A 17 year old boy has had 6 episodes of DKA in the past year. He receives Lantus q.a.m. and Humalog insulin with each meal. Physical exam is unremarkable, TSH 1.5 uU/ml, T4 8.9ug/dl, and HbA1c 11.7%
1. What is the most appropriate next step?

A  Start Symlin b.i.d..
B  Consult a psychologist.
C  Nuclear study of gastric emptying.
D  C-peptide analysis after ingestion of Sustacal.

2

• A 15 year old girl presents with the sudden onset of polydipsia and polyuria. She has not had a menstrual cycle for 4 months. Physical exam is unremarkable.

• Fasting a.m. tests: glucose 86 mg/dl, cortisol 18 ug/dl, prolactin 21 ng/ml, T4 7.9 ug/dl, FSH 3 mIU/ml, serum osmolality 304 mosm/kg, urine osmolality 77 mosm/kg. A 24 hour urine collection was 3145 ml.
2. Her MRI scan shows thickening of the pituitary stalk. A lytic lesion is seen in her mandible.

The most likely diagnosis is:
   A  Sarcoidosis
   B  Tuberculosis
   C  Craniopharyngioma
   D  Histiocytosis

3. A nine year old boy has grown 3.2 cm in the past year.

Which test is **NOT** helpful:
   A  random growth hormone level
   B  random IGF-I level
   C  post exercise growth hormone level
   D  growth hormone level obtained 1 hour after the ingestion of 0.2 mg of clonidine.
• An obese adolescent African-American male is seen 1 month after presenting in diabetic ketoacidosis. Acanthosis nigricans is noted around his neck and in the axilla. Which of the following observations is most likely:

4. Which of the following observations is most likely:

A  his daily insulin dose is over 1.5 U/kg/day.
B  he no longer requires insulin.
C  he has lost 40#
D  his HbA1c is in the non-diabetic range.
A 15 year old boy presents with increased thirst, polyuria, and a 10# weight loss over the past month. His blood glucose is 468 mg/dl, and his urine is positive for both glucose and ketones. Before starting insulin therapy you should obtain:

5. Before starting insulin therapy you should obtain:

A islet cell and insulin autoantibody titers
B glutamic acid decarboxylase (GAD) antibody titer
C plasma insulin level
D plasma C-peptide level
E none of the above.
6

• A 3 year old boy is noted to have pubic hair, 6cc testicles, adult body odor and penile enlargement. Aqueous Lupron 500 mcg is given S.C.

• Blood obtained 120 minutes later revealed:
  - LH 10.2 mIU/ml
  - TSH 1.3 uU/ml
  - HCG < 5 mIU/ml
  - DHEAs 40 ug/dl

6. The most likely diagnosis is:

A idiopathic central precocious puberty
B hypothalamic hamartoma
C dysgerminoma
D craniopharyngioma
E late onset congenital adrenal hyperplasia.
6.5
The second most likely diagnosis is:

A idiopathic central precocious puberty
B hypothalamic hamartoma
C dysgerminoma
D craniopharyngioma
E late onset congenital adrenal hyperplasia.

7 & 8

• A 10 year old boy has previously tracked at the 75% height for age. At a recent physical exam, it was noted that he had grown 1.8 cm in the last year, and was now at the 50%. He gained 5 kg in the last year.
7. The most likely diagnosis is:

A Crohn’s disease  
B Kallman’s syndrome  
C constitutional delayed onset of growth  
D familial short stature  
E growth hormone deficiency

8. Free T4 and TSH levels are normal. Growth hormone levels are low in response to stimulation with Glucagon and Clonidine. Which tumor is most likely to cause this boy’s problem:

A dysgerminoma  
B hamartoma  
C astrocytoma  
D craniopharyngioma
9

- A slender 9 year old boy presents because of concern about the size of his penis. His height is at the 75% for age. His penis is 4 cm long, and the testicles are 1 cc each. He cannot identify the odors of coffee or garlic.

9. This boy most likely:

A. has an androgen receptor defect
B. has a genetic defect in the LH beta subunit
C. has congenital adrenal hyperplasia due to a 3 beta-ol deficiency
D. lacks GnRH pulse generating cells in the hypothalamus.
10

• A 2 1/2 year old child with type 1 diabetes for 6 months has had 2 severe insulin reactions in the past month. Each hypoglycemic episode was associated with a seizure, and required glucagon. The child's growth and weight (13 kg) are normal. A HbA1c level is 6.6%. The parents want the child to have “normal” blood sugar levels.
  – Insulin:
    • Bedtime  3 units Lantus
    • Meals     1 unit Novolog per 25 grams CHO

10. The most appropriate plan at this point is:

A  continue current regimen
B  decrease the insulin dose
C  convince the parents that “tight control” in a 2 year old child will not help prevent diabetic complications
D  obtain a neurology consult to evaluate for a seizure disorder
E  obtain a cortisol and ACTH level.
11

• A 14 year old girl has had type 1 diabetes for 5 years. Recently she has been experiencing frequent hypoglycemic episodes, in spite of reducing her insulin doses. She is now only receiving 4 units of Lantus in the morning before breakfast.
• Fasting laboratory studies reveal:
  – C-peptide < 0.2 ng/ml
  – free insulin 54 mU/l
  – glucose 42 mg/dl
  – cortisol 24 ug/dl

11. The most likely cause of this girl’s hypoglycemia is:

A  functioning insulinoma
B  Addison’s disease
C  surreptitious insulin use
D  perirenal mesothelioma
E  remission of type 1 diabetes
12

• A 14 year old girl presents because of acne, hirsutism and secondary amenorrhea. She experienced menarche at 9 years of age. Mother remembers that she had pubic hair when she started kindergarten.

