

Moyamoya

- Progressive narrowing of distal ICA/proximal circle of Willis (COW) vessels with secondary collateralization
 - Collateral vessels → cloud-like "puff of smoke" (moyamoya) at angiography
- **Moyamoya disease** = primary (idiopathic) moyamoya
 - Marked East-West gradient (more common in Japan, Korea)
- Moyamoya pattern of collaterals = secondary to many etiologies

Moyamoya

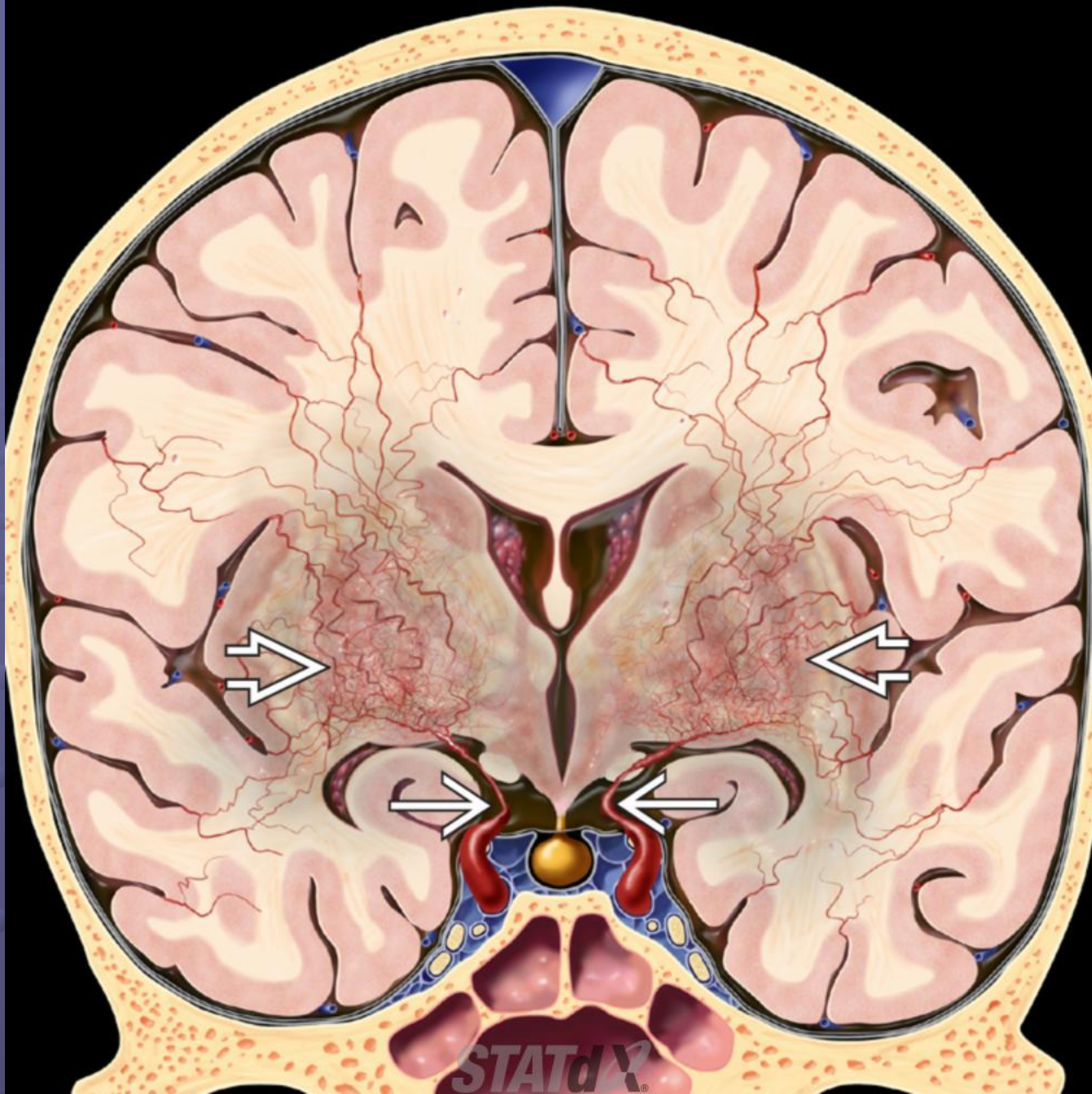
- Characterized by chronic thrombosis of cerebrovasculature, predominantly the terminal portion of the bilateral internal carotid arteries (ICAs), with resultant collateral arterial formation.
- Collateral vessels predispose patients to cerebral aneurysms as well as hemorrhagic and ischemic strokes.
- Most commonly affects females of Asian descent,
- Peak incidence in childhood and early adulthood.
- Etiology of MMD is not well understood, however there appears to be a familial basis of inheritance

Moyamoya

- MMD can be asymptomatic
- Majority initially present with stroke symptoms.
- **Angiography is diagnostic**
- In some cases the classic “puff of smoke” appearance of the collateral vessels is not identified on CT angiography or MR angiography, and only seen in conventional angiography.
- Imaging findings suggestive of MMD
 - Stenosis or occlusion of the terminal ICAs
 - Proximal anterior cerebral arteries (ACAs)
 - And/or middle cerebral arteries (MCAs), and
 - Collateral formation near the occlusive or stenotic lesions.

