

# Gilles de la Tourette's syndrome

**Y**OUR starter for three: What links prominent 18th-century literary figure Dr Samuel Johnson and Manchester United's new goalkeeper Tim Howard? Answer: Both have been afflicted with Tourette's syndrome (TS). Next question: How much do you know about TS, and why should it concern you as a psychologist?

I believe it is imperative that all healthcare professionals are educated about TS correctly, so they can comment without propagating the many 'media-friendly' myths. The media often misrepresents TS, giving the impression that all sufferers make outbursts of swearing (the *Daily Sport* welcomed Tim Howard with the headline 'No f\*\*\*ing problem'), make the V-sign in public, bark like dogs, have a poor prognosis and are not amenable to treatment. In fact, I feel privileged to have worked with individuals with TS for over 20 years. They are a group of people who are often frustrated not only with their affliction, but also with the public, who



**MARY M. ROBERTSON** with an update and comment on psychobehavioural therapies.

really do not understand TS. For all professionals and people involved with TS in any way, it is their duty to fight the stigma.

## History

The first medical description of TS was in 1825, when Itard reported the case of a French noblewoman, the Marquise de Dampierre. She developed symptoms at the age of seven, and because of her socially unacceptable utterances was forced to live as a recluse until she died at the age of 85. In 1885 Georges Gilles de la Tourette described nine cases of TS, emphasising the triad of multiple tics, coprolalia (swearing) and echolalia (meaningless

repetition) which has gained him eponymous fame. The first reported case of TS in the UK may well have been Mary Hall of Gadsden, described in 1663 by William Drage (Lees *et al.*, 1984). Several notable figures may have had TS (see box below).

From 1900 to 1965 the TS literature was predominantly psychoanalytical, with many classic descriptions by early writers such as Mahler, Kanner, Boncour and Reich. Notions on cause included repressed sexual desires and a moral conflict between ego and superego (for a review see Robertson, 1989).

In my opinion, some of the early descriptions remain the best. They also indulge in primarily psychological explanations, with a curiously biological mixture at other times. Meige and Feindel (1907) discuss echophenomena exquisitely:

*In spite of their frequency among those who are addicted to tic, echolalia and echokinesis cannot be enumerated with the tics, seeing that their exhibition is dependent on the actions of others, whereas once a tic is established it requires no stimulus from without for its manifestation. Of course their affinity to the tics is very close: they spring from*

## NOTABLE INDIVIDUALS

Dr Samuel Johnson (Murray, 1979)

Wolfgang Amadeus Mozart (Simkin, 1992), but this has been challenged (Karhausen, 1998) (Personally,

I think it is doubtful: Mozart had no typical tics, no ADHD, no obsessive compulsions and no relevant family history. Moreover, the obscenities in his writings were written on purpose and must be taken in context.)

Leo Tolstoy's brother Dmitri, portrayed as Nicolai Levin in *Anna Karenina* (Hurst & Hurst, 1994)

Julius Wechter, a marimba player in Herb Alpert's Tijuana Brass band (Carroll & Robertson, 2000)

Jim Eisenreich, major league baseball player (Carroll & Robertson, 2000)

Tim Howard, Manchester United goalkeeper

*the same soil; they represent in the adult the persistence and amplification of the child's propensity for imitation, and therefore in their own way postulate a degree of mental infantilism.* (p.217)

*There is no necessary connection, as a matter of fact, between tic and coprolalia, though of course they may co-exist, sometimes in association with other syndromes; they are in reality only episodic syndromes of hereditary insanity. A distinction ought to be drawn between coprolalia and the use of trivial or inconvenient terms, words with which even some well-educated persons are wont to garnish their conversation.* (p.219)

### The scale and nature of the problem

TS used to be considered a very rare condition, with only case reports documented in the literature (see Robertson, 1989, 1994). The generally accepted prevalence for TS was thought for some time to be 5/10,000 (Bruun, 1984). Throughout the late 1980s and into the 21st century, the medical literature on TS has mushroomed. Large cohorts of patients have been described, including several recent epidemiological studies. Thus it has been demonstrated that TS may be more common than previously suggested, as evidenced by several large-scale investigations (e.g. Kurlan *et al.*, 2001).

The results of the latter studies suggest that a more realistic figure for the prevalence of TS may well be around 1 per cent of mainstream schoolchildren between the ages of six and 17 years. The prevalence of TS in children with special educational needs (Eapen *et al.*, 1997) and autistic spectrum disorders (Baron-Cohen *et al.*, 1999) is even higher. TS is found in all cultures, countries and racial groups and occurs three to four times more commonly in males (Freeman *et al.*, 2000; Robertson 1989, 1994).

TS usually brings with it a range of other problems for sufferers to deal with. Clinic patients have significant non-obscene complex socially inappropriate behaviours (NOSI: Kurlan *et al.*, 1996), plus a variety of psychopathologies (see box 'What is Tourette syndrome?'; see also Robertson, 2000). In one large international study (Freeman *et al.*, 2000) only 12 per cent of TS patients had no other

## WHAT IS TOURETTE'S SYNDROME?

- An inherited neuropsychiatric disorder characterised by the presence of both multiple motor tics (twitches, 'habits') and one or more vocal tics (noises).
- Approximately 1 per cent of mainstream schoolchildren may be affected.
- Symptoms last longer than one year: wax and wane in severity, increase during stress, and decrease when playing sport or a musical instrument; decrease but do not necessarily disappear when asleep.
- Onset under 18 years of age (mean 6.7 years; range 1–17), with motor tics (e.g. eye blinking) usually followed by vocal tics (e.g. throat clearing, sniffing) around the age of nine (Shapiro *et al.*, 1988).
- Associated symptoms and psychopathology commonly include echophenomena (copying behaviours), palilalia (repeating oneself), non-obscene socially inappropriate behaviours, obsessive compulsive behaviours, ADHD and depression. Coprolalia is seen in around 10 per cent of patients), and self-injurious behaviour (with an obsessional quality) in around 30 per cent.
- Cause is genetic in most cases.
- Treatment is by medication and psychological manoeuvres.

psychopathology or comorbidity. However, due to different clinicians using different diagnostic criteria and measurements, estimates of comorbidity of specific disorders vary considerably: for example, ADHD occurs in 20–90 per cent of clinic patients (Robertson & Eapen, 1992), and OCD in anything from 11 per cent to 80 per cent (see Robertson, 1989).

### Searching for a cause

Most evidence suggests that TS is a genetic disorder, transmitted via a single major gene locus with autosomal dominant inheritance. But more recently other mechanisms of inheritance have been suggested, and to date no single gene has

may be due to the lack of variation in the families, since all were selected to have at least one individual with TS.

Other causal factors have also been invoked, including prenatal and perinatal events (Leckman *et al.*, 1990) and streptococcal infections (Swedo *et al.*, 1998); and a further argument for the predominantly autoimmune aetiology in a subset of TS patients has been demonstrated by Taylor *et al.* (2002).

### Treatment

A recent study has suggested an earlier age at onset of TS (5.6 years), severe tics at around 10 years, and the majority of symptoms disappearing in half of the patients by the age of 18 years (Leckman *et al.*, 1998). This relatively good prognosis is important for healthcare professionals to know and to impart to their TS patients.

As with early aetiological theories, early treatments were predominantly psychoanalytical. The pendulum swung to mainly biological aetiologies, and for many years medication was the main method of treatment. Thorough reviews of the pharmacological management of TS appear elsewhere (e.g. Robertson, 2000) and are largely beyond the scope of this article, but medication is the mainstay of treatment of the motor and vocal tics and many of the behaviours of TS.

The commonly used agents are the dopamine-antagonists, including the 'typical' and more recent 'atypical' antipsychotics. The doses used in TS are much smaller than those used in the functional psychoses such as schizophrenia. However, patients may also need stimulants for the ADHD symptoms (if the tics worsen, one can add an antipsychotic) and antidepressants to counter depression and obsessions.

## 'for many years medication was the main method of treatment'

been identified. The recent first genome scan (Tourette Syndrome Association International Consortium for Genetics, 1999) identified areas of interest on chromosomes 4 and 8. A recent factor analysis of TS individuals identified four significant factors: (a) aggressive phenomena (e.g. kicking, tempers); (b) purely motor and phonic tics symptoms; (c) compulsive phenomena (e.g. touching of others/objects); and (d) tapping and the absence of grunting. These four factors accounted for 61 per cent of the observed clinical variation in the TS individuals and their first-degree relatives (Alsobrook & Pauls, 2002). Three of the four factors (a, b and d) were significantly correlated, suggesting that they are inherited. The only factor that did not show significant correlation was the 'pure tic' factor. This

Nicotine transdermal patches, botulinum toxin injections to localised sites (e.g. vocal cords) and cannabinoids have all been documented as useful for symptoms. Clinicians may have to give more than one medication for particular symptom profiles, and clearly they must be familiar with symptom profiles and take great care when giving children medication.

Currently, experts acknowledge a biological aetiology but think that a holistic approach in treatment is probably the best way forward. This approach is reflected in a multidisciplinary and multifaceted approach to treating patients with TS. In expert clinics, management can range from psychoeducation and supportive reassurance, to intricate pharmacological interventions. Psychoeducation is mandatory; in my clinic, personalised factsheets, reading lists, and 'question and answer' leaflets are routinely given to all patients. In many patients with mild symptomatology, and for many children, education and reassurance and supportive psychotherapy for the patients and relatives may be sufficient.

Although psychological methods are helpful with many symptoms of TS, documentations of success have, until quite recently, been mainly individual case reports or small series of patients. The box below gives the types of psychological

## PSYCHOLOGICAL THERAPIES

Psychotherapy (MacKay & Heimlich, 1972)

Hypnotherapy (Clements, 1972)

Massed negative practice (e.g. Storms, 1985) – over-rehearsal of target tic, e.g. blinking, to result in fatigue and render the patient unable to produce the tic involuntarily.

Brief behaviour therapy (Carr & Bailey, 1996)

EEG sensorimotor rhythm biofeedback training (Tansey, 1986)

Assertiveness training (Mansdorf, 1986)

Self-monitoring (Billings, 1978)

Social skills group (Lambert & Christie, 1998)

Relaxation imagery (self-hypnosis) (Kohen & Botts, 1987)

Cognitive-behavioural therapy

Habit reversal – a multicomponent procedure consisting of awareness training, self-monitoring, relaxation, competing response training and contingency management



Georges Gilles de la Tourette

therapies that have been documented to be successful in treating patients with TS. On the other hand, behavioural techniques and relaxation therapy have not proved useful in the treatment of tics in TS (e.g. Bergin *et al.*, 1998).

Despite these modest beginnings, there are an increasing number of useful papers about psychological therapies used for treating TS. Peterson and Cohen (1998) discuss the multimodal developmental intervention in TS, and the importance of psychoeducation and supportive psychotherapy. They also note that early reports of successful treatment of tics with psychoanalysis have not been replicated, stating that dynamic psychotherapies have little or no efficacy in reducing tic symptoms.

Behaviour therapy or cognitive behaviour therapy (CBT) is well described as being successful in the treatment of some disorders such as obsessive compulsive disorder (OCD) (e.g. Baxter *et al.*, 1992). In the Tourette's patient with OCD it is worth using CBT along with a selective serotonin reuptake inhibitor and a small dose of neuroleptic. Future studies should examine further the use of CBT in individuals with TS.

Habit reversal or habit reversal training (HRT) is a multicomponent procedure developed by Azrin and Nunn (1973) for the treatment of nervous habits and tics. It consists of awareness training, self-monitoring, relaxation training, competing response training and contingency management (e.g. Wilhelm *et al.*, 2003). Several authors then described HRT as successful in a single case (Tolchard, 1995) and studies using small numbers of patients with TS (e.g. Peterson & Azrin, 1992; Woods *et al.*, 2003). The momentum in this type of treatment grew, and a large series of 47 patients with chronic tic disorder indicated successful use of HRT (O'Connor *et al.*, 2001).

The first controlled study of HRT was small ( $N = 10$ ) but successful (Azrin & Peterson, 1990). Finally, success with this modality of treatment was recently documented by Wilhelm *et al.* (2003) in an elegant randomised controlled trial of HRT versus supportive psychotherapy in 32 patients with TS, who were carefully selected, assessed and allocated to an HRT group or a supportive psychotherapy group.

Seven patients in each group were medicated. Changes in the severity of tic symptoms (as measured by the Yale Global Tic Severity Scale) as well as psychosocial impairment (Clinical Global Impression Improvement Scale) were examined over the course of a 14-session treatment for 29 patients who completed at least eight treatment sessions. Results indicated the 16 patients in the HRT group improved significantly better than the 13 patients in the supportive psychotherapy group at 10-month follow-up. This provides great encouragement for the future – it does seem that HRT is a useful behavioural therapy for TS. (See Piacentini & Chang, 2001, for an earlier review of the literature on behavioural methods of treatment.)

Guidance to teachers and educators about youngsters and pupils with TS is also important (e.g. Stefl & Rubin, 1985). Carroll and Robertson (2000) highlight the importance of raising pupils' self-esteem and self-confidence, the importance of skill building, refocusing attention, and teaching the child to understand anger while acknowledging the pain caused by peer rejection. They also discuss a whole-school approach to bullying as well as ways of promoting language and social skills.

Many patients have faith in a host of complementary therapies because they are seen to do good and have no adverse side-effects. But there is little if any evidence for them – more research is needed.

### Conclusion

Although more recent studies suggest that Tourette's syndrome is quite common, it remains an elusive and controversial disorder. This is an interesting and rewarding area, and psychologists should not keep leaving the interest and rewards

to psychiatrists and neurologists. Yes, medication is important, but a multidisciplinary approach is in vogue, and many psychologists could contribute to helping ease the frustration of people with TS.

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## WEBLINKS

Voluntary organisations play a vital role in educating the public and the media, and helping families, patients, teachers and family practitioners.

Tourette Syndrome (UK) Association:

[www.tsa.org.uk](http://www.tsa.org.uk)

Tourette Syndrome Association Inc.:

[tsa.mgh.harvard.edu](http://tsa.mgh.harvard.edu)

### References

- Alsobrook, J.P. & Pauls, D.L. (2002). A factor analysis of tic symptoms in Gilles de la Tourette's syndrome. *American Journal of Psychiatry*, 159, 291–296.
- Azrin, N.H. & Nunn, R.G. (1973). Habit reversal: A method of eliminating nervous habits and tics. *Behaviour Research and Therapy*, 11, 619–628.
- Azrin, N.H. & Peterson, A.L. (1990). Treatment of Tourette syndrome by habit reversal. *Behavior Therapy*, 21, 305–318.
- Baron-Cohen, S., Scahill, V.L., Izaguirre, J., Hornsey, H. & Robertson, M.M. (1999). The prevalence of Gilles de la Tourette syndrome in children and adolescents with autism: A large scale study. *Psychological Medicine*, 29, 1151–1159.
- Baxter, L.R., Schwartz, J.M., Bergman, K.S. et al. (1992). Caudate glucose metabolic rate changes with both drug and behaviour therapy for obsessive-compulsive disorder. *Archives of General Psychiatry*, 49, 681–689.
- Billings, A. (1978). Self-monitoring in the treatment of tics. *Journal of Behavior Therapy and Experimental Psychiatry*, 9, 339–342.
- Bergin, A., Waranch, H.R., Brown, J., Carson, K. & Singer, H.S. (1998). Relaxation therapy in Tourette syndrome: A pilot study. *Pediatric Neurology*, 18, 136–142.
- Bruun, R.D. (1984). Gilles de la Tourette's syndrome: An overview of clinical experience. *Journal of the American Academy of Child and Adolescent Psychiatry*, 23, 126–133.
- Carr, J.E. & Bailey, J.S. (1996). A brief behavior therapy protocol for Tourette syndrome. *Journal of Behavior Therapy and Experimental Psychiatry*, 1, 33–40.
- Carroll, A. & Robertson, M.M. (2000). *Tourette syndrome: A practical guide for teachers, parents and carers*. London: David Fulton.
- Clements, R.O. (1972). Gilles de la Tourette's syndrome – An overview of development and treatment of a case, using hypnotherapy, haloperidol, and psychotherapy. *American Journal of Clinical Hypnosis*, 14(3), 167–172.
- Eapen, V., Robertson, M.M., Zeitlin, H. & Kurlan, R. (1997). Gilles de la Tourette's syndrome in special education schools. *Journal of Neurology*, 244, 378–382.
- Freeman, R.D., Fast, D.K., Burd, L., Kerbeshian, J., Robertson, M.M. & Sandor, P. (2000). Tourette Syndrome International Database Consortium. *Developmental and Medical Child Neurology*, 42, 436–447.
- Hurst, M.J. & Hurst, D.L. (1994). Tolstoy's description of Tourette syndrome in Anna Karenina. *Journal of Child Neurology*, 94, 366–367.
- Karhausen, L.R. (1998). Weeding Mozart's medical history. *Journal of the Royal Society of Medicine*, 91, 546–550.
- Kohen, D.P. & Botts, P. (1987). Relaxation-imagery (selfhypnosis) in Tourette syndrome. *Journal of Clinical Hypnosis*, 29, 227–237.
- Kurlan, R., Daragjati, C., Como, P.G., McDermott, M.P., Trinidad, K.S., Roddy, S. et al. (1996). Non-obscene complex socially inappropriate behavior in Tourette's syndrome. *Journal of Neuropsychiatric and Clinical Neurosciences*, 8, 311–317.
- Kurlan, R., McDermott, M.P., Deeley, C., Como, P.G., Brower, C., Eapen, S. et al. (2001). Prevalence of tics in schoolchildren and association with placement in special education. *Neurology*, 57, 1383–1388.
- Lambert, S. & Christie, D. (1998). A social skills group for boys with Tourette's syndrome. *Clinical Child Psychology and Psychiatry*, 3, 267–277.
- Leckman, J.F., Dolnansky, E.S., Hardin, M.T., Clubb, M., Walkup, J.T., Stevenson, J. et al. (1990). Perinatal factors in the expression of Tourette's syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry*, 29, 220–226.
- Leckman, J.F., Zhang, H., Vitale, A., Lahnin, F., Lynch, K., Bondi, C. et al. (1998). Course of tic severity in Tourette syndrome: The first two decades. *Pediatrics*, 102, 14–19.
- Lees, A.J., Robertson, M.M., Trimble, M.R. & Murray, N.M.F. (1984). A clinical study of Gilles de la Tourette syndrome in the United Kingdom. *Journal of Neurological and Neurosurgical Psychiatry*, 47, 1–8.
- MacKay, M.C. & Heimlich, E.P. (1972). Psychotherapy with paraverbal therapy in a case of Gilles de la Tourette syndrome. *American Journal of Psychotherapy*, 26, 571–577.
- Mansdorf, I.J. (1986). Assertiveness training in the treatment of a child's tics. *Journal of Behavior Therapy and Experimental Psychiatry*, 17, 29–32.
- Meige, H. & Feindel, E. (1907). *Tics and their treatment* (Trans. S.A.K. Wilson). London: Sidney Appleton.
- Murray, T.J. (1979). Dr Samuel Johnson's movement disorder. *British Medical Journal*, 1, 1610–1614.
- O'Connor, K.P., Brault, M., Robillard, S., Loiselle, J., Borgeat, F. & Stip, E. (2001). Evaluation of a cognitive-behavioural program for the management of chronic tic and habit disorders. *Behaviour Research and Therapy*, 39, 667–681.
- Peterson, A.L. & Azrin, N.H. (1992). An evaluation of behavioral treatments for Tourette syndrome. *Behaviour Research and Therapy*, 30, 167–174.
- Peterson, B.S. & Cohen, D.J. (1998). The treatment of Tourette's syndrome: Multimodal, developmental intervention. *Journal of Clinical Psychiatry*, 59(Suppl), 62–72.
- Piacentini, J. & Chang, S. (2001). Behavioral treatments for Tourette syndrome and tic disorders. *Advances in Neurology*, 85, 319–331.
- Robertson, M.M. (1989). The Gilles de la Tourette syndrome: The current status. *British Journal of Psychiatry*, 154, 147–169.
- Robertson, M.M. (1994). Annotation: Gilles de la Tourette syndrome – An update. *Journal of Child Psychology and Psychiatry*, 35, 597–611.
- Robertson, M.M. (2000). Tourette syndrome, associated conditions and the complexities of treatment. *Brain*, 123, 425–462.
- Robertson, M.M. & Eapen, V. (1992). Pharmacologic controversy of CNS stimulants in Gilles de la Tourette's syndrome. *Clinical Neuropharmacology*, 15, 408–425.
- Shapiro, A.K., Shapiro, E.S., Young, J.G. & Feinberg, T.E. (1988). *Gilles de la Tourette syndrome* (2nd edn). New York: Raven Press.
- Simkin, B. (1992). Mozart's scatological disorder. *British Medical Journal*, 305, 1563–1567.
- Steffl, M.E. & Rubin, M. (1985). Tourette syndrome in the classroom. *Journal of School Health*, 55, 72–75.
- Storms, L. (1985). Massed negative practice as a behavioral treatment for Gilles de la Tourette's syndrome. *American Journal of Psychotherapy*, 39, 277–281.
- Swedo, S.E., Leonard, L.H., Garvey, M., Mittleman, B.B., Allen, A.J. & Perlmutter S. (1998). Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections: Clinical description of the first 50 cases. *American Journal of Psychiatry*, 155, 264–271.
- Tansey, M.A. (1986). A simple and complex tic (Gilles de la Tourette's syndrome): Their response to EEG sensorimotor rhythm biofeedback training. *International Journal of Psychophysiology*, 4, 91–97.
- Taylor, J.R., Morshed, S.A., Parveen, S., Mercandante, M.T., Scahill, L., Peterson, B.S. et al. (2002). An animal model of Tourette's syndrome. *American Journal of Psychiatry*, 159, 657–660.
- Tolchard, B. (1995). Treatment of Gilles de la Tourette syndrome using behavioural psychotherapy: A single case example. *Journal of Psychiatric and Mental Health Nursing*, 2(4), 233–236.
- Tourette Syndrome Association International Consortium for Genetics (1999). A complete genome scan in sib-pairs affected with Gilles de la Tourette syndrome. *American Journal of Human Genetics*, 65, 1428–1436.
- Wilhelm, S., Deckersbach, T., Coffey, B., Bohné, A., Peterson, A. & Baer, L. (2003). Habit reversal versus supportive psychotherapy for Tourette's disorder: A randomized controlled trial. *American Journal of Psychiatry*, 160, 1175–1177.
- Woods, D.V., Twohig, M.P., Flessner, C.A. & Roloff, T.J. (2003). Treatment of vocal tics in children with Tourette syndrome: Investigating the efficacy of habit reversal. *Journal of Applied Behavior Analysis*, 36(1), 109–112.