

3.4.2

## BLOCK 3.4: 'Life Cycle II' International Bachelor Medicine Groningen (IBMG)

**Date:** 27 June 2013  
**Location:** Exam Hal  
**Time:** 18.30 – 21.15  
**Examiner:** Drs. M.D. Talsma



This assessment consists of two parts (to be filled in on 2 separate UOCG forms):

- (a) a **closed-book** part, from **18.30-19.15**, consisting of – 43 items
- (b) an **open-book** part, from **19.15-21.15**, consisting of – 44 items

The total number of pages is 13 (front page excluded).

### **Urgent advice about the open book part of this assessment**

First complete all items; do not start looking up information immediately. Looking up information is very time consuming which may result in not completing the assessment on time. Look up answers you have left blank and, if you have enough time, answers you are not sure about.

The following (study) material is allowed during the open-book test

- British National Formulary
- Beckmann: Obstetrics and Gynecology
- Burkitt: Essential Surgery
- Singer: The Cambridge Textbook of Bioethics
- Jorde: Medical Genetics
- Kliegman: Nelson Essentials of Pediatrics
- Rang: Rang & Dale's Pharmacology
- Kumar: Clinical Medicine
- Kumar (Robbins): Basic Pathology
- Readers block 3.4
- language dictionaries (no medical dictionaries)

You are not allowed to use other material (including copies of other books than the obligatory books, old examination papers) or electronic aids (e.g. mobile phone, PDA).

### Answer form instructions

- First of all, read the instructions on the UOCG form itself how to complete the UOCG form.
- Erasing is NOT allowed (erasing may cause read-in errors).
- In preference use an HB pencil to complete the UOCG form.
- Fill in your **name** and **student number**; mark the examination with the codes 'closed book part 3.4.2' and 'open book part 3.4.2' respectively.
- First answer the questions on the exam paper by circling the most appropriate answer.
- Then fill in your answers on one of the attached UOCG forms. Double check afterwards!
- The answer sheet for the closed-book part will be collected after **45 minutes**. You are allowed to begin with the open book part as soon as you have finished the closed-book part. However, you are **not** allowed to use your study material **before** the invigilator has given the signal to do so.
- If you have a question about these instructions ask the invigilator or lecturer.

### After the assessment

- Hand in your second UOCG form and/or evaluation forms to one of the invigilators.
- The answers will be published on Nestor asap.
- The results will be published on Nestor within 10 working days.

**GOOD LUCK!**

## Closed Book part

1. In which body fluid is PSA (prostate-specific antigen) measured?
  - a. urine
  - b. saliva
  - c. serum
  - d. sperm

*A 63-year-old man has metastasized prostate cancer.*

2. What is the drug therapy of choice?
  - a. chemotherapy
  - b. endocrine therapy
  - c. prednisone in conjunction with an opiate

*A 54-year-old woman had her last menstruation at the age of 46. She still suffers from regular hot flushes.*

3. This is a reason for further investigation.
  - a. correct
  - b. incorrect

*An 83-year-old man with advanced COPD, which has deteriorated over the last six months, presents to the Emergency Department with severe breathing difficulty, cough and fever. The doctors suspect a bacterial chest infection. Because of the patient's old age and his severe and deteriorating COPD, the doctors ask the patient whether he would like to be treated for the infection or not. After careful consideration, the patient and his family decide that they only want symptomatic relief and no curative treatment (i.e. antibiotics) for the chest infection. The patient dies the next day.*

4. Would this be considered a death of natural causes?
  - a. yes
  - b. no

*Urge incontinence has various characteristic features. One of these is:*

5. painful micturition
  - a. correct
  - b. incorrect

*The doctor is convinced that the patient's request for death is supported by his/her next of kin.*

6. Is this one of the due cautions criteria for euthanasia in The Netherlands?
  - a. yes
  - b. no

*A two-year-old boy has microcytic anaemia. His serum iron is low; his iron-binding capacity and serum ferritin are normal.*

