DOI: 10.14260/jemds/2015/1749

ORIGINAL ARTICLE

A STUDY OF MULLERIAN DUCT ANOMALY ON REPRODUCTIVE AGE GROUP

Debjani Roy[1]

HOW TO CITE THIS ARTICLE:
Debjani Roy. "A Study of Mullerian Duct Anomaly on Reproductive Age Group". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 70, August 31; Page: 12143-12151, DOI: 10.14260/jemds/2015/1749

ABSTRACT: Congenital anomaly of reproductive tract has varied modes of presentation which may present at any time from birth to menopausal age, mostly at reproductive age group. Though its incidence is rare, it is still a major topic of discussion among pediatricians, gynecologists, embryologists, pathologists, anatomists, endocrinologists, radiologists and psychiatrists etc. From the present study, one may have a guideline regarding the approach & outcome of a patient with anomaly of reproductive tract. **METHODS:** Patients were selected randomly from GOPD and emergency OT from June 2006 to Aug. 2007 with various problems of mullerian duct anomaly asked for treatment at Gynae & Obst., R.G.K. M.C.H., Kolkata with collaboration of radiology department (For radiological investigations) and (Pathology department for diagnosis of sex chromatin). A series of 54 cases of patients with congenital anomalies of reproductive organ in reproductive age group (12-50 years) were studied clinically, radiologically along with detection of sex chromatin from buccal smear with/without karyotyping and hormonal assay and outcome of these patients were studied when corrective measures given. **RESULTS:** Among 54 patients, maximum patient's cluster (33) at peripubertal age (12-20 years). With gynaecological problems (28). After receiving corrective measures outcome were good for almost 88% which depicts the p value 0.03. Ultrasonography showed almost 96.3% accuracy to detect the congenital anomaly. The study also shows Prevalence of congenital anomaly at G.O.P.D. of a regional medical college hospital of West BENGAL is 0.15 which is very near to value given in literature (0.16%). **CONCLUSION:** The aim of the study was to search out the different congenital anomalies of female reproductive organ and to improve the qualities of life of those patients with different new treatment modalities. Malformations of uterus may not be stamped as rare as it is probable that many malformations are never diagnosed because many patients with such malformations are able to conceive and proceed to term without complication. Mullerian duct anomalies encompass a wide spectrum of clinical and imaging findings, and in certain malformations surgical intervention is justified.

KEYWORDS: Mullerian Duct Anomaly (MDA), Atresia, Agenesis, Turner, Rockitnasky-Kauser-Hauser (RKH) syndrome.

INTRODUCTION: The mullerian ducts are the primordial anlage of the female reproductive tract; hence a wide variety of malformations can occur when this system is disrupted. They range from uterine and vaginal agenesis to duplication of the uterus and vagina to minor uterine cavity abnormalities. Mullerian abnormalities are frequently associated with abnormalities of renal and axial skeletal systems; mullerian abnormalities are often first encountered when patients are examined for those associated conditions. (Anomalies of Vertibras & Ribs; a Developmental Field Defect).

Most mullerian duct abnormalities (MDAs) are associated with functioning ovaries and age appropriate external genitalia. These abnormalities are often recognized after the onset of puberty.

DOI: 10.14260/jemds/2015/1749

In the prepubertal period, normal external genitalia and age appropriate developmental milestones often mask abnormalities of the internal reproductive organs. After the onset of puberty, young women often present to the gynaecologists with menstrual disorder; late presentations include infertility and obstetric complications & also psychiatric problem.[1]

The aim of the study of different cases of gynaecological and obstetrical problems of reproductive age group associated with developmental anomalies of reproductive organ by different diagnostic method (Hysterosalpingography (HSG), ultrasonography (USG), hysteroscopy, laparoscopy and biochemical test where feasible) & to throw light on the frequency of their presence in normal population & also follow up of such cases after application of corrective measure, if at all correctable.

MATERIALS AND METHODS: The study was performed at G.O.P.D. & infertility clinic of a regional medical college hospital of West BENGAL with prior permission of authority.

Subjects of this study were selected from Department of Obstetrics and Gynaecology (Both from inpatients and outpatient department) and infertility clinic who came with complaints of primary amenorrhoea, primary infertility, post caesarean recurrent breech, pain abdomen, urinary retention, habitual abortion, primi with transverse lie, etc. The age limit of the subjects was taken from 12 years to 50 years.

