

•> CASE 42

A 7-year-old boy in second grade is brought to a pediatrician by his parents for an evaluation of his eyes. They state that he blinks them repeatedly, and that this behavior seems to be worsening. They first noticed it a year or more ago, but it became very obvious in the past several weeks. They note that their son cannot control the blinking, and that it appears worse at some times of the day compared to others. The boy's teacher reports that other children tease him because of his rapid eye blinking. The pediatrician observes that in addition to blinking, the child seems to clear his throat frequently, although his nose and throat appear normal on physical examination. The parents report that this behavior occurs several times daily as well. The patient is doing well at school, although he sometimes has trouble completing his homework. The patient's father has a history of attention-deficit/hyperactivity disorder (ADHD).

- **What is the most likely diagnosis?**
- **What is the best therapy for this condition?**

ANSWERS TO CASE 42: Tourette Disorder

Summary: A 7-year-old boy presents to a pediatrician with a year-long history of uncontrollable blinking, which has worsened over the past several weeks. The blinking is worse at some times than at others, and the boy is teased at school because of it. The child also clears his throat repeatedly, although nothing physically wrong can be found. He is doing well at school although he sometimes has trouble completing his homework. The patient has a paternal history of ADHD.

- **Most likely diagnosis:** Tourette disorder.
- **Best treatment:** The first line of somatic treatment involves the use of a dopamine-2 receptor antagonist such as haloperidol or pimozide. If side effects prohibit their use, clonidine, a selective alpha-2-adrenergic receptor agonist can also be of use.

Analysis

Objectives

1. Recognize Tourette disorder in a patient (see Table 42-1 for diagnostic criteria).
2. Describe the basic evaluation and treatment of this disorder.

Considerations

A 7-year-old boy shows signs of a motor tic in the form of eye blinking. This behavior has been present to some degree for more than a year and has recently worsened. The tics affect how other children at school interact with him. He also exhibits a vocal tic in the form of throat clearing, which has been present for a long period of time. This combination of multiple motor and vocal tics occurring for **at least 1 year** is consistent with Tourette disorder.

Table 42-1
DIAGNOSTIC CRITERIA FOR TOURETTE DISORDER

1. Presence of both motor and vocal tics during the course of the illness but not necessarily at the same time.
2. Tics occurring almost every day for a period of at **least a year**, and during this year there is never a tic-free period lasting more than three consecutive months.
3. The disturbance must cause marked distress or impairment in functioning.
4. Onset must be before 18 years of age.
5. The disorder must not be due to a substance or a general medical condition.

There is a genetic predisposition to this disease, probably via autosomal dominant inheritance. There are also relationships among Tourette disorder, obsessive-compulsive disorder (OCD), and attention-deficit disorder. Haloperidol is the most commonly prescribed agent for treating this problem. It is important to note that symptoms such as these in a child could also be related to an environmental allergen, and this should be thoroughly worked up before diagnosing Tourette disorder and beginning treatment.

APPROACH TO TOURETTE DISORDER

Definitions

Athetoid movements: Slow, irregular, writhing movements.

Choreiform movements: Dancing, random, irregular, nonrepetitive movements.

Coprolalia: Vocal tic involving the involuntary vocalization of obscenities.

Dystonic movements: Slower than choreiform movements, these are twisting motions interspersed with prolonged states of muscular tension.

Hemiballistic movements: Intermittent, coarse, large-amplitude, unilateral movements of the limbs.

Myoclonic movements: Brief, shocklike muscle contractions.

Tic: A sudden, rapid, recurrent, nonrhythmic, stereotyped motor movement or vocalization.

Clinical Approach

The lifetime prevalence of Tourette disorder is approximately 4 to 5 per 10,000 in the general population, and it tends to be more common in boys. The motor component (eye blinking, shoulder shrugging, neck jerking) usually emerges by 7 years of age, and the vocal component (grunting, sniffing, snorting, using obscene words) by 11 years of age. Epidemiologic studies involving twins indicate a strong genetic etiology, probably via autosomal dominant inheritance. There is a strong relationship between Tourette disorder, OCD, and ADHD. The dopamine system can be involved in the tic disorders; thus, dopamine antagonists such as haloperidol suppress the tics.

