



# Value of MRI in the diagnosis and classification of oblique vaginal septum syndrome

**Objective :** To analyze the MRI of oblique vaginal septum syndrome (OVSS) and to assess the application of MRI in the diagnosis of OVSS.

**Methods :** Clinical and imaging data of 18 patients with OVSS that was confirmed by surgery were retrospectively analyzed. The patients underwent MRI examinations preoperatively. All cases involved conventional MRI (transverse T1 WI and T2 WI, coronal T2WI, and sagittal T2WI) and contrast enhancement. The manifestations of OVSS were summarized based on MRI examinations.

**Results :** The primary clinical symptoms of OVSS were painful menstruation and menorrhagia. The distribution of OVSS cases was as follows : type I, 1; type II, 12; type III, 5; and type IV, 0. Resections of the oblique vaginal septa were performed. 18 patients were diagnosed with OVSS based on MRI scans, and were confirmed to have OVSS intraoperatively surgery. Thus, the accuracy of MRI diagnosis was 94.7%(17/18). The MRI scans showed uterus didelphys, hydrocolpos, or hematocolpos and revealed ipsilateral renal agenesis in all 18 cases.

**Conclusion:** MRI has an important clinical role in the diagnosis, classification, and evaluation of patients with OVSS

**KEYWORDS:** oblique vaginal septum syndrome ■ vaginal disorders ■ magnetic resonance imaging ■ diagnosis

## Introduction

Oblique Vaginal Septum Syndrome (OVSS) is also referred to as Herlyn-Werner-Wunderlich Syndrome (HWWWS) [1]. OVSS is a disorder that occurs as a result of abnormal development of Muller's canal. OVSS is characterized by a bilateral uterus, double cervix, and vaginal septum with ipsilateral renal agenesis. The primary anatomic feature is an oblique septum that originates between the cervix, is attached to one side of the vagina wall, blocks the lateral passage, and leads to a series of vaginal obstruction symptoms [2]. OVSS was first reported abroad in 1922, and later in China by Meilu [3] in 1985. This type of malformation involving the reproductive tract is not rare. OVSS is often diagnosed in young patients. The clinical manifestations of OVSS are diverse, complex, nonspecific, and occult, so OVSS is difficult to diagnose accurately. MRI has good soft tissue resolution, no radiation, and multi-parameter, multi-planar imaging. OVSS has a double uterus and cervix, posterior oblique septal effusion and hemorrhage, and ipsilateral renal agenesis, which together facilitate the diagnosis. This study retrospectively analyzed the clinical data and MRI images of 18 patients with OVSS who were treated in our hospital, and the application of MRI in patients with OVSS is discussed.

## Methods

### ■ Clinical data

18 patients with OVSS confirmed by MRI and confirmed during surgery were collected from our hospital. The patients were 11–28 years of age (average,  $19.22 \pm 5.67$  years). OVSS developed in all patients after menarche; the age of menarche was 10–15 years and the average age of menarche was  $12.61 \pm 1.24$  years. The clinical manifestations of patients with OVSS include menstrual pain, abnormal menstruation, menorrhagia, and other symptoms.

### ■ Scanning instruments and parameters

MRI examinations were performed using a 3.0 TMRI scanning instrument Siemens Prisma (Magnetom Prisma, Siemens, Erlangen, Germany and GE Discovery MR750 (GE Medical Systems, Milwaukee, WI) with eight-channel phased array coils. The MRI sequences included a fast spin-echo sequence (TSE), line T1WI, T2WI axial, and T2WI sagittal and coronal images of diffusion-weighted imaging axial and coronal scans parallel to the long axis of the uterine fundus. The sequence parameters and scanning details were as follows : sagittal T1WI (TR/TE :500 ms/11 ms; field, 200 mm × 200 mm, matrix, 512 × 512; depth, 4 mm; and

