Am J Hum Genet 37:435–450, 1985

Tourette Syndrome: Clinical and Psychological Aspects of 250 Cases

DAVID E. COMINGS[1] AND BRENDA G. COMINGS[2]

SUMMARY

Tourette syndrome is a common hereditary neuropsychiatric disorder consisting of multiple tics and vocal noises. We summarize here clinical aspects of 250 consecutive cases seen over a period of 3 years. The sex ratio was four males to one female, and the mean age of onset was 6.9 years. Only 10% were Jewish, indicating that it is not more prevalent in Ashkenazi Jews. Only 33% had compulsive swearing (coprolalia), indicating that this is not necessary for the diagnosis. The most frequent initial symptoms were rapid eye-blinking, facial grimacing, and throat-clearing. In this series, it was clear that Tourette syndrome is a psychiatric as well as a neurological disorder. Significant discipline problems and/or problems with anger and violence occurred in 61%, and 54% had attention-deficit disorder with hyperactivity. Some degree of exhibitionism was present in 15.9% of males and 6.1% of females. Obsessive-compulsive behavior was seen in 32%. Other than tics and vocal noises, the most common parental complaints were of short temper and everything being a confrontation. There were no significant clinical differences between familial and sporadic cases.

Whenever a child presents with a learning disorder, attention-deficit disorder, or significant discipline or emotional problems, the parents should be questioned about the presence of tics or vocal noises in the patient and other family members.

INTRODUCTION

Tourette syndrome (TS) [1–7] is a hereditary neuropsychiatric disorder characterized by muscular tics and vocal noises. Family studies indicate that TS and

Received November 9, 1984; revised January 7, 1985.

[1] Department of Medical Genetics, City of Hope National Medical Center, 1500 E. Duarte Rd., Duarte, CA 91010.

[2] Private practice.

chronic multiple tics (MT) (motor tics alone or vocal noises alone) are genetically related [4–6] and due to a single major gene [6, 7]. It is far more common than previously thought, with recent estimates of a lifetime risk for the TS-MT trait of 0.5%–1.6% [5, 6]. It continues to be an underrecognized disorder. This review examines clinical and psychiatric aspects of 250 cases seen over a period of 3 years.

METHODS

All patients were interviewed using a structured, standard questionnaire. Over 95% of the patients were self-referred, referred by school districts, or referred by the Tourette Syndrome Association. The Tourette Syndrome Clinic is part of the Department of Medical Genetics (City of Hope) and, as such, sees patients without financial considerations or the usual institutional separation of adult and pediatric cases. All consecutive patients, fitting the DSM-III criteria of TS, were included in the series. As a result of the method of referral, the series is not preselected for severity, resistance to treatment, age, sex, psychiatric problems, family history, or any other characteristic and patients are prospectively examined in a uniform manner. Inclusion in the series required satisfying the six DSM-III [8] criteria: (1) onset between ages 2 and 15, (2) multiple motor tics, (3) vocal tics, (4) waxing and waning of symptoms, (5) ability to suppress symptoms for varying periods of time, and (6) presence of symptoms for more than 1 year.

The presence of attention-deficit disorder (ADD) with hyperactivity was diagnosed as described previously [9]. It was divided into three grades. In grade 0, it was not present. In grade 1, the patient satisfied the DSM-III criteria [8] for ADD with hyperactivity based on interviewing the parent or patient and examining school and psychologist records. In grade 2, the diagnosis had been made previously by another physician and the patient had been placed on stimulant medication. Using this same group, we have previously reported a detailed analysis of the relationship between TS and ADD and the role, if any, of stimulant medication in the production of tics [9].

To minimize the subjectivity of classification, the cases were placed into only three classes of severity (table 1). In grade 1 (12%), the tics were too mild to require treatment. This was either because the symptoms had always been mild or because there had been a significant decrease in the severity of the symptoms in the past several years. In grade 2 (59%), the tics were sufficiently severe to require treatment. Grade 3 cases (29%) were those in which the tics required treatment and there was a significant interference with the patient's life. This was easiest to determine in older patients who were supported by welfare or other social services because of their inability to hold a job. Younger cases were placed in this category when there were long-term significant problems in school. To avoid circular reasoning, it was not based on the presence of discipline problems or violence. Shapiro et al. [10] divided their cases into four categories: mild, 23% (similar to our grade 1); moderate, 34%; marked, 33%; and severe, 11%.

