



Mullerian Malformations and Reconstructive Surgery: Clinicians' Approach

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Abstract

Mullerian malformations are not uncommon. Overall incidence ranges widely between 1 in 200 women and 4 in 100 women (Chan et al. in Hum Reprod Update 17:761–771, 2011; Grimbizis et al. in Hum Reprod 28:2032–2044, 2013). Other way, these observations suggest presently an increased number of Mullerian abnormalities are diagnosed with more details, following the use of newer diagnostic modalities. Most classifications that are available have limitations. Diagnosis was based on imaging studies that had low diagnostic accuracy. It was focused mainly on the anomalies of the uterus. Less is known about the anomalies of the cervix or the vagina in isolation or in combination with the utero-cervical and vaginal malformations. Improved diagnostic modalities and incorporation of assisted reproductive technology have improved the outcome further. Therefore, a more expanded classification needs to be introduced with wider criteria for the clinicians. This will make clinicians' approach for the management more simple.

Keywords Mullerian malformations \cdot Clinical presentations \cdot Diagnosis \cdot Reconstructive surgery \cdot Menstrual \cdot Sexual and reproductive functions

Summary

Development of the Mullerian system is a complex process that occurs over a period between 5 and 15 weeks of embryonic life. Development and progressive differentiation of the two Mullerian ducts (mesodermal) are under the influence of many factors (molecular, genetic, chromosomal, hormonal

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and environmental). Paramesonephric ducts developments are in continuum with the development of urogenital sinus (urinary bladder), urorectal septum, ureteric buds, cloacal membrane and the endodermal cloaca. Maldevelopment of the Mullerian system thus leads to the non-function or dysfunction of other organs too. Successful development of the Mullerian system is essential for normal menstrual function, sexual function and finally the reproductive function. A wide range of malformations have been observed affecting the genital organs either in isolation or in combination with other organs. Interestingly in the present days, major changes have been observed in all the areas of management of Mullerian abnormalities. Microarray technology can provide insight into the genomic expression of developmental control. Newer diagnostic modalities provide details of structural information for any type of Mullerian malformation. More and more reconstructive and organ-sparing surgery is now possible to restore the menstrual as well as the sexual function. Endoscopy is used as an alternative method of open surgery with all its advantages. Presently, there is remarkable development in the field of reproductive endocrinology and assisted reproductive technology. Combined together, a significant change is observed in the management outcome of patients with Mullerian malformations. There is a need to update our understanding in the management of this problem based on current evidences [1, 2].

Development of Mullerian System

Male and female reproductive tracts were described by Wolf in 1760 and Muller in 1830, respectively [3, 4]. The generic terms in common usages are "mesonephric" or Wolffian and paramesonephric or "Mullerian" ducts. The ducts appear between 5 and 15 weeks of embryonic life. Theories of sexual differential towards the male confirm the presence of SRY gene on the Y chromosome, testicular determining facts (TDF) to favour gonadal cortical regression and anti-Mullerian hormone to cause regression of the Mullerian ducts. Otherwise, absence of these factors favours the development of female gonads and Mullerian ducts with regression of the Wolffian system. This explanation is apparently more simple to accept. However, the role of other genes or molecules like HOXA, SOX9, DAX1, WT1, SF1 is too complex to understand. Craniocaudal expression of HOXA genes along the Mullerian ducts directs the development of the female reproductive organs. Commonly involved HOXA genes are: HOXA-9, 10, 11 and 13. Congenital anomalies of the reproductive tract are thought to be associated with the genetic and molecular abnormalities. Variable expression of genetic defect may be precipitated by teratogenic exposure. Absence of anti-Mullerian hormone or oestrogen receptors in the Mullerian ducts is also implicated. Till date, it is accepted that Mullerian malformations are multifactorial, polygenic and familial.

Clinical Scenario

Data regarding the prevalence of Mullerian malformations vary considerably. The reasons are: (a) there is lack of consensus in the criteria to define the anomalies, (b) imaging methods used earlier for the diagnosis had low accuracy, and (c) newer modalities of diagnosis with higher accuracy not only make the diagnosis more but also with specific type.

