Received:
10 January 2019

Revised:
20 February 2019

Accepted:
03 March 2019

<https://doi.org/10.1259/bjr.20190045>

Cite this article as:

Bhayana A, Ghasi RG. MRI evaluation of pelvis in Mayer-Rokitansky-Kuster-Hauser syndrome: interobserver agreement for surgically relevant structures. *Br J Radiol* 2019; **92**: 20190045.

FULL PAPER

MRI evaluation of pelvis in Mayer-Rokitansky-Kuster-Hauser syndrome: interobserver agreement for surgically relevant structures

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Objectives: Diagnostic role of MRI in Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is well documented. Recent studies have shown the relevance of MRI in detailing the pelvic anatomy, particularly small Mullerian remnants. The results are, however, not consistent and reproducibility of MRI features has not been confirmed. The aim of our study was to evaluate the detailed pelvic anatomy in patients with MRKH syndrome and to calculate the interobserver agreement of the MRI features.

Methods: The study was carried out on female patients, aged 15–30 years, with primary amenorrhea. Following clinical, gynaecological, endocrinological, ultrasound and chromosomal work-up and a provisional diagnosis of MRKH syndrome, MRI pelvis was performed on 25 patients using a 1.5 T scanner. MRI images were retrospectively reviewed by two radiologists for a detailed evaluation of Mullerian structures (uterine buds, fibrous

bands, midline triangular soft tissue), vagina, and ovaries. κ coefficient was calculated as a measure of interobserver agreement.

Results: Interobserver agreement was good to excellent (κ : 0.7788 ± 0.2168 to 1 ± 0) for uterine buds and their characteristics, vagina, ovaries and associated renal/vertebral anomalies; poor to fair for fibrous bands (κ : 0.2857 ± 0.3273 to 0.6032 ± 0.2149) and good for midline triangular soft tissue (κ : 0.7826 ± 0.1474).

Conclusion: MRI is capable of providing key pre-operative anatomical information in MRKH syndrome non-invasively. There is good to excellent interobserver agreement for MRI features of most of the surgical relevant structures.

Advances in knowledge: MRI features of salient preoperative anatomical structures are reproducible between observers.

INTRODUCTION

The paired Mullerian or paramesonephric ducts fuse between 6 and 11 weeks of gestation to form uterus, cervix and upper two-thirds of vagina. Mullerian duct anomalies (MDA) result from disruption in any of the three stages of uterine development, *i.e.* organogenesis, fusion or septal resorption.¹ Ovaries develop from primordial germ cells, while lower one-third of vagina develops from sinovaginal bulb. Owing to the different embryological origins, ovaries and lower third of vagina are not involved in MDA.²

The prevalence of MDA is reported to be 1–5%.² American Fertility Society (AFS), has classified MDA into seven basic types, from Type I to Type VII.³ Out of these seven types, Type I MDA also called as Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, which reportedly accounts for nearly 15% of total case load.² MRKH syndrome is characterized by complete/partial agenesis of uterus and upper two-thirds of vagina, with normal bilateral ovaries,

fallopian tubes and 46 XX karyotype.² Although, uterine absence is a rule, there may be presence of bilateral Mullerian remnants, *i.e.* the rudimentary uterine buds, seen as solid muscular tissues along the lateral pelvic walls. These Mullerian remnants may contain the utero-ovarian and round ligaments.^{4–7}

Clinical presentation is with primary amenorrhoea. The normal secondary sexual characteristics and a normal external phenotype is the major cause for delayed diagnosis. The clinical diagnosis of MRKH is exclusion based, with normal genetic and endocrine workup. Ultrasound is first-line imaging tool for MDA and is usually diagnostic of MRKH in the appropriate clinical setting. The management decisions are, however, not limited to a diagnosis. The ultimate goal of surgery has traditionally been restoration of sexual function using any of the vaginoplasty techniques.^{5,6} With advancements in reproductive surgeries and infertility treatment, more ambitious attempts at reproduction

can be made. There lies now a need for an exhaustive anatomical evaluation of the rudimentary uterine buds, especially cavitation and volumes, with additional information required pertaining to the location of ovaries. Laparoscopy with intraoperative ultrasound has been used to provide this information.^{2,4-8} MRI is the only non-invasive modality capable of dispensing these details.

In the past decade, there has been an increasing interest in the role of MRI in pre-operative assessment of MRKH syndrome.⁵⁻¹¹ Reproducibility of MRI features in intricate evaluation of anatomy including Mullerian remnants has still not been extensively researched. The purpose of this study was to assess the interobserver reproducibility of pelvic MRI anatomy in MRKH syndrome and substantiate its role as a non-invasive alternative to diagnostic laparoscopy.

