital uterine anomalies (CONUTA) for a more effective way of classifying and managing Mullerian anomalies.

ASRM Mullerian anomalies classification 2021 is another classification given by American society for reproductive medicine for classifying and managing the case.

Treatment, surgical or non-surgical can be planned on the basis of the presentation, investigations and classification.

II. CASE DESCRIPTIONS

Case 1

A 13 year old patient presented with complain of hypomenorrhea since menarche. On examination, secondary sexual characteristics were well developed according to age. Local examination revealed an oblique vaginal septum while on per rectal examination uterus, along with a mass in right fornix, could be palpated. USG and MRI findings confirmed didelphys uterus with hematocolpos, hematometra and and oblique vaginal septum. She was taken for vaginal exploration. Oblique septum was resected and right hematocolpos, hematometra was drained. Didelphys uterus was confirmed with uterine

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sound. Patient was explained about follow up after next menses.

Case 2

A 20 year old patient was admitted with history of 2 months of amenorrhea and complain of backache. Examination suggested 10-12 weeks size uterus. MRI confirmed pregnancy of 11 weeks 5 days in rudimentary horn, not communicating with uterus. Patient was taken for laparotomy followed by rudimentary horn excision.

Case 3

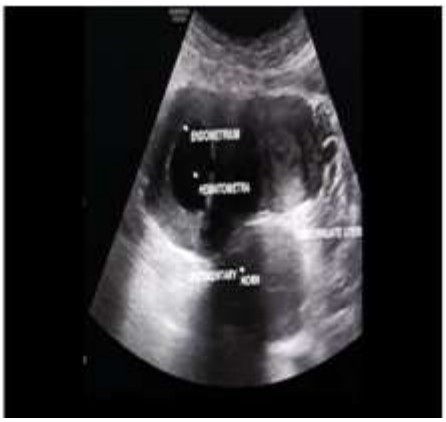
A 13 year old patient presented with complain of primary amenorrhea. On examination, bluish bulge seen on per speculum and shallow vagina on per vaginum examination. USG and MRI confirmed hematocolpos with transverse vaginal septum and hypoplastic lower 1/3 vagina. Patient was taken for vaginal examination under anaesthesia followed by hematocolpos drainage and septum resection with double breasting of vaginal mucosa. Patient was advised for vaginal mould placement regularly. On follow up after 2 months, patient had complain of cyclical pain with scanty menses and urinary incontinence. She had been placing the mould wrongly in urethra leading to dilated urethra and hematometra formation. She was taken for vaginal examination under anaesthesia followed by hematometra drainage and plication of bladder neck. She was further advised on correct placement of vaginal mould regularly under guidance.

Case 4

A 16 year old patient presented with complain of primary amenorrhea and cyclical abdominal pain since 2 years. Patient had and history of hematometra , hematocolpos drainage before 4 months. On per rectum examination, uterus was bulky. USG and MRI suggested hematometra and hypoplastic vagina. Patient was taken for diagnostic laparoscopy which confirmed unicornuate uterus with rudimentary horn. Per vaginal exploration revealed midvaginal septum with bluish bulge. Spetum was punctured with a long needle and blood was aspirated. Thus after cruciate incision, hematocolpos was drained. Cervical os was not seen: instead a small dimple was visualized. Hegar dilation was inserted and collection from uterus was drained. Vaginal mould was inserted and patient was instructed to use it regularly.

Case 5

A 20 year old patient presented with complain of dysmenorrhea. Examination revealed tenderness on lower abdomen. Per rectum examination suggested bulky uterus. USG was advised which showed unicornuate uterus with a collection filled rudimentary horn not communicating with uterus. Patient was taken for laparotomy and rudimentary horn was resected. Patient was advised follow up after 6 months.



Case 6

A 16 year old female was admitted with complain of cyclical abdominal pain. Patient had similar complain before one year for which she had undergone examination under general anesthesia followed by hematometra drainage. USG and MRI was suggestive of uterine didelphys with hematometra in right horn and a normal left horn. Patient was taken for diagnostic hysterolaparoscopy followed by laparotomy for resection of right horn and right cervix.

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Case 7

A 13 year old patient presented with complain of cyclical abdominal pain and primary amenorrhea. On examination, she had a shallow vagina and per rectal examination revealed bulky uterus. MRI suggested hematocolpos with hematometra with possibility of transverse vaginal septum. Patient was taken for vaginal exploration under anaesthesia followed by transverse vaginal septum resection and hematometra drainage. Patient was instructed for vaginal mould placement regularly.

Case 8

A 16 year old female was admitted with complain lower abdominal pain. On examination, left forniceal fullness present. Per rectum examination showed boggy mass over left fornix. MRI showed uterine didelphys with longitudinal vaginal septum and left hematocolpos. Also there was absent left kidney pointing towards OHVIRA diagnosis. Patient was taken for diagnostic laparoscopy followed by left hemihysterectomy and drainage of left hematocolpos followed by vault closure. Patient was advised follow up after 6 months with USG, if any complain.

Case 9

A 17 year old presented with complain of cyclical abdominal pain and scanty menses. On per rectal examination, patient had a 10 week size uterus and bulging over right side. USG and MRI showed bicornuate uterus with longitudinal vaginal septum and hematometra and hematocolpos on right side. Patient underwent per vaginum septum resection followed by drainage. Patient was advised vaginal mould placement regularly.

