



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.

\_\_\_\_\_ and \_\_\_\_\_

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?

\_\_\_\_\_

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

\_\_\_\_\_

\_\_\_\_\_

4. Galactosemia patients need the supplementation of \_\_\_\_\_

because \_\_\_\_\_.

5. Reducing the load on the affected pathway by dietary restriction is a method of treatment in which disorder. Give one example: \_\_\_\_\_.

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

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

\_\_\_\_\_

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

\_\_\_\_\_

\_\_\_\_\_

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.