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interval, 1.2 mm); T2WI (TR/TE :6760 ms / 123 ms; field, 200 mm × 200 mm; matrix, 320 × 320 mm; depth, 4 mm; and interval, 1.2 mm); T2WI (TR/TE :8430 ms/117 ms; field, 280 mm × 280 mm; matrix, 320 × 320; depth, 4.5 mm; and interval, 1.2 mm); diffusion-weighted imaging (TR/TE :6415 ms/92 ms; field, 238 mm × 280 mm; matrix, 160 × 136; depth, 4.5 mm; and interval, 1.2 mm); diffusion-weighted imaging axis (TR/TE :6415 ms/92 ms; field, 238 mm × 280 mm; matrix, 160 × 136; depth, 4.5 mm; and interval, 1.2 mm); coronary FLASH-T2WI fat suppression (TR/TE :7730 ms/123 ms; field, 200 mm × 200 mm; matrix, 320 × 320 mm; depth, 4 mm; and interval, 1.2mm);contrast agent (meglumine gadolinium [Gd-DTPA],0.1 mmol/kg); T1-VIBE-TRAE (TR, 3.9-4.6 ms; TE, 1.4-2.2 ms; FOV, 228-358 mm × 272-359 mm; matrix, 320-431 × 260-392; and depth, 3.5-5 mm); T1-VIBE-Corr (TR, 3.3-3.9 ms; TE, 1.2-1.6 ms; FOV, 339 × 339; matrix, 287 × 288; and depth, 2.5-3.5 mm). Before examination, the patients did not void and the bladder was filled during imaging.

### ■ Image analysis

All the imaging data were analyzed by three experienced senior diagnostic physicians who read the films independently, and observed the location of the oblique septum, the size of the posterior septal cavity, the signal, the relationship between the posterior septal cavity and the cervix, the development of the uterus and the contralateral vagina, and the accompanying changes of the bilateral appendages and urinary tract. The diagnostic standard was as follows : double uterus; double cervix; double vagina and unilateral vaginal atresia; and urinary system malformation on the side with vaginal atresia.

### ■ Clinical classification

According to the 1980 Rock and Jones Classification Criteria [4], the deformity was divided into the following three types : type I, oblique vaginal septum with complete atresia and incompatibility between the uterus; type II, oblique vaginal septum with incomplete atresia, including more than one opening ; and type III, oblique vaginal septum with complete atresia, but the two uteri are connected. In 2011, the Peking Union Medical College Hospital added type IV [5]. The characteristics of the type IV deformity were as follows : no foramen oblique septum; the posterior uterus has ipsilateral uterine adnexa; the contralateral uterine adnexa was absent; the cervical canal was blind; the other uterus had an ipsilateral uterine adnexa;

the contralateral uterine adnexa was absent; the cervix developed normally; the two uteri were completely separated and the ipsilateral kidney was absent; and the cervical end of the uterus was completely atretic distal to the septate uterus, so the patient had undergone many years of swelling, which flowed back to the pelvic cavity and may be accompanied by progressive dysmenorrhea and recurrent ovarian endometriotic cysts.

## Results

The age range of 18 patients was 11–28 years and the average age was 19.2 years. All of the patients developed OVSS after menarche. The age of menarche was 10–15 years and the average age of menarche was 12.6 years. The clinical manifestations of patients with OVSS included menstrual pain, abnormal menstruation, menorrhagia, and other symptoms. The clinical symptoms and MRI findings are shown in TABLE 1. The MRI findings were as follows : double uterus; double cervix deformity in 18 patients; anteverted, anteflexed uterus in 13 patients; and posterior uterus in 5 patients. 10 patients had a right oblique septum complicated with absence of the right kidney and 8 patients had a left oblique septum with absence of the left kidney. Thirteen patients had hemorrhage in the posterior aspect of the oblique septum and 5 patients of posterior septum of oblique septum. 18 patients had obstructive hemorrhage and effusions (uterus, cervix, posterior septum, and pelvic cavity), and clearly showed ipsilateral kidney absence. The classification of the 18 patients was as follows : type I, 1; type II, 12; type III, 5; and type IV, 0. All 18 patients were diagnosed with OVSS by MRI pre-operatively (TABLES 1-5) (FIGURES 1-4, 5A-5E).

