



TOURETTE'S WITH DIGNITY DON'T TIC ME OFF

Paul Sandor MD, FRCPC **Mary Robertson** MBChB, MD, DSc (Med), DPM, FRCPCH, FRCP, FRCPsych
Stuart Ellis-Myers **Sharon Chung** PhD
Colin Shapiro BSc (Med), BSc (Hons), MBBCh, PhD, MRCPsych, FRCP(C)

TOURETTE'S WITH DIGNITY DON'T TIC ME OFF

Paul Sandor MD, FRCPC

Mary Robertson MBChB, MD, DSc (Med), DPM, FRCPCH, FRCP, FRCPsych

Stuart Ellis-Myers Sharon Chung PhD

Colin Shapiro BSc (Med), BSc (Hons), MBBCh, PhD, MRCPsych, FRCP(C)

Tourette Syndrome is much more common than was previously appreciated. It causes significant distress to both patients and their family members. It results in lasting psychological effects even when the key features in many patients substantially subside.

There are now effective medical and behavioural treatments and the management should ideally include a team of parents, teachers, administrators, nurses, doctors, psychologists, and other patients with the disorder.

We hope this booklet is a good start to providing you with the key information you need. In line with this view, one of the authors, Stuart Ellis-Myers, writes from a personal perspective. This is not necessarily indicative of how things are for all patients with Tourette's.

Index

| | | |
|-------------------|---|----|
| <i>Chapter 1</i> | <i>I Have Tourette's - A Patient's View</i> | 4 |
| <i>Chapter 2</i> | <i>What is Tourette Syndrome?</i> | 8 |
| <i>Chapter 3</i> | <i>What Causes Tourette Syndrome?</i> | 10 |
| <i>Chapter 4</i> | <i>How Common is it?</i> | 13 |
| <i>Chapter 5</i> | <i>How do You Recognize Tourette Syndrome?</i> | 17 |
| <i>Chapter 6</i> | <i>Methods Used to Assess Patients with Tourette Syndrome</i> | 22 |
| <i>Chapter 7</i> | <i>What are the Medical Treatments?</i> | 29 |
| <i>Chapter 8</i> | <i>Sleep in Tourette Syndrome</i> | 37 |
| <i>Chapter 9</i> | <i>What are the Social Problems? - A Patient's View</i> | 42 |
| <i>Chapter 10</i> | <i>School-Related Problems</i> | 45 |
| <i>Chapter 11</i> | <i>What are the Social Solutions? - A Patient's View</i> | 49 |
| <i>Chapter 12</i> | <i>How can Schools Help?</i> | 52 |
| <i>Chapter 13</i> | <i>How much Difference do the Solutions and Treatments Make? - A Patient's View</i> | 57 |
| <i>Chapter 14</i> | <i>Resources for Patients, Parents, Schools and Health Care Professionals</i> | 60 |

1. I Have Tourette's – A Patient's View

WHAT DOES IT FEEL LIKE

If you are truly going to understand what it is like to live with Tourette's, I (S. E-M.) believe that first you have to be able to turn off your rational mind and think back to your own personal experiences of feeling stressed, about to be hurt, and broken by forces way outside of your control.



I truly believe that if it were possible for you to know what Tourette's feels like, within minutes you would be screaming out for a tranquilizer or any way to turn off that experience. For those of you wanting to truly know what Tourette's feels like, imagine doing this:

- 1. Quickly guzzle down five Red Bulls and five shots of espresso***
- 2. Now grab two pieces of metal wire and stick them into an electrical socket until your hair starts to smoke!***

These two actions would collectively pulverize your sanity as your mind and nervous system erupt into an excruciatingly high level of anxiety that is completely blinding and overwhelming. Your body would twitch uncontrollably and many of you would start to swear loudly as this helps to express and release the overwhelming anxiousness that boils within. It is impossible to focus on anything except the trauma that is being experienced.

Most people fail to realize that the hidden inner Tourette's journey is the toughest part to live with. It is all too easy to get caught up and focus in on the drama of diagnosing our bizarre movements and sounds. Our twitches and tics are nothing more than expressions of how we feel inside and our bodies reacting to neural disordering.

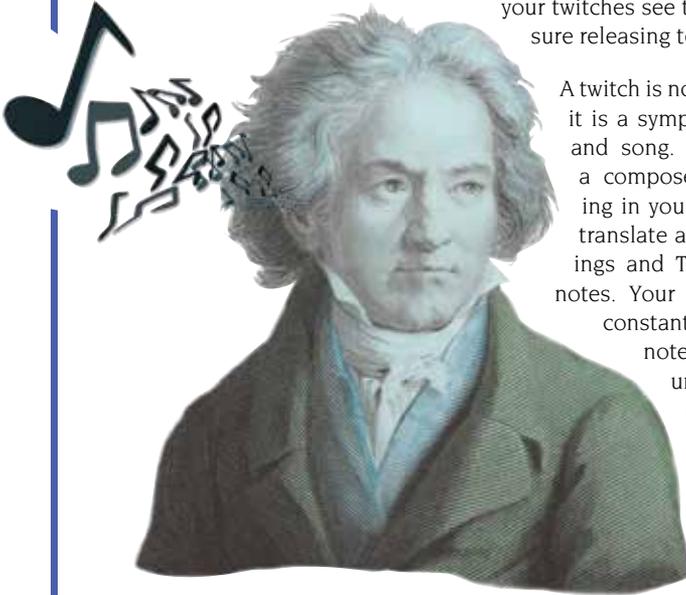
My experience with Tourette's has always been about dealing with what is going on behind the twitches, vocal sounds and bizarre movements. For people like me living with Tourette's, the true feeling resides in the never ending sense of depression, obsessive compulsive and anxious thoughts that constantly flow in and out of my mind. I have learned over and over again that if you try to fight against this never-ending flow of dark thoughts, this action just gives them more power to consume and burns you up.

All of us living with Tourette's fear the moments when these constant negative feelings overtake and bring us down. As I type these words there are two constant forces at work. I can literally feel them at work. It's like being constantly set out on a small, shallow open boat floating precariously in a dark stormy sea. It is perpetual night as nasty oozy black waves of anxious, depressed and obsessive thoughts constantly washed in wanting to capsize and pull the boat down. I have learned through many years of experience, medication and self-help to first accept that these situational feelings are never ending and in doing so I am now able to calmly bail out these massively uncomfortable feelings.

The Tourette's experience is uniquely different for every person living with it. Initially it looks like we all have the same type of twitches. If you look closer you will see that every time we twitch we do so differently and uniquely. Our tics, twitches and bizarre movements are just our way of naturally venting and expressing the never ending conveyor belt of anxiousness that travels through our every waking moment. For me there is a constant sense of the world coming to an end. Worse, that something really nasty is about to occur. My rational mind knows that such feelings are completely wrong. However my irrational mind is constant in reminding me that my rational mind is not to be trusted!

The toughest challenge we all have to face is to learn how to embrace and accept that we will be twitching and feeling both obsessive and anxious for the rest of our life. The crazy part is once you accept this reality, it truly does get better because in the instant you stop fighting Tourette's, it gets better!

It took me many years to realize that my twitches and vocal sounds are not embarrassing but are actually valuable and important! This is because I have learned how to value them for the way they both express and release all of the anxiety I am feeling. Instead of thinking badly about your twitches see them as priceless pressure releasing tools!



A twitch is not a single note. Rather, it is a symphony of sound, music and song. Imagine that there is a composer like Beethoven living in your brain. It is his job to translate all of your anxious feelings and Tourette's into musical notes. Your composer is there to constantly write down musical notes that explain your Tourette feelings. He then hands these musical notes to a wildly gesturing conductor who stands before an orchestra made up of your muscles and vocal chords. Just to make things interesting.

We all have a scramble of "notes" in our brain which we are trying to put into some order. For patients with Tourette's this can be a particular challenge

Your (OCD) obsessive, compulsive disorder needs to

constantly change and disrupt everything by demanding that your Beethoven needs to change some of your freshly composed musical notes already written and conducted and sometimes even after they have been played! When the orchestra, made up of your muscles and vocal chords, performs these musical notes correctly by twitching and sounding we instantly feel a very brief sense of momentary calm. Then the entire process starts all over again and never stops!

I also have twitches and panic attacks that have nothing to do with this musical process. They just show up, do their thing and leave!

Most people living with Tourette's have a favourite set of twitches. Many tics, sounds and movements come and go and are forgotten like winter, spring, summer and fall. When I was a kid I would have to constantly blow on my hand, blink, snap my fingers and then quickly stretch my arm in and out while making chirping

sounds. I would also have to look behind as I raised my foot three to five times to look under my shoe. I took a lot of bullying and rejection at school for these twitches. Every time I would constantly look under my shoe, the kids at school would call me 'Dog Poo Stu'. Interestingly, if I go for an early morning walk or sometimes when I am in an elevator I am compelled to look under my shoe again!

The Chicken Slap is my favourite twitch that I have used and performed for the past four decades. It is highly efficient at helping me get the twitch movements and sounds performed quickly and efficiently.

Here's what I do and perform within my chicken slap twitch: I first need to scrunch my neck, close my eyes, then bob my head to the left a few times as I make different toning sounds. Then my eyes join in and close up tight as my right arm muscles tighten drawing in and then my tics and movements accelerate repeating all of these same movements over and over again. Then I have to add in my tongue needing to make a squeaking sound depressing itself down on one of my back right large molar teeth. I also have to rhythmically and rapidly snap my right hand little finger against my hand. Next the arm slapping evolves from a side to side action into becoming more of a forward and back movement. I stop once the storm of needing to twitch is past by performing these movements as composed and conducted.

What is wonderful is that I have never actually thought about taking the time as I am now to describe this twitch in such detail. Even after living with Tourette's for over forty years it feels as if I am still getting to know my own twitches better. The truth is we all twitch so many times a day we do not focus or remember much of what is going on in a twitch as it is being performed.

2. What is Tourette Syndrome?

Tourette Syndrome (TS) was named after Georges Gilles de la Tourette, a French neurologist who published a paper describing a series of nine patients with this condition in 1885. These were rather severe cases, not unlike the people with Tourette Syndrome depicted in recent times on television and on film. Such severe cases are relatively uncommon as a majority of patients with Tourette Syndrome have only mild to moderate symptoms.

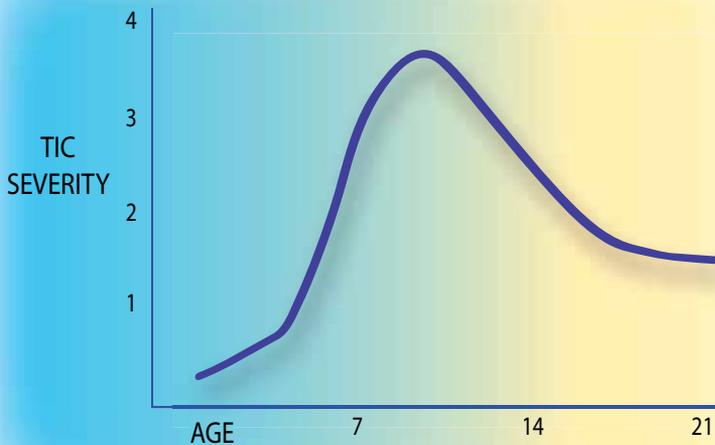
Tourette Syndrome is an inherited neurodevelopmental disorder with childhood onset. Let us consider this statement in more detail. Tourette Syndrome is inherited i.e. it tends to occur more often in certain families and it is passed from generation to generation. There is much evidence that genetic predisposition plays a major role in determining who develops Tourette Syndrome. There are also several lines of evidence e.g. neuropathology and imaging, which suggest that this genetic predisposition has subtle but definitive effects on how the brain development proceeds in affected individuals. The signs and symptoms of Tourette Syndrome begin in childhood and always before age 18.

As a result of atypical brain development, a person with Tourette Syndrome has difficulty inhibiting certain movements and sounds. The most common and earliest signs of Tourette Syndrome include frequent blinking, squinting, eye rolling and head and neck movements. These are jerky, sudden and occur at irregular intervals. Phonic tics may include sniffing i.e. a sharp intake of breath through the nose or snorting i.e. pushing the air out through the nose, throat clearing, coughing and squeaking. These tics can be termed simple motor or simple phonic tics. Many, but not all people with Tourette Syndrome, also experience complex tics that resemble a voluntary movement such as tapping, touching people or objects and brushing one's hair repeatedly. Examples of complex phonic tics include uttering words, sentences and most infamously swear words. It is important to note that no more than 10% of people with Tourette Syndrome will experience involuntary swearing (coprolalia) and, like all other tics, this one may be present for a period and then disappear. It is an important feature of Tourette Syndrome that tics change over time with some disappearing and new ones appearing periodically. It is also not uncommon that old tics that have disappeared may come back another time.

The majority of patients are able to describe an unpleasant sensation in an area of the body where the tic occurs, a so-called premonitory urge, which is relieved temporarily by performing the tic. However, many patients experience a widely distributed vague, yet distressing premonitory urge that is not localized to any particular area. A premonitory urge occurs more often before complex tics and less often before simple tics. It is also important to note that children who are younger than 9 or 10 are often unaware of their tics and may not experience, or report, any premonitory urges.

Emotions have a prominent impact on frequency and severity of tics in Tourette Syndrome patients. Not only negative emotions (anger, fear) but also positive ones (excitement, joy) can increase the frequency and the intensity of tics while the emotional arousal lasts. The tics may take a few hours or even several days to settle down to a baseline after the emotional arousal ends.

Tics usually emerge around age 5 but may be present as early as age 1 or 2. It may begin in the teens, but by accepted expert agreement must begin before age 18. The frequency and the intensity of tics tend to increase in the pre-pubertal and pubertal period, roughly between age 10 and 14 (see below). Once the child enters the late teens, there is a high probability of improvement i.e. the frequency and severity of tics tends to decrease significantly in about 80% of the patients.