7. What is the most likely diagnosis?
  - a. anaemia of chronic disease
  - b. thalassaemia
  - c. iron deficiency anaemia

*A 40-year-old woman suffers from severe urinary urgency with inability to hold her urine. Her menstruation is still regular.*

8. What is the treatment of choice?
  - a. colposuspension surgery
  - b. pelvic floor physiotherapy
  - c. oral anticholinergic agent
  - d. vaginal oestriol cream

*A 45-year-old man dies as a result of an accident. He has given consent for organ and tissue donation. His wife is against this.*

9. She has the right of veto under Dutch law.
- a. correct
  - b. incorrect

*During a liver transplant operation the heart-lung machine is used to pump blood from the lower half of the body to the heart. Unfortunately, when the machine is connected a large air embolism develops and the patient dies.*

10. This is a case of death from natural causes.
- a. correct
  - b. incorrect

11. Brain death may only be diagnosed in a case of fatal brain injury the cause of which is known.
- a. correct
  - b. incorrect

*Achondroplasia is an abnormality that is due to a de novo mutation in 90% of cases.*

12. When two children in a family with parents of normal stature are born with achondroplasia, this is likely to be a case of:
- a. heteroplasmy
  - b. meiotic non-disjunction
  - c. genotypic mosaicism
  - d. genomic imprinting

13. Prostate cancer metastases are most commonly found in the:
- a. lungs
  - b. liver
  - c. skeleton

*A two-year-old girl is suffering from fatigue and is conspicuously pale. Physical examination reveals no abnormalities other than moderate injection of the mucosa.*

*Lab tests show normochromic normocytic anaemia. The HbF is low, fetal antigen I is absent.*

14. What is the treatment of choice?
- a. treat with androgens
  - b. bone marrow transplant
  - c. do not treat
  - d. treat with prednisolone
15. What is the deciding factor in the prognosis of **non**-metastasized prostate cancer?
- a. degree of local spread
  - b. degree of malignancy-
  - c. PSA level

16. Which of the following findings is the most reliable indication of death?
- a. areflexia
  - b. hypothermia
  - c. cardiac arrest
  - d. pupil deformability

*An 83-year-old woman has advanced dementia. She lives in a nursing home. She needs to be referred to a hospital as she has sustained a hip fracture after a fall.*

*When the dementia started a few years ago both the woman and her husband designated a nephew as her representative, but the husband is still in good health and of sound mind.*

17. Who should be asked for consent for her hospitalization under Dutch legal jurisdiction?
- a. the nephew
  - b. her husband
  - c. no consent is required

18. Biopsies of the prostate to rule out cancer are taken via the:
- urethra
  - perineum
  - rectum
  - abdomen

*A six-year-old boy is seen for conspicuous pallor and anuria. A few days ago he was vomiting and had bloody diarrhoea. Lab tests reveal severe anaemia and markedly increased urea and creatinine.*

19. What is the most likely diagnosis?
- haemolytic uremic syndrome
  - renal-vein thrombosis
  - acute glomerulonephritis

*A boy of foreign extraction has spells of yellow pallor. The mother has noticed that this occurs particularly a few days after he has been ill with fever or has eaten broad beans. At these times the blood picture shows red cells that 'look as if a bite has been taken out of them'.*

20. What is the most likely diagnosis?
- congenital spherocytosis
  - $\beta$ -thalassaemia
  - G6PD deficiency
  - sickle cell anaemia

21. Ability to make decisions depends on the nature of the decision and the context.
- correct
  - incorrect

*Coagulation factor deficiencies usually cause an increased bleeding tendency.*

22. Deficiency of which of the following factors causes the most severe bleeds?
- Factor 9
  - Factor 11
  - Factor 12

*Virtually all types of childhood cancer are treated with chemotherapy.*

23. Why?
- most tumours in childhood are not responsive to radiotherapy
  - in childhood cancer there is a major risk of micrometastases already being present at the time of diagnosis
  - children suffer less from the side effects of chemotherapy than adults

