

GLOSSARY

Anxiety - Can show as sleep difficulties, tension habits, motor unrest (fidgeting), phobias, worries, poor concentration, or panic attacks.

Attention Deficit Hyperactivity Disorder (ADHD) - Children may show signs of hyperactivity before TS symptoms appear. Signs include: poor concentration; failing to finish what is started; not listening; being easily distracted; clumsiness; often acting before thinking; shifting constantly from one activity to another; and general fidgeting. This starts before the age of seven and causes problems in school and at home. Adults too may show signs of ADHD, but instead of hyperactivity the signs tend to be restlessness, difficulty relaxing, impulsive behaviour and difficulty concentrating. Work and relationships may prove troublesome for adults with ADHD.

Conduct disorder - Can show as persistent and repetitive lying, stealing, truancy, starting fires, vandalism, fighting, or cruelty to animals.

Copropaxia and coprolalia - Copropaxia means making obscene or otherwise unacceptable movements or gestures. Coprolalia means using obscene or unacceptable language. This may involve swearing (though only 10-15% of people with TS have coprolalia) or racist remarks. Coprolalia can cause serious problems at school, in society and at work. It is particularly sad that the words uttered usually bear no relation to the true feelings of the speaker.

Depression - Should always be taken seriously. It is treatable and medical advice should be sought. In TS depression is most commonly seen in people with severe tics, sleep disturbances or OCD.

Echophenomena - Repeating other people's words (echolalia) and other people's gestures (echopraxia). Common in TS.

NOSI - Non-Obscene Socially Inappropriate behaviour, involves saying things that are socially unacceptable, for example personal remarks about height, weight or facial features.

Obsessive Compulsive and Ritualistic Behaviours - A person feels that something must be done over and over. Examples include touching an object with one hand after touching it with the other hand to 'even things up' or repeatedly flicking the light switch on and off. In more serious cases, the obsession may be around sexual, violent, religious or aggressive themes. Compulsions typically include checking, ordering, counting, repeating, getting things 'just right' or symmetrical, or forced touching. This is a different spectrum from the symptoms of 'pure' obsessive compulsive disorder (OCD).

Other psychopathology - Examples include rage attacks/aggression, oppositional defiant disorder, inappropriate sexual behaviour.

Rage attacks - Can be frightening and destructive. Once begun, a rage attack has to be left to run its course. Rage may be linked to tic suppression.

Paliphenomena - Similar to echophenomena. Involves the person with TS repeating their own words and actions such as "Hello, I came here by bus bus bus bus".

SIB - Self-injurious behaviour. It includes punching and slapping the head, face or body, or scratching or sticking sharp objects into the body, including the eyes. It can be an obsessional behaviour.

Sleep Disorders - These include frequent awakenings, or walking or talking in your sleep. They are fairly common among people with TS.

Symptoms

The symptoms of Tourette Syndrome (TS) are tics (repeated movements and sounds). It is important to understand that these are chronic (long-term) and involuntary. Someone with TS may be able to suppress tics for a period but eventually they have to let them out.

Tics usually start in childhood around the age of seven, and are usually worst between 10-12 years. However, in approximately half of TS patients, most symptoms disappear by the age of 18. TS is a persistent disorder but not always greatly disabling.

The first tics often start around the head and face, like blinking and/or grimacing. Vocal tics tend to appear later, around age 11. The different symptoms can be simple, such as blinking, or complex, like touching or jumping. Examples of vocal tics besides uttering words or making sounds are throat clearing, sniffing and/or coughing.

Even within the same person, the tics vary in many ways:

- they wax and wane; they get better and worse over time
- they change; one tic stops and another starts
- they may be made worse by stress and anxiety
- they may be alleviated with relaxation or concentration on an absorbing task

These changes are completely unpredictable. However, just before a tic is about to happen it is common to experience so called premonitory sensations. These sensations can be either localised - in the area where the tic is about to happen - or generalised. It is often very difficult, even for family, friends, teachers and employers of a person with TS, to believe that their actions or vocal utterances are involuntary, but they are.

Over 85 percent of people with TS have more than just tics. Additional conditions ('co-morbidities') include obsessive compulsive disorder (OCD) and/or attention deficit hyperactivity disorder (ADHD). Children and adults may also suffer from 'rages'. Co-morbidities often present more problems than the tics and can be less visible.

Types of tic

The range of tics in TS is very broad. The range of simple and complex tics is shown in Table 1 below.

table 1	Examples of simple tics	Examples of complex tics
Movement	Eye blinking, head jerking, shoulder shrugging and facial grimacing	Jumping, touching other people or things, smelling, twirling, and sometimes hitting or biting oneself
Sound	Throat clearing, yelping and other noises, sniffing, coughing and tongue clicking	Uttering words or phrases out of context, coprolalia (saying socially unacceptable words), and echolalia (repeating a sound, word, or phrase just heard)

Types of TS

TS can be classified into roughly three types, corresponding to 3 levels of severity and complexity. Attention deficit disorders (ADHD) and obsessive compulsive behaviours (OCB) often occur together with TS, and make it even harder to deal with.

This is illustrated in Table 2 below.

medical name		
pure tourettes	full blown tourettes	tourettes plus
		OCB or OCD, ADHD, depression, anxiety, SIB, sleep disorders, personality disorders, other psychopathology, conduct disorder
	paliphenomena echophenomena copropraxia/coprolalia, NOSI	paliphenomena echophenomena copropraxia/coprolalia, NOSI
movement and sound tics only	movement and sound tics	movement and sound tics

The table is for general guidance only. Tics and behaviours vary widely. It is perfectly possible, for example, to have OCB and sleep disorders without coprolalia.

Causes

The cause of TS has not yet been established, although it appears to involve an imbalance in the function of the neurotransmitters (chemical messengers in the brain) dopamine and serotonin. It is also likely to involve abnormalities in other neurotransmitter systems of the brain.

Brain scanning has revealed that there are some areas of the brain that appear to be different in TS individuals, for example some structures in the basal ganglia part of the brain, and in the fronto-temporal brain areas.

In the majority of cases, TS is inherited. A person with TS has roughly a 50 percent chance of passing on the gene to each of their children. However, the child may not inherit the parent's type of tics, but may show others instead. So far no single gene has been convincingly identified, and exactly how TS is inherited is not clear. The genetics of TS is complex and many researchers now believe that an individual may inherit a vulnerability to a spectrum disorder that includes TS, OCB, and perhaps also ADHD.

Research has shown that different environmental factors may, in some cases, contribute to the onset or affect the severity of TS. Streptococcal throat infections seem to play a role in triggering off TS in some individuals, by causing an abnormal immune reaction. The streptococcus bacteria do not cause TS by itself, but some individuals may well inherit susceptibility to both the syndrome and the way they react to some infections. Other possible environmental factors that may contribute to the onset or severity of TS include pregnancy and birth related problems such as complications during pregnancy, smoking during pregnancy, severe nausea and/or vomiting during the first trimester and premature low birth weight children.

In conclusion, the cause of TS is complex. All factors mentioned above most likely interact to produce the very special profile of symptoms and severity that is unique to each person with TS.

Diagnosis

For TS to be diagnosed, multiple motor tics and at least one vocal tic must be present over a period of at least twelve months, without a break of more than three months.

Some doctors may use an EEG, MRI, CT scan, or certain blood tests to rule out other conditions that might be confused with TS such as epilepsy, autism, dystonia and Sydenham's chorea. TS can only be diagnosed by observing and evaluating the symptoms. There are rating scales to help with the assessment of tic severity.

Treatments

There is currently no cure for TS. Most people with TS are not significantly affected by their tics or behaviours, and so do not require medication. There are medications which can help control the tics when they cause problems. However, while they may be very successful in some cases, they don't work for everybody, or they may have undesirable side effects (eg weight gain.)

Other types of therapy may also be helpful. Psychotherapy can assist a person with TS and help the family cope. Some behaviour therapies can teach the substitution of one tic for another that is more socially acceptable, or less painful. In addition, the use of relaxation techniques can alleviate stress that would otherwise make tics worse.

Special educational needs

School children with TS as a group have the same IQ range as the population at large, but the prevalence of TS in special educational needs groups is higher than across the whole school population. TS, especially combined with attention deficits, often calls for special educational assistance. Tape recorders or computers for reading and writing problems, untimed exams in a private room, and permission to leave the classroom when tics become overwhelming are often helpful.

It is important that TS is recognised and understood in the school so that children with TS are not, for example, punished for their tics.

Statistics

- TS is now believed to affect up to one in every 100 school children, although many have mild symptoms that may not require medical attention.
- Coprolalia (involuntary bad language) only affects about 10% of people with TS.
- TS is three to four times more common in males than in females.

Reference: This edition written by Judith Kidd and Claire Ball, updated by Dr Linnea Larsson.

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For more information on TS and our services, visit www.tourettes-action.org.uk.

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