• Laboratory studies 1 hour after an I.M. injection of Cortrosyn 0.25mg:
  – free testosterone 11 ng/dl
  – DHEA-S 3200 ng/ml
  – 17-OH progesterone 1682 ng/dl

12. The most likely diagnosis is:

A arrhenoblastoma
B polycystic ovarian syndrome
C Cushing syndrome
D adrenal cortical adenoma
E late onset congenital adrenal hyperplasia
13. A 2 year old boy with asthma has ingested 40 yellow synthroid tablets.

Treatment options which might be helpful include:
A potassium iodide
B methimazole
C propranolol
D prednisone
E careful observation

14

• A 16 year old girl has had breast development since age 10, but has never menstruated. She denies sexual activity. Her breast are Tanner stage 5, without galactorrhea. Pubic hair is absent. She refuses to allow a vaginal exam.
14. Which of the following hormones is most likely to be elevated in this 16 year old girl:

A  testosterone
B  prolactin
C  follicle-stimulating hormone
D  thyroid stimulating hormone
E  17-OH progesterone

15 & 16

• A 13 year old honors student presents because of a lack of breast development. Pubic hair has been present for about 1 year. Her parents are of normal stature and had normal pubertal development. Her height and weight are both below the 5% for age.
• Physical exam reveals Tanner 1 breast, Tanner 3 pubic hair, and up-curving finger nails.
15. Which of the following hormone levels is most likely to be elevated:

A  testosterone
B  prolactin
C  follicle-stimulating hormone
D  thyroid stimulating hormone
E  17-OH progesterone
16. Which of the following karyotypes is least likely to be found with this patient:

A  45 XO
B  46 XX / 45 XO
C  46 XY / 45 XO
D  46 X, i(Xq)
E  46 X, r(X)
F  46 X,Xq-

17. Which of the following has the highest lifetime risk of developing type 1 diabetes:

A  the father of a 4 year old type 1 diabetic
B  the brother of a 6 year diabetic girl
C  the fraternal twin of a diabetic child
D  the child of a type 1 diabetic father
E  the child of a type 1 diabetic mother
18. Which of the following genotypes is protective against type 1 diabetes:

A  DQA*0501 / DQB*0201  
B  DQA*0301 / DQB*0201  
C  DQA*0102 / DQB*0602

19. An 11 year old boy has had type 1 diabetes for 5 years. He does his own blood glucose test, and gives his own insulin injections. Which of the following changes in his management should NOT occur as he enters puberty:

A  yearly ophthalmology exams  
B  yearly urine albumin:creatinine ratios  
C  increasing his dose of Lantus in the evening  
D  decreasing the total daily insulin dose from about 0.9 units/kg/day to 0.8 units/kg/day.
• A 15 year old boy presents because of bilateral breast enlargement for more than 2 years. The breast are very noticeable, and prevent him from participating in sports. On exam, the testes are 20cc with out nodules. Pubic hair is Tanner 5, and the penis is normal.

20. Which of the following groups of tests might uncover a problem other than adolescent gynecomastia:

A prolactin, FSH, TSH, testosterone
B prolactin, estradiol, 17-OH progesterone
C FSH, total estrogens
D 17-OH progesterone, DHEA-S, prolactin
21. Which is the best treatment option for this adolescent with gynecomastia:

A  depotestosterone injections
B  treatment with letrozole
C  treatment with anastrozole and tamoxifen
D  plastic surgery
E  observation
22. This growth chart most likely represents a boy with:

A  Crohn’s disease
B  Growth hormone deficiency
C  Precocious puberty
D  Prader-Willi syndrome
E  Renal tubular acidosis
23. This growth chart most likely represents a boy with:

A  Growth hormone deficiency  
B  Hypothyroidism  
C  Delayed onset of puberty  
D  Celiac disease
24. This growth chart most likely represents a girl with:

A  Acromegaly  
B  Exogenous obesity  
C  Precocious puberty  
D  Cushing’s disease  
E  Hypothyroidism
25. This growth chart most likely represents a boy with:

A. Cushing’s disease
B. Growth hormone deficiency
C. Precocious puberty
D. Renal tubular acidosis
26. This growth chart most likely represents a boy with:

A  Crohn’s disease
B  Growth hormone deficiency
C  Precocious puberty
D  Prader-Willi syndrome
E  Renal tubular acidosis

Question 27
This boy has:

A  just broken a neighbor’s window with his baseball
B  just seen his first Playboy centerfold
C  just watched the movie “Halloween”
D  Graves’ eye disease
28. This adolescent should be screened with which blood test on a yearly basis?

A   TSH  
B   Transglutaminase IgA  
C   Random glucose  
D   A&B  
E   A&C

29. A term infant is born with ambiguous genitalia. No testicles are palpable in the genital area. A pelvic ultrasound reveals a uterus. Which of the following is most likely to be elevated on the initial blood tests.

A. 11-deoxycortisol  
B. Deoxycorticosterone  
C. 17-hydroxyprogesterone  
D. Cortisol  
E. Sodium
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