

to be poorer (Robertson, 1989). Golden (1984) reviewed the neuropsychological aspects of the syndrome and found that there was a specific problem with arithmetic, which had previously been identified in studies by Joschko & Rourke (1982) and Incagnoli & Kane (1982). It should be noted, however, that the numbers of cases in these studies were small, and that this area remains to be fully investigated.

There is evidence that such cognitive and, perhaps more surprisingly, behavioural disturbances can, as in this case, persist after the clinical syndrome has virtually disappeared (Leckman & Cohen, 1983).

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Tourette's Syndrome in New Zealand

A Postal Survey

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It has become increasingly evident that Tourette's syndrome (TS) is not as rare as was once thought and substantial cohorts from various parts of the world have been reported. The clinical characteristics seem independent of culture as they appear to occur with some degree of uniformity irrespective of the country of origin. We investigated the point prevalence and report the clinical characteristics of TS in New Zealand. Forty probable cases were identified and the symptoms were similar to those described in cohorts from other parts of the world.

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Tourette's syndrome (TS) is characterised by multiple motor and one or more vocal tics which usually occur

many times a day and must be present for a period of more than one year. Classically, the anatomical location, number, frequency, complexity and severity of the tics change over time, and the age of onset is usually before 21 years (DSM-III-R; American Psychiatric Association, 1987).

The most common motor symptoms are blinking of the eyes, head shaking and shoulder shrugging, while sniffing, coughing and clearing of the throat are common vocalisations. Obsessive thoughts and compulsive behaviour are now being recognised as important parts of the disorder, as are coprophenomena (inappropriate swearing and making of obscene gestures) and echophenomena (copying behaviours), while other associated, but less common

behaviours include self-injurious behaviour (SIB), aggression to people or property, attention and concentration problems, hyperactivity and learning difficulties. It is usually these behavioural symptoms which prompt a request for medical help, when the diagnosis may be made.

The characteristics of TS seem to be largely independent of culture, as the phenomenology in the various countries where large cohorts have been investigated has some degree of uniformity (for a review see Robertson, 1989). This cross-cultural similarity emphasises the biological factor in the aetiology of the syndrome, which is thought to be largely genetic and due to an autosomal dominant gene with varying penetrance (Robertson, 1989).

TS is no longer the rarity it was once thought to be, as evidenced by the documentation of substantial cohorts from many countries including the USA, UK, Soviet Union, Japan, China, Netherlands and Denmark (Robertson, 1989). However, in some parts of the world there may be under-reporting of individuals, which could perpetuate the belief that TS is extremely rare. This may be the case in New Zealand (NZ), where only three patients had been documented (Shenken, 1980, 1984) before we carried out this survey.

The NZ government's 1988 population estimate is 3 347 300. If it is assumed that the prevalence of TS in NZ is similar to that in the USA and Europe, where there is a wide variation (estimates varying from 0.005 per 1000 (Lucas *et al*, 1982) to 0.50 per 1000 (Bruun, 1984)), then between 17 and 1674 cases of TS would be expected. We anticipated that there may be an untapped cohort of TS sufferers in NZ. The purpose of the present study was to find as many cases as possible and to document their clinical features.

Method

A postal survey was undertaken to determine whether the prevalence of TS in NZ might be similar to reported rates in other countries and to compare the clinical characteristics of the patients identified with those in other documented cohorts.

Data were collected by two of the authors (MV, MM), while on medical student electives in NZ. Because the study period was limited to the eight weeks of an elective, a postal survey was considered to be the most effective form of data collection.

Many TS sufferers are referred to neurology or psychiatry clinics with isolated complaints, and only then are the syndromic features of TS recognised. However, since TS tends to have its onset in childhood, and many of the symptoms of the disorder are most visible before and during teenage years, we expected many cases also to be seen and managed by paediatricians. A list of psychiatrists,

neurologists and paediatricians was compiled from the NZ Medical Register.

Each doctor on the list received a personal letter explaining our study, information about TS, a questionnaire asking if he/she knew of any cases, two copies of an assessment sheet for documentation of clinical features, and a stamped addressed envelope for replies. When cases were identified, the doctors were asked to complete the assessment sheet. An additional source of information was a self-help group of parents of afflicted children – the Tourette's Syndrome Support, NZ (TSS, NZ).

The assessment sheet included the most commonly seen features of TS, including motor and vocal tics, obsessive-compulsive behaviour, childhood hyperactivity, SIB, sleep disturbances and aggression, and could elicit as many features as were necessary for a probable diagnosis to be made. The assessment sheet included enough personal data to allow identification of duplicate records. Replies were sent to one of the authors (BJ) at the Department of Health in Wellington, NZ.

The NZ population data are based on the NZ government department of statistics estimates, published in the 1990 NZ yearbook (Department of Statistics, 1990). School-age children are taken to be the number enrolled in primary and secondary education in 1988.

Results

A total of 291 letters were sent out to doctors (166 psychiatrists, 99 paediatricians, 26 neurologists). The response rates were: psychiatrists, 85 (51.2%); paediatricians, 58 (58.6%); neurologists, 12 (46.2%); total, 155 (53.3%).

