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ORIGINAL COMMUNICATION

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Gilles de la Tourette's syndrome in special education schools: a United Kingdom study

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Abstract In order to determine the prevalence of tic disorders in children with severe school problems requiring a residential facility and comparison groups of children in regular day schools, we performed direct clinical examinations for the presence of tics and Gilles de la Tourette's syndrome (GTS) in 20 children from a residential school for emotional and behavioral difficulties (EBD); 25 children from a residential school for learning disabilities; 17 "problem" children (PC) (identified by teachers as having academic or behaviour problems) and 19 normal children (NC) selected at random (using random numbers) from a regular school. Of the EBD students, 65% were judged to have definite tics as compared with 24% of students with learning difficulties ($P < 0.05$), 6% of PC ($P < 0.003$) and none of the NC ($P < 0.0006$) group. Most of the affected students met diagnostic criteria for GTS. Our findings suggest that GTS is commonly associated with the need for special education and that this association is particularly robust for children with severe school problems. In these children, the presence of tics may be an indicator of an underlying dysfunction of neurological development.

Introduction

Tics are involuntary, sudden, recurrent non-rhythmic stereotyped movements (motor tics) or vocalizations (vocal tics) (American Psychiatric Association 1987). A variety of primary tic disorders have been described that are based on the duration and pattern of tics (American Psychiatric Association 1987; Tourette Syndrome Classification Study Group 1993). Transient tic disorder refers to single or multiple motor or vocal tics with a duration of less than 1 year. When tics (motor or vocal but not both) have been present for more than a year, chronic tic disorder is the diagnosis. Individuals are diagnosed as having Gilles de la Tourette syndrome (GTS) if multiple motor and one or more vocal tics have been present for at least 1 year.

Motor and vocal tics have been commonly observed in children, and reported in around 10% of children between the ages of 6 to 12 years (Fallon and Schwab-Stone 1992). However, the generally accepted prevalence of the GTS is much lower at around 0.5/1000 (Brunn 1984). It is unclear what proportion of children observed to have tics actually fulfill diagnostic criteria for GTS or one of the other primary tic disorders. It should also be noted that published prevalence figures for GTS are likely to be underestimates, as they are based on clinic or other targeted populations that focus on severe cases and overlook the mild cases who do not come to medical attention (Kurlan et al. 1987; Robertson and Gourdie 1990). Further contributing to underestimation is the reliance on historical information or data obtained from questionnaires rather than direct interviews.

In addition to tics, GTS is often associated with behavioral disturbances such as coprophenomena (obscene words or gestures), echophenomena (copying behaviors), self-injurious behaviors (SIB), obsessive compulsive behaviors (OCB) and attention deficit hyperactivity disorder (ADHD) (Kurlan 1989; Robertson 1989). Taken together, it is well known that this spectrum of clinical symptoms commonly contributes to school problems in children with GTS (Kurlan et al. 1991). Thus, Erenberg et al. found that up to 36% of 200 child and adolescent cases of GTS experienced learning problems, including learning disability (22%), needing to repeat a grade (12%), poor grades (18%) and requiring full-time special education classes (12%) (Erenberg et al. 1986). Since GTS has generally been considered to be rare, its possible contribution to school problems in the general childhood population has received little consideration. However, Comings et al. studied students referred for psychoeducational assessment in a single California school district and reported that 12% of all children in special education had definite GTS and 28% had a broader tic disorder diagnosis (Comings et al. 1990). In a community-based pilot study using direct clinical examinations and involving 70 children attending public schools in one district, Kurlan and colleagues found that of the special education sample, 26% had definite or probable tics as compared with 6% of regular classroom students (Kurlan et al. 1994).

The present study attempts to confirm and extend the results of Kurlan's pilot study by focusing on children with severe school problems, requiring placement in a residential facility. We investigated two different residential schools, one for children classified as emotionally/behaviorally disturbed (EBD) and one for children classified as learning disabled (LD). We hypothesized that any association between tics and special education should be particularly evident for such severely affected children. To enhance the validity of our findings, in comparison with Kurlan's pilot study which used a trained neuropsychological technician, each student in this study was examined by a rater (VE, MMR, RK) who was expert in the clinical diagnosis of GTS.

Subjects and methods

We interviewed and examined all children attending one residential school for EBD and one residential school for LD in a suburban district (West Essex) outside the metropolitan area to the east of London, England. Clinical diagnoses made by other physicians prior to the study and medications prescribed for children attending these residential schools are summarized in Table 1. No children were receiving psychostimulants or other medications known to induce tics (the subject receiving haloperidol was prescribed this medication to treat GTS). In addition, from one non-residential comprehensive primary school in the same district, we asked teachers to select problem children (PC) who were having academic or behavior difficulties (but were still in regular classroom programs), and normal children (NC) were selected randomly using random numbers. Since both the EBD and LD children were attending primary schools, we chose a comprehensive primary school for comparison and selected students at random from the 1st and 4th grades. However, there was considerable discrepancy in the ages of children thus selected, with the age of children from the comprehensive school ranging from 5 to 10 years, and that in the special education schools ranging from 8 to 16, as the majority had repeated grades several times. All the children in the EBD school were boys but since those in the LD school had a male-to-female sex ratio of around 2 : 1, the same sex ratio was chosen for the PC and NC groups as well. The population of West Essex is about 250,000 with around 49,000 being children under the age of 16 years. There was only one residential school for EBD and one for LD children in the district, and hence these were selected for the study while the regular school was chosen at random from the 26 in the same area. Children in the residential schools were referred there when they were unable to tolerate standard day schools. They live in residential facilities Mondays through Fridays, going home to parents on weekends.

Table 1 Clinical diagnoses and medications for children in residential schools

	EBD	LD
Clinical diagnosis		
Mixed disorder of conduct and emotion	17	0
ADHD	2	3
Gilles de la Tourette's syndrome	1	0
Mental retardation	0	25
Obsessive-compulsive disorder	0	2
Medications		
Fluoxetine	0	2
Amitriptyline	0	1
Haloperidol	1	0

Direct clinical examinations were performed for the presence of tics and associated behaviors in 20 EBD, 25 LD, 17 PC and 19 NC. Informed consent was obtained from the parents. All students were personally examined using The National Hospital Interview Schedule for the assessment of GTS and related behaviors, widely used in previous clinical and family studies of GTS (Robertson and Gourdie 1990; Robertson et al. 1988; Eapen et al. 1993), and the reliability and validity of which have been established (Robertson and Eapen 1996).

Children were observed during the interview for 20 min and during mealtime when they were unaware that they were being observed. The examiners (VE, MMR, RK) are all well acquainted with the schedule and its administration in the assessment of tics and GTS. Research diagnostic criteria for classifying observed abnormal movements and noises as tics and for diagnosing tic disorders were employed (Tourette Syndrome Classification Study Group 1993). The occurrence of ADHD was ascertained using DSM-III-R criteria, further corroborated by the teachers. In addition, all children completed the Leyton Obsessional Inventory – child version (LOI) (Berg et al. 1986), and scoring of this instrument was based on "yes" responses. Information was obtained from teachers, but not from parents, nor were medical records reviewed.

For pairwise comparisons between the subject groups regarding the occurrence of tic disorders, 2-sided Fisher's exact tests were employed, using data in the two groups of interest (e.g. EBD vs LD). The Bonferroni correction was used to adjust for multiple comparisons.

Table 2 Summary of clinical findings for the four school populations studied

	Emotionally/ behaviorally disturbed	Learning disabled	Regular school problem	Regular school random
Number of subjects	20	25	17	19
Gender (male/female)	20/0	18/7	11/6	12/7
Mean age (range, years)	9.7 (8–11)	13.4 (10–16)	7.9 (5–10)	7.5 (5–10)
Clinical features tic disorders				
i. Gilles de la Tourette's syndrome (GTS)				
a. Definite GTS	11 (55%)	5 (20%)	1 (6%)	0
b. Possible GTS	2 (10%)	6 (24%)	2 (12%)	0
ii. Chronic motor tic disorder	1 (5%)	0	0	0
iii. Transient tic disorder	0	0	0	1 (5%)
iv. Definite tic disorder, diagnosis deferred	1 (5%)	1 (4%)	0	1 (5%)
Associated behaviors				
Coprophenomena	3	0	0	0
Echophenomena	6	2	0	0
Self-injurious behavior	3	0	0	0
Obsessive-compulsive behavior (Leyton score ≥ 10)	3	7	0	1
Attention deficit, hyperactivity disorder	11	12	4	5

