Tourette Syndrome and Tic Disorders in a Swedish School Population

Prevalence, Clinical Assessment, Background, Psychopathology, and Cognitive Function

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Dissertation presented at Uppsala University to be publicly examined in Auditoriet, Gustavianum, 753 10 Uppsala, Friday, March 10, 2006 at 09:15 for the degree of Doctor of Philosophy (Faculty of Medicine). The examination will be conducted in Swedish.

Abstract

A total population of 4,479 children (7-15 years of age) attended school in Ludvika & Smedjebacken in 2000. All the school children and their parents were asked to fill in a questionnaire concerning different tics. A three-stage procedure was used: tic identification, interview, and clinical assessment.

Tourette syndrome, according to DSM IV criteria was found in 25 (0.6%) of the children, another 34 (0.8%) suffered from chronic motor tics (CMT), 24 (0.4%) from chronic vocal tics (CVT) and 214 (4.8%) children had had transient tics (TT) during the last year. Altogether 297 (6.6%) children had or had had some tic disorder.

Twenty-five controls without tics and 25 children with TT of the same age, sex and school as the TS children were randomly chosen. They were together with the 34 children with CMT and the 24 children with CVT examined with use of a broad battery of instruments.

The mean age of the first symptoms of TS was significantly lower than the onset of chronic motor/vocal tics. A younger age of onset of TS indicated more severe tics. Eighty per cent had a first-degree relative with a psychiatric disorder such as tic disorder, obsessive-compulsive behaviour, attentiondeficit/hyperactivity disorder (ADHD), or depression. A non-significant increase with regard to reduced optimality score in the pre-, peri-, or neonatal periods was found in children with TS compared to controls. No differences were found concerning socio-economic status. Psychiatric comorbid disorders were found in 92% of the children with TS. ADHD was most common. Patterns of psychiatric comorbidity were similar in children with TS and CVT. Children with TS perform poorer than the population in general with respect to cognitive functioning and self-perception.

The results are discussed as they relate to the need for case identification, diagnosis, intervention, and treatment.

Keywords: Tourette syndrome, tic disorders, population study, psychopathology, heredity, perinatal complications, cognitive function

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To the memory of my father

&

To the real sunshines of my life

Shirin, Jamen, Joan
List of publications


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<th>Abbreviation</th>
<th>Full Form</th>
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<tbody>
<tr>
<td>ADHD</td>
<td>Attention Deficit Hyperactivity Disorder</td>
</tr>
<tr>
<td>APA</td>
<td>American Psychiatric Association</td>
</tr>
<tr>
<td>ASSQ</td>
<td>Asperger Syndrome Screening Questionnaire</td>
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<tr>
<td>CBCL</td>
<td>Child Behavior Checklist</td>
</tr>
<tr>
<td>CD</td>
<td>Conduct Disorder</td>
</tr>
<tr>
<td>CDI</td>
<td>Children’s Depression Inventory</td>
</tr>
<tr>
<td>CMT</td>
<td>Chronic Motor Tics</td>
</tr>
<tr>
<td>CMVT</td>
<td>Chronic Motor/Vocal Tics</td>
</tr>
<tr>
<td>CT</td>
<td>Chronic Tics</td>
</tr>
<tr>
<td>CVT</td>
<td>Chronic Vocal Tics</td>
</tr>
<tr>
<td>CY-BOCS</td>
<td>Children’s Yale-Brown Obsessive Compulsive Scale</td>
</tr>
<tr>
<td>DCD</td>
<td>Developmental Coordination Disorder</td>
</tr>
<tr>
<td>DSM-III</td>
<td>Diagnostic and Statistical Manual of Mental Disorders. Third edition</td>
</tr>
<tr>
<td>DSM-IV</td>
<td>Diagnostic and Statistical Manual of Mental Disorders. Fourth edition</td>
</tr>
<tr>
<td>ICD-10</td>
<td>International Classification of Diseases. Tenth edition</td>
</tr>
<tr>
<td>LD</td>
<td>Learning Disorders</td>
</tr>
<tr>
<td>MR</td>
<td>Mental Retardation</td>
</tr>
<tr>
<td>NEPSY</td>
<td>NEPSY A Developmental Neuropsychological Assessment</td>
</tr>
<tr>
<td>OCB</td>
<td>Obsessive Compulsive Behaviour</td>
</tr>
<tr>
<td>OCD</td>
<td>Obsessive Compulsive Disorder</td>
</tr>
<tr>
<td>ODD</td>
<td>Oppositional Defiant Disorder</td>
</tr>
<tr>
<td>PDD</td>
<td>Pervasive Developmental Disorder</td>
</tr>
<tr>
<td>RCFT</td>
<td>Rey Complex Figure Test</td>
</tr>
<tr>
<td>TS</td>
<td>Tourette Syndrome</td>
</tr>
<tr>
<td>TSSS</td>
<td>Tourette Syndrome Severity Scale</td>
</tr>
<tr>
<td>TT</td>
<td>Transient Tics</td>
</tr>
<tr>
<td>WIPPSI</td>
<td>Wechsler Pre-school and Primary Scale of Intelligence for Children</td>
</tr>
<tr>
<td>WISC-III</td>
<td>Wechsler Intelligence Scale for Children. Third edition</td>
</tr>
</tbody>
</table>
Introduction

Background

Gilles de la Tourette’s syndrome (TS) is a childhood onset inherited neuropsychiatric disorder, characterised by the presence of both multiple motor tics and one or more vocal tics, which last longer than a year. The generally accepted research definition being taken from DSM-IV. Children with TS often exhibit a variety of behavioural symptoms, particularly attention deficit hyperactivity disorder (ADHD), learning problems and obsessive compulsive disorder (OCD).

TS is characterized by marked fluctuations in severity and frequency during the disorder course, and has a wide variation among patients. The disorder typically begins between 3-7 years of age and half of the patients are tic-free by 18 years. Prevalence rates have been estimated to vary between 0.5-3% in a school population. Evidence indicates a developmental disorder of synaptic neurotransmission as the underlying etiology.

To date a TS gene has not been isolated. Environmental factors may play an important role in the expression of tics, and a poststreptococcal autoimmune cause has been proposed, but is unproven. However, a prefrontal dopaminergic abnormality is suggested. Brain imaging, neuropsychological and post mortem studies support involvement of cortico-striatal-thalamocortical pathways, but definitive pathophysiological mechanism or neurotransmitter abnormality is unknown.

The diagnosis of TS is based on a history and an observation of tics. The first step in the management is education of the patients, family members and teachers. Medication is considered when symptoms are functionally disabling. Classes of medication used in the treatment of patients with TS include neuroleptics, central nervous system (CNS) stimulants, and selective serotonin re-uptake inhibitors (SSRI).

History

The first medical description of TS was in 1825, when Itard reported the case of a French nobliewoman, the Marquise de Dampierre. She developed symptoms at the age of seven and because of her socially unacceptable utterances was forced to live as a recluse until she died at the age of 85 (1, 2).
Sixty years later, in 1885, Georges Gilles de la Tourette, the French neurologist and student of Charcot at the Salpêtrière Hospital in Paris, described Itard's original case and added eight more cases of TS emphasising the triad of multiple tics, coprolalia (swearing) and echolalia (meaningless repetition), which gained him eponymous fame (3). Gilles de la Tourette considered the disorder, that he described, to be hereditary. There have been translations of some of these case reports and the history of Tourette syndrome (4, 5).

From 1900 to 1965 the TS literature was predominantly psychoanalytical. Notions on cause included repressed sexual desires and a moral conflict between ego and superego. When the beneficial effects of neuroleptic drugs on the symptoms of the syndrome began to be recognized, the observation helped to refocus attention from psychogenic causes to primarily organic etiologies involving a dysfunction of the central nervous system (6).

The word tic or tique appeared in the French language to describe an unsightly muscular caprice. The term was first used to describe certain trick movements in horses in 1665 (7). Then it was used to refer to distasteful motor acts in humans, but in 1756 the term tic douloureux was coined (trigeminal neuralgia in today's lexicon). In his elaborated classification of tonic spasms and cramps, the eighteenth century Montpellier physician Boissier de Sauvages considered 19 varieties of tics (also including trismus, bruxism, and tic douloureux). The term “hypochondriac tics” used to describe some facial movements that resemble what we now term tics. The term convulsive tic, as used by Charcot and Gilles de la Tourette, connotes the “abruptness and momentariness” of the abnormal movement. Until the end of the 19th century movement disorders, referred to as motor incoordinations, were diagnosed as chorea (chorea=Greek for “dance”) (8, 9).

Many notable historical figures have been thought to be afflicted with TS. Among them are Claudius, the third Roman Emperor, who manifested tics and stuttering as well as compulsive behavioural traits. Wolfgang Amadeus Mozart is also a possible TS person. According to the memoirs of various people that met Mozart, he had various motor and vocal tics. Scatology was, furthermore, abundant in his letters (10). Dr Samuel Johnson was also thought to suffer from TS and severe obsessive compulsive disorder (OCD). According to contemporary descriptions, Dr Johnson performed complex gestures when crossing a threshold, involuntarily touched specific objects, and felt impelled to measure his footsteps (11).

Definition and clinical characteristics

Tics, which represent the clinical hallmark of Tourette syndrome, are sudden, repetitive stereotyped motor movements or phonic productions that involve discrete muscle groups (Table 1). Tics may be seen as fragments of normal motor action or vocal productions that are misplaced in context and
that can be easily mimicked and at times confused with goal directed behaviour (12).

Clinicians characterize tics by their anatomical location, number, frequency, intensity, and complexity. Many patients with tic disorders report the presence of associated sensory phenomena including premonitory urges that incessantly prompt tics (13, 14). The age of the onset of TS symptoms starts at age two and cases with an onset at 18 years have been reported. The mean age of onset is 7 years of age, and symptoms usually begin with motor tics. The symptoms fluctuate markedly in severity and frequency over time, and vary from one individual to another.

Coprolalia (the inappropriate and involuntary uttering of obscenities) occurs in less than one third of the clinical population (15). One suggestion is that this may be culturally determined, as only 4% have true coprolalia in Japan (16) and in UK the percentage is 43% (17). Although TS was originally proposed to be a lifelong disorder, many individuals have spontaneous remission or marked improvement without use of medication. In a study of 58 adolescents, tics virtually disappeared in 26%, diminished considerably in 46%, remained stable in 14%, and increased in 14% (18). Another study has confirmed a decline in tic severity and decrease in comorbidity over the time (19).

**Table 1. Motor and vocal tics**

<table>
<thead>
<tr>
<th>Motor tics</th>
<th>Vocal tics</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Simple</strong></td>
<td><strong>Simple</strong></td>
</tr>
<tr>
<td>Eye-blinking</td>
<td>Clearing throat</td>
</tr>
<tr>
<td>Head-jerking</td>
<td>Sniffing</td>
</tr>
<tr>
<td>Grimacing</td>
<td>Coughing</td>
</tr>
<tr>
<td>Lip-pouting</td>
<td>Whistling</td>
</tr>
<tr>
<td>Shoulder-shrugging</td>
<td>Hissing</td>
</tr>
<tr>
<td>Abdominal tensing</td>
<td>Grunting</td>
</tr>
<tr>
<td>Frowning</td>
<td>Tongue clicking</td>
</tr>
<tr>
<td>Limb-jerking</td>
<td>Animal sounds and other noises</td>
</tr>
<tr>
<td><strong>Complex</strong></td>
<td><strong>Complex</strong></td>
</tr>
<tr>
<td>Hopping, clapping, squatting</td>
<td>Repeating words or phrases</td>
</tr>
<tr>
<td>Touching objects or other people</td>
<td>Unusual rhythms, tone or volume</td>
</tr>
<tr>
<td>Kissing</td>
<td>Mimicking accents</td>
</tr>
<tr>
<td>Picking at clothing</td>
<td>Echolalia (repeating the sound or words of another person)</td>
</tr>
<tr>
<td>Self–injurious</td>
<td>Palilalia (repeating one’s own sound or words)</td>
</tr>
<tr>
<td>Biting oneself</td>
<td>Coprolalia (inappropriate uttering of obscenities)</td>
</tr>
<tr>
<td>Copropraxia (inappropriate making of obscenence gestures)</td>
<td></td>
</tr>
<tr>
<td>Echopraxia (imitating other people’s gestures or movements)</td>
<td></td>
</tr>
</tbody>
</table>
Diagnostic criteria for Tourette syndrome and other tic disorders

TS is one disorder in a spectrum of disorders, ranging from a mild transient form to TS that has tics as its main feature. Tic disorders can be divided into two main categories according to their duration: transient (present for less than 12 months) and chronic (present for more than 12 months). Transient tics (TT) is the mildest and most common form. The diagnoses of TS and tic disorders are based on a history and observation of tics, often supported by the presence of coexisting behavioural disorders and a family history of similar symptoms. There is no diagnostic laboratory test.

Today the diagnosis of TS and other tic disorders is spelled out in the DSM-IV (20) (Table 2) and ICD-10 (21) (Table 3). The diagnostic criteria for diagnosing TS in DSM changed with time. In the DSM-IV current diagnostic criteria, the onset must be before the age of 18 years. In DSM-III the onset must be before the age of 21 years (22) (Table 4). In ICD-10 and DSM-III no requirement for the level of severity nor it is necessary to exclude specific other causes in order to make a diagnosis according to these manuals. It seems feasible to require the symptoms to be an obstacle in daily life.
Table 2. DSM-IV criteria for tic disorders and Tourette syndrome

**Transient Tic Disorder**
A. Single or multiple motor and/or vocal tics (i.e., sudden, rapid, recurrent, non-rhythmic, stereotyped motor movements, or vocalizations).
B. The tics occur many times a day, nearly every day for at least four weeks, but for no longer than 12 consecutive months.
C. The disturbance causes marked distress or significant impairment in social, occupational, or other important areas of functioning.
D. The onset is before the age 18 years.
E. The disturbance is not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington’s disease or postviral encephalitis).
F. Criteria have never been met for Tourette’s syndrome or chronic motor or vocal tic disorder.

**Chronic Motor or Vocal Tic Disorder**
A. Single or multiple motor or vocal tics (i.e., sudden, rapid, recurrent, non-rhythmic, stereotyped motor movements, or vocalizations), but not both, have been present at some time during the illness.
B. The tics occur many times a day nearly every day or intermittently throughout a period of more than one year, and during this period there was never a tic-free period of more than three consecutive months.
C. Same as above.
D. Same as above.
E. Same as above.
F. Criteria have never been met for Tourette syndrome.

**Tourette Syndrome**
A. Both multiple motor and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently (a tic is a sudden, rapid, recurrent, non-rhythmic, stereotyped motor movement, or vocalization).
B. The tics occur many times a day (usually in bouts) nearly every day or intermittently throughout a period of more than one year, and during this period there was never a tic-free period of more than three consecutive months.
C. Same as above.
D. Same as above.
E. Same as above.

Table 3. ICD-10 criteria for Gilles de la Tourette syndrome
A. Multiple motor tics and one or more vocal tics that have been present at some time during the disorder, but not necessarily concurrently.
B. The frequency of tics must be many times a day, nearly every day for more than one year, with no period of remission during that year lasting longer than two months.
C. Onset before 18 years of age.

Table 4. DSM-III criteria for Tourette syndrome
A. Both multiple motor and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently.
B. The tics occur many times a day (usually in bouts) nearly every day or intermittently throughout a period of more than one year.
C. Location, number, frequency, complexity and severity of tics change over time.
D. Onset before age 21.
E. Not exclusively occurring during intoxication with psychoactive substance or CNS disease, such as Huntington disease or postviral encephalitis.
Epidemiology

TS and other tic disorders have been reported in all races, ethnic groups and socio-economic classes. The prevalence of tic disorders varies widely in published reports. Discrepancies can be explained by differences in distribution, diagnostic criteria, inclusion thresholds, varied study designs, and methodologies. Simple motor and vocal tics are very common in all school-age children, occurring in 4–24% of them (3, 23-24, 27). Chronic tic disorders are believed to affect 1-3% of the population and many actually represent a mild form of TS (23, 25-27) The prevalence of Tourette syndrome varies in different studies, with a range of 0.05-3% (23, 25-35). TS is predominantly a disorder among males. The male to female ratio is 1.6–10:1 (26, 30-31, 33-35). In studies of children with special educational needs, in particular, the prevalence of TS has been very high (30%), (26, 36) (Table 5 shows the prevalence estimates).

Table 5. Prevalence of TS in school populations

<table>
<thead>
<tr>
<th>Author/Year</th>
<th>Age (years)</th>
<th>N</th>
<th>Informant</th>
<th>Diagnostic Criteria</th>
<th>Prevalence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Comings et al.</td>
<td>5-13</td>
<td>3,034</td>
<td>Observation + teacher/parent interview + examination</td>
<td>DSM-III</td>
<td>0.6</td>
</tr>
<tr>
<td>Nomoto et al.</td>
<td>4-12</td>
<td>1,218</td>
<td>Parent Q + tel. interview</td>
<td>DSM-III</td>
<td>0.5</td>
</tr>
<tr>
<td>Mason et al.</td>
<td>13-14</td>
<td>166</td>
<td>Self report Q + parent/teacher Q + observation</td>
<td>DSM-III</td>
<td>2.9</td>
</tr>
<tr>
<td>Kadesjö &amp; Gillberg.</td>
<td>7</td>
<td>435</td>
<td>Parent/teacher Q + clinical examination</td>
<td>DSM-IV</td>
<td>1.1</td>
</tr>
<tr>
<td>Hornsey et al.</td>
<td>13-14</td>
<td>918</td>
<td>Self-report, parent/teacher Q + two interviews</td>
<td>DSM-III</td>
<td>0.8</td>
</tr>
<tr>
<td>Kurlan et al.</td>
<td>8-17</td>
<td>1,596</td>
<td>Parent/teacher interview + observation</td>
<td>DSM-IV</td>
<td>0.8-1.5</td>
</tr>
<tr>
<td>Wang et al.</td>
<td>6-12</td>
<td>2,000</td>
<td>Self report Q + clinical interview</td>
<td>DSM-IV</td>
<td>0.6</td>
</tr>
<tr>
<td>Lanzi et al.</td>
<td>6-11</td>
<td>2,347</td>
<td>Classroom observation</td>
<td>DSM-IV</td>
<td>0.7</td>
</tr>
</tbody>
</table>

Q= questionnaire
Aetiology

Various biochemical, imaging, neurophysiologic, and genetic studies support the notion that TS is an inherited, developmental disorder of synaptic neurotransmission resulting in disinhibition of the cortico-striatal-thalamic-cortical circuity (37). The basal ganglia, particularly the caudate nucleus, and inferior prefrontal cortex, have been implicated in the pathogenesis of TS, as well as OCD and ADHD (6). The distribution of classic neurotransmitters within these circuits raises the possibility that a variety of transmitters, including dopamine, serotonin, glutamate and GABA, may be involved in the pathobiology of TS (38).

To date, the TS gene has not been isolated (6, 39). Family and twin studies support a role for genetic factors in the transmission and expression of TS (40-42). Comorbidity with OCD and ADHD are common among patients with TS. If OCD, ADHD and panic attacks are included, the rate of familiar transmission is 15-82% (40-51). In 1986, Pauls and Leckman (40) used segregation analysis of affected families to indicate that TS is inherited in an autosomal dominant pattern with a variable expression that also includes chronic tic disorder and OCD.

The role of adverse pre- and perinatal events in the pathogenesis of tic disorders has been considered since 1956, when Pasamanick and Kawi (52) found that mothers of children with tics were 1.5 times more likely to have experienced a complication during pregnancy than the mothers of children without tics. The severity of maternal life stress during pregnancy, severe nausea and/or vomiting during the first trimester, premature low birthweight children, low Apgar scores and more frequent maternal prenatal visits suggested to be risk factors for developing Tourette syndrome (53-54).

Recently there have been suggestions that a variety of other factors are also involved in the aetiopathogenesis of TS, including gender- and stress-related hormonal factors (55-56) and various bacterial and viral infections. In the USA Swedo et al. (57) described a group of 50 children with OCD and tic disorders, whom they designated as having paediatric autoimmune neuropsychiatric disorders, associated with streptococcus (PANDAS) with evidence of group A B-haemolytic streptococcal (GABHS) infections. The diagnostic criteria included presence of OCD and/or a tic disorder, prepubertal symptom onset (usually acute and dramatic), association with GABHS infections, episodic course of symptom severity, and association with neurological abnormalities.

The idea of PANDAS is still speculative and only a few cases have been described which support the hypothesis. Kurlan (58) suggests that further research is required to establish more clearly the role of post-infectious and immune-mediated mechanisms in TS. He also suggests that with the present state of knowledge, testing and treating of patients should not form part of a
routine clinical work-up and should only be used in the context of research protocols.

Comorbidity

A number of clinically and population based studies have reported a high rate of comorbidity in tic disorders (17, 23, 30, 31, 33, 59-68). The most frequent ones have been ADHD, learning disability, obsessive-compulsive disorder (OCD), pervasive developmental disorder (PDD), depression and self-injurious behaviours. Several studies have shown that learning difficulties are common in children with TS and that tics are more common in children attending special education classes (36, 43, 60, 66).

The children with TS are less popular among friends and more withdrawn. They have low self-perception and the TS has been linked to a range of everyday problems suggestive of social inappropriateness and getting into trouble with the law (69-70). Individuals with TS have higher scores on externalizing and internalizing behaviours and a high risk for a variety of social and emotional difficulties (71, 72).

Neuropsychological function

Cognitive disturbances, especially low IQs have been reported in children with TS (73), but impairment occurs in only a portion of them. Mostly it is associated with comorbidity for ADHD, OCD and learning disability (3, 48, 74-77). Results differ concerning discrepancies between verbal IQ (VIQ) and Performance IQ (PIQ) in individuals with TS. Several studies have found that individuals with TS have significantly lower PIQ than VIQ scores suggesting a greater difficulty on visuomotor and visuoperceptual than on verbal tasks, with as many as 30-40% of patients with TS having significantly lower PIQ than VIQ (3, 78). However, Bornstein (79) did not find a significant PIQ-VIQ discrepancy in a large study of 100 children with TS.

According to Como (80) executive function (EF) deficits are common among TS patients and some cases can be more debilitating than the tics characterising the syndrome. To date studies of EF ability in TS reveal puzzlingly inconsistent findings. Some authors have found evidence of EF deficits (82, 83), although there are some negative findings (81, 84). Several authors suggest that executive dysfunctions in TS are typically associated with comorbid disorders, especially ADHD and OCD (83, 85, 86-88). Some studies have reported significant associations between tic severity, behavioural problems and inversely related neuropsychological function, but these findings are inconsistent (79, 87).
Treatment of tics

The decision to treat a patient should be made from the findings of an initial comprehensive assessment that includes analysis of tics, documentation of comorbid disorders, assessment of severity, as well as the resulting impairment. The treatment of tics and any comorbidities should be prioritised according to the impairment caused by each problem. The first step of a treatment is a proper education of the patient, family, teachers, and other persons who interact with the patient.

Medications are usually considered when symptoms begin to interfere with peer relationships, social interaction, academic or job performance, or activities of daily life (89). There is no cure for tics and all pharmacotherapy must be regarded as symptomatic therapy. If a tic-suppressing drug is indicated, a two-tiered approach is recommended: firstly, non–neuroleptic drugs for mild tics, and secondly, typical or atypical neuroleptics for more severe tics. The goal of the treatment is not a complete suppression of all motor and phonic tics, but to reduce them such that they no longer cause substantial psychosocial disturbance (89, 90).

Important issues in research of TS and tics

There are at least four perspectives that need to be considered when discussing the importance of these studies.

1- Research: There has been considerable variation in reported prevalence rates for different studies. Discrepancies can be explained by differences in diagnostic criteria and inclusion thresholds, and varied study designs and methodologies. It is now recognized that TS is not uncommon. Although clinically a heterogeneous disorder with high rates of comorbid condition, ADHD and obsessive compulsive behaviours (OCB) are most common. To my knowledge no Swedish comprehensive, population-based study of school children with TS and other tic disorders has been published with respect to the backgrounds, clinical characteristics and comorbidity. In addition there is no study that has examined cognitive and emotional performance in population samples. This would have major implications for future studies of these conditions.

2- Health care: Knowledge with respect to the prevalence of TS and different tic disorders, clinical course, background factor, psychopathology and cognitive function is critical for planning and early support in health care.
3- **School**: To identify appropriate screening methods for caretaking of children with TS, which generally affects adjustment and school achievement, is essential for planning and early support in education and school health care.

4- **Family**: Parents often feel helpless and at a loss to know what to do when their children have tics. Helping parents both to adjust the diagnosis and to manage the negative reaction of peers and the public can be empowering to families. The parents shall have the right help and guidance in order to be able to treat their children adequately.

**Ethics**

All children and their parents gave their informed consent according to the Declaration of Helsinki. The study was approved by the Ethics Committee of the Medical Faculty of Uppsala University.
Aims of the present study

1- To estimate the prevalence of Tourette syndrome and other tic disorders in elementary school children 7-15 years old. (Study I)

2- To describe the symptoms and tic severity in Tourette syndrome and other tic disorders, and age of onset in a total population of children. (Study II-IV)

3- To study the familial loading of mental disorders in first-degree relatives to children with Tourette syndrome and other tic disorders. (Study II)

4- To evaluate the role of adverse pre- and perinatal events in the development of Tourette syndrome, and to compare the socio-economic status between the groups. (Study II)

5- To describe the psychiatric comorbidity of Tourette syndrome and other tic disorders found in a general population of children 7-15 years old. (Study III)

6- To evaluate if children with TS have any specific cognitive deficits or low self-perception. (Study IV)
Material

Study population and identified cases (I-IV)
The study was carried out in January 2000 in a town in the middle of Sweden (Ludvika-Smedjebacken). The total population was 40,000 and 4,479 were children (2,158 girls: 2,321 boys) who attended school. The male to female ratio was 1.04:1.

In Sweden children start the first grade when they are 6-7 years old and leave the ninth grade at 15-16 years of age. Both regular schools (normal classes and three classes for children with language disorder and severely disturbed behaviour) and special schools for children with mental retardation were included in the system. The study was carried out in three stages, tic identification, interview, and clinical examination.

Clinical material and controls (I)
Step 1. Identification of children with tics
All 4,479 children and their parents were asked to fill in a questionnaire (Appendix II) concerning different tics. Four children did not participate in this study because they attended school away from their hometown, two children had severe language disability, one child had severe cerebral palsy, while another child had autism.

Step 2. Interview
The parents who answered that their children had possible tics (n=298) were interviewed via telephone (60 children) and via personal interview (238 children) by the author to confirm or refute the diagnosis.

Step 3. Clinical assessment
Only 238 (80%) of the children with tics accepted a physical examination.
Identified cases and controls (II, III, IV)

Twenty-five controls without tics and 25 children with TT were randomly chosen with same age, sex and school as the TS children. Their mean age was 10.4±2.2 years. The children with CMT or CVT, 18 girls and 40 boys, had the mean age of 10.2±0.7 years. Two children (one of each sex) with TS did not want to be personally examined. They were both known by the author and gave their permission to use all documents and medical records.

Instead of using the 1980thies Swedish norms for the “I think I am” self-perception test, new comparison groups were used, one for middle school and one for elementary school children, with 25 children with TS. Twenty-nine children, 86% boys, constituted the middle school comparison group, and 27 children, 78% boys, constituted the elementary school comparison group. The comparison groups and TS group did not significantly differ in sex distribution (Table 6).

Table 6. The population based cases identified and controls in studies I-IV.

<table>
<thead>
<tr>
<th>Population (Study I)</th>
</tr>
</thead>
<tbody>
<tr>
<td>4,479 children (2,158 girls: 2,321 boys) aged 7-15 years, who attended schools in the community of Ludvika-Smedjebacken</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Cases identified (Study I)</th>
</tr>
</thead>
<tbody>
<tr>
<td>297 children with tics, 107 girls and 190 boys. (25 children had Tourette syndrome, 34 children had chronic motor tics, 24 children had chronic vocal tics, 214 children had transient tics)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Cases identified and controls (Study II-III)</th>
</tr>
</thead>
<tbody>
<tr>
<td>25 children with TS, 34 with chronic motor tics, 24 with chronic vocal tics, 25 with transient tics and 25 without tics</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Cases identified and controls (Study IV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>25 with TS and 27+29 children without tics</td>
</tr>
</tbody>
</table>

23
Methods

Case-finding procedure

Questionnaire

A screening questionnaire concerning different tics was developed on the basis of a literature review, clinical experience and DSM-IV criteria. This questionnaire was designed by the author for the parents and teachers in order to evaluate the child’s motor tics in its face, upper part of the body and extremities, as well as vocal tics with sounds and words/phrases. The questionnaire consisted of six items (See Appendix I-II).

A pilot study was carried out in a nearby city (Borlänge) in September 1999 among children in the 4th grade who were 10 to 11 years old (n=70). The tic-screening questionnaire together with common information about tic disorders and specific information about the study were sent to the children and their parents. The questionnaire was also given to the teachers after the parents and children’s approval. Responses from 56 parents were received. Four of them did not want the teachers to participate. Thus 52, teachers completed the questionnaire.

According to parental reports, 12 students had definite tics, two had probable tics and 42 had no tics at all. The teachers reported only five students with tics. Three of them coincided with parental answers while the teachers reported two students with tics, which the parents did not. The possible tic disorder was 14 of 16 according to the parents and 5 of 16 according to the teachers.

A separate question at the end of the questionnaire in the pilot study dealt with the clarity of the descriptions given. Both parents and teachers answered that it was easy to understand the descriptions. As the parents seemed to be more adequate informants, it was considered sufficient for the purpose of this study to ask only the parents together with their child to fill in the tic-screening questionnaire.

Procedure (I)

Both oral and written general information about the tic disorders and TS, including a description of the conditions to be studied, was given to the teacher, school nurse, school physician, school psychologist, and school
social worker. Additional written information with a specified description of the study was given to the school administrators in each community, as well as to the parents and children.

The class teacher distributed the tic-screening questionnaire to the parents. The parents were instructed to fill in the questionnaire together with the child. In the first round, answers from 2,007 parents were received (44.8%). In the second round, a new letter together with the screening questionnaire was sent directly to the parents whose children did not return the questionnaire. The postages for the letters were pre-paid, and another 639 questionnaires were received (14.3%). Thus the proportion of answers in the second round was 59.1%. In order to find out the proportion of tic disorders among the 40.9% of the children and parents who did not answer at all, a new letter was sent to every fifth child (n=368) and answers from 298 parents were received in this third round. Thus the total number of questionnaires fully completed was 2,944 (65.7%), 1,528 males and 1,416 females (Table 7).

Table 7. Questionnaire response from the parents

<table>
<thead>
<tr>
<th>Total population</th>
<th>4,479</th>
</tr>
</thead>
<tbody>
<tr>
<td>Responses:</td>
<td></td>
</tr>
<tr>
<td>1st round</td>
<td>2,007 (44.8%)</td>
</tr>
<tr>
<td>2nd round</td>
<td>639 (14.3%)</td>
</tr>
<tr>
<td>3rd round sent to every 5th child who did not answer (n=368)</td>
<td>298 (6.6%)</td>
</tr>
<tr>
<td>Total response</td>
<td>2,944 (65.7%)</td>
</tr>
</tbody>
</table>

Parent and child interview (I-III)

The parents who answered that their children had possible tics (n=298) were interviewed by the author to confirm or refute the diagnosis. Only 238 (80%) of the children accepted a personal interview and physical examination. Sixty parents were interviewed only via telephone by the author. All sixty reported that the tics had disappeared. The duration of the tics had been a few months only. Two children (one boy and one girl) with TS participated in the first interview but they did not want to participate in the extensive examination. They were both known by the author and gave their permission to use all documents and medical records for information in the study.

The parents of each child had a semi-structured interview (91), which lasted about 90 minutes. The interview was longer if the child had major problems. It was systematically focused on heredity, pregnancy, obstetrical and neonatal circumstances, milestones of development, current behaviour, social as well as motor performance. The child was present during the interview. A clinical interview together with Swedish translations of different rating scales was used (see below). Information was also collected about mental disorders through systematic interviews with a checklist based on
DSM-IV criteria as well as observations of psychopathology during the interview.

Clinical assessment (I-IV)

*Physical examination (I-III)*

Each child was examined by a routine neuropaediatric and a general physical examination using a modified version of the neurodevelopmental-screening test (92). The children were all (238) examined by the author (except two who were examined by professor A-L von Knorring). The examination was comprised of a routine paediatric examination such as weight, height, head circumference, speech, language, activity level, skull, mouth, lungs, heart, blood pressure, abdomen, skin, and puberty stage (93).

A more detailed neurological examination was carried out. The examination included the testing of cranial nerves, fine motor skills, diadochokinesis, standing on the left and right leg for 20 seconds, walking on heels and toes, Fog’s test, Prechtl’s sign, the Romberg test, jumping back and forth across a line, the fine motor test, stereognosis, and Babinski and tendon reflexes.

The total physical and neurodevelopmental score ranged from 0 to 32. Each item was scored on a scale from 0 to 2 (0=no abnormality, 1=some abnormality, and 2=major abnormality). The examination lasted one and a half hour and 19 children had been followed by the author for 4-9 years prior to the study. A physiotherapist examined children showing marked gross motor problems. All the children with TS were also examined by an occupational therapist concerning fine motor skills.

*Psychological examination (IV)*

The children with TS completed a psychological test battery in a uniform order, across two or three testing sessions, each about 1.5 hour in duration. The battery included an intelligence test (WISC III) (94), neuropsychological assessment with the Rey complex figure test (95), verbal fluency (NEPSY) (96), and a self-rating scale (I think I am) (97). Child mean age at testing was 11 years (SD=2.7).

Fifteen children were tested with Rey complex figure test (NIMES), seventeen children were tested with the verbal fluency test (NEPSY) and eighteen children were tested with “jag tycker jag är” (I think I am) test, 24 of the children were tested with WISC III and one child with WPPSI, the preschool version of the WISC test.

*Assessment measures (I-IV)*

The assessment involved a comprehensive interview about medical history, placement of the child and behaviour problems based on a questionnaire (91)
and Swedish translations of different rating scales. The scales were Children’s Yale-Brown Obsessive Compulsive Scale [CY-BOCS] (98), Conners parent and teacher scales (99), Child Behaviour Checklist [CBCL] (100), Asperger Syndrome Screening Questionnaire [ASSQ] (101), and Children’s Depression Inventory [CDI] (102). Information about mental disorders was also collected through systematic interviews with a checklist based on DSM-IV criteria. Tic type and severity were assessed by clinical examination and by the Shapiro Tourette Syndrome Severity Scale [TSSS] (3, 8).

Pregnancy, obstetrical and other relevant medical records were collected concerning the 25 children with TS and their matched controls. The method used for rating “reduced optimality in the pre-, peri- and neonatal periods” is described in Gillberg and Gillberg (103). It is a modified version of Prechtl’s list of optimal conditions (104). For each factor with optimal conditions a score of 0 was given, and for each condition outside the optimal range a score of 1 was given. All scores for individual items were then added to obtain a total score for reduced optimality in the pre-, peri- and neonatal period (Table 8).

**Tic severity scale**

The Tourette syndrome severity scale (TSSS), developed by Shapiro and Shapiro (3, 8) for use in a clinical trial of pimozide, includes a composite clinincinal rating of severity comprising five factors. Scores for the five items are summed and converted to a qualitative global severity rating. The scoring of tic severity range was from 0 (none) to 9 (very severe) (Appendix III). The TSSS is simple to use, appears valid, is highly reliable when used by physicians and provides an overall index of tic severity. This scale is limited in that it fails to assess the width of tic characteristics.

**I think I am**

This is a Swedish instrument to measure self-perception, which is widely used in clinical settings and in research in Sweden. There is one version for elementary school children and one version for middle school and high school children. The elementary school version has 32 items and the middle school version has 72 items. The elementary school version has scales for physical well-being (6 items), for achievement (6 items), psychological well-being (8 items), relations to parents and family (6 items), and relations to others (6 items). The middle school version has scales for physical well-being (14 items), achievement (14 items), psychological well-being (16 items), relations to parents and family (14 items), and relations to others (14 items). Scale scores are computed as the sum of items. High scores reflect a positive self-evaluation and the total score is assumed to reflect the child’s global self-evaluation (97).
**Wechsler Intelligence Scale for Children (WISC-III)**
This is a commonly used version of the popular intelligence test, for which Swedish norms are available. The following measures were used: Index scores for the four factors (1) verbal comprehension (VCI), (2) perceptual organisation (POI), (3) freedom from distractibility (FDI) and (4) processing speed (PSI) as well as verbal IQ (VIQ) scores, performance IQ (PIQ) scores and full scale IQ (FSIQ) scores. Scale scores for digit span forward and back-wards were used to measure working memory (94).

**Rey Complex Figure Test (RCFT)**
This is a test used to measure executive functions, i.e., visuospatial construction ability, visual planning and visual memory. The child is asked to copy a geometric figure. After a couple of minutes the child is asked to recall and draw the figure, and after thirty minutes the child is again asked to recall and draw the figure. Four measures were extracted: copying, short term memory, delayed recall, and organisation. Australian norms were used and Stanine scores for each measure were used in the calculations (95).

**Verbal fluency (NEPSY)**
This is a test of semantic and phonological flow. As a measure of semantic flow, the child is asked to produce as many animal words as possible within one minute, and then to produce as many words as possible for things one can eat or drink within one minute. As a measure of phonological flow, the child is then asked to produce as many words as possible beginning with an S and as many words as possible beginning with an F, respectively, within one minute each. Three scores were computed as the sum of semantic, of phonological, and as a total score of semantic plus phonological words, respectively. Swedish norms were used, and the child’s performance was expressed in terms of the percentile for his/her age (96).

**Social class (II)**
Social class was determined using the classification of the Swedish Central Bureau of Statistics (105), which takes into account the breadwinner occupational status. The mother’s level of education was also included in this study.

**Diagnostic classification (I-IV)**
For each child the neuro-developmental and/or psychiatric diagnoses were made by the author (except two made by professor A-L von Knorring) on the basis of the information and findings obtained at the child’s examination and a history of the symptoms, based on the interview with the child and his/her parent, from questionnaires (see above) and previous documents. Psychiatric diagnosis according to DSM-IV criteria was used.
Table 8. Instruments and statistics

<table>
<thead>
<tr>
<th>Study</th>
<th>Instruments</th>
<th>Statistics used</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Child Tic Screening Questionnaire, DSM IV, Neurodevelopmental examination, Semi-structured interview</td>
<td>Chi-square test</td>
</tr>
<tr>
<td>II</td>
<td>DSM IV, Optimality score in the pre-peri and neonatal period, Semi-structured interview, Social class</td>
<td>Chi-square test, Oneway ANOVA, Pearson’s correlation</td>
</tr>
<tr>
<td>III</td>
<td>ASSQ parent and teacher, CBCL, CDI, Conners parent and teacher scales, CY-BOCS, DSM IV, TSSS</td>
<td>Chi-square test, Oneway ANOVA, Pearson’s correlation</td>
</tr>
<tr>
<td>IV</td>
<td>I think I am, RCFT, TSSS, Verbal fluency, WISC III, WIPPSI</td>
<td>Chi-square test, Mann-Whitney U-test, Pearson’s correlation</td>
</tr>
</tbody>
</table>

Statistics (I-IV)

The SPSS for Macintosh 10.0 statistical package was used. Comparisons of proportions between groups were performed using the chi-square test. Comparisons of mean values between more than two groups were performed with oneway ANOVA. Mann-Whitney test were used to compare distributions for small samples. Pearson’s correlation coefficient was used to examine the linear association between variables. A significance level of 5% was applied throughout (Table 8).
Results

Prevalence of Tourette syndrome and other tic disorders (I)

Two hundred and thirty-nine children with tics were reported in the first round of the screening: 19 (0.9%) with TS, 27 (1.3%) with chronic motor tics, 19 (0.9%) with chronic vocal tics, and 174 (8.7%) with transient tics. The second screening round involved 639 children, 57 of whom were reported to have tics: six with TS (0.9%), seven with chronic motor (1.1%), and five with chronic vocal tics (0.8%), and 39 with transient tics (6.1%). Finally, the third screening round found only one child with transient tics (0.3%) (Table 9).

Table 9. Prevalence of the different tic disorders in the three rounds. Tourette syndrome (TS), chronic motor tics (CMT), chronic vocal tics (CVT) and children with transient tics (TT).

<table>
<thead>
<tr>
<th>Response round</th>
<th>TS</th>
<th>CMT</th>
<th>CVT</th>
<th>TT</th>
</tr>
</thead>
<tbody>
<tr>
<td>The first round n=2,007</td>
<td>19 (0.9%)</td>
<td>27 (1.3%)</td>
<td>19 (0.9%)</td>
<td>174 (8.7%)</td>
</tr>
<tr>
<td>The second round n=639</td>
<td>6 (0.9%)</td>
<td>7 (1.1%)</td>
<td>5 (0.8%)</td>
<td>39 (6.1%)</td>
</tr>
<tr>
<td>The third round n=298</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1 (0.3%)</td>
</tr>
<tr>
<td>Total n=2,944</td>
<td>25 (0.8%)</td>
<td>34 (1.1%)</td>
<td>24 (0.8%)</td>
<td>214 (7.3%)</td>
</tr>
</tbody>
</table>

Sixty children with transient tics did not want to come to the clinical examination. They did not significantly differ according to age, sex, and level of tic severity or number of periods compared with the 154 individuals with transient tics who participated in the clinical examination.

As seen in table 10, in the total population, 297 children (6.6%) suffered from tics, the male to female ratio was 1.6:1. Twenty-five children (0.6%) had Tourette syndrome; the male to female ratio of TS was 9:1. Thirty four (0.8%) children had chronic motor tics with a male to female ratio of 2:1.
Twenty-four children had chronic vocal tics (0.5%), with the male to female ratio 2.3:1. Two hundred and fourteen children (4.8%) had transient tics during the last year. The male to female ratio 4:1 and 87% had only one tic period, 13% had two or more episodes. One child who reported tics in the questionnaire was found to suffer from asthma when clinically examined. These results were counted on the total population and not only on the population who answered the questionnaire.

The results would be even higher if only those who answered had been counted (Table 11). The frequency of different tic disorders in different age groups showed the same pattern; higher frequencies were found among boys, although this was less pronounced for transient tics than Tourette syndrome and chronic tics. The highest prevalence was found among the youngest children, which was the case for both boys and girls.

In the special school/classes nineteen children (46.3%) with tics were found. This is a significantly higher proportion than the 6.3% in the regular schools (p<0.001).

Table 10. One-year prevalence of different tic disorders of school children, 7-15 years.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Sex</th>
<th>7-9 y</th>
<th>10-12 y</th>
<th>13-15 y</th>
<th>TOTAL</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n (%)</td>
<td>n (%)</td>
<td>n (%)</td>
<td>n (%)</td>
<td></td>
</tr>
<tr>
<td>Tourette Syndrome</td>
<td>Female</td>
<td>1 (0.1)</td>
<td>0</td>
<td>2 (0.3)</td>
<td>3 (0.1)</td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>10 (1.2)</td>
<td>10 (1.3)</td>
<td>2 (0.3)</td>
<td>22 (0.9)</td>
</tr>
<tr>
<td>Chronic Motor Tics</td>
<td>Female</td>
<td>3 (0.4)</td>
<td>2 (0.3)</td>
<td>6 (0.9)</td>
<td>11 (0.5)</td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>14 (1.7)</td>
<td>6 (0.8)</td>
<td>3 (0.4)</td>
<td>23 (1.0)</td>
</tr>
<tr>
<td>Chronic Vocal Tics</td>
<td>Female</td>
<td>4 (0.5)</td>
<td>2 (0.2)</td>
<td>1 (0.2)</td>
<td>7 (0.3)</td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>6 (0.7)</td>
<td>5 (0.7)</td>
<td>6 (0.8)</td>
<td>17 (0.7)</td>
</tr>
<tr>
<td>Transient Tics</td>
<td>Female</td>
<td>47 (6.0)</td>
<td>24 (3.3)</td>
<td>15 (2.3)</td>
<td>86 (4.0)</td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>56 (6.7)</td>
<td>41 (5.4)</td>
<td>31 (4.3)</td>
<td>128 (5.5)</td>
</tr>
<tr>
<td>Tics (total)</td>
<td>Female</td>
<td>55 (7.8)</td>
<td>28 (3.9)</td>
<td>24 (3.7)</td>
<td>107 (5.0)</td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>86 (10.3)</td>
<td>62 (8.2)</td>
<td>42 (5.8)</td>
<td>190 (8.2)</td>
</tr>
</tbody>
</table>
Table 11. Comparison between prevalences the total population and the population answering the questionnaire, Tourette syndrome (TS), chronic motor tics (CMT), chronic vocal tics (CVT) and children with transient tics (TT).

<table>
<thead>
<tr>
<th></th>
<th>Special school</th>
<th>Regular school</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total population</td>
<td>Response population</td>
<td>Total population</td>
<td>Response population</td>
</tr>
<tr>
<td></td>
<td>N=41</td>
<td>N=36</td>
<td>N= 4,438</td>
<td>N=2,908</td>
</tr>
<tr>
<td>TS</td>
<td>3 (7.3%)</td>
<td>3 (8.3%)</td>
<td>22 (0.5%)</td>
<td>22 (0.8%)</td>
</tr>
<tr>
<td>CMT</td>
<td>4 (9.8%)</td>
<td>4 (11%)</td>
<td>30 (0.7%)</td>
<td>30 (1.0%)</td>
</tr>
<tr>
<td>CVT</td>
<td>7 (17.1%)</td>
<td>7 (19.4%)</td>
<td>17 (0.4%)</td>
<td>17 (0.6%)</td>
</tr>
<tr>
<td>TT</td>
<td>5 (12.2%)</td>
<td>5 (13.9%)</td>
<td>209 (4.7%)</td>
<td>209 (7.2%)</td>
</tr>
</tbody>
</table>

Symptoms, severity, and age of onset (II)

Symptoms
In 18 (72%) cases with TS, the initial symptom started with tics in the head and face. Vocalizations were reported as the initial symptom in four (16%) mostly repeated throat clearing. At the time of examination, all children with TS showed simple tics. Complex motor tics were present in 10 (40%) children with TS. Vocal tics were predominantly of the simple type, such as sniffing, throat clearing, blowing, and whistling. The distribution of tics at the time of the examination can be seen in table 12.

Table 12. Prevalence of different symptoms at the time of examination in children with Tourette syndrome, chronic motor tics (CMT), chronic vocal tics (CVT) and transient tics (TT).

<table>
<thead>
<tr>
<th></th>
<th>TS n (%)</th>
<th>CMT n (%)</th>
<th>CVT n (%)</th>
<th>TT n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Motor tics (simple and complex)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eyes, face or head</td>
<td>18 (72)</td>
<td>26 (76)</td>
<td>8 (64)</td>
<td></td>
</tr>
<tr>
<td>Shoulder, neck, arm</td>
<td>14 (56)</td>
<td>15 (44)</td>
<td>4 (16)</td>
<td></td>
</tr>
<tr>
<td>Trunk, legs</td>
<td>11 (44)</td>
<td>6 (20)</td>
<td>3 (12)</td>
<td></td>
</tr>
<tr>
<td>Simple vocal tics</td>
<td>18 (72)</td>
<td>16 (66)</td>
<td>3 (12)</td>
<td></td>
</tr>
<tr>
<td>Complex vocal tics</td>
<td>13 (52)</td>
<td>12 (50)</td>
<td>2 (8)</td>
<td></td>
</tr>
<tr>
<td>Coprolalia</td>
<td>11 (44)</td>
<td>8 (32)</td>
<td>1 (4)</td>
<td></td>
</tr>
<tr>
<td>Compulsion</td>
<td>9 (36)</td>
<td>2 (6)</td>
<td>2 (8)</td>
<td>0</td>
</tr>
</tbody>
</table>

Age at onset and age at first diagnosis
The first neurological symptoms of TS appeared between 3 and 10 years of age (mean 4.6±2.9 y). The first symptoms of children with CMT were observed at 7.4±2.6 years and CVT at 8.2±2.1 years (CMT versus CVT: n.s.).
The 25 matched children with transient tics had onset of tics at 8.5±3.0 years. The age of onset of children with TS was significantly lower than the age of onset of chronic motor or chronic vocal tics (CMVT) (F=13.4, p=0.000) (Figure 1).

![The age of the onset of tics](image1)

*Figure 1. The age of the onset of tics*

The average interval between the onset of the first symptoms and diagnosis of TS was 4 years, ranging from 1 to 8 years (Figure 2).

![Age at onset and age at diagnosis](image2)

*Figure 2. Age at onset and at diagnosis of 25 children with TS*
Experience with counseling services

Prior to the study, three of the TS cases had been in contact with outpatient services of child psychiatry, six children had been treated by a child neurologist, and one child had been treated by a paediatrician. Another fourteen had been in contact with at least two of the previously mentioned services/specialists.

The children with TS were referred with different symptoms (table 13). Nine children were diagnosed with TS during the study, and eight of them had been in contact with the medical service. The only child without contact with the medical service was the sibling of a child already diagnosed as suffering from Tourette syndrome. The parents to the children with TS were also asked to rate how helpful they had found those services to be. The majority of the parents thought that the services were not helpful enough, and they were especially dissatisfied with the child psychiatry outpatient services and from the school staff.

<table>
<thead>
<tr>
<th>Diagnoses</th>
<th>N</th>
<th>(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tourette syndrome</td>
<td>3</td>
<td>(12)</td>
</tr>
<tr>
<td>Tourette syndrome with comorbidity</td>
<td>13</td>
<td>(52)</td>
</tr>
<tr>
<td>Language disturbance + OCD</td>
<td>1</td>
<td>(4 )</td>
</tr>
<tr>
<td>ADHD</td>
<td>4</td>
<td>(16)</td>
</tr>
<tr>
<td>Asperger syndrome</td>
<td>1</td>
<td>(4 )</td>
</tr>
<tr>
<td>Defiance and child-parent problems</td>
<td>2</td>
<td>(8 )</td>
</tr>
<tr>
<td>No contact with medical service</td>
<td>1</td>
<td>(4 )</td>
</tr>
</tbody>
</table>

Familial loading of mental disorders (II)

Questions about psychiatric illnesses were asked three generations back. Eighty percent of TS children had first-degree relatives with a psychiatric illness. The most common were different tic disorders, ADHD, OCD, and depression. Only 20% of the control children were reported to have first-degree relatives with psychiatric illness (TS=20 vs. controls=5, $\chi^2=18.0$, P<0.001) (Table 14).
Table 14. Number of children with tic disorders and matched controls who have a first-degree relative with psychiatric disorder

<table>
<thead>
<tr>
<th>Disorder</th>
<th>TS n (%)</th>
<th>CT n (%)</th>
<th>TT n (%)</th>
<th>C n (%)</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tic Disorder</td>
<td>13 (52)</td>
<td>6 (10)</td>
<td>6 (24)</td>
<td>3 (12)</td>
<td>TS &gt; CT ***</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS &gt; TT *</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS &gt; C**</td>
</tr>
<tr>
<td>OCD /Anxiety Disorder</td>
<td>8 (32)</td>
<td>2 (4)</td>
<td>0 (0)</td>
<td>2 (8)</td>
<td>TS &gt; CT**</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS &gt; TT**</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS &gt; C*</td>
</tr>
<tr>
<td>Depressive Disorders</td>
<td>6 (24)</td>
<td>6 (10)</td>
<td>0 (0)</td>
<td>1 (4)</td>
<td>TS &gt; TT **</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS &gt; C *</td>
</tr>
<tr>
<td>ADHD/Learning Disability</td>
<td>14 (56)</td>
<td>14 (24)</td>
<td>3 (12)</td>
<td>5 (20)</td>
<td>TS &gt; CT **</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS &gt; TT**</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS &gt; C **</td>
</tr>
<tr>
<td>Pervasive Development Disorders</td>
<td>3 (12)</td>
<td>1 (2)</td>
<td>0 (0)</td>
<td>1 (4)</td>
<td>n.s.</td>
</tr>
</tbody>
</table>

*** = p < 0.001  ** = p < 0.01  * = p < 0.05

Tourette syndrome (TS), chronic motor tics (CMT), chronic vocal tics (CVT), transient tics (TT), and controls (C)

Pre/perinatal events and socio-economic status (II)

There were no statistical differences between the children with TS and the control children with regard to reduced optimality scores in the pre-, peri-, and neonatal periods. The mothers of the children with TS were twice as likely to have complications during pregnancy and were younger than the control-mothers at the delivery (mean 20±1.4 years vs. 23±3.4 years, p<0.01). Socio-economic status, parental education or divorce rate did not differ significantly between the children with TS, CMT, CVT, TT, and controls.

Comorbidity (III)

Physical and developmental comorbidity at the time of examination

A significantly higher proportion of children with TS (20%) were motor clumsy and/or had deficient coordination disorder (DCD) compared to the control children where no one was found with DCD. Only a few with CMT or CVT had DCD. Language disorder was found more often in children with CVT (21%) compared to controls. Children with and without tics had no differences in the frequency of migraine, asthma, eczema, enuresis or enco-presis. Eleven children with TS (44%) were over-sensitive to olfactory im-
pressions, e.g. they had to smell the food before eating it. The comorbidity is reviewed in table 15.

**Table 15.** Comorbid physical and developmental diagnoses at the time of examination in children with Tourette syndrome (TS), chronic motor tics (CMT), chronic vocal tics (CVT), transient tics (TT), and controls (C).

<table>
<thead>
<tr>
<th>Physical disorder/developmental disorder</th>
<th>TS n (%)</th>
<th>CMT n (%)</th>
<th>CVT n (%)</th>
<th>TT n (%)</th>
<th>Controls n (%)</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>2 (8)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>ns</td>
</tr>
<tr>
<td>Language disability</td>
<td>1 (4)</td>
<td>2 (6)</td>
<td>5 (21)</td>
<td>1 (4)</td>
<td>0</td>
<td>CVT&gt;C*</td>
</tr>
<tr>
<td>DCD</td>
<td>5 (20)</td>
<td>2 (6)</td>
<td>2 (8)</td>
<td>0</td>
<td>0</td>
<td>TS&gt;TT, C*</td>
</tr>
<tr>
<td>Atopic disorder/asthma</td>
<td>8 (32)</td>
<td>7 (21)</td>
<td>2 (8)</td>
<td>7 (28)</td>
<td>ns</td>
<td></td>
</tr>
<tr>
<td>Epilepsy</td>
<td>1 (4)</td>
<td>1 (3)</td>
<td>0</td>
<td>1 (4)</td>
<td>ns</td>
<td></td>
</tr>
<tr>
<td>Mental retardation</td>
<td>4 (16)</td>
<td>3 (9)</td>
<td>5 (21)</td>
<td>1 (4)</td>
<td>1 (4)</td>
<td>ns</td>
</tr>
<tr>
<td>Learning disability</td>
<td>4 (17)</td>
<td>1 (3)</td>
<td>1 (4)</td>
<td>2 (8)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Sleep disorder</td>
<td>7 (28)</td>
<td>2 (6)</td>
<td>2 (8)</td>
<td>4 (16)</td>
<td>TS&gt;TT**, TS&gt;CMT*, C&gt;TT*</td>
<td></td>
</tr>
</tbody>
</table>

*** = p< 0.001    ** = p< 0.01    * = p< 0.05

**Psychiatric comorbidity at time of examination**

The frequency of psychiatric comorbidity differed significantly between TS and other tic disorders and controls. The children with TS generally had many more associated problems than children with other tic disorders and controls both in terms of school adjustment and behavioural dysfunction. Ninety-two percent of the TS children had one or more diagnoses. Children with CVT had also increased psychiatric morbidity that differed from children with TT and controls (Table 16).

**Table 16.** Comorbid psychiatric diagnoses at the time of examination in children with Tourette syndrome (TS), chronic motor tics (CMT), chronic vocal tics (CVT), transient tics (TT) and controls (C).

<table>
<thead>
<tr>
<th>Psychiatric disorders</th>
<th>TS n (%)</th>
<th>CMT n (%)</th>
<th>CVT n (%)</th>
<th>TT n (%)</th>
<th>Controls n (%)</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>ADHD</td>
<td>17 (68)</td>
<td>4 (12)</td>
<td>8 (33)</td>
<td>1 (4)</td>
<td>2 (8)</td>
<td>TS&gt;CMT, TT, C***</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>TS&gt;CVT*, CVT&gt;CMT, TT, C**</td>
</tr>
<tr>
<td>Autism spectrum</td>
<td>5 (20)</td>
<td>1 (3)</td>
<td>1 (4)</td>
<td>1 (4)</td>
<td>0</td>
<td>TS&gt;CMT, C*</td>
</tr>
<tr>
<td>Depression</td>
<td>5 (20)</td>
<td>2 (6)</td>
<td>1 (4)</td>
<td>0</td>
<td>0</td>
<td>TS&gt;TT, C*</td>
</tr>
<tr>
<td>OCD</td>
<td>4 (16)</td>
<td>0</td>
<td>2 (8)</td>
<td>0</td>
<td>0</td>
<td>TS&gt;CMT, TT, C*</td>
</tr>
<tr>
<td>CD</td>
<td>2 (8)</td>
<td>0</td>
<td>0</td>
<td>1 (4)</td>
<td>ns</td>
<td></td>
</tr>
</tbody>
</table>

*** = p< 0.001    ** = p< 0.01    * = p< 0.05
Symptom ranking
Families considered rage symptom, attention deficit and learning difficulties to be most important, while motor and vocal tics were least important (Table 17).

Table 17. Percentage of the parents of the children with TS ranking symptom as very important

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rage</td>
<td>36%</td>
</tr>
<tr>
<td>Hyperactivity/ Attention deficit</td>
<td>20%</td>
</tr>
<tr>
<td>Learning difficulties</td>
<td>20%</td>
</tr>
<tr>
<td>Obsessions/Compulsion</td>
<td>12%</td>
</tr>
<tr>
<td>Tics</td>
<td>12%</td>
</tr>
</tbody>
</table>

Overall levels of behavioural and emotional problems
Children with TS showed significantly more often behavioural and adaptive problems than children with other tic disorders or controls. The TS group had significantly higher scores on all the subscales of CBCL. The children with TS recorded significantly higher mean total scores on CDI, Conners, ASSQ and CY-BOCS than the CMT, TT and control groups. Teacher’s and parent’s Conners rating scales (total scores) correlated significantly with total CBCL scores (Pearson r=0.31 and 0.26 respectively, p<0.001 and p<0.01). A similar level of correlation was seen in respect of teacher’s and parent’s ASSQ rating scales with total CBCL scores (Pearson r=0.28 and 0.23 respectively p<0.01 in both instances). No correlations were found between CDI or CY-BOCS and CBCL (Table 18).

Correlates of tic severity (II-IV)

Tic severity of children with TS was significantly higher compared with other groups of tic disorders. Mean score of TSSS for TS was 5.0±1.9 (marked tic severity), CMT 1.4±1.1 (mild) and TT 0.4±0.6 (very mild) (F-value 95.8, p<0.001). Children with severe TS were younger at the onset of the disorder (Pearson r=0.56, p<0.01). No significant correlation was found between the age of the onset and the severity of CMVT. No correlation was found between tic severity and IQ in the TS children. The tic severity scale did not show significant correlation with the total or sub-scores of CBCL.
Table 18. Mean score of behaviour problems (CBCL; Child Behaviour Checklist) in children with Tourette syndrome (TS), chronic motor tic disorder (CMT), chronic vocal tic disorder (CVT), transient tic disorder (TT) and controls.

<table>
<thead>
<tr>
<th>Measure</th>
<th>TS M (SD)</th>
<th>CMT M (SD)</th>
<th>CVT M (SD)</th>
<th>TT M (SD)</th>
<th>Control M (SD)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total score</td>
<td>40.5 (28.2)</td>
<td>13.4 (22.0)</td>
<td>19.5 (18.9)</td>
<td>9.9 (17.8)</td>
<td>13.8 (18.4)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Attention problems</td>
<td>7.1 (3.2)</td>
<td>2.8 (4.2)</td>
<td>4.3 (4.3)</td>
<td>1.6 (2.7)</td>
<td>2.9 (4.0)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Social problems</td>
<td>3.2 (2.8)</td>
<td>1.3 (2.4)</td>
<td>2.2 (2.9)</td>
<td>1.0 (2.0)</td>
<td>1.8 (2.6)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Thought problems</td>
<td>1.4 (2.0)</td>
<td>0.7 (1.5)</td>
<td>0.8 (1.8)</td>
<td>0.2 (0.8)</td>
<td>0.7 (1.4)</td>
<td>ns</td>
</tr>
<tr>
<td>Externalizing behaviours</td>
<td>15.7 (12.0)</td>
<td>4.2 (7.9)</td>
<td>7.1 (8.2)</td>
<td>4.3 (9.4)</td>
<td>4.6 (6.5)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Delinquent behaviour</td>
<td>3.4 (3.4)</td>
<td>1.3 (2.8)</td>
<td>1.6 (2.3)</td>
<td>1.2 (2.4)</td>
<td>1.2 (2.5)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Aggressive behaviour</td>
<td>12.2 (9.2)</td>
<td>2.9 (5.5)</td>
<td>5.5 (6.1)</td>
<td>3.1 (7.2)</td>
<td>3.4 (4.7)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Internalizing behaviours</td>
<td>8.5 (9.6)</td>
<td>2.6 (4.8)</td>
<td>3.0 (4.1)</td>
<td>1.4 (3.4)</td>
<td>2.6 (3.5)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Somatic complaints</td>
<td>2.0 (2.7)</td>
<td>0.9 (1.8)</td>
<td>0.5 (1.0)</td>
<td>0.3 (0.6)</td>
<td>0.5 (1.0)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Anxious/ Depressed</td>
<td>4.4 (5.9)</td>
<td>1.0 (2.0)</td>
<td>1.4 (2.7)</td>
<td>0.8 (2.1)</td>
<td>1.2 (2.1)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Withdrawn</td>
<td>2.4 (2.3)</td>
<td>0.7 (1.8)</td>
<td>1.1 (1.7)</td>
<td>0.4 (1.2)</td>
<td>0.8 (1.5)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Cognition and self-perception (IV)

Cognitive function

Children with TS attained slightly below average on WISC-scores. The variation was very large. Three out of the 21 children with a complete test protocol (14.3%) had a full scale IQ ≤ 70. Furthermore, one child for which detailed WISC-data was missing was diagnosed as mentally retarded, and a child tested with WIPPSI was mildly retarded. There were also a few children scoring 2 SD above average.

One child had a verbal IQ of 135 and one child had a performance IQ of 133. As seen in figure 3, the profile in index scores for the WISC factors was somewhat uneven. Paired t-tests of the differences revealed that the mean perceptual organization score differed significantly from the freedom from distractibility and processing speed scores, p<0.05. The difference between the perceptual organisation score and the verbal comprehension score was not significant, p= 0.14.

As regards executive functions, the TS group scored at or above average on the RCFT tests. However, variation was large, and between 33 and 40%
had ≤ stanine 3 on the memory and spatial organization tests. Only one child scored that low on the RCFT copying test. Half of the group scored in the lowest 25 percent, or worse, on the verbal fluency tests. The WISC digit span scores demonstrated that the children also had difficulties with verbal working memory. As many as 79% of those tested scored below the normal range on the digit span backward test, and 32% scored below the normal range on the digit span forward test (Table 19).

Table 19. Results of cognitive tests

<table>
<thead>
<tr>
<th>Tests</th>
<th>Mdn</th>
<th>Variation</th>
</tr>
</thead>
<tbody>
<tr>
<td>WISC, n = 21-22</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Verbal IQ</td>
<td>94</td>
<td>53–135</td>
</tr>
<tr>
<td>Performance IQ</td>
<td>97.5</td>
<td>76–133</td>
</tr>
<tr>
<td>Full scale IQ</td>
<td>95</td>
<td>57–125</td>
</tr>
<tr>
<td>RCFT, n = 15, stanine scores</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Copying</td>
<td>7</td>
<td>2–9</td>
</tr>
<tr>
<td>Short term memory</td>
<td>5</td>
<td>2–9</td>
</tr>
<tr>
<td>Delayed recall</td>
<td>5</td>
<td>1–9</td>
</tr>
<tr>
<td>Organization</td>
<td>7</td>
<td>1–9</td>
</tr>
<tr>
<td>Verbal fluency, n = 17&lt;sup&gt;a&lt;/sup&gt;</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Semantic categories</td>
<td>11–25</td>
<td>&lt;10–75/100</td>
</tr>
<tr>
<td>Phonological categories</td>
<td>3–10</td>
<td>&lt;10–75/100</td>
</tr>
<tr>
<td>Total</td>
<td>11–25</td>
<td>&lt;10–75/100</td>
</tr>
<tr>
<td>Verbal working memory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WISC digit span forward&lt;sup&gt;b&lt;/sup&gt;</td>
<td>7</td>
<td>4–11</td>
</tr>
<tr>
<td>WISC digit span backward&lt;sup&gt;b&lt;/sup&gt;</td>
<td>4</td>
<td>2–14</td>
</tr>
</tbody>
</table>

<sup>a</sup> Results expressed in percentiles

<sup>b</sup> Results expressed in scale scores, M = 10, normal range 7 – 13

![Figure 3. Mean index scores on the Verbal Comprehension (VCI), Perceptual Organization (POI), Freedom from Distractibility (FDI) and Processing Speed (PSI) WISC factors](image-url)
Correlation between IQ and medication

The children taking medication (n=11) had lower full IQ, Mdn=81.5, than children who were not (n=10), Mdn=101, p<0.05.

Self-perception

Children with TS had in several respects less positive self-perception as well as a lower total score than children in the comparison groups. The variation in scores was again substantial, and about the same in the TS and the comparison groups. Children in the high and low tics severity groups did not differ in self-perception, nor did the age at onset have relations with self-perception, p>0.10. Finally, relation between cognitive performance and self-perception were investigated. Performance IQ was negatively related to total self-perception score, p<0.05. Higher IQ was associated with less positive self-perception (Table 20).

Table 20. Self-perception among children with TS and comparison children

<table>
<thead>
<tr>
<th>Self-perception aspect</th>
<th>TS Group Middle school N = 14</th>
<th>Comparison Group Middle school N = 29</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical well-being</td>
<td>Mdn 10.5 Variation – 3–15</td>
<td>Mdn 17 Variation 3–27</td>
<td>0.06</td>
</tr>
<tr>
<td>Achievement</td>
<td>Mdn 11.0 Variation – 9–22</td>
<td>Mdn 15 Variation 8–26</td>
<td>0.05</td>
</tr>
<tr>
<td>Psychological well-being</td>
<td>Mdn 9.5 Variation – 4–26</td>
<td>Mdn 18 Variation 4–31</td>
<td>0.10</td>
</tr>
<tr>
<td>Family relations</td>
<td>Mdn 14.5 Variation – 7–20</td>
<td>Mdn 21 Variation 15–27</td>
<td>0.01</td>
</tr>
<tr>
<td>Social relations</td>
<td>Mdn 14.5 Variation 0–22</td>
<td>Mdn 17 Variation 2–26</td>
<td>0.29</td>
</tr>
<tr>
<td>Total score</td>
<td>Mdn 50.5 Variation – 17–99</td>
<td>Mdn 93 Variation 12–126</td>
<td>0.01</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Self-perception aspect</th>
<th>TS Group Elementary school N = 4</th>
<th>Comparison Group Elementary school N = 27</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical well-being</td>
<td>Mdn 6 Variation 4–6</td>
<td>Mdn 6 Variation 2–6</td>
<td>0.68</td>
</tr>
<tr>
<td>Achievement</td>
<td>Mdn 3 Variation 1–6</td>
<td>Mdn 4 Variation 4–6</td>
<td>0.91</td>
</tr>
<tr>
<td>Psychological well-being</td>
<td>Mdn 3 Variation 2–4</td>
<td>Mdn 8 Variation 2–8</td>
<td>0.01</td>
</tr>
<tr>
<td>Family relations</td>
<td>Mdn 2 Variation – 2–2</td>
<td>Mdn 6 Variation 2–6</td>
<td>0.01</td>
</tr>
<tr>
<td>Social relations</td>
<td>Mdn 3 Variation 2–4</td>
<td>Mdn 6 Variation 2–6</td>
<td>0.01</td>
</tr>
<tr>
<td>Total score</td>
<td>Mdn 14.5 Variation 14–20</td>
<td>Mdn 26 Variation 11–32</td>
<td>0.01</td>
</tr>
</tbody>
</table>
Discussion

This is a first comprehensive and descriptive study concerning different tic disorders in respect to background factors (hereditary, family and social factors), clinical assessment and comorbidity. In addition it is the first controlled study of cognitive function and self-perception in TS in a total school population in a small Swedish community.

Representativity and generalisability

The population-based group is likely to be representative of all Swedish school children with TS. Ludvika-Smedjebacken is a typical Swedish community: a middle-sized town with a total population of 40,000. The socio-economic status in Ludvika-Smedjebacken is comparable to the Swedish average. The number of higher educated people is lower, but the average income is the same as in the whole country. There is no great difference concerning the proportion of immigrants (statistics Sweden 2005).

The study was carried out through a careful case identification procedure (three-stages); tic identification, interview, and clinical assessment. The findings are based on published criteria for diagnosis (DSM-IV) and on reliable measures.

Non-response

About one third did not answer the screening questionnaire. Furthermore, 60 children of those who had TT did not want to have a clinical examination. Before the start of the study I informed the staff at outpatient service of child and adolescent psychiatry, outpatient paediatric clinic, habilitation centre, the school authorities and the school medical service about the study. Then a written information was sent to the children and their parents. Therefore, probably all the children with tics, which affect their function, have answered. This I have already seen in the parent’s answers. In the third round of the tic identification there was only one child with TT who was identified. Thus I believe it was sufficient enough to ask every fifth child who had not answered the earlier two rounds. The reason for asking only every fifth child in the third round was to save time and resources.
As a result of the decision of the ethics committee, I was not allowed to ask the teachers about their pupil’s tics in the main study. Some cases may on the one hand have been missed. On the other hand, the number of missed cases would be small as the parents were the best reporters of their children’s tics, according to results from the pilot study. The screening questionnaire was apparently easy for the parents to complete, although some were not native Swedish speakers. Professional translators helped the parents who did not speak Swedish. It was, therefore, probably that very few cases were missed due to insufficient understanding of the tic-screening questionnaire.

Validity and reliability aspects

Diagnostic criteria and procedure

The criteria of the DSM-IV were used throughout this study in assigning psychiatric diagnoses. I chose to use the DSM-IV criteria rather than the DSM-III, DSM IV-TR or ICD-10 because it is clinically relevant to take impairment and distress into account when making a diagnosis. If the DSM-III, ICD-10 or DSM IV-TR had been used, a higher prevalence of children diagnosed with the disorder would probably have been found.

The diagnosis was considered positive only if a consensus was achieved that criteria met to a degree that would consider the child to be impaired by the diagnostic symptoms in daily life activities. When it was difficult to make a diagnosis, the child was either discussed with the habilitation centre team (child neurologist, psychologist, physical and/or occupational therapist, special teacher/speech pathology therapist) or with my supervisor, professor A-L von Knorring. In a few cases the child was referred to the super-specialized child psychiatry centre at Uppsala University Hospital in order to get a more accurate evaluation.

The tic screening questionnaire was developed on the basis of a literature review, clinical experience, and DSM-IV criteria. This questionnaire had been used in another town before this study. I used many different rating scales, such as Conners, ASSQ, CDI, CY-BOCS, CBCL, which have been well validated in many countries.

All interviews and examinations, except two, which had been done by professor A-L von Knorring, were performed by myself during the course of about four years. The author was not blind to the subjects and no specific test of inter-rater reliability was done in connection with this study. However, most children with TS have previously been investigated by other physicians and almost two thirds by the same one. The fact that the same physician performed both the telephone interview and the clinical assessment also give strength to the study. It reduces measurement errors and false positive cases to a minimum.
Both the semi-structured questionnaires used for eliciting information and the schedule for psychiatric and physical/neurodevelopmental examinations have been subjected to inter-rater reliability studies (91, 92). These two instruments have been used in many Swedish studies. At the time of data collection no structured psychiatric interview covering all the relevant DSM-IV diagnoses had been translated into Swedish. However, this has been taken into account in this study as the diagnoses were made after consensus by the team evaluating the child.

The small sample size of tic disorders can hide some of the differences not found between the groups due to type-2 errors. This sample is, however, representative of Swedish school children.

The control group was selected to match the TS group for age, sex and school. They were examined in exactly the same fashion as the tic disorders groups in the three studies (I, II, III), which increases the validity of the findings obtained.

No control group was used for the cognitive tests. Swedish norms were available for most of these tests, except in the case of the RCFT, Australian norms were used. Although there is no reason to suspect cultural differences in the rather basic functions measured with this test, non Swedish norms are clearly not ideal. Further, there was large internal attrition with regard to cognitive tests, with the exception of WISC III, and to the self-perception test. However, as the missing data in many cases in study IV was due to the failure, despite attempts, to secure valid test results from these very troubled children, it is highly unlikely that the problems of my TS children were inflated. Rather, they may have been deflated.

Main findings

Prevalence (I)

This study has identified prevalence estimates for TS and other tic disorders that are higher than those generally considered in both regular and special schools. The one-year prevalence of TS in 7-15 year-old school children in this study was 0.6%. Another 0.8% had CMT and 0.5% had CVT. As many as 4.8% had TT within the past year. In total, slightly less than 7% of the school children were found to have some kind of recent tic disorder. The prevalence figures are calculated for the total school population (4,479), irrespective of that about a third did not answer the questionnaire. If only those who answered had been counted the prevalence would be still higher.

The present study showed a prevalence close to the prevalence of TS found in some studies (25, 26, 31). This study shows a frequency higher than the Israeli study (33) but lower than another Swedish study (30). The differences can be explained by the different ages included in the two studies. This
study had a wider age range (7-15 years) than the Israeli study (33), which
studied a population of conscripts 17 years old, and the other Swedish study
(30) included much younger children 7 years. As the prevalence of TS de-
creases with age this can explain why the Israeli study found a lower fre-
quency and the other Swedish study found a higher frequency than I did.
However, my study showed a prevalence peak during the first school years
in line with others. There are also other differences between the above men-
tioned studies. Apter et al. (33) used point prevalence measures and Comings
(26) used a 2-year observation time. The inclusion criteria in my study were
that tics had occurred some time during the last 12 months.

Compared with the studies by Mason and coworkers (29) and Kurlan and
colleagues (23), my prevalence is much lower. This results from the fact that
Mason and collaborators used DSM-III criteria and I used DSM-IV criteria.
The DSM-IV requires that the tics cause a marked disturbance or significant
functional impairment, which DSM-III does not. Kurlan and colleagues (22)
found a lower prevalence when they used DSM-IV criteria. When the doubt-
ful cases were excluded, the results are similar to my study.

The present study also found a lower frequency of TT than earlier studies,
which could be explained by the fact that the earlier studies used DSM-III
criteria. The definition in DSM-III requires a minimum duration of two
weeks while the DSM-IV criteria have a minimum duration of four weeks.
The proportion of children with TT not participating in the study was con-
siderably high. They did, however, not differ from the children with transient
tics participating in the study.

Sex Ratio (I)
The male/female ratio of TS in the present study was 9:1, which is compara-
ble with other studies (32, 34). Some studies have found both a lower rela-
tive frequency of males (30, 31, 33) and a higher (35). Although the ratio
differs greatly in different studies, they all show that males are more likely
than females to suffer from this disorder. The dominance of males may be
caused by the influence of sex hormone on the central nervous system in the
early development course (55).

Symptoms (II)
Motor and vocal tic symptoms showed a consistent pattern with previous
studies (3, 14, 32, 67). Motor tics, especially those of the head, were usually
the first symptoms to manifest. The onset of TS typically occurs at 4 to 5
years of age for children with TS and significantly later, at 7 to 8 years, for
children with chronic motor or vocal tics. Symptoms in the general popula-
tion were similar to those described in clinical populations. Children with
severe TS had a younger age of onset of the disorder, but this correlation was
not found in chronic motor/vocal tics. Retrospective information about the
onset of tic symptoms is not always totally reliable. So the information about onset in this study can be questionnable.

Coprolalia occurred in 44% of children with TS. The prevalence of coprolalia in this study was comparable with some other studies (17, 30, 31, 67). The lowest prevalence of coprolalia (4%) was found in a Japanese study of prepubertal children in a pediatric neurological clinic (16). On the other hand, a much higher prevalence (43%) was found in another Japanese study, that included adolescent and adult with TS in psychiatric clinics (67). So it seems as if the cultural aspect of the symptoms of TS are not very important.

Familial loading (II)
A high heredity for psychiatric illness was found in first grade relatives, which has also been reported in earlier studies (43-45). Eighty percent of the TS children had parent or sibiling who suffered from different psychiatric illnesses. The most common were tic disorders, ADHD, OCD and depression. The very high familial loading of tics and OCD supports the earlier proposal that the cause of Tourette syndrome is highly genetic. Family studies are never conclusive concerning the genetic basis of disorders. In order to study gene and environmental factors other methods are needed i.e., twin studies and molecular genetic studies.

Pre/perinatal events (II)
No increase of pre-, peri- and neonatal complications were found in children with TS. This is opposite to the findings of Leckman (53). His group found that the degree of maternal stress and severity of nausea and vomiting in the first trimester might be important. There was, however, no control group, so it is not possible to know whether there are more pregnancy complications in children with TS than in healthy children.

Comorbidity (III)
The results showed that children with TS differed from other children with other tic disorders and controls in a wide range of the psychiatric comorbidity. TS children had higher rates of ADHD, PDD, depression, DCD, OCD and sleep disorder. Similar findings have been reported by others (17, 30, 63, 65, 106-108).

Tic disorders, obsession and compulsion
Almost one third of the subjects with TS had OCB and more than half of them suffered from OCD. Many have reported rates of OCD features in 3-85% of individuals with TS (30, 33, 62, 63, 107-109). This wide range reflects differences in the populations assessed, the diagnostic criteria em-
ployed and evaluation methods used. The high prevalence of OCD/OCB symptoms has been observed not only in clinical samples of TS, but also in community samples of persons identified with tics.

The present study showed a lower frequency than other community studies, (30, 33). Apter (33) reported that 42% of 12 Israeliie military recruits found to have tics had associated obsessive-compulsive symptoms. Kadesjö (30) reported that 38% of school children with TS had major compulsive behaviours. In contrast, my study has shown a higher rate than Kurlan and colleagues (63) and Caine and colleagues (32). Kurlan reported that 11% of TS children have OCD and Caine found that about 50% of children with TS had OCB, but only 7% had OCD. My study, like Kurlan’s, was restricted to children and to those subjects meeting full DSM-IV criteria for OCD. Probably the symptoms of OCD may not have appeared yet. Several studies (110-112) suggested that OCD in TS changes with age and the duration of TS. The frequency of OCD increases with the duration of TS. The sample of TS in my study was young and had a relative short history of tics, which can be another explanation of the low prevalence of OCD. This is supported by the finding that most children with OCD/OCB were twelve years or older.

**Tic disorders and ADHD**

Two thirds of TS and one third of CVT children met the criteria for ADHD and had major learning/school problems. The rates of comorbidity between TS and ADHD identified in this sample were consistent with the findings in other studies (29, 30, 63, 113-116). The only exception is the Israeli study (33) reporting ADHD in 8.3% of conscripts with TS, which can be explained by the older age and the screening procedure.

**Tic disorders and pervasive developmental disorders**

I found 20% with pervasive developmental disorders (PDD) in TS, which is comparable to previous studies (30, 101, 116). Two children with TS had, previous to the study, been diagnosed with Asperger syndrome. One of them also had a comorbid ADHD diagnosis. Three children with TS had a first-degree relative with pervasive developmental disorders.

**Tic disorders and learning problems**

Dyslexia had previously been diagnosed in 20% of the TS children. However, 80% of TS children without a diagnosis of specific learning disorders had school related difficulties. This is similar to the finding by others (23, 60, 66, 108,118).

**Tic disorders and other problems**

This study found that one fifth of the children with TS suffered from current depression. This is comparable to a previous study (119). The pattern of comorbidity in children with CVT was similar to children with TS but CVT
children had more language disorders (21%) and four also had Down syn-
drome (DS). In this study no child with TS had DS. Kerbeshian et al re-
ported 2% of TS children with DS (120).

Overall levels of behavioural and emotional problems
Total problem scores according to CBCL rates in children with TS were
considerably higher than the mean score found in Swedish children of the
general population (121). The children with TS were more likely to be af-
flicted with inattention, social problems, aggression and delinquent behav-
iours. Internalising and externalising symptoms were also increased. This in
line with other investigators who have found an increased frequency of ag-
gression, attention and social difficulties in subjects with TS (63, 122, 123).

This study did not find any significant correlation between tic severity
and CBCL scores, which is in line with some studies (30, 116) but opposite
to other clinical studies reporting significant association of tic severity and
behavioural disturbance (114, 122). I did not find any correlation between
CDI and CBCL scores. However, there are differences between the various
tests used to quantify depressive traits. The CDI explores thoughts and de-
pressive feeling occurring in the two weeks preceding the test, while the
CBCL considers a longer period of time (the past sex months). Another im-
portant element is that the CDI is a self-assessment questionnaire, filled di-
rectly by the child and that the CBCL is a questionnaire filled in by parents.

Cognition and self-perception (IV)
One of the most important findings was that TS, for many children, was
associated with subnormal cognitive abilities and negative self-perception.
Children with TS attained slightly below-average WISC-scores. Twenty per-
cent had an IQ lower than 71, but only 16% received a diagnosis of mental
retardation (MR). This was because one child had IQ 70 in WISC but, when
the team at the habilitation centre evaluated him he had no limitations in
adaptive functioning. When he was tested with Leiter’s test he had IQ 85.
However, 16% percent of children with TS had MR, which is 20-40 times
higher than the prevalence of MR in the Swedish population (124-126).

The TS group demonstrated no discrepancy between VIQ-PIQ, thus sup-
porting some earlier findings (78, 79, 85) and contrasting with others (3,
127). However, the profile of index scores on the WISC factors was some-
what uneven, with the freedom from distractibility and processing speed
factors presenting the lowest median scores. This is in line with the findings
of previous studies of attention deficits in the TS group, attention being
measured with complex tasks (74, 76, 85, 128). Thus, the present data gave
additional evidence of attention problems among children with TS. How-
ever, there is controversy to be found in literature, whether these problems
are the results of the neuropathology of the TS disorder (84) or of ADHD
comorbidity in TS patients (77, 86). Other studies (130, 131) found that individuals with pure TS had difficulty with timed executive tests, while the ADHD comorbid group had problems with tasks necessitating the ability to maintain “preparedness to act”. Singer and co-workers (131) concluded that cognitive slowing (bradyphrenia) is the executive functions (EF) problem specific to TS, and that it is a deficit independent of comorbidity. The WISC profiles in this study indicate both speed and attention problems in the TS group, but due to the small sample size and the high prevalence of ADHD comorbidity, I could not address the issue of pure TS versus ADHD comorbid TS in relation to these deficits.

I hypothesised that patients with TS would show weak performance also in tests of executive functions. This was in part confirmed. Verbal fluency, memory and spatial organization tests were significantly impaired in the children with TS, which is comparable to previous studies (76, 85, 127). Again, however, the high ADHD comorbidity rates do not allow conclusions with regard to the effects of TS itself, since EF deficits are very common in children with ADHD (132).

Children with TS had less positive self-perception in several respects than did children in the comparison groups, pointing to severe social and emotional consequences of the disorder. The vulnerability in terms of poor peer relations was demonstrated in a study by Bawden and co-workers (72) finding that children with TS were consistently ranked low in terms of popularity and were at increased risk for poor peer relationships. The younger children in this study were also unsatisfied with their social relations. Higher performance IQ was associated with more negative self-perception. Another study has reported similar results (71). Perhaps, TS children with adequate cognitive abilities may be particularly aware of their difficulties and feel badly about themselves. It should be noted that negative self-perception is found also among children with ADHD (134). So again, the effect of ADHD comorbidity is unclear.

Relation between medication and IQ
Children who were taking medication had lower full IQ scores than children who were not. These results may be attributed to medication effects or to the neuropathology of severe TS. It should be noted that while medication for children with emotional and behaviour disorders is quite rare in Sweden, almost half of the present sample were receiving pharmacological treatment. These children took their medicine because they had severe symptoms, and they had also responded with reduction in symptoms. Probably, the lower IQ among children receiving medication was due to the severity of the disorder. Previous studies have shown that the common medications given to patients with TS have no impact on their neuropsychological test performance (133, 135). Also, as discussed above, many children in this study had ADHD, and
it should be remembered that children with TS and comorbid ADHD often have low IQ (74, 85).

Relation between tic severity and IQ
As regards tic severity and age at onset, I did not find any relation to any of the cognitive measures or to self-perception, which lends some support to the negative findings regarding these aspects of the disorder and relations to IQ (87). However, efforts to clarify the role of tic severity and the duration of symptoms for various outcomes should continue, as other studies have found such effects (79).

Comments
All except one (96%) with TS had been in contact with medical services. About a third had been in contact with child psychiatry prior to the study, but surprisingly none of them had a diagnosis of Tourette syndrome. It is well known from earlier clinical descriptions that recognition and diagnosis of tics and Tourette syndrome often occur very late (136, 137). This was also the case in this study; e.g. 1/3 had no diagnosis of TS prior to the study, although they had tried to find help for different kinds of symptoms and problems. The results of the present study gave even more evidence of this.

Such relatively high misdiagnosis indicates that the medical professional did not have complete understanding of this disorder or that the parents did not recognize their children’s abnormal behaviour. The majority of children with TS are not diagnosed in their school settings and 40% reported they were never counselling with school psychologist and 20% reported that counselling was provided but did not help.

Walter and Carter (138) suggested that the school psychologist has a valuable role to play when it comes to behaviour modification of tics because school psychologists are ideally positioned to collect necessary information to help develop a plan and then to monitor it efficiency. School psychologist also have a valuable role to play in other ways, they support the teacher in service of the child.

Less than half the children with chronic tics and only a minority with TT (12%) had sought help from medical services. One explanation of this might be that the tics were mostly mild for children with CMVT and very mild for those with TT and the contact with health services was due to other causes than tics, such as delayed psychomotor development, attention problems and learning difficulties.
Conclusions

Tourette syndrome and other tic disorders are common in childhood. The prevalence is higher in younger school-age children and in boys. The highest prevalence is in special schools/classes. Tic severity is correlated with early age of onset. In a total population of children with TS almost all have sought help from medical services, but this counseling was generally not perceived as being very helpful.

A high rate of psychiatric disorders is prevalent in parents and siblings of children with Tourette syndrome. The findings draw attention to the importance of thorough investigation of both parents and siblings of patients with TS for evidence of tics, OCD, depression, or ADHD.

Psychiatric comorbid disorders are common even in community-based samples of children with TS and chronic tic disorders (CT). These findings seem to indicate that TS and CT are part of the same disease entity, TS being a more severe and complex form of tic disorder.

Tourette syndrome in the population is associated with negative self-perceptions and problematic cognitive abilities although, in the latter case, there are large individual variations. Clinicians need to be aware of the cognitive deficits that may pose a greater obstacle for children with tics than the tic disorder itself.

One of the most important findings of this study is that behavioural, learning, and psychiatric problems, which can be, associated with tics in children and which should lead to thorough investigation for the presence of comorbidities, in children with chronic tics.

Early behavioural interventions to treat the comorbid conditions of TS symptoms should be undertaken. It would seem that interventions such as mental health counseling are necessary. These children need to be identified, diagnosed, treated and supported for many years.

The findings of this study imply that TS is a heterogeneous neuropsychiatric disorder. The pattern of comorbidity reflects the possible neurobiological and genetic links between chronic tic disorders, ADHD and OCD.

Recommendations

Tourette syndrome (TS) is more common in schoolchildren than has been hitherto accepted. School health authorities and primary care need to be always aware that TS is a common problem. They should be able to identify children with TS at a considerably earlier age than is generally the case today and thereby make a contribution to the development of adequate support and treatment facilities for the children and their families. Earlier identification and intervention in children with different tic disorders may reduce the level of impairment and help reduce the high rate of psychosocial and aca-
demic dysfunction evident in many children with TS through childhood and into adult life.

The clinicians need to be more aware of the behavioural, learning and psychiatric problems that can be associated with tics in children. This awareness should lead to thorough investigation for the presence of comorbidities, which are often much more impairing than tics per se. Thus, tics should serve as “a red flag” when evaluating children.

This study supports the need for vigilance concerning TS in school-age children in primary care and educational settings, so that children with a potentially serious disorder can be identified and assessed. Then effective management packages can be formulated to address their needs and develop a multi-faceted and integrated approach to plan an individual education programme for the student with TS. A multidisciplinary therapeutic programme must be established in close collaboration with the child and its parent’s.
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Svensk sammanfattning

Tics och Tourette syndrom hos grundskolebarn

Bakgrund


Förlöpet av TS under uppväxttiden är mycket varierande, från mycket svåra utbredda frekventa tics under långa perioder, till diskreta glesa besvär som kan försvinna efter adolescensen.

Den första kända kliniska beskrivningen av TS härrör från Itard. Itard beskrev i Frankrike 1825 en markisinna som vid 7 års ålder började få motoriska och vokala tics. Syndromet fick sitt namn efter förslag av Jean Martin Charcot för att hedra Georges Gilles de la Tourette som 1885 var den förste att publicera ett samlat patientmaterial.

Den moderna Tourette forskningen började på 1960-talet efter det att man i Europa publicerat fall, där man lyckats behandla patienter med TS med Haloperidol. Detta ledde till stor optimism angående TS, att det var en neurokemisk balansstörning som skulle kunna botas med hjälp av medicin.


Den aktuella synen på TS kan sammanfattas som följande: En genetisk grundad vulnerabilitet som berör utvecklingen av ett system centrat i hjärnan omfattande basala ganglier, särskilt striatum, delar av limbiska systemet samt sensori-motoriska cortex. Framför allt dopaminerga och serotoninerga bansystem är engagerade. Den genetiska vulnerabiliteten påverkas av miljöfaktorer som man tror skall vara både av infektiös natur som t.ex Streptokockinfektioner (i analogi med Sydenhamns Chorea), psykosocialstress och
könhormonmedlierade. Flera undersökningar tyder på att det finns ett samband mellan TS och andra sjukdomsgrupper som tvångssyndrom, ångestsyndrom, affektiva syndrom, autistiska syndrom, ADHD/DAMP och inlärningssvårigheter.


De epidemiologiska undersökningarna om TS och tics som hittills finns har brister. Prevalensen av TS anslås enligt olika undersökningar mellan 0,05-3 procent. En populationsbaserad undersökning av barn har funnit en prevalens av tics på ca 10-20%. Många rapporter visar att TS är vanligt i grupper med specialundervisning, upp till 1/3 av eleverna där har TS.

Frågeställningar
1- att kartlägga epidemiologien av tics och TS hos grundskolebarn i Sverige
2- att studera backgrundsfaktorer samt beskriva symptomatologin hos barn med tics och TS
3- att studera förekomsten av tilläggsproblem i form av uppmärksamhetstörningar, tvångstankar och tvångshandlingar, inlärningssvårigheter, språkproblem och beteendeavvikelse hos barn med tics och TS
4- att studera de kognitiva funktioner och självkänsla hos barn med TS

Material
Grundskolepopulation (n= 4479) i Ludvika/Smedjebacken, åk 1-9, ålder 7-15.

Metod

Föräldrar, lärare och elevvården informerades om tics och om studien. Barnen tillsammans med sina föräldrar fick fylla i tics screening formulär. Alla barn som har svarat att de har tics kallades till skolhälsovårdslokalen, för intervju och läkarundersökning, för att bekräfta eller avvisa diagnosen tics/TS. DSM IV kriterier har använts. Barnen blev undersöpta av under-tecknad. Barnen delades upp i TS, kronisk motorisk tics (KMT), kronisk

Resultat

Min undersökning visade att förekomsten av TS är 0.6% i grundskolepopulationen men 0.8% hos de screenade barnen. Detta är vanligare än vad man tidigare har rapporterat. Däremot tycks förekomsten av övergående tics vara mindre (6.6%) än vad som angivits i tidigare internationella studier.


Däremot har barn med övergående tics inte mer psykiska problem än kontrollbarnen. Barnen med TS har kognitiva problem och dålig självkänsla jämfört med populationen i övrigt. 20% av barnen har ett IQ ≤ 70, men det finns också barn som har IQ > 130. Till yttermera visso har många med TS inte fått adekvat hjälp i skolan eller av sjukvården trots att alla utom en har kontakt med sjukvården.

Sammanfattning

1. Tourette syndrom och de andra tics tillstånd förekommer hos grundskolebarn population 6.6%. TS förekommer mycket mindre än de andra tics tillstånd (0.6%), den är mer vanlig än man har trott. De yngre barnen har högre tics/TS prevalens och speciellt hos pojkare. Högsta prevalensen av tics/TS är hos särskolebarn (46%).
2. Ticssvårighetsgrad är relaterad till tidig debut ålder.
3. Föräldrar och syskon till Barn med TS har hög frekvens av neuropsykiatriska tillstånd. Detta bör uppmärksammas för att kunna erbjuda dem den hjälp de behöver.
4. Ingen signifikant skillnad mellan TS och kontroll grupp med avseende pre/perinataala faktorer.
5. Barnen med TS lider av andra psykiatriska och neurologiska diagnoser som i sig kan vara svårare än själva ticsen.
7. Barn med TS ska upptäckas tidigt för att de ska få adekvat pedagogik och rätt hjälp från sjukvården annars finns det stor risk att de får svåra psykiska symtom och misslyckas i skolan.
References

1. Itard JMG (1825). Memoire sur quelqes fonctions involontaires des appareils de la locomotion de la prehension et de la voix. Arch Gen Med 8:385-407


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Appendix I
Formulär om Tics till lärare

Barnets namn ......................................... ålder: år.............. mån .........

Skolklass .................................................. Skola .........................

Pojke □    Flicka □

Formuläret ifyllt av ....................................................
Nedan finns en lista på olika typer av vanor och tics som barnet kan ha haft det senaste året. Det finns tre svarsalternativ: Säkert, Kanske och Inte alls. Sätt ett kryss (X) för det alternativ som Du tycker stämmer bäst.

Har barnet under året haft?

<table>
<thead>
<tr>
<th></th>
<th>Säkert</th>
<th>Kanske</th>
<th>Inte alls</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Upprepade ryckningar och liknande i ansiktet, t ex blinkningar, grimaser, räcka ut tungan, slic-kande runt munnen, spottningar (motoriska tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Upprepade ryckningar och liknande i nacken, axlar eller bål, t ex vridningar, nickningar, axelryckningar, vridande i bålen (motoriska tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Upprepade ryckningar och liknande i armar, händer, ben eller fötter, t ex handklappningar på olika ställen på kroppen på sig själv eller andra, sparkar med benen eller fotterna (motoriska tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Upprepade ljud av olika slag, t ex hosta, harkla, fräsa, snörvla eller lätten som liknar djurljud som skälla, gny, gnäll (vokala tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Upprepade ord och fraser som t ex svordomar, könsord, kränkande eller andra fula ord (vokala tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Andra annorlunda ritualer eller vanor.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Hur länge har barnet haft tics? Beskriv
Har eleven stödundervisning? Ja □    Nej □
Kommentar .......................................................
Appendix II

Formulär om Tics till föräldrar

Barnets namn ……………………………. ålder: år………….. mån ……..

Skolklass ……………………………………… Skola …………………

Pojke □  Flicka □

Formuläret ifyllt av ……………………………..……………………………

Nedan finns en lista på olika typer av vanor och tics som barnet kan ha haft det senaste året. Det finns tre svarsalternativ: Säkert, Kanske och Inte alls. Sätt ett kryss (X) för det alternativ som Du tycker stämmer bäst.

<table>
<thead>
<tr>
<th></th>
<th>Säkert</th>
<th>Kanske</th>
<th>Inte alls</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Upprepade ryckningar och liknande i ansiktet, t ex blinkningar, grimaser, räcka ut tungan, slic-kande runt munnen, spottningar (motoriska tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Upprepade ryckningar och liknande i nacken, axlar eller bål, t ex vridningar, nickningar, axelryckningar, vridande i bålen (motoriska tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Upprepade ryckningar och liknande i armar, händer, ben eller fötter, t ex handklappningar på olika ställen på kroppen på sig själv eller andra, sparkar med benen eller fötterna (motoriska tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Upprepade ljud av olika slag, t ex hosta, harkla, fräsa, snövlra eller läten som liknar djurljud som skälla, gny, gnäll (vokala tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Upprepade ord och fraser som t ex svordomar, könsord, kränkande eller andra fula ord (vokala tics).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Andra annorlunda ritualer eller vanor.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Hur länge har barnet haft tics? Beskriv

Har eleven stödundervisning? Ja □ Nej □

Kommentar …………………………………………………………………

Najah Khalifa, 1998
## Appendix III

### Tourette Syndrome Severity Scale (TSSS)

<table>
<thead>
<tr>
<th>Tics noticeable to others</th>
<th>Tics elicit comments or curiosity</th>
<th>Patient considered odd or bizarre</th>
<th>Tics interfere with functioning</th>
<th>Total score sum of ratings</th>
<th>Severity rating</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non present (0)</td>
<td>_</td>
<td>_</td>
<td>-</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Very few (0.5)</td>
<td>No (0)</td>
<td>No (0)</td>
<td>No (0)</td>
<td>0.5</td>
<td>Very mild</td>
</tr>
<tr>
<td>Some (1)</td>
<td>No (0)</td>
<td>No (0)</td>
<td>No (0)</td>
<td>1- &lt; 2</td>
<td>Mild</td>
</tr>
<tr>
<td>Most (2)</td>
<td>Yes (1)</td>
<td>No (0)</td>
<td>No (0)</td>
<td>2- &lt; 4</td>
<td>Moderate</td>
</tr>
<tr>
<td>All (3)</td>
<td>Yes (1)</td>
<td>Possibly (1)</td>
<td>Might (1)</td>
<td>4- &lt;6</td>
<td>Marked</td>
</tr>
<tr>
<td>All (3)</td>
<td>Yes (1)</td>
<td>Yes (2)</td>
<td>Yes (2)</td>
<td>6 - 8</td>
<td>Severe</td>
</tr>
<tr>
<td>All (3)</td>
<td>Yes (1)</td>
<td>Yes (2)</td>
<td>Yes (2)</td>
<td>9</td>
<td>Very severe^a</td>
</tr>
</tbody>
</table>

^aAll of above in addition to being incapacitated, requiring hospitalization, or remaining at home.
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