Most patients with Tourette's show tics by the age of 7 years and many are much better by 14 years

3. What Causes Tourette Syndrome?

As mentioned previously, there is much evidence from family and segregation studies as well as twin studies that genetic factors play a very significant role in determining who will manifest the symptoms of Tourette Syndrome. Family adoption studies indicate that this is not a learned behaviour because even adopted away children who were raised in families free of tics developed this disorder. In addition we learned from twin studies that Tourette Syndrome is strongly heritable. This follows from the observation that when one identical twin has the diagnosis of Tourette Syndrome, then in about 50% of cases his/her identical twin will also meet diagnostic criteria for this disorder. In contrast, it has been observed that when one non-identical (fraternal) twin receives diagnosis of Tourette Syndrome, the other twin has only about an 8% chance of having the same condition. The fact that even genetically identical twins both meet the diagnostic criteria for Tourette Syndrome only about half the time, indicates that there are non-genetic (environmental) factors at play. What these factors are has not been established, although there is some evidence suggesting that adverse events during pregnancy and delivery may be relevant. In addition physiological stress during gestation, labour and the immediate postpartum period is more common in people with Tourette Syndrome than in controls. The observation that the smaller twin is usually the one who is more severely affected, also indicates that adverse conditions during gestation interact with genetic predisposing factors and may lead to a more severe manifestation of the disorder.

Family studies have also indicated that first degree relatives of a person diagnosed with Tourette Syndrome have an increased risk of having tics or Tourette Syndrome. Incidentally, they also have an increased risk of having OCD (Obsessive Compulsive Disorder) and ADHD (Attention Deficit Hyperactivity Disorder).

Early segregation studies indicated that Tourette Syndrome may be caused by a single major (autosomal dominant) gene, but more recent studies have shown that Tourette Syndrome is likely a result of an interaction between several genes with a minor affect. It is therefore common that a child with Tourette Syndrome receives some of the genetic predisposition from each parent. This may also at least partially explain why there is such a wide range of tic severity among Tourette Syndrome patients, since inheriting a large number of affected genes is likely associated with more severe conditions.

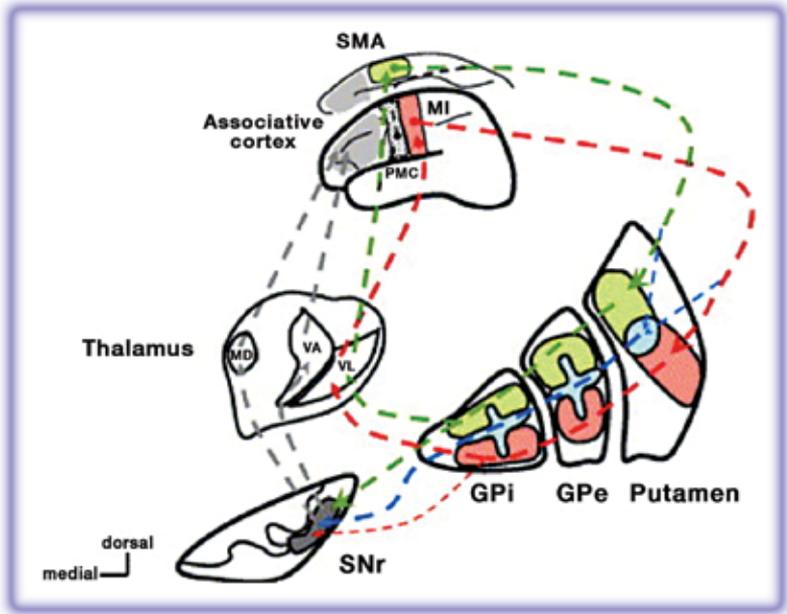


If one identical twin is diagnosed with Tourette's, the likelihood of the other twin having it is 53%. For a fraternal twin it is 8%. In the general population the risk is 1%

It appears that these genetic factors lead to subtle deviations from the normal path of brain development that eventually manifests with the various involuntary movements and sounds (and possibly other co-morbid conditions e.g. ADHD, OCD and Learning Disabilities). Fortunately in many cases the neural development process repairs or compensates for the initial deviation from normal brain development, often leading to a marked improvement in symptoms by the end of teens.

Brain imaging studies have shown that people with Tourette Syndrome have a subtly different brain structure. These abnormalities have been found in several parts of the brain pathways which together are called the cortico-striatal-thalamic-cortical circuit (CSTC) and these include reduced volumes of caudate nucleus, changes in the volume of dorsolateral prefrontal cortex and thinning of the sensory motor cortex. (See fig. on next page). How these changes affect brain function is not entirely clear. Several studies using various forms of functional imaging such as functional MRI and PET scanning certainly have detected differences between patients with Tourette Syndrome and control subjects. There is research suggesting that the minority of people who continue to have significant tics in adult life are the ones whose brain has failed to develop adaptive changes i.e. failed to compensate for the earlier neurodevelopmental abnormalities.

It has been well-known from pharmacological studies that medications that block dopamine receptors (brain receptors) tend to improve tics substantially and those that increase dopamine levels in the brain may increase the tic frequency and severity. The important role of increased dopamine activity in Tourette Syndrome is also supported by evidence from imaging studies using PET scanning.



Cortico-striato-thalamo-cortical circuit

The cortico-striato-nigro-thalamo-cortical loop is thought to be important in planning, selecting and producing various normal actions. Subtle abnormalities in the brain structures and/or connections between them are believed to lead to tics.

(SMA, supplementary motor area; pre-SMA, pre-supplementary motor area; MI, primary motor cortex; PMC, premotor cortex; GPi, globus pallidus pars internus; GPe, globus pallidus pars externus; SNr, substantia nigra pars reticulata; VA, ventroanterior thalamus; VL, ventrolateral thalamus; MD, mediodorsal thalamus).

4. How Common is it?

Tourette Syndrome (TS) is very common. This is only a relatively recent “discovery”, and even within the medical fraternity there appears to be some dissent or disagreement at times. TS was once thought to be a rare and bizarre curiosity really only known to very few. Now the majority of doctors, health professionals and educators believe that TS occurs in about 1% of the population. Why has there been this change?

First the understanding of what TS actually is, has helped. In the past many people (even doctors) believed that coprolalia (the swearing tic) had to be present for the diagnosis of Tourette’s. This is not so, and only about 10% of TS patients have coprolalia; even in specialist TS clinics only around a third of patients have this symptom. So, years ago, if there was no coprolalia, the patient might not have been diagnosed. In addition there has been an increased awareness of TS through medical education, the media and also the National support groups. Examples of support groups include Tourette Syndrome Foundation of Canada, Tourette Syndrome Association (USA), Tourette’s Action (UK) and a Pan European Professional Group called the European Society for the Study of Tourette Syndrome (ESSTS).

When discussing how common TS is, researchers use two terms (i) prevalence and (ii) epidemiology. Prevalence refers to how common the disorder is, while epidemiology indicates where in the world the disorder (TS) occurs. As disorders may occur with varying frequencies in various populations in the world (and as the world today has mass migration, this applies to sub-populations within a country), it is important to know where the investigations have been undertaken.

Before epidemiological and prevalence studies were carried out, patients with TS were reported in medical papers in most parts of the world (Australia, Canada, Europe (e.g. Austria, Denmark, France, Germany, Italy, The Netherlands, Poland, Spain, Sweden, UK), India, New Zealand, Papua New Guinea, South America (e.g. Brazil, Argentina), China, Japan, the USA and the former USSR. The only exceptions were the Caribbean Islands and Sub-Saharan Black Africa – where TS had never been described. This is not because it is not recognized (in Africa an atypical case was described and, in Trinidad, a case of “Tardive Tourette’s” was described in a patient with schizophrenia, following long-term use with neuroleptic medication). In other words, in countries where predominantly black individuals live, TS probably does not exist. In an unpublished PhD thesis, in South Africa (white cases) TS was found, and in a prevalence study in the Xhosa people (black) a few were found. The reasons for this have been described elsewhere including that these cases were genetically mixed, as in the case with numerous peoples of all racial groups in South Africa. For example the early white Afrikaans settlers (from

the Netherlands) and their slaves, the indigenous Khoisan people, the Black African people and subsequent French Huguenot, English & other European settlers all inter-married – particularly in earlier times (1655 – 1800s). Of interest is that ADHD has been described in Sub-Saharan Africa and in an “OCD initiative Group” in Nigeria.



Parts of the world where Tourette's has not been reported

Parts of the world where Tourette's has been reported

Parts of the world where Tourette's has been reported and where prevalence studies have been conducted

Subsequent studies specifically examined the prevalence of TS, and the epidemiology was also deduced. The studies which explored how common TS was, were conducted in many of the western (developed) world's countries (e.g. Italy, Poland, Sweden, the UK, the USA, as well as in China, Taiwan, and Japan). The studies which examined youngsters in the community (i.e. at school or in a community sample) between ages 5 and 18 and in which the methods included a direct interview with the young person, suggested that TS affects between 0.4% and 3.8% for youngsters; a calculated figure suggested that a ball-park figure is about 1% of most populations worldwide. In addition, the studies were similar in that they were conducted in mainstream schools/communities. Most also used similar multi-staged methods, with both direct observations of the youngsters (in almost

all studies) and questionnaires about pupils, as well as obtaining information from parents and/or teachers, and in some instances, both. None of the studies involved individuals who had already been identified. They were also conducted by clinicians with a special interest in TS. In the majority of the “cases” identified, TS was mostly undiagnosed and mild, without distress, impairment or coprolalia.

The prevalence (i.e. how common it is) of TS in special educational populations, such as those individuals with learning difficulties (LD), emotional and behavioural disorders or autistic spectrum disorders (ASD), is even higher. In some cases they are as high as 25% in LD and between 10% and 15% in ASD. These studies took place in Italy, the UK, and the USA.

Boys and men are more commonly affected, with the male to female ratio being 3:1. Clinical characteristics are similar irrespective of the country of origin, highlighting the biological nature of TS. In some instances it seems that within families, the affected males have tic symptoms, whereas the females have OCB.

There are some cultural differences. In the Far East, the figures have been slightly less than in the west, but whether or not this is a true reflection of lower prevalence, or because different diagnostic criteria were used, is unclear. In Sub-Saharan Africa there have been no case reports and only one study has been conducted which showed a very low prevalence. It has been suggested that TS may not occur at all in the area, and one of the explanations is genetic (see chapter 3).

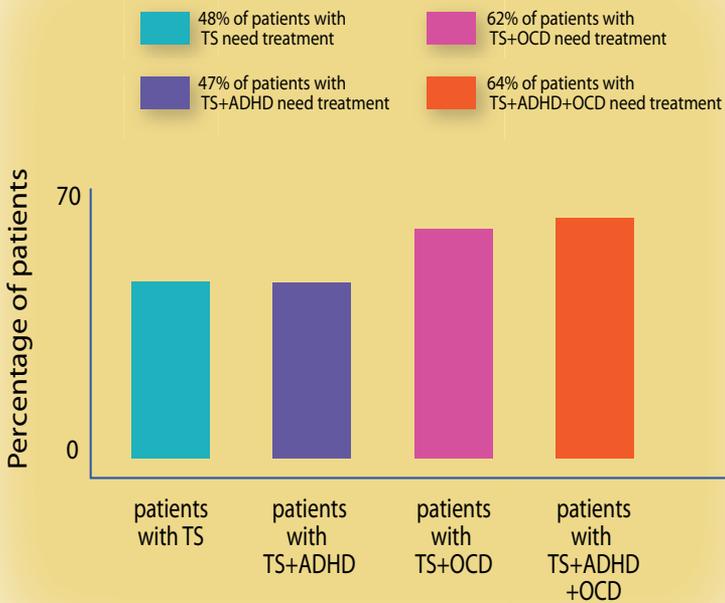
Let us look at what a prevalence of about 1% means in various countries. Canada has a population of 30,000,000 (30 million), which means that about 300,000 individuals may have TS, even if they have not been diagnosed, many are mild and do not require treatment. The USA has a population of approximately 300,000,000 (300 million) and so again approximately 3,000,000 (3 million) may have TS. In cultures which are ethnically similar and from European descent (having migrated to North America) these figures will probably be true. However, in some populations (e.g. Sub-Sahara Black Africans, African-Americans, Amerindians and possibly Hispanics in the USA), TS is less frequent.

There are many reasons for the varying figures in prevalence including the age of the individuals in the studies giving the ranges of 0.4%-3.8%. These include whether or not the individual (young person/child) was actually interviewed, the length of the interview, whether multi-staged methods were used, the age group evaluated, and whether or not there were other informants (e.g. teachers, parents, videos etc.). In some of the earlier studies which showed low prevalence, studies included patients who were hospitalized or already known to doctors and also older individuals.

It was initially believed that TS was life-long and the severity remained the same, but several studies all reported that tic severity reduced during adolescence; only

increased tic severity in childhood was associated with increased tic severity as adults. Many adults with TS think that their symptoms have disappeared, but it has been shown using video studies that 90% of the adults still had mild tics. It was also shown that tics were much more severe among children (about 80% requiring medication), whereas only around 12% of adults needed medication for tics and their distress was less. Also, it has recently been shown that phenotype (see Chapter 5) changes with age. This illustrates how the age of the individuals used in a prevalence investigation is crucial (ideally 5-18 years) and certainly in schools and community, not in clinics.

PERCENTAGE OF PATIENTS THAT NEED TREATMENT



5. How do You Recognize Tourette Syndrome?

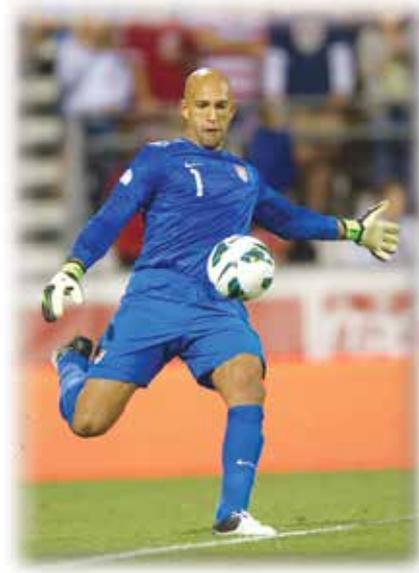
As noted in previous chapters, Tourette Syndrome (TS) is a childhood onset movement disorder: the cardinal features are multiple motor tics (“twitches”) and one or more vocal/phonic tics (noises) lasting longer than a year. The age at onset ranges from 2-21 years, with most youngsters having their first tics (motor tics) at the age of 7; the onset of the vocalizations is usually later, commonly around 11 years. Tics can be simple (e.g. blinking, eye rolling, eye-brow raising, nose twitching, head nodding, mouth pouting, facial grimacing) or complex (e.g. touching, squatting, jumping, hopping).

Premonitory “urges” are common and may be either localized (around the area of the tic) or generalized (covering a wide area of the body). This may be compared to a sneeze, in that it feels uncomfortable to the individual, and the feeling is relieved after the tic.

Tics usually begin in the head and face: eye blinking is often the first and one of the most common tics, seen in the majority of individuals. Common simple vocalizations include sniffing, throat clearing, gulping, snorting and coughing. Complex vocal tics include barking, making animal noises and uttering words. Other characteristic features include echolalia (copying what other people say), echopraxia (copying what other people do) and palilalia (repeating the last word or part of sentence said by the individual).

Coprolalia (inappropriate, involuntary, swearing, which is often disguised by the patient) is uncommon, occurring in only approximately 10% of patients, mainly starting around 15 years. Many physicians are still under the misapprehension that coprolalia must be present in order to make the diagnosis. The media highlights the swearing aspect of TS, as it has “public sensational appeal”. It is also important to understand that the coprolalia of TS is different from “social swearing”. “Social swearing” is usually directed, for example “F... off” if the person is angry with the other; used as an adjective, for example “Bloody marvelous” to highlight how good something is; or in exasperation or frustration, for example “damn it!”. The individual saying these words is not embarrassed and usually feels justified in saying the word. This is in contrast to coprolalia. The person with TS is often embarrassed by it and so, instead of the whole swear word, many say only parts of the word (Fu Fi Shi Cu), and disguise it (by coughing, saying something or covering their mouths). Coprolalia is also out of context (“Hello it is wonderful to meet you, you c...; sorry I didn’t mean that”). A good example of coprolalia is “Perfect Pete” (winner of “Big Brother” 2006 in the UK). He would be talking normally when suddenly he would say “wanker” and stick the third finger of his right hand in the air – usually continuing his otherwise normal conversation afterwards.

Other complex words can appear quite random. In the UK, a well-known young woman with TS says the word “biscuit” hundreds of times a day; other than that her speech and conversation are normal. Also in the UK, a young man says “chicken” in the middle of an otherwise normal sentence.



Famous and successful people do have Tourette's

It is also important to note that many famous/successful people have TS. An excellent example is Tim Howard, the football goal-keeper for the USA (Sydney Olympic Games), and Manchester United and subsequently Everton goalkeeper. He is a fabulous man, who is a great ambassador for TS and if one did not know he had TS one would probably not notice his tics at all.

genetically related and occur much more commonly together than by chance. The common co-existent psychopathologies with TS include depression, depressive symptomatology, learning difficulties and personality disorder. These occur commonly but there are several reasons for the co-occurrence. The relationships between the co-morbid disorders and the psychopathology and TS are complex. The relationships between TS and the co-morbidities/psychopathologies are summarized in the figure on page 20. In many individuals it is these disorders which cause more disadvantage than the tics per se.

In a young person with TS and ADHD, a child (more often boys than girls)

CO-MORBIDITIES AND ASSOCIATED CONDITIONS

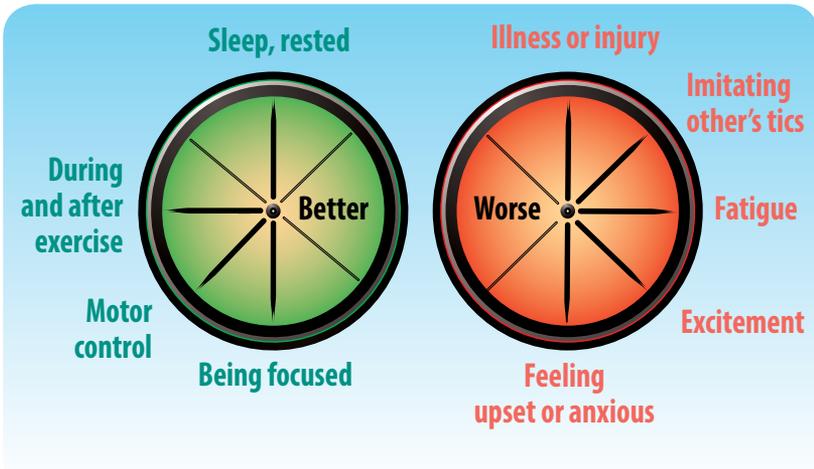
The predominant co-morbid disorders associated with TS include Attention deficit-hyperactivity disorder (ADHD), Obsessive Compulsive Behaviour (OCB), Obsessive Compulsive Disorder (OCD) and probably autism spectrum disorders (ASD). They are



One of the authors, Dr. Mary Robertson with Tim Howard

may be hyperactive (getting out of his chair at school and maybe running around), have poor attention (often “day-dreaming”), be impulsive (blurting into others’ conversations/queue) and therefore disturbing his peers. A young person with ODB/OCD may take ages to do his/her homework (as it “has to be perfect”, and thus he/she will re-write sentences), spend ages making things symmetrical, counting numbers over (and the “favourite number” has to be reached and “feel good”). They can also have doubts and check things repeatedly. They may hoard things (collect and keep unnecessary objects) or be overly concerned with cleanliness, dirt, germs and contamination. Many of these problems can be kept personal and secret – so some feel they “suffer in silence” as their difficulties may not be noticed unless they are questioned directly. Others have ruminations and rituals and if these are not performed “correctly” the aftermath can be awful for them. For instance, there is a lovely young boy with TS and OCD who thought he was responsible for 9/11, as his usual ritual was not correctly performed that day.

SOME FACTORS THAT MAKE TICS BETTER OR WORSE

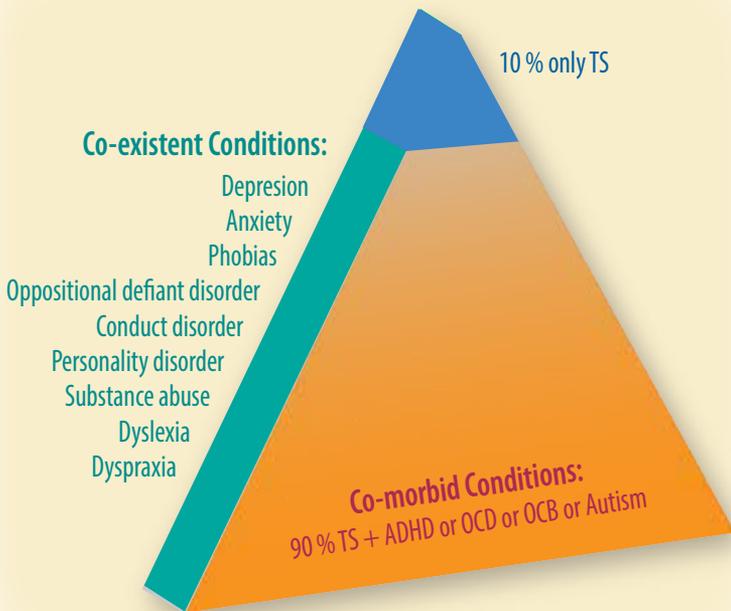


What is important is that in community and clinical studies about 90% of people with TS have co-morbidity/psychopathology. As an example, one large world-wide study of 3,500 TS clinic patients showed that about 90% at all ages, had co-morbidity/psychopathology. The most common was ADHD, followed by OCB,

and OCD. Anger control problems, sleep difficulties, coprolalia, and SIB (self-injurious behaviours) only reached high levels in patients with other difficulties (co-morbidity). Males were more likely than females to have co-morbid disorders. In one excellent community study, of the 90% of TS individuals who had other diagnoses, 36% of the individuals had three or more diagnoses.

DETECTION

How do we recognize people with TS? Moderately to severely affected (TS patients with obvious motor tics, loud vocalizations, constant barking, coprolalia and copropraxia), are easily recognized and diagnosed. But if a person is affected with mild simple tics only, the doctor, the public, and even the individual may NOT know that the person has TS. For example, if a young boy has persistent



Only a minority have pure Tourette's. The majority has a co-morbid condition with some genetic linkage. All patients (pure TS or TS + co-morbidity) can have a co-existent condition such as depression

eye blinking, often his parents will take him to an optician, optometrist, or an allergist only to be told that “nothing is wrong with his eyes”. The professional is correct – nothing is wrong with the eyes. Often, however, if the blinking continues the parents then seek the advice of an ophthalmologist. Again they are told (possibly reassured) that nothing is wrong with the eyes. Many people are satisfied with that explanation. When they seek advice from a TS specialist, only then the diagnosis of TS may be made.

The international diagnostic criteria used to assume and stipulate that TS is a unitary (single) condition. Recent studies have challenged this. The studies using fancy statistical tests have shown that TS has more than one type (phenotype/factor/class). All studies have shown that one type consists of tics only (i.e. TS-only) which we have seen occurs in about 10% of all TS patients; the other 90% have other difficulties/disorders.

Relatively recently, researchers have analyzed TS individuals on the basis of clinical symptoms into subgroups, specifically separating those with and without ADHD, demonstrating significant differences. In almost all studies patients with TS-only (tics only) had virtually no other behavioural problems, while those with TS and ADHD had an increased chance of having various behavioural problems (increased aggression, delinquency or conduct difficulties) than healthy controls.

People with TS have a higher risk of obsessional symptomatology than healthy control subjects and OCB (Obsessive Compulsive Behaviour) encountered in TS has been shown to be statistically and clinically somewhat different than behaviours found in “primary” OCD (Obsessive Compulsive Disorder).

In conclusion, an expert can recognize TS easily, but understanding of the complexities often present is important – otherwise TS cases may be missed. Living with undiagnosed TS and/or other neurodevelopmental conditions can have serious life-long consequences on the individual’s psychological development, educational success and social acceptance and as a result, the individual’s education and socialization may be jeopardized.

6. Methods Used to Assess Patients with Tourette Syndrome

Medical assessment has several components: history, physical examination and investigations. The first two are important in making a diagnosis of TS. Investigations are rarely necessary, since there is no laboratory blood test, imaging or EEG technique that would help in making the TS diagnosis. Occasionally in uncertain cases these techniques can be used to make sure that the physician is not mistaking another condition for TS. It must be said that such confusion is quite rare.



Asking the patient is one component of diagnosis

So if the patient can provide history that two or more motor tics and at least one phonic tic have been present on and off for longer than a year, and that these have been variable in anatomical location and frequency over time, that is crucial information. If the physician can observe motor and/or phonic tics in the office (or often in the waiting room), that reinforces the diagnostic conclusion.

In many situations, standardized questionnaires can be used to systematically elicit important information. When assessing a patient with TS it is helpful to use a questionnaire that lists various tics, obsessive compulsive symptoms and ADHD behaviours to help patient recall various symptoms that may have been present in the past.



Doing a simple test can give you valuable information about a diagnosis

In Tourette's it is common to rely on a variety of sources of information especially when dealing with children – parents, guardians and teachers can provide invaluable information since many children and some adults may only be aware of some (or even none) of their symptoms. It is useful to obtain information about the range of different tics, the complexity of the tics, if they result in movements or sounds, how frequently they occur and how bothersome they are to the person experiencing them. Many individuals have tics but seem to be oblivious to them. The usual way to ascertain this information is to use a standardized questionnaire. There are many such questionnaires which vary in scope, detail and length and will provide different amounts of detail and will be qualitatively different (this would be similar to the different scales for measuring depression or fatigue).

The first one is the Yale Global Tic Severity Scale (YGTSS) which is available on the internet at: <http://dcf.psychiatry.ufl.edu/files/2011/06/TIC-YGTSS-Clinician.pdf>

This questionnaire is based on both subjective accounts and objective observation but the questionnaire should be viewed as an objection instrument analogous to the barometer (blood pressure device) for measuring the level of blood pressure. The questionnaire provides a validated method of recording the severity of tics that can be helpful in monitoring the treatment outcomes. (Refer to figure on page 20).

Sample questions from the Yale Global Tic Severity Scale (YGTSS) follow on the next page.

YGTSS

YALE GLOBAL TIC SEVERITY SCALE

Sample questions from the Questionnaire

MOTOR TIC SYMPTOM CHECKLIST

The patient has experienced, or others have noticed, involuntary and apparently purposeless bouts of:

Eye movements

Nose, mouth, tongue movements, or facial grimacing

Head jerks/movements

Shoulder jerks/movements

Arm or hand movements

Leg, foot or toe movements

Abdominal/trunk/pelvis movements

Other simple motor tics

Other complex motor tics: touching; tapping; picking; evening-up; rude/obscene gestures; obscene finger/hand gestures; unusual postures

PHONIC TIC SYMPTOM CHECKLIST

The patient has experienced, or others have noticed, bouts of involuntary and apparently purposeless utterance of:

Coughing; throat clearing; sniffing; whistling; animal or bird noises

Other simple phonic tics such as syllables; words

Rude or obscene words or phrases

Repeating what someone else said, either sounds, single words or sentences.
Perhaps repeating what's said on TV (echolalia)

Repeating something the patient said over and over again (palilalia)

Other tic-like speech problems, such as sudden changes in volume or pitch

SEVERITY RATINGS

Number

Frequency

Intensity

Complexity

Interference

Impairment

In some medical conditions there are co-occurring conditions or facets of the condition that are very important to establish and measure. An example may be in patients suspected of a breathing disorder in their sleep, where one uses a questionnaire to establish the likelihood of sleep apnea, one may also need to quantify the level of sleepiness and/or the level of alertness (they are not reciprocal) that the person has. One may use a questionnaire to measure sleepiness and another to measure alertness.

As described in the previous chapter, there are two conditions that very frequently co-occur in Tourette's. These are attention deficit hyperactivity disorder (ADHD) and Obsessive Compulsive Disorder (OCD). In the next three pages, you will see extracts from two important questionnaires/checklists that help diagnose these disorders.

The first of these conditions is OCD. On pages 26 and 27 is the widely used and comprehensive assessment of OCD called the Yale Brown Obsessive Compulsive Scale (Y-BOCS). If you wish to see the full questionnaire, which is 22 pages long, the link on the internet is: <http://www.stlocd.org/handouts/YBOC-Symptom-Checklist.pdf>

With regard to ADHD, a sample tool that is widely used is the SNAP-IV which is often done by the teacher and the parent. This will give an indication of the likelihood of ADHD being a significant issue. A sample of this 18-item scale is provided on page 28. The full version is available on the internet at: <http://www.myadhd.com/snap-iv-6160-18sampl.html>

In conclusion, a detailed assessment to establish the fine details of Tourette's requires considerable time and a well-trained and experienced clinician. The greatest problem, however, is that associated conditions and problem behaviours are often missed or neglected by parents, teachers, nurses and family doctors as well as many specialists who do not treat Tourette's routinely even though these often cause much more difficulty to the patient and his family than do the tics.

