American Academy of Special Education Professionals

AASEP

STAFF DEVELOPMENT in SPECIAL EDUCATION

Tourette Syndrome

Course Objectives

- To provide a definition of Tourette syndrome
- To present an overview of the symptoms of Tourette syndrome
- To discuss the course of Tourette syndrome
- To determine whether people with Tourette syndrome can control their tics
- To discuss the possible causes of Tourette syndrome
- To discuss disorders associated with Tourette syndrome
- To examine how Tourette syndrome is diagnosed
- To examine how Tourette syndrome is treated
- To determine whether Tourette syndrome is inherited
- To discuss the prognosis of Tourette syndrome
- To examine appropriate educational settings for children with Tourette syndrome

Definition of Tourette syndrome

Tourette syndrome (TS) is a neurological disorder characterized by repetitive, stereotyped, involuntary movements and vocalizations called tics. The disorder is named for Dr. Georges Gilles de la Tourette, the pioneering French neurologist who in 1885 first described the condition in an 86-year-old French noblewoman.

The early symptoms of TS are almost always noticed first in childhood, with the average onset between the ages of 7 and 10 years. TS occurs in people from all ethnic groups; males are affected about three to four times more often than females. It is estimated that 200,000 Americans have the most severe form of TS, and as many as one in 100 exhibit milder and less complex symptoms such as chronic motor or vocal tics or transient tics of childhood. Although TS can be a chronic condition with symptoms lasting a lifetime, most people with the condition experience their worst symptoms in their early teens, with improvement occurring in the late teens and continuing into adulthood.

Symptoms of Tourette syndrome

Tics are classified as either simple or complex.

Simple Motor Tics

Simple motor tics are sudden, brief, repetitive movements that involve a limited number of muscle groups. Some of the more common simple tics include eye blinking and other vision irregularities, facial grimacing, shoulder shrugging, and head or shoulder jerking. Simple vocalizations might include repetitive throat-clearing, sniffing, or grunting sounds.

Complex Tics

Complex tics are distinct, coordinated patterns of movements involving several muscle groups. Complex motor tics might include facial grimacing combined with a head twist and a shoulder shrug. Other complex motor tics may actually appear purposeful, including sniffing or touching objects, hopping, jumping, bending, or twisting. Simple vocal tics may include throat-clearing, sniffing/snorting, grunting, or barking. More complex vocal tics include words or phrases.

Other Symptoms of TS

Perhaps the most dramatic and disabling tics include motor movements that result in self-harm such as punching oneself in the face or vocal tics including coprolalia (uttering swear words) or echolalia (repeating the words or phrases of others). Some tics are preceded by an urge or sensation in the affected muscle group, commonly called a premonitory urge. Some with TS will describe a need to complete a tic in a certain way or a certain number of times in order to relieve the urge or decrease the sensation.
Tics are often worse with excitement or anxiety and better during calm, focused activities. Certain physical experiences can trigger or worsen tics, for example tight collars may trigger neck tics, or hearing another person sniff or throat-clear may trigger similar sounds. Tics do not go away during sleep but are often significantly diminished.

The Course of TS

Tics come and go over time, varying in type, frequency, location, and severity. The first symptoms usually occur in the head and neck area and may progress to include muscles of the trunk and extremities. Motor tics generally precede the development of vocal tics and simple tics often precede complex tics. Most patients experience peak tic severity before the mid-teen years with improvement for the majority of patients in the late teen years and early adulthood. Approximately 10 percent of those affected have a progressive or disabling course that lasts into adulthood.

Control of Tics

Although the symptoms of TS are involuntary, some people can sometimes suppress, camouflage, or otherwise manage their tics in an effort to minimize their impact on functioning. However, people with TS often report a substantial buildup in tension when suppressing their tics to the point where they feel that the tic must be expressed. Tics in response to an environmental trigger can appear to be voluntary or purposeful but are not.

Causes of TS

Although the cause of TS is unknown, current research points to abnormalities in certain brain regions (including the basal ganglia, frontal lobes, and cortex), the circuits that interconnect these regions, and the neurotransmitters (dopamine, serotonin, and norepinephrine) responsible for communication among nerve cells. Given the often complex presentation of TS, the cause of the disorder is likely to be equally complex.

Disorders are associated with TS

Many with TS experience additional neurobehavioral problems including inattention; hyperactivity and impulsivity (attention deficit hyperactivity disorder—ADHD) and related problems with reading, writing, and arithmetic; and obsessive-compulsive symptoms such as intrusive thoughts/worries and repetitive behaviors. For example, worries about dirt and germs may be associated with repetitive hand-washing, and concerns about bad things happening may be associated with ritualistic behaviors such as counting, repeating, or ordering and arranging.

People with TS have also reported problems with depression or anxiety disorders, as well as other difficulties with living, that may or may not be directly related to TS. Given the range of potential complications, people with TS are best served by receiving medical care that provides a comprehensive treatment plan.

Diagnosis of TS

TS is a diagnosis that doctors make after verifying that the patient has had both motor and vocal tics for at least 1 year. The existence of other neurological or psychiatric conditions can also help doctors arrive at a diagnosis. Common tics are not often misdiagnosed by knowledgeable clinicians. But atypical symptoms or atypical presentation (for example, onset of symptoms in adulthood) may require specific specialty expertise for diagnosis. These include childhood-onset involuntary movement disorders such as dystonia, or psychiatric disorders characterized by repetitive behaviors/movements (for example, stereotypic behaviors in autism and compulsive behaviors in obsessive-compulsive disorder — OCD).

