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A 74-year-old patient is hospitalized with a right middle cerebral artery stroke. During the work-up, the carotid duplex reveals a stenosis of 55% in the ipsilateral carotid artery.

Is carotid endarterectomy (CE) indicated for this patient?

A. Yes  
B. No
The North American Symptomatic Carotid Endarterectomy Trial (NASCET) showed a significant benefit of CE for patients with $\geq 70\%$ symptomatic stenosis. Two-year ipsilateral stroke risk was 26% in the patients treated with medication and 9% in the CE group ($P < .001$).

A combined analysis of trials of symptomatic patients showed a benefit of CE for 50% to 69% stenosis (adjusted relative risk [ARR], 4.6% over 5 years) and for $\geq 70\%$ (ARR, 16% over 5 years). The overall rate of stroke or death for all surgical patients within 30 days of trial surgery was 7.1%.

For most of the following patients, surgical removal of an intracranial hemorrhage (ICH) with craniotomy is indicated or should be considered. For which patients is routine evacuation with craniotomy NOT recommended?

A. Patients with supratentorial ICH who are within 96 hours of ictus
B. Patients with lobar clots within 1 cm of the surface
C. Patients with cerebellar hemorrhage > 3 cm in diameter who are deteriorating neurologically
D. Patients with brainstem compression or ventricular obstruction resulting from hemorrhage
A. Patients with supratentorial ICH who are within 96 hours of ictus.

The decision about whether and when to operate on ICH remains controversial. The following recommendations are based on the latest American Heart Association and American Stroke Association guidelines. Patients with small hemorrhages or minimal neurologic deficits should be treated medically because they generally do well with medical treatment alone. Patients with a GCS score \( \leq 4 \) should also be treated medically because their outcome is generally not improved by surgery. Patients with a cerebellar hemorrhage \( \geq 3 \) cm in diameter who are deteriorating neurologically or who have brainstem compression and hydrocephalus should have surgery as soon as possible. Routine evacuation of supratentorial hemorrhage by standard craniotomy is not recommended within 96 hours of ictus. However, patients with lobar clots within 1 cm of the surface may be considered for surgical evacuation.

An 86-year-old man presents to the emergency room within 1 hour of onset of right hemiplegia and global aphasia. He has no significant medical history, and his laboratory data are all normal. Neuroimaging finds no hemorrhage and no signs of acute ischemia. The emergency room physician is unsure whether to give intravenous (IV) tissue plasminogen activator (tPA). He asks for your recommendation on giving IV tPA.

Should he give IV tPA to this patient?

A. Yes
B. No
Although information on the safety and efficacy of IV tPA in very old patients with acute ischemia is scarce, some data suggest that the risk for bleeding is not increased in elderly patients. A study showed that the rate of parenchymal hemorrhage was not significantly higher in elderly patients (>= 80 years of age) compared with younger patients (6.3% vs 5.3%; P = 1.000). The rate of symptomatic intracerebral hemorrhage was similar between the groups (2.6% vs 2.6%; P = 1.000).

A 45-year-old woman presents to the emergency room with the worst headache of her life. A lumbar puncture reveals many red blood cells, and the head CT shows a subarachnoid hemorrhage. Intracranial CT angiography reveals a left posterior communicating aneurysm.

What is the most appropriate treatment for this patient?

A. Watchful waiting
B. Craniotomy and clipping of the aneurysm
C. Endovascular coiling
D. Hypothermia
C. Endovascular coiling.

The International Subarachnoid Trial (ISAT) was stopped early when use of endovascular coils for the treatment of subarachnoid hemorrhage demonstrated significant benefit compared with craniotomy and the use of vascular clips. In this study of 2143 patients, the rate of death or dependence (modified Rankin scale score of 3 to 6) was 30.6% in the neurosurgery group and 23.7% in the endovascular group ($P = .0019$). Mortality was 10.1% for the neurosurgical group and 8.1% for the endovascular group. The study suggests that the most appropriate treatment for patients with subarachnoid hemorrhage related to a ruptured intracranial aneurysm usually is endovascular coiling.

What is the approximate 7-day risk for stroke after transient ischemic attack (TIA)?

A. 0.05%
B. 1%
C. 5%
D. 20%
E. 40%
C. 5%.

A meta-analysis by Giles and Rothwell showed that the risk for major stroke after TIA is 5.2% at 7 days. The investigators reviewed 18 cohort studies with a total of 10,126 patients who had experienced a TIA. This was the first meta-analysis of stroke risk for the early period after TIA; results of previous, individual studies have been inconsistent.

What percentage of patients with Parkinson's disease (PD) suffer from depression?

A. 1%
B. Less than 10%
C. More than 25%
D. Up to 40%
E. 70%
D. Up to 40%.

Depression is underdiagnosed in patients with PD. It can affect up to 40% of patients and is the most common neuropsychiatric manifestation of PD. Indeed, symptoms of depression can precede the onset of motor manifestations of PD. The diagnosis of depression in PD is challenging because many symptoms of depression (apathy, anhedonia, sleep and appetite disturbances, and changes in libido) overlap with the primary symptoms of PD. The most commonly used scales to screen for depression are the Beck Depression Scale, Hamilton Depression Scale, and the Geriatric Depression Scale.

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A 65-year-old man presents to your clinic after falling several times within the past few months. On examination, his most notable findings are an unstable, wide-based gait and marked retropulsion. He does have bradykinesia with masked facies and dysarthria. You find no evidence of cogwheeling or resting tremor. His symptoms do not respond to levodopa/carbidopa. During the next few months, his eye movements are notable for slowing of vertical saccades and fast phases.

What is your diagnosis?

A. L-dopa-resistant Parkinson’s disease
B. Progressive supranuclear palsy
C. Normal pressure hydrocephalus
D. Amyotrophic lateral sclerosis
B. Progressive supranuclear palsy.

Progressive supranuclear palsy (PSP) is a neurodegenerative disease that affects eye movements, posture, and cognition. It was first described in 1964. The cardinal manifestations of PSP are supranuclear ophthalmoplegia, pseudobulbar palsy, prominent neck dystonia, parkinsonism, behavioral and cognitive disturbances, and gait imbalance. The mean age at onset is 63 years. The diagnosis is purely clinical; PSP is associated with no specific laboratory or imaging findings, although neuroimaging studies may be used to rule out other disorders. Medication is generally ineffective, and the response in patients who do respond is short-lived and incomplete. No medication is effective in halting the progression of PSP.

Q

Which of the following statements about impulse control disorder (ICD) and Parkinson's disease (PD) is true?

A. Patients with PD only display problems with impulse control when they are on dopamine agonists
B. The incidence of ICD is higher in patients with PD who are not taking dopamine agonists
C. The incidence of ICD is higher in patients with PD when they are taking dopamine agonists
D. The problem of ICD is not typically associated with PD
According to the American Psychiatric Association's Diagnostic and Statistical Manual (DSM), ICDs are characterized by "the failure to resist an impulse, drive, or temptation to perform an act that is harmful to the person or to others." Although the incidence of ICD in patients with PD is as high as 13.7% when they are taking a dopamine agonist, several studies have shown that 6% to 7% of patients with PD meet the criteria for an ICD even when they are not receiving treatment with a dopamine agonist. The most common ICD behaviors reported in association with PD are gambling, sex, shopping, and eating. Both pathologic gambling and compulsive sexual behavior or hypersexuality occurred in approximately 2% to 4% of PD patients.

What dose of coenzyme Q10 (CoQ10) has been shown to have an effect on early Parkinson's disease?

A. 75 mg  
B. 150 mg  
C. 300 mg  
D. 600 mg  
E. 1200 mg
CoQ10 is an antioxidant and a mitochondrial energy stabilizer. Mitochondrial dysfunction is present in PD and is theorized to be one of the factors that can initiate the apoptosis cascade. Patients with PD have been reported to have significantly lower serum CoQ10 levels than age-comparable patients. A preliminary study found that subjects with early PD receiving 1200 mg of CoQ10 daily experience significantly less decline than patients treated with placebo. A more recent study showed that patients with midstage PD receiving 300 mg daily did not note improvements in PD symptoms. A large study of CoQ10 in patients with early PD is planned.


A 52-year-old man comes to your office at the urging of his wife, who cannot get a good night’s sleep because the patient gets out of bed several times each night and paces the room. He also occasionally kicks his wife inadvertently in the middle of the night. He states that he feels like ants are crawling over his legs, particularly when he is trying to sit still and at night.

**What is the diagnosis?**

A. Peripheral neuropathy with paresthesias  
B. Restless legs syndrome with periodic limb movements of sleep  
C. Parasomnias  
D. Parkinson’s disease with paresthesias
A

Restless legs syndrome (RLS) is characterized by paresthesias or dysesthesias, an urge to move the limbs, and motor restlessness. The onset or exacerbation of symptoms occurs in the evening and improves with limb movement or walking. Despite its common occurrence (2% to 15%), it is underrecognized and misdiagnosed. In order to make a diagnosis of RLS, patients should meet the criteria of the International RLS Study Group and the National Institutes of Health: (1) an urge to move the legs; (2) temporary relief with movement; (3) onset or worsening of symptoms with rest or inactivity; and (4) worsening or onset of symptoms in the evening or night. Many patients with RLS also have periodic limb movements of sleep. These are stereotyped, repetitive flexion movements of the legs more than the arms, which may arouse patients from sleep and can interfere with the bed partner’s ability to sleep.


B. Restless legs syndrome with periodic limb movements of sleep.
Can the diagnosis of relapsing-remitting multiple sclerosis (MS) be made with imaging alone?

A. Yes
B. No
A

A. Yes.

The findings from MRI were officially incorporated into the International Panel Criteria, the diagnostic criteria of MS, in 2001. The International Panel Criteria incorporated the McDonald Criteria as the criteria for the diagnosis of MS based on MRI. In order to make the diagnosis of MS based on MRI findings, there must be demonstration of dissemination in time or space on the MRI.

For MRI lesions disseminated in space, at least 3 of the following criteria must be met:

- One gadolinium-enhancing lesion or at least 9 T2-hyperintense lesions if there is no gadolinium-enhancing lesion
- At least 1 infratentorial lesion
- At least 1 juxtacortical lesion
- At least 3 periventricular lesions

For MRI lesions disseminated in time:

- If MRI is done more than 3 months after the clinical event, then a gadolinium-enhancing lesion at a site different from the original clinical event is sufficient; if there is no gadolinium enhancement, then a follow-up scan must show a new T2 or gadolinium-enhancing lesion.
- If the first MRI scan occurs less than 3 months after the onset of the clinical event, then a second scan more than 3 months later showing a new gadolinium-enhancing lesion or a new T2 lesion fulfills the requirement.

An 18-year-old woman is referred to your office after branch retinal artery occlusions occurred in each eye several months apart. Her examination reveals moderate hearing loss in the right ear in addition to visual field cuts in each eye. A brain MRI reveals multiple small cortical strokes in the frontal lobes and left parietal lobe.

What is the diagnosis?

A. Multiple sclerosis
B. Acute disseminated encephalomyelitis
C. Susac syndrome
D. Conversion reaction
C. Susac syndrome.

Susac syndrome is a microangiopathy involving the arterioles of the brain, the retina, and the cochlea and clinically presents with subacute encephalopathy, branch retinal artery occlusion, and sensorineural hearing loss.

A 25-year-old woman presents to an emergency room having had blurred vision in her right eye for several days. She denies any other neurologic symptoms. You see her in consultation and find a visual acuity of 20/200 in the right eye, an afferent papillary defect, and a pale right optic nerve. A brain MRI reveals several T2/FLAIR lesions in the supratentorial white matter, but no other lesions. She is diagnosed with a clinically isolated syndrome (CIS). You review the different treatment options with her. She comes back to your office and states that she is unsure about starting therapy due to the cost of medication and inconvenience of treatments. She states that since she does not have multiple sclerosis, the medications are not needed.

Is her perception correct?

A. Yes
B. No
The Betaferon/Betaseron in Newly Emerging MS for Initial Treatment (BENEFIT) study, which evaluated interferon-beta-1b in patients with CIS, demonstrated that with early treatment, the risk for a second relapse diagnostic of clinically definite MS (CDMS) was reduced by 50% compared with delaying treatment for 2 years or until the next event. A second phase of the study showed that patients treated sooner had a lower risk for disability progression based on the Expanded Disability Status Scale score. Early treatment not only delayed the diagnosis of CDMS but reduced disability progression after 3 years.

Which of the following features of early multiple sclerosis are not predictive of long-term disability? (More than one answer may be correct.)

A. Frequent relapses during the first 2 to 5 years
B. Optic neuritis occurs within the first 2 years
C. Incomplete recovery from relapses
D. Large lesion volume
E. Interval between relapses during the first 2 years
Both clinical and MRI measures of disease activity during the first 2 to 5 years after a clinically isolated syndrome are important predictors of long-term disability. Natural history studies have shown that frequency of relapses during the first 2 to 5 years, intervals between relapses during the first 2 years, and incomplete recovery from relapses during the first 5 years are robust predictors of long-term disability. In a study of 79 patients with clinically isolated syndrome, the most robust predictor of disease progression was large lesion volume. No study has shown that the occurrence of optic neuritis within the first 2 years predicts long-term disability.

A 72-year-old man comes to your office because he is worried about his memory. He states that he has been having more difficulty remembering where he places items. He has already had to replace his reading glasses several times because he misplaced them. He is still working as an English professor and has not had any difficulties at work related to his memory problems. Cognitive testing reveals only moderate retrieval deficit with all other cognitive functions intact.

**What is the diagnosis?**

A. Pick’s disease  
B. Frontal lobe dementia  
C. Mild cognitive impairment  
D. Vascular dementia
C. Mild cognitive impairment.

Mild cognitive impairment (MCI) refers to the transitional state between the cognitive changes of normal aging and the fully developed clinical features of dementia. MCI has garnered much attention from researchers as a treatment target for neurodegenerative processes before extensive damage has occurred. The 2 subtypes of MCI are amnestic (including memory impairment) and nonamnestic (other nonmemory cognitive domains impaired). MCI is diagnosed when a patient has a cognitive symptom that is not normal for age and not attributable to dementia, with essentially normal functional activities.

Is the following statement true or false? Serologic evaluations, CSF analysis, and neuroimaging with CT/MRI are ordered in the work-up of dementia to find biologic markers and diagnostic imaging patterns for the diagnosis of Alzheimer’s disease (AD).

A. True
B. False
Serologic evaluations, CSF analysis, and neuroimaging with CT/MRI are ordered in the work-up of dementia to exclude alternate causes of dementia, especially reversible or treatable causes. Currently, no laboratory test can confirm the diagnosis of AD. Although many markers for AD are being investigated, no marker has been shown to significantly improve diagnostic accuracy. CT and MRI are limited in that the common radiologic findings of AD are neither sensitive nor specific to AD. Other imaging modalities, specifically positron emission tomography (PET) and single-photon emission computed tomography (SPECT) have shown promise in providing better sensitivity and specificity for AD, but neither has been validated with pathologic comparisons.

What is the second most common type of degenerative dementia?

A. Alzheimer’s dementia
B. Dementia with Lewy bodies
C. Frontotemporal dementia
D. Vascular dementia
B. Dementia with Lewy bodies.

Dementia with Lewy bodies (DLB) is the second most common type of degenerative dementia after Alzheimer's dementia, which accounts for 50% to 60% of cases of dementia. Roughly 10% to 15% of patients with degenerative dementia have DLB at autopsy. Core clinical features of DLB are fluctuating cognitive impairment (in 50% to 75% of patients); visual hallucinations; and parkinsonism, seen in 25% to 50% of patients at diagnosis. Features that support this diagnosis include repeated falls, syncope, transient loss of consciousness, neuroleptic sensitivity, hallucinations in other modalities, REM sleep behavior disorder, and depression. Although vascular dementia is more common than DLB, it is not a degenerative dementia.

A 52-year-old woman presents with behavioral changes over the course of 2 years. Her concentration is decreasing, making it difficult to accomplish her work and she does not care about her poor performance. She is eating more and has gained 25 pounds in 4 months. She is being charged with sexual harassment at work related to telling inappropriate dirty jokes. She has stopped bathing and refuses to take care of her personal hygiene. On examination, she has grasp and palmomental reflexes but no other abnormalities. Her Folstein MMSE is 29/30 but her clock drawing test was poor.

What is the diagnosis?

A. Vascular dementia
B. Frontotemporal dementia
C. Normal pressure hydrocephalus
D. Presenile dementia
B. Frontotemporal dementia.

Frontotemporal dementia (FTD) is a degenerative dementia that is characterized by prominent personality and behavioral changes greater than prominent memory loss early in the disease course. Common behavior and conduct disturbances include a loss of personal awareness, loss of social comportment, disinhibition, impulsivity, hyperorality, social withdrawal, stereotyped or preservative behavior, and speech output changes. Physical examination often reveals early prominent primitive reflexes. The 3 principal varieties are frontal variant FTD, semantic dementia, and progressive nonfluent aphasia.

Is the following statement true or false? Atypical antipsychotic drugs are associated with an increased risk for death in elderly patients with dementia.

A. True
B. False
A. True.

Atypical antipsychotic drugs are used to treat agitation in elderly patients with dementia. These agents accounted for approximately 83% of the antipsychotic drugs dispensed in 2002. Atypical antipsychotic drugs are associated with an increased risk for death in elderly patients with dementia. The mortality rate is approximately 1.6 to 1.7 times higher with atypical antipsychotics than with placebo at 30 days of treatment, and the elevated risk continued to 180 days. Based upon this information, the FDA issued a public advisory that use of atypical antipsychotic drugs in elderly patients with dementia was associated with increased risk for death compared with placebo. However, use of conventional antipsychotic drugs was associated with an even greater risk for death than an atypical antipsychotic drug at 30 days (adjusted hazard ratio, 1.26 to 1.55).

All of the following statements about autism spectrum disorders (ASD) are true EXCEPT:

A. Affected children usually lack social reciprocity
B. Profound language delay is universal
C. Rett syndrome is a genetic disease
D. Pervasive developmental disorder not otherwise specified (PDD-NOS) is a commonly diagnosed ASD
E. ASD and PDD are synonymous
ASDs comprise a group of developmental disorders that share profound, early deficits in social reciprocity and communication and the presence of repetitive or restricted behaviors and interests. In toddlers, these difficulties are manifested by reduced frequency of vocalization; failure to point or use eye contact; and limited integration of vocalization, gestures, and facial expressions. This category of disorders is called pervasive developmental disorders in the DSM-IV. Autism is the most well-studied. Patients with Asperger syndrome do not have language or cognitive deficits. Rett syndrome is a rare progressive disorder with a known genetic cause. PDD-NOS is a category devised for patients whose disorder does not quite meet criteria for autism or another ASD but who have sufficient impairment to need services. It is as common as autism.

A 23-year-old woman with classic migraine symptoms is referred to your office for management of her headaches. Her migraines are infrequent enough that prophylactic therapy for migraine is not needed. The patient was prescribed a short-acting triptan by her primary care doctor, with partial but incomplete relief.

Q

What further treatment options are available?

A. A nonsteroidal anti-inflammatory drug
B. A higher dose of triptan
C. A different triptan
D. All of the above
The patient should be tried on a concurrent regimen of the triptan and a nonsteroidal anti-inflammatory drug. A study by Burstein and colleagues suggests that treatment with medications that target serotonin dysmodulation and inflammation in migraine may improve outcomes compared with monotherapy. They evaluated the efficacy and tolerability of a short-acting triptan and a long-acting nonsteroidal anti-inflammatory drug separately and in combination. The study showed significant symptomatic relief of the combination compared with either drug alone or to placebo at 2 hours and 4 hours. Using medications that target multiple mechanisms may be more effective than monotherapy without increasing the incidence of adverse events. Other options include using a higher dosage of the triptan if available and trying another triptan.

A 35-year-old man is referred for headaches. Over the past few weeks, he has developed bilateral, intense, throbbing headaches that occur whenever he goes to the gym and lifts weights. He states that when he stops exercising, the headache recedes within about 15 minutes. He has stopped lifting weights. He states that he is able to do cardiovascular exercise without any headache.

What is the diagnosis?

A. Cluster headache  
B. Atypical migraine headache  
C. Exertional headache  
D. Tension headache
C. Exertional headache.

Exertional headache occurs in roughly 1% of the general population. It can be unilateral or bilateral, throbbing at the onset, and may develop migrainous features. Exertional headache can be prevented by a warm-up period or by preventive medications, such as indomethacin or beta-blockers. The headaches usually resolve on their own within 5 minutes to 24 hours.

Is the following sentence true or false? Medication overuse headache is now a recognized subtype of chronic daily headache.

A. True
B. False
A. True.

The most recent International Classification of Headache Disorders (ICDH-II) distinguishes subtypes of chronic daily headache, especially chronic migraine and medication overuse headache. The diagnosis of medication overuse headache is made if medication overuse is present and the headache developed or worsened with medication overuse.

A 32-year-old woman with complex partial seizures has been seizure-free for 1 year on antiepileptic drugs (AEDs), but she wants to discontinue the drugs. She has normal intelligence and a normal neurologic examination. Her EEG is normal.

Which aspect of her profile does not guarantee successful withdrawal from medication?

A. Seizure-free for 1 year on AEDs
B. Single type of partial or generalized seizure
C. Normal neurologic examination
D. Normal intelligence
E. EEG normalized with treatment
A. Seizure-free for 1 year on AEDs.

The Quality Standards Subcommittee of the American Academy of Neurology (AAN) developed a practice parameter guideline for discontinuing antiepileptic drugs in seizure-free patients. Based upon their analysis, the profile that yields the greatest chance for successful drug withdrawal is (1) seizure-free for 2 to 5 years on AEDs; (2) single type of partial or generalized seizure; (3) normal neurologic examination and normal IQ; and (4) EEG normalized with treatment. Children meeting this profile have at least a 69% chance of successful withdrawal and adults a 61% chance.

Q

Status epilepticus is a medical emergency associated with significant mortality and morbidity that requires immediate and effective treatment. Benzodiazepines are often the first agents used.

Are all benzodiazepines equally effective for treating status epilepticus?

A. Yes
B. No
Lorazepam has been found to be more effective than diazepam for cessation of seizures (relative risk, 0.64 [95% CI, 0.45-0.90]) and provides a lower risk for continuation of status epilepticus requiring a different drug or general anesthesia (relative risk, 0.63 [95% CI, 0.45-0.88]).

A boy with intractable seizures is referred to you for seizure management. A work-up reveals mesial temporal lobe epilepsy with unilateral hippocampal sclerosis.

What is the most appropriate treatment?

A. Continuous intravenous benzodiazepines
B. Temporal lobe resection
C. Ketogenic diet
D. High-dose phenytoin
B. Temporal lobe resection.

Patients with mesial temporal lobe epilepsy are excellent surgical candidates. The results of surgery are good: Long-term freedom from seizures is achieved in 60% to 80% of patients. In a randomized trial comparing surgery with medical management, 58% of surgical patients achieved seizure freedom compared with 8% of the medically treated patients. Longer-term outcomes are consistent with these short-term data.

All of the following are hallmark symptoms of narcolepsy EXCEPT:

A. Persistent daytime sleepiness
B. Loud snoring
C. Cataplexy
D. Hypnagogic hallucinations
E. Sleep paralysis
Narcolepsy is a lifelong condition with a peak onset in the second decade of life. Hallmark symptoms of narcolepsy include persistent daytime sleepiness; cataplexy (ie, an abrupt paralysis or paresis of skeletal muscles that usually follows emotional experiences); hypnagogic hallucinations (ie, vivid and often frightening dreams that occur shortly after falling asleep); and sleep paralysis, a transient global paralysis of voluntary muscles that also usually occurs shortly after falling asleep and lasts a few seconds to minutes. Loud snoring can be a symptom of obstructive sleep apnea but not of narcolepsy.

A 43-year-old man presents to the emergency room with pain in his right shoulder as well as weakness in his right hand and forearm. He states that he awoke with the pain and that it is sharp and intense. His examination reveals weakness that is not localizable to a dermatome or nerve root and involves many different peripheral nerve territories. He also has numbness in the hand.

What is the diagnosis?

A. Delayed-onset multiple sclerosis
B. Acute disseminated encephalomyelitis
C. Stroke
D. Parsonage-Turner syndrome (brachial neuritis)
D. Parsonage-Turner syndrome (brachial neuritis).

Parsonage-Turner syndrome is a rare syndrome of unknown cause that affects the lower neurons of the brachial plexus; the motor neurons are disproportionately affected compared with the sensory nerves. The syndrome is characterized by acute onset of excruciating unilateral shoulder pain followed by ipsilateral arm weakness. Treatment is limited to physical therapy and symptomatic pain relief.

A 69-year-old woman presents to your clinic with vertigo. She states that she started having vertigo 2 nights ago while in bed. She reports that every time she would roll over, she would get vertigo, which would be worse when she rolled to the left than to the right. The vertigo would last for about 40 seconds and then resolve spontaneously.

**What is the best treatment for this patient?**

A. Epley maneuver
B. Complete, supine bedrest until the vertigo resolves
C. Nonsteroidal anti-inflammatory drugs
D. Avoid bright lights until vertigo resolves
A. Epley maneuver.

The patient has benign paroxysmal positional vertigo. It is caused by free-moving canaliths in the posterior semicircular canal. The vertigo is relieved by particle repositioning with the Epley maneuver. The Epley maneuver is simple, can be done in the office, and involves moving the particles by rotating the head in a particular sequence. It can be repeated if the vertigo is improved but not resolved. The patient must avoid lying completely flat for 24 to 48 hours and should sleep with the head elevated on a few pillows or upright.

Which steroid should be given to patients with bacterial meningitis as adjunctive therapy?

A. Intravenous Solu-Medrol
B. Oral prednisone
C. Intravenous dexamethasone
D. None of the above
Bacterial meningitis causes an intense inflammatory response in the enclosed spaces of the brain and spinal cord. This unchecked inflammatory response can lead to significant morbidity and mortality, despite effective antibiotic therapy. Dexamethasone is the only adjunctive therapy for the treatment of patients with bacterial meningitis that has proven clinical efficacy.

A 19-year-old man is hospitalized in a psychiatric ward after developing visual and auditory hallucinations. He has a seizure soon after being admitted and is transferred to the emergency room. There, he is postictal and noted to have a fever and meningeal signs.

What is the most likely diagnosis?

A. Herpes encephalitis  
B. Cerebral toxoplasmosis  
C. Temporal lobe epilepsy  
D. None of the above
A. Herpes encephalitis.

Herpes encephalitis is caused by HSV-1 or HSV-2. Psychiatric symptoms are sometimes the initial and only signs of herpes encephalitis. Headaches, seizures, and aphasia may also occur. Rapid diagnosis of herpes encephalitis is imperative and should not delay treatment. Work-up includes neuroimaging studies, lumbar puncture, and perhaps an EEG. The goals of therapy are to reduce morbidity and to prevent complications. Intravenous acyclovir should be initiated immediately.

Friedreich ataxia is caused by a mutation of a gene locus on which chromosome?

A. Chromosome 4  
B. Chromosome 9  
C. Chromosome 14  
D. Chromosome 19
Friedreich ataxia is an autosomal recessive disorder resulting from a mutation of a gene locus on chromosome 9. It was the first of the inherited ataxias to be recognized and accounts for approximately 50% of cases of hereditary ataxias. Incidence ranges from 1 in 22,000 to 1 in 100,000.

Unintentional falls account for 16,000 deaths in the United States per year. All of the following are risk factors for falls EXCEPT:

A. Advanced age
B. Age-associated frailty
C. Arthritis
D. Hypertension
E. Depression
D. Hypertension.

Patients with dementia, stroke, or gait and balance disorders are at greatest risk for falls. Increased risk for falls is also probable among individuals with Parkinson’s disease, peripheral neuropathy, lower-extremity weakness or sensory loss, and substantial loss of vision. General risk factors for falls include advanced age, age-associated frailty, arthritis, impairments in activities of daily living, depression, and use of psychoactive medications.

What is the estimated prevalence of neuropathic pain in the general population?

A. 0.5%
B. 1.5%
C. 5%
D. 15%
E. 25%
Neuropathic pain has an estimated prevalence in the general population of 1.5% and includes patients suffering from postherpetic neuralgia, diabetic neuropathy, or pain from low back disorders. The most common cause of neuropathic pain in the developed world is diabetes.

Corticosteroids are often used in patients with brain tumors to reduce tissue swelling and to control signs and symptoms. These drugs can improve neurologic symptoms and reduce cerebral edema within the first 8 to 48 hours. Dexamethasone is the corticosteroid of choice for brain tumor patients.

Is the dosing of oral and parenteral forms of dexamethasone equivalent?

A. Yes
B. No
A. Yes.

Dosing of oral and parental glucocorticoids is equivalent because of rapid and complete absorption of the oral formulation in the gastrointestinal tract. Intravenous therapy should be converted to oral therapy as soon as possible.

What percentage of patients with brain tumors can achieve complete seizure freedom with levetiracetam?

A. 10%
B. 20%
C. 40%
D. 60%
E. 80%
A large retrospective study showed that complete freedom from seizures could be achieved in 80% of patients with brain tumors who took levetiracetam. In addition to being effective, this drug is less likely to interfere with the efficacy of cancer treatment because it is not metabolized in the liver like the chemotherapeutic agents.

Is it true that aspirin has been shown to be an effective primary preventive agent for ischemic stroke in women but not in men?

A. Yes
B. No
A. Yes.

Aspirin has been shown to be an effective primary preventive agent for ischemic stroke, but only in women. Although aspirin has been shown to reduce the risk for myocardial infarction (MI) in men, there are no data to show that it helps with primary prevention of stroke in men. Recently, the Women’s Health Study found that women who took 100 mg of aspirin every other day had a 17% reduction in overall stroke risk ($P = .04$) as well as a 24% reduction in ischemic stroke ($P = .0009$) without increased risk for hemorrhagic stroke ($P = .31$). Surprisingly, aspirin did not significantly reduce the risk for MI in women.

What is the hallmark electroencephalographic (EEG) pattern of primary generalized epilepsy?

A. Rhythmic, anterior-dominant, generalized, bisynchronous, 3-Hz spike waves superimposed on a normal background
B. Irregular, rapid spike wave activity
C. Arrhythmic asynchronous spike waves
Classically, the presence of rhythmic, anterior-dominant, generalized, bisynchronous, 3-Hz spike-wave discharges superimposed on a normal background is considered to be the EEG hallmark of primary generalized epilepsy. However, the most common EEG abnormality associated with this disorder is the so-called "irregular," "atypical," or "rapid spike" wave activity. This is characterized by generalized paroxysms of spikes or spike wave complexes occurring with an irregular frequency of about 3 to 5 Hz.

All of the following meet clinical diagnostic criteria for neurofibromatosis type 2 (NF2) EXCEPT:

A. Bilateral cranial nerve (CN) VIII masses
B. A first-degree relative with NF2 and a unilateral CN VIII mass
C. A first-degree relative with NF2 and neurofibroma and glioma
D. A first-degree relative with NF2 and meningioma and glioma
E. A first-degree relative with NF2 and juvenile posterior subcapsular lenticular opacity
The clinical diagnostic criteria for NF2 are:

- Bilateral CN VIII masses; or
- A first-degree relative with NF2; and either
  - Unilateral CN VIII mass; or
  - Two of the following: neurofibroma, meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity.

What percentage of patients with tuberous sclerosis complex suffer from seizures?

A. 3%
B. 30%
C. 60%
D. 90%
E. 100%
Seizures are the most common neurologic symptom of tuberous sclerosis complex, occurring in about 90% of patients. Epilepsy in tuberous sclerosis complex often begins during the first year of life and, in most cases, in the very first months. Seizures in infancy present with epileptic spasms and hypsarrhythmia — abnormal, interictal, high-amplitude waves with a background of irregular spikes on EEG.

A 54-year-old man is referred to your clinic for headaches. He has been having headaches since his 40s. The headaches occur 1 to 8 times per day and are unilateral, severe, and localized to the orbit. The pain is a sharp, stabbing pain that is associated with lacrimation and conjunctival injection.

**What is the diagnosis?**

A. A typical migraine  
B. Tension headache  
C. Medication overuse headache  
D. Cluster headache
D. Cluster headache.

Cluster headaches are an uncommon type of headache, occurring in about 0.4% of the population. They are 5 times more common in men than in women. Age of onset is usually in the third or fourth decade of life. They are marked by clustering of headaches with periods of remission and are usually unilateral and sharp in quality. More than 97% of cases include autonomic symptoms. Lacrimation and conjunctival injection are each present in about 80% of cases. Ipsilateral nasal congestion is present in 75%. A partial Horner syndrome with a slight ipsilateral ptosis or miosis or a combination of both is present in 65% of cases.

What is the most common neurologic presentation of neurocysticercosis?

A. Headaches  
B. Seizures  
C. Dementia  
D. Blindness  
E. Hearing loss
Neurocysticercosis is the most common helminthic disease of the nervous system and currently represents a major public health problem. The disease occurs when humans become the intermediate host in the life cycle of *Taenia solium* by ingesting eggs from contaminated food. The clinical presentation is pleomorphic because of individual differences in the number and location of the lesions within the nervous system. Epilepsy is the most common clinical manifestation and occurs in 70% of patients. Seizures are most often generalized tonic-clonic or simple partial.

A 37-year-old woman with no history of medical problems presents to the emergency room with severe headache and blurred vision. She was sick a week before presentation with nausea and emesis. She has had the headache for several days, but the pain worsened last night. Neurologic examination is normal except for mild optic nerve bulging. She is thin and smokes. Her only medication is oral contraception.

What is her diagnosis?

A. Temporal arteritis  
B. Orbital tumor  
C. Cerebral venous thrombosis  
D. Carcinomatous meningitis
C. Cerebral venous thrombosis.

The patient has cerebral vein thrombosis related to clot formation in the cerebral vein or sinus. The occluded vein/sinus leads to venous congestion that can increase intracranial pressure (ICP) and/or regional ischemia. The most frequently occluded venous sinuses are the superior sagittal and the 2 transverse sinuses. Headache is the most common presenting sign, followed closely by paresis.

What is the culprit gene in Hallervorden-Spatz syndrome?

A. PANK1
B. PANK2
C. STANG1
D. STANG2
E. CABO1
Hallervorden-Spatz syndrome is an autosomal recessive disorder characterized by dystonia, parkinsonism, and iron accumulation in the brain. PANK2 encodes a pantothenate kinase, a key regulatory enzyme in the biosynthesis of coenzyme A.

All of the following medications are used to treat restless legs syndrome EXCEPT:

A. Amantadine  
B. Levodopa  
C. Ropinirole  
D. Carbamazepine  
E. Clonazepam
A. Amantadine.

Carbidopa-levodopa and dopamine agonists have been used to improve the sensory symptoms of restless legs syndrome. Clonazepam can alleviate sensory symptoms and periodic limb movements of sleep. Carbamazepine has been shown to be effective in reducing sensory symptoms of restless legs syndrome. Amantadine has not been studied nor is it used in patients with restless legs syndrome.

A 72-year-old man is referred to you for dementia. His wife reports that he has been more confused and forgetful over the past few months. She states that he has started wearing diapers because of urinary incontinence. His examination is notable for a moderate recall deficit at 10 minutes as well as a “magnetic” gait.

What is the most likely diagnosis and work-up?

A. Normal pressure hydrocephalus  
B. Parkinson’s dementia  
C. Dementia with Lewy bodies  
D. Alzheimer’s dementia
A

A. Normal pressure hydrocephalus.

The patient most likely has normal pressure hydrocephalus (NPH). NPH is caused by overproduction of CSF by the ependymal cells of the ventricles. This patient has the classic triad of symptoms of NPH — dementia, gait apraxia, and urinary incontinence. He needs neuroimaging to assess for ventriculomegaly, which may be difficult to distinguish from normal aging for an elderly patient. If imaging is inconclusive, CSF drainage can be performed. Clinical improvement subsequent to CSF drainage is diagnostic of NPH.

Question: Where are the therapeutic deep brain stimulation leads placed for essential tremor?

A. Subthalamic nucleus  
B. Caudate  
C. Thalamus  
D. Globus pallidus  
E. Hypothalamus
C. Thalamus.

The target for deep brain stimulation (DBS) in patients with essential tremor is the thalamus. DBS can be unilateral or bilateral depending upon the patient’s symptoms. DBS has gained wide acceptance and is preferred over thalamotomy due to advantages related to its nonablative and adjustable nature. The specific mechanism of action of DBS in essential tremor remains unknown.

Q

Is the risk for stroke in patients who have migraine higher than in those who do not?

A. Yes
B. No
A. Yes.

The relative risk for stroke in patients who have migraine compared with those who do not was 2.2 (95% CI, 1.7-2.9). The relative risk for transient ischemic attack in migraine patients compared with others was 2.4 (95% CI, 1.8-3.3).

Q

Is skin biopsy a reliable tool for diagnosing small-fiber neuropathy?

A. Yes
B. No
Skin biopsy analysis is a reliable tool for diagnosing small-fiber neuropathy. It is a safe, painless procedure that allows somatic fibers carrying temperature and pain sensation to be differentiated from autonomic fibers. Biopsy of glabrous skin can be performed to examine large sensory fibers in immune-mediated and inherited demyelinating neuropathies. Loss of intraepidermal nerve fibers correlates with increased severity of neuropathy and a higher risk for neuropathic pain. Skin biopsy can be repeated in proximity to a previous biopsy to assess progression of neuropathy and response to neuroprotective treatments.

What is the annual risk for stroke among patients with atrial fibrillation but no history of anticoagulation or cerebrovascular disease?

A. 0.1% to 0.2%
B. 2.5% to 4%
C. 8% to 10%
D. 15% to 20%
Atrial fibrillation is the most common cause of cardioembolic stroke. A meta-analysis showed that the annual risk for stroke in patients without exposure to anticoagulation or history of previous stroke/TIA is between 2.5% and 4%. The risk for stroke in the setting of atrial fibrillation was most significantly elevated in patients with a history of stroke or TIA. Other factors that increase the risk for stroke in patients with atrial fibrillation are advanced age, hypertension, or diabetes.