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Питання Європейського іспиту

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European Board Examination in Neurology
Variants of questions with answers

Closed Book
Questions related to the guidelines

1. Which of the following would be first-line therapy for sialorrhoea in Amyotrophic Lateral Sclerosis, according to EFNS guidelines (2012)?

A. Botulin toxin into the salivary glands
B. Irradiation of salivary glands
C. Submandibular duct transposition
D. Transdermal hyoscine
E. Oral trihexiphenidyl

2. Which of the following has been recommended to be performed every 6 months in a patient with definite amyotrophic lateral sclerosis, according to the EFNS guidelines (2012)?

A. Expiratory peak flow
B. Mini Mental State Examination
C. Serum albumin
D. Assessment of speech function
E. Chest X-ray

3. In which of the following regions of the world is dentatorubral-pallidoluysian-atrophy (DRPLA) most prevalent?

A. Africa
B. Asia
C. Australia
D. Europe
E. South America

4. Which of the following diseases is most likely given the finding of 14-3-3 proteins in the CSF?

A. Amyotrophic lateral sclerosis
B. Alzheimer's disease
C. Creutzfeld-Jakob disease
D. Frontotemporal dementia
E. Progressive supranuclear palsy

5. Which of the following imaging techniques is useful to distinguish between the various Parkinsonism-Dementia Syndromes?

A. Diffusion–tensor Imaging (DTI) MRI
B. Diffusion Weighted Image (DWI) MRI
C. Fluid-attenuated inversion recovery (FLAIR) MRI
D. SPECT perfusion imaging
E. T1-weighted MRI with gadolinium

6. Which of the following is in the exclusion-criteria for Parkinson's disease, according to the EFNS-Guideline (2013)?

A. Hyposmia
B. Severe L-dopa-induced chorea
C. Strictly unilateral involvement after 3 years
D. Symmetric presentation of symptoms
E. Visual hallucinations

7. Which of the following test is most sensitive for the diagnosis ocular myasthenia gravis?

A. AChR antibody detection
B. The edrophonium test
C. The ice-pack test
D. Repetitive stimulation of a cranial nerve
E. Single fibre EMG of a facial muscle

8. For which of the following anti-epileptic drugs is there some evidence of effectiveness in the treatment of primary restless legs syndrome?

A. Lamotrigine
B. Levetiracetam
C. Oxcarbazepine
D. Phenytoin
E. Pregablin

9. Which of the following non-pharmaceutical therapies has some effectiveness in the short-term treatment of restless legs syndrome?

A. Aerobic training
B. Infrared light therapy
C. Physiotherapy
D. Yoga
E. None of the above

10. Which of the following statements with regard to Polymerase Chain Reaction (PCR) in the diagnosis of viral CNS infections is most valid?

A. Due to technical restrictions only one virus within one CSF sample can be detected
B. Adequate specimen storage is important for optimal reliability of this test
C. The sensitivity of a PCR for Herpes Simplex Encephalitis decreases after 3 days
D. The specificity of a PCR for viral CNS diseases generally is lower than 80 %
E. Nowadays with PCR, brain biopsy for the diagnosis of progressive multifocal leukoencephalopathy (PML) is obsolete

11. Which of the following is the most prominent effect of pallidotomy in Parkinson's disease?

A. Reduction of bradykinesia
B. Reduction of dyskinesia
C. Reduction of freezing
D. Reduction of rigidity
E. Reduction of tremor

12. Which drug is recommended in the EFNS guidelines as first line treatment for convulsive status epilepticus in an adult person?

