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Review Article

## MRKH syndrome: a review of literature

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### ABSTRACT

Primary amenorrhea is defined as failure to achieve menarche till age of 14 years in absence of normal secondary sexual characters or till 16 years irrespective of secondary sexual characters. The most common cause of primary amenorrhea is gonadal pathology followed by Mayer-Rokitansky-Küster-Hauser syndrome (MRKH syndrome). MRKH syndrome is a rare congenital disorder characterised by uterine and vaginal aplasia. It occurs due to failure of development of Müllerian duct. Its incidence is 1 per 4500 female births. Mostly girls present with primary amenorrhea. It is characterised by presence of normal secondary sexual characteristics, normal 46 XX genotype, normal ovarian function in most of the cases and absent or underdeveloped uterus and upper part (2/3) of vagina. It is of two types: type A is isolated type while type B is associated with other renal/skeletal/cardiac anomalies. Treatment includes psychological counselling and vaginoplasty. Vaginoplasty can be done by various non-surgical and surgical techniques. The authors hereby review the literature of MRKH syndrome regarding its embryology, etiopathogenesis, approach to work up and management.

**Keywords:** Amenorrhea, MRKH syndrome, Müllerian duct

### INTRODUCTION

MRKH syndrome is a rare congenital disorder characterised by uterine and vaginal aplasia. It occurs due to failure of development of Müllerian duct. Its incidence is 1 per 4500 female births.<sup>1-3</sup>

Typically, young girls with this syndrome presents with primary amenorrhea. It is characterised by normal secondary sexual characteristics, normal 46 XX genotype, normal ovarian function in most of the cases and absent or underdeveloped uterus and upper part (2/3) of vagina.

It is of two types: type A and Type B. Type A is an isolated variety in which uterus, cervix and upper 2/3 of vagina is absent while bilateral ovaries are normal. No other anomaly is associated. Type B is associated with various other anomalies such as renal anomalies, skeletal anomalies and cardiac anomalies. Bilateral ovaries might

be absent in type B variety. Most common anomaly in type B syndrome is renal anomalies.

Diagnosis is based on radiological findings. Ultrasound pelvis is the first investigation of choice. Absence of uterus and cervix is found on sonography. MRI pelvis is done to confirm the diagnosis. MRI abdomen is also done to rule out associated anomalies. Skeletal survey is done to rule out skeletal anomalies.

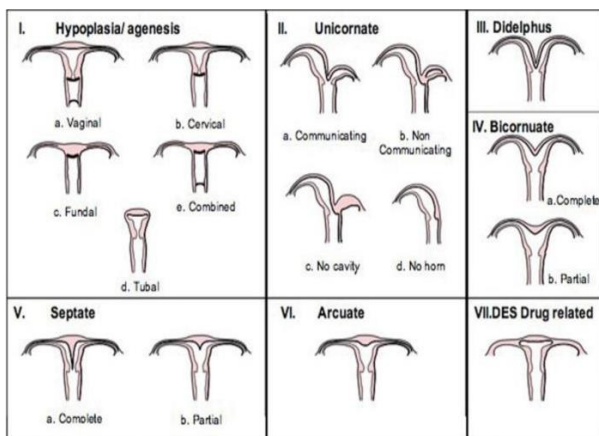
Management includes psychological counselling along with creation of neovagina. Neovagina can be achieved by non-invasive, non-surgical methods by serial dilatation of vagina with vaginal dilators. However, it can be done only if vaginal dimple is present. In absence of vaginal dimple/ failure of non-surgical method or patient refusal to dilators, surgery is done to create neovagina. Various techniques of vaginoplasty are available which are based either on traction or on grafts. Surgery can now be done laparoscopically also.

The authors hereby review the literature of MRKH syndrome regarding its embryology, etiopathogenesis, approach to work up, management and future prognosis about pregnancy.

## EMBRYOLOGY

The female reproductive tract develops from a pair of Müllerian ducts.<sup>4</sup> Following structures are derived from Müllerian duct: uterus, cervix, fallopian tube and the upper two-thirds of the vagina. The ovaries and lower third of the vagina have different embryological origins. Ovaries are derived from germ cells that migrate from the primitive yolk sac while lower one third of vagina is derived from sino-vaginal bulb. Normal development of the Müllerian ducts occurs in three phases: organogenesis, fusion and septal resorption. During first phase i.e. Organogenesis, bilateral Müllerian duct is formed. Failure of this phase results in agenesis or hypoplasia of uterus or unicornuate uterus. During second phase, fusion of Müllerian ducts leads to formation of uterus with a central septum. Failure of this step results in a bicornuate or didelphys uterus. Septal resorption is the third phase during which resorption of the central septum occurs. Failure of this stage results in a septate or arcuate uterus.

Based on the type of malformation, The American fertility association has classified Müllerian duct anomalies into 7 types as shown in Figure 1.<sup>5</sup> Of this, MRKH syndrome is class I Müllerian duct anomaly.



**Figure 1: Classification of Mullerian duct anomalies by American fertility society.<sup>5</sup>**

## ETIOPATHOGENESIS

Various theories have been proposed for its pathogenesis, which are as follows:

Girdhani et al has described that MRKH syndrome occurs due to deficiency of estrogen or gestogen receptors.<sup>6</sup> This deficiency leads to inhibition of development of Müllerian duct and hence prevents formation of structures which are derived from Müllerian duct.

Another theory was proposed by Pavanello et al who described the genetic basis of syndrome.<sup>7</sup> They found the genetic cause of MRKH syndrome especially type B, in which renal association is found in form of renal agenesis (unilateral/ bilateral). The gene was found to be single and autosomal dominant with variable expression.

Cramer et al proposed the biological basis of MRKH syndrome.<sup>8</sup> According to this theory, vaginal agenesis occurs due to decreased activity of Galactose-1-phosphate uridyl transferase enzyme (GALT). They found that fetal/ maternal mutation of GALT enzyme leads to its decreased activity which causes vaginal agenesis.

Nodale C et al explained the gene expression profile of patient with MRKH syndrome.<sup>9</sup> It was found that multiple genes are involved but two of them have strongest association which are HOXA gene and WNT-4 gene. HOXA-10 gene is associated with development of uterus, HOXA-11 gene with fallopian tube and cervix, HOXA-13 gene with vagina. Also, HOXA gene is found to be associated with development of kidney, bone and vascular structures, thus explaining the association of MRKH syndrome with other anomalies.

In another study by Bingham C et al and Linder TH et al, genital malformations such as bicornuate uterus, uterus didelphys and Müllerian aplasia were occasionally found associated with renal anomalies in some familial aggregates showing mutations within the TCF2 gene.<sup>11</sup> The TCF2 gene (formerly v-HNF1 or HNF-1  $\beta$ ) was originally found associated with MODY-type diabetes and with diabetes mellitus, renal cysts and other renal developmental disorders. So, Defects of this gene can thus account for some rare cases of Müllerian malformations associated with familial cases with history of diabetes or renal disorders.

## CLASSIFICATION OF MRKH SYNDROME

Müllerian duct anomalies have been classified into different forms. Following are the classifications.

The American fertility association has classified Müllerian duct anomalies into 7 types as shown in Figure 1.<sup>5</sup> Of this, MRKH syndrome is class I Müllerian duct anomaly. It is further classified into:

### Type A

Isolated syndrome characterised by absent uterus and upper two third of vagina with normal bilateral ovaries and no associated anomalies.

### Type B

Associated with ovarian agenesis, renal anomalies, skeletal abnormalities, cardiac defects and hearing/ ocular anomalies. Renal anomalies can be renal agenesis,

ectopic kidney, hyperplasia, malrotation or horse shoe kidney.

In study by Oppelt et al, associated malformations were found in almost 50% of patients, of which renal and skeletal complications were most frequent.<sup>12</sup> Renal anomalies were found in 30% of cases of which most common was renal agenesis.

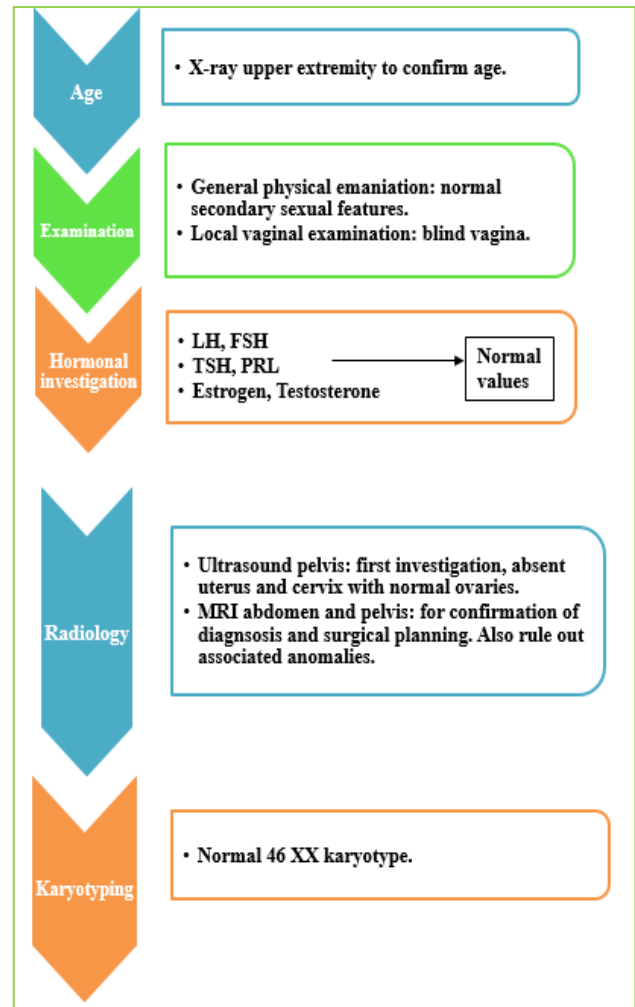
Tarry and Duckett has classified Müllerian duct anomalies according to physical findings, ultrasound report and findings on laparoscopy.<sup>13</sup> Grade 0-4 was assigned referring to extent of Müllerian duct system affected. Each side is graded separately. Letter 'M' was used to refer Müllerian duct. Following is the grading:

- M0: unilateral system normally formed but unfused/retained septum.
- M1: vaginal agenesis alone. Uterus normal.
- M2: vaginal and uterine agenesis.
- M3: total Müllerian agenesis.
- M4: Müllerian and ovarian agenesis.

**APPROACH TO WORK UP**

Typically, a young girl presents to OPD with complaint of primary amenorrhea. Primary amenorrhea is defined as failure to achieve menarche till age of 14 years in absence of normal secondary sexual characters or till 16 years irrespective of secondary sexual characters.<sup>14</sup> The most common cause of primary amenorrhea is gonadal pathology followed by MRKH syndrome.<sup>15</sup> First of all, in such girls, age should be confirmed. X ray of upper extremity is done for age confirmation. Secondly, general physical examination and local vaginal examination is done to look for secondary sexual characteristics and vaginal opening respectively. In MRKH syndrome, usually all secondary sexual characteristics are normal and blind vagina is found.

After history and examination, investigations are done including endocrine test, radiological evaluation and karyotyping study. Endocrine evaluation includes hormonal profile i.e. LH, FSH, Estrogen, Testosterone, TSH and prolactin. Usually, in MRKH syndrome, hormonal profile is normal. On radiological evaluation, ultrasound is done on which absent uterus and cervix is found with mostly normal bilateral ovaries. For confirmation of diagnosis and surgical planning, MRI abdomen and pelvis is the investigation of choice. On MRI, most common finding is absent uterus and cervix with absence of upper 2/3<sup>rd</sup> of vagina. Bilateral ovaries and lower vagina are found to be normal. MRI abdomen helps to see the associated anomalies. Skeletal survey is done to rule out skeletal anomalies. On karyotyping, normal 46 XX karyotype is found in these girls. In 1982, Smith et al reported that MRKH girls have normal 46 XX genotype.<sup>16</sup> Similar findings were reported by Cabra et al and Orozo Sanchez et al.<sup>17,18</sup> The whole approach is summarised in Figure 2.



**Figure 2: Flowchart showing approach to work up the patient of primary amenorrhea.**

**DIFFERENTIAL DIAGNOSIS**

The immediate differential diagnosis of MRKH syndrome is androgen insensitivity syndrome (AIS). AIS occur due to inactivating mutation of androgen receptor thus, leading to absence of development of male characters. In AIS, young girl presents with primary amenorrhea with scarce pubic and axillary hair growth. On investigation, no uterus, cervix and ovaries are seen, rather intraabdominal testicles are found. On karyotyping, genotype 46 XY is found. In AIS, surgery needs to be done to remove gonads due to risk of gonadal malignancy.

**ROLE OF LAPAROSCOPY**

Laparoscopy is usually not required for diagnosis of these cases, since it is expensive and invasive. However, in cases where patient presents with pelvic symptoms, diagnostic laparoscopy is done. Surgical treatment in form of vaginoplasty is now-a-days is combined with laparoscopy procedures. Recently, authors encountered a case of a young 17-year-old girl, who presented to our

OPD with complaint of primary amenorrhea. Complete work was done, and she was diagnosed with MRKH syndrome type A with an ovarian cyst. Patient and family were counselled and advised surgery for cystectomy. Vaginoplasty was advised at a later age.

However, parents wanted to get combined surgery of cystectomy and vaginoplasty thus took the decision of surgery at later age only. But, after few months, she presented to emergency department with complaint of acute pain in lower abdomen. On MRI, huge thin walled complex ovarian cyst of size 97 x 87 x 93 mm was found (Figure 3).

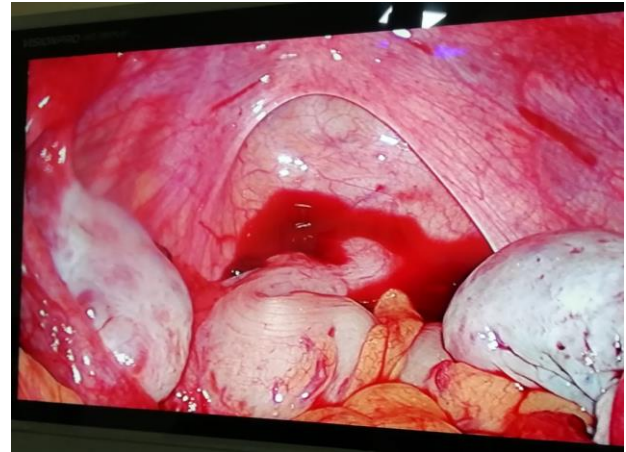


**Figure 3: Image of MRI pelvis showing absence of uterus, cervix and vagina along with presence of huge ovarian cyst.**

Laparoscopy was done, on which huge ovarian cyst was found with torsion (Figure 4 and 5). Ovary was detorted, and cystectomy was done.



**Figure 4: Image on laparoscopy showing huge ovarian cyst.**



**Figure 5: Absence of uterus and cervix with normal bilateral ovary after cystectomy.**

## MANAGEMENT

Management of MRKH syndrome includes both psychological counselling and surgical treatment. When a young girl is diagnosed with such syndrome, it is a big psychological distress to both patient and the whole family. Thus, it is recommended that patient and family should be counselled before and after treatment. Group programs are also helpful for further re-assurance.

### *Non-surgical treatment*

Non-surgical management includes serial dilatation with vaginal dilators. Different techniques have been described, of which most commonly used technique is Frank's technique.

Frank's described a method in which vaginal dilatation is done by vaginal mould daily for 20 minutes by patient herself.<sup>19</sup> Progressively, length and width of dilator is increased. Usually, six months is required to reach functional depth and width.

Ingram described a method in which progressive dilators are attached on a bicycle seat and patient is asked to provide perineal pressure by sitting and slightly leaning forward.<sup>20</sup>

Few disadvantages associated with these methods are urethritis, cystitis, vesico-vaginal fistula formation or secondary prolapse. But being a non-invasive method with high success rate (78% to 92%), it is recommended as a first line therapy.<sup>21,22</sup> However, it can be done in only those cases where vaginal dimple of at least 2-4 cm length is present.

### *Surgical treatment*

Various surgical techniques have been described. These techniques are of two types:

- a. Traction based: Vecchiotti procedure (laparoscopic/open)
- b. Graft based: Abbe-McIndoe technique (vaginal approach, skin grafting), Intestinal vaginoplasty (combined vaginal and open/laparoscopic approach), Davydov technique (combined vaginal and laparoscopic approach, peritoneal graft).

In Vecchiotti procedure, laparoscopically, an olive is attached to the vaginal dimple, the thread attached to this olive is passed to the female pelvis via vesico-rectal space, and then taken out through anterior abdominal wall, thereby connected to the traction device.<sup>23</sup> Thus, traction is giving to olive which helps in creating new vagina. Slowly, this traction is increased day by day. Once functional length of 7-8cm is achieved, the olive is removed, and girl is advised to either start intercourse or use vaginal dilators to maintain the length. Since this procedure requires very careful dissection, so it is done in only surgically naive tissue.

In graft-based techniques, a graft is used to create neo vagina. This graft can be skin graft, bowel segment or a bioengineered tissue. Various techniques obtain graft from different sites.

The Abbe-McIndoe technique is the most commonly used technique for vaginoplasty.<sup>24</sup> In this technique, after creating a space between bladder and rectum, vaginal mould is placed. This vaginal mould is placed by putting skin graft over a vaginal stent. Split thickness tissue graft is obtained from anterior thigh or buttock. After healing of tissues, vaginal dilators are used, or intercourse is done to maintain the vagina.

In intestinal vaginoplasty, segment of sigmoid colon, ileum or jejunum is obtained via open or laparoscopic approach. The intestinal tissue provides lubrication. The disadvantage of this method is excessive vaginal discharge can occur and secondly, obtaining intestinal tissue via bowel resection and anastomosis increases the morbidities.<sup>25</sup>

In Davydov procedure, an autologous peritoneal graft is used for vaginoplasty.<sup>26</sup> Laparoscopically, peritoneum graft from pouch of Douglas is dissected and mobilised. After creating vaginal space, peritoneum is reached and then mobilised peritoneal sac is opened and pulled downward to connect the vaginal epithelium with graft taken from peritoneum of pouch of Douglas.

Various studies have been done to compare results of surgical and non-surgical techniques and also compare different surgical procedures. These are the results of various studies:

Morcel K et al did a study to compare sexual and functional results after creating neovagina via non-surgical and surgical techniques in girls with MRKH syndrome.<sup>27</sup> Out of 40 women who enrolled in the study,

20 were treated by Frank's method while other 20 were managed by surgical techniques (12 cases via sigmoid vaginoplasty and 8 cases via Davydov procedure). It was found that functional sexual outcomes were similar after surgical and non-surgical techniques. Thus, it was concluded that Frank's method should be proposed as first line therapy because it is less invasive than surgical procedures. Surgical reconstruction should be done in the case of failure of non-surgical technique or of refusal by the patient.

A study was done by Willemsen WN et al, to compare long term results after nonsurgical and surgical treatment (Davydov procedure) of vaginal agenesis in MRKH syndrome.<sup>28</sup> It was found that long-term outcomes of both Frank dilation and Davydov procedure in experienced hands were good. However, when surgical management was preceded by non-surgical Frank's dilatation method, no significant difference was found. The major complications found after surgical therapy were formation of granulation tissue (23%) and tendency to obliterate neovagina (12%).

In a study done Ding JX et al, comparison was done between laparoscopic peritoneal vaginoplasty (Davydov procedure) and acellular porcine small intestinal submucosa (SIS) graft.<sup>29</sup> 34 patients were operated via SIS graft and 41 patients underwent Davydov procedure. It was found that the operating time and estimated per-operative blood loss was significantly lesser while using SIS graft. However, SIS graft is very costly (\$3525 per graft), thus significantly increasing the cost of surgery. Overall, it was concluded that vaginoplasty using SIS graft is successfully achieved in women with MRKHS, and the anatomical and functional outcomes of this procedure are comparable to laparoscopic peritoneal vaginoplasty.

Kuessel L et al published a study in which neovagina was created in a 20-year-old woman with MRKH syndrome, by using Wharton-Sheares-George method.<sup>30</sup> In this technique, the rudimentary Müllerian ducts were dilated incrementally by pushing Hegar dilators in the direction of the pelvic axis, and the resulting median raphe was then intersected using diathermy. Subsequently, a vaginal mold is inserted into the newly created cavity and held in position by two sutures. A mean vaginal length of 8.3 cm and width of 3.3 cm was achieved. No major Intraoperative and post-operative complications or prolapse were reported till date. Overall, it was concluded that Wharton-Sheares-George method of vaginoplasty is a minimally invasive, quick and safe surgical option that does not require allogenic or autologous transplants, nor does it require traction devices or specialized surgical equipment and provides anatomically and functionally successful outcomes.

Callens N et al gave an update on surgical and non-surgical treatment for vaginoplasty.<sup>19</sup> A research of all published literature was conducted from 1898 to March

2013 using Pubmed, Cochrane Library and Web of Science. A total of 190 studies are analysed. It was found that when anatomical success was defined as a length of  $\geq 7$  cm and functional success as coitus, all vaginoplasty techniques yielded significantly higher success rates ( $>90\%$ ) as compared to non-surgical techniques (75%). However, overall complication rates were significantly lower within the vaginal dilation groups when compared with the different vaginoplasty techniques. Thus, vaginal dilation recommended as first choice treatment was found to be justified. Among surgical techniques, Traction vaginoplasty seems to have the highest anatomical (99%) and functional success rates (96%), whereas both split- and full-thickness skin graft procedures and intestinal procedures have the lowest successful outcomes (83-95%). Thus, the laparoscopic Vecchietti procedure is considered an appropriate surgical option in patients who are poorly compliant and had failed dilation therapy, or for those who do not want to start with vaginal dilation therapy.

### FUTURE ISSUE OF PREGNANCY

Girls with MRKH syndrome have two issues of major concern, one is about sexual activity and other is future pregnancy. The sexual activity is achieved by creating neovagina by either non-surgical or surgical techniques as described in management segment. However, another matter of concern is to bear children. In women with normal ovarian function, pregnancy can be achieved by new fertilization techniques i.e. in-vitro fertilization followed by embryo transfer in surrogate mother. The response to treatment in terms of number of oocytes retrieved, fertilization rate, embryo quality and pregnancy rate has been reported to be slightly lower than average in comparison for infertile patients.<sup>31</sup>

### CONCLUSION

MRKH syndrome is a rare congenital disorder characterized by uterine and vaginal aplasia. Typically, presentation is primary amenorrhea with normal secondary sexual characters, blind vagina, absence of uterus and cervix on ultrasonography with normal 46 XX karyotype. Patient and family need urgent psychological counselling. Neovagina is firstly created by serial vaginal dilators. In failure of non-surgical techniques or women refusal to do so, vaginoplasty is done by either open/laparoscopic techniques. Pregnancy can be achieved by using latest in vitro fertilization techniques and surrogacy.

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