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School-Based Occupational Therapy Services for Children with Tourette Syndrome

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Some people may call it "The Cursing Disease," but whatever you call it, Tourette Syndrome is a baffling disorder that can create havoc with a child's development. Studies indicate that over 70% of children and adolescents diagnosed with Tourette Syndrome require special education services. Children with Tourette Syndrome are clearly a high-risk population for developing special education needs, yet studies indicate that only 25% of them are even identified. Furthermore, of those who are identified or referred to their school district's Committee on Special Education for evaluation, even fewer are referred to occupational therapists for evaluation or treatment. To date, there has been no research addressing interventions for the motoric and organizational deficits that are often observed in this population. In this article, we will describe the spectrum of problems associated with TS, with emphasis on those symptoms and associated features that occupational therapists are uniquely qualified to address.

OVERVIEW OF TOURETTE SYNDROME

Tourette Syndrome (TS) is a neurobiological disorder characterized by involuntary movements and sounds ("tics"). Recent research suggests that most cases of TS are genetically transmitted. While the exact neurological basis is not clear, dysfunction in the dopamine system is most often implicated as the cause of the tics. TS is one of a number of tic disorders. Before a physician can make the differential diagnosis of TS, the following criteria from the DSM-IV must all be met:

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 3 consecutive months.
3. The 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).

There are no neurological or hematological tests that can confirm the diagnosis of TS. Diagnosis is made on the basis of the physician's observation of the patient and/or on the reported history by a reliable observer.

Once thought to be extremely rare, there is now growing evidence that TS may be a fairly common pediatric-onset disorder that goes undiagnosed or misdiagnosed. TS appears to be more prevalent in children than in adults, and may be present in approximately 1.6% of a pediatric population. The symptoms of TS generally emerge between the ages of 5 and 10 years, with a mean age of onset of 7 years. In over 60% of patients, the first tics are usually simple motor tics of the head, neck, or face region. Simple motor tics include eye blinking, eye rolling, squinting, shoulder shrugging, facial grimacing, and tossing hair out of eyes. Approximately 13% of patients report simple phonic tics as their first symptom. Simple phonic tics include throat clearing sounds, grunting, sniffing, spitting, and coughing. Almost every study on TS has found that TS is more prevalent in males than in females. In adults, the male:female ratio is approximately 3:1; in children, it is 9:1.

The first tic of TS usually appears suddenly, lasts a few weeks, and then disappears, only to be replaced a few weeks or months later by a different tic. Apart from the variability in the anatomic location of the tic(s), there is also variability in the frequency and severity of the ticcing. This inherent variability is referred to as the "waxing and waning" of TS. Individuals with TS will generally experience bouts of severe and frequent ticcing alternating with periods of total or relative remission. Because of the variability in the frequency and location of tics, the early tics of TS are usually misdiagnosed, thereby delaying referral to pediatric neurologists. For example, children with eye blinking tics are often referred for vision examinations, while children with sniffing or coughing tics are referred to pediatricians or allergists for treatment of "colds" or suspected allergies. Because simple tics are misdiagnosed, it often takes five years from the onset of symptoms before a child is finally correctly diagnosed.

As the symptoms of TS emerge, simple motor tics of the head and neck region may be replaced by more complex tics of the extremities and trunk. Complex motor tics are movements that involve the coordinated (but involuntary) sequence or activation of two or more muscle groups. Some common complex tics of TS include touching people or objects, smelling fingers or objects, jumping or skipping, poking or jabbing, punching, kicking, hopping, flapping arms, twirling around, tensing muscle groups, thrusting movements of the groin or torso, twirling hair, and imitating the movements of others. Complex phonic tics are linguistically meaningful (but involuntary) utterances. Some commonly observed complex phonic tics include making animal-like sounds, unusual changes in pitch or volume of voice, stuttering, and echo phenomena such as repeating one's own last words or sounds (palilalia) or imitating the last words or sounds of others (echolalia). In a child who is diagnosed as having TS, almost any unusual and repetitive movement or utterance may be a complex tic.

Complex tics are frequently misinterpreted as voluntary misbehavior. Because most children with TS are not diagnosed, it is not uncommon for school districts to misclassify them as "Emotionally Disturbed." If the behaviors are symptoms of a neurological disorder, then such children are appropriately classified as "Other Health Impaired" under the Individuals with Disabilities Education Act (I.D.E.A.). Because most cases are undiagnosed, occupational therapists evaluating children referred by the Committee on Special Education should be particularly alert to the presence of tics. If they are observed or suspected, the occupational therapist should recommend that the C.S.E. refer the child to a knowledgeable pediatric neurologist or psychiatrist for further evaluation.

