ISSN 1998-4235 (Print) • ISSN 2522-1183 (Online)
Український неврологічний журнал.— 2018.— № 1.— С. 64—71.
УДК 616.8(079.1)(4)

At the suggestion of MD, PhD, Professor of Neurology Viktoriia Gryb, the Head of the Department of Neurology and Neurosurgery at the Ivano-Frankivsk Medical University, and the Assistant Professor of Neurosurgery, neurosurgeon Vyacheslav Botyev, the selection of Multiple Choice Question (MCQ) and Extended Matching Question (EMQ) are published.

European Board Examination in Neurology
Variants of questions with answers. Examples 3

EXTENDED MATCHING QUESTIONS

Each option may be used once, more than once or not at all.

1. HEADACHE

Each of the patients below has headache. Choose the best diagnosis from the options given for each of the clinical scenarios below.

A. Sinusitis
B. Migraine
C. Iatrogenic
D. Meningitis
E. Brain tumor
F. Cluster headache
G. Temporal arteritis
H. Tension headache
I. Trigeminal neuralgia
J. Subarachnoid hemorrhage

1.1. A 22-year-old woman goes to see her GP because of a unilateral headache with nausea and vomiting. She is photophobic and says that she can see flashing and shimmering lights when she closes her eyes. Her pulse is 70 beats/min, temperature 36.7 °C with neurological examination entirely unremarkable.

1.2. A 73-year-old man goes to see his GP with a right-sided headache. He says the pain gets worse when he eats.

1.3. A 53-year-old woman complains of short attacks of what she says feels like an electric shock over her left cheek. She says that the pain is intolerable and can be triggered by brushing her teeth.

1.4. A 33-year-old man goes to see his doctor with a severe headache that affects the left side of his head and is associated with a red eye, ptosis and a running left nostril. He says that he has the same symptoms every March.

1.5. A 29-year-old man is brought to see his GP by his wife. She is worried as he is having headaches every morning when he wakes up. They seem to get better spontaneously through the course of the day. While examining the patient, the GP asks the man to touch his toes, which seems to make the headache worse.

2. NEUROANATOMY

Select the area of the brain most likely to be responsible for the defects described below.

A. Pons
B. Pituitary
C. Cerebellum
D. Pineal gland
E. Broca's area
F. Olfactory bulb
G. Temporal lobe
H. Substantia nigra
I. Wernicke's area
J. Aqueduct of Sylvius

2.1. A 76-year-old man is admitted with non-communicating hydrocephalus.

2.2. An 84-year-old woman is admitted after having had a stroke. She can understand the instructions of the doctor but is unable to speak.

2.3. A 61-year-old woman with Parkinson's disease wants to know where in her brain are the defective neurones located.

2.4. A 29-year-old man, who lost his sense of smell after a car accident, wants to know which part of his brain is not working.

2.5. A 59-year-old woman goes to see her GP asking for some sleeping tablets to help her with jet-lag. He patiently explains to her that a part of the brain has a clock to help the body know whether it is day or night.

3. CRANIAL NERVE LESIONS

For each of the patients with a cranial nerve lesion, choose the defective nerve from the *options given below.*

A. II
B. III
C. IV
D. V
E. VI
F. VII
G. VIII
H. IX
I. X
J. XI

3.1. An obese 23-year-old woman goes to see her GP with headache and visual loss that is worse after bending over. The cranial nerve examination reveals that she cannot look tothe right with her right eye.

3.2. An 83-year-old woman presents to the emergency department with a stroke. The doctor touches the back of her throat with a tongue depressor; the patient says that she can feel nothing.

3.3. A 69-year-old man complains that his sense of taste has gone. Careful examination revealsno taste sensation in the anterior two-thirds of his tongue. The posterior one-third is functioning normally.

3.4. An 83-year old man complains of diarrhoea. His GP says that this is the result of one of his nerves being cut when he had an operation for an ulcer 40 years ago.

