Tourette Syndrome

Want to know more?

WHAT MAKES US TIC?

Tourettes action

www.tourettes-action.org.uk

This publication is intended as a general introduction for those recently diagnosed with Tourette Syndrome, their families and colleagues, and for those who wish to know more about the condition.

Diagnosis may be a stressful time and questions about the condition may be detailed and specific. For further information and support, please contact the Tourettes Action free Helpdesk on 0300 777 8427 or
email help@tourettes-action.org.uk
Alternatively go to www.tourettes-action.org.uk

Helpline open 9am - 5pm.
Monday - Friday.
Call charges apply.

www.tourettes-action.org.uk

What is Tourette Syndrome?

Tourette Syndrome is an inherited neurological condition. It affects one schoolchild in every hundred and more than 300,000 children and adults in the UK. The key feature is tics – involuntary and uncontrollable sounds and movements.

It is a complex condition and covers an extraordinarily wide spectrum. People may have a very mild form of Tourette Syndrome (TS). They and those close to them may not even be aware that they have TS. At the other end of the scale, medical symptoms are extreme and the social, educational and economic effects are serious. These are the examples that generate media interest.

Symptoms

The key features are tics, repeated movements and sounds. It is important to understand that these are chronic (long-term) and involuntary. Someone with Tourette Syndrome may be able to suppress them for a period but eventually they have to let the tics out.

Tourette Syndrome is a neurological condition of unknown origin. It is a complex condition and covers an extraordinarily wide spectrum. People may have a very mild form of TS. They and those close to them may not even be aware that they have TS.

Tics usually start in childhood around the age of seven, and are usually worst between 10-12 years. However, in approximately half of people with TS, most symptoms disappear by the age of 18. TS is a persistent disorder but not always greatly disabling.

The first tics often start around the head and face, like blinking and/or grimacing. Vocal tics tend to appear later, around age 11. The different tics can be simple, such as blinking, or complex, like touching or jumping. Examples of vocal tics besides uttering words or making sounds are throat clearing, sniffing and/or coughing.

Although the word 'involuntary' is generally used to describe

the unwanted movements and sounds, most people with TS can suppress their tics for a short time. This could be compared to the experience of sneezing, or trying to suppress a sneeze. Often there is a premonitory feeling or urge which is relieved by the action of ticcing. This sensation has been described as like 'itchy blood', or 'insects crawling under the skin'.

Over 85 percent of people with TS have more than just tics. Additional conditions (comorbidities) include obsessive compulsive disorder (OCD) and/or attention deficit hyperactivity disorder (ADHD). Children and adults may also suffer from 'rages'. Comorbidities often present more problems than the tics and can be less visible.

Coprolalia – involuntary swearing

The well-known feature of coprolalia or involuntary swearing is present in only about 10 percent of cases. Some people with TS also made rude gestures – copropraxia.

More recently a phenomenon known as Non-Obscene Socially Inappropriate (NOSI) behaviour has been identified where people with TS may have a compulsion to do or say the wrong thing, which may be socially unacceptable; for example, touching something they should not touch or making an insulting comment.

Vocal tics can take form of random words or phrases or repeating what you or someone else has just said echo/palli – lalia.

Living with Tourette Syndrome

There have been many studies on Tourette Syndrome and, taken together, they indicate that one schoolchild in 100 has Tourette Syndrome. Boys are affected two to four times more commonly than girls. Tourette Syndrome is significantly more common in special educational needs (SEN) environments.

Using available figures, the number of people with diagnosable Tourette Syndrome in the UK is thought to be between 200,000 and 330,000. The majority of them probably have a mild disorder of little medical significance, especially in adulthood.

TS is probably less common in adults than in children, and for most adults symptoms become less severe. On average, symptoms peak at age 11 (when many children face the

additional disruption of moving school), although there is great variation. Towards the end of adolescence, about half of teenagers with TS find that their symptoms have effectively disappeared or that they are no longer significant.

