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Research Article

Analysis of Mullerian developmental defects in a tertiary care hospital: a four year experience

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ABSTRACT

Background: Mullerian duct anomalies are congenital anomalies of the female genital tract. Mullerian duct anomalies affect the reproductive health of the female to a varied extent. This may present as obstructive or as non-obstructive anomalies. When clinically suspected, proper investigations are required to confirm the diagnosis including ultrasonography, laparoscopy and hysteroscopy.

Methods: In the present study, all the patients presenting with Mullerian duct anomalies to the gynaecology OPD at General Hospital, Sola, GMERS Medical College, Ahmedabad during the four year duration from 2011 to 2014 were included. These patients were analysed with respect to their incidence, presenting complain, age at presentation, classified according to AFS classification after proper diagnosis and managed individually.

Results: The incidence of Mullerian duct anomalies was found to be 0.084%. 52.9% belonged to the age group of 21 to 25 years. Most of these patients (73.5%) presented with chronic complains. Only a few presented with acute symptoms (14.7%) whereas a few were asymptomatic (11.8%). Septate uterus (20.6%) was the most common anomaly diagnosed at our setup followed by bicornuate uterus (17.7%) and transverse vaginal septum (17.6%). History along with clinical and USG examination were adequate in diagnosis of patients with vaginal agenesis, didelphic, bicornuate uterus, TVS and imperforate hymen. Laparoscopy was needed to confirm cases of MRKH & unicornuate uterus and hysteroscopy for cases of septate uterus. Arcuate uterus was diagnosed during LSCS.

Conclusions: It was thus concluded that with timely evaluation, diagnosis and optimal management, their menstrual disturbances are relieved early in adolescence period recuperating their psychiatric health & improving their reproductive carrier. Delay in management may cause serious complications and potential infertility.

Keywords: Mullerian anomalies, TVS, Unicornuate uterus, Bicornuate uterus

INTRODUCTION

Mullerian duct anomalies arise due to a variety of embryological disruption during its (Paramesonephric duct) development. This may be due to complete agenesis, defective vertical or lateral fusion, or resorption failure. The prevalence ranges from 0.001 to 10% in general population and 8-10% in those with adverse reproductive history.^{1,2} Mullerian Duct Anomalies are associated with renal anomalies (25%), skeletal

anomalies (12%) and GIT anomalies (12%) due to its close association during its development.^{3,4} Among the cases of Mullerian duct anomalies, a few present to the emergency as acute abdominal pain or urinary retention where as a majority present with chronic complains related to menstruation and fertility. Thus this may present as obstructive or as non-obstructive anomalies. When clinically suspected, proper investigations are required to confirm the diagnosis including ultrasonography, laparoscopy and hysteroscopy. Though all the cases are classified into 7 groups according to

AFS,³ each and every case requires individualisation for optimal management. With good surgical skills, most of these conditions can be surgically treated with finest outcome.

Aims and objectives

1. To determine the incidence of Mullerian duct anomalies
2. Analysis of various clinical presentation in cases with Mullerian duct anomalies
3. Formulating the optimal management in diagnosed patients when required

METHODS

Our study was a prospective longitudinal observational study conducted in at General hospital, Sola, GMERS Medical College, Ahmedabad between the time period from 2011 to 2014.

Among 40000 patients reporting to the Obstetrics & Gynaecology OPD in the above mentioned period, 34 cases were found to have Mullerian duct anomalies suspected through history and diagnosed by clinical examination, radiological procedures (mainly USG) and endoscopic procedures (Laparoscopy & hysteroscopy). None of these underwent MRI. A few were diagnosed to have Mullerian duct anomalies during laparoscopy - hysteroscopy during infertility investigation. Few were diagnosed during obstetric operative procedure mainly LSCS. Results were analysed based on their age at presentation, chief complaint with which the patient reported, type of anomaly diagnosed, investigation helpful in diagnosis, management and outcome followed upto 1 year.



Figure 1: Hematocolpos on USG.