Moyamoya Disease

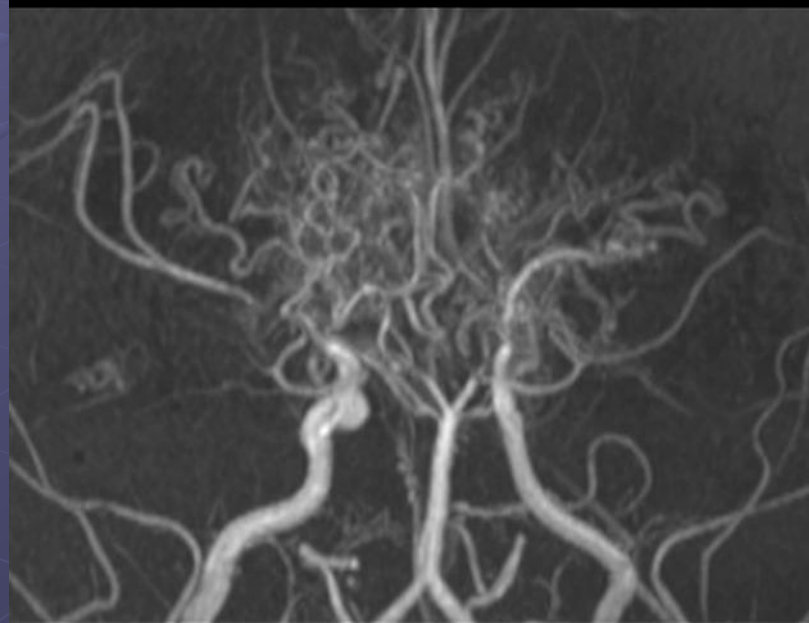
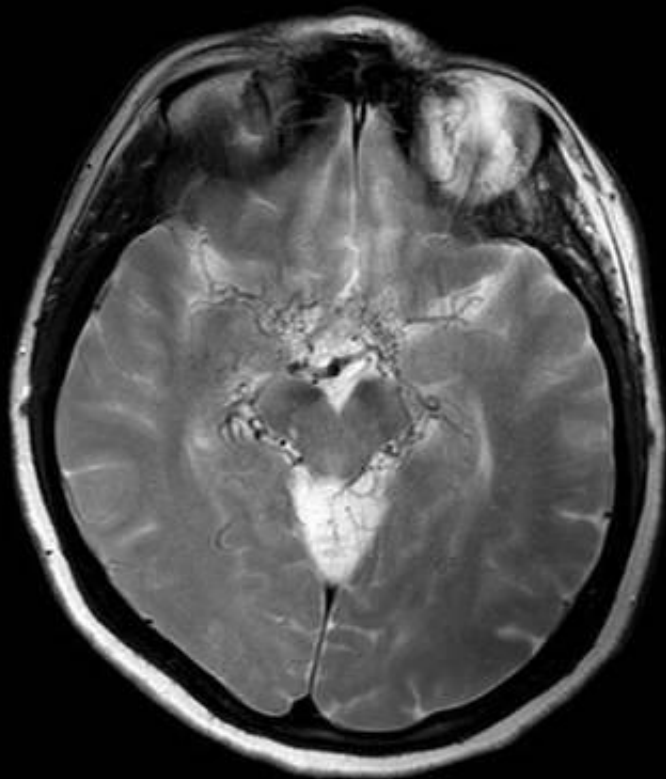
- Idiopathic
- Secondary (Moyamoya syndrome)
 - NF1
 - Radiation vasculitis
 - Radiation as a child
 - Sickle cell disease
 - Downs syndrome
 - Thyroid disease,
 - Systemic erythematosus lupus