Some people with TS have a severe disorder persisting into adulthood. Although the tics may improve, in some instances co-morbidities may persist or even get worse. However,we do not have good tools to predict whether an individual child with TS will become an adult with TS.

People with TS have the same range of IQ as the rest of the population, with some subtle differences. There is a tendency towards lower verbal IQ (intelligence that depends on verbal reasoning using language) in relation to performance IQ (puzzle solving, mathematics, for example).

TS is very variable in its severity and co-morbidities. Some people with TS have symptoms that are only mildly troubling. Others have symptoms that are intensely disabling:

- physically (tics can cause damage to joints or self-injury, eg hitting oneself)
- educationally (TS can make it impossible to follow what is going on in class)
- economically (TS can be a barrier to employment)
- socially (TS can lead to ridicule, bullying and social exclusion)

Other common features

These include depression and disturbed sleep. Medical treatment with drugs can be helpful for these features, but the effects are variable.

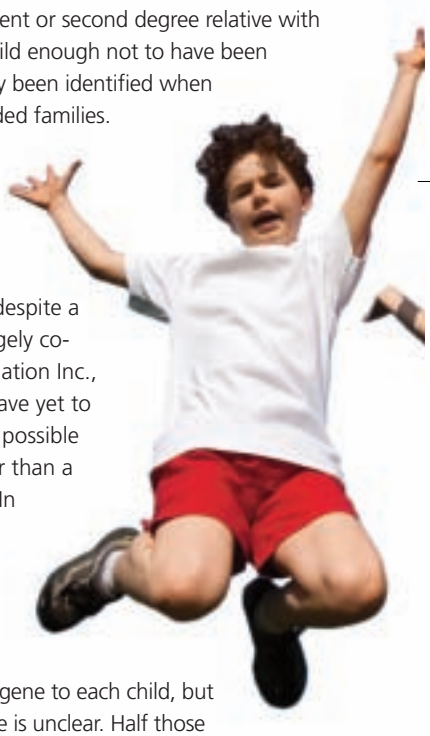
About 12 percent of patients seen in clinics have a syndrome consisting of tics alone. The rest have additional conditions ('co-morbidities'), most commonly obsessive compulsive disorder (OCD) and/or attention deficit hyperactivity disorder (ADHD). Children and adults may suffer from 'rages' with little provocation and usually followed by remorse. Co-morbidities often present more practical problems than the tics, and can be less visible.

Causes of Tourette Syndrome

Some doctors still think of Tourette Syndrome as a psychological condition. In fact and without doubt, Tourette Syndrome is a neurological disorder, due to a disturbance of underlying brain function.

From the 1980s it became increasingly apparent that TS is mainly a genetic (inherited) condition.

There were documented cases of large families where many members had TS. Consultants became aware that most of the cases of TS they saw in clinics were hereditary. Such cases are not always easy to identify. Some very mild features, such as a parent with mild obsessive tendencies, may be related to the same genetic cause. In most cases there is a more compelling family history than this, such as a parent or second degree relative with tics. Sometimes, the tics are mild enough not to have been previously noted and have only been identified when researchers interviewed extended families. So there is considerable evidence that TS is a genetic disorder with a wide range of severity.



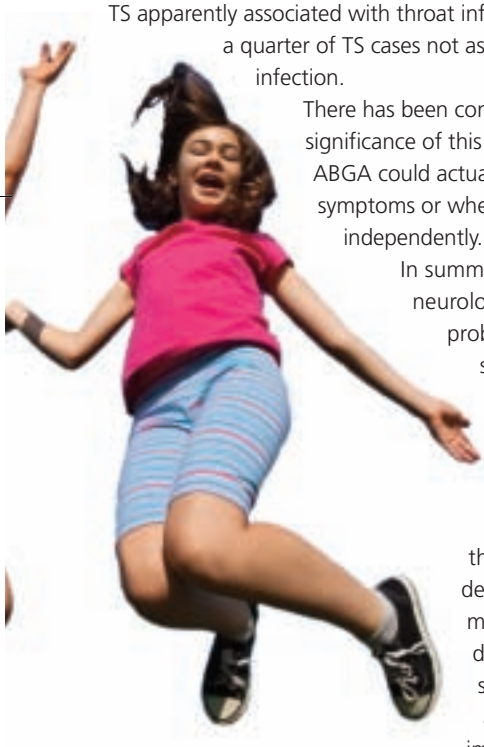
Unlike many other genetic neurological conditions, and despite a committed research effort largely co-ordinated by the US TS Association Inc., the major genes causing TS have yet to be identified. There are many possible reasons. Multiple genes rather than a single gene may be involved. In addition, there may be more than one kind of TS, each caused by different genetic factors.

