6 y/o boy presenting with arm numbness and mild weakness

Martin Ollenschleger, MD

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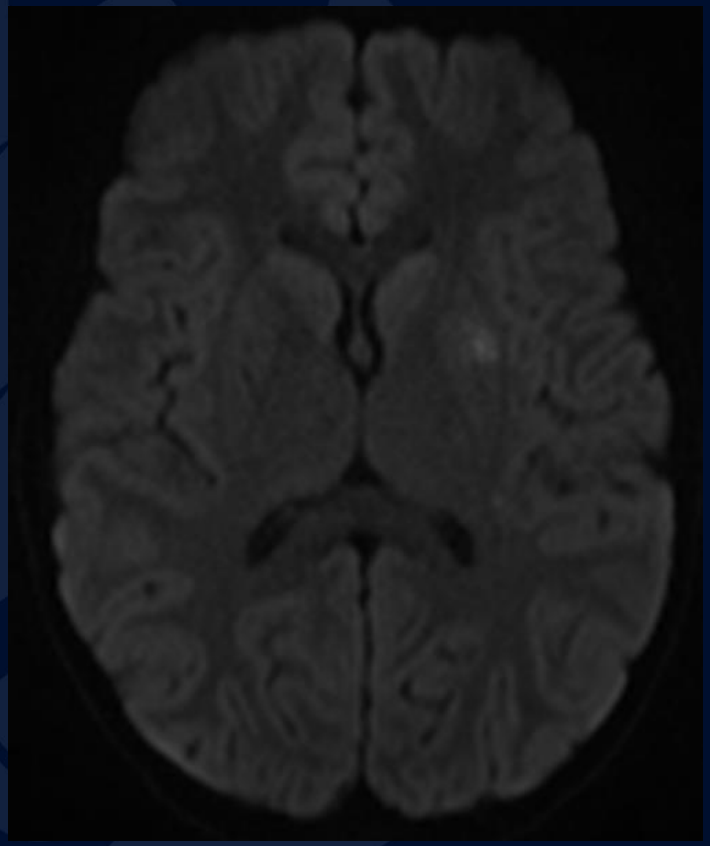

