Childhood tic disorders: diagnosis and management

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Abstract

Tics are considered to be benign but often cause great distress to the child and family, especially when there is a lack of understanding about their nature and the related conditions. This article is a clinical guide to assessment, diagnosis and management; with focus on practical aspects, caveats and catches and most importantly recognising and managing developmental and psychiatric co-morbidities in Tourette Syndrome and other Tic conditions.

Keywords Hyperkinetic movements; movement disorders; neuropsychiatric disorder; paroxysmal movements; stereotypies; tics; tourette syndrome

Introduction: what are tics and tic disorders?

Tics are common in childhood and are defined as sudden, rapid, repetitive, non-rhythmic, inapposite, irresistible, muscle movements (motor tics) or vocalizations (vocal tics), which can be classified as simple or complex tics (see Table 1). The term *Tourette Syndrome* (named after French neurologist Gilles de la Tourette) is classified under the motor disorders section of neurodevelopmental disorders (DSM-5, Diagnostic and statistical manual – 5th edition. American Psychiatric Association 2013). It is defined by the presence of multiple (2 or more) motor tics and one or more vocal tic; both present at the same time (but not necessarily simultaneously) for at least 1 year; a waxing and waning course and onset before 18 years are part of the diagnostic criteria. For this diagnosis the tics should not be attributable to substance use or medical conditions. Tics are devided in to motor and vocal tics (see Table 1).

Tics are often benign and no research (to date) indicates that they cause harm to or a delay in brain development. It is however important not to confuse this fact to mean that their presence can be ignored and dismissed. Tic presence can be considered as a 'surface marker' for wider neuro-developmental, neuropsychological and psychiatric difficulties. It is with this in mind that this article has been written aimed at the general paediatrician meeting children with 'Tourette Spectrum Difficulties', a term we like to use in our clinical service.

In children with Tics there is a general presence of impulse control difficulties, subtle differences in neuropsychological and motor functioning, as well as a high rate of psychiatric/developmental co-morbidities such as ADHD (30–60%), compulsions (or obsessive-compulsive behaviours 30–40%), anxiety (25%), disruptive behaviour (10–30%), depression/mood difficulties (10%), Obsessive-Compulsive Disorder (5–8 %), Autism Spectrum Disorder (5%) and motor co-ordination difficulties. Some children also have episodic rage. Quality of life is usually affected by these co-morbidities rather than the tics themselves.

Practice Point: when you see a child with tics screen for the common comorbidities, as they may need more active management than the tics.

Classification of tic disorders (DSM-5)

1. **Provisional (transient) Tic disorder:** Motor or vocal tics or both which have been present for less than a year.
2. **Chronic Tic disorder:** Either single/multiple motor tics or vocal tics present for over a year.
3. **Tourette Syndrome:** Multiple motor tics along with vocal tics lasting for over a year, they do not have to be concurrently present; they follow a waxing waning pattern.

A diagnosis of Tourette syndrome indicates neither the severity nor the complexity of the tic condition. A young person can have severe and multiple motor tics without any vocal tics thus not meeting criteria for a diagnosis of Tourette's, whilst another young person may have two very mild motor tics and a vocal tic and have Tourette syndrome. Similarly the complexity of a tic conditions is most often secondary to the types and number of co-morbidities rather than from Tourette vs Tic disorder diagnosis. The term Tourette syndrome is an constructed label that has found its way into the media and wider public as denoting repetitive swearing (coprolalila). This is a misconception as in reality only 5–8 % of people with Tourette's will have coprolalia. Nevertheless the term Tourette syndrome continues to be used by most clinicians to honour the nineteenth century French Neurologist Georges Gilles de la Tourette. We use this historical information to help to allay the natural anxiety that can come with the use of the label.

Practice Point: it can help to discuss 'Monsieur Tourette' to allay the stigma of the diagnostic labels.

Characteristics of tics

Children with tics often describe them as somewhat involuntary but voluntary participation is often required in response to an urge to tic. This premonitory urge preceding a tic is not very clear to younger children but in our experience usually emerges around aged 8 years (the ability to perceive the urge and the

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strength of the urge can vary between individuals). The premonitory urge appears to be the involuntary part of a tic and often the movement is done to alleviate the urge but many younger children with rapid tics will describe them as happening very suddenly without much warning or much voluntary participation. Following the tic movements, the urge reduces but may quickly increase again.

Tics are usually suppressible; to some extent the ability to supress, and the duration of suppression can vary between individuals. Suppressibility also depends on the amount of practice one has done to hold/supress tics. Interestingly not all suppression is effortful and some children report that "they just stop" in some situations especially on walking into the clinic room for example. Tics are very suggestible hence talking about these or drawing attention to these, for instance by mimicking the tic-action in front of someone with tics will worsen them. Similarly, stimulation, massage, warm compresses or irritation of parts of the body where the tics are occurring will usually exacerbate tics.

In addition to the co-existing developmental and psychiatric disorders there are also some interesting neurological phenomenon known to occur in some but not all individuals with tics. These include:

- Echolalia - repeating someone else's speech after hearing it due to a strong/irresistible urge to do so (often done under the breath)
- Echopraxia — copying an observed physical action (of another person), an irresistible urge is triggered by observing such an action which only is relinquished by reproducing that action in some way.
- Coprolalia — irrepressible urge to swear resulting in actual repetitive and often out of context swearing or use of other inappropriate words/sentences. They will often modify suppress this and look a bit annoyed or embarrassed by it often but not always explaining/apologising to a new observer.
- Copropraxia is the irrepressible use of inappropriate and rude gestures such as repetitive display of the middle finger in a social context where it draws attention and/or potentially embarrasses the patient. Most (but not all) will try to suppress, modify or conceal it and try to explain or apologise for using these gestures.
- Misophonia — refers to negative emotional arousal in response to certain sounds. This has recently been recognised in young people with tics, often triggering anger and an exacerbation in tics.

Practice Point: Ask about whether certain movements, sounds and situations trigger or exacerbate the tics.

Epidemiology and course of tics

Tics usually start between the ages of 4—7 years. For most children the first tics are in the form of repetitive blinking, sniffing, throat clearing or coughing. Tics fluctuate considerably in severity and frequency. Many children with minor and transient tics between ages of 4—6 will not come to medical attention. Transient tics are very common. Chronic Persistent Tics and Tourette Syndrome affect 1—2 % children.

Tics will typically fluctuate, waxing and waning over time, and moving around the body. New tics emerge and earlier ones disappear and this is part of the expected course. Tics get worse with anxiety, tiredness, illness, excitement and anecdotally probably with excessive screen time. Tics tend to reduce when a child is engaged in a cognitively demanding and interesting task. Exercise will typically reduce tics especially whilst engaged in the physical activity. Tics should not stop voluntary action and they will not interfere with important actions or result in falls or injuries and any presentation of such tics (also called blocking tics) should alert a clinician to the possibility of a functional component/tic-like attacks.

Tics peak in severity between ages 8 and 12 years. After 13 they tend to reduce in a fluctuating manner. In about 55—60% of young people tics will be minimal by late teens - early adulthood. In another 20—25% tics become infrequent but occasional. In around 20% the tics continue into adulthood (some of whom will report a worsening of tics).

Aetiology

In the past tics were considered behavioural or stress-related and often referred to as 'nervous habits' or 'twitches', it is now known that tics are neurological movements that may be worsened by anxiety but this is not causal. Tics have a complex multigenetic aetiology and they are highly heritable — concordance between monozygotic twins is 87% (and *de novo* presentations are rare) interested readers are advised to review our reading list recommendations.

Underlying mechanisms involve several neural networks in the brain, primarily between the cortex and basal-ganglia (fronto-striatal-thalamic circuits) but also involving other brain areas such as the limbic system, mid brain and cerebellum. Abnormalities in interoceptive awareness and central sensorimotor processing have also been described.

Different or deficient response-inhibition circuits have been identified, as well as the role of various neurotransmitters, receptor sensitivities and size of the caudate. Readers interested in the underlying neurobiology should see work by Worbe, Hartmann and colleagues in our suggested reading list.

History taking

In the history it is helpful to consider the points listed in Box 1.

Practice Point: be alert for the story of the child with recurrent visits to the optician or the allergy/ENT clinic as blinking, sniffing and throat clearing. These may be early undiagnosed tics.

Examination

Conduct a full neurological examination — examine for cerebellar signs, myoclonus, dystonia, chorea and tremor. If the movements are unusual look for features of functional movements (see below). Motor co-ordination problems can be co-morbid. Look for signs of neuro-cutaneous syndromes i.e. café au lait spots for Neurofibromatosis, and hypo-pigmented macules for tuberous sclerosis. Establish whether certain movements lead to dyskinesia (kinesogenic dyskinesia) or are exercise induced.

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