OBJECTIVES

• Describe the normal anatomical and physiologic development of the ovary during fetal life, infancy, childhood, and adolescence.
• Discuss the etiology, diagnosis, and management of ovarian cysts at different ages.
• Discuss the diagnosis and management of ovarian torsion.

FETAL AND NEONATAL OVARIAN CYSTS
INCIDENCE

- Ovarian cysts estimated to be present in 34% of fetuses
- Increasing in detection by TVS prevalence
- Diagnosis most often made in 3rd trimester
- Symptomatic cysts in 1/2500 live births

ETIOLOGY

- 2nd to excessive maternal hormone stimulation from hCG
- FSH from the fetal pituitary beginning at 20 weeks gestation increases the number and size of follicles
- Probable pathology is disordered folliculogenesis
- After birth, E2 and hCG decrease rapidly
- FSH declines more slowly

DIFFERENTIAL DIAGNOSIS

- Mesenteric cyst
- Enteric duplication
- GI obstruction
- GU obstruction
- Renal cyst
- Anterior meningocele
- Neoplastic masses
  - Cystadenoma
  - Cystic teratoma
  - Granulobastoma
  - Nephroblastoma
  - Hemangioma
  - Lymphangioma
- Complex cyst 31 weeks, 6 cm

MANAGEMENT: Antenatal

- Conservative approach favored (Müller-Leisse 1992)
  - Spontaneous resolution during neonatal period in most cases (Bagolan 2002)
  - Low risk of antenatal complications
    - Pulmonary hypoplasia, labor dystocia, polyhydramnios
    - cyst rupture, torsion

Luzzato et al: 27 patients with antenatal ovarian cyst
  * 5 operated in newborn period
  22 followed
  - 10 simple all regressed
    10 followed
  - bilateral ovaries
  - 12 complex
    - 8 unilateral
    - 2 bilateral
    - ovary ovary

Pediatric Surgery Int. 2000; 16:35-39
2 wks later, appears cystic 4 cm

2 wks later, 1.5 cm

MANAGEMENT:
Antenatal Aspiration

- Theoretical advantages:
  - May prevent torsion and auto-amputation
  - Fluid analysis
  - May prevent laparotomy
- Concerns regarding accurate diagnosis
- Neonatal treatment preferred

NEONATAL CYST

NEONATAL OVARY

1-2 million oocytes
1 cm diameter
Primordial follicles
Elevated circulating FSH levels

NEONATAL CYSTS

- Simple ovarian cysts are follicular in origin
- Complex ovarian masses may represent in-utero or neonatal torsion or hemorrhage
- Malignancy with cystic lesions extremely rare < 2 yo
  - One reported case
  (Doeling, 2000)
NEONATAL CYST
Differential Diagnosis

1. GU (uterine anomaly, renal cyst, urologic obstruction, mega ureter, urachal cyst)
2. GI (meconium, mesenteric or pancreatic cyst, bowel obstruction)
3. Pelvic tumors (presacral teratoma, neuroblastoma, menigocele, retroperitoneal tumor)

NEONATAL OUTCOME

- Both simple and complex cysts undergo regression within the first few months ~75%
  - Proportional to cyst size


- Complications:
  - Torsion
  - Hemorrhage
  - Mass effect

COMPLICATIONS

- Torsion:
  - Risk ∝ size cyst
  - Approximately 25% if > 5 cm
  - Neonatal torsion may be asymptomatic or associated with pain, N, V, Fever

- Hemorrhagic Cyst
  - Due to torsion and associated ischemia

NON-OVARIAN COMPLICATIONS

- Intestinal obstruction from visceral compression
- Thoracic compression (pulmonary hypoplasia)
- GU obstruction

NEONATAL OVARIAN CYST

Asymptomatic

Diagnosis Ovarian Cyst
Complex or Simple

Resolution

Recurrence

Recurrence

Development of Symptoms

Surgery

Strickland 2002
• Fetal and neonatal ovarian cysts are common, usually physiologic
• Spontaneous resolution in ~ 75% cases within 6 months of life
• Conservative management advocated
• Neonatal percutaneous aspiration of large cysts may prevent torsion in rare cases
• If surgery is indicated, aim for the most conservative treatment

THE OVARY IN CHILDHOOD
• Follicular development to antral stage
• Follicular atresia
• Low FSH, low estradiol
• Stromal growth, progressive ovarian enlargement

Ovarian cysts in childhood

Millar et al
• 1818 prepubertal ultrasounds
• 2-5% - unilocular cysts < 1 cm
• Cysts > 2 cm were rare
• 5% large cysts associated with pseudopuberty
• Ovarian cysts < 5 cm - follow conservatively