Results

We examined a total of 81 students. Demographic data and results are summarized in Table 2. For the EBD group, 13 (65%) subjects were found to have a definite tic disorder, including GTS (n = 11.55%), chronic motor tic disorder, (n = 1.5%), and definite tic disorder, diagnosis deferred (n = 1, 5%). For the LD group, 6 (24%) subjects were found to have a definite tic disorder, including GTS (n = 5, 20%) and definite tic disorder, diagnosis deferred (n = 1.4%). Diagnosis was deferred when tics were observed on examination, but no reliable history could be obtained. An additional 6 (24%) subjects from the LD group, exhibited tic-like behaviour, where the movements observed by the examiners were slower and lower in amplitude and intensity than typical tics and were therefore not included. Of the PC group, one student (6%) had definite GTS while none from the NC group received a diagnosis of tic disorder. The proportion of subjects with a definite tic disorder was significantly greater in the EBD group when compared with the LD ($P < 0.003$) and NC ($P < 0.0006$) groups. No other comparisons between groups attained statistical significance.

Fifty-five percent of the EBD students and 48% of the LD had evidence of ADHD, compared with 24% of PC and 12% of NC from the regular school. While OCB (Leyton score > 10) was noted in 15% of EBD, and 28% of LD, only one subject from the NC group and none from the PC group exhibited similar behaviors. From the EBD group, of the 13 children with a definite tic disorder, 3 (23%) had OCB and 8 (62%) had ADHD. Of the 6 subjects from the LD group with a definite tic disorder diagnosis, 3 (50%) had OCB and 5 (83%) had ADHD.

The associated behaviors of coprophenomena, echophenomena and SIB were observed more commonly in the EBD group (3,6,3 subjects, respectively) than the LD group (0,2,0 subjects, respectively).

Discussion

The results of this study indicate that tic disorders occur commonly in children requiring special education, the rates being high in EBD (55%) and LD (24%) when compared with "problem" (6%) and random (0%) children in regular school. These rates in the special education groups are higher than previously reported figures for childhood tics, and this is true even when the rate is corrected for the sex ratio in the present sample. Our results, however, must be interpreted in light of the possible limitations of the study. It is difficult to draw firm conclusions from the small sample size, and this study must be considered preliminary in nature. Examiners were not blinded to the educational placement of the subjects, which could have biased the diagnosis of tics. Given the scarcity of resources available for special education and considering the fact that there was only one school (for EBD and LD) of its kind (offering residential placement) in the whole district, students referred to these schools invariably represent the most severe end of the behavioral spectrum. The higher rates observed may be, at least in part, a reflection of this referral bias. However, our methodology, which utilized a randomly selected or complete sample of the population

of interest and which included face-to-face evaluation, along with the use of questionnaires and corroborative history from the teachers, increases the validity of our findings.

Our study is further limited in that the groups were not ideally matched for age and sex, and it may be argued that since tic disorders are more common in males, the higher prevalence of tics observed in the EBD group is at least partially accounted for by the fact that all children were boys. In addition, our diagnoses of tic disorders were based on current symptoms only, did not include subjects with a prior history of tics, and were not based on historical information from parents. Thus, we may have overlooked some cases with a tic disorder.

Despite these limitations, our findings are in agreement with those of Kurlan and colleagues who found a significantly higher prevalence of tics (26%) in children requiring special educational services in a regular day school setting when compared with children in a normal classroom program (6%) (Kurlan et al. 1994). The current study extends these observations and indicates that the relationship between tics and special education is even more robust for severely impaired children. The rate of tic disorders observed by Comings et al. (28%) for children in special education also agrees with our findings (Comings et al. 1990). Our findings that the rate of observed definite tics was higher in EBD (65%) than LD (24%) and that behaviors characteristically associated with GTS, including coprophenomena, echophenomena and self-injurious behavior, are seen more commonly in the EBD group suggest the possibility that GTS is linked to specific patterns of behavioral disturbance and perhaps selective abnormalities in brain function and development. Further study in this area is needed before definite conclusions can be reached.

It is interesting to note that of the 20 subjects in our sample identified as having a definite tic disorder, only one child had been previously diagnosed as having GTS. It therefore seems that although tic disorders are common in this population, they are often overlooked, possibly since they are occurring in the context of other difficluties such as behavioral and emotional problems or learning disability. However, family genetic studies that have largely included subjects with mild, uncomplicated cases of tics also indicate that many cases of tic disorder diagnosed by examiners are unrecognized or overlooked by the subjects themselves and their relatives (Kurlan et al. 1987; Robertson and Gourdie 1990). Furthermore, cases referred for medical evaluation are often misdiagnosed (Robertson and Gourdie 1990). The experience reported in both school and family studies thus indicates that accurate determination of prevalence rates for tic disorders in a population of interest cannot rely on patient self-report questionnaires or the diagnoses of health professional, but rather requires a direct interview by experienced or trained observers. This may explain why a recent study of the Israeli defense force, which relied on subject self-reports, yielded a much lower prevalence estimate for GTS in adolescents of 4.3 per 10,000 (Apter et al. 1993).

It remains unclear whether the tics observed in our childhood populations represent a disorder that is identical with or etiologically related to GTS. On the one hand, it may be that all cases of childhood tics do in fact represent GTS and are related to the same genetic mechanism (Kurlan 1994). On the other hand, it is possible that some of the subjects do not have a disorder that is related to GTS. This is in keeping with the finding from a segregation analysis study of families affected by tics which indicated that motor tics (chronic or transient) may not always be genetically related to GTS and are perhaps in these circumstances, phenocopies (Eapen et al. 1993). Clarification of the true relationship between childhood tics and the disorder GTS awaits the availability of reliable neurobiological or genetic markers for the condition.

About 1/6 to 1/8 of the students from both special education schools with tics had evidence of ADHD, while OCB was present in about a quarter to half of cases with tics, representing rates similar to those observed in GTS patients (Frankel et al. 1986; Robertson et al. 1988). Both OCB and ADHD can lead to difficulties in classroom performance, and it is not surprising that these children were placed in special education schools. This representation of multiple problems in our subjects may be related to the increased likelihood of such children being in need of special facilities.

A comparison of our findings in the two groups of special education students supports the notion that GTS is linked to a specific pattern of behavioral disturbance. We found a distinct difference between the EBD and LD groups in the type, frequency, and quality of tics observed. The rate of observed definite tics was higher in EBD (65%) than LD (24%). In addition, in 6 subjects from the LD group, we observed tic-like behavior that was atypical, lacking the usual “GTS quality” in that these were slow and having lower amplitude. Furthermore, based on the evidence from direct interview with the child and the information from the teachers, it seemed that they lacked other GTS features such as the waxing and waning, brief periods of voluntary suppression, premonitory sensation and a sense of relief after the tic. This is in keeping with the fact that tic-like stereotypes and mannerisms are common in a mentally retarded population. In addition, behaviors characteristically associated with GTS, including coprophenomena, echophenomena, and self-injurious behavior, were observed more commonly in the EBD group. Thus, despite the fact that LD are more likely to have evidence of central neurological dysfunction, symptoms most closely referable to GTS were more commonly associated with EBD. These observations suggest that tics may have a specific association with emotional/behavioral disturbance rather than being a sign of global neurological impairment. Thus, it could be postulated that the

underlying neuronal abnormality in GTS is linked to selective areas in the brain, resulting in specific behavioral manifestations. Indeed, available evidence, especially from neuroimaging studies, suggest that the abnormality in GTS is specific, involving basal ganglia and frontal areas of the brain (Chase et al. 1984; George et al. 1992).

In summary, our results indicate that GTS and related tic disorders occur commonly in children requiring special education. This link appears to be particularly robust in children with severe school difficulties. The presence of tics may be an observable sign of underlying neurological dysfunction that contributes to learning problems and a specific pattern of emotional/behavioral disturbance. The recognition of GTS in children requiring special education may be of practical importance. Since many of the symptoms (i.e., tics, OCB, ADHD) are treatable, institution of appropriate therapy might improve academic performance and behavior in these children. Further study of tics in children with learning, behavior and other developmental disorders is needed to better clarify the interrelationships of these conditions.

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