METHODS AND MATERIALS

Subjects and ethics approval

Institutional review board approval was obtained. A study was carried out at an institutional level on consecutive female patients, in the age group of 15–30 years, who presented to Department of Gynaecology, with history of primary amenorrhoea, with or without pelvic pain, from November 2014 to September 2018. Among these, MRKH syndrome was suspected in patients with normal secondary sexual characteristics, normal external genitalia, normal hormonal levels (FSH and LH), 46 XX karyotype and lack of well-developed uterus on ultrasound. Patients were then referred for MRI pelvis. A detailed retrospective analysis of MRI images of 25 patients finally diagnosed as MRKH syndrome was undertaken for the study.

MRI procedure

Following clinical, gynaecological, endocrinological, chromosomal work-up, and ultrasound pelvis, these patients were referred for MRI Pelvis, for adequate and detailed evaluation of Mullerian structures, vagina, and ovaries. MRI was performed on 1.5 T scanner (PHILIPS ACHIEVA Netherlands), using a pelvic phased array coil. We did not use any bowel preparation, fasting or antiperistaltic agents. We did not use intravaginal gel for vaginal canal distension to maintain simplicity of the examination. We did not use fat saturation in any of the sequences because the Mullerian remnants can be hypointense on T_2 weighted images. MRI protocol is summarized in Table 1.

MRI image analysis

The MRI images were independently reviewed by two radiologists with 15 years and 5 years of experience in MRI respectively.

Radiologists were blinded to the ultrasound findings. The following MRI features were analysed.

(1) Uterine buds:

1. Presence or absence (If present : (a) unilateral or bilateral. (b) Volumes of the buds. (c) Signal intensity on T_2)
2. Location, *i.e.* relationship with respect to ovaries (cranial/caudal/at same plane, anterior/posterior/at same plane).
3. Presence of bud cavitation (layered differentiation).
4. Presence of intraluminal blood.
5. Presence of a triangular midline soft tissue structure. (If present: define T_2 signal intensity).
6. Presence or absence of fibrous band connecting two uterine buds. (If present: Thin/Thick and T_2 signal intensity).

(2) Vagina:

The presence of two embryologically separate parts of vagina, *i.e.* upper two thirds and lower one-thirds was evaluated on T_2 weighted sagittal and axial MR images. The vagina was identified by its hyperintense mucosa surrounded by hypointense fibromuscular wall and hyperintense perivaginal venous plexus.

(3) Ovaries:

1. Presence
2. Location: pelvic or extra pelvic. Ovaries were considered extrapelvic if they were located in abdomen, inguinal canal, anterior or anterolateral to iliac vessels/psoas muscles.
3. Associated mass or cyst

(4) Associated anomalies:

1. Renal anomalies
2. Lumbosacral vertebral anomalies

STATISTICAL ANALYSIS

Interobserver agreement was calculated for each MRI parameter. κ coefficient with standard error was calculated as a measure of inter observer agreement for each parameter. Additionally, Lin's concordance coefficient was calculated, for interobserver agreement between bilateral uterine bud volumes. The statistical analysis used in the study was performed on MS Excel, using SPSS v. 25 (IBM, Portsmouth, UK). The MRI imaging findings, for both observers, with κ values is summarized in Table 2.

Table 1. MRI protocol

Sequence	Plane	TR/TE (ms)	Flip angle (°)	FOV (mm) /RFOV (%)	Slice width /Interval(mm)	NSA
T1- FSE	Axial	780/12	90	300-320/80	4/0.4	3–4
T2-FSE	Axial, coronal, sagittal	4800-5000/120	90	300-320/80	4/0.4	3–4
T2- FSE	Coronal	4800/120	90	Large FOV to cover renal areas	5/1	3

FOV, field of view; FSE, fast spin echo; TE, echo time; TR, repetition time.

Table 2. MRI results by two observers with κ coefficient.

