

Gilles de la Tourette Syndrome (GTS)

What is Tourette Syndrome?

Tourette Syndrome is an inherited, neurological disorder characterized by repeated and involuntary body movements (tics) and uncontrollable vocal sounds. In a minority of cases, the vocalizations can include socially inappropriate words and phrases -- called coprolalia. These outbursts are neither intentional nor purposeful. Involuntary symptoms can include eye blinking, repeated throat clearing or sniffing, arm thrusting, kicking movements, shoulder shrugging or jumping. Coprolalia, once thought to be common, only occurs in a minority of TS cases.

These and other symptoms typically appear before the age of 18 and the condition occurs in all ethnic groups with males affected 3 to 4 times more often than females. Although the symptoms of TS vary from person to person and range from very mild to severe, the majority of cases fall into the mild category. Associated conditions can include obsessions, attention problems, impulsiveness, and sudden mood shifts and emotional outbursts. It is not unusual for a touterter (someone with Tourettes) to also have obsessive compulsive disorder (OCD) or attention deficit/hyperactivity disorder (ADHD).

GTS occurs in all populations and all ethnicities around the world. In the 1960's and 1970's some studies were published that incorrectly suggested a higher prevalence among individuals of Eastern European descent. This is a classic example of ascertainment bias: the studies were performed in clinics that served large local ethnic populations.

Most people with TS lead productive lives and participate in all professions. Increased public understanding and tolerance of TS symptoms are of paramount importance to people with Tourette syndrome.

An extremely thorough, up-to-date review is by Mary M. Robertson and Jeremy S. Stern, "The Gilles de la Tourette Syndrome," *Critical Reviews in Neurobiology*, 11(1):1-19 (1997).

My child has GTS. What did I do wrong?

NOTHING. There is no evidence that anything that you ever did, either during pregnancy or as a parent, had anything to do with your child developing GTS. There is nothing you could have done to prevent it as far as anybody knows.

Prevalence

The numbers quoted vary depending on how the calculation was performed. Some of the more recent numbers are listed in the following table.

Prevalence	Reference
5/10,000	Usual figure. ¹
0.77/10,000 (boys)	North Dakota. ²
0.22/10,000 (girls)	
105/10,000 (boys)	Duarte, CA. ³
13/10,000 (girls)	
4.9/10,000 (boys)	Israel. ⁴
3.1/10,000 (girls)	
2.87/10,000	Upstate NY. ⁵

¹Brunn (1984), *J. Am. Acad. Ch. Psychiat.*, 23:126

²Burd (1986), *Am. J. Psychiat.* 143:787

³Comings (1990), *J. Clin. Psychiat.*, 51:463

⁴Apter (1992), *Adv. in Neurology*, 58:67

⁵Caine (1988), *Neurology*, 38:472

Tics

A **tic** is usually defined as sudden and uncontrollable body movement (**motor tic**) or utterance (**vocal tic**). However, not all tics are sudden; some are slow, with an almost dance-like, seemingly choreographed motion (e.g., choreic tics). In Tourettes, most tics are more sudden. A **tonic tic** is marked by prolonged muscular contraction; a **clonic tic** is marked by a series of repeated contractions and relaxations.

Touretic tics may be **simple tics** that consist of a single movement - an eye blink, a head shrug (e.g., the "hair out of the eyes tic" is extremely common in GTS), a knee jerk, a snuffling or sniffing sound, throat clearing, snorting, coughing. While they are involuntary, one to two thirds of Touretters experience premonitory urges. They are aware that the tic is coming but can do nothing about it, and can delay it, but for only a short time. These can be compared to the urge to sneeze or to go to the bathroom. A **complex tic** is a series of simple tics or movements that are repeated as a set, or a seemingly purposeful motion, like hitting, pinching,

poking, smelling, or saying recognizable words or phrases.

Complex vocal tics are the most popularized fact regarding Tourettes. But only a small percentage of Touretters - probably fewer than 25% - every experience them. The most common occurrences include suddenly saying or repeating words or phrases completely out of context. They need not be unusual or unacceptable.

Echolalia, the repeating of words or phrases, such as "jingles" in a commercial, can occur anywhere from seconds to months, or even years later. (Consider the character in the movie *Forget Paris* who keeps repeating the Toyota jingle and reading street signs as they pass). **Palilalia** is the repeating of ones own words and phrases.

Coprolalia is the uttering of socially inappropriate words or phrases.

GTS tics range from very mild to very severe. They tend to wax and wane during the course of a Touretter's life. They may encompass the whole body - almost convulsive movements at times - and then seem to disappear for days, weeks, or months. It is not uncommon for tics to increase in severity during the teenage years and then almost disappear in the early twenties. Tics rarely disappear permanently.

Diagnostic Criteria

The standard clinical diagnostic criteria are

1. **Both** motor and vocal tics present at some time of life, not necessarily at the same time.
2. Tics are frequent, usually in "bouts" **and** occur nearly daily for some period **or** intermittently for a year **and** no more than three months without a tic.
3. Tics cause distress **or** interfere with social life **or** job function **or** school **or** something else that the ticer or his or her family considers important.
4. Onset before age 18.
5. Not drug-induced or due to another medical condition.

If only motor or only vocal tics are present, or they are not present for a full year, or they first occur after age 21, the diagnosis is more likely to be chronic motor (vocal) tic syndrome (CMTS),

CVTS). It is widely believed that CMTS and CVTS are just milder forms of Tourette Syndrome. The treatment is identical.

Is GTS Psychological?

NO. Not a single study has ever been published that provided any evidence of a psychological basis to GTS. There is considerable evidence for a physiological origin, however.

Genetics of GTS

The GTS gene is multifactorial – i.e., there are multiple genes involved, and it is the accumulation of a sufficient number of these genes that makes one susceptible to GTS. Genes that have been identified as contributing to GTS include several that encode dopamine transporter proteins.

Although it is multifactorial, many texts still state that GTS is caused by a single autosomal dominant gene with reduced penetrance. Autosomal genes are genes that are not on the sex chromosome (but are on one of the other twenty-two chromosomes). A gene is dominant if inheritance from a single parent is sufficient to express the protein that the gene encodes. When a gene is partially penetrant then there is less than a 100% chance of its expression, even if the gene is inherited; other factors, probably environmental, are required to "set it off."

There is evidence that viral infections (particularly streptococcus A) can do this. GTS may result from small brain lesions caused by antibodies to certain proteins in the brain (this probably also occurs in MS). When a touretter has an early childhood strep infection, his or her immune system develops antibodies that recognize specific proteins on the surface of the virus. These proteins are very similar to proteins in the basal ganglia, an organ of the brain that is involved in the control of movement. This is usually not a problem because the blood-brain barrier normally prevents antibodies from entering the brain. However, for (presumably genetic) reasons these antibodies migrate to the brain in susceptible individuals. Such antibodies have been identified in approximately two thirds of touretters tested, while less than one percent of non-touretters have them.

Why is GTS so frequently misdiagnosed?

Many physicians still incorrectly believe that GTS is rare and that coprolalia is required for diagnosis. Both of these beliefs are false. Strangely, many of the misdiagnoses are for truly rare disorders.

Some of the common misdiagnoses are acanthocytosis, amphetamine usage, Aspergers syndrome, athetoid cerebral palsy, bad parenting, Sydenham's Chorea, Huntington's disease, demonic possession, dystonia, encephalitis lethargica, epilepsy, Hallervorden-Spatz disease hyperparathyroidism, Jacob-Creutzfeld Disease, the "Jumping Frenchmen of Maine," klazomania, lack of exercise, latah, manganese poisoning, Meige's syndrome, myoclonus, myriatchit, neonatal karnicterus, Pelizaeus-Merzbacher disease, phenothiozine intoxication, phenylketonuria, psychosis, schizophrenia, spasmodic torticollis, status dysmyelinatus, and Wilson's disease.

Famous people with GTS

Some famous people with Tourettes include Samuel Johnson (1709-1784), the inventor of the English dictionary, the composer Wolfgang Mozart (1756-1791), major league baseball player Jim Eisenreich, NBA basketball player Mahmoud Abdul-Rauf (formerly Chris Jackson), surgeon Carl Bennett, actor Peter Antico, musician David Aldridge, novelist Kurt Tidmore, and late musician Julius Wechter (founded Baja Marimba Band, played with the Tijuana Brass). Kurt Cobain (1968-1994), the lead singer of Nirvana, may have had GTS, but there is no published evidence of that fact (he did write the song *Tourettes* published on the *In Utero* album). In fiction, the character Nikolai Levin in *Anna Karenina* was modeled after author Leo Tolstoy's brother Dmitry.

Why is it called Tourette Syndrome?

George Giles de la Tourette, who lived from 1857 to 1904, published the first paper on GTS in the medical literature.

For Further Reading

- Brunn RD, Brunn B, *A Mind of Its Own*, Oxford University Press, 1994.
- Comings DE, *Tourette syndrome and human behavior*, Hope Press, 1990.
- Haerle T, ed., *Children with Tourette syndrome: A parent's guide*, Woodbine House, 1992.
- Hughes S, *RYAN: A Mother's Story of Her Hyperactive/Tourette Syndrome Child*, Hope Press, 1990.
- Hughes S, *What Makes Ryan Tick? A Family's Triumph over Tourette Syndrome and Attention Deficit Hyperactivity Disorder*, Hope Press, 1996.
- Johnson-Hamer S, *Raising Joshua- One Mother's Account of the Challenges of Parenting a Child with Tourette Syndrome*. Hope Press, 1997.
- Landau E, *Tourette Syndrome*, Venture Books, 1998.
- Naseef R, *Special Children, Challenged Parents - The Struggles of Raising a Child with a Disability*, Carol Publishing Group, 1997.
- Robertson MM, Baron-Cohen S, *Tourette Syndrome: The Facts*, Oxford Medical Publications, 1998.
- Shimberg EF, *Living with Tourette Syndrome*, Fireside Books, 1995.

Text for Educators

- Dombush MP, Pruitt SK, *Teaching the Tiger A Handbook for Individuals Involved in the Education of Students with Attention Deficit Disorder, Tourette Syndrome or Obsessive-Compulsive Disorder*, Hope Press, 1995.

Medical Texts

- Chase TN, Friedhoff AJ, Cohen DJ, eds., "Tourette Syndrome." *Advances in Neurology*, Vol. 58, Raven Press, 1992.
- Jankovic J, "Tourette Syndrome." Special Issue of *Neurologic Clinics*, 15(2), May 1997, W. B. Saunders.
- Kurlan R, *Handbook of Tourette's Syndrome and Related Tic and Behavioral Disorders*, Marcel Dekker, 1993.
- Leckman JF, Cohen DJ, *Tourettes Syndrome: Tics, Obsessions, Compulsions, Developmental Psychopathology and Clinical Care*, Wiley, 1998.