KANSAS INITIATIVE FOR STROKE SURVIVAL

A PROJECT BY AND FOR KANSANS

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Cerebral vasculopathies

"First Tuesdays" Lecture Series

Sabreena Slavin, MD

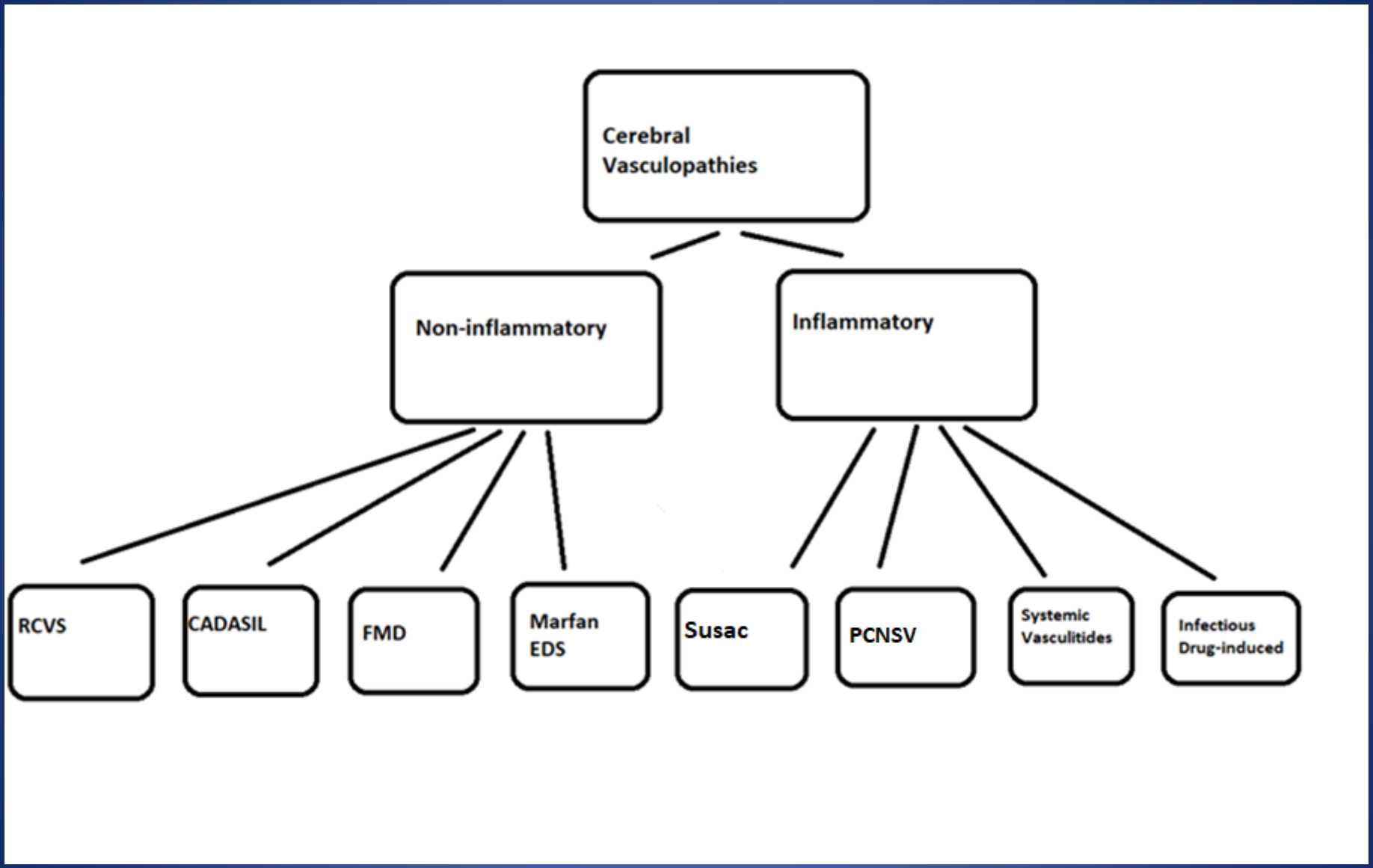
Introduction and Goal of “First Tuesdays”

- Didactic lecture series as part of the Kansas Initiative for Stroke Survival (KISS)
- Updates in Practice and FAQ’s on Acute Stroke Care
- 20 minute didactic, 10 minutes for questions/discussion

Epidemiology of non-atherosclerotic vasculopathy and stroke in the young

- Comprises 7-25% of stroke in the young
  - Arterial dissection: 20–25%
  - Infectious vasculitis: 7%
  - Moyamoya disease in Asian populations: 6-15%
  - Remaining causes: less than 1% each

Griffiths et al, *Stroke Res Treat* 2011


Cerebral Vasculopathies
Non-inflammatory
Inflammatory
RCVS
CADASIL
FMD
Marfan EDS
Susac
PCNSV
Systemic Vasculitides
Infectious Drug-induced