3.5. A 71-year-old man tells his GP that he has been unable to lift his right shoulder since a stroke 6 months ago.

4. DRUGS USED IN NEUROLOGICAL DISEASE

Select the most appropriate drug for each of the indications below

A. Riluzole
B. Donepezil
C. Fluoxetine
D. Lorazepam
E. Temazepam
F. Ethosuximide
G. Amitriptyline
H. Carbamazepine
I. Methylphenidate
J. Prochlorperazine

4.1. A 31-year-old epileptic woman has been having a witnessed seizure for the past 30 min.The ambulance crew has helpfully sited an intravenous cannula.

4.2. A 22-year-old man with amyotrophic lateral sclerosis.

4.3. A 7-year-old boy with attention-deficit hyperactivity disorder.

4.4. A 9-year-old girl who suddenly seems to stop concentrating in class for 3—4 s before she returns to normal.

4.5. A 43-year-old woman who attends the emergency department with dizziness, tinnitusand vomiting. The doctor diagnoses benign paroxysmal positional vertigo and offers her some medication which he says will help.

5. DRUGS USED IN MOVEMENT DISORDERS

Select the most appropriate drug for each of the indications below

A. Levodopa
B. Selegiline
C. Piracetam
D. Entacapone
E. Cabergoline
F. Propranolol
G. Benserazide
H. Amantadine
I. Apomorphine
J. Tetrabenazine

5.1. A 37-year-old man with Huntington's disease presents to his neurologist with hemiballismus. He has not taken any medication to control his symptoms to date.

5.2. An 81-year-old man reads the patient information leaflet with his L-dopa tablets. It says that there are two drugs in the medication: he wants to know the name of the other one.

5.3. A 29-year-old doctor drops into the neurology clinic to see the consultant. He explains that he has had a tremor all his life but has managed perfectly well until now. However, his consultant, a plastic surgeon, is becoming increasingly frustrated with his suturing abilities.

5.4. A 71-year-old woman with newly diagnosed Parkinson's disease is started on L-dopatherapy. She tolerates this poorly and discontinues the medication. Her GP says that he will give her a different sort of tablet, which he calls a dopamine agonist.

5.5. A 75-year-old man has end-stage Parkinson's disease. His wife is extremely frustratedas he just sits in a chair all day long. He is on a cocktail of oral medications which only slightly improve his symptoms. The neurologist says that he thinks it may be time to move on to a powerful injection.

6. OFFENDING ARTERY

A. Facial artery
B. Basilar arteries
C. Vertebral artery
D. Ophthalmic artery
E. Internal carotid artery
F. Anterior choroid artery
G. Posterior cerebral artery
H. Superior cerebellar artery
I. Anterior communicating artery
J. Posterior communicating artery
K. Anterior inferior cerebellar artery
L. Posterior inferior cerebellar artery
M. M3 portion of middle cerebral artery
N. A1 portion of anterior cerebral artery

For each of the following descriptions, select the most appropriate answers from the list above.
Each answer may be used once, more than once or not at all.

6.1. Glossopharyngeal neuralgia

6.2. Trigeminal neuralgia

6.3. Hemifacial spasm

6.4. Horner's syndrome

6.5. CN III palsy

7. PARANEOPLASTIC DISORDERS

A. Hypercalcemia
B. Myasthenia gravis
C. Stiffman syndrome

D. Limbic encephalitis
E. Motor neuron disease
F. Opsoclonus-myoclonus
G. Guillain-Barré syndrome
H. Dorsal root ganglionopathy
I. Paraproteinemic neuropathy
J. Lambert-Eaton myasthenic syndrome
K. Paraneoplastic cerebellar degeneration

For each of the following descriptions, select the most appropriate answers from the list above.
Each answer may be used once, more than once, or not at all.

7.1. A 67-year-old female has a 2 month history of progressive gait disturbance. On examination, she has dysmetria of the limbs, awide-based, unsteady gait; and hypermetric saccades. A hard, firm breast lump is discovered.

7.2. A 70-year-old male with a history of lungcancer develops nausea and vomiting and then becomes lethargic. On examination, he is lethargic but arousable, disoriented, and inattentive. He is weak proximally and has diminished reflexes.

