

MOYAMOYA DISEASE

Background

Moyamoya disease (MMD) is a progressive vascular disease causing blood vessels (specifically the internal carotid arteries) in the brain to narrow or close. To compensate, the brain creates pathways of tiny new blood vessel networks. The term “moyamoya” means puff of smoke in Japanese to describe the look of the tangled, fragile vessels that form as emergency backup. The disruption of oxygen rich blood flow increases the risk of a transient ischemic attack (TIA) and stroke. Long-term neurological conditions can include problems with learning, language, behavior and seizures.

The symptoms of MMD are progressive and can develop over the course of months to years. Children are more commonly affected than adults; with a peak age of diagnosis between 5-10 years and a 2:1 female-to-male ratio. MMD has no identifiable cause, but research supports a genetic defect or traumatic injury trigger. MMD may also be diagnosed in children with other pre-existing conditions that cause vascular damage to blood vessels in the brain, including sickle cell disease and Down syndrome.

Children with MMD will present with symptoms caused by reduced oxygen rich blood supply. Most strokes in children present suddenly with symptoms including:

- Weakness or numbness on one side of the body
- Balance issues
- Dizziness
- Difficulty speaking
- Vision changes

There is no cure for MMD, but treatment is focused on preventing vascular events and managing symptoms of the disease. Medication alone will not stop the progression of the narrowed blood vessels, but Aspirin may be prescribed to prevent or reduce the development of small blood clots. Anti-seizure medication may also be indicated. Surgery to restore blood flow in the affected area of the brain is the only long-term treatment for MMD to reduce the subsequent risks of stroke and protect the intellectual abilities of the individual. The overall prognosis for individuals with MMD depends on age of onset and how rapidly vascular blockage occurs.



Top Takeaways for School Considerations

MMD is a blood flow disorder in the brain and a major cause of stroke in childhood. Staff should be trained in the signs and symptoms of potentially life-threatening medical emergencies.

MMD is three times more common in people with Down syndrome.

Neurological deficits will be variable and dependent upon the specific area of the brain affected.

The student may experience cognitive impairments especially in executive functioning including planning and organization.

Mathematics has been identified as an area of specific learning need.

Kennedy Krieger Institute's Specialized Health Needs Interagency Collaboration

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



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Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of risk for ineffective cerebral tissue perfusion, impaired physical mobility, risk for disturbed sensory perception and impaired thought process
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Nutrition interventions and equipment needs (consider brand/size of feeding tube, tube replacement, water flushes, fluid intake goal and supplements); note school district policy on tube replacement and consider keeping backup feeding tube kit at school if applicable
- Activity, positioning, transferring (consider precautions and/or restrictions)
- Equipment troubleshooting (consider equipment/device user manual, battery, charger)
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Has the school staff been trained to implement the student-specific emergency plan?
2. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
3. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule?
4. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
5. Would schedule flexibility support the student?
6. Does the student need additional adult support to access the academic curriculum in the least restrictive environment?
7. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?
8. Does the classroom environment support the student's needs and/or equipment (e.g., desk/seating options, maneuverability space, electrical outlets, flash pass for bathroom or nurse)?

Resources

Kennedy Krieger Institute: Neurology and Neurogenetics Clinics
kennedykrieger.org

Child Neurology Foundation
childneurologyfoundation.org

American Stroke Association
stroke.org

Children's Hemiplegia and Stroke Association
chasa.org



Scan QR code or visit
KennedyKrieger.org/HealthInformation for more information.