RESULTS AND DISCUSSION

General Features

Table 1 provides a summary of a number of different features seen in this series. The male/female sex ratio was 4:1. The modal age in 5-year intervals, for both sexes, was 10–15 years. There were no significant preceding illnesses that might have triggered the onset of symptoms.

Age of Onset

The average age of onset of tics or vocal noises was 6.9 years. In nine prior studies of 221 patients, the mean age of onset was 7.3 years [2]. The distribution is shown in figure 1.

TABLE 1

SUMMARY OF VARIOUS FEATURES OF THE 250 CASES OF TS

Feature	Males		Females		All	
Sex—No. (%)	201	(80)	49	(20)	250	(100)
Age—mean (SD)	17.0	(10.0)	26.3	(17.1)	18.2	(12.3)
Age-onset of symptoms—mean (SD)	7.0	(3.0)	6.7	(2.5)	6.9	(3.0)
Years from onset to diagnosis—mean (SD)	9.5	(9.5)	18.0	(16.5)	11.2	(11.7)
Severity of TS—No. (%):						
Grade 1	28	(14)	3	(6)	31	(12)
Grade 2	116	(58)	31	(63)	147	(59)
Grade 3	57	(28)	15	(31)	72	(29)
Hyperactivity—No. (%):						
Grade 0	86	(43)	29	(59)	115	(46)
Grade 1	57	(28)	17	(35)	74	(30)
Grade 2	58	(29)	3	(6)	61	(24)
Grade 1 + 2	115	(57)	20	(41)	135	(54)
Jewish—No. (%)	21	(10)	5	(10)	26	(10)
Coprolalia—No. (%)	56	(28)	19	(39)	75	(33)
Age of onset coprolalia—mean (SD)	10.6	(5.6)	10.2	(4.7)	10.5	(5.4)
Years from onset of symptoms to onset of coprolalia—mean (SD)	3.8	(5.8)	4.0	(4.8)	3.8	(5.6)
Discipline problems—No. (%):						
Grade 0	110	(55)	29	(59)	139	(56)
Grade 1	57	(28)	10	(20)	67	(27)
Grade 2	29	(14)	9	(18)	38	(15)
Grade 3	5	(2)	1	(2)	6	(2)
Grade 1–3	91	(45)	20	(41)	111	(44)
Anger and violence—No. (%):						
Grade 0	116	(58)	29	(59)	145	(58)
Grade 1	42	(21)	13	(26)	55	(22)
Grade 2	41	(20)	7	(14)	48	(19)
Grade 3	2	(1)	0	(0)	2	(1)
Grade 1–3	85	(42)	20	(40)	105	(42)
Exhibitionism—No. (%):						
Grade 0	169	(84)	46	(94)	215	(86)
Grade 1	10	(5)	2	(4)	12	(5)
Grade 2	6	(3)	1	(2)	7	(3)
Grade 3	16	(8)	0	(0)	16	(6)
Grade 1–3	32	(16)	3	(6)	35	(14)

The age of onset of symptoms in relation to the grade of severity of TS and the grade of hyperactivity is shown in table 2. For each grade of severity of TS, the age of onset of symptoms was earlier for patients with ADD and hyperactivity than for patients without hyperactivity. The average age of onset was progressively higher going from grade 1 TS (6.08 years) to grade 2 (6.83) years and grade 3 (7.53) years. This trend was present in both the patients with and without hyperactivity. Thus, the extremes were an average age of onset of 4.65 years for grade 1 TS with hyperactivity to 8.14 years for grade 3 TS without hyperactivity. While this might suggest that patients with a late age of onset tend to end up with more severe symptoms and more likely to need treatment