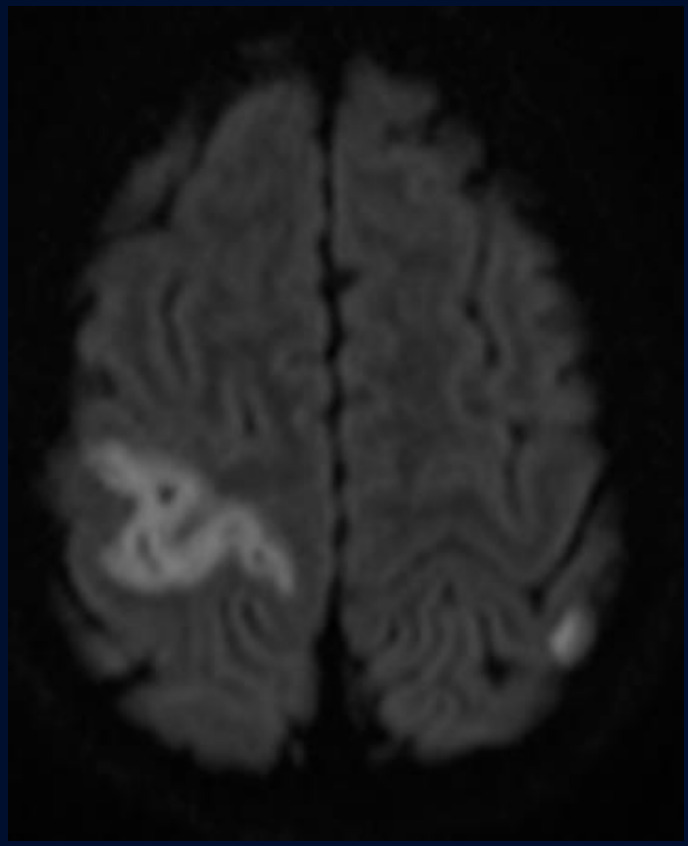

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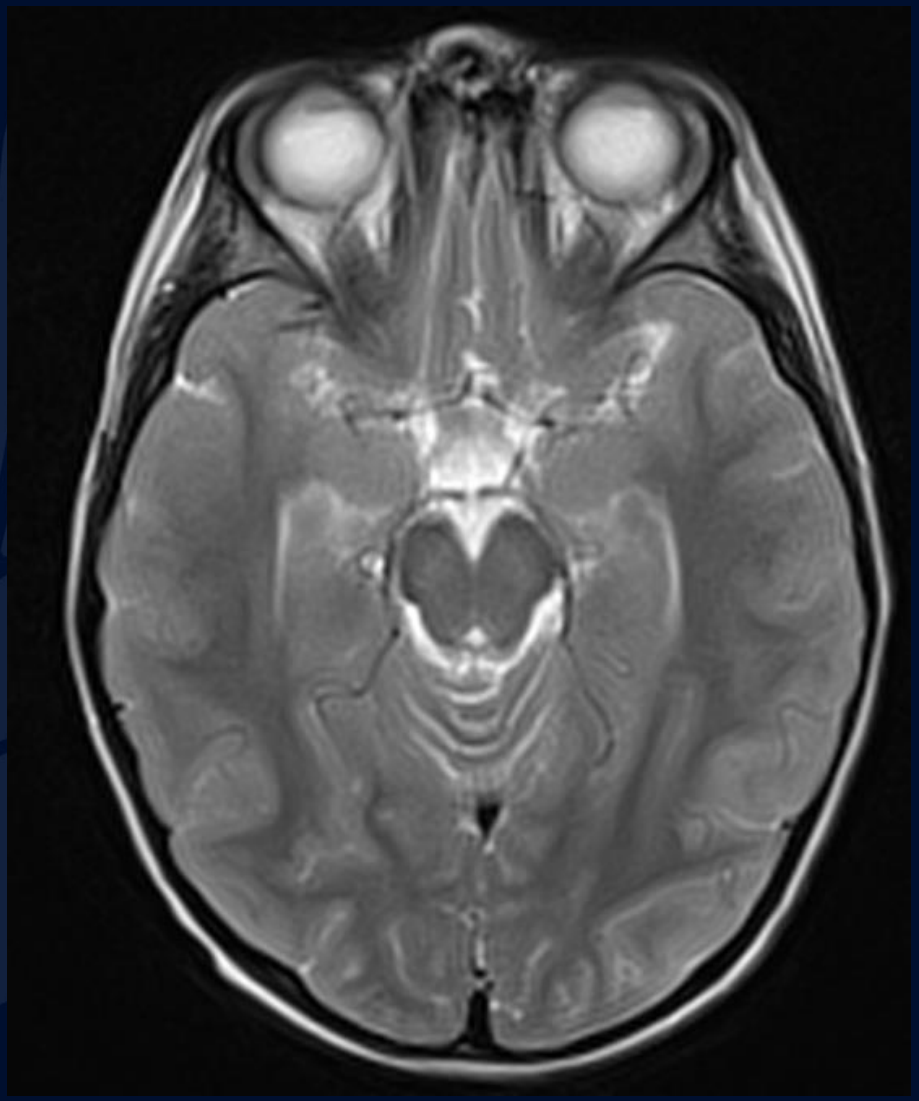

