Discussion & Reference Manual

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EXAM OBJECTIVES

The Residency In-service Training Examination (RITE) is sponsored by the American Academy of Neurology (AAN) in order to achieve the following:

- To provide residents the opportunity to assess their knowledge in neurology and neuroscience;
- To sharpen residents' knowledge base by identifying areas for potential growth;
- To serve as a tool for further education in neurology by providing the references and discussions for each item.

This examination is not designed to be a certifying or qualifying examination, and its use in that manner is vigorously discouraged by both the Residency Examination Subcommittee and the AAN Board of Directors.

FREQUENTLY CITED REFERENCES


FREQUENTLY CITED REFERENCES (continued)


Tucker GJ. Psychiatry for the neurologist part A. Continuum 1997 Nov;3(6).

Question(s) 1: Anatomy
Discussion:
The supraspinatus muscle initiates abduction of the fully adducted upper extremity.

Reference:

Question(s) 2: Physiology
Discussion:
Wave V is generated by auditory structures in the area of the inferior colliculus.

Reference:

Question(s) 3: Radiology
Discussion:
The Chiari type I malformation is defined as congenital displacement of cerebellar tonsils (with or without vermian displacement) into the cervical spinal canal. It is frequently associated with a narrow foramen magnum. Hydrocephalus is not a common finding. More than 2-3 mm downward displacement of the tonsils is considered abnormal. The reference point is the posterior lip of the foramen magnum. Herniation beyond C3 is rare. The fourth ventricle is in its normal position in the posterior fossa. Spinal dysraphism, an open tentorial incisura, and a narrow tectum are associated with Chiari type II (but not with Chiari type I) malformation. Chiari type I malformation is often asymptomatic, especially when small.

Reference:

Question(s) 4: Behavioral
Discussion:
a lesion in the language dominant hemisphere temporal-parietal area can impair the ability to link symbolic meaning to a word that can be repeated correctly, the disorder known as transcortical sensory aphasia.

Reference:

Question(s) 5: Physiology
Discussion:
End-plate activity as recorded by needle EMG represents miniature end-plate potentials recorded extracellularly.

Reference:

Question(s) 6: Anatomy
Discussion:
The abducens nerves emerge from the ventral surface of the brainstem at the junction between the pons and medulla.

Reference:

Question(s) 7: Pharmacology/Chemistry
Discussion:
Ornithine carbamoyltransferase deficiency is X-linked recessive and typically presents in infancy with coma and hyperammonemia. In females, the presentation is heterogenous and may be delayed into adulthood.

Reference:

Question(s) 8: Behavioral
Discussion:
The hallmark of conduction aphasia is impaired repetition. While any type of paraphasia may be seen in conduction aphasia, the vast majority of substitutions will involve phonemes resulting in production of literal (phonemic) paraphasic errors. Word substitution errors (semantic paraphasia) are less characteristic of conduction aphasia.
Question(s) 9: Pharmacology/Chemistry
Discussion:
Niemann-Pick disease type C is characterized by autosomal recessive inheritance with gradual and progressive neurological deterioration. Patients may have hepatosplenomegaly, neurological dysfunction, including vertical supranuclear (usually downgaze) palsy, ataxia dystonia and cognitive impairment.

Reference:

Question(s) 10: Clinical Adult
Discussion:
Pseudotumor cerebri is much more common in women and is characterized by normal CSF composition and normal ventricles on neuroimaging. The neurologic examination is typically normal, but sixth nerve palsies may occur.

Reference:

Question(s) 11: Pharmacology/Chemistry
Discussion:
Praziquantel is effective in treating cerebral cysticercosis. Active, uninflamed cysts are responsive. Praziquantel will produce inflammation and edema as it kills the larva, which may temporarily result in symptoms such as headache.

Reference:

Question(s) 12: Clinical Pediatrics
Discussion:
Spasmus nutans usually begins in the first year of life. Children are neurologically normal, although some may have coexisting amblyopia or strabismus. The triad of abnormal head position(s), head nodding and nystagmus is characteristic, although not all components need be present simultaneously, and the nystagmus may sometimes be monocular. The major differential diagnosis is tumors of the optic apparatus. Another important distinction is that between spasmus nutans and the chaotic, but conjugate saccadic eye movements frequently seen in association with ataxia and myoclonus in children with neuroblastoma. The syndrome usually remits in one to two years.

Reference:

Question(s) 13: Anatomy
Discussion:
Visual, auditory, gustatory and nociceptive sensory modalities reach the cerebral cortex through the thalamus. Olfactory pathways bypass the thalamus.

Reference:

Question(s) 14: Behavioral
Discussion:
The syndrome in which an individual's non-paralyzed hand appears to carry out activities (usually grasping) that cannot be controlled by the individual has been called the alien hand syndrome. Damage to the corpus callosum usually underlies the alien hand syndrome. When it is secondary to infarct, it is in the anterior cerebral artery territory distribution.

Reference:
Question(s) 15: Pharmacology/Chemistry
Discussion:
Dopamine agonists such as ropinirole and pramipexole may produce sudden irresistible attacks of sleep while driving. Neurologists who prescribe these drugs and patients who take them should be aware of this possible side effect.

Reference:

Question(s) 16: Physiology
Discussion:
Centrotemporal spikes in the EEG of a child are associated with benign focal epilepsy of childhood, a genetically-determined disorder that remits during adolescence.

Reference:

Question(s) 17: Pathology
Discussion:
Clinically, patients with diffuse axonal injury are usually unconscious immediately after injury and remain comatose or develop a persistent vegetative state.

Reference:

Question(s) 18: Physiology
Discussion:
When abnormal, sensory nerve action potentials indicate dysfunction distal to the level of the dorsal root ganglion and thus can distinguish between pre-ganglionic (root) and post-ganglionic (plexus, nerve) lesions.

Reference:

Question(s) 19: Physiology
Discussion:
The characteristic electrophysiologic findings in Lambert-Eaton myasthenic syndrome are: decreased amplitude of compound muscle action potentials (CMAP) with further amplitude reduction (decrement) to repetitive nerve stimulation between 1 and 5 Hz; marked increase in CMAP size in response to repetitive stimulation at 20-50 Hz and an increase in CMAP size after brief maximal voluntary contraction.

Reference:

Question(s) 20: Behavioral
Discussion:
Simultanagnosia is an abnormality of visual attention in which the patient is unable to see a panorama but has good visual acuity. This occurs predominantly in patients with high parietal lobe disease such as infarcts in the posterior watershed, sagittal sinus thromboses and in some patients with Alzheimer's disease.

Reference:

Question(s) 21: Pathology
Discussion:
Although the etiology of Ammon's horn sclerosis (AHS) is controversial, with some authors contending the lesions are the cause and others the result of the seizures, the lesion is nonetheless the most common pathology found in patients with tissue resected for temporal lobe epilepsy. Seizure control after temporal lobectomy is best with AHS but is also very good with neoplasms and vascular malformations, although these are less common causes of temporal lobe epilepsy. End folium sclerosis with neuronal loss confined to the CA4 sector of the hippocampus is rare, as is Rasmussen's encephalitis.
Reference:

Question(s) 22: Pharmacology/Chemistry
Discussion:
Drugs of addiction increase levels of dopamine released at projections of the ventral tegmental area. Cocaine and amphetamine raise dopamine levels in the nucleus accumbens by blocking the dopamine transporter, prolonging the duration of action of dopamine in the synaptic cleft. Nicotine enhances release of dopamine by acting on the presynaptic cholinergic receptors, and mu-opioids inhibit dopaminergic neurons that suppress dopaminergic neurons in the ventral tegmental area.

Reference:

Question(s) 23: Anatomy
Discussion:
The most reliable marker for neuronal differentiation is synaptophysin. Despite the promising name, antibodies raised against neuron-specific enolase have proven to be of very low specificity for neuronal differentiation. Desmin is a marker for muscle differentiation, S-100 protein is abundant in most glial cells, and vimentin is a ubiquitous intermediate filament protein found in a wide variety of cell types.

Reference:

Question(s) 24: Behavioral
Discussion:
The salient feature of an acute confusional state is a disturbance of attention and concentration.

Reference:

Question(s) 25: Pathology
Discussion:
The histologic hallmarks of Alzheimer's disease are neuritic plaques, neurofibrillary tangles, Hirano bodies, granulovacuolar degeneration of neurons, and the deposition of amyloid in the walls of blood vessels in the leptomeninges and cerebral cortex. Alzheimer type II astrocytes, in contrast, are reactive astrocytes that are seen in hyperammonemonic conditions and are not a feature of Alzheimer's disease. Ballooned neurons are a feature of corticobasal degeneration.

Reference:

Question(s) 26: Physiology
Discussion:
Nightmares and cluster headache typically arise from REM sleep. REM occurs at onset of neonatal sleep. Pavor nocturnus, or night terrors, is an arousal from stage 4 sleep commonly seen in children.

Reference:

Question(s) 27: Anatomy
Discussion:
The facial nerve conveys fibers to the stapedius, orbicularis oculi, platysma and zygomaticus muscles, and preganglionic parasympathetic fibers to the submandibular ganglion. The trigeminal nerve conveys special visceral efferent fibers to the muscles of mastication, the tensor tympani and tensor veli palatini.

Reference:
Question(s) 28: Pharmacology/Chemistry

Discussion:
Some patients with autonomic failure exhibit a degree of hypoproliferative anemia. Administration of recombinant human erythropoietin-alpha, 25-75 U/kg subcutaneously two to three times a week for six weeks, rapidly corrects the anemia and improves orthostatic tolerance.

Reference:
Hoeldtke RD, Streeten DHP. Treatment of orthostatic hypotension with erythropoietin. NEJM 1993;329:611-615.

Question(s) 29: Pharmacology/Chemistry

Discussion:
Patients with lesions in the medial portion of the frontal lobes may have an uninhibited bladder, characterized by inability to voluntarily inhibit an otherwise normal micturition reflex. In patients with detrusor hyperreflexia without outlet obstruction, the drugs of choice are anticholinergics or direct smooth muscle relaxants, such as oxybutynin or tolterodine.

Reference:

Question(s) 30: Physiology

Discussion:
Significant membrane depolarization is caused by elevated extracellular potassium ions.

Reference:

Question(s) 31: Anatomy

Discussion:
The facial (intermediate) nerve provides preganglionic inputs from the superior salivatory nucleus to the sphenopalatine (pterygopalatine) ganglion, which innervates the lacrimal gland. The otic ganglion receives preganglionic inputs from the glossohyparyngeal nerve. The nodose, petrosal, and geniculate ganglia are sensory ganglia and do not receive preganglionic inputs.

Reference:

Question(s) 32: Clinical Adult

Discussion:
In a large scale epidemiologic assessment of apparently healthy men followed prospectively in the Physician's Health Study, the relative risk of venous thrombosis was 2.7 among men with a point mutation in the gene coding for coagulation factor V resulting in resistance to activated protein C. The prevalence of the heterozygous state for this mutation is 6.0%, making this the most common inherited condition predisposing to venous thrombosis identified to date. The mutation was not associated with an increased risk of myocardial infarction or stroke.

Reference:

Question(s) 33: Radiology

Discussion:
Luxury perfusion refers to increased perfusion in the periphery of an ischemic area. It is best documented with PET or SPECT. Some authors have used the term to refer to gray matter enhancement, which is best seen on MRI three days to several weeks after a cerebral infarct and results from contrast leaking into the perivascular space in an area of ischemic insult.

Reference:
Question(s) 34: Physiology
Discussion:
Calcium ions are necessary for production of excitation-contraction coupling in striated muscle.

Reference:

Question(s) 35: Pharmacology/Chemistry
Discussion:
The Lambert-Eaton myasthenic syndrome occurs most often, but not always, in patients with small cell cancer of the lung. These patients have proximal muscle weakness that may improve with exercise. The pathogenesis is an antibody directed against the presynaptic voltage-gated P/Q type calcium channel.

Reference:

Question(s) 36: Clinical Adult
Discussion:
Wernicke's encephalopathy, which results from deficiency of thiamine (vitamin B1), may be precipitated in at-risk patients by intravenous glucose administration or carbohydrate loading.

Reference:

Question(s) 37: Pharmacology/Chemistry
Discussion:
Fragile-X syndrome is characterized by X-linked mental retardation. Affected males are moderately to severely mentally retarded and have a characteristic facial appearance. Ninety percent have enlarged testes (macroorchidism).

Reference:

Question(s) 38: Behavioral
Discussion:
Carbamazepine and amitriptyline have been used for emotional lability. However, carbamazepine is the better choice in persistent post-traumatic agitation because it does not have the anticholinergic and mild epileptogenic properties possessed by amitriptyline.

Reference:

Question(s) 39: Physiology
Discussion:
A reduced P100 amplitude of a pattern reversal evoked potential on one side is most likely due to decreased visual acuity in that eye.

Reference:

Question(s) 40: Anatomy
Discussion:
Neural crest derivatives include cells of the dorsal root ganglia (involved in transmission of pain) and sympathetic ganglia. These derivatives require nerve growth factor (NGF) for survival. Other neural crest derivatives are Schwann cells and melanocytes.

Reference:
Question(s) 41: Clinical Adult
Discussion:
Romberg's sign, assessed in the course of gait and station testing, invariably is associated with a loss of position sense in the feet and legs (sensory rather than cerebellar ataxia). Subacute combined degeneration produces Romberg's sign by damaging the dorsal columns. Of course, subacute combined degeneration will also produce motor disturbances due to lateral column damage.

Reference:

Question(s) 42: Anatomy
Discussion:
The end organs involved in perception of vibration are Pacinian corpuscles found in subcutaneous connective tissue and in periosteum.

Reference:

Question(s) 43: Pharmacology/Chemistry
Discussion:
Periodic alternating nystagmus (PAN) is characterized by horizontal nystagmus that reverses direction every 90 to 120 seconds. Downbeat nystagmus and square wave jerks may be seen in the null period while the nystagmus changes direction. The nystagmus is little affected by visual fixation, but may be interrupted by vestibular stimuli. PAN has been reported in association with structural abnormalities of the hindbrain, multiple sclerosis, degenerative diseases, acquired metabolic encephalopathies and intoxications with antiepileptic drugs including phenobarbital, primidone and phenytoin. The gamma aminobutyric acid agonist baclofen abolishes PAN in most patients. Alcohol is not known to improve PAN.

Reference:

Question(s) 44: Clinical Pediatrics
Discussion:
The most common presentation of HIV-1 encephalopathy in children is neurodevelopmental delay. Acquired microcephaly, ataxia and seizures are less common but can occur.

Reference:

Question(s) 45: Pharmacology/Chemistry
Discussion:
Imipramine is a tertiary amine tricyclic antidepressant that has potent anticholinergic as well as local anesthetic effects; this increases the risk of cardiac block.

Reference:
Tucker GJ. Psychiatry for the neurologist part A. Continuum 1997 Nov;3(6).

Question(s) 46: Anatomy
Discussion:
The anterior choroidal artery supplies the lateral part of the medial segment of the globus pallidus, the target of pallidotomy in patients with Parkinson's disease. The posterior communicating artery supplies the medial part of the medial segment.

Reference:

Question(s) 47: Pharmacology/Chemistry
Discussion:
Capsaicin binds to the vanilloid receptor (VA1) which is a cation channel also activated by heat and extracellular acidosis. This channel is inhibited by capsazepine.

Reference:
**Question(s) 48: Clinical Adult**

**Discussion:**
In the syndrome of alexia without agraphia, a complete right homonymous hemianopsia is present in many cases, but there are exceptions. Impaired naming and understanding of color names in the presence of intact or nearly intact color vision is common although some patients suffer from an actual impairment of color vision. Mild anomia is common but not always present. In most cases, there are no other aphasic disturbances or abnormalities of the primary motor or sensory systems. The most frequently reported pathology is occlusion of the dominant (left) posterior cerebral artery.

**Reference:**

**Question(s) 49: Pathology**

**Discussion:**
The AIDS dementia complex is characterized neuropathologically by pallor of the cerebral white matter, cerebral atrophy, perivascular macrophages, and multinucleated giant cells. Intense vasculitis is uncommon. Cerebral atrophy and numerous Rosenthal fibers describe Alexander's disease. Spongiform cortical changes are seen in prion diseases and glial cytoplasmic inclusions are seen in multiple system atrophy, not AIDS dementia complex.

**Reference:**

**Question(s) 50: Behavioral**

**Discussion:**
Alexia without agraphia results from damage in the left visual cortex, classically accompanied by damage to the splenium of the corpus callosum.

**Reference:**

**Question(s) 51: Pathology**

**Discussion:**
The chordoma is the most common neural crest-derived tumor of the sacrum. The chordoma is derived from notochordal remnants and occurs predominantly in the sacrum or the clivus, the two ends of the notochord. Myxopapillary ependymomas arising in the filum terminale or tumors of the nerve roots and their coverings such as neurofibromas and schwannomas can attain sufficient size to erode the sacrum but this is uncommon. Teratomas can also erode the sacrum, but do so less commonly than chordomas. Ewing's sarcoma is rare in this location.

**Reference:**

**Question(s) 52: Pharmacology/Chemistry**

**Discussion:**
Nimodipine is a dihydropyridine class of L-type voltage-gated calcium-channel blocker that is used to prevent vasospasm in patients with subarachnoid hemorrhage.

