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# Evaluation of the abdominopelvic region using MRI in patients with primary amenorrhea

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## Abstract

**Background:** This study aimed to evaluate the imaging findings of patients who underwent an abdominal and pelvic magnetic resonance imaging (MRI) due to primary amenorrhea.

**Methods:** The pelvic and abdominal images of 34 female patients (mean age 15.61 years, range 14–19 years) were retrospectively analyzed by a single radiologist blinded to the clinical and laboratory data of the patients (other than primary amenorrhea) to evaluate the etiology of primary amenorrhea. The anatomy and anomalies of the internal genital organs and other accompanying abdominopelvic anomalies were investigated.

**Results:** Gonadal dysgenesis was present in 14 patients (41.17%) and Müllerian duct anomalies (MDAs) were present in 20 (58.83%) (Mayer-Rokitansky-Kuster-Hauser [MRKH] syndrome in 13 [65%], distal vaginal obstruction [DVO] findings in five [25%], and obstructed hemivagina and ipsilateral renal anomaly [OHVIRA] syndrome in two [10%]). Seven patients with MRKH (53.84%) were of type 1 and six (46.15%) were of type 2. A total of eight additional anomalies (vertebral and renal) were detected, six in MRKH and two in OHVIRA syndrome cases. Endometrioma and hematosalpinx were observed in one of the five patients with DVO (5%).

**Conclusions:** Primary amenorrhea is a common symptom that affects both the physical and psychosocial status of individuals. Determination of the underlying etiology is the first step in planning treatment. The evaluation of internal genital organ anomalies involved in the etiology is important for sexual function and fertility. MRI is a

non-invasive imaging modality that should be preferred in these cases as it provides detailed data about the anatomy and anomalies of internal genital organs due to its high soft tissue contrast resolution.

**Keywords:** gonadal dysgenesis; magnetic resonance imaging; Mullerian duct anomaly; primary amenorrhea.

## Introduction

Primary amenorrhea, seen in 2–5% of adolescent girls, refers to the absence of menstruation by 16 years of age if there is normal development of secondary sexual characters, or by 14 years of age in the absence of normal growth [1–4]. Primary amenorrhea is a symptom, and various pathologies may be involved in its etiology [1, 3, 4]. For normal menstruation to occur, both a properly functioning hypothalamo-hypophyseal axis and normal female genital anatomy are necessary. A problem with either of these components leads to the symptoms of amenorrhea [1]. It is possible that the factors involved in this condition can be reversed by simple medical or surgical interventions, but sometimes more complex situations may arise, making menstruation completely impossible. Determining the cause is very important in order to select the appropriate treatment. Due to the variety of etiology, different methods are used to reach a diagnosis of primary amenorrhea [1, 2, 4]. Hormone and karyotype analyses are used to determine endocrine and cytogenetic anomalies, respectively [1, 2, 4]. Radiological imaging methods are utilized to identify anatomical disorders.

The imaging method often used in the visualization of the female genital organ anatomy and anomalies is ultrasonography (US) [2, 5, 6]. The advantages of US include easy availability, low cost and the absence of ionizing radiation; however, gas artifacts may cause limitations in the field of view (FOV) [5, 6]. In this case, magnetic resonance imaging (MRI) is the preferred reference method for advanced imaging with its high soft tissue resolution, non-requirement for ionizing radiation, multiplanar imaging capability and non-invasive nature in the detailed evaluation of the anatomy [5, 7, 8].

In this study, we aimed to evaluate the anatomy and anomalies of the internal genital organs and any other

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associated systemic finding of patients who underwent an abdominal MRI due to primary amenorrhea.

## Materials and methods

Prior to the study, the approval of the Ethics Committee of the Eskişehir Osmangazi University Medical Faculty was obtained.

### Patients

All patients were referred to our clinic for an abdominal and pelvic MRI to investigate the etiology of primary amenorrhea. The age of the patients ranged from 14 to 19 years (mean 15.61 years). None of the patients had a history of pelvic surgery.

The patients were evaluated using pelvic US before the MRI examination. All the patients had been referred to our clinic for further examination. The primary complaint was primary amenorrhea in 29 patients and acute pelvic pain accompanied by primary amenorrhea in five patients.

Twenty-nine patients were referred to MRI because the normal genital organs could not be visualized using US (uterus in four patients, ovaries in three patients and both in 22 patients). Five patients were scheduled for an MRI due to the pre-diagnosis of cystic mass-vaginal obstruction for further evaluation of the cystic mass lesion accompanied by acute pelvic pain and primary amenorrhea.

