had intermittent difficulties with anxiety and panic, and successfully takes an SSRI for this.

The treatment of Jenny involved decreasing secondary gain for not talking (education of her classmates and a preset schedule for play dates), requesting her sister not to speak for her, and family involvement in making expectations clear (especially from both her mother and her father). Paternal attention seemed particularly helpful in reinforcing expectations and helping Jenny get attention from her father in an adaptive manner. Additionally, an SSRI was initiated for treatment of anxiety. The combination of treatments was effective in allowing Jenny to begin to speak in school and in public.

PART D
Tic Disorders

Tourette Disorder and Other Tic Disorders

Essential Concepts

- Motor tics are repetitive, involuntary movements of discrete muscle groups.
- Phonic (or vocal) tics are involuntary sounds.
- The tics of Tourette disorder are temporarily suppressible and preceded by a premonitory urge.
- The tics associated with Tourette disorder characteristically wax and wane.
- High emotions typically exacerbate tics.

CLINICAL DESCRIPTION

There are four tic disorders described in the Diagnostic and Statistical Manual (DSM-IV-TR): 1) Tourette disorder (also known as Tourette syndrome or TS); 2) chronic motor or vocal tic disorder; 3) transient tic disorder; and 4) tic disorder not otherwise specified. Tourette disorder (which will be referred to as TS) is an inherited neurological disorder with onset in childhood, characterized by the presence of multiple motor tics and at least one phonic (vocal) tic. The tics characteristically wax and wane. TS was once considered a rare and bizarre syndrome, with a psychogenic cause. The eponym for the disorder was bestowed by Jean-Martin Charcot on behalf of his resident, Georges Gilles de la Tourette, a French neurologist who published an account of nine patients with the unusual movement disorder in 1885.

For chronic motor or vocal tic disorder, there are single or multiple motor or vocal tics, but not both. Transient tic disorder is single or multiple motor and/or vocal tics for no

more than 12 consecutive months. For all of the tic disorders, the tics must occur many times a day, nearly every day, and there must never be a tic-free period of more than three consecutive months. The disturbance must cause distress and impairment and have an onset before the age of 18.

Epidemiology

Estimates of the disorder have increased over time, with the ascertainment of milder forms of tics. It is estimated that 1–10 per 1,000 children suffer from a diagnosable tic disorder. A large, community-based study suggested that over 19% of school-aged children have had tics of some type, with almost 4% of children in regular education fulfilling the diagnostic criteria for TS. As many as 1 in 100 people may experience some form of tic disorder, which includes transient tics, chronic tics, or TS. Males are affected three to four times more often than females. This number decreases in adulthood, as the tic symptoms frequently resolve in the milder forms of the disorder.

KEY POINT

Coprolalia (the spontaneous utterance of socially objectionable or curse words or phrases) is the most publicized symptom of Tourette disorder, but it is not required for a diagnosis of TS. Fewer than 15% of TS patients exhibit coprolalia.

TIP

Children with TS describe "premonitory sensory phenomena" or a vague, although sometimes powerful, feeling of needing to tic. Some children liken the feeling to the "need to sneeze" or to "scratch an itch." They are able to suppress the urge for a period of time, but the need to tic seems to build up as irresistible tension. Performing the tic tends to relieve the tension, at least for a time. High levels of either positive or negative emotions tend to exacerbate tics. Concentration in an absorbing activity often leads to a decrease in tics. There are surgeons with TS. They are able to suppress tics for long periods of time for very complex procedures.

Etiology

Genetic studies have proven that the overwhelming majority of cases of TS are inherited. Recent research suggests that a small number of TS cases may be caused by a defect on chromosome 13 of gene SLITRK1. TS is most likely the result of an additive model involving multiple genes in most affected individuals.

The inherited vulnerability to tic disorders may produce varying symptoms in different family members. A person with TS has about a 50% chance of passing the genes to one of his or her children. The genes may express as TS, as a milder tic disorder, or as obsessive-compulsive symptoms with no tics at all. Boys are more likely to demonstrate tics, and females are more likely to demonstrate obsessive-compulsive (OCD) symptoms. Only a minority of the children who inherit the genes will have symptoms severe enough to require medical attention.

While a genetic mechanism is clear, for at least some children with the disorder the exact mechanism has not been established. Research presents considerable evidence that abnormal activity of the brain neurotransmitter, dopamine is involved. Neuroanatomic models implicate abnormalities in the complex cycling cascade of dopaminergic neuronal functions and influences between the brain's cortex and subcortex (especially the striatum and thalamus).

Other factors that have been implicated include perinatal events, such as low birth weight, maternal stress, and obstetric complications. Autoimmune processes may affect tic onset and exacerbation in some cases; the unproven but interesting hypothesis that pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS) play a role in the onset of tic disorders and OCD is a current focus of research.

Assessment

The assessment includes a standard complete psychiatric evaluation. Neurological disorders, such as hyperkinetic movement disorders, including stereotypic behavior, dystonias, choreiform disorders, and myoclonus, may be confused with tics, and should be ruled out (Table 10.1).

