Professor Mary Robertson Prize 2012,

Jonathan Mills

I certify that this work is my own, written of my own accord following an elective placement in Child and Adolescent Psychiatry.

Jonathan Mills

11/7/2012

Professor Mary Robertson Prize 2012,

Jonathan Mills

Abstract

The objective of this essay is to examine emerging, novel evidence of improved functioning in individuals with Tourette's syndrome, highlighting cases where people have excelled in their respective fields despite a diagnosis of Tourette's syndrome. The neuroscience behind function in Tourette's will be discussed, and an argument made that we should change our attitudes towards those with Tourette's, respecting them for their merit, capability and contribution to society.

Introduction

Tourette Syndrome has existed for centuries, with one of the earliest recordings of probable Tourette's dating to the 15th century, describing the case of a priest 'possessed by the devil' (Finger, 1994 as quoted by Teive et al, 2008). It was only towards the end of the 19th century before Tourette's was formally recognised.

It is thought Tourette's is an inherited, developmental neuropsychiatric disorder that first presents in childhood, with motor and vocal tics that typically last over a year, starting around the age of 7 with a degree of resolution into adolescence (APA, 2000; Leckman et al. 1998; Bloch et al, 2006). Genetic and environmental factors contribute substantially to an individual's susceptibility, though the mode of inheritance has yet to be identified (Bloch et al. 2011), the genetic factors implicated show variable degrees of penetrance and expression (Van de Weterine and Heutink, 1993). Thus a child with a parent affected by Tourette's may not go on to develop the condition, equally a child with parent's who do not suffer with Tourette's may also develop the condition.

The presentation of Tourette's is one of involuntary tics, motor and vocal that can be innocuous such as blinking or throat clearing, causing no problems to the individual, whilst at the other end of the spectrum, the uncommon presentation of coprolalia (uttering of obscene language) which is highly apparent and considerably distressing to the patient. What characterises Tourette's is the improvement of tics whilst the patient sleeps and whilst the patient consciously attempts to suppress them or whilst absorbed in activities (Wood et al. 2003; O'Connor et al. 2003). Suppression however results in inner tension that resolves when the tic is discharged, with some patients reporting premonitions and the ability to identify when they feel a tic developing (Kwak et al. 2003; Banaschewski et al, 2003).

Whilst Tourette's is associated with a number of social difficulties that will be discussed within this essay, there is suggestion that Tourette's confers certain advantages to the individual (Leckman and Cohen, 1999). The levels of increased function will be the focus of this essay, and propose that is society, stigma and misconceptions surrounding Tourette's that causes the disability associated with Tourette's, when many patients are in fact very abled.

Portrayal of Tourette's in the public eye

"I hate feeling like a lower-class citizen, a freak."

'Chopper', Tourette's: I swear I can't help it.

It is worth considering the stigma associated with Tourette's, identifying the public perception surrounding the condition. It is a sad reflection of society that Tourette's is portrayed negatively by the media, with those suffering from the condition being the object of fun. One of the commonest portrayals demonstrated is that of coprolalia- the shouting of obscene language. Yet this is an uncommon presentation with only around 10% of suffers exhibiting coprolalia as a symptom (Singer, 2005). Why is it then, that an uncommon symptom attracts much attention and has come to portray Tourette's to a wider audience? John Davidson, who featured on a BBC documentary regarding Tourette's suggests much of the focus stems from Tourette's being considered comical, querying "If you ask anybody what's the funniest illness, most people will say Tourette's." (BBC, 2009). Certainly for a television show portraying the average sufferer of Tourette's as they are- normal- it would not make for good viewing. But, portraying the rarer, more extreme presentation of a child blurting out profanity in the most inappropriate of places makes for audience figures and consequently the stereotype is fuelled. Whilst it is fortunate that some media outlets have channelled their energies for the benefit of raising awareness and challenging the stigma surrounding Tourette's, other shows continue to hold outdated and ignorant views, such as suggesting those with Tourette's be ostracised from important events and sensationalising the condition.