7.3. A 57-year-old female with a history of smoking has a 3-month history of hip and shoulder weakness. She also complains of xerostomia. There are no sensory symptoms, and she is cognitively intact. On examination, she is orthostatic. There is proximal muscle weakness, but she has increasing muscle strength with repetitiveactivity of her muscles. Eye movements are normal.

7.4. A 65-year-old female develops pain and paresthesias in her feet. On examination, she has stocking distribution sensory loss, and mild distal weakness with areflexia. Serum protein electrophoresis reveals amonoclonal gammopathy, and bone marrow biopsy reveals plasma cell dyscrasia.

8. LEUKODYSTROPHIES

A. CADASIL
B. Krabbe disease
C. Refsum disease
D. Canavan disease
E. Alexander disease
F. Zellweger syndrome
G. Pelizaeus-Merzbacher disease
H. Metachromatic leukodystrophy
I. X-linked adrenoleukodystrophy
J. Cerebrotendinous xanthomatosis
K. Acute disseminated encephalomyelitis

For each of the following descriptions, select the most appropriate answers from the list above.
Each answer may be used once, more than once, or not at all.

8.1. Two brothers, 4 and 6 years of age, but not their 9-year-old sister, exhibit limb ataxia, nystagmus, and learning disability, and haveabnormally low serum cortisol levels.

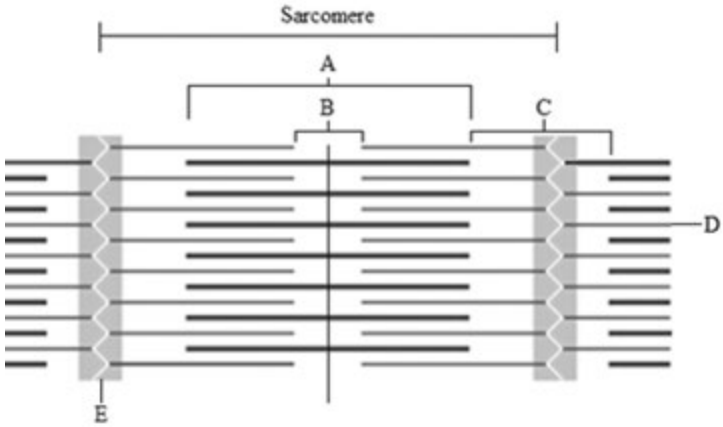
8.2. A 3-month-old boy exhibits nystagmus and limb tremors unassociated with seizures. Over the next few years, he develops opticatrophy, choreoathetotic limb movements, seizures, and gait ataxia. He dies during status epilepticus and at autopsy is found to have widespread myelin breakdown with myelin preservation in islands about the blood vessels. The pathologist diagnoses as udanophilic leukodystrophy to describethe pattern of staining observed on slidesprepared to look for myelin breakdown products.

8.3. A 17-month-old boy had developed normally until approximately 13 months of age, when he began having progressive gait problems. On examination, the patient is spastic, yet nerve conduction studies (NCS) reveal slowed motor and sensory conduction velocities. Cerebrospinal fluid (CSF) protein is elevated. MRI revealswhite matter abnormalities. Leukocyte testing reveals deficient arylsulfatase A activity.

8.4. A 6-month-old child has a rapid regression of psychomotor function and loss of sight. There is increased urinary excretion of N-acetyl-L-aspartic acid.

9. SARCOMERE

Directions: The questions below consist of lettered headings within figure, depicting the sarcomere, followed by a set of numbered items. For each numbered item, select one or more than one heading with which it is closely associated. Each lettered heading may be used once, more than once, or not at all.



9.1. Composed solely of actin filaments

9.2. Shortens during muscle contraction

9.3. H band

9.4. A band

9.5. Z disc

10. TREMOR

A. Palatal
B. Holme's
C. Dystonic
D. Essential
E. Cerebellar
F. Neuropathic
G. Psychogenic
H. Parkinsonian
I. Physiological
J. Drug-induced
K. Primary orthostatic

For each of the following descriptions, select the most appropriate answers from the list above.
Each answer may be used once, more than once or not at all.

