The prevalence of Gilles de la Tourette syndrome in children and adolescents with autism: a large scale study

S. BARON-COHEN,1 V. L. SCAHILL, J. IZAGUIRRE, H. HORNSEY and M. M. ROBERTSON

From the Department of Experimental Psychology and Psychiatry, University of Cambridge; Children’s Centre, Southport; and Department of Psychiatry and Behavioural Sciences, University College London and the National Hospital for Neurology and Neurosurgery, London

ABSTRACT

Background. An earlier small-scale study of children with autism revealed that 8.1% of such patients were co-morbid for Gilles de la Tourette syndrome (GTS). The present study is a large scale test of whether this result replicates.

Method. Four hundred and forty-seven pupils from nine schools for children and adolescents with autism were screened for the presence of motor and vocal tics.

Results. Subsequent family interviews confirmed the co-morbid diagnosis of definite GTS in 19 children, giving a prevalence rate of 4.3%. A further 10 children were diagnosed with probable GTS (2.2%).

Conclusions. These results indicate that the rate of GTS in autism exceeds that expected by chance, and the combined rate (6.5%) is similar to the rates found in the smaller-scale study. Methodological considerations and alternative explanations for an increased prevalence are discussed.

INTRODUCTION

Gilles de la Tourette syndrome, or Tourette syndrome (GTS) is a neurodevelopmental disorder defined by the presence of chronic, multiple motor and vocal tics of childhood onset (American Psychiatric Association, 1987). The average age of onset is reported to be 5 years of age (Leckman et al. 1998). The tics show a fluctuating course and may decrease in severity during adulthood. The precise aetiology of GTS is unknown. In most cases however, GTS appears to be genetically transmitted (Curtis et al. 1992), although the exact pattern of inheritance is still unknown. Implicated neurological abnormalities include dysfunction of the basal ganglia and/or the prefrontal cortex (reviewed in Chase et al. 1986), and biochemical abnormalities of the dopamine and serotonin neurotransmitter systems (reviewed in Baker et al. 1995). GTS is often accompanied by obsessive–compulsive behaviours (Frankel et al. 1986; Eapen et al. 1997a). These may be an alternative expression of the putative GTS gene(s) (Pauls et al. 1986).

Autism is also a neurodevelopmental disorder, itself defined by abnormal social and communication development, with a pattern of restricted and repetitive interests and activities (American Psychiatric Association, 1994). Autism has an earlier age of onset than GTS (usually by 18 months of age) and often shows a chronic course. The precise aetiology of autism is also unknown, although possible aetiological factors include genetic (Folstein & Rutter, 1977; Bailey et al. 1995), neurological (Bauman & Kemper, 1994) and cognitive (Frith, 1989; Baron-Cohen, 1995) abnormalities.

Recently, a growing number of case-reports have documented the co-occurrence of autism and GTS in the same individuals. Realmuto &
Main 1982) were the first to report the development of GTS in a child with autism. These authors interpreted this as a chance association, as did a subsequent report of GTS in autism (Barabas & Matthews, 1983). However, Barabas & Matthews also discussed the possibility of a common neurochemical abnormality.

The estimated general population prevalence of autism is 1 per 1000 (Baron-Cohen et al., 1996) and the estimated general population prevalence of GTS is 2 per 10000 (Robertson, 1994). This is likely to be an underestimate, as GTS may go undetected. Hence, if autism and GTS are truly independent, the rate of co-occurrence expected by chance would be 2 per 10 million of the general population, 1 per 1000 individuals with GTS, and 2 per 10000 individuals with autism.

A few studies have documented the prevalence of GTS in populations of individuals with autism spectrum disorders. Kerbeshian & Burd (1986) reported a clinical series of six individuals with Asperger’s syndrome, of whom 3 (50–0%) had co-morbid GTS. The same group (Burd et al., 1987) reported a rate of 20–3% of GTS in an ascertained sample of 59 individuals meeting DSM-III criteria for infantile autism or pervasive developmental disorder (PDD). Of the 12 children with co-morbid GTS, 10 had an atypical PDD, while only two had autism. This group also showed significantly higher IQ than the group with PDD without GTS, and significantly higher measures of receptive and expressive language. Burd et al. believed this to be an indication that co-morbid GTS in children with autism provided a marker for improved developmental outcome. Kano et al. (1987, 1988) described two children with autism and GTS, drawn from a sample of 76 children with autism, suggesting a much lower prevalence of 2.6%.

We are not aware of any studies reporting the rate of co-morbid autism and GTS in the general population. However, Sverd (1991) reported 10 children with autism or PDD and co-morbid GTS. Given the population statistics for the geographical regions from which these children were clinically ascertained, Sverd argues that these children represent a rate of co-morbidity exceeding that of chance.

Although distinct disorders, autism and GTS share several behavioural features. Here we list some of these shared features, while pointing out how these may differ in the two syndromes: (1) echolalia and palilalia are common in both GTS and autism, although in autism, unlike GTS, these behaviours may be appropriate to the level of speech development; (2) types of obsessive–compulsive behaviours are frequently seen in both autism and GTS, although in autism these may be better described as rigid and ritualistic behaviours, such as an insistence upon sameness and resistance to change; (3) like GTS, autism is associated with abnormal motor behaviours, although in autism these often take the form of stereotypies, such as spinning, rocking and hand flapping.

A study carried out in a special school for children with autism (Baron-Cohen, et al. 1999), found that three out of the 37 pupils (8.1%) had co-morbid GTS. Previous studies had generated prevalence rates retrospectively from clinical series. A special school population of children and adolescents with autism was used, with a prospective, multi-stage design, using direct observation in the classroom, and, later, both pupil and family interviews. This may have produced a more accurate estimate of the prevalence of GTS in children with autism than previous studies. However, the sample size was small. The current study aimed to replicate the earlier study, to establish the rate of GTS in a special school population, but this time with a much larger sample.

METHOD

Participants

Thirty-three schools for children with autism from around England were invited to take part in the study. Of these, nine schools agreed to participate within the timescale of the study. We have no reason to suspect these were not representative of children with autism spectrum conditions more generally, as three schools were in the north, four were in the midlands, and two were in the Greater London area. The schools catered for a mixture of day and residential students. The total number of children with autism within these schools was 458, with a mean age of 11:1 (years:months; range 3:6 to 19:8) and a sex ratio of 49:1 (male:female). The children’s parents were contacted, via the schools, to request their consent for them to take part in the study. The parents of two children
(0.4%) declined to take part. Nine children for whom parental consent was given were unavailable for observation at the time of the study. Hence the total number of children participating was 447, with a mean age of 11:1 (years:months; range 4:2 to 19:8), and a sex ratio of 5:1. The children’s diagnoses showed varying degrees of autism: 280 were diagnosed as having autism; 141 received a diagnosis of autism spectrum condition; and 26 were diagnosed as having Asperger’s syndrome. These diagnoses were not made by our team but were taken from the school notes, in all cases from reports by a child psychiatrist or paediatrician.

Procedure

Stage 1
The records of all children included in the study were reviewed to identify any children who had previously been assessed for a tic disorder, or had previously received a diagnosis of GTS.

Stage 2
Where possible, the symptoms of GTS were discussed with teaching staff, using an advisory leaflet adapted from the Tourette Syndrome Association, UK. Some teachers were then asked to comment on the habits and movements of children in their classes.

Stage 3
An observer (V.S., a psychologist) carried out observations of all participating children. Children were observed at school, in their classrooms and during their usual school activities. Class sizes ranged from three children to nine, with an average class size of six. Each child was observed for at least 10 min, using a time-interval sampling observational procedure. The presence of motor and vocal tics was recorded. Each child was subsequently classified as having motor tics, motor and vocal tics, or no tics, on observation. Children classified as having motor and vocal tics were entered directly into Stage 6. Children classified as having only motor or vocal tics were entered into Stage 4.

Stage 4
Children classified as having motor or vocal tics (but not motor and vocal tics) at Stage 3 were re-observed at their school by the observer, for a further 10 mins each, a few days later. Children for whom both motor and vocal tics had been observed, when all observations (Stages 3, 4 and 5) were pooled, were also entered into Stage 6.

Stage 5
Some children (N = 55) were later observed by an independent observer (H.H., also a psychologist) for 10 mins each. Of these children, 23 had been classified by the first observer as showing tics, and 32 had been classified as showing no tics. The names given to this observer were presented in a random order, with no indication of their initial tic classification.

Stage 6
The parents of the children classified as showing both motor and vocal tics on observation were invited to meet a psychiatrist (M.M.R. or J.I.) to discuss their child’s possible diagnosis of GTS, and to obtain a family history. Children and parents were interviewed using a short version of the National Hospital Interview Schedule (NHIS) (Robertson & Eapen, 1996). The Yale Global Tic Severity Scale (YGTSS) (Leckman et al. 1989) was also used. The diagnostic interviews were conducted at the children’s schools, or at University College London Middlesex Hospital (N = 1). Interviews were conducted at the child’s school as far as was possible, to reduce anxiety. Each child was also observed for 30–45 min, as part of the interview. DSM-III-R (APA, 1987) criteria for GTS were used in preference to DSM-IV (APA, 1994) criteria, as DSM-IV requires the additional criterion that tic symptoms must cause marked distress or significant impairment. The adoption of this subjective criterion is inappropriate for many research purposes (Freeman et al. 1995; Erenberg & Fahn, 1996; Kurlan et al. 1997). A particular issue for this study is that impairment and distress may be difficult to establish in children with limited communication and in some children with learning disability.

If the parents were unable to attend the interview, but wished their child to be included in this stage of the study, the child was accompanied by his or her keyworker from the school, and a phone interview with the parents was conducted later by a psychiatrist. It should be noted that in a study of this kind there is a risk that stereotypies will be confused as tics.
The distinction between these is hard to draw, but if either of the raters or clinicians thought a behaviour could be a stereotypy, this was discounted as a tic, in order to err on the side of being conservative.

RESULTS

Stage 1 – review of notes
No evidence for previous assessments or diagnoses of GTS or alternative tic disorders was found in the records of any of the participating children.

Stage 2 – teacher discussion
Our previous study (Baron-Cohen et al. 1999) showed teachers’ reports to be fairly reliable. Therefore, the children identified by their teachers as showing tic-like behaviours were noted and this data was used to help classify children.

Stage 3 – initial observation (V.S.)
Thirty children were identified as having motor and vocal tics by the first observer (V.S.) after Stage 3. These children were entered directly into Stage 6. A further 114 children were identified for whom motor or vocal tics were observed by the first observer. These children were entered into Stage 4.

Stage 4 – re-observation (V.S.)
Of the 114 children who were entered into Stage 4, 18 were unavailable for re-observation by the first observer. Their classification therefore remained the same. From the remaining 96 children, a further 10 were identified for whom both motor and vocal tics were observed by the first observer. These children were also entered into Stage 6.

Stage 5 – re-observation (H.H)
Fifty-five children were seen by the second observer (H.H.), 1 to 2 weeks after the initial observations. All observations, from Stages 3, 4 and 5, were pooled (for example, if one observer saw motor tics and the other vocal tics, that child was classified as showing motor and vocal tics). Agreement on tics v. no tics between the two observers was 70-4%. The two observers agreed that 12 out of the 55 children were showing tic-like behaviours, 26 showed no tic-like behaviours. They disagreed on the classification of 16 children. Both observers were trained in the identification of tics by MMR at the Tourette Clinic, at the National Hospital for Neurology and Neurosurgery, Queen Square, London. The implications of this result are discussed below. When the observations of both observers were combined, the final results were as follows: out of the 447 children entered into the study, 43 children were identified as showing both motor and vocal tics, 98 were identified as showing motor tics only, and 11 were identified as showing vocal tics only.

Stage 6
Forty-three children entered Stage 6 of the study. The parents of 10 children declined to be interviewed. One child was unavailable at the time of the appointment. For the remaining children, at least one parent was interviewed. Twenty-three children were accompanied by their parents to interview, nine were accompanied by their keyworker or teacher, and the parents later interviewed by phone. The diagnosis of definite GTS was confirmed after family interview for 19 of the 32 children. A further 10 children were diagnosed as having probable GTS. If the child showed symptoms at interview and there was a personal history of symptoms, a diagnosis of definite GTS was made. If one of these two criteria was met, but not the other, a diagnosis of probable GTS was made. For the remaining children, two were diagnosed with chronic motor tic disorder, and one was diagnosed as having Rett syndrome. This is a pervasive developmental disorder, characterized by the development of autism, dementia, apraxia of gait and stereotyped use of the hands, following a period of at least 5 months of normal functioning after birth (Hagberg et al. 1983). For definite cases, the rate of true positives was 59-38%, although if we include probable GTS cases, the true positive rate rises to 90-63%. The rate of false positives was 9-38%. The method employed in this study does not allow the calculation of true and false negatives.

Medication information was provided by the parents, and six of the 32 children were taking psychotropic medication (four definite, two probable GTS), three were taking anti-
**Table 1. Diagnosis, severity of tics, and family history**

<table>
<thead>
<tr>
<th>Child</th>
<th>Diagnosis 1</th>
<th>Fam. history</th>
<th>Yale score (%)</th>
<th>Diagnosis 2</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Autism</td>
<td>M</td>
<td>6</td>
<td>Mild GTS</td>
</tr>
<tr>
<td>2</td>
<td>Autism</td>
<td>x</td>
<td>15</td>
<td>Probable GTS</td>
</tr>
<tr>
<td>3</td>
<td>Autism</td>
<td>P</td>
<td>15</td>
<td>Mild GTS</td>
</tr>
<tr>
<td>4</td>
<td>Spectrum</td>
<td>x</td>
<td>17</td>
<td>Probable GTS*</td>
</tr>
<tr>
<td>5</td>
<td>Autism</td>
<td>M</td>
<td>18</td>
<td>Definite CMT</td>
</tr>
<tr>
<td>6</td>
<td>Autism</td>
<td>P</td>
<td>48</td>
<td>Mild GTS</td>
</tr>
<tr>
<td>7</td>
<td>Spectrum</td>
<td>M</td>
<td>17</td>
<td>Definite GTS</td>
</tr>
<tr>
<td>8</td>
<td>Autism</td>
<td>x</td>
<td>40</td>
<td>Probable GTS</td>
</tr>
<tr>
<td>9</td>
<td>Spectrum</td>
<td>x</td>
<td>26</td>
<td>Probable GTS</td>
</tr>
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<td>10</td>
<td>Autism</td>
<td>P</td>
<td>41</td>
<td>Severe GTS</td>
</tr>
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<td>Autism</td>
<td>M</td>
<td>54</td>
<td>Mild GTS</td>
</tr>
<tr>
<td>12</td>
<td>Autism</td>
<td>x</td>
<td>40</td>
<td>Severe GTS</td>
</tr>
<tr>
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<td>Autism</td>
<td>M</td>
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<td>62</td>
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<tr>
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<td>x</td>
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</tr>
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<td>35</td>
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<tr>
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<td>PM</td>
<td>7</td>
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<td>P</td>
<td>30</td>
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<td>M</td>
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<tr>
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<td>P</td>
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<td>Mild GTS</td>
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<td>M</td>
<td>37</td>
<td>Mild GTS</td>
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<tr>
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</tr>
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<td>P</td>
<td>32</td>
<td>Probable GTS</td>
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<td>15</td>
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<tr>
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<td>Autism</td>
<td>PM</td>
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<td>Probable GTS</td>
</tr>
<tr>
<td>32</td>
<td>AS</td>
<td>M</td>
<td>18</td>
<td>Mild GTS</td>
</tr>
</tbody>
</table>

*Child 4, probable GTS, definite chronic motor tic disorder (CMT); Child 18, probable CMT, definite Rett’s syndrome; Child 22, probable GTS, definite chronic vocal tic disorder.

Family history: Tourette syndrome/tics/obsessive–compulsive behaviours; M, history on maternal side of family; P, history on paternal side of family; x, no family history.

AS, Asperger’s syndrome.

Convulsants (two probable GTS, one Rett syndrome plus probable chronic motor tic disorder) and one was taking ritalin (definite GTS). The onset of tics in this child predated the commencement of medication.

YGTSS scores were calculated for all 32 children. The scores ranged from 4% to 63%, with a mean of 28% (S.D. = 16.7).

For the children found to have co-morbid GTS and autism, 20 had been diagnosed with autism (7.1% of children with autism had co-morbid GTS), seven had been diagnosed with an autism spectrum condition (5.0% of children with an autism spectrum condition had co-morbid GTS) and two had been diagnosed with Asperger’s syndrome (7.7% of children with Asperger syndrome had co-morbid GTS).

When the frequencies of co-morbid GTS in children with autism, autism spectrum conditions and Asperger’s syndrome were compared, there were no significant differences ($\chi^2 = 0.43$, df = 2, NS).

Family histories were collected for all 32 children. For 25 of the 32 children (78%), there was found to be a paternal and/or maternal family history of tics and/or obsessive–compulsive behaviours. These results are shown in Table 1.

**DISCUSSION**

The observed rate of 6.5% of GTS (including ‘probable’ GTS cases) in this special school population of children with autism far exceeds that expected by chance. This study, with its prospective, multi-stage design, using combined observational and family interview/history methods, and large sample size, is likely to have
yielded the most accurate estimate yet of the prevalence of GTS in children with autism. Previous studies have retrospectively assessed the rate of GTS in clinical series of individuals with autism, and the earlier study carried out by this team (Baron-Cohen et al. 1999) had a much smaller sample size, although it used a similar method. The rates found in these two studies are also similar ($\chi^2 = 0.43$, df = 1, NS). Thus, the current study supports the findings of the smaller study, and the methods used.

It was suggested (Burd et al. 1987) that concomitant GTS in children with autistic-type conditions provides a marker for improved developmental outcome. The present study was unable to collect data on IQ, and measures of receptive and expressive language, and so cannot be directly compared with Burd et al.’s study. However, the fact that GTS was found to be equally common in children with autism, children with autistic spectrum conditions and children with Asperger’s syndrome shows that GTS is not related to the severity of autism in the child.

It is also notable that a significant proportion of children not identified as having GTS did, however, show motor or vocal tics (but not both motor and vocal tics) on observation. One hundred and nine children (24.4%) were showing tics on observation, but did not show full GTS symptoms. Children with GTS show a fluctuating course of tic expression, and so it is plausible that some of those children showing only motor or vocal tics, if observed for a longer period, would have shown both motor and vocal tics, and so would have entered the final stage of the study. Indeed, this is demonstrated by the 10 children who were identified as having both motor and vocal tics only after re-observation at Stage 4. This is also evident in the four children who were identified as having only motor or vocal tics, or even no tics in one case, by one observer, but as having both motor and vocal tics by the other, either a week later or a week earlier, and the one child who was observed as having motor tics only by one observer, and vocal tics only by the other. This may point to the need for a longer period of observation, to increase the chances of identifying those children with GTS. The observed rate of 6.5% of GTS in children with autism may, therefore, be an underestimate.

The high rate of tics observed in these children (340% of children were classified as showing tics) is interesting, as no children had previously been diagnosed as having a tic disorder. This may indicate that children with autism also show a higher prevalence of alternative tic disorder, chronic motor tic disorder and chronic vocal tic disorder, but that this is being overlooked, possibly since they are occurring in the context of the other problems associated with autism. The possibility of an increased rate of other tic disorders in autism has relevance for genetic studies.

Is it also possible that the high observed rate of tics in children with autism reflects the difficulty in distinguishing tics from other abnormal movements and noises in this population? This differential diagnosis problem has been previously documented (Burd et al. 1987). In particular, complex motor tics can be difficult to distinguish from stereotypes in the absence of self-reported subjective experiential information, particularly regarding the volitional nature of behaviours. Differentiating vocal tics from the wide range of vocal productions in children with autism may be even more problematical. Apart from the clear-cut tics and stereotypes, there are quite a number of behaviours that can only be understood through careful enquiry about the nature of the movement or noise, its longitudinal course, and possible alternative explanation for the symptom. For example, a vocalization, which was initially noted as a sniffing tic, was later discovered, after questioning, to be a repetitive imitation of a TV character with which the child was currently obsessed. However, as mentioned in the Burd et al. study, tics, in contrast to stereotypes, are typically short-lived, contextually inappropriate and interrupt the flow of behaviour or speech. It was also attempted to systematize the difference between motor tics and stereotypes by their topography (tics tend to be clearer in the face, neck shoulders and arms, compared with hands and fingers), their nature (spasmodic versus

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1 This prevalence includes those children: diagnosed with probable or definite, GTS (29) and CMT (3); with parents who declined to be interviewed in Stage 6 (10) and one child unavailable for diagnosis (these 11 children were classified as either motor or vocal, as their tics had not been verified by a psychiatrist); who were showing motor or vocal tics on observation by V.S. and/or H.M. (109). Therefore, the total number of children showing tics was 152.
rhythmic) and also by the quality of the subjective experience and their response to psychosocial factors (Kano et al. 1988). These factors were taken into account when the observations were made, and we are therefore confident that the result obtained represents true co-morbidity.

It is well-known that the tics encountered in GTS wax and wane in severity and fluctuate with time. Stress or anxiety may increase tics, whereas concentrating on a task may reduce tics. Thus, if the raters observed the children with autism at different times, for up to 10 min, and on different days (sometimes more than a week apart), it is quite conceivable that different tics occurred at different times. The levels of stress and anxiety may also have been different, and the children may also have been observed at different times during different tasks (e.g. concentrating). The raters were trained at the Tourette clinic at the National Hospital Queen Square during both new-patient and follow-up clinics. Inter-rater reliability was assessed on the tic observation section of the NHIS. Training was carried out for approximately 6 weeks, seeing about 40 GTS patients (V.S.), while the other psychologist (H.H.) sat in on the clinic and rated many more GTS patients over a year's duration. Agreement between the two raters was only modest, and this may well have been due to factors described above.

A study by Eapen et al. (1997b) found that children in a special school population had an increased prevalence of GTS. Fifty-five per cent of emotionally/behaviourally disturbed children (EBD) and 20% of children classed as learning disabled (LD) were diagnosed as having GTS. Whereas the EBD result could indicate that children with EBD have such problems due to the disruptive effects that GTS can have on the individual, the LD result could point to a larger picture – that children with learning difficulties are more prone to GTS. IQ data were not available for the children in the current study, thus we cannot say whether the results we obtained, of 6.5% GTS in children with autism, can be attributed to their more general learning difficulties. Further research in this area is necessary to better clarify the links between these conditions.

The generally accepted prevalence figure for GTS is around 0.5 per 1000 (Bruun, 1984); this figure has also been reached in a careful epidemiological study (Apter et al. 1993). A more recent pilot study by Banerjee et al. (1998) in the UK yielded higher results and found the rate of GTS in a mainstream school population to be around 3%. This study, however, had a small sample size (N = 166), and the identified cases were not re-assessed and formally diagnosed by an expert. It has been criticized (Traverse, 1998) and stirred debate (Banerjee et al. 1998). The actual prevalence rate of GTS has, therefore, yet to be determined, but the currently accepted figure remains around 0.5 per 1000. We await the results of our larger definitive study in a UK school population.

The YGTSS scores in our GTS individuals ranged from 4% to 63% with a mean of 29% (see Table 1). The majority of the scores indicate mild to moderate severity. The scale range is 0–100%. In the only UK GTS clinic study published using the scale (a modified version with the total range being 0–55) the GTS cases scored a mean of 26.2 (range 11–55) (Robertson et al. 1997). This would be between 45% and 55% using the currently used version of the scale. In another study (Robertson et al. 1999), on 280 consecutive GTS clinic cases, the YGTSS scores ranged from 1 to 100% (mean 49%; s.d. = 23). Both these studies indicate that the GTS/autism individuals are not nearly as severe as clinic patients. In the present study, some of the YGTSS global tic severity scores may have been higher than expected in this population due to the presence of, for example, echophenomena, which occur in both GTS and autism and which symptom receives a separate score on the YGTSS.

The short version of the NHIS (Robertson & Eapen, 1996) was used in this study. This goes into detail with regard to individual tics, and the interviewer records whether specific tics have taken place in the past (ever), in the past week (which allows the YGTSS to be completed), as well as those observed at interview. Tics include simple ones such as frowning, raising eyebrows, blinking, winking, eye movements, nasal twitches, mouth twitching, pouting and opening, tongue protrusion, facial grimacing, platysma tightening, head nodding and shaking, shoulder shrugging and flicking the hair out of the eyes; simple vocalizations include grunting throat clearing, sniffing, snorting, grunting and
coughing. These were the majority of tics noted in the study. Complex tics (motor and vocal) and stereotypies may, however, be difficult to differentiate from each other, and include, for example, hand-flapping (common in people with autism; rare in people with GTS, and not regarded as a tic in this study), twirling (also not encountered in this study), vocalizations, inappropriate fluctuations in pitch of the voice, and squawling. Of course, some symptoms such as echolalia and echopraxia, are common in both autism and GTS, and may be indistinguishable from each other phenomenologically in each condition. It is acknowledged that the relatively higher scores in the complexity score of the YGTSS may well have been partly due to the presence of symptoms such as ecophenomena; one author (M. M. R.) rated the pupils on the YGTSS and included only what she considered to be tics, apart from the ecophenomena. It must be noted that 18 pupils had echolalia, while 12 had echopraxia (10 of these had echolalia as well). We acknowledge the difficulties in differentiating between the two.

Our new screening method suggests that a larger scale cognitive study of the effects of co-morbidity would now be possible. These effects were investigated in the Baron-Cohen and Robertson (1995) case studies. A child with autism, a child with GTS and a child with both conditions were tested for theory of mind, intention-editing and executive function deficits. The predictions, that the child with autism would show a deficit in theory of mind tests, that the child with GTS would show a deficit in intention-editing tests, and that the child with both conditions would show deficits in both these areas, were supported. A larger scale study, with similar methods, may help to confirm these results, which have the natural limitations of single case studies.

One of the purposes of this study was to find children who were being overlooked for treatment. In co-morbid cases of autism and GTS, the latter is often overlooked (as in 100% of our cases) and the symptoms are, therefore, left untreated. Finding these co-morbid cases allows the children’s pharmacological management to be reviewed, and alleviation of tics can help to improve their quality of life. Three such cases were found in this study. By themselves, these cases provide a clinical justification for such screening. For the other children, diagnosed with mild GTS, and not needing medication, knowledge of their condition can help parents and teachers in their continuing support for the child.

The observed elevated rate of GTS in children with autism is consistent with the operation of common aetiological factors, and does not support a chance co-occurrence. Possible common aetiological factors include neurochemical and frontal lobe abnormalities. That there is a substantial family history of GTS or GTS spectrum disorders suggests that there may also be independent genetic mechanisms at play. Future work addressing these possibilities will be important in furthering our understanding of the respective pathogeneses of these two neurodevelopmental disorders.

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REFERENCES


