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


**Question(s) 1:** Physiology  
**Discussion:**  
The clinical description is classic for carpal tunnel syndrome. Of the choices, palmar sensory studies are probably the most sensitive in mild carpal tunnel syndrome.

**Reference:**  

**Question(s) 2:** Behavioral  
**Discussion:**  
Patients with prosopagnosia often fail to identify objects as well as faces, particularly when there are many members in a category (cars), and when unique identification (my car) is required. Such patients typically have no difficulty identifying gender, age, and emotional state. Famous and well known individuals may be identified by voice, although visual identification may be impossible. Such patients may not be able to identify immediate family by sight alone, but a unique physical characteristic or the sound of their voice may provide clues that enable identification.

**Reference:**  

**Question(s) 3:** Anatomy  
**Discussion:**  
Myelination begins in the peripheral nervous system. The motor roots myelinate before the sensory roots. Still before birth, myelin appears in the central nervous system in components of major sensory systems and components of major motor systems. Myelination in commissures continues to occur well after birth.

**Reference:**  

**Question(s) 4:** Pathology  
**Discussion:**  
The histologic hallmarks of Alzheimer's disease are neuritic plaques and 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 is also usually present. Alzheimer type II astrocytes, in contrast, are reactive astrocytes that are seen in hyperammonemic conditions and are not a feature of Alzheimer's disease. Ballooned neurons are a histological feature of corticobasal degeneration.

**Reference:**  

**Question(s) 5:** Pathology  
**Discussion:**  
Peripheral neuropathies can be caused by inorganic arsenic, lead, mercury and thallium. Aluminum intoxication can produce an encephalopathy, but does not produce a neuropathy. Cyanide, manganese and bismuth do not cause peripheral neuropathy.

**Reference:**  

**Question(s) 6:** Anatomy  
**Discussion:**  
The radial glia participate in cell-cell interactions necessary for neuronal migration.

**Reference:**  

**Question(s) 7:** Anatomy  
**Discussion:**  
Facial pain, thermal sensibility, fine touch and position sense all converge for the first time in the ventroposterior medial nucleus of the contralateral thalamus. The chief sensory nucleus of cranial nerve V serves fine touch, the spinal nucleus of cranial nerve V serves pain and thermal sensibility.
and the mesencephalic nucleus of cranial nerve V serves position sense. These sensory modalities converge in the contralateral thalamus in the ventroposterior medial nucleus of the thalamus while sensory information from the contralateral trunk and extremities converges on the ventroposterior lateral nucleus of the thalamus.

Reference:

Question(s) 8: Clinical Adult
Discussion:
Primary CNS lymphoma are multiple in at least 50% of cases. Meningiomas are multiple in less than 10% and multicentric glioblastoma multiforme occur in 5% of the cases.

Reference:

Question(s) 9: Clinical Adult
Discussion:
Donepezil has been reported to improve cognitive function in patients with MS.

Reference:

Question(s) 10: Anatomy
Discussion:
The medial septal nucleus provides cholinergic innervation to the hippocampus. The nucleus basalis of Meynert provides cholinergic inputs to the amygdala and the neocortex.

Reference:

Question(s) 11: Anatomy
Discussion:
Both the superior orbital fissure and the cavernous sinus contain cranial nerves III, IV, VI, V1, and sympathetic nerve fibers. V2 does not travel through the superior orbital fissure. A lesion in neither location would produce facial anhidrosis.

Reference:

Question(s) 12: Pharmacology/Chemistry
Discussion:
Horseradish peroxidase is taken up at nerve terminals by endocytosis, and is subsequently transported to cell bodies by retrograde axonal transport. Its reaction product is readily visualized histochemically.

Reference:

Question(s) 13: Clinical Pediatrics
Discussion:
This pattern of weakness is due to hypoplasia of the depressor anguli oris muscle. This can be associated with cardiac involvement (cardiofacial syndrome or “Cayler syndrome”).

Reference:

Question(s) 14: Physiology
Discussion:
At the resting membrane potential, potassium has the highest intracellular concentration, with sodium next, then
chloride, then calcium.

Reference:

Question(s) 15: Clinical Adult Discussion:
Cluster headache is characterized by excruciating unilateral pain, usually centered on one eye or temple, lasting 30-120 minutes. Patients average 1-2 headaches a day for a period of 4-8 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) 16: Behavioral Discussion:
The Wisconsin Card Sort Test, which challenges a patient to change cognitive sets without warning, is particularly sensitive to frontal damage. Language skills may be unaffected, and vocabulary is often spared. Face recognition and visual perception abnormalities such as hemi-inattention syndromes are most often associated with damage to the parietal, temporal, or occipital lobes, rather than the frontal lobes. Therefore, the best answer is the WCST.

Reference:

Question(s) 17: Pharmacology/Chemistry Discussion:
Neurons synthesizing nitric oxide (NO) are abundant in the nervous system. They utilize L-arginine as a substrate and activity of NO synthase is activated by intracellular Ca2+/calmodulin. They are selectively spared in hypoxia-ischemia and Huntington’s disease.

Reference:

Question(s) 18: Clinical Adult Discussion:
The most common side effects of modafinil are mania and depression.

Reference:

Question(s) 19: Pharmacology/Chemistry Discussion:
Heparin interacts with antithrombin III and accelerates the rate of neutralization of thrombin.

Reference:

Question(s) 20: 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) 21: 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) 22: Physiology
Discussion:
Significant membrane depolarization is caused by elevated extracellular potassium ions.

Reference:

Question(s) 23: Behavioral
Discussion:
Bilateral tonic or clonic movements in association with preserved consciousness is characteristic of supplementary motor area seizures. Bicycling movements and other asynchronous bilateral movements may occur with this type of epilepsy. The gradual onset and prolonged duration of a spell is more suggestive of pseudoseizures, and are not common with supplementary motor area seizures. Pelvic thrusting and side-to-side head movements are suggestive of pseudoseizures.

Reference:

Question(s) 24: Anatomy
Discussion:
A unilateral lesion of the vagus nerve produces ipsilateral paralysis of the soft palate, pharynx and larynx.

Reference:

Question(s) 25: 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) 26: Clinical Adult
Discussion:
Laceration of the middle meningeal artery is the typical basis for epidural hematoma.

Reference:

Question(s) 27: Behavioral
Discussion:
Presence of the E4 allele for apolipoprotein E is associated with earlier age of onset of Alzheimer's disease, with the presence of one E4 allele associated with a typical onset in the mid-70's and the presence of two E4 alleles associated with typical onset before age 70.

Reference:

Question(s) 28: Clinical Adult
Discussion:
Tacrine and donepezil are approved for the treatment of probable Alzheimer's disease. Both produce a small but significant improvement in cognitive function.
Reference:

Question(s) 29: Behavioral
Discussion:
Selective serotonin reuptake inhibitors (SSRI) have emerged as the treatment of choice for obsessive-compulsive disorder.

Reference:

Question(s) 30: Physiology
Discussion:
The gamma loop functions by changing the length of the intrafusal fibers.

Reference:

Question(s) 31: Physiology
Discussion:
The clinical presentation is typical for hereditary neuropathy with liability to pressure palsy and inherited brachial plexus neuropathy-two genetically distinct disorders. Mayo Clin Proc 19954;70:743-746.


Question(s) 32: Pharmacology/Chemistry
Discussion:
Postganglionic parasympathetic neurons innervating the exocrine glands utilize acetylcholine, which activates secretion via muscarinic M3 receptors. In addition, these neurons release vasoactive intestinal polypeptide (VIP), which potentiates secretion due to its potent vasodilator effect.

Reference:

Question(s) 33: Behavioral
Discussion:
Aphemia is distinguished from Broca's aphasia, transcortical motor aphasia (TCMA), and mixed transcortical aphasia (MTCA) by preserved writing. Writing is impaired in Broca's, TCMA and MTCA.

Reference:

Question(s) 34: Clinical Adult
Discussion:
Restless legs syndrome is characterized by unpleasant limb sensations that are worst at rest, resulting in a compulsion to move the limbs. Most patients have insomnia. About 80% of patients also experience periodic limb movements of sleep. Medications that have been shown to be effective in restless legs syndrome include carbidopa-levodopa, dopamine agonists, opioids, benzodiazepines, anticonvulsants, and clonidine hydrochloride.

Question(s) 35: 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. Affective expression is better preserved in TLE than in schizophrenia, where affecting blunting is common.


Question(s) 36: Physiology
Discussion: The patient’s examination localizes to the right L5 root. Both the gluteus medius and peroneus longus muscles receive their major innervation from the L5 nerve root.


Question(s) 37: Anatomy
Discussion: 14-3-3 protein is elevated in prion disorders in contrast to most other dementing disorders. The protein elevation reflects neuronal destruction and so can be elevated in any disease that results in rapid neuronal death such as trauma, stroke, or encephalitis; therefore, consideration of the clinical context is important.


Question(s) 38: Anatomy
Discussion: The upper thoracic (T1-T4) and the first lumbar segments are in vascular watershed zones and are, therefore, the most vulnerable areas of the cord to ischemic insult.


Question(s) 39: Anatomy
Discussion: Verbal asomatognosia is a category of neglect in which the patient denies ownership of a limb contralateral to a lesion of the supramarginal gyrus of the parietal lobe (usually nondominant).


Question(s) 40: Anatomy
Discussion: A lesion of the pathway connecting the red nucleus, inferior olivary nucleus, and dentate nucleus (Guillain-Mollaret triangle) results in palatal myoclonus.


Question(s) 41: Anatomy
Discussion: Visual attention is thought to be mediated by structures, including the pulvinar, claustrum, and superior colliculus.

Reference: Kandel ER, Schwartz JH, Jessel TM.
Question(s) 42: Anatomy
Discussion:
Dressing apraxia is seen in non-dominant parietal lobe lesions.

Reference:

Question(s) 43: Physiology
Discussion:
The EEG in the postictal state after a generalized seizure shows a low amplitude record with irregular slowing.

Reference:

Question(s) 44: Anatomy
Discussion:
The blood supply of the corticospinal tract is via the anterior spinal artery which is fed by the vertebral and radicular arteries. There is a prominent anastomotic area near T4, not T10.

Reference:

Question(s) 45: Anatomy
Discussion:
The radial nerve supplies the extensor pollicis longus and brevis, abductor pollicis longus, and extensor digitorum longus. The abductor pollicis brevis is innervated by the median nerve.

Reference:

Question(s) 46: Anatomy
Discussion:
The mesencephalic nucleus of the trigeminal nerve mediates jaw proprioception.

Reference:

Question(s) 47: Clinical Adult
Discussion:
Leprosy affects more than 10 million people world-wide, and in all cases peripheral nerves are affected. It is most common in Africa, Asia, and Latin America.

Reference:

Question(s) 48: Behavioral
Discussion:
Bromocriptine is a dopamine receptor agonist, which activates post-synaptic dopamine receptors. It is the treatment of choice for neuroleptic malignant syndrome, a life-threatening reaction to neuroleptic medication to which elderly patients have a relative vulnerability.

Reference:

Question(s) 49: Neuroimaging
Discussion:
Neurofibromatosis is an autosomal dominant syndrome with two distinct types: 1 and 2. Neurofibromatosis type 1 (NF1) (von Recklinghausen's disease): cutaneous cafe au lait spots, hamartomas of iris and multiple subcutaneous nodules (peripheral nerve neurofibromas) are characteristic
features. Abnormal high signal areas especially in the basal ganglia are often seen. They are most likely hamartomas. The abnormal gene is in the long arm of 17. Neurofibromatosis 2 (NF2): bilateral acoustic Schwannomas other cranial nerve tumors, spinal meningiomas, paraspinal neurofibromas and spinal cord ependymomas. NF2 is more rare. Defect: deletion of genetic material from chromosome 22. This incidence is 1 in 50,000, whereas NF1 is 1 in 3,000. There is higher incidence of aneurysms, lytic bone tissue lesions (but not sclerotic). Corpus callosum is not affected.

Reference:

Question(s) 50: Pharmacology/Chemistry
Discussion:
Midodrine is a prodrug that is converted in the liver to an alpha agonist. Its predictable absorption and pharmacokinetics makes it the drug of choice for treatment of orthostatic hypotension unresponsive to fludrocortisone.

Reference:

Question(s) 51: Anatomy
Discussion:
One of the rare but fascinating manifestations of frontotemporal dementia is the emergence of new artistic abilities.

Reference:

Question(s) 52: Physiology
Discussion:
Photoreceptors are normally depolarized in the dark because of persistent opening of the Na+ channels.

Reference:

Question(s) 53: Physiology
Discussion:
Myotonic dystrophy has distal greater than proximal weakness.

