The Rare Cases of Mullerian Duct Anomalies and Pregnancy Losses

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I. OVERVIEW

Mullerian anomalies (MDLs) refer to congenital abnormalities in which the Mullerian (paramesonephric) ducts do not develop properly. It could be due to complete agenesis or defective vertical or lateral fuse, or resorption failure.

The mullerian system developmental anomalies are some of the most interesting disorders that gynecologists and obstetricians come across. The primordial anlage of the female reproductive system is the mullerian. They divide to form the fallopian tubes and uterus. This system can lead to a variety of malformations. These malformations can range from uterine or vaginal agenesis, duplication of the vagina and uterus, to minor uterine cavities abnormalities and duplication of some uterine structures. Mullerian malformations often occur in association with abnormalities of the axial and renal skeletal systems. They are often first seen when patients are examined for other conditions. Mullerian duct anomalies (MDAs), in most cases, are associated with functioning ovaries or age-appropriate external genitalia. These abnormalities can be recognized as soon as puberty begins.

II. PREVELANCE-

The existence of mullerian defect dates back to antiquity around 300 BC. Columbo was the first to report a case of vaginal birth (uterus and vagina), in the 16th century. Uterine abnormalities often go unreported and are not recognized at birth. Reproductive malfunctions are often reported more frequently during childbearing years.

The study will determine the incidence rate. Incidence rates range from 0.1-3.5% according to most authors. The incidence of mullerian anomalies in women with fertility issues is slightly higher at 36%. Recurrent abortions are more common than 5-10% in women. Patients who have had third-trimester miscarriages are most likely to experience mullerian defects. [4,10] Septate, arcuate and didelphys are the most common mullerian-duct anomalies. The study and the geographical location will determine the exact distribution.

Reports varying from 0.16 to 10% indicate that mullerian anomalies are also common. [12, 13, 14, 15, 16, 17, 18] It has been found that 8-10% of women who undergo hysterosalpingography (HSG) with recurrent pregnancy loss have mullerian anomalies. [13 15.] This is in contrast to a prevalence rate of 2-3% among elective hysteroscopy patients, who are thought to be more representative of the general population. [16] A Danish study of 622 women aged 20-74 years, all of whom were subject to saline contrast sonohysterography revealed a 9.8% prevalence of mullerian anomalies in the general populace. This was especially true for nulliparous and oligomenorrhea.

III. EMBRYOLOGY

A pair of Mullerian tubes forms the female reproductive tract. These structures inude the fallopian tube and uterus. The embryological origins of the ovaries and the lower third of the vagina are different. They were formed from germ cells that migrate from both the primitive yolk sac or the sinovaginal bulbs, respectively.

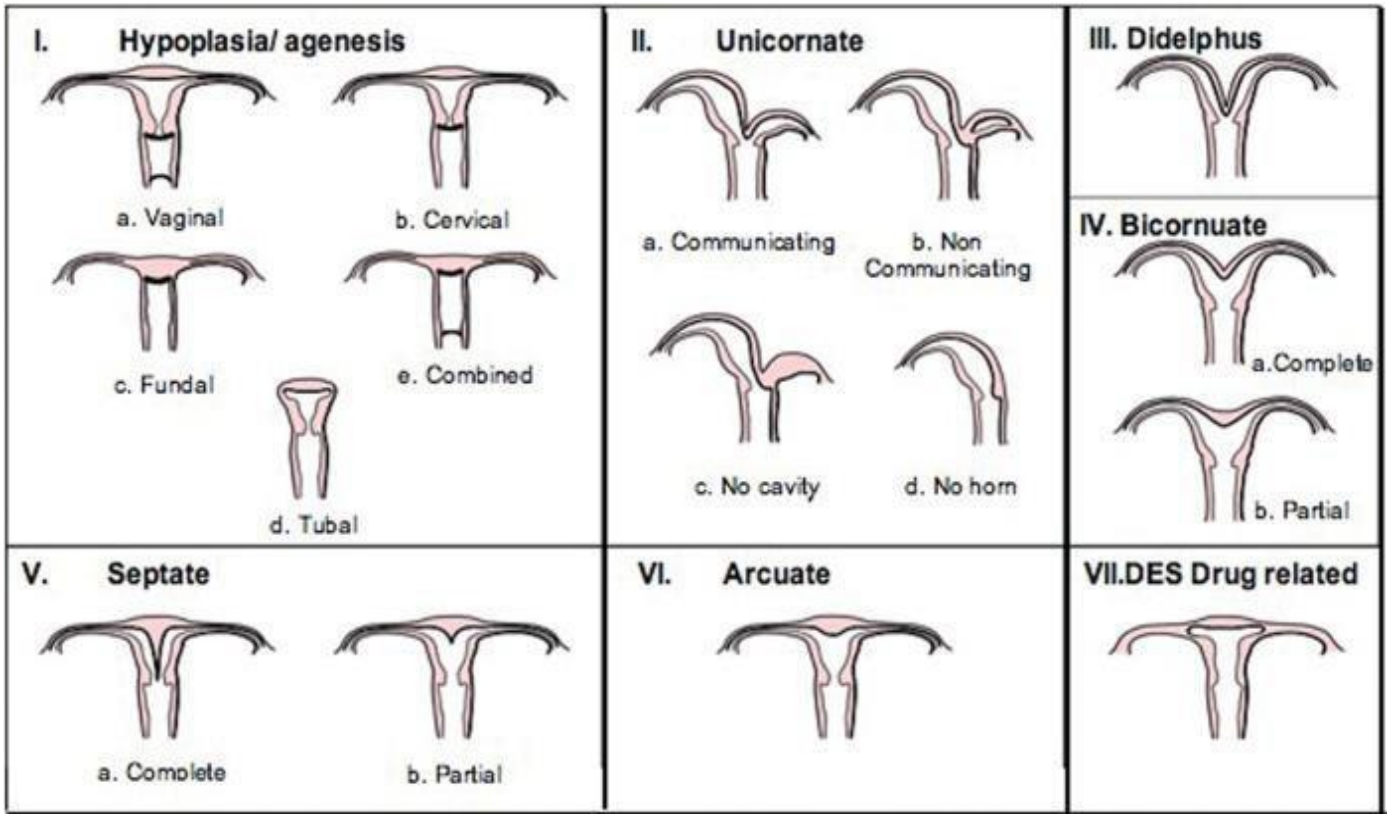
The normal development of Mullerian ducts is dependent on completion of three phases: organogenesis (fusion), and septal resorption. The formation of both Mullerian conduits is an indication that organogenesis has occurred. This can lead to uterine hypoplasia/uterine agenesis, or a unicornuate-uterus. Fusion refers to the fusion of the ducts that form the uterus.

This can lead to a didelphys or bicornuate. After the ducts have fused, septal resorption is the subsequent resorption. A septate or arcuate-uterus is formed when there are defects at this stage.

IV. CLASSIFICATION-

Because of the potential for poor pregnancy outcomes and treatment, it is crucial to properly classify MDA [1]. The American Society of Reproductive Medicine is the most commonly used classification system.

[1] (Figure 1.). This classification system is only a guideline. Not all anomalies will fit into any of the categories. It is important to describe each component of a case accurately in cases that don't fit within a particular category.



[FIGURE1- CLASSIFICATION OF MDA ACCORDING TO AMERICAN SOCIETY OF REPRODUCTIVE MEDICINE]

V. SURGICAL MANAGEMENT OF DEFECTS-

Each mullerian anomaly in the mullerian tube is described and organized according its AFS classification.

Class I - Vaginal Agenesis

Both surgical and nonsurgical treatment options have been tried. Gradually dilating devices are used to create a new vagina. It may take several months, or even years for a functional vagina to form. Surgery is the best treatment option for mullerian dysplasia.

It is crucial to choose the right time for a vaginoplasty. Only consider surgical treatment if the patient is able to participate in the decision-making process and is motivated to continue using a vaginal prosthesis for at least several months.

Class II - Unicornuate Uterus

Unicornuate uterus is rare. This is evident in the lack of data in literature on surgical strategies, particularly for subtypes. The development of specific surgical management plans has not been significant. Reconstruction metroplasty is not recommended for women with unicornuate uterus. [6, 41, 25,] Endometrium found in the accessory horn is a sign that surgery should be performed. Laparoscopic hemihysterectomy is the best option. [20, 21] When the rudimentary endometrium is absent, surgical treatment is not recommended.

III - Uterus Didelphys

An indication to have the vaginal septum resected is an obstructed unilateral vulvum. To preserve reproductive ability and prevent damage to the uterus or tubes, surgery is required [9,17].

Surgical techniques-

- Uterine didelphys with obstructed unilateral vagina

  - The preferred method is to complete excision and marsupialization the vaginal septum. This is done as one procedure. Laparoscopy is possible to treat any adhesions or endometriosis that may be associated with the septum after it has been removed. [16, 37, 38]
  - To prevent bleeding from the vaginal mucosa, it is necessary to leave a large pedicle after removing an obstructed septum. [41, 32]
  - For the best chance of successful reproduction, neither a hemi-hysterectomy nor a salpingo-oophorectomy is recommended.
- Uterus didelphys, nonobstructed

  - There are very few indications for septum resection of the nonobstructed didelphys. These patients are not suitable for surgical unification. These patients are not likely to experience fertility-related problems. Obstetric complications can be minimal if the woman has a full-term pregnancy.
  - Metroplasty is a decision that must be made individually. Only selected patients may be eligible for surgical reconstruction. The benefits of surgery in this setting are unclear as most reports are speculative.
  - The Strassmann mesotherapy is recommended. [5] This procedure unites the uterine cavities at their fundus while leaving the cervices intact. Below is a detailed description of this procedure: Surgical techniques to bicornuate the uterus.

CLASS IV - BICORNUATE UTERUS

Rarely does a bicornuate uterus require surgical reconstruction.[14] Metroplasty's benefits have not been evaluated in prospective trials using data from observational studies.[12,35] Women who have had recurrent spontaneous abortions, birth defects, or midtrimester losses should not be considered for metroplasty.

There are many options for metroplasty, but the Strassmann method is the best option to unify the bicornuate & didelphys.

In this situation, transcervical lysis is not recommended for other anomalies. It can cause uterine perforation.[14] The Strassmann procedure involves the removal of the septum via wedge resection and subsequent unification.

CLASS V - SEPTATE UTERUS

Hysteroscopic metroplasty combined with concurrent laparoscopy is the preferred surgical procedure. [17, 14 and 37] Laparoscopy reduces the chance of uterine perforation after septal incision. [18] An alternative to hysteroscopic mesoplasty is transcervical ultrasound guidance.

- Laparoscopy

  - Double-puncture is used. A probe is passed through the inferior incision. To keep the uterine fundus visible, the probe is used.
  - An examination of the uterus may be helpful in confirming the septate uterus' concave exterior shape. This will help to distinguish it from a bicornuate. It can also be used to detect unexpected pelvic diseases.

- Hysteroscopy

  - The laparoscope and hysteroscopy begin. The cervix is dilapidated to 6 mm and the hysteroscope is inserted at the level of the external Os. Under direct vision, it is advanced into the cavity of the uterine lining. The instruments used to remove the septum will determine the distention media.
  - You can perform a hysteroscopic operation using microscissors or electrosurgery.

CLASS VI - ARCUATE UTERUS

Arcuate uterus can be managed in a similar manner to septate. Only patients with poor reproductive performance are eligible for surgery correction.

VI. CONCLUSION-

Mullerian anomalies, a group of developmental disorders affecting the female reproductive tract, are morphologically varied. It is important to make a diagnosis in order to plan treatment and manage strategies. Mullerian duct anomalies can be corrected using a surgical procedure that is specific to each type of malformation. This may differ for certain groups. The critical indicator of the surgical

procedure's worth is the ability of the patient to have healthy sexual relations after the procedure and to achieve successful reproductive outcomes.

REFERENCES

[1] Litta P, Pozzan C, Merlin F, Sacco G, Saccardi C, Ambrosini G. Hysteroscopic metroplasty under laparoscopic guidance in infertile women with septate uteri: follow-up of reproductive outcome. J Reprod Med. 2004 Apr. 49(4):274-8.

[2] Fedele L, Bianchi S. Hysteroscopic metroplasty for septate uterus. Obstet Gynecol Clin North Am. 1995 Sep. 22(3):473-89.

[3] Donnez J, Nisolle M. Hysteroscopic surgery. Curr Opin Obstet Gynecol. 1992 Jun. 4(3):439-46.

[4] Querleu D, Brasme TL, Parmentier D. Ultrasound-guided transcervical metroplasty. Fertil Steril. 1990 Dec. 54(6):995-8.

[5] Parsanezhad ME, Alborzi S, Zarei A, Dehbashi S, Shirazi LG, Rajaeefard A, et al. Hysteroscopic metroplasty of the complete uterine septum, duplicate cervix, and vaginal septum. Fertil Steril. 2006 May. 85(5):1473-7.

[6] Rock JA, Murphy AA, Cooper WH 4th. Resectoscopic techniques for the lysis of a class V: complete uterine septum. Fertil Steril. 1987 Sep. 48(3):495-6.

[7] Fedele L, Bianchi S, Marchini M, Mezzopane R, Di Nola G, Tozzi L. Residual uterine septum of less than 1 cm after hysteroscopic metroplasty does not impair reproductive outcome. Hum Reprod. 1996 Apr. 11(4):727-9.

[8] Fayez JA. Comparison between abdominal and hysteroscopic metroplasty. Obstet Gynecol. 1986 Sep. 68(3):399-403.

[9] March CM, Israel R. Hysteroscopic management of recurrent abortion caused by septate uterus. Am J Obstet Gynecol. 1987 Apr. 156(4):834-42.

[10] Vercellini P, Fedele L, Arcaini L, Rognoni MT, Candiani GB. Value of intrauterine device insertion and estrogen administration after hysteroscopic metroplasty. J Reprod Med. 1989 Jul. 34(7):447-50.

[11] Dabirashrafi H, Mohammad K, Moghadami-Tabrizi N, Zandinejad K, Moghadami-Tabrizi

[12] M. Is estrogen necessary after hysteroscopic incision of the uterine septum?. J Am Assoc Gynecol Laparosc. 1996 Aug. 3(4):623-5.

[13] Barbot J. Hysteroscopy and hysterography. Obstet Gynecol Clin North Am. 1995 Sep. 22(3):591-603.

[14] Valle RF, Sciarra JJ. Hysteroscopic treatment of the septate uterus. Obstet Gynecol. 1986 Feb. 67(2):253-7.

[15] Lobaugh ML, Bammel BM, Duke D, Webster BW. Uterine rupture during pregnancy in a patient with a history of hysteroscopic metroplasty. Obstet Gynecol. 1994 May. 83(5 Pt 2):838-40.

[16] DeCherney AH, Russell JB, Graebe RA, Polan ML. Resectoscopic management of mullerian fusion defects. Fertil Steril. 1986 May. 45(5):726-8.

[17] Gray SE, Roberts DK, Franklin RR. Fertility after metroplasty of the septate uterus. J Reprod Med. 1984 Mar. 29(3):185-8.

[18] Hickok LR. Hysteroscopic treatment of the uterine septum: a clinician's experience. Am J Obstet Gynecol. 2000 Jun. 182(6):1414-20.

[19] Daly DC, Walters CA, Soto-Albors CE, Riddick DH. Hysteroscopic metroplasty: surgical technique and obstetric outcome. Fertil Steril. 1983 May. 39(5):623-8.

[20] Zanetti E, Ferrari LR, Rossi G. Classification and radiographic features of uterine malformations: hysterosalpingographic study. Br J Radiol. 1978 Mar. 51(603):161-70.

[21] Heinonen PK. Complete septate uterus with longitudinal vaginal septum. Fertil Steril. 2006 Mar. 85(3):700-5.

[22] Rock JA, Zacur HA, Dlugi AM, et al. Pregnancy success following surgical correction of imperforate hymen and complete transverse vaginal septum. Obstet Gynecol. 1982 Apr. 59(4):448-51.

[23] McKusick VA, Bauer L, Koap CE, Scott RB. Hydrometrocolpos as a simply inherited malformation. JAMA. 1964 Sep 14. 189:813-6.

[24] Suidan FG, Azoury RS. The transverse vaginal septum: a clinicopathologic evaluation. Obstet Gynecol. 1979 Sep. 54(3):278-83.

[25] Banerjee AK, Clarke O, MacDonald LM. Sonographic detection of neonatal hydrometrocolpos. Br J Radiol. 1992 Mar. 65(771):268-71.

[26] Shatzkes DR, Haller JO, Velcek FT. Imaging of uterovaginal anomalies in the pediatric patient. Urol Radiol. 1991. 13(1):58-66.

[27] Khunda SS, Al-Omari S. A new approach in the management of lower Mullerian atresia. J Obstet Gynaecol. 1998 Nov. 18(6):566-8.

[28] Turner G. A second family with renal, vaginal, and middle ear anomalies. J Pediatr. 1970 Apr. 76(4):641.

[29] Tugay M, Tugay S, Inan N, Ozkan SO, Anik Y. Distal vaginal agenesis in monozygotic twins: case report. J Pediatr Surg. 2008 Apr. 43(4):765-7.

[30] Imamoglu M, Cay A, Sarihan H, Kosucu P, Ozdemir O. Two cases of pyometrocolpos due to distal vaginal atresia. Pediatr Surg Int. 2005 Mar. 21(3):217-9.

[31] Dursun I, Gunduz Z, Kucukaydin M, Yildirim A, Yilmaz A, Poyrazoglu HM. Distal vaginal atresia resulting in obstructive uropathy accompanied by acute renal failure. Clin Exp Nephrol. 2007 Sep. 11(3):244-6.

[32] Scanlan KA, Pozniak MA, Fagerholm M, Shapiro S. Value of transperineal sonography in the assessment of vaginal atresia. AJR Am J Roentgenol. 1990 Mar. 154(3):545-8.

[33] Tolhurst DE, van der Helm TW. The treatment of vaginal atresia. Surg Gynecol Obstet. 1991 May. 172(5):407-14.

[34] Acien P, Acien M, Sanchez-Ferrer ML. Müllerian anomalies "without a classification": from the didelphys-unicollis uterus to the bicervical uterus with or without septate vagina. Fertil Steril. 2008 Mar 24.

[35] Acien P, Sanchez-Ferrer M, Mayol-Belda MJ. Unilateral cervico-vaginal atresia with ipsilateral renal agenesis. Eur J Obstet Gynecol Reprod Biol. 2004 Dec 1. 117(2):249-51.

[36] Chen YT, Mattison DR, Feigenbaum L, Fukui H, Schulman JD. Reduction in oocyte number following prenatal exposure to a diet high in galactose. Science. 1981 Dec 4. 214(4525):1145-7.

[37] Cramer DW, Ravnikar VA, Craighill M, Ng WG, Goldstein DP, Reilly R. Müllerian aplasia associated with maternal deficiency of galactose-1-phosphate uridyl transferase. Fertil Steril. 1987 Jun. 47(6):930-4.

[38] Davydov SN. [Colpopoeisis from the peritoneum of the uterorectal space]. Akush Ginekol (Mosk). 1969 Dec. 45(12):55-7.

[39] Fedele L, Bianchi S, Frontino G. Septums and synechiae: approaches to surgical correction. Clin Obstet Gynecol. 2006 Dec. 49(4):767-88.

[40] Fedele L, Zamberletti D, Vercellini P, Dorta M, Candiani GB. Reproductive performance of women with unicornuate uterus. Fertil Steril. 1987 Mar. 47(3):416-9.

[41] Franz RC. Sigmoid colon vaginoplasty: a modified method. Br J Obstet Gynaecol. 1996 Nov. 103(11):1148-55.

[42] Lavanya R, Urala MS. Segmental absence of the fallopian tube--a rare anomaly. Int J Gynaecol Obstet. 1994 Nov. 47(2):175-6.

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