NF1 • GUIDE FOR EDUCATORS

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CHILDREN'S TUMOR FOUNDATION
ENDING NF THROUGH RESEARCH

NEUROFIBROMATOSIS TYPE 1

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INTRODUCTION

A diagnosis in a child of neurofibromatosis type 1 (NF1) raises many important issues for care, whether in the doctor's office, at home, or in school. Children with NF1 can enjoy good health and academic success, but many have special needs—especially in a school environment. Proper attention to these needs can dramatically increase the chances that children with NF1 will do well, both in school and in later life.

Classroom and special education teachers, school psychologists and counselors, occupational therapists, speech pathologists, and school nurses can make a significant difference in the lives of children with NF1 by gaining knowledge about the disorder, and providing early intervention when and if needed.

We thank you for your concern and interest in reading this brochure. This is a time of exciting progress in our understanding of NF1, and we welcome you to the care team.

ABOUT NEUROFIBROMATOSIS

Neurofibromatosis is a term that encompasses three distinct disorders: neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2), and schwannomatosis. All are genetic conditions that cause tumors to form on nerves. This brochure deals exclusively with NF1, which is, in addition to tumors, commonly associated with learning disabilities. In contrast, NF2—a largely adult-onset disorder that can cause loss of hearing and balance—only rarely shows effects in children, and is usually not associated with cognitive disabilities. Schwannomatosis typically causes symptoms of severe chronic pain in adulthood.

NF1 strikes 1 in every 3,000 births and can lead to a wide range of medical problems. The features of NF1 vary greatly from one person to the next. Some children are quite severely affected, while most have considerably milder cases. Although many children with NF1 generally enjoy good health, the disorder can lead to blindness, disfigurement, bone abnormalities, learning disabilities, disabling pain, and cancer.

Research indicates that approximately 50-60% of all children with NF1 have some form of learning disability. Many will require special education services to address learning difficulties, speech problems, motor deficits, or psychosocial problems. There is also a higher incidence of attention deficit disorder among children with NF1 compared to the general population. In addition to the potential for cognitive difficulties, some children may have visible manifestations of NF1 that draw attention and cause added stress for them in social situations.

WHAT CAUSES NF1?

NF1 is caused by a change, or mutation, in the structure of a gene. Because of this, a child can inherit NF1 from a parent who has the disorder. About half of the time, however, a child with NF1 is the only person in the family who has the disorder. In such instances, the NF1 gene change occurred as the result of "spontaneous mutation"—a random error in the process of copying genetic information.

NF1 is not the consequence of drug, alcohol, or X-ray exposure, or any other factor under the control of the child's parents. NF1 is not contagious. Contact between an affected child and an unaffected child can not transmit the condition.

DIAGNOSING NF1

NF1 is generally diagnosed by a geneticist or pediatric neurologist using defined clinical criteria. However, school nurses and special education teachers can play an important role in identifying children who may have NF1, and who should be referred to a physician knowledgeable in NF for medical evaluation and assessment.

A diagnosis of NF1 is often made by the presence of six or more pigmented spots on the skin (called café-au-lait spots), in combination with other hallmark features of the disorder. Genetic testing is available to confirm most cases of asymptomatic NF1 in children (where there are no visible or clinical features).

A separate brochure ("Newly Diagnosed with NF1") is available from the Children's Tumor Foundation at www.ctf.org; it provides more specific information on the diagnosis and management of NF1.

MEDICAL COMPLICATIONS OF NF1

Generally, it is the job of medical professionals, not school personnel, to monitor the health of children with NF1. However, it is also helpful for educators to be aware of some of the potential complications and manifestations of NF1 that can affect children.

Tumors: Because NF1 is associated with tumor formation, one of the most feared complications is cancer. Fortunately, the most common tumors associated with NF1 (called neurofibromas) are not typically malignant. They may, however, require surgery to allay the chance that serious complications could develop. It is important to note that any sudden growth or pain in a neurofibroma can be a sign of malignancy onset and should be brought to the attention of a medical specialist.

Some children with NF1 develop brain tumors. The most common of these are optic gliomas, which involve the optic nerve controlling vision. Typically, these are asymptomatic and do not require treatment, although they can cause problems with vision or stimulate the early onset of puberty. In such cases, effective treatments are available. Other types of brain tumors are rare in children with NF1, and may be preceded by headaches, seizures, or changes in behavior. Any or all of these should be medically evaluated as soon as possible after onset. Most such changes are not indications of a brain tumor, but medical evaluation is important.

Migraines: Some children with NF1 have a migraine syndrome that can include a headache, abdominal pain, nausea, vomiting, malaise, fatigue, or dizziness. The headache may be a minor feature, or may not be present at all. These children may miss many days of school or be sent frequently to the school nurse. A medical evaluation should be initiated if these symptoms are present, in order to rule out, for example, that there is a brain tumor. The good news is that children with NF1 can respond dramatically to appropriate medications for prevention and treatment of migraines.

Scoliosis: Children with NF1 are at higher-than-average risk for scoliosis (abnormal curvature of the spine) which can appear at a much younger age than is typical in the general population. This can be detected by regular screening, but early management is critical to preventing serious complications.

Bone defects: Abnormal bone development may occur in some individuals with NF1. Most bone defects of NF1 will be evident at birth or shortly thereafter (some, such as vertebral defects, can occur later). They can occur in almost any bone, but are seen most often in the skull and limbs.

High blood pressure: Children with NF1 are also at a higher-than-average risk for high blood pressure, and again, this can appear at a much younger age than in the general population. This can be detected by regular screening and appropriately managed.

Speech and motor deficits: These are often associated with NF1. Children exhibiting these problems tend to benefit greatly from early intervention through speech and occupational therapy.

PHYSICAL ACTIVITIES FOR CHILDREN WITH NF1

In general, children with NF1 are not unusually fragile and do not require special protection. They are capable of participation in a full range of normal activities. The only exception is for those who have specific complications (for example those relating to bone defects, scoliosis, or tumors) that may place them at risk for injury. In these cases, the child's physician will point out any restrictions on physical activity.

COGNITIVE & BEHAVIORAL CONSEQUENCES OF NF1

Ensuring early recognition of cognitive or behavioral problems in children with NF1 is a critical piece of a family's partnership with school personnel. It is important to remember that at least half of all children with NF1 have some degree of cognitive or behavioral difficulties.

The possibility of such problems should be kept in mind for any child with NF1. Many physicians suggest that all children with NF1 be formally evaluated for cognitive function—either through diagnostic testing in the schools or a neuropsychological examination by a medical specialist.

It is believed that cognitive and behavioral problems may be caused by changes in the structure and/or function of the brain due to NF1. No specific profile of cognitive or behavioral impairment seems to be unique to NF1. Rather, problems overlap with those seen in the general population—and children with NF1 respond to the same interventions that are used for children with cognitive or behavioral impairment who do not have NF1.

However, it is equally critical to recognize that roughly half of all children with NF1 have no cognitive or behavioral complications. There can be a danger of "over-diagnosing" or "over-analyzing" the condition and assuming that such problems will be present in a child with NF1. For this and many other reasons, it is important for educators and parents to work together in evaluating whether a problem exists.

INTELLIGENCE & NF1

As in the general population, intelligence in children with NF1 spans the entire range, from below average to above average. Studies have shown a tendency for IQ scores in children with NF1 to be "shifted to the left" (i.e. lowered) to the mid-80s. However, any one individual may score below or above this.

There does not seem to be a consistent discrepancy between verbal and performance IQ in NF1. Severe problems (classifiable as intellectual disability) are rare and these are generally obvious in the first few years of life.

NF1-ASSOCIATED LEARNING DISABILITIES

A learning disability is defined as a problem with a specific cognitive function that is necessary for learning in individuals with average- or even above-average intelligence. As in the general population, there is a wide range in the character and severity of learning disabilities that can be seen in children with NF1—including both verbal and non-verbal disabilities.

The learning disabilities seen in NF1 can include dysfunction in visual or auditory perception (not to be confused with visual or auditory acuity); information integration (such as sequencing, abstraction, or organization); memory; language; gross motor, fine motor, or oral motor skills (causing clumsiness, poor handwriting, or poor articulation); and social skills or behavior (including misperception of social cues, attention deficit, or hyperactivity). They may present problems in reading, spelling, math, spatial ability, neatness, test taking, speech, ability to make friends, or many other functions.

Children with NF1-associated learning disabilities can benefit greatly from evaluating areas of strength and weakness, and from an Individualized Education Program (IEP) tailored to the unique needs of the child. With early and appropriate intervention, these children can succeed in school.

A separate brochure ("NF1: About Learning Disabilities") is available from the Children's Tumor Foundation at www.ctf.org; it outlines specific learning disabilities and practical suggestions for classroom modifications that may be helpful.

BEHAVIORAL PROBLEMS

Behavioral problems associated with NF1 may include attention deficit/hyperactivity disorder (ADHD), as occurs in the general population. Although this problem is a direct result of this complex genetic condition, children with NF1 who have behavioral problems respond to the same interventions as any child: individualized attention, behavioral modification, and, in some cases, medication. Medication should never be used as a sole approach, but it can be helpful to some children with NF1.

PSYCHOSOCIAL CONCERNS

Children with learning disorders or other disabilities may find themselves bullied by fellow students. The rare disfiguring complications of NF1 in childhood are often subject to questions or teasing from classmates, requiring a high level of sensitivity to both the child with NF1 and the other students. This behavior often is a result of ignorance among their peer group, and can be counteracted by providing accurate, age-appropriate information.

Children with NF1 often suffer from social isolation, poor self-esteem, anxiety, or depression. For those coping with the combined burden of medical, learning, and social issues associated with NF1, the typical school day can be psychologically exhausting.

MAINSTREAM CLASSES OR SPECIAL EDUCATION?

Many children with NF1 benefit from special education services. If learning disabilities are present, there is no reason to wait to see if children will "outgrow" their deficits. Early intervention usually is available through the public school system as early as age three, and from birth in some school districts, and should be sought out as early as possible (see the "Finding Help" section).

Children with learning disabilities resulting from NF1 may learn academic skills at a slower rate than their peers; however research informs us that they will, in time, learn the necessary material—just as their classmates do. They often struggle with rote foundation skills despite having adequate comprehension abilities. Educators and parents should maintain high expectations for the child, but not demand mastery to the point of frustration. Children enrolled in special education classrooms should be evaluated regularly to assess whether special services continue to be needed, or if the child is increasingly capable of succeeding in mainstream education classes.

FINDING HELP

Some children with learning deficits may not meet eligibility criteria for receiving special services based solely on evaluation within the school system. However, children with NF1 are eligible for special education under the category of "Other Health Impairment" as verified by a physician. Speech and occupational therapy may be provided as "Related Services" if needed. Assistive technology services, such as access to laptop computers if handwriting is a problem, are also available and helpful for many children.

COMMUNICATION AMONG PARENTS, CHILDREN & EDUCATORS

Because of the variability of features in NF1, some children are more obviously affected by the disorder whereas others are not. It is important for educators to discuss with a child's parents what the child knows about his or her condition. Many parents wait to tell their child about NF until they feel the child is able to handle the information emotionally. How much and what to tell a child must be individualized to his or her particular complications, age, maturity, and level of understanding.

Parents often have mixed feelings about sharing their child's diagnosis of NF1 with school personnel. Some parents fear that a child known to have NF1 will be assumed to have medical problems or learning disabilities, even if he or she does not. They worry that an assumption of learning disabilities may create a "self-fulfilling prophesy" that can lower a teacher's expectations of the child—and, in turn, lower a child's self-esteem and performance—even when no learning problems are present. However, for anyone involved with a child who has NF1, knowledge typically translates to better care.

Thank you for taking the time to learn about neurofibromatosis.

CHILDREN'S TUMOR FOUNDATION
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Founded in 1978, the Children's Tumor Foundation (CTF) is a highly recognized national nonprofit 501(c)(3) organization dedicated to finding treatments for NF.

Our Mission:
Drive research, expand knowledge, and advance care for the NF community.

Our Vision:
End NF.

To become involved and learn about local Foundation activities in your area, please visit our website or contact us at the address or number below.

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