

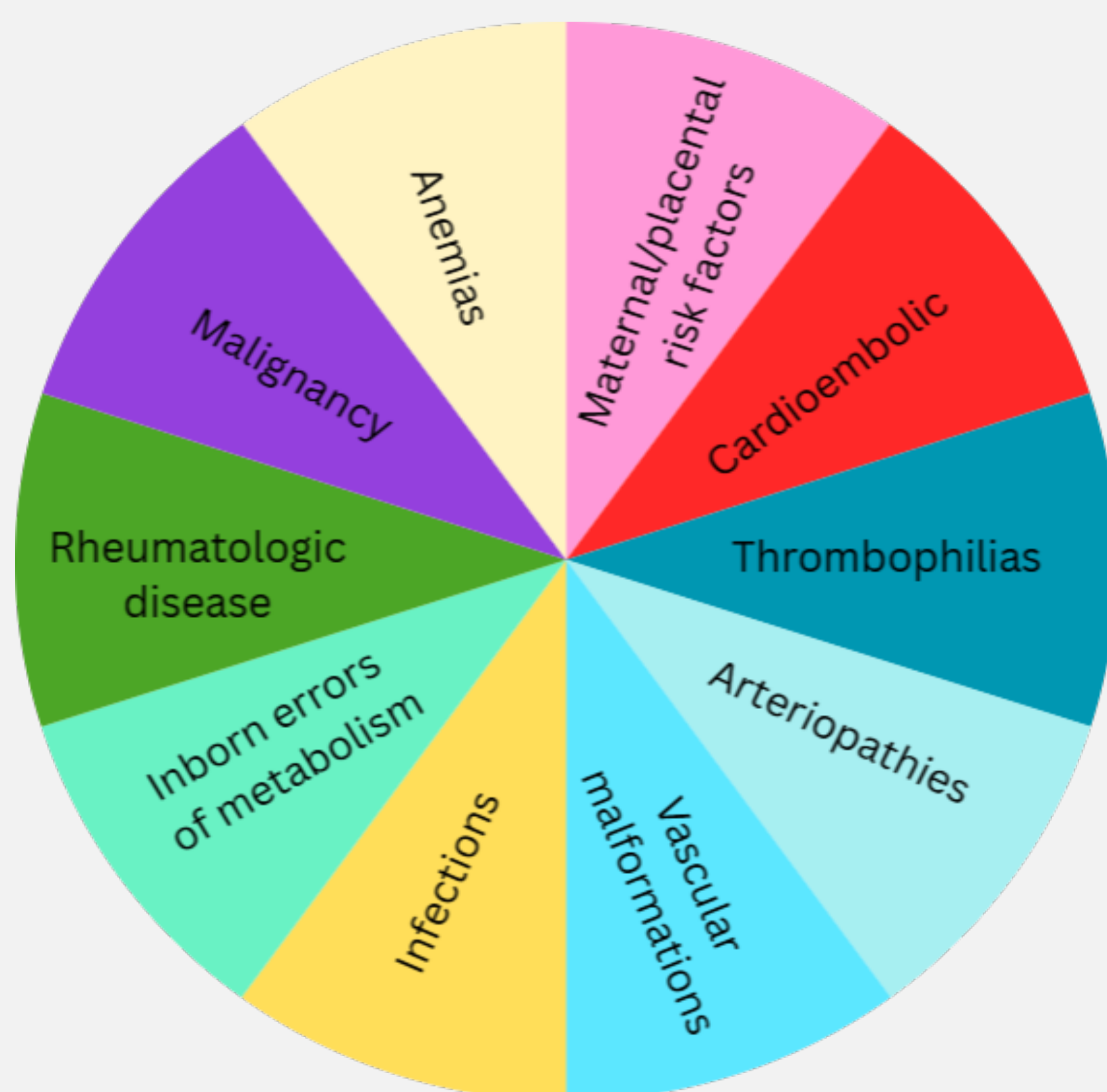
Chronic Monofocal Cerebral Arteriopathy in a Toddler: When Etiology Remains Elusive

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INTRODUCTION

- Pediatric arterial ischemic strokes have an incidence of 1-13 per 100,000 children. They are associated with numerous perinatal and childhood risk factors distinct from adult ischemic strokes¹.
- Etiology of pediatric stroke may be more complex than initial evaluation and imaging may indicate.