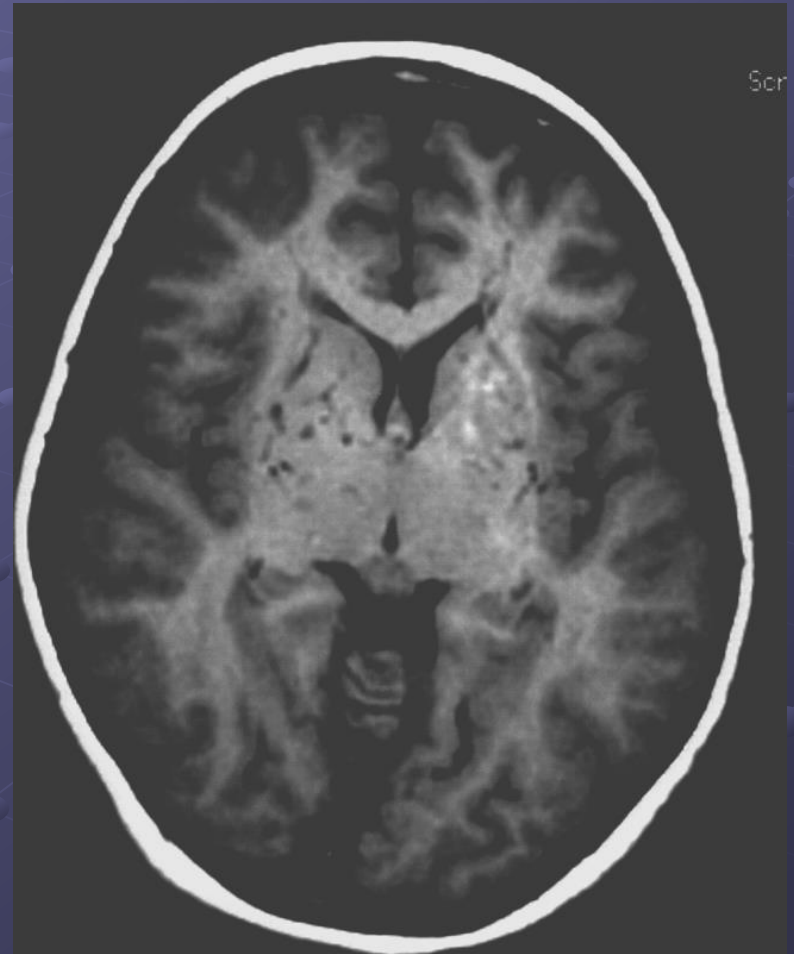
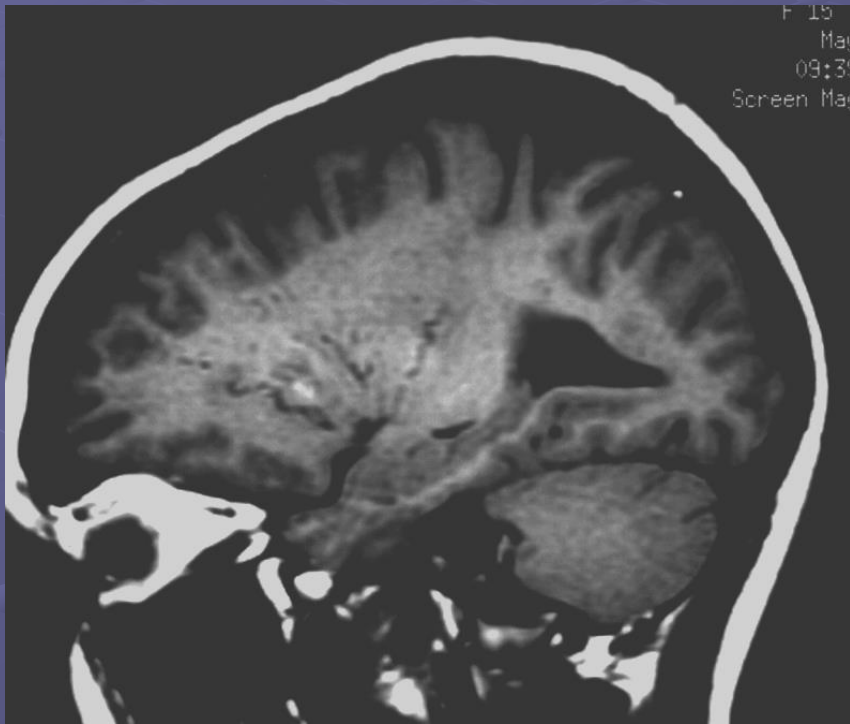
Coronal graphic shows severe tapering of both distal internal carotid arteries (white solid arrow) and strikingly enlarged lenticulostriate arteries (white open arrow) coursing through basal ganglia. This is the puff of smoke (moyamoya) pattern.



Axial T2WI MR shows curvilinear net-like filling defects (black open arrow) within the ambient (circummesencephalic) cistern corresponding to collateral moyamoya vessels. Note the asymmetric atrophy.



Moyamoya Disease



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