

# Unilateral ovarian hypoplasia—A report of two cases

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## ABSTRACT

**Unilateral ovarian hypoplasia is a rare event with unknown true incidence. Our knowledge about this developmental anomaly is limited and based primarily on case reports. Ovarian hypoplasia is usually asymptomatic; it is most commonly diagnosed from laparoscopy for other indications. Here we report two cases of unilateral ovarian hypoplasia in patients who presented to the Continuum Reproductive Center with primary infertility. After initial infertility workup, both patients underwent diagnostic laparoscopy for suspected tubal disease. In each case, operative findings were consistent with unilateral ovarian hypoplasia. Following the surgery, one patient underwent *in vitro* fertilization and achieved an ongoing pregnancy. The other conceived spontaneously, but was diagnosed with an ectopic pregnancy.**

**Keywords:** Ovarian Hypoplasia; Infertility

## 1. INTRODUCTION

Unilateral ovarian hypoplasia is a rare event. This developmental abnormality is usually asymptomatic and is most commonly diagnosed from laparoscopy for other indications. One theory regarding the etiology of congenital ovarian hypoplasia is the loss of an ovary as a result of ischemia secondary to torsion of an ovarian pedicle during fetal or early postnatal life; a second theory is adnexal hypoplasia resulting from developmental agenesis or aplasia [1-6]. In general, developmental abnormalities of the female genital tract include malformations of the ovaries, fallopian tubes, uterus, cervix, vagina, or external genitalia. Mullerian anomalies, estimated to be present in 3% - 5% of women [7], represent the majority of female genital tract malformations and include uterine didelphys as well as bicornuate, septate and unicornuate uteri. Occasionally, Mullerian-tract abnormalities coexist with gonadal developmental disorders [8-12].

Several authors have reported congenital ovarian agenesis in the setting of normal or abnormal fallopian tube

development [1-3,13-18]. However, the number of reports is small and knowledge about these developmental anomalies is limited. There are few data in the literature on unilateral ovarian hypoplasia [12,15].

We report two cases of ovarian hypoplasia. Both cases were diagnosed with laparoscopy for suspected tubal disease in patients with primary infertility.

## 2. CASE REPORTS

### 2.1. Case 1

A 33-year-old African-American nulligravida presented to the Continuum Reproductive Center after trying to conceive for ten months (See **Table 1**, Case 1 for summary data). She had regular menstrual periods with mild dysmenorrhea, hypertension well-controlled on alpha methyl dopa, and mild intermittent asthma. She had no significant family history of infertility, recurrent miscarriages or anomalies. The patient's partner was a healthy 35-year-old African-American man with a twelve-year-old child from a prior relationship.

On physical examination, the patient had normal external female genitalia, a normal cervix, and a retroverted uterus. The right ovary was palpated and normal in size but the left ovary could not be palpated. A transvaginal pelvic sonogram revealed a normal uterus and right ovary, and a right hydrosalpinx measuring 27 mm × 7 mm. The left ovary could not be visualized by transvaginal or transabdominal sonography. A hysterosalpingogram revealed a normal uterine cavity, a right hydrosalpinx and bilaterally-occluded fallopian tubes. Laboratory tests revealed a serum anti-mullerian hormone level of 5.1 ng/ml and follicle-stimulating hormone and estradiol levels of 5.7 mIU/ml and 55 pg/ml, respectively. A semen analysis was within normal limits by WHO criteria [19]. The patient underwent operative laparoscopy and findings included a right hydrosalpinx 20 mm × 10 mm, endometriotic implants along the left uterosacral ligament, and an infantile left ovary and fallopian tube that together measured 10 mm × 10 mm. Bilateral fallopian-tube occlusion was confirmed. A right salpingectomy and fulguration of endometriosis were performed. Pathological examination confirmed a right hydrosalpinx with

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chronic inflammation.

Two months after surgery the patient underwent in vitro fertilization using luteal-phase Leuprolide Acetate, follitropin and recombinant human chorionic gonadotropin. Twenty-two oocytes were retrieved and inseminated, 18 fertilized normally, and two high-grade blastocysts were transferred to the uterus on post-retrieval day five. A single viable intrauterine pregnancy was achieved and is currently ongoing.

## 2.2. Case 2

A 35-year-old nulligravid Indian woman presented with primary infertility of three years' duration (See **Table 1**, Case 2, for summary data). She had regular menstrual cycles and her gynecological, medical and surgical history was unremarkable. Her partner's medical and urological history was unremarkable and a semen analysis was normal by WHO criteria [19]. While under the care of a prior physician, the patient had a hysterosalpingogram which revealed a normal uterine cavity, patency of the right fallopian tube, and minimal patency of the left fallopian tube. The couple had completed six cycles of Clomiphene Citrate, with the addition of intrauterine inseminations in two cycles. No pregnancy was achieved, despite documented ovulation in all treatment cycles.

The patient's physical examination revealed normal external genitalia, an anteverted uterus of normal size, and a normal cervix. A saline hysterosonogram revealed a normal-appearing uterine cavity and a 5-mm × 5-mm endometrial polyp. Transvaginal and transabdominal

sonography failed to visualize a left ovary. Laboratory tests revealed a serum anti-mullerian hormone level of 5.5 ng/ml and follicle-stimulating hormone and estradiol levels of 5.9 mIU/ml and 45 pg/ml, respectively. Laparoscopy revealed a normal uterus, right ovary and right fallopian tube. The left fallopian tube was normal and the left ovary was atrophic (<10 mm maximum diameter). The left ovary was adherent to the left pelvic sidewall. Filmy adhesions were lysed between the left cornual region and the left anterolateral sidewall. Bilateral tubal patency was revealed by chromopertubation. A small endometrial polyp was excised by hysteroscopy. Two months following laparoscopy, the patient was diagnosed with a right ectopic pregnancy that was treated effectively with intramuscular methotrexate.