**Reference:**

**Question(s) 53: Behavioral**

**Discussion:**
Of the most common residua of traumatic brain injury, altered personality is the most likely to persist and to interfere with rehabilitation.

**Reference:**
Question(s) 54: Clinical Adult

Discussion:
Progressive multifocal leukoencephalopathy is caused by the JC virus, a papova virus. This disease usually develops in an immune-compromised person, with AIDS being the most common associated disease. Before AIDS, progressive multifocal leukoencephalopathy was most commonly associated with chronic lymphocytic leukemia.

Reference:

Question(s) 55: Anatomy

Discussion:
The area postrema is the only paired circumventricular organ and may act as a chemoemetic "center."

Reference:

Question(s) 56: Physiology

Discussion:
Long term use of chlorpromazine has been reported to lead to generalized paroxysmal bursts in EEGs.

Reference:

Question(s) 57: Physiology

Discussion:
Periodic sharp waves are most likely to be seen in Creutzfeldt-Jakob disease.

Reference:

Question(s) 58: Physiology

Discussion:
Type I fibers are easily recruited fibers that react strongly to nicotinamide adenine dinucleotide dehydrogenase (NADH) but weakly to ATPase. They are the slow twitch fibers and have a high resistance to fatigue.

Reference:

Question(s) 59: Physiology

Discussion:
Procaine block of fusimotor fibers decreases the phasic stretch reflex.

Reference:

Question(s) 60: Pathology

Discussion:
Germinomas are the most frequent tumor in the pineal region. These tumors are composed of large germ cells and smaller lymphocytes.

Reference:

Question(s) 61: Behavioral

Discussion:
Many features of factitious disorder and malingering are behaviorally similar. However, the key differentiating feature is that factitious disorder is motivated by the need to assume the sick role while the motivation for malingering is an external incentive such as avoidance of duty of work or financial compensation.
Question(s) 62: Behavioral
Discussion:
While anxiety is present in many psychiatric disorders, it is not the major criterion in most. An acute and severe anxiety response is, however, the defining characteristic of a simple phobia.

Reference:

Question(s) 63: Pathology
Discussion:
In Wernicke-Korsakoff syndrome, lesions are located in the periventricular gray matter, periaqueductal gray matter, mamillary bodies, and floor of the fourth ventricle.

Reference:

Question(s) 64: Anatomy
Discussion:
Functional neuroimaging shows thermal pain is associated with activation of the anterior cingulate gyrus.

Reference:

Question(s) 65: Physiology
Discussion:
The hair cells are innervated by bipolar neurons of the spiral ganglion of the cochlea.

Reference:

Question(s) 66: Pathology
Discussion:
In neuromyelitis optica, women are affected up to four times more often than men and the clinical course of the disease is frequently rapidly progressive. Devic disease is much more common in Asia, with up to 8% of multiple sclerosis cases in Japan being of the Devic type. Oligoclonal bands are absent from the CSF in most cases. The presence of necrosis in spinal cord virtually defines the entity.

Reference:

Question(s) 67: Clinical Adult
Discussion:
The acute neurotoxic effects of organophosphates include miosis, salivation, sweating and fasciculations.

Reference:

Question(s) 68: Pharmacology/Chemistry
Discussion:
The current guidelines recommend treating ischemic stroke patients who present within 3 hours of symptom onset with recombinant tissue plasminogen activator (rt-PA) at a dose of 0.9 mg/kg (maximum dose 90 mg) within 3 hours of symptom onset with 10% of the dose given as bolus followed by infusion lasting 60 minutes. Exclusion criteria include prior intracranial hemorrhage; history of intracranial neoplasm, aneurysm or arteriovenous malformation; stroke or head trauma within the previous three months; major surgery or biopsy of a parenchymal organ within preceding 14 days; gastrointestinal or urinary bleeding within the previous 21 days; recent myocardial infarction; seizure at onset of stroke; history of known hereditary of acquired abnormal hemostasis; current
use of oral anticoagulants with prothrombin time > 15 seconds, use of heparin in previous 48 hours with prolonged partial thromboplastin time; platelet count < 100,000/mm³, and evidence on CT of major hypodensity or sulcal effacement (> 1/3 of middle cerebral artery territory), and systolic blood pressure > 185 mmHg or diastolic blood pressure > 110 mmHg, and blood glucose < 50 mg/dl or > 400 mg/dl.

Reference:

Question(s) 69: Anatomy
Discussion:
The bulk of fibers from the dentate nucleus project mainly to the ventral lateral thalamic nucleus.

Reference:

Question(s) 70: Clinical Pediatrics
Discussion:
Optic gliomas are the most common CNS tumor for children with neurofibromatosis type 1, but other cranial nerves can be affected by neurofibromas or schwannomas. Astrocytomas, meningiomas, medulloblastomas, ependymomas and hamartomas also occur with increased frequency in patients with neurofibromatosis type 1.

Reference:

Question(s) 71: Physiology
Discussion:
Hypsarrhythmia is a distinctive EEG pattern seen in young children that is often associated with infantile spasms.

Reference:

Question(s) 72: Clinical Pediatrics
Discussion:
Rett syndrome is characterized by developmental regression and autistic behavior in girls after a period of normal development during the first 7-18 months of life. Gait apraxia, truncal ataxia, loss of purposeful use of the hands, microcephaly, hyperventilation and seizures are seen later in the disease. The genetic basis of this disorder has recently been elucidated.

Reference:

Question(s) 73: Physiology
Discussion:
The Achilles tendon reflex is a monosynaptic reflex with the efferent limb in the tibial nerve.

Reference:

Question(s) 74: Anatomy
Discussion:
The Martin-Gruber anastomosis occurs in 15-30% of the population. It consists of a communicating branch from the median nerve to the ulnar nerve in the forearm to supply the first dorsal interosseous, adductor pollicis, and abductor digiti minimi.

Reference:

Question(s) 75: Radiology
Discussion:
The internal cerebral vein, which is one of the major components of the deep draining system of the brain, is located in the roof of the third ventricle between the leaflets of the veli interpositi.
Reference:

Question(s) 76: Clinical Adult
Discussion:
A spastic (or reflex neurogenic) bladder is characterized by precipitant micturition and incontinence. It occurs with upper spinal cord lesions and is associated with other signs of upper motor neuron dysfunction.

Reference:

Question(s) 77: Pharmacology/Chemistry
Discussion:
Two to three percent of patients given the antipsychotic drug clozapine experience bone marrow suppression.

Reference:

Question(s) 78: Physiology
Discussion:
During attacks of hypokalemic periodic paralysis, the amplitude of the compound muscle action potential falls progressively as paralysis develops and may be absent when the muscle membrane becomes completely unexcitable. The sensory nerve conduction studies remain normal as the defect involves the muscle membrane. Between attacks, electromyographic studies show no abnormal spontaneous activity, unless a chronic myopathy develops.

Reference:

Question(s) 79: Physiology
Discussion:
Substance P is probably the transmitter released by small unmyelinated afferent pain fibers.

Reference:

Question(s) 80: Anatomy
Discussion:
The following structures travel through the various foramina: foramen rotundum - maxillary nerve; foramen ovale - mandibular nerve; foramen spinosum - middle meningeal artery; foramen lacerum - internal carotid artery; jugular foramen - glossopharyngeal nerve, vagal nerve, spinal accessory nerve. The internal carotid artery would be affected by a fracture through the foramen lacerum.

Reference:

Question(s) 81: Physiology
Discussion:
The burst suppression pattern has the poorest prognosis. Most of the patients with this type of pattern following an anoxic insult either die or do not regain useful neurologic function.

Reference:

Question(s) 82: Behavioral
Discussion:
One of the most striking characteristics of a patient with Pick's disease is a tendency to make the same statements repetitively (gramophone syndrome).

Reference:
Question(s) 83: Behavioral
Discussion:
All dietary products that contain tyramine are a potential danger to the patient taking MAO inhibitors. Cottage cheese does not contain tyramine.

Reference:

Question(s) 84: Physiology
Discussion:
Activation of membrane phospholipases triggers critical signal transduction pathways for a variety of neurotransmitters, hormones, and other signals. Hydrolysis of phosphatidylinositol diphosphate (PIP2) by phospholipase C results in production of inositol triphosphate (IP3), a signal for calcium release from intracellular stores, and protein kinase C, which mediates phosphorylation of a variety of receptor, cytoskeletal, and nuclear proteins.

Reference:

Question(s) 85: Clinical Pediatrics
Discussion:
This is a classic presentation of benign paroxysmal vertigo, a syndrome that occurs in children in the first five years of life, and is a frequent precursor of migraine. Investigations (MRI, EEG) are normal. The episodes usually resolve by the end of the first decade.

Reference:

Question(s) 86: Physiology
Discussion:
The only predominantly axonal mixed sensorimotor polyneuropathies listed as an option is uremic polyneuropathy. Dejerine-Sottas disease, Guillain-Barré syndrome, and paraproteinemnic neuropathies are predominantly demyelinating diseases, and spinal muscle atrophy affects anterior horn cells.

Reference:

Question(s) 87: Pathology
Discussion:
The pathognomonic feature of high intracranial pressure and transtentorial herniation is a wedge of pressure necrosis in one or both parahippocampal gyri. Hemorrhagic infarction due to entrapment of the posterior cerebral artery, and entrapment of the third cranial nerves also frequently occur. Secondary brainstem hemorrhages and infarctions are maximal in the midbrain and pons, and rarely extend to the medulla.

Reference:

Question(s) 88: Physiology
Discussion:
Sleep markedly enhances the presence of temporal spikes.

Reference:

Question(s) 89: Clinical Adult
Discussion:
Tendency to malignant hyperthermia is an inherited trait. Central core myopathy, in some instances, predisposes to malignant hyperthermia. It may be induced with halothane or succinylcholine and is treated with IV dantrolene.

Reference:
Question(s) 90: Anatomy
Discussion:
Functional imaging studies indicate that the contralateral lateral hemisphere of the cerebellum is activated during cerebral hemispheric cognitive activity.

Reference:

Question(s) 91: Anatomy
Discussion:
Tyrosine hydroxylase is a marker of dopamine-synthesizing cells. A subset of amacrine cells synthesize dopamine, which is important in modulation of retinal circuits in response to variations in light intensity.

Reference:

Question(s) 92: Behavioral
Discussion:
Contralateral neglect has been associated with injury to the right parietal lobe, right frontal lobe, cingulate and thalamus, but not the right occipital lobe.

Reference:

Question(s) 93: Pathology
Discussion:
Peripheral neuropathies can be caused by arsenic, lead, mercury and thallium. Aluminum intoxication can produce an encephalopathy, but does not produce a neuropathy. Copper and bismuth also do not cause peripheral neuropathy.

Reference:

Question(s) 94: Behavioral
Discussion:
Good spontaneous and repetitive affective prosody, as well as spontaneous gesturing with poor affective comprehension of language and gesture is known as transcortical sensory aprosodia.

Reference:

Question(s) 95: Pharmacology/Chemistry
Discussion:
Midodrine is a prodrug that is metabolized in the liver to des-glymidodrine, a potent alpha-1 adrenergic agonist that produces arterial and venous vasoconstriction. Unlike other sympathomimetics, midodrine has a predictable absorption and plasma half life. It is currently the drug of second choice after fludrocortisone for management of neurogenic orthostatic hypotension. The dose is 10-40 mg/day. It should not be administered in the evening given the risk of supine hypertension. Scalp pruritus is a common side effect.

Reference:

Question(s) 96: Pharmacology/Chemistry
Discussion:
Propranolol and primidone are both of proven value in treating essential tremor. In individuals with bronchoconstriction or congestive heart failure, beta blockers such as propranolol are contraindicated.

Reference:
Question(s) 97: Behavioral
Discussion:
Focal cortical abnormalities such as aphasia are not typical features of hydrocephalic dementia.

Reference:

Question(s) 98: Physiology
Discussion:
Acetylcholine is an excitatory neurotransmitter. Strychnine is a competitive inhibitor of the inhibitory neurotransmitter glycine. Excitatory transmitters open ligand gated sodium channels that are structurally different from the voltage gated sodium channels that propagate the action potential. Opening of chloride channels hyperpolarizes the cell membrane and is characteristic of inhibitory neurotransmission. Calcium channel blockers decrease excitatory synaptic transmission.

Reference:

Question(s) 99: Anatomy
Discussion:
The suprachiasmatic nucleus receives direct inputs from the retina (retinohypothalamic tract). These inputs are thought to entrain circadian rhythms on the basis of the light/dark cycle.

Reference:

Question(s) 100: Pathology
Discussion:
Analysis of 14-3-3 protein has diagnostic utility for Creutzfeldt-Jakob disease, and shows high specificity and sensitivity, albeit not 100%, only when done in the appropriate clinical context. A variety of neurological disorders leading to tissue damage such as stroke, encephalitis and trauma are also associated with elevated CSF 14-3-3 protein. The test is done on CSF.

Reference:

Question(s) 101: Anatomy
Discussion:
The posterior columns contain ascending fibers from spinal ganglia (first order neurons) concerned with tactile and kinesthetic sense. The fasciculus gracilis contains the fibers from sacral, lumbar and the lower six thoracic dorsal roots. The second order neurons for these fibers are located in the nucleus gracilis.

Reference:

Question(s) 102: Pathology
Discussion:
Both granulovacuolar degeneration and Hirano bodies are prominent in Ammon's horn of the hippocampus.

Reference:

Question(s) 103: Clinical Adult
Discussion:
Disturbance of the S1 nerve root results in pain radiating down the posterior aspect of the lower extremity, sensory disturbance of the little toe, lateral foot, and most of the sole of the foot, weakness in several muscles including the gastrocnemius and soleus muscles, and reduction or absence of the ankle jerk.

Reference:
**Question(s) 104: Clinical Pediatrics**

**Discussion:**
The congenital myasthenic syndromes are not related to an immune process, but are caused by genetic defects affecting the neuromuscular junction. These include defects in acetylcholine synthesis and packaging (familial infantile MG), end-plate deficiency of acetylcholinesterase, acetylcholine receptor deficiency, and the slow channel syndrome.

**Reference:**

**Question(s) 105: Pathology**

**Discussion:**
Brains at autopsy taken from schizophrenic patients show an increase, not decrease, in ventricular size, especially of the temporal horns of the ventricles due to loss of cortical gray matter in medial temporal lobes. Although cerebellar abnormalities of the vermis were reported in a single study, they were not verified in subsequent work. More dramatic changes such as heterotopias and major gyral abnormalities such as polymicrogyria are not generally seen in schizophrenic patients, either premortem on radiographic studies, or post-mortem.

**Reference:**

**Question(s) 106: Pathology**

**Discussion:**
A heavy metal screen tests for chronic heavy metal poisoning and exposure, and the test is almost always done on urine. Blood screening for heavy metals, such as arsenic, is done only for the uncommon situation where acute poisoning is suspected. Since arsenic is rapidly removed from blood, blood is not the specimen of choice to rule out chronic arsenic exposure. The heavy metal screen will almost always include tests for lead, arsenic, and inorganic mercury, but toluene and methanol are not heavy metals and must be analyzed by separate methods. Organic mercury poisoning is a unique situation where tests need to be done on whole blood since this form of mercury is mainly located in erythrocytes.

**Reference:**

**Question(s) 107: Physiology**

**Discussion:**
The combination of fast activity (12-14 Hz activity) and delta slowing is most often seen as an anesthetic effect or drug overdose.

**Reference:**

**Question(s) 108: Clinical Adult**

**Discussion:**
Although memory impairment has been attributed to frontal lobe dysfunction, true amnesia is uncommon with frontal lobe lesions. Inattention, distractibility, poor motivation, and impaired strategy formation lead to the forgetfulness associated with frontal lobe lesions. Lesions of the temporal lobe have been associated with pure amnesia and lesions of the occipital and parietal lobes or cerebellum do not produce forgetfulness.

**Reference:**
Question(s) 110: Behavioral
Discussion:
Patients with obsessive-compulsive disorder (OCD) usually do have insight that their obsessions and compulsions are excessive and unreasonable. The condition may be seen associated with Tourette’s syndrome. PET and SPECT studies have demonstrated abnormally high metabolism and blood flow respectively in the orbital frontal and striatal areas of the brain. SSRI's (selective serotonin reuptake inhibitors) such as fluvoxamine may be effective in ameliorating OCD symptoms.
Reference:

Question(s) 111: Clinical Pediatrics
Discussion:
Ataxia-telangiectasia is an autosomal recessive disorder characterized by ataxia, oculocutaneous telangiectasias and immunoincompetency. Choreoathetoid movements are seen even at an early age.
Reference:

Question(s) 112: Physiology
Discussion:
The majority of retinal axons terminate in the lateral geniculate nucleus, the principal subcortical region that processes perceptual visual information.
Reference:

Question(s) 113: Physiology
Discussion:
The F-wave is produced by supramaximal stimulation, is easily derived from most major nerves, and is involved early in Guillain-Barré syndrome.
Reference:

Question(s) 114: Pathology
Discussion:
Sturge-Weber disease is characterized by a meningeal vascular malformation and calcification in the second and third layers of the underlying cortex.
Reference:

Question(s) 115: Anatomy
Discussion:
The amygdala plays a critical role in emotional responses, including conditioned fear.
Reference:

Question(s) 116: Anatomy
Discussion:
The intralaminar nuclei, including the centromedian/parafascicular complex, provide glutamatergic inputs to the striatum, particularly to its local cholinergic neurons. This constitutes a feedback loop between the striatum, pallidum, and thalamus.
Reference:

Question(s) 117: Clinical Adult
Discussion:
A majority of epileptics experience significant depression at some time during the course of the disease. The suicide rate is over five times higher in epileptics than in the population as a whole.
Reference:

Question(s) 118: Behavioral
Discussion:
Munchausen’s syndrome is commonly manifested by pseudologia phantastica. This is a form of lying in which the person appears to believe in the reality of their own fantasies and acts on them. This would represent the repeated feigning of illness. Ailurophobia is the dread of cats and noesis is the sense that one has been chosen to lead and command.