Etiology of Pediatric Ischemic Strokes



CASE DESCRIPTION

- Our case describes a two-year-old female presenting with acute left-sided weakness, found to have an acute right middle cerebral artery distribution stroke on a rapid stroke MRI. She received alteplase in the emergency department due to initial concerns of an acute embolic stroke given concerns of a significantly narrowed terminal internal carotid artery.
- Head CT and CT angiogram obtained one hour after initial imaging noted diffuse encephalomalacia of the right hemisphere and a small, remote deep frontal lobe white matter infarct. Upon revisiting the initial MRI, the volume loss of the right hemisphere and remote right frontal deep white matter stroke was present but not commented on in the report.
- She was subsequently felt to have focal cerebral arteriopathy and treated with one dose of high-dose intravenous methylprednisolone as part of the NIH FOCAS trial before being excluded due to the small remote stroke and chronic cerebral atrophy seen on the MRI and CT suggesting pre-existing vascular insufficiency.
- Diagnostic catheter-directed cerebral angiogram confirmed a moderately severe stenosis of the right M1 associated with a linear filling defect extending into the right M1 and right A1 branches. There was also an intimal flap, suggestive of an acute intracranial dissection.
- She received anticoagulation with intravenous heparin followed by low-molecular weight heparin and aspirin due to concern for chronic vasculopathy and progressive symptoms clinically (stable radiologically).
- Echocardiogram and thrombophilia work-up were normal. Whole exome sequencing as part of her arteriopathy workup showed an unexpected incidental de novo pathogenic variant in DYNC1H1 gene unrelated to her presenting phenotype, but was otherwise normal.

