MOYAMOYA DISEASE

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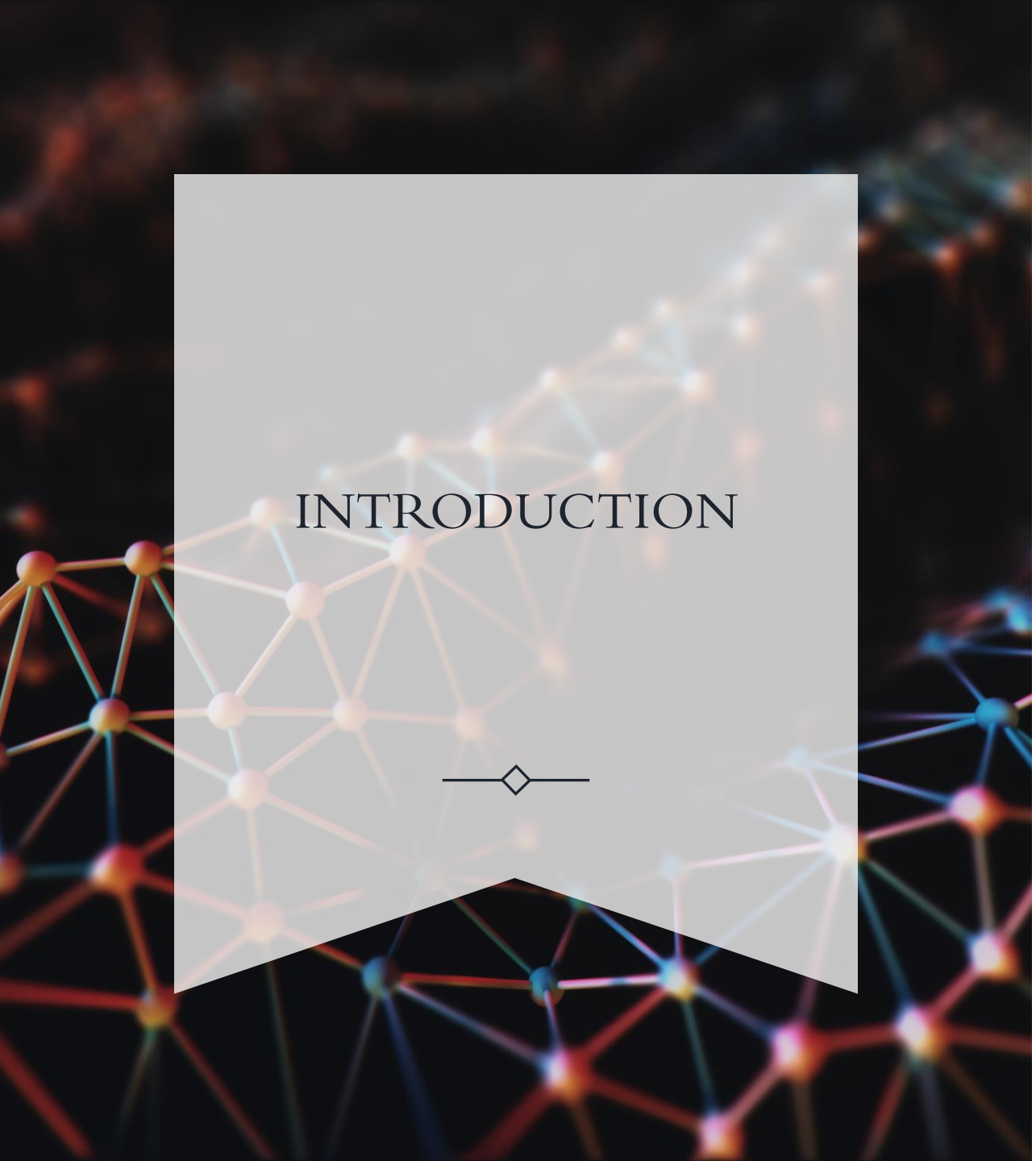
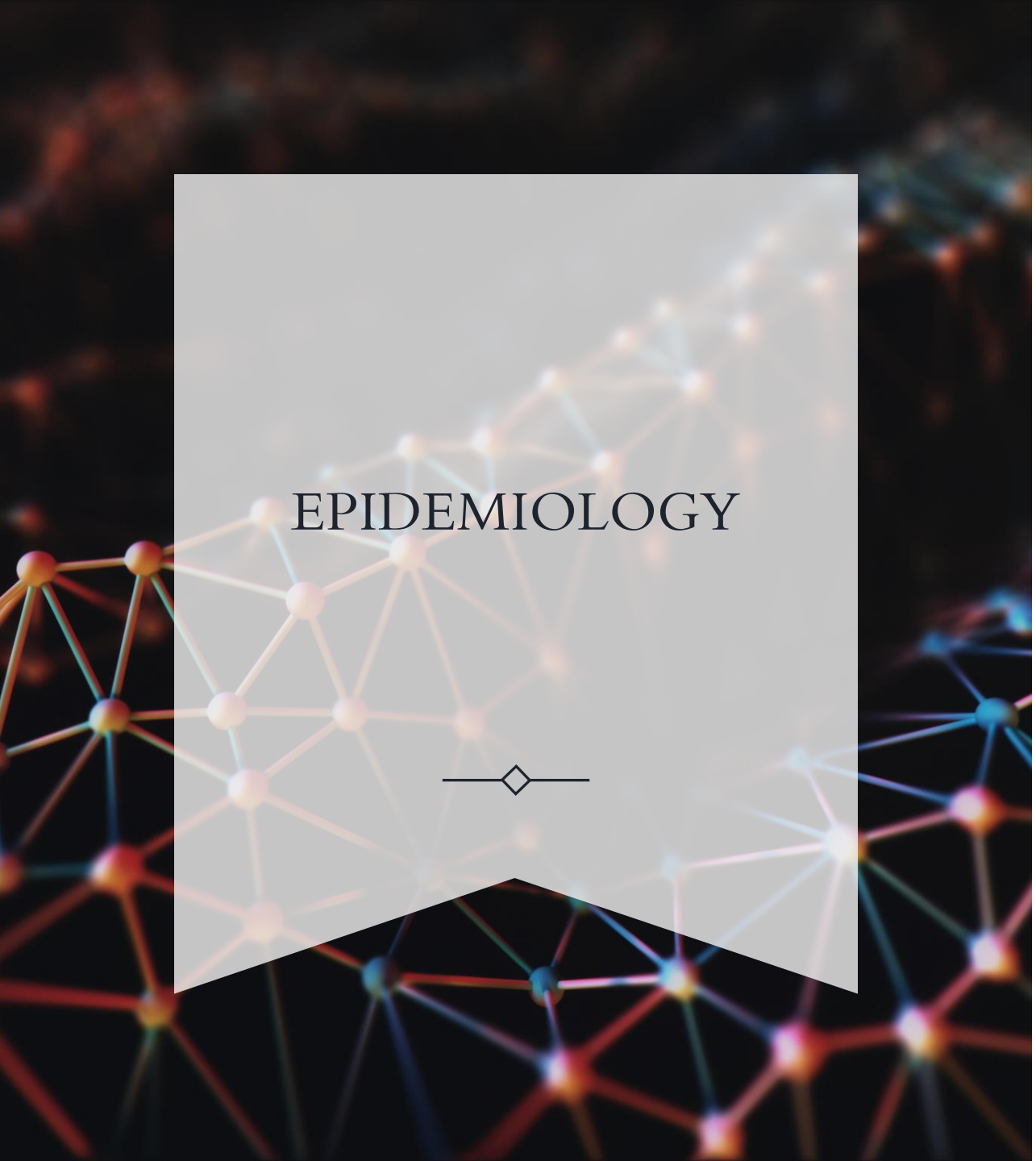


