Puberty

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Puberty Dr. Ashraf Fouda Damietta General Hospital Puberty It is a physiological phase lasting 2 to 5 years, during which the genital organs mature. Manifestations of puberty in the female include:
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It is a physiological phase lasting 2 to 5 years, during which the genital organs mature.
Manifestations of puberty in the female include:

1) Menarche
2) Appearance of secondary sex characters
3) Physical development and
4) Psychological changes
Secondary sex characters include:
Development of the breast, appearance of pubic and axillary hair.
• The first sign of pubertal development is usually breast growth (thelarche), followed by appearance of pubic hair (pubarche), then (axillary hair), then (menarche).

• The mean interval between breast budding and menarche is 2.5 years with a standard deviation of about one year.
Adrenarche
The increased activity of the suprarenal cortex at puberty with increased production of adrenal androgens which lead to appearance of pubic and axillary hair.
Cause of Puberty

- During childhood, the hypothalamus is extremely sensitive to the negative feedback exerted by the small quantities of estradiol & testosterone produced by the child's ovaries.
- As puberty approaches, the sensitivity of the hypothalamus is decreased and subsequently, it increase the pulsatile GnRH secretion.
The anterior pituitary responds by progressive secretion of FSH and LH associated with increased secretion of growth hormone.
The ovaries respond to the increase Gonadotrophin secretion by follicular development & estrogen secretion.
• Estrogen causes development of the genital organs and the appearance of the secondary sexual characters.

• With increased estrogen secretion, menarche and cyclic estrogen secretion occurs.
Factors Affecting the Initiation of Pubertal Development

1 - Height and weight ratio (nutritional factors)
2 - Maturation of the hypothalamus
3 - Increased neurotransmitter output in CNS
4 - Onset of adrenal androgen activity
Deposition of SC Fat
17% to menstruate & 22% to ovulate
Genital Organ Changes

- Mons pubes, labia majora & minora: increase in size
- Vagina:
  1. length: increase, appearance of the rugae
  2. Epithelium: thick, stratified squamous, containing glycogen
  3. pH: acidic
Genital Organ Changes

- **Uterus:**
  1) enlarge, uterus / cervix : 2 / 1

- **Ovaries:**
  1. Increase in size, almond shape
  2. 300 thousands primary follicle at menarche (2 million at birth)
Adolescence

- *Is the period of life during which the child becomes an adult person*
- *i.e. the physical, sexual and psychological development are complete.*
- *Puberty represents the first part of adolescence.*

Adolescence
Is the period of life during which the child becomes an adult person i.e. the physical, sexual and psychological development are complete.
Puberty represents the first part of adolescence.
Abnormalities of Puberty

1 - Precocious puberty
2 - Delayed puberty
3 - Growth problems: during adolescence e.g. short stature or tall stature, marked obesity and menstrual disorders at puberty.
Definition
It means menarche or appearance of any of the secondary sexual characters before the age of 8 years.
Types

1 - **True** precocious puberty
2 - **False** (pseudo-precocious puberty)
3 - **Incomplete** precocious puberty
1. True (central, cerebral) Precocious Puberty
It is due to increased production of pituitary gonadotropins.
2. False (peripheral) Precocious Puberty

It is of peripheral origin. It is due to secretion of sex hormones; (estrogen or androgen) which is not dependent on pituitary gonadotropins as in case of estrogenic or androgenic ovarian tumors.
2. False (peripheral) precocious puberty

- False precocious puberty may be isosexual or heterosexual.
- A girl who feminizes early is defined as having isosexual precocious puberty.
- A girl who virilized early is defined as having heterosexual precocious puberty. (female pseudohermaphrodite)
3. Incomplete Precocious Puberty

- In this case only one pubertal change as breast development is present before the age of 8 years without the presence of any other pubertal changes and in absence of increased estrogen production.
- The other pubertal changes occur at the normal age.
3. Incomplete Precocious Puberty

Incomplete forms of precocious puberty include premature thelarche (unilateral or bilateral), premature pubarche and premature adrenarche with appearance of pubic and axillary hair.
Etiology of Precocious Puberty

1. Constitutional or idiopathic:
   • In most cases of precocious puberty (90%), no cause is found.
   • For some unknown reason the hypothalamus stimulates the pituitary gland to secrete its gonadotrophic hormones.
   • There is normal menstruation and ovulation.
   • Pregnancy can occur at young age.
Etiology of Precocious Puberty

2. Organic lesions of the brain:
   • The next common cause.
   • Organic lesions affecting the midbrain, hypothalamus, pineal body, or pituitary gland may lead to premature release of pituitary gonadotrophins.
   • Examples include traumatic brain injury, meningitis, encephalitis, brain abscess, brain tumor as glioma, craniopharyngioma, and hamartomas.
Etiology of Precocious Puberty

3. McCune-Albright Syndrome
4. Adrenal causes:
   (a) Hyperplasia, adenoma, or carcinoma of suprarenal cortex. *Congenital adrenal hyperplasia* and *Cushing syndrome* lead to precocious puberty in the male direction, i.e. heterosexual precocious puberty;
   (b) Estrogen secreting adrenal tumor which is very rare.

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Etiology of Precocious Puberty

5. Ovarian causes:

(a) Estrogen producing tumors as granulosa and theca cell tumor;
(b) Androgen producing tumors as androblastoma;
(c) Choriocarcinoma because it secretes human chorionic gonadotrophin (HCG) which may stimulate the ovaries to secrete estrogen;
(d) Dysgerminoma if it secretes HCG.
Etiology of Precocious Puberty

6. **Juvenile hypothyroidism:**
Lack of thyroxine leads to increased production of thyroid stimulating hormone and the secretion of pituitary gonadotrophins may also be increased.

7. **Drugs:**
- Iatrogenic may follow oral or local administration of estrogen.
- A long course of estrogen cream used for treatment of vulvovaginitis of children may lead to breast development or withdrawal bleeding.

8. **Silver syndrome:** Small stature, retarded bone age and increased Gonadotrophin levels.
Diagnosis of Precocious Puberty

1. History:

- It excludes iatrogenic source of estrogen or androgen.
- It differentiates between isosexual and heterosexual precocious puberty.
Diagnosis of Precocious Puberty
2. Physical examination:
   • It diagnoses McCune-Albright syndrome.
   • Neurologic and ophthalmologic examinations exclude organic lesions of the brain.

Female Precocious Puberty
3. Special Investigations:
   These are done according to the history and clinical findings and include:
3. Special Investigations

a. **X-ray examination of the hand and wrist** to determine bone age.

- Estrogen stimulates growth of bone but causes early fusion of the epiphysis.
- So the child is taller than her peers during childhood, but she is short during adult life.
3. Special Investigations

b. **Hormonal assay:**
including serum FSH, LH, prolactin, estradiol, testosterone, 17α-hydroxyprogesterone, TSH, and human chorionic gonadotrophin to diagnose Choriocarcinoma.
3. **Special Investigations**

c. **Ultrasonography** to diagnose ovarian or adrenal tumor.

d. **CT or MRI** to diagnose an organic lesion of the brain, or adrenal tumor.
Hypothyroidism retards bone age. It is the only condition of precocious puberty in which bone age is retarded.
Idiopathic Precocious Puberty is diagnosed after excluding all other causes.
Treatment of Precocious Puberty

Objectives:

1. Arrest maturation until normal pubertal age.
2. Attenuate & diminish established precocious characteristics.
3. Maximize adult height.
4. Avoid abuse, reduce emotional & social problems
Treatment of Precocious Puberty

1. Treatment of the cause e.g., thyroxin for hypothyroidism, removal of ovarian and adrenal tumors.

2. Incomplete forms of precocious puberty do not require treatment, as estrogen production is not increased.
3. McCune-Albright Syndrome
• is treated with testolactone oral tablets.
• The drug inhibits the formation of estrogen from its precursors, so reduces estrogen level.
• The dose is 20 mg/kg body weight in 4 divided doses and increased to 40 mg/kg body weight during a 3 week interval.
4. Idiopathic Type is treated by explanation and reassurance and by giving one of the following drugs which inhibit the secretion of gonadotrophins:

(a) Gonadotrophin releasing hormone analogues which are given as daily nasal spray, intramuscular, or subcutaneous injections every 4 weeks.

(b) Medroxyprogesterone acetate tablets (Provera tablets) or intramuscular injection (Depo-Provera);

(c) Danazol capsules;

d) Cyproterone acetate tablets (Androcur).
4. Idiopathic Type
Treatment is given until the age of 12 years (mean age of pubertal development).
### Gonadotrophin Releasing Hormone Analogues

**Drug of choice because it achieves all objectives:**

1. It acts by binding to the anterior pituitary receptors causing down-regulation & desensitization of the pituitary
2. Regression of symptoms occurs in the first year
3. Delayed epiphyseal fusion; treatment more effective if begun before bone age >12 yrs
4. Maintain E2 at <10 pg/mL
5. Children require higher doses than adults for suppression
6. Adrenarche will continue

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**Gonadotrophin Releasing Hormone Analogues**

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2) Regression of symptoms occurs in the first year
3) Delayed epiphyseal fusion; treatment more effective if begun before bone age >12 yrs
4) Maintain E2 at <40 pg/mL
5) Children require higher doses than adults for suppression
6) Adrenarche will continue
McCune-Albright Syndrome

- The disease is found more frequently in girls.
- It consists of a triad of:
  1. Precocious puberty,
  2. Cystic changes in bones, and
- The cause of precocious puberty is autonomous production of estrogen by the ovaries.
- FSH and LH levels are low.
- The treatment is testolactone oral tablets which inhibit ovarian steroidogenesis.

McCune-Albright Syndrome

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- It consists of a triad of:
  1) Precocious puberty
  2) Cystic changes in bones and
  3) Cafe-au-lait patches of the skin
- The cause of precocious puberty is autonomous production of estrogen by the ovaries.
- FSH and LH levels are low
- The treatment is testolactone oral tablets which inhibit ovarian steroidogenesis
Delayed Puberty
Secondary Sexual Characters do not develop by the age of 14 years or no menstruation until age of 16 years
Delayed Puberty

It is either:

* **Delayed onset:** Breast bud does not appear until 13 years or menarche does not occur till 16 years
  
or

* **Delayed progression:** Menarche does not occur within 5 years after breast bud.
### Etiology of Delayed Puberty

1. **Constitutional**
   - with +ve family history, short stature & normal fertility.

2. **Hypergonadotropic hypogonadism**
   - (FSH > 40) = ovarian causes of primary amenorrhea = primary ovarian failure & secondary ovarian failure (if occurs before puberty).

3. **Hypogonadotropic hypogonadism**
   - hypothalamic & pituitary causes of primary amenorrhea e.g. Kallman's syndrome, Anorexia nervosa.

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**Etiology of Delayed Puberty**

1. Constitutional:
   - with positive family history, short stature and normal fertility
2. Hypergonadotropic:
   - (FSH > 40) = ovarian causes of primary amenorrhea = primary ovarian failure and secondary ovarian failure (if occurs before puberty)
3. Hypogonadotropic hypogonadism = hypothalamic and pituitary causes of primary amenorrhea e.g. Kallman’s syndrome, Anorexia nervosa
Etiology of Delayed Puberty

4 - Normogonadotropic hypogonadism
   = end organ defects = uterine causes
   (Mullerian agenesis and testicular feminization syndrome), imperforate hymen
   (c/o = delayed menarche + normal other aspects of puberty), PCOD and Virilizing ovarian adrenal tumors.

5 - General causes of amenorrhea
   (endocrinal or non-endocrinal especially malnutrition) if occurred before puberty & decreased GH and steroid synthesis defects.
Investigations of Delayed Puberty

History:

1 - Family history, nutritional history, any systemic diseases (e.g. history of endocrinal disturbance).

2 - Clinical picture of space occupying lesion in the ovary, adrenal, pituitary & hypothalamus.

3 - Periodic pain and positive secondary sexual characteristics in imperforate hymen.
Investigations of Delayed Puberty

Examination:

(A) Body measurement for causes of amenorrhea + increased or decreased weight, short or tall stature, proportions (upper / lower segment ratio & arm span / height ratio).

(B) Tanner staging of breast, pubic & axillary hair if present.

(C) Clinical picture of Turner, Mullerian agenesis & imperforate hymen.

(D) Neurological examination for smell sense (Kallman's syndrome), visual field & other cranial nerve lesions.
Special Investigations

1. **FSH & LH assay** important to differentiate level of the lesion & progesterone assay in 17 OH deficiency
2. **Chromosomal study** if short stature or hypergonadotropic type
3. **Radiological bone age study** & radiologic study for pituitary adenoma

Special Investigations
1) FSH & LH assay important to differentiate level of the lesion & progesterone assay in 17 OH deficiency
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3) Radiological bone age study & radiologic study for pituitary adenoma
**Treatment of Delayed Puberty**

* **Constitutional:** Reassurance

* **Treatment of the cause (if treatable) or cyclic estrogen-progesterone hormone replacement therapy if the cause is not treatable, for 3 cycles:** Norethistrone acetate 5 mg twice daily for 21 d or OCP

* **Patient with Y chromosome cell line:** Gonadectomy + hormone replacement therapy
Thank you

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