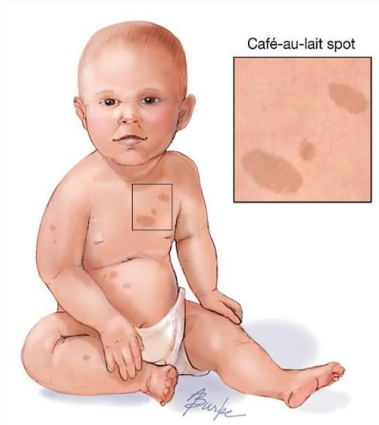
Kennedy Krieger Institute

Maryland State Department of Education
Preparing World Class Students

Factsheet: Neurofibromatosis

SHNIC
Specialized Health Needs
Interagency Collaboration

What is it?



Neurofibromatosis (NF) is progressive genetic disorder of the nervous system. Genetic mutations cause the body to produce the growth of benign tumors, called neurofibromas, on nerve tissue. Skin and skeletal abnormalities are most often seen but tumors can also grow inside the body, pressing on nerves and vital organs affecting their function. This disease, with its many forms, affects all races and sexes equally and is the most common genetic disorder in the US.

Genetic mutations can either be inherited or occur through spontaneous mutation. Chromosome 17 (NF 1) and chromosome 22 (NF 2) are each responsible for producing proteins that support cell growth. When NF affects these chromosomes, cell growth is uncontrolled.

NF 1 is characterized by 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 often seen. Small spots, like freckles, are also observed in the armpits or groin. Other medical concerns of NF 1 include learning disability, bone deformity, large head size and short stature. There is also a high co-morbidity between NF 1 and ADHD as NF 1 children are 3 times more likely to have ADHD than their siblings.

NF 2 is far less common than NF 1. NF 2 is characterized by benign tumors that develop in the ear. The placement of the tumor disrupts sound and balance cues from the inner ear to the brain. Signs and symptoms are usually seen in late teens and early adulthood and include hearing loss, ringing in ears and poor balance. In some cases, these tumors can also further develop on cranial, optic, spinal and peripheral nerves.

Treatment is aimed at removal of tumors and treating the underlying medical complications. These complications can include:

- Seizures
- High blood pressure
- Scoliosis
- Hearing /Speech impairment
- Early or delayed puberty
- Pain
- Vision problems and/or blindness
- Numbness
- Cancer

Effects on learning

The relationship of why student's with NF struggle with school is not clear. Regardless, it is important for the school staff to understand and prepare for a child's potential behavior in the classroom. Without support and structure, a child with NF can be falsely labeled as disruptive.

- Difficulty concentrating
- Affected gross and fine motor skills
- Poor handwriting
- Inconsistent memory
- Require repeated verbal information
- Restless and fidgety
- Poor visual-spatial skills
- Problems with executive function that include planning, problem solving, abstract formation, and reasoning
- Poor control of own movement
- Poor balance
- Clumsy, bumping into things
- Impulsive
- Fast talker
- Impaired social skills
- Poor time management
- Poor organization
- Expressive and receptive language deficits

Suggested school accommodations

- Invite the class to learn about NF per student and family's request
- Position in front of classroom with minimal distractions
- Position in classroom for volume considerations
- Home to school diary for communication
- Colored tape to designate personal work space on all desks
- Stranger danger tools
- Tablet or computer program for alternative recording methods
- Offer short, concise, limited instruction
- Planner with daily schedule
- Supplement instruction with visual aid
- Block off parts of page not in use
- Additional support in cafeteria
- Support with equipment
- Use of "tangle toy" for fidgety behavior
- Offer rules, checklists, schedules
- Allow extra time for work
- Reiterate starting point to help with sequencing
- Supervision as may not recognize or anticipate danger
- Role play to demonstrate behavior, facial expressions, body language
- Visual timer for time management
- Recognition of strengths
- Repeated instructions, ask for feedback of understanding
- Use ruler or notecard for visual tracking

SHNIC school nurses information:

Specific health issues for individual health care plans

- Diagnosis including classification and characteristics
- Current medication list including PRN pain medications
- Surgical history with plan for updates
- Seizure action plan, if applicable
- Baseline cardiac history, blood pressure parameters
- Baseline renal history
- Contraindications for positioning, mobility
- CPR considerations related to tumors and/or surgical interventions
- Supervision in halls, stairs

Resources & Manuals

Neurofibromatosis Network
http://www.nfnetwork.org/understanding-nf/what-is-nf/nf-2

Addressing academic concerns:
A guide for parents of children with neurofibromatosis 1
https://nfcenter.wustl.edu/wp-content/uploads/2010/10/SLC6848_AcademicConcernsNeurofibromatosisR4.pdf

Addressing executive function:
A guide for parents of children with neurofibromatosis 1
https://nfcenter.wustl.edu/wp-content/uploads/2011/06/Executive-Function-Brochure.pdf

The Neuro Foundation– NF1 information sheets
http://www.nfauk.org/what-is-neurofibromatosis/nf-type-1/nf1-info-sheets/