Approximately 25–30% of women with Mullerian anomalies have impaired reproductive behaviour. Overall incidence of miscarriage is about 10–15% of all clinical pregnancies. Most studies [5] support that arcuate and didelphic form of anomalies does not contribute to recurrent pregnancy loss (RPL), whereas septate, unicornuate and bicornuate anomalies do.

Structural abnormalities of the uterus have been observed in 0.1 to 4% of all women. Uterine structural anomalies may cause infertility, repeated pregnancy loss, malpresentation, fetal growth restriction or preterm births. Presentations vary depending on the type and severity of the malformation.

At the same time, improved outcome has been observed with newer modalities of therapy including MIS. Therefore, the syndrome needs to be redefined with wider criteria, based on the newer diagnostic modalities, along with the current therapeutic approaches.

Classification of Mullerian System

Many classification systems for Mullerian abnormalities have been suggested since Kauffmann's description in 1930s [6]. The importance of a classification system is manifold. It needs to define (a) a specific anomaly in isolation or in combination, (b) its impact on menstrual, sexual or reproductive function, (c) involvement of any other system anomaly in association, (d) specific intervention needed for diagnosis and management and ultimately, and (e) the expected outcome benefits obtained, in terms of menstruation, sexual function and reproduction.

Classification systems of Mullerian malformations underwent successive modifications [7, 8]. Unfortunately to meet the universal agreement in any system of classification is absent. Reality is that malformation of an organ system does not follow any specific rule. Abnormalities of Mullerian systems may involve the uterus, cervix, vagina and the tubes (Fig. 1). Anomalies may be in isolation or combination with other parts as in cervico-vaginal agenesis. Often other system anomalies (renal) are associated. Diagnosis and therapeutic benefits are Mullerian anomalies are aimed with successful restoration of the following areas:

- (a) Near normal anatomy
- (b) Normal sexual function
- (c) Cyclic menstrual function
- (d) Successful reproductive function

Amidst complexities of classifications, a simple approach to view the Mullerian abnormalities is based on the normal embryological understanding. Important area of fusion is that of united lower vertical parts the Mullerian ducts



Fig. 1 Laparotomy showing uterine knobs (rudimentary) one on either side. Normal ovaries are seen (arrow)

(Mullerian eminence) with the dorsal wall of the urogenital sinus (sinovaginal bulb). The disintegration of the central cells of Mullerian eminence allows vaginal canal to open into the urogenital sinus. Ureteric buds develop on the dorsal wall of the urogenital sinus and to meet the metanephric kidney. It is not uncommon to see the anomalies related to urorectal septum and the cloaca. Congenital vesico-vaginal fistula occurs in women when Mullerian eminence ruptures into the vesico-urethral part of the cloaca instead of urogenital sinus. Based on the sequence of Mullerian duct differentiation and fusion (lateral and vertical) along with that of urogenital sinus differentiation, clinicians need a classification of Mullerian anomalies. Mullerian maldevelopment



Group A (a) Complete agenesis, (MRKH syndrome)

may affect the function of all the following organs: uterus, cervix, vagina, parts of cloaca (urinary bladder and the rectum) fallopian tubes and the renal system.

Classification of Mullerian Anomalies Based on Therapeutic Approach

Group A: Agenesis: Absent uterus

- (a) Complete agenesis (MRKH syndrome)
- (b) Unilateral agenesis (unicornuate uterus)



Group B:

Mullerian abnormalities with the problems of vertical fusion (canalization): Functioning uterus:

- (a) Obstructive (cervico-vaginal agenesis)
- (b) Non-obstructive (communicating horn)



Group B (a) Cervicovaginal agenesis

Group B (b) Nonobstructive

Group C:

Mullerian abnormalities with the problems of lateral fusion: Functioning uterus:

- (a) Obstructive (non-communicating horn);
- (b) Non-obstructive (septate, bicornuate uterus)

rectovesical ligament (Fig. 2b). Surprisingly, the obstetric outcome is uneventful, in the majority (Fig. 2a–c). Management approach for a clinician is difficult when the anomaly is complex with the involvement of two or more segments (Mullerian, cloacal, vesicourethral or anorectal).