INTRODUCTION

- Moyamoya disease (a hazy puff of smoke) in japanese, an apt name considering it is a chronic, occlusive cerebrovascular disease affecting ICA bilaterally and / or ACA and MCA.
- Moyamoya syndrome is the moyamoya vasculopathy associated with underlying conditions such as sickle cell disease, down's syndrome, noonan syndrome, neurofibromatosis type 1 etc.

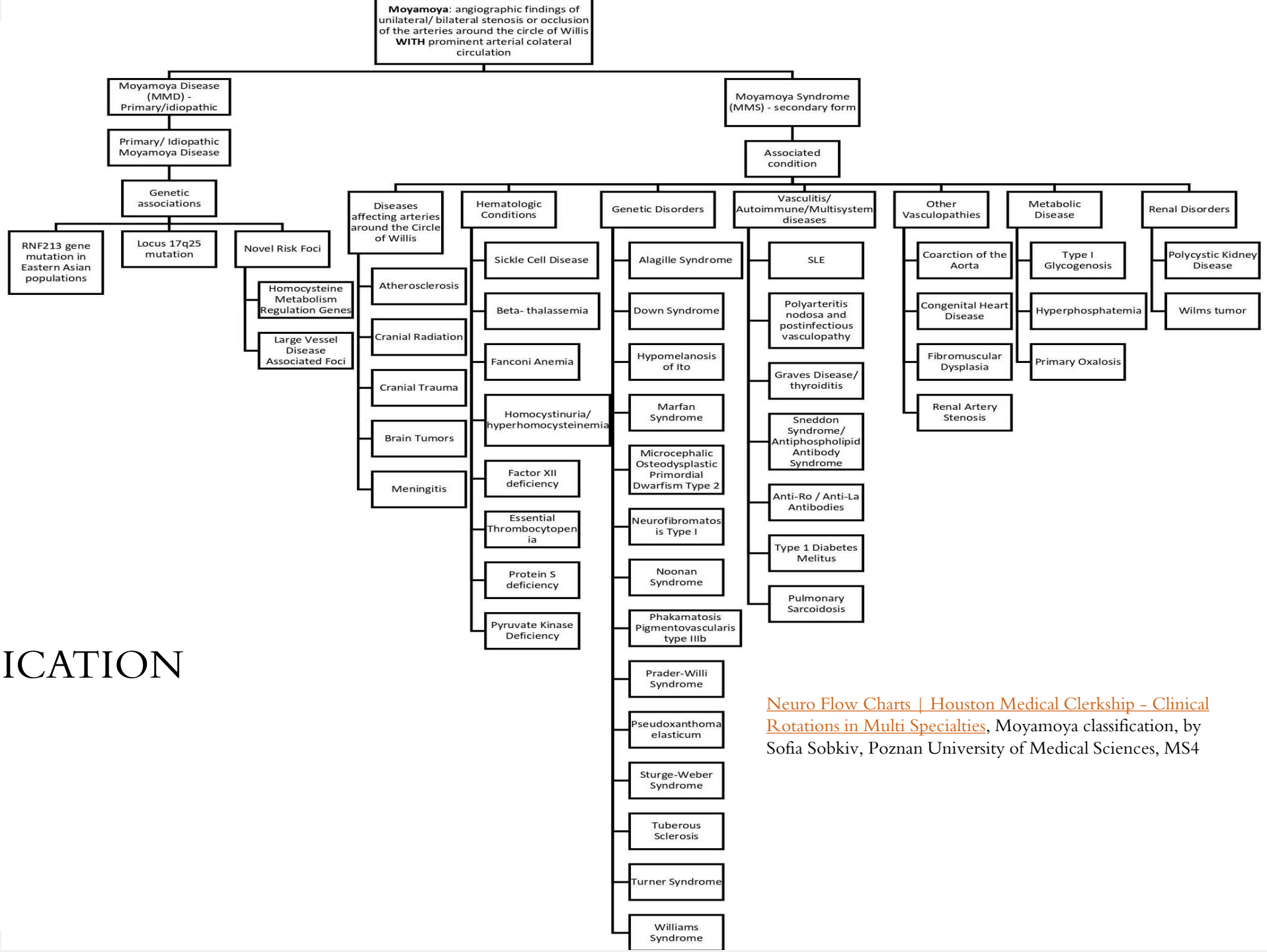


EPIDEMIOLOGY

- Regional preferences are seen with this disease as it affects East Asia most prominently, specifically Japan, South Korea with prevalence rates of 10.5/100,00 and 16.1/100,000 individuals respectively.
- Upward trend is seen in the US. On average, incidence of 0.09/10,000 is reported.

Zhang H, Zheng L, Feng L. Epidemiology, diagnosis and treatment of moyamoya disease. Exp Ther Med. 2019 Mar;17(3):1977-1984. doi: 10.3892/etm.2019.7198. Epub 2019 Jan 25. PMID: 30867689; PMCID: PMC6395994

CLASSIFICATION



Neuro Flow Charts | Houston Medical Clerkship - Clinical Rotations in Multi Specialties, Moyamoya classification, by Sofia Sobkiv, Poznan University of Medical Sciences, MS4

THE BASICS

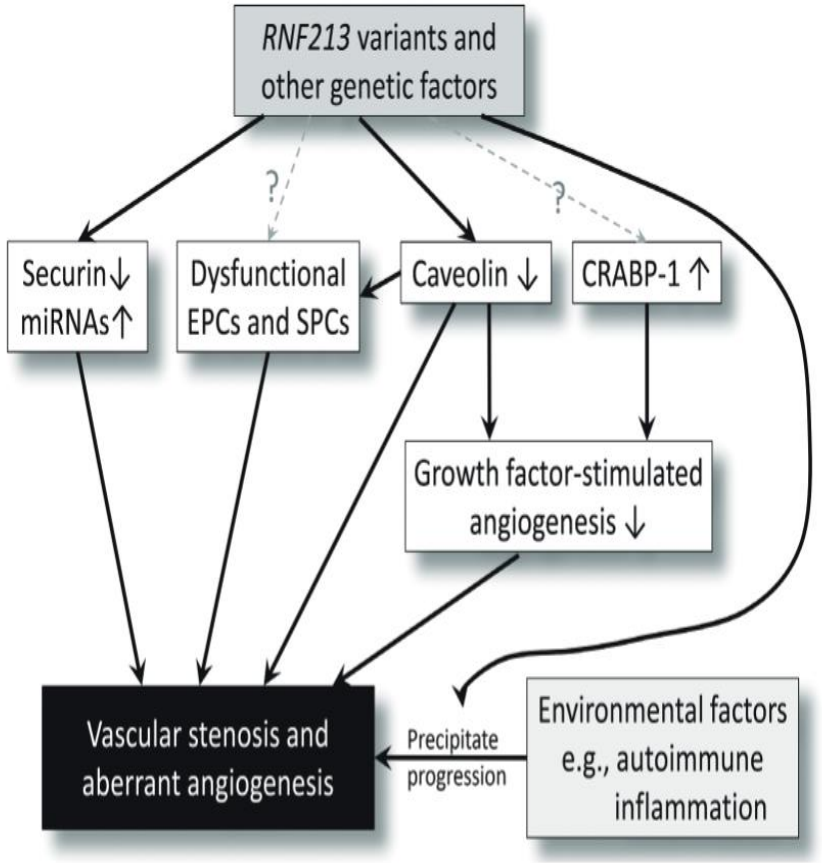


Figure 1. Potential mechanisms of moyamoya disease. The association between genetic, circulating, and environmental factors. *RNF213, Ring finger 213;* EPCs, endothelial progenitor cells; SPCs, smooth muscle progenitor cells; miRNAs, microRNAs; CRABP-1, cellular retinoic acid-binding protein-I.

