

***Original article*****□ Tourette syndrome: a bizarre history**

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**SUMMARY:** In 1885 the French neurologist Gilles de la Tourette (1857-1904) published an article in the journal "Archives de neurologie" describing nine cases of a bizarre malady that he had encountered in his medical practice. These patients presented with multiple tics and involuntary movements, particularly of the face and upper limbs, accompanied by brief, violent and irrepressible verbal outbursts, usually insults and expletives. To the six cases he had examined in person, he added three already described by other physicians. One of the latter was the case of the Marquise de Dampierre, recently deceased at the age of 85, whose symptoms had previously been described, in 1825, by Jean Marc Gaspard Itard (1775-1838). Her clinical condition, masterfully described by Gilles de la Tourette, became an emblematic example of the "maladie des tic multiples". The investigative method he had learned under Jean Martin Charcot (1825-1896), and the epistemological evolution that the approach to "nervous and mental disease" was undergoing at the end of the 19th century, were the ideal ground upon which Gilles de la Tourette was able to construct the nosographical framework of the new syndrome which still today bears his name. By examining the contributions of the Authors who described the Marquise's symptoms, we can see how the afflicted woman was co-opted as proof positive of the various explanations for mental illness that were proffered throughout history.

**KEY WORDS:** History of Medicine, Tics, Tourette syndrome.

**□ INTRODUCTION**

What we today know as Tourette's syndrome represents a fascinating episode in the long and meandering story of tic classification<sup>(10)</sup>. Named after the man who described the first patient series, calling it "*la maladie des tics multiples*", Tourette's features a startling and variable array of symptoms comprising motor, phonic and behavioural tics. Its peculiar clinical manifestation reflects the complexity of its pathology - few other human conditions display such an intricate mesh of neurobiological, anatomical and functional aspects plaguing both the mind (both neurological and mental) and the body - which can affect manifest in many and varied ways within a single individual.

**□ GILLES DE LA TOURETTE'S ARTICLE**

In 1885, the journal *Archives de neurologie* published an article by a young French neurologist, Gilles de la Tourette (1857-1904), a pupil of the great Jean Martin Charcot (1825-1896), describing nine cases of a bizarre malady that he happened to have encountered during his medical practice<sup>(5)</sup>. These patients all presented with multiple tics and involuntary movements, particularly of the face and upper arms, accompanied by brief, violent and irrepressible verbal outbursts, usually insults and expletives. de la Tourette had observed six of these cases in person, while the remaining three had been referred to him by other physicians.

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**Figure 1.** Jean Marc Gaspard Itard (1775–1838), who, in 1825, was the first to describe the strange behaviour of the Marquise de Dampierre.

One of the latter was a certain woman of noble origins, the Marquise de Dampierre, who had passed away but a few months earlier, in August 1884, at the grand old age of 85. The Parisian newspapers of the day were full of the news of the death of the hapless Marquise, who had been the talk of high society during her lifetime due to her bizarre behaviour, which had been the source of considerable amazement and gossip for well over half a century. Indeed, the Marquise was a ticqueur, and began to display the first symptoms of her singular disturbance when she was only 7 years old - tics and violent movements of the mouth, face, neck and arms. Several years later, these were joined by short but uncontrollable cries and guttural barks, and explosive episodes of obscenity and swearing quickly followed. This unfortunate condition forced the poor lady to live confined to her residence “as if in a prison” until death relieved her of her afflictions.

Her story had originally been brought to public attention by Jean Marc Gaspard Itard (1775-1838), head physician of the Royal Institute for Deaf-Mutes in Paris, who described her condition in an article published in the 1825 *Archives générales de médecine*<sup>(6)</sup>.

“In the middle of a conversation in which she was particularly interested,” wrote Itard, “she suddenly and without warning interrupts what she is saying or listening to with a bizarre cry, pronouncing words that are even more extraordinary and that contrast deplorably with her intelligence and fine manners. These words are generally vulgar imprecations, obscene epithets and, no less embarrassing for her and her audience, an extremely crude expression formulating an opinion regarding some member of the assembled company.”

