

# MOYAMOYA DISEASE

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**Abstract:** *Moyamoya disease is a rare, progressive blood vessel (vascular) disorder in which the carotid artery in the skull becomes blocked or narrowed, reducing blood flow to the brain. Tiny blood vessels then open up at the base of the brain in an attempt to supply the brain with blood. The word "moyamoya" means "puff of smoke" in Japanese, a term describing the appearance of cluster of tiny blood vessels. These tiny clusters of blood vessels cannot supply the necessary blood and oxygen to the brain, resulting in temporary or permanent brain injury. The condition may cause a ministroke (transient ischemic attack, or TIA), stroke, bulge or ballooning in a blood vessel (aneurysm) or bleeding in the brain. It also affects the brain function and cause cognitive and developmental delays or disability. Moyamoya disease most commonly affects children, but adults may have the condition. This disease is found all over the world, but more common in East Asian countries, especially Korea, Japan and China and in people of East Asian descent.*

**KEYWORDS;** *Moyamoya disease, stroke, disability.*

## DEFINITION

**Moyamoya disease** is a disease in which certain arteries in the brain are constricted. Blood flow is blocked by the constriction, and also by blood clots (thrombosis). A collateral circulation develops around the blocked vessels to compensate for the blockage, but the collateral vessels are small, weak, and prone to bleeding, aneurysm and thrombosis.

## CAUSES

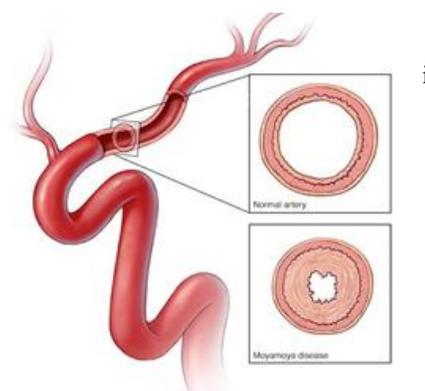
- The exact cause of moyamoya disease is **unknown**.
- Researchers believe that the higher concentration of this disease in these Asian countries strongly suggests the disease may have **genetic causes**.
- Moyamoya is also associated with certain conditions, such as **Down syndrome, sickle cell anemia, neurofibromatosis type 1 and hyperthyroidism**.

## SYMPTOMS

Moyamoya disease may occur at any age, though symptoms most commonly occur between 5 and 10 years of age in children and between 30 and 50 years of age in adults.

## First symptom of moyamoya disease include

- stroke or recurrent transient ischemic attacks (TIAs),
- bleeding in the brain (hemorrhagic stroke) from abnormal brain vessels.



**Accompanying signs and symptoms** related to reduced blood flow to the brain include:

- Headache
- Seizures

- Weakness, numbness or paralysis in face, arm or leg, typically on one side of the body
- Visual disturbances
- Difficulties with speaking or understanding others (aphasia)
- Developmental delays
- Involuntary movements
- Cognitive decline

These symptoms can be triggered by exercise, crying, coughing, straining or fever.

**SEEK IMMEDIATE MEDICAL ATTENTION** If there is any signs or symptoms of a stroke even if they seem to fluctuate or disappear. Think "**FAST**" and do the following:

- **Face.** Ask the person to smile. Does one side of the face droop?
- **Arms.** Ask the person to raise both arms. Does one arm drift downward? Or is one arm unable to raise up?
- **Speech.** Ask the person to repeat a simple phrase. Is his or her speech slurred or strange?
- **Time.** If you observe any of these signs, call ambulance immediately.

Every minute counts. The longer a stroke goes untreated, the greater the potential for brain damage and disability.

### RISK FACTORS

- **Being of Asian descent.** Moyamoya disease is found all over the world, but it's more common in East Asian countries, especially Korea, Japan and China. This may possibly be due to certain genetic factors in those populations. Higher rates of moyamoya disease have

been documented among Asians living in western countries.

- **Having a family history of moyamoya disease.** If anyone have a family member with moyamoya disease, the risk of having the condition is 30 to 40 times higher than the general population — a factor that strongly suggests a genetic component to the disease and may justify screening of family members.
- **Having a certain medical condition.** Moyamoya disease sometimes occurs in association with another disorder, including neurofibromatosis type 1, sickle cell anemia and Down syndrome, among others.
- **Being female.** Females have a slightly higher incidence of moyamoya disease.
- **Being young.** Though adults can have moyamoya disease, children younger than 15 years old are most commonly affected.

