

The Life Cycle of Tics

An informative discussion of Tourette syndrome, tics, urges, obsessions, compulsions, habits, and inattentiveness. What they are, why they happen and what they feel like.

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Objectives

- Understand what motor and phonic tics are
- Understand what sensory urges and premonitory sensations are and what they feel like
- How do we label tics?
- How do we make the diagnosis of Tourette syndrome?
- What are the associated behaviors that we see with Tourette syndrome?
- Why is it important to identify the associated behaviors in Tourette syndrome
- Attention deficit hyperactivity disorder and obsessive-compulsive behavior
- Managing TS, ADHD and OCD in the educational setting
- What is the common timeline we see in Tourette syndrome and the associated behaviors?
- Where is the problem in the brain related to Tourette syndrome
- Pharmacologic and non-pharmacologic therapies for Tourette syndrome and related disorders

Georges Gilles de la Tourette and TS



- Worked at the famous Salpêtrière Hospital in Paris under Charcot
- Published his famous article in Archives de Neurologie in 1885
- Referred to the condition as "maladie des tics"
- Initially treated with haloperidol (Haldol) in the 1960s by Arthur Shapiro and colleagues

So just what is a tic?

- ♦ Any sudden, involuntary or semivoluntary, non-rhythmic, repetitive movement, gesture or utterance that mimics some fragment of normal behavior
- ♦ A motor tic is just what it sounds like. They can be brief, jerking movements (clonic tics), slow movements of sustained posture (dystonic tics) or an isometric contraction (tonic tics)
- ♦ A phonic, or vocal, tic includes any sound produced or, more simply, anything that arises from the movement of air through the windpipe

Sensory urges and premonitory sensations

- ♦ A premonitory sensation or sensory urge is a focal perception or mental awareness that precedes a tic
- ♦ Patients may describe the urge as a specific sensation such as an “itch,” “a dry throat,” “a tickle in the throat,” or that their “clothes are uncomfortable”
- ♦ Or, they may describe a non-specific urge or feeling such as “anxiety,” “a spring tightening,” or “a rubber band winding up”
- ♦ The actual tic is often described as a release of this tension and that the tic actually relieves the uncomfortable sensation that we refer to as the premonitory sensation
- ♦ Approximately 80% of patients with Tourette syndrome describe having sensory urges or premonitory sensations that precede their motor and phonic tics

What are the different types of tics?

- ♦ **Motor tics**
 - Simple motor tics – fast, darting, meaningless. Examples include eye blinking, head jerking, grimacing, jaw snapping, shoulder shrugs, neck tightening
 - Complex motor tics – slower, cluster of simple movements or a coordinated sequence of movements that may seem to be purposeful. Examples include hopping, clapping, touching, tapping, poking, smelling, kissing, brushing the hair out of one’s eyes
- ♦ **Phonic tics**
 - Simple phonic tics – fast, darting, meaningless. Examples include throat clearing, coughing, grunting, yelping, humming, sniffing, sniffing, animal noises
 - Complex phonic tics – linguistically meaningful utterances. Examples include words, syllables, phrases, statements, mutterings, expressions
 - Specific phonic tics
 - Echolalia – repeating what someone else says
 - Pallilalia – repeating one’s own words or phrases
 - Coprolalia – utterance of obscene words or socially inappropriate

DSM-5 Tic Disorders

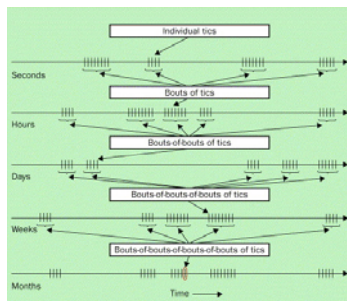
- ♦ Tourette syndrome
- ♦ Persistent (chronic) motor or vocal tic disorder
 - One or more motor tic or vocal tics, but *NOT* both
 - Tics that occur many times a day, nearly every day or on and off throughout the period of more than a year
 - Tics that start before 18 years of age
 - Symptoms are not due to medications or another medical condition
 - Not diagnosed with Tourette syndrome
- ♦ Provisional tic disorder
 - One or more motor tics or vocal tics, but *NOT* both
 - Present for no longer than 12 months in a row
 - Onset before the age of 18
 - Symptoms are not due to medications or another medical condition
 - Not diagnosed with Tourette syndrome or persistent motor or vocal tic disorder

DSM-5 Diagnostic Criteria for TS

- ♦ Have both multiple motor tics *AND* vocal tics, although they might not always happen at the same time
- ♦ Have had tics for at least one year. The tics can occur many times a day (usually in bouts) nearly every day, or on and off
- ♦ Have tics that begin prior to the age of 18 years
- ♦ Have symptoms that are not due to taking medicine or other drugs or due to having another medical condition



The elusive nature of tics...



- ♦ Tics come and go and are often worse without clear triggers
- ♦ Tics will commonly worsen with stress, anxiety or fatigue
- ♦ Tics have a waxing and waning course that is impossible to predict with any certainty
- ♦ This unpredictable nature of tics makes determination of treatment success very difficult to assess

Lechman, James F, Tourette's syndrome. Lancet 2002; 369: 1577-86

The disabling nature of tics...

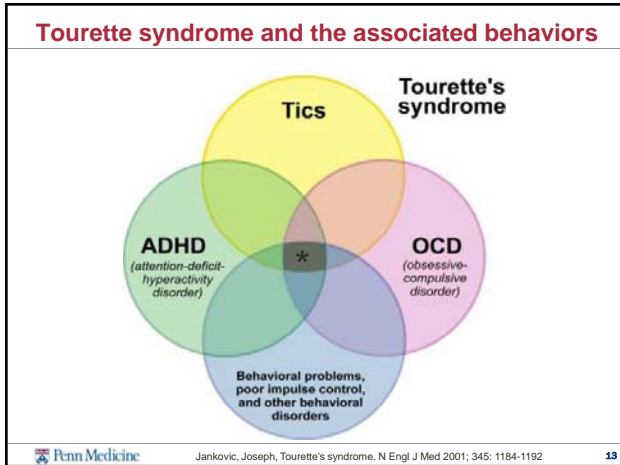
- ♦ The disabling nature of tics is not always as obvious as it seems
- ♦ Though tics can be disturbing to patients, it is often the mental energy expended to suppress tics and premonitory urges or the urges alone that are tremendously more disabling than the tics themselves
- ♦ School children will suppress their tics at school and then release them at home after getting off the school bus
- ♦ Premonitory urges and tics interfere with attention and focus which is only exacerbated by the effort to suppress the urges and tics
- ♦ Tics will often increase in frequency when they are brought to one's attention or are the focus of conversation

Many tics are physically uncomfortable...

- ♦ Severe neck pain from neck snapping or head tossing that can over time cause severe arthritis and even spinal cord injury
- ♦ Constant throat clearing or coughing causing pain and irritation
- ♦ Severe eye pain caused by constant squinting or blinking
- ♦ Tooth trauma from constant snapping of the jaw
- ♦ Punching or slapping causing injuries

The associated behaviors of TS...

- ♦ Inattention, disorganization, hyperactivity, impulsivity (ADHD)
- ♦ "The Sticky Brain" – hyperfocusing, "trouble letting go"
- ♦ Obsessions and compulsions (OCB and OCD)
- ♦ Chronic anxiety – social, generalized, specific phobias
- ♦ Emotional instability/lability – dysregulation of affect
- ♦ Mood disorders – depression, dysthymia, bipolar disorder
- ♦ Mirror phenomenon – echolalia, pallilalia, echopraxia
- ♦ Learning differences / disabilities
- ♦ Executive dysfunction
- ♦ Sensory processing issues
- ♦ Poor impulse control



Why is it important to identify these behaviors?

- ♦ The associated behaviors are often more disabling than the tics, especially in children and adolescents
- ♦ ADHD accounts for almost all of the aggression and delinquency seen in the TS population (i.e. TS-only patients show same rates of these problems as the control population)
- ♦ Academic, social and occupational difficulties seen in ADHD+TS is virtually identical to those seen in ADHD-only patients
- ♦ While ADHD is highly co-morbid with learning disabilities, TS-only patients do not have higher rates of LD than control population
- ♦ ADHD carries significant risk for lifelong psychiatric morbidity in TS
- ♦ 50-90% of patients with TS who present for treatment exhibit signs of ADHD and accompanying ODD or CD
- ♦ The community prevalence of ADHD is 5%; the rate among children with tics is roughly 10 fold that of the general population

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Tourette syndrome and OCD

- ♦ Approximately 50% of TS patients will exhibit symptoms of obsessive-compulsive behavior
- ♦ Obsessive-compulsive behavior and OCD are an anxiety disorder and are characterized by:
 - Recurrent and unwanted obtrusive thoughts and images (obsessions), and/or
 - Repetitive behaviors (compulsions) that are done with the hope of preventing or reducing the obsessions
 - Performing the "rituals" provides only temporary relief
 - Not performing them significantly increases anxiety
- ♦ The rituals interfere with daily life and can be isolating and embarrassing
- ♦ Common problems seen in school as a result of OCD include:
 - Difficulty transitioning from one activity to the next
 - Not completing work or drifting to another activity
 - Constant erasing
 - Inability to tolerate mistakes

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Managing TS, OCD and ADHD in school

♦ **Organizational skills and executive function are heavily impacted by both ADHD and OCD in children and adolescents, often greatly impacting their educational performance**

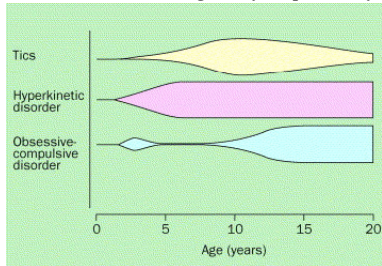
- Impaired attention and focus and easy distractibility
- Difficulty with short-term memory
- Difficulty starting and completing projects or staying on task

♦ **Some common strategies to help with ADHD and OCD in school**

- Extended time for testing or testing in an alternative environment
- Extra set of books for home
- Reducing amount of homework (odd or even answers) or projects
- Emailing assignments home each day
- Allowing children to step out of class when necessary
- Having a safe place for children to go when necessary
- Reassuring a child is staying on task
- Audio books for children with difficulty focusing when reading

What is the common timeline we see in TS?

- ♦ **ADHD presents earlier than tics (4-6 years of age)**
- ♦ **Simple motor tics begin at 6-8 years of age with vocal tics later**
- ♦ **OCB or OCD typically presents shortly after tics (7-9 years of age)**
- ♦ **ADHD and OCD remain life-long as they are personality traits**



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Where is the problem in the brain related to TS?

♦ **Neurotransmitter hypotheses in TS**

- Dopamine – response to neuroleptics and exacerbation by stimulants
- Serotonin – response to SSRI antidepressants
- Norepinephrine – exacerbation by stress/anxiety and positive response to medications such as clonidine and guanfacine

♦ **The basal ganglia and TS**


- The basal ganglia are hypothesized to participate in the selection, activation and termination of innate and learned motor programs (i.e. habits)
- In addition, the basal ganglia facilitate the learning of appropriate context-dependent motor behavior, e.g. obtaining a reward or avoiding danger in a familiar situation. Once learned, such actions may become subconscious, i.e. habitual or automatic

♦ **The cortex and TS**

- The prefrontal cortex has been implicated in TS, OCD and ADHD


Principles of Intervention

- ♦ Clarify the clinical syndrome (course, prognosis)
- ♦ Target specific symptoms
- ♦ Use medications only for psychosocial or physical disability stemming from symptoms
- ♦ Consider co-morbid conditions in choice and priority of any interventions
- ♦ Always employ a multi-modal treatment approach
 - Education of child, family, school, community
 - Parent and teacher training – behavior management
 - School interventions
 - Cognitive behavioral interventions
 - Medications (for target symptoms)
 - Social skills groups
 - Support groups
 - Complementary treatments
 - Results of the Pediatric OCD Treatment Study (POTS) Randomized Controlled Trial

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Pharmacologic therapies for TS+

- ♦ All medications for TS and related behaviors are symptomatic; i.e. they are used only to mitigate the symptoms and are not curative
- ♦ Medications are chosen to treat the specific symptom that is disabling
- ♦ Some medications are effective in treating multiple symptoms associated with Tourette syndrome
- ♦ Common medications for tics include:
 - Clonidine and guanfacine
 - Topiramate
 - Risperidone, Aripiperzole
 - Haloperidol, pimozide, fluphenazine
- ♦ Common medications for ADHD include:
 - Clonidine and guanfacine
 - Atomoxetine
 - Methylphenidate and amphetamine compounds
- ♦ Common medications for obsessive-compulsive behavior include:
 - SSRI and SNRI antidepressants
 - Clomiprimine

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Non-pharmacologic therapies for TS+

- ♦ Habit Reversal Training (HRT)
 - Awareness training, self-monitoring, competing response training (where a movement is performed that is opposite to a particular tic)
- ♦ Comprehensive Behavioral Intervention for Tics (CBI-T)
 - Therapy increasing awareness of tics and the urge to tic, training to do competing behaviors when the urge to tic is felt and changing day to day activities that can be helpful in reducing tics
- ♦ Comprehensive Behavioral Therapy
 - The mainstay for non-pharmacologic treatment of obsessive-compulsive behavior and anxiety, but also valuable for ADHD
- ♦ Neurofeedback for ADHD
 - Typically involves computer-based exercises providing feedback regarding attention levels to promote self-regulation and enable behavioral training

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