Does Arterial Spin Labeling Get Your Head Spinning?: A Radiologist's Guide to Cerebrovascular Reactivity MRI

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Teaching Points

1. Identify normal cerebrovascular anatomy.

2. Brief review of Moyamoya disease and Moyamoya syndrome, including common pathologies and options for neurosurgical treatment.

3. Fundamentals of ASL imaging as an MR perfusion technique.

4. Basics of a cerebrovascular reactivity challenge, including the pharmacological rationale of vasodilation and cerebrovascular reserve.

5. Common imaging presentations of chronic cerebral ischemia undergoing acetazolamide challenge CVR MRI.

Lexicon

ACZ	Acetazolamide
CBV	Cerebral blood volume
CBF	Cerebral blood flow
CPP	Cerebral perfusion pressure
CVR	Cerebrovascular reserve
MTT	Mean transit time
TMAX	Time to maximum
OEF	Oxygen extraction fraction
$CMRO_2$	Cerebral metabolic rate of oxygen
COW	Circle of Willis
SOD	Steno-occlusive disease
MMD/S	Moyamoya disease/syndrome
ASL	Arterial spin labeling
BOLD	Blood oxygen level dependent
DSC	Dynamic susceptibility contrast
PC	Phase contrast
DSA	Digital subtraction angiography
ECIC	Extracranial-intracranial

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Goal of Stroke Prevention

- Well-established correlate between compromised cerebrovascular reserve & an elevated risk of stroke
  - ✓ To estimate CVR, we can measure vascular flow response provoked by a vasoactive stimulus
- Abnormal CVR, defined as a poor CBF response or "steal", reveals the hemodynamic significance of a steno-occlusive lesion
  - x Blood vessels downstream of a blockage cannot dilate further in order to match tissue blood supply against increased demand from the "stressor"
- Although a variety of imaging modalities (CT/SPECT/PET/US) can achieve CVR mapping, MRI is a nonionizing method that also allows *in vivo* quantification

Fisher JA, Venkatraghavan L, Mikulis DJ. Magnetic Resonance Imaging-Based Cerebrovascular Reactivity and Hemodynamic Reserve. Stroke. 2018 Aug;49(8):2011-2018.

Pathophysiology of Cerebrovascular Disease

Ability to maintain normal CBF by reducing vascular resistance is compromised

Chronic hypoperfusion = hemodynamic progression of ↓ CBF resulting in ↓ CPP, triggering compensatory mechanisms (ie, autoregulation)

① **Early stage (autoregulatory vasodilation)** = occurs at the level of distal arterioles to ↓ vascular resistance (↑ in CBV and MTT), with no significant ↓ in CBF or OEF

② **Mid stage (autoregulatory failure)** = hypoperfusion exceeds maximal vasodilation, leading to an initial ↓ in CBF and a compensatory ↑ in OEF to preserve $CRMO_2$ (also known as "misery perfusion")

③ **Late stage (autoregulatory failure)** = advanced hypoperfusion (dramatic ↓ in CBF and CPP) exceeds maximal O_2 extraction and neurons are unable to maintain normal metabolic function (↓ $CRMO_2$) → ***cerebral ischemia***

Vagal AS, Leach JL, Fernandez-Ulloa M, Zuccarello M. The acetazolamide challenge: techniques and applications in the evaluation of chronic cerebral ischemia. AJNR Am J Neuroradiol. 2009 May;30(5):876-84.

Assessment of Chronic Steno-occlusive Disease

SOD = Chronic arteriopathy characterized by high-grade stenosis or occlusion of either the cervical carotid arteries or intracranial COW

Three main approaches (with associated modalities) to evaluate:

CTP, MRP

CT, SPECT, PET, MRI, TCUS

PET

① Measure the CBV/CBF ratio (mathematically = to MTT)

② Measure the degree of cerebral flow reserve (CVR)

$$CVR\ (\%) = \frac{CBF\ (Post\ ACZ) - CBF\ (Pre\ ACZ)}{CBF\ (Pre\ ACZ)} \times 100$$

③ Measure OEF directly

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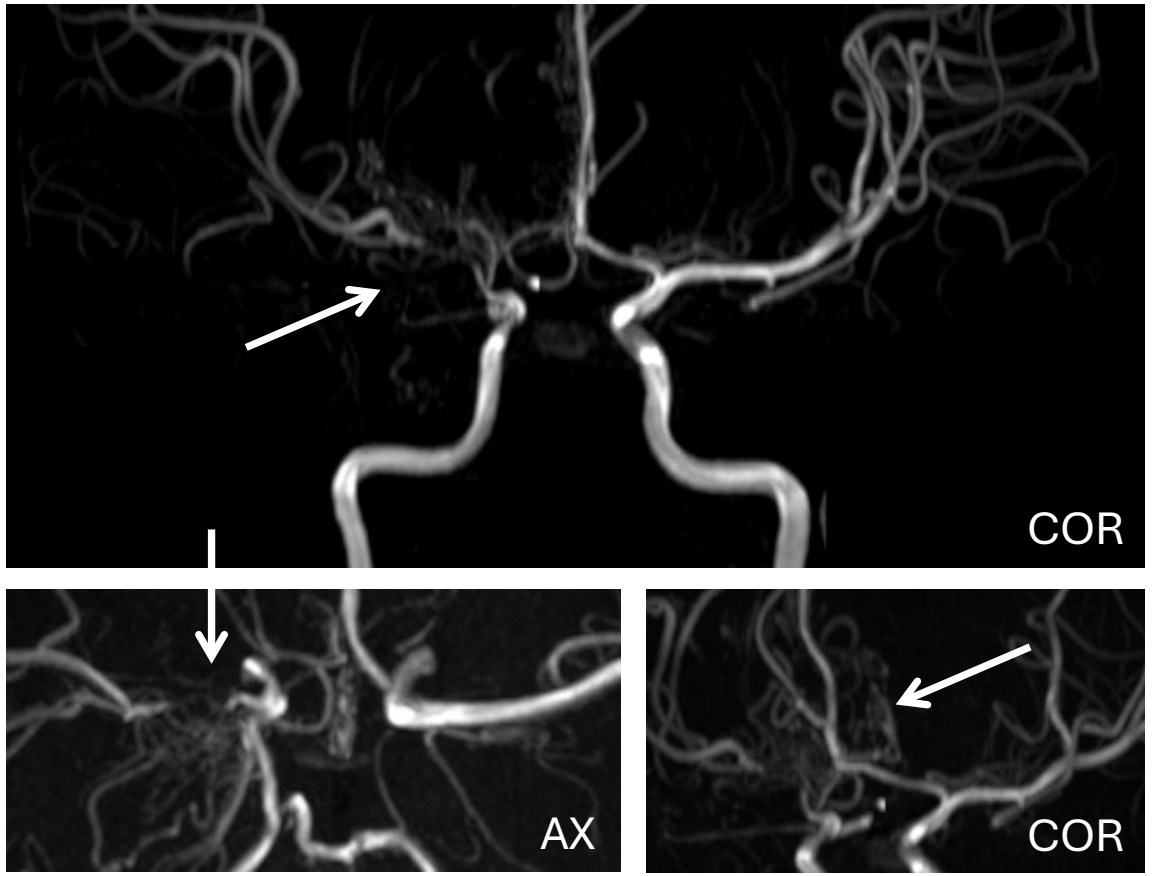
Moyamoya Angiopathy

- ✓Progressive stenosis or occlusion of supraclinoid ICA & major branches
- ✓Most commonly bilateral but can present with unilateral involvement
- ✓Affects both pediatric & adult populations
- ✓Frequently leads to cerebral atrophy & watershed infarctions (significantly ↓ CBF and ↑ CBV, MTT, OEF)
- ✓*Hallmark* = recruitment of collateral vessels to an area of chronic ischemia

Neovascularization



Chronic steno-occlusive disease (SOD) of the right anterior circulation. Moyamoya ("puff of smoke") appearance of the surrounding area, with collateralization of lenticulostriate vessels.

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Moyamoya Angiopathy

Etiologies:

- ✓ Blood dyscrasias (eg, SCD)
- ✓ Phakomatoses (NF1, TSC)
- ✓ Connective tissues disorders (SLE, Marfan, Ehlers-Danlos)
- ✓ Vasculopathies (eg, FMD)
- ✓ Vasculitides (eg, post-infectious, meningitis)
- ✓ Cranial irradiation
- ✓ Down syndrome
- ✓ Thyroid disease

➢ Disease vs. Syndrome? = Common endpoint of pathogenesis

- ✓ **MMD** = Idiopathic, sometimes familial, non-inflammatory & non-atherosclerotic
- ✓ **MMS** = Broadly-defined arteriopathy that mimics Moyamoya-like appearance ("phenomenon" or "pattern") with associated risk factors, such as atherosclerotic or inflammatory/vasculitis

➢ Treatment = S*urgical revascularization* due to ineffectiveness of medical therapy

- ✓ Predominantly ECIC bypass surgery, including direct & indirect revascularization:
  1. Direct superficial temporal artery (STA)-MCA bypass commonly performed in adults
  2. Indirect revascularization techniques (eg, EDAS) are preferred in pediatrics, as vessels are generally too small for direct anastomosis

➢ *Suzuki grading system* to characterize and monitor progression on imaging

- ✓ Catheter angiography remains the gold standard for diagnosis & postoperative surveillance

Li J, Jin M, Sun X, Li J, Liu Y, Xi Y, Wang Q, Zhao W, Huang Y. Imaging of Moyamoya Disease and Moyamoya Syndrome: Current Status. J Comput Assist Tomogr. 2019 Mar/Apr;43(2):257-263.

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Evolution of Moyamoya Angiopathy

Suzuki grading system by conventional DSA:

Stage	Manifestation
I	“Narrowing of the carotid fork” (ICA terminus / bifurcation)
II	“Initiation of the moyamoya” change (with dilation of the major MCA / ACA branches)
III	“Intensification of the moyamoya” change (with further narrowing of ICA & now MCA / ACA)
IV	“Minimization of the moyamoya” change (with occlusion of ICA & near-occlusion of MCA / ACA)
V	“Reduction of the moyamoya” change (with occlusion of ICA / MCA / ACA)
VI	“Disappearance of the moyamoya” change (with ICA elimination and cerebral supply from ECA)

**Moyamoya* = Japanese for “puff of smoke” to describe collateralization

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Terms of Cerebrovascular Reactivity

Reactivity = the magnitude of CBF response before (ie, baseline) and after a vasodilatory stimulus

$$\textbf{CVR} = \frac{\text{CBF (Stimulus)} - \text{CBF (Baseline)}}{\text{CBF (Baseline)}}$$

Augmentation phenomenon = normal baseline CBF with expected ↑ in CBF after vasodilatory stimulus (ie, *normal reserve*)

Limited augmentation = areas of normal CBF at baseline with less than expected ↑ in CBF after vasodilatory stimulus (ie, *poor reserve*)

Steal phenomenon = areas of ↓ CBF at baseline with a paradoxical continued ↓ in CBF after vasodilatory stimulus (ie, *absent reserve*)

Resultant ↓ in blood flow from regions already maximally dilated

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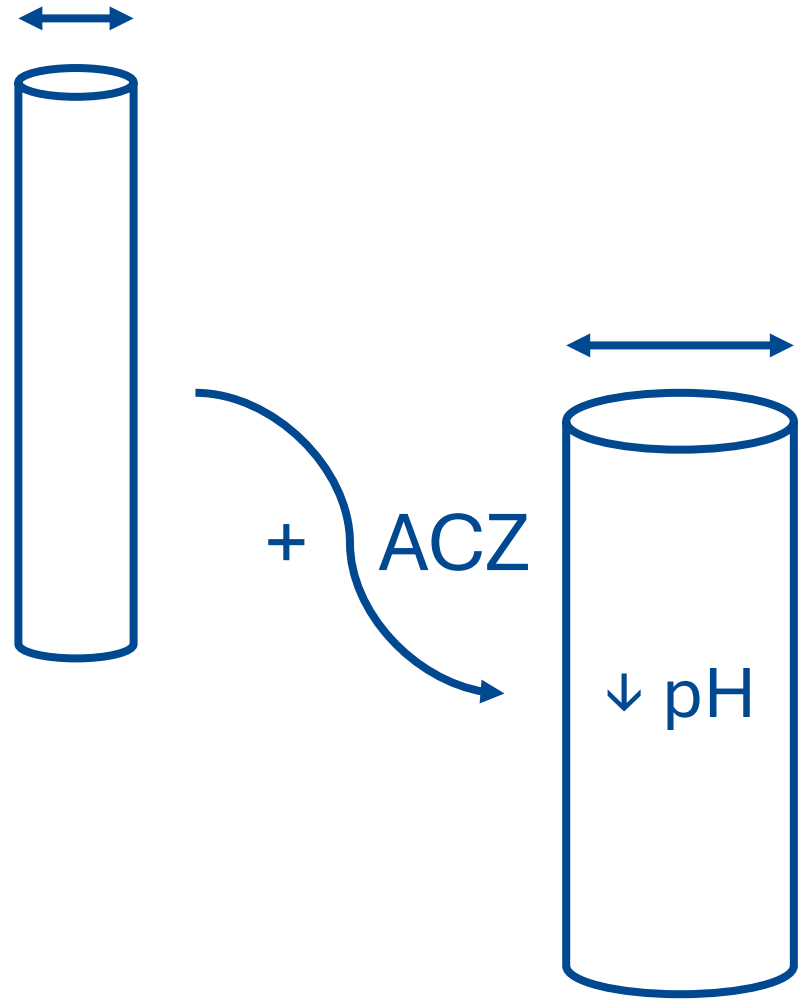
Acetazolamide Challenge

- Drug: Acts as a global vasodilatory agent
- Dose: Standard dose of 1000 mg (ie, 1 g) intravenously for the ACZ challenge test
- Blood-brain barrier: Able to cross BBB but permeates slowly
- Common side effects: Acute & transient, including transient circumoral numbness, paresthesias, malaise & headache
- Contraindications: ***Sulfa hypersensitivity**, electrolyte imbalances, hepatorenal disease, adrenocortical insufficiency, longstanding use for another condition (eg, glaucoma therapy)

  ***Note:** No longer an absolute contraindication but relative consideration, given new data regarding lack of cross-reactivity



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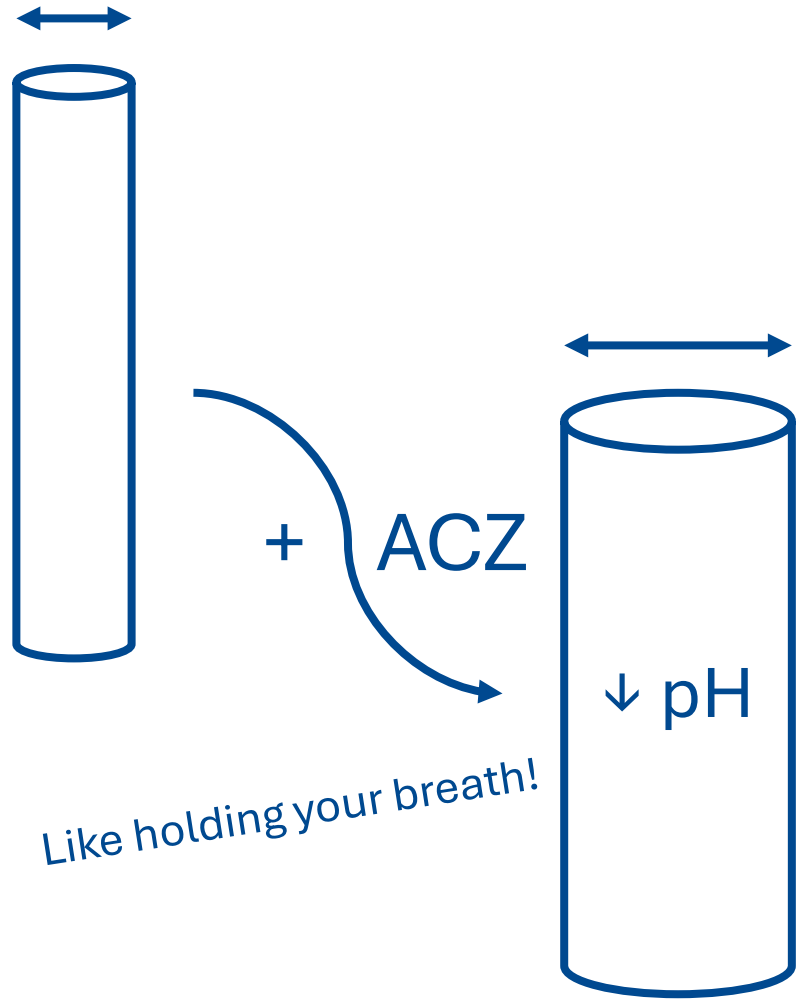
Acetazolamide Challenge

- Mechanism of action: Inhibitor of the carbonic anhydrase enzyme, important for acid-base homeostasis
  - → Inhibition leads to carbonic acidosis (↓ pH)
  - → Reactive vasodilation via smooth muscle relaxation occurs to increase blood flow (↑ CBF)
  - → Hyperemia "washes away" excess acid & restores normal pH
- Peak augmentation: 10-15 min after IV bolus
- Expected response: approx. 30-60% ↑ in CBF among healthy subjects (ie, *normal reserve*)
- Failed response: <10% ↑ in absolute CBF *or* an absolute change of <10 mL/100 g/min (ie, *abnormal reserve*)



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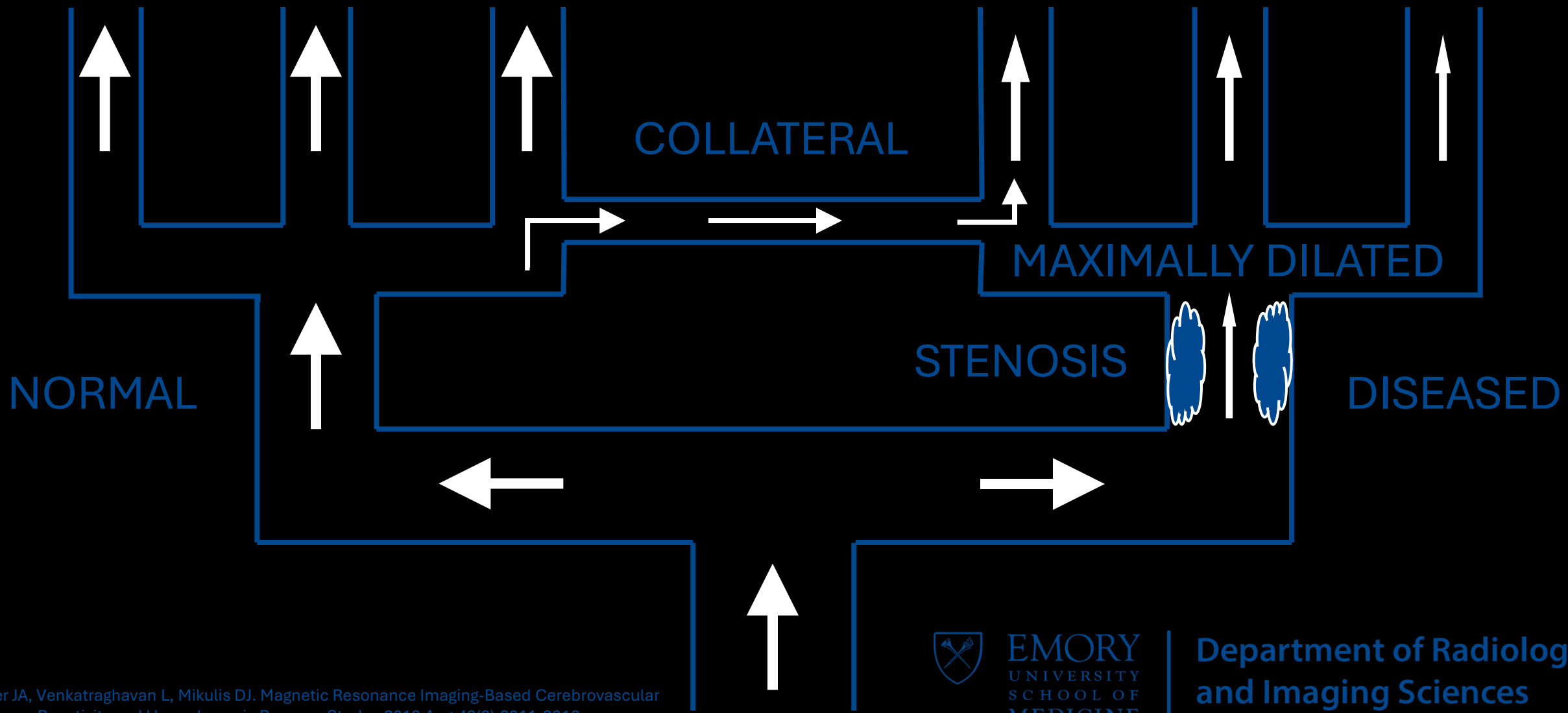
Steno-occlusive Disease at Baseline



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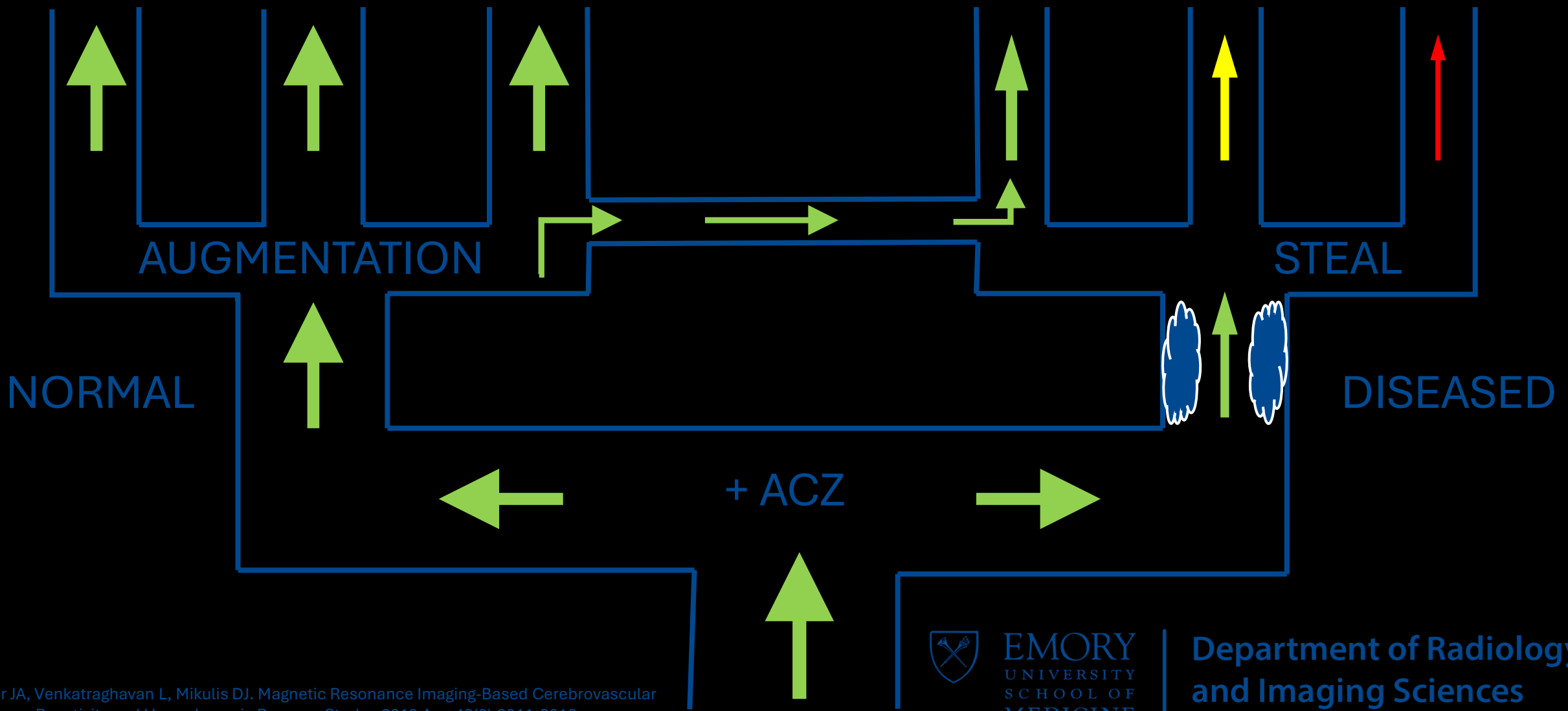
Steno-occlusive Disease with Challenge



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Arterial Spin Labeling

ASL = Noninvasive MR perfusion technique capable of quantifying CVR

- ✓Takes advantage of water in arterial blood as a freely diffusible tracer
  - →Spin labelling or "tagging" = *no exogenous contrast administration required*
- ✓Directly measures CBF in tissue (absolute physiological units of mL/100 g/min)
- ✓Comparable to gold-standard PET for mapping CBF in healthy subjects

Multiple MRI techniques are available for quantification:

- ✓Most common are BOLD, ASL and dual-echo ASL/BOLD
  - →BOLD = measures oxy- vs. deoxyhemoglobin (*well-correlated with CVR-ASL*)
  - →DSC = measures both CBF and CBV
  - →PC = measures CBF in large vessels

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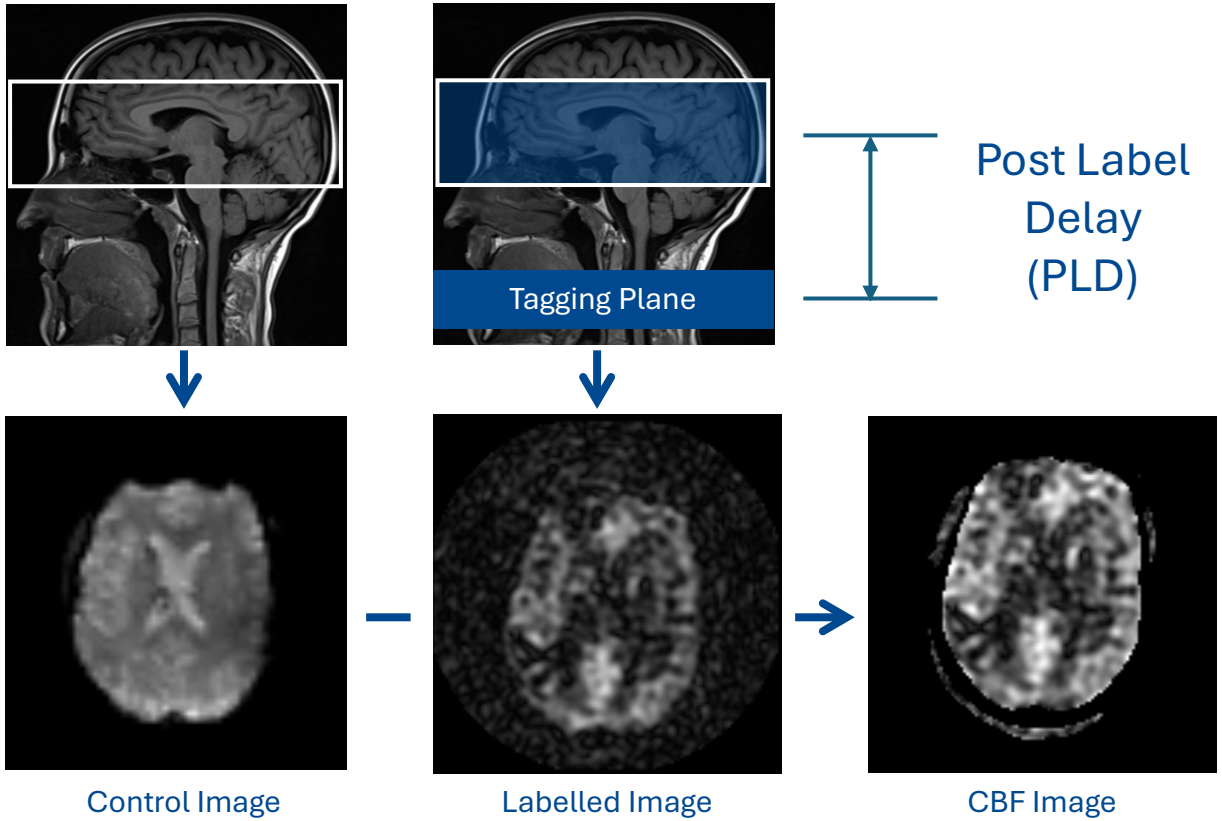
Arterial Spin Labeling



Different labeling methods:

1. Pulsed ASL (PASL)
2. Continuous ASL (CASL)
3. Pseudocontinuous ASL (pCASL)

Limitations that can potentially affect CVR estimation include:

- ✓Low signal-to-noise ratio (SNR)
- ✓Differences in labelling duration and efficiency
- ✓Bolus arrival time = distortion of flow measure due to changes in transit times

PLD too short → generates artifact
PLD too long → suffers from low SNR

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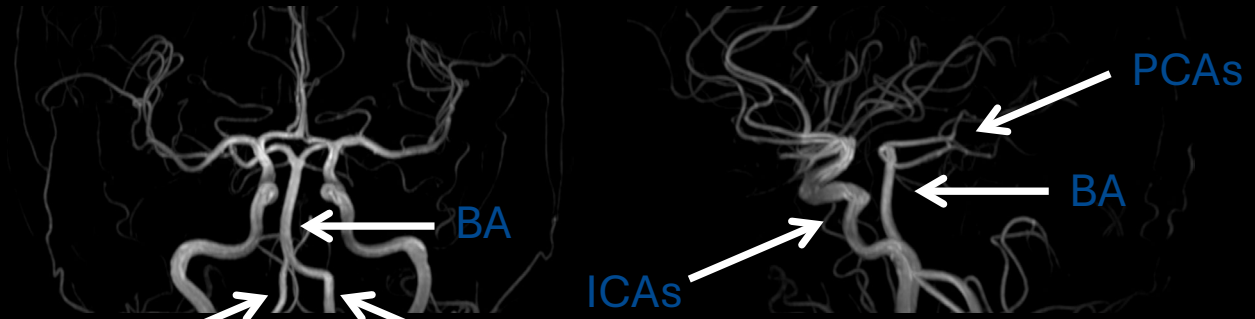
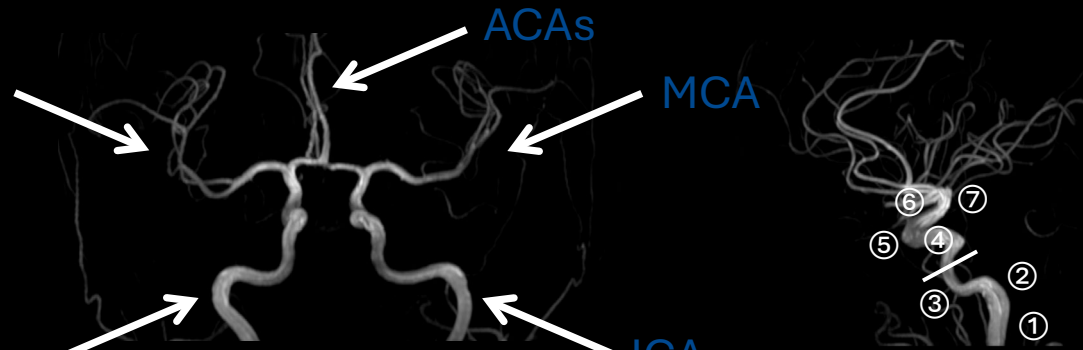
Cerebrovascular Anatomy

COW = Circle of Willis
ICA = Internal Carotid Artery
BA = Basilary Artery VA = Vertebral Artery
ACA / MCA / PCA = Anterior / Middle / Posterior Cerebral Artery

ANT+ POST CIRCULATION

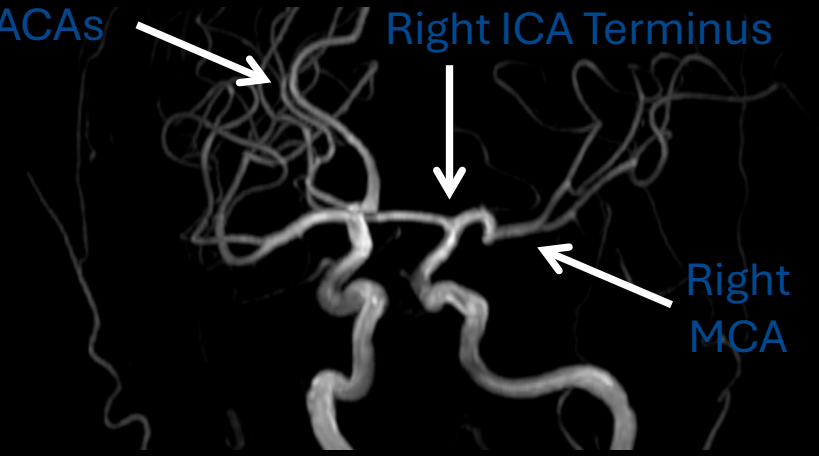


MRA COR MIP MRA SAG MIP



ICA Segments:
① Cervical
② Petrous
③ Laceral
④ Cavernous
⑤ Clinoid
⑥ Ophthalmic (Paraclinoid)
⑦ Communicating (Supraclinoid)

ANT CIRCULATION (POST-PROCESSING)



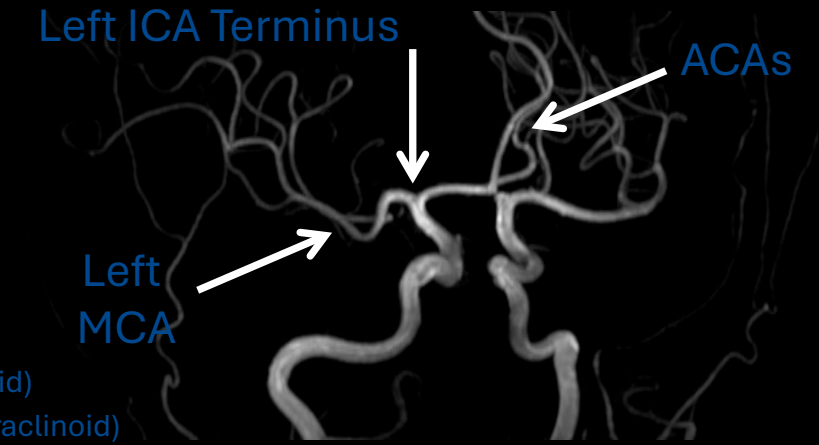


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Figure 1. *Right anterior circulation chronic steno-occlusive disease, compatible with moyamoya. Poor cerebrovascular reserve in the right MCA and ACA territories*.

(**T2**) Loss of right ICA flow void. (**Initial FLAIR/DWI**) Acute on chronic watershed infarcts in the right frontoparietal as well as temporo-occipital regions, indicating MCA/ACA and MCA/PCA border zones. (**CVR-DWI**) No new areas of infarction after acetazolamide challenge. (**3D-TOF MRA**) Absent flow-related signal in the distal right ICA. Reconstitution of flow-related signal in the ACA and MCA, likely due to collateral supply from the ACOM.

(**Dynamic BOLD imaging**) Impaired augmentation in the right MCA and ACA territories. Additionally, decreased signal in the left anterior MCA and ACA territories, which may be artifactual, given discordance with MRA and ASL findings. However, this could also represent steal phenomenon from collateral shunting across the ACOM. (**Pre- and post-acetazolamide ASL perfusion**) Impaired augmentation in the right MCA and anterior ACA territories. (**Post-acetazolamide DSC perfusion**) Severely prolonged Tmax in the right MCA territory and MCA-ACA border zone, with corresponding reduction in CBF.

Case 1

48-year-old male with left facial numbness and left arm weakness

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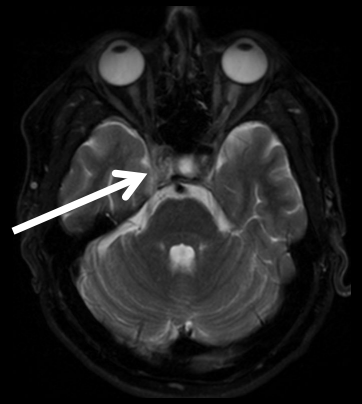
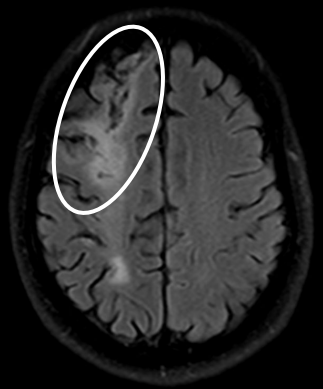
Case 1

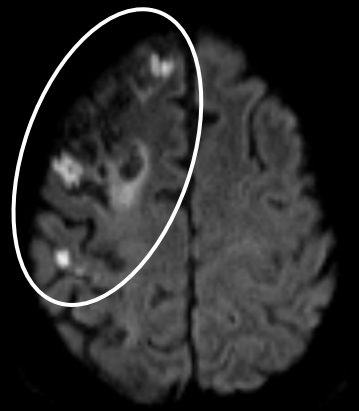
CVR DWI performed 3 days later



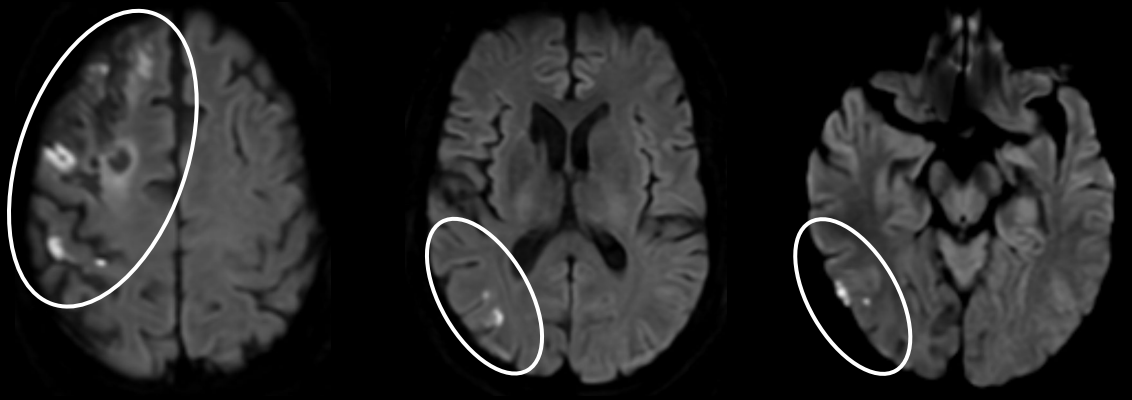
Initial T2



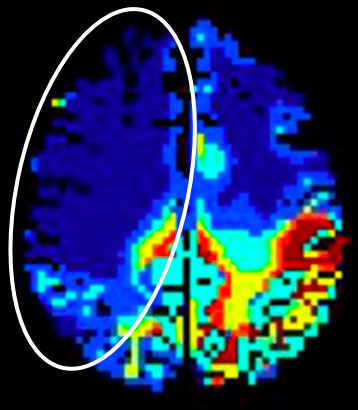
Initial FLAIR



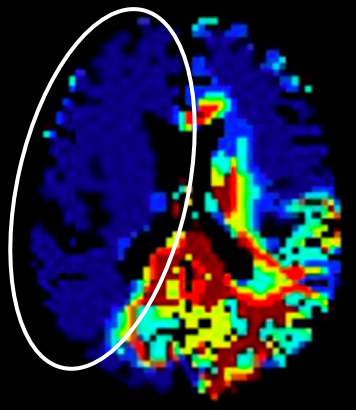
Initial DWI



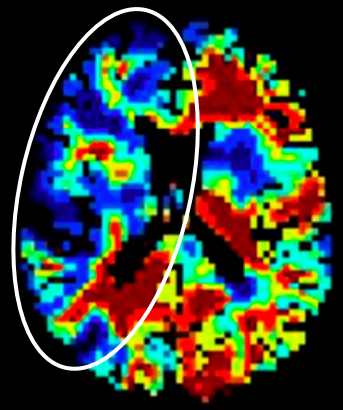
CVR BOLD

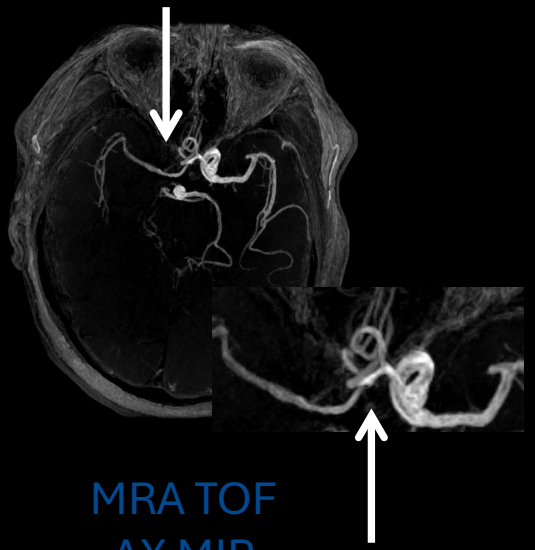


CVR BOLD

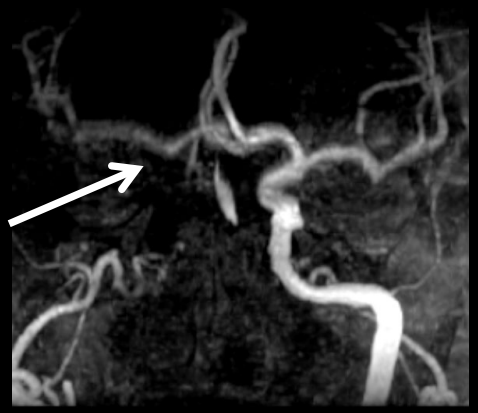


CVR ASL





MRA TOF
AX MIP



MRA TOF
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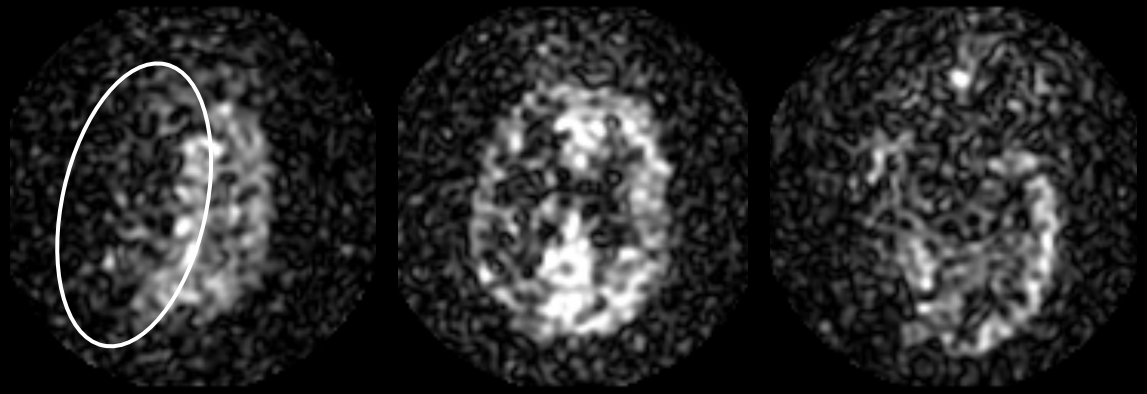
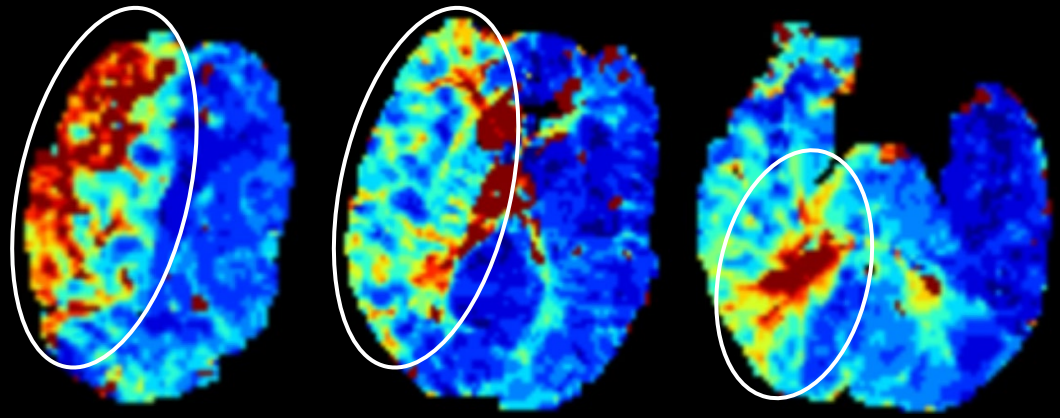
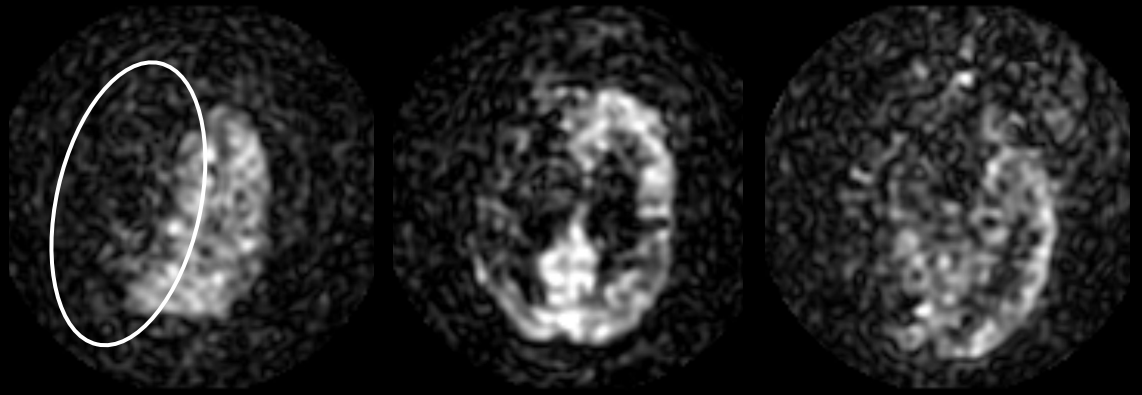
Case 1



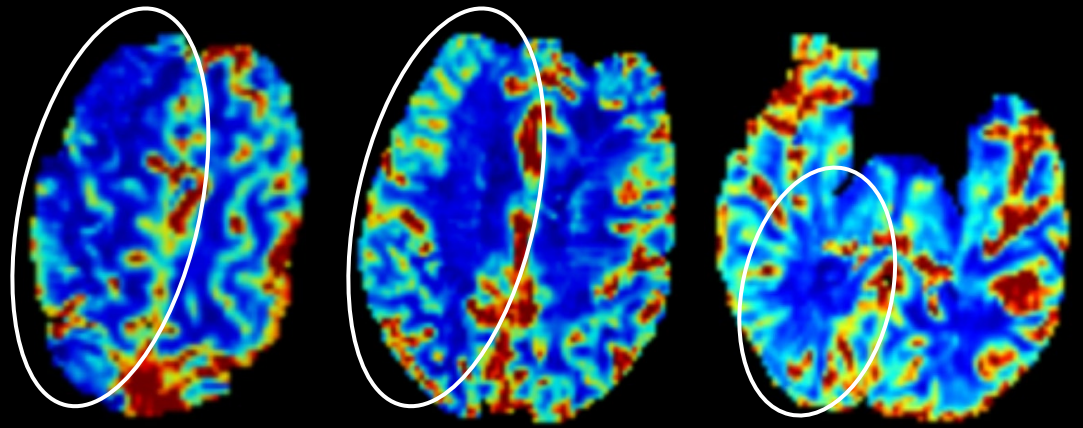
ASL PRE



TMAX



ASL POST



CBF

Case 2

Figure 2. *Bilateral anterior circulation chronic steno-occlusive disease, compatible with moyamoya. New finding of poor cerebrovascular reserve in the left MCA territory.*

(**Prior MRA**) High-grade stenoses of the distal left ICA as well as proximal MCA and ACA. (**Current MRA**) Interval progression of now absent flow-related signal of the distal left ICA as well as proximal MCA and ACA, compatible with steno-occlusive disease.

(**Current DSA**) Bilateral Suzuki grade 2 moyamoya changes, more severe on the right as evidenced by complete MCA occlusion. Pial collateralization arising from the bilateral PCA territories, supplying the right MCA and left ACA territories.

(**DWI/FLAIR/POSTCON**) Scattered small subacute infarcts with enhancement in the left frontoparietal region, indicating the MCA-ACA border zone. Chronic right frontal watershed infarcts. (**ASL perfusion, not shown**) Nondiagnostic due to artifact. (**Post-acetazolamide DSC perfusion**) Severely prolonged Tmax in the bilateral MCA-ACA border zones. (**Dynamic BOLD imaging**) Impaired augmentation in the left MCA territory. Impaired augmentation on the right side from remote infarction.

54-year-old female with multiple myeloma, now several weeks of worsening lower extremity weakness (R > L)

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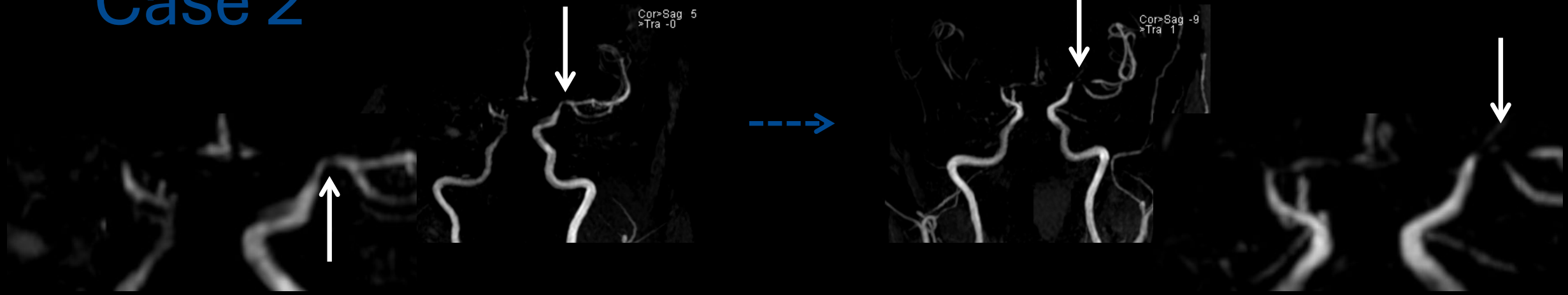
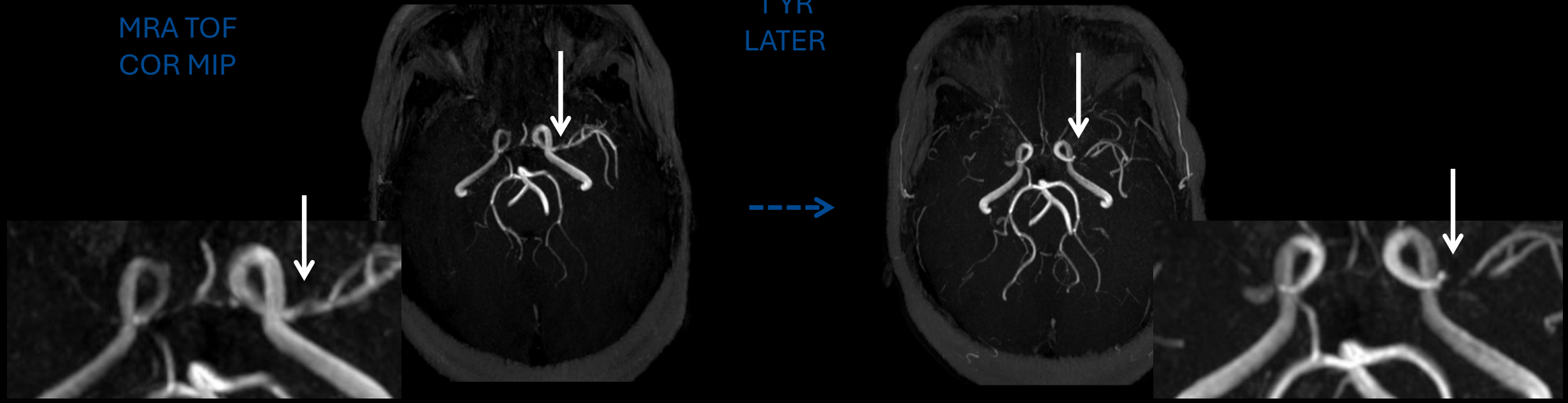
Case 2