Reference:

Question(s) 119: Behavioral
Discussion:
Nefazodone does not suppress REM sleep; it actually may increase REM sleep.

Reference:

Question(s) 120: Anatomy
Discussion:
Neurilemmal sheath cells of all peripheral nerves, ganglia capsule cells, sympathetic ganglia, and chromaffin cells are all neural crest derivatives. Astrocytes are derived from neuroectoderm.

Reference:

Question(s) 121: Pathology
Discussion:
The CSF findings of pressure elevation, hypoglycorrhachia, mildly elevated protein, and mononuclear pleocytosis are most consistent with chronic meningitis due to fungi or mycobacteria. The reduced glucose is especially helpful in distinguishing viral infections such as herpes encephalitis and progressive multifocal leukoencephalopathy from chronic fungal or tuberculous meningitis.

Reference:

Question(s) 124: Anatomy
Discussion:
The optic nerve is part of the central nervous system and, hence, its myelin is derived from oligodendroglia as opposed to Schwann cells that provide myelin to the other cranial and peripheral nerves.

Reference:

Question(s) 122: Clinical Adult
Discussion:
A lesion of the third nerve nucleus produces weakness of muscles innervated by that third nerve, but also bilateral weakness of the superior rectus, levator palpebrae, and pupillary constrictor muscles. A lesion of the third nerve itself does not affect any contralateral muscles. Thus, ptosis of the left eyelid could result from lesions of the left third nerve or either left or right third nerve nucleus. Cortical lesions have also been reported to cause either contralateral or bilateral ptosis on occasion.
Reference:

Question(s) 125: Pathology
Discussion:
Polymerase chain reaction (PCR) is a now widely available molecular diagnostic method that permits the selective amplification of target sequences of DNA. For the diagnosis of infectious diseases, primer sequences are selected that permit the targeting of unique organism-specific DNA sequences; if the organism's DNA is present in the tissue or CSF, then an infection is present. PCR may be useful for the diagnosis of mycobacteria and some viral diseases such as cytomegalovirus, progressive multifocal leukoencephalopathy, and Herpes simplex in which cultures are slow or cumbersome.

Reference:

Question(s) 126: Pathology
Discussion:
Most of the rare familial Alzheimer's disease kindreds show autosomal dominant inheritance patterns. While mutations in the amyloid precursor protein gene on chromosome 21 were the first to be discovered, mutations in this gene account for less than 10% of all familial Alzheimer disease pedigrees. Presenilin mutations on chromosomes 1 and 14 may account for more of the familial cases. How presenilins exactly participate in causing Alzheimer's disease is unknown, but ultimately they mediate an increase in the APP/ßA4 amyloid protein. Late-onset Alzheimer's disease may be influenced by a gene on chromosome 19 that encodes for allelic variants of ApoE proteins.

Reference:

Question(s) 127: Behavioral
Discussion:
Decreased repetition is noted in all the presylvian aphasias, including Broca's, Wernicke's, conduction, and global aphasia. Repetition is generally spared in the transcortical aphasias.

Reference:

Question(s) 128: Clinical Adult
Discussion:
For patients with relapsing and remitting multiple sclerosis, treatment with interferon ß1b reduces the frequency of relapses, but does not eliminate them.

Reference:

Question(s) 129: Physiology
Discussion:
The most sensitive test for myasthenia gravis is single fiber EMG of the frontalis muscle.

Reference:

Question(s) 130: Pharmacology/Chemistry
Discussion:
Thrombotic thrombocytopenic purpura has been reported rarely with ticlopidine and clopidogrel. Ticlopidine may also result in leucopenia. Aspirin, dipyridimole, low molecular weight heparin and sulfinpyrazone have not been associated with this complication.

Reference:
Question(s) 131: Clinical Adult
Discussion:
The syndrome of spinocerebellar degeneration is most characteristic of prolonged chronic vitamin E deficiency.

Reference:

Question(s) 132: Pharmacology/Chemistry
Discussion:
Unlike other antiepileptic drugs, gabapentin is not metabolized in the liver and therefore is less likely to interfere with the metabolism of porphyrins or other drugs.

Reference:

Question(s) 133: Physiology
Discussion:
Small sharp spikes, which constitute a benign EEG pattern, are seen in adults and usually occur during drowsiness and light sleep. They are best seen in the temporal electrodes.

Reference:

Question(s) 134: Anatomy
Discussion:
The superior cervical ganglion provides noradrenergic inputs to the pineal gland. Stimulation of beta adrenergic receptors in the pineal gland stimulates synthesis of melatonin.

Reference:

Question(s) 135: Clinical Pediatrics
Discussion:
Glutaric aciduria typically presents with macrocephaly, with or without developmental delay, hypotonia or a hyperkinetic movement disorder (the latter may slowly progress or present acutely as a catastrophic deterioration, usually in the context of intercurrent infection). Treatment with carnitine may be effective in preventing deterioration. Krabbe's disease and Rett syndrome are associated with microcephaly; obstructive hydrocephalus is associated with enlarged ventricles; and MERRF (Myoclonus Epilepsy with Ragged Red Fibers) is associated with ataxia, myoclonus, seizures and myopathy.

Reference:

Question(s) 136: Physiology
Discussion:
The presence of periodic sharp waves in a neonate with seizures is strongly suggestive of neonatal herpes simplex encephalitis.

Reference:

Question(s) 137: Behavioral
Discussion:
Speech remains intact as language deteriorates with advancing Alzheimer's dementia, eventually producing an aphasia in which the patient is fluent and paraphasic, their speech is empty, and they have limited comprehension but repeat well, which is typical of transcortical sensory aphasia.

Reference:
**Question(s) 138**: Pharmacology/Chemistry

**Discussion:**
Methanol intoxication results in severe metabolic acidosis due to oxidation of methanol to formic acid. Dilated, unreactive pupils and reduced vision are typical, due to destruction of retinal ganglion cells. Bicarbonate is the keystone of treatment of methanol poisoning. Concomitant administration of ethanol, a competitive substrate of alcohol dehydrogenase, may have some benefit. A loading dose (0.6 g/kg) should be administered as soon as the diagnosis of methanol poisoning is made.

**Reference:**

**Question(s) 139**: Pharmacology/Chemistry

**Discussion:**
Many viper, pit viper, and Australasian elapid venoms contain factors that produce a consumption coagulopathy and defibrination, which may persist for some weeks. This may result in intracranial hemorrhage, classically occurring several days following the bite.

**Reference:**

**Question(s) 140**: Pathology

**Discussion:**
Medulloblastoma is especially prone to widespread leptomeningeal metastases. Occasionally, this can be seen with glioblastoma and ependymoma.

**Reference:**

**Question(s) 141**: Anatomy

**Discussion:**
The radial glia participate in cell-cell interactions necessary for neuronal migration.

**Reference:**

**Question(s) 142**: Anatomy

**Discussion:**
The ciliary muscle receives postganglionic parasympathetic innervation via the third nerve. The superior cervical ganglion provides the sympathetic innervation to the facial sweat glands, carotid artery, pineal gland, and tarsal muscle.

**Reference:**

**Question(s) 143**: Physiology

**Discussion:**
K+ influx during a nerve action potential limits the duration of the action potential.

**Reference:**

**Question(s) 144**: Pathology

**Discussion:**
Paranodal giant axonal enlargements or "balloons" are seen in neuropathy due to n-hexane or methyl n-butyl ketone exposure. They are also encountered in hereditary giant axonal neuropathy.

**Reference:**

**Question(s) 145**: Behavioral

**Discussion:**
A lesion affecting the left occipital lobe and the splenium of the corpus callosum will produce a right visual field defect and alexia without agraphia. A disturbance of color recognition, either color agnosia or achromatopsia, may or may not be present.
Reference:

Question(s) 146: Physiology
Discussion:
Sawtooth waves are seen on the EEG during REM sleep.

Reference:

Question(s) 147: Pathology
Discussion:
In normal pressure hydrocephalus, even though the lateral ventricles are enlarged, the mean CSF pressure at lumbar puncture is not elevated. Known possible antecedent causes include head injury, subarachnoid (but not subdural) hemorrhage, or meningitis, although in some cases no cause can be identified even after careful autopsy examination. It has been suggested that the causative mechanism in normal pressure hydrocephalus is an increase in CSF outflow resistance.

Reference:

Question(s) 148: Anatomy
Discussion:
The medial preoptic area plays an important role in regulation of gonadotropin secretion. It includes the sexually dimorphic nucleus, which contains twice as many neurons in men as in women.

Reference:

Question(s) 149: Anatomy
Discussion:
Dressing apraxia is seen in non-dominant parietal lobe lesions.

Reference:

Question(s) 150: Anatomy
Discussion:
The mammillothalamic tract projects to the anterior nuclear group of the thalamus.

Reference:

Question(s) 151: Pharmacology/Chemistry
Discussion:
Fabry's disease is an X-linked defect in the alpha-galactosidase. It is characterized by painful peripheral neuropathy with autonomic manifestations, a typical rash in the lower half of the body, and accumulation of glycolipids in the endothelium of cerebral vessels and glomerular arterioles.

Reference:

Question(s) 152: Physiology
Discussion:
Nonconvulsive status epilepticus can be prolonged and is associated with fairly abrupt deterioration in mental function and a paroxysmal EEG. These features differentiate non-convulsive status from the dementias.

Reference:
Question(s) 153: Clinical Adult

Discussion:
The pupils react normally in Horner’s syndrome, while lesions of the parasympathetic pathway are associated with poor or absent reaction of the pupil to light.

Reference:

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Question(s) 154: Clinical Adult

Discussion:
Post-partum women are at increased risk for sagittal sinus thrombosis due to their hypercoagulable state. Headache, delirium, and focal seizures are frequent early symptoms.

Reference:

Question(s) 155: Physiology

Discussion:
The N13 potential is generated at the cervical cord level, and an absent response would suggest a lesion involving the cervical cord.

Reference:

Question(s) 156: Clinical Adult

Discussion:
The combination of early visual hallucinations, markedly fluctuating mental status, sensitivity to neuroleptics and lack of response to levodopa are characteristic of dementia with Lewy bodies.

Reference:

Question(s) 157: Anatomy

Discussion:
Sonic hedgehog is a diffusible factor produced by the notochord and the floor plate, and is critical for differentiation of the ventral portions of the neural tube, including cranial and spinal motor neurons.

Reference:

Question(s) 158: Pathology

Discussion:
Bromocriptine is a dopamine agonist that functions by the same mechanism as Prolactin Inhibitory Factor (PIF, which is dopamine), to inhibit prolactin synthesis and release by prolactin-secreting pituitary adenomas (prolactinomas). Prolactinoma size reduction is effected by decreasing tumor cell cytoplasmic volume without actual tumor killing; therefore, the prolactinoma will re-expand if bromocriptine is withdrawn. Bromocriptine has no significant role in neuro-oncology other than in suppression of prolactin-secreting tumors.

Reference:

Question(s) 159: Anatomy

Discussion:
The tuberomammillary nucleus contains histamine-synthesizing neurons that project to the cerebral cortex and are involved in arousal mechanisms.

Reference:
Question(s) 160: Anatomy
Discussion:
Axons from neurons in the gracile and cuneate nuclei sweep ventromedially as the internal arcuate fibers before ascending as the medial lemniscus. They are involved with tactile and kinesthetic sense. The other structures listed are components of the auditory pathway and lesions of these will affect auditory evoked responses.

Reference:

Question(s) 161: Anatomy
Discussion:
The subthalamic nucleus modulates (suppresses) ipsilateral basal ganglionic activity, which in turn modulates cortical motor outflow to the contralateral effector muscles. In general, basal ganglionic lesions have contralateral motoric effects. In the case of the subthalamic nucleus, contralateral hemiballismus – high amplitude flinging appendicular movements – ensues. A right subthalamic infarct leads to left sided hemiballismus.

Reference:

Question(s) 162: Pathology
Discussion:
Significant keys to pathogenesis in multiple sclerosis may be associated with the topographic features of demyelinative plaques. Older plaques tend to be sharply, not poorly, demarcated from surrounding brain. In two thirds of cases, spinal cord, cerebrum, and optic nerves are equally involved, with sparing of one or another of these regions in less than 15% of autopsied patients. Gray matter, including cerebral cortex, is not spared even though the largest or most grossly visible plaques are in deep white matter. Plaques extend along veins, not arteries, in finger-like projections called Dawson's fingers. Subpial plaques are classically wedge-shaped, with a broad base near CSF pathways.

Reference:

Question(s) 163: Pathology
Discussion:
Connective tissue (collagen) in the brain is produced only by fibroblasts of blood vessels' adventitia. Glia do not produce collagen.

Reference:

Question(s) 164: Pharmacology/Chemistry
Discussion:
Pemoline and methylphenidate have both been of some benefit in the treatment of attention deficit disorder in children. Clonidine, a central agonist of the alpha-2 adrenoreceptors, is helpful to decrease hyperactivity and impulsivity in these patients.

Reference:

Question(s) 165: Radiology
Discussion:
Perfusion-weighted MR imaging (PWI) allows an assessment of perfusion of the brain microvasculature. PWI requires intravenous gadolinium contrast that causes a paramagnetic susceptibility effect. MRI signal declines as gadolinium transits the microvasculature. One of the uses of PWI is to complement the information obtained by diffusion-weighted MRI (DWI) in acute stroke patients. A variety of perfusion properties of the contrast bolus can be calculated, such as cerebral blood volume, cerebral blood flow, mean transit time, time-to-peak, and time of arrival. Flow-related enhancement is used in MR angiography. Changes in venous oxygenation are used in functional MRI. Magnetization transfer pulses, such as those used in MR angiography, measure magnetization transfer effects. MR spectroscopy measures brain metabolites such as choline in parts per million (ppm).
Reference:

Question(s) 166: Behavioral
Discussion:
Subcortical dementia is characterized clinically by psychomotor slowing, forgetfulness, cognitive decline, visuospatial impairment and personality changes, especially in mood. Bradyphrenia (slowness of mental processing) is very common.

Reference:

Question(s) 167: Behavioral
Discussion:
No single diagnostic approach provides an absolute diagnosis of Alzheimer's disease. At present, the most accurate diagnostic tool remains the carefully performed evaluation by a knowledgeable clinician.

Reference:

Question(s) 168: Behavioral
Discussion:
Inability to produce melody in verbal output, amelodia, produces a significant loss of non-verbal affective expression (affective motor aprosodia). The causative lesion is most commonly found in the posterior inferior frontal lobe of the right hemisphere, the region in the right frontal lobe analogous to Broca's area in the left.

Reference:

Question(s) 169: Anatomy
Discussion:
The sulcus limitans marks the lateral junction of basal and alar plates.

Reference:

Question(s) 170: Anatomy
Discussion:
The cholinergic input to the thalamic relay nuclei arises from the mesopontine tegmentum, including the pedunculopontine and laterodorsal tegmental nuclei. The basal forebrain nuclei provide cholinergic inputs to the cerebral cortex rather than the thalamic relay nuclei. The reticular nuclei contain local neurons that are GABAergic. Corticothalamic inputs are glutamatergic.

Reference:

Question(s) 171: Behavioral
Discussion:
A variety of behavioral problems can occur during the course of Alzheimer's disease. Almost never, however, does the Alzheimer patient show the rapid thought processing that characterizes mania.

Reference:

Question(s) 172: Physiology
Discussion:
Electrophysiologic findings seen in acquired demyelinating neuropathies include slowing of conduction velocity, prolonged distal motor latencies, prolonged F-wave latencies and conduction block/abnormal temporal dispersion.

Reference:
Question(s) 173: Anatomy
Discussion:
The thalamus is supplied mainly by the branches of the posterior cerebral, posterior communicating, and posterior choroidal arteries.

Reference:

Question(s) 174: Pharmacology/Chemistry
Discussion:
Bupropion can reduce the seizure threshold, particularly in patients with bulimia and anorexia nervosa.

Reference:
Tucker GJ. Psychiatry for the neurologist part A. Continuum 1997 Nov;3(6).

Question(s) 175: Radiology
Discussion:
Normal brain myelination is a dynamic process that begins during fetal life and continues after birth in a predictable manner. The progress of the myelination is seen as changes in white matter signal on the MRI image. In the term infant, myelination is best assessed on the T1-weighted image where it is seen as high signal. Normal adult patterns are seen by 18 months of age. Some areas, such as the white matter dorsal and superior to the ventricular trigone, have persistent high signal on T2-weighted images, which should be considered normal. These association fibers often become myelinated by the second or third decade of life.

Reference:

Question(s) 176: Clinical Adult
Discussion:
In tuberculous meningitis, CSF protein is typically elevated, glucose is low, and cells are moderately increased (predominantly lymphocytes).