Y-BOCS SYMPTOM CHECKLIST

Sample questions from the Questionnaire

| AGGRESSIVE OBSESSIONS | EXAMPLES |
|--|---|
| 1. Fear might harm self | Fear of eating with a knife or fork, fear of handling sharp objects, fear of walking near glass windows |
| 4. Fear of blurring out obscenities or insults | Fear of shouting obscenities in public situations like in church, fear of writing obscenities |
| CONTAMINATION OBSESSIONS | EXAMPLES |
| 12. Concern with dirt or germs | Fear of picking up germs from sitting in certain chairs, shaking hands, or touching door handles |
| 16. Bothered by sticky substances or residues | Fear of eating with a knife or fork, fear of handling sharp objects, fear of walking near glass windows |
| SEXUAL OBSESSIONS | EXAMPLES |
| 21. Content involves children or incest | Unwanted thoughts about sexually molesting either your own children or other children |
| HOARDING / SAVING OBSESSIONS | EXAMPLES |
| 25. I have obsessions about hoarding or saving things | Worries about throwing away seemingly unimportant things that you might need in the future, urges to pick up and collect useless things |
| RELIGIOUS OBSESSIONS | EXAMPLES |
| 26. (Scrupulosity) concerned with sacrilege and blasphemy | Worries about having blasphemous thoughts, saying blasphemous things, or being punished for such things |
| 27. Excess concern with right/wrong, morality | Worries about always doing "the right thing", having told a lie, or having cheated someone |
| OBSESSION WITH NEED FOR SYMMETRY OR EXACTNESS | EXAMPLES |
| 29. (Accompanied by magical thinking, concerned the mother will have an accident unless things are in the right place) Obsessions about symmetry or exactness | Worries about throwing away seemingly unimportant things that you might need in the future, urges to pick up and collect useless things |
| MISCELLANEOUS OBSESSIONS | EXAMPLES |
| 31. Need to know or remember certain things | Belief that you need to remember insignificant things like license plate numbers, the names of actors on television shows |

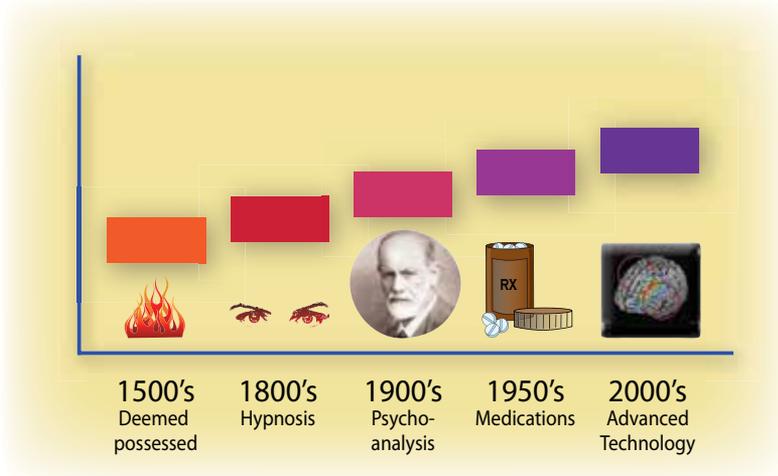
| | |
|--|--|
| SOMATIC OBSESSIONS | EXAMPLES |
| 55. Concern with illness or disease | Worries that you have an illness like cancer, heart disease or AIDS, despite reassurance from doctors that you do not |
| CLEANING/WASHING COMPULSIONS | EXAMPLES |
| 45. Excessive or ritualized cleaning of household items or other inanimate objects | Excessive cleaning of faucets, toilets, floors, kitchen counters, or kitchen utensils |
| CHECKING COMPULSIONS | EXAMPLES |
| 51. Checking that nothing terrible did/will happen | Searching the newspaper or listening to the radio or television for news about some catastrophe that you believe you caused |
| REPEATING COMPULSIONS | EXAMPLES |
| 55. Re-reading or re-writing | Taking hours to read a few pages in a book or to write a short letter because you get caught in a cycle of reading and rereading |
| COUNTING COMPULSIONS | EXAMPLES |
| 58. Need to count and recount | Counting objects like ceiling or floor tiles, books in a bookcase, nails in a wall, or even grains of sand on a beach |
| ORDERING / ARRANGING COMPULSIONS | EXAMPLES |
| 59. Need to order and reorder, arrange and rearrange items | Straightening paper and pens on a desktop or books in a bookcase |
| HOARDING / COLLECTING COMPULSIONS | EXAMPLES |
| 60. Compulsions to hoard or collect things | Saving old newspapers, notes, cans, paper towels, wrappers, and empty bottles for fear that if you throw them away you may one day need them |
| MISCELLANEOUS COMPULSIONS | EXAMPLES |
| 63. Need to touch, tap, or rub | Giving in to the urge to touch rough surfaces, like wood, or hot surfaces, like a stove top; giving in to the urge to lightly touch other people |

Sample SNAP-IV Teacher and Parent Rating Scale # 6160

| For each item, select the box that best describes this child. Put only one check per item. | | Not at all (0) | Just a Little (1) | Quite A Bit (2) | Very Much (3) |
|---|--|-------------------|----------------------|--------------------|------------------|
| 1. | Often fails to give close attention to details or makes careless mistakes in schoolwork, work, or other activities | | | | |
| 2. | Often has difficulty sustaining attention in tasks or play activities | | | | |
| 3. | Often does not seem to listen when spoken to directly | | | | |
| 4. | Often does not follow through on instructions and fails to finish schoolwork, chores, or duties | | | | |
| 5. | Often has difficulty organizing tasks and activities | | | | |
| 6. | Often avoids, dislikes, or is reluctant to engage in tasks that require sustained mental effort (e.g., schoolwork or homework) | | | | |
| 7. | Often loses things necessary for tasks or activities (e.g., toys, school assignments, pencils, books, or tools) | | | | |
| 8. | Often is distracted by extraneous stimuli | | | | |
| 9. | Often is forgetful in daily activities | | | | |
| 10. | Often fidgets with hands or feet or squirms in seat | | | | |
| 11. | Often leaves seat in classroom or in other situations in which remaining seated is expected | | | | |
| 12. | Often runs about or climbs excessively in situations in which it is inappropriate | | | | |
| 13. | Often has difficulty playing or engaging in leisure activities quietly | | | | |
| 14. | Often is "on the go" or often acts as if "driven by a motor" | | | | |
| 15. | Often talks excessively | | | | |
| 16. | Often blurts out answers before questions have been completed | | | | |
| 17. | Often has difficulty awaiting turn | | | | |
| 18. | Often interrupts or intrudes on others (e.g., butts into conversations/games) | | | | |

7. What are the Medical Treatments?

TOURETTE'S TREATMENTS THROUGH THE AGES



1. People in the middle ages with TS were deemed possessed by evil spirits, or the devil, and “treated” with exorcism and/or burned at the stake.
2. 19th century; Dr. Charcot may have tried hypnosis.
3. Early 20th century: Freud’s disciples applied their approach, most notably Sandor Ferenczi reported a case of TS treated with psycho-analysis. It did not work.
4. Second half of the 20th century: most classes of medications have been tried, only a few have been shown to work.
5. 21st century: the use of current “cutting edge” technology-rTMS (repetitive transcranial magnetic stimulation) applying strong magnetic field to certain areas of the brain, DBS (deep brain stimulation) with electrodes implanted in the brain

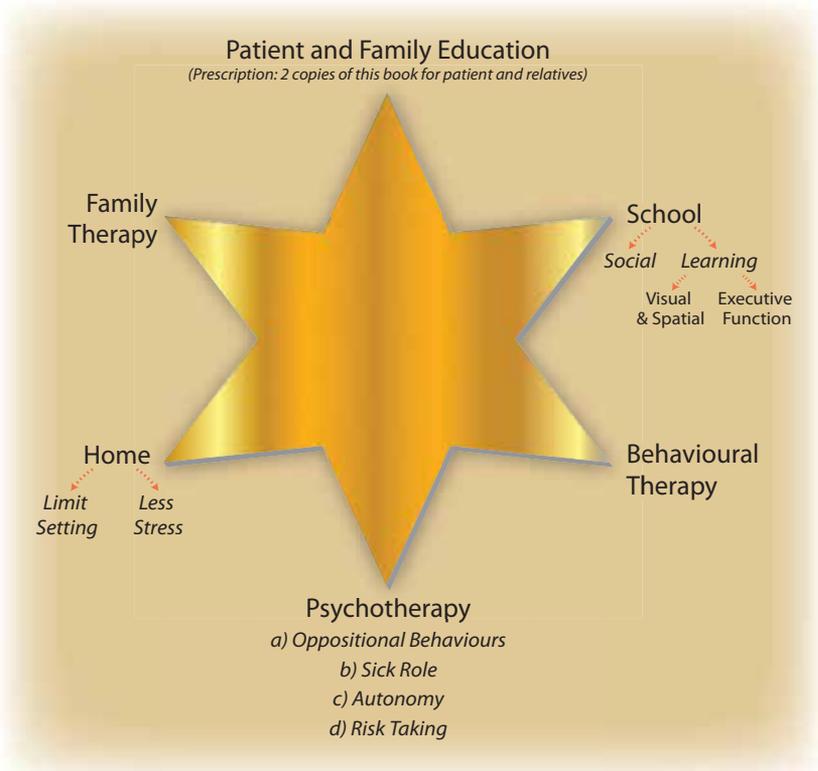
In textbooks of psychiatry it was common to see explanations of the cause of Tourette Syndrome ranging from the psycho-analytic to the biological. While the former is given less credence now, it is still widely recognized that there are both psychological and biological factors contribute to the manifestation of Tourette's. This being the case, it is appropriate to have a treatment/management plan that deals with both the biological and psychological components. Some have suggested that Tourette's is the ultimate psychosomatic condition emphasizing both components, and there are studies that suggest that in some cases, a psychological treatment (a form of “talk therapy”) and a biological intervention (the use of a medication) can induce similar changes in brain function in patients with Tourette's.

Many psychiatrists today think of conditions in terms of the “bio-psychosocial model” emphasizing that all of these components need to be addressed. An important part of the psychological intervention is to facilitate acceptance of the conditions especially by children and adolescents.

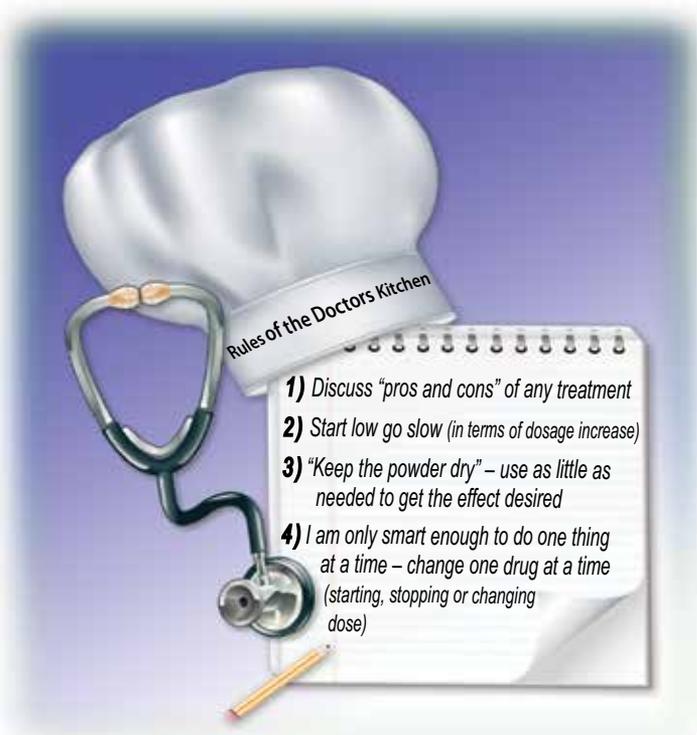
There is also the role of behavioural attempts to manage the frequency and manifestations of tics. It is a common observation that the expression of tics at school and at home may be very different – i.e. that some degree of control can be applied in some circumstances. This behavioural/psychological control can be enhanced.

The purpose of this chapter is to enhance the awareness of the range of treatments – not to map these out in detail.

GOLD STAR PSYCHOLOGICAL TREATMENT



Further information about psychotherapy is provided at the end of this chapter



General rules about the use of medications

Regarding medications and the labels appended to medications, it is important to bear in mind that although a single medication or group of drugs have been known, under one description, the same drug may have different actions and be used for multiple purposes. The most common example may be the medication “aspirin” that was first known as an analgesic (to decrease pain). However today it is probably more widely used (at a different dosage) to thin the blood in order to decrease the chance of a heart attack or stroke. It also is used at times to help bring down a fever. The key issue is that these drugs have the role of altering the amounts of certain brain chemicals which may have many different effects. An example may be the use of anti-parkinson medications for the far more “innocent” condition of “restless legs” in teenagers.