Parents with TS have a 50 percent chance of passing the gene to each child, but the exact pattern of inheritance is unclear. Half those

children with the gene will show signs of TS, and many of them will have only mild features.

Group A streptococcal throat infections can cause an immunological reaction resulting in rheumatic fever in some children. This is now very rare in the UK but occurs in certain non-Western countries, such as Brazil. Rheumatic fever is also associated with neurological reactions causing the movement disorder Sydenham's chorea (St. Vitus's dance). Unlike TS, it is more common in girls. It has been suggested that in some cases, where a streptococcus infection has triggered an explosive onset of TS, the cause may be a similar neurological reaction.

The theory is that the streptococcal infection produces antibodies against the basal ganglia, which is a part of the brain responsible for controlling movement. Anti basal ganglia antibodies (ABGA) are present in Sydenham's chorea, in cases of TS apparently associated with throat infection and also in about a quarter of TS cases not associated with known infection.



There has been controversy over the significance of this finding and whether ABGA could actually cause TS-like symptoms or whether they occur independently.

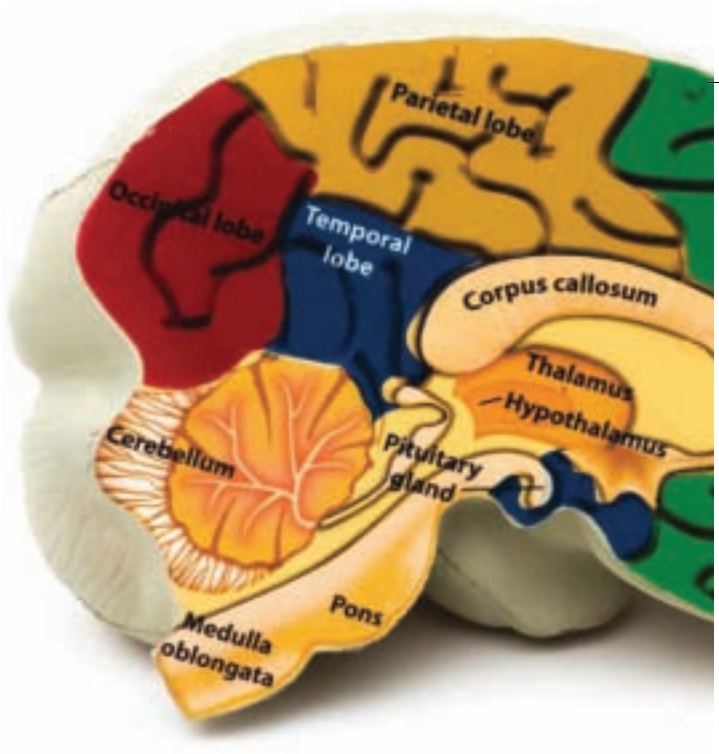
In summary, TS is a genetic neurological condition, most probably with a complex set of controlling genes. The ultimate goal of TS research would be to identify important genes, define what effects they have on the developing brain and to match them up with the different types or symptoms of TS. There are other potentially important environmental factors, for example immunological, which could interact with genes in a complex manner.

Neurosurgery

Neurosurgical treatments for Parkinson's disease (PD) and some other movement disorders are now well established. From the 1990s, small areas of the basal ganglia were lesioned (cut) using a technique called stereotactic surgery. A recent development in treatment for PD is deep brain stimulation (DBS). DBS involves implants which deliver high frequency electrical pulses to specific areas of the brain.

