Clearing the Smoke: Key Findings in the Evaluation, Treatment, and Follow-Up of Moyamoya

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Moyamoya means **puff of smoke**. This angiographic appearance is caused by filling of lenticulostriate collaterals that form around the MCAs and ACAs when the distal ICAs and proximal major vessels are or narrowed or occluded. Laterality does not distinguish **Moyamoya Disease** from **Moyamoya Syndrome** as unilateral involvement can occur in either. An **underlying associated condition** indicates **Syndrome** rather than Disease.

- These include:**
- Trisomy 21
 - Atherosclerosis
 - Autoimmune Vasculitis
 - Traumatic Brain Injury
 - Neoplasm
 - Sickle Cell Disease
 - Cranial Radiation Therapy
 - Neurofibromatosis, type 1
 - Thyroid disease
 - Meningitis

<small>Modified from Fujimura & Tominaga, Neurol Med Chir (Tokyo) 2015;55:189–93.</small>	Definitive Moyamoya Disease	Probable Moyamoya Disease	Moyamoya Syndrome (a.k.a. Quasi-Moyamoya)
Laterality of Stenosis	Bilateral	Unilateral	Uni- or Bi-lateral
Underlying Condition	No	No	Yes

Occlusion of the right distal ICA (arrow). Moyamoya collaterals (arrowheads).

Pathology specimens in patients with **Moyamoya Disease** reveal a characteristic pattern of intimal thickening and medial thinning. Less is understood about the pathophysiology of **Moyamoya Syndrome**, which is presumed to vary based on the underlying cause.

The Suzuki Stages: Transition from ICA- to ECA-based intracranial blood supply

Moyamoya Disease and **Moyamoya Syndrome** differ in etiologies, but are similar in progression and treatment—all of which is **surgical**.

Direct surgical treatments anastomose an ECA branch to an ICA branch

Superficial Temporal Artery-to-MCA anastomosis

- Posterior STA limb to M4 branch.

Occipital Artery-PCA

- Rare, PCA is involved in only 30% of Moyamoya cases and still fewer require specific posterior revascularization.
- Usually supplements other surgical treatments.

There are many variations of **indirect surgical treatments**; all stimulate trans-dural or trans-pial, superficial-to-deep, ECA-fed **collateral formation**.

Multiple burr holes with periosteal flaps sutured to the dural or arachnoid surface

Post-operative cross-table lateral XR showing burr holes and pneumocephalus

Pial synangiosis

The dura is opened and the superficial temporal artery (STA) adventitia is sutured into pia mater

Pre- and 6mo post pial synangiosis in same patient. Post procedure, new collaterals stem from synangiosis site (arrowhead). STA has hypertrophied (arrow)

CT images obtained 1-4 days post right EDAMS show enlargement of intracranial temporalis flap. MR obtained on day 4 confirms that there is no intracranial hematoma (it is easy to mistake the flap for hematoma!) Flap edema is expected. Note that a previously-performed left sided EDAMS flap has atrophied—as expected.

Detecting complications:

- hyperperfusion syndrome – in patients treated with direct bypass Perfusion imaging should be used to exclude hyperperfusion syndrome, as this may otherwise have a similar appearance to infarct.
- stroke or hemorrhage – can occur periprocedurally or post-procedurally
- donor artery occlusion
- wound infection
- CSF leak