*A neonate has retrognathia with a protruding occiput. His heels protrude at the rear, making the feet look like rocking chairs. When he makes a fist, the ring finger and little finger bend over the other fingers.*

24. What is the most likely cause of this clinical picture?
- chromosome 15 duplication
  - deletion of the short arm of chromosome 7
  - trisomy of chromosome 13
  - trisomy of chromosome 18

*A two-year-old girl is seen for sudden-onset left-sided hemiplegia. She has a history of episodes of frequent vomiting and has had three seizures. The girl's sister has cardiomyopathy and her mother is deaf.*

25. What type of hereditary disorder is most likely here?
- autosomal dominant
  - autosomal recessive
  - sex-linked dominant
  - mitochondrial

*A surgeon at the Emergency Department is treating a five-year-old boy who has been badly beaten up by his ten-year-old cousin. The surgeon would like to report this to the AMK (Child Abuse Advice and Reporting Centre) in line with the Child Abuse Reporting Code in The Netherlands.*

26. To do this he requires the permission of the five-year-old boy's parents.
- a. correct
  - b. incorrect

*A man of Egyptian extraction has G6PD deficiency.*

27. What is the probability that his sons will be heterozygous for G6PD?
- a. 0%
  - b. 25%
  - c. 50%

28. Which tier of government in The Netherlands is responsible for organizing youth health care?

- a. the municipal authority
- b. the provincial authority
- c. central government

29. Pulmonary hypoplasia in Potter's syndrome is an example of:

- a. anticipation
- b. association
- c. sequence

30. The national vaccination programme in The Netherlands includes vaccinations to prevent cancer.

- a. correct
- b. incorrect

*An 11-year-old boy has regular tachycardia of 260 bpm with broad QRS complexes. He is very pale, his blood pressure is 55/30 and his capillary refill is prolonged.*

31. What is the treatment of choice?
- a. intravenous adenosine
  - b. intravenous amiodarone
  - c. asynchronous electrical cardioversion
  - d. synchronous electrical cardioversion

*A two-week old baby has a transcutaneously measured saturation of 88%. Physical examination reveals increased right atrial impulse, widely split second heart sound, grade 3 systolic ejection murmur at 2 left and mid-diastolic murmur at 4-5 left. The chest X-ray shows a large heart and increased pulmonary vascular markings.*

32. The diastolic murmur in this baby is most likely to be due to relative stenosis of the:

- a. aortic valve
- b. mitral valve
- c. pulmonary valve
- d. tricuspid valve

*A two-year-old toddler suffers from shortness of breath in the evening. His temperature is 38.3 °C. On physical examination he is found to be hoarse with a barking cough. There is moderate dyspnoea with audible inspiration.*

33. What is the treatment of choice for this clinical picture?

- a. intravenous amoxicillin
- b. oral dexamethasone
- c. inhaled salbutamol

34. In patients suffering from otitis media the onset of symptoms is gradual.

- a. correct
- b. incorrect

35. Which of the following symptoms is most commonly seen first in distributive shock?
- anuria
  - hypotension
  - cold extremities
  - tachycardia

*A new-born baby girl dribbles and blows bubbles. There was a strikingly large amount of amniotic fluid.*

36. What is the test of choice to diagnose the problem?
- abdominal ultrasound scan
  - chest X-ray with gastric catheter at maximum reach
  - barium gastric X-ray
  - oesophagogastroscopy

*A seven-month-old baby has bloody mucus in his stools. He is vomiting and the abdomen is distended and tender.*

37. Which of the following is the most likely diagnosis?
- anal fissure
  - invagination
  - cow's milk allergy
  - gastric ulcer

*A 14-year-old girl has liver failure as a result of taking an overdose of paracetamol. She is confused and her speech is incoherent. Her breathing is rapid and deep. Neurological tests show increased tendon reflexes.*