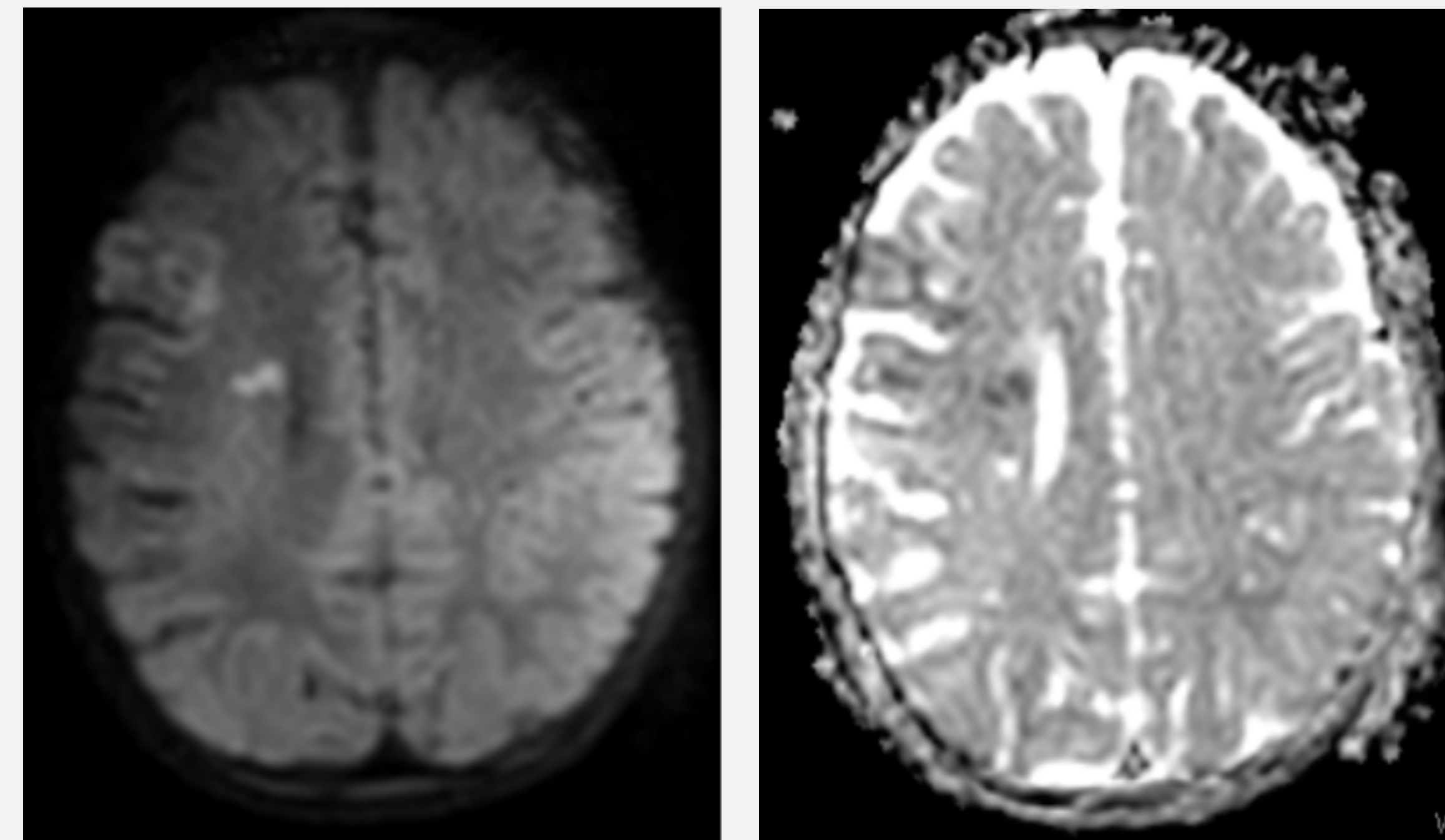


FIGURE 1: Initial MR stroke protocol imaging axial images with the following findings: Diffusion restriction sequence showing hyperintensity in the right MCA distribution (left). Correlated hypointensity in the right MCA distribution on apparent diffusion coefficient sequence (right).

