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# Neurology Test Review

# Question 1.

- B; Measurement of serum 25-hydroxyvitamin D level

# Diagnose vitamin D deficiency in a patient with multiple sclerosis

## Presenting signs and symptoms

- **Optic neuritis**
  - Subacute visual deficit in one eye
  - Painful eye movement
  - Afferent pupil defect
- **Myelitis**
  - Sensory/Motor symptoms with spinal level
  - Lhermitte sign
- **Brain stem/Cerebellar**
  - Ataxia/Vertigo
  - Dysconjugate eye movements/nystagmus/INO
- **Cognitive Dysfunction/Fatigue**
- **Uthoff phenomenon**
  - Transient worsening of baseline symptoms with elevated body temperature

## Diagnostic Criteria

McDonald Criteria:

Evidence of CNS Demyelination disseminated in space and time

- MRI findings (5)
- Electrophysiologic Testing
  - Visual evoked potentials
  - Auditory evoked potentials
  - Somatosensory evoked potentials
  - Optical coherence tomography
- Cerebrospinal fluid analysis
  - Oligoclonal bands (85-90%)
  - IgG index

## Importance of Vitamin D in MS

- **Reduced serum vitamin D levels predict future accumulation of new lesions on MRI**
- **Various factors reduce bone mineral density in patients with MS by 3-6x**
- Smoking cessation is also strongly encouraged in patients with MS to reduce conversion to secondary progressive disease.

## Question 2.

- B; High-dose methylprednisolone

# Treat acute spinal cord injury

## Presenting signs and symptoms

- Neck or back pain
- Weakness, sensory changes, bowel and bladder dysfunction
- Corticospinal tracts affected:
  - Spastic paresis/paralysis
  - Hyperreflexia
  - Extensor plantar response
  - Sensory level
- Cauda equine syndrome (distal cord/lower roots)
  - Flaccid paralysis
  - Areflexia
  - Loss of perianal sensation

## Diagnosis

- MRI cord compression protocol immediately
- CT myelography if MRI contraindicated (uses contrast)



## Treatment

- High dose intravenous administration of glucocorticoids within the first 8 hours of traumatic spinal cord injury improves outcomes
- Urgent surgical decompression followed by steroids and radiation therapy is typically required for metastatic cord compression
- Radiosensitive tumors such as leukemia, lymphoma, myeloma, and germ cell tumors may respond to urgent radiation alone

## Question 3.

- A; Lumbar puncture

# Diagnose a subarachnoid hemorrhage with lumbar puncture

## Presenting signs and symptoms

- Sudden onset severe headache “thunderclap headache”
- Nausea and vomiting
- Meningismus
- Loss of consciousness
- Seizure <10%
- Sudden death

## Diagnosis

- Non contrast CT scan of brain
  - Sensitive highest 6-8 hours after bleed (95%) but declines to 58% after 5 days
- Lumbar puncture
  - Elevated opening pressure
  - Elevated RBC count that does not decrease from tube 1 to tube 4
  - Xanthochromia indicates blood has been in CSF for 2 hours
  - Xanthochromia can be seen with:
    - Elevated CSF protein > 150 mg/dL
    - Elevated serum bilirubin >10
    - Elevated CSF RBC >100,000

## Missed Diagnosis

- Failure to appreciate the clinical spectrum of presentation
- Failure to obtain a head CT or understand its limitations
- Failure to perform a lumbar puncture and/or correctly interpret the results

