

Tics and Tourette Syndrome in Children — Symptoms and Treatment

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Tics and related diseases represent a set of neurobehavioral disorders of childhood that are characterized by sudden, repeated, nonrhythmic, muscle movements that are accompanied by sounds or vocalizations. Tourette syndrome is the most severe form with the symptoms running a chronic course of more than one year. The disease is mainly diagnosed by history taking and physical examination of the characteristic tic phenomenon. Treatment involves behavioral therapy, antipsychotics, and treatment of comorbid conditions such as ADHD and OCD. The disease begins in childhood, achieves full expression in adolescence, and disappears in teenage.



Definition of Tics and Tourette Syndrome

Tics represent a neurobehavioral disorder of childhood that is **characterized by sudden, repeated, nonrhythmic, muscle movements that are accompanied by sounds or vocalizations**. Tourette's syndrome represents the same symptoms and

lasts for more than one year. The activities do not interfere with a child's intended activities or self-image.

Epidemiology of Tics and Tourette Syndrome

Tics affect 20 % of children worldwide, many of whom are not diagnosed or treated.

The severe form (Tourette's syndrome) affects 3—8 per 1000 children.

The disease has a slight male predilection with a male to female ratio of 3:1.

The disease takes a typical variation mainly affecting children aged 4—9 years while the improvement is noted as the patient grows older and disappears in adulthood for most of the cases. The average age of onset of symptoms is 7.8 years. More than half of children suffering from tics and related disorders will lead a normal adult life.

Tourette syndrome is commonly associated with other comorbidities such as obsessive-compulsive disorder, depression, anxiety, attention-deficit/hyperactivity disorder, and self-injurious behavior. The strong association with obsessive-compulsive disorder intrigued medical doctors and surgeons as it meant a common target for deep brain stimulation might be involved in both conditions.

Etiology and Pathophysiology of Tics and Tourette Syndrome

The causative pathway of the disease remains largely unknown but several theories have been put forward to explain the cause of the disease which includes:

Idiopathic theory

In most of the cases, the cause is unknown and there is no exhaustive explanation of the pathogenesis of the disease.

Genetic influence theory

Tics are thought to arise from a genetic mutation with the developmental period. This is supported by the fact that **there is a 45 % chance that a couple with tics or its variant will give birth to a child with any of the variants of tics** and a 15 % chance of developing Tourette syndrome. This idea was put forward by Georges Gilles la Tourette in 1800 who suggested an autoimmune dominant pattern with incomplete penetrance of inheritance.

Autoimmune theory

Some cases of tics are preceded by an infection with group A β hemolytic streptococci. It was thus renamed a pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection (PANDAS). Other associated infections include Herpes simplex virus infection, HIV, mycoplasma pneumonia and varicella Zoster Virus.

Neurobiological theory

The disease is thought to be a **disorder of the fibers that pass from the frontal cortex to the deeper areas of the brain** such as the thalamus. This pathway is known as the cortical-striatal-thalamal-cortical pathway (CSTC). Pathology in this pathway causes

manifestations of tics/Tourette syndrome. Multiple neurotransmitters such as dopamine, GABA, glutamate, serotonin, and acetylcholine are incriminated in the causative pathway of the disease. The most characterized one is dopamine as improvement is noted on the administration of neuroleptics.

In 2012, the **International Deep Brain Stimulation Database and Registry** was launched. One of the goals of this database was to pool data and identify novel pathophysiologic mechanisms and new therapeutic targets in Tourette syndrome. The **centromedian thalamic** region, **anterior globus pallidus internus, posterior globus pallidus internus, and the anterior limb of the internal capsule** were the most common targets for deep brain stimulation in Tourette syndrome. Accordingly, it is currently hypothesized that these brain regions are the ones responsible for the motor and phonic symptoms of Tourette syndrome.

Clinical features of Tics and Tourette Syndrome

Tics can present with various features such as they may be:

1. **Simple tics:** represents a brief movement or vocalization with no social meaning
2. **Complex tics:** tics lasting longer and have a social meaning
3. The tics can also take a **predominantly motor or sensory form** as summarized below:

	Motor Tics	Sensory Tics
Simple Tics	<ul style="list-style-type: none"> • Eye blinking • Eye rolling • Limb and head jerking. • Grimacing. • Shoulder shrugging 	<ul style="list-style-type: none"> • Throat clearing. • Whistling • Coughs • Grunting • Animal sounds
Complex Tics	<ul style="list-style-type: none"> • Jumping • Touching objects and other people • Copropraxia • Echopraxia 	<ul style="list-style-type: none"> • Coprolalia • Echolalia

Other tic characteristics

Fluctuating course

The **tics have a waxing and waning pattern** where they are more prominent when the patient is resting, concentrating, emotionally pleased and less prominent when the patient is engaged in an activity but are always absent during sleep. Tics are exacerbated by periods of anticipation, emotional upset (e.g. stress, anxiety, excitement, anger), or fatigue.

Presence in typical locations

The most common first symptom is a facial tic (eye blink, nose twitch, and grimace) and they almost always involve the head and neck region. Some tics may appear to have a social meaning and may be thought to represent a normal function.

Voluntary suppressibility

This active suppression of tics represents the ability to voluntarily suppress the tic, however, is often associated with a build-up of inner tension that resolves when the tic is

permitted to occur.

Premonitory sensations

Premonitory sensations are sensory **phenomena that occur before a motor or phonic tic and take the form of an urge**, impulse, tension, pressure, itch, or tingle. The recognition that tics may represent a voluntary response to an involuntary sensation has led some investigators to classify tics as “involuntary” rather than involuntary. Premonitory sensations are more common in adults.

Clinical features of associated conditions

Attention deficit hyperactivity disorder(ADHD) patients present with inattentiveness, hyperactivity, and impulsiveness.

Obsessive-compulsive (OCD) disorder

Obsessive-compulsive disorder (OCD) patients present with preoccupation, multiple obsessional thoughts, and compulsions that interfere with normal activity. Patients with concurrent anxiety have extreme worry and tension.

Classification of Tics and Tourette Syndrome

According to the diagnostic and statistical manual of mental illnesses, tic disorders can be classified into:

- Provisional tic disorder
- Chronic tic disorder
- Tourette syndrome
- Substance-induced tic disorder
- Tic disorder due to general medical condition
- Tic disorder otherwise non-specified

Investigations of Tics and Tourette Syndrome

The diagnosis of tics is usually clinical with the identification of specific tic phenomenology and their classification. However, **some tests are carried out to rule out differential diagnosis and for research purposes to characterize the disease further.**

To characterize a finding as a tic it must meet the **DSM V criteria that provides for:**

- Involuntary, sudden, unprovoked, repetitive, and nonrhythmic movements or vocalizations
- The Absence of other etiologies such as drug and substance abuse
- A duration of more than one year of symptoms indicates Tourette syndrome

Tic phenomenology

Simple motor tics are movements that involve a single muscle or one muscle group. Complex motor tics are a cluster of simple actions or coordinated movements dystonic character that involves holding in a position for long. Simple vocal tics include grunts, barks, and hoofs. Mainly simple sounds.

Complex vocal tics are vocalizations that involve **repetition of words** of other people

such as:

- **Echolalia:** repetition of words said by others
- **Palilalia:** repetition of one's own words
- **Coprolalia:** use of obscene words in public
- **Copropraxia:** making obscene gestures in public or touching genitals
- **Echopraxia:** imitating the gestures of others

The tests to rule out differential diagnosis include:

Test	Findings
Functional MRI	Used in research of tics and Tourette syndrome to demonstrate brain function during the occurrence of tics. Shows reduced caudate volumes.
CT Scan	Has been employed in investigating any abnormalities and may show reduced caudate volumes.
EEG	Needed in the study of tics in the sleep period to ascertain the nature of brain activity during an occurrence.
PET Scan	Shows increased brain activity in the areas involved such as thalamus and striatum.
Blood workups	Are needed before the administration of certain medications and necessary to rule out chronic diseases.

Differential Diagnosis of Tics and Tourette Syndrome

Diagnostic considerations in tic diseases should include:

- Anxiety disorders
- ADHD
- Autism
- Drug and substance abuse (cocaine toxicity)
- Obsessive compulsion disorder(OCD)
- Huntington disease dementia

Management of Tics and Tourette Syndrome

Treatment is only needed when the disease interferes with the child's image, social activity, or education. The **available options** include:

Cognitive behavioral therapy (CBT)

This option **involves a series of psychological interventions known as Comprehensive Behavioral Intervention for Tics (CBT)** is a combination of the following elements:

Habit Reversal Therapy (HRT)

The first stage of HRT is tic description, awareness and characterizing the tic. Understanding where they occur in the body, which muscles are involved and associated premonitory signs that indicate a tic episode is likely to occur. The next stage is finding a competing response which when employed reduces the chances of occurrence of the tics.

Education about tics

Teaching about Tic Disorders and Tourette syndrome should be taught to the affected persons and the people who take care of the patient. The information that is typically included in psychoeducation is:

- Understanding the causes
- Understanding tics as a neurobehavioral disease
- Understanding the course of the disease and the co-occurring conditions that often occur in individuals with Tourette syndrome

The knowledge helps to cope with having a tic disorder and makes it easier for helper or teachers to handle these patients.

Social Support and Reward System

The Presence of educated and willing caregivers helps to reduce the associated psychiatric conditions such as depression and anxiety that result from social detachment of the patient.

Functional Intervention

This involves the identification of environmental events that make tics worse or maintain tics for an individual. It may **include reactions to situation, thoughts, or feelings that a person has in a place**. This can be done with help from a trained therapist who can also offer strategies for alternative management of the tics.

Relaxation Training

Relaxation is used to reduce the stress which could be a likely trigger to the worsening of tics. Relaxation has been shown to reduce the frequency of occurrence of the tics.

Pharmacological therapy/antipsychotics

Examples of medications in use include clonidine, risperidol, haloperidol and olanzapine. Dopamine receptor antagonist has been widely used with improvement due to the associated neurobiology of the disease.

Deep brain stimulation (DBS)

This **involves implantation of electrodes and passage of stimulatory current to selected areas of the brain** whose increased activity is likely to give tics. These regions include the thalamus and Globus pallidus. The efficacy of deep brain stimulation is the same after the stimulation of the four brain-region targets mentioned before. The adverse-events associated with deep brain stimulation can be classified as device-related, surgery-related and stimulation-related.

- Device-related adverse-events include the implantation of the electrodes outside the target region or the removal of the pulse generation. This is reported in 2% of the patients undergoing DBS.
- The most common surgery-related adverse-events are infections (3%) and hemorrhages (1%).
- The stimulation-related adverse-events are plenty. They include bradykinesia, depression, dysarthria, paresthesias, lethargy, and nausea or vertigo. The most common adverse-events that the patient should be warned about are dysarthria, nausea, and paresthesias.

Other interventions

Yoga, meditation, nutritional therapy, and hypnotherapy have been tried in the management of tics and Tourette syndrome with some level of success reported.

Treatment of comorbid conditions

ADHD is treated with low dose stimulants and monitoring for tics done after that. This is done together with classroom and social accommodation such as offering extra time to complete tasks or one on one tutorials for the affected children. Administration of antidepressants such as SSRIs (fluoxetine and clomipramine) treats OCD. Anxiety and behavioral conditions are treated only when they can fit in the DSM diagnostic criteria.

Prognosis of Tics and Tourette Syndrome

Tics have a defined course with diagnosis done at younger ages of 9–13 years and the disease progress to full expression in the adolescent period where the symptoms begin normalizing and **the patient's clinical condition improves in teenage years**. Less than half will have residual disease in adulthood.

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