Diagnostic modalities

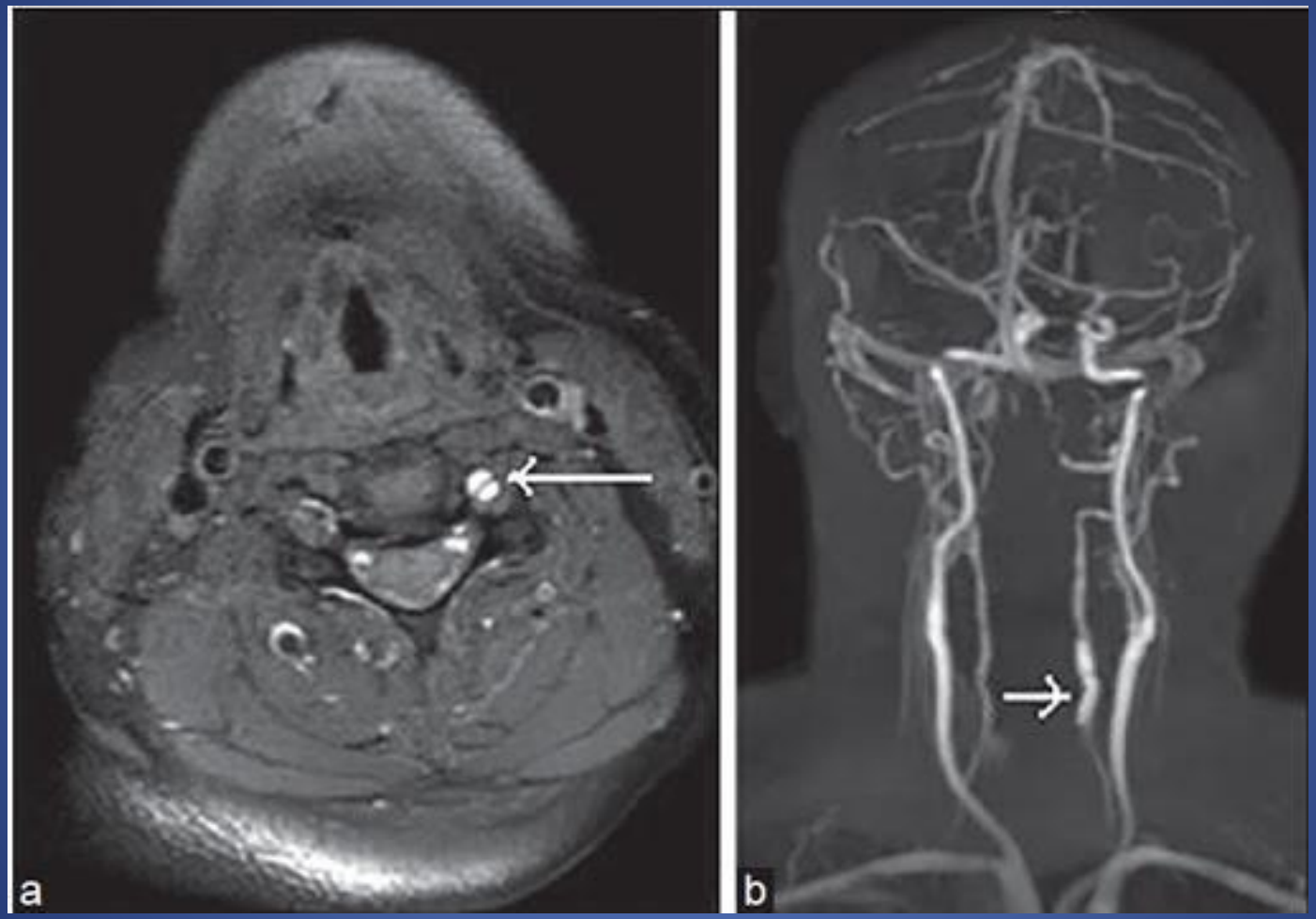
- MRI brain
  - Territories affected, genetic syndromes
- Noninvasive vessel imaging
  - Territories affected, inflammatory vs noninflammatory
- Systemic workup
- CSF
  - Inflammatory vs noninflammatory
- Angiogram
  - Confirmation of vasculopathy, territories affected, reversibility
- Biopsy
  - Gold standard diagnosis?

Arterial dissection

- Can involve cervical portion of internal carotid artery or vertebral artery
- Represents about 1/5 of stroke causes in patients younger than 45
- Traumatic or spontaneous – connective tissue disease such as Marfan syndrome and EDS can predispose
- Clinical symptoms
  - head/neck/facial pain
  - cerebral and/or retinal symptoms
  - Horner's syndrome with carotid; Vertigo/nystagmus with vertebral