Reference:

Question(s) 54: Pathology
Discussion:
Angiotropic (intravascular) lymphoma is a disease of multiple organs that involves the central nervous system in greater than 30% of cases. It is usually a B cell lymphoma by immunophenotyping, and causes multiple small brain infarcts, often of deep white matter. A patient with CNS involvement by angiotropic lymphoma may show lymphoma cells only within the blood vessels and may not have either intraparenchymal lymphomatous masses or significant meningeal involvement. The old name for this tumor prior to the advent of specific immunohistochemical markers for lymphoma was "malignant angioendotheliomatosis", reflecting the erroneous belief that the tumor cells were endothelial in origin.

Reference:

Question(s) 55: Behavioral
Discussion:
Approximately 70% of Rett's patients develop seizures in early childhood. In autism, about 25% of patients develop
seizures usually in later adolescence.

Reference:

Question(s) 56: Physiology
Discussion:
Myotonic potentials are the result of repetitive discharge of the muscle membrane.

Reference:

Question(s) 57: Behavioral
Discussion:
Asperger's syndrome is manifested by impairments in two core features: social interaction and motor behavior which often results in repetitive stereotopies or mannerisms or preoccupation with particular items of interest. These patients have a very difficult time "reading" the emotional gestures or body language of others and have significant impairment in abstract concepts such as getting the punch line of a joke. They usually have normal intellectual and language skills and do not have the thought disorder, psychosis, or paranoia often seen in schizophrenia and related disorders. They lack the autonomic features and anxiety that often accompanies a social phobia.

Reference:

Question(s) 58: Physiology
Discussion:
Slow repetitive stimulation (2 to 3 Hz) in a hand muscle causes a decremental response in the majority of patients with LES. Brief sustained exercise and rapid (20 to 30 Hz) repetitive stimulation are both likely to result in facilitation of the compound muscle action potential amplitude in a patient with LES.

Reference:

Question(s) 59: 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) 60: Behavioral
Discussion:
Defective repetition may be associated with Broca's aphasia, Wernicke's aphasia, conduction aphasia and global aphasia. Intact repetition is preserved in transcortical sensory aphasia.

Reference:

Question(s) 61: Behavioral
Discussion:
Toluene toxicity is often the result of "recreational" abuse by sniffing and inhaling fumes from spray paint cans. Patients who chronically inhale toluene vapors develop dementia (clinically consistent with subcortical dementia), cerebellar ataxia and long tract findings, and in some cases cranial nerve palsies. It is not uncommon for these patients to also manifest a paranoid psychosis. MRI demonstrates a diffuse leukoencephalopathy, cerebral atrophy, and T2 hypointense lesions of the thalamus and/or the basal ganglia.

Reference:
Filley CM. The behavioral neurology of white matter. New York: Oxford University Press,
Question(s) 62: Neuroimaging
Discussion:
In the extracellular space there is relatively free diffusion of protons whereas in the intracellular space diffusion is somewhat restricted. The effective diffusion coefficient of tissue water is determined by a weighted average, which reflects the volume fractions between intracellular and extracellular water. The interruption of cerebral blood flow results in rapid (within minutes) breakdown of energy metabolism and ion exchange pumps. This leads to a massive shift of water from the extracellular into the intracellular compartment (cytotoxic edema) and produces a typical high-intensity area on diffusion-weighted images. Early (within the first 6 hours after stroke) CT signs of brain ischemia are subtle and difficult to detect. On conventional MR images, early (within the first 6 hours after stroke) morphologic signs (produced by tissue swelling) are detected in 50% of acute infarctions; however, signal abnormalities are often not detected. With diffusion-weighted imaging of acute infarction (within the first 6 hours after stroke), 94% sensitivity and 100% specificity have been reported.

Reference:

Question(s) 63: 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) 64: 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) 65: Pharmacology/Chemistry
Discussion:
The N-methyl-D-aspartate (NMDA) receptor requires binding of glycine for its activation, is inhibited by Mg2+ in a voltage-dependent manner, is activated by polyamines and inhibited by Zn2+. Ligands to the phencyclidine-like anesthetic site, e.g., MK-801, inhibit the NMDA receptor.

Reference:

Question(s) 66: 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) 67: Clinical Pediatrics
Discussion:
Rasmussen's syndrome usually presents in childhood with uncontrollable focal seizures that rarely, if ever, respond to anticonvulsants or other medical treatments.
Epilepsia partialis continua is a frequent presentation of this disorder. The disease is progressive and usually involves only one hemisphere. Histologically, perivascular lymphocytic infiltrates with vascular injury, astrogliosis, neuronal loss and cortical atrophy are seen. Hemispherectomy is the best established treatment but there is increased interest in plasmapheresis because anti-glutamate receptor antibodies are seen in some cases.

Reference:

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**Question(s) 68: Pathology**

**Discussion:**
When the axon of a spinal nerve is severed, the axon and its myelin sheath break down and myelin fragments are found in macrophages. The neuronal cell body swells as smooth endoplasmic reticulum, dense bodies, Golgi membranes, vesicles and neurofilaments accumulate and push the nucleus and the Nissl substance to the margins of the cell, a process called central chromatolysis.

Reference:

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**Question(s) 69: Behavioral**

**Discussion:**
The failure to interpret another's facial gestures or "read" another's body language or moods is usually do to impairment in the right orbitofrontal and medial temporal areas (especially with involvement of the right amygdala). Hypoorality is not a trait of the frontotemporal dementias, and semantic aphasia, verbal memory deficits, and nascent artistic traits are all features of left frontotemporal involvement.

Reference:

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**Question(s) 70: Anatomy**

**Discussion:**
The sciatic nerve passes underneath the piriformis muscle and may be entrapped at that location.

Reference:

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**Question(s) 71: Anatomy**

**Discussion:**
The medial forebrain bundle conveys impulses from the lateral hypothalamus rostrally to the nuclei of the diagonal band and to the medial septal nuclei.

Reference:

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**Question(s) 72: Anatomy**

**Discussion:**
The caudal (posterior) neuropore closes on about day 25 or 26 (25 to 26 somite stage).

Reference:

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**Question(s) 73: 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) 74: Anatomy
Discussion:
The Mollaret cell is an activated monocyte series cell. It reflects inflammation and is not an absolutely specific CSF finding in recurrent aseptic meningitis.

Reference:

Question(s) 75: Behavioral
Discussion:
Methyl mercury intoxication may occur through the ingestion of fish that have been exposed to the compound in industrial waste. It causes a subacute syndrome manifested by confusion, apathy, lethargy, dementia, irritability, personality change, constricted visual fields, and a prominent diffuse cerebellar syndrome due to destruction of granule cells of the cerebellum. It can be treated with penicillamine or dimercaptosuccinic acid. BAL should not be used because it causes increased concentration of mercury in the brain.

Reference:

Question(s) 76: Anatomy
Discussion:
Dopamine is the neurotransmitter of a subgroup of amacrine cells.

Reference:

Question(s) 77: Neuroimaging
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 prostheses 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) 78: Anatomy
Discussion:
Unilateral interruption of the medial longitudinal fasciculus (MLF) will result in failure of ipsilateral eye adduction on attempted horizontal gaze. The MLF links together the cranial nerve nuclei concerned with ocular motility and thereby permits conjugate gaze. Conjugate gaze movements occur in response to the cerebral cortex, vestibular nuclei and head and neck proprioception. When the frontal eye fields initiate volitional horizontal gaze, the command is relayed to the contralateral paramedian pontine reticular formation near the sixth nerve nucleus leading to abduction. The command immediately crosses the midline and ascends in the MLF to the oculomotor nucleus to effect adduction. Interruption of these ascending fibers of the MLF results in failure of adduction of the eye ipsilateral to the MLF defect.

Reference:
2. Brazis PW, Masdeu JC, Biller J.

Question(s) 79: Pharmacology/Chemistry
Discussion:
The excitatory amino acid neurotransmitters glutamate and aspartate are the primary neurotransmitters employed by the projection neurons of the cerebral cortex. Acetylcholine and dopamine are used by the extra-thalamic afferent pathways from nuclei of the brainstem and basal forebrain. Glycine is the primary inhibitory amino acid neurotransmitter used in the spinal cord. Nitric oxide is used for local circuit modulation rather than projection.

Reference:

Question(s) 80: Anatomy
Discussion:
Fibers of the autonomic sympathetic nervous system that innervate the pupillodilator muscle leave the neuraxis at C8-T2.

Reference:

Question(s) 81: Physiology
Discussion:
Patients with sensory neuronopathy and associated lung carcinoma frequently have anti-Hu antibodies. Anti-Jo1 antibodies are associated with polymyositis, acetylcholine receptor antibodies with myasthenia gravis, calcium channel antibodies with Lambert-Eaton myasthenic syndrome, and anti-amphiphysin antibodies with stiffman's syndrome.

Reference:

Question(s) 82: Pharmacology/Chemistry
Discussion:
Up to 1% percent of patients treated with the antipsychotic drug clozapine for several months experience bone marrow suppression.

Reference:

Question(s) 83: Clinical Adult
Discussion:
Adrenoleukodystrophy is an x-linked disorder characterized by defective beta oxidation and accumulations of very-long chain fatty acids. This metabolic defect is detectable in heterozygotes and prenatally. The adult onset phenotype (progressive spastic paraparesis) is called adrenomyeloneuropathy.

Reference:

Question(s) 84: 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 areas, occipital-parietal infarction, sagittal sinus thromboses and in some patients with Alzheimer's disease. It is not seen in frontotemporal dementia.

Reference:
Question(s) 85: Behavioral
Discussion:
Disturbance with the recognition of color in one visual field, hemiachromatopsia, occurs only with inferior, posterior occipital lesions.

Reference:

Question(s) 86: Pathology
Discussion:
Pituitary carcinomas by definition must show systemic spread or cerebrospinal metastases. Even in invasive pituitary adenomas, histological features are not reliable predictors of biological behavior.

Reference:

Question(s) 87: 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) 88: Pharmacology/Chemistry
Discussion:
The clinical picture of fever with progressive mental status changes, especially including bizarre behavior and olfactory or gustatory hallucinations, along with fever and focal neurologic abnormalities strongly suggests herpes simplex encephalitis. The laboratory findings that are helpful in determining the diagnosis are: periodic lateralized epileptiform discharges (PLEDs) in the EEG, temporal lobe abnormalities on CT or MRI, and mild hypoglycemia. Acyclovir is the treatment of choice.

Reference:

Question(s) 89: Physiology
Discussion:
An incremental response to tetanic stimulation indicates a presynaptic defect at the neuromuscular junction. In an infant, the most likely reason for developing this is from intestinal botulism.

Reference:

Question(s) 90: Pathology
Discussion:
Curvilinear profiles and fingerprint bodies in sweat glands and endothelial cells, as seen by electron microscopy, are diagnostic for Batten's disease (neuronal ceroid lipofuscinosis).

Reference:

Question(s) 91: Physiology
Discussion:
Abnormal jitter and blocking on SFEMG is seen in patients with generalized myasthenia gravis. Denervation potentials are rarely seen. Conduction block is seen in demyelinating neuropathies. CMAP facilitation >100% after exercise is indicative of Lambert-Eaton syndrome.
Reference:

Question(s) 92: Anatomy
Discussion:
The subependymal zone is now referred to as “brain marrow” because it is the site of neuropoiesis in the adult brain. Pluripotential stem cells reside in this area immediately beneath the ependyma, and these cells may spawn new neurons and glia. Their contribution to the total population of cells in the mature brain is very modest at best, but they are of intense interest as potential agents of repair or as cells of origin of primary brain tumors.

Reference:

Question(s) 93: 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) 94: Behavioral
Discussion:
Apathy is uncommon with orbitofrontal lesions. Those patients are usually disinhibited or hypomanic. Apathy is more prominent than depression in Alzheimer’s patients, and it is more common in late onset Alzheimer’s. It is a common feature of progressive supranuclear palsy and in patients with Alzheimers disease who also manifest extrapyramidal features.

Reference:

Question(s) 95: Physiology
Discussion:
The combination of fast activity (12 to 14 Hz activity) and delta slowing is most often seen as an anesthetic effect or drug overdose.

Reference:

Question(s) 96: 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) 97: 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:
Discussion:
Long term use of chlorpromazine has been reported to lead to generalized paroxysmal bursts in EEGs.

Reference:

Question(s) 99: Behavioral
Discussion:
Apraxia in the non-dominant limbs to verbal command in the presence of intact strength in all limbs and no language disturbance localizes to the corpus callosum.