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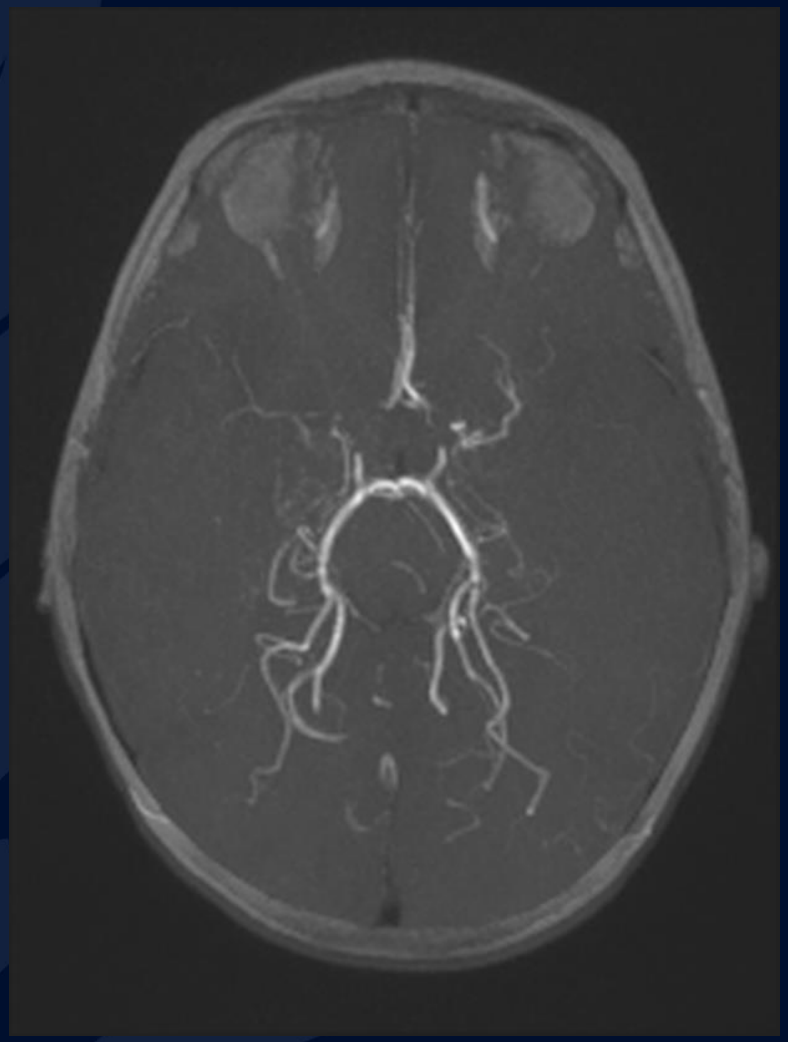