Reference:

Question(s) 177: Pharmacology/Chemistry
Discussion:
The musculocutaneous and median nerves are branches of the lateral cord of the brachial plexus.

Reference:

Question(s) 178: Clinical Adult
Discussion:
Toxoplasma gondii is a major cause of encephalitis in patients with AIDS. The characteristic CT scan shows multiple (occasionally single) ring and/or nodular enhancing lesions. The use of MRI may enhance detection of lesions.

Reference:

Question(s) 179: Physiology
Discussion:
Somatosensory evoked potentials can be elicited in the absence of a recordable peripheral sensory nerve action potential because the CNS can amplify the incoming responses.

Reference:

Question(s) 180: Behavioral
Discussion:
Ganser syndrome is sometimes known as the "syndrome of approximate answers," or "pseudostupidity." Most often seen in psychiatric disorders, it has also been reported following neurologic conditions such as head trauma and neurosyphilis.
Reference:  

Question(s) 181: Behavioral  
Discussion:  
Patients with auditory receptive aphasia experience difficulty verbalizing their thoughts because of output errors at the level of words (semantic paraphasias) and parts of words (phonemic paraphasias).

Reference:  

Question(s) 182: Pharmacology/Chemistry  
Discussion:  
Topiramate and zonisamide are weak inhibitors of the carbonic anhydrase, and therefore may increase urinary pH and decrease urinary citrate excretion. Both effects may predispose to kidney stones. In addition, both drugs have been associated with impaired concentration and memory, and could potentially affect school performance.

Reference:  

Question(s) 183: Anatomy  
Discussion:  
A right third nerve palsy and left hemiparesis are seen in a right ventral rostral mesencephalic (midbrain) syndrome of Weber.

Reference:  

Question(s) 184: Radiology  
Discussion:  
Of the medical devices and conditions listed, only cardiac pacemakers constitute an absolute contraindication for MRI. Many heart valves are MR-compatible, particularly those produced in the last decade. Middle ear prosthesis are not considered hazardous to patients, but the device itself can be damaged. An MRI can be performed in a patient with an aneurysm clip depending on the material used for the clip. Pregnancy is a relative but not absolute contradiction to MRI. It should be used with caution in the first trimester.

Reference:  

Question(s) 185: Clinical Pediatrics  
Discussion:  
Instability of the first and second cervical vertebrae or atlantoaxial dislocation is seen in 15-40% of patients with Down’s syndrome. If these individuals plan to participate in athletic competition, they should have imaging of the cervical spine.

Reference:  

Question(s) 186: Anatomy  
Discussion:  
The ventral posterolateral nucleus projects to areas three, one and two, the primary somesthetic area of the parietal lobe.

Reference:  

Question(s) 187: Behavioral  
Discussion:  
Aphasia is indicative of cortical dysfunction and is not characteristic of subcortical disorders.

Reference:  
Question(s) 188: Pharmacology/Chemistry
Discussion:
The psychosis induced by dopaminergic drugs in Parkinson’s disease may be hard to manage. The traditional dopamine blocking agent like haloperidol and chlorpromazine may worsen parkinsonism. Olanzapine and risperidone are less likely to do so but are not entirely free of this problem. Clozapine is probably the most suitable drug but requires white blood count monitoring. Quetiapine, an atypical neurolptic, can be useful in this setting.

Reference:

Question(s) 189: Clinical Adult
Discussion:
Transient episodes of monocular or bilateral visual loss lasting seconds are characteristic of increased intracranial pressure and may be precipitated by changes in posture.

Reference:

Question(s) 190: Clinical Adult
Discussion:
Diazepam is the mainstay of treatment for the stiff-person syndrome, which occurs about as often in women as in men. Onset in adult life, proximal distribution of stiffness, development of lordosis, and precipitation by motion or emotion are typical. Diabetes and organ-specific autoimmune disorders are common. Seizures sometimes occur.

Reference:

Question(s) 191: Pathology
Discussion:
Contrecoup contusions occur in the brain away from the point of impact and do not directly underlie the site of skull fractures. Orbital surfaces of the frontal lobe are the most common location and contrecoup contusions at this site usually occur following a fall on the occiput with the sudden deceleration of the head. parasagittal contusions, also called gliding contusions, occur in association with diffuse brain damage and diffuse axonal injury.

Reference:

Question(s) 192: Anatomy
Discussion:
Taste sensibility is represented in the parietal operculum (area 43) and adjacent parainsular cortex.

Reference:

Question(s) 193: Anatomy
Discussion:
Erb-Duchenne upper plexus paralysis occurs secondary to damage to the fifth and sixth cervical roots or upper trunk of the brachial plexus. It is a common deficit and is usually due to traumatic separation of the head and shoulder but may also be due to pressure on the shoulder, birth injuries, or idiopathic plexitis.

Reference:

Question(s) 194: Physiology
Discussion:
A femoral neuropathy causes quadriceps weakness with sparing of the adductors and iliopsoas muscles, absent or decreased ankle jerk, and loss of sensation of the anteromedial thigh and medial lower leg.

Reference:
Question(s) 195: Clinical Adult
Discussion:
The onset of orofacial dyskinesias with lingual and oral dystonia in a 30-year-old patient is characteristic of neuroacanthocytosis which may also be associated with chorea and peripheral polyneuropathy.

Reference:

Question(s) 196: Physiology
Discussion:
In normal subjects, the maximum difference between right and left median nerve motor distal latencies (stimulating at the wrist) is 0.7 msec. The maximum difference for median sensory distal latencies is 0.5 msec. When the differences between right and left are greater than these values, one can confidently diagnose median mononeuropathy at the wrist (carpal tunnel syndrome) on the side with the longer latencies. The difference between right and left is not helpful in assessing bilateral compression. Conduction velocity between elbow and wrist is not affected in this condition.

Reference:

Question(s) 197: Pathology
Discussion:
Paraneoplastic syndromes predominantly affect the brainstem, cerebellum, dorsal root ganglia and medial temporal lobe.

Reference:

Question(s) 198: Anatomy
Discussion:
A lesion of the accessory cuneate nucleus would spare pain and temperature. Cells of origin of the lateral spinothalamic tract are present in laminae I, IV, and V of the dorsal horn. They project to ventral posterolateral and intralaminar and posterior nuclei of the thalamus. Further projection to the cortex is to areas three, one, and two and to the secondary somatic sensory area.

Reference:

Question(s) 199: Anatomy
Discussion:
The sacral parasympathetic nucleus, located in the intermediolateral cell column at S2-S4 levels of the spinal cord, contains the preganglionic parasympathetic neurons innervating the bladder detrusor muscle. A lesion of the sacral parasympathetic nucleus can result in a hypotonic bladder.

Reference:

Question(s) 200: Anatomy
Discussion:
The abducens nerve lies immediately adjacent to the internal carotid artery in the cavernous sinus.

Reference:

Question(s) 201: Behavioral
Discussion:
Both auditory and visual hallucinations, persecutory delusions, and ideas of reference are seen in both temporal lobe epilepsy (TLE) and schizophrenia. A positive family history of schizophrenia, schizoid personality, or schizotypal personality is often present in schizophrenia but usually not in TLE. Affect is better preserved in TLE than in schizophrenia.

Reference:
Question(s) 202: Behavioral
Discussion:
Attachment theory investigates secure and insecure parent-child bonding in early life and its consequences for later development.

Reference:

Question(s) 203: Pathology
Discussion:
Dolichoectasia (fusiform aneurysm) most commonly affects the supraclinoid segment of the internal carotid artery and the basilar artery, and is seen in patients with advanced cerebral atherosclerosis. Berry aneurysms are associated with both coarctation of the aorta and arteriovenous malformations (3-9% of patients with intracranial AVMs have berry aneurysms), but fusiform aneurysms are not. Fungi cause arteritis and aneurysmal change but not the elongated vessel distention of dolichoectasia. Human immunodeficiency virus is associated with, but perhaps not the direct cause of, smaller vessel vasculitis.

Reference:

Question(s) 204: Pharmacology/Chemistry
Discussion:
Paraneoplastic cancer-associated retinopathy (CAR) occurs in patients with small cell lung carcinoma, melanoma, breast carcinoma, and a variety of gynecological tumors. Anti-CAR antibodies stain the inner and outer segment layers and the outer nuclear layer of the retina. These antibodies react with the 23-kd calcium-binding protein recoverin, which functions in the light adaptation process associated with the phototransduction cascade and initiated by light activation of rhodopsin.

Reference:

Question(s) 205: Pathology
Discussion:
Kinetic energy as defined by classical mechanics is equal to one-half the product of the mass of the projectile multiplied by the square of its velocity. Kinetic energy increases linearly with projectile mass, but increases as the square of the velocity; therefore, velocity is the single most important determinant of tissue injury during missile passage. Tumble and fragmentation can determine the rate at which kinetic energy is transferred and can alter the amount of damage done by a bullet possessed of a given kinetic energy.

Reference:

Question(s) 206: Pharmacology/Chemistry
Discussion:
Hyperphosphorylated tau is the primary component of the paired helical filaments. Tau hyperphosphorylation can affect microtubule polymerization and thus intracellular transport.

Reference:

Question(s) 207: Behavioral
Discussion:
Alexia without agraphia is often seen with lesions of the left occipital cortex that frequently are seen to extend into the splenium of the ipsilateral corpus callosum. Because of the occipital cortical involvement and splenial involvement, there may be associated color anomia and right homonymous hemianopia.
Reference:

Question(s) 208: Clinical Adult Discussion:
Severe pain and muscle wasting in the thigh (asymmetrical proximal motor neuropathy) is a relatively frequent occurrence in diabetes.

Reference:

Question(s) 209: Clinical Adult Discussion:
Herpes simplex encephalitis is the most common form of acute sporadic encephalitis. It is characterized by lymphocytic pleocytosis and evidence of mild hemorrhage in the CSF, and frequent focal encephalopathic symptoms.

Reference:

Question(s) 210: Clinical Pediatrics Discussion:
ADHD and obsessive-compulsive symptoms are often associated with Tourette's syndrome. Boys are more often affected than girls. An increase in seizures is not seen. Coprolalia is not necessary for the diagnosis.

Reference:

Question(s) 211: Physiology Discussion:
Radiating dysesthesias into the little finger suggest a C-8 radiculopathy. The flexor carpi ulnaris is largely innervated by the C-8 nerve roots.

Reference:

Question(s) 212: Pharmacology/Chemistry Discussion:
Paralytic shell-fish poison (saxitoxin) resembles tetrodotoxin in its ability to reversibly bind to membrane voltage-gated sodium channels.

Reference:

Question(s) 213: Anatomy Discussion:
The synapse of the afferent axons for muscle stretch reflexes is located in the anterior (ventral) horn.

Reference:

Question(s) 214: Behavioral Discussion:
Selective serotonin reuptake inhibitors (SSRI) have emerged as the treatment of choice for obsessive compulsive disorder.

Reference:

Question(s) 215: Behavioral Discussion:
An aphasia is fluent if: word output per minute is high; there are often five or more words per phrase; content per phrase is low; paraphasias are present; and speech is non-dysarthric with normal prosody.

Reference:
Question(s) 216: Anatomy
Discussion:
Axons of the Purkinje cells are the only ones to emerge from the cerebellar cortex and project to the deep cerebellar nuclei.

Reference:

Question(s) 217: Pathology
Discussion:
Corticobasal ganglionic degeneration has ballooned achromatic cortical neurons. Cortical Lewy bodies are seen in diffuse Lewy body dementia. Granulovacuolar degeneration is seen in aging and Alzheimer's disease. Hirano bodies are seen in Alzheimer's disease and Cowdry A inclusions are usually intranuclear viral inclusions.

Reference:

Question(s) 218: Physiology
Discussion:
Reduced cerebral blood flow is associated with arterial partial pressure of carbon dioxide of 25 mm Hg.

Reference:

Question(s) 219: Anatomy
Discussion:
The tibialis anterior, peroneus brevis, short head of biceps femoris and tibialis posterior are all innervated by L5. The abductor hallucis is innervated by S1.

Reference:

Question(s) 220: Physiology
Discussion:
Electrocerebral silence on EEG must be established by careful attention to technical detail. Electrocerebral silence does not exclude potential for recovery as it may be seen in reversible drug intoxication. Conversely, while brain death is defined as the irreversible loss of all brain function and therefore implies electrocerebral silence, it is not necessary to document electrocerebral silence before pronouncing death. The criteria for recording EEGs in brain death include recording at a sensitivity of 2 microvolts/mm and long interelectrode distances.

Reference:

Question(s) 221: Behavioral
Discussion:
Atypical parkinsonism, retrocollis, subcortical dementia, and supranuclear vertical gaze palsy are all features of progressive supranuclear palsy (PSP). Transcortical motor aphasia is not a characteristic clinical feature of PSP.

Reference:

Question(s) 222: Clinical Adult
Discussion:
Cluster headache is characterized by excruciating unilateral pain, usually centered on one eye or temple, lasting 30 to 120 minutes. Patients average one or two headaches a day for a period of four to eight weeks, and the attacks tend to recur at the same time each day, often during REM sleep. An ipsilateral partial Horner's syndrome often accompanies the pain.

Reference:
**Question(s) 223:** Physiology

**Discussion:**
The clinical and EEG findings in supplementary motor seizures are distinctive, with tonic posturing, focal symptoms in the legs, not hands, and usually preserved consciousness. The seizures are often misdiagnosed as psychogenic seizures and the EEG findings are often subtle or absent.

**Reference:**

**Question(s) 224:** Behavioral

**Discussion:**
A lesion in the left angular gyrus, which is situated in the inferior parietal lobule, can cause alexia and agraphia. The latter feature is a component of Gerstmann's syndrome.

**Reference:**

**Question(s) 225:** Anatomy

**Discussion:**
On the side of a spinal cord hemisection there would be an upper motor neuron syndrome, greatly impaired discriminatory tactile sense, loss of kinesthetic sense, and reduced muscle tone. Contralateral to the lesion there would be loss of pain and temperature due to interruption of the ascending spinothalamic tracts.

**Reference:**

**Question(s) 226:** Physiology

**Discussion:**
The generalized 3 Hz spike-and-wave pattern is seen in children with absence seizures from three to ten years of age.

**Reference:**

**Question(s) 227:** Pharmacology/Chemistry

**Discussion:**
In proton magnetic resonance spectroscopy, the N-acetyl aspartate peak is an index of neuronal integrity; lactate acid of anaerobic glycolysis, phosphocreatine of energy metabolism, and choline of membrane synthesis and degradation.

**Reference:**

**Question(s) 228:** Behavioral

**Discussion:**
Anton's syndrome, the denial of blindness, is typically associated with bilateral posterior cerebral artery territory infarction producing "cortical" blindness plus memory impairment.

**Reference:**

**Question(s) 229:** Anatomy

**Discussion:**
The insular cortex receives visceral nociceptive input via the ventromedial posterior (VMPO) thalamic nucleus.

**Reference:**

**Question(s) 230:** Clinical Adult

**Discussion:**
The anterior spinal artery syndrome is characterized by loss of motor function, bladder and bowel control, and pain and temperature sensation below the level of the lesion, with preservation of position and vibration sensation. It most commonly results from aortic dissection, aortic surgery, aortic atherosclerosis, vasculitis, or spinal trauma.

**Reference:**
Question(s) 231: Clinical Adult

Discussion:
Laceration of the middle meningeal artery is the typical basis for epidural hematoma.

Reference:

Question(s) 232: Behavioral

Discussion:
The presentation of disinhibited, poorly monitored verbal output that is socially unacceptable has been termed verbal dysdecorum and is seen most frequently following damage to the right frontal convexity.

Reference:

Question(s) 233: Physiology

Discussion:
When a strong nociceptive stimulus is given to one leg, it will cause inhibition of extensors in the stimulated leg, inhibition of flexors in the contralateral leg, a crossed extension reflex, and a contralateral positive supporting reaction.

Reference:

Question(s) 234: Behavioral

Discussion:
Schizophrenia is associated with pathological cellular changes in the dorsomedial thalamus and the dorsolateral prefrontal cortex, thereby affecting the pathway between the two sites. The role of pedunculopontine, nigrostriatal, hippocampal-fornical-mamillary, and amygdala-orbitofrontal pathways is not clearly established in schizophrenia.

Reference:

Question(s) 235: Clinical Adult

Discussion:
Carotid artery dissection may be associated with an ipsilateral Horner's syndrome and face or head pain.

Reference:

Question(s) 236: Clinical Adult

Discussion:
Hypothyroid myopathy is commonly associated with painful cramps and impressive elevations in serum CK levels. Muscle percussion commonly results in a slow, prolonged, electrically silent local mounding called myoedema. Polymyositis is not typically accompanied by painful cramps.

Reference:

Question(s) 237: Physiology

Discussion:
With regard to nerve conduction studies, a steady depolarization of excitable membranes slows conduction velocity. Axonal loss produces reduced compound motor action potential amplitudes.

Reference:

Question(s) 238: Clinical Adult

Discussion:
In complete section of the corpus callosum, left limb apraxia occurs because pathways connecting language areas in the left hemisphere to the motor areas of the right hemisphere are interrupted. Motor, sensory and visual functions of the right hemisphere are otherwise unaffected. Alexia without agraphia occurs with lesions of the left occipital lobe that involve fibers crossing from the right hemisphere through the splenium of the corpus callosum.
Reference:  

Question(s) 239: Behavioral  
Discussion:  
Disturbance with the recognition of color in one visual field -- hemiachromatopsia -- occurs only with inferior, posterior occipital lesions.

