


Tourette syndrome

Tourette syndrome (TS or simply **Tourette's**) is a common neurodevelopmental disorder with onset in childhood,^[4] characterized by multiple motor tics and at least one vocal (phonic) tic. Some common tics are eye blinking, coughing, throat clearing, sniffing, and facial movements. These tics characteristically wax and wane, can be suppressed temporarily, and are typically preceded by an unwanted urge or sensation in the affected muscles. Tics are often unnoticed by casual observers.

Tourette's was once considered a rare and bizarre syndrome, most often associated with coprolalia (the utterance of obscene words or socially inappropriate and derogatory remarks), but this symptom is present in only a small minority of people with Tourette's.^[3] It is no longer considered a rare condition; about 1% of school-age children and adolescents have Tourette's.^[1] Many individuals with Tourette's go undiagnosed or never seek medical care. There are no specific tests for diagnosing Tourette's; it is not always correctly identified because most cases are mild and the severity of tics decreases for most children as they pass through adolescence. Extreme Tourette's in adulthood, though sensationalized in the media, is a rarity. Tourette's does not adversely affect intelligence or life expectancy.

In most cases, medication for tics is not necessary. Education is an important part of any treatment plan, and explanation and reassurance alone are often sufficient treatment.^{[1][5]} Among those who are seen in specialty clinics, attention-deficit hyperactivity disorder (ADHD) and obsessive-compulsive disorder (OCD) are present at higher rates. These co-occurring diagnoses often cause more impairment to the individual than the tics; hence, it is important to correctly identify associated conditions and treat them.^[6]

Tourette's is defined as part of a spectrum of tic disorders, which includes provisional, transient and persistent (chronic) tics. While the exact cause is unknown, it is believed to involve a combination of genetic and environmental factors. The condition was named by Jean-Martin Charcot (1825–1893) on

Tourette syndrome	
Other names	Tourette's syndrome, Tourette's disorder, Gilles de la Tourette syndrome (GTS)
	
Georges Gilles de la Tourette (1857–1904), namesake of Tourette syndrome	
Specialty	Pediatrics, neurology
Symptoms	Tics ^[1]
Usual onset	Typically in childhood ^[1]
Duration	Long term ^[2]
Causes	Genetic with environmental influence ^[2]
Diagnostic method	Based on history and symptoms ^[1]
Treatment	Education, behavioral therapy ^{[1][3]}
Medication	Usually none, occasionally antipsychotics and noradrenergics ^[1]
Prognosis	Improvement to disappearance of tics beginning in late teens ^[2]
Frequency	About 1% ^[1]

behalf of his resident, Georges Albert Édouard Brutus Gilles de la Tourette (1857–1904), a French neurologist, who published an account of nine patients with Tourette's in 1885.

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Characteristics

Tics



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Examples of motor tics

Tics are movements or sounds "that occur intermittently and unpredictably out of a background of normal motor activity",^[7] having the appearance of "normal behaviors gone wrong".^[8] They are sudden, repetitive, nonrhythmic movements (motor tics) and utterances (phonic tics) that involve discrete muscle groups.^[9] Joseph Jankovic describes vocal or phonic tics as "motor tics that involve respiratory, laryngeal, pharyngeal, oral, and nasal musculature".^[10] Tics associated with Tourette's change in number, frequency, severity and anatomical location. Waxing and waning—the ongoing increase and decrease in severity and frequency of tics—occurs differently in each individual. Tics may also occur in "bouts of bouts", which vary for each person.^[9] The variation in tic severity may be hourly, daily or weekly.^[11] Tics may increase

when an individual is experiencing stress, fatigue or anxiety,^[12] or when an individual is engaged in relaxing activities like watching TV; they sometimes decrease when an individual is engrossed in or focused on an activity like playing a musical instrument.^[13]

Coprolalia (the spontaneous utterance of socially objectionable or taboo words or phrases) is the most publicized symptom of Tourette's, but it is not required for a diagnosis of Tourette's, and only about 10% of Tourette's patients exhibit it.^{[1][3]} Echolalia (repeating the words of others) and palilalia (repeating one's own words) occur in a minority of cases,^[9] while the most common initial motor and vocal tics are, respectively, eye blinking and throat clearing.^[14]

In contrast to the abnormal movements of other movement disorders such as choreas, dystonias, myoclonus, and dyskinesias, the tics of Tourette's are temporarily suppressible, nonrhythmic, and often preceded by an unwanted urge.^[15] Over time, about 90% of individuals with Tourette's are aware of an urge that precedes tic onset,^[11] similar to the need to sneeze or scratch an itch. Individuals describe the need to tic as a buildup of tension, pressure, or energy^{[16][17]} which they consciously choose to release, as if they "had to do it"^[18] to relieve the sensation^[16] or until it feels "just right".^{[18][19]} Examples of this urge are the feeling of having something in one's throat, or a localized discomfort in the shoulders, leading to the need to clear one's throat or shrug the shoulders. The actual tic may be felt as relieving this tension or sensation, similar to scratching an itch. Another example is blinking to relieve an uncomfortable sensation in the eye. These urges and sensations, preceding the expression of the movement or vocalization as a tic, are referred to as "premonitory sensory phenomena" or premonitory urges. Because of the urges that precede them, tics are described as semi-voluntary or "*unvoluntary*",^{[1][7]} rather than specifically *involuntary*; they may be experienced as a *voluntary*, suppressible response to the unwanted premonitory urge.^{[3][13]} Published descriptions of the tics of Tourette's identify sensory phenomena as the core symptom of the syndrome, even though they are not included in the diagnostic criteria.^{[17][20][21]}