Although the symptoms of involuntary obscenities (coprolalia) or involuntary obscene gestures (copropraxia) have attracted the most interest, they are generally misunderstood by school personnel. Therapists working with children who exhibit coprolalia or copropraxia need to remember that: (1) the utterances or movements do not reflect the child's true thoughts or feelings, and (2) less than 30% of all patients with TS will develop either of these symptoms. Even if they do appear, they are unlikely to persist for long. As with all tics, they will come and go. By communicating to the child or adolescent that the therapist understands the involuntary nature of the symptoms, the therapist validates the child's experience of being out of control of his body and helps foster a positive therapeutic relationship. One of the greatest sources of distress for children and adolescents with TS is their recognition that their peers and significant adults in their life simply don't understand what it feels like to live in a body that does unpredictable and unwanted things.

Most patients with TS can suppress tics for varying amounts of time, from a few seconds to hours, and this suppressibility often misleads school personnel into believing that the symptoms are voluntary in nature. Suppressing tics is generally undesireable because it will lead to a more explosive release of tics later. Patients who consciously or unconsciously suppress tics throughout the day will often come home and appear to "explode" in tics until they can get them out of their system. Such explosive bouts of suppressed tics are often accompanied by emotional behaviors. The severity or frequency of ticcing may also vary across settings or activities within school. Children are generally less likely to tic in the classroom than they are in one-on-one situations. For that reason, occupational therapists are more likely to observe tics than the classroom teacher.

In the natural progression of the disorder, symptom severity tends to increase over the first decade from onset of symptoms. For about 70% of patients with TS, the severity of the tics or their frequency will diminish significantly in the late teen years. While tic symptoms do not necessarily get more severe during adolescence, there is some evidence that they tend to become more complex and more unpredictable. In fact, if coprolalia or copropraxia emerge, it is generally between the ages of 10 and 13 years.

Although the symptoms of TS follow an inherently waxing and waning course, there are some factors that have been identified as producing a worsening of symptoms. Of all of these factors, stress is the single most potent factor exacerbating the symptoms of TS. Some stressors that have been identified in the clinical literature include: (1) time pressure (particularly time pressure during tests), (2) environments or programs that do not provide adequate opportunity to discharge symptoms or dissipate excessive motor activity, (3) anticipation of upcoming exciting events, (4) school re-opening in September, (5) environmental factors such as allergy seasons or hot weather, (6) specific settings where children are expected to sit quietly (such as library, study hall, concerts, religious services), and (7) fatigue. While tics tend to worsen under conditions of stress or fatigue, relaxation does not necessarily decrease tics. Children observed relaxing and watching TV often display a significant number of tics. One possible explanation is that there is a decreased effort to suppress tics as the child relaxes. Many patients report that symptom severity abates under conditions where they are nonanxiously engrossed or constructively engaged in a novel or challenging task. This observation is of particular relevance for occupational therapists in selecting treatment activities for the child with TS.

For many children, the tics of TS may be the least of their problems. Children with TS are more likely than their non-TS peers to have other neurobiological disorders as well as TS. One of the most frequently observed co-morbid disorders is Obsessive-Compulsive Disorder (OCD) or obsessive-compulsive symptoms. Approximately 40 - 50% of young patients who have TS will also have OCD or obsessive-compulsive symptoms, although the symptoms of OCD tend to emerge after the tics. In young children with TS, the most frequent compulsive-type behaviors relate to “evening up” (for example, tug on one ear then tug on the other), symmetry, touching, arranging things in a particular order, and smelling things. Other common compulsive behaviors exhibited by children with TS are identical to those observed in children who have OCD without any tics: checking (or re-checking a fixed number of times), counting, and rituals involving hygiene. Children with either disorder may also engage in repetitive questioning or reassurance-seeking.

The second most commonly observed co-morbid disorder is Attention Deficit Disorder with Hyperactivity (ADHD). ADHD is present in approximately 30% of cases of mild TS, more than 50% of cases of moderate TS, and 70 - 80% of cases of severe TS. The symptoms of ADHD may precede the emergence of the tics by two to three years. Children who have ADHD appear inattentive, impulsive, distractible, and hyperactive. These children tend to have significant difficulty in a regular classroom and in their peer relationships.

In addition to OCD and ADHD, children with TS are also more likely to experience problems from Oppositional Defiant Disorder, depression, anxiety disorders (including phobias and panic attacks), explosive outbursts of aggressive behavior, and self-injurious behaviors such as hitting and biting. As noted earlier, children with TS also appear to be more likely to have learning disabilities.

Because there is no single profile that accurately describes children who have TS, occupational therapists asked to evaluate a diagnosed child should request to see the treating neurologist’s or psychiatrist’s report. The report will provide essential information about any suspected motor impairment, and will be useful in understanding the child’s symptoms and problems, as well as providing information on medications and possible side effects.

Because tics are of neurologic origin, they are generally not amenable to behavior modification techniques, and the first line of treatment will usually be pharmacological. The most commonly prescribed medications for tic management are neuroleptics such as haloperidol (Haldol®), pimozide (Orap®), and fluphenazine (Prolixin®), although some investigators are now exploring the use of respiridone (Respiridol®). While there are a variety of possible side effects associated with neuroleptic use, the most commonly observed side effects involve extrapyramidal symptoms (e.g., dyskinesias and tremor). Neuroleptics may also produce lethargy, cognitive dulling, and subtle personality changes, including increased irritability. Physicians may prescribe the anti-hypertensive clonidine (Catapres®) for children who have TS in combination with Attention Deficit Disorder. The most commonly observed side effect of clonidine is lethargy. The management of Attention Deficit Disorder (with or without hyperactivity) in the presence of TS is often complicated. Some children with TS will experience a worsening of tics if placed on stimulant medications such as methylphenidate (Ritalin®). For children who have obsessive-compulsive symptoms or full-blown obsessive-compulsive disorder, there are a number of anti-depressants that may be prescribed. Until recently, Anafranil® was the most frequently employed. Anafranil® can produce tremor and sweating, among other possible side effects. In very high doses, there is a slight risk of epileptic seizure. Recently approved anti-depressants have increased the range of choices, and occupational therapists may need to obtain information from the child's treating physician on side effects profiles.

SCHOOL-BASED PROBLEMS AND OCCUPATIONAL THERAPY

Neuropsychological studies of children with TS indicate that although TS is not correlated with intellectual potential, children with TS are more likely to have deficits in the areas of fine motor skills, handwriting, and visuomotor integration. Academically, they may display deficits in written mathematics, passage comprehension (in timed or group-administered tests), spelling, and written expressive language. Many of these children also have significantly impaired organizational skills and are described as "terminally disorganized" in terms of time management, materials management, and space management. They may lose track of papers, assignments, and possessions, and have significant difficulty in completing projects.

As noted earlier, most cases of TS are undiagnosed, and occupational therapists evaluating children need to be alert to the presence of any tics so that children can be referred for further neurological evaluation. This is especially true when evaluating children with known ADHD or behavioral problems, but also applies to children with significant learning disabilities. If the child has already been diagnosed, the occupational therapist may find the Bender-Gestalt VMI, Beery-Buktenica, Bruininks-Oseretsky, Purdue Pegboard, and handwriting speed tests to be of particular clinical value. Because there have been no formal studies of children with TS using a battery of the evaluation techniques, unequivocal statements are not possible. The deficits described below, however, are based on the authors' experiences with children with TS, and are meant to serve as guidelines for occupational therapists who are asked to evaluate a diagnosed child.

1. Occupational therapists evaluating or working with younger children may find decreased fine motor speed and decreased dexterity. Children with these problems will have difficulty manipulating objects, using scissors, tying shoelaces, etc. Many parents of such children report significant difficulties in activities of daily living such as using eating utensils and dressing.

2. In children and adolescents, dysgraphia, or poor handwriting, is the most commonly observed problem. Letters may be poorly formed, there may be inconsistency in size of the letters, and there may be a lack of fluidity to the writing. The lack of fluidity may be secondary to increased upper extremity tension, and some children do appear to use excessive pressure on the writing instrument. A number of these children also appear to employ an improper pencil or pen grip.

3. Another feature related to the impaired handwriting is the poor use of the available workspace. Many children do not stay within the margins and do not employ normal spacing. They may leave almost no space between words and/or run the line off the edge of the page. They may start out at

the left margin but then shift the left margin successively to the right over the course of the page. Other children appear to totally disregard the ruled horizontal lines as they write.

When the child has ADHD as well as TS, school personnel frequently misattribute graphomotor or visuospatial problems to impulsivity and/or lack of motivation. To the contrary, these problems usually represent a true deficit, and occupational therapists should be alert to their presence. Because children with TS often do have low frustration tolerance, however, therapists should make every effort to pace the session appropriately and to introduce enjoyable tasks.

4. Slow handwriting speed is also observed in a significant percentage of children we have observed. Slow handwriting speed may reflect interference from several possible sources. First, arm tics may directly interfere with handwriting or result in a slower overall production rate. Second, a child who has OCD or obsessive-compulsive symptoms may have compulsive rituals that interfere with writing. A child may need to compulsively rework her writing until it looks “just right,” or she may have to perseverate on a particular letter. Other nonobservable compulsive rituals may also interfere. Imagine how your handwriting speed would be affected if you had to mentally count how many times the letter “E” occurred as you were writing a sentence or paragraph. Third, there may be independent visual processing deficits (e.g., decreased ocular motility) that reduce graphomotor speed. Finally, as noted earlier, many of the neuroleptic medications used to treat tics may produce motoric side effects that interfere with handwriting. Occupational therapists working with a diagnosed child are advised to find out what medications the child is taking because if the problems are due to medication side effects, the therapist’s approach to remediating deficits may be different. Occupational therapists should also ask the school nurse to keep them apprised of any and all changes in a child’s medication regimen. Many of these children undergo frequent changes in medications and/or doses.

Because there may be a variety of factors contributing to graphomotor impairment or decreased speed, occupational therapists will generally need to obtain additional information to help them determine the cause of the problem. Some problems, such as improper pencil or pen grip or increased muscle tone can be addressed, as can the visuospatial organizational deficits. Other problems, such as the compulsive reworking of letters or counting rituals, are unlikely to be successfully remediated through occupational therapy alone, and the O.T. should consult with the child’s treating psychologist or physician. Children whose obsessive-compulsive writing rituals are so severe as to preclude a functional handwriting may require alternative means of expressions (such as tape recording, dictating to a scribe, or word processing).

5. Impaired visuomotor integration is also present in a significant percentage of children and adolescents with TS. Delays of one to two years on the Bender Gestalt VMI or Beery VMI are fairly common. In one psychological study, approximately 40% of the sample showed impairment on the Coding Subscale of the Wechsler Intelligence Scale for Children. Some children will also display relative deficits on other Performance subscales of the WISC. Unfortunately, many school psychologists are not aware that these patterns of deficit should lead the school psychologist to recommend evaluation by an O.T.

Dysgraphia and poor visuomotor integration probably both contribute to the common observation that children with TS tend to have problems with written arithmetic (but not necessarily mental math or applied problems). Many of these children cannot fit their work neatly into the available workspace on the page, or cannot line up their columns of numbers correctly to arrive at the correct solution. Occupational therapists working with children with TS also assist the child by serving as a consultant to the classroom teachers. Occupational therapists can advise teachers to have the child turn lined paper sideways so that the lines serve as column guides. Occupational therapists should also encourage the teacher to allow the child a larger workspace for performing their computations. Enlarging the ditto or workpage by using the enlargement feature on the office copier will provide extra space for the student who needs it. Occupational therapists should also advise the classroom teacher if the child needs to have a reduced amount of copying from the board or text to paper. Many of these children will also require testing accommodations that permit them to record their answers directly in test booklets.

For the child with TS and ADD, occupational therapists may want to consider providing services in the classroom. Children who have ADD typically have a problem with carryover of skills into the classroom. One possible explanation is that the demands of the classroom are so overwhelming to the child that he cannot even begin to think about incorporating the developing skills or techniques he is acquiring in a pull-out therapy situation. If school-based occupational therapists cannot provide some of the services in the classroom, then they should try to allocate time to serve as an ongoing consultant to the classroom teachers. The O.T.'s advice with respect to the following questions can make a significant difference to the child:

(1) How much handwritten work should the child be expected to do at one time?
(2) Should the teacher grade for neatness or ignore it?
(3) Should the teacher reduce copying activities from the board for the child with TS?
(4) Does the child need assistive technology to complete his assignments? Many (if not most) children with TS require word processors in the classroom and at home to enable them to produce written work that is commensurate with their knowledge and potential. The occupational therapist is uniquely qualified to determine if the child should be taught keyboarding, and if so, what system of keyboarding should be taught (touch typing, hunt and peck) and what postural adaptations may be necessary. The use of computers also permits the occupational therapist to teach the child time management skills through the use of commercially available software. Children with TS who are academically on grade level but who need a computer as assistive technology are eligible to receive it under Section 504 of the Rehabilitation Act and the Americans with Disabilities Act.

SUMMARY

Children with TS often display fine motor impairment, decreased psychomotor speed, handwriting deficits, visuomotor integration problems, and spatial organizational deficits. While the child with TS may appear to be emotionally disturbed, there is a neurobiological basis for the child's symptoms and emotional dysregulation. The frustration produced by sensorimotor or sensoriperceptual problems may contribute to some of the emotional behaviors observed in school settings. Occupational therapists who recognize tics in undiagnosed children and refer them for further neurological evaluation provide an invaluable service. For children who are already diagnosed, occupational therapists may be an important part of a comprehensive individualized education plan. It is the authors' hope that this article will produce greater awareness of this medical disorder and stimulate research on the efficacy of school-based occupational therapy interventions for children with this disorder. For further information on Tourette Syndrome, or to contact your local chapter, write or call:

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END NOTES

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