## 3. DISCUSSION

We present two cases of unilateral ovarian hypoplasia, one with concomitant ipsilateral fallopian-tube hypoplasia and the other with a normal-appearing ipsilateral fallopian tube. In both patients, diagnosis was made with laparoscopy. The incidence of unilateral ovarian hypoplasia is unknown. Published reports include cases of adnexal agenesis with the absence of the ovary and/or fallopian tube discovered at the time of laparoscopy [1,3,9,13-15, 20]. The incidence of ovarian agenesis has been suggested to be one case in 11,241 women (0.0089%) as reported by Sivanesaratnam in 1985 [3]. This estimate is based on two cases of unilateral absence of the ovary and fallopian tube diagnosed at one center.

**Table 1.** Data summary on 2 cases with unilateral ovarian hypoplasia.

	Case 1	Case 2
<b>Age</b>	33	35
<b>Presenting symptom</b>	Trying to conceive for 10 months.	3 years infertility.
<b>Physical examination</b>	Normal, except that left ovary not palpated.	Normal.
<b>Ultrasound</b>	Normal uterus and right ovary, right hydrosalpinx (27 mm × 7 mm), left ovary not visualized.	Normal except for failure to visualize left ovary. Saline hysterosonogram revealed normal uterine cavity with 5-mm × 5-mm endometrial polyp.
<b>Laboratory tests</b>		
AMH, ng/ml	5.1	5.5
E <sub>2</sub> , pg/ml	55	45
FSH, mIU/ml	5.7	5.9
<b>Laparoscopic findings</b>	Right hydrosalpinx (20 mm × 10 mm); endometriotic implants along the left uterosacral ligament; infantile left ovary and fallopian tube, together measuring 10 mm × 10 mm; bilateral fallopian-tube occlusion.	Normal left fallopian tube; atrophic left ovary, with maximum diameter <10 mm, and adherence to the left pelvic sidewall; bilateral tubal patency on chromopertubation.
<b>Reproductive outcome after laparoscopy</b>	GnRH agonist IVF-ET resulted in singleton viable intrauterine pregnancy.	Spontaneous pregnancy resulted in right ectopic pregnancy treated with methotrexate.

The suggested etiologies for ovarian agenesis or hypoplasia include torsion of the ovarian pedicle in the fetus or early neonate resulting in ischemia and atresia [4-6, 16,21]. Ovarian hypoplasia has also been suggested to result from a failure in embryologic development [1-3]. Ovarian torsion in adults is often associated with abdominal pain, nausea and vomiting. Surgical detorsion often allows for the preservation of the affected ovary. If torsion occurs during fetal life, the absence of reported symptoms might result in ovarian injury and hypogenesis. In adults, adnexal torsion might result from a variety of factors including ovarian enlargement by cysts or tumors, trauma, or by venous congestion or pregnancy. An inflammatory process in the pelvis or previous surgery and consequent adhesions might also predispose to torsion. Women with mullerian-duct anomalies might be more prone to ovarian torsion due to abnormal anatomic connections between the ovary and the pelvic sidewall [9]. Ovarian torsion might lead to organ auto-amputation [5, 15]. Sebastian *et al* reported a case in which calcified ovarian tissue was found within free-floating structures in the pelvis during laparoscopy [5]. Uckuyu *et al.* described parasitic ovaries attached to the omentum; this situation likely resulted from ovarian torsion followed by amputation and re-implantation [15].

Embryological theories on the etiology of unilateral adnexal agenesis and hypoplasia point to embryologic defects in the development of the genital ridge and the caudal end of the mullerian (paramesonephric) duct [15, 22]. Failure in the canalization of the mullerian ducts is believed to result in failed fallopian-tube development, while unicornuate uterus is a putative result of failed mullerian-duct fusion [3,23]. It has also been postulated that inadequate blood flow to the caudal portion of the mullerian duct can cause gonadal agenesis by an aberrancy in autocrine and paracrine signaling [13]. Compromised vascular flow to the caudal portion of a paramesonephric duct during its embryologic descent might also result in abnormal tubal development [1].

Ipsilateral fallopian tube anomalies range from underdeveloped fallopian tubes to complete tubal agenesis [1, 13]. Haydardeoglu *et al.* reported on a patient with left ovarian and renal agenesis and a unicornuate uterus [8]. It was presumed in that case that the most likely etiology was an insult to the unilateral urogenital ridge. Both Mulayim *et al.* and Demir *et al.* reported cases in which a patient presented with a unicornuate uterus and unilateral ovarian agenesis [10,11]. Nigam *et al.* reported a patient with a septate uterus, hypoplastic left adnexa, cervical duplication and longitudinal vaginal septum [12]. In our second reported case herein, pelvic adhesions and a hypoplastic ovary were found on the same side of the pelvis. It is conceivable that the patient underwent torsion of an ovarian pedicle early in fetal life or during very early

childhood. Absence of other congenital reproductive organ or renal anomalies makes the hypothesis of an embryological anomaly in either of our reported cases less likely.

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