From a legal perspective, there is no evidence to suggest those suffering from Tourette's are more likely to be criminals, or fail to understand and comply with the law (Jankovic et al, 2006). Yet, Tourette's is one condition where injustice and exclusion have been allowed to continue as a consequence of poor understanding of the cause. An explanation for how the stigma has arisen was suggested that perception of those seen to be disruptive to public

order attracts negative judgement due to the opinion they are 'out of control'(Davis et al. 2004). The public attitude towards behavioural disorders lends itself that those with behavioural disorders such as Tourette's are thought to be unpredictable and difficult to speak to (Crisp et al. 2000).

Having outlined examples of stigma and misconceptions surrounding Tourette's, and possible reasons for why these views are allowed to proliferate, it is worth moving to consider positive figures with tourettes.

Social impact and famous people

In challenging stigma and misconceptions, it is worth demonstrating that individuals can lead successful lives and gain respect from their skills and talents despite the burden of Tourette's. Perhaps the most notable individual to have had Tourette's is believed to be Samuel Johnson (Pearce, 1994), credited with the concept of creating the dictionary. During the 18th century, before Tourette's became formally identified, Johnson had gained fame for his wit and writing despite physical disability. His work *'A Dictionary of the English Language'* published in 1755 brought Johnson great success and was described by Bate (1977) as "one of the greatest single achievements of scholarship". Though Tourette's was not formally described until a century after Johnson's death, what is believed to have been tics were eloquently described and recorded by his close companion Boswell in Hibbert (1971), "He commonly held his head to one side, moving his body backwards and forwards, rubbing his left knee in the same direction with the palm of his hand". Indeed Johnson in addition to this description of motor tics also demonstrated vocal tics described by Boswell as making "various sounds like a half whistle or as if clucking like a hen". It is difficult to imagine how those with Tourette's historically may have been treated, whilst it could be suggested a man of Johnson's standing was shielded from being ostracised, lack of understanding is likely to have resulted in ill treatment.

In the field of Politics, André Malraux (1901-1976) an accomplished writer and artist rose to become Minister of State and Minister for cultural affairs under Charles De Gaulle, preserving his nations heritage. Such was Malraux's status, following an official visit to the United States, Jacqueline Kennedy, wife of President John F Kennedy was noted to describe him as "the most fascinating man I've ever talked to." (New York Times, 2011). It is speculated that Malraux had Tourette's, having been described as having vocal and motor tics during childhood (Todd, 2005). Whether Malraux actually had Tourette's or another movement disorder remains to be determined as he never formally acknowledged the condition. This aside, demonstrates that despite adversity, an individual can rise to achieve great things.

Everton FC goalkeeper, Tim Howard has publically acknowledged he suffers with Tourette's, though this has not stopped him playing football at the international level. As one example of someone with Tourette's who has excelled at sport, Howard became the fourth

goalkeeper in the history of the premier league to score a goal (BBC sport). Howard is just one example of many sports personalities to suffer with the condition, yet perform at a high level.

Neurologist Oliver Sacks described a case of a surgeon and pilot referred to as 'Carl Bennett' in his work "*The Anthropologist from Mars" (Sacks, 1995).* 'Bennett' spoke of finding medical school difficult, his tics resulting in reading things over and over again, and times in clinic repeat words hundreds of times. Sacks wrote about how during lengthy operations 'Bennett' was capable of such concentration he was able to operate without a tic. 'Bennett' gained a reputation as being a particularly skilled and capable surgeon, lecturing on anatomy.

These cases represent just a handful of individuals who have excelled in their respective fields, overcoming the problems associated with Tourette's. Indeed a number of highly successful people who suffer from Tourette's appear to be able to suppress their tics when required to do so, using this ability to further their performance.

<u>Evidence of improved function</u>

"It's like a miracle," one of them says. "The way the Tourette's disappears."

'Carl Bennett', A surgeon's life.

Having highlighted specific individuals in a variety of fields with considerable talent, it is worth reviewing the literature for evidence of wider applicability. One such area demonstrating temporal advantage in those with Tourette's was a study comparing children with Tourette's only against matched controls without Tourette's. The finding of the study by Vicario et al. (2010) found that the lower the severity of tics, the faster and more precise the ability to respond to a timing interval. The paper hypothesised this may be a pathophysiological consequence of Tourette's, or may be a compensatory mechanism associated with the conscious exertion to supress tics. Mueller et al. (2006) used an oculomotor switching task whereby subjects had to look towards one direction if a stimulus was one colour, and away from it if it were another. It was found that the Tourette's group were more accurate without compromising response time. The study showed that individuals with Tourette's had greater levels of cognitive control than their age-matched counterparts, with the suggestion the need for tic suppression may result in enhanced executive processes for inhibitory control. The study was later supported by Jackson et al., (2007) who examined whether patients with Tourette's performed better when the order of tasks occurred randomly. The study found that when those with Tourette's and the control group were 'briefed' about the task, they both benefited. However, when both groups were

