

Mini Review

Undescended Ovaries: A Clinical Review

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Abstract. Undescended ovaries are frequently seen in conjunction with uterine malformations and are typically found during the course of an infertility evaluation. Other important clinical signs may prompt evaluation in an adolescent patient, though this may also be an incidental finding. An understanding of embryologic features is critical for management. It is additionally important to explain to patients and families that this is a normal developmental variant of the ovary that does not require removal in the absence of concerning pathology.

Background and Epidemiology

Undescended ovaries and fallopian tubes occur infrequently. They are typically characterized by attachment above the common iliac vessels, leading to their maldescent. Ovarian maldescent has a reported prevalence of 0.3–2%. Undescended ovaries and fallopian tubes are more commonly seen in conjunction with uterine anomalies with up to 20% occurring in patients with Mayer-Rokitansky-Kuster-Hauser syndrome and as high as 42% in cases of unicornuate uterus.¹ Several classification systems have been developed to describe complex urogenital anomalies; however, neither the Vagina Cervix Uterus Adnex-Associated Malformation system² nor the Acien modified embryological classification scheme³ address disorders of ovarian descent. Pubmed and Medline searches on key words “undescended ovary” and “ectopic ovary” revealed a total of 26 cases. The age of diagnosis ranged from 13 to 36 with an average age of 26. At least 15 of these 26 cases (73%) were associated with Mullerian anomalies. Adolescent cases comprised 1.2% of cases. Maldescent was much more common in

patients with unicornuate uteri compared to bicornuate uteri, with 63% of cases with Mullerian anomalies associated with unicornuate variants. Among the reported cases, 35% were found to be right sided, 31% were left sided, and 23% exhibited bilateral maldescent. Finally, the prevalence of associated renal anomalies was 23% in this identified group (see Table 1).

Embryology

A review of embryologic development is important for a complete understanding of this disorder. The gonad initially develops as a proliferated set of cells located on the medial surface of the urogenital ridge during the 5th week of development. The urogenital ridge is located on the posterior wall of the peritoneal cavity during development. The gonad then migrates to its “true” pelvic position due to chemotactic mechanisms. Ultimately, a normally positioned ovary lies below the pelvic brim or in the “true” pelvis, between the utero-ovarian and infundibulopelvic ligaments.^{4–7} Typically, descent of the ovary is guided by the gubernaculum, a cord of mesenchyme connected to the lower pole of the gonad. The gubernaculum then attaches to the uterus and eventually becomes the utero-ovarian and round ligaments. In contrast, the suspensory ligament attaches to the upper pole of the ovary and becomes the infundibulopelvic ligament as development progresses. The mechanism underlying maldescent of the ovary is unclear, but it has been suggested that it may be due to lack of caudal descent or growth restriction of a specific portion of the genital ridge. Other explanations for maldescent are also possible. For example, mullerian anomalies are typically thought to be a multifactorial process leading to a developmental insult at 6–9 weeks. A similar process may occur at 12 weeks leading to ovarian maldescent.^{8–12}

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Table 1. Case Review

Source	Cases	Age	Ectopic Pregnancy (Yes/No)	Infertility (Yes/No)	Mullerian Anomalies	Renal Anomalies (Yes/No)	Side Affected	Means of Diagnosis
Seoud et al ¹⁵	1	28	Yes	No	None	No	Left	Exploratory Laparotomy
McCullough et al ¹¹	1	27	No	Yes	None	No	Right	Laparoscopy
Nichols et al ¹⁰	1	36	No	No	None	Yes (ipsilateral renal agenesis)	Right	Exploratory Laparotomy
Ombelet et al ⁹	1	not listed	No	Yes	Unicornuate	No	Right	Laparoscopy, Hysteroscopy, MRI
Kives et al ⁸	1	13	No	No	Bicornuate	No	Bilateral	CT, Laparoscopy
Ombelet et al ¹⁷	1	not listed	No	Yes	Unicornuate	No	Left	HSG, MRI
	2	26	No	Yes	Unicornuate	No	Left	Hysteroscopy, Laparoscopy, HSG
	3	not listed	No	Yes	Unicornuate	Yes (Ipsilateral renal agenesis)	left	HSG, Laparoscopy, MRI
	4	not listed	No	Yes	Unicornuate	Yes (Ipsilateral renal agenesis)	Right	HSG, Laparoscopy, MRI
	5	not listed	No	Yes	Unicornuate	No	Bilateral	MRI
	6	not listed	No	Yes	Unicornuate	No	none	MRI
	7	not listed	No	Yes	Unicornuate	No	none	MRI
	8	not listed	No	Yes	Unicornuate	No	none	MRI
Trinidad et al ¹³	1	20	No	No	None	No	Right	MRI
	2	14	No	No	Vaginal Agnesis	Yes (Ipsilateral renal agenesis)	Left	MRI
Rock et al ¹²	1	not listed	No	Yes	Septate uterus with cervical duplication	No	Right	Unknown
	2	not listed	No	Yes	Unicornuate	Malrotation	Left	Unknown
	3	not listed	No	Yes	Unicornuate	Ipsilateral renal agenesis	Left	Unknown
	4	not listed	No	No	Cervical agnesis	No	Bilateral	Unknown
Gorgen et al ¹⁶	1	19	No	Yes	Bicornuate	No	Bilateral	Laparoscopy
Van Voorhis et al ¹⁴	1	35	No	Yes	Bicornuate	No	Bilateral	Laparoscopy, Ultrasound
Verkauf et al ¹	1	27	No	Yes	Bicornuate	No	Bilateral	Laparoscopy
	2	31	No	Yes	None	No	Left	Laparoscopy
	3	33	No	Yes	Unicornuate	No	Right	Laparoscopy
	4	32	No	Yes	None	No	Right	Laparoscopy
	5	35	No	Yes	None	No	Right	Laparoscopy