A. Diazepam
B. Levetiracetam
C. Lorazepam
D. Phenytoin
E. Valproate

Closed book
General questions

1. What kind of identification inability does prosopagnosia reflect?

A. Facial expression
B. Familiar faces
C. Melody in language
D. Non-musical sounds
E. Pitch of a melody

2. *Which of the following symptoms is found with increased frequency in patients with neuropathic pain as compared with patients with nociceptive pain?*
   A. Deep aching pain
   B. Lancinating pain
   C. Pain evoked by local pressure
   D. Pain on exercise
   E. Sensation of pins and needles

3. *What do laser evoked potentials mainly assess in patients with neuropathic pain?*
   A. The autonomous reflex loop
   B. The function of lemniscal pathway
   C. The function of spinothalamic pathway
   D. The function of spinal interneurons
   E. The interaction between lemniscal and spinothalamic pathways

4. *Which of the following drugs is recommended as the first line treatment of painful diabetic neuropathy?*
   A. Alpha1 blockers
   B. Chloride channel inhibitors
   C. Dopaminergic agents
   D. Serotonergic agents
   E. GABA-antagonists

5. *Which is the most common skeletal deformity associated with Charcot-Marie-Tooth disease?*
   A. Pes cavus (high arched foot)
   B. Pes equinus (horse foot)
   C. Pes equinovarus (club foot)
   D. Pes planus (flat foot)
   E. Pes valgovarus (skew foot)

6. *Which of the following drugs is the most effective in preventing attacks in Menière's disease ?*
   A. A glutamate antagonist
   B. A histaminergic drug
   C. A postassium-channel blocker
   D. A serotonergic drug
   E. A sodium-channel blocker

7. *Which of the following signs/syndromes is most likely to occur in a carotid artery dissection?*
   A. Adie syndrome (tonic pupil)
   B. Argyll Robertson pupil (pupillary dysfunction with near-light dissociation)
   C. Horner's syndrome (ptosis and miosis)
   D. Marcus-Gunn pupil (relative afferent pupillary defect)
   E. Parinaud syndrome (Vertical eye movement restriction with pupillary dysfunction)

8. *What is the correct term when a patient reports the experience of colours when hearing sounds?*
   A. Dysaesthesia
   B. Hyperaesthesia
   C. Paraesthesia
   D. Polyaesthesia
   E. Synaesthesia

9. *A bedbound and thin 81-year-old female presented with footdrop (weakened dorsiflexion and eversion of the foot). During the examination she also had sensory deficit in the anterolateral aspect of the lower leg and the dorsum of the foot including the web space between the 1st and 2nd metatarsals. Hip abduction by the gluteus medius was unaffected. Which nerve is most likely to be affected?*
   A. The femoral nerve
   B. The peroneal nerve
   C. The posterior tibial nerve
   D. The saphenous nerve
   E. The sciatic nerve

10. Which of the following statements relating to torticollis (cervical dystonia) is correct?
A. Torticollis is the most common form of focal dystonia
B. Torticollis is most common in men
C. Torticollis in young women is mostly associated with an autoimmune disease
D. The first line treatment of torticollis is an anticholinergic drugs
E. Torticollis is most often associated with an isolated genetic mutation

11. Which one of the following statements is most valid for primary torsion dystonia (PTD)?
A. PTD mostly starts after the age of 40 years
B. PTD can successfully be treated with botulinum toxin
C. PTD can successfully be treated with subthalamic nucleus deep brain stimulation
D. A cerebral MRI shows no abnormalities specific to PTD
E. More than half of the patients with PTD show pyramidal signs

12. Which of the following signs is a feature of progressive supranuclear palsy (PSP)?
A. Atrophy of the tongue
B. Broad based gait
C. Extensor plantar responses
D. Miosis
E. Spastic hypernasal speech

13. Which of the following structures undergoes degeneration that leads to neuropathic symptoms in Friedreich's ataxia?
A. Anterior roots
B. Dorsal root ganglia
C. Posterior columns
D. Posterior roots
E. Spinocerebellar tracts

14. In which region of the cortex is alpha-activity most prominent in an EEG done with the eyes closed?
A. Central
B. Frontal
C. Occipital
D. Parietal
E. Temporal

15. Which component of somatosensory evoked potentials (SEPs or SSEPs) indicates radicular-medullary damage?
A. N13
B. P40
C. N70
D. P100
E. P300

16. Which of the following drugs should be chosen preferentially for treating vestibular paroxysmia due to vascular compression of the vestibular nerve?
A. Acetazolamide (Carbo-Anhydrase-Inhibitor)
B. Aminopyridine (Potassium Channel Blocker)
C. Baclofen (GABA-ergic Drug)
D. Carbamazepine (Sodium Channel Blocker)
E. Diazepam (Chloride Channel Activator)

