Neurofibromatosis

Symptoms or Behaviors

- Skinfold freckling between the ages of 3 and 5
- Optic pathway gliomas, peak incidence between 4 and 6 years of age
- Two or more Lisch nodules (half have by 5 years)
- High blood pressure
- Migraine headaches
- Nausea and abdominal pain
- Plexiform neurofibromas (grown under skin surface and deeper areas of the body)
- Learning difficulties
- Attention deficits
- Dermal neurofibromas
- Scoliosis
- Seizures are a known complication of NF
- Short stature as a group
- Early or late puberty
- Itching related to skin neurofibromas (heat makes it worse)
- Enlarged head

Minnesota Low Incidence Project

About the Disorder

Neurofibromatosis (NF) is a genetic disorder. There are two forms of NF, Type I and II. They are caused by different genes which are located on different chromosomes. The gene may be inherited from a family member or if there is no family history of NF as a result of gene change called spontaneous mutation. Both forms of NF are characterized by the growth of benign tumors called neurofibromas. Neurofibromas are benign tumors composed of nerve, blood vessel and fibrous tissue. These tumors can grow anywhere in the body where there are nerve cells. This can be just under the skin, as well as deeper within the body, spinal cord and/or brain. About 1:4,000 people in the United States are affected with NF 1, and 1:35,000 are affected with NF II. Males and females are affected equally and it is found in all races. There is no predictable course and no certain prognosis.

The most common form is NF 1, also known as von Recklinghasen NF or Peripheral NF. In diagnosing NF 1, doctors look for the following: six or more café-au-lait spots (brown spots on the skin that increase in size over time), two or more neurofibromas of any type, freckling in the armpit or groin area, optic pathway tumors, two or more Lisch nodules (flecks of pigment on the iris), bone abnormalities, or a family member with NF 1. In NF 1, tumors commonly grow on the skin or in the nerve to the eye. Tumors that affect the eye are called optic gliomas and if they grow large enough may cause problems with vision, including blindness. About 50% of the individuals with NF I have some type of learning disability.

NF 2 is also called Bilateral Acoustic Neurofibromatosis or Central Bilateral Acoustic NF. NF 2 is characterized by multiple tumors on the nerves to the ears called acoustic neuromas. If the acoustic neuromas become large enough, they can lead to deafness. Ringing in the ear, problems with balance, and hearing loss usually occur in the teenage years or early twenties. Individuals with NF 2 are at high risk for developing brain tumors.

Treatment for both NF 1 and NF 2 is aimed at controlling symptoms. In NF 1 surgery may help with bone malformations or to remove painful or disfiguring tumors. The tumors may grow back in even greater numbers. Tumors become malignant in 3-5% of the cases and treatment then includes surgery, radiation or chemotherapy. For NF 2, partial surgical removal of tumors and radiation may be used.

Neurofibromatosis can be stressful for many affected individuals. Some experience social isolation and loneliness. The general public's reaction to disfigurement and the unfounded fear that NF is contagious can result in unpleasant situations. Uncertainty about future complications poses additional stress.

Developed with Minnesota Low Incidence funds, Region 10 Physical Health Disabilities Network, and School Nurse Organization of Minnesota (SNOM).

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Educational Implications

The degree that a student is affected by Neurofibromatosis will make a difference on what is needed in the educational setting. Vision and hearing problems due to tumors, discomfort from bone deformities and possible seizure disorders would all impact a child's education. Fifty per cent of the children with NF 1 have some type of learning disability which may involve language development, visual-motor, visual-spatial, reading and organizational/attending difficulties. Studies have indicated a greater prevalence of cognitive deficits in children with NF 1 than the general population. There is no predictable course and because of that students may experience symptoms appearing at different ages during their school career. Staff should be alert for signs of anxiety and depression.

Instructional Strategies and Classroom Accommodations

- Twice a year screening of vision and hearing
- If vision/hearing problems are present, accommodations specific to those needs must be addressed such as preferential seating and specialized equipment
- Screen for scoliosis; monitor physical comfort
- If seizures are present, the school nurse in cooperation with general and special education staff will develop plans/accommodations that meet student's specific needs
- Assist the child to cope with disfigurements from facial tumors and bone deformities
- Educate the class about Neurofibromatosis (obtain approval from parents and student)
- Support good social relationships and self-esteem; observe for signs of anxiety or depression
- Address attentional and organizational difficulties (preferential seating, frequent checks to make sure assignments are recorded, completed and filed, get the child's attention before giving clear and simple directions, give frequent positive feedback, etc.)
- Assess to determine if or what type of learning difficulties are present; accommodations need to be made based on specific areas of difficulty, similar to LD.
- Physical education program may need to be modified.

Resources

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Achieving in Spite of… Learning Disabilities
A Booklet on Learning Disabilities published by National Neurofibromatosis Foundation, Inc.

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