

# Spooky Strokes: an overview of rare and atypical cerebral vascular diseases

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# Objectives

- Summarize classic stroke types and mechanisms
- Describe unusual stroke etiologies (including PACNS, RCVS, PRES, Moya moya, CVST)
- Treatment and management of atypical strokes



# TOAST Criteria

- Trial of Org 10172 in Acute Stroke Treatment (TOAST) in 1993
- 5 stroke subtypes
  - Large-artery atherosclerosis
  - Cardioembolic
  - Small-vessel
  - Stroke of other determined etiology
  - Stroke of undetermined etiology

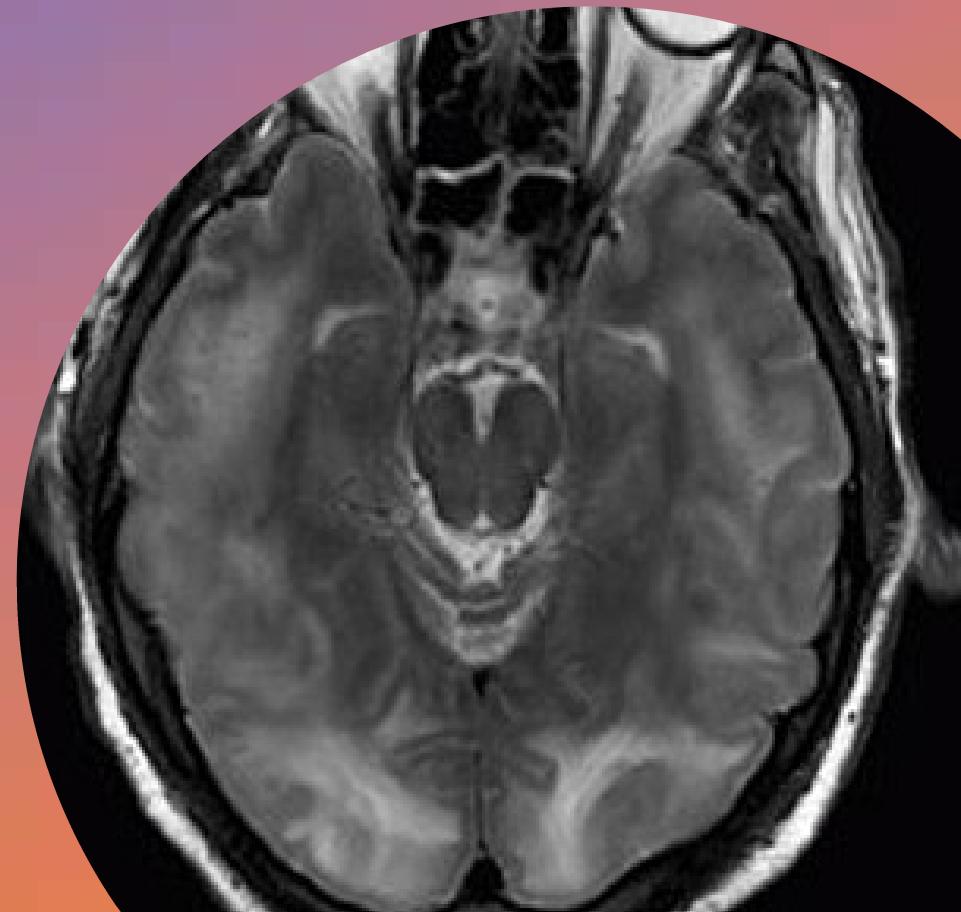
# Certified jump-scare: Atypical strokes

- PRES
- RCVS
- CVST
- Moyamoya
- Cerebral air embolism



-PRIMROSE

- + . Posterior Reversible  
Encephalopathy  
Syndrome (PRES)
- o

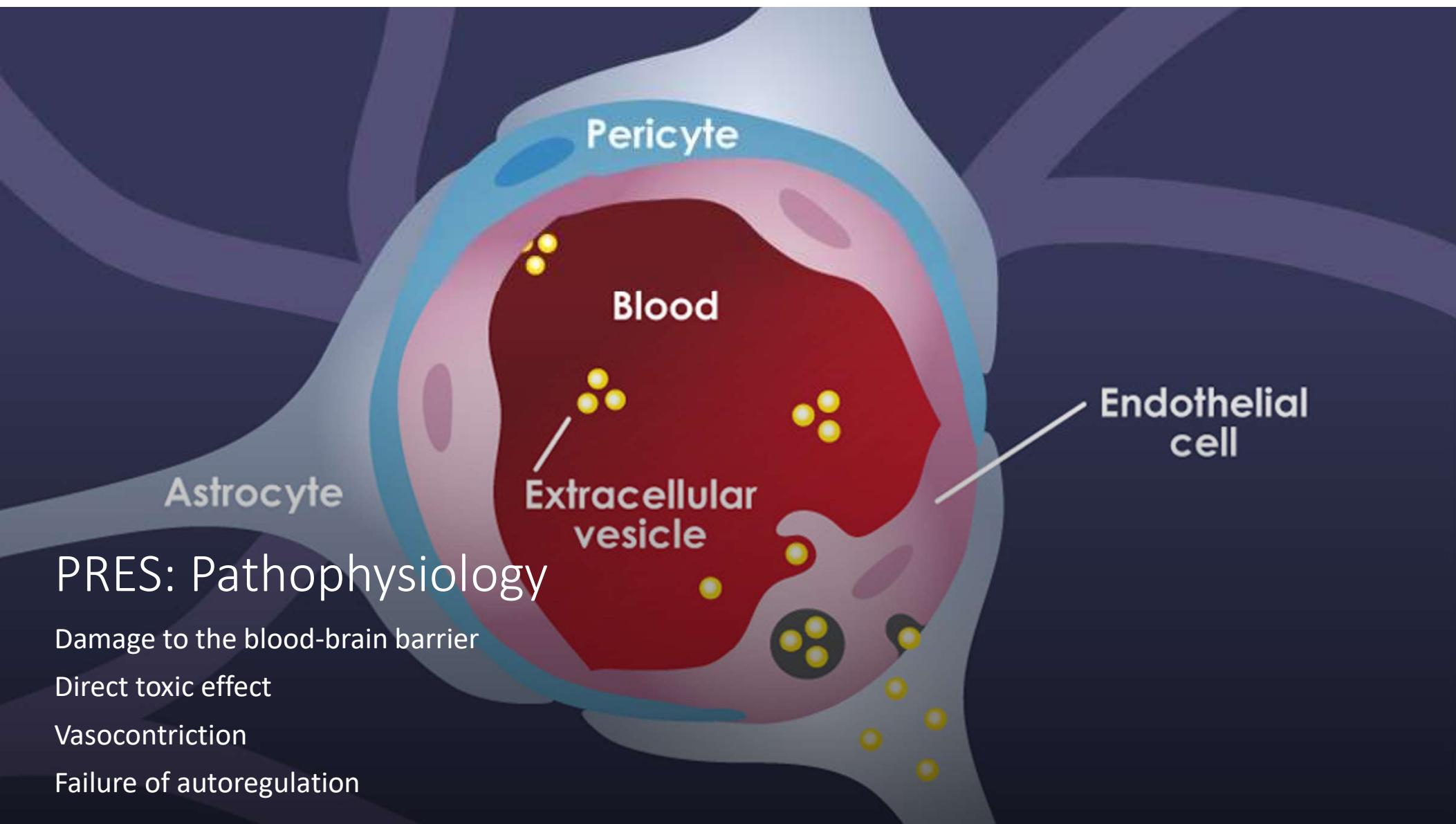


# PRES

- Syndrome of headaches, altered mental status, seizures, and visual loss (cortical blindness)
- Associated white matter changes on imaging
- **Misleading: it may not be "reversible"!!!**

