Pediatric Ovarian Torsion

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INTRODUCTION

Adnexal torsion is a surgical emergency, and is reported to be the fifth most common gynecologic emergency with a prevalence of 2.7% and incidence of 4.9 per 100,000 in women younger than 20 years.1–4 It is more likely to occur in women of reproductive age, but can be seen at any age.5,6 Adnexal torsion in the pediatric and adolescent population accounts for approximately 15% of all cases of torsion.7 Although adnexal torsion can occur at any age in children (infants to 18 years), up to 52% of torsion cases in children occur between the ages of 9 and 14 years of age, with a median age of 11 years. Neonatal ovarian torsion is rare, with only 16% of cases occurring in girls younger than 1 year.7,8

Disclosure: None.

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http://dx.doi.org/10.1016/j.suc.2016.08.008
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ANATOMY

The adnexal structures include bilateral ovaries and fallopian tubes. These structures are contained within multiple folds of the peritoneum called the broad ligament, making them relatively mobile structures. The mesometrium, mesovarium, and mesosalpinx together make up the broad ligament. The ovary has a dual blood supply from the ovarian artery coursing through the suspensory ligament, which courses through the peritoneum attaching laterally to the ovary, and the ovarian branch of the uterine artery within the cardinal ligament. These 2 arteries anastomose at the lateral margins of the uterus. The ovarian ligament attaches the uterus to the ovary medially (Fig. 1).

Isolated ovarian torsion is twisting of the ovary alone by twisting on the mesovarium. Adnexal torsion involves twisting of all the adnexal components (fallopian tube and ovary) causing impairment of blood flow (Fig. 2). Adnexal torsion is more common than isolated ovarian torsion, being present in up to 67% of torsion cases. Isolated tubal torsion is defined as tubal torsion in the absence of torsion of the ovary, and is rare with an incidence of only 1.0 per 1.5 million. Torsion is more common on the right because of the hypermobility of the cecum and ileum and the slightly longer mesosalpinx and utero-ovarian ligament on the right, allowing more mobility of the adnexa.

PATHOPHYSIOLOGY

Up to 25% of pediatric patients with adnexal torsion may have normal ovaries. This increased propensity for adnexal torsion in this population may be due to the small size of the uterus in pediatric patients and the relatively long utero-ovarian ligaments leading to excess mobility of the adnexa. More commonly, 51% to 84% of pediatric adnexal torsion cases occur due to adnexal pathology, including cystic teratomas or dermoids (31%), follicular or hemorrhagic ovarian cysts (23%-33%), and,
less frequently, paraovarian/paratubal cysts, cystadenomas, or hydrosalpinx. The risk of adnexal torsion increases when the mass is benign and the size is 5 cm or larger. Malignancies are often fixed to adjacent tissues and thus are less likely to be associated with torsion. Polycystic ovarian syndrome can result in enlarged ovaries leading to abdominal pain; however, the increased weight and size of the ovaries also can be a risk factor for torsion. Congenital anomalies, such as agenesis, hypoplasia, or other abnormal development of müllerian structures, can lead to clinical examination findings consistent with torsion; therefore, it is important to corroborate the ultrasound and other imaging results with physical examination findings to ensure an appropriate diagnosis.

Torsion of the ovarian pedicle first compressed venous blood flow, followed by arterial flow because the walls of arteries are thicker and more resistant to compression. The impairment in venous blood flow causes ovarian edema and enlargement of the ovary. If torsion persists, arterial blood flow is then affected due to increased pressure within the ovary, leading to ovarian ischemia and necrosis. Complications such as pelvic thrombophlebitis, hemorrhage, infection, and peritonitis can then occur, although this is extremely rare and may be more of a theoretic concern.

**CLINICAL PRESENTATION**

Physical examination findings in patients with adnexal torsion are typically nonspecific and can include normal temperature or a low-grade fever and/or mild tachycardia. The most common symptom of adnexal torsion is the acute onset of pelvic or abdominal pain. The pain can be variable in nature, including nonradiating, constant, or intermittent (depending on whether the torsion is partial or complete); mild or intense; and of variable duration (days to months), but is often isolated to one side. The patient also
may voice a history of transient episodes of similar pain indicating previous events of partial, intermittent torsion. Other commonly associated symptoms include nausea and vomiting due to peritoneal reflexes, flank pain, and anorexia. Vaginal bleeding and bowel or bladder abnormalities are other rare associated symptoms. Pelvic examinations are generally not performed in this patient population unless the patient has a specific vaginal complaint or is sexually active. Also, it is not necessary to perform a pelvic examination to make the diagnosis. Clinical symptoms in neonates can be difficult to interpret due to the lack of specific symptoms and limitations in assessing pain; however, they may present with an abdominal mass and/or feeding intolerance.

