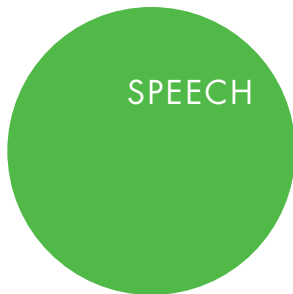




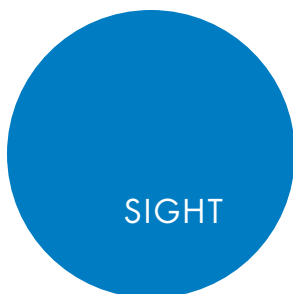
**A SHORT COURSE**



**ON A DISTURBING**



**AND ELUSIVE DISEASE:**



**NEUROFIBROMATOSIS**

- **A serious and misunderstood disorder** 1/2
- **What is neurofibromatosis?** 3/4
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## ● **A SERIOUS AND MISUNDERSTOOD DISORDER**

Each year, thousands of children across the country are affected by neurofibromatosis {NF}, a progressive genetic disorder that causes tumors to form on nerves and other tissue throughout the body. NF is not a rare disorder—it's the most common neurological disease caused by a single gene. Sadly, the majority of those affected are the very young and school age children.

Educators and others who observe and interact with children on a daily basis play a vital role in helping NF children achieve their full potential. Symptoms vary greatly from child to child, and early caregivers should be alert to subtle signs that may help diagnose or manage the disease.

Children and families need particular support from educators because of the high occurrence of learning disabilities {LD} among those with NF. About half of all children diagnosed with the more common form of NF have cognitive deficits or learning disabilities that qualify them for educational assistance. By identifying specific classroom challenges, parents and educators together can help children avoid and counteract common pitfalls such as social isolation, anxiety and depression.



Approximately 50% of  
children with {NF}

1/2

have cognitive deficits  
or learning disabilities  
that qualify them for  
educational assistance.



SPEECH  
MOTOR  
ATTENTION  
VISUOSPATIAL



## ● WHAT IS NEUROFIBROMATOSIS?

**Neurofibromatosis** (pronounced neuro-fi-bro-ma-to-sis and often called NF) is one of the most common genetic disorders, even though many people are unfamiliar with it. NF can affect anyone, regardless of family history, race, gender or ethnic background.

Neurofibromatosis 1 **{NF1}** affects approximately one in every 3,000 individuals. The more rare form, Neurofibromatosis 2 **{NF2}**, affects approximately one in every 35,000 individuals.

Both forms of NF are characterized by the growth of benign tumors called neurofibromas. These tumors are composed of tissue from the nervous system (neuro) and fibrous tissue (fibroma). In NF1, neurofibromas most commonly grow on the skin or on the nerve to the eye. In NF2, neurofibromas most commonly occur in deeper areas of the body such as the spinal cord, and/or brain.

NF is a disorder of the nervous system that can affect many different systems of the body in various ways. Each case is different and requires a unique set of medical specialists and a plan for management.

The gene for NF is always present at birth – passed on by a parent or caused by a spontaneous change in the genetic material at conception. There is nothing a parent can do or not do to cause this change. NF is an autosomal dominant genetic condition, meaning you can not catch it from other people.

