Tics and Tourette syndrome

a guide to management in childhood

Most children with tics or Tourette syndrome need no treatment and their symptoms subside by late adolescence. Medical therapy should only be used if absolutely necessary or if there is complicated Tourette syndrome.

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Tics are the most common movement disorder of children and as many as 10 to 24% of boys will have a tic at some time in their life. Tourette syndrome is a neuropsychiatric disorder characterised by multiple motor tics and one or more vocal tics.

IN SUMMARY

- **Tics are common in children and particularly involve facial muscles.**
- **Tics are temporarily suppressible but this produces a build up of tension and an increasing urge to perform the tic.**
- **Tourette syndrome is defined by the presence of multiple motor and one or more vocal tics, which typically wax and wane in severity.**
- **The diagnosis of Tourette syndrome can usually be made on the clinical features alone and there is no need for investigation.**
- **Most children with tics or Tourette syndrome need no treatment, and in most the symptoms subside by late adolescence.**
- **There is a subgroup of children with Tourette syndrome who have associated disorders and who require psychiatric management.**

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Historical features

There is a long list of historical figures who may have had tics or Tourette syndrome, including Claudius, Napoleon Bonaparte, Molière, Peter the Great and Samuel Johnson. More recent Tourette syndrome sufferers include Kurt Cobain from the pop group Nirvana, and Paul Gascoigne, the colourful English soccer player.

Discussion of Tourette syndrome is incomplete without mention of the life of the man who first recognised the syndrome and after whom it is named, Georges Albert Edouard Brutus Gilles de la Tourette. Tourette was born in 1857 in a village near Loudun, a town famous because of events in 1634 when Father Grandier was accused of demonically possessing a whole convent of nuns, including the Mother Superior. Charged with inciting them to utter obscenities and engage in lewd and wanton behaviour, Grandier was found guilty and burnt alive.

Tourette was a brilliant student but restless and difficult. He became the pupil of the great French neurologist, Jean-Martin Charcot. Although admired by Charcot, he made many enemies and was described by the 19th century French author Daudet as 'neither good nor bad, neither studious

Figure. The English author Samuel Johnson may have suffered from a form of Tourette syndrome, based on contemporary descriptions of his facial tics and strange vocalisations. Dr Johnson (1709–1784) at Cave's the Publisher, 1854 (oil on canvas), by Henry Wallis (1830–1916).

nor lazy, neither intelligent nor foolish and he vacillated with his confused and malicious mind between a multitude of faults without lingering'. In 1893, while sitting in his office, Tourette was shot three times by a young woman who claimed she was under his hypnotic influence. One bullet hit him in the occipital region but he recovered. Subsequently, his behaviour became increasingly disturbed and he died in 1904 in an asylum, not from sequelae of his head wound but from general paresis of the insane.

In 1885, Tourette reported nine patients with his 'syndrome'. In contrast to many original descriptions of syndromes, most of the features he recognised are still considered typical. Tourette regarded coprolalia (the involuntary muttering of obscenities or profanities) as pathognomonic. It occurred in five of his cases. He believed the condition was incurable and the cause hereditary. Although the life span was not affected, it was 'a deplorable companion throughout life'.

Tourette's first case, the Marquise de Dampierre, had already been described by the French physician Itard 60 years earlier and by other authors in the intervening years. A woman of 'distinguished manners and background', in the middle of an interesting conversation, she would suddenly emit 'horrible screams' and 'words even more extraordinary than her screams' in particular 'merde' and 'foutu cochon'. An obsessional component was noted: 'The more revolting these explosions are, the more tormented she becomes by the fear she will say them again; and this obsession forces these words into her mind and to the tip of her tongue so that she can no longer control them'.

What are tics?