### MRI examination

The MRI images of 34 female patients who had been referred to our MRI unit with the diagnosis of primary amenorrhea between January 2015 and December 2018 were retrospectively evaluated from the hospital image archive system. Image quality was adequate

in 32 patients and low in two patients due to motion artifacts. As all images were of sufficient diagnostic quality, they were all included in analysis. In addition to a pelvic MRI, all patients underwent an upper abdominal MRI to identify possible accompanying anomalies.

The MRI examination of 27 patients was undertaken using a 1.5 Tesla MRI (Siemens, Magnetom Avanto, Germany) device with six-channel array coils, while for seven patients, a 3 Tesla MRI (General Electric, Milwaukee, WI, USA) device with 48-channel array coils was used. For both machines, the imaging protocol for the pelvic MRI comprised T1-weighted, T2-weighted, and T1- and T2-weighted fat suppressed sequences in the axial plane and T2-weighted sequences in the coronal and sagittal planes. The parameters of the MRI examination are summarized in Table 1.

### Analysis of images

The MRI images were evaluated by a single radiologist with experience in abdominal radiology using a dedicated workstation (GE, Advantage Workstation 4.3, Boston, MA, USA). The radiologist performed this evaluation blinded to the other clinical findings of patients (apart from primary amenorrhea) and the results of karyotype and biochemical analyses, as well as the operative results of the patients who underwent surgery.

The pelvic examination included the evaluation of the presence and morphology of the uterus, ovaries and vagina. The uterus was examined in terms of normal growth and developmental abnormalities, such as rudimentary uterus, agenesis and hypoplasia. The ovaries were evaluated in terms of normal development, location anomalies, agenesis, presence of ovotestis and hypoplasia. In cases where the ovaries could not be bilaterally visualized, the presence of inguinal or abdominal undescended testis was investigated considering the possibility of androgen insensitivity syndrome. Patients without normal ovarian and testicular tissues were classified as gonadal dysgenesis. Concomitant uterine and vaginal anomalies were recorded. Patients with ovarian tissue showing normal, hypoplastic, unilateral agenesis or location anomalies were examined for

**Table 1:** Imaging parameters of MRI.

Sequence	T1	T2	T1 (fat saturation)	T2 (fat saturation)
1.5 Tesla MRI				
TR/TE	507/69.8	7500/94	754/9.8	4940/96
Matrix size	256×117	256×127	256×117	256×127
NEX	1	1.5	1	1.5
FOV, cm	43	43	43	43
Slice thickness, mm	4	4	4	4
Intersection gap, mm	1	1	1	1
3 Tesla MR				
TR/TE	807/8.9	6959/93	574/7.9	6605/93
Matrix size	320×256	320×320	288×244	320×320
NEX	1	1.5	1	1.5
FOV, cm	40	40	42	42
Slice thickness, mm	4	4	4	4
Intersection gap, mm	1	1	1	1

FOV, field of view; MRI, magnetic resonance imaging; NEX, number of excitations; TE, echo time; TR, repetition time.

Müllerian duct anomalies (MDAs). These patients were subdivided according to the accompanying uterine and vaginal anomalies.

The pelvic images were investigated in terms of endometrioma, hemato-hydrosalpinx and other space-occupying adnexal masses. Both the upper abdomen and pelvic images were examined to identify urinary and vertebral anomalies, which may accompany the anomalies of the genital system. After the MRI examinations were completed, the coexistence of MRI and other findings, including the results of karyotype analysis, surgical results and important clinical data (syndromic cases, etc.) was analyzed in those cases for whom these data were accessible.

## Statistical analysis

SPSS software v. 22.0 (IBM, Chicago, IL, USA) was used for statistical analysis. Descriptive statistics on continuous data were obtained as mean, standard deviation, median, and minimum and maximum values, and for discrete data, the percentage values were used.

## Results

Gonadal dysgenesis was found in 14 of 34 patients (41.17%), and MDAs were found in 20 (58.83%) (Mayer-Rokitansky-Kuster-Hauser [MRKH] syndrome in 13 [65%], distal vaginal obstruction [DVO] findings in five [25%], and obstructed hemivagina and ipsilateral renal anomaly [OHVIRA] syndrome in two [10%]) (Table 2). None of the patients had inguinal or abdominal undescended testes.