17. Where do the cholinergic neurons of the nucleus basalis Meynert mainly project to?
A. Corpus pineale
B. Frontal neocortex
C. Nucleus subthalamicus
D. Putamen
E. Thalamus

18. *What does gadolinium enhancement reflect in patients with MS?*
   A. Active antigen presentation
   B. Acute axonal damage
   C. Blood brain barrier permeability abnormalities
   D. Presence of macrophages
   E. Vanishing white matter

19. *What can be measured using proton MR spectroscopy?*
   A. Axonal density
   B. The molecular composition of tissue
   C. The relative proportions of free and bound water
   D. Tissue anisotropy
   E. Water molecular motion and tissue anisotropy

20. *What does the cerebrospinal fluid (CSF) albumin / serum albumin ratio reflect?*
   A. Total protein content
   B. Cerebrospinal fluid density
   C. Blood-brain barrier status
   D. Neurodegeneration
   E. Immunological status

ANSWERS

Closed Book
Questions related to the guidelines

1. *D.* Transdermal hyoscine.

Sialorrhoea Sialorrhoea (drooling or excessive salivation) is common and may be socially disabling. Amitriptyline is often used, but there are no formal studies proving its efficacy. Oral doses of 10 mg three times a day are often sufficient. Atropine drops, 0.5 % or 1 %, administered three or four times a day sublingually have the advantage of a short duration of action — valuable in patients who suffer from sialorrhoea alternating with an uncomfortably dry mouth. Transdermal hyoscine (scopolamine), 1.5 mg every third day, reduces salivary flow. Care is needed in elderly patients, because of the frequent side effects of confusion or loss of bladder control. Caution is needed in patients with significant bulbar palsy as increased dysphagia may occur, with serious consequences. Another option is external irradiation of the salivary glands, with four studies showing satisfactory results.

2. *D.* Assessment of speech function.

1. Regular assessment (i. e. every 3—6 months) of speech and language function by a trained speech and language therapist is recommended (GCPP). 2. Those with evidence of early language deficits should undergo full neuropsychological testing (GCPP). 3. The use of appropriate communication support systems (ranging from pointing boards with figures or words, to computerized speech synthesizers) should be individualized and appropriate training and support provided as required (GCPP).

3. *B.* Asia.

Dentatorubral pallidoluysian atrophy (DRPLA), also known as Naito-Oyanagi disease, Haw River syndrome, or myoclonic choreoathetosis with epilepsy is a hereditary neurodegenerative disease that is especially prevalent among individuals of Japanese origin, is a rare subtype of type I autosomal dominant cerebellar ataxia (ADCA type I). It is characterized by involuntary movements, ataxia, epilepsy, mental disorders, cognitive decline and prominent anticipation.

4. *C.* Creutzfeld-Jakob disease.

The 14-3-3 protein is a normal neuronal protein that is released into cerebrospinal fluid (CSF) in association with acute neuronal injury. It has been suggested that the presence of 14—3-3 protein in CSF is a reliable marker for Creutzfeldt-Jakob disease (CJD), with sensitivity and specificity for this protein reported as high as 96 % and 93 % to 100 %, respectively. These reports have recently led the World Health Organization to revise its diagnostic criteria for probable sporadic CJD (sCJD) to allow substitution of a positive 14—3-3 test for a positive electroencephalogram, provided the disease has less than a 2-year duration. Yet, a series of studies has suggested that both sensitivity and specificity for sCJD are lacking with this test.

5. *E.* T1-weighted MRI with gadolinium.

MR imaging in Parkinson disease reveals loss of the substantia nigra pars compacta, with reduced width in a lateral to medial gradient (most prominently of the more lateral portion), best evaluated on dedicated inversion recovery T1-weighted sequences.

6. *C.* Strictly unilateral involvement after 3 years.