The model can be criticised in a number of ways. The theory that the treatment works by suppressing the area of brain to which it is applied may be simplistic. However, clinical trials show conclusively that these approaches can work in reducing the symptoms of PD.

TS is very different. We do not have a compelling model of what parts of the brain may be causing the symptoms because, unlike PD, there are no discernible changes in brain structure. However, like PD, TS is also a disorder of movement in which the

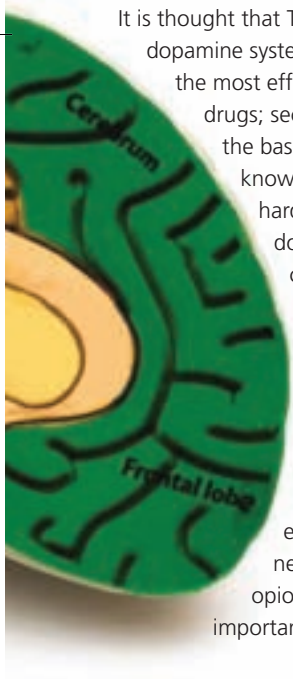


basal ganglia are implicated. There has been great interest in evaluating the effect of DBS to this part of the brain. Internationally, fewer than 50 DBS operations have been performed to date, the vast majority in adults. The largest study published so far consists of 18 individuals. Guidelines for how patients should be selected have also been published.

So the experience to date is encouraging but still experimental. No operations done in the UK have been documented to date. DBS could in future become an important treatment for people with severe TS who is not responsive to any other treatment, but this will only be established with further well documented experience.

Dopamine & other neurotransmitters

Dopamine is a chemical, related to adrenaline, which is secreted by the brain cells (neurons) to communicate with other neurons. Dopamine affects the basal ganglia system, which plays a major role in controlling movement. It is also involved in the limbic (emotional) part of the brain. Dopamine is the neurochemical that is deficient in Parkinson's disease (PD).



It is thought that TS may involve a dysfunction of the dopamine system. There are two reasons for this: firstly, the most effective drugs for tics are dopamine-blocking drugs; secondly, because TS is a movement disorder, the basal ganglia are implicated. This is sometimes known as the 'dopamine hypothesis'. It has been hard to assemble convincing evidence that the dopamine system plays a major role in causing symptoms rather than merely suppressing them. There has been little neuropathological (post-mortem) data to prove this. Some brain scanning studies have supported this hypothesis, whilst others have not. It is certainly possible that dopamine is important, despite negative studies in some cases. It is also eminently possible that other neurotransmitters, such as serotonin and opioids, are involved or are even more important.

Research and treatment

Research into medical treatment of Tourette Syndrome is exceedingly difficult for three main reasons. Firstly, the severity of symptoms is very hard to measure objectively. Rating scales exist but they all have limitations. Secondly, the severity of Tourette Syndrome fluctuates naturally without treatment. Thirdly, individuals have hugely varied responses to medicines.

Drug treatment

There are numerous reports of different drugs being used successfully in a small number of TS patients. In other areas of medicine, this evidence would not be considered strong enough to judge the effectiveness of a treatment.

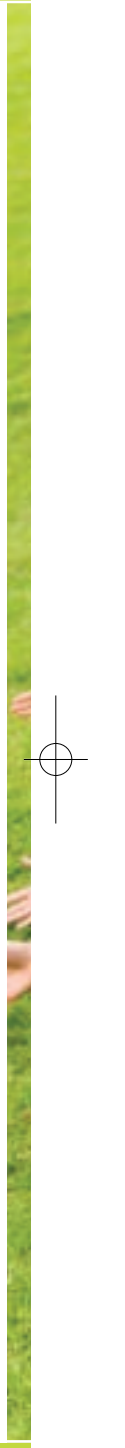
Currently, drugs are prescribed to relieve symptoms; to treat tics, for ADHD and for OCD. There are standard medical treatments for ADHD and OCD which are also used in TS. Often, ADHD and OCD are more important targets for treatment than the tics as they are often more debilitating than the tics. Treatments for ADHD was thought to make tics worse and were traditionally not recommended but recent trials suggest this is incorrect.