http://j-stroke.org, http://dx.doi.org/10.5853/jos.2015.01760

THE BASICS

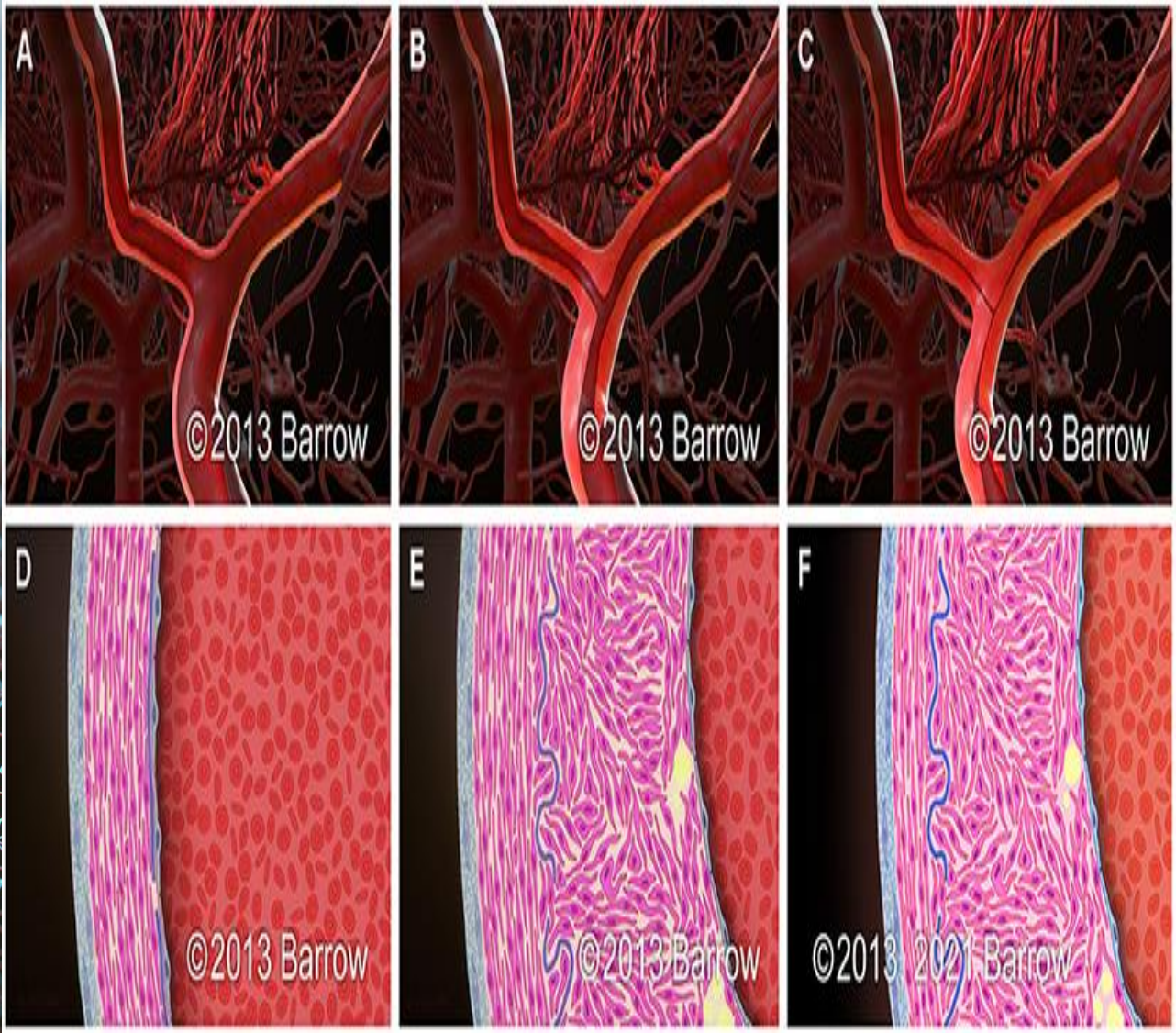
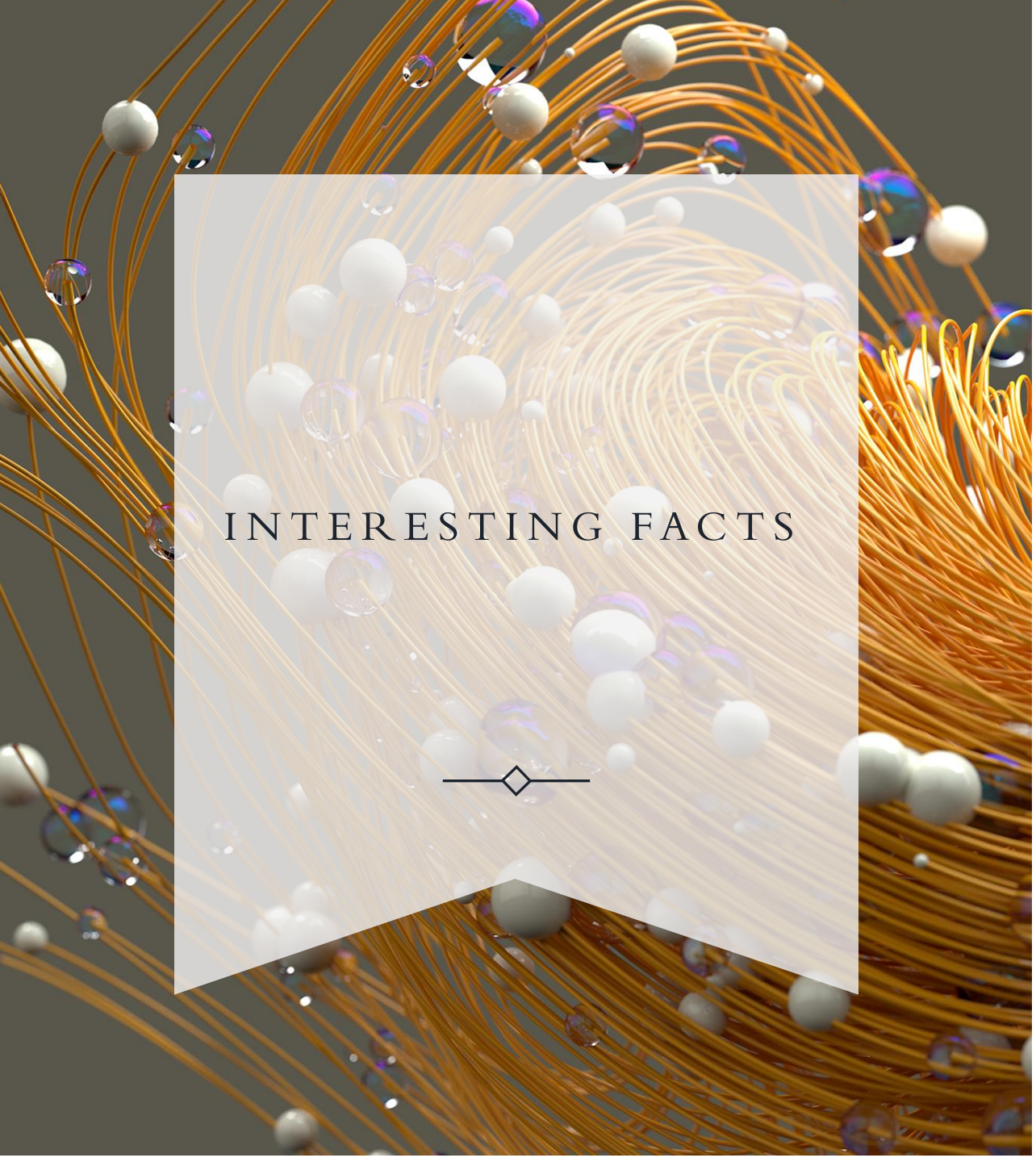


