Pediatric Gynecology/
REI
Prolog Questions
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Straddle Injury

A 7yo girl presents to ED w/ perineal injury that is actively bleeding. You are told she fell on the crossbar of her brother’s bike. History obtained from pt and her mother is inconsistent. PE shows no bruises over the skin and external genitalia w/ defect involving the fourchette and extending to perirectal area. Anal orifice is intact. What is the best next step?

- A) Observation in the ED
- B) Laparoscopy
- D) Pelvic sonogram
- C) Exam under anesthesia
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Straddle Injury

- **Answer:** Exam under anesthesia
- **EUA** is best choice b/c she is actively bleeding and EUA will allow assessment of lacerations that may need repair
- Straddle injuries can result in vulvar hematomas that can impair voiding due to urethral spasm. Must ensure she can void or will need foley.
- **Treatment of vulvar hematoma:** pyridium for dysuria, sitz baths. Large hematomas may need I&D. If pain persists → evaluate for pelvic fracture.
Straddle Injury

- Posterior vaginal tears suspicious for sexual abuse and penile penetration will cause hymenal tears between 4 and 8 o’clock

- Child protective services must be notified if any suspicion of abuse arises. Exam must be properly documented.
Vulvar disease

In a pediatric patient, for which of the following conditions would you prescribe clobetosol propionate and inquire about the possibility of sexual abuse?

- A) Vulvar adhesions
- B) Lichen sclerosus
- D) Embryonal rhabdomyosarcoma
- C) Mullerian agenesis
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Lichen sclerosus

- **Answer:** Lichen sclerosus

- Lichen sclerosus is an inflammatory skin condition in adults and children that can cause itching and soreness. 4-5% of adult cases have an associated squamous cell carcinoma.

- PE shows ivory-colored papules, plaques, fissuring, erosions and hyperkeratosis.

- Path: hypotrophic dystrophy and flattening of rete pegs in the subdermal layer, hyalinization and keratinization.

- **Tx:** 1-2% hydrocortisone cream. If this fails, clobetesol propionate bid followed by taper. Surgery indicated only if clitoris is buried, labia are fused or introitus stenosed.

- Koebner’s phenomena: when genital trauma (such as from abuse) induces lichen sclerosus secondary to friction and scarring.
Labial Adhesions

- Thought to result from local irritation and scratching
- PE shows a thin avascular line of fusion in the midline
- Usually asymptomatic but can impair urination/menstruation
- Tx: estrogen cream, zinc oxide or weak corticosteroid for 10-14 days. If this fails, can use a lubricated swab or sound to break down the membrane
Embryonal carcinoma of the vagina (Sarcoma botryoides)

- Very rare tumor, but specific to children
- Usually originates from cervix or upper vagina, arising from undifferentiated mesenchymal tissue
- PE: tumors have a grapelike appearance
- Spread rapidly, extending through subvaginal tissues
- Tx: chemo followed by surgery or irradiation
- Survival rate as high as 90%
Mullerian Agenesis

- aka Mayer-Rokitansky-Kuster-Hauser syndrome
- Failure of the upper vagina and uterus to form
- PE: blind vagina
- Tx: Progressive dilatation or surgical vaginoplasty
- All patients with mullerian anomalies should be assessed for renal anomalies as well
A sexually active 15 yo female c/o significant lower abdominal pain w/ menses, L>R. Menarche age 14.5, thelarche 16.5, growth curves WNL. PE sig for LLQ tenderness, no RB, guarding. Pelvic exam showed 1 cervix and L adnexal tenderness. Recent tests for STDs neg. What is most appropriate next step in management?

- A) Abdominal radiography
- B) Pelvic sonogram
- C) Expectant management
- D) MRI
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Mullerian Anomalies

- **Answer: Pelvic Sonogram**

- Pelvic sonogram can better define pelvic organs, including identification of a Mullerian anomaly, which should be considered in absence of STD, PID or pregnancy

- The pt in this case actually had symptoms due to a didelphic uterus
Mullerian Tract Development

- 6 wks gestation – Mullerian tracts identified
- 9 wks gestation – elongation occurs to the level of the UG sinus at Muller’s tubercle
- Uterovaginal canal then forms and inserts into UG sinus at Muller’s tubercle
- Fusion of the ducts occurs from caudal to cephalic
- 20 wks gestation – by this time canalization and resorption has occurred
Mullerian Anomalies