## Discussion

### ■ Etiology and pathogenesis

OVSS was first reported by Purslow [6] in 1922. Herlyn and Werner [7] and Wunderlich [8] described OVSS in 1971 and 1976. Therefore, this disease is also referred to as Herlyn-Werner-Wunderlich syndrome. This developmental disorder is also referred to in the literature as obstructed hemivagina and ipsilateral renal anomaly syndrome (OHVIRAS). The pathogenesis of OVSS is not completely clear, but most scholars agree that the original ducts of the urinary and reproductive systems are mesonephric and accessory mesonephric ducts. During embryonic development, both ducts (mesonephric and accessory mesonephric ducts) originate in the urogenital ridge, and

**Table 1. Clinical data and MRI findings of 18 patients with OVSS.**

Case	Age (years old)	Menarche (years old)	Clinical symptom	Posterior septal effluents	The oblique vaginal septum, Urinary system	Clinical classification
1	12	12	Abdominal pain	hematocele	Right cervix, Absence of right kidney	Type 2
2	26	13	Abdominal pain	hematocele	Right cervix, Absence of right kidney	Type 2
3	11	10	Abdominal pain with fever	hematocele	Left cervix, vagina, Absence of left kidney	Type 2
4	20	13	Menostaxis, lower abdominal mass	hematocele	Right cervix, Absence of right kidney	Type 1
5	26	11	menostaxis	hematocele	Left cervix, Absence of left kidney	Type 2
6	25	13	Abdominal pain	effusion	Right cervix, Absence of right kidney	Type 2
7	24	14	Abdominal pain	effusion(hematocele)	Left cervix, vagina , Absence of left kidney	Type 2
8	23	15	Abdominal pain	hematocele/empyemata	Left cervix, vagina , Absence of left kidney	Type 2
9	21	12	Abdominal pain	hematocele	Left cervix, Absence of left kidney	Type 3
10	28	12	Abdominal pain	hematocele	Right cervix, vagina, Absence of right kidney	Type 2
11	14	12	Menstrual irregularity	hematocele	Right cervix, vagina ,Absence of right kidney	Type 2
12	18	13	Abdominal pain	hematocele	Left cervix, vagina , Absence of left kidney	Type 3
13	19	13	Menstrual irregularity	hematocele	Right cervix, Absence of right kidney	Type 2
14	12	11	Abdominal pain, Menstrual irregularity	Hematocele, effusion	Left cervix, Absence of left kidney	Type 2
15	16	14	Menstrual irregularity	hematocele	Right cervix, vagina , Absence of right kidney	Type 3
16	24	14	Menstrual irregularity	hematocele	Right cervix, vagina , Absence of right kidney	Type 3
17	12	13	Menstrual irregularity	effusion, hematocele	Right cervix, vagina , Absence of right kidney	Type 3
18	15	12	Menstrual irregularity	hematocele	Left cervix, Absence of left kidney	Type 2

**Table 2. Composition of clinical symptoms.**

Clinical symptom	Number of cases	Composition (%)
Abdominal pain	8	44.4%
Abdominal pain with fever	1	5.5%
Menostaxis, lower abdominal mass	1	5.5%
Menostaxis	1	5.5%
Menstrual irregularity	6	33.3%
Abdominal pain, Menstrual irregularity	1	5.5%

**Table 3. Composition of posterior septal effluents.**

Posterior septal effluents	Number of cases	Composition (%)
Hematocele	13	72%
Effusion	1	5.5%
Hematocele/empyemata	1	5.5%
Effusion (hematocele)	1	5.5%
Hematocele, Effusion	2	11.1%

**Table 4. Composition of the oblique vaginal septum and urinary system.**

Oblique vaginal septum, Urinary system	Number of cases	Composition (%)
Right cervix ,Absence of right kidney	5	27%
Right cervix, vagina ,Absence of right kidney	5	27%
Left cervix, Absence of left kidney	4	22%
Left cervix, vagina, Absence of left kidney	4	22%