Group D:

Rare abnormalities: Complete/segmental absence of fallopian tubes, congenital vesico-vaginal fistula, rectovaginal fistula, (cloacal dystrophy), narrow introitus with ectopia vesicae, (exostrophy of the bladder). The new classification system [9] has been described as simple, clear, accurate, comprehensive, user friendly and correlates with patient's presentation. Uterus didelphys is a distinct anomaly due to the complete failure of lateral fusion. Uterine body, cervix and the vagina are double with the presence of the



Group D Segmental absence of fallopian tube



Fig. 2 a Uterus didelphys, seen following caesarean delivery, viewed from the anterior surface. Left gravid horn; right non-gravid horn. Caesarean wound is seen on lower segment of the uterus, the left gravid horn (arrow). **b** The same, uterus didelphys as seen above.

View from the posterior surface. Left is the gravid horn. Rectovesical ligament is seen (arrow). \mathbf{c} The same patient with uterus didelphys (as above). The two cervices are seen from below

Unilateral obstruction is almost always associated with agenesis of the ipsilateral kidney. Bilateral obstruction with bilateral kidney agenesis is associated with non-viable embryo.

Obstructive lesions need early diagnosis and early interventions. This is to prevent deterioration of organ function due to retained blood or the back flow of menstrual blood in the peritoneal cavity to develop pelvic endometriosis.

Diagnosis of Mullerian Abnormalities

Diagnosis is predominantly made on clinical history and examination. X-ray and hysterosalpingography have limitations. 2-D ultrasound (US) provides reliable, objective and measurable information for the anatomy of the genital organs. Hysterosalpingo-contrast sonography (HYS CO SY) focuses mainly on the intracavitary abnormalities. It has some added benefits over the 2–D US.

3-D US is highly reliable. Geometric measurements of the uterus in different planes (sagittal, coronal, axial) and line drawings (interostial, fundal) can detect the arcuate, septate, bicornuate and didelphic uterus. 3-D image acquisition and post-processing is essential for the diagnosis of the complex anomalies. For 3-D US, it is implied that sonologist has the special training, experience and adequate background knowledge of the uro-genital system. Even a 3-D US diagnosis needs confirmation with endoscopic evaluation before any surgical intervention, as most noninvasive methods are examiner dependent.

MRI gives highly reliable and objective information. The author feels gynaecologists should be trained to read the MRI images and work closely with the radiologist. Endoscopy (laparo-hysteroscopy) is suitable for more complex anomalies and also for other associated pelvic pathology. In India, most gynaecologists are conversant with laparohysteroscopy procedure and prefer to go for confirmation of diagnosis and the management. A prior US or MRI knowledge is helpful though. Adolescents are much benefitted with use of 2-D US, 3-D US, MRI or endoscopic procedures either alone or in combination for the management.

Vaginal agenesis and rudimentary uterus or presence of uterine knobs commonly goes by the names of persons whose work has enriched the knowledge with this problem. Realdus Columbus (1559) first described congenital absence of the vagina. Mayer (1829) described the congenital absence of vagina with a rudimentary uterus in still born infants. Rokitansky (1838) and Kustner (1910) described an entity of congenital absence of vagina, rudimentary uterus, and normal ovaries with renal and skeletal abnormalities. Hauser (1973) described the frequency of the disorder and the full spectrum of the abnormalities. The terminology Mayer Rokitansky-Kuster syndrome and vaginal agenesis are commonly used. Overall incidence of vaginal agenesis varies from 1 in 4000 to 1 in 10,000 female births. MRKH syndrome is considered as the second common cause of primary amenorrhoea in an adolescent girl next to gonadal dysgenesis (45X0/45X0/XX).

Clinical Presentation

Familial clustering has been observed. Vaginal agenesis is associated with amenorrhoea in almost all of the patients. However, a broad spectrum of other variable anomalies is also observed. In one series, the presence of uterine knobs (25%), partial vaginal agenesis (1%) and hematometra (7%) has been mentioned [10]. Author has the experience of seeing such cases with the presence of uterus or uterine knobs having islands of endometrium with variable extent of vaginal agenesis. Imaging studies (MRI) and laparoscopic evolution are of definite value to these women. Prognostically, these are the favourable cases where reconstructive surgery can establish her menstrual and sexual function, if not the



Fig. 3 Case of congenital cervico-vaginal agenesis. Uterus is bulky due to hematometra. Uterus ends abruptly with the absence of the cervix

reproductive function (Fig. 3). Ovaries are normal both histologically and functionally in all the cases with MRKH syndrome. These girls may present with history of cyclic pelvic pain (20%) with development of hematometra.