- Imperforate hymen: due to incomplete canalization of the urogenital sinus. Incidence 0.1%. Pts present w/ amenorrhea, cyclical pelvic pain and bluish hue to hymen. Valsalva will produce a bulge.
Mullerian Anomalies

- **Transverse Vaginal Septum:** Present w/ amenorrhea, cyclical pelvic pain. MRI can help differentiate from imperforate hymen. Valsalva will not produce a bulge. Believed to be sex-linked autosomal recessive. Associated anomalies include coarc of aorta, ASD, L-spine malformations.
Mullerian Anomalies

- **Longitudinal Vaginal Septum:** Not always symptomatic. Believed to be due to embryonic arrest of mullerian and metanephric ducts at 8 wks gestation. Often associated w/ uterus didelphys and associated renal anomaly.
Mullerian Anomalies

- **Rudimentary Horns**: Non-communicating. May be associated with pelvic pain secondary to outflow tract obstruction when there is functional endometrium. Require resection when symptomatic.
Mullerian Anomalies

- **Uterus Didelphus**: Vertical fusion defect. May be associated with a hemivagina and blind vaginal pouch. May presents with pelvic pain. Resection of the vaginal septum relieves the outflow tract obstruction.
A 17 yo HS senior is sent to ED by school nurse for malaise, decreased appetite and abdominal pain. History notable for irregular periods. She is sexually active w/ a negative hCG. PE notable for RLQ pain and rebound tenderness. What is the next appropriate step?

- A) Laparoscopy
- B) CT Abd/Pelvis
- C) GI consult
- D) Administer Azithromycin/Cipro
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Adolescent with Appendicitis

- **Answer:** CT Abd/Pelvis
- Pt is classic presentation of appendicitis and CT is most sensitive imaging modality
- Appendicitis parallels development of lymphoid system and peaks in young adulthood
  - Slightly more prevalent in males w/ male-to-female ratio 1.3:1
- Important signs/symptoms (although invariable): diffuse midepigastric pain that localized to RLQ, anorexia, obstipation, mild leukocytosis, low-grade fever
While rounding, you are asked to come delivery room to evaluate term neonate just delivered with undetermined gender. Mother is repeatedly asking if newborn is boy or girl and your associate wants your opinion. What is the most important next step?

- A) Testing serum androgens
- B) Testing serum electrolytes
- C) Karyotype determination
- D) Abdominal sonogram
- E) MRI
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Ambiguous Genitalia

- **Answer:** Testing serum electrolytes

- **All cases of ambiguous genitalia should be treated as congenital adrenal hyperplasia (CAH) until proven otherwise**

- **Must follow closely for salt-wasting (hyperkalemia, hyponatremia) and signs of adrenal insufficiency (N/V, diarrhea, dehydration, shock)**
Ambiguous Genitalia

- Most common cause of CAH causes 21-hydroxylase deficiency; 17-hydroxyprogesterone levels will be elevated
  - Next most common cause of CAH is 11-ß-hydroxylase deficiency; 11-desoxycortisol levels will be elevated
- The other choices may be useful in determining gender (karyotype being 100% accurate) but metabolic issues most important initially.
You are called to ED to evaluate 10 yo premenarchal girl w/ abdominal pain. PE limited by voluntary guarding. Abdominal sono shows bilateral solid adnexal masses each 10cm x 10cm in size. hCG level is 62 and LDH is 137. What is the most likely diagnosis?

- A) Endodermal sinus tumor
- B) Immature teratoma
- C) Dysgerminoma
- D) Theca-lutein cyst
- E) Mature cystic teratoma
Premenarchal Pelvic Mass

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Premenarchal Pelvic Mass

- **Answer:** Dysgerminoma

- Solid and solid/cystic adnexal tumors in children are usually dysgerminomas or immature teratomas
  - Dysgerminomas account for 50% of all cases
Dysgerminomas

- Consist of germ cells that have not differentiated to form embryonic or extraembryonic structures
- Dysgerminomas are bilateral in 10-15%
- Majority of patients have Stage I disease
- hCG and LDH can be elevated
- Associated w/ gonadoblastoma in a small percentage of pts
- Chromosomal analysis is key b/c Y chromosome necessitates oophorectomy
Other pelvic masses mentioned