# Question 4.

- B; Large volume lumbar puncture



# Diagnose normal pressure hydrocephalus

## Presenting signs and symptoms

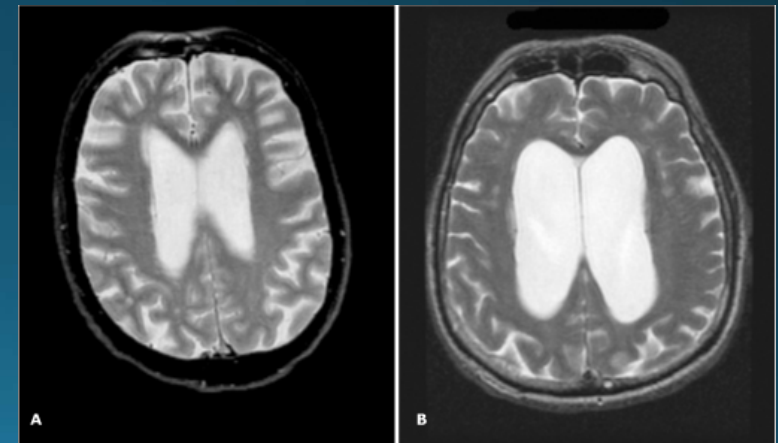
- Gait difficulty “magnetic gait” as if the feet are stuck to the floor
- Cognitive disturbance of psychomotor slowing, decreased attention and concentration, and apathy (decrease in executive function)
- Urinary urgency precedes urinary incontinence
- Spasticity, postural tremor and rigidity

## Diagnosis

- MRI of the brain preferred imaging
  - Ventriculomegaly without sulci enlargement
  - White matter signal abnormality
  - Aqueduct flow void
- Neuropsychological testing
- **Large volume lumbar puncture**

## Treatment

- Ventricular shunting
- Repeat large volume lumbar puncture



# Question 5.

- A; Carotid artery dissection

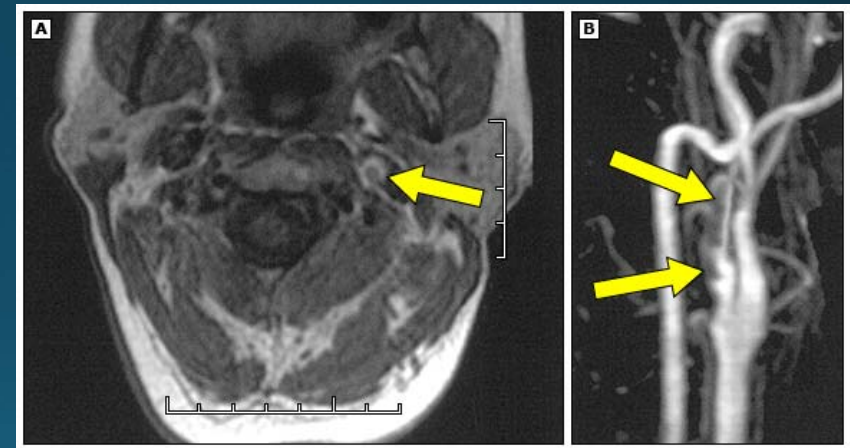
# Diagnose carotid artery dissection

## Presenting signs and symptoms

- Head and neck pain (thunderclap in 20%) 80% gradual
- Partial Horner's syndrome (25%)
  - Ptosis
  - Miosis
  - Anhydrosis
- Isolated orbital or periocular pain (rare)
- Ischemia (TIA or stroke)
- Retinal artery occlusion or ischemic optic neuropathy

## Diagnosis

- Carotid duplex
  - 68-95% sensitive
  - Misses lesions near skull base and vertebral artery dissections within the transverse foramina
- MRA or CTA
  - Crescent sign of intramural hematoma
  - String sign
- Conventional angiography is the gold standard and should be performed if non-invasive studies are negative with high suspicion