Associated anomalies are common (40%). The author had the experience of seeing patients with multiple anomalies including renal (pelvic kidney) and skeletal (scoliosis, fused vertebrae) system (Fig. 4). Common abnormalities observed are: pelvic kidney, horseshoe kidney, agenesis of ipsilateral kidney in cases with unicornuate uterus, along with vertebral scoliosis, fused lumbosacral vertebra and segmental loss of fallopian tube. Occasionally, Turner's features have been observed with MRKH syndrome [11]. These days, more abnormalities are observed to be associated with subsequent investigations including cervical, spinal dysplasia, tetralogy of Fallot, ventricular septal defects, neurosensory hearing loss cloacal dysplasia and ectopia vesicae. Mullerian abnormalities have been observed as a component of multiple malformation syndrome. The hand-foot-genital syndrome is an autosomal dominant disorder. It is manifested with skeletal abnormalities of the hands and feet (Fig. 5). The individual presents with small feet, short great toes, and abnormal thumbs. The associated Mullerian anomalies in the hand-foot-genital syndrome are bicornuate uterus and septate uterus [12]. Therefore, the syndrome needs to be redefined with wider criteria as being observed by many subsequent workers.

Management of Vaginal Agenesis

The principle of management is to create neovagina with a depth of 6-8 cm which is adequate for sexual function. Sexual and menstrual function may be restored. Reproductive



Fig. 4 Multiple skeletal malformations showing kypho-scoliosis of lumbosacral vertebrae, fused lumbosacral vertebrae and pelvic bone deformity

function in the absence of a functioning uterus cannot be achieved. Surrogacy is the option for achieving a genetic motherhood. Delivery of a viable baby has been reported following uterine transplantation in a woman with MRKH syndrome. What is more important is to select a case for creation of neovagina. Potential complications of the procedure need to be explained. Uterine transplantation [13] or surrogacy for reproduction is thought to be the alternative. However, creation of neovagina is a prerequisite for restoration of complete reproductive function.

Surgical procedures that are commonly performed for neovagina creation are many. McIndoe-Reed procedures (1938) are to create a space between the bladder in front and rectum behind. Split thickness skin graft is used over a mould. Wharton (1938) placed a condom-covered mould instead of skin graft in the neovaginal space. William's vulvo-vaginoplasty (1976) is to create a vaginal pouch from the skin flaps of labia majora in the midline. Author's experience of vaginoplasty with or without amnion graft appears to be more simple. Space is created between the bladder and the rectum extending up to the peritoneum of the POD. A mould is placed in the space for a variable period of time during the post-operative phase. Follow-up is maintained



Fig. 5 17-year-old girl with hand-foot-genital syndrome (multiple skeletal malformations) with cervico vaginal anomalies

till sexual life is established. Regular use of amnion graft is not mandatory.

Currently, it is being done laparoscopically. Following creation of neovagina, pelvic peritoneum is dissected off laparoscopycally. It is used as a graft for the neovagina so created. The neovaginal vault is closed with a purse string stitch. Vecchietti and Devydov's procedure is a laparoscopic method of neovagina creation. Vecchietti approach entails the risk of bladder or rectal injury where wire negotiation is done.

Functional success of patients following vaginoplasty, with long-term follow-up, has been found to have satisfactory sexual function in majority (90%) [14]. MRKH syndrome when seen in the outpatient department needs to be differentiated from other conditions like (a) labial agglutination, (b) imperforate hymen and (c) incomplete variety of testicular feminization syndrome. Each case needs clinical examination and imaging studies. Karyotyping may be needed in some cases to confirm the diagnosis.

Mullerian anomalies: Functioning uterus with complete outflow tract obstruction (failure of vertical fusion).

Obstruction of the outflow tract may be at any level below the body of the uterus. Agenesis of cervix is rare, and congenital cervico-vaginal agenesis is extremely rare (Fig. 3). Management option has changed from hysterectomy to conservative surgery in these days with the availability of newer modalities of imaging studies. Management approach has improved further with surgery including laparoscopy and ART.