OVARIAN CYSTS IN CHILDHOOD
• Retrospective analysis of management of 51 children with ovarian cysts > 5cm
  – 28 resolved spontaneously
  – 23 required surgery:
    • 6 Teratomas
    • 2 Cystadenomas
    • 1 Granulosa cell
    • 1 Sertoli-Leydig cell
• < 5.5cm - all resolved
OVARIAN CYSTS IN CHILDREN

Reasons for concern:
- > 5 cm
- solid components
- septations, complex internal echoes
- endocrine symptoms or signs
- pelvic pain

OVARIAN CYSTS IN CHILDHOOD

Considerations:
- Hormonally active - pseudo puberty
- McCune – Albright Syndrome
- Precocious Puberty
- Thyroid Disease
- Congenital paraovarian structure
- Germ Cell Tumors / Malignancy
- Torsion

OVARIAN TUMORS IN CHILDREN

- Germ Cell
  - Mature Teratoma
  - Immature Teratoma
  - Endodermal sinus
  - Embryonal
  - Dysgerminoma
- Epithelial
  - Serous
  - Mucinous

- Stromal/Sex chord
  - Granulosa cell
  - Thecoma
  - Sertoli-Leydig
  - Fibroma

OVARIAN TUMORS IN CHILDHOOD

<table>
<thead>
<tr>
<th>Ovarian cyst</th>
<th>Benign tumors</th>
<th>Malignant tumors</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>57.9%</td>
<td>30%</td>
<td>7.9%</td>
<td>4.3%</td>
</tr>
<tr>
<td>Corpus luteum</td>
<td>Teratoma</td>
<td>Dysgerminoma</td>
<td></td>
</tr>
<tr>
<td>Follicular</td>
<td>Cystadenoma</td>
<td>Teratoma</td>
<td></td>
</tr>
<tr>
<td>Paratubal</td>
<td></td>
<td>Sertoli-Leydig</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Undifferentiated</td>
<td></td>
</tr>
</tbody>
</table>

OVARIAN TUMOR MARKERS

- CA-125
  - epithelial tumors
- LDH
  - dysgerminoma
- AFP
  - Endodermal sinus, embryonal carcinoma, immature teratoma, mixed germ cell
- Estradiol
  - thecomas, dysgerminoma
- Testosterone
  - fibroma, Sertoli-Leydig
- HCG
  - choriocarcinoma, Germ cell, embryonal cell
EVALUATION OF A PREPUBERTAL OVARIAN CYST

- CBC
- Thyroid screening
- Imaging
  - Ultrasound with doppler flow
  - CT scan
  - MRI
- Tumor markers
  - AFP, HCG, LDH, Inhibin A,B
- Hormonal markers
  - FSH, LH, Estradiol, Testosterone

BENIGN CYSTIC TERATOMA

DERMOID CYST

- Most common germ cell and benign tumor of childhood
- Bilateral 0-9% in children
- Incidence of malignancy decreases with age
- Has distinct X-ray and ultrasound appearance
- Conservative surgery is indicated
- Surgical approach remains controversial
THE OVARY IN ADOLESCENCE

- 300,000 germ cells
- Follicular growth
- Progressive increase in estrogen, progesterone, LH, FSH production with increased ovulatory cycles
- Frequent ovulatory defects

OVARIAN CYST BEYOND PUBERTY

- Most occur from failure of ovulation or involution (functional)
- Increased incidence in adolescence
- May be associated with:
  - Menstrual irregularities
  - Pelvic discomfort and pain
  - Rupture and hemorrhage
  - Torsion
- simple or complex
Cysts that aren’t “cysts”
– 1 - 2 cm ovarian follicles which are often called “simple cysts
– A normal physiologic occurrence

Hemorrhagic corpus luteum
develops in luteal phase
Rupture of this cyst can rarely cause hemoperitoneum
• Other cysts can also rupture: dermoid, cystadenoma, endometrioma

HEMORRHAGIC CYST

OVARIAN CYST: SYMPTOMS
• PAIN with:
  – Torsion
  – Rapidly expanding
  – Leaks or ruptures
• Ruptured corpus luteum can have the an acute presentation with sudden onset of pain with or without orthostatic symptoms

SYMPTOMATIC OVARIAN CYST: DX and RX
• Dx: CBC, βHCG, U/S
• Rule out other emergencies:
  – Appendicitis
  – Ovarian torsion
  – Ectopic pregnancy
• Rx:
  – Observation, analgesic (NSAID +/- narcotics)
  – Laparoscopy for persistent pain and ↓Hct / Hg
  – Role of OCP (to prevent new cysts)
**TABLE 4. Differential Diagnosis of Adnexal Masses in Adolescent Girls**