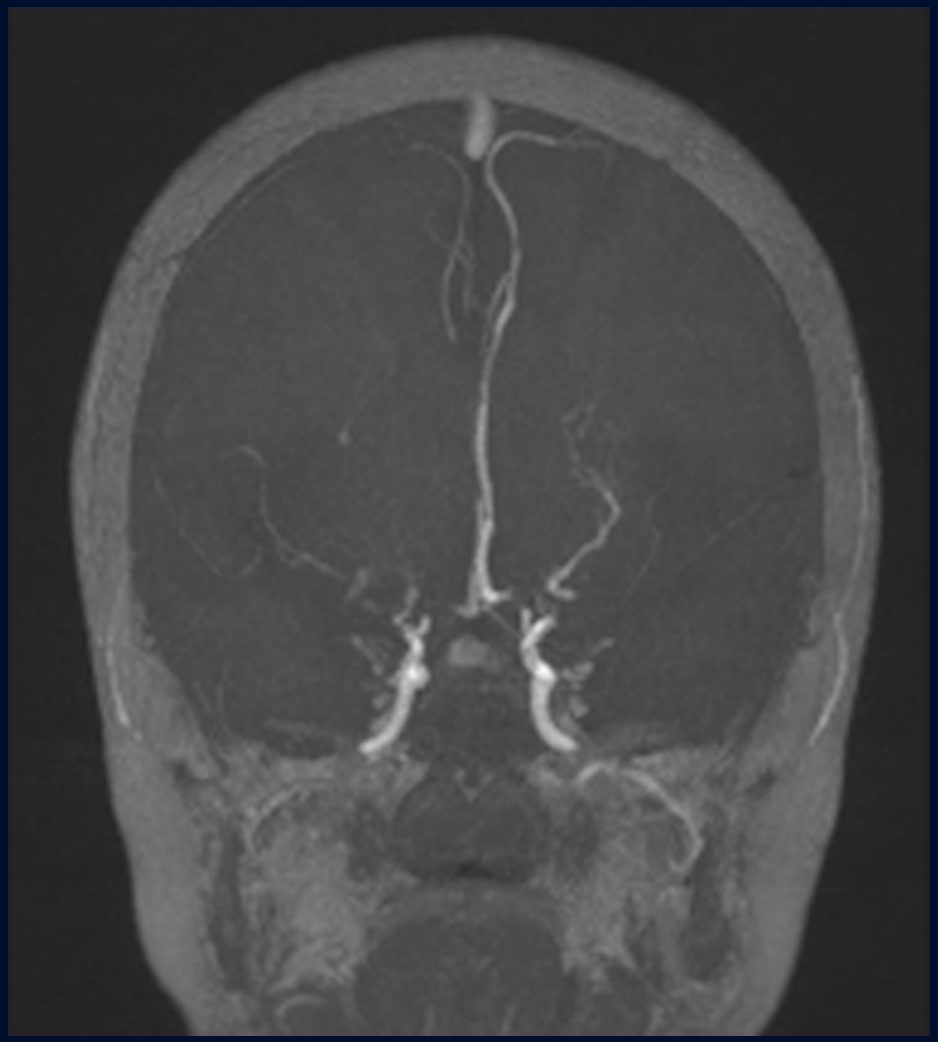

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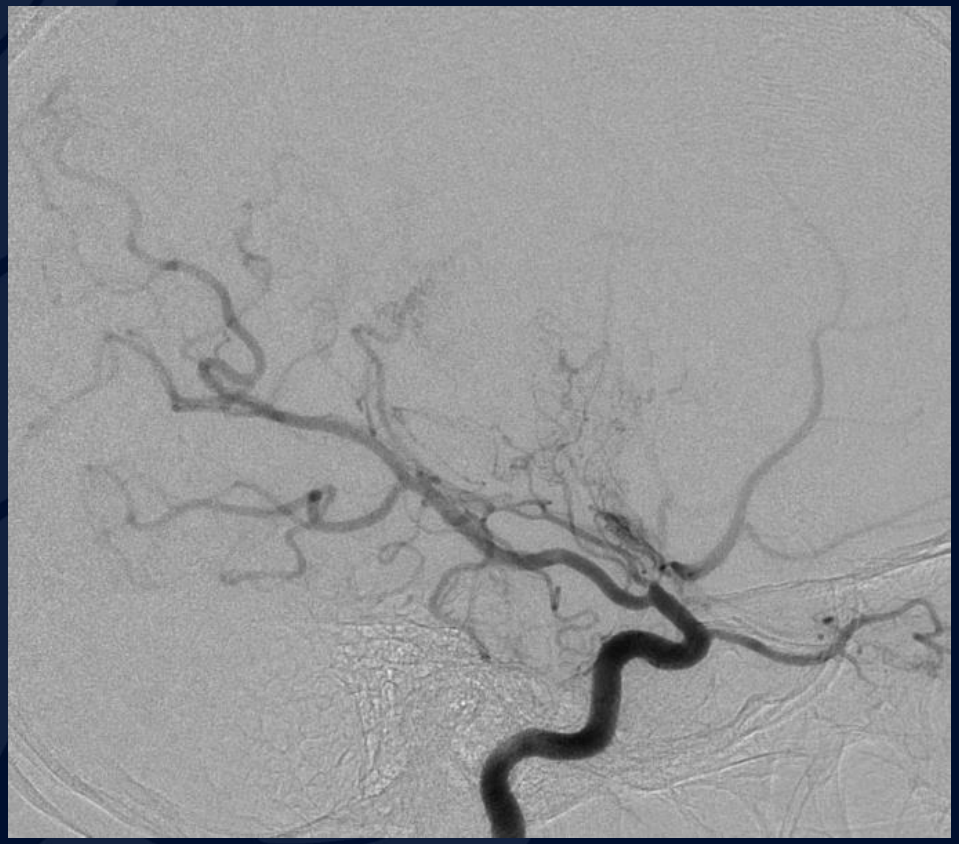