# Question 6.

- B; Normalization of blood pressure

# Treat reversible cerebral vasoconstriction syndrome (RVCS)

## Presenting signs and symptoms

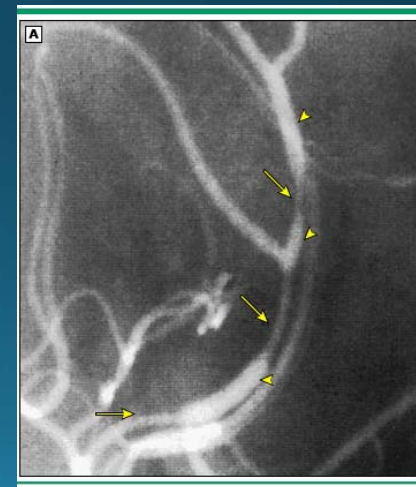
- Sudden, excruciating thunderclap headaches triggered by physical exertion, coughing, sneezing that are recurrent over days to weeks
- Headaches are diffuse or vertex, associated with nausea and vomiting, photophobia
- 1/3 of patients develop strokes
- Risk factors: pregnancy, migraine, vasoconstrictive drugs, anti-migraine drugs, tacrolimus, cyclophosphamide, and illicit drugs

## Diagnosis

- Recurrent acute, severe headaches with or without neurologic signs/symptoms
- Initial brain imaging is normal in 30-70% of patients, despite diffuse vasoconstriction
- No aneurysmal subarachnoid hemorrhage
- Normal or near-normal CSF analysis
- Angiography by fluoroscopy, CT, or MRA should reveal vasoconstriction
- Reversibility of angiographic abnormalities documented within 12 weeks

## Management

- Opioids for pain
- No vasodilators helpful
- Usually resolves in weeks spontaneously



# Question 7.

- A; Aspirin

# Treat ischemic stroke in a patient with atrial fibrillation

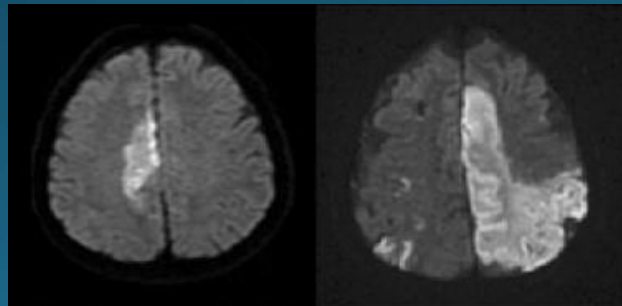
## Presenting signs and symptoms

Clues that a stroke is cardioembolic in origin:

- Involvement of the cortical surface in the absence of cerebral artery atherosclerosis (80% anterior, 20% posterior circulation)
- Presence of more than one infarct, especially if bilateral or involving the territory of both the internal carotid and vertebrobasilar arteries
- Involvement of an entire territory of a large intracranial artery
- EKG or telemetry monitoring showing paroxysmal, persistent, or permanent afib

## Diagnosis

- EKG and telemetry monitoring (may require monitor for up to 30 days)
- Transthoracic echocardiogram
- Transesophageal echocardiogram on a selective basis



## • Treatment

- Acute stroke within the window for lytics and no contraindication should receive the lytics
- **If no lytics advised, BRIDGING therapy with aspirin/antiplatelet agents until discharge from the hospital**
- Avoid full dose anticoagulation acutely, especially if the stroke is  $> 1/3$  of the MCA distribution, and if there is no petechiae on neuroimaging

# Question 8.

- C; Surgical resection of the brain lesion



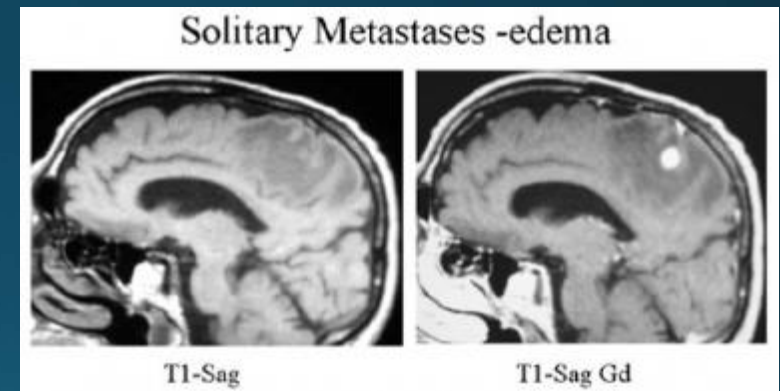
# Treat a solitary metastatic brain tumor