**Table 5. Composition of clinical classification.**

Oblique vaginal septum, Urinary system	Number of cases	Composition (%)
Type 1	1	5.50%
Type 2	12	72%
Type 3	5	27.70%

the development of Müllerian ducts (accessory mesonephric duct) depend on the development of the mesonephric duct. Development of the ipsilateral Mullerian duct is affected when the unilateral mesonephros is hypoplastic in the embryonic stage. OVSS may be due to unilateral mesonephric dysplasia (thus causing ipsilateral urinary malformations), which results in failure of bilateral Mullerian tube fusion, and the development and formation of a double uterus and cervix, and a vaginal malformation. Contact between the Mullerian nodule and genitourinary sinus is also affected, resulting in the formation of an unobstructed vagina on the ipsilateral side [9]. The typical manifestations are a double uterus, double cervix, double vagina, and unilateral vagina with complete or incomplete atresia, accompanied by a ipsilateral urinary system malformation (ipsilateral renal absence is more common) [10]. Because the reproductive and urinary systems interact with each other during the development of the primitive embryo, patients with OVSS are often complicated by urinary system malformations, and most of which are an oblique septum,

ipsilateral renal absence and contralateral renal compensatory enlargement [11,12]. Among the 18 patients, all were complicated by urinary system malformations and ipsilateral kidney absence.

### ■ Clinical characteristics

The clinical manifestations of OVSS are closely related to the degree of oblique vaginal septum occlusion. The clinical manifestations are also derived from the basis of obstruction. Posterior hemorrhage, empyema or poor drainage can cause a series of symptoms and signs, and even induce acute abdomen [13]. During menstruation, the descending passage of menstrual blood is blocked and accumulated in the blind cavity formed by oblique septum. Increased amount of hematocele will lead to hematocele in the affected side of uterus, and further through the fallopian tube lead to pelvic hematocele [14]. The main clinical manifestations are periodic lower abdominal pain, dysmenorrhea, irregular menstruation, vaginal purulence, lower abdominal mass and a series of symptoms caused by pelvic inflammation.

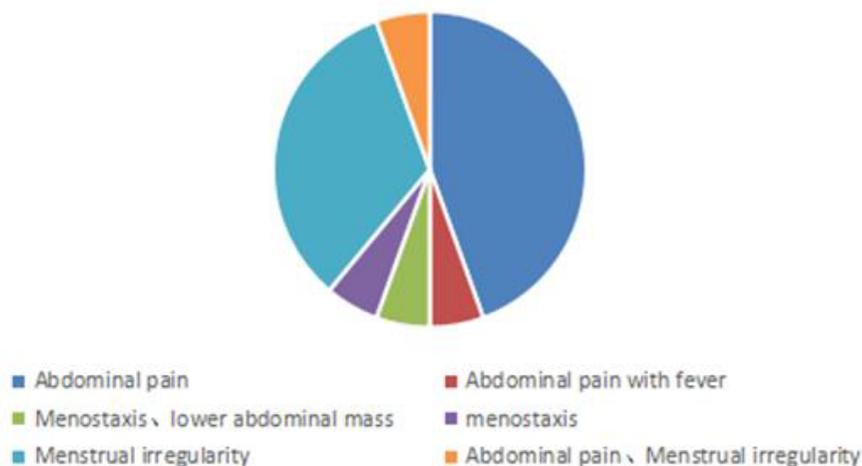


Figure 1. Composition of clinical symptoms.

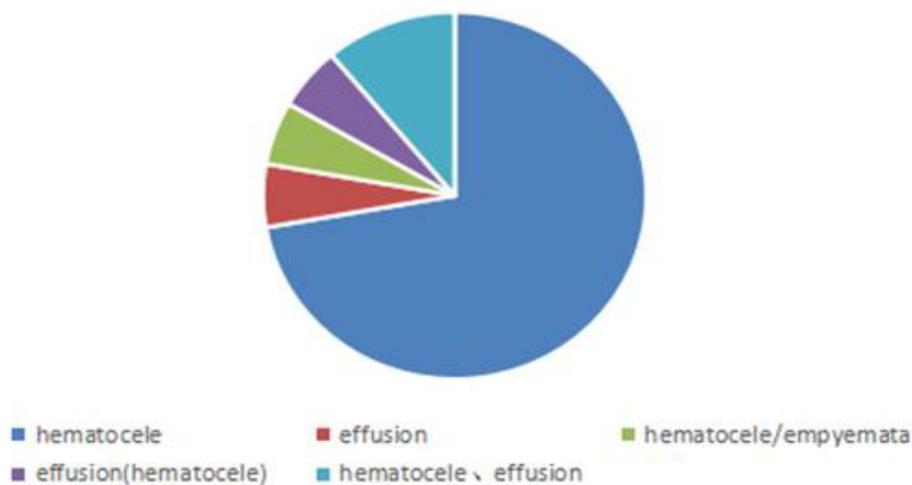


Figure 2. Composition of posterior septal effluents.

