Patient Handouts

Tourette Syndrome

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What is 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.

What are the symptoms?

Tics are classified as either simple or complex. 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 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. 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.

What is 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.

Can people with TS control their 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.

What causes 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.

What disorders are associated with TS?

Many individuals 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.

How is TS diagnosed?

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[1] 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. 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 when the history or clinical examination is atypical.

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.

How is TS treated?

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 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. However, given the lower side effect risk assocaited with these medications, they are often used as first-line agents before proceeding to treatment with neuroleptics.

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.

Behavioral treatments such as awareness training and competing response training can also be used to reduce tics. A recent NIH-funded, multi-center randomized control trial called Cognitive Behavioral Intervention for Tics, or CBIT, showed that training to voluntarily move in response to a premonitory urge can reduce tic symptoms. Other behavioral therapies, such as biofeedback or supportive therapy, have not been shown to reduce tic symptoms. However, supportive therapy can help a person with TS better cope with the disorder and deal with the secondary social and emotional problems that sometimes occur

Is TS inherited?

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 gender 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.

Genetic counseling of individuals with TS should include a full review of all potentially hereditary conditions in the family.

What is the prognosis?

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.

What is the best educational setting for children with Tourette syndrome?

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.

What research is being done?

Within the Federal government, the leading supporter of research on TS and other neurological disorders is the National Institute of Neurological Disorders and Stroke (NINDS). The NINDS, a part of the National Institutes of Health (NIH), is responsible for supporting and conducting research on the brain and central nervous system. The NINDS and other NIH components, such as the National Institute of Mental Health, the Eunice Kennedy Shriver National Institute of Child Health and Human Development, the National Institute on Drug Abuse, and the National Institute on Deafness and Other Communication Disorders, support research of relevance to TS, either at NIH laboratories or through grants to major research institutions across the country. Another component of the Department of Health and Human Services, the Centers for Disease Control and Prevention, funds professional education programs as well as TS research.

Knowledge about TS comes from studies across a number of medical and scientific disciplines, including genetics, neuroimaging, neuropathology, clinical trials (medication and non-medication), epidemiology, neurophysiology, neuroimmunology, and descriptive/diagnostic clinical science.

Genetic studies. Currently, NIH-funded investigators are conducting a variety of large-scale genetic studies. Rapid advances in the technology of gene finding will allow for genome-wide screening approaches in TS, and finding a gene or genes for TS would be a major step toward understanding genetic risk factors. In addition, understanding the genetics of TS genes will strengthen clinical diagnosis, improve genetic counseling, lead to the clarification of pathophysiology, and provide clues for more effective therapies.

Neuroimaging studies. Within the past 5 years, advances in imaging technology and an increase in trained investigators have led to an increasing use of novel and powerful techniques to identify brain regions, circuitry, and neurochemical factors important in TS and related conditions.

Neuropathology. Within the past 5 years, there has been an increase in the number and quality of donated postmortem brains from TS patients available for research purposes. This increase, coupled with advances in neuropathological techniques, has led to initial findings with implications for neuroimaging studies and animal models of TS.

Clinical trials. A number of clinical trials in TS have recently been completed or are currently underway. These include studies of stimulant treatment of ADHD in TS and behavioral treatments for reducing tic severity in children and adults. Smaller trials of novel approaches to treatment such as dopamine agonist and GABAergic medications also show promise.

Epidemiology and clinical science. Careful epidemiological studies now estimate the prevalence of TS to be substantially higher than previously thought with a wider range of clinical severity. Furthermore, clinical studies are providing new findings regarding TS and co-existing conditions. These include subtyping studies of TS and OCD, an examination of the link between ADHD and learning problems in children with TS, a new appreciation of sensory tics, and the role of co-existing disorders in rage attacks. One of the most important and controversial areas of TS science involves the relationship between TS and autoimmune brain injury associated with group A beta-hemolytic streptococcal infections or other infectious processes. There are a number of epidemiological and clinical investigations currently underway in this intriguing area.

Where can I go for more information?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN
P.O. Box 5801
Bethesda, MD 20824
(800) 352-9424
http://www.ninds.nih.gov

Information also is available from the following organizations:

Tourette Syndrome Association
42-40 Bell Boulevard, Suite 205
Bayside, NY 11361-2820
(718) 224-2999, (888) 4-TOURET (486-8738)
http://www.tsa-usa.org

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National Institute of Neurological Disorders and Stroke. Tourette Syndrome Fact Sheet. Available at: http://www.ninds.nih.gov/disorders/tourette/detail_tourette.htm. Accessed January 16, 2014.

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