أمراض الغدد الصم

4

أ. م. د. زينب العرفي

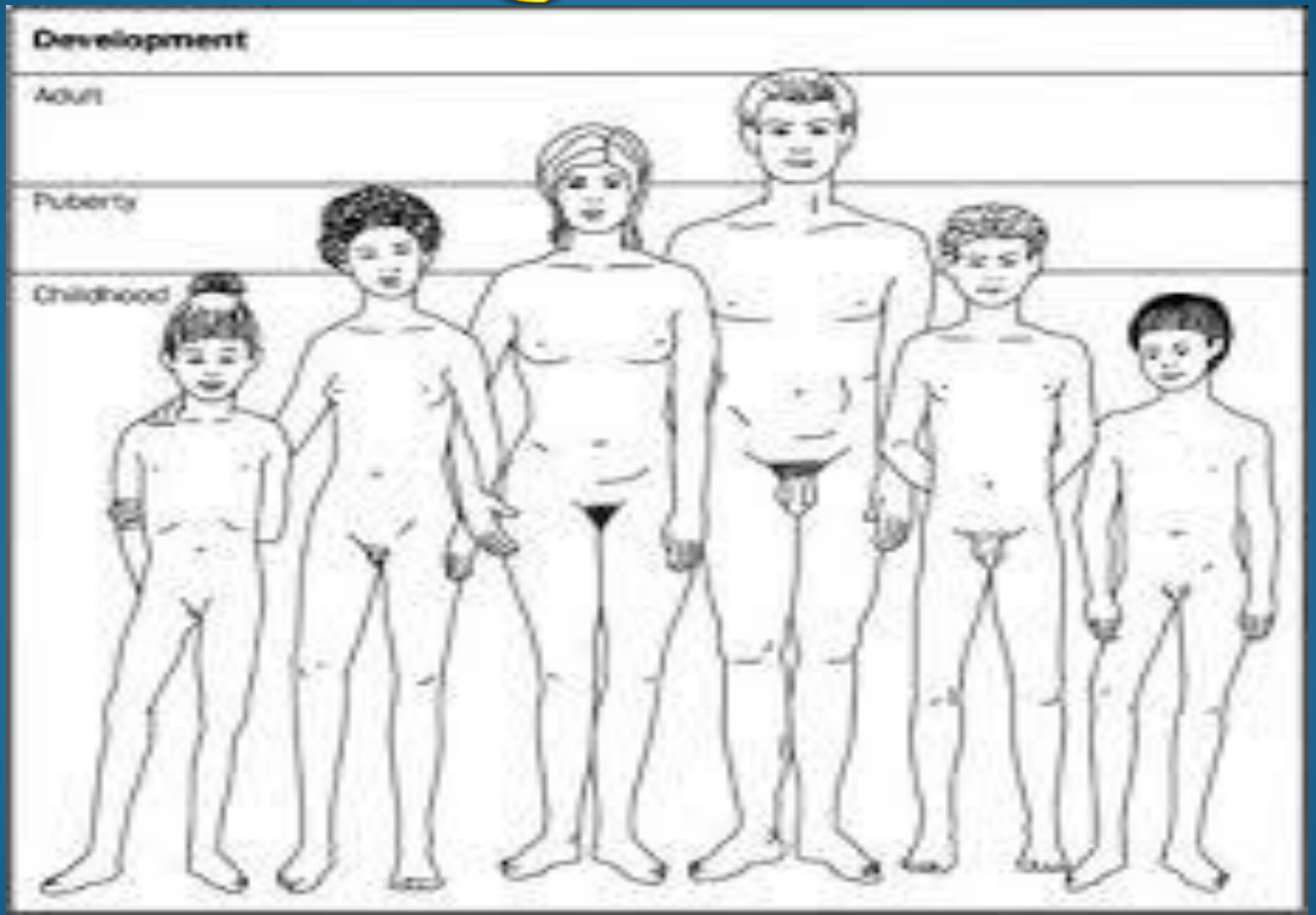
القزامة النخامية

- الذكاء طبيعي
- نسبة الجذع للأطراف سوية
- الصحة العامة طبيعية
- تأخر العمر العظمي عن الزمني لكنه مناسب للعمر الطولي
- قد يرافقها تأخر البلوغ

التشخيص التفريقي

- تأخر البلوغ الفيزيولوجي
- القزامة العائلية
- تناذر لارون (مقاومة على مستوى المستقبلات
- قصور الدرق البدئي
- تناذر تورنر
- سوء التغذية والأمراض الجهازية
- تناذر الطفل المضطهد

البلوغ



Tanner Staging مقياس تانر لمراحل البلوغ
Sexual Maturation Rating (SMR)

مرحلة البلوغ: تبدلات فيزيولوجية وشكلية (المظاهر الجنسية الثانوية) تتطور هذه المظاهر تدريجياً وبترتيب معين.