While individuals with tics are sometimes able to suppress their tics for limited periods of time, doing so often results in tension or mental exhaustion.^{[1][3]} People with Tourette's may seek a secluded spot to release their symptoms, or there may be a marked increase in tics after a period of suppression at school or at work.^{[8][11]} Some people with Tourette's may not be aware of the premonitory urge. Children may be less aware of the premonitory urge associated with tics than are adults,^[11] but their awareness tends to increase with maturity;^[7] by the age of ten, most children recognize the premonitory urge.^[13] They may have tics for several years before becoming aware of premonitory urges. Children may suppress tics while in the doctor's office, so they may need to be observed while they are not aware they are being watched.^[22] The ability to suppress tics varies among individuals, and may be more developed in adults than children.

Onset and progression

Although there is no such thing as a typical case of Tourette syndrome,^[23] the condition follows a fairly reliable course in terms of the age of onset and the history of the severity of symptoms. Tics may appear up to the age of eighteen, but the most typical age of onset is from five to seven.^[1] A 1998 study published by Leckman and colleagues from the Yale Child Study Center^[24] showed that the ages of highest tic severity are eight to twelve (with an average of age ten), with tics steadily declining for most patients as they pass through adolescence.^{[11][25]}

The most common, first-presenting tics are eye blinking, facial movements, sniffing and throat clearing. Initial tics present most frequently in midline body regions where there are many muscles, usually the head, neck and facial region.^{[11][23]} This can be contrasted with the stereotyped movements of other disorders (such as stims and stereotypies of the autism spectrum disorders), which typically have an earlier age of onset; are more symmetrical; rhythmical and bilateral; and involve the extremities, for example, flapping the hands.^[26] Vocal tics usually appear years after motor tics, although they can appear first.^[27] Tics that appear early in the course of the condition are frequently confused with other conditions, such as allergies, asthma, and vision problems: pediatricians, allergists and ophthalmologists are typically the first to identify a child as having tics.^[9] In individuals with a greater severity of tics, complex tics may develop, such as "arm straightening, touching, tapping, jumping, hopping and twirling".^[11]

The severity of symptoms varies widely among people with Tourette's, and many cases may be undetected.^{[9][27][28]} Most cases of Tourette's in older individuals are mild and almost unnoticeable.^[29] Adults with TS presenting in clinics are atypical.^[1]

Co-occurring conditions

When symptoms are severe enough to warrant referral to clinics, attention-deficit hyperactivity disorder (ADHD) and obsessive-compulsive disorder (OCD) are often found in association with Tourette's.^[1] In specialty clinics, 30% of individuals with TS also have mood or anxiety disorders, or disruptive behaviors.^{[11][30]} When ADHD is present along with tics, the occurrence of conduct disorder and oppositional defiant disorder increase in likelihood.^[11] Aggressive behaviors and angry outbursts in persons with TS are not well understood; they are not associated with severe tics, but are associated with the presence of ADHD.^[31]

Compulsions resembling tics are present in some individuals with OCD; "tic-related OCD" is hypothesized to be a subgroup of OCD, distinguished from non-tic related OCD by the type and nature of obsessions and compulsions.^[32] Compared to the more typical contamination compulsions found in OCD without tics, tic-related OCD presents with more "counting, aggressive thoughts, symmetry and touching" compulsions.^[11] Compulsions associated with OCD without tics are usually related to obsessions and anxiety, while those in tic-related OCD are more likely to be a response to a premonitory urge.^[11]

Not all persons with Tourette's have ADHD or OCD or other comorbid conditions, although in clinical populations, a high percentage of patients presenting for care do have ADHD.^{[19][33]} Over time, Dale (2017) states that 85% of people with Tourette's have some co-occurring condition.^[11] Denckla (2006) reported that a ten-year overview of patient records revealed about 40% of people with Tourette's have "TS-only" or "pure TS", referring to Tourette syndrome in the absence of ADHD, OCD and other disorders.^{[34][35]} Dure and DeWolfe (2006) reported that 57% of individuals presenting with tic disorders had uncomplicated tics, while 43% had tics plus comorbid conditions.^[8] Sukhodolsky, et al (2017) stated that 37% of individuals in clinical samples have pure TS.^[33]

People with "full-blown Tourette's" have significant comorbid conditions in addition to tics.^[8] Among individuals with TS studied in clinics, between 2.9 and 20% have been reported to have autism spectrum disorders,^[36] but one study indicates that a high association of autism and TS may be partly due to difficulties in distinguishing between tics and tic-like behaviors or OCD symptoms seen in people with autism.^[37]

Classification

Tourette's was classified by the fourth version of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV-TR) as one of several tic disorders "usually first diagnosed in infancy, childhood, or adolescence" according to type (motor or phonic tics) and duration (transient or chronic). Transient tic disorders consisted of multiple motor tics, phonic tics or both, with a duration between four weeks and twelve months. Chronic tic disorder was either single or multiple, motor or phonic tics (but not both), which were present for more than a year.^[9] Tourette's was diagnosed when multiple motor tics, and at least one phonic tic, are present for more than a year.^[38]

The fifth version of the DSM (DSM-5), published in May 2013, reclassified Tourette's and tic disorders as motor disorders listed in the neurodevelopmental disorder category, and replaced transient tic disorder with provisional tic disorder, but made few other significant changes.^{[39][40][41]} Tic disorders are defined only slightly differently by the World Health Organization International Statistical Classification of Diseases and Related Health Problems, ICD-10; code F95.2 is for combined vocal and multiple motor tic disorder [de la Tourette].^{[42][43]}

Although Tourette's is the more severe expression of the spectrum of tic disorders,^[44] most cases are mild^[45] and many individuals with TS do not come to clinical attention.^{[1][28]}

Between 2008 and 2014, studies suggested that Tourette's is not a unitary condition as described in the existing classification systems.^{[11][46]} Distinguishing between pure TS, and TS when it is accompanied by comorbid conditions, has implications in the management of symptoms.^[46] Some experts believe that TS and chronic tic disorder should be considered the same disorder, because vocal tics are also due to muscular contractions, and should not be distinguished from motor tics.^[27]

Causes

The exact cause of Tourette's is unknown, but it is well established that both genetic and environmental factors are involved.^[47] Genetic epidemiology studies have shown that the overwhelming majority of cases of Tourette's are inherited, although the exact mode of inheritance is not yet known and no gene has been identified.^{[6][48][49]} In other cases, tics are associated with disorders other than Tourette's, a phenomenon known as tourettism.^[50]

A person with Tourette's has about a 50% chance of passing the gene(s) to one of his or her children, but Tourette's is a condition of variable expression and incomplete penetrance.^[51] Thus, not everyone who inherits the genetic vulnerability will show symptoms; even close family members may show different severities of symptoms, or no symptoms at all. The gene(s) may express as Tourette's, as a milder tic disorder (provisional or chronic tics), or as obsessive-compulsive symptoms without tics. Only a minority of the children who inherit the gene(s) have symptoms severe enough to require medical attention.^[52] Gender appears to have a role in the expression of the genetic vulnerability: males are more likely than females to express tics.^[22]

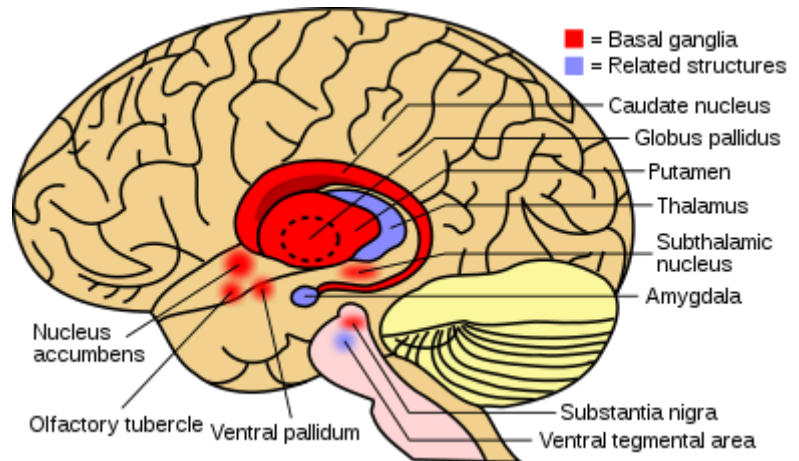
Non-genetic, environmental, post-infectious, or psychosocial factors—while not causing Tourette's—can influence its severity.^[23] Autoimmune processes may affect tic onset and exacerbation in some cases. Both obsessive-compulsive disorder (OCD) and tic disorders may arise in a subset of children as a result of a poststreptococcal autoimmune process.^[45] Children who meet five diagnostic criteria are classified, according to the hypothesis, as having Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS).^{[53][54]} The controversial PANDAS hypothesis includes the newer

PANS hypothesis, and both are the focus of clinical and laboratory research, but remain unproven; there is a broader hypothesis linking immune system abnormalities and immune dysregulation with TS.^{[3][11][45]}

Some forms of OCD may be genetically linked to Tourette's.^{[19][55]} A subset of OCD is thought to be causally related to Tourette's and may be a different expression of the same factors that are important for the expression of tics.^[56] The genetic relationship of ADHD to Tourette syndrome, however, has not been fully established.^[35]

