The Epidemiology of Tourette's Syndrome: A Pilot Study

by Janine Jagger, Brigitte A. Prusoff, Donald J. Cohen, Kenneth K. Kidd, Claudia M. Carbonari, and Karen John

Abstract

A new, precoded, self-report questionnaire was used to collect information on 75 patient members of the Tourette Syndrome Association. This diverse group of Tourette's syndrome (TS) patients supplied by mail detailed medical, symptom, developmental, family, and personal history information. Analysis of these data supported the usefulness and validity of the questionnaire, and also indicated that the diagnosis of TS is being made earlier; attentional difficulties may be basic to the disorder; haloperidol is an effective treatment for many patients, but alternative treatments are needed because unpleasant side effects of haloperidol often lead to discontinued use, and TS is disabling for many with most patients reporting serious problems in one or more areas of functioning.

In 1885, Gilles de la Tourette vividly described a neuropsychiatric syndrome with a unique constellation of clinical features: multiple motor tics, phonic symptoms, compulsions, and other behavioral phenomena, such as the irresistable urge to echo or imitate. He observed its predominance in males, early onset, and lifelong duration, and speculated that its cause might lie in perhaps inherited neurological vulnerability. Following Tourette's report, with increasing frequency in the past 20 years, hundreds of cases and series of patients have been described in the world literature, leaving little doubt that Tourette's syndrome (TS) is a clinically recognizable disorder with a stable pattern of expression across cultures (Abuzzahab and Anderson 1973; Shapiro et al. 1978). While each patient presents his own set and pattern of symptoms, there is a remarkable similarity in clinical phenomenology and natural history, with a waxing and waning course in which symptoms disappear and are replaced by new ones.

Tourette's syndrome was once thought to be a rare clinical curiosity. However, the discovery of the therapeutic value of haloperidol in the mid-1960s (Shapiro and Shapiro 1968), and the exceptional public information campaigns of the national Tourette Syndrome Association have led to increasing recognition of the disorder and more frequent, earlier diagnosis (Shapiro et al. 1978). During the past decade, the use of new biochemical strategies in the study and treatment of TS and other neuropsychiatric disorders (van Woert et al. 1976; Cohen et al. 1978; Friedhoff and Alpert 1978; Cohen et al. 1979*a*) has generated increasing interest in the possible neurochemical basis and correlates of TS. This new research requires more refined understanding of the clinical characteristics of TS in large, representative populations of patients.

To a remarkable extent, research on TS has been facilitated by the Tourette Syndrome Association (Lake et al. 1977; Wassman et al. 1978), and the present study was undertaken in collaboration with them. Access to this national group (whose registry includes nearly 2,000 purported TS patients) afforded us the opportunity

Reprint requests should be sent to Dr. B.A. Prusoff, Yale University School of Medicine, Department of Psychiatry, 904 Howard Avenue, New Haven, CT 06519.

to study a geographic, racial, and ethnic cross-section of TS patients. In this article, we present the results of a pilot study, which employed our newly developed Tourette's Syndrome Questionnaire (TSQ). The pilot study objectives were to gather systematically a spectrum of information on TS patients and to assess the usefulness and validity of the TSQ. Concurrently, the TSQ was used with a clinically diagnosed population of TS patients which supported its validity; the results of the clinical project have been reported separately (Cohen et al. 1979*b*).

Methods

Questionnaire Design. The precoded, self-report TSQ was designed to elicit a patient's detailed medical history and clinical picture in the context of the individual's social and psychological development and personal experiences. The questions contained in the 35 pages of the TSQ are categorized and arranged under the following subject headings: Patient Personal Data Inventory; General Medical History; History of Tourette Symptomatology; Treatment History; Family History; Prenatal and Birth History; and Developmental History. The patient's mother was requested to complete the birth and prenatal history questions, and parents were asked to assist with the developmental and family history questions. However, the entire questionnaire may have been completed by or with the assistance of the patient's parents. Content of the questions was based on numerous sources including patient interviews, Tourette Syndrome Association member suggestions, literature reports, and contributions from researchers and clinicians in the fields of child psychiatry, psychology, neuropharmacology, epidemiology, and genetics.

Subjects. Ten percent of the Tourette Syndrome Association patient membership was selected for the questionnaire survey by a systematic sampling technique on an alphabetic file. Anonymity of respondents was ensured by a numerical identification system controlled by the Tourette Syndrome Association (TSA). Questionnaires were mailed to the 200 selected patients. A followup letter was sent to nonrespondents. The TSA contacted those who had still not responded by telephone to encourage response, to assess reasons for failure to respond, and to determine characteristics of nonresponders.

Results

Response Rate. After the initial mailing, 61 completed questionnaires were returned. Three more were returned after the followup letter, and an additional 11 after the telephone survey, bringing the total to 75 completed questionnaires (37.5 percent of those sent).

