Understanding Moyamoya Disease

What is Moyamoya Disease?

Moyamoya disease is caused by blocked arteries found at the base of the brain. The blocked arteries cause tiny blood vessels to form and look tangled. Moyamoya means "puff of smoke" in Japanese which is used to describe the tangled blood vessels.

Moyamoya disease can happen to anyone at any age. It mostly affects children ages 5 to 15 and adults ages 30 to 40.

The exact cause of Moyamoya disease is not known. It is possible that this disease runs in families (inherited or genetic disease).

What are the Symptoms?

The most common symptoms of Moyamoya disease are:

- **stroke.** A stroke happens when blood and oxygen flow to the brain is stopped or interrupted. This happens because of a ruptured or blocked blood vessel.

- **hemorrhagic stroke.** This type of stroke happens when a blood vessel in the brain ruptures and bleeds. It can be caused by a weakened area in the wall of a blood vessel or by abnormal branching of blood vessels. This is known as an arteriovenous malformation (AVM). It can also be caused by dangerously high blood pressure.

- **transient ischemic attacks (TIAs).** Transient ischemic attacks are caused by a temporary loss of blood flow to a part of the brain. These are sometimes called "mini-strokes."

Symptoms of a stroke and TIA include:

- sudden or temporary weakness

- numbness or paralysis in an arm, hand, leg or facial muscles (usually on one side of the body)

- sudden blurred double vision or dimness in your eye(s)

- sudden confusion

- trouble speaking or understanding language, or thinking

- sudden clumsiness

- loss of balance

- dizziness

- sudden severe headache.

How is Moyamoya Disease Found?

To confirm Moyamoya disease, the doctor may order:

- **CT or CAT scan (computerized axial tomography).** This test uses X-ray and a computer for an in-depth, cross-sectional look at the brain.

- **cerebral angiography.** This is a special X-ray of the blood vessels using dye.

- **MRI scan (magnetic resonance imaging).** This test uses a magnetic field to get a 3-D view of your brain. This scan can give information about the tissues and blood flow within your brain.

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- **Angiogram.** This uses X-ray to see your blood vessels. A radiologist (doctor of X-ray) inserts a tube (catheter) into an artery in your groin and threads it to the arteries in your neck and head. They inject contrast into your blood vessels to help them show up on the X-ray. This test helps rule out problems with blood vessels in the neck or brain.

- **Ultrasound.** This painless, safe test uses high-frequency sound waves to create an image of your arteries and blood flow on a computer screen. This lets your doctor see if your arteries are narrowed or damaged. Ultrasound is most often done on the carotid arteries in your neck.

- **Magnetic resonance angiogram (MRA) and CT angiography.** These are tests that use MRI or CT technology to get a 3-D view of your blood vessels.

- **Trans cranial doppler (TCD).** This is a test that measures blood flow through the major vessels in the brain.

You will receive information about what will happen during the tests, and what to expect after the tests.

How is Moyamoya Disease Treated?

Surgery is needed to restore blood flow to the brain. There are several different procedures that can open narrowed blood vessels or bypass blocked arteries. Your doctor will tell you about these procedures and you will work together to decide which type of surgery is best for you.