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Magnetic resonance imaging in patients of Mayer–Rokitansky–Kuster–Hauser (MRKH) syndrome presenting with primary amenorrhoea: a prospective study

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Abstract

Background This study aims to characterize the spectrum of imaging findings in patients of Mayer–Rokitansky–Kuster–Hauser (MRKH) syndrome presenting with primary amenorrhoea.

Objectives (1) To aid in clinical diagnosis of MRKH syndrome. (2) To detect the associated non gynecological anomalies. (3) To arrive at optimal management options.

Results This prospective observational study was performed in 14 subjects who presented with primary amenorrhoea and diagnosed as MRKH on MRI from 1st January, 2021 to 30 June, 2022. Out of 14 patients, 4 patients showed hypoplastic uterus while remaining 10 showed complete uterine agenesis. Mean volume of hypoplastic uterus was 5.3 ml. Out of 10 patients with complete uterine agenesis, 9 showed bilateral rudimentary uterine buds connected by a fibrous band while 1 showed unilateral bud. 3 uterine buds showed three layered zonal differentiation of which 2 showed blood within lumen suggesting functioning endometrium. 1 uterine bud was located outside pelvis. Bilateral ovaries were noted in all 14 patients of which 4 ovaries were located outside pelvis and 1 ovary showed endometrioma within. 10 patients showed complete agenesis of proximal 2/3rd of vagina. Among non-gynecological findings, 2 showed unilateral renal agenesis and 1 showed unilateral hypoplastic kidney. 1 patient showed dorsal dermal sinus.

Conclusions The findings in MRKH patients varies from complete uterine agenesis with rudimentary uterine buds or normal positioned hypoplastic uterus. The uterine buds can be functional with collection within endometrium and could lead to endometrioma formation due to retrograde menstruation. Ovaries and uterine buds can have an extrapelvic location.

Keywords Mullerian agenesis, Primary amenorrhoea, Paramesonephric duct, Uterine bud, Functional endometrium, Zonal differentiation of uterus

Background

MRKH (Mayer–Rokitansky–Kuster–Hauser) syndrome which is also known as mullerian duct agenesis is a rare congenital condition occurring due to interrupted normal in utero development of paramesonephric duct. It is characterized by aplasia or hypoplasia of structures developing from mullerian duct comprising of uterus and upper two third of vagina. Fallopian tubes and ovaries show normal development in almost all cases [1]. These

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females are XX karyotypically. As secondary sexual characters are normally developed in these patients, they go undetected until late adolescence when they present with primary amenorrhoea. It has been classified as class I mullerian duct anomaly according to American Fertility society [2] and as mullerian agenesis according to ASRM (American Society for Reproductive Medicine) mullerian anomalies classification 2021 [3]. It has as incidence of around 1 in 4500 female live births [4]. This syndrome is classically categorized into two different types, type I or typical MRKH and type II or atypical MRKH on the basis of absence or presence of associated upper urinary tract, skeletal, middle ear or cardiac anomalies respectively [5].

The clinical information of primary amenorrhoea with normal secondary sexual characteristics can be used to reach to a definite diagnosis with the help of imaging findings. Ultrasound and MRI of pelvis are the noninvasive preoperative diagnostic techniques [6]. USG pelvis is the first line investigation. The findings can be further confirmed on MRI. Gold standard investigation for the diagnosis of MRKH is diagnostic laproscopy.

MRKH greatly affects the psychological health of patients given its impact on sexual relations and fertility therefore the therapeutic methods aim at managing them [7]. The American College of Obstetricians and Gynaecologists lays stress on non-surgical approach for primary management of these patients. The first line treatment is vaginal dilator. As these patients have functional ovaries, transvestibular vaginoplasty with pelvic peritoneum is considered as a desired therapeutic option given that surrogacy or assisted reproductive techniques are used for fertility [8].

Methods

The study was carried out after getting approval from the institute's ethical committee (IEC). A detailed observational study was done in 14 patients coming with chief complaints of primary amenorrhea. These patients were selected after being clinically and pathologically examined for normal secondary sexual characteristics (normal axillary and pubic hair and breast development with respect to age), normal hormonal levels (FSH, LH, AMH), and XX karyotype.

These were then evaluated ultrasonographically by SAMSUNG H60 Ultrasound Machine using 5 MHz frequency curvilinear probe in the supine position. All these patients were then sent for MRI pelvis. A detailed and informed consent was taken before undergoing MRI.

MRI was done on 1.5 Tesla superconducting machine, GE (Optima MR360, United States). The sequences which were taken were T1 Fast spin echo (FSE) in axial plane, T2 FSE axial, coronal and sagittal planes after application of phased array pelvic coil. T1 FSE images were obtained at TR: 800–900 ms and TE: 10-12 ms at FOV of 180–200 mm. T2 FSE images were obtained at TR: 4800–5800 ms and TE: 85-100 ms at FOV of 180–200 mm, Slice thickness was taken as 4 mm for these images with slice interval of 1 mm.

Abdominal axial T2 SS FSE sections of slice thickness 5 mm and slice interval 1.5 mm after proper breath-hold were also taken specially to look for associated renal anomalies. Intravenous or local contrast was not used.

The MRI images were assessed for the mullerian structures including uterus, vagina and ovaries.

Uterus was evaluated for its location in pelvis with respect to midline, volume of the hypoplastic uterus was calculated in millilitres (ml), zonal differentiation into myometrium, functional zone and endometrium was looked for, and endometrial cavity was looked for any collection.

If absent, the uterine buds were evaluated for its presence, differentiation into different layers, volume in ml, location in pelvis and relative position with respect to ovaries. The presence of retrovesicle triangular cord sign and converging band between the two uterine buds was also noted.

The ovaries were assessed for their presence, location (pelvic or extrapelvic), relative position with respect to each other, volume in ml and for the presence of any dominant follicle.

The vagina was assessed for length, and for the presence of upper 2/3rd and lower 1/3rd parts separately.

Detailed evaluation of abdominal sections were done to look for associated other anomalies like renal and skeletal anomalies.

Statistical analysis

The data were transferred to a Microsoft Excel 2010 sheet and statistical analysis was done using IBM SPSS for Windows version 22.0 (SPSS, Inc., Chicago, IL, USA).

Results

Mean Age of patients in the study was 18 years (Fig. 1).

Assessment of uterus and uterine buds (Table 1).

Among the 14 patients who were studied, hypoplastic uterus was present in 4 patients of which 3 were noted in midline in pelvis, while 1 was located off midline, slightly towards right. Three layered zonal differentiation of hypoplastic uterus was noted in 2 patients (Fig. 2) while the other 2 showed absence of any zonal differentiation. None of these showed any collection in endometrial cavity.

Remaining 10 patients showed complete agenesis of uterus and proximal 2/3rd of vagina. Bilateral uterine buds were noted in 9 of 10 patients (Fig. 1) while 1 patient showed only one uterine bud showing three

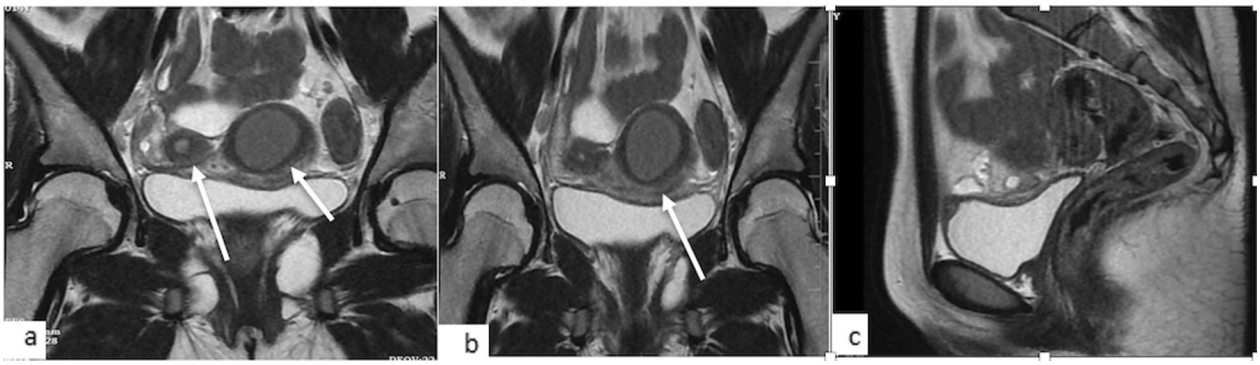

Fig. 1 **a**–**c** A 19-year-old female coming with primary amenorrhoea **a** T2 weighted coronal image showing bilateral functional uterine buds in pelvis with collection within endometrium (straight arrows). **b** T2 weighted coronal image showing converging band between the two buds (straight arrow). **c** T2 weighted sagittal image showing non visualization of normal uterus and upper two-third of vagina in midline

layered zonal differentiation within it (Fig. 4). This bud was located high up in false pelvis. One of the patients showed zonal differentiation in both uterine buds with hyperintense collection within (Fig. 1). Mean volume of hypoplastic uterus was 5.37 ml. Mean volume of right uterine bud was 3.63 ml ± 1.689 (SD). Mean volume of left uterine bud was 8 ml ± 11.01 (SD).

Converging band connecting the uterine buds was noted in all 9 patients with complete agenesis of uterus (Figs. 1, 3). Also retrovesicle triangular cord was observed in 5 of 9 patients with complete agenesis of uterus (Fig. 3).

While most of the patients showed constant caudal relation of uterine buds with respect to ovaries, 1 patient showed cranial relation of both uterine buds and 2 patient showed cranial relation of one of the buds (Fig. 3).

Assessment of ovaries (Table 2).

Two ovaries were noted in all 14 patients, of which left ovary was extra-pelvic in 3 patients and right ovary in 1 patient. One of the patients who had right hypoplastic uterus showed both ovaries towards right side in pelvis. 3 patients showed dominant follicles. One of the patients with zonal differentiation of left uterine bud showed multiple endometriomas in left ovary (Fig. 4) while the other showed multiple simple cysts in left ovary.

Assessment of vagina (Table 3).

The nine patients with complete agenesis of uterus showed absent upper two-third of vagina. Remaining four patients with hypoplastic uterus showed presence of complete vagina. Mean length of vagina was 2.80 cm ± 1.01 (SD).

Presence of associated anomalies (Table 4).

Mullerian agenesis is also associated with other non-gynaecological anomalies like urinary tract anomalies. In our study, three patients had associated renal tract anomalies and one patient also had Dorsal dermal sinus (Fig. 4).

Discussion

MRKH is a rare congenital condition in which mullerian ducts fail to develop normally. The aetiology of it is still not clear and is thought to be multifactorial [9]. It classically presents as hypoplastic uterus or aplastic uterus with rudimentary Mullerian remnants as was noted in most of our patients. The diagnosis of MRKH is of exclusion, after the endocrine and genetic work up of patients are normal. Transabdominal sonography of pelvis is the first line imaging method for its diagnosis while MRI is the noninvasive imaging modality of choice and confirms the diagnosis when USG findings are inconclusive or equivocal.

Hypoplastic uterus was noted in 10 of 14 patients which is in contrary to the misconception that there is complete agenesis of uterus in MRKH patients. This has also been described in many previous literatures as stated by Hall-Craggs et al. [10]. However these uterus lack the typical zonal differentiation and even if three layer differentiation is present they lack the fertility potential [11]. No case of hypoplastic uterus was reported by Yoo et al. and Wang et al. in their studies [11, 12].

Those patients with complete agenesis of uterus typically show bilateral rudimentary uterine buds connected by a fibrous band [12]. These are identified as solid round to oval isointense structures on T2WI. Bilateral uterine buds were noted in 9 of total patients while only 1 showed unilateral bud. Out of 19 uterine buds, 16 showed no zonal differentiation while 3 of the buds showed three layered zonal differentiation within giving a target pattern of which 2 showed hematometra within. The occurrence of cavitation in uterine buds suggest functioning

Table 1 showing the data of patients with assessment of uterus, uterine buds, zonal differentiation, presence of absence of converging band and retrovesicle cord sign

S. No.	Age (year)	Uterus (if present, volume in ml)	Location of uterus in pelvis	Zonal differentiation of hypoplastic uterus	Right uterine bud (volume in ml)	Left uterine bud (volume in ml)	Zonal differentiation of uterine buds	Collection within uterine buds	Converging band connecting uterine buds	Retrovescicle triangular cord sign
1	19	A	–	–	P (3.37 ml)	P (3.8 ml)	A	A	P	A
2	17	A	–	–	P (5.11 ml)	P (4.7 ml)	A	A	P	P
3	18	A	–	–	P (2.8 ml)	P (2.69 ml)	A	A	P	P
4	16	A	–	–	P (0.99 ml)	P (1.88 ml)	A	A	P	A
5	16	A	–	–	P (1.4 ml)	P (3 ml)	A	A	P	P
6	16	P (Hypoplastic, 9.1 ml)	Midline	P (3 layers)	A	A	–	–	–	–
7	20	A	–	–	P (5.8 ml)	P (2.3 ml)	A	A	P	P
8	18	A	–	–	P (4.5 ml)	P (2.8 ml)	A	A	P	P
9	26	A	–	–	P (3.5 ml)	P (3.2 ml)	A	A	A	A
10	15	P (Hypoplastic, 1 ml)	Midline	A	A	A	–	–	A	A
11	19	P (Hypoplastic, 6 ml)	Midline	P (3 Layers)	A	A	–	–	A	A
12	18	P (Hypoplastic, 5.4 ml)	Towards right	A	A	A	–	–	A	A
13	20	A	–	–	A	P (20.7 ml)	P (left uterine bud)	A	A	A
14	19	A	–	–	P (5.2 ml)	P (35 ml)	P (both right and left)	P (right-0.14 ml, Left-12.3 ml)	P	A

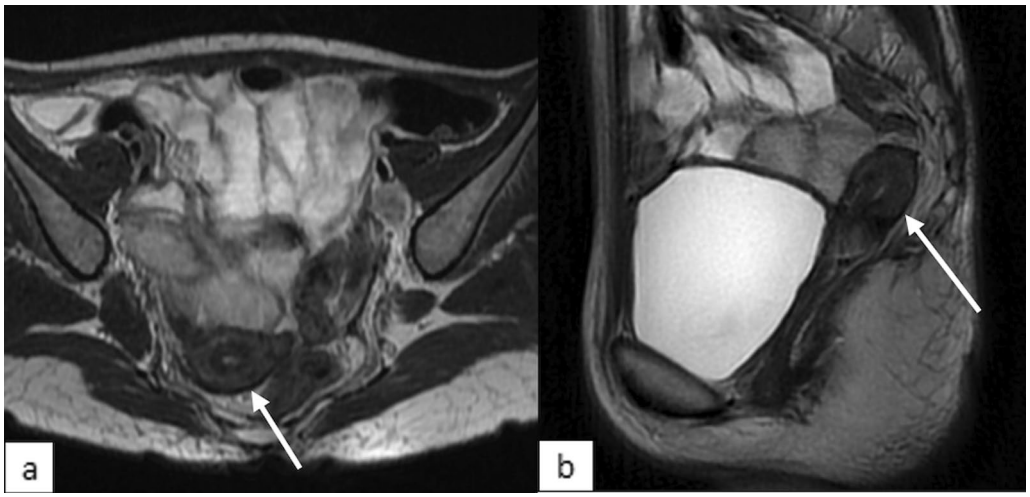

Fig. 2 **a**, **b** A 16-year-old female presenting with primary amenorrhoea (**a**) and (**b**) Axial and sagittal T2 WI showing normal positioned hypoplastic uterus with three layered zonal differentiation (straight arrows)

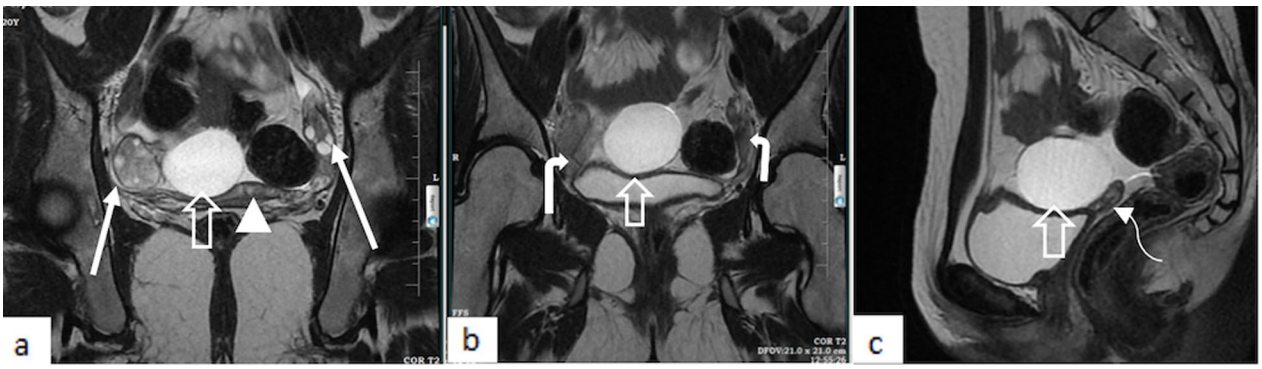

Fig. 3 **a**–**c** A 16-year-old female with primary amenorrhoea. **a**, **b** coronal T2 WI showing bilateral normal ovaries in pelvis [Straight arrows in (**a**)]. Bilateral non differentiated uterine buds located inferiorly and medially with respect to ovaries {curved arrows in (**b**)}. A converging band can be seen between them (arrowhead). A midline cystic structure can be seen which was considered to be collection in mullerian remnant [Block arrows in (**a**), (**b**) and (**c**)]. (**c**) Sagittal T2 WI showing triangular cord sign in retrovescicle location (S shaped arrow)

endometrium. These functioning endometrium cause these patients to present with cyclical pelvic pain in post pubertal period [9]. This is also important as this a criterion for surgical removal of these buds. Therefore knowledge about the location of uterine buds is essential for surgical approach. Two of buds were located extra-pelvically. This was also noted earlier [10, 13].

In contrast to Boruah DK [13] and Hall-Craggs et al. [10], the uterine buds showed a constant caudal relationship to ovaries [8], this constant relationship was not noted and the uterine buds were seen to be located superior, inferior or at the same level as ovaries. Similar findings were also described by Yoo et al. [11].

A connecting band was noted in all nine patients showing bilateral uterine buds seen to connect these buds. It is seen as iso to hypointense linear structure on T2 weighted images better appreciated on coronal sections. These formed a retro vesicle small triangular structure in most of these patients described as retro vesicle triangular cord sign. This is seen as a T2 hypointense structure in midline posterior to urinary bladder in upper part better appreciated on sagittal sections.

All 14 patients showed normal development of bilateral ovaries suggestive of different embryonic development of ovaries and uterus. The ovaries are better appreciated on T2 weighted images with follicles appearing hyperintense in hypointense stroma. Four ovaries were located in extra-pelvic location. Extra-pelvic location of some ovaries in MRKH patients was noted in most of the previous literatures [6, 10–13] suggesting increased incidence

Table 2 showing the data of patients with assessment of location and size of ovaries and presence of any ovarian pathology

S. No.	Age (year)	Right ovary (volume in ml)	Location and side of right ovary	Left ovary (volume in ml)	Location and side of left ovary	Relative position of ovaries with respect to each other	Presence of ovarian pathology
1	19	5	Pelvis, R	3.7	Pelvis, L	Same	–
2	17	2.4	Pelvis, R	8.2	Extra pelvic, L	Different	–
3	18	12.5	Pelvis, R	13	Pelvis, L	Same	–
4	16	10.6	Pelvis, R	11.6	Pelvis, L	Different	–
5	16	8.89	Pelvis, R	3.53	Pelvis, L	Same	–
6	16	1.6	Pelvis, R	5	Pelvis, L	Same	–
7	20	9	Pelvis, R	4.1	Pelvis, L	Same	–
8	18	14.4	Pelvis, R	9	Pelvis, L	Same	–
9	26	9.2	Pelvis, R	21	Extra pelvic, L	Different	–
10	15	2.3	Pelvis, R	3.6	Pelvis, L	Same	–
11	19	3.5	Pelvis, R	6	Pelvis, L	Same	–
12	18	2.8	Pelvis, R	10	Pelvis, R	Same	–
13	20	12.6	Extra pelvic, R	46	Extra pelvic, L	Same	Endometrioma left ovary
14	19	9.1	Pelvis, R	22	Pelvis, R	Same	Simple cyst left ovary

Mean volume of right ovary was 7.42 ml ± 4.38 (SD)

Mean volume of left ovary was 11.909 ml ± 11.51 (SD)

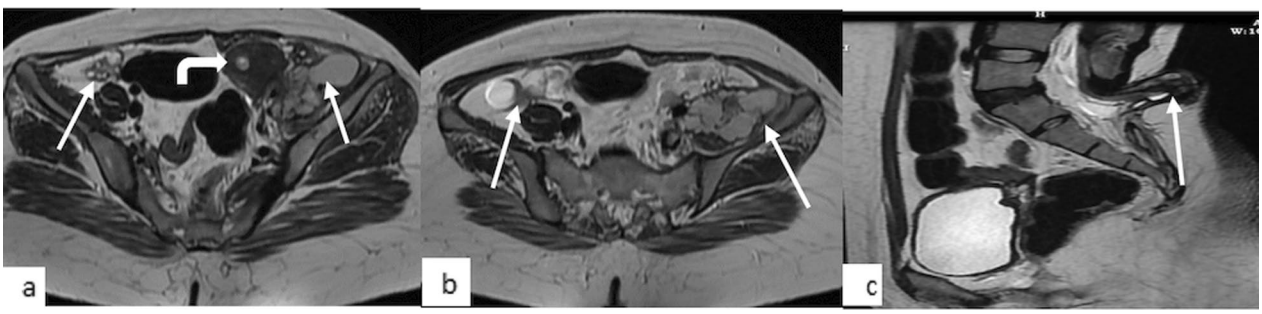

Fig. 4 (**a**–**c**) A 20-year-old female with type II MRKH syndrome (**a**), (**b**) Axial T2 WI showing unilateral left uterine bud in extrapelvic location [Curved arrow in (**a**)]with three layered differentiation. Bilateral ovaries can be seen in extrapelvic location with left ovary containing endometriomas [Straight arrows in (**a**) and (**b**)]. **c** T2 sagittal image showing sacral dorsal dermal sinus (Straight arrow)

of abnormal location of ovaries in MRKH syndrome [14]. The description of this fact is important in relation to fertility management [14]. One of the patients showing zonal differentiation of uterine bud showed multiple endometriomas in same sided ovary suggesting functioning endometrium in mullerian remnant causing reversed flow of menstrual blood as stated by Kawano et al. [15]. Similar finding was also seen in study by Reinhold et al. [16].

A midline cystic structure posterior to uterus was noted in one of our cases. Similar finding was described by Hall-Craggs et al. [10] in which on laparoscopy, it was proved to be proteinaceous material filled in mullerian remnant; however, no laparoscopic follow-up was done in our patient.

MRI is also an appropriate imaging tool for the diagnosis of associated anomalies in MRKH patients. Renal and vertebral anomalies are the most common non-gynaecological anomalies seen in association with MRKH syndrome. Among the renal anomalies, unilateral renal agenesis is the most common occurrence [9, 17, 18]. Two patients showed agenesis of one of the kidneys and 1 of them showed small one kidney with normal other kidney. No case of vertebral anomaly was noted in our study. One patient showed spinal dysraphism as sacral dorsal dermal sinus.

Diagnostic laproscopy is the gold standard investigation for MRKH.

Management of MRKH patients is primarily important for establishing the normal sexual functions which

Table 3 showing the length of vagina in 14 subjects

S. No.	Age (years)	Length of vagina (cm)
1	19	1.9
2	17	2.5
3	18	2.1
4	16	2.1
5	16	2.4
6	16	4.7
7	20	2.2
8	18	2.0
9	26	2.4
10	15	4.1
11	19	4.3
12	18	4.2
13	20	2.1
14	19	2.3

Table 4 tabulates the associated anomalies of urinary tract or spine in 14 subjects

S. No.	Age (years)	Associated anomalies
1	19	–
2	17	Absent right kidney
3	18	–
4	16	–
5	16	–
6	16	–
7	20	–
8	18	Right renal agenesis
9	26	–
10	15	–
11	19	–
12	18	–
13	20	Small left kidney, dorsal dermal sinus
14	19	–

is done by creation of a good anatomical and functional neovagina. This can be done either non-surgically by vaginal dilatation or surgically by various types of vaginoplasties. Vaginoplasty can be done using various autografts like bowel, peritoneum or labia majora [19].

Vaginal dilatation is the first line of management as stated by American college of Obstetricians and Gynecologists (ACOG) which also states that surgery should only be done in those cases who have failed the dilatation method.

Uterine transplantation has now emerged as the first true infertility management for these patients [20].

Surgical excision of uterine buds is done in cases of functional endometrium with obstructive bleeding as these lead to typical cyclical or atypical abdominal pain due to accumulation of haemorrhagic content within them. In other asymptomatic cases or those with unobstructive bleeding surgery may not be needed [21].

Limitations

- The present study was conducted in a sample of limited size. Thus future studies are required on more number of patients in order to generalize the results.
- There was a selection bias to typical cases as not every patient underwent imaging.
- No interobserver agreement for MR images evaluation was calculated.
- The study was limited to our hospital centre only.
- Laparoscopic correlation of the imaging findings was not done as some of the patients denied the investigation or rest were lost to follow-up.

Despite these limitations, this study was able to clearly confirm the diagnosis of MRKH ruling out other causes of primary amenorrhoea and also delineates the spectrum of findings in MRKH patients both related to mullerian remnants as well as the most of the associated non-gynaecological anomalies thus helping in managing these patients. Also this study is an add on to the previous studies with different patients presenting with primary amenorrhoea in this region.

Though laparoscopic correlation of the imaging findings was not done, findings were still similar to those studies in which the correlation was done as by Pomili et al. [22].

Conclusions

We conclude that MRI is the investigation of choice in patients who are suspected to have Mullerian anomalies. The findings in MRKH patients varies from complete uterine agenesis with rudimentary uterine buds or normal positioned hypoplastic uterus. The uterine buds can be functional with collection within endometrium and could lead to endometrioma formation due to retrograde menstruation. Ovaries and uterine buds can have an extra-pelvic location. Associated genitourinary and skeletal anomalies can also be diagnosed in the same sitting.

Abbreviations

MRI	Magnetic resonance imaging
MRKH	Mayer–Rokitansky–Kuster–Hauser
ASRM	American Society for Reproductive Medicine
LH	Luteinising hormone
FSH	Follicle stimulating hormone
AMH	Anti-mullerian hormone

FSE	Fast spin echo
SS FSE	Single shot fast spin echo
ml	Millilitres
SD	Standard deviation
P	Present
A	Absent
R	Right
L	Left

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Author contributions
SJ contributed to preparation of manuscript and analysis of data; UP contributed to review of manuscript; RG contributed to editing of manuscript and references; MK contributed to preparation of manuscript and images. All authors have read and approved the manuscript.

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Availability of data and materials
Complete data of the patient can be retrieved from record keeping department of the institute.

Declarations

Ethics approval and consent to participate
Study approval was taken from the Institutes ethics committee and informed consent was taken from all participants.

Consent for publication
Written consent for publication was taken from the patients who participated in the study.

Competing interests
The authors declare that they have no competing interests.

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