Reference:  

Question(s) 240: Behavioral  
Discussion:  
Retrograde amnesia is usually associated with an ongoing learning defect (anterograde amnesia) although there have been a few case reports of pure retrograde amnesia. Retrograde amnesia can shrink to short periods (measured in seconds) with return of learning ability.

Reference:  

Question(s) 241: Behavioral  
Discussion:  
Damage to the non-dominant parietal lobe results in prominent neglect or denial of the contralateral half of the body and space and dressing apraxia. Right/left confusion occurs with dominant parietal lobe lesions.

Reference:  

Question(s) 242: Clinical Pediatrics  
Discussion:  
Pompe's disease is a lysosomal glycogen storage disease that affects practically all tissues and results from a defect of 1,4-glucosidase (acid maltase). Hypotonia, failure to thrive, and decreased reflexes develop during the first few months of life. Cardiomegaly is prominent in infantile forms, which more commonly present with pulmonary insufficiency. Unlike other glycogeneses, the liver is normal in size or only slightly enlarged and there are no abnormalities of glucose homeostasis. PAS-positive glycogen is seen in membrane-bound vacuoles in muscle, hepatocytes, and Schwann cells, but no abnormalities are seen in myelin sheaths.

Reference:  

Question(s) 243: Clinical Adult  
Discussion:  
When the examiner is testing the biceps strength, the subject's forearm should be held in supination to eliminate elbow flexion force produced by the brachioradialis muscle.

Reference:  

Question(s) 244: Pathology  
Discussion:  
Vasogenic edema is characterized by blood-brain barrier breakdown and accumulation of plasma filtrate, including plasma proteins, in enlarged extravascular spaces. It is maximal in white matter. The other major type of cerebral edema, in contrast, is cytotoxic edema, which has its underlying pathogenesis in impairment of the cellular sodium-potassium membrane pump and causes intracellular swelling of neurons or glia, mostly in gray matter. Cerebral edema results in a reduction, not an increase, in ventricular size.

Reference:  
Question(s) 245: Behavioral  
Discussion:  
Akinetic mutism, with somnolence, indicates a disorder affecting the reticular projections from the mesencephalon to the thalamus.

Reference:  

Question(s) 246: Behavioral  
Discussion:  
Quetiapine has a very low anticholinergic side effect profile. Mesoridazine, thioridazine and clozapine have a very high anticholinergic side effect profile, and chlorpromazine a moderate profile.

Reference:  

Question(s) 247: Physiology  
Discussion:  
Electrodiagnostic characteristics of neuropathy include decreased motor unit action potential (MUAP) recruitment, increased MUAP amplitude, and fibrillation potentials with a distal to proximal gradient.

Reference:  

Question(s) 248: Pharmacology/Chemistry  
Discussion:  
Point mutations of the peripheral myelin protein-22 gene in chromosome 17p are responsible for Charcot-Marie-Tooth disease type 1A. Charcot-Marie-Tooth disease type 1B, linked to chromosome 1, is associated with point mutations of the Po gene that encodes for myelin protein zero.

Reference:  

Question(s) 249: Behavioral  
Discussion:  
Alzheimer's disease spares the primary sensory and motor areas of the brain until very late in the disease and usually does not cause focal weakness. Myoclonus is a well established concomitant feature in some cases.

Reference:  

Question(s) 250: Physiology  
Discussion:  
The presence of pain in an evolving brachial plexopathy is characteristic of carcinomatous invasion of the brachial plexus.

Reference:  

Question(s) 251: Physiology  
Discussion:  
The Martin-Gruber anastomosis produces a characteristic nerve conduction pattern of a larger median motor amplitude with proximal stimulation (compared with distal stimulation) and a significantly smaller ulnar motor amplitude with proximal stimulation (when compared with distal stimulation).

Reference:  
**Question(s) 252:** Anatomy  
**Discussion:**  
The infundibulum carries compounds from the hypothalamus to the neurohypophysis and adenohypophysis modulating the activities of these structures. The hypothalamic derived compound that tonically inhibits pituitary gland synthesis and release of prolactin is dopamine (also known as prolactin inhibitory factor). Regulation of other pituitary gland hormones is under positive control (i.e., hormone releasing factors) that are proteins. Interruption of the infundibulum leads to loss of tonic inhibition of prolactin secretion by dopamine and elevation of prolactin. Similarly, dopamine receptor antagonists will elevate prolactin and dopamine receptor agonists can suppress prolactin secretion and have been used in the medical management of prolactin secretion pituitary adenomas.

**Reference:**  

**Question(s) 253:** Clinical Adult  
**Discussion:**  
Hereditary paroxysmal cerebellar ataxia is an autosomal dominant disorder characterized by recurrent attacks of unsteady gait, limb ataxia, and dysarthria, lasting minutes to hours. Between attacks, patients may have nystagmus and mild gait ataxia, but the neurologic examination is otherwise normal. Oral acetazolamide usually prevents or reduces the frequency of the attacks.

**Reference:**  

**Question(s) 254:** Pathology  
**Discussion:**  
There are decreased amounts of choline acetyltransferase in brains of patients with Alzheimer's disease (AD). While several neurotransmitters are decreased in AD, cholinergic deficiency is the dominant neurochemical change.

**Reference:**  

**Question(s) 255:** Physiology  
**Discussion:**  
CSF is secreted in humans at about 0.3 ml per minute.

**Reference:**  

**Question(s) 256:** Pharmacology/Chemistry  
**Discussion:**  
Trazodone inhibits serotonin reuptake and blocks postsynaptic alpha-1 receptors. It is relatively safe but can produce priapism and orthostatic hypotension.

**Reference:**  
Tucker, GJ. Psychiatry for the neurologist part A. 1997 Nov;3(6).

**Question(s) 257:** Clinical Adult  
**Discussion:**  
The pronator quadratus is supplied by the anterior interosseous nerve, a branch of the median nerve. To test this muscle while minimizing the action of the pronator teres, pronation is tested with the elbow flexed.

**Reference:**  
Question(s) 258: Behavioral

Discussion:
This patient had nutmeg toxicity. Although a common ingredient in some pies, an excessive amount may be toxic. Nutmeg contains myristicin, which is a hallucinogen. It may produce hallucinations, severe headache and agitation, flushing, palpatations and extremity numbness. The symptoms usually resolve within several hours; however, the patient may require supportive therapy for the headache and autonomic features. A benzodiazepine may also help the agitation.

Reference:

Question(s) 259: Clinical Pediatrics

Discussion:
Rasmussen's syndrome usually presents in childhood with uncontrollable focal seizures that almost always do not respond to anticonvulsants or other medical treatments. The disease is progressive and usually is unilateral, involving only one hemisphere. Histologically, perivascular lymphocytic infiltrates with vascular injury, astrogliosis, neuronal loss and cortical atrophy are seen. Hemispherectomy is the best known treatment but there is increased interest in plasmapheresis because anti-glutamate receptor antibodies are seen in some cases.

Reference:

Question(s) 260: Anatomy

Discussion:
The tibial division of the sciatic nerve supplies the tibialis posterior muscle. The tibialis anterior, peroneus longus, peroneus brevis and extensor digitorum brevis are supplied by the common peroneal nerve.

Reference:

Question(s) 261: Pathology

Discussion:
In hydranencephaly, portions of the frontal, temporal and parietal lobes are replaced by a thin-walled cyst in the distribution of the middle and anterior cerebral arteries (internal carotid arteries).

Reference:

Question(s) 262: Anatomy

Discussion:
The ventral posteromedial thalamic nucleus receives afferent input from the contralateral spinal trigeminal nucleus, which is responsible for pain and temperature modalities of the face.

Reference:

Question(s) 263: Pathology

Discussion:
Most (72%) acute subdural hematomas occur as the result of falls or assaults; only 24% occur in relation to a motor vehicle accident. Extradural (epidural) hemorrhages are associated with skull fractures and bleeding results from tears in the meningeal vessels fed off the external carotid arteries. Although occasional acute subdural hematomas may be due to bleeding from cortical arteries, most appear to be secondary to rupture of the bridging veins.

Reference:

Question(s) 264: Anatomy

Discussion:
Visual attention is thought to be mediated by structures, including the pulvinar, claustrum, and superior colliculus.

Reference:
Question(s) 265: Physiology
Discussion:
Vertex or V-waves are high voltage sharp contoured waveforms that can occur with phase reversals on a bipolar montage over the central areas.

Reference:

Question(s) 266: Pharmacology/Chemistry
Discussion:
Gamma aminobutyric acid is found in high concentrations in inhibitory interneurons throughout the brain, and is the neurotransmitter of the output neurons of the striatum, globus pallidus, and cerebellar Purkinje cells.

Reference:

Question(s) 267: Physiology
Discussion:
Wicket waves, or wicket spikes, constitute a benign pattern of uncertain clinical significance, occurring predominantly in the EEG of older adults during light sleep.

Reference:

Question(s) 268: Anatomy
Discussion:
Derivatives of the alar plate include the cerebellum, inferior olivary complex, quadrigeminal plate and the red nucleus. Derivatives of the basal plate include motor nuclei of cranial nerves in the nucleus ambiguus that provides motor neurons to the striated muscles of the larynx and pharynx via the vagal, glossopharyngeal and accessory nerves.

Reference:

Question(s) 269: Physiology
Discussion:
Brainstem auditory evoked potentials would be normal in a patient with cortical deafness because the responses are recorded from the brainstem, not the cortex.

Reference:

Question(s) 270: Clinical Adult
Discussion:
Microaerophilic streptococcus species are the most common pathogens responsible for brain abscesses (60-70%) followed by Bacteroides (20-40%) and aerobic Gram negatives (25-35%). A mixture of two or more organisms is found in 30-60% of cases.

Reference:

Question(s) 271: Anatomy
Discussion:
The rubrospinal tract is concerned with control of tone in flexor muscles. This tract arises from the red nucleus, crosses in the ventral tegmental decussation, and lies anterior to the corticospinal tract in the lateral funiculus.

Reference:
**Question(s) 272:** Pharmacology/Chemistry

**Discussion:**
Streptococcus pneumoniae is a common cause of bacterial meningitis. As several causes of ceftriaxone resistant Streptococcus pneumonia have been reported, the initial treatment in suspect cases should include vancomycin, until the results of drug sensitivities are available.

**Reference:**

**Question(s) 273:** Clinical Adult

**Discussion:**
Anticholinergic (scopolamine) toxicity is associated with large, poorly reactive pupils.

**Reference:**

**Question(s) 274:** Anatomy

**Discussion:**
Microglia are the resident macrophages of the brain. They share antigens with circulating monocytes and both cell lines are probably derived largely from the same bone marrow precursors. The microglia are activated by injury to become large scavengers identical to macrophages. When activated they may undergo mitosis. These cells play no role in maintaining the blood-brain-barrier, but are conspicuous at the circumventricular organs where the blood-brain-barrier is absent.

**Reference:**

**Question(s) 275:** Physiology

**Discussion:**
Dermatomyositis causes weakness with an erythematous rash on the dorsum of the hands and on the elbows, knees, neck, and face. Needle EMG subacutely shows fibrillation potentials and small polyphasic motor unit action potentials.

**Reference:**

**Question(s) 276:** Physiology

**Discussion:**
The breach rhythm consists of EEG activity that is increased in amplitude and prominence recorded over the area of a skull defect. The breach rhythm is most prominent when seen over the central and temporal regions.

**Reference:**

**Question(s) 277:** Clinical Pediatrics

**Discussion:**
Friedreich ataxia is an autosomal recessive progressive spinocerebellar ataxia associated with expansion of both alleles of the frataxin gene in 98% of cases; rare cases with expansion of triplet repeats in one allele and point mutations in the other allele have been reported. Decrease in the density and number of myelinated fibers is present in sural nerve biopsies in most children, even early in the course of the illness.

**Reference:**
Reference:

Question(s) 280: Clinical Pediatrics
Discussion:
Prenatal cytomegalovirus infection can cause severe cerebral injury, such as microcephaly, microgyria, cerebral calcifications and chorioretinitis. Minimally affected children can have only hearing loss.

Reference:

Question(s) 281 - 284: Clinical Adult
Discussion:
Current therapy for Bassen-Kornzweig disease includes high dose vitamin E; for Hartnup disease, nicotinic acid; for Refsum's disease, dietary restriction of phytanic acid; and for multiple carboxylase deficiency, biotin.

Reference:

Question(s) 285 - 289: Anatomy
Discussion:
Posterior and lateral columns are selectively damaged in vitamin B12 deficiency and HIV vascular myelopathy. Tabes dorsalis results in posterior column dysfunction while anterior spinal artery occlusion spares this area. Spinal muscular atrophy affects anterior horn cells.

Reference:

Question(s) 290 - 294: Pharmacology/Chemistry
Discussion:
Advances in molecular genetics have provided insight into several genetic epileptic syndromes. Several channelopathies underlie genetic forms of epilepsy. Mutations in the alpha-4 subunit of the neuronal nicotinic acetylcholine receptor (chromosome 20q3.12) occur in autosomal dominant nocturnal frontal lobe epilepsy; mutations in the KCNQ2 (chromosome 20 q13.3)/KCNQ3 (chromosome 8q24) subtypes of voltage-gated potassium channels (which combine to conduct the M-current ion neurons) are associated with benign familial neonatal convulsions; and mutations in the beta subunit of the neuronal voltage-gated sodium channel (SCN1B, chromosome 19q13.1) with generalized epilepsy with febrile seizures. X-linked disorders of neuronal migration, including bilateral periventricular nodular heterotopia, associated with mutation in filamin-1 an actin-binding protein and lissencephaly with subcortical band heterotopia (linked to a mutation of the doublecortin gene) are also associated with seizures. Genetic causes of progressive myoclonic epilepsy include mutations of the cystatin B gene in the Unverricht-Lundborg type; and laforin (a intracellular protein tyrosine phosphatase) in progressive myoclonic epilepsy with Lafora bodies.

Reference:

Question(s) 295 - 299: Pharmacology/Chemistry
Discussion:
Levetiracetam does not have a clinically significant effect on liver enzymes and is inactivated to a small extent by the liver. It is mostly eliminated unchanged by the kidneys. Valproate inhibits the metabolism of other anticonvulsants, including lamotrigine and the epoxide metabolite of carbamazepine. Tiagabine has been reported to produce non-convulsive status epilepticus and absence seizures. Hyponatremia, ranging from asymptomatic to hyponatremic coma, may occur during oxcarbazepine use, particularly in the elderly. Urinary tract calculi may occur in patients while taking zonisamide (4%) or topiramate (1.5%). Weight loss with or without diminished appetite is more likely to occur with felbamate or topiramate.

Reference:
Question(s) 300 - 302: Pharmacology/Chemistry
Discussion:
Large fiber type peripheral neuropathy is seen in several inherited enzyme deficiencies, including Krabbe's disease, which results from galactosylceramidase deficiency. Hepatosplenomegaly also occurs in a number of storage diseases, including Niemann-Pick's disease, type A and B (acid sphingomyelinase deficiency), but not in Krabbe's disease or Fabry's disease. Cornea verticillata is unique to Fabry's disease, which also produces a small fiber peripheral neuropathy. Hexosaminidase A deficiency causes GM2 gangliosidosis and is not associated with corneal changes, hepatosplenomegaly or peripheral neuropathy.

Reference:

Question(s) 303 - 305: Clinical Pediatrics
Discussion:
The eye findings in pediatric neurologic disorders can be very helpful in the diagnosis. Kayser-Fleischer rings can be seen with a slit lamp well before neurologic findings appear in Wilson's disease. Brushfield spots are frequently seen in newborns with Down's syndrome, while Lisch nodules usually develop in neurofibromatosis during school age years or later.

Reference:

Question(s) 306 - 309: Pathology
Discussion:
Muscle biopsy findings reflect the underlying biochemical abnormalities. In carnitine deficiency, there is a defect of transport of fatty acids into the mitochondria, and lipid therefore accumulates in the muscle. Myophosphorylase deficiency impairs carbohydrate utilization leading to accumulation of glycogen. In mitochondrial myopathies, subsarcolemmal accumulations of numerous malformed mitochondria are seen as red accumulations on the modified Gomori trichrome stain ("ragged red fibers"). Inclusion body myositis is an indolent myositis of late adult life characterized by rimmed vacuoles in muscle fibers best seen on cryostat sections. Ultrastructurally, filamentous whorls are seen within the rimmed vacuoles.

Reference:

Question(s) 310 - 312: Pharmacology/Chemistry
Discussion:
Pyridoxine deficiency results from dietary deficiency of pyridoxine, and has occurred in infants given formulas in which the sterilization process has destroyed pyridoxine, or in milk from unusual sources that is intrinsically deficient in pyridoxine (such as goat's milk). It should be distinguished from pyridoxine dependency, an autosomal recessive disorder in which mutations producing conformational changes in glutamic acid decarboxylase cause early onset seizures that are controlled by pharmacologic doses of pyridoxine. Cobalamin (vitamin B12) deficiency may follow inadequate dietary intake (as in some vegetarian diets), or malabsorption with or without intrinsic factor deficiency. Patients typically have a slowly evolving syndrome of loss of posterior column function, with paresthesias, loss of vibration, proprioception and two point discrimination, and eventually spasticity and cognitive impairment. Thiamine deficiency most often occurs because of inadequate diet, as in alcoholics. Individuals with marginal diets may develop overt symptoms of thiamine deficiency if their remaining reserves are depleted by glucose loading. Acute thiamine deficiency causes Wernicke's encephalopathy, characterized by ataxia, confusion and abnormalities of eye movements.

