



Attention and Executive Function Concerns in Children with Noonan Syndrome

A GUIDE FOR TEACHERS

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What Is Noonan Syndrome?

Noonan syndrome (NS) is a relatively common genetic syndrome caused by changes in one of several genes that are essential for a child's growth and development.

NS can be inherited from a parent or it can be sporadic (de novo), meaning the child is the first person in the family to be diagnosed with NS.

The variations in genes associated with NS can influence the child's overall development, including their brain development. While NS can potentially impact many different aspects of development (e.g., physical, social, emotional, behavioral, etc.), the specific areas that are affected tends to vary widely from one child to another.

Some of the more common characteristics include:

Physical appearance and growth

- Short stature and growth challenges
- Distinctive facial features
- Skeletal issues
- Eye or skin conditions

Risk for medical problems

- Heart disease
- Bleeding disorders
- Hearing loss
- Vision problems
- Chronic muscle and joint pain
- Feeding and gastrointestinal problems



Risk for cognitive, social, emotional, and behavioral difficulties

- Motor, speech, and language delays
- Learning disabilities
- Attention and executive function problems (e.g., ADHD)
- Difficulties with social functioning:
 - Can appear socially immature (act younger than peers)
 - Might have interests of a younger child
 - May have difficulty making and maintaining friendships
- School attendance and performance can be negatively impacted by frequent medical appointments and/or health problems

What Is Attention Deficit Hyperactivity Disorder?

Attention Deficit Hyperactivity Disorder (ADHD) is a common neurodevelopmental condition which is characterized by hyperactive, inattentive, and/or impulsive behaviors that begin early in life. These symptoms occur at home, at school, and in the community.

Difficulty focusing on activities, especially non-preferred activities, is a key component to ADHD. While some children with ADHD can attend appropriately to something they enjoy (e.g., screen time, Legos), it may be challenging to direct their attention away from that activity. As such, children with ADHD may seem oppositional, when in fact they are struggling to shift their attention to more challenging tasks (e.g., homework).

Inattentive behaviors can include:

- Becoming distracted by others talking or someone walking by the door
- Having trouble getting started on a task
- Forgetting steps in a task

Hyperactive and impulsive behaviors can include:

- Frequently interrupting others
- Acting without thinking
- Blurting out answers without a filter
- Getting up and walking around the classroom
- Being unable to sit still



What Are Executive Functions?

“Executive functions” (or EF) are skills that help children organize and complete goal-oriented behaviors. These skills continue to develop throughout childhood into early adulthood.

EF skills help a child:

- Figure out how to begin a task
- Plan out the steps needed to complete multi-step tasks (e.g., follow directions, complete a math problem, clean-up following activities)
- Pay attention to multiple things at once (e.g., take notes from the board while the teacher is talking)
- Shift attention from one task to another
- Hold attention for extended periods of time and resist distraction (e.g., peers talking, computer screens)
- Grasp and understand a complex situation
- Think before doing something (inhibitory control)
 - Examples of poor inhibitory control can include saying things out loud, yelling, getting up, and touching objects or peers
- Control their behavior in the face of frustrating tasks
 - Examples of poor frustration tolerance include giving up easily or exhibiting large emotional/behavioral outbursts when things aren't going well
- Use problem-solving skills to be flexible and adapt to changes in the environment

A significant delay in the development of EF skills can impact a child's academic, social, and emotional functioning.



A helpful analogy is to think of EF as an orchestra conductor. A conductor tells each musician what and when to do something, in order for the instruments to start and stop so they can play in the correct sequence. EF skills are the “conductor” for the brain.

What Is the Relationship Between Attention, Executive Functions & Noonan Syndrome?

ADHD symptoms can interfere with the ability of some children with NS to learn and hold onto information. They may struggle to consistently show their best abilities in educational and testing environments. Teachers more frequently report increased difficulty with planning, organization, and overall time management in children with NS.

Medical problems common in children with NS, such as poor hearing, fatigue, and chronic pain, can also interfere with a child's ability to learn and pay attention in class.

While difficulties with attention and executive functioning are common in children with NS, *not* all children with NS have executive functioning difficulties and/or are diagnosed with ADHD. Each child with NS can have different symptoms with varying degrees of difficulty associated with each symptom. Therefore, it is important to evaluate and support each child according to their individual needs.



About 1 in 3 children with NS meet diagnostic criteria for ADHD

Some important functions that might be impacted in a child with ADHD and/or EF problems:

Working memory

- Forgets steps within a task
- Has difficulty completing multi-step tasks (e.g., word problems, cleaning up an area)
- Has trouble concentrating on chores and activities

Self-monitoring

- Unaware of how a behavior affects others
- Does not check work for mistakes
- Has a poor understanding of one's own strengths/weaknesses

Inhibition

- Responds with an initial reaction without thinking

Attention

- Has problems with attending to and following verbal instructions
- Cannot sustain attention for more than a few minutes

How Do I Know If a Child with NS is Struggling with Attention and Executive Functions?

Signs of attention and EF problems may look different depending on the age of the child. Common examples include:

In early childhood

- Has problems with emotional control
- Exhibits low frustration tolerance
- Demonstrates disruptive or unsafe behaviors that frequently require caregiver attention and supervision
- Has problems understanding social cues
- Asks repetitive questions even though they have been given the answer
- Has difficulties adjusting to unexpected changes in plans
- Struggles to organize thoughts and engage in conversation



In adolescence

- Makes frequent, careless errors and does not check their work
- Often requires help and reminders to complete multi-step instructions or projects and daily activities
- Is slow to start assignments and tasks
- Does not use a checklist or planner, even when one is available
- Requires more time than peers during times of transition (e.g. moving between classes)
- Has problems waiting in line (e.g., moving too slow or too fast)
- Gets distracted easily
- Struggles to gather materials and keep things organized
- Is unable to keep his/her room or desk clean

Primary components of attention problems are:

1. Inconsistent ability for children to show what they have learned
2. Inability to complete tasks

Because of this, teachers and caregivers may perceive that a child is purposefully ignoring or defying them. While this may sometimes be the case, more often the child wants to do well, but their inability to control their emotions and behaviors interferes with their ability to demonstrate their knowledge and skills.



How Can I Provide Support for Attention or EF Challenges in the Classroom?

For students in the United States, the following interventions can be included in a student's Individualized Education Program (IEP) or 504 Plan. They can also be provided as informal supports in a classroom. Teachers can also request a psychoeducational or neurodevelopmental evaluation to have a student's abilities assessed. Reports from these evaluations can provide individualized recommendations.

General recommendations for all ages

- Provide preferential seating. For example, seat the child closer to the teacher and away from distractions, such as windows, doors, and other talkative children.
- Break down multi-step tasks into smaller, more manageable components.
- Have the child repeat and/or rephrase verbal directions whenever realistically possible. This will help ensure they have attended and comprehended instructions.
- Clearly identify transitions and provide incremental warnings.
- Provide explicit instruction on how to organize, study, and take notes.
- Educate the child on how to use a planner to document tasks and assignments.
- Create a routine for turning in homework.
- Provide frequent, scheduled breaks in which the child is allowed to move around and expend energy
- Should the child require disciplinary action or additional instruction time, *removing recess privileges is not recommended.*
- Present information in a multi-modal (e.g., visual & auditory) format whenever possible.
- Ensure regular communication between home and school. Daily, weekly, or monthly plans can be developed to monitor the child's behavior and schoolwork.
- Focus on how to change the outcome rather than pointing out the problem behavior. Also, be sure to focus on the child's positive moments throughout the day.
- Utilize a hands-on instruction method. Teachers might utilize a "tell me, show me, let me try", or "show me again" instructional technique.
- It may be helpful for teachers to model or demonstrate specific steps of a task. Saying each step out loud can also be effective for those children who learn better with self-talk strategies.
- Provide instruction on social skills. Have the child practice problem solving different social scenarios or discuss different ways to handle a situation.



Recommendations for preschool and early childhood

- Repeat information to ensure that the child has heard and attended to directions in a large group setting.
- Provide supervision for independent work time.
- Give one direction at a time. For example:
 1. First take one crayon out
 2. Color part of your picture with that crayon
 3. Put the crayon away
 4. Take out a new crayon
 5. Now color another part of your picture
- Provide a “cubby” or “focus fort” for independent work time.
- Provide a picture checklist of tasks.



Recommendations for adolescence

- Provide extended time and a minimally distracting environment for timed tests and independent assignments.
- Use a visual schedule of activities/tasks, and check off items on the lesson outline as material is presented.
- Utilize “naturally interesting” material whenever possible. This is especially important when teaching new or difficult skills. For example, when learning essay writing, let the adolescent write about their favorite topic.
- Provide close monitoring and intermittent, discrete prompting to ensure that the adolescent stays on task.
- Give “stop and think” reminders before the adolescent responds to task demands.
- Use mastery learning to decrease the homework workload. Once an adolescent has mastered the skill, they should then be able to stop the assignment.
- Redirect the adolescent to more appropriate behavior through verbal and visual cueing when negative behaviors emerge.

POINTS FOR TEACHERS TO KEEP IN MIND

- Children and adolescents with NS will likely need help from teachers to develop and improve executive function skills over time.
- Continued practice of skills is important to show growth in learning these concepts.
- Focus on the quality of skills gained rather than the quantity.
- Change is gradual and educators will need to be flexible in their approach to working with students who struggle with these behaviors.



Learning and Memory Strategies

- Some children with NS have difficulty with retrieval (i.e., free recall) of information. Teachers may be able to better assess learning and knowledge by providing response choices rather than requiring the child to generate their own answers or respond to open-ended questions.
- Many children with NS learn better with a “contextual story” format rather than formats requiring memorization of unrelated information.
- Elaborated memory strategies may assist students with NS in retaining information. Examples include drawing pictures to accompany spelling words, relating new information to everyday life or concepts they already understand, and identifying and emphasizing relationships between words or concepts.
- Studying about a new concept or topic over multiple shorter learning sessions will be more effective than a single longer session.

Other Classroom Considerations

Accommodations for snacks/feeding

- Some children with NS may require food-related accommodations, such as more frequent snacks, reminders to eat or drink, or other mealtime support (e.g., feeding through a g-tube). These accommodations can help children to meet growth goals and also support better energy and attention during instruction time.

Breaks and naps

- Due to health issues and fatigue which are common in NS, it may be important to schedule frequent breaks and/or longer naps for young children with NS, or to allow naps at an older age than is typical.

Communication

Good communication with parents of your student with NS is essential. We especially recommend sharing with parents if any of the following classroom concerns are observed:

- Frequent headaches.
- Severe fatigue.
- Falls or other injuries that may occur at school. Some children with NS have bleeding problems that cause easy bruising or clotting problems.
- Teasing/bullying. Children with NS may experience physical or growth differences that may affect them socially. Maintaining a healthy school environment will require awareness and support from teachers and peers.
- Medication effects. Teacher feedback and help to inform treatment planning with regard to effectiveness of medications for ADHD or other behavioral difficulties in school, as well as potential side effects.

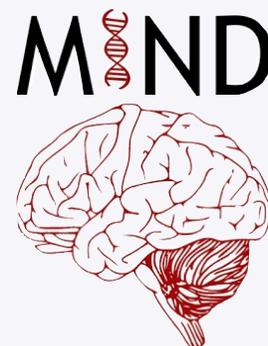
Resources

Noonan Syndrome

- Genetics Education Materials for School Success (GEMSS): gemssforschools.org
- NS Resources, Wessland Family Site: wessland.com/noonansyndrome.htm
- GeneReviews: ncbi.nlm.nih.gov/books/NBK1124
- Noonan Syndrome Foundation: teamnoonan.org

ADHD and Executive Function

- *Smart but Scattered: The Revolutionary Executive Skills Approach to Helping Children Reach Their Potential* by Peg Dawson, Ed.D. and Richard Guare, Ph.D.
- *The Organized Student: Teaching Children the Skills for Success in School and Beyond* by Donna Goldberg.
- *Super Study Skills: The Ultimate Guide to Tests and Studying* by Laurie Rozakis, Ph.D.
- National Resource Center on ADHD: help4adhd.org
- Children and Adults with Attention Deficit Hyperactivity Disorder (CHADD): chadd.org



WRITTEN BY

Meghan Clifford, Psy.D. & Rene Pierpont, Ph.D., L.P.

The MIND Group at the University of Minnesota

<https://z.umn.edu/mindgroup>

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