The standard treatment for tics is neuroleptics, dopamine receptor blocking drugs. Their main use is to treat psychosis but there is no clinical link between TS and psychosis. Individual patients can respond to different TS drugs in a variety of ways and it is not known why.

Non-drug treatment

Drug treatment is not the only option. Psychological cognitive-behavioural approaches have been shown to be as effective as drugs to treat TS. This is standard treatment for OCD, that can also be applied to tics using a package of measures known as Habit Reversal Therapy. Although there is only a very patchy service in the UK, Tourettes Action is working towards increasing access to it.

There is little evidence for other non-drug treatments. Patients and parents are often interested in the influence of diet, for which we have little reliable data so far.



Glossary and further information

Anxiety – Can show as sleep difficulties, tension habits, motor unrest, phobias, worries, poor concentration, or panic attacks.

ADHD – Attention Deficit Hyperactivity Disorder – Condition characterised by an impaired ability to regulate activity level (hyperactivity), attend to tasks (inattention), and inhibit behaviour (impulsivity). For a diagnosis of ADHD, the behaviours must appear before an individual reaches age seven, continue for at least six months, be more frequent than in other children of the same age, and cause impairment in at least two areas of life (school, home, work, or social function). Adults too may show signs of ADHD such as overly impulsive behaviour and concentration difficulties.

Basal Ganglia – Several large clusters of nerve cells, including the striatum and the substantia nigra, deep in the brain below the cerebral hemispheres; responsible for motor movements.

CBT – Cognitive Behavioural Therapy – Psychological treatment for mental health conditions. Treatment usually takes between eight and 20 sessions. A combination of cognitive and behavioural therapies. CBT is based on the assumption that most unwanted thinking patterns and emotional and behavioural reactions are learned over a long period of time. The aim is to identify the thinking causing the unwanted feelings and behaviours and to learn to replace this thinking with more positive thoughts. The therapist does not focus on past events (such as childhood) but on current difficulties. The goal is to teach new skills and ways of reacting.

Comorbidity – Presence of more than one disease or health condition in an individual at a given time. OCD and ADHD are often comorbid with TS.

Copropraxia and coprolalia – Copropraxia: making obscene or otherwise unacceptable movements or gestures. Coprolalia: using obscene or unacceptable language. This may involve swearing or racist remarks. Coprolalia can cause serious problems at school, in society and at work, and it is particularly sad that the words uttered usually bear no relation to the true feelings of the person saying them.

DBS Deep Brain Stimulation – Electrodes are implanted in the brain and stimulated by a surgically implanted pulse generator in the upper chest. Several studies have shown that this surgical intervention may aid in the amelioration of involuntary movements in patients with Parkinson's Disease and Essential Tremor. More recent studies have shown promise for other disorders including Dystonia (a movement disorder which causes involuntary contractions of the muscles, resulting in twisting and repetitive movements and can be very painful). Early experience with DBS for tics in TS has been mixed. While some individuals have experienced a reduction in symptoms, others have not. There is no long-term follow-up yet to indicate whether symptoms will return. There might be serious risks involved, including cerebral bleeding and infection.

Depression – In TS depression is most commonly seen in people with severe tics, sleep disturbances or OCD. Clinical depression is a common psychiatric disorder, characterised by a persistent lowering of mood, loss of interest in usual activities and diminished ability to experience pleasure. Depression should always be taken seriously. It is treatable and medical advice should be sought.

Dopamine – A neurotransmitter (naturally produced chemicals by which nerve cells communicate) that controls movement and balance. Essential to the proper functioning of the central nervous system (CNS). Dopamine assists in the effective transmission of electrochemical signals from one nerve cell (neuron) to another.

Dopamine antagonist – Binds to and blocks the action of dopamine receptors, essentially hindering receptor activity by preventing stimulation by dopamine. Antagonists can prevent or reverse the actions of dopamine by keeping dopamine from attaching to receptors.