“The more disgusted she is by the vulgarity of these expressions,” Itard went on to explain, “the more she is tormented by the fear of pronouncing them again, and this worry is precisely what drives the words onto the tip of her tongue, where she is no longer able to prevent their escape.”<sup>(6)</sup>

Sixty years later, the young Gilles de la Tourette selected the Marquise as a kind of poster girl - an emblematic case of the *maladie des tics*. His description of the clinical presentation of this disorder was so precise that it is still - with a few small adjustments - valid today. It is therefore no small wonder that he remains forever associated with the “syndrome of Gilles de la Tourette”, as it was baptized several years later by the young neurologist’s mentor, Jean Martin Charcot.

However, although Gilles de la Tourette identified and masterfully described several cases that could be traced to a single syndrome, the malady that took his name had been observed before. Indeed, in the century before, Louis de Bourbon, Prince of Condé (1621-1686) and noble of the court of Louis XIV of France, known as the Sun King, was known to try and stifle his involuntary outbursts by stuffing his mouth with stones. Even further back in time, the *Malleus Maleficarum* - the inquisitors’ ‘bible’ and handbook for witch- and demon-hunters everywhere across Europe - written in 1486 by J. Sprenger and H. Kramer, contains a singular description of the disease. Its Authors refer an anecdote regarding the case of a Bohemian farmer, recounted by one of the inquisitors in the time of Pope Pius II. This farmer went to Rome to see his only son - a priest - in order that the latter liberate him from the demonic possession that he deemed himself to be afflicted with. Indeed, when he passed before any church, the devil would gain possession of him, and through his tongue begin to utter blasphemous words. When asked whether he was unable to refrain from pronouncing such blasphemies, the farmer replied, “I cannot in any way

help myself. Even if I use all my limbs, my neck, my tongue - all of my strength, in other words - I am unable to stop myself, and I almost feel pleasure in talking and exclaiming that which I say. I can feel that it is I pronouncing the words, but I am completely unable to prevent them, and when I try to help myself through prayer, the devil attacks me with greater force, escaping through my tongue and making me say things that I would not about the worst lustful vices". In the end, a bishop took pity on him and apparently managed to obtain God's grace through prayer, exorcism and subjecting him to a diet of bread and water for fifty days, upon which the farmer was sent home "completely cured" and free of possession<sup>(7,8)</sup>. It is not difficult to see how such a bizarre syndrome, characterized by brusque involuntary movements and incongruous, often scurrilous, vocalizations should be mistaken by the medieval mind for demonic possession. The Bohemian farmer mentioned in the *Malleus Maleficarum* was fortunate enough to escape from the inquisitorial flames, apparently cured by a successful exorcism, but it is likely that many more such persons afflicted by Tourette's met a far less felicitous end.

#### □ THE CASE OF THE MARQUISE DE DAMPIERRE, AND ITARD'S INTERPRETATION OF THE SAME

The story of the Marquise of Dampierre provides an interesting and effective model through which to introduce the reader to the history and natural evolution of this disease. At the same time, it sheds light on how interpretations of such a complex syndrome have been strongly influenced by the prevailing ideas of the various ages regarding how the brain works and the mind expresses itself.

The account of Itard's article furnished by Gilles de la Tourette is incomplete. However, Itard himself only described the case when the patient was 26 years old - 18 years after the onset of symptoms - and neither he nor de la Tourette provide any information regarding the progression of the disease from 1825 (when Itard's article was published) to 1884 (when the Marquise left this mortal coil). What is more, neither Itard nor de la Tourette, still less Charcot, had occasion to examine the patient while she was still among us.

Thus the reconstruction of her clinical case has largely been made through indirect observations provided by third parties, rather than direct examination by a trained physician, a fact which reveals the powerful

role of accounts and literary reconstruction in the identification of clinical syndromes and emblematic cases. The history of Tourette's syndrome is both a chapter in the medical history of a disease, but also a striking example illustration of how the creation and interpretation of the clinical history of particular cases comes about.

