

UW Nuclear Medicine Clinical Case Conference – DS1226 CME Disclosure Statement



I, Dr. *Yuyang Zhang* have no financial relationships to disclose.

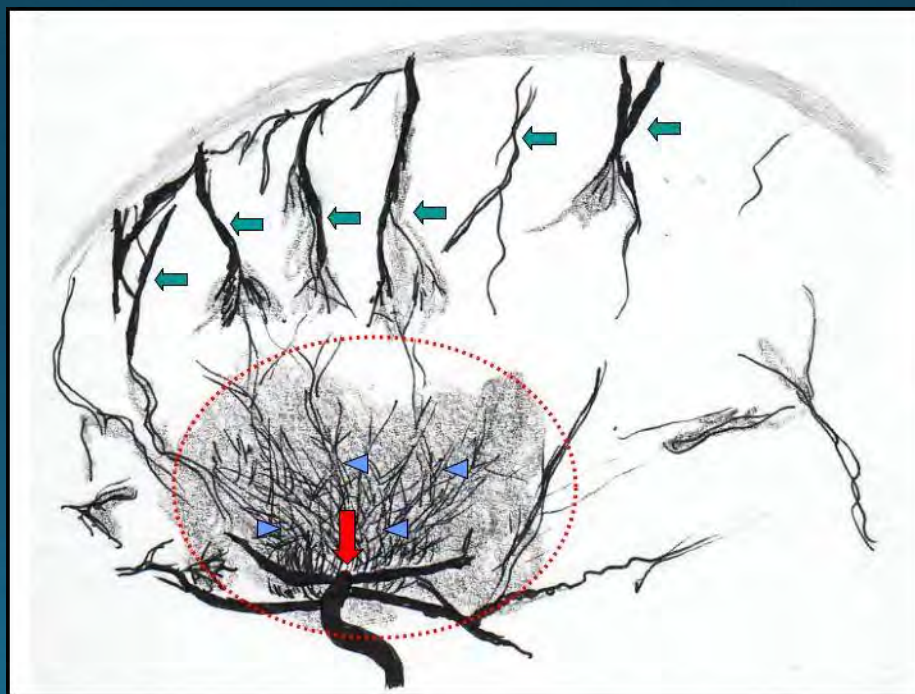
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Moyamoya disease

- What the meaning of Moyamoya?

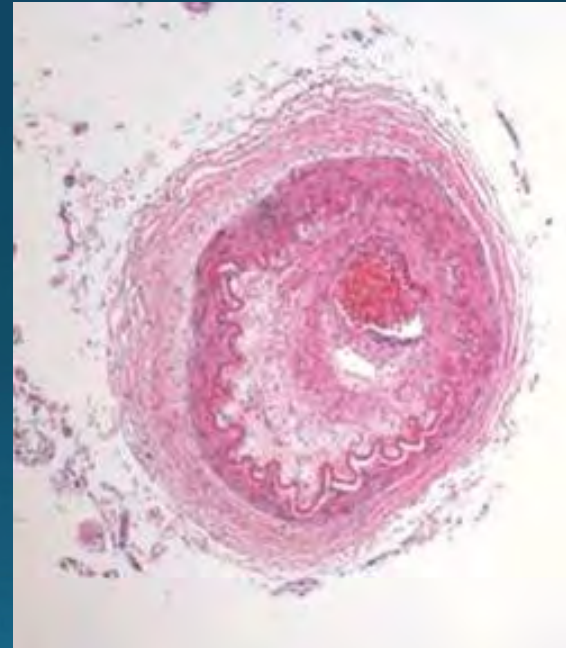
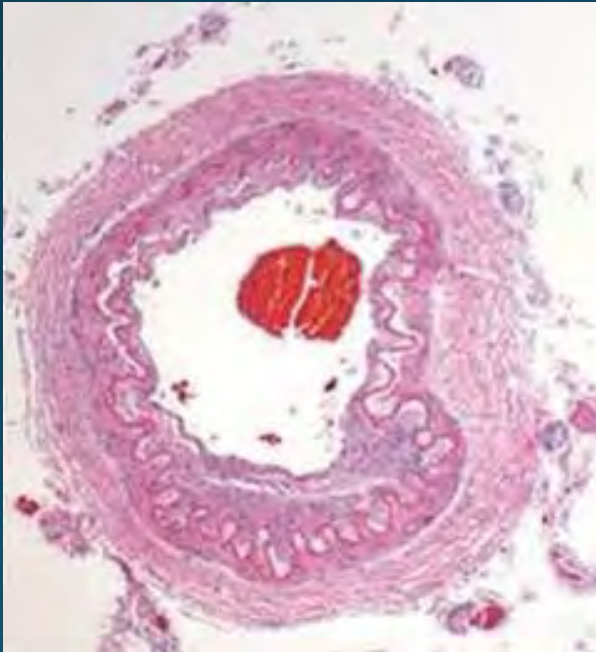
- It is Japanese.
- Angio have the appearance of a "puff of smoke" ("もやもや (moyamoya)" in Japanese) due to small collaterals



Jiro Suzuki, Akira Takaku. Cerebrovascular "Moyamoya" Disease: Disease Showing Abnormal Net-Like Vessels in Base of Brain. Arch Neurol. 1969;20(3):288-299.

Moyamoya disease

- progressive cerebrovascular disorder caused by blocked arteries in the basal ganglia, primarily in the internal carotid artery
- primarily affects children, also occur in adults
- the first symptom of Moyamoya disease: stroke, or recurrent transient ischemic attacks
- disturbed consciousness, speech deficits (usually aphasia), sensory and cognitive impairments, involuntary movements, and vision problems
- inherited genetic abnormalities or acquired

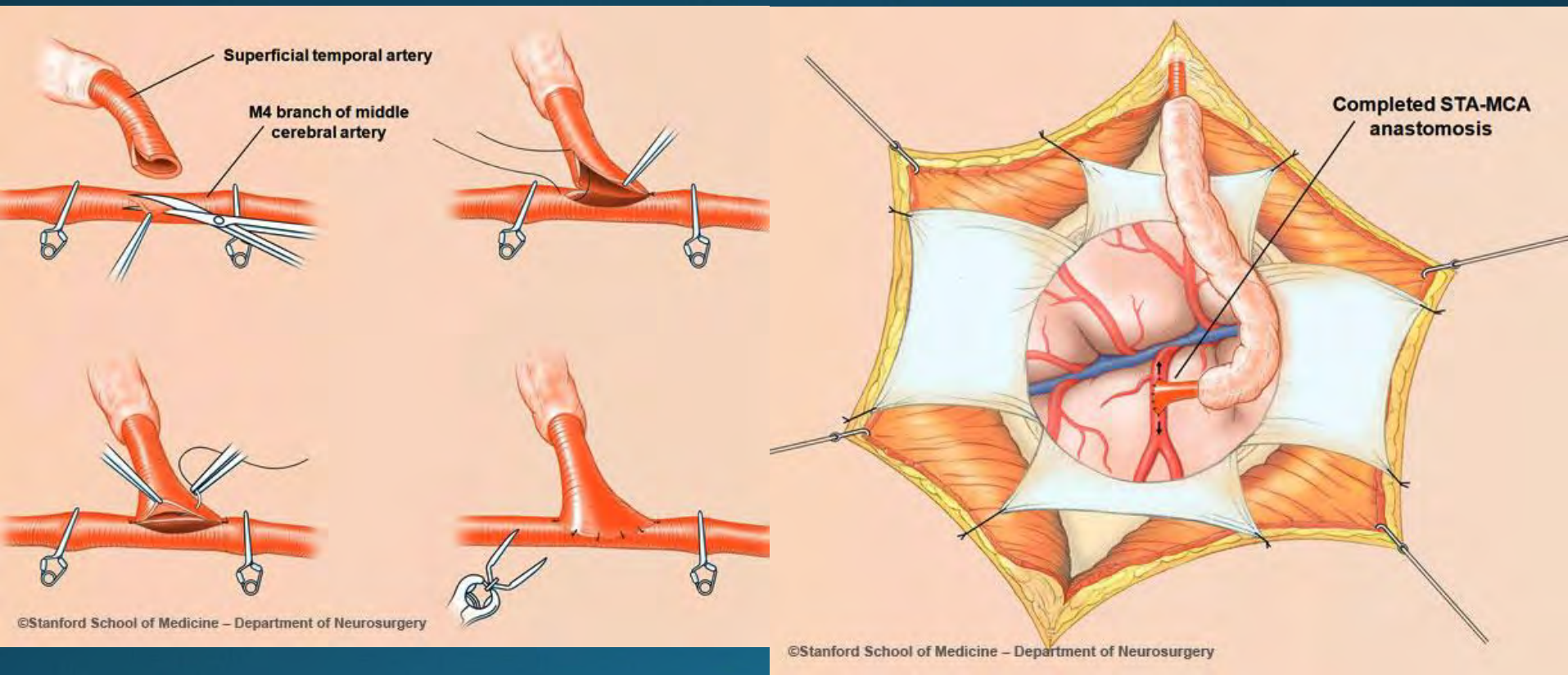


From Stanford moyamoya surgery website

treatments

- The same concept: a blood and oxygen "starved" brain reaching out to grasp and develop new and more efficient means of bringing blood to the brain and bypassing the areas of blockage
- **Direct revascularization.**
 - **STA-MCA** procedure (the treatment of choice) - the scalp artery (superficial temporal artery or STA) is directly sutured to an artery on the surface of the brain (middle cerebral artery or MCA)

