



College of Applied Medical Science

Second Semester 1435/1436

Community Health Department

Midterm exam

Clinical Nutrition Program

Metabolic Integration and Disorders

CHS 344

Student Name: _____

Student Number: _____

Time: 1 hour 15 minutes.

Sections	Total possible score	Student's score
1	14	
2	6	
Total	20	

Section 1. List your full answer, using the available space only. (14 points)

1. Give two examples of disorders that involve energy metabolism.

Glycogen storage disease and Fatty acid disorders such as MCAD

2. We want newborn screening tests to be highly sensitive and specific, but the higher the sensitivity of a test the lower its specificity. What does this mean?

This means the more sensitive the test in detecting any abnormality in the blood levels will lead to reducing any false negative cases, this leads to less specificity in actually detecting only the affected patients, in turn this could increase false positive cases (lower specificity).

3. What are the enzymes deficient in the disorders of galactose metabolism?

Galactose-1-phosphate uridyl transferase (GALT), galactokinase (GALK), and galactose epimerase (GALE).

4. Galactosemia patients need the supplementation of Calcium

because it is deficient in their diet.

5. Reducing the load on the affected pathway by dietary restriction is a method of treatment in which disorder. Give one example: ___PKU, MSUD, tyrosinaemia___.

6. You have a newborn patient newly diagnosed with GSD I.

6a. What is the main goal of this dietary treatment?

To prevent hypoglycemia.

6b. What should the parents do in terms of diet during infections and illness? WHY?

Increase glucose supply. Because during infections, a frequent supply of exogenous glucose must be maintained. Furthermore, glucose metabolism is increased with fever.

Extra details: Replacement of meals and snacks by glucose polymer drinks is often needed. Nasogastric drip feeding 24 h a day may be necessary. If this is not tolerated, a hospital admission is indicated for intravenous therapy.

Section 2. Multiple choice questions, choose the best suitable answer. (1 point each)

1. Suha is a 9 year old patient with GSD Ib, What is the best option for her treatment to ensure metabolic control through the night:

- A. Frequent carbohydrate-enriched meals during the night.
- B. Use gastrostomy tube for continuous nocturnal gastric drip feeding.
- C. Uncooked cornstarch every 4 hours.**
- D. Nasogastric continuous nocturnal drip feeding.

2. For Suha and other GSD patients, carbohydrate metabolism is increased, therefore they need an adequate supply of:

- A. Vitamin D.
- B. Vitamin B1.**
- C. Vitamin C.
- D. Calcium.

3. The deficiency of which enzyme causes GSD 1b:

- A. Glucose-6-phosphate translocase.**
- B. Galactokinase (GALK).
- C. Glucose-6-phosphatase.
- D. Uridine diphosphate galactose4-epimerase (GALE).

4. In classic galactosemia, the renal and hepatic disturbances are probably related to the accumulation of:

A. Galactose-1-phosphate

B. UDPgalactose

C. Galactonate

D. Glucose

5. Taking a blood sample for newborn screening in which following period could lead to a false negative result:

A. One hour after birth.

B. One day after birth

C. One month after birth.

D. One year after birth.

6. Liver glycogenesis in general presents with this group of symptoms during infancy:

A. Hepatomegaly, delayed growth, and hyperglycemia.

B. Hypoglycemia, stunted growth, and hepatomegaly.

C. Liver cirrhosis, myopathy, and hypoglycemia.

D. Myopathy, hyperglycemia, and hyperlactatemia.