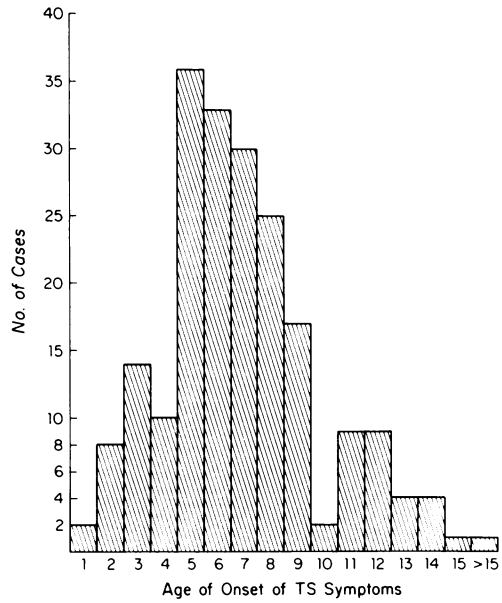

FIG. 1.—Distribution of age of onset of the 250 cases of TS

than those with an early age of onset, there may be an ascertainment bias contributing to these results. For example, older patients are somewhat more likely to be classified as grade 3, since they have a longer time period over which to judge if TS has resulted in a significant interference in their life.

In almost all cases, the symptoms had been present for a number of years before a diagnosis was made. This averaged 9.5 years for males, 18.0 years for females, and 11.2 years for all cases. The maximum was 65 years. While it

TABLE 2

AGE OF ONSET OF SYMPTOMS IN RELATION TO SEVERITY OF TS AND GRADE OF HYPERACTIVITY

	GRADE OF HYPERACTIVITY								
	0			1 OR 2			ALL		
GRADE OF TS	No.	Average	SD	No.	Average	SD	No.	Average	SD
1	21	6.76	2.93	10	4.65	1.84	31	6.08	2.80
2	72	7.10	2.92	75	6.57	2.41	147	6.83	2.69
3	22	8.14	3.28	50	7.26	3.41	72	7.53	3.34
1–3 ...	115	7.23	3.03	135	6.68	2.87	250	6.94	2.94

NOTE: Significant comparisons were—
Grade 1 or 2 hyperactivity:
Grade 1 vs. Grade 2 TS: $t = 2.42$, df = 83, $P = < .02$.
Grade 1 vs. Grade 3 TS: $t = 2.34$, df = 58, $P = < .025$.
All cases:
Grade 1 vs. Grade 3 TS: $t = 2.11$, df = 101, $P = < .05$.

would be anticipated that the increased attention given to the syndrome in recent years would result in a more rapid diagnosis, this figure is not significantly different from that of 12.1 years given by Shapiro et al. [1] in their summary of 145 cases seen from 1965 to 1974. Although virtually every patient had been to some type of physician with their complaints, in 76%, the presumptive diagnosis was made only after they had heard of the syndrome in the mass media or from a friend. Others have reported as high as 80% self-diagnosis [10]. When the diagnosis was missed, some of the most frequently encountered statements were: "Don't worry, it is just a habit and will go away," or "It is psychological in origin."

Reasons for Missed Diagnosis

We have observed three major factors for the failure of professionals to make a diagnosis of TS: (1) Lack of familiarity with the syndrome. In the majority of cases when the patients asked their physician about Tourette syndrome, the physician had not heard of it before. Lack of familiarity with the syndrome also meant that the appropriate questions that are required to make a diagnosis are not asked. (2) Suppressibility of symptoms. Most patients, especially those with relatively mild symptoms, are able to completely suppress both the motor and vocal tics for periods of up to several hours. A frequent complaint of the parent was that their child was ticing in the car on the way to the doctor's office, stopped ticing during the visit, then resumed ticing on the way home. The doctor, meanwhile, believes that the mother is imagining things or that if they did not see the tics themselves the patient could not possibly have them. This suppressibility, however, is one of the essentials for making the diagnosis rather than a reason to reject it. Because of suppressibility, the diagnosis often rests on the history alone. (3) Belief that coprolalia must be present. Parents have often been told that their child did not have TS because he is not having compulsive swearing. As discussed below, coprolalia is actually present in only one-third of the cases and is not a requirement for diagnosis. Golden [11] listed similar reasons for missed diagnoses including confusion with transient tic of childhood with the parent being told they will go away, misinterpretation of chronic coughing, sniffing, and throat-clearing as indicative of respiratory tract disease or allergies, and believing that the symptoms are psychological in origin. In children with significant behavioral problems, a common misdiagnosis was "childhood schizophrenia."

Presenting Symptoms

Tics of the facial muscles were the most frequent presenting symptom, occurring in 66% of patients. Of these, eye-blinking was most prominent (48%). With the onset of eye-blinking, parents often took their child to the pediatrician or ophthalmologist and were usually told it was due to an allergy and would disappear with time. Facial grimaces consisted of a wide range of movements including nose-twitching, stretching of the mouth in any direction, pursing the lips, and all combinations of these. Mouth-opening was the presenting symptom in 7.6%. Licking of the lips occasionally resulted in chronic discoloration

or open lesions around the mouth (that disappeared with haloperidol treatment).

Vocal noises occurred as a presenting symptom in 32% of cases. Of these, the most frequent was repeated throat-clearing (13%). Coprolalia, as a presenting symptom, was unusual, occurring in only 3%.

Motor and Vocal Tics

The frequency of motor and vocal tics throughout the course of TS is shown in table 3. Again, facial tics were the most frequent, occurring in 82%, and included rapid eye-blinking (56%), mouth-opening (38%), and facial grimacing (29%). Head and neck tics were present in 63%. The most characteristic of these was a "hair-out-of-eyes tic." Excessive use of the finger sign (3%) is termed copropraxia. Diaphragmatic tics were usually accompanied by grunting noises. Skipping tics often resulted in the children being labeled "queer" by their peers. Hitting themselves on almost any part of their body was present in 7%.

Other tics occurred in 19% of males and 29% of females. Some examples are rocking, blowing on or kissing their hands, twirling hair, often to the point of producing a bald spot, and chewing on clothing. A rearrangement of clothing as if it was too tight or didn't fit right was a frequent theme.

Self-abusive behavior was present in 33 patients (13%). In some, biting themselves on the hand was so frequent that marked callouses developed. Based on a questionnaire sent to 111 TS families, VanWoert et al. [12] reported head banging in 13; hand, tongue, cheek and/or lip biting in eight; hitting themselves in eight; and producing skin ulcerations by scratching in four.

Complex tics are those in which a tic in one part of the body is reproducibly followed by a secondary tic. These occurred in 21% of males and 24% of females. The most frequent class was a head tic followed by a tic of one of the extremities. The ritualistic nature of some is indicated by one patient who, whenever he took a shower, first touched the floor, then turned around exactly four times.

Throat-clearing was the most frequent vocal tic, occurring in 56% of patients. Sniffing was often misdiagnosed as a nasal allergy. Spitting was the most unpleasant of the tics. Unlike motor tics, which can be ignored by looking away, parents or spouses especially complain about vocal tics. Even relatively quiet ones begin to grate on one's nerves by virtue of their constant presence. They are also the symptoms that most frequently get children in trouble in the classroom. If teachers do not understand their origin, they often assume the children are purposely trying to either get attention or be disruptive.

Coprolalia was taken as being present only if the use of swear words was impulsive and totally inappropriate to the situation. Simple swearing when angry is not coprolalia. In this series, 28% of males, 39% of females, and 33% of all patients had coprolalia. The difference between males and females was significant at $P = .01$. TS can be viewed as a "disinhibition syndrome" due to release of the inhibiting neuronal pathways. The increased frequency of coprolalia in females may be because swear words may be more strongly inhibited

TABLE 3

TYPE AND FREQUENCY OF MOTOR AND VOCAL TICS THROUGHOUT THE COURSE OF TS

Motor tics					
	No.	%		No.	%
Facial	82	33	Shoulder	32	13
Eyeblinking	56	22	Arms	23	9
Mouth-opening	38	15			
Facial grimacing	29	12	Hands	15	6
Rolling eyes	9	4	Flexing-clasping	5	2
Squinting eyes	4	2	Piano fingers	6	2
Opening eyes	3	1	Copropraxia	3	1
Closing eyes while driving	3	1	Other	6	2
Sticking tongue out	6	2			
Licking lips	4	2	Diaphragm	12	5
Licking shoulder	3	1			
Biting tongue	1	.5	Legs and feet	21	8
Looking at the sun	1	.5	Kicking	7	3
Grinding teeth	3	1	Hopping-skipping	7	1
			Flexing feet	3	3
Head and neck			Other	8	3
Hair out of eyes	63	25			
Horizontal head tic	12	5	Hitting self	7	3
Vertical head tic	20	8			
Chin on shoulder	9	4			
	3	2			

Vocal tics					
	No.	%		No.	%
Throat-clearing	139	56	Humming	14	6
Grunting	79	32	Yelling-screaming	17	7
Sniffing	37	15	Blow-out breath	16	6
Spitting	22	9	Suck-in breath	11	4
Barking	21	8	Whistling	5	2
Snorting	16	6	Other	52	21
Squeeking	15	6	No Vocal tics	9	4
Coughing	17	7			

in females than in males. The duration from onset of symptoms to onset of coprolalia averaged 5.4 years. In the study of 75 TS patients by Jagger et al. [13] using mailed questionnaires, 37% had coprolalia with 3.5 years from onset of symptoms to onset of coprolalia.

Jewish vs. Non-Jewish

Tourette syndrome has been reported as a disorder especially common in Ashkenazi Jews. In the three series described by Shapiro et al. [1] of 34, 80, and 31 patients, 61%, 49%, and 16% were Jewish. Despite the lower frequency in the third series, 45% of the total series were Jewish. Wassman et al. [14] also found that a high proportion of 14 families in Minnesota were of Jewish and

Eastern European ancestry. This contrasts sharply with the present series in which only 10% were Jewish, which reflects the approximate proportion of Jews living in Los Angeles. The high frequency of Jews in the Shapiro et al. [1] series is probably simply a reflection of the high percentage of Jews in the New York area. This is consistent with the report of Golden [15] who found that 28% of patients from a New York series were Jewish while only 2.7% from a Texas series were Jewish. Nee et al. [16] also found no preponderance of families with a Jewish or Eastern European background.

Nationality

As opposed to other series suggesting a high frequency of patients of Eastern European ancestry, the most frequent ancestry in this series was English (13%), German (13%), and Irish (14%). There were few nationalities that were not included. Notable inclusions were blacks (1.8%) and Mexicans (8.9%).

Discipline Problems

One of the major recurrent themes in interviewing Tourette families is the presence of discipline problems. These were divided into four grades.

In grade 0, there were no discipline problems identified by the parents or remembered by the older patients.

In grade 1, parents or patients believed that problems with discipline were significantly outside the realm of normal, long-term, and clearly greater than with unaffected siblings. Typical parental comments were: "every interaction escalates into a major confrontation," "he seems to have been angry ever since the onset of the tics," "unpredictable," "overreacts to minor things," "always finds a negative aspect in every thing," and "life with him is a constant battle."

In grade 2, the discipline problems were severe and often led to feelings of total helplessness on the part of the parents. Marital problems due to constant conflict on how to handle the child were common. Typical comments were: "want's immediate gratification," "he is impossible to discipline, it is easier to just ignore him" (learned helplessness) [17], "he is always stealing things from us and our friends," "Jekyll-and-Hyde personality," "shows a wanton disregard of the rules and does the opposite of what he is told," "reacts violently to criticism," "very explosive," and "I'm afraid I'm going to become a child abuser."

In grade 3, the problems were so great that the patient had to be institutionalized at some time in his or her life either in a juvenile correction facility or mental hospital.

Table 4*A* shows the relationship between grade of discipline problems in males and females and the presence or absence of hyperactivity. This indicates that: (1) the discipline problems are comparable in frequency in both males (45.3%) and females (40.8%), (2) there is some relationship to the presence of hyperactivity in that the frequency of grades 1–3 discipline problems is approximately two times greater in those with hyperactivity compared to those without, (3) the significant frequency of discipline problems in those without hyperactivity suggests that this is a feature of TS even in the absence of hyperactivity.

TABLE 4

DISCIPLINE PROBLEMS IN TS

A. Discipline problems in TS patients with and without hyperactivity

GRADE OF DISCIPLINE PROBLEM	WITH HYPERACTIVITY			WITHOUT HYPERACTIVITY			ALL		
	No.	%		No.	%		No.	%	
Males									
0	52	45.2		58	67.4		110	54.7	
1	37	32.2		20	23.3		57	28.4	
2	22	19.1	54.8	7	8.2	32.6	29	14.4	45.3
3	4	3.5		1	1.1		5	2.5	
Total	115	100.0		86	100.0		201	100.0	
Females									
0	8	40.0		21	72.0		29	59.2	
1	4	20.0		6	20.1		10	20.4	
2	7	35.0	60.0	2	6.9	27.0	9	18.4	40.8
3	1	5.0		0	0.0		1	2.0	
Total	20	100.0		29	100.0		49	100.0	