The most basic classification of Mullerian duct defect consists of agenesis & hypoplasia, defect of vertical fusion & lateral fusion. In 1979, Buttram & Gibbons.[2] proposed a classification of MDA that was based on the degree of failure of normal development, & they separated these anomalies into classes that demonstrate similar clinical presentations, treatment & prognosis.[3] for fetal salvage. Modified in 1988 by a sub-committee of American Fertility Society the most widely accepted classification.

AFS Classification of Anomalies of the Mullerian Duct.[4]

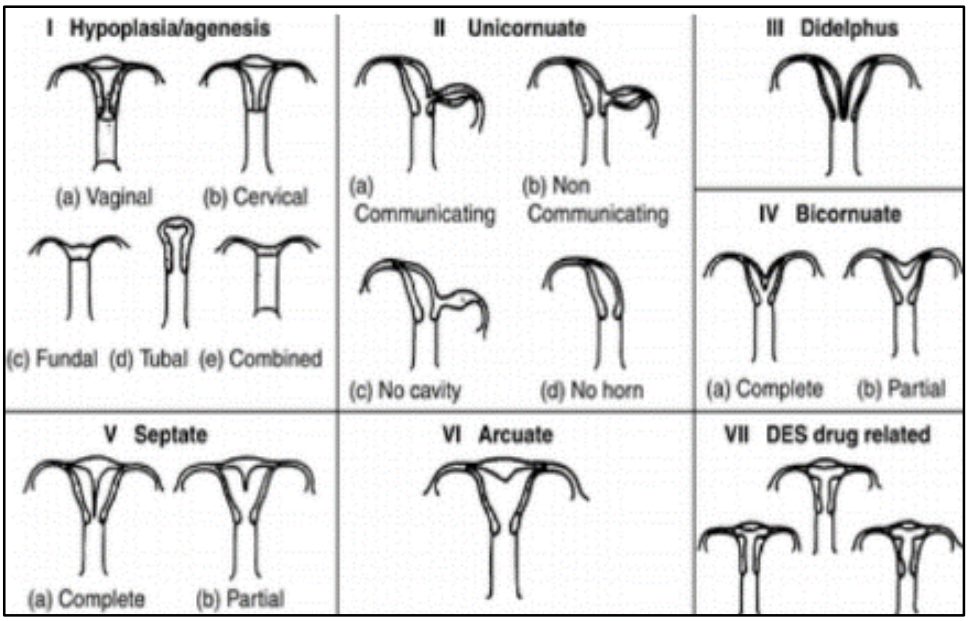

Fig. 1

DOI: 10.14260/jemds/2015/1749

Classification	Clinical Finding	Description
I	Segmental or complete agenesis or hypoplasia	Agenesis and hypoplasia may involve the vagina, cervix, fundus, tubes, or any combination of these structures. Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is the most common example in this category.
II	Unicornuate uterus with or without a rudimentary horn	When an associated horn is present, this class is subdivided into communicating (Continuity with the main uterine cavity is evident) and noncommunicating (No continuity with the main uterine cavity). The noncommunicating type is further subdivided on the basis of whether an endometrial cavity is present in the rudimentary horn. These malformations have previously been classified under asymmetric lateral fusion defects. The clinical significance of this classification is that they are invariably accompanied by ipsilateral renal and ureter agenesis (Buttram[2], 1979; Acien, 1992; Wagner, 1994).
III	Didelphys uterus	Complete or partial duplication of the vagina, cervix, and uterus characterizes this anomaly.
IV	Complete or partial bicornuate uterus	Complete bicornuate uterus is characterized by a uterine septum that extends from the fundus to the cervical os. The partial bicornuate uterus demonstrates a septum, which is located at the fundus. In both variants, the vagina and cervix each have a single chamber.
V	Complete or partial septate uterus	A complete or partial midline septum is present within a single uterus.
VI	Arcuate uterus	A small septate indentation is present at the fundus.
VII	DES-related abnormalities	A T-shaped uterine cavity with or without dilated horns is evident.

STATISTICS: The author uses the following formulas[5] for calculation of study results.

1. Standard deviation is "Root-Means-Square-Deviation" denoted by the Greek letter sigma σ or standard deviation.

$$\text{Standard Deviation} = \sqrt{\frac{\sum (x - \bar{x})^2}{(\eta)}}$$

When $\sum (x - \bar{x})^2$ square of summation of duration of Each value form arithmetic mean $(x - \bar{x})^2$.

n- Is the sample size.