Predictors of outcome with metastatic brain lesions

- Karnofsky performance score
  - 0-40 Dead to bedbound
  - 50-70 Special care required to home bound but cares for self
  - 80-100 Cares for self to Normal
- Type of cancer
  - Non-small cell lung CA
  - Melanoma
  - Breast
  - Renal
  - Gastrointestinal
- Age of patient
- Extracranial metastases

## Management

- Resect lesion by surgery or stereotactic radiosurgery with following whole brain radiation
- Best candidates are:
  - KFS > 70
  - < 3 metastases
  - Age < 65 yo
  - Controlled primary tumor
  - No extracranial metastases
- Median survival 7.1 months



# Question 9.

- D; Medication overuse headache

# Diagnose medication overuse headache

## Presenting signs and symptoms

- Headache that occurs  $\geq 15$  days per month in a patient with a pre-existing headache disorder
- Regular intake  $\geq 10$  days/month for  $> 3$  months of ergotamines, triptans, NSAIDs, opioids, or combination medications
- Regular intake  $\geq 15$  days/month for  $> 3$  months of simple analgesic (acetaminophen, aspirin, or NSAID)

## Risks of medication induced MOH

- HIGH
  - Opioids, Butalbital-containing analgesics, and aspirin/acetaminophen/caffeine combinations
- INTERMEDIATE
  - Triptans
- LOW
  - NSAIDs

## Treatment

- Outpatient:
  - Stop overused medication cold-turkey
  - Bridge with NSAIDs or prednisone or clonidine if opioid
  - Start a preventative medication
  - (NOT appropriate for butalbital users; must use phenobarbital wean)
- Inpatient:
  - Stop overused medication cold-turkey or wean opioids
  - Bridge with methylprednisolone, dihydroergotamine, clonidine
  - Start prevention medication

# Question 10.

- D; No pharmacologic treatment is required

# Manage hypertension in a patient with an acute ischemic stroke

## Stroke prevention

- Relative Risk Reduction
  - 50% RR in heart failure
  - 30-40% RR in stroke
  - 25% RR in MI
- Absolute Risk Reduction
  - BP therapy for 4-5 years will prevent a complication in 2 out of 100 patients
  - Probable underestimate of benefit

## Acute ischemic stroke *with* TPA

- Get blood pressure **< 185/110** to give lytics
- Keep blood pressure **< 180/105** for 24 hours after lytics
- **MAP = DP + 1/3 (SBP-DBP)**

## Acute ischemic stroke *without* TPA

- **Get blood pressure < 220/120 without lytics**
- **Treat blood pressure if patient is having hypertensive emergency such as aortic dissection, AKI, hypertensive encephalopathy, ACS, or acute heart failure**
- **Reduce blood pressure only by 15% in the first 24 hour (MAP 150 to 130)**

# Question 11.

- C; MRI of the brain

# Evaluate facial nerve palsy with incomplete $\gamma$ after 3 months

## Presenting signs and symptoms:

- Complete unilateral facial paralysis over hours to days
- Spares the forehead
- Hyperacusis and impaired taste may occur



## Differential Diagnosis:

- Herpes virus
- Lyme disease
- HIV infection
- Sarcoid
- Stroke
- Sjogren's disease
- Tumor

## Treatment:

- Oral prednisone within the first 72 hours
- Antiviral therapy alone has no benefit to shorten or lessen the severity
- Artificial tears and nighttime eye patch
- Complete recovery in 70-90% of patients within weeks
- If symptoms worsen or do not improve by 2 months, will need MRI to look and serologies to look for secondary causes

## Question 12.

- C; Discontinue combined oral contraceptive



# Treat migraine with typical aura

## Presenting signs and symptoms

- Aura (25% of migraines)
  - Usually 5 minute onset and precedes or concomitant with headache
  - Visual/sensory/speech (first positive then negative symptoms)
- Headache
  - Unilateral, throbbing, photophobia/phonophobia/ nausea/vomiting
- Precipitating factors
  - Stress, hormonal fluctuations, fasting, sleep disturbance, etc.