Exclusion criteria for Parkinson's disease:
- History of repeated strokes with stepwise progression of parkinsonian features
- History of repeated head injury

- History of definite encephalitis
- Oculogyric crises
- Neuroleptic treatment at onset of symptoms
- More than one affected relative
- Sustained remission
- Strictly unilateral features after 3 years
- Supranuclear gaze palsy
- Cerebellar signs
- Early severe autonomic involvement
- Early severe dementia with disturbances of memory, language and praxis
- Babinski sign
- Presence of a cerebral tumor or communicating hydrocephalus on CT scan
- Negative response to large doses of L-dopa (if malabsorption excluded)
- MPTP exposure

7. *E.* Single fibre EMG of a facial muscle.

8. *E.* Pregablin.

The need for treatment should be based on the frequency of troublesome symptoms and the effect that these have on sleep quality. For patients with infrequent bouts of RLS, treatment on an as-required basis may be appropriate. Avoidance of aggravating substances, such as nicotine, caffeine, and alcohol, should be advised. Stretching and light exercise may also help. The following recommendations are in line with advice provided by the American Academy of Sleep Medicine and the EFNS/ European Neurological Society (ENS)/European Sleep Research Society (ESRS) joint guidelines:

- First line: dopamine agonists (DAs, e. g. pramipexole, ropinirole, and rotigotine);
- Second line: levodopa, gabapentin enacarbil, or opioids;
- Third line: cabergoline, pregabalin, or carbamazepine;
- Other: benzodiazepines (short term to improve sleep quality).

Additionally, iron supplementation in those known to be deficient can improve symptoms.

Seventy per cent of patients treated with levodopa, and to a lesser extent dopamine agonist (DA), are associated with the phenomenon of «augmentation». Patients report the onset of symptoms earlier in the day, spread to the trunk and upper limbs, and reduced duration of treatment effect.

9. *E.* None of the above.

10. *B.* Adequate specimen storage is important for optimal reliability of this test.

Both the timing of the CSF sample and the physical conditions such as specimen storage can be important confounding variables in PCR diagnosis, so close attention needs to be given to these. Related to this, a self-evident pre-requisite for obtaining a very high specificity for viral PCR is that the laboratory carrying out the assay is fully experienced in this technique; in particular, it is vital to avoid contamination.

11. *B.* Reduction of dyskinesia.

Pallidotomy

- Reduction of dyskinesia associated with levodopa therapy.

Indications

Advanced idiopathic Parkinson disease with predominant symptoms of rigidity and tremor and «on time» dyskinesia.

Contraindications

- Significant dementia, cognitive, swallowing, or speech difficulties
- High risk for hemorrhage, coagulopathy, poorly controlled hypertension, and advanced age
- Procedure should not be performed bilaterally.
- Ipsilateral hemianopsia
- Short duration of disease

12. *C.* Lorazepam.

The preferred treatment pathway for generalised convulsive status epilepticus (GCSE) is intravenous (i.v.) administration of 4—8 mg lorazepam or 10 mg diazepam directly followed by 18 mg/kg phenytoin. If seizures continue more than 10 min after first injection, another 4 mg lorazepam or 10 mg diazepam is recommended.

Closed book
General questions

1. *B.* Familiar faces.

Prosopagnosia, also called face blindness, is a cognitive disorder of face perception in which the ability to recognize familiar faces, including one's own face (self-recognition), is impaired, while other aspects of visual processing (e. g., object discrimination) and intellectual functioning (e. g., decision making) remain intact.

Patients do not recognize familiar faces, including those of family members. This syndrome is usually due to lesions of the right inferior occipitotemporal medial temporal (fusiform gyrus) region.

Prosopagnosia is a form of visual agnosia characterized by an inability to recognize previously known human faces or equivalent stimuli (hence, a retrograde defect) and to learn new ones (anterograde defect).

As with more pervasive visual agnosia, this may be

- apperceptive: due to faulty perceptual analysis of faces; or
- associative: a semantic defect in recognition.

Familiar individuals may be recognized by their voices or clothing or hair; hence, the defect may be one of visually triggered episodic memory. It is important to note that the defect is not limited solely to faces; it may encompass animals ('zooagnosia') or cars.

