Tic Disorders in Childhood: Physician Awareness is Key

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Tic disorders are a common occurrence in school-age children and constitute the most common movement disorder brought to the attention of the care provider by families and teachers. The spectrum and severity range from mild, transient simple motor tics to the potentially devastating Tourette Syndrome.

Tics are defined as sudden, rapid, brief, repetitive and purposeless involuntary stereotyped movements or utterances. They frequently are referred to as "habit spasms" or "nervous twitches." An appreciation and understanding of the spectrum as well as the associated disorders allows care providers to counsel parents and to treat the child appropriately when indicated.

Table 1
Tic Classification

	Simple	Complex
Motor	Eye blinking Head twitching Head thrusting Shoulder shrugging Mouth opening	Facial grimacing Touching Smelling Jumping Echokinesis Copropraxia
Vocal	Sniffing Snorting Coughing Throat clearing Grunting Barking	Echolalia Palilalia Coprolalia

Tic Classification

Tics are typically classified as either motor or vocal. Both motor and vocal tics can be either "simple" or "complex" [1] (Table 1). The most common simple motor tics are repetitive eye blinking, head twitching or thrusting, shoulder shrugging, and mouth opening. Complex motor tics consist of motor actions such as facial grimacing, touching, smelling, jumping, echokinesis (imitating other people's movements) and copropraxia (obscene gestures).

Similarly, simple vocal tics consist of sniffing, snorting, coughing, grunting, barking, throat clearing, clicking, squeaking and hissing. Complex vocal tics consist of echolalia (repeating other people's words), palilalia (repeating one's own words), and most distressing, coprolalia. There is actually a third type of tic called sensory tics. These include an uncomfortable sensation such as a tickle, irritation, temperature change or unusual feeling causing the patient to produce a voluntary movement or sound. Although uncommon in children, they have been reported in up to 40 percent of adult Tourette patients [2].

In general, tics have several common characteristics to assist in their diagnosis. Tics typically wax and wane and frequently occur less in an office setting than by history. In this situation a video diary supplied by the family can be helpful to the physician.

Tics frequently increase with anxiety, emotional stress, anger, excitement and fatigue. While they may occur during sleep, they are usually less noticeable and they decrease with relaxation. They also frequently attenuate during absorbing activities. Tics are briefly suppressible, although attempts to do so frequently result in an "inner (emotional) tension," which is relieved by "releasing" the tic. Despite these facts, it is important to recognize that tics are an involuntary movement and are not caused by stress.

Once the diagnosis of tics has been made, an attempt to classify the disorder must be undertaken. This approach divides tic disorders into three groups (Table 2):

- "transient" (lasting less than one year)
- "chronic" (lasting more than 1 year)
- "nonspecific"

Transient tic disorder is the mildest and most common tic disorder occurring in up to 25 percent of school children [3]. Transient tics are simple tics and usually of a motor

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Table 2
Tic Syndrome Classification

I. Transient Tic Disorder
(duration < 1 year)

II. Chronic Tic Disorder
(duration > 1 year)
A) Chronic Single Tics
B) Chronic Multiple Motor or Vocal Tics
C) Tourette Syndrome

III. Nonspecific Tic Disorder

variety, although transient vocal tics can occur. Due to their mild and benign nature, treatment is not necessary or recommended.

Chronic tic disorders include three subtypes: chronic single tic disorder, chronic multiple motor or vocal tic disorder and Tourette syndrome. Chronic single tic disorder requires the occurrence of only a single motor, or less commonly, vocal tic for more than one year. Chronic multiple tic disorder (CMTD) consists of multiple tics, either all motor or less frequently, all vocal, but not both, occurring over one year.

Several studies have shown that CMTD tends to be a mild form of Tourette syndrome and that both are transmitted as inherited traits in the same families [4].

Tourette syndrome (TS) is a chronic, complex, fluctuating tic disorder of variable severity characterized by both motor and vocal tics of both simple and complex types. Current essential diagnostic criteria for the diagnosis of Tourette syndrome include (Table 3):

- onset before age 21 years,
- multiple motor tics,
- one or more vocal tics,
- a waxing and waning course,
- the gradual replacement of old tics with new ones,
- the absence of other medical explanations for tics, and
- duration for more than one year.

Tourette syndrome is more common in males than females (3:1). The mean age of onset is between 6 to 7 years of age with most patients presenting before age 13 years. Eye blinking, facial grimacing or head twitches are the most common initial tics.

Although Tourette syndrome has been thought to be a life-long disorder, in 30 to 40 percent of children with TS all tic symptoms will disappear by late adolescence and an additional 30 percent will experience a marked improvement. The remaining patients will have symptoms persist into adulthood [5]. At present there are no reliable prognostic characteristics that permit identification of those patients who will have spontaneous improvement in their symptoms. Remission appears to be independent of the use of medications.

Nonspecific tic disorder is a category for children who do not meet the criteria for a specific tic disorder. Included in this group are children who develop tics in response to medications such as stimulants or neuroleptic withdrawal and those who have onset of their tics following an acute insult such as herpes encephalitis. The recently described PANDAS (Pediatric Autoimmune Neurologic Disorders Associated with Streptococcal infection) may also be in this category [6].