While tics are generally easy to recognise, they are difficult to define. Typically they start in the facial region with movements such as blinking, nose twitching or facial grimacing. Although they are classified among the involuntary movement disorders, there is usually a volitional component. This has resulted in convoluted attempts at definition, including terms such as 'intentional involuntary actions', 'semivoluntary or involuntary movements' and 'involuntary but suppressible movements'. A premonitory sensation, usually a sense of discomfort or tightness, is present in the affected body part in both motor and vocal tics in up to 80% of people with tics. This can be suppressed, but it causes a build up of tension and an increasing urge to perform the tic. Once the tic has occurred, there is temporary relief but the discomfort and urge then return. The premonitory sensation prior to the tic is of particular interest as it may be the 'involuntary' component. Children as young as 6 or 7 years can describe the premonitory feelings, usually in terms of the affected body part being 'sore'.

Children with tics are very suggestible and simply mentioning a particular tic to a child may be enough to immediately provoke it. Tics tend to be decreased by distraction or intense concentration. Commonly they increase during relaxation after a period of stress. They may be particularly florid while the child is watching television in the evening after a hard day at school. In Tourette syndrome, the tics wax and wane and there are periods of remission. They frequently change character – one tic is dominant for a time, then subsides and another takes over. Unlike most other movement disorders, tics may persist in sleep.

Tics can be subdivided, although somewhat arbitrarily, into motor and vocal (or phonic) tics and simple and complex tics:

- simple motor tics – abrupt jerky movements of small groups of muscles, for example, blinking and facial twitches
- complex motor tics – involve multiple muscle groups, are more co-ordinated and may appear

Table 1. Children with tics seen over a 10-year period

Characteristics of the children with tics seen by the author during 1989 to 1999

- number of children = 153
- 84% boys; 16% girls
- age range, 4 to 16 years
- 86% were 12 years or younger

The children fell into two broad groups:

Group 1

- 5% simple tics
- 20% multiple tics
- 35% uncomplicated Tourette syndrome
- 3% moderate-severe Tourette syndrome

Group 2

- 30% Tourette syndrome and psychiatric comorbidity
- 2% single tics and psychiatric comorbidity
- 5% multiple tics and psychiatric comorbidity

voluntary; examples are touching, hitting or jumping

- simple vocal tics – simple inarticulate sounds such as throat clearing, sniffing or coughing
- complex vocal tics – usually consist of words, for example echolalia (the involuntary repeating of words heard) or coprolalia. Although swearing is the most widely known symptom of Tourette syndrome, it is relatively rare in childhood.

Table 1 summarises the characteristics of children with tics or Tourette syndrome seen by the author over a 10-year period (1989 to 1999) in a teaching hospital. The striking male predominance is obvious (84% boys *v.* 16% girls). The children formed two broad groups: approximately 60% of the children had tics or uncomplicated Tourette syndrome (that is, the tics were not disabling and the child was otherwise well), and the remainder had tics that were overshadowed by other problems such as attention deficit hyperactivity disorder, obsessive symptoms, conduct disorder, anxiety, depression or major family and social problems. Most of the children in the second group were referred for formal psychiatric management of these other problems.

There is a large amount of recent neuroimaging and neurophysiological data about tics suggesting they result from the disturbance of circuits between the basal ganglia and frontal lobes, but discussion of this is beyond the scope of this article.

How common is Tourette syndrome?

Tourette syndrome was once regarded as a rare and exotic condition but it is now recognised that mild forms are common. To some extent this is the result of a liberalisation of the diagnostic criteria. Recent studies have suggested a prevalence of Tourette syndrome of between 0.05% and 2.9% of the population.

Diagnosis of the syndrome

The essential criteria for the diagnosis of Tourette syndrome are the presence of multiple motor tics and one or more vocal tics for more than a year with other neurological conditions excluded. Formal attempts at definition have varied as different criteria have been used. For example, the various revisions and editions of the American Psychiatric Association's 'Diagnostic and statistical manual of mental disorders' (DSM-III, DSM-IIIR and DSM-IV) all have slightly different criteria. Table 2 shows the criteria used in the classification system suggested by the US Tourette Syndrome Classification Study Group.