MRA TOF
COR MIP

1 YR
LATER



MRA TOF
AX MIP

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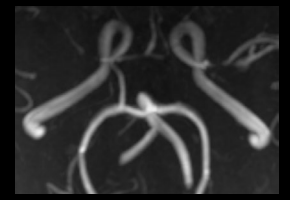
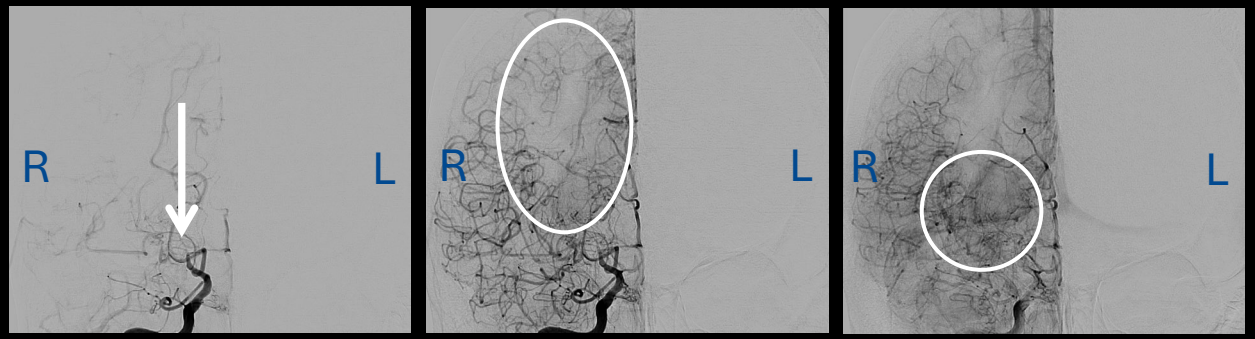
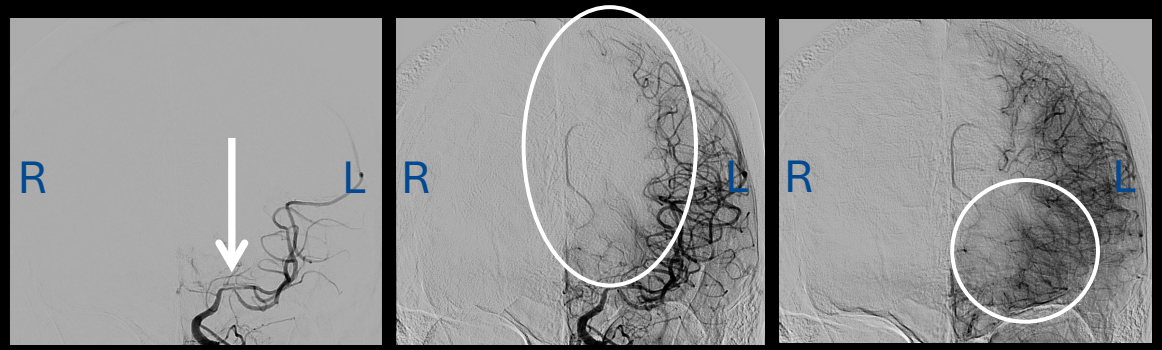
Case 2





DSA
RIGHT FRONTAL



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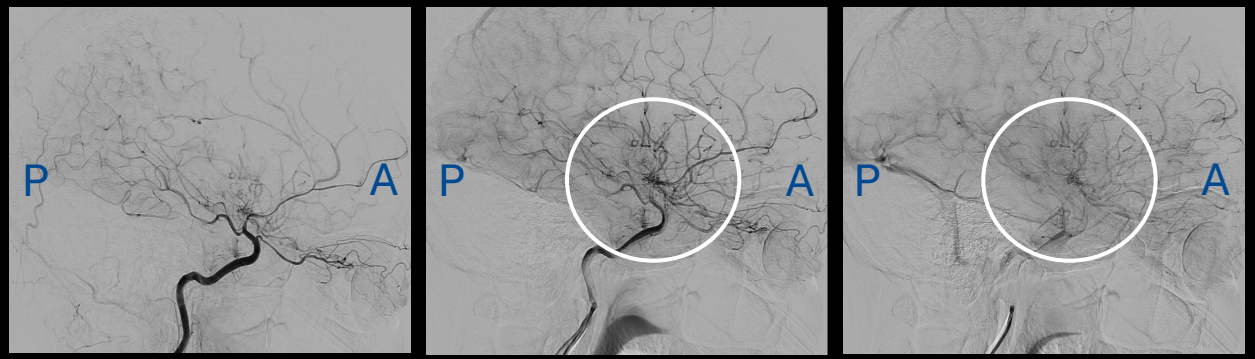

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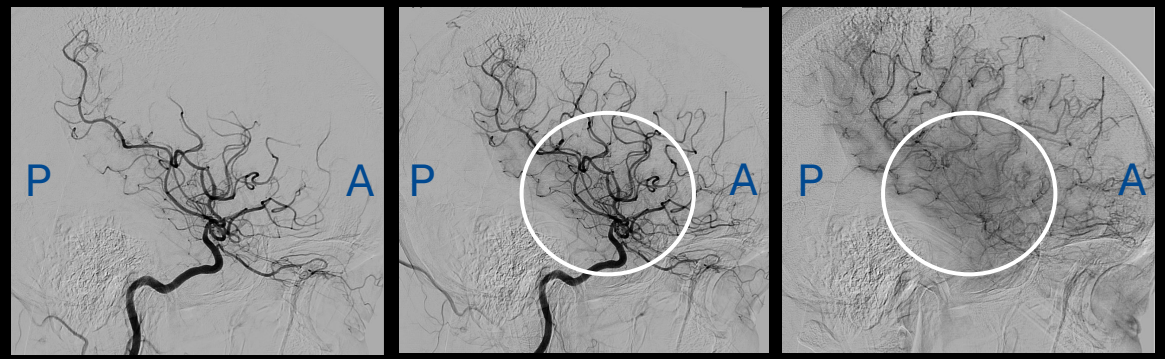
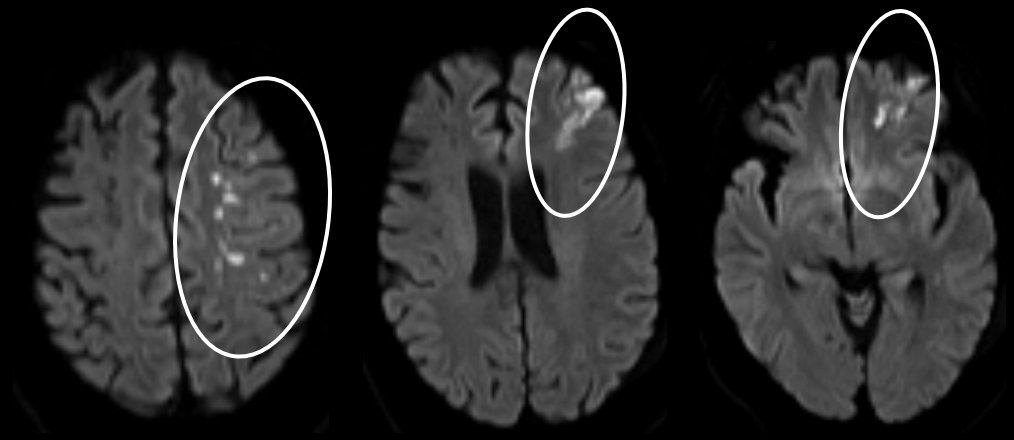
RIGHT LATERAL

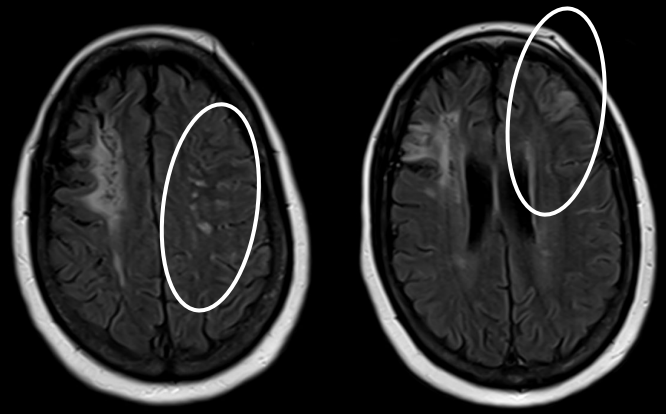


DSA
LEFT LATERAL

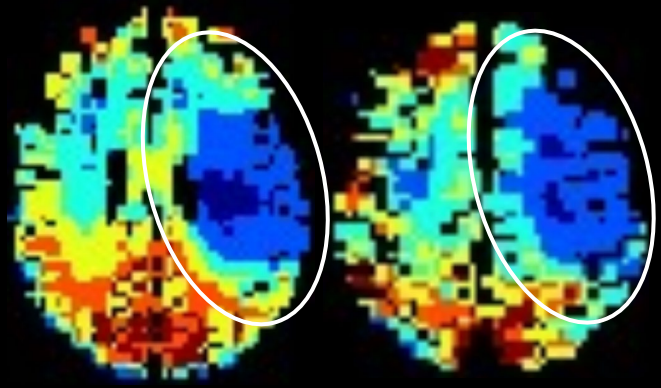
Case 2



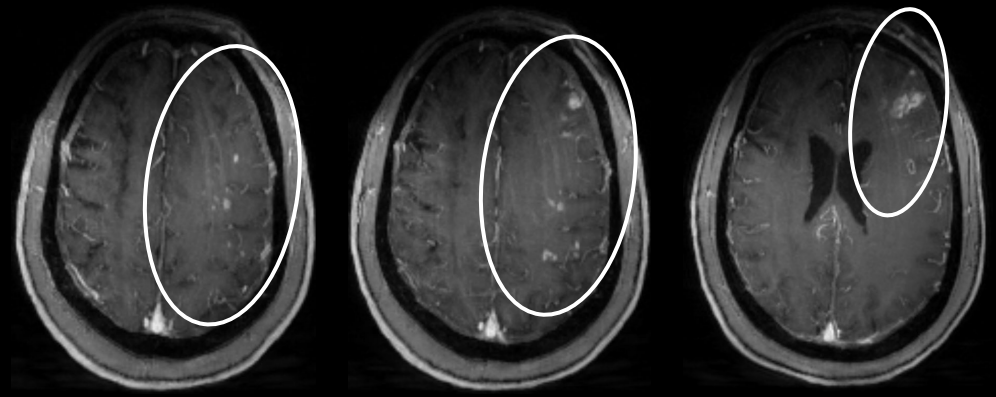
DWI



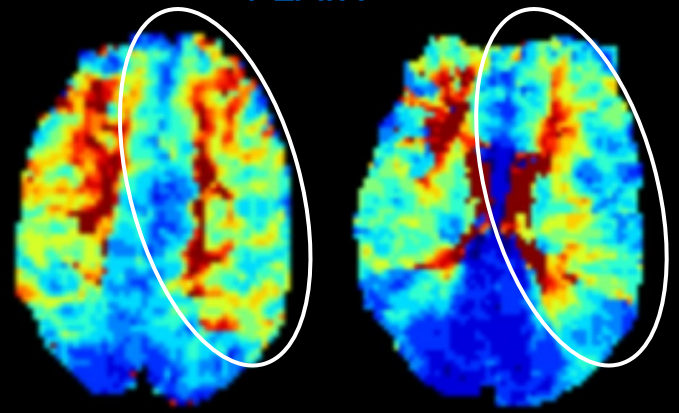
FLAIR



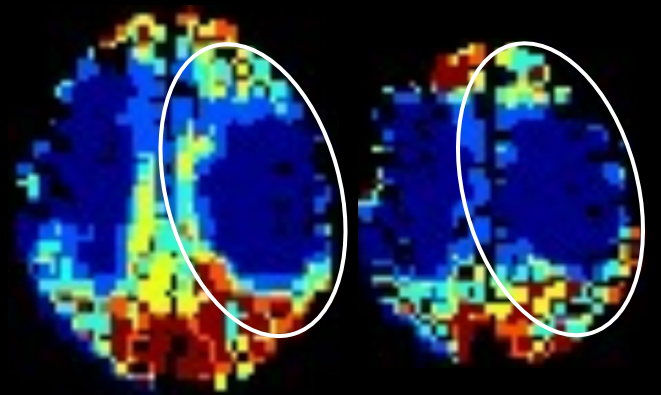
BOLD



POSTCON



TMAX



BOLD (DELAYED)

Figure 3. *Bilateral anterior circulation chronic steno-occlusive disease, compatible with moyamoya. New finding of poor cerebrovascular reserve in the left anterior circulation.*

(**Prior MRA**) Right anterior circulation chronic steno-occlusive disease with lenticulostriate moyamoya changes. (**Current MRA**) Status post right-sided EDAS. Interval development of left anterior circulation steno-occlusive disease, involving the distal left ICA as well as proximal MCA and ACA.

(**Pre- and post-acetazolamide ASL perfusion**) Impaired augmentation throughout the majority of the left MCA and ACA territories, most significant anteriorly. (**Post-acetazolamide DSC perfusion**) Subthreshold areas of prolonged Tmax in the bilateral ACA territories, right greater than left, with a threshold area in the anterior right ACA territory. (**Dynamic BOLD imaging**) Impaired augmentation throughout the bilateral ACA territories, as well as majority of the left MCA territory.