## Diagnosis of Migraine with Aura

- B) One or more of the following fully reversible aura symptoms:
- Visual
  - Sensory
  - Speech and/or language
  - Motor
  - Brainstem
  - Retinal
- C) At least two of the following four characteristics:
- At least one aura symptom spreads gradually over  $\geq 5$  minutes, and/or two or more symptoms occur in succession
  - Each individual aura symptom lasts 5 to 60 minutes
  - At least one aura symptom is unilateral
  - The aura is accompanied, or followed within 60 minutes, by headache

## Treatment

- Migraine with aura is the second greatest risk factor for stroke in women after hypertension, surpassing diabetes.
- Oral contraceptives containing estrogen are contraindicated in women with migraine with aura. Progesterone-only contraception is okay.
- Basilar and hemiplegic migraines are contraindications for triptan therapy

# Question 13.

- A; Intravenous labetalol

# Treat hypertension after an intracerebral hemorrhage

## Types of Intracerebral Hemorrhages:

- Epidural
  - Head trauma/ skull fx: 75-95%
  - Middle meningeal artery trauma
  - "lucid interval"
- Subdural
  - Head trauma
  - Acute vs. chronic
  - Bridging veins but arterial 20-30%
- Subarachnoid
  - Spontaneous "the worst headache of my life"
  - Aneurysm rupture/AVMs
  - May need to repeat CTA if first one negative
- Intraparenchymal
  - Hypertensive, AVM, amyloid angiopathy
  - Thalamic, Basal ganglia, Cortex. Pons

**Table 6. Suggested Recommended Guidelines for Treating Elevated BP in Spontaneous ICH**

1. If SBP is  $>200$  mm Hg or MAP is  $>150$  mm Hg, then consider aggressive reduction of BP with continuous intravenous infusion, with frequent BP monitoring every 5 min.
2. If SBP is  $>180$  mm Hg or MAP is  $>130$  mm Hg and there is the possibility of elevated ICP, then consider monitoring ICP and reducing BP using intermittent or continuous intravenous medications while maintaining a cerebral perfusion pressure  $\geq 60$  mm Hg.
3. If SBP is  $>180$  mm Hg or MAP is  $>130$  mm Hg and there is not evidence of elevated ICP, then consider a modest reduction of BP (eg, MAP of 110 mm Hg or target BP of 160/90 mm Hg) using intermittent or continuous intravenous medications to control BP and clinically reexamine the patient every 15 min.

Note that these recommendations are Class C. SBP indicates systolic blood pressure; MAP, mean arterial pressure.

# Question 14.

- A; Decompression surgery

# Treat carpal tunnel syndrome

## Presenting signs and symptoms

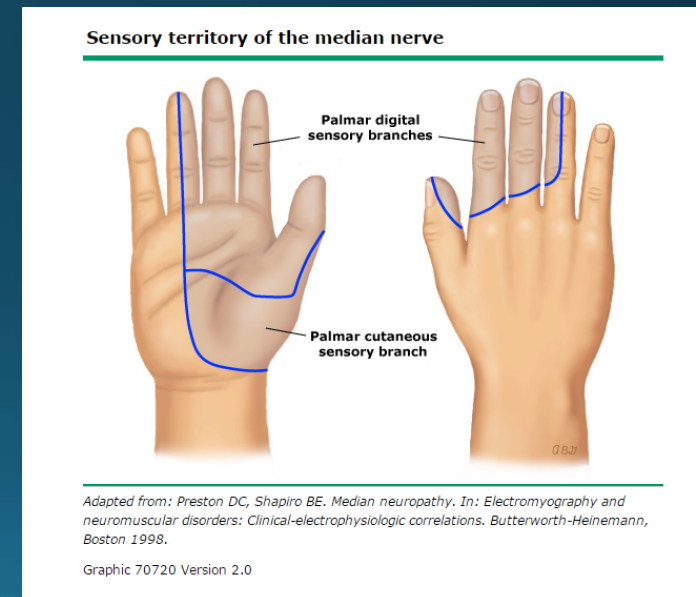
- Dull, aching, numbness, paresthesias, weakness or clumsiness in the medial nerve distribution
- Worsened by repetitive movements of the wrist or sleep
- Mitigated by changes in hand posture or shaking of the hand
- Electrodiagnostic testing is used for diagnosis (in uncertain cases) and to gauge severity and prognosis (and timing of surgery)

## Indications for surgery

- Persistent numbness or pain
- Motor dysfunction with diminished grasp or pinch grip
- Thenar eminence flattening

## Conservative Management

- Wrist splinting, especially during sleep
- Glucocorticoid injection



# Question 15.

- B; Chronic inflammatory polyneuropathy

# Diagnose chronic inflammatory demyelinating polyradiculoneuropathy

## Presenting signs and symptoms

- Symmetric proximal and distal weakness
- Vibration and position symmetric sensory impairment > temperature and pain sensory impairment > autonomic
- Diminished reflexes
- **Slow, progressive decline or relapsing-remitting over 2 months to years**

## Diagnosis

- **Electrodiagnostic or pathologic evidence of demyelination**
- Elevated protein in CSF seen in 90% of patients
- CSF WBC > 10 suggests an alternative diagnosis
- Nerve biopsy in atypical cases to exclude amyloidosis, sarcoidosis or vasculitis

## Treatment

- IVIG (contraindicated in CKD)
- Glucocorticoids
- Plasma exchange
- Immune modulators (steroid sparing)

# Question 16.

- C; Evaluate for concurrent illness



# Identify the cause of delirium in a patient with dementia

## Confusion assessment method (CAM) for the diagnosis of delirium\*

Feature	Assessment
1. Acute onset and fluctuating course	Usually obtained from a family member or nurse and shown by positive responses to the following questions: "Is there evidence of an acute change in mental status from the patient's baseline?"; "Did the abnormal behavior fluctuate during the day, that is, tend to come and go, or increase and decrease in severity?"
2. Inattention	Shown by a positive response to the following: "Did the patient have difficulty focusing attention, for example, being easily distractible or having difficulty keeping track of what was being said?"
3. Disorganized thinking	Shown by a positive response to the following: "Was the patient's thinking disorganized or incoherent, such as rambling or irrelevant conversation, unclear or illogical flow of ideas, or unpredictable switching from subject to subject?"
4. Altered level of consciousness	Shown by any answer other than "alert" to the following: "Overall, how would you rate this patient's level of consciousness?" Normal = alert Hyperalert = vigilant Drowsy, easily aroused = lethargic Difficult to arouse = stupor Unarousable = coma

\*The diagnosis of delirium requires the presence of features 1 AND 2 plus either 3 OR 4.

## ACUTE AGITATED CONFUSION IN AN OLDER PATIENT

### PRESCRIBING GUIDELINE

#### Look for possible precipitants

Metabolic problems - sodium, calcium, hypoxia, hypoglycaemia?	Is your patient in pain?
Is there infection in chest, urine, skin, joints, or meninges?	Is alcohol withdrawal a possibility?
Is benzodiazepine withdrawal a possibility?	Is urinary retention a possibility?