Double-blind placebo-controlled (trial) – The gold standard for clinical trials. The placebo (inactive substance) is given to one group of participants, while the treatment being tested is given to another group. Neither patients nor those administering know which group receives the placebo, so their expectations cannot influence the outcome.

Echophenomena – Echolalia: repeating other people's words. Echopraxia: repeating other people's gestures. Common in TS.

Full Blown – Although this term implies that the disease or disorder cannot be developed any further, it is used to describe the middle level of severity of TS. This can be misleading, as the severity of TS can increase to become 'Tourette's Plus'.

Genetics – Field of science that looks at how traits are passed down from one generation to another, through the genes.

Limbic – Related to the part of the brain involved in emotion, motivation, and emotional association with memory.

Neuroleptic drug – An antipsychotic drug that may produce a state of apathy, lack of initiative and limited range of emotion. In psychotic patients, neuroleptic drugs cause a reduction in confusion and agitation and tend to normalise psychomotor activity.

Neurology – Diagnosis and treatment of diseases and disorders of the nervous system.

Neurosurgery – Surgical treatment of diseases and disorders of the brain and nervous system.

Neurotransmitters – Chemicals that are used to relay, amplify and modulate signals between a neuron and another cell.

NOSI – Non-Obscene Socially Inappropriate behaviour – Falls short of swearing, but involves saying or doing things that are socially unacceptable; for example, personal remarks about height, weight or facial features.

OCD Obsessive Compulsive Disorder – Anxiety disorder characterised by repeated intrusive thoughts and associated ritualised behaviours intended to alleviate that anxiety. Compulsions typically include checking, ordering, counting, repeating, getting things 'just right' or symmetrical, or forced touching which is a different spectrum from the symptoms of 'pure' OCD. Examples include touching an object with one hand after touching it with the other hand to 'even things up' or repeatedly flicking the light switch on and off. In more serious cases, the obsession may have sexual, violent, religious or aggressive themes.

Paliphenomena – Similar to echophenomena but involves the person with TS repeating their own words and actions eg "Hello, I came here by bus bus bus bus".

PANDAs Paediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections – Disorders affecting some children with OCD and/or tic disorders such as TS. Their symptoms worsen following strep infections such as strep throat or scarlet fever. The children can have dramatic onset of symptoms, including motor or vocal tics, obsessions, and/or compulsions. Children may also become moody, irritable or show concerns about separating from parents or loved ones.

Parkinson's Disease – A chronic and progressive neurological disorder affecting control over movement, balance, coordination and speech.

Perinatal – During and immediately after childbirth.

Placebo – Inactive substance or treatment that looks the same as, and is given the same way as, an active drug or treatment being tested. The effects of the active drug or treatment are compared to the effects of the placebo.

Prevalence – Proportion of cases in the population; calculated by dividing the total number of cases in the population by the number of individuals in the population, eg one percent of schoolchildren are affected by TS.

Prognosis – Likely or expected development of a disease or the chances of getting better.

Psychology – The scientific study of human and animal behaviour.

Psychopathology – Refers to either the study of mental illness or mental distress or the manifestation of behaviours and experiences which may be indicative of mental illness or psychological impairment. Examples of psychopathology sometimes found in people with TS include rage attacks/aggression, oppositional defiant disorder and inappropriate sexual behaviour.

Psychopharmacology – Study of the effects of drugs on mood, sensation, consciousness, or other psychological or behavioural functions.

Psychosis – A mental state often described as involving a 'loss of contact with reality'.

Pure Tourette Syndrome – A proposed sub-type of TS, defining TS with no comorbid features.

Rage attacks – Frightening and destructive violent outburst, often without provocation or disproportionate to the trigger. Once begun, a rage attack has to be left to run its course. Rage may be linked to tic suppression.

Ratings Scales – Designed to assess the severity of tics. There are a number, all with limitations. The most well known include The Yale Global Tic Severity Scale (YGTSS; Leckman et al., 1989) and the Motor tic, Obsessions and compulsions, Vocal tic, Evaluation Survey (MOVES; Gaffney, Sieg, & Hellings, 1994). There are many others.