There are no laboratory tests that have been proven to establish a diagnosis of adnexal torsion; however, a pregnancy test, complete blood cell count, and electrolyte values are helpful in clinical assessment and differential diagnosis. A complete blood count is useful to assess white blood cell count as a sign of inflammatory reaction or infection and a urine pregnancy test to rule out pregnancy or ectopic pregnancy. Most laboratory findings are normal in patients with adnexal torsion; however, a slight leukocytosis sometimes can occur. Adnexal torsion must be differentiated from other diagnoses, including appendicitis, kidney stone, gastroenteritis, hemorrhagic ovarian cyst rupture, pelvic inflammatory disease, and ectopic pregnancy. If a complex ovarian mass (septations and/or solid components) is present on imaging, serum of tumor markers, including HCG (human chorionic gonadotropin), AFP (alpha-fetoprotein), CA125 (cancer antigen 125), and LDH (lactate dehydrogenase) can be useful in assessing malignancy risk.

DIAGNOSTIC PROCEDURES

The diagnosis of adnexal torsion is often difficult due to the vague and variable clinical presentation as well as nonspecific imaging findings. The most commonly used and accurate imaging study used to assist in the diagnosis of adnexal torsion in pediatric and adolescent patients is pelvic ultrasonography with color Doppler to evaluate blood flow to the ovaries. The abdominal approach for pelvic ultrasound is most often used in the pediatric and adolescent population compared with the transvaginal approach used in the adult population. Ultrasound findings in adnexal torsion can include a unilaterally enlarged ovary or asymmetric ovarian enlargement, heterogeneous appearance of one ovary due to edema, the presence of a simple or complex adnexal mass, present or diminished/absent flow on color Doppler, peripherally displaced follicles due to stromal edema from ischemia, medialization of the ovary, displacement of the uterus from the midline, free pelvic fluid, and the whirlpool sign, defined as twisting of the ovarian pedicle causing twisting of vessels (Figs. 3–6). The most frequently observed adnexal lesions found during torsion include ovarian cystic teratomas, follicular or hemorrhagic cyst, paraovarian/paratubal cysts, cystadenoma, and hydrosalpinx. A torsed fallopian tube can appear dilated and edematous. The risk of adnexal torsion also increases when the adnexal mass is 5 cm or larger; however, children can experience adnexal torsion with completely normal size ovaries as well.

Unfortunately, torsion cannot be absolutely confirmed or excluded based on the presence or absence of Doppler flow on ultrasound. Although the absence of vascular flow is highly suspicious for torsion, the sensitivity of absent arterial flow is as low as 40% to 73%; however, venous compression is evident in up to 93% of torsion cases. There are cases of torsion in which completely obstructed vascular flow is not present on ultrasound Doppler as well (Fig. 7). Computed tomography and
Fig. 3. Ovarian torsion with edema and peripheral displacement of the ovarian follicles. The image demonstrates an enlarged left ovary with scattered small peripheral follicles and a paraovarian cyst. The presence of peripheral follicles confirms that the structure is ovarian in origin. Unilateral ovarian enlargement with peripheral displacement of the follicles suggests torsion. (From Ngo AV, Otjen JP, Parisi MT, et al. Pediatric ovarian torsion: a pictorial review. Pediatr Radiol 2015;45:1849; with permission.)

Fig. 4. Massively enlarged, featureless torsed ovary in a 4-year-old girl with abdominal pain for 1 week. Ultrasound image demonstrates a heterogeneous, hyperechoic avascular mass in the midabdomen. No peripheral follicles are present to aid in the identification of ovarian tissue. This is an example of a case in which a torsed ovary (arrow) became massively enlarged and edematous/necrotic, leading to loss of the normal imaging features of an ovary and making it difficult to distinguish ovarian torsion from an adnexal mass. (From Ngo AV, Otjen JP, Parisi MT, et al. Pediatric ovarian torsion: a pictorial review. Pediatr Radiol 2015;45:1853; with permission.)
MRI are not first-line imaging modalities, but can be used to further delineate anatomy, if ultrasound is not available, or if other pathologies remain in the differential diagnosis, such as appendicitis. MRI is the gold standard imaging modality for müllerian anomalies.18

Fig. 5. (A) Ultrasound image of a 6-year-old with left ovarian torsion. The left ovary is larger than the right ovary. Note peripheral follicles in the left ovary from ovarian edema. (B) Power Doppler image of the left ovary with no intraovarian flow identified. (From Ngo AV, Otjen JP, Parisi MT, et al. Pediatric ovarian torsion: a pictorial review. Pediatr Radiol 2015;45:1848; with permission.)

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Fig. 6. Ultrasound whirlpool sign in ovarian torsion. (A, B) Color flow on Doppler ultrasound images demonstrates the twisted pedicle (arrows) in a 12-year-old girl with a large, mature cystic teratoma (T) arising from the left adnexa, representing the lead point for left adnexal torsion. (From Ngo AV, Otjen JP, Parisi MT, et al. Pediatric ovarian torsion: a pictorial review. Pediatr Radiol 2015;45:1849; with permission.)
DIAGNOSIS AND TREATMENT

Adnexal torsion is a clinical diagnosis. If clinical features, patient history, and/or imaging suggest a high suspicion for adnexal torsion, the final diagnosis is made by immediate exploratory surgery. Prompt diagnosis and operation are beneficial so as to prevent irreversible adnexal damage, salvaging the torsed adnexa, and maximizing the success of ovarian conservation. Pain lasting more than 10 hours is associated with an increased rate of tissue necrosis. Laparoscopic surgery is considered the best diagnostic and therapeutic approach in the pediatric population (Fig. 8).

Fig. 7. Flow variation in 2 girls with surgically confirmed ovarian torsion. (A) Color Doppler ultrasound image demonstrates an enlarged right ovary measuring $9.7 \times 9.9 \times 4.8$ cm for a total volume of 242 mL. No color or spectral flow is identified. (B) Color Doppler image with both arterial and venous flows are present in the enlarged, torsed right ovary, which measures $5.17 \times 4.6 \times 3.2$ cm for a total volume of 39.2 mL. (From Ngo AV, Otjen JP, Parisi MT, et al. Pediatric ovarian torsion: a pictorial review. Pediatr Radiol 2015;45:1849; with permission.)

Fig. 8. Left adnexal torsion visualized during diagnostic laparoscopy on a young patient who presented with acute-onset severe abdominal pain and was found to have a left ovarian cyst.
Traditionally, surgeons performed oophorectomy in children and adolescents if a torsed ovary appeared necrotic.\textsuperscript{30–32} There were also concerns that the inflammatory effect from the necrotic ovary could lead to adhesive disease and subsequent bowel obstruction, along with increased risk of venous thrombosis once the ovary was detorsed.\textsuperscript{30,31,33} Evidence has shown that even necrotic-appearing ovaries, which are black-blue in color, appear to improve after detorsing, showing signs of recovery (Fig. 9). In addition, follow-up ultrasound postoperatively demonstrates normal Doppler flow, and follicular development after only 6 weeks.\textsuperscript{21,22,34–36} Another commonly cited reason for oophorectomy has been due to concern for malignancy. Fortunately, ovarian malignancy in children also is extremely rare; thus, the probability of occurrence with torsion is rare. It is still important to identify malignancy risk ahead of time. Studies have reinforced that oophorectomy should be reserved for grossly abnormal ovaries and/or elevated tumor markers including HCG, AFP, and CA125 drawn preoperatively.\textsuperscript{4,15} Recent literature supports conservative management, including detorsion, ovarian or paraovarian/paratubal cystectomy to reduce cyst recurrence (not simple drainage of the cyst), and preservation of the adnexa (ovary and fallopian tube) even if necrotic, as a safe and effective approach to the management of adnexal torsion.\textsuperscript{16,30,33,34,37–40} Even though evidence reinforces conservative management, oophorectomy is still performed in a significant number of cases, with national data showing rates unchanged from 2000 (61%) to 2006 (58%).\textsuperscript{41}

Ovarian or paraovarian/paratubal cystectomies as opposed to simple cyst drainage should be performed for all cysts confirmed as nonfunctional or physiologic types (hemorrhagic or follicular cyst).\textsuperscript{42,43} Ovarian or paraovarian/paratubal cystectomy if indicated can be safely performed using a bipolar device such as the Harmonic Scalpel or a monopolar device to incise the ovarian capsule or mesosalpinx of the paraovarian/paratubal cyst, and with blunt and sharp dissection, the cyst can be easily removed from the cyst bed. Occasionally the cyst bed needs to be coagulated with electrocautery to provide hemostasis.

