Tourette syndrome: an analysis of 200 pediatric and adolescent cases[1]

Gerald Erenberg, M.D.
Robert P. Cruse, D.O.
A. David Rothner, M.D.

The clinical profiles of 200 children and adolescents with Tourette syndrome are reviewed. The mean age of onset was 6.3 years, and the mean lag between onset of symptoms and diagnosis was 4.5 years. Thirty-seven percent of these patients had another family member with a tic disorder. A strikingly large number (58%) of these children had learning problems, behavioral problems, or both. These problems often caused more difficulties in everyday living than the tics and noises themselves. It is not sufficient simply to diagnose this complex disorder. These patients and their families require both help and understanding in dealing with the tics and noises as well as the associated behavioral and learning difficulties.

Index term: Gilles de la Tourette's disease

Cleve Clin Q 53:127–131, Summer 1986

Tourette syndrome is a disorder characterized by the childhood onset of multiple motor tics and involuntary vocalizations. Georges Gilles de la Tourette first delineated the disorder in 1885 when he described nine cases,[1] although one of his cases had been described previously by Itard in 1825. Only a few additional cases had been described until the 1960s, when the number of cases diagnosed increased rapidly.[2–4] Much of this increase was because of heightened public awareness, which led patients who recognized the syndrome in themselves or in family members to seek medical help.

We report the clinical profiles of 200 children and adolescents with Tourette syndrome seen at The Cleveland Clinic Foundation over a five-year period. The complexity

[1] Departments of Pediatric and Adolescent Medicine and Neurology, The Cleveland Clinic Foundation. Submitted for publication Aug 1985; accepted Jan 1986. pa

0009-8787/86/02/0127/05/$2.25/0

Table. Characteristics of 200 children with Tourette syndrome

	Number	Percent
Sex		
Male	165	82
Female	35	18
Race		
White	194	97
Black	6	3
Family history of tics	74	37
Severity		
Mild	89	44
Moderate	99	50
Severe	12	6
Neurologic disorders	10	5
Febrile seizures	7	4
Afebrile seizures	3	2
Cerebral palsy	0	0
Mental retardation	6	3
Neuromuscular disorder	0	0
Learning problems	72	36
Learning disability	43	22
Repeated grade	24	12
Poor grades	35	18
Full-time special class	17	8
Part-time special class	24	12
Behavior problems	95	48
Attention deficit disorder	70	35
Adjustment reaction	20	10
Neurosis or psychosis	17	8
Abnormal neurologic examination	5	3

of this disorder has become increasingly evident as we have gained experience with large numbers of patients.

Patient population

Starting in 1977, all patients under 19 years of age with Tourette syndrome were seen by one of us and prospectively added to the Tourette syndrome registry. We made the diagnosis based on the following DSM-III criteria:[5] age at onset between 2 and 15 years; presence of recurrent, involuntary, repetitive, rapid, purposeless motor movements affecting multiple muscle groups; multiple vocal tics; ability to suppress movements voluntarily for minutes to hours; variations of the intensity of the symptoms over weeks or months; duration of symptoms for more than one year. We took an extensive history from each patient and examined them thoroughly. We did not routinely perform laboratory tests for patients thought to have classical Tourette syndrome.

We obtained the family history by interviewing the parents and did not routinely examine other family members. In the absence of a standardized grading system, we judged the severity of the disorder as follows: mild cases were those where the tics were not continuously present or were not usually noticed by other people. We judged the disorder as moderate if the tics were continuously present and led to stress, embarrassment, and teasing. Severe cases had constant tics that were considered bizarre by those who saw them and severely limited the person's ability to lead a normal life.

We identified school problems from the history given by the parents. School reports and results of psychological tests were available for most children who were experiencing learning problems, although no uniform method of measurement was used. Children were also divided into three groups according to behavior problems: The first group was made up of children with attention deficit disorder with or without hyperactivity, based on the criteria listed in the DSM-III.[6] The second group comprised children whose behavioral difficulties were mild and thought to reflect reactive problems in adjusting to the burden of exhibiting involuntary tics and noises. The third group consisted of patients with neurotic or psychotic behaviors. These behaviors always included an element of obsessions and compulsions and were usually more disruptive than the tic components of the disorder. Most patients in the latter group were seen in consultation by a staff psychiatrist.

Results

The *Table* summarizes some of the major findings in the 200 children with Tourette syndrome. The male-to-female ratio was 4.7, which is higher than in most previous studies.[2-4] The small number of nonwhites has been consistently noted in the past.[3] The mean age of onset was 6.3 years, and 90% began having symptoms before age 10. The average lag between onset of symptoms and diagnosis was 4.5 years, and most patients had been seen by several physicians before the correct diagnosis was made. No religious or ethnic group was disproportionately represented; the religious

and ethnic background of the patients was found to be the same as that seen in the geographic areas served. Thirty-seven percent reported another family member with a tic disorder. The percentage of those with a mild form of Tourette syndrome was higher than previously reported.[7]

Only 15 patients (8%) had experienced previous encephalopathic events, such as neonatal asphyxia or severe head trauma, that might have altered brain function. In general, the group enjoyed good health, and most patients had no significant medical illnesses in their background. The medical disorders found in 32 patients (16%) included a wide variety of problems such as asthma, chronic abdominal pain, and recurrent urinary tract infections. The incidence of febrile seizures, afebrile seizures, and mental retardation was no different from that seen in the population as a whole. Except for the presence of tics, the neurological examination was normal in 195 patients (98%). The abnormalities seen in the remaining five patients were minor and included reflex asymmetry and motor incoordination.

Of the 200 patients, 115 (58%) had learning problems, behavior problems, or both. Learning disabilities and attention deficit disorder with or without hyperactivity were the most common findings. Seventeen patients (9%) had neurotic or psychotic behavior that severely limited their everyday functioning. Coprolalia, the involuntary use of obscenities, had occurred in only 15 patients (8%).

Following the original analysis of the entire group, we compared males and females, patients with mild tics and patients with moderate and severe tics, patients with and without a positive family history of tics, patients with and without learning problems, and patients with and without behavior problems. We found few significant differences between these groups except for the relationships between behavior problems, learning problems, and gender. Patients with either behavior problems or a family history of tics had an earlier age of onset of their disorder. As expected, behavior and learning problems were frequently found in the same patients. The greatest difference between groups was the much higher incidence of behavior and learning problems in males than females. This does not differ from the male-to-female ratio usually found in children with behavior and learning problems. On the other hand, the presence or absence of behavior and learning problems did not correlate significantly with the severity of the tics. Coprolalia, however, was more frequent in those with moderate and severe tics.

Discussion

Tics are a common disturbance of childhood and are estimated to occur at one time or another in up to 24% of children.[8] Tic disorders can be placed on a spectrum with transient, simple motor tics of childhood at one end.[9] At the other end of the spectrum is Tourette syndrome, a disorder of chronic, multiple motor tics with vocalizations.

Although the true incidence of Tourette syndrome is unknown, it is clearly not a rare disorder. Recent increases in case reporting have led to speculation that Tourette syndrome may occur in up to 2.6% of the general population.[8] As more persons with Tourette syndrome are identified, the complexity of this disorder is becoming increasingly apparent.[10] In fact, as stated by Cohen et al.,[11] Tourette syndrome appears to be an extremely complex biological and psychological disorder reflecting the interaction among genetic, neurological, behavioral, and environmental factors that change in their expression during development.

The proposed etiology for tic disorders has changed over time. Until recently, tics were believed to be caused by emotional or psychiatric factors, although no specific psychopathology has uniformly been found in persons with tic disorders.[12] Currently, emphasis is being placed on an organic etiology, although the exact mechanism has not been delineated. An organic cause for Tourette syndrome is supported by experience with medications that interact with neurotransmitters[13] as well as by biochemical studies.[14] Tourette syndrome can be precipitated at times by the administration of psychostimulants that increase the release of catecholamines.[15] This finding, along with the beneficial response to haloperidol, a dopaminergic receptor blocker, has led to the hypothesis that excessive dopamine or increased sensitivity to dopamine action is the basis of the disorder. Several investigators have found diminished homovanillic acid in the cerebrospinal fluid of patients with Tourette syndrome.[16,17] Since homovanillic acid is a metabolite of dopamine, a lowered level was thought to indicate that excessive dopaminergic effect is due to supersensitive dopamine receptors and not to an overproduction of dopamine.

In the absence of a biochemical or electrophysiological marker,[18] the diagnosis of Tourette syndrome remains a clinical one. Currently available laboratory tests do not aid diagnosis.[8] Essential clinical features include involuntary motor and vocal tics beginning in childhood or adolescence, and the mean age of onset in our group was 6.3 years. Motor tics are more often the initial manifestation of Tourette syndrome than involuntary vocalizations. Although the onset may be explosive, the problem usually begins as simple, repetitive tics involving the face or head and evolves over time. The list of possible motor tics is almost endless and may include both simple and complicated movements. Organized, ritualistic, compulsive actions are common, including touching or smelling one's own or another person's body. The vocal disturbances may be usual noisemaking done in excess, unusual noises, uttering partial or full words, repeating words or sentences, or stammering.

Although frequently reported in earlier series,[19] only 8% of our patients had ever experienced coprolalia. It is possible that the incidence of coprolalia will increase as our patients become older. In addition, the relatively small number with coprolalia may reflect the higher-than-expected number of patients whose symptoms were milder than previously reported.[7] Initial case reports almost always described persons with severe symptoms. The full spectrum of possible severity, from mild to severe, is only now being appreciated as larger numbers of cases are being diagnosed. Ten patients with Tourette syndrome were seen in 1978, and only one (10%) had the mild form.[20] After intense publicity in the local media, we saw 98 cases in 1981, and 62 (63%) were considered mild. Many patients with mild forms had previously been diagnosed as persons with nervous habits, and the correct diagnosis of Tourette syndrome had not been made.

As in other series, we found more boys to be affected than girls. Although Tourette syndrome has been reported in all races, ethnic groups, and socioeconomic classes,[21] it occurs much more frequently in white persons than in black persons. Initial reports revealed a high percentage of patients to be of Ashkenazi Jewish or of eastern European origin.[22,23] In our cases, however, the percentage of Tourette syndrome patients with Jewish or eastern European background was the same as the general population.

There is increasing evidence that Tourette syndrome and other tic syndromes may have a genetic origin. Originally, anecdotal reports of multiple family members with Tourette syndrome were the only evidence for this.[24,25] More recently, systematic studies of Tourette syndrome families have been reported.[26,27] The 37% of our patients who reported another family member with a tic disorder is undoubtedly an underestimation since we did not evaluate other family members. Although it has been suggested that female patients show a positive family history more often than males,[8] we did not find such a difference in our series. Current studies have not yet clarified the mode of genetic inheritance, and there is no consistent pattern of inheritance.[28]

Review of medical histories did not reveal any unusual patterns of illness in our patients. Patients with Tourette syndrome were not found to have a higher-than-expected incidence of medical or neurological disorders. What is striking, however, is the high incidence of behavior and learning problems in the children with Tourette syndrome. In our series, over 50% of the patients had accompanying difficulties in the form of learning disabilities, attention deficit disorder with or without hyperactivity, and other behavioral disorders. A relationship between behavioral factors and the basic component of Tourette syndrome (tics and noises) has been recognized and discussed almost from the time the syndrome was first described.[29] Until recently, however, attention was focused on how emotional and behavioral problems might be the cause of Tourette syndrome. We now believe that these problems are not the cause of Tourette syndrome but are an inherent part of the disorder for some patients, except for those who experience adjustment reactions because of social embarrassment.

The reason for the behavioral and learning problems is unknown. They occurred as frequently in children with mild tics and noises as they did in those whose involuntary movements were more severe. The incidence was the same in those with and without a family history of tic disorders. There was, however, a much higher incidence of such problems in males with Tourette syndrome than in females. A strong relationship between these two problems existed: the majority of patients with learning problems had behavioral problems and vice versa. When present, the behavioral problems usually preceded the onset of the tics. For many children, the associated behavior and learning problems caused more difficulties in their everyday life than the tics and noises. The number of patients

with significant psychiatric disease may increase as these patients are followed for a longer time. Recent evidence has indicated that more severe symptoms will develop in Tourette syndrome patients with mild obsessive-compulsive symptoms as they become older.[30]

For children with Tourette syndrome, the severity of their tics was often a barometer of their emotional state. Complex relationships were found between the emotions and tic phenomena, even in children whose day-to-day behavior was not out of the ordinary. Feelings, both positive and negative, tremendously influenced the outward manifestations of tic behavior. Periods of increased symptoms often corresponded to times of excitement and increased emotional tension. Examples included worsening of symptoms before returning to school, before school examinations, and in anticipation of birthdays, Christmas, and other special events.

Simply making the diagnosis of Tourette syndrome was a major accomplishment five years ago, and efforts at that time were directed toward recognizing the disorder. Today, many families have already made the correct diagnosis by the time they come to see the physician. What these families want is help, not only for the tics and noises, but also for the learning, behavioral, and social problems that may be present. Such help, however, will require increased understanding and knowledge of the biological and psychological bases of Tourette syndrome.

References

1. Gilles de la Tourette G. Étude sur une affection nerveuse, caracterisée par de l'incoordination motrice, accompagnée de'écholalie et de coprolalie. Arch Neurol (Paris) 1885; **9:**19–42, 158–200.
2. Nee LE, Caine ED, Polinsky RJ, Eldridge R, Ebert MH. Gilles de la Tourette syndrome: clinical and family study of 50 cases. Ann Neurol 1980; **7:**41–49.
3. Golden GS. Movement disorders in children: Tourette syndrome. JDBP 1982; **3:**209–216.
4. Golden GS. Tourette syndrome: the pediatric perspective. Am J Dis Child 1977; **131:**531–534.
5. American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, ed 3. Washington, DC: American Psychiatric Association, 1980, pp 76–77.
6. American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, ed 3. Washington, DC, American Psychiatric Association, 1980, pp 41–45.
7. Shapiro AK, Shapiro ES, Bruun RD, Sweet RD. Gilles de la Tourette Syndrome. New York, Raven Press, 1978, p 130.
8. Shapiro E, Shapiro AK. Tic disorders. JAMA 1981; **245:**1583–1585.
9. Golden GS. Tics and Tourette's: a continuum of symptoms? Ann Neurol 1978; **4:**145–148.
10. O'Quinn AN, Thompson RJ Jr. Tourette's syndrome: an expanded view. Pediatrics 1980; **66:**420–424.
11. Cohen DJ, Detlor J, Shaywitz BA, Leckman JF. Interaction of biological and psychological factors in the natural history of Tourette syndrome: a paradigm for childhood neuropsychiatric disorders. Adv Neruol 1982; **35:**31–40.
12. Shapiro AK, Shapiro E, Wayne H, Clarkin J. The psychopathology of Gilles de la Tourette's syndrome. Am J Psychiatry 1972; **129:**427–434.
13. Feinberg M, Carroll BJ. Effects of dopamine agonists and antagonists in Tourette's disease. Arch Gen Psychiatry 1979; **36:**979–985.
14. Cohen DJ, Shaywitz BA, Young JG, et al. Central biogenic amine metabolism in children with the syndrome of chronic multiple tics of Gilles de la Tourette. J Am Acad Child Psychiatry 1979; **18:**320–341.
15. Golden GS. The effect of central nervous system stimulants on Tourette syndrome. Ann Neurol 1977; **2:**69–70.
16. Singer HS. Tics and Tourette syndrome. Johns Hopkins Med J 1982; **151:**30–35.
17. Singer HS, Butler IJ, Tune LE, Seifert WE Jr, Coyle JT. Dopaminergic dysfunction in Tourette syndrome. Ann Neurol 1982; **12:**361–366.
18. Krumholz A, Singer HS, Niedermeyer E, Burnite R, Harris K. Electrophysiological studies in Tourette's syndrome. Ann Neurol 1983; **14:**638–641.
19. Shapiro AK, Shapiro ES, Bruun RD, Sweet RD. Gilles de la Tourette Syndrome. New York, Raven Press, 1978, pp 146–149.
20. Erenberg G, Rothner AD. Tourette syndrome; a childhood disorder. Cleve Clin Q 1978; **45:**207–212.
21. Shapiro AK, Shapiro ES, Bruun RD, Sweet RD. Gilles de la Tourette Syndrome. New York, Raven Press, 1978, pp 117–118.
22. Eldridge R, Sweet R, Lake CR, Ziegler M, Shapiro AK. Gilles de la Tourette's syndrome: clinical, genetic, psychologic, and biochemical aspects in 21 selected families. Neurology 1977; **27:**115–124.
23. Wassman ER, Eldridge R, Abuzzahab S, Nee L. Gilles de la Tourette syndrome: Clinical and genetic studies in a Midwestern city. Neurology 1978; **28:**304–307.
24. Friel PB. Familial incidence of Gilles de la Tourette's disease, with observations on aetiology and treatment. Br J Psychiatry 1973; **122:**655–658.
25. Guggenheim MA. Familial Tourette syndrome. Ann Neurol 1979; **5:**104.
26. Kidd KK, Prusoff BA, Cohen DJ. Familial pattern of Gilles de la Tourette syndrome. Arch Gen Psychiatry 1980; **37:**1336–1339.
27. Pauls DL, Cohen DJ, Heimbuck R, Detlor J, Kidd K. Familial pattern and transmission of Gilles de la Tourette syndrome and multiple tics. Arch Gen Psychiatry 1981; **38:**1091–1093.
28. Kidd KK, Pauls DL. Genetic hypotheses for Tourette syndrome. Adv Neurol 1982; **35:**243–249.
29. Shapiro AK, Shapiro ES, Bruun RD, Sweet RD. Gilles de la Tourette Syndrome. New York, Raven Press, 1978, pp 11–14.
30. Montgomery MA, Clayton PJ, Friedhoff AJ. Psychiatric illness in Tourette syndrome patients and first-degree relatives. Adv Neurol 1982; **35:**335–339.

Gerald Erenberg, M.D.
The Cleveland Clinic Foundation
9500 Euclid Avenue
Cleveland, OH 44106