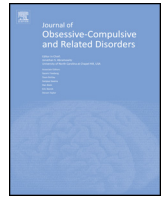




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Clinical features of Tourette syndrome and tic disorders



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ABSTRACT

Background: Tourette syndrome (TS) is a neurodevelopmental disorder characterized by motor and vocal tics—rapid, repetitive, stereotyped movements or vocalizations lasting at least one year in duration. The goal of this article was to review the long-term clinical course of tics and frequently co-occurring conditions in children with TS.

Methods: We conducted a traditional literature search to locate relevant articles regarding long-term outcome and prognosis in TS and tic disorders.

Results: Tics typically have an onset between the ages of 4 and 6 years and reach their worst-ever severity between the ages of 10 and 12 years. By age 10 years, most children are aware of nearly irresistible somatosensory urges that precede the tics. A momentary sense of relief typically follows the completion of a tic. Over the course of hours, tics occur in bouts, with a regular inter-tic interval. Tics increase during periods of psychosocial stress, emotional excitement and fatigue. Tics can become “complex” in nature and appear to be purposeful. Tics can be willfully suppressed for brief intervals and can be evoked by the mere mention of them. Tics typically diminish during periods of goal-directed behavior. Over the course of months, tics wax and wane. By early adulthood, roughly three-quarters of children with TS will have greatly diminished tic symptoms and more than one-third will be virtually tic free.

Conclusion: Although tics are the defining aspect of TS, they are often not the most enduring or impairing symptoms in children with TS. Indeed in TS tics rarely occur in isolation, and other coexisting conditions—such as behavioral disinhibition, hypersensitivity to a broad range of sensory stimuli, problems with visual motor integration, procedural learning difficulties, attention-deficit hyperactivity disorder, obsessive-compulsive disorder, depression, anxiety, and emotional instability—are often a greater source of impairment than the tics themselves. Measures used to enhance self-esteem, such as encouraging independence, strong friendships and the exploration of interests, are crucial to ensuring positive adulthood outcome.

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1. Introduction

“I finally apprehend the magnitude of the background noise that I have been experiencing for decades... the people around me do not share my tics because they do not hear the drum-beat. They do not feel the sensations without sources, do not have irresistible urges to pause in midsentence, do not receive strict internal commands to trace with their eyes in midair the shape of each of Aristotle’s regular solids, and so on in endless, bewildering variety... Finally and most important, I feel convinced that this complex challenging enigmatic internal world is the obvious core of Tourette’s (Hollenbeck, 2001).”

Tic disorders have been the subject of speculation for at least 300 years. In the past 25 years, Tourette syndrome (TS) has come

to be recognized as a model developmental disorder occupying the nexus of neurology and psychiatry (Kushner, 2000). TS is a childhood-onset neuropsychiatric disorder characterized by multiple motor and vocal tics that last at least a year in duration (Walkup, Ferrão, & Leckman, 2010). Once thought to be a rare condition, TS is known to affect four to six in every 1000 children (Khalifa & von Knorring, 2003, 2006; Robertson, Eapen, & Cavanna, 2009; Scahill, Dalsgaard, & Bradbury, 2013). Transient tic disorder, characterized by tics that are present for less than a year in duration, and chronic tic disorders, which are characterized by either motor or vocal tics, but not both, have a prevalence of approximately 20% and 5%, respectively, in children (Khalifa & von Knorring, 2003, 2006; Robertson et al., 2009; Scahill et al., 2013). Individuals with TS commonly experience comorbid psychiatric disorders such as attention-deficit hyperactivity disorder (ADHD) and obsessive-compulsive disorder (OCD) (Ferrão et al., 2013; Khalifa & von Knorring, 2003, 2006; Leckman, 2002; Leckman, Bloch, Sukhodolsky, Scahill, & King, 2013; Rothenberger & Roessner, 2013;

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Robertson et al., 2009; Scahill, Sukhodolsky, Williams, & Leckman, 2005; Scahill et al., 2013). These comorbid conditions often cause more impairment in individuals with TS than the tics themselves.

The purpose of this article is to review the clinical course of individuals with TS. We begin by focusing on the nature and course of tic symptoms in TS, then focus on the clinical course of common comorbid conditions and finish by discussing the adulthood global functioning and psychosocial outcome of children with TS (Course, 2013). The conclusion will summarize, for clinicians, the prognostic information we use to explain the expected clinical course of TS to families seeking evaluation in our clinic.

2. Tic symptoms

A tic is a sudden, repetitive, non-rhythmic, stereotyped motor movement or vocalization involving discrete muscle groups (Leckman et al., 2013). Tics can be described based on their anatomical location, number, frequency, and duration (Leckman, Riddle, & Hardin, 1989). Another useful descriptor is the intensity or “forcefulness” of the tic, as some tics call attention to themselves simply by virtue of their exaggerated, forceful character (Leckman et al., 1989). Tics can also be described in terms of their “complexity” (Leckman et al., 1989). Complexity refers to how simple or involved a movement or sound is, ranging from brief, meaningless, abrupt fragments (simple tics) to ones that are longer, more involved, and seemingly more purposive in character (complex tics). Simple tics are sudden, brief (usually less than 1 s in duration), meaningless movements or sounds. Complex tics are sudden, more purposive appearing, stereotyped movements of longer duration that can include “orchestrated” combinations of motor or vocal or motor and vocal tics. The observed range of tics is extraordinary, so that virtually any voluntary motor movement or vocalization can emerge as a tic.

Tics usually have their onset in the first decade of life around the age of 4–6 years with simple motor movements such as eye blinking, nose twitching, or facial grimaces (Leckman et al., 2013). Motor tics usually progress in a rostrocaudal direction with time (Leckman, Zhang, & Vitale, 1998). Vocal tics, when they appear, typically first manifest themselves a year or two after the onset of motor tics (Leckman et al., 1998). Vocal tics begin as simple vocalizations such as throat clearing, sniffing, or fractions of words (Leckman et al., 1998).

Many TS patients experience premonitory urges (Leckman, Walker, & Cohen, 1993; Woods, Piacentini, Himle, & Chang, 2005). A premonitory urge is a sensory phenomenon that occurs immediately prior to a tic, similar to the need to sneeze or scratch an itch (Leckman et al., 1993; Woods et al., 2005). Individuals often describe the need to tic as the buildup of tension, usually in the body location where the tic is about to occur, but in some individuals the tension occurs throughout the body (Leckman et al., 1993; Woods et al., 2005). Depending on the intensity of the urge the individual may consciously decide to tic or not to tic. However, if the urge is very strong, it can be impossible to resist. The actual tic may be felt as relieving this tension or sensation, similar to scratching an itch. After a tic is done, there is often a fleeting and incomplete sense of relief. Awareness of premonitory urges increases with age and is present in as many as 90% of adolescents with TS (Leckman et al., 1993; Woods et al., 2005). Gaining an awareness of these difficult to describe premonitory urges lies at the heart of habit reversal training which has proven to be one of the most efficacious treatments for TS (Piacentini, Woods, & Scahill, 2010; Wilhelm et al., 2012).

During the course of a day tic symptoms of TS generally occur in bouts and over weeks to months they wax and wane in severity (Leckman, 2002; Peterson & Leckman, 1998). Factors such as

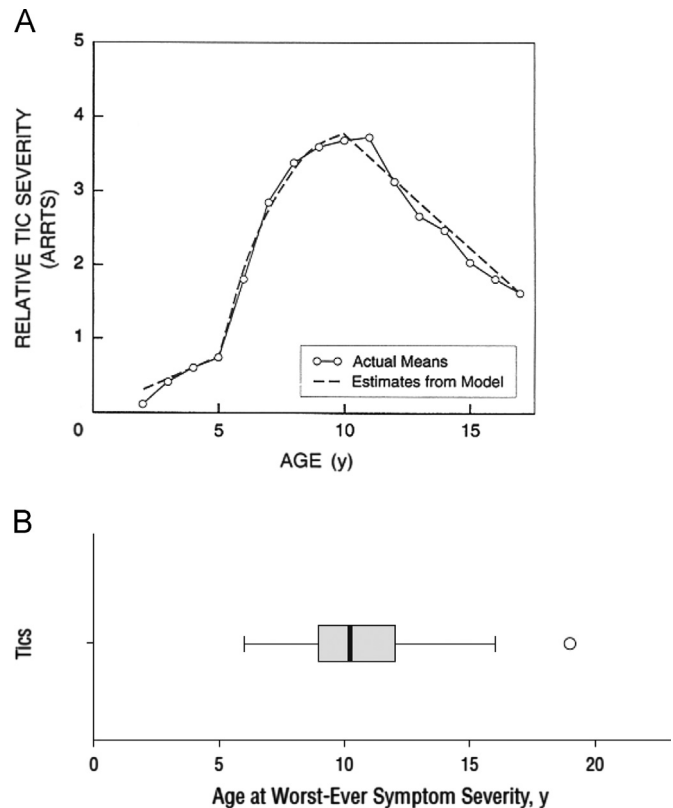


Fig. 1. Course of tic severity over the first two decades. (A) Data on the relative severity of tics are presented for 42 individuals with Tourette syndrome (A–D) (Leckman 2002). The relative tic severity scale goes from 0 (no tics) to 5 (severe tics). Estimates from the patient and a respective parent were obtained independently. (B) Box plot representing age when tic disorder symptoms were at their worst ($N=46$) (Leckman et al., 1993).

psychosocial stress, excitement, anxiety, social events and fatigue are known to exacerbate tics in the moment and psychosocial stress has been shown to be predictive of future tic severity (Buse, Kirschbaum, Leckman, Münchau, & Roessner, 2014; Conelea & Woods, 2008; Lin, Katsovich, & Ghebremichael, 2007; Steinberg et al., 2013). However, for many individuals, focused concentration, especially involving fine-motor control such as playing a musical instrument, dancing, or playing sports, can lead to a marked reduction of tics (Leckman et al., 2013). Many TS patients are able to temporarily suppress tics, but often at the expense of concentration and exhaustion (Leckman et al., 2013). As children get older they typically develop a greater ability to suppress tics (Leckman et al., 2013). Interestingly, recent studies suggest that environments conducive to tic suppression result in reduced tic frequency without adverse consequences or a rebound in tic frequency or severity (Specht et al., 2013).

When Georges Gilles de la Tourette originally described the clinical syndrome that now bears his name, tics were hypothesized to be persistent and lifelong; our current knowledge suggests this is generally not the case (Bloch, Peterson et al., 2006; Course, 2013; Gilles de la Tourette, 1885; Leckman et al., 1998). Tics typically emerge around the age of 4–6 years, reach their worst ever point early in the second decade of life, and then, on average, improve during adolescence (Bloch et al., 2006; Course, 2013; Leckman et al., 1998). Fig. 1 demonstrates the clinical course of tic severity in two cohorts of 42 and 46 children, respectively, with TS followed until early adulthood (Bloch et al., 2006; Leckman et al., 1998).

Fig. 2 describes the early adulthood tic outcome of more than 80 children followed from initial evaluation (average age 11 years) to

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