38. What is the treatment of choice?
- intravenous glucose
  - lactulose by gastric catheter
  - intravenous diuretics
  - intravenous fresh frozen plasma

*A six-year-old girl from the biblebelt area has had a cough, red eyes and a cold in the nose for several days. A few greyish-white spots are found in her mouth, in the cheek.*

39. These symptoms are most likely to be caused by:
- human herpes virus type 6
  - measles virus
  - parvovirus B19
  - rubella virus

*A six-year-old boy suffers from epilepsy. He is markedly retarded. He has some flesh-coloured papules on and around his nose.*

40. What is the most likely cause of his clinical picture?
- a deletion on chromosome 22
  - mutation in one of the genes TSC1/TSC2
  - congenital herpes simplex virus infection
  - mutation in the NF2 gene

41. Somatostatin inhibits the release of growth hormone.
- correct
  - incorrect

*The Dutch Medical Treatment Agreement Act codifies the basic rights and obligations in the physician-patient relationship. (Questions 42-43)*

42. One of the rights of patients of 16 years and older is that they may express their wishes in an advanced directive.
- true
  - false

43. A patient below the age of 12 is **not** required to consent to medical treatment.
- a. true
  - b. false

**End of Closed Book part**

**Continue with Open Book part on page 7**

## Open Book part

*A 55-year-old woman has difficulty holding her stools. When she feels the urge, she has to go to the toilet within three to five minutes, otherwise she is incontinent.*

1. Her problem is most likely to be with the:
  - a. pudendal nerve
  - b. external anal sphincter
  - c. puborectalis muscle

*A ten-year-old boy has some painless swollen glands in the left axilla and left groin. Four months ago he weighed 40 kg but he has lost weight and now weighs only 35.5 kg. Biopsies from an axillary and a groin gland show nodular glomerulosclerosis with Reed-Sternberg cells. A diagnosis is made.*

2. What stage has the disease reached, given the above data?
  - a. IIA
  - b. IIIA
  - c. IIIB
  - d. IVB

*An eight-year-old boy is developmentally retarded (IQ 55), although it is noticeable that his social skills are relatively well developed. He talks easily and likes music. He has typical facial characteristics with a sunken bridge of the nose, curved eyebrows and a blue iris with scattered pigmentation in both eyes. He also has a heart defect, supraaortic stenosis, and probably stenoses in both pulmonary arteries.*

3. What anomaly is most likely in his case?
  - a. acidosis with elevated propionic acid
  - b. elevated urinary pipercolic acid
  - c. chromosome 7q11 deletion
  - d. chromosome 5p deletion

*A 28-year-old woman has a regular menstrual cycle but with vaginal bleeding between periods. She has no fever or lower abdominal pain. She is found to have an infection.*

4. What is the most likely pathogen?
  - a. Chlamydia trachomatis
  - b. Neisseria gonorrhoeae
  - c. Treponema pallidum
5. A male and female cousin are married. In comparison with those of non-consanguineous marriages, their children are at increased risk of:
  - a. autosomal dominant disease
  - b. autosomal recessive disease
  - c. X-linked dominant disease

*A 54-year-old woman has broken her right wrist.*

6. Does a bone mineral density test need to be carried out according to the ACOG?
  - a. yes
  - b. no

*Postmenopausal women are sometimes prescribed hormone therapy to prevent osteoporosis and cardiovascular disease.*

7. This therapy results in a reduction in the number of cerebral infarctions.
  - a. correct
  - b. incorrect

An 85-year-old woman has atrial fibrillation, for which she is taking acenocoumarol (an anticoagulant). She is given a temporary course of ibuprofen (an NSAID) for arthritis. A week later she is admitted to hospital with gastric bleeding.