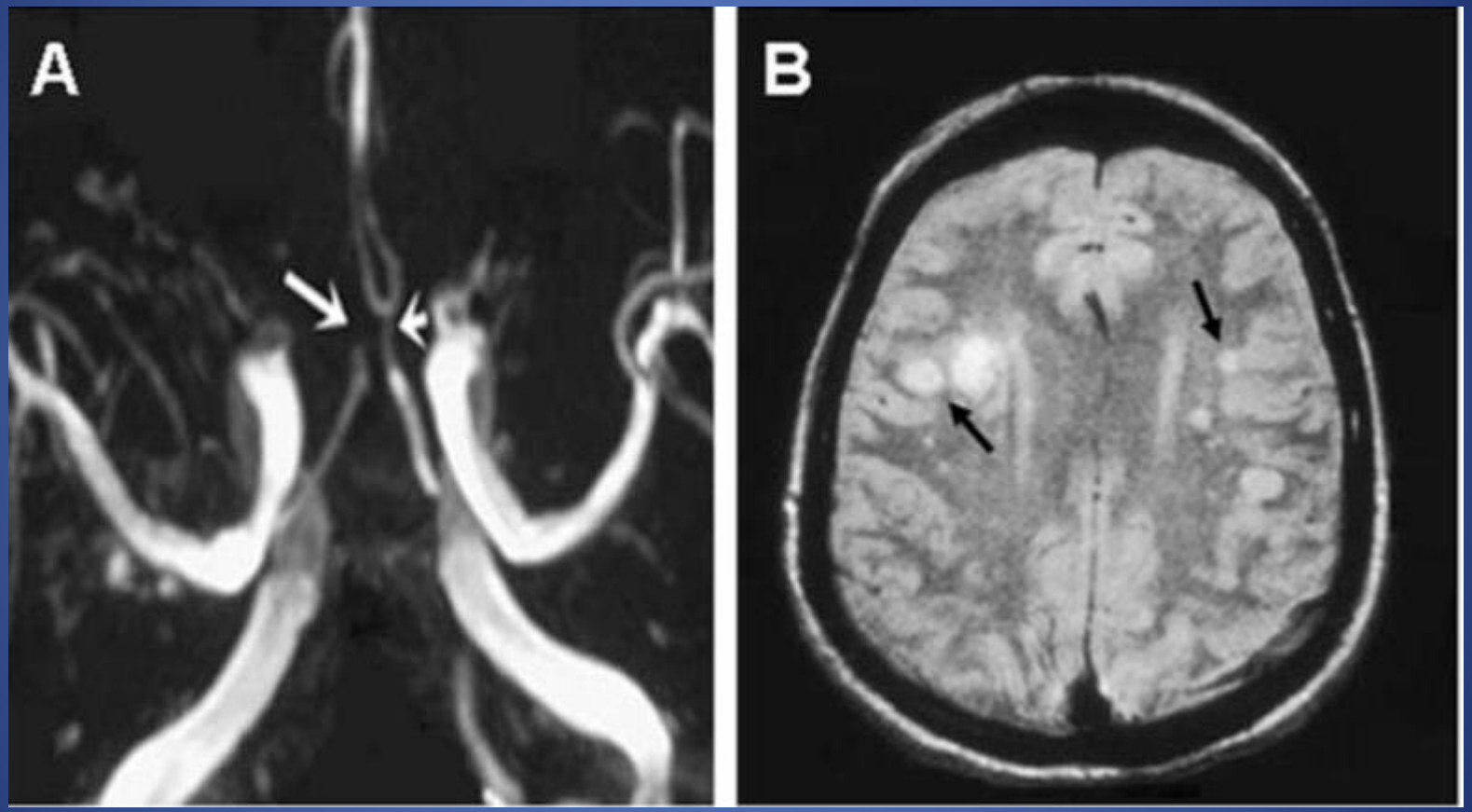
Peng et al, *Biomed Res Int* 2017

Vertebral dissection

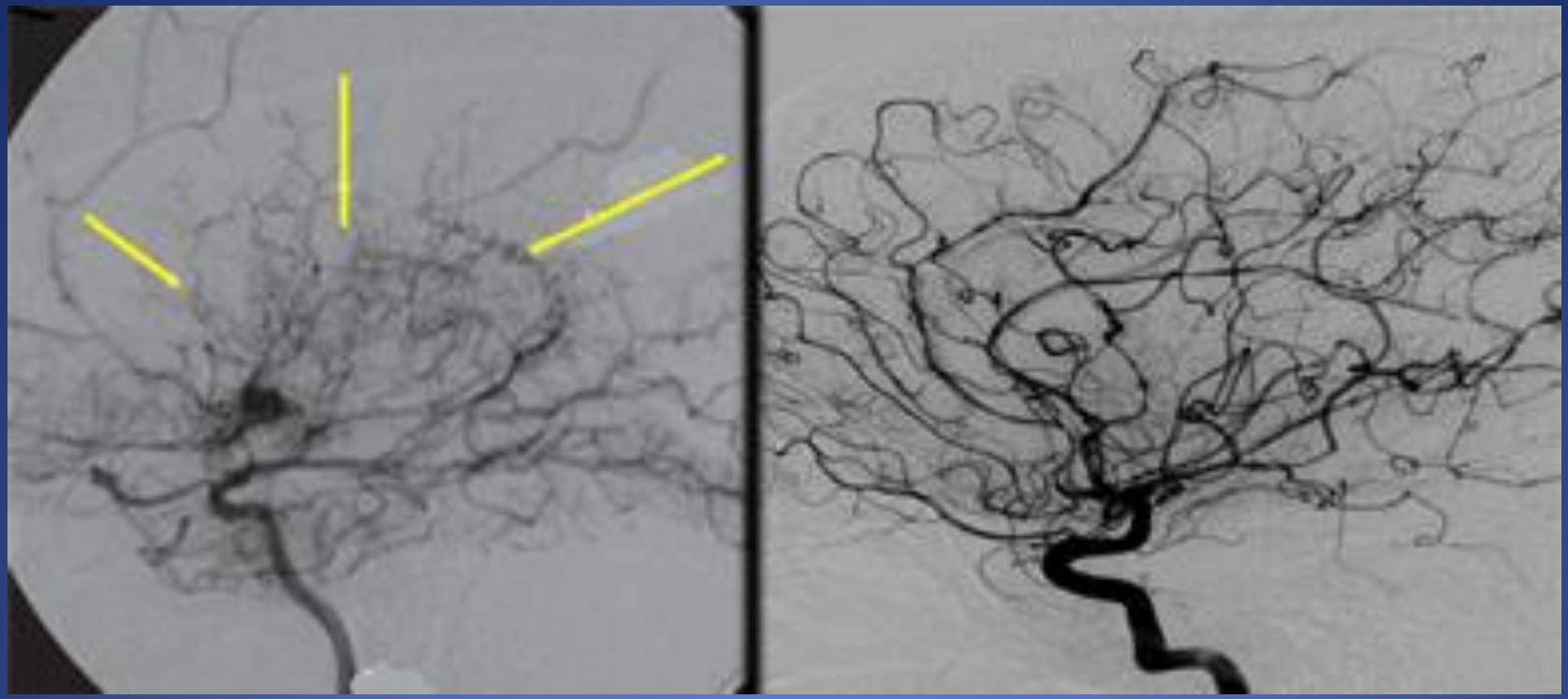


Deshmukh et al, *J Craniovertebr Junction Spine* 2015

VZV Vasculitis



Nagal et al, *Neurology* 2008

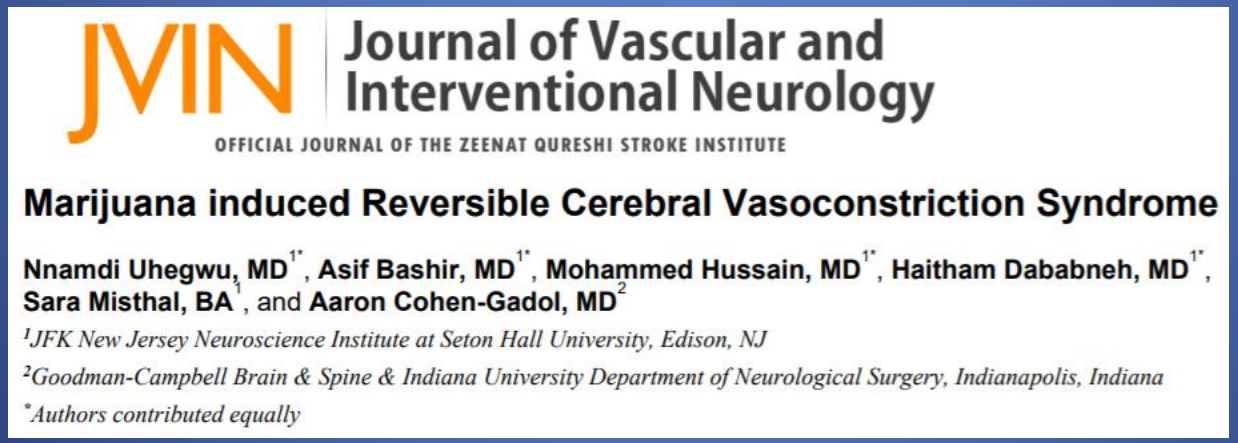


- Moyamoya disease vs normal
- Best outcomes with surgical treatment: most commonly STA-MCA bypass

Kim et al, *J Stroke* 2016

Most common drugs to cause vasculopathies

- Amphetamines
- Cocaine
- Marijuana/impurities?



JVIN Journal of Vascular and Interventional Neurology

OFFICIAL JOURNAL OF THE ZEENAT QURESHI STROKE INSTITUTE

Marijuana induced Reversible Cerebral Vasoconstriction Syndrome

Nnamdi Uhegwu, MD[1*], Asif Bashir, MD[1*], Mohammed Hussain, MD[1*], Haitham Dababneh, MD[1*], Sara Misthal, BA[1], and Aaron Cohen-Gadol, MD[2]

