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The prevalence and epidemiology of Gilles de la Tourette syndrome Part 2: Tentative explanations for differing prevalence figures in GTS, including the possible effects of psychopathology, aetiology, cultural differences, and differing phenotypes

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Abstract

As has been demonstrated, Gilles de la Tourette Syndrome (GTS) occurs in at least 1% of the population worldwide. However, earlier studies suggested a lower prevalence. In addition, the prevalence figures for different studies very between 0.4% and 3.8%. Moreover, the prevalence appears to vary in some parts of the world and races, with a lower rate in Afro-Americans and sub-Saharan black Africans. In this the second part of the review, possible reasons for the differences in prevalence and epidemiology are discussed. Tentative explanations for differing prevalence figures in GTS include problems with the diagnosis of GTS, the multidimensional nature of tics, as well as other tic factors including the waxing and waning of symptoms and the suppressibility of symptoms. Other factors inherent to GTS include the fact that there is no diagnostic test and indeed no definitive diagnosis other than clinical, the fact that psychosocial stresses can lead to increased tic severity, and that comorbid disorders may mask tics. The varying methods of study employed can also effect prevalence. There may be some regional differences in GTS as well, which may be due to a lack of awareness of GTS, or it may be a true reflection of low prevalence as in some populations GTS does appear rare. With regard to the sub-Saharan Africa data and possibly the African American data, matters are much more complex than meets the eye. The following reasons are all possible for the apparent rarity in these populations and include (i) other medical priorities and less propensity to seek health care, (ii) lack of awareness of GTS, (iii) chance, (iv) ethnic and epigenetic differences and reasons, (v) genetic and allelic differences in different races, and (vi) an admixture of races. The aetiology of GTS is also complex, with influences from complex genetic mechanisms, pre- and perinatal difficulties and, in a subgroup, some infections, possibly by epigenetic mechanisms. These may well affect phenotype and, thus, prevalence. There have even been suggestions that people with GTS are increasing. Recent data suggests that GTS is not a unitary condition and that there may well be different types of GTS. The prevalence of GTS in these individual subtypes is unknown. It is suggested that a new nomenclature be adopted for GTS in future, pending further genetic and phenomenological studies. To what extent the aetiology affects the phenotype and, thus, the prevalence is still unclear.

Keywords: Gilles de la Tourette Syndrome; Prevalence; Phenotype; Psychopathology; Aetiology; Cultural differences

Introduction

The generally accepted international diagnostic criteria for Gilles de la Tourette Syndrome (GTS), a childhood-onset neuropsychiatric disorder, include multiple motor tics and one or more phonic (vocal) tics or noises, lasting longer than a year [1,2]. The prevalence and epidemiology of GTS are more complex than was once thought. In a recent study and review by the present author, Robertson (Part 1 of this duo [3]) collected the data from 14 studies undertaken in mainstream school and school-age youngsters in the community using similar multistage methods, scrutinized the data, and reported

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prevalence figures for GTS of between 0.4% and 3.8% for youngsters between the ages of 5 and 18 years. Of the 420,312 young people studied internationally, 3989 (0.949%) were calculated as having been diagnosed as having GTS. It was therefore suggested that a figure of 1% would be appropriate for the overall international GTS prevalence figure. There were, however, "outliers" to the figure: for instance, GTS does seem to be substantially rarer in the Xhosa youngsters (Black South African people). With regards epidemiology, GTS is found in all cultures, although to differing degrees, and is not common in the North American "African-American" and has not been reported in sub-Saharan black Africa countries, that is, other than a few GTS individuals identified in the South African Xhosa study. In all cultures where GTS has been reported, the phenomenology is similar, highlighting the biological underpinnings of the disorder [3].

In order to understand the prevalence and epidemiology of GTS, one must take the disorder in context, with regard to clinical phenomenology, psychopathology, and possible phenotypes, as well as the complex aetiological theories, as they almost certainly all affect the prevalence data.

Tentative explanations for differing prevalence figures in GTS

What are the reasons for these differing results in prevalence? Suggestions have included problems with the diagnosis of GTS; the fact that tics are probably multidimensional in nature; which dimensions run along varying continuums including intensity of symptoms (from mild to severe), frequency of symptoms (from rare to constant), a variety of symptoms (single and/or multiple tic groups), complexity of tics (simple to highly complex), and comorbid psychiatric disorders (from none to multiple: which in turn affect disability); and the fact that there is no diagnostic test and indeed no definitive diagnosis other than clinical; the varying methods of study employed; the fact that symptom intensity and frequency decreases with age and affected individuals are often unaware of their tics, as reported by Leckman et al. [4], Robertson and Gourdie [5], Pappert et al. [6], Tanner and Goldman [7], Kuperman [8], Stefanoff and Mazurek [9], and Scahill et al. [10]. In addition, psychosocial stresses can lead to increased tic severity [11]. Other factors such as the waxing and waning of symptoms, the suppressibility of symptoms, the fact that comorbid disorders may mask tics, and the fact that tics may reduce with the treatment of comorbid disorders complicate research further. Nevertheless, the overall prevalence rate worldwide, apart from sub-Saharan black Africa, in studies using similar multistage methods, is 1% of youngsters.

Some problems with the nosology and diagnosis of GTS

As mentioned briefly in Part 1 by Robertson [3], for a diagnosis of GTS to be made, the current international diagnostic criteria demand both motor and phonic tics. However, this may be well somewhat arbitrary as, for example, sniffing is certainly a sound. It was once considered to be a motor tic but "evolved" to become a vocal/phonic tic. Only tics which are actually from the vocal cords are truly vocal, such as sounds including coprophenomena, echophenomena, and actual words. Throat clearing, coughing, and gulping are probably somewhere in the middle but are now always considered as vocal/phonic tics. It is for that reason that "sound" tics are often currently referred to as phonic rather than vocal—so as not to imply the vocal cords.

In addition, the *Diagnostic and Statistical Manual of Mental Disorders* (*DSM*) American Psychological Association [1] criteria have changed over time. For example, it has changed the upper age limit (under which GTS must start) suggesting 15, 21, and 18 years, which is somewhat arbitrary. Also, in *DSM*, Fourth Edition (*DSM-IV*), distress and impairment were added to the criteria which met with much opposition. Finally, in *DSM-IV* it was stated that "during the (1 year) there was never a tic-free period of more than 3 consecutive months"; there seemed to be no evidence base for this, and thus, yet another arbitrary criterion had been introduced. *DSM-IV* came under such criticism that *DSM-IV, Text Revision* [1] changed, and the three above criteria were eliminated. Many researchers, in the intervening periods, used *DSM, Revised Third Edition* or other standard [e.g., World Health Organization (WHO)] criteria [2].

In addition, in the WHO [2] criteria, there has never been an age of onset stipulation, with the result that some "adult onset tic disorders" may be included in the GTS umbrella in some studies in Europe, while similar cases will be excluded in the United States and other countries employing *DSM* criteria. These "adult-onset tic disorders" have indeed been documented from Canada [12] and the United Kingdom, [13], but they often had different aetiologies, such as being secondary to infections, trauma, or noxious agents. In other words, they were different to "pure or primary" GTS. However, had the age of onset of these patients been within the *DSM* requirements at the time, they may have received a diagnosis of GTS. Furthermore, others have described GTS commencing before 1 year of age [14]. These data highlight the complexity of diagnosis of GTS and related tics disorders even further.

In addition, many clinicians erroneously believe that coprolalia must be present for the diagnosis to be made; therefore, the diagnosis is still not made fairly frequently by inexperienced clinicians. Thus, it is well known that many patients with GTS have attracted a variety of incorrect diagnoses including myoclonic epilepsy before the correct diagnosis by an expert specialist [15,16].

The lifetime prognosis of GTS and its effect on symptoms and, thus, prevalence

It was initially thought that GTS was lifelong, as mentioned in Part 1, but then Erenberg et al. [17] first