10.1. A 40-year-old literary agent has had worsening tremor of the hands. This has been present for 2 years, but has increasingly impaired her work ability because she is frequently required to take her clients to lunch, and she is embarrassed by perinability to eat and drink normally. A glass of wine with the meal typically helps somewhat. On examination, there is amild head tremor, but no rest tremor of the hands. When she holds a pen by the tip at arm's length, however, a coarse tremor is readily apparent. Examinationis otherwise normal.

10.2. A 64-year-old man has noticed dragging of the right leg and tremor and stiffness of the right hand. On examination, he has a tremor of the right hand, which disappears when he reaches to grab a pen. Movements are slower on the right than the left. He has cogwheel rigidity of the right arm.

10.3. A 56-year-old presents 4 months after athalamic stroke with a left arm 3 Hz action tremor with a «wing-beating» appearance.

10.4. A 47-year-old patient presents with a jerky, low-frequency 2 Hz high-amplitude actiontremor. He also had impairment in finger-nose and heel-shin resting.

ANSWERS

1. HEADACHE

1.1. *B.* This young woman has a typical history for migraine with teichopsia, nausea and vomiting associated with a unilateral headache. The lack of fever makes meningitisunlikely.

1.2. *G.* Jaw claudication is a specific sign of temporal arteritis: urgent treatment with steroids is indicated.

1.3. *I.* An electric shock-like pain over one of the branches of the trigeminal nerve is a feature of trigeminal neuralgia. Attacks can be triggered by light touch, shaving, facewashing, or even brushing the teeth. The pain lasts up to 2 min, after which time it spontaneously stops.

1.4. *F.* Sufferers of cluster headache experience unilateral episodic, short attacks with no aura (unlike migraine). There is often associated ipsilateral lacrimation with nasalcongestion.

1.5. *E.* This patient has signs of raised intracranial pressure: morning headaches resolving through the day, worse on stooping. The only option which is compatible with along history and raised intracranial pressure is brain tumor.

2. NEUROANATOMY

2.1. *J.* The aqueduct of Sylvius, the communicating canal between the third and fourth ventricles, is particularly narrow and the refore susceptible to blockage, resulting inhydrocephalus.

2.2. *E.* This patient has an expressive dysphasia: she is able to understand but unable to speak. The lesion is in Broca's area, in the dominant (usually the left) frontal lobe.

2.3. *H.* The substantia nigra is an area of the midbrain with predominantly dopaminergic neurones. It is involved in the control of voluntary movement. Degeneration of thin area leads to the characteristic features of Parkinson's disease.

2.4. *F.* The olfactory bulb is located in the limbic system and is involved in the recognitionof smell.

2.5. *D.* A physiological function of the pineal gland is to secrete melatonin, a hormone which controls the body's diurnal rhythm. This hormone is available as a drug insome countries for jet-lag, but is not licensed in the UK.

3. CRANIAL NERVE LESIONS

3.1. *E.* VI. The history here is suggestive of benign intracranial hypertension. The inability togaze laterally occurs in abducens nerve palsy, a false localizing sign of raised intracranial pressure.

3.2. *H.* IX. There are two components to the gag reflex. The afferent (sensory) part involvesthe glossopharyngeal nerve, whereas the motor component is mediated via thevagus nerve.

3.3. *F.* VII. The facial nerve is responsible for taste in the anterior two-thirds of the tongue; the rest of the tongue is innervated by the glossopharyngeal nerve.

3.4. *I.* X. Before the advent of medication such as H_2-blockers (e.g. ranitidine) and proton pump inhibitors (e.g. omeprazole), highly selective vagotomy was a wellrecognized treatment for reducing acid secretion.

3.5. *J.* XI. The accessory nerve innervates the trapezius and sternocleidomastoid. The trapezius is responsible for elevating the shoulders.

4. DRUGS USED IN NEUROLOGICAL DISEASE

4.1. *D.* This patient is in status epilepticus, a medical emergency. First-line treatment, ifintravenous access is available, is with lorazepam. An alternative is rectal diazepam.

4.2. *A.* Riluzole is an approved treatment for amyotrophic lateral sclerosis. It can prolong the time patients have before they become dependent on mechanical ventilation.

4.3. *I.* Methylphenidate (Ritalin) is a stimulant that paradoxically works in attention deficit hyperactivity disorder. It should only be used as part of an overall strategy totreat the condition.

4.4. *F.* The history here is suggestive of absence seizures, with a temporary loss of awareness of the surroundings. Ethosuximide can be used to treat simple absence seizures.

4.5. *J.* Prochlorperazine, a phenothiazine, is a dopamine antagonist and acts on the centralnervous system to suppress nausea and vomiting.

5. DRUGS USED IN MOVEMENT DISORDERS

5.1. *J.* Tetrabenazine is thought to deplete nerve endings of dopamine and can improve symptoms in some patients with Huntington's disease.

5.2. *G.* Benserazide. Levodopa is always given with a dopamine decarboxylase inhibitor such asbenserazide, which does not cross the blood–brain barrier. This stops the peripheral metabolism of the levodopa and permits the utilization of smaller doses, which havefewer side effects.

5.3. *F.* Propranolol.The long history and lack of progression are indicative of benign essential tremor. Propranolol (and alcohol) can give symptomatic relief.

5.4. *E.* Cabergoline is an oral dopamine receptor agonist and is sometimes used as a first-line therapy for idiopathic Parkinson's disease.

5.5. *I.* Apomorphine is a powerful dopamine agonist that is only used in advanced Parkinson's disease owing to its need to be administered parenterally, and its potential to cause severe vomiting.

6. OFFENDING ARTERY

6.1. *L.* **6.2.** *H.* **6.3.** *K.* **6.4.** *E.* **6.5.** *J.*

7. PARANEOPLASTIC DISORDERS

7.1. *K.* Paraneoplastic cerebellar degeneration. Characterized by subacute, progressiveataxia, dysarthria, and nystagmus. Myoclonus, opsoclonus (irregular jerking of the eyesin all directions), diplopia, vertigo, and hearing loss may also occur. The most common associated tumor types are small cell carcinoma of the lung, ovarian/breast carcinoma, and lymphoma. Anti-Purkinje cell antibodies (anti-Yo antibodies) may be present in 50 %. Paraneoplastic cerebellar degeneration may precede the symptoms ofthe underlying tumor itself.

7.2. *A.* Hypercalcemia. It may be a result of parathyroid-related peptide secreted by the tumor itself (usually lung cancer) or of bone destructionby metastatic disease. The elevated serumcalcium decreases membrane excitability, leading to the clinical syndrome of fatigability, lethargy, generalized weakness, and areflexia progressing to coma and even convulsions. Symptoms usually do not occurs until levels reach 14 mg/dL (3 mmol/L) or higher.

7.3. *J.* Lambert-Eaton myasthenic syndrome. Lambert-Eaton myasthenic syndrome (LEMS) shows subacute proximalmuscle weakness and spares the bulbar musculature, and is due to presynaptic blockade of voltage gated calcium channels by autoantibodies. A characteristic feature is the increase in strength briefly after repeated muscle activation. Most cases are associated with small cell lung cancer, or in the context of other autoimmune diseases.

7.4. *I.* Paraproteinemic neuropathy. Polyneuropathy mayoccur in up to 15 % of patients with multiplemyeloma presents as a chronic distal symmetrical sensory or sensorimotor neuropathy. CSF protein may be elevated if there is a chronic inflammatory demyelinating polyneuropathy-like picture. Up to 20 % of patients referred for evaluation of polyneuropathy may a monoclonal gammopathy of undetermined significance, but a hematologic malignancy may later declare itself.

8. LEUKODYSTROPHIES

8.1. *I.* X-linked adrenoleukodystrophy. Produces rapidly evolving brain damage in maleinfants or boys, with survival from onset of symptoms usually limited to 3 years. Longchain fatty acids accumulate in adrenal cortical and other cells, resulting in adrenal insufficiency and CNS disease.

