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Review article

Famous people with Gilles de la Tourette syndrome?

Francesco Monaco[a], Serena Servo[a], Andrea Eugenio Cavanna[b,c,*]

[a]Department of Neurology, Amedeo Avogadro University, Novara, Italy
[b]Department of Neuropsychiatry, University of Birmingham and BSMHFT, Birmingham, United Kingdom
[c]Sobell Department of Motor Neuroscience and Movement Disorders, Institute of Neurology, UCL, London, United Kingdom

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Abstract

Virtually no neurologist nor psychiatrist today can be unaware of the diagnosis of Gilles de la Tourette syndrome (GTS). Although the eponymous description by Dr. Georges Gilles de la Tourette was published in 1885, familiarity with this syndrome has been achieved only recently. In this article, the two most renown accounts of exceptional individuals retrospectively diagnosed with GTS are critically analyzed: British lexicographer Samuel Johnson and Austrian musician Wolfgang Amadeus Mozart. In both cases, clinical descriptions have been retrieved from written documents predating Gilles de la Tourette's original publication. The case for Samuel Johnson having GTS is strong, mainly based on Boswell's extensive biographical account. Johnson was reported to have a great range of tics and compulsions, including involuntary utterances, repetitive ejaculations, and echo-phenomena. On the other hand, there is circumstantial evidence that Mozart may have had hyperactivity, restlessness, sudden impulses, odd motor behaviors, echo/palilalia, love of nonsense words, and scatology, the latter being documented in autograph letters ("coprographia"). However, the evidence supporting the core features of GTS, i.e., motor and vocal tics, is rather inconsistent. Thus, GTS seems to be an implausible diagnosis in Mozart's medical history and completely unrelated to his undisputed musical genius.

Keywords: Gilles de la Tourette syndrome; Tics; Famous people; Samuel Johnson; Wolfgang Amadeus Mozart

Famous people, famous diseases: an artificial association

Myths and legends have always followed the lives of famous people throughout history, so that also their diseases are often matter of deep interest, but at the same time of extreme controversy. In a number of cases, specific issues make the exact posthumous diagnosis of the presumed clinical syndrome at least questionable. First of all, as we go back in time, the medical witnesses become more and more confuse, and even in the biographies of people who lived in the last two centuries, there is a lot of inaccuracy and unreliability. Secondly, medical diagnoses were formulated, until the last decades of the 19th century, mostly or solely on the basis of the mere clinical examination, without almost no instrumental tool at all. Thirdly, both overemphasis and undercriticism may often influence the coeval biographers, not rarely conditioned by subjective feelings and political prejudices about the cultural atmosphere of that particular era. Lastly, but probably most importantly, many syndromes have been described and named by the authors in more "modern" times, and it seems quite artificial and at times arbitrary to "label" famous personalities of the past with more or less exotic eponyms just to enroll a new and illustrious testimonial. This behavior of the scientific community can somehow be defined as a forced "medical repéchage."

An exemplar case is the one of the Italian violinist and composer Niccolò Paganini (1782–1840), who was supposed to be affected by Marfan syndrome [1], a hereditary mesodermic dystrophy characterized by multiple physical abnormalities, including long hyperflexible hands with

* Corresponding author. Department of Neuropsychiatry, Birmingham and Solihull Mental Health NHS Foundation Trust, Barberry Building, B152FG Birmingham, United Kingdom.
E-mail address: a.cavanna@ion.ucl.ac.uk (A.E. Cavanna).

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spider-like fingers. The syndrome was first described in 1896, i.e., 56 years after the death of Paganini, but convincing evidence of this condition in the famous musician has always been lacking [2]. For instance, a recent contribution suggested that Ehlers–Danlos syndrome is more plausible than Marfan syndrome [3]. For other famous people of the past, the retrospective diagnosis is supported by more consistent evidence. This is the case with Russian writer Fiodor Dostojevskij (1821–1881), who suffered from epilepsy and was able to provide us with accurate descriptions of subjective ictal phenomenology in his novels [4,5].

