King Saud University Medical City
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Department of Obstetrics & Gynecology
Course GYN 482

PUBERTY

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Objectives

- Recognize sexual differentiation
- Know the role of sex chromosome in sexual differentiation .
- To know the normal phases of female puberty
- Recognize the causes of abnormal puberty

Sexual Differentiation

- The embryo differentiates into female or male through the sex chromosomes .
- Sex chromosomes XY is a male and XX is a female.
- Gonadal sex is the differentiation of either ovaries or testes .
- Subsequent development of the internal and external genitalia give phenotypic sex .

- In the presence of Y chromosome the undifferentiated gonad will become a testis .
- Absence of Y chromosome will result in the development of the ovaries .
- We need at least one X chromosome for embryo development .
- The testis produce androgen and mullerian inhibitor

- The undifferentiated embryo contain both Wolffian and Mullerian ducts .
- Wolffian duct will develop the male internal organs .
- Mullerian duct will develop female internal organs .
- The leydig cell produces testosterone that promotes the development of the Wolffian duct will lead to vas deferens, epididymis and the seminal vesicles

- Dihydrotestosterone acts on the cloaca to form the penis and the scrotum .
- Absence of the testosterone means the wolffian duct will regress and the cloaca will be an external female genitalia

Normal Puberty

- It happens as the result of the maturation of the hypothalamo pitutary ovarian axis .
- The gonadotrophin releasing hormones is produced and the gonadotrophin FSH and LH will increase in frequency and amplitude .
- This will lead to full establishment of the normal ovulatory menstrual cycle .
- Puberty occurs over a period of 5 to 10 years

Physiology of Puberty

- This is result in the physical changes resulting in female adult life in these sequences .
- Growth spurts .
- Breast development .
- Pubic hair growth .
- Menarche .
- Finally axillary hair growth .
- This sequences occurs in 70% of female and variation may happens .

- Growth spurts starts at the age of 11
- 6-10 cm per year .
- By the age of 15 most girl will achieve their final height.
- Menstrual cycles in the region between 9 and 16 and usually are irregular because of the immaturity of the axis .

Precocious Puberty

- Puberty before the age of 9 years .
- Causes.
- Idiopathic .
- MaCune Albright syndrome.
- Tumor of adrenal and ovary producing hormones.
- Cerebral tumor.
- Ingestion of exogenous estrogens.

- The commonest cause is simply is premature maturation of hypothalamus and production of the gonadotropin releasing hormones.
- This can be treated with gonadotropin releasing hormones agonist GnRHa .
- But other serous causes should be excluded like brain tumor

Delayed Puberty

- Mostly patient come because of delay in the menstruation .
- It is important to establish whether puberty itself is delayed.
- Detailed history is taken about other secondary sexual characters.
- Exclude chronic illness .
- Family history.

Investigations

- Gonadotropins level FSH and LH .
- Karyotyping .
- Pelvic ultrasound to confirm the presence of the uterus and ovaries .
- Possibly X- ray to determine bone age.
- Other like thyroid function test prolactin and 17-alpha-hydroxy-progesterone .

Hypogonadotropic hypogonadism

- Majority is constitutional delay in puberty.
- May be secondary to chronic illness and improvement of underlying condition is the treatment.
- Anorexia nervosa at young age have low levels of gonadotrophin .
- Athletic girls .
- Congenital deficiency of gonadotropin with hypoplasia of olfactory lobe Kallman syndrome

- Acquired damage to hypothalamus and pituitary by tumor, trauma ,infection , radiation , secondary to hydrocephalus and hemochromatosis due to repeated transfusion in sickle cell disease , thalassemia and willson disease .
- In all cases the ultrasound will confirm the immature uterus and small inactive ovaries,

- Most girls with constitutional delay will proceed to normal development if left untreated.
- Otherwise treatment is replacement with gonadotropin or estrogen and progesterone .

Hypergonadotropic hypogonadism

- Failure of gonadal development.
- No negative feed back from the gonads .
- Commonest cause is Turner syndrome 45xo .
- Damage to the ovaries by infection , irradiation, chemotherapy, or surgery .
- Autoimmune disease such as Adison , vitiligo, and hypothyroidism.

- Turner syndrome.
- Features.
- Wide carrying angle of the arms .
- Webbed neck .
- Broad chest and widely spaced nipples .
- May have color blindness, co-arctation of the aorta .
- Streak ovaries and may be a small uterus.

- Treatment by hormone replacement therapy estrogen and progesterone.
- Gonadal causes carries a bad prognosis for pregnanacy .

- Anatomical causes ,
- Normal puberty but no menstrual cycle .
- Imperforate hymen or transverse vaginal septum.
- 1-They present with amenorrhea ,cyclical pain and sometime retention of urine .
- Treat with incision of the hymen or the septum .
- 2- mullarian agenesis , no uterus ,fallopian tubes and vagina.
- Exclude urinary tract anomalies.

Androgen insensitivity syndrome

- Normal breast but scanty or absent pubic hair.
- This is due androgen insensitivity syndrome .
- The karyotype (genotype) is XY and phenotype is a female .
- They have testes .
- There is no uterus, fallopian tubes, and upper two third of the vagina.

- Management .
- The patient is brought up as a female .
- Remove the testes because of the risk of malignant transformation .
- Start hormonal replacement therapy .
- Create a vagina for satisfactory sexual intercourse .

- Abnormal uterine bleeding .
- Is common after the menarche .
- Mainly due the un-ovulatory cycles .
- In case of menorrhagia treat if it is affecting the general condition of the patient .
- Exclude other blood diseases hemophilia and Vonwillibrand disease .
- Oligo menorrhea reassure the patient .
- It is usually improve spontaneously with time .

Congenital adrenal hyperplasia CAH

- Autosomal recessive .
- Mainly 21 hydroxylase deficiency
- Excessive androgen sex hormones .

- Clinical picture .
- Ambiguous genitalia.
- Either delayed or preconscious puberty .
- Excessive fascial hair .
- Virilization clitoromegaly .
- menstrual disorder .
- Infertility.

- Investigations.
- Hormonal assay .
- 17-hydroxyrogesterone is high .