8. What is the most likely cause of the gastric bleeding?
- naproxen reduces the metabolism of acenocoumarol, thus increasing the acenocoumarol level
  - in elderly patients in particular, naproxen damages the gastric mucosa, thus increasing the risk of gastric bleeding
  - naproxen causes a decrease in renal function, resulting in a decrease in acenocoumarol excretion, thus increasing the risk of bleeding

A 75-year-old man is treated with atorvastatin (a cholesterol-reducing drug). While being treated for a respiratory infection with erythromycin he complains of muscle pain, a known side effect of atorvastatin.

9. What is the most likely mechanism causing these symptoms?
- pharmacokinetic interaction between erythromycin and atorvastatin
  - pharmacodynamic interaction between erythromycin and atorvastatin
  - the respiratory infection

Elderly men sometimes develop gynaecomastia.

10. What is the most common cause?
- increased production of sex hormones in the adrenal gland
  - increased production of sex hormones in the testicles
  - changed metabolism of sex hormones in the liver
  - increased sensitivity of breast tissue to sex hormones

Although glomerular filtration rate and renal perfusion decline in the elderly, their serum creatinine is often normal.

11. What causes this?
- change in liver function in the elderly
  - reduced muscle mass in the elderly
  - active tubular secretion of creatinine in the elderly
12. Elderly people have a lower level of scavenging enzymes (molecules that protect cells against free oxygen radicals) than younger people.
- correct
  - incorrect

Judging the efficacy of treatments is part of a physician's expertise.

13. In The Netherlands, who decides whether a treatment is disproportionate?
- the physician
  - the physician and the patient
  - the patient
  - the patient and his/her family

A 46-year-old woman with diabetes mellitus and epilepsy has died suddenly. Her brother, who found the body, was not present when she died. Physical examination does not yield any explanation for her death, but there are no indications of death from other than natural causes. The duty GP issues a certificate of death from natural causes.

14. This is permissible in The Netherlands because the diseases that the woman was suffering from provide a likely explanation for her death.
- correct
  - incorrect

A man is found dead. There is lividity that disappears when pressed at the edges but not in the centre. Rigor mortis is complete. The ambient temperature is 16°C.

15. How long ago is the man most likely to have died?
- 6 hours
  - 18 hours
  - 36 hours

A 58-year-old woman complains of painful frequent micturition and urine loss when she coughs.

16. What is the most likely cause of these symptoms?

- a. atrophy of the urethral epithelium
- b. enterocele
- c. rectocele

An 80-year-old woman is being treated with Sinthrome (a coumarin derivative). A natural healer prescribes her a remedy containing St John's wort.

17. What effect is the St John's wort most likely to have on the amount of Sinthrome needed for adequate anticoagulation?

- a. she will need to take more Sinthrome to achieve the desired effect
- b. she will need to take less Sinthrome to achieve the desired effect
- c. the Sinthrome dosage does not need to be changed

A mother has stopped giving her six-month old baby, who had been developing well, night feeds because she gradually found she could not cope. A few days after she stops breastfeeding, the baby is found more or less comatose in bed in the morning. Physical examination reveals a markedly enlarged liver, which feels soft to the touch. The lab tests show increased lactate, glucose measurement was unsuccessful, and there is uncompensated metabolic acidosis.

18. What disease is most likely?

- a. type I tyrosinaemia
- b. type I glycogen storage disease
- c. organic acidaemia
- d. galactosaemia

An eight-year-old girl has severe epilepsy, for which she is being treated with phenytoin and valproate. She is seen for fatigue. On physical examination she is found to be conspicuously pale. Blood tests show an Hb of 4.5 mmol/l (7.2 g/dl), a leukocyte count of  $9.6 \times 10^9/l$  and a thrombocyte count of  $213 \times 10^9/l$ . The red blood cells are conspicuously large.

19. What is the most likely cause of the anaemia?

- a. myelodysplasia
- b. vitamin B12 deficiency
- c. folic acid deficiency
- d. Diamond-Blackfan syndrome

A neonate has an Hb of 6 mmol/l. A blood smear shows conspicuously small red blood cells with spherocytes, elliptocytes and fragmented erythrocytes.

