NEUROFIBROMATOSIS

What is it?

Neurofibromatosis (NF) is a genetic neurological disorder affecting the brain, spinal cord, nerves and skin. Genetic mutations cause the body to grow benign tumors, called neurofibromas, along the body's nerves or under the skin. Skin and skeletal abnormalities are most visible but the tumors can also grow inside the body; pressing on organs and nerves affecting their function. There are three types of Neurofibromas: NF1, NF2 and NF3.

NF1 is caused by mutations in the gene that controls production of a protein called neurofibromin. This gene is believed to function as a tumor suppressor. The characteristics of this genetic condition is distinctive skin features that appear in childhood, especially during puberty. Flat, brown spotting on the skin (café-au-lait spots, totaling more than 6) and soft bumps on or under the skin are most commonly seen. Small spots in the armpits and groin are also present. Other medical concerns include learning disabilities, ADHD, bone deformity, short stature, and larger head size.

NF2 results from mutations in a different tumor-suppressing gene. NF2 is far less common than NF1. Characteristics of NF2 are benign tumors on the nerves that transmit balance and hearing impulses from the ear to the brain. The disruption of the impulses can cause ringing in the ear(s) and headaches in early childhood, adolescence or adulthood. Some children with NF2 also have other abnormalities of the eye including visual impairments, clouding of the lenses, and increase tumor growth lining the brain, and spinal cord.

NF3 is caused by various mutations on chromosome 22 but presents as a disease during adulthood and is characterized by chronic pain.

What are the complications?

Treatment is aimed at removal of tumors and treating the underlying medical complications. These complications can include: seizures, high blood pressure, pain, scoliosis, early or delayed puberty, vision problems, hearing and/or speech impairment, numbness, or cancer.

What are the effects on learning ?

About 50 to 60% of children with NF has some type of learning disability that affects executive function, processing, reading, and math. Many of these children also have difficulty with fine motor and gross motor activities because of the tumors. Developing a 504/IEP is something that the team should consider as NF can affect the following:

- Concentration
- Impulsivity, restlessness, fidgety
- Poor visual-spatial skills
- Affected gross and fine motor skills, poor balance, poor handwriting
- Inconsistent memory, requires repeated verbal information
- Difficulties with planning, problem solving, abstract formation, and reasoning
- Expressive and receptive language deficits
- Poor organization, poor time management
- Impaired social skills, fast talker

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 this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT/SLP evaluations
- Audiology/impaired vision consultations
- Modified or flexible school day
- Plan for absences and make-up work
- Assistive technology
- Executive function supports
- Preferential seating in classroom for hearing/vision needs
- Repeated instructions, check for understanding
- Offer short, concise, limited instruction
- Supplement instruction with visual aid
- Use of multi modalities for teaching material
- Establish a home and school communication method
- Additional support in cafeteria if needed
- Support with equipment
- Supervision as may not recognize or anticipate danger
- Role play to demonstrate behavior, facial expressions, body language
- Recognition of strengths
- Emotional support
- Peer awareness and education about disease
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including classification and characteristics
- Current medication list including PRN pain medications
- Surgical history with plan for communicating updates
- Baseline cardiac and renal history, blood pressure parameters
- Contraindications for positioning, mobility
- CPR considerations related to tumors and/or surgical interventions
- Supervision in halls, stairs
- Restrictions regarding physical activities
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

Johns Hopkins Medicine: Comprehensive Neurofibromatosis (NF) Center
https://www.hopkinsmedicine.org/neurology_neurosurgery/centers_clinics/neurofibromatosis/

Washington University– Neurofibromatosis Center
https://nfcenter.wustl.edu/

St. Louis Children's: A guide for parents of children with neurofibromatosis-1
https://nfnetwork.org/data/uploads/nf1-educational-materials/addressing-executive-function-nf1.pdf

NF Network: Understanding neurofibromatosis
https://nfnetwork.org/data/uploads/nf1-educational-materials/understanding-nf-3rd-edition.pdf