(**Current DSA**) End-stage moyamoya changes in the right anterior circulation with patent EDAS. Evidence of developing moyamoya changes in the left anterior circulation. Interval development of high-grade stenosis of the distal left ICA, and to a lesser extent narrowing of the proximal MCA and ACA. (**Subsequent DSA**) Left-sided injection only. Essentially unchanged moyamoya appearance. Status post left EDAS with excellent synangiosis.

Case 3

22-year-old female with known right-sided Moyamoya arteriopathy, status post EDAS about 6 years prior

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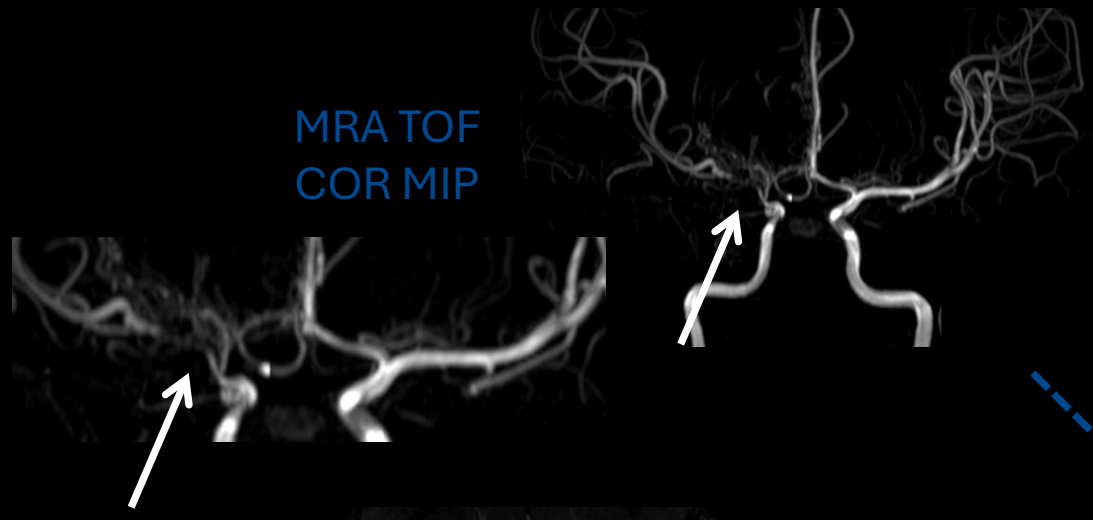
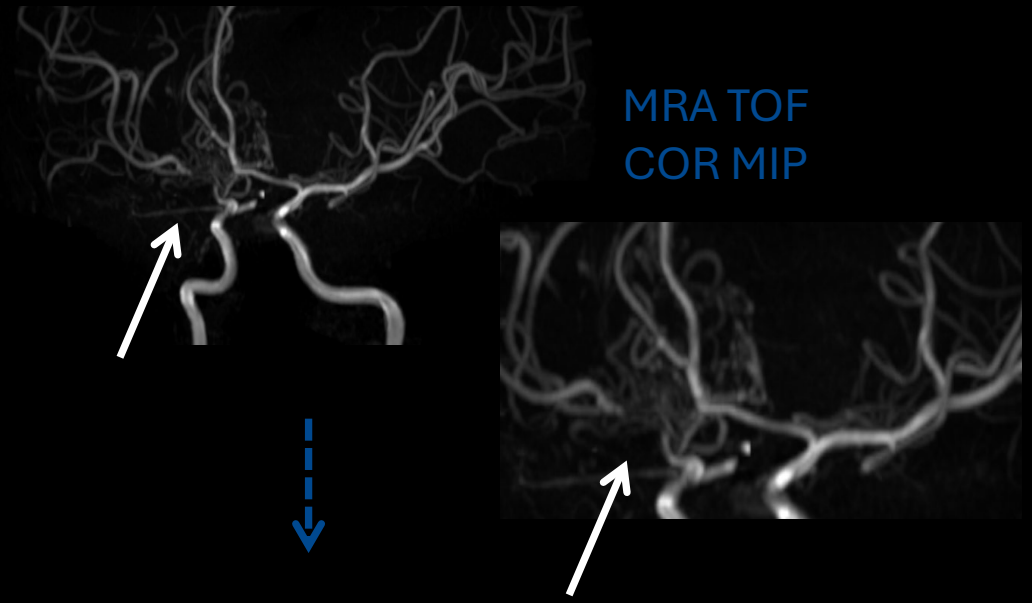
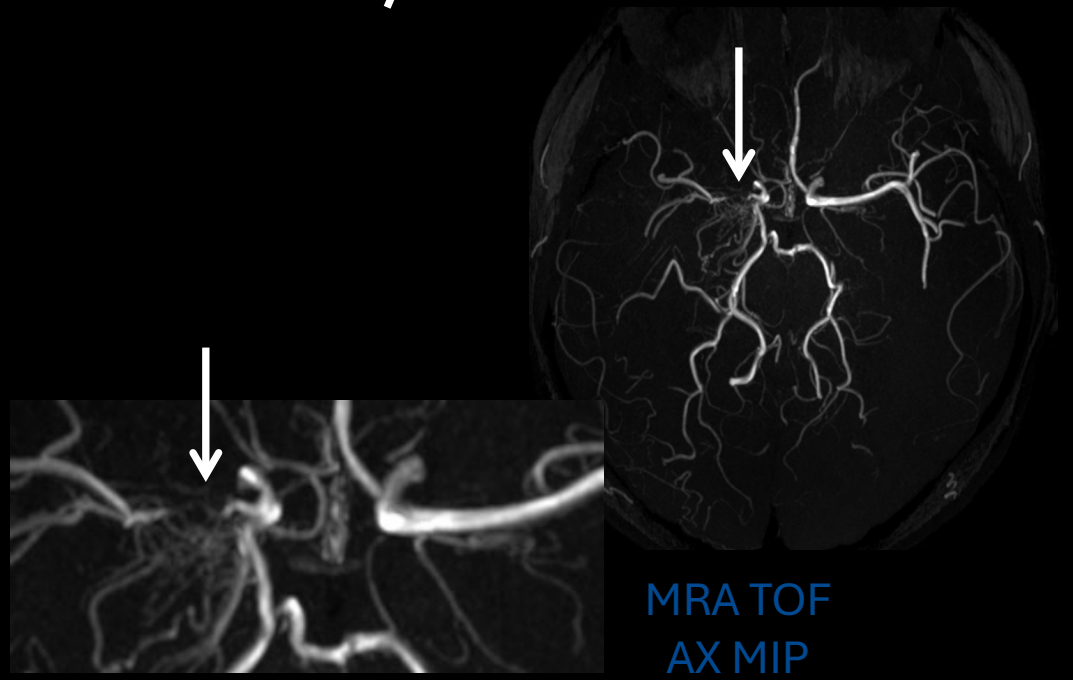
Case 3

MRA TOF
COR MIP

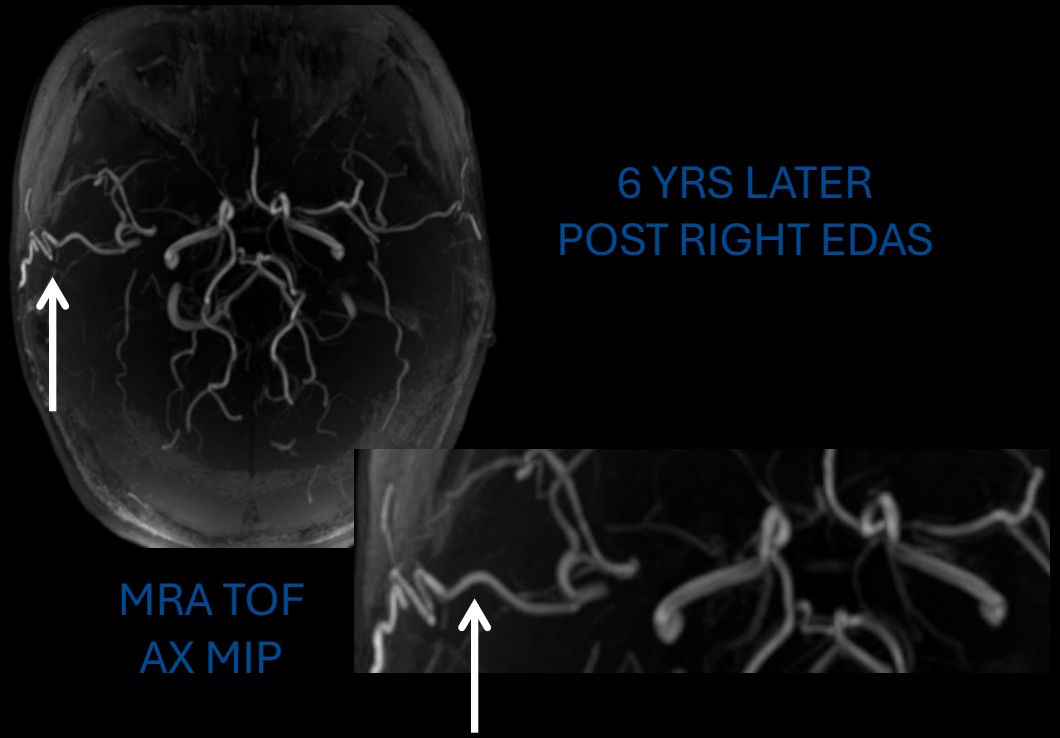


MRA TOF
COR MIP





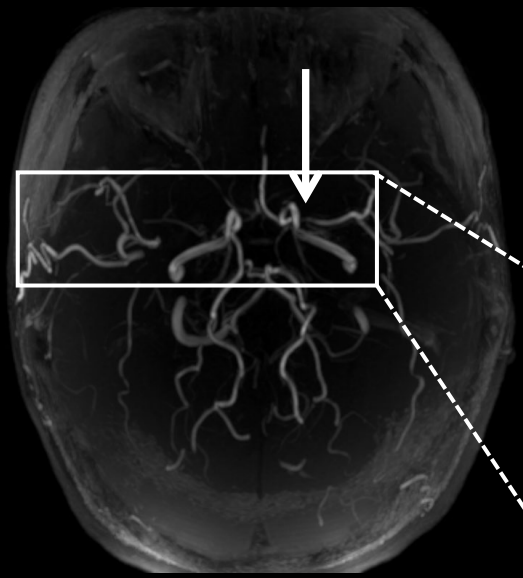
MRA TOF
AX MIP

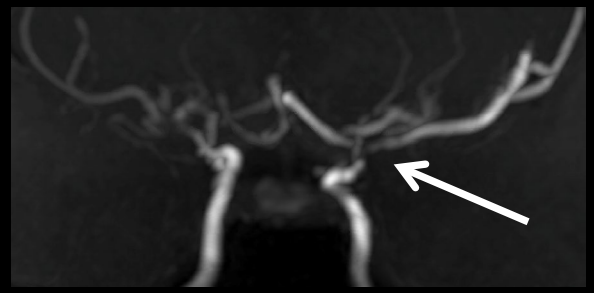


MRA TOF
AX MIP

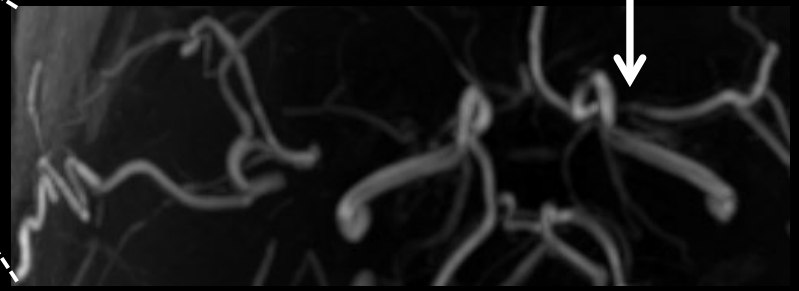
6 YRS LATER
POST RIGHT EDAS

Case 3





MRA TOF (AX AND COR)