If sample size <30; n is replaced by (η - 1)

DOI: 10.14260/jemds/2015/1749

2. Standard Error of the Mean:

$$S.E. \quad \bar{x} = S/\sqrt{(\eta)}$$

= S stands for standard deviation of mean

$\eta \rightarrow$ stands for total no. of sample.

3. P value:

$$3.\ \text{P value - P} = \frac{\sum x - x^1}{\sum x} \times 100$$

$\sum x \rightarrow$ Total no. of sample provides treatment.

$X^1 \rightarrow$ Person with good outcome.

If P. value is < 0.05 → Significant at 5 percent level.
If P. value is = 0.05 → just significant at 5 percent level.
If P. value > 0.05 → not significant at 5 percent level.

A test of significance will measure the probability of result by mean. If the probability of occurrence of such a difference is less than 5 times out of 100, it is said to be significant or statistically significant, i.e. , P<0.05. In that case, the difference will be due to the treatment modalities in more than 95% of such experiments. The sample estimate is then said to be significant or significantly different at 5% level of significance. This difference could be by chance only once in twenty trials. The difference is highly significant if the probability of chance occurrence is less than 1%. (P<0.01).

RESULTS: The Study has been performed at outpatient dept. of Gynae & Obst., Regional. M.C.H., WB with collaboration of radiology department (for radiological investigations) and (pathology department for diagnosis of sex chromatin). Patients were selected randomly from GOPD and emergency OT. From June 2006 to Aug-2007. 54 patients with various problems of mullerian duct anomaly asked for treatment.

The analysis of clinical data, etiology and the result of outcome are given below:

Table-1: Incidence According to age distribution (Frequency Distribution table)

Age Group (yrs.)	Cases	Percentage
12-20	33 (28 with Gynaecological problem) (5 with Obstetrical problem)	61.11%
21 – 30	20 (10 with Gynaecological problem) (10 with Obstetrical problem)	37.33%
31-40	Nil	
41-50	1	1.85%

DOI: 10.14260/jemds/2015/1749

It shows maximum patients clusters at the peripubertal age; patients mainly with Gynaecological problem come during this age group while patients with Obstetrical problem come at the most fertile period (21-30 yrs).Standard Deviation from this table of age distribution is 15.9.

Table-2: Incidence According to Caste

Caste	Cases	Percentage
Hindu	39	72.23%
Muslim	15	27.77%
Others	Nil	

It shows that maximum patients asking for treatment is of Hindu category. Standard Deviation from this table of Caste distribution is 16.6.

Table-3: According to the Gross problem (Gynaecological /Obstetrical)

Gross Problem	Cases	Percentage
Gynaecological	39	72.23%
Obstetrical	15	27.77%

Standard Deviation from this table on the basis of Gross problem is 16.6.

Table-4: According to cases come with different Menstrual problem Among 54 cases

Menstrual Problem	Cases	Percentage
Primary amenorrhoea	18	33.33%
Cryptomenorrhoea	10	18.51%
Dysmenorrhoea	6	11.11%
Others(normal menstrual history)	20	37.30%

It shows maximum patients come without any menstrual problem but with other problems (primary infertility; recurrent abortion; retention of urine; dyspaurenia; incidental findings etc.) followed by primary amenorrhoea.

Standard Deviation from this table on the basis of menstrual problem is 6.6 which show standard error with the range of approx. 3.8.

Table-5: Patient present with vaginal atresia (19cases) can be divided According to etiology as

Etiology of Vaginal Atresia	Cases	Percentage
Rockitnasky- Kauser-Hauser Synd (R.K.H.).	9	47.36%
Turner Synd	4	21.05%

It signifies maximum patients with vaginal atresia are associated with Rockitansky-Kauser-Hauser syndrome.

Standard Deviation from this table on the basis of problem of vaginal atresia is 3.1. Which shows standard error with the range of approx. 1.55.

DOI: 10.14260/jemds/2015/1749

Table-6: Patients present with obstetrical problem (15 cases) can be distributed according to diagnosis

Final Diagnosis	Cases	Percentage
Pregnancy in rudimentary horn	2	13.33%
Abnormal presentation	3	20%
Fetal loss	6	40%
Others (Menorrhagia, uterine cavity defect with DES, incidental finding during M.T.P./LUCS)	4	26.66%

Standard Deviation from this table on the basis of obstetrical problem is 1.7. Which shows standard error with the range of approx. 0.98.

