

Obstructed Hemivagina and Ipsilateral Renal Anomaly—A Reproductive Surgical Unit's Experience

Dear Editor,

Uterus didelphys with an imperforate hemivagina is an embryonic malformation of the genitourinary system of the female that occurs between the 12th and 16th weeks of pregnancy. This defect may manifest itself as a duplication of all or part of the female reproductive system. The obstruction of one hemivagina will block outflow, resulting in complications such as haematocolpos and haematometra. The persistence of this situation may also be complicated by the occurrence of endometriosis as a result of blood reflux into the abdominal cavity. These anomalies are frequently accompanied by kidney and urinary tract malformations (i.e. kidney agenesis and dysplasia, double collecting system, ectopic ureter) on the same side as the defect.¹

Materials and Methods

A retrospective case review revealed 4 patients being diagnosed with uterine didelphys with obstructed hemivagina from August 2010 to May 2012. All patients underwent examination under anesthesia, hysteroscopic resection of the obstructing vaginal septum with therapeutic laparoscopy.²

Results

Please refer to Table 1. The mean age at diagnosis was 15 years (range, 12 to 22 years). Menarche ranged from 12 to 13 years of age, with a mean of 12.5 years. All patients had a history of regular menses with cyclic pelvic pain. Two patients had suspected uterine didelphys discovered through the presence of other developmental anomalies that prompted earlier workup. All patients had the diagnosis made after radiological investigations. Three of our patients, in their teenage years, presented with compressive symptoms such as backache and acute retention of urine to the paediatrics department. All had obstructed hemivagina, ipsilateral renal anomaly and 1 had bilateral renal anomaly.

Discussion

Ultrasound and magnetic resonance imaging (MRI) were the most frequently used modalities for initial imaging. MRI, the most sensitive imaging modality for congenital anomalies, was used in only 2 cases. In previous studies,

MRI was found to be 100% sensitive and 83% to 100% specific.³

Because of the interesting clinical picture, didelphys uterus with unilateral obstruction is often published in the literature. This anomaly usually presents with acute or chronic pelvic pain following menarche. There is a large variation in time to accurate diagnosis. This may be due to several factors. Since only one uterine horn is obstructed, the patient menstruates regularly from the other horn delaying the diagnosis of outflow obstruction that would more readily be made had she presented with amenorrhea and cyclic pain. As it is rare, it is not on the list of differential diagnosis that is considered. When these patients present to doctors with abdominal pain, they may be given analgesics e.g. nonsteroidal anti-inflammatory drugs (NSAIDs). This may inadvertently cause a delay in diagnosis because they diminish or eliminate menses.⁴ In this report, we have observed that this method fails. That patient was sent to the orthopaedic surgeon who was not conversant in congenital anomalies, and subsequently, failed to make the correct diagnosis. It is common for these patients to have cyclic pelvic pain which in the presence of menstrual outflow obstruction results in retrograde flow, haematometra, haematosalpinx, and endometriosis.

The review of the literature in the management of such cases have revealed several approaches. Traditionally, it has been under examination under anaesthesia with scapel and blade for resection of the vaginal septum, resulting in hymenectomy.⁵ In some cases, because of the abdominal symptoms, laparotomy has been done inadvertently, with a hysterectomy in one case.⁴

The aims of the management is to confirm the diagnosis, treat symptoms, improve reproductive potential and preserve hymen (important in some cultures).

Resection of the vaginal septum can be done with hysteroscopy with ultrasound guidance or with additional laparoscopy. Under hysteroscopic resection, a resectoscope is used. The benefits include resection under direct visualisation, preservation of the hymen and the ability to resect a high small haematocolpus. Patient 1 was a challenging case. She had a high small haematocolpus with a small button hole opening which was picked up under magnification by hysteroscopy.

Table 1. Summary of the Results from Imaging Modalities Compared with Intra-operative Findings and Follow-up.

Patient*	Imaging Modality	Scan Findings	Operative Findings	Follow-up
1	U/S	Bicornuate uterus, possible haematocolpus and absent left kidney.	Vaginoscopy/hysteroscopy—Right cervix seen, small button hole in mid point of septum with possible pus drainage.	29 months: well
	MRI	Uterine didelphys with left haematometrocolpus, hypoplastic ectopic left kidney. Possible ectopic left ureter insertion in vagina.	Laparoscopy—Uterine didelphys, both tubes normal with slight left peritubal adhesions. Cystoscopy—Right ureter orifice seen, left ureteric orifice not seen.	
2	U/S	Uterine didelphys, obstruction of right vagina by septum.	Vaginoscopy/hysteroscopy—Low haematocolpus (obstructed), complete septum. Laparoscopy—Uterine didelphys, right mildly dilated haematosalpinx.	50 months: well
3	U/S	Uterine didelphys with obstruction of the right vagina, left uterine horn appears unaffected, possible vaginal septum obstructing the right side. Haematocolpus with bicornuate uterus, absent right kidney.	Vaginoscopy/hysteroscopy—Left 13 cm haematocolpus (obstructed). Laparoscopy—Bicornuate uterus, both tubes and ovaries normal (Figs. 1 and 2).	20 months: well
	CT	Haematocolpus with bicornuate uterus, absent right kidney.		
4	MRI	Complete septate uterus with left haematometrocolpos and left haematosalpinx, left renal agenesis and lumbar scoliosis (Fig. 3).	Vaginoscopy/hysteroscopy—10 cm left haematocolpus, complete septum. Laparoscopy—Bicornuate uterus with left blocked haematosalpinx- neosalpingostomy done, endometriosis on posterior uterine surface and left ovarian fossa.	29 months: well

U/S: Ultrasound; MRI: Magnetic resonance imaging; CT: Computed tomography
*In all cases, hymen was preserved.