The uterus was normal in three of the patients with gonadal dysgenesis. Four patients had rudimentary or hypoplastic uterus and seven patients did not have a

uterus. The operative results of the four patients with gonadal dysgenesis were available, all showing the presence of streak gonads with no findings indicating malignancy. In these patients, the streak gonads had not been visualized by MRI. Karyotype analysis had been performed in 10 patients (71.42%) with gonadal dysgenesis. The karyotype was determined as 46, XX in seven patients, 46, XY in two patients and 45, X in one patient. Gonadal dysgenesis coexisted with syndromes in six patients (Swyer syndrome in two patients, and Prader-Willi, Perrault, Cornelia de Lange and Turner syndromes in one patient each). The findings of patients with gonadal dysgenesis are summarized in Table 3.

Seven of the patients with MRKH (53.84%) were of type 1 and six (46.15%) were type 2. Furthermore, of the MRKH cases, three (23.07%) had a rudimentary uterus and 10 (76.92%) had uterine agenesis. The ovaries had normal appearance in seven patients (Figure 1). In one case, unilateral normal ovary was observed with a missing ovary on the other side. Two patients had slightly decreased ovarian

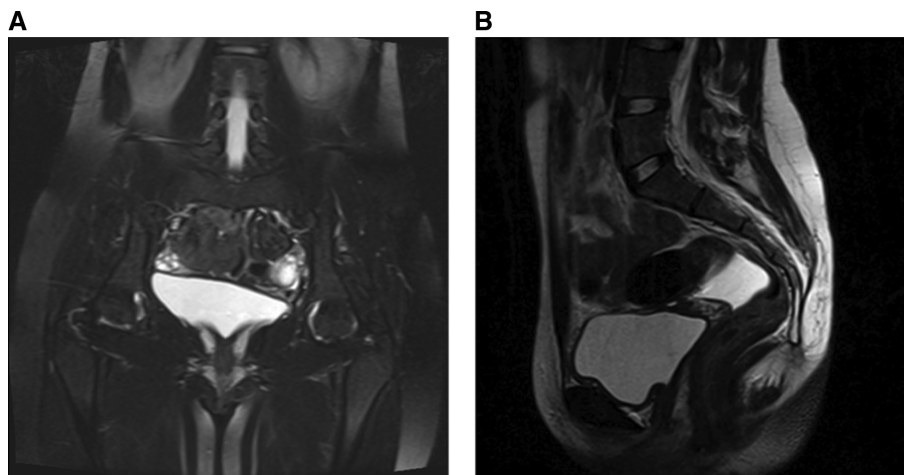
**Table 2:** Causes of primary amenorrhea detected by MRI.

Cause	Number of patients, n	Number of patients, %
Gonadal dysgenesis	14	41.17
Müllerian duct anomalies	20	58.83
Müllerian agenesis	13	65
Distal vaginal obstruction	5	25
OHVIRA syndrome <sup>a</sup>	2	10
Total	34	100

<sup>a</sup>OHVIRA syndrome, obstructed hemivagina and ipsilateral renal anomaly syndrome.

**Table 3:** Features of patients with gonadal dysgenesis.

Patient number	Uterus	Karyotype	Syndrome-clinical diagnosis	Result of surgery
1	Agenesis	46, XX		
2	Agenesis	46, XY	Swyer syndrome	
3	Agenesis	46, XY	Swyer syndrome	
4	Rudimentary		Prader-Willi syndrome	
5	Normal	46, XX	Hypergonadotropic hypogonadism	
6	Agenesis	46, XX	Perrault syndrome	Streak gonad
7	Rudimentary	46, XX		
8	Normal			Streak gonad
9	Agenesis	46, XX	Cornelia de Lange syndrome	Streak gonad
10	Rudimentary			
11	Normal	46, XX	Premature ovarian failure	Streak gonad
12	Agenesis			
13	Rudimentary	45, X	Turner syndrome	
14	Agenesis			



**Figure 1:** MRI showing (A) the normal right and left ovaries and (B) absence of uterus. The normal sized right and left ovaries are just above the bladder (A), no uterus is identified in normal position (B).