Differential diagnosis

In most children with tics or Tourette syndrome, the diagnosis can be made on the clinical features and there is no need to order investigations. Rarely there may be problems differentiating Sydenham's chorea or multifocal myoclonus from simple tics. The phenomenology of complex tics may be identical to that of compulsions or stereotypic movements but usually associated features such as the presence of simple tics or obsessional thoughts help in the differentiation. Tics can occur as sequelae of encephalitis. (They were particularly associated with the pandemic of Von Economo's disease in the years 1916 to 1927.) Psychoactive substance abuse should be considered in the older child or teenager who suddenly develops tics. Diseases involving the basal ganglia, such as Huntington's disease or neuroacanthocytosis, may produce tics as well as other movement disorders.

Table 2. Criteria used in defining Tourette syndrome

The Tourette Syndrome Classification Study Group's criteria for classifying tics as part of Tourette syndrome:

- Both multiple motor and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently
- The tics occur many times a day, nearly every day, or intermittently throughout a period of more than a year
- The anatomical location, number, frequency, complexity, type and severity of tics change over time
- The onset is before the age of 21
- Involuntary movements and noises cannot be explained by other medical conditions
- Motor and/or vocal tics must be witnessed by a reliable examiner directly at some point in the illness or be recorded by videotape or cinematography

Associated disorders

Attention deficit hyperactivity disorder, obsessive compulsive disorder and self-injurious behaviour are commonly associated with Tourette syndrome. Other problems that may occur include anxiety, depression, conduct disorder, rage attacks, learning difficulties, personality disorders and nonobscene socially inappropriate behaviours (such as making hurtful or derogatory comments about another person's race, height or weight).

What causes Tourette syndrome?

Genetics

Tourette syndrome seems to be a genetic disorder, but its exact mode of inheritance is at present uncertain. Linkage studies involving 80% of the human genome have failed to find the 'Tourette syndrome gene'. It may be inherited as an autosomal dominant condition with sex specific penetrance, but in some families inheritance appears to be bilineal (that is, from both the mother and the father). Polygenic inheritance has also been suggested. Twin studies have shown 50 to 90% concordance with monozygotic twins; in those cases where there is discordance, the lower birthweight twin is most often affected. As with many conditions, Tourette syndrome may result from a genetic predisposition interacting with environmental factors.

PANDAS

In a parallel to Sydenham's chorea it has been suggested that in some children, obsessive compulsive disorder or tics may be precipitated by streptococcal infection. This theory has resulted in the use of aggressive treatments including plasmapharesis and intravenous gamma globulin infusions. The status of this proposed syndrome – paediatric autoimmune neuropsychiatric disorder associated with streptoccocal infection (PANDAS) – is at present uncertain. A major problem with the hypothesis is the frequency of both streptococcal infections and asymptomatic carriage of streptococcus in childhood. Around 20% of normal children have a positive throat swab for streptococcus at any given time. There is also no evidence of an association between obsessive compulsive disorder and valvular heart disease, which would be expected if there was a true parallel with Sydenham's chorea. At this time, in the absence of more compelling evidence, it does not seem reasonable to use aggressive immunomodulatory therapies to treat either Tourette syndrome or tics.

Course of the disorder and its prognosis

It seems that in around 60 to 70% of cases of Tourette syndrome, the tics cease or are substantially reduced by late adolescence. A recent review of 42 children with Tourette syndrome found that the disorder was at its most severe at around the age of 10 years. By 18 years, nearly half of the children were almost free of tics and only 10% continued to have tics that were of moderate to marked severity.

Treatment

Tics are mainly a social disability and themselves rarely harm the child. Usually, it is the reaction of other people to the child with tics that is the problem. There may be teasing, ridicule and ostracism, with consequent poor self-esteem. Certain tics are more troublesome; for example, persistent throat clearing or sniffing can be extremely irritating to others, whereas facial blinking or grimacing may simply be regarded as a curiosity. Loud swearing is a problem in any social setting away from the home.