**Drugs** - be suspicious of all prescribed drugs and check that none have been suddenly stopped.

#### Can you modify the environment?

One to one nursing - discuss extra staff with the directorate manager.	Try to find a quiet, well lit, side room.
Can family stay with the patient for some of the time?	Provide an understanding nurse.

Is your patient too hot, too cold, or hungry?

#### Drug Treatment

**N.B. Only use drugs if your patient is at risk of causing harm to themselves or others.**

If alcohol or benzodiazepine withdrawal is a possibility refer to the alcohol withdrawal guideline.

In other cases use:

1. Haloperidol 0.5-1mg orally if possible Wait 20 mins at least
2. If no response 0.5-2mg orally or IM Wait 20 mins at least repeat Haloperidol
3. If no response discuss with a senior member of your team
4. If agitation remains an acute problem discuss with on-call psychiatric staff. (Out of hours contact via REH switchboard on Ext.7600)

An alternative to haloperidol in patients in whom this is unsuitable (eg, Parkinson's disease, dementia with Lewy Bodies) is lorazepam 0.5mg orally or im, using same regime as for haloperidol. Use as little as possible: benzodiazepines prolong delirium and may be associated with a worse outcome.

#### REMEMBER

This is a general guideline - your patients have *individual* problems

Seek and treat participants      Try to modify the environment

Give drugs time to work

Table 4 Standard investigations in delirium. In many situations in a neuroscience unit the likely cause will be known and investigations will not be required. The list below is suggested as a 'standard' screening battery only

Exclude hypoglycaemia and hypoxia at the bedside.  
 Urine and electrolytes, Ca  
 Liver function tests  
 Full blood count  
 Erythrocyte sedimentation rate and C reactive protein  
 Troponin  
 Glucose  
 Blood cultures if any evidence of infection  
 Urinalysis ± MSU  
 Chest x-ray  
 ECG  
 Consider CT ± lumbar puncture if delirium persist without known precipitant  
 Further investigations will be indicated depending on underlying neurological state

Carson, Alan; Ryan, Tracy. Practical-Neurology 2010; 10:67-81

# Question 17.

- C; Naproxen

# Treat migraine without aura with an NSAID

## Presenting signs and symptoms

- A. At least 5 attacks fulfilling criteria B-D
- B. Headache attacks lasting **4-72 hours** (untreated or treated)
- C. Headache with at least two of the following 4 characteristics:
  - **Unilateral location**
  - **Pulsating quality**
  - **Moderate or severe pain that inhibits or prohibits daily activity**
  - **Aggravated by physical activity**
- D. During the headache, occurrence of at least one of the following symptoms:
  - **Nausea/vomiting**
  - **Photophobia/Phonophobia**
- E. Not better accounted for by another diagnosis

## Treatment

- Goal of therapy to resolve migraine and return the patient to normal function within 1-2 hours of treatment
- Pt to remain free of headache for 24 hours
- **NSAID**, triptan, dihydroergotamine (DHE)
- **NSAIDS are preferred as first line because of cost-effectiveness, and are most effective for mild-moderate migraine**
- **Administer rescue medication within 1 hour of onset**
- Refractory migraine: >72 hours; short course of glucocorticoids, IV DHE, IV prochlorperazine with diphenhydramine, ketoralac

# Question 18.

- C; Smoking cessation counselling

# Counsel a patient with an unruptured cerebral aneurysm about smoking cessation

## Risk factors for cerebral aneurysms

- Hereditary
  - Elers-Danlos
  - Pseudoxanthoma elasticum
  - Autosomal dominant polycystic kidney disease (6X RR)
- Familial
- **Cigarette smoking (3-5X RR)**
- Hypertension
- Estrogen deficiency
- Coarctation of the aorta

## 5 year: risk of aneurysm rupture

- Location
  - LOW : Cavernous carotid artery
  - INTERMEDIATE: Anterior circulation
  - HIGH: Posterior circulation
- Size > 7 mm increases risk of rupture
  - 7-12 mm (2.6%)
  - 13-24 mm (14.5 %)
  - >25 mm (40%)