**Table 4:** Imaging findings of patients with MRKH syndrome.

Patient number	Uterus	Right ovary	Left ovary	Concomitant anomaly
1	Rudimentary	Hypoplasia	Hypoplasia	No
2	Agenesis	Hypoplasia	Hypoplasia	No
3	Rudimentary	Normal	Normal	No
4	Agenesis	Abnormal localization	Abnormal localization	Right renal ectopy-left renal agenesis
5	Agenesis	Hypoplasia	Normal	Left renal ectopy
6	Agenesis	Abnormal localization	Abnormal localization	Right renal agenesis
7	Agenesis	Hypoplasia	Hypoplasia	Right renal ectopy
8	Agenesis	Normal	Normal	No
9	Agenesis	Normal	Normal	Vertebral anomaly
10	Rudimentary	Normal	Normal	No
11	Agenesis	Normal	Normal	No
12	Agenesis	Normal	Normal	No
13	Agenesis	Normal	Normal	Right renal agenesis

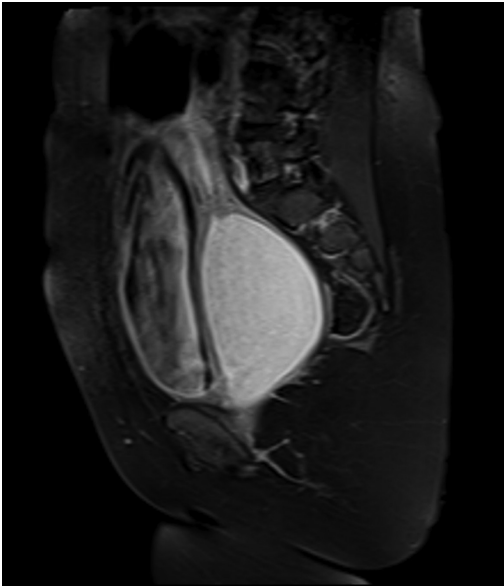
MRKH, Mayer-Rokitansky-Kuster-Hauser.

volume and the ovaries were in a higher position. In three patients, both ovaries were hypoplastic. There were seven additional findings among six patients with type 2 MRKH: ectopic kidney in three, renal agenesis in three and lumbosacral transitional vertebra in one. One patient presented with both renal agenesis and renal ectopia. Table 4 shows the findings of the patients with MRKH.

Five cases in which the proximal vagina and cervical cavity were distended by hemorrhagic content were evaluated in favor of DVO. Among these cases, hematometocolpos was detected in three patients (60%) and hematocolpos was detected in two (40%). In one patient (20%), atresia was observed in the middle and distal vagina. For the remaining four patients (80%), the distended vagina extended to the bottom of the pubococcygeal line and the obstruction level was determined as a distal vagina.

The surgical results were available for these five patients, revealing distal vaginal atresia in three (60%) and an imperforate hymen in two (40%) (Figure 2). None of these patients had any uterine structural anomalies. The ovaries were normal in four patients, while a unilateral endometrioma and hematosalpinx were co-present in one patient. Table 5 presents the findings of the patients with DVO.

Müllerian duct fusion anomaly was observed in two patients, both having uterus didelphys coexisting with a unilateral (left in one patient and right in the other) hematometocolpos. Renal agenesis was present on the same side as the hematometocolpos. Based on these findings, both patients were diagnosed with OHVIRA syndrome (Figure 3). The surgical results of only one patient were available, revealing obstructed hemivagina as the cause of hematometocolpos.



**Figure 2:** MRI showing the distal vagina distended with hemorrhagic content.

In all five patients who had been referred to MRI with a pre-diagnosis of vaginal obstruction-mass on US, the images revealed hematocolpos. No other cystic mass was observed.

In 17 of 26 patients, for whom the uterus could not be visualized on US, no uterus was detected in MRI, either. These patients were diagnosed with uterine agenesis, confirming the US findings. Among the remaining nine cases in the same group (no uterus visualization on US), the MRI showed a rudimentary uterus in seven patients, and uterus didelphys and obstructed hemivagina in two who had been diagnosed with uterine agenesis by US.

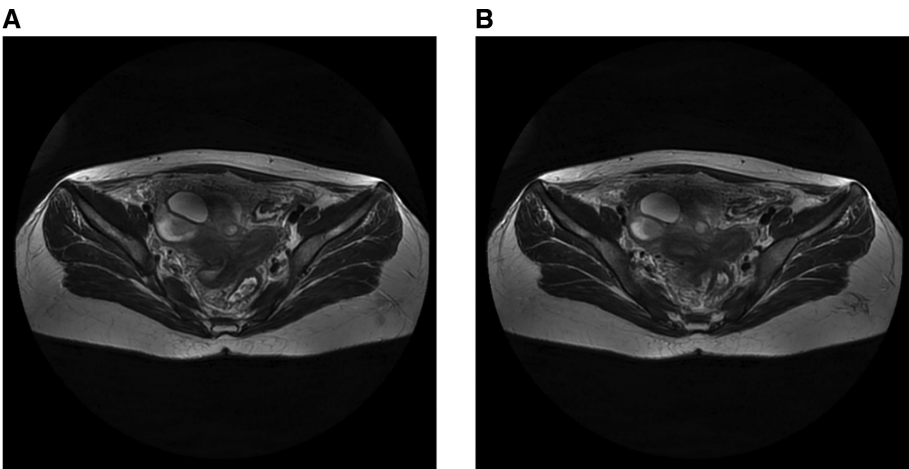
In 25 patients, the ovaries were not visualized by US. For 14 of these patients, MRI also could not detect the ovaries; thus, the patients were diagnosed with gonadal dysgenesis. Of the patients whose ovaries could not be evaluated on US, three had bilateral hypoplastic ovaries, one had a unilateral hypoplastic ovary, two had abnormal high position ovaries and five had normal ovaries. This group of patients were evaluated as having MDAs.