Reference:
Question(s) 313 - 315: Pathology
Discussion:
Up to 50% of patients with dysplastic gangliocytoma of the cerebellum have the stigmata of Cowden syndrome (multiple hamartoma syndrome), which include oral mucosa fibromas, multiple trichilemmomas, hamartomatous colon polyps, thyroid neoplasms, and breast cancer. About 5% of patients with Gorlin syndrome (nevoid basal cell carcinoma syndrome) develop cerebellar medulloblastoma, particularly the desmoplastic subtype. Type 2 neurofibromatosis patients are prone to develop spinal cord ependymomas, whereas subependymal giant cell astrocytomas are characteristic of tuberous sclerosis.

Reference:

Question(s) 316 - 317: Clinical Adult
Discussion:
Hyperkalemic periodic paralysis and paramyotonia congenita are associated with mutations in the gene coding for the alpha subunit of the skeletal muscle sodium channel. About 70-80% of patients with the clinical diagnosis of Charcot-Marie-Tooth disease type 1A have a region of DNA duplication on chromosome 17; deletion of this same region is associated with hereditary neuropathy with liability to pressure palsies.

Reference:

Question(s) 318 - 321: Pathology
Discussion:
Intracytoplasmic inclusions in oligodendroglia, termed glial cytoplasmic inclusions (GCIs), are found in multiple system atrophy (MSA). GCIs are immunopositive for alpha-synuclein and MSA is considered by some to be a “synucleinopathy.” Neuronal intranuclear inclusions are found in Huntington’s disease and other CAG-polyglutamine expansion diseases. Intranuclear “ground glass” inclusions in oligodendrocytes are characteristic of progressive multifocal leukoencephalopathy (JC virus infection). The characteristic inclusion body seen in rabies encephalitis is the Negri body, which is found in the cytoplasm of large neurons.

Reference:

Question(s) 322 - 324: Physiology
Discussion:
The vestibulo-ocular reflex is mediated by the semicircular canals. Optokinetic nystagmus requires foveal fixation and smooth pursuit. Both have saccadic eye movements as a component.

Reference:

Question(s) 325 - 328: Anatomy
Discussion:
The superior colliculus projects to the pulvinar, forming the extrageniculate visual pathway. The inferior colliculus projects to the medial geniculate nucleus to relay auditory information. The basolateral nuclear complex of the amygdala projects to the dorsomedial thalamic nucleus that relays limbic inputs to the prefrontal cortex. The dentate nucleus projects to the ventral lateral nucleus, which relays cerebellar information to the motor cortex.

Reference:

Question(s) 329 - 330: Physiology
Discussion:
Sleep paralysis is seen in patients with narcolepsy. Morning headaches are seen in patients with obstructive sleep apnea.

Reference:
**Question(s) 331 - 333: Pharmacology/Chemistry**
**Discussion:**
Sumatriptan is an agonist of 5-HT1D (and perhaps 1F) receptors, and olanzepine and other atypical antipsychotic agents are antagonists of 5-HT2A and 5HT2C receptors. Lorazepam is an agonist of gamma aminobutyric acid receptors. Ondansetron is the best known therapeutic ligand of 5-HT3 receptors.

**Reference:**

**Question(s) 334 - 337: Pathology**
**Discussion:**
Takayasu's arteritis affects predominantly the aortic arch in young patients (age 15-45 years) of Asian heritage. Giant cell arteritis, in contrast, affects those over age 55 years, and causes a very high erythrocyte sedimentation rate. The vessel wall elastic lamina appears to be the main target of the inflammatory response. Antiphospholipid antibodies are particularly found with the CNS vasculitis associated with systemic lupus erythematous, with a prevalence of 7-58%, although they also occur with other diseases and may be present as an independent syndrome. The diagnosis of Wegener's granulomatosis is based on the presence of anti-neutrophil cytoplasmic autoantibodies. Polyarteritis nodosa is associated with seropositivity for hepatitis B in 10-50% of cases.

**Reference:**

**Question(s) 338 - 342: Clinical Adult**
**Discussion:**
The most common neurologic symptom of significant hyponatremia is obtundation. The rapid correction of hyponatremia is associated with central pontine myelinolysis. Severe hypokalemia is associated with muscle weakness. Hyperkalemia is neurologically asymptomatic although life-threatening cardiac arrhythmias may occur. Hypocalcemia is associated with tetany. Hypercalcemia commonly produces obtundation.

**Reference:**

**Question(s) 343 - 344: Physiology**
**Discussion:**
The serotonergic neurons of the raphe nuclei are thought to mediate NREM sleep. Both NREM and REM sleep states are retained with a transection at the cervico-medullary junction, as the structures mediating sleep states are located above the lower medullary level of the brainstem.

**Reference:**

**Question(s) 345 - 350: Clinical Pediatrics**
**Discussion:**
Tuberous sclerosis and myotonic dystrophy are autosomal dominant disorders, ataxia telangiectasia is an autosomal recessive disorder, Duchenne's muscular dystrophy is an X-linked recessive disorder, myoclonus epilepsy with ragged red fibers is a mitochondrial disorder, while Sturge-Weber syndrome is usually not an inherited disorder.

**Reference:**

**Question(s) 351 - 355: Pharmacology/Chemistry**
**Discussion:**
Familial amyloid neuropathy is associated with mutations in the transthyretin gene. Two clinical phenotypes have been described. Type I, the more common, typically presents between the ages of 25-35 with sensory impairment in the legs (often accompanied by lancinating pain and paresthesias) and autonomic dysfunction. Affected individuals often succumb to renal or cardiac failure within 10-15 years. Type II presents somewhat later and is slowly progressive. Carpal tunnel syndrome is often the presenting feature; autonomic dysfunction is not present. Tangier disease is a rare autosomal recessive condition that appears during childhood and is characterized by the presence of orange tonsils and a sensorimotor neuropathy that can predominantly affect the distal upper extremities, producing intrinsic hand muscle atrophy. There may also be relative preservation of position and
vibration perception. Ptosis, ophthalmoplegia and hepatosplenomegaly are less common. Tangier disease is associated with apolipoprotein A deficiency and very low HDL levels. Fabry’s disease, due to a defect in alpha galactosidase A (ceramide trihexosidase ), is an X-linked recessive disorder with onset during childhood or adolescent years. Intermittent lancinating pains and dysesthesias, affecting the distal limbs, may be triggered by fever, hot weather, and vigorous exercise. Diffuse vascular involvement produces hypertension, cardiomegaly, cardiac ischemia, renal damage, and stroke. A dark, red, macular and papular rash, often clustered over thighs and lower trunk, is a characteristic feature. Refsum’s disease is an autosomal recessive disorder characterized by retinitis pigmentosa, cerebellar ataxia, and chronic polyneuropathy. Cardiomyopathy and neurogenic deafness develop in most; ichthyotic skin changes, pupillary abnormalities, and cataracts in some. Anosmia and night blindness may precede the neuropathy. Refsum's disease is due to a defect in phytanic acid oxidase. Hereditary coproporphyria is due to a defect in coproporphyrinogen oxidase. It is inherited in an autosomal dominant fashion. Skin photosensitivity is present and blistering occurs with excessive sun exposure or mild trauma, leading to scarring. Neurologic features are similar to those seen in acute intermittent porphyria.

Reference:

Question(s) 361 - 363: Clinical Adult Discussion:
Chorea is one of the major features in the Jones diagnostic criteria for rheumatic fever. It typically arises several months after the acute group A streptococcal infection. The median interval between acute measles infection and onset of subacute sclerosing panencephalitis is 8 years, with a range from 2 to 12 years. Acute motor axonal neuropathy occurs in epidemics in northern China during summer months, and it has been associated with antecedent Campylobacter jejuni infection.

Reference:

Question(s) 364 - 366: Clinical Adult Discussion:
Villaret’s syndrome (involvement of cranial nerves IX, X, XI, XII, and the cervical sympathetic chain) is due to a lesion in the retroparotid or retropharyngeal space. Glossopharyngeal neuralgia may result. Glossopharyngeal neuralgia is sometimes associated with syncope; the pain often responds to carbamazepine whether or not there is associated syncope.

Reference:

Question(s) 367 - 368: Clinical Adult Discussion:
Lateral medullary infarctions are most often associated with vertebral artery occlusive disease, either atherosclerotic or cardioembolic. Typical features are vertigo, facial pain, headache, nausea, vomiting, and ataxia; examination findings include ipsilateral Horner’s syndrome, reduced pain and temperature sensation in the ipsilateral face and contralateral body.

Reference:
Question(s) 369 - 372: Pharmacology/Chemistry Discussion:
The patient is suffering from cervical dystonia (spasmodic torticollis) for which botulinum toxin injection is the treatment of choice. Botulinum toxin acts by preventing acetylcholine release. It typically takes one week for its clinical effect to appear; the beneficial effect usually lasts for several months. It is generally well tolerated, but dysphagia is the most frequent complication following sternocleidomastoid or other cervical muscle injections, occurring in 15-25% of patients. Prolonged treatment can result in antibody formation with declining efficacy, especially if frequent doses and "booster" injections are employed.

Reference:

Question(s) 373 - 375: Pharmacology/Chemistry Discussion:
The discovery of elevated CSF glutamate levels in patients with ALS led to the study of glutamate release inhibitors, such as riluzole, in the treatment of ALS. Studies have shown that riluzole has an 8% benefit on survival at 12 months and a 9% benefit on survival at 18 months. Both baclofen and dantrolene are effective in the symptomatic treatment of spasticity. However, baclofen has fewer adverse effects and is, therefore, a more appropriate initial therapeutic choice. Diazepam or tizanidine could also be used.

Reference:

Question(s) 376 - 378: Clinical Pediatrics Discussion:
In globoid cell leukodystrophy (Krabbe's disease), there is demyelination of the brain, spinal cord and peripheral nerves due to a deficiency of galactosylceramidase. Onset of the classic infantile form is four to six months of age with dementia, increased muscle tone and loss of deep tendon reflexes due to peripheral neuropathy. MRI of the brain reveals demyelination; CSF protein is elevated; nerve conduction velocities are slowed; optic nerves are pale. The onset of classic metachromatic leukodystrophy due to a deficiency of arylsulfatase is later in the second year of life. Cherry red macular is seen in several of the sphingolipidoses but not the leukodystrophies.

Reference:

Question(s) 379 - 381: Clinical Pediatrics Discussion:
The gene for Duchenne's and Becker's muscular dystrophy resides in the X chromosome, but approximately one-third of single cases in a family result from new mutations. The clinical onset of Duchenne's dystrophy is around age four years and Becker's dystrophy presents later in the first decade, but there is clinical overlap. Serum CK (creatine kinase) and muscle histology does not differentiate the two disorders. The muscle protein, dystrophin, is absent in Duchenne's and of abnormal size in Becker's providing a tool for differential diagnosis. About 70% of cases with Duchenne's and Becker's dystrophy have a deletion on the X chromosome and the remainder of cases have a point mutation. If a deletion is present in the index case, the carrier status of the mother and other female relatives can be ascertained. If a male with mild Becker's dystrophy has children, all of his daughters will be carriers and his sons will not be affected.

Reference:

Question(s) 382 - 383: Pharmacology/Chemistry Discussion:
Ataxia-telangiectasia (A-T) is an autosomal recessive disorder that characterized by progressive childhood onset ataxia, with subsequent development of chorea, myoclonus, anterior horn cell disease, and both cellular and humoral immunodeficiency. Patients typically develop bulbar telangiectasias within the first decade, as well as premature graying, loss of subcutaneous fat, vitiligo and café-au-lait patches. Heterozygotes have an increased incidence of
cancer, particularly lymphomas. Patients with advanced disease have cerebellar atrophy on MRI and CT, and blood tests typically show elevated alpha-fetoprotein and carcinoembryonic antigen, and variable deficiencies of IgA, IgE and IgG subclasses. Expanded GAA repeats in both alleles of the frataxin gene are seen in Friedreich's ataxia, and result in abnormal mitochondrial iron transport. Low copper and ceruloplasmin are seen in Wilson's disease, that has recently been found to have abnormalities of mitochondrial copper trafficking. SURF-1 is a nuclear gene that is required for normal assembly of cytochrome C oxidase (COX). Tocopherol transport protein transports alpha-tocopherol (Vitamin E) in the plasma; its deficiency produces a Friedreich ataxia-like phenotype. The gene product in A-T, ATM is a kinase with carboxy-terminal phosphatidylinostitol 3-kinase-like domain, that has recently been found to Phosphorylate the Nijmegen breakage syndrome protein (NBS1). This action appears to be critical for DNA repair, and likely explains the protein manifestations of A-T.

Reference:

Question(s) 384 - 386: Pharmacology/Chemistry Discussion:
Dopa-responsive dystonia may result from a number of inherited enzyme deficiencies. GTP cyclohydrolase deficiency is transmitted as an autosomal dominant trait, with variable expressivity. Tyrosine hydroxylase and aromatic-L-amino acid decarboxylase deficiency are autosomal recessive disorders. Monoamine oxidase deficiency is an X-linked disorder that presents with aggressive behavior in hemizygotes; dystonia is not a part of the phenotype. Patients with GTP cyclohydrolase deficiency typically remain responsive to levodopa, and do not develop cognitive impairment or stroke-like episodes. Children typically present with asymmetric gait dystonia with diurnal fluctuation, but adults may present with focal upper extremity signs or mild parkinsonism. CSF analysis in GTP cyclohydrolase deficiency reflects the impaired production of the protein co-factors required for the activity of the hydroxylases that catalyze the production of biogenic amines. Thus, levels of proteins, HVA and 5-HIAA will be low prior to treatment.

Reference:

Question(s) 387: Radiology Discussion:
In the coronal image, bilateral electrodes can be seen creating an artifact at the vertex that extends down to the medial pallidal region. The cross sections of the tracts of the electrodes project symmetrical dark dots in the T2 axial image. The dots are located in the globus pallidus. Bilateral or unilateral pallidal stimulation is used for the treatment of Parkinson's disease. Thalamic stimulators can be used for essential tremor; however, in this case, the stimulators are not in the thalamus.

Reference:

Question(s) 388: Radiology Discussion:
Proton density and T2-weighted images in the coronal plane show a bithalamic mass lesion, larger on the left than on the right. The lesion elevates the floors of the bodies of the lateral ventricles, more so on the left than on the right, and displaces the internal cerebral vein from left to right. The temporal horns of the lateral ventricles are slightly dilated, but the surrounding tissue appears normal. The corpus callosum appears intact. Given the age of the patient (17 years) and the finding of a bithalamic mass lesion, a tumor is the most likely diagnosis. Infarction in the distribution of both thalami would be extremely unusual, especially at this young age.
Reference:

Question(s) 389: Radiology
Discussion:
Although the history suggests a metastasis, inspection of the lesion reveals a mass in the sella turcica that is expanding in all directions, particularly towards the left with compression of the trigeminal and oculomotor nerves in the cavernous sinus. Dural enhancement is present. The mass itself enhances relatively little, which would be very unusual for a metastasis but much more common for a pituitary adenoma. On the left side of the adenoma, there is a low-intensity area that may correspond to necrotic tissue. These findings are characteristic of pituitary apoplexy, which is infarction of a pituitary adenoma with resultant rapid swelling and pressure exertion on neighboring structures. Pituitary apoplexy may also occur secondary to hemorrhage into an adenoma. Sarcoidosis tends to enhance uniformly.

Reference:

Question(s) 390: Radiology
Discussion:
The fourth ventricle is of normal size without cysts and the corpus callosum is definitely present. The tectal area is normal. The cerebellar tonsils descend below the foramen magnum. The patient has Chiari type I malformation.

Reference:

Question(s) 391: Radiology
Discussion:
The MRI flair image shows a recurrent artery of Heubner infarct. This artery supplies the anteroinferior portion of the caudate nucleus, the putamen, and the anterior limb of the internal capsule.

Reference:

Question(s) 392: Pathology
Discussion:
The image shows a colloid cyst of the third ventricle.

Reference:

Question(s) 393: Pathology
Discussion:
This muscle biopsy demonstrates degenerating fibers undergoing phagocytosis, hypercontracted fibers, excessive fibrosis and variation in fiber size. The proliferation of connective tissue, especially, is most consistent with a dystrophic process, in this case Duchenne’s muscular dystrophy. Kugelberg-Welander syndrome is a spinal motor neuron atrophy which gives a neurogenic appearance in muscle biopsies. Nemaline rod myopathy contains classic eosinophilic inclusions in the muscle fibers. Inclusion body myositis, very uncommon in children, demonstrates rimmed vacuoles and an inflammatory picture, while Pompe’s disease is manifested by glycogen storage in the myofibers.
Reference:

Question(s) 394: Radiology
Discussion:
The most likely possibility is an aneurysm because of the lesion's negligible signal intensity due to fast flow.