Continues on page 34

The following 2 pages lists the side effects of 6 medications, 4 of which are used for the treatment of Tourette's. The side effects of aspirin i.e. a very well-known medication, is given for the purpose of comparison. The side-effects of Ritalin (Methylphenidate) which is very often used in children to treat ADHD, is also given. This is for comparative purposes and to reassure prescribers, parents and patients. It is notable that one of the side effects of Ritalin, is to potentially worsen tics.

| MEDICATION | ASPIRIN (ASPIRIN) | METHYLPHENIDATE (RITALIN) | CLONIDINE (CATAPRES) |
|-----------------------------------|--|---|---|
| COMMON SIDE EFFECTS | Upset stomach, heartburn, loss of appetite, blood in the stool, bruising more easily, confusion, dizziness, fainting, nausea or vomiting, pain, buzzing or ringing in ears, abdominal or stomach pain, cramping, tiredness or weakness, hepatotoxicity | Nervousness and insomnia, loss of appetite, abdominal pain, weight loss, nausea, dizziness, tachycardia, angina, blood pressure and pulse changes, cardiac arrhythmias | Lightheadedness, dry mouth, fatigue, dizziness, somnolence, hypotension, constipation |
| LESS FREQUENT SIDE EFFECTS | Rashes, hives, fever, vision problems, liver damage, thirst, stomach ulcers and bleeding, trouble breathing, Reye's syndrome, allergic reaction. Hypoglycemia (which has been reported in children) and hyperglycemia | Hypersensitivity (including skin rash, urticarial, fever, arthralgia, exfoliative dermatitis, erythema multiforme with histopathological findings of necrotizing vasculitis, and thrombocytopenic purpural) | Feeling short of breath, even with mild exertion, swelling, rapid weight gain, arrhythmias, flu symptoms, insomnia, confusion, headache, ear pain, nausea, diarrhea |
| RARE SIDE EFFECTS | Kidney impairment, vertigo | May increase tic severity in Tourette's syndrome. Toxic psychosis has also been reported | Mood changes, fever, hallucinations, hyperglycemia, blurred vision, decreased lacrimation (tears). Overdose or intravenous use can cause hypertension |

| MEDICATION | ARIPIPRAZOLE (ABILIFY) | PIMOZIDE (ORAP) | RISPERIDONE (RISPERDAL) |
|-----------------------------------|---|--|--|
| COMMON SIDE EFFECTS | Feeling sleepy, headache, vomiting, fatigue, increased appetite, insomnia, nausea, restlessness tremor, muscle stiffness | Headache, dizziness, dry mouth, restlessness | Insomnia, dystonia, akathisia, extrapyramidal symptoms, headache, dizziness, Parkinsonism, asthenia. Increased dream activity, nervousness, impaired concentration, increased sleep duration |
| LESS FREQUENT SIDE EFFECTS | Constipation, insomnia, uncontrollable twitching or jerking movements, cough | Prolongation of QT-interval. Gingival hyperplasia (increase in size of the gums) which is reversible after discontinuing the drug. Fever, excitement, agitation, irritability, tension, anxiety and nightmares. Extrapyramidal side effects (especially akathisia) | Somnolence, and hypoesthesia, leg cramps, torticollis, hypotonia, migraine, hyperreflexia, choreoathetosis |
| RARE SIDE EFFECTS | Neuroleptic Malignant Syndrome, allergic reaction (such as swelling in the mouth or throat, itching, rash), speech disorder, nervousness, fainting, reports of abnormal liver test values, inflammation of the pancreas | Parkinson's-like symptoms. Neuroleptic Malignant Syndrome, (fever, sweating, very stiff muscles, and confusion). Seizures at higher doses | Confusion, coma, sleep-related eating disorder (SRED), seizures, neuroleptic malignant syndrome, tardive dyskinesia |

Continued from page 31

The two main groups of medications that have been used include clonidine and antipsychotics. Clonidine is described as a centrally active alpha-adrenergic agent. It is still widely used. Both clonidine and antipsychotics were used over forty years ago. The older antipsychotics had some significant and important long-term side effects. For this reason they fell out of favour and were largely replaced with second generation antipsychotics that have become available with significantly less side effects.

At a time when the choice was between clonidine and a group of drugs in which the side effect of tardive dyskinesia (a long-term change in motor behaviour) was relatively common, the preference was to start with clonidine. Now, new agents e.g. Aripiprazole, show great promise in large controlled studies.

DOSES OF DRUGS THAT ARE USED FOR TOURETTE'S

| A. Alpha-noradrenergic agent | |
|---|--|
| a) Clonidine 0.1mg | 0.3mg/day in 3 divided doses |
| b) Guantacine 1mg | 3mg/day in 3 divided doses |
| B. Benzodiazepine agents | |
| a) Clonazepam | Used as an adjunctive treatment 0.5 – 2mg/day |
| C. Atypical antipsychotics | |
| a) Olanzapine (Zyprexa) | 5-10mg |
| b) Risperidone (Risperdol) | 0.5-2mg |
| c) Ziprazidone (Zeldox) | 20-80mg |
| d) Aripiprazole (Abilify) | 2mg – 20mg |
| D. Orap | |
| Pimozide | Adults: 1-2mg/day, taken in divided doses Children 12 years old +: 0.05mg/day per kg of weight Children younger than 12 years old: use and dose must be determined by the doctor |
| E. Botulinum toxin | |
| For specific tics – is of limited use | |
| <i>There are claims for many other agents but those cited above have been most prominent</i> | |

BEHAVIOURAL TREATMENT

The goal of the behavioural treatment of Tourette Syndrome is to help patients to effectively manage their tics. Research studies have provided initial evidence to support the potential efficacy of specific behavioural interventions in reducing tics. The following behavioural interventions are promising stand-alone treatments for Tourette's but they are usually applied in combination, as a treatment package.

| One specific approach is HRT This is akin to a cognitive behavioural approach with steps of: | |
|---|--|
| a) | Self-monitoring of both tics and the urge to have a tic leading to increased self-awareness |
| b) | Creating "set piece" alternatives i.e. movements that cannot co-occur with tics |
| c) | Relaxation or meditation training partly to decrease the stress trigger and partly to gain greater control |
| d) | The process monitors the possibility of a tic occurring with the aim to decrease occurrence of tics |

Habit reversal training (HRT) evolved into comprehensive behavioural intervention for tics (CBIT), which combines habit reversal training with relaxation training, psychoeducation, social support, and functional intervention

1. Awareness training:

Patients have to get to know their tics in order to be able to manage them. The first step in getting familiar with the tics is to identify and describe them in detail. Then the patient has to learn to notice when a tic happens. The therapist and parents (or other supportive individuals whom the patient involves in the treatment) can help the patient in the beginning to detect when a tic occurs. The next phase of the awareness training is self-monitoring. The self-monitoring serves two main purposes: first, it gives a good measure of the frequency and intensity of tics so that changes during the treatment can be tracked. Secondly, by monitoring the situations where the tics occur and recording the events that happen before the tic (for example certain activities, emotions, thoughts, or physical sensations) or after the tics (for example other people's reactions) can reveal which factors make the tics more likely to happen (these factors are called triggers and reinforcements).

2. Relaxation training

The tics and, for some patients, the anxiety related to their disorder, may cause increased muscle tension. Relaxation training helps the patients to reduce muscle tension and anxiety. For example, in progressive muscle relaxation the patient learns to recognize the difference between tension and relaxation in 16 specific

muscle groups. The patient then gradually reduces the number of muscle groups to 8 and then 4 – until he/she learns to achieve deep relaxation by just thinking of a cue word. Patients are encouraged to apply relaxation before entering a situation that is expected to trigger tics (based on the self-monitoring) and when they experience the events and sensations that typically precede tics. It is important to emphasize that relaxation is a skill that can be mastered only by regular practice.

3. Competing response training

The competing response training involves choosing and practicing a behaviour that a patient can perform instead of the tic and which is incompatible with a particular tic. The competing response is usually a subtle contraction of muscles that are opposite to the tic movement. For example, if the tic involves lifting the arm in front of the face, the competing response can be pushing the shoulders down and gently pressing the arm to touch the body. The competing response is always a behaviour that appears to be “natural” and can be performed without the interruption of ongoing activities. Again, as is the case with relaxation, performing the competing response effortlessly requires practice and awareness of the triggers of the tic.

4. Contingency management

Accurate self-monitoring and a detailed analysis of the sequence of events that precede and follow each specific tic helps to reveal the environmental factors that maintain the tics. Some of these factors tend to happen before the tics and will make the tics more likely to occur. Others tend to follow the tics and will give a “positive feedback” to the tics so that they will continue to happen. For example, a monitoring log may show that a tic happens more frequently when a child is doing his homework. When the tics intensify, the parent consoles the child and gives him a break from the homework. In this case, the child and the parent can agree in advance on the duration of the homework sessions and the breaks in between the sessions. The child is also encouraged to ask for help if he has difficulties with the homework and to engage in active problem solving if he finds the homework difficult. The parent will encourage the child to continue working even if he has tics and will praise the child for completing his homework. In other words, the child will learn to stay on the task and engage in effective problem solving instead of being immersed in boredom/tension which may trigger the tics. Also, the tics will not be maintained by the parent's affection and by a break from the homework; instead a desired outcome (completing the homework) will be naturally rewarded.

During treatment the patient and the therapist focus on one tic at a time. Usually, the patient and the therapist make a tic hierarchy based on how distressing each tic is for the patient. The patient then chooses one tic from the top (“most distressing”) range of the hierarchy and works to manage that tic until it comes under control.

The psychological treatment of Tourette Syndrome can be well combined with medication. The advantage of the behavioural treatment is that it helps the patient to be an agent in his treatment; to feel confident that he can control the tics instead of the tics controlling his life.

8. Sleep in Tourette Syndrome

Sleep disorders in children are a significantly under-treated health problem world-wide. Mood, the regulation of attention, the ability to remain alert and problem behaviours have been found to be affected by the amount and quality of sleep. There are many associations among the amount and quality of sleep, mood, the regulation of attention and arousal and social behaviour during wakefulness. Health professionals are recognizing that not only are sleep disorders common in children, but that the presentation in children, compared to adults, can differ significantly. Depending on their age, children sleep as much as 50-65% of their lives, and even small improvements in sleep quality have been shown to manifest in better functional daytime behaviour and improved quality of life.

| All of the sleep disorders listed below can cause poor sleep | Consequences of poor sleep | |
|--|---|--|
|  Large tonsils in a child with Sleep Apnea |  Insomnia |  Hyperactivity |
|  Excessive daytime sleepiness |  Bruxism |  Temper outburst |
|  Periodic Limb Movement Disorder/ Restless Leg Syndrome |  Nightmares |  Conduct disorder |
|  Parasomnia |  Nocturnal Enuresis |  Depression and/ or Anxiety |

There are many causes of sleep problems in children in general

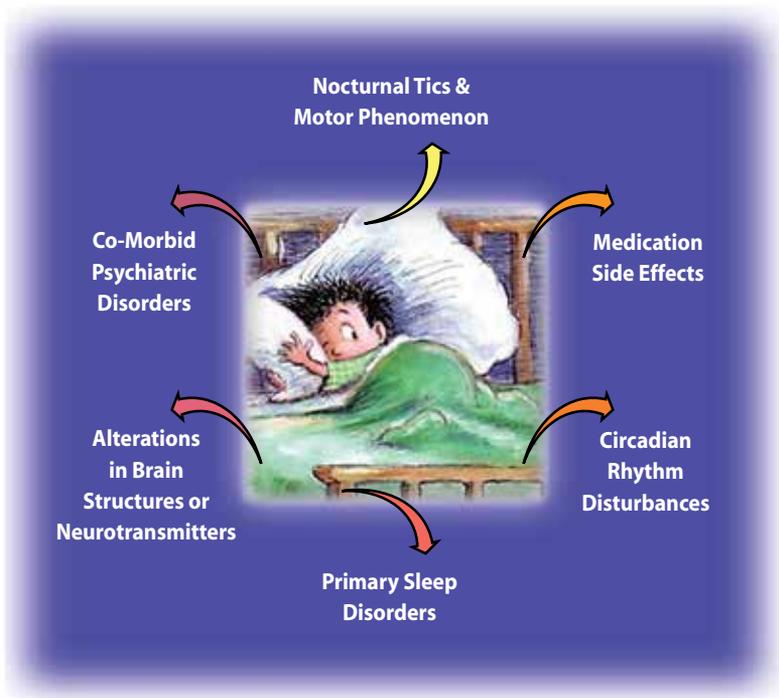
Problem behaviours in children can be a result of, or exacerbated by, disrupted sleep. This reasoning is based on several lines of evidence. Poor sleep has been strongly linked with deregulation of emotion and difficulty with impulse modulation, even in children with no history of behavioural problems. Sleep disordered breathing, restless legs syndrome or disturbed sleep in children can present with hyperactivity, inattention and temper outbursts and mimic attention deficit hyperactivity disorder (ADHD). Lastly, notwithstanding any neurodevelopmental disorder, behavioural issues and attentional difficulties secondary to sleep disturbances can resolve with treatment of the sleep disorder. Hence, the presence of an undiagnosed or untreated sleep disorder can complicate the management of behavioural issues in children with neurodevelopmental disorders.

Among children with Tourette Syndrome (TS), up to as many as 60% of children and their parents report problems with sleep. Several sleep complaints (both subjective and objective) have been reported in children with TS. Children with TS report more sleep disturbances than adults with TS, and this has been thought to be a result of a delay in the normal processes of maturing. However, adults who complain of insomnia often report their symptoms starting during teenage years or younger. In general, parental reports of sleep disturbances in children with TS have been largely unsubstantiated by objective sleep assessment. It is well-established that subjective experiences of sleep such as sleep quality and how refreshing sleep is, are poorly correlated with sleep architectural findings from polysomnography. To complicate matters, to date there are limited numbers of research studies utilizing overnight polysomnography in children with Tourette's. Unmedicated children with TS, but without co-morbid conditions, have been even less well studied.

Sleep disturbances in children with TS can stem from one or more sources (see figure on next page).

- 1) Tourette Syndrome motor abnormalities impinge on sleep at night. Tics have been shown to occur in all sleep stages and are higher in REM (Rapid Eye Movement sleep is often associated with dreaming) compared to non-REM sleep. Forced vocalizations (words, phrases, grunts or bird-like calls) during deep sleep are also frequently seen in TS. Furthermore, there is a reciprocal relationship between sleep and TS symptoms. Sleep is more disturbed in individuals with more severe TS-related symptoms during the day. Likewise, more sleep problems in TS have been shown to result in a greater severity of tics as well as more behavioural problems during daytime hours.
- 2) About 9 in 10 children with TS have a co-morbid psychiatric disorder. The rate of sleep disturbances in TS is greater if children with TS also have co-morbid ADHD or obsessive-compulsive disorder. Co-existent ADHD and TS have been noted to alter sleep in an additive manner, that is, the presence of both disorders results in a greater negative impact on sleep compared to children with a diagnosis of only TS or ADHD alone. Depressive symptoms are also commonly found in TS. Sleep disturbances and depression are closely

CAUSES OF SLEEP DISORDERS IN TOURETTE SYNDROME



interwoven; poor sleep is a hallmark symptom of depression and disturbed sleep is a strong risk factor for developing depression or increases the risk of a relapse of a depressive illness. Anxiety disorders are also seen in TS. Childhood anxiety is linked to sleep resistance (problems with going to bed), increased sleep onset latency, childhood insomnia, nightmares and a greater number of arousals from and/or awakenings during sleep.