Figure 2. Schematic representation of the paradigmatic progression pattern of arterial stenosis in moyamoya on the macroscopic **(A–C)** and microscopic **(D–F)** scale. **(A,D)** Normal terminal ICA. **(B,E)** Early in the progression of moyamoya, intimal hyperplasia leads to the development of luminal stenosis without a significant change in the outer diameter of the arterial segment. **(C,F)** As luminal stenosis continues to progress in moyamoya, attenuation of the medial layer of the artery leads to a reduction in the outer arterial diameter.

Front. Neurol., 03 September 2021
Sec. Stroke
Volume 12 - 2021 | https://doi.org/10.3389/fneur.2021.661578



INTERESTING FACTS

- In most moyamoya patients, stenosis occurs in the bilateral proximal portions of the anterior cerebral circulation and involves the terminal supraclinoid ICA and the proximal MCA and ACA.
- The posterior circulation is spared in most moyamoya patients. Interestingly involvement of posterior circulation is associated with poor prognosis.
- When present, stenosis of the posterior circulation typically affects the posterior communicating artery and the P2 segment of the posterior cerebral artery (PCA), while sparing the basilar artery and P1 segment of the PCA

STAGES

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Suzuki stages of moyamoya disease

Stage	Appearance
1	Bilateral ICA stenosis
2	Collateral vessels begin to form
3	Prominence of collateral vessels
4	Severe stenosis/complete occlusion of circle of Willis, moyamoya vessels narrow, extracranial collaterals begin to form
5	Prominence of extracranial collaterals
6	Complete carotid occlusion

