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Basic medical science faculty

Tourette syndrome and How Do We control It

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Abstract:
Tourette syndrome (TS) is a childhood onset neurodevelopment condition characterized by multiple motor tics and at least one phonic tic that have persisted for more than one year since their onset .commonly associated with other co-morbid conditions – such as Obsessive-Compulsive Disorder (OCD), Attention-Deficit and Hyperactivity Disorder (ADHD). Its pathophysiology most likely involves basal ganglia and frontocortical circuits.
TS suppression is common in physicians' offices, and the best time to look for tics is when the patient is walking into or out of the examination room and theres a criteria for it that will be discussed later on for its diagnosis.
As for its treatment it includes pharmacological therapy, local intramuscular injection of botulinum toxin is one of the therapeutic options and surgical treatment with deep-brain stimulation has recently been used in patients with TS who have disabling tics that are refractory to medication.

Introduction

Tourette syndrome (TS) is a childhood onset neurodevelopment condition characterized by multiple motor tics and at least one phonic tic that have persisted for more than one year since their onset(1) . Tics are defined as paroxysmal , rapid, non-rhythmic movements motor tics or vocalisation phonic tics(2). Motor tics range from single, short, sudden movements, such as eye blinking or nose twitching to complex behavioral sequences such as squatting, jumping, or even obscene gestures (copropraxia) Vocal tics can range from inarticulate single vocalizations to echolalia , one repeats the words of other paralilia , one repeats his/her own words or even the controversial feature of coprolalia “the utterance of obscenities(3).

TS is commonly associated with other co-morbid conditions – such as Obsessive-Compulsive Disorder (OCD), Attention-Deficit and Hyperactivity Disorder (ADHD), anxiety and affective disorder – in approximately 90% of people with TS, also document that these co-occurring conditions might affect and diminish the quality of life of individuals with TS more than the actual tics(4). The cause of Tourette’s syndrome is unknown, but the pathophysiology most likely involves basal ganglia and Frontocortical circuits a useful scheme of basal ganglia dysfunction(5).

A mutation in histidine decarboxylase (Hdc), the key enzyme for the biosynthesis of histamine (HA), has been implicated as a rare genetic cause (6).

This report aims to to broaden your knowledge about what this syndrome is in a whole, how to diagnose it and how it is managed. Also how patients reacted to different medications during numerous trials and what type of treatment turned out to be the most therapeutic.

Materials and methods

A clinical trial was carried out over an eight week period that compared habit-reversal treatment with supportive therapy, the score was measured by Yale Global Tic Severity Scale (YGTSS) which was made by Yale Child Study Center. October 1992 its psychological measure designed to assess the severity and frequency of symptoms

of disorders such as tic disorder, Tourette syndrome, and obsessive-compulsive disorder, in children and adolescents between ages 6 to 17 The higher score indicates a greater disability.

Result was 7.6 vs 3.5 indicating supportive therapy was more beneficial.

Discussion

pathophysiology most likely involves basal ganglia and frontocortical circuits. A useful scheme of basal ganglia dysfunction should be able to account for the features that make Tourette's syndrome unique, in addition to the features that Tourette's syndrome shares with other disorders. Recent advances in knowledge of basal ganglia functional anatomy and physiology make it possible to hypothesize how specific neural mechanisms relate to specific clinical manifestations of Tourette's syndrome. A model of selection and suppression of competing behaviors by the basal ganglia is presented. The functional anatomy of basal ganglia circuits and new information on dopamine modulation of those circuits provide the basis for hypotheses of basal ganglia dysfunction in Tourette's syndrome .(5)

A mutation in histidine decarboxylase (Hdc), the key enzyme for the biosynthesis of histamine(HA), has been implicated as a rare genetic cause.(6) The diagnosis of Tourette's syndrome is made when motor and vocal tics have been present for at least 1 year.(3) The temporal criterion is used to distinguish the tics of Tourette's syndrome from identical-appearing tics that can occur transiently during normal development. (3)

Traditional diagnostic distinctions between Tourette's syndrome and conditions such as chronic motor tic disorder and chronic vocal tic disorder in which there are motor or vocal tics, but not both, are probably invalid from a neurobiological perspective, since chronic motor or vocal tic disorder and Tourette's syndrome are believed to result from similar mechanisms. (3)

Thus, most clinicians now consider patients who have chronic motor or vocal tics or both types of tics to have Tourette's syndrome, Tics should be distinguished from compulsions.(3)

Unlike tics, compulsions occur in response to an obsession (e.g: hand washing due to fear of contamination), according to rules (e.g: a certain number of times or in a certain order), or to ward off harm to self or others. However, tics and compulsions commonly coexist and have phenomenologies that are so similar that sometimes it is difficult to distinguish between them.(3)

Tics commonly accompany developmental disorders such as mental retardation, autism, and Asperger's syndrome, and many experts do not diagnose Tourette's syndrome when these other disorders are present. In these cases, the tics are considered to be secondary to the developmental disorder. Other neurologic disorders can also cause tics, but these disorders are rare.(4)

In most children, the diagnosis of Tourette's syndrome is made clinically;
neuroimaging or other laboratory testing is not necessary to establish the diagnosis.(3)

Tic suppression is common in physicians' offices, and the best time to look for tics is when the patient is walking into or out of the examination room.(3)

Table 1. Diagnostic Criteria for Tourette's Syndrome.*

Both multiple motor tics and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently.
The tics occur many times a day (usually in bouts) nearly every day or intermittently throughout a period of more than 3 consecutive months.
The onset is before 18 years of age.
The disturbance is not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington's disease or postviral encephalitis).

* Criteria are from the *Diagnostic and Statistical Manual of Psychiatry*, 4th edition.[3]

In many children with Tourette's syndrome, tics are mild and not disabling, and education about the condition with some supportive counseling is sufficient. A key focus is on maintaining and strengthening the child's self-confidence and self-esteem. Supportive counseling may be helpful for these purposes, although it has not been rigorously studied. Tics can be disabling by causing social embarrassment, isolation, and sometimes conflict (e.g., because of vocal insults). Some tics (e.g., neck jerking) are painful, and some (e.g., scratching and poking) can be self-injurious. When tics are disabling, tic-suppressing therapy is indicated. General approaches to treating Tourette's syndrome and its common coexisting conditions are summarized in Table2

Table 2. Treatment Options for the Tourette's Syndrome Triad.*

Tics	OCD	ADHD
Habit reversal	Cognitive behavioral therapy	Behavioral therapy
Alpha-agonists	Selective serotonin-reuptake inhibitors	Alpha-agonists
Tetrabenazine	Atypical antipsychotic agents	Atomoxetine
Atypical antipsychotic agents	Deep brain stimulation	Methylphenidate
Classic neuroleptic agents		
Botulinum toxin		
Deep-brain stimulation?		

* ADHD denotes attention deficit–hyperactivity disorder, and OCD obsessive–compulsive disorder.

Behavioral Therapy

Clinical trials have shown that a form of cognitive behavioral therapy termed habit-reversal treatment is efficacious in suppressing tics.

This form of therapy involves training patients to monitor their tics and premonitory sensations and to respond to them with a voluntary behavior that is physically incompatible with the tics.
In a clinical trial that compared habit-reversal treatment with supportive therapy and education, habit reversal carried out in eight weekly sessions resulted in modestly greater improvement as measured by the score on the Yale Global Tic Severity Scale at 10 weeks (mean score reduction, 7.6 vs. 3.5 points).
This two-part scale measures the severity of tics and overall impairment; scores for each part range from 0 to 50, with higher scores indicating greater disability, and a change in the total score of 2.5 to 5.0 points is considered a minimal clinically significant difference.
Potential shortcomings of habit-reversal therapy are that it is not widely available, it is time-consuming, and its long-term benefits have not been examined. Its clinical value remains controversial, but some experts advocate trying this therapy before initiating medication in cases that are not severe.(2)

Pharmacotherapy

Table 3. Tic-Suppressing Medications.*

Medication	Daily Dose	Common Side Effects	Comments
	mg		
Alpha-agonists			
Clonidine	0.05–0.5	Sedation, dizziness, headache, irritability	Oral and transdermal forms available
Guanfacine	0.5–4.0	Sedation, dizziness, headache, irritability	Less sedation than with clonidine, and fewer doses needed
Antipsychotic agents			
Haloperidol†	0.5–20.0	Sedation, depression, increased appetite, parkinsonism	
Fluphenazine	0.5–20.0	Sedation, depression, increased appetite, parkinsonism	
Pimozide†	0.5–10.0	Sedation, depression, increased appetite, parkinsonism	ECG monitoring of the QT interval needed
Risperidone	0.5–16.0	Sedation, weight gain, glucose intolerance, parkinsonism	
Dopamine depleter			
Tetrabenazine	12.5–100.0	Sedation, insomnia, depression, restlessness	Expensive, dose influenced by CYP2D6 genotype

* This list is not comprehensive. ECG denotes electrocardiographic.
† This drug is approved by the Food and Drug Administration for this indication.

Botulinum Toxin

Local intramuscular injection of botulinum toxin is a therapeutic option for some types of particularly bothersome tics, although data from controlled trials are lacking. Case series indicate that this treatment can reduce tics as well as associated premonitory sensations and pain.
Botulinum toxin is used most frequently for eye blinking and neck and shoulder tics. The benefits are temporary, lasting 3 to 6 months.(4)

Deep-Brain Stimulation

Surgical treatment with deep-brain stimulation has recently been used in patients with Tourette's syndrome who have disabling tics that are refractory to medication.

The results of double-blind trials of thalamic stimulation with the use of a crossover design (comparing stimulation on with stimulation off) indicate that some patients have a substantial benefit.

The largest published trial showed a mean reduction of 29% in the score on the Yale Global Tic Severity Scale during stimulation. However, the criteria for identifying patients with Tourette's syndrome who will have the greatest benefit from deep-brain stimulation have not been established, and the optimal location for the electrodes in such patients is not clear; the globus pallidus, putamen, subthalamic nucleus, and other areas have been used.

Deep-brain stimulation can be complicated by stroke, infection, and side effects during stimulation such as paresthesia, visual symptoms, and dysarthria.(4)

Conclusion:

Tourette syndrome is a model disorder of the interaction of developmental, genetic, neurobiologic, and behavioral influences. Behavioral approaches are always appropriate, but they are often inadequate or inaccessible. There is no entirely safe psychopharmacologic agent, but one must also consider the risks of not getting treatment. Children with severe Tourette syndrome have difficulty learning due to the adverse impact of constant tics and other coexisting conditions, and they are often socially ostracized. It is clear that tic reduction and efforts to address serious comorbid conditions can dramatically improve quality of life. Medication treatment should never be the first approach and should always be performed in conjunction with behavioral and other supportive services. Adverse drug reactions should not discourage the clinician from considering other medication trials.

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