Question(s) 1: Anatomy
Discussion:
The Martin-Gruber anastomosis occurs in 15% to 30% of the population. It consists of a communicating branch from the median nerve to the ulnar nerve in the forearm to supply the first dorsal interosseous, adductor pollicis, and abductor digiti minimi.

Reference:

Question(s) 2: Anatomy
Discussion:
The prosencephalon divides into the diencephalon and the telencephalon. The diencephalon gives rise to the thalamus and hypothalamus. The telencephalon gives rise to the striatum and cerebral cortex.

Reference:

Question(s) 3: Clinical Pediatrics
Discussion:
Group B streptococcus causes almost 50% of cases of neonatal bacterial meningitis and is the most common organism. E. coli and L. monocytogenes are the second and third most common organisms, respectively.

Reference:

Question(s) 4: Clinical Adult
Discussion:
The first treatment for tardive phenomenon is elimination of the precipitating medication.

Reference:

Question(s) 5: Clinical Adult
Discussion:
Limb, cranial nerve and autonomic dysfunction commonly accompany Guillain-Barré syndrome, but sensory loss is rare. In those who may not have received vaccination, the diagnosis of diphtheria should be considered. This is especially true when symptoms are preceded by pharyngeal exudation. Diphtheria typically begins with a pharyngeal infection and exudate, sometimes followed by local palatal neuropathy. This may be followed by paralysis of pupillary accommodation. About 10% of patients develop a diffuse motor and sensory polyneuropathy about 8-12 weeks after the pharyngeal infection. Cerebrospinal fluid protein is usually elevated.

Reference:

Question(s) 6: Clinical Pediatrics
Discussion:
Ataxia-telangiectasia is an autosomal recessive disorder characterized by ataxia, oculocutaneous telangiectasias, and immunoincompetency. Choreoathetoid movements are seen even at an early age.

Reference:

Question(s) 7: Physiology
Discussion:
Periodic lateralized epileptiform discharges (PLEDs) are seen after an acute or subacute cerebral insult such as a stroke and are often associated with focal seizures.

Reference:
Niedermeyer E, Lopes da Silva F. 

Question(s) 8: Pharmacology/Chemistry
Discussion: 
Dopamine agonists such as ropinirole and pramipexole may produce sudden irresistible attacks of sleep while driving. Neurologists who prescribe these drugs and patients who take them should be aware of this possible side effect. 

Reference: 

Question(s) 9: Pharmacology/Chemistry
Discussion: 
Tolcapone and entacapone increase the duration of clinical response with levodopa. These drugs act by inhibiting COMT enzyme in the periphery and decreasing conversion of levodopa to 3-O-methyl dopa. Tolcapone is associated with a small risk of liver damage, and its use requires liver enzyme monitoring. Both may result in severe diarrhea and cause discoloration of urine.

Reference: 

Question(s) 10: Clinical Adult
Discussion: 
The primary cancers most responsible for spinal cord metastasis are lung (49%), breast (15%), lymphoma (9%), colorectal (7%), renal and head and neck (6% each).

Reference: 

Question(s) 11: Anatomy
Discussion: 
A posterior communicating artery aneurysm may compress the third cranial nerve leading to ipsilateral pupillary dilatation and ophthalmoparesis.

Reference: 

Question(s) 12: Pharmacology/Chemistry
Discussion: 
Drugs that can raise carbamazepine levels include isoniazid, erythromycin, cimetidine, calcium channel blockers (such as verapamil), and propoxyphene. Carbamazepine levels are lowered by phenobarbital, phenytoin, and primidone. Warfarin, chlorpromazine, digoxin, and gabapentin have no significant effect on carbamazepine levels.

Reference: 
Holland KD. Efficacy, pharmacology, and adverse effects of antiepileptic drugs. Neurology Clin 2001;19:313-345

Question(s) 13: Anatomy
Discussion: 
Unlike the relay and association nuclei of the thalamus (dorsal thalamus), the reticular nucleus (ventral thalamus) does not project to the cerebral cortex. It receives input from the cortex and projects to the other thalamic nuclei and is critical for thalamocortical synchronization, particularly generation of sleep spindles.

Reference: 
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams &
Question(s) 14: Physiology
Discussion:
Activity in the 8 Hz range can be seen in comatose patients with pontine infarctions, ie, an alpha coma pattern.

Reference:

Question(s) 15: Clinical Adult
Discussion:
Anterior ischemic optic neuropathy (AION) usually causes severe visual loss and optic disc swelling. Pain upon eye movement is not a clinical feature of AION.

Reference:

Question(s) 16: Pharmacology/Chemistry
Discussion:
The selective serotonin transporter (SERT) blockers, or SSRIs, include fluoxetine, fluvoxamine, paroxetine, sertraline and citalopram. Paroxetine is the most potent SERT blocker, citalopram the most selective, and fluoxetine the longest lasting. Sertraline is also a potent blocker of the dopamine transporter. Venlafaxine is an SSRI at low dose and at high dose also blocks the norepinephrine transporter (NET).

Reference:

Question(s) 17: Clinical Adult
Discussion:
Axillary and inguinal freckling, cafe au lait spots, and cutaneous neurofibromas are characteristic skin findings in patients with neurofibromatosis type 1

Reference:

Question(s) 18: Pharmacology/Chemistry
Discussion:
Cerebral autosomal dominant arteriopathy with cerebral infarcts and leukoencephalopathy (CADASIL) is due to a mutation of the Notch-3 gene, which encodes for a protein that is critically involved in neural determination and neuritogenesis. Notch is a transmembrane receptor that is internalized after cleavage of its cytoplasmic domain by presenilin. Notch is then translocated to the nucleus where it controls expression of a variety of transcription factors. Studies in zebrafish, mice, and humans indicate that Notch works in conjunction with other angiogenic pathways to pattern and stabilize the vasculature.

Reference:
Kalaria RN. Advances in molecular genetics and pathology of cerebrovascular disorders. Trends Neurosci 2001;24:392-400.


Question(s) 19: Anatomy
Discussion:
Sensation to the umbilicus is carried by T10.

Reference:

Question(s) 20: Clinical Pediatrics
Discussion:
Pompe's disease is a lysosomal glycogen storage disease that affects practically all tissues and results from a defect of 1,4-glucosidase (acid maltase). Hypotonia,
failure to thrive, and decreased reflexes develop during the first few months of life. Cardiomegaly is prominent in infantile forms, which more commonly present with pulmonary insufficiency. Unlike other glycogenoses, the liver is normal in size or only slightly enlarged, and there are no abnormalities of glucose homeostasis. PAS-positive glycogen is seen in membrane-bound vacuoles in muscle, hepatocytes, and Schwann cells, but no abnormalities are seen in myelin sheaths.

Reference:

Question(s) 21: Physiology
Discussion:
Normal motor conduction velocities in a six month old child’s upper limb are around 20 - 36 m/sec

Reference:

Question(s) 22: Pharmacology/Chemistry
Discussion:
Glucose transporter type 1 deficiency may present with intractable seizures associated with low cerebrospinal fluid (CSF) glucose and low or low-normal CSF lactate. Patients respond to an alternative fuel source in the form of ketone bodies, as supplied by the ketogenic diet. The CSF profile may be differentiated from that seen in mitochondrial cytopathies, where low glucose is typically accompanied by high lactate. Glycine encephalopathy can present with intractable seizures from birth and is associated with normal CSF glucose and lactate, and with a high CSF glycine (and elevated CSF:plasma glycine ratio). Glycine encephalopathy does not respond to the ketogenic diet.

Reference:

Question(s) 23: Clinical Adult
Discussion:
The collateral blood supply to the mid-thoracic (T5-T7) cord is relatively tenuous making this region of the spinal cord most vulnerable to ischemia.

Reference:

Question(s) 24: Clinical Adult
Discussion:
Myokymia on EMG is very suggestive of radiation-induced plexopathy. Presence of Horner's syndrome, a discrete mass on CT scanning, lower trunk involvement, and the presence of pain all favor neoplastic over radiation-induced brachial plexopathy.

Reference:

Question(s) 25: Behavioral
Discussion:
Patients with AIDS dementia complex have a subcortical dementia with psychomotor slowing, difficulty concentrating, especially in conducting serial 7’s, impaired reading, and forgetfulness.

Reference:
Question(s) 26: Clinical Adult

Discussion:
Reactivation of herpes zoster virus in the geniculate ganglion (Ramsay Hunt syndrome) produces an acute CN VII palsy, sometimes with associated hearing loss. Usually there is a characteristic vesicular rash on the external ear and sometimes on the ipsilateral side of the tongue.

Reference:

Question(s) 27: Clinical Adult

Discussion:
The primarily demyelinating form of hereditary motor and sensory neuropathy is designated as HMSN I, or Charcot-Marie-Tooth disease type 1A (CMT 1). In the majority of families with autosomal dominant CMT 1, the disease is associated with duplication of a 1.5 megabase pair region on chromosome 17; these families are classified as CMT 1A. Deletion of the same chromosomal region has been demonstrated in patients with hereditary neuropathy with liability to pressure palsies (HNPP), also called tomaculous neuropathy. This is also inherited in an autosomal dominant pattern.

Reference:

Question(s) 28: Clinical Adult

Discussion:
Early treatment with dexamethasone improves the outcome in adults with acute bacterial meningitis (particularly pneumococcal meningitis) and does not increase the risk of gastrointestinal bleeding.

Reference:

Question(s) 29: Anatomy

Discussion:
The "locked-in" (deafferented state) consists of quadriplegia, aphonia and horizontal gaze impairment. Corticospinal, corticobulbar and corticopontine tracts are all involved. The quadriplegia is due to bilateral corticospinal tract involvement. Aphonia is due to involvement of the corticobulbar tract destined to the lower cranial nerves. Horizontal gaze paralysis is due to involvement of the fascicles of cranial nerve VI. Because the reticular formation is not usually affected, the patient is awake. In addition, since the supranuclear oculomotor pathways are dorsal and in the midbrain, the patient can look up and blink. A remarkable first hand account of this horrific syndrome is given in "The Diving Bell and the Butterfly" by Jean-Dominique Bauby.

Reference:

Question(s) 30: Anatomy

Discussion:
Mastication is spared with a facial nerve lesion. The facial nerve conveys special visceral efferent fibers (to the buccinator), general visceral efferent fibers (to parasympathetic ganglia), special visceral afferent fibers (from taste buds), and general somatic afferent fibers (from the skin of the external auditory canal).

Reference:

Question(s) 31: Anatomy
Discussion:
The dentate nucleus provides GABAergic innervation to the inferior olivary nucleus (nucleo-olivary tract). This pathway constitutes a negative feedback loop that uncouples the inferior olivary oscillator. The other components of this circuit are the olivocerebellar and olivonuclear pathways and the dentro-rubro-olivary pathway.

Reference:

Question(s) 32: Anatomy
Discussion:
The neurohypophysis contains fenestrated capillary endothelium with tight junctions. It is a circumventricular organ that has no blood-brain barrier.

Reference:

Question(s) 33: Clinical Adult
Discussion:
Optic neuritis is an inflammatory or autoimmune disease process affecting the optic nerve causing relatively acute impaired vision, progressing over hours to days. It is more common in women and affects patients who are 20 to 50 years of age. The optic disc is normal in approximately two-thirds of patients and swollen in one-third. Pain in the eye, often exacerbated by movement, occurs in greater than 90 percent of patients.

Reference:

Question(s) 34: Physiology
Discussion:
Fusimotor fibers transmit impulses to muscles spindles.

Reference:

Question(s) 35: Pharmacology/Chemistry
Discussion:
Periodic movements of sleep (nocturnal myoclonus) consist of the occurrence during sleep of periodic episodes of highly stereotypic leg muscle jerks. If severe enough to warrant therapy, pergolide, pramipexole, or controlled release levodopa/carbidopa at bedtime is often effective.

Reference:

Question(s) 36: Physiology
Discussion:
A unilateral P100 abnormality indicates an ipsilateral lesion of the visual pathway anterior to the optic chiasm such as a unilateral demyelinating process or optic nerve glioma. A tumor of the occipital lobe or a thalamic hemorrhage would cause a bilateral abnormality or little or no effect on the P100 latency.

Reference:

Question(s) 37: Pharmacology/Chemistry
Discussion:
Tardive dyskinesia results from chronic treatment with dopaminergic D2 receptor blockers. These include the classical high-potency neuroleptics, but also antiemetic agents such as prochlorperazine (Compazine) or metoclopramide (Reglan).

Reference:
Tucker GJ. Psychiatry for the neurologist.
Question(s) 38: Clinical Adult
Discussion:
CADASIL (cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy) is a hereditary disorder that presents with ischemic strokes, dementia, migraine with aura, and emotional-intellectual disturbances. Specific diagnosis can be made by skin-muscle biopsy which demonstrates thickening of smooth arteriopathic muscle cells that eventually degenerate. Electron microscopy can diagnose the disease by identifying granular, osmophilic materials in arterial smooth muscle.

Reference:

Question(s) 39: Anatomy
Discussion:
A lesion in the base of the midbrain (bas peduncular region) will produce an ipsilateral third nerve palsy and contralateral hemiparesis (Weber's syndrome).

Reference:

Question(s) 40: Behavioral
Discussion:
Inability to produce melody in verbal output, amelodia, produces a significant loss of non-verbal affective expression (affective motor aprosodia). The causative lesion is most commonly found in the posterior inferior frontal lobe of the right hemisphere, the region in the right frontal lobe analogous to Broca's area in the left.

Reference:

Question(s) 41: Anatomy
Discussion:
The common peroneal nerve divides into two branches, the deep and superficial peroneal nerves. The deep peroneal nerve innervates the tibialis anterior and extensor digitorum brevis. The peroneus longus and brevis are supplied by the superficial peroneal nerve. The soleus is innervated by the tibial nerve.

Reference:

Question(s) 42: Behavioral
Discussion:
Callosal apraxia results from a lesion in the genu of the corpus callosum. This results in a left limb kinetic apraxia. Tactile and auditory input cross the corpus callosum posteriorly and are therefore unaffected by a genu lesion. Alexia without agraphia results from a left occipital splenium of the corpus callosum lesion.

Reference:

Question(s) 43: Pathology
Discussion:
This patient was deficient for vitamin B12. Intrinsic factor is made by the parietal cells of the stomach and is necessary for B12 absorption. B12 deficiency causes subacute combined degeneration of the spinal cord. The cord will show spongy vacuolization of the posterior columns and lateral corticospinal tracts.

Reference:
Graham DI, Lantos PL. Greenfield's Neuropathology. 7th ed. London: Arnold,
Question(s) 44: Physiology
Discussion:
Compression of a large diameter nerve fiber results in preferential loss of fine discriminative touch as opposed to loss of pain or temperature sensations.

Reference:

Question(s) 45: Pathology
Discussion:
Hypertensive hemorrhages occur most commonly in the putamen. Less commonly they occur in the thalamus, pons and cerebellar hemispheres.

Reference:

Question(s) 46: Pathology
Discussion:
The pathognomonic feature of high intracranial pressure and transtentorial herniation is a wedge of pressure necrosis in one or both parahippocampal gyri. Hemorrhagic infarction due to entrapment of the posterior cerebral artery, and entrapment of the third cranial nerves also frequently occur. Secondary brainstem hemorrhages and infarctions are maximal in the midbrain and pons, and rarely extend to the medulla.

Reference:

Question(s) 47: Pharmacology/Chemistry
Discussion:
In patients with detrusor hyperreflexia without outlet obstruction or urinary retention, anticholinergic drugs, including oxybutynin, are the most appropriate treatment. If retention occurs, this should be combined with intermittent self-catheterization.

Reference:

Question(s) 48: Anatomy
Discussion:
The anterior cingulate gyrus has an inhibitory influence on the micturition reflex and would be the most likely site of impairment.

Reference:

Question(s) 49: Pathology
Discussion:
Mycotic aneurysms are most commonly found in the distal branches of the middle cerebral artery.

Reference:

Question(s) 50: Pathology
Discussion:
Findings from several multicenter trials have confirmed that folic acid supplements reduce the risk of both first event and recurrent neural tube defects in babies, with the estimated reduction rate of 60 and 72%, respectively.

Reference:

Question(s) 51: Physiology
Discussion:
The presence of normal facial nerve CMAP on the weak side, with absence of R1 and R2 responses on the affected side indicates a demyelinating lesion without axonal loss. The prognosis for recovery of function is excellent.

Reference:

Question(s) 52: Pathology
Discussion:
Tomaculous neuropathy (also known as pressure sensitive neuropathy) is characterized by focal hypermyelination with redundant myelin folds best seen on teased nerve preparations. Afflicted individuals have susceptibility to pressure palsies following relatively trivial compression.

Reference:

Question(s) 53: Physiology
Discussion:
Multifocal motor neuropathy (MMN) will have focal conduction blocks in two or more motor nerve fibers, usually distinct from common entrapment sites. Fibrillation potentials, fasciculation potentials, normal sensory responses and large motor unit action potentials can be seen in either MMN or ALS.

Reference:

Question(s) 54: Pathology
Discussion:
Saccular aneurysms usually become manifest over age 40, with less than 5% encountered under age 20. Familial cases are rare. About 80-90% occur on the internal carotid artery or on anterior branches of the circle of Willis. Septic aneurysms exist but are not saccular (berry) aneurysms and most occur on distal vessel branches, not proximally as do saccular aneurysms. It is important to recognize the frequency of multiple saccular aneurysms in patients with subarachnoid hemorrhage. The reported frequency is 8.6-31%, with most studies citing 20-25% of patients harboring multiple saccular aneurysms.

Reference:

Question(s) 55: Clinical Pediatrics
Discussion:
Tuberous sclerosis complex is an autosomal dominant syndrome resulting from mutations in one of two tumor suppressor genes, TSC 1 or TSC 2. Characteristic lesions include both hamartias and hamartomas. Both of these are circumscribed groups of cells that are misaligned or otherwise disrupted in their architectural relationships, and often have dysplastic features. The cell types are, however, appropriate for the tissue in which they are found. Hamartias have no growth potential. Lesions in this category include
cortical tubers, retinal flat lesions and renal cysts; hamartomas have the potential to grow. Examples of the latter include facial angiofibromas, subependymal giant cell astrocytomas and renal angiomylipomas. Mental retardation occurs as a result of poorly controlled seizures, and intellect is usually normal in the absence of seizures. Hypomelanotic macules are a frequent skin finding, but are not pathognomonic of the disease. Some patients, particularly women, develop pulmonary involvement that may be manifest as recurrent pneumothoraces.


**Question(s) 56: Behavioral Discussion:**
Major depression is frequent in Parkinson’s disease, occurring in 40 to 60% of patients during the course of their illness. It is less common in dementia of the Alzheimer type, Lewy body dementia, frontotemporal dementia, and NPH.


**Question(s) 57: Clinical Adult Discussion:**
In the presence of a positive HIV test the most probable brain mass lesion would be either toxoplasmosis or B-cell non-Hodgkin’s lymphoma. Kaposi’s sarcoma rarely metastasizes to brain.


**Question(s) 58: Clinical Adult Discussion:**
In complete section of the corpus callosum, left limb apraxia occurs because pathways connecting language areas in the left hemisphere to the motor areas of the right hemisphere are interrupted. Motor, sensory and visual functions of the right hemisphere are otherwise unaffected. Alexia without agraphia occurs with lesions of the left occipital lobe that involve fibers crossing from the right hemisphere through the splenium of the corpus callosum.


**Question(s) 59: Clinical Pediatrics Discussion:**
The clinical and laboratory features are typical of molybdenum cofactor deficiency. The cofactor is essential for the normal function of sulfite oxidase, xanthine oxidase and aldehyde oxidase. Wilson disease may produce hypouricemia, but does not present neurologically this early in life. Menkes disease is an X-linked copper transport disorder that presents in boys. Homocystinuria causes dislocated lenses, but does not account for the metabolic abnormalities and early, aggressive onset of disease. Pyridoxine dependency is associated with intractable seizures, but not the rest of the clinical or metabolic features described.


**Question(s) 60: Pharmacology/Chemistry Discussion:**
Multiple sclerosis (MS) is an autoimmune inflammatory demyelinating disease of the central nervous system (CNS) directed by autoreactive CD4+ T cells with proinflammatory properties recognizing myelin antigens. Nonglycosylated interferon-(IFN-) 1b has proven to be beneficial in relapsing-remitting multiple sclerosis (RRMS) because of its ability to reduce relapse rate, as well as activity and burden of the disease as assessed by magnetic resonance imaging (MRI).

Question(s) 61: Physiology
Discussion: Metachromatic leukodystrophy is associated with demyelinating sensorimotor polyneuropathy.


Question(s) 62: Pathology
Discussion: Onion bulbs are the result of repeated episodes of demyelination and remyelination and are composed of concentric rings of Schwann cells. They are prominent in demyelinating neuropathies such as Charcot-Marie-Tooth disease.


Question(s) 63: Physiology
Discussion: The proximity of the sciatic nerve explains its vulnerability during hip replacement surgery. The peroneal division of this nerve is often more severely affected. The abnormalities of the superficial peroneal and sural SNAPs serve to localize the lesion distal to the dorsal root ganglion, and argue against a lumbosacral root lesion. The innervation of the short head of the biceps femoris and anterior tibialis muscles both come from the peroneal division of the sciatic nerve.


Question(s) 64: Physiolog
Discussion: Burst suppression pattern in hypoxic ischemic coma carries an extremely poor prognosis.


Question(s) 65: Physiology
Discussion: Focal polymorphic delta slowing can be seen with focal cerebral lesions such as a supratentorial tumor, a cerebral infarction or cerebral abscess. Pseudotumor cerebri is usually associated with mild generalized slowing or a normal EEG. Lacunar infarctions are not generally associated with prominent EEG findings.


Question(s) 66: Behavioral
Discussion: Over half of epileptics have one or more episodes of significant depression during the course of the disorder and the suicide rate is considerably greater than in the general population.

Reference: Mendez MF, Cummings JL, Benson DF. Depression in epilepsy. Significance and phenomenology. Arch Neurol 1986;43:766-
Question(s) 67: Clinical Pediatrics

Discussion:
Night terrors (pavor nocturnus) in children are an arousal during slow-wave sleep and characteristically occur during the first half of the night 30 minutes after onset of sleep. The child often cries out, is agitated and is uncommunicative. Treatment is not necessary, but often the episodes are confused with nocturnal complex partial seizures and sedative medication can temporarily suppress the episodes.

Reference:

Question(s) 68: Physiology

Discussion:
Fasciculation potentials result from irritation of the alpha motor neuron, and are seen in a variety of neurogenic conditions, but can occur in normal individuals. They are frequently seen in motor neuron disease, but usually in association with other abnormalities of spontaneous activity, motor unit action potential morphology or recruitment.

Reference:

Question(s) 69: Anatomy

Discussion:
The sweat glands are innervated by postganglionic sympathetic cholinergic fibers. Cholinergic inputs stimulate sweat production via M3 type muscarinic receptors.

Reference:

Question(s) 70: Behavioral

Discussion:
Reduplication of place (reduplicative paramnesia) has been associated with combined lesions in the right parietal and bifrontal areas.

Reference:

Question(s) 71: Anatomy

Discussion:
With a right unilateral lesion of the medial longitudinal fasciculus, the patient can abduct the left eye on attempted gaze to the left but the right eye cannot be adducted. Nystagmus occurs in the left (abducting) eye.

Reference:

Question(s) 72: Anatomy

Discussion:
The oculomotor nerves pass between the proximal portions of the posterior cerebral and superior cerebellar arteries.

Reference:

Question(s) 73: Anatomy

Discussion:
Compression of the optic apparatus at the junction of the optic nerve and the optic chiasm will damage fibers streaming to the chiasm from the ipsilateral eye as well as fibers from the contralateral eye's nasal
retina subserving temporal visual space. The patient has a junctional scotoma consisting of ipsilateral visual loss and a contralateral temporal visual field defect. For those inclined toward eponyms, this is known as the anterior chiasmal syndrome of Traquair, and the looping forward fibers from the contralateral eye are known as Von Willebrand's knee.

Reference:

Question(s) 74: Anatomy
Discussion:
The occipital horn exhibits a high degree of variability and asymmetry and is often rudimentary. Knowledge of this variability is important in order to avoid attributing asymmetry of the occipital horns to disease.

Reference:

Question(s) 75: Clinical Adult
Discussion:
The anti-MuSK antibody is a newly discovered antibody found to be the cause of myasthenic syndromes in some patients previously considered 'seronegative'. Testing for this antibody marker is now commercially available. Until recently, the clinical phenotype of this disease and features distinct from myasthenia gravis due to acetylcholine receptor antibodies was unknown.

Reference:

Question(s) 76: Clinical Adult
Discussion:
Autoantibodies to GAD (glutamic acid decarboxylase) may occur with stiff man (stiff person) syndrome.

Reference:

Question(s) 77: Clinical Adult
Discussion:
Alexia without agraphia (pure alexia) results from damage to the pathways conveying visual information inputs from both hemispheres to the dominant angular gyrus, which itself remains intact but disconnected from visual regions. This syndrome is usually due to combined lesions of the dominant medial occipital region and the inferior fibers of the splenium of the corpus callosum. It can also be seen with a single lesion of the dominant occipitoparietal periventricular white matter beneath and beside the occipital horn of the lateral ventricle.

Reference:

Question(s) 78: Pathology
Discussion:
Neonatal meningitis is due mainly to group B streptococci (Streptococcus agalactiae) and Escherichia coli.

Reference:


Question(s) 79: Anatomy
Discussion:
Most of the parasympathetic neurons in the ciliary ganglion project fibers to the ciliary muscles to change the shape of the lens for accommodation. A small number of fibers
project to the iris sphincter to constrict the pupil.

Reference:

Question(s) 80: Clinical Adult
Discussion:
The Hallpike maneuver is most likely to be abnormal in a patient with benign paroxysmal positional vertigo.

Reference:

Question(s) 81: Behavioral
Discussion:
Quetiapine is an effective antipsychotic with very little dopaminergic blocking activity. Hence, it is useful in Parkinson's disease patients with hallucinations or delusions. It acts predominantly on serotonin receptors. Risperidone, perphenazine, thioridazine, and haloperidol all have more dopamine blocking properties than quetiapine.

Reference:

Question(s) 82: Pathology
Discussion:
Vacuole formation in muscle fibers is the most common change in hypokalemic periodic paralysis. The vacuoles are most prominent during the attack and are much less prominent between attacks. Group atrophy is indicative of neurogenic atrophy. Increased numbers of mitochondria may be seen in mitochondrial myopathies, which may also exhibit abnormally shaped mitochondria with paracrystalline inclusions. Lymphorrhages are small collections of lymphocytes seen at the motor end plate in myasthenia gravis. Abnormalities of the Z band are seen in nemaline myopathy.

Reference:

Question(s) 83: Physiology
Discussion:
Wicket waves, or wicket spikes, constitute a benign pattern of uncertain clinical significance, occurring predominantly in the EEG of older adults during light sleep.

Reference:

Question(s) 84: Clinical Adult
Discussion:
A spinocerebellar syndrome with ataxia, ophthalmoparesis, and Babinski signs developing in a patient with a long history of fat malabsorption suggests a deficiency of vitamin E.

Reference:

Question(s) 85: Anatomy
Discussion:
The adductor pollicis normally brings the thumb toward the palm. When the ulnar nerve is non-functional, the flexor pollicis assumes the role of adducting the metacarpal.

Reference:
Discussion:
Dopaminergic agonists and inhibitors of dopamine reuptake have been used to treat the behavioral traits associated with medial frontal syndrome. Methylphenidate is such a drug. This syndrome may be seen after ischemia in the distribution of the anterior cerebral artery.

Reference:

Question(s) 87: Anatomy
Discussion:
To reach its area of distribution, the posterior cerebral artery crosses the border of the tentorium cerebelli from the infratentorial compartment into the supratentorial compartment. Increased intracranial pressure in the supratentorial compartment may compress the posterior cerebral artery against the tentorium cerebelli leading to compromised blood flow to the visual cortex.

Reference:

Question(s) 88: Physiology
Discussion:
The N13 waveform of the median somatosensory evoked potential is referable to postsynaptic activity in the cervical cord.

Reference:

Question(s) 89: Pathology
Discussion:
Ammon's horn sclerosis is seen in chronic temporal lobe epilepsy and may reflect hypoxic or excitotoxic injury to this structure.

Reference:

Question(s) 90: Physiology
Discussion:
Valproic acid is the drug of choice for patients with absence and tonic clonic seizures and the 3Hz spike-and-wave pattern on the EEG. Ethosuximide, while effective for absence seizures, will not effectively treat the generalized tonic clonic seizures. Carbamazepine and phenytoin are less effective against absence seizures.

Reference:

Question(s) 91: Physiology
Discussion:
The afferent limb of the blink reflex is carried via the trigeminal nerve ipsilateral to the stimulus. The efferent limb is transmitted through bilateral facial nerves. A pattern of absent R1 and ipsilateral R2 response on stimulation of one side and absent contralateral R2 to stimulation of the opposite side is consistent with a lesion of the facial nerve.

Reference:

Question(s) 92: Pathology
Discussion:
During the early clinical phases of most neurovirologic diseases, the viral DNA is present. Only polymerase chain reaction testing on cerebrospinal fluid (CSF) is useful. CSF tests for herpes virus DNA are highly specific and sensitive. The CSF IgG antibody response is unlikely to be well-developed by three days after the onset of clinical symptoms. Serum IgG antibody tests are not informative, since they are positive in
most non-neurologically ill, adult individuals. Cultures are virtually never positive for herpes simplex virus in patients with herpes simplex encephalitis.

**Reference:**

**Question(s) 93:** Pathology

**Discussion:**
Trinucleotide repeats underlie myotonic dystrophy, Huntington's disease, and fragile X mental retardation, among other conditions. A small number of trinucleotide repeats are present in normal individuals, increased numbers of repeats lead to these autosomal dominant disorders, and transgenerational increase in the number of trinucleotide repeats are associated with earlier onset of disease.

**Reference:**

**Question(s) 94:** Anatomy

**Discussion:**
The middle cerebellar peduncle consists of crossed afferent fibers from the pontine nucleus, the pontocerebellar tract.

**Reference:**

**Question(s) 95:** Physiology

**Discussion:**
Sleep markedly enhances the presence of temporal spikes.

**Reference:**

**Question(s) 96:** Pathology

**Discussion:**
Synaptophysin is the most specific and useful immunohistochemical marker for assessing neuronal differentiation; other useful neuronal markers include neurofilament proteins, chromogranin, and Neu-N. S-100 protein is positive in neural crest derivatives, but is not specific for neurons because Schwann cells and CNS glial cells (astrocytes, oligodendrocytes and ependymal cells) are also strongly immunoreactive. Similarly, despite the optimistic name, neuron-specific enolase has a wide expression distribution beyond the nervous system and glial tumors such as glioblastoma are often immunopositive. Neu-N is a more recently introduced neuronal marker with nuclear and cytoplasmic localization. Vimentin is a cytoskeletal intermediate filament protein and cells of many lineages exhibit immunoreactivity for this protein.

**Reference:**

**Question(s) 97:** Anatomy

**Discussion:**
Visual, auditory, gustatory and nociceptive sensory modalities reach the cerebral cortex through the thalamus. Olfactory pathways bypass the thalamus.

**Reference:**

**Question(s) 98:** Behavioral

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

Reference:

Question(s) 99: Pathology
Discussion:
Nerve specimens from patients with Krabbe disease and metachromatic leukodystrophy contain characteristic deposits of sphingolipids. Lead intoxication produces a demyelinating neuropathy. Alcoholic neuropathy is an axonal neuropathy with secondary demyelination. Canavan's disease shows characteristic spongiform leukodystrophy only in the brain.

Reference:

Question(s) 100: Pharmacology/Chemistry
Discussion:
Paroxysmal kinesigenic dyskinesia is characterized by asymmetrical dystonic or choreic movements brought on by startle or sudden movements. These attacks respond well to low dose anticonvulsants.

Reference:

Question(s) 101: Pathology
Discussion:
Saccular (“berry”) aneurysms of the circle of Willis are present in 10-30% of patients with adult polycystic kidney disease, in whom they are responsible for significant morbidity and an important cause of death. Saccular aneurysms are also associated with fibromuscular dysplasia, coarctation of the aorta and moyamoya disease.

Reference:

Question(s) 102: Anatomy
Discussion:
The central nucleus of the inferior colliculus, medial geniculate body, nucleus of the trapezoid body, and superior olivary nucleus are part of the auditory pathway. The inferior olive is not.

Reference:

Question(s) 103: Pathology
Discussion:
The current World Health Organization (WHO 2000) criteria constitute one of three grading systems for the fibrillary (diffuse) astrocytomas that are currently used internationally. The other two systems are the St. Anne/Mayo classification and the modified Ringertz system. Under the WHO 2000 criteria, diffuse astrocytomas that exhibit nuclear atypia and marked mitotic activity are classified as anaplastic astrocytomas (WHO grade III). If either vascular proliferation or necrosis had been present, the neoplasm would be upgraded to glioblastoma (WHO grade IV), but these features were not seen in this case.

Reference:

Question(s) 104: Anatomy
Discussion:
The infundibulum carries compounds from the hypothalamus to the neurohypophysis and adenohypophysis modulating the activities of these structures. The hypothalamic derived compound that tonically inhibits pituitary gland synthesis and release of prolactin is dopamine (also known as prolactin inhibitory factor). Regulation of other pituitary gland hormones is under positive control (i.e., hormone releasing factors) that are proteins. Interruption of the infundibulum leads to loss of tonic inhibition of prolactin secretion by dopamine and elevation of prolactin. Similarly, dopamine receptor antagonists will elevate prolactin and dopamine receptor agonists can suppress prolactin secretion and have been used in the medical management of prolactin secretion pituitary adenomas.

Reference:

Question(s) 105: Behavioral Discussion:
Patients with REM behavior disorder have lack of limb paralysis that normally accompanies REM sleep. Therefore, they act out their dreams, which can often involve aggressive or violent behavior.

Reference:

Question(s) 106: Physiology Discussion:
Periodic lateralized sharp wave discharges in a patient with a several day history of fever, seizures and obtundation would most likely suggest herpes simplex encephalitis.

Reference:
Elkind MS. The role of warfarin and aspirin in secondary prevention of stroke. Curr
Behavioral Discussion:
The memory disorder of early Alzheimer's disease involves impairment in word recall (recent memory) with normal digit span (immediate memory), and relatively spared remote memory.

Reference:

Clinical Adult Discussion:
Acute dystonic reactions are often observed after treatment with potent dopamine D2-receptor antagonists including metoclopramide. Cervical and limb dystonia is most common. Laryngeal dystonia may be a life-threatening form of dystonic reaction in these patients. Anticholinergic medication including diphenhydramine is the most appropriate treatment.

Reference:

Anatomy Discussion:
The arcuate fasciculus connects the frontal gyri with parts of the temporal lobe.

Reference:

Clinical Adult Discussion:
In benign paroxysmal positional vertigo, there is a latent period before the development of vertigo and nystagmus (vertical, torsional, or a combination) when the affected ear is in the down position on Hallpike-Dix testing, and resolution of symptoms within 60 seconds. The diagnosis is based on history and physical, with no need for further diagnostic testing in straightforward cases. Canalith repositioning treatment and cupololith repositioning treatment both produce remission in 60-70% of cases, and improvement in an additional 20-30% of cases.

Reference:

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

Reference:

Pharmacology/Chemistry Discussion:
Aspirin/sustained release dipyridamole (Aggrenox®) is an effective antiplatelet therapy. The most frequent side effect that may limit its use is headache.

Reference:

Clinical Adult Discussion:
Metoclopramide has been implicated as a cause of the neuroleptic malignant syndrome in some cases.

Reference:
Pharmacology/Chemistry

**Discussion:**
Zidovudine (AZT), used for treatment of HIV infection, is a thymidine analog that inhibits reverse transcriptase and mitochondrial DNS polymerase, leading to depletion of mitochondrial DNA. Myalgia, weakness and elevated CK levels occur after 6-11 months of exposure with doses of 800-1200 mg/day. The diagnosis requires muscle biopsy, which shows ragged red fibers, reflecting mitochondrial proliferation. Recovery occurs over several months after discontinuation of AZT.

**Reference:**

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**Question(s) 118:** Clinical Pediatrics

**Discussion:**
The findings are most suggestive of pantothenate kinase associated neurologic disorder, associated with mutations in PANK2. Frataxin is associated with Friedreich ataxia, SOD with familial ALS, PDH with a syndrome of lactic acidosis, seizures and neurologic deterioration, and ATM with ataxia telangiectasia.

**Reference:**

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**Question(s) 119:** Pharmacology/Chemistry

**Discussion:**
Combining levodopa with the catechol-O-methyltransferase (COMT) inhibitor entacapone has been shown to be an effective strategy in the management of Parkinson's disease (PD) patients experiencing motor fluctuations. Results show the most common dopaminergic side effects to be dyskinesia and nausea, which result from the increased bioavailability of levodopa and can be readily managed. Non-dopaminergic side effects include diarrhea and harmless urine discoloration.

**Reference:**

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**Question(s) 120:** Pathology

**Discussion:**
Central pontine myelinolysis (CPM) has been most closely associated with the too-rapid correction of profound hyponatremia. It has been described in a variety of patients including alcoholics with Wernicke-Korsakoff syndrome, but many non-alcoholic patient groups have also been reported. The latter include liver transplant recipients and patients with severe burns, malnutrition, severe electrolyte disorders, and anorexia. Hypophosphatemia, but not thyroid disorders, has also been postulated to be related to CPM.

**Reference:**

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**Question(s) 121:** Pathology

**Discussion:**
Sturge-Weber syndrome is characterized by facial vascular nevus in a trigeminal distribution and leptomeningeal vascular malformation of the occipital lobe. Some patients also have ocular choroidal angiomas.

**Reference:**

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**Question(s) 122:** Clinical Adult

**Discussion:**
Cerebral vasospasm is a serious complication of subarachnoid hemorrhage typically occurring between the 4th and 14th day after hemorrhage. It often presents with focal neurological deficits and drowsiness.

**Reference:**

Question(s) 123: Anatomy
Discussion:
The facial nerve conveys fibers to the stapedius, orbicularis oculi, platysma and zygomaticus muscles, and preganglionic parasympathetic fibers to the submandibular ganglion. The trigeminal nerve conveys special visceral efferent fibers to the muscles of mastication, the tensor tympani and tensor veli palatini.

Reference:

Question(s) 124: Pathology
Discussion:
Intimal fibroplasia is the major histopathological finding in the vessels in moyamoya syndrome. Neither complicated atherosclerotic plaques nor vasculitis are histological features. PAS-positive granules are seen in cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL).

Reference:

Question(s) 125: Clinical Pediatrics
Discussion:
It is estimated that the incidence of symptomatic optic pathway gliomas in NF1 is 1.5 to 7.5%. The vast majority of these tumors present before the age of 6. It is recommended that all young children with NF1 be followed closely by an ophthalmologist with an expertise in NF1 and that MRI scans to detect optic nerve gliomas be performed only in children who have ophthalmologic abnormalities. Optic pathway tumors in children with NF1 usually do not progress after they have come to medical attention. Surgical or medical intervention therefore is only indicated when there is evidence of progression.

Reference:

Question(s) 126: Anatomy
Discussion:
The upper visual fields ultimately project to the inferior lip of the contralateral calcarine sulcus.

Reference:

Question(s) 127: Pathology
Discussion:
PMP22 and P0 are peripheral nerve myelin proteins that are abnormal in the hereditary motor and sensory neuropathies (Charcot-Marie-Tooth disease: CMT). Overexpression of PMP22 occurs because of duplication of the PMP22 gene in CMT1A while mutations of the gene encoding P0 protein occur in CMT1B.

Reference:

Question(s) 128: Pathology
Discussion:
Chronic steroid myopathy may develop in Cushing's disease or during chronic steroid treatment for a variety of diseases. Moderate to severe atrophy of type 2 fibers
is a constant finding in both iatrogenic steroid myopathy and in Cushing's disease with myopathy.

Reference:

Question(s) 129: Anatomy
Discussion:
The anterior choroidal artery supplies the lateral part of the medial segment of the globus pallidus, the target of pallidotomy in patients with Parkinson's disease. The posterior communicating artery supplies the medial part of the medial segment.

Reference:

Question(s) 130: Physiology
Discussion:
In diseases of the sensory system that damage sensory fibers proximal to the dorsal root ganglia, sensation is impaired but the sensory nerve action potential remains normal.

Reference:

Question(s) 131: Anatomy
Discussion:
Light first encounters the innermost layer of the retina, the ganglion cell layer.

Reference:

Question(s) 132: Physiology
Discussion:
With acute (less than 1 week) nerve root compression, the only EMG abnormality may be a reduced recruitment of motor unit potentials. Fibrillation potentials are usually not seen for 7 - 10 days after onset of injury. Fasciculation potentials and large, polyphasic motor unit action potentials reflect chronicity.

Reference:

Question(s) 133: Anatomy
Discussion:
Unilateral deafness could result from lesions in the cochlea, cochlear branch of the eighth nerve, or both the dorsal and ventral cochlear nuclei.

Reference:

Question(s) 134: Pathology
Discussion:
Metastatic choriocarcinoma is frequently hemorrhagic and would be a major consideration in a young woman, particularly following a recent miscarriage which may be related to gestational choriocarcinoma. The hemorrhagic metastatic tumors include melanoma, renal cell carcinoma, choriocarcinoma, and lung carcinoma. Breast carcinoma is not commonly hemorrhagic, nor are meningiomas. CNS lymphoma in an otherwise healthy young woman would be unusual.

Reference:

Question(s) 135: Behavioral
Discussion:
Borderline personality disorder is manifested by a wide range of psychiatric and behavioral features, but it has a single consistent characteristic - instability of mood.

Reference:
American Psychiatric Association.
Question(s) 136:
Pharmacology/Chemistry
Discussion:
The pedunculopontine nucleus (PPN) receives inputs from the globus pallidus and subthalamic nucleus, and projects to the substantia nigra, thalamus, and medullary reticulospinal neurons. The PPN contains one population of cholinergic and another population of glutamatergic neurons. It has been implicated as a component of the mesencephalic locomotor region. There is loss of PPN neurons in Parkinson's disease, and this has been correlated to the gait abnormalities in this disorder.

Reference:

Question(s) 137:
Anatomy
Discussion:
The synapse of the afferent axons for muscle stretch reflexes is located in the anterior (ventral) horn. The muscle stretch reflex is monosynaptic directly from sensory neuron to anterior horn cell.

Reference:

Question(s) 138:
Clinical Pediatrics
Discussion:
Zellweger syndrome presents in infancy, and affected individuals have characteristic facial features with high forehead, hypoplastic supraorbital ridges and midface hypoplasia. Severe weakness, hypotonia and seizures are common. Diagnosis is confirmed by elevated levels of saturated and unsaturated very long-chain fatty acids in body fluids.

Reference:

Question(s) 139:
Pharmacology/Chemistry
Discussion:
Fetal vitamin K deficiency with hemorrhagic complications occurs in 10% of neonates born to mothers receiving antiepileptic drugs that induce liver metabolism of vitamin K, including phenobarbital and phenytoin. Women taking enzyme-inducing antiepileptic drugs should be treated with vitamin K1, 10-20 mg daily during the last month of pregnancy. Infants should receive 1 mg intramuscularly at birth and, if needed, fresh frozen plasma.

Reference:

Question(s) 140:
Physiology
Discussion:
The Achilles tendon reflex is a monosynaptic reflex with the efferent limb in the tibial nerve.

Reference:

Question(s) 141:
Anatomy
Discussion:
The first major branch of the internal carotid artery is the ophthalmic artery.

Reference:

Question(s) 142:
Behavioral
Discussion: The Gerstmann's syndrome - agraphia, acalculia, right/left disorientation, finger agnosia - strongly suggests damage in the left (dominant) parietal lobe.


Question(s) 143: Behavioral Discussion: Anosodiaphoria may be seen with right hemisphere lesions as in this patient. It is indifference to their condition despite the recognition of the deficit.


Question(s) 144: Pharmacology/Chemistry Discussion: Wernicke's encephalopathy may be precipitated by a carbohydrate load in the face of chronic, severe malnutrition. The characteristic clinical symptoms include mental status changes, ophthalmoplegia, nystagmus and ataxia. Parenteral thiamine can produce dramatic improvement.


Question(s) 145: Anatomy Discussion: The dominant hemisphere is predominantly responsible for language - social communication by means of symbols - regardless of whether the communication relies on visual or auditory information.

Families are sometimes frustrated when individuals with aphasia also are unable to communicate in writing until they understand that language has many facets that reside in the same part of the brain.


Question(s) 146: Physiology Discussion: The frequency of orthostatic tremor is uniquely high (13-18 Hz) which distinguishes it from other types of tremor which may occur while the patient is standing. Legs and lumbar paraspinal muscles are affected.


Question(s) 147: Behavioral Discussion: Patients with dementia of the Alzheimer's type most commonly have a fluent aphasia with progressive emptiness in spontaneous speech, preserved repetition, relatively better oral comprehension and diminished reading comprehension with better ability to read aloud.


Question(s) 148: 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 receives its blood supply from the anterior choroidal artery.
Reference:

Question(s) 149: Clinical Adult
Discussion:
Seizures are most likely after head trauma if there has been a penetrating head injury, especially in the central (parietal) area. The risk of posttraumatic seizures is greatest in patients with a seizure immediately after the head injury. Prophylactic anticonvulsant medications do not decrease the risk of posttraumatic epilepsy.

Reference:

Question(s) 150: Clinical Adult
Discussion:
The diaphragm is innervated by C3-C5, not C6. The brachioradialis muscle is innervated primarily by C5-C6 roots. The subscapular nerve innervates the teres major muscle and the radial nerve innervates the supinator muscle. The first dorsal interosseus is innervated by the ulnar nerve.

Reference:

Question(s) 151: Anatomy
Discussion:
The Babinski sign, an abnormal superficial plantar reflex, is elicited by stroking the sole of the foot, which is in the S1 dermatome.

Reference:

Question(s) 152: Anatomy
Discussion:
Obstructive hydrocephalus occurs when there is a physical obstruction to CSF flow within the ventricular system. The most common site of such obstruction is the cerebral aqueduct.

Reference:

Question(s) 153: Behavioral
Discussion:
Witzelsucht (inappropriate jocularity) is seen in patients with orbitofrontal cortex lesions. Lesions in the orbitofrontal cortex also include disinhibited and antisocial behavior.

Reference:

Question(s) 154: Pathology
Discussion:
The potential for invasion of mesenchymal tissues, including the dura (and superior sagittal sinus), cranial bones, muscles and scalp, is an intrinsic property that is frequently seen to a greater or lesser extent in many ordinary (WHO grade I) meningiomas. Invasion of these mesenchymal tissues may complicate surgical resection but does not constitute a criterion for upgrading to atypical (WHO grade II) or anaplastic (WHO grade III) meningioma. Brain invasion, in contrast, is much less commonly seen and its presence in the absence of prior surgical procedures warrants a diagnosis of atypical (WHO grade II) meningioma. Highly vascular meningiomas are sometimes referred to as angiomatos and have no association with aggressive behavior. The older term "angioblastic meningioma" encompassed a heterogeneous group of vascular dural-based tumors that included angiomatos meningioma, hemangiopericytoma and hemangioblastoma; it is an imprecise and obsolete term that should be avoided. Four histologic subtypes merit upgrading based on their potential for early recurrence and/or aggressive clinical behavior: clear cell (WHO grade II), chordoid (WHO grade II), rhabdoid (WHO grade III), and papillary (WHO grade...
III). In contrast, the remaining nine WHO-recognized meningioma subtypes are classified as low grade (WHO grade I): meningothelial, fibrous, transitional, psammomatous, angiomatous, microcystic, secretory, lymphoplasmacyte-rich, and metaplastic.

Reference:

Question(s) 155: Anatomy
Discussion:
Axons of the Purkinje cells are the only ones to emerge from the cerebellar cortex and project to the deep cerebellar nuclei.

Reference:

Question(s) 156: Anatomy
Discussion:
The abducens nerve is contained in the sheath of the internal carotid artery at the level of the cavernous sinus. The oculomotor (III), trochlear (IV) and ophthalmic (VI) nerves are located in the lateral wall of the cavernous sinus. The optic nerve occupies the optic foramen but not the cavernous sinus.

Reference:

Question(s) 157: Clinical Adult
Discussion:
Symptoms of perilymph fistula are variable, but may include recurrent vestibulopathy. Characteristic precipitating factors include cough, sneeze, straining, and exercise.

Reference:

Question(s) 158: Physiology
Discussion:
The combination of an abnormality of wave I and subsequent waves on the ipsilateral side and a contralateral prolongation of III-V is most often seen with a cerebellopontine angle tumor that compresses the brainstem.

Reference:

Question(s) 159: Anatomy
Discussion:
The hypocretin/orexin neurons of the lateral hypothalamus innervate the cholinergic and monoaminergic cell groups of the brainstem involved in control of REM sleep. Experimental and clinical evidence indicate that hypocretin neurons are affected in narcolepsy.

Reference:

Question(s) 160: Anatomy
Discussion:
The foramen ovale transmits the mandibular division of the trigeminal (V) nerve. The foramen rotundum transmits the maxillary division of the trigeminal (V) nerve.

Reference:
Discussion:
The internal arcuate fibers from the dorsal column nuclei (cuneate and gracile nuclei) decussate completely to form the medial lemniscus.

Reference:

Question(s) 162: Pathology
Discussion:
Target fibers, which are best seen with trichrome or NADH stains, are characteristic of acute denervation.

Reference:

Question(s) 163: Anatomy
Discussion:
On the side of a spinal cord hemisection there would be an upper motor neuron syndrome, greatly impaired discriminatory tactile sense, loss of kinesthetic sense, and reduced muscle tone. Contralateral to the lesion there would be loss of pain and temperature due to interruption of the ascending spinothalamic tracts.

Reference:

Question(s) 164: Pharmacology/Chemistry
Discussion:
Midodrine raises blood pressure by stimulating alpha-adrenergic receptors. It is a pro-drug with a short half-life and does not cause fluid retention like the mineralocorticoids.

Reference:

Question(s) 165: Pathology
Discussion:
Methanol intoxication causes necrosis of the bilateral optic and putamen.

Reference:

Question(s) 166: Physiology
Discussion:
Nonconvulsive status epilepticus can be prolonged and is associated with fairly abrupt deterioration in mental function and a paroxysmal EEG. These features differentiate non-convulsive status from the dementias.

Reference:

Question(s) 167: Clinical Adult
Discussion:
Pseudotumor cerebri is much more common in women and is characterized by normal CSF composition and normal ventricles on neuroimaging. The neurologic examination is typically normal, but sixth nerve palsies and enlarged blind spots may occur.

Reference:
Question(s) 168: Pharmacology/Chemistry

Discussion:
Niemann-Pick disease type C is characterized by autosomal recessive inheritance with gradual and progressive neurological deterioration. Patients have variable hepatosplenomegaly, and progressive neurodegeneration. Findings include vertical supranuclear (usually downgaze) palsy, ataxia, dystonia, seizures, gelastic cataplexy and dementia.

Reference:

Question(s) 169: Anatomy

Discussion:
Alexia without agraphia follows combined damage to the dominant medial occipital region and the inferior fibers of the splenium of the corpus callosum. This is in the distribution of the posterior cerebral artery.

Reference:

Question(s) 170: Pathology

Discussion:
Although increased internal nuclei are seen in many myopathies, this alteration is particularly noteworthy in myotonic dystrophy in which up to 30% of myofibers may bear internal nuclei. As in all muscle biopsies, care must be taken not to sample the muscle too close to a tendinous insertion as this area is normally rich in internal nuclei.

Reference:

Question(s) 171: Physiology

Discussion:
Paramyotonia congenita is a hereditary disorder with muscle stiffness that worsens with repetitive movements. It is exacerbated by cold temperatures. The EMG shows myotonia. Symptoms can last for hours but usually are too mild to require treatment.

Reference:

Question(s) 172: Pharmacology/Chemistry

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

Reference:


Question(s) 173: Behavioral

Discussion:
Misreaching under visual guidance (optic ataxia) and failure to scan and integrate an entire visual scene or picture(simultanagnosia) is part of Balint's syndrome. When these symptoms occur together the occipito-parietal region is the most common area affected. The third
clinical sign seen in Balint's syndrome is ocular apraxia.

Reference:

Question(s) 174: Pharmacology/Chemistry
Discussion:
Cerebral cysticercosis is a common cause of focal seizures in the Hispanic population living in the southern United States. The acute lesions typically resolve with formation of a granuloma, and therefore expectant treatment with anticonvulsants is suggested. If the cyst and/or edema persist in the two to three month follow-up CT or MRI then cysticidal therapy is indicated. The drugs of choice are praziquantel or albendazole. Dexamethasone is added to the treatment to reduce the inflammatory reaction induced by the acute destruction of the parasite.

Reference:

Question(s) 175: Anatomy
Discussion:
In C7 lesions, paresthesias may involve the index and middle finger and even the thumb, with atrophy and weakness in the triceps, wrist extensors, and pectoral muscles, and a parallel triceps reflex depression.

Reference:

Question(s) 176: Clinical Pediatrics
Discussion:
MELAS - mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes is one of several mitochondrial encephalopathies. In cases where a clear maternal inheritance has been established, there is a mutation in the mitochondrial tRNA leu. Cranial CT and MR scans in patients with MELAs reveal infarct-like areas bilaterally in the posterior cerebrum and almost always hypodensities or calcifications in the basal ganglia region.

Reference:

Question(s) 177: Physiology
Discussion:
Carpal tunnel syndrome (median neuropathy at the wrist) is commonly bilateral, worse in the dominant hand, has numbness involving the thumb, index, third, and frequently radial half of the fourth finger with sparing over the thenar eminence, weakness of the abductor pollicis brevis muscle with denervation here in more severe cases.

Reference:

Question(s) 178: Behavioral
Discussion:
This patient has a vascular dementia. This is based on the patient's history as well as the pattern of a subcortical type of dementia. Recent research has demonstrated that the cholinesterase inhibitors such as donepezil, rivastigmine, and galantamine are also effective in treating the cognitive deficits of vascular dementia. The cholinesterase inhibitors have also demonstrated efficacy in treating non-cognitive behavioral symptoms especially apathy.

Reference:
Cummings JL and Mega MS. Neuropsychiatry and Behavioral Neuroscience. New York: Oxford University
Question(s) 179: Behavioral Discussion:
Many authorities consider prosopagnosia as a more subtle form of visual agnosia and almost all patients with visual agnosia show difficulty in recognizing familiar faces.

Reference:

Question(s) 180: Anatomy Discussion:
The chemoreceptor trigger zone is located in the area postrema.

Reference:

Question(s) 181: Clinical Adult Discussion:
The serotonin syndrome displays myoclonus, fever, confusion, ataxia, movement problems, sweating, and shivering. Prominent myoclonus helps differentiate it from the neuroleptic malignant syndrome.

Reference:


Question(s) 182: Clinical Adult Discussion:
Alpha coma refers to the rare occurrence of clinical coma associated with EEG activity in the alpha frequency range (8-13 Hz) that is unresponsive to external stimuli. It has been reported with hypoxia, drug overdose, and lesions in the pontomesencephalic region. Prognosis depends on etiology.

Reference:

Question(s) 183: Pathology Discussion:
Brains at autopsy taken from schizophrenic patients show an increase, not decrease, in ventricular size, especially of the temporal horns of the ventricles due to loss of cortical gray matter in medial temporal lobes. Although cerebellar abnormalities of the vermis were reported in a single study, they were not verified in subsequent work. More dramatic changes such as heterotopias and major gyral abnormalities such as polymicrogyria are not generally seen in schizophrenic patients, either premortem on radiographic studies, or post-mortem.

Reference:

Question(s) 184: Anatomy Discussion:
The “frontal eye field” in humans is located in the caudal part of the middle frontal gyrus.

Reference:

Question(s) 185: Clinical Adult Discussion:
Extracranial vertebral artery dissections have most often been recognized in patients who had either trauma or chiropractic manipulation. Neurologic symptoms may be delayed by hours, days or weeks. Transient ischemic attacks (TIA’s) most often include
dizziness, diplopia, veering, staggering, and dysarthria. The diagnosis of arterial dissection has traditionally been made using standard catheter angiography, although MRI/MRA is very sensitive.

Reference:

Question(s) 186: Clinical Adult Discussion:
Herpes simplex type 1 is one of the most common causes of encephalitis in immunocompetent adults. It is due to reactivation of latent virus in the trigeminal ganglion and thus initially involves the overlying temporal lobes.

Reference:

Question(s) 187: Pharmacology/Chemistry Discussion:
Parkin parkinsonism is inherited in an autosomal recessive manner and is characterized by early onset parkinsonism and dystonia. The responsible gene product, parkin, is a ubiquitin ligase involved in the degradation of many proteins including alpha-synuclein.

Reference:

Question(s) 188: Anatomy Discussion:
Ballism is associated with discrete lesions in the subthalamic nucleus. The dyskinesia occurs contralateral to the lesion and is associated with hypotonia.

Reference:

Question(s) 189: Physiology Discussion:
In collateral reinnervation, surviving motor neurons reinnervate orphaned muscle fibers by sprouting terminal processes. Reinnervating motor units cannot increase their territory within a muscle. The transmission of electrical impulses from muscle fiber to muscle fiber is an example of ephaptic transmission and is the mechanism for the generation of complex repetitive discharges.

Reference:

Question(s) 190: Pathology Discussion:
Toxoplasmosis is a special hazard in patients with HIV infection and often appears as multiple ring-enhancing lesions on CT examination.

Reference:

Question(s) 191: Pharmacology/Chemistry Discussion:
Lamotrigine is efficacious for treatment of partial and generalized seizures, including the Lennox-Gastaut syndrome. It is metabolized in the liver. Valproate inhibits
metabolism and increases considerably the half life of lamotrigine. Lamotrigine is started at a dose of 50 mg/d for two weeks in patients not receiving valproate, and 25 mg every other day for two weeks in patients receiving the drug. The incidence of serious rash is 0.3% in adults and 1% in children. Co-administration with valproate and rapid escalation of the dose increase the risk of this complication.

Reference:

Question(s) 192: Pathology

Discussion:
The most common lesion attributed to sickle-cell disease is cerebral infarction. This is often due to occlusion of large arteries. The second most common complication is intracerebral hemorrhage. Subarachnoid hemorrhage is less common and attributed to rupture of aneurysms. Sagittal sinus thrombosis is rare.

Reference:

Question(s) 193: Anatomy

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

Reference:

Question(s) 194: Anatomy

Discussion:
The suprachiasmatic nucleus of the hypothalamus receives direct inputs from the retina. This retino-hypothalamic tract mediates influences of the day-night light cycle on the circadian pacemaker.

Reference:

Question(s) 195: Physiology

Discussion:
Spinal muscular atrophy is the most common cause of progressive motor neuron disease in infancy. Mutations of the survival motor neuron gene is present in 95% of children with spinal muscular atrophy type 1.

Reference:

Question(s) 196: Pharmacology/Chemistry

Discussion:
Niemann-Pick disease, type C (NPC) is an autosomal recessive lipidosis, in which impaired intracellular trafficking of cholesterol and glycolipids lead to intracellular sequestration of unesterfied cholesterol, and increased concentrations of glycolipids in the brain. Secondary responses to exogenous LDL-cholesterol loading, including down regulation of LDL receptors and HMG CoA reductase and upregulation of ACAT are impaired. There is no primary deficiency in these enzymes. At Least 95% of cases are associated with mutations in NPC1; the remainder are associated with mutations in NPC2 (HE1).

Reference:

2000;290:2298-2301.

Question(s) 197: Anatomy
Discussion:
The nucleus ambiguus contains motor neurons that supply striated muscles of the palate, pharynx and larynx; therefore, disruption of this nucleus will impair phonation.

Reference:

Question(s) 198: Pharmacology/Chemistry
Discussion:
Lovastatin (and other statins) induced necrotizing myopathy is attributed to the effects of the drug in inhibiting synthesis of mevalonic acid, a precursor of several essential metabolites, including coenzyme Q10 (ubiquinone). This toxic action is potentiated by clofibrate, gemfibrozil, nicotinic acid, and cyclosporine.

Reference:

Question(s) 199: Behavioral
Discussion:
The supplementary motor area (SMA) is thought to act as a pacemaker for verbal output. Damage to the SMA or to its connections with Broca's area produces the clinical picture of transcortical motor aphasia.

Reference:

Question(s) 200: Anatomy
Discussion:
Erb-Duchenne upper plexus paralysis occurs secondary to damage to the fifth and sixth cervical roots or upper trunk of the brachial plexus. It is a common deficit and is usually due to traumatic separation of the head and shoulder but may also be due to pressure on the shoulder, birth injuries, or idiopathic plexitis.

Reference:

Question(s) 201: Behavioral
Discussion:
Decreased hemoglobin and elevated mean corpuscular volume are compatible with a macrocytic anemia that may be secondary to vitamin B12 deficiency. Vitamin B12 deficiency, in turn may cause combined system degeneration (affecting posterior columns and lateral corticospinal tracts). Dementia and peripheral neuropathy may also be seen.

Reference:

Question(s) 202: Clinical Adult
Discussion:
Orthostatic tremor or “shaky leg” syndrome is a disorder of middle-aged or elderly people and is characterized by feelings of unsteadiness in the legs and a fear of falling when standing. Other associated symptoms include difficulty in initiating walking and leg discomfort when standing. The symptoms are attenuated by walking, abolished by sitting, and due to a high frequency tremor in the weight-bearing muscles.

Reference:
Discussion: Tardive dyskinesia, dystonic reactions, and Parkinsonian symptoms are hazards of metoclopramide administration, especially in the elderly.


Question(s) 204: Physiology

Discussion: Patients with increased intracranial pressure, such as that caused by ventricular outflow obstruction, typically have EEGs which show rhythmic slow activity in the theta-delta frequency range. The site is often distant from the site of obstruction. These EEG findings are not specific.


Question(s) 205: Clinical Adult

Discussion: REM-sleep behavior disorder is characterized by motor activity, often violent, accompanying dreams. Polysomnography reveals persistent EMG activity during periods that are otherwise typical of REM sleep. The condition is most common in elderly men, and usually responds well to clonazepam.


Question(s) 206: Behavioral

Discussion: This history depicts a case of herpes simplex virus encephalitis. The treatment of choice is acyclovir.


Question(s) 207: Behavioral

Discussion: Achromatopsia follows lesions that involve the occipital cortex inferior to the calcarine sulcus. Damage in this area produces a superior visual field defect (upper quadrant anopsia), and loss of color vision in the preserved inferior visual field.


Question(s) 208: Physiology

Discussion: Shoulder dislocation can result in injury of the axillary nerve. The clinical picture is of isolated deltoid weakness with numbness over the deltoid.


Question(s) 209: Physiology

Discussion: Typical triphasic waves would most likely suggest the presence of hepatic coma.


Question(s) 210: Clinical Adult

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

Reference:

Question(s) 211: Clinical Pediatrics
Discussion:
The combination of dysconjugate, highly variable nystagmus, head nodding and head tilt without ophthalmologic abnormalities, and with normal neuroimaging, is diagnostic of spasmus nutans. Latent nystagmus is a jerk nystagmus that is evoked or enhanced by covering one eye. Congenital nystagmus is usually conjugate and suppressed by convergence (in contrast to spasmus nutans, that is typically increased by convergence). Opsoclonus describes chaotic, conjugate saccades, seen classically in the paraneoplastic syndrome associated with neuroblastoma. Whipple's disease produces a convergence-divergence nystagmus with associated movements of the muscles of the head and neck (oculomasticatory myorhythmia).

Reference:

Question(s) 212: Clinical Adult
Discussion:
The most common cause of an isolated oculomotor (3rd) nerve palsy is an intracranial aneurysm usually arising from the junction of the internal carotid and posterior communicating arteries. This is especially true if the (3rd) nerve palsy is "complete" and involves the pupil as well as the extraocular muscles. When the pupillary light reflex is spared, ischemia to the oculomotor nerve is more likely and is most commonly associated with underlying diabetes mellitus.

Reference:

Question(s) 213: Physiology
Discussion:
Paraspinal muscle denervation usually indicates a lesion at the level of the ventral nerve root.

Reference:

Question(s) 214: Pharmacology/Chemistry
Discussion:
Tyrosine hydroxylase (TH) deficiency is an autosomal recessive disorder of biogenic amine synthesis in which the conversion of L-tyrosine to L-dihydroxyphenylalanine (L-dopa) is impaired. L-dopa is subsequently metabolized to dopamine, and to homovanillic acid (HVA) and 3-methoxy-4-hydroxyphenylethanol (via norepinephrine). Thus, HVA and 5-hydroxyindole acetic acid (5-HIAA) levels in cerebrospinal fluid are diminished in TH deficiency, although 5-HIAA, a metabolite of serotonin, is normal. Clinical manifestations include parkinsonian and dystonic features presenting in infancy. Improvement is seen with the administration of oral L-dopa.

Reference:

Question(s) 215: Anatomy
Discussion:
The long thoracic nerve innervates the serratus anterior muscle. Winging of the scapula results when the patient pushes against a wall, especially with the shoulder adducted. There may also be inability to abduct the arm beyond 90 degrees.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia:
Question(s) 216: Clinical Adult
Discussion:
The syndrome of spinocerebellar degeneration is most characteristic of prolonged chronic vitamin E deficiency.

Reference:

Question(s) 217: Clinical Adult
Discussion:
Lesions of the fifth lumbar nerve root produce denervation that is evident clinically in toe extensor, foot dorsiflexor, and foot invertor (posterior tibial) muscles. Involvement of paraspinous muscles (electromyographically) confirms that the site of the lesion is at the level of the nerve root.

Reference:

Question(s) 218: Anatomy
Discussion:
Secondary auditory fibers from the cochlear nuclei form the dorsal, intermediate, and ventral acoustic striae. The dorsal and intermediate striae cross the midline and enter the lateral lemniscus. The fibers of the ventral striae terminate in the superior olivary nuclei and the nucleus of the trapezoid body. These nuclei give rise to tertiary fibers that enter the lateral lemnisci. The lateral lemniscus ascends to the midbrain where most of the fibers terminate in the inferior colliculi.

Reference:

Question(s) 219: Behavioral
Discussion:
Electroconvulsive therapy (ECT) may improve both mood and motor symptoms in patients with advanced Parkinson's disease and major depression. ECT is safer than tricyclic antidepressants in patients with cardiac conduction disturbances. Nefazodone, amitriptyline, and imipramine would be poor choices because of their potential cardiac side effects and in the case of the later two drugs their anticholinergic properties. Anticholinergics, such as benztropine, are not effective in treating major depression and may cause cardiac side effects, memory loss, and confusion in the elderly.

Reference:

Question(s) 220: Pharmacology/Chemistry
Discussion:
Homocystinuria is inherited as an autosomal recessive disorder due to deficiency of cystathionine synthase. It is associated with long limbs, arachnodactyly, and downward dislocation of the lens (ectopia lentis). Approximately 50% of the patients have mental retardation. Most neurologic features result from cerebral thromboembolic disease. Approximately 40% of the patients respond to pyridoxine.

Reference:
Question(s) 221: Clinical Adult
Discussion:
A parasagittal frontal lesion may produce spastic paraparesis, and be mistaken for a spinal cord lesion.

Reference:

Question(s) 222: Anatomy
Discussion:
The fourth cervical dermatome comes to lie adjacent to the second thoracic dermatome on the trunk.

Reference:

Question(s) 223: Anatomy
Discussion:
The cortical projections of the anterior nucleus of the thalamus are to the cingulate gyrus. This is part of the classical Papez circuit that formed the basis of the concept of the limbic system.

Reference:

Question(s) 224: Clinical Adult
Discussion:
A femoral neuropathy or high lumbar plexus lesion associated with retroperitoneal pain in a hemophiliac with prolonged PTT suggests a retroperitoneal hemorrhage. While imaging and physiological tests might confirm the diagnosis, definitive treatment requires replacement with Factor VIII concentrates. The amount administered depends on the patient's Factor VIII level. When adequately replaced, surgical drainage based on imaging studies and his clinical condition could be considered. While antiphospholipid antibodies are an important cause of elevated prothrombin times, they would not be an important diagnostic possibility in a patient presenting with spontaneous hemorrhage at a young age.

Reference:

Question(s) 225: Anatomy
Discussion:
The suprachiasmatic nucleus receives direct bilateral projections from the retina and functions as a biologic clock.

Reference:

Question(s) 226: Behavioral
Discussion:
Topographic disorientation (impaired orientation and navigation in the environment) is most strongly associated with damage in the right posterior parahippocampal region or the infracalcarine cortex. Right parietal damage may cause a milder form of the disorder.

Reference:

Question(s) 227: Clinical Pediatrics
Discussion:
A partial third nerve palsy associated with headache in young children is most commonly due to ophthalmoplegic migraine. Cerebral aneurysms presenting as a partial third nerve palsy would be extremely unusual in young children. Myasthenia gravis does not present with pain and pupillary involvement would be unusual. Guillain-Barre, Miller Fisher Syndrome, and
a brain stem tumor would have other findings on careful neurologic examination.

Reference:

Question(s) 228: Anatomy
Discussion:
Functional imaging studies indicate that the contralateral lateral hemisphere of the cerebellum is activated during cerebral hemispheric cognitive activity.

Reference:

Question(s) 229: Clinical Adult
Discussion:
Rhinocerebral mucormycosis is a fulminant disease involving the frontal and nasal cavities with direct extension to the cranial cavity. Typically, this disease occurs in patients with uncontrolled diabetes and invades the juxtaposed cerebrum after local spread in the nasal-orbital cavities.

Reference:

Question(s) 230: Pharmacology/Chemistry
Discussion:
Facial nerve palsy, frequently bilateral, is the most common neurological sign of sarcoidosis. It is often accompanied by parotid gland swelling. Loss of taste is also common in conjunction with the facial weakness and indicates that the lesion is above the exit of the chorda tympani. Sarcoidosis is often responsive to corticosteroid therapy.

Reference:

Question(s) 231: Anatomy
Discussion:
The superior olivary nuclei are part of the brain stem auditory pathways and are involved in the function of the 8th cranial nerve.

Reference:

Question(s) 232: Clinical Adult
Discussion:
Respiratory muscle weakness with ventilatory failure is a potentially serious complication in patients with AIDP. Spirometry is the most effective measure of respiratory muscle function in these patients.

Reference:

Question(s) 233: Pathology
Discussion:
Bone fractures are associated with release of fatty bone marrow into the systemic circulation. Pulmonary circulation may be compromised, and emboli may travel to the brain leading to ball hemorrhages around small vessels. The symptoms described in this patient also fit best with fat embolism, rather than diffuse axonal injury.

Reference:
Question(s) 234: Pathology
Discussion:
Zellweger's cerebro-hepato-renal syndrome presents with hypotonia and cranio-facial dysmorphic features and the brain shows widespread neuronal migration defects, especially pachygyria and cerebellar abnormalities. Wolman's disease, due to acid lipase deficiency, presents with diarrhea, vomiting, failure to thrive, hepatosplenomegaly and adrenal calcification, but minimal CNS abnormalities. Farber's disease is characterized by painful swelling of the joints and subcutaneous nodules; neurons show stored material but widespread migrational abnormalities are rarely found in the brain. Pompe's disease is primarily a disorder of muscle and presents with hypotonia, and while neurons and astrocytes may show increased storage of glycogen, there are no associated migrational disorders in the brain. Krabbe's leukodystrophy demonstrates no migrational disorders in the brain.

Reference:

Question(s) 235: Anatomy
Discussion:
The amygdala plays a major role in the appreciation of fear. Individuals with bilateral amygdala dysfunction cannot recognize fearful facial expressions but have no difficulty with recognition of other emotions.

Reference:

Question(s) 236: Clinical Adult
Discussion:
The most important initial study to obtain in a patient over the age of 55 suspected of having temporal arthritis is an erythrocyte sedimentation rate.

Reference:

Question(s) 237: Pathology
Discussion:
Kinetic energy as defined by classical mechanics is equal to one-half the product of the mass of the projectile multiplied by the square of its velocity. Kinetic energy increases linearly with projectile mass, but increases as the square of the velocity; therefore, velocity is the single most important determinant of tissue injury during missile passage. Tumble and fragmentation can determine the rate at which kinetic energy is transferred and can alter the amount of damage done by a bullet possessed of a given kinetic energy.

Reference:

Question(s) 238: Physiology
Discussion:
Radiculopathy of the C5 root causes weakness of the supraspinatus, deltoid and rhomboid muscles, but to a lesser extent the biceps, brachioradialis and brachialis muscles. Pronator teres receives its innervation from the C6, C7 roots and is least likely to be involved in a C5 radiculopathy.

Reference:

Question(s) 239: Pathology
Discussion:
The plaque in multiple sclerosis is an area of destruction of myelin and oligodendroglia, with relative sparing of the axons and neurons. Perivascular mononuclear cell
infiltrates may be present but neither fibrinoid necrosis of vessels nor microglial clusters are found in typical patients. No intranuclear inclusions (as are seen with viral infections) are present in multiple sclerosis. Bizarre nuclear features in astrocytes and intranuclear inclusions in oligodendrocytes typify progressive multifocal leukoencephalopathy, not multiple sclerosis.

Reference:

Question(s) 240: Anatomy
Discussion:
In looking downward to the left, when the eye is turned outward, the depressor is the left inferior rectus. When eye is turned inward, the depressor is the right superior oblique.

Reference:

Question(s) 241: Physiology
Discussion:
14- and 6-per-second positive spikes are a benign phenomenon in the EEG and are seen over posterior and temporal head regions during drowsiness and light sleep.

Reference:

Question(s) 242: Anatomy
Discussion:
This patient has a cervical central spinal cord syndrome manifested by lower motor findings in his hands, upper motor neuron hyper-reflexia in his legs and a classical sensory disturbance in which secondary order neurons subserving pain and thermal sense are interrupted as they decussate in the central spinal cord. The primary order neuronal axons in the dorsal columns subserving fine touch and proprioception are spared. The central spinal cord syndrome most commonly results from syringomyelia or intrinsic tumors.

Reference:

Question(s) 243: Anatomy
Discussion:
Normal CSF pressure (lumbar) measured in recumbent position is 10 to 15 cm H2O.

Reference:

Question(s) 244: Pharmacology/Chemistry
Discussion:
Seizures and myoclonus are associated with use of high doses of meperidine. The excitability is due to a metabolite normeperidine, which has twice the CNS stimulant effect of meperidine.

Reference:

Question(s) 245: Pathology
Discussion:
The genetic basis and pathophysiology of most of the leukodystrophies have been known for some time. Light has recently
been shed on one of the last holdouts: Alexander disease. The morphologic hallmark of Alexander disease is the presence of profuse numbers of Rosenthal fibers. Rosenthal fibers are composed of densely compacted glial intermediate filaments made of glial fibrillary acidic protein (GFAP). Recent studies have demonstrated that a large percentage of Alexander disease cases are associated with a mutation in the GFAP gene.

Reference:


Question(s) 246: Clinical Adult Discussion:
The pain that occurs with meralgia paresthetica (damage to the lateral cutaneous nerve of the thigh) is often widely distributed and can include the low back, buttock, anterolateral thigh, and lateral knee regions. It is often aggravated by standing and relieved by sitting. Paresthesias and hypesthesia usually involve a much more limited area in the anterolateral thigh. The syndrome is often associated with obesity or pregnancy, and symptoms disappear with weight loss or delivery of the baby.

Reference:

Question(s) 247: Anatomy Discussion:
The fornix is the main efferent fiber system from the hippocampal formation. The precommissural fibers are distributed to the septal nuclei, lateral preoptic area, anterior part of the hypothalamus and nucleus of the diagonal band.

Reference:


Question(s) 254 - 258: Clinical Pediatrics

Discussion:
Aicardi syndrome and incontinentia pigmenti are X linked dominant disorders that are considered to be lethal in males. Becker muscular dystrophy is an X linked recessive disorder, biotinidase deficiency is felt to be a autosomal recessive disorder and Kearnes-Sayre is a mitochondrial disorder.

Reference:

Question(s) 259 - 263: Physiology

Discussion:
Emery-Dreifuss muscular dystrophy is associated with contractures of the neck and elbows. Dermatomyositis is associated with a rash on the dorsum of the fingers and of the eyelids. Myotonic dystrophy is associated with frontal balding, cataracts, endocrinopathies, cardiac defects, and mental retardation. Botulism has associated autonomic abnormalities including fixed, dilated pupils. ALS is often associated with frequent cramps.

Reference:

Question(s) 264 - 268: Physiology

Discussion:
With an acute neurogenic lesion, the only EMG abnormality may be a reduced recruitment pattern. Myokymic discharges are a feature of radiation plexopathy. Myotonic discharges are seen in a variety of myopathies, and may be observed with or without clinical myotonia. In adult acid maltase deficiency, myotonic discharges are often seen, and occur in the absence of clinical myotonia. Morphology and recruitment of MUAPs are often normal in endocrine myopathies and myasthenia gravis (MG). There may be increased variability of MUAP morphology on consecutive firings in MG.

Reference:

Question(s) 269 - 273: Physiology

Discussion:
The following diseases should have these electrodiagnostic findings: Friedreich's ataxia - sensory polyneuropathy, Lambert-Eaton myasthenic syndrome - small CMAPs, incremental response to 10 sec exercise, myasthenia gravis with cholenergic crisis - repetitive motor responses after single supramaximal motor stimulus and decrement with repetitive stimulation at 2 Hz, Charcot-Marie-Tooth disease Type 1A - prolonged motor and sensory responses with very slow conduction velocities, hereditary neuropathy with susceptibility to pressure palsies (PMP22 gene defect) - conduction blocks.

Reference:

Question(s) 274 - 278: Pharmacology/Chemistry

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

Reference:


Question(s) 279 - 281: Clinical Pediatrics Discussion:
Symptoms in opsoclonus-myoclonus syndrome typically precede detection of occult neuroblastoma. Medulloblastoma, some ependymomas and ependymoblastomas have the capability to seed along the CSF pathways. Children with neurofibromatosis have an increased incidence of optic pathway gliomas.


Question(s) 282 - 285: Pathology Discussion:
Patients with von Hippel-Lindau syndrome have hemangioblastomas of the central nervous system (CNS). Patients with tuberous sclerosis may develop subependymal giant cell astrocytomas that protrude into the ventricular system. Patients with neurofibromatosis often have neurofibromas and schwannomas. Children with the "bathing trunk" epidermal nevus syndrome may develop cutaneous and CNS melanomas.


Question(s) 286 - 288: Physiology Discussion:
Breach rhythms are enhanced cerebral rhythms over the area of a skull defect. Mu activity is located over the central regions and is reactive to movement and touch. Rolandic spikes are seen in children with benign rolandic epilepsy.


Question(s) 289 - 293: Clinical Pediatrics Discussion:
The eye findings in pediatric neurologic disorders can be very helpful in the diagnosis. Kayser-Fleischer rings can be seen with a slit lamp well before neurologic findings appear in Wilson's disease. Brushfield spots are frequently seen in newborns with Down's syndrome, while Lisch nodules usually develop in neurofibromatosis during school age years or later. Corneal clouding secondary to glycosaminoglycan accumulation is typical of MPSI (Hurler disease, alpha-iduronidase deficiency) and conjunctival, facial and
antecubital telangiectasias occur in ataxia
telangiectasia, albeit sometimes many years
after onset of ataxia in early childhood.

Reference:
Menkes JH, Sarnat HB, eds. Child
neurology. Philadelphia: Lippincott Williams
& Wilkins, 2000.

Question(s) 294:
Pharmacology/Chemistry

Discussion:
Amitriptyline is 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:
Portenoy RK, Kanner RM. Non-opioid and
adjuvant analgesics. In: Portenoy RK,
Kanner RM, eds. Pain management: theory
and practice. Philadelphia: F. A. Davis,
1996.

Question(s) 295: Clinical Adult

Discussion:
The diagnosis of multiple sclerosis (MS) is
considered to be clinically definite when
there have been two attacks and there is
clinical evidence of two separate lesions.

This evidence must be an abnormality on
physical examination or on paraclinical
testing such as evoked potential or imaging
techniques. Pattern shift visual evoked
responses reveal abnormalities in over 90%
of patients with a history of optic neuritis,
even when visual acuity has returned to
normal. In a well-designed clinical trial,
subcutaneous beta interferon was
demonstrated to reduce the relapse rate of
certain relapsing-remitting patients. No
beneficial effect on ultimate disability was
demonstrated over the two to three year
follow-up period. An expert consensus panel
has concluded that beta interferon may be
helpful for patients with clinically definite MS
who have had at least two acute
exacerbations in the previous two years.

Reference:
1. Bradley WG, Daroff RB, Fenichel GM, et

Randomized comparative study of interferon
beta-1a treatment regimens in MS: the
EVIDENCE Trial. Neurology 2002;59:1496-
1506.

Question(s) 296:
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:
1. Brooks BR. Emerging directions in ALS
therapeutics: palliative therapies at the
advent of the twenty-first century. Clin

2. Mitsumoto H, Chad DA, Pioro EP.

Question(s) 297 - 299: Clinical Pediatrics Discussion:
Typical childhood absence seizures do not have auras or post-ictal confusion. Automatisms can be seen. The usual EEG shows 3-4 Hz generalized spike-and-wave discharges and imaging studies of the brain are normal. The initial drug of choice is ethosuximide or valproate.

Reference:

Question(s) 300:
Pharmacology/Chemistry Discussion:
Nitrous oxide abuse can produce myeloneuropathy that is clinically virtually indistinguishable from that produced by vitamin B12 deficiency. Typical symptoms include paresthesias in the hands and feet, gait ataxia, and leg weakness. A so-called reverse Lhermitte's sign, in which neck flexion induces electrical shock-like sensations traveling from the feet upwards, may occur. In the setting of nitrous oxide abuse the serum B12 level and the Schilling test are almost always normal. Because nitrous oxide interferes with the vitamin B12-dependent conversion of homocysteine to methionine and not with the conversion of methylmalonyl CoA to succinyl coA, only the homocysteine level will be elevated, in contrast to vitamin B12 deficiency where both homocysteine and methylmalonic acid levels will be elevated.

Reference:

Question(s) 301:
Pharmacology/Chemistry Discussion:
Paralytic shellfish poisoning is caused by consumption of bivalve mollusks (mussels, clams, scallops, oysters) contaminated with the dinoflagellate-produced toxin called saxitoxin, which blocks sodium channels and, thus, interferes with impulse conduction in peripheral nerves and muscles. Respiratory failure can result. A number of other marine toxins can produce very similar symptoms. Ciguatera toxin is another dinoflagellate-derived toxin that concentrates in certain large fish. Neurotoxic shellfish poisoning is due to consumption of oysters and clams contaminated with brevotoxin B, yet another dinoflagellate toxin. Domoic acid poisoning occurs following consumption of mussels contaminated with the marine diatom Nitzchia pungens. Neurologic features include confusion and altered states of arousal. Seizures may also occur. Domoic acid is a glutamate agonist. Tetrodotoxin, unlike the previously mentioned toxins, is produced by the puffer fish itself. Saxitoxin poisoning occurs in the Northeast and Northwest USA, ciguatoxin poisoning in Florida and Hawaii, brevotoxin B poisoning in the Gulf of Mexico and Caribbean Sea, domoic acid poisoning in eastern Canada and tetrodotoxin poisoning wherever puffer fish are eaten, primarily in Japan.

Reference:

Question(s) 302 - 303:
Pharmacology/Chemistry Discussion:
Lead has direct effects on porphyrin metabolism, by inhibiting gamma-aminolevulinic acid dehydrase. Lead intoxication produces a motor neuropathy that affects predominantly, but not exclusively, the radial nerve. Associated features include abdominal pain, bluish discoloration of the gums just below the teeth, microcytic hypochromic anemia with basophilic stippling of the red cells, and increased coproporphyrin levels. Arsenic reacts with sulfhydryl groups of proteins and
interferes with several steps of oxidative metabolism in the neuron, producing dying back type axonal degeneration, particularly in myelinated fibers. Thallium ions act interchangeably with potassium in respect to their transport by the Na/K ATPase system. Alopecia, and cranial nerve and autonomic involvement help to distinguish thallium from arsenic intoxication. Organophosphates (OP) inhibit acetylcholinesterase and OP intoxication produces an early syndrome (type I) of excessive muscarinic activation, followed by an intermediate syndrome (type II) of excessive nicotinic activation and muscle weakness. In addition, OP induces a delayed axonal, predominantly motor neuropathy associated with manifestations of central nervous system involvement (ataxia, spasticity); this is attributed to phosphorylation and inhibition of a neuropathy target esterase (NTE) and impairment of axonal transport. Acrylamide impairs axonal transport causing accumulation of neurofilaments and paranoidal swelling mostly in large myelinated axons. This produces a dying back axonopathy, affecting both the peripheral nerves and the central tracts. (eg., gracile tract and dorsal spinocerebellar).

Reference:

Question(s) 304: Pharmacology/Chemistry
Discussion:
Cluster headache will often respond acutely to oxygen inhalation at a flow rate of 8-10 l/Min via face mask. It may be triggered or exacerbated by vasodilating substances such as nitroglycerin, histamine and ethanol. Drugs such as dihydroergotamine may also be helpful in acute headache management. Verapamil, lithium and methysergide can be effective prophylactic agents for cluster headaches. Oral analgesics, such as codeine, are generally ineffective in treating cluster headache, as are anticonvulsants and diuretics.

Reference:

Question(s) 305: Clinical Pediatrics
Discussion:
The gene for Duchenne's and Becker's muscular dystrophy resides in the X chromosome, but approximately one-third of single cases in a family result from new mutations. The clinical onset of Duchenne's dystrophy is around age four years and Becker's dystrophy presents later in the first decade, but there is clinical overlap. Serum CK (creatine kinase) and muscle histology does not differentiate the two disorders. The muscle protein, dystrophin, is absent in Duchenne's and of abnormal size in Becker's providing a tool for differential diagnosis. About 70% of cases with Duchenne's and Becker's dystrophy have a deletion on the X chromosome and the remainder of cases have a point mutation. If a deletion is present in the index case, the carrier status of the mother and other female relatives can be ascertained. If a male with mild Becker's dystrophy has children, all of his daughters will be carriers and his sons will not be affected.

Reference:

Question(s) 306: Pharmacology/Chemistry
Discussion:
Fabry disease is an X-linked recessive disorder caused by deficiency of alpha-galactosidase A. This enzyme deficiency causes accumulation of globotriaosylceramide (GB3) in multiple tissues. The most debilitating symptom is pain, that may occur as attacks of burning pain in the palms and soles, or as a chronic nagging pain in the digits. This is a manifestation of small fiber neuropathy, that may also produce autonomic dysfunction manifest as nausea, diarrhea, vomiting and vascular instability of the extremities. GB3 is also deposited in the skin and its vessels, leading to the formation of angiokeratomas, that may be sparse and restricted to the
scrotum and umbilicus. The eyes, kidneys, heart and cerebral circulation are also involved to a variable extent. Phenytoin and/or carbamazepine in combination are effective in treating the pain of Fabry disease, and the other systemic manifestations may also be treated symptomatically. Enzyme replacement therapy has been shown to be effective for relief of pain and other manifestations.

Reference:


Question(s) 307: Clinical Pediatrics
Discussion:
The spinal muscular atrophies are a group of autosomal recessively inherited disorders, characterized by progressive weakness and atrophy of muscles due to degeneration of anterior horn cells in the spinal cord. The examination reveals weakness and loss of deep tendon reflexes. Serum CK is normal or slightly elevated. EMG and muscle biopsy are consistent with denervation. The majority of children have a homozygous deletion of exon 7 and 8 on chromosome 5. Early onset SMA (1 and 2) have rapidly progressive weakness with severely shortened life-span, while SMA 3 (Kugelberg-Welander disease) is a milder form of the disease with onset after 18 months of age, often with survival into adult life.

Reference:

Question(s) 308: Clinical Pediatrics
Discussion:
Creatine deficiency syndromes are a newly described group of inborn errors of creatine synthesis (arginine:glycine amidinotransferase (AGAT) deficiency and guanidoacacetate methyltransferase (GAMT) deficiency) and creatine transport (creatine transporter (CRTR) deficiency). The common clinical denominator of creatine deficiency syndromes is mental retardation and epilepsy, suggesting the main involvement of cerebral grey matter (grey matter disease). Patients with GAMT deficiency exhibit a more complex clinical phenotype with dystonic hyperkinetic movement disorder and epilepsy that in some cases is unresponsive to pharmacological treatment. The common biochemical denominator of creatine deficiency syndromes is cerebral creatine deficiency which is demonstrated by in vivo proton magnetic resonance spectroscopy. Measurement of guanidoacetate in body fluids may discriminate GAMT (high concentration), AGAT (low concentration) and CRTR (normal concentration). Further biochemical characteristics include changes in creatine and creatinine concentrations in body fluids. GAMT and AGAT deficiency are treatable by oral creatine supplementation, while patients with CRTR deficiency do not respond to this type of treatment.

Reference:

Question(s) 309 - 311: Physiology
Discussion:
The description of acutely evolving symmetric weakness with hyporeactive reflexes in a 45 year old man should suggest the diagnosis of acute inflammatory demyelinating polyneuropathy (AIDP) or Guillain-Barre syndrome. Extremity pain may be a presenting feature. Nerve conduction studies are the best test to perform in this case since they will demonstrate the electrophysiologic signatures of segmental demyelination in the form of motor conduction block and abnormal temporal dispersion of compound muscle action potentials. Early in the course of the disease, prolonged or absent F-wave...
responses may be the only abnormalities indicating proximal conduction block or delay. Needle EMG of affected muscles in this case will likely demonstrate reduced recruitment only. Most of the other choices (fibrillation potentials, polyphasic motor unit potentials, and unstable motor units) indicate axonal loss or reinnervation. CRDs may be seen in either primary nerve or muscle disease and are usually observed in chronic processes. Single fiber EMG in this case would demonstrate normal jitter and fiber density.

Reference:

Question(s) 312 - 314: Clinical Pediatrics Discussion:
Since he did not lose consciousness but his symptoms lasted greater than 15 minutes, this would be considered a grade 2 concussion and he should not be allowed to return to the game and should be reexamined frequently. If no abnormalities, he still should be reevaluated the next day. If this was his first grade 2 concussion, he should be allowed to return to athletics after being symptom free with exercise for one week. If he has had two grade 2 concussions, he should be withheld for two symptom free weeks. Any grade 2 concussion who continues to have symptoms for more than a week should have neuroimaging.

Reference:

Question(s) 315 - 318: Clinical Pediatrics Discussion:
An acutely ill child with over 1000 wbc's in the CSF which are predominantly polymorphonuclear cells should be considered to have bacterial meningitis and treated for this. The most likely etiology in this child would be either s. pneumoniae or influenzae would be very unlikely in an immunized child. Treatment should be cefotaxime or ceftriaxone with vancomycin because many strains of s. pneumonia are resistant to penicillins, cefotaxime and ceftriaxone. Seizures occur in 20-50% of children with meningitis and can be caused by cytokine release/inflammation, vasculitis, infarction, fever, electrolyte imbalance and extra-axial fluid, or often more than one of the preceding. Chronic seizures are unusual after recovery from meningitis.

Reference:

Question(s) 319 - 320: Physiology Discussion:
Hypokalemic periodic paralysis usually presents in the 2nd decade of life. Low K+ in the serum may be found during the paralysis. Respiratory muscles are usually spared. Paralysis is often precipitated by a carbohydrate load the night before or excessive exercise. This is a channelopathy, with the L-type Ca++ channel abnormal.

Reference:

Question(s) 321:
Pharmacology/Chemistry Discussion:
Absence seizures are brief epileptic seizures which present in childhood and adolescence. They are characterised by sudden loss of awareness and an electroencephalogram (EEG) typically shows generalised spike wave discharges at three cycles per second. Automatisms, such as eye fluttering or eye opening, are commonly associated with these episodes. Ethosuximide, valproate and lamotrigine are currently used to treat absence seizures.
Reference:

Question(s) 322:
Pharmacology/Chemistry
Discussion:
The majority of patients with relapse-onset multiple sclerosis (MS) will go on to develop secondary-progressive MS (SPMS) disease, with approximately 50% developing SPMS after 10 years. It remains unknown whether the relapsing and progressive phases of MS differ qualitatively. Immunosuppressive therapies, such as methyleprednisolone, which are capable of reducing or stopping clinical relapses and suppressing MRI activity, generally do not stop disease progression. At present, the only disease-modifying therapies licensed for use in secondary-progressive multiple sclerosis are interferon-beta-1b in Europe and the US, and mitoxantrone in the US.

Reference:

Question(s) 323:
Pharmacology/Chemistry
Discussion:
Mitoxantrone (Novantrone), a synthetic anthracenedione derivative, is an antineoplastic, immunomodulatory agent. Its presumed mechanism of action in patients with multiple sclerosis (MS) is via immunomodulatory mechanisms, although these remain to be fully elucidated. Intravenous mitoxantrone treatment improved neurological disability and delayed progression of MS in patients with worsening relapsing-remitting (RR) [also termed progressive-relapsing (PR) MS] or secondary-progressive (SP) disease. At the recommended dosage, mitoxantrone appears to have a low potential to cause cardiotoxicity, although potential cardiotoxicity limits the total cumulative dose to 140 mg/m(2).

Reference:

Question(s) 324 - 326:
Pharmacology/Chemistry
Discussion:
Malignant hyperthermia is an autosomal-dominant inherited disorder of the skeletal muscle cell characterized by a hypermetabolic response to all commonly used inhalational anaesthetics and depolarizing muscle relaxants. The clinical syndrome includes muscle rigidity, hypercapnia, tachycardia and myoglobinuria as result of increased carbon dioxide production, oxygen consumption and muscle membrane breakdown.

Reference:

Question(s) 327 - 328:
Pharmacology/Chemistry
Discussion:
Valproic acid has been associated with an increased frequency of polycystic ovary syndrome, as has carbamazepine and epilepsy itself. Fetuses exposed to valproate are at increased risk of neural tube defects.

Reference:


Question(s) 329:
Pharmacology/Chemistry
Discussion:
Topiramate is a weak carbonic anhydrase inhibitor used for epilepsy and migraine that can cause hypohidrosis and hyperthermia, in individuals, particularly children, who...
exercise in hot weather. It produces mental slowing in moderate to high doses, and frequently induces weight loss.

Reference:

Question(s) 330: Pharmacology/Chemistry
Discussion:
The Lambert-Eaton myasthenic syndrome presents with proximal weakness, partly repaired by exercise, ataxia and autonomic dysfunction (dry mouth, impotence). It is caused by antibodies directed against pre-synaptic P/Q type calcium channels. About 60% of cases are associated with small cell lung cancer.

Reference:

Question(s) 331: Pharmacology/Chemistry
Discussion:
Fabry disease is an x-linked sphingolipidosis that is characterized by the storage of ceramide trihexoside in vascular endothelium, leading to small fiber neuropathy, stroke, cardiomyopathy and renal impairment. It is also associated with corneal opacities (cornea verticillata) and cataracts. The deficient enzyme, alpha-galactosidase A, can be replaced intravenously (as agalsidase alpha), and has been shown to reverse the principal manifestations of the disease.

Reference:
Desnick RJ. Enzyme replacement therapy for Fabry disease: lessons from two alpha-galactosidase A orphan products and one FDA approval. Expert Opin Biol Ther 2004;4:1167-1176

Question(s) 332 - 333: Pharmacology/Chemistry
Discussion:
Tuberous sclerosis complex (TSC) is an autosomal dominant hamartiosis and hamartomatosis, strongly associated with infantile spasms (IS). Vigabatrin is widely recommended as the first line therapy for IS in TSC, although class I evidence is lacking. Vigabatrin is associated with retinal toxicity, which may be reversible if detected early; screening is recommended, although there are no firm data on appropriate frequency. Somewhat stronger data support the use of ACTH, although it also has significant toxicity related to its steroidogenic actions..

Reference:

Question(s) 334 - 335: Pharmacology/Chemistry
Discussion:
Gingival overgrowth occurs mainly as a result of certain anti-seizure, immunosuppressive, or antihypertensive drug therapies. Phenytoin is well recognized to induce gum hypertrophy, but the frequency is also high with calcium channel blocking agents, which also cause constipation. Drug-induced perturbation of cytokine balance in the gingival fluid is the most likely mechanism. Cathepsin-L inhibition has been suggested as one mechanism, based on experimental animal data.

Reference:


Question(s) 336:
Pharmacology/Chemistry

**Discussion:**
Gabapentin was approved as adjunctive therapy for partial seizures with and without secondary generalizations in 1993, but more than 80% of prescriptions are for off-label uses including neuropathic pain, migraine headache, spasticity, and bipolar disorder. Gabapentin possesses several desirable pharmacokinetic properties: it does not undergo hepatic metabolism, is excreted unchanged in urine, and does not affect the plasma concentrations of other drugs which are primarily metabolized by the liver.

**Reference:**

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**Question(s) 337 - 339: Behavioral**

**Discussion:**
Recent research suggests that Type II diabetes raises the odds of developing Alzheimer's disease by 65%. The speed at which diabetics make perceptual comparisons is one of the most sensitive cognitive tests in diabetics with dementia. The speed of this performance declines by over 40% over time. Treatment of a patient with cognitive decline and type II diabetes calls for aggressive treatment of the disease including the use of insulin.

**Reference:**

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**Question(s) 340: Behavioral**

**Discussion:**
This is the history of a young man with AIDS. He subacutely developed progressive multifocal leukoencephalopathy (PML), a JC papovavirus that affects immunocompromised patients. It has a predilection for the white matter in the bilateral occipito-parietal regions. This is seen on T2 images of an MRI as large hyperintense white matter lesions in the occipitoparietal regions. Visual agnosia and Balint's syndrome are some of the clinical manifestations of the disease.

**Reference:**


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**Question(s) 343: Pathology**

**Discussion:**
The histologic figure shows a distinctive form of keratin composed of clusters of plump keratinocytes that is referred to as "wet" keratin. Among tumors of the central nervous system, this type of keratin is unique to the adamantinomatous craniopharyngioma. Wet keratin nodules frequently undergo dystrophic calcification. In contrast, the keratin seen in epidermoid and dermoid cysts consists of layers of very thin, flat, flaky anucleate squamous cells. Papillary craniopharyngioma arises primarily in adults rather than children and is composed of squamous epithelium that, in contrast to that of the adamantinomatous variant, does not form significant amounts of keratin. The lining of Rathke cleft cysts consists of pseudostratified ciliated columnar epithelium with goblet cells; although focal squamous metaplasia can occur, the distinctive nodules of wet keratin characteristic of adamantinomatous craniopharyngioma are not seen.

**Reference:**
1. Kleihues P, Cavenee WK, eds. Pathology


Question(s) 344: Neuroimaging
Discussion:
Multiple calcified lesions are seen the brain parenchyma, involving both cerebral hemispheres and the cerebellum, primarily at the gray-white junction. This is a typical appearance and location for healed toxoplasmosis lesions, in this case seen in a patient with AIDS. Additional CT characteristics of HIV encephalopathy are also seen in this patient, including generalized atrophy and diffuse, confluent low-density of the white matter. Sarcoidosis can be manifested on imaging studies as either a diffuse meningeal process or multiple parenchymal lesion; however, calcification is not a feature. Sturge-Weber syndrome results in intracranial calcification secondary to pial angiomatosis, with curvilinear calcification following the contour of the cortex, typically in the occipital or parietal-occipital lobes. Calcification secondary to hyperparathyroidism occurs in a symmetric fashion in the basal ganglia, dentate nuclei of the cerebellum, and periventricular white matter. The calcifications in tuberous sclerosis occur in the subependymal tubers lining the ventricles.

Reference:

Question(s) 345: Pathology
Discussion:
The image shows diffuse, widespread myelin loss from a case of X-linked adrenoleukodystrophy.

Reference:

Question(s) 346: Neuroimaging
Discussion:
Axial MRI scans demonstrate the high signal intensity lesion which is of low signal intensity in the T1-weighted image. The lesion is located in the vascular territory of the anterior (superior) branches of the middle cerebral artery. The ability of the MRI scan to detect ischemic lesions is mainly related to changes in water content of developing infarct. Earliest changes are related to cytotoxic edema which is followed by vasogenic edema. Encephalitis is usually not restricted to a vascular territory.

Reference:

Question(s) 347: Neuroimaging
Discussion:
On close inspection, there is a hyperintense rim at the depth and margins of both central or Rolandic sulci. This finding is highly suggestive of ulegryria, or cortical necrosis with gliosis at the depth of the sulci. Ulegryria is one of the varieties of perinatal hypoxic-ischemic encephalopathy. The patient had had clumsy hands for his entire life, and with aging the clumsiness had worsened. There was no other evidence to suggest MELAS. The other options are even less likely.

Reference:

Question(s) 348: Neuroimaging
Discussion:
Metachromatic leukodystrophy (MLD) is the correct response. MLD is characterized by extensive white matter demyelination that
spares the subcortical U-fibers. Confluent periventricular demyelination is present. The anterior white matter is more severely affected. Ceroid lipofuscinosis spares white matter relative to grey matter. Leigh's disease primarily affects basal ganglia structures.Binswanger's disease occurs in the adult population due to chronic cerebrovascular disease.

Reference:

Question(s) 349: Pathology
Discussion:
Degeneration restricted to the dorsal columns is characteristic of tabes dorsalis.

Reference:

Question(s) 350: Pathology
Discussion:
Examination of the cortex shows the characteristic inclusion of myoclonic epilepsy, the Lafora body.

Reference:

Question(s) 351: Clinical Adult
Discussion:
Patients with graves ophthalmopathy often present with diplopia with or without proptosis. MRI shows enlarged extraocular muscles.

Reference:

Question(s) 352: Pathology
Discussion:
The image shows a colloid cyst of the third ventricle.

Reference:

Question(s) 353: Pathology
Discussion:
The tissue section from the resected intraventricular mass showed large cells with abundant eosinophilic cytoplasm. Many also multiple nuclei. Although these features closely resemble those of a number of primary central nervous system tumors, including gemistocytic astrocytoma and ganglion cell tumor, the intraventricular location eliminates these entities, which are intraparenchymal tumors, and points to subependymal giant cell astrocytoma as the correct diagnosis. Of the remaining choices listed, the differential diagnosis of an intraventricular tumor would include subependymoma and choroid plexus papilloma; however, the tumor in the present case does not display the papillary architecture of choroid plexus papilloma or the multilobulation and abundant fibrillar matrix of subependymoma. Subependymal giant cell astrocytomas are commonly associated with tuberous sclerosis and may be the presenting clinical feature, as in the present case.

Reference:

Question(s) 354: Neuroimaging
Discussion:
The non-enhanced CT scan demonstrates midline shift from right to left. On the right side, there is noted extra-axial fluid collection. The subdural hematoma is crescent shaped. The extra-cerebral mass typically locates over the frontal and parietal convexity. There are two different density regions: dependent portion is higher density...
than the brain tissue or the upper part of the hematoma. With unclotted blood the density level may be seen from red blood cells from settling into the dependent region. There is no need for further testing and craniotomy should be performed.

Reference:

Question(s) 355: Pathology
Discussion:
The face demonstrates cyclopia with a fused single eyeball and a superior proboscis. Cyclopia occurs with midline cleavage defects and clefting of the lip or the palate is usually also present. Alobar holoprosencephaly is also part of this spectrum of midline cleavage abnormalities with a single, globular hemisphere, a single ventricle and fused basal ganglia.

Reference:

Question(s) 356: Pathology
Discussion:
Bilateral deep gray matter hemorrhage is seen with straight sinus venous thrombosis. The lesion may be either bilaterally symmetrical or slightly asymmetrical, as in the illustrated case. Venous thrombosis is commonly associated with hypercoagulable states and severe dehydration. Fat embolism typically affects the white matter not the gray matter. Metastases usually do not show the symmetry seen here.

Reference:

Question(s) 357: Pathology
Discussion:
The location of this hemorrhage, with its epicenter in the basal ganglia, is the most common location for hypertensive hemorrhages. Over 60% of hypertensive hemorrhages are located here; other less common places include the pons and cerebellum.

Reference:

Question(s) 358: Physiology
Discussion:
Benign Rolandic epilepsy is characterized by nocturnal focal or generalized seizures and bilateral independent centrotemporal spikes.

Reference:

Question(s) 359: Pathology
Discussion:
The photograph shows a biphasic cellular population consisting of small reactive lymphocytes and large neoplastic germ cells, which is characteristic of germinoma - the most common pineal region tumor. Pineocytomas, in contrast, are composed of small mature pineocytes that form large rosettes with fibrillary cores. Pineoblastomas are densely cellular primitive neuroectodermal tumors that often form Homer Wright (medulloblastoma-type) rosettes and occasionally fleurettes. Pineal astrocytomas show prominent eosinophilic cytoplasmic processes. Regarding terminology, the designation "pinealoma" is an inaccurate, arcane and imprecise historical relic that should not be used; it dates to a time before a distinction was made between germinoma and pineoblastoma.

Reference:
Question(s) 360: Neuroimaging
Discussion:
Sagittal T1 and axial T2-weighted MRI show a hyperintense rounded mass at the anterosuperior third ventricle consistent with a colloid cyst. Approximately 60% of colloid cysts exhibit short T1 and variable shortening of T2 relaxation times. This MR appearance may be related to heavy protein or mucin content. The signal of many colloid cysts allows differentiation from most gliomas (long T1 and T2 relaxation times), aneurysms (flow-related signal void) and meningiomas (isointense to the brain in T1 and T2). Third ventricle craniopharyngiomas are rare. They may exhibit short T1 and long T2 relaxation times.

Reference:

Question(s) 361: Pathology
Discussion:
A coronal section of the brain at autopsy shows mamillary bodies that are congested and discolored, which is a characteristic finding in thiamine deficiency manifesting as Wernicke's encephalopathy.

Reference:

Question(s) 362: Neuroimaging
Discussion:
Sagittal T1-weighted images demonstrate a loculated fluid collection in the posterior epidural space, extending for several levels in the mid-thoracic spine. The collection causes significant compression and anterior displacement of the spinal cord. Note on both sagittal and axial images that the subarachnoid space is also compressed, placing this in the epidural, not intradural space (excluding meningioma). This is a typical appearance of a loculated, encapsulated epidural abscess. A ten-day-old hematoma should be hyperintense on T1-weighted images. Epidural metastases and myeloma are of soft tissue signal, generally nearly isointense to neural tissue and usually are anteriorly located, extending posteriorly from vertebral body lesions.

Reference:

Question(s) 363: Anatomy
Discussion:
The photograph shows a cross section of the brain stem at the superior pontine level. The locus ceruleus is identified by arrows bilaterally. The locus ceruleus is the major source of norepinephrine projections in the central nervous system.

Reference:

Question(s) 364: Physiology
Discussion:
Nerve conduction studies with temporal dispersion, conduction block, and slow conduction are seen in chronic inflammatory demyelinating polyneuropathy. In Charcot-Marie-Tooth disease, there are slow latencies and nerve conduction velocities, but no temporal dispersion or block. In amyotrophic lateral sclerosis, nerve conduction studies may be normal or with decreased compound muscle action potential amplitudes. Nerve conduction studies are normal in fascioscapulohumeral dystrophy and myasthenia gravis.

Reference:
Question(s) 365: Neuroimaging
Discussion:
Noncontrast axial CT reveals a lens shaped well localized hyperdense extraaxial lesion with some surrounding edema. Findings are consistent with epidural hematoma. Epidural hematoma is typically a localized hyperdense lesion because dura is adherent to the skull. Acute subdural hematoma is crescent shaped and more widespread. Chronic subdural has low density on CT.

Reference:

Question(s) 366: Neuroimaging
Discussion:
The signal of the deformity producing tissue is the same as in cortex, thus this lesion represents heterotopic gray matter. Neoplasm would have had a high signal. The lesion is not limited to the basal ganglia, thus eliminating a hemorrhage. There is no sign of chronic infarction. The study does not include the hippocampi. Therefore, mesial temporal sclerosis cannot be diagnosed.

Reference:

Question(s) 367: Neuroimaging
Discussion:
Multiple sclerosis plaques are shown. The thoracic sensory level relates to plaques in the thoracic cord.

Reference:

Question(s) 368: Pathology
Discussion:
In the figure, myelin-stained cross sections of the cervical and lumbar spinal cord show degeneration of the corticospinal tracts with sparing of the posterior columns and spinocerebellar tracts. These findings are most compatible with amyotrophic lateral sclerosis.

Reference:

Question(s) 369: Physiology
Discussion:
In a sensory nerve action potential recording, the initial positivity corresponds to the action potential passing beneath the active recording electrode, and is used to measure the onset latency for terminal conduction velocity measures.

Reference:

Question(s) 370: Pathology
Discussion:
The dysembryoplastic neuroepithelial tumor (DNT) is composed of neurons and oligodendroglial-like cells clustered within intracortical nodules. A prominent myxoid stroma is typically present within the tumor nodules. This indolent tumor is frequently associated with cortical migration abnormalities. Surgery is curative.

Reference:

Question(s) 371: Pathology
Discussion:
Bilateral necrosis of the globus pallidus is often due to carbon monoxide intoxication.

Reference:
Question(s) 372: Neuroimaging
Discussion:
T2-weighted images show hypointense signal changes on the pial surface of the brain stem, temporal lobes and cerebellar vermis. Susceptibility effect with T2 shortening and hypointensity is due to hemosiderin from prior repeated subarachnoid hemorrhage. The etiology of the hemorrhagic process was a cervical neoplasm.

Reference:

Question(s) 373: Neuroimaging
Discussion:
The correct answer is intracranial hypotension secondary to a persistent cerebrospinal fluid leak, resulting in prominent, abnormal meningeal enhancement. The clinical findings are not consistent with acute pyogenic meningitis. Tuberculosis meningitis is usually a basilar meningitis.

Reference:

Question(s) 374: Pathology
Discussion:
Germinal matrix hemorrhage is a common complication of premature birth. The germinal matrix, located in the walls of the lateral ventricles, is a cellular area which is quite vascular and delicate. These hemorrhages are graded clinically into four different subtypes: Grade I (localized to the germinal matrix only), Grade II (ruptured into the ventricle without ventricular dilatation), Grade III (ruptured into the ventricle with ventricular dilatation), and Grade IV (ruptured into the ventricular system and also into the cerebral parenchyma). Clinical outcome in neonates with the Grade III and IV hemorrhages is much worse that those with Grades I and II.

Reference:

Question(s) 375: Neuroimaging
Discussion:
On histology, this lesion was a glioblastoma multiforme. An astrocytoma is unlikely to show enhancement. On the absence of risk factors and with a normal physical examination, the probability of toxoplasmosis or an abscess are lower than for a glioblastoma, although either lesion can have a similar morphology. Demyelinating lesions can show a ring of enhancement, but it is more frequently open than closed, as with this lesion.

Reference:

Question(s) 376: Pathology
Discussion:
The characteristic inclusion body of rabies (Negri body) is classically seen in large pyramidal neurons of the hippocampus and is located in the cytoplasm. In contrast, Herpes simplex and cytomegalovirus produce Cowdry A intranuclear inclusions. Poliovirus produces Cowdry B intranuclear inclusions during the acute phase (not seen in post-polio syndrome). Viral inclusions are not present in eastern equine encephalitis and many other viral encephalitides in which only the non-specific findings of perivascular lymphocytic infiltrates, microglial nodules
and granular ependymitis are seen.

Reference:

Question(s) 377: Neuroimaging
Discussion:
The uniform filling of this large vascular anomaly and its position identify it as a vein of Galen aneurysm which is a congenital anomaly.

Reference:

Question(s) 378: Neuroimaging
Discussion:
This abnormality is clearly a herniated disc. The herniated material is contiguous with disc material (non-sequestered).

Reference:

Question(s) 379: Neuroimaging
Discussion:
The most likely cause is a chiasmal glioma. The lesion clearly does not arise from the sella. It contains no cystic region or area of signal void that would be consistent with a craniopharyngioma.

Reference:

Question(s) 380: Pathology
Discussion:
The lesion is a porencephalic cyst. The lesion is partially covered by arachnoid and extends from the subarachnoid space to the ventricular system. The lesion is typically smooth walled and surrounded by abnormal radially oriented gyri reflecting its development early in life.

Reference:

Question(s) 381: Pathology
Discussion:
The photograph of a carotid endarterectomy specimen shows multiple dark irregular brown ulcerated plaques characteristic of complicated atherosclerosis. In simple (uncomplicated) atherosclerosis there are no plaque ulcerations.

Reference:

Question(s) 382: Pathology
Discussion:
Neuritic plaques consist of an amyloid core and dystrophic neurites, with reactive astrocytes and microglia. They are a classic finding in Alzheimer's disease; in the commonly used CERAD grading system, neuritic plaque counts are used as histologic criteria for making the diagnosis of Alzheimer's disease.

Reference:

Question(s) 383: Neuroimaging
Discussion:
Round soft tissue mass within the spinal canal is located in the intradural, extramedullary compartment. This is
evidenced by widening of the subarachnoid space above and below the mass, and compression rather than enlargement of the spinal cord. Of the choices mentioned, meningioma is the only consistent intradural extramedullary mass; ependymoma, astrocytoma and hemangioblastoma are intramedullary lesions. Lipomas are usually hyperintense on T1 weighted images.

Reference:

Question(s) 384: Neuroimaging
Discussion:
The most likely diagnosis is toxoplasmosis. On MRI, toxoplasmosis lesions are most commonly multiple and are located in the deep central grey nuclei, or lobar grey-white junction. Other common locations include the posterior fossa, cerebral cortex, and paraventricular white matter. Toxoplasmosis lesions appear isointense to hypointense on T1-weighted images and mildly to markedly hyperintense on T2-weighted images, exerting marked mass effect. Copious edema is often noted which is out-of-proportion to the lesion size. After contrast administration, ring-like or nodular enhancement patterns are most commonly noted. Cytomegalovirus encephalitis and progressive multifocal leukoencephalopathy are usually not enhancing in this manner. Toxoplasmosis may be difficult to distinguish from primary CNS lymphoma. Nocardia and glioblastoma are much less likely in this clinical setting.

Reference:

Question(s) 385: Anatomy
Discussion:
A lesion of the fourth cranial nerve results in superior oblique weakness with resultant diplopia.

Reference:


Question(s) 386: Neuroimaging
Discussion:
Both images demonstrate the missing vermis thus the 4th ventricle connects with the cisterna magna, the hallmark of Dandy-Walker malformation. The posterior fossa is enlarged.

Reference:

Question(s) 387: Clinical Adult
Discussion:
When an optic nerve lesion extends posteriorly to involve the anterior chasm, the earliest indication is the occurrence of a temporal field defect in the contralateral eye (usually, but not always in the upper temporal field).

Reference:

Question(s) 388: Neuroimaging
Discussion:
T1-weighted MRI shows a mixed signal destructive mass involving the clivus and nasopharynx extending to the spinal canal. The lesion is consistent with a clivus chordoma. The ventral pontine surface is compressed. Primary tumors, such as chordomas and cartilaginous tumors of the
skull base are rare. Chordomas arise from remnants of the embryonic notochord which is a mesodermal derivative. Cranial chordomas are most common in the third and fourth decades of life and males are affected more often than females. Cartilaginous tumors occur between 20 and 60 years of age. They are extradural and over half arise in or adjacent to the body of the sphenoid bone. Radiologically the normal, high signal marrow cavity of the clivus is replaced by lower signal intensity tumor. Large areas of calcification may be seen as void-phenomenon. Radiographically the distinction between chordomas, chondrosarcomas and chondromas may be impossible. Meningiomas are isointense to the brain before contrast. This is an extra pontine lesion; the brainstem is not involved. The pituitary gland is normal. Parapharyngeal abscesses are usually smoothly contoured.

Reference:


Question(s) 389: Neuroimaging
Discussion:
Axial T2-weighted images 6 months apart demonstrates development of abnormal signal in the globus pallidus and white matter disease adjacent to the trigone of the right lateral ventricle. Cerebral atrophy and enlarged CSF spaces are also seen to develop. The ventricular and subarachnoid space each have increased, consistent with parenchymal tissue loss. The findings are consistent with anoxic injury occurring shortly after the first scan. The globus pallidus is especially vulnerable to anoxic injury. Differential diagnosis includes chronic hypoglycemic injury, carbon monoxide poisoning, AIDS, Wilson's disease and menigitis. Gliomatosis cerebri would result in multiple T2 hyperintense parenchymal lesions and mass effect. Krabbe's usually presents in infancy. Both Canavan's and Krabbe's are expected to cause widespread white matter lesions.

Reference:

Question(s) 390: Pathology
Discussion:
The graphic illustrates bilateral, nearly symmetric, parasagittal, cortical and basal ganglia venous infarctions due to thrombosis of the superior sagittal sinus and deep cerebral veins.

Reference:

Question(s) 391: Neuroimaging
Discussion:
The biopsy of this lesion showed demyelination. It corresponds to an unusual case of Balo's concentric sclerosis. The open ring seen of enhancement in the first study is more frequent with demyelinating lesions than with tumors or infections.

Reference:


Question(s) 392: Neuroimaging
Discussion:
A large cystic, rim-enhancing mass is present with a more solid component along the anterior margin. Porencephaly would not
enhance, nor would a lipoma. Abscess would be more homogeneously enhancing around the rim, and produce more prominent hemiparesis. Hemangioblastomas typically occur in the posterior fossa. The best answer is cystic astrocytoma.

Reference:

Question(s) 393: Pathology
Discussion:
The tumor illustrated is a low grade oligodendroglioma; the calcifications and "fried-egg cells" are characteristic of oligodendroglioma. These tumors typically present in patients in their 30s or 40s. Due to the high tendency of these tumors to invade the overlying cortex, seizures (often of one to five years duration) are one of the most frequent clinical presentations. A dural-based mass in a 60-year-old woman is most likely to be a meningioma. The prototypical profile of a smoker with a lung mass is metastatic lung carcinoma and the male with a posterior fossa mass is more likely to be a patient with medulloblastoma.

Reference:

Question(s) 394: Neuroimaging
Discussion:
The tumor is located in the region of the sella turcica, ruling out glioblastoma, ependymoma and schwannoma. Meningiomas seldom are this symmetrical. Pituitary adenoma is the most likely diagnosis.

Reference:

Question(s) 395: Neuroimaging
Discussion:
The most likely diagnosis is Leigh’s disease. The abnormal areas are seen as high signal intensity within the brain stem and basal ganglia. They are symmetric. Such abnormalities are found in patients with metabolic acidosis and elevated lactate including Leigh’s disease. Herpes usually involves the medial temporal lobe, insular cortex, and inferior frontal lobes, areas that are not involved in this case. There is also no brain swelling or mass effect, findings that are common in herpes. Carbon monoxide poisoning involves principally the globus pallidus and does not involve the brain stem. Infarctions in sickle cell disease do not have such symmetry and are uncommon in the brainstem.

Reference:

Question(s) 396: Neuroimaging
Discussion:
Right carotid arteriogram (AP and lateral view, arterial phase). The arteriogram demonstrates a "string of beads" in the midportion of the internal carotid artery. The appearance is quite characteristic for fibromuscular dysplasia (FMD). FMD is a nonatheromatous angiopathy of unknown etiology. Characteristically FMD extend from C1-2 level to the entrance of the internal carotid artery into petrous carotid canal. FMD is bilateral in 65% of cases. Nearly one third is of the cases are associated with intracranial aneurysms.

Reference:

Question(s) 397: Neuroimaging
Discussion:
Both the location of the lesion and the presence of precocious puberty favor a hamartoma in the tuber cinereum. In the sagittal image the intact pituitary gland can be seen in the sella, below the suprasellar
cistern.

Reference:

Question(s) 398: Pathology
Discussion:
The cells shown contain neurofibrillary tangles - an intraneuronal intracytoplasmic inclusion composed of paired helical filaments. The section has been stained with a silver stain. Neurofibrillary tangles are seen in Alzheimer disease, as well as a variety of other conditions including Pick disease, progressive supranuclear palsy, Down syndrome, Parkinson-dementia complex of Guam, and post-encephalitic Parkinsonism.

Reference:

Question(s) 399: Neuroimaging
Discussion:
Axial T2 weighted image shows a diffuse high signal intensity throughout all of the white matter that is shown within the semicentrum ovale. The sulci are enlarged for the patient's age. Thus the patient has a diffuse process involving the white matter that does not produce mass effect. This is not normal, not typical of demyelinating disease in the sense of multiple sclerosis, making the subacute form of AIDS the most likely diagnosis. Pick's disease would involve more of the frontal and temporal lobes, while a glioblastoma would be a focal mass lesion.

Reference:
Zimmerman RA, Bilaniuk LT, Sze G.


Question(s) 400: Pathology
Discussion:
The photomicrographs show classic perivascular pseudorosettes characteristic of ependymoma.

Reference:

Question(s) 401: Neuroimaging
Discussion:
In the sagittal T1-weighted image the normal flow void (low signal) is replaced by intraluminal high signal related to thrombosed superior sagittal sinus. The signal intensity of the thrombus over time has the same evolution pattern as intracerebral hematomas.

Reference:

Question(s) 402: Pathology
Discussion:
The photomicrograph shows acutely branching septate hyphae which most likely represent an opportunistic Aspergillosis infection. Patients with Aspergillosis frequently have neutropenia and are receiving immunosuppressive therapy for neoplasms. The organism initially infects the lung and then spreads hematogenously to involve other organs. The brain is second only to the lung as a site of visceral involvement, and frequently manifests as hemorrhagic infarctions.

Reference:
Question(s) 403: Pathology
Discussion:
The LFB-PAS sections demonstrate numerous enlarged, round axonal swellings which stain positively with the silver stain. This is the histologic picture of axonal spheroids, the finding in diffuse axonal injury. Also known as "shear injury," patients with this condition are rendered immediately unconscious with trauma, and subsequently have a course of chronic "closed head injury."

Reference:

Question(s) 404: Pathology
Discussion:
The brain shows downward herniation of the cerebellar tonsils. This is consistent with a Chiari I malformation.

Reference:


Question(s) 405: Neuroimaging
Discussion:
The middle cerebral artery (MCA) is occluded after its first proximal branches. The abrupt cutoff of the MCA trunk is seen on both anterior-posterior and lateral angiograms; absence of the MCA trunk, normally seen in the sylvian fissure, is best appreciated on a lateral view.

Reference:

Question(s) 406: Pathology
Discussion:
The image shows numerous small gyri over the surface of the brain. The correct diagnostic term is polymicrogyria.

Reference:

Question(s) 407: Pathology
Discussion:
Toxoplasmosis is a parasitic infection seen most commonly in immunosuppressed patients. Both tachyzoites and cysts containing bradyzoites may be present.

Reference:

Question(s) 408: Neuroimaging
Discussion:
The dilated third ventricle projects superiorly between bodies of the lateral ventricles. A corpus callosum is not seen, therefore this is agenesis of the corpus callosum.

Reference:

Question(s) 409: Pathology
Discussion:
There is severe multicystic encephalomalacia due to intrauterine hypoxia/ischemia. The formation of cysts reflects cavitation which is the end stage of maturation of infarcts. The infarcts must be weeks to months old, and therefore, reflect intrauterine events.
**Question(s) 410: Neuroimaging**

**Discussion:**
The scan shows sequelae of chronic herpes simplex virus encephalitis. Encephalomalacia is noted in both temporal lobes, more prominent on the left with cystic change and tissue loss. The lesions are too extensive for mesial temporal sclerosis. The lesions cross vascular boundaries and are in an unusual location for strokes. Epidermoid and arachnoid cysts are usually unilateral and not associated with surrounding parenchymal hyperintensities.

**Reference:**

**Question(s) 411: Pathology**

**Discussion:**
The myelin-stained axial section of the brain stem at the level of the inferior olivary nuclei shows infarction (with cavitation and surrounding pallor on this myelin stain) of a wedge-shaped area of the dorsolateral portion of the medulla in the distribution of the posterior inferior cerebellar artery. This usually results from occlusion of the vertebral artery.

**Reference:**

**Question(s) 412: Neuroimaging**

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

**Reference:**

**Question(s) 413: Neuroimaging**

**Discussion:**
There is symmetric high signal intensity involvement of the putamen and thalami bilaterally. The globus pallidus is also involved but not exclusively as it is in many patients with carbon monoxide poisoning. The heads of the caudate and nuclei appear normal and there is no significant overall atrophy of the brain. These are findings that tend to exclude Huntington’s, while the high signal intensity within the globus pallidus and putamen is atypical for Parkinson’s disease. Gliomatosis cerebri, an infiltrating astrocytoma of the white matter is excluded by the fact that the disease process spares the white matter where the tumor occurs. The correct response is Wilson’s disease.

**Reference:**

**Question(s) 414: Physiology**

**Discussion:**
The illustration shows sleep spindles which are thought to be generated by the reticular thalamic nucleus.

**Reference:**
Shaul N. The fundamental neural mechanisms of electroencephalography.
Question(s) 415: Pathology
Discussion:
Centronuclear myopathy is characterized by the presence of centrally located nuclei, often accompanied by a perinuclear "halo". The muscle may show type I myofiber predominance.

Reference:

Question(s) 416: Pathology
Discussion:
The biopsy shows a severe vacuolar myopathy due to extensive glycogen storage. These findings are consistent with Pompe's disease (Glycogen storage disease type II), which is caused by a genetic deficiency of acid-alpha-glucosidase (acid maltase). In its most severe form, the disease presents in infancy with cardiomegaly, hepatomegaly, progressive muscle weakness, macroGLOSSia, and hypotonia ("floppy baby"). Of the choices provided, Pompe's disease is the only disorder that presents findings of vacuolar myopathy on a muscle biopsy. Pathologic findings of Haltia-Santavuori disease (infantile neuronal ceroid lipofuscinosis), Alexander's disease primarily affect the brain. Krabbe's disease is a leukodystrophy affecting CNS white matter. Kuf's disease is an adult form of neuronal ceroid lipofuscinosis.

Reference:

Question(s) 417: Neuroimaging
Discussion:
The MRI shows bilateral medial temporal cortical dysplasia. There is no dysplastic tissue in the other regions mentioned in the question.

Reference:

Question(s) 418: Neuroimaging
Discussion:
The scans show schizencephaly with septo-optic dysplasia. Porencephalic cyst would be lined with white matter, whereas the cavity in schizencephaly is lined by heterotopic grey matter. Lissencephaly refers to brains with absent or extremely poor sulcation. Holoprosencephaly results from the failure of lateral cleavage into distinct cerebral hemispheres and failure of transverse cleavage into diencephalon and telencephalon.

Reference:

Question(s) 419: Pathology
Discussion:
The photograph shows remote bilateral frontal contusions with a slight orange tinge secondary to the presence of residual hemosiderin-laden macrophages. The olfactory nerves are disrupted, which undoubtedly would have led to anosmia.

Reference:

Question(s) 420: Neuroimaging
Discussion:
Contrast-enhanced CT shows a densely enhancing mass projecting into the upper margin of the sella turcica. This could be either an intra- or suprasellar lesion based on the CT findings. As such it could be a pituitary adenoma, meningioma, aneurysm, or possibly another lesion. The sagittal T1 weighted MR image shows a hypointense flow void, such as is found in the lumen of
an aneurysm. The only other thing that could mimic this on the MR scan would be a collection of air or cortical bone. The combination of the CT and the MR indicate an aneurysm. The pituitary gland can be seen below the aneurysm within the sella.

Reference:

Question(s) 421: Pathology
Discussion:
An anterior cavum septum pellucidum lies between the transverse fibers of the genu of the corpus callosum and the anterior commissure. Constantly present in the human fetus, it tends to become obliterated near term. In adults, the cavity is found in about 20% of brains studied at autopsy. It has no proven clinical significance.

Reference:

Question(s) 422: Pathology
Discussion:
Multiple periventricular areas of demyelination are characteristically seen in multiple sclerosis. These plaques have a grey hue because of the loss of myelin that normally confers a glistening white appearance to the white matter.

Reference:

Question(s) 423: Pathology
Discussion:
The biopsy of this cystic mass showed layers of flattened, anucleate squamous cells. This type of flaky keratin is characteristic of epidermoid cysts. In contrast, the cyst contents of adamantinomatous craniopharyngiomas typically consist of nodular clusters of plump necrotic keratinocytes referred to as wet keratin. The lining of Rathke cleft cysts, colloid cysts, and neurenteric cysts consists of ciliated pseudostratified columnar epithelium with scattered goblet cells. These cysts have watery or mucinous contents, but no keratin.

Reference:

Question(s) 424: Neuroimaging
Discussion:
The histology of this temporal lobe lesion proved it to be a glioblastoma multiforme. An astrocytoma is unlikely to show enhancement. A choroid-plexus papilloma is much more rare in this age group that either of the previous tumors, and this tumor is not in the ventricle, but around the ventricle. Demyelinating lesions can show an open ring of enhancement, as with this lesion, but this sign is not reliable in post-surgical cases. Incisural sclerosis does not show enhancement.

Reference:


Question(s) 425: Neuroimaging
Discussion:
Sagittal T1 and axial proton density images show abnormal hyperintensity filling the vein of Galen and straight sinus, instead of the normal flow void expected. This is secondary to thrombus in these structures, and have resulted in hemorrhagic venous
infarction in the thalami. Mass effect from the infarctions has compressed the third ventricle, resulting in obstructive hydrocephalus of the lateral ventricles. The axial image shows prominent basal veins of Rosenthal (seen on either side of the vein of Galen as flow void), likely providing collateral venous drainage. The pineal gland is normal in appearance, but displaced inferiorly by the thrombus and mass effect.

Reference:

Question(s) 426: Neuroimaging
Discussion:
The scan shows Canavan's disease. The abnormality is the high signal intensity in the white matter that goes from the periventricular region to the cortex in a diffuse fashion, involving all of the white matter without sparing subcortical U-fibers. Periventricular leukomalacia is in the periventricular region in the frontal parietal region. Metachromatic leukodystrophy does not extend out into the subcortical U-fibers, while adrenoleukodystrophy favors the occipital and parietal regions sparing the frontal lobes in all but a small percentage of cases. Adrenoleukodystrophy also tends to spare the subcortical U-fibers. Krabbe's disease has significant atrophy.

Reference:

Question(s) 427: Pathology
Discussion:
The illustration depicts myofibrils with an increased number of subsarcolemmal mitochondria, imparting a red coloration to the muscle fiber: the "ragged red fiber." Although ragged red fibers can occasionally be seen in other conditions, they are most commonly observed in the mitochondrial myopathies. Electron microscopic examination in such cases confirms the presence of abnormal or too-numerous mitochondria. Of the choices offered in the question, only Kearns-Sayre myopathy is a mitochondrial disease. Werdnig-Hoffman is a motor neuron disease; Duchenne's is an X-linked-inherited myopathy involving abnormalities of dystrophin; central core myopathy is a genetic condition where muscle biopsy demonstrates the presence of core-structures in the myofibrils; and dermatomyositis is an autoimmune inflammatory myopathy.

Reference:
Question(s) 430: Neuroimaging
Discussion:
The images show a cystic cerebellar hemispheric mass with an enhancing mural nodule. T2-weighted images suggest the presence of small vessels within the mural nodule as hypointense flow voids. The mass is intra-axial and infratentorial. The differential diagnosis includes ganglioglioma, medulloblastoma, cystic astrocytoma, and abscess. Demyelinating plaques may present as neoplasms, but this is unusual. The finding is most consistent with a cerebellar cystic hemangioblastoma.

Reference:

Question(s) 431: Physiology
Discussion:
Myokymic discharges can be seen as myokymia on the skin surface, are rhythmic and may be seen as singlets or multiplets. They are sometimes associated with potassium channel antibodies.

Reference:

Question(s) 432: Neuroimaging
Discussion:
The axial T1 weighted image shows an extra-axial hyperintense lesion that lies between the hypointense inner table of the skull and the isointense brain parenchyma. The sulci are displaced and the cortex appears intact. There is a corresponding displacement of the ventricular system. The high signal intensity (T1 shortening) is consistent with methemoglobin-containing subacute clot in the subdural space. Epidural hematoma is excluded because the blood collection crosses suture lines. Subdural hygroma and empyema are excluded because the signal is not consistent with CSF or pus. En-plaque meningioma is usually iso to hypointense on noncontrast T1-weighted images, although it could present a similar appearance on postcontrast images.

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

Reference:

Question(s) 442 - 444: Pathology
Discussion:
The photomicrograph illustrates the classic spongiform changes of Creutzfeldt-Jakob disease. These changes are visible only under the microscope, not grossly. The cerebrospinal fluid is usually normal. The gross brain characteristically shows shows only a mild degree of overall atrophy or no atrophy at all. The hippocampal formation is usually well preserved even in cases with severe overall brain atrophy, a feature distinct from Alzheimer's disease which shows early and severe hippocampal involvement.

Reference:

Question(s) 445 - 447: Pathology
Discussion:
The photomicrograph illustrates severe atrophy of the cord with myelin loss in the posterior columns and dorsal spinocerebellar tracts. These changes are classic for Friedreich's Ataxia. Systemic findings include cardiomyopathy and adult onset diabetes mellitus. Thickening of the skull to 2 to 3 times normal is characteristic for dentatorubral-pallidoluysian atrophy. Gynecomastia and testicular atrophy are seen in spinal and bulbar muscular atrophy. The disease shows autosomal recessive inheritance and in more than 95% of cases is due to GAA repeat expansion in the frataxin gene.

Reference:

Question(s) 448 - 451: Pathology
Discussion:
Progressive supranuclear palsy presents with falls, postural instability, retrocolis, bradykinesia, pseudobulbar palsy and dementia. At autopsy, globoid neurofibrillary tangles are characteristically found in substantia nigra, brainstem tegmentum, putamen, and select cortical areas. These are immunoreactive for tau. Also seen are tau-positive astrocytic inclusions, so called tufted astrocytes. Tufted astrocytes are almost exclusively seen in PSP and can be found in striatum, thalamus, subthalamic nucleus and precentral gyrus and are rare to absent in a similar disorder, corticobasal degeneration.

Reference:
Discussion:
Kernicterus, meaning yellow nuclei, is seen in infants who die with severe neonatal jaundice. Bilirubin is a breakdown product of blood that is normally conjugated in the liver to form bilirubin diglucuronide, which then is carried into bile and the intestine and converted to urobilinogens by gut flora. Neonates have more unconjugated bilirubin since they initially lack the enzymes necessary to convert the unconjugated bilirubin. The unconjugated bilirubin passes into selected nuclei, especially globus pallidus, subthalamus and Ammon's horn in term infants. How precisely this causes cellular injury is uncertain.

Reference:

Question(s) 455 - 457: Pathology

Discussion:
Wilson's disease is an autosomal recessive condition due to derangement of copper metabolism. The cornea shows copper deposition (Kayser-Fleischer rings) and the brain shows cavitation and atrophy of the caudate and putamen in rapidly progressive forms. Alzheimer type II, metabolic astrocytes are abundant.

Reference:

Question(s) 458: Neuroimaging

Discussion:
This is a typical presentation for CADASIL syndrome, in which an autosomal dominant pattern of cerebrovascular disease and associated migraine headaches are present. The anterior temporal damage apparent bilaterally in Figure A is strongly suggestive of the CADASIL syndrome, which was proven through appropriate genetic testing in this case. The anterior temporal regions are unlikely to be affected in this manner by any of the other disorders. Multiple sclerosis is consistent with Figure B despite the presence of some gray matter involvement, but the anterior temporal lesions are uncharacteristic and multiple sclerosis lacks the strong association with migraine that is part of the CADASIL syndrome. Nonspecific MRI abnormalities associated with migraine are typically small scattered foci of nonspecific gliosis rather than the large confluent areas of damage shown in these images. AlthoughBinswanger's disease is entirely consistent with the images shown, the CADASIL syndrome is still a better choice due to the family history, the associated migraines, and the bitemporal damage, as already discussed.

Reference:

Question(s) 459 - 460: Neuroimaging

Discussion:
There is increasing evidence supporting early stroke treatment with TPA, even when early ischemic changes are present throughout more than one third of the middle cerebral territory. The issue remains controversial, and D is therefore the best answer. TPA and heparin must not be started simultaneously, due to the high risk of severe brain hemorrhage. The presence of edema (which accounts for the CT changes) is not conclusive for infarction, as ischemic tissue may swell. DWI findings in the left hemisphere are those of a rim of hyperintensity around the rim of a chronic infarction (4-5 months previously). The absence of ADC abnormalities indicates that this rim of high signal is likely to represent T2 shine through. Although the bihemispheric strokes are suggestive of a cardioembolic mechanism, the strokes are not about the same age. DWI imaging abnormalities may persist up to 57 days post-ictus, and are therefore not expected to resolve within 10 days in this patient. Therefore, the best answer is that DWI hyperintensity may persist for two months following stroke. Although DWI abnormalities typically do correlate with the ischemic core, indicative of irreversible and permanent damage, DWI signal resolution with tissue recovery is commonly seen. This is particularly true in subacute infarction, in areas where T2 shine through accounts for 70%
some of the DWI abnormality. The ischemic penumbra is best defined as the mismatch between the brain tissue that is being hypoperfused, but where DWI abnormalities are not evident. This typically occurs outside the ischemic core that is best represented by the zone of DWI abnormality. Cytotoxic edema, rather than vasogenic edema, accounts for DWI abnormalities, and in fact DWI imaging is helpful in differentiating between these two types of edema. There is a definite tendency for the area of DWI abnormality to become the ischemic core, so the statement that there is no relationship is false.

Reference:
2. Von Kummer R. Early major ischemic changes on computed tomography should preclude use of tissue plasminogen activator. Stroke. 2003;34:820-821.
Lyden P. Early major ischemic changes on computed tomography should not preclude use of tissue plasminogen activator. Stroke. 2003;34:821-822.

Question(s) 461 - 463: Neuroimaging
Discussion:
Lipomas are homogeneous in signal intensity (unlike a dermoid tumor), and do not contain calcium deposits (like dural dysplasia typically does), teeth or nails (like a teratoma), or rich vascularity (like a menigioma). Lipomatous transformation of meningioma produces tumors that are also heterogeneous in comparison to lipomas. The imaging characteristics of this lesion are therefore most consistent with lipoma. Cranial nerve IV emerges from the dorsal surface of the brainstem in close proximity to the lesion shown. The remaining cranial nerves are inferior to this location (e.g., the cerebellopontine angle for cranial nerves VII and VIII) or lower pons. Thus, this lipoma, situated in the quadrigeminal plate cistern, is most likely to affect cranial nerve IV. A lipoma is considered a hamartoma, and is not a true neoplasm. There is no sex preference for the prevalence of lipoma, and the rate of malignant transformation is exceedingly rare. Therefore drop metastases do not occur. The high signal on T1, T2, and FLAIR sequences suggest tissue type, but does not have a relationship to neoplastic potential.

Reference:

Question(s) 464 - 468: Anatomy
Discussion:
Subarachnoid hemorrhage headache is characterized by sudden onset, severe, headache which may be associated with photophobia and neck stiffness. Any patient presenting with _the worst headache_ in her life, particularly if associated with photophobia or neck stiffness, must be emphatically evaluated for subarachnoid hemorrhage. Temporal arteritis results from granulomatous inflammation of the arteries of the head and neck. Blood flow is compromised through the inflamed vessels resulting in ischemia that may lead to retinal infarction and jaw claudication. Classic migraine is characterized by an aura that may consist of fortification spectra in the visual fields. The aura is followed by throbbing, often hemicranial headache. Common migraine is devoid of aura. Trigeminal neuralgia is characterized by lacinating pain in the distribution of branches of the trigeminal nerve. The pain is severe, episodic and may be provoked by minor stimulation of trigger zones. Cluster headache is an episodic severe hemicranial vascular headache usually lasting less than an hour but occurring in groups. The attacks may occur at the same time of day for several days and then stop for several weeks or months only to return. During the attack, the patient may have nasal stuffiness, lacrimation, and a Horner_s syndrome.
Question(s) 469 - 473: Anatomy
Discussion:
Abductor pollicis brevis is innervated by the median nerve with radicular derivation from C8 and T1. The abductor digiti minimi is innervated by the ulnar nerve but also has radicular contributions from C8 and T1. Presence of fibrillation potentials indicating denervation in the abductor digiti minimi coupled with the absence of fibrillation potentials in the abductor pollicis brevis indicates that an ulnar mononeuropathy is much more likely in this case than a radiculopathy. The serratus anterior muscle is innervated by the long thoracic nerve and stabilizes the scapula for forward pushing and arm elevation. No sensory loss is evident and on examination scapular winging is evident. Radial mononeuropathy as in this case most commonly occur at the midhumeral level near the spiral groove. The triceps is spared because branches innervating this muscle originate proximal to the spiral groove. Radial mononeuropathy presents predominantly with wrist and finger drop with little or no pain. The infraspinatus muscle is innervated by the suprascapular nerve distal to the spinoglenoid notch of the scapula. More proximally, the suprascapular nerve innervate the supraspinatus muscle.

Reference:
http://www.teleemg.com/ EMG and Nerve Conductions Velocity Homepage by Joe F. Jabre, M.D.

Question(s) 474 - 477: Anatomy
Discussion:
Bilateral putamenal necrosis is classic for methanol intoxication. Patients may manifest parkinsonism due to this selective basal ganglia damage, as well as blindness secondary to retinal ganglion cell degeneration. Ingestion of ethylene glycol can either be purposeful as a result of a suicide attempt or accidental when containers that once were used to haul antifreeze are not properly cleaned before they are used for carrying water. Ethylene glycol ingestion may be fatal and at autopsy the brain shows generalized edema, petechial hemorrhages, and oxalate crystal deposition. Inorganic arsenic causes a peripheral sensory motor neuropathy with axonal degeneration and demyelination. Muscle biopsy is devoid of vacuolar myopathy, unlike the findings in colchicine poisoning. Urban or Native American youth are groups of individuals who are particularly prone to glue sniffing as a form of recreational drug use. Toluene exposure over time causes cognitive deficits and white matter abnormalities in cerebral white matter.

Reference:

Question(s) 478 - 480: Anatomy
Discussion:
Cerebral cysticercosis often presents with seizures as the parasite dies and incites a vigorous inflammatory response often associated with local edema. Imaging often shows multiple cystic lesions with a nodule. The most common form of fungal meningitis in immunocompromised patients is cryptococcal meningitis. This chronic meningitis is associated with headache, cognitive decline, and cranial neuropathies. CSF analysis shows elevated protein, reduced glucose, and elevated lymphocyte count. CNS Lyme disease is associated with a low grade meningoencephalitis and neuropathies. Bilateral facial nerve palsies...
may be seen. The major differential diagnostic considerations in bilateral facial palsy are Lyme disease, sarcoidosis, and Guillain-Barre syndrome.

**Reference:**