STA-MCA bypass surgery illustrations

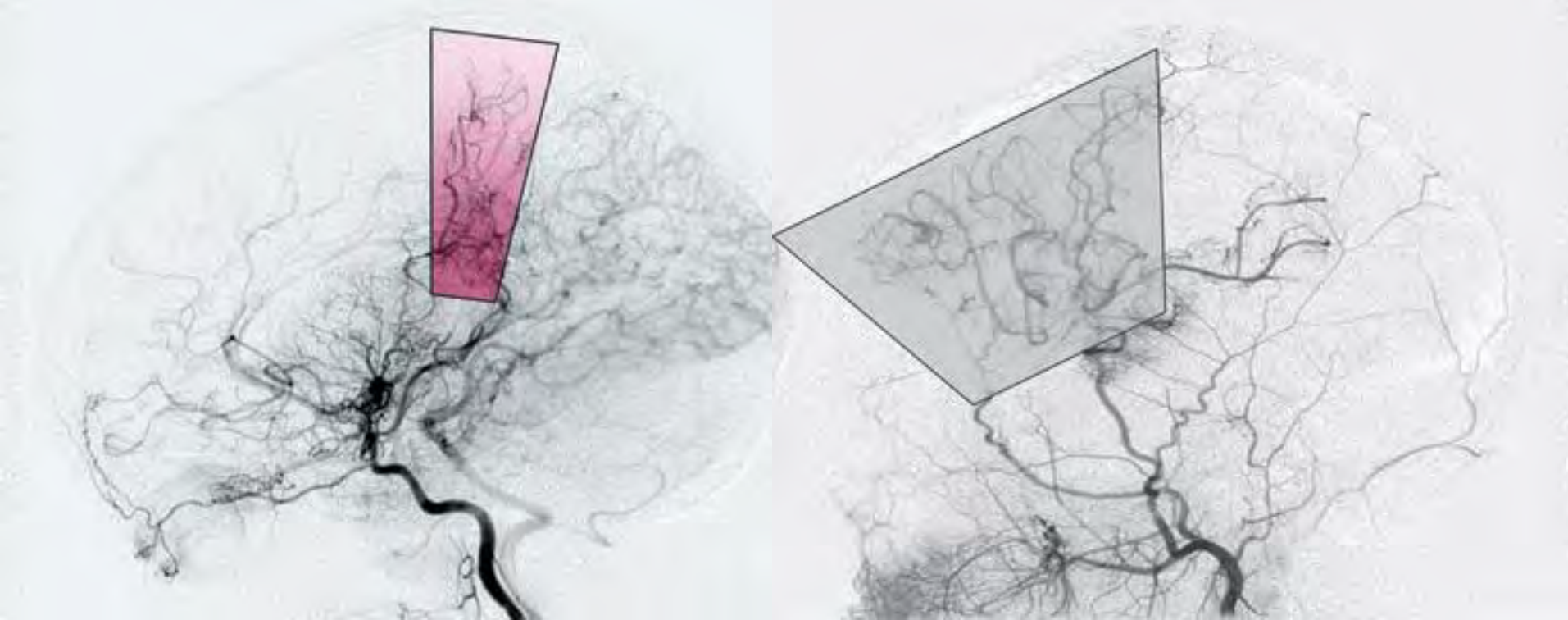


From Stanford moyamoya surgery website

• Indirect revascularization

- **EDAS** (encephaloduroarteriosynangiosis)
- the **EMS** (encephalomyosynangiosis)
- **multiple burr holes** procedure

