Information from your family doctor. Tourette's syndrome: what it is and how it's treated.

**American Academy of Family Physicians.**

Publication Types:

- Patient Education Handout

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Tourette's syndrome.

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Tourette's syndrome is a movement disorder most commonly seen in school-age children. The incidence peaks around preadolescence with one half of cases resolving in early adulthood. Tourette's syndrome is the most common cause of tics, which are involuntary or semivoluntary, sudden, brief, intermittent, repetitive movements (motor tics) or sounds (phonic tics). It is often associated with psychiatric comorbidities, mainly attention-deficit/hyperactivity disorder and obsessive-compulsive disorder. Given its diverse presentation, Tourette's syndrome can mimic many hyperkinetic disorders, making the diagnosis challenging at times. The etiology of this syndrome is thought to be related to basal ganglia dysfunction. Treatment can be behavioral, pharmacologic, or surgical, and is dictated by the most incapacitating symptoms. Alpha2-adrenergic agonists are the first line of pharmacologic therapy, but dopamine-receptor-blocking drugs are required for multiple, complex tics. Dopamine-receptor-blocking drugs are associated with potential side effects including sedation, weight gain, acute dystonic reactions, and tardive dyskinesia. Appropriate diagnosis and treatment can substantially improve quality of life and psychosocial functioning in affected children.

Publication Types:

- Review

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Developmental psychopathology of children and adolescents with Tourette syndrome—impact of ADHD.

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BACKGROUND: In Tourette syndrome (TS) as a neurodevelopmental disorder not only the tics but also the comorbid conditions change with increasing age. ADHD is highly comorbid with TS and usually impairs psychosocial functioning more than the tics. Its impact on further comorbidity during development is important for clinical practice and still a matter of debate. METHOD: Aspects of developmental psychopathology considering the impact of ADHD were examined by logistic regression (year wisely) in a cross-sectional sample of children and adolescents (n = 5060) from the TIC database. RESULTS: In TS+ADHD (compared to TS-ADHD) higher rates of comorbid conditions like OCD, anxiety disorders, CD/ODD and mood disorders were found in children (5-10 years). In adolescents (11-17 years) higher comorbidity rates in TS+ADHD remained only for CD/ODD and mood disorders. Accordingly, for OCD and anxiety disorders there was a steeper year wise increase of these comorbidities in TS-ADHD while it was a similar for CD/ODD and mood disorders in TS-ADHD as well as TS+ADHD. CONCLUSION: Children with TS+ADHD have more comorbidities than the TS-ADHD group, whereas in both adolescent groups this did no longer hold for OCD and anxiety disorders. These findings indicate that in TS comorbid ADHD is associated with high rates of externalizing and internalizing problems, whereas TS without ADHD is associated only with internalizing problems in adolescence.

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**Tic disorders and ADHD: answers from a world-wide clinical dataset on Tourette syndrome.**

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BACKGROUND: Tourette syndrome (TS) is a neurodevelopmental disorder with frequent comorbidity with Attention-deficit-Hyperactivity disorder (ADHD). The impact of this association is still a matter of debate. METHOD: Using the TIC database containing 6,805 cases, the clinical differences were ascertained between subjects with and without ADHD. RESULTS: The reported prevalence of ADHD in TS was 55%, within the range of many other reports. If the proband was diagnosed with ADHD, a family history of ADHD was much more likely. ADHD was associated with earlier diagnosis of TS and a much higher rate of anger control problems, sleep problems, specific learning disability, OCD, Oppositional-defiant disorder, mood disorder, social skill deficits, sexually inappropriate behaviour, and self-injurious behaviour. Subjects with seizures and with Developmental Coordination Disorder also had high rates of ADHD. Anxiety disorder, however, was not more frequent. Preliminary data suggest that most behavioural difficulties in ADHD are associated with the Combined or Hyperactive-Impulsive Subtypes of ADHD. Every large site (>200 cases) had a significantly increased rate of anger control problems in cases with ADHD. CONCLUSION: Subjects with TS have high rates of ADHD and complex associations with other disorders. Clinically the findings confirm other research indicating the importance of ADHD in understanding the behavioural problems often associated with the diagnosis of TS. Additional ADHD comorbidity should be taken into account in diagnosis, management, and training.

PMID: 17665279 [PubMed - indexed for MEDLINE]

**Neural correlates of tic severity and cognitive control in children with Tourette syndrome.**

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Tourette syndrome (TS) is a neurodevelopmental disorder characterized by involuntary motor and phonic tics. It is hypothesized that excess dopamine leads to an imbalance in the pathways through the basal ganglia, resulting in unchecked movements via thalamic disinhibition. It has been unclear whether TS is associated with cognitive control deficits as well as pure motor control deficits, or whether cognitive deficits are associated with the presence of comorbid conditions. Furthermore, little is known about the neural underpinnings of TS in childhood, prior to the long-term effects of medication on brain function. Here, children with TS and typically developing children performed a cognitive control task during
event-related fMRI data acquisition. The study included 18 native English-speaking 7-13-year-old children with TS (M = 10.42; 15 males), and 19 healthy, age-matched native English-speaking volunteers (M = 10.33; 11 males). The task involved three separate manipulations of cognitive control. Behaviourally, higher tic severity was correlated with slower task performance on the most demanding task conditions. Neurally, higher tic severity was associated with enhanced activation of dopaminergic nuclei (substantia nigra/ventral tegmental area) and cortical, striatal and thalamic regions in the direct pathway. Heightened tic severity was also associated with greater engagement of the subthalamic nucleus area, suggestive of a compensatory mechanism. Overall, patients engaged left prefrontal cortex more strongly than typicals during task performance. These data suggest that children aged 7-13 unmedicated for TS exhibit increased activation in the direct pathway through the basal ganglia, as well as increased compensatory activation in prefrontal cortex and the subthalamic nucleus.

Publication Types:

- Research Support, N.I.H., Extramural
- Research Support, Non-U.S. Gov't

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Factors influencing diagnosis delay in children with Tourette syndrome.

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BACKGROUND: Tourette syndrome (TS) is a chronic disorder characterized by motor and vocal tics. Previous studies reported a substantial lag period between disease onset and diagnosis ranging from 3 to 11.9 years. AIMS: To determine the lag period and factors associated with diagnosis delay of TS. METHODS: All files of 185 children with TS attending one neuropediatric unit in Jerusalem were reviewed. Lag time between disease onset, according to DSM criteria, and diagnosis was determined and the contributions of the disease course, comorbidities and epidemiological factors were assessed. RESULTS: A relatively short lag to diagnosis following the onset of diagnosable TS was documented (mean 13.2 +/- 15.9 months, median 6 months). A relatively longer gap was associated with older age at TS onset (r=0.161, p<0.05) and vocal tics as the first manifestation rather than motor or combined motor and vocal tics (mean=20.3+16.3 months vs 11.9+16.5 and 12.6+15.2, respectively, p<0.05). A relatively shorter gap was associated with tic severity (r=0.13, p<0.05) and presence of comorbid obsessive-compulsive disorder (OCD) (9.5+14.7 months vs. 14.1+16 without OCD, p<0.05). CONCLUSIONS: Lag time to diagnosis is relatively short in our population. Factors associated with a shorter lag (early age of TS onset, motor tics as the first manifestation, greater tics severity and the presence of OCD) may be perceived as disruptive, prompting patient and families to seek medical care. Conversely, vocal tics as the first manifestation, associated with a longer lag, may be misdiagnosed as features of common pediatric conditions, thus delaying diagnosis.

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Clinical analysis of Gilles de la Tourette syndrome based on 126 cases.

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BACKGROUND AND PURPOSE: Gilles de la Tourette syndrome (TS) is characterized by the presence of multiple motor and vocal tics, as well as other neuropsychiatric disorders. The aim of the study was to evaluate the frequency of particular clinical symptoms in patients diagnosed with TS. MATERIAL AND METHODS: A hundred twenty-six individuals were studied. A brief questionnaire including data from the medical history and neurological examination was used. RESULTS: TS was much more frequent in males (80%; 101/126) than in females. The mean age at onset was 7.6 (2-17) years. The onset of the disease was usually slow. Abrupt onset of the disease, usually after infection, was noted in 11% (12/114) of patients. The mean delay in diagnosis was 3.9 years. In most patients tics were moderate (64%; 81/126). Mild and severe intensity of tics were reported in 15% (19/126) and 21% (26/126) of patients, respectively. 77% (97/126) of individuals with TS had comorbidities. The mean comorbidity score was 2.79 per patient. Anger control problems, sleep difficulties,
self-injurious behaviour and coprolalia were strongly associated with comorbidity. The most common reported comorbidity was attention deficit hyperactivity disorder (59%; 74/126). Family history was positive in 46% (57/125) of patients, most often in TS patients with onset between ages 2 and 4 years (70%; 14/20). Haloperidol was the most commonly used medication in our cohort (60%; 57/95). 22% (27/122) of patients did not receive any symptomatic treatment.

CONCLUSIONS: The appropriate diagnosis was delayed for about four years after the onset of the disease. Comorbidity and behavioural problems were frequent features of TS. Genetic factors can play an important role in the aetiology of TS.

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Neurocognitive correlates of child obsessive compulsive disorder and Tourette syndrome.

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This study investigated the neurocognitive correlates of childhood OCD and TS, which are purported to share frontal-striatal dysfunction. Neurocognitive measures tapping frontal-striatal functions such as executive, attention/memory, and visuomotor abilities were administered to three groups of participants, OCD without comorbid TS (OCD), TS without comorbid OCD (TS), and normal controls. Results suggested that OCD group demonstrated deficits in the area of spatial attention relative to healthy controls. The OCD participants demonstrated no cognitive deficits compared to the TS group. TS participants showed trends towards impairments in the areas of response inhibition, divided attention, and cognitive flexibility relative to the OCD and normal control groups. Spatial attention deficits for the OCD group are partially consistent with adult OCD studies indicating deficits in spatial memory. TS findings were less robust and may be construed tentatively as suggestive of executive function deficits. Future research is needed to delineate the influence of development on neurocognitive deficits associated with OCD and TS.

Publication Types:

- Research Support, N.I.H., Extramural

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The study addressed the issue of arithmetic deficiencies in children with Tourette syndrome (TS) as well as explanations for such deficiencies. A total of 47 children with TS were assigned to three subgroups based on a composite attention score from the Test of Variables of Attention (TOVA). These children, along with 17 normal controls between 8 and 16 years of age, were tested on standardized measures of IQ, attention, visuospatial ability, and arithmetic achievement. The children also were administered an experimental calculation task with two levels of structure. Children with TS scored below controls on tests of IQ, attention, and arithmetic achievement but not visuospatial ability. The TS subgroup with the greatest impairment of attention accounted for most of the differences in arithmetic achievement. Regression analysis, based on the 47 children with TS, indicated that IQ and TOVA scores were the best predictors of arithmetic achievement. Likewise, the experimental calculation task indicated that the poor performance of some children with TS could be attributed to deficient attention. Irrespective of structure, children in the TS subgroup with the greatest attentional impairment made more attention (but not visuospatial) errors than did controls on the experimental task. Thus, on both the standardized and the experimental tasks, poor arithmetic skill was found only in children with TS who had significant attentional deficits.

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Living with Tourette's syndrome.

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Illnesses in children are difficult enough, but for children and adolescents with Tourette's syndrome, the emergence of unwelcome motor and vocal tics in social situations can lead to not only embarrassment, but also increased self-consciousness, social isolation, thoughts of persecution, and physical pain. This article offers an overview of Tourette's syndrome and focuses on DSM-IV-TR criteria, severity, prevalence and course, etiology and epidemiology, indications for medications, psychosocial therapies, and nursing implications.

Publication Types:

- Review

PMID: 17848040 [PubMed - indexed for MEDLINE]

Parenting stress and related factors in parents of children with Tourette syndrome.

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The objective of this study was to assess the stress of parents and its influencing factors in caring for children with Tourette syndrome. A total of 150 subjects, either fathers or mothers of children diagnosed with Tourette syndrome between the ages of 6 and 12, were recruited by purposive sampling from the membership roster of the Taiwan Tourette Family Association. Study tools included a Parenting Stress Index Form and Social Support Index Form. The standardized score for parent perception of parenting stress was 83.5. The main stressor of parents of children with Tourette syndrome was found to be child care difficulties. A correlation was found between parenting stress and child gender, age, school situation and disease severity; parent age and family income. A significant negative correlation (r=-.459, p<.01) was found between social support and parenting stress. It was revealed that social support had a significant effect on parenting stress in this study. Multiple linear regression analysis found disease severity and family income to be the variables with the greatest predictive power for parenting stress, explaining 42% of total variance. Results showed that factors affecting parenting stress included family income and disease severity. These findings should help clinical professionals develop more effective health care strategies to address the needs of children with Tourette syndrome and their parents.

Publication Types:

- Research Support, Non-U.S. Gov't

PMID: 17806033 [PubMed - indexed for MEDLINE]
Chronic tic disorders are characterized by involuntary motor and vocal tics, which are influenced by contextual factors. Recent research has shown that (a) children can suppress tics for brief periods of time, (b) suppression is enhanced when programmed reinforcement is provided for tic-free intervals, and (c) short periods of suppression do not result in a paradoxical "rebound" in tic frequency when active suppression has ceased. The current study extended existing research in three important ways. First, we examined whether tic suppression ability decreased as suppression duration increased from 5 to 25 to 40 min. Second, we examined post-suppression tic frequency to test whether longer periods of suppression were more likely to be associated with a rebound effect. Finally, we explored neuropsychological predictors of tic suppression. Thirteen children with Tourette syndrome or a chronic tic disorder completed the study. Results showed that (a) tic suppression was sustained for all of the suppression durations, (b) rebound effects were not observed following any of the suppression durations, and (c) ability to suppress was correlated with omission, but not commission errors on a continuous performance task. Implications of these findings are discussed.
number analysis of the SLITRK1 gene, and haplotype analysis. We found tics or other behavioral manifestations in 15 subjects. Of these, 5 received a diagnosis of definite TS, 5 were classified as having definite CMT, 2 had definite nonspecific tic disorder, and 3 patients had obsessive-compulsive disorder without motor or phonic tics. Tics mainly involved the craniocebral district. Many patients with tics had coexisting psychiatric disorders, especially obsessive-compulsive disorder, performed poorly at school and had social problems. Direct sequencing and copy number analysis of the SLITRK1 gene, and haplotype analysis suggested that the SLITRK1 locus was not involved in this family. In conclusion, the distinctive clinical features in this family are the motor tics mainly involving the face and the neck and the severe coexisting psychiatric disorders. The negative results of the SLITRK1 analysis point to genetic heterogeneity in TS. (c) 2007 Movement Disorder Society.

PMID: 17712845 [PubMed - indexed for MEDLINE]

Related Articles, Links

Tourette syndrome and tic disorders: a decade of progress.

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OBJECTIVE: This is a review of progress made in the understanding of Tourette syndrome (TS) during the past decade including models of pathogenesis, state-of-the-art assessment techniques, and treatment. METHOD: Computerized literature searches were conducted under the key words "Tourette syndrome," "Tourette disorder," and "tics." Only references from 1996-2006 were included. RESULTS: Studies have documented the natural history of TS and the finding that tics usually improve by the end of the second decade of life. It has also become clear that TS frequently co-occurs with attention-deficit/hyperactivity disorder, obsessive-compulsive disorder, and a range of other mood and anxiety disorders. These comorbid conditions are often the major source of impairment for the affected child. Advances have also been made in understanding the underlying neurobiology of TS using in vivo neuroimaging and neurophysiology techniques. Progress on the genetic front has been less rapid. Proper diagnosis and education (involving the affected child and his or her parents, teachers, and peers) are essential prerequisites to the successful management of children with TS. When necessary, modestly effective antitonic medications are available, although intervening to treat the comorbid attention-deficit/hyperactivity disorder, obsessive-compulsive disorder and/or obsessive-compulsive disorder is usually the place to start. CONCLUSIONS: Prospective longitudinal studies and randomized clinical trials have led to the refinement of several models of pathogenesis and advanced our evidence base regarding treatment options. However, fully explanatory models are needed that would allow for more accurate prognosis and the development of targeted and efficacious treatments.

Publication Types:
• Review

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16: Mov Disord. 2007 Sep 15;22(12):1743-50.
Related Articles, Links

Malignant Tourette syndrome.

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The aim of this work was to draw attention to potentially life-threatening symptoms associated with Tourette syndrome (TS) and to explore their relationship to TS comorbidities. Medical records of all patients with TS evaluated at our Movement Disorders Clinic between July 2003 and July 2006 were reviewed. Data on patients with malignant TS, defined as >or=2 emergency room (ER) visits or >or=1 hospitalizations for TS symptoms or its associated behavioral comorbidities, were entered into a dataset and analyzed. Five illustrative cases are described. Of 333 TS patients evaluated during the 3-year period, 17 (5.1%) met the criteria for malignant TS. Hospital admission or ER visits were for tic-related injuries, self-injurious behavior (SIB), uncontrollable violence and temper, and suicidal ideation/attempts. Compared with patients with nonmalignant TS, those with malignant TS were significantly more likely to have a personal history of obsessive compulsive behavior/disorder (OCB/OCD), complex phonic tics, coprolalia, copropraxia, SIB, mood disorder, suicidal ideation, and poor response to medications. Although TS is rarely a disabling disorder, about 5% of patients
referred to a specialty clinic have life-threatening symptoms. Malignant TS is associated with greater severity of motor symptoms and the presence of >or=2 behavioral comorbidities. OCD/OCB in particular may play a central role in malignant TS; obsessive compulsive qualities were associated with life-threatening tics, SIB, and suicidal ideation. Malignant TS is more refractory to medical treatment than nonmalignant TS. (c) 2007 Movement Disorder Society.

Publication Types:
- Case Reports
- Review

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Sleep patterns in children with attention-deficit/hyperactivity disorder, tic disorder, and comorbidity.

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BACKGROUND: In children, attention-deficit/hyperactivity disorder (ADHD), tic disorder (TD), and their coexistence (ADHD + TD comorbidity) are very common and clinically important. Associated sleep patterns and their clinical role are still insufficiently investigated. This study aimed at characterizing these sleep patterns in children with ADHD, TD, and ADHD + TD comorbidity and determining whether, in ADHD + TD, the factors ADHD and TD may affect the sleep pattern in an independent (additive) or in a complex (interactive) manner. METHOD: By means of polysomnography, sleep patterns were investigated in 4 groups of unmedicated 8.0-16.4-year-old children (healthy controls, ADHD-only, TD-only, and ADHD + TD). Each group consisted of 18 subjects matched for age, gender, and intelligence. RESULTS: ADHD was primarily characterized by increase in rapid eye movement (REM) sleep, whereas TD patients displayed lower sleep efficiency and elevated arousal index in sleep. In children with ADHD + TD, both effects appeared. No interaction between the ADHD and TD factors was found for any of the sleep parameters. Significant correlations between sleep patterns and clinical symptoms were found. CONCLUSIONS: ADHD and TD are characterized by specific sleep alterations. When coexisting, the two disorders alter the sleep pattern in an additive manner, suggesting a high impact on clinical and therapeutic perspectives.

Publication Types:
- Research Support, Non-U.S. Gov't

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Tie suppression in the treatment of Tourette's syndrome with exposure therapy: the rebound phenomenon reconsidered.

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Exposure and response prevention (ER), a behavioral treatment program consisting of exposure to premonitory sensory experiences during prolonged tic suppression, was shown to be a promising new treatment for tics in Tourette's syndrome (TS). In this study, the commonly reported paradoxical increase in tic frequency following voluntary tic suppression, i.e., rebound phenomenon, was examined. Tic frequency was rated in 20 TS patients during 15-minute videotaped conversations taken both before and following 10 ER sessions. In addition, tic frequency was obtained at home by family members of the patients during 15-minute daily tic frequency registrations. Ratings following ER sessions were compared with ratings obtained before the sessions. Neither the ratings at the institute nor the ratings at home supported a rebound effect following ER tic suppression. Copyright (c) 2007 Movement Disorder Society.

A measure of functional impairment in youth with Tourette's syndrome.

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OBJECTIVE: Tourette's Syndrome (TS) during childhood is linked to varied behavioral and psychological difficulties and functional impairment. The current study was undertaken to examine both tic-related impairment and impairment from other psychological problems in 59 youth (mean age 11.4 years, 69% male) with TS. METHODS: Caretakers completed a checklist about the impact of tics and other psychological difficulties on family, school, and social functioning. In addition, a clinician administered a measure of tic severity to families. RESULTS: Over half of the sample reported one significant problem area due to the presence of tics, with over a third reporting two or more problem areas. Problems were heterogeneous in nature, with no report of a particular problem area in more than 25% of the children. The rate of nontic-related impairment was very high, with 70% of parents reporting at least one problem area. CONCLUSIONS: Treating both tic and nontic-related impairments concurrently may improve functioning more so than treating the symptoms separately.

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Case reports and review of Tourette syndrome.

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Tourette syndrome (TS) is characterized by at least one motor and one verbal tic with duration of more than one year with no 3-month period without tics. It is one of the most common childhood movement disorders and often causes significant morbidity in the children it affects. TS as well as its numerous comorbidities are often under-recognized if the clinical suspicion of the physician is not high. In this review, we describe two patients with TS that varied in their degree of symptoms and treatment. We would like to emphasize with this review that TS occurs more commonly than it is diagnosed and is often confused with other conditions, such as seasonal allergies, sinusitis, and seizures. Correct diagnosis is important in order to allow appropriate treatment and to improve the quality of life for these patients.

Publication Types:

- Case Reports
- Review

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