

**TIC DISORDERS**

**INCLUDING**

**TOURETTE**

**SYNDROME**

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<p><b>Material in this handout is taken from various publications from the Tourette Syndrome Association.</b></p>
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## TOURETTE SYNDROME

This information has been prepared specifically for the patient, parent, friend, relative, or teacher, who has recently encountered one of the tic disorders including Tourette syndrome (TS). You can deal with these conditions best if you know as much as possible about its cause (or causes), its symptoms, the types of treatment available and what to expect in the future. In this discussion the term Tourette syndrome is used to cover all tic disorders.

### HISTORY

In 1885, a French doctor named George Gilles de la Tourette described nine patients with the syndrome that now bears his name. The first reported case of Tourette syndrome was by another French doctor named Itard in 1825. He described the case of Marquise de Dampierre, a woman of nobility who was quite an important person in her time. The Marquise had had symptoms consisting of tics involving many parts of her body, plus coprolalia (obscene speech) and echolalia (echo speech) from the age of seven. These symptoms would appear explosively so as to interrupt her in the middle of a conversation. The obscene words and movements were very much unlike her usual character. As Itard described it:

"The more she herself thinks her vulgarities will be revolting, the more she is tortured by the fear that she will utter them, and it is precisely this preoccupation, that when she can no longer control it, puts these words at the tip of her tongue."

Because of the social problems these symptoms caused, this lady lived the last 70 years of her life as a recluse. She lived to be over 90 years old and was one of the patients that Gilles de la Tourette saw some 60 years after Itard.

### WHAT IS IT?

Gilles de la Tourette syndrome is a neurological disorder which becomes evident in early childhood or adolescence between the ages of 2 and 15. Tourette syndrome is defined by multiple motor and vocal tics lasting for more than one year. Many people have only motor tics or only vocal tics. The first symptoms usually are involuntary movements (tics) of the face, arms, limbs or trunk. These tics are frequent, repetitive and rapid. The most common first symptom is a facial tic (eye blink, nose twitch, grimace), and is replaced or added to by other tics of the neck, trunk, and limbs.

These involuntary (outside the patient's control) tics may also be complicated, involving the entire body, such as kicking and stamping. Other symptoms such as touching, repetitive thoughts and movements and compulsions can occur.

There are also verbal tics. These verbal tics (vocalizations) usually occur with the movements; later they may replace one or more motor tics. These vocalizations include grunting, throat clearing, shouting and barking. The verbal tics may also be expressed as coprolalia (the involuntary use of obscene words).

Neither echolalia (echo speech) or coprolalia is necessary for the diagnosis of Tourette syndrome. All patients have involuntary movements and some have vocalizations. Echo phenomena are also reported, although less frequently. These may include repeating word of others (echolalia), repeating ones own words, and repeating movements of others.

### THE TYPICAL CASE

The first symptom that appears is usually a tic on a small area of the face, usually around the eyes or mouth. A single simple tic is not an unusual occurrence and it may occur in many people, adults as well as children. Doctors familiar with tic problems state that 20 to 25% of the population have tics at some point during their lives. In most people, tics are temporary and disappear after a short time. With Tourette syndrome the tics remain and are gradually accompanied by other tics involving other muscles of the face, neck, arms, legs and trunk. Vocal tics are always present with Tourette syndrome.. As time passes, tics frequently increase in number so that many sites may show rapid muscular spasms at the same time. Tics can wax and wane in severity and have remissions and exacerbation. It is important to remember that the frequency and intensity of the tics can vary greatly from one patient to another and also in the same patient at different times. Severity of tics may impact on the patient's daily functioning and efforts to overcome the tics.

In most patients emotional stress, tension, anxiety or fatigue may cause an increase of symptoms. Concentration on a non-anxious task usually results in a decrease of symptoms. Tics usually do not occur during sleep or are greatly reduced.

After muscular tics have been present for some time, the patient is noted to make a soft, unintelligible noise that appears to be involuntary. This may take the form of sniffing, throat-clearing, hiccuping, grunting, barking, a nonsense sound or a specific word or phrase. The term "vocalization" is used to describe all these sounds. Usually the sound becomes louder and more distinct, and in some cases, the sound that was at first unintelligible is now clearly an obscenity.

The word "explosive" describes how the vocalizations are emitted. Vocalizations usually become worse under the same circumstances (tension, stress or fatigue) that cause muscular tics to be more pronounced. Many patients do not progress to the point of explosive outbursts, and some may conceal an obscene utterance by changing it slightly when speaking.

Coprolalia (involuntary utterance of obscene words) and echolalia are present in less than 10% of patients.

All patients do not follow the previously described "typical case". In some patients vocalizations come first and in others muscular tics and vocalizations begin at the same time. There is also a great variation in the time required for different aspects of the symptoms to appear. Several symptoms may alert at the same time or it may be several years between the onset of one symptom and the onset of others.

Other abnormal movements or behavior patterns can also occur. Some of these include stuttering, sticking out the tongue constantly, pounding the chest, stomach or head with the hand or fist, grinding the teeth, handling the genitals constantly, abnormal walking, temper tantrums, echopraxia (imitating the movements of others), or compulsive touching. These are examples given to point out the wide variety of symptoms that may appear and to make it clear that additional symptoms are common with all tic disorders including Tourette syndrome.

## CAUSE

The precise cause has not been definitely established. Researchers feel it is due to a biochemical imbalance in the part of the brain called the basal ganglia. Basal ganglia sends pathways to the cortex of the brain; these pathways influence and control motor movements of the body. Tic disorders appear to involve the chemical neurotransmitters dopamine and serotonin and their effect on these parts of the central nervous system. This is why dopamine-blocking agents, such as haloperidol, pimozide and Risperidone are being used successfully in treating TS.

## OTHER FACTS

Tics disappear during sleep and during intense sexual arousal. The number of boys with tic disorder outnumbers girls by at least three to one (no known reason). Some patients can voluntarily inhibit symptoms for a period of time (depending on the patient) for just a moment to as long as several hours. This voluntary effort may increase tension which ultimately requires discharge of tic symptoms.

There have been studies to determine the nature of the heredity of Tourette syndrome. Perhaps it is an autosomal dominant gene with high sex-specific (male>female) penetrance. This means that an individual would have to have only one gene (inherited from either parent) in order to be susceptible to Tourette syndrome. These studies have been inconclusive. Other researchers feel that several genes working together may be responsible. Some authorities feel that some varieties of the disorder may not be genetic. As many as 10% of TS cases occur in families with no known previous cases.

Normally some symptoms remain throughout life. There is usually a waxing and waning of symptoms. Symptoms frequently decrease through adolescence.

Tourette syndrome is:

- Not mentally or physically degenerative
- Not physically crippling
- Not physically disabling
- Not contagious
- Not infectious

It does not diminish intellectual capacity. It is not terminal. No one ever dies from Tourette syndrome.

There is no diagnostic test to confirm a patient has Tourette syndrome. A diagnosis is made if the following conditions are present:

1. Age for onset between 2 and 15 years.
2. Multiple involuntary muscular and verbal tics (in younger patients verbal tics may not have developed yet).
3. Tic severity and frequency wax and wane (increase and decrease), but are present for more than one year.

The following are confirmatory but are not essential for the diagnosis:

1. Coprolalia (involuntary use of obscene words).
2. Copropraxia (involuntary obscene gesturing).
3. Echolalia (involuntary repetition of sounds from self or others).
4. Echopraxia (involuntary imitation of the movements of others).

There are certain behavioral problems noted to be associated with TS; i.e. obsessive-compulsive disorder (OCD), attention-deficit hyperactivity disorder (ADHD), and oppositional defiant disorder (ODD).

Some consider Tourette syndrome to be one of the several tic disorders. These are described as:

1. Acute or transient tic of childhood that begins usually before age of 12, involves one muscle group only, often an eye blink and stops within one year.
2. Chronic simple tic usually begins before the age of 15, involves one or two muscle groups. Tics persist throughout life but there is little or no change in the pattern of tics and generally consists of either motor or vocal tics but not both. Chronic motor tics and Tourette syndrome are probably different manifestations of an autosomal dominant gene with high penetrance.

## TREATMENT

Tourette syndrome is not as rare as was originally thought, although it may not be encountered by most doctors. Few disorders have had as many varied types of treatments. If you can name a treatment, some Tourette patient has probably had it prescribed as a cure for his condition. When tics first appear in a child the most common advice is, "ignore it and it will disappear." If the patient has a tic disorder or Tourette syndrome the tics may not go away or "disappear."

Another common incorrect diagnosis for the patient is psychological conflict over inhibited aggression. Common treatments (all of which are ineffective in the vast majority of cases) are: psychotherapy, group therapy, relaxation, hypnosis, bio-feedback, behavior modification, chiropractic, a variety of vitamin diets, herbs, trace elements, herbs, and many drugs.

There are medications that will reduce tics; none are a cure. Haloperidol (Haldol) is effective for 60% to 90% of patients. Other medications in this group called neuroleptic drugs that have the same effect on tics are pimozide (Orap), risperidone (Risperdal), olanzapine (Zyprexa), and ziprasidone (Geodon). Response to any one of these drugs is quite variable from patient to patient.

Drug dosage that patients require can be quite variable even for children of similar age and size. There is no one set dose for an individual. The medication is started at a low dose and must be slowly increased until the tics are reduced or until side effects are encountered. The usual side effects are reversible; they disappear when the medication is decreased or discontinued.

Most patients do not obtain 100% relief of symptoms (tics and vocalizations) with medication. Many patients will experience a 90% reduction in tics; other 50-60 %.

If the patient stops medication, symptoms usually return within two to seven days. On reinstitution of medication, symptoms will decrease to the previous level of control.

Drug treatment of Tourette syndrome can be complicated and should be supervised by a physician experienced in the management of this condition. Even then, extreme patience is required of the patient, family, school and physician as the drug is introduced or dose changed. There are now several drugs available with more being studied. The problem of side effects is complicated and of concern to all. Your doctor will discuss medication pros and cons with you. Please ask questions if you do not understand.

## **THE PROGNOSIS - WHAT WILL HAPPEN?**

### **THE PATIENT AND HIS ENVIRONMENT**

Years ago, with little or no knowledge of Tourette syndrome, the prognosis was vague. During the past 20 years more and more patients have been followed by doctors so that a considerable amount of information is now available. Based on this, patients may expect to lead fairly normal lives, even those receiving no treatment for reduction of symptoms considering the severity of tics.

Four factors are important: 1) the severity of symptoms, 2) how well-adjusted the patient was prior to onset of symptoms, 3) the relationship between the patient and his family, and 4) the treatment used, how soon treatment was begun after onset of symptoms and the patient's response to treatment.

It is important to remember that patients are the same as anybody else except for the symptoms which are beyond their control. Others respond to the patient differently depending on how they feel about the patient and how much they understand the problem. Those who do not understand may ridicule the patient or stay away because they feel uncomfortable. It is not difficult to understand why many patients prefer to spend much of their time alone. It is also not difficult to realize that a tolerant and understanding attitude is important in dealing with these patients. They require support and encouragement from all people, especially from their family and their school.

Parents should give the child the chance to exert as much independence as possible and encourage him to express himself. They should not attempt to structure the child's life excessively, for this results in hostility on both sides. The child should not be punished for exhibiting his symptoms nor should he be threatened in an attempt to stop tics or vocalizations.

Tourette patients have the same capacity for physical activity as other people. Sports can be very beneficial and should be encouraged.

Some patients learn to use the symptoms to control their environment. It is difficult, but parents should attempt to recognize these times and refuse to permit it, not by punishment, but by being firm and kind and in not giving in to his desires. These children do need structure in their life.

## SCHOOLS AND TEACHERS

Depending on severity of tics and the presence of learning problems, the Tourette patient can present a difficult problem in school. The teacher has the entire class to consider. However, the patient should be treated just like his classmates whenever possible. Intellectually, he may present problems since symptoms cause a decrease in scholastic progress. Rarely is there any change in the child's basic mental capability. The decrease in progress more likely represents reduced ability to concentrate and synthesize information because of the disruptive effect of tics and/or the presence of associated learning problems such as attention deficit disorder with or without hyperactivity.

Drowsiness may be the effect of excessive medication and should be reported to the parents and the patient's doctor. Aggressive behavior may arise because of classmates making fun of the patient. It helps to encourage the patient by reminding him of his positive attributes so he understands his capabilities are comparable to those of his classmates. Tutoring (one-on-one teaching) may also be helpful. Very helpful information on educational techniques is available through the Tourette syndrome Association. An educational session on the disorders for the child's class may be very helpful.

The most important point for the family and teacher to remember is that the patient, like everybody else, needs love, support and understanding. He has extra problems so he needs extra love and understanding to be able to cope with his problems.

## THE TOURETTE SYNDROME ASSOCIATION

The Tourette syndrome Association is a voluntary, non-profit organization composed of individuals with this disorder, their relatives, friends and other interested people including professionals treating this disorder. The goals of the association are:

- To educate the public and physicians about the symptomology of this disorder and current treatment available.
- To promote research toward finding the cause and, ultimately, a cure.
- To be an advocate for the patient.

The Association holds periodic meetings where the latest medical developments are discussed and where patients and their families can discuss common problems and offer support to one another. TSA publishes a newsletter and other literature. They prepare programs for radio and television; educational videotapes are available.

The headquarters of the Association is in Bayside, New York (outside New York City). There are regional chapters throughout the country to conduct the affairs of the Association in their local area. You are invited and encouraged to attend meetings and other activities and to participate directly in the work of the TSA.