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Surgical Management of Mullerian Development Anomalies: 5 Years' Experience of a Tertiary Care Center

Sah MK*, Basnet T, Yadav P, Yadav J

Department of Obstetrics and Gynaecology, B.P Koirala Institute of Health Sciences, Dharan, Nepal. *Corresponding Author's Email: drgmanoj@gmail.com

ABSTRACT

Maldevelopment of the mullerian duct occurs in variety of forms and each anomaly is distinctive. This study was conducted to study clinical profile of patient with mullarian anomalies, different modalities used in diagnosis, and surgery performed to patients with mullerian duct anomalies. This was retrospective study of 23 patients who underwent surgery for different mullerian duct anomalies in last 5 years. There presenting problem, diagnostic modalities used, types of anomalies, other associated anomalies and different surgical management were evaluated. As per American Fertility society 6 patients belong to class I, 12 in class II and 5 in class III. Most of patients with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome had primary amenorrhea, patient of vaginal septum usually presents with cyclical abdominal pain, primary subfertility or dyspareunia. Patients with septate uterus had recurrent abortion 2D Ultrasonography (USG), Magnetic Resonance imaging (MRI) and hysteroscopy were modalities used for diagnosis of these defects. Patient of MRKH syndrome and vaginal agnesis were managed with vaginoplasty using amnion graft. Patients with septate uterus were managed with hysteroscopic septal resection. Septum resection was performed in all patients with transverse vaginal septum. Uterine anomalies are different groups of malformations with a broad spectrum of presentations. Management of each anomaly is individualized depending on the symptoms and fertility concerns.

Key words: Diagnosis, Mullerian duct anomalies, Surgical management.

INTRODUCTION

Mullerian duct anomalies consist of structural malformations resulting from abnormal development of the paramesonephric or Mullerian ducts (MDs). MDs are mesodermal in origin and differentiate to form uterus, fallopian tubes, cervix and upper one third of the vagina. Lower two thirds of the vagina is derived from urogenital sinus that has an endodermal origin. These anomalies arise whenever there is dysregulation or interruption in any of the processes of differentiation, migration, fusion and canalization of the two primary structures Mullerian duct and urogenital sinus. If there is Failure in the development of one or both Mullerian ducts, it leads to uterine agenesis whereas incomplete development results in uterine hypoplasia or a unicornuate uterus.

Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH syndrome) is a condition with the absence of uterus, cervix and upper part of the vagina. When there is incomplete fusion of the caudal portion of the Mullerian duct it results in lateral fusion defects, i.e., uterus didelphys, bicornuate uterus and arcuate uterus. Failure of reabsorption of the central septum results in a septate and sub-septate uterus, while vertical fusion defects lead to anomalies such as an imperforate hymen, transverse vaginal septum, oblique vaginal septum, or the absence of the cervix (1). The prevalence of these anomalies ranges from 0.001 to 10% in the general population. The embryological development of the female reproductive system is closely related to the development of the urinary system, and anomalies in both systems may occur in up to 25% of these patients.

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Other associated malformations may affect the gastrointestinal tract (12%) or musculoskeletal system (10-12%) (2). The initial diagnostic method is the two-dimensional ultrasound (US2D), but there are various other modalities like three-dimensional ultrasound (US3D), MRI, hysterosalpingo-contrast-sonography, X-ray hystero-salpingography, video-hysteroscopy and video-laparoscopy (3). Some Mullerian malformations are healed with surgery and the success of the treatment depends on accurate diagnosis and the choice of the best technique. Vaginal agenesis can be treated with dilatation or vaginoplasty. When the dilatation is unsuccessful, vaginoplasty could be done by various techniques (4). Congenital cervical and vaginal atresia is controversial in terms of treatment, since there are no guidelines or randomized studies that point to the best surgery. It is a gynecological urgency with various risks and hysterectomy is the principal option for treatment according to several authors, principally due to its innumerous benefits (5).

Unicornuate uterus needs surgical correction if there is a cavitated noncommunicating rudimentary uterine horn. It needs resection due to the pain that results from the impediment of menstrual flow (6). Uterus didelphys only needs intervention in a condition when one of the vagina is obliterated and the septum between them needs to be resected to drain the hematocolpos and hematometra, as well as it allows for the outflow of normal menstrual flow. Bicornuate uterus cause recurrent miscarriage in the 2nd trimester of pregnancy and premature birth. Strassman's metroplasty can be recommended, with good results and a 90% rate of full term pregnancy. Septate uterus is the most common mullerian malformation. It is associated with miscarriages in the first and second trimesters, although the treatment is relatively easy. The chosen treatment is hysteroscopy metroplasty, with good results (7).

Transverse vaginal septum should be treated with surgical resection and anastomosis of the proximal and distal vaginas. The choice of the technique depends on its localization and thickness, which is diagnosed in a physical exam, ultrasonography and MRI. The lower vaginal septum with lesser thickness and perforated types has the good results. It can leads to few complications like stenosis, re-obstruction, dyspareunia and psychological difficulties. Vaginal dilatation is generally recommended after surgery to improve the result (3).

METHODOLOGY

This was retrospective descriptive study. All the patients who had undergone surgery at B.P Koirala Institute of Health Sciences, Dharan, Nepal for mullerian anomalies in last 5 years from January 2017 to December 2021 were included in this study. Any patient whose data cannot be retrieved was excluded from the study. Permission was taken from the medical review board for the retrieval of case records for conducting this research. All the details of patient were obtained from case records that include age, parity. Detail history was reviewed to know the clinical presentation and specific past, personal and family history were also obtained. All examination finding was also reviews that include general physical examination, systemic examination and pelvic examination. Specific diagnostic modalities used for diagnosis was obtained and any associated other congenital anomalies was noted. Surgery performed to the patients was also recorded.

Ethical Approval

Ethics approval was obtained from Institutional Review Committee before conducting the study (IRC/2243/022).

Statistical analyses

Statistical analysis was conducted with SPSS version 21. Categorical variables expressed as percentage.

RESULT

We have included 23 patients with mullarian anomalies who underwent surgery at our centre in last 5 years from January 2017 to January 2022. All patients were of 11 to 27 years (Table 1).

All patients presented with different complain, primary amenorrhea was most common complain seen in 43.47% of cases, followed by cyclical abdominal pain in 30.43%, primary subfertility in 8.7%, Recurrent abortion in 8.7%, Irregular Cycle in 4.35%, Dyspareunia in 4.35% (Table 2).

Table 1: Distribution of Patients according to Age

Age Groups	Number of patients N (%)
11-16	7(30.43)
17-21	9(39.14)
22-27	7(30.43)

Table 2: Distributions of patients according to clinical presentations

Presenting symptoms	N (%)
Primary Amenorrhea	10(43.47)
Cyclical abdominal pain	7(30.43)
Primary Subfertility	2(8.70)
Recurrent abortion	2(8.70)
Irregular cycle	1(4.35)
Dyspareunia	1(4.35)

Table 3: Distribution of patients according to the American Fertility Society classification of utero-vaginal Anomalies

Class	N (%)
I	6(26.08)
II	12(52.17)
III	5(21.73)
IV	0

We found six patients who presented with primary amenorrhea had MRKH syndrome (Class I anomaly). They were advised for USG and later on MRI was done. All patients underwent vaginoplasty and amnion graft was used in all patients. All of them were advised to use vaginal dilator in post-operative periods to keep vaginal canal patent. Patients with vaginal atresia underwent vaginoplasty.

Ten patients found to have vaginal septum and two patients had vaginal agenesis (Class II anomaly), most of them presented with cyclical abdominal pain. USG and MRI were done which show vaginal septum at different level with hematometra or hematometrocolpos. All patients underwent septum resection and drainage of hematometra. Two patients had vaginal agenesis and vaginoplasty done.

One patient who presented with primary subfertility was advised for USG and MRI. She had unicornuate uterus with non-communicating rudimantory (Class III anomalies) along with a large endometriotic cyst. Excision of rudimentary horn and left salpingectomy was done along with cystectomy for endometrioma. Two patients who had history of recurrent pregnancy loss and a patient with primary subfertility underwent diagnostic hysteroscopy and diagnosed as uterine septum (Class III anomalies) and later on hysterescopic septum resection was performed.

We also had one patient with Uterine didelphys with transverse vaginal wall septum in anterior vaginal who presented with complain of pain abdomen and history of a spontaneous pregnancy loss. Septum resection was performed in this patient.