Comorbid problems

A variety of comorbid behavioral problems are identified in children with Tourette syndrome [7]. The two most prevalent and often problematic are obsessive-compulsive disorder and attention-deficit hyperactivity disorder. Additionally, learning difficulties, speech and language disorders and sleep problems have commonly been observed.

Common compulsive symptoms include ordering and arranging habits, checking rituals, frequent counting, and rituals to decontaminate objects or body parts such as hand washing. Obsessive-compulsive behavior has been reported in more than 40 percent of children with Tourette's [8]. Attention-deficit hyperactivity disorder has been noted in 50 percent of children with Tourette's and may precede the onset of the tics.

Determining impact

Determining the major source of difficulty is essential before any specific recommendations and treatment can be instituted. Tics rarely cause significant physical discomfort or damage, however their adverse effect on psychosocial development and interpersonal relationships can be immense.

It is essential for the care provider to inquire not only about the number, intensity and frequency of the tics, but also to question the degree of impairment and impact they have on the child's activities and social interactions. It must also be determined whether there are any associated problems such as attention-deficit hyperactivity disorder, obsessive-compulsive disorder or behavioral difficulties.

After a complete assessment the care provider should educate the patient and family about the disorder and define the target symptoms that need to be addressed. Just because a symptom is present does not mean it requires pharmacotherapy.

Table 3
Tourette Syndrome Criteria

1. Onset prior to 21 years of age
2. Presence of multiple types of motor tics
3. Presence of one or more type of vocal tic
4. A waxing and waning course
5. Gradual replacement of old tics with new ones
6. Absence of other medical explanation for tics
7. Duration for more than 1 year

Treatment Options

The pharmacologic treatment of tic disorders is strictly symptomatic and not curative. Treatment should be reserved for those children with significant tics causing psychosocial or functionally disabling problems. The goal is to reduce the tics to a tolerable level, realizing that irradication may not be possible. While alternative nonpharmacologic treatments have been used, including conditioning techniques, relaxation, biofeedback and hypnosis, pharmacotherapy is most frequently employed.

At present there are two main classes of medications to suppress tics: alpha-adrenergic agonists and neuroleptics (Table 4). Other agents including benzodiazepines, calcium channel blockers, catecholamine-depleting agents and opiate antagonists have been used with variable success. Clonidine is effective in treating tics in approximately 50 percent to a somewhat overly optimistic 70 percent of patients with Tourette's [9].

While its efficacy is less than the neuroleptics (80 percent), it is frequently the initial medication prescribed due to a relatively benign side-effect profile. It can be administered as a tablet or trans-dermal patch. An alternative alpha-adrenergic agonists, guanfacine (Tenex), may be tried if behavioral problems are prominent. Neuroleptics are the most effective tic-suppressing agents although their side-effect profile may limit their use. The most commonly used agents include: haloperidol, pimozide and fluphenazine. They all appear to be approximately equipotent in suppressing the tics.

In children, generally pimozide and fluphenazine are used before haloperidol because they have a slightly lower incidence of side-effects and are tolerated somewhat better.

Treatment of any comorbid problems needs to be undertaken with careful consideration to the total patient care. The approach to the attention-deficit hyperactive disorder is somewhat controversial because psychostimulant medications may provoke or intensify tics. If behavioral and educational approaches fail, desipramine and alpha-adrenergic agonists may be tried. If necessary a brief trial of central stimulants can be undertaken with careful attention to tic exacerbation.

In Tourette patients with disabling obsessive-compulsive disorder, additional psychiatric intervention and behavioral modification techniques should be considered. Two psychotropic drugs, clomipramine and fluoxetine, have been helpful in treating severe obsessive-compulsive manifestations.

Table 4
Treatment of Tics and Comorbid Problems

Tics

Alpha-Adrenergic agonists
- Clonidine
- Guanfacine (Tenex)

Neuroleptics
- Pimozide (Orap)
- Fluphenazine (Prolixin)
- Haloperidol (Haldol)

Attention-Deficit Hyperactivity Disorder

Behavioral and educational interventions

Pharmacologic treatment
- Desipramine
- Alpha-Adrenergic agonists
  - Clonidine
  - Guanfacine (Tenex)
- Central stimulants
  - Ritalin
  - Dexedrine
  - Pemoline (Cylert)

Obsessive-Compulsive Disorder

Fluoxetine (Prozac)

Clomipramine (Anafranil)

Conclusion

Physician awareness is the key to properly diagnosing and treating pediatric tic disorders. The spectrum is wide, ranging from benign transient simple motor tics to Tourette syndrome. Recognition of this spectrum and familiarity with tic classification and tic syndrome classification allows the care provider to properly educate the family and ultimately determine whether the major source of distress is related directly to tics or associated behaviors.

Once this has been done, then an individualized multimodal treatment plan can be developed. A valuable resource for additional information for care providers, parents and patients is the nonprofit voluntary Tourette Syndrome Association, 42-40 Bell Blvd., Bayside, New York, 11361-2874; (718) 224-2999; and on the Internet at tourette@ix.netcom.com.

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