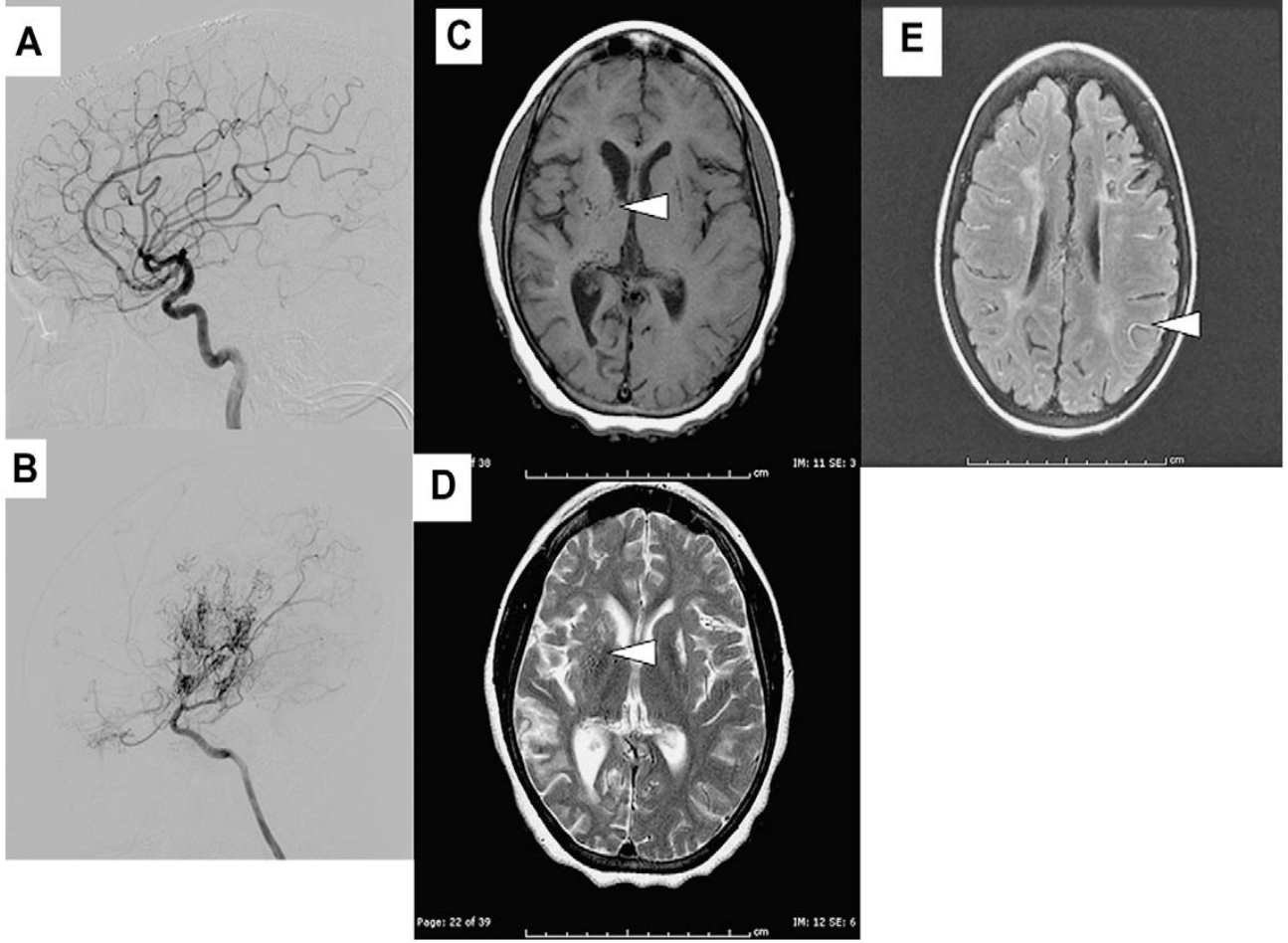
@article{Smith2010MoyamoyaEP, title={Moyamoya: epidemiology, presentation, and diagnosis.}, author={Edward R. Smith and R. Michael Scott}, journal={Neurosurgery clinics of North America}, year={2010}, volume={21 3}, pages={ 543-51 }

PRESENTATION

Symptoms at presentation	
Common	
Ischemic stroke	50–75
Transient ischemic attack (including drop attacks)	50–75
Hemorrhage (in adults)	10–40
Less common	
Seizures	
Headache	
Rare	
Choreiform movements	
Cognitive or psychiatric changes	
Associated characteristics and conditions	
Common	50–75
Angiographic findings of moyamoya without other disease	
Asian heritage	
Less common (moyamoya syndrome)	10–20
Sickle cell disease	
Neurofibromatosis type 1	
Cranial therapeutic irradiation	
Down's syndrome	
Rare (moyamoya syndrome)	<10
Congenital cardiac anomaly	
Renal-artery stenosis	