Of the 136 initial nonresponders, only 36 (26 percent) were reached by telephone. Among those, seven stated that they did not have Tourette syndrome; six reported that they had never been formally diagnosed, but believed they had TS. Twenty-one stated that they had been formally diagnosed as having TS, and 11 of those patients subsequently returned questionnaires. Reasons given for nonresponse included: lost or did not receive questionnaire (28 percent); were not interested in the questionnaire (22 percent); felt unable to complete the questionnaire (19 percent); did not have Tourette syndrome (11 percent); believed the questionnaire to be too long (8 percent); and felt the questionnaire to be an invasion of privacy (6 percent). Six percent of those contacted offered no reason for not completing the questionnaire.

Demographic Findings. Fifty-seven of the respondents were male (76 percent), and 18 (24 percent) were female. Ages of the patients ranged from 8 to 57 years of age: 28 percent were 12 years or younger, 37 percent were 13 through 19, and 35 percent were 20 or over. All but two of the patients were white; one black and one oriental patient responded. Of the patients 18 years of age or older, 33 percent were married, 53 percent were single, and 14 percent were separated or divorced. The educational level of patient households was high: Among patients living with parents, 60 percent of the heads of household had at least some college. Thirty-eight percent of the patients were in Hollingshead's (1965) social class III, 16 percent in I, 19 percent in II, 19 percent in IV, and 7 percent in V (mean social class 2.8).

General Medical History. Patients reported having had a variety of common childhood illnesses, but in the absence of a control group no statistical evaluation was possible. No marked frequency of an unusual disease was noted.

Medical Disorders and Tourette Syndrome Onset. Thirty-six percent of the patients indicated that

a specific medical problem immediately preceded and may have been associated with the onset of TS symptoms. However, the problems cited were thinly distributed among such diverse "precipitators" as rheumatic fever, head injury, respiratory disorder, measles, digestive disorder, and use of a drug.

Onset and Diagnosis. The first signs of TS were noticed by family members as early as 2 years and as late as 16 years of age, with a mean of 7 years of age. The average interval between the onset of symptoms and the determination of a TS diagnosis was 7.6 years. Younger patients, those under 21 years of age, reported a considerably shorter period between onset and diagnosis (3.5 years) than did older patients (15.5 years). Based on their TS symptoms, 39 percent of the patients had been assigned one diagosis other than TS, and 45 percent had been given two or more different diagnoses. Only 16 percent received a Tourette's syndrome diagnosis the first time they were diagnosed. The most frequent of the other diagnoses reported were emotional problems, transient tics of childhood, chorea, hyperactivity, "nervousness," and residual effects of a physical illness. Most patients were told they had TS by either a neurologist (60 percent) or a psychiarist (32 percent).

More than half of the patients (51 percent) first suspected that their disorder was TS after hearing about TS from the mass media, while only 40 percent were first alerted by a physician. The most common first symptom noticed by family members or others was reported to be a tic of the eyes or face (57 percent); other much less frequent first signs included tics of the extremities and trunk, and a variety of vocal tics and compulsive actions.

Symptomatology. In order to ascertain the ages at which a number of symptoms first appear, patients were asked to indicate when (or if) each of 23 symptoms listed was first experienced. (See table 1 for the symptoms as they appeared on the questionnaire, and the number and percentage of patients who "ever" experienced each of them.) Space was provided for additional symptoms not included on the list. Patients reported a virtually inexhaustible variety of tics and other symptoms, and because it often was not productive to consider each separately, we will report symptom findings in three broad categories: (1) motor tics, (2) vocal tics, and (3) compulsive actions.

Motor tics of the eyes, face, and head were reported to have occurred earliest (mean 7.2 years). In addition, more patients (95 percent) reported having had these tics than any other symptom. Consistent with the description by Mahler, Luke, and Daltroff (1945) of the progression of symptoms over time, our findings suggest a cephalo-caudal sequence in the development of motor tics with the following mean ages of onset recorded: eyes, face, or head (7.2 years); shoulder or neck (8.7 years); arms or hands (9.1 years); trunk (9.5 years); and legs (10.1 years). While the symptom progression appeared to be continuous, the differences in mean ages were slight and significance levels for these differences have not been determined. (Figure 1 graphically displays the ages of onset of these and other symptoms.) It should be noted that these are means and that not all patients experienced each motor tic in turn, if at all. Additional analyses of within-patient onset data are needed to confirm the symptom progression found across patients.

When viewed as a group of symptoms, motor tics first appear at an earlier mean age (8.7 years) than either vocal tics (10.7 years) or compulsive actions (10.8 years). Of particular interest to patients is the finding that 37 percent of the patients in our sample claimed to have had coprolalia. However, since the mean age of first occurrence was 11.1 years, it is possible that the younger patients in our sample who had not had the symptom will have it in the future. The most frequent vocal tic was the uttering of low noises (84 percent of the patients).

Fewer patients claimed to demonstrate compulsive actions than either motor or vocal tics. The most frequent compulsive action was that of touching objects (55 percent of the patients). Touching of sexual organs of self or others was reported by 36 percent of the patients, with a mean onset age of 11.7. Self-destructive behaviors were serious problems for some patients; 12 percent reported biting themselves and 17 percent reported that they banged their heads.

One-fourth of the patients indicated that they had experienced an internal tic, that is, one they feel but do not express. One-third stated that they presently feel they have tics more on one side of the body than the other; however, neither side was systematically favored.

Seventy-one percent of the pa-

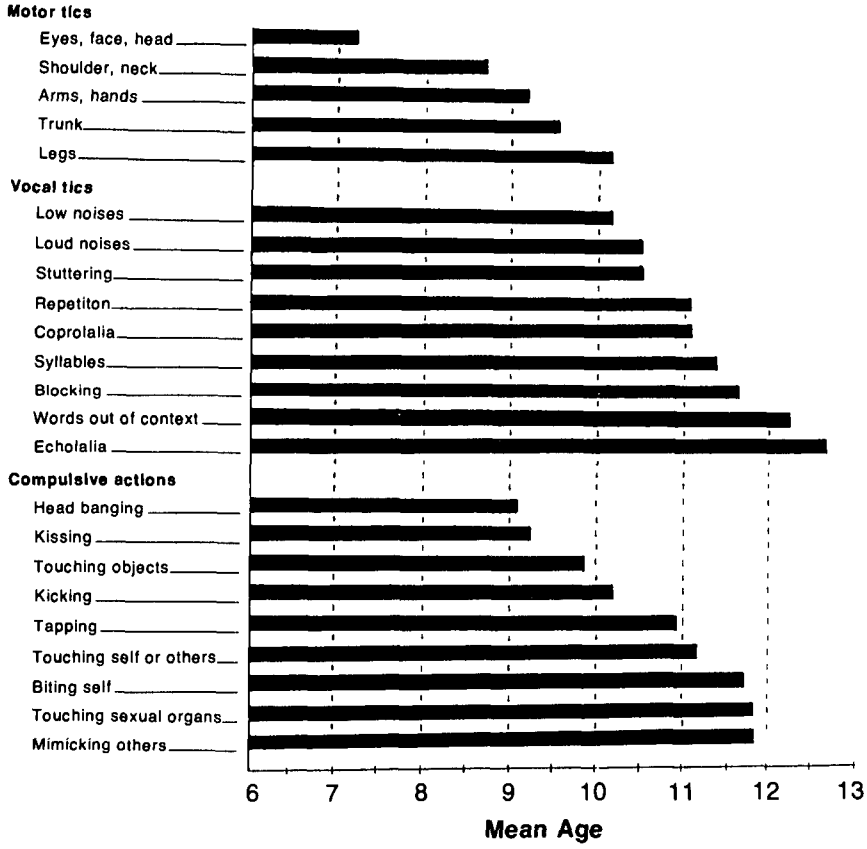
Table 1. Symptom abatement across patients ($n=75$)

Symptom	Patients ever experiencing		Patients ever experienced who are not now experiencing	
	Number	Percent	Number	Percent
Motor tics				
Eyes, face, or head	71	94.7	7	10.0
Shoulder or neck	69	92.0	18	26.1
Arms or hands	62	82.7	12	19.4
Trunk	46	61.3	9	19.6
Legs	46	61.3	16	34.8
Vocal tics				
Low noises	63	84.0	16	25.4
Loud noises	50	66.7	22	44.0
Syllables	20	26.7	8	40.0
Words (out of context)	22	29.3	3	13.6
Repetition	41	54.7	16	39.0
Echolalia	28	37.3	12	42.9
Stuttering	20	26.7	3	15.0
Blocking	28	37.3	4	14.3
Coprolalia	28	37.3	7	25.0
Compulsive actions				
Touching objects	41	54.7	6	14.6
Touching self or others	39	52.0	9	23.1
Mimicking others physically	16	21.3	7	43.8
Tapping	32	42.7	8	25.0
Kicking	27	36.0	10	37.0
Kissing	11	14.7	1	9.1
Touching sexual organs, self, or others	27	36.0	7	25.9
Biting self	9	12.0	2	22.2
Head banging	13	17.3	7	53.8

tients claimed that certain parts of the body currently were affected more severely by TS symptoms. The eyes, face, and head were named by 62 percent, the arms or hands by 21 percent, and the shoulders or neck by 13 percent. When patients were asked to rate the relative severity of 23 symptoms listed, which they currently were experiencing, no symptom was rated consistently as more severe than the others on the list. The mean severity of most symptoms ranged from mild to moderate. Several patients wrote in and rated the severity of symptoms which did not appear on our list, but in many cases the symptom that they specified was an example of one of the 23 symptoms listed. Some of the exceptions were turning in circles while standing or walking, sniffing objects or people; smelling fingers; mouthing, but not uttering, obscene words; spitting; pulling hair; irregular breathing patterns; mouthing objects; rocking self to sleep; and hitting self. The severity of these "other" symptoms was rated as slightly greater than those symptoms that were presented on our list. However, most of the "other" symptoms were mentioned by only one patient, and that patient may well have specified the symptom because it was severe or particularly troublesome.

The current severity question allowed us to compute the number of patients currently experiencing each symptom, and the number of symptoms currently being experienced by patients by age. Motor tics were reported most frequently (95 percent of the patients currently were experiencing one or more),

Figure 1. Mean age of occurrence of Tourette tics



with tics of the eyes or face currently being experienced by 85 percent of the patients, tics of the shoulders or neck by 68 percent, tics of the arms or hands by 67 percent, and tics of the trunk by 50 percent. Seventy-six percent of the patients currently were experiencing one or more vocal tics, and the uttering of low noises was the most frequently cited among them (63 percent of the patients). Compulsive actions were experienced currently by two-thirds of the patients; the most common compulsive action was touching objects (47 percent of the patients). Statistical analysis revealed no significant age differences in the type (motor tics, vocal tics, or compulsive actions) or number of tics reported as currently being experienced by patients.

Changes in Symptomatology. Fifty-two (69 percent) of the patients had experienced a period of time greater than 1 week during which symptoms were greatly reduced or disappeared entirely. Among 41 patients who described at least one such period, 24 (59 percent) were taking medication at the time and 17 (41 percent) were not.

Further, an examination of whether a patient who had ever experienced a particular symptom was currently experiencing the symptoms revealed that specific symptoms disappear slightly more often when patients are on medication but not significantly so. Table 1 presents the number and percentage of patients who ever experienced each symptom and the number and percentage of patients who had no longer had each symptom. Percentages of patients who no longer had a symptom ranged from a high of 54 percent (patients who once experienced head banging but did not at the time they were surveyed) to a low of 9 percent (patients who once experienced kissing but did not at the time they were surveyed), indicating that some symptoms were much more likely to abate than others. However, this did not appear to be related to the commonality of the symptom (e.g., motor tics of eyes, face, or head) or type of symptom (e.g., motor tic, vocal tic, or compulsive action).

Patients were requested to indicate how each of a number of situations affected their symptoms—that is, whether the situation did not affect symptoms, increased symptoms, or decreased symptoms. The general inconsistency of responses indicated marked differences in the reactions of individual patients to most situations. However, relative agreement among patients was noted for four situations: Symptoms increased during emotional trauma, when patients were fatigued, and when upset or anxious; symptoms decreased only during sleep.

Clinical Procedures. Patients reported undergoing a variety of clinical procedures. Seventy-three percent of the patients had been treated by either a psychiatrist or a psychologist for TS symptoms. Although only 16 percent felt that this treatment had helped in reducing symptoms, 38 percent

did believe that it had helped them cope with their symptoms. Twenty-seven percent of patients treated by a psychiatrist or psychologist had undergone psychotherapy or psychoanalysis, and the majority of them (75 percent) felt it had neither a positive nor a negative effect. A small number of patients had undergone behavior modification (12), hypnosis (9), physical therapy (5), or treatment by a chiropractor (10), and in each of these cases at least two-thirds believed that there was no beneficial effect. The one patient who had undergone electroshock therapy believed that the treatment had been harmful.

Electroencephalographic (EEG) Results. Sixty-two (83 percent) of the patients had had at least one EEG done. Of these, 58 percent reported a normal result on their most recent EEG, 11 percent reported a borderline result, and only 8 percent a clearly abnormal result; 19 percent did not know the result of their EEGs. Three patients failed to report their most recent EEG results.

Pharmacotherapy. Fifty-one (68 percent) of the patients indicated that they currently were taking medication for their TS symptoms; forty-six (63 percent) of the patients currently were taking haloperidol. The nature of the questionnaire did not enable us to determine the medication currently taken by the remaining five patients.

Since haloperidol has been the drug of choice for the treatment of TS, the patterns of characteristics of haloperidol usage among the patients sampled were assessed. Sixty-eight (91 percent) of the patients in our sample had taken haloperidol at some time (for a mean duration of 31 months), and 88 percent of those patients had experienced a reduction in symptoms. Of the patients who experienced a reduction in symptoms, 43 percent felt that their symptom reduction was uniform and not symptom-specific, while 50 percent stated that certain symptoms were helped more than others. The symptoms that patients cited as being helped more varied widely, but motor tics were mentioned slightly more frequently than other symptoms.

One or more undesirable side effects were noted by 80 percent of the patients who had ever taken haloperidol, with lethargy, impaired functioning, and depression accounting for the majority (60 percent) of the complaints (see table 2). Twenty (29 percent) of the patients who had ever taken haloperidol stopped taking it. The majority (80 percent) had discontinued use because of unpleasant side effects. Only two patients (10 percent of those who stopped) had stopped because haloperidol had been ineffective in treating symptoms.

Patients were asked to indicate on a list of other drugs those that they had taken in the past and whether those drugs had had a good or bad effect on their TS symptoms. For most drugs listed,

Table 2. Unpleasant side effects experienced by patients taking haloperidol

Side effect	Number reporting side effect ($n=63$)[1]	Percent of those responding to question
None	8	12.7
Lethargy	34	54.0
Impaired functioning	17	27.0
Depression	7	11.1
Parkinsonian symptoms	9	14.3
Weight gain	5	7.9
Impaired motor control	2	3.2
Restlessness	2	3.2
Headache	1	1.6
Slurred speech	1	1.6
Repetition of words	1	1.6
Loss of memory	1	1.6
Schizophrenic behavior	1	1.6
Numbness of tongue	1	1.6
Facial paralysis	1	1.6
Frequent urination	1	1.6
Got "high"	1	1.6
Drowsiness	1	1.6

[1] Side effects are not mutually exclusive. Some patients reported as many as three side effects. Sixty-eight patients had taken haloperidol at some time, but only 63 responded to this question.

patients noticed no effect on their symptoms. Exceptions for which a "good effect" was reported as frequently or more frequently than "no effect" were diazepam (45 percent) and benztropine mesylate (57 percent). A "bad effect" was reported as frequently or more frequently than "no effect" for phenytoin sodium (50 percent), methylphenidate hydrochloride (57 percent), and amphetamine derivatives (71 percent).

Haloperidol Usage and Symptom Presence. As mentioned above, most (92 percent) of the patients who were currently taking medication for their TS symptoms were taking haloperidol. We compared patients taking haloperidol with patients not taking it to see if the groups differed with regard to specific symptoms ever experienced that were "not" now experienced. There was a trend for patients taking haloperidol to have lost specific symptoms more than those not taking haloperidol, but there were no significant differences between the groups. It should be kept in mind that this analysis was based on the presence/absence of symptoms and not on the relative severity or frequency of a symptom or symptoms.

Family History. One section of the questionnaire dealt with the family history of TS and/or multiple tics. Each respondent was asked about all of his first-degree relatives (parents, siblings, children), his spouse, his aunts and uncles (by specific relationship), and his half-siblings (specifying shared parent for each). The information solicited on each relative was (1) whether that relative had ever had TS and (2) whether that relative had ever had multiple tics. Responses were "no," "yes," and "don't know."

Analysis of these data on parents and siblings of the TS patient disclosed that in families of TS patients the syndrome of multiple tics occurs more frequently in male than in female relatives. This finding offers a possible explanation for the preponderance of male patients in our sample. Further, our analysis revealed that the sex difference is related to the genetic transmission of susceptibility to TS. The complete results of this analysis are described elsewhere (Kidd, Prusoff, and Cohen 1980).

Prenatal and Birth History. Sixty-seven mothers of the patients surveyed completed this section of the questionnaire. These data have not been compared to similar data obtained from a control population, but the preliminary analysis found little out of the ordinary that would warrant such a comparison study.

Infancy and Early Childhood. The reported early development of the patients in our sample was unremarkable. During the first month of life they were active and strong (97 percent) and attentive to sights and sounds (100 percent). Developmental landmarks were achieved relatively early by most patients: 73 percent sat without assistance at 6 months or younger; 52 percent said words by 10 months and 92 percent by 17 months; walking began by 11 months for 43 percent and by 13 months for 85 percent. Ninety-two percent of the patients completed bowel and urinary training by about 3 years of age. The most frequently noted behavioral problems when patients were between 1 and 5 were sleeping problems (32 percent), being difficult to manage (27 percent), bladder and bowel problems (25 percent), and unusual fears (24 percent).

Ages 5 to 12. Enuresis was a problem for 27 percent of the patients beyond the age of 5, for 19 percent past age 6, and for 11 percent past age 9. Parent assessment of patient school performance from ages 5 through 12 indicated that similar numbers of patients were judged to have achieved at below average, average, and above-average levels in arithmetic, writing, and spelling. However, 51 percent of the respondents felt that the patient's school performance was not what it should have been. Thirteen percent of the patients had been kept back in school, and 6 percent had been advanced a grade sometime during this age period.

Classroom and learning difficulties when patients were between the ages 5 to 12 were frequently cited. Sixty-nine percent reported that teachers had complained of listening and attention problems, and parents of 50 percent of the patients concurred with this assessment. Forty-two percent of the patients had had teachers say that they were hard to control in the classroom. It should be noted that the following were not mutually exclusive: twenty-seven percent of the patients had been placed in special classes, 21 percent had been assigned a special teacher, and 19 percent had been diagnosed as having a learning disability. Forty percent of the patients were judged to be more awkward and clumsier than other children in the 5 to 12 age bracket. Twenty-five percent of the patients had

been diagnosed hyperactive by a physician between the ages of 1 and 11, and 12 percent were diagnosed as having minimal brain dysfunction (MBD) between the ages of 5 and 12; some patients had received both diagnoses at different times.

The social stigma of TS at these ages was evidenced by the high percentage (75 percent) of patients who were teased about their symptoms by classmates. However, 72 percent of the patients stated that they had had a best friend when they were between the ages of 5 and 12, and 92 percent had some friends.

Ages 12 to 18. Between the ages of 12 and 18, patients again were distributed similarly in below-average, average, and above-average groups for arithmetic, writing, and spelling. However, respondents reported that for 66 percent of the patients, school performance was not what it should have been compared with 51 percent judged to be performing below ability from ages 5 to 12. Fewer patients were said to have been hard to control in the classroom between ages 12 and 18 (27 percent) than between ages 5 and 12 (42 percent). Listening and attention continued to be problems for patients between ages 12 and 18 (49 percent). Thirty-one percent of the patients were in special classes and 22 percent were assigned a special teacher during this age period. Thirty-five percent of patients were reported to be more awkward or clumsier than others between the ages of 12 and 18. However, 83 percent participated in sports, and of those participating, 79 percent were considered to be at least average athletes.

Seventy-seven percent of the patients reported that they had had a best friend, and 51 percent had dated when they were between the ages of 12 and 18. Virtually all (98 percent) of the patients had some friends, but 73 percent had been teased by peers about TS symptoms at these ages. Forty-eight percent of the patients felt that teachers had been unfair or difficult to deal with, and 23 percent reported that they had been teased or ridiculed by teachers about TS symptoms.

Other Developmental Findings. Fifty-six percent of the patients were reported to be above average in overall intelligence, and 41 percent were average. Questions concerning patient handedness revealed that 82 percent were right handed and 17 percent left handed. No patient reported having been forced to change handedness. There were no unusual findings concerning age of onset of puberty.

Emotional Status. Sixty-seven percent of the patients had seen a mental health professional for emotional or psychological problems; 13 percent had been hospitalized for psychiatric reasons. Off medication, the aggressiveness of the patients in our sample was reported to be above average for 39 percent of the patients, average for 32 percent, and below average for 29 percent. Patients' sensitivity to pain off medication was considered to be normal for 54 percent of the patients, above average for 29 percent, and below average for 16 percent.

Effects on Family Relationships. Forty percent of the patients with siblings felt that their relationships with brothers and sisters were affected for the worse by TS, but the majority (58 percent) felt that these relationships were not affected at all. The relationships between patients and parents were found to be affected slightly more often, with 50 percent claiming that the relationship was affected for the worse and 46 percent not at all.

Effects on Social Contact. As can be seen in table 3, when patients were asked how the quality of their social contact was affected by TS, 12 percent claimed not at all, 32 percent only a little, 30 percent

Table 3. Degree to which Tourette syndrome affects patients' social contact, school performance, and job performance

Degree affected	Social contact outside family (percent)	School performance (percent)	Job performance (percent)
Not at all	11.6	15.1	29.6
Only a little	31.9	32.1	37.0
Moderately	30.4	20.8	11.1
Substantially	14.5	20.8	14.8
Severely	11.6	11.3	7.4
Number responding	$n = 69$	$n = 53$	$n = 27$

moderately, 14 percent substantially, and 12 percent severely.

Effects on School and Job Performance. Among student patients, school performance was not felt to be affected by 15 percent, was felt to be affected only a little by 32 percent, moderately by 21 percent, substantially by 21 percent, and severely by 11 percent. Among patients with full-time occupations, 30 percent felt their job performance was not affected by TS, 37 percent that it was slightly affected, 11 percent moderately, 15 percent substantially, and 7 percent felt that their job performance was severely affected (see table 3).

Specific Difficulties Encountered by TS Patients. Patients were asked to respond to questions concerning specific difficulties they have faced. Among those who had the necessary experience to respond to each question, 14 percent encountered difficulties in obtaining health insurance, 13 percent in obtaining life insurance, 13 percent in obtaining a driver's license, 25 percent in obtaining employment, 34 percent in holding a job, and 9 percent in gaining admission to college or other higher education (see table 4).

Patient Attitude Toward Having TS. When patients were asked to characterize their current attitude toward having TS (table 5), 16 percent said that they felt like an average person, 49 percent said that TS is a problem but one they face as a challenge, 24 percent said that it is a serious problem but that they compensate when possible, and 10 percent that they had serious shortcomings and were pessimistic about the future.

Table 4. Percentage of patients experiencing specific difficulties as a result of Tourette syndrome

Area of difficulty	Percent experiencing difficulty	Number responding
Obtaining health insurance	13.9	36
Obtaining life insurance	12.9	31
Obtaining a driver's license	13.3	30
Obtaining employment	25.0	28
Holding a job	34.5	29
Gaining admission to higher education	8.7	23

Table 5. Patient attitude toward having Tourette syndrome (n = 67)

Attitude	Percent
Feels like an average person	16.4
Is a problem but faces it as a challenge	49.3
Is a serious problem but compensates when possible	23.9
Feels serious shortcomings, is pessimistic about the future	10.5
Withdrawn, ashamed, overcome	0

Discussion

The major objective of this study was to gather a broad spectrum of information on TS patients. The low (37.5 percent) response rate to our pilot survey was disappointing; nevertheless, our sample comprises a diverse group of TS patients. While response to the questionnaire may have been related to the demographic characteristics of the sample, there is no reason to believe that nonresponse was related to the history, symptomatology, or treatment of the disorder.

Validation of the TSQ. Both the literature and our clinical experience support the usefulness and validity of the TSQ. Indeed, our survey findings are remarkably similar to those reported (Shapiro et al. 1978; Abuzzahab and Anderson 1973; Nee et al. 1980) and our clinical project findings (Cohen et al. 1980), namely: the 3.2:1 male to female ratio; the age of onset between 2 and 16 years, with a mean of 7 years; the suggestion of a cephalo-caudal sequence in the development of motor tics; overall symptomatology and course; treatment history; and genetic susceptibility to TS.