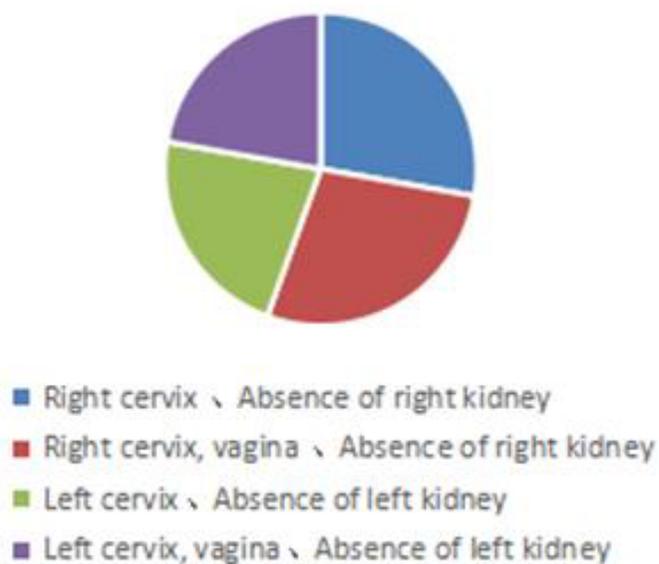
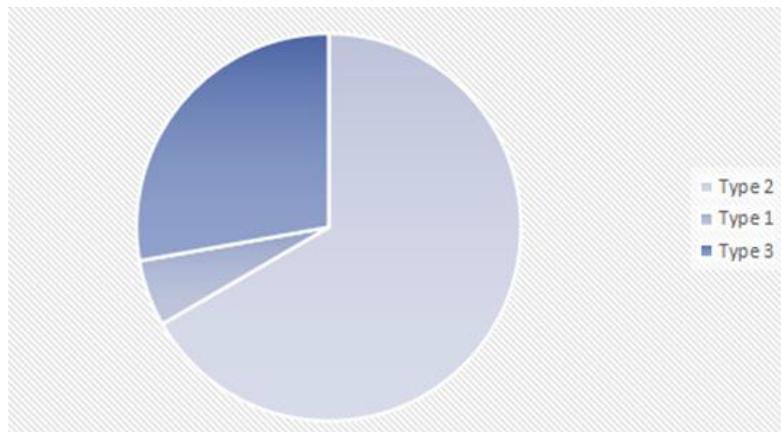
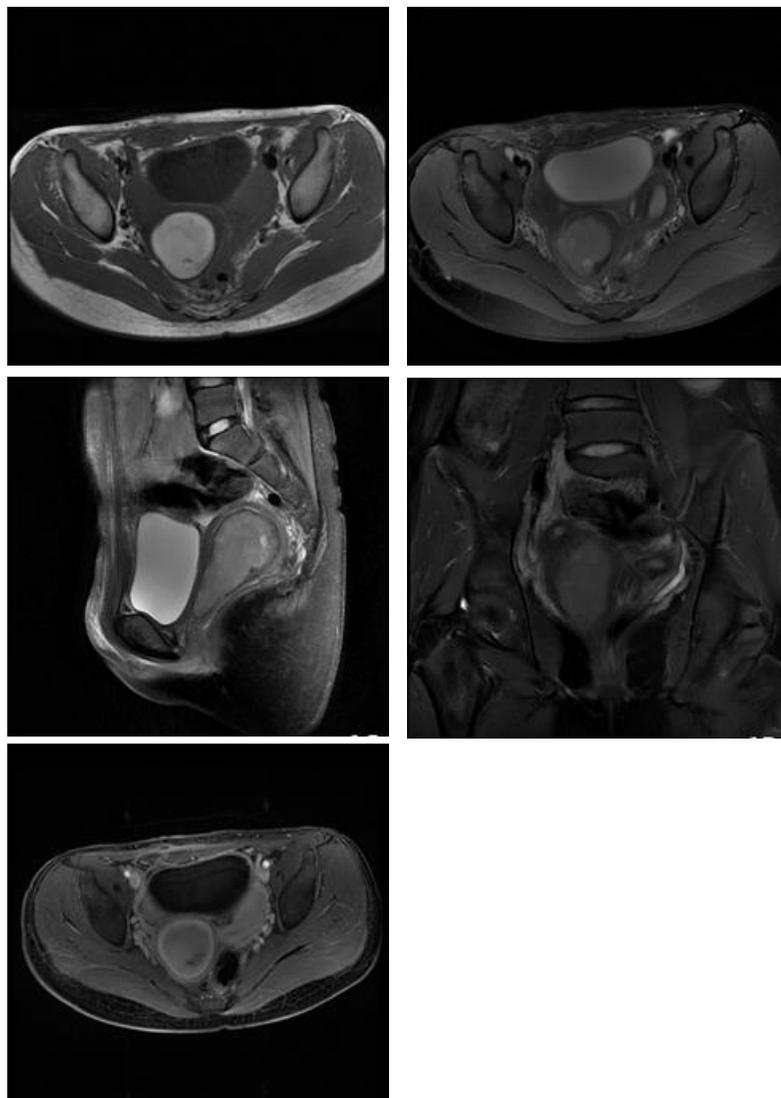