Table 4: Types of anomalies, Diagnostic modalities, Surgical treatment provided and Associated anomalies

Type of Anomalies	No of patients	Presenting Symptoms	Diagnostic modalities	Surgical treatments	Associated anomalies
Transverse vaginal septum	10	Cyclical abdominal pain	USG and MRI	Septum Resection	Renal agenesis in a patient
MRKH Syndrome	6	Primary Amenorrhea	USG and MRI	Vaginoplasy	
Unicornuate Uterus with non-communicating rudimentary horn with endometriosis	1	Primary Subfertility	USG	Excision of rudimentary horn with left salpingectomy and cystectomy for endometrioma	
Septate Uterus	3	Recurrent abortion and primary subfertility	USG and hysteroscopy	Hysteroscopic resection of septum	
Uterine Didelphys with transverse vaginal septum	1	Cyclical pain	USG and MRI	Septum Resection	
Vaginal agenesis	2	Primary Amenorrhea	USG and MRI	Vaginoplasty	

DISCUSSION

Uterine anomalies are different groups of malformations with a broad spectrum of presentations (8). This study was aimed to study clinical profile of patients with mullerian anomalies, diagnostic modalities used and surgical treatment provided to patients with mullerian duct anomalies.

Patients with mullarian development anomalies mostly present with primary amenorrhea, cyclical abdominal pain, primary subfertility and recurrent abortion. Which is consistent with study conducted by Goyal *et al*. (1).

All suspected patient should be advised for ultrasonography. Patients in this study were investigated with 2D USG and MRI. MRI is the most accurate imaging study for uterine anomalies; it reaches accuracy to 100%, compared to the results of combined hysteroscopy and laparoscopy (9). Study done by Sahar Mahmoud *et al*. found 3D TVS proved to be highly accurate in the diagnosis of uterine anomalies and showed strong agreement with MRI as both can provide valuable information about both the internal details and external contour of the uterus (10). We found only 1 (4.34%) had associated renal agenesis contrast to Eline *et al.* where they found 64.4% of renal agenesis were associated with mullarian duct anomalies (11).

In our study, we encountered six patients with MRKH syndrome. The treatment goal in these women is the creation of an artificial vagina surgically, that allows sexual functioning. McIndoe procedure is the surgical creation of an artificial vagina using a split skin graft or an amnion graft (12). In our study, we had also performed McIndoe vaginoplasty using autologous amnion grafts and there was no any intraoperative and postoperative complication. Williams *et al.* has conducted the study where they had used amnion graft in 28

patients. They had one patient who has rectal injury during procedure (13). Goyal *et al.* had used both skin and amnion graft (1).

Transverse vagina septum was most common anomaly we found. Transverse vaginal septum is classified based on septum location. The location of the septum is based on the distance from the vaginal introitus to the distal end of the septum and it can be low (< 3 cm), mid (3–6 cm) or high (> 6 cm). Most of the patients in our study had the septum located in the lower part of the vagina. This finding is similar to the study by Williams *et al.* (13) and Sardesai *et al.* (14). There are various techniques of managing vaginal septum, which include simple excision through the vagina, incision and drainage of hematocolpos, vaginoscopy resection of septum, abdominoperineal vaginoplasty (14). Most of the patients in this study had simple excision of the septum as majority had a low type that was easy to approach through the vagina.

Raga *et al.* reviewed all pregnancies among a population with congenital uterine malformations and found a 25.5% incidence of early miscarriage (≤13 weeks) and 6.2% incidence of late miscarriage (14–22 weeks) in the group with a septate uterus (15). Moreover, De Cherney *et al.* showed that hysteroscopic removal of the septum in women with recurrent (≥3) first trimester miscarriage improved reproductive outcome (16). We had two patients who had recurrent pregnancy loss and was diagnosed to have septate uterus. Septum was resected via hysteroscopy approach.

Didelphys uterus is a very rare uterine anomaly, can be associated with a longitudinal vaginal septum. Combination with transverse vaginal septum makes it even rarer (17). We managed one patient of didelphys uterus with transverse uterine septum where septum resection was performed.

CONCLUSION

Uterine anomalies are different groups of malformations with a broad spectrum of presentations. Management of each anomalies are individualized depending on the symptoms and fertility concerns.

Limitations

This was retrospective descriptive study. We could not follow up patients and surgical outcome could not be evaluated in this study.

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CONFLICT OF INTEREST

None declared.

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