## Diagnosis of NF

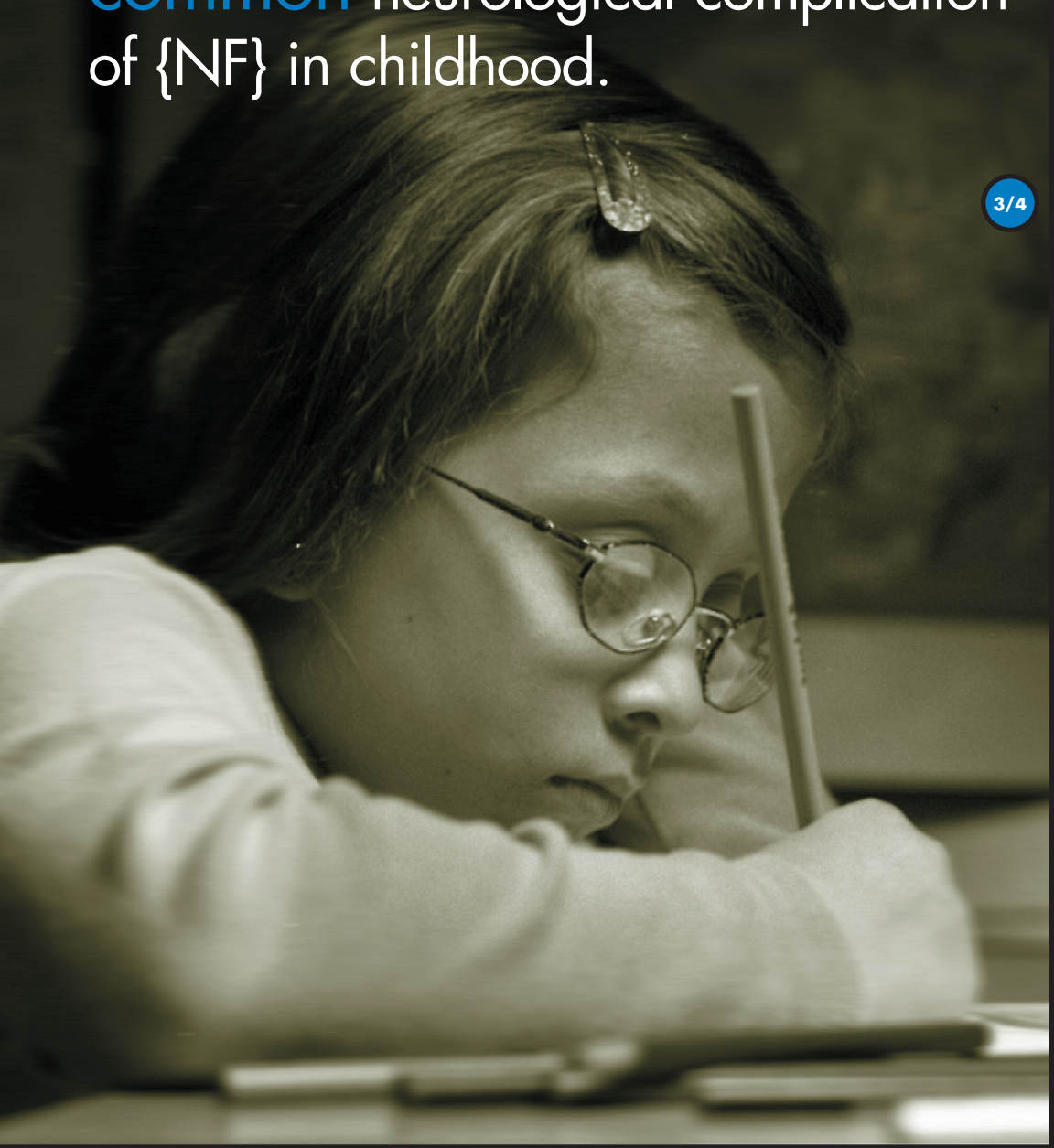
Parents, educators and medical professionals all play a role in recognizing the telltale signs of a child with NF.

The appearance of six or more café-au-lait spots (light tan in color) is the most common sign of NF. The spots are flat, pigmented spots on the skin. In darker-skinned people, café-au-lait spots appear darker in color than surrounding skin. Occasionally, café-au-lait spots may be so pale that they go unnoticed in ordinary light. An examination by a physician familiar with the signs of NF is the best way now available to determine whether NF is present.

Café-au-lait spots are usually present at birth in children who have NF or, generally, appear by two years of age. The number of spots may increase in childhood and occasionally later in life. The spots may be very light in color in infants and usually darken as the child gets older. With NF, café-au-lait spots and freckling can often be seen in the armpit (axilla), where small spots are called axillary freckling, and the groin.

Individuals with neurofibromatosis 2 (NF2) show fewer outward signs of the disease. Tumors affecting both of the auditory nerves are the hallmark. Hearing loss beginning in the teens or early twenties is generally the first symptom.

Cognitive deficits and academic learning difficulties are the most **common** neurological complication of {NF} in childhood.





OBSERVE  
STRENGTHS  
WEAKNESSES  
ASSESS



## ● WHAT EDUCATORS SHOULD LOOK FOR IN CHILDREN WITH NF

### Aside from physical vulnerabilities,

children with NF face social, emotional and cognitive adjustments and challenges. Specifically, NF is known to cause problems with language-related tasks, spatial relationships and fine and gross motor skills. Many parents and teachers also report that NF children may display behavior associated with Attention Deficit Hyperactivity Disorder.

Children with NF and their parents share the same desire for full and healthy development as their peer group. Active support and encouragement from parents, educators and administrators can mean the difference between a thriving, well-adjusted boy or girl and a child who feels isolated and depressed. These simple steps will help anyone become an advocate and ally for a child/family dealing with NF:

### OBSERVE

No two NF children look, sound, act or learn in the exact same manner. Like all kids, children with NF display patterns of individual strengths and weaknesses, in and out of the classroom. Warning signs can range from subtle developmental gaps to fairly obvious dysfunction.

NF children often find difficulty with the following:

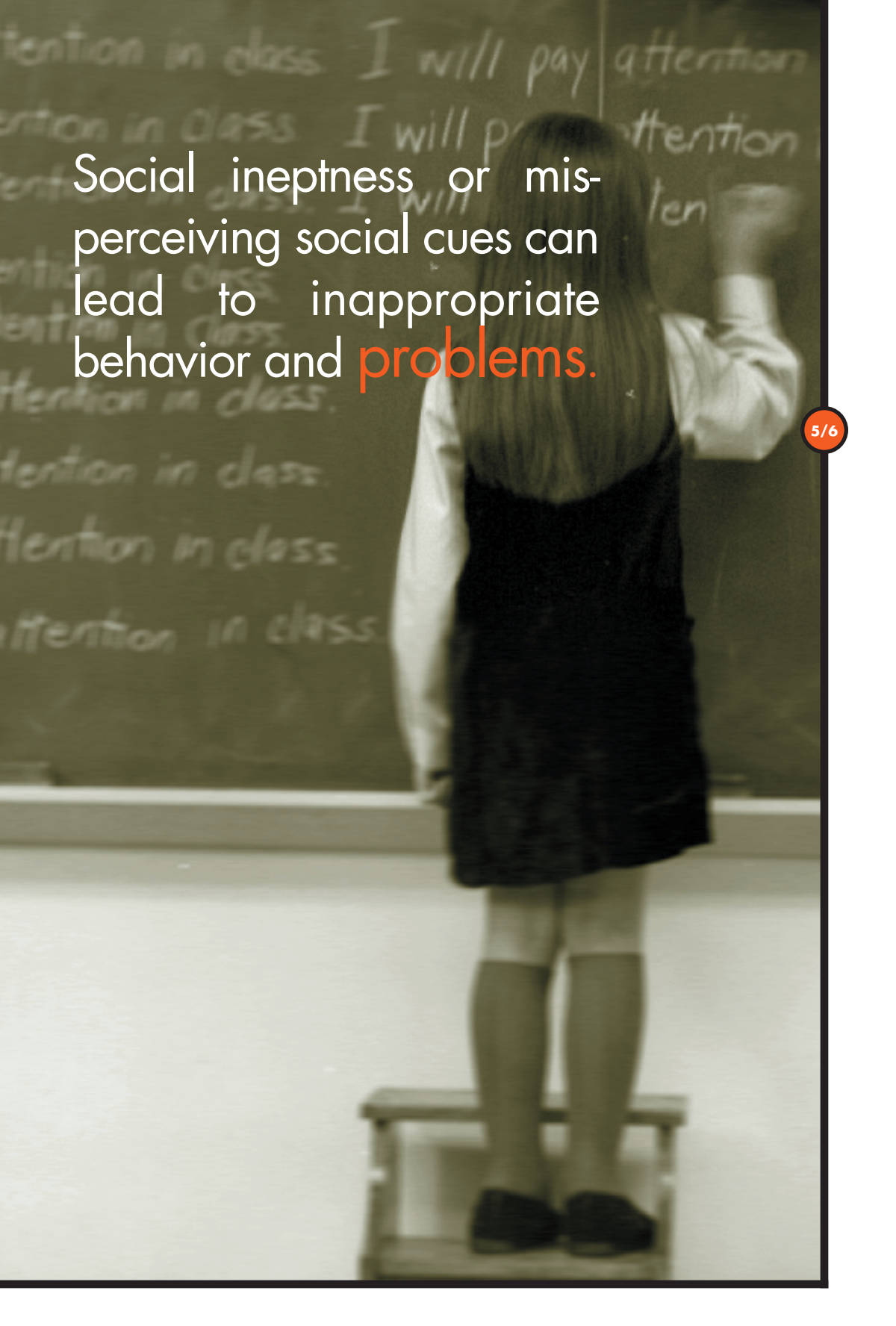
- Language issues such as sound-to-symbol association in reading and phonics, and overall verbal acuity, memory and problem solving.
- Fine motor skills and visual-motor integration such as writing, copying and drawing skills.
- Social and behavioral struggles, often stemming from trouble with listening and interpreting messages, especially in social situations where much talking occurs.

### ASSESS

When a child is diagnosed with NF during the first year of life, a neurodevelopmental evaluation can provide the basis for early intervention and set the stage for future learning strategies.

As the child approaches school age, a thorough psychoeducational assessment can pinpoint a child's unique skills and weaknesses. Parents, social workers and educators can then develop a plan to help the child manage and overcome the effects of NF. A careful assessment can be performed by a school-assigned psychologist or by an independent professional. Many health insurers will cover the cost of an assessment related to an underlying medical condition such as NF.

