

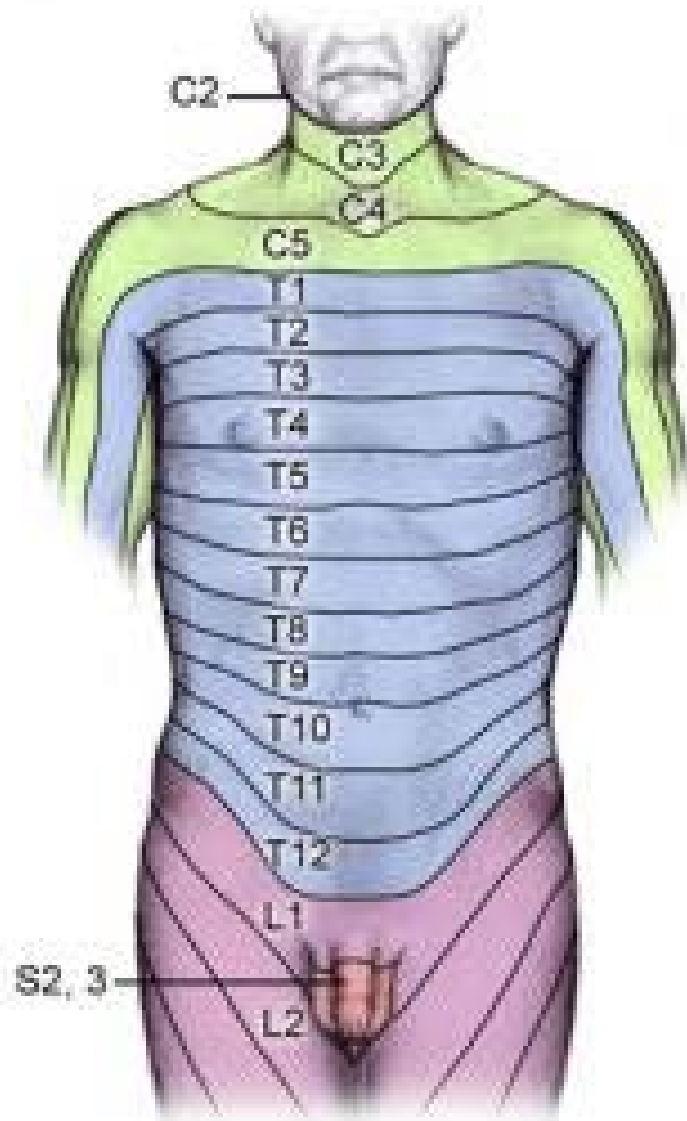
Neurology MKSAP Test 2017

1

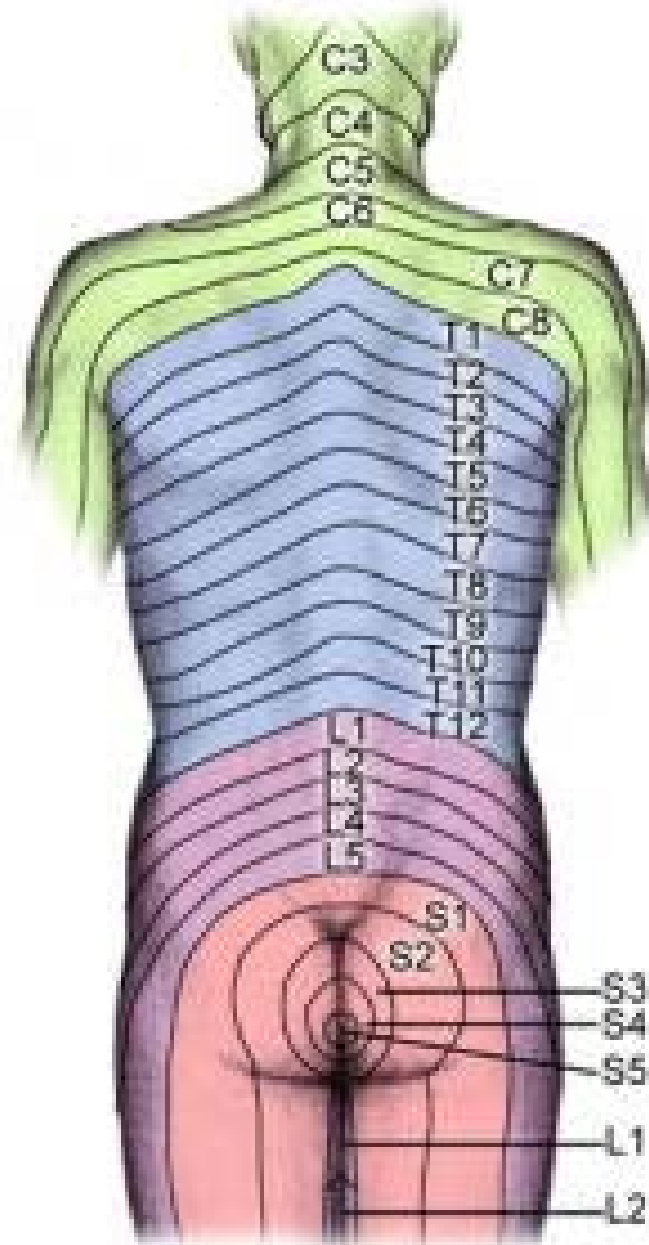
In addition to a 5-day infusion of intravenous methylprednisolone, which of the following is the most appropriate next step in management?

- a. Discontinuation of natalizumab
- b. Measurement of serum 25-OH vitamin D level
- c. MRI of lumbar spine
- d. Oral TMP-SMX for 5 days

Anterior view



Posterior view



Vit D Deficiency in MS

- <30 = insufficient
- <20 = deficiency

- Adjunctive Vit D tx in MS superior to disease-modifying therapy alone, even if NOT deficient
- MS patients have increased risk of osteoporosis

Golan D, Halhal B, Glass-Marmor L, et al. Vitamin D supplementation for patients with multiple sclerosis treated with interferon-beta: a randomized controlled trial assessing the effect on flu-like symptoms and immunomodulatory properties. BMC Neurol. 2013 Jun 14;13:60

MS Meds

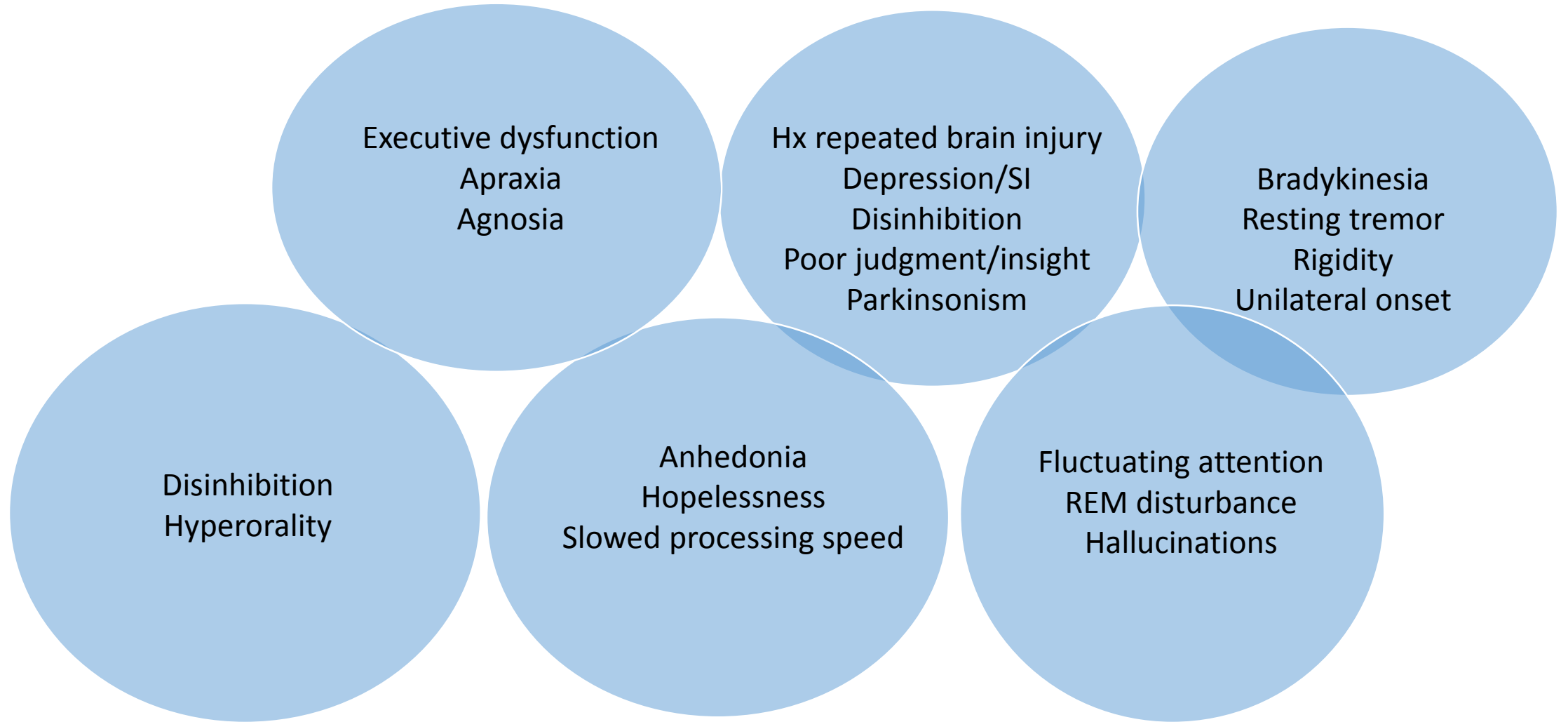
- Control the disease
 - Steroids
 - Biologics
 - Vitamin D
- Control disease-related symptoms
 - Fatigue Amantadine
 - Spasticity Dalfampyridine
 - Urinary incontinence Oxybutynin
 - Depression Bupropion
 - Osteoporosis Vitamin D
- Control side effects of disease/meds

7

Which of the following is the most likely diagnosis?

- a. Chronic traumatic encephalopathy
- b. Dementia with Lewy bodies
- c. Depression-related cognitive impairment
- d. Parkinson disease

Selected causes of Memory Impairment



Sensitivity and specificity of diagnostic tests for dementia

Diagnostic test	Sensitivity (percent)	Specificity (percent)
Mini-Mental State Exam*	87	82
Montreal Cognitive Assessment	≥94	≤60
Short Portable Mental Status Questionnaire*		
- Any dementia	82	92
- Mild dementia	55	96
NINCDS criteria¶	92	65
DSM-IV criteria¶	76	80
Clinical judgment¶	85	82

NINCDS: National Institute of Neurological and Communicative Diseases; DSM-IV: Diagnostic and Statistical Manual of Mental Disorders, 4th ed.

* Diagnosis of dementia.

¶ Diagnosis of Alzheimer disease.

12

Which of the following is the most appropriate next diagnostic test?

- a. LP
- b. MRA brain
- c. MRV brain
- d. MRI brain

Unilateral Dilated Unreactive Pupil

- Suggests external compression of the superficial pupil fibers of the ipsilateral oculomotor (III) nerve in the subarachnoid space
 - Tumor
 - Aneurysm

 - Cerebral herniation
 - ICP

26

Which of the following is the most appropriate next step in treatment?

- a. Chemotherapy
- b. Continued high-dose glucocorticoids only
- c. XRT
- d. Surgery

- Plasmacytoma and myeloma respond very well to radiation
- Nuance here is spinal stability, only mild neuro deficits



The right answer
will always stand
alone

Tsutsumi S, Yasumoto Y, Ito M. Solitary spinal extradural plasmacytoma: a case report and literature review. Clin Neuroradiol. 2013 Mar;23(1):5-9.

30

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

- a. Glucose tolerance test
- b. MRI lumbosacral spine
- c. Serum vitamin D measurement
- d. Sural nerve biopsy

Small-fiber Neuropathy

- Unmyelinated peripheral nerves that carry sharp pain, temperature, and autonomic nerve fibers

DDx Small-fiber Neuropathy

- **Impaired glucose metabolism**
- Vitamin B₁₂ deficiency
- HIV infection
- Amyloidosis
- Sjögren syndrome
- Paraproteinemia
- Celiac disease
- Sarcoidosis

35

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

- a. Carotid endarterectomy
- b. MRA neck
- c. Resumption of statin therapy
- d. Substitution of clopidogrel for aspirin

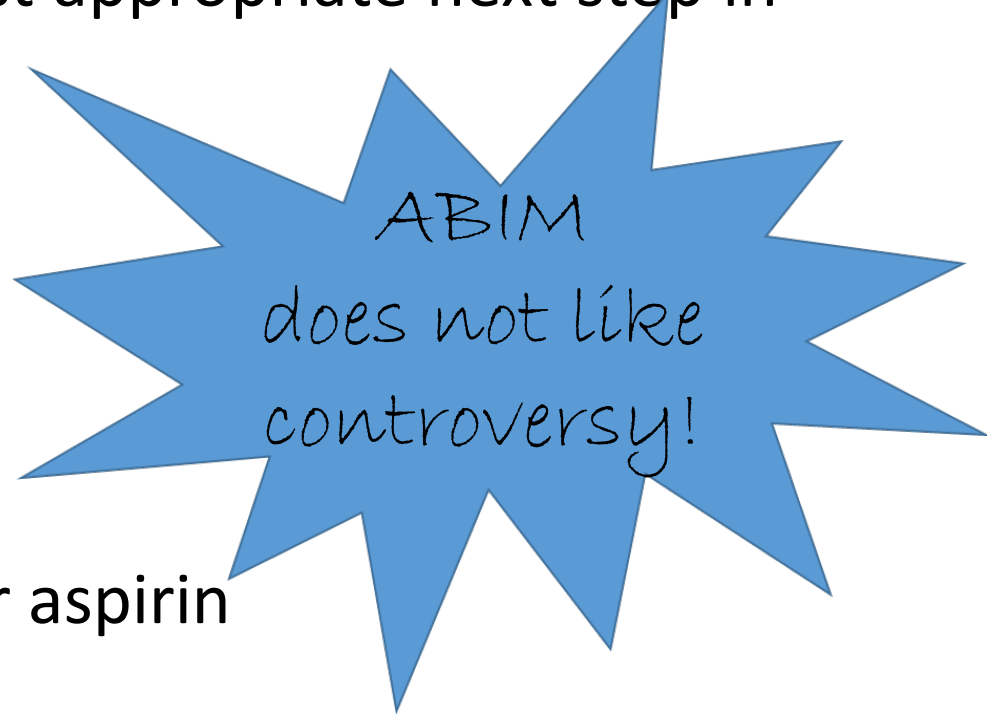


TABLE 1

Four Statin Benefit Groups and Major Recommendations From the 2013 ACC/AHA Guideline on the Treatment of Blood Cholesterol to Reduce Atherosclerotic Cardiovascular Risk in Adults (1)

Patient Group	Major Recommendations
1. Adults aged ≥ 21 years with clinical ASCVD (including history of or current acute coronary syndrome, myocardial infarction, stable or unstable angina, coronary or other arterial revascularization, stroke, TIA, or peripheral arterial disease presumed to be of atherosclerotic origin)	<ol style="list-style-type: none"> For patients age ≤ 75 years, high-intensity statin (or moderate-intensity statin if not a candidate for high-intensity statin due to safety concerns) For patients age > 75 years, moderate-intensity statin
2. Adults aged ≥ 21 years with LDL-C ≥ 190 mg/dL (not due to modifiable secondary causes)	<ol style="list-style-type: none"> High-intensity statin therapy to achieve $\geq 50\%$ reduction in LDL-C statin (or moderate-intensity statin if not a candidate for high-intensity statin due to safety concerns) May consider combining statin and non-statin therapy to further reduce LDL-C Cascade screening of close biologic relatives should be performed to identify others with the disease who would benefit from treatment.
3. Adults aged 40–75 years without ASCVD but with diabetes and with LDL-C 70–189 mg/dL	<ol style="list-style-type: none"> Moderate-intensity statin If 10-year ASCVD risk $\geq 7.5\%$, consider high-intensity statin.
4. Adults aged 40–75 years without ASCVD or diabetes, and with LDL-C 70–189 mg/dL and an estimated 10-year risk for ASCVD of $\geq 7.5\%$	<ol style="list-style-type: none"> Estimate 10-year ASCVD risk using Pooled Cohort Equations (2): <ol style="list-style-type: none"> If $\geq 7.5\%$, moderate- or high-intensity statin; If ≥ 5 to $< 7.5\%$, consider moderate-intensity statin. In selected individuals with 10-year ASCVD risk $< 5\%$, or age < 40 or > 75 years, individualize decisions based on presence of other high-risk features.* Before initiation of statin therapy for primary prevention, it is reasonable for clinicians and patients to engage in a discussion that considers the potential for ASCVD risk-reduction benefits and for adverse effects and drug-drug interactions, as well as patient preferences for treatment.

*The 2013 ACC/AHA guideline recommends consideration of other ASCVD risk factors (LDL-C ≥ 160 mg/dL, family history of premature ASCVD, hs-CRP ≥ 2.0 mg/L, CAC score ≥ 300 Agatston units, ABI < 0.9 , and high lifetime ASCVD risk).

ABI indicates ankle-brachial index; ACC, American College of Cardiology; AHA, American Heart Association; ASCVD, atherosclerotic cardiovascular disease; CAC, coronary artery calcification; hs-CRP, high-sensitivity C-reactive protein; and LDL-C, low-density lipoprotein cholesterol.

2016 ACC Expert Consensus Decision Pathway on the Role of Non-Statin Therapies for LDL-Cholesterol Lowering in the Management of Atherosclerotic Cardiovascular Disease Risk A Report of the American College of Cardiology Task Force on Clinical Expert Consensus Documents Writing Committee, Donald M. Lloyd-Jones, Pamela B. Morris, Christie M. Ballantyne, Kim K. Birtcher, David D. Daly Jr., Sondra M. DePalma, Margo B. Minissian, Carl E. Orringer and Sidney C. Smith Jr. Journal of the American College of Cardiology Volume 68, Issue 1, 5 July 2016, Pages 92-125

TABLE 2**Examples of High-, Moderate-, and Low-Intensity Statin Therapy (Adapted From 2013 ACC/AHA Guideline on the Treatment of Blood Cholesterol to Reduce Atherosclerotic Cardiovascular Risk in Adults)**

High-Intensity Statin Therapy	Moderate-Intensity Statin Therapy	Low-Intensity Statin Therapy
Daily dose lowers LDL-C, on average, by approximately $\geq 50\%$.	Daily dose lowers LDL-C, on average, by approximately 30% to $< 50\%$.	Daily dose lowers LDL-C, on average, by $< 30\%$.
Atorvastatin 40-80 mg Rosuvastatin 20-40 mg	Atorvastatin 10-20 mg Fluvastatin 40 mg twice daily Fluvastatin XL 80 mg Lovastatin 40 mg Pitavastatin 2-4 mg Pravastatin 40-80 mg Rosuvastatin 5-10 mg Simvastatin 20-40 mg	Fluvastatin 20-40 mg Lovastatin 20 mg Pitavastatin 1 mg Pravastatin 10-20 mg Simvastatin 10 mg

Bold face type indicates statins and doses that were evaluated in RCTs included in the 2013 ACC/AHA guideline.

ACC indicates American College of Cardiology; AHA, American Heart Association; LDL-C, low-density lipoprotein cholesterol; and RCT, randomized controlled trial.

2016 ACC Expert Consensus Decision Pathway on the Role of Non-Statin Therapies for LDL-Cholesterol Lowering in the Management of Atherosclerotic Cardiovascular Disease Risk A Report of the American College of Cardiology Task Force on Clinical Expert Consensus Documents Writing Committee, Donald M. Lloyd-Jones, Pamela B. Morris, Christie M. Ballantyne, Kim K. Birtcher, David D. Daly Jr., Sondra M. DePalma, Margo B. Minissian, Carl E. Orringer and Sidney C. Smith Jr. *Journal of the American College of Cardiology* Volume 68, Issue 1, 5 July 2016, Pages 92-125

- Using a statin to treat patients with asymptomatic internal carotid artery stenosis is associated with a stroke risk of less than 2% per year.

36

Which of the following is the most appropriate treatment?

- a. Indomethacin
- b. Normalization of blood pressure
- c. tPA
- d. warfarin

Format: Abstract ▾

Send to ▾

Headache. 2013 Mar;53(3):570-6. doi: 10.1111/head.12040.

Reversible cerebral vasoconstriction syndrome.

Yancy H¹, Lee-Iannotti JK, Schwedt TJ, Dodick DW.

⊕ Author information

Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is a cerebrovascular disorder with a clinical picture that continues to be refined. It has presented to multiple subspecialties over the past several decades, bringing with it many questions regarding risk factors, diagnosis, and management. Answers have been forthcoming but many questions remain. RCVS presents with recurrent, secondary thunderclap headaches and predominantly affects young women. The mechanism of vasoconstriction is unclear, but there has been speculation regarding a hyperadrenergic state. Diagnosis requires physician awareness, vascular imaging, and knowledge of the differential. The hallmark of its diagnosis is reversibility. Management is empiric, usually with calcium-channel blockers, as there are no controlled treatment trials for RCVS. Randomized controlled trials are needed.

PMID: 23489219 DOI: [10.1111/head.12040](https://doi.org/10.1111/head.12040)

[Indexed for MEDLINE]



47

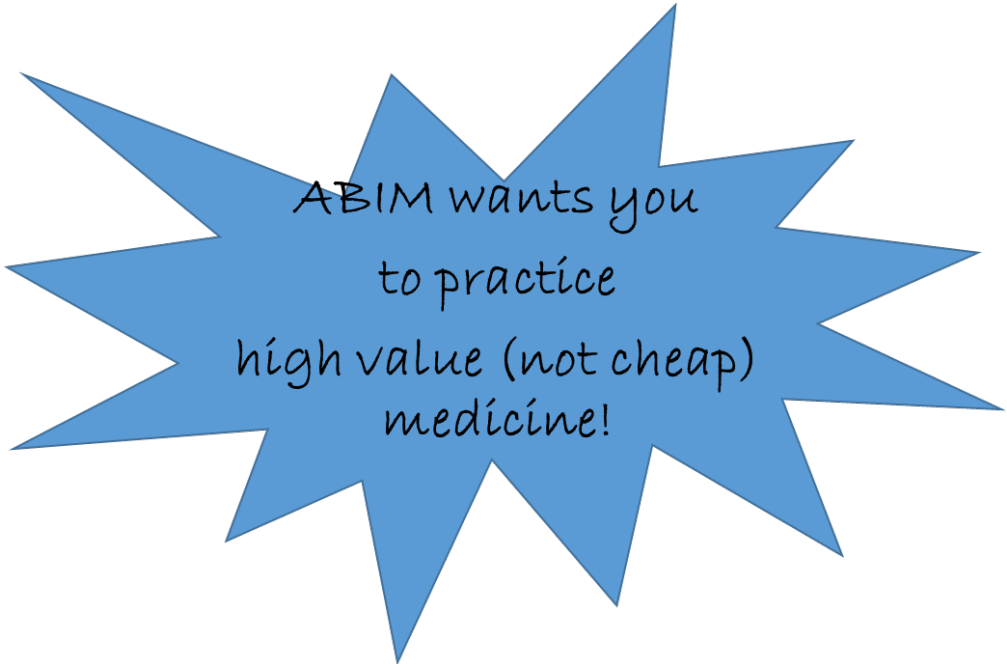
Which of the following is the most appropriate next step in management?

- a. Ambulatory EEG monitoring
- b. Carbamazepine
- c. Clinical observation
- d. LP

Treat Based on Risk of Recurrence

- 2-year risk of recurrence after a single unprovoked seizure is approximately 40%
- 80–90% of individuals who recur do so within 2 years of the initial seizure
- Risk Factors
 - previous head trauma with loss of consciousness
 - focal brain lesion on MRI
 - postictal Todd paralysis of the right arm (focal weakness after a seizure) or other abnormality on neuro exam
 - Abnormal EEG

Evaluate a first seizure in a patient at high risk of recurrent seizure.



ABIM wants you
to practice
high value (not cheap)
medicine!

48

Which of the following is the most appropriate management?

- a. CT head
- b. EEG
- c. ESR measurement
- d. MRI brain
- e. sumatriptan

Migraine without Aura Diagnostic criteria

- A. At least five attacks fulfilling criteria B–D
- B. Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)
- C. Headache has at least two of the following four characteristics:
 - 1. unilateral location
 - 2. pulsating quality
 - 3. moderate or severe pain intensity
 - 4. aggravation by or causing avoidance of routine physical activity (e.g. walking or climbing stairs)
- D. During headache at least one of the following:
 - 1. nausea and/or vomiting
 - 2. photophobia and phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis.

Migraine with Aura Diagnostic criteria

- A. At least two attacks fulfilling criteria B and C
- B. One or more of the following fully reversible aura symptoms:
 - 1. visual
 - 2. sensory
 - 3. speech and/or language
 - 4. motor
 - 5. brainstem
 - 6. retinal
- C. At least two of the following four characteristics:
 - 1. at least one aura symptom spreads gradually over 5 minutes, and/or two or more symptoms occur in succession
 - 2. each individual aura symptom lasts 5-60 minutes
 - 3. at least one aura symptom is unilateral
 - 4. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.



Treating migraine headaches

Some drugs should rarely be used

AAN Guideline: Acute Management of Migraine

- Must Offer (Level A)
 - None
- Should Offer (Level B)
 - Intravenous metoclopramide side effects including akathisia and drowsiness
 - Intravenous prochlorperazine side effects including akathisia and drowsiness
 - Subcutaneous sumatriptan
 - May be less efficacious than intravenous anti-dopaminergics
 - Contraindications
 - Those who have used ergotamine, DHE, or a triptan medication within the previous 24 hours.

AAN Guideline: Migraine Treatment in ED

- **AVOID**
- Intravenous **diphenhydramine** may be AVOIDED in adults who present to an ED with acute migraine (May avoid–Level C). The efficacy of diphenhydramine with regard to treatment of akathisia was beyond the scope of this work.
- Intravenous **hydromorphone** may be AVOIDED in adults who present to an ED with acute migraine (May avoid–Level C).
- Intravenous **lidocaine** may be AVOIDED in adults who present to an ED with acute migraine (May avoid–Level C).
- Intravenous **morphine** may be AVOIDED in adults who present to an ED with acute migraine (May avoid–Level C).
- Intravenous **octreotide** may be AVOIDED in adults who present to an ED with acute migraine (May avoid–Level C).

Orr, S. L., Friedman, B. W., Christie, S., Minen, M. T., Bamford, C., Kelley, N. E. and Tepper, D. (2016), Management of Adults With Acute Migraine in the Emergency Department: The American Headache Society Evidence Assessment of Parenteral Pharmacotherapies. *Headache: The Journal of Head and Face Pain*, 56: 911–940. doi:10.1111/head.12835

53

Which of the following is the most likely diagnosis?

- a. Multiple system atrophy
- b. Parkinson disease
- c. Progressive supranuclear palsy
- d. Vascular parkinsonism

Parkinson-Plus Syndromes

- Multiple system atrophy
 - Cerebellar dysfunction (olivopontocerebellar degeneration)
 - **Autonomic dysfunction** (Shy-Drager)
 - Cognition often spared
- Progressive supranuclear palsy
 - Postural instability
 - Cognitive impairment more likely
 - Staring eyes, axial rigidity
 - **Eye movements impaired** (downward then upward)
- Diffuse Lewy body disease
 - **Visual hallucination**
 - **Sleep** disturbance
 - Cognitive fluctuation
 - Eventual parkinsonism
- Corticobasal degeneration
 - **Apraxia and dystonia** are true hallmarks

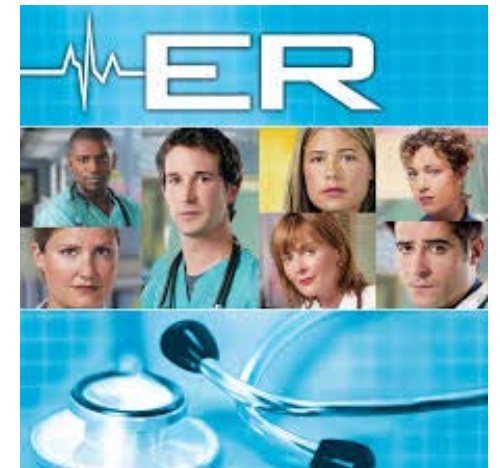


Table 1: Clinical features differentiating PPS from Idiopathic PD

	Typical PD	Park. Plus Syndrome
1. Pattern of onset	Asymmetrical	Usual symmetrical (exc. CBGD)
2. i) Rigidity	Peripheral > Axial	Axial > Peripheral
ii) Rest tremor	Present	Absent or atypical
iii) Assoc. Features (early dementia, vertical gaze abnormality, pyramidal tract lesion, cerebellar features, early autono. disturb, cortical sensory impairment and myoclonus)	Absent	Usually present depending upon the type
3. Response to levodopa	Excellent	Mild to moderate response initially and then gradually wanes after one year
4. Progress of the disease	Slow	Usually rapid

Red Flags: When parkinsonism Isn't Parkinsons

- History of severe cerebral trauma, stroke, exposure to neurotoxins or anti-dopaminergic agents
- No rest tremor
- Symmetrical signs
- Early falls
- Associated ophthalmoplegia, pyramidal or cerebellar signs
- Associated autonomic dysfunction
- Rapid disease progression
- Poor response to levodopa

Neurology Test Review

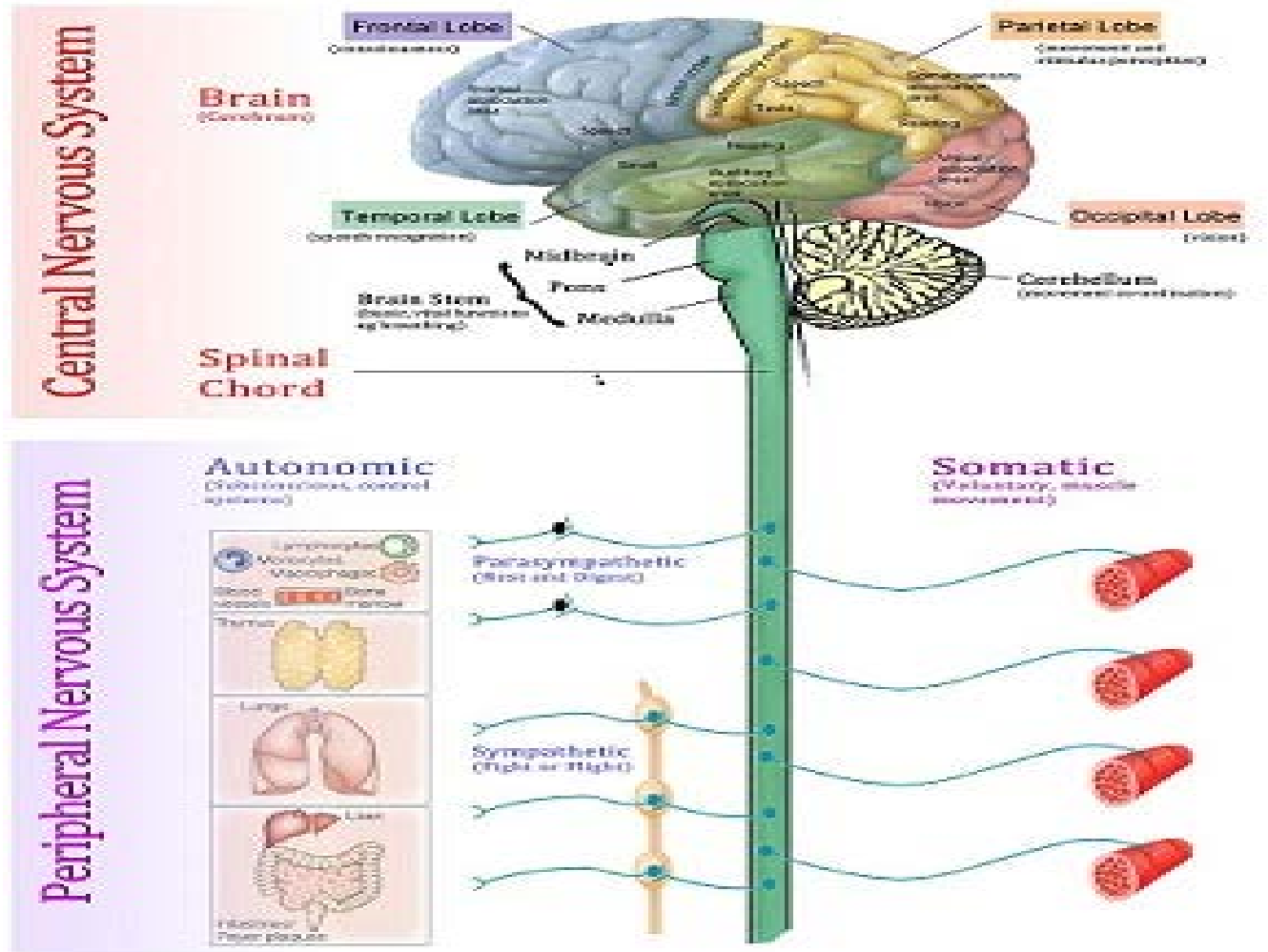
Akshjot Puri

PGY 3

Internal Medicine

QUESTION 11

The Nervous System



Peripheral neuropathy

Mononeuropathy

- Vasculitis
- Meralgia paresthetica
- Carpal tunnel syndrome
- Bells palsy

Brachial and Lumbosacral plexopathy

Polyneuropathy

- DM
- Hereditary-CMT-1,2
- Inflammatory- GBS, CIDP

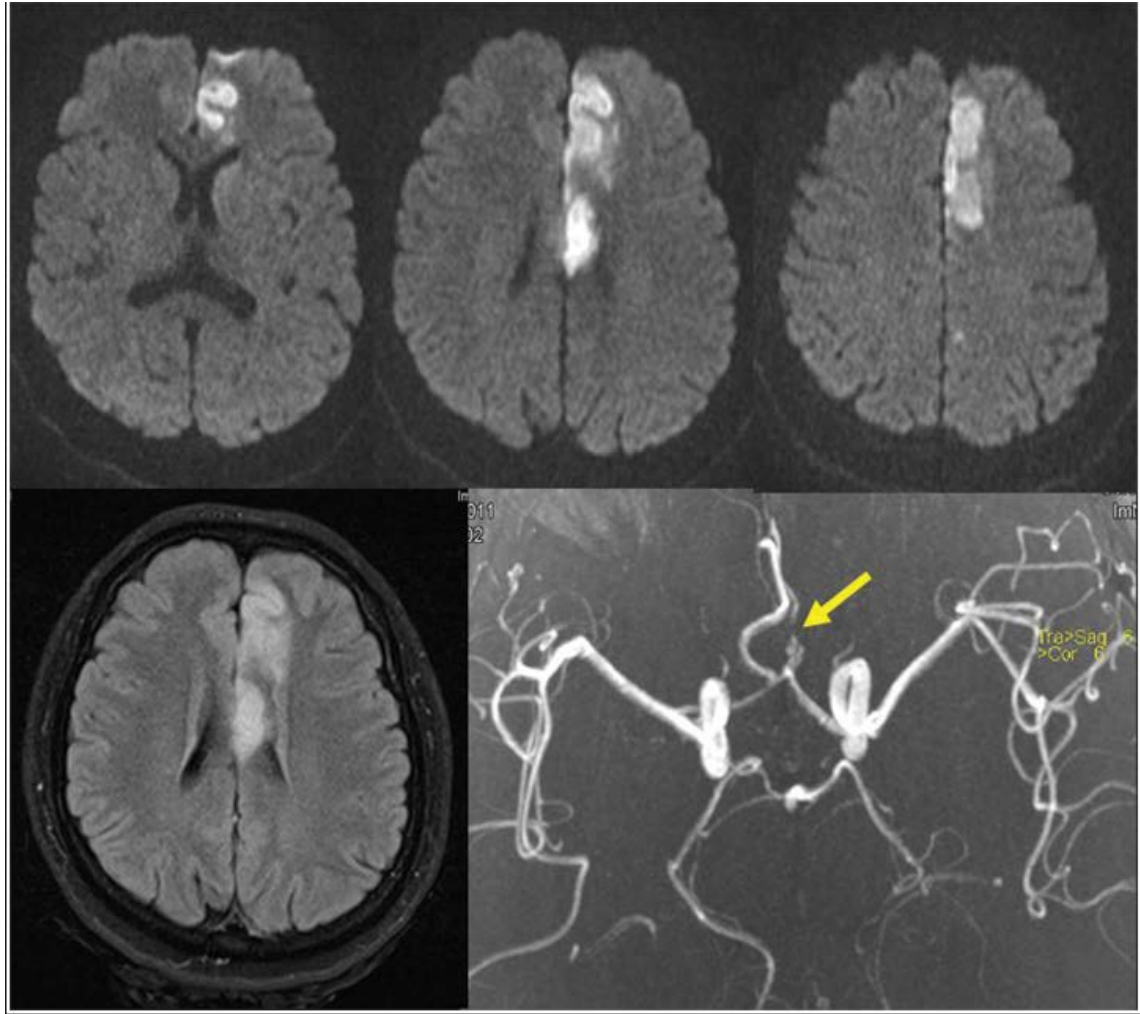
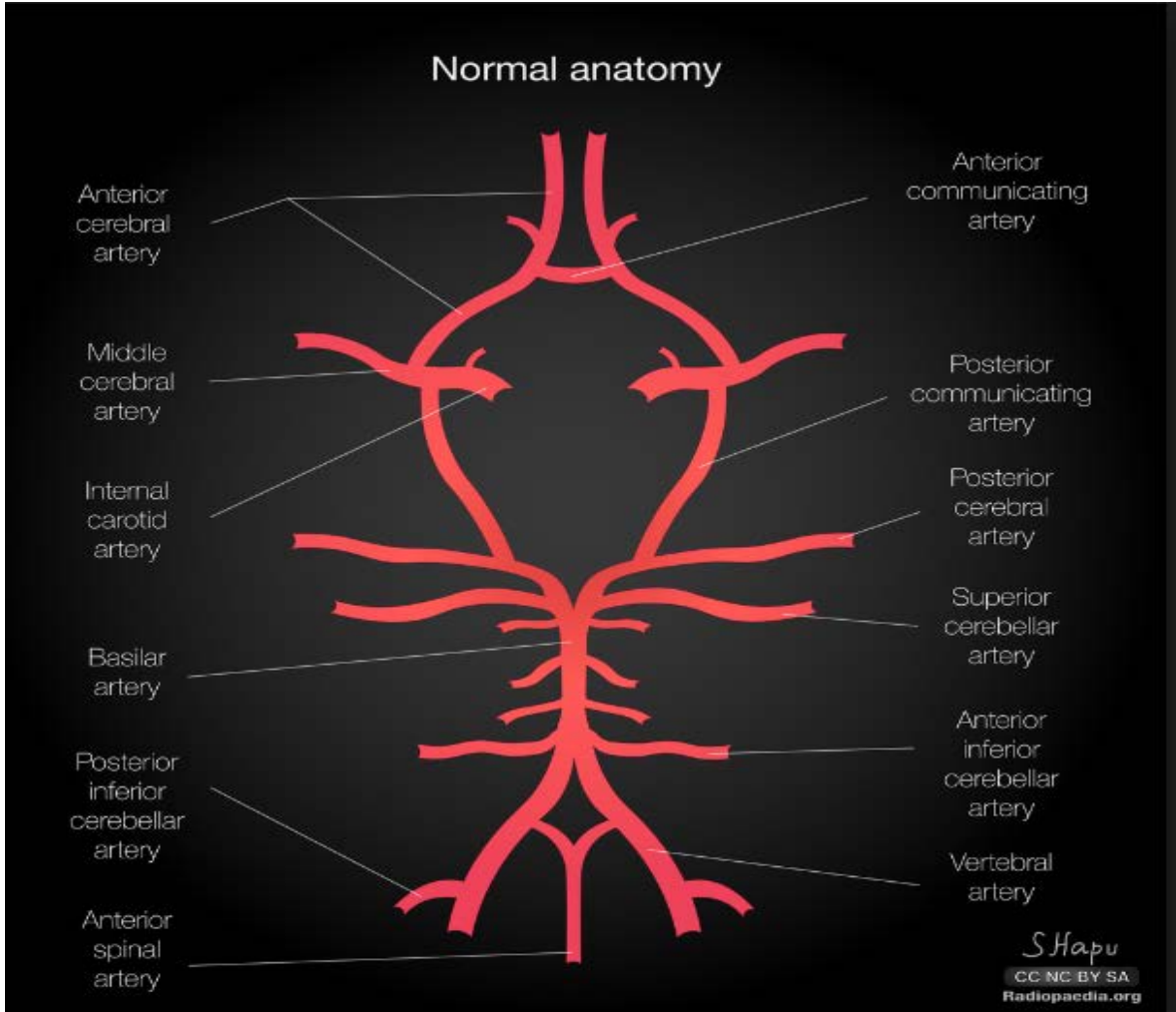
Critical Illness neuropathy

Systemic

Paraprotein-Amyloid, MGUS

- 5 months worsening
- Pain in thighs
- Presyncope
- Diffuse areflexia
- Demyelinating pattern- slowing of conduction velocities
- Idiopathic/DM/paraproteinemia/HIV
- Responds to steroids and immunomodulators

QUESTION 12



- 25% of cryptogenic ischemic strokes have PAF on 30 day cardiac monitoring
- PACs/Ectopy more likely
- Isolated PFO-incidental finding
- AC- only when proven A fib
- DAPT no evidence

QUESTION 13

- Acute dopamine agonist withdrawal syndrome- PARKINSONIAN HYPERPYREXIA SYNDROME
- AMS/hyperthermia/rhabdo/EPS- **dystonia, rigidity**
- Mortality rates 4%

MYOCLONUS

RIGIDITY

SPASTICITY

HYPERREFLEXIA

DYSTONIA

SEROTONIN SYNDROME

- SSRI/SNRI/MAO/TCA/Tramadol/amphetamine/metoclopramide/ondansetron/triptans
- Hyperthermia, AMS , **hyperreflexia**, tremors, **myoclonus**, rhabdo, seizures
- Sweating, diarrhea, dilated pupils
- Trt- stop offending agent, BZD, Cyproheptadine

NEUROLEPTIC MALIGNANT SYNDROME

- Typical antipsychotics
- Hyperthermia, AMS, **rigidity**, rhabdo, seizures
- Diarrhea, tachycardia, variable BP
- Trt- Bromocriptine, dantrolene, BZD

MALIGNANT HYPERTHERMIA

- RYR 1 genetic mutation, Sch(depolarizing, volatile)
- Hyperthermia, **rigidity**,
- Tachycardia
- Trt-dantrolene

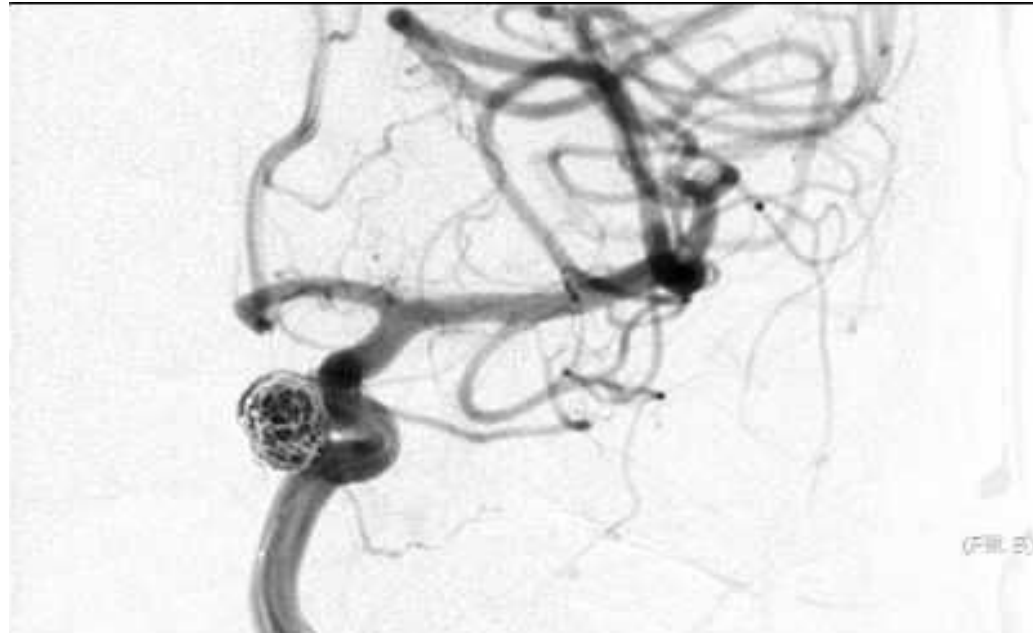
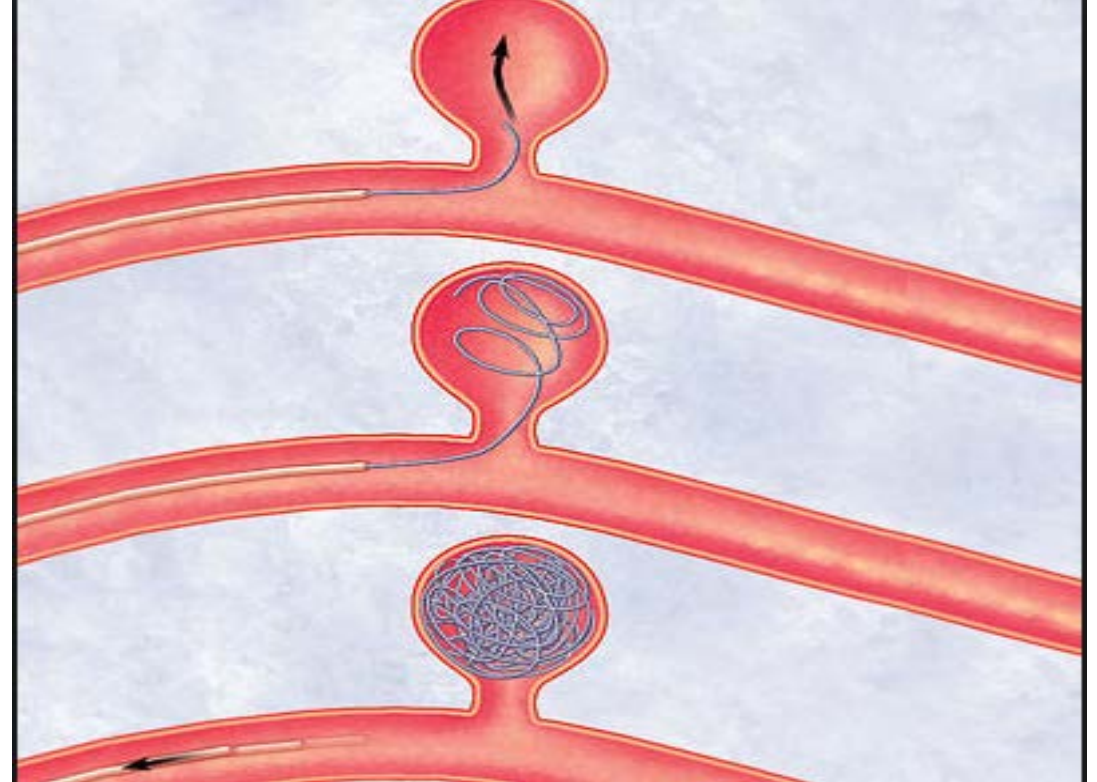
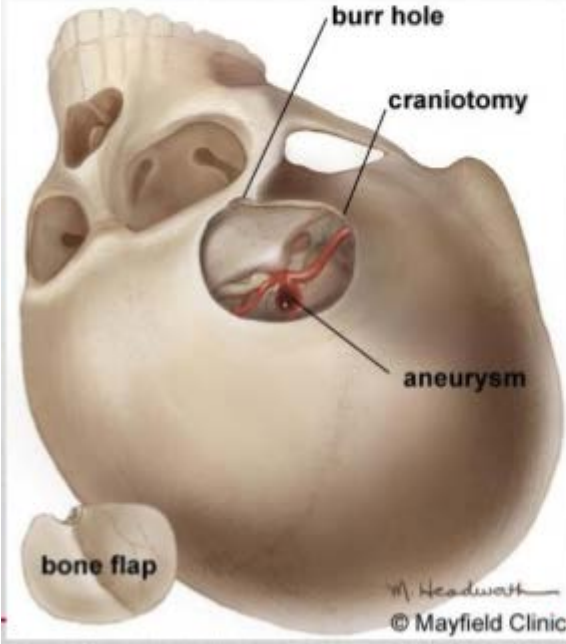
QUESTION 14

- Decreases vasospasm
- Decreases morbidity, mortality even in absence of vasospasm
- Neuroprotective- Ca influx blocked
- 21 Days- Aneurysmal SAH
- Transcranial US neg
- IV dopamine, vasopressor not indicated without signs of active vasospasm

COMPLICATIONS SAH

1. Rebleeding
2. Hydrocephalus
3. Cerebral edema
4. Seizures

Clipping



Mayer SA, Chong JY. Critical care management of increased intracranial pressure. J Intensive Care Med 2002;17:55-67

1. CT head STAT
2. Call Neurosurgery/Neurology/ICU
3. Elevate head of the bed 30 degrees
4. Avoid any fevers
5. Paco₂ 25-30
6. Pao₂ <50
7. MAP maintenance (MAP-ICP=CPP)
8. Mannitol q6h IV or hypertonic saline 3%
9. Sedation
10. Pentobarbital coma
11. Hypothermia 32-34C
12. Seizure prophylaxis
13. Steroids

QUESTION 15

- Common to have sinus pressure/drainage
- Triggers- stress, hormonal, weather, sleep pattern, meal pattern, light, noise, odor
- Trt- NSAIDS (1st line- Cost effectiveness), triptans , dihydroergotamine

Table 4. ICHD-2 Diagnostic Criteria for Migraine Without Aura

At least five episodes fulfilling the following criteria:

Headache episodes lasting four to 72 hours (untreated or unsuccessfully treated)

Headache has at least two of the following characteristics: unilateral location, pulsating quality, moderate or severe pain intensity, aggravated by (or causes avoidance of) routine physical activity such as walking or climbing stairs

During the headache, the patient experiences at least one of the following: nausea or vomiting; and photophobia and phonophobia

Headache is not attributed to another disorder

ICHD-2 = International Classification of Headache Disorders, 2nd ed.

Adapted with permission from the American Academy of Neurology: Lipton RB, Bigal ME, Steiner TJ, et al. Classification of primary headaches. Neurology. 2004;63(3):428. Table 2. ICHD-2 diagnostic criteria for 1.1 Migraine without aura. <http://www.neurology.org/content/63/3/427.abstract>.

QUESTION 16

MYOPATHY

1. Inflammatory- PM, DM, IBM
2. Endocrine- Steroid, thyroid, acromegaly, hyper PTH, Addison's , Vitamin D def
3. Toxic-Statins(+gemfibrozil/fenofibrate/CYP3A4 -), Etoh, Interferon, HAART
4. Inherited

Muscle dystrophy (Beckers & Emery Dreifuss- adulthood, cardiac complications)

Myotonic dystrophy (Myotonia, delayed hand grip release)

Mitochondrial – ophthalmoplegia

Glycogen storage disease, fatty acid oxidation pathways



QUESTION 17

- BP before rtPA 185/110 after rtPA 180/105
- Complication- ICH 6% (Risks- large cerebral CVA, high NIHSS, cardioembolic, protocol violations)
- Hold antiplatelet/AC 24 hrs, till post 24 hr CT confirms no hemorrhage
- Choice- IV nicardipine or labetalol
(S/L nitro- unable to titrate BP, raises ICP)

Establish absolute exclusion criteria:

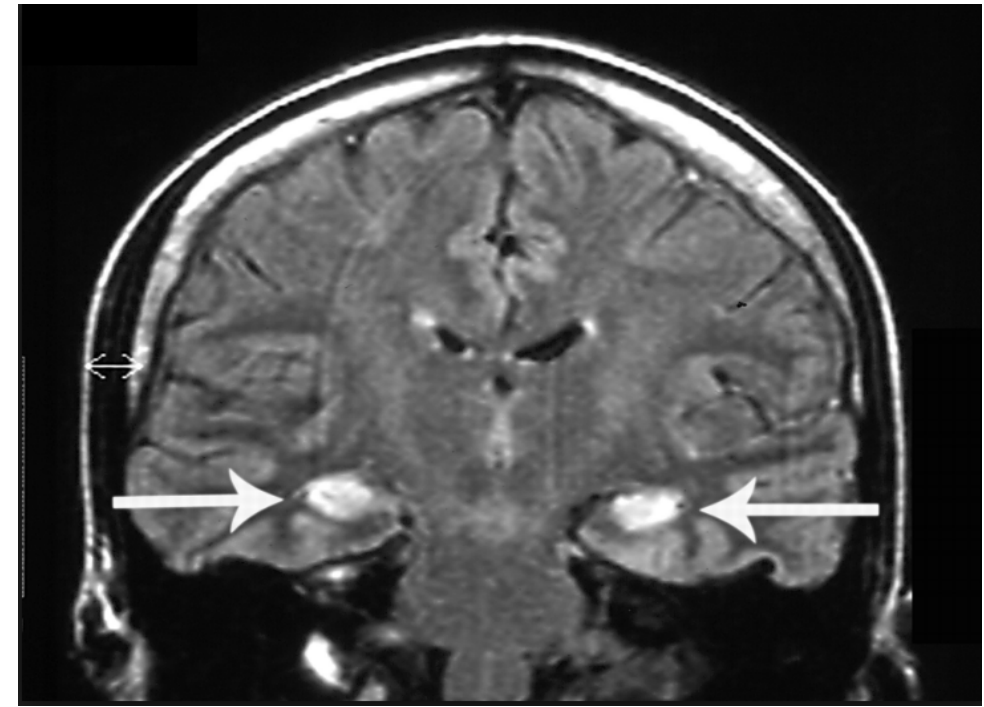
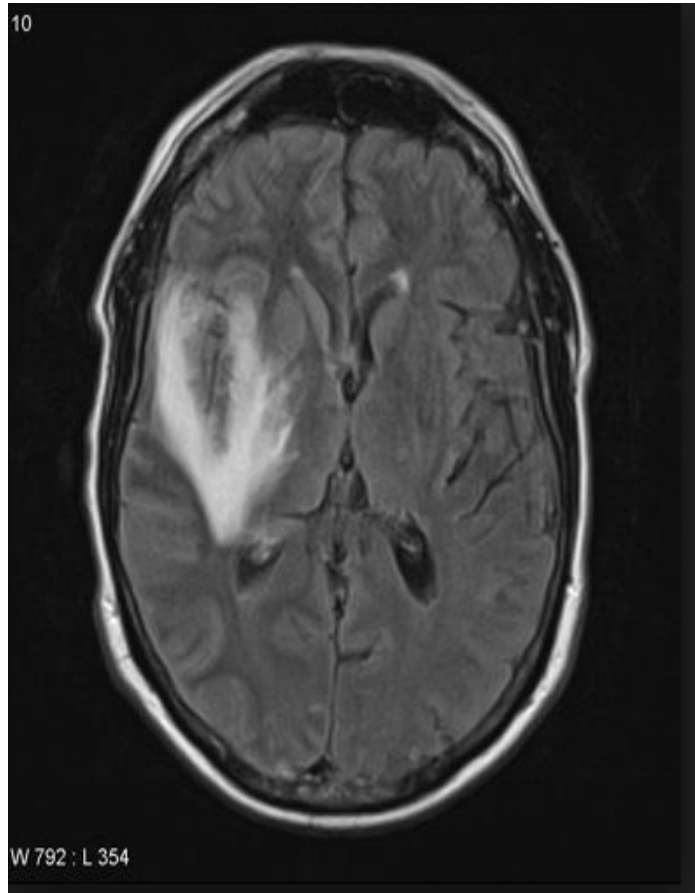
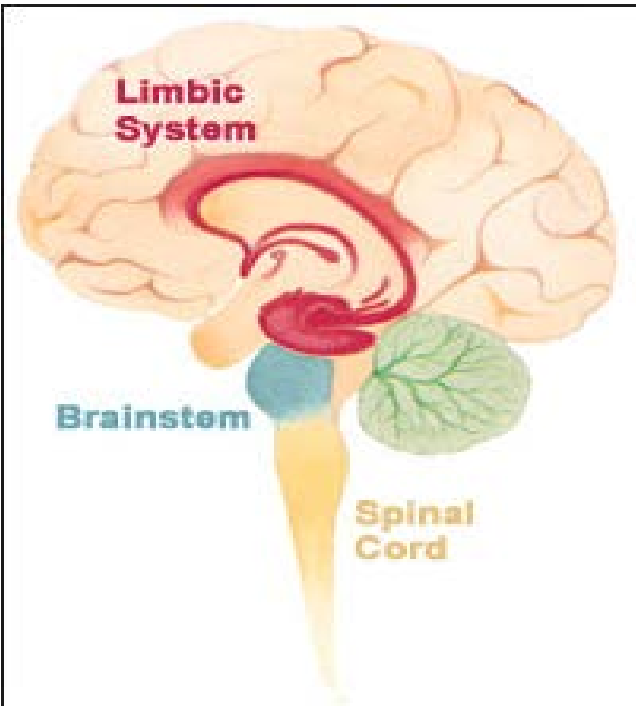
- Significant head trauma or stroke within 3 months
- Suspicion of subarachnoid hemorrhage
- Arterial puncture at noncompressible site within 7 days
- Previous intracranial hemorrhage
- Intracranial neoplasm, vascular malformation, or aneurysm
- Recent intracranial or spinal surgery
- Inability to control blood pressure to < 185/110 mm Hg
- Active internal bleeding
- Acute bleeding risk: platelet count < 100,000/ μ L (100×10^9 /L), heparin administered within 48 hours with greater than normal aPTT, anticoagulant use with INR > 1.7 or PT > 15 s, current use of direct thrombin inhibitors or direct factor Xa inhibitors with elevated results on blood tests (aPTT, INR, platelet count, ecarin clotting time, thrombin time, or anti-factor Xa level)
- Hypodensity evident in greater than one third of the middle cerebral artery territory on head CT

Relative exclusion criteria²:

- Minor or rapidly improving symptoms
- Pregnancy
- Seizure at onset
- Major surgery or trauma within 14 days
- Gastrointestinal or urinary tract hemorrhage within 21 days
- Recent acute myocardial infarction within 3 months

QUESTION 18

- **Short term memory loss-MMSE**, mood-personality changes, sleep, delusions, psychosis, seizures, dementia
- Etiology- paraneoplastic (tumor or Ab hu/LGII) or non paraneoplastic
- Progress over weeks-months
- EEG- Epileptic activity u/l or b/l temporal lobes
- MRI-Hyperintense signals medial part of u/l or b/l temporal lobes
- CSF- Lymphocytic pleocytosis
- Differential-
Infectious- HSV1/2, HHV 6, VZV, CMV, Treponema pallidum
Neoplastic- paraneoplastic, mets
Autoimmune- SLE, sjogrens, cerebral vasculitis
Metabolic- Hashimoto, Korsakoff
- Reversible



QUESTION 19

- HYDROPHILIC STATINS- Rosuvastatin, pravastatin, fluvastatin
LIPOPHILIC STATINS – Atorvastatin, simvastatin, lovastatin
- Immune mediated necrotizing form of statin myopathy associated with anti HMGCR autoantibodies, progressive, immunosuppression
- Increased risk with –gemfibrozil, CYP3A4 –

QUESTION 20

PCNSLymphoma/NHL

- Immunocompromised-HIV/Immunocompetent
- 90% DLBCL, 10% rest- burkitts, T cell
- Any locations CNS, ocular, leptomeninges, multiple
- Stereotactic biopsy(gold standard), CSF + 15%
- Trt- Steroids, **Methotrexate based**, WBRT

No surgery

Virchow robin perivascular spaces