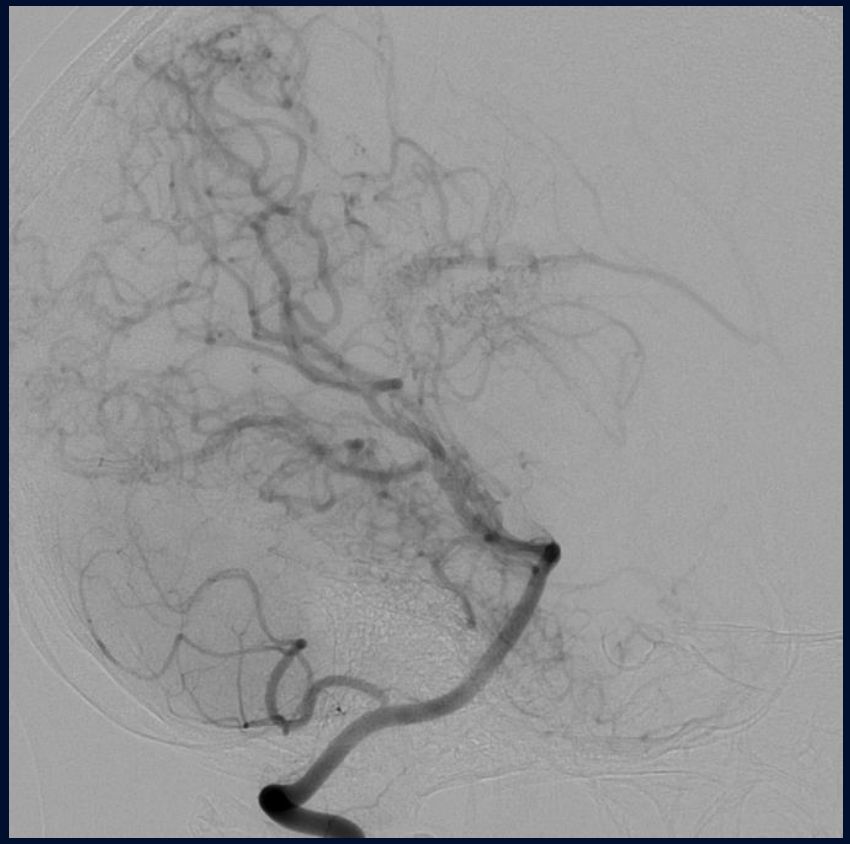

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Moyamoya

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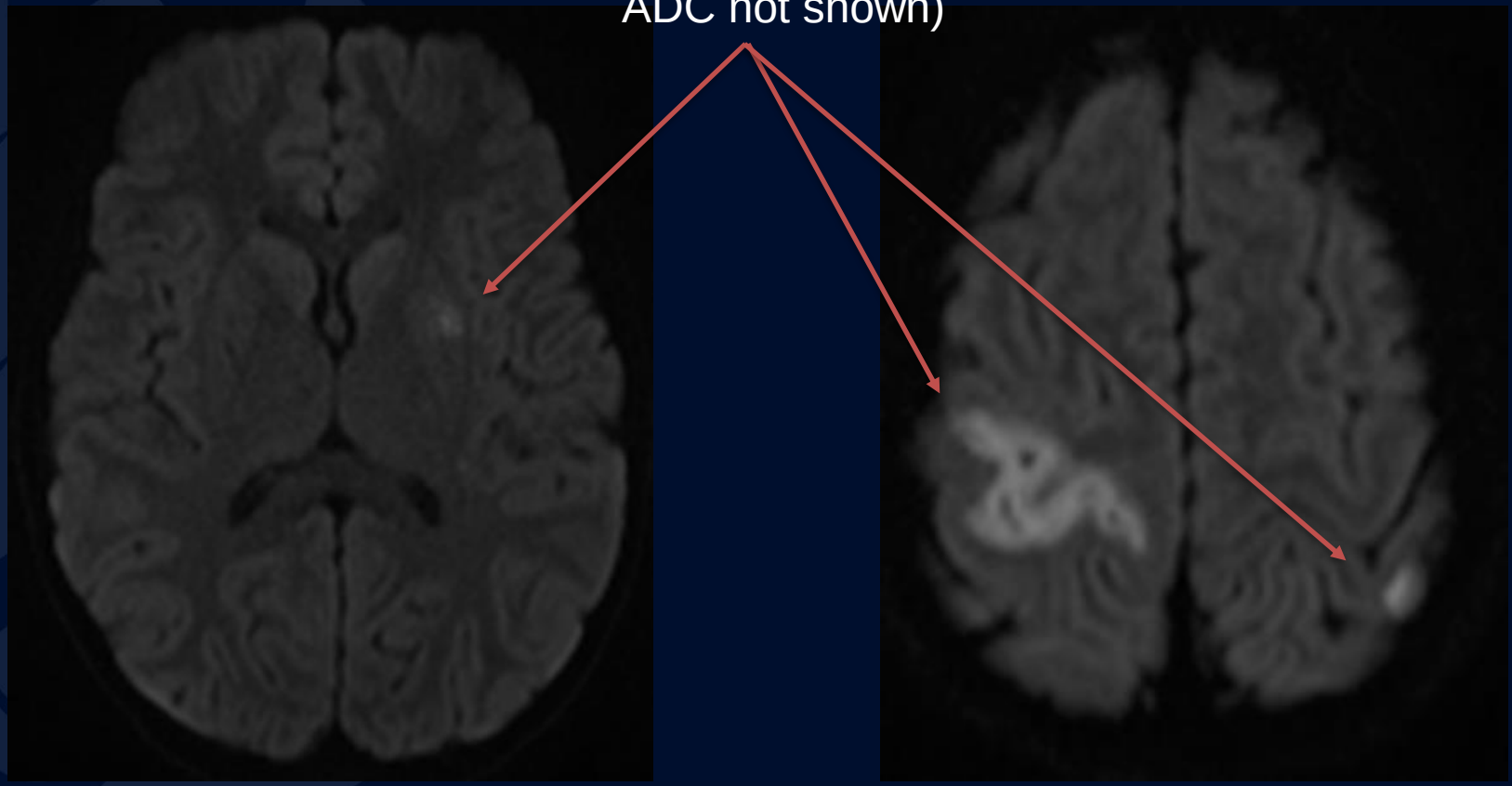
Areas of restricted diffusion consistent with acute infarction bilaterally (corresponding low signal ADC not shown)



