OVARIAN CYSTS AND TORSION

Epidemiology:
- <5% incidence in girls from birth to age 8 years old
- 10-12% incidence in adolescents, most of which spontaneously disappear
- ovarian torsion diagnosed in up to 2.7% of females with acute abdominal pain

The easiest way to contextualize ovarian cysts in the pediatric population is by age group:

<table>
<thead>
<tr>
<th>Fetus</th>
<th>Neonate</th>
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<tbody>
<tr>
<td>- Follicular cyst</td>
<td>- Physiologic cyst (maternal hormonal stimulation in-utero)</td>
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<tr>
<td>Infants/Pre-pubertal</td>
<td>Adolescent</td>
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<tr>
<td>- Simple ovarian (enlarged cystic follicle)</td>
<td>- Simple/complex ovarian cysts</td>
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<tr>
<td>- Sex/Cord Stromal</td>
<td>- Follicular cyst, hemorrhagic</td>
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<tr>
<td>- Granulosa cell tumor</td>
<td>- Corpus luteum cyst</td>
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<td>- Sertoli/Leydig cell tumor</td>
<td>- Luteoma of pregnancy</td>
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<td>- Germ Cell tumor</td>
<td>- Chocolate cyst of endometriosis</td>
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<tr>
<td>- Endodermal sinus tumor</td>
<td>- Theca-lutein cyst</td>
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<tr>
<td>- Dysgerminoma</td>
<td>- Excessive HCG state such as with molar pregnancy</td>
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<tr>
<td>- Chorionicarcinoma</td>
<td>- Cystic teratoma</td>
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**McCune Albright Syndrome presentation: recurrent follicular cysts
- Classic triad of precocious puberty, “café – au – lait” skin spots, and polyostotic fibrous dysplasia

Ovarian Torsion – almost always associated with a cyst
- Defined as the abnormal twisting of the involved ovary on its ligamentous support
- Results in venous congestion, hemorrhage, and eventually ischemia
- Prolonged ischemia leads to necrosis, death of ovary, and possible infection or peritonitis

*Note: Embryologically, ovary migrates from T10 to true pelvis by puberty.*
Therefore, early in life it is an abdominal organ and more susceptible to torsion.

History/Physical Examination
Ovarian cysts can often be **asymptomatic**

*Signs of precocious puberty:*
- vaginal bleeding, axillary/pubic hair, breast development (advanced Tanner stage for age)

*Menstrual and Sexual history (post-pubertal girls)*
- last menstrual period, regularity, flow, pain
- onset of symptoms in relation to period

*Somatic signs: chronic abdominal aching pain*
- **Tumor:** increasing abdominal girth, early satiety, nausea/vomiting
**Torsion** – pelvic pain, nausea, fever

**Differential Diagnosis**

<table>
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<tr>
<th>Younger Age</th>
<th>Older Age</th>
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<tbody>
<tr>
<td>- GU: Wilms tumor, urinary tract obstruction, urachal cyst</td>
<td>ECTOPIC PREGNANCY</td>
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<td>- GI: mesenteric/omental cyst, volvulus, colonic atresia, intestinal duplication, Meckel’s diverticulum</td>
<td>Tubo-ovarian Abscess</td>
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<tr>
<td>- Choledochal/splenic/pancreatic cyst</td>
<td>(below DDx for <strong>ovarian torsion</strong>)</td>
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<tr>
<td>- lymphangioma</td>
<td>Appendicitis, small bowel obstruction</td>
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<tr>
<td></td>
<td>Pyelonephritis</td>
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<td></td>
<td>Pelvic inflammatory disease</td>
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<td>Gastroenteritis</td>
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**Evaluation**

Abdominal exam – difficult to detect ovarian masses. Tenderness, rebound/guarding when present are helpful

Pelvic exam- most reliable exam for ovarian mass, but often not tolerated in pediatric population

**Pelvic Ultrasound** – to assess for whether cyst is simple (clear, fluid-filled) or complex (septa, debris)

- Size
  - Simple cysts < 2 cm in diameter are considered physiologic
- Character
  - Anechogenic and fluid-filled? -> favors functional cyst
  - Thick-walled? Calcifications? Septations? -> may indicate malignancy
- Free fluid in pelvis? Hematoperitoneum?
- Addition of Doppler
  - Not always conclusive, but Doppler flow can be compromised in ovarian torsion

**Serum Markers**

- AFP (endodermal sinus tumor)
- LDH (dysgerminoma)
- hCG (molar pregnancy, choriocarcinoma)
- Estrogen (Granulosa cell tumor)
- Testosterone (Sertoli-Leydig Cell)
- CA-125 (epithelial ovarian cancers, less likely to be seen in pediatrics)

**CT/MRI** only if ultrasound is equivocal or malignancy is strongly suspected

**Management/Treatment**

**Fetus** -> expectant management since both simple and complex cysts will likely spontaneously regress

- Serial ultrasounds: antenatally every 3-4 weeks
Neonate

- Expectant management is one option: serial ultrasounds at birth, q4-6 weeks thereafter
- **Aspiration**: shown to have low risk of adverse effects for cysts > 4-5 cm
- Surgery: if complex, symptomatic, increasing in size, or persists for > 4-6 months

Infants/Pre-pubertal

- Absence of complex features (septation or calcification) can be observed, follow-up ultrasound
- Otherwise surgery, especially in times of torsion

Adolescents

- Observation, NSAIDs for pain, **oral contraceptive pills for < 6 cm**
- >6 cm, laparoscopic cystectomy may be warranted

**TORSION**

- First ensure the patient is hemodynamically stable
  - Resuscitate with fluids, blood transfusion if bleeding intraperitoneally
- Surgery **immediately** to salvage torsed ovary
  - **Oophoropexy**: performed in children **without evidence of an ovarian mass**.
    - Involves either shortening the utero-ovarian ligament
    - or suturing the ovary to the utero-sacral ligament

**Caption**: Image of ovarian torsion. Ovarian vessels are twisted 3 times.
http://www.bonnmd.com/bonnmd/Ovarian_Torsion_files/torsion1_thumb.jpg
An Overview of Polycystic Ovary Syndrome (PCOS)

PATHOPHYSIOLOGY
Excess intra-ovarian androgen production caused by likely a combination of multiple proposed theories:
1) Abnormal pituitary function (elevated LH/FSH ratio)
2) Primary ovarian dysfunction
3) Problem of adrenal steroidogenesis
4) Insulin resistance
   a. Can be in the context of the metabolic syndrome

CLINICAL DIAGNOSTIC CRITERIA
The Rotterdam criteria is validated and utilized in the diagnosis of adults, but not in adolescents
The NIH criteria for diagnosis of adolescents with PCOS requires both:
1) Hyperandrogenism confirmed by biochemical testing
2) Abnormal menstrual pattern

CLINICAL FEATURES
- Hirsutism: sexual hair that appears in a male pattern
  - Hirsutism equivalents: acne vulgaris, pattern alopecia, seborrhea, hyperhidrosis, hidradenitis suppuritiva
- Anovulation: primary amenorrhea, oligomenorrhea, irregular bleeding
- Polycystic ovaries: usually diagnosed by ultrasound
- Obesity and Insulin resistance
  - Large waist circumference, acanthosis nigricans
  - Abnormal labs: serum triglycerides, HDL, glucose, blood pressure

RECOMMENDED DIAGNOSTICS TESTS
- Serum free testosterone
  - Prolactin, IGF-1, TSH, cortisol, 17-hydroxyprogesterone
    - To rule out other causes of hyperandrogenism
- Pelvic ultrasonography

TREATMENT
1) Combination oral contraceptive pills to regulate menstrual cycles
2) Spironolactone: safest and most potent anti-androgen therapy
3) Obesity/Insulin resistance
   a. Diet and exercise for goal of weight reduction
   b. Metformin favored in adolescents to combat insulin resistance
References