- 3) Sleep can also be affected by medications used to treat TS or a co-morbid condition. Anti-dopaminergic drugs are considered to be the standard pharmacological treatment for TS but blockade of the dopaminergic system can result in a number of sleep disturbances, including insomnia, increased light sleep, reduced amounts of deep sleep and REM, excessive daytime sleepiness, restless legs syndrome and periodic limb movements in sleep. To the contrary, treatment with risperidone, an atypical antipsychotic, has been shown to actually improve sleep in children with TS by decreasing the sleep onset latency, reducing the number of arousals and awakenings and lessening the number of movements in both REM and non-REM sleep. Therefore, medications in children with TS must be carefully selected so as to minimize disruption of sleep, and where possible, to ameliorate sleep.

- 4) Tourette Syndrome is characterized by neurodevelopmental alterations in brain structures or neurotransmitters. The manifestation of symptoms of TS is believed to be a consequence of alterations to the cortico-striato-thalamo-cortical circuit in the brain (see chapter 3). Decreased neural activation at the level of the basal ganglia have also been demonstrated. TS may also involve a delay in development or maturation of brain structures. Some authors of this booklet, together with colleagues, have demonstrated the presence of REM behaviour disorder in children with TS and have concluded that TS is characterized in functional alterations of the neural pathways that control motor activity in REM sleep. Others have also implicated a disturbance in the dopamine system for the high incidence of restless legs syndrome and periodic limb movements in sleep in TS.
- 5) Although not well studied, there is evidence to suggest a circadian rhythm disruption in children with TS. Body temperature and sleep are closely linked and interdependent circadian phenomena and circadian dysregulation of body temperature has been observed in TS. Alterations in the circadian rhythm of the hormone cortisol (also called the stress hormone) have also been reported in TS. An altered cortisol rhythm as a consequence of disturbances in the hypothalamic-pituitary-adrenocortical axis has also been reported in TS. Lastly, others have hypothesized that pineal calcification and disturbances in melatonin secretion are linked to the pathophysiology of TS. Further studies are needed to determine if children, and especially teens, with TS have circadian rhythm disturbances, if they more prone to circadian sleep phase disturbances than their peers and whether sleep can be improved with melatonin administration or bright light treatment.
- 6) Disturbed sleep in children with TS may be unrelated to the above factors. About 4% of children are diagnosed with a primary sleep disorder. However, sleep disorders in children remain largely undetected and there are estimates that up to one-third of children may have a sleep disorder or complaint at some point during their childhood. Commonly seen childhood sleep disorders include behavioural insomnia of childhood (bedtime resistance, delayed sleep onset and prolonged night awakenings), sleep breathing disorders including obstructive sleep apnea, night terrors/nightmares, sleep walking or sleep talking, and nocturnal enuresis (bed-wetting). Given the high incidence of sleep disorders in children with TS, a thorough clinical history with an overnight polysomnographic evaluation and daytime assessment of excessive daytime sleepiness before, are essential to characterize the nature of the sleep disorder and determine treatment options. Regular sleep specialist follow-up in children with TS is also needed to detect any changes in sleep with the onset puberty and teenage years.

It is well recognized that children and adolescents with disturbed or insufficient sleep have poor academic performance, impaired memory and concentration, greater behavioural and conduct problems, more mood disorders and poorer quality of life. There is a clear association between sleep disturbances and TS in

childhood. Regardless of the cause of the sleep disturbance, it is crucial that sleep disturbances in children with TS are aggressively diagnosed and managed so as to minimize use of medications that can further disrupt sleep with the goal of achieving optimal management of behavioural and motor disorders in TS.

REPORTED SLEEP COMPLAINTS IN TOURETTE SYNDROME

Difficulty falling asleep

Alterations in sleep duration (too short or too long)

Increased sleep latency

Increased number of awakenings and arousals from sleep (sleep fragmentation)

Prolonged wakefulness after sleep onset

Reduced or absent amounts of deep sleep (also called slow wave sleep)

Obstructive sleep apnea

Sudden intense and partial arousals from slow wave sleep

Greater number of movements during sleep

Periodic limb movements in sleep

Restless legs syndrome

Alterations in REM sleep (decreased or increased)

Increased REM sleep latency

REM sleep behaviour disorder

Sleep talking

Sleep walking

Night terrors

Bedwetting at night (nocturnal enuresis)

Poor sleep quality

Non-refreshing sleep

9. What are the Social Problems? – A Patient's View



I have a friend with full-blown Tourette's who has to shout out profanities every thirty seconds. He also has to physically touch people in the genital area. Fortunately only ten percent of people living with Tourette's live with this extreme severity level. People with full blown Tourette's tend to surround themselves with family members and friends who know about, and fully support, their completely antisocial behaviours.

Their social problems occur only when they are in public. Train rides, movie theatres and being in any crowd can be dangerous for them because there will always be people who have absolutely no clue what Tourette's is. They feel threatened by the wild bizarre movements, shouts, swearing and rude gestures that my full-blown TS peers constantly exhibit.

The social problems I live with apply more to the remaining 90% of us who do not live with coprolalia i.e. swearing. What I am about to speak about resides in the realm of shame. Therefore what you are about to read is something even I normally keep hidden and secret.

Thanks to my childhood experience of constant schoolyard bullying, rejection and ridicule, I am left with a deep and abiding belief that I am no good and completely unlikable. Please take into account that when everyone in your school peer group rejects and ridicules you and then physically abuses you, you start to believe that you are completely unworthy and unlikable. Furthermore you unknowingly start to reject and ridicule yourself.

Living with Tourette's at the very least means that conversations and interaction with me are as fractured as they are intense. In the period of five minutes it feels completely normal for me to ricochet through five to ten different subject matters expressing emotions from laughter to anger. All of this feels completely normal and in social situations I have to forever focus on trying to slow down and focus on one subject that is being discussed.



Children often ridicule a peer with Tourette's

As remarkably merciless and hurtful children were to me, I find that adults are the complete opposite. Thanks to the media showing people living with Tourette's, it has been my extreme pleasure to experience that just about everyone I meet is either fascinated by or is pleased to actually meet someone living with the disorder. Thanks to this experience, I decided twelve years ago to become a professional speaker. From a solitary platform I am able to reach out to thousands of people and use my Tourette's to inspire and change people for the better.

Tourette's is no different from any other social disorder because it forces an individual to either deal or not deal with their situation. My epiphany breakthrough came when I was participating in a group therapy session. I was deeply depressed and actively suicidal. During a morning session it felt as if I was struck by lightning with a new knowing to focus my life on helping other people before helping myself. I know people with Tourette's who hate their Tourette's and are socially abhorred, and others with the same level of affliction who become very social and successful.

This does not change the experience that in every new social situation I always feel as if the people around me are about to find out what a truly offensive person I am. The message within me is always screaming, "it's just a question of time until these new people start to realize how unlikable you are!" My childhood tormentors tattooed this response deeply into me. As a kid I chose to fully and unquestionably believe them.

Deep within the shame element of Tourette's lies all of my anxious and obsessive compulsive feelings. I feel anxious every moment of my life. The only reason I am socially acceptable is because I have somehow learned to embrace and come to terms with my social clumsiness. Today as an adult I know what I need to do and observe in order to appear normal around non-twitchy people.

Every moment of my life feels as if I have just done something terrible and something awful is about to happen. I know rationally that this is not the case. I also now know these irrational thoughts feel absolutely true and real.

My suggestion to kids is this: if you are experiencing these types of thoughts and feelings, try opening up and talking about your Tourette's, your twitches and your inner feelings to your parents and doctors. Try sharing how you are feeling inside just once! Your mum and dad unconditionally love you and you can always trust a doctor. How I wish I had known this when I was a kid living with Tourette's. I foolishly kept all of my bad feelings and anxious thoughts hidden away inside of me. In my own silly way I thought these feelings were normal and at the same time I felt so shameful and badly about these thoughts that I just kept them hidden away. I foolishly thought that if I shared that I was thinking sad, anxious and negative thoughts that my parents would no longer love me. I was so wrong. I also remember never telling my parents about how every day at school I was being bullied, rejected and ridiculed. Trust me and learn from my mistakes. If you share your anxious thoughts and feelings with others that you either trust or love something wonderful happens. You have a way to release and learn how to, at the very least, minimize and lessen your anxious and obsessive thoughts.

10. School-Related Problems

Unlike the experiences described in the previous chapter, the majority of children with Tourette Syndrome whose symptoms are relatively mild, may go undetected – and will not suffer the social and psychological torment. These children with ‘milder’ TS may have some academic difficulties but are more likely to have organizational deficits, issues with handwriting or fine motor problems, be more aggressive or oppositional than their peers, have difficulty relating to peers, and be more unhappy or depressed. As a consequence, despite the impact on learning and social interaction with peers, these children may ‘fly under the radar’ and may not be brought to the attention of guidance counsellors or school psychologists which is a common pathway to medical detection and care. On the contrary, children with more severe Tourette’s or those with one or more associated disorders (e.g., ADHD, OCD) are more likely to receive diagnosis and management when they overwhelm their teachers’ ability to cope with their learning difficulties, behavioural challenges and emotional dysregulation (emotional response that is poorly modulated). In a previously presented vignette geared towards providing teachers with information about TS, the authors describe a 10-year-old boy, Z., living in a rural area, who for the past two years had been made to sit at the back of the class so that his phonic tics would not disrupt the class. The teacher would remind students regularly to notify her immediately if Z. was irritating them, etc. As a consequence, over the years Z. had not been invited to any play dates or parties by his fellow classmates. Moreover, Z.’s IQ dropped by 20 points on subsequent retesting.



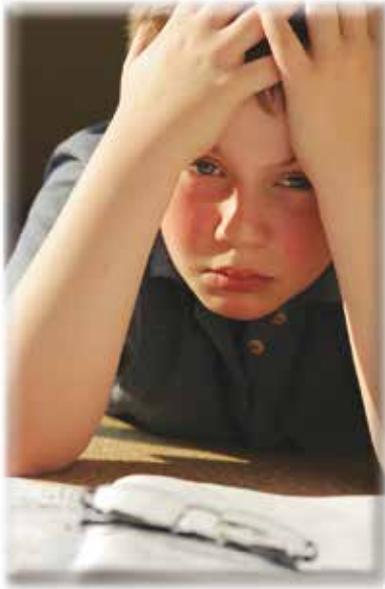
PEER PROBLEMS AND SOCIAL WITHDRAWAL

Children with TS are embarrassed by, and frustrated with, trying to suppress their tics. More severe forms of TS and associated disorders such as ADHD have a well-recognized negative impact on children’s ability to interact with their peers. However, a research study

Teasing about Tourette’s can lead to social withdrawal and low self esteem

exploring social adjustment and peer relationships noted that even children with mild and moderate TS may be more withdrawn, more aggressive and less popular than their classmates. Moreover, these social problems are not related to the severity of tics and the presence of co-morbid ADHD by itself was not a predictor of poor social adjustment.

The vignette regarding the boy Z emphasizes the social isolation experienced by children with TS. Ironically, being labelled as the class pariah and feeling looked down upon by others may make children with TS act out in school to be noticed and get attention, regardless of the self-defeating aspects of this behaviour. Social development and ability to interact positively with peers may be further hampered if children with TS are forced to undergo home instruction because of their disruptive behaviours and inability to function in the classroom.



Attending in class is often problematic

LEARNING/ATTENTION

Children with Tourette's, particularly those with attention problems, will have difficulty focusing on the school task at hand as they are easily distracted by activities occurring outside the window or by other children in the class. Constant instructions to pay attention will not be helpful to a child with a neurodevelopmental disorder such as Tourette's, and will further degrade an already low self-esteem by drawing the ridicule of classmates. Despite evidence of normal intelligence and IQ scores in children with TS, they are more likely to experience a number of learning difficulties and visual-motor impairments. These may include trouble with handwriting, difficulty with copying from the board or a book, poor mathematical skills, spelling deficits, issues with reading comprehension and impairments with written expressive language. Deficits

in executive function can lead to children with TS appearing lazy, careless, irresponsible and disorganized. As a consequence, children with TS may find routine school work to be overwhelming and consequently have trouble keeping up with the rest of the class, be unable to complete and turn in homework assignments, forget to give parents important notices from the teacher or school and regularly lose track of possessions.

DISRUPTIVE BEHAVIOURS

Multiple motor, vocal or phonic tics that occur throughout the day, especially in children with more severe TS, can be very disruptive to discipline in the classroom. Facial grimaces and grunting noises that are sometimes punctuated by loud animal-like noises or curse words, can result in fellow classmates laughing and making rude comments. Children with TS are also prone to aggression and temper outbursts. Getting a child with TS to sit still in class can be almost impossible for those with attention deficits or hyperactive tendencies. Impulse control issues that result in socially inappropriate acts such as constantly touching other students or their possessions can result in physical altercations in the classroom. Furthermore, tics

and other disruptive behaviours can be enhanced during periods of increased stress such as during oral recitations in front of the class and during tests. The usual course of action is for schools to suspend children with TS for disrupting the class. In a survey of children with TS, three-quarters of respondents stated that tics posed a moderate to significant interference with school and almost half of the children attributed the peer rejection they experienced to their tics.



Bullying, including on social media, can lead to depression and an added burden to Tourette's

VICTIM OF TEASING & BULLYING

Because of their neurodevelopmental challenges (most notably tics, hyperactivity, impulsivity and social skills deficits), children with TS are at greater risk of being the victims of teasing and/or bullying. Types of bullying behaviour include: threatening gestures; pushing, shoving or poking; imitating the child; verbal abuse (includes incessant or malicious teasing); and physical confrontation. Furthermore, bullying can be very subtle and remain unnoticed by the teacher. Children who are victims of bullying experience isolation, low self-esteem, depression, and avoidance which is not helped by other disorders that are more common in TS kids, such as ADHD and OCD. They may also suf-

fer from side effects from medication they need to take for Tourette's, such as weight gain. In the worst case scenario, these children may even have suicidal thoughts. These negative emotions compound the low self-esteem and feelings of social isolation commonly experienced by children with TS. Children will often not report that they are being bullied because they are embarrassed, wish to protect their parents from hurt, fear that they won't be believed or are afraid that the abuse will worsen if other children find out that they have 'tattled'. It often falls to teachers to monitor events in their classroom and to pay special attention to children who appear to be loners and who are having difficulty making friends. However, larger class sizes and increasing percentages of children with behavioural issues place considerable demands on teachers' time, thereby rendering it less likely that teachers will detect incidences of bullying in their classes.

LATENESS DUE TO EXCESSIVENESS DAYTIME SLEEPINESS (EDS)

As detailed in chapter 8, children with TS experience significant issues with sleep and daytime wakefulness. Parents may find it difficult to awaken children with TS in the morning and hard to stop them from falling back asleep instead of getting ready for school. Children with TS may find themselves being sent often



to the principal's office and collecting numerous disciplinary citations for repeated lateness. Even those who are able to get to school on time may find it difficult to remain awake and focused during the first few periods. This problem may be exacerbated in later childhood and adolescence where there is the tendency for most children, even those without mental health concerns, to want to go to bed later and awaken well past school start times.

11. What are the Social Solutions? – A Patient's View

Doctors, parents and kids living with Tourette's please let me remind you: many people react badly to a person with Tourette's. This is because our twitches, sounds and wild movements scare them. I invite you to see Tourette's from their perspective. Many adults have never met anyone who is twitchy before. For some people it's alarming, interesting, amusing - and for some it is scary.

As you grow up there will be many people who test, reject and ridicule you. In those moments you will feel lost, angry and alone. I know that I did, so let me promise you this: as you get older it does get better and more importantly, please know that there is a special power and strength that is growing inside of you.



There is a South African expression: " 'n Boer maak 'n plan", which means "A farmer makes a plan". It conveys the idea that one can always find a solution for a problem. This is what this graphic conveys, that a TS patient can always find a solution

For those of us living with full-blown swearing Tourette's, the solution is to plan your social time and situations where you have as much control as possible. My full-blown friends do not travel alone on trains or busses. They always have a friend along who can explain to anyone what is happening.



Our third author, Stuart Ellis-Myers, who has Tourette's, wrote several chapters in this book

enjoy being a living, breathing solution for the mass socialization of Tourette's because my work allows me to reach out, speak and be understood by thousands of people.

For adults living with Tourette's the social solutions have already been found. Just about every person I have met over twenty years of age has come to understand and at the very least know how to manage the social aspects of this disorder.

Where the social solution remains a problem is within the school system. Bullying is a runaway epidemic without a solution. Social media has exacerbated the

They prefer their social time to be with family members and friends who love and support them. In many cases new friends can be easily made by being introduced to acquaintances of friends. There are many people who are brought into the Tourette community because when first exposed to the uniqueness of our bizarre behaviours, these non-twitching people think that we are cool and fun to be around.

I chose to become a professional speaker twelve years ago. In the beginning I foolishly tried to hide my Tourette's away. Today I proudly bill myself out as, 'Robin Williams with a Twitch!' I

situation and many children who have been constantly bullied at school have resorted to suicide.

The most important social solution is for newly-diagnosed kids and their families to get involved with their local Tourette Syndrome Association chapters.

Over the years I have met and worked with many mums and dads of children living with Tourette's. When I speak at professional conferences to large groups of seven hundred people or more, there are always a few people who come up to meet me after my performance. These are either parents or people related to someone living with Tourette's who feel they are somehow to blame for a child's condition. My message to these people is: Stop! Just Stop! Forgive yourself for having a kid with Tourette's because there is nothing to forgive! This situation just happened to you and it was not purposely created by you! I also ask TS parents to just let go. Just to let go and relax! I invite them and others involved with Tourette's kids, to realize and accept just what an amazing job they did/are doing raising and supporting a kid with Tourette's.

Most parents I know feel terrible when their perfect non-twitchy child starts to twitch and is ultimately diagnosed with Tourette's. I invite all mums and dads living with newly-diagnosed kids living with Tourette's to learn to accept the new situation. If there is one thing that I do hate about Tourette's it is this: Tourette's does not show itself straight away. There are always minor telltale signs and symptoms that a child has Tourette's but these symptoms can so easily be diagnosed as a bad cough or another illness that can explain away a tic or a twitch.

Whenever I meet mums and dads of a newly-diagnosed Tourette's kid, I like to lightly knock them a few times on their head to take notice and remember that their child living with TS will have an okay and probably wonderful life.

REACH OUT & CONNECT

Please consider contacting your local Tourette Syndrome Association Chapter for support. These are listed for you in the chapter on resources (chapter 14). Here you can find many mums, dads and other children happily living with Tourette's. They share their experiences and give advice on how you too can live happily with Tourette's. I have planned many a bowling event for families with Tourette's. There is nothing better than to welcome kids newly diagnosed with Tourette's to one of these events. 'First timers' always show up feeling nervous thinking badly about their Tourette's. In minutes of being surrounded by other kids all happily bowling, twitching and sounding off, they begin to realize that they are not alone and that Tourette's is actually not that bad!

12. How can Schools Help?

Despite the bleak educational picture associated with having Tourette Syndrome, children with TS and co-morbid disorders can be intellectually gifted, extremely creative and/or possessing an obsessive perfectionism that manifests in extremely detailed projects and papers in schools. With proper schooling, children with TS can look forward to exceptional careers. One actor who had Tourette Syndrome as a child, is Dan Aykroyd. Historical figures including Samuel Johnson, the renowned British writer, exhibited symptoms consistent with TS. It is incumbent on school officials, educational administrators and teachers to provide a school environment that encourages children with TS to learn so as to minimize the negative aspects of their disorder and achieve their maximal academic potential and social development.

Given the relatively low prevalence of TS and the unique manifestations of the syndrome in each child, most school psychologists, teachers, guidance counselors and nurses may only encounter a handful of children with severe TS during their career. Nonetheless, early educational experiences are a crucial framework for later years as symptoms in the majority of children with TS will significantly improve or remit by late adolescence. The challenge for educators is to find innovative ways to deliver the curriculum in the face of learning or behavioural difficulties while promoting acceptance and building self-esteem in children with TS. Failure to do so will produce dysfunctional adults with low self-esteem, poor social skills and few marketable skills to employers.

SCHOOL ADMINISTRATORS & OFFICIALS

School officials and administrators need to provide opportunities for – and encourage – teachers, guidance counsellors and nurses to avail themselves of specialized training in identifying and managing children with mental health disorders such as Tourette Syndrome. Management of children with



TS needs to also involve allied educational staff including playground attendants and school bus drivers as problem behaviours are not limited to the classroom and bullying and fights often occur outside of class. Finally, most school boards, in conjunction with local or regional governments have set up an Identification, Placement and Review committee (also called the special Education Testing and Advisory committee) composed of representatives from local and community associations and school board officials who employ necessary healthcare and educational professional resources to develop and implement a special education plan. The following are generally the aim of these plans: identifying the academic behavioural or social problems a particular child is having; conducting a functional assessment of the child that can guide intervention and set goals for instruction; planning the intervention strategy to enable the child to learn and function in an academic and social environment; determining the appropriate school placement for the child; and, arranging for continuous monitoring of the child's progress based on the goals set forward in the student care plan. Parents, and students older than 16 years of age, are generally allowed to be present and to participate at all committee discussions and when decisions about identification and placement are made.

SCHOOL PLACEMENT OPTIONS FOR KIDS WITH TS

- Most kids with TS are in a regular class and doing well, some even excelling
- Placement in a regular class but with the teacher receiving specialized consultative services
- Placement in a regular class for most of the day but with the child receiving additional specialized instruction by a special education teacher within the regular classroom setting
- Placement in a regular class for half of the school day and instruction from a special education teacher for the remainder of the day
- Placement in a regular class for at least one period followed by attendance at a special education class with a low student to teacher ratio
- Placement in a full-time special education class for the entire day
- For children with severe learning disabilities, specialized schools have been set up to cater to their learning needs. However, assessment by a higher level committee is required to determine eligibility for admission

Principals who, based on feedback from teachers or guidance counsellors, judge that a child will benefit from a special education program, are the commonest route of referral for the above-mentioned committees for assessment. The assessment determines the needs of the child. However, many school boards also mandate that a principal must request a meeting between a child and the Identification, Placement and Review committee upon receiving a written request from parents.

Apart from the special education plan and regardless of placement, it is a good idea for school officials to take advantage of initiatives that are geared towards improving the school experience for the child with TS. Examples of such initiatives are listed below.



BULLYING

School officers need to set up and enforce a zero tolerance policy on teasing and bullying and inform the student body that there will be severe consequences for the above mentioned behaviour. The Olweus Bully Prevention Program is an evidence-based best practice model of bully prevention that can be implemented by schools to set rules against bullying, to develop sanctions against bullying that can be consistently followed by school staff, and to create school-wide education programs on what bullying is and what to do when bullying occurs. Furthermore, schools can provide a support group comprised of guidance counsellors, school social workers, nurses and psychologists who are trained in bullying intervention techniques, and who can communicate with and monitor children who are most susceptible to being bullied or teased. It is equally important for schools to impart to children, including those with TS, that they do not deserve to be bullied and that the school is willing and ready to work with them and the bullies to end the bullying. Administrators need to identify individuals at the school that bullied children can speak to if the problem continues and they need to work with the school counsellors and teachers to develop a safety plan.

STIGMA

Schools can help reduce the stigma of having TS (or other developmental disorders such as ADHD) by promoting awareness and by encouraging input from parents and health care professionals. One possibility could be to provide information flyers geared towards classmates and their parents learning more about TS and could include social events such as fundraising bake sales and a themed yearly mental health awareness day.

LATER SCHOOL START TIMES

At the onset of early teen years, adolescents will frequently start to exhibit behaviours of wanting to go to sleep later and awaken well past school start times. This sleep phase behaviour leads to frequent conflicts among sleepy teens, parents who find it a chore to awaken their children in time for school, and teachers who find students wandering in well after the start of class, especially in the first two periods of the day. Children with TS face similar issues with poor sleep and difficulty awakening. A number of schools in Canada and the US have successfully adopted later start times to better accommodate sleep phase delay behaviour in adolescence. Later start times have been shown to result in less late days, better grades, and an overall improvement in school functioning of the student body. In particular, children with TS and other mental health challenges would also greatly benefit from a shift to later school start times. While it may be difficult, it is not impossible to implement later school start times especially in the face of very positive findings from schools that have done so. At the very least, special education programs for children with TS could selectively feature later school start times.

TEACHERS, GUIDANCE COUNSELLORS, NURSES AND SCHOOL PSYCHOLOGISTS

It can be very frustrating for school employees to deal with the learning and behavioural issues in a child with TS. However, teachers need to realize that disruptive behaviours are occurring involuntarily and to not react with anger or annoyance. School educational and healthcare workers can empower themselves by



Extra help for kids with Tourette's may be needed

taking advantage of training programs that have been developed by local chapters of Tourette Syndrome Associations (e.g., "Educating Children with Tourette Syndrome: Understanding and Educating Children with a Neurobiological Disorder" developed by the Long Island Chapter of the Tourette Syndrome Association Inc.). Many of these documents are available online and have been developed to provide teachers and school healthcare workers with information and resources to better meet the educational and social needs of children with TS and to better cope with the impact of TS on children's academic functioning and interaction with peers.

Giving the child with TS short breaks out of the classroom can reduce class disruption by helping the child to relax, release tics that they have been trying to suppress and enhance their ability to focus. Allowing children with TS to take tests in a private room and eliminating recitations in front of the class can help reduce their stress. A number of accommodations can be made to help children with writing difficulties. A few of the more notable ones include: reducing the number of assignments that need to be copied from the board; allowing the child to present a taped rather than written assignment; assigning a note-taking buddy to provide a carbon copy of class notes to help students with visual-motor problems get important information on paper; providing both written and oral instructions; and eliminating the stressful time restriction from tests.

Teachers and guidance counsellors should also make a point of speaking to the class of a child with TS. Providing classmates with a better understanding of what TS is will go a long way towards setting a tone of understanding and acceptance in the classroom. Teachers should also encourage children to tell them or guidance counsellors about any difficulty with bullying or teasing they are experiencing.

Frequent communication between educational workers and parents of a child with TS is essential. It is all too easy for educational workers to react negatively when meeting with parents of children with TS. Teachers, in particular, may either feel that parents are being critical of their skills in managing the classroom or are simply making excuses for a problem child. The reality is that parents are often as frustrated as the teachers in trying to manage a child with TS, but parents are the best source of information about their child, and educational workers can use the parents' experiences with a child with TS to better help manage the child at school. Parents can also be asked to address the class to help classmates understand problems that their child with TS is having.

To summarize, students with TS pose a significant challenge for school officials and educational staff. Ideas to help school boards and teachers address this challenge are listed above. The goal of educational professionals, parents and healthcare providers is for children with TS to find school to be a safe, nurturing and a therapeutic environment for learning.

13. How Much Difference do the Solutions and Treatments Make? – A Patient's View



Social interaction with other people with Tourette's can be very helpful

I have been around Tourette's for many years. The best solution I have ever seen is where we hold local bowling events where newly diagnosed kids with Tourette's are forced by their parents to attend. The last thing any newly diagnosed kid with Tourette's wants to do is to be around other people living with the same condition. This is because they have inadvertently and wrongly chosen to see Tourette's as a bad thing. How can anyone blame a kid for feeling otherwise? See it from their perspective. One day they are normal and then over a period of time that is differently experienced, generally around the age of 7, everything changes.

You start to feel anxious, stressed, angry, bewildered and uncomfortable in your body. The obsessive thoughts make it difficult to even put a sock on as the lie of stitching across the toes can feel absolutely awful! Then you start to twitch and make vocal sounds.

Time after time when we take a newly diagnosed kid and unceremoniously drop them into a large group of twitching people the result is absolutely magical. A newly-diagnosed twitchy kid when surrounded by others just like him having a fun

time is always a life-changing experience. They leave our bowling events smiling knowing that they are not alone in this affliction. They also keenly observe parents and twitchy old people like me doing well in life. They instantly start to let go of the shame normally associated with Tourette's and replace that darkness with the knowledge that they are now a part of a group of people who can, and are, living life well.

When it comes to the medical community I blame the current system which is over-stressed and under-resourced. Doctors only have 10 minutes to see a patient and can so easily misdiagnose Tourette's. Correct diagnosis can take years as the symptoms of constant throat clearing are misdiagnosed as a local physical



Sometimes Doctors are in too much of a rush

problem versus a neurological one. Thanks to the relative rarity of the disorder, specialists are few and far between and normally found by conferring with other parents who have children with Tourette's. The worst part is seeing so many kids being given needlessly high levels of medication.