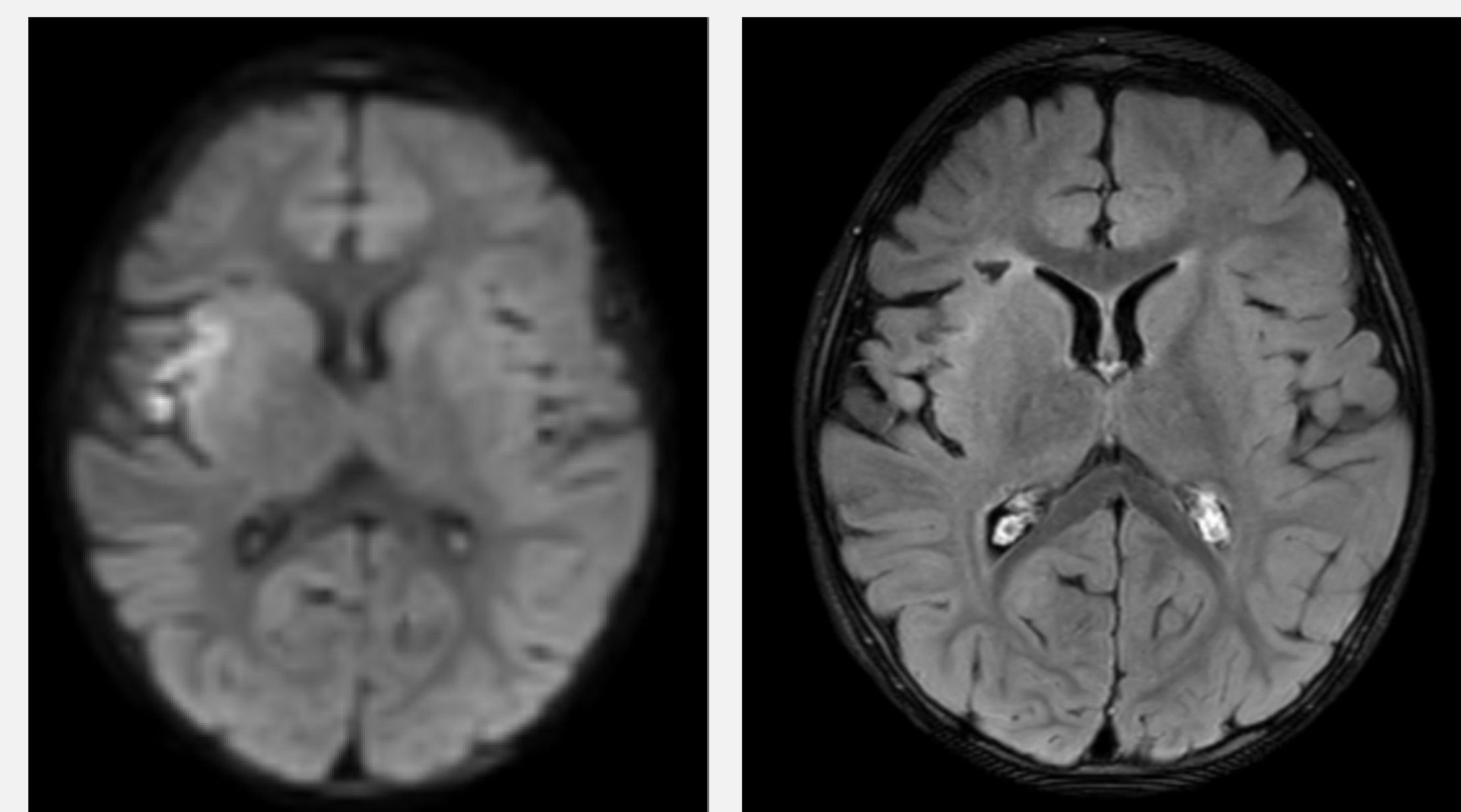


FIGURE 2: MRI brain with and without contrast on hospital day 2 showing evidence of acute right frontal lobe ischemia as well as right cerebral volume loss and gliosis consistent with chronic ischemia. Axial diffusion restriction (left) and FLAIR (right) sequences shown.

