Key: = important notes
= reference to torsion
= reference to malignancy

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:

Fetus	Neonate
- **Follicular cyst**	- **Physiologic cyst (maternal hormonal stimulation in-utero)**
Infants/Pre-pubertal	**Adolescent**
- **Simple ovarian (enlarged cystic follicle)** - **Sex/Cord Stromal** **-Granulosa cell tumor** **-Sertoli/Leydig cell tumor** - **Germ Cell tumor** **-endodermal sinus tumor** **-dysgerminoma** **-choriocarcinoma** - **Cystic teratoma**	- **Simple/complex ovarian cysts** - **Follicular cyst, hemorrhagic** - **Corpus luteum cyst** - **Luteoma of pregnancy** - **Chocolate cyst of endometriosis** - **Theca-lutein cyst** **-excessive HCG state such as with molar pregnancy** - **Cystic teratoma**

**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 by 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

Younger Age	Older Age
- **GU: Wilm's tumor, urinary tract obstruction, urachal cyst** - **GI: mesenteric/omental cyst, volvulus, colonic atresia, intestinal duplication, Meckel's diverticulum** - **Choledochal/splenic/pancreatic cyst** - **lymphangioma**	**ECTOPIC PREGNANCY** **Tubo-ovarian Abscess** **(below DDx for ovarian torsion)** **Appendicitis, small bowel obstruction** **Pyelonephritis** **Pelvic inflammatory disease** **Gastroenteritis**

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 ->

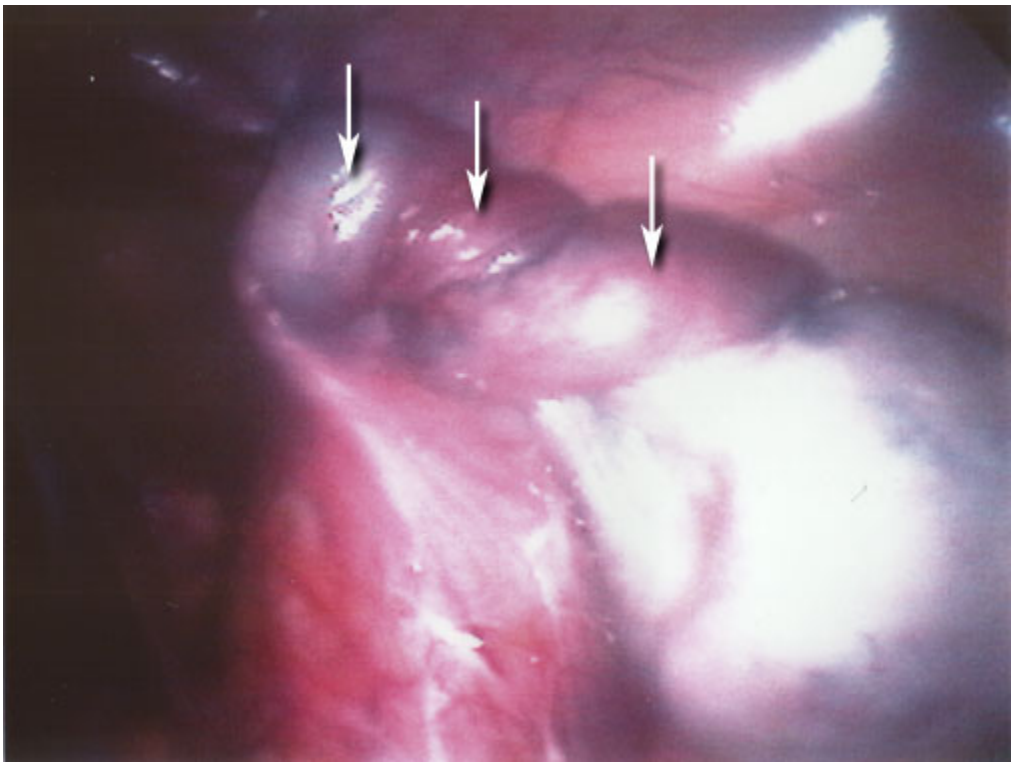
- Absence of complex features (septation or calcification) can be observed, follow-up ultrasound
- Otherwise surgery, especially in times of torsion

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

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