



ATTENTION DEFICIT HYPERACTIVITY DISORDER AND TUBEROUS SCLEROSIS COMPLEX

What is ADHD?

Attention Deficit Hyperactivity Disorder (ADHD) is a common neurobehavioral disorder. It is usually first diagnosed in childhood and often lasts into adulthood. Children with ADHD have trouble paying attention, have impulsive behaviors (may act without thinking about what the result will be), and in some cases are overly active.

Between 5% and 7% of school age children and 2-5% of adults meet criteria for ADHD and have symptoms that interfere with academic performance, work, and social activities. In tuberous sclerosis complex (TSC), ADHD may be even more common. As many as 25% to 50% of children with TSC may experience ADHD. ADHD often accompanies epilepsy and intellectual disability, and many children with autistic spectrum disorders will have problems with inattention, hyperactivity and/or impulsivity.

What Are the Symptoms of ADHD?

The main groups of symptoms seen in ADHD are inattention-related behaviors, hyperactivity-related behaviors and impulsivity-related behaviors. The hyperactivity and impulsivity usually appear first, often beginning prior to the school age years. Inattention may not be recognized until elementary school. The child with hyperactivity may fidget and squirm, can't stay seated, runs when he or she should walk, is noisy, and is often described as constantly on the go. The impulsivity is seen as trouble waiting their turn, frequently interrupting others, and excessive talking even when expected to be quiet. The child with inattention has trouble concentrating and is easily distracted, makes frequent careless errors, appears not to listen and seldom completes work, is disorganized and tends to procrastinate, and is forgetful and frequently loses books, homework or clothes.

As people with ADHD get older, the symptoms change in subtle ways. The teenager with hyperactivity and impulsivity feels restless and moves quickly from one task to the next, frequently failing to complete projects. While most people have some of these symptoms some of the time, the person with ADHD has many of these symptoms most of the time.

There are three different types of ADHD, depending on which symptoms are strongest in the individual:

- 1. Predominantly Inattentive Type:** It is hard for the individual to organize or finish a task, to pay attention to details, or to follow instructions or conversations. The person is easily distracted or forgets details of daily routines.
- 2. Predominantly Hyperactive-Impulsive Type:** The person fidgets and talks a lot and has trouble sitting still for long (e.g., for a meal or while doing homework). Smaller children may run, jump or climb constantly. The individual feels restless and has trouble with impulsivity. Someone who is impulsive may interrupt others a lot, grab things from people, or

speak at inappropriate times. It is hard for the person to wait their turn or listen to directions. A person with impulsiveness may have more accidents and injuries than others.

3. Combined Type: Symptoms of the above two types are equally present in the person.

How Is the Diagnosis of ADHD Made?

The criteria for the diagnosis of ADHD are found in the Diagnostic and Statistical Manual of Mental Disorders, 5th Edition (DSM-5). It lists nine symptoms of inattention and nine of hyperactivity and impulsivity. To make a diagnosis of ADHD in children or adolescents, there must be six or more symptoms of either inattention or hyperactivity/impulsivity present for at least six months. Adults need five or more symptoms of inattention or hyperactivity/impulsivity. Some of the symptoms should have begun prior to age 12 years+, the symptoms should cause impaired functioning in at least two settings, and the symptoms cannot be better explained by another diagnosis. If the child has both inattention and hyperactivity-impulsivity, the diagnosis will be ADHD, combined type. If there is inattention but not hyperactivity-impulsivity, the diagnosis is ADHD, predominantly inattentive type. The inattentive type of ADHD is more commonly diagnosed in adolescents and adults with ADHD.

The diagnosis of ADHD is made by physicians—including family practitioners, pediatricians, neurologists and psychiatrists—and by psychologists. The physician can also prescribe medication if needed for treatment. The psychologist can perform further testing to determine if the person with a diagnosis of ADHD has specific learning disorders (such as in reading, writing, spelling or math) or has global intellectual disability (IQ<70). Child and adolescent psychiatrists are specialists in neurodevelopmental disorders such as ADHD and are a good resource if there are questions about the diagnosis, particularly when there may be a combination of diagnoses.

At present, there are no specific laboratory tests for the diagnosis of ADHD. The diagnosis is based on discussion with the parents, the child or the adult themselves, and other sources (such as the child's teacher), and by observation of the individual by the physician or psychologist. The evaluating professional will ask about symptoms of both ADHD and other behavioral and cognitive disorders that may accompany or mimic ADHD. Specific questionnaires, such as the Conners Parent Rating Scale and Teacher Rating Scale or the ADHD Rating Scale-IV by DuPaul et al., may be used to help with assessment and to follow response to therapy. Some professionals may use computerized tests such as the continuous performance test to look for evidence of ADHD. Neuroimaging and specialized neurophysiological tests are helping to better understand ADHD but are not used routinely for diagnosis.

Are There Other Problems That Might Look Like ADHD?

It is important to be careful in making a diagnosis of ADHD. Almost everyone has occasional problems with forgetfulness, lack of concentration or difficulty sitting still. Some children are temperamentally more active and enthusiastic than the average child. If there is no impairment, the child should not be given a diagnosis of ADHD. Also, our ability to pay attention, sit still and control our impulses changes with age and with our developmental level.

Many children with global intellectual disability (IQ<70) will be more inattentive, overactive and impulsive than others of their age. However, the behaviors may be in keeping with their developmental level. Some children with specific learning disorders (such as reading, writing, spelling, math) may appear to have trouble concentrating or paying attention because they have given up listening to lessons they do not understand or because they have to concentrate so hard to learn that their attention wears out faster. Their failure to complete tasks may be a sign of not knowing how to do the work instead of a sign of ADHD. Other children may be bored by work they have already mastered and thus appear inattentive and are disruptive. Psychoeducational testing can help to differentiate some of these presentations.

Children who are depressed or anxious may have trouble concentrating or sleeping and have agitation that results in excessive movements. Children with sleep disorders may be fatigued and drowsy during the daytime, thus appearing inattentive. Children that are living in a chaotic home and children that are being abused may look distracted and fail to concentrate on schoolwork.

Seizures can sometimes be confused with ADHD. Absence seizures and complex partial seizures may cause repeated episodes of brief loss of attention. Antiepileptic drugs may contribute to difficulties with attention or may cause hyperactivity. If a child has TSC, seizures are always a possibility. Chronic headaches from increased intracranial pressure due to a subependymal giant cell astrocytoma (SEGA) can cause lethargy and inattention and should not be confused with ADHD.

Children with TSC may have difficulty paying attention without the other behavioral problems of ADHD. We would refer to these as neuropsychological attention deficits. A very high proportion of people with TSC have some specific attention deficits, even if they don't meet criteria for ADHD. A comprehensive neuropsychological assessment by a clinical psychologist or neuropsychologist may help identify these types of problems with attention.

Are There Other Behavioral or Cognitive Problems That Accompany ADHD?

Individuals with ADHD are at an increased risk of a number of other problems. Children with ADHD have an increased risk for oppositional defiant disorder (ODD), conduct disorder, anxiety disorder, depression, bipolar disorder, Tourette's syndrome, and a range of learning disorders.

Children with ODD refuse to obey, frequently annoy others, and are often easily annoyed. They are angry children who have frequent temper tantrums. In comparison, children with conduct disorder get into trouble for major violations of the rules. They are aggressive toward others, destructive, steal and lie, are truant from school and run away from home. Approximately 30% to 40% of children with ADHD have ODD or conduct disorder. The children with both ADHD and ODD or conduct disorder are the ones at risk for developing substance abuse or delinquency.

Approximately 20% to 25% of children with ADHD will have additional problems with anxiety disorder or depression. Fearfulness or excessive worrying suggests anxiety disorder, and sadness or irritability characterizes childhood depression. Bipolar disorder is uncommon in children, but almost all the children who develop bipolar disorder before their teenage years

will have symptoms of ADHD. Grandiosity, a decreased need for sleep, hypersexuality, racing thoughts, severe irritability and rapid mood swings are symptoms of bipolar disorder.

An estimated 25-40% of children with ADHD will have specific learning disorders. In those cases, treatment of ADHD will not be sufficient to improve their academic performance. These children will require special educational programs or individualized educational plans to help them learn.

What Causes ADHD?

ADHD is a multifactorial disorder and can be caused by a range of factors including genetic and neurological problems. No one gene causes ADHD but multiple genes have been found that increase vulnerability to developing symptoms of ADHD. ADHD definitely runs in families. If a parent has ADHD, approximately half of his or her children will have symptoms of ADHD.

Some of the evidence for neurological involvement comes from imaging techniques such as PET scanning and functional MRI. These studies show that individuals with ADHD have less activity in the frontal lobes than people with no evidence of ADHD. This is consistent with psychological studies that show problems in executive functioning, or tasks that involve the frontal lobes. The executive functions are important in planning, self-control and analysis of behavior—tasks that are difficult for the child with ADHD. Recent research has shown significant difficulties with executive functioning in individuals with TSC.

How Is ADHD Treated?

The two main components of treating ADHD are non-pharmacological (including talking and behavioral treatments) and pharmacological (using medications). Usually both are required for the child with significant problems.

Non-pharmacological treatments

Behavioral or psychosocial treatments involve the child, parents and the school system. The first step is to provide psycho-education to everyone to make sure people understand what ADHD is and how ADHD affects the individual. This is an essential first step in order to come up with a plan to help the person with ADHD. The next important step is to make modifications or accommodations in the school and to train parents and families. Basic modifications in school might include reducing distractions, frequent reminders to stay on tasks, help with organization, rewards for attention and concentration and extra time for tests. Parent management training instructs the parents on establishing structure in the child's environment, using effective discipline for impulsive behaviors, rewarding attention, reducing oppositional behavior, promoting positive parent-child interaction and often relaxation techniques to help deal with the parents' stress.

If the child has alienated other people by impulsive or disruptive behaviors, social skills training may be needed. Some children will benefit from cognitive behavioral training in organization, planning and social responses. If the first modifications in the school setting are not successful, the child may need smaller classes or one-on-one instruction. An individualized educational plan (IEP) is essential for the child with severe ADHD. Many children with severe ADHD will qualify for special educational services under the Individuals with Disabilities Educational Act or section 504 of the National Rehabilitation Act.

Pharmacological treatments

Medication is helpful for the child with ADHD. Parents may worry that these drugs are addictive, but the truth is that appropriate use of stimulants may help prevent later substance abuse. Parents also expect that their child will be off medication by adolescence. This may happen, but some teenagers and adults may find that stimulants continue to be beneficial and that stopping medication leads to difficulties at school or work. Finally, the response to medication cannot be used to determine the diagnosis. Children with ADHD and children without ADHD may have an increase in attention span on stimulants.

Stimulant medications are the first-line drugs used to treat ADHD. Examples include methylphenidate (Ritalin, Concerta, Focalin) and dextroamphetamine (Adderall, Dexedrine, Vyvanse). These drugs work in almost 80 percent of children with ADHD. The medication takes effect within 30 minutes. The duration varies by the medication used from an average of four hours for regular Ritalin or Dexedrine, six hours for Adderall, to 10 to 12 hours for Concerta, Focalin XR, Adderall XR, and Vyvanse. The main side effects are decreased appetite and trouble sleeping. Some children complain of headaches, stomachaches, depression or irritability.

If the stimulants are not effective or cause unacceptable side effects, second-line treatments including atomoxetine (Strattera) and antidepressants, including the tricyclic antidepressants (desipramine, nortriptyline) and bupropion (Wellbutrin), can be used. Atomoxetine and the antidepressants are effective in 60% to 70% of children with ADHD. Atomoxetine may cause upset stomach and lethargy. Tricyclic antidepressants may cause dry mouth, constipation, lethargy, increased heart rate and increased blood pressure. Bupropion may cause the same side effects as both the stimulants and the tricyclic antidepressants. In high doses, bupropion has caused seizures. Clonidine and guanfacine have been used to reduce hyperactivity and to promote sleep, but they are less effective for improving attention. The main side effects are lethargy, fatigue, and decreased blood pressure.

For the child with TSC and ADHD, extra care must be taken in choosing a medication. Bupropion can lower the seizure threshold. There is a minimal risk of increase in number of seizures with the tricyclic antidepressants. Stimulants do not seem to cause seizures as long as the dose is appropriate. If there is a history of heart problems in TSC, tricyclic antidepressants and clonidine must be used cautiously. The tricyclics can affect cardiac conduction and clonidine can lower the blood pressure.

Do Children Outgrow ADHD?

In the past, we thought ADHD disappeared during adolescence. Better follow-up studies have disproven this. Though 30% to 40% of children with ADHD will improve significantly, approximately 50% to 60% will continue to have some symptoms of ADHD, and an additional 10% to 15% will have more severe problems. Adults with ADHD do not have the obvious hyperactivity seen with childhood ADHD; however, they may have restlessness, distractibility and disorganization. They will respond to many of the same therapies used successfully to treat childhood ADHD.

TSC and ADHD

The symptoms of ADHD and the criteria for the diagnosis of ADHD are the same for children with TSC and children unaffected by TSC. The symptoms of ADHD are more common in children with TSC. Inattention in children with TSC may be caused by seizures, central nervous system tumors and hydrocephalus, by cortical tubers, or by white matter abnormalities disrupting CNS pathways involved in attention. It is even possible that the molecular abnormalities caused by TSC could be enough to cause some attention problems. Children with TSC and epilepsy are at an increased risk for autism spectrum disorder. Autism spectrum disorder may cause hyperactivity and inattention, and some children may have both autism and ADHD.

As listed above, there are a number of factors to consider with ADHD in TSC. Seizures may make a person appear inattentive, but seizures can also increase the likelihood of ADHD. Headaches associated with SEGAs may cause inattention and concentration problems that could be confused with ADHD. Also, many people with TSC have neuropsychological attention deficits even when they don't appear from the outside to have obvious ADHD symptoms.

Children with ADHD and TSC are also at risk for other problems. Both global intellectual disability (IQ<70) and specific learning disorders (reading, writing, spelling, math) are more common in the child with TSC. Anxiety and depression may accompany many chronic illnesses and may be more common in children with TSC than unaffected children.

The treatment of ADHD is the same in children with or without TSC, with certain cautions. Drugs that lower the seizure threshold and drugs that affect the heart must be carefully monitored. Unfortunately, children with both ADHD and epilepsy do not respond as well to medication for ADHD as do children with ADHD alone. When treatments don't seem to be working well, it is important to consider re-evaluating the person with TSC taking into consideration the physical, neurological, cognitive, educational and social factors.

Resources

Centers for Disease Control and Prevention
Attention-Deficit/Hyperactivity Disorder Information:
<http://www.cdc.gov/ncbddd/adhd/index.html>

CDC ADHD Fact Sheet:
http://www.cdc.gov/ncbddd/actearly/pdf/parents_pdfs/ADHDFactSheet.pdf

Children and Adults with Attention Deficit/Hyperactivity Disorder (CHADD)
8181 Professional Place - Suite 150
Landover, MD 20785
Tel: 301-306-7070
Fax: 301-306-7090
Website: www.chadd.org

National Resource Center on AD/HD
Phone: 1-800-233-4050
Website: www.help4adhd.org

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***This publication from the Tuberous Sclerosis Alliance is intended to provide basic information about tuberous sclerosis complex (TSC). It is not intended to, nor does it, constitute medical or other advice. Readers are warned not to take any action with regard to medical treatment without first consulting a health care provider. The TS Alliance does not promote or recommend any treatment, therapy, institution or health care plan.*

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