Pediatric and Adolescent Gynecology

- Normal sexual differentiation
- Abnormalities of the external genitalia
- Normal internal genitalia
- Abnormalities of the internal genitalia
- Tips on the adolescent and pediatric exam
- Normal hormonal changes neonatal to completion of puberty
NORMAL SEXUAL DIFFERENTIATION

**XX**
- Primordial Gonad [4 wks]
- Ovary [8-9 wks]
- Development of Mullerian System
  - internal genitalia
  - not under hormonal control

**XY**
- Testis [6-7 wks]
- Leydig cells
- Testosterone
- local testosterone
- Wolffian Duct development
- Placental HGC
- 5-α reductase
- DHT
- androgen receptor
- masculization of external genitalia

Expression of SRY
External Genitalia

• Hormone dependent, complete 18-20 wks
• Absence of androgens
  – genital tubercle=clitoris
  – labio-scrotal folds=labia majora
  – urethral folds=labia minora
  – urogenital sinus=lower 1/3 vagina (endoderm)
  – sinovaginal bulb+urogenital sinus=hymen
Abnormal External Genitalia
Female

• Primarily a result of abnormal androgen exposure
• The degree and timing of androgen exposure can lead to varying degrees of virulization ranging from clitoral hypertrophy to complete virulization
  – eg. CAH, maternal androgen, androgen secreting tumours, placental aromatase deficiency
Abnormal External Genitalia

Male

• Complete or partial insensitivity to androgens
• Most common are androgen insensitivity and 5 alpha reductase deficiency
ANDROGEN INSENSITIVITY

• Complete: Bilateral testes, females external genitalia, short blind vagina, no Mullerian structures, 50% inguinal hernias,

• Orchiectomy post puberty, risk of malignancy very low prior to 25 years

• Incomplete: Orchiectomy as above or early if ambiguous genitalia or virulization in early puberty
5 alpha REDUCTASE DEFICIENCY

- No mullerian structures
- Ambiguous or female external genitalia at birth (DHT needed in utero)
- Puberty, virulization as normal male can occur with testosterone alone
- Dx: increase T:DHT ratio after HCG stimulation
NORMAL MULLERIAN DEVELOPMENT

- Upper 1/3 mullerian duct not fused = fallopian tubes
- Lower 2/3 fused = uterus, cervix, upper 2/3 vagina (mesoderm)
Mullerian Anomalies

• Modified AFS Classification
• 4 groups based on embryologic considerations
Class I: Segmental Mullerian Agenesis or Hypoplasia
Class II: Unicornuate Uterus with/without Rudimentary Horn

A-1-a. Communicating

A-1-b. Non-communicating

A-2. No cavity

B. No horn
Class III: Uterine Didelphys
Class IV: Bicornuate Uterus
Class V: Septate Uterus

A. Complete

B. Partial
Class VI: DES Anomalies

A. Constriction bands

B. T-shaped

C. Widening of lower two-thirds of uterine cavity
ASSOCIATED ANOMALIES

- Urologic: 30%
- Skeletal: 12%
- Auditory: 33%, sensorineural, high frequency
PRESENTATION

• Neonatal:
  – No vaginal opening, vaginal dimple
  – mucocele, distended uterus
  – result of maternal estrogen stimulation

• Childhood:
  – unlikely to present
PRESENTATION

• Amenorrhea, Dysmenorrhea, Menorrhagia
• Difficulty with tampons
• Dysparunia
• Infertility, recurrent abortion
• Poor obstetrical outcomes
• Incidental
IMPERFORATE HYMEN

• Most common anomaly, <=1%
• DDX: labial agglutination, vaginal agenesis, low transverse septum
• If mucocele in newborn period, need to excise, if not, do in the 2 year window between breast budding and menarche
• Cruciate incisions followed by excision and reapproximation, pre&post premarin cream
EXAMINATION AND HORMONE STATUS

NEONATE TO COMPLETION OF PUBERTY
## EXAMINATION

<table>
<thead>
<tr>
<th></th>
<th>INFANT</th>
<th>CHILD</th>
<th>ADOLESCENT</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hymen</strong></td>
<td>thickened, pale</td>
<td>translucent</td>
<td>redundant, thick</td>
</tr>
<tr>
<td><strong>Labia</strong></td>
<td>prominent, thick</td>
<td>small, flat</td>
<td>opposed, prominent</td>
</tr>
<tr>
<td><strong>Vagina</strong></td>
<td>acidic, rugated 5cm</td>
<td>neutral, thin 8cm</td>
<td>acidic, rugated 10-12cm</td>
</tr>
<tr>
<td><strong>Flora</strong></td>
<td>lactobacillus</td>
<td>mixed, pH↑</td>
<td>lactobacillus</td>
</tr>
<tr>
<td><strong>Clitoris</strong></td>
<td>≤0.6 cm²</td>
<td>&lt;0.6 cm²</td>
<td>2-4 mm wide</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>10 mm virulization</td>
</tr>
</tbody>
</table>
EXAMINATION

• Put child at ease, allow her control

• Positions:
  – Frog-legged: supine or in mother’s lap sitting or lithotomy
  – Knee chest (prone)

• Hand-held mirror

• Magnifier
Table 12-1. Suggestions for Genital Examinations of Young and Adolescent Girls

Obtain the patient’s cooperation.
This is necessary to create teachable moments and to afford adequate examinations.

Get the patient to comply.
She must have control over the examination.
She must be promised no physical pain.
She must know what information is being sought.

Special measures in the prepubertal female’s examination:
Inform the child that the genital examination is sanctioned.
Involve the child with a hand-held mirror and a magnification device.
Position the child with her feet in the stirrups, on the examiner’s lap, or sitting on a parent’s lap. Drape her legs over the parent’s thighs.
Attempt to examine in both the supine and knee-chest positions.
Be knowledgeable about the various spread methods to reveal the vestibule.
Use instruments only when necessary.

Special aspects of the peripubertal genital examination:
Understand the impact of estrogen.
Take advantage of the stimulus phenomenon.
Choose a speculum of proper width after the introitus is evaluated.
EXAMINATION

- Index finger on labia minora bilaterally
- Light lateral downward pressure
- Suckling, deep breath or cough allows vestibular tissue to fan out
- Recall ovaries are abdominal
- Be sure all swabs are lubricated
EXAMINATION

• Flexible hysteroscope or pediatric cystoscope best (irrigate to distend and flush), vaginoscope, nasal speculum, test tube

• Catheter-in-a-catheter: attach 1cc tuberculin syringe to proximal 6.5 cm of a butterfly tubing placed in distal 6.5 cm of a soft, sized 12 bladder catheter
HYMEN: Variations, alterations and configurations

- Many variations on the normal hymen
- Describe the configuration
- Key Points:
  - Contour most variable in the suburethral area
  - Majority have a smooth free margin from 3 to 9 o’clock with no irregularities, mounds or indentations
HYMEN: Variations, alterations and configurations

- Interpretation of bumps and breaks of the hymen controversial
- Most common at 6 o’clock
- 3x more likely to be sexual abuse than other pediatric gyne conditions eg. Remnant of septate hymen, inflammatory granulation tissue, HPV
NEONATAL

• Estrogen hormonal influence remains for approximately one month
  – high as a result of placental conversion of fetal and maternal C19 steroids to estrogens
• Vaginal discharge normal, may be slightly bloody for the first 10 days of life (10%)
• Enlargement of breast tissue, may secrete colostrum or milk
NEONATAL

- Palpate for gonads, inguinal hernias
- Identify vaginal opening
- Lubricated pediatric feeding tube for probe
- Clitoris, especially in preterm infants, may appear disproportionally enlarged, observe for other evidence of androgen excess
NEONATAL

- Hymenal tags are not uncommon
  - 1-3 mm isolated exophytic mucosal lesions
  - Vast majority resolve
- Periurethral cysts
  - Often confused with small hydromucocolpos and bulging imperforate hymen
  - Resolve over the first month of life
  - Treat only if infection or voiding concerns
NEONATAL

- Ovaries may contain follicular cysts:
  - 5x5 cm average size at diagnosis
  - >99% functional and benign
  - conservative management: serial U/S
  - 80% resolve within 4 months
  - Warn re: risk of torsion
  - If >4 cm consider aspiration (can do prenatally)
NEONATAL

- Hormone profile:
  - High estradiol, low FSH/LH
  - As estradiol levels fall FSH/LH levels increase similar to those in the adult life
  - First one to two years, FSH/LH levels intermittently rise and fall
CHILDHOOD

• Little or no hormonal stimulation
• Genitalia growth proportional to general growth
• Small follicular cysts common
• FSH and LH begin to fall age 3
• Resetting of “gonadostat”, low levels of estradiol inhibit GnRH, FSH, LH
PUBERTY

- Tanner staging and growth curves
- Growth spurt
- Early secondary sexual characteristics
- Menarche
- Completion of secondary sexual characteristics
- Completion of growth
HORMONE CONTROL

- Development of positive feedback mechanism of HPO axis, low levels of estradiol increase FSH and follicular recruitment begins
HORMONAL CONTROL

- Exact mechanism of activation of H-P-O axis unknown
- Nocturnal pulsatile GnRH first
- Increase in LH > FSH secretion
- Increase in FSH leads to increase estradiol
- Growth spurt, Leukorrhea, Breast budding
HORMONAL CONTROL

- Initial increase in FSH and LH not cyclic
- Estradiol levels fluctuate, only intermittently high enough to lead to LH surge i.e. ovulation
- Initial ovulatory cycles subnormal progesterone, shortened luteal phase, gradual shift to normal corpus luteal function
HORMONAL CONTROL

- First post-menarchal year, 50% of cycles ovulatory
- Investigate if ovulatory cycles not established by 3 years
ADRENARCHE

- Primarily adrenal, likely independent from thelarche and gonadarche
- DHEAS and androstenedione under control of ACTH stimulation of zona reticularis
- Control of this unknown
SUMMARY

• Initial growth spurt
• Breast bud 9.8 years
• Onset of adrenarche 10.5 years
• Peak in growth velocity 11.4 years
• Menarche 12.8 years
• Mean time interval thelarche to menarche 3 years, abnormal if greater than 4 years
SUMMARY

- Growth potential post menarche 6-8 cm
- Adult pubic hair 13.7 years
- Adult breast 14.6 years