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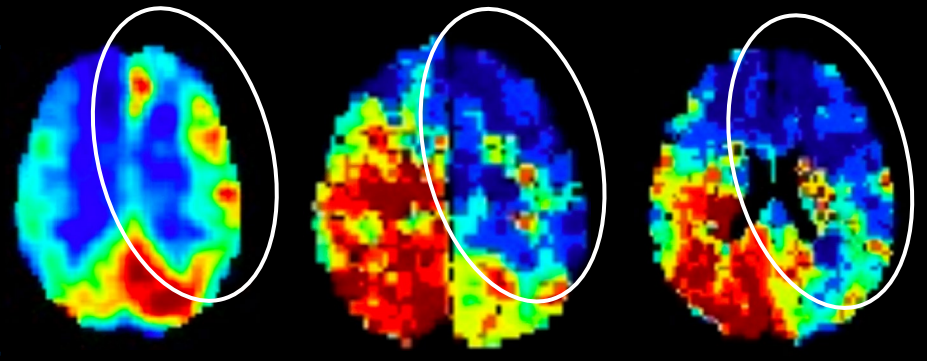
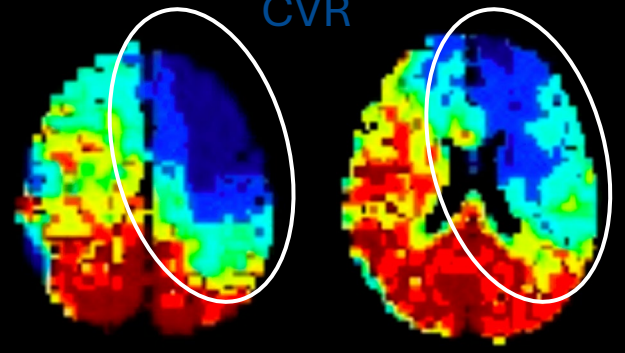
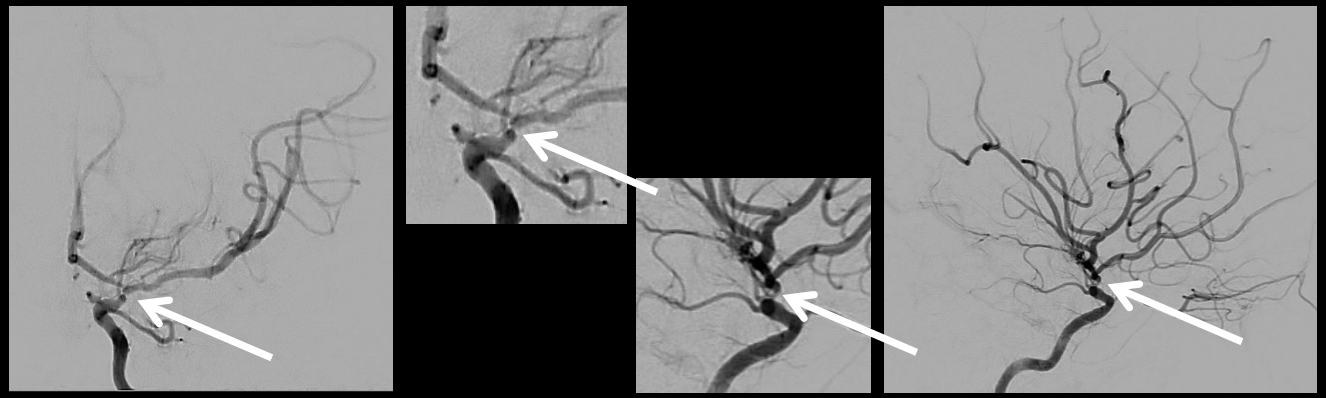


CBF

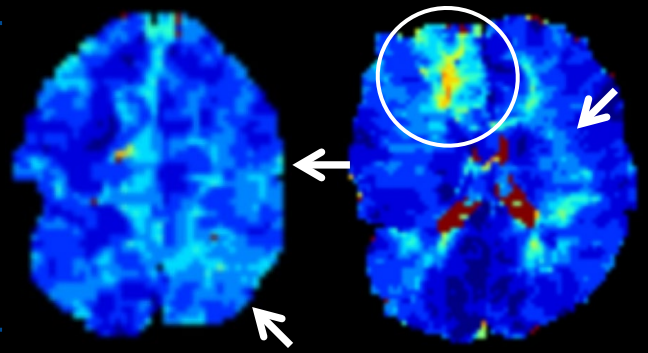
CVR

BOLD





TMAX



DSA (FRONTAL AND LATERAL)

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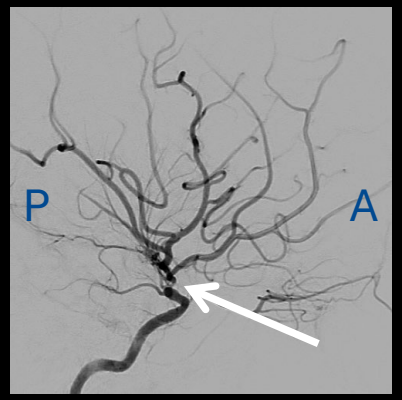
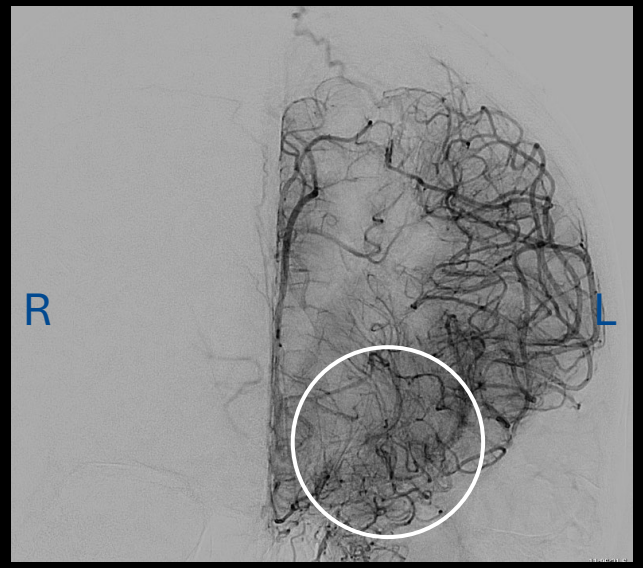
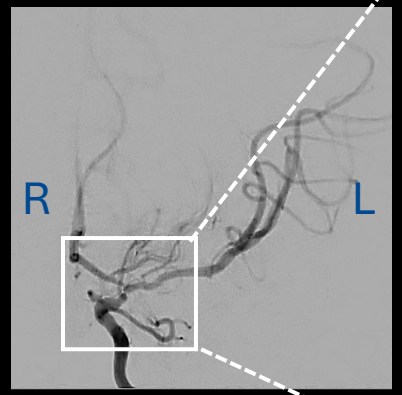
Case 3

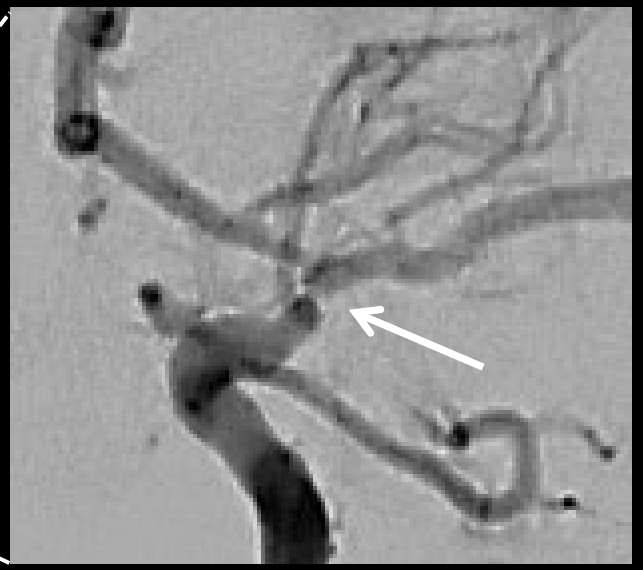


LEFT
LATERAL





LEFT
FRONTAL



6 MO
AFTER
LEFT
EDAS

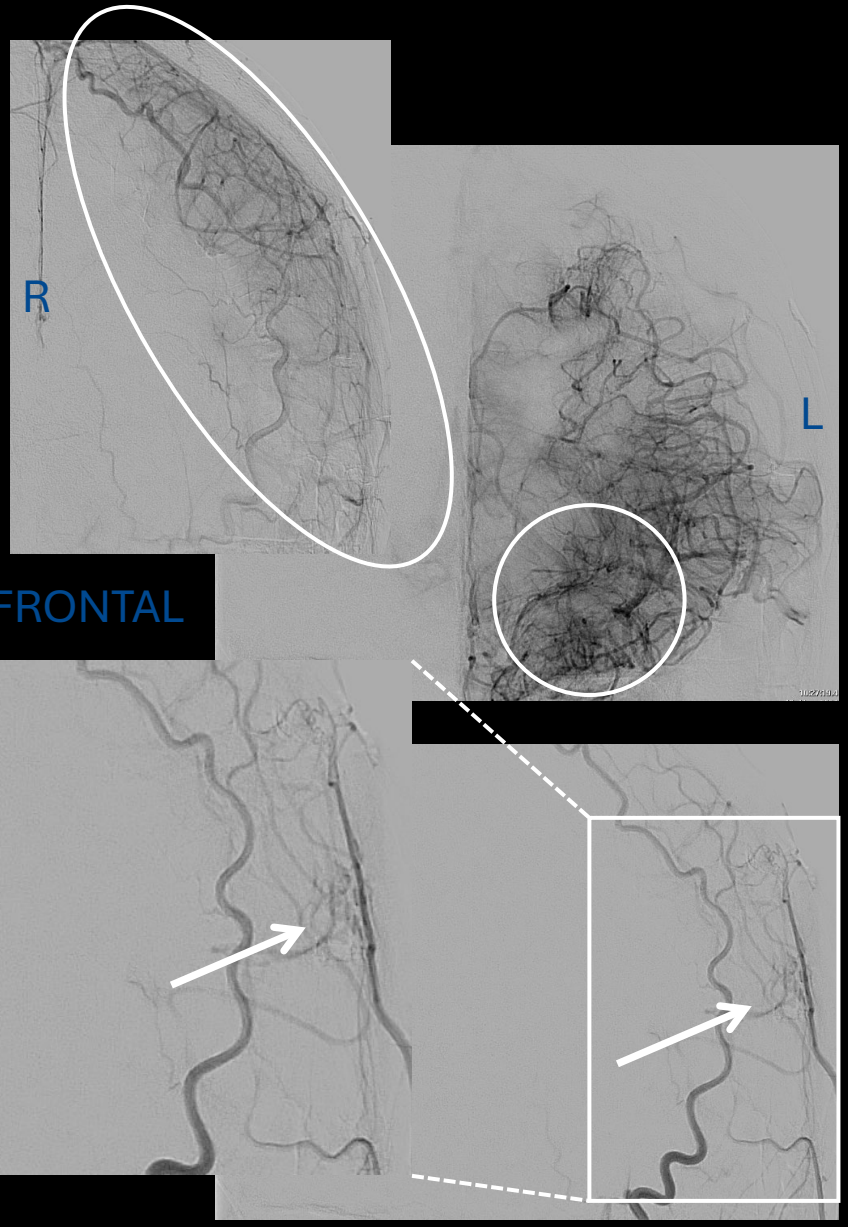


Figure 4. *Right anterior circulation chronic steno-occlusive disease, compatible with moyamoya. Poor cerebrovascular reserve in the anterior right MCA territory.*

(**Prior DSA**) Right moyamoya change, consistent with Suzuki stage 2. Pial collateralization from the posterior circulation to the MCA and ACA territories.

(**Current MRA**) Absent flow-related signal involving the distal right ICA as well as MCA and proximal ACA, with lenticulostriate collateralization. Findings compatible with steno-occlusive disease and moyamoya changes. (**FLAIR**) Chronic microvascular ischemic changes and lacunar infarcts in the bilateral centrum semiovale, right greater than left.

(**Pre- and post-acetazolamide ASL perfusion**) Absent labeling in the left cerebral hemisphere due to artifact from left carotid stent. Reduced baseline CBF in the right anterior MCA and ACA territories. Impaired augmentation in the right anterior MCA territory. (**Post-acetazolamide DSC perfusion**) Prolonged Tmax in the right MCA territory. (**Dynamic BOLD imaging, not shown**) No expected increase in signal, likely due to technical failure.

Case 4

44-year-old female with known bilateral Moyamoya arteriopathy (R > L), status post left CCA angioplasty and stenting about 3 months prior

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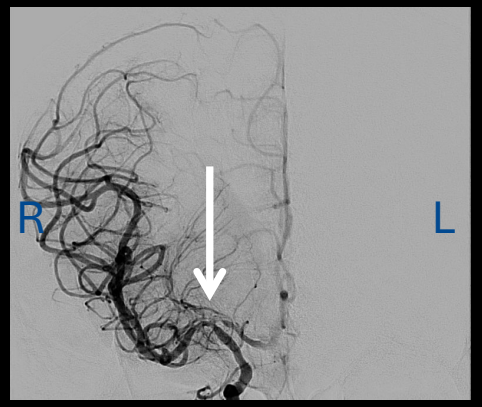
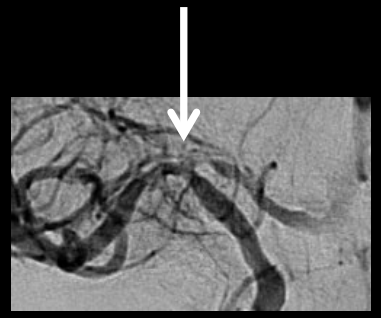
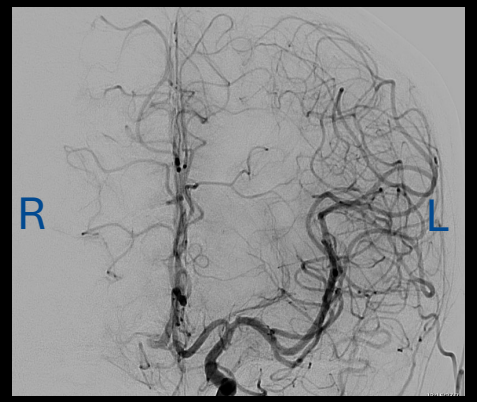
Case 4



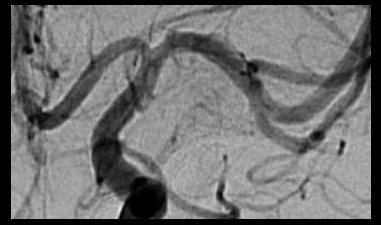
RIGHT FRONTAL

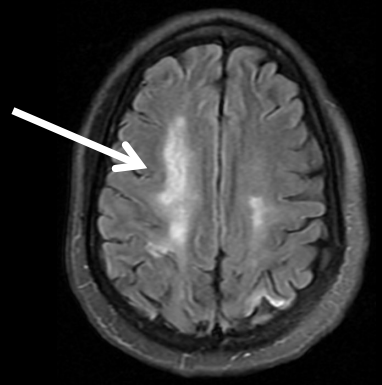


3 MO AFTER
LEFT CCA
STENTING

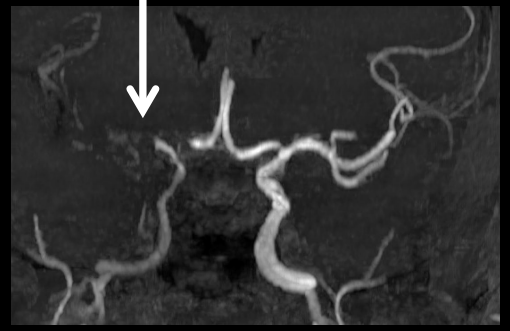


LEFT FRONTAL

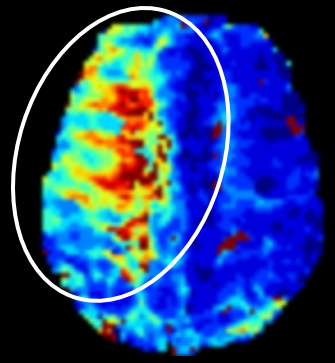




FLAIR

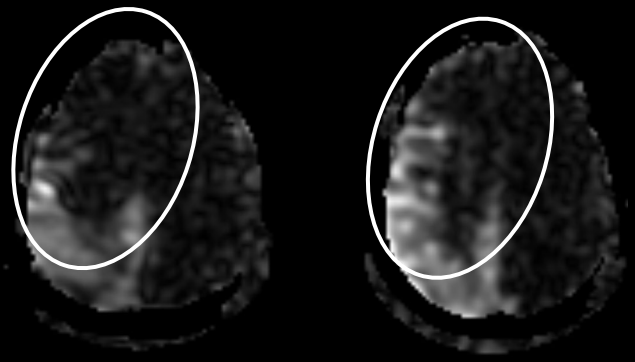


MRA TOF COR

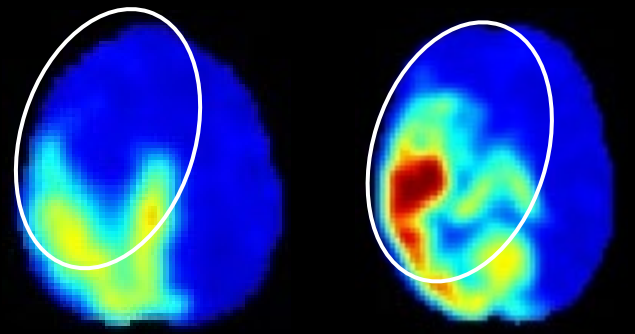


TMAX

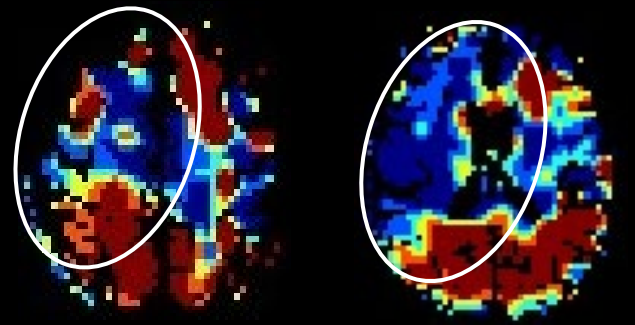
ASL
PRE



ASL
CBF



ASL
CVR



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Case 5

Figure 5. *Bilateral anterior circulation chronic steno-occlusive disease, compatible with moyamoya. Poor cerebrovascular reserve in the anterior right MCA and ACA territories.*

(**Pre- and post-acetazolamide ASL perfusion**) Severely impaired augmentation in the right anterior MCA and ACA territories. Preserved augmentation posteriorly, likely due to PCA to MCA collaterals. (**Post-acetazolamide DSC perfusion, not shown**) Symmetric CBF and CBV. (**Dynamic BOLD imaging**) Severely impaired augmentation in the right anterior MCA and ACA territories.

(**Current MRA**) Absent flow-related signal involving the proximal MCA and ACA, compatible with steno-occlusive disease. (**FLAIR**) Chronic infarct in the high right frontal lobe. (**Current DSA**) High-grade stenosis of the right proximal MCA with lenticulostriate collateralization, compatible with steno-occlusive disease and moyamoya changes.

(**Subsequent DSA**) Patent right-sided STA-MCA bypass graft, which appropriately supplies the right MCA territory. Interval progression of moyamoya changes bilaterally, now with near-occlusive right proximal MCA stenosis as well as high-grade left proximal MCA and ACA stenoses.

37-year-old female with known right Moyamoya arteriopathy (discovered after an episode of post-gestational weakness and confusion, at that time MRI revealed a small right frontal infarct). Now status post right STA-MCA revascularization about 6 months prior.

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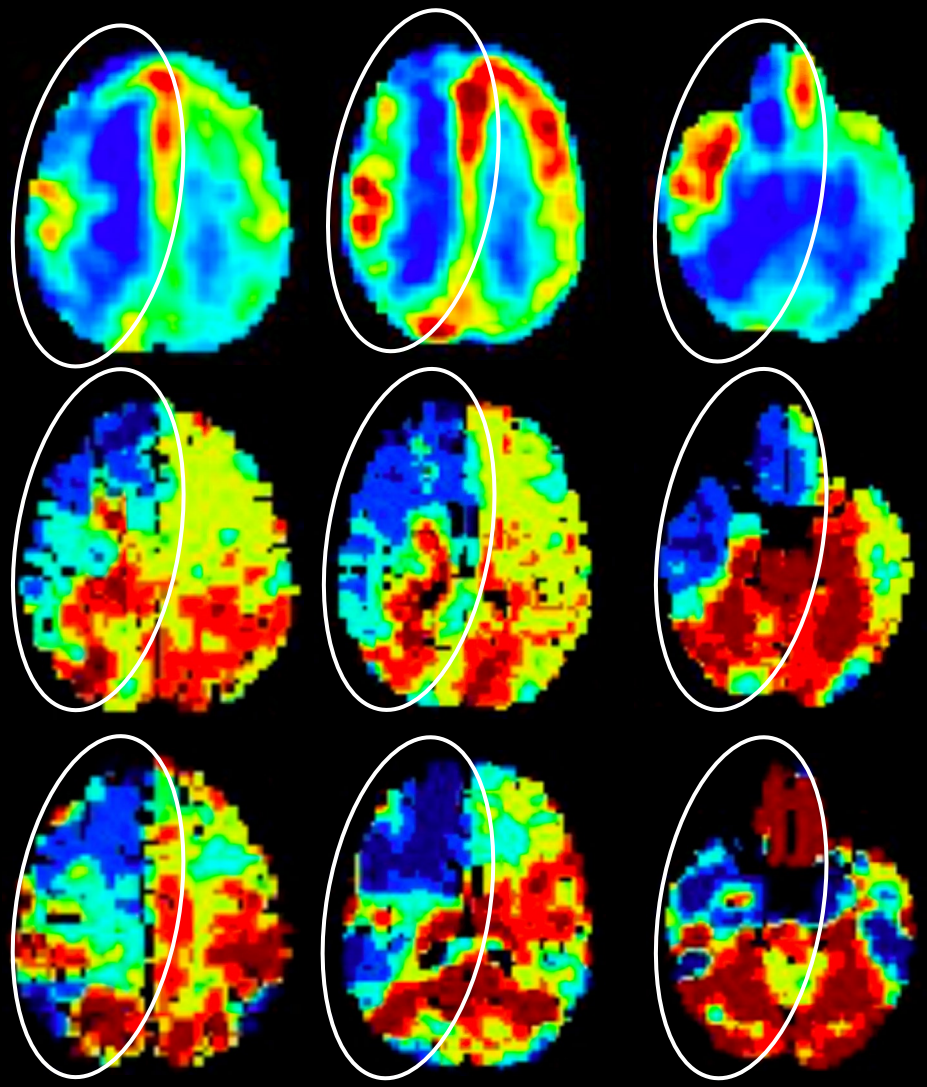
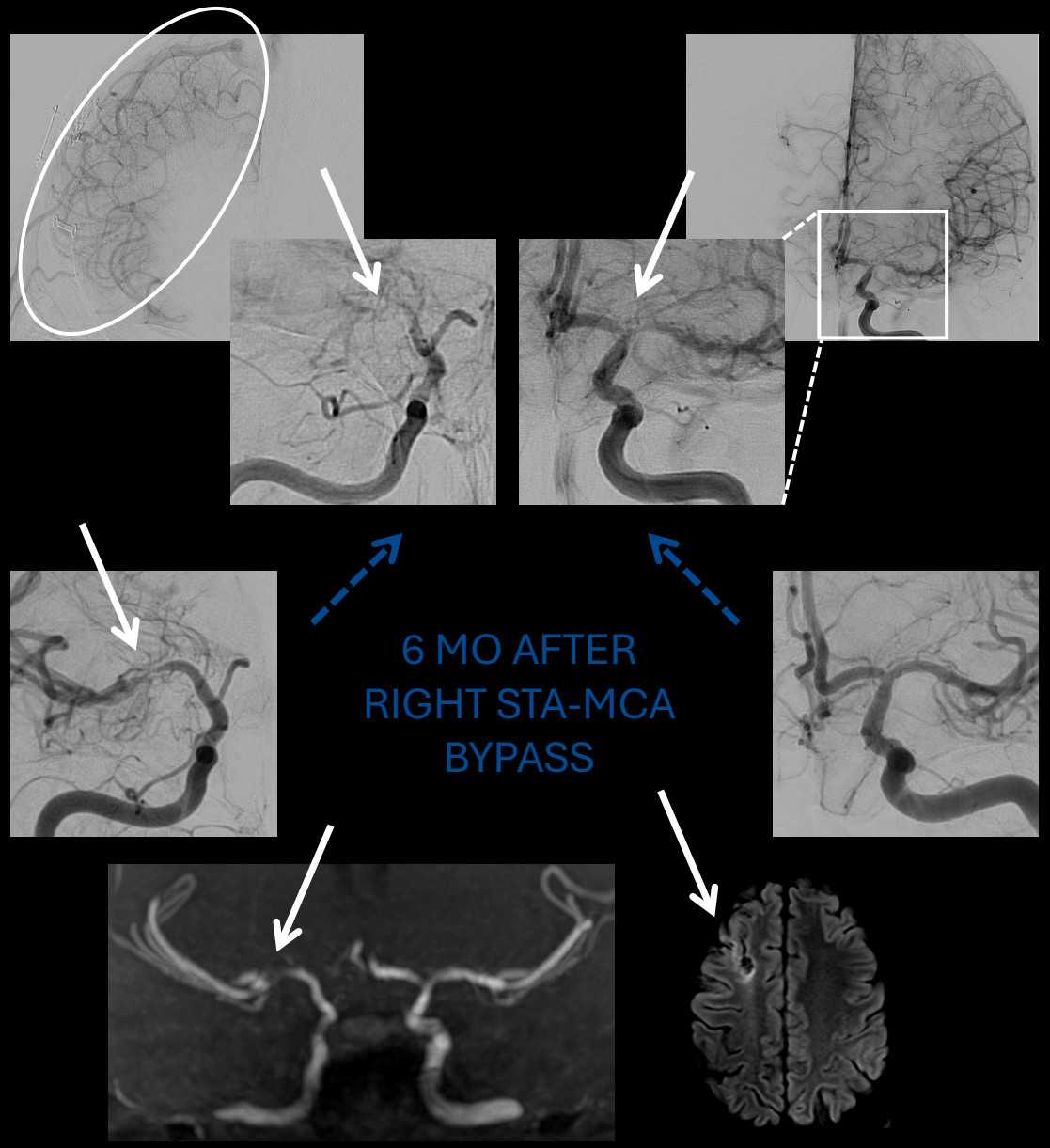
Case 5



ASL CBF

ASL CVR

BOLD CVR



6 MO AFTER RIGHT STA-MCA BYPASS

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Takeaways & Pitfalls

- ✓Impaired augmentation on the right with possible steal from the left shown on BOLD imaging (***Case 1***)
- ✓Although ASL was nondiagnostic due to artifact, assessment of CVR could still be made based on BOLD imaging (***Case 2***)
- ✓DSC perfusion can underestimate the severity and extent of poor CVR (***Case 3***)
- ✓Although ASL labeling failure occurred on the left side due to carotid stenting, assessment of CVR could still be made on the right side (***Case 4***)
- ✓Moyamoya can progress on the order of months* (***Case 5***)

***Query:** Due to less demand from the bypass?

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4. Li J, Jin M, Sun X, Li J, Liu Y, Xi Y, Wang Q, Zhao W, Huang Y. Imaging of Moyamoya Disease and Moyamoya Syndrome: Current Status. J Comput Assist Tomogr. 2019 Mar/Apr;43(2):257-263. doi: 10.1097/RCT.0000000000000834. PMID: 30589721; PMCID: PMC6426357.

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