Tourette’s Syndrome: Medical, Educational and Behavior Therapy

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Abstract

The French physician, Gill de la Tourette, was the first person to define Tourette’s Syndrome (TS) in 1885 and has brought much attention to it in Western countries for the past few decades. Unfortunately, most Asian countries are not familiar with Tourette’s Syndrome. Tourette’s Syndrome is regarded as a disorder of the nerve system that often appears with involuntary tic behaviors. The purpose of this article is to give a better understanding of Tourette’s Syndrome, starting with a brief background, followed by its pathogenesis and an analysis of treatment, including medication and education. The related studies of Tourette’s syndrome are also discussed in this study.

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Introduction

Tourette's Syndrome is regarded as a disorder of the nerve system that often appears with involuntary tic behaviors, such as repetitive eye-blinking, shoulder shrugging, throat-clearing and some other complex motor and vocal tics [1] [2] [3] [4]. According to the “Diagnostic and Statistical Manual of Mental Disorders, 4th edition (DSM-IV)”, if the tics occur many times a day, nearly every day for at least four weeks, or intermittently throughout a period of more than a year without a tic-free interval of more than three months, the diagnosis of Tourette syndrome should be considered [5].

Tics are the motor movements which are rapid, sudden, unexpected, irresistible, inappropriate, repetitive, stereotypic activity, and occur irregularly. There are four major types of tics, including simple motor tics, such as eye blinking, nose twitching, head jerks, shoulder shrugs; complex motor tics with more purposeful movements, such as sustained looks, facial gestures, and obscene gestures. The remaining two tics are phonic tics, including simple phonic tics, such as throat clearing, sniffling, barking, gurgling, and innumerable other sounds; complex phonics with more meaningful utterances, such as the echo phenomenon, immediate repetition of other’s words, or inappropriate and aggressive statements [6].

Unlike other diseases, most people are not familiar with Tourette's Syndrome and may take its uncontrollable tics for intentional misbehaviors. Tourette’s Syndrome has a prevalence rate of approximately 3 in every 1000 people and the ratio of men to women is estimated to be 3:1 [7]. A recent study revealed the increased prevalence of Tourette's Syndrome with a prevalence rate about 0.85% to 1%. The study further indicated that Tourette Syndrome affects 1% to 3.8% of western young children and affects 0.46 to 0.56% Taiwanese and Chinese young children [8] [9]. According to The Tourette International Consortium report on the characteristics of 6805 patients with TS, the male to female ratio is 4.4–1 and the mean age of onset is 6.4 years [10]. According to Tourette's Action, which is the Tourette foundation in the United Kingdom, the average age for diagnosis in the UK is seven. Previous studies indicated that most of the children with TS typically start in childhood at the age of six-years-old and may occur in earlier years [11] [12].

Most children with mild TS and without any comorbid conditions will usually not be diagnosed and there is no need for treatment. The percentage of TS children who experience a reduction in the symptoms by the end of their teenage years is 50 percent. Accordingly, about half of children with TS will carry the condition into adulthood [13]. Robertson pointed out that the symptoms of Tourette’s Syndrome might be controlled or improved, but usually won’t completely disappear with age [14]. However, another study reported that approximately half of patients, tics disappeared in their adulthood, 40 - 45% of cases are improved, and only 5–10% of patients remain the symptom of tics [15] [16]. Although Tourette's syndrome has a general trend and certain external symptoms, many specialists pointed out that Tourette's symptoms are highly variable from person to person. These tics may fluctuate in type, frequency, severity, response to stress and other features. Some Tourette's children appear to have fewer incidents of tics when they encounter high stress situations, while some others might have more complex tics symptoms [17].

The causes of Tourette’s Syndrome

There is more and more evidence to support the concept that Tourette's Syndrome has a genetic basis and it may originate in early brain development [18]. It is regarded as a hereditary disease, but there is no
Evidence to certify which gene is precisely connected with it. Tourette's syndrome is a genetically neuropsychiatric disorder and 51.7% of TS patients have family history [19]. Although the mechanism of TS remains unclear, imbalanced neurotransmitters (dopamine and serotonin) seem to play a role. Basal Ganglia, a typical part of the brain which controls body movements, has also been linked to TS [20], [21]. It is reported that certain ethnic groups, such as sub-Saharan black African, African-American, and American Hispanic ethnic origins are less commonly associated with Tourette's syndrome [22].

Some studies have also indicated that different environmental factors may be relevant to Tourette's Syndrome, such as complications, drinking or smoking during pregnancy, children born either prematurely or with low birth weight, and other possible environmental factors which may be relevant to TS [23], [24], [25]. A recent study indicated that inadequate maternal weight gain and parity, cannabis use during pregnancy, and maternal alcohol were found significantly associated with Tourette's Syndrome. However, this study also showed that birth weight and prenatal maternal smoking, were not significantly associated with Tourette syndrome tic disorder [26].