20. The blood cell abnormalities are most likely to be caused by a change in:

- a. the haemoglobin alpha chain
- b. the haemoglobin beta chain
- c. NADPH formation
- d. spectrin

A six-year-old boy is seen for bleeding in the skin and mucosa. Tests show a prothrombin time (PT) of 11 seconds, an activated partial thromboplastin time (aPTT) of 32 seconds and a bleeding time of 12 minutes. The thrombocyte count is  $180 \times 10^9/l$ . The ristocetin cofactor assay is normal.

21. What is the most likely cause of the bleeding tendency?

- a. parvovirus B19 infection
- b. Factor 8 deficiency
- c. von Willebrand factor deficiency
- d. glycoprotein IIb-IIIa deficiency

*A five-year-old boy has eight coffee-coloured patches 2-8 cm in diameter on his skin. He also has multiple small brown nodules in both eyes. He is now being seen for a progressive swelling around the eye with protuberance of the eye. A malignant condition is suspected.*

22. What malignant condition is most likely here?

- a. Ewing's sarcoma
- b. Hodgkin's
- c. rhabdomyosarcoma
- d. neuroblastoma

*A neonate has skin bleeding and necrosis of the fingers and toes. An ultrasound scan shows thrombosis of the portal vein and an aortic thrombus. An infection is ruled out. A hereditary disease is suspected.*

23. What type of hereditary disease is most likely?

- a. autosomal dominant
- b. autosomal recessive
- c. X-linked recessive

*A twelve-year-old boy has an Hb of 4 mmol/l. Spur cells are found in the blood smear.*

24. What is the most likely cause of the anaemia?

- a. autoimmune haemolysis
- b. vitamin E deficiency
- c. elevated serum cholesterol
- d. disseminated intravascular coagulation

*A six-year-old boy has had several spells of severe pain in his legs. He has also had a cerebral infarction with paralysis of the left half of the body. At a check-up his Hb was 4 mmol/l.*

25. What is the treatment of choice for this boy to prevent new problems?

- a. oral coumarin
- b. oral hydroxyurea
- c. fractionated subcutaneous heparin

*A six-day-old neonate has elevated blood guanidinoacetate.*

26. He is most likely to have a disorder of:

- a. purine synthesis
- b. carbohydrate metabolism
- c. creatine synthesis
- d. fatty acid oxidation

*An eight-week-old boy is seen for twitching in the shoulders, arms and legs. He has a striking appearance with a high forehead and flat orbital rims and large fontanelles. He weighs 3.9 kg (birth weight 3.4 kg). There is marked hypotonia.*

27. What is the best test to reach a diagnosis?

- a. blood leucine
- b. urinary propionic acid
- c. urinary pipecolic acid
- d. urinary galactose

*A three-month-old girl is 56 cm tall, weighs 3.8 kg and has a cranial circumference of 35 cm. She also has hypotonia, hypertelorism, low ear position and laryngeal hypoplasia.*

28. What is the most likely cause of this clinical picture?

- a. deletion of part of chromosome 5
- b. organic acidaemia
- c. a lysosomal disease
- d. trisomy of chromosome 9

*A two-year-old boy is seen for growth retardation, in particular insufficient weight gain. His hearing is poor. Blood tests show a pH of 7.22 with an anion gap of 8. The urinary pH is 7.*

29. What is the next test of choice?
- blood lactate level
  - urinary calcium level
  - ketone bodies in the blood and urine

*A 40-year-old woman has undergone a hysterectomy for cervical carcinoma. She is now suffering from loss of libido.*

30. What is the treatment of choice?
- a progestogen
  - an oestrogen
  - tibolone

*The possibility of sexual abuse in a two-year-old girl is being considered. A vaginal culture shows chlamydial growth.*

31. If chlamydial growth is found, the physician should report child abuse to the AMK (*Child Abuse Advice and Reporting Centre*).
- correct
  - incorrect

*The Dutch Professions in Health Care Act recognizes professions that are subject to a system of constitutive registration.*

32. Is pharmacy one of these professions?
- yes
  - no

*A 16-year-old boy is unconscious after a car accident and needs mechanical ventilation. After consulting a neurologist, who states that neurological improvement is unlikely, the attending Emergency Department (ED) physician decides in consultation with the boy's parent, that further treatment is futile and discontinues mechanical ventilation. To reduce suffering, the ED physician administers morphine.*

33. Under Dutch criminal law, is the ED physician likely to be prosecuted for unjustified euthanasia?
- true
  - false

*Physicians may be confronted with three types of liability: civil, criminal and disciplinary liability.*

34. A physician who, because of inadequate knowledge of the content of the X-ray files, amputates the wrong leg of a patient is likely to be held accountable in all three spheres of liability.
- true
  - false

*A six-month-old girl has a heart defect that causes slight cyanosis at rest. If she becomes agitated she first cries inconsolably, then develops deep cyanosis and tachypnoea and eventually loses consciousness.*

35. How can these attacks be dealt with? By:
- increasing pulmonary vascular resistance
  - decreasing pulmonary vascular resistance
  - increasing systemic arterial resistance
  - decreasing systemic arterial resistance

*An eight-year-old boy is found unconscious, lying on his back with legs extended, and with flexed, supinated arms and clenched fists.*

36. This posture points to uninhibited activity of the:
- cortex
  - basal ganglia
  - mesencephalon
  - pons

An eight-year-old boy is being given medication and parenteral nutrition through a line from the jugular vein into the right atrium. He develops fever (39.2°C) and looks ill. A bacterial infection is suspected.

37. What is the most likely pathogen?
- coagulase-negative Staphylococcus
  - E. coli
  - Staphylococcus aureus
  - Pseudomonas aeruginosa

A four-year-old boy is seen for kidney stones. He also has growth retardation and deafness. Blood tests show reduced serum potassium and a pH of 7.25 with sodium bicarbonate of 8 mmol/l. A urine test shows an elevated calcium level.

38. This is most likely a case of:
- autosomal dominant hereditary disease
  - autosomal recessive hereditary disease
  - X-linked recessive disease

A two-year-old girl has oliguria of 0.5 ml/kg/h. Urine osmolality is 700 mOsm/l, urine sodium level is 10 mmol/l.

39. What is the most likely location of the cause of the oliguria?
- prerenal
  - renal

An 11-year-old boy has recurring nasal cavity infections and chronic productive cough and shortness of breath, sometimes combined with fever.

On physical examination the cardiac impulse is felt in the right medioclavicular line. The NO level in the nose is markedly reduced.

40. How high is the risk of this boy later in life having a child with the same condition?
- 0%
  - 25%
  - 50%
  - all the sons, none of the daughters

A one-year-old girl has marked developmental retardation and deafness. A cranial X-ray shows periventricular calcification.

41. What is the most likely cause of the abnormalities?
- congenital toxoplasmosis
  - a deletion on chromosome 15
  - CMV infection
  - trisomy 13

A four-year-old boy has a bout of chickenpox. A week later it is noticeable that his gait is unsteady and he often falls. He finds it difficult to talk and is somewhat drowsy. A lumbar puncture is performed.

42. What is most likely to be found in the cerebrospinal fluid?
- increased segmented leukocyte count
  - increased lymphocyte count
  - markedly increased protein level and relatively low glucose level
  - increased erythrocyte count

43. Reduced reactivity of the autonomous nervous system is found in children with:

- ADHD
- PDD-NOS
- conduct disorder
- Asperger's syndrome

A young male infant is diagnosed with hypocortisolism. His blood pressure is raised, his genitalia are normal.

44. What is the most likely diagnosis?

- a. 21-hydroxylase deficiency
- b. 11-hydroxylase deficiency
- c. 3-beta hydroxysteroid dehydrogenase deficiency
- d. congenital adrenal hypoplasia

**End of assessment**