<table>
<thead>
<tr>
<th>Organ</th>
<th>Differential Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ovary</td>
<td>Physiologic cyst, follicular cyst, simple cyst, corpus luteum, tubo-ovarian abscess, endometriosis, dermoid, benign, malignant</td>
</tr>
<tr>
<td>Fallopian tube</td>
<td>Ectopic pregnancy, tubo-ovarian abscess, complex hydatid cyst, hemorrhagic cyst, lipoma, dermoid, molar pregnancy, endometriosis, interstitial, uterine anatomy</td>
</tr>
<tr>
<td>Broad ligament</td>
<td>Hydrosalpinx, hemoperitoneum, hemorrhagic cyst, dermoid, malignant, benign</td>
</tr>
<tr>
<td>Uterus</td>
<td>Uterine fibroid, endometriosis, leiomyoma, leiomyosarcoma, lipoma, endometriosis, mesentericodem, retropertioneal lipoma</td>
</tr>
<tr>
<td>Miscellaneous neoplasms</td>
<td>Sertoli-Leydig tumor, Brenner tumor, choriocarcinoma, dermoid, neuroblastoma, germ cell tumor</td>
</tr>
</tbody>
</table>

**MANAGEMENT OF ADNEXAL MASSES IN ADOLESCENTS**

**Simple Cyst**
- Repeat ultrasound 4-8 weeks
- Resolution:停手 surgery, monophasic OC, observation
- Growth: persistence, partial resolution
- Persistence: no change, stop observation

**Complex Cyst**
- Exclude pregnancy related diagnosis
- Repeat ultrasound 4-8 weeks
- Resolution: surgery, partial resolution
- No change: stop, observation

**INDICATIONS FOR IMMEDIATE SURGICAL INTERVENTION**

- Suspicion of torsion
- Solid mass
- Hemodynamic instability
- Refractory pain
- High suspicion of malignancy

**Surgical techniques available**

- Aspiration (diagnostic cytology only)
- Fenestration and biopsy or enucleation (hemorrhagic cysts, endometriomas)
- Cystectomy (preferred method)
- Oophorectomy
TORSION ADNEXA
• Can involve ovary, fallopian tube, paratubal cyst
• Result in ischemia and rapid onset acute pelvic pain
• Right side may be more commonly
• Dermoid cyst most commonly involved
• May occur in normal ovary, especially in younger girls (7-10)

TORSION ADNEXA: SX
• Severe pain
• Constant (complete torsion) or intermittent (partial torsion)
• Onset coincide with lifting, exercise or coitus
• Autonomic reflex (N, V)

TORSION ADNEXA: SIGNS
• Localized tenderness
• +/- acute abdominal signs
• Pelvic mass
• Mild fever and leukocytosis

TORSION ADNEXA: DX
• Real time Ultrasound:
  Complex mass with echogenic rim
• Color doppler ultrasound
  Absent arterial/venous flow
  low velocity peripheral flow
  central venous flow without arterial ovarian enlargement
• CT/MRI
• Direct visualization

Diagnosis of torsion
Correlation of sonographic findings
• Real time sonography
  – Complex mass 73%
  – Cystic mass 20%
  – Cul-de-sac fluid 87%
• Doppler abnormalities 91%
  – No A/V flow 40%; decreased 13%
  – Decreased venous/absent arterial flow 33%
  – Decreased arterial/absent venous flow 7%
  Russell et al J Ultra Med 2001
TORSION ADNEXA: DDX

- Acute appendicitis
- Pregnancy related complications
- Rupture of ovarian cyst
- Ovulation
- Urinary colic
- Intestinal rupture/obstruction

CONSERVATIVE SURGICAL INTERVENTION

- Laparoscopic confirmation of diagnosis
- Identify point of torsion
- Detorse adnexa & conserve ovary
- If cyst or neoplasm present - manage appropriately
- +/- Oophoropexy
Surgical Management: Adnexal Torsion (cont’d)

- Resumption ovarian function 87 - 95%
- No evidence that detorsion ↑ risk of pulmonary embolism (*McGovern et al., 1999)*
- Aspiration or cystectomy if necessary

IN SUMMARY: the main points

- The ovary is an endocrine and reproductive organ that is active at all ages
- Ovarian cysts < 5 cm can be seen at all stages of development; most regress without treatment
- Ovarian Cysts are common; increasing with age
- Ultrasound is the mainstay of ovarian diagnosis
- Torsion should be diagnosed urgently; managed conservatively