Women with functioning uterus with complete outflow tract obstruction may have three subtypes. There may be agenesis of (a) cervix or (b) vagina or (c) combined cervix and vagina or only with a (d) transverse vaginal septum. These patients present with primary amenorrhoea (cryptomenorrhoea) during adolescence. This group needs early intervention for reconstructive surgery; otherwise, they often lose their fertility potential.

The uterus may be normal and/or at times malformed (bicornuate or unicornuate). The incidence of complete cervico-vaginal agenesis is unknown. It is based on isolated case reports only. Diagnosis of this abnormality is difficult. Majority of the patients have cyclic abdominal pain, presence of hematometra and/or endometriosis. High transverse vaginal septum and cervical agenesis need to be differentiated when the vagina is patent.

Cervical agenesis has been described differently by different workers. In reality, anomalies do not follow any rule. It is clinically divided into two main groups depending upon the presence or absence of cervical stroma. At the same time, the presence or absence of the vagina. Women with presence of cervical stroma and patent vagina have the best outcome in terms of restoration of menstrual and reproductive function. Author has the experience of seeing such cases mostly associated with vaginal agenesis. It is mainly due to agenesis of the combined Mullerian ducts and/or their differentiation and failure of fusion or canalization.

Management

Review of literature through the 1960s and 1970s revealed that hysterectomy is the only option for congenital absence of the cervix. The main concern is the infection. Presently, most of us feel the option for hysterectomy should be deferred until appropriate evaluation has been made. Consideration may be given for creating a cervico-vaginal fistula. A variety of methods have been described based on individual team expertise and resources to create a cervicovaginal fistula tract between the lower uterine cavities with the upper part of the neovagina. Post-operative management is equally essential to maintain the patency of the tract. Currently, we are placing a polyethylene stent (tube) in the tract to maintain the patency of the canal. Introduction of assisted reproductive technologies has prompted more of *conservative and 'organ-sparing'* approach. Good prognosis has been reported following ART (ZIFT) as well as open surgery and IVF-ET [15]. Stent has been used to maintain the patency of the fistula tract for variable period 2-4 months, though 2 weeks time may be adequate in majority of the cases. Hysterectomy may be needed in cases with prior repeated failed attempts of developing a fistula tract or to maintain it. In such a case, gestational surrogacy is the option. Author's experience of managing such cases is the combined abdomino-perineal approach. Successful pregnancies have been reported following reconstructive surgery of patients with congenital cervico–vaginal agenesis [15].

Mullerian Abnormalities with Failure of Vertical Fusion (Obstructive)

Transverse vaginal septum is due to failure of vertical fusion. No reliable data are available regarding the incidence of the transverse vaginal septum. Commonly, it is present in upper third of the vagina. However, it may be observed in mid- or lower third of vagina. Patient often presents after puberty with cyclic lower abdominal pain, central lower abdominal and pelvic mass. Patient suffers amenorrhoea (cryptomenorrhoea). Sometimes, a small opening may be present in the centre of the septum. There may be the presence of haematosalpinx, hemoperitoneum and pelvic endometriosis. Rare cases may be associated with upper vaginal or cervical agenesis (Fig. 6).

Management consists of assessment of hematometra or the hematocolpos with imaging studies. A transverse incision is made on the septum, and a probe is introduced



Catheter in situ

Fig.6 Upper vaginal and cervical agenesis. Dissection was done through the vaginal route. Collected blood was allowed to drain. Vaginal mould was used to prevent restenosis

through the septum. Dissection of the septum is guided by palpating the catheter anteriorly, and a double-gloved finger placed in the rectum. When the dissection is continued upwards, cervix is reached. The margins of the septum are excised. A vaginal mould can be placed for few days to prevent restenosis.