- **Endodermal sinus** – AFP usually increased, usually unilateral
- **Immature teratomas** – usually unilateral, hCG not usually elevated
- **Theca-lutein cysts** – hCG levels will be very high; cysts are usually multicystic and bilateral
- **Mature cystic teratomas** – bilateral in 10-15%, cyst lumen contains mature elements with differentiation of tissues from all 3 germ layers
A mother brings her 6 yo daughter to ED for white discharge and vulvar erythema for past 2 wks. Child is hesitant to allow her mother to touch her vulvar area. Mother informs you that recently the child is reluctant to go near, or be touched by, an uncle who lives w/ the family. The physical finding most likely to be found this child is:

- A) Posterior hymenal tear
- B) Vulvar hemangioma
- C) Urethral caruncle
- D) Lichen sclerosus
- E) Labial agglutination
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Sexual Molestation

- **Answer:** Posterior hymenal tear

- In absence of obvious signs of trauma, physical finding most likely to be found in recently sexually abused child is a posterior hymenal tear or hymenal transection

  - A deep posterior hymenal notch or healed tear, an increase in transhymenal diameter, or a deep notch or concavity in the anterior half of the hymen are other studies that increase suspicion of sexual abuse
Acute Pelvic Pain in Young Women

An ED physician requests consultation for a non-sexually active 14 yo young woman w/ bilateral LQ pain, R>L. Pain is described as sharp and stabbing, radiating down the R leg. Pain has been intermittent over the past 24 h. She has taken 800 mg Ibuprofen w/o relief. Menarche 6 mo ago and she is currently at the end of her menstrual period. PE remarkable for decreased bowel sounds and tenderness w/ rebound in the LLQ. Bimanual exam limited by patient discomfort. Beta hcg and STD tests are negative. What is next appropriate step?

- A) Laparoscopy
- B) Spiral CT
- C) Pelvic sono
- D) MRI
- E) Laparotomy
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Acute Pelvic Pain in Young Women

- **Answer:** Pelvic Sonogram
9. Precocious Puberty

A 71/2 year-old African-American girl is referred for evaluation of precocious puberty. The appearance of pubic hair was noted at 6 years 11 months and breast budding 1 month ago. She is otherwise in excellent health and without any additional symptoms. Examinations reveals Tanner stage III pubic hair and Tanner stage II breast development. Longitudinal growth has increased from the 55th to the 60th percentile. Her growth velocity chart demonstrates that she has moved from 4-cm to 5.5-cm growth per year. The most appropriate initial management is

(A) observation only
(B) bone age X-ray of the hand only
(C) magnetic resonance imaging (MRI) of the head only
(D) adrenocorticotropic hormone (ACTH) challenge test
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Normal puberty

General order:
--Acceleration of growth velocity
--Secondary sexual characteristics (ages 9-11)
--Thelarche
--Adrenarche
--Adolescent growth spurt (growth velocity increases from 4 cm to 9cm per year)
--menstruation (mean age 12.8 years)
Precocious Puberty

- Historical definition: sexual development before age 8 years
- workup if:
  - African Americans < age 6 (and adrenarche often precedes thelarche)
  - whites < age 7
  - CNS of behavioral changes
11. Emergency Contraception

A 29-year-old woman comes to your office with a history of unprotected intercourse within the past 24 hours. She requests emergency contraception therapy. The oral steroid hormone treatment that would provide her best option for emergency contraception is

(A) ethinyl estradiol and norgestrel
(B) mestranol
(C) desogestrel
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Emergency Contraception

2 FDA approved products:

--Preven kit (combination ethinyl estradiol and levonorgestrel)

--Plan B: levonorgestrel 0.75 mg PO q 12 h x 2 doses within 72 hours of intercourse
A 25-year-old woman received a diagnosis of premature ovarian failure. Her karyotype is 46, XX. Which of the following laboratory studies is most likely to be abnormal in this patient?