One year post surgery

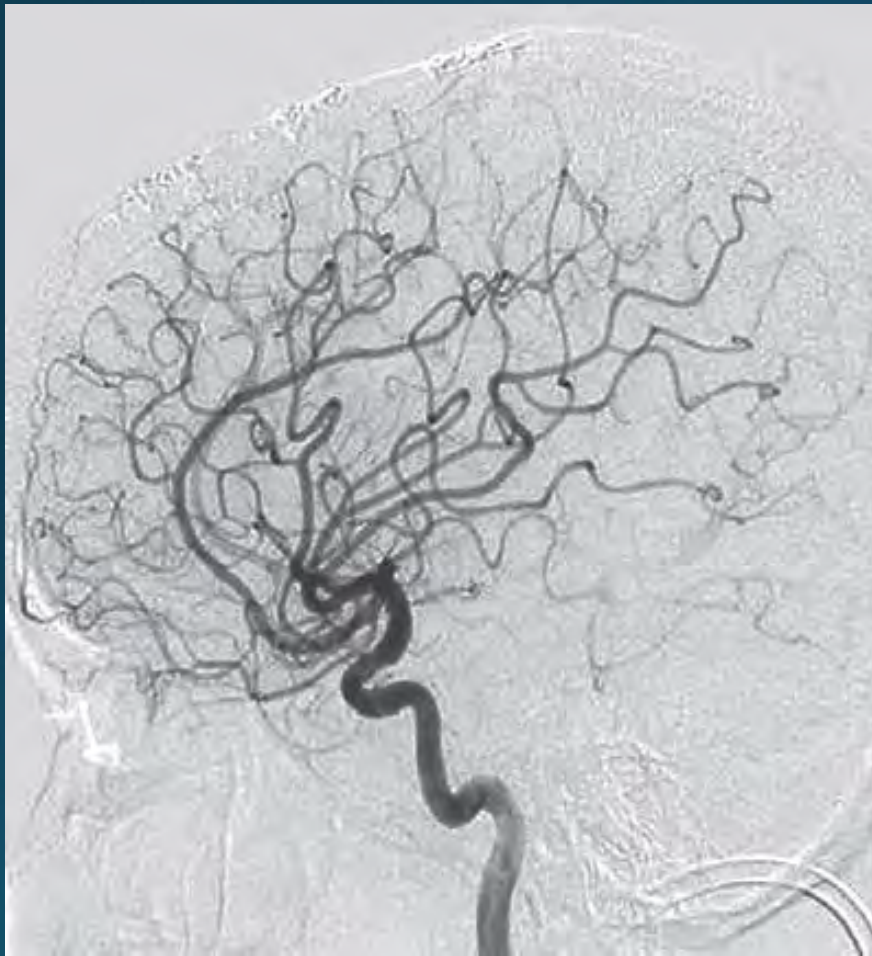


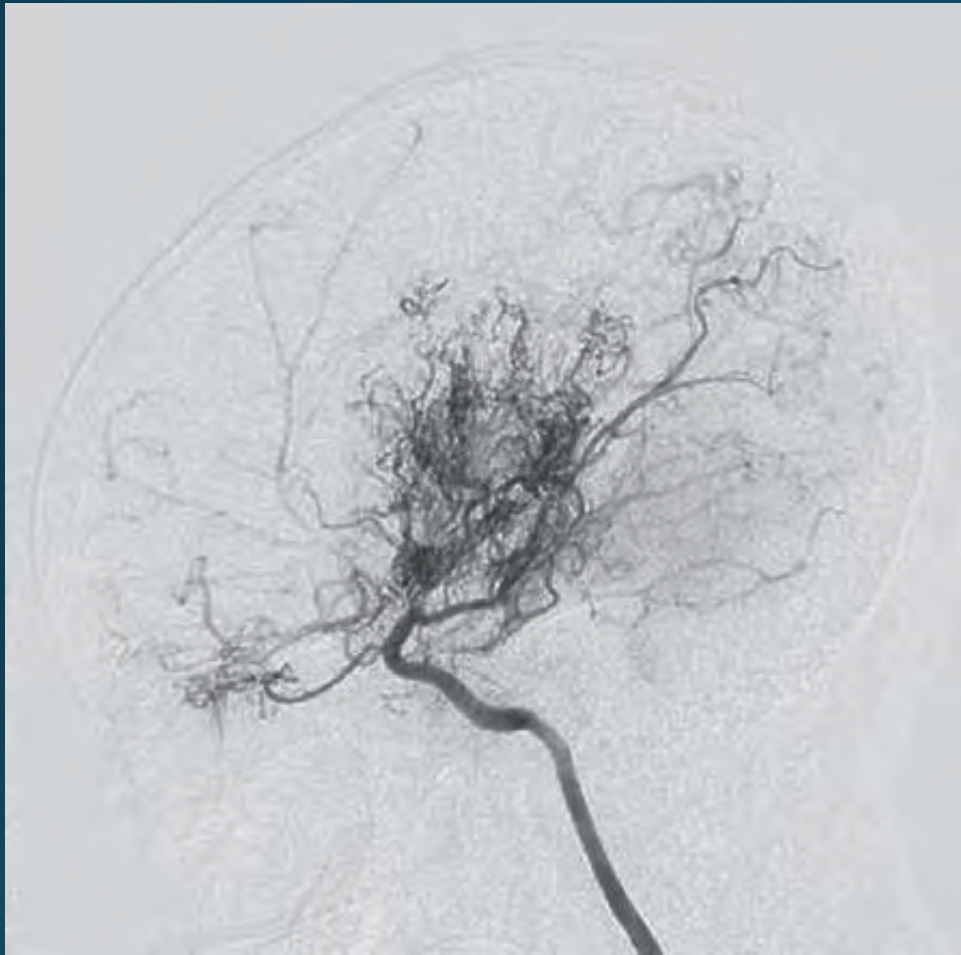
Moyamoya Disease and Moyamoya Syndrome
N Engl J Med 2009;360:1226-37

Diagnosis

- initially suggested by CT, MRI, or angiogram
- MRI and MRA should be performed for the diagnosis and follow-up of moyamoya disease
- Diffusion-weighted imaging useful to follow up
- nuclear medicine studies such as SPECT
 - to demonstrate the decreased blood and oxygen supply to
 - areas of the brain involved with moyamoya disease
- Conventional angiography is the gold standard