Prosopagnosia is often found in association with a visual field defect, most often a left superior quadrantanopia or even hemianopia, although for the diagnosis of prosopagnosia to be made this should not be sufficient to produce a perceptual deficit. Alexia and achromatopsia may also be present, depending on the exact extent of the underlying lesion.

Anatomically, prosopagnosia occurs most often in association with bilateral occipito-temporal lesions involving the inferior and mesial visual association cortices in the lingual and fusiform gyri, sometimes with subjacent white matter. Unilateral non-dominant (right) hemisphere lesions have occasionally been associated with prosopagnosia, and a syndrome of progressive prosopagnosia associated with selective focal atrophy of the right temporal lobe has been reported. Involvement of the periventricular region on the left side may explain accompanying alexia, and disconnection of the inferior visual association cortex (area V4) may explain achromatopsia.

Pathological causes of prosopagnosia include

- Cerebrovascular disease: by far the most common cause;
- Tumour, e.g. glioma, extending from one hemisphere to the other via the splenium of the corpus callosum;
- Epilepsy (paroxysmal prosopagnosia), due to bilateral foci or spread from one occipital focus to the contralateral hemisphere;
- Focal right temporal lobe atrophy;
- Herpes simplex encephalitis, usually as part of an extensive amnesic syndrome (although memory impairment may put this outwith the operational criteria for an agnosia);
- Developmental (or 'congenital') prosopagnosia; suggests that facial recognition is a separate neuropsychological function (the acquired pathologies do not respect functional boundaries).

2. *E.* Sensation of pins and needles.

If we start with the concept of pain as a disease, then treatment begins with a pain diagnosis. Pain pathophysiology comprises 2 categories: nociceptive and neuropathic pain. Nociceptive pain is further divided into visceral and somatic pain, and neuropathic pain is divided into peripheral and central neuropathic pain.

Nociceptive, or somatic, pain is the common discomfort we have all experienced as a result of injury — a paper cut, a broken bone, or appendicitis, among other things. Somatic pain makes sense to us; we can understand the patient's pain.

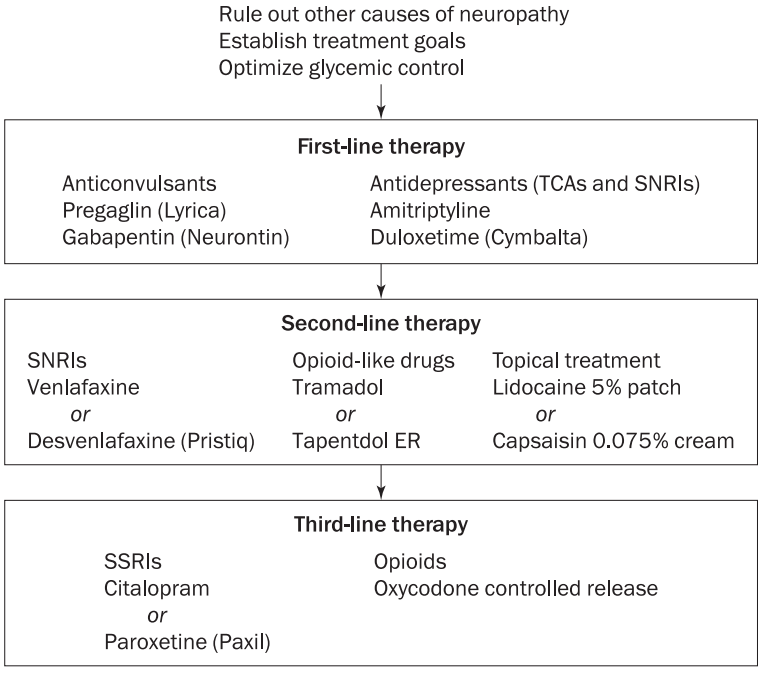
Neuropathic pain is associated with injury to a nerve or the central nervous system. Such injuries can give rise to paresthesias, such as numbness, tingling, or electrical sensations. Neuropathic pain can also generate unusual symptoms, such as anesthesia dolorosa, in which the area producing the pain is numb to the touch. This symptom is often puzzling to patients, and some have questioned their own sanity because painful numbness makes no sense. They can pinch the area and not feel the pinch, yet the area is excruciatingly painful. In addition, with nerve, spinal cord, or brain injuries, syndromes (such as allodynia) can develop and ordinarily non-noxious stimuli (such as light pressure or stroking) cause pain.

