Knowledge Classification Sample Questions

Questions on the RITE are now also designated as Core Knowledge (more than 90% of residents should have encountered the clinical scenario described in the question), Intermediate Knowledge (between 50% and 90% of residents should have encountered the scenario described in the question), and Subspecialty Knowledge (50% of residents or more will NOT have the scenario described in the question). Total percent of questions designated Core and Subspecialty Knowledge are reported here but not by specific content area. Note these designations are not included in the ABPN Certification Examination in Neurology; rather, they were developed by the RITE Panel as a means of providing additional information about progress throughout residency. These sample questions appeared on the 2021 RITE and will not appear on future administrations of the RITE.

Item Number: 1 **Knowledge Classification:** Core

Question: A neurologist reviews a study to determine if patients taking interferon beta for MS have an increased likelihood of relapse due the presence of neutralizing antibodies. Patients are enrolled separately in two groups: those who had relapses since starting therapy and those who did not. Both groups are tested for the presence of neutralizing antibodies at study entry. The frequency and titers of neutralizing antibodies were found to be higher in the patients who had a relapse. Which of the following best describes the design of this study?

A. case control
B. cross-sectional
C. double-blinded placebo-controlled
D. prospective cohort
E. retrospective cohort

Rationale: The key feature of a case-control study is that participants are ascertained based on outcome, in this case the presence of MS relapses. All patients in this study have MS and are taking interferon beta: patients in group 1 have had relapses since starting therapy and those in group 2 have not. The direction of inquiry of the research question is from outcome to exposure. The “exposure” here is the presence of neutralizing antibodies. The investigators inquired about the frequency of neutralizing antibodies among the cases (patients with relapses) and the controls (patients without relapses). In a cross-sectional study, all members of the sample are observed at a single point in time.

Item Number: 3 **Knowledge Classification:** Core

Question: A 15-year-old boy presents with fever, lethargy, and a generalized clonic seizure. CSF analysis shows 35 white blood cells/mm3 (all lymphocytes), protein 48 mg/100 mL, and glucose of 65 mg/100 mL. EEG abnormalities are shown. Which of the following IV medications is the most appropriate management?

A. acyclovir
B. amphoterecin
C. ceftriaxone
D. dexamethasone
E. phenytoin

Rationale: The EEG shows periodic lateralizing epileptiform discharges in the left temporal leads. The clinical presentation, EEG findings, and CSF abnormalities are typical of herpes simplex encephalitis. The treatment of choice is IV acyclovir.

Item Number: 6 **Knowledge Classification:** Intermediate

Question: A 34-year-old woman who has had frequent headaches over the past year recently developed bifacial weakness, diplopia, worsening headache, stiff neck, and mild encephalopathy. MRI scan with contrast reveals basilar leptomeningeal enhancement. Which of the following additional abnormal findings is most likely to be seen?

A. bilateral hilar adenopathy on chest radiograph
B. cardiac thrombus on echocardiogram
C. positive CSF JC virus DNA
D. positive serum HIV antibody
E. positive CSF PCR for herpes simplex virus

Rationale: Bilateral hilar adenopathy, bilateral cranial nerve palsies, and encephalopathy due to a basilar meningoencephalitis are typical of neurosarcoid. Progressive multifocal leukoencephalopathy due to the JC virus, herpes simplex virus encephalitis, and HIV dementia all are characterized by white matter or cortical lesions on MRI. The case is inconsistent with strokes due to cardiac thrombi.

Item Number: 7 **Knowledge Classification:** Subspecialty

Question: A 60-year-old man with restless legs syndrome is initially treated with pramipexole at 10:00 PM daily and has had a good response to the medication for the past year. He then develops symptoms occurring earlier in the evening, and for the past month also during the day. He does not take the medication earlier in the day as he fears it may stop working at night. Which of the following mechanisms best describes his symptoms?

A. augmentation
B. denervation
C. rebound
D. receptor supersensitivity
E. underdosing

Rationale: Augmentation in restless legs syndrome refers to symptoms occurring earlier in the day and sometimes spreading from the legs to the arms. This should be distinguished from rebound by calculating the duration of therapy and the time the medication is taken. In this case, rebound is unlikely given the one-time dosing and half-life of the drug.

Knowledge Classification: Subspecialty

Item Number: 9 **Knowledge Classification:** Subspecialty

Question: A 33-year-old man presents with left-sided facial weakness, hyperacusis, and left periauricular discomfort that has progressed over the past 24 hours. Examination reveals a moderate degree of left lower motor neuron facial weakness. Which of the following interventions is most likely to improve his outcome?

A. acyclovir
B. doxycycline
C. facial nerve decompression
D. physical therapy
E. corticosteroids

Rationale: Early use of corticosteroids has been widely reported to increase the probability of recovery in patients with new-onset Bell palsy. When administered early, the addition of an antiviral agent to a steroid may modestly further increase the probability of recovery. Antivirals alone, however, have no value in this setting. Neither physical therapy modalities nor facial nerve decompression has been shown to be of value for the treatment of acute Bell palsy. In the absence of a tick bite, erythema migrans rash, positive Lyme serology, or other clinical suggestion of Lyme disease, it is not appropriate to prescribe doxycycline for Bell palsy.

Knowledge Classification: Core

Item Number: 10 **Knowledge Classification:** Core

Question: A 32-year-old woman arrives at the emergency department in status epilepticus. She has a history of well-controlled diabetes mellitus for which she takes insulin. Vital signs are as follows: BP 130/85 mm Hg, HR 110 beats/min, RR 15 breaths/min, temperature 98.6°F (37°C). She is mildly cyanotic and having generalized tonic-clonic movements. An oral airway and IV are in place and she is given oxygen by mask at 8 L/min. Administration of which of the following IV medications is the most appropriate next step?

A. acyclovir
B. dextrose
C. mannitol
D. propofol
E. valproic acid

Rationale: In a patient with status epilepticus, hypoglycemia should be immediately considered as an underlying etiology, particularly in a patient with diabetes. If serum glucose is not immediately available or the value is uncertain, 50 mL of IV 50% dextrose should be administered concurrently with 100 mg of IV thiamine.

Item Number: 12 **Knowledge Classification:** Core

Question: The diffusion-weighted images of a 22-year-old man who was found unresponsive show cytotoxic edema in which of the following structures?

F. bilateral hippocampi and globus pallidi
G. bilateral amygdala and lentiform nuclei
H. bilateral unci and putamina
I. bilateral internal capsules and thalami
J. bilateral caudates and putamina

Rationale: The images show sequelae of hypoxic injury, presumably due to drug overdose and/or hypoventilation. Recognizing the involvement of the susceptible gray matter structures is crucial for diagnosis.

Item Number: 16 **Knowledge Classification:** Subspecialty

Question: Alpha-glucosidase alfa enzyme replacement has been approved for the treatment of which of the following conditions?

K. Gaucher disease
L. Krabbe disease
M. Leigh syndrome
N. McArdle disease
O. Pompe disease

Rationale: Alglucosidase alfa is an FDA-approved treatment for late-onset (age >8 years) (noninfantile) Pompe disease, a rare genetic disorder that occurs in an estimated 1 in every 40,000 to 300,000 births. Principal signs and symptoms are cardiac and skeletal muscle weakness that progresses to respiratory weakness and death from respiratory failure. A mutation of the GAA gene prevents the body from making an enzyme, or making enough of the enzyme, necessary for lysosomal degradation of muscle glycogen. Without the enzyme action, glycogen builds up in the lysosomes. Alglucosidase alfa is believed to work by replacing the deficient GAA, thereby reducing the accumulated glycogen in cardiac and skeletal muscle cells. Multiple brands of alglucosidase alfa are available, with some directed at the treatment of infants and children with Pompe disease because younger patients generally have a much more aggressive form of the disease.

Item Number: 18 **Knowledge Classification:** Intermediate

Question: An obese 16-year-old girl presents following a single generalized tonic-clonic seizure upon awakening. Medical history reveals episodes of morning “twitchiness.” She is sexually active and takes no medications. Neurologic examination is normal. EEG shows 4-Hz spike-and-wave discharges. Which of the following medications is the most appropriate management for this patient?

P. carbamazepine
Q. lamotrigine
R. phenytoin
S. topiramate
T. valproic acid

Rationale: This patient most likely has juvenile myoclonic epilepsy (JME), and although she has had only a single seizure, the morning “twitchiness” is most likely myoclonus. With this type of epilepsy, she is at high risk for having further events. Lamotrigine must be started at a very low dose and gradually increased. Risk of allergic reaction is increased with higher starting doses or rapid titration. In some cases, it can increase the myoclonus. Phenytoin and carbamazepine often worsen myoclonus in JME. Valproic acid provides good control but has embryotoxic effects, may increase intellectual disability in offspring, and often contributes to obesity; thus, it is a poor choice in an obese adolescent girl who is sexually active, as no form of contraception is infallible. Topiramate could be considered but is poorly tolerated when started at full dose and has not been labeled by the FDA for JME. Levetiracetam, however, is FDA approved as adjunctive treatment of JME.

Item Number: 19 **Knowledge Classification:** Subspecialty

Question: A right-handed, 70-year-old man presents to the emergency department at noon with global aphasia and right-sided hemiplegia. Medical history includes atrial fibrillation treated with dabigitran. His wife witnessed his symptom onset 20 minutes prior to arrival but is unsure if he took his dabigitran this morning or last night. Which of the following laboratory studies best evaluates the activity of dabigatran in this patient?

A. anti-Xa activity
B. aPTT
C. dilute Russell viper venom time (dRVVT)
D. PT
E. thrombin time

Rationale: Dabigitran is a direct thrombin inhibitor that is FDA approved for reduction of stroke risk in patients with nonvalvular atrial fibrillation and for treatment of deep venous thrombosis and pulmonary embolism in patients who have been treated with a parenteral anticoagulant for 5 to 10 days. Peak levels can increase PT and aPTT, but these studies are not indicative of dabigitran activity at nonpeak levels. Thrombin time is the most sensitive and widely available assay in detecting anticoagulatory activity of dabigatran. A normal thrombin time virtually excludes a clinically relevant anticoagulatory effect of dabigatran in plasma. At the time of peak levels, thrombin time measurements are extremely prolonged or unmeasurable. Dabigitran activity also can be assessed by ecarin clotting time. Ecarin activates prothrombin, but this activation is inhibited in a linear manner by direct thrombin inhibitors such as dabigitran. Rivaroxaban and apixiban are factor Xa inhibitors that are FDA approved for reduction of stroke risk in patients with nonvalvular atrial fibrillation. Anti-factor Xa activity tests correlate with the concentration of factor Xa inhibitors.

Item Number: 21 **Knowledge Classification:** Core

Question: For the past 6 months, a 55-year-old woman has experienced vertigo when she lies on her right side or when she cradles a phone on her right shoulder. She develops severe vertigo after a latency of a few seconds in the right head down position during the Dix-Hallpike maneuver. The vertigo and nystagmus subside within 20 seconds. The remainder of her examination is normal. Which of the following is the most appropriate next step in management?

A. audiometry
B. canalith repositioning maneuver
C. administration of meclizine
D. MRI of the brain
E. vestibulonystagmography (VNG)

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

Item Number: 22 **Knowledge Classification:** Intermediate

Question: A 72-year-old man presents with a 3-year history of progressive difficulty with thinking. He works as a college professor and often becomes confused if he is distracted by students' questions. His wife reports he sees intruders in the house when they are home alone. He scores 21/30 on the Mini-Mental Status Examination. Examination reveals mild rigidity in both arms with passive range of motion and slowing of gait. Which of the following is the most likely diagnosis?

A. Creutzfeldt-Jakob disease
B. dementia of Alzheimer type
C. dementia with Lewy bodies
D. frontotemporal dementia
E. Parkinson disease

Rationale: The history, time course of the progressive dementia, and hallucinations are consistent with Lewy body dementia. Dementia of Alzheimer type typically does not cause vivid hallucinations. Creutzfeldt-Jakob disease is typically more rapid in its tempo of decline. Although the rigidity of limbs and slow gait may be suggestive of Parkinson disease, the combination of these findings with vivid visual hallucinations, fluctuating cognition and attention, and early dementia is more consistent with dementia with Lewy bodies.

Item Number: 23 **Knowledge Classification:** Intermediate

Question: A 36-year-old woman taking venlafaxine and mirtazapine for depression presents with tremulousness, limb jerking, confusion, and tachycardia 4 days after starting tramadol for a recent diagnosis of fibromyalgia. This neurologic condition is being mediated by which of the following mechanisms?

A. decreased transmission of GABA inhibitors
B. downregulation of dopamine D2 receptor
C. increased activation of cholinergic M1 and M3 receptors
D. increased noradrenergic neurotransmission
E. increased activation of serotonin 5-HT1A and 5-HT2A receptors

Rationale: Serotonin syndrome is a potentially life-threatening adverse drug reaction caused by excessive serotonergic agonism of CNS and peripheral nervous system serotonergic receptors. Symptoms are characterized by a triad of features: neuromuscular hyperactivity (tremor, clonus, myoclonus, hyperreflexia, and in advanced stages, pyramidal rigidity); autonomic hyperactivity (diaphoresis, fever, tachycardia and tachypnea); and altered mental status (agitation, excitement, and in advanced stages, confusion). Monoamine oxidase inhibitors and opioid analgesics can cause serotonin toxicity when serotonin neurotransmission is increased at postsynaptic 5-HT1A and 5-HT2A receptors through increased serotonin synthesis, decreased serotonin metabolism, increased serotonin release, inhibition of serotonin reuptake, or direct agonism of the serotonin receptors. Many other medications and supplements have been implicated in this syndrome, including SSRIs, SNRIs, bupropion, monoamine oxidase inhibitors, triptans, opioid analgesics, and lithium. Illicit drugs such as LSD, cocaine, and amphetamines, herbal supplements such as St. John's wort and ginseng, dextromethorphan, ritonavir, linezolid, and some antiemetics such as metoclopramide, ondansetron, and granisetron are also associated with serotonin syndrome. Etiology often includes therapeutic medication use, intentional overdosing of serotonergic agents, or complex interactions between medications that directly or indirectly modulate the serotonin system. In this patient, the etiology is concurrent administration of tramadol, venlafaxine, and mirtazapine. In this patient, the activation of 5-HT1A receptors by mirtazapine, the combined serotonin reuptake inhibition by venlafaxine and tramadol, as well as possible serotonin release by tramadol, were contributing factors. Neuroleptic malignant syndrome (NMS) is caused almost exclusively by antipsychotics, including all types of neuroleptic medicines along with newer antipsychotic drugs. The higher the dose, the more common the occurrence. Rapid and large increases in dose can also trigger this syndrome. Other medications, environmental or psychological factors, hereditary conditions, and specific demographics are associated with greater risk for this syndrome, but to date no conclusive evidence has been found. This disorder typically develops within 2 weeks of initial treatment but may develop at any time the medication is being taken.

Item Number: 26 **Knowledge Classification:** Subspecialty

Question: An ambulatory 5-year-old girl who is intellectually disabled and lacks verbal communication skills presents with a history of generalized epilepsy. Examination reveals microcephaly, an abnormal gait with jerky movements of the limbs, wide-spaced teeth, a wide smile, and an excessively cheerful and social demeanor. Which of the following studies is most likely to reveal the etiology?

A. DNA test for deletion of chromosome 15q11-q13
B. DNA test for deletion of MECP2 gene
C. mitochondrial DNA panel for large deletions
D. MRI of the brain for structural malformations
E. quantitative urine organic acid for elevations of methylmalonic acid

Rationale: The clinical description is of a child with Angelman syndrome, a disorder that results from loss of function of the imprinted UBE3A gene on chromosome 15q11.2-q13. This loss of function can be caused by a mutation on the maternal allele (the most common cause), a 5- to 7-Mb deletion of the maternally inherited chromosomal region, paternal uniparental disomy of chromosome 15, or an imprinting defect. The MECP2 gene is associated with Rett syndrome. The clinical description does not fit with a mitochondrial disorder. Though cortical malformations may be associated with epilepsy, this does not explain the larger syndrome. Methylmalonic aciduria is associated with exacerbations during intercurrent illnesses.

Item Number: 27 **Knowledge Classification:** Subspecialty

Question: An adult patient with an intellectual disability, adenoma sebaceum, and subungual fibromas presents with headache and papilledema. MRI scan shows obstructive hydrocephalus resulting from an intraventricular tumor at the foramen of Monro. Which of the following is the most likely diagnosis?

A. central neurocytoma
B. choroid plexus papilloma
C. colloid cyst
D. ependymoma
E. subependymal giant cell astrocytoma

Rationale: Patients with tuberous sclerosis have a variety of systemic and CNS tumors. In the brain, astrocytic hamartomas called tubers and candle gutterings are present. Subependymal giant cell astrocytomas (SEGA) are true neoplasms arising in the vicinity of the foramen of Monro. These tumors may produce CSF obstruction and rarely undergo malignant degeneration.

Item Number: 28 **Knowledge Classification:** Core

Question: An 82-year-old woman presents with left-sided homonymous hemianopia. The axial FLAIR sequence MRI scan is shown. Five years later she presents with complete loss of vision, although she confabulates answers when asked to name objects presented visually. Her current axial noncontrast CT scan is also shown. Which of the following is the most likely diagnosis?

A. cerebral amyloid angiopathy
B. hemorrhagic conversion of cerebral infarcts
C. hemorrhagic venous infarction from cerebral sinus venous thrombosis
D. hemorrhagic metastases
E. hypertensive intracranial hemorrhage

Rationale: This patient has consecutive occipital lobar hemorrhages. Multiple hemorrhages restricted to lobar, cortical, or cortico-subcortical regions are characteristic of probable cerebral amyloid angiopathy according to the Modified Boston Criteria. The diagnosis also can be suggested by a single lobar hemorrhage accompanied by focal or disseminated superficial siderosis in a patient age >55 years in the absence of other likely causes of hemorrhage or superficial siderosis. The 5-year interval between hemorrhages in this patient argues against hemorrhagic venous infarction from cerebral sinus venous thrombosis or hemorrhagic metastases. The lobar location of the lesions is atypical for hypertensive intracranial hemorrhage. Hemorrhagic conversion of cerebral infarcts is possible but less likely than cerebral amyloid angiopathy given the appearance of the hemorrhage.

Item Number: 30 **Knowledge Classification:** Intermediate

Question: A 63-year-old woman with a history of hypertension presents with a sudden onset of confusion. During the episode, she was able to recognize her son but forgot he was in a motor vehicle crash 2 months ago. She repeatedly asks why she is in the emergency department and states she needs to pick up her daughter's wedding dress. Her forward digit span is 6. Neurologic examination and functioning are normal. Which of the following is the most likely diagnosis?

A. acute confusional state
B. partial complex seizure
C. transient global amnesia
D. transient ischemic attack involving the dominant hippocampus
E. Wernicke encephalopathy

Rationale: This patient is unable to form new memories, which is classic for an amnestic disturbance. She has retrograde and anterograde amnesia. Although the event had a sudden onset similar to a stroke or transient ischemic attack, bilateral impairment is required for amnesia. Acute confusional states are manifested by impaired attention and are inconsistent with a forward digit span of 6. Partial complex seizures are manifested by altered awareness. Wernicke encephalopathy is usually associated with ophthalmoplegia and gait disturbance in the setting of malnutrition.

Item Number: 32 **Knowledge Classification:** Subspecialty

Question: A 19-year-old woman presents with a history of psychosis, tonic-clonic seizures, and encephalopathy. Her EEG is shown. Which of the following diagnostic tests should be ordered to confirm the etiology of these findings?

A. CSF 14-3-3 protein
B. continuous EEG monitoring
C. hepatic transaminases and ammonia
D. NMDA receptor antibody
E. somatosensory evoked potentials

Rationale: This patient's symptoms are consistent with a typical presentation of N-methyl-D-aspartate (NMDA) receptor encephalitis. The EEG reveals extreme delta brushes with rhythmic delta activity and superimposed beta, which has been seen in close association with this syndrome. Antibodies in the CSF can confirm the diagnosis. Although continuous EEG can aid in capturing potential seizures, it will not help in confirming the diagnosis. Liver function tests and ammonia levels would be beneficial in confirming a metabolic encephalopathy, and the EEG may have revealed triphasic waves. Somatosensory evoked potentials may be helpful in identifying hypoxic injury in which case the EEG may also reveal diffuse slow waves but not extreme delta brushes. Analysis of 14-3-3 protein may be helpful in the diagnosis of Creutzfeldt–Jakob disease, and the EEG may reveal periodic discharges that may be asymmetric depending on the timing of the disease.

Item Number: 33 **Knowledge Classification:** Subspecialty

Question: A 46-year-old man presents with a 4-year history of weakness that started in the left lower extremity then progressed to the left hand and right leg over the next 2 months. He reports his muscles "feel jumpy." Examination reveals pronator drift in the right upper extremity, diffuse 4-/5 weakness in the other three limbs, diffuse hyperreflexia in all four extremities with upgoing toes, a snout reflex, and fasciculations and atrophy in all four limbs. Which of the following findings is most likely to be seen on EMG/nerve conduction studies?

A. enlarged polyphasic motor unit action potentials
B. reduced sensory and motor responses
C. slowed conduction velocity without temporal dispersion
D. motor amplitude decrement with 2-Hz repetitive stimulation
E. myotonic discharges

Rationale: Patients with ALS have normal sensory responses but decreased motor amplitudes on nerve conduction studies. EMG shows enlarged polyphasic motor unit potentials and reduced recruitment. Reduced sensory and motor responses are seen in association with peripheral nerve lesion distal to the dorsal root ganglia. Slowed conduction velocity without temporal dispersion is seen in type 1 Charcot-Marie-Tooth disease. A motor amplitude decrement with 2-Hz repetitive stimulation is seen in neuromuscular junction disorders. Myotonic discharges are seen most frequently in myotonic dystrophies, congenital myotonic syndromes, acid maltase deficiency, and hyperkalemic periodic paralysis.

Item Number: 34 **Knowledge Classification:** Subspecialty

Question: A 69-year-old woman with a history of chronic migraine, anxiety, and depression is admitted for severe headache. She takes chlorpromazine, bupropion, and lorazepam daily. Intravenous dihydroergotamine is recommended. Which of the following is the most appropriate next step in management?

A. initiate an infusion of dihydroergotamine without an ECG as dihydroergotamine does not affect cardiac conduction
B. order an ECG to assess for an increased PR interval from bupropion as dihydroergotamine can increase the PR interval further
C. order an ECG to assess for an increased PR interval from lorazepam as dihydroergotamine can increase the PR interval further
D. order an ECG to assess for an increased QTc interval from bupropion as dihydroergotamine can increase the QTc interval further
E. order an ECG to assess for an increased QTc interval from chlorpromazine as dihydroergotamine can increase the QTc interval further

Rationale: Antiemetics, including metoclopramide, chlorpromazine, and prochlorperazine, are used to treat nausea associated with migraine. However, these medications are also associated with an increased risk of QT interval prolongation and torsades de pointes. Risk factors for QT prolongation with antiemetics include high drug concentrations from frequent use, concurrent use with other medications that cause QT prolongation, such as dihydroergotamine, a baseline QT prolongation or T wave lability, bradycardia, electrolyte disturbances such as hypokalemia, and hypomagnesaemia, impaired hepatic and renal function, and underlying heart disease.

Item Number: 35 **Knowledge Classification:** Subspecialty

Question: Needle EMG of which of the following muscles would be most helpful in distinguishing a common peroneal neuropathy in the thigh from a peroneal neuropathy at the fibular head?

A. adductor magnus
B. peroneus longus
C. short head of the biceps femoris
D. tibialis anterior
E. vastus lateralis

Rationale: The short head of the biceps femoris is innervated by the common peroneal nerve in the thigh. This muscle would be spared in a lesion of the peroneal nerve at the fibular head.

Item Number: 36 **Knowledge Classification:** Subspecialty

Question: A 41-year-old woman presents with a 7-year history of progressive ataxia of the lower extremities with recent involvement of the upper extremities. Gait is wide-based and dysmetria is present in both upper and lower extremities. She has impaired smooth ocular pursuits and horizontal nystagmus. Family history reveals episodic ataxia in a first cousin. Which of the following entities is most likely affected in this disorder?

A. GABA receptor
B. inward-rectifier potassium channel
C. N-methyl-D-aspartic acid glutamate receptor
D. voltage-gated calcium channel
E. voltage-gated sodium channel

Rationale: Spinocerebellar ataxia type 6 (SCA6) is a degenerative disorder of the cerebellum with a autosomal dominant inheritance pattern and characterized by nearly selective and progressive death of Purkinje cells. The underlying gene mutation consists of an expansion of a trinucleotide CAG repeat in the 3' region of the CACNA1A gene, encoding the 1A subunit of the neuronal P/Q-type voltage-gated calcium channel. Affected individuals have 20 to 33 CAG repeats. Characteristic findings include adult onset (mean, 43 to 52 years), slowly progressive cerebellar ataxia, dysarthria, and nystagmus. Patients often present with gait unsteadiness, stumbling, imbalance (~90%), and dysarthria (~10%). Gait ataxia, upper limb incoordination, intention tremor, and dysarthria eventually develop in all patients. Dysphagia and choking are also common. Visual disturbances may result from diplopia, difficulty fixating on moving objects, horizontal gaze-evoked nystagmus, and vertical nystagmus. Hyperreflexia and extensor plantar responses occur in up to 50% of patients. Basal ganglia signs, including dystonia and blepharospasm, occur in up to 25%. Mental status is generally preserved. Episodic ataxia type 2 is also due to point mutations in the CACNA1A gene coding for the alpha 1A voltage-dependent calcium channel subunit. However, small repeat expansions of the CAG motif have been found in family members presenting with either spinocerebellar ataxia or episodic ataxia.

Item Number: 39 **Knowledge Classification:** Core

Question: A 62-year-old woman presents 1 hour after developing right-sided weakness and language difficulties. Examination reveals global aphasia, left gaze preference, and right-sided hemiplegia. Blood pressure is 140/95 mm Hg. She takes no medications. A CT scan of the head is normal. Which of the following tests should be performed prior to administration of IV tissue plasminogen activator (tPA)?

A. chest radiography
B. echocardiography
C. MRI of the brain
D. serum glucose
E. stool guaiac

Rationale: Hypoglycemia can present with signs of focal neurologic dysfunction and mimic stroke. Blood glucose should always be checked prior to administration of IV tPA, whereas stool guaiac, chest radiography, brain MRI, and echocardiography are not required.

Item Number: 40 **Knowledge Classification:** Intermediate

Question: A 43-year-old woman is admitted with a 3-month history of progressive cognitive decline, apraxia, and aphasia. Examination reveals myoclonus and rigidity. Which of the following MRI sequences is the most likely to show an abnormality?

A. diffusion-weighted imaging
B. fast-spin echo
C. gradient echo
D. T1-weighted with contrast
E. T2-weighted without contrast

Rationale: In patients with Creutzfeldt-Jakob disease, MRI has been shown to have higher specificity and sensitivity than EEG and CSF testing for 14-3-3 protein. Of the sequences available, the diffusion-weighted/apparent diffusion coefficient sequence is likely to show all abnormalities, including hyperintensity in the cortical gyri (cortical ribboning), caudate, and thalamus. Fast-spin echo and gradient echo identify blood products. T1 with contrast shows lesions where the blood-brain barrier has broken down. T2 without contrast is of limited utility except to highlight atrophy that would not be expected in this type of rapidly progressive dementia.

Item Number: 42 **Knowledge Classification:** Subspecialty

Question: A 6-year-old boy presents with multiple episodes a day of twisting of the right arm and leg. The episodes are brief and brought on by sudden movement. Which of the following is the most appropriate management?

A. acetazolamide
B. carbamazepine
C. methylphenidate
D. psychological counseling
E. trihexyphenidyl

Rationale: Paroxysmal kinesigenic dyskinesia may be sporadic or inherited in an autosomal dominant fashion. It responds well to low-dose anticonvulsants, such as carbamazepine.

Item Number: 43 **Knowledge Classification:** Intermediate

Question: A senior resident is working with a junior resident who is known to be rude when services call for a consult. While in the emergency department, the senior resident witnesses the junior resident refusing to admit a patient and yelling at the attending physician. Which of the following is the most appropriate course of action?

A. Direct the junior resident to manage patients on the ward while the senior resident assumes responsibility for this patient.
B. Ask the program director if the junior resident can be assigned to a different service.
C. Notify the neurology attending about the situation at the end of the rotation.
D. Immediately discuss the case privately with the junior resident, then notify the program director soon after.
E. Mandate that the junior resident attend anger management training.

Rationale: The situation should be discussed in private immediately, then reported to the program director. This resident's pattern of behavior needs a higher level of intervention.

Item Number: 44 **Knowledge Classification:** Subspecialty

Question: A 12-year-old girl presents with subacute decline in cognition, psychotic symptoms, and bilateral Babinski signs. Her MRI scan is shown. Which of the following is the most likely diagnosis?

A. 18q-syndrome
B. Alexander disease
C. juvenile-onset neuronal ceroid lipofuscinosis
D. metachromatic leukodystrophy
E. vanishing white matter disease

Rationale: Metachromatic leukodystrophy is an autosomal recessive disorder in which neurocognitive symptoms can be the primary presentation in the late juvenile onset form, with relatively subtle signs on neurologic examination. Long tract signs alone are common. Imaging studies usually demonstrate involvement of the periventricular and deep white matter with relative sparing of the U fibers.

Item Number: 45 **Knowledge Classification:** Subspecialty

Question: A 13-year-old boy with epilepsy develops confusion and fatigue after walking home on a hot day. No seizure activity was witnessed. His only medication is zonisamide. His temperature is 107.6°F (42°C), but his neurologic and general medical evaluation is otherwise normal. Which of following is the most likely mechanism of his increased body temperature?

A. peripheral numbness and tingling
B. hypohidrosis
C. Stevens Johnson syndrome
D. urinary stone
E. decreased appetite

Rationale: Hypohidrosis is an infrequent side effect of zonisamide, particularly in children and can lead to symptoms of heatstroke due to impaired heat dissipation mechanisms and produce hyperthermia.

Item Number: 46 **Knowledge Classification:** Core

Question: A 27-year-old woman develops weakness in the right arm and hand after fracturing her humerus. She has intact elbow extension but weakness of supination, wrist extension, and finger extension. Sensory loss is noted over the dorsal aspect of the hand between the thumb and index finger. Which of the following nerves most likely has been injured?

A. anterior interosseous
B. median
C. posterior interosseous
D. radial
E. ulnar

Rationale: A radial neuropathy is characterized by wrist drop, finger drop, weakness of elbow flexion when the arm is half pronated (brachioradialis), weakness of elbow supination (supinator), and sensory loss on the dorsal lateral aspect of the hand. Extension at the elbow (triceps) is preserved with radial nerve injury at this level. Finger abduction can appear to be weak when tested in a wrist drop position and may mistakenly suggest ulnar nerve involvement. It is important to extend the wrist passively to test finger abduction properly.

Item Number: 50 **Knowledge Classification:** Intermediate

Question: A 65-year-old woman with a history of hypertension is concerned about her risk for Alzheimer disease and asks for advice regarding nonprescription interventions that may help decrease the risk. Which of the following interventions has been demonstrated to provide cognitive protection in this patient population?

A. elimination of dairy from her diet
B. feeding and caring for a pet
C. increasing cardiorespiratory fitness
D. regular use of coconut oil
E. regular use of ginkgo biloba

Rationale: Use of supplements such as ginkgo biloba and coconut oil has not been proven to provide cognitive protection, but improving cardiorespiratory fitness has been shown in many studies to help reduce the risk of dementia, possibly by improving white matter integrity. A dairy-free diet and feeding and caring for a pet do not reduce the risk of developing cognitive issues.

Item Number: 53 **Knowledge Classification:** Intermediate

Question: 19-year-old woman who presents with a severe persistent headache for the past 2 days postpartum reports pain whenever she tries to rise to a sitting or standing position. History reveals she received epidural anesthesia during labor and delivery. Axial T1 post-gadolinium, T2-weighted, and coronal T1 post-gadolinium MRI scans are shown.

A. bacterial meningitis
B. eclampsia
C. HELLP (hemolysis, elevated liver enzyme levels, and low platelet count) syndrome
D. intracranial hypotension
E. posterior reversible encephalopathy syndrome

Rationale: Intracranial hypotension is the diagnosis. As is typical, the MRI scans show prominent thickening and enhancement in the dura, with small subdural effusions seen on the T2-weighted axial image. The patient reports symptoms typical of a postural headache and had epidural anesthesia during her recent labor and delivery.

Eclampsia and posterior reversible encephalopathy syndrome are related in that eclampsia is a specific form of posterior reversible encephalopathy that occurs exclusively in pregnancy. HELLP syndrome is a complication of pregnancy characterized by hepatic and hematologic abnormalities in a small fraction of patients with eclampsia. The brain imaging findings for these diagnoses are identical and consist of regions of abnormal hyperintense T2 and FLAIR signal in cortical and subcortical tissue, typically in the posterior portions of the cerebral hemispheres. These findings are not present in the images shown. Rather, the abnormality is diffuse non-nodular dural thickening. Meningitis is not a good option based on the clinical presentation of postural headache; with meningitis, the headache is not postural and is typically associated with nuchal rigidity, which not present in this patient. Also, the pattern of abnormal enhancement is not in the subarachnoid space, which typically is present in about half of patients with meningitis; the other half may not have abnormal findings on imaging, explaining the importance of CSF analysis in meningitis.

Item Number: 60 **Knowledge Classification:** Intermediate

Question: A 17-year-old college student is brought to the emergency department after having a seizure the morning after staying up all night studying. She has no history of epilepsy but recalled having several episodes of hand jerking upon wakening when she was a teenager. Which of the following EEG findings is most consistent with her clinical presentation?

A. anterior temporal spike and wave transients
B. generalized 3-Hz spike wave discharges
C. generalized paroxysmal fast wave activity
D. generalized polyspikes
E. temporal intermittent rhythmic delta

Rationale: Patients with juvenile myoclonic epilepsy (JME) may have a variety of interictal abnormalities, but polyspikes are specific for this disorder. JME comprises up to 10% of all epilepsies, with median age at onset 15 years. Hallmark features are myoclonic jerks with retained consciousness and generalized tonic-clonic seizures. Only one third of patients with JME have absence seizures.

Item Number: 62 **Knowledge Classification:** Core

Question: A 19-year-old woman presents with excessive daytime drowsiness. Approximately 1 year ago she began to note episodes of extreme sleepiness during the day, necessitating a midday nap. Over the last 3 months, she has had vivid dreams when falling asleep and upon awakening. On a few occasions, she has been unable to move her arms and legs for several seconds after awakening. Her neurologic examination is normal. Which of the following additional historical features is most likely to be present?

A. childhood physical or sexual ab
B. kicking her legs in her sleep
C. loss of muscle tone with emotional upset
D. loud snoring after 3 hours of sleep
E. sleepwalking

Rationale: This patient gives a classic history for narcolepsy. She has three of the typical clinical features of narcolepsy: excessive daytime sleepiness, hypnagogic hallucinations, and sleep paralysis. She should be questioned about the presence of cataplexy, which is present in narcolepsy type 1. Narcolepsy type 2 does not have cataplexy and usually has normal CSF levels of hypocretin.

Item Number: 71 **Knowledge Classification:** Core

Question: A 15-year-old girl experiences acute onset of leg weakness without any changes in mental status. She is found to have 20/80 visual acuity in the left eye, hyperreflexia in the legs, upgoing plantar responses, and vibratory sensation loss at the great toes. MRI scans show T2-hyperintense lesions in the corpus callosum, periventricular white matter, left optic nerve, and C2-3 region of the cord. Which of the following is the most likely diagnosis?

A. acute disseminated encephalomyelitis
B. adrenoleukodystrophy
C. combined system degeneration
D. multiple sclerosis
E. neuromyelitis optica

Rationale: This combination of findings represents classic multiple sclerosis. Posterior column findings are particularly important diagnostically. Although acute disseminated encephalomyelitis may occur in adolescents and adults, encephalopathy is characteristically present. Neuromyelitis optica spectrum disorder has less significant brain involvement, and spinal cord changes are typically more longitudinally extensive. Combined system degeneration spares the eyes and does not produce the imaging changes noted. Adrenoleukodystrophy with adolescent onset is usually myelitic but almost always affects young men and does not manifest the additional findings or imaging changes noted here.

Item Number: 73 **Knowledge Classification:** Core

Question: A 78-year-old woman presents after abrupt development of difficulty with vision 1 hour ago. Examination reveals a blood pressure of 170/90 mm Hg and a right-sided homonymous hemianopsia. CT scan reveals a left occipital intraparenchymal hemorrhage, and gradient-echo MRI scan shows two old microbleeds in the left frontal and right parietal lobes. Routine coagulation studies are normal. Which of the following is the most likely etiology?

A. amyloid angiopathy
B. arteriovenous malformation
C. capillary telangiectasia
D. hypertensive hemorrhage
E. dural arteriovenous fistula

Rationale: Cerebral amyloid angiopathy is a common cause of lobar intracerebral hemorrhage in elderly patients and is implicated as a potential cause of chronic microbleeds on gradient echo MRI. Hypertensive hemorrhages usually affect the small penetrating arteries and cause deep brain hemorrhages. Arteriovenous malformations usually present between the ages of 10 to 40 years. In dural arteriovenous fistulas, MRI findings are usually normal. Capillary telangiectasia is most commonly found in the pons and associated with a small amount of hemorrhage and gliosis.

Item Number: 75 **Knowledge Classification:** Core

Question: A 25-year-old man reports the onset of ascending paresthesias in the arms and legs 2 days ago, followed by weakness in both arms and legs 24 hours ago. Today he notes shortness of breath. Vital signs are as follows: BP 120/70 mm Hg, HR 110 beats/min and regular, RR 25 breaths/min, temperature 98.6°F (37°C), oxygen saturation 97%. He is dyspneic and uses accessory muscles for breathing. Neurologic examination reveals facial weakness bilaterally, 4/5 strength in the upper and lower extremities, absent deep tendon reflexes, and mild abnormalities on sensory examination. Which of the following is the most appropriate immediate next step in management?

A. chest radiograph
B. lumbar puncture
C. MRI of the spine
D. nerve conduction studies
E. spirometry

Rationale: Respiratory muscle weakness associated with ventilatory failure is a potentially serious complication in patients with acute inflammatory demyelinating polyradiculopathy. Spirometry is the most effective measure of respiratory muscle function in these patients. Oxygen saturation is not a sensitive marker for impending respiratory dysfunction in neuromuscular disorders.

Item Number: 76 **Knowledge Classification:** Intermediate

Question: A 64-year-old woman presents with a rapidly progressing dementing illness associated with myoclonic jerking. There is no family history of a similar disorder. MRI scans show increased signal in the caudate and putamen, and EEG reveals periodic sharp wave complexes. Which of the following is the most likely route of disease acquisition in this patient?

A. iatrogenic
B. ingestion
C. inhalation
D. inherited
E. sporadic

Rationale: Creutzfeldt-Jakob disease is a rapidly progressive dementing illness with characteristic hyperintensity of the caudate and putamen seen on MRI studies, positive 14-3-3 protein, and periodic sharp wave complexes on EEG. The variant form, which can be caused by ingestion or iatrogenic exposure to contaminated material, typically has a psychiatric presentation. The sporadic form is the most common and presents with multifocal neurologic symptoms and dementia. In a patient with no family history, the sporadic form is the most likely etiology.