1. True statements pertaining to the assessment of the nutritional status of children include:
A. A simple means of assessing body fat stores is by measuring the triceps skinfold thickness.
B. Clinical signs of undernutrition usually precede biochemical abnormalities.
C. A sensitive index of lean body mass is the determination of the upper arm muscle circumference.
D. A deficit in height-for-age suggests chronic undernutrition.
E. A weight-for-height deficit suggests acute undernutrition.

2. Children with severe protein-calorie malnutrition typically have which of the following immunologic deficiencies:
A. Hypogammaglobulinemia.
B. Depressed cell-mediated immunity.
C. Decreased phagocytic activity of polymorphonuclear leukocytes.
D. Decreased total hemolytic complement activity.
E. Decreased total peripheral lymphocyte count.

3. Clinical signs associated with vitamin C deficiency include:
A. Nasolabial seborrhea.
B. Swollen, bleeding gums.
C. Epiphyseal enlargement.
D. Ecchymoses.
E. Cardiac enlargement.

4. In a neonate with hyperammonemia, which of the following would suggest the diagnosis of transient hyperammonemia of prematurity?
A. Gestational age less than 36 weeks.
B. History of a similar disease in a sibling.
C. Early onset of respiratory distress.
D. Severe perinatal asphyxia.
E. Specific histopathologic findings on liver biopsy.

5. Typical findings in a neonate with a urea cycle enzyme deficiency include:
A. Increased concentration of plasma ammonia.
B. Elevated value for anion gap.
C. Vomiting.
D. Seizures.
E. Stupor.

6. An immediate goal in the treatment of neonatal hyperammonemia is the rapid removal of ammonia. The single most effective means of doing this is:
A. Exchange transfusion.
B. Peritoneal dialysis.
C. Hemodialysis.
D. Intravenous sodium benzotate.
E. Protein-free diet.

7. True statements pertaining to neonatal forms of urea cycle disorders include:
A. Some forms can be diagnosed in utero.
B. Most forms are transmitted as X-linked recessives.
C. Carriers for some of the disorders can be identified.
D. Despite treatment, some forms are almost always lethal.
E. Most forms commonly present in the first two days of life.

8. The neurologic and developmental outcomes of the neonatal forms of urea cycle disorders appear to be related to:
A. Early aggressive treatment.
B. Magnitude of hyperammonemia.
C. Duration of coma.
D. Rate of neurologic improvement.
E. Specific enzyme deficiency.

9. A 4-year-old girl presents with a life-long history of urinary dribbling. She voids regularly and has a good urinary stream. She has no problems with bowel control. Physical examination is unremarkable except that the vulvar area is damp and smells of urine. Of the following, the single most likely diagnosis is:
A. Spinal dysraphism.
B. Sacral agenesis.
C. Posterior urethral valves.
D. Ectopic ureter.
E. Urachal anomaly.

10. Among the following, which is the most definitive test in the diagnosis of Wilson disease?
A. Serum ceruloplasmin.
B. 24-hour urine copper.
C. Serum copper.
D. Hepatic copper.
E. Liver histology.

11. True statements pertaining to Wilson disease include:
A. Kayser-Fleischer rings are pathognomonic.
B. There is autosomal recessive inheritance.
C. Abrupt increases in serum copper levels cause hemolysis.
D. Heterozygotes have no biochemical manifestations of the disease.
E. Symptoms often start in early childhood.

12. Which of the following should be used in the treatment of most patients with Wilson disease?
A. Penicillamine.
B. Pyridoxine.
C. Triethylene tetramine 2HCl.
D. BAL.
E. Diet limiting copper-rich foods.

13. True statements about the neuropsychiatric disorder associated with Wilson disease include:
A. It usually occurs in a younger age group than the liver disease.
B. Improvement in neurologic symptoms occurs with appropriate therapy.
C. Subtle personality changes may occur.
D. Parkinsonian symptoms are typical.
E. Patients are commonly sent to a psychiatrist initially.