RCT Randomised Control Trial – One of the most commonly reported methods for evaluating the effectiveness of treatments. People are allocated at random to receive one of several clinical interventions. One of these interventions acts as a comparison to provide a benchmark. Randomised controlled trials are the most rigorous way of determining whether a cause-effect relation exists between treatment and outcome and for assessing the cost effectiveness of a treatment.

Selective Serotonin Reuptake Inhibitors (SSRIs) – A drug that blocks the removal of serotonin from the synapse; thereby prolonging and increasing the effects of serotonin.

SIB – Self-injurious behaviour – It includes punching and slapping the head, face or body, or scratching or sticking sharp objects into the body, including the eyes. It can be an obsessional behaviour.

Sleep Disorders – A group of syndromes characterised by disturbance in the patient's amount of sleep, quality or timing of sleep, or behaviours or physiological conditions associated with sleep. Frequent awakenings, sleep talking or walking are fairly common among people with TS.

Stimulant Medication – Drugs that increase the release or block the reabsorption of dopamine and norepinephrine, two brain neurotransmitters. In adults, they have the effect of making people more alert, active and awake. In children, they can increase attention and reduce hyperactivity, and are used as one part of the treatment for hyperkinetic disorder and ADHD.

Tourette Syndrome Plus – Proposed sub-type of TS, which includes TS with ADHD, OCB or OCD, and/or Self-Injurious Behaviours. Includes TS patients with depression, anxiety, personality disorders, Oppositional Defiant Disorder, Conduct Disorder and any other learning problems.

Waxing and waning – Commonly used to describe the fluctuations in tic severity with TS. Tics can worsen (wax) and lessen (wane) with a frustrating lack of predictability.

Further reading

Busy Body: My Life with Tourette's Syndrome Nick van Bloss

Coping with Tourette Syndrome: A Workbook for Kids With Tic Disorders Sandra Buffolano

Front of the Class: How Tourette Syndrome Made Me the Teacher I Never Had Brad Cohen, Lisa Wysocky

Hi, I'm Adam: A Child's Book about Tourette Syndrome Adam Buehrens

I Can't Stop: A story about Tourette Syndrome Holly L. Niner

Kids in the Syndrome Mix of ADHD, LD, Asperger's, Tourette's, Bipolar and More!: The One Stop Guide for Parents, Teachers and Other Professionals Martin L., M.D. Kutscher, Robert R. Wolff

Life, Interrupted. The memoir of a nearly Person James McConnel

Making Allowances: Autobiographical Accounts of People with Tourette Syndrome Chris Mansley

Managing Tourette Syndrome: A Behavioural Intervention for Children and Adults Therapist Guide (Treatments That Work) Douglas W. Woods, John Piacentini, Susanna Chang, Thilo Deckersbach, Golda Ginsburg, Alan Peterson, Lawrence D Scahill, John T Walkup, Sabine Wilhelm

Quit It Marcia Byalick

The Good Schools Guide to Special Educational Needs 2006 Sandra Hutchinson

Tics and Tourette Syndrome: A Handbook for Parents and Professionals Isobel Heyman and Uttom Chowdhury.

Tictionary: A Reference Guide to the World of Tourette Syndrome, AS, AD/HD and Obsessive Compulsive Disorder for Parents and Professionals Becky Ottinger.

Tic Talk: Living with Tourette Syndrome Dylan Peters.

Tourette Syndrome: A Practical Guide for Teachers, Parents and Carers Amber Carroll and Mary Robertson

Tourette Syndrome – The Facts (Second Edition) Mary Robertson and Andrea Cavanna.

Why do you do that? A Book about Tourette Syndrome for Children and Young People Uttom Chowdhury and Mary Robertson.

Tourettes Action is the UK's leading support and research charity for people with Tourette Syndrome (TS) and their families.

We want people with TS to receive the practical support and social acceptance they need to help them live their lives to the full. We deal with all forms of TS including the most challenging.

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