8.2. *G.* Pelizaeus-Merzbacher disease. Leukodystrophy with significant Sudan-staining typically become symptomatic during the first months of life, but survival may extend into the third decade of life. Most affected persons are male.

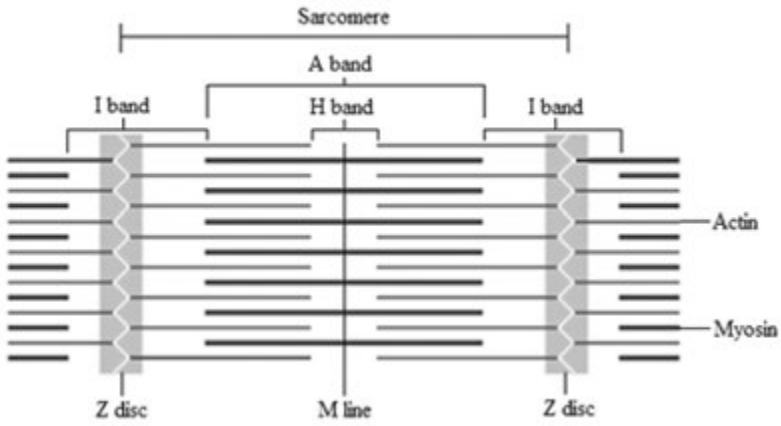
8.3. *H.* Metachromatic leukodystrophy. Sphingolipidosis due to arylsulfatase-A deficiency resulting in accumulation of galactosyl sulfatides. The affected person usually has retardation, ataxia, spasticity, and sensory disturbances usually symptomatic during infancy.

8.4. *D.* Canavan disease. Canavan disease may produce developmental regression at about 6 month of age, with extensor posturing, rigidity and myoclonic seizures may develop. There is accumulation of N-acetylaspartic acid in the blood and urine, but elevated levels in the brain establish the diagnosis.

9. SARCOMERE

9.1. *C.* **9.2.** *B, C.* **9.3.** *B.* **9.4.** *A.* **9.5.** *E.*

A single muscle cell contains several myofibrils, each of which consists of linear chains of solitary contractile units, or sarcomeres. Myofibrils are connected to one anothervia desmin intermediate filaments, and this complex is then anchored to the muscle cell sarcolemma by several proteins, including dystrophin. Within the myofibril, each sarcomere is connected to the adjacent sarcomere at the Z disc (E), and actin (thin) filaments radiate from the Z disc toward the center of the sarcomere. Myosin (thick) filaments are found interspersed between adjacent actin filaments. The dark band, or A band (A), is the region of the sarcomere that is composed primarily of myosin filaments. The light band, or I band (C), is the region that is composed solely of actin filaments and is centered on a Z disc. The H band (B) is the region of the A band where myosin filaments are not overlapped by actin filaments in the resting state, and it is centered on the M line. With myofibril contraction, the actin and myosin filaments form successive cross bridges that facilitate sliding across one another. This functionally shortens the sarcomere during contraction and consequently results in H band and I band shortening. This process of



muscle contraction requires the presence of cytosolic Ca^{2+}. In the resting state, tropomyosin and a troponin complex (troponins I, C, and T) are bound to actin. After the sarcoplasmatic reticulum releases Ca^{2+} in response to an action potential, troponin C binds four molecules of Ca^{2+}, which subsequently relieves the inhibition of the myosin binding site on action. Myosin heads are then free to bind actin and form cross bridges. The myosin head, which has intrinsic ATPase activity, then rotates, pulling the actin filaments longitudinally and increasing the overlap between the thick and thin filaments. ATP then binds to the myosin head, which stimulates the release of the cross bridge between actin. The subsequent hydrolysis of ATP «cocks» the myosin head, which then forms a second cross bridge, contracts, and further increases the overlap between thick and thin filaments. ADP is released, and the process is repeated as long as ATP and Ca^{2+} are present in the cytosol. In this manner, the myosin heads «walk» along the actin filaments during contraction, effectively shortening the sarcomere and myofibril.

10. TREMOR

10.1. *D.* **10.2.** *H.* **10.3.** *B.* **10.4.** *E.*