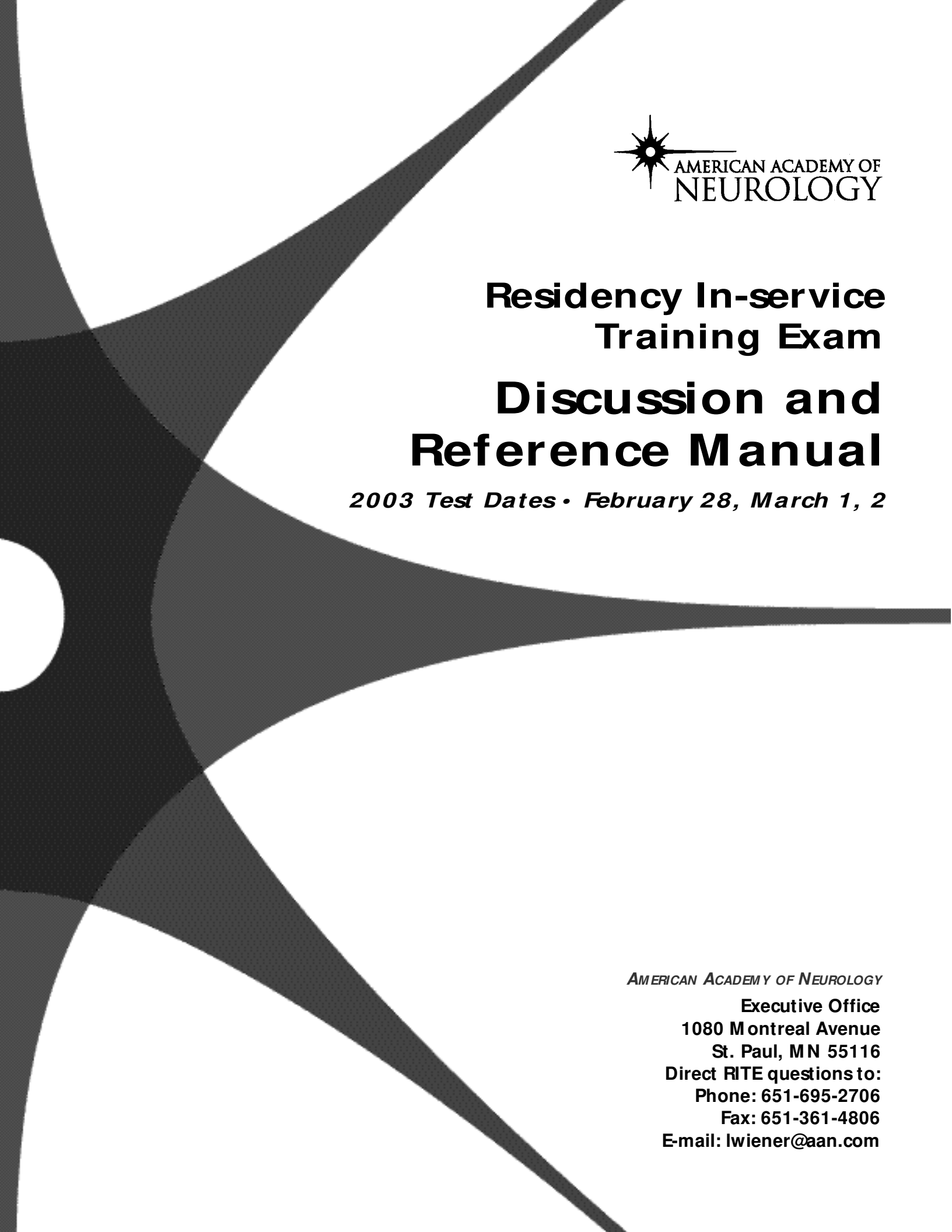

Residency In-service Training Exam

Discussion and Reference Manual

2003 Test Dates • February 28, March 1, 2

AMERICAN ACADEMY OF NEUROLOGY

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EXAM OBJECTIVES

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

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

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

FREQUENTLY CITED REFERENCES

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Menkes JH, Sarnat HB, editors. Child neurology. 6th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

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Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Scriver CR, Beaudet AL, Sly WS, et al, editors. The molecular and metabolic bases of inherited disease. 8th ed. New York: McGraw-Hill, 2001.

Swaiman KF, Ashwal S, editors. Pediatric neurology. 3rd ed. St. Louis: Mosby, 1999.

Question(s) 1: Behavioral

Discussion:
Mild memory impairment or subtle changes in other cognitive functions that do not interfere with daily activities and for which no other underlying cause can be found constitute Mild Cognitve Impairment (MCI).

Reference:
Bennett DA, Wilson RS, Schneider JA, et al. Natural history of mild cognitive impairment in older persons. Neurology 2002;59:198-205.

Question(s) 2: Clinical Pediatrics

Discussion:
Ataxia telangiectasia commonly presents with recurrent infections before the onset of conjunctival and malar telangiectasia, which develop before nystagmus and ataxia. Patients with ataxia telangiectasia, unlike those with other immunodeficiencies, develop B-cell neoplasia such as lymphoma and Hodgkin's disease. Bassen-Kornzweig syndrome is characterized by malabsorption, peripheral neuropathy and pigmentory retinopathy that responds to therapy with vitamin E. Friedreich's ataxia is a progressive, autosomal recessive disorder presenting between age 10 and 12. Spinocerebellar ataxia type III is a trinucleotide repeat disorder that presents in adulthood. Refsum disease (phytanic acid oxidase deficiency) presents with ataxia, retinitis pigmentosa and icthyosis.

Reference:
1. Oski FA, DeAngelis CD, Feigin RD, et al. Principles and practice of pediatrics. Philadelphia: JB Lipincott, 1990.
2. Menkes JH, Sarnat HB, editors. Child neurology. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 3: Clinical Adult

Discussion:
When the eye is rotated outward about 23 degrees, the superior rectus is a pure elevator.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 4: Physiology

Discussion:
In acquired demyelinating polyneuropathy, multifocal demyelination results in abnormal temporal dispersion of the compound muscle action potential.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 5: Physiology

Discussion:
The 3 Hz spike-and-wave pattern is facilitated by hyperventilation, alkalosis, hypoglycemia and drowsiness, but is diminished during REM sleep.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998

Question(s) 6: Clinical Adult

Discussion:
Dementia is now recognized as an important feature of Parkinson's disease, eventually occurring in about a third of patients (though reported incidence varies widely between studies). Dementia is also a major symptom of progressive supranuclear palsy (PSP), but the manifestations may be overlooked or attributed to the dysarthria and eye movement abnormalities. Mental deterioration occurs in about 85% of patients with juvenile Huntington's disease. Higher cortical functions are often altered in Wilson's disease. Machado-Joseph disease is a hereditary disorder with variable manifestations that can include dystonia, athetosis, rigidity, spasticity, lower motor neuron signs, and ataxia, but intelligence remains intact.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 7: Clinical Adult

Discussion:
The pronator quadratus is responsible for pronation of the forearm when the elbow is flexed. It is supplied by the anterior interosseous nerve, a branch of the median nerve.

Reference:
Brown WF, Bolton CF, editors. Clinical electromyography. 2nd ed. Boston: Butterworth-Heinemann, 1993.

Question(s) 8: Physiology

Discussion:
Bilateral absence of cortical responses (N20) on median somatosensory evoked potentials is associated with a poor outcome in comatose patients.

Reference:
Chiappa KH. Evoked potentials in clinical medicine. 3rd ed. New York: Lippincott-Raven Press, 1997.

Question(s) 9: Pharmacology/Chemistry

Discussion:
The prominent manifestation of congenital deficit of dopamine-beta-hydroxylase is orthostatic hypotension. Elevated levels of plasma dopamine and undetectable levels of plasma norepinephrine are diagnostic features of this disorder. DL threo-dihydroxyphenylserine (DL-threo-DOPS) is an unantural amino acid that is converted to norepinephrine through a single decarboxylation step by the dopa decarboxylase, bypassing the dopamine-beta-hydroxylase step, and is very effective for management of orthostatic hypotension in this disorder.

Reference:
Biaggioni I, Roberstson D. Endogenous restoration of noradrenaline by precursor therapy in dopamine-beta-hydroxylase deficiency. A novel orthostatic syndrome. Lancet 1987;1(8526):183-188.

Question(s) 10: Clinical Adult

Discussion:
Leukemics have a very high incidence of leptomeningeal involvement (up to 70% of patients) although prophylactic CNS treatment has markedly reduced the incidence of symptomatic disease.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 11: Physiology

Discussion:
Injury to the deep peroneal nerve results in weakness of foot and toe dorsiflexion with decreased recruitment in the anterior tibialis muscle and sensory loss in the web space between the first two toes.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 12: Anatomy

Discussion:
Light first encounters the innermost layer of the retina, the ganglion cell layer.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 13: Clinical Adult

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

Reference:
Bousser MG, Tournier-Lasserve EJ. Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy: from stroke to vessel wall phsiology. J Neurol Neurosurg Psychiatry 2001;70:285-287.

Question(s) 14: Physiology

Discussion:
Sensory responses would be expected to be affected in lesions distal to the dorsal root ganglion (ie., plexopathies); while they would be spared in lesions proximal to the dorsal root ganglion (i.e., radiculopathies). Thus, they are quite useful in distinguishing between root and plexus localizations.

Reference:
Brown WF, Bolton CF, editors. Clinical electromyography. 2nd ed. Boston: Butterworth-Heinemann, 1993.

Question(s) 15: Anatomy

Discussion:
The armpit's dermatome is usually T2.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 16: Physiology

Discussion:
Mu activity is attenuated by movement of an extremity.

Reference:
Fisch B. Spehlmann's EEG primer. 2nd ed. Amsterdam: Elsevier Science Publications, 1991.

Question(s) 17: Behavioral

Discussion:
Motor and verbal tics are required for a clinical diagnosis of Tourette's syndrome. Motor tics alone are not sufficient for the diagnosis of Tourette's syndrome and coprolalia is not required (as it occurs in only approximately 30% of cases).

Reference:
Moore DP. Textbook of clinical neuropsychiatry. New York: Oxford University Press, 2001.

Question(s) 18: Clinical Adult

Discussion:
A drowsy patient with a right hemiplegia and eyes deviated to the left that conjugately cross the midline with the doll's eye maneuver is more likely due to a frontal lesion than a lateral medullary, occipital, pontine, or thalamic lesion.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 19: Behavioral

Discussion:
Mirtazapine is sedating even at lower doses. It would not be safe to use it in a cross-country truck driver. Fluoxetine, bupropion, desipramine, and sertraline are far less sedating than mirtazapine.

Reference:
Arana GW, Rosenbaum JF. Handbook of psychiatric drug therapy. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 20: Physiology

Discussion:
In a lesion of the peroneal nerve above the knee, the short head of the biceps femoris muscle will show evidence of denervation in addition to the tibialis anterior and peroneus longus muscles.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 21: Physiology

Discussion:
The femoral nerve innervates muscles involved in hip flexion and knee extension. Its sensory territory includes the territory of the saphenous nerve below the knee.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 22: Clinical Adult

Discussion:
Tardive dyskinesia and parkinsonian symptoms are hazards of metoclopramide administration, especially in the elderly.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 23: Clinical Adult

Discussion:
The most useful laboratory study for diagnosing amyotrophic lateral sclerosis (ALS) is the EMG, which shows widespread changes of denervation. Spinal fluid is usually normal, but mild protein elevation (less than 100 mg/dl) may occur. While serum creatine kinase levels are typically normal, they may be increased to two to three times normal in almost half the patients with ALS. Sedimentation rate is usually normal.

Reference:
Brooke MH. A clinician's view of neuromuscular diseases. 2nd ed. Baltimore: Williams & Wilkins, 1986.

Question(s) 24: Physiology

Discussion:
In an acute lumbar radiculopathy, the only abnormal findings on a needle EMG examination performed within a few days of onset of symptoms may be reduced recruitment of motor unit potentials.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 25: Anatomy

Discussion:
Microglial cells are of mesodermal origin and have a phagocytic function when central nervous system tissue is injured. Schwann cells, ependymal cells, tanycytes and oligodendrocytes are of ectodermal origin.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 26: Physiology

Discussion:
The peripheral neuropathy associated with monoclonal gammopathy is demyelinating in character. Nerve conduction velocities in the teens or twenties would be characteristic of this entity.

Reference:
Dyck PJ, Thomas PK, Griffin JW, et al, editors. Peripheral neuropathy. 3rd ed. Philadelphia: WB Saunders, 1993.

Question(s) 27: Behavioral

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

Reference:
American Psychiatric Association. Diagnostic and statistical manual of mental disorders. 4th rev. ed. Washington: American Psychiatric Association, 2000.

Question(s) 28: Clinical Adult

Discussion:
Hypothyroid myopathy is commonly associated with painful cramps, normal reflexes, and impressive elevations in serum creatine kinase (CK) levels. Percussion of muscle commonly causes a slow, prolonged, electrically silent local mounding called myoedema.

Reference:
Aminoff MJ. Neurology and general medicine. 3rd ed. New York: Churchill Livingston, 2001.

Question(s) 29: Clinical Pediatrics

Discussion:
Myotonic dystrophy is an autosomal dominant disorder that is associated with CTG repeats at the 19q13.3 locus. Maternal inheritance can lead to a severe form of neonatal myotonic dystrophy. Testicular atrophy is usually not associated with sterility.

Reference:
Swaiman KF, Ashwal S, editors. Pediatric neurology. 3rd ed. St. Louis: Mosby, 1999.

Question(s) 30: Pharmacology/Chemistry

Discussion:
The main site of disposal of manganese in the body is biliary excretion. Patients with biliary atresia, chronic liver disease, or exposure to high dose of manganese during prolonged parenteral nutrition, are prone to develop manganese intoxication. Clinically, it is characterized by parkinsonism and dystonia, which do not respond to levodopa. T1-weighted MRI shows hyperintensity in the globus pallidus, striatum, and midbrain. The primary site of damage is the globus pallidus.

Reference:
Pal PK, Samili A, Calne DB. Manganese neurotoxicology: a review of clinical features, imaging, and pathology. Neurotoxicology 1999;20:227-238.

Question(s) 31: Pharmacology/Chemistry

Discussion:
Rotenone is commonly used as an herbicide and is a potent inhibitor of mitochondrial complex I. Chronic administration of rotenone in rats reproduces several findings of Parkinson's disease, including selective loss of susbtantia nigra compacta neurons and accumulation of inclusions resembling Lewy bodies.

Reference:
Betarbet R, Sherer TB, MacKenzie G, et al. Chronic systemic pesticide exposure reproduces features of Parkinson's disease. Nat Neurosci 2000;3:1301-1306.

Question(s) 32: Physiology

Discussion:
Fusimotor fibers transmit impulses to muscles spindles.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 33: Physiology

Discussion:
The amplitude of the facial nerve CMAP is correlated with the number of functional axons in the facial nerve. The lower the CMAP amplitude the more axonal degeneration and the poorer the prognosis.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 34: Physiology

Discussion:
Postcentral cortical cells that have small receptive fields are likely to have their receptive fields on the distal parts of contralateral extremities.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 35: Physiology

Discussion:
Temporal lobe seizures are associated with confusion, contralateral posturing of the hand, automatisms, and difficulty speaking. This constellation of findings suggests an origin in the left temporal lobe.

Reference:
1. Walker M, Shorvon S. Partial epilepsy syndromes in adults. In: Porter RJ, Chadwick D, editors. The epilepsies 2. Boston: Butterworth-Heinemann, 1997.
2. Williamson PD, Engel J. Complex partial seizures. In: Engel J, Pedley TA, editors. Epilepsy: a comprehensive textbook. New York: Lippincott-Raven, 1998.

Question(s) 36: Pathology

Discussion:
Vitamin B12 deficiency produces combined system degeneration, with damage to the posterior columns and lateral columns. Associated megaloblastic anemia may be present. Amyotrophic lateral sclerosis classically produces neuroaxonal loss of the upper and lower motor neurons with sparing of the sensory columns; posterior column involvement is rarely seen in familial ALS. Anterior spinal artery thrombosis spares the posterior columns supplied by the paired posterior spinal arteries. Neurosyphilis produces tabes dorsalis as its primary spinal cord manifestation, with demyelination and axonal loss in the posterior columns but with sparing of the anterior corticospinal tracts. Vitamin E deficiency produces dystrophic axons in the posterior columns.

Reference:
Fuller GN, Goodman JC. Practical review of neuropathology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 37: Behavioral

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

Reference:
Rasmussen K, Abrams R. Treatment of Parkinson's disease with electroconvulsive therapy. (Review) Psychiatric Clinics of North America. Philadelphia: WB Saunders, Co., 1991.

Question(s) 38: Anatomy

Discussion:
Ballism is associated with discrete lesions in the subthalamic nucleus. The dyskinesia occurs contralateral to the lesion and is associated with hypotonia.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 39: Pathology

Discussion:
In males suspected of having Duchenne's or Becker's muscular dystrophy, dystrophin analysis may reveal the presence of a mutation in affected individuals. Carrier analysis can also be performed. The precise location and size of the dystrophin mutation determines whether the more severe Duchenne's phenotype or the milder Becker's phenotype will be seen.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 40: Physiology

Discussion:
Averaging in evoked potential testing increases the signal to noise ratio.

Reference:
Harper CM. Somatosensory evoked potentials. In: Daube JR, editor. Clinical neurophysiology. Philadelphia: FA Davis Company, 1996.