Case 10

A 30 year old female presented with complain of primary infertility. On examination, there was single cervix with normal size uterus. USG was suggestive of normal findings. On diagnostic hystero-laparoscopy for infertility, there was septate uterus. Patient was then opened for laparotomy followed by septum resection and metroplasty. Per operative IUCD was placed in situ.\

Case 11

A 31 year old female presented with complain of primary infertility since 4 years. Examination as well as investigations had normal findings. USG showed subseptate uterus with normal adnexa. Patient was taken for diagnostic hystero-laparoscopy. Hysteroscopic septum resection was done and patient was advised follow up.

Case 12

A 14 year old female presented with complain of lower abdominal pain. Patient had documented transverse vaginal septum and had undergone drainage multiple times. USG and MRI were suggestive of transeverse vaginal septum with hematometra and hematocolpos; normal bilateral kidneys. Patient was taken for vaginal septum resection and hematometra, hematocolpos drainage followed by double breasting of vaginal mucosa. Patient was advised regular placement of vaginal mould.

Case number	Clinical presentation	Anomaly	Management	Asrm classification	ESHRE
Case 1	Hypomenorrhea	Uterine didelphys with hematometra hematocolpos oblique vaginal septum	EUA resection of oblique vaginal septum		U3cC2V2
Case 2	Rudimentary horn ectopic pregnancy with backache	Unicornuate uterus with right rudimentary	Rudimentary horn excision	U4aU	U4aC0V0

		horn			
Case 3	Primary amenorrhea	Transverse vaginal septum with hypoplastic lower vagina	EUA Septum resection double breasting of vaginal mucosa Vaginal mould placement		U0C0 V3
Case 4	Primary amenorrhea with cyclical abdominal pain	Unicornuate uterus with rudimentary horn with transverse vaginal septum	Diagnostic laparoscopy Followed by Resection of transverse vaginal septum Vaginal mould placement		U4aCoV3
Case 5	Dysmenorrhea	Unicornuate uterus with rudimentary horn	Rudimentary horn resection		U4aC0 V0
Case 6	Cyclical abdominal pain	Uterine didelphys	Diagnostic hystero-laproscopy followed by laparotomy resection of right horn and right cervix(hemi-hysterectomy)		U3cC2V0
Case 7	Cyclical abdominal pain with primary amenorrhea	Transverse vaginal septum	Resection of transverse vaginal septum Vaginal mould placement		U0C0V3
Case 8	Lower abdominal pain	Uterine didelphys with left hematocolpos longitudinal vaginal septum Absent left kidney	Diagnostic laparoscopy followed resection of left horn and left cervix(hemi-hysterectomy) With drainage of hematocolpos followed by vault closure		U3cC2V2
Case 9	Cyclical abdominal pain and scanty menses	Bicornuate uterus Longitudinal vaginal septum Hematometra Hematocolpos	Resection of longitudinal vaginal septum		U3bC0V2
Case 10	Primary infertility	Septate uterus	Diagnostic hystero-laproscopy followed by Laparotomy for		U2bC0V0

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			septum resection and metroplasty with inert intrauterine device placement		
Case 11	Primary infertility	Subseptate uterus	Diagnostic hystero-laproscopy followed by hysteroscopic septum resection		U2aC0V0
Case 12	Lower abdominal pain	Transverse vaginal septum with hematometra hematocolpos	Resection of transverse vaginal septum and double breasting of vaginal mucosa Vaginal mould placement		U0C0V3

Age of diagnosis	**Number of cases**	**Percentage**
<15 years	4	33.33%
15-20 years	6	50%
>20 years	2	16.67%

Presentation		
Hypomenorrhea	1	7.14%
Primary amenorrhea	3	21.43%
Cyclical abdominal pain	5	35.71%
infertility	2	14.28%
Others	1	7.14%

III. DISCUSSION

Müllerian duct abnormalities (MDAs) are a broad and complex spectrum of defects that are often associated with primary amenorrhea, infertility, endometriosis, and obstetric complications. Women with congenital uterine malformation usually experience a higher incidence of complications during pregnancy and delivery. An obstetrician should have a high index of suspicion for an undiagnosed congenital defect such as MDA in the presence of infertility with a background of maternal congenital malformations. Early diagnosis and recognition of the condition may allow proper planning of treatment to ensure a favorable obstetric outcome.

Congenital anomalies of the female reproductive tract may involve the uterus, cervix, fallopian tubes or vagina. Uterine anomalies are the most common mullerian anomalies but the true incidence is not known as many women are asymptomatic and sensitive imaging modalities have only recently become available. Most cases are diagnosed during evaluation for obstetric or gynaecological problems but in the absence of symptoms, most anomalies remain undiagnosed. Reported population prevalence rates have varied between 0.06% and 38%. As nearly 47% of women with uterine defects have successful fertility and pregnancy, the true incidence of congenital mullerian defects is significantly understated. Mullerian anomalies are known to be associated with renal anomalies in 30-50% of cases and defects include renal agenesis, severe renal hypoplasia, ectopic or duplicate ureters.

This case series comprises of 12 cases of Mullerian anomalies, diagnosed through clinical history, physical examination, radiological scans or surgery. The case series is multiparametric in design and focuses on multiple parameters of an individual case. Parameters like age, clinical presentation, hormonal profile, Ultrasonography (USG), Magnetic resonance imaging (MRI),

Computed tomography (CT) scan, IVP (Intravenous pyelogram) findings & mode of treatment were analyzed. Through the present case series we aim to evaluate the various clinical presentations of the Mullerian duct anomalies, classify them as per ESHRE classification system, assess the other congenital anomalies associated with them and also study the optimum mode of management used for the respective cases.

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