The MRI results of the 10 operated patients (four gonadal dysgenesis, one OHVIRA syndrome and five DVO cases) were compared with the surgical results. The MRI of the streak gonads in patients with gonadal dysgenesis was not successful, but malignancy was excluded. The accuracy rate of MRI was found to be 100% for the uterine findings in these patients. Similarly, the accuracy rate of MRI for the diagnosis of DVO, determination of the obstruction

**Table 5:** MRI and surgical findings of patients with DVO.

Patient number	Uterus	Cervix	Vagina	Surgery
1	Hematometocolpos	Hematometocolpos	Hematometocolpos	Imperforate hymen
2	Normal	Hematocolpos	Hematocolpos	Distal vaginal atresia
3	Hematometocolpos	Hematometocolpos	Hematometocolpos	Distal vaginal atresia
4	Normal	Hematocolpos	Hematocolpos	Imperforate hymen
5	Hematometocolpos	Hematometocolpos	Distal vaginal atresia	Distal vaginal atresia

DVO, distal vaginal obstruction.



**Figure 3:** MR image showing (A) unilateral obstructive hemivagina and (B) uterus didelphys. Right hemivagina is fluid filled (A), two separated uterin cavity is seen (B).



level, and detection of OHVIRA syndrome was found to be 100%.

## Discussion

Primary amenorrhea is a problem with the potential to affect the individual not only physically but also socially and psychologically [1]. It has greater importance due to being observed and diagnosed in adolescence. It is crucial to determine the most appropriate approach in this process. In patients presenting with primary amenorrhea, the first step is to determine the etiology after elimination of the possibility of pregnancy. The medical history, detailed physical examination determining secondary sex characteristics and hormonal analysis should be evaluated in all patients [2, 6–8]. Pelvic imaging, cytogenetic studies and karyotype analysis should also be performed where necessary [7, 8].

In various studies investigating the etiology of primary amenorrhea, the two most commonly implicated factors worldwide are gonadal dysgenesis and MDAs [1, 4, 9, 10]. Gonadal dysgenesis ranks first in the etiology in cases reported in western countries, whereas MDAs are the most prominent in cases from Asia and Africa [1, 4, 9, 10]. In the current imaging study, MDAs were found to be the most common factor in the etiology of primary amenorrhea. In a study conducted with the Thai population in Thailand, Müllerian agenesis was detected in 39.7% of the patients, gonadal dysgenesis in 35.3% and hypogonadotropic hypogonadism in 9.2% [4]. In our study, abdominal and pelvic MRIs were examined, and therefore, the hypogonadotropic hypogonadism etiology was not included in analysis. We found the rate of etiological factors to be 58.83% for MDAs and 41.17% for gonadal dysgenesis, which indicates similar percentages and frequency order compared to the Thai study. In another study undertaken in India, Kriplani et al. reported MDAs in 47% of the patients, followed by gonadal dysgenesis [1]. To the best of our knowledge, there are no wide-range studies investigating the etiology of primary amenorrhea in Turkey. Çakmak et al. examined the MRI of seven patients and found findings of MDAs in four and gonadal dysgenesis in two [11], consistent with our results.