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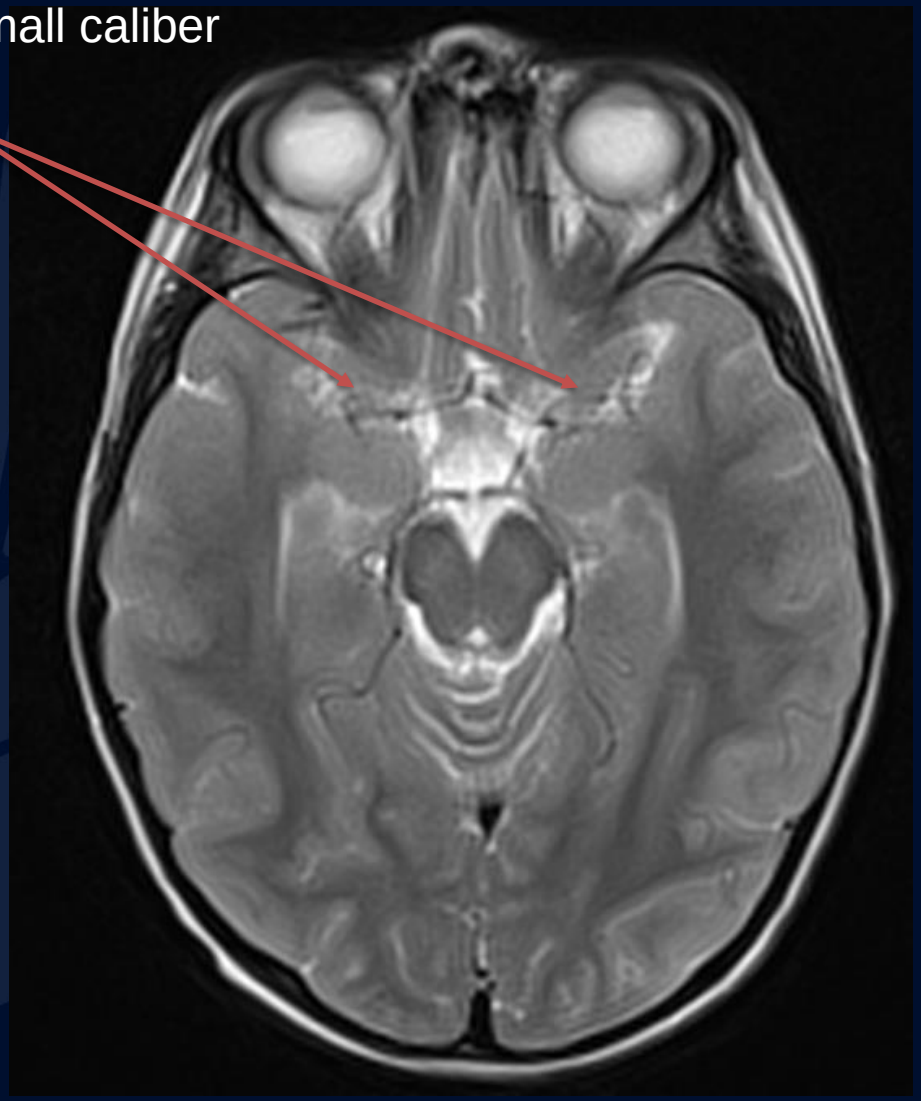
Abnormal MCA flow void with multiple small caliber vessels



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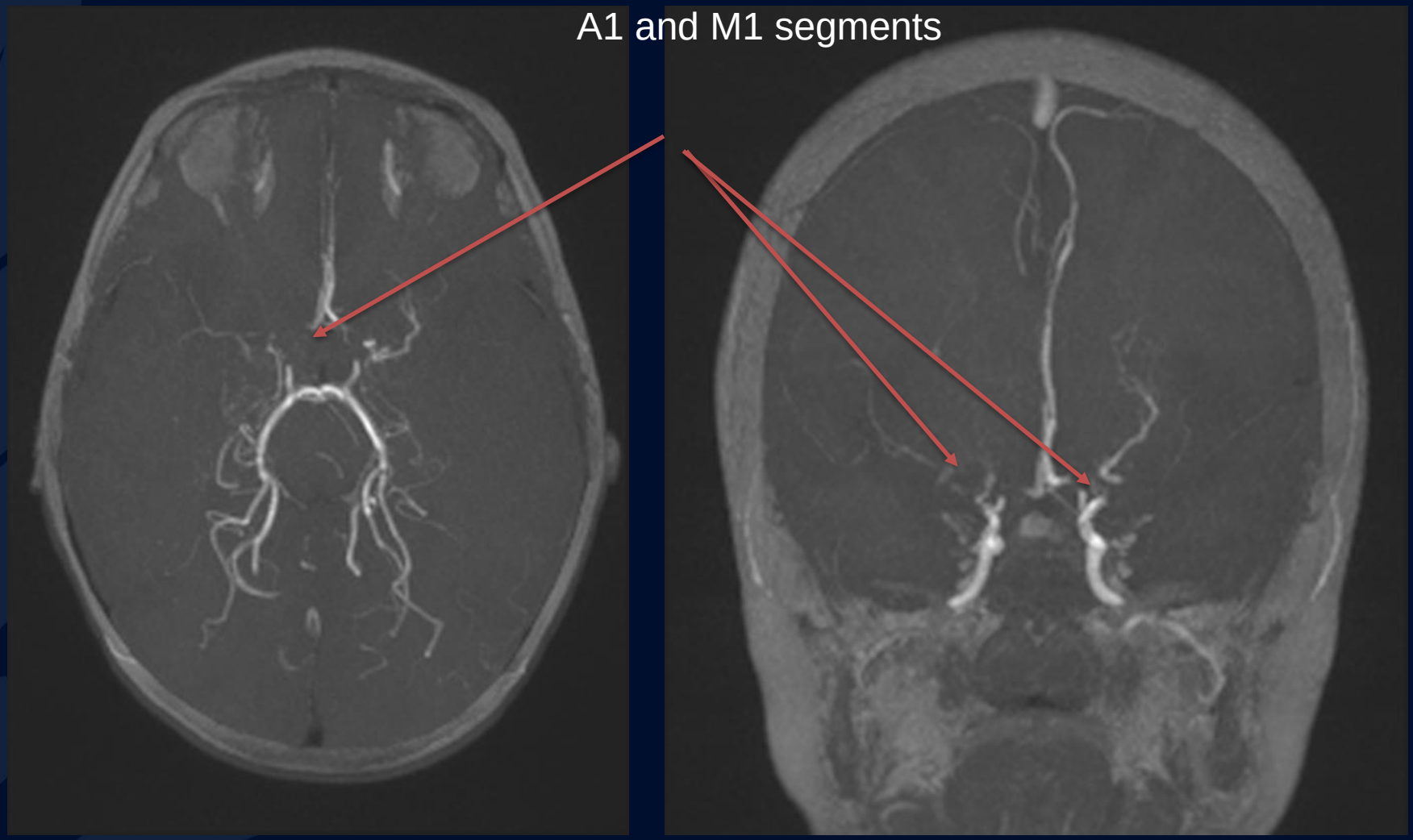
Severe, near occlusive stenosis of distal ICA and A1 and M1 segments