يبدأ البلوغ عند :

الأنثى بظهور برعم الثدي

الذكر ببدء كبر حجم الخصيتين.

ثم تمر هذه المرحلة بتطورات تدريجية حتى الاكتمال.

لإيجاد لغة طبية لوصف المظاهر دون الحاجة لتفاصيل أو صور ودون الاعتماد على العمر قام كل من W.A.MARSHALL and J. M.TANNER

بعمل هام جداً انتهى بوضع مقياس للبلوغ والنضج الجنسي

GIRLS

Sexual Maturity Rating

Breast Development

1 Prepubertal; nipple elevation only

2 Small, raised breast bud

3 General enlargement of raising of breast and areola

4 Further enlargement with projection of areola and nipple as secondary mound

5 Mature, adult contour, with areola in same contour as breast, and only nipple projecting

Source: Data from Tanner JM. Growth at adolescence. Oxford: Blackwell Scientific Publications, 1962.

BOYS

Sexual Maturity Rating

Genital Development

1. Prepubertal; no change in size or proportion of testes, scrotum and penis from early childhood
2. Enlargement of scrotum and testes; reddening and change in texture in skin of scrotum; little or no penis enlargement
3. Increase first in length then width of penis; growth of testes and scrotum
4. Enlargement of penis with growth in breadth and development of glands; further growth of testes and scrotum, darkening of scrotal skin
5. Adult size and shape genitalia

Source: Data from Tanner JM. Growth at adolescence. Oxford: Blackwell Scientific Publications, 1962.