Many children whose movements would have been diagnosed 30 years ago as 'just a habit' now carry a diagnosis of Tourette syndrome. This should not result in them automatically receiving medication. The first step in management is to explain the nature of tics to the child and the family, emphasising that although there is some control of tics, this is limited – 'It's like holding your breath' is a useful analogy. Tics are typically made worse by stress, and looking at ways of reducing this can be helpful. (The Marquise de Dampierre went into remission after taking a cure in Switzerland involving frequent milk baths, but relapsed after her return to France and marriage.) The parents should be warned that punishing the child for the tics or repeatedly drawing attention to them is likely to upset the child and actually make the tics worse. It is important to ensure that throat clearing or sniffing tics are not attributed to allergy and then treated with removal of the tonsils and adenoids.

Liaison with the school may be important in preventing harsh treatment from teachers and bullying from other children. The diagnosis of Tourette syndrome may be useful in getting access to additional educational resources. Behavioural techniques may help obsessive compulsive symptomatology. Relaxation exercises, massed practice (voluntarily performing a particular tic a large number of times in front of a mirror) and habit reversal

(performing a completely different movement when the premonitory symptoms occur) may help individual patients, but there is limited evidence of their overall effectiveness.

Drug therapy

Although being near a child who constantly sniffs or clears his throat can test the patience of the calmest of individuals, it must be remembered that there is no drug that stops all tics and has no side effects. As a general rule, medical therapy should only be used to suppress tics if they are significantly disturbing the child's life and other approaches have not worked. Tics are a more immediately obvious symptom than attention deficit hyperactivity disorder, obsessive compulsive disorder, anxiety disorder and family dysfunction. It can be less threatening to the family and easier for the doctor to focus treatment on the number of tics occurring each day rather than these other problems, but it may not be in the best interest of the child. A certain amount of tolerance of human diversity and a sense of humour are helpful.

My practice is to use clonidine (Catapres) initially if, despite other measures, the child and the family feel the tics are intolerable, the child's life is miserable because of teasing, or there is a marked exacerbation of the tics at a time of stress, such as before exams. I try to restrict the use of the stronger neuroleptic agents such as haloperidol (Serenace) to those rare instances where the tics themselves threaten to injure the child (for example, violent neck-jerking tics which can damage the cervical spinal cord), or where the tics are so bizarre or so severe that the child is unable to function away from the home. In complex cases, early referral for psychological counselling or psychiatric evaluation is helpful.

Clonidine

Clonidine is an α_2-adrenoreceptor agonist which was first used as an antihypertensive but later found to have other additional actions. The starting dose is 25 to 50 μg per day given in the evening, and doses of up to 150 μg nocte can be tolerated. It may be particularly useful in children with Tourette syndrome and attention deficit hyperactivity disorder. There may be a delay of four to six weeks before there is an onset of action. The response rate varies: in some children clonidine can be very helpful, but in others it is no use at all. The main complication is drowsiness. Bradycardia and nocturnal restlessness can also occur. If clonidine is suddenly withdrawn, there are the risks of worsening of the tics, increased restlessness and increased heart rate and blood pressure.

Haloperidol

Haloperidol is the most commonly prescribed drug for Tourette syndrome and has been used for its treatment since 1961. It acts primarily as a dopamine D2 receptor blocker. A response rate of 70 to 80% is generally cited, although a recent placebo controlled study found that it was no better than placebo. Between 67 and 80% of patients taking haloperidol find the side effects intolerable and only 10 to 30% take the drug for extended periods of time. (One of my patients referred to it as 'kryptonite' – because the green-coloured tablets weakened him.)

Potential problems with haloperidol include acute dystonic reactions and parkinsonism. Drowsiness, irritability and depression can be major problems. There may be school and social phobias, and excessive appetite and weight gain may occur. Worrying side effects, which are more common in adults but can occur in children, include akathisia, tardive dyskinesia, tardive dystonia and the neuroleptic malignant syndrome. If haloperidol is used in conjunction with tricyclic antidepressants, there is a risk of cardiac arrhythmia, and there have been reports of irreversible encephalopathy when it is combined with lithium.