Comorbidity and Differential Diagnosis

Individuals with TS are at increased risk of suffering from other psychiatric disorders. The most common comorbidities

TABLE 10.1. Assessment Essentials for Tic Disorders

1. The chief complaint should be obtained from the child and guardian to guide a successful postevaluation discussion and treatment plan formulation.
2. History gathering should include questions that differentiate tics from other movement disorders or stereotypic movements of autism spectrum disorders or stereotyped movement disorder. Ask about "saw tooth" symptom pattern with episodic presentation with abrupt onset and gradual spontaneous reduction associated with group A beta-hemolytic streptococcal infection suggestive of PANDAS.
3. A directed interview will include questions about premonitory urges and ability to temporarily suppress the movements. Additionally, ask questions targeting comorbid conditions, with particular emphasis on symptoms of OCD, ADHD, and learning problems. Testing to rule out learning disorder should be considered if it is suspected.
4. The physical examination should focus on cutaneous abnormalities (suggestive of neurofibromatosis), infectious conditions, and the general neurological exam.
5. Uncomplicated and classical tic disorders do not require other extensive testing. However, for concerns about a movement disorder inconsistent with tics, neuroimaging may be required. Laboratory testing should occur when considering possible metabolic or infectious causes. If PANDAS are implicated, antistreptococcal antibody levels should be drawn.

are OCD and attention deficit hyperactivity disorder (ADHD). Learning disabilities, sleep disorders, disruptive behavior disorders, and mood disorders may also complicate the clinical condition of children and adolescents with TS.

Treatment

TS is a complex and variable disorder in terms of severity and functional disability. Knowledge, understanding, and support are some of the best treatments for tics. The child may be frightened and humiliated by the symptoms. Teachers may not understand the behaviors. Helping educate children and adults alike can go a very long way in terms of destigmatizing the disorder and helping the child avoid the assaults to self-esteem that many children with TS suffer (Table 10.2).

TABLE 10.2. Essentials of Treatment for Tic Disorders

Psychosocial Treatments

Parent and child psychoeducation—education about the disorder for the child and family helps destigmatize and decrease secondary depression, avoidance, and behavioral problems. Additionally, stress reduction and family treatment may be useful to decrease high levels of anxiety or emotion.

Behavioral intervention—habit reversal training (HRT) is a specific and evidence-based behavioral technique used to reduce repetitive behavior by a cooperative and invested child or adolescent. This involves awareness of tics, learning a competitive motor behavior instead of the tic, relaxation techniques, and positive feedback for improvements.

Supportive psychotherapy—this may be helpful for a child who is struggling with issues of self-esteem and other tic- or non-tic–related issues.

Educational/School Related Interventions

Psychoeducational assessment—cognitive and academic assessment is recommended if learning disorders are suspected.

Teacher and class psychoeducation—helping classroom teachers and the other students understand the disorder to minimize teasing or being ostracized may be very helpful in terms of social functioning and self-esteem.

Medication Treatment for Tics

Alpha-2a agonists (first line)
Clonidine, guanfacine

Neuroleptics
Atypical (second line)
Risperidone, ziprasidone
Typical (third line)
Pimozide,[a] haloperidol

Nicotine receptor antagonist (neuroleptic augmentation)
Mecamylamine

[a]Black box warning for potential of prolonged cardiac conduction (QTc).

Medication treatment of tics plus ADHD is complicated because stimulant medication and bupropion may exacerbate tics. If there is significant and impairing ADHD, however, stimulant medications can be used cautiously. Several studies have shown that stimulants do not exacerbate tics any more than placebo does. However, the "start low and go slow"

advice remains. Be advised that the Physicians Desk Reference does include an FDA warning that stimulants should not be used in the presence of tic disorders. Atomoxetine and tricyclic antidepressants may be effective if alpha agonists and stimulant medications do not appropriately treat a severe tic disorder with ADHD.

CASE VIGNETTE

Toby is a 6-year-old boy referred by his pediatrician to you after work-up revealed no medical etiology for Toby's recurrent dry cough. His mother was initially concerned and then annoyed with his repetitive cough, which she described "as though he's forcing it to happen or something." This cough seemed unrelated to runny nose or any illness. It had gone on for months at a time—just when his mother thought it was going away, it would re-emerge. Additionally, Toby was hyperactive and inattentive. His teacher noted that he could not sit still in his seat, he blurted out answers, could not wait his turn, and seemed as though "his motor was running on high speed." She also noted that Toby had an eye-blinking habit that the other children had noticed and some began to mimic.

On examination, Toby was an attractive and well-developed 6-year-old boy, who had a knack for precarious acrobatics in the waiting room and in the office. He presented as bright and articulate, but quite hypermotoric. Neurological examination was essentially normal. There were periods of rapid eye blinking and a dry cough. Toby's father displayed subtle sniffing and eye-blinking movements. Family history was positive for a paternal grandmother who was a "control freak" and did not let anyone sit on the white furniture in the living room. Toby's father and paternal uncle had ADHD.

You diagnose a tic disorder (not yet TS, as it has not been a year) and ADHD. You set up the following treatment plan:

1. Education about TS as a disorder, the cause, and the treatments. You stress that many children never need medication for TS, although there are medications if the symptoms are severe. You discuss the possibility of a mild tic disorder in the father, as well. You discuss the fact that TS and ADHD often occur together.
2. Further evaluation, including school-based evaluation (cognitive and academic) to rule out learning disability. You talk to school personnel about the tic disorder and how to talk about the tic disorder with other students. You discuss with the school social worker the possibility of having a session on medical disorders (such as TS, diabetes, etc.) for the other students so that they may understand Toby better. The school sets a strict policy about teasing and ensures that students who mimic Toby are educated about TS and about the harmful effects of teasing. You send ADHD scales to the teacher. You have the parents fill out an ADHD scale. You consider the need for a Section 504 plan for extra support services for Toby's ADHD and tic disorder.
3. Behavioral structure and support plan at home for ADHD symptoms, including structure, consistency, decreased stimulation, and opportunities for "noncontingent" positive regard and engagement.
4. Pharmacologic intervention with guanfacine or clonidine, which may address hyperkinesis and tics, or atomoxetine for the treatment of ADHD. If severe and functionally impairing ADHD symptoms persist, consider a "start low and go slow" trial of a stimulant medication.