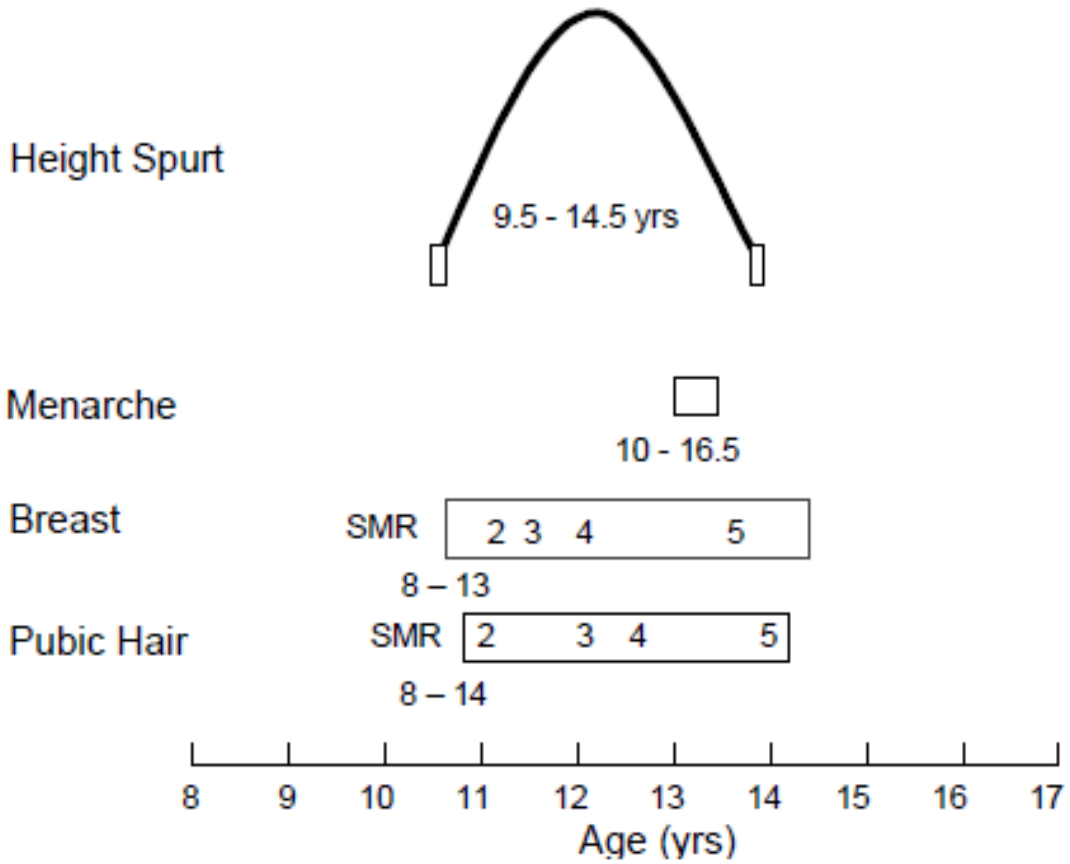
Pubic Hair Growth

Sexual Maturity Rating

	BOYS		GIRLS
1	Prepubertal; no pubic hair	1	Prepubertal; no pubic hair
2	Sparse growth of hair at base of penis	2	Sparse growth of hair along labia
3	Darkening, coarsening and curling, increase in amount	3	Pigmentation, coarsening and curling, with an increase in amount
4	Hair resembles adult type, but not spread to medial thighs	4	Hair resembles adult type, but not spread to medial thighs
5	Adult type and quantity, spread to medial thighs	5	Adult type and quantity, spread to medial thighs

Source: Data from Tanner JM. Growth at adolescence. Oxford: Blackwell Scientific Publications, 1962.

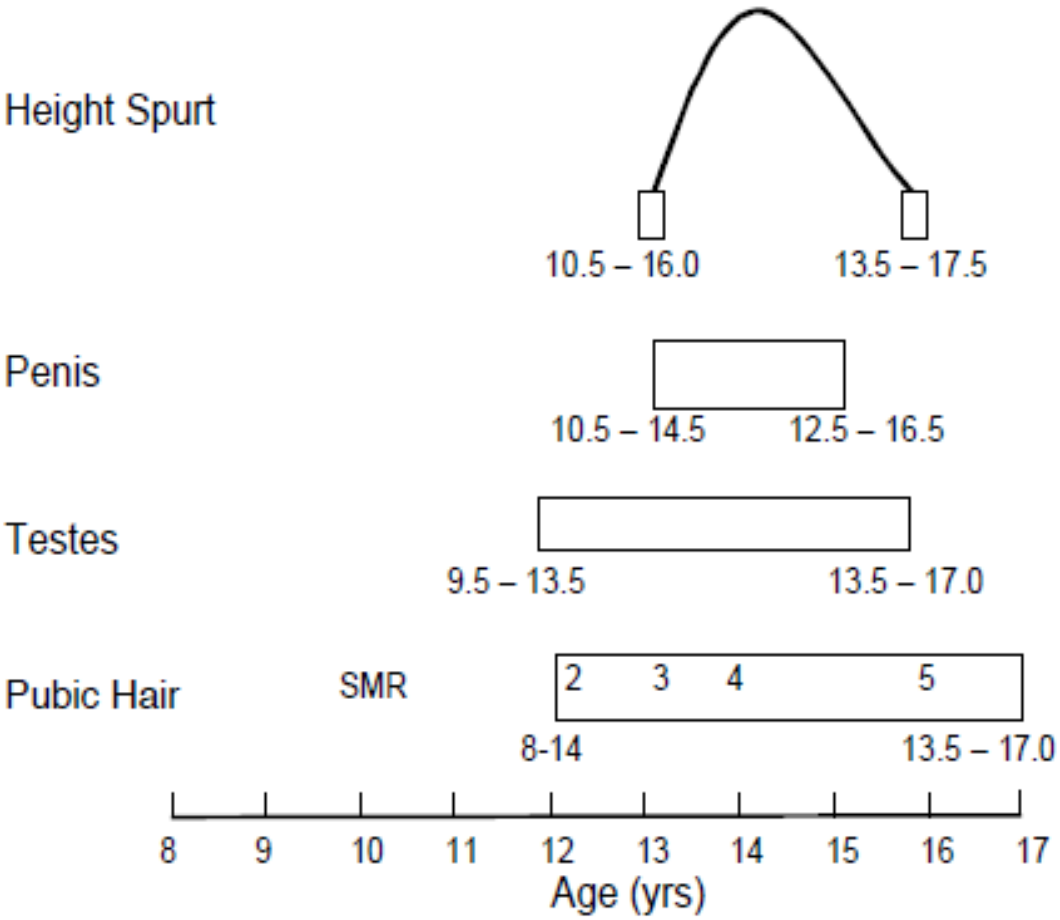
Sequence of Physiological Changes During Puberty in Females