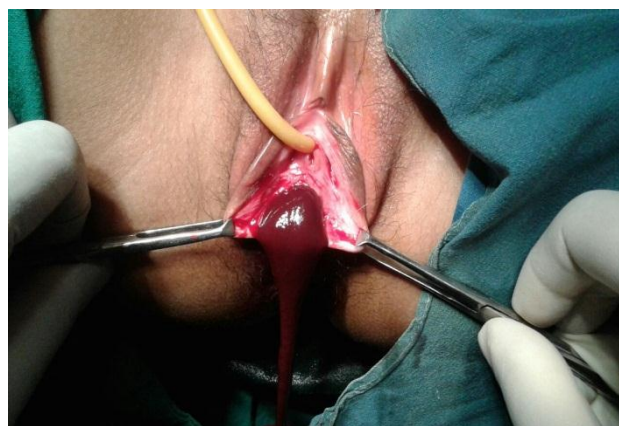


Figure 2: Hematocolpos drainage.

RESULTS

Out of the 40000 patients attending the OPD, 34 patients were found to have Mullerian duct anomalies. Hence the incidence was found to be 0.084%.

Out of the total 34 patients, 26.47% (9/34) were unmarried at the time of presentation where as 73.52% (25/34) were married.

Table 1: Age at presentation.

Age (years)	No. of patients (n=34)	% of patients
10-15	5	14.7
16-20	5	14.7
21-25	18	52.9
26-3	4	11.8
31-35	2	5.9

In our study, 52.9% belonged to the age group of 21 to 25 years. 14.7% belonged to the age group of 16 to 20 years and 10-15 years where as 11.8% belonged to the age group of 26-30 years each. The earliest presentation was at 13 years of age and the maximum age among the study population was a case at 33 years.

Most of these patients (73.5%) presented with chronic complains. Only a few presented with acute symptoms (14.7%) whereas a few were asymptomatic (11.8%).

Table 2a: Mode of presentation.

Acute	Chronic	Asymptomatic
5	25	4

Most of these presented with more than one complains. The principle complaint of these patients were analysed which brought them to the hospital or which prompted the diagnosis of Mullerian anomaly.

Table 2b: Chief complaint.

Chief complaint	No. of patients (n = 34)	% of patients
Acute abdominal pain	4	11.8
Chronic abdominal pain	2	5.9
Cyclical dysmenorrhoea	2	5.9
Primary amenorrhea	13	38.2
Primary infertility	10	29.4
Secondary amenorrhea	2	5.9
Secondary infertility	1	2.9
Urinary retention	2	5.9
Abnormal labour	3	8.8
Asymptomatic	4	11.8

Among the 4 cases that presented with acute abdominal pain, 2 were diagnosed as TVS (one high, one low) 1 was that of unruptured hymen and 1 of upper vaginal agenesis. Both the cases of unruptured hymen and low TVS were associated with urinary retention from a hematocolpos causing acute abdominal pain.

Among the 2 cases of chronic abdominal pain, one was a case of high TVS. Other was a case of unruptured hymen.

Both the cases of cyclic dysmenorrhoeal were those of unicornuate uterus with non-communicating functional horn that caused the cyclical pain due to the haematometra from the functional endometrium in the contra lateral horn. In both the cases, horn was excised.

Among the 13 cases of primary amenorrhea, 4 were MRKH syndrome. 6 were those of TVS which was excised and 1 was that of upper vaginal agenesis and a pull through operation was performed in it. 2 was a case of unruptured hymen.

Among the 10 cases of primary infertility, 6 were of complete septate uterus. 3 were bicornuate uterus with no associated complain. One was a low TVS with recurrent fibrosis post-surgery (operated thrice earlier) with recurrent secondary amenorrhea.

2 cases were of secondary amenorrhea where both had a TVS with recurrent fibrosis.

One case that presented as secondary infertility (H/o spontaneous abortions) was a case of complete septate uterus.

Among the 2 cases those were admitted in emergency department with urinary retention, one was of a low TVS and the other of unruptured hymen, both with haematometra, hematosalpinx and haematocolpos.

3 cases were diagnosed during LSCS that were performed for abnormal progress of labour. Persistent occipitoposterior position was seen in a case of arcuate

uterus, breech with NPOL in case of unicornuate uterus and NPOL in a case of arcuate uterus.

Among the 6 asymptomatic cases, one was a didelphic uterus diagnosed when the patient presented for a bartholins cyst. 2 cases were those of unicornuate uterus with non-communication non-functional horn - one diagnosed during LSCS (ERCS) and other on USG for early pregnancy. 3 cases were bicornuate uterus diagnosed during USG in early pregnancy.

Table 3: Type of Mullerian duct anomalies (According to AFS classification).

Type of anomaly (AFS classification)	No. of patients	%
Hypoplasia/ aplasia		
1a vaginal agenesis	1	14.7%
1e combined agenesis	4	
Unicornuate		
2b non communicating	4	14.7%
2d No horn	1	
Didelphus	1	2.9%
Bicornuate		
4a complete	6	17.7%
Septate		
5a complete	6	20.6%
5b incomplete	1	
Arcuate	2	5.9%
7 DES related	0	
Transverse vaginal septum	6	17.6%
Unruptured hymen	2	5.9%

Septate uterus (20.6%) was the most common anomaly diagnosed at our setup followed by bicornuate uterus (17.7%) and transverse vaginal septum (17.6%).

Table 4: Type of Mullerian duct anomalies with principle investigation of diagnosis.

Type of anomaly (AFS classification)	Diagnostic procedure
Hypoplasia/ aplasia	
1a vaginal agenesis	CE, USG
1e combined agenesis	CE, Laparoscopy
Unicornuate	1 - USG,
2b non communicating	1 - LSCS
2d No horn	2 - USG, Laparoscopy
	1 - LSCS
Didelphus	CE, USG
Bicornuate	4 - USG
	2 - USG, Laparoscopy
Septate	Hysteroscopy
Arcuate	LSCS
7 DES related	0
TVS	CE, USG
Unruptured hymen	CE, USG

History along with clinical and USG examination were adequate in diagnosis of patients with vaginal agenesis, didelphic, bicornuate uterus, TVS and imperforate Hymen. Laparoscopy was needed to confirm cases of MRKH & Unicornuate uterus and Hysteroscopy for cases of septate Uterus. Arcuate Uterus was diagnosed during LSCS.

DISCUSSION

The incidence in the present study was 34 cases out of 40000 total patients (0.084%). In the study conducted by JayatiNath et al.⁵ the incidence was 0.1% and similar in the study by Rock et al.^{1,3} by Crook et al.⁶ Most authors report incidence between 0.1%-3.5 %.⁷⁻⁹ Grimbizis et al.¹⁰ reported mean incidence of 4.3% in general population.

In our study, majority of cases presented in the age of 21 to 25 years (52.9%). In the study by Jayati Nath et al.,⁵ 80% of patients presented between 10 to 19 years. In the study by Mane et al.¹¹ the mean age of presentation was 17 years. Reindollar et al.¹² reported majority of cases in adolescence. Banerjee et al.¹³ reported 52.6% of cases in the age group of 15-17 years.

In our study, 73.5% of patients presented with chronic complains whereas only 14.7% presented to the emergency department with acute symptoms like abdominal pain or urinary retention. Among those with chronic complains, 5.9% had chronic abdominal pain, primary amenorrhoea in 26.5%, cyclic dysmenorrhoea was presenting symptom in 5.9 %, 5.9% were with chronic abdominal pain and 32.3% had infertility. In the Jayati Nath et al.,⁵ incidence of above complains was 50%, 43.7%, 25%, 18% and 31.2% where as in Jeon et al.¹⁴ the incidence was 10.8%, 11.5%, 12.15%, 12.4% and 8.6% respectively. Banerjee et al.¹³ reported abdominal pain, primary amenorrhoea and infertility in 37%, 31.5% and 31.5% respectively. Among the cases that presented with primary amenorrhea, 4 were those of MRKH and 6 of TVS. The results were quite similar to those cited by Parikh et al.¹⁵ and Banerjee et al.¹³ who also found these 2 conditions were most common in causes of primary amenorrhoea.

In the present study, 14.7% has agenesis, 14.7% had unicornuate uterus, 2.9% didelphic uterus, 17.7% had bicornuate uterus, 20.6% had septate uterus & 5.9% had arcuate uterus. The % of each of these abnormalities in the study by Jayati et al.⁵ were 18.7, 25, 6.25, 18.7, 6.25 & 6.25 respectively whereas that quoted by Ayush et al.¹⁶ were 6-25% for unicornuate uterus, 5-11% for didelphic uterus, 10-39% for bicornuate uterus, 34-55% for septate uterus and 7% for arcuate uterus. 17.6% of cases had TVS and 5.9% had unruptured hymen in the present study where as in the study by Jayati et al.⁵ there were 18.75% cases of TVS.

When asymptomatic, these require no treatment. However those with Obstruction require early treatment and also those with infertility. Vaginoplasty was

performed in 3 cases of MRKH where as it was reserved for future in 1. A Pull through operation was performed in the case of upper vaginal agenesis. 2 cases of unicornuate uterus with functional non communication horn were treated with excision of the horn. All the cases septate uterus were treated with hysteroscopic resection of the septum. All the cases of TVS were treated with resection of the septum followed by mould placement. Unruptured hymen was incised to drain the haematocolpos. No treatment was offered in asymptomatic patients with arcuate uterus, didelphic uterus & asymptomatic bicornuate uterus.

CONCLUSION

Out of 40000 OPD patients, 34 were diagnosed with Mullerian duct anomalies with an incidence of 0.084.52.9% patients belonged to the age group of 21 to 25 years. The commonest presenting complain in the reproductive age group was primary infertility whereas that in adolescence was primary amenorrhoea. Maximum number of cases belonged to bicornuate uterus, septate uterus and transverse vaginal septum. Cases presenting with recurrent fibrosis are prevalent in TVS following faulty techniques of surgery. Treatment modality was tailored according to the case diagnosed.

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Ethical approval: The study was approved by the institutional ethics committee

REFERENCES

1. John A. Rock, Lesley L. Breech. Surgery for anomalies of Mullerian ducts. In: Rock JA, Jones HW, eds. Te Linde's Operative Gynaecology. 10th ed. Philadelphia: Lippincott Williams Wilkins; 2013: 539-584.
2. Rackow BW, Arici A. Reproductive performance of women with Mullerian anomalies. *Curr Opin Obstet Gynaecol.* 2007;19:229-37.
3. John A. Rock, Lesley L. Breech. Surgery for anomalies of Mullerian ducts. In: Rock JA, Jones HW, eds. Te Linde's Operative Gynaecology. 10th ed. Philadelphia: Lippincott Williams Wilkins; 2013: 539-584.
4. Evans TN, Poland ML, Boving RL. Vaginal malformations. *Am J Obstet Gynaecol.* 1981;141:910-20.
5. Jayati Nath, Nayana Pathak. A study of Mullerian anomalies in a tertiary care teaching hospital of North India. *Int J Sci Res.* 2015;4:1020-2.
6. Crook DO, John B, Gebbert MD. Congenital anomalies of the female urogenital tract. *J Pelvic Med Surg.* 2005;11:165-81.
7. Strassmann EO. Operations for double uterus and endometrial atresia. *Clin Obstet Gynecol.* 1961;4:240.

8. Strassmann EO. Fertility and unification of double uterus. *Fertil Steril.* 1966 Mar-Apr;17(2):165-76.
9. Golan A, Langer R, Bukovsky I, Caspi E. Congenital anomalies of Mullerian system. *Fertil Steril.* 1989 May;51(5):747-55.
10. Grimbizis GF, Camus M, Tarlatzis BC, Bontis JN, Devroey P. Clinical implications of uterine malformations and hysteroscopic treatment results. *Hum Reprod Update.* 2001;7:161-74.
11. Mane SB, Shastri. Our 10-year experience of variable Mullerian anomalies and its management. *Pediatr Surg Int.* 2010;26:795-800.
12. Reindollar RH, Byrd JR, McDonough PG. Delayed sexual development: a study of 252 patients. *Am J Obstet Gynaecol.* 1981;140:371-80.
13. Banerjee I, Mondal SC, Dam P, Roy P. Case series of Mullerian developmental defects encountered in a tertiary care hospital: a one-year experience. *Open J Obstet Gynecol.* 2014;4:733-44.
14. Jeon GH, Park YR, Shin YJ. Clinical characteristics of women with Mullerian anomaly: twenty years of experience at Asan medical center. *Korean J Obstet Gynaecol.* 2010;53:626-32.
15. Parikh RM, Nakum K, Kadikar GK, Gokhle AV. Mullerian anomalies: a cause of primary amenorrhea. *Int J Reprod Contracept Obstet Gynecol.* 2013;2:393-7.
16. Ayush Goel, Frank Gaillard. Mullerian duct anomaly classification. Available at: <http://radiopedia.org/articles/mullerian-duct-anomaly-classification>.

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