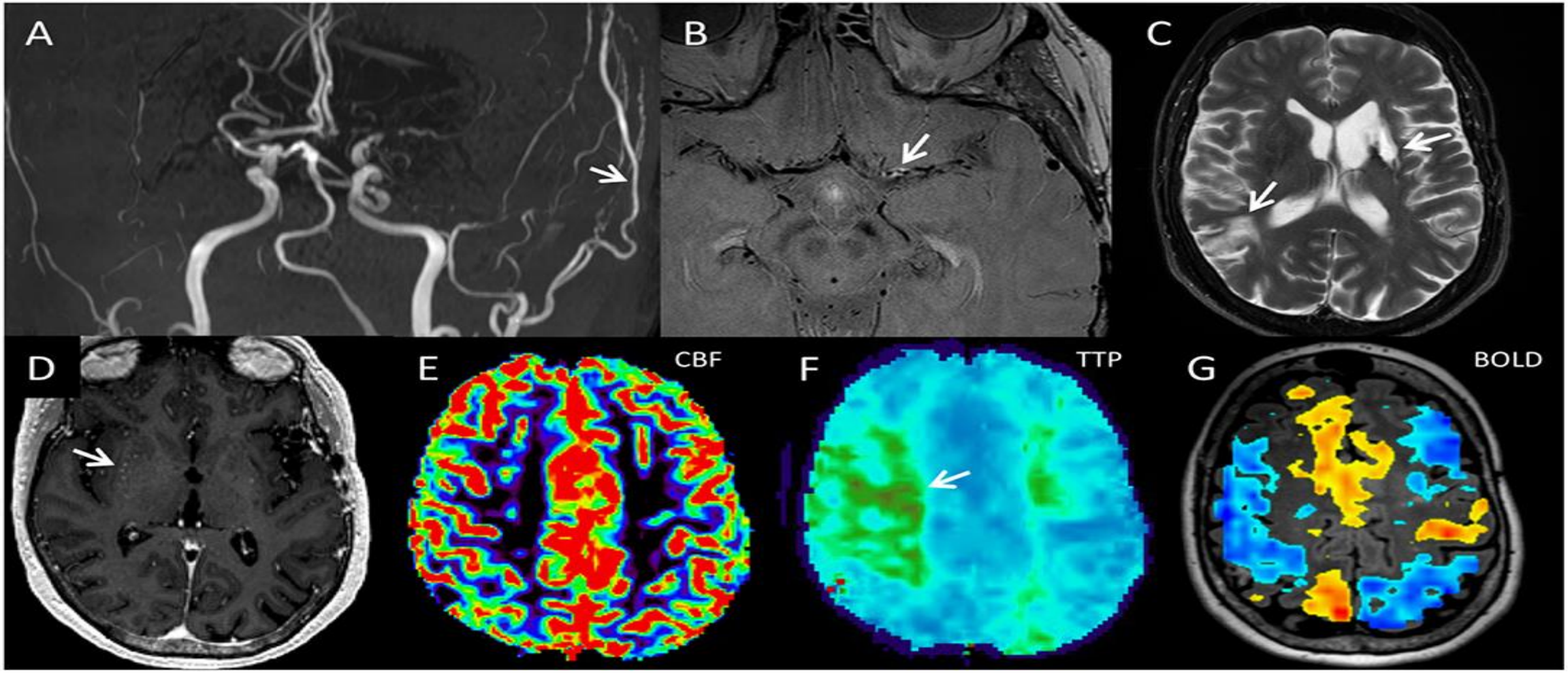
March 19, 2009, N Engl J Med 2009; 360:1226-1237,DOI: 10.1056/NEJMra0804622

DIAGNOSIS



Radiographic findings in moyamoya. Representative angiograms illustrating a normal study compared with moyamoya. (A) Normal lateral projection angiogram with injection of the internal carotid artery (ICA). (B) Suzuki grade III to IV with significant ICA narrowing and characteristic puff-of-smoke collaterals; note diminished cortical perfusion compared with (A). (C–E) Typical MRI images of moyamoya. (C) T1- and (D) T2-weighted studies reveal cortical atrophy, old infarcts, and flow void signals resulting from basal collaterals (white arrowheads). (E) FLAIR images demonstrating ivy sign consistent with bilateral ischemia (white arrowhead).