B. Discipline problems and severity of TS

GRADE OF DISCIPLINE PROBLEM	SEVERITY OF TS								
	1			2			3		
	No.	%		No.	%		No.	%	
Males									
0	19	67.8		65	56.0		26	45.6	
1	5	17.9		37	31.9		15	26.3	
2	4	14.3	32.2	14	12.1	44.0	11	19.3	54.4
3	0	0.0		0	0.0		5	8.8	
Total	28	100.0		116	100.0		57	100.0	
Females									
0	2	66.6		18	58.1		9	60.0	
1	1	33.3		7	22.6		2	13.3	
2	0	0.0	33.0	6	19.3	41.9	3	20.0	40.0
3	0	0.0		0	0.0		1	6.7	
Total	3	100.0		31	100.0		15	100.0	

Table 4*B* shows the relationship between discipline problems and the severity of TS in males and females. It was not unexpected that when TS is too mild to treat, the ancillary problems such as difficulty with discipline are also less frequent and that all of the patients with grade 3 discipline problems were also in the highest category, grade 3, of severity of TS. This also gives an internal control in that male patients with grade 1 severity of TS would be closest to a non-TS control, and here 32.2% had discipline problems. Grade 2 was different from grade 1 only in that the tics required treatment, and here 44% had discipline problems. Finally, in grade 3, 54.4% had discipline problems. By chi-square, these are significantly different at $P = .05$.

Anger and Violence

If there was a single word that best characterized the behavioral problems in TS it would be "anger." Many patients were described as having a "short temper" and were "easy to anger." The responses to the question: "Have there been problems with violence?" were divided into three grades. In grade 0, there were no problems. In grade 1, the problems were clearly outside the realm of normal in severity, but did not involve destruction of property or physical attacks on other persons. Some typical responses were: he or she "has many temper tantrums, yells and screams, and is easily upset," "pounds on walls and slams doors," and "has a violent temper."

In grade 2, the expression of anger took the form of destruction of property, killing animals, and injuring other people. Some examples are: "he ripped off the closet doors and knocked holes in the wall," "beats up on his mother constantly," and "burned down his sister's doll house." The expression of anger by physical destruction was seen often enough that one of the questions asked was: "Are there holes in the walls of his or her room?"

In grade 3, the patient's anger and violence resulted in legal problems.

Table 1 summarizes the frequency of the different grades of problems with anger and violence. For all cases, 42% had some problems. In relationship to hyperactivity, it increased from 24% for those without hyperactivity to 47% for those with grade 1 and 59% with grade 2 hyperactivity. Despite being the "gentler" sex, females had as many problems with anger and violence as did males (see table 1).

We believe that there are both intrinsic and extrinsic causes of the anger. The extrinsic causes refer to anger secondary to being unable to control bodily movements, being ridiculed by peers, teachers, and parents, and frustration over concomitant problems such as ADD with learning disorders, with consequent failure in school. However, anger is also intrinsic to the disease. Three observations lead us to this conclusion: (1) Many parents of children who suddenly, over a period of 1 or 2 days, developed tics and vocal noises tell us that their child just as rapidly changed from easy-going to irritable, short-tempered, and angry. In many cases, when the haloperidol reached a dose that controlled the tics, their personality also reverted to a previous pleasant disposition (although this is not always long-lasting). The personality changes were too rapid and too closely synchronized with the motor and vocal tics to be

secondary to long-term problems of coping with the symptoms. (2) Anger and short-temper occurred in some TS patient's even when their symptoms were relatively mild and there was no peer ridicule or associated problems. (3) In animal experiments, dopaminergic agonists elicit not only tics and stereotyped behavior [18], but also enhance aggressive behavior and fighting [19]. (4) The problems with anger are consistent with TS being a disorder of "dysinhibition."

Shapiro et al. [1] reported that in a blind study a group of TS patients did not significantly differ from general *psychiatric outpatients* on factors such as overt and underlying psychosis, obsessive-compulsive traits, inhibition of hostility, hysteria, and general maladaption. They did not believe TS patients were characterized by more aggressivity, impulsive sexuality, self-destructiveness, enuresis, or antisocial behavior. This is in contrast to our observations, and we believe that a comparison to a general psychiatric outpatient population is not the proper control group. Wilson et al. [20] examined the question of behavior disturbance in TS using a Behavior Problem Checklist and other tests. The 21 TS patients studied showed considerably more disturbance than 17 unselected public school children. The TS scores were comparable to those seen in problem children attending special classes. The pattern of the scores was nonspecific and comparable to those across problem-class subtypes of aggressive, hyperactive, and withdrawn children. Until comparison to a suitable control group is made for this series (in progress), we can get a simulated internal control group by proposing that among these 250 patients the group closest to a normal population would be those with grade 1 TS without ADD. There were 21 such patients, and 33% had either grade 1–3 discipline problems and/or grade 1–3 problems with violence. By contrast, among all 250 patients, 61% had grade 1–3 discipline problems and/or grade 1–3 problems with violence. These are significantly different at $P = .01$ (t-test).

It is difficult to obtain a totally neutral control group where the parents are willing to come in and be interviewed for an hour. While there are potential problems with an internal control, there are also some advantages such as being ascertained and interviewed in an identical manner. The internal controls were separated from the rest simply on the basis of the severity of tics and criteria for a diagnosis of ADD. These are independent of problems with discipline and anger or violence.

We see many similarities between some patients with TS and the heterogenous group of conduct disorders of childhood [8, 21]. The overlap between ADD with hyperactivity and conduct disorder is well documented [22]. Patients with both these disorders should be carefully questioned for TS symptoms in themselves and their relatives.

Exhibitionism

Eldridge et al. [3], Nee et al. [16], and Comings and Comings [23] reported the presence of troublesome sexual and aggressive impulses and exhibitionism in TS. Exhibitionism was classified as follows: in grade 0, it was absent; in grade 1, it involved sexual touching (themselves or others); in grade 2, exhibi-

tionism was limited to family members in the home; and in grade 3, there was public exhibitionism. Table 1 gives the distribution of these grades in males and female patients with TS. Some form of exhibitionism was present in 16% of males and 6% of females. In grade 1, a typical comment is: "He is always touching his crotch." In grade 2, one comment was: "I have exhibitionistic urges in the home and go around without pants on. They are so strong that I can't go to the beach because I know I will take my suit off." In grade 3, some comments were: "I often flash by taking my shorts down for a second when playing basketball," and "My husband urinates in the backyard even though the bathroom is only a few feet away."

Enuresis

Later in the course of the series, it became apparent that there was a high frequency of enuresis. Of 77 patients specifically questioned, 28 or 36% had problems with enuresis. This compares with 27% enuresis reported by Jagger et al. [13].

Obsessive-Compulsive Behavior

One of the striking features of TS is the presence of ritualistic actions and obsessive-compulsive behavior. Table 5 lists the ritualistic actions and ritualistic thoughts present in these patients. Needing to touch things "until it felt right," is a common theme. Obsessive thoughts sometimes centered around aggressive, sexual, or violent matters. Touching or counting things often has to be done in units of 2's or 3's. In many families, first-degree relatives of TS patients describe obsessive-compulsive thoughts and actions in the absence of tics or vocal noises, suggesting that the TS gene may be expressed as an obsessive-compulsive personality only [24]. Some obsessive-compulsive be-

TABLE 5

OBSESSIVE-COMPULSIVE SYMPTOMS

Symptom	No. patients
Ritualistic actions:	
Touching things just right	19
Objects placed just right or in right place	20
Touching things exact number of times	5
Washing hands until raw	5
Rechecking things many times	9
Smelling things	4
Other	12
Ritualistic thoughts:	
About violence and torture	15
Counting things (often by units of 2, 3, or 4)	12
Mental echololia or paralalia	10
About sex	9
Spell things forward of backward	3
Other	5

havior was present in 45% of females, 29% of males, and 32% for all. In some series, up to 68% had obsessive-compulsive behavior [13, 16].

Positive vs. Negative Family History

It has frequently been suggested that certain clinical features are more or less common in familial as opposed to sporadic cases [16, 25, 26]. The large number of unselected cases in the present series allowed us to determine if familial cases are different from sporadic ones. Patients with a positive and negative family history were compared for sex, age of onset, years from onset of symptoms to diagnosis, severity of TS, presence of ADD with hyperactivity, being Jewish, coprolalia, exhibitionism, obsessive-compulsive behavior, and problems with discipline or violence. None of these features was significantly different in familial vs. sporadic cases.

