

MOYAMOYA DISEASE

What is it?

Moyamoya disease is a rare, progressive blood vessel disease. The blood vessels which supply blood to the brain become narrowed. The narrowing limits the flow of blood to the brain and puts the child at risk for a stroke. The brain attempts to make up for the reduced blood flow by growing new blood vessels, called collaterals. In Japanese, the name "Moyamoya" means "puff of smoke". The tangled blood vessels that form cause a blockage that looks like a puff of smoke. The newly formed blood vessels work temporarily by increasing blood flow to the brain but they eventually stop working. In an attempt to supply the brain with blood, tiny blood vessels at the base of the brain open up. The condition can cause a transient ischemic attack, stroke, or bleeding in the brain.

Brain function is affected causing cognitive and developmental delays and disability. Children are most often affected by this disease. Moyamoya has no identifiable cause but it is believed to be caused by a genetic defect or traumatic injury that triggers the disease. Moyamoya is slightly more common in girls and is somewhat more common in children of Asian descent. The disease progresses over time so treatment is necessary to reduce the risk of stroke.

What are the characteristics or complications?

Similar to most diseases, symptoms will vary from child to child. Some of the most common symptoms could include:

- Seizures
- Headaches
- Vision problems
- Involuntary movements
- Muscular weakness or paralysis affecting one side of the body
- Sensory and cognitive impairment
- Speech deficits
- Risk for blood clots, strokes and transient ischemic events
- Brain hemorrhage (rare in children, more often occurs in adults)

What is the treatment?

There is no cure for Moyamoya disease. Treatment is focused on reducing the risk of stroke and treating the symptoms of the disease. No medication stops the progression of the cerebral artery narrowing and the disease continues to progress in most cases. Medications such as aspirin might be prescribed to prevent or reduce the development of small blood clots forming in the narrowed vessels. Anti-seizure medication might be prescribed if indicated. Surgery is the only treatment that can reduce the risk of a life threatening stroke. Surgery can restore blood flow to the brain. A team of medical and developmental specialists are key to the best possible outcome.



Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

Supporting students with Moyamoya disease in the school setting requires the educators and parents/guardians to work as a team. A well coordinated plan promotes success for the student in the educational setting. Some accommodations to consider for an 504/IEP might include:

- Early intervention assessment for young children
- PT/OT/SLP/Vision specialist consults
- Emotional support
- Education about seizure management for staff
- Allow rest breaks as fatigue is common side effect of seizures and medication
- Provide a private area to rest or recover after a seizure
- Assess need for a protective helmet
- Consider assistive technology
- Extended time for testing
- Accommodations if student is having surgery
- Revisiting work if student has seizures
- Extra time for assignments as needed
- Offer emotional support
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including all affected systems and symptoms
- Communicate with school staff, parents, and provider any changes or concerns about the disease
- Current medication list for home and school
- Documentation/log of seizures including type of seizure, description of, typical length, characteristics, triggers, warning signs, how often seizures occur, and student's behavior following a seizure
- Orders for emergency medications, when to administer, dose, route
- Orders from provider for any physical activity restrictions to prevent overexertion
- PT/OT/SLP/Vision services
- Plan for monitoring neurological changes, headaches, anxiety
- Orders for fluid intake goals for student to stay hydrated
- Provide rest area if needed
- Emergency Care Plan (ECP) related to seizures and other medical needs in the school setting and staff training as appropriate for each

Resources & Manuals

Boston Children's Hospital Moyamoya

<http://www.childrenshospital.org/conditions-and-treatments/conditions/m/moyamoya-disease>

National Organization for Rare Disorders (NORD)

<https://rarediseases.org/rare-diseases/moyamoya-disease/>

NIH Genetic and Rare Disease Information Center

<https://rarediseases.info.nih.gov/diseases/7064/moyamoya-disease>

NIH National Institute of Neurological Disorders and Stroke-Moyamoya Disease Information page

<https://www.ninds.nih.gov/disorders/all-disorders/Moyamoya-disease-information-page>