Table-7-Patients Present With Mullerian Duct Abnormalities (54 Cases)

Mullerian Duct Abnormality	Cases	Percentages
Bicornuate uterus(bicornuate-9,bicornuate bicollis with septate vagina-3)	12	22.22%
Septate uterus -complete septum from fundus to vagina-1, septate uterus-2, septate uterus and cervix-2	5	9.25%
Arcuate uterus	1	1.85%
Unicornuate uterus	1	1.85%
DES associated abnormality	1	1.85%
Cervico vaginal dysgenesis	6	11.11%

It can be concluded that majority of patient with mullerian abnormality have bicornuate uterus.

Standard Deviation from this table on the basis of Mullerian duct abnormalities is 9.7.

Table-8: Distribution of Turner and RKH syndrome on the basis of Caste

Syndrome	Caste	Percentages
Turner(4)	Hindu- 2 Muslim-2	50% 50%
RKH(9)	Hindu- 5 Muslim-4	55.56% 44.44%

So, whether these congenital anomalies have association with consanguineous marriage cannot be ascertained.

Table-9: According to outcome after giving corrective measures where Possible

	Good (88.23%)	Poor (2.9%)	Defaulter (5.8%)	Awaiting	Tubal ligation	Unrelated cause (2.9%)
Obstetrics (15)	7	1	2	2	2	
Gynaecological (39)	23			6 (for marriage)		1 Ovarian tumor TAH BSO done

DOI: 10.14260/jemds/2015/1749

So it can be concluded that majority of patients whether coming with gynaecological or obstetrical problems were satisfied with the treatment. i.e., out of 54 patients, 38 have surgical/medical measurement (2 awaiting, 30 good result, 3 fair, 2 tubal ligation, 1 poor), 5 awaiting, 2 defaulter, 9 others (DES, Diagnosed male, waiting for delayed puberty).

On the basis of outcome the above table depict the p value which is test of significance of the result is 0.03. (So, $p<0.05$).

Table-10: Accuracy of Ultrasonography to detect the congenital anomaly among 54 cases

Provisional Diagnosis	Cases	Percentages
True Positive	52	96.3%
False Positive	2	3.7%

It shows Ultrasonography is accurate upto 96.3% to detect the congenital anomaly.

Table-11: Prevalence of congenital anomaly at G.O.P.D. of R. G. K. M.C.H (from June 2006 – August 2007)

	Total Patient Attended at GOPD	Congenital Anomaly	Percentages
Gynaec. Prob.	20160	39	0.19%
Obst. Prob.	14932	15	0.1%

Prevalence of congenital anomaly at G.O.P.D. of R. G. K. M.C.H is 0.15 which is very near to value given in literature (0.16%;).[6]

Standard Error of estimation of prevalence is 6.25%.

DISCUSSION: The aim of the study was to search out the different congenital anomalies of female reproductive organ and to improve the qualities of life of those patients with different new treatment modalities. While performing the study for last fourteen months the author faced several problems owing to the poor economic status of the patients and socio cultural condition.

In fact, there was amalgamation of experiences with which the study reached to a point to refresh our previous experience about the incidence of these cases (May not be true reflection of actual prevalence or incidence); it also gave an idea about how to tackle those poor soul who are suffering from such structural, morphological, sexual and above all psychological ailments.

Malformations of uterus may not be stamped as rare as it is probable that many malformations are never diagnosed because many patients with such malformations are able to conceive and proceed to term without complication.[7]

In certain malformations surgical intervention is justified such as patient having habitual abortion, preterm delivery with no living issue and pent up menstrual discharge with/without infection due to failure of canalization.[8]

Mullerian duct anomalies encompass a wide spectrum of clinical and imaging findings, and while many of the anomalies will be diagnosed initially at HSG or two-dimensional US, further imaging with MR and, potentially, three-dimensional US will often be required for a definitive diagnosis.

DOI: 10.14260/jemds/2015/1749

Magnetic Resonance imaging (MRI) currently is the study of choice because of its high accuracy and detailed elaboration of uterovaginal and ovarian anatomy. Laparoscopy and hysteroscopy are then reserved for women, in whom interventional therapy is being undertaken, thus reducing health care expenditures and sparing women from invasive diagnostic procedures.