Question(s) 41: Physiology

Discussion:
Paraspinal muscle denervation usually indicates a lesion at the level of the ventral nerve root.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 42: Clinical Adult

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

Reference:
Stewart JD. Focal peripheral neuropathies. New York: Elsevier, 1987.

Question(s) 43: Anatomy

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

Reference:
Patten J. Neurological differential diagnosis. 2nd ed. New York: Springer, 1996.

Question(s) 44: Physiology

Discussion:
Damage to autonomic fibers in the leg can result in tachycardia, vasodilation, hypotension, loss of sweating, and loss of nerve terminals that produce acetylcholine.

Reference:
Low PA. Clinical autonomic disorders: evaluation and management. 2nd ed. New York: Lippincott-Raven, 1997.

Question(s) 45: Physiology

Discussion:
Diminished slow component peak velocity with cool and warm water caloric testing on one side during ENG indicate an ipsilateral peripheral vestibular abnormality.

Reference:
Brey RH. Vertigo and balance. In: Daube JR, editor. Clinical neurophysiology. Philadelphia: FA Davis Company, 1996.

Question(s) 46: Clinical Adult

Discussion:
The blood supply to the mid-thoracic (T4-T6) cord is relatively tenuous making this region of the spinal cord most vulnerable to ischemia.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 47: Clinical Adult

Discussion:
Cerebellum is a more frequent site of CNS metastases than brain stem, leptomeninges, thalamus or spinal cord.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 48: Pathology

Discussion:
Devic's disease is also known as neuromyelitis optica; the spinal cord and optic nerves are predominantly involved although other regions of the CNS may show lesions indistinguishable from classic multiple sclerosis.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 49: Clinical Adult

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

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 50: Clinical Adult

Discussion:
The suprascapular nerve may be damaged by trauma or by compression within the suprascapular notch (e.g., by a ganglion or by the suprascapular ligament). The affected muscles are the supraspinatus and infraspinatus, responsible for initial abduction and external rotation of the shoulder, respectively.

Reference:
Stewart JD. Focal peripheral neuropathies. New York: Elsevier, 1987.

Question(s) 51: Pharmacology/Chemistry

Discussion:
Suboptimal doses of carbidopa is a frequent cause of nausea/vomiting at the initiation of levodopa therapy. It takes 100-150 mg of carbidopa per day to saturate the peripheral aromatic aminoacid decarboxylase enzyme to prevent peripheral side effects of levodopa.

Reference:
Roger E, Kurlan JB. Treatment of movement disorders. Philadelphia: Lippincott Co., 1995.

Question(s) 52: Anatomy

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

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 53: Behavioral

Discussion:
The hallmark of the acute confusional state is impaired attention.

Reference:
Taylor MA. The fundamentals of clinical neuropsychiatry. New York: Oxford University Press, 1999.

Question(s) 54: Physiology

Discussion:
Wave V of the brainstem auditory evoked potential is thought to be generated at the level of the inferior colliculus and an absence of wave V would suggest a lesion of the lower midbrain at the level of the inferior colliculus.

Reference:
Chiappa KH. Evoked potentials in clinical medicine. 3rd ed. New York: Lippincott-Raven Press, 1997.

Question(s) 55: Physiology

Discussion:
In a patient with myxedema, a single stimulus to a nerve results in slow contraction and relaxation of the muscle.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 56: Anatomy

Discussion:
The area postrema, a circumventricular organ, lacks a blood-brain barrier. It acts as a chemoemetic center.

Reference:
Fuller GN, Goodman JC. Practical review of neuropathology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 57: Anatomy

Discussion:
Although they are part of the sympathetic nervous system, the postganglionic fibers to the sweat glands (sudomotor fibers) are cholinergic.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 58: Clinical Adult

Discussion:
Patients with severe hypermagnesemia may also have low calcium and cardiac conduction abnormalities. Treatment involves administration of intravenous calcium gluconate. Patients should be monitored closely for impending respiratory failure, and if it occurs, they should be immediately intubated and mechanically ventilated (preferably with simultaneous calcium administration). Saline and calcium are used together if the patient is not in renal failure. Hypermagnesemia associated with renal failure requires hemodialysis for treatment.

Reference:
Riggs JE, Neurologic manifestations of fluid and electrolyte disturbances. Neurol Clin 1989;7:509.

Question(s) 59: Physiology

Discussion:
A myoclonic seizure is the most common type of seizure induced by photic stimulation in patients with light sensitivity.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 60: Clinical Adult

Discussion:
While not always the earliest finding, the diagnosis of progressive supranuclear palsy is confirmed by the appearance of vertical gaze paresis.

Reference:
Watts RL, Koller WC. Movement disorders: neurologic principles and practice. New York: McGraw-Hill, Co., 1997.

Question(s) 61: Anatomy

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

Reference:
Patten J. Neurological differential diagnosis. 2nd ed. New York: Springer, 1996.

Question(s) 62: Physiology

Discussion:
Phenytoin reduces post-tetanic potentiation.

Reference:
Levy RH, Mattson RH, Meldrum BS, editors. Antiepileptic drugs. 4th ed. New York: Raven Press, 1995.

Question(s) 63: Clinical Adult

Discussion:
Cluster headache is characterized by severe unilateral headache associated with autonomic symptoms. Headache attacks typically last 15 minutes to two to three hours. A Horner's syndrome may occur on the side of the headache. The entity is much more common in men than women and usually occurs in young individuals (mean onset 28 years).

Reference:
Raskin NH. Headache. 2nd ed. New York: Churchill-Livingstone, 1998.

Question(s) 64: Physiology

Discussion:
The cerebellum exerts its effect on synergy of movement by way of the dentatorubrothalamic pathway.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 65: Anatomy

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

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 66: Anatomy

Discussion:
The terminal branches of the posterior cord of the brachial plexus are the radial and axillary nerves.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 67: Pharmacology/Chemistry

Discussion:
Anandamide is an endogenous ligand for the cannabinoid receptor, which has been implicated in several functions including pain control. In addition, anandamide is an endogenous ligand of the vanilloid 1 receptor (VR1), which is activated by heat and low pH and mediates the effects of capsaicin on primary nociceptive afferents.

Reference:
Di Marzo V, Bisogno T, De Petrocellis L. Anandamide: some like it hot. Trends Pharmacol Sci 2001;22:346-349.

Question(s) 68: Clinical Adult

Discussion:
Symptoms of perilymph fistula are variable, but may include recurrent vestibulopathy. Characteristic precipitating factors include cough, sneeze, straining, and exercise.

Reference:
Baloh RW, Honrubia V. Clinical neurophysiology of the vestibular system. Philadelphia: FA Davis Co., 1990.

Question(s) 69: Behavioral

Discussion:
The rapid course of the dementia with myoclonus, visual and cerebellar impairment (wide-based gait) is suggestive of Creutzfeldt-Jakob disease. EEG often reveals pseudoperiodic high voltage sharp and slow waves superimposed on an increasingly slow and low voltage background. These sharp waves are usually synchronous with myoclonus, but may persist in the absence of myoclonus. CSF immunoassay of peptide fragments of brain proteins known as 14-3-3, (although not pathognomonic for a specific disease), in this clinical setting, is consistent with Creutzfeldt-Jakob disease.

Reference:
Victor M, Ropper A. Principles of neurology, 7th ed. New York: McGraw-Hill, 2001.

Question(s) 70: Clinical Adult

Discussion:
Saddle anesthesia, sphincter loss and loss of ankle reflexes after a fall signify midline disk herniation with compression of the caudal equina.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 71: Behavioral

Discussion:
Anticholinergic side effects are of considerable significance in the pharmacologic treatment of depression. Among those that are most likely to cause these side effects are the tricyclic antidepressants, especially the tertiary amines such as amitriptyline.

Reference:
Arana GW, Rosenbaum JF. Handbook of psychiatric drug therapy. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 72: Behavioral

Discussion:
Alexia without agraphia (also known as pure word blindness or acquired pure alexia) is most often produced by left posterior cerebral artery occlusion. A frequent associated finding is a right homonymous hemianopsia.

Reference:
Damasio A, Tranel D, Rizzo M. Disorders of complex visual processing. In: Mesulam MM, editor. Principles of behavioral and cognitive neurology. 2nd ed. New York: Oxford University Press, 2000.

Question(s) 73: Pharmacology/Chemistry

Discussion:
Reduced concentrations of coenzyme Q-10 (CoQ-10) have been found in patients with myopathy complicating the use of statins. Primary CoQ-10 deficiency produces either a myopathy or progressive cerebellar degeneration, reflecting the key role of CoQ-10 as an electron shuttle between complexes I or II and complex III in the respiratory chain.

Reference:
1. Musumeci O, Naini A, Slonim AE. Familiar cerebellar ataxia with muscle coenzyme Q-10 deficiency. Neurology 2001;56: 849-855.
2. Farmer JA. Learning from the cerivastatin experience. Lancet 2001;358:1383-1384.

Question(s) 74: Clinical Pediatrics

Discussion:
Between 60% and 75% of children with epilepsy who have been seizure-free for more than two years on medications will remain seizure-free for 2 years when anti-epileptic drugs are withdrawn.

Reference:
Pellock JM, Dodson WE, Bourgeois BFD, editors. Pediatric epilepsy, diagnosis and treatment. 2nd ed. New York: Demos, 2001.

Question(s) 75: Anatomy

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

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 76: Clinical Adult

Discussion:
A lesion of the posterior interosseous nerve typically results in a "finger drop" rather than wrist drop, because the branches to the extensor carpi radialis brevis and longus muscles usually arise from the main trunk of the radial nerve proximal to the origin of the posterior interosseous nerve. The triceps and brachioradialis muscles are spared for similar reasons. In contrast, the supinator muscle is innervated by the posterior interosseous nerve (as it pierces the muscle).

Reference:
Stewart JD. Focal peripheral neuropathies. New York: Elsevier, 1987.

Question(s) 77: Physiology

Discussion:
Periodic sharp waves and/or spikes are most likely to be seen with a post-anoxic state.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 78: Clinical Adult

Discussion:
Chemical meningitis is a rare complication of craniopharyngioma in which seeding of cyst material into the subarachnoid space occurs.

Reference:
Youmans JR, editor. Neurological surgery: a comprehensive reference guide to the diagnosis and management of neurosurgical problems. Philadelphia: WB Saunders Company, 1996.

Question(s) 79: Pharmacology/Chemistry

Discussion:
Recurrent stupor is a rare disorder associated with excessive accumulation of endozepine-4, an endogenous ligand of the benzodiazepine receptor. These patients show elevated endozepine levels in blood and cerebrospional fluid. The electroenecphalogram during the episodes resemble that found in patients taking benzodiazepines. Flumazenil induces transient (10-15 min) of wakefulness in these patients.

Reference:
Lugaresi E, Montagna P, Tinuper P, et al. Endozepine stupor: recuring stupor linked to endozepine-4 accumulation. Brain 1998;121:127-133.

Question(s) 80: Physiology

Discussion:
The EEG in Huntington's chorea shows a low voltage background.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 81: Clinical Adult

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

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 82: Behavioral

Discussion:
Similarities between Alzheimer's disease and Down's syndrome include the presence of typical neuropathological changes (plaques and neurofibrillary tangles) and abnormalities of chromosome 21. Down's syndrome individuals routinely develop the pathology of Alzheimer's disease by their late 30s/early 40s. By the time Down's patients are age 50 years, over 50% will experience seizures. In those with dementia the figure approaches 80%.

Reference:
Moore DP. Textbook of clinical neuropsychiatry. New York: Oxford University Press, 2001.

Question(s) 83: Behavioral

Discussion:
Transcortical motor and sensory aphasia patients retain the ability to repeat. Broca's, Wernicke's, and conduction aphasia as well as pure word deafness all have impairments of repetition.

Reference:
Benson DF. Aphasia, alexia, and agraphia. New York: Churchill Livingston, 1979.

Question(s) 84: Anatomy

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

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 85: Behavioral

Discussion:
In most instances, the duration of transient global amnesia is less than 12 hours, produces no other focal neurologic abnormalities and usually disappears without residua. The amnesia affects recall of recent events and impairs new learning, but spares digit span (immediate recall).

Reference:
Hodges JR, Warlow CP. Syndromes of transient amnesia: towards a classification. A study of 153 cases. Neurol, Neurosurg, and Psychiatry 1990;53:834-843.

Question(s) 86: Clinical Adult

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

Reference:
Aminoff MJ. Neurology and general medicine. 3rd ed. New York: Churchill Livingston, 2001.

Question(s) 87: Clinical Adult

Discussion:
Following ischemic stroke, motor recovery tends to plateau more quickly than functional recovery with only small changes seen after 8-12 weeks. Arm movement generally recovers less than leg movement. Hemiparesis recovers significantly over the course of a year following stroke with severe hemiplegia persisting in only 9% of patients. More than 50% of patients are able to sit unsupported in the first week after stroke.

Reference:
Dombovy M. Rehabilitation and the course of recovery after stroke. In: Whisnant JP, editor. Stroke: populations, cohorts, and clinical trials. Boston: Butterworth-Heinemann, 1993.

Question(s) 88: Behavioral

Discussion:
Any Alzheimer's patient who acutely develops symptoms of an acute confusional state and behavioral changes first warrants a workup to look for the underlying cause. Even mild changes in metabolic status, medications, or an infection such as a urinary tract infection may precipitate confusion and behavioral changes.

Reference:
Cummings JL, Benson DF. Dementia: a clinical approach. Boston: Butterworth-Heinemann, 1992.

Question(s) 89: Pharmacology/Chemistry

Discussion:
3,4-diaminopyrimidine (3,4-DAP) is effective in improving both neuromuscular and autonomic manifestations of the Lambert-Eaton myasthenic syndrome. This disorder is due to an immune-mediated blockade of prejunctional P/Q type voltage-gated calcium channels responsible for acetylcholine release. 3,4-DAP blocks the voltage-gated potassium channels responsible for repolarization of the action potential, and thus increases action potential duration and opening of the P/Q channels.

Reference:
McEvoy KM, Windebank AJ, Daube JR, et al. 3,4-diaminopyridine in the treatment of Lambert-Eaton myasthenic syndrome. NEJM 1989;321:1567-1571.

Question(s) 90: Anatomy

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

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 91: Anatomy

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 92: Clinical Adult

Discussion:
Patients with McArdle's disease and phosphofructokinase deficiency do not have a normal rise in serum lactate with the ischemic exercise test.

Reference:
Engel AG, Franzini-Armstrong C. Myology. New York: McGraw Hill, 1994.

Question(s) 93: Pathology

Discussion:
Lewy bodies, both cortical and brainstem type, stain with antibodies directed against alpha synuclein. The gene for synuclein, located on chromosome 4, is mutated in rare familial forms of Parkinson's disease. Various types of inclusions in sporadic forms of Parkinson's disease, diffuse Lewy body disease, and multiple system atrophy can be identified by alpha-synuclein staining; these three disorders are considered synucleinopathies. In contrast, "tauopathies" include Alzheimer's disease, Pick's disease, frontotemporal dementia with parkinsonism (FTDP-17), progressive supranuclear palsy, and corticobasal ganglionic degeneration.

Reference:
1. Dickson, DW. Tau and synuclein and their role in neuropathology. Brain Pathol 1999;9:657-661.
2. Fuller GN, Goodman JC. Practical review of neuropathology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 94: Anatomy

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 95: Clinical Adult

Discussion:
The initial ocular motor deficit in patients with progressive supranuclear palsy consists of impairment of vertical saccades, with downward saccades usually affected first. Tremor is rare. Axial rigidity is greater than extremity rigidity, postural instability with falls is common, and dementia is often mild.

Reference:
1. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.
2. Victor M, Ropper A. Adams and Victor's principles of neurology. 7th ed. New York: McGraw-Hill, Co., 2001.

Question(s) 96: Behavioral

Discussion:
Major depression is frequent in Parkinson's disease, occurring in 40 to 60% of patients during the course of their illness. It is less common in dementia of the Alzheimer type or Pick's disease. In addition, depression is not associated with Anton's syndrome or Kluver-Bucy syndrome.

Reference:
Cummings JL, Benson DF. Dementia: a clinical approach. Boston: Butterworth-Heinemann, 1992.

Question(s) 97: Behavioral

Discussion:
Nortriptyline is an antidepressant with a well-defined therapeutic window. It is less effective at both very low and very high doses.

Reference:
Arana GW, Rosenbaum JF. Handbook of psychiatric drug therapy. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 98: Physiology

Discussion:
The parasympathetic nerves exiting at the S2-S4 dorsal roots mediate the urge to urinate.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 99: Clinical Adult

Discussion:
Clinical features with Horner's syndrome include miosis and mild ptosis. The light and near responses of the pupil are normal. The anisocoria is worse in the dark and less evident in bright light. A solution of 10% cocaine will dilate a normal pupil but will not dilate the pupil in a patient with Horner's syndrome.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 100:
Pharmacology/Chemistry

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

Reference:
Wald JJ. The effects of toxins on muscle. Neurology Clin 2000;18:695-717.

Question(s) 101: Clinical Adult

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

Reference:
Lupski JR, Chance PF, Garcia CA. Inherited primary peripheral neuropathies. Molecular genetics and clinical implications of CMT 1A and HNPP. JAMA 1993;270:2326-2330.

Question(s) 102: Anatomy

Discussion:
The posterior tuberomammillary nucleus of the hypothalamus provides diffuse histaminergic innervation to the cerebral cortex.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 103: Physiology

Discussion:
Tetrodotoxin blocks sodium channels.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 104: Pathology

Discussion:
Methanol intoxication causes necrosis of the optic nerves and necrosis of the putamina bilaterally.

Reference:
1. Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.
2. Fuller GN, Goodman JC. Practical review of neuropathology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 105: Anatomy

Discussion:
The medial temporal cortex projects to the mammillary bodies via the fornix.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 106: Clinical Adult

Discussion:
Functional recovery from the effects of an ischemic stroke is most significant during the first few weeks following stroke. There is some continued improvement between six months and one year. After one year, the proportion of patients with deterioration in function increases and by three to five years after stroke, more survivors experience increased disability than improvement, possibly due to the effects of co-morbidity and increased age.

Reference:
Dombovy M. Rehabilitation and the course of recovery after stroke. In: Whisnant JP, editor. Stroke: populations, cohorts, and clinical trials. Boston: Butterworth-Heinemann, 1993.

Question(s) 107: Behavioral

Discussion:
Major depression occurs in over 40% of Huntington's disease patients and suicide occurs in close to 10% of patients. Suicide is far less common in Alzheimer's disease, olivo-pontocerebellar degeneration, and amyotrophic lateral sclerosis and is rarely associated with right middle cerebral artery infarction.

Reference:
Moore DP. Textbook of clinical neuropsychiatry. New York: Oxford University Press, 2001.

Question(s) 108:
Pharmacology/Chemistry

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

Reference:
1. Victor M, Sieb JP. Myopathies due to drugs, toxins, and nutritional deficiencies. In: Engel AW, Franzini-Armstrong C, editors. Myology. 2nd ed. New York: McGraw-Hill, 1994.
2. Karpati G, Hilton-Jones D, Griggs RC. Disorders of voluntary muscle. 7th ed. New York: Churchill Livingstone, 2001.

Question(s) 109: Anatomy

Discussion:
The group of long association fibers interconnecting the superior and middle frontal gyri with the posterior superior temporal gyrus is the arcuate fasciculus. In the dominant hemisphere, interruption of this fasciculus results in conduction aphasia.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 110: Clinical Adult

Discussion:
Symptoms of Parkinson's disease usually begin asymmetrically. Myoclonus is not a feature. A variety of autonomic symptoms may occur, but they are rarely disabling. Vomiting is uncommon. Oculomotor abnormalities may also occur, but usually not of the magnitude seen in progressive suranuclear palsy. Hallucinations are uncommon except in association with medications.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 111: Pathology

Discussion:
The principal entities in the differential diagnosis of a solitary tumor of the lumbar cistern are schwannoma, meningioma, myxopapillary ependymoma, and paraganglioma of the filum terminale. Solitary metastasis may also occur. Schwannomas arise from the nerve roots of the cauda equina or from small peripheral nerve fascicles in the filum terminale. Two unique neoplasms arise from the filum terminale: myxopapillary ependymoma and paraganglioma of the filum terminale. Both tumors form well-circumscribed fusiform masses. Myxopapillary ependymomas are immunopositive for glial fibrillary acidic protein and S-100 protein while paragangliomas, as in this case, are strongly positive for the neuronal markers synaptophysin and chromogranin. Of the other two choices listed, central neurocytomas arise in the lateral and third ventricles and dysembryoplastic neuroepithelial tumors are found in the cerebral cortex.

Reference:
Fuller GN, Goodman JC. Practical review of neuropathology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 112: Physiology

Discussion:
REM sleep is characterized by fluctuations in blood pressure, muscle twitches, dreaming, a low voltage EEG pattern and a decrease in muscle tone. Susceptability to apnea is increased during REM.

Reference:
Kryger MH, Roth T, Dement WC. Principles and practice of sleep medicine. 3rd ed. Philadelphia: WB Saunders Company, 2000.

Question(s) 113: Clinical Adult

Discussion:
Polymyositis often presents as an acquired progressive proximal weakness with elevated creatine kinase levels and EMG demonstrating fibrillations and small polyphasic potentials.

Reference:
Engel AG, Franzini-Armstrong C. Myology. New York: McGraw Hill, 1994.

Question(s) 114:
Pharmacology/Chemistry

Discussion:
The hypocretin/orexin neurons of the posterior lateral hypothalamus play a critical role in preventing abrupt transitions between wakefulness and sleep. These neurons send excitatory projections to the cholinergic and monoaminergic brainstem neurons involved in arousal, and projections that inhibit the ventrolateral preoptic neurons involved in sleep onset. Impaired activity of the hypocretin/orexin neurons is responsible for narcolepsy.

Reference:
Sutcliffe JG, de Lecea L. The hypocretins: setting the arousal threshold. Nat Rev Neurosci 2002;3:339-349.

Question(s) 115: Anatomy

Discussion:
The anterior choroidal artery supplies the anteromedial part of the head of the caudate nucleus. The internal carotid artery sends branches directly to the genu of the internal capsule. The anterior and medial parts of the thalamus are supplied by posteromedial (thalamoperforating) arteries. The hippocampal formation receives its blood supply from the anterior choroidal artery.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 116:
Pharmacology/Chemistry

Discussion:
HIV-1 infection is initiated by the interaction of virion envelope glycoproteins, such as gp120, with a least two cellular receptors, the CD4 molecule and a chemokine receptor. Macrophage-tropic strains of HIV-1 replicate in macrophages and CD4+ T cells and use the CC chemokine receptor CCR5, whereas T cell-tropic HIV-1 strains replicated primarily in CD4+ T cells and use the CXC chemokine receptor CXCR4. HIV-1 enters the brain in association with infected macrophages soon after infection. HIV does not replicate in neurons and infects astrocytes and oligodendrocytes nonproductively, but replicate actively in the microglia, as well as infected macrophages that migrated to the brain.

Reference:
Power C, Gill MJ, Johnson RT. The neuropathogenesis of HIV infection: host-virus interaction and the impact of therapy. Can J Neurol Sci 2002;29:19-32.

Question(s) 117: Anatomy

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

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 118: Clinical Adult

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

Reference:
Harper CM, Thomas JE, Cascino TL, et al. Distinction between neoplastic and radiation induced brachial plexopathy, with emphasis on the role of EMG. Neurology 1989;39:502-506.

Question(s) 119: Clinical Adult

Discussion:
In the presence of a positive HIV test the most probable brain mass lesion would be either toxoplasmosis or B-cell non-Hodgkin's lymphoma. Kaposi's sarcoma rarely metastasizes to brain.

Reference:
MacArthur J. NeuroAIDS: diagnosis and management. Hosp Pract 1997;32:73-74.

Question(s) 120: Clinical Adult

Discussion:
Patients with Duchenne muscular dystrophy may develop tight heel cord muscles due to paresis of dorsiflexors of the foot. These children should be referred to physical therapy for a daily muscle-stretching program and should attempt to maintain their activity level.

Reference:
Karpati G, Hilton-Jones D, and Griggs RC. Disorders of voluntary muscle. 7th ed. New York: Churchill Livingston, 2001.

Question(s) 121: Anatomy

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

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 122: Behavioral

Discussion:
Among the defining characteristics of post traumatic stress disorder are duration more than a month, the occurrence of flashbacks, reminiscences of the episode of severe stress that the patient suffered, and persistent symptoms of increased arousal, including an exaggerated startle response.

Reference:
American Psychiatric Association. Diagnostic and statistical manual of mental disorders (DSM-IV-R). 4th ed. Washington: American Psychiatric Association, 2000.

Question(s) 123: Anatomy

Discussion:
The sural nerve is a pure sensory nerve; therefore, all of its axons arise from cell bodies located in the dorsal root ganglion. The dorsal root ganglion that contributes the majority of axons to the sural nerve is S1.

Reference:
Midroni G, Bilbao JM. Biopsy diagnosis of peripheral neuropathy. Boston: Butterworth-Heinemann, 1995.

Question(s) 124: Anatomy

Discussion:
Dorsal midbrain tegmental lesions injure the red nucleus, the third nerve nucleus, and the brachium conjunctivum, resulting in a Claude syndrome.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 125: Physiology

Discussion:
The region of the brain with the lowest threshold to electrical stimulation is the hippocampus.

Reference:
Wyllie E. Treatment of epilepsy: principles and practice. 3rd ed. Philadelphia: Lea & Febiger, 2001.

Question(s) 126: Anatomy

Discussion:
The superior cervical ganglion provides the noradrenergic innervation to the pineal gland. These inputs are important to entrain melatonin secretion with the light-dark cycle.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 127: Pathology

Discussion:
Intracranial epidural hematomas usually arise from a laceration of the meningeal artery secondary to a skull fracture. The patient may be fully lucid for an extended period of time following the injury while blood accumulates in the epidural space.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 128: Physiology

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

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 129:
Pharmacology/Chemistry

Discussion:
The patient is suffering from restless legs syndrome. Oxycodone, carbidopa/levodopa and pergolide are all effective treatments, but dopamine agonists, such as pergolide, do not produce the daytime rebound restlessness that can occur with carbidopa/levodopa and do not have the addictive potential of oxycodone. Benzodiazepines can be effective, but daytime drowsiness may be a problem for elderly patients placed on diazepam. Pergolide would be the drug of choice.

Reference:
Jankovic J, Tolosa E, Scott-Conner CE, editors. Parkinson's disease and movement disorders. 4rd ed. Baltimore: Williams & Wilkins, 2002.

Question(s) 130: Anatomy

Discussion:
The paraventricular nucleus of the hypothalamus provides a major excitatory input to the preganglionic sympathetic neurons and plays a critical role in integrated responses to stress.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 131: Clinical Adult

Discussion:
The most important initial study to obtain in a patient over the age of 55 suspected of having temporal arthritis is an erythrocyte sedimentation rate.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 132: Physiology

Discussion:
Vertex waves are first seen at 2-3 months of age. The full term neonate has a discontinuous rhythm in quiet sleep (trace alternant). Delta brushes are a pattern of prematurity but may also be seen in a full-term infant. Rare temporal sharp waves may also been seen in the premature infant. Rhythmic anterior slow waves can be seen during sleep in full-term infants.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 133: Pathology

Discussion:
Fatal familial insomnia is a human prion disease. The cause of ALS and corticobasal ganglionic degeneration remains obscure, but transmission studies have failed to support prion etiology. Familial dentatopallidoluyseal degeneration is a genetic disorder characterized by trinucleotide repeat amplification. Striatonigral degeneration is a component of multiple system atrophy.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 134: Clinical Adult

Discussion:
Organophosphate poisoning may occur in patients exposed to pesticides. Intramuscular atropine administration is standard initial therapy.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 135: Anatomy

Discussion:
Normal median somatosensory evoked potentials would be seen with a lesion of the lateral cuneate nucleus. The fasciculus cuneatus contains long ascending branches of the upper six thoracic and all cervical dorsal roots. Axons from the second order neurons in the nucleus cuneatus cross the midline as the internal arcuate fibers and ascend in the medial lemniscus to the ventral posterolateral nucleus. From here, third order neurons send axons through thalami-cortical radiations to the sensory cortex.

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 136: Behavioral

Discussion:
Bupropion has had a low incidence of erectile dysfunction associated with its use. All of the selective serotonin reuptake inhibitors (SSRIs) have been reported to have erectile dysfunction as a side effect. Amitriptyline and venlafaxine also cause erectile dysfunction.

Reference:
Arana GW, Rosenbaum JF. Handbook of psychiatric drug therapy. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 137: Physiology

Discussion:
The sense of vibration is dependent on Pacinian corpuscles.

Reference:
Kandel ER, Schwartz JM, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 138: Behavioral

Discussion:
Lithium, haloperidol, valproate, and electroconvulsive therapy are effective for acute mania. Selegeline is not known to benefit acute mania and may exacerbate the problem because of its monoamine oxidase inhibitory effects.

Reference:
Goldman HH. General psychiatry. Norwalk: Appleton and Lange, 1995.

Question(s) 139: Physiology

Discussion:
Calcium binding in the membrane facilitates acetylcholine release following an action potential.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 140:
Pharmacology/Chemistry

Discussion:
The structural basis of the blood brain barrier (BBB) is the presence of tight junctions (zonula occludens) between capillary endothelial cells. Tight junctions consists of macromolecular complexes, including the transmembrane proteins occludin and claudin, and the associated zonula occludens (ZO) proteins and cingulin.

Reference:
Huber JD, Egleton R, Davis TP. Molecular physiology of tight junctions in the blood-brain barrier. Trends Neurosci 2001;24:719-726.

Question(s) 141:
Pharmacology/Chemistry

Discussion:
Baroreflex failure may occur with peripheral or central lesions that interrupt the baroreceptor reflex circuit, which in normal conditions serves as a buffer against marked oscillations of blood pressure. Causes of baroreflex failure include bilateral carotid sinus denervation, as occurs following surgery or in disorders such as the Guillain-Barré syndrome, or with lesions that affect the nucleus tractus solitarius bilaterally, such as syringobulbia. The syndrome is characterized by episodes of fluctuating hypertension, resembling a pheochromocytoma. Clonidine, a central alpha-2 receptor agonist that inhibits the brainstem sympathoexcitatory drive, is effective in controlling these episodes.

Reference:
Robertson D, Hollister AS, Biaggioni I, et al. The diagnosis and treatment of baroreflex failure. NEJM 1993;329:1449-1455.

Question(s) 142:
Pharmacology/Chemistry

Discussion:
Midodrine is a prodrug that is transformed in the liver to a potent agonist of alpha receptors in arteries and veins. Given its predictable absorption and peak effect, it is the sympathomimetic of choice for treatment of neurogenic orthostatic hypotension. One common side effect is scalp pruritus, a manifestation of alpha adrenergic mediated piloerection. Its most potentially serious side effect is supine hypertension. Midodrine should not be administered late in the evening and patients taking the drug should avoid lying flat.

Reference:
1. Jankovik J, Gilden JL, Hiner BC, et al. Neurogenic orthostatic hypotension: a double-blind, placebo-controlled study with midodrine. Am J Med 1993;95:38-48.
2. Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 143: Anatomy

Discussion:
The preganglionic sympathetic neurons are located in the intermediolateral cell column in all thoracic and the upper two lumbar spinal segments. Thus, they are formed from the neural tube.

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 144: Physiology

Discussion:
Decerebrate rigidity is associated with tonic neuronal activity in the vestibulospinal and pontine reticulospinal nuclei.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 145: Physiology

Discussion:
Permanent diabetes insipidus is produced by destruction of the supraoptic and paraventricular nuclei, which release the antidiuretic hormone vasopressin.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 146: Pathology

Discussion:
Carbon monoxide poisoning may produce bilateral lesions of the globus pallidus. Methanol toxicity may produce lesions of the putamen. Bilateral petechial hemorrhages of the mammillary bodies are seen in acute Wernicke encephalopathy.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 147: Anatomy

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

Reference:
1. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 148: Physiology

Discussion:
Interneurons in the spinal cord are necessary for flexion, inverse myotatic and crossed extension reflexes and for reciprocal innervation. No interneurons are required for the tendon stretch reflex which is monosynaptic.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 149: Anatomy

Discussion:
The largest cerebellar efferent bundle, the superior cerebellar peduncle, is formed by fibers from the dentate, emboliform, and globose nuclei.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 150: Clinical Adult

Discussion:
Lhermitte's sign (an abnormal sensation, usually described as "like an electric shock", precipitated by neck flexion and traveling rapidly down the spine) has been described in a variety of conditions, most of which are associated with demyelination in the posterior columns of the cervical spinal cord. Common associated conditions include multiple sclerosis, radiation myelopathy and subacute combined degeneration of the cord (vitamin B12 deficiency).

Reference:
1. Kanchandani R, Howe JG. Lhermitte's sign in multiple sclerosis: a clinical survey and review of the literature. J Neurol Neurosurg Psych 1982;45:308-312.
2. Gautier-Smith PC. Lhermitte's sign in subacute combined degeneration of the cord. J Neurol Neurosurg Psych 1973;36:861-863.
3. Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 151: Anatomy

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

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 152: Physiology

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

Reference:
1. Ebersole JS, Pedley TA. Current practice of clinical electroencephalography. 3nd ed. New York: Lippincott Williams Wilkins, 2003.
2. Niedermeyer E, Lopes da Silva F. Electroencephalography. 4th ed. Baltimore: Lippincott, Williams & Wilkins, 1998.

Question(s) 153:
Pharmacology/Chemistry

Discussion:
Horner's syndrome results from impaired sympathetic innervation of the eye. Lesions may involve the central (first-order) neurons of the hypothalamus, brainstem and cord, the preganglionic (second-order) neurons of the C8-T2 roots to the level of the superior cervical ganglion or the postganglionic (third-order) neurons as they follow the internal and external carotid arteries to innervate the orbit and vessels of the face respectively. All forms of Horner's syndrome show failure to dilate on exposure to cocaine, which is a norepinephrine reuptake inhibitor. Preganglionic lesions show normal or exaggerated response to hydroxyamphetamine, which releases norepinephrine from nerve terminals. The response is absent or greatly attenuated in postganglionic lesions, where the nerve terminals are destroyed.

Reference:
Miller NR, Newman NJ. Walsh and Hoyt's Clinical neuro-ophthalmology. The essentials. 5th ed. Philadelphia: Lippincott, Williams and Wilkins, 1999.

Question(s) 154: Anatomy

Discussion:
The globus pallidus projects to the ventral anterior and ventral lateral (pars oralis) nuclei of the thalamus.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 155: Anatomy

Discussion:
The amygdalofugal fibers pass through the inferior thalamic peduncle to the magnocellular division of the mediodorsal nucleus.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 156: Clinical Adult

Discussion:
Rest tremor is more common in idiopathic Parkinson's disease (PD) than in diffuse Lewy body disease (DLBD), but can occur in either. Other parkinsonian features occur with equal frequency in the two conditions. Most, but not all, patients with DLBD initially respond to dopaminergic agents, whereas almost all patients with PD respond. Cognitive or psychiatric abnormalities are less common in PD than in DLBD, which typically presents with such symptoms.

Reference:
Louis ED, et al. Parkinsonian features of eight pathologically diagnosed cases of diffuse Lewy body disease. Movement Disorders 1995.

Question(s) 157: Anatomy

Discussion:
The suprachiasmatic nucleus receives direct bilateral projections from the retina and functions as a biologic clock.

Reference:
1. Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 158: Clinical Adult

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 159: Clinical Adult

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

Reference:
1. Caplan LR. Caplan's stroke, a clinical approach. 3rd ed. Boston: Butterworth-Heinemann, 2000.
2. Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 160: Anatomy

Discussion:
The cephalad portion of the neural tube initially forms three vesicles (prosencephalon, mesencephalon, and rhombencephalon) that subsequently segment further to form the telencephalon, diencephalon, mesencephalon, metencephalon, and myelencephalon. The pons arises from the metencephalon.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 161: Pathology

Discussion:
Prion diseases have long incubation times and evoke no host immune response. Immunocompromised individuals are not preferentially affected by prion diseases. The CSF does not show pleocytosis (as it does in most other infections) and neither inflammation or microglial nodules are seen in the brain. Spongiform change, intense gliosis, and neuronal loss unassociated with mononuclear cell inflammation is the characteristic histologic pattern for Creutzfeldt-Jakob disease.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 162: Anatomy

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 163: Behavioral

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

Reference:
Damasio A, Tranel D, Rizzo M. Disorders of complex visual processing. In: Mesulam MM, editor. Principles of behavioral and cognitive neurology. 2nd ed. New York: Oxford University Press, 2000.

Question(s) 164: Anatomy

Discussion:
The anterior lobe of the cerebellum projects to the nucleus interpositus (globose and emboliform nuclei) and fastigial nucleus, regulates ipsilateral extensor muscle tone, receives some input form the pontine nuclei via pontocerebellar tracts, and has its output from the cerebellum via the superior cerebellar peduncle. It has direct (cortical) connections and connections through deep nuclei to the vestibular system.

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 165: Physiology

Discussion:
Activity in the 8 Hz range can be seen in comatose patients with pontine infarctions, i.e., an alpha coma pattern.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 166: Anatomy

Discussion:
The dorsal (posterior) spinocerebellar tract is an uncrossed ascending tract that arises from the large neurons of Clarke's column (dorsal nucleus), which extends from L3 to C8. Clarke's nucleus receives afferent fibers from all parts of the body except the head and neck but is functionally related primarily to the hind limb and caudal parts of the body.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 167: Physiology

Discussion:
Development of a steady force is characteristic of a type I muscle fiber.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 168: Anatomy

Discussion:
Posterior cord lesions would affect muscles innervated by subscapular, thoracodorsal, radial and axillary nerves. The flexor carpi ulnaris is innervated by the ulnar nerve of the medial cord.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 169: Clinical Pediatrics

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

Reference:
Gomez MR, editor. Tuberous sclerosis complex. 3rd ed. New York: Oxford University Press, 1999.

Question(s) 170: Anatomy

Discussion:
In looking downward to the left, when the eye is turned outward, the depressor is the left inferior rectus. When eye is turned inward, the depressor is the right superior oblique.

Reference:
1. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 171: Pathology

Discussion:
The two most common adult tumors in the cerebellopontine angle region are vestibular schwannoma and meningioma. In this case, a meningioma was present. Most meningiomas are strongly positive for epithelial membrane antigen (EMA). In contrast, schwannomas are negative for EMA but strongly positive for S-100 protein. Ultrastructurally, meningiomas show elongated intertwined cell processes joined by large numbers of desmosomes. Schwannomas also show elongated cell processes by electron microcopy, but do not have the prominent intercellular junctions seen in meningiomas; rather, a layer of basal lamina covers the cell processes of schwannoma. Another distinctive ultrastructural feature seen in many schwannomas is the presence of Luse bodies ("long-spacing collagen"), which are fusiform cross-striated structures composed of collagen that has an increased banding periodicity. Antoni type A tissue (densely cellular tumor) and Antoni type B tissue (loose, hypocellular tumor) are characteristic of schwannoma. Palisades of nuclei separated by fibrillar anuclear zones are referred to as Verocay bodies and are typically seen in Antoni type A areas of schwannomas.

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 172:
Pharmacology/Chemistry

Discussion:
Quetiapine is an atypical neuroleptic that, like clozapine, has 5-HT2 receptor blocking activity lower D2 blocking potency than the typical neuroleptic such as haloperidol or chlorpromazine. Therefore, quetiapine and clozapine are the neuroleptic of choice for management of hallucinations in patients with Parkinson's disease. Risperidone, another atypical neuroleptic, has higher D2 receptor blocking activity and thus higher risk of extrapyramidal side effects.

Reference:
Olanow CW, Watts RL, Koller WC. An algorithm (decision tree) for the management of Parkinson's disease (2001). Neurology 2001;56(Suppl 5):S1-S88.

Question(s) 173: Anatomy

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

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 174:
Pharmacology/Chemistry

Discussion:
The medium spiny neurons of the striatum contain, in addition to D1 and D2 dopamine receptors, several other receptors that can potentiate or antagonize the effects of dopamine. The A2 adenosine receptors antagonize the effects of activation of D2 receptors. This may have therapeutic implications, as adenosine receptors are blocked by caffeine.

Reference:
Richardson PJ, Kase H, Jenner PG. Adenosine A2a receptor antagonists as new agents for the treatment of Parkinson's disease. Trends Pharmacol Sci 1997;18:338-344.

Question(s) 175: Physiology

Discussion:
The most common presentation of nemaline myopathy is congenital hypotonia. Affected children are usually quite bright but exhibit a characteristic open-mouth appearance. Extra-ocular muscles are not affected and muscle weakness is static throughout life.

Reference:
Griggs RC, Mendell JR, Miller RG. Congenital myopathies. In: Griggs RC, Miller RG, editors. Evaluation and treatment of myopathies. Philadelphia: FA Davis Co., 1995.

Question(s) 176: Clinical Adult

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 177: Behavioral

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

Reference:
Mendez MF, Cummings JL, Benson DF. Depression in epilepsy. Archives of Neurology 1996;43:766-770.

Question(s) 178: Pathology

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

Reference:
Fuller GN, Goodman JC. Practical review of neuropathology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 179: Behavioral

Discussion:
Wernicke's aphasia is characterized by impairments in comprehension, repetition, reading, writing, and naming, with fluent paraphasic speech.

Reference:
Benson DF. Aphasia, alexia, and agraphia. New York: Churchill Livingston, 1979.

Question(s) 180:
Pharmacology/Chemistry

Discussion:
Tick paralysis usually occurs in young individuals. The paralysis is reversible upon removal of the tick. It is thought to result from a presynaptic neuromuscular block. This results from an impairment of excitation-secretion coupling due to reduced calcium availability.

Reference:
1. Cooper BJ, Spence I. Temperature-dependent inhibition of evoked acetyl choline release in tick paralysis. Nature 1976;263:693-695.
2. Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 181: Behavioral

Discussion:
Topographagnosia, usually combined with some degree of unilateral neglect, most often follows right parietal damage.

Reference:
Grusser O-J, Landis T. Visual agnosias and other disturbances of visual perception and cognition. London: MacMillan Press, 1991.

Question(s) 182: Clinical Adult

Discussion:
Cataplexy, a loss of muscle tone following emotional stimulation, occurs in about 70% of patients with narcolepsy. It is not associated with other conditions that cause excessive daytime somnolence.

Reference:
Kryger MH, Roth T, Dement WC. Principles and practice of sleep medicine. 3rd ed. Philadelphia: WB Saunders Company, 2000.

Question(s) 183: Physiology

Discussion:
During muscle contraction, the filaments slide over one another and the Z bands move closer together.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 184: Behavioral

Discussion:
Dopamine agonists such as bromocriptine have been used to treat the behavioral traits associated with medial frontal syndrome. This syndrome may be seen after ischemia in the distribution of the anterior cerebral artery.

Reference:
Muller U, von Cramon DY. The therapeutic potential of bromocriptine in neuropsychological rehabilitation of patients with acquired brain damage. Prog Neuropsypharm Biol Psych 1994;18:1103-20.

Question(s) 185: Behavioral

Discussion:
Alien hand phenomenon is one of the associated clinical features of corticobasal ganglionic degeneration.

Reference:
Riley DE, Lang AE, Lewis A, et al. Corticobasal ganglionic degeneration. Neurology 1990;40:1203-1212.

Question(s) 186: Behavioral

Discussion:
Flumazenil may be used to reverse the sedation associated with benzodiazepines. This patient was comatose and ventilator-dependent prior to flumazenil and became alert and "weanable" after treatment.

Reference:
Kaplan HI, Sadock BJ. Pocket handbook of psychiatric drug treatment. 2nd ed. Baltimore: Williams and Wilkins, 1996.

Question(s) 187: Behavioral

Discussion:
Alexia without agraphia, also known as pure word blindness, is most often produced by left posterior cerebral artery occlusion that can cause a right homonymous hemianopsia.

Reference:
Mesulam MM. Principles of behavioral and cognitive neurology. 2nd ed. New York: Oxford University Press, 2000.

Question(s) 188: Physiology

Discussion:
Preganglionic autonomic nerve fibers conduct at a velocity of approximately 3-15 meters/second.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 189: Behavioral

Discussion:
Patients with frontotemporal dementia have been shown to manifest a variety of behavioral changes including hoarding of items and nascent musical and/or artistic expression. This combination of behaviors is usually not seen in other degenerative dementias.

Reference:
Miller BL, Cummings JL, Boone K, et al. Emergence of artistic talent in frontotemporal dementia. Neurology 1998;51:978-981.

Question(s) 190: Pathology

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

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 191:
Pharmacology/Chemistry

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

Reference:
diMauro S, De Vivo DC. Diseases of carbohydrate, fatty acid and mitochondrial metabolism. In: Siegel GJ, Agranoff BW, Albers RW, et al, editors. Basic neurochemistry. 6th ed. Philadelphia: Lippincott-Raven, 1999.

Question(s) 192: Physiology

Discussion:
An asymmetric slowing of conduction velocities of pattern reversal visual evoked responses with a unilateral delay of 30 msec is most likely to be associated with optic neuritis.

Reference:
Nuwer MR. Fundamentals of evoked potentials and common clinical applications today. Electroencephalogr Clin Neurophysiol 1998;106:142-148.

Question(s) 193: Physiology

Discussion:
A highly epileptogenic EEG pattern that has a high incidence of associated seizures is the 3 Hz spike-and-wave. Benign EEG patterns that are unassociated with seizures include 6 Hz spike-and-wave, small sharp spikes and wicket spikes.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 194: Clinical Adult

Discussion:
Nitrous oxide abuse may result in a clinical syndrome of myeloneuropathy indistinguishable from that of vitamin B12 deficiency. The mechanism appears to be interference with the vitamin B12-dependent conversion of homocysteine to methionine.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 195: Physiology

Discussion:
Nerve conduction velocities are about half the adult value in term infants and slower in premature infants. They reach adult values by age two.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 196: Physiology

Discussion:
In healthy young adults, REM sleep is approximately 20-25% of total sleep: stage 1 is 5%, stage 2 is 45%, and stage 3 and 4 combined is 25%.

Reference:
Kryger MH, Roth T, Dement WC. Principles and practice of sleep medicine. 3rd ed. Philadelphia: WB Saunders Company, 2000.

Question(s) 197:
Pharmacology/Chemistry

Discussion:
Pyridoxine-dependent seizures are believed to result from diminished activity of glutamic acid decarboxylase (GAD), which is responsive to pharmacologic doses of its cofactor, pyridoxine (vitamin B6). Deficient action of GAD would be expected to produce elevated levels of the excitatory neurotransmitter glutamic acid, with corresponding low levels of gamma aminobutyric acid, the major inhibitory neurotransmitter in the brain. Such findings have been reported in patients with pyridoxine-dependent seizures, both prior to and when off therapy. The cerebrospinal fluid glucose concentration is normal in this condition.

Reference:
Gospe SM. Current perspectives on pyridoxine-dependent seizures. J Pediatr 1998;132:919-923.

Question(s) 198: Physiology

Discussion:
Increased fiber density on single fiber EMG occurs when there has been denervation and reinnervation, resulting in enlargement of the motor unit and a greater likelihood that two or more muscle fibers innervated by a motor unit will be within the recording radius of the single fiber EMG electrode. The correct answer is ALS.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 199: Behavioral

Discussion:
Clinically significant obsessions and compulsions are seen in about half of the individuals with Tourette's syndrome.

Reference:
Frankel M, Cummings JL, Robertson MM, et al. Obsessions and compulsions in Gilles de la Tourette's syndrome. Neurology 1986;36:378-382.

Question(s) 200:
Pharmacology/Chemistry

Discussion:
The termination of synaptic actions of L-glutamate, as with GABA and most monoamines, depends on its active uptake by neurons and glia. The glutamate transporter is energy-dependent and coupled to co-transport of Na+ and K+. Loss of glutamate uptake occurs in patients with amyotrophic lateral sclerosis. The glutamate transporter has been cloned.

Reference:
1. Rothstein JD, Martin LJ, Kuncl RW. Decreased glutamate transport by the brain and spinal cord in amyotrophic lateral sclerosis. NEJM 1992;326:1464-1468.
2. Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 201: Physiology

Discussion:
50% decrease in CMAP amplitude at the elbow compared with the wrist is consistent with conduction block below the proximal point of stimulation and above the distal point of stimulation.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 202: Anatomy

Discussion:
The cranial preganglionic outflow arises from visceral cell groups of the brain stem associated with the oculomotor, facial, glossopharyngeal and vagus nerves. All of these are parasympathetic.

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 203: Anatomy

Discussion:
The contralateral cerebellar hemisphere (in this case, the left cerebellar hemisphere) is engaged with motor tasks planned and executed from the opposite cerebral hemisphere. Activation of a cerebral cortical motor strip is accompanied by activation of the contralateral cerebellar hemisphere. Conversely, if the motor areas of a cerebral hemisphere are damaged, the contralateral cerebellar hemisphere shows a reduction in activity.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 204:
Pharmacology/Chemistry

Discussion:
Ceramide is released in conditions of cell stress and binding of cytokines to death receptors and can activate caspase 3, probably through release of cytochrome c.

Reference:
McDonald ES, Windebank AJ. Mechanisms of neurotoxic injury and cell death. Neurology Clinics 2000;3:525-540.

Question(s) 205:
Pharmacology/Chemistry

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

Reference:
Richelson E. Pharmacology of antidepressants. Mayo Clin Proc 2001;76:511-527.

Question(s) 206:
Pharmacology/Chemistry

Discussion:
The CD4+ helper cells (Th) exist as two distinct subsets, Th1 and Th2. Th1 cells produce proinflammatory cytokines, such as interleukin-2 (IL)-2, tumor necrosis factor (TNF) alpha, and interferon (IF) gamma, which are the major players in cell-mediated immunity and proinflammatory responses. In contrast, Th2 cells produce IL-10, which inhibits the inflammatory effectors of Th1 cells. Interferon beta is produced by fibroblasts and may affect immune balance in favor of an ant-inflammatory responses, by inhibiting production of Th1 cytokines, such as IL-2 and increasing that of Th2 cytokines, such as IL-10.

Reference:
Dalakas M. Basic aspects of neuroimmunology as they relate to immunotherapeutic targets: present and future prospects. Ann Neurol 1995;37(supplement 1):S2-S13.

Question(s) 207: Clinical Adult

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

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 208: Clinical Pediatrics

Discussion:
ADHD and obsessive-compulsive symptoms are often associated with Tourette's syndrome. Boys are more often affected than girls; 96% have symptoms before 11 years. An increase in seizures is not seen. Coprolalia is not necessary for the diagnosis.

Reference:
1. Berg BO, editor. Principles of child neurology. New York: McGraw-Hill, 1996.
2. Menkes JH, Sarnat HB, editors. Child neurology. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 209: Anatomy

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 210: Anatomy

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

Reference:
1. Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.
2. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 3rd ed. Boston: Little, Brown and Co., 1996.
3. Bauby JD. The diving bell and the butterfly. New York: Alfred A. Knopf, 1997.

Question(s) 211: Physiology

Discussion:
A common anomaly of innervation in the leg involves the accessory peroneal nerve. It is recognized when the amplitude of the compound muscle action potential is larger with stimulation at the knee than it is with stimulation at the ankle and confirmed by obtaining a response with stimulation behind the ankle.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 212: Physiology

Discussion:
A lesion in the area ventral to the locus ceruleus in the pontine tegmentum causes REM sleep without atonia.

Reference:
Kryger MH, Roth T, Dement WC. Principles and practice of sleep medicine. 3rd ed. Philadelphia: WB Saunders Company, 2000.

Question(s) 213: Behavioral

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

Reference:
Damasio A, Tranel D, Rizzo M. Disorders of complex visual processing. In: Mesulam MM, editor. Principles of behavioral and cognitive neurology. 2nd ed. New York: Oxford University Press, 2000.

Question(s) 214: Pathology

Discussion:
Seizure disorders frequently are associated with neuronal loss in the hippocampus, in both end-folium and Sommer's sector. The condition is known as mesial temporal sclerosis.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 215: Physiology

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

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 216: Anatomy

Discussion:
A unilateral lesion of the ventrocaudal pons results in ipsilateral lateral rectus and facial paresis and a contralateral facial sparing hemiparesis. This is known as the Millard-Gubler syndrome.

Reference:
1. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 217: Clinical Adult

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

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 218: Anatomy

Discussion:
The locus ceruleus is the principal location of noradrenergic neurons in the central nervous system (CNS). These noradrenergic neurons project widely throughout the CNS and appear to be important in regulation of the sleep-wake cycle and attentional mechanisms.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 219: Clinical Pediatrics

Discussion:
At birth the median head circumference of a normal term infant is 35 cm. A head circumference of 45 cm is greater than the 98th percentile. In the absence of other cerebral malformations, congenital aqueductal stenosis is the most common cause of noncommunicating hydrocephalus. A familial X-linked form accounts for 2% of these. Increased intracranial pressure in the context of hypoxic ischemic encephalopathy does not cause increased head circumference at birth. Klippel-Feil syndrome (fusion of the cervical vertebra) may be associated with Chiari malformation but is a much less common cause of hydrocephalus. Choroid plexus papillomas are typically located in one lateral ventricle and become symptomatic after the perinatal period, usually by obstructing ventricular outflow. Lissencephaly is typically associated with microcephaly.

Reference:
Fenichel GM. Clinical pediatric neurology: a signs and symptoms approach. Philadelphia: WB Saunders, 1997.

Question(s) 220: Neuroimaging

Discussion:
Enhancing ring lesion is a non-specific finding and may be due to abscess, tumor, cysticercosis cyst or resolving hematoma. Although there are certain neuroimaging characteristics that suggest the cause, clinical history or pathology findings are needed for definite diagnosis.

Reference:
Salzman C, Tuazon CU. Value of the ring enhancing sign in differentiating intracerebral hematoma and brain abscess. Arch Intern Med 1987;147:951-2.

Question(s) 221: Physiology

Discussion:
For depolarization to occur, Na+ flows inward and K+ flows outward.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 222: Pathology

Discussion:
Ethylene glycol toxicity has birefringent calcium oxalate deposits in and around blood vessels. Oxalic acid crystals may be detected in the urine.

Reference:
Ellison D, Love S. Neuropathology: a reference text of CNS pathology. Chicago: CV Mosby, Inc., 1998.

Question(s) 223: Behavioral

Discussion:
Impulsive and antisocial behavior are often seen after lesions of the orbitofrontal cortex. Akinetic mutism is seen with medial frontal lesions. Depression and apathy are more commonly seen with left dorsolateral frontal lesions or subcortical caudate lesions. Hyposexuality is more common with temporal lobe lesions.

Reference:
Feinberg TE, Farah MJ. Behavioral neurology and neuropsychology. New York: McGraw Hill, 1997.

Question(s) 224: Behavioral

Discussion:
The majority of structural lesions associated with development of obsessive-compulsive behavior have involved the frontal lobe and/or frontal-basal ganglia network connections.

Reference:
Swoboda KJ, Jenike MA. Frontal abnormalities in a patient with obsessive-compulsive disorder. Neurology 1995;45:2130-2134.

Question(s) 225: Clinical Adult

Discussion:
A spinocerebellar syndrome with ataxia, ophthalmoparesis, and Babinski signs developing in a patient with a long history of fat malabsorption suggests a deficiency of vitamin E.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 226: Pathology

Discussion:
Paracrystalline intramitochondrial inclusions are the most definitive ultrastructural indicators of abnormal mitochondria. Tubular cristae, calcium inclusions and fibrillary background may be seen in normal mitochondria. Viral inclusions are not seen in mitochondria.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 227: Anatomy

Discussion:
Ascending fibers that reach the medulla via the fasciculus gracilis and cuneatus terminate in the nucleus cuneatus and nucleus gracilis. From there, axons sweep ventromedially as the internal arcuate fibers, cross the midline and then continue upward as the medial lemniscus.

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 228: Clinical Adult

Discussion:
Cerebral malaria most commonly manifests with seizures and altered consciousness. Focal deficits and movement disorders occur less commonly. Cerebral malaria is almost always caused by P. falciparum. It should be suspected in patients with a history of travel to endemic areas.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 229: Behavioral

Discussion:
Many authorities consider prosopagnosia as a more subtle form of visual agnosia and almost all patients with visual agnosia show difficulty in recognizing familiar faces.

Reference:
Damasio A, Tranel D, Rizzo M. Disorders of complex visual processing. In: Mesulam MM, editor. Principles of behavioral and cognitive neurology. 2nd ed. New York: Oxford University Press, 2000.

Question(s) 230: Physiology

Discussion:
A nocturnal seizure in an eight-year-old child is most likely due to a benign focal epilepsy of childhood, associated with centrotemporal spikes on the EEG.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 231: Anatomy

Discussion:
The central tegmental tract consists of descending fibers from midbrain nuclei that project to the inferior olivary complex and ascending fibers from the lower brain stem that project to thalamic nuclei.

Reference:
1. Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 232: Clinical Adult

Discussion:
Chronic paroxysmal hemicrania is characterized by frequent, short-lived, unilateral, fronto-temporal headaches with ocular and nasal autonomic symptoms. Jaw claudication is a feature of temporal arteritis. Horner’s syndrome is present in chronic paroxysmal hemicrania but abducent nerve palsies do not occur.

Reference:
Victor M, Ropper A. Adams and Victor's principles of neurology. 7th ed. New York: McGraw-Hill, Co., 2001.

Question(s) 233: Anatomy

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

Reference:
1. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.
2. Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 234: Physiology

Discussion:
The diagnosis is brachial plexopathy of the upper trunk. In brachial plexopathy, needle EMG demonstrates abnormality in muscles innervated by a trunk or cord and paraspinal muscles are spared.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 235: Clinical Adult

Discussion:
Essential tremor is typically characterized by bilateral postural tremor involving the upper extremities. Head titubations and vocal tremor are also seen.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 236: Anatomy

Discussion:
In the adult, spinal cord segments are not all adjacent to the corresponding vertebral body. The tenth thoracic spinal cord segment is adjacent to the eighth thoracic vertebral body.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 237: Clinical Adult

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 238: Clinical Pediatrics

Discussion:
While spinal muscular atrophy usually does not present with arthrogryposis (congenital joint contractures), it is one of the most common etiologies of this finding. Many other neuromuscular conditions can present with arthrogryposis including myotonic dystrophy and congenital fiber type disproportion. Uterine restraint and uterine bands are also frequent causes of arthrogryposis, but these conditions do not usually have severe hypotonia.

Reference:
Volpe JJ. Neurology of the newborn. 4th ed. Philadelphia: WB Saunders, 2001.

Question(s) 239:
Pharmacology/Chemistry

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

Reference:
Wevers RA, De Rijk-Van Andel JF, et al. A review of biochemical and molecular genetic aspects of tyrosine hydroxylase deficiency including a novel mutation (291delC). J Inherit Metab Dis 1999;22:364-373.

Question(s) 240: Clinical Adult

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

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 241: Pathology

Discussion:
Combined loss of heterozygosity for chromosomes 1p and 19q in anaplastic oligodendrogliomas is associated with increased responsiveness to chemotherapy.

Reference:
Kleihues P, Cavenee WK, editors. Pathology and genetics of tumours of the nervous system. Lyon: International Agency for Research on Cancer, 2000.

Question(s) 242: Physiology

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

Reference:
Chiappa KH. Evoked potentials in clinical medicine. 3rd ed. New York: Lippincott-Raven Press, 1997.

Question(s) 243: Behavioral

Discussion:
Diffuse Lewy body dementia is characterized by dementia with cortical impairments, parkinsonism, vivid visual hallucinations, heightened sensitivity to neuroleptics, and a fluctuating clinical course with periods of episodic confusion mixed with periods of lucidity.

Reference:
McKeith IG, Galasko D, Levsaka K, et al. Consensus guidelines for the clinical and pathological diagnosis of dementia with Lewy bodies (DLB): report of the consortium on DLB international workshop. Neurology 1996;47:1113-1124.

Question(s) 244: Physiology

Discussion:
The convention in EEG is that if the first electrode of the pair is surface negative, the deflection goes up. If there is a focal area of surface negativity or positivity, there is a phase reversal in adjacent channels at the common electrode.

Reference:
Fisch B. Spehlmann's EEG primer. 2nd ed. Amsterdam: Elsevier Science Publications, 1991.

Question(s) 245: Pathology

Discussion:
Visual disturbances due to lesions around the optic nerve; and polydipsia, polyuria, somnolence or obesity due to involvement of the hypothalamus and pituitary gland are among the most common manifestations of central nervous system sarcoidosis.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 246: Physiology

Discussion:
A given skeletal muscle fiber discharges once in response to an action potential, fibrillates following denervation, is part of only one motor unit, has a conduction velocity of about 5 m/sec, and is innervated by either a 'fast' or 'slow' axon, but not both.

Reference:
Kandel ER, Schwartz JH, Jessel TM. Principles of neural science. 4th ed. New York: McGraw-Hill, 2000.

Question(s) 247: Clinical Pediatrics

Discussion:
DSM IV criteria for the diagnosis of Asperger's disorder include: A. Qualitative impairment in social interaction as manifested by at least two of the following: 1. Marked impairment in the use of multiple nonverbal behaviors, such as eye-to-eye gaze, facial expression, body postures and gestures to regulate social interaction. 2. Failure to develop peer relationships appropriate to developmental level 3. A lack of spontaneous seeking to share enjoyment, interests or achievements with other people 4. Lack of social or emotional reciprocity B. Restricted repetitive and stereotyped patterns of behavior, interests and activities, as manifested by at least one of the following: 1. Encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus, 2. Apparently inflexible adherence to specific, nonfunctional routines or rituals, 3. Stereotyped and repetitive motor mannerisms (e.g. hand or finger flapping or twisting, or complex whole body movements), 4. Persistent preoccupation with parts of objects C. The disturbance causes clinically significant impairment in social, occupational, or other important areas of functioning D. There is no clinically significant general delay in language (i.e. single words used by age 2 years, communicative phrases used by age 3 years). E. There is no clinically delay in cognitive development or in the development of age-appropriate self-help skills, adaptive behavior (other than in social interaction), and curiosity about the environment in childhood. F. Criteria are not met for another specific Pervasive Developmental Disorder or Schizophrenia. The criteria do not include delay in cognitive development, language acquisition, or in development of self-help adaptive skills. Stereotyped and repetitive use of language is not a criterion for Asperger syndrome. These are all differences that distinguish Asperger syndrome from autistic disorder.

Reference:
American Psychiatric Association. Diagnostic and statistical manual of mental disorders. 4th rev. ed. Washington: American Psychiatric Association, 2000.

Question(s) 248:
Pharmacology/Chemistry

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

Reference:
Grewal RP. Stroke in Fabry's disease. J Neurol 1994;241:153-156.

Question(s) 249: Physiology

Discussion:
Generalized beta activity (activity over 14 Hz) can be seen as a consequence of drug use, particularly benzodiazepines or barbiturates.

Reference:
Niedermeyer E, Lopes da Silva F, editors. Electroencephalography: basic principles, clinical applications, and related fields. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 250: Anatomy

Discussion:
Descending fibers in the spinal trigeminal tract convey impulses concerned with pain, thermal and tactile sense from the face, forehead and mucous membranes of the nose and mouth. The spinal trigeminal tract and nucleus, pars caudalis, are the only parts uniquely concerned with the perception of pain and thermal sense.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 251: Anatomy

Discussion:
The cochlear nerve originates from the spiral ganglion, enters the brainstem at the cerebellopontine angle and terminates on the dorsal and ventral cochlear nuclei. The pattern of tonotopic localization is evident in both the nerve fibers and their termination in the cochlear nuclei.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 252: Pathology

Discussion:
Central neurocytoma is a mature neuronal intraventricular tumor, which is microscopically indistinguishable from oligodendroglioma. Most tumors called intraventricular oligodendrogliomas in the past were probably central neurocytomas. Confirmation is provided by immunohistochemical demonstration of neuronal antigens or electron microscopic identification of neuronal features, such as cytoplasmic dense core granules.

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 253:
Pharmacology/Chemistry

Discussion:
Gilles de la Tourette's syndrome may be worsened by stimulants such as methylphenidate. Drugs used for treatment include haloperidol, pimozide and clonidine.

Reference:
Jankovic J. Tourette's syndrome. NEJM 2001;345:1184-1192.

Question(s) 254: Physiology

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

Reference:
Ebersole JS, Pedley TA. Current practice of clinical electroencephalography. 3rd ed. New York: Lippincott Williams and Wilkins. 2002.

Question(s) 255: Pathology

Discussion:
Homer Wright rosettes contain the cytoplasmic processes (of the tumor cells) in the center of the rosette. This type of rosette is commonly seen in neuroblastoma, medulloblastoma, and other PNETs. In contrast, Flexner-Wintersteiner rosettes have a central lumen formed by the tumor cells. This type of rosette is seen in retinoblastoma as well as other types of PNET.

Reference:
Fuller GN, Goodman JC. Practical review of neuropathology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 256: Clinical Pediatrics

Discussion:
Prenatal cytomegalovirus infection can cause severe cerebral injury, such as microcephaly, microgyria, cerebral calcifications and chorioretinitis. Minimally affected children can have only hearing loss.

Reference:
Menkes JH, Sarnat HB, editors. Child neurology. 6th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 257: Clinical Pediatrics

Discussion:
Neonatal tetanus results from infection of the umbilical stump by Clostridium tetani, whose exotoxin selectively inhibits inhibitory synapses in the CNS. The enhanced excitability of the nervous system produces the characteristic muscular spasms with trismus, facial rigidity, opisthotonus and poor feeding. Children born at home to unimmunized mothers are at greatest risk. The pupils are spared in neonatal tetanus, in contrast to infantile botulism, another Clostridial disease.

Reference:
Volpe JJ. Neurology of the newborn. 4th ed. Philadelphia: WB Saunders, 2001.

Question(s) 258: Clinical Pediatrics

Discussion:
A minority of children with perinatal asphyxia causing selective injury to the basal ganglia does not develop symptoms and signs for periods ranging for 7-14 years. (Mean 12.9). Nearly one half of these children have early normal development, and 80% have normal intellect. Dystonia continues to progress for a mean of 7 years after onset, although most children do not become wheelchair bound.

Reference:
Volpe JJ. Neurology of the newborn. 4th ed. Philadelphia: WB Saunders, 2001.

Question(s) 259: Clinical Pediatrics

Discussion:
Fragile X syndrome exhibits a transmission pattern of X-linked inheritance with no male to male transmission. However, both males and females can be affected. In contradistinction to most X-linked recessive disorders, males carrying the mutant gene do not always have manifestations of the disorder but can pass the gene on to their daughters.

Reference:
Swaiman KF, Ashwal S, editors. Pediatric neurology. 3rd ed. St. Louis: Mosby, 1999.

Question(s) 260: Clinical Pediatrics

Discussion:
Pyridoxine (vitamin B6) dependent seizures usually begin in the neonatal period but can appear up to a year of age. The diagnosis is made in the infant by cessation of seizures after parenteral administration of 100 mg of pyridoxine. It is an autosomal recessive disorder and requires life-long treatment with pyridoxine.

Reference:
Berg BO. Principles of child neurology. New York: McGraw-Hill, 1996.

Question(s) 261: Clinical Pediatrics

Discussion:
The ketogenic diet is a mainstay in the treatment of glucose transporter type 1 defect. Acetazolamide in conjuction with the diet can produce a symptomatic metabolic acidosis and therefore should should not be used when the diet is initiated and only with caution while a child is on the diet. Topirimate also is a weak carbonic anhydrase inhibitor and the physician should be aware that it also can lead to a metabolic acidosis.

Reference:
Swaiman KF, Ashwal S, editors. Pediatric neurology. 3rd ed. St. Louis: Mosby, 1999.

Question(s) 262 - 264: Physiology

Discussion:
Low amplitude compound muscle action potentials are characteristic of Lambert-Eaton myasthenic syndrome. An amplitude decrement of compound muscle action potentials to low rates of repetitive stimulation is characteristically seen in myasthenia gravis and Lambert-Eaton myasthenic syndrome. F-waves are usually present in neuromuscular junction disorders.

Reference:
Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 265 - 269:
Pharmacology/Chemistry

Discussion:
Several brain neoplasms are associated with either impaired expression of tumor suppressor genes or increased activity of oncogenes. Mutations of the p53 gene (chromosome 17p) occur in 40% of all astrocytic tumors, particularly in young adults. Mutations in the pRB gene (chromosome 13q), encoding for a protein that controls the G1-S phase transition during the cell cycle, occurs in patients with retinoblastoma. Mutations of the NF-1 gene (chromosome 17q) encoding for neurofibromin (which inhibits the ras-oncogene pathway via activation of ras-GTPase) occurs in neurofibromatosis type 1. Other genetic disorders associated with brain tumors are neurofibromatosis type 2, associated with mutations of the gene in chromosome 22q encoding for merlin (a cytoskeletal protein), and tuberous sclerosis, associated with mutations in the gene encoding for tuberin, a protein of unknown function.

Reference:
Hill JR, Kuriyama N, Kuriyama H, et al. Molecular genetics of brain tumors. Arch Neurol 1999;56:439-441.

Question(s) 270 - 271: Physiology

Discussion:
The F-wave has a variable latency. The H-reflex is dependent on the integrity of the proximal segment of the sensory and motor roots.

Reference:
1. Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.
2. Kleihues P, Cavenee WK, editors. Pathology and genetics of tumors of the nervous system. 2nd ed. New York: Oxford University Press, 2000.

Question(s) 272 - 274: Pathology

Discussion:
Temporal lobe (uncal) herniation compresses the posterior cerebral artery, causing posterior cerebral artery infarction and cortical blindess. Leg weakness follows cingulate herniation and entrapment of the anterior cerebral artery with occasional infarctions. Tonsillar herniation will result in cardiorespiratory arrest.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 275 - 279:
Pharmacology/Chemistry

Discussion:
Genetic disorders affecting distinct subunits of muscle ion channels are associated with specific disturbances of muscle membrane excitability. Hyperkalemic periodic paralysis and paramyotonia congenita are due to different mutations of the alpha subunit of the voltage-gated sodium channel. Hypokalemic periodic paralysis is due to a mutation in the dihydropyridine (DHP) receptor. Myotonia congenita is associated with a mutation of the chloride channel.

Reference:
Lehmann-Horn F, Jurkatt-Roth K. Voltage-gated ion channels and hereditary disease. Physiol Rev 1999;79:1317-1372.

Question(s) 280 - 282: Clinical Pediatrics

Discussion:
Niemann-Pick disease type A can have cherry red macula, associated with hepatosplenomegaly and mental deterioration. Children with the neuronal ceroid lipofuscinosis exhibit progressive dementia and seizures. All of the various forms have abnormal visual evoked potentials and electroretinograms. Hurler syndrome is one of the mucopolysacharidoses, characterized by slowed development, coarse facies and bony abnormalities. Kyphosis appears early.

Reference:
Menkes JH, Sarnat HB, editors. Child neurology. 6th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 283 - 287: Behavioral

Discussion:
Lesions in the hippocampus produce a declarative memory deficit. Docility may be seen with bilateral lesions of the amygdala (part of the Klüver-Bucy Syndrome). Lesions of the anterior cingulate may result in loss of motivation. Anosognosia (denial of illness or impairment) is seen with right parietal lesions. Depression is commonly associated with left prefrontal lesions.

Reference:
1. Feinberg TE, Farah M. Behavioral neurology and neuropsychology. New York: McGraw-Hill, 1997.
2. Mesulam MM. Principles of behavioral and cognitive neurology. 2nd ed. New York: Oxford University Press, 2000.

Question(s) 288 - 291: Pathology

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

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 292 - 295: Pathology

Discussion:
Medulloblastomas frequently disseminate via the subarachnoid space to the spinal cord, forming "drop metastases." Ependymomas comprise about 60% of primary spinal cord neoplasms. Most originate from within the cord proper; however, the myxopapillary ependymoma usually arises from the filum terminale. Oligodendrogliomas account for 10-15% of all gliomas. They are often slow growing and calcified and may undergo spontaneous hemorrhage. The most common glioma of older adults is glioblastoma.

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 296 - 299: Clinical Pediatrics

Discussion:
Krabbe's disease and metachromatic leukodystrophy are autosomal recessive lysosomal enzyme disorders, occuring in both males and females. In addition to a central myelin abnormality the myelin of peripheral nerves is also affected, so that motor nerve conduction velocities are slowed and CSF protein concentration is elevated. Seizures are not prominent in either disorder and if present occur late in the disease.

Reference:
Menkes JH, Sarnat HB, editors. Child neurology. 6th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 300 - 302: Pathology

Discussion:
Muscle biopsy findings reflect the underlying biochemical abnormalities. In carnitine deficiency, there is a defect of transport of fatty acids into the mitochondria, and therefore lipid accumulates in the muscle. In progressive external ophthalmoplegia, large scale deletions of mitochondrial DNA lead to the subsarcolemmal accumulations seen on modified Gomori trichrome stain as ragged red fibers. In the periodic paralyses, there is myofiber vacuolization during and immediately following attacks; the muscle biopsy may be normal in between episodes of weakness. The molecular basis of hypokalemic periodic paralysis is sodium channel mutation, but the cause of the vacuoles is still unclear.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 303 - 306:
Pharmacology/Chemistry

Discussion:
Several biological toxins can produce acute or subacute neurologic manifestations. Botulinum toxin prevents release of acetylcholine by hydrolyzing proteins in the synaptic vesicle (synaptobrevin) or the terminal membrane (SNAP-25, syntaxin) involved in exocytosis. Saxitoxin and tetrodotoxin block voltage-gated sodium channels. Mushroom toxins, such as those from Amanita muscaria, block both cholinergic and GABA synapses. Latrotoxin, the venom of the black widow spider, produces acute release of acetylcholine from synaptic terminals. Toxins of the chickpea (Lathyrus) including alpha-amino-beta-oxalaminopropionic acid activate glutamate receptors resulting in excitotoxicity.

Reference:
Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 307 - 311: Behavioral

Discussion:
Valproate is commonly used to treat bipolar disorder. Paroxetine may be used to treat obsessive-compulsive disorders. Buspirone is an anxiolytic agent. Benztropine is indicated for acute dystonic reactions. Bromocriptine may be used to treat neuroleptic malignant syndrome.

Reference:
Arana GW, Rosenbaum JF. Handbook of psychiatric drug therapy. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 312 - 317:
Pharmacology/Chemistry

Discussion:
Several inherited neurologic disorders are due to mutations in genes encoding for critical proteins. Familial hemiplegic migraine (as well as episodic ataxia type 2, EA-2) is related to mutation in the gene encoding for the alpha subunit of the P/Q calcium channel. Spinocerebellar degeneration occurs in families with a genetic defect in vitamin E metabolism. Charcot-Marie-Tooth disease may occur as a manifestation of several genetic defects. The X-linked form is due to mutation in the connexin-2 gene (connexin is a gap junction protein of peripheral myelin). DOPA-responsive dystonia is due to a defect in guanosine triphosphate (GTP) cyclohydrolase; this enzyme is necessary for formation of tetrahydrobiopterin, the cofactor for tyrosine hydroxylase (the key enzyme for dopamine synthesis). Emerin is a protein that localizes to the inner nuclear membrane, and is deficient in Emery-Dreifuss muscular dystrophy, an X-linked disorder that presents in the first decade with toe walking, followed by progressive joint contractures and cardiac conduction defects that may lead to sudden death. Hamartin is the gene product of TSC-1, one of two genes that are associated with the tuberous sclerosis complex. facial angiofibromas, hypopigmented macules, periungual and subungual fibromas, cavities, cortical tubers, renal angiomyolipomas, cardiac rhabdomyomas and subependymal giant cell astrocytomas are included in the diverse manifestations of this multisystem disease.

Reference:
1. Terwindt GM, Ophoff RA, Haan J, et al. Familial hemiplegic migraine: a clinical comparison of families linked and unlinked to chromosome 19. DMG RG. Cephalagia 1996;16:153-155.
2. Murakami T, Garcia CA, Reiter LT, et al. Charcot-Marie-Tooth disease and related inherited neuropathies. Medicine 1996;75:233-250.
3. Ouahchi K. Arita M, Kayden H, et al. Ataxia with isolated vitamin E deficiency is caused by mutations in the alpha-tocopherol transfer protein. Nat Genet 1995;9:141-145.
4. Worton RG, Molnar MJ, Brais B, et al. The muscular dystrophies. In: Scriver CR, Beaudet AL, Sly WS, et al, editors. The molecular and metabolic bases of inherited diseaase. 8th ed. New York: McGraw-Hill, 2001.
5. Sampson JR. Tuberous sclerosis. In: Scriver CR, Beaudet AL, Sly WS, et al, editors. The molecular and metabolic bases of inherited diseaase. 8th ed. New York: McGraw-Hill, 2001.

Question(s) 318 - 321: Pathology

Discussion:
Epidural hematomas are associated with lacerations of the meningeal arteries (most often the middle meningeal artery). They are associated with skull fractures in approximately 85% of cases. Subdural hematomas are more common than epidural hematomas, and they are associated with tearing of bridging veins. Subdural hematomas can be associated with skull fractures as well, but this is a less frequent association (when compared to epidural hemorrhage). Subarachnoid hemorrhage is seen in traumatic brain injury, but important differential diagnostic considerations include rupture of a saccular aneurysm or vascular malformation.

Reference:
Prayson R. Neuropathology review. Totowa: Humana Press Inc., 2001.

Question(s) 322 - 326: Anatomy

Discussion:
The entorhinal cortex is the gateway for neocortical multimodal information to enter the hippocampal circuit. It projects via the perforant pathway to the dentate gyrus, and granule cells of this region give rise to mossy fibers that innervate the CA3 region. CA3 neurons project to CA1 neurons via the Schaffer collaterals. The subiculum, and not the CA1 area, provides most of the axons of the fornix. The mammillary bodies project to the anterior thalamic nuclei via the mamillothalamic tracts.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 327 - 331:
Pharmacology/Chemistry

Discussion:
The management of parkinsonism includes non-dopaminergic drugs as adjuvants for a variety of non-motor manifestations. Midodrine (and fludrocortisone) helps in the management of orthostatic hypotension in patients with multiple system atrophy; clonazepam helps patients with rapid eye movement sleep behavior disorder; trazodone is helpful for management of insomnia and depression; quetiapine or olanzapine aid management of psychosis; and anticholinergic drugs, such as trihexiphenydil, may help reduce drooling. Anticholinergic drugs are relatively contraindicated in elderly patients as they can worsen cognitive function and trigger urinary retention.

Reference:
Olanow CW, Koller WC, editors. An algorithm (decison tree) for the management of Parkinson's disease. Treatment guidelines. Neurology 1998;50(suppl 1):S1-S57.

Question(s) 332 - 334: Behavioral

Discussion:
Apolipoprotein E4 associated Alzheimer's disease is linked to chromosome 19. Frontotemporal dementia has been linked to chromosome 17. Presenilin 2 associated dementia is linked to chromosome 1.

Reference:
1. Coslett HB. Behavioral neurology/higher cortical function. Seminars in Neurology 2000;20:4.
2. Grabowski TJ, Anderson SW, Cooper GE. Disorders of cognitive function. Continuum 2002;8:2.

Question(s) 335 - 339: Anatomy

Discussion:
The posterior cord of the brachial plexus gives off the subscapular, thoracodorsal, radial and axillary nerves. The medial cord gives off the medial head of the median nerve and ulnar nerve. The lateral cord gives off the lateral head of the median nerve and the musculocutaneous nerve.

Reference:
Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. 4th ed. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 340 - 342: Clinical Pediatrics

Discussion:

The seizures in the benign rolandic epilepsy of childhood characteristically occur in sleep with facial twitching and drooling. Spike discharges arise from the central or centrotemporal region. In Lennox-Gastaut syndrome, the EEG typically demonstrates 2 Hz spike-and-wave abnormalities; there often multiple types of seizures (tonic, atonic and atypical absence) and most patients are mentally retarded. The classic EEG abnormality seen in the early stages of infantile spasms is hypsarrhythmia. 10-16 Hz polyspikes, maximal over frontal areas is seen in juvenile myoclonic epilepsy. 3 Hz spike-and-wave is characteristic of the EEG in childhood absence epilepsy.

Reference:

Pellock JM, Dodson WE, Bourgeois BFD, editors. Pediatric epilepsy, diagnosis and treatment. 2nd ed. New York: Demos, 2001.

Question(s) 343 - 348: Clinical Pediatrics

Discussion:

Sodium valproate is a reasonable medication to use first in generalized absence seizures, juvenile myoclonic epilepsy, and Lennox Gastaut syndrome. Some authorities recommend ethosuximide as the first line medication for generalized absence seizures because of its low side effect profile, but it does not protect against generalized tonic, clonic, or tonic clonic seizures. Lennox Gastaut is very difficult to treat but sodium valproate is a reasonable first line medication. Carbemazepine is a reasonable first line medication for simple and complex partial seizures. Treatment with anticonvulsants may not be necessary in benign rolandic epilepsy. ACTH is the usual starting treatment for infantile spasms in the United States. Vigabatron also has been proven efficacious for infantile spasms.

Reference:

Swaiman KF, Ashwal S, editors. Pediatric neurology. 3rd ed. St. Louis: Mosby, 1999.

Question(s) 349 - 354:
Pharmacology/Chemistry

Discussion:

Ataxia telangiectasia presents in childhood, and is associated with scleral telangiectasias, oculomotor apraxia and chorea. Blood levels of alpha-fetoprotein and carcinoembryonic antigen are high, with variable depression of 1gA, 1gE and 1gG subclasses. Wilson disease may present in children with hepatic disease, hemolytic anemia or renal tubular dysfunction. Neurologic dysfunction usually begins in the teenage years, with basal ganglia and cognitive dysfunction. Kayser-Fleischer rings (representing copper deposition in Descemet's membrane of the cornea) are characteristic. Serum copper and ceruloplasmin levels are low.Relsum disease results from phytanic acid oxidase deficiency, and is associated with elevated levels of phytanic acid in the blood. Patients develop peripheral neuropathy with ataxia, deafness and visual loss with pigmentary retinopathy.Metachromatic leukodystrophy classically presents in late infancy or early childhood with ataxia, spasticity, optic atrophy and dementia. Reflexes are lost because of demyelinating peripheral neuropathy. Arylsulfatase A deficiency is diagnostic, but must be distinguished from pseudodeficiency. Mitochondrial cytopathies have protean manifestations, with progressive external ophthalmoplegia, deafness, pigmentary retinopathy, ataxia and short stature as frequent findings. Deletions or mutations in mitochondrial or nuclear DNA cause impaired oxidative phosphorylation, usually associated with elevated levels of lactic acid in blood and CSF. Both primary sulfite oxidase deficiency, and deficiency of this enzyme secondary to molybdenum cofactor deficiency are associated with early onset, often intractable seizures, profound developmental delay, microcephaly and ectopia lentis (often not apparent until two years of age). Sulfites are typically present in fresh urine specimens.

Reference:

Scriver CR, Beaudet AL, Sly WS, et al, editors. The molecular and metabolic bases of inherited disease. 8th ed. New York: McGraw-Hill, 2001.

Question(s) 355 - 360:
Pharmacology/Chemistry

Discussion:
The complex, interacting motor control systems responsible for eye movements are susceptible to the effects of drugs and toxins at many levels. The following associations have been reported:benzodiazepines - divergence paralysis; carbamazepine - downbeat paralysis; cocaine – opsoclonus; phenothiazines – internuclear ophthalmoplegia; phenytoin – periodic alternating nystagmus; tobacco – upbeat nystagmus.It should be remembered that some agents (such as phenytoin) might produce several types of abnormal eye movements. The correct answers to this item represent the best matches between the drugs or toxins and the eye movements listed.

Reference:
Leigh RJ, Zee DS. The neurology of eye movements. 3rd ed. New York: Oxford University Press, 1999.

Question(s) 361 - 363: Clinical Pediatrics

Discussion:
Subdural effusions are commonly seen with bacterial meningitis in children under the age of two, regardless of the organism. Tuberculous meningitis most commonly manifests as a caseous meningitis resulting from hematogenous spread from a pulmonary focus with frequent cranial nerve involvement. Pyogenic abscesses result from one of three routes of infection: (1) bloodstream, either from sepsis or from cardiopulmonary shunt, most often cyanotic congenital heart disease; (2) contiguous infections such as those of the sinuses or middle ear; or (3) penetrating wounds.

Reference:
Menkes JH, Sarnat HB, editors. Child neurology. 6th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 364 - 366: Clinical Pediatrics

Discussion:
Neurofibromatosis type I and tuberous sclerosisis are transmitted by autosomal dominant inheritance. Incontinentia pigmenti is an X-linked dominant disorder (lethal in males).

Reference:
Menkes JH, Sarnat HB, editors. Child neurology. 6th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 367: Clinical Adult

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

Reference:
1. Bradley WG, Daroff RB, Fenichel GM, et al. Neurology in clinical practice. 3nd ed. Boston: Butterworth-Heinemann, 1999.
2. Panitch H. et al. Randomized comparative study of interferon beta-1a treatment regimens in MS - the EVIDENCE trial. Neurology 2002;59:1507-1517.

Question(s) 368 - 369: Clinical Adult

Discussion:

Trigeminal neuralgia is characterized by paroxysms of shock-like pains lasting about a second and often occurring in volleys, usually beginning at the maxilla or mandible. The pain can often be provoked by light touch in a trigger zone. Baclofen and a variety of anticonvulsant medications are often effective treatment. Trigeminal neuralgia most often begins in the sixth or seventh decade of life; onset below age 40 increases the likelihood that multiple sclerosis is the cause.

Reference:

Bradley WG, Daroff RB, Fenichel GM, et al, editors. Neurology in clinical practice. 3rd ed. New York: Butterworth-Heinemann, 1999.

Question(s) 370:

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:

1. O'Donoghue JL. Neurotoxicity of industrial and commercial chemicals. Boca Raton: CRC Press, 1985.
2. Heimburger DC. Nutrition's interface with health and disease. In: Bennett JC, Plum F, editors. Cecil textbook of medicine. Philadelphia: WB Saunders, 1996.

Question(s) 371 - 372: Behavioral

Discussion:

Alzheimer's disease is the most common degenerative dementia. It is progressive with deficits occurring in memory, visual spatial skills, language, higher executive function, and personality. Primary motor and sensory functions are usually preserved until later in the disease. Language dysfunction early in the disease often resembles a transcortical sensory aphasia. Frontotemporal dementia usually presents with profound changes in personality with relative early preservation of other cognitive functions. Progressive posterior cortical atrophy would have profound visual spatial impairment with relative sparing of other cognitive functions. Diffuse Lewy body dementia has prominent extrapyramidal features and visual hallucinations. Pick's disease demonstrates a profound change in personality and behavior while other cognitive functions continue to remain intact early.SPECT scan early in Alzheimer's disease demonstrates hypoperfusion in bilateral temporal parietal areas. Anticholinesterases such as donepezil, rivastigmine, and galantamine, as well as alpha-tocopherol (vitamin E) are recommended treatments of Alzheimer's disease.

Reference:

1. Cummings JL, Benson DF. Dementia: a clinical approach. Boston: Butterworth-Heinemann, 1992.
2. Moore DP. Textbook of clinical neuropsychiatry. New York: Oxford University Press, 2001.

Question(s) 373:
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:
Lockwood AH. Toxic and metabolic encephalopathies. In: Bradley WG, Daroff RB, Fenichel GM, et al. Neurology in clinical practice. 2nd ed. Boston: Butterworth-Heinemann, 1996.

Question(s) 374:
Pharmacology/Chemistry

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

Reference:
Windebank A. Metal neuropathy. In: Dyck PJ, Thomas PK, editors. Peripheral neuropathy. 3rd ed. Philadelphia: WB Saunders, 1993.

Question(s) 375 - 377: Clinical Pediatrics

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

Reference:
Menkes JH, Sarnat HB, editors. Child neurology. 6th ed. Philadelphia: Lippincott, Williams & Wilkins, 2000.

Question(s) 378:
Pharmacology/Chemistry

Discussion:
Methanol is metabolized to formaldehyde and formic acid by alcohol dehydrogenase. Formic acid is believed to be the cause of retinal toxicity that mainly affects the ganglion cells. Patients usually have metabolic acidosis, but Kussmaul respiration is uncommon because of the respiratory depression caused by the intoxication. Fomepizole is an inhibitor of alcohol dehydrogenase that appears to be safe and effective in managing methanol poisoning.

Reference:
Brent J, McMartin K, Phillips S, et al. Fomepizole for the treatment of methanol poisoning. NEJM 2001;344:424-429.

Question(s) 379: Clinical Pediatrics

Discussion:
Infantile neuroaxonal dystrophy is an inherited neurodegenerative disorder characterized by psychomotor regression, usually beginning in infancy or early childhood, with progression to dementia, blindness (associated with optic atrophy) and tetraplegia. Axonal neuropathy is characteristic, as is the presence of axonal spheroids in peripheral nerve and brain. MRI shows cerebellar atrophy, with signal hypointensity in the pallida and nigra.

Reference:
Nardocci N, Zorzi G, Farina L. Infantile neuroaxonal dystrophy: clinical spectrum and diagnostic criteria. Neurology 1999;52:1472-1482

Question(s) 380: Clinical Adult

Discussion:
Primary central nervous system lymphoma in AIDS patients is highly associated with positive PCR for EBV-DNA in the CSF. Progressive Multifocal Leukoencephalopathy (PML) is associated with positive PCR for JC virus DNA in the CSF. Thallium SPECT scan is positive in CNS Lymphoma. Increased 14-3-3 protein in the CSF is a feature of Creutzfeldt-Jakob disease. PML is visible on CT scans and MRIs as a non-enhancing brain lesion with no mass effect.

Reference:
Belman AL, Preston T, Milazzo M. Human immunodeficiency virus and acquired immunodeficiency syndrome. In: Goetz GG, Pappert EJ, editors. Textbook of clinical neurology. Philadelphia: WB Saunders Company, 1999.

Question(s) 381: Behavioral

Discussion:
The history is suggestive of early Alzheimer's disease. The MMSE is not a very sensitive test for making a diagnosis of probable Alzheimer's disease in highly intelligent individuals. Patients should be started early on cholinesterase inhibitors and vitamin E.

Reference:
Hake AM. The treatment of Alzheimer's disease: the approach from a clinical specialist in the trenches. In: Pascuzzi RM, Roos KL. Therapy in neurology expert clinicians' views. Seminars in Neurology 2002;22:71-74.

Question(s) 382: Pathology

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

Reference:
Ellison D, Love S. Neuropathology: a reference text of CNS pathology. Chicago: CV Mosby, Inc., 1998.

Question(s) 383: Neuroimaging

Discussion:
The tumor is located in the region of the sella turcica, ruling out glioblastoma, ependymoma and schwannoma. Meningiomas seldom are this symmetrical.

Reference:
Stark DD, Bradley WG. Magnetic resonance imaging. St. Louis: C.V. Mosby, 1988.

Question(s) 384: Neuroimaging

Discussion:
Bony erosion and opacification of the mastoid air cells is seen on the right. The erosion is adjacent to the proximal aspect of the styloid process, which marks the stylomastoid foramen, exit for cranial nerve VII from the skull base. Thus, CN VII is the most likely involved. Erosion is also immediately adjacent to the jugular foramen, which carries CN IX, X, and XI. The hypoglossal canal, which carries CN XII is located more medial to the erosion and much less likely involved. CN III, IV and VI pass through the cavernous sinus and are not near the imaged area.

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 385: Neuroimaging

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

Reference:
Zimmerman RA. Medical imaging of pediatric supratentorial tumors. Seminars in roentgenology 1990;2;225-248.

Question(s) 386: Neuroimaging

Discussion:
The MRI demonstrates posterior frontal and superior temporal atrophy. The anterior frontal lobes are without significant atrophy. The medial temporal temporal lobes are intact, a common area for atrophy in Alzheimer's dementia. There is no evidence of vascular insults or normal pressure hydrocephalus. The personality change is also more indicative of frontal temporal dementia than Alzheimer's.

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 387: Pathology

Discussion:
The photo shows a well-demarcated mass at the cerebellopontine angle. The most common tumor at this site is a schwannoma of the vestibular branch of the 8th cranial nerve.

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 388: Physiology

Discussion:
14 and 6 positive spikes are sharply contoured, and occur in the posterior head regions during light sleep. They are best demonstrated on referential EEG montages, and are most common in adolescent patients.

Reference:
Klass DW, Westmoreland BF. Electroencephalography: general principles and adult electroencephalograms. In: Daube JR. Clinical neurophysiology. Philadelphia: FA Davis Company, 1996.

Question(s) 389: Neuroimaging

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

Reference:
1. Bakshi R, Lindsay BD, Kinkel PR. Brain magnetic resonance imaging in clinical neurology. In: Joynt RJ, Griggs RC, editors. Clinical neurology. Philadelphia: Lippincott-Raven, 1997.
2. DeHaan J, Grossman RI, Civitello L, et al. High field MRI of Wilson's disease. J Comput Tomogr 1987;11:132-135.

Question(s) 390: Neuroimaging

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

Reference:
1. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.
2. Bakshi R, Glass J, Louis DN, et al. Magnetic resonance imaging features of solitary inflammatory brain masses. Journal of Neuroimaging 1998;8:8-14.

Question(s) 391: Neuroimaging

Discussion:
CT is consistent with subacute subdural hematoma with a significant mass effect upon the ventricles. Surgical intervention is appropriate.

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 392: Neuroimaging

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

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 393: Neuroimaging

Discussion:

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

Reference:

1. Czervionke LF, Daniels DL, Meyer GA, et al. Neuroepithelial cysts of the lateral ventricles: MR appearance. AJNR 1987;8:609-613.

2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 394: Physiology

Discussion:

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

Reference:

Kimura J. Electrodiagnosis in disease of nerve and muscle. 3rd ed. New York: Oxford University Press, 2001.

Question(s) 395: Neuroimaging

Discussion:

Surgical treatment alone for an AVM this large likely carries an inappropriate degree of morbidity. An intravascular embolization procedure could substantially reduce the blood flow within the AVM, so that the other interventions such as surgical excision or radiation therapy might be used more effectively. The malformation extends into both temporal lobes. Right carotid artery ligation will not eliminate filling from the other hemisphere, or the posterior circulation (both present in this case). Surgical therapy followed by radiation fails for the same reason mentioned above. Prior to surgery or embolization, the amytal test can help evaluate the possible outcomes.

Reference:

Rauch RA, Vinuela F, Dion J, et al. Preembolization functional evaluation in brain arteriovenous malformations: the superselective Amytal test. AJNR 1992;13:303-308.

Question(s) 396: Neuroimaging

Discussion:
Of the options mentioned, to have the patient breath an oxygen-rich mixture, were the headache to return, is the most reasonable one. This patient had his first episode of cluster headache, when an incidental congenital arachnoid cyst of the right middle fossa was found on MRI. The cyst does not create mass effect and therefore neither decompression nor drainage is warranted. Angiography might be a good idea to rule out an aneurysm causing a subarachnoid hemorrhage, but a lumbar puncture should be done before this procedure. The absence of mass effect precludes risk with an LP. It was specifically mentioned that the relevant findings are shown and therefore no aneurysm was seen on MRI. There is nothing in the history to suggest an internal retinal artery occlusion. The right middle cerebral artery is not occluded, but, lifted by the cyst, fades into the higher section.

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 397: Neuroimaging

Discussion:
The enhancing, nodular, patchy, diffuse abnormalities shown are most characteristic of neurosarcoidosis. Toxoplasmosis produces granulomas, which may enhance, but these are rarely restricted to the gray matter as in this case. Cerebrotendinous xanthomatosis is a white matter disease. Hemorrhagic encephalopathy typically shows areas of both low and high signal intensity, and is not uniformly enhancing. The patient's age and race are also suggestive of neurosarcoidosis.

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 398: Neuroimaging

Discussion:
CT shows multiple parenchymal calcifications with mild atrophy; this is characteristic of a host of infections including toxoplasmosis, rubella, cytomegalovirus, and herpes. Calcification seen in Sturge-Weber syndrome is typically cortical and gyriform. Although lipomas may calcify, they are extra-axial, usually single, and are hypodense on CT. The scan does not show malformations of the sulci or ventricles to suggest schizencephaly or colpocephaly.

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 399: Neuroimaging

Discussion:
Note the hyperdense pineal. Normally, the pineal is not calcified at age 2. The earliest pineal calcifications occur on CT around 6-1/2 years of age. The patient has calcifications in both orbits that indicate congenital retinoblastoma. The calcification in the pineal region indicates that a third tumor, a pineoblastoma is developing.

Reference:
Zimmerman RA, Bilaniuk LT. Age related incidence of pineal calcification detected by CT. Radiology 1982;142:659-662.

Question(s) 400: Pathology

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

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 401: Neuroimaging

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

Reference:
1. Gomori JM, Grossman RI, Goldberg HI, et al. Intracranial hematomas: imaging by high field MR. Radiology 1985;157:87-90.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 402: Neuroimaging

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

Reference:
Bakshi R, Mechtler LL, Kamran S, et al. MRI findings in lumbar puncture headache syndrome: abnormal dural-meningeal and dural venous sinus enhancement. Clinical Imaging 1999;23:73-76.

Question(s) 403: Neuroimaging

Discussion:
The correct answer is epidural metastasis. The bone and especially inner table demonstrates bony irregularity and infiltration of tumor into epidural space. Homogenous enhancement signifies disruption of inner and outer table with infiltration of tumor (metastatic prostate cancer) into extradural space. Epidural hematoma, focal pachymeningitis, and hygroma with hemorrhage into it would present with a smooth medial surface.

Reference:
Woodruff WW. Fundamentals of neuroimaging. Philadelphia: W.B. Saunders Company, 1993.

Question(s) 404: Physiology

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

Reference:
Shaul N. The fundamental neural mechanisms of electroencephalography. Electroencephalogr Clin Neurophysiol 1998;106:101-107.

Question(s) 405: Pathology

Discussion:
The photomicrograph shows a trichinella organism acquired by ingestion of undercooked pork.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 406: Neuroimaging

Discussion:
The correct answer is a Chiari malformation with syringomyelia, both of which are clearly visible on the image and often associated in the same patient. Hydrocephalus is also frequently present with a Chiari malformation (small posterior fossa, with the cerebellar tonsils descended across the foramen magnum), so that aqueductal atresia need not be postulated to explain the enlarged lateral ventricles. The aqueduct is not seen in this image slightly off midline. Neither of the other two possible answers is present on the MRI.

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 407: Neuroimaging

Discussion:
Multiple basal ganglia high signal foci are seen bilaterally. They are more numerous than normal Virchow-Robin spaces. Toxoplasmosis resides also in basal ganglia, but then there is usually also mass effect. Cryptococcus enters the intracranial space through the perivascular spaces and the organisms are surrounded by gelatinous material that is seen as high signal foci. This correlates with "soap-bubble" abscesses seen grossly. The correct answer is cryptococcus infection.

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 408: Neuroimaging

Discussion:
The MRA demonstrates no flow in the basilar artery. Good flow is noted in the vertebral arteries bilaterally. Both of the anterior cerebral arteries originate off the left anterior circulation. The internal and common carotid arteries demonstrate good flow without a dissection or stenoses.

Reference:
1. Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.
2. Osborne A. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 409: Pathology

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

Reference:
1. Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.
2. Duckett S. Pediatric neuropathology. Baltimore: Williams and Wilkins, 1995.

Question(s) 410: Neuroimaging

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

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 411: Pathology

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

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 412: Pathology

Discussion:
Methotrexate neurotoxicity is associated with either high dosage or intrathecal administration, particularly in conjunction with irradiation. It may cause multiple discrete or confluent foci of necrosis in the cerebral or spinal white matter. The lesions frequently are periventricular in location.

Reference:
Ellison D, Love S. Neuropathology: a reference text of CNS pathology. Chicago: CV Mosby, Inc., 1998.

Question(s) 413: Neuroimaging

Discussion:
Tethered cord syndrome often presents with back pain and imaging studies usually demonstate a thickened filum terminale, widening of the spinal canal, posterior cord lipoma, and a low lying spinal cord. The axial MRI demonstrates good cord caliber at L1-L2 where you should be seeing filum terminale only. The lipoma can be seen in the proton density sagittal views.

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 414: Pathology

Discussion:
The myelin-stained axial section of the brain stem at the level of the inferior olivary nuclei shows infarction of a wedge-shaped area of the dorsolateral portion of the medulla in the distribution of the posterior inferior cerebellar artery. This usually results from occlusion of the vertebral artery.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 415: Pathology

Discussion:
The photomicrograph shows numerous Rosenthal fibers clustered most densely around blood vessels. They are a characteristic feature of Alexander's disease.

Reference:
Duckett S. Pediatric neuropathology. Baltimore: Williams & Wilkins. 1995.

Question(s) 416: Pathology

Discussion:
Formation of onion bulbs results from repeated episodes of demyelination and remyelination. Onion bulbs are especially conspicuous in hypertrophic Charcot-Marie-Tooth disease, Dejerine-Sottas disease and Refsum disease. In addition, approximately one half of the cases of chronic inflammatory demyelinative polyneuropathy show substantial numbers of onion bulbs.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 417: Neuroimaging

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

Reference:
Barkovich AJ. Pediatric neuroimaging. 2nd ed. New York: Raven Press, 1994.

Question(s) 418: Pathology

Discussion:
The photomicrograph shows acutely branching septate hyphae which most likely represent an opportunistic Aspergillosis infection. The organism initially infects the lung and then spreads hematogenously to involve other organs. The brain is second only to the lung as a site of visceral involvement, and frequently manifests as hemorrhagic infarctions, as shown.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 419: Neuroimaging

Discussion:
The scan shows typical findings of Dandy-Walker malformation, with absent vermis, large posterior fossa cyst and absent posterior corpus callosum.

Reference:
Barkovich AJ, Maroldo TV. Magnetic resonance imaging of normal and abnormal brain development. Top Magn Reson Imaging 1993:5;96-122.

Question(s) 420: Neuroimaging

Discussion:
The axial T2-weighted image shows large cerebrospinal fluid collection expanding the posterior fossa with absence of the inferior vermis and hypoplasia of the two cerebellar hemispheres. In an arachnoid cyst, the cerebellar hemispheres should be relatively well preserved as to size and the inferior vermis would not be absent. Cystic astrocytoma is not a consideration as the fluid is extra-axial. A giant cisterna magna would not be associated with both vermian and bilateral cerebellar hypoplasia. Schizencephaly (a pial ependymal cleft seen in the supratentorial space) is not present.

Reference:
1. Barkovich AJ, Kjos BO, Norman D, et al. Revised classification of posterior fossa cysts and cyst-like malformations based on the results of multiplanar MR imaging. AJNR 1989;10:977.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 421: Pathology

Discussion:
Degeneration restricted to the dorsal columns is characteristic of tabes dorsalis.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 422: Neuroimaging

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

Reference:
1. Barkovich AJ. Pediatric neuroimaging. New York: Raven Press, 1990.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 423: Pathology

Discussion:
Colloid cyst of the third ventricle is a round circumscribed grape-like lesion occurring in the third ventricle where it may produce position-dependent CSF obstruction due to ball-valving. Acute lethal hydrocephalus may occur leading to sudden death.

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 424: Pathology

Discussion:
In this picture, the collections of multiple bubbly cysts in the white matter, caudate and putamen are typical of parenchymal involvement by cryptococcal meningitis. Aspergillosis produces gray irregular necrotic masses. Tuberculoma and metastases tend to be discrete solid firm masses.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 425: Neuroimaging

Discussion:
The patient presented here has multiple neurofibromas of spinal nerves, and a plexiform neurofibroma infiltrating the nerves forming the left brachial plexus. Although multiple spinal tumors can be seen in both NF-1 (pathology is neurofibroma) and in NF-2 (pathology is schwannoma), plexiform neurofibromas are not seen in NF-2. The remaining diagnoses are not consistent with the images.

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 426: Pathology

Discussion:
The graphic shows a circumscribed collection of purulent material (empyema) on the surface of the brain (i.e., subdural as opposed to epidural or subarachnoid). Subdural empyema may complicate head injury or sinus infections.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 427: Anatomy

Discussion:
The arrow is pointing to the head of the caudate nucleus. Lesions of this structure are associated with chorea. A lesion of the subthalamic nucleus leads to hemiballismus. Lesions of the substantia nigra result in parkinsonism. Hemiparesis results from interruption of the upper motor neurons anywhere in their path above the pons.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 428: Pathology

Discussion:
The atrophic cerebellar vermis and loss of both Purkinje and granule cell neurons indicate a degeneration of neurons seen in alcoholic cerebellar degeneration. No tumor or volume-expanding lesion is present. Dandy Walker causes agenesis of the cerebellar vermis with formation of a cyst. Methanol intoxication affects deep white matter and putamen, not cerebellar vermis.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 429: Neuroimaging

Discussion:

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

Reference:

1. Ham JS, Huss RG, Benson JE, et al. MRI imaging of the skull base. J Comput Assist Tomogr 1984;8:944-952.

2. McGinnis BD, Brady TJ, New PF, et al. MR imaging of tumors of the posterior fossa. J Comput Assist Tomogr 1984;7:575-584.

Question(s) 430: Neuroimaging

Discussion:

Both the location of the lesion and the presence of precocious puberty favor a hamartoma in the tuber cinereum.

Reference:

1. Williams AL, Mosby VM. Cranial computed tomography: a comprehensive text. St. Louis: C.V. Mosby, 1985.

2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

3. Osborne AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 431: Neuroimaging

Discussion:

There is a hypodense, CSF density, collection that lies between the two leaves of the septum pellucidum, separating the lateral ventricles to each side of the midline. The finding is consistent with a cavum septum pellucidum and cavum vergae. A colloid cyst would occur only in the region of the foramen of Monro and be slightly to markedly hyperdense; a craniopharyngioma would not lie between the leaves of the septum pellucidum, but would displace the ventricle around the mass of the craniopharyngioma, and would likely have calcification in its wall. An intraventricular meningioma would lie adjacent to the choroid plexus of the lateral ventricle or that within the third ventricle, would be hyperdense in a significant portion of its mass. An arachnoid cyst would lie external to the ventricular system.

Reference:

1. Rao K, Harwood-Nash D. Craniocerebral anomalies. In: Lee SH, Rao K, editors. Cranial computed tomography and MRI. New York: McGraw-Hill, 1987.

2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 432: Neuroimaging

Discussion:
The MRIs show mass in the prepontine space indenting the pons. The signal intensity characteristics are those of fat, the finding being most consistent with a lipoma. The bright line posterior to the mass is due to chemical shift artifact, which is usually best seen on conventional spin-echo T2-weighted image.

Reference:
1. Truwit CL, Barkovich AJ. Pathogenesis of intracranial lipoma: an MR study in 42 patients. AJNR 1990;11:665.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 433: Neuroimaging

Discussion:
The extensive enhancement of the ependyma is most suggestive of ventriculitis. Choroid plexus papilloma with CSF spread, ependymoma, transependymal flow of CSF, and tuberous sclerosis with candle guttering do not produce this pattern, with the possible exception of transependymal CSF flow (which is non-enhancing). Enhancement of the choroid plexus is normal and does not suggest a papilloma. Candle guttering has the appearance of enhancing, strings or beads that tend to protrude into the ventricles.

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 434: Neuroimaging

Discussion:
MR is effective in demonstrating a displaced optic chiasm because the optic chiasm is usually slightly more intense than pituitary tumors.

Reference:
1. Daniels DL, Haughton VM, Czervionke LF. MR of the skull base. In: Bradley WG, Stark D. Magnetic resonance imaging. St. Louis: CV Mosby, Inc., 1988.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 435: Anatomy

Discussion:
The structure indicated by the arrow is the lateral geniculate body.

Reference:
Haines DE. Fundamental neuroscience. 2nd ed. New York: W.B. Saunders, 2002.

Question(s) 436: Pathology

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

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 437: Neuroimaging

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

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 438: Pathology

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

Reference:
Duckett S. Pediatric neuropathology. Baltimore: Williams & Wilkins, 1995.

Question(s) 439: Neuroimaging

Discussion:
The diffusion and ADC images give clear evidence of an acute stroke. The hyperintensity on the diffusion with corresponding hypointensity on the ADC map are shown. These findings would not be present on the other choices.

Reference:
Bakshi R, Ketonen L. Brain MRI in clinical neurology. In: Joynt RJ, Griggs RC, editors. Baker's clinical neurology. Philadelphia: Lippincott, Williams & Wilkins, 2001.

Question(s) 440: Neuroimaging

Discussion:
Predicting the chances of a good outcome is notoriously inaccurate, but duration of dementia is one of the more important considerations. Depressive symptoms may improve with shunting, but this may occur whether or not his dementia syndrome improves. If his shunt results in a decrease in ventricular size, that is indeed favorable, but the duration of dementia is a more critical variable. Persons with chronic NPH may not respond to the shunt, even if follow-up CT scans suggest that the hydrocephalus is improved. The duration of urinary incontinence reflects the degree of damage to the fibers of the medial frontal lobe micturition centers. The bilateral Babinski signs, severe gait abnormality, and spasticity all suggest the same type of damage. The duration of incontinence has little predictive value. Finally, subcortical types of dementia are more likely to be treatable than cortical varieties. This by itself is no guarantee that the shunt will produce clinical improvement in the dementia syndrome.

Reference:
Cummings JL, Benson DF. Dementia: a clinical approach. Boston: Butterworth-Heinemann, 1992.

Question(s) 441: Pathology

Discussion:

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

Reference:

Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 442: Neuroimaging

Discussion:

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

Reference:

1. Atlas S. Magnetic resonance imaging of the brain and spine. New York: Raven Press, 1991.
2. Bakshi R, Lindsay BD, Kinkel PR. Brain magnetic resonance imaging in clinical neurology. In: Joynt RJ, Griggs RC, editors. Clinical neurology. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 443: Pathology

Discussion:

The biopsy of this cystic mass showed layers of flattened, anucleate squames. This type of “flaky” keratin is characteristic of epidermoid and dermoid cysts. In contrast, the keratin formed by adamantinomatous craniopharyngiomas typically consists of nodular clusters of very plump keratinocytes referred to as “wet” keratin. The lining of Rathke cleft cysts, colloid cysts, and neurenteric cysts consists of ciliated pseudostratified columnar epithelium with scattered goblet cells.

Reference:

Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 444: Pathology

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

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 445: Pathology

Discussion:
The photo shows a poorly circumscribed variegated mass with foci of necrosis and hemorrhage most consistent with glioblastoma multiforme. The lack of central purulent material is against an abscess. The lesion is not confined within a single vascular territory, nor is it hemorrhagic, arguing against embolic infarct. The intra-axial location argues against most meningiomas. The lesion is a mass, ruling out Huntington's disease.

Reference:
Burger PC, Scheithauer BW, Vogel FS. Surgical pathology of the nervous system and its coverings. 4th ed. New York: Churchill Livingston, 2002.

Question(s) 446: Neuroimaging

Discussion:
Combining the history and the image, a Tolosa-Hunt syndrome is the most likely diagnosis. A meningioma "en plaque" would not have caused a similar syndrome two years previously in the contralateral eye leaving no trace. Myasthenia and the Miller-Fisher syndrome do not show on MRI the enhancing lesion in right cavernous sinus and meninges present in this case. A pseudotumor of the orbit sometime extends into the retro-orbital meninges, but in this case there is no mass effect in the intraconal fat and the process clearly extends much beyond the orbit.

Reference:
Orrison WW. Neuroimaging. Philadelphia: WB Saunders, 2000.

Question(s) 447: Neuroimaging

Discussion:
The sagittal T1-weighted image shows a small pons and cerebellar atrophy. The axial T2-weighted image through the pons shows the pons to be reduced in size. The axial image through the medulla and lower cerebellum show that the cerebellum is of high signal intensity relative to the more posteriorly placed occipital lobes, and that the medullary olives are not normal in size. Thus the findings are that of cerebellar, pontine and olivary atrophy. There is no intrinsic high signal intensity lesion within the substance of the pons so that an infarct is not present. The hallmark of olivopontocerebellar atrophy is loss of the belly of pons, which is also clearly affected. Friedreich's ataxia is usually primarily seen with spinal atrophy.

Reference:
1. Nabatame H, Fukuyama H, Akiguchi I, et al. Spinocerebellar degeneration: qualitative and quantitative MR analysis of atrophy. JCAT 1988;12:298.
2. Bakshi R, Lindsay BD, Kinkel PR. Brain magnetic resonance imaging in clinical neurology. In: Joynt RJ, Griggs RC, editors. Clinical neurology. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 448: Pathology

Discussion:
The graphic shows severe pontine atrophy and would be most likely seen in olivopontocerebellar atrophy (OPCA). OPCA is part of multiple system atrophy. Glia (especially oligodendroglia) in multiple system atrophy exhibit cytoplasmic, flame-shaped, silver-positive inclusions. The inclusions are alpha-synuclein immunoreactive.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 449: Neuroimaging

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

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 450: Neuroimaging

Discussion:
The sagittal MR images post-gadolinium injection show multiple round enhancing lesions that are on the surface of the cervical spinal cord. Elongated irregular plaques of tumor enhance along the dorsal aspect of the upper thoracic cord. The findings are consistent with tumor seeding. Post-operative changes are present in the posterior fossa. Astrocytoma of the cervical cord would expand the cord and enhance more homogeneously throughout. Syringohydromyelia would be a cavity within the cord that expands it. Cord contusions would have high signal intensity within the substance of the cord. Arachnoiditis can enhance, but does not have a nodular appearance as in this case.

Reference:
1. Sze G, Abramson A, Krol G, et al. Gadolinium-DTPA in the evaluation of intradural extramedullary spinal disease. AJNR 1988;9:153.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 451: Neuroimaging

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

Reference:
Offenbacher H, Fazekas F, Schmidt R, et al. Superficial siderosis of the central nervous system: MRI findings and clinical significance. Neuroradiology 1996;38:S51-S56.

Question(s) 452: Pathology

Discussion:
Centronuclear myopathy is characterized by the presence of centrally located nuclei, often accompanied by a perinuclear vacuole. The muscle usually shows type I myofiber predominance.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 453: Anatomy

Discussion:
The arrow is pointing to the substantia nigra which normally functions to faciliatate voluntary motor activity originating in the prefrontal and motor cortex ipsilateral to the substantia nigra. Damage to one substantia nigra results in hemiparkinsonism. Since the upper motor neurons of the motor strip are the motor output elements and these cross at the medullary decussation, the hemiparkinsonism resulting from a unilateral substantia nigra lesion is contralateral to the lesion.

Reference:
Benarroch EE, Westmoreland BF, Daube JR, et al. Medical neurosciences - an approach to anatomy, pathology and physiology by systems and levels. 4th ed. New York: Lippincott, Williams & Wilkins, 1999.

Question(s) 454: Anatomy

Discussion:
The photomicrograph shows a coronal section of the corpus callosum and cingulate gyri. The arrows identify the indusium griseum. The indusium (also called the supracallosal gyrus) is a direct continuation of the hippocampal formation located dorsal to the corpus callosum.

Reference:
Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 455: Neuroimaging

Discussion:
The correct response is thrombosed aneurysm. Arachnoid cyst would be filled with cerebrospinal fluid-like signal on all sequences. Hamartomas of tuber cinereum are isointense with grey matter on T1 sequences and hyperintense on T2. A lipoma would also show hyperintensity on T1 and could occur in this location, however, they are usually associated with chemical shift artifact on conventional spin-echo T2. This thrombosed aneurysm contains blood breakdown products in the early subacute stage.

Reference:
Bakshi R, Lindsay BD, Kinkel PR. Brain magnetic resonance imaging in clinical neurology. In: Joynt RJ, Griggs RC, editors. Clinical neurology. Philadelphia: Lippincott, Williams & Wilkins, 1998.

Question(s) 456: Neuroimaging

Discussion:
The contrast enhanced axial CT demonstrates ill-defined ring enhancement with significant edema. Findings are typical for malignant brain neoplasm. The axial proton density MR image demonstrates an ill-defined mixed signal lesion in the right posterior parietal region. Intratumoral hyperintense necrotic foci are seen in addition to hyperintense edema. Mass effect upon the right lateral ventricle is seen. Some tubular signal void regions are seen, they are most likely consistent with vascular structures. The ring enhancement in brain abscess is fairly uniform in thickness and the ring enhancement appears to be under tension. Herpes encephalitis usually originates in temporal lobes. Major brain infarction usually also involves the gray matter, which is spared in this patient.

Reference:
1. Kelly WM, Brant Zawadski M. Magnetic resonance imaging and computed tomography of supratentorial tumors. In: Radiology: diagnosis-imaging intervention. Philadelphia: J.B. Lippincott Co., 1986.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 457: Pathology

Discussion:
Herniation of the cerebellar vermis and medulla, with kinking of the upper cervical cord and beaking of the quadrigeminal plate are components of the Arnold-Chiari malformation.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 458: Neuroimaging

Discussion:
MR angiogram demonstrating good flow in all major vessels. An anatomical variant of both anterior cerebral arteries originating from the left internal carotid artery is shown. In addition, the right posterior cerebral artery originates from the anterior circulation (fetal origin). There are no aneurysms or stenoses evident.

Reference:
Osborn AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 459: Pathology

Discussion:
The illustration shows an eosinophilic intracytoplasmic inclusion in a cortical neuron. This is a Negri body and is diagnostic of rabies encephalitis.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 460: Neuroimaging

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

Reference:
1. Geyer CA, Sartor KJ, Prensky AJ, et al. Leigh's disease: CT & MR in five cases. J Comput Assist Tomogr 1988;12:40-44.
2. Osborne AG. Diagnostic neuroradiology. St. Louis: CV Mosby, Inc., 1994.

Question(s) 461: Neuroimaging

Discussion:
Ependymoma is the most likely diagnosis, confirmed here by surgery. The tumor arises from the spinal canal and molds the vertebral bodies. A chordoma would arise from the vertebral bodies, or more likely, from the sacrum, and compress the spinal canal. Although at the sacral level it may be difficult to appreciate the origin of the tumor, it clearly appears intramedullary in higher sections. Similar arguments could be made against the diagnosis of osteochondroma. The appearance of intramedullary coccidiomycosis is different and in general, infections tend to affect the intervertebral discs, relatively spared by this tumor.

Reference:
Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 462: Pathology

Discussion:
The picture shows marked caudate atrophy diagnostic of Huntington disease, which results from a trinucleotide repeat amplification mutation in the huntingtin gene residing on chromosome 4.

Reference:
Graham DI, Lantos PL. Greenfield's neuropathology. 7th ed. New York: Arnold Press, 2002.

Question(s) 463: Neuroimaging

Discussion:
Noncontrast CT reveals a nearly isointense extra-axial mass in left parietal area. The enhancement is homogeneous and intense consistent with meningioma.

Reference:
1. Yock DH. Imaging of CNS disease: a CT and MR teaching file. Baltimore: Mosby, 1991.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 464: Neuroimaging

Discussion:
Common carotid artery injection (arterial phase, lateral view) reveals a flame-shaped "stump" of internal carotid artery consistent with dissection. The external carotid artery fills with contrast. The internal carotid artery (ICA) is relatively unprotected in its cervical course and therefore vulnerable to trauma. Trauma to ICA may result in thrombosis, intimal tears, stenosis, arteriovenous fistulas, pseudoaneurysms or dissection.

Reference:
1. Stringer WL, Kelly DL Jr. Traumatic dissection of the extracranial internal carotid artery. Neurosurg 1980;6:123-130.
2. Greenberg JO. Neuroimaging: a companion to Adams and Victor's principles of neurology. New York: McGraw-Hill, Co., 1999.

Question(s) 465 - 469: Neuroimaging

Discussion:
MR midline sagittal images show signal void due to flowing blood in the internal cerebral vein. Thin soft tissue forming the roof of the cranial aspect of the fourth ventricle represents the superior medullary velum.

Reference:
Daniels DL, Haughton VM, Naidich T. Cranial and spinal magnetic resonance imaging: atlas and guide. New York: Raven Press, 1987.

Question(s) 470 - 474: Neuroimaging

Discussion:
Axial T2-weighted image through suprasellar cistern. Normal anatomy.

Reference:
Hanaway J, Woolsey TA, Gado MH, et al. The brain atlas. Baltimore: Fitzgerald Science Press, 1998.

Question(s) 475 - 479: Neuroimaging

Discussion:
Straightforward identification of cervical and cerebral vessels.

Reference:
1. Netter FH. The CIBA collection of medical illustrations: nervous system. Volume 1, Part 1. New York: Donnelley & Sons Company, 1986.
2. Parent A. Carpenter's human neuroanatomy. 9th ed. Baltimore: Williams & Wilkins, 1996.

Question(s) 480: Anatomy

Discussion:
The visual field defect is a quadrantic sectoranopia and localization is the lateral geniculate nucleus of the thalamus. The anterior choroidal artery is the main artery to the lateral geniculate.

Reference:
1. Parent A. Carpenter's human neuroanatomy. Baltimore: Williams & Wilkins, 1996.
2. Brazis PW, Masdeu JC, Biller J. Localization in clinical neurology. Boston: Little, Brown & Co., 1996.

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