Reference:

Question(s) 395: Radiology
Discussion:
The post-injection T1-weighted MR image shows enhancement of an extra-axial small mass that displaces the brain parenchyma away from the inner table of the skull. The inner table of the skull is focally expanded at the center of the mass. The findings are characteristic of a meningioma. A "dural tail sign" is present on the post-contrast image. An epidural hematoma would have been of high signal intensity on the pre-injection study. Without expansion of the bone, a depressed fracture would have a plane of cleavage within the calvarium and may also have had an underlying cortical contusion.

Reference:

Question(s) 396: Radiology
Discussion:
The MRI images are consistent with an acute hemorrhage. The bright ring on T1, which does not enhance with gadolinium, corresponds to intracellular methemoglobin, which appears dark in the T2-weighted image on the far left. In this image, edema can be seen as a bright signal area surrounding the acute hemorrhage. The core of the hemorrhage, containing intracellular deoxyhemoglobin, appears gray on both T1 and T2-weighted images. As the hemorrhage becomes organized, the ring will become bright on both pulse sequences as methemoglobin becomes extracellular.

Reference:

Question(s) 397: Radiology
Discussion:
The sagittal MRI shows a round, sharply marginated, nonenhancing mass in the hypothalamus superior to the infundibulum, consistent with a hypothalamic hamartoma. Hamartomas are usually isointense to gray matter on all sequences, do not enhance, and can be sessile or pedunculated. An aneurysm would demonstrate either signal void due to flowing blood, or a complex arrangement of high and low signal layers due to clot within the aneurysm. Sarcoidosis, craniopharyngioma, and meningioma would enhance with contrast.

Reference:

Question(s) 398: Radiology
Discussion:
A straightforward case of an arteriovenous malformation is seen in the MRI scans. The low signal region is characteristic of the flow void typical of arteriovenous malformation. Mass effect is absent.

Reference:

Question(s) 399: Radiology
Discussion:
The MR findings are those of a herniated L5-S1 disc. The disc extends beyond the endplate of the adjacent vertebral bodies into the epidural fat and in the axial projection is shown to compress and displace the left nerve root. Meningiomas are rare at the lumbar canal and arise as a broad-based lesion from the dura. Metastases do not involve the disc space directly, but rather arise from the dense
bone of the pedicle and vertebra, and often dissect into the epidural fat. Neurofibromas may have a variety of appearances depending upon whether they are solitary or plexiform, but are intradural extramedullary lesions and can have an extradural component as well. They tend to pass through the neural foramina. Lymphoma originates either within the vertebral body with extension posteriorly into the epidural region or within the paravertebral lymph nodes with extension through the neural foramina into the epidural space leading to compression of the thecal sac. Of all these lesions, only the herniated disc arises within the disc space and extends posteriorly in the fashion seen in the scans.

Reference:

Question(s) 400: Radiology
Discussion:
The correct answer is HIV encephalitis. A progressive subacute encephalitis due to a direct cerebral HIV infection (previously known as AIDS dementia complex) may be visualized by MRI. Cortical atrophy, seen as enlargement of the supratentorial subarachnoid and ventricular spaces, is the most common MRI finding, and is present in virtually all patients with clinically significant HIV encephalitis. Associated demyelinated and vacuolated lesions are isointense to mildly hypointense on T1-weighted images and markedly hyperintense on T2-weighted images. These typically begin in the paraventricular white matter and centrum semiovale and spare the subcortical U-fibers. The lesions may be symmetric or asymmetric and appear "fluffy" or "cotton-like." They are poorly circumscribed and often become confluent and diffuse. No mass effect or parenchymal enhancement is noted. Lymphoma lesions are usually contrast-enhancing. The lesions of progressive multifocal leukoencephalopathy most commonly extend to the subcortical U-fibers and appear hypointense on T1-weighted images. Creutzfeldt-Jakob disease has a variable appearance on MRI that often includes prominent basal ganglia involvement.

Reference:

Question(s) 401: Physiology
Discussion:
An EEG showing triphasic waves is consistent with hepatic encephalopathy.

Reference:

Question(s) 402: Radiology
Discussion:
The correct answer is mesial temporal sclerosis (MTS), also known as hippocampal sclerosis. MRI may show three major findings in MTS: (1) volume loss of mesial temporal lobe parenchyma; (2) signal alterations of the existing mesial temporal lobe gray matter (hypointensities on T1-weighted images and hyperintensities on T2-weighted images); and (3) disruption of the normal hippocampal architecture. The evaluation of volume loss (atrophy) of the hippocampus relies on a comparison of the parenchyma and CSF spaces of the affected side to the contralateral side. The anterior hippocampus is preferentially affected. Hypointensities on T1-weighted images and hyperintensities on proton density- and T2-weighted images usually involve the gray matter more prominently than the white matter. Careful examination of the proton density images is especially helpful to differentiate abnormal hippocampal signal from normal CSF in the temporal horn or choroidal fissure. When signal alterations are seen in combination with hippocampal architectural disruption or atrophy, the specificity of diagnosing MTS becomes more favorable.

Reference:
Question(s) 403: Pathology

Discussion:
The cell origin of the hemangioblastoma is still unknown, despite many investigative efforts. The tumor arises independently or in conjunction with von Hippel-Lindau disease. The most common location is in the cerebellum, but the spinal cord can also be a site of origin. Microscopically, the tumor is a capillary-rich neoplasm with abundant foamy cells containing lipid. Pilocytic astrocytomas, also commonly found in the cerebellum, are composed of astrocytes with long, fibrillar processes. Medulloblastomas are malignant small blue cell tumors of neuronal origin. Ependymomas are glial tumors composed of ependymal cells, which contain fairly uniform nuclei and characteristic perivascular pseudorosettes.

Reference:

Question(s) 404: Radiology

Discussion:
The image shows a subacute subdural hematoma (SDH). SDHs conform to the shape of convexity and tend to have a lens or "banana" shape. They freely cross cranial sutures but do not cross the midline insertion of the falx or tentorium. SDHs evolve on MRI at a slower rate compared to intraparenchymal hematomas (IPHs). This occurs because the subdural space lacks a blood-brain-barrier and the adjacent dura matter remains well vascularized; thus, the oxygen concentration within an SDH remains higher than that expected for an IPH of the same age. This prolongs the evolution of the hemoglobin degradation process by days to weeks at each stage.

Reference:

Question(s) 405: Radiology

Discussion:
The lesion affects both white matter and overlying cerebral cortex, consistent with operculofrontal middle cerebral artery infarction. Intermixed hyperdensities are consistent with hemorrhagic conversion. There is no diffuse subarachnoid hyperdensity to suggest subarachnoid bleeding. There is no vasogenic edema pattern to suggest high-grade primary or metastatic neoplasm. There is no hyperdensity in the superior sagittal sinus to suggest venous thrombosis. Amyloid angiopathy usually causes large lobar hemorrhages.

Reference:

Question(s) 406: Radiology

Discussion:
The scans show fusiform cord enlargement, best appreciated on T2-weighted images, extending over several spinal levels. Increased intramedullary T2-signal is noted in areas corresponding to cord enlargement. The findings are not entirely specific, but are most consistent with acute myelitis as compared to the other choices. The MRI findings in various forms of myelitis are non-specific, including those of idiopathic myelitis and those of myelitis associated with identifiable causes such as infections, post vaccination, post-infectious, and collagen vascular diseases. Such findings typically include spinal cord enlargement, intramedullary increased T2-signal lesions, and variable enhancement involving several spinal levels. The lack of a hemorrhagic component weighs against a spinal cord contusion. The lack of enhancement could be seen with myelitis, but argues strongly against ependymoma and would be unusual for astrocytoma.

Reference:
Question(s) 407: Radiology
Discussion:
The findings seen are consistent with an acute corona radiata stroke showing high signal on flair and diffusion images and decreased signal on apparent diffusion coefficient mapping.

Reference:

Question(s) 408: Radiology
Discussion:
The rounded lesion that enhances with gadolinium is located in the center of the conus medullaris, which is a location preferred by ependymomas, particularly the myxopapillary variant. Metastasis here is less frequent, as is the inflammatory lesion of schistosomiasis. Herniated discs and neurofibromata are extramedullary lesions.

Reference:

Question(s) 409: Radiology
Discussion:
The contrast-enhanced sagittal midline MRI shows multiple nodular lesions and drop metastases that are most often seen with medulloblastoma. Ependymomas, anaplastic gliomas, germinomas, and choroid plexus tumors can also seed the neuraxis through the CSF.

Reference:

Question(s) 410: Radiology
Discussion:
The magnetic resonance venograms clearly show the transverse sinus (A), superior sagittal sinus (C), and the jugular vein (F).

Reference:
Nemer FH. Nervous system anatomy and physiology. New Jersey: CIBA Medical Education Division, 1986.

Question(s) 411: Pathology
Discussion:
The image shows numerous small gyri over the surface of the brain. The correct diagnostic term is polygyria.

Reference:

Question(s) 412: Radiology
Discussion:
The rounded lesion in the right cerebellar hemisphere demonstrates hypointensity on T1-weighted and T2-weighted images, and a peripheral ring of hyperintensity on T1-weighted images. These findings are characteristic of early subacute blood (deoxyhemoglobin centrally and intracellular methemoglobin peripherally). On the T2-weighted image, thin linear structures demonstrating signal void (flow) are seen anterior to the hematoma, representing the arteriovenous malformation that ruptured.

Reference:

Question(s) 413: Radiology
Discussion:
The findings demonstrate right occipital encephalomalacia most consistent with chronic changes following a right posterior cerebral artery infarct.

Reference:
Question(s) 414: Pathology
Discussion:
An eosinophilic cytoplasmic inclusion in a neuron with melanin granules is a Lewy body. Negri bodies are intracytoplasmic inclusions seen in rabies but these are not common in pigmented neurons.

Reference:

Question(s) 415: Radiology
Discussion:
The lesion in the left hippocampus has the imaging characteristics of a cavernous angioma, with a rim of low-intensity hemosiderin surrounding a cluster of high-intensity, fluid-containing vessels. The absence of draining veins rules out arteriovenous malformation and venous angioma. Astrocytoma and oligodendroglioma do not have the imaging characteristics present in this MRI.

Reference:

Question(s) 416: Radiology
Discussion:
The pre- and post-contrast MR images show a mass at the neural foramen of the spine that enhances after gadolinium administration. Of the choices offered, the most likely is neurofibroma. Ependymoma is an intramedullary tumor while the illustrated lesion is not. Extruded discs are located within the spinal canal but do not extend outside of it. The same applies to meningiomas. A metastasis would most likely involve the vertebral body and would not be nearly as frequent as a neurofibroma in the location shown in this image.

Reference:

Question(s) 417: Radiology
Discussion:
The linear high density structure seen on the non-contrast head CT lies at the expected location of the proximal portion of the left middle cerebral artery. The artery should not be so dense without intravenous contrast present; therefore, intraluminal thrombosis should be presumed. The T2-weighted MR image in a slightly higher cut demonstrates hemorrhagic infarction in the perisylvian region and lentiform nucleus in the portions of the brain supplied by the lateral striate arteries, which arise from the M1 segment of the middle cerebral artery (MCA) and proximal cortical branches of the MCA.

Reference:

Question(s) 418: Radiology
Discussion:
The pituitary lesion shown on the left side of the gland demonstrates classic features of a pituitary microadenoma on the noncontrast MRI, with focal hypointensity less than one centimeter in size, displacement of the pituitary stalk, remodeling of the sellar floor, and upward convexity of the superior margin of the gland. On the image shown, there is lateral bowing of the medial wall of the cavernous sinus abutting the lesion; however, MR is not a sensitive method to determine invasion of the cavernous sinus, which can be diagnosed with certainty only when carotid arterial encasement is seen, which is not seen on this image. Pituitary apoplexy would show high signal (MET-HG) on the T1-weighted image. Epidermoid is a possibility but much less likely. The neurohypophysis is usually hyperintense in T1-weighted image.

Reference:

Question(s) 419: Radiology
Discussion:
A normal spectrum is shown. The tallest peak is N-Acetyl-Aspartate (NAA) and represents neuronal activity (normal). To the left of the NAA peak are the (normal) creatinine and (normal, smaller) choline peaks.

Reference:
Question(s) 420: Pathology
Discussion:
In patients with Pick's disease, the atrophy is most pronounced in the frontal and temporal lobes. Pick's disease is a member of a larger group of disorders classified as frontotemporal dementias.

Reference:

Question(s) 421: Radiology
Discussion:
The lesion predominantly affects white matter, consistent with a vasogenic edema pattern due to high-grade primary or metastatic neoplasm. There is no hyperdensity to suggest bleeding. The relative sparing of the overlying cerebral cortex would be unusual for an operculofrontal middle cerebral artery infarction. A contrast-enhanced MRI is indicated for further clarification.

Reference:

Question(s) 422: Radiology
Discussion:
Leptomeningeal enhancement, parenchymal enhancement, and, especially, pituitary stalk/hypothalamic involvement in a patient with cranial nerve symptoms is characteristic for neurosarcoidosis. About 5-10% of sarcoidosis patients develop CNS involvement. Lack of hemiatrophy and prominent (asymmetric) choroid plexus enhancement make the diagnosis of Sturge-Weber syndrome less likely. No gyral enhancement is present. Low CSF pressure would not be limited to one hemisphere and would not have mass-like and leptomeningeal enhancement. The enhancing lesion is intra-axial and therefore is not a meningioma.

Reference:

Question(s) 423: Radiology
Discussion:
The pre-and post-injection gadolinium enhanced MR studies show a round contrast-enhancing lesion in the medial left temporal lobe. There is also a fullness and slight mass effect within the uncus of the same side. Abscesses tend to be ring lesions and mesial temporal sclerosis does not enhance. The lesion is not cystic and is also in the wrong location for a colloid cyst. Thus, the most likely diagnosis is tumor.

Reference:

Question(s) 424: Pathology
Discussion:
There is complete degeneration of one medullary pyramid and atrophy of the ipsilateral cerebral peduncle indicating Wallerian degeneration secondary to destruction of the corticospinal tract above this level.

Reference:

Question(s) 425: Radiology
Discussion:
Pick's disease is one of the frontotemporal dementias characterized by atrophy in the frontal and temporal lobes. The atrophy is bilateral in 30% of cases, left-sided predominant in 50%, and right-sided predominant in 20%.

Reference:
Question(s) 426: Radiology
Discussion:
The differential diagnosis of ring-enhancing lesions includes metastatic disease, abscess, and subacute infarct or hematoma. In this case, the presence of multiple lesions is most suggestive of metastasis, which is the most common neoplasm of the posterior fossa in adults. An abscess is not excluded; however, abscess was not provided as a choice. A resolving hematoma would generally not have as much edema as seen in these scans.

Reference:

Question(s) 427: Radiology
Discussion:
The parasagittal, uniformly-enhancing, dural-based mass seen in the MR scans is most consistent with a meningioma. Regarding the other choices, a lipoma would be hyperintense on pre-contrast scans; the sagittal sinus is clearly not obstructed in the post-contrast coronal images; there is no caudate atrophy to suggest Huntington's disease; and toxoplasmosis usually manifests as multiple "ring-enhancing" parenchymal lesions.

Reference:

Question(s) 428: Radiology
Discussion:
Anoxia causes increased T2 signal in the globus pallidus, deep portions of the sulci, and superior cerebellum, which are all anatomical regions that are susceptible to hypoxia. The necrotic globus pallidus enhances after gadolinium administration.

Reference:

Question(s) 429: Pathology
Discussion:
Failure of cleavage of the prosencephalon results in holoprosencephaly and is associated with midline facial defects. The malformation may be due to chromosomal abnormalities in some cases.

Reference:

Question(s) 430: Pathology
Discussion:
The lesion shown in this image is a remote infarct. The preservation of the outer layer of the cortex would argue against a remote contusion. An old hematoma cavity would be smoother, and both an abscess and metastatic carcinoma would be accompanied by brain swelling rather than shrinkage (note the enlarged frontal horn).

Reference:

Question(s) 431: Radiology
Discussion:
The basilar artery is markedly dilated and can be seen in its typical location anterior to the midbrain. The left middle cerebral artery can be seen to supply large feeders to the arteriovenous malformation. The arteriovenous malformation has a typical appearance that is not seen in neoplasia. Soft neurological findings are common in this setting. The basilar artery can be seen clearly and there is no corresponding vein to be confused with this structure. Therefore the arteriovenous malformation likely involves all three major intracerebral vessels.

Reference:

Question(s) 432: Pathology
Discussion:
The image is a coronal section through the splenium of the corpus callosum. There are sharply demarcated plaques of demyelination outside the walls of both lateral ventricles.

Reference:

Question(s) 433: Pathology
Discussion:
The image shows atrophy of the caudate nuclei with compensatory ventricular enlargement characteristic of Huntington's disease. This autosomal dominant trinucleotide repeat amplification disorder is associated with all permutations of movement disorders including chorea, athetosis, dystonia and rigidity, coupled with dementia.

Reference:

Question(s) 434: Radiology
Discussion:
The scans show active spondylitis at L4-5 and remote spondylitis at L2-3. Spondylitis may include osteomyelitis, discitis and epidural abscess formation, each of which are seen in the current case at L4-5. The enhancement and T2 hyperintensity of the L4-5 disc is consistent with active discitis. In contrast, metastatic disease usually spares and does not cross the disc space. Remote spondylitis often results in the changes seen in the current case at L2-3; the disc space is markedly narrowed and the adjacent vertebral bodies show degenerative changes. Incidental note is made of scoliosis, which is maximal at L2.