Underlying the history of the strange case of the Marquise is the eternal tussle between free will and biological reductionism, i.e., the mind-body problem. The Marquise's symptoms were the subject of much discussion concerning the link between the functioning of the brain and the existence of the "mind", even before Gilles de la Tourette described her case in 1885. For Itard, she was merely the latest in a series of patients presenting with involuntary disturbances of movement and language, but who otherwise seemed to be in complete control of their faculties. His series comprised 7 men and 3 women, all of whom manifested bizarre behaviour, which would doubtless have been ascribed to possession by demon in the preceding century. All of a sudden, without any apparent



**Figure 3.** Title page of an edition of the *Malleus Maleficarum* dated 1520. The Latin title is generally translated into English as "The hammer of witches which destroyeth witches and their heresy as with a two-edged sword".

cause, these individuals would be compelled to run, jump, cry out or swear, and then, just as rapidly, their symptoms would disappear. It was as if, wrote Itard, “the brain was suddenly stripped of its *control* over the muscular apparatus [...], which, instead of ceding, as one sees in ordinary convulsions, with alternating increases and reductions in contractions, commences to execute more or less regular movements, [performing] several functions [...] as if they were under the exclusive influence of will”<sup>(6)</sup>.

Itard’s understanding of the causes of these behaviours was influenced by the theories of the French philosopher Etienne de Condillac (1715-1780), who maintained that the acquisition of consciousness depended on the stimulation of the senses. Take, for example, Victor, the “crazy child”, or the deaf-mutes in his care - it was Itard’s conviction that people unable to speak since birth had experienced an emotional or social trauma so great that it had arrested the physiological development of their brains. Itard believed that such an impediment to growth (hypoplasia) of the cerebral cortex could be cured, or at least partially overcome, by careful and persistent re-education. In his view, the development of language required reinforcement through experience and sensations; likewise human will. A weak will, such as that he ascribed to Tourette’s patients, could be strengthened through training, for instance by increasing the state of shame that their incongruous outbursts provoked. This form of therapy, or *moral treatment*, was based on the belief that good health depended on an adequately balanced combination of diet, atmosphere, climate, work and lifestyle. Disease (a category that, for Itard, included brain lesions) was the result of an imbalance of these factors. Thus, in a rather oppressive society, the incongruous behaviour of “unbalanced” people, self-discipline, rather than external authority, was the key to moral re-education, the purpose being to create “balanced” individuals, better able to fit in with normal society.

Although Itard did admit that some behavioural alterations were caused by irreparable physical damage to the nervous system, he selected the case of the Marquise de Dampierre to demonstrate that her fellow sufferers of “cerebral irritation” could benefit from his moral treatment. Unlike his seven male patients, whom he believed had suffered irreversible damage to the brain, Itard felt that his three female patients were instead victims of domestic unhappiness - the underlying cause of their altered, or “unbalanced”, nervous system. Such “cerebral irritation” weakened

their will, in turn resulting in the motor tics and involuntary vocalizations that afflicted them. Itard was therefore convinced that the condition of his female patients could be worsened or improved by modifications to the role of women, wives and mothers in society. In other words, their condition was caused by their experience interfering with the natural development of their feminine sensibilities.

This interpretation, outlandish as it may seem today, was in line with the medical thought of Itard’s contemporaries, who attributed the high incidence of maladies and complaints among women to the alienation determined by their place in a patriarchal society. Hence, with the right intervention, this arrested female development, the cause of so many ills, could be halted, or even reversed.

Itard connected the tic manifestations of his first patient, characterized by a “sudden involuntary movement” during which she “hit herself in the stomach [...] very vigorously”, to her attempted suicide, “after two years in a profoundly unhappy marriage, afflicted by pains and jealous torments”<sup>(6)</sup>. Itard never revealed the environmental privations behind the weakened will of the Marquise de Dampierre, but he did assert that her suffering would have been diminished had she adapted to the role of wife and mother. He explained that although she married “happily”, “the marriage, instead of strengthening and completing her cure as hoped, very quickly led to the appearance of her malady”. But, in his eyes, this was because the “Marquise de Dampierre [...], being childless, was deprived of the favourable benefits [-] physical and moral [- that] motherhood would have ordinarily have provided”.

For Itard, the Marquise’ symptoms were the best demonstration that the mysterious “voluntary” behaviours that she displayed were a kind of rebellion against a subjugated will. According to Itard, the faculty of will was not localized in any specific area of the brain, but, like speaking or comprehension, was a function of the brain as a whole. