EXAM OBJECTIVES

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

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

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

FREQUENTLY CITED REFERENCES


Question(s) 1: Pharmacology/Chemistry

Discussion:
Vigabatrin is an irreversible inhibitor of the GABA (gamma-aminobutyric acid) transaminase, which is especially effective in treatment of infantile spasms. Despite its favorable pharmacokinetic profile, vigabatrin has not been approved in the United States because it produces visual field defects, particularly progressive constriction with nasal sparing. This has been attributed to selective vulnerability of cells of the inner retina that use GABA as a neurotransmitter.

Reference:

Question(s) 2: Neuroradiology

Discussion:
The enhancement described is most characteristic of meningioma. Typically, in glioblastoma and arteriovenous malformation, the enhancement occurs in the arterial phase and by the venous phase, the enhancement has disappeared. The other lesions often do not show tumor blush on angiography.

Reference:

Question(s) 3: Pharmacology/Chemistry

Discussion:
Calpain-3 is a muscle specific calcium-activated neutral protease. Mutations producing deficiency of this enzyme produce an autosomal recessive form of limb-girdle muscular dystrophy (LGMD2A). Other LGMDs are due to dysferlinopathy (LGMD2B) and sarcoglycanopathies (LBMD2C-2F).

Reference:

Question(s) 4: Behavioral

Discussion:
Sedation with antidepressants is usually dose dependent with higher doses causing more sedation. Mirtazapine is unique in that it has less sedation when a dose greater than 15 mg/d is administered.

Reference:

Question(s) 5: Physiology

Discussion:
Electrocerebral silence can be seen in patients with a drug overdose or intoxication who later recover. The recordings should be done at a sensitivity of 2 micro-V/mm and an impedance of greater than 100 ohms, and using long inter-electrode distances.

Reference:

Question(s) 6: Anatomy

Discussion:
The mammillothalamic tract ends in the anterior nuclear group of the thalamus.

Reference:
Question(s) 7: Pathology
Discussion:
Intraventricular hemorrhage in premature infants originates from rupture of the fragile blood vessels in the subependymal germinal matrix. The vascular germinal matrix spawns the neuroectodermal constituents of the brain and involutes at maturity. In the premature, this structure is still present, and in the face of hemodynamic instability or hypoxemia may hemorrhage.

Reference:

Question(s) 8: 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) 9: 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) 10: 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) 11: Physiology
Discussion:
Valproic acid is the drug of choice for patients with absence and tonic-clonic seizures and the 3Hz spike 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) 12: Pharmacology/Chemistry
Discussion:
The aim of treatment of severe hypertension in neurologic catastrophes associated with increased intracranial pressure is to reduce the blood pressure to safe levels avoiding further increase in intracranial pressure. The drugs of choice include labetalol, esmolol, and enalapril. Clonidine may be used in some cases. Vasodilators, such as calcium channel blockers or nitrates, are best avoided as they may increase intracranial pressure in the setting of reduced systemic blood pressure, thus reducing cerebral perfusion pressure in areas with poor cerebral autoregulation.

Question(s) 13: Clinical Adult
Discussion: Sudden death is more often a manifestation of cardiac disease than of neurologic disease.


Question(s) 14: 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 paraspinal muscles (electromyographically) confirms that the site of the lesion is at the level of the nerve root.


Question(s) 15: Pharmacology/Chemistry
Discussion: REM sleep behavior disorder (RBD) occurs in patients with neurodegenerative disorders, such as Parkinson's disease and multiple system atrophy. Clonazepam (0.5-1.0 mg at bedtime) is very effective in controlling both the behavior and the dream components of RBD.


Question(s) 16: Clinical Adult
Discussion: Oral potassium in repeated doses is recommended for an attack of hypokalemic periodic paralysis.


Question(s) 17: Pathology
Discussion: The presence of demyelination with inclusion bodies in oligodendrocytes is characteristic of progressive multifocal leukoencephalopathy (PML). The astrocytes in the areas of demyelination may be large and contain atypical bizarre nuclei. PML is caused by infection of oligodendroglia with the JC virus, which is a member of the papova group of DNA viruses. The virus receives its name from the initials of the first patient from whom it was isolated and has no relationship to Jakob-Creutzfeldt disease (more commonly termed Creutzfeldt-Jakob disease, CJD), which is a prion disorder, not a viral disease.


Question(s) 18: Anatomy
Discussion: The short head of the biceps femoris is the only muscle proximal to the knee innervated by the peroneal division of the sciatic nerve.

Reference: Delagi EF, Perotto A. Anatomic guide for the electromyographer 1980;172.
Question(s) 19: Pathology
Discussion:
Hemorrhage and necrosis of the central core of the spinal cord usually results from traumatic subluxation injury of the cervical spine. Rarely, hematomyelia can also result from intraspinal vascular malformations.

Reference:

Question(s) 20: Clinical Adult
Discussion:
Although the pattern of recovery following a stroke may be variable, the earliest evidence of a return of neurological function is usually an increase in tone. This generally occurs before improvement in strength or a return of reflexes.

Reference:

Question(s) 21: Clinical Adult
Discussion:
Hypnic headache is a rare nocturnal headache disorder affecting the elderly. Women are more often affected than men. Attacks of pain may be unilateral or bilateral and are usually not accompanied by autonomic symptoms. The headaches occur two to four hours after sleep onset and last 15 to 30 minutes, occasionally hours. Some patients have several attacks per night.

Reference:
Dodick DW, Mosek AC, Campbell JK. The hypnic ("alarm clock") headache syndrome. Cephalgia 1998;18:152-156.

Question(s) 22: Pathology
Discussion:
Chloroquine toxicity is associated with long-term administration of the drug at doses greater than 200 mg/day; the primary target of damage is skeletal muscle, with less frequent involvement of cardiac muscle or retina. Methotrexate toxicity causes a necrotizing leukoencephalopathy, phenytoin damages Purkinje cells (either primarilly or from intermittent hypoxia associated with repeated seizures), taxol toxicity causes a dose-dependent peripheral neuropathy with painful dysesthesias, and vincristine also causes a neuropathy, usually initially a sensory neuropathy.

Reference:

Question(s) 23: Neuroradiology
Discussion:
Enhancement in the internal auditory canal is a non-specific finding as seen in many conditions, in addition to acoustic schwannoma.

Reference:

Question(s) 24: Pharmacology/Chemistry
Discussion:
Several growth/differentiation factors have been shown to increase survival of specific neuronal groups. The neurotrophin family including nerve growth factor (NGF), brain derived neurotrophic factor (BDNF) and neurotrophin-3 (NT-3) act via receptor tyrosine kinase. For example, NGF, acting via trkA receptors, increases survival of nociceptive, sympathetic, and basal forebrain cholinergic neurons. Glial derived neurotrophic factor (GDNF) acts via a receptor complex that includes the transmembrane tyrosine kinase Ret and is critical for development of some nociceptive, parasympathetic, enteric, and dopaminergic substantia nigra neurons. Ciliary neurotrophic factor (CNTF) is a member of the cytokine family that acts via a receptor complex that may also recognize interleukin-6. CNTF increases survival of motor neurons and parasympathetic neurons. Activation of the cytokine class receptor by
CNTF accounts for the severe side effects observed when this agent was administered in clinical trials for amyotrophic lateral sclerosis.

Reference:

Question(s) 25: Pharmacology/Chemistry
Discussion:
Neuroleptic malignant syndrome is felt to result from insufficient stimulation of dopamine receptors and is typically precipitated by treatment with high potency neuroleptics. Dopamine agonist drugs, such as bromocriptine, have proven effective in treating this potentially lethal condition.

Reference:

Question(s) 26: 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 is called Parkin and it is involved in protein breakdown. It is a ubiquitin ligase and is involved in the degradation of many proteins including alpha-synuclein.

Reference:

Question(s) 27: Physiology
Discussion:
Breach rhythm is a finding seen in patients with skull defect, which alters the conductance between the brain and the recording electrode over the scalp.

Reference:

Question(s) 28: Clinical Pediatrics
Discussion:
Patients with Lennox-Gastaut syndrome have multiple types of seizures, such as tonic, atypical absence, and atonic with age of onset of one to eight years. The EEG reveals slow spike-wave discharges, 1.5 to 2 Hz. Most children are mentally retarded and approximately 70% have an identifiable cause for the retardation and epilepsy. Hypsarrhythmia is an abnormal EEG pattern usually associated with infantile spasms in infants less than one year of age.

Reference:

Question(s) 29: Behavioral
Discussion:
An anterior callosal lesion disconnects the left hand (right motor strip) from the verbal left hemisphere, yielding an apraxia to verbal commands confined to the left hand (alien hand syndrome).

Reference:
Question(s) 30: Clinical Adult

Discussion:
CNS toxoplasmosis is the most commonly encountered neurologic opportunistic infection in AIDS patients. Pyrimethamine and sulfadiazine are of initial benefit in up to 90% of patients. The CSF profile can be identical to that of HIV encephalopathy. Blood and CSF serologies are not in general diagnostically reliable. Most adults in the U.S. have antibodies indicative of prior exposure to toxoplasmosis and cerebral involvement is thought to represent reactivation of a late primary infection. A course of empiric therapy for toxoplasmosis is recommended in many cases.

Reference:

Question(s) 31: Anatomy

Discussion:
Innervation of the detrusor muscle is parasympathetic (S2-S4).

Reference:

Question(s) 32: Physiology

Discussion:
F-responses can be used to assess the entire length of the motor axon and can be absent or delayed in demyelinating diseases of the peripheral nerve, such as in Guillain Barré syndrome.

Reference:

Question(s) 33: 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) 34: 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) 35: Clinical Adult

Discussion:
The most common neurologic manifestation of Legionnaires' disease is delirium (found in 30% of cases). Ataxia, transverse myelitis, and CSF pleocytosis are less common manifestations.

Reference:
Question(s) 36: Clinical Adult
Discussion:
Episodic dyscontrol is used to identify short periods of rage (violence) in an otherwise peaceful individual. The patient often expresses disbelief and/or remorse concerning the incident; some degree of intoxication (most often alcohol) is frequently but not invariably present.

Reference:

Question(s) 37: Clinical Adult
Discussion:
Partial seizures are those for which there is behavioral or EEG evidence of onset in a focal area of the brain limited to one lobe or hemisphere. The seizure focus determines whether the seizure has sensory manifestations, motor manifestations or both. Partial seizures are classified as complex when there is impairment of consciousness. Partial complex seizures may occur at any age. Partial complex seizures most often arise from the temporal lobe but may also arise from other lobes of the brain, particularly the frontal lobe. They respond to a number of different anticonvulsants, though they tend to be more refractory to medical treatment than most other types of seizure.

Reference:

Question(s) 38: Behavioral
Discussion:
Late-life delusional disorder is characterized by the occurrence of non-bizarre delusions, predominantly involving erotic, grandiose, jealous or persecutory themes. Average age of onset for women is 60-69 years and the minimum duration of symptoms is one month.

Reference:

Question(s) 39: Behavioral
Discussion:
Anosodiaphoria may be seen with right hemisphere lesions as in this patient. It is indifference to their condition despite the recognition that they may have a hemiparesis and/or hemisensory deficit.

Reference:

Question(s) 40: Clinical Adult
Discussion:
ALS is a disorder of upper and lower motor neurons. The associated dysarthria reflects both flaccid and spastic features.

Reference:

Question(s) 41: 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) 42: Pharmacology/Chemistry
Discussion:
Malignant hyperthermia and central core disease are autosomal dominant disorders associated with mutations in the muscle ryanodine receptor (RYR). The RYR is the calcium release channel of the sarcoplasmic reticulum of the skeletal muscle.

Reference:

Question(s) 43: Clinical Adult
Discussion:
Marburg's variant of multiple sclerosis is among the more acute of the idiopathic demyelinating disorders. It may need to be included in the differential diagnosis of stupor and coma.

Reference:

Question(s) 44: Clinical Adult
Discussion:
Duchenne's muscular dystrophy is associated with high creatine kinase (CK) levels. It is a sex-linked recessive disorder resulting in abnormal amounts of a protein, dystrophin, in muscle.

Reference:

Question(s) 45: Pharmacology/Chemistry
Discussion:
Nimodipine and other dihydropyridines block calcium influx through L-type channels. These channels are involved in slow depolarization and excitation-contraction coupling of the vascular smooth muscle. Calcium influx through N- and P/Q channels triggers neurotransmitter release.

Reference:

Question(s) 46: Behavioral
Discussion:
Carbamazepine and amitriptyline have both been used for emotional lability. However, carbamazepine is the better choice in persistent post-traumatic agitation because it does not have the anticholinergic and mild epileptogenic properties that amitriptyline has.

Reference:

Question(s) 47: Physiology
Discussion:
An EEG pattern consisting of generalized, bisynchronous periodic sharp-and-slow wave complexes recurring at 4- to 15-second intervals is characteristic of subacute sclerosing panencephalitis.

Reference:
Question(s) 48: Clinical Adult
Discussion:
Progressive myelopathy with chiefly motor signs, occurring in various endemic foci (Jamaica, South India, South Africa, Colombia, Japan) is associated with the retrovirus HTLV-I. Multiple sclerosis is suggested by recurrent optic neuritis. Cycad consumption may be associated with Guamanian ALS and the parkinsonism-dementia complex.

Reference:

Question(s) 49: 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 angiomatous and have no association with aggressive behavior. The older term "angioblastic meningioma" encompassed a heterogeneous group of vascular dural-based tumors that included angiomatous 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) 50: 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) 51: 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:
Question(s) 52: Pharmacology/Chemistry
Discussion:
Galantamine (Reminyl) is a drug recently approved in the United States for treatment of Alzheimer's disease. Unlike donepezil, rivastigmine, and tacrine, galantamine not only blocks acetylcholinesterase but also activates presynaptic nicotinic receptors by binding to an allosteric site of the molecule. This may result in increased release of neurotransmitters.

Reference:

Question(s) 53: 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:

Question(s) 54: Pharmacology/Chemistry
Discussion:
Fluconazole, together with itraconazole and ketoconazole, belongs to the triazole class of antifungal drugs that inhibit the synthesis of ergosterol, with the accumulation of substituted sterols that interfere with the synthesis and permeability of the fungal cell membrane. Like other triazoles, fluconazole inhibits cytochrome P450 - dependent liver metabolism and consequently increases plasma concentration of phenytoin, warfarin, cyclosporine, zidovudine, oral hypoglycemic agents, and other drugs. Fluconazole is the most widely used oral agent for CNS fungal infections. One of its advantages is that it crosses the blood brain barrier well, with CSF concentrations that are 50-90% of the plasma. Its main disadvantage is that it has a lower cure rate than amphotericin B for most CNS fungal infections, including cryptococcal meningitis.

Reference:

Question(s) 55: 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) 56: Physiology
Discussion:
In secondary bilateral synchrony, the focal epileptiform discharge spreads to relevant subcortical and forebrain structures and then to the contralateral cortex.

Reference:

Question(s) 57: Physiology
Discussion:
Inclusion body myositis produces a characteristic pattern of weakness involving finger flexor and quadriceps muscles most severely. Muscle enzymes are normal or minimally increased, and EMG studies typically reveal an inflammatory myopathy. The muscle biopsy finding most typical of inclusion body myositis is vacuole formation surrounded by a basophilic rim.

Reference:
**Question(s) 58: Physiology**

**Discussion:**
Median somatosensory evoked potentials in cervical spondylosis may show diminished amplitude of N13. Arnold-Chiari malformation is associated with normal amplitude of N13 and increased N13-N20 interpeak latency. In amyotrophic lateral sclerosis, median SEP's are typically normal.

**Reference:**

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**Question(s) 59: Neuroradiology**

**Discussion:**
MRI does not involve gamma rays, positrons, sodium or potassium. Only protons are involved. Nuclei with paired electrons in the outer shell do not give an MR signal.

**Reference:**

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**Question(s) 60: Pharmacology/Chemistry**

**Discussion:**
Pantothenate kinase is essential to coenzyme A (CoA) synthesis. Deficiency of CoA may lead to impaired membrane biosynthesis, for example in rod photoreceptors, that degenerate in HSS. Phosphopantothenate, the product of pantothenate kinase, is deficient, leading to cysteine accumulation, that in turn predisposes to free radical production and lipid peroxidation in the presence of iron.

**Reference:**

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**Question(s) 61: Pathology**

**Discussion:**
Malignant peripheral nerve sheath tumors most commonly arise in a neurofibroma, often of the plexiform type. They may also arise de novo in a normal nerve. Malignant transformation of a schwannoma or ganglioneuroma is rare. It is extremely rare to find these malignant peripheral nerve sheath tumors involving or arising from cranial nerves or cranial nerve neurofibromas.

**Reference:**

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**Question(s) 62: Physiology**

**Discussion:**
Vasopressin is secreted by cells of the supraoptic nuclei.

**Reference:**

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**Question(s) 63: Neuroradiology**

**Discussion:**
Cavernous hemangioma, one of the most common malformations identified on MRI, is best visualized on gradient echo and T2-weighted images with high field strength scanners. They typically have popcorn appearance with mixed signal core and hemosiderin rim. Pontine glioma, ruptured aneurysm, tuberculosis or venous angioma do not contain calcium or hemosiderin rim.

**Reference:**
Question(s) 64: Pharmacology/Chemistry
Discussion:
The serotonin syndrome consists of a combination of mental and behavioral changes, motor hyperactivity, and autonomic lability, that occurs following use of potent serotonimetic agents alone or in combination with non-specific monoamine oxidase inhibitors (MAOIs). This syndrome can occur in patients taking various combinations of drugs, including serotonin precursors (e.g. tryptophan), serotonin reuptake inhibitors (e.g. fluoxetine, sertraline, clomipramine, imipramine, nortriptyline, trazodone), MAOIs (clorgyline, phenelzine, tranylcypromine, iproniazid).

Reference:

Question(s) 65: Clinical Adult
Discussion:
In barbiturate coma, ciliospinal and oculovestibular reflexes may be absent, the EEG may be flat, and there may be loss of spontaneous respirations. Characteristically, the pupils remain reactive.

Reference:

Question(s) 66: Physiology
Discussion:
EMG needle examination evidence of fibrillation potentials and decreased recruitment of motor units in the iliopsoas, vastus medialis, adductor longus and vastus lateralis muscles without other abnormality localizes the lesion to the lumbar plexus.

Reference:

Question(s) 67: Pharmacology/Chemistry
Discussion:
Complex partial status epilepticus presents with altered awareness and responsiveness. Automatisms may be present. EEG will demonstrate characteristic epileptiform discharges. This form of status epilepticus usually responds well to intravenous benzodiazepines, such as lorazepam, which must be followed by anticonvulsants effective in chronic seizure control, such as carbamazepine.

Reference:

Question(s) 68: Physiology
Discussion:
In BAEP's, absence of wave I with intact wave V is most commonly due to peripheral hearing loss.

Reference:

Question(s) 69: Anatomy
Discussion:
The anterior choroidal artery arises from the internal carotid artery distal to the origin of the posterior communicating artery. It has a long subarachnoid course, enters the inferior horn of the lateral ventricle through the choroidal fissure, and supplies the amygdaloid complex, hippocampal formation, globus pallidus, and the ventrolateral portion of the posterior limb and the entire retrolenticular portion of the internal capsule.

Reference:
Question(s) 70: Anatomy
Discussion:
The inferior cerebellar peduncle connects the medulla to the cerebellum and contains the dorsal spinocerebellar tract, cuneocerebellar tract, olivocerebellar tract, and the vestibulocerebellar tract. The trigeminocerebellar tract lies within the superior cerebellar peduncle and would be spared in a lesion confined to the inferior cerebellar peduncle.

Reference:

Question(s) 71: Pathology
Discussion:
Sturge-Weber syndrome is characterized by facial vascular nevi in a trigeminal distribution and leptomeningeal vascular malformation of the occipital lobe. Some patients also have ocular choroidal angiomas.

Reference:

Question(s) 72: Clinical Adult
Discussion:
Paroxysmal dyskinesias consist of episodes of involuntary movements and are classified according to phenomenology, duration of attacks, and etiology. With paroxysmal kinesigenic dyskinesia (paroxysmal kinesigenic choreoathetosis) abnormal involuntary movements lasting seconds to minutes occur abruptly after a sudden voluntary movement. With paroxysmal nonkinesigenic dyskinesia (paroxysmal dystonic choreoathetosis) attacks of involuntary movements last minutes to hours and occur spontaneously.

Reference:

Question(s) 73: Anatomy
Discussion:
The ciliary muscle receives post ganglionic parasympathetic innervation via the 3rd (oculomotor) cranial nerve. The superior cervical ganglion provides the sympathetic innervation to the facial sweat glands, carotid artery, pineal gland and tarsal muscle.

Reference:

Question(s) 74: Physiology
Discussion:
One of the most important technical aspects of recording evoked potentials is replication of the waveforms.

Reference:

Question(s) 75: Pharmacology/Chemistry
Discussion:
Demyelination results in dysregulation of expression of voltage gated sodium channels in the axon. There is upregulation of some sodium channel subtypes and homogeneous redistribution of the sodium channels along the axon. This allows recovery of function, but at the expense of abnormal axonal excitability.

Reference:
Question(s) 76: Clinical Adult
Discussion:
Aphasia is a disorder of language, to be distinguished from disorders of speech (dysarthria, dysphonia) and disorders of thought (e.g., dementia, confabulation, perseveration, agnosia). Aphasic utterances can involve nonexistent word forms called neologisms.

Reference:

Question(s) 77: Pharmacology/Chemistry
Discussion:
Sudden sensorineural hearing loss (SSNHL) has an incidence of 1:3000. A study of 603 patients with this diagnosis compared the outcomes in patients treated with IV pentoxifylline alone, IV glucocorticoids alone, and the combination of IV pentoxifylline and IV glucocorticoids. None of the therapies appeared to be harmful; the combination of steroids and pentoxifylline produced more improvement at all frequencies than pentoxifylline alone, and glucocorticoids alone produced better results than non-steroidal therapy. Antiviral therapy for SSNHL has not been evaluated in this fashion.

Reference:

Question(s) 78: Pathology
Discussion:
Apoptosis is an active energy-dependent process in which individual cells discretely die without provoking the hubbub of inflammation. Proteases, endonucleases and lipases are activated, destroying the cell from within. One hallmark of apoptosis is fragmentation of the nucleus into chunks called apoptotic bodies. Cellular and organelle swelling are more common in necrosis in which the cell explodes, thereby releasing its contents into its environment and provoking an inflammatory response.

Reference:

Question(s) 79: 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) 80: Clinical Pediatrics
Discussion:
A study of 160 children from 36 weeks gestation to 18 years of age with radiographic confirmation of cerebral sinovenous thrombosis found that 58 percent of children had seizures, 76 percent had diffuse neurologic signs, and 42 percent had focal neurologic signs. 43 percent of patients were neonates, and 54 percent were under one year of age. Risk factors were present in 98 percent of cases, and included acute systemic illness, head and neck disorders and abnormal tests for prothrombotic disorders (in 38 percent). Seizures at presentation and the presence of venous infarcts predicted adverse neurologic outcomes.

Reference:
Question(s) 81: Anatomy
Discussion:
The upper visual fields ultimately project to the inferior lip of the contralateral calcarine sulcus.

Reference:

Question(s) 82: Physiology
Discussion:
A patient with a recent stroke and seizures would most likely show periodic lateralized sharp wave discharges on the EEG.

Reference:

Question(s) 83: 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) 84: Pharmacology/Chemistry
Discussion:
Cocaine, like amphetamine, binds to the presynaptic dopamine transporter (DAT) inhibiting dopamine reuptake and increasing dopamine concentrations in the nucleus accumbens. This mechanism underlies the acute reinforcing effects of these drugs of addiction.

Reference:

Question(s) 85: 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) 86: Anatomy
Discussion:
The internal arcuate fibers from the dorsal column nuclei (cuneate and gracile nuclei) decussate completely to form the medial lemniscus.

Reference:

Question(s) 87: Behavioral
Discussion:
Digit span, which involves attentional processes, immediate recall and ability to sequence bits of information, may be reduced following lesions of dorsolateral frontal cortex. Lesions of the fornix, mammillary bodies and medial dorsal nucleus of the thalamus, on the other hand, cause amnesia (impaired secondary memory) without a reduction of digit span.

Reference:
**Question(s) 88: Clinical Pediatrics**

**Discussion:**
Hypoxic-ischemic encephalopathy is the most common cause of early seizures in both premature and full-term infants. Intracranial hemorrhage may occur anytime during the first week and is a more common cause of seizures in premature infants than in term infants. Developmental abnormalities of the brain may be associated with early or late seizures. Kernicterus initially presents with hypotonia, cerebral depression, and seizures are uncommon. Pyridoxine dependency is a very rare cause of intractable seizures.

**Reference:**

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**Question(s) 89: Physiology**

**Discussion:**
A progressive increase in latency, duration, amplitude and area of motor and sensory nerve action potentials accompanies a physiologic decline in temperature. This is reversed by warming.

**Reference:**

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**Question(s) 90: Pharmacology/Chemistry**

**Discussion:**
Toxic leukoencephalopathy is associated with exposure to therapeutic agents (particularly antineoplastic drugs), drugs of abuse and environmental toxins. Hyperintensity of white matter on T-2 weighted sequences is characteristic, and the diagnosis should not be made in the absence of such changes. Methotrexate and carmustine are the anticancer drugs most commonly implicated in toxic leukoencephalopathy. Carbon monoxide poisoning typically produces demyelination that begins days to weeks after exposure. The mechanism is not known, although prolonged depression of oxygenation and circulation has been proposed. Alcoholism produces a number of changes, including an excessive number of white matter hyperintensities, loss of total white matter volume and preferential involvement of the frontal white matter that correlates with observed neuropsychological deficits.

**Reference:**

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**Question(s) 91: 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:**

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**Question(s) 92: Clinical Adult**

**Discussion:**
Epidural metastases (carcinoma, lymphoma, myeloma) are the most common spinal tumors in the elderly.

**Reference:**

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**Question(s) 93: Clinical Adult**

**Discussion:**
Prosopagnosia (inability to identify faces visually) is most often seen after bilateral inferior occipito-temporal lesions affecting both fusiform gyri. Similar bilateral lesions (or a nondominant lesion) of the fusiform and lingual gyri account for cerebral achromatopsia. Palinopsia (recurrence of a visual image after diverting gaze or when the stimulus object is withdrawn) occurs with occipito-temporal disease, often epileptogenic. Alexia without agraphia (inability to read but with preserved writing ability) occurs with combined lesions of the
dominant occipital lobe and the inferior splenium of the corpus callosum or with a single lesion of the dominant occipito-temporal paraventricular white matter behind, beneath, and beside the occipital horn of the lateral ventricle. Alexia with agraphia (central alexia) is usually due to an angular gyrus (parietal lobe) lesion.

Reference:

Question(s) 94: Pharmacology/Chemistry
Discussion:
3,4-diaminopyridine (3,4-DAP) blocks a K+ channel and increases acetylcholine release. It has been used for treatment of Lambert-Eaton myasthenic syndrome.

Reference:

Question(s) 95: Physiology
Discussion:
Absence seizures are characteristically seen in children 5-15 years of age associated with staring and may have automatisms such as repetitive chewing movements. Similar features may be seen in complex partial seizures, although absence seizures are shorter and may occur more frequently. The characteristic electroencephalogram during an absence seizure would show 3 Hz spike-and-wave discharges.

Reference:

Question(s) 96: 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) 97: 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) 98: 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 inputs from the cortex and projects to the other thalamic nuclei, and is critical for thalamocortical synchronization, particularly generation of sleep spindles.

Reference:

Question(s) 99: Pathology
Discussion:
Lesions associated with Friedreich ataxia include degeneration of the posterior columns, corticospinal tracts, spinocerebellar tracts and dorsal roots. Anterior motor neurons are not affected.

Reference:
Question(s) 100: Anatomy
Discussion:
The most prominent efferent pathway from the amygdaloid nuclear complex is the stria terminalis. Most of the fibers arise from the corticomediaal part of the nuclear complex. They arch along the medial border of the caudate near its junction with the thalamus. The largest termination is the nucleus of the stria terminalis. This is located lateral to the columns of the fornix and dorsal to the anterior commissure. Other terminations include the anterior hypothalamic nuclei and the medial preoptic area.

Reference:

Question(s) 101: Clinical Adult
Discussion:
A lesion of the medial longitudinal fasciculus results in impaired adduction of the ipsilateral eye, and not a conjugate deviation of the eyes.

Reference:

Question(s) 102: Clinical Adult
Discussion:
Takayasu’s syndrome is a nonspecific arteritis involving mainly the aorta and the large arteries arising from its arch. Most of the patients have been young Asian females. The exact etiology has never been ascertained but an autoimmune mechanism has been suspected.

Reference:

Question(s) 103: Pharmacology/Chemistry
Discussion:
The vanilloid receptor (VR1) is a member of the transient receptor potential (TRP) family of cation channels, which are particularly permeable to calcium. The VR1 receptor is activated by capsaicin, noxious heat, and acid pH.

Reference:

Question(s) 104: Physiology
Discussion:
Botulism results in an impaired release of acetylcholine from the nerve terminal.

Reference:

Question(s) 105: Behavioral
Discussion:
Visual hallucinations may be experienced but are uncommon in the dementia of depression, Alzheimer’s disease, progressive supranuclear palsy, Pick’s disease, and normal pressure hydrocephalus. They are commonly associated with diffuse Lewy body disease.

Reference:

Question(s) 106: Behavioral
Discussion:
Prosopagnosia results from bilateral occipitotemporal junction lesions. It is often associated with achromatopsia because of involvement of the fibers projecting from the inferior lip of the occipital lobe.
Reference:

Question(s) 107: Pathology
Discussion:
Normal pressure hydrocephalus may easily be mistaken clinically for Binswanger's or Alzheimer's disease, and the latter may co-exist with normal pressure hydrocephalus but do not cause it. The two most common antecedent conditions that may have occurred years before the development of normal pressure hydrocephalus and subarachnoid hemorrhage and head injury.

Reference:

Question(s) 108: Pathology
Discussion:
The predominant pathological lesion in patients with homocystinuria is thromboembolic disease. Involvement of cerebral blood vessels results in infarcts in the cerebrum, cerebellum, midbrain, and thalamus. Thrombi may also be seen in dural venous sinuses. Arterial walls of affected patients may show fibrous intimal thickening.

Reference:

Question(s) 109: Pathology
Discussion:
Examination of the cortex shows the characteristic inclusion of myoclonic epilepsy, the Lafora body.

Reference:

Question(s) 110: Neuroradiology
Discussion:
The Chiari type I malformation is defined as congenital displacement of cerebellar tonsils (with or without vermian displacement) into the cervical spine canal. It is frequently associated with narrow foramen magnum and syringohydromyelia (syrinx). Hydrocephalus is not a common finding. More than 2-3 mm downward displacement of the tonsils is considered abnormal. The reference point is posterior lip of foramen magnum. Herniation beyond C3 is rare. The 4th ventricle locates in its normal position in the posterior fossa. Spinal dysraphism, open tentorial incisura and narrow tectum are associated with Chiari II (but not with Chiari I malformation). Chiari I malformation is often asymptomatic, especially if small. 2-3 mm herniation is a very common finding in MRI. The correct answer is syrinx.

Reference:

Question(s) 111: Anatomy
Discussion:
Cytoplasmic filaments are classified into three categories based on cross sectional diameter: thin (5-7 nm), intermediate (10-12 nm), and thick (16 nm). Thin filaments are composed of actin and thick filaments are composed of myosin. There are several different types of intermediate filaments, which differ in their protein composition. The intermediate filament proteins are: GFAP, neurofilament proteins, cytokeratins, desmin, vimentin, and nestin. Microtubules, which are composed of tubulin, are larger than thick filaments, with a diameter of 24 nm.

Reference:
Question(s) 112: Behavioral Discussion:
Approximately 40-50% of Alzheimer's disease patients will experience delusions during the course of their disease. Most common are the persecutory delusions such as theft or infidelity. About ten percent of patients may experience hallucinations, usually visual or auditory. Grandiose delusions are rare, as are olfactory hallucinations.

Reference:

Question(s) 113: Pathology Discussion:
Head injury patients with diffuse axonal injury are usually comatose from the time of injury and recovery is minimal. Experimental studies have shown that the histological hallmark of this condition - axonal spheroids or axonal retraction balls - develop over a period of hours to days.

Reference:

Question(s) 114: Anatomy Discussion:
The lower midbrain tegmentum contains the fibers of the superior cerebellar peduncle that decussate to reach the contralateral ventral lateral thalamic nucleus.

Reference:

Question(s) 115: Pathology Discussion:
Autosomal dominant cerebral cavernous malformation (CCM1) syndrome occurs in Hispanic individuals and is characterized by intraparenchymal cavernous malformations that can produce seizures, impairment of function, or hemorrhage.

Reference:

Question(s) 116: Pathology Discussion:
Meningoangiomatosis may be a cause of seizures or may be an asymptomatic condition. Like multiple meningiomas, meningoangiomatosis is associated with neurofibromatosis type 2.

Reference:

Question(s) 117: Anatomy Discussion:
The nucleus tractus solitarius is the first central relay station for both the vasodepressor and cardioinhibitory components of the baroreflex. The dorsal nucleus of the vagus and particularly the nucleus ambiguus mediate the cardioinhibitory effect. Inhibition of neurons of the ventrolateral medullary reticular formation is involved in the vasodepressor response.

Reference:
Question(s) 118: Anatomy
Discussion:
The posterior cord splits into the radial and axillary nerves.

Reference:

Question(s) 119: 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) 120: Behavioral
Discussion:
The triad of optic ataxia, ocular apraxia, and simultanagnosia (Balint’s syndrome) is usually the result of bilateral occipitoparietal junction lesions.

Reference:

Question(s) 121:
Pharmacology/Chemistry
Discussion:
Htt is a cytoplasmic protein, highly expressed in the cortex and striatum, that contains increased numbers of glutamine residues in Huntington Disease (HD), reflecting expansion of CAG repeats in the Htt gene. Mutant Htt undergoes proteolysis in the cell, and the amino-terminal fragments containing the glutamine expansions form aggregates, visible as nuclear and cytoplasmic inclusions in the brain. The mechanism of neurotoxicity of Htt is not proven, but several observations have been made. Mutant Htt activates caspase, thus promoting apoptosis, and also reduces transcription of brain-derived neurotrophic factor (BDNF), which normally protects striatal neurons from glutamate-induced excitotoxicity. Wild-type Htt has been shown to reduce the toxicity of mutant Htt in vivo.

Reference:

Question(s) 122: Clinical Adult
Discussion:
Spinal bulbar muscular atrophy (Kennedy’s disease) is characterized by lower motor neuron findings, especially affecting cranial musculature associated with decreased libido and gynecomastia. Female carriers are asymptomatic. Muscle biopsy shows non-diagnostic neuropathic changes. Testicular biopsy shows diminished spermatozoids with abnormal motility and mild elevation of FSH and LH. Diagnosis is by DNA analysis which shows expanded CAG repeats translated into polyglutamine repeats.

Reference:
Question(s) 123:
Pharmacology/Chemistry

Discussion:
Astrocyte-derived lactic acid is the main source of energy to the neuron. During periods of activity, the neuron releases glutamate, which is taken up by the astrocyte. Astrocyte uptake depends on the Na,K, ATPase. The fall in ATP levels activates glycolysis in the astrocyte, with production of lactate, which is released. Lactate is taken up by the neuron and converted to pyruvate via the lactate dehydrogenase, thus providing a source for neuronal aerobic metabolism.

Reference:

Question(s) 124: Pathology

Discussion:
Target fibers, which are best seen with trichrome or NADH stains, are characteristic of acute denervation.

Reference:

Question(s) 125: 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) 126: Anatomy

Discussion:
The insular cortex receives inputs from the parvocellular portion of the ventromedial nucleus of the thalamus, relaying taste and visceral sensation. In addition, it receives nociceptive-specific and thermal inputs from the posterior ventromedial nucleus, which is the site of termination of spinothalamic pathways arising from lamina I of the dorsal horn.

Reference:

Question(s) 127: Behavioral

Discussion:
Bilateral lesions of the amygdala result in the Kluver-Bucy syndrome. This syndrome is seen in herpes simplex encephalitis, Pick's disease, anoxic-ischemic lesions in the anterior medial temporal lobes, and after bilateral temporal lobectomy. It is rarely, if ever, seen as a manifestation of Creutzfeldt-Jakob, Alzheimer's or Huntington's disease.

Reference:

Question(s) 128: Clinical Adult

Discussion:
Allodynia is the perception of a non-noxious stimulus as being painful.

Reference:

Question(s) 129: Anatomy

Discussion:
The nucleus ambiguus contains motor neurons that supply striated muscles of the palate, pharynx and larynx.

Reference:
Question(s) 130:
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 tend to 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) 131:
Clinical Pediatrics

Discussion:
Hemiplegic cerebral palsy (CHP) is the most common CP syndrome among children born at term and is most commonly the result of cerebral infarction. Scoliosis occurs in only 10-20% of children with CHP, and is often mild. Likewise, general growth (height and weight) is not impaired, and the head circumference is normal in 85-90%. About 20% of children with CHP have learning disabilities.

Reference:

Question(s) 132:
Behavioral

Discussion:
The nonfluent cortical aphasias include Broca's, global, mixed transcortical, and transcortical motor.

Reference:

Question(s) 133:
Clinical Adult

Discussion:
Paroxysmal hemicrania is a disorder, more common in women, characterized by frequent (7 to 22 per day) episodes of unilateral severe but short-lasting (five to 45 minutes) headaches associated with ipsilateral autonomic manifestations. Indomethacin is the treatment of choice.

Reference:

Question(s) 134:
Behavioral

Discussion:
Significant pathological dilatation of the cerebral ventricular system, especially of the lateral and third ventricles, has been demonstrated in schizophrenic patients.

Reference:

Question(s) 135:
Clinical Adult

Discussion:
Amitriptyline relieves pain, but can have adverse cardiac effects. Sympathectomy relieves pain associated with reflex sympathetic dystrophy. Gabapentin may relieve neuropathic pain.

Reference:
**Question(s) 136: Anatomy**

**Discussion:**
The "frontal eye field" in humans is located in the caudal part of the middle frontal gyrus.

**Reference:**

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**Question(s) 137: Behavioral**

**Discussion:**
Clinical features of catatonia include psychosocial withdrawal, stupor, mutism, immobility, generalized analgesia, posturing, waxy flexibility, mannerisms, rituals, grimacing, and shoulder shrugging.

**Reference:**

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

**Discussion:**
Refsum's disease is a potentially treatable neuropathy due to defective alpha oxidation of phytanic acid in the microsomes. Diet low in this branched fatty acid (less than 10 mg/d) and, in severe cases, plasma exchange, can lead to improvement of the neuropathy.

**Reference:**

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**Question(s) 139: 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:**

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**Question(s) 140: Clinical Pediatrics**

**Discussion:**
Seizures, blindness, psychomotor retardation are cardinal features of Tay-Sachs disease. Tay-Sachs disease is an autosomal recessive disorder in which hexosaminidase A is deficient and hexosaminidase B is increased. The disorder is panethnic, but is more frequent in Ashkenazy Jews. Prenatal diagnosis is now available. Hepatosplenomegaly is not a feature of Tay-Sachs disease, but does occur in Sandhoff disease (deficiency of hexosaminidase A and B). Macrocephaly is frequent in GM2 gangliosidoses.

**Reference:**

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**Question(s) 141: Clinical Adult**

**Discussion:**
The most common visual field defect seen in early compressive lesions of the optic nerve is a central scotoma in the ipsilateral eye.

**Reference:**

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**Question(s) 142: Neuroradiology**

**Discussion:**
There is almost 100% association of meningomyelocele with Chiari II (Arnold-Chiari malformation) but not with Chiari I (tonsillar herniation). Association of midline dysraphism is present, thoraco-lumbar kyphoscoliosis and diastematomyelia are seen in approximately 30%.
Reference:

Question(s) 143: Clinical Pediatrics
Discussion:
Children with Tourette’s syndrome very frequently have an associated attention deficit disorder and obsessive-compulsive disorder. Coprolalia is quite unusual in children. Most children with Tourette’s do not need medication and often symptoms subside after adolescence. Motor tics and vocalizations can be suppressed for varying periods of time, which is helpful in distinguishing tics from chorea. The disorder is felt to be an autosomal dominant disorder but expression of the gene is very complex.

Reference:

Question(s) 144: Clinical Adult
Discussion:
Patients with spinal stenosis may have leg pain on exertion, which is relieved by rest (neurogenic claudication). This pain is worse with back hyperextension, relieved by leaning forward when walking (e.g. when using a shopping cart), and often not present when riding a bicycle. Absent peripheral arterial pulses suggests vascular claudication rather than neurogenic claudication.

Reference:

Question(s) 145: Physiology
Discussion:
The H-reflex is the electrophysiological equivalent of the monosynaptic tendon stretch reflex at the ankle.

Reference:

Question(s) 146: 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) 147: 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) 148: Anatomy
Discussion:
The dermatome of the thumb is C6.

Reference:
**Question(s) 149:**
Pharmacology/Chemistry

**Discussion:**
Friedreich's ataxia is the most common autosomal recessive cerebellar ataxia. It is due to an unstable expansion of the GAA repeat in the first intron in the frataxin gene in chromosome 9q. Frataxin is located in the mitochondria, where it appears to exert a protective effect against iron accumulation and generation of reactive oxygen species. Thus, it has been proposed that Friedreich's ataxia is due to oxidative stress due to free radicals generated in iron-filled mitochondria.

**Reference:**

**Question(s) 150:** Clinical Adult

**Discussion:**
Gastric resection is a cause of B12 deficiency due to loss of production of intrinsic factor.

**Reference:**

**Question(s) 151:** Physiology

**Discussion:**
Decerebrate rigidity is produced in response to a lesion in the brainstem below the level of the red nucleus.

**Reference:**

**Question(s) 152:** Pathology

**Discussion:**
The nucleus of Onufrowicz of the sacral cord is spared in motor neuron disease. The remaining motor nuclei are involved to varying degrees in ALS.

**Reference:**

**Question(s) 153:** Behavioral

**Discussion:**
Galantamine is a therapeutic agent for Alzheimer's disease that acts by both inhibiting AChE and allosterically modulating nicotinic ACh receptors. Rivastigmine acts by inhibiting AChE and butyrycholinesterase. Choline acetyltransferase is decreased in AD. Chromosomes 1, 14, 19, 21 and possibly 12 have been associated with the genetics of AD but not 3. Alpha secretase cleaves APP normally. Beta and gamma secretase have been implicated in the production of toxic beta amyloid.

**Reference:**

**Question(s) 154:** Clinical Adult

**Discussion:**
Crocodile tears, a sign of aberrant regeneration, is most commonly seen in association with Bell's palsy.

**Reference:**
**Question(s) 155: Pathology**

**Discussion:**
Triethyltin produces severe white matter edema due to accumulation of fluid in vacuoles within myelin sheaths. There is splitting of myelin along intraperiod lines. Other toxic causes of intramyelin edema include hexachlorophene, 5-fluorouracil, and lithium.

**Reference:**

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**Question(s) 156: Behavioral**

**Discussion:**
A diagnosis of major depression requires the presence of five out of nine depressive symptoms listed in DSM IV for at least two weeks. The diagnosis suggests a biologic pathophysiology and is usually an indication for pharmacologic intervention.

**Reference:**

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**Question(s) 157: Anatomy**

**Discussion:**
The anterior choroidal artery usually arises from the internal carotid artery.

**Reference:**

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**Question(s) 158: Physiology**

**Discussion:**
Motor conduction block, abnormal temporal dispersion, slowed conduction velocities, and prolonged late responses are the electrodiagnostic features of demyelination.

**Reference:**

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**Question(s) 159: Anatomy**

**Discussion:**
The subfornical organ is a circumventricular structure with no blood-brain-barrier. Although the subcommissural organ is circumventricular in nature, it possesses a capillary endothelium with tight junctions.

**Reference:**

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**Question(s) 160: 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) 161: Pathology
Discussion:
About 90% of patients with idiopathic syringomyelia have Chiari type I malformation. None of the spinal cord degenerative disorders are strongly associated with syringomyelia, nor is subacute combined degeneration. Megalencephaly is not associated with syringomyelia.

Reference:

Question(s) 162: Pharmacology/Chemistry
Discussion:
Oxcarbazepine is a derivative of carbamazepine and shares many similarities with carbamazepine, including its mechanism of action, ability to induce hepatic metabolism of oral contraceptives and other drugs, risk of hyponatremia, and indication for treatment of partial epilepsy. Unlike carbamazepine, oxcarbazepine is reduced to 10-monohydroxy-carbamazepine and does not undergo oxidation to an epoxide. This may explain its fewer side effects as compared to carbamazepine.

Reference:

Question(s) 163: Pharmacology/Chemistry
Discussion:
First episode demyelination in presumed multiple sclerosis should be treated with beta-interferon.

Reference:

Question(s) 164: Pharmacology/Chemistry
Discussion:
Levodopa crosses the blood-brain-barrier via a transport system shared by neutral amino acids of the diet. Lipid soluble drugs readily pass through barrier without a transport system.

Reference:

Question(s) 165: Physiology
Discussion:
Lennox-Gastaut syndrome is characterized clinically by frequent generalized seizures in association with mental retardation and a slow spike-and-wave EEG pattern.

Reference:

Question(s) 166: 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) 167: Pharmacology/Chemistry
Discussion: Abrupt withdrawal of CNS depressants, such as barbiturates and benzodiazepines that have been used at high doses for prolonged periods of time can produce seizures and other manifestations of central nervous system hyperexcitability that may be lethal. These effects are less common with drugs that have high binding to plasma proteins and long half-life.


Question(s) 168: Anatomy
Discussion: The little toe's dermatome is S1.


Question(s) 169: Physiology
Discussion: Light sensitivity manifested by photomyoclonus induced by photic stimulation can occur with abrupt alcohol withdrawal.


Question(s) 170: Anatomy
Discussion: The nucleus of the tractus solitarius (NTS) contains the first central neuron for the baroreceptor afferents. Lesions involving the NTS produce fluctuating hypertension mimicking a pheochromocytoma.


Question(s) 171: Pathology
Discussion: Ammon's horn sclerosis is seen in chronic temporal lobe epilepsy and may reflect hypoxic or excitotoxic injury to this structure.


Question(s) 172: Pathology
Discussion: CSF oligoclonal bands are seen in nearly all patients with subacute sclerosing panencephalitis and 83-94 percent of patients with definite multiple sclerosis. They are also seen in 25-50 percent of patients with neuroborreliosis, cryptococcal meningitis, Guillain-Barre syndrome, and other conditions. Many patients with neuromyelitis optica fail to have oligoclonal bands in CSF.


Question(s) 173: 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.

**Question(s) 174: Pathology**

**Discussion:**
Laminar necrosis results from hypoxic/ischemic injury as may occur in a large variety of conditions producing ischemia or hypoxia. Multiple sclerosis may involve the cortex with focal demyelination best seen with myelin stains. Marchiafava-Bignami disease is necrosis of the corpus callosum initially associated with consumption of red wine but subsequently seen in malnourished, chronically ill individuals. Hyponatremia is related to central pontine myelinolysis.

**Reference:**

**Question(s) 175: Pathology**

**Discussion:**
Angioinvasive branching fungal hyphae are characteristic of aspergillosis and mucormycosis. In contrast, in blastomycosis, coccidioidomycosis, cryptococcosis, and histoplasmosis rounded yeast forms are seen.

**Reference:**

**Question(s) 176: Behavioral**

**Discussion:**
Contusion of the orbitofrontal cortex is associated with social disinhibition. Apathy, depression and loss of task set is more commonly seen in dorsolateral prefrontal lesions. Akinetic mutism is more commonly associated with medial frontal lesions.

**Reference:**

**Question(s) 177: Anatomy**

**Discussion:**
The pedunculopontine nucleus (PPN) is an integral component of the motor circuit of the basal ganglia. It receives inputs from the globus pallidus and subthalamic nucleus and projects to reticulospinal medullary neurons that control locomotion. There is depletion of PPN neurons in Parkinson's disease.

**Reference:**

**Question(s) 178: Pharmacology/Chemistry**

**Discussion:**
The hypocretin/orexin system of the lateral hypothalamus has been implicated in the mechanisms of narcolepsy. These neurons project to cholinergic and monoaminergic cell groups involved in regulation of REM sleep. Reduced CSF levels of hypocretin and reduced numbers of hypocretin cells have been found in patients with narcolepsy.

**Reference:**

**Question(s) 179: Behavioral**

**Discussion:**
Problems with interpersonal relationships, fear of abandonment, waxing and waning between idealizing and devaluing people close to them, impulsive outbursts of anger, suicidal gestures, associated depression, and often dissociative states are all commonly seen in the patient with borderline personality disorder.

**Reference:**
Question(s) 180: Clinical Adult
Discussion:
Human prion diseases include kuru, sporadic Creutzfeldt-Jakob disease, iatrogenic Creutzfeldt-Jakob disease, new variant Creutzfeldt-Jakob disease, Gerstmann-Straussler-Scheinker disease, and fatal familial insomnia.

Reference:

Question(s) 181: 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) 182: Pathology
Discussion:
Critical care myopathy is an under-recognized disorder characterized by selective loss of myosin from myofibers. Also known as myosin losing myopathy, this disorder is most commonly seen in critically ill patients treated with corticosteroids and neuromuscular blockers.

Reference:

Question(s) 183: Physiology
Discussion:
Rapid, irregular, asynchronous movements of the legs and trunk occurring while standing is called orthostatic tremor or "shaky legs syndrome". This disorder usually occurs in middle-aged or elderly people and is characterized by feelings of unsteadiness in the legs and fear of falling when standing. Walking is normal and symptoms are abolished by walking or sitting.

Reference:

Question(s) 184: 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.

Reference:
Question(s) 185: Physiology
Discussion:
Low frequency sounds are best picked up by the apex of the basilar membrane.

Reference:

Question(s) 186: Pharmacology/Chemistry
Discussion:
There are several families of neuroactive peptides, that include the neurohypophyseal hormones (vasopressin, oxytocin, neurophysin), opioids (opiocortins, enkephalins, dynorphins, FMRFamide), tachykinins (substance P, physalaemin, kassinin, uoperolein, eledoisin, bombesin, substance K), secretins (secretin, glucagon, VIP, GIP, GHRF, peptide histidine isoleucineamide), insulins (insulin, IGF 1 and 2), somatostatins (somatostatins, pancreatic polypeptide) and gastrins (gastrin, chlecystokinin).

Reference:

Question(s) 187: Clinical Pediatrics
Discussion:
Infantile botulism usually presents between 3 and 18 weeks of age. The disease is caused by the C. botulinum toxin which blocks acetylcholine release. Clinical features include constipation, hypotonia, areflexia, poor suck, impaired pupillary response to light and ophthalmoplegia. The infants are often breast-fed. Diagnosis is made by EMG with repetitive nerve stimulation, causing an incremental response and isolation of C. botulinum toxin in the stool.

Reference:

Question(s) 188: Physiology
Discussion:
Children who develop infantile botulism typically are normal at birth and develop normally until the second to fifth month of life. Hypotonia then develops, accompanied by constipation. On examination the patient is quite weak and areflexic. Compound muscle action potential recording in response to 50 Hertz stimulation produces a diagnostic incrementing response.

Reference:

Question(s) 189: Clinical Adult
Discussion:
Patients that have suffered severe head injury with basilar skull fracture are at increased risk for the development of diabetes insipidus (DI). Serum sodium should be followed carefully in patients who are potential organ donors following severe head injury since failure to recognize and treat DI can render donor organs unusable due to damage from hypotension and hypernatremia.

Reference:

Question(s) 190: Pathology
Discussion:
In tabes dorsalis, the degeneration is confined to the dorsal columns, whereas in AIDS myelopathy, amyotrophic lateral sclerosis, Friedreich's ataxia, and pernicious anemia, degeneration occurs in other tracts.

Reference:
Question(s) 191: Neuroradiology
Discussion:
The posterior cerebral artery supplies parts of the temporal lobe, parietal lobe, occipital lobe, part of the thalamus, midbrain, choroid plexus and ependyma of third and lateral ventricles, but not the globus pallidus.

Reference:

Question(s) 192: Pharmacology/Chemistry
Discussion:
Ethambutol is the drug most frequently associated with toxic optic neuropathy. Intoxication most commonly occurs with dosages of at least 25 mg/kg/d, and may be more common in patients with renal tuberculosis. Visual loss is insidious and usually symmetric. Yellow-green dyschromatopsia is the earliest symptom. There is no treatment other than withdrawal of the drug.

Reference:

Question(s) 193: Behavioral
Discussion:
Prosopagnosia (in which visual perception is intact but there is an impaired ability to recognize the identity of the perceived figure by vision alone) is nearly always associated with bilateral lesions involving the occipitotemporal junctional area.

Reference:

Question(s) 194: 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.

Reference:

Question(s) 195: Pharmacology/Chemistry
Discussion:
Mirtazapine is a new antidepressant drug that blocks the serotonin 5HT2A and the histamine H1 receptors. Blockade of H1 receptors accounts for sedation, drowsiness, and weight gain that occur with this and other antidepressants such as amitriptyline and doxepin.

Reference:

Question(s) 196: Behavioral
Discussion:
Korsakoff amnestic syndrome causes impairment in declarative memory (anterograde amnesia) and forgetting of recent events (retrograde amnesia) with sparing of motor memory and semantic memory (memory for meaning of words). Digit span remains normal in this syndrome.

Reference:
Question(s) 197: Physiology
Discussion:
Periodic limb movements of sleep (nocturnal myoclonus) occur during light sleep.

Reference:

Question(s) 198: Behavioral
Discussion:
Depression is the most common psychiatric disorder in epilepsy; in one study depression accounted for 80% of psychiatric hospital admissions of epileptics.

Reference:

Question(s) 199: Neuroradiology
Discussion:
Enhancing meninges is a nonspecific finding, and can be seen with a variety of findings, including granulomatous diseases of the meninges, meningitis (usually bacterial more than viral), prior hemorrhage or surgery, metastatic meningeal disease and idiopathic intracranial hypotension. It is not seen in pseudotumor cerebri, in which imaging studies are typically normal.

Reference:

Question(s) 200: Pathology
Discussion:
Choline acetyltransferase is the synthetic enzyme for acetylcholine, and is a marker of the axonal termini of cholinergic neurons. The neurons of the basal nucleus are the major cholinergic projections to the cerebral cortex, and with depopulation of this projection nucleus, there is a corresponding reduction in cortical choline acetyltransferase.

Reference:

Question(s) 201: Pathology
Discussion:
Malignant hyperthermia is a potential complication seen in patients with central core myopathy associated with the administration of certain anesthetic drugs such as succinylcholine and halothane.

Reference:

Question(s) 202: Physiology
Discussion:
The short head of the biceps femoris muscle is the only muscle proximal to the knee to receive innervation via the peroneal division of the sciatic nerve.

Reference:

Question(s) 203: Physiology
Discussion:
Large motor unit potentials result from collateral sprouting.

Reference:
**Question(s) 204:** Anatomy  
**Discussion:**  
The dorsal columns of the spinal cord constitute part of a highly specific sensory pathway with respect to place.

**Reference:**  

**Question(s) 205:** Clinical Adult  
**Discussion:**  
About 1% of patients with strokes due to vertebrobasilar occlusive disease have sudden bilateral hearing impairment.

**Reference:**  
Huang MH, Huang CC, Ryu SJ, et al.  

**Question(s) 206:** Pathology  
**Discussion:**  
Rosenthal fibers are opaque, homogeneous, eosinophilic structures composed of compressed GFAP intermediate filaments that are commonly associated with slow growing low grade tumors, including pilocytic astrocytoma, pleomorphic xanthoastrocytoma, and ganglion cell tumors. They may be seen adjacent to any chronic compressive process and are abundant in Alexander disease (which in a large number of cases occurs secondary to a GFAP gene mutation).

**Reference:**  

**Question(s) 207:** Physiology  
**Discussion:**  
The P-9 of the somatosensory evoked potential originates in the brachial plexus.

**Reference:**  

**Question(s) 208:** Pathology  
**Discussion:**  
Hallervorden-Spatz disease is a progressive neurodegenerative disease with neuroaxonal dystrophy, rust-brown discoloration of globus pallidus and pars reticularis of substantia nigra due to accumulation of iron-containing pigment, and an onset usually before age 15 years. Nasu-Hakola disease is characterized by repeated bone fractures, bone cysts, and mineralization of the basal ganglia.

**Reference:**  

**Question(s) 209:** Clinical Adult  
**Discussion:**  
The most common cognitive deficit after a closed head injury is impairment of memory.

**Reference:**  

**Question(s) 210:** 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.

**Reference:**  
Question(s) 211: 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) 212: Anatomy

Discussion:
A lateral medullary stroke spares the hypoglossal nucleus, medullary pyramid and the medial longitudinal fasciculus.

Reference:

Question(s) 213: 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) 214: 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) 215: Pharmacology/Chemistry

Discussion:
Astrocytes contain glutamine synthase, an ATP-dependent enzyme that utilizes glutamate and ammonia (NH3) to produce glutamine. Therefore, astrocytes have two important functions (1) detoxification of NH3; and (2) providing glutamine to the neurons as a substrate for glutamate production by action of glutaminase.

Reference:

Question(s) 216: Physiology

Discussion:
Triceps, anconeus and pronator teres (C6-C7) have significant contributions from the C-7 root.

Reference:
Question(s) 217: Pathology

Discussion:
Anencephaly is convincingly associated with maternal folic acid deficiency and reduced rates of this congenital abnormality occur in women given folic acid. The thymus is enlarged in anencephalics due to lack of adrenal driven involution, which in turn results from absence or derangement of the hypothalamic-pituitary axis. The risk of anencephaly in subsequent pregnancies is 5-7%, females are more commonly afflicted, and there is no compelling evidence of a role of maternal exposure to toxins in the genesis of this condition.

Reference:

Question(s) 218: 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) 219: Behavioral

Discussion:
Nefazodone does not suppress REM sleep. It actually may increase REM sleep.

Reference:

Question(s) 220: Behavioral

Discussion:
The apolipoprotein E (APOE) genotype associated with the greatest risk of developing Alzheimer's disease is the 4/4 allele.

Reference:
Question(s) 224: 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) 225: Behavioral
Discussion:
Catatonia may be seen in a number of medical, neurological, and psychiatric conditions. Of the psychiatric causes, bipolar disorder is most common. Patients may present with a host of clinical features including akinesia, mutism, catalepsy, waxy flexibility, echopraxia and echolalia, utilization behavior, and despite extreme negativism may manifest automatic obedient behavior. When the patient is severely impaired, ECT is a treatment of choice. IV benzodiazepines such as lorazepam may also be beneficial.

Reference:

Question(s) 226: 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) 229: Physiology

Discussion:
The utricle and saccule of the vestibular system are associated with linear acceleration.

Reference:

Question(s) 230: Behavioral

Discussion:
Hypoperfusion in the left internal carotid artery distribution causes watershed infarcts affecting the borderzone area of the middle and anterior cerebral arteries, and the posterior borderzone between the middle and posterior cerebral arteries. The patient's aphasia is a mixed transcortical aphasia, manifested by absent spontaneous speech and impaired comprehension. Intact repetition is secondary to sparing of the presylvian area.

Reference:

Question(s) 231: Behavioral

Discussion:
Factitious disorder is defined as a syndrome of intentional production of psychological or physical symptoms in the absence of external incentives but in the presence of a psychological need to assume the sick role. When there are external incentives for the behavior then "malingering" is the likely diagnosis.

Reference:

Question(s) 232: Pharmacology/Chemistry

Discussion:
Like other projection neurons in relay systems of the brain, neurons of the nucleus gracilis projecting to the thalamus use L-glutamate.

Reference:

Question(s) 233: 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:

Question(s) 234: Anatomy

Discussion:
In the developing neural tube, the basal plate becomes anterior horn gray matter and the alar plate becomes the posterior horn gray matter.

Reference:
**Question(s) 235: Physiology**
**Discussion:**
3 Hz slow wave activity can be seen as a normal finding during hyperventilation in a child. Simple absence seizures are associated with 3 Hz spike and wave discharges during hyperventilation.

**Reference:**

**Question(s) 236: Anatomy**
**Discussion:**
Normal CSF pressure (lumbar) measured in recumbent position is 10 to 15 cm H2O.

**Reference:**

**Question(s) 237: Pharmacology/Chemistry**
**Discussion:**
Apoptosis is a mechanism of cell death that may occur in many neurodegenerative disorders. It depends on activation of caspases and is characterized by chromatin condensation and cell desintegration in the absence of inflammation. Apoptosis may be triggered by exogenous signals such as activation of "death receptors" by the Fas-ligand, by p53 in response of DNA damage, or by release of mitochondrial cytochrome c, which activates an apoptosis activating factor (Apaf-1) and then the caspase cascade. Release of cytochrome c is inhibited by Bcl-2, one of the antiapoptotic members of the Bcl-2 family. Binding of the proapoptotic member Bax to Bcl-2 prevents its protective effect and triggers apoptosis.

**Reference:**

**Question(s) 238: Physiology**
**Discussion:**
Juvenile myoclonic epilepsy typically begins in adolescence. The EEG is characterized by frontally predominant, 4-6 Hz generalized spike-and-wave discharges. Myoclonic and generalized seizures are the predominant seizure types. Absence seizures occur in approximately one-third of patients. The disorder is often familial, and patients are cognitively normal.

**Reference:**

**Question(s) 239: Physiology**
**Discussion:**
With acute (less than 1 week) nerve root compression, the only EMG abnormality may be a reduced recruitment of motor unit potentials.

**Reference:**

**Question(s) 240: Pathology**
**Discussion:**
When nerve cells undergo rapid death, the cell bodies and proximal dendrites are phagocytized by cells derived from microglia, which give rise to macrophages.

**Reference:**
Question(s) 241: Pharmacology/Chemistry
Discussion:
Nifedipine and other calcium channel blockers can cause increased weakness in myasthenic patients. A variety of drugs induce or worsen myasthenic symptoms through actions on pre- or post-synaptic mechanisms. Certain antibiotics impair transmitter release by interfering with calcium flux. Other drugs affect metabolism of acetylcholine or cholinergic receptor function. Bromocriptine, chloramphenicol, fluoxetine and gabapentin do not adversely affect the myasthenic patient.

Reference:

Question(s) 244: Physiology
Discussion:
Polymorphic delta activity is produced by processes involving subcortical white matter.

Reference:

Question(s) 245: Clinical Adult
Discussion:
Aseptic meningitis, cranial neuritis, and radiculoneuritis are the most characteristic neurologic complications of the second stage of Lyme disease.

Reference:

Question(s) 246: Pathology
Discussion:
Paraneoplastic cerebellar degeneration is most commonly associated with ovarian carcinoma and some breast tumors. It can also be seen with Hodgkin's lymphoma.

Reference:

Question(s) 247: Physiology
Discussion:
Intermittent generalized slow wave bursts can be seen as a normal feature of the EEG in children during drowsiness and hyperventilation.

Reference:
Question(s) 248: Behavioral
Discussion:
Olanzapine is an atypical antipsychotic that frequently causes significant weight gain. Quetiapine, risperidone, haloperidol, and molindone are less likely to do so.

Reference:

Question(s) 249: Physiology
Discussion:
Renshaw cells are interneurons that have a direct input to alpha motoneurons.

Reference:

Question(s) 250: Pathology
Discussion:
Meningiomas are associated with monosomy 22, leading to loss of a tumor suppressor gene (merlin). Even when the genetic loss is not as gross as loss of an entire chromosome, there is a loss of heterozygosity for this locus. Meningiomas, especially spinal, are much more common in women. They constitute one fourth to one third of all primary intraspinal tumors. There is a marked gender asymmetry in meningiomas of the spinal cord with a 10-20:1 female:male ratio.

Reference:

Question(s) 251: Pharmacology/Chemistry
Discussion:
Mutations of the FIL-1 gene in chromosome Xq28, encoding for filamin-1, produce periventricular heterotopy. This reflects a complete failure of neuronal migration that may cause seizures. It is dominant in females and lethal in males. Filamin-1 interacts with actin and is critically involved in the early phases of migration.

Reference:

Question(s) 252: Clinical Adult
Discussion:
The classic syndrome of vitamin A toxicity is pseudotumor cerebri, with headache, papilledema, and occasionally abducens palsy.

Reference:

Question(s) 253: Physiology
Discussion:
The most common EEG finding in patients who have narcolepsy is sleep onset REM sleep.

Reference:

Question(s) 254: 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) 255: 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) 256: 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. The pathogenic relationship between Notch-3 mutation and development of arteriopathy is still undetermined.

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

Question(s) 257: Clinical Adult

Discussion:
Selective impairment of vertical eye movement is most characteristic of a lesion in the midbrain.

Reference:

Question(s) 258: Clinical Adult

Discussion:
Autoantibodies to GAD (glutamic acid decarboxylase) may occur with stiff man (stiff person) syndrome.

Reference:

Question(s) 259: Physiology

Discussion:
Hyperkalemic periodic paralysis is inherited in an autosomal dominant pattern. A distinct familial form has prominent clinical and electromyographic evidence of myotonia as an associated feature.

Reference:

Question(s) 260: Clinical Adult

Discussion:
In order to standardize terminology around the world, the World Health Organization defines impairment as the clinical signs and symptoms produce by damage to the nervous system. Disability is the personal limitation imposed on the activities of daily living by neurological impairment, and handicap is the social or environmental effects of the disability or impairment.

Reference:
Burks JS, Johnson KP. Multiple sclerosis: Medical management and rehabilitation. New York: Demos, 2000;223.
**Question(s) 261: Anatomy**

**Discussion:**
The nucleus tractus solitarius is the site of termination of afferents from the baroreceptors. Involvement of this structure results in paroxysmal hypertension resembling a pheochromocytoma.

**Reference:**

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**Question(s) 262: Pathology**

**Discussion:**
The most helpful immunostain for confirming a diagnosis of meningioma is epithelial membrane antigen (EMA). S-100 protein is positive in gliomas and schwannomas. Transthyretin is positive in choroid plexus papillomas.

**Reference:**

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**Question(s) 263: Physiology**

**Discussion:**
Pain is mediated by free nerve endings.

**Reference:**

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**Question(s) 264: Anatomy**

**Discussion:**
The basal nucleus of Meynert is in the substantia innominata and contains clusters of large cholinergic neurons that have widespread projections to the cortex and amygdala.

**Reference:**

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**Question(s) 265: Physiology**

**Discussion:**
The sodium-potassium-ATP pump maintains the resting membrane potential despite passive diffusion of sodium into and potassium out of the cell. Inhibition of this pump would result in an increase in intracellular sodium concentration, a decrease in intracellular potassium and a resulting depolarization of the cell membrane.

**Reference:**

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**Question(s) 266: Clinical Adult**

**Discussion:**
Locked-in syndrome is a state in which quadriplegia prevents the usual means of gestural or verbal communication. Usually the patient can communicate by way of vertical eye movements or blinking and can demonstrate full comprehension of his plight. A sleep wake cycle is often preserved. The most common vascular lesion underlying the locked-in syndrome is basilar artery occlusion with extensive destruction of the pontine base.

**Reference:**
Question(s) 267: Pharmacology/Chemistry

Discussion:
Mutations in the alpha-synuclein gene have been found in some families with autosomal dominant Parkinson’s disease. The abnormal alpha-synuclein is present in the ubiquitin-positive Lewy bodies. Synuclein is a normal protein in the nucleus and synaptic terminals of neurons, but its role is still incompletely defined.

Reference:

Question(s) 268: Pharmacology/Chemistry

Discussion:
Periodic movements of sleep (nocturnal myoclonus) consists 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) 269: Pharmacology/Chemistry

Discussion:
Nerve growth factor, acting via trk A receptors, is critical for survival of nociceptive and sympathetic neurons. Mutations of the Trk A gene are associated with the phenotype of congenital insensitivity to pain and anhidrosis, as seen in hereditary sensory and autonomic neuropathies types IV and V.

Reference:

Question(s) 270: Behavioral

Discussion:
Anosognosia (unawareness of deficit or illness) is usually seen associated with non-dominant parietal lobe lesions. Achromatopsia is found after lesions of the inferior lip of the occipital lobe. Limb kinetic apraxia is seen after lesions of the anterior corpus callosum. Expressive aprosodia is seen after right frontal lesions. Semantic aphasia is seen after dominant hemisphere lesions.

Reference:

Question(s) 271: 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) 272: Physiology

Discussion:
Multifocal motor neuropathy with conduction block presents with slowly progressive, asymmetric weakness, atrophy and reflex loss with normal or minimally abnormal sensation. Nerve conduction studies involving clinically weak muscles demonstrate evidence of partial conduction block, but sensory nerve conduction studies are typically normal.
Reference:

Question(s) 273: 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) 274: Behavioral
Discussion:
Of clozapine, quetiapine, phenelzine, olanzapine, and risperidone the one most likely to produce extrapyramidal side effects is risperidone.

Reference:

Question(s) 275: 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) 276: Pharmacology/Chemistry
Discussion:
Topiramate is a weak inhibitor of carbonic anhydrase, and therefore may increase urinary pH and decrease urinary citrate excretion. Both actions may predispose to kidney stones.

Reference:

Question(s) 277: Behavioral
Discussion:
Good spontaneous and repetitive affective prosody as well as spontaneous gesturing with poor affective comprehension of language and gesture is known as a transcortical sensory aprosodia.

Reference:

Question(s) 278: 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:
Question(s) 279: Behavioral Discussion:
Obsessive-compulsive disorder is often associated with Tourette's syndrome.

Reference:

Question(s) 280: 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) 281: 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) 282: Physiology Discussion:
REM or active sleep accounts for 50% of sleep in neonates.

Reference:

Question(s) 283: Anatomy Discussion:
The dorsomedial (or mediodorsal) nucleus of the thalamus provides innervation to the whole prefrontal cortex, which consists of a paralimbic orbitofrontal region involved in control of affect and behavior, and an associative dorsolateral region involved in executive functions.

Reference:

Question(s) 284: Clinical Adult Discussion:
Myeloma, especially of the osteosclerotic type, has a frequent association with a demyelinating polyneuropathy.

Reference:

Question(s) 285: Physiology Discussion:
Cerebral blood flow is increased most effectively by administration of 5% carbon dioxide.

Reference:
Question(s) 286: Pathology

Discussion:
Shadow plaques in multiple sclerosis were formerly thought to represent areas of partial, arrested, or incomplete demyelination. They are now recognized to be areas of partial remyelination. There is little agreement on any characteristic precursor or nascent lesion in multiple sclerosis; ultrastructural studies fail to detect changes in myelin sheaths or oligodendrocytes that clearly precede the sudden appearance of macrophages in the plaque lesions.

Reference:

Question(s) 287: Anatomy

Discussion:
The pulvinar receives fibers from the superior colliculus and projects to areas 17, 18 and 19. Both the inferior and lateral pulvinar have reciprocal connections with the occipital cortex.

Reference:

Question(s) 288: 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, part A. Continuum 1997 Nov;3(6).

Question(s) 289: 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 unesterified 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:

Question(s) 290: Pathology

Discussion:
It is important to remember that although retinal hemorrhages are well described in non-accidental trauma (most notably in "shaken baby syndrome") there are many other causes of retinal hemorrhages, including following accidental trauma, in coagulopathies, and in newborns after vaginal delivery. Retinal hemorrhage after resuscitation is rare, but has been described.

Reference:
Question(s) 291: 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) 292: 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) 293: Behavioral Discussion:
Patients with obsessive-compulsive disorder have been shown to respond to treatment with certain selective serotonin reuptake inhibitors (SSRIs), including fluoxetine and fluvoxamine.

Reference:

Question(s) 294: Behavioral Discussion:
Chronic alcoholic hallucinosis is characterized by auditory hallucinations with a clear sensorium.

Reference:

Question(s) 295: Clinical Adult Discussion:
A history of abrupt loss of consciousness and examination findings of pinpoint pupils, ocular bobbing, and absent oculovestibular responses would suggest an acute, severe pontine lesion, such as pontine hematoma.

Reference:

Question(s) 296: Behavioral Discussion:
The pathophysiology of schizophrenia as associated with pathological changes in the dorsomedial thalamus and the dorsolateral prefrontal cortex thereby affecting the pathway between the two sites. Any role of pedunculopontine, nigrostriatal, hippocampal-fornical-mamillary, and amygdala-orbitofrontal pathways is not clearly established in schizophrenia if in fact there is a role in these pathways.

Reference:
Question(s) 297: Behavioral
Discussion:
Bupropion has essentially no anticholinergic, sedating, sexual dysfunction, or orthostatic hypotension side effects. Fluoxetine, citalopram, and venlafaxine have a high to very high risk of sexual dysfunction. Mirtazapine is sedating. None of the above medications have anticholinergic or significant orthostatic hypotensive side effects.

Reference:

Question(s) 298: Anatomy
Discussion:
The chemoreceptor trigger zone is located in the area postrema.

Reference:

Question(s) 299: 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) 300: Behavioral
Discussion:
This history depicts a case of herpes simplex virus encephalitis. The treatment of choice is acyclovir.

Reference:

Question(s) 301: Pharmacology/Chemistry
Discussion:
Fetal vitamin K deficiency with hemorrhagic complications occurs in 10% of neonates born from 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) 302: Pathology
Discussion:
Mycotic aneurysms are most commonly found in the distal branches of the middle cerebral artery.

Reference:

Question(s) 303: Clinical Adult
Discussion:
Numerous studies of the cognitive disturbance in multiple sclerosis demonstrate the absence of classic cortical (e.g., language, cognition, visuospatial, memory) impairments but clearly show a slowing of information processing.

Reference:
Question(s) 304: Pharmacology/Chemistry
Discussion:
Streptococcus pneumoniae is a common cause of bacterial meningitis. As several causes of ceftriaxone resistant streptococcus pneumoniae have been reported, the initial treatment in suspect cases should include vancomycin, until the results of drug sensitivities are available.

Reference:

Question(s) 305: 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) 306: Pathology
Discussion:
Spontaneous or traumatic dissections occur most frequently in the carotid artery at the neck. Vertebral artery dissections may occur after chiropractic manipulations of the neck.

Reference:

Question(s) 307: Behavioral
Discussion:
The loss of remote memory including autobiographical memory in the face of intact new learning ability is consistent with psychogenic amnesia.

Reference:
Question(s) 311: 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) 312: Anatomy Discussion:
Sensation to the umbilicus is carried by T10.

Reference:

Question(s) 313: Physiology Discussion:
Ictal EEG is the most important factor in determining the site of seizure onset on chronic, intracranial EEG monitoring.

Reference:

Question(s) 314: Anatomy Discussion:
The optic nerve provides inputs to the superior colliculus (for reflex saccades), lateral geniculate nucleus (relay of the visual pathway), pretectal nucleus (relay of the light reflex) and the suprachiasmatic nucleus (the circadian pacemaker). The medial preoptic nuclei would be spared by enucleation of the eye.

Reference:

Question(s) 315: Clinical Pediatrics Discussion:
In both Duchenne's and Becker's muscular dystrophy the dystrophin gene is affected but the molecular mechanisms differ in the two disorders. Absence of dystrophin in the muscle confirms the diagnosis of Duchenne's muscular dystrophy. Whereas altered or reduced dystrophin in the muscle is consistent with Becker's muscular dystrophy.

Reference:

Question(s) 316: Clinical Pediatrics Discussion:
The cerebrospinal fluid in preterm newborn infants without bacterial meningitis and without other disease can have a protein of 65-150 mg/dl and the white blood cell count can have a range of 0-29 cells/cu mm with a mean of nine.

Reference:

Question(s) 317: Pathology Discussion:
The brain is very sensitive to the effects of carbon monoxide intoxication. A cherry-red color may be seen in many tissues (including the brain) due to the presence of carboxyhemoglobin. With survival of 24-48 hours, petechial hemorrhages may be seen in the white matter along with more significant hemorrhage in the globus pallidus. Cavitation is seen in longer-term survivors.

Reference:
**Question(s) 318:** Physiology
**Discussion:**
Acetylcholine is the neurotransmitter of preganglionic sympathetic and vagal nerve fibers.

**Reference:**

**Question(s) 319:** Clinical Pediatrics
**Discussion:**
The incidence of lamotrigine-associated severe rash is estimated to be one in 1,000 adults and one in 200 children. The risk in children is increased with a rapid increase in dose and when given with valproate.

**Reference:**

**Question(s) 320:** Clinical Pediatrics
**Discussion:**
The movements of Sydenham chorea cannot be suppressed by the patient, whereas tics can be suppressed voluntarily for at least a short period of time. Tics are stereotyped with a limited variety of movements at any one time. In contrast chorea is very variable.

**Reference:**

**Question(s) 321:** Clinical Pediatrics
**Discussion:**
Most cases of newborn infants with myelomeningocele have a Chiari type II malformation. If hydrocephalus develops, overt clinical signs most commonly appear two to three weeks after birth.

**Reference:**

**Question(s) 322:** Pathology
**Discussion:**
Approximately 90-95% of epidural hematomas are associated with a skull fracture. They are usually unilateral, disc-shaped, and are most often due to fractures of the squamous-temporal bone with laceration of the middle meningeal artery.

**Reference:**

**Question(s) 323 - 325:** Clinical Pediatrics
**Discussion:**
The majority of normal children will pick up a small object with a thumb and forefinger (pincer) grasp by one year of age, can walk unsupported by 15 months and when asked, will point to several body parts correctly by two years of age.

**Reference:**
Question(s) 326 - 330: Anatomy
Discussion:
The inferior cerebellar peduncle contains primary and secondary vestibulocerebellar fibers and certain cerebellovestibular (juxtarestiform) fibers from the nodulus, uvula, and fastigial nuclei. The middle cerebellar peduncle carries all pontocerebellar fibers. The superior cerebellar peduncle carries the dentatorubral, dentatothalamic and anterior (ventral) and spinothalamic tracts.

Reference:

Question(s) 331 - 334: Clinical Pediatrics
Discussion:
The skin lesions of tuberous sclerosis include hypopigmented macules, which may require a Wood's lamp to visualize, adenoma sebaceum, shagreen patch, subungal and periungual fibromas. Rocky mountain spotted fever classically has a generalized petechial rash and Lyme disease frequently has erythema migrans as the typical rash. Rheumatic disease has erythema marginatum, subcutaneous nodules, carditis, polyarthritis, and Sydenham chorea as major manifestations.

Reference:

Question(s) 335 - 337: Behavioral
Discussion:
The amygdala is associated with emotional memory; the hippocampus with declarative memory and the pulvinar with visual attention.

Reference:

Question(s) 338 - 340: Physiology
Discussion:
AMPA, kainate and NMDA receptors are ionotropic glutamate receptors. Domoic acid is an agonist at the kainate receptor. NMDA receptor responses are blocked by magnesium ions. Metabotropic glutamate receptors are coupled to second messengers such as phospholipase A, adenylate cyclase and G-proteins.

Reference:

Question(s) 341 - 342: Clinical Adult
Discussion:
14-3-3 protein has been reported in the CSF of 90% of patients with classical Creutzfeldt-Jakob disease. pcr of CSF for Epstein Barr Virus DNA is frequently positive in primary central nervous system lymphoma, particularly in patients with AIDS.

Reference:
Question(s) 343 - 345: 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.

Reference:

Question(s) 346 - 348: Clinical Pediatrics
Discussion:
The Walker-Warburg syndrome is a type II lissencephaly with a congenital muscular dystrophy and has autosomal recessive inheritance. Rett syndrome is characterized by progressive microcephaly and developmental regression in the first two years of life. The disorder occurs almost exclusively in females. Individuals with Angelman syndrome, also known as the "happy puppet" syndrome, have mental retardation, jerky movements, and seizures. The diagnosis can be confirmed by demonstrating a deletion on chromosome 15.

Reference:

Question(s) 349 - 352: Anatomy
Discussion:
The motor nucleus of the trigeminal nerve sends fibers to muscles of mastication. The spinal trigeminal nucleus subserves pain and temperature functions. The mesencephalic nucleus contains primary sensory neurons involved with pressure and kinesthetic sense of teeth, periodontium, hard palate, and joint capsules. The principal (main) sensory nucleus is concerned with tactile and pressure sense of the face.

Reference:

Question(s) 353 - 356: Anatomy
Discussion:
Hemisection of the spinal cord (Brown-Sequard syndrome) results in ipsilateral paralysis and loss of position sense, and contralateral loss of pain sensation below the lesion. The bladder is only temporarily affected.

Reference:

Question(s) 357 - 360: Anatomy
Discussion:
The gluteus medius is innervated by the superior gluteal nerve. The gluteus maximus is innervated by the inferior gluteal nerve. The short head of the biceps femoris is innervated by the peroneal division of the sciatic nerve. The tibialis posterior is innervated by the tibial nerve.

Reference:
Question(s) 361 - 364: Clinical Pediatrics
Discussion:
Leber’s optic atrophy is transmitted via maternal mitochondria. Becker’s muscular dystrophy is inherited in a sex linked recessive fashion. Spinal muscular atrophy is an autosomal recessive disorder. Tuberous sclerosis can be inherited in an autosomal dominant mode but approximately one-half of the cases are due to new mutations.

Reference:

Question(s) 365 - 369: Physiology
Discussion:
Drug-induced muscle disorders include myalgia, myotonia, type 2 atrophy (e.g., prednisone), and focal (e.g., oxycodone), necrotizing, inflammatory (e.g., L-tryptophan), mitochondrial (e.g., zidovudine) or autophagic (e.g., amiodarone) myopathies.

Reference:

Question(s) 370 - 373: Physiology
Discussion:
Serotonin is associated with the dorsal raphe nuclei; norepinephrine is associated with the locus ceruleus; acetylcholine with the basal forebrain and parasympathetic neurons of cranial nerves, and histamine with the hypothalamus.

Reference:

Question(s) 374: Clinical Adult
Discussion:
In an HIV positive patient, an enhancing brain mass, which does not respond to antitoxoplasmosis therapy, is most likely a B-cell lymphoma. Toxoplasmosis is the most common cause of focal intracranial mass lesions in patients with AIDS. Lymphoma is the second most common cause. Less common causes include pyogenic abscess, syphilitic gumma, Candida or Nocardia abscess, cryptococcoses, cryptococcal pseudocyst, other fungal and parasitic infections, and infarction. About 80% of patients with established CNS toxoplasmosis have positive toxoplasma titers in serum, and about 85% of patients with CNS toxoplasmosis have multiple lesions on brain MRI. Thus, patients with negative titers and only a single lesion on MRI should have a stereotactic biopsy, but patients with either positive titers or multiple MRI lesions should be treated empirically with antitoxoplasmosis therapy for 2 to 3 weeks and monitored clinically and radiologically.

Reference:
Question(s) 375: Clinical Adult
Discussion:
Confusion, multiple cerebral lesions, multiple sensory and perceptual deficits indicate a poorer prognosis than hemiparesis alone. Preventive measures for decubitus ulcers, pulmonary embolism, contractures, dysphagia and aspiration are regularly employed in most stroke facilities. Although it is important to treat depression when it is present, prophylactic medication is not indicated.

Reference:

Question(s) 376: 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:

Question(s) 377: Pharmacology/Chemistry
Discussion:
The most common form of disulfiram neurotoxicity is peripheral polyneuropathy which may on occasion affect proximal muscles first, leading to potential misdiagnosis as myopathy. The neuropathy typically appears 5-6 months after institution of disulfiram treatment. Disulfiram-induced CNS toxicity is less common. A progressive neuropathy first appearing 6 months after stopping alcohol consumption is unlikely to be alcohol-induced. Although thiamine deficiency can result in neuropathy, it should not occur on thiamine replacement unless there is some block of intestinal absorption. Vitamin E deficiency can produce posterior column, spinocerebellar, and peripheral nerve dysfunction, but this also would not occur in the face of replacement therapy unless a malabsorption process is present. Abetalipoproteinemia (Bassen-Kornzweig disease) is a process where malabsorption results in severe Vitamin E deficiency. Thiamine is an important coenzyme for a number of cellular reactions, including those responsible for ATP generation. The combination of disulfiram and alcohol produces the "Antabuse syndrome" or "acetaldehyde syndrome", characterized by the unpleasant combination of vasodilatation of the face and body, headache, tachycardia, respiratory difficulty, vomiting, weakness, and hypotension. It is probably produced by buildup of acetaldehyde due to inhibition of ethanol metabolism.

Reference:
**Question(s) 378 - 380: 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-wave discharges and imaging studies of the brain are normal. The initial drug of choice is ethosuximide or valproate.

**Reference:**

**Question(s) 381 - 383: Clinical Pediatrics**

**Discussion:**
Congenital myotonic dystrophy is inherited from the mother, who may be relatively asymptomatic. EMG and muscle biopsy of the baby is not helpful. EMG of the mother can support the diagnosis. The diagnosis of the baby is confirmed by finding an abnormal expansion of a trinucleotide repeat on chromosome 19. Children with congenital myotonic dystrophy have delayed motor milestones. Mental retardation is common.

**Reference:**

**Question(s) 384: Clinical Pediatrics**

**Discussion:**
Children, who have simple febrile seizures, have a 2-4% risk of later, unprovoked non-febrile seizures (epilepsy). The risk is 7% when these patients are followed to age 25 years.

**Reference:**

**Question(s) 385: Behavioral**

**Discussion:**
Lewy Body disease is a progressive dementing illness manifested by combined cortical and subcortical clinical signs and symptoms, and fluctuating episodes of acute confusion. Psychosis with delusions and visual hallucinations are common. Parkinsonism and gait disturbance are prominent. Deficits in memory, executive function, visual spatial and language are usual. Patients with this disorder have significant susceptibility to adverse effects of even low doses of neuroleptics. Lewy body disease patients with hallucinations demonstrate a relatively greater reduction in choline acetyltransferase activity compared to those patients without hallucinations.

**Reference:**

**Question(s) 386: 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) 387:
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) 388:
Pharmacology/Chemistry
Discussion:
Temporal arteritis typically affects the elderly and is ushered in by headaches. Jaw claudication is a characteristic symptom and low grade fever is often present. The erythrocyte sedimentation rate is usually markedly elevated. Treatment of temporal arteritis consists of high dose prednisone therapy. Low dose therapy will not protect from temporal arteritis-induced blindness.

Reference:

Question(s) 389 - 390:
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 angiookeratomas 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) 391 - 393:
Pharmacology/Chemistry
Discussion:
Dimethylmercury is extremely toxic, and may be absorbed transdermally, or by inhalation. Latex gloves are not protective. Dimethylmercury is rapidly converted to methylmercury, which has a half-life of about 78 days in humans. Long latent periods can occur between exposure and the onset of symptoms in humans and monkeys. Pathologically, the injury involves the cerebral cortex, and cerebellum. Neuronal necrosis and gliosis is seen. Chelation therapy with succimer will markedly increase elimination of mercury from the body, but may not be clinically beneficial unless it is begun soon after exposure. Arsenic exposure produces severe gastrointestinal symptoms with or without encephalopathy acutely, and a progressive polyneuropathy with cutaneous changes, anemia and jaundice with chronic exposure. Cisplatinum may produce an acute encephalopathy and a more chronic cerebellar syndrome with peripheral neuropathy and renal impairment. Lead poisoning in adults occurs with industrial exposures to lead salts, and in conjunction with inhalation of gasoline fumes (tetraethyl and tetraneethyl lead). A delirium may develop. Organic lead poisoning is usually reversible. Thallium exposure is now commonly associated with ingestion of rodenticides. It may produce a lethal encephalopathy acutely, or a painful progressive sensorimotor polyneuropathy with optic atrophy and ophthalmoplegia. Diffuse alopecia occurs two to four weeks after ingestion.

Reference:

Question(s) 394: 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) 395: 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.
**Question(s) 396: Neuroradiology**

**Discussion:**
Axial T2 weighted image shows a diffuse high signal intensity throughout all of the white matter that is shown within the semi-centrum 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:**
Question(s) 399: 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) 400: Neuroradiology
Discussion:
The lesion in the right paracentral lobule is likely to be demyelinating. It affects the white matter, but the ring of enhancement opens when it touches the cortex of the medial aspect of the hemisphere. The open ring appearance of the ring of enhancement (“open-ring sign”) is characteristic of demyelinating lesions. Histology was confirmed by biopsy. Glioblastoma, lymphoma and abscess tend to have a closed ring of enhancement. Astrocytoma generally does not enhance.

Reference:

Question(s) 401: Neuroradiology
Discussion:
The lesion lies in the motor cortex, immediately anterior to the Rolandic (central) sulcus. Therefore, the correct answer is frontal lobe.

Reference:

Question(s) 402: Neuroradiology
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:
Question(s) 403: Neuroradiology
Discussion:
The AP and lateral contrast myelogram demonstrates narrowing of the dural sac at the disc space of multiple levels. The narrowing of the dural sac is limited to the disc space consistent with spinal stenosis which is the combination of disc herniation/bulge and degenerative changes of the posterior elements. Arachnoiditis would not be limited to the disc space. The roots are well seen without clustering. AV malformation is usually associated with tortuous, tangle of vessels. The spinal ependymoma in this region usually occurs at the end of conus. In this particular patient, there is no evidence for neoplastic lesions.

Reference:

Question(s) 404: Neuroradiology
Discussion:
Axial T2-weighted image shows a symmetric brain with normal size ventricles. The frontal lobe cortex is flat, and the white matter that is hypointense does not show the normal interdigitation into the gray matter in this region. Thus the cortex is thick, and the patient has pachygyria. Such an abnormality develops during the period of neuronal migration, occurring during the second trimester.

Reference:

Question(s) 405: Neuroradiology
Discussion:
The middle cerebral artery and its branches are absent. The anterior cerebral artery and the posterior communicating and posterior cerebral artery are present.

Reference:

Question(s) 406: Neuroradiology
Discussion:
The patient was treated for an obstructive hydrocephalus by means of a ventriculo-peritoneal shunt. The shunt track can be seen in the right peri-atrial white matter and the shunt can be seen entering the atrium of the lateral ventricle.

Reference:

Question(s) 407: 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) 408: Neuroradiology
Discussion:
The small and medium sized vessels are ragged and demonstrate the segmented pattern of vasculitis. While vasospasm is possible, this typically affects larger vessels than those affected in this case. This appearance is known as "beads on a string".

Reference:
Question(s) 409: Pathology

Discussion:
The characteristic inclusion body of rabies (Negri body) is classically seen in large pyramidal neurons 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) 410: Anatomy

Discussion:
The photomicrograph showed numerous subpial corpora amylacea. Corpora amylacea are polyglucosan bodies that accumulate with age in astrocyte cytoplasmic processes. They are most numerous in the subpial and perivascular regions. Among the other answer choices, Cowdry A inclusions are brightly eosinophilic intranuclear inclusions seen in several different viral infections, including herpes simplex, varcella zoster, cytomegalovirus and subacute sclerosing panencephalitis; Hirano bodies are oval-to-oblong eosinophilic inclusions seen in the hippocampus in Alzheimer's disease (and to a lesser extent in some normal aged individuals); and Negri bodies are eosinophilic cytoplasmic inclusions seen in rabies.

Reference:

Question(s) 411: Neuroradiology

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) 412: Neuroradiology

Discussion:
Both of the MR images are post-injection of gadolinium DTPA. Both images show separate and discrete ring-enhancing lesions, one in the right occipital pole, and one in the left frontoparietal region. Mass effect is clearly demonstrated on the left side with compression and displacement of the ventricular system. The right occipital lesion has a thin wall, while that on the left is somewhat thicker and more irregular. Both lesions arise at the corticomedullary junctures. Given the age of the patient, clinical presentation, and the appearance of the two lesions, multiple brain abscesses would be the most likely diagnosis. Infarcts tend to be more cortical, demyelinating diseases, more periventricular and more homogeneous in enhancement, occult vascular malformations may enhance, but it is the core of the lesion, and not marginal enhancement, while resolving hematomas would have central high signal intensity on a T1-weighted image.

Reference:
Question(s) 413: Clinical Adult Discussion:
Hereditary motor and sensory neuropathy, type I (HMSN - 1) is characterized by marked slowing of nerve conduction velocities. It is most often inherited in an autosomal dominant pattern, with variable (but nearly complete) expressivity. The age of onset is variable; family members who appear to be unaffected may simply be pre-symptomatic, and may have slow nerve conduction velocities.

Reference:

Question(s) 414: Anatomy Discussion:
The figure shows neurohypophyseal tissue obtained by transphenoidal biopsy of the pituitary. The arrows point to Herring bodies. Herring bodies are axonal storage sites for the hormones oxytocin and vasopressin.

Reference:

Question(s) 415: Neuroradiology Discussion:
Chiari type II malformation is associated with significant beaking or bending of the brainstem, not evident in the current case. The Dandy-Walker malformation occurs in the midline and typically includes maldevelopment of the cerebellar vermis with separation of the cerebellar hemispheres. There are three types of hemangioblastomas, two of which may contain cysts. This cyst is contiguous with the fourth ventricle, and resulted from surgical excision of a cerebellar astrocytoma. Arachnoid cysts occur extrinsic to the brain, rather than within the ventricular system.

Reference:

Question(s) 416: Neuroradiology Discussion:
The findings are consistent with glioblastoma. Glioblastomas have characteristic imaging features on postcontrast studies, which show intense, inhomogeneous, nodular, ring-like enhancement, which encloses a central isointense necrotic core, and delineates the gross tumor margin. Anaplastic astrocytoma, fibrillary astrocytoma, meningioma, and oligodendroglioma tumors typically do not show this bizarre ring-like enhancement.

Reference:

Question(s) 417: Anatomy Discussion:
The filum terminale is the terminal continuation of the spinal cord beginning at the lower end of the conus medullaris. It is composed principally of collagen from the pia mater and blood vessels. In addition, small nerve fascicles are present (whose investing Schwann cells occasionally give rise to schwannomas) and, most importantly, a remnant of the spinal cord central canal is also present, usually in the form of an eccentrically-placed cluster of ependymal cell nests and rosettes. The significance of this central canal remnant lies in the fact that these are the cells of origin of myxopapillary ependymomas, which arise almost exclusively from the conus and filum.

Reference:
Question(s) 418: Physiology
Discussion:
Focal conduction block of compound muscle action potentials from demyelination can be seen in chronic inflammatory demyelinating polyneuropathy, causing decreased amplitudes without axonal involvement always occurring.

Reference:

Question(s) 419: Neuroradiology
Discussion:
High density mass within parietal lobe with marked associated edema consistent with intraparenchymal hematoma.

Reference:

Question(s) 420: 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) 421: Neuroradiology
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. Biswnanger’s disease occurs in the adult population due to chronic cerebrovascular disease.

Reference:

Question(s) 422: Neuroradiology
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) 423: Neuroradiology
Discussion:
The most likely diagnosis is autosomal dominant spinocerebellar ataxia type 6 (proved by genetic testing). These patients have pure cerebellar atrophy, affecting predominantly the superior vermis. There is no pontine atrophy, a feature of both olivo-ponto-cerebellar atrophy and Machado Joseph disease. There is no downword displacement of the cerebellar tonsils, and the posterior fossa is not smaller than normal, ruling out Chiari type I malformation. Finally, the cerebellar atrophy results in a widened supracerebellar cistern, but without any evidence of cerebellar compression, which would be present with a subarachnoid cyst.

Reference:
Question(s) 424: Neuroradiology
Discussion:
The location and dense contrast enhancement strongly suggest an anterior communicating artery aneurysm. Dolichectasia usually results in fusiform aneurysm formation, most commonly in the posterior circulation.

Reference:

Question(s) 425: Neuroradiology
Discussion:
T2 weighted axial and sagittal images are shown. There is abnormally increased signal intensity in the parasagittal parietal and occipital lobes, extending into the splenium of the corpus callosum, where it crosses the midline. This is typical appearance of an infiltrating neoplasm, most commonly astrocytoma. Posterior cerebral artery infarct can involve the hemisphere and splenium, but would not cross the midline into the vascular territory of the right PCA. Schilder’s disease is a demyelinating process and would not involve gray matter as is seen in this case. There are no abnormal vessels present to suggest AVM. An acute hematoma has the opposite signal characteristics than in this case, with hypointensity seen on both T1 and T2 weighted images.

Reference:

Question(s) 426: Neuroradiology
Discussion:
Marked enlargement of the lateral and third ventricles is noted with small sulci indicating obstructive (non-communicating) hydrocephalus which may be seen in aqueductal stenosis. Changes in the CSF spaces are not seen to indicate colpocephaly or schizencephaly. Chiari type I is characterized by syringomyelia. Ex-vacuo hydrocephalus refers to ventricular enlargement due to brain atrophy, with concurrent sulcal/fissure expansion, not seen in this case.

Reference:

Question(s) 427: Neuroradiology
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) 428: 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) 429: Pathology
Discussion:
Lacunar infarcts, as seen in the gross photograph in the thalamus, result from arteriolar hyalinization caused by long-standing hypertension.

Reference:

Question(s) 430: Pathology
Discussion:
The biopsy of a right parietal lobe mass showed a tumor composed of cells with monotonous round nuclei surrounded by prominent perinuclear halos (“fried egg” appearance). These features are characteristic of oligodendroglioma. Of the other choices listed, ependymomas and astroblastomas characteristically show perivascular pseudorosettes, with stout non-tapering cytoplasmic processes in the latter. Fibrillary astrocytomas and gemistocytic astrocytomas show eosinophilic cytoplasm, rather than the clear perinuclear halos of oligodendrogliomas, with multiple elongated cytoplasmic processes in fibrillary astrocytomas and large rounded globular cytoplasm in gemistocytic astrocytomas.

Reference:

Question(s) 431: Neuroradiology
Discussion:
Hypertensive encephalopathy, a cause of reversible posterior leukoencephalopathy syndrome (RPLS), is a rapidly evolving condition that develops in patients with malignant hypertension. If not promptly recognized and aggressively treated it leads to progressive neurologic deterioration and death. Renal failure and eclampsia are contributory causes. Similar changes are also seen in toxicity due to immunosuppressive agents. MRI and CT shows edema located in the occipital and parietal lobes, and other posterior regions. These findings are seen in 85% of patients studied within 72 hours after onset of symptoms. The relative lack of sympathetic innervation in vertebrobasilar vasculature may predispose this region to the development of the cerebral edema.

Reference:

Question(s) 432: Neuroradiology
Discussion:
Axial image demonstrates small, constricted spinal canal. This is mainly due to degenerative facet joint disease. No disc herniation or synovial cyst is present. The nerve roots are coiled above the narrow area, since they cannot slide freely though the narrow part. Distal nerve roots are stretched. The correct answer is spinal stenosis.

Reference:

Question(s) 433: Neuroradiology
Discussion:
This abnormality is clearly a herniated disc. The herniated material is contiguous with disc material (non-sequestered).

Reference:
**Question(s) 434: Pathology**  
**Discussion:**  
The photomicrograph of the lumbar cistern mass shows large perivascular cuffs of mucin with adjacent ependymal cells. These features are diagnostic of the myxopapillary ependymoma, which arises from ependymal remnants of the central canal in the filum terminale.

**Reference:**  

**Question(s) 435: Neuroradiology**  
**Discussion:**  
Contrast enhanced CT scan demonstrates a large, lobulated, densely enhancing structure medial to the Sylvian fissure, with a large serpiginous structure leading from the lesion posteriorly towards the vein of Galen. This represents the nidus and a large draining vein of an arteriovenous malformation. Although AVM's can be easily diagnosed with enhanced CT and with MRI, angiography remains a necessity for complete evaluation of these lesions. Selective injection of the intracerebral vessels allows precise delineation of the feeding arteries and draining veins, something that cannot be determined on MRA or digital subtraction venous angiography - which are non-selective studies. Retrograde venography would not provide any information of the arterial supply to the lesion.

**Reference:**  

**Question(s) 436: Neuroradiology**  
**Discussion:**  
The correct diagnosis is "communicating hydrocephalus." Dilation of the Sylvian fissure and other sulci may suggest atrophy. However, this feature is not uncommon in shunt-responsive hydrocephalus, as documented in the references. On the coronal images note that the sulci at the high parietal convexity are compressed, as compared to the markedly dilated posterior extent of the Sylvian fissure.

**Reference:**  

**Question(s) 437: Neuroradiology**  
**Discussion:**  
This patient had documented E. faecalis osteomyelitis affecting the L2 and L3 vertebrae. There is a ring-enhancing lesion, affecting both of these vertebrae, rather than simply the disc. Thus, there is more pathology than can be explained simply by discitis or expanding L3 neoplasm. There is no evidence of an anterior or posterior spinal approach to suggest surgery. The expanding quality and ring enhancement are not expected with hematoma and compression fracture.

**Reference:**  

**Question(s) 438: 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.
**Question(s) 439:** Neuroradiology  
**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:**

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**Question(s) 441:** Pathology  
**Discussion:**
The astrocyte illustrated in the photomicrograph is a Creutzfeldt astrocyte. These reactive astrocytes are characterized by multiple small nuclei (micronuclei) and abundant eosinophilic cytoplasm. Although not pathognomonic, they are typically seen in demyelinating diseases and serve as a red flag to alert the pathologist to consider demyelinating disorders in the differential diagnosis. There are a number of other types of astrocytes with distinctive morphology. The Bergmann astrocytes of the cerebellar cortex send long cytoplasmic processes from the Purkinje cell layer to the pial surface and show striking proliferation in response to ischemic insult. Alzheimer type II astrocytes can be seen in many types of liver failure that result in hyperammonemia. They differ from all other reactive astrocytes in lacking discernible cytoplasm; rather, the nuclei are enlarged, pale, with only a thin marginal rim of chromatin, and are often irregular in shape. Superficial reactive astrocytosis occurring in the normally very hypocellular molecular layer is referred to as Chaslin’s subpial gliosis. Finally, highly atypical, bizarre reactive astrocytes can be seen in progressive multifocal leukoencephalopathy (caused by infection with the JC virus).

**Reference:**
Question(s) 442: 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 (neuroblastoma-type) rosettes and occasionally fluerettes. 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) 443: Pathology
Discussion:
Infarction with hemorrhage over an extensive area of cortex and in the underlying white matter of the cerebral hemisphere is consistent with sagittal sinus venous thrombosis. The lesion may be either bilaterally symmetrical or prominently asymmetrical, as in the illustrated case. Venous thrombosis is commonly associated with hypercoagulable states and severe dehydration.

Reference:

Question(s) 444: Anatomy
Discussion:
The photomicrographs show pigmented cells in the leptomeninges that have elongated cell processes. Of the various pigmented cells that might be encountered in the central nervous system, the long bipolar cytoplasmic processes and location in the leptomeninges identify these cells as leptomeningeal melanocytes. These cells are normal constituents of the central nervous system and are found predominantly in the leptomeninges of the upper cervical cord, medulla, and mesial basal aspects of the brain stem and brain as far rostrally as the gyri recti.
Question(s) 447: Pathology

Discussion:
The dysembryoplastic neuroepithelial tumor (DNT) is composed of neurons and oligodendrogial-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) 448: Neuroradiology

Discussion:
Of the available options, only the suppurative ventriculitis would cause abnormal ependymal enhancement and the CSF to appear hyperdense. Hemorrhage into ventricle could have a similar appearance. The white matter shows marked edema.

Reference:

Question(s) 449: Neuroradiology

Discussion:
Multiple sclerosis plaques are shown. The thoracic sensory level relates to plaques in the thoracic cord.

Reference:

Question(s) 450: Anatomy

Discussion:
The photomicrograph shows a meningothelial (arachnoid) cell cluster in the fibrovascular stroma of the choroid plexus of the lateral ventricular trigone (glomus choroideum). Small nests of meningothelial cells like these are commonly found in the stroma throughout the choroid plexus and are a result of the developmental mode of formation of the choroid in which the vascular leptomeninges (including arachnoid cells) invaginate into the ventricular cavity. The clinical significance of these small arachnoid cell rests is that they occasionally give rise to meningiomas (intraventricular meningioma).

Reference:

Question(s) 451: Neuroradiology

Discussion:
Pleomorphic xanthoastrocytoma typically produce a large cyst with superficially positioned mural nodule. Most lesions arise within the temporal or parietal lobes. This lesion is surrounded with minimal hemosiderin rim, but no edema and this excludes abscess and hematomas. The signal characteristics of cavernous hemangioma are heterogeneous (popcorn). The mass effect is too little for a hematoma of this size. Porencephalic cysts do not exert mass effect.

Reference:
Question(s) 452: Neuroradiology

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) 453: Neuroradiology

Discussion:
Axial CTs show a large mass lesion filling the right orbit, compressing and deforming the posterior margin of the globe. The inferior cut through the maxillary sinus shows that the lateral wall of the maxilla is destroyed and is associated with the soft tissue mass. Metastatic disease commonly involves bone and adjacent soft tissue, as in this case. Facial trauma does not produce a disappearance of bone, but fractures it. Fibrous dysplasia expands bone but does not destroy it. Hemangioma may involve bone and soft tissue, but is not a destructive process. In the orbit, capillary hemangiomas are commonly found in the cutaneous tissues and periorbita, but do not extend retroglober and do not destroy bone. Cavernous hemangiomas are discrete masses that may be found intra- or extracolonic, but do not usually cross these margins.

Reference:

Question(s) 454: Neuroradiology

Discussion:
There is an intradural, extramedullary mass in the thoracic spine causing significant cord compression. This represents either neurofibroma or meningioma, and requires surgical removal. Neither tumor is radiosensitive. Imaging with CT or myelography is not likely to add any further useful information.

Reference:

Question(s) 455: Neuroradiology

Discussion:
Focal, poorly marginated cortical lesion is seen in the parasagittal left posterior frontal lobe. The high signal on T1-weighted image is due to T1 shortening and is consistent with the presence of intracellular or extracellular methemoglobin in a subacute hemorrhage. Of the lesions mentioned, only hemorrhagic infarct and AVM would contain subacute blood. The small size and cortical location of the blood are more consistent with infarct than with vascular malformation. The blood is in a vascular territory, most likely due to embolic occlusion of a paracentral branch of the anterior cerebral artery.

Reference:
Question(s) 456: 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) 457: 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.

References:

Question(s) 458: Neuroradiology

Discussion:
This non-contrast CT reveals a hyperdense tubular region in the proximal segment of the left middle cerebral artery (MCA), consistent with acute thrombosis (arrow). Also noted are subtle left cerebral sulcal effacement, hypodensity of the left basal ganglia, frontal, anterior parietal, and superior temporal lobes. These findings are consistent with an acute MCA infarction. This so-called dense MCA sign or hyperdense MCA sign has been correlated angiographically with embolic or atherothrombotic MCA occlusion. The hyperdensity is most likely due to either calcific or hemorrhagic components of the acute plaque. The hyperdense MCA sign is nonspecific when present in isolation. False positive hyperdense MCAs have been noted in asymptomatic patients with high hematocrit or calcific atherosclerotic disease; these are usually bilateral.

Reference:

Question(s) 459: Neuroradiology

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 usual location for strokes. Epidermoid and arachnoid cysts are usually unilateral and not associated with surrounding parenchymal hyperintensities.

References:

Question(s) 460: Pathology

Discussion:
The cell shown contains a neurofibrillary tangle - an intraneuronal intracytoplasmic inclusion composed of paired helical filaments. The section has been stained with a silver stain. Neurofibrillary tangles can be seen in a variety of conditions including Alzheimer disease, Pick disease, progressive supranuclear palsy, Down syndrome, Parkinson-dementia complex of Guam, and post-encephalitic Parkinsonism.

Reference:
**Question(s) 461: Neuroradiology**

**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:**

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**Question(s) 463: Neuroradiology**

**Discussion:**
Sagittal T1-weighted and coronal proton density weighted images show a high signal intensity cortical-subcortical lesion in the temporal lobe. This has the appearance of methemoglobin in that the lesion is bright on the T1 and the proton density image. Fat on the proton density image is becoming dark as is seen in the scalp fat. In addition, there is a rim of low signal intensity surrounding the hematoma on the proton density image. This is due to the presence of hemosiderin in macrophages surrounding the hematoma. For this phenomenon to have occurred, the hematoma has to be approximately two weeks or more in age.

**Reference:**

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**Question(s) 464: Physiology**

**Discussion:**
The illustration shows typical centrotemporal spikes, often associated with benign rolandic epilepsy of childhood, which is manifested by clonic movements of the face and hand that often progress to a more generalized seizure.

**Reference:**

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**Question(s) 465: Neuroradiology**

**Discussion:**
The angiogram shows the distal common carotid artery and its branches. The proximal internal carotid artery has a rounded stump with no distal flow, consistent with occlusion to atherosclerosis. A dissection usually has a tapered stump, not a rounded stump. Vessel occlusion in moya-moya is usually intracranial.

**Reference:**
**Question(s) 466:** Neuroradiology  
**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) 467:** Neuroradiology  
**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) 468:** Neuroradiology  
**Discussion:**  
The lesion at the tip of the left temporal lobe spares the cortical ribbon and is clearly extra-axial, ruling out old head trauma or herpes simplex encephalitis. The signal characteristics are typical of CSF, ruling out a metastasis. The lesion on the right perimesencephalic cistern cannot be incisural sclerosis, characterized by gliosis, not by a CSF-like lesion like the one shown. Both lesions are arachnoid cysts.

**Reference:**  

**Reference:**  

**Question(s) 469:** Neuroradiology  
**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) 470:** Pathology  
**Discussion:**  
The photomicrographs show classic perivascular pseudorosettes characteristic of ependymoma.

**Reference:**  

**Question(s) 471:** 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) 472: Physiology**

**Discussion:**
Jitter on single fiber EMG can be seen with diseases of neuromuscular transmission such as myasthenia gravis as well as a denervating neuropathy such as amyotrophic lateral sclerosis.

**Reference:**

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**Question(s) 473 - 477: Neuroradiology**

**Discussion:**
The petrosal segment of the carotid artery (C) has a vertical and horizontal segment, is just distal to the cervical portion of the carotid, and is followed by the cavernous portion of the carotid artery. The ophthalmic artery arises from the anterior surface of the carotid artery and is a good marker for the carotid artery piercing the dura. The posterior communicating artery (A) connects the distal internal carotid artery (B) and proximal posterior cerebral artery. Anterior cerebral artery (D) and anterior communicating artery (E) are also shown.

**Reference:**

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**Question(s) 478 - 480: Neuroradiology**

**Discussion:**
The images are MR T1-weighted images (long TR/short TE). This is apparent because of the marked hypointensity of the CSF in relation to brain parenchyma. T2-weighted images, with long TR and long TE, would show hyperintense CSF. Fat saturation MR images would suppress the thin bright rim overlying the skull (subcutaneous fat). The structure pointed to by (A) shows the sylvian (lateral) fissure/circular sulcus region. This separates the frontal and temporal lobes. The structure pointed to by (B) shows the trigone/atrium of lateral ventricle. The structure marked by (C) shows the genu of corpus callosum. The structure marked by (D) shows the frontal paranasal sinus. The bright superficial rim around the skull is subcutaneous fat (E), because it is bright on T1-weighted image, due to T1 shortening effect. None of the other structures typically cause T1 shortening. Hyperintensity on T1-weighted image results from shortening of T1 relaxation time. This is seen in a variety of tissues on noncontrast images, most commonly, blood, fat, calcium, and protein-rich tissue.

**Reference:**