## Management of unruptured cerebral aneurysm

- **Avoid smoking**
- Avoid alcohol
- Avoid stimulants and illicit drugs
- Avoid excessive straining and Valsalva maneuvers

**Consider treatment for asymptomatic aneurysms 7-10 mm**

# Question 19.

- D; No further testing

# Diagnose early Parkinson disease

## Presenting signs and symptoms

- Resting tremor
- Bradykinesia
- Cogwheel rigidity
- Gait/Postural impairment
  
- Cognitive slowing
- Depression, anxiety, apathy, psychosis
- Sleep fragmentation, REM sleep disorder
- Autonomic dysfunction
- Dystonia

## Diagnosis of Parkinsons Disease

- Presence of bradykinesia with at least one other cardinal feature in the absence of red flags for atypical forms of Parkinsonism

Disorders that can mimic Parkinson disease
<b>Neurodegenerative causes</b>
Alzheimer disease
Corticobasal degeneration
Dementia with Lewy bodies
Frontotemporal dementia
Huntington disease
Multiple system atrophy
Parkinsonism-dementia-ALS complex of Guam
Progressive supranuclear palsy
Spinocerebellar ataxias

## Red Flags for Atypical Disease

- Severe dysautonomia
- Abnormal eye movements
- Prominent early cognitive impairment
- Visual hallucinations
- Early prominent postural instability and falls
- Prominent ataxia or cerebellar deficit
- Symmetric or extremely asymmetric involvement
- Rapid and stepwise deterioration rather than gradual



# Question 20.

- C; Dementia with Lewy Bodies

# Diagnose dementia with Lewy bodies

Clinical and radiologic features of dementia with Lewy bodies (DLB)	
	Frequency in DLB (percent)*
<b>Central feature (essential for the diagnosis) <sup>¶</sup></b>	
Progressive cognitive decline, dementia	100
<b>Core features (two features essential for diagnosis of probable DLB, one for possible DLB) <sup>¶</sup></b>	
Fluctuating cognition	60-80
Recurrent well-formed, detailed visual hallucinations	50-75
Spontaneous features of parkinsonism	80-90
<b>Suggestive features (one suggestive feature with one core feature may diagnose probable DLB, one or more suggestive features may diagnose possible DLB) <sup>¶</sup></b>	
REM sleep disorder	85
Severe neuroleptic sensitivity	30-50
Low dopamine transporter uptake in basal ganglia on SPECT or PET	
<b>Supportive features (common features with undetermined diagnostic specificity) <sup>¶</sup></b>	
Repeated falls	33
Syncope or transient loss of consciousness	
Severe autonomic dysfunction	
Hallucinations in other modalities	20
Systematized delusions	55-75
Depression	30-40
Relative preservation of medial temporal lobe on MRI or CT	
Generalized low uptake on SPECT or PET perfusion imaging with reduced occipital activity	
Abnormal (low uptake) MIBG myocardial scintigraphy	
Prominent slow wave activity and temporal lobe transient sharp waves on EEG	
<b>Conflicting features (features which make DLB less likely) <sup>¶</sup></b>	
Cerebrovascular disease evidenced by focal neurologic signs or neuroimaging	
Other physical illness or brain disorder which is consistent with some or all of clinical features	
First appearance of parkinsonism at late stage (severe) dementia	
<b>Temporal sequence (feature which distinguishes DLB from Parkinson disease dementia) <sup>¶</sup></b>	
Dementia should occur before or concurrently with onset of parkinsonism	

# Holiday Fun

- 11:15-11:45
- Groups 1 and 2 Gingerbread House Creation Contest
- Groups 3 and 4 Holiday Card making Stations
  
- 11:45-12:15
- Switch!!
  
- 12:15 Gingerbread House Winner Announced!