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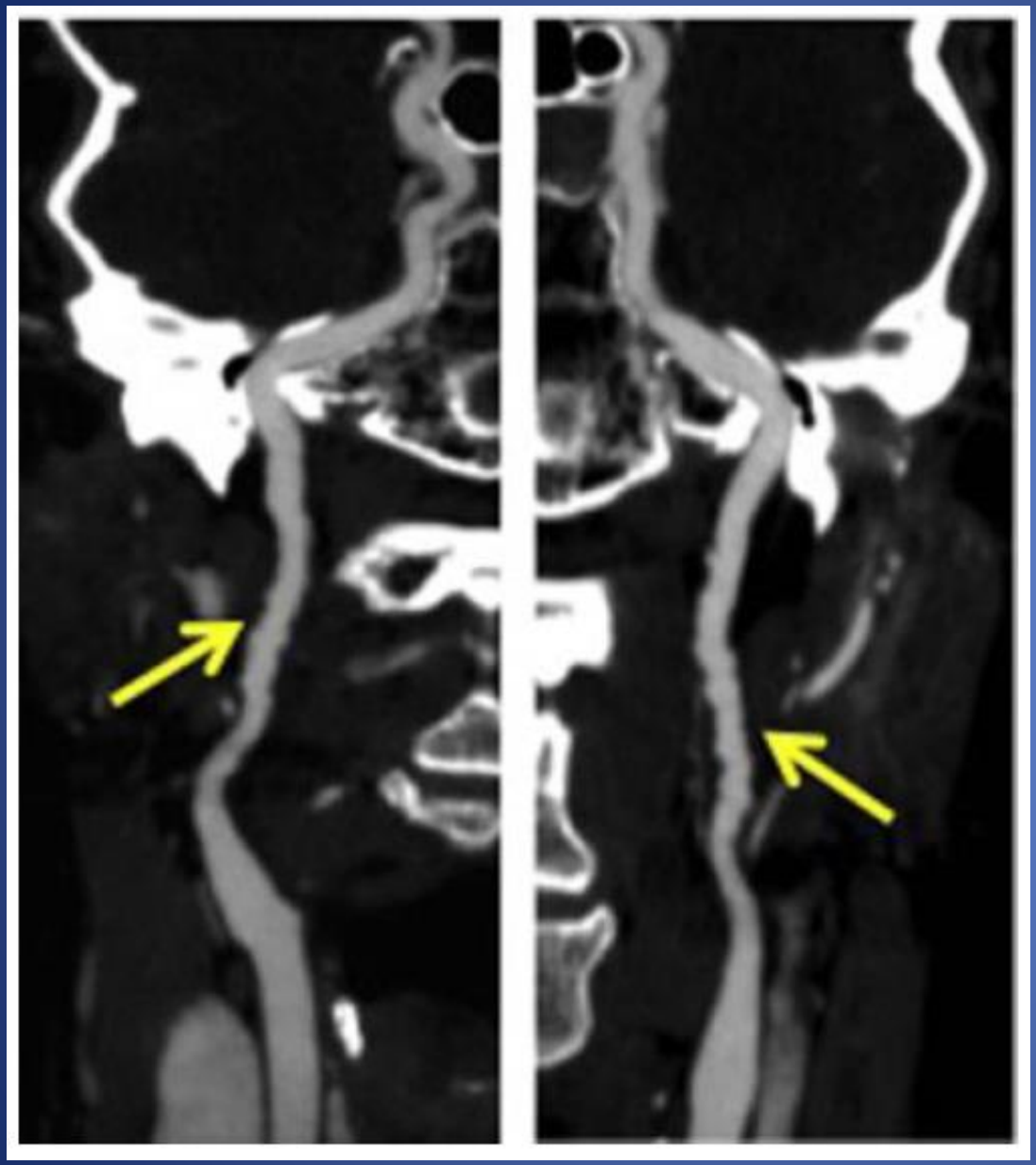
[*]Authors contributed equally

Fonseca et al, *Curr Neurol Neurosci Rep* 2013; Uhegwu et al, *J Vasc Interv Neurol* 2015



Fibromuscular dysplasia

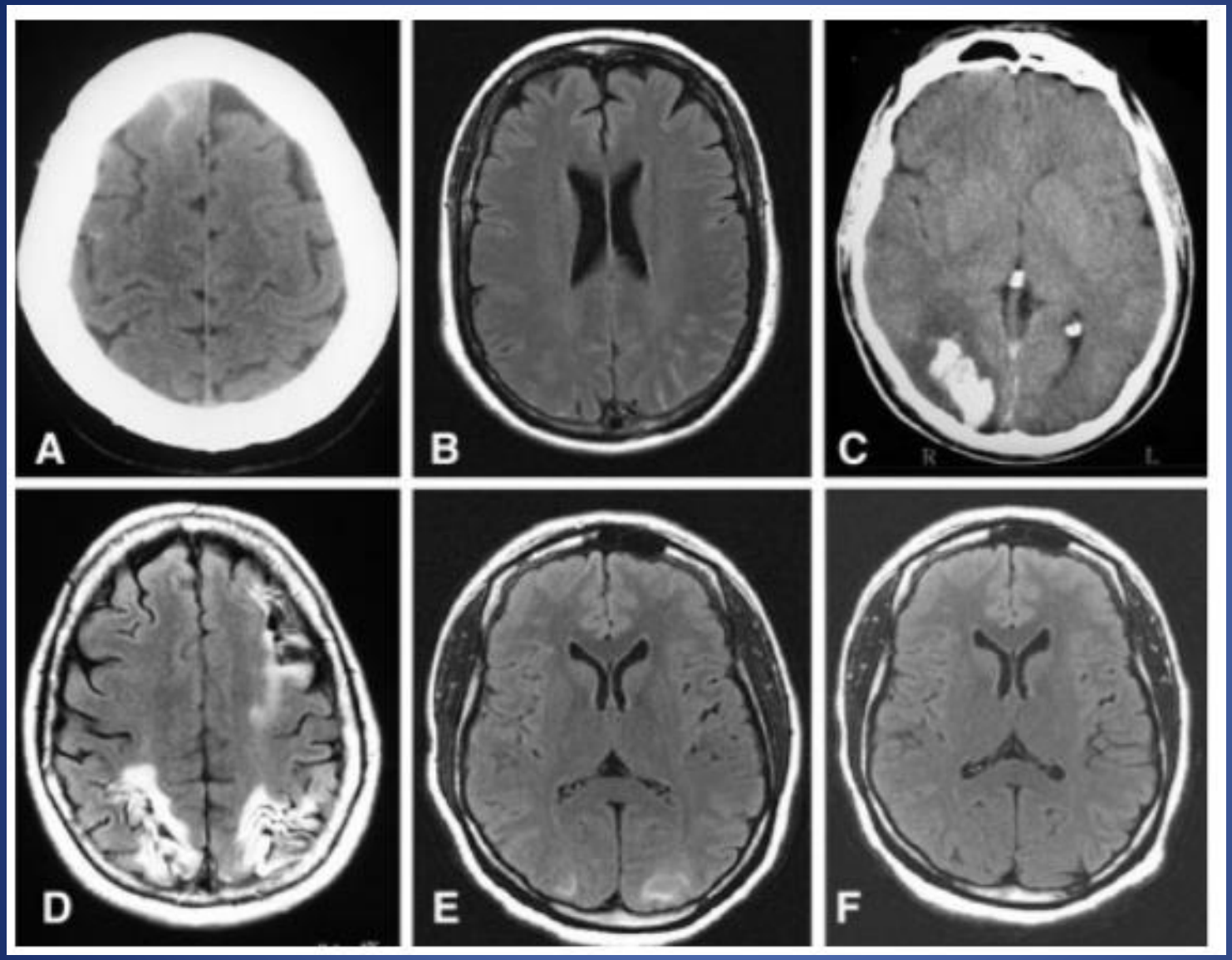
- Renal artery: 80%
- Cervical ICA: 75%
- Vertebral artery: 37%