(A) Fasting glucose
(B) Calcium
(C) Cortisol
(D) Thyroid-stimulating hormone (TSH)
(E) Vitamin B12
1. Premature ovarian failure

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Premature ovarian failure

- loss of ovarian function before age 40

- Causes --> idiopathic, abnormal karyotype (Turners, Fragile X), viral infections (mumps), injuries to ovaries (radiation, chemo, surgery), and autoimmune

- think about autoimmune diseases (hypothyroidism, diabetes, adrenal failure, hypoparathyroidism, pernicious anemia)…think about screening for these annually in idiopathic POF
A 43-year-old nulligravid woman requests a second opinion regarding treatment for osteoporosis. Osteoporosis has been diagnosed by dual-energy X-ray absorptiometry (DXA), and she has begun to take alendronate at her physician’s recommendation. She takes calcium supplements, 1,500 mg per day, with vitamin D, 400 IU per day. She runs approximately 16 km (10 mile) per week and bikes 80.5 km (50 mile) every weekend. On physical examination, she is 1.7 m (5 ft 8 in) tall, she weighs 57.2 (126 lb), her blood pressure level is 96/60 mm Hg, and pulse rate is 64 beats per minute. The remainder of her examination is unremarkable. Laboratory studies, including TSH, estradiol, FSH and serum prolactin levels, are normal. In addition, her electrolyte, human parathyroid hormone, serum calcium, and 24-hour urinary free calcium levels obtained while she is off calcium supplementation also are normal. To monitor her response to alendronate, the most appropriate next step in her management is

(A) repeat DXA scan
(B) urinary N-telopeptide level
(C) lateral X-ray of spine
(D) urinary calcium excretion rate
(E) ultrasonography of calcaneus
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Osteoporosis

- bone resorption by osteoclasts
- bone formation by osteoblasts
- urinary N-telopeptide to follow bone degradation (low levels indicate bone stabilization/compliance with therapy)
- DXA scan
  - T-score between -1 and -2.5 → osteopenia
  - T score <-2.5 → osteoporosis
  - treat at T score <-2, or <-1.5 with risk factors
  - tx: Ca, vit D, bisphosphonate
A 13-year-old adolescent comes in for evaluation of “lack of sexual development”. The patient’s medical history and family history are unremarkable. Vital signs are normal. Physical examination reveals a height of 142 cm (56 in), Tanner stage I breast and pubic hair development, webbed neck, high-arched palate, broad chest, small uterus, normal cervix, and nonpalpable ovaries. Bone age is 11.2 years. Laboratory analysis reveals a FSH level of 41 mIU/mL, a luteinizing hormone level of 24 mIU/mL, estradiol level of less than 10 pg/mL, and karyotype of 45,X. Appropriate initial management for this patient is

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(B) calcium
(C) recombinant human growth hormone
(D) medroxyprogesterone acetate
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Turners Facts

- XO, or mosaic 45X/46XX or 46XY
- Order: IV pyelogram/renal US, echocardiogram, hearing exam, TSH/free T4, fasting plasma glucose
- Treat: growth hormone treatment and later with estrogen therapy
A 19-year-old Hispanic woman with a history of hepatitis comes in for management of hirsutism present since puberty that has become more severe despite therapy with oral contraceptives. She has dark facial hair on the sides of her face, upper lip, and chin. She has neither acanthosis nigricans nor clitoromegaly. Her DHEAS, serum testosterone, and androstenedione levels are at the upper limits of normal. TSH and prolactin concentrations are normal. Her basal 17α-hydroxyprogesterone level is 230 ng/dL. Following administration of corticotropin, her 17α-hydroxyprogesterone level increases to 510 ng/dL. The next best step in management is to continue OCs and prescribe

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Hirsutism

- PCOS
- androgen-producing tumors
- Nonclassic CAH
PCOS

- DHEAS, testosterone, and androstenedione are within normal range/upper limit
- TREATMENT: OCPs, spironolactone, hair removal agents
Androgen producing tumors

- Testosterone > 200 ng/dL
- Hilus cell tumors of ovary, adrenal tumor, etc.
- TREATMENT: surgery
Nonclassic CAH

- 17α-hydroxyprogesterone for screening and diagnostic testing
- if random 17α-hydroxyprogesterone > 200 ng/dL (SCREENING) then do an acute adrenal stim test for DIAGNOSTIC PURPOSES

- Acute adrenal stim test:
  - give corticotropin, measure 17α-hydroxyprogesterone in 30-60 min
  - if 17α-hydroxyprogesterone > 1,000 ng/dL → CAH
Ferriman-Gallwey

