Tourette's Disorder

Gilles de la Tourette Syndrome and Tic Disorders

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The Harvard Medical School Mental Health Letter, May 1989

The characteristic symptoms of Gilles de la Tourette syndrome (abbreviated Tourette syndrome or TS) are tics: abrupt, brief, rapid, recurrent, involuntary movements and vocal sounds that occur at irregular intervals. Our remarks on this disorder are based on a detailed study of 666 patients with TS and clinical experience with over 2,000 other patients suffering from tics.

Tics may be simple or complex. Simple motor tics consist of isolated muscle contractions such as a jerking movement of the arm. Complex motor tics involve several coordinated muscle movements; for example, a person may twirl or do deep knee bends while walking. Simple vocal tics include grunts, barks, other inarticulate noises, stuttering, stammering, and word garbling, as well as unusual emphasis in speech (altered volume, pitch, or clarity of certain words or phrases). Complex vocal tics include coprolalia (socially unacceptable or obscene language or sounds), echolalia (repetition of the previous speaker's last sound, word, or sentence), and palilalia (repetition of one's own last sound, word, or sentence). Complex motor tics include copropraxia (socially unacceptable or obscene gestures) and echopraxia (imitating or echoing the movements of others).

Tics become more severe in the evening and in the presence of family or close friends. Anxiety, fatigue, pleasurable anticipation, and television watching also make them worse. Tics are reduced by absorption in a task. They also decrease in the morning, at work, in school, and in the presence of strangers; they usually disappear in sleep.

TS is diagnosed if a patient has multiple motor tics and at least one vocal tic beginning before age 21, lasting more than a year, and spontaneously varying in severity and type. The symptoms first appear at an average age of six or seven; in 90 percent of cases the disorder begins before age ten. Up to 82 different symptoms may appear in the first week. In our study, the most common of these were eye movements, inarticulate vocal sounds, and head or neck motions. Other common initial tics are throat clearing, grimaces, shrugging, sniffing, lip and mouth motions, hand, finger, and arm movements, and motions of the torso. Early symptoms may be misdiagnosed because they have many other causes; for example, throat clearing, sniffing, coughing, snorting, hissing, and noisy breathing are often mistaken for allergy symptoms.

A third of the patients we studied developed two to ten symptoms during their lives; 44 percent had 11 to 20, and the rest had 21 or more. The largest number of different tics we found in any one patient was 60. Eighty percent developed the most common simple motor tic, an eye movement. The other common simple motor tics, in descending order of frequency, were horizontal neck movements, shoulder shrugs, vertical head movements, arm movements, grimaces, mouth opening, hand or finger motions, leg movements, lip motions, and movements of the torso. Other types of simple motor tics affected fewer than 20 percent of patients.

More than two-thirds of the patients had one or more of 54 complex motor tics. The most common,

in descending order, were hitting themselves, jumping, touching themselves, touching others, making obscene gestures, smelling their hands, and smelling other objects. Patients had 77 different vocal tics; the most common, in order, were throat clearing, grunts, squeals, shrieks, yelps, and other high-pitched cries, sniffing, coughing, screaming, and snorting.

In a relatively unfamiliar type of TS, the symptoms are sensations rather than (or in addition to) movements and sounds. Patients have annoying feelings of heaviness, lightness, tickling, heat and cold in their joints, bones, muscles, and other parts of the body. Squeezing the muscles brings only temporary relief.

Outcome Uncertain

It is impossible to predict the outcome of TS. The frequency, types, and severity of tics often change. Twenty-seven percent of TS patients lose their tics for periods from a week to seven years; others suffer all their lives. Five to eight percent recover completely and permanently, usually during puberty or adolescence. Symptoms become less severe (sometimes much less severe) in about 35 percent of patients during adolescence and in most patients when they become adults. Yet a few people whose early symptoms are mild develop severe tics in their 20s or early 30s.

TS was once thought to cause insanity, but it does not. Patients suffer no intellectual deterioration or psychosis and they do not require hospitalization. Nor is TS associated with any particular character trait or personality type or any other physical or psychiatric illness. The symptoms ordinarily do not interfere with physical activity or with work, school, and recreation, but they may be severe enough to affect social functioning and emotional health, especially when the patient lacks resources for coping with them.

Until recently TS was considered rare. Now diagnosis has become more accurate and the disorder is often detected early, so the number of reported cases has increased. A conservative estimate is that one in every 200 persons has TS. The rate does not vary with race, nationality, social class, or the general medical and psychiatric history of the patient or the patient's family. The only factors known to make a difference are sex (about 75 percent of patients with TS are male) and a family history of tic disorders.

TS is the most serious of several tic disorders. A chronic disorder with only motor tics is twice as common as TS, and transient symptoms lasting less than a year may arise in as much as 25 percent of the population. All tic disorders seem to occur at about the same rate in all cultures. All begin at about the same age and respond to treatment in the same way. They probably result from a single disease process of which TS is the most severe form.

Causes of TS

Some forms of TS and chronic motor tic disorder seem to be inherited. Eight percent of patients in our sample had one or more family members with TS; 47 percent had one or more family members with some tic disorder. The concordance rate for these disorders in monozygotic (genetically identical) twins is very high; if one twin has a tic disorder, there is about an 80 percent chance that the other will have it too. In dizygotic (fraternal) twins of the same sex, the concordance rate is only 25 percent. Chronic tic disorders are probably transmitted by an autosomal dominant gene; this means that a child has a 50 percent chance of inheriting the disorder if one parent carries the necessary genetic material. But the expression of the gene is apparently influenced by sex: tics are more common in the families of female TS patients than in those of male TS patients.

TS was once attributed to unconscious emotional conflicts derived from childhood experience. In the 1960s, neuroleptic drugs were found to be an effective treatment, and TS is now thought to be an organic neurological disorder. The cause could be hypersensitivity of certain receptors in the brain for the neurotransmitter dopamine. Neuroleptics block dopamine receptors, and drugs that enhance the effects of dopamine make the symptoms of TS worse. The drugs may act on the basal ganglia, a brain region involved in the regulation of body movements. This theory has not been proved experimentally, and it may be too simple to explain TS fully.

One controversial issue in the study of TS is its relationship to the symptoms of impulsiveness, restlessness, and short attention span that characterize attention deficit hyperactivity disorder (ADHD). A very large proportion of patients with TS are reported to have ADHD -- as high as 60 percent among those seen by specialists. But this may be an illusion; patients with more severe symptoms, or those who happen to have both TS and ADHD, are more likely to be diagnosed and referred to specialists and especially to research centers. When milder cases of TS are considered, the percentage who also have ADHD drops considerably.

There has been considerable confusion about this. Much behavior regarded as a sign of TS is in fact usually a sign of ADHD. People with ADHD often have abnormal neuropsychological tests and abnormal EEGs; they often suffer from conduct disorders and other psychopathology. People with TS alone do not have these problems.

Some researchers have found a relationship between TS and obsessive-compulsive disorder -- unwanted intrusive thoughts creating tension that is relieved by meaningless repetitive acts like handwashing, counting, and checking. But many of these studies are poorly controlled and otherwise inadequate; for example, if complex tics are not distinguished from compulsive actions, TS and obsessive-compulsive disorder will seem closely related almost by definition. More careful research on this subject is needed -- especially random surveys of the general population to determine whether obsessive-compulsive disorder and TS occur together at a rate higher than chance. Our own clinical observations and controlled studies suggest that obsessive-compulsive disorder is not associated with TS.

Treatment

Treatment of TS and other tic disorders is needed only if the symptoms interfere with psychosocial, academic, and vocational functioning. Because the progress of the disorder varies so much and temporary or permanent spontaneous remissions are so common, carefully controlled experiments are needed to evaluate treatments. Thirty to 50 percent of patients will have a placebo response in most studies. The drug most commonly used in treating TS is haloperidol (Haldol ®), although other neuroleptics such as pimozide (Orap ®) and fluphenazine (Prolixin ®) are also effective at low doses. The aim is to reduce tics at least 70 percent while the patient is in school, at work, among strangers, or in social situations. The more severe symptoms that occur at home and in the evening can be controlled by increasing the dose, although that may also heighten side effects.

The most common side effects are extrapyramidal symptoms -- muscle spasms, restless pacing and fidgeting, tremors in the hands and feet, slowing of body movements, and abnormal posture. Other potential adverse effects are irritability, listlessness, depression, loss of motivation, and cognitive impairment. Adverse effects are usually most severe in the first three months. None of our patients has developed tardive dyskinesia, the chronic disorder of body movement sometimes produced by

neuroleptic drugs.

Lowering the dose is the most obvious way to reduce side effects. Extrapyramidal symptoms can be treated with antiparkinsonian drugs, and other symptoms sometimes respond to stimulant or antianxiety drugs. Although many clinicians and researchers disagree, we do not believe that amphetamines and other stimulants cause TS or make it worse. In fact, we find that a stimulant used to treat hyperactive patients who also have TS often improves their tics and at the same time reduces the adverse effects of neuroleptics. We often prescribe stimulants when patients become lethargic and depressed, gain weight, or develop phobias on low doses of neuroleptics.

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