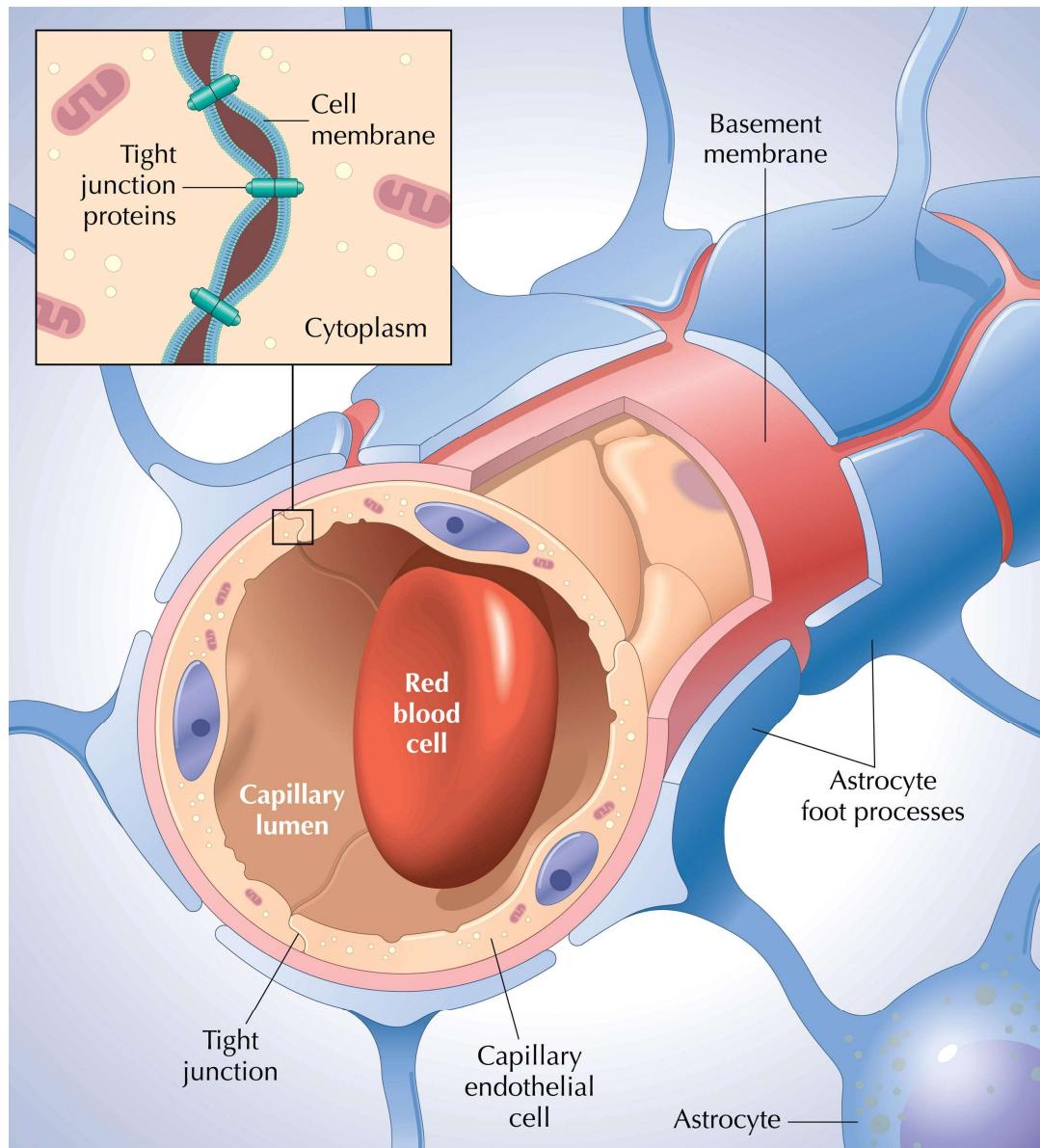
## PRES: Etiology

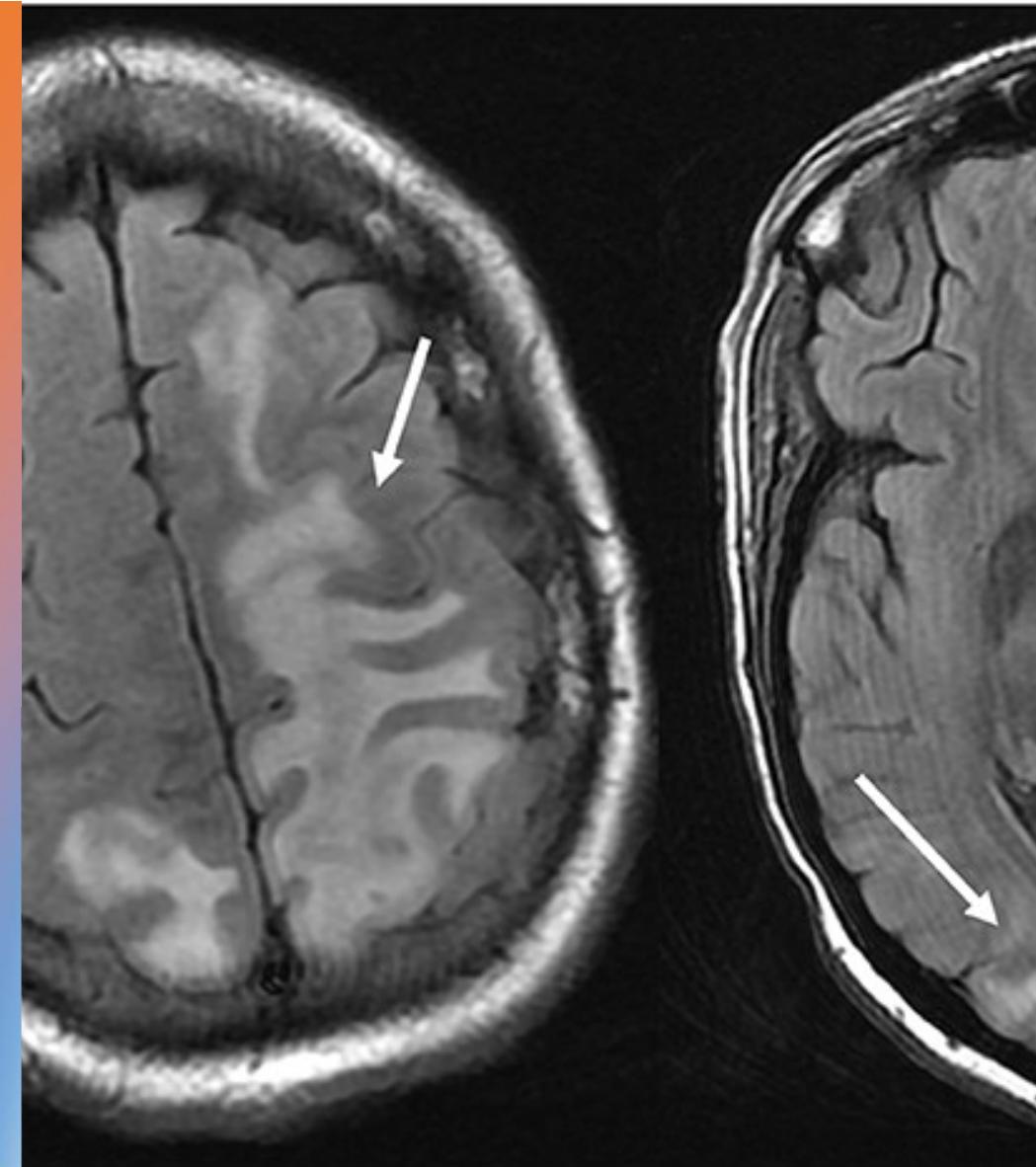
- Hypertension
- Eclampsia
- Immunosuppressive therapy after transplant
- Chemotherapy for cancer
- Sepsis
- Opiate/drug withdrawal, stimulants
- Hypomagnesemia, hyponatremia, hypercalcemia, renal/hepatic dysfunction



## PRES: Pathophysiology

- Damage to the blood-brain barrier
- Direct toxic effect
- Vasoconstriction
- Failure of autoregulation