The case of Gilles de la Tourette syndrome

In the light of the aforementioned motivations, Gilles de la Tourette syndrome (GTS) seems quite paradigmatic. GTS is recognized as a medical condition of unknown cause, characterized by the presence of multiple motor tics (e.g., blinking, grimacing, or jumping) and at least one vocal/phonic tic (e.g., grunting or sniffing) with onset before age 18 years [6,7]. Originally GTS was thought to be a rarity, but several studies indicate that this disorder is not rare (the prevalence has now been documented to be around one per cent of youngsters), although often underestimated, due to the complex clinical condition and the co-occurring psychiatric disorders, like attention deficit hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD) and impulse control problems [7,8]. The pathophysiology of GTS is not fully understood, although genetic epidemiology studies support the existence of susceptibility genes, and heritability patterns may become relevant to the diagnosis, especially in unclear cases. In fact, there is no diagnostic test for GTS other than expert clinical observation and careful history taking, and it should be pointed out that often clinicians erroneously believe that coprolalia (uttering of obscene words) must be present for the diagnosis to be made. A number of people may also have mild GTS, which does not prompt referral to specialists [9].

Nervous tics, hyperactivity, and obsessive–compulsive behaviors are among the most frequently portrayed aspects of modern existence. Thus, it is not surprising that GTS has progressively gained scientific recognition, theoretical revisitation and, at times, remarkable mediatic visibility. We have now the chance of reading newspaper or electronic articles, seeing movies, TV fiction serials, and even popular cartoons (quoting from *The Simpsons*: “Bart, to avoid this test, you’ve had smallpox, the bends, and that unfortunate bout of Tourette’s syndrome” [10]), where GTS is quoted and/or attributed *tout court* to a character, sometimes in a simplistic and incorrect way. With regards to famous people of the past, the retrospective diagnosis of GTS poses specific challenges. For example, while the “phenomenology” can be fairly accurately described by biographers, key elements such as the subjective “urges” which characteristically precede tic expression, the transient post-tic relief, and the variable degrees of tic suppressibility, are more difficult to prove in the absence of direct accounts from the individuals themselves [7]. It seems therefore advisable and necessary to reconsider critically the medical history, among the others, of two extraordinary “presumed” patients of the virtual “hall of fame” of GTS, i.e., the British literate Samuel Johnson and the *princeps musicae* Wolfgang Amadeus Mozart. The comparison between these two famous “patients” is also of didactic value, as shown by Kammer [11] in his original pathography article.

Samuel Johnson (Lichfield 1709–London 1784)

Of modest origins, the English literate Samuel Johnson (Fig. 1) was never able to get a doctoral degree, although later in his life, he was awarded several degrees *honoris causa*. He was described as a surly, ugly, and bad-tempered man, often tormented by moral and religious issues. He married at 26 with a poor and shrewish 50-year-old widow, and maintained in his home a quack doctor and four indigent women. He wrote all of his own entire journals, like “The Rambler” and “The Idler.” He also wrote the tragedy “Irine,” the philosophical romance “The History of Rasselas, Prince of Abyssinia,” and a number of treatises, including ”The Vanity of human wishes” and “Lives of the Poets.” His most

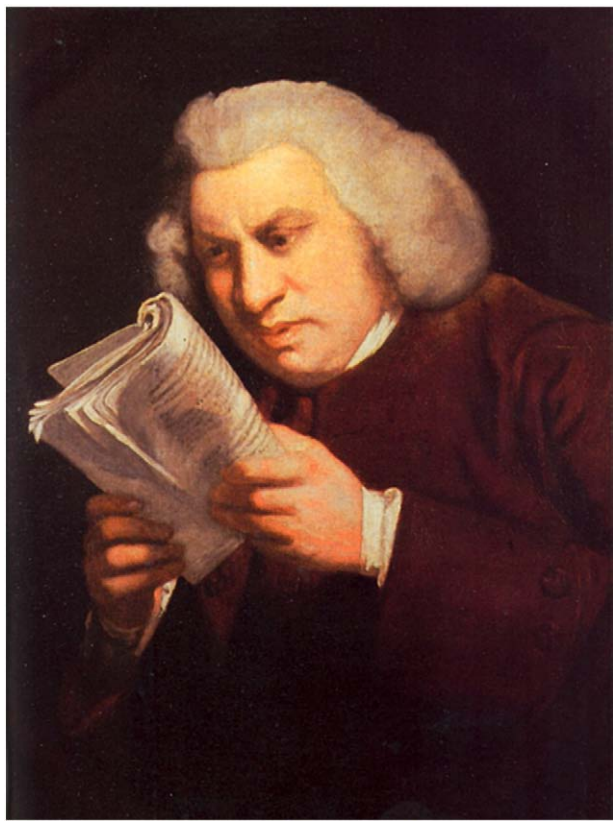

Fig. 1. Samuel Johnson (1709–1784).