exposed to a random and 'unbriefed' task, the control group demonstrated a significant reduction in performance whilst the Tourette's group did display significant reduction in performance. The study by Jackson et al. added support to the emerging evidence that those with Tourette's have paradoxically greater levels of cognitive control compared to an age matched control group.

In a study by Bialystok and Senman (2004), bilingual children were compared to monolingual children. The children who spoke two languages were better at answering 'reality' questions when given an object that looked like something else (appearance questions); so the example of a pen shaped like a whale, both monolingual and bilingual children could answer what the object looked like (a whale), but on asking what the object really was (a pen), bilingual children performed more competently. Though the study did not use children with Tourette's, the study provides evidence that being able to suppress one language over another provides greater executive control and thus, greater performance. By extension, the hypothesis that suppression of tics (like suppression of another language) leads to greater executive functioning.

Other studies have proved equivocal, suggesting no difference in a go/no-go performance task between those with Tourette's and age-matched controls (Roessner et al., 2008), whilst other studies employing the same go/no-go performance task showed delayed response time with no difference in accuracy (Eichele et al., 2010). What the literature demonstrates however, is how poorly we understand Tourette's, and that suggestions that an individual with Tourette's is less capable than their peers is incorrect. Further research may yield a more definitive answer, but certainly some evidence emerging is encouraging that those affected by Tourette's are every bit as capable (if not more so) than others in society.

Physiology of function

It is worth exploring the science behind function within Tourette's, as a better understanding of the physiology and neuroanatomy may lead us to greater appreciation of an individual affected by Tourette's. Albin and Mink's (2006) study provided evidence that abnormalities within the basal ganglia were responsible for altered function within Tourette's. The nuclei that comprise the basal ganglia such as the striatum, sub-thalamic nucleus, globus pallidus and substantia nigra and their respective neural circuits are associated with sensorimotor and cognitive functioning (Alexander et al. 1986). An individual with Tourette's is likely to have smaller basal ganglia nuclei than an individual without Tourette's, particularly the caudate nucleus as demonstrated by MRI studies (Peterson et al. 1998). It is difficult however, to distinguish between whether the structural changes are as a result of or as a response to Tourette's as the basal ganglia influence and are influenced by the cortex (Jackson, 2006; Albin and Mink, 2006). Additionally, changes evident in childhood are not apparent in adulthood, as prefrontal volume is greater in

children though the reverse is true in adults with Tourette's when compared to controls (Peterson et al., 1998). Dopaminergic pathways are thought to have a role in the causation of tics, particularly ascending pathways from the midbrain to striatum and cortex (Swain et al., 2007), and in particular the meso-cortical and meso-striatal dopaminergic pathways are particularly significant in action planning and decision making (Meck, 1996). Abnormalities within the dopaminergic transmission of the meso-striatal and meso-cortical pathways, in addition to abnormal connection between the basal ganglia and prefrontal cotex are also apparent in individuals with Tourette's, and there is evidence from functional imaging studies that support changes to cerebellar structures during tics (Lerner et al., 2007; Bohlhalter et al., 2006)

The Dorsolateral Prefrontal Cortex appears implicated in regulating inhibitory control, whilst the increased prefrontal cortical volumes observed in children with Tourette's are thought to represent structural changes within the cortex that voluntarily suppress tics (Baym et al., 2008; Stern et al,. 2008). Sowell et al., (2008) reported primary sensorimotor cortical thickness correlated inversely to tic severity, an explanation favoured by Vicario et al. (2010) as an explanation for their study finding faster response times in the Tourette's group. However, a great deal of the neuroanatomy and neurophysiology behind Tourette's still remains to be elucidated.

<u>In support of those affected by Tourette's.</u>

"Tourette's Syndrome is not a problem. It is part of my life."

Tim Howard, Goalkeeper Everton F. C.