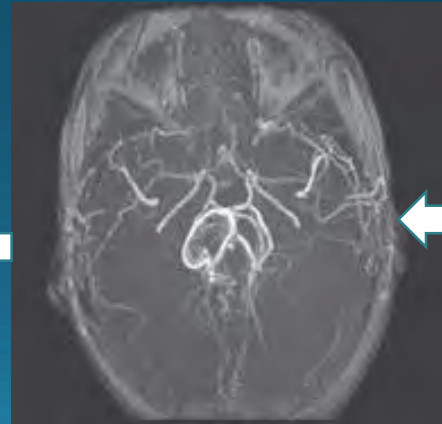
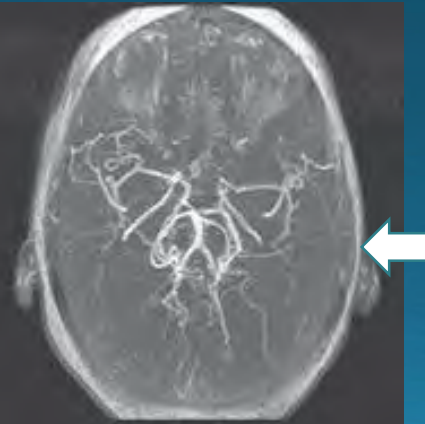
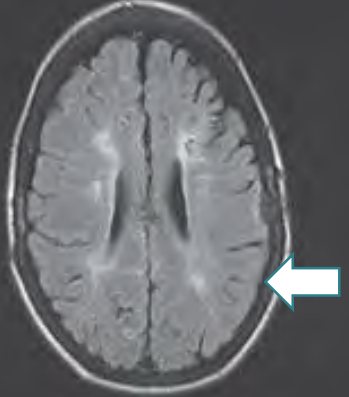
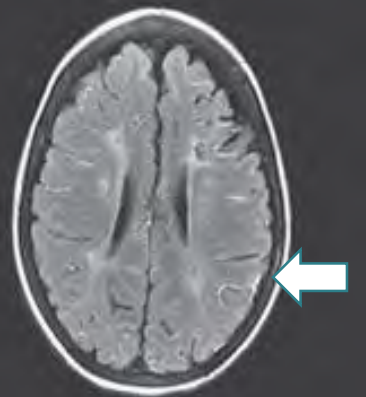
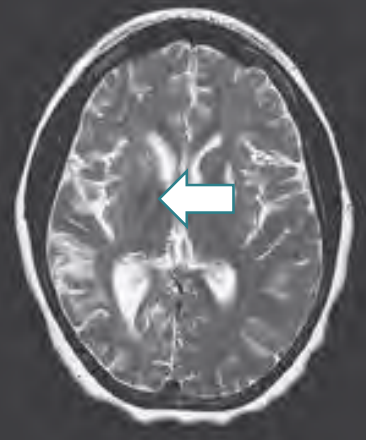
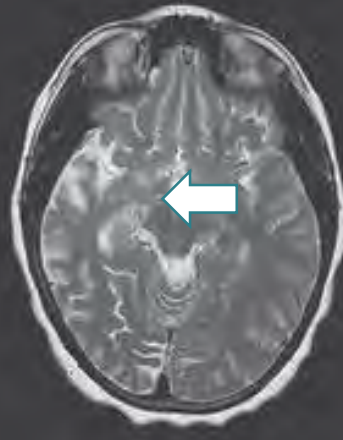
Cerebral Angiogram