Reference:

Question(s) 100: Pathology
Discussion:
Autosomal dominant variants of cavernous angiomas are associated with KRIT1 gene mutation on chromosome 7q. Cavernous angiomas are seldom associated with venous malformations. Von Hippel-Lindau disease is associated with capillary hemangioblastomas, not cavernous angiomas.

Reference:

Question(s) 101: Anatomy
Discussion:
Spinal cord ischemia is a recognized complication of aortic aneurysm repair. The anterior spinal artery territory is very commonly affected resulting in disruption of descending corticospinal tracts and ascending spinothalamic tracts. The dorsal columns are spared with preservation of fine touch and joint position sense.

Reference:

Question(s) 102: Physiology
Discussion:
Hereditary neuropathy with liability to pressure palsies may present in childhood. Other family members may or may not be symptomatic. Electrophysiologic studies reveal conduction blocks at sites of pressure and mild diffuse slowing.

Reference:

Question(s) 103: Anatomy
Discussion:
Homonymous hemianopsia results from lesions occurring in the visual radiations or occipital cortex. As the occipital cortex is approached the visual field defects become increasingly congruent. In addition, occipital cortex lesions characteristically produce macular sparing.

Reference:

Question(s) 104: Behavioral
Discussion:
As a general rule the most common neurobehavioral features seen in patients with mild traumatic brain injury and post-concussion syndrome are difficulty with attention, memory impairment, and executive dysfunction.

Reference:

Question(s) 105: Physiology
Discussion:
The presence of periodic lateralized sharp wave discharges in a patient with a recent febrile illness and onset of seizures would strongly suggest herpes simplex
encephalitis.

Reference:

Question(s) 106: 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) 107: 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) 108: Clinical Adult Discussion:
Cerebral amyloidosis usually affects the elderly and accounts for up to 10% of intracranial hemorrhages. The typical location of the hemorrhages is in the lobar areas. The deposition of beta amyloid protein in the media and adventitia of small meningeal and cortical vessels result in lobar hemorrhages that frequently recur.

Reference:

Question(s) 109: 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) 110: 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) 111: Physiology Discussion:
Sensory nerve action potentials are preserved in radiculopathy but not in lesions distal to the dorsal root ganglion (i.e., brachial plexopathy, ulnar mononeuropathy).

Reference:
Question(s) 112: Behavioral Discussion:
Parkinsonism, nascent artistic ability, and semantic aphasia have all been linked with frontotemporal dementia. Tauopathy is a prevalent finding in many of the frontotemporal forms of dementia. Achromatopsia is not an associated feature. In fact, many FTD patients utilize very vivid colors in their paintings.

Reference: 

Question(s) 113: Behavioral Discussion:
Pick's disease is usually manifested by disinhibition, socially inappropriate behavior, and sometimes the Kluver Bucy syndrome. Although some patients may develop depression, it is far less likely than the incidence of depression in Parkinson's, Wilson's, Huntington's disease and multiple sclerosis.

Reference: 

Question(s) 114: Anatomy Discussion:
The medial lemniscus ascends through the brainstem and terminates in the ventral posterolateral nucleus of the thalamus.

Reference: 

Question(s) 115: 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) 116: Clinical Adult Discussion:
Propranolol is effective for both migraine and essential tremor.

Reference: 

Question(s) 117: Physiology Discussion:
Acetylcholine is the primary postganglionic neurotransmitter at all parasympathetic neuroeffector junctions. Norepinephrine is the primary postganglionic neurotransmitter at most sympathetic neuroeffector junctions.

Reference: 

Question(s) 118: Behavioral Discussion:
Dorsolateral association cortex monitors, judges, facilitates and inhibits activity of distributed complex cortical networks. This is referred to as the "executive function" of the frontal lobes.

Reference: 

**Question(s) 119: Physiology**

**Discussion:**
Both GBS and CIDP are acquired demyelinating autoimmune neuropathies. The CSF and electrodiagnostic findings are quite similar, and cannot distinguish one from the other. The interval between onset of symptoms and disease plateau is the main distinguishing feature, with the vast majority of GBS patients reaching a plateau within four weeks of disease onset.

**Reference:**

**Question(s) 120: Pharmacology/Chemistry**

**Discussion:**
Increased levels of carbamazepine may occur within 1-2 days of the administration of erythromycin; this interaction is clinically important as it may lead to carbamazepine toxicity.

**Reference:**
Leppick, IE. Metabolism of antiepileptic medication: Newborn to elderly. Epilepsia 1992;33 Suppl (4);S32-S40.

**Question(s) 121: Clinical Adult**

**Discussion:**
The best indicator of a benign prognosis in MS is early age of disease onset.

**Reference:**

**Question(s) 122: Pathology**

**Discussion:**
Rhabdoid meningioma is a recently described variant of meningioma that often has aggressive clinical behavior with numerous recurrences and the potential for metastasis. It is a World Health Organization grade III tumor. Many meningiomas have distinctive histological appearances, such as secretory, fibrous, and psammomatous subtypes, but have no adverse prognosis associated specifically with their morphology and are usually WHO grade I tumors.

**Reference:**

**Question(s) 123: 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) 124: Anatomy**

**Discussion:**
Reactivation of Herpes Zoster virus is a common neurologic complication in immunocompromised patients. The virus is latent in sensory ganglia including the sensory ganglia of the facial nerve, the geniculate ganglia. Reactivation of virus in this location produces facial paralysis and occasionally hearing loss and vertigo from viral spread to the 8th nerve. A vesicular rash is typically seen in the external acoustic meatus and lateral tongue.

**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. It is most widely applied for the detection of viruses in the CSF. 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 CSF, then an infection is present. PCR has been most frequently utilized for the diagnosis of mycobacterial and viral diseases, such as those caused by cytomegalovirus, the JC virus of progressive multifocal leukoencephalopathy, varicella zoster virus and Herpes simplex virus I and II.

Reference:

Question(s) 126: Physiology
Discussion:
This patient presented with progressive painless weakness and atrophy in the absence of sensory symptoms. He was found to have both upper and lower motor neuron signs in the symptomatic left arm, but was also found to be weak in the left leg which was essentially asymptomatic. The presentation is very suggestive of motor neuron disease or ALS. The distribution of weakness is not that of a myopathy (proximal), so muscle biopsy and serum CK evaluation would not be helpful. The absence of sensory symptoms or signs argues against a peripheral neuropathy, so nerve biopsy, while likely showing axonal loss, would not be of high yield. Electromyography would allow for demonstration of the distribution of neurogenic abnormalities, and if it showed changes in multiple body regions would be evidence in favor of ALS. Only 5-10% of ALS cases are familial so genetic testing at this stage is not indicated.

Reference:

Question(s) 127: Clinical Adult
Discussion:
Large abnormal mitochondria producing ragged-red fibers are observed in AZT-treated patients suggesting that AZT can induce a toxic mitochondrial myopathy.

Reference:

Question(s) 128: Pathology
Discussion:
Medulloblastoma is especially prone to widespread leptomeningeal dissemination. Occasionally, this can also occur with glioblastoma and ependymoma.

Reference:

Question(s) 129: 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) 130: Physiology
Discussion:
Carbamazepines act by blocking voltage
dependent Na+ channels of the action potential.

Reference:

Question(s) 131: Physiology
Discussion:
Decreased number of large motor units, firing rapidly, is consistent with a chronic motor neuropathy, as seen in old polio. HMSN would have sensory abnormalities in NCS, as would CIDP and tabes dorsalis. Cerebral palsy would give decreased number of normal size MUAPs, firing slowly.

Reference:

Question(s) 132: Clinical Adult
Discussion:
Evidence-based medicine refers to the practice of clinical therapeutics related to information learned from randomized, controlled trials, systematic review or practice guideline generated from comprehensive review of the literature.

Reference:

Question(s) 133: Anatomy
Discussion:
The hypophysis is supplied by two sets of arteries, the superior and inferior hypophyseal arteries. These are branches of the internal carotid arteries.

Reference:

Pharmacology/Chemistry
Discussion:
Lamotrigine does not affect metabolism of carbamazepine or other antiepileptic drugs and therefore is the least likely in this group to be associated with toxicity during polytherapy.

Reference:

Question(s) 135: Pharmacology/Chemistry
Discussion:
Dystrophin is a membrane-bound protein distributed along the intracellular surface of the sarcolemma, and is a member of the superfamily of cytoskeletal proteins; dystrophin interacts with actin and may contribute to structural and functional stability of the plasma membrane.

Reference:

Question(s) 136: Physiology
Discussion:
In the resting neuron, increasing sodium permeability depolarizes the cell.

Reference:

Question(s) 137: Behavioral
Discussion:
Galantamine, memantine, lithium, and vitamin E have all been shown to have some benefit in Alzheimer's disease or experimental animal models of Alzheimer's disease. The combination of estrogen/progestin was shown to have deleterious effects in Alzheimer's disease.

Question(s) 138: Physiology
Discussion:
The decremental response to 2 to 3 Hz repetitive nerve stimulation in patients with myasthenia gravis is due to failure of neuromuscular transmission at a number of end-plates resulting in fewer muscle fiber action potentials contributing to the compound muscle action potential.

Reference:

Question(s) 139: Physiology
Discussion:
N13 wave on tibial SEPs may be absent or attenuated in cervical spondylosis, even when there is minimal clinical evidence on exam. Brainstem lesions or foramen magnum lesions may have prolonged N13 to N20 interval.

Reference:

Question(s) 140: 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) 141: 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) 142: Behavioral
Discussion:
The hyperventilation syndrome is characterized by dizziness and paresthesias. These symptoms may be reproduced by three minutes of hyperventilation in many patients. Depression, migraine, and vertigo are unusual manifestations. Syncope can occur, but is less common than dizziness.

Reference:

Question(s) 143: Behavioral
Discussion:
Pick’s disease may be associated with Kluver-Bucy syndrome and therefore with docility, hyperorality, hypersexuality, hypermetamorphosis, and “psychic blindness”.

Reference:
Question(s) 144: Pharmacology/Chemistry
Discussion:
Nimodipine is a member of the dihydropyridine class of L-type voltage-gated calcium-channel blockers that is used to prevent vasospasm in patients with subarachnoid hemorrhage.

Reference:

Question(s) 145: Anatomy
Discussion:
Hyperprolactinemia results in galactorhea and menstrual cycle disruption in women, and impotence and loss of libido in men.

Reference:

Question(s) 146: Physiology
Discussion:
Hyperventilation reduces intracranial pressure by causing vasoconstriction.

Reference:

Question(s) 147: 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) 148: Anatomy
Discussion:
Functional neuroimaging shows thermal pain is associated with activation of the anterior cingulate gyrus.

Reference:

Question(s) 149: Behavioral
Discussion:
Anterograde episodic memory is most affected in transient global amnesia. Semantic memory (knowledge of worldly facts) is a form of explicit memory. A deficit in semantic memory may be associated with episodic memory deficits when the lesion involves more extensive neocortex especially the inferotemporal association areas.

Reference:

Question(s) 150: Pathology
Discussion:
The injury described above fits best with a contrecoup type injury. The individual decelerates when he falls and strikes the back of his head on the floor. Because the head was in motion and he struck his occiput, the contrecoup sites will be the frontal lobes and temporal tips. The irregular bony contours within the cranial vault in the anterior and middle fossae exacerbate the injury.
Reference:

Question(s) 151: Clinical Adult
Discussion:
The formation of an epidural hematoma may be delayed by minutes to 16 days after injury. Epidural hematomas are more common in younger than in elderly patients because the dura is more adherent to the inner table of the skull in elderly patients.

Reference:

Question(s) 152: 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) 153: 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) 154: Physiology
Discussion:
The most likely finding in a patient with early stages of Alzheimer's disease is a normal background or mild slowing of alpha activity (8 to 9 Hz).

Reference:

Question(s) 155: 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:

Question(s) 156: Anatomy
Discussion:
The anterior choroidal artery is a branch of the internal carotid artery. In addition to the choroid plexus, this artery supplies the optic tract, lateral geniculate nucleus, and much of the posterior limb of the internal capsule. Hemiparesis from involvement of the corticospinal tract and hemisensory loss from involvement of thalamic radiations within the posterior limb of the internal capsule are the most common findings. An incongruous hemianopia may result from involvement of the optic tract.

Reference:

Question(s) 157: Clinical Pediatrics
Discussion:
Trinucleotide repeat expansions have been found in all of these diseases. Friedreich, Huntington, myotonic dystrophy and
dentatorubro-pallidoluysian atrophy (DRPLA) are progressive disorders. Fragile X syndrome is non-progressive.