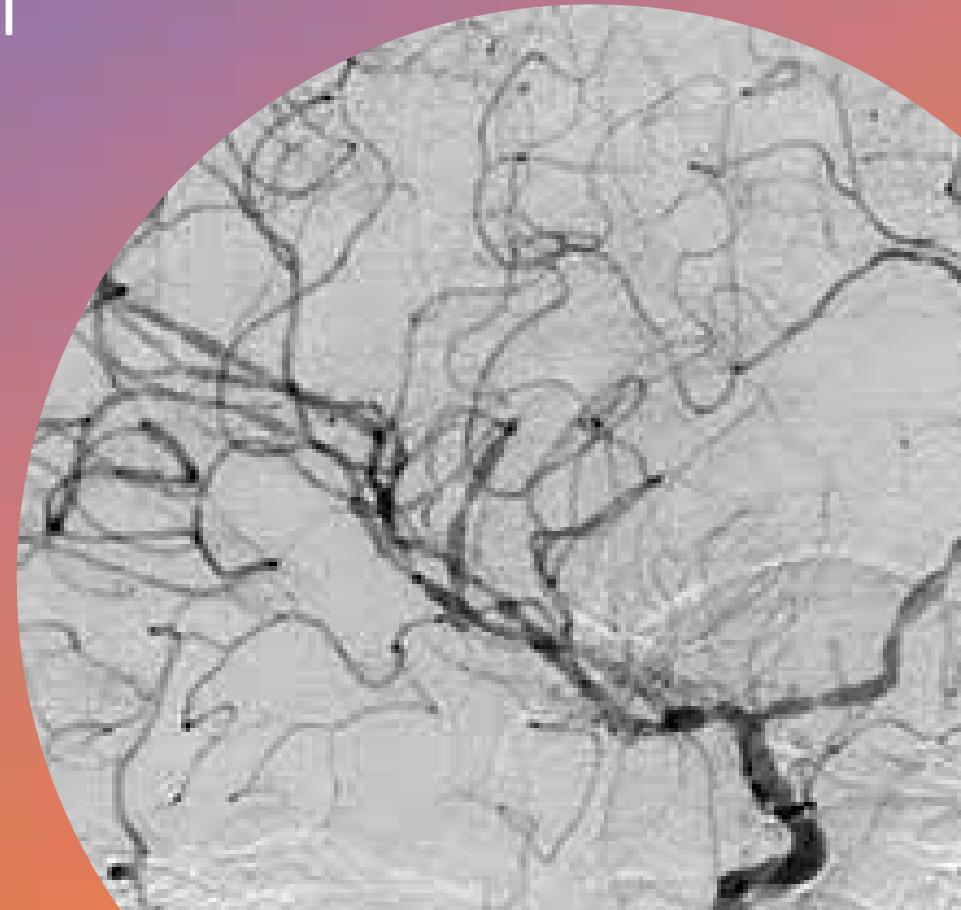
## PRES: Imaging

- MRI w/o contrast is gold-standard
- 3 patterns
  - **Parietal-occipital**
  - Superior frontal sulcus pattern
  - Holohemispheric watershed pattern

## PRES: Treatment

- Usually require ICU admission
- Lower BP no more than 20% in first 24 hours
  - Clevipipine, Nicardipine, sodium nitroprusside gtts (fast-on/fast-off, easy to titrate), Labetalol PRN
- Treat seizures with anti-seizure medications
- Pre-eclampsia – delivery
- Sepsis – treatment of infectious source
- Discontinue offending agent

- # Reversible Cerebral Vasoconstriction Syndrome (RCVS)



## RCVS

- Recurrent thunderclap headaches (screaming, agitation, collapse)
- Trigger factors: orgasm, physical exertion, acute stress/strong emotion, sneezing, swimming
- Ischemic and hemorrhagic strokes (non-aneurysmal SAH)
- Multifocal reversible narrowing of cerebral arteries
- Monophasic course – most resolve in ~3 months
- 2:1 to 10:1 female-to-male ratio
- Peak age is 42 years

## RCVS2 Score

- Range –1 to +10
- Distinguishes patients with RCVS from other intracranial arteriopathies

## RCVS<sub>2</sub> Score for Reversible Cerebral Vasoconstriction Syndrome ★

Distinguishes reversible cerebral vasoconstriction syndrome from other intracranial arteriopathies at admission.

### INSTRUCTIONS

To use the score, patients must have a first presentation of abnormal intracranial vascular imaging and be 18–55 years old. The score has not been tested outside this age range.

	When to Use ▾	Pearls/Pitfalls ▾	Why Use ▾
Recurrent or single thunderclap headache	Absent 0	Present +5	
Intracranial carotid artery involvement	Not affected 0	Affected -2	
Vasoconstrictive trigger identified	No 0	Yes +3	
Sex	Male 0	Female +1	
Subarachnoid hemorrhage present on imaging	Absent 0	Present +1	

## RCVS: Etiology

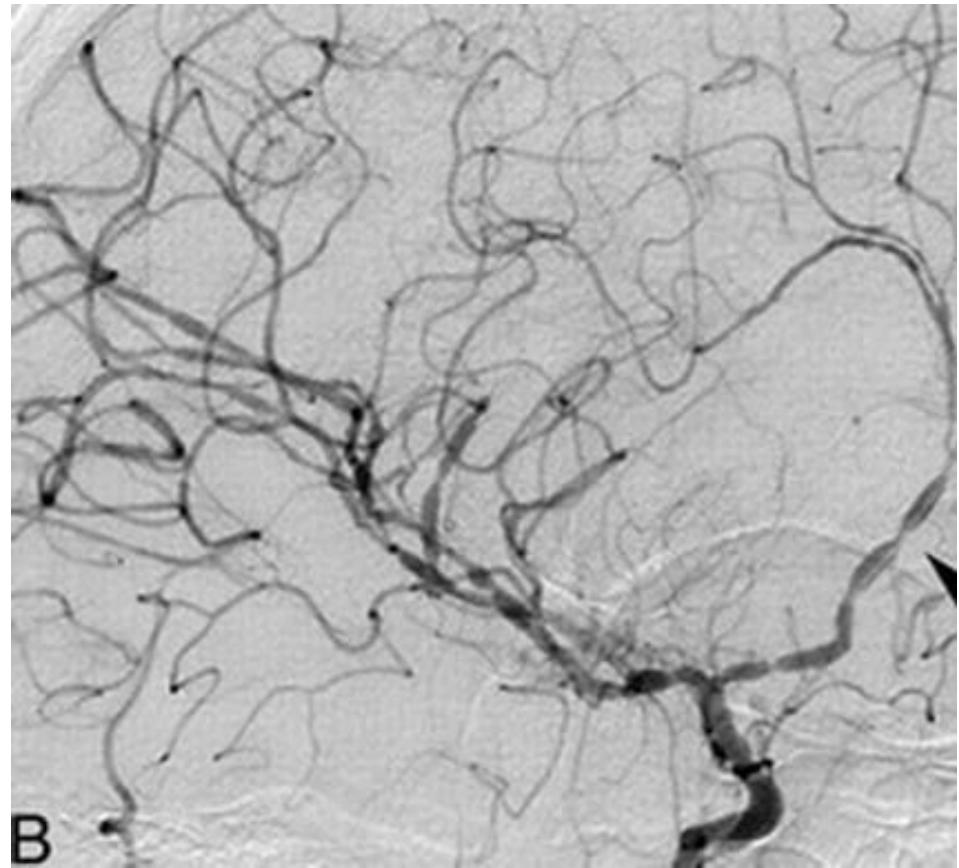
- Idiopathic
- Medications, drugs, blood products
  - Amphetamine, cocaine, marijuana, ecstasy, LSD
  - SSRIs, chemotherapy, oral contraceptives, pseudoephedrine
  - RBC transfusion
- Pregnancy
  - Eclampsia, pre-eclampsia, delayed post-partum eclampsia
- Miscellaneous

