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Clinical Descriptions and Diagnostic Requirements for ICD-11 Mental, Behavioural or Neurodevelopmental Disorders Tourette syndrome Essential (required) features • The presence of both motor tics and phonic tics that may or may not manifest concurrently or continuously during the symptomatic course is required for diagnosis. • Motor and phonic tics are defined as sudden, rapid, non-rhythmic and recurrent movements or vocalizations, respectively. • Motor and phonic tics have been present for at least 1 year with onset during the developmental period. • The symptoms are not a manifestation of another medical condition (e.g. Huntington disease), and are not due to the effects of a substance or medication on the central nervous system (e.g. amphetamine), including withdrawal effects (e.g. from benzodiazepines). Additional clinical features • Tourette syndrome frequently co-occurs with attention deficit hyperactivity disorder, and impulsivity, disinhibition, anxiety and immature behaviour may be associated features of both diagnoses. • Motor and phonic tics in Tourette syndrome may be voluntarily suppressed for short periods of time, may be exacerbated by stress, and may diminish during sleep or during periods of focused enjoyable activity. • Tics are often highly suggestible – for example, when an individual with Tourette syndrome is asked about specific symptoms, old tics that have been absent for some time may transiently reappear. Boundary with normality (threshold) • Transient motor or phonic tics (e.g. eye blinking, throat clearing) are common during childhood, and are differentiated from Tourette syndrome by their transient nature. 8A05.00 Neurodevelopmental disorders | Secondary-parented categories in neurodevelopmental disorders

155 Neurodevelopmental disorders Course features • The onset of Tourette syndrome commonly occurs during childhood (between the ages of 4 and 6 years), with peak symptom severity occurring between the ages of 8 and 12 years. Across adolescence, there is decreasing likelihood of onset. Onset during adulthood is rare and most often associated with severe psychosocial stressors, use of specific drugs (e.g. cocaine) or an insult to the central nervous system (e.g. post-viral encephalitis). • The onset of Tourette syndrome is typically characterized by transient bouts of simple motor tics such as eye blinking or head jerks. Phonic tics usually begin 1-2 years after the onset of motor symptoms and initially tend to be simple in character (e.g. throat clearing, grunting, or squeaking), but then may gradually develop into more complex vocal symptoms that include repetition of one's own or another person's speech or obscene utterances (i.e. coprolalia). Sometimes the latter is associated with gestural echopraxia, which also may be of an obscene

nature (i.e. copropraxia). • Vocal and/or motor tics may wax and wane in severity, with some individuals experiencing remission of symptoms for weeks or months at a time. Eventually the symptoms become more persistent, and can be accompanied by detrimental effects to personal, family, social, educational, occupational or other important areas of functioning. • The majority of individuals with Tourette syndrome will experience significantly diminished symptoms by early adulthood, with more than one third experiencing a full remission of symptoms. • Evidence suggests a good long-term clinical course for individuals with a solitary diagnosis of Tourette syndrome. Those with co-occurring conditions (e.g. obsessive-compulsive disorder, attention deficit hyperactivity disorder, anxiety and fear-related disorders, depressive disorders) tend to exhibit a poorer prognosis. Developmental presentations • The prevalence rate of Tourette syndrome among school-aged children has been estimated at approximately 0.5%. • Motor and phonic tics in Tourette syndrome tend to be most severe between the ages of 8 and 12 years, gradually diminishing throughout adolescence. By late childhood (approximately 10 years of age), most children become aware of premonitory urges (bodily sensations) and increased discomfort preceding – and relief of tension following – motor and vocal tics. • The vocal symptom of coprolalia (inappropriate swearing, experienced involuntarily) is uncommon, affecting only 10–15% of individuals with Tourette syndrome, and tends to emerge in mid-adolescence. • Many adults with childhood-onset Tourette syndrome report attenuated symptoms, though a small number of adults will continue to experience severe tic symptoms. Neurodevelopmental disorders | Secondary-parented categories in neurodevelopmental disorders

Clinical Descriptions and Diagnostic Requirements for ICD-11 Mental, Behavioural or Neurodevelopmental Disorders • The pattern of co-occurring disorders appears to vary with developmental stage. Children with Tourette syndrome are more likely to experience attention deficit hyperactivity disorder, obsessive-compulsive disorder, autism spectrum disorder and separation anxiety disorder compared to adolescents and adults. Adolescents and adults are more likely than children to develop a depressive disorder, a disorder due to substance use or a bipolar disorder. Culture-related features • Symptoms of Tourette syndrome are consistent across cultural groups. • If vocalizations or movements have a specific function or meaning in the context of an individual's culture and are used in ways that are consistent with that cultural function or meaning, they should not be considered evidence of Tourette syndrome. Sex- and/or gender-related features • Tourette syndrome is more common among males than females (gender ratio ranging from 2:1 to 4:1). • Course and symptom presentation do not vary by gender. • Women with persistent tic disorders may be more likely to experience co-occurring anxiety and fear-related disorders and depressive disorders. Boundaries with other disorders and conditions (differential diagnosis) Boundary with autism spectrum disorder and stereotyped movement disorder Repetitive and stereotyped motor movements such as whole-body movements (e.g. rocking) and unusual hand or finger movements can be a characteristic feature of autism spectrum disorder and of stereotyped movement disorder. These behaviours can appear similar to tics, but are differentiated because they tend to be more stereotyped, last longer than the duration of a typical tic, tend to emerge at a younger age, are not characterized by premonitory sensory urges, are often experienced by the individual as soothing or rewarding, and can generally be interrupted with distraction. Neurodevelopmental disorders | Secondary-parented categories in neurodevelopmental disorders

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