UNDERSTANDING MOYAMOYA

A condition affecting women's health

A rare vascular condition of the brain, moyamoya affects women three times more than men. This progressive condition causes the brain's main artery — the carotid — to become very narrow, decreasing blood and oxygen flow. A new cluster of tiny, unstable blood vessels opens at the base of the brain to compensate for the decrease in blood supply. These cause an increased risk for a stroke either from blood vessel blockage or rupture that can lead to short-term or permanent brain injury.

Moyamoya means "puff of smoke" in Japanese, describing the blood vessel cluster's appearance. While the cause of moyamoya is not fully understood, there is a higher incidence in Japan, Korea and China than in Europe and North America. These cases seemingly have a genetic connection and patients are mostly young children and teens. In Japan, one in 300,000 people are diagnosed — usually females younger than 20 (source, National Organization for Rare Disorders).

The term "moyamoya syndrome" is for moyamoya due to an identifiable other condition and the term "moyamoya disease" is used when no underlying other condition that can cause these vascular changes is present. Recently, moyamoya syndrome diagnoses have increased in perimenopausal women in their 40s and 50s, leading to hormone-related research. And, the Mayo Clinic states that a moyamoya diagnosis in a family member increases likelihood by 30 to 40 times.

"Preconditions can contribute to moyamoya syndrome, and hypertension is number one," says

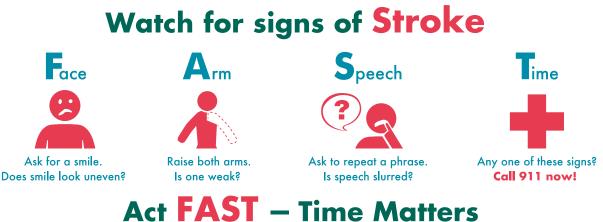
Diana Rojas-Soto, MD, Neurology, Dartmouth-Hitchcock Medical Center. "High blood pressure, cholesterol and diabetes also cause blood vessel damage, so we usually find moyamoya patients have at least one of these risk factors."

Most people don't know they have moyamoya until they experience stroke-like symptoms, such as numbness on one side of the body, trouble speaking or vision issues. Some patients might experience worsening headaches.

"Lots of people get headaches. But if they become unusually debilitating and incremental in intensity and duration, the recommendation is to be checked by a physician," says Rojas-Soto. "The only way to diagnose moyamoya is with imaging, and MRI and contrast enhanced CT scan of the arteries is the most sensitive."

Once moyamoya is diagnosed, providers can reduce the risk of stroke with medication and/or surgery. Aspirin can help reduce the risk of blood clot formation in the narrowed cluster vessels, and some patients may benefit from anti-seizure or headache-reducing medications. While there is no cure for moyamoya, revascularization surgery can increase blood flow to the brain and has stroke prevention benefits.

"It's important to know that even with rare conditions like this, there's always a way to manage risk. We must control the factors we can: diet, exercise and not smoking. Treat cholesterol and blood pressure aggressively, and seek smoking cessation counseling if you're a smoker," Rojas-Soto recommends.



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