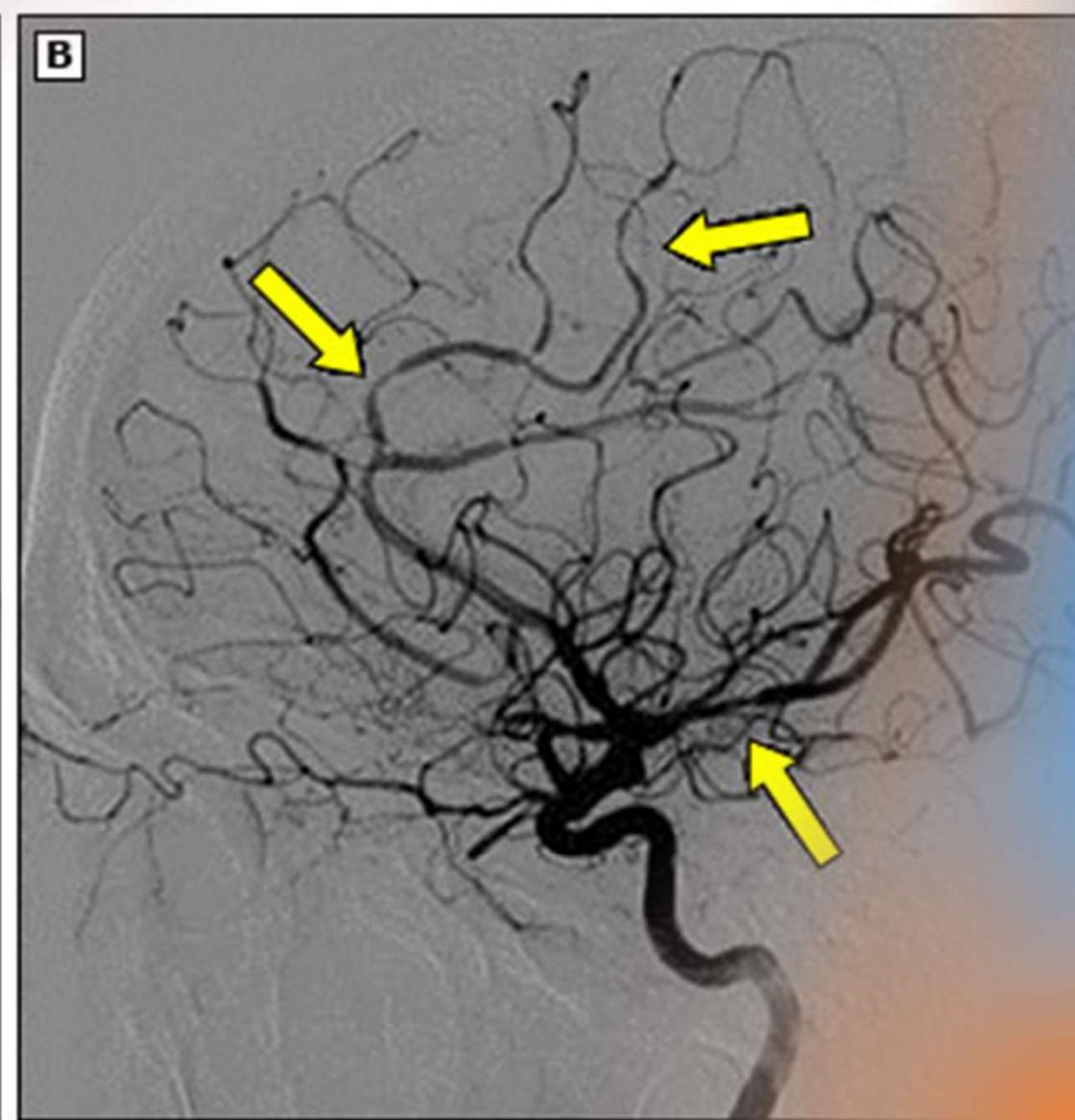
## RCVS: Pathophysiology

- Transient disturbance in cerebral arterial tone
- Sympathetic overactivity

## RCVS: Imaging

- "Sausage on a string" appearance of cerebral arteries
- Convexity SAH





## RCVS: Treatment

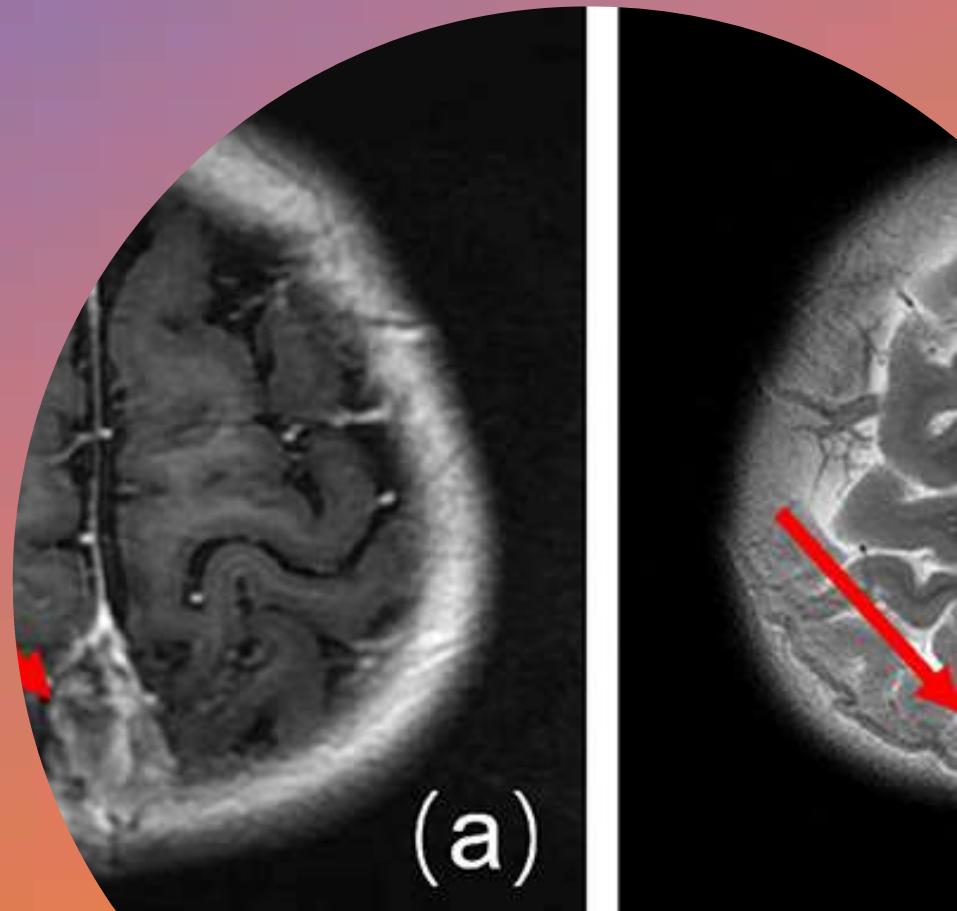
- Nimodipine 4-12 weeks, Verapamil, and magnesium sulfate
- Intra-arterial Verapamil in single location reverses vasoconstriction in multiple arteries – diagnostic and therapeutic
- Benzodiazepines may be helpful to reduce agitation
- Self-limited course – observation and symptomatic management

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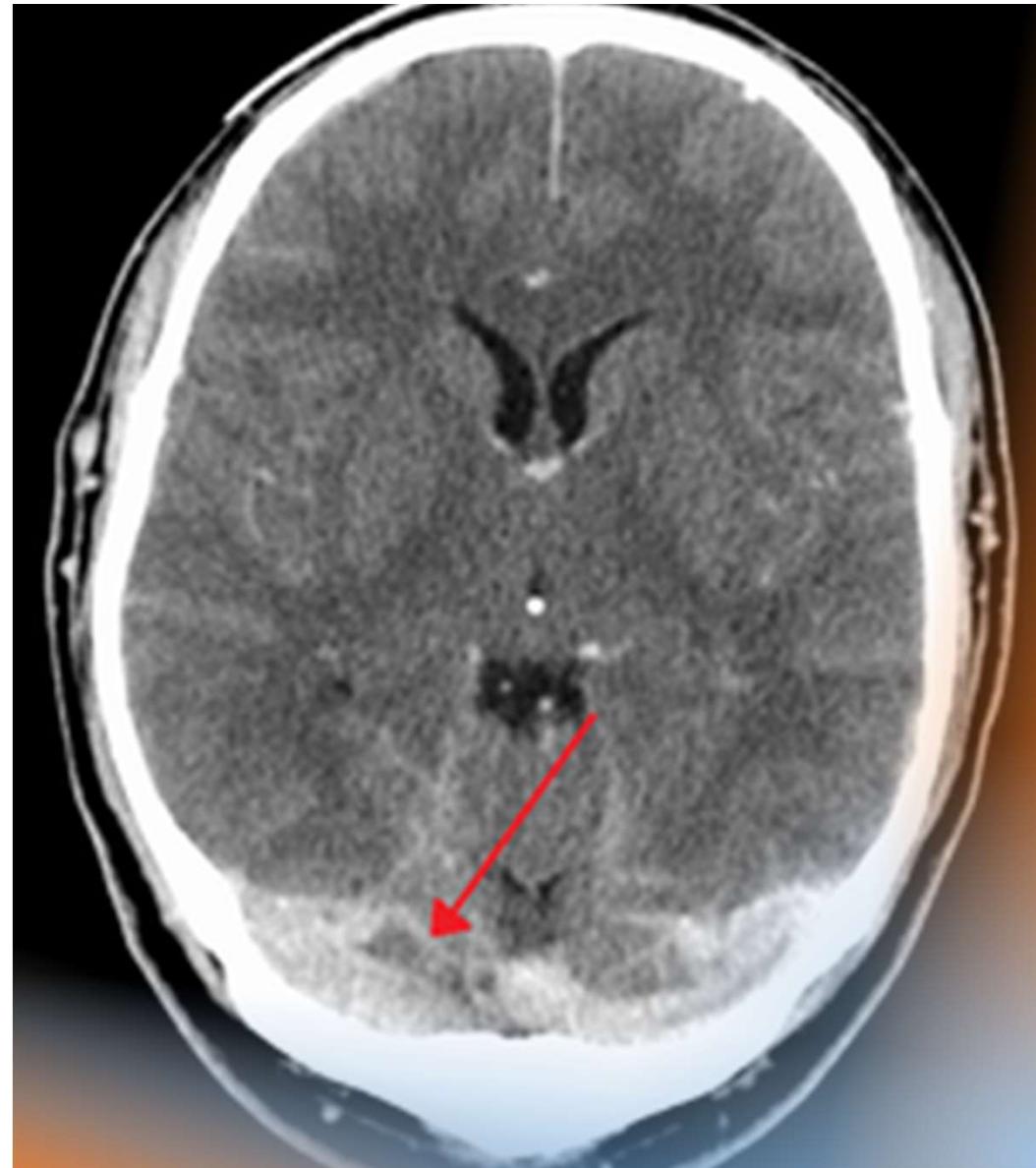
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# Cerebral Venous Sinus Thrombus (CVST)