Right uterine bud		Observer 1	Observer 2	κ coefficient $\pm$ standard error
	Presence/absence ($N = 25$)	22/3	23/2	0.7788 ± 0.2168
	Craniocaudal position w.r.t ovary: ($N = 22$) Caudal to ovary/At same position/ Cranial	21/1/0	21/1/0	1 ± 0
	Anteroposterior position w.r.t. ovary: ($N = 22$) Anterior to ovary/Posterior/Same plane	20/2/0	20/2/0	1 ± 0
	Signal intensity: ($N = 22$) Hyperintense/Hypointense	21/1	21/1	1 ± 0
	Cavitation: ($N = 22$) Present/Absent	6/16	6/16	1 ± 0
	Intraluminal blood : ($N = 22$) Present/Absent	0/22	0/22	1 ± 0
	Uterine mass: ($N = 22$) Present/Absent	0/22	0/22	1 ± 0
Left uterine bud				
	Presence/absence ($N = 25$)	18/7	18/7	1 ± 0
	Craniocaudal position w.r.t ovary: ($N = 18$) Caudal to ovary/At same position/ Cranial	14/4/0	14/4/0	1 ± 0
	Anteroposterior position w.r.t. ovary: ($N = 18$) Anterior to ovary/Posterior/Same plane	14/4/0	14/4/0	1 ± 0
	Signal intensity: ($N = 18$) Hyperintense/Hypointense	16/2	16/2	1 ± 0
	Cavitation: ($N = 18$) Present/Absent	3/15	3/15	1 ± 0
	Intraluminal blood: ($N = 18$) Present/Absent	0/18	0/18	1 ± 0
	Uterine mass: ($N = 18$) Present/Absent	0/18	0/18	1 ± 0
Right fibrous band	Present/absent ($N = 25$)	24/1	20/5	0.2857 ± 0.3273
	Signal intensity: ($N = 20$) Hyperintense/Hypointense	4/16	4/16	1 ± 0
	Thick/Thin ($N = 20$)	6/14	6/14	1 ± 0
Left fibrous band	Present/absent ($N = 25$)	22/3	19/6	0.6032 ± 0.2149
	Signal intensity: ($N = 19$) Hyperintense/Hypointense	8/11	8/11	1 ± 0
	Thick/Thin ($N = 19$)	6/13	6/13	1 ± 0
Midline triangular soft tissue	Present/absent ($N = 25$)	20/5	18/7	0.7826 ± 0.1474
	Signal intensity: ($N = 18$) Hyperintense/Hypointense	13/5	13/5	1 ± 0
Ovary (Right)	Present/absent ($N = 25$)	25/0	25/0	1 ± 0
	Intrapelvic /Extrapelvic ($N = 25$)	19/6	19/6	1 ± 0
	Ovarian mass ($N = 25$) Present/absent	0/25	0/25	1 ± 0
Ovary (Left)	Present/absent ($N = 25$)	25/0	25/0	1 ± 0
	Intrapelvic/Extrapelvic ($N = 25$)	15/10	15/10	1 ± 0
	Ovarian mass ($N = 25$) Present/absent	0/25	0/25	1 ± 0

(Continued)

Table 2 (Continued)

Right uterine bud		Observer 1	Observer 2	κ coefficient $\pm$ standard error
Vagina	Upper third Present/absent	0/25	0/25	1 $\pm$ 0
	Lower third Present/absent	25/0	25/0	1 $\pm$ 0
Renal anomalies (N = 25)	Renal agenesis: Present/absent	3/22	3/22	1 $\pm$ 0
	Ectopic pelvic kidney Present/absent	1/24	1/24	1 $\pm$ 0
	Solitary ectopic pelvic kidney Present/absent	1/24	1/24	1 $\pm$ 0
	Horseshoe kidney Present/absent	0/25	0/25	1 $\pm$ 0
Vertebral anomalies (N = 25)	Lumbosacral transition vertebrae Present/absent	2/23	2/23	1 $\pm$ 0
	Sacral agenesis Present/absent	1/24	1/24	1 $\pm$ 0
	Scoliosis Present/absent	1/24	1/24	1 $\pm$ 0

RESULTS

Uterine buds/remnants

The right rudimentary uterine bud was observed to be present in 22/25 cases by Observer 1 and 23/25 cases by Observer 2. This suggested a good interobserver agreement (κ : 0.7788 $\pm$ 0.2168). The left rudimentary uterine bud, was reported to be present in 18/25 cases by both observers, giving an excellent interobserver agreement (κ : 1 $\pm$ 0). Bilateral uterine bud volumes were calculated using formula for prolate ellipsoid, *i.e.* Volume = 0.52 (Length X width X height). Lin's concordance coefficient was calculated for bilateral uterine bud volumes, for both observers, which was 0.6244 (confidence interval = 0.4411–0.7577), suggesting a good correlation. Excellent inter observer agreement (κ : 1 $\pm$ 0) was obtained for relative position of buds and ovaries with majority of buds being caudal and anterior to the ipsilateral ovary (Figure 1). Both observers were in complete agreement with regard to bud cavitation. (Figure 2).

Fibrous bands and midline triangular soft tissue structure

The presence of fibrous bands, extending from uterine buds and converging onto a midline triangular soft tissue structure were documented by both observers (Figure 1). The right fibrous band was observed to be present in 24/25 cases by Observer 1 and in 20/25 cases by Observer 2. This finding showed a poor interobserver agreement with a low κ value (0.2857 $\pm$ 0.3273). However, there was a fair interobserver agreement for left fibrous band (κ : 0.6032 $\pm$ 0.2149). Further, both observers documented the signal intensities and thickness of these bilaterally present fibrous bands, which showed an excellent inter observer agreement (κ : 1 $\pm$ 0). Presence of a retrovesical midline triangular soft tissue structure was noted by both observers, with good interobserver agreement (κ :

0.7826 $\pm$ 0.1474) (Figure 1). Also, the signal intensity of these observed midline structures revealed excellent agreement (κ : 1 $\pm$ 0).