Furthermore, some researchers indicated that the tics of TS may be an acquired consequence of autoimmune mechanisms following a Group A-hemolytic streptococcal (GABHS) infection, because the antibodies wrongly attack the area that is responsible for movement, called the Basal Ganglia [27].

The Comorbidities of Tourette's Syndrome

Many TS children have comorbid conditions of either Attention Deficit Hyperactivity Disorder (ADHD) or Obsessive-compulsive Disorder (OCD). In fact, tics do not cause many learning problems for TS children, but these two major comorbid conditions of TS have caused many learning and adjustment problems. Some other psychiatric comorbidities are also found among TS children, such as learning difficulties, migraine, depression and anxiety disorders [28], [29], [30], [31]. Unfortunately, these comorbidities of Tourette's Syndrome usually happen to the TS children at the age between 4 and 10 years old [32].

The previous studies indicated that the comorbidities of Tourette Syndrome have a high risk of epilepsy [33], [34]. A study was conducted in Taiwan to determine the risk of epilepsy in children with Tourette syndrome. The researchers analyzed 1,000,000 randomly selected subjects from the Taiwan National Health Insurance Research Database. There were 1062 TS patients under aged 18 chosen as the experimental group, while the control group consisted of subjects of typically development totaling three times the number of experimental group subjects matched by age and gender. The results of this study indicated that a much higher incidence of epilepsy in TS children’s group was prevalent. This study further indicated that the increased risk was not associated with comorbidities, but with Tourette’s Syndrome itself. In the other words, Tourette syndrome is associated with a higher risk of epilepsy, meaning TS children are much more likely to develop epilepsy than the children without Tourette's Syndrome [35], [36].

Learning abilities and Social Adjustment of Children with TS

Many studies revealed that half of children with TS have problems with learning disabilities. Studies which evaluated children’s academic achievement also suggested that many TS children have learning disabilities [37], [38], [39]. However, other research indicated that children with Tourette’s Syndrome are intelligent and their intelligence is not different from other children of typically development [40], [41]. It is worth mentioning that the academic achievement and
learning potential of TS children are very often limited because many of them have to endure negative peer relationships and biased treatment by classmates and teachers.

A Taiwanese study indicated that most of the parents of TS children (77.4%) claimed their children had outstanding or above average academic performance, but 49.4% of the TS parents felt their children suffered rejection at school. Such results indicated that most Taiwanese parents of the children with Tourette's Syndrome worried about their children's adjustments in school [42]. The findings of this study were consistent with previous findings that TS children had more emotional and behavioral problems than children of normal development. TS children experienced more personal distress, peer rejection, and learning difficulties [43] [44].

TS children who are also diagnosed with ADHD are always at risk for developing a complex array of emotional and behavioral problems [45]. Not surprisingly, TS children always have difficulty with making friends and sometimes have poor relationships with teachers. The unusual motor or vocal tics of TS children can cause other pupils to avoid or tease them. It is unavoidable that children with TS often suffer from peer rejection and experience more frustration [46]. The feeling of being “weird, different, rejected, unacceptable”, not only causes painful learning experiences for TS children, but also damages these children's self-esteem.

A greater understanding and sensitivity toward children with TS will assist teachers and parents as they strive to meet the educational and behavioral needs of TS children. In fact, when a child has TS, not only does he/she have to deal with it, but also the people around him/her, such as the teachers, peers, parents and siblings. Like other chronic disorders, Tourette's Syndrome is quite often considered as a major stressor that affects the quality of life for the whole family [47]. The tics and possible comorbid conditions, undoubtedly, cause many emotional adjustment problems for TS children who are, most likely, the major focus for teachers and parents. The parents of TS children need to spend more time and energy taking care of them. This usually makes the other siblings in the family feel less important. Sometimes, the other siblings worry that he/she might be the next TS child [48].

**Medical treatment of Tourette's Syndrome**

In addition to education, the treatment of TS involves proper medication. As mentioned previously, the key symptom of TS is tics. Although medical treatments can control tics, the patients with very mild symptoms do not need any medication, unless they are thought to be seriously disturbed by these symptoms, or if they feel that the frequency or intensity of the tics is rising. If medication is indicated, it is important to diagnose appropriately, and it may take a long time for doctors to find the proper dosage which can help control tics of the TS patients effectively but without causing serious adverse effects. Therefore, when cooperating with doctors, it requires great patience. Pimozide and Haloperidol, the classic neuroleptic antipsychotic drugs, which can effectively block the receptors of D2 dopamine and alter the effect of dopamine to a normal level, are the only two FDA-approved tic-suppressing medications.

However, Many TS patients are not willing to take these drugs because they worry about the side effects which including sedation, depression, and weight gain. Because of these side effects, other medications may be considered as first-line drugs for TS patients. Previous studies revealed that the alpha-2 adrenergic drugs, which inhibit the sympathetic nervous system's postganglionic functioning, such as clonidine and guanfacine, have high efficacy in reducing tics [49] [50]. These alpha-2 adrenergic drugs are also effective for
ADHD, so many doctors would try to prescribe alpha-2 adrenergic drugs for TS children who have comorbid conditions of ADHD. However, these drugs also have some side effects such as headache, drowsiness, and moodiness [51].

A study with 38 TS patients was performed to assess the efficacy and safety of risperidone for the treatment of Tourette’s Syndrome. At the end of the 4-week trial, 22 patients (58%) experienced improvement, while 7 patients (18%) had no significant improvement in their symptoms of tics. The results suggested that risperidone could be one of the promising medications for the treatment of tics [52].

Along with oral medication treatments, local intramuscular injections of botulinum toxin can be another option, when dealing with some complex and bothersome tics. It can reduce tics and mitigate uncomfortable sensations [53] [54]. Unfortunately, the effectiveness of botulinum toxin therapy is less than six months. Deep brain stimulation (DBS) is another therapeutic option for TS patients who cannot optimally control their symptoms with medications. However, deep brain stimulation is intended only for adults, not children, and is usually chosen only when other options are exhausted, due to the potential for complications such as stroke and infection [55].

It is certainly the case that many TS patients need to deal with their tics as well as the comorbidities. Usually, the doctors will evaluate the possible interfering conditions first, and then consult with the patients or the TS child’s parents, to decide whether to use a combination therapy of tic-suppressing, anti-OCD and anti-ADHD medications, or to merely take care of the more immediately pressing symptom [59].

**Educational assistance and behavior therapy**

Although some studies revealed that pharmacological treatments can usually reduce approximately 50% of the frequency of tics, very often side-effects will occur [60]. Therefore, for those who only have mild symptoms, behavior therapy can be considered. Or even for those who have complex and severe symptoms, behavior therapy can also be used alongside other medical treatments.

One of the effective behavior therapies for TS combines several methods which is called comprehensive behavioral intervention for tics (CBIT) [61]. There are several steps in CBIT. The first step is to use the perspective of psychoeducation which means that we need to have a better understanding of Tourette’s Syndrome and Tic disorder. The second step is to apply functional intervention which is to identify environmental events that cause tics symptoms to get worse. Then, under this circumstance the therapist will work with the patient to reduce or control these tic increasing situations.

In addition to CBIT, habit reversal therapy is also very helpful for TS patients and it is followed by several procedures. Initially, we need to identify all of the tics in detail and choose the tic which is most bothersome. Then, the therapist will help the patient become aware of situations that trigger the tics before they appear. The next procedure is to find a diversionary response. For example, if somebody has a motor tic which involves eye-blinking, he can be taught to channel it into more
favorable movements, such as closing his eyes for a few seconds before the blinking starts [62].

Social support is also very important especially for TS children. Therefore parents and teachers should have a good understanding of Tourette’s Syndrome. Last, but not least, is relaxation training which is used to reduce the stress that a person with tic disorders experiences. For instance, deep breathing combined with progressively tensing and relaxing the muscle groups in the TS patient’s body. Once patients know how to use these methods, they can control their tics successfully by themselves.

In educational settings, behavioral therapies, such as habit reversal training, functional intervention, relaxation training or some other positive reinforcement programs, can successfully reduce the frequency of tics [63]. According to the Tourette action reports as well as some other research, even though behavior therapies cannot cure Tourette’s Syndrome, they can be very effective in helping TS children to control their tics. Therefore, CBIT is recommended as first line therapy by Tourette Canada as well as other major Tourette associations.

However, it is important that this behavioral therapy is conducted by specially trained therapists. In order to reduce tics in TS children, it is extremely important to help them cope with their emotional and social adjustments. Parents’ attitudes toward TS are the key point in helping TS children’s emotional adjustment. Interaction between parents and TS children appear to be the best predictor of the children’s emotional adjustment [64]. Also, the understanding and flexibility of school teachers, and parent-teacher communication can facilitate the social acceptance of TS children. Teachers and parents play key roles in helping TS children to maintain and reinforce their self-confidence and self-esteem. To date, there is no direct comparison study between behavioral therapy and medical treatment. It is vitally important to cooperate and communicate closely with therapists and consultants during behavioral therapy and/or medical treatment.

**Conclusion**

Although there are still many challenges and tasks which researchers and educators need to conquer in the future, it is hoped that, through the understanding of Tourette’s Syndrome, our society can provide these TS children with an appropriate learning environment to develop their strengths and abilities. There is a need for teachers and parents to understand the superior intelligence of TS children and then use this as a guide for behavioral therapy. For instance, if the TS children are good at sports, then they should participate more in sporting activities; if TS children are good at visual arts, then drawing should be the core course of behavioral therapy. By doing so, the tics of TS children can be controlled effectively and they can build self-esteem and confidence. However, we need more empirical researches to support what is suggested here.

Previous researches of Tourette’s Syndrome have provided a valuable data base, but the related institutions should sponsor some educational conferences about Tourette’s Syndrome and share their findings from these previous studies. It is important to educate parents and teachers regarding the necessary practical skills which are important in helping children with TS through practical-oriented workshops. In addition, raising public awareness of Tourette’s Syndrome through the media is also necessary because TS patients need to be understood.

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