School Problems in TS

A major focus of problems in children with TS centers around the school. The ADD contributes to a significant proportion of their difficulties. Prior to diagnosis and treatment, uncontrollable tics and vocal noises may be so distracting that reading and learning is difficult. In occasional cases, the tics may be so severe that it is difficult for TS patients to keep their eyes on the paper. The ridicule of their peers, and sometimes teachers, may further compound their dislike of school. One bright college student was able to identify a number of factors that made school difficult for her. These were: (1) an inability to filter out extraneous noise, (2) compulsive re-reading of the same sentence over and over, (3) inability to settle down to study since everything had to be in just the right place and have the right "feel," (4) feeling so overwhelmed that she could not organize or prioritize her work, (5) panic on timed tests, and (6) racing thoughts entering her head so fast they were gone before adequately grasped.

These problems combine to produce low self-esteem, frequent acting-out, and, in many cases, school phobia. We see the latter both before and after treatment and do not believe it occurs predominantly after treatment with haloperidol, as a side-effect of the medication, as Mikkelsen et al. [27] suggest. It usually disappears when the parents learn the importance of not supporting the child's fears by allowing him or her to stay home.

Treatment

Although clonidine can be useful in some cases [28], in our experience, haloperidol [1, 2] is still the drug of choice, and we find it is more effective in children than in adults. However, it is very important to start at low doses and increase it gradually. We routinely begin the patient at 0.5 mg each evening (occasionally 0.25 mg for young children) for 1 week and then increase by 0.5 mg at weekly intervals. This is continued until there is either a 70%–90% reduction in symptoms or occurrence of side effects that cannot be eliminated with benztropine 0.5–2 mg tid. Our goal is 70%–90% suppression of the tics, not 100%. The advantage of this is that if tics disappear completely for several weeks, the dose is then decreased by 0.5 mg every 2 weeks until they return. If

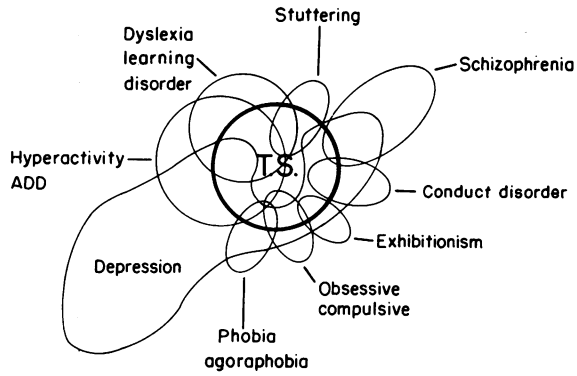

FIG. 2.—Interrelationships between various neuropsychiatric disorders and TS. *Central circle* represents those in the population with the TS gene. *Other circles* indicate that a portion of individuals with the given diagnosis will have the problem due to the presence of a TS gene.

the patient has gone into a prolonged remission, the medication can be terminated. If 100% suppression of the tics was the end point, there would be more side effects and then no cue for decreasing the dose. Pimozide, a haloperidol-like drug, is now available in the United States. For a discussion of the advantages and disadvantages of this drug in the treatment of TS, see Shapiro et al. [29]. The role of stimulants in the treatment of ADD in TS is discussed in detail elsewhere [8].

For the complete treatment of TS, it must be thought of as a family disorder since no one in the household is unaffected. One of the greatest problems comes from excusing misbehavior (but not the tics and vocal noises) because the child has TS. If anything, these children require a much more structured environment than other children, with immediate, nonphysical consequences for inappropriate behavior.

In summary, Tourette syndrome is a common genetic disorder with 1% of the population estimated to be gene carriers and 50% of these expressing the gene as either TS or motor or vocal tics [6]. While approximately half of our cases of TS had predominately tics and vocal noises, the other half had additional problems including attention-deficit disorder with hyperactivity, learning disorders, and varying degrees of behavioral problems or conduct disorder. These interrelationships are illustrated in figure 2. Whenever a child presents with these disorders, the parents should be questioned about the presence of tics or vocal noises in the patient and other family members.

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Annual Meeting
American Society of Human Genetics
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