3. *C.* The function of spinothalamic pathway.

Until the 1980s, central neuropathic pain (CNP) was mainly viewed as a 'release' phenomenon caused by a lesion that removed inhibitory influences of the lemniscal pathways on pain systems. However, detailed sensory analysis of CNP patients later demonstrated that almost all cases had lesions affecting the major pathways for temperature and pain sensation (the spino-thalamo-cortical pathways), while concomitant injury to the medial lemniscal system was not essential for the development of CNP. Accordingly, impairment of spinothalamic pathways is now regarded as a crucial or even sine qua non condition for the occurrence of CNP.

4. *D.* Serotonergic agents.

Management of Painful Diabetic Peripheral Neuropathy



ER = extended release; SNRI = serotonin-norepinephrine reuptake inhibitor; SSRI = selective serotonin reuptake inhibitor; TCA = tricyclic antidepressant.

Pregabalin (Lyrica), gabapentin (Neurontin), amitriptyline (except in older adults), or duloxetine (Cymbalta) should be used as first-line treatment for painful diabetic peripheral neuropathy.

5. *A.* Pes cavus (high arched foot).

In CMT1, symptoms often begin during the first or second decade of life. It is characterized by slowly progressive weakness, muscular wasting, and sensory impairment predominantly involving the distal legs. Foot deformities and difficulties in running or walking resulting from symmetrical weakness and wasting in the intrinsic foot, peroneal, and anterior tibial muscles are often present. In two-thirds of patients, the upper limbs are involved later in life. Inspection reveals pes cavus and hammer toes in nearly three-quarters of adult patients, mild kyphosis in approximately a tenth, and palpably enlarge hypertrophic peripheral nerves in a quarter. The foot deformities occur because of long-term muscular weakness and imbalance between the intrinsic extensor and long extensor muscles of the feet and toes (a similar process cases clawing of the fingers in more advanced cases).

6. *B.* A histaminergic drug.

Treatment

There is no cure for Ménière's disease; hence the aim of treatment is to reduce symptom load.

- Acute: treatment of Ménière's disease involves the control of vertigo and nausea in the acute setting with antiemetics, antihistamines, and other centrally acting sedative medication.
- Long-term treatment:
  - First line: salt restriction and diuretics;
  - Second line: betahistine;
  - Third line: steroids and gentamicin administered via intratympanic injection.

7. *C.* Horner's syndrome (ptosis and miosis).

How does carotid dissection present? Sudden neck pain, ipsilateral Horner syndrome, and sometimes lower cranial nerve palsies.

Where do intracranial dissections most commonly occur? Vertebral >> basilar > internal carotid > MCA > ACA, PCA, PICA.

How do spontaneous dissections present? Vertebral artery: neck pain, generalized severe headache, stroke, TIA, SAH

ICA: ipsilateral headache (often orbital), and incomplete Horner's syndrome (aka oculosympathetic palsy; ptosis and miosis without anhidrosis), bruit.

8. *E.* Synaesthesia.

Synaesthesia is a perceptual experience in one sensory modality following stimulation of another sensory modality. The most commonly encountered example is colour-word synaesthesia ('coloured hearing' or chromaesthesia), experiencing a visual colour sensation on hearing a particular word. Synaesthesia occurs in a small percentage of the normal population. Known synaesthetes include the composers Messiaen and Scriabin, the artist Kandinsky, and the author Nabokov. There may be concurrent excellent memory (hypermnesia), sometimes of a photographic nature (eidetic memory). Symptomatic synaesthesia is rare but has been described with epileptic seizures of temporal lobe origin and with drug use (LSD). Characteristics ascribed to synaesthetic experience include its involuntary or automatic nature, consistency, generic or categorical and affect-laden quality. Neuropsychologically, this phenomenon has been conceptualized as a break down of modularity. Functional imaging studies of colour-word synaesthetes show activation of visual associative areas of cortex (but not primary visual cortex), as well as perisylvian language areas, when listening to words which evoke the experience of colour.