Mechanism

The exact mechanism affecting the inherited vulnerability to Tourette's has not been established, and the precise cause is unknown. Tics are believed to result from dysfunction in cortical and subcortical regions, the thalamus, basal ganglia and frontal cortex.^[47] Neuroanatomic models implicate failures in circuits connecting the brain's cortex and subcortex.^[23] Imaging techniques implicate the basal ganglia and frontal cortex.^[48] After 2010, the role of histamine and the H3-receptor came into focus in the pathophysiology of TS,^[57] as "key modulators of striatal circuitry".^{[58][59]} A reduced level of histamine in the H3-receptor may disrupt other neurotransmitters, causing tics.^[60]



The basal ganglia and thalamus are implicated in Tourette syndrome.

Diagnosis

According to the fifth edition of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-5), Tourette's may be diagnosed when a person exhibits both multiple motor and one or more vocal tics over the period of a year; the motor and vocal tics need not be concurrent. The onset must have occurred before the age of 18, and cannot be attributed to the effects of another condition or substance (such as cocaine).^[38] Hence, other medical conditions that include tics or tic-like movements—such as autism or other causes of tourettism—must be ruled out before conferring a Tourette's diagnosis. Since 2000, the DSM has recognized that clinicians see patients who meet all the other criteria for Tourette's, but do not have distress or impairment.^{[61][62][63]}

There are no specific medical or screening tests that can be used in diagnosing Tourette's;^[19] it is frequently misdiagnosed or underdiagnosed, partly because of the wide expression of severity, ranging from mild (the majority of cases) or moderate, to severe (the rare, but more widely recognized and publicized cases).^[24] Coughing, eye blinking, and tics that mimic unrelated conditions such as asthma are commonly misdiagnosed.^[3] The diagnosis is made based on observation of the individual's symptoms and family history,^[3] and after ruling out secondary causes of tic disorders.^[52] In patients with a typical onset and a family history of tics or obsessive-compulsive disorder, a basic physical and neurological examination may be sufficient.^[44]

There is no requirement that other comorbid conditions, such as ADHD or OCD, be present,^[3] but if a physician believes that there may be another condition present that could explain tics, tests may be carried out to rule out that condition. An example of this is when diagnostic confusion between tics and seizure activity exists, which would call for an EEG, or if there are symptoms that indicate an MRI to rule out brain abnormalities.^[64] TSH levels can be measured to rule out hypothyroidism, which can be a cause of tics. Brain imaging studies are not usually warranted.^[64] In teenagers and adults presenting with a sudden onset of tics and other behavioral symptoms, a urine drug screen for cocaine and stimulants might be necessary. If a family history of liver disease is present, serum copper and ceruloplasmin levels can rule out Wilson's disease.^[44] Most cases are diagnosed by merely observing a history of tics.^[23]

Secondary causes of tics (not related to inherited Tourette syndrome) are commonly referred to as tourettism.^[50] Dystonias, choreas, other genetic conditions, and secondary causes of tics should be ruled out in the differential diagnosis for Tourette syndrome.^[44] Other conditions that may manifest tics or stereotyped movements include developmental disorders; autism spectrum disorders^[65] and stereotypic movement disorder;^{[66][67]} Sydenham's chorea; idiopathic dystonia; and genetic conditions such as Huntington's disease, neuroacanthocytosis, pantothenate kinase-associated neurodegeneration, Duchenne muscular dystrophy, Wilson's disease, and tuberous sclerosis. Other possibilities include chromosomal disorders such as Down syndrome, Klinefelter syndrome, XXY syndrome and fragile X syndrome. Acquired causes of tics include drug-induced tics, head trauma, encephalitis, stroke, and carbon monoxide poisoning.^{[44][50]} The symptoms of Lesch-Nyhan syndrome may also be confused with Tourette syndrome.^[26] Most of these conditions are rarer than tic disorders, and a thorough history and examination may be enough to rule them out, without medical or screening tests.^[23]

Screening

Although not all people with Tourette's have comorbid conditions, most Tourette's patients presenting for clinical care at specialty referral centers may exhibit symptoms of other conditions along with their motor and phonic tics.^[35] Along with ADHD and OCD, learning disabilities and sleep disorders may be present.^[3] Disruptive behaviors, impaired functioning, or cognitive impairment in patients with comorbid Tourette's and ADHD may be accounted for by the comorbid ADHD, highlighting the importance of identifying and treating comorbid conditions.^{[3][6][19][68]} Disruption from tics is commonly overshadowed by comorbid conditions that present greater interference to the child.^[23] Tic disorders in the absence of ADHD do not appear to be associated with disruptive behavior or functional impairment,^[69] while impairment in school, family, or peer relations is greater in patients who have more comorbid conditions and often determines whether therapy is needed.^[8]

Because comorbid conditions such as OCD and ADHD can be more impairing than tics, these conditions are included in an evaluation of patients presenting with tics. "It is critical to note that the comorbid conditions may determine functional status more strongly than the tic disorder", according to Samuel Zinner, MD.^[23] Because comorbid conditions such as ADHD or OCD can cause greater impact on overall functioning than tics, a thorough evaluation for comorbidity is called for when symptoms and impairment warrant.^[44] The initial assessment of a patient referred for a tic disorder should include a thorough evaluation, including a family history of tics, ADHD, obsessive-compulsive symptoms, and other chronic medical, psychiatric and neurological conditions. Children and adolescents with TS who have learning difficulties are candidates for psychoeducational testing, particularly if the child also has ADHD.^[64] Undiagnosed comorbid conditions may result in functional impairment, and it is necessary to

identify and treat these conditions to improve functioning. Complications may include depression, sleep problems, social discomfort, self-injury,^[44] anxiety, personality disorders, oppositional defiant disorder, and conduct disorders.^[70]

Management

Treatment of Tourette's is individualized and involves a collaboration between the clinician, individual with TS, and caregivers where applicable.^[25] It is focused on identifying and helping the individual manage the most troubling or impairing symptoms.^[3] Most cases of Tourette's are mild, and do not require pharmacological treatment;^[52] instead, psychobehavioral therapy, education, and reassurance may be sufficient,^[71] and "watchful waiting is an acceptable approach" for those without "functional impairment from their tics".^[25] Treatments, where warranted, can be divided into those that target tics and comorbid conditions, which, when present, are often a larger source of impairment than the tics themselves.^[64] Not all people with tics have comorbid conditions,^[35] but when those conditions are present, they often take treatment priority.^[25]

There is no cure for Tourette's and no medication that works universally for all individuals without significant adverse effects. Knowledge, education and understanding are uppermost in management plans for tic disorders.^[3] The management of the symptoms of Tourette's may include pharmacological, behavioral and psychological therapies. While pharmacological intervention is reserved for more severe symptoms, other treatments, such as supportive psychotherapy or cognitive behavioral therapy, may help to avoid or ameliorate depression and social isolation, and to improve family support. Educating a patient, family, and surrounding community (such as friends, school, and church) is a key treatment strategy, and may be all that is required in mild cases.^{[3][72]} Practice guidelines for the treatment of tics were published by the American Academy of Neurology in 2019.^[25]



Clonidine (or the clonidine patch) is one of the medications typically tried first when medication is needed for Tourette's.

Medication is available to help when symptoms interfere with functioning.^[52] The classes of medication with the most proven efficacy in treating tics—typical and atypical neuroleptics including risperidone, ziprasidone, haloperidol, pimozide and fluphenazine—can have long-term and short-term adverse effects.^[64] The antihypertensive agents clonidine and guanfacine are also used to treat tics; studies show variable efficacy, but a lower side effect profile than the neuroleptics.^[73] Stimulants and other medications may be useful in treating ADHD when it co-occurs with tic disorders. Drugs from several other classes of medications can be used when stimulants fail, including guanfacine, atomoxetine and tricyclic antidepressants.

Clomipramine, a tricyclic, and SSRIs—a class of antidepressants including fluoxetine, sertraline, and fluvoxamine—may be prescribed when a Tourette's patient also has symptoms of obsessive-compulsive disorder. Several other medications have been tried, but evidence to support their use is unconvincing.^[64]

Because children with tics often present to physicians when their tics are most severe, and because of the waxing and waning nature of tics, it is recommended that medication not be started immediately or changed often.^[23] Frequently, the tics subside with explanation, reassurance, understanding of the condition and a supportive environment.^[23] When medication is used, the goal is not to eliminate symptoms: it is used at the lowest dose that manages symptoms without adverse effects, given that these may be more disturbing than the symptoms for which the medication was prescribed.^[23]

Cognitive behavioral therapy (CBT) is a useful treatment when OCD is present.^[74] Other behavioral therapies including habit reversal training (HRT)/comprehensive behavioral intervention (CBIT) and exposure and response prevention (ERP) are first-line interventions,^[75] subject to some limitations: children younger than ten may not understand the treatment, people with severe tics or ADHD may not be able to suppress their tics or sustain the required focus to benefit from behavioral treatments, and there is a lack of therapists trained in behavioral interventions.^[76]

Relaxation techniques, such as exercise, yoga or meditation, may be useful in relieving the stress that may aggravate tics, but the majority of behavioral interventions (such as relaxation training and biofeedback, with the exception of habit reversal) have not been systematically evaluated and are not empirically supported therapies for Tourette's.^[77] Complementary and alternative medicine approaches, such as dietary modification, allergy testing and allergen control, and neurofeedback, have popular appeal, but no role has been proven for any of these in the treatment of Tourette syndrome.^{[78][79]} In spite of no evidence base supporting dietary approaches to management of TS symptoms as of 2018, anecdotal reports indicate that parents, caregivers, and individuals with TS are using dietary approaches and nutritional supplements.^[12]

Deep brain stimulation (DBS) has become a valid option for individuals with severe symptoms that do not respond to conventional therapy and treatment.^{[80][81]} There is low-quality, limited evidence that DBS is safe, well tolerated, and yields symptom reduction ranging from no change to complete remission.^[80] Selecting candidates who may benefit from DBS is challenging, and "age, tic severity, and treatment refractoriness are important factors to consider", according to Fraint and Pal (2016).^[81] The ideal brain location to target has not been identified as of 2016.^{[80][81][82]}

Prognosis

Tourette syndrome is a spectrum disorder—its severity ranges over a spectrum from mild to severe. The majority of cases are mild and require no treatment.^[52] Many people with TS may not realize they have tics; because tics are more commonly expressed in private, TS may go unrecognized or undetected.^[88] In these cases, the impact of symptoms on the individual may be mild, to the extent that casual observers might not know of their condition. Symptoms typically subside as children pass through adolescence;^[80] the overall prognosis is positive, but a minority of children with Tourette syndrome have severe symptoms that persist into adulthood.^[47]

Regardless of symptom severity, individuals with Tourette's have a normal life span. Although the symptoms may be lifelong and chronic for some, the condition is not degenerative or life-threatening. Intelligence is normal in those with Tourette's, although there may be learning disabilities.^[3] Severity of tics early in life does not predict tic severity in later life,^[3] and prognosis is generally favorable,^[89] although there is no reliable means of predicting the outcome for a particular individual. The



André Malraux (1901–1976) was a French author, adventurer and Minister of Culture who had Tourette syndrome.^{[83][84][85][86]} President Kennedy, Marie-Madeleine Lioux, Malraux, Jackie Kennedy and Vice President Johnson were photographed at the unveiling of the *Mona Lisa* at the National Gallery of Art, Washington, DC, in 1963.^[87]

gene or genes associated with Tourette's have not been identified, and there is no potential cure.^[89] A higher rate of migraines than the general population and sleep disturbances are reported.^[89]

By the time they reach adulthood, three-fourths of individuals with Tourette's experience a reduction in the severity of their tics.^[11] Several studies have demonstrated that the condition in most children improves with maturity. Tics may be at their highest severity at the time that they are diagnosed, and often improve with understanding of the condition by individuals and their families and friends. The statistical age of highest tic severity is typically between eight and twelve, with most individuals experiencing steadily declining tic severity as they pass through adolescence. One study showed no correlation between tic severity and the onset of puberty, in contrast with the popular belief that tics increase at puberty. In many cases, a complete remission of tic symptoms occurs after adolescence.^{[24][90]} However, a study using videotape to record tics in adults found that, although tics diminished in comparison with childhood, and all measures of tic severity improved by adulthood, 90% of adults still had tics. Half of the adults who considered themselves tic-free displayed evidence of mild tics.^{[11][91]}



Tim Howard, goalkeeper, described in 2019 by a staff writer for the *Los Angeles Times* as the "greatest goalkeeper in U.S. soccer history",^[92] attributes his success in association football to his Tourette's.^[93]

Children with Tourette's may suffer socially if their tics are viewed as "bizarre". If a child has disabling tics, or tics that interfere with social or academic functioning, supportive psychotherapy or school accommodations can be helpful.^[52] In children with tics, the additional presence of ADHD is associated with functional impairment, disruptive behavior, and tic severity.^[70]

Decreased measures in quality of life are observed in children whose tics are accompanied by ADHD, which can severely impact the child's well-being in all realms, and extend into adulthood.^[94] As ADHD symptoms improve with maturity, adults report less negative impact in their occupational lives than do children in their educational lives.^[94] Adults are more likely to report a reduced quality of life due to depression or anxiety.^[94]

A supportive environment and family generally gives those with Tourette's the skills to manage the disorder.^{[94][95][96]} People with Tourette's may learn to camouflage socially inappropriate tics or to channel the energy of their tics into a functional endeavor. Accomplished musicians, athletes, public speakers, and professionals from all walks of life are found among people with Tourette's. Outcomes in adulthood are associated more with the perceived significance of having severe tics as a child than with the actual severity of the tics. A person who was misunderstood, punished, or teased at home or at school is likely to fare worse than a child who enjoyed an understanding and supportive environment.^[9]

A study of 46 subjects at 19 years of age found that the symptoms of 80% had minimum to mild impact on their overall functioning, and that the other 20% experienced at least a moderate impact on their overall functioning.^[9] The rare minority of severe cases can inhibit or prevent individuals from holding a job or having a fulfilling social life. In a follow-up study of thirty-one adults with Tourette's, all patients completed high school, 52% finished at least two years of college, and 71% were full-time employed or were pursuing higher education.^[91]

Epidemiology

Tourette syndrome is found among all social, racial and ethnic groups and has been reported in all parts of the world;^{[3][19]} it is three to four times more frequent among males than among females.^[33] The tics of Tourette syndrome begin in childhood and tend to remit or subside with maturity; thus, a diagnosis may no longer be warranted for many adults, and observed prevalence rates are higher among children than adults.^[24] As children pass through adolescence, about one-quarter become tic-free, almost one-half see their tics diminish to a minimal or mild level, and less than one-quarter have persistent tics. Only 5 to 14% of adults experience worse tics in adulthood than in childhood.^{[3][6]} Up to 1% of the overall population experiences tic disorders, including chronic tics and transient tics of childhood.^[69] Chronic tics affect 5% of children, and transient tics affect up to 20%.^{[33][97]}

Most individuals with tics do not seek medical attention, so epidemiological studies of TS "reflect a strong ascertainment bias" towards those with co-occurring conditions.^[48] The reported prevalence of TS varies "according to the source, age, and sex of the sample; the ascertainment procedures; and diagnostic system",^[19] with a range reported between 0.15% and 3.0% for children and adolescents.^[33] Sukhodolsky, et al wrote in 2017 that the best estimate of TS prevalence in children was 1.4%,^[33] and Stern stated in 2018 that the prevalence in children was 1%.^[1] Prevalence rates in special education populations are higher.^[45]

Singer (2011) states the prevalence of TS in the overall population at any time is 0.1% for impairing cases and 0.6% for all cases,^[3] while Bloch and colleagues (2011) state the overall prevalence as between 0.3 and 1%.^[48] Robertson (2011) also suggests that the rate of Tourette's in the general population is 1%.^[45] Using year 2000 census data, a prevalence range of 0.1 to 1% yields an estimate of 53,000–530,000 school-age children with Tourette's in the US,^[69] and a prevalence estimate of .1% means that in 2001 about 553,000 people in the UK age five or older would have Tourette's.^[29]

Tourette syndrome was once thought to be rare: in 1972, the US National Institutes of Health (NIH) believed there were fewer than 100 cases in the United States,^[98] and a 1973 registry reported only 485 cases worldwide.^[99] However, multiple studies published since 2000 have consistently demonstrated that the prevalence is much higher than previously thought.^[100] Discrepancies between current and prior prevalence estimates come from several factors: ascertainment bias in earlier samples drawn from clinically referred cases; assessment methods that may fail to detect milder cases; and differences in diagnostic criteria and thresholds.^[100] There were few broad-based community studies published before 2000 and until the 1980s, most epidemiological studies of Tourette syndrome were based on individuals referred to tertiary care or specialty clinics.^[101] Individuals with mild symptoms may not seek treatment and physicians may not confer an official diagnosis of TS on children out of concern for stigmatization;^[88] children with milder symptoms are unlikely to be referred to specialty clinics, so prevalence studies have an inherent bias towards more severe cases.^[102] Studies of Tourette syndrome are vulnerable to error because tics vary in intensity and expression, are often intermittent, and are not always recognized by clinicians, patients, family members, friends or teachers.^{[23][103]} Approximately 20% of persons with Tourette syndrome do not recognize that they have tics.^[23] Newer studies—recognizing that tics may often be undiagnosed and hard to detect—use direct classroom observation and multiple informants (parents, teachers, and trained observers), and therefore record more cases than older studies relying on referrals.^{[72][104]} As the diagnostic threshold and assessment methodology have moved towards recognition of milder cases, there has been an increase in estimated prevalence.^[100]

History

The first presentation of Tourette syndrome is thought to be in the book, *Malleus Maleficarum* (Witch's Hammer) by Jakob Sprenger and Heinrich Kraemer, published in the late 15th century and describing a priest whose tics were "believed to be related to possession by the devil".^{[105][106]} A French doctor, Jean Marc Gaspard Itard, reported the first case of Tourette syndrome in 1825,^[107] describing the Marquise de Dampierre, an important woman of nobility in her time.^[108] Jean-Martin Charcot, an influential French physician, assigned his resident Georges Albert Édouard Brutus Gilles de la Tourette, a French physician and neurologist, to study patients at the Salpêtrière Hospital, with the goal of defining an illness distinct from hysteria and chorea.^[22]



Jean-Martin Charcot (1825–1893) was a French neurologist and professor who bestowed the eponym for Tourette syndrome on behalf of his resident, Georges Albert Édouard Brutus Gilles de la Tourette.

In 1885, Gilles de la Tourette published an account in *Study of a Nervous Affliction* describing nine persons with "convulsive tic disorder", concluding that a new clinical category should be defined.^{[109][110]} The eponym was later bestowed by Charcot after and on behalf of Gilles de la Tourette.^{[22][111]}

Little progress was made over the next century in explaining or treating tics, and a psychogenic view prevailed well into the 20th century.^[22] The possibility that movement disorders, including Tourette syndrome, might have an organic origin was raised when an encephalitis epidemic from 1918–1926 led to a subsequent epidemic of tic disorders.^[112]

During the 1960s and 1970s, as the beneficial effects of haloperidol (Haldol) on tics became known, the psychoanalytic approach to Tourette syndrome was questioned.^[113] The turning point came in 1965, when Arthur K. Shapiro—described as "the father of modern tic disorder research"^[114]—treated a Tourette's patient with haloperidol, and published a paper criticizing the psychoanalytic approach.^[112]

Since the 1990s, a more neutral view of Tourette's has emerged, in which biological vulnerability and adverse environmental events are seen to interact.^{[22][23]} In 2000, the American Psychiatric Association published the DSM-IV-TR, revising the text of DSM-IV to no longer require that symptoms of tic disorders cause distress or impair functioning,^[61] recognizing that clinicians often see patients who meet all the other criteria for Tourette's, but do not have distress or impairment.^{[62][63]}

Findings since 1999 have advanced TS science in the areas of genetics, neuroimaging, neurophysiology, and neuropathology. Questions remain regarding how best to classify Tourette syndrome, and how closely Tourette's is related to other movement or psychiatric disorders. Good epidemiologic data is still lacking, and available treatments are not risk free and not always well tolerated.^[115] High-profile media coverage focuses on treatments that do not have established safety or efficacy, such as deep brain stimulation, and alternative therapies involving unstudied efficacy and side effects are pursued by many parents.^[116]

Society and culture

Not everyone with Tourette's wants treatment or a "cure", especially if that means they may "lose" something else in the process.^{[117][118]} Researchers Leckman and Cohen, and former US Tourette Syndrome Association (TSA) national board member Kathryn Taubert, believe that there may be latent advantages associated with an individual's genetic vulnerability to developing Tourette syndrome, such as a heightened awareness and increased attention to detail and surroundings that may have adaptive

value.^[119] There is evidence to support the clinical lore that children with "TS-only" (Tourette's in the absence of comorbid conditions) are unusually gifted: neuropsychological studies have identified advantages in children with TS-only.^{[35][120]} Children with TS-only are faster than the average for their age group on timed tests of motor coordination.^{[35][121]}

Notable individuals with Tourette syndrome are found in all walks of life, including musicians, athletes, media figures, teachers, physicians, and authors.^[122] A well-known example of a person who may have used obsessive-compulsive traits to advantage is Samuel Johnson, the 18th-century English man of letters, who likely had Tourette syndrome as evidenced by the writings of James Boswell.^{[123][124]} Johnson wrote A Dictionary of the English Language in 1747, and was a prolific writer, poet, and critic. Tim Howard, described by the *Chicago Tribune* as the "rarest of creatures—an American soccer hero",^[125] and by the TSA as the "most notable individual with Tourette Syndrome around the world",^[126] says that his neurological makeup gave him an enhanced perception and an ability to hyper-focus that contributed to his success on the field.^[93]



Samuel Johnson (1709–1784) c. 1772. Johnson is likely to have had Tourette syndrome.

Although it has been speculated that Mozart had Tourette's,^{[127][128]} no Tourette's expert or organization has presented credible evidence to support such a conclusion,^[128] and there are problems with the arguments supporting the diagnosis: tics are not transferred to the written form, as is supposed with Mozart's scatological writings; the medical history in retrospect is not thorough; side effects due to other conditions may be misinterpreted; "it is not proven whether written documents can account for the existence of a vocal tic" and "the evidence of motor tics in Mozart's life is doubtful".^[129]

Pre-dating Gilles de la Tourette's 1885 publication, likely portrayals of TS or tic disorders in fictional literature are Mr. Pancks in *Little Dorrit* by Charles Dickens and Nikolai Levin in *Anna Karenina* by Leo Tolstoy.^[130] The entertainment industry has been criticized for depicting those with Tourette syndrome as social misfits whose only tic is coprolalia, which has furthered stigmatization and the public's misunderstanding of those with Tourette's.^[131] The coprolalic symptoms of Tourette's are also fodder for radio and television talk shows in the US^[132] and for the British media.^[133]

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External links

- [Tourette syndrome \(https://curlie.org/Health/Conditions_and_Diseases/Neurological_Disorders/Tourette_Syndrome/\)](https://curlie.org/Health/Conditions_and_Diseases/Neurological_Disorders/Tourette_Syndrome/) at Curlie

Classification **ICD-10:** F95.2 (<http://apps.who.int/classifications/icd10/browse/2016/en#/F95.2>) • **ICD-9-CM:** 307.23 (<http://www.icd9data.com/getICD9Code.ashx?icd9=307.23>) • **OMIM:** 137580 (<https://omim.org/entry/137580>) • **MeSH:** D005879 D005879, D005879 (https://www.nlm.nih.gov/cgi/mesh/2015/MB_cgi?field=uid&term=D005879.) • **DiseasesDB:** 5220 (<http://www.diseasesdatabase.com/ddb5220.htm>)

External resources **MedlinePlus:** 000733 (<https://www.nlm.nih.gov/medlineplus/ency/article/000733.htm>) • **eMedicine:** med/3107 (<https://emedicine.medscape.com/med/3107-overview>) neuro/664 (<http://www.emedicine.com/neuro/topic664.htm#>) •

GeneReviews:

Tourette Disorder (https://www.ncbi.nlm.nih.gov/books/NBK21138/)

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