Soun et al, *Radiol Clin North Amer* 2019

Reversible cerebral vasoconstriction syndrome

Precipitating factors
Postpartum
Postpartum alone[a], postpartum + exposure to drugs[a], eclampsia, preeclampsia
Exposure to drugs, alcohol, medications and blood products
Cannabis[a], cocaine[a], ecstasy, amphetamine derivates, lysergic acid diethylamine
Binge alcohol drinking[b]
Selective serotonin reuptake inhibitors[a]
Nasal decongestants[a], phenylpropanolamine[a], pseudoephedrine[a], ephedrine[a]
Ergotamine tartrate, methergine, bromocriptine[a], lisuride, sumatriptan, isometheptine
Tacrolimus (FK-506), cyclophosphamide, erythropoietin, intravenous immune globulins, red blood cell transfusion, interferon alpha[b]
Nicotine patches[a]
Catecholamine-secreting tumour
Pheochromocytoma, bronchial carcinoid tumour
Miscellaneous
Hypercalcemia, porphyria, head trauma, spinal subdural hematoma, postcarotid endarterectomy, neurosurgical procedures

Ducros et al, *Brain* 2007



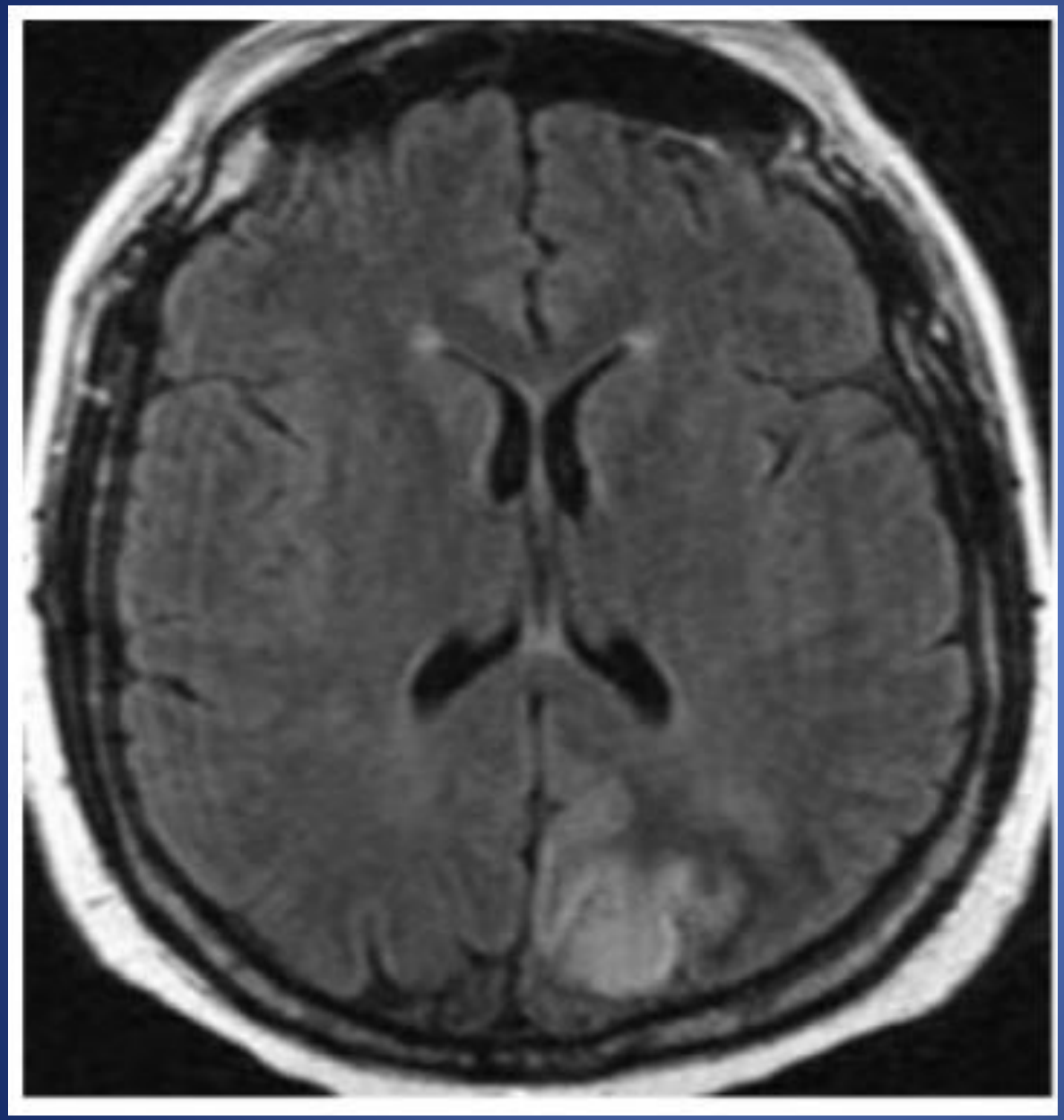
RCVS

Ducros et al, *Brain* 2007

Treatment of RCVS

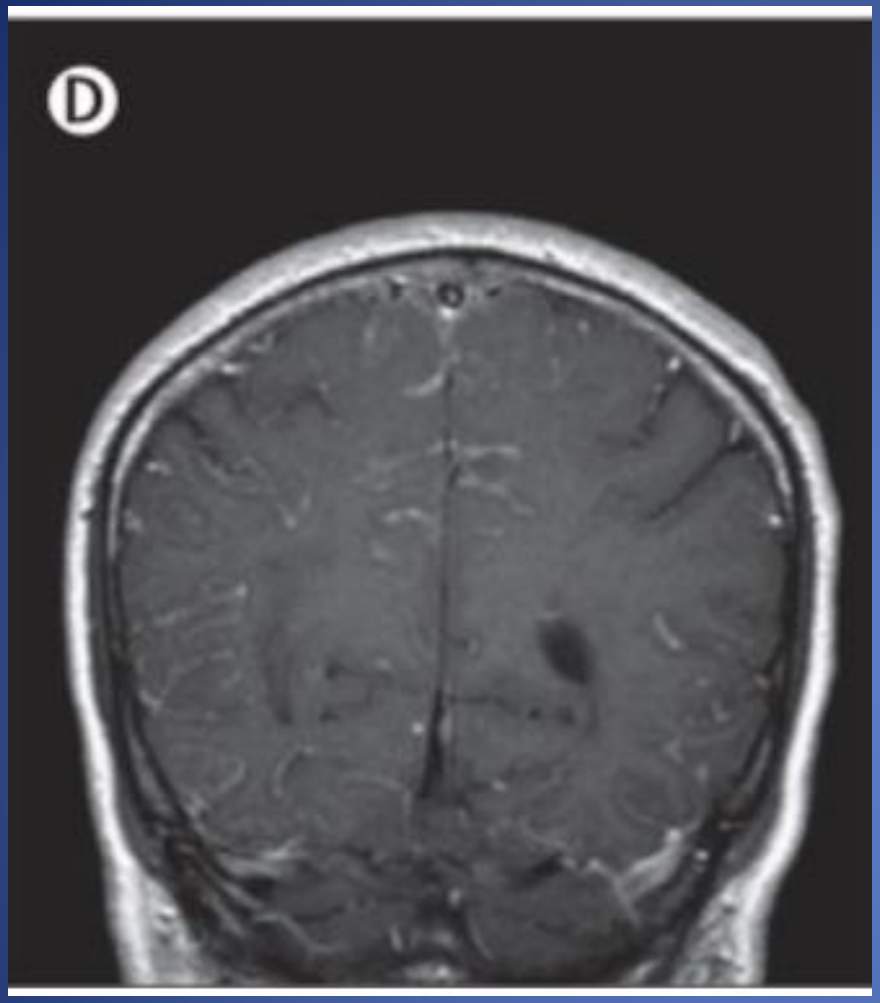
- No RCT's
- Supportive, avoidance of triggers
- Mostly Nimodipine; also Verapamil, Nicardipine, Milrinone, and Magnesium sulfate
- Duration of treatment: 4-12 weeks
- In one study with 162 patients with RCVS, 28% got steroids. Worsening occurred in 37% of patients treated with steroids vs only 5% in patients not treated ($p < 0.001$)[1]

Quitby et al, *Semin Neurol* 2020; 1. Singhal et al, *Neurology* 2017



PCNSV

Abdel Razek et al, *Radiographics*. 2014; Ducros et al, *Brain* 2007



Leptomeningeal enhancement in PCNSV

Treatment of PCNSV

- Study with 112 patients diagnosed with PCNSV
  - Combinations of steroids and immunosuppressant and/or maintenance therapy
  - Overall, 63% of patients achieved prolonged remission without recurrence with avg f/u of 53 months (range 0-198)
  - Maintenance therapy independently associated with no recurrence with OR 8.09