Social ineptness or misperceiving social cues can lead to inappropriate behavior and **problems.**





ADJUST  
ENVIRONMENTS  
MODIFY  
SIMPLIFY



An assessment will isolate and measure some key brain functions:

- **Motor Skills:** gross functions such as balance and overall coordination, as well as fine motor skills such as the “pincer grasp” function and hand-eye coordination.
- **Learning and Memory:** general cognitive ability (IQ) as well as submeasures of language, visual perception and orientation, reading and certain math skills.
- **Attention and Social Skills:** ability to listen to and carry out instructions, overall attention span and ability to track, perceive and respond to social cues.

## ADJUST

Children with LD often do not understand the nature of their struggles. Parents and teachers can help by identifying the problem in clear, straightforward terms. Open discussion about a child's strengths and weaknesses can alleviate anxiety and counteract peer labels. NF children, while faced with specific cognitive challenges, are more than capable of performing in a full range of learning environments.

These teaching strategies have proven effective in working with children with various NF-related learning disabilities:

- Tell the student everything
- Encourage verbal feedback from the child
- Seek cooperative learning situations with opportunities for active verbalization and conversational turn taking
- Provide additional time and verbal assistance for transition times
- Associate verbal labels with concrete experience
- Verbally teach cognitive strategies for conversational pragmatics
- Observe and expand existing coping strategies
- Apply a reciprocal teaching approach in teaching new tasks
- Encourage verbal mediation so that the child can hear what the teacher is thinking
- Use active verbalization and sub-vocalization to enhance memory tasks
- Simplify layout of assignments
- Keep pencil and paper tasks to a minimum
- Use computer word processor or Alphasmart keyboard
- Provide assistance for tasks requiring fine motor and visual perceptual skills
- Modify timed assignments
- Make all expectations explicit



A diagnosis of NF should alert **clinicians, parents and teachers** to monitor for learning disabilities.



ALLOW TIME  
ENCOURAGE  
UNDERSTAND  
BE POSITIVE



## ENCOURAGE

Children and adolescents with NF share many of the learning and developmental problems of other kids, while also dealing with the physical signs and medical complications of their disorder. The foundation for all intervention is the ability of parents, teachers and others to foster and maintain a hopeful attitude of acceptance and respect for the developing child. The primary goal for everyone involved is to champion the child's successes, encourage his or her talents and to avoid the disappointment trap. It's important for the child to experience a sense of personal worth and accomplishment in at least one area – some special skill that the child works hard at developing. A taste of personal mastery can develop a well of confidence to meet other life goals.

Everyone involved – parents, peers and educators – can also apply these simple intervention strategies to help children with NF transition in and out of the classroom and maintain a positive and respectful attitude:

- Allow extra time to get places
- Develop appropriate expectations
- Provide verbal compensatory strategies (especially for new situations)
- Avoid power struggles and threats

- Take a positive approach
- Provide daily, non-threatening opportunities for contact with non-disabled peers
- Provide an environment marked by consistent and established routines.

## Social and emotional problems in children with NF

Like any child affected by a chronic illness and/or learning disability, children with NF also face a variety of social and emotional adjustment obstacles. Educators and parents can work together to anticipate problems and minimize stress and social isolation.

Some general characteristics to watch for include:

- Appearance of immaturity
- Fine/gross motor awkwardness and uncoordination
- Difficulty transitioning from one motor task to another
- Dependence on “verbal mediation” (i.e., talking themselves through things)
- Social ineptness or misperceiving social cues which can lead to inappropriate behavior and problems with peer relationships
- Difficulty adapting to new situations

## ● RESOURCES FOR PARENTS AND EDUCATORS:

9/10

The NFCA (Neurofibromatosis Clinics Association, Inc.) acts as an information resource for health care professionals, educators, patients and the general public throughout southwestern Pennsylvania and surrounding regions. NFCA contributes funds to support the continued operation and eventual expansion of the NF Clinic in the Neurology Department at Children's Hospital of Pittsburgh.

### **The NF Clinic**

The NF Clinic is located on the second floor in the Neurology Department at Children's Hospital of Pittsburgh. NF patients are evaluated on the fourth Wednesday of every month.

An NF Social worker familiar with all aspects of the disorder, including the need to anticipate classroom challenges, is available for individual, family, or group support. Please contact the NF social worker at Children's Hospital of Pittsburgh at (412) 692-6544 (voice mail) or (412) 692-5255.

### **The Neurofibromatosis Clinics Association**

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**nfca**

*Neurofibromatosis Clinics Association*

Growing awareness. Seeking hope.

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