# Neurology Month Exam Review

A 60-year-old man is evaluated for a **3-year history of gradual, progressive decline in his cognition, behavior, and motor skills**. Both he and his wife have noticed these changes. He becomes agitated easily, and his wife reports that his personality is much more subdued than it used to be. He no longer enjoys socializing. **Memory loss is not a prominent symptom**. The patient played **professional football for 5 years after college graduation**. He has hypertension and hyperlipidemia. His medications are lisinopril, hydrochlorothiazide, and atorvastatin.

On physical examination, vital signs are normal. There is slightly increased tone in the bilateral upper extremities. He has a slow, shuffling gait.

MRI of the brain reveals **global atrophy.** 

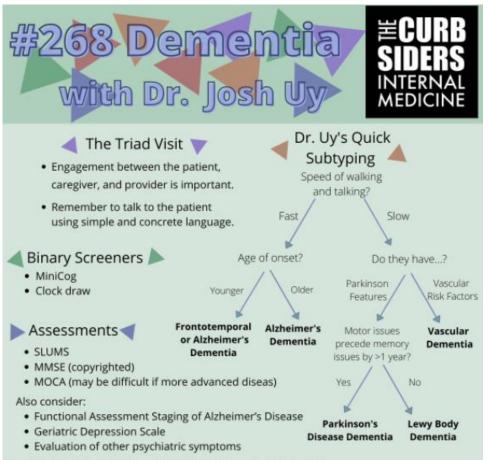
Neuropsychological testing shows marked cognitive slowing and a disorganized thought process.

#### Which of the following is the most likely diagnosis?

- A Alzheimer disease
- B Behavioral-variant frontotemporal dementia
- C Traumatic encephalopathy syndrome
- D Vascular cognitive impairment

# **Key Point**

 The cognitive pattern typical of traumatic encephalopathy syndrome is often one of cognitive slowing and disorganized thought processing, with less involvement of memory and visuospatial function early in the disease course; as the disease progresses, symptoms of mild parkinsonism can occur.



A 73-year-old woman is evaluated for **progressive left arm weakness**. One year earlier, she noted difficulty with **fine movements of her left hand**. Within a few months, her entire **left upper** extremity became stiff and slow, such that she now cannot use this limb. Her left leg has also become rigid. She notes **brief jerky movements of her left arm**. She takes no medication.

On physical examination, vital signs are normal. Language and memory are intact, but she speaks slowly and has difficulty with multistep commands. Results of cranial nerve testing, including extraocular movements, are normal. Muscle strength is preserved throughout. Her left upper and lower extremities are rigid and slow. Her left hand is forced into a bent position and cannot be opened by the examiner. She needs support during ambulation because of stiffness of her left leg. Her sensory perception is preserved, but she cannot identify an object placed into her left hand when her eyes are closed. Cerebellar examination findings, plantar response, and deep tendon reflexes are unremarkable.

#### Which of the following is the most likely diagnosis?

- A Amyotrophic lateral sclerosis
- **B** Corticobasal degeneration
- C Multiple system atrophy
- D Parkinson disease
- E Progressive supranuclear palsy

### Parkinson-plus syndromes

Parkinson-plus syndromes often are more rapidly progressive than Parkinson disease, have additional features that are atypical for idiopathic Parkinson disease, and are less responsive to standard antiparkinsonian therapies, particularly levodopa.

#### Primary Parkinsonism

- · Parkinson's disease
- · Juvenile parkinsonism

#### Secondary Parkinsonism

- Drug-induced
- Stroke
- Infection
- Tumor
- Hypoxia
- Psychogenic
- · Normal pressure hydrocephalus
- Toxins (eg Mn++, CO, MPT)

#### Parkinsonism

#### Parkinson-plus Syndrome

- · Cortico-Basal Ganglionic Degeneration
- · Multiple System Atrophy
- · Progressive Supranuclear Palsy
- · Dementia with Lewy Bodies
- · Primary pallidal atrophy
- · Lytico-bodig