Reference:
Question(s) 438: Pathology
Discussion:
Syringomyelia is a cystic lesion in the spinal cord that interrupts the crossing pain fibers and damages anterior horn cells. Muscle atrophy, not hypertrophy, often results. Vibratory, position, and touch senses are usually spared.

Reference:

Question(s) 439: Radiology
Discussion:
The correct answer is enlarged perivascular spaces (PVS; also known as Virchow-Robin spaces). PVSs surround and accompany cerebral vessels as they penetrate the brain from the subarachnoid space. They contain CSF and probably represent an extension of the subarachnoid space. With advancing age, PVSs are found with increasing frequency and enlarging size on MRI. PVSs are isointense with CSF on T1-weighted images, proton density-weighted images, and T2-weighted images. In the transverse plane, they are usually bilateral, symmetric, round, well-circumscribed, and less than 4 mm in diameter. In the longitudinal plane, they appear curvilinear or tubular. PVSs are most commonly noted adjacent to the anterior commissure at the level of the inferior one-third of the basal ganglia and extending through the high-convexity cortex, subcortical, and paraventricular white matter. Lesions in vasculitis, demyelinating disease, and Binswanger's disease are typically hyperintense on proton-density images.

Reference:

Question(s) 440: Pathology
Discussion:
Multiple, well-circumscribed pigmented lesions are most likely metastatic melanoma. Metastases tend to occur at the grey-white junction, but they may be seen anywhere in the neuraxis. Lung, breast, melanoma, kidney, and gastrointestinal tumors are the major sources of CNS metastases.

Reference:

Question(s) 441: Pathology
Discussion:
The intense astrocytic reaction with spongiform change in the cortex shown in the hematoxylin-and-eosin stained section is characteristic of Creutzfeldt-Jakob disease.

Reference:

Question(s) 442: Physiology
Discussion:
The EEG shows generalized theta activity, which could be seen as a normal drowsy pattern in a child.

Reference:

Question(s) 443: Pathology
Discussion:
The image shows a subdural hematoma.

Reference:
Question(s) 444: Radiology
Discussion:
The scan demonstrates findings highly suggestive of acute or subacute Herpes simplex virus (HSV) encephalitis involving the temporal lobe. The lesion crosses the vascular boundaries of the middle and posterior cerebral arteries, making an ischemic insult unlikely. The contrast-enhancing pattern would be unusual for glioblastoma or lymphoma. A contusion could occur in this location, but would contain evidence of hemorrhage and MR evidence of parenchymal blood degradation products, which is not seen in this case. HSV encephalitis primarily involves limbic structures. MR typically shows temporal and inferior frontal lobe swelling, with low signal on T1-weighted images and high signal on T2-weighted images. Post-contrast studies in the acute and subacute setting often show cortical and leptomeningeal enhancement. The lesion may not be hemorrhagic in the early stages.

Reference:

Question(s) 445: Radiology
Discussion:
The axial T1-weighted MRI demonstrates multiple low signal lesions in the right hemisphere, which is also smaller than the left hemisphere. The right trigone is larger than the left and the volume of the white matter on the right side is smaller. The lesions extend from the central gray nuclei all the way to the cortex and are most likely of an ischemic nature. The phase-contrast coronal arteriogram does not demonstrate any flow in the M1 segment of the right middle cerebral artery, confirming the presence of chronic infarction of the right cerebral hemisphere.

Reference:

Question(s) 446: Physiology
Discussion:
Subacute sclerosing panencephalitis is associated with periodic slow wave complexes in the EEG that recur every 4 to 15 seconds.

Reference:

Question(s) 447: Radiology
Discussion:
The pre-and post-injection gadolinium T1-weighted images show an intra- and extracanicular mass lesion that is extra-axial and compresses the pons. These features are consistent with an acoustic neuroma. The lesion is adequately seen on the pre-injection study and the post-injection study need not have been done. The differential diagnosis is limited to an acoustic schwannoma (also known as vestibular schwannoma, acoustic neuroma, or acoustic neuromina), with little likelihood that it could be a meningioma.

Reference:

Question(s) 448: Pathology
Discussion:
A combination of hydrocephalus and meningomyelocele is a common feature of an Arnold-Chiari malformation (Chiari type II malformation) of the cerebellum and medulla.

Reference:

Question(s) 449: Radiology
Discussion:
Intracranial lipomas most commonly occur near the midline, usually along the corpus callosum as in this case. The CT shows a midline lipoma in the interhemispheric space. There is calcification associated with the lipoma and hydrocephalus is present. Midline lipomas are thought to represent a form of dysraphism.
**Reference:**

**Question(s) 450:** Pathology

**Discussion:**
Toxoplasmosis is a parasitic infection seen most commonly in immunosuppressed patients. Both tachyzoites and cysts containing bradyzoites may be present.

**Reference:**

**Question(s) 451:** Radiology

**Discussion:**
The CT scan shows a collection of air in the right frontal lobe, with the bottom portion having an air fluid level. The finding is not consistent with an artifact nor with a vascular event. Infection could be present; however, the way in which the air would gain access to the intracranial compartment would have to involve a break in the skull, most often in the air containing sinuses of the cranial base. In this case, the air collection entered through a fracture of the frontal sinus. In order for the air to occupy as much space within the brain parenchyma as it does, there would have to be a loss of substance of the brain focally at the site. This is consistent with the patient having had a loss of brain substance (volume) due to a prior focal hemorrhagic contusion.

**Reference:**

**Question(s) 452:** Radiology

**Discussion:**
The CT and MRI scan findings are highly consistent with Sturge-Weber syndrome (SWS). Cerebral lesions in SWS are most frequently found in the unilateral parieto-occipital area. On CT scans, parenchymal calcification is found adjacent to the leptomeningeal lesions in a pericapillary distribution in the fourth cortical layer, possibly secondary to chronic tissue hypoxia. Non-contrast MR images in SWS typically show unilateral cerebral atrophy with enlargement of the ipsilateral subarachnoid spaces and ventricular cavities. Enlarged tubular regions of flow void and gyriform hypointensities (decreased signal on T1-weighted and T2-weighted images) reflect venous collateralization and cortical "serpentine" calcification, respectively. Contrast-enhanced MRI is best for demonstrating the salient CNS vascular abnormalities in SWS. The leptomeninges show intense enhancement overlying the involved cortical region secondary to leptomeningeal (pial) angiogenesis or a compromised blood-brain-barrier secondary to chronic cortical ischemia. This meningeal enhancement often extends well beyond the region of parenchymal atrophy.

**Reference:**

**Question(s) 453:** Pathology

**Discussion:**
The photomicrographs show large groups of atrophic fibers as well as smaller groups of normal-to-hypertrophic fibers. Histochemical sections show grouping of fiber types and both types of fibers are atrophic. These findings are characteristic of denervation, such as that seen in spinal muscular atrophy.

**Reference:**

**Question(s) 454:** Pathology

**Discussion:**
Bilateral necrosis of the globus pallidus is often due to carbon monoxide intoxication.

**Reference:**

**Question(s) 455:** Radiology

**Discussion:**
The magnetic resonance venograms clearly show the straight sinus (D), basal vein of Rosenthal (E), and internal cerebral vein (G).
Question(s) 456: Radiology
Discussion:
The dark foci in the axial T1-weighted image represent flow void ("black blood") in collateral vessels due to occluded distal internal carotid arteries and collateral circulation. The MRA demonstrates lack of flow in the distal internal carotid artery and multiple collateral vessels in the region of the flow void seen in the axial T1-weighted image. These are classic findings in moyamoya syndrome.

Reference:

Question(s) 457: Radiology
Discussion:
The features seen on the T2-weighted MR images are classic for multiple sclerosis (MS). Numerous periventricular lesions are appreciated above and below the tentorium cerebelli. The lesions are greater than 5 mm. The corpus callosum is also involved. These findings confer a greater than 95% specificity for MS. Emboli would affect the corticomedullary junction. Normal pressure hydrocephalus would demonstrate periventricular increased signal uptake rather than multiple lesions.

Reference:

Question(s) 458: Radiology
Discussion:
The unenhanced CT images demonstrate a large amount of blood located in the subarachnoid space of the Sylvian fissure and adjacent sulci. On the more inferior of the two images, there is a rounded density consisting of a middle cerebral artery aneurysm.

Reference:

Question(s) 459: Radiology
Discussion:
The sudden onset following head trauma excludes early onset of Alzheimer's disease. Unfortunately, the patient was misdiagnosed as early Alzheimer's disease and went untreated. Because there is no gradual deterioration following trauma, it is not post-concussion syndrome nor traumatic brain injury with residual dementia. Although alcohol-related dementia is associated with enlargement of the ventricles, as shown, there was no drinking history. The most likely diagnosis is hydrocephalic dementia. The CTs clearly show enlargement of the ventricles over the three years between scans as well as dilated temporal horns (which are supportive of the normal pressure hydrocephalus diagnosis). The history of onset of dementia is of great importance in the diagnosis. In addition, the patient had severe "frontal lobe" signs, grasp reflexes, spastic paraparesis, incontinence, severe ataxia, and bilateral Babinski signs when he was eventually diagnosed. The fact that there was only a minimal degree of cortical atrophy present on follow-up imaging also militates against the Alzheimer's disease diagnosis he was given.

Reference:

Question(s) 460: Radiology
Discussion:
The axial fat-suppressed gadolinium-enhanced T1-weighted image shows marked enhancement of the entire length of the optic nerve, including the optic nerve head. In addition, there is an area of middle cerebellar peduncle enhancement on the contralateral side. The presence of a second lesion in the middle cerebellar peduncle makes multiple sclerosis the most likely disease. In Devic's disease, the brain is usually normal on MRI studies.

Reference:
Question(s) 461: Radiology
Discussion:
The enhanced CT image demonstrates multiple irregular ring-enhancing lesions adjacent to and compressing the lateral ventricles. There is extensive white matter vasogenic edema in both hemispheres. This appearance is most consistent with a glioblastoma that has spread through the corpus callosum to involve both hemispheres. In immunocompromised patients, lymphoma can have a similar appearance, but in immunocompetent patients, lymphoma is most commonly seen as a homogenously enhancing mass. Herpes encephalitis in adults nearly always affects the temporal lobes. Neurocysticercosis can occur in the brain parenchyma or as cysts within the ventricles. First seen as cysts in the brain, cysticercosis can incite an inflammatory reaction when the larva dies, but the lesions are usually small, well circumscribed, and located at the corticomedullary junction.

Reference:

Question(s) 462: Radiology
Discussion:
The lesion seen on the CT scan is a colloid cyst. Colloid cysts originate in the primitive neuroepithelium of the tela choroidea. They are virtually always located in the anterior third ventricle at the foramen of Monro, where they may cause unilateral or bilateral symptomatic hydrocephalus, most often in the third to fifth decade of life. Pathologically, colloid cysts are smooth, well-circumscribed cysts attached to the third ventricular roof, filled with a viscous material that is composed of secretory and catabolic products derived from the epithelial lining cells such as fat, cholesterol crystals, blood degradation products, and CSF. Colloid cysts are usually hyperdense on noncontrast CT scans. They have a highly variable MRI appearance but are most commonly homogeneously hyperintense on T1-weighted images, proton density-weighted images and flair sequences, and are hypointense on T2-weighted images. Variations in the contents of colloid cysts account for their variable MRI appearance. A mixed signal lesion, with a hyperintense rim on T1- and T2-weighted images and a core that is isointense on T1-weighted images and hypointense on T2-weighted images is also common. When obstructive hydrocephalus occurs, a rim of hyperintensity on proton density- and T2-weighted images is found surrounding the lateral ventricles, representing aberrant transependymal CSF flow.

Reference:

Question(s) 463: Anatomy
Discussion:
A lesion of the fourth cranial nerve results in superior oblique weakness with resultant diplopia.

Reference:

Question(s) 464: Radiology
Discussion:
The scans show multiple small ring-enhancing lesions, consistent with cerebral abscesses, located at the supratentorial gray-white junction. In addition, a posterior occipital enhancing lesion is noted. The appearance is most consistent with septic emboli. By the imaging appearance alone, cerebral metastases are not ruled out. In the current case, the patient had endocarditis secondary to intravenous drug abuse. Uncomplicated pyogenic meningitis typically shows intense meningeal enhancement of the cerebral convexity, tentorium, and falx.

Reference:
**Question(s) 465**: Radiology  
**Discussion**:  
The sagittal two-dimensional phase contrast venogram with a velocity encoding of 15 cm/s is tailored to demonstrate slow flow such as that in the dural venous sinuses. The scan demonstrates practically no flow in the superior sagittal sinus (SSS). One prominent cortical vein shows flow. Just adjacent to this cortical vein, minor flow is seen in the SSS. This finding is consistent with near occlusion of the SSS. Higher velocity encodings are needed to demonstrate arterial flow.

**Reference**:  

**Question(s) 466**: Radiology  
**Discussion**:  
The axial and sagittal MRI scans show a Chiari type I malformation, with cerebellar tonsil herniation greater than 5 mm through the foramen magnum. The additional anatomical malformations that would be seen with the other types of Chiari malformations are not present in this case.

**Reference**:  

**Question(s) 467**: Radiology  
**Discussion**:  
Bilateral white matter lesions are identified that relatively spare the cortex and do not exert mass effect. These most likely represent progressive multifocal leukoencephalopathy. HIV encephalitis usually spares the U-fibers. Toxoplasmosis and primary CNS lymphoma would be associated with some mass effect.

**Reference**:  

**Question(s) 468**: Physiology  
**Discussion**:  
Viscerosensory auras consist of abdominal or epigastric sensations, and are characteristic of the onset of temporal lobe seizures. These may be followed by loss of awareness, oral-alimentary or manual automatisms, or dystonic postures. Dystonic postures are usually contralateral to the ictal focus.

**Reference**:  

**Question(s) 469**: Anatomy  
**Discussion**:  
Climbing fibers originate in the inferior olivary complex and appear to have glutamate as their neurotransmitter. Each climbing fiber possesses an extensive all-or-none excitatory connection with Purkinje cell dendrites in the cerebellar cortex.

**Reference**:  

**Question(s) 470 - 472**: Anatomy  
**Discussion**:  
The cutaneous sensory distribution of the deep peroneal, saphenous, and anterior cutaneous (femoral) nerves, are illustrated.

**Reference**:  

**Question(s) 473 - 475**: Radiology  
**Discussion**:  
The lesion is a cavernous angioma (CA), a type of intracranial vascular malformation. CAs, also known as cavernous hemangiomas or cavernomas, are congenital anomalies in blood vessel development, resulting in a tight collection of sinusoidal vasculature, containing hemorrhage in multiple stages of evolution. 80% are supratentorial, commonly in a lobar location close to the subarachnoid space or ventricles. On pathologic examination, CAs consist of a compact
cluster of calcific vascular channels, containing thrombosis and recurrent, seeping hemorrhages, with little or no intervening normal brain parenchyma. The immediately surrounding parenchyma is hemosiderin-stained. MRI findings in CAs that have not developed a frank acute hematoma are highly characteristic. A reticulated core of hyperintensities is noted on T1-weighted, PD, and T2-weighted image, representing extracellular methemoglobin ("popcorn" or "honeycomb" appearance). Complete rings of hypointensities on T1-weighted, PD, and T2-weighted image are noted, representing hemosiderin. The hypointense rim is most robust on T2-weighted image, due to susceptibility effect. There is usually no mass effect, edema, or evidence for associated aberrant feeding or draining vessels.

Reference:

Question(s) 476 - 478: Radiology
Discussion:
MRI lumbar spinal anatomy questions showing the inferior articular facet (solid black arrow), thecal sac (open white arrow) (the spinal cord does not normally descend to this level), and the S1 root (open black arrow).

Reference:

Question(s) 479 - 480: Radiology
Discussion:
The scans show multiple cerebral abscesses. Abscesses have distinct MRI features of the core and rim. The core, containing central liquefactive necrosis and pus is usually hypointense on T1-weighted image and hyperintense on T2-weighted image, with signal characteristics similar to the surrounding edema. The core is not as hypointense as CSF on T1-weighted image. The rim, containing granulation and fibrocollagenous tissue, often has a distinguishing feature in the subacute, mature stage on MRI. It is isointense to mildly hypointense on T1-weighted image and iso-intense to hypointense on T2-weighted image. Therefore, on T2-weighted image, the rim is well visualized as a separator between the abscess core and the surrounding vasogenic parenchymal edema. This feature may help to distinguish an abscess from other ring-enhancing masses such as glioblastomas. Various mechanisms have been proposed to explain the relative hypointensity of the abscess capsule on T2-weighted image. Fibrous collagen, hemorrhage, and iron have been proposed as possible causes. However, more likely, this hypointensity on T2-weighted image reflects the continuous production of paramagnetic substances, such as free radicals or enzymes, by phagocytosing macrophages. The associated edema is usually copious, even if the abscess itself is small. After contrast administration, intense enhancement of the capsule is noted, without enhancement of the core. The enhancement is smooth and usually fully encloses the core ("complete ring"). The presence of a daughter abscess, connected to the larger parent abscess, is a very helpful clue to the diagnosis.

Reference: