

Tics and Tourette Syndrome in Children and Adolescents: Information for Primary Care and Psychiatrists



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Sommaire : Children/youth may present with brief motor episodes, but not all are tics. Tics are neurologic in basis, and have specific features (e.g. suggestible, suppressible, worsen with stress and have an associated premonitory urge). Most movement difficulties and tic disorders improve with time. Management includes starting with education of the family about the nature of tics and informing the school. Further steps might include (referral to) habit reversal therapy and other psychological interventions. Should tics persist and cause impairment, medication treatments include clonidine and guanfacine. Otherwise, antipsychotics such as Risperidone or Aripiprazole are often used. Look for and manage any comorbid conditions such as attention deficit hyperactivity disorder (ADHD) and obsessive compulsive disorder (OCD) as they often cause more significant issues than the tics themselves. Should tics persist, or should it be a complicated presentation, refer to specialty care.

Case, Part 1

You are seeing a 15-yo teenager whose mother has brought him to see you due to frequent blinking and grunting. He tends to do this more when he is watching TV, and less when he is focused on a particular activity such as playing guitar. Unfortunately, his siblings do not understand and are getting annoyed at him for grunting and making noises.

When you ask him about his main concerns, he states he is more concerned about his siblings teasing him for his grunting, as well as troubles paying attention in class.

What are Tics?

Tics are sudden, repetitive stereotyped movements or noises.

Features include:

- Involuntary, in that the patient may have no control over them
- Semi-voluntary, in that the patient be able to suppress them for short periods. The fact that tics are 'semi-voluntary' and appear to be under the control of the person can sometimes lead to parents and family members putting pressure on the patient to 'control' their tics.

Older children and adults may report a premonitory sensation such as paresthesia prior to the tic.

After suppressing tics all day, a child may experience temporary "release" of tics upon returning home.

Triggers include:

- Stress
- Fatigue
- Excitement
- Talking about tics

Relieving factors

- May improve when the patient is concentrating on something or distracted, e.g. watching TV, playing video games, performing neurosurgery (in a famous case of a neurosurgeon with Tourette's)

Classification of Tics

	Simple	Complex
Motor	<ul style="list-style-type: none"> Eye blinking Head twitching Head thrusting Shoulder shrugging Mouth opening 	<ul style="list-style-type: none"> Facial grimacing Touching Smelling Jumping Coprophaxia (obscene gestures) Echopraxia aka. Echokinesis (imitating another person's gestures)
Vocal	<ul style="list-style-type: none"> Sniffing Snorting Coughing Throat clearing Grunting Barking 	<ul style="list-style-type: none"> Echolalia (echoing what other's say) Palilalia (repeating sounds, words, numbers spoken by oneself) Coprolalia (swearing, obscenities)

Source: Koch, 2016

Epidemiology

Tics are common - 20% of children show tics at some point in their life

Age of onset: Usually tics start around age 5-6 years, but patients are often not brought to see a physician until later around 9-10 years (Mills, 2014).

Course and Prognosis

Tics tend to come and go and change over time. Even when they are particularly bad, they often improve again after several weeks.

Tics may worsen and are at the worst from age 8-12 years (Verdellen, 2011).

As the youth ages:

- Most cases (85-90%) show improvement in tics by late teens and early adulthood (NINDS, 2012).
- A minority (10-15%) may have a worsening case of tics/Tourette that lasts into their adult years (NINDS, 2012).

Pathophysiology

Tics are felt to be due to basal ganglia dysfunction and immaturity. As a result, tics generally improve as the basal ganglia and other relevant systems develop and mature.

Presentation

In most cases, the tics themselves are not the main cause of impairment, but rather the comorbid conditions such as attention deficit hyperactivity disorder (ADHD), obsessive compulsive disorder (OCD) and executive skills dysfunction that can occur as well.

Parents may bring the child to be seen for symptoms of impulsivity, behavioural issues or obsessive compulsive symptoms.

History

Take a movement history:

Age of onset	At what age did the unusual movements start?
Location	Where in the body were these movements?
Course	How did the movements progress over time?
Worst severity	At what age were they the worst?
Impairment or distress	Any distress, problems or impairment at home, school or elsewhere because of these movements? E.g. Is it distressing to the person? Or mainly to other people? E.g. Is it causing social problems, such as at school?
Severity of impairment or distress	Between 0 and 10, if 0 is no distress at 10, and 10 is the most distressing, how distressing are the tics / movements?
Premonitory urges	Any feelings or urges before a movement?

DSM-5 Tic Disorders

Provisional Tic Disorder (formerly known as Transient Tic Disorder in DSM-IV)	<p>Features:</p> <ul style="list-style-type: none"> <input type="checkbox"/> Ongoing fluctuating motor and/or vocal tics <input type="checkbox"/> Onset before age 18 years <input type="checkbox"/> Duration for less than one year.
Chronic Motor or Vocal Tic Disorder (CMVTD)	<p>Features</p> <ul style="list-style-type: none"> <input type="checkbox"/> Fluctuating tics that are either entirely motor or, less commonly, solely vocal tics. <input type="checkbox"/> Onset start before age 18 <input type="checkbox"/> Duration for more than 12-months <input type="checkbox"/> CMVTD is similar to TS, except CMVTD lacks vocal tics

Tourette's Disorder (aka Tourette syndrome)	<p>Features</p> <ul style="list-style-type: none"> <input type="checkbox"/> Multiple motor and at least one vocal tic, although not necessarily concurrently. <input type="radio"/> Coprolalia (10-19% of patients), the use of obscene words or socially unacceptable language, is one of the most socially distressing symptoms, but is not a diagnostic criterion. <input type="checkbox"/> Onset before age 18 years <input type="checkbox"/> Duration for over a year <p>Additional features</p> <ul style="list-style-type: none"> ● Tics occur many times a day, nearly every day or intermittently throughout a period of one year. ● Not due to use of a substance or general medical condition. <p>Course</p> <ul style="list-style-type: none"> ● Symptoms wax and wane over days, weeks or months <p>Comorbidity</p> <ul style="list-style-type: none"> ● ADHD by age 4 <input type="radio"/> Executive dysfunction causes problems with school and social function ● OCD by age 7 ● Depression, anxiety, behaviour problems <p>Prognosis</p> <ul style="list-style-type: none"> ● Tics improve by age such that by age 18, most have improvement in tics, and 50% have complete resolution of symptoms, as the nigrostriatal system matures
Substance-Induced Tic Disorder	<p>Features</p> <ul style="list-style-type: none"> <input type="checkbox"/> Onset during or within one month of substance intoxication or withdrawal which suggests a causative role of the substance <p>Stimulants are not considered a good example since there is existing evidence that they are no more associated with tics as an adverse event than placebo or other medications.</p>
Tic Disorder Due to a General Medical Condition	<p>Features</p> <ul style="list-style-type: none"> <input type="checkbox"/> General medical condition (e.g. infection, toxins, stroke, head trauma, surgery) that causes the tic <input type="checkbox"/> Variety of sporadic, genetic, and neurodegenerative disorders, such as neuroacanthocytosis, Huntington's disease, and Creutzfeldt-Jakob.
Tic Disorder, Not Otherwise Specified	<p>Features</p> <ul style="list-style-type: none"> <input type="checkbox"/> Movements/vocalizations do not meet criteria for a specific tic disorder because they are <input type="checkbox"/> Atypical clinical presentation. <input type="checkbox"/> Atypical in age of onset (i.e., adult onset) or <p>Symptoms starting in adulthood tend to:</p> <ul style="list-style-type: none"> ● Have environmental triggers ● Have more severe social impairment ● Respond more poorly to medications

Differential Diagnosis

Doing the comprehensive differential diagnosis is most likely beyond the scope of a typical primary care practice. The differential diagnosis for unusual movements is extensive, and includes the following:

<p>Are there movements such as:</p> <ul style="list-style-type: none"> • hand flapping or twisting, • body rocking, • head banging, • face or mouth stretching such as a marked grimace. 	==>	<p>Consider motor stereotypy, a repetitive, non-functional motor disorder which interferes with normal activities or results in injury. Commonly seen in ASD.</p>
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<p>Are there movements such as:</p> <ul style="list-style-type: none"> • pacing up and down, • rubbing the legs, face or scalp with the hands. 	<p>==></p>	<p>Consider akathisia, a feeling of discomfort, and as a result, individual feels a need to walk or move to ease the discomfort</p> <p>Akathisia can occur as a result of:</p> <ul style="list-style-type: none"> • Iron deficiency • Thyroid disorders • Side effect of drugs (e.g. neuroleptic medications such as Haloperidol or Pimozide).
<p>Is there</p> <p>1) An abrupt, dramatic onset of obsessive-compulsive disorder or severely restricted food intake;</p> <p>2) Concurrent presence of at least two additional neuropsychiatric symptoms, with similarly severe and acute onset such as:</p> <ul style="list-style-type: none"> • Anxiety, emotional lability and/or depression; • Irritability, aggression and/or severely oppositional behaviors; behavioral (developmental) regression; • Deterioration in school performance; sensory or motor abnormalities; • Somatic signs and symptoms, including sleep disturbances (night terrors, difficulty falling or staying asleep or waking too early), or urinary frequency. 	<p>==></p>	<p>Consider pediatric acute-onset neuropsychiatric syndrome (PANS), a dramatic (often overnight) onset of neuropsychiatric symptoms classically with OCD symptoms or food restriction, but can also involve tics.</p> <ul style="list-style-type: none"> • ● Speak with neurology or psychiatrist about where to refer.
<p>Is there the following constellation including:</p> <ul style="list-style-type: none"> • General: Fatigue that stops a person from engaging with previously enjoyed activities; impairs school function. • Neuropsychiatric: New onset of mood or anxiety problems; mood swings; poor concentration; insomnia; headaches; vocal and/or motor tics, sound/light sensitivity • GI: Gastrointestinal problems, • MSK: joint and muscle pain. 		<p>Consider Lyme disease.</p> <ul style="list-style-type: none"> • Children with Lyme disease may present with neuropsychiatric symptoms such as tics, anxiety and ADHD (Riedel, 1998) and thus easily be misdiagnosed. • Majority of patients with Lyme disease complain of unrelenting fatigue, difficulties engaging in previously enjoyed activities and decline in school performance.
<p>When triggered by unexpected noise, movement or touch, does the person have:</p> <ul style="list-style-type: none"> • Excessive startle response such as eye blinking or body spasms? • Extreme muscle tension (stiffness or hypertonia) that prevent voluntary movement and can cause the affected person to fall stiffly, like a log, without loss of consciousness. • Exaggeration of reflexes (hyperreflexia) • Unstable way of walking (gait) may also occur. 		<p>Consider excessive startle (hyperekplexia)</p> <ul style="list-style-type: none"> • A rare, hereditary neurologic disorder. • Potentially dangerous, e.g. individual may choke, fall or drown.

For more details, please see the decision tree from the ESSTS (Cath, 2011).

Comorbid Conditions

Comorbid conditions are present in the majority (80%) of those with tic disorders and Tourette Syndrome. Screen for comorbid conditions, which often cause a greater impact than the tics themselves:

<p>ADHD (in up to 70%) (Freeman, 2007)</p>	<p>Any problems with inattention? Any problems with hyperactivity? Any problems with impulsivity?</p>
<p>Obsessive compulsive disorder (OCD) (in over 80%) (Robertson, 2000)</p>	<p>Any repetitive movements? (i.e. compulsions)? Any troubling thoughts that you can't get off your mind? (i.e. obsessions)</p>

Anxiety (in about 30%) (Stefl, 1984)	Any problems with worries? Are these worries excessive?
Anger, rage and behavioural problems	Any problems with low frustration tolerance? Anger? When frustrated, any problems with: <ul style="list-style-type: none"> ● Verbal aggression? E.g. screaming ● Physical aggression? E.g. damaging property
Learning disability	Any learning problems?

Physical Exam

A full physical including neurologic exam is important to rule out progressive neurological disorders (Mill, 2014).

Head/Neck	Any involuntary motor movements, or verbalization? E.g. sniffing, blinking, grunting, etc. Any Kayser-Fleisher rings that would suggest Wilson's disease?
MSK	Any involuntary movements in arms / legs / trunk?
Skin	Any skin issues that would suggest neurocutaneous syndromes such as tuberous sclerosis and neurofibromatosis?
Neurologic	Other than the tics, children with tics will have an otherwise normal neurologic exam. Abnormal gait would suggest other conditions.

It is also possible that the child's tics may be suppressed during the office visit. If this is the case, consider asking parents to videotape the tics.

Investigations

Are there classic tic or Tourette features? → Further investigations are not required.

Are there atypical features? (E.g. adult onset, uncharacteristic deterioration, progressive worsening of symptoms)

- If YES, then further investigations should include
 - EEG
 - Neuro-imaging

Management of Tics

Management of tics starts with non-medication interventions such as:

- Education about tics for the family, teachers and classmates to help others to understand the behaviours, so that the patient can be more accepted
- Otherwise, often patients may be teased, ridiculed, punished over their tics
- Key points
 - Reassure parents that tics are common, occur in 20% of children
 - Don't call attention to tics or pressure the child to stop their tics, which may increase stress and thus worsen tics
 - Patients can be taught behavioural strategies to help with their tics
 - In more severe situations, medications may be helpful
 - Nonetheless, in most cases tics do not need to be treated, and they usually resolve over time
- Provide patient / family education materials and advocacy / support groups such as
 - Tourette Canada (www.tourette.ca)

Psychological interventions including the following:

- Habit reversal therapy (Deckersbach, 2006; Woods, 1995) is a non-medication approach to helping people with tics.
- Relaxation training

School Liaison

Consider writing a letter to the school that documents the diagnosis of tics or Tourette Disorder. This helps educators understand the nature of the child with tics and Tourette, in order to help create a more understanding environment. When teachers (or peers) do not understand tics, they may inadvertently tease, ridicule or discipline a child for the tics, in particular vocal tics such as grunting or swearing.

There are various resources for teachers on how to support a student with tics/Tourette in the classroom https://tourette.ca/wp-content/home/ementalhealth/ementalhealth.ca/frontend/uploads/2016/09/10_Helpful_Tips_for_Educators-1.pdf

Medications for Tics

Do the tics meet any of the following indications for medications? (Roessner, 2011)

- Tics causing pain or discomfort, such as intense or frequent tics that cause pain, or even injury.
- Tics causing social problems, such as coprolalia or complex motor tics that lead to teasing, bullying or social isolation.
- Tics causing psychological problems such as depression or anxiety
- Tics causing impairment, such as concentration and fatigue from the effort of having to suppress suppress his/her tics

Medications for Treatment of Tics in Children/Youth

Medication	Dosing / Comments
Alpha2-adrenergic agonist	First-line for tics, given less likelihood of side effects compared to other medications
● Clonidine (Catapres)	Start at 0.05 mg bedtime Therapeutic target of 0.1 mg three times a day Maximum 0.2 mg three times a day
● Guanfacine (Intuniv XR; Tenex)	Start at 0.5 mg bedtime Therapeutic target of 1 mg twice daily Maximum 1 mg three times a day
Dopamine receptor blocking drugs	Consider for multiple, or complex tics, however not first-line due to side effects
● Aripiprazole (Abilify)	6-18 years (<50 kg) <ul style="list-style-type: none"> • Start 2 mg daily • Target dose of 5 mg daily after 2 days • Maximum 10 mg daily, increasing in weekly intervals 6-18 years (≥50 kg) <ul style="list-style-type: none"> • Start at 2 mg daily x 2-days • Target dosage of 5-10 mg daily after 1-2 weeks • Maximum 20 mg daily; increase gradually by 5 mg at weekly intervals
● Pimozide (Orap)	Start at 0.5 mg bedtime (or 0.5 mg/kg/day) Therapeutic target of 1 mg twice daily Maximum 3 mg twice daily

● Risperidone (Risperdal)	Start at 0.25 mg bedtime Therapeutic target of 1 mg twice daily Maximum 2 mg twice daily
● Olanzapine (Zyprexa)	Start at 1.25 mg bedtime Therapeutic target of 2.5 mg twice daily Maximum 5 mg twice daily

Reference: Kenney, 2008; Roessner, 2011

Management Based on Target Symptoms

Issue	Possible Recommendations
Tics	Atypical antipsychotic (e.g. risperidone (Risperdal), aripiprazole (Abilify))
Tics and ADHD	Clonidine
ADHD symptoms	Consider alpha2-adrenergic agonists ADHD stimulants Atypical neuroleptics
OCD symptoms	1st line: Fluoxetine and other SSRIs, otherwise 2nd line: Clomipramine Consider referring to psychologist, general paediatrician or child psychiatrist
Learning difficulties suggestive of learning disability	Refer for psychoed testing such as <ul style="list-style-type: none"> ● Privately; ● Refer to psychoeducational testing through school board by writing a letter recommending psychoeducational testing
Depression	SSRI (e.g. Fluoxetine)

When to Refer

Are there any of the following?

- Unusual physical features
- Learning difficulties
- Autism spectrum disorder

If so, then consider referral to

- Paediatrician
- Neurologist
- Clinical geneticist
- Specialized motor disorder / tic / Tourette clinic if available

When to Refer to Neurology

If tics are causing distress at home and school, consider referring to:

- Neurology,
- Psychiatrist comfortable in managing tics, or
- Specialty Tourette / Tic Clinic if available.

Case, Part 2

You are seeing a 15-yo teenager whose parents have brought him in due to frequent blinking. Your assessment shows that aside from the blinking and grunting, he does not have any other unusual movements, and that his movements appear to be typical of tics. You give him a diagnosis of provisional tic disorder. You provide some information to the family, which will hopefully help the family (i.e. siblings) be more accepting.

Given his complaints of attention however, you ask them to come back in a few weeks time to explore further.

Clinical Practice Guidelines

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* Indicates references of particular interest to primary care..

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About this Document

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