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StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan-.

Tourette Syndrome And Other Tic Disorders

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Last Update: July 19, 2021.

Continuing Education Activity

Tourette syndrome, also referred to as Tourette disorder, is a common neurodevelopmental disorder affecting up to 1 percent of the population. It is characterized by multiple motor and vocal tics that begin in childhood. Children with Tourette syndrome may have additional comorbid neuropsychiatric conditions such as attention deficit hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD), anxiety disorder, and oppositional defiant disorder (ODD). The prognosis for Tourette syndrome is generally positive, as tics typically remit by adulthood, but other neuropsychiatric conditions may persist. This activity reviews the evaluation and management of patients with Tourette syndrome and other tic disorders and highlights the role of interprofessional team members in collaborating to provide well-coordinated care and enhance outcomes for affected patients.

Objectives:

- Identify the etiology of Tourette syndrome.
- Describe the population at risk for developing Tourette syndrome.
- Explain the management strategies for Tourette syndrome.
- Explain the importance of improving coordination amongst the interprofessional team to enhance the delivery of care for patients affected by Tourette syndrome.

Access free multiple choice questions on this topic.

Introduction

Tourette syndrome referred to as Tourette disorder in the recently updated *Diagnostic and Statistical Manual of Mental Disorders (DSM–5)*, is a common neurodevelopmental disorder affecting up to 1% of the population. It is characterized by multiple motor and vocal tics and starts in childhood. Children with Gilles de la Tourette syndrome (GTS) sometimes experience physical pain, social isolation, emotional disturbance, and are at risk for underachievement. It is difficult to distinguish whether these experiences are the result of the tics themselves or the fact that most children with Tourette syndrome also have other comorbid neuropsychiatric conditions such as attention deficit hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD), anxiety disorder, or oppositional defiant disorder (ODD). The pathophysiology and genetic basis for GTS are not well understood despite many in-depth studies. Pharmacological treatment is not usually indicated, as children respond well to comprehensive behavioral intervention for tics (CBIT). Teacher and caregiver education can have a positive impact on the social experience of a child struggling with Tourette syndrome. The prognosis is variable, as tics typically remit by adulthood, but the other neuropsychiatric conditions may persist.[1][2][3][4]

Etiology

In his initial paper, Georges Gilles de la Tourette reported that the condition aggregated in families. Since then, researchers have learned that children with GTS have a family history 52% of the time. They have a first-degree relative with GTS at a rate 10 times the general population, and the ratio of concordance in monozygotic twins is 5:1 compared to dizygotic twins. Tourette syndrome has demonstrated to be one of the most heritable, non-Mendelian,

neuropsychiatric disorders. However, multiple large studies have yet to identify a causal gene. GTS appears to be highly polygenic with environmental factors affecting ideographic phenotype. There have been some replicated studies identifying gene abnormalities that lead to disruption of the cortico-striato-thalamo-cortical circuits. Mounting evidence also points to significant environmental contributors such as infection, autoimmunity, or perinatal injury.

Epidemiology

Over the last 30 years, many studies have attempted to determine the prevalence of Tourette syndrome. The large variation in methodology has led to a reported prevalence between 3:1000 and 8:1000 children. Males are more commonly affected than females with a ratio of 3:1 to 4:1. The Center for Disease Control and Prevention (CDC) in the United States reports that GTS is twice as common in Caucasian children as compared to Hispanic or African-American children, but at this time it is not clear whether differences in access to care bias this figure.

Pathophysiology

Motor and vocal tics have 3 components:

1. Premonitory urge,
2. Physical expression of the tic,
3. Sense of relief experienced afterward

Further, there are exacerbating factors such as anxiety and voluntary suppression that influence the expression of tics. These various components involve multiple circuits in the brain, which connect the frontal cortex to subcortical structures (thalamus and basal ganglia).

Antipsychotics have been shown to reduce tics, pointing toward a major role for dopamine in GTS. Dopamine receptors are involved in both the excitatory and inhibitory pathways of the basal ganglia as well as the frontal cortex and ventral striatum. Theories suggest abnormalities in dopamine pathways at the presynaptic, intrasynaptic, and postsynaptic levels, but additional research is needed for further clarification. GABA (gamma-aminobutyric acid) disruption has been demonstrated in pathological post-mortem studies of Tourette syndrome patients. This disruption may result in the disinhibition of cortico-basal ganglia loops. Other neurotransmitters have been studied including glutamate, acetylcholine, serotonin, noradrenaline, and histamine and been found to have varying degrees of evidence supporting involvement in GTS.

History and Physical

Motor tics are sudden, rapid, recurrent, nonrhythmic stereotyped motor movements, generally preceded by an urge. They can affect any part of the body but are by far most common in the face, head, and neck region. Vocal tics include any tic that produces noise: sniffing, grunting, humming, clicking, yelling words repetitively. Coprolalia, shouting expletives or other obscenities, affects less than 10% of patients with GTS.[5][6][7][8]

Criteria for Tourette Disorder

Diagnostic and Statistical Manual of Mental Disorders, Fifth Ed. (DSM-5)

- Multiple motor tics and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently
- The tics may wax and wane in frequency but have persisted for more than 1 year since the onset of the first tic
- Onset is before age 18
- The disturbance is not attributable to the physiological effects of a substance or another medical condition

The DSM-5 also lists criteria for persistent (chronic) motor or vocal tic disorder, which includes all criteria for GTS except that the child has only had either motor or vocal tics, not both. Provisional tic disorder meets all criteria for GTS except that the tics have not lasted longer than a year. There is no clear pathophysiological difference between these 3 disorders.

The typical age of tic onset is 4 to 6 years old, with symptoms peaking around 10 to 12 years of age. Simple motor tics are typically diagnosed early, while more complex tics tend to develop later. The individual tics have a rapid onset, occurring multiple times daily. They escalate over the next several days to weeks. Then, they plateau for a variable amount of time, weeks to years, before gradually tapering to a stop. For each patient, different tics can persist for a variable amount of time, and often there is overlap with multiple different types of tics occurring concurrently.

Most patients report a premonitory urge, which they usually describe as a vague sense that they need to perform the movement, followed by a sense of relief after the tic. About 20% of patients report a sensory component with the urge, usually as itching, tingling, or aching. Patients consistently report that this premonitory urge is the factor that is most bothersome in Tourette syndrome. Younger children do not report this urge as frequently as older children for reasons that are not clearly understood. The movements themselves are involuntary in the sense that they are mostly subconscious and a patient cannot typically make them stop completely, but they are under voluntary influence. Most patients report the ability to suppress their tics for varying lengths of time. However, this active suppression can take a tremendous amount of focus and energy. Patients report that suppressing tics causes the premonitory urge to build to a level that can be quite distressing. Some patients are hesitant to attempt to suppress tics because it can lead to worsening tics or a sense that they lose control of their tics. We call this phenomenon “purging.” Parents often report that children have very frequent motor and vocal tics upon arrival home from school, but when they confer with the child’s teacher, they have not been having a problem at school. This is an adaptive coping strategy children learn, and parents should be encouraged to make the home a safe place for a child to freely release the tics.

Children with Tourette syndrome have high rates of comorbid neurobehavioral disorders. ADHD or OCD is seen in most patients. Children with GTS often experience a lot of anxiety, sleep abnormalities, poor impulse control, or other behavioral disorders. These comorbid neurobehavioral disorders often cause more psychosocial impairment for the child than the severity of the tics.

Echophenomenon is present in over half of children with GTS. Echolalia refers to repeating words, while echopraxia is repeating movements. This is prevalent in Tourette syndrome patients who have comorbid OCD. This can become a problem when dealing with authority figures, like teachers or police officers, who do not understand GTS.

Evaluation

Detailed history and physical examination by an experienced clinician can lead to an appropriate diagnosis. Neurologic examination should be normal other than the presence of motor or vocal tics. History or exam findings that would raise concern for a different diagnosis include a change in the child’s cognition, tics occurring while the child is asleep, or constant movement. The Yale Global Tic Severity Scale is the gold standard validated instrument that is used in most Tourette studies and can be used in the clinic. It is important to evaluate for evidence of comorbid conditions as well, including ADHD, OCD, anxiety disorder, or other behavioral problems. Often, these are quite disruptive for the child’s social functioning. Treatment of these conditions can indirectly lead to a reduction in tic frequency and severity.[9][10][11]

Currently, there is no specific laboratory or genetic test available to diagnose Tourette Syndrome. Brain MRI or CT scan is typically normal. Recent MRI studies recently have shown subtle reduced caudate volume in patients with GTS, and the degree of this volume loss correlates to OCD symptoms. However, these results were obtained by rigorously detailed measuring, and the techniques are not currently available to most community-based providers.

Even when children seem to have tics under good control at school or in other social environments, they may be expending a considerable amount of mental effort to do so. This can interfere with their ability to concentrate on lessons or fully participate in conversations. Asking questions to gauge this mental aspect can help a provider have a clearer understanding of how disruptive GTS is for the patient.

Treatment / Management

Often, the primary reason families of children with mild GTS request consultation is concern about the possibility of another etiology for the tics. Identifying the family’s goals for a visit is an important first step, as reassurance of the correct diagnosis may be all that is needed. Thoroughly educating the child and family about the motor and vocal tics, the natural history of the syndrome, common comorbid conditions, coping strategies, prognosis, and treatment options comes next. Given the high rate of anxiety disorder and OCD in family members of children with GTS, this conversation can sometimes be difficult and require extra reassurance. Parents are often concerned about the psychosocial impact GTS can have on their child’s life. This has been explored in many studies, and there are

effective strategies for improving a child's quality of life. The most significant influential factor in a child's experience with GTS is whether or not they have a supportive teacher at the school. Barriers include the teacher not understanding the nature of tic disorder, the child being unaware of the tics, the tics being exacerbated when they are pointed out or when the child is anxious, and that disciplining a child for tics is not an effective strategy. Providing educational material for the parents, which can be shared with teachers, can be helpful. A study showed that a 5-minute generic presentation to students to educate them about tic disorder significantly reduced bullying and had a positive impact on the social experience of students with GTS. Ultimately, clear goals must be set for the treatment of children with Tourette syndrome. Expecting the child to sit quietly and still for the entire school day is probably not a realistic goal, so there needs to be a clear plan in place to accommodate the child's needs.[12][13][13]

The neurobehavioral comorbidities often are more disruptive for children with GTS than the tics themselves, particularly ADHD and OCD symptoms. Children should be thoroughly evaluated for symptoms of ADHD, OCD, depression, oppositional defiant disorder (ODD), autism spectrum disorder, and generalized anxiety disorder. These comorbidities need to be evaluated separately for severity and symptom-related disability. The treatment plan for tics should take into account whether or not the child has these other disorders, as there is evidence that children with different comorbid conditions respond differently to various therapy options or medication.

Behavior therapy and/or pharmacological treatment of Tourette syndrome may be indicated if the child is experiencing an impaired quality of life. This may be the result of pain from repetitive movements or whiplash tics. There may be difficulty with specific motor tasks secondary to disruptive tics, or difficulty transitioning to sleep at night because of frequent movement. The child may experience social isolation or distress and is at high risk for mood disturbance. Considering the potential side effects associated with the medication, behavior therapy should be the first-line treatment option. Habit-reversal training (HRT), usually as the main component of comprehensive behavioral intervention for tics (CBIT) is the recommended treatment of choice in the United States practice parameter. This involves multiple approaches to treatment, including training in recognizing and redirecting tics as well as intervention for influencing factors, such as relaxation techniques for anxiety. This therapy enhances the patient's recognition of the premonitory urge and offers a competing response or a motor movement that is not compatible with the tic. This therapy is very effective for the right patients. Studies have shown significant improvement in tic control compared to supportive psychotherapy, and over 80% of children maintained this control after 6 months of follow-up. Younger children are often less aware of the premonitory urges in tics, which may make it more difficult for them to fully understand the treatment. Other limitations include difficulty finding qualified therapists, cost, and lack of coverage by various insurance plans.

For more severe cases, alpha-2-adrenergic agonists and antipsychotics are first-line pharmacologic choices. Guanfacine and clonidine are recommended in the United States practice parameters. A couple of small trials showed equal efficacy between clonidine and risperidone and another between clonidine and haloperidol. However, there is some evidence that clonidine may only be effective in children with comorbid ADHD. Sedation is by far the most common reason children do not tolerate the alpha-2-adrenergic agonists. Other side effects include orthostatic hypotension, bradycardia, or irritability. They should be titrated off slowly to prevent rebound hypertension.

Antipsychotic medications have been the most extensively studied. Haloperidol and pimozide are the first-generation antipsychotics with the most data showing efficacy in reducing tic severity. However, their use is limited by potentially severe side effects such as sedation, acute dystonia, and other drug-induced movement disorders like weight gain, and prolonged QTc interval (pimozide). Newer atypical antipsychotic medications such as risperidone and aripiprazole both have data that demonstrate efficacy. There is a lower incidence of drug-induced movement disorders, but they are still high side effect medications. Risperidone can cause sedation, weight gain, and metabolic disturbances including hyperprolactinemia. Aripiprazole causes sedation, nausea, and weight gain. There is weak evidence showing that topiramate reduces tic burden, with a better side effect profile, so it may be a consideration if the child cannot tolerate the other two classes of medication.

For children refractory to CBIT and pharmacotherapy, there are a couple of other options. Botulinum toxin injections may be useful for motor tics that are particularly disabling or painful. These are logistically difficult and require an experienced specialist to administer. Deep brain stimulation has been used in patients with severe GTS refractory to all of the above treatment strategies. There is debate over the appropriate age to implant a device, as some patients show improvement by adulthood. However, adolescence is a formative developmental time in a child's life, so an argument could be made that waiting until adulthood would be missing the most significant treatment window.

Differential Diagnosis

Absence Seizures with Eyelid Myoclonus

Childhood absence epilepsy has an onset in the same age range as motor tics. Considering that the first motor tic typically involves eye blinking, it could be difficult to differentiate between these two. Motor tics are not associated with altered consciousness or staring and are usually much shorter in duration than absence seizures. Anxiety does not typically exacerbate seizures. Routine EEG is not usually indicated for children with motor tics, but if there is a concern for altered consciousness, this study should be able to differentiate between the 2 diagnoses.

Stereotypies

Rare, rhythmic, repetitive, fixed, predictable, purposeless movements that occur in children who are otherwise developing normally. Examples of primary motor stereotypies are flapping and waving of the arms, hand flapping, head nodding, and rocking back and forth. The difference is that these typically start before 3 years of age, often in infancy. Children tend to have a single repetitive movement, as opposed to multiple motor tics. These are easier to voluntarily control and tend to occur primarily when the child is excited. They significantly decrease in frequency as the child reaches elementary school age.

Chorea

Characterized by jerky, involuntary movements affecting especially the shoulders, hips, and face. The movements can be erratic but are more constant than motor tics. This occurs much less in childhood than do motor tics.

Paroxysmal Dyskinesias (PDK)

Abnormal movements often precipitated by a startle or a sudden movement. The attacks are usually short, only lasting seconds or minutes. An unusual sensation in the limbs may precede the symptoms. Most people with PKD have dystonia, and some have a combination of chorea and dystonia or ballism. This is usually autosomal dominant but can be sporadic.

Akathisia

Movement disorder characterized by a feeling of inner restlessness and inability to stay still. This is commonly a side effect of antipsychotic, antidepressant, or antiemetic medications. Typically, akathisia occurs in adults but is being seen more frequently in children as these medications are being regularly prescribed.

Obsessive-Compulsive Disorder

Distinguishing between motor tics and repetitive movements of OCD can be very difficult, and there is often overlap between these two illnesses. OCD movements are performed to relieve significant anxiety and are often associated with compulsive thoughts (“If I don’t do this exactly right, something terrible will happen”). Motor tics are typically preceded by a vague urge that is not as clearly defined and are usually more subconscious. The treatment approach is similar, so distinguishing between each movement is not usually necessary.

Prognosis

A general approximation used to offer families and patients insight regarding the prognosis of GTS is the suggestion that one-third will resolve completely, one-third will improve, and another third will continue without any attenuation. [14] Furthermore, tics appear to progressively decline during adolescence, as one study demonstrated a yearly decline in this demographic. In regards to the 'third' that experiences persistence of GTS into adulthood, it has been postulated that coexistent psychopathologies are more likely present (e.g. ADH, OCD).[15]

Complications

Complications of Tourette syndrome include behavioral and psychosocial impairments. As mentioned previously, GTS can precipitate embarrassment and shame. Additionally, GTS has been associated with increased diatheses of anxiety disorders, disruptive behaviors, mood disorders, learning disabilities, and sleep disorders.[16] In the most extreme settings, GTS-induced mood perturbations have been postulated to be the impetus for suicide.[17]

Deterrence and Patient Education

Patient education cannot be emphasized enough in the setting of Tourette syndrome. Tics can be dentrimental to appropriate psychosocial development and thus parents and teachers must be cognizant and supportive. Current

guidelines recommend habit reversal training with CBIT before initiating pharmacotherapy. In the event that CBIT is inaccessible or ineffective, medications may be necessary; these include tetrabenazine, risperidone, fluphenazine, or clonidine. Addressing comorbid psychiatric disorders will also help to alleviate the overall morbidity.

Enhancing Healthcare Team Outcomes

Tourette syndrome is best managed by an interprofessional team that includes a psychiatry nurse, pediatrician, primary care provider, psychiatrist, school teacher, and a social worker. The diagnosis is clinical and children with Tourette syndrome do not routinely need brain MRI. Only consider getting this test if the child has an abnormal neurologic exam or there are red flags in the history that may point toward a different diagnosis.

The provider should take a holistic approach to the treatment of children with GTS, considering comorbid neurobehavioral conditions. Families should be provided with extensive education, reassurance, and support.

Studies show that the school teacher can have a significant impact on the psychosocial experience of a child with Tourette syndrome, positive or negative. Taking time to educate the teacher about the condition and encouraging the education of the students in the child's classroom can significantly change a child’s life.[16]

Review Questions

- Access free multiple choice questions on this topic.
- Comment on this article.

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Bookshelf ID: NBK499958 PMID: 29763133