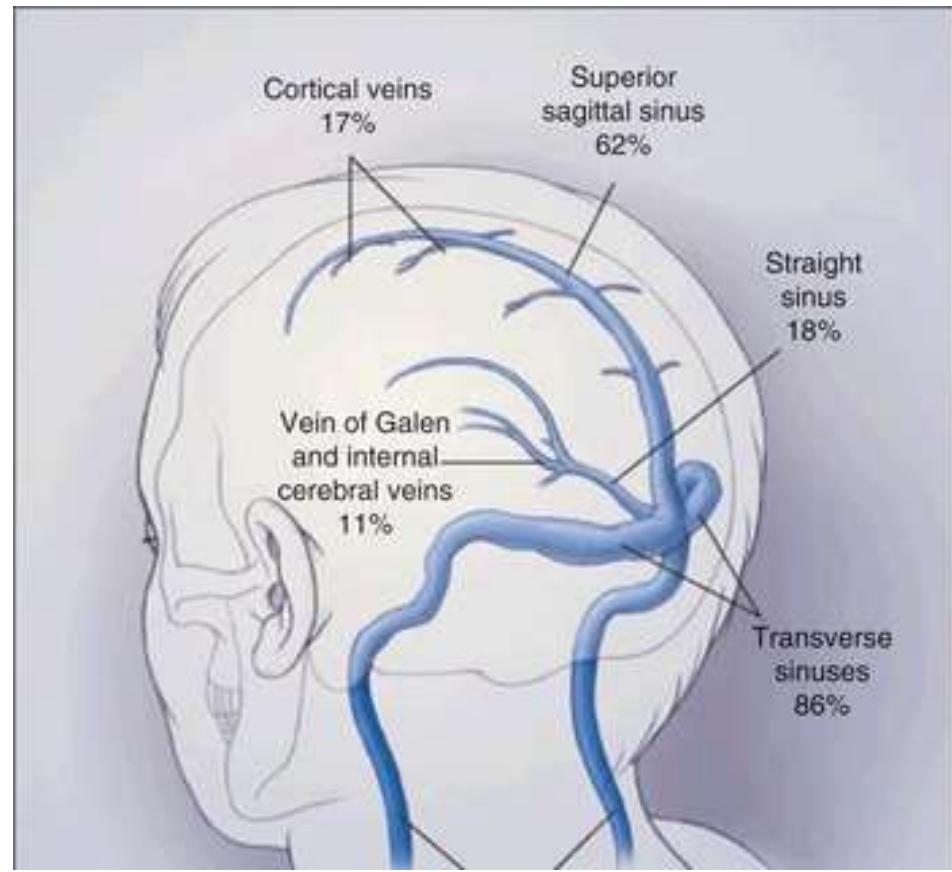
## CVST

- Clot (thrombus) in venous sinuses
- Present with headache, seizures, altered mental status, cranial nerve palsies
- Intracranial venous hemorrhages
- 2.9:1 female-to-male ratio
- Median age 37 years



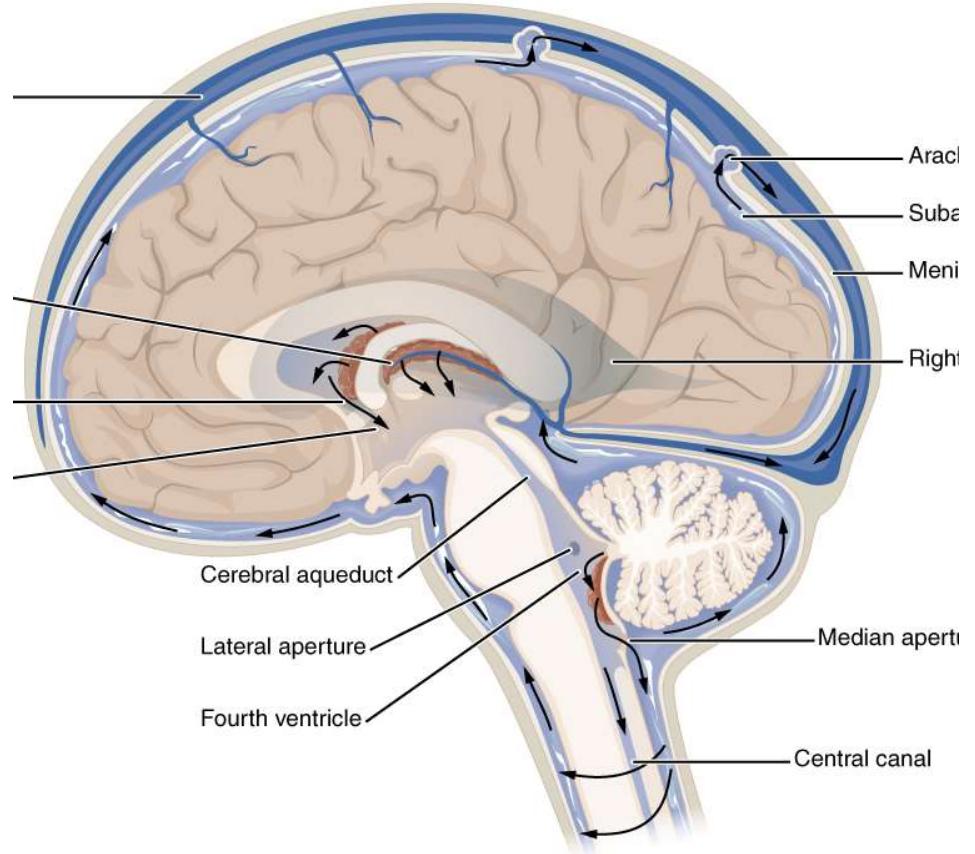
# CVST

- Cerebral venous system



## CVST

- Arachnoid granulations drain CSF into the venous system



## CVST: Etiology

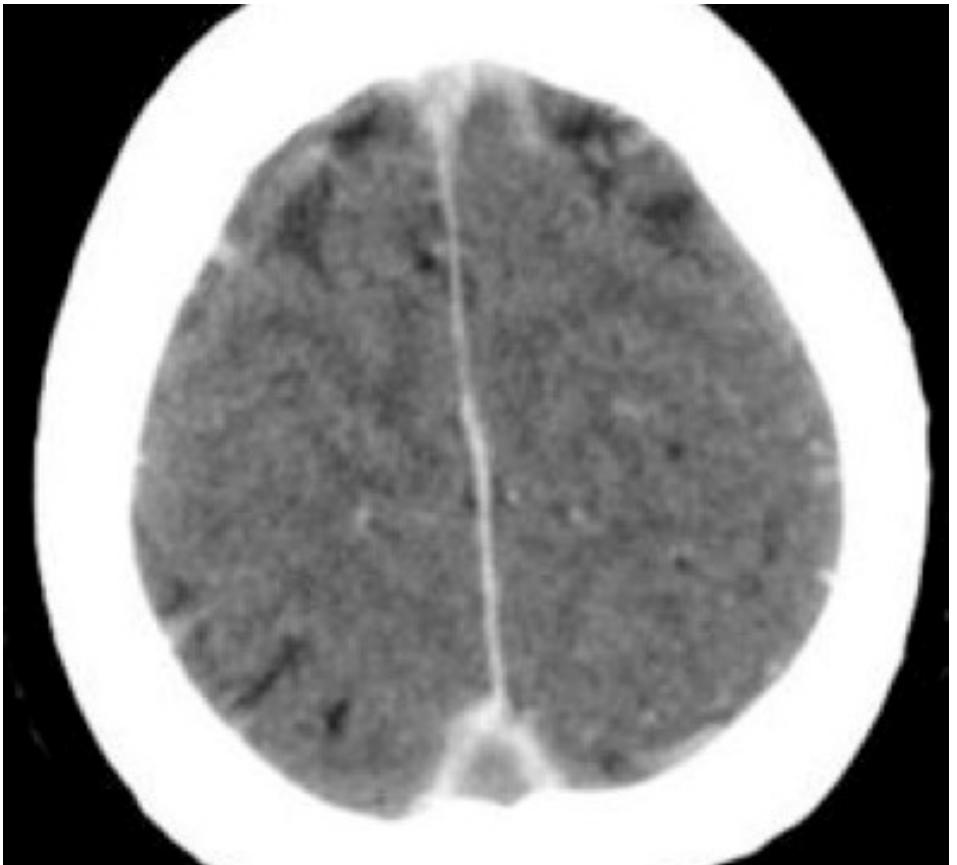
- Occlusion with disruption of venous drainage
- Venous congestion
- Increased venous pressure leads to venous or capillary rupture
- Brain hemorrhage
- Impaired CSF circulation can cause intracranial hypertension
- Cytotoxic and vasogenic edema

# CVST: Etiology

- Sinus infections/meningitis
- Pregnancy
- Hypercoagulability: malignancy
- Dehydration
- Drugs: Oral contraceptives, Sildenafil
- Head trauma
- Lumbar puncture
- Diabetic ketoacidosis

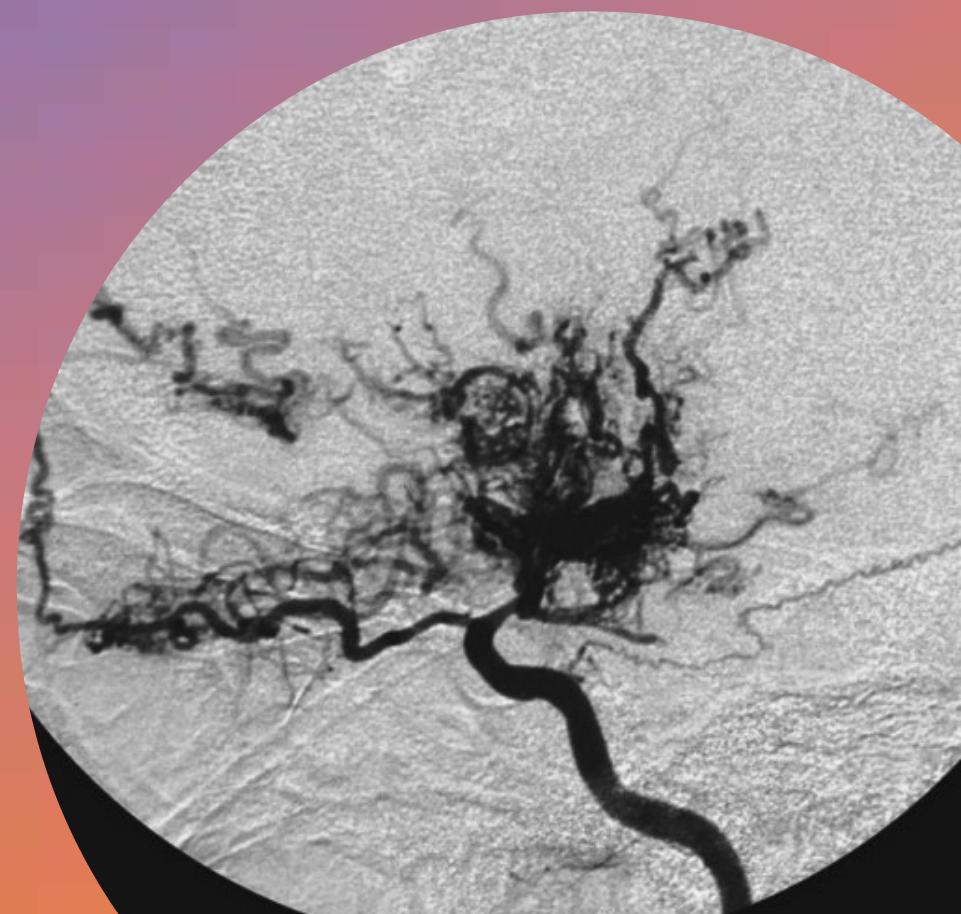
## CVST: Imaging

- CTA/CTV
- MRA/MRV
- Empty delta sign



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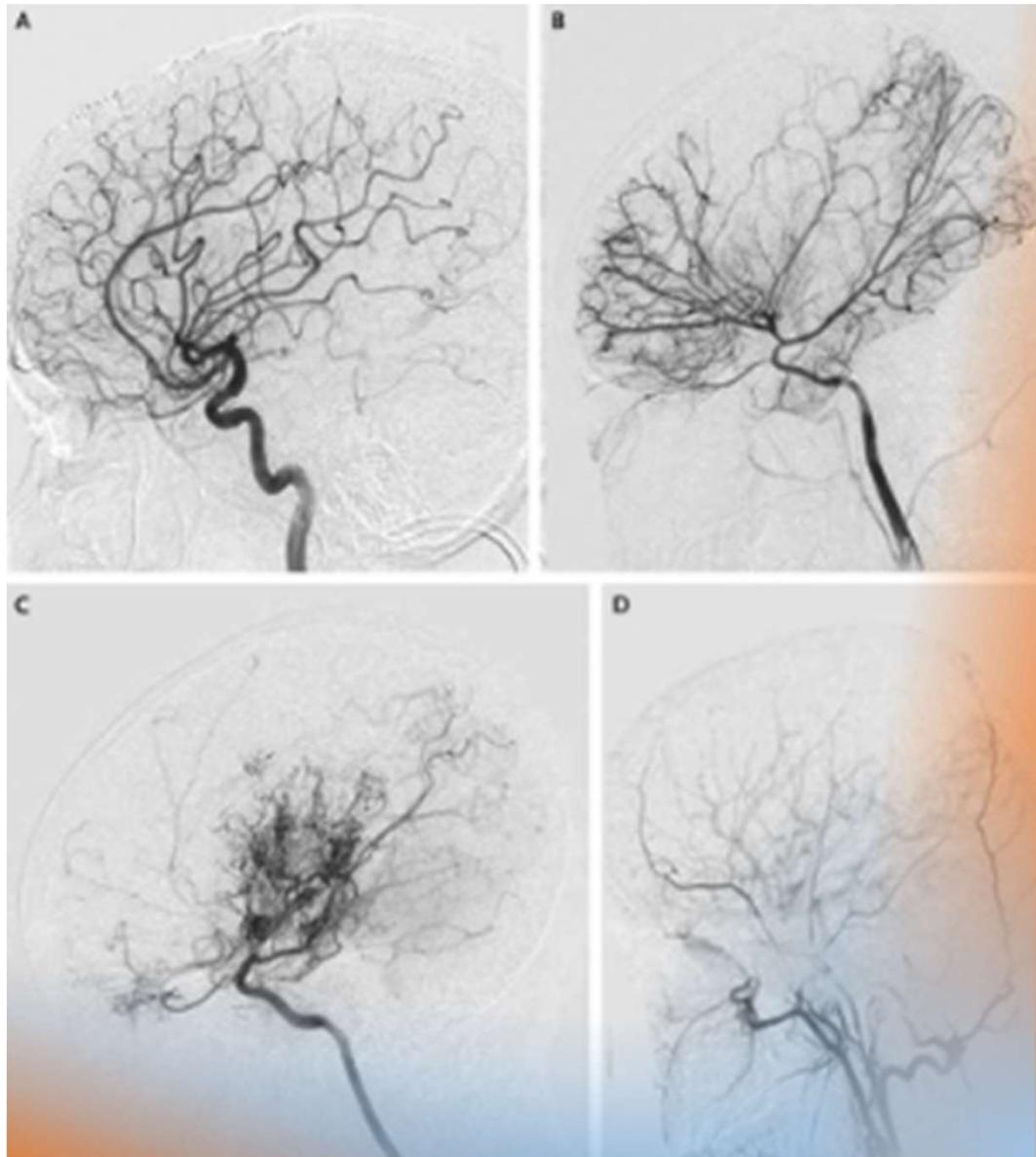
# Moyamoya





