Tourette Syndrome[(1)]

Tourette Syndrome ("TS") is a **tic disorder** that is characterized by multiple motor and vocal tics. According to the following criteria from the Diagnostic and Statistical Manual of Mental Disorders ("DSM"), TS requires:

1. 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, nonrhythmic, stereotyped motor movement or vocalization.)
2. The tics occur many times a day (usually in bouts) nearly every day or intermittently throughout a period of more than 1 year; and during this period there was never a tic-free period of more than three consecutive months.
3. This disturbance causes marked distress or significant impairment in social, occupational, or other important areas of functioning.
4. The onset is before age 18 years.
5. 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).

TS occurs in approximately 1 in 2000 people. The syndrome is four times more common in boys than in girls, and more frequent among family members than in the general population (there may be a form of the syndrome that is inherited in an autosomal dominant pattern).

Tics may be voluntarily suppressed for short periods of time, vary in severity, and may be exacerbated by stress. Motor tics often involve the head and neck (eye blinks, shrugs, grimaces, or jaw stretches), but may include other parts of the body, especially the torso and upper limbs. Complex motor tics may be present such as touching, squatting, deep knee bends, retracing steps, and twirling when walking.

Vocal tics include words or sounds such as clicks, grunts, yelps, barks, sniffs, or coughs. **Coprolalia** (involuntary uttering of obscene words) is present in less than 10% of the cases. Other associated features include **echopraxia** (involuntary imitation of motions), **echolalia** (involuntary repetition of words), obsessive thoughts, and compulsions. Hyperactivity, distractibility, and impulsivity are also relatively common. Rare complications of TS include physical injury, such as blindness due to retinal detachment (from banging head or striking oneself), orthopedic problems (from knee bending, neck jerking, or head turning), and skin problems (from picking).

I. Comorbid Disorders

Attention Deficit Hyperactivity Disorder ("ADHD") is present in greater than 60% of cases and is commonly the inattentive type. The symptoms of ADHD sometimes predate the symptoms for TS . Therefore, tics may become noticeable after stimulant treatment for ADHD. **Obsessive Compulsive Behavior** (obsessions and compulsions are present but do not fall under the DSM-IV criteria for Obsessive Compulsive Disorder) is found in 67% of cases. Other comorbid disorders include anxiety disorder, oppositional defiant disorder, depression, learning disability, sleep disturbance, and difficulty with speech.

II. Special Issues Advocates May Wish to Develop in Individual Cases: Functional Impairments Often Manifested by Children with TS

1. Social discomfort, shame, self-consciousness, and depression frequently occur.
2. Social, academic, and occupational functioning may be impaired due to anxiety about tic occurrence in social situations or due to social rejection.
3. If the tics are severe enough, they may directly interfere with activities of daily living (e.g., reading or writing).
4. For many with TS, the comorbid disorders are the most difficult aspect of the condition. For example, the presence of ADHD is a significant predictor of academic problems.

5. The duration of the disorder is usually lifelong, although there may be periods of remission. Often, the severity of the tics decreases during adolescence and adulthood and may disappear entirely. However, the comorbid disorders tend to persist throughout adulthood.
6. Examples of broad areas of functioning (cognition/communication, motor, social, personal development, concentration/persistence/pace) affected by TS include the following:

Motor: For school-age children and adolescents (6-18), functioning in the motor area of development involves the ability to use fine motor skills in order to engage in the physical activities involved in normal mobility, school work, play, physical education, sports, and other physically related daily activities other than self-care. Clients with TS may have motor impairments as a result from the tics.

Social: For school-age children (6 to 12) functioning in the social area of development involves the ability to play alone, to play with another child and in a group, to initiate and develop friendships, to respond to social environments through appropriate and increasingly complex interpersonal behaviors, such as empathizing with others and tolerating differences, and to relate appropriately to individuals and in group situations (e.g., siblings, parents or caregivers, peers, teachers, school classes, neighborhood groups). For adolescents (12-18), functioning in the social area of development involves the ability to initiate and develop friendships, to relate appropriately to individual peers and adults and to peer and adult groups, and to reconcile conflicts between self and peers, family members or other adults outside the family. Clients with TS may have social impairments if the child requires emotional support at school, or has behavioral problems due to comorbid disorders.

Personal development: For school-age children and adolescents, functioning in the area of personal development involves the ability to help oneself and to cooperate with others in taking care of personal needs, health, and safety (for school-age children - eating, dressing, maintaining personal hygiene, following safety precautions, for adolescents - dressing, bathing, doing laundry, adhering to medication or therapy regimens). Children with TS may have personal development impairments due to the tics or due to comorbid conditions, such as sleepwalking or enuresis (urinary incontinence).

Concentration, persistence, or pace: For school-age children and adolescents, functioning in the area of concentration, persistence, or pace involves the ability to engage in an activity, and to sustain the activity for a period of time and at a reasonable pace. Clients with TS may have concentration/persistence/pace impairments due to the tics, comorbid conditions (e.g., ADHD), or due to side effects of the medications (e.g., fatigue).

III. Other Tic Disorders

1. Transient tic disorder does not last longer than twelve consecutive months.
2. Chronic Motor Tic Disorder occurs in 1/200 persons. It includes the presence of motor or vocal tics, but not both. Otherwise, it has the same criteria as TS (also, the person must never have been diagnosed with TS). The severity of the symptoms and functional impairments are usually much less than those found in TS.
3. Tics may be caused by a disorder known as PANDAS (pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection). This autoimmune disease may present with tics, obsessions, compulsions, or encephalitis.

IV. Pharmacological Treatments

1. **<u>Adrenergic agonist</u>**

Guanfacine (*Tenex*), Clonidine (*Catapres*)

<u>Action:</u> decrease activity of locus ceruleus, decrease anxiety (attention-enhancing properties are not as good as psychostimulants)

Side Effects: sedation (guanfacine has a lower incidence of sedation), fatigue, drying of nasal mucosa, orthostatic hypotension (rebound hypertension if withdrawn suddenly), transdermal patch may cause irritation or redness

1. **Neuroleptics**

Haloperidol (*Haldol*), Olanzapine (*Zyprexa*), Fluphenazine, Risperidone (*Risperdal* - lower incidence of extrapyramidal side effects), Pimozide (*Orap* - if symptoms are severe or there is a failed response to Haloperidol)

Action: Dopamine or Serotonin Blockers

Side Effects: tremors, orthostatic hypotension, urinary retention, confusion, sexual dysfunction, fatigue, weight gain, cognitive blunting, restlessness, mood or mental changes (including aggressive behavior, agitation, difficulty in concentration and memory problems), insomnia, extrapyramidal effects - parkinsonism (difficulty in speaking or swallowing, loss of balance control, mask-like face, shuffling walk, stiffness of arms or legs, trembling and shaking of hands and fingers), extrapyramidal effects - dystonic (muscle spasms of face, neck, and back; tic-like or twitching movements, twisting movements of body; inability to move eyes, weakness of arms and legs), tardive dyskinesia (rare)

1. **Benzodiazepines**

Clonazepam (*Klonopin*)

Action: enhance the affinity of GABA receptors for GABA

Side Effects: anterograde amnesia, drowsiness, fatigue, ataxia, dizziness, behavior changes, tachycardia/palpitation, anxiety, confusion, mental depression, psychological or physical dependence (withdrawal seizures)

1. **Monoamine Oxidase B Inhibitor**

Deprenyl (*Selegiline*)

Action: monoamine oxidase B Inhibitors

Side Effects: hallucinations, confusion, nausea, hypotension

Note: Obsessive Compulsive Disorder can be treated with selective serotonin reuptake inhibitors (Fluoxetine (*Prozac*), Sertriline (*Zoloft*), or Paroxetine (*Paxil*)), serotonin and norepinephrine reuptake inhibitors (Venlafaxine (*Effexor*)), or a Tricyclic Antidepressant (Clomiprimine).

References:

1. Rapaport, Judith L., M.D., Ismond, Deborah R., M.A. DSM-IV Training: Diagnosis of Childhood Disorders (Brunner Mazel, 1996).
2. Brown, Lawrence, M.D. "TS ." University of Pennsylvania School of Medicine. Philadelphia, Nov.3, 1998.
3. Diagnostic and Statistical Manual of Mental Disorders (American Psychiatric Press, 4th ed. 1994) ("DSM-IV").

Endnote

1.This description of current medical treatment of TS, as it relates to SSA's Childhood Listings of Impairments, was authored by Lily Kernagis. During the Summer of 1999, Lily was a student at the University of Pennsylvania School of Medicine who worked with the Advocating on Behalf of Children Project of Community Legal Services, Inc., as a participant in the Bridging the Gap Program at Penn.