#### Other Neurodegenerative Disorders

- · Alzheimer's disease
- · Spinocerebellar ataxias
- Inherited dystonic syndromes (eg X-linked Dystonia Parkinsonism, Rapid Onset Dystonia Parkinsonism)
- Genetic Metabolic Abnormalities (eg Wilson's disease, Neurodegeneration with Brain Iron Accumulation)
- · Dentato-rubro-pallido-luysian Atrophy
- · Pick's Disease

A 66-year-old woman is evaluated for a 6-month history of slowed gait and difficulty lifting her feet off of the ground when she first starts to walk. She becomes incontinent without feeling the urge to urinate. Her family reports that she seems to be thinking more slowly.

On physical examination, vital signs are normal. Gait is magnetic, with hesitancy and shuffling.

MRI of the brain shows **ventriculomegaly in the lateral ventricles and third ventricle**. There is no obstruction in the cerebral aqueduct.

#### Which of the following is the most appropriate next step in management?

- A High-volume cerebrospinal fluid removal
- B Lumboperitoneal shunt procedure
- C Placement of a temporary lumbar drain
- D Ventriculoperitoneal shunt procedure

# Normal pressure hydrocephalus

- Normal pressure hydrocephalus is characterized by the triad of gait changes, urinary incontinence, and cognitive impairment.
- Normal pressure hydrocephalus cannot be diagnosed without evidence of ventriculomegaly (enlarged cerebral ventricles) on a brain MRI or CT scan of the head; cerebrospinal fluid diversion through a ventriculoperitoneal shunt is the definitive treatment.

A 60-year-old woman comes to the office to discuss the **best strategy to prevent cognitive impairment.** She has no medical problems and takes no medications. She does not use tobacco, has **1 to 2 alcoholic drinks per week, and is sedentary.** 

On physical examination, vital signs and other findings are unremarkable. Results of cognitive screening testing are normal.

#### Which of the following is the most effective preventive measure?

- A Cognitive training
- **B** Donepezil
- **C** Physical exercise training
- D Vitamin E supplementation

# Preventing Cognitive Impairment

- Physical exercise is the most important modifiable lifestyle factor to prevent the onset of cognitive impairment.
- Medications, vitamin, and herbal supplements do not prevent cognitive decline in healthy people or in patients with mild cognitive impairment.

A 78-year-old woman is evaluated for a **1-year history of progressively** worsening memory impairment. She can no longer handle her finances, as she doesn't remember how to fill out checks properly. She lives with her spouse and is independent in all other basic activities of daily living.

On physical examination, vital signs are normal. **Montreal Cognitive Assessment score** is **21/30**, with difficulty copying the cube, recalling the five words, and naming the day, date, and month noted.

Results of laboratory evaluation are normal.

#### Which of the following is the most appropriate imaging test to perform next?

- A Amyloid PET scan
- B Fluorodeoxyglucose PET scan
- C Head CT with contrast
- **D** MRI of the brain without contrast

### Dementia and Imaging

- Either a brain MRI or a head CT without contrast (Option D) should be done in the evaluation of dementia. Guidelines recommend that patients with evidence of dementia symptoms for less than 3 years should have advanced neuroimaging with noncontrast CT or MRI.
- Neuroimaging can identify conditions that may be remediable or modifiable, such as cerebrovascular disease, hemorrhage, tumor, and abscess. Neuroimaging can also support the diagnoses of specific dementias, such as Alzheimer disease, Creutzfeldt-Jakob disease, and hydrocephalus.

A 30-year-old man is evaluated for a 1-hour history of a severe headache. He has had similar headaches for the past 2 weeks that start in the evening, last 2 to 3 hours, and are characterized by intense left-sided pain that is periorbital and piercing. Associated features are photophobia, nausea, and ipsilateral tearing. Nasal sumatriptan, acetaminophen, and ibuprofen have not relieved the pain. He has a 7-pack-year smoking history.

On physical examination, vital signs are within normal limits. Left ptosis and miosis are noted.

Results of laboratory studies and MRI of the brain with contrast are normal.

#### Which of the following is the most appropriate next step in management?

- A Carbamazepine
- B Indomethacin
- C Magnetic resonance angiography of the head and neck
- D Subcutaneous sumatriptan

# Table 9. International Headache Society Criteria for Cluster Headache A. At least five attacks fulfilling criteria B-D B. Severe or very severe unilateral orbital, supraorbital, and/or temporal pain lasting 15-180 minutes (when untreated)

- C. Either or both of the following:
  - a) conjunctival injection and/or lacrimation

b) nasal congestion and/or rhinorrhea

- -

c) eyelid edema

d) forehead and facial sweating

e) miosis and/or ptosis

2. A sense of restlessness or agitation

1. At least one of the following symptoms or signs, ipsilateral to the headache:

- D. Attack frequency from one every other day to eight per day when the disorder is active
- E. Headache not better accounted for by another ICHD-3 diagnosis

ICHD-3 = International Classification of Headache Disorders, 3rd edition.

Adapted with permission of SAGE Publications LTD., London, Los Angeles, New Delhi, Singapore and Washington DC, from Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018 Jan;38(1):41. [29368949] doi:10.1177/0333102417738202.

- Cluster headache and the other trigeminal autonomic cephalalgias are the most severe and stereotypic primary headache disorders and are characterized by trigeminal pain and ipsilateral cranial autonomic features.
- Cluster headache is treated with oxygen inhalation and subcutaneous sumatriptan.

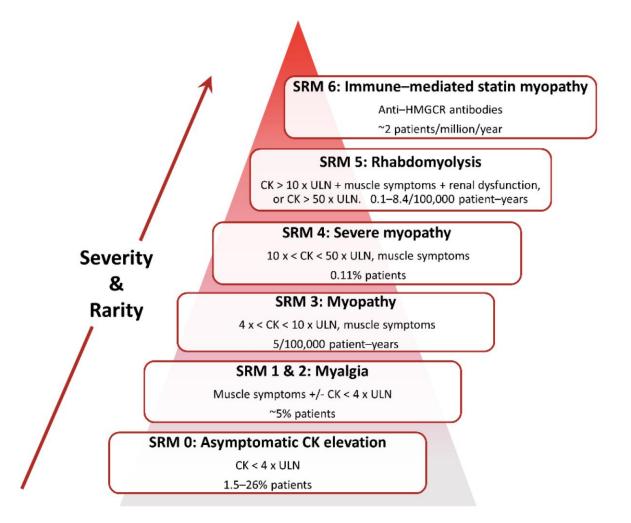
A **63-year-old man** is evaluated for **myalgia and proximal weakness in both upper and lower extremities**. He has diabetes, hypertension, and coronary artery disease. Current medications include **metformin**, **empagliflozin**, **lisinopril**, **and atorvastatin**.

On physical examination, blood pressure is 132/82 mm Hg; other vital signs are normal. **Proximal muscle tenderness** is noted. All other findings, including those from the neurologic examination, are normal.

Laboratory studies show an erythrocyte sedimentation rate of 15 mm/h, hemoglobin A1c level of 6.8%, and serum **creatine kinase level of 250 U/L.** 

#### Which of the following is most likely the cause of this patient's symptoms?

- A Immune-mediated necrotizing myopathy
- B Polymyalgia rheumatica
- C Proximal lumbosacral radiculoneuropathy
- D Statin-induced myopathy



Turner, R. M., & Pirmohamed, M. (2020). Statin-related myotoxicity: a comprehensive review of pharmacokinetic, pharmacogenomic and muscle components. Journal of clinical medicine, 9(1), 22.

An 84-year-old man is evaluated for an 18-month history of **increasing forgetfulness** and one episode of **getting lost while driving** on a familiar route. He lost his car keys and had to have a new set made. He lives with his spouse and is independent in all of his instrumental and basic activities of daily living. He and his spouse report no signs and symptoms of depression. In high school and in college he was on the boxing team. He is otherwise well and takes no medications.

On physical examination, vital signs are normal. His **Montreal Cognitive Assessment score is 21/30**, with difficulty copying the cube, recalling the five words, and naming the day, date, and month noted.

Depression screening is negative.

Brain MRI shows significant hippocampal atrophy bilaterally with minimal white matter hyperintensities.

#### Which of the following is the most likely diagnosis?

- A Alzheimer dementia
- **B** Mild cognitive impairment
- C Traumatic encephalopathy syndrome
- D Vascular cognitive impairment

# Mild Cognitive Impairment

 The formal criteria for MCI are a subjective report (from either the patient or a witness) of a decline in cognitive abilities with relative preservation of day-to-day function and evidence of cognitive impairment on cognitive testing. Because of wide variability in the risk of progression, steps should be taken to confirm the underlying cause of the symptoms.

A 49-year-old woman is evaluated in the emergency department for **sudden-onset severe headache of 24 hours' duration**. The headache **reached its peak intensity within 1 minute**. She is otherwise well and takes no medications.

On physical examination, vital signs are normal. Nuchal rigidity is present. Findings on mental status and funduscopic examinations are normal. There are no cranial nerve deficits.

A CT scan of the head is normal.

#### Which of the following is the most appropriate next step in management?

- A Cerebral catheter angiography
- **B** Lumbar puncture
- C Magnetic resonance venography of the brain
- D Oral sumatriptan

Condition	Features	Diagnostic Testing	
Subarachnoid hemorrhage	Sudden onset/maximal at onset, neck pain (LR 4.1) or stiffness (LR 6.6)	Head CT (w/in 6h), LP if delayed presentation to ED (onset >6h) or low hematocrit (<30)	
Reversible cerebral vasoconstriction syndrome	Headache recurs daily or every few days over several week period. Shorter duration headache than SAH (hours vs days). Look for triggers as a clue: postpartum, autoimmune, sex, drugs, (rock 'n' roll).	Head CTA or MRA (falsely negative imaging in the first week)	
Cerebral venous thrombosis	Often <50y, female, hypercoagulable state, recent postpartum or surgery. Papilledema, AMS, seizures, neuro deficit, elevated ICP	CT venogram or MR venogram	
Cervical artery dissection	Headache + neck pain, recent neck trauma or manipulation, connective tissue disease, HTN	Head and neck CTA	
Posterior reversible encephalopathy (PRES)	Headache, seizures, visual loss, extreme HTN	Brain MRI (w/o)	
Spontaneous intracranial hypotension	Recent spinal procedure, supine positioning improves pain, minor trauma: falls, lifting, coughing	Brain MRI (w/ and w/o)	
Pituitary apoplexy	Usually h/o pituitary adenoma. Consider in pregnancy and dopamine agonist therapy. Ophthalmoplegia, vomiting, visual deficits	Head CT (w/o for acute bleed in the first 2h), brain MRI if negative	Schwedt, T. J., Matharu, M. S., & Dodick, D. W. (2006). Thunderclap headache. The Lancet Neurology, 5(7), 621- 631.
Third ventricle colloid cyst	Lasts seconds to a day. Then rapidly resolves, signs of hydrocephalus or increased ICP	Brain MRI	

### SAH

- Thunderclap headache is a severe headache that reaches maximum intensity within 1 minute and is a medical emergency that warrants immediate evaluation with a head CT scan.
- Patients with suspected subarachnoid hemorrhage but a normal head CT scan require a lumbar puncture.

A 54-year-old man is evaluated for **difficulty swallowing** of 6 months' duration. He initially had occasional choking with liquids, which has progressed to dysphagia with both liquids and solids. His voice has become hoarse. He reports no changes in extremity strength but sometimes finds it hard to lift his head. He takes no medications.

On physical examination, vital signs are normal. Lower facial muscles are weak. Tongue is atrophied and moves slowly; fasciculations are present. Neck flexion and right-hand finger flexion are mildly weak. Jaw jerk and patellar reflexes are brisk, and right ankle clonus is present. Extraocular movements are intact.

Creatine kinase level is 250 U/L.

MRI of the brain and cervical spine and chest radiograph are normal.

#### Which of the following is the most likely diagnosis?

- A Amyotrophic lateral sclerosis
- B Chronic inflammatory demyelinating polyradiculoneuropathy
- C Inclusion body myositis
- D Myasthenia gravis
- E Neuromyelitis optica

### **ALS**

- Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease of motor neurons that causes progressive weakness, atrophy, and early death; 20% of patients with ALS will develop frontotemporal dementia.
- Riluzole and edavarone are FDA-approved treatments for ALS, but they offer only modest benefits in term of survival and functional decline.

A **43-year-old** woman is seen in a follow-up evaluation of myasthenia gravis diagnosed 2 years ago. Current treatment with daily prednisone and pyridostigmine partially controls her symptoms. She is experiencing **weakness with prolonged activity** and in the evening. She wishes to **minimize long-term need for glucocorticoids and other medications** that may suppress her immune system.

On physical examination, vital signs are normal. She has mild bilateral ptosis and impaired left eye adduction and bilateral eye abduction. Speech is nasal. Proximal upper and lower extremity muscle strength and neck flexion are mildly weak.

Acetylcholine receptor—binding antibody is positive.

CT of the chest is normal.

#### Which of the following is the most appropriate treatment?

- A Eculizumab
- B Ipilimumab
- C Plasma exchange
- D Reduction in prednisone dose
- **E** Thymectomy

# MG and Thymectomy

- Thymectomy should be performed in all patients with thymoma and offered as an option to (potentially) avoid or minimize immunotherapy in other patients with active disease who are younger than 65 years and within 3 years of diagnosis.
- Onset of myasthenia gravis (MG) most commonly occurs in the third decade of life in women and after age 50 years in men, with ptosis, diplopia, and fluctuating painless weakness without sensory loss being typical symptoms; all patients with MG should undergo chest CT to screen for thymoma, a tumor associated with the disease.
- Symptomatic treatment of ocular and mild generalized myasthenia gravis usually starts with the cholinesterase inhibitor pyridostigmine; in those with more advanced disease, immunosuppressive therapy is required, as is thymectomy for those with thymoma.

A 70-year-old man is evaluated for a 2-day history of agitation. He has not slept and has been rummaging about the house. He has dementia due to Alzheimer disease. His caregiver reports that he has not started any new medications and has not shown any obvious signs of infection or injury. He has not been constipated and has been able to urinate regularly. The patient is unable to give a symptom history. His only medication is donepezil.

On physical examination, vital signs are normal and other findings are unremarkable.

A complete blood count and measurement of serum electrolytes, serum creatinine, and blood urea nitrogen will be obtained.

#### Which of the following is the most appropriate additional diagnostic test?

- A Arterial blood gas analysis
- B Head CT scan
- C Lumbar puncture with cerebrospinal fluid analysis
- **D** Urinalysis

### Hyperactive delirium

- The leading causes of delirium are fluid and electrolyte disturbances, infection, drug toxicity, metabolic disturbances, and sensory/environmental disturbances.
- The top three classes of drugs most commonly implicated in precipitating delirium are opioids, benzodiazepines, and drugs with anticholinergic properties.

A 60-year-old woman is evaluated for **constant movements of her tongue and lips that she cannot suppress.** She also has difficulty with keeping her jaw closed, and food spills out of her mouth during meals. The movements **started 2 years ago following treatment with metoclopramide for diabetic gastroparesis.** She **stopped the medication**, but movements persisted. Medical history is also significant for panic disorder. She has no history of depression and no family history of abnormal movements or dementia. **Current medications are metformin, sitagliptin, canagliflozin, and clonazepam.** 

On physical examination, vital signs are normal. She displays **frequent**, **fast**, **and variable twitching of the lips and facial muscles and intermittent tongue protrusion**. She cannot keep her mouth open more than a few seconds because of forced closure of the jaw, with associated hyperactivity of the masseter muscles. Minimal writhing finger flexion movements are noted during ambulation. MRI of the brain is unremarkable.

#### Which of the following is the most appropriate treatment?

- A Add aripiprazole
- B Add ropinirole
- C Add valbenazine
- D Begin cognitive behavioral therapy
- E Discontinue clonazepam

### Tardive dyskinesia

- Tardive dyskinesia is characterized by choreiform and dystonic craniofacial movements, which often involve other body parts, such as the neck and trunk -- an extrapyramidal complication of dopamine receptor—blocking medications.
- First step in management should be removal of the offending drug.
- If dyskinesia persists and causes functional or social impairment, treatment with a vesicular monoamine transporter 2 inhibitor, such as valbenazine, deutetrabenazine, or tetrabenazine, can be helpful.
  - Potential adverse effects include sedation, depression, and suicidality; patients should be monitored closely for psychiatric side effects. In trials, valbenazine and deutetrabenazine have shown a lower rate of psychiatric adverse effects compared with tetrabenazine.
  - Other options include amantadine, clonazepam, and, in the case of tardive dystonia, botulinum toxin injection. In refractory cases, deep-brain stimulation can be beneficial.

41-year-old woman is evaluated for a **2-day history of worsening imbalance and clumsiness of the left arm**. She has **relapsing-remitting multiple sclerosis**. Medications are **teriflunomide and a vitamin D supplement**.

On physical examination, vital signs are normal. Neurologic examination shows **new findings of dysmetria and tremor of the left arm.** The patient has **a wide-based, ataxic gait and is unable to perform tandem gait.** 

An MRI of the brain shows multiple periventricular and juxtacortical lesions and one lesion in the left cerebellum. The **cerebellar lesion enhances** with contrast administration.

#### Which of the following is the most appropriate treatment?

- A Intramuscular adrenocorticotropin hormone gel
- **B** Intravenous glucocorticoids
- C Intravenous immunoglobulin
- B Ocrelizumab infusion
- E Plasmapheresis

# Multiple Sclerosis Relapses

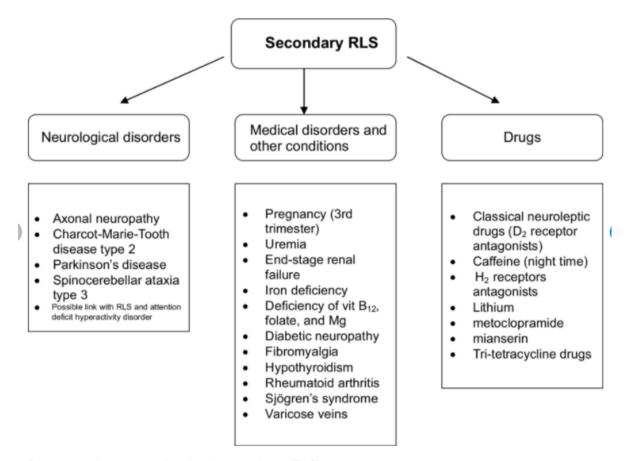
- The first-line treatment for multiple sclerosis relapses is high-dose glucocorticoids, typically administered as intravenous methylprednisolone (1 g/d for 3 to 5 days).
- Intramuscular adrenocorticotropin hormone gel and plasmapheresis are typically reserved as second-line treatments for multiple sclerosis relapses refractory to glucocorticoid treatment.

A 45-year-old woman is evaluated for sleeping difficulties. Her main symptom is an uncomfortable urge to move her legs when she is at rest or in bed at night. This may result in difficulty falling asleep or sometimes in maintaining sleep. She may wake up three to five times a night and then walk around the house for a few minutes to relieve her symptoms before attempting to go back to sleep. Her husband reports no snoring or witnessed apneic episodes or daytime sleepiness.

On physical examination, vital signs and the remainder of the physical examination are normal.

Which of the following is the most appropriate management?

- A Cognitive behavioral therapy for insomnia
- **B** Ferritin measurement
- C Polysomnography
- D Zolpidem



Some secondary causes of restless legs syndrome (RLS).

A **25-year-old woman** is evaluated during a follow-up visit for a generalized tonic-clonic seizure diagnosed yesterday after a comprehensive evaluation. She reports having a similar seizure 1 year ago for which she did not seek medical attention. She also reports that for the past year she has had brief episodes of muscle twitches on awakening that cause her to drop her coffee cup. She has no other medical problems and takes no medications. **She is planning a pregnancy.** 

CT scan of the head without contrast and follow-up MRI obtained yesterday were normal. An electroencephalogram showed **4- to 6-Hz spike-and-wave discharges consistent with generalized epilepsy.** 

Pregnancy test is negative.

#### Which of the following is the most appropriate management?

- A Gabapentin
- **B** Levetiracetam
- C Topiramate
- D Valproic acid

# Juvenile myoclonic epilepsy

- In adults, juvenile myoclonic epilepsy is the most common form of idiopathic generalized epilepsy; associated myoclonic seizures are often called "college seizures" because of the age of onset (teens or twenties) and associated triggers (sleep deprivation, alcohol use, and stress).
- In a woman with childbearing potential, lamotrigine and levetiracetam are the safest antiepileptic drug options.

**17 78-year-old man** is evaluated for **two generalized tonic-clonic seizures** that occurred 2 weeks apart during the past month. Medical history includes hypertension and type 2 diabetes mellitus and a recent diagnosis of mild cognitive impairment. Medications are hydrochlorothiazide and metformin.

On physical examination, vital signs are normal. All other examination findings, including those from a neurologic examination, are unremarkable.

Serum creatinine, electrolytes, and liver chemistry tests are normal. Findings on brain MRI and electroencephalography are normal.

#### Which of the following is the most appropriate treatment?

- A Lamotrigine
- B Oxcarbazepine
- C Valproic acid
- D No seizure medication is necessary

### Seizures and Older Adults

- Lamotrigine, gabapentin, and levetiracetam are better-tolerated and equally effective seizure medications when compared with older antiepileptic drugs in treatment of older adults.
- Incidence of new-onset epilepsy is highest in adults older than 60 years.
- Major risk factors for seizure recurrence include stroke and dementia, but about one-third to one-half of cases are of unknown cause. The diagnosis of seizures in older adults may be difficult because of atypical presentations that may mimic delirium, transient ischemic attack, or syncope. Possible seizures should always be included in the differential diagnosis of older patients with intermittent or fluctuating confusional states of unclear etiology.
- Antiepileptic drug therapy should be started in older adults following two clearly documented unprovoked seizures (e.g., seizures not related to alcohol metabolic derangements, medication, or medication withdrawal).

A 58-year-old man is evaluated for **disabling rhythmic movements involving his upper extremities.** These movements started 8 years ago and now interfere with his job as a chemist. He reports that his handwriting has become illegible. Previous treatment with **propranolol, primidone, and clonazepam** did not improve these symptoms. He also has a history of kidney stones. Family history includes **tremor in his brother and Parkinson disease in his uncle.** He takes no medications.

On physical examination, vital signs are normal. The patient's voice quivers during prolonged vowels. Bilateral upper extremities exhibit a high-amplitude rhythmic movement during the finger-to-nose task and with outstretched position. There are no involuntary movements when hands are placed on the chair arms. Rapid alternative movements, tone, and gait are normal.

#### Which of the following is the most appropriate treatment?

- A Botulinum toxin injection
- **B** Deep-brain stimulation
- C Levodopa
- D Occupational therapy
- E Topiramate

### **Essential Tremor**

- First-line pharmacologic treatments for essential tremor include propranolol, primidone, and topiramate.
- Surgical therapies, including deep-brain stimulation and focused ultrasound thalamotomy, can control tremor in refractory essential tremor associated with functional disability.

Features	Parkinson's tremor	<b>Essential tremor</b>
Tremor	At rest, increases with walking. Decreases with posture holding or action	Posture holding or action
Frequency	3-6 Hz	5-12 Hz
Distribution	Asymmetrical	Symmetrical (mostly)
Body parts	Hands and legs	Hands, head, voice
Writing	Small and Illeligible	Tremulous
Course	Progressive	Stable or slowly progressive
Family History	Less common (1%)	Often (30%-50%)
Other neurological signs	Bradykinesia, rigidity, loss of postural reflexes	None
Substances that improve tremor	Levodopa, anticholinergics	Alcohol, propranolol, primidone
Surgical treatment	Patients usually have other Parkinsonian features requiring subthalamic nucleus or internal globus pallidus deep brain stimulation (DBS)	Thalamic VIM DBS or thalamotomy

A 22-year-old woman is evaluated for a 5-year history of seizures that occur twice monthly, last 2 minutes, and are characterized by staring, lip smacking, and confusion. These seizures have continued despite treatment with two adequately dosed antiepileptic medications (carbamazepine, followed by levetiracetam, both of which have been discontinued owing to insufficient ability to control seizures). Her current medication is lamotrigine at maximum dose, which she tolerates well with no adverse effects.

On physical examination, vital signs are normal. All other examination findings, including those from a neurologic examination, are unremarkable.

Results of outpatient routine electroencephalography (EEG) are normal.

#### Which of the following is the next most appropriate management?

- A Phenytoin
- B Temporal lobectomy
- C Vagus nerve stimulator
- D Video EEG monitoring

# Drug-resistant epilepsy

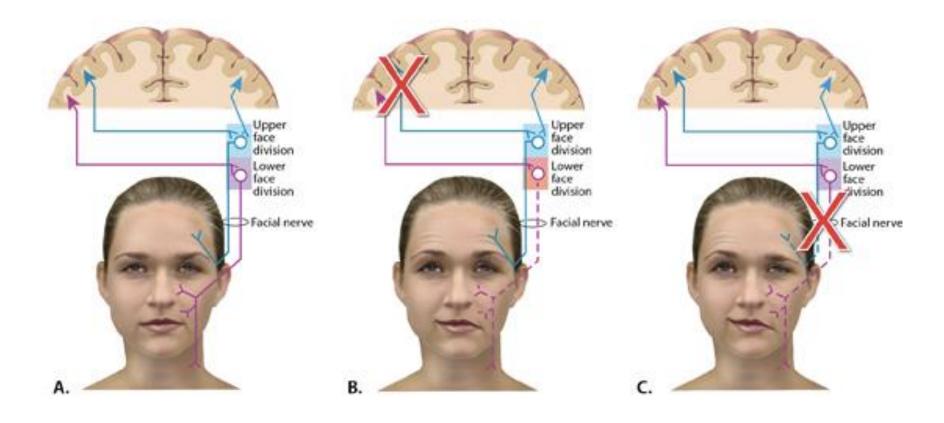
- Drug-resistant epilepsy is defined as having ongoing seizures despite treatment with two tolerated, appropriately chosen, and adequately dosed antiepileptic medications.
- Video electroencephalography is the first step in determining candidacy for epilepsy surgery in patients with drug-resistant epilepsy followed by advanced brain imaging.

A 36-year-old woman is evaluated for left-sided facial weakness. Over the last **10 days she has developed progressive weakness** on the left side of her face, which is apparent with eye closure, smiling, and chewing. She has also reduced tearing and diplopia. She is otherwise well and takes no medications.

On physical examination, vital signs are normal. Left upper and lower facial muscles and muscles of mastication are weak. She has difficulty closing her left eye and diminished leftward gaze. Left corneal reflex is impaired. Facial sensation is preserved. The rest of the neurologic examination is unremarkable.

#### Which of the following is the most appropriate next step in management?

- A Electromyography of face muscles
- B MRI of brain
- C Oral acyclovir
- D Prednisone
- E No further testing or intervention



- Atypical Bell palsy, including subacute onset or involvement of multiple facial nerves, requires additional evaluation with imaging and other laboratory testing.
- Classic Bell palsy, characterized by the acute onset of a peripheral facial nerve neuropathy, is a clinical diagnosis requiring no additional evaluation.
- The most appropriate next step in management is MRI of the brain .