Differential Diagnosis

Tic disorders must be differentiated from general medical conditions that can cause abnormal movements. Involuntary movements such as myoclonus, athetosis, dystonias, and hemiballismus can be seen in such diseases as Sydenham chorea, Huntington chorea, Wilson disease, and stroke. The long-term use of a typical antipsychotic such as haloperidol can cause tardive dyskinesia, another involuntary movement disorder. The presence of

a family history of these disorders, findings on a physical examination, **or** a history of long-term use of antipsychotic medication can help rule out these conditions.

Tics must be differentiated from compulsions seen in OCD. Compulsions are typically fairly complex behaviors performed to ward off the anxiety of an obsession or according to a rigid set of behavioral rules. Certain vocal and motor tics such as barking, coprolalia, or echolalia must be distinguished from the psychotic behavior seen in schizophrenia. However, in the latter case patients have other findings congruent with psychosis, such as hallucinations or delusions. Transient tic disorders last at least 4 weeks but for no longer than 1 year. Patients with a chronic motor or vocal tic disorder can have it for more than 1 year, but there is an absence of multiple motor tics and/or motor and vocal tics occurring simultaneously.

Treatment

The treatment of Tourette disorder involves **both somatic therapies and psychotherapies**. In children, tics are often worsened or triggered by anxiety-producing events. Children and families can be taught to reduce the manifested anxiety at home, which in turn can help to reduce the triggers for tics. A child can be taught relaxation techniques that can help in the reduction of this anxiety.

In addition to anxiety reduction and trigger reduction, there are medications that are helpful in controlling tics associated with Tourette disorder. First-line medications currently used include **clonidine and guanfacine**. Approximately two thirds of children with tics respond to reasonable doses of these agents. In addition to these two classes of medications, some antipsychotic medications can be effective in the treatment of tic disorders. The most commonly used medication in this category is **haloperidol**. Most patients have a favorable response to antipsychotics, but their long-term side effect profiles are causes for concern. Other methods of symptom reduction should be tried prior to using this potentially risky agent.

Comprehension Questions

- [42.1] Which of the following disorders has a higher likelihood of developing in a patient with Tourette disorder?
- A. Obsessive-compulsive disorder
 - B. Sleep terror disorder
 - C. Primary insomnia
 - D. Developmental disability
 - E. Sydenham chorea

- [42.2] Which of the following pathologies would be most important to rule out before starting treatment of Tourette disorder in a child presenting as described earlier?
- A. Streptococcus infection
 - B. Environmental allergies
 - C. Autism
 - D. Marijuana abuse
 - E. Rett disorder
- [42.3] Which of the following medications sometimes used to treat Tourette disorder has the least likelihood of causing tardive dyskinesia?
- A. Pimozide
 - B. Clonidine
 - C. Risperdal (risperidone)
 - D. Haloperidol
 - E. Clozaril (clozapine)
- [42.4] A patient with ADHD is treated with methylphenidate during the school year. After several months of treatment, his teachers and parents note that he has developed both motor and vocal tics. What should be the first line of treatment for these symptoms?
- A. Begin treatment with haloperidol
 - B. Discontinue the use of methylphenidate
 - C. Refer the child for family therapy
 - D. Reduce the dose of methylphenidate
 - E. Administer an anticonvulsant

Answers

- [42.1] A. Obsessive-compulsive disorder has been associated with Tourette disorder, along with ADHD and other learning disorders.
- [42.2] B. Environmental allergies can have a presentation most resembling this presentation of Tourette disorder. Although the other pathologies mentioned are important, they are not usually considered as part of the differential diagnoses for Tourette disorder so probably warrant less consideration in the active work up of this particular patient.
- [42.3] B. Clonidine is an alpha-2 agonist that does not cause tardive dyskinesia as a side effect. It is moderately effective in the treatment of vocal and motor tics, although not as effective as some antipsychotics.
- [42.3] D. The development of tics as a side effect of stimulant medication is relatively common. These tics diminish in severity or cease when the dose is reduced, or the medication is discontinued. Because stimulants

are quite effective in treating ADHD, a reduction in the dose of the stimulant or management of its side effects should be tried before the medication is discontinued.

CLINICAL PEARLS

A diagnosis of Tourette disorder requires both vocal and motor tics and a duration of 1 year.

Vocal tics can consist not only of words but also throat clearing, grunting, and squeaking.

Motor tics can involve complicated movements such as brushing hair or repetitive and intricate hand or arm movements.

REFERENCE

Kaplan H, Sadock B. Synopsis of psychiatry, 9th ed. Baltimore: Lippincott Williams & Wilkins, 2003:1421-1422.