Vagina

Our study showed that the lower one-third of vagina was seen in 100% of cases, with absent upper third in all 25 cases, by both the observers, using T2 sagittal and axial MRI images (κ : 1 $\pm$ 0) (Figure 1).

Ovaries

Both the observers reported normal morphology bilateral ovaries to be present in all the 25 cases (Figure 1). 6 right ovaries and 10 left ovaries were reported to be extrapelvic (κ : 1 $\pm$ 0), (Figure 3). One case with inguinal left ovary was reported by both the observers (Figure 4). No ovarian mass or cyst was reported by either of the observers (κ : 1 $\pm$ 0).

Associated anomalies

There was excellent agreement between the two observers for associated renal anomalies (κ : 1 $\pm$ 0). Both the observers reported three cases of left renal agenesis (Figure 2), one case of a solitary ectopic malrotated pelvic right kidney and one case of ectopic malrotated left pelvic kidney. Also, there was excellent agreement for associated vertebral anomalies (κ : 1 $\pm$ 0). Both the observers reported two cases of lumbosacral transition vertebra (Castellvi's Type II A in one and Type II B in second), and one case of levoscoliosis of lumbosacral spine with partial sacral agenesis. Combined renal and vertebral anomalies were also present (Figures 2 and 5). Therefore, there were 6 cases of MRKH type II and 19 cases of MRKH Type I in our study.

Figure 1 (a–d). Type I MRKH syndrome in an 18-year-old female. (a) T₂W sagittal MRI image shows absence of a normal morphology uterus, cervix between bladder and rectum. A triangular midline soft tissue structure is seen above the dome of urinary bladder (thick yellow arrow). Lower third of vagina is seen (thin yellow arrow). (b) T₂W coronal MRI image shows normal bilateral ovaries (yellow arrows). (c) T₂W coronal MRI image shows bilateral hyperintense rudimentary uterine remnants, abutting and lying in caudal relationship to both ovaries (dotted yellow arrow). (d) T₂W axial MRI image shows a thick hyperintense band (red arrow), connecting the bilateral uterine remnants (yellow asterisks). T₂W, T₂ weighted.

DISCUSSION

The initial classification of Mullerian duct anomalies was proposed by Buttram and Gibbons, with subsequent modification by American Fertility Society.^{3,12} MRKH syndrome is a Type I MDA, with an incidence of about 1:4000–1:5000 female births.² There are two types of MRKH syndrome; i.e., Type I/Type A/isolated/Typical MRKH syndrome is characterized by isolated utero-vaginal abnormality, while Type II/Type B/Atypical MRKH syndrome is characterized by presence of associated non-gynaecological anomalies namely; renal, skeletal, ovarian, middle ear, cardiac and tubal anomalies. Type B is reported to be more common.¹³ However, we observed 6 cases of MRKH Type II and 19 cases of MRKH Type I in our study. The atypical MRKH syndrome, which comprises of anomalies of all three systems, i.e. Mullerian, renal and cervico-thoracic somite dysplasia is called MURCS syndrome.^{4,13,14}

Ultrasound is established as the initial imaging modality for Mullerian anomalies including MRKH. However, transvaginal three-dimensional ultrasound is not feasible in paediatric or sexually inactive young patients. A partially developed small rudimentary uterine horn may be misinterpreted as a small tubular prepubertal uterus. Visualization of extrapelvic ovaries can be challenging.^{2,4–7} Occasionally, the retrovesical

Figure 2 (a–f). Type II MRKH syndrome in a 17-year-old female. (a) T₂W sagittal MRI image shows absence of uterus, cervix and vagina between bladder and rectum (yellow arrow). (b) T₂W axial MRI image shows a cavitating rudimentary right uterine remnant, with three layered differentiation. Left uterine remnant is absent. (c) T₂W sagittal MRI image shows normal right ovary (yellow solid arrow), with right rudimentary uterine remnant, abutting and lying in caudal relationship to ovary (dotted yellow arrow). (d) T₂W axial MRI image shows normal right ovary. (e) T₂W coronal MRI image shows extra pelvic, ectopically located left ovary, lateral to the psoas muscle, above the pelvic brim (red arrow). (f) T₂W coronal MRI image shows normal right kidney in right renal fossa (red arrow), with absent left kidney in left renal fossa (yellow asterisk). T₂W, T₂ weighted.