9. *B.* The peroneal nerve.

The deep fibular (peroneal) nerve is a terminal branch of the common fibular nerve that supplies motor innervation to the four muscles of the anterior compartment of the leg: (1) tibialis anterior, (2) extensor digitorum longus, (3) extensor hallucis longus, and (4) fibularis (peroneus) tertius. These four muscles are responsible for dorsiflexion of the ankle. The deep fibular nerve also innervates the extensor digitorum brevis and extensor hallucis brevis, which are intrinsic muscles of the foot, sends articular branches to joints it crosses, and supplies cutaneous innervation to the first interdigital cleft. The deep fibular nerve is responsible for dorsiflexion of the foot at the angle joint, and the segmental innervation of this movement is L4 and L5.

10. *A.* Torticollis is the most common form of focal dystonia.

Cervical dystonia (or spasmodic torticollis) is the most common form of focal or localized dystonia.

11. *D.* A cerebral MRI shows no abnormalities specific to PTD.

MRI of the brains of patients with DYT1 dystonia is usually normal.

12. *E.* Spastic hypernasal speech.

Signs & Symptoms

The signs and symptoms of PSP vary from person to person, but patients generally fall into one of four clinical syndromes (phenotypes): Richardson syndrome, atypical Parkinsonism, corticobasal syndrome, and pure akinesia and gait freezing. Less commonly, patients present with cognitive loss and no motor signs.

The most common presentation is the Richardson syndrome, consisting of gait and balance impairment, a wide-eyed staring facial expression, abnormal speech, memory and cognitive impairment and a slowing or loss of voluntary eye movement, particularly in the downward direction (supranuclear ophthalmoplegia). Cognitive symptoms include forgetfulness and personality changes, such as loss of interest in formerly pleasurable activities (apathy), impaired attention and concentration, depression, and increased irritability.

Fewer than half of all PSP patients are initially diagnosed correctly because many patients do not present with the classic Richardson syndrome. Many of these patients are initially slow and have muscle rigidity and occasionally tremor, resembling Parkinson disease, and they may initially respond somewhat to levodopa. Other patients present with bizarre stiffening (rigidity and dystonia) and loss of voluntary function in one upper limb, as is seen in corticobasal degeneration. Rarely, patients exhibit the syndrome of primary akinesia and gait freezing. These patients exhibit hesitant initiation of gait and a tendency to freeze or stop when turning and when crossing thresholds (doorways). Their eye movements and cognition

are normal. Small handwriting and low-volume rapid, mumbling speech (tachyphemia or cluttered speech) are typical and are similar to that which occur in Parkinson disease, but in contrast to Parkinson disease, there is no slowness (bradykinesia) or muscle stiffness (rigidity). Finally, some patients with PSP present with cognitive impairment, resembling Alzheimer disease or frontotemporal dementia. Most patients with atypical presentations ultimately develop abnormalities of eye movement, speech, swallowing and gait (Richardson syndrome) in a few years. Thus, the diagnosis of PSP typically becomes more certain as the disease progresses.

13. *B.* Dorsal root ganglia.

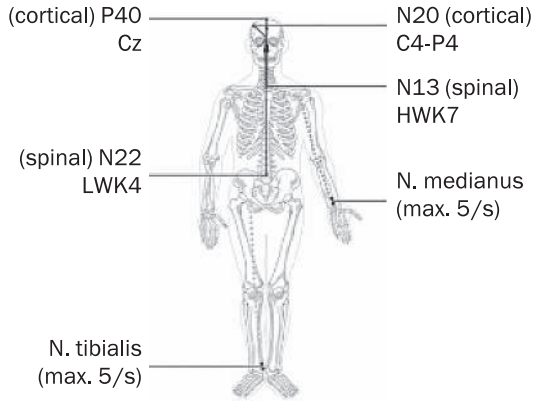
In a post scriptum to his 4 previous articles on ataxia, Friedreich (1825—1882) gave detailed gross and microscopic descriptions of the spinal cord, spinal gray matter, dorsal spinal roots, and medulla oblongata. This review of the neuropathology of Friedreich ataxia stresses the critical role of hypoplasia and superimposed atrophy of dorsal root ganglia.