- The Ferriman-Gallwey score is a method of evaluating and quantifying hirsutism.
- Hair growth is rated from 0 (no growth of terminal hair) to 4 (complete and heavy cover) in nine locations: upper lip, chin, chest, upper back, lower back, upper abdomen, lower abdomen, the upper arms and the thighs, giving a maximum score of 36.
- In Caucasian women, a score of 8 or higher is regarded as indicative of androgen excess.
- With other ethnic groups, the amount of hair expected for that race should be considered.
4. Bulimia

A 22-year-old woman who desires pregnancy presents with a 4-year history of amenorrhea. She is 1.6 m (5 ft 4 in.) tall and weighs 55.3 kg (122 lb). She lost 9.1 kg (20 lb) about the time her menses ceased. Her weight gradually increased with a total weight gain of more than 6.8 kg (15 lb) over the next 2 years, but menses did not resume. She admits to vomiting several times each week. The treatment most likely to effect resumption of menses is

(A) phenothiazine
(B) nutritional counseling
(C) selective serotonin reuptake inhibitors (SSRIs)
(D) cognitive-behavioral therapy
(E) monoamine oxidase inhibitors
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The patient in the figure presents with a 6 month history of amenorrhea, 9.1 kg (20 lb) weight gain, fatigue, and occasional headaches. Based on this patient’s presentation, the best screening test for her condition is

(A) 24-hour urinary free cortisol excretion test
(B) high-dose dexamethasone (8 mg) suppression test
(C) overnight dexamethasone (1 mg) suppression test
(D) plasma adrenocorticotropic hormone (ACTH) concentration
(E) 4:00 PM serum cortisol concentration
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19. Cushings

- Too much cortisol

**SCREENING TEST**
- low-dose dexamethasone suppression test
- 1 mg dexta at 11 PM, plasma cortisol at 8 AM
- \(<5 \mu g/dL \rightarrow normal\)
- \(\geq 10 \mu g/day \rightarrow Cushings\)

**DIAGNOSTIC TEST**
- 24-hour urinary free cortisol excretion test
- normal is \(<100 \mu g/day\)
- \(\geq 250 \mu g/day \rightarrow CUSHINGS\)
Random cortisol levels

- < 5 µg/dL → adrenal insufficiency
- 17 → normal adrenal reserve
- 5-17 → do not rule out adrenal insufficiency

**NOTE: these are random levels and not for Cushings 😊**
A 13-year-old adolescent comes in for evaluation and treatment of rapid weight gain and irregular menstrual cycles. She has a family history of obesity and diabetes mellitus in her father and endometriosis in her aunt. On physical examination, she is morbidly obese and has evidence of acanthosis nigricans. Her waist circumference is 90 cm, her fasting blood glucose level is 175 mg/dL, and her total cholesterol level is 300 mg/dL. The most common pathophysiologic basis for her disorder is

(A) hyperlipidemia  
(B) hypothalamic dysfunction  
(C) hyperandrogenemia  
(D) insulin resistance  
(E) leptin gene mutation
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Metabolic Syndrome

- insulin resistance syndrome = metabolic syndrome X

- PATHOPHYSIOLOGY - acquired insulin resistance --> a postreceptor defect causes lack of response to insulin action leading to elevated levels of insulin and subsequent metabolic abnormalities
Metabolic Syndrome

**DIAGNOSIS (3 or more criteria)**
- central obesity (waist circumference $>88$ cm)
- elevated triglyceride level
- low high-density lipoprotein (HDL)
- elevated blood pressure
- elevated fasting blood glucose level
Metabolic Syndrome

- TREATMENT: weight loss, diabetes and cardiac meds as appropriate
- if really high BMI → bariatric surgery
117. Primary amenorrhea

A 16-year-old adolescent comes in for evaluation of primary amenorrhea and lack of breast development. She denies being sexually active and has no history of vaginal bleeding. Her height is 134.6 cm (53 in.). She has Tanner stage 1 breast development and Tanner stage 1 pubic hair. She has a normal vaginal length, visible cervix, small palpable uterus, and no palpable ovaries. Initial lab evaluation reveals a negative serum pregnancy test result and normal levels of prolactin and TSH. The serum measurement that would provide the most information to make the diagnosis is

(A) FSH
(B) estradiol
(C) testosterone
(D) DHEAS
(E) 17-hydroxyprogesterone
A 16-year-old adolescent comes in for evaluation of primary amenorrhea and lack of breast development. She denies being sexually active and has no history of vaginal bleeding. Her height is 134.6 cm (53 in.). She has Tanner stage 1 breast development and Tanner stage 1 pubic hair. She has a normal vaginal length, visible cervix, small palpable uterus, and no palpable ovaries. Initial lab evaluation reveals a negative serum pregnancy test result and normal levels of prolactin and TSH. The serum measurement that would provide the most information to make the diagnosis is

(A) FSH
(B) estradiol
(C) testosterone
(D) DHEAS
(E) 17-hydroxyprogesterone
117. Primary amenorrhea

- DEFINITIONS:
  --Primary - absence of menarche by age 16
  --Secondary - absence of menses for more than 3 cycles or 6 months in women who were previously menstruation

- THINK ABOUT:
  --Dysfunction of the hypothalamus, pituitary gland, ovaries, uterus, or vagina.

- Primary amenorrhea is often genetic or anatomic abnormality
Primary amenorrhea

WORK-UP

- PHYSICAL EXAMINATION
- LABORATORY TESTING
- KARYOTYPE
Primary amenorrhea

WORK-UP

- PHYSICAL EXAMINATION
  - pubic hair? --> adrenals working, testosterone receptors present
  - breast? --> evidence of estrogen/working ovaries
  - imperforate hymen/vaginal septum?
  - no uterus? --> mullerian agenesis
  - blind vagina (palpable testes) --> androgen insensitivity syndrome (breasts, no pubic hair)
Primary amenorrhea

WORK-UP

- **LAB TESTING**
  - $\beta$HCG --> pregnant
  - FSH
    - --> hypergonadotropin hypogonadism (ovarian failure, minimal breast development, normal pubic hair)
    - --> hypogonadotropin hypogonadism (hypothalamic or pituitary problem, CT/MRI)
  - prolactin --> pituitary adenoma
  - TSH --> hypothyroidism/hyperthyroidism
Primary amenorrhea

WORK-UP

- KARYOTYPE
  - 45 X  --> Turners
  - 45 X, 46 XY  --> gonadal dysgenesis (46 XY=Swyer)
    - 20-30% gonadoblastoma, dysgerminoma rate, so take out gonads
  - 46 XY  --> androgen insensitivity
A 16-year-old adolescent with no history of sexual activity is being evaluated for primary amenorrhea. On physical examination, she has Tanner stage IV breast development. Tanner stage II pubic hair, a 2 cm vaginal pouch, no visible cervix, and no palpable uterus or ovaries. Lab analysis reveals a FSH level of 8.3 mIU/mL, testosterone level of 811 ng/dL, and a 46,XY karyotype. The next step in management for this patient is

(A) breast augmentation  
(B) estrogen therapy  
(C) vaginoplasty  
(D) psychologic counseling
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(A) breast augmentation
(B) estrogen therapy
(C) vaginoplasty
(D) psychologic counseling
Complete Androgen Insensitivity

- Phenotypic female (breasts), no pubic hair, no uterus, 46XY karyotype, serum testosterone in male range
- Have testes
- Lack androgen tissue receptors (no pubic hair)
- Peripheral aromatization of androgens to estrogen (breast)
Complete Androgen Insensitivity

TREATMENT

--counseling
--gonadectomy after sexual maturation is complete
--estrogen therapy as needed
--vaginoplasty/dilators
111. Pituitary microadenoma

A 28-year-old woman presents with a history of infrequent menstrual periods. Her last period was 4 months ago. Physical examination reveals a body mass index (BMI) of 26 kg/m² and galactorrhea bilaterally. Lab results show a negative β-hcg level, a normal TSH level, a DHEAS level of 180 µg/dL, and a fasting prolactin level of 75 ng/mL. A repeat prolactin test confirms the prior elevated level. A MRI study of her pituitary gland is normal. The most appropriate next step in management is

(A) clomiphene citrate (Clomid, Serophene)
(B) dexamethasone (Decadron)
(C) weight reduction
(D) metformin (Glucophage)
(E) bromocriptine mesylate (Parlodel)
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