Twenty-three cases of TS fulfilling DSM-III-R criteria were identified from these replies and an additional 14 individuals from TSS, NZ were recognised. Three of the cases from TSS, NZ were duplicates of confirmed cases notified by psychiatrists. Excluding these three individuals, there were 34 TS cases (psychiatrists, 13; paediatricians, 2; neurologists, 8; TSS, NZ, 11). There were another six individuals in whom the diagnosis was suspected but not confirmed by the notifying doctor (paediatricians, 5; neurologists, 1). A total of 40 possible patients were thus identified.

In all the six suspected cases, assessment sheets were completed which supported a TS diagnosis. Although attempts were made to complete assessment sheets in all the 34 cases originally identified, it was possible in only 18 cases, making a total of 24 cases in whom assessment sheets were completed (psychiatrists, 8; paediatricians, 7; neurologists, 1; TSS, NZ, 8). More than half of the cases were identified by psychiatrists and neurologists, whom we had expected to be most familiar with the syndrome. However, in the absence of direct clinical interviews, and because assessment sheets were completed in only 24 cases, the authors refer to the whole group of 40 individuals as probable cases of TS.

The mean age of these cases was 16 years 0 months. The mean age at onset of TS was eight years seven months, and the mean duration of symptoms was seven years five months. The male to female ratio for those individuals where gender was known was 4:1.

Discussion

While the data presented here are limited owing to the restrictions of our study, they probably form a substantial sample of TS cases in NZ. Given the method of ascertainment, the short period of data collection, the modest response rate to the survey (overall 53%), and the lack of reminder letters, the 40 probable TS sufferers may underestimate the number of affected individuals in NZ. The study is further limited by the fact that general practitioners were not included: however, in our experience in the UK and USA, the majority of general practitioners are not well acquainted with TS.

Table 1
Symptom totals from assessment sheets (24 completed)

	<i>n</i>	% of cases
Motor tics		
excessive eye-blinking	22	85
head tics	20	77
neck movements	12	46
opening of mouth	12	46
shoulder shrugging	11	42
whole body jerking	10	38
rolling eyes	10	38
interrupted walking	7	27
Vocalisations		
grunting	17	65
throat clearing	15	58
excessive sniffing	14	54
coprolalia	11	42
excessive coughing	7	27
Associated phenomena		
unable to attend or concentrate	16	62
impulsive actions and decisions	12	46
hyperactive as child	10	38
sleep talking	9	35
forced to touch	9	35
excessive aggression	8	31
obsessive or recurrent thoughts	7	27
nightmares	7	27
stammered/stuttered	5	19
copying behaviours (echophenomena)	5	19
obsessional tidiness	4	15
SIB	4	15
compulsive rituals	3	12
need to count things	3	12
panic attacks	3	12
obsession with numbers	2	8
sleep walking	2	8
bed wetting	2	8
night terrors	1	4
Phobias		
crowded places	5	19
heights	4	15
closed spaces	4	15
the dark	4	15
spiders/insects	4	15

All but two of the cases we identified were between the ages of 10 and 20 years. We have therefore estimated the prevalence in that age range. There were 1 090 500 people below the age of 20 in NZ in 1990. If it is assumed that half of them are between the ages of 10 and 20, the point prevalence estimated from the 38 probable patients between 10 and 20 is 0.7 per 10 000.

A possible source of bias in the identification of cases is the fact that we know that the majority had been seen by a doctor or other health care professional. From our experience with the UK and USA Tourette Associations, it is likely that all the patients from the TSS, NZ had also seen a doctor. Several studies have shown that only 25–50% of TS sufferers seek professional help (Pauls *et al*, 1990; Robertson & Gourdie, 1990). It is therefore possible that, by targeting doctors, we have missed more than half of the NZ cases, and therefore have underestimated the prevalence. Despite this, the estimated point prevalence of TS that we have found from our data is within the range we predicted at the outset of our study, that is 17–1674 (Lucas *et al*, 1982; Bruun, 1984).

The clinical features of the 24 cases that we were able to document fully are shown in Table 1. They are similar to cohorts of TS patients in other parts of the world (see Table 2). This cross-cultural

Table 2
Summary of findings of previous studies

Reference	Number of patients with TS	Mean age of onset: years	% with coprolalia	% with obsessive-compulsive behaviour ¹
Wassman <i>et al</i> (1978)	15	5.0	53	NS ²
Eldridge <i>et al</i> (1977)	21	7.7	NS	NS
Montgomery <i>et al</i> (1982)	15	7.3	20	67
Nee <i>et al</i> (1980)	50	7.0	58	68
Shapiro <i>et al</i> (1978)	145	7.2	69	–
Moldofsky <i>et al</i> (1974)	15	7.3	60	NS
Lucas <i>et al</i> (1982)	25	8.0	NS	NS
Comings & Comings (1985)	250	6.9	33	32
Steff (1984)	431	NS	36	74
Robertson <i>et al</i> (1988)	90	7.0	33	37
Present study	24	8.7	46	50

1. Thoughts also included.
2. NS = Not stated.

similarity provides further support for the suggestion that the aetiology of the disorder is culture independent and likely to be biological (Robertson & Trimble, 1991).

We conclude that there are a substantial number of TS sufferers in NZ and that the symptoms reported for TS in NZ is similar to that reported in other clinic samples around the world. The prevalence we have calculated is of the same order as in previous studies. Additional work might identify more affected individuals in NZ, and provide a better estimate of prevalence.

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