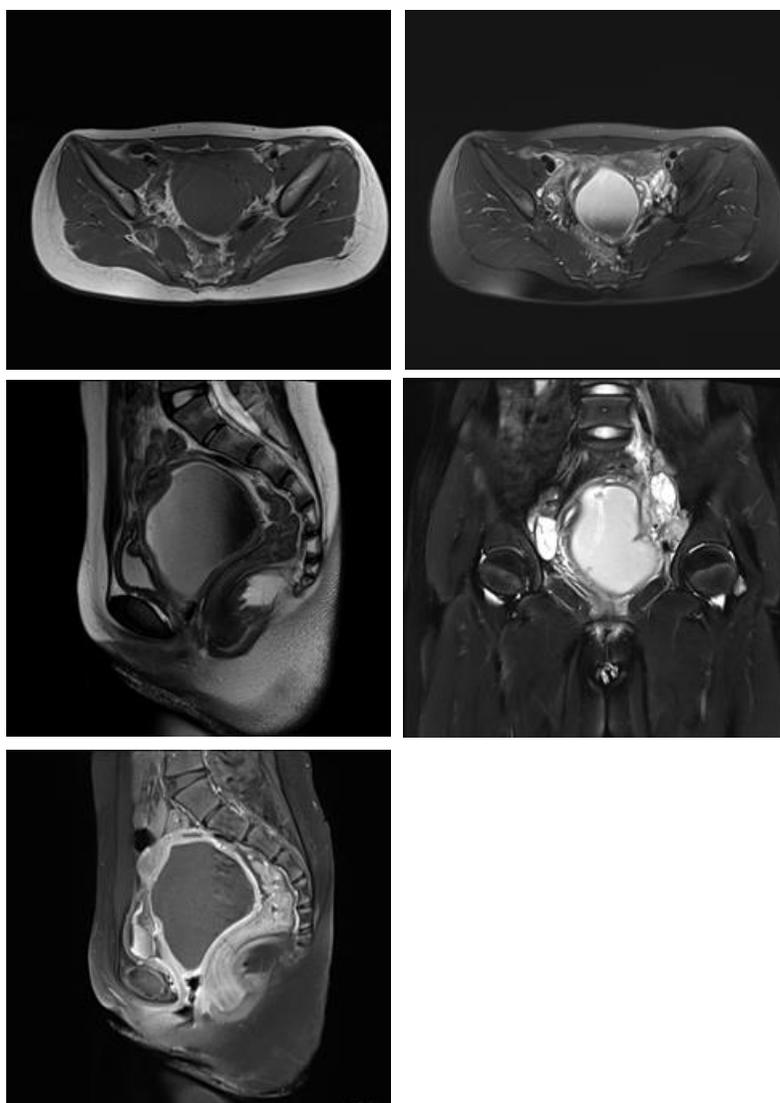
Figure 3. Percentage of oblique vaginal septum and urinary systems.



