JOURNAL OF NEUROLOGY AND PSYCHOLOGY RESEARCH

CASE STUDY Open Access

# Tourette's syndrome: II. Diagnosis and Symptoms Management

**Dr. Alain L. Fymat***

Professor, International Institute of Medicine & Science, California, USA.

**Received date:** July 11, 2023, **Accepted date:** July 16, 2023, **Published date:** July 21, 2023.



***Corresponding Author:** Dr. Alain L. Professor, International Institute of Medicine & Science, California, USA.

## Abstract

The diagnosis of Tourette's syndrome (TS) is clinically based on the combination of signs and symptoms and medical history. There are no blood, laboratory, or imaging tests needed for diagnosis. In this Article, I will elaborate on the difficulties of diagnosing TS and the associated requirements and conditions to be diagnosed with TS, persistent (chronic) motor or vocal/phonic tic disorder, or else provisional (transient) tic disorder. I will also address the issues of timely or delayed diagnosis and even misdiagnosis, and the sudden onset of tic-like behaviors. I will further discuss tourettism, which encompasses tics that are associated with other disorders, such as the abnormal movements of chorea, dyskinesia, dystonia, and myoclonus, and developmental and autism spectrum disorder (ASD). I will additionally consider the diagnostic confusion between tics and seizure activity, functional tic-like movements, other conditions and possibilities, and acquired causes of tics. When symptoms are severe enough, one must also consider screening for co-occurring conditions such as attention-deficit hyperactivity disorder (ADHD), obsessive compulsive disorder (OCD), and other neurophysiological conditions. In the absence of a cure or an effective medication for treating all symptoms, I will review the avenues available for the management of TS symptoms including the touted behavioral therapy (BT) and comprehensive behavioral intervention therapy (CBIT) and present a systematic approach/algorithm for behavioral and psychobehavioral therapy of TS patients. Lastly, the clinical practice guidelines issued by the several professional and regulatory organizations will be summarized.

## Abbreviations

AAN: American Academy of Neurology; ADHD: Attention-Deficit Hyperactivity Disorder; ASD: Autism Spectrum Disorder; AT: Awareness Training; BT: Behavioral Therapy; CBIT: Comprehensive Behavioral Intervention Therapy; CD: Compulsive Disorder; CRT:

Competing Response Ttraining; CT: Computerized tomography; DMD: Duchenne's Muscular Dystrophy; DS: Down's Syndrome; DSM: (AAN's) Diagnostic & Statistical Manual of Mental Disorders; EEG: Electroencephalography; ERP: Exposure and Response Prevention; EU: European Union; HD: Huntington's Disease; HRT: Habit Reversal Training; ICD: (WHO's) International Classification of Diseases; IED: Intermittent Explosive Disorder; KS: Klinefelter's Syndrome; LNS: Lesch-Nyhan Syndrome; MRI: Magnetic Resonance Imaging; NCBDDD: (CDC&P's) National Center on Birth Defects and Developmental Disabilities; OCD: Obsessive–Compulsive Disorder; ODD: Oppositional Defiant Disorder; PANDAS: Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections; PBT: Psychobehavioral Therapy; PKAN: Pantothenate Kinase-Associated Neurodegeneration; PT: Psychotherapy; RCT: Randomized controlled trials; SC: Sydenham's Chorea; SSRI: Selective Serotonin Re-uptake Inhibitors; TAA: Tourette Association of America; THEO: Tourette Health and Education Outreach; TS: Tourette's syndrome; TS: Tuberous Sclerosis; TS-BTI: Tourette Syndrome Behavior Training Institutes; TSH: Thyroid-Stimulating Hormone; WD: Wilson's Disease; WHO: World Health Organization; YGTSS: Yale Global Tic Severity Scale.

## Keywords

Attention-deficit hyperactivity disorder; Autism spectrum disorder; Comprehensive behavioral intervention therapy; Down's syndrome; Duchenne's muscular dystrophy; Habit reversal training; Huntington's disease; Klinefelter's syndrome; Lesch-Nyhan syndrome; Neuroacanthocytosis; Obsessive–compulsive disorder; Pantothenate kinase-associated neurodegeneration; Sydenham's chorea; Tourette diagnosis; Tourette symptoms management; Tourettism; Tuberous sclerosis; Wilson's disease.

## Introduction

The diagnosis of Tourette's syndrome (TS) is clinically based on the combination of signs and symptoms and medical history. There are no blood, laboratory, or imaging tests needed for diagnosis. In rare cases, neuroimaging studies such as magnetic resonance imaging (MRI), computerized tomography (CT), electroencephalography (EEG) or certain blood tests may be used to rule out other conditions that might be confused with TS, but hey are not needed to establish TS as their results usually come back normal for TS.

## Diagnosing TS

Common tics are often diagnosed by a primary care provider, a pediatrician, or a mental healthcare specialist. However, atypical (non-classical) symptoms or atypical presentations (for example, symptoms that begun in adulthood) may require additional expertise for diagnosis. There are three criteria that health professionals will use to diagnose TS or other tic disorders:

1. **Presence of motor and vocal/phonic tics:** The presence, not necessarily at the same time, of both motor (at least 2) and vocal/phonic (at least 1) tics, including their onset (before age 18), occurrence pattern, and frequency (several times a day, every day, intermittently).

2. **Waxing-and-waning of tics:** Tics may wax and wane in frequency but have occurred for more than 1 year.

3. **Tics not caused by medical conditions or medications** other than the use of substances (particularly toxicants).

Tic disorders differ from each other in terms of the type of tics present (motor or vocal, or a combination of

both), their character (chronic/persistent or transient/provisional), and duration of the symptoms (more than 1 year for chronic tics and less than 1 year for transient tics).

**To be diagnosed with TS**

To be diagnosed with TS, a person must have:

• Two or more motor tics (for example, blinking or shrugging the shoulders) and at least one vocal tic (for example, humming, clearing the throat, or yelling-out a word or phrase), although they might not always happen at the same time;

• Had tics for at least a year. The tics can occur many times a day (usually in bouts) nearly every day, or off and on;

• Tics began before age 18 years;

• Symptoms are not due to taking medicine or other drugs or due to having another medical condition, for example, seizures, Huntington's disease (HD), or postviral encephalitis that can cause tics;

• There should be no underlying neurological symptoms; and

• There should be no intake of certain substances (toxicants).

It may take some time to receive a formal diagnosis of TS. Families and physicians unfamiliar with the disorder might think that mild and even moderate tic symptoms may be insignificant, a part of a developmental phase, or the result of another condition. For example, some parents may think that eye blinking is related to vision problems or that sniffing is related to seasonal allergies.

**To be diagnosed with a persistent (chronic) motor or vocal tic disorder**

To be diagnosed with a persistent tic disorder, a person must have:

• One or more motor or vocal tics, but not both;

• Tics that occur many times a day nearly every day or on-and-off throughout a period of more than a year;

• Tics that start before age 18 years;

• Symptoms that are not due to taking medicines or other drugs, or due to having a medical condition that can cause tics; and

• Not been diagnosed with TS.

**To be diagnosed with a provisional (transient) tic disorder**

To be diagnosed with a provisional (transient) tic disorder, a person must have:

• One or more motor or vocal tics;

• Been present for no longer than 12 months in a row;

• Tics that start before age 18 years;

• Symptoms that are not due to taking medicine or other drugs, or due to having a medical condition that can cause tics; and

• Not been diagnosed with TS or a persistent motor or vocal tic disorder.

In some children, tics may suddenly appear, or suddenly become worse, following a streptococcal (strep) infection, such as strep throat or scarlet fever.

Table 1 summarizes the comparative criteria for the diagnosis of the above three types of tic disorders:

| | TS | Persistent motor or vocal tic disorder | Provisional motor or vocal tic disorder |
|---|---|---|---|
| **Motor; vocal tics** | **2** or more; **1** or more | **2** or more; **1** or more | **1** or more motor or vocal |
| **Tic duration** | At least **1** year | Many times a day for at least **1** year | Less than **1** year |
| **Beginning of tics** | Before age **18** | Before age **18** | Before age **18** |
| **Symptoms** | Not due to medicines, drugs or other condition that can cause tics | Not due to medicines, drugs or other condition that can cause tics | Not due to medicines, drugs or other condition that can cause tics |
| **Other condition(s)** | No underlying neurological symptoms and no intake of certain substances | Not diagnosed with TS | Not diagnosed with TS, or persistent/chronic motor or vocal tic disorder |

**Table 1: Comparative diagnostic criteria of TS, persistent, or provisional tic disorders**

**Timely diagnosis**

According to the Diagnostic & Statistical Manual of Mental Disorders, version 5 (DSM-5) (see Article I in this series), TS may be diagnosed when a person exhibits both multiple motor tics and one or more vocal tics over a period of one 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). Hence, other medical conditions that include tics or tic-like movements—for example, autism or other causes of tics—must be ruled out.

Patients referred for a tic disorder are assessed based on their family history of tics, vulnerability to attention-deficit hyperactivity disorder (ADHD), obsessive–compulsive disorder (OCD), and a number of other chronic medical, psychiatric, and neurological conditions. In individuals with a typical onset and a family history of tics or OCD, a basic physical and neurological examination may be sufficient.

Unfortunately, there are no specific medical or screening tests that can be used to diagnose TS. The diagnosis is usually made based on observation of the individual's symptoms and family history, and after ruling out secondary causes of tic disorders (so-called 'tourettism').

**Delayed diagnosis**

Delayed diagnosis often occurs because professionals mistakenly believe that TS is rare, always involves coprolalia (the spontaneous utterance of obscene words or socially inappropriate and derogatory remarks, or taboo words or phrases), or must be severely impairing. However, since 2000, the DSM had recognized that many individuals with TS do not have significant impairment. Diagnosis of TS does not require the presence of coprolalia or a co-morbid condition such as ADHD or OCD.

**Misdiagnosis**

TS may be misdiagnosed because of the wide expression of its severity, ranging from mild or moderate (in the majority of cases) to severe (the rare but more widely recognized and publicized cases). Further, about 20% of people with TS do not realize that they have tics.

Tics that appear early in the course of TS are often confused with allergies, asthma, vision problems, and other conditions. Pediatricians, allergists, and ophthalmologists are among the first to see or identify a

child as having tics, although the majority of tics are first identified by the child's parents. Coughing, blinking, and tics that mimic unrelated conditions such as asthma are commonly misdiagnosed. An average delay of three years may take place between symptom onset and diagnosis.

**Sudden onset of tic-like behaviors**

Sometimes people have tic-like behaviors that look like tics, but that are distinctly different from those typically seen in TS and other tic disorders. They are often experiencing movement symptoms for the first time with no reported history of tics. These types of tic-like behaviors are more common among teenagers and more often seen in girls than boys. Sometimes these tics can happen in groups of children. Many experts believe these sudden onset tic-like behaviors can happen for different reasons, and tic-like behaviors may need different treatment compared to tic disorders including TS. The best first step is usually to talk to a healthcare provider who is familiar with tics and get a thorough assessment.

## Differential diagnosis

Tics that may appear to mimic those of TS but are associated with other disorders are known as 'tourettism'. 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. They include:

**Abnormal movements associated with chorea, dyskinesia, dystonia, and myoclonus**

Chorea is an abnormal involuntary body movement disorder, one of a group of neurological disorders called dyskinesias. Dystonia is a neurological hyperkinetic movement disorder in which sustained or repetitive muscle contractions result in twisting and repetitive movements or abnormal fixed postures. Myoclonus is a brief, involuntary, non-rhythmic, twitching of a muscle, a joint, or a group of muscles, different from the rhythmic clonus and dyskinesias. These movements are distinct from the tics of TS in that they are more rhythmic, not suppressible, and not preceded by an unwanted urge.

**Developmental and autism spectrum disorder (ASD)**

They may manifest tics, other stereotyped movements, and stereotypic movement disorders. The stereotyped movements associated with autism typically have an earlier age of onset; are more symmetrical, rhythmical, and bilateral; and involve the extremities (for example, flapping the hands). Among individuals with TS studied in clinics, between 2.9% and 20% had ASD, but this may partly be due to difficulties distinguishing between tics and tic-like behaviors or OCD symptoms seen in people with autism.

**Diagnostic confusion between tics and seizure activity**

If another condition might better explain the tics, tests may be done. For example, if there is diagnostic confusion between tics and seizure activity, an EEG may be ordered. Further, an MRI can rule out brain abnormalities, but such brain imaging studies are not usually warranted. In addition:

- **Hypothyroidism:** Measuring thyroid-stimulating hormone (TSH) blood levels can rule out hypothyroidism, which can be a cause of tics.

- **Wilson's disease (WD):** If there is a family history of liver disease, serum, copper, and ceruloplasmin levels can rule out Wilson's disease.

**Stimulants**

The typical age of onset of TS is before adolescence. In teenagers and adults with an abrupt onset of tics and other behavioral symptoms, a urine drug screen for

stimulants might be requested.

**Functional tic-like movements**

They can be difficult to distinguish from tics that have an organic (rather than a psychological) cause. They are "not fully stereotypical", do not respond to medications, do not demonstrate the classic waxing-and-waning pattern of TS tics, and do not progress in the typical fashion in which tics often first appear in the face and gradually move to the limbs. They may occur alone or co-exist in individuals with tic disorders. These tics are inconsistent with the classic tics of TS in the following several ways:

• **Premonitory urge:** Present in 90% of those with tic disorders, premonitory urge is absent in functional tic-like movements;
• **Onset:** It is more abrupt than typical with movements that are more suggestible;
• **Suppressibility:** While seen in tic disorders, it is here lacking;
• **Family or childhood history of tics:** None;
• **Female predominance:** It occurs with a later-than-typical age of first presentation;
• **OCD and ADHD:** They are less co-occurring; and
• **Co-occurring disorders:** There are more of these.

**Other conditions**

Other conditions that may manifest tics include:

1. **Sydenham's chorea (SC)**;
2. **Idiopathic dystonia**; and
3. **Genetic conditions** such as:
    - **Huntington's disease (HD);**
    - **Neuroacanthocytosis;**
    - **Pantothenate kinase-associated neurodegeneration (PKAN);**
    - **Duchenne's muscular dystrophy (DMD);**
    - **Wilson's disease (WD);** and
    - **Tuberous sclerosis (TS).**

**Other possibilities**

Other possibilities include chromosomal disorders such as:

- Down's syndrome (DS);
- Klinefelter's syndrome (KS);
- XYY syndrome; and
- Fragile-X syndrome.

**Acquired causes of tics**

Acquired causes of tics include:

- **Drug-induced tics;**
- **Head trauma;**
- **Encephalitis;**
- **Stroke:** and
- **Carbon monoxide poisoning.**

**Self-injurious behaviors**

The extreme self-injurious behaviors of **Lesch-Nyhan syndrome (LNS)** may be confused with TS or stereotypes, but self-injury is rare in TS even in cases of violent tics.

## Co-occurring conditions

Because people with milder symptoms are unlikely to be referred to specialty clinics, studies of TS have an inherent bias towards more severe cases. When symptoms are severe enough to warrant referral to clinics, ADHD and OCD are often also found. In specialty clinics, 30% of those with TS also have mood or anxiety disorders or disruptive behaviors. There are increased rates of anxiety and depression in those adults with TS who also have OCD.

**ADHD**

In the absence of ADHD, tic disorders do not appear to be associated with disruptive behavior or functional

impairment, while impairment in school, family, or peer relations is greater in those who have more co-morbid conditions. When ADHD is present along with tics, the occurrence of conduct disorder (CD) and oppositional defiant disorder (ODD) increases.

Aggressive behaviors and angry outbursts in people with TS are not well understood; they are not associated with severe tics, but are connected with the presence of ADHD. ADHD may also contribute to higher rates of anxiety, and aggression and anger control problems are more likely when both OCD and ADHD co-occur with TS.

**OCD**

There are two types of OCD: "pure or non-tic-related" OCD and "tic-related" OCD, the latter being hypothesized to be a distinct subgroup of the former and distinguished from it by the type and nature of obsessions and compulsions.

• **Pure OCD:** Compulsions that resemble tics are present in some individuals with OCD and relate to contamination, obsession, and anxiety.

• **Tic-related OCD:** Compulsions are present with more counting, aggressive thoughts, symmetry and touching. They are more likely to be a response to a premonitory urge.

Not all people with TS have ADHD or OCD or other co-morbid conditions, and estimates of the rate of pure TS (or TS-only) vary from 15% to 57%. In clinical populations, a high percentage of those under care do have ADHD. Children and adolescents with pure TS are not significantly different from their peers without TS on ratings of aggressive behaviors or conduct disorders, or on measures of social adaptation. Similarly, adults with pure TS do not appear to have the social difficulties present in those with TS-plus-ADHD.

Among those with an older age of onset, more substance abuse and mood disorders are found, and there may be self-injurious tics. Adults who have severe, often treatment-resistant tics are more likely to also have mood disorders and OCD. Coprolalia is more likely in people with severe tics plus multiple co-morbid conditions.

## Screening for other conditions

Although not all those with TS have co-morbid conditions, most presenting for clinical care exhibit symptoms of other conditions along with their tics. ADHD and OCD are the most common, but ASD or anxiety, mood, personality, oppositional defiant, and conduct disorders may also be present.

Learning disabilities and sleep disorders may be present with reported higher rates of sleep disturbance and migraine than in the general population. A thorough evaluation for co-morbidity is called for when symptoms and impairment warrant, and careful assessment of people with TS includes comprehensive screening for these conditions.

Co-morbid conditions such as OCD and ADHD can be more impairing than tics and cause greater impact on overall functioning. Disruptive behaviors, impaired functioning, or cognitive impairment in individuals with co-morbid Tourette's and ADHD may be accounted for by ADHD, highlighting the importance of identifying co-morbid conditions. Children and adolescents with TS who have learning difficulties are candidates for psycho-educational testing, particularly if the child also has ADHD.

## Neuropsychological Function

There are no major impairments in neuropsychological function among people with TS, but conditions that occur along with tics may cause variation in neurocognitive function. A better understanding of co-

morbid conditions is needed to untangle any neuropsychological differences between TS-only individuals and those with co-morbid conditions.

**Intellectual ability, attentional ability, and nonverbal memory**

Only slight impairments are found in intellectual ability, attentional ability, and nonverbal memory—but ADHD, other co-morbid disorders, or tic severity could account for these differences.

**Visuomotor and visuoconstructive skills**

In contrast with earlier findings, visuomotor integration and visuoconstructive skills are not found to be impaired, while co-morbid conditions may have a small effect on motor skills.

**Verbal fluency**

Co-morbid conditions and severity of tics may account for variable results in verbal fluency, which can be slightly impaired.

**Social cognition**

There might be slight impairment in social cognition, but not in the ability to plan or make decisions. Children with TS-only do not show cognitive deficits. They are faster than average for their age on timed tests of motor coordination, and constant tic suppression may lead to an advantage in switching between tasks because of increased inhibitory control.

**Learning disabilities**

Learning disabilities may be present, but whether they are due to tics or to co-morbid conditions is controversial. Older studies that reported higher rates of learning disability did not control well for the presence of co-morbid conditions. There are often difficulties with handwriting, and disabilities in written expression and math are reported in those with TS plus other conditions.

## Management of Symptoms

As already indicated, there is no cure for TS, no one effective medication for treating all symptoms, and no single most effective medication. Further, most medications prescribed for tics have not been approved for that use (they have been repurposed), and no medication is without the risk of significant adverse effects. Treatment is therefore initially focused on identifying the most troubling or impairing symptoms and helping the individual manage them. Because co-morbid conditions are often a larger source of impairment than tics, they are a priority in treatment.

People with TS 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. Even children with milder tics may be angry, depressed or have low self-esteem as a result of increased teasing, bullying, rejection by peers or social stigmatization, and this can lead to social withdrawal. Some children feel empowered by presenting a peer awareness program to their classmates. It can be helpful to educate teachers and school staff about typical tics, how they fluctuate during the day, how they impact the child, and how to distinguish tics from naughty behavior.

By learning to identify tics, adults can refrain from asking or expecting a child to stop ticking, because tic suppression can be exhausting, unpleasant, attention-demanding, and can result in a subsequent rebound bout of tics. Adults with TS may withdraw socially to avoid stigmatization and discrimination because of their tics. Depending on their country's healthcare system, they may receive social services or help from support groups.

**Behavioral therapy (BT)**

Behavior problems can be highly disruptive for the child and others in the child's life. It is important to get a diagnosis and treatment plan from a mental health professional as soon as possible. Effective treatments for disruptive behaviors include behavior therapy training for parents. In this regard, BT is a treatment that teaches people with TS ways to manage their tics. It is not a cure for tics. However, it can help reduce the number of tics, their severity, their impact, or a combination of all of these. It is important to understand that even though BT might help reduce the severity of tics, this does not mean that tics are just psychological or that anyone with tics should be able to control them.

**Habit reversal training (HRT)**

HRT is one of the most studied behavioral interventions for people with tics. It has two main parts:

• **Awareness training (AT):** Here, people identify each tic out loud.

• **Competing response training (CRT):** People learn to do a new behavior that cannot happen at the same time as the tic. For example, if the person with TS has a tic that involves head rubbing, a new behavior might be for that person to place their hands on their knees, or to cross their arms so that the head rubbing cannot take place.

**Comprehensive behavioral intervention for tics (CBIT)**

For many years, medication was the only real treatment option for children and adults with TS. However, whereas medication can help control tics, it does not always work and can cause side effects that make people feel bad and can lead to other health problems. Working with the Tourette Association of America (TSA), researchers sought an alternative treatment that does not use medication. They begun developing and testing a promising treatment option called comprehensive behavioral intervention for tics (CBIT), which has been compared to rehabilitation after a stroke. The first large study of CBIT in children was published in 2010. During the past decade, work to learn more about this treatment, and to make it more widely available, has continued.

CBIT is an evidence-based type of behavioral therapy for TS and chronic tic disorders. It includes habit reversal in addition to other strategies, including education about tics and relaxation techniques. It teaches a person to become aware of their behavior and helps them change how they behave. It is very systematic and carefully planned, depending on each person's specific needs and symptoms, and typically takes place in a therapist's office. It has been compared to rehabilitation after a stroke.

CBIT procedures combine elements of habit reversal training (HRT) with psycho-education and function-based behavioral interventions. Results of a child study showed that 52.5% of the children who received CBIT showed significant symptom improvement and 38% for adults. Improvement can be sustained for at least 6 months after the end of the treatment. During CBIT, the therapist helps individuals with tics learn to: Become more aware of their tics and the urge to tic; carefully choose and practice a new behavior instead of the tic. This "new behavior" (competing response) helps reduce and, in some cases, even eliminate the tic; identify situations that can make tics worse and find ways to change them; and learn ways to prevent or cope with stress.

When this behavioral treatment was first developed, health professionals were concerned that it could cause more misunderstandings about tics. Just because tics can be managed by changing behavior does not mean that tics are done by choice.

During the past decade, there has been increasing evidence that CBIT can be effective at reducing tic symptoms and tic-related impairment among children and adults with TS. Experts now suggest using it as the first approach to treatment because its effectiveness is similar to medications but with fewer side effects. But, the evidence also shows that CBIT is not a "cure" for TS and does not help everybody; rather, it is a tool that, when used appropriately, can help many people with TS to manage their tics better and reduce the impact that tics may have on their lives. Researchers continue to learn more about how and for whom this treatment is most effective.

In CBIT, a therapist will work with a child (and his/her parents) or an adult with TS to better understand the types of tics the person is having and to understand the situations in which the tics are at their worst. Changes to the surroundings may be made, if possible, and the person with TS will also learn to do a new behavior instead of the tic (habit reversal training, HRT). For example, if a child with TS often has a certain tic during math class, the math teacher can be educated about TS, and perhaps the child's seat can be changed so that the tics are not as visible. In addition, the child also can work with a psychologist to learn HR techniques. This helps to decrease how often the tic occurs by doing a new behavior (like putting their hands on their knees when an urge to perform the tic happens). CBIT skills can be learned with practice, with the help of an experienced therapist, and with the support and encouragement of those close to the person with TS.

In recent years, more health professionals have recognized that CBIT can be very effective in managing the symptoms of TS. So far, few clinicians have been trained in these types of treatments specifically for TS and tic disorders. The CDC&P and the TSA have been working to educate more health professionals in this approach to managing TS symptoms.

**Who can benefit from learning CBIT?**

CBIT is most appropriate for practitioners with behavioral and clinical expertise, including physicians, psychologists, behavioral therapists, occupational therapists, social workers, and other allied health professionals.

**Is CBIT for everybody who has TS or tics?**

It is important to understand that not everyone will benefit from CBIT. Teaching this technique to children with untreated ADHD may be more difficult because of problems with focusing, impulsivity, and low frustration tolerance. Likewise, any other psychiatric or social problem that gets in the way of participating in treatment may make CBIT more difficult. CBIT can be used without treating co-occurring conditions, but it is recommended that patients address any co-occurring conditions with an evaluation. These conditions or symptoms may hinder their ability to practice CBIT. The Tourette Association of America (TAA) offers two options for practitioners to explore CBIT: No-Cost CBIT Overview Workshops and Tourette Syndrome Behavior Training Institutes (TS-BTIs):

• **TS education and training:** Diagnosis and treatment of people with TS can be improved when health care providers and educators have up-to-date and accurate information about this disorder. Education and training for these professionals will help them to better identify, diagnose, and refer for treatment people with TS. Educating the community (for example, friends and family, educators, and coworkers) about TS can increase understanding of the symptoms, reduce teasing, and decrease stress for people living with TS. People with TS cannot help having tics, and are not being disruptive on purpose. When others understand these facts, people with TS might receive more support, which might help lessen some tic symptoms. Many organizations including the CDC&P's National Center on Birth Defects and Developmental Disabilities

(NCBDDD) are committed to raising awareness about TS and to improving the education of those who care for and work with people living with TS. In doing so, they support the following educational activities.

• **Tourette health and education outreach (THEO):** CDC&P 's NCBDDD has partnered with the TAA to provide much needed educational programs. The programs educate physicians, allied professionals, and school personnel, as well as those who have TS, their families, and the general public about TS. The programs provide accurate, up-to-date, science-based information about the recognition, diagnosis, and treatment of TS. The goal of this outreach is to increase awareness and provide information that will help people with TS receive needed health services, be more accepted by those around them, and have the opportunity to succeed in school and at work.

The CDC-TSA partnership also has a special focus on under-served communities, providing educational programs in all geographical areas and reaching out to organizations that serve cultural minorities or communities that are at risk for health disparities. It has conducted more than 1,350 education programs for professionals and community members in all 50 U.S. States as well as in Washington, D.C., the U.S. Virgin Islands, Puerto Rico, St. Maarten, and Canada. Additionally, more than 50 programs were held virtually with no state affiliation.

**Education and outreach: Increasing online support and telehealth**

CDC&P supported 120 CBIT programs with about 3,700 attendees. TSA also supported 33 training institutes for CBIT with close to 300 practitioners becoming licensed to provide CBIT to their patients. With limited options for direct access to training and treatment in the current environment, there is more need than ever to find ways to use telehealth approaches. For this purpose, TSA has worked with experts to identify ways to provide teletraining to professionals who want to learn about CBIT and also assist professionals who have been trained on CBIT so that they can learn to provide the therapy to their clients via telehealth. In the long term, telehealth can help families who live in rural areas, who have difficulty finding time to visit with a provider, or who have trouble finding a trained provider by giving them options for accessing treatment.

## Aggressive acts of children with compulsive disorder (CD)

Children with CD act aggressively toward others and break rules, laws, and social norms. They might have more injuries and difficulty with friends. In addition, the symptoms of CD happen in more than one area in the child's life (for example, at home, in the community, and at school).

**Rage**

Some people with TS have anger that is out of control, or episodes of "rage." Rage that happens repeatedly and is disproportionate to the situation that triggers it may be diagnosed as a mood disorder, like intermittent explosive disorder (IED). Symptoms might include extreme verbal or physical aggression. Examples of verbal aggression include extreme yelling, screaming, and cursing. Examples of physical aggression include extreme shoving, kicking, hitting, biting, and throwing objects. Rage symptoms are more likely to occur among those with other behavioral disorders such as ADHD, oppositional defiant disorder (ODD), or CD.

Among people with TS, symptoms of rage are more likely to occur at home than outside the home. Treatment can include behavior therapy, learning how to relax, and social skills training. Some of these methods will help individuals and families better understand what can cause the symptoms of rage, how to avoid encouraging these behaviors, and how to use

appropriate discipline for these behaviors. In addition, treating other behavioral disorders that the person might have, such as ADHD, ODD, or CD can help to reduce symptoms of rage.

**Anxiety**

There are many different types of anxiety disorders with many different causes and symptoms. These include generalized anxiety disorder, OCD, panic disorder, post-traumatic stress disorder (PTSD), separation anxiety, and different types of phobias. Separation anxiety is most common among young children. These children feel very worried when they are apart from their parents.

**Depression**

Everyone feels worried, anxious, sad, or stressed from time to time. However, if these feelings do not go away and they interfere with daily life (for example, keeping a child home from school or other activities, or keeping an adult from working or attending social activities), a person might have depression. Having either a depressed mood or a loss of interest or pleasure for at least weeks might mean that someone has depression. Children and teens with depression might be irritable instead of sad.

To be diagnosed with depression, other symptoms also must be present, such as: Changes in eating habits or weight gain or loss; changes in sleep habits; changes in activity level (others notice increased activity or that the person has slowed down); less energy; feelings of worthlessness or guilt; difficulty thinking, concentrating, or making decisions; repeated thoughts of death; and thoughts or plans about suicide, or a suicide attempt. Depression can be treated with counseling and medication.

**Other health concerns**

Children with TS can also have other health conditions that require care. Among the more common health conditions that can occur with TS are: Asthma; hearing loss or vision problems; bone, joint, or muscle problems; and brain injury or concussion.

A CDC&P study showed that the rates of asthma and hearing or vision problems were similar to children without TS, but bone, joint, or muscle problems as well as brain injury or concussion were higher for children with TS. Children with TS were also less likely to receive effective coordination of care or have a medical home, which means a primary care setting where a team of providers provides health care and preventive services.

**Educational concerns**

As a group, people with TS have levels of intelligence similar to those of people without TS. However, people with TS might be more likely to have learning differences, a learning disability, or a developmental delay that affects their ability to learn.

Many people with TS have problems with writing, organizing, and paying attention. They might have problems processing what they hear or see. This can affect the person's ability to learn by listening to, or watching, a teacher. Or, the person might have problems with their other senses (such as how things feel, smell, taste, and movement) that affect learning and behavior. Children with TS might have trouble with social skills that affect their ability to interact with others.

As a result of these challenges, children with TS might need extra help in school. Many times, these concerns can be addressed with accommodations and behavioral interventions (for example, help with social skills). Accommodations can include things such as providing a different testing location or extra testing time, providing tips on how to be more organized, giving the child less homework, or letting the child use a

computer to take notes in class. Children also might need behavioral intervention therapy, or they may need to learn strategies to help with stress, paying attention, or other symptoms.

## Algorithm for the treatment of patients with TS

Symptoms management is individualized and involves shared decision-making between the clinician, patient, family, and caregivers. It will be helpful to follow the various steps outlined in the flow chart of Figure 1.

**Step 1: Evaluation and identification of tic disorder**

Through a comprehensive evaluation, sometimes involving consultation with a pediatrician or a pediatric neurologist, a child and adolescent psychiatrist can determine whether a child or adolescent has TS or another tic disorder. A child's parents are typically the first to notice their tics; they may feel worried, imagine that they are somehow responsible (they are in part, genetically), or feel burdened by misinformation about TS. Effectively, educating parents about the diagnosis and providing social support can ease their anxiety. This support can also lower the chance that their child will be unnecessarily medicated or experience an exacerbation of tics due to their parents' emotional state.

For students with TS, although they often function well in the regular classroom, co-morbid conditions such as ADHD, learning disabilities, obsessive-compulsive symptoms, and frequent tics can greatly interfere with their academic performance or social adjustment. After a comprehensive assessment, they should have access to an educational setting that meets their individual needs.

They may require tutoring, smaller or special classes, private study areas, exams outside the regular classroom, other individual performance accommodations and, in some cases. special schools.

**Step 2: Psychoeducation and social support**

Psychoeducation, i.e., knowledge, education, reassurance, and understanding, is uppermost in management plans for tic disorders and often sufficient for the majority of cases. In particular, psychoeducation targeting the patient and their family and surrounding community is a key management strategy.

**Step 3: Indication for treatment?**

Step # 2 will determine whether treatment, if any, is indicated. If not, monitoring the individual with tics may be sufficient. “Watchful waiting" may be an acceptable approach for those who are not functionally impaired.

**Step 4: Have co-morbid disorders treatment priority?**

If treatment is indicated, have co-morbid disorders and tics have priority? If not, proceed with treatment approach (step 5). Otherwise, provide treatment of co-morbidities and tics.

**Step 5: Have behavioral and psychobehavioral (PBT) therapy preference?**

If not, proceed with treatment approach (step 6) with preference for behavioral therapy (BT). Otherwise, in the absence of co-morbid disorders, tic treatment is indicated with preference for (and availability of) behavioral and psychobehavioral therapy. Having been shown to be effective, behavioral therapies using habit reversal training (HRT) and exposure and response prevention (ERP) will be first-line interventions in the management of TS. In HRT, the child learns to develop greater awareness of the tics and comes up with a different behavior to take its place.

Because tics are somewhat suppressible, when people with TS are aware of the premonitory urge that precedes them, they can be trained to develop a response to the urge that competes with the tics. Comprehensive behavioral intervention for tics (CBIT) is based on HRT, the best researched behavioral therapy for tics.

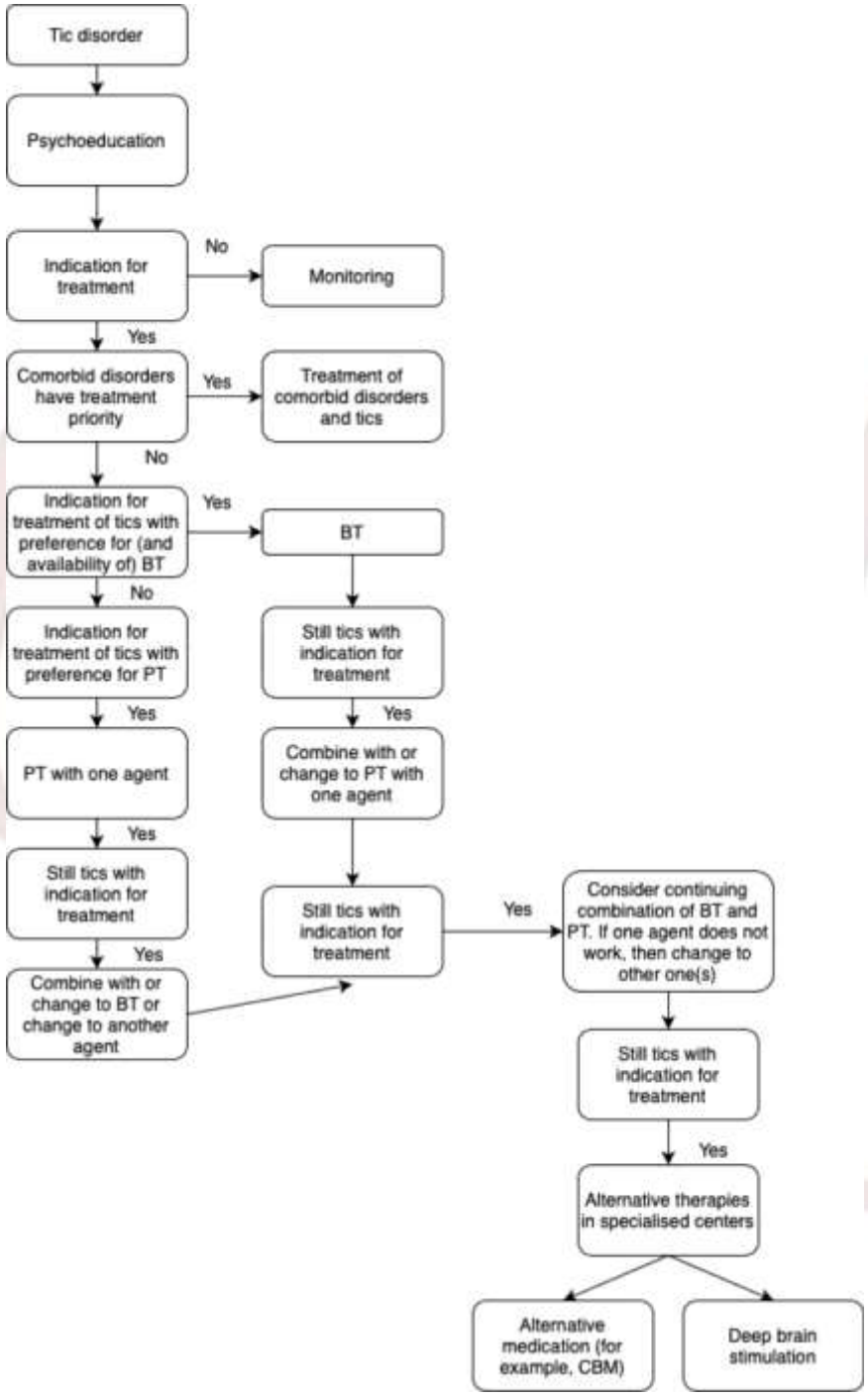


**Figure 1: Algorithm for behavioral and psychobehavioral therapy of patients with TS**
**based on shared clinician patient decision making**

*Key:* BT: Behavior therapy; CBM: Cannabis-based medicine; PT: Pharmacotherapy
*Reference: Kirsten, Müller-Vahl, Szejko et al. (2022). (Reproduced by permission)*

However, TS experts still debate whether increasing a child's awareness of tics with HRT/CBIT (as opposed to ignoring tics) can lead to more tics later in life.

When disruptive behaviors related to co-morbid conditions exist, anger control training, and parent management training can be effective. Cognitive behavioral therapy (CBT) is a useful treatment when OCD is present. Relaxation techniques, such as exercise, yoga, and meditation may be useful in relieving the stress that can aggravate tics. Beyond HRT, the majority of behavioral interventions for TS (for example, relaxation training, and biofeedback) have not been systematically evaluated and are not empirically supported.

**Step 6: Preference for psychotherapy (PT)**

**Step 7: Psychotherapy (PT) with one agent**

If BT is not effective, PBT may be combined with, or changed to, PT with one agent.

**Step 8: Still indication for tic treatment**

If yes, proceed with treatment approach (step 9).

**Step 9: Combine with or change to BT or to another agent**

**Step 10: Consider continuing combination of BT with PT**

If one agent does not work, then change to other agent(s).

**Step 11: Indication for treatment still needed**

If tics still indicate the need for treatment, then proceed with the next step.

**Step 12: Possible application of alternative therapies**

Such alternative therapies should be provided in specialized centers.

**Step 13: Alternative therapies**

Alternative therapies may consist of alternative medications, for example CBM, or neurostimulation such as deep brain stimulation (DBS) that will be considered more closely in Article III in this series.

## Clinical practice guidelines

Clinical practice guidelines for the treatment of tics were separately published by the American Academy of Neurology (AAN), the European Union (EU), and the World Health Organization (WHO). I will review here these guidelines, particularly for the management of symptoms of TS, reserving Article II in this series for treatment options *per se*.

**The AAN guidelines**

In 2019, the AAN's Guideline Committee published a practice guidelines document for the treatment of tics in people with TS and chronic tic disorders. I provide in this section only a synopsis of it and urge the interested reader to consult the complete document, which can be found as a data supplement at links.lww.com/WNL/A882.

As a preamble, it must be emphasized that the treatment of tics should be individualized and prioritized, particularly since many people with tic disorders may also present psychiatric co-morbidities. It should be attained on the basis of collaborative discussions made among clinicians, patients, and caregivers. The recommended guidelines also considered the questions as to when should clinicians and patients pursue treatment and, if treatment is deemed required, how should clinicians and patients

elect the most pertinent options and in what combination or sequence it should be administered.

When the benefit-to-harm ratio is favorable for any given intervention, three recommendation designations are provided (**A** for 'must'; **B** for 'should'; and **C** for 'may'), each designation denoting the strength level of the said recommendation, A being relatively the strongest and C the weakest.

Thirteen categories of recommendations have been formulated. Here, I will only summarize the first seven of them that relate to symptoms management. They are summarized in Tables 2 through 8, respectively for counseling, psychoeducation, assessment and treatment of ADHD in children with tics, assessment and treatment of OCD in children with tics, other psychiatric co-morbidities, assessment of tic severity and treatment expectations, and behavioral treatments. The other six recommendations for pharmacological treatment and brain neurostimulation will be reviewed in Article III in this series.

| Recommendation # | Level | Clinician's role |
|---|---|---|
| **1a** | A | Must inform patients and their caregivers about the natural history of tic disorders |
| **1b** | A | Must evaluate functional impairment related to tics from the perspective of the patient and, if applicable, the caregiver |
| **1c** | B | Should inform patients and caregivers that watchful waiting is an acceptable approach in people who do not experience functional impairment from their tics |
| **1d** | C | May prescribe CBIT as an initial treatment option relative to watchful waiting for people with tics who do not experience functional impairment if they are motivated to attempt treatment |
| **1e** | A | Must periodically re-evaluate the need for ongoing medical treatment (For physicians prescribing medications for tics) |

**Rationale:** Providing information about the natural history of TS to help inform treatment decisions.

## Table 2: Counseling recommendations

| Recommendation # | Level | Clinician's role |
|---|---|---|
| **2** | B | Should refer people with TS to resources for psychoeducation for teachers and peers, such as the Tourette Association of America (TAA) |

**Rationale:** Psychoeducation about TS (a) with peers, can result in more positive attitudes toward a person with TS; (b) with teachers, can improve knowledge about the condition; and (c) may positively affect people with TS by improving peers' attitudes about, and teachers' knowledge of, TS.

## Table 3: Psychoeducation recommendations

| Recommendation # | Level | Clinician's role |
|---|---|---|
| **3a** | B | Should ensure an assessment for co-morbid ADHD is performed |
| **3b** | B | Should evaluate the burden of ADHD symptoms |
| **3c** | B | Should ensure appropriate ADHD treatment is provided in cases of functionally impairing ADHD |

**Rationale:** Co-morbid ADHD is common in people with TS. Several trials have specifically addressed the medical treatment of both ADHD and tics in children with both disorders. While ADHD symptoms may improve in adolescence, adults with TS may require ongoing care for this co-morbidity.

**Table 4: Assessment and treatment of ADHD in children with tics**

| Recommendation # | Level | Clinician's role |
|---|---|---|
| **4a** | B | Clinicians should ensure an assessment for co-morbid OCD is performed in people with tics |
| **4b** | B | In people with tics and OCD, clinicians should ensure appropriate OCD treatment is provided |

**Rationale:** OCD behaviors are common in people with TS. Children with tics may not respond as well as those without tics to selective serotonin re-uptake inhibitors (SSRIs). Cognitive behavioral therapy (CBT) is considered a first-line treatment of OCD in individuals with tic disorders.

**Table 5: Assessment and treatment of OCD in children with tics**

| Recommendation # | Level | Clinician's role |
|---|---|---|
| **5a** | A | Clinicians must ensure appropriate screening for anxiety, mood, and disruptive behavior disorders is performed in people with tics |
| **5b** | A | Clinicians must inquire about suicidal thoughts and suicide attempts in people with TS and refer to appropriate resources if present |

**Rationale:** People with TS are at high-risk of other psychiatric co-morbidities, including anxiety disorders, oppositional defiant disorder, and mood disorders. Co-morbid mood disorders appear more prevalent in adolescents and adults than children and in those with greater tic severity. There is an increased risk of dying by suicide and attempting suicide in people with TS. Persistence of tics beyond young adulthood, previous suicide attempts, and co-morbid personality disorders increase the risk of death by suicide.

**Table 6: Other psychiatric co-morbidities**

| Recommendation # | Level | Clinician's role |
|---|---|---|
| **6a** | C | May measure tic severity using a valid scale to assess treatment effects |
| **6b** | A | Must counsel patients that treatments for tics infrequently result in complete cessation of tics |

**Rationale:** Several rating scales are available for measuring tic severity, the Yale Global Tic Severity Scale (YGTSS) being the most extensively deployed and validated. The use of validated scales to measure tic severity can aid the evaluation of treatment response in the clinical setting. While medications, behavioral therapy, and neurostimulation can result in meaningful reduction in tics, these interventions rarely result in complete cessation of tics.

## Table 7: Assessment of tic severity and treatment expectations

| Recommendation # | Level | Clinician's role |
|---|---|---|
| **7a** | B | Should prescribe CBIT as an initial treatment option relative to other psychosocial/behavioral interventions |
| **7b** | B | Should offer CBIT as an initial treatment option relative to medication |
| **7c** | C | May prescribe CBIT delivered over teleconference or secure voice over Internet protocol delivery systems if face-to-face options are unavailable. If CBIT is unavailable, other behavioral interventions for tics may be acceptable, such as exposure and response prevention |

**Rationale:** People with tics receiving CBIT are more likely than those receiving psychoeducation and supportive therapy to have reduced tic severity. Given the effort required from patients or their families, along with its benign safety profile, CBIT is an acceptable intervention for people with tics that lead to psychosocial or physical impairment and who are motivated to participate in treatment.

## Table 8: Behavioral treatments

**The E.U. Guidelines**

In 2022, the European Union (EU) issued its clinical practice guidelines for the treatment of patients with TS. Despite some overlap with the AAN guidelines, there are notable differences regarding the assessment and interventions in clinical practice. This is reflected by differences in: (i) health care organization and use, (ii) patient preferences, (iii) first choice of pharmacological agents, (iv) availability and application of behavioral treatments, (v) costs of treatment, and (vi) approval status.

The EU guidelines have incorporated the newly implemented DSM-5 and the WHO's ICD-11 criteria (see below). In the DSM-5, TS is classified as a "neurodevelopmental disorder" alongside ADHD, intellectual disabilities, and autism spectrum disorder (ASD). In contrast, in the ICD-11, tic disorders have been assigned to 'movement disorders'. However, in the opinion of some clinician researchers, this "disregards the growing body of both genetic and clinical evidence that tic disorders are related to developmental and psychiatric disorders rather than to neurological disorders … furthermore, there is no scientific evidence to support the introduction of infectious or

post-infectious tics" … and, still further, do not recommend assessment of children for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections (PANDAS)-related TS".

With respect to behavioral interventions, in Part II of the EU Guidelines - Psychological interventions, substantial progress has been made since 2011. Most importantly, since then, several randomized controlled trials (RCTs) have been published on habit reversal treatment (HRT) in both children and adults. As a result, behavioral treatment is currently regarded as the symptomatic treatment of choice in reducing tics. Further, internet-based modules of established behavioral treatments have been developed to render behavior therapy more accessible. In addition, adaptations have been made to broaden the focus of behavioral treatment from reducing tic severity only to improving the individual's overall quality of life.

**The WHO's ICD-11 guidelines**

The WHO's International Classification of Diseases, version 11 (ICD-11) includes a chapter, Chapter 06 for "Mental, behavioral or neurodevelopmental disorders". These disorders are syndromes characterized by clinically-significant disturbances in an individual's cognition, emotional regulation, or behavior that reflect a dysfunction in the psychological, biological, or developmental processes that underlie mental and behavioral functioning. The disturbances are usually associated with distress or impairment in personal, family, social, educational, occupational, or other important areas of functioning. Chapter 06 excludes acute stress reaction and uncomplicated bereavement.

## Summary and Conclusions

There are no specific screening or medical tests that can be used for the diagnosis of TS, which remains clinically-based on the combination of signs and symptoms and medical history. There are no blood, laboratory, or imaging tests needed for diagnosis. Three criteria are employed by health professionals to diagnose TS or other tic disorders (presence of motor and vocal/phonic tics; waxing-and-waning of tics; and tics not caused by medical conditions or medications, other than the use of substances/toxicants). Delayed diagnosis often occurs because professionals mistakenly believe that TS is rare, always involves coprolalia, or must be severely impairing. TS may also be misdiagnosed because of the wide expression of severity of tics. Other tics (categorized as tourettism) may appear to mimic those of TS but are associated with other disorders. When symptoms are severe enough to warrant referral to clinics, ADHD and OCD are often found as co-occurring conditions.

Symptoms management may include behavioral, psychological, and pharmacological therapies to help control the symptoms, and a type of behavioral therapy called 'habit reversal training'. Pharmacological intervention is reserved for more severe symptoms. Psychotherapy (PT) or cognitive behavioral therapy (CBT) may ameliorate depression and social isolation, and improve family support. The decision to use behavioral or pharmacological treatment is "usually made after the educational and supportive interventions have been in place for a period of months, and it is clear that the tic symptoms are persistently severe and are themselves a source of impairment in terms of self-esteem, relationships with the family or peers, or school performance. The child and adolescent psychiatrist can also advise families about how to provide emotional support and the appropriate educational environment for their child or adolescent.

For many years, medication was the only real treatment option for children and adults with TS. However, while medication can help control tics, it does not always work and can cause side effects that make people feel bad and can lead to other health problems. Researchers developed and tested a promising treatment option called comprehensive behavioral intervention for tics

(CBIT), a behavioral therapy treatment that teaches people with TS ways to manage their tics. It is not a cure for tics but it can help reduce the number of tics, their severity, their impact, or a combination of all of these. It is important to understand that even though behavioral therapies might help reduce the severity of tics, this does not mean that tics are just psychological or that anyone with tics should be able to control them.

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