14. *C.* Occipital.

Alpha rhythm may have a frequency of between 8 and 13 Hz, but in most adults it is between 9 and 11 Hz. This rhythm is found most typically over the posterior portions of the head during wakefulnes. Alpha rhythm is seen best when the patient is resting with the eyes closed. Immediately after eye closure, its frequency may be increased transiently (the «squeak» phenomenon).

15. *A.* N13.

N13 is a near-field potential recorded at the C5–C7 spinous process using a noncephalic reference, felt to be generated by the postsynaptic activity in the large myelinated fibers in the dorsal horn.



A larger and more consistent component recorded over the lower cervical spine is N13. N13 has a horizontally oriented voltage field, negative dorsally and positive ventrally (see image below), and is generated by postsynaptic activity of neurons in the gray matter of the lower cervical spinal cord. It sometimes is called the stationary cervical potential, because its latency is not affected by changes in the cervical recording electrode location.

16. *D.* Carbamazepine (Sodium Channel Blocker).

Vestibular paroxysmia

This is a rare, but important, syndrome characterized by brief (milliseconds to seconds) attacks of vestibular and auditory deficits, including vertigo, imbalance, and tinnitus. It is thought to arise from neurovascular cross-compression of the vestibular nerve and an offending vessel. It is important to consider the diagnosis in patients presenting with paroxysmal spontaneous vertigo, as it is readily treatable.

Treatment

- First line: carbamazepine or oxcarbazepine.
- Second line: gabapentin, lamotrigine, phenytoin, or sodium valproate.
- Refractory symptoms: microvascular neurosurgical decompression of the eighth cranial nerve.

17. *B.* Frontal neocortex.

Basal Forebrain Nuclei and Septal Area

Several poorly defined cell islands, located beneath the basal ganglia deep in the hemisphere, project widely to the cortex. These cell islands include the basal forebrain nuclei (also known as the nuclei of Meynert or substantia innominata), which send widespread cholinergic projections throughout the cerebral cortex. Located just laterally are the septal nuclei, which receive afferent fibers from the hippocampal formation and reticular system and send axons to the hippocampus, hypothalamus, and midbrain.

Major cholinergic pathways

1. Septal nuclei
   - project via the fornix to the hippocampal formation.
2. Basal nucleus of Meynert
   - is located in the substantia innominata of the basal forebrain, between the *globus pallidus* and the anterior perforated substance.
   - projects to the entire neocortex.
   - receives input from the locus ceruleus, raphe nuclei, substantia nigra, amygdaloid *nucleus*, and orbitofrontal and temporal cortices.
   - degenerates in Alzheimer's disease.

The basal *nucleus* of Meynert projects to the entire cortex; this *nucleus* degenerates in Alzheimer's disease. Striatal ACh-local-circuit neurons degenerate in Huntington's chorea.

18. *C.* Blood brain barrier permeability abnormalities.

Gadolinium-enhanced MR imaging appears to be more sensitive than HICT in the detection of the transient abnormalities of the blood-brain barrier that occur in patients with active MS and appears capable of distinguishing active lesions that may correspond to the anatomic regions responsible for abnormal clinical findings.

It is generally believed that because acute MS lesions are associated with a transient breakdown of the blood-brain barrier (BBB), gadolinium contrast agents would produce enhancement of these lesions on T1-weighted images. Beginning with the earliest magnetic resonance studies of MS, it became clear that the correlation between enhanced lesions and clinical disease activity is modest at best.

19. *B.* The molecular composition of tissue.

What is MRS based on? The detection of metabolites that have a concentration of at least 0.5 to 1.0 mM (millimolar).

How does MRS work? By placement of a voxel in the region of interest and comparing the relative concentration of metabolites in that region.

20. *C.* Blood-brain barrier status.

Albumin is produced exclusively in the liver, so all albumin detected in the CSF originates by definition from blood. Therefore, it serves as an ideal parameter to evaluate the permeability of the blood-CSF barrier.