**Figure 4. Percentage of clinical classification.**



**Figure 5. OVSS patient is a 13-year-old female. (Figures 5A-5D)** Axial T1WI, axial T2WI, sagittal T2WI, coronal T2WI, and enhanced posterior axis. **(Figure 5E)** Two uteri are seen in the pelvic cavity on both sides; The right uterine cavity, cervical tube, and vaginal segment are dilated, and hemorrhage (black arrow) was noted in the pelvic cavity.



**Figure 6. OVSS patient is a 16-year-old female. (Figures 6A-6D)** Axial T1WI, axial T2WI, sagittal T2WI, coronal T2WI, and 5E enhanced posterior axis, respectively. The pelvic cavity showed two uterine bodies. The lower part of the left cervix is obviously widened, and a large amount of hemorrhage and effusion are noted in the left cervix. The vaginal septum (**Figures 6C & 6E black arrow**) appeared to be compressed and displaced to the right cervix.

### ■ Clinical classification

According to the 1980 classification criteria of Rock and Jones [15], the deformity was classified into three types : type I , oblique vaginal septum is completely atretic and the two uteri are not connected; type II , the oblique vaginal septum is incompletely atretic with more than one opening ; and type III , the oblique vaginal septum is completely atretic, but the two uteri are connected Type I, because of the diagonal septate uterus, the cervix and the opposite side of the cervix are completely disconnected, and the blood cannot be discharged through the uterus. The time interval between clinical onset and menarche is short. Types II and III

because of the oblique septum or the existence of a fistula between the cervix, play a role in drainage of transvaginal blood. As a result, the interval between the onset and menarche is longer. The distribution of anomalies was as follows : type I,1; type II, 12; and type III, 5. In 2011, the type IV classification was advanced in Beijing Union Hospital [16]. The characteristics of type IV are as follows : no foramen oblique septum; the posterior uterus has ipsilateral uterine adnexa; the contralateral uterine adnexa was absent; the cervical canal was blind; the other uterus had an ipsilateral uterine adnexa; the contralateral uterine adnexa was absent; the cervix developed normally; the two uteri were completely separated and the ipsilateral kidney

was absent; and the cervical end of the uterus was completely atretic distal to the septate uterus. There were no type IV cases in this group of patients.

### ■ MRI features

The early detection and diagnosis of OVSS based on MRI features is beneficial for the prevention of associated diseases and protection of fertility and urinary system function. The diversity of clinical manifestations increases the difficulty of diagnosis and treatment. The key to successful treatment is a correct preoperative diagnosis [17]. MRI has the advantages of being noninvasive, non-ionizing radiation, painless, high soft tissue resolution, and scanning with multiple parameters, arbitrary plane, and azimuth. MRI can display the changes in uterine shape and signal, eliminate the interference of other organs in the pelvic cavity, clearly display the tissue signals of each layer of the uterus, and clearly display the inner structure of the uterine cavity and the outer contour of the uterine fundus. MRI can demonstrate the relationship between the posterior septal cavity effusion and cervix, and has unique advantages in diagnosing OVSS. The characteristics of OVSS include a bilateral uterus, double cervix, hemorrhage in the septal cavity, and a contralateral normal vagina [18]. MRI can show the type of uterine malformation, the location and course of the oblique septum, and the relationship between the septum and the cervical effusion. Diagnosis of the endometrial and cervical development and determination of the nature of the effusion were more advantageous in displaying the oblique septum and vaginal panorama. The hind cavity hemorrhage presented a hyperintensity in T1WI or a hypointensity in T2WI, and empyema in the septum was a hyperintensity in DWI. There were still some difficulties in the differential diagnosis of MRI with or without perforation and a fistula involving the oblique septum. The characteristics of the fluid signal in the posterior septum of the 18 cases were summarized as follows: hypointensity in T1WI and hyperintensity in T2WI; hyperintensity in DWI; first considering empyema; hyperintensity in T1WI and hypointensity in T2WI considering hemorrhage, but complicated and variable hemorrhage signal; and hemorrhage and pus can co-exist. In the sagittal position, the lower end of the effusion was sharp and was distinguished from hymen atresia (the lower end is wider and flat). The MRI findings were as follows: double uterus; double cervix deformity

in 18 cases, including an anterior flexed uterus; a double cervix deformity in 18 cases, including 13 cases of anteverted, anterior flexed uterus; and 5 cases of a posterior uterus. There were 10 patients with a right oblique septum, 8 patients with a left oblique septum, 13 patients with a posterior septal hemorrhage, and 5 patients with a posterior septal septum. In this study, all patients were diagnosed by MRI preoperatively, and involvement of the uterus, cervix, and vagina was clearly demonstrated (FIGURES 6A-6E).

### ■ Differential diagnosis

**1) Hymen atresia:** Due to menarche atresia, menstrual blood cannot be discharged after menarche, resulting in uterine and fallopian tube bleeding. "Acute urinary retention" is often seen in the clinic setting. MRI and ultrasound show obvious dilation of the uterine cavity, a thin uterine wall, direct continuation of a dilated uterine cavity, and a dilated vagina. The vagina was oblong and distended below the level of the perineum.

**2) Vaginal wall cyst:** Most of the patients had no self-conscious symptoms, cystic masses were located under the vaginal mucosa, the surface was smooth, palpation did not elicit pain, and OVSS patients have progressive dysmenorrhea.

**3) Pelvic inflammation and abscess:** The main manifestation of the patient was lower abdominal pain, increased vaginal secretions, fever, ultrasound examination revealed a pelvic effusion, and the pelvic mass boundary was not clear.

**4) Endometriosis:** Most of the symptoms were secondary, dysmenorrhea was progressive, single or multiple cystic masses were noted by ultrasound, the margin was irregular, and adhesions and an effusion were present. Diagnosis needs to be combined with the medical history.

**5) Dysfunctional uterine bleeding:** The clinical symptom was similar to types II and III disease and the deformity of the genitourinary and urinary systems.

**6) Bilateral uterus with vaginal mediastinum, double uterus, bilateral vaginal patency, and no intravaginal blood retention:** There was no posterior cavity of the vaginal septum and renal absence of oblique septum. The main manifestations of type I OVSS were dysmenorrhea, which were easily misdiagnosed as primary dysmenorrhea, a vaginal wall cyst,

and a pelvic mass. Type II and III patients were mainly characterized by vaginal bleeding or a purulent secretion, and the patients with type I OVSS had dysmenorrhea and were easily misdiagnosed with primary dysmenorrhea, a vaginal cyst, a pelvic mass, vaginitis, pelvic inflammation, and adolescent dysfunctional uterine bleeding. The main distinguishing points were female genital and urinary system malformations that were characterized by a double uterus, double cervix, double vagina, complete or incomplete atresia of one side of the vagina, and often accompanied by an ipsilateral kidney and ureter absence or other urinary system malformations [19].

### ■ Treatment

For OVSS patients, vaginal oblique septum resection is the most effective treatment. Oblique septum resection as early as possible is the focus of diagnosis and treatment [20]. The purpose of the operation is to relieve the obstruction of the genital tract and avoid the occurrence of severe complications. The best time for the operation is during the menstrual period. The tension of the vaginal wall mass is large and easy to locate.

### Conclusion

In summary, OVSS is a special type of female

genitourinary system malformation. The clinical symptoms are complex and changeable, and the symptoms are not specific, so can be easily misdiagnosed in the clinic. MRI imaging can obtain arbitrary plane images with good soft tissue resolution. MRI is sensitive to soft tissue and liquid components, and can clearly show the relationship between a posterior septum effusion and the cervix, and can make a preliminary judgement on the composition of the intracavitary effusion, which is of great significance for the diagnosis of OVSS. MRI examination preoperatively can clearly display the abnormal structure of the uterus and vagina, evaluate the condition of effusion in the posterior septum cavity, whether or not it is complicated with other abnormalities and urinary system malformations, and provide reference information for the formulation of an operative plan.

### Acknowledgement

The authors of the present study would like to thank the patients and their families for support and collaboration, and we thank International Science Editing ( <http://www.internationalscienceediting.com>) for editing this manuscript.

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