With time and greater understanding of Tourettes, achievements by talented individuals will erode the stigma associated with Tourette's. As more research emerges over skill and performance in those with Tourette's, it is possible we may find that misconceptions are corrected and achievements (perhaps as a consequence of Tourette's) will be celebrated.

So in summary, we have looked at the stigma and misconceptions facing the patient with Tourette's, challenging how the condition is portrayed in the media. Famous individuals with Tourette's have also been discussed along with their contributions and talents. The evidence that is emerging of individuals with Tourette's having higher levels of functioning and reasons behind this has also been explored along with the neuroscience behind why this may occur.

Perhaps, given some of the evidence that is emerging, it is time to make the suggestion that we move from labelling people with Tourette's as 'different', stigmatising them and correct misconceptions of the person with Tourette's as being a coprolaliac who shouldn't attend

social engagements. Instead, we should be focussing on the merits of the individual, respecting them for the person they are.

Word Count: 2564

References

- Albin RL, Mink JW (2006) Recent advances in Tourette Syndrome research. Trends Neurosci 29:175–182
- Alexander GE, DeLong MR, Strick PL (1986) Parallel organization of functionally segregated circuits linking basal ganglia and cortex. Annu Rev Neurosci 9:357–38
- APA Diagnostic and Statistical Manual of Mental Disorders (4th Edition, Text Revision (DSM-IV-TR))American Psychiatric Association, Washington, DC (2000)
- Bate, Walter Jackson (1977), *Samuel Johnson,* New York: Harcourt Brace Jovanovich
- Baym CL, Corbett BA, Wright SB, Bunge SA, 2008, Neural correlates of tic severity and cognitive control in children with Tourette syndrome, Brain, 131:165-179
- Bialystok E, Senman L (2004) Executive processes in appearance-reality tasks: the role of inhibition of attention and symbolic representation. Child Dev 75:562–579
- BL Wood, K Klebba, O Gbadebo, D Lichter, R Kurlan, B Miller Pilot study of effect of emotional stimuli on tic severity in children with Tourette's syndrome Mov Disord, 18 (2003), pp. 1392–1395
- Bloch M, State M, Pittenger C. "Recent advances in Tourette syndrome". *Curr. Opin. Neurol.* 2011 Apr;24(2):119–25. doi:10.1097/WCO.0b013e328344648c PMID 21386676
- Bohlhalter S, Goldfine A, Matteson S, Garraux G, Hanakawa T, Kansaku T et al. 2006, Neural correlates of tic generation in Tourette syndrome: An event-related functional MRI study, Brain, 129: 2029-2037
- C Kwak, K Dat Vuong, J Jankovic Premonitory sensory phenomenon in Tourette's syndrome Mov Disord, 18 (2003), pp. 1530–1533
- Crisp AH, Gelder MG, Rix S *et al*. Stigmatisation of people with mental illness. *Brit J Psychiatry* 2000;177:4-7.
- Davis KK, Davis JS, Dowler L. In motion, out of place: the public space(s) of Tourette Syndrome.*Soc Sci & Med* 2004;59: 103-112
- Eichele H, Eichele T, Hammar A, Freyberger HJ, Hugdahl K, et al. (2010) Go/No-go performance in boys with Tourette syndrome. Child neuropsychology 16: 162–168.
- Hibbert, Christopher (1971), The personal History of Samuel Johnson, New York: Harper & Row, ISBN: 0-06-011879-2
- http://www.bbc.co.uk/sport/0/football/16425923 (last accessed 11/7/2012)
- In tapes, Candid Talk by Young Kennedy Widow, article in The New York Times, September 11th 2011.
- J.F. Leckman, H. Zhang, A. Vitale *et al.* Course of tic severity in Tourette syndrome: the first two decades Pediatrics, 102 (1998), pp. 14–19
- Jackson GM (2006) Tourette's Syndrome. Curr Biol 16:443–444