Ultrasonography imaging should be performed during the secretory phase of the menstrual cycle, when the endometrial thickness and echo complex are better characterized. US has a reported pooled accuracy of approximately 90%-92% Endovaginal US has advantage of improved spatial resolution, although at the expense of a decreased field of view.

Hysterosonography, with infusion of saline into the endometrial canal, provides improved delineation of the endometrium and internal uterine morphology;

Hysterosalphinogogram (HSG) is indicated in the early stages of evaluation of the infertile couple. The examination provides a morphologic assessment of the endometrial and the endocervical canals and supplies important information regarding tubal patency. Characterization of uterine anomalies can be difficult; notably with regard to differentiation of a septate from a bicornuate uterus. And the inability to evaluate the external uterine contour adequately HSG also entail exposure to ionizing radiation in these typically young women.

Three Dimensional Ultrasonography with surface and transparent mode reconstruction of the uterus has reported advantageous over conventional two dimensional scanning. In experienced hand, a sensitivity of 93% and specificity of 100% have been achieved.

Magnetic Resonance Imaging Technique has a reported accuracy up to 100% in the evaluation of mullerian duct anomalies. Although MRI is more expensive than US, its great accuracy makes it more trusted by many gynaecologist. MRI provides clean delineation of internal and external uterine conture. MRI depicts utero vaginal morphology, thus aiding in classification of anomalies.

The primary goals of surgical intervention[9] are to relieve obstruction and pain, to restore a normal sex life, and to preserve the patient's reproductive potential. The timing of surgery depends on the patient's anatomic configuration and on the presence or absence of functional endometrial tissue. In the patient with functional ovarian tissue but an absent uterus and vagina, reconstruction of the genital tract is not medically urgent.[10]

Variations in patients' anatomic findings and the lack of a standardized surgical technique complicate the outcomes and prognosis. One of the major limitations encountered in this study was the inability to make an etiological diagnosis for all patients. The author feels the imperative need of future research programme in order to find out the appropriate cause for these sets of patients. In every case one should always be careful about the psychological aspect of the patient; keeping this in mind it may be wiser not to explain the true fact to the patient.

REFERENCES:

1. Speroff L: The uterus. In: Mitchell C. ed. Clinical Gynaecologic Endocrinology & Infertility. 7th edition. Philadelphia, Pa Lippincott Williams & Wilkins; 2005:113.
2. Buttram VC Jr, Gibbons WE: Mullerian anomalies: a proposed classification. (An analysis of 144 cases 0. Fertil Steril 1979 Jul; 32(1):40-6.
3. Buttram VC Jr, Mullerian anomalies and their management. Fertil Steril 1983 Aug; 40(2):159-63.

DOI: 10.14260/jemds/2015/1749

4. The American Fertility Society classifications of adnexal adhesions, distal tubal obstructions, tubal occlusion secondary to tubal ligation, tubal pregnancies, mullerian anomalies and intrauterine adhesions. Fertil Steril 1988; 49:944-955.
5. Park, s Text Book of Preventive and Social Medicine. (18th ed.)2005. Ch18; p649-650.
6. Novak's Gynaecology. 13th Ed. By Jonatharn S. Berek. Uterine abnormalities. Espsm (pg-1010-1020).
7. Gray's Anatomy 39thed.1995:Ch.3.Embriology&Development. Williams, Warwick Dyson, Bannisher et al. (Pg204-217).
8. Te Linde's, Operative Gynaecology. 9th Ed. Ch.29 Surgery for Anomalies of Mullerian Duct by John A Rock Lessly L Breech (pg705-749).
9. Chakraborty BN & associates: congenital absence of the vagina and uterus-simaltaneous Vaginoplasty & hysteroplasty. Jobstet & gynecol (India) 2000, 27:627.
10. Munro Kerr's operative obstetrics (Tenth Edition), Ch.16 Dystosia Associated with Malformations of the uterus and vagina (Pg. 187-201).

AUTHORS:
1. Debjani Roy

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Anatomy, IPGME&R, Kolkata, West Bengal.

FINANCIAL OR OTHER COMPETING INTERESTS: None

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Debjani Roy,
Flat No. 2C, 2nd Floor,
Sakuntala Apartment, Kaikhali Ghosh Para,
Near VIP Road, P.O., Airport,
Kolkata-700052, West Bengal.
E-mail: anatomydoctor1975@gmail.com

Date of Submission: 22/08/2015.
Date of Peer Review: 23/08/2015.
Date of Acceptance: 25/08/2015.
Date of Publishing: 28/08/2015.