Reference:

Question(s) 158: Clinical Pediatrics
Discussion:
The risk of a recurrent seizure within 24 months for a child with a single, idiopathic seizure and a normal EEG is 23%.

Reference:

Question(s) 159: Clinical Adult
Discussion:
The acute neurotoxic effects of organophosphates include miosis, salivation, sweating and fasciculations.

Reference:

Question(s) 160: 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) 161: Behavioral
Discussion:
Kluver-Bucy syndrome results from bilateral temporal lesions involving the amyndala nuclei. Clinical features include hypermetamorphosis, hyperorality, hypersexuality, visual agnosia, and blunted emotional affect. Aggression is not a component of the syndrome. Hypermetamorphosis occurs when an individual is overly sensitive or acute aware of minute stimuli in the environment, like a speck of lint on someone's shirt, or a scrap of paper on the floor. Patient's with Kluver-Bucy syndrome may become preoccupied with these stimuli, by touching, picking or examining them, symptoms which are described as hypermetamorphosis.

Reference:

Question(s) 162: Physiology
Discussion:
Both doll's head eye movements and caloric responses require an intact brainstem. Doll's head eye movements have a proprioceptive component and may be present when caloric responses are absent. They are not usually seen in normally awake individuals.

Reference:

Question(s) 163: Physiology
Discussion:
In a patient with suspected multiple sclerosis and a single neurologic event, evoked potentials (especially visual and somatosensory evoked potentials) may reveal a previous demyelinating lesion that may have had minimal if any clinical manifestation and may not be seen on MRI scan.

Reference:
Chiappa KH. Evoked potentials in clinical medicine. 3rd ed. New York: Lippincott
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:

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:

The most common neurologic complication of scleroderma is carpal tunnel syndrome.

Reference:

The tight junctions of the cerebrovascular endothelial cells prevent molecular traffic between endothelial cells, and since CNS endothelial cells are not fenestrated, only molecules that can be transported or are lipophilic can enter the brain. Astrocytes are important in that they signal the endothelial cells to express the unique phenotype seen in the CNS. Pericytes and smooth muscle cells are not major structural elements of the blood brain barrier.

Reference:

Radiating dysesthesias into the little finger suggest a C8 radiculopathy. The flexor carpi ulnaris is largely innervated by the C8 nerve roots.

Reference:

The amygdala plays a critical role in emotional responses, particularly including conditioned fear.

Reference:

Zellweger's syndrome classically presents with dysmorphic facies, seizures, and severe hypotonia and the infants usually die by age six months. The brains often show the relatively unique combination of both pachygyria and polymicrogyria, as well as simplification of the dentate and olivary nuclei.

Reference:
Discussion:
The supraspinatus muscle initiates abduction of the fully adducted upper extremity.

Reference:

Question(s) 172: Clinical Adult
Discussion:
The American Medical Association has recently published guidelines regarding the ethical use of industry-sponsored funds for social gatherings and other offerings provided by pharmaceutical companies to physicians in the interest of endorsing products. Pharmaceutical companies are restricted from providing strictly meals or entertainment to physician families. It is appropriate to accept educational grants from pharmaceutical companies if the source is disclosed and free from bias.

Reference:

Question(s) 173: Clinical Adult
Discussion:
Most idiopathic brain abscesses contain multiple pathogenic organisms all present simultaneously.

Reference:

Question(s) 174: Physiology
Discussion:
Increased jitter is seen in a variety of neuromuscular disorders and is not specific to any particular disease. Jitter is not increased in metabolic myopathies and steroid myopathies.

Reference:

Question(s) 175: Pharmacology/Chemistry
Discussion:
The predominant effect of acetylcholine in the cerebral pyramidal neurons is postsynaptic excitation, mediated via M1 muscarinic receptors. The effector mechanism is inhibition of a variety of potassium currents.

Reference:
Halliwell JV. Physiological mechanisms of cholinergic action in the hippocampus. Prog Brain Res. 1990;84:11-20.

Question(s) 176: 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 a meaningful degree of neurologic function.

Reference:

Question(s) 177: Physiology
Discussion:
Rhythmic 10 to 40 Hz fast activity is most characteristic of the onset of generalized tonic clonic seizures.

Reference:

Question(s) 178: Pharmacology/Chemistry
Discussion:
The anti-GQ1 b antibody is associated with the Miller-Fisher syndrome (ataxia, ophthalmoparesis, and areflexia).
Question(s) 179: Clinical Pediatrics

Discussion:
Therapy of vascular hemangiomas in childhood with interferon alpha has been associated with the development of spastic diplegia. The role of interferon in this process is supported by data showing increased concentrations of interferon in some children with spastic CP, the Aicardi-Goutieries syndrome, and animal studies.

Reference:

Question(s) 180: Behavioral

Discussion:
Speak to the violent patient in a non-threatening manner, and avoid provocative physical gestures. Excessive firmness or demands may provoke additional violence. Physical force should be left to security personnel, who should be involved as soon as possible when a patient escalates to the point of violence. Prolonged, direct eye contact should be avoided.

Reference:

Question(s) 181: 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) 182: Clinical Adult

Discussion:
Mitoxantrone is the only drug approved by the FDA for treatment of secondary progressive MS.

Reference:

Question(s) 183: 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) 184: 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) 185: Physiology
Discussion:
The clinical syndrome of frequent daytime sleepiness and cataplexy is consistent with narcolepsy. Diagnostic criteria for narcolepsy include a mean sleep latency <5 minutes and REM sleep in at least two naps.

Reference:

Question(s) 186: Clinical Adult
Discussion:
Anticholinergic (scopolamine) toxicity is associated with large, poorly reactive pupils.

Reference:

Question(s) 187: Pathology
Discussion:
Corticobasal degeneration classically presents with progressive asymmetric apraxias, rigidity, and/or aphasia due to neuronal loss and atrophy of the peri-Sylvian region. Astrocytic plaques with tau-immunoreactivity are a diagnostic feature of corticobasal degeneration.

Reference:

Question(s) 188: Anatomy
Discussion:
The lateral spinothalamic tract is laminated so that the most lateral and most posterior fibers represent the lowest portion of the body. The entire lateral spinothalamic tract is ventral to the plane of the dentate ligament. This anatomic relationship guides the neurosurgical ablation of the spinothalamic tracts for pain control.

Reference:

Question(s) 189: Behavioral
Discussion:
The occipito-temporal or ventrally directed visual processing pathways are involved in object recognition or the "what" of identification.

Reference:

Question(s) 190: Physiology
Discussion:
P14 is generated in the brainstem.

Reference:

Question(s) 191: 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) 192: 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) 193: Pathology
Discussion:
Rabies spreads to the brain along the nerves. Cytomegalovirus, eastern equine encephalitis, poliomyelitis and subacute sclerosing panencephalitis spread via hematogenous routes in naturally occurring infections.

Reference:

Question(s) 194: 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) 195: Pathology
Discussion:
Alpha-synuclein is a presynaptic protein that forms the major component of the filaments in brainstem Lewy bodies, cortical Lewy bodies, and Lewy neurites seen in Parkinson's disease and diffuse Lewy body disease.

Reference:

Question(s) 196: Pathology
Discussion:
Diffuse axonal injury (DAI) results from shearing forces at the time of trauma. DAI is unrelated to ischemic events, contusions, or penetrating trauma.

Reference:

Question(s) 197: Pathology
Discussion:
Familial frontotemporal dementia that arises from a mutation on chromosome 17 results in a mutation of the gene encoding for tau protein.

Reference:

Question(s) 198: Pathology
Discussion:
PARK1 is parkinsonism caused by point mutations in the alpha synuclein gene on chromosome 4 and shows younger age of onset and a greater degree of dementia than sporadic Parkinson's disease.
**Reference:**

**Question(s) 199:** Physiology

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

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**Reference:**

**Question(s) 200:** Behavioral

**Discussion:**
Bilateral strokes may cause pseudobulbar palsy, which can include pseudobulbar affect (emotional incontinence), dysarthria, dysphagia, and spastic paraparesis. Absence of anhedonia or depressed mood makes major depression, dysthymic disorder, or adjustment disorder with depressed mood unlikely.

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**Reference:**

**Question(s) 201:** Pathology

**Discussion:**
Desmoplastic infantile ganglioglioma (DIG) and desmoplastic infantile astrocytoma (DIA) are large cystic tumors of infants, usually attached to dura, with a generally good prognosis after surgical resection. They are often frim due to the rich reticulin network found throughout the tumor, a feature that gives rise to the name "desmoplastic". Dysembryoplastic neuroepithelial tumors (DNT) usually occur in patients with seizures and occur in older children and young adults; they are not attached to dura. No type of meningioma is characterized by common occurrence in children under two years. Central neurocytoma occurs near the ventricles, with a mean patient age of 29 years.

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**Reference:**

**Question(s) 202:** Anatomy

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

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**Reference:**

**Question(s) 203:** 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.

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

**Question(s) 204:** Physiology

**Discussion:**
Although periodic waveforms can be seen in other conditions, the presence of generalized periodic 1 Hz sharp waves with progressive dementia is strongly suggestive of Creutzfeldt-Jakob disease.

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**Reference:**

**Question(s) 205:** 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, helps to decrease hyperactivity and impulsivity in these patients.

Reference:

Question(s) 206: Pharmacology/Chemistry
Discussion:
Clozapine is the antidopaminergic drug of choice for treatment of psychosis in patients with Parkinson’s disease due to its relative lack of extrapyramidal side effects. It acts on D4 receptors, which are concentrated in the frontal cortex but not in the basal ganglia.

Reference:

Question(s) 207: Physiology
Discussion:
The suprachiasmatic nuclei of the hypothalamus regulates the circadian rhythm.

Reference:

Question(s) 208: Pathology
Discussion:
Central nuclei and preferential type I myofiber atrophy are characteristic features of myotonic dystrophy.

Reference:

Question(s) 209: Clinical Adult
Discussion:
Most medications are ineffective for neurogenic bladder problems of detrusor-sphincter dysynergia and intermittent catheterization is usually the best management.

Reference:

Question(s) 210: Physiology
Discussion:
The musculocutaneous and median nerves are branches of the lateral cord of the brachial plexus.

Reference:

Question(s) 211: Neuroimaging
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) 212:** Behavioral Discussion:
The occipital-parietal or dorsally directed visual processing pathways are functionally correlated with identifying the location of an object in space. This is the “where” pathway of visual processing.

**Reference:**

**Question(s) 213:** Clinical Adult Discussion:
Brain metastases are rare from prostatic neoplasm. Malignant melanoma is uncommon in African-Americans. Meningioma is most commonly dural-based and may not enhance with contrast. Glioblastoma multiforme occurs in older age groups, accounts for more than 50% of all primary brain tumors, and frequently presents with a ring-enhancing lesion on neuroimaging.

**Reference:**

**Question(s) 214:** Anatomy Discussion:
The mammillothalamic tract projects to the anterior nuclear group of the thalamus.

**Reference:**

**Question(s) 215:** Pharmacology/Chemistry Discussion:
Type 1 (Andrade) familial amyloidosis is inherited as an autosomal dominant and is associated with amyloid deposition in peripheral nerves. It is characterized by progressive loss of pain and temperature sensation, lancinating pain, and severe generalized autonomic failure. Mutations have been demonstrated in the gene that codes for transthyretin, the protein that transports thyroxine and retinol binding protein.

**Reference:**

**Question(s) 216:** 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). Bromocriptine treatment causes a decrease the tumor cell cytoplasmic volume without actual killing the adenoma cells. Therefore, the prolactinoma may 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) 217:** 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:**
Discussion:
Valproate can produce hair loss that rarely requires discontinuation of the drug.

Reference:

Question(s) 219: Pathology
Discussion:
Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) results from missense mutations of the Notch 3 gene on chromosome 19. The small deep white matter vessels show replacement of the media by eosinophilic, periodic acid-Schiff positive, Congo red negative, granular material. Ultrastructurally, there is compact electron-dense material known as granular osmophilic material surrounding myocytes in the arterial media. Although the predominant effects of CADASIL are on the central nervous system, it is a systemic vasculopathy that can be appreciated on skin or nerve biopsy.

Reference:

Question(s) 220: 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) 221: 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) 222: Neuroimaging
Discussion:
The acute state of hemorrhage is related to oxyhemoglobin, which appears isointense on T1 weighted imaging, and iso- to hyperintense on proton density and T2-weighted images. Chronic infarction is characterized by ferritin and hemosiderin accumulation, which cause hypointensity on all three imaging sequences.

Reference:

Question(s) 223: Anatomy
Discussion:
The abducens nerves emerge from the ventral surface of the brainstem at the junction between the pons and medulla. Cranial nerves VII and VIII also emerge at the pontomedullary interface, but they do so much more laterally.

Reference:

Question(s) 224: 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.

Reference:

Question(s) 225: Anatomy
Discussion:
Established projections to the globus pallidus arise from the striatum and subthalamic nuclei. The pallidum does not receive direct afferents from the cerebral cortex, dentate nucleus, or thalamus.

Reference:

Question(s) 226: 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) 227: Behavioral
Discussion:
Atypical parkinsonism, eyelid opening apraxia, retrocollis, subcortical dementia including slowness of mental processing and memory retrieval deficit, depression, 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) 228: Behavioral
Discussion:
Nefazodone inhibits activity of cytochrome P450IIIA4, the isoenzyme responsible for metabolism of alprazolam (and also triazolam).

Reference:

Question(s) 229: 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) 230: Physiology
Discussion:
Sawtooth waves are seen on the EEG during REM sleep.

Reference:

Question(s) 231: Physiology
Discussion:
Ethosuximide, used to treat absence seizures, acts on calcium channels in the
thalamus.

Reference:

Question(s) 232: 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) 233: Anatomy
Discussion:
The majority of axons of cerebellar Purkinje cells synapse with neurons in the deep cerebellar nuclei. A few from the vermis and flocculonodular lobule project directly to the vestibular nuclei.

Reference:

Question(s) 234: Clinical Adult
Discussion:
Triptans are now known to cause medication overuse headache.

Reference:

Question(s) 235: 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:

Reference:

Question(s) 236: Clinical Pediatrics
Discussion:
Rett syndrome is seen in young girls and is characterized by developmental arrest, stereotypic hand movements, deceleration of head growth and loss of communication skills. It is an x-linked disorder and 80-85% of classic Rett syndrome patients will have an abnormality in the MECP2 gene, while approximately 50% of "atypical" Rett patients will have an abnormality of this gene.

Reference:

Question(s) 237: Anatomy
Discussion:
The stapedius, buccinator, posterior belly of the digastric, frontalis, as well as other muscles of facial expression are all innervated by the facial nerve. The masseter, temporalis, medial and lateral pterygoids, tensor vell palati, tensor tympani, anterior belly of the digastic and mylohyoid are innervated by the trigeminal nerve.

Reference:

Question(s) 238: Pathology
Discussion:
Degeneration of the cerebellar granular cell neurons is one of the major histopathological findings in organic mercury derivative intoxication.

Reference:
Question(s) 239: 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 occasionally is associated with but perhaps not the direct cause of smaller vessel vasculitis.

Reference:

Question(s) 240: Neuroimaging
Discussion:
The classical secondary sign of neurofibroma is the widening of neural foramina and erosion of pedicle, all seen in plain X-rays, CT or MRI.

Reference:

Question(s) 241: 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) 242: Physiology
Discussion:
In a patient with a traumatic brachial plexopathy, absent ulnar motor and sensory responses and motor median responses would be consistent with lower trunk brachial plexopathy. With C7, T1 root avulsion, denervation in weak, atrophic muscles are found, but there is no abnormality of sensory evoked potentials as the dorsal root ganglia are not involved. Mononeuropathies could have motor and sensory responses involved for each mixed nerve mononeuropathy. A cervical myelopathy would have normal motor and sensory responses.

Reference:

Question(s) 243: Pathology
Discussion:
Atypical teratoid/rhabdoid tumors are typically associated with a signature chromosomal change, ie. chromosome 22 loss. This is associated with an INI-1 gene defect.

Reference:

Question(s) 244: Behavioral
Discussion:
Executive functions of the brain are formulated in heteromodal cortex. Emotional properties of information processing are formulated in paralimbic regions, and specific sensory and simple motor processing occurs in unimodal cortices.

Reference:
Question(s) 245: Clinical Pediatrics  
Discussion:  
Neonatal seizures usually have a serious underlying reason. Treatable causes need to be recognized since delay in treatment can aggravate serious neurologic compromise. Group B Streptococci is the most common cause of bacterial meningitis/septicemia in the neonate and has about a 25% mortality rate and of the survivors about 20% will have severe deficits. If seizures in the neonate are secondary to subarachnoid hemorrhage, there usually is a good outcome. Benign epileptic syndromes in the newborn can present before the third day of life.

Reference:  

Question(s) 246: Physiology  
Discussion:  
The clinical presentation is suggestive of the diagnosis of myotonic dystrophy. Myotonic potentials (spontaneous potentials with waxing and waning amplitude and frequency) on needle electromyography would confirm the diagnostic suspicion. Complex repetitive discharges and fibrillation potentials may be seen in either myopathic or neurogenic disease. Large polyphasic motor unit action potentials with an increased firing rate would be expected in a chronic neurogenic condition. Fasciculation potentials may be seen in the setting of radiculopathy or anterior horn cell disease, but may also be seen in normal subjects.

Reference:  

Question(s) 247: Anatomy  
Discussion:  
The indolamine serotonin occurs largely in neurons whose cell bodies form the median raphe nuclei of the brainstem.

Reference:  

Question(s) 248: 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) 249: Pathology  
Discussion:  
Eosinophilic granular bodies histological features most commonly found in low grade gliomas, such as pleomorphic xanthoastrocytoma. They are small aggregates of eosinophilic, hyaline material situated in the processes of astrocytes.

Reference:  

Question(s) 250: Anatomy  
Discussion:  
The suboccipital nerve derived from C1 is usually purely motor. It terminates in the short posterior muscles of the head (rectus capitis and obliquus capitis).

Reference:  
Question(s) 251: 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) 252: Anatomy
Discussion:
Fornical damage interrupts Papez’s circuit and results in loss of the ability to form new memories.

Reference:

Question(s) 253: Pharmacology/Chemistry
Discussion:
Disorders of vitamin E metabolism are associated with adult-onset cerebellar ataxia. A mutation in the gene encoding for alpha-tocopherol-transfer protein (a-TTP) has been identified in several ataxia families living on a small Japanese island.

Reference:

Question(s) 254: Physiology
Discussion:
Scotopic vision is most sensitive in the dark-adapted eye.

Reference:

Question(s) 255: Physiology
Discussion:
The afferent limb of the reflex involves V1 (trigeminal) and the efferent limb is the motor axons of the facial nerve.

Reference:

Question(s) 256: 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) 257: Physiology
Discussion:
The H reflex is best elicited by submaximal, long-duration stimuli, preferentially exciting Ia afferent fibers. The F wave is maximal with supramaximal stimulation.

Reference:

Question(s) 258: Physiology
Discussion:
Although the H reflex is analogous to the ankle jerk reflex, it bypasses the muscle spindle component. The F wave is produced by antidromic activation of a pool of anterior
horn cells, and does not involve the muscle spindle.

Reference:

Question(s) 259: Clinical Pediatrics
Discussion:
Idiosyncratic toxic reactions to valproic acid include thrombocytopenia, hepatotoxicity and pancreatitis.

Reference:

Question(s) 260: Clinical Pediatrics
Discussion:
Friedreich’s 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:

Question(s) 261: 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) 262: Clinical Pediatrics
Discussion:
Intraventricular hemorrhage in the preterm infant originates almost exclusively from the germinal matrix. In contrast, at term intraventricular hemorrhage arises primarily from veins of the choroid plexus.

Reference:

Question(s) 263 - 266: Anatomy
Discussion:
Cholinergic neurons are in the substantia innominata. Noradrenergic neurons are in the locus ceruleus. Serotonergic neurons are in the median and dorsal nuclei of the raphe. The ventral tegmental area contains dopaminergic neurons.

Reference:

Question(s) 267 - 270: Clinical Adult
Discussion:
The GME Core Competencies for residency education were created by the Accreditation Council for Graduate Medical Education (ACGME) in 1999 and include six areas: Patient Care, Medical Knowledge, Interpersonal Skills and Communication, Professionalism, Practice-based Learning and Improvement and Systems-based Practice in which physician skills are to be taught and monitored during residency training. Practice-based Learning and Improvement involves recognizing individual limitations and the need for life-long learning as a practicing physician, ability to obtain up-to-date information from the literature to assist in quality care of patients and the ability to evaluate case load and practice experience in a systematic manner. Systems-based Practice involves an
awareness of community, national and allied health resources that may enhance patient quality of life, ability to lead a health care team, appropriate utilization of consultations, and awareness of the importance of accurate medical data in effective management of patients.

Reference:

Question(s) 271 - 275: Pharmacology/Chemistry
Discussion:
After presynaptic reuptake, neurotransmitters are metabolized by specific enzymes. Some of the metabolites can be measured in the cerebrospinal fluid as an indirect index of activity of the corresponding neurons. GABA is metabolized by GABA-transaminase to succinic semialdehyde, which serves as a precursor in the Krebs cycle. Monoamines are metabolized by monoamine-oxidases and methyltransferases. The final metabolite of dopamine is homovainillic acid; of norepinephrine is 3-methoxy-4-hydroxy-phenylglycol (MHPG), of serotonin 5-hydroxyindoleacetic acid (5-HIAA), and of histamine methyl-imidazolacetic acid (MIAA)

Reference:

Question(s) 276 - 278: 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 erythematosus, 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) 279 - 284: Pharmacology/Chemistry
Discussion:
Antiepileptic drugs affect the function of neuronal ion channels. Phenytoin, carbamazepine and lamotrigine inhibit Na+ channels (phenytoin by both voltage- and use-dependent mechanisms), ethosuximide reduces the low threshold Ca++ currents (T currents) in thalamic neurons, and benzodiazepines activate the GABA A receptor-coupled Cl- channel. Gabapentin increases the promoted release of GABA, and Vigabatrin inhibits GABA transaminase.

Reference:

Question(s) 285 - 289: Behavioral
Discussion:
Flumazenil is used to reverse overdose of bezodiazepines. Quetiapine is an antipsychotic that may be effective in treating the psychosis associated with dementia. Valproate has been used to treat the rapid cycling of bipolar disorder. Dopaminergic agonists such as methylphenidate has been used to stimulate motivational intent in akinetic mutism patients with medial frontal lesions. The SSRIs such as paroxetine have been effective in treating obsessive-compulsive traits.
Question(s) 290 - 292: Physiology

Discussion:
A pattern of 3 Hz spike-and-wave is consistent with absence seizure. A hypsarrhythmic pattern is seen with infantile spasms. The slow spike-and-wave is seen with the Lennox-Gastaut syndrome. Triphasic waves can be seen with metabolic encephalopathies.

Reference:

Question(s) 293 - 297: 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) 298 - 299: 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:
Bradley WG, Daroff RB, Fenichel GM, et al.

Question(s) 310 - 313: Clinical Adult Discussion:
Systemic lupus erythematosus (SLE) affects the nervous system at every level. Strokes occur in 15% of SLE patients, more often associated with coagulopathy or cardiac valve abnormality than with central nervous system (CNS) vasculitis. At some point in the disease course, 30-54% of SLE patients have seizures. Signs of meningeal inflammation are present at autopsy in 18% of patients, but clinically significant aseptic meningitis is rare. In contrast, symptoms and signs of aseptic meningitis occur in 10-49% of patients with Behcet's disease. Sjogren's syndrome can also affect the nervous system at any level, but peripheral nervous system (PNS) manifestations are more common than CNS manifestations. A variety of patterns of peripheral nerve involvement occur; one of the most distinctive (but not the most common) is pure sensory neuropathy (dorsal root ganglionopathy). Rheumatoid arthritis affects the CNS and PNS primarily by direct compression from skeletal abnormalities. Up to 70% of patients with advanced disease have cervical spine abnormalities, which may have no neurologic manifestations or may produce signs and symptoms of myelopathy.

Reference:

Question(s) 314 - 317: 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) 318 - 319: Anatomy Discussion:
The anterior choroidal artery supplies the anteromedial part of the head of the caudate nucleus. The internal carotid artery sends branches directly to the genu of the internal capsule. The anterior and medial parts of the thalamus are supplied by posteromedial (thalamoperforating) arteries. The hippocampal formation and amygdala receive their blood supply from the anterior choroidal artery.

Reference:

Question(s) 320 - 323: Physiology Discussion:
Hypsarrhythmia is seen in patients with infantile spasms which are often characterized as flexor spasms. The 3 Hz spike-and-wave is seen in patients with absence seizures which are often manifested by staring and eye flutter. Centrottemporal spikes are seen with benign epilepsy of childhood. The slow spike-and-wave is part of the Lennox-Gastaut syndrome which is characterized by slow spike-and-wave, frequent generalized seizures including atypical absence, atonic and myoclonic seizures, and mental retardation. The 6 Hz spike-and-wave is a benign variant that is not associated with epilepsy.

Reference:
Question(s) 324 - 327: 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.


Question(s) 328 - 330: 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 2-12 years. Acute motor axonal neuropathy occurs in epidemics in northern China during summer months, and it has been associated with antecedent Campylobacter jejuni infection.


Question(s) 331: Clinical Adult Discussion: Olfactory manifestations are typical of partial seizures of temporal lobe onset. Carbamazepine, phenobarbital, phenytoin, primidone, and valproic acid are all effective in controlling partial seizures, but carbamazepine was better tolerated and more effective for complex partial seizures than valproic acid in one large study, and phenobarbital and primidone have a significantly higher rate of adverse effects than carbamazepine and phenytoin. Thus, carbamazepine and phenytoin are generally considered first-line drugs for partial complex seizures, but in patients whose seizures are controlled without toxicity on phenobarbital, primidone, or valproic acid there is no reason to change medications.

effective in the treatment of painful diabetic neuropathy. Newer selective serotonin reuptake inhibitors, such as sertraline, have not been convincingly demonstrated to be effective, nor have neuroleptics or beta blockers. Narcotic analgesics are not appropriate first-line therapy. Oral administration of local anesthetic agents, such as mexiletine, has also been demonstrated to be effective in painful diabetic neuropathy. Chlorzoxazone is a muscle relaxant, while phenoxybenzamine is a sympatholytic drug that has been utilized for causalgia. In this patient, dexamethasone might be expected to aggravate the diabetes. Amantadine has no analgesic properties. Capsaicin causes initial release and then subsequent depletion of small peptides, including substance P, in primary afferent neurons. The initial release may be responsible for development of a local burning sensation, which is one of the adverse effects of this drug. The subsequent depletion of substance P presumably results in reduced transfer of nociceptive information to the central nervous system.

Reference:

Question(s) 339 - 340:
Pharmacology/Chemistry
Discussion:
While diazepam, dantrolene, and baclofen all may produce drowsiness, fatigue and nausea, dantrolene is the drug that has a well-defined potential for hepatotoxicity, which occurs in an estimated 9/100,000 prescriptions. Clonidine is an alpha-2 adrenergic agonist that has been shown to be effective in reducing spasticity (especially extensor spasms and clonus) when used as an adjunct to baclofen.

Reference:

Question(s) 341:
Pharmacology/Chemistry
Discussion:
Cyclosporine toxicity may produce a wide array of neurotoxic effects, including cortical blindness, tremor, mental status changes, and many others. Seizures are another significant complication of cyclosporine therapy, typically occurring in the setting of an encephalopathy. Hypomagnesemia may contribute to the development of seizures in patients receiving cyclosporine. This is the result of inappropriate renal wasting of magnesium due to a direct cyclosporine-induced toxic effect on the renal tubule.

Reference:

Question(s) 342 - 343:
Pharmacology/Chemistry
Discussion:
Because elderly individuals - especially those who are already displaying some cognitive impairment - are particularly prone to anticholinergic toxicity, nortriptyline would be a better antidepressant choice than amitriptyline because nortriptyline's anticholinergic properties are much less. Amoxapine may actually exacerbate parkinsonism, while non-selective MAO inhibitors like tranilcypramine are contraindicated in persons on levodopa. There have been reports of the combination of selegiline and fluoxetine producing the "serotonin syndrome", consisting of hypertension, diaphoresis, tremulousness and confusion. Similarly, the combination of selegiline and meperidine may produce a "central excitatory syndrome" with similar symptoms. Macrolide antibiotics, such as erythromycin, can dramatically elevate plasma bromocriptine levels and, thus, could precipitate dyskinesia. It is possible that the same phenomenon could occur with other ergot derivatives. Amantadine can produce livedo reticularis, in which the skin (typically the legs) takes on a purplish mottled appearance. This is not harmful, but may be
frightening to patients.

Reference:

Question(s) 344 - 345:
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) 346:
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:
The 12-year-old boy has a characteristic history for juvenile myoclonic epilepsy with early morning myoclonic and generalized tonic clonic seizures, beginning in the second decade of life. Absence seizures may also occur. The EEG reveals generalized, fast (4 to 6 Hz) spike-and-wave complexes. Valproic acid is the treatment of choice and recurrence of seizures is likely to occur if treatment is stopped.

Reference:

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:

Aicardi-Goutieres syndrome is a progressive encephalopathy characterized by acquired microcephaly, with white matter changes, basal ganglia calcifications and spasticity or dystonia, associated with CSF lymphocytosis and elevated interferon alpha levels. Experimental data suggest that interferon has a protective role against neurotropic viruses, but is itself toxic in elevated concentrations. Familial cases consistent with autosomal recessive transmission have been reported. Congenital CMV infection is associated with positive TORCH serology, classically periventricular calcifications and a static encephalopathy. Pantothenate-kinase associated neurological disorder causes a progressive dystonia, variably associated with acanthocytosis, retinal degeneration or hypoprebetalipoproteinemia (HARP variant), and the ‘eye-of-the-tiger’ sign on MRI. Pseudohypoparathyroidism causes symmetric basal ganglia calcification without CSF changes, short stature and bony anomalies. Tuberous sclerosis complex (TSC) produces calcification of subependymal nodules, but not the basal ganglia.

Reference:

Question(s) 359 - 361: Clinical Pediatrics
Discussion:
This child has tuberous sclerosis complex, an autosomal dominant disorder associated with mutations in the tuberin and merlin genes. Intellectual outcome is best correlated with seizure control. Vigabatrin is a GABA transaminase inhibitor that potentiates the effect of endogenous GABA. It is not approved by the FDA for use in the
US, owing to a high frequency of visual loss associated with its use. European studies suggest that it is the drug of choice for infantile spasms in tuberous sclerosis complex.

Reference:

Question(s) 358 - 360: Clinical Pediatrics
Discussion:
The clinical history is classical for childhood ataxia with central hypomyelination (CACH), also known as vanishing white matter disease. Mutations in any of five genes coding for translation initiation factor 2B (eIF2B) have been associated with this syndrome. The alternatives are associated with the following diseases: Ataxin – SCA 1; ATM – ataxia telangiectasia; Frataxin – Friedreich ataxia; MLC 1 – Megencephalic leucoencephalopathy with subcortical cysts.

Reference:

Question(s) 361 - 362: Clinical Pediatrics
Discussion:
Asperger syndrome is classified with the autistic spectrum disorders (pervasive developmental disorders). It shares the features of autistic disorder, except that language is spared. Children with Asperger syndrome are often referred to as 'little professors' owing to their extensive knowledge of very restricted fields of interest.

Reference:


Question(s) 362 - 363: Pathology
Discussion:
The clinical features described are best attributed to periventricular leukomalacia, a white matter injury that may be due to ischemia and reperfusion or cytotoxic injury during infections. The greatest period of vulnerability for the premature infant is during the 24th-32nd week of gestation. The most common location for the sharply demarcated areas of white matter necrosis are anterior to the frontal horn, lateral corners of the ventricles at the level of the foramen of Monro, and the lateral regions of the trigone and occipital horn. White matter involvement in these areas can lead to spasticity involving the lower extremities more than upper. The injury is usually static and nonprogressive.

Reference:

Question(s) 364: Pharmacology/Chemistry
Discussion:
The findings are most consistent with propofol infusion syndrome. Propofol impairs mitochondrial fatty acid oxidation and oxidative phosphorylation. Propofol raises levels of malonylcarnitine that inhibits CPT1, blocking long chain fatty acid transport into mitochondria. Complex II function is secondarily inhibited. Children have limited carbohydrate stores, and require larger doses of propofol for sedation, thus placing them at higher risk of this syndrome than adults. Inadequate caloric intake before or during the infusion further increases the risk, by leading to inability to meet metabolic demands and suppress fat metabolism.
Hemofiltration has been shown to correct the metabolic abnormalities in propofol infusion syndrome. Carbohydrate intakes of 6-8 mg/kg/min during propofol infusion might prevent it.

Reference:

Question(s) 365: Pharmacology/Chemistry
Discussion:
Benzodiazepines are regarded as the treatment of choice for alcohol withdrawal on the basis of several controlled trials, reducing the risk of all symptoms including delirium tremens and seizures. The other agents listed have shown varying degrees of effectiveness in relieving symptoms of withdrawal, as primary or adjunctive agents, but none are as effective as benzodiazepines.

Reference:

Question(s) 366: Pharmacology/Chemistry
Discussion:
Opioid withdrawal resembles severe influenza, with the additional features of mydriasis, lacrimation, rhinorrhea, piloerection, yawning, sneezing, anorexia nausea, vomiting and diarrhea. Delirium tremens and seizures do not occur. Classic withdrawal techniques utilize long acting oral agents such as methadone. Studies of aggressive (rapid) withdrawal regimens used over five days have found that the least severe symptoms and shortest duration of withdrawal was achieved with a combination of naltrexone, clonidine and buprenorphine.

Reference:

Question(s) 367 - 368: Pharmacology/Chemistry
Discussion:
The most appropriate way to counsel patients regarding changes in their therapy would be to include the referring physician in the decision-making process.

Reference:

Question(s) 369: Clinical Adult
Discussion:
Use of intravenous TPA is a proven treatment for acute ischemic stroke in the appropriate patient candidate if given within the three hour time window from symptom onset. Verbal informed consent from the patient or next of kin if the patient can not consent is necessary to administer the drug. Informed consent for the use of TPA includes discussion of the risks and benefits of therapy including the risk of brain hemorrhage or death.

Reference:

Question(s) 370 - 371: Pharmacology/Chemistry
Discussion:
Autosomal dominant hypokalemic periodic paralysis typically presents in the first three decades with episodes of weakness occurring following rest after exercise,
particularly if the subject is exposed to high carbohydrate meals, emotional stress or cold. The respiratory and cardiac muscles are almost always spared, and attacks last for hours. Symptoms often resolve in later life, although patients may be left with residual weakness. Most patients have mutations in the skeletal muscle calcium channel gene, although mutations may be found in sodium and potassium channels in the remainder. Secondary hypokalemic paralysis occurs in older patients with chronic medical problems predisposing them to hypokalemia. Preventive treatment with acetazolamide is usually effective in abolishing or attenuating episodes, and oral potassium supplements may be helpful during acute attacks. Some patients require the use of potassium sparing diuretics such as spironolactone or triamterene. Nephrolithiasis is a complication of long-term therapy with acetazolamide, and should be screened for with annual ultrasound examinations.

Reference:

Question(s) 372 - 374: Clinical Pediatrics Discussion:
Benign focal epilepsy of childhood with centrotemporal sharp waves presents from 2 to 13 years of age. About 20% of patients only have one seizure and in 2/3 of the cases seizures are infrequent. Children with this disorder are neurologically normal and the majority of patients outgrow the disorder. Carbemazepine is the treatment of choice if treatment is necessary.

Reference:
Wyllie E. The treatment of epilepsy, principles and practice. 3rd ed. (city?): Lippincott Williams & Wilkins, 2001

Question(s) 375 - 376: Pharmacology/Chemistry Discussion:
Wilson’s disease is an autosomal recessive trait associated with mutations of the copper-transporting ATPase gene on chromosome 13q14.3. Clinical features are neurological (40%), hepatic (40%), and psychiatric (15%). Patients present in the second or third decade of life. Neurologic features include an akinetic-rigid syndrome resembling parkinsonism, generalized dystonia, or postural tremor with ataxia. Dysarthria and clumsiness of the hands are common presenting features. Kayser-Fleischer rings are present in virtually all patients with neurological features. Symptoms of liver disease include a history of prior or concurrent liver disease. The pathologic abnormalities are primarily in the basal ganglia, with cavitory necrosis of the putamen and caudate, in addition to cortical atrophy. The liver develops a nodular cirrhosis. In most cases Wilson’s disease can be diagnosed by measurement of the serum concentration of the copper protein, ceruloplasmin, which is often low (<20mg/dl). Serum total copper is low in many patients and urinary copper excretion is always raised. Definitive investigation is a liver biopsy with measurement of copper concentration. D-penicillamine with pyridoxine is the gold standard of treatment. Alternative therapies include trientene, zinc, tetrathiomolybdate, dimercapol, and liver transplantation. Symptomatic treatment with antiparkinsonism drugs may be of benefit.

Reference:
tics in this age group, but the latter are not
contraindicated.