Moyamoya Disease and Moyamoya Syndrome

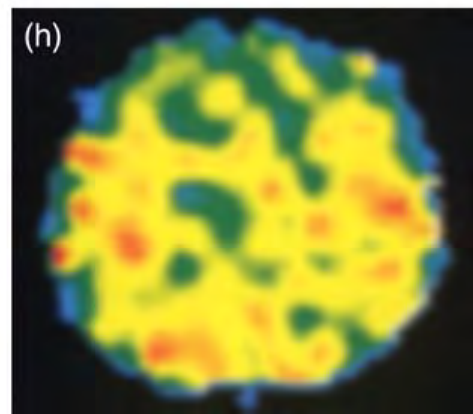
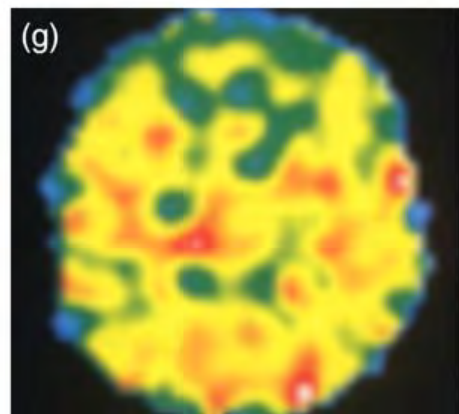
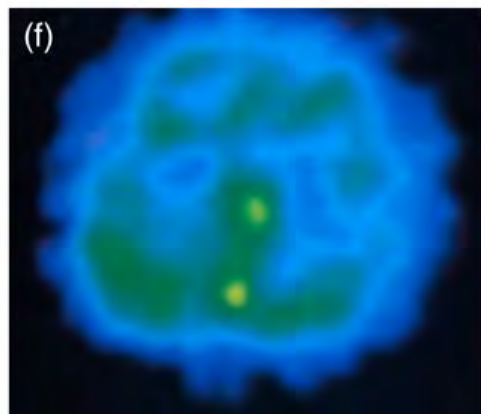
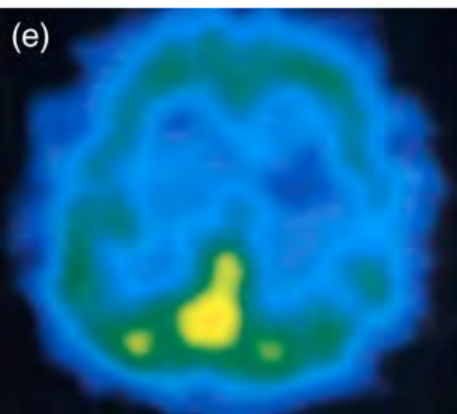
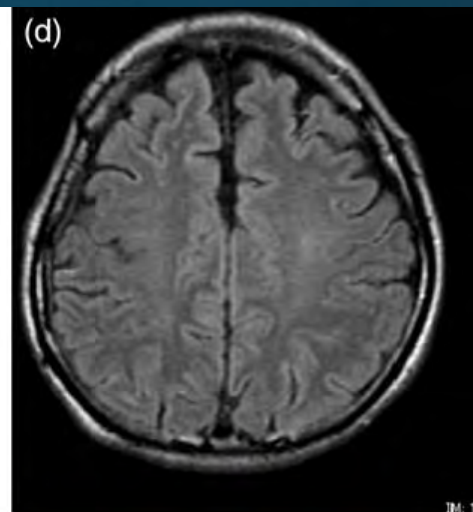
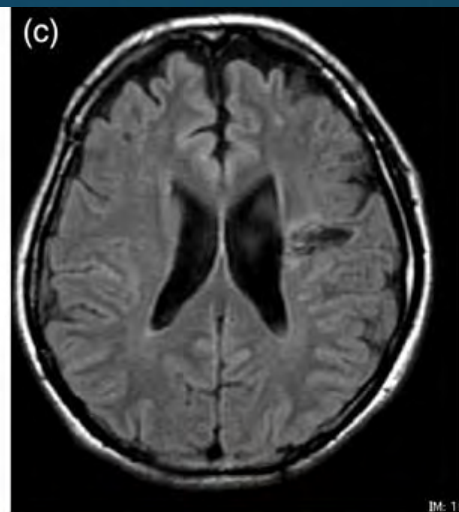
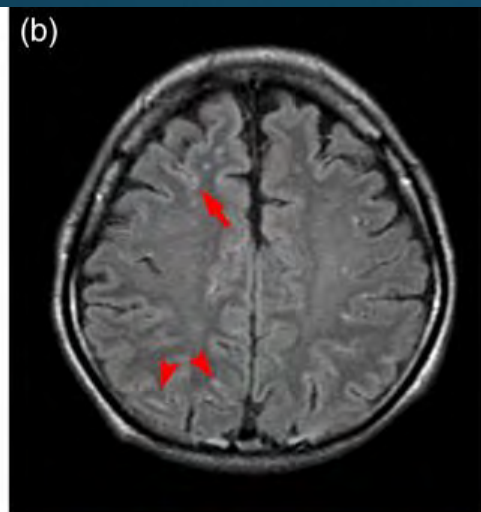
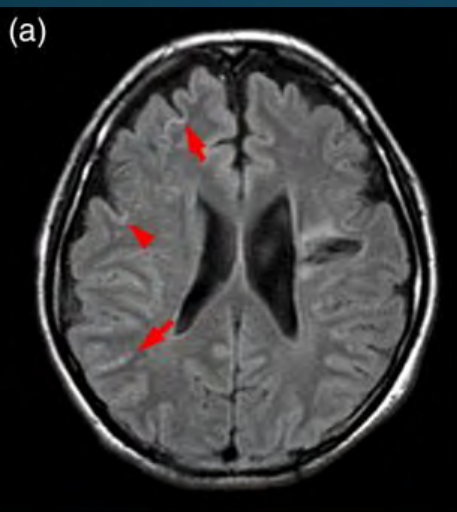
N Engl J Med 2009;360:1226-37