Fig. 9. Enlarged congested left ovary after detorsion. (From Ngo AV, Otjen JP, Parisi MT, et al. Pediatric ovarian torsion: a pictorial review. Pediatr Radiol 2015;45:1848; with permission.)
A study by Styer and Laufer also advocated for ovarian bivalving after detorsion cases of severely hemorrhagic and edematous adnexa. During this procedure, they describe using a linear incision along the antimesenteric aspect of the affected ovary after untwisting. This method serves to confirm viable tissue within hemorrhagic, ischemic areas, and releases the increased pressure of the edematous ovarian capsule to facilitate lymphatic and venous drainage and allow for arterial flow, thus reducing ischemia of the ovary after detorsion.

Oophoropexy after detorsing the adnexa is a controversial surgical technique that can be used to limit ovarian mobility and prevent retorsion. Several techniques for this procedure have been described, including fixation of the ovary to the peritoneum on the pelvic sidewall, uterosacral ligaments, or round ligament, shortening the utero-ovarian ligament via plication, and suturing the ovary to the back of the uterus. This is not a procedure that is routinely done for every case of adnexal torsion because there is no widely agreed consensus on the topic and removal of adnexal masses that could have precipitated the torsion usually prevent retorsion. Use of this procedure can be considered in management of recurrent torsion of an affected ovary to decrease the likelihood of subsequent retorsion or can be performed if only one ovary remains due to prior oophorectomy. The only reported disadvantage of oophoropexy is the possibility of anatomic distortion between the ovary and fallopian tube and the possibility of reduced fertility due to this anatomic stability change to prevent retorsion. However, this type of fertility reduction has been described only in the lateralizing type of oophoropexy (eg, fixation to the pelvic sidewall), and thus medial oophoropexy (eg, utero-ovarian ligament plication) may be a better choice. Overall, the efficacy and safety of this ovarian fixation procedure are not well established.

CLINICAL OUTCOMES

There is variability in the literature about the late effects of conservative surgical management versus more radical techniques as treatment for ovarian torsion. In the pediatric population, menstrual cycles are used to assess ovarian function. Some studies have suggested women who undergo conservative procedures show an increase in menstrual irregularity and painful menses. Certain studies suggest that removal of one ovary does not significantly worsen female fertility, whereas others state that resection of the affected ovary can have a negative impact on future fertility. Prior studies evaluating long-term follow-up of patients with ovarian torsion managed conservatively showed rates of normal-sized ovaries and follicular function as high as 91% to 98%. These studies reinforce that most detorsed ovaries recover function and show follicular development after only 6 weeks even when they appear necrotic intraoperatively. Despite the contradictory literature, studies overall agree that conservative management of adnexal torsion via laparoscopy is the preferred management and will preserve ovarian function in pediatric patients, allowing for subsequent normal progression through puberty and future fertility.

Surveillance ultrasounds can be performed to monitor for cyst recurrence and ovarian integrity 3 months after a procedure for adnexal torsion and/or cystectomy and then every 6 months to 1 year for long-term surveillance. Menarchal patients with history of functional ovarian cysts (follicular or corpus luteal, which develop from the follicle containing the egg during menstrual cycle) can be placed on hormonal therapy to reduce ovulation and recurrence of ovarian cysts, thereby reducing the risk of subsequent adnexal torsion. Hormonal therapy does not reduce recurrence of para-ovarian/paratubal cysts or dermoid ovarian cysts because these are not formed as a
consequence of ovulation. Therapeutic options for functional ovarian cysts include combined hormonal contraception (estrogen and progesterone) methods, such as oral contraceptive pills, patch, or vaginal ring or progesterone-only options including depo medroxyprogesterone acetate or etonogestrel implants, all of which suppress ovulation and subsequent functional cyst formation.60,61

SUMMARY

In summary, adnexal torsion is a surgical emergency and the fifth most common gynecologic emergency presenting with variable examination and radiologic findings. Early diagnosis and surgical intervention with diagnostic laparoscopy and conservative management with ovarian preservation are important for maintenance of ovarian function and preservation of future fertility in girls and adolescents.

REFERENCES