quadrangular vestigial lamina being misinterpreted as hypoplastic uterus.¹³

MRI is reported to be almost 100% sensitive and specific for diagnosis of Mullerian duct anomalies.⁵ Earlier, the management options were limited to vaginoplasty with the aim of restoring sexual function. The development of assisted reproductive

Figure 3 (a–e). Type I MRKH syndrome in a 20-year-old female. (a) T₂W sagittal MRI image shows absence of a normal morphology uterus, cervix between bladder and rectum. A small triangular midline soft tissue structure is seen above the dome of urinary bladder (yellow arrow). (b) T₂W sagittal MRI image shows normal lower third of vagina (yellow arrow). (c) T₂W axial MRI image shows normal morphology of bilateral ovaries, with extra pelvic location, seen anterolateral to bilateral iliac vessels (dotted yellow arrows). (d) T₂W sagittal MRI image shows hyperintense right rudimentary uterine remnant, lying in caudal relationship to ovary (red arrow). (e) T₂W sagittal MRI image shows hyperintense left rudimentary uterine remnant, lying in caudal relationship to ovary (red arrow). MRKH, Mayer-Rokitansky-Kuster-Hauser; T₂W, T₂ weighted

Figure 4 (a,b).Type I MRKH syndrome in a 24-year-old female. (a) T_2W sagittal MRI image shows left ovary in inguinal canal (dotted red arrow). (b) T_2W axial MRI image shows hyperintense rudimentary right uterine remnant in right inguinal canal, with inguinal left ovary and rudimentary left uterine remnant abutting the left ovary (red arrows).

technologies now allows for possible creation of a small functioning uterus.⁵ Also, oocyte retrieval methods require accurate localization of extrapelvic ovaries. Laparoscopy with intraoperative ultrasound was the only method to provide detailed information about Mullerian bud size, location, cavitation and position of ovaries. It has been proposed that MRI can provide such information non-invasively.⁵ In the past decade, a number of studies have studied rudimentary Mullerian remnants in MRKH patients.^{5–11}

Presence of rudimentary uterine buds was documented by both the readers with good interobserver agreement. Uterine buds were mostly bilateral. Signal intensity was usually hyperintense on T_2 weighted images. There was good interobserver correlation regarding the uterine bud volumes. Presence of cavitation within rudimentary uteri was shown with excellent inter

Figure 5 (a–d).Type II MRKH syndrome in a 22-year-old female. (a) T_2W axial MRI image shows normal morphology of bilateral ovaries, with extra pelvic location, seen anterolateral to bilateral iliac vessels and psoas muscles (green arrows). (b) T_2W coronal MRI image shows bilateral hyperintense rudimentary uterine remnants, abutting and lying in caudal relationship to both ovaries (yellow arrows). (c) T_2W coronal MRI image shows absent bilateral kidneys in renal fossae (yellow asterisks), with malrotated solitary ectopic pelvic kidney (red arrow). (d) T_2W coronal MRI image shows pseudarthrosis of bilateral L5 transverse processes with S1 (Lumbosacral Transition vertebrae Type II B; dotted yellow arrows). MRKH, Mayer-Rokitansky-Kuster-Hauser; T_2W , T_2 weighted.

observer agreement. Although not a constant relationship; buds were usually antero inferior to ipsilateral ovaries. This knowledge may assist in intraoperative localization.⁸ These findings are important because cavitated buds can potentially cause cyclical pain or endometriosis and may need removal. Also, presence of a reasonably sized cavitated uterine bud may be surgically reconstructed to create a functional uterus. No other diagnostic modality has been found accurate enough to provide this information non-invasively.

The Mullerian remnants described in MRKH include a midline triangular soft tissue structure, lying above the bladder dome, and bilateral fibrous bands, extending from rudimentary uterine buds and converging onto this triangular structure in the midline. There was good interobserver agreement in detection of the midline retrovesical remnant and excellent agreement regarding its signal intensity. The interobserver agreement in the detection of fibrous bands was poor to fair. This may be due to their fine structure and difficult visualization by a less experienced observer. The importance of identifying these structures pre-operatively is guiding a better laparoscopic resection of the uterine buds. The surgical resection involves medial traction of the laterally situated uterine buds, to prevent injury to the pelvic side-wall structures and ureter.^{15,16} The resection plane should ideally proceed along these fibrous bands across the midline.¹⁶

In reference to vagina, our study showed that the lower one-third of vagina was seen in all our cases. We did not attempt to measure the length of the vaginal canal because we did not use any intra-vaginal gel or contrast during the study.