*An average female is represented: the range of ages within which some of the events may occur is given by the figures placed directly below them.

Source: Adapted from Tanner JM. Growth at adolescence. Oxford: Blackwell Scientific Publications; 1962. Reprinted with permission. http://www.blackwell-synergy.com/

Sequence of Physiological Changes During Puberty in Males



An average male is represented: the range of ages within which each event charted may begin and end is given by the figures placed directly below its start and finish.

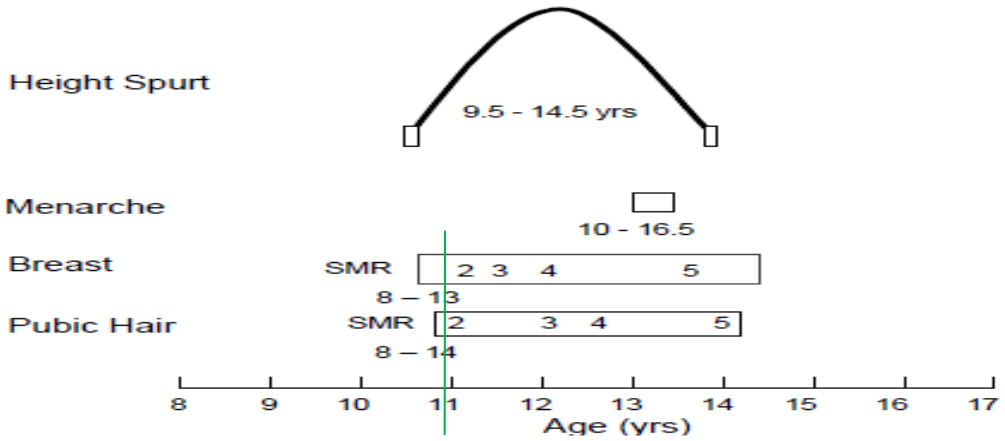
Source: Adapted from Tanner JM. Growth at adolescence. Oxford: Blackwell Scientific Publications; 1962. Reprinted with permission. http://www.blackwell-synergy.com/

Sexual Maturity Rating

GIRLS

Breast Development	Stage	Pubic Hair Growth
Prepubertal; nipple elevation only	1	Prepubertal; no pubic hair
Small, raised breast bud	2	Sparse growth of hair along labia
General enlargement of raising of breast and areola	3	Pigmentation, coarsening and curling, with an increase in amount
Further enlargement with projection of areola and nipple as secondary mound	4	Hair resembles adult type, but not spread to medial thighs
Mature, adult contour, with areola in same contour as breast, and only nipple projecting	5	Adult type and quantity, spread to medial thighs

Sequence of Physiological Changes During Puberty in Females



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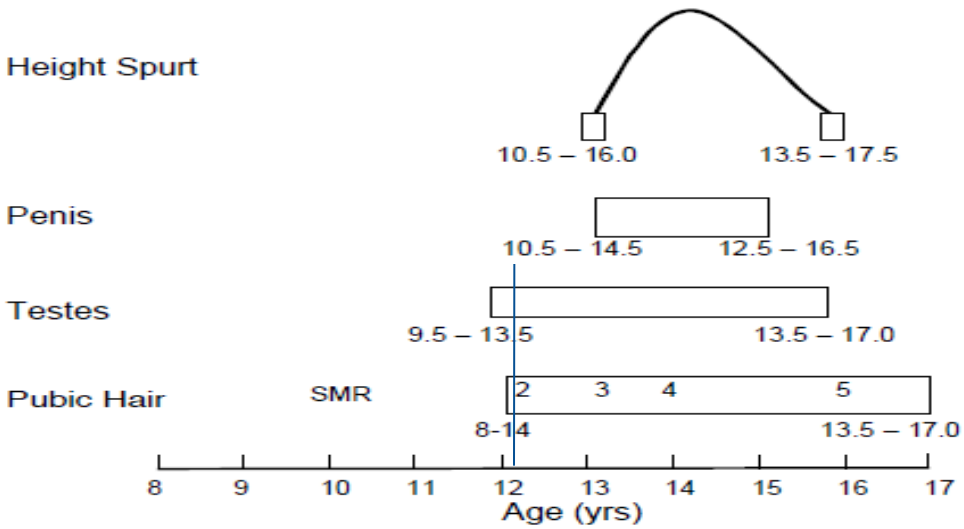
BOYS