Diagnosis of TS. While any delay in the diagnosis of TS can lead to unnecessary bewilderment and in-

appropriate treatment, our data show that patients now are likely to go undiagnosed for much shorter periods. However, most patients had been assigned at least one diagnosis other than TS. Insidious onset and clinicians' lack of experience with TS may account for some of the "misdiagnosis" (e.g., transient tics of childhood and chorea), but in other cases patients may well have met the criteria for the diagnosis assigned them. For example, a number of patients had been diagnosed hyperactive, with a learning disability, or with minimal brain dysfunction (MBD)—diagnoses which do not share essential criteria with TS, but which have been associated with it (Moldofsky, Tullis, and Lamon 1974; Shapiro et al. 1978). The *DSM-III* (American Psychiatric Association 1980) recognizes that a patient may meet criteria for more than one disorder and provides for multiple Axis I and II diagnoses. For the TS patient, concomitant diagnoses are of particular value in the implementation of a therapeutic program (e.g., special education). Further research is needed to delineate the relationships between the diagnostic criteria for TS and other disorders that appear frequently in association with it.

Subgroups of Patients. The TSQ and the clinical data support the notion of heterogeneity, on clinical and genetic grounds, among TS patients. However, whether there is currently any strong evidence for this heterogeneity having clinical or biological implications is not known. We do not yet know if there are important biological or therapeutic response differences between patients with or without coprolalia, with or without family history, or with or without EEG abnormalities. To determine the meaning of these within-diagnosis differences, future research should use increasingly refined typologies including family history, symptom pattern, associated or other diagnoses, and response to medication.

EEG Results. Eighty-three percent of the patients had had at least one EEG; of these, only 11 percent reported a borderline result and 8 percent a clearly abnormal result. Since physicians tend to minimize EEG results to patients, and since a large number of the patients (19 percent) did not know the results of their EEGs, these findings may represent an underreporting of the actual incidence of borderline and abnormal EEGs among the patients in this sample.

Haloperidol Usage. Our data show that haloperidol is effective in the treatment of TS and is useful for some patients for whom side effects are minimal and the therapeutic gains worth the inconvenience they may experience. The fact that haloperidol was thought to be the "magic cure" for TS has held up research into other possible treatments and, clearly, for many of the patients in our sample alternatives are needed.

Effects of TS on Patient Functioning. Patient responses to a number of questions regarding the effects of TS on their attitudes toward life, school, and job performances, family and social relationships, and other areas of functioning revealed that some patients are remarkably optimistic about the future and have managed to maintain "normal" lives in spite of TS. However, our data suggest that TS has caused serious problems for a majority of patients and has been debilitating for many. The reported differences in patient attitudes and patient functioning may reflect actual differences in the severity of their symptoms. In addition, the fact that symptoms had disappeared or were greatly reduced for at least one period lasting 1 week or more (reported by 69 percent of the patients) may have contributed to the optimism expressed by some patients.

School and Work Problems. Ninety-seven percent of the patients in our sample were reported to be of average or above-average intelligence, yet the majority of them reported school and attentional difficulties. Many had required special educational provisions. A number of patients had had difficulty obtaining and/or holding a job, and among those with full-time occupations, one-third claimed that their job performance was moderately to severely affected by TS. On the basis of these data and our clinical findings, we believe that these reported school- and work-related problems are due not only to the limitations imposed by TS symptoms, e.g., noises and compulsions, but also to basic attentional difficulties, which may be exacerbated by medication side effects.

Social and Family Relationships. Most respondents reported that social and/or family relationships were negatively affected by TS. Three-quarters had been teased by peers, nearly half had found teachers unfair or difficult to deal

with, and nearly one-fourth had been teased or ridiculed by teachers. Social contact was reported to be moderately to severely affected by more than half of the patients. While TS would seem to strain family relationships, many patients felt that relationships with siblings and parents had not been affected at all by their disorder.

References

Abuzzahab, F.E., Sr., and Anderson, F.O. Gilles de la Tourette's syndrome: International Registry. *Minnesota Medicine,* 56:492–496, 1973.

American Psychiatric Association. *DSM–III: Diagnostic and Statistical Manual of Mental Disorders*. 3rd ed. Washington, D.C.: APA, 1980.

Cohen, D.J.; Detlor, J.; Young, J.G.; and Shaywitz, B.A. Clonidine ameliorates Gilles de la Tourette syndrome. *Archives of General Psychiatry,* 37:1350–1357, 1980.

Cohen, D.J.; Shaywitz, B.A.; Caparulo, B.K.; Young, J.G.; and Bowers, M.B., Jr. Chronic, multiple tics of Gilles de la Tourette's disease: CSF monoamine metabolites after probenecid administration. *Archives of General Psychiatry,* 35:245–250, 1978.

Cohen, D.J.: Shaywitz, B.A.; Young, J.G.; Carbonari, C.M.; Nathanson, J.A.; Lieberman, D.; Bowers, M.B., Jr.; and Maas, J.W. Central biogenic amine metabolism in children with the syndrome of chronic multiple tics of Gilles de la Tourette. *Journal of the American Academy of Child Psychiatry,* 18:320–341, 1979*a*.

Cohen, D.J.; Young, J.G.; Nathanson, J.A.; and Shaywitz, B.A. Clonidine in Tourette's syndrome. *Lancet,* II:551–553, 1979*b*.

Friedhoff, A.J., and Alpert, M. Receptor sensitivity modification as a potential treatment. In: Lipton, M.A.; DiMascio, A.; and Killam, K.F., eds. *Psychopharmacology: A Generation of Progress*. New York: Raven Press, 1978. pp. 797–801.

Gilles de la Tourette, G. Étude sur une affection nerveuse caracterisée par de l'incoordination motrice accompagnée d'echolalie et de coprolalie (jumping, latah, myriachit). *Archives Internationales de Neurologie de Maladies Hereditares de Medicine Mentale et Psychosomatique,* 9:19–42, 158–200, 1885.

Hollingshead, A.B. *Two Factor Index of Social Position*. New Haven: Yale Station, 1965.

Kidd, K.K., Prusoff, B.A.; and Cohen, D.J. The familial pattern of Tourette syndrome. *Archives of General Psychiatry,* 37:1336–1339, 1980.

Lake, C.R.; Ziegler, M.C.; Eldridge, R.; and Murphy, D.L. Catecholamine metabolism in Gilles de la Tourette's syndrome. *American Journal of Psychiatry,* 134:257–260, 1977.

Mahler, M.S.; Luke, J.A.; and Daltroff, W. Clinical and follow-up study of the tic syndrome in children. *American Journal of Orthopsychiatry,* 15:631–647, 1945.

Moldofsky, H.; Tullis, C.; and Lamon, R. Multiple tic syndrome (Gilles de la Tourette's syndrome). *Journal of Nervous and Mental Disease,* 159:282–292, 1974.

Nee, L.; Caine, E.D.; Polinsky, R.J.; Eldridge, R.; and Ebert, M.H. Gilles de la Tourette syndrome: Clinical and family study of 50 cases. *Annals of Neurology,* 7(1):41–49, 1980.

Shapiro, A.K., and Shapiro, E. Treatment of Gilles de la Tourette's syndrome with haloperidol. *British Journal of Psychiatry,* 114:345–350, 1968.

Shapiro, A.K.; Shapiro, E.; Bruun, R.D.; and Sweet, R.D. *Gilles de la Tourette Syndrome*. New York: Raven Press, 1978.

van Woert, M.H.; Jutkowitz, R.; Rosenbaum, D.; and Bowers, M.B., Jr. Gilles de la Tourette's syndrome: Biochemical approaches. In: Yahr, M.D., ed. *Basal Ganglia*. New York: Raven Press, 1976. pp. 459–465.

Wassman, E.R.; Eldridge, R.; Abuzzahab, F.S.; and Nee, L. Gilles de la Tourette syndrome: Clinical and genetic studies in a mid-western city. *Neurology,* 28:304–307, 1978.

Acknowledgment

The authors thank the Tourette Syndrome Association, Inc., for their generous assistance with the pretesting of an earlier version of the questionnaire and with collecting these pilot study data. Gratitude is also extended to Margaret Fogarty for her assistance with data analysis. This project was supported by the Yale Mental Health Clinical Research Center (MHCRC) grant MH–30929.

The Authors

Janine Jagger, M.P.H., is Instructor and student, Division of Health Services Research, University of Virginia Medical Center, Richmond, VA. At the time this research was carried out, Ms. Jagger was Associate in Research, De-

partment of Psychiatry, Yale University School of Medicine. Brigitte A. Prusoff, Ph.D., is Senior Research Associate, Department of Psychiatry; Donald J. Cohen, M.D., is Professor of Pediatrics, Psychiatry, and Psychology, Child Study Center; Kenneth K. Kidd, Ph.D., is Associate Professor of Human Genetics and Psychiatry; Claudia M. Carbonari, M.S., is Associate in Research, Child Study Center; and Karen John, B.A., is Associate in Research, Department of Psychiatry, Yale University School of Medicine, New Haven, CT.

Ninth World Congress of Social Psychiatry

The **Ninth World Congress of Social Psychiatry** will be held in **Paris, France, July 5–9, 1982**. The Congress is being organized by L'Association Française de Psychiatrie et de Psychopathologie Sociales (President of the Congress: P. Jean; President of the Scientific Council: P. Sivadon). Its main aims are:

- To take stock of the most topical issue in the field of mental health.
- To provide a forum for discussion among representatives of different cultures and all professionals concerned with the mental health field.

The program will focus on psychopathological problems associated with underemployment (unemployment, labor mobility, technological development, social behavior of young people, the aging worker, and dismissal); attitudes of the general public toward health and prevention measures (contraception, "corner street groups," prevention campaigns, mental hygiene, and alternatives to medical care); social indicators and mental health indicators (sects, alcoholism, suicide, fatigue, violence, criminality, and toxicomania); psychopathological aspects of the consumption of drugs (iatrogenic processes, use of drugs, prescription of drugs, free health care, and self-medication).

For futher information, contact the **Secretariat, PMV, 9^{e} Congrès Mondial de Psychiatrie Sociale, BP 246, 92205 Neuilly-sur-Seine (France)**.