All our 25 cases (100%) had normally developed bilateral ovaries, which is expected due to the different embryonic origins of the ovaries and uterus. However, 24% (6/25) of right ovaries and 40% (10/25) of the left ovaries were ectopic/extra pelvic in location in our study, which is in concordance with the previously published studies. The exact location of the ovaries is extremely helpful for *in-vitro* fertilization or surrogacy.⁸ Transvaginal ovum retrieval becomes difficult in extrapelvic ovaries and abdominal route needs to be opted.⁸

The ovaries in MRKH syndrome are commonly extrapelvic, located at or above the level of pelvic brims, far anterior or anterolateral to the psoas muscles, anterolateral to the iliac vessels, or in iliac fossae or in abdomen.⁸ The embryological explanation proposed is female gubernaculum dysfunction, with elongated utero-ovarian ligaments.¹⁷ The T_2 coronal larger field of view MRI image, plays an important role in detecting extra pelvic ovaries (Figure 2). Rarely, the ovaries can be found in bilateral inguinal canals, as shown in one of our patients (Figure 4).^{18,19} Reporting this fact remains important, to prevent torsion, incarceration and ischaemia of these ectopic inguinal ovaries.¹⁰ Excessive mobility of ovaries and weak suspensory ligaments have been hypothesized as possible causes for inguinal location of ovaries.^{19–21}

Renal anomalies are known to be one of the commonly associated non-gynaecologic anomalies in Type II MRKH syndrome.^{2,4–9}

Table 3. Comparative analysis with previous studies

Ref. no.	Sample size, surgical correlation	Scanner, protocol, observers	Results	Comparison with results of present study
Pompii et al., 2009 (5)	56, Yes	1.0 T Multiplanar T_2W , T_1W two experienced reviewers, Consensus reporting	Uterine buds: Present (78%), bilateral (61%). 100% accurate for bud cavitation. Fibrous remnants: not evaluated Ovaries: 14.5% ectopic Vagina: Lower 1/3 rd vagina present in 17/56 patients Renal anomalies: 11% patients MRI sensitivity- 81.42 %, good agreement with Laparoscopy (κ - 0.55).	Similar MRI results. Larger sample Surgical correlation+ Precise location of uterine buds, volume, not assessed. No information about fibrous remnants.
Yoo RE et al., 2013 (6)	15, No	1.5 T & 3.0 T Multiplanar T_2W , T_1W imaging two experienced gynaecological radiologists, Consensus reporting	Uterine buds: Present bilaterally in 100% patients, cavitation in 13%, Fibrous remnants: Bands present in 100%, midline remnant in 86% patients Ovaries: 42% ectopic Vagina: lower 1/3rd present in 93% patients Renal/vertebral anomalies: 40% patients	Higher incidence of Mullerian remnants. Smaller sample Precise location of uterine buds, volume, not assessed
Kara et al., 2013 (7)	16, No	1.5 T Multiplanar T_2W , T_1W No 3D scanning two reviewers with specialized experience, Consensus reporting	Uterine buds: Present (68%) Precise location cavitation, volume not assessed Fibrous remnants: not evaluated Ovaries: 12.5% ectopic Vagina: Blind ending in 100% Renal anomalies: 25% Vertebral anomalies: 12.5%	Smaller sample. Precise location of uterine buds, cavitation, volume, was not calculated. No information about fibrous remnants.
Craggs et al., 2013 (8)	66, No	1.5 T Multiplanar T_2W , T_1W images One observer with specialized experience	Uterine buds: Present (92%), bilateral (82%), cavitation (21%), location caudal to ovaries(100%), mean bud volume 6.4 ml Fibrous remnants: Bands(6%) Ovaries: ectopic 42% Vagina: Canal present in 66%, mean length 2 cm, dimple in 33% Renal anomalies: 20%	Larger sample size The location of buds constantly caudal to ovary. Fibrous remnants uncommon.
Preibsch H et al., 2014 (9)	115, Yes	Retrospective review of routine pre-operative MRI, no interobserver rating	Uterine buds: Present (90%), bilateral (75%), cavitation (33%) Fibrous remnants: not evaluated Ovaries: MRI accurate in 93% patients Renal anomalies: 27%	Much larger sample size with surgical correlation. No information about fibrous remnants, bud volumes or precise location.
Boruah DK et al., 2017 (10)	11, No	two reviewers, consensus reporting	Uterine buds: Presence in 81%, all bilateral, Location caudal to ovary(100%), mean bud volume 2.26 ml (right) & 1.2 ml (left), cavitation (5.6%) Fibrous remnants: Bands (77.8%), midline remnant (44.4%) Ovaries: Ectopic (9%) Vagina: Mean length 2.6 cm Renal anomalies: 9% Vertebral: 27%	Similar results, smaller sample. No unilateral buds detected.
Wang Y et al., 2017 (11)	92, No	3 T Variable MRI protocol two experienced gynaecological reviewers, consensus reporting	Uterine buds: Present (78%), bilateral (68%), cavitation in 50% Fibrous remnants: 48% of patients with bilateral buds Ovaries: Ectopic 38% Vagina: lower 1/3rd (17%), mean length 2.4 cm Associated anomalies: 50%	Larger sample size Similar results, although mean bud volumes are much larger. No information about fibrous remnants

Presence of ectopic pelvic kidney may lead to increased ureteral injuries while performing laparoscopic vaginoplasty or laparoscopic resection of obstructed uterine remnants.^{4,5,15} We observed three cases of unilateral renal agenesis (3/25; 12%),

one solitary ectopic pelvic kidney (1/25; 4%) and one unilateral ectopic pelvic kidney (1/25; 4%). Among the various vertebral anomalies, we observed one case of scoliosis with partial sacral

agenesis (1/25; 4%) and two cases of lumbosacral transition vertebrae (2/25; 8%).

A comparative analysis of our study with other major similar studies has been summarized in Table 3. Our results and those of most other studies are in agreement that uterine buds are quite commonly found in MRKH syndrome and that bilateral buds are more frequent. The incidence of ectopic ovaries and associated anomalies is also remarkably similar in all the studies. The presence of fibrous remnants is much more variably reported, wherein our results are nearly similar to Yoo RE et al, and at variance with others (Hall-Cragg MA et al, Baruah DK et al, Wang Y et al). Most others have not specifically analysed MRI for fibrous remnants. All the studies largely conclude that MRI does have the ability to non-invasively elaborate upon the detailed pelvic anatomy in MRKH patients and can be used for preoperative assessment. Our study adds objectivity to available literature, because most studies are based on consensus reporting, while we have calculated interobserver agreement for each of the MRI features, suggesting an overall good reproducibility.

The strength of our study was the homogeneity of data. All patients were imaged on same 1.5 T MRI machine and study population was quite homogeneous, with similar age range.^{6,8} We did not use any specialized MRI sequences or three-dimensional

imaging, because we aimed to assess the reproducibility of MRI findings in a routine MRI protocol. Additionally, the images were reported and reviewed by two radiologists, with statistical calculation of the interobserver agreement for all the imaging parameters. Unlike many previous studies, our observers did not have any specialized experience in gynaecological imaging, as our objective was to assess if MRI can be routinely used for anatomical evaluation in MRKH syndrome. Our study revealed good to excellent interobserver agreement for characteristics of uterine buds, vagina, ovaries and associated renal or vertebral anomalies.

However, there were some limitations of our study. Firstly, we did not correlate MRI results with diagnostic laparoscopy or surgery because it was a cross-sectional study and we did not follow up the patients. Secondly, our sample size was small.

CONCLUSION

Rudimentary uterine buds and fibrous remnants are frequently present in MRKH syndrome and there is a high incidence of extrapelvic ovaries. Renal and lumbosacral vertebral anomalies are common. The good to excellent inter observer agreement for most of the structures the reproducibility of MRI features. MRI can be used as a preoperative assessment tool as a non-invasive alternative to diagnostic laparoscopy in MRKH syndrome.

REFERENCES

- Gould SW, Epelman M. Magnetic resonance imaging of developmental anomalies of the uterus and the vagina in pediatric patients. *Seminars in Ultrasound, CT and MRI* 2015; **36**: 332–47. doi: <https://doi.org/10.1053/j.sult.2015.05.002>
- Behr SC, Courtier JL, Qayyum A. Imaging of Müllerian duct anomalies. *Radiographics* 2012; **32**: E233–E250. doi: <https://doi.org/10.1148/rg.326125515>
- The American Fertility Society The American fertility Society classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, Müllerian anomalies and intrauterine adhesions. *Fertil Steril* 1988; **49**: 944–55. doi: [https://doi.org/10.1016/S0015-0282\(16\)59942-7](https://doi.org/10.1016/S0015-0282(16)59942-7)
- Rousset P, Raudrant D, Peyron N, Buy J-N, Valette P-J, Hoeffel C. Ultrasonography and MRI features of the Mayer-Rokitansky-Küster-Hauser syndrome. *Clin Radiol* 2013; **68**: 945–52. doi: <https://doi.org/10.1016/j.crad.2013.04.005>
- Pompili G, Munari A, Franceschelli G, Flor N, Meroni R, Frontino G, et al. Magnetic resonance imaging in the preoperative assessment of Mayer-Rokitansky-Küster-Hauser syndrome. *Radiol Med* 2009; **114**: 811–26. doi: <https://doi.org/10.1007/s11547-009-0407-5>
- Yoo R-E, Cho JY, Kim SY, Kim SH. Magnetic resonance evaluation of Müllerian remnants in Mayer-Rokitansky-Küster-Hauser syndrome. *Korean J Radiol* 2013; **14**: 233–9. doi: <https://doi.org/10.3348/kjr.2013.14.2.233>
- Kara T, Acu B, Beyhan M, Gökçe E. MRI in the diagnosis of Mayer-Rokitansky-Küster-Hauser syndrome. *Diagn Interv Radiol* 2013; **19**: 227–32.
- Hall-Craggs MA, Williams CE, Pattison SH, Kirkham AP, Creighton SM. Mayer-Rokitansky-Küster-Hauser syndrome: diagnosis with MR imaging. *Radiology* 2013; **269**: 787–92. doi: <https://doi.org/10.1148/radiol.13130211>
- Preibsch H, Rall K, Wietek BM, Brucker SY, Staebler A, Claussen CD, et al. Clinical value of magnetic resonance imaging in patients with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome: diagnosis of associated malformations, uterine rudiments and intrauterine endometrium. *Eur Radiol* 2014; **24**: 1621–7. doi: <https://doi.org/10.1007/s00330-014-3156-3>
- Boruah DK, Sanyal S, Gogoi BB, Mahanta K, Prakash A, Augustine A, et al. Spectrum of MRI appearance of Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome in primary amenorrhea patients. *J Clin Diagn Res* 2017; **11**: TC30–TC35. doi: <https://doi.org/10.7860/JCDR/2017/29016.10317>
- Wang Y, Lu J, Zhu L, Sun Z, Jiang B, Feng F, et al. Evaluation of Mayer-Rokitansky-Küster-Hauser syndrome with magnetic resonance imaging: three patterns of uterine remnants and related anatomical features and clinical settings. *Eur Radiol* 2017; **27**: 5215–24. doi: <https://doi.org/10.1007/s00330-017-4919-4>
- Buttram VC Jr, Gibbons WE. Müllerian anomalies: a proposed classification. *Fertil Steril* 1979; **32**: 40–6 (An analysis of 144 cases.).
- Fiaschetti V, Taglieri A, Gisone V, Coco I, Simonetti G. Mayer-Rokitansky-Küster-Hauser syndrome diagnosed by magnetic resonance imaging. Role of imaging to identify and evaluate the uncommon variation in development of the female genital tract. *J Radiol Case Rep* 2012; **6**: 17–24. doi: <https://doi.org/10.3941/jrcr.v6i4.992>
- Duncan PA, Shapiro LR, Stangel JJ, Klein RM, Addonizio JC. The MURCS association: Müllerian duct aplasia, renal aplasia, and cervicothoracic somite dysplasia. *J Pediatr* 1979; **95**: 399–402. doi: [https://doi.org/10.1016/S0022-3476\(79\)80514-4](https://doi.org/10.1016/S0022-3476(79)80514-4)

15. Strübbe EH, Willemsen WN, Lemmens JA, Thijn CJ, Rolland R. Mayer-Rokitansky-Küster-Hauser syndrome: distinction between two forms based on excretory urographic, sonographic, and laparoscopic findings. *AJR Am J Roentgenol* 1993; **160**: 331–4. doi: <https://doi.org/10.2214/ajr.160.2.8424345>
16. Hilger WS, Magrina JF. Removal of pelvic leiomyomata and endometriosis five years after supracervical hysterectomy. *Obstet Gynecol* 2006; **108**(3 Pt 2): 772–4. doi: <https://doi.org/10.1097/01.AOG.0000209187.90019.d3>
17. Acién P, Sánchez del Campo F, Mayol M-J, Acién M, Pedro A, Francisco SC, Maria-Jose M, Maribel A. The female gubernaculum: role in the embryology and development of the genital tract and in the possible genesis of malformations. *Eur J Obstet Gynecol Reprod Biol* 2011; **159**: 426–32. doi: <https://doi.org/10.1016/j.ejogrb.2011.07.040>
18. Al Omari W, Hashimi H, Al Bassam MK. Inguinal uterus, fallopian tube, and ovary associated with adult Mayer-Rokitansky-Küster-Hauser syndrome. *FertilSteril* 2011; **95**: 1119. e1–1119.e4.
19. Okada T, Sasaki S, Honda S, Miyagi H, Minato M, Todo S. Irreducible indirect inguinal hernia containing uterus, ovaries, and fallopian tubes. *Hernia* 2012; **16**: 471–3. doi: <https://doi.org/10.1007/s10029-010-0764-y>
20. Thomson GR. Complete congenital absence of the vagina associated with bilateral herniae of uterus, tubes, and ovaries. *Br J Surg* 1948; **36**: 99–100. doi: <https://doi.org/10.1002/bjs.18003614124>
21. Fowler CL. Sliding indirect hernia containing both ovaries. *J Pediatr Surg* 2005; **40**: e13–14. doi: <https://doi.org/10.1016/j.jpedsurg.2005.05.066>