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Review

Clinical assessment of Tourette syndrome and tic disorders

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ABSTRACT

Tourette syndrome (TS) is a neuropsychiatric disorder involving multiple motor and phonic tics. Tics, which usually begin between the ages of 6 and 8, are sudden, rapid, stereotyped, and apparently purposeless movements or sounds that involve discrete muscle groups. Individuals with TS experience a variety of different sensory phenomena, including premonitory urges prior to tics and somatic hypersensitivity due to impaired sensorimotor gating. In addition to other conditions, stress, anxiety, fatigue, or other heightened emotional states tend to exacerbate tics, while relaxation, playing sports, and focused concentration on a specific task tend to alleviate tic symptoms. Ninety percent of children with TS also have comorbid conditions, such as attention deficit hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD), or an impulse control disorder. These disorders often cause more problems for the child both at home and at school than tics do alone. Proper diagnosis and treatment of TS involves appropriate evaluation and recognition, not only of tics, but also of these associated conditions.

Contents

1. Introduction

Tourette syndrome (TS) was first described by the French neurologist, Gilles de la Tourette, in 1885 as a "maladie des tics." In his original case series describing the syndrome that now bears his name, Gilles de la Tourette wrote about many of the characteristics of the syndrome including: involuntary movements and

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sounds, markedly enhanced startle reactions, a tendency to repeat both vocalizations (echolalia) and movements (echopraxia), and uncontrollable verbal obscenities (coprolalia) (Lajonchere et al., 1996). Since then, our knowledge of TS has progressed significantly, including advances in our understanding of tics, their surrounding sensory phenomena, and the central role that other co-occurring diseases, such as attention deficit hyperactivity disorder (ADHD) and obsessive-compulsive disorder (OCD), have on the overall clinical course of the disorder. This review will focus on our current understanding of the diagnosis, clinical characterization and assessment of tics as well as their clinical course. Other reviews will focus on the evidence-based treatment and neurobiology of tic disorders.

2. Definition of tics

Tics appear as sudden, rapid, purposeless motor movements or sounds that involve discrete muscle groups. They are also stereotyped in that they will occur in a similar manner each time they are performed. In comparison to some movement disorders or psychiatric conditions (e.g. sterotypies, chorea, or dyskinesia), patients with tics report the ability to suppress them, even if only for a short duration. However, they report that suppression often causes discomfort. Almost any movement, sound, or combination therein that the body can make can become a tic. Although some tics are more mild (i.e. eye blinking), others can be more severe to the point of causing pain to the patient (i.e. head or neck jerk). Apart from the physical consequences incurred by them, tics and their associated neuropsychiatric symptoms can diminish patients' quality of life, social and academic function, and lifetime achievements. They can also be very troubling and disruptive to the patients' family, and many times the entire family needs care and counseling (Leckman, 2012). Oftentimes, the tics themselves have less adverse effects than the co-occurring disorders. For instance, a 2011 study measuring quality of life (QoL) in fifty youth with TS found that symptoms of depression, OCD, and ADHD appeared to have a widespread negative impact on QoL; however, increased tic severity and poor QoL were not associated (Eddy et al., 2011).

3. Tourette syndrome and other tic disorders

The prevalence of TS varies based on study design and location. An international prevalence of 0.6–1% has been reported for mainstream schoolchildren, with the disorder being 3–4 times more common in males than in females (Cavanna and Termine, 2012). Data from the 2007 National Survey of Children's Health (NSCH) showed an estimated prevalence of 0.3% among U.S. children aged 6–17 years (Scahill et al., 2009). This number may represent an underestimate of TS prevalence since data were gathered from a parent-reported survey, and detection might be imperfect for children with fluctuating levels of symptoms or limited access to specialty health-care services (Scahill et al., 2009). Alternatively, TS prevalence may differ in prevalence worldwide due to either genetic or environmental differences. For example, TS has been reported to be less common in African-American people and has been reported only very rarely in sub-Saharan black African people (Robertson, 2008a). Regardless, the phenomenology of TS is similar in all cultures in which it has been reported (Robertson, 2008a).

TS is defined by the pediatric onset of both motor and vocal tics, lasting for at least one year. Although TS is the most notorious cause of chronic tics, there are types of tic disorders that are more common in children. Based on the Diagnostic and Statistical Manual – IV (DSM-IV) of the American Psychiatric Association, other tic disorders include: chronic motor tic disorder (CMT) and chronic vocal tic disorder (CVT), which are defined as having motor or phonic tics (but not both) for at least one year; and transient tic disorder (TTD), which is characterized by tics (either motor and/or vocal) for a duration of less than one year (DSM-IV-TR, 2000). Transient tics affect 15–25% of school-aged children with the majority experiencing resolution of tics within several months (Khalifa and von Knorring, 2003; Scahill et al., 2005; Robertson, 2008a,b). With the advent of DSM-V, the category of TTD is likely to be replaced by "Provisional Tic Disorder," as this designation is more accurate than TTD for patients with ongoing tic symptoms of less than one-year duration since onset (Walkup et al., 2010). Tic Disorders Not Otherwise Specified is the diagnostic term used for tic disorders that begin after age 18 or are secondary to other factors such as substance use (e.g. cocaine), toxins (e.g. carbon monoxide poisoning), or head trauma (e.g. physical trauma, stroke, or encephalitis) (Table 1).

Tics also exhibit several characteristics that distinguish them from other common childhood movement disorder such as stereotypies, choreas and dystonias. The distinguishing characteristics of tics include (1) they wax-and-wane in severity, (2) the character of the movements changes over time, (3) they are temporarily suppressible and (4) they are typically associated with sensory phenomena. Table 2 contrasts TS with other common movement and childhood psychiatric disorders confused with TS.

4. Characterization of tics

Tics are characterized by their anatomical location, number, frequency, and duration. They are also further described by their forcefulness or intensity and by their complexity (ranging from simple to complex). The most widely-used rating scale of tic severity is the Yale Global Tic Severity Scale (YGTSS), which includes separate scores from 0 to 5 for number, frequency, intensity, complexity, and interference (the degree to which planned actions or speech are interrupted by tics) of both motor and phonic tics (Leckman et al., 1989). This tool has allowed for the standardization of tic severity across different studies and research groups, aiding in the characterization and quantification of symptoms.

Additionally, because the clinical characteristics of TS make it hard for clinicians to diagnose and assess the severity of the condition, the Tourette Syndrome Diagnostic Confidence Index (DCI) was created through a collaborative effort of an expert group of clinicians. Based on the range and complexity of tics, their changeable nature, the temporal features of tic expression, and associated subjective and cognitive experiences, the DCI assigns a score from 0 to 100, which reflects the likelihood of having or ever having had TS (Robertson et al., 1999).

Other rating scales include the Shapiro Tourette Syndrome Severity Scale, Tourette's Syndrome-Clinical Global Impression Scale, and the Hopkins Motor and Vocal Tic Scale (Walkup et al., 1992). Standardized video recordings can also be used to count tics (Tanner et al., 1982). See Table 3 for a detailed comparison of various rating scales. For a detailed discussion on these rating scales, we suggest reading a recently published review (McGuire et al., 2012).

5. Natural history

The natural history of TS has been established based on clinical observations. There is a clear progression of the disorder from the onset of symptoms to, in most cases, full or partial regression of symptoms. Tics usually begin around 6–8 years of age, and 90–95% of TS cases have an onset of tics between the ages of 4 and 13 (Leckman et al., 1998). Simple motor tics involving the eyes or face are usually the first to appear in a child with TS. They are called simple because they involve a single contraction, such as a shoulder shrug or neck stretch. Motor tics will typically progress