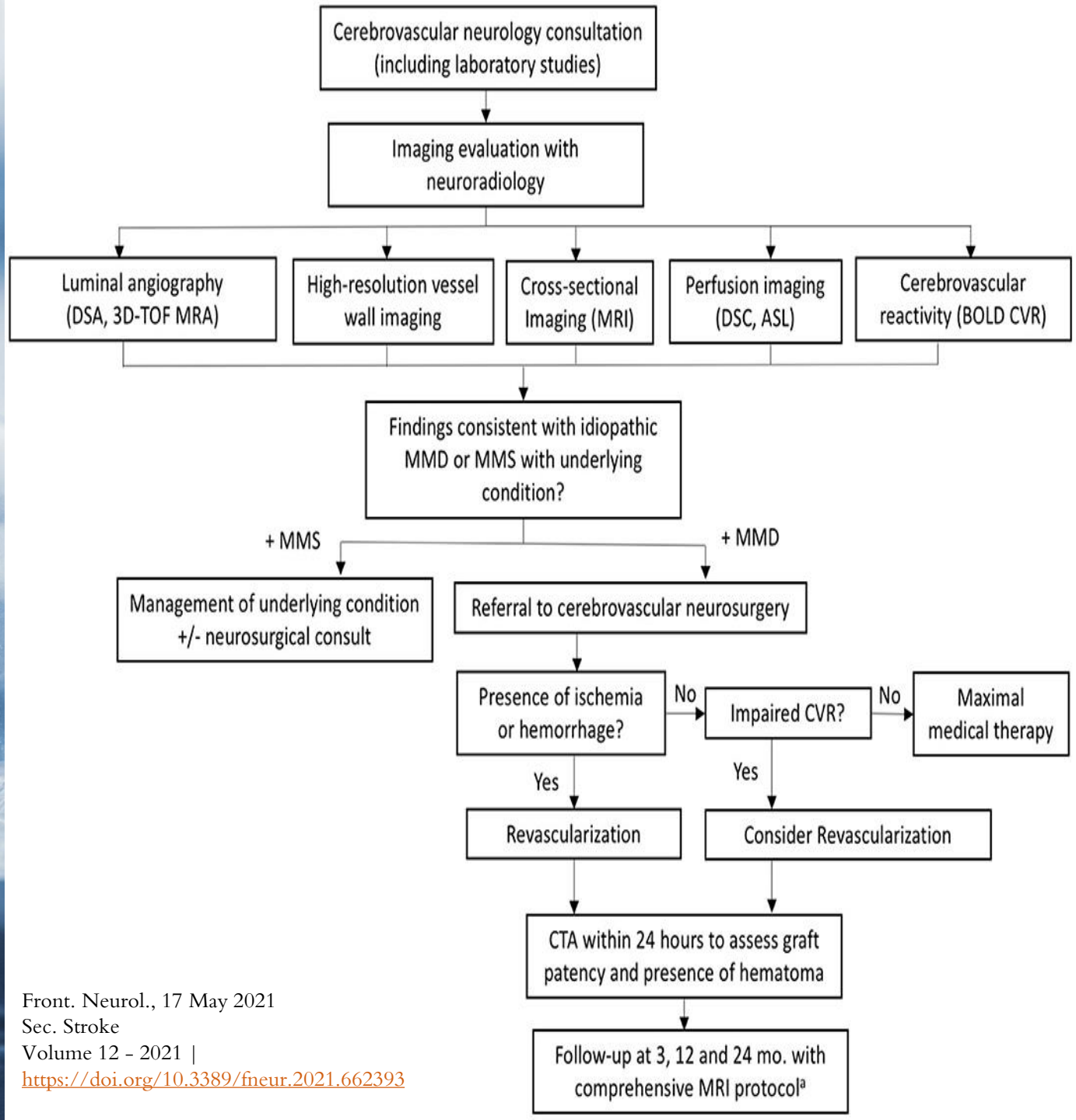
Front. Neurol., 03 September 2021
Sec. Stroke ,Volume 12 - 2021 | https://doi.org/10.3389/fneur.2021.661578



(A) demonstrates narrowing of the bilateral supraclinoid ICAs, M1 segments, and left A1 segment with decreased to absent flow. There is a patent left direct STA-MCA bypass with relatively prominent flow in the left STA (white arrow). Axial proton density vessel wall imaging demonstrates circumferential enhancement of the proximal left M1 segment (white arrow) with negative remodeling **(B)**. **(C)** Axial T2-weighted fast spin echo image demonstrates chronic infarcts in the left basal ganglia and right parietal lobe (arrows). **(D)** Axial post-gadoliniumT1 weighted MPRAGE (magnetization prepared gradient echo) image demonstrates dots of enhancement in the right basal ganglia consistent with moyamoya collateral arteries (white arrow). **(E)** Axial DSC CBF images demonstrates preserved blood flow throughout. **(F)** Axial TTP image demonstrates regional delayed transit time within the posterior right frontal lobe and left centrum semiovale region (region of green color, white arrow), consistent with delayed, but preserved flow supplied from collateral moyamoya vessels. (G) A 20 s breath-hold BOLD cerebral vascular reactivity map superimposed on a 3D axial T2 FLAIR image shows large regions of decreased reactivity in both MCA territories including areas that had normal CBF as well as increased reactivity in other regions including the ACA territories. That is, decreased reactivity could either show up on the map as non-activation, which represents decreased reactivity without steal, or blue, which represents decreased reactivity with steal.

Front. Neurol., 17 May 2021
Sec. Stroke Volume 12 – 2021,https://doi.org/10.3389/fneur.2021.662393

TREATMENT



Front. Neurol., 17 May 2021
Sec. Stroke
Volume 12 - 2021 |
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TREATMENT

REVASCULARIZATION TYPES

- Direct Revascularization: Direct cerebral revascularization consists of immediate flow augmentation to the affected hemisphere by supplying an additional source of blood flow via a direct surgical anastomosis between an extracranial donor vessel and intracranial recipient. Superior to indirect bypass in decreasing post operative cerebral events, improving hemodynamic status.
- Indirect Revascularization: Branch of the donor artery(usually superficial temporal artery) is placed on the pial surface of the brain without actual anastomosis being made. The artery will develop effective collaterals over time.

THANK YOU