I was asked recently if the treatment of the tics and

twitches was as important as dealing with the anxiety. My response as someone living with Tourette's for forty-six years would be to say "absolutely". If you can relieve and diminish the tics, twitches and bizarre sounds and gestures we make, then something magical happens. The kids living with Tourette's have a chance to feel more normal and their natural stress level is reduced.

In my case I take an antidepressant as it assists my dopamine level and helps me reduce my anxiety level. I was foolish in thinking that "toughing it out" was the best solution. I tried many different types of drugs, and then only when I was very anxious and depressed. In other words I am suggesting that if you live with Tourette's, try experimenting with the different medications that are available by prescription when you are feeling less twitchy and anxious. Most people give up too soon as the process of finding the correct cocktail of medications and prescription levels can take years to get right and then the balance needs to be constantly maintained or updated.

For the past twenty years I have been a devotee of group therapy and self-help. This started right after the day my first wife told me she was not happy in our relationship and no longer loved me. What was worse she had felt this way now for over a year. Behind all this was her family who did not want her to have “twitchy babies”.

I share this because in everyone’s life comes an absolutely devastating situation that can cause one to break. I believe that Tourette’s simply amplifies the anxiety and depression that comes with any traumatic situation. Cognitive therapy was my first foray into working on the actively suicidal thoughts that came out of that experience.

Looking back over my life with Tourette’s, I am now a firm believer that there needs to be a constant balance of medication and psychological support. In my case I have learned how to go from failure to failure with great enthusiasm. I tried sitting around feeling twitchy and sorry for myself. That choice did not work. Fortunately for me I chose to try the medical and psychological options that were available.



Group therapy with or without other Tourette’s patients can be very beneficial

14. Resources for Patients, Parents, Schools and Health Care Professionals

Parents, especially mothers, consistently state that they are under considerable stress trying to parent their child who has TS, care for their family and ensure that their child receives the necessary services to enable them to do well in school and society. One of the primary causes of frustration and stress is the ongoing search for information about TS and what resources are available to children and their families. Hence, it is crucial for agents and caregivers of children with TS (or children suspected of having TS) to obtain easy access to information about what community, healthcare, counselling, school and government services are available for them and their child.



Tourette Syndrome
Foundation of Canada
La Fondation canadienne
du syndrome de la Tourette

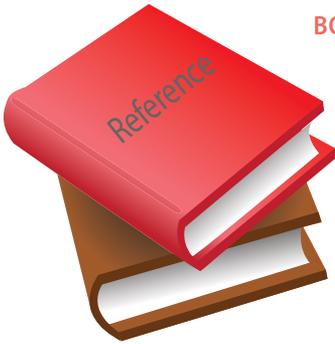
Tourettes ✖
action

ASSOCIATIONS

The best and most authoritative source of information in Canada is the Tourette Syndrome Foundation of Canada: www.tourette.ca. In the USA it is the Tourette Syndrome Association Inc.: www.tsa-usa.org. In the UK: <http://www.tourettes-action.org.uk>. In Europe: <http://tourette-eu.org/en/european-society-for-the-study-of-tourettesyndrome.html>. Each of these organizations have local or regional chapters that provide information about TS, resources, newsletters, videos, etc. The TSA local chapters can also help parents and edu-

cational staff locate local health care providers who have experience diagnosing and treating children with TS. Local TSA chapters are also invaluable in providing support and identifying community resources to help parents cope with managing their child with TS, point out counselling services available to children and their families, provide tips on how to enhance their child's quality of life and assist with meeting the needs of other family members.

Various country-wide, regional and local child and youth mental health associations are also available (usually found by internet) to assist families in finding local agencies, hospitals and clinics that specialize in assessment, consultation, social support and rehabilitation services for children with TS. These agencies can provide access to a variety of mental health services including primary health care services (physicians, nurse practitioners), community mental health services, hospital services and forensic mental health services for individuals who are having issues with the criminal justice system.



BOOKS

There are in excess of 2,700 books on sale that cover various topics related to Tourette Syndrome and are geared towards a variety of audiences. A few of these are also available in Kindle and Audio format. Parents, caregivers and educators of children with TS need information on Tourette Syndrome to better understand the disorder, to assist them in parenting their child, to make better health and school choices and to be a better advocate for their

child; books are one of the best resources for that purpose, and in particular, self-help books containing practical tips and ideas can prove invaluable. Children with TS are also better served by learning more about their disorder and there are many books geared towards informing children about TS and sharing the experiences of other children with TS. One book seller, Caversham Books (<http://www.cavershambooksellers.com>), focusses on mental health issues. Public libraries are a valuable resource but most are limited in the number of books they have available for loan. Book stores such as Chapters and Amazon have the widest selection of books for sale. Although children's books featuring TS tend to be less costly, the average cost of a self-help book for TS is around \$25.00. The drawback is that very few books providing information on TS are below \$15.00, and the vast majority cost well in excess of \$20.00. Used books, where available, are a much cheaper option. The ideal self-help book would be informative, fairly comprehensive and easy to read while costing around \$15.00 (this book, for example!). The names of a few of the many books available are: *Coping with Tourette Syndrome – A Workbook for Kids with Tic Disorders*, by Sandra Buffolano, *Nix your Tics* by B. Duncan McKinlay, *The Tourette Syndrome & OCD Checklist* by Susan Conners, *Educating Children with Tourette Syndrome: Understanding and Educating Children with a Neurobiological Disorder*, developed by the Long Island Chapter of the Tourette Syndrome Association Inc., and *A Family's Guide to Tourette Syndrome* edited by John T. Walkup, Jonathan W. Mink and Kevin St. P. McNaught.

INTERNET

The Internet contains a plethora of information about TS, and as a plus, most resources featured on the internet are free. Be mindful though, that not all information from the internet is correct or supported by scientific evidence. For families who do not own computers or do not have internet service, public libraries often have computers available at no cost for those with a library card (most recommend reserving a computer to avoid long wait times) and



librarians can assist novices with searching the internet. Individuals with laptops can find free Wi-Fi internet access at several popular fast food venues.

Parents and educators can, for example, search for and view strategies employed by other educators (http://www.tsa-usa.org/aeduc_advoc/educ_ed_strat_main.htm); obtain tips on dealing with tics in the classroom (<http://www.schoolbehavior.com/disorders/tourettes-syndrome/>); and find out more about individual education programs available from their local school boards (<http://www.edu.gov.on.ca/eng/general/elemsec/speced/iep/iep.html>). They can also get news on academic studies, for example <http://hms.harvard.edu/news/reducing-tourette-tics-8-8-12>.

The internet can also help to reduce the stigma associated with TS by identifying sites that share the experiences, poetry and writings of children with TS and their parents (e.g., Poetry: <http://www.tourettesyndrome.net/index.php?s=poetry>; Voices of Tourette Syndrome: (<http://well.blogs.nytimes.com/200.02/10/the-voices-of-tourettes-syndrome/>). Facebook and Twitter are also increasingly being used to bring about awareness and understanding of TS by putting a face to the disorder and alerting others about TS-themed social and fundraising events being organized by individuals or organizations.

Used under parents' supervision, sites aimed at children (e.g., Kids Quest – Tourette Syndrome operated by the Centre for Disease Control and Prevention: (<http://www.cdc.gov/ncbddd/kids/tourette.html>) can provide children with TS and their peers with kid-friendly information about TS, help them locate other kid-friendly resources and identify movies and books that can help them further with their study of TS.

1-800-668-6868 KIDS HELP PHONE / HELP LINE

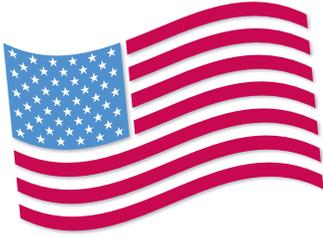
1-800-55-1800

0300-777-8427

0800-543-754

Most countries/cities have community-based charitable organizations that provide toll-free, 24-hour, anonymous and confidential phone and on-line professional counselling services for children and youth. Youth are encouraged to call and the 'anything goes' mission offers children a range of assistance for various issues ranging from homework difficulties to thoughts of suicide.

These organizations are also able to connect kids with community professional counsellors. The anonymity of the phone call has the added benefit of helping to erase the hesitancy to reach out due to the stigma of mental health problems while providing immediate reassurance and feedback for children who are shy or reluctant about discussing their mental health concerns. In Canada the Kids Help Line number is **1-800-668-6868**. The TS helpline in the UK is **0300-777-8427**, Australia **1-800-55-1800**, New Zealand **0800-543-754** and the USA **1-800 55-1800**.



GOVERNMENT-BASED RESOURCES

A number of clinical community healthcare agencies, such as the Youthdale Treatment Centres in Toronto, Ontario, Canada are non-profit and receive funding from the government to provide comprehensive and integrated outpatient and inpatient assessment, consultation and management, crisis support services and residential treatment programs for children with mental health challenges. Such facilities are able to provide children and their families access to services of psychiatrists, child youth workers, social workers and psychologists whose fees are covered by government healthcare funding (in Canada). Youthdale also works closely with educators to ensure that children in residential treatment programs continue to receive education specially geared towards their needs.

Some governments also provide funding for the education of students who need specialized programs, services and equipment. For children whose educational needs cannot be served in the public schools, private special-needs schools with small and individualized classes offer alternative education and specialized support for students with any kind of physical disability, behavioural disorder, mental disability or communicative disorder. Such special education private schools are well-equipped to support children with Tourette Syndrome. The major drawback is that if it is determined that a child needs to be placed in a private special education school, the tuition for these schools can pose a financial burden for the family. The Canada Revenue Agency allows a certain portion of tuition paid to attend such private schools to be available as a charitable donation. For children with special needs who attend special education schools with specialized staff, parents may qualify for a medical tax break on tuition. The Internal Revenue Service in the US has deemed that tuition for special schooling for children with disabilities can be claimed as a deductible medical expense. Along these lines, most countries also allow tax credits and deductions for children with special needs. Many private schools also offer scholarships and bursaries to help families cover tuition costs.

There are many government-sponsored programs and services to help children with special needs and their families. Each program has its own eligibility requirements and more information can be obtained from the nearest regional office of the Ministry of Children and Youth Services. In addition, the Ministry of Community and Social Services in Canada funds many services to children and youth. In the United States, the Administration for Children and Families oversees Federal programs that promote the economic and social well-being of families and children. Major

programs include the Administration on Developmental Disabilities and the Family and Youth Services Bureau. Disability.gov is the federal US government website that provides information on community disability programs and services. Worldwide, almost all government organizations have created programs at all levels of administration (e.g., countrywide to local offices) to assist families of children with special needs. Most services are delivered through community-based agencies. The services include: respite care (in-home and outside the home); specialized community supports to help children with developmental disabilities live in their communities; and, residential services. Included in the available programs is a Behavioural Management Program.

FURTHER RESOURCES

The Tourette Syndrome Associations websites, Canada: www.tourette.ca; USA: www.tsa-usa.org; UK: <http://www.tourettes-action.org.uk>; Europe: <http://tourette-eu.org/en/european-society-for-the-study-of-tourette-syndrome.html>). Similarly Life's a twitch! website: www.lifesatwitch.com/

NOTES



Dr. Paul Sandor has been involved in treating patients with Tourette syndrome and conducting research regarding this condition and other neurodevelopmental disorders for the past 30 years. He is a Professor of Psychiatry and Wolf Family Chair in Neurodevelopmental Psychiatry at the University of Toronto, University Health Network and Youthdale Treatment Centers in Toronto, Ontario, Canada. His research included genetics, imaging, neurophysiology and clinical pharmacology. He has over 60 peer-reviewed publications. In his capacity as the Director of Tourette Syndrome Neurodevelopmental Clinic at the Toronto Western Hospital, University Health Network, Toronto, Ontario, Canada he has assessed and managed several thousand patients with Tourette syndrome. Dr. Sandor is the Chairperson of the Professional Advisory Board, Tourette Syndrome Foundation of Canada.



Dr. Mary May Robertson is an Emeritus Professor at University College London. She has co-authored 4 books, edited 2 books and has over 350 medical/scientific publications. Her many awards include a DSc (Med) in Neuropsychiatry entitled "What makes us Tic?" (2006). It is a rare degree and she was the 9th person to receive it at the University. Her biography was entered into the American International Publication Marquis WHOSE WHO in 2010, in 2011 she was awarded the International Neuropsychiatric Association's W. Alwyn Lishman lifetime achievement award, in 2012 she was the first recipient of the Professor Mary M. Robertson ESSTS (European Society for the Study of Tourette Syndrome) Lifetime Achievement Award; in 2012 she was also invited to be a patron of Tourette's Action in UK. She has also been awarded the University of Cape Town Gold medal for philanthropy.



Stuart Ellis-Myers aka "Twitchy" is a dramatic model of self-inspiration and determination. When he speaks of improving lives, he speaks from personal experience. Since the age of nine Stuart has "suffered" from Tourette Syndrome. Despite this disability, Stuart has become an internationally known and respected keynote speaker on communication leadership, sales and how to overcome any adversity. He might even look familiar as he has appeared in twelve television commercials.



Dr. Sharon Chung is the Research Manager and a Staff Scientist at the Sleep Research Unit at the Toronto Western Hospital, University Health Network and a Clinical Researcher at the Youthdale Child and Adolescent Treatment Centres. Her other duties include supervising students and fellows and organizing professional educational events. She has published a number of research papers and is actively involved in sleep and child and adolescent research. Her sleep research interests include sleep education, sleep apnea, shift work, and patient compliance with treatment. She has previously participated in a number of educational events providing shift work coping strategies to workers at various locations. Her current child and adolescent research involves an assessment of the clinical record of young offenders and investigating gender and psychopathological effects on adolescent harassment and bullying.



Dr. Colin Shapiro has co-authored over 25 books and booklets and over 350 medical and science publications. He studied medicine in Johannesburg, South Africa, and subsequently did his PhD and Psychiatry training in Edinburgh, Scotland. He was appointed as a full professor in the department of psychiatry at the University of Toronto at the young age of 37 and was subsequently cross-appointed to the department of ophthalmology. He co-founded the British Sleep Society and later the International Neuropsychiatry Association. He has a strong interest in educating both family physicians and patients and has recently set up a program to educate school children about sleep and sleep disorders.

ISBN 978-0-9683576-8-2



9 780968 357682