Mullerian Abnormalities with Failure of Development of One Duct

Unicornuate uterus is a very rare type of anomaly. It is due to the absence of development or hypoplasia of one Mullerian duct. It may present with or without a rudimentary horn; this rudimentary horn may or may not be communicating to the contralateral uterine cavity. Moreover, the rudimentary horn may or may not have a cavity. Lining of endometrium in the cavity of the rudimentary horn may or may not be present. Reproductive outcomes in a patient with unicornuate uterus are often poor. Repeated miscarriages, preterm births and cervical incompetence are common. Author has the experience of managing one such a rare case with long-term hospital admission ending planned preterm delivery with satisfactory maternal and perinatal outcome. In another case, author had to face difficulties in the management when a ruptured rudimentary horn pregnancy was encountered as an emergency [16]. Ectopic pregnancy can occur in a rudimentary horn even if the rudimentary horn is non-communicating. This is due to transperitoneal migration of sperm. Rudimentary horn needs to be excised, if it is non-communicating and the endometrium is present. When the horn is communicating, but is small, it should be removed.

Problems of infertility, in such a patient, could be overcome with ovulation induction or with ART, as indicated. Gestational surrogacy is an option for women with repeated pregnancy loss due to reduced uterine cavity space.

Mullerian Abnormalities Due to Failure of Lateral Fusion (Non-obstructive)

Bicornuate uterus develops when the two Mullerian ducts remain separated at their cranial ends but are fused at their caudal ends. In this case, there are two horns with an indentation in between the two unfused horns. Imaging studies show the V-shaped indentation of the external contour of the fundus. It should be kept in mind that failure of fusion of the two ducts may occur at any point and for any distance in this process. Patients with bicornuate uterus often suffer infertility, recurrent pregnancy loss, preterm labour or malpesentation. However, it is not uncommon to see a woman with bicornuate uterus with an uneventful reproductive behaviour only to be detected during hysterectomy because of heavy menstrual bleeding and dysmenorrhoea.

Women with bicornuate uterus may need metroplasty operation for unification of the two horns. Commonly performed operation is Strassmann unification. Unification operation can be performed laparoscopically also with its advantages [17]. Successful pregnancies have been reported in all the procedures of metroplasty.

Septate uterus develops when the two Mullerian ducts fuse completely, but the tissue between the two ducts is not resorbed partly. A complete septum extends from the fundus up to the internal cervical os. Management of cases with septate uterus has been much simplified these days. Hysteroscopic resection of the septum is done by using scissors, or by a resectoscope. Ablation of the septum may be done using laser fibres or electro-coagulation. Successful pregnancies have been observed in 70–95% of cases. Term delivery rate after hysteroscopic metroplasty in patients with prior recurrent miscarriages has been reported [18].

Uterus didelphys develops when the two Mullerian ducts fail to fuse and each duct gives rise to a separate uterine horn with its own cervix and vagina. Usually, uterus didelphys does not need any treatment. However, metroplasty has been reported similar to bicornuate uterus, for patients with RPL, where no other cause has been detected [18]. Author has the experience of managing such patients ending successful pregnancy with conservative management (Fig. 2a–c).

Others

Arcuate Uterus

There is slight indentation over the fundus in its external architecture. Some consider arcuate uterus as the mild form of bicornuate uterus. However, the most consider it as a variation of normal uterine morphology. The effect of arcuate uterus on reproductive outcome is controversial. Metroplasty for arcuate uterus is rarely recommended.

Mullerian Development and Diethylstilbestrol (DES)

Uterine body becomes deformed. The cavity appears T-shaped and reduced in size. The DES-related anomaly is not discussed here as DES is discontinued after 1971 [9].

Conclusion

Mullerian system malformations are not uncommon. A wide range of malformations are observed either in isolation or in combination. All these may affect menstrual function, sexual function and/or the reproductive function adversely. Significant changes have undergone in the areas of diagnosis. It is possible to detect the type and extent of the abnormality (ies), precisely. Organ-sparing reconstructive surgery is possible with the use of open and/or endoscopic surgery. Contribution of assisted reproductive technology in the management is enormous. Most women can have successful reproductive out come with their own genetic material. There is a need to update the understanding in the management based on the progress of knowledge and the current evidences.

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Compliance with Ethical Standards

Conflict of interest The author declares that he has no conflict of interest.

Ethical Standards All procedures followed were in accordance with the ethical standards of the responsible committee on the human participants (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Informed Consent Informed consent was obtained from the eligible women. Additional informed consent was obtained from all individual participants for whom identifying information is included in this study.

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