Genital Development	Stage	Pubic Hair Growth
Prepubertal; no change in size or proportion of testes, scrotum and penis from early childhood	1	Prepubertal; no pubic hair
Enlargement of scrotum and testes; reddening and change in texture in skin of scrotum; little or no penis enlargement	2	Sparse growth of hair at base of penis
Increase first in length then width of penis; growth of testes and scrotum	3	Darkening, coarsening and curling, increase in amount
Enlargement of penis with growth in breadth and development of glands; further growth of testes and scrotum, darkening of scrotal skin	4	Hair resembles adult type, but not spread to medial thighs
Adult size and shape genitalia	5	Adult type and quantity, spread to medial thighs



An average male is represented: the range of ages within which each event charted may begin and end is given by the figures placed directly below its start and finish.

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Delayed puberty

Puberty is considered to be delayed if the onset of the physical features of sexual maturation has not occurred by a chronological age that is 2.5 standard deviations (SD) above the national average. In the UK, this is by the age of 14 in boys and 13 in girls. Genetic factors have a major influence in determining the timing of the onset

20.22 Causes of delayed puberty and hypogonadism

Constitutional delay

Hypogonadotrophic hypogonadism

- Structural hypothalamic/pituitary disease (see Box 20.59, p. 787)
- Functional gonadotrophin deficiency
  - Chronic systemic illness (e.g. asthma, malabsorption, coeliac disease, cystic fibrosis, renal failure)
  - Psychological stress
  - Anorexia nervosa
  - Excessive physical exercise
  - Hyperprolactinaemia
  - Other endocrine disease (e.g. Cushing's syndrome, primary hypothyroidism)
- Isolated gonadotrophin deficiency (Kallmann's syndrome)

Hypergonadotrophic hypogonadism

- Acquired gonadal damage
  - Chemotherapy/radiotherapy to gonads
  - Trauma/surgery to gonads
  - Autoimmune gonadal failure
  - Mumps orchitis
  - Tuberculosis
  - Haemochromatosis
- Developmental/congenital gonadal disorders
  - Steroid biosynthetic defects
  - Anorchidism/cryptorchidism in males
  - Klinefelter's syndrome (47XXY, male phenotype)
  - Turner's syndrome (45XO, female phenotype)

Anorchidism: congenital absence of one or both testes

النخامى العصبية

النخامى العصبية

- خزان لمفرز النواتين جانب البطينية وفوق البصرية:
  - Vasopressine Antidiuretic hormone (ADH)
  - Oxytocin

فازوبريسين الهرمون المضاد للإدرار
Anti-Diuretic Hormon (ADH)

- ينظمه: الضغط الحلولي وحجم المصورة
- يزيد افرازه: الخوف والألم والجهد والحرارة والمورفين والتدخين والنزف الشديد والايثر
- ينقص افرازه: الكحول والأدرينالين والبرد

البيلة التفهة
Diabitis Insipidis

- نقص إفراز الفازوبرسين (الهرمون المضاد للإدرار)
- بول ممدد (نقص أوزمولية البول) - بوال
- زيادة أوزمولية الدم – سهاف
- مركزية وكلوية

20.65 Causes of diabetes insipidus

Cranial

Structural hypothalamic or high stalk lesion

- See Box 20.59

Idiopathic

Genetic defect

- Dominant (*AVP* gene mutation)
- Recessive (DIDMOAD syndrome – association of diabetes insipidus with diabetes mellitus, optic atrophy, deafness)