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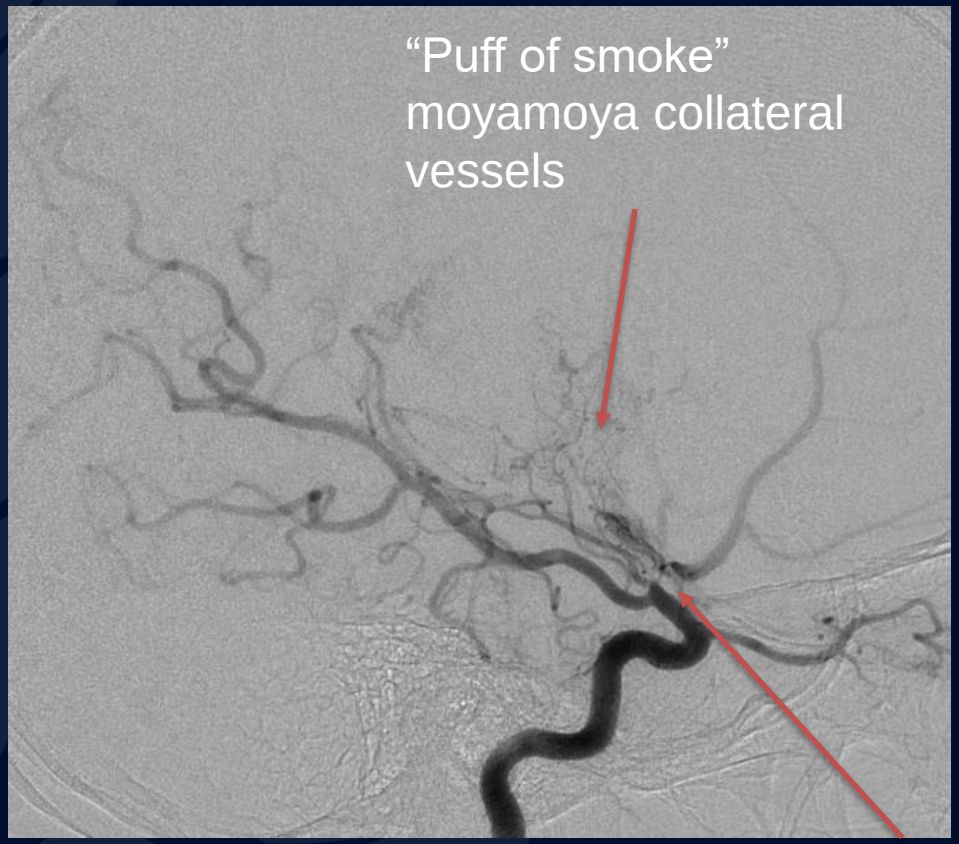
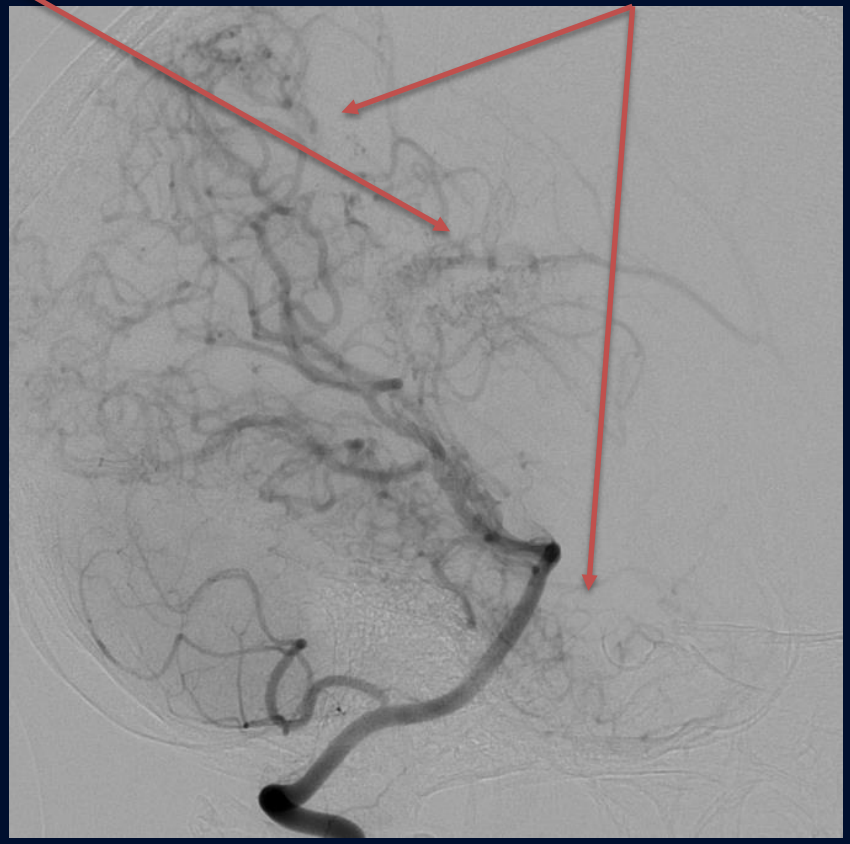
Peri-splenial branches back-filling pericallosal artery