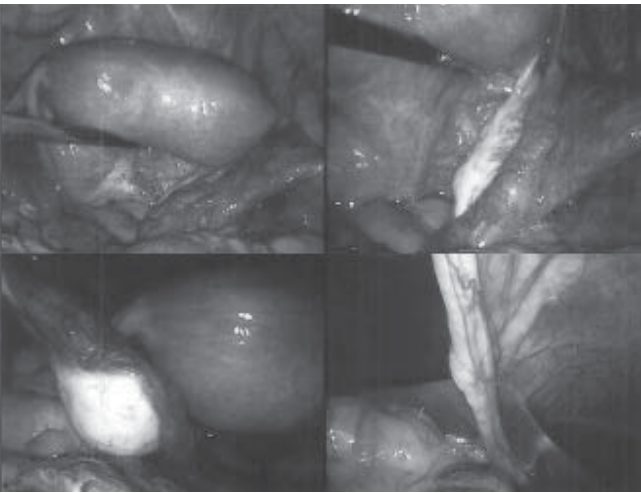


Fig. 1. Bicornuate uterus as seen laparoscopically during surgery.

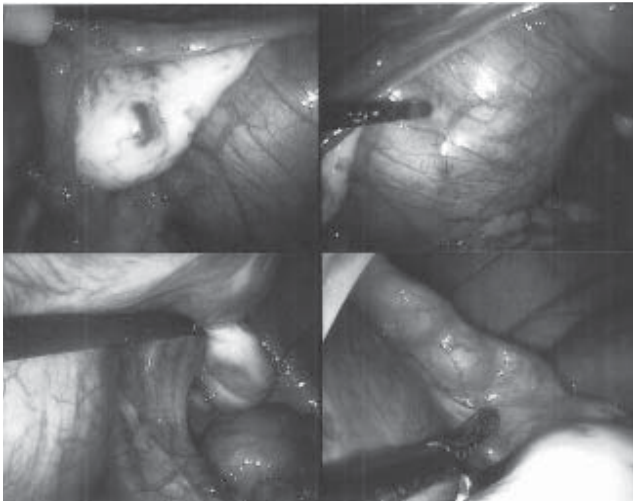


Fig. 2. Obstructed hemivagina as seen laparoscopically during surgery.

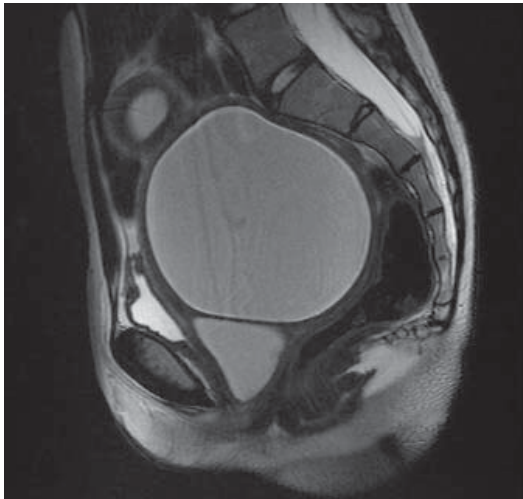


Fig. 3. MRI of left haematometocolpos.

The other issue is the discordance between ultrasound findings and operative findings. In Patient 1, besides bicornuate uterus and possible haematocolpos, there was nothing else reported on the ultrasound. Laparoscopically, there was slight peritubal adhesions which were resected. In Patient 2, the tubes were reported to be normal but laparoscopically, there was mild right haematosalpinx. We advocate the addition of routine laparoscopy as part of the surgical procedure because it allows the diagnosis of Mullerian abnormalities and treatment of conditions such as endometriosis and pelvic adhesions. The presence of hydrosalpinx on ultrasound would strongly support the inclusion of laparoscopy. It also allows maximising the fertility potential with measures such as tubal surgery and endometriotic ablation (possibly from retrograde menses).

One interesting case that we have in our series is Patient 1, a 22-year-old female who presented past 14, which is the usual age of presentation. She presented with decreased menstrual flow. She presented late because she had a button-hole defect in the vaginal septum which allowed limited flow of menses and hence no overt symptoms. The limitation faced during surgery was that there was no obvious bulge on hysteroscopy. We used the button hole⁵ that was extruding pus-like material as a guide for resection. We made a 4-cm incision originating from the button hole. Hysteroscopy allowed us a magnified vision to identify the button hole which the naked eye would have missed. On resection during surgery, there was pus in the hemivagina which was probably the result of an ascending infection.

Conclusion

In view of current evidence, hysteroscopic resection should be the modality of choice. We advocate the use of hysteroscopic resection because it allows a magnification of

visual field for surgical procedure and better identification of the anatomy with allowing the preservation of hymen. We advocate laparoscopy because it allows the confirmation of the diagnosis, allows the diagnosis and treatment of conditions e.g. endometriosis not seen on ultrasound, allows the maximisation of reproductive potential e.g. neosalpingostomy (since this will not unblock on its own, and early intervention prevents chronic damage to tubes) and preservation of the hymen.

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