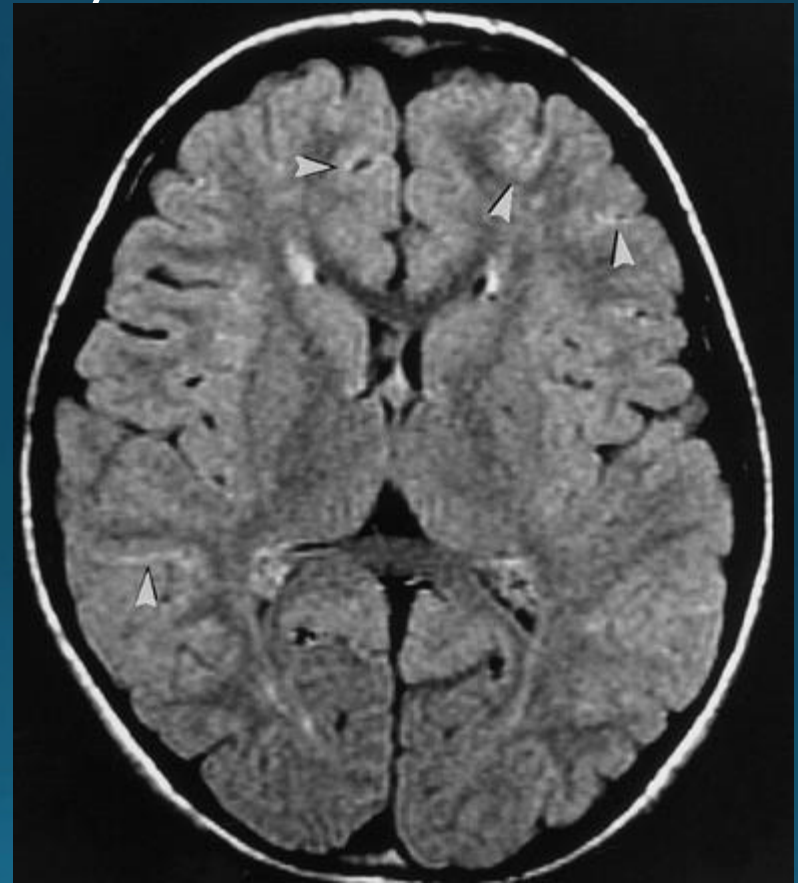
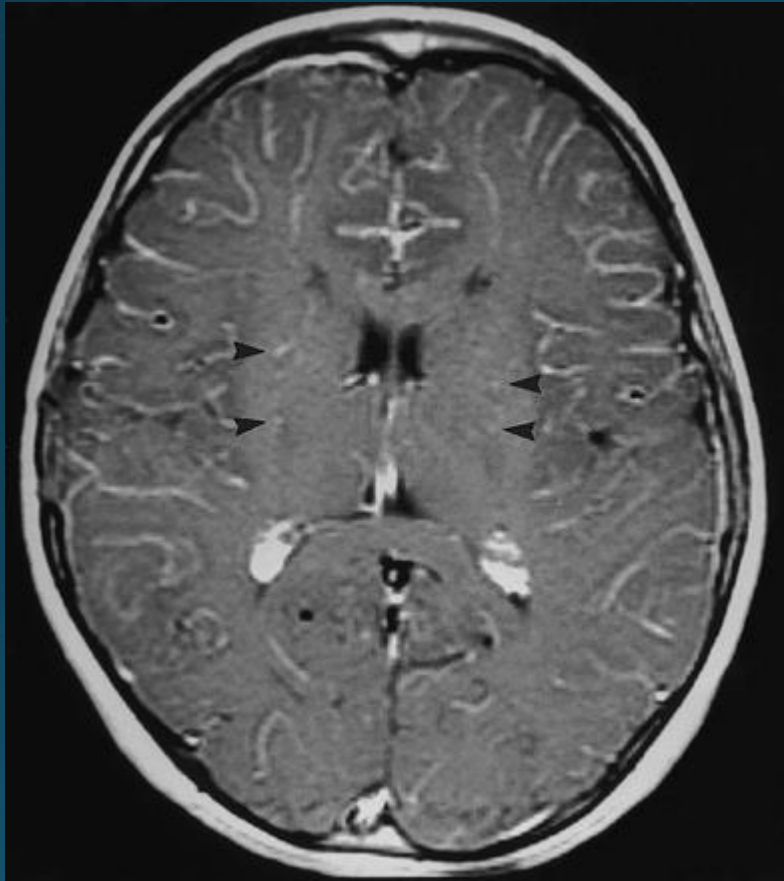
'ivy sign'

- diffuse leptomeningeal enhancement on post-contrast MR or FLAIR images
- the pattern of contrast enhancement resembled ivy creeping across stones
- disappears or decreases after successful bypass surgery