### COMPLICATIONS

- Vision problems
- Weakness (hemiparesis)
- Language disturbance (aphasia)
- Movement disorders
- Learning or developmental issues
- Seizures

### DIAGNOSIS

- **Thorough Physical Examination**
- **Magnetic resonance imaging (MRI).** An MRI uses powerful magnets and radio waves to create detailed images of brain. Physician inject a dye into a blood vessel to view the arteries and veins and highlights blood circulation (magnetic resonance angiography). Sometimes perfusion MRI is recommended( a type

of imaging that can measure the amount of blood passing through the vessels)

- **Computerized tomography (CT) scan.** A CT scan uses a series of X-rays to create a detailed image of the brain. CT angiogram ( inject a dye into a blood vessel to highlight blood flow in arteries and veins.
- **Cerebral angiography.** During a cerebral angiogram (doctor inserts a long, thin tube (catheter) into a blood vessel in groin and guides it to brain using X-ray imaging, then injects dye through the catheter into the blood vessels of brain to make them visible under X-ray imaging.
- **Transcranial Doppler ultrasound.** In a transcranial Doppler ultrasound, sound waves are used to obtain images of brain. This test is to obtain information about the blood vessels in the brain.
- **Positron emission tomography (PET) scan or single-photon emission computerized tomography (SPECT).** In these tests, injects a small amount of a safe radioactive material and places emission detectors over the brain. PET provides visual images of brain activity. SPECT measures blood flow to various regions of the brain.
- **Electroencephalography (EEG).** An EEG monitors the electrical activity in brain via a series of electrodes attached to scalp. Children with moyamoya disease often exhibit abnormalities on EEG.

## TREATMENT

The goal of treatment is to reduce symptoms, improve the blood flow and lowers the risk of serious complications such as an ischemic stroke caused by a lack of blood flow, bleeding in brain (intracerebral hemorrhage) or death.

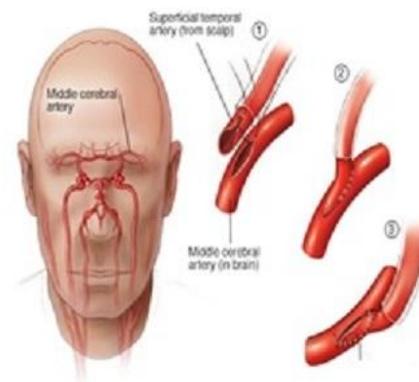
## MEDICATION

- **Blood thinners.** After diagnosis, aspirin or other blood thinner to prevent strokes.
- **Calcium channel blockers.** Also known as calcium antagonists, this type of medication may improve symptoms of headache and possibly reduce symptoms related to transient ischemic attacks.
- **Anti-seizure medications.** For treating seizure disorder.

## PROGNOSIS

Without surgery, the majority of individuals with Moyamoya disease will experience mental decline and multiple strokes because of the progressive narrowing of arteries. Without treatment, Moyamoya disease can be fatal as the result of intracerebral hemorrhage (bleeding within the brain).

## Revascularization surgery



If the symptoms become worse or if tests show evidence of low blood flow revascularization surgery is recommended. In revascularization surgery, surgeons bypass blocked arteries to help restore blood flow to the brain. Doctor may use direct or indirect revascularization procedures, or a combination of both.

- **Direct revascularization procedures.** In direct revascularization surgery, surgeons stitch (suture) the

scalp artery directly to a brain artery (superficial temporal artery to middle cerebral artery bypass surgery) to increase blood flow to the brain immediately. Direct bypass surgery may be difficult to perform in children, due to the size of the blood vessels to be attached. Direct revascularization surgery has a risk of complications, including stroke.

- **Indirect revascularization procedures.** In indirect revascularization, the goal is to increase blood flow to the brain gradually over time.

Types of indirect revascularization procedures include

- **Encephaloduroarteriosynangiosis (EDAS)**
- **Encephalomyosynangiosis (EMS)**
- **Combination of both EDAS + EMS**

## THERAPY

To address the physical and psychological effects of stroke other therapies are recommended.

- **Physical and occupational therapy** can help to attempt to regain any lost physical function caused by stroke.
- **Speech therapy** also may be recommended if needed.
- **Cognitive behavioral therapy** can help address emotional issues related to have moyamoya disease, such as how to cope with fears and uncertainties about future stroke.

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