Reference:

Question(s) 380 - 381: Clinical Adult
Discussion:
The single best predictor of survival in
patients with advanced cancer is
performance status as a measure of
functional ability. Once patients with solid
tumors spend >50% of their time in bed, the
median survival is approximately 2-3
months. Home hospice care is an important
benefit under Medicare regulations that
provides patient and care taker support
during the dying process. Under federal law,
Medicare-certified home hospice agencies
may not use the DNR status as a criteria for
admission. Establishing a physician of
record, expected prognosis of less than six
month survival and use of a palliative
approach to care are required.

Reference:
1. Lassauniere JM, Vinant P. Prognostic
factors, survival and advanced cancer.
2. Beresford L. The hospice handbook-a
complete guide. Boston: Little Brown and

Question(s) 382 - 384: Behavioral
Discussion:
Catatonia is a syndrome manifested by a
number of motor and neuro behavioral
features. It may have a “retarded-stuporous”
form or an “excited-delirious” form. It may be
seen in over 10% of inpatient psychiatric
patients. Catatonia is more prevalent in
mood disorders than in schizophrenia. The
most common mood disorder in which it is
seen is bipolar. Catatasy, waxy flexibility,
echophenomena, and negativism including
mutism are common. Many neurological and
systemic illnesses may also present as
catatonia. Treatments include
benzodiazepines, barbiturates, and ECT.
Dopamine antagonists as well as
baclofen may worsen the condition.

Question(s) 385: Pharmacology/Chemistry
Discussion:
Modafinil is a non-stimulant wake-promoting
drug which has shown benefit in treating
daytime hypersomnia associated with
Parkinson's disease, myotonic dystrophy,
and fatigue associated with multiple
sclerosis. Presently, it is FDA approved only
for the treatment of hypersomnia of
narcolepsy. The site of action of modafinil is
not known with certainty. The most likely
sites of action are neurons of the anterior
hypothalamus, tuberomammillary nucleus
(TMN) and orexin (hypocretin) neurons,
involved in regulating the sleep-wake cycle.

Reference:
Randomized trial for modafinil for treating
subjective daytime sleepiness in patients
with Parkinson's disease. Movement
Disorders 2003;18:287-293.
2. Scammell TE, Estabrooke IV, McCarthy
MT, et al. Hypothalamic arousal regions are
activated during modafinil-induced
wakefulness. J Neurosci 2000;20:8620-
8628.

Question(s) 386: Pathology
Discussion:
Congenital toxoplasmosis produces necrosis
and calcification in periventricular and
cortical areas of premature brains.

Reference:
Graham DI, Lantos PL. Greenfield's
neuropathology. 7th ed. New York: Arnold

Question(s) 387: Neuroimaging
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) 388: Neuroimaging
Discussion:
There is evidence of right parietal watershed infarction on the CT scan. The characteristic findings include loss of the markings of the gray-white interface, subtle reduction in sulcal markings, and the typical distribution between the middle cerebral and posterior cerebral artery territories. The other lesion locations do not fit with these findings. The midline structure is a calcified pineal gland, and not a colloid cyst, which would be hypodense on the CT scan. Cerebral contusions are hyperdense on CT due to the presence of blood. Thus, watershed infarction is the best answer.

Reference:

Question(s) 389: Neuroimaging
Discussion:
Of the given diagnoses, the most likely is a late onset neurodegeneration with brain iron accumulation, type 1 (NBIA 1), or Hallervorden-Spatz syndrome. In the globus pallidus can be observed the “eye-of-the-tiger” sign, described originally in this disorder and later found also in cortico-basal-ganglionic degeneration and in progressive supranuclear palsy. The other diagnoses do not fit the imaging findings.

Reference:

Question(s) 390: Neuroimaging
Discussion:
Dissection and total occlusion of the left internal carotid are incorrect because there is no evidence for dissection. Emergency surgery is not required and it is clear that there is a large ulcerated plaque, not a dissection. The ulcerated plaque could have resulted from the accident, but it is far more likely that it happened spontaneously. If plaque material from within a crater this large had broken off and embolized during the accident, the patient would have had serious neurological morbidity. Therefore, the correct answer is that the accident did not cause the findings.

Reference:

Question(s) 391: Anatomy
Discussion:
The structure is the substantia nigra which projects to and receives fibers predominantly from the caudate and putamen.
Reference:


Question(s) 392: Neuroimaging
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) 393: Neuroimaging
Discussion:
Sagittal T1 weighted image shows a hypointense flow void that is linear, extending from the pial surface down into the white matter of the parietal lobe. Radiating hypointense tributaries are present within the white matter. The findings are consistent with that of an enlarged vascular structure such as a vein. Axial scan done for T1 blood flow effects shows the tributaries to be of high signal intensity within the white matter. This pattern of white matter tributaries to a trans-cerebral draining vascular structure is characteristic of a venous angioma. A tumor would not appear as a tubular hypointense flow void, an infarct would not have a long linear pattern extending from cortex deep into white matter in a tubular fashion, while Sturge Weber Disease is associated with hemiatrophy, which is not present in this case. Schizencephaly is an ependymal pial seam associated with dilatation of the ventricle focally, and is lined by heterotopic gray matter. The seam and the heterotopic gray matter are not present, and the ventricle is normal. An acute hemorrhage would show mass effect.

Reference:

Question(s) 394: Neuroimaging
Discussion:
The small, mostly intracanalicular lesion on the right, enhances with gadolinium and has the typical appearance of an acoustic neurinoma. There is no dural tail, typical of meningiomas. Cholesteatomas have different signal characteristics and do not have a uniform enhancement pattern. Chordomas appear in the clivus, not at this location.

Reference:

Question(s) 395: Neuroimaging
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) 396: Neuroimaging
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) 397: Pathology
Discussion:
The graphic shows an acute subdural hematoma. Subdural hematomas result from disruption of bridging veins and are seen more commonly in older persons with cortical atrophy with resultant tension on these veins. The inciting trauma often is a fall, but may not be recollected.

Reference:

Question(s) 398: Neuroimaging
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) 399: Neuroimaging
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 left image, there is a rounded density consisting of a middle cerebral artery aneurysm.

Reference:

Question(s) 400: Neuroimaging
Discussion:
Both the clinical and the imaging pictures suggest embolization of the pulmonary venous circulation by microspheres, which occluded some of the thin arterial perforators in the brain. Metastases, abscesses and hemorrhages have a different MRI appearance.

Reference:

Question(s) 401: Pathology
Discussion:
The photograph shows syringomyelia. 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.

Question(s) 402: Neuroimaging
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 and would not cause the irregular displacement of the top of the thalami. Multiple sclerosis would not cause this amount of mass effect.


Question(s) 403: 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, such as 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.


Question(s) 404: Pathology
Discussion: This brain shows multiple cavities involving the deeper cortical layers and underlying white matter, a condition known as multicystic encephalopathy or multicystic encephalomalacia. The lesions are most pronounced in the distribution of the anterior and middle cerebral arteries. They are the result of circulatory disturbances during the latter half of pregnancy or the neonatal period.


Question(s) 405: Physiology
Discussion: Progressive weakness and dry mouth with marked increase in compound muscle action potential amplitude after brief exercise is consistent with Lambert Eaton myasthenic syndrome. Calcium channel antibodies block calcium uptake presynaptically and hinder acetylcholine release. Although a similar electrophysiologic finding can be seen with botulism, the clinical history is not consistent with this.


Question(s) 406: Neuroimaging
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) 407: Neuroimaging
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) 408: Neuroimaging
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) 409: Pathology
Discussion:
The kodachrome shows epithelial cords or strings of vacuolated "physaliphorous" cells within a mucoid matrix. Chordoma is the most common neural crest-derived tumor of the sacrum and almost always arises within bone. Chordoma is derived from notochordal remnants and occurs predominantly in the sacrum or the skull base near the clivus. Myxopapillary ependymomas arise in the filum terminale and neurofibromas and schwannomas arise in the nerve roots. While these types of tumors can also attain sufficient size to erode the sacrum, it is uncommon. Teratomas can also erode the sacrum, but do so less frequently than chordomas. Ewing's sarcoma is a tumor type that is rare in this location.

Reference:

Question(s) 410: Neuroimaging
Discussion:
Cortical dysplasia can be seen at the posterior extent of the left sylvian fissure, as a thick cortical ribbon. Near the lesion is the angular branch of the middle cerebral artery, of a normal appearance. There is no evidence of an AVM. Cerebral infarction would cause cortical thinning, not thickening. The nodular appearance of an oligodendroglioma and the cystic appearance of porencephaly are not present in this image.

Reference:

Question(s) 411: Pathology
Discussion:
Agyria and lissencephaly are terms used to describe relatively smooth brains with a reduced number of gyri. Agyria is thought to result from an arrest of neuronal migration that leads to an abnormally thick cortex with improper layering. Neuronal heterotopia can be seen in the white matter as well.

Reference:

Question(s) 412: Neuroimaging
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) 413: Pathology
Discussion:
The graphic illustrated a tuber, characterized by a broad, pale gyrus in the frontal lobe with loss of the gray-white matter delineation. Cortical tubers are one of the characteristics of tuberous sclerosis.

Reference:

Question(s) 414: Pathology
Discussion:
The photograph shows a combination of hydrocephalus and meningomyelocele in this infant, features of Arnold-Chiari malformation (Chiari type II malformation).

Reference:

Question(s) 415: 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) 416: Neuroimaging
Discussion:
There is marked enlargement of the third and lateral ventricles, with a normal sized fourth ventricle. Note also the peculiar configuration of the quadrigeminal plate, which appears displaced and compressed.
superiorly, but normally inferiorly, reflecting dilation of the proximal portion of the aqueduct only. The axial T2 image through the midbrain also fails to demonstrate the normal flow void seen at the level of the aqueduct that originated from high velocity of CSF traveling through the patent aqueduct. These findings are consistent with a diagnosis of obstructive hydrocephalus, with obstruction at the level of the aqueduct (excluding the choices of atrophy or communicating hydrocephalus). In this case, there is no evidence of a mass in the quadrigeminal plate, and obstruction is due to stenosis.

Reference:

**Question(s) 419**: Neuroimaging

**Discussion:**
The line points to a lipoma located immediately superior a myelomeningocele. CSF is dark on T1 and hyperintense on T2 images. The lipoma is identified by being bright on both T1 and T2 images, in contrast to CSF which is dark on T1. Although hemorrhage and certain types of metastatic tumors may be bright on T1, the circular shape and location are most consistent with lipoma (fat).

Reference:

**Question(s) 420**: Pathology

**Discussion:**
The coronal brain section shows a lesion with a necrotic center and a capsule. The surrounding tissue is edematous. This is grossly consistent with a bacterial brain abscess. This abscess has ruptured into the lateral ventricle.

Reference:

**Question(s) 421**: Pathology

**Discussion:**
This mid-sagittal section of cerebellum shows atrophy of the superior vermis. This is characteristic of alcoholic cerebellar degeneration, but can also be seen with aging.

Reference:
Question(s) 422: Pathology
Discussion:
This patient had acute bacterial endocarditis with destruction of one leaf of his aortic valve and a large vegetation on other leaflets. These vegetations contained neutrophils and fibrin, as seen on the low power photomicrograph in B. His brain contained infarctions, hemorrhages, and early abscess formation, features seen in patients with acute bacterial endocarditis, particularly in cases due to virulent organisms such as Staphlococcus sp.

Reference:

Question(s) 423: Neuroimaging
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) 424: Neuroimaging
Discussion:
An intra-axial pontine lesion is noted that heterogeneously enhances. Moderate mass effect is present. This most likely is a glioma.

Reference:

Question(s) 425: Neuroimaging
Discussion:
The image shows heterotopic grey matter in the left frontal region. Deformity of the left frontal horn is seen with ex-vacuo hydrocephalus. No mass effect is seen to suggest tumor. Rather, the lesion is destructive. Heterotopic grey matter refers to cortical neurons in abnormal location. Heterotopia is caused by migrational arrest of the affected neuroblasts. The cause of the incomplete migration is unknown. Three kinds of heterotopic grey matter have been described: 1) nodular (periventricular), 2) laminar (in deep white matter), like this patient, 3) band heterotopia (double cortex).

Reference:

Question(s) 426: Pathology
Discussion:
The microscopic appearance is that of a capillary-rich neoplasm with abundant foamy cells containing lipid. This is a hemangioblastoma. 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. Pilocytic astrocytomas, also commonly found in the cerebellum, are composed of astrocytes with elongate, bipolar, eosinophilic cell processes; these tumors often show Rosenthal fibers. Medulloblastomas are malignant small blue cell tumors of neuronal origin that manifest little visible cytoplasm. Ependymomas are glial tumors which show formation of ependymal canals, or more commonly, perivascular pseudorosettes.

