ABSTRACT

The triad of uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare entity. These cases are difficult to diagnose due to heterogeneous presentation and rarity of the condition. Ultrasonography and magnetic resonance imaging are important for initial diagnosis; however, laparoscopy is required for the confirmation of the diagnosis. Early and accurate diagnosis of this condition followed by prompt surgical therapy is essential to prevent complications and to preserve future fertility. Here we report a case of uterus didelphys with hemivagina and ipsilateral renal agenesis with hematometra and hematosalpinx in a young girl.

Keywords: Didelphic uterus, Hematometra, Hematosalpinx, Hemivagina, Renal agenesis.


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INTRODUCTION

Mullerian duct anomalies (MDA) are developmental defects involving female reproductive system with presentations varying from undiagnosed very mild segmental abnormalities to unicornuate, bicornuate, didelphic uterus, and to even absence of uterus and vagina. Incidence of MDA is 0.5 to 5%, while of didelphic uterus is 1 in 300,000.1,2 Mullerian anomalies with renal agenesis occur as a result of simultaneous arrest of development of mullerian and metanephric ducts at 8 weeks of gestational age.3 Didelphic uterus with obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is quite a rare anomaly. Incidence of this rare anomaly is not well known but it ranges between 0.1 and 3.8%.4 Such patients usually present with pelvic pain at menarche, while others present with features of abscess formation like pain, fever, tenderness. A high index of suspicion is required for timely diagnoses and management of this rare condition so as to prevent its further complications.5 Here, we present a rare case of uterine didelphys with hemivagina with ipsilateral renal agenesis in a 13-year-old girl who presented with left hematometra and hematosalpinx.
normal location. The patient was catheterized. Vulvovaginal examination revealed normal uterus with $5 \times 5$ cm cystic to firm mass in left fornix, complete vertical vaginal septum, and a transverse vaginal septum on left side. Vaginoscopy with hysteroscopy showed vagina, right cervix, right uterine cavity normal with right normal cornual opening as shown in Figures 5 and 6. Laparoscopy confirmed the findings of left hematometra with hematosalpinx
Management of Hematometra in Single Horn of Didelphic Uterus with OHVIRA Syndrome

Vaginoplasty was planned on the left side. Laparotomy was performed and a small vertical midline incision was made on left horn of uterus following which old chocolate-colored blood was drained out, as shown in Figure 8. Hysteroscopy was done from uterine incision through abdominal route which showed normal endometrium. Vaginoplasty was done by Abbe-McIndoe method. Dilator was inserted into the uterine cavity through abdominal route and felt in new vaginal plane. Incision was given over dilator and a new opening was created, thus connecting cervix to neovagina. Malecots catheter was kept in uterine cavity through the vaginal route to maintain the patency of the tract. Split thickness graft (STG) was taken from medial aspect of thigh; vaginal mould was prepared; and mould inserted in left vagina. Uterine incision was closed with vicryl 1-0. After 8 days, vaginoscopy revealed well-epithelialized left neovagina and hysteroscopy revealed normal cavity with proliferative endometrium on left side, as shown in Figures 9 and 10. Right-sided natural vagina could be easily differentiated from left-sided neovagina by a vertical septum and by different types of epithelium, as shown in Figure 11. Right-sided cervix and endometrial
cavity were normal. Malecots catheter was kept *in situ* for 3 months after which vaginal septum resection was done. Vaginoscopy with hysteroscopy showed single vagina with two separate cervices with two uterine cavities with healthy endometrium on either side. On follow-up, patient was asymptomatic with normal regular menstrual cycles.

**DISCUSSION**

Stassart et al⁶ observed from 1953 to 1991, a series of 15 such cases and concluded that the most common presentation in these patients is postmenarchal pelvic pain and/or dysmenorrhea with the presence or absence of palpable pelvic mass and/or vaginal discharge if a communication exists between the two hemivaginas. Various diagnostic modalities include transvaginal sonography (TVS), hysterosalpingography (HSG), computed tomography scan, MRI, laparoscopy, and hysteroscopy. Transvaginal sonography can accurately classify mullerian anomalies in 85 to 92% cases, while MRI has got a diagnostic accuracy in 96 to 100% of cases.⁷ Orazi et al⁸ observed 11 adolescent cases and concluded that though ultrasonography may help in the diagnosis of this rare entity, but MRI definitely has an upper hand as it provides detailed uterine morphological details, while also providing correct diagnosis about vaginal channel, i.e., obstructed or nonobstructed. Salim et al⁹ worked on the diagnostic accuracy of three-dimensional (3D) sonography in congenital uterine anomalies and found that it can diagnose them with acceptable reproducibility. Zurawin et al¹⁰ recommended laparoscopy to be considered as gold standard in diagnosing mullerian anomalies.

If an absent or dysplastic kidney is seen in a female fetus on an antenatal scan or a girl postnatally, then it is advisable to screen her for mullerian anomalies also.³ This OHVIRA syndrome also known as Herlyn Werner Wunderlich syndrome is rare and consists of a triad of didelphic uterus, obstructed hemivagina, and ipsilateral renal agenesis.¹¹ Wilson¹², first described the association of didelphys uterus with obstructed hemivagina and absent ipsilateral kidney.

Didelphic uterus has maximum association of renal abnormality of about 20% with it, as compared to any other mullerian anomaly.¹³ Their presentation varies from cyclical abdominal pain, vaginal mass, vaginal discharge to urinary symptoms. The treatment of choice for OHVIRA syndrome involves transverse vaginal septal resection with preservation of normal anatomy with reproductive capabilities.¹⁴ Modifications in this as well as advancements have always been reported, like hysteroscopic resection of vaginal septum to preserve hymenal integrity.¹⁵ Purandare et al¹⁶ also conserved both the uteri of the patient who presented to them with similar complaints. Various methods of resection of transverse vaginal septum have been proposed with resultant successful term pregnancy. Its management involves correct diagnosis, counseling of the patient and parents, and then undertaking definitive corrective surgery, which would allow her to have healthy sexual life with successful reproductive outcomes.¹⁶ In this case, we tried to conserve both of her uteri by establishing a communication between the left uterine cavity and left neovagina so as to prevent formation of hematometra. In earlier times, hemihysterectomy was the management for such patients, but it is no longer preferred as pregnancy has been reported in both horns with equal incidence after surgical correction. Altchek and Paciuç¹⁷ reported pregnancy occurring twice in a previously obstructed didelphys uterus after surgical correction. Therefore, every effort should be made to preserve the obstructed uterus.

**CONCLUSION**

Obstructed hemivagina and ipsilateral renal agenesis syndrome is a rare congenital anomaly, which is often missed clinically and is difficult to diagnose. Magnetic resonance imaging is the diagnostic modality of choice. Diagnostic laparoscopy provides additional information regarding pelvic anatomy. A prompt and accurate diagnosis with appropriate surgery aimed at conserving the uterus is required to relieve the symptoms, to prevent complications, and to preserve the sexual and reproductive function.

**REFERENCES**