## Moyamoya

- **Chronic progressive** bilateral occlusion/stenosis of the terminal internal carotid arteries (ICA)
- Abnormal vascular network
- Two general types of stroke:
  - Ischemic from stenosis
  - Hemorrhagic from fragile collaterals
- 1.8:1 female-to-male ratio
- Two peaks: ~5 years and ~40 years



# Moyamoya: Etiology

- Moyamoya disease vs moyamoya syndrome
- Moyamoya disease – ***no*** associated risk factors
- Moyamoya syndrome – well-recognized associated conditions

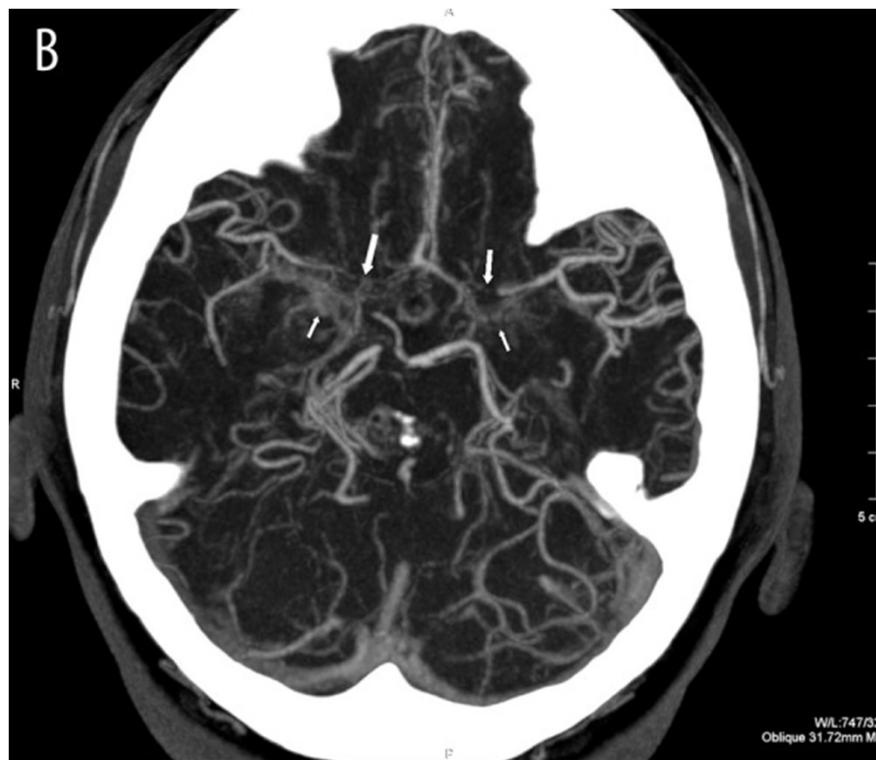
# Moyamoya: Etiology

<b>Current causes of acquired MMS</b>	<b>Key diagnosis features</b>
Cephalic or neck irradiation <sup>112</sup>	Interview
Skull base tumor <sup>111</sup>	Imaging of the skull base
Atherosclerosis of skull base arteries	Complete imaging of cervical arteries
Chronic meningitis (especially tuberculosis meningitis), cerebral vasculitis <sup>110</sup>	Cerebrospinal fluid analysis
Autoimmune angiitis <sup>113</sup>	Autoimmune blood tests (anti-nuclear antibodies)
Prothrombotic disorders <sup>114</sup>	Antithrombin, protein C/protein S, activated protein C resistance, factor V Leiden mutation, prothrombin mutation
<b>Current causes of inherited MMS</b>	
Sickle cell disease or trait <sup>136–138</sup>	Hemoglobin electrophoresis (if from African or Caribbean origin)

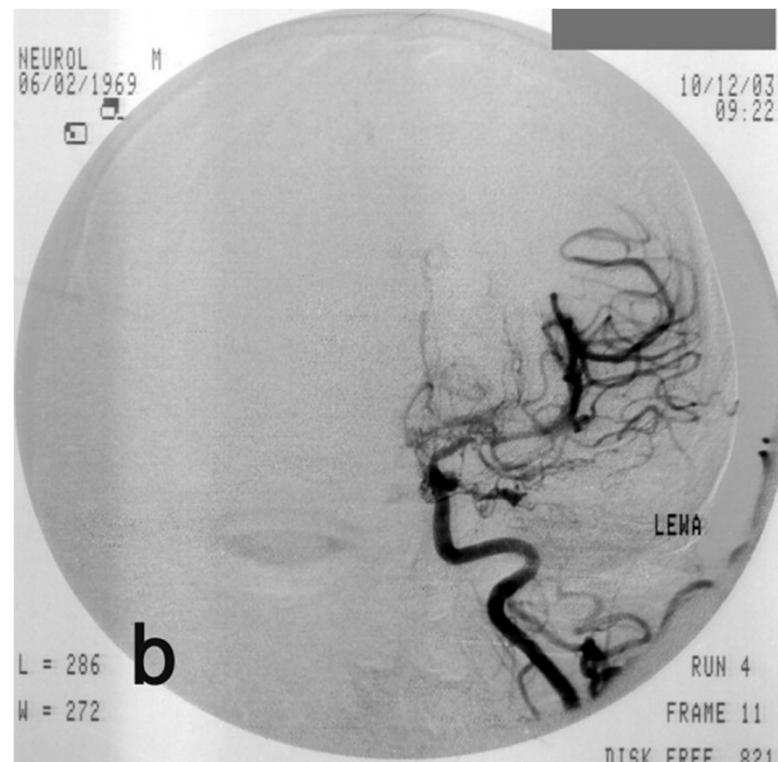
# Moyamoya: Imaging

- "Puff of smoke" = *moyamoya* in Japanese
- CTA and MRA
- Small, twisting wide vessels penetrating toward base of brain

# Moyamoya: Imaging



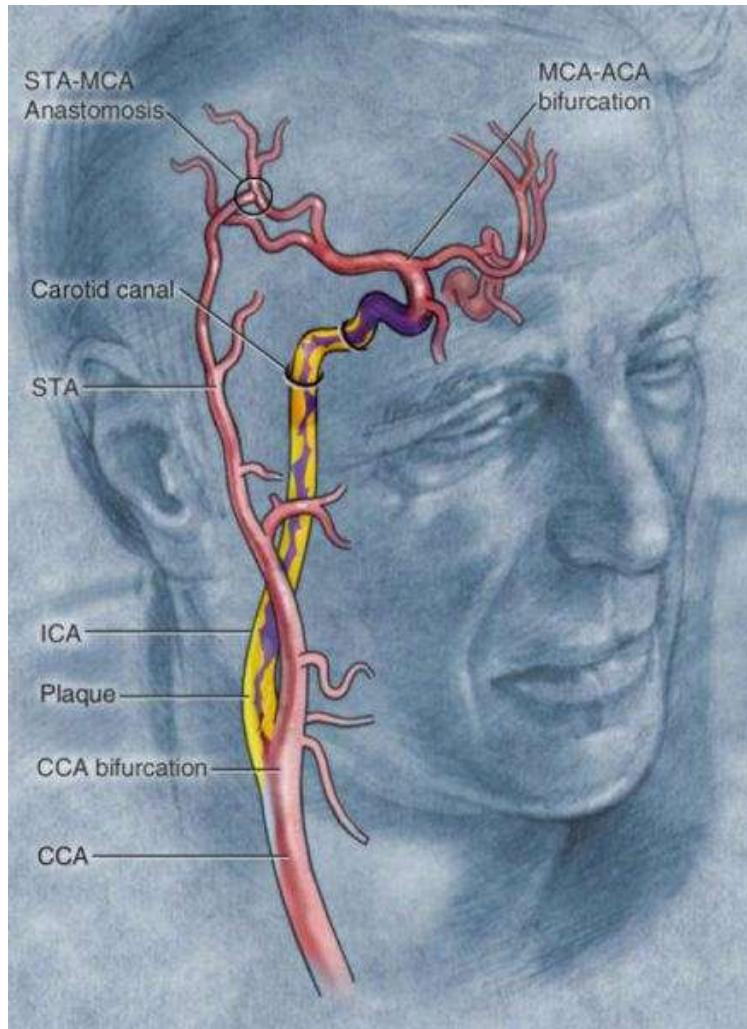
# Moyamoya: Imaging



# Moyamoya: Treatment

- EC/IC bypass
- Superficial temporal artery to middle cerebral artery (STA-MCA) bypass



