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MADISON NEUROLOGY LLC

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Neurology Care 7 days a Week. Because Your Brain Matters.

Moyamoya Disease

Patient education guide · Stroke & Vascular · Madison Neurology LLC

What is it?

Moyamoya disease is a rare, progressive narrowing of the main arteries at the base of the brain (the internal carotid arteries and their major branches). In response, the brain grows a network of tiny fragile collateral vessels that look like a 'puff of smoke' (moyamoya in Japanese) on angiography. It can cause strokes, TIAs, or brain bleeds.

How does it happen?

The exact cause is unknown; genetic factors play a role (it is more common in people of East Asian ancestry and sometimes runs in families). When it occurs with another condition such as sickle cell disease, Down syndrome, or prior radiation, it is called moyamoya syndrome.

Common symptoms

- Recurrent TIAs or ischemic strokes, sometimes triggered by crying, coughing, or hyperventilation
- Headaches
- Seizures
- Brain hemorrhage (more common in adults)
- Involuntary movements (chorea) or cognitive slowing

How is it diagnosed?

MRI and MR angiography show the narrowed arteries and collateral vessels. Catheter angiography confirms the diagnosis and stages severity; blood flow studies (such as perfusion imaging) help guide treatment.

How is it treated?

The definitive treatment is surgical revascularization — either direct bypass (connecting a scalp artery to a brain artery) or indirect procedures that encourage new vessel growth. Aspirin is often prescribed to reduce clot risk. Good hydration and avoiding activities that cause deep rapid breathing help reduce TIA triggers.

What does the future hold?

Without treatment, repeated strokes are common and cumulative. Surgical revascularization substantially reduces stroke risk and most patients stabilize well afterward. Lifelong follow-up with a cerebrovascular specialist is recommended.

Questions patients commonly ask

Q: Is moyamoya hereditary?

A: About 10-15% of cases are familial. Screening of close relatives with MRI/MRA may be offered in familial cases.

Q: Why do my symptoms happen when I cry or blow a straw?

A: Deep rapid breathing lowers blood carbon dioxide, which constricts brain arteries and can temporarily reduce flow — a classic moyamoya trigger.

Q: Can children and adults both be treated with surgery?

A: Yes. Bypass surgery is effective across ages and is the standard of care for symptomatic disease.

National support groups & resources

- **American Stroke Association** — stroke.org — 1-888-478-7653
- **American Academy of Neurology — Brain & Life patient resources** — brainandlife.org
- **National Organization for Rare Disorders** — rarediseases.org — 1-800-999-6673

(Organization contact details were current at the time of compilation; please verify online.)

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