Reference:
Question(s) 427: Neuroimaging
Discussion:
The correct answer is congenital abnormality. MR images demonstrate dilatation of the cord around a central cystic cavity, consistent with syringohydromyelia. Note also on the sagittal images that the inferior portion of the cerebellum has herniated down into the upper cervical canal, diagnostic of a Chiari malformation. Hydromyelia is commonly seen in association with both Chiari type I and Chiari type II malformation.

Reference:

Question(s) 428: Neuroimaging
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) 429: Neuroimaging
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) 430: Neuroimaging
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) 431: Neuroimaging
Discussion:
Meningeal carcinomatosis is suggested by the ring of enhancement around the brain stem sections and by the enhancement of the meninges in the cerebellar sulci. Hemorrhages and superficial siderosis cause a different signal intensity. Tuberculous meningitis could produce similar imaging findings, but the history suggests pial involvement by malignant melanoma.

Reference:

Question(s) 432: Neuroimaging
Discussion:
The most likely possibility is an aneurysm because of the lesion's negligible signal intensity due to fast flow.

Reference:

Question(s) 433: Pathology
Discussion:
The photograph shows a massive basal ganglionic hemorrhage with rupture into the ventricular system. The location of this hematoma is common for hypertensive intracerebral hemorrhage. Hypertension is a major risk factor for this type of hemorrhage, and hypertension is associated with pathologic and electrocardiographic evidence of left ventricular hypertrophy. Prostate cancer only rarely metastasizes to brain parenchyma and is usually not hemorrhagic. Visceral cysts are not relevant to intracerebral hemorrhage, but are frequently seen in von Hippel-Lindau disease with associated CNS hemangioblastoma.

Reference:

Question(s) 434: Pathology
Discussion:
The photograph shows Wallerian degeneration, with shrinkage of one medullary pyramid and atrophy of the ipsilateral cerebral peduncle. The Wallerian degeneration is secondary to destruction of the corticospinal tract above this level, such as an infarction in the posterior limb of the internal capsule.

Reference:

Question(s) 435: Neuroimaging
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) 436: Neuroimaging
Discussion:
CT scans demonstrate marked calcification of the basal ganglia and in the cerebellum, confined on MRI by the presence of hypointensity in those same areas on the T2-weighted image. All of the possibilities listed can cause calcifications. Leigh's disease causes necrosis of the basal
ganglia, particularly putamen, and results in low density of CT, and hyperintensity on T2-weighted images. With amyloid angiopathy, hyperparathyroidism, and Wilson's disease, there is a known disorder associated with the calcifications. In conditions where no such metabolic derangement can be found to explain the calcifications, the diagnosis is idiopathic calcification of the basal ganglia, or Fahr's disease.

Reference:

Question(s) 437: Neuroimaging
Discussion:
Although abscesses, gliomas, lymphomas and multiple sclerosis can all present with an enhancement ring similar to the one shown, abscesses, lymphomas and multiple sclerosis evolve quicker than this lesion. It could be a glioma, but the proximity to the lesion and the presence of other lesions in the path of the radiation portals strongly suggests radiation necrosis.

Reference:

Question(s) 438: Neuroimaging
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) 439: Neuroimaging
Discussion:
There is gadolinium enhancement of meningeal structures, suggesting hypotension, carcinomatosis or lymphoma. The orthostatic character of the headache and the absence of other findings favor intracranial hypotension, also favored by the imaging finding of descended tonsils present in the sagital image.

Reference:

Question(s) 440: Neuroimaging
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) 441: Neuroimaging
Discussion:
Sagittal T1-weighted image shows a Chiari II malformation with deformity of the tectum of the mesencephalon, caudalization of the cerebellar vermis into the cervical spinal canal, and a deformity of the medial aspect of the cerebral hemisphere, with an absent posterior corpus callosum. The cerebral aqueduct is not visualized and the ventricles do not appear enlarged, suggesting there is aqueductal stenosis and that a functioning shunt should be present as was the case. There is a high degree of association between the cerebral changes of Chiari II malformation and the presence of a lumbar myelomeningocele, thus the lumbar spine should be and was dysraphic. Patients with
Chiari II malformations do not have to be and are frequently not mentally retarded. There is no evidence of contusion, subdural hematoma, or other forms of trauma on the section shown.

Reference:

Question(s) 442: Neuroimaging
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) 444: Neuroimaging
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 nonspecific, 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) 445: Physiology
Discussion:
An EEG showing triphasic waves is consistent with a metabolic encephalopathy, most commonly hepatic encephalopathy.

Reference:

Question(s) 446: Neuroimaging
Discussion:
The distribution of the changes,
predominantly in the superior frontal gyrus, and the stump of the occluded anterior cerebral artery, visible on MR angiography, indicate that the cause of the infarct is an anterior cerebral artery occlusion.

Reference:

Question(s) 447: Neuroimaging
Discussion:
The location of this lesion is clearly too cephalad to be a giant aneurysm, and it falls outside of the corpus callosum. There is no surrounding edema, so metastatic disease is unlikely. Lipomas do not enhance. The homogenous enhancement, isointensity on T1 images, and location are characteristic of a falcine meningioma.

Reference:

Question(s) 448: Pathology
Discussion:
The brain shows severe atherosclerosis of intracranial vessels as well as bilateral remote anterior cerebral artery infarctions. The ventricles are slightly enlarged due to hydrocephalus ex vacuo. This patient actually had a single, unpaired anterior cerebral artery, accounting for the bilateral nature of the infarctions.

Reference:

Question(s) 451: Neuroimaging
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 (viewed with a fluorescent microscope) is diagnostic of cerebral amyloid angiopathy. Neither primary or secondary systemic amyloidoses cause amyloid deposits within cerebral blood vessels. CADASIL deposits are would not stain with thioflavin S or Congo red since they are not amyloid. Chronic hypertension causes thickened walls of small blood vessels due to hyalinization and lipohyalinosis; no amyloid is deposited.

Reference:
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) angiomatosis 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) 452: 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 would not necessarily be confined to the vascular distribution territory of a branch of the middle cerebral artery as this lesion is. Both an abscess and metastatic carcinoma would be accompanied by brain swelling rather than shrinkage; note the lack of mass effect on the ventricular system.

Reference:

Question(s) 453: Neuroimaging
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) 454: Neuroimaging
Discussion:
There is a calcified soft tissue mass in the pineal region, most consistent with a pineoblastoma. Mass effect on the aqueduct results in hydrocephalus. Because of the high incidence of subarachnoid seeding in these tumors, contrast enhanced imaging of the entire spine is indicated to assess for "drop metastases." Chordoma usually arises from clivus, which is intact. No pituitary calcification is present to suggest craniopharyngioma.

Reference:

Question(s) 455: Neuroimaging
Discussion:
Developmental venous anomaly (DVA) (venous angioma) are commonly identified in the work-up of patients with neurological symptoms. Headache is the most common symptom. Seizures are also commonly associated. There is no real evidence, however, that either headaches or seizures are caused by the DVA and in most instances the DVA is incidental. It is very important to understand that the DVA represents the venous drainage of the brain tissue in which it is situated. Removal of the DVA thus is associated with venous infarction.

Reference:
Rigamonti D, Hsu FPK, Huhn S. Angiographically occult vascular malformations in neurovascular surgery. In: Carter PL, Spetzler RF, editors. (need
Question(s) 456: Physiology
Discussion:
Subacute sclerosing panencephalitis is associated with periodic slow wave complexes in the EEG that recur every 4-15 seconds.

Reference:

Question(s) 457: Neuroimaging
Discussion:
Although it is possible for this lesion to be neoplastic or infectious, the enhancement ring in these disorders involves both the gray and the white matter, and most often is a complete, closed ring. By contrast, the enhancement ring of “giant plaques,” like the one shown here, is often open and affects only the white matter at the border of the lesion.

Reference:

Question(s) 458: Pathology
Discussion:
The photograph shows abundant exudate, especially along vessels, in the subarachnoid space on the dorsal surface of the brain. This is most consistent with purulent leptomeningitis. Hemophilus influenzae rarely causes meningitis in the adult, and pneumococcal meningitis is more common than meningococcal meningitis in the elderly patient. With this history of alcoholism and asplenism, the most likely diagnosis is pneumococcal meningitis.

Reference:

Question(s) 459: Pathology
Discussion:
Lobar atrophy of the frontal and temporal lobes is typical of Pick's disease. Microscopically, intracytoplasmic argyrophilic neuronal inclusions and gliosis are observed in Pick body Pick's disease.

Reference:

Question(s) 460: Neuroimaging
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) 461: Pathology
Discussion:
The biopsy of this AIDS patient disclosed the presence of toxoplasma gondii in the bradyzoite form.

Reference:
Question(s) 462: Pathology

Discussion:
The muscle biopsy specimen shows a necrotizing vasculitis with fibrinoid necrosis of the vessel walls, not amyloid angiopathy. Vasculitis may be seen in amphetamine-induced vasculitis, polyarteritis nodosa, rheumatoid vasculitis, and Wegener's granulomatosis, but not in polymyalgia rheumatica which is associated with temporal arteritis and usually type II fiber atrophy in the muscle biopsy, not vasculitis. Takayasu's arteritis affects large blood vessels such as aortic arch, not small intramuscular vessels. ALS causes no inflammation of intramuscular blood vessels, but does show neurogenic atrophy.

Reference:

Question(s) 463: Physiology

Discussion:
The EEG sample of an eight-month-old infant shows sleep spindles.

Reference:

Question(s) 464: Physiology

Discussion:
A 3 Hz spike-and-wave pattern is seen in absence seizures. Some patients also have generalized tonic clonic seizures which may persist without treatment but tend to be easily controlled. Both seizure types are controlled by valproate. Carbamazepine may exacerbate absence seizures. Automatisms may occur during absence seizures, particularly when they are prolonged.

Reference:

Question(s) 465: Neuroimaging

Discussion:
The MRI reveals far too much white matter disease for Alzheimer's disease. Metachromatic leukodystrophy typically produces by this age more diffuse and confluent areas of demyelination than those shown. Progressive multifocal leukoencephalopathy is less common, and large regions (rather than small plaques) of white matter signal change are expected. Given the diffuse white matter lesions that are characteristic of MS plaques, her age, and gender, MS is the most likely diagnosis. Vasculitis typically involves the gray/white matter junction.

Reference:

Question(s) 466: 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) 467: Pathology

Discussion:
Fungal infections with Coccidioides occur in regions of semi-arid climate. In the United States, the Southwest and California are the most common locations for these infections. Mature fungi in tissue are endospores, such as the one pictured, which are round and have a refractile wall.

Reference:
Graham DI, Lantos PL. Greenfield's

**Question(s) 468: 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) 469: Neuroimaging**

**Discussion:**
The high subcortical signal in T2, affecting multiple gyri, suggests increased vascular permeability, as it happens in all the conditions listed as possible answers. However, only eclampsia fails to have other findings and causes white matter changes with the topography and shape present in the images. Multiple sclerosis tends to cause sharper lesions, with a periventricular distribution. In abscesses, there are round, cystic collections. There is no evidence of nodular deposits of choriocarcinoma. Emboli in these locations would be accompanied by cortical necrosis, not evident in this image.

**Reference:**

**Question(s) 470: Neuroimaging**

**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) 471: Neuroimaging**

**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) 472: Neuroimaging**

**Discussion:**
This is a typical location for a colloid cyst. This usually causes low signal in the T2-weighted image, however, the MRI signal can be variable.

**Reference:**

**Question(s) 473: Neuroimaging**

**Discussion:**
The large left cerebrospinal fluid collection represents typical arachnoid cyst: the medial part of the Sylvian fissure is "box" shaped.
The larger hemicranium proofs that the underlying pathology is long standing. Chronic subdural hematoma and subdural effusion would have flattening effect upon the sulci. Hemimegalencephaly is a parenchymal malformation, although there is enlargement of the hemisphere.

Reference:

Question(s) 474: Neuroimaging
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) 475 - 479: Neuroimaging
Discussion:
This is a straightforward question about normal vascular anatomy. The innominate artery joins the right subclavian to the aorta. The left subclavian emerges directly from the aorta. The left vertebral artery typically originates from the left subclavian. The right posterior cerebral artery originates from the basilar tip. The external carotid is recognized by the presence of branches prior to entering the cranium.

Reference:

Question(s) 480: Anatomy
Discussion:
The arrow points to the neurohypophysis (pituitary stalk) which is a direct extension of the neuraxis. This structure contains axons arising from the supraoptic and paraventricular nuclei. These axons carry oxytocin and vasopressin (antidiuretic hormone ADH); therefore, damage to the infundibulum will result in diabetes insipidus.

Reference: