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The prevalence of tic disorders and clinical characteristics in children

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ABSTRACT

Background: Prevalence is a simple statement about the frequency of a disease in the population. For many medical conditions, including Tourette syndrome, there are true cases that have not been previously diagnosed due to problems of access to appropriate clinical services. Therefore, to obtain a trustworthy estimate of prevalence, it is necessary to go beyond cases identified in clinical settings and evaluate community samples.
Method: We reviewed 11 community surveys in children with Tourette syndrome (TS) published since 2000. We also examined the frequency of co-occurring psychiatric conditions in community samples and large clinically-ascertained samples.
Results: Transient tics are relatively common affecting as many as 20% of school-age children. The 11 studies reviewed here offer a wide range of estimates from 2.6 to 38 per 1000 children for TS. Six studies provide estimates in a narrower range from 4.3 to 7.6 per 1000 but the confidence interval around this narrower range remains wide. Six studies provided results on chronic tic disorders ranging from 3 to 50 per 1000 for Chronic Motor Tic Disorder and 2.5 to 9.4 per 1000 for Chronic Vocal Tic Disorder. Community samples and large clinically-ascertained samples consistently show high rates of ADHD, disruptive behavior and anxiety disorders in children with TS.
Conclusions: The wide range of prevalence estimates for TS and chronic tic disorders is likely due to differences in sample size and assessment methods. The best estimate of prevalence for TS in school-age children is likely to fall between 4 and 8 cases per 1000. Clinical assessment of children with chronic tic disorders warrants examination of other problems such as ADHD, disruptive behavior and anxiety.

1. Introduction

Epidemiology considers the prevalence of disease and compares the characteristics of those affected by the condition to those that are not affected. Characteristics that are associated with the disorder under study may prove to be factors that increase risk for the disorder upon further study. Prevalence, which is a simple statement about the frequency of the condition, is an essential metric for understanding the public health impact of the condition of interest. Other essential elements in the assessment of public health impact are the morbidity and mortality associated with the condition. For example, a highly prevalent condition associated with mild disability would have less public impact than a condition of similar frequency marked by greater disability. In chronic neuropsychiatric disorders such as Tourette syndrome (TS), the focus is on morbidity (i.e. disability) rather than mortality.

Obtaining a trustworthy estimate of prevalence of TS faces several challenges. First, there is no objective test for the diagnosis. Second, known and unknown biases can influence who comes to clinical attention. Therefore, counting clinical cases is not a valid approach for estimating prevalence because cases in the community that have not been diagnosed will be missed. Third, by extension, valid estimates of prevalence rely on community surveys. However, the identification and assessment of a large community sample to evaluate prevalence of TS is an expensive proposition. Fourth, TS resides on a spectrum from mild to severe. Separating affected individuals from unaffected may be difficult. For example, a child with a few motor and vocal tics that come and go and do not cause a problem may not be a case of TS. This issue here is whether impairment due to tics should be included in the case definition.

An additional benefit of community sampling is that associations of TS identified in these samples can confirm or contest the observed associations in clinical samples. For example, ADHD is a common co-occurring condition in clinical samples of children with TS. The strength of this association maybe exaggerated because children with TS and ADHD maybe more likely to come to clinical attention than children with TS alone. Results from community samples may help to settle the matter on the association of TS and ADHD. Finally, examination of the full TS spectrum

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in community samples may also uncover gaps in mental health services. It may be that even children with mild TS identified in a community survey have health service needs that are not being met. Once uncovered, appropriate steps can be taken to fill these gaps and reduce the disability that accompanies chronic tic disorders in children. The purpose of this paper is to describe the prevalence and associated disability of TS in children. To these ends, we review community surveys on the prevalence of TS and tic disorders in children since 2000. The review also examines the co-occurrence of other psychiatric disorders in community samples and selected large clinically-ascertained samples.

2. Prevalence of tic disorders in childhood

Isolated and transient tics are relatively common in school age children ranging from 11% to 20% (Cubo et al., 2011; Kurlan et al., 2001; Linazasoro, Van Blercom & de Zarate, 2006; Snider, Seligman & Ketchen, 2002) with a male to female ratios between 2 to 1 and 3.5 to 1. For most of these children, the tics were mild. Because most studies were cross-sectional, it is impossible to know how many children with transient tics would become cases of TS or chronic tic disorder over time. Because there are no diagnostic tests for tic disorders, the diagnosis relies on history and observation. The Diagnostic and Statistical Manual – Fifth Edition – revised (American Psychiatric Association, 2013) defines three tic disorders of interest here. Provisional Tic Disorder consists of motor tics, vocal tics or both lasting less than one year. Persistent Tic Disorder is defined by the presence of motor or vocal tics (but not both) lasting for more than a year. Tourette's Disorder (also known as Tourette syndrome) includes multiple motor tics and at least one vocal tic lasting for a more than a year. For each of these tic disorders, the diagnostic criteria specify the onset of tics before 18 years of age. Although tics may be chronic, tics often show a fluctuating course with a commonly observed tendency to rise and fall in frequency and intensity over time. Tics are suppressible – for at least brief periods of time. By the age 10 years, most patients describe a warning or urge before some or all of their tics and momentary relief after the execution of the tic (Leckman, 2002).

Community surveys conducted in various countries over the past 20 years provide estimates of prevalence for TS ranging from 0.5 to 38 cases per 1000 children (reviews Scahill, Sukhodolsky, Williams &Leckman, 2005; Hirtz et al., 2007; Robertson, 2008). The lower bound of 0.5 per 1000 came from a survey of Israeli army inductees (Apter et al., 1993). This survey (not listed in Table 1) relied on self-reports from inductees in the Israeli army. The subjects were 16 to 18 years of age, when tics decline is a high percentage of cases (Bloch et al., 2006). It is likely that a parent interview focused on lifetime diagnosis would have identified more cases. The upper bound of 38 per 1000 came from a study of 1255 school children (Kurlan et al., 2001). Similarly, Cubo et al. (2011) reported a prevalence of 36.4 per 1000. These two studies each used two approaches to define cases: TS with impairment and TS without impairment. Not surprisingly, when impairment was included in the case definition, the prevalence estimate went down to 8 per 1000 (Kurlan et al., 2001) and 16 per 1000 (Cubo et al., 2011). If these estimates at the extremes are disregarded, the resulting range of prevalence from studies conducted over the past decade is 1 to 16 per 1000. Although narrower than the extremely broad range of 0.5 to 38 per 1000, 1 to 16 per 1000 remains imprecise and insufficient to guide estimates of service needs for affected children. For example, the population of children between 6 and 18 years in the United States is roughly 50 million. If the prevalence is 1 per 1000, that would translate into 50,000 cases of TS nationwide. If the prevalence is 10 per 1000, the number of cases jumps to 500,000. The variation in estimates across these community studies is likely due to differences in sampling method, sample size, the rate of subject participation, assessment methods and diagnostic threshold used to define cases.

That the estimate of prevalence would be influenced by the symptom threshold used to define the disorder is clear. Simply stated, if children with mild forms of TS are defined as true cases, the prevalence will increase. If the severity threshold is set higher or includes a requirement of impairment, the prevalence will be lower. What may be less clear is the impact of case definition on associated features. For example, it may worthwhile to determine whether specific associations such as ADHD, anxiety or learning disability hold across the range of severity from mild to more extreme.

Using Medline, we searched with several key words (Tourette syndrome, tic disorders, epidemiology, and prevalence) to identify prevalence studies published since 2000. To identify studies missed by our literature search, we also consulted recent reviews (Scahill et al., 2005; Hirtz et al., 2007; Robertson, 2008). Studies included in the review were those that provided a lifetime diagnosis of one or more tic disorders. We calculated the 95% confidence interval from the data provided in each report to allow comparison across studies (the legend in Table 1 shows the formula used).

Table 1 presents the lifetime prevalence estimates for TS (per 1000) from 11 community surveys from various countries. All but two of the studies in Table 1 used a multi-stage design. In the typical two-stage design, the sample is screened for tic disorders (Stage One) followed by a diagnostic assessment (Stage Two). The ideal screening procedure is simple and relatively inexpensive. At the same time, the screening procedure should not “miss” many cases (false negatives) and not have too many false positives (cases those screen positive but are not true cases). A screen that does not miss many cases has high sensitivity. On the other hand, a screen with a high percentage of false positives has low specificity. Low specificity is a problem because the more detailed and expensive diagnostic assessment would be conducted on a large number of unaffected subjects. No screen is perfect. But an efficient screen must somehow avoid missing cases (false negatives) and avoid the expense of conducting unnecessary diagnostic assessments on unaffected subjects (false positives). In a disorder such as TS, the screening test may be a brief set of questions to the parent about the presence of tics in the child. Available data suggest that parents and teachers do not agree and that teachers miss more cases of tic disorders than parents (Hornsey, Banerjee, Zeitlin & Robertson, 2001). To protect against the possibility of missing cases in the screening phase, a well-designed community survey includes evaluation of at least a subsample of randomly selected false negatives. In TS community surveys, however, this has rarely been done.

The wide range of sample sizes across studies presented in Table 1 is striking (range 435–9712). For example, consider two imaginary studies each with a prevalence estimate of 7 cases of TS per 1000 children. A study with a sample size of 1000 would have a 95% confidence interval of the other with 5000. If the observed prevalence was 7 cases per 1000 children, the 95% confidence interval for the sample size of 1000 would be 2–12 cases per 1000 compared to 6–8 cases per 1000 in a sample of 5000.

Studies with smaller sample sizes prompt obvious questions concerning the representativeness of the sample and, as noted, result in wide confidence interval. For example, Kadesjo and Gillberg (2000) estimated a prevalence of 11 per 1000, which is greater than the upper bound of the 95% confidence interval (CI) for all but three studies presented in Table 2. The 95% CI of 4 to 27 per 1000 indicates that 4 per 1000 is equally plausible as 27 per 1000. The 4 per 1000 figure is consistent with findings of several studies presented in Table 1. By contrast, the upper limit of the confidence interval (27 per 1000) is exceeded by only two studies (Cubo et al., 2011; Kurlan et al., 2001). Taken together, these