Nephrogenic

Genetic defect

- V2 receptor mutation
- Aquaporin-2 mutation
- Cystinosis

Metabolic abnormality

- Hypokalaemia
- Hypercalcaemia

Drug therapy

- Lithium
- Demeclocycline

Poisoning

- Heavy metals

Chronic kidney disease

- Polycystic kidney disease
- Sickle-cell anaemia
- Infiltrative disease

20.59 Causes of anterior pituitary hormone deficiency

Structural

- Primary pituitary tumour
  - Adenoma*
  - Carcinoma (exceptionally rare)
- Craniopharyngioma*
- Meningioma*
- Secondary tumour (including leukaemia and lymphoma)
- Chordoma
- Germinoma (pinealoma)
- Arachnoid cyst
- Rathke's cleft cyst
- Haemorrhage (apoplexy)

Inflammatory/infiltrative

- Sarcoidosis
- Infections, e.g. pituitary abscess, tuberculosis, syphilis, encephalitis
- Lymphocytic hypophysitis
- Haemochromatosis
- Langerhans cell histiocytosis

Congenital deficiencies

- GnRH (Kallmann's syndrome)* – gonadotrophin-releasing hormone
- GHRH* – growth hormone-releasing hormone
- TRH – thyrotrophin-releasing hormone
- CRH – corticotrophin-releasing hormone

Functional*

- Chronic systemic illness
- Anorexia nervosa
- Excessive exercise

Other

- Head injury*
- (Para)sellar surgery*
- (Para)sellar radiotherapy*
- Post-partum necrosis (Sheehan's syndrome)
- Opiate analgesia

*The most common causes of pituitary hormone deficiency.

20.66 How and when to do a water deprivation test

Use

- To establish a diagnosis of diabetes insipidus and to differentiate cranial from nephrogenic causes

Protocol

- No coffee, tea or smoking on the test day
- Free fluids until 0730 hrs on the morning of the test, but discourage patients from 'stocking up' with extra fluid in anticipation of fluid deprivation
- No fluids from 0730 hrs
- Attend at 0830 hrs for body weight, plasma and urine osmolality
- Record body weight, urine volume, urine and plasma osmolality and thirst score on a visual analogue scale every 2 hrs for up to 8 hrs
- Stop the test if the patient loses 3% of body weight
- If plasma osmolality reaches > 300 mOsm/kg and urine osmolality < 600 mOsm/kg, then administer DDAVP (see text) 2 μg IM

Interpretation

- Diabetes insipidus is confirmed by a plasma osmolality > 300 mOsm/kg with a urine osmolality < 600 mOsm/kg
- Cranial diabetes insipidus is confirmed if urine osmolality rises by at least 50% after DDAVP
- Nephrogenic diabetes insipidus is confirmed if DDAVP does not concentrate the urine
- Primary polydipsia is suggested by low plasma osmolality at the start of the test

وظيفة

1. ترجمة الأشكال والجداول
2. كيفية حساب أوزمولية البلازما
3. التشخيص التفريقي لبوال وسهاف

مريض 50 سنة شخص له سرطان بنكرياس بدأ يشكو من قلق وعدم توجه وتخليط ذهني. قارب

Syndrome of Inappropriate Secretion of Antiduretic Hormone (SIADH)

SIADH

فرط إفراز الهرمون المضاد للإبالة

- فرط إماهة
- نقص ضغط المصورة الحلولي
- نقص تركيز صوديوم الدم

الأسباب

- وطائية: التهابية، أو ورمية، أو ارتشاحية
- فرط افرازغير غدي ل ADHأو مادة شبيهة به: سرطان الرئة صغير الخلايا، وسرطان بنكرياس، وسرطان بروستات، وتيموما
- دوائي: كلوربروباميد- كاربامازبين – فنكرستين – نيكوتين – فينوتيازين – سيكلوفوسفاميد
- أمراض رئوية: الخراجة الرئوية، وذات الرئة
- الأمراض العصبية: التهاب السحايا الدرني والتهاب الدماغ ونزف تحت العنكبوت ورضوض الرأس وخراجة الدماغ
- معالجة مفرطة بالفازوبرسين أو مقلداته

الوظيفة: معالجة SIADH


شكراً
لإصغائكم