- Jackson GM, Mueller SC, Hambleton K, Hollis CP (2007), Enhanced cognitive control in Tourette Syndrome during task uncertainty. Exp Brain Res. 182(3): 357-64.
- Jankovic J, Kwak C, Frankoff R. Tourette's syndrome and the law. *J Neuropsychiatry Clin Neurosci.* 2006 Winter;18(1):86–95.doi:10.1176/appi.neuropsych.18.1.86 PMID 16525075
- K O'Connor, H Briseboise, M Brault, S Robillard, J Loiselle Behavioral activity associated with onset in chronic tic and habit disorder Behav Res Ther, 41 (2003), pp. 241–249
- Leckman JF, Cohen DJ. *Tourette's Syndrome—Tics, Obsessions, Compulsions: Developmental Psychopathology and Clinical Care.* John Wiley & Sons, Inc., New York. 1999:18–19, 148–151; 408. ISBN 0-471-16037-7
- Lerner, A, Bagic A, Boudreau EA, Hanakawa T, Pagan F, Mari Z et al. 2007, Neuroimaging of neuronal circuits involved in tic generation in patients with Tourette syndrome, Neurology, 68, 1979-1987
- M.H. Bloch, B.S. Peterson, L. Scahill *et al.* Adulthood outcome of tic and obsessive-compulsive symptom severity in children with Tourette syndrome Arch Pediatr Adolesc Med, 160 (2006), pp. 65–69
- Malraux: A life, Olivier Todd, 2005, Knopf ISBN:0375407022
- Meck, WH, 2996, Neuropharmacology of timing and time perception, Brain Research: Cognitive Brain Research, 3:227-242
- Mueller SC, Jackson GM, Dhalla R, Datsopoulos S, Hollis CP (2006), Enhanced cognitive control in young people with Tourette's Syndrome. Curr Biol 16:570–573
- Pearce JM. "Doctor Samuel Johnson: 'the great convulsionary' a victim of Gilles de la Tourette's syndrome" (PDF). *Journal of the Royal Society of Medicine*. 1994 Jul;87(7):396–9
- Peterson BS, Skudlarski P, Anderson AW, Zhang H, Gatenby JC, Lacadie CM, Leckman JF, Gore JC (1998) A functional magnetic resonance imaging study of tic suppression in Tourette Syndrome. Arch Gen Psychiatry 55:326–333
- Roessner V, Albrecht B, Dechent P, Baudewig J, Rothenberger A. Normal response inhibition in boys with Tourette syndrome. Behavioral and Brain Functions. 2008;4:29
- Singer HS (March 2005). "Tourette's syndrome: from behaviour to biology". *Lancet Neurol* 4 (3): 149–59. doi:10.1016/S1474-4422(05)01012-4.PMID 15721825.
- Sowell ER, Kan E, Yoshii J, Thompson PM, Bansal R, Xu D et al. 2008, Thinning of sensorimotor cortices in children with Tourette syndrome, Nature Neuroscience, 11: 637-639
- Stern ER, Blair C, Peterson BS, 2008, Inhibitory deficits in Tourette's syndrome, Developmental Psychobiology, 50: 9-18
- Swain, JE, Scahill L, Lombroso PJ, King RA, Leckman JF, 2007, Tourette syndrome and tic disorders: A decade of progress, Journal of American Academy of Child & Adolescent Psychiatry, 46, 947-968
- T Banaschewski, W Woerner, A Rothenberger Premonitory sensory phenomena and suppressibility of tics in Tourette syndrome: developmental aspects in children and adolescents Dev Med Child Neurol, 45 (2003), pp. 700–703
- Teive HA, Chien HF, Munhoz RP, Barbosa ER. "Charcot's contribution to the study of Tourette's syndrome". *Arq Neuropsiquiatr*. 2008 Dec;66(4):918–21. PMID

19099145as reported in Finger S. "Some movement disorders." In Finger S (ed). *Origins of neuroscience: the history of explorations into brain function.* New York: Oxford University Press, 1994:220–239.

- The Anthropologist from Mars, Oliver Sacks, "A Surgeon's Life", 1995, Picador, ISBN: 0330343475
- Tourette's: I swear I can't help it. Television show, BBC, 2009. Stuart Colquhoun (also known as Chopper).
- Tourette's: I swear I can't help it. Television show. BBC. 2009. John Davidson
- van de Wetering BJ, Heutink P. "The genetics of the Gilles de la Tourette syndrome: a review". *J Lab Clin Med.* 1993 May;121(5):638–45
- Vicario CM, Martino D, Spata F, Defazio G, Giacchè R, Martino V, Rappo G, Pepi AM, Silvestri PR, Cardona F, Time processing in children with Tourette's syndrome, Brain and Cognition, Volume 73, Issue 1, June 2010, Pages 28-34.