

# Neuro fibroma tosis

ASSOCIATION OF IRELAND



Learning & Cognitive Difficulties  
in Neurofibromatosis Type 1

**NF1**

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### Disclaimer

*Every care has been taken to ensure the accuracy of the information contained in this brochure. The NF association cannot however accept responsibility for errors or omissions, but where such are brought to our attention the information will be amended accordingly. The author and publisher accept no responsibility for any loss, damage, injury or inconvenience sustained or caused as a result of information supplied in this brochure. It is recommended that anyone who has concerns about Neurofibromatosis first speak to their doctor.*



**Professor Green**  
**Director, Centre of**  
**Medical Genetics**

*"I am delighted to support and endorse the new information sheets for people with Neurofibromatosis 1 and their families. The information will be of great help to the many families in Ireland with NF1, and will help those families to understand better the many ways in which Neurofibromatosis 1 can affect people. The National Centre for Medical Genetics is delighted to be associated with the Neurofibromatosis Association of Ireland, and that the Neurofibromatosis Association of Ireland has funded a genetic counsellor to run a specialised NF clinic in the NCMG. The NCMG has a wealth of experience with Neurofibromatosis and sees many families with the condition throughout Ireland. The NCMG holds genetics clinics in Dublin, Cork, Limerick and Galway, and is happy to see families with NF1, with a referral from their own doctor.*

**Prof. Andrew Green**

## **NEUROFIBROMATOSIS CLINIC**

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## INTRODUCTION

*Explanations and Practical Strategies  
A Resource Guide for Parents and Educators*

## ACKNOWLEDGEMENT

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## HOW TO READ THIS GUIDE

This guide has been designed specifically for parents and teachers of children with Neurofibromatosis type 1 (NF1) and highlights the common learning and cognitive difficulties experienced by many of these children. It also contains a number of strategies targeting specific areas of weakness to help children, parents and educators manage these difficulties.

Areas of weakness in the child's learning and cognition can be strengthened with proper intervention. It is important to recognise that children with NF1 learning difficulties can succeed in school with the help of an individualised education plan and appropriate accommodations in the classroom.

Not all of the strategies contained in this guide will apply to all children. While there are some common learning and cognitive difficulties experienced by children with NF1, these difficulties can affect each individual differently. Therefore, some of the descriptions and strategies may not be relevant to your child. The diagnosis and treatment of learning difficulties should be individually tailored and conducted in consultation with the child's family, school, and relevant health professionals. It is important to note that not all children with NF1 have learning difficulties. This guide is divided into two sections. The first outlines the common areas of learning difficulty for children with NF1: literacy and numeracy. The second section describes areas of cognitive weakness that may underlie these learning difficulties. For example, a difficulty in maths may result from a weakness in any number of cognitive domains, such as language, visual perceptual skills, or the ability to manipulate information in immediate memory.

### **ABOUT NEUROFIBROMATOSIS**

Neurofibromatosis is a term that is given to three different genetic conditions: Neurofibromatosis type 1 (NF1), formerly known as von Recklinghausen's disease, Neurofibromatosis type 2 (NF2), and Schwannomatosis. Each of these genetic conditions causes tumours to form on nerves. Although NF1 and NF2 share a common name, they are caused by abnormalities in two different and unrelated genes. This guide will focus on learning and cognitive difficulties associated with NF1.

#### **Common Features of NF1**

NF1 is a common genetic disorder that occurs in approximately one in 3,000 individuals. It can affect multiple systems in the human body, including the

nervous system, skin, eyes, skeleton, endocrine system, and circulatory system. Common diagnostic features include:

- **Café-au-lait patches:** Individuals with NF1 often have flat, pigmented spots on the skin that usually become obvious during the first two years of life. These vary in size and shape, but do not cause any medical problems.
- **Freckling:** This usually occurs in the armpits or groin.
- **Cutaneous neurofibromas:** These are small, benign tumours that grow around small nerves and develop in or under the skin. They generally begin to appear around puberty and continue to increase in number throughout adolescence and adulthood. The number of neurofibromas varies from person to person.
- **Lisch nodules:** These are harmless brown, disc-like bumps on the iris (coloured part of the eye) that do not affect vision.

### **Complications of NF1:**

NF1 is a complex disorder with a variety of manifestations. A characteristic of NF1 is its diversity of clinical complications from one person to the next. While some people are quite severely affected, others may go through life with such mild cases they never even realise that they have the disorder. Some of the most common complications of NF1 include:

- **Learning difficulties:** Although intelligence is usually within the normal range, 50-60% of children with NF1 experience learning difficulties at school. Some children may require special assistance with school work.
- **Optic gliomas:** Optic gliomas are benign tumours that develop on the nerve pathway from the eyes to the brain. These are relatively common in children with NF1. Although they are usually slow growing and remain stable over many years, some may cause problems with vision and require treatment.
- **Plexiform neurofibromas:** Plexiform neurofibromas are larger, often internal tumours involving a

group of nerves and blood vessels. They can occur in or under the skin or they may grow inside the body and are often difficult to remove completely. These tumours are benign and often do not cause any problems, but they can affect the function of the involved nerve. Rarely, they can undergo a change and become malignant.

- **Scoliosis:** Scoliosis, or curvature of the spine, is typically identified in early childhood and may require bracing or surgical intervention.
- **Bone abnormalities:** Children with NF1 may show abnormal bone development, such as bowing or fracture of the long bones – particularly the lower leg. In rare cases, repeated fracture and incomplete healing may lead to the formation of a false joint in the bone, also known as “pseudarthrosis”.

## LEARNING DIFFICULTIES IN NF1

Learning difficulties represent one of the most common and challenging complications of NF1. Research has revealed that about 50-60% of school-aged children with NF1 are academic underachievers, with many children experiencing marked difficulties in academic areas such as reading, spelling, arithmetic, and written expression. Estimates of the incidence of learning difficulties in NF1 vary considerably, attributed to the use of different terms, diagnostic criteria, and biased study samples (e.g. participants obtained from learning disorders clinics). Despite these varying estimates, without intervention children with NF1 are at a far greater risk of academic underachievement during their school years than children without NF1.

At school, many children with NF1 struggle to acquire the basic skills necessary for:

1. Literacy
2. Numeracy

## LITERACY SKILLS IN NF1

To become a skilled reader, children need to learn a number of basic reading sub-skills such as letter recognition, whole word recognition, knowledge of letter-to-sound rules, and the ability to access meaning from the printed word. Recent findings suggest that a high percentage of children with NF1 experience difficulties with one or more of these reading sub-skills. Many children with NF1 display a specific weakness in their ability to sound out words when reading, known as developmental phonological dyslexia. Children with developmental phonological dyslexia find it difficult to learn phonics. That is, learning the rules about which sounds correspond with which letters. These children experience considerable difficulty reading unfamiliar words and nonsense words (e.g. “ganten” or “gurve”) and tend to over-rely on their word recognition skills (often guessing a word based on its appearance) as they are unable to translate unfamiliar printed words into speech. Some children with NF1 also present with developmental mixed dyslexia, experiencing difficulties both with sounding out words and word recognition.

Problems with reading can lead to the child becoming further behind with their school work and beginning to lack motivation, confidence, and self-esteem.

Children with reading difficulties often also have problems in spelling. Spelling can be particularly difficult for children with phonological dyslexia because the same weaknesses that impact the ability to sound out words when reading also influence spelling.

### **Strategies to Assist Children with: Reading Difficulties**

Often, children with reading difficulties, particularly those with phonological dyslexia, require explicit instruction in concepts such as phonological



awareness (the ability to perceive and manipulate sounds of language). Children must discover that spoken words can be simplified into smaller units of sound, that letters represent these sounds, and that written words have the same number and sequence of sounds as in the spoken form of the word. Children with reading difficulties may benefit from the following strategies:

- Provide systematic and highly structured training exercises, such as:
  - Identifying rhyming and non-rhyming word pairs.
  - Practice learning letter-to-sound rules.
  - Blending isolated sounds to form words.
  - Segmenting spoken words into their individual sounds.
  - Playing language games such as "I spy."
- For children who are struggling with the phonics of reading (those with phonological dyslexia), there are numerous commercially available, computer-based phonics training programs. The effectiveness of some of these has been scientifically documented in treatment studies.
- Avoid situations where the child has to read aloud in class, as this may reduce confidence and motivation. Reserve reading aloud for one-on-one time with the classroom teacher. Alternatively, give the child additional time to read pre-chosen reading material and to rehearse at home the day before. This will help ensure that the child is seen to be able to read aloud, along with their peers.
- Practice reading stories, both to allow the application of newly acquired decoding skills to read words in context and to experience reading for meaning.
- Provide additional time for reading.

- Provide access to lesson information to allow the child to review ahead of time the content of reading material.
- Offer alternatives to multiple-choice tests, such as orally administered tests and a separate, quiet room for taking tests.
- Children who also experience word recognition difficulties may benefit from:
  - o Practice with flashcard reading.
  - o Use of mnemonic strategies – teaching the child associations between words and their meanings by showing flashcards with a word and a mnemonic cue related to the word meaning. These should start out as common words, 2-4 letters in length, and be taught in small groups of 6-10 words. For example, the mnemonic for “each” might show each child receiving a lollipop; or “over” might illustrate going over a bridge.

### **Strategies to Assist Children with: Spelling Difficulties**

- Teach common, irregular words (words that do not follow normal spelling-to-sound rules, such as “what” and “of”) from the earliest stages of spelling.
- Encourage the child to keep a file of frequently misspelled words for reference when writing and provide spelling aids for the child to use (e.g. list of spelling rules such as “i before e except after c”).
- Encourage proof reading, which can be useful for initial correction of spelling. Children with reading difficulties often do not correct their spelling spontaneously as they write, but they can be trained to look for errors that are specific to them.
- Encourage activities that involve building printed words with letter tiles, rather than oral spelling.

- Underline all misspelled words and give the child a chance to correct mistakes before assigning a final grade.

## NUMERACY SKILLS IN NF1

Reading and language difficulties, even subtle ones, can make it difficult for the child with NF1 to acquire numeracy skills. Mathematics has its own language and this can cause a number of problems, particularly for children with reading difficulties. While some children who experience reading difficulties are good at maths, a large number of children with reading difficulties also experience problems in at least some areas of maths, such as those that require many steps or place a heavy load on working memory (e.g. algebra or long division). The effects of maths problems throughout schooling can lead to maths illiteracy in adulthood, impacting upon daily living and vocational prospects.

Children who experience maths difficulties associated with NF1 often have trouble remembering basic number facts in mathematical operations (+ - / x) even though they may understand them and expend great effort trying to do so. For example, they may continue to require concrete aids such as counting on fingers and appear to have difficulty developing efficient memory strategies on their own. Additionally, the presence of other cognitive difficulties such as visual perceptual problems (see below) may result in a specific difficulty with pictorial presentations, confused arrangement of numerals and signs, and difficulty forming representations of abstract information.

### **Strategies to Assist Children with: Maths Difficulties**

- When introducing new maths skills and concepts, begin with concrete examples (e.g. Cuisenaire rods,

abacus) and later move to more abstract concepts when the child demonstrates understanding.

- General mathematical terminology needs to be understood clearly before it can be applied to calculations (e.g. “addition,” “plus,” “sum of,” and “total” all describe a single mathematical process).
- Group similar problems together (e.g. all addition in one section).
- Children with NF1 are often impulsive and may misread problems and maths signs. These children should be encouraged to double-check their work for accuracy.
- Encourage the child to verbalise, or and talk their way through, each step of the problem.
- Once the child demonstrates understanding, encourage frequent repetition and practice of newly learned concepts, as well as practice in reciting basic maths facts (e.g. times tables).
- Rehearse mathematical vocabulary constantly, using multisensory or kinaesthetic methods (e.g. pictures and graphics). However, children with visual perceptual difficulties may become overwhelmed by the presence of complex diagrams or graphs. For these children, it is important to minimise or complement complex visual information with verbal explanations.
- Read and explain story problems, or break problems into smaller steps.
- Put key words into a card index system or on the inside of the student’s maths book so that it can be used for reference and revision.

- Encourage the student to ask questions and explain their understanding of maths concepts as they work.
- Use mnemonic instruction – linking new information to prior knowledge through the use of visual and/or auditory cues, using strategies such as keywords (linking new information to keywords that the child already knows) and pegwords (using a consistent set of rhyming words to represent numbers).
- Encourage the use of estimation, particularly for children who also experience reading difficulties. Teach the child to form the habit of checking their answers against the question when they have finished the calculation. This is a way of “proof reading” calculations and checking whether the answer is possible, sensible, or silly.
- Encourage the child (particularly relevant for those with reading difficulties) to use a calculator.
- Use graph paper to help organise ideas on paper and to help align columns when completing maths problems, particularly for children with visual perceptual difficulties.
- Encourage the child to self-chart their progress (e.g. keeping track of how many and which principles have been learned and how many remain to be learned).
- Remind the child that maths problems are worked from right to left, unlike reading, which is left to right.

### **THE LINK BETWEEN LEARNING AND COGNITIVE DIFFICULTIES IN NF1**

Research suggests that many areas of cognitive functioning contribute to academic success. Children

with NF1 often experience difficulties in a number of cognitive domains such as visual perception, attention, language, and higher level (executive) skills – and these in turn impact their ability to learn in the classroom. These difficulties lead to disadvantages for the child, such as difficulties with comprehension, following task instructions, copying from the blackboard, and handwriting.

## **COMMON COGNITIVE DIFFICULTIES IN NF1**

***Visual Perception, Language, Executive Functions, Attention, Motor Skills, Behaviour.***

### **Visual Perceptual Skills**

When we look around, our eyes are taking in information about what we can see. Our brain then puts these pieces of information together so that we can recognise objects and people. It then goes a step further to help us figure out where things are in relation to each other.

One of the most common cognitive problems seen in children with NF1 is difficulty processing visual information efficiently. Unlike language-based difficulties that are usually readily apparent to parents and teachers, difficulties with visual perceptual skills can often go unrecognised and unaided in the classroom. The way children with visual perceptual difficulties 'view the word' is different from other children. It is thought that they focus on the detail of what they see and fail to grasp the visual image as a whole. Children with visual perceptual difficulties often have problems coordinating what they see with their motor skills (visual motor integration) and may have difficulty figuring out where and how to place written responses on a sheet of paper. Additionally, they tend to not be drawn to toys and activities that rely on building and

construction (e.g. Lego), and they may have difficulty telling the time from an analogue clock.

On neuropsychological tests, children with NF1 often have difficulties with tasks such as:

- Judging the orientation of lines.
- Assembling puzzles.
- Copying shapes.

The problem can even extend as far as:

- Difficulty matching shapes and sizes.

Visual perceptual weaknesses can affect performance in several areas in the classroom. Parents frequently report that children with NF1 have significant problems in the following areas:

- Copying information from a whiteboard or blackboard.
- Judging relationships between objects and shapes in space.
- Reading words in the correct order.
- Aligning columns correctly when completing maths problems.
- Tend to skip lines, reverse or rotate letters, and omit words when reading and writing.
- Arithmetic weaknesses can also result from deficits in visual perceptual skills (e.g. aligning columns of numbers, observing directionality) and in organising their work. In addition to these visual processing difficulties, some children with NF1 have fine motor difficulties and awkward pen

grips, which compound the problem and make it difficult for them to keep up on written tasks in the classroom.

### **Strategies to Assist Children with: Visual Perceptual Difficulties**

- School assignments that rely heavily on copying text may need to be modified due to their load on the child's visual perceptual skills. For example, allow the child to record their responses or check work frequently for visual errors.
- Give the child a simple overview or summary of what will be learned before each lesson, so that some of the confusing visual information can be more easily understood.
- Provide worksheets and tests that are clear, as simple as possible, and well-spaced with less clutter (e.g. fewer problems presented on each page etc.).
- Additional time should be given to complete tasks, especially those requiring complex visual skills. Children with visual perceptual difficulties must compensate when processing visual information, leading to tasks being completed at a slower rate. It is often counterproductive to place time constraints on these students as they tend to become overwhelmed.
- Check work for mistakes, particularly in situations where the child is required to work from the whiteboard/blackboard. If possible, the child may benefit from having the notes provided for them by the teacher.
- Practice tracing shapes and copying pictures. It is important to provide feedback as the child may not be aware of any mistakes.



- Give verbal descriptions to promote understanding and reinforcement of visual information.
- Use graph paper when doing math problems to help keep numbers in line.
- When reading, a note card could be placed under each line to help the child keep their place. When writing on the chalkboard, help the child keep place by writing each line in a different colour of chalk.
- Tasks requiring folding, cutting with scissors, and working with information that is visually complex (e.g. maps and graphs), are likely to require one-to-one assistance.
- Mark the child's desk with "left" and "right" indicators.

### Language

In early studies of the cognitive profile of NF1, it was assumed that language skills were relatively well preserved. This assumption was due, in part, to a greater emphasis on the better documented visual perceptual weaknesses of the disorder. More recent studies have clarified that language skills may be an area of cognitive weakness for children with NF1. On neuropsychological tests, children with NF1 have been found to experience difficulties with tasks of:

1. **Receptive language** – the ability to process either spoken or written language.
2. **Expressive language** – the ability to convey meaning through either speaking or writing.

#### 1. Receptive Language Difficulties

Children with receptive language difficulties may have problems in the following areas:

- Following directions or instructions.
- Understanding complex sentence structures (e.g. passive sentences).
- Understanding the meaning and content of speech.
- Discriminating between sounds.
- Understanding word meanings.
- Understanding lengthy or complex speech.
- May appear not to be listening or ignoring people at times.
- Not keeping up with their classmates, either academically or socially.
- May cause behavioural problems or acting up in class.
- May be easily distracted or drift off when listening to speech or reading stories.
- May appear to be forgetful. For example, they may only complete part of an instruction or remember part of a shopping list.

## 2. Expressive Language Difficulties

Children with expressive language disorders often present with symptoms such as:

- Poor grammatical or sentence structure.
- Limited content in speech.
- Confused meaning and grammar.
- General use of short, simple sentences.

- Difficulty coming to the point or organising speech.
- Difficulty participating in conversations.
- Difficulty recalling or retelling information.
- Difficulty completing oral and written narratives and/or assignments.
- Difficulty finding the right word when speaking.

Other language difficulties, particularly more subtle aspects of language processing such as the ability to perceive and manipulate sounds of language (phonological awareness), have also been found in children with NF1. These may affect success in reading and spelling. Children with poor phonological awareness do not seem to be aware of the sound structure of language, and often:

- Experience difficulty learning rhymes.
- Do not seem to hear sounds properly.
- Find it difficult to segment words into syllables and individual units of sound (phonemes).
- Can't manipulate phonemes.
- Can't recognise words with common phonemes.

Children with this core phonological processing problem may also have poor short-term memory for verbal material. Therefore, they have difficulty remembering instructions (particularly those with a number of steps), lists of words, and spelling. Speech articulation difficulties may interfere with the normal development of phonological awareness and processing. Recent findings also suggest that weaknesses in reading tend to co-occur with weaker verbal skills in areas such as verbal comprehension,

verbal learning, rapid word generation skills, and phonological short term memory.

### **Ways to Help Children with Language Difficulties**

A speech therapist will be able to assess whether a child is experiencing language difficulties or whether other difficulties with speech and language development are present. A formal language assessment will help to identify any specific problems and areas of relative strength. A neuropsychological assessment may also be helpful in identifying any other cognitive problems that are impacting language function.

Some children with language difficulties may require one-to-one therapy to initially manage and develop specific language skills.

Schools and special education teachers can help set up individualised education programs for children experiencing language difficulties. Classroom assistant support may also be required for children with severe difficulties.

### **Strategies to Promote: Language Understanding**

- Gain the child's attention, using their name.
- Avoid abstract language and use simple, short sentences.
- Use visual prompts, pictures, and gestures.
- Use the child's experiences and interests.
- Allow the child extra thinking time (e.g. break up teaching sessions into chunks to allow time for information to be processed).
- Repeat instructions and demonstrate meaning with actions.

- Provide extra time after group sessions to check that the content of the lesson has been understood, and encourage the child to ask for instructions to be repeated, simplified, or written down if they misunderstand.
- It is often helpful to provide a lesson summary, or learning intentions and success criteria in advance or at the start of the lesson, so that the child can concentrate on listening and understanding rather than taking notes.
- Recording lectures and tutorials can be very helpful for children who require repetition of information in order to make accurate notes and to discuss the content with others at a later date.
- If a teaching session introduces a large amount of new terminology, provision of a glossary of key terms (keywords) can be very useful.

### **Strategies to promote: Spoken Language Development**

- Provide ample time for the child to respond.
- Model a short sentence or word, and do not expect the child to repeat or copy it.
- Avoid using questions which require yes/no answers.
- Increase the child's self-confidence by calling on them when you know that they know the answer. Ask non-threatening questions which require only a short answer or opinion.
- Offer games involving naming (e.g. parts of the body, action words).
- Use forced alternatives (e.g. "Do you want to play with the blocks or the sand?") so that the child hears the words to help model their answers.

- Encourage the child to repeat questions to him/herself before responding.
- Seat the child at the front of the room to reduce embarrassment when speaking or responding to questions.
- Prepare the student by saying his/her name before asking questions.
- If the child finds it difficult to respond, assist him/her with clues.
- Reduce anxiety by providing opportunities for small group discussion and participation.
- Allow time to rehearse and respond, particularly during oral presentations.

### **Executive Functions**

Executive functions may be best understood as an umbrella term that encompasses a number of related skills necessary for purposeful, goal-directed activity. Executive functions play an important role in a child's intellectual development, academic achievement, personality, social skills, relationships, and communication. These are important skills which continue to develop into early adulthood.

Executive functions include the ability to:

- Initiate behaviour.
- Inhibit competing actions or stimuli.
- Select relevant task goals.
- Organise a means to solve complex problems.
- Shift problem-solving strategies in a flexible manner.

- Monitor and evaluate behaviour and emotions.
- Manipulate information in immediate memory (working memory).

While not routinely investigated in early studies, executive functions are now a recognised area of weakness for children with NF1. On educational psychology tests, children with NF1 often have difficulties with tasks such as:

1. **Working memory** – Holding information in mind for the purpose of completing a task.
2. **Planning/organisation** – Identifying and organising the steps and elements necessary to carry out an intention or achieve a goal.
3. **Complex problem-solving** – Thinking of strategies, forming abstract concepts, and utilising feedback to generate alternative strategies when one strategy is ineffective.

Weaknesses in working memory may impact all areas of a child's learning. In the classroom, a child may experience difficulty comprehending large amounts of verbal information and completing complex mental arithmetic. These children are often described as forgetful (e.g. have difficulty on tasks with multiple steps) and lose track of what they are doing or saying even after a few seconds. These problems may also impact other academic skills, such as reading and spelling (e.g. losing track while completing reading comprehension tasks).

Planning/organisation weaknesses may result in difficulties finding an appropriate starting point (on classroom tasks and activities around the home), underestimating the time needed to finish a task, problems completing work in an organised manner, becoming overwhelmed by large and complex projects,

and getting caught up in the details of the task. Children with weaknesses in complex problem solving are often described as rather inflexible and concrete in their thinking.

Difficulties in executive functions are often a hidden source of academic underachievement. They are more subtle and less well understood than other cognitive or learning difficulties. And they rarely qualify a child for additional education support.

## **Strategies for Children with: Executive Function Difficulties**

### **Working Memory Difficulties**

- Simplify verbal information and explain concepts clearly. For example, the child may benefit from large chunks of information being presented in a point-by-point format.
- Provide visual instructions and cues, as well as verbal instructions.
- Provide frequent repetition of information, reinforcing the information in an alternative way if possible.
- Keep an extra folder of handouts so a child can easily replace lost ones.
- Ask the child to repeat instructions to check their understanding. This also helps to reinforce instructions in the child's memory.

### **Planning/Organisation and Complex Problem-Solving Difficulties**

- Help the child find an appropriate starting point, particularly on complex tasks.



- Provide close supervision during the planning stages of school-based tasks or assignments.

This may involve:

- o Breaking larger tasks down into smaller, more manageable steps.
  - o Providing a list of instructions for the child to follow.
  - o Assessing understanding of task requirements at each step.
  - o Cueing with prompts if the child continues to experience difficulty knowing how to approach the next step.
  - o As the child begins to manage these small steps, tasks can gradually increase in length and complexity.
- Ask the child to explain their approach to a new task or assignment.
  - Offer encouragement and rewards for good ideas.
  - Use a calendar to keep track of important events, such as assignment due dates, so that appropriate planning can be implemented.
  - Organise everyday needed items. Have a place for everything and keep everything in its place. This includes clothing, school bags, and school items.
  - At the end of the school day, help the child check that he or she has everything needed for homework. The same can be done at home when getting ready for school. Checklists of materials needed are helpful for the child to use.
  - Create a schedule for homework, including a plan for when assignments should be completed. Check periodically on the status of the child's assignments to ensure that they are following the plan.

- Use a different coloured notebook for each subject or a folder with dividers and pockets for each class, to keep the child organised at school.
- Teach the child to be responsible for keeping their notebook organised, assignments recorded, and homework turned in by graphing, charting, or rewarding when he or she is successful.

### **Complex Problem-Solving Difficulties**

- Use concrete and manipulative materials to demonstrate concepts. Allow the child to use fingers and other aids that are useful.
- When assigning long-term independent tasks, such as book reports or term papers, provide a sequential list of tasks for the child to follow. Help the child outline the steps needed to complete each task. This will also help them learn to plan and manage time more effectively.
- A confused child may not know which part of a task is confusing. Help the child learn how to determine those parts and how to be specific when asking for help. It may be just one word or phrase that is confusing them, and not the whole task.
- Explain words and phrases that have multiple or subtle meanings, such as idioms.

### **Attention**

Attention is the process of selectively concentrating on one aspect of the environment while ignoring other things. Current research suggests that there are different types of attention that rely on different brain systems. Therefore, it is possible that a child may experience difficulty with one aspect of attention and not with another. Attention difficulties are among the most frequent concerns raised by parents of children with NF1 and pose significant challenges to academic performance and achievement.

On educational psychology tests, children with NF1 often have difficulties on tasks of:

- Sustained attention – the ability to focus attention and concentrate for extended periods of time.
- Switching attention – the ability to shift attention from one task (or aspect of a task) to another in a flexible and efficient way.

Children with sustained attention difficulties are likely to find it hard to concentrate and pay attention to one task for any length of time. In a classroom situation, instructions and teacher directions can fade out of the child's awareness because a minor or more compelling task or idea has captured their attention. Conversely, a child's attention can become quite fixated, where they persevere with a course of action and find it difficult to switch their attention to a new activity or idea.

Difficulties with sustained attention are likely to make it difficult for the child to complete mentally effortful or lengthy tasks in the classroom, or homework.

### **What is Attention Deficit Hyperactivity Disorder?**

ADHD is a common developmental and behavioural disorder that is characterized by poor concentration, distractibility, hyperactivity, and impulsiveness that are inconsistent with the child's age. Children and adults with ADHD are easily distracted by sights and sounds in their environment, cannot concentrate for long periods of time, are restless and impulsive, or have a tendency to daydream and be slow to complete tasks.

Different symptoms of ADHD may appear in different settings, depending on the demands the situation may pose for the child's self control. A child who "can't sit still" or is otherwise disruptive will be

noticeable in school, but the inattentive daydreamer may be overlooked. An impulsive child who acts before thinking may be considered just a “discipline problem,” while the child who is passive or sluggish may be viewed as merely unmotivated. Yet, both may have different types of ADHD.

Although all children can be restless, act without thinking, and daydream, it is when the level of hyperactivity, distractibility, poor concentration or impulsivity begins to affect performance in school or behaviour at home, that a diagnosis of ADHD may be considered.

Symptoms of ADHD often become apparent in preschool or early school years and can cause serious difficulties. There are three recognised subtypes of ADHD:

**1) Hyperactive-Impulsive** – Children with this subtype of ADHD often exhibit behaviours such as:

- Frequent fidgeting and squirming when required to sit still.
- Running about or climbing in situations where it is inappropriate.
- Difficulties playing or engaging in activities quietly.

Concerns regarding social skills and peer interactions have often been reported in studies of children with NF1. Common descriptions of the behavioural profile of children with NF1 include being shy, a loner, awkward around peers, and having difficulty forming relationships with peers. However, very few studies have explored these issues in detail. Children with NF1 experience significantly greater levels of anxiety, withdrawal, depression, and somatic complaints, and they have poorer overall social skills than their peers. In addition, learning difficulties and low academic

achievement are associated with less developed behavioural and social skills. Some of the characteristic behaviours seen in children with NF1 include:

- Impulsivity – Difficulty controlling urges and impulses.
- Hyperactivity – Unusually high rate of purposeless motor activity.
- Inattention – Inability to focus on one activity for a reasonable length of time.
- Social skills deficits – Immature or inappropriate responses in social encounters and difficulty perceiving what other people are doing, saying or demonstrating, such as “reading between the lines” or noticing and interpreting facial expression and body language.
- Inflexibility – Overly excited or unsettled by changes in routine, or an inability to shift easily from one activity to another.

In NF1, these behavioural difficulties are often related to the presence of problems with attention, such as ADHD. It is important to note that not all children with NF1 will have behavioural or social difficulties, and not all children who display the characteristic behaviours above will have ADHD.

It is also important to fully investigate and rule out conditions that may cause these behaviours as the treatments may be very different. As discussed above, attention problems can interfere with a child’s ability to learn in the classroom. They may also impact the child’s relationship with their peers and their peers’ relationships with them. Children with social skills deficits often have trouble reading subtle social nuances, such as realising that someone is annoyed or frustrated with them. They can also be shunned by their classmates because of their inappropriate

behaviour. They may experience trouble making and sustaining friendships. Without friends, a child feels isolated and may withdraw from social situations, including school.

### **Strategies to Help Children with: Behavioural Difficulties**

- The most important way to eliminate problem behaviours is to help the child develop confidence. Ensure success by focusing on the child's strengths rather than their weaknesses.
- Positive reinforcement has been shown to be very effective in increasing desired behaviours.

Thus, when a desired behaviour is exhibited, respond in a way that is likely to increase its reoccurrence. Although punishment suppresses unwanted behaviours, positive reinforcement is much more effective in helping children develop alternative, more functional behaviours.

Some ways to help set up positive reinforcement schedules are as follows:

- o Use a sticker chart, so that a small reward is given to the child after the sticker chart is filled up.
- o Reinforce by using praise, giving the child access to a desired game for 10 minutes, or a small toy they want.
- o Draw up a hierarchy of behaviours you want the child to exhibit. For example, if a hyperactive child is always out of their seat, one reinforcer could be given for sitting in their seat, while a second could be given for working while sitting.
- o Reinforce immediately. It is important to note that the longer it takes to reinforce a wanted behaviour, the less effective the reinforcer will be. Immediate reinforcement (e.g. once the sticker chart is filled up) works best.

- o Build anticipation for earning the reinforcer – this can help motivate children. The reinforcer should be varied, as the child is likely to get tired of the same thing over and over again.
- o Describe the appropriate behaviour to the child. Unless explicitly told, the child may not be aware of what behaviours are reinforced. Describing the behaviour can also help review your specific expectations.
- “Response cost” could be used in conjunction with positive reinforcement. Response cost is losing what you have earned for good behaviour if inappropriate behaviour returns. To be effective, however, more reinforcers must be earned than lost; so there should be more opportunities for success rather than failure.
- Be consistent with directions, rules, discipline and organisation.
- Try to accommodate inflexibility by setting limits for the child and telling them specifically what you want.

### **Strategies to Help Children with: Social Skill Difficulties**

- Rather than assume a child will pick up appropriate social behaviours, actually teach them to the child.
- Demonstrate or model appropriate ways to act rather than simply telling the child.
- Role-play different social situations a child may be in and discuss the possible consequences of their hypothetical choices.
- Observe the child interacting with peers and try to identify the situations he or she is having difficulty in and why.

- Teach children how to play games so when they are with peers they will know how to play them.
- Teach the child to recognise facial expressions, body language and moods.

### **Motor Skills**

Well-developed motor skills are the result of an intricate set of cognitive and physical processes which result in smooth, targeted, and precise actions. Motor skills play an important role in a number of areas such as activities of daily living (e.g. self-care skills, opening doors with knobs, using a knife and fork, and cutting paper with scissors). Basic motor skills, such as grasping and manipulation, develop during the infant and toddler years. These skills are subsequently refined during preschool and early school years.

#### **Motor skills can be classified into:**

- 1. Gross motor skills** – simple, large muscle group actions such as running, ball skills, and riding a bike.
- 2. Fine motor skills** – the coordination of small muscle groups which typically involve the hands and fingers for activities such as grasping, releasing, and picking up small objects.

Both gross and fine motor difficulties are common in children with NF1, with estimates at approximately 50%. A significant proportion of children with NF1 would benefit from receiving occupational therapy at some stage during their childhood.

Children with gross motor difficulties may:

- Bump into things.
- Appear clumsy.



- Fall over frequently.
- Have trouble with sports and other physical activities.
- Appear to have poor balance and coordination.

**Children with fine motor difficulties may have difficulties with tasks such as:**

- Handwriting.
- Drawing and copying.
- Manipulating items of clothing such as shoe laces, buttons, and zippers.
- Coordinating the use of cutlery.
- Cutting around shapes with scissors.

Gross and fine motor difficulties may impact a child's overall school experience in a number of areas, such as sporting success, keeping up with peers on the playground, and completing written tasks efficiently in the classroom.

### **Strategies for Children with: Motor Difficulties**

- Posture is an extremely important element to consider in assessing gross motor skills. Adequate posture may make all the difference between being able to execute an action or not.
- Use manipulative activities to increase fine motor control. For example, practice threading small beads on a string or manipulating pieces of Lego. Additionally, the child could be encouraged to make small animals or other objects from play dough or clay.
- Practice cutting pictures out from newspapers or magazines. If a child is finding this difficult, try

drawing an outline around the area with a thick black marker.

- Occupational therapy could be considered for younger children to help teach pencil-grip and basic handwriting skills. In some cases, the use of a computer word processor may be beneficial for school written assignments. Word processors are particularly useful when several revisions are required.
- Allow the child to dictate longer writing tasks to someone who can do the writing, or let the child record themselves on an MP3 player.
- Proper posture is important for good writing. Make sure the height of the child's chair and table is appropriate for him or her. Furthermore, pencil grip devices can be helpful for many children.
- Provide children with alphabet and number charts to help them remember how to form the symbols when they are writing.
- Have the child write on every other line, as it's easier for them to make corrections.
- Allow the child to trace if he or she has trouble drawing.

### **Other Cognitive Difficulties**

The presence of memory difficulties in children with NF1 is an area of controversy and they have not been routinely documented in studies. At present, it is unclear whether the memory deficits that have been reported may more accurately reflect primary language or visual perceptual difficulties, rather than a pure memory deficit. Further research is needed to clarify the role that NF1 plays in memory function.

### **What if the Strategies Don't Seem to be Working?**

Selecting and monitoring the effectiveness of implemented strategies should be an ongoing process. If, after monitoring the child's progress, the accommodations don't appear to be helpful changes should be made with the involvement of the child, parents, educators, and relevant health care professional. It is essential that the strategies address the child's specific areas of cognitive weakness and they be tailored to the individual learning requirements of the child. An educational psychology assessment is the optimal way to fully understand the child's cognitive strengths and weaknesses and, thus, help with implementing strategies that will most benefit the child with NF1.

### **HOPE THROUGH RESEARCH**

The understanding of NF1 learning difficulties has made dramatic advances in the last few years. Research studies have unravelled some of the changes that can occur in the brain in NF1, and as a result are identifying approaches for drug treatment for learning difficulties. For the latest information on learning difficulties and other complications of NF1 visit the website of the American Children's Tumor Foundation. [www.ctf.org](http://www.ctf.org)

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### **NF1 BROCHURES**

- OVERVIEW OF NEUROFIBROMATOSIS TYPE 1
- A GUIDE FOR EDUCATORS
- LEARNING & COGNITIVE DIFFICULTIES
- NEUROFIBROMATOSIS TYPE 1 FOR TEENS
- THE CHILD WITH NEUROFIBROMATOSIS TYPE 1
- TALKING TO YOUR CHILD
- READERS 100 QUESTIONS ANSWERED

### **LEAFLETS**

- NF1 REVIEW CHECKLIST FOR CHILDREN & ADULTS
- NEUROFIBROMATOSIS – A BRIEF INTRODUCTION
- SCHWANNOMATOSIS
- CONTACT FORM

### **CLINICAL GUIDELINES FOR MANAGING NF1**

- FOR ADULTS
- FOR HEALTH PROFESSIONALS

### **NEUROFIBROMATOSIS TYPE 2 BROCHURES**

- FOR FAMILIES
- FOR HEALTH PROFESSIONALS

### **HANDBOOK**

- NF IRELAND HANDBOOK

## Neurofibromatosis is a Little Known Genetic Condition and Can Manifest Itself in a Whole Lot of Different Ways

The care of persons with NF is made complex by the wide range of expression of the disorder. It is difficult to predict the specific problems that will occur in a particular individual. Diagnosis is made if an individual has two or more of the following features.

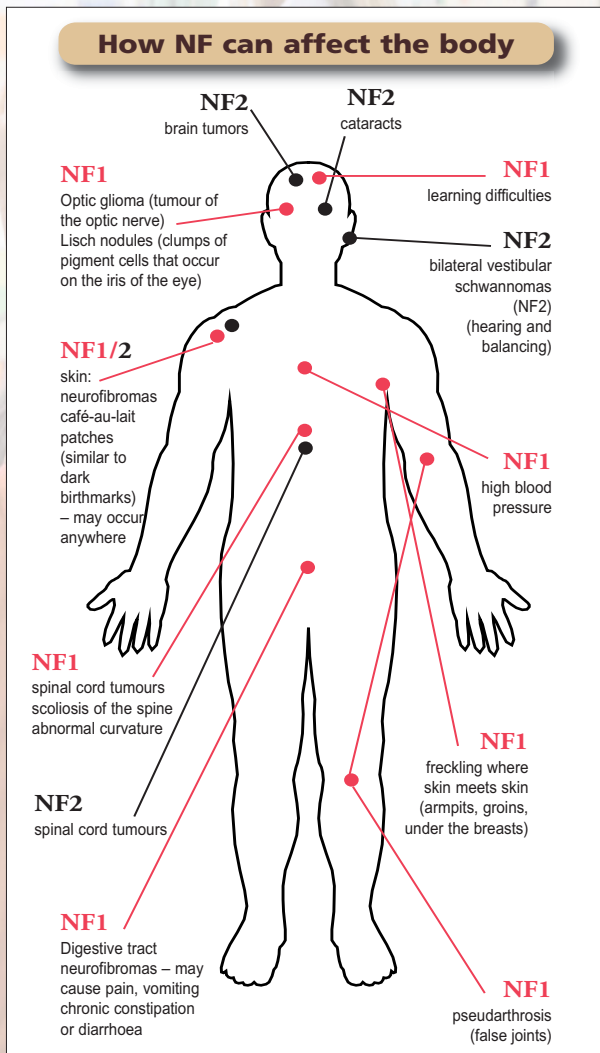
The diagnosis is based on the following clinical criteria:

1. **Six or more (café au lait)** coffee coloured patches sized 5mm or over in pubertal individuals and over 15mm in size in post pubertal individuals.
2. **Freckling** under the **arm** or in the **groin** area.
3. **Two or more Neurofibromas** of any type (growth of tumours on nerve tissue anywhere on the body) usually first seen on the skin.
4. **Plexiform Neurofibromas** – large bundle of nerves are thickened and appear as a soft tissue mass under the skin, these growths often large, can change the normal shape of the body.
5. **Optic Glioma** – thickening of the optic nerve.
6. **Lisch Nodules** – clumps of pigment cells that occur on the iris of the eye.
7. **Orthopaedic** problems include **scoliosis** (curvature of the spine) **abnormal bone development**, such as overgrowth in long bones causing bowing and deformity that result in fractures, which fail to heal.
8. **First-degree relative with NF** e.g. parent, sibling, offspring.
9. **Learning Difficulties**. As many as 50% of children with NF have short attention span, appear clumsy and uncoordinated. Problems particularly with arithmetic and spelling are common.

## Neurofibromatosis Type 2

Another rarer type of Neurofibromatosis and distinct in its clinical feature is **NF2**. The gene for **NF2** is located on chromosome 22, Features include:

**Vestibular schwannomas** (tumour on hearing nerve). **Schwannoma** (type of tumour of the substance that covers nerve fibres). **Meningiomas** (tumour of the covering of the brain). **Cataract**.



A photograph of a young man's back, showing several light brown café-au-lait patches. Three red arrows point from a text box above to these patches. The background is a bright blue sky with white clouds and a green field.

## CAFÉ-AU-LAIT PATCHES

**NEUROFIBROMATOSIS: Tell-Tale Signs**  
*Café-au-lait patches,*  
*6 or more consult your Doctor*



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