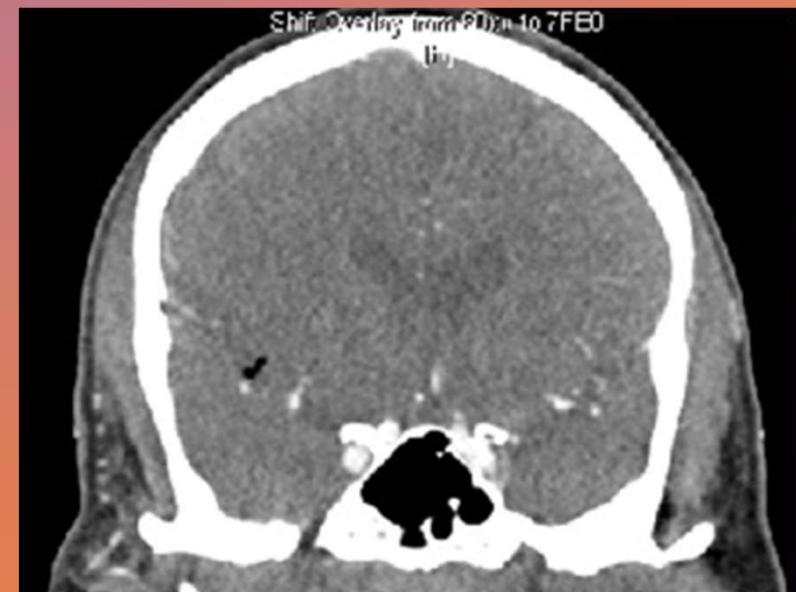


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# Cerebral Air Embolism



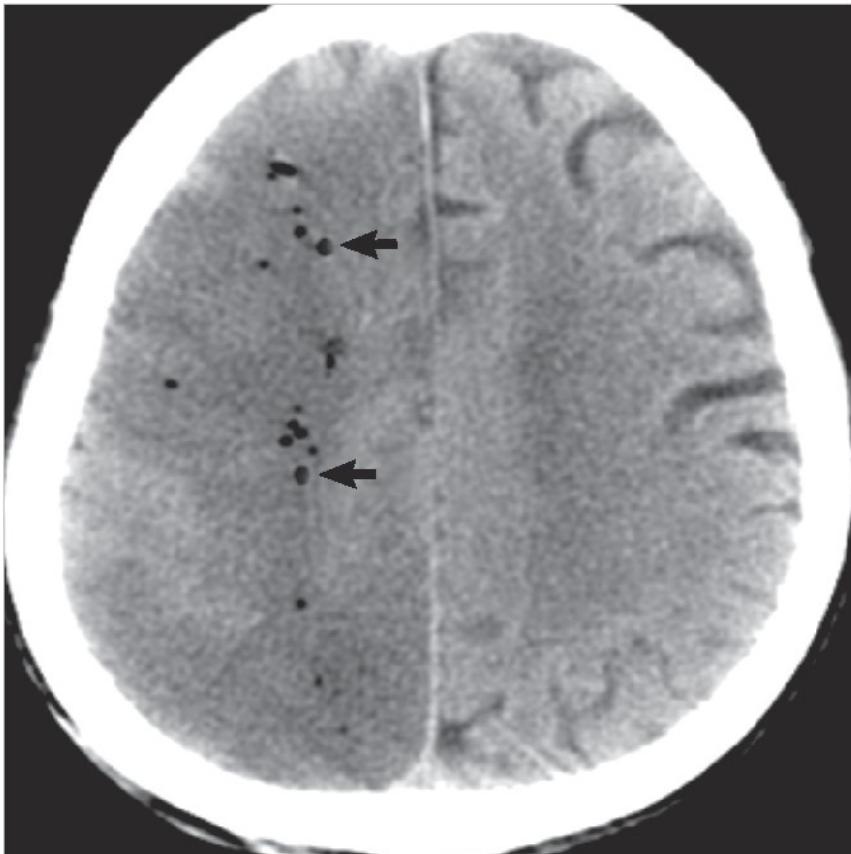


Fig. 2. Immediate non-contrast brain CT showed an extensive lesion with low attenuation

## Cerebral Air Embolism

- Air enters the venous or arterial circulation, and travels to the brain
- Biggest medical source is air in lines
- May be fatal
- Associated with long-term cognitive deficits

# Cerebral Air Embolism: Etiology

- Endovascular therapies
- Dialysis
- Tracheal-esophageal fistula (radiofrequency ablation for Afib)
- Contrast injection during radiographic procedures
- Disruption of venous system with central venous catheters
- Right (venous) to left (arterial) shunt, e.g. PFO
- Scuba diving
- Positive pressure ventilation
- Insufflation during laparoscopic surgeries



## Cerebral Air Embolism

- Venous air embolism
  - Pulmonary pressure increase with cardiovascular collapse
- Arterial air embolism/paradoxical arterial air embolism

# Cerebral Air Embolism: Pathophysiology

- Air in the arterial system can induce platelet aggregation by triggering release of plasminogen-activator inhibitor
- Release of cytokines
- Activation of vascular endothelium
- Microthrombi, end-organ damage

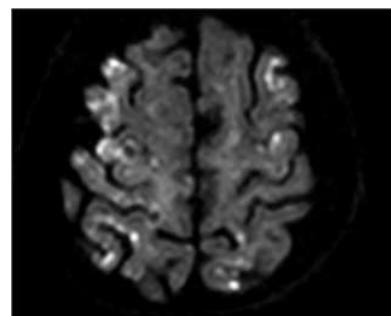
## Cerebral Air Embolism: Imaging

- Non-contrast CT head
- High convexity along the cortical grooves
- Sulcal subarachnoid gyriform pattern

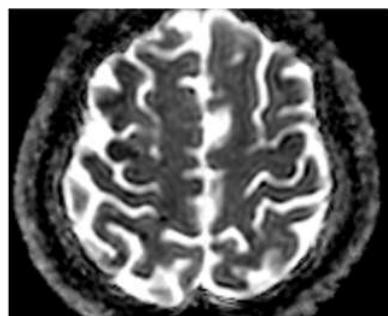


# Cerebral Air Embolism: Imaging

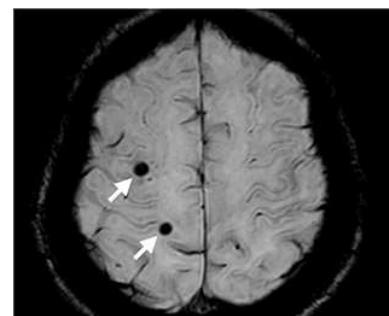




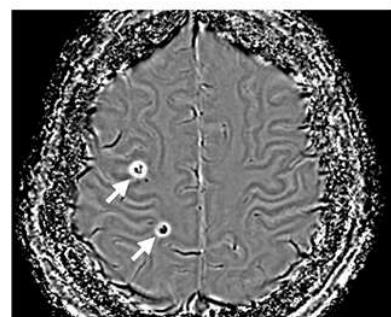
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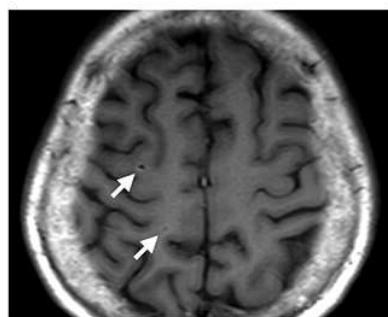
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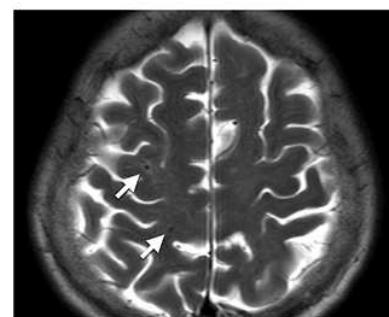
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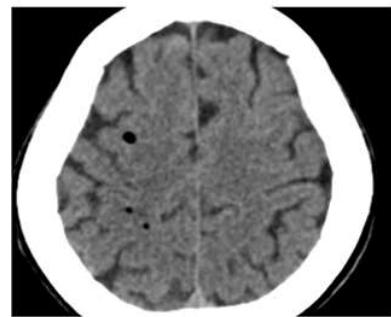
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# Cerebral Air Embolism: Treatment

- Zero-tolerance policy for air in lines
- Hyperbaric oxygen chamber
  - Increases arterial partial pressure of oxygen helps dissolve oxygen into plasma and increases its diffusion into the tissue
- Endovascular aspiration

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