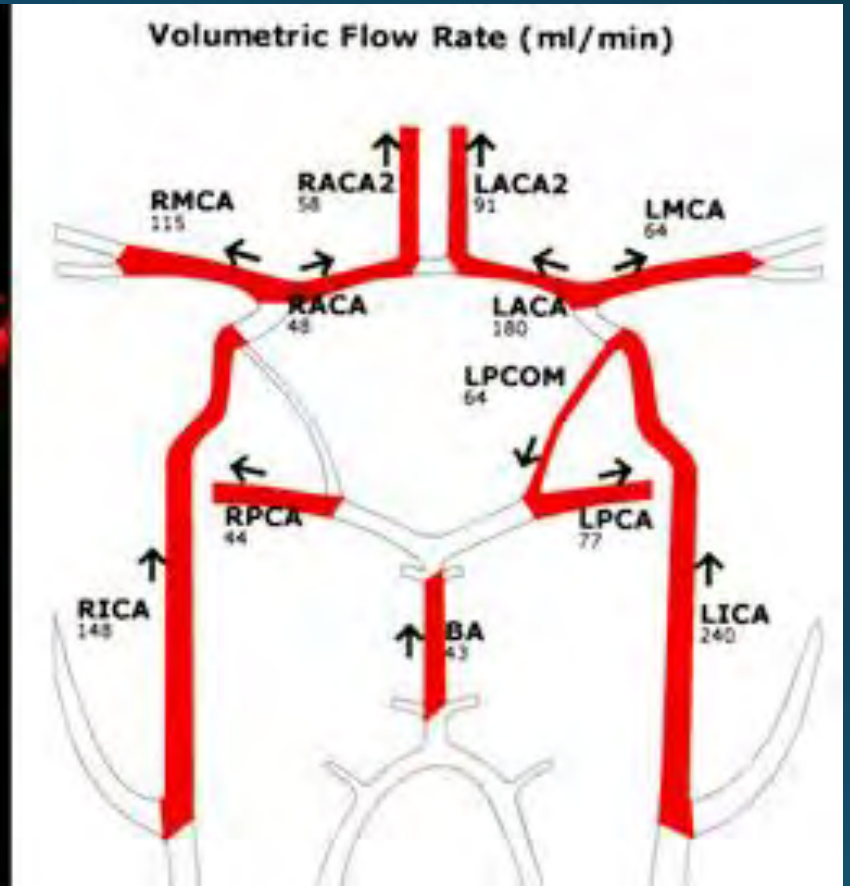
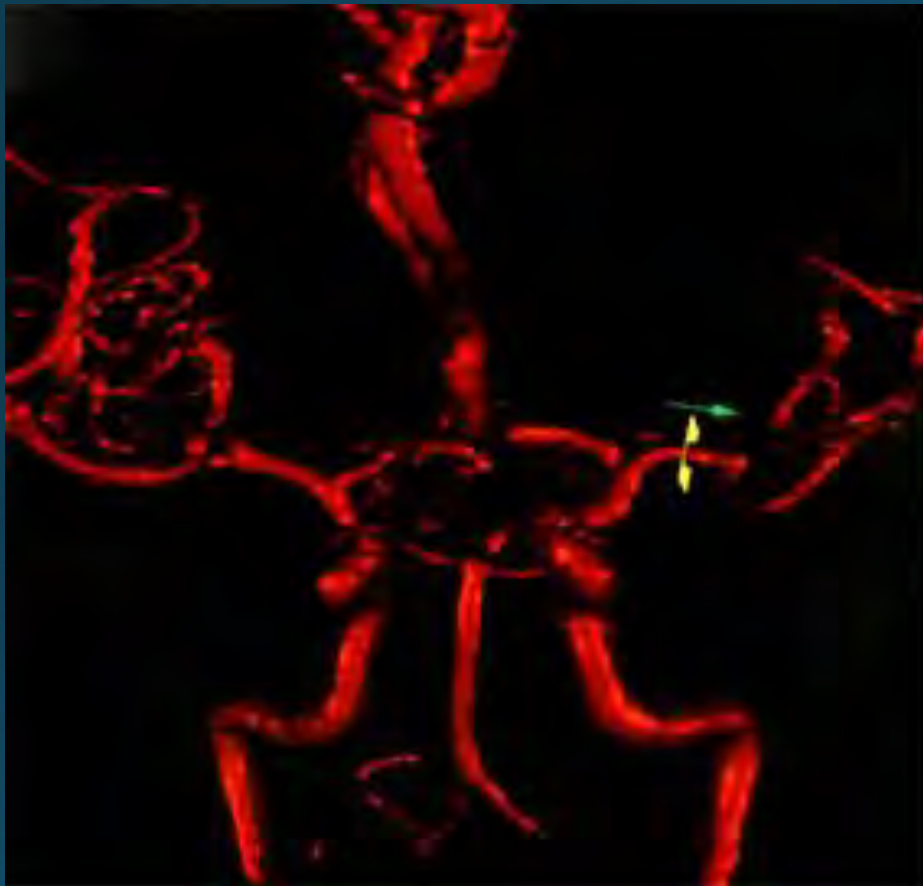


Gadolinium-enhanced T₁-weighted images are superior to FLAIR MR images for depicting the ivy sign in patients with moyamoya disease.



Ivy signs on FLAIR images before and after STA-MCA anastomosis in patients with Moyamoya disease. *Acta Radiol.* 2011 Apr 1;52(3):291-6

MRI/Nova



from Stanford moyamoya surgery website

Summary

- Moyamoya is Japanese = puff of smoke
- 6 grades by severity
- Direct or indirect bypass surgery
- Ivy sign on MR
- Post contrast T₁ is superior to FLAIR in detecting Ivy sign

References

- Moyamoya Disease and Moyamoya Syndrome
N Engl J Med 2009;360:1226-37.
- **Ivy signs on FLAIR images before and after STA-MCA anastomosis in patients with Moyamoya disease.** Acta Radiol. 2011 Apr 1;52(3):291-6
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Thanks!