There are no blood or laboratory tests needed for diagnosis, but neuroimaging studies, such as magnetic resonance imaging (MRI), computerized tomography (CT), and electroencephalogram (EEG) scans, or certain blood tests may be used to rule out other conditions that might be confused with TS.

It is not uncommon for patients to obtain a formal diagnosis of TS only after symptoms have been present for some time. The reasons for this are many. For families and physicians unfamiliar with TS, mild and even moderate tic symptoms may be considered inconsequential, part of a developmental phase, or the result of another condition. For example, parents may think that eye blinking is related to vision problems or that sniffing is related to seasonal allergies. Many patients are self-diagnosed after they, their parents, other relatives, or friends read or hear about TS from others.

Treatment of TS

Neuroleptic Medications

Because tic symptoms do not often cause impairment, the majority of people with TS require no medication for tic suppression. However, effective medications are available for those whose symptoms interfere with functioning. Neuroleptics are the most consistently useful medications for tic suppression; a number are available but some are more effective than others (for example, haloperidol and pimozide).

Unfortunately, there is no one medication that is helpful to all people with TS, nor does any medication completely eliminate symptoms. In addition, all medications have side effects. Most neuroleptic side effects can be managed by initiating treatment slowly and reducing the dose when side effects occur. The most common side effects of neuroleptics include sedation, weight gain, and cognitive dulling. Neurological side effects such as tremor, dystonic reactions (twisting movements or postures), parkinsonian-like symptoms, and other dyskinetic (involuntary) movements are less common and are readily managed with dose reduction.

Discontinuing neuroleptics after long-term use must be done slowly to avoid rebound increases in tics and withdrawal dyskinesias. One form of withdrawal dyskinesia called tardive dykinesia is a movement disorder distinct from TS that may result from the chronic use of neuroleptics. The risk of this side effect can be reduced by using lower doses of neuroleptics for shorter periods of time.

Other Medications
Other medications may also be useful for reducing tic severity, but most have not been as extensively studied or shown to be as consistently useful as neuroleptics. Additional medications with demonstrated efficacy include alpha-adrenergic agonists such as clonidine and guanfacine. These medications are used primarily for hypertension but are also used in the treatment of tics. The most common side effect from these medications that precludes their use is sedation.

Effective medications are also available to treat some of the associated neurobehavioral disorders that can occur in patients with TS. Recent research shows that stimulant medications such as methylphenidate and dextroamphetamine can lessen ADHD symptoms in people with TS without causing tics to become more severe. However, the product labeling for stimulants currently contraindicates the use of these drugs in children with tics/TS and those with a family history of tics. Scientists hope that future studies will include a thorough discussion of the risks and benefits of stimulants in those with TS or a family history of TS and will clarify this issue. For obsessive-compulsive symptoms that significantly disrupt daily functioning, the serotonin reuptake inhibitors (clomipramine, fluoxetine, fluvoxamine, paroxetine, and sertraline) have been proven effective in some patients.

Psychotherapy
Psychotherapy may also be helpful. Although psychological problems do not cause TS, such problems may result from TS. Psychotherapy can help the person with TS better cope with the disorder and deal with the secondary social and emotional problems that sometimes occur. More recently, specific behavioral treatments that include awareness training and competing response training, such as voluntarily moving in response to a premonitory urge, have shown effectiveness in small controlled trials.

Research on the Inheritance of TS
Evidence from twin and family studies suggests that TS is an inherited disorder. Although early family studies suggested an autosomal dominant mode of inheritance (an autosomal dominant disorder is one in which only one copy of the defective gene, inherited from one parent, is necessary to produce the disorder), more recent studies suggest that the pattern of inheritance is much more complex.

Although there may be a few genes with substantial effects, it is also possible that many genes with smaller effects and environmental factors may play a role in the development of TS. Genetic studies also suggest that some forms of ADHD and OCD are genetically related to TS, but there is less evidence for a genetic relationship between TS and other neurobehavioral problems that commonly co-occur with TS. It is important for families to understand that genetic predisposition may not necessarily result in full-blown TS; instead, it may express itself as a milder tic disorder or as obsessive-compulsive behaviors. It is also possible that the gene-carrying offspring will not develop any TS symptoms.

The sex of the person also plays an important role in TS gene expression. At-risk males are more likely to have tics and at-risk females are more likely to have obsessive-compulsive symptoms.

People with TS may have genetic risks for other neurobehavioral disorders such as depression or substance abuse. Genetic counseling of individuals with TS should include a full review of all potentially hereditary conditions in the family.

Prognosis for Individuals with TS

Although there is no cure for TS, the condition in many individuals improves in the late teens and early 20s. As a result, some may actually become symptom-free or no longer need medication for tic suppression. Although the disorder is generally lifelong and chronic, it is not a degenerative condition. Individuals with TS have a normal life expectancy. TS does not impair intelligence. Although tic symptoms tend to decrease with age, it is possible that neurobehavioral disorders such as depression, panic attacks, mood swings, and antisocial behaviors can persist and cause impairment in adult life.

Educational Settings for Children with TS

Although students with TS often function well in the regular classroom, ADHD, learning disabilities, obsessive-compulsive symptoms, and frequent tics can greatly interfere with academic performance or social adjustment. After a comprehensive assessment, students should be placed in an educational setting that meets their individual needs. Students may require tutoring, smaller or special classes, and in some cases special schools.

All students with TS need a tolerant and compassionate setting that both encourages them to work to their full potential and is flexible enough to accommodate their special needs. This setting may include a private study area, exams outside the regular classroom, or even oral exams when the child's symptoms interfere with his or her ability to write. Untimed testing reduces stress for students with TS.