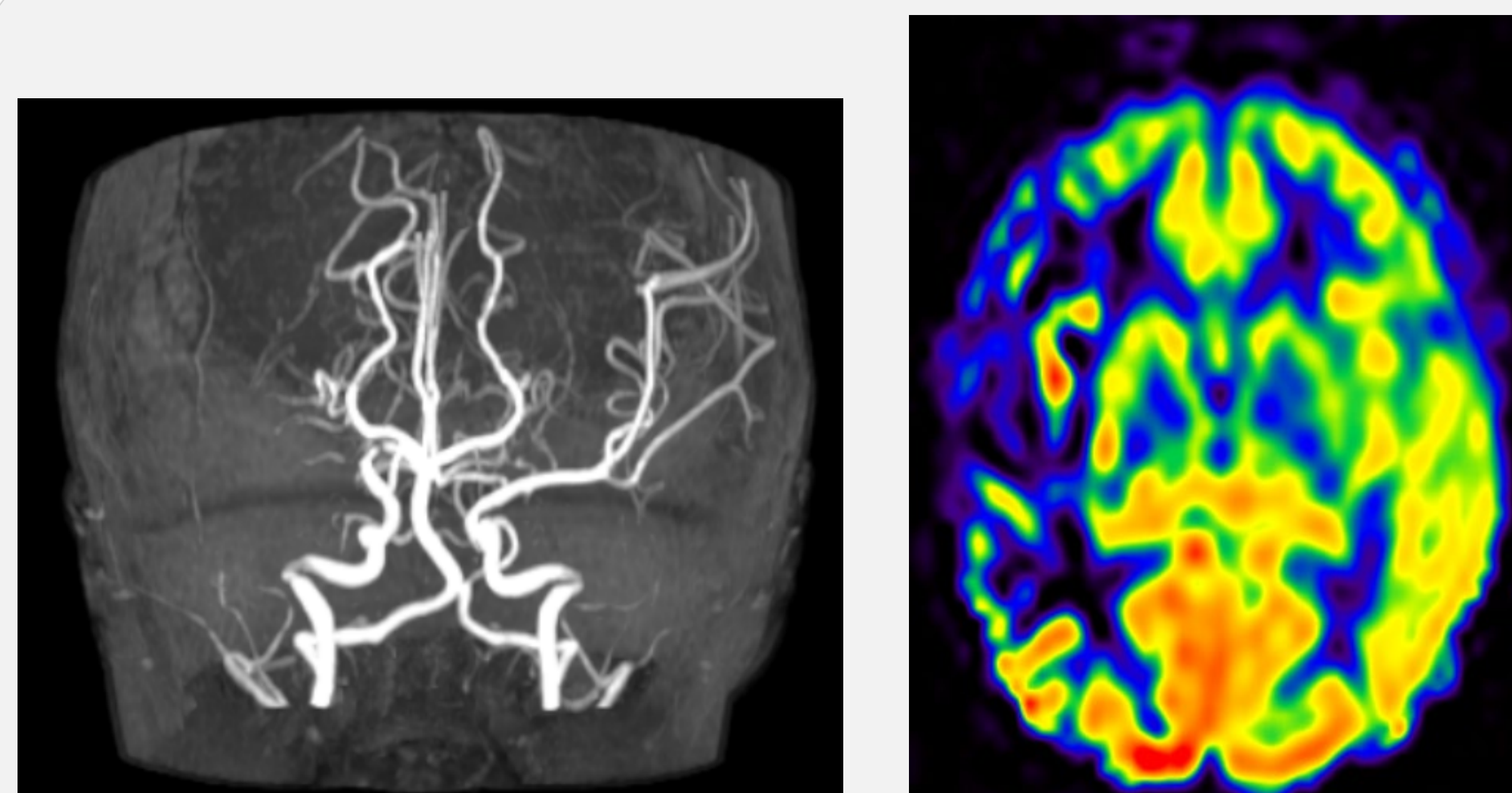


FIGURE 3: MRA head on hospital day 2 showing absence of right middle cerebral artery flow (left). Imaging was also noted to have vessel wall enhancement. Arterial spin labeling shows absent flow to the right frontal and parietal lobes (right).

DISCUSSION

- Identifying ischemic stroke etiology is important, not just for treatment, but for communicating risk of recurrence and prognosis
- Persistent neurologic deficits occur in 70% of children who have sustained a stroke, of which 36% are mild, 23% are moderate, and 10% are severe². Arterial ischemic strokes have a recurrence rate of 12-20% during childhood³.
- In regard to craniocervical arterial dissection, those with genetic or acquired connective tissue disorders, male gender, or trauma are more susceptible to dissection¹. To the best of our knowledge, our patient did not have these risk factors.
- Vessel dissection in the pediatric population most commonly affects extracranial vessels, including vertebral artery and posterior circulation¹. Our patient had an intimal flap within an intracranial vessel, which may be associated with an underlying malformation given the presence of stenosed vessels.
- Although the patient was excluded from the FOCAS trial, she did have findings consistent with a transient cerebral arteriopathy on angiography. These findings are more common in children with abnormal lipid profiles, familial cardiovascular disease, and viral infections¹. This may be a further line of inquiry in the assessment of etiology and comorbidities.
- Through our patient evaluation, we discovered acute and chronic changes that we believe may be due to repeat ischemic insults. Our patient's etiology remains elusive, but identification and management of her stenosed vessels and acute intracranial dissection may have improved her outcome, as she only has mild left-sided hemiplegia.

REFERENCES

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3. Mastrangelo, M., Giordo, L., Ricciardi, G. et al. Acute ischemic stroke in childhood: a comprehensive review. *Eur J Pediatr* 181, 45–58 (2022). <https://doi.org/10.1007/s00431-021-04212-x>