One of the problems of drug therapy for Tourette syndrome and other neuropsychiatric syndromes is that polypharmacy may gradually develop in an attempt to combat side effects of the original drug. Suggested 'antidotes' to the side effects of haloperidol include:

- benztropine (Cogentin) for the acute dystonic reactions
- anticholinergic drugs for parkinsonism
- beta blockers, clonidine, cyproheptadine (Periactin) and high dose vitamin E for akathisia
- antidepressants for depression
- caffeine in the morning to reduce drowsiness
- exercise and diet to combat weight gain.

This list emphasises the importance of being convinced that drug therapy is necessary before it is begun.

Other drugs

Pimozide (Orap) and atypical neuroleptics such as risperidone (Risperdal) have a similar side effect profile to haloperidol, although the side effects are probably less frequent and less severe. Pimozide has the potential to cause cardiac arrhythmias. Risperidone is increasingly being used for Tourette syndrome, although its PBS listing is for schizophrenia. Weight gain seems more common with risperidone than with haloperidol and it is 10 times as expensive.

Useful contacts

Further information on Tourette syndrome can be obtained from:

Tourette Syndrome Association of Australia
Telephone: (02) 9382 3726, fax: (02) 9382 3764, email: info@tourette.org.au, website: http://www.tourette.org.au (with links to the various State Tourette syndrome associations). This also acts as the local association for NSW.

Tourette Syndrome Association of Queensland
Telephone: (07) 3358 4988

Tourette Syndrome Association of Victoria
Telephone: (03) 9828 7218

Tourette Syndrome Association of Tasmania
Telephone: (03) 6244 7052

Western Australian Tourette Syndrome Organisation (WATSO)
Telephone: (08) 9388 3486

South Australian Tourette Syndrome Association
Telephone: (08) 8357 8909

The large number of other medications that have been tried in Tourette syndrome is testimony to the lack of an 'ideal' drug for the treatment of tics. These have included selective serotonin reuptake inhibitors, tetrabenazine, dopamine agonists, benzodiazepines, nicotine patches, calcium channel blockers, naltrexone, lithium, carbamazepine and marijuana.

Other treatments

Botulinum toxin injections have been given into the muscles producing tics. In veterinary practice, botulinum toxin is injected into the vocal cords to 'de-bark' dogs. A successful vocal cord injection in an adolescent who had severe coprolalia has been reported, but this seems an extreme measure and a rather mechanistic approach.

Various forms of psychosurgery have been tried. Overall experience is limited to less than 40 operations and it should only be regarded as an experimental treatment reserved for exceptional circumstances.

Tourette syndrome and stimulant medication

Tics and Tourette syndrome have been regarded as contraindications to the use of stimulant medications such as methylphenidate (Attenta, Ritalin) and dexamphetamine (Dexamphetamine Tablets), based on reports of onset or worsening of tics when these medications are introduced. However, a number of recent studies have shown that in children with attention deficit hyperactivity disorder, between 10 and 20% developed tics if followed over several years, even without exposure to stimulant medication. Stimulants may make tics worse in some children, particularly if they are taken in high doses. In other children, their use results in fewer tics, presumably because of increased confidence, diminished anxiety and improved self-esteem.

Conclusion

The bizarre syndrome described by Tourette is rare in childhood, but milder forms of Tourette syndrome are common. There are problems with all the drugs that are used to treat tics, and potent neuroleptic agents should not be prescribed automatically. In children who have disabling Tourette syndrome, the tics are often the least of their problems. Their difficulties arise from the associated attention deficit hyperactivity disorder, obsessive compulsive disorder, self-injurious behaviour, conduct disorder or family problems. These children are best managed by a child psychiatrist. MT

Further reading

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