Prominent leptomeningeal collateral vessels from PCA supplying parietal and temporal portion of MCA





Steno-occlusive disease of ICA terminal segment beyond anterior choroidal artery

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Moyamoya

- Moyamoya is a progressive steno-occlusive vasculopathy involving the internal carotid artery terminus and proximal anterior and middle cerebral arteries.
- The process is usually bilateral, though often appears asymmetric.
- Moyamoya disease is most common in Asian populations, though can be seen in other ethnicities.
- Moyamoya has a bimodal distribution with peak age at 5 years of age and at 40 years of age.
- Given the early age peak, moyamoya should be considered in the evaluation of pediatric stroke patients.

Moyamoya

Moyamoya syndrome manifests with moyamoya-like changes within the intracranial vessels, though is determined to be secondary to another underlying disease. Common diseases associated with moyamoya syndrome include:

- Sickle cell disease
- Down Syndrome
- Tuberous Sclerosis
- Neurofibromatosis Type 1
- Thyroid disease
- History of cranial irradiation

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Moyamoya (Imaging)

MRI:

- Evidence of acute, sub-acute or chronic infarction; usually small, subcortical infarcts bilaterally
- Abnormal fine caliber flow voids at the skull base and penetrating into basal ganglia
- Ivy Sign on FLAIR imaging – Linear high signal with cortical sulci
- Microbleeds on gradient echo imaging

Moyamoya (Imaging)

MRA:

- Stenosis or occlusion of the distal ICA extending into the proximal ACA & MCA
- Small caliber collateral vessels surrounding ICA terminus and MCA
- Rapid decrease in caliber of the post-bulbar cervical ICA (champagne bottle neck sign)

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Moyamoya (Imaging)

Cerebral Angiography:

- Ideal for visualizing the degree and extent of stenotic lesions of the distal ICA and proximal ACA/MCA and surrounding moyamoya collaterals (puff of smoke)
- Evaluate degree of collateral supply from posterior circulation leptomeningeal and ECA dural collaterals
- Evaluate caliber and location of major STA branches for bypass

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Moyamoya

- Progression of disease is classified into 6 stages by Suzuki, et al:

TABLE 1. Angiographic Suzuki Grading System of MMD

Stage	Manifestation
Stage I	Narrowing of the carotid Fork
Stage II	Initiation of the Moyamoya with dilatation of the intracerebral main arteries
Stage III	Intensification of the Moyamoya with partial disappearance of the intracerebral main arteries
Stage IV	Minimization of the Moyamoya., advanced steno-occlusive ICA, ACA and MCA with gradually enlarged collateral from the extracranial area
Stage V	Reduction of the Moyamoya, disappearance of ACA and MCA with increased collateral from the external carotid artery
Stage VI	Disappearance of the Moyamoya with cerebral circulation maintained only by the external carotid artery or the vertebral artery.

- Treatment
  - Symptomatic moyamoya is treated surgically to avoid progression of ischemia
  - Techniques
    - Indirect Bypass, most commonly Encephaloduroarteriosynangiosis (EDAS)
    - Direct STA-MCA bypass

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References

- Li, et al. *Imaging of Moyamoya Disease and Moyamoya Syndrome: Current Status*. J Comput Assist Tomogr 2019 (43): 257–263.
- Grotta, et al. Stroke: Diagnosis, Pathophysiology and Mangement. *Chapter 40: Moyamoya Disease*
- StatDX
- Citation: Ollenschleger M. Moyamoya. Radiology Online. 2021.

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