Diagnosis

Diagnosis of undescended ovaries is often difficult. Because an ovary is not in the typical location, it may easily be mischaracterized. Differential diagnosis for abdominal pain occurring above the “true” pelvis should include the following: appendicitis, cholelithiasis, cholecystitis, pyelonephritis, nephrolithiasis,

inflammatory bowel disease, pancreatitis, tumors, or gastroenteritis. Clearly this type of abdominal pain could result in a rather extensive evaluation. Key features of the history that may compel a clinician to suspect undescended ovary include cyclic abdominal pain above the “true” pelvis, dysmenorrhea, a history of infertility, or history of uterine or renal malformations. Of the cases reviewed from our recent Medline



Fig. 1. Mayer-Rokitansky-Kuster-Hauser syndrome. Coronal T2-weighted MT image of the pelvis of a 16-year-old girl demonstrating incomplete descent of the ovaries (large arrows). No uterine or vaginal tissue was identified. Black asterisk, urinary bladder, short arrows, femoral heads. Published with permission from: Mitchell CS, Goske MJ, Applegate K: Imaging of mullerian anomalies. In: Congenital Malformations of the Female Genital Tract: Diagnosis and Management. Edited by G Gidwani, T Falcone Philadelphia, Lippincott Williams & Wilkins, 1999, Figure 8, p 64.

and Pubmed searches, the majority (54%) were diagnosed surgically, though radiologic studies suggested the diagnosis in most cases prior to surgical intervention. Specificity and sensitivity of various radiological tests have not been reported previously; however, MRI was the most frequently used modality in the available literature reviewed. MRI is traditionally thought of as the most sensitive imaging modality for evaluation of Mullerian anomalies.¹³ MRI data have therefore been extrapolated to include disorders of gonadal descent from a level above the pelvic brim to a level beyond the inguinal canal.

Infertility Concerns

Our present literature search revealed that 77% of cases were diagnosed at the time of an infertility evaluation. The remaining 23% of individuals presented during the adolescent years or had notable incidental findings in adulthood. Many reproductive endocrinologists may suspect a diagnosis on the basis of hysterosalpingogram studies that demonstrate elongated fallopian tubes or an inability to locate an ovary on pelvic ultrasound.¹⁴ Other providers may not suspect this developmental variant because it is exceedingly

rare. If this diagnosis is not suspected, a biopsy may be performed to confirm the presence of ovarian tissue. There is no need to remove an undescended ovary in the presence of benign appearing tissue and absence of malignant concerns. Once a diagnosis is made, patients and/or parents should be fully educated about this ovarian variant. For example, there has been no association between premature ovarian failure, torsion, polycystic ovaries, endometriosis or other gynecological conditions with maldescent reported in the literature to date. Patients should understand that it is still possible to become pregnant spontaneously; however, in vitro fertilization techniques may be required for couples as well. There have been at least 10 cases reported with successful stimulation of undescended ovaries with either clomiphene or gonadotropins, followed by successful retrieval of oocytes for IVF. Ultimately, eight successful pregnancies resulted, with term deliveries occurring in three individuals. It is also important to remember that ectopic pregnancies are common in this population, with a prevalence of 4% in this reviewed group.¹⁵ Finally, because many patients have associated Mullerian anomalies, miscarriage is much more common in this population.^{16,17} The importance of a complete fertility evaluation among these individuals becomes paramount for successful pregnancy.

Conclusions

Undescended ovaries are uncommon among the general population, but may be more frequently seen among individuals with infertility or in the presence of developmental anomalies of the uterus or renal collecting system. Accurate diagnosis remains important to avoid unnecessary removal of a normal ovary. Parents and patients should additionally be counseled about potential risks for future fertility.

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