Boysson et al, *Stroke* 2018

PCNSV vs RCVS

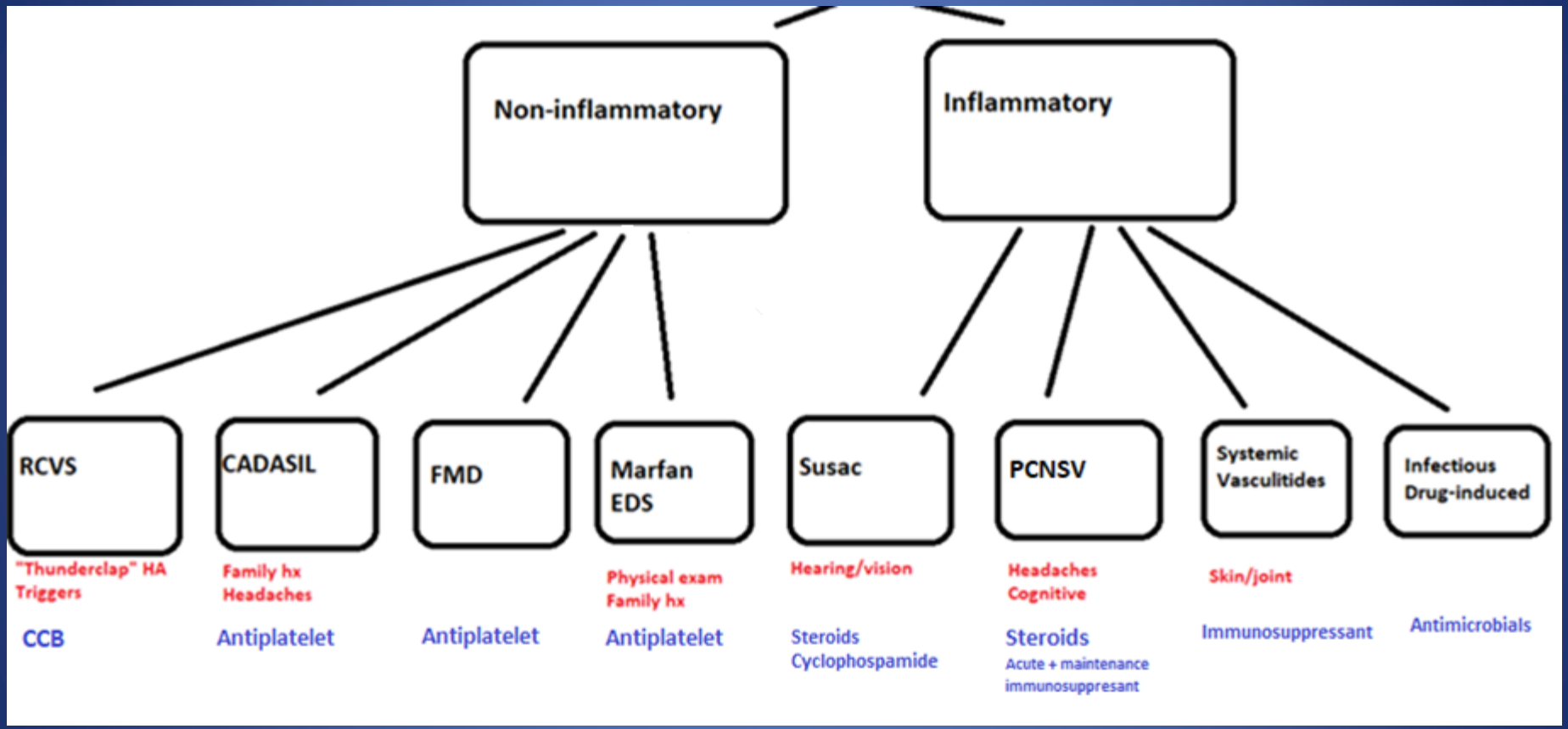
	PCNSV	RCVS
Precipitating factor	None	Post-partum onset or onset after exposure to vasoactive substances
Onset	More insidious, progressive course	Acute onset followed by a monophasic course
Headaches	Chronic and progressive	Acute, thunderclap type
CSF findings	Abnormal (leucocytosis and high total protein concentration)	Normal to near normal
MRI	Abnormal in almost all patients	Normal in 70% of patients
Angiography	Possibly normal; otherwise, diffuse abnormalities are often indistinguishable from RCVS; irregular and asymmetrical arterial stenoses or multiple occlusions are more suggestive of PCNSV; abnormalities might be irreversible	Always abnormal, strings of beads appearance of cerebral arteries; abnormalities reversible within 6–12 weeks
Cerebral biopsy	Vasculitis	No vasculitic changes
Drug treatment	Prednisone with or without cytotoxic agents	Nimodipine

PCNSV=primary CNS vasculitis. RCVS=reversible cerebral vasoconstriction syndrome. CSF=cerebrospinal fluid.

Table 2: **Characteristics of primary CNS vasculitis and reversible cerebral vasoconstriction syndrome**

Salvarani et al, *Lancet* 2012

Summary of clinical signs and treatment:



Questions?

- Call for help anytime!
- http://www.kissnetwork.us/
- email at sslavin2@kumc.edu