In the literature on imaging findings related to gonadal dysgenesis, most publications are in the form of case reports [12, 13]. The results reported vary depending on the karyotype [13, 14]; however, the common finding is the lack of normal ovarian and testicular tissues [14]. It is possible to observe bilateral streak gonads, bilateral gonadal dysgenesis or unilateral streak gonad

accompanied by gonadal dysgenesis on the other side [14]. The uterus can be normal or rudimentary or may not be developed. For the imaging of these patients, US is the first preferred modality. However, US may not be sufficient due to its known disadvantages. In such cases, the preferred imaging modality is MRI. In the study conducted in patients with Turner syndrome, MRI allowed a more detailed and accurate evaluation of the internal genital organs compared to US [15]. Streak gonads were not detected by MRI in that study or other case reports in the literature [15]. In the current study, the gonads could not be visualized on MRI in any of the 14 patients with gonadal dysgenesis, and streak gonads were identified in four patients who underwent surgery. It is not usually possible to demonstrate streak gonads on MRI if there is no evidence of concomitant malignancy. However, considering that streak gonads have malignancy potential [12, 14], an MRI examination can be performed to exclude malignancy and identify cases that require biopsy. In our patients group, laparoscopy was preferred as the surgical method as there was no suspicion of malignancy. If there is a risk of malignancy, the inguinal approach or laparotomy may be necessary. MRI was effective in making a decision concerning the type of operation by excluding the possibility of a malignancy. The uterus and its internal structure (myometrium and endometrium) can be easily evaluated by MRI. In the presence of a normal uterus, pregnancy is possible through oocyte transfer or hormonal therapy [12]. A normal uterus finding is important for the management of the process in patients with gonadal dysgenesis. In the current study, the uterus was completely normal in three of the patients with gonadal dysgenesis.

MDAs are one of the most common etiologic factors in primary amenorrhea [1, 4, 9, 10]. The incidence of MDAs in the general population is reported to be 1–5% [5], which can increase to 15–25% in recurrent miscarriage and infertility cases [5] and reach 50% in primary amenorrhea [1]. In studies investigating the imaging findings related to MDAs, the most commonly investigated anomaly was MRKH syndrome, which is characterized by the absence of the uterus and the upper two-third segment of the vagina [16–20]. However, a rudimentary uterus and functional endometrium may be present in some cases [16, 19–21]. MRKH syndrome is the cause of primary amenorrhea in approximately 15% of cases [20]. In the current study, this rate was found to be 38.23%. Our higher rate can be attributed to the evaluation of only patients who had undergone a pelvic MRI, rather than all primary amenorrhea cases. The difference in these rates can be explained by the fact that pelvic imaging was not performed in cases where the patients were diagnosed

using the hormone panel, physical examination findings and medical history, and where the evaluation of pelvic anatomy is not required.

There are two types of MRKH syndrome: Type 1 is associated with isolated MDAs, and type 2 is accompanied by renal, ovarian and vertebral anomalies [17–20]. US can be used to differentiate the two types, but it is not sufficient in certain cases; e.g. in the presence of vertebral anomalies or ovaries that were in a higher position. Vertebral anomalies can also be detected by X-ray; however, this procedure involves exposure to radiation. In the evaluation of renal anomalies, it is not always possible to make a diagnosis using US because of gas superposition. In addition, in cases of renal agenesis, further investigation is necessary to rule out renal ectopia-hypoplasia or atrophy. MRI is the only imaging method that can simultaneously detect ovarian, renal and vertebral anomalies without the use of contrast media or exposure to ionizing radiation. MRI can detect uterine and vaginal anomalies with high accuracy. Pompili et al., who compared preoperative MRI and laparoscopy findings, reported the sensitivity and specificity of MRI as 100% [22]. In the current study, the patients did not have laparoscopy findings. However, in accordance with the literature, a rudimentary uterus was detected by MRI in three of the 13 patients, and MRI was also used to identify the type of MRKH. As in the literature, we detected renal anomalies (ectopia and agenesis) as the most common accompanying anomaly. We found vertebral anomalies in one patient. In MRKH syndrome, the ovaries are expected to be normal; however, in the literature, a variety of ovarian anomalies have been reported in cases with this syndrome (high position, hypoplasia, unilateral absence, etc.) [16–20, 22]. In our cases, high position and hypoplasia of ovaries were also observed, but none of the patients had bilateral absence of ovaries. In cases where the absence of uterus is accompanied by bilateral ovarian absence, androgen insensitivity syndrome should be considered in the differential diagnosis of MRKH, and the presence of abdominal and inguinal undescended testes should be investigated. None of the current cases presented with bilateral absence of ovaries or undescended testes.

Another cause of primary amenorrhea is DVO. In cases of primary amenorrhea, patients generally present with acute pelvic pain in addition to primary amenorrhea [1, 23, 24]. In these patients, the first diagnostic modality is usually US [23]. However, MRI is also increasingly used to determine the possible accompanying anomalies and perform a detailed evaluation of anatomy [23, 24]. Hematocolpos-hematometocolpos can also be detected in detail on MRI. It is also possible to determine the level of

obstruction. The level of obstruction is at the distal vagina in cases of imperforate hymen, most cases of transverse vaginal septum and in patients with distal vaginal atresia [24]. It may be impossible to differentiate the causes of obstruction, but the level can always be determined. In the current study, two of the five patients with DVO had an imperforate hymen and three had distal vaginal atresia. MRI revealed obstruction in all five patients, and one patient had a higher level of obstruction preoperatively. This patient was diagnosed with distal vaginal atresia, and her surgical results were consistent with the MRI findings. Endometrioma and hematosalpinx can also be seen in vaginal obstruction cases due to retrograde menstruation [23]. In the present study, endometrioma and hematosalpinx coexisted with DVO in one patient.

OHVIRA syndrome is a rare variant of MDAs, characterized by uterus didelphys, blind hemivagina and same-sided renal agenesis [25]. Patients usually present with cyclic pain caused by blind hemivagina. Primary amenorrhea is not an expected finding in this syndrome. However, the literature contains case reports on pediatric OHVIRA and coexistence of primary amenorrhea [25]. In our study, blind hemivagina, uterus didelphys and renal agenesis were detected on the MRI images of two patients who had clinical indications of primary amenorrhea. Due to blind hemivagina, the proximal vagina and uterine cavity on the same side were distended with hemorrhagic content.

The excellent soft tissue resolution of MRI allows a detailed evaluation of the pelvic anatomy. In this study, uterine and cervical morphology were evaluated in detail by MRI. There was no difficulty in evaluating these structures; however, in patients with DVO, it may be difficult to differentiate between transverse vaginal septum, atresic distal vagina and imperforate hymen by MRI. In this study, we were not able to differentiate between imperforate hymen and short segment distal vaginal atresia in one patient, but the level of vaginal obstruction was accurately detected. It is known that laparoscopy may not be sufficient to evaluate subperitoneal structures in MRKH syndrome [26]. Furthermore, it is not possible to evaluate endometrium directly by laparoscopy [26, 27]. Fedele et al. found MRI superior to laparoscopy in MRKH syndrome [27]. In our study, no diagnostic difficulties were encountered in these patients, and uterine morphology and internal structures were easily evaluated. In gonadal dysgenesis, there are still difficulties in the detection of streak gonads by MRI despite the developments in technology. Distinguishing gonads from intestinal loops is still challenging [26]. We were not successful in displaying streak gonads, as reported in the literature. This problem can be overcome in MRI systems with high magnetic field

strength or higher homogeneity. It can also be possible to obtain more detailed images by using a flex abdomen coil or pelvic multi-coil.

One of the limitations of our study is the small number of patients. Other limitations include the laparoscopic findings, karyotype analysis or cytogenetic studies not being available for all patients and the consequent inability to compare the MRI findings with these results.

## Conclusions

Primary amenorrhea is an important condition affecting the social and psychosexual status of the individual. Determining the appropriate surgical method is important to ensure sexual functioning and, if possible, fertility. Detailed anatomical data is critical in making an informed decision on surgery. The preoperative radiological method should be MRI, which allows visualizing the pelvic anatomy in detail and identifying possible anomalies.

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## References

- Kriplani A, Goyal M, Kachhawa G, Mahey R, Kulshrestha V. Etiology and management of primary amenorrhoea: a study of 102 cases at tertiary centre. *Taiwan J Obstet Gynecol* 2017;56:761–4.
- Deligeorgiou E, Athanasopoulos N, Tsimaris P, Dimopoulos KD, Vrachnis N, et al. Evaluation and management of adolescent amenorrhea. *Ann N Y Acad Sci* 2010;1205:23–32.
- Geckinli BB, Toksoy G, Sayar C, Soylemez MA, Yesil G, et al. Prevalence of X-aneuploidies, X-structural abnormalities and 46,XY sex reversal in Turkish women with primary amenorrhea or premature ovarian insufficiency. *Eur J Obstet Gynecol Reprod Biol* 2014;182:211–5.
- Tanmahasamut P, Rattanachaiyanont M, Dangrat C, Indhavivadhana S, Angsuwattana S, et al. Causes of primary amenorrhea: a report of 295 cases in Thailand. *J Obstet Gynaecol Res* 2012;38:297–301.
- Behr SC, Courtier JL, Qayyum A. Imaging of müllerian duct anomalies. *Radiographics* 2012;32:E233–50.
- Rosenberg HK. Sonography of the pelvis in patients with primary amenorrhea. *Endocrinol Metab Clin North Am* 2009;38:739–60.
- Al Jurayyan NA, Al-Jurayyan RN, Mohamed SH, Babiker AM, Al Otaibi HM. Radiological imaging of disorders of sex development (DSD). *Sudan J Paediatr* 2013;13:10–6.
- Klein DA, Poth MA. Amenorrhea: an approach to diagnosis and management. *Am Fam Physician* 2013;87:781–8.
- Reindollar RH, Tho SP, McDonough PG. Delayed puberty: an update study of 326 patients. *Trans Am Gynecol Obstet Soc* 1989;8:146–62.
- Reindollar RH, Byrd JR, McDonough PG. Delayed sexual development: a study of 252 patients. *Am J Obstet Gynecol* 1981;140:371–80.
- Çakmak V, Karabulut N, Çakmak P, Özari N, Kiroğlu Y. The efficiency of magnetic resonance imaging in the diagnosis of primary amenorrhea. *Pamukkale Med J* 2008;1:132–6.
- Azidah A, Nik Hazlina N, Aishah M. Swyer syndrome in a woman with pure 46, XY gonadal dysgenesis and a hypoplastic uterus. *Malays Fam Physician* 2013;8:58–61.
- Mannaerts D, Muys J, Blaumeiser B, Jacquemyn Y. A rare cause of primary amenorrhoea, the XY female with gonadal dysgenesis. *BMJ Case Rep* 2015;2015:bcr2014206609.
- Berberoğlu M, Şıklar Z, Ankara University Dsd Ethic Committee. The evaluation of cases with Y-chromosome gonadal dysgenesis: clinical experience over 18 years. *J Clin Res Pediatr Endocrinol* 2018;10:30–7.
- Maggio MC, De Pietro A, Porcelli P, Serraino F, Angileri T, et al. The predictive role of pelvic magnetic resonance in the follow up of spontaneous or induced puberty in turner syndrome. *Ital J Pediatr* 2018;44:24.
- Hall-Craggs MA, Williams CE, Pattison SH, Kirkham AP, Creighton SM. Mayer-Rokitansky-Kuster-Hauser syndrome: diagnosis with MR imaging. *Radiology* 2013;269:787–92.
- Giusti S, Fruzzetti E, Perini D, Fruzzetti F, Giusti P, et al. Diagnosis of a variant of Mayer-Rokitansky-Kuster-Hauser syndrome: useful MRI findings. *Abdom Imaging* 2011;36:753–5.
- Fiaschetti V, Taglieri A, Gisone V, Coco I, Simonetti G. Mayer-Rokitansky-Kuster-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.
- Rousset P, Raudrant D, Peyron N, Buy JN, Valette PJ, et al. Ultrasonography and MRI features of the Mayer-Rokitansky-Kuster-Hauser syndrome. *Clin Radiol* 2013;68:945–52.
- Kara T, Acu B, Beyhan M, Gökçe E. MRI in the diagnosis of Mayer-Rokitansky-Kuster-Hauser syndrome. *Diagn Interv Radiol* 2013;19:227–32.
- Boruah DK, Sanyal S, Gogoi BB, Mahanta K, Prakash A, et al. Spectrum of MRI appearance of Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome in primary amenorrhea patients. *J Clin Diagn Res* 2017;11:30–5.
- Pompili G, Munari A, Franceschelli G, Flor N, Meroni R, et al. Magnetic resonance imaging in the preoperative assessment of Mayer-Rokitansky-Kuster-Hauser syndrome. *Radiol Med* 2009;114:811–26.
- Economy KE, Barnewolt C, Laufer MR. A comparison of MRI and laparoscopy in detecting pelvic structures in cases of vaginal agenesis. *J Pediatr Adolesc Gynecol* 2002;15:101–4.
- Yoo RE, Cho JY, Kim SY, Kim SH. A systematic approach to the magnetic resonance imaging-based differential diagnosis of



- congenital Müllerian duct anomalies and their mimics. *Abdom Imaging* 2015;40:192–206.
25. Angotti R, Molinaro F, Bulotta AL, Bindi E, Cerchia E, et al. Herlyn-Werner-Wunderlich syndrome: an “early” onset case report and review of Literature. *Int J Surg Case Rep* 2015;11:59–63.
26. Reinhold C, Hricak H, Forstner R, Ascher SM, Bret PM, et al. Primary amenorrhea: evaluation with MR imaging. *Radiology* 1997;203:383–90.
27. Fedele L, Dorta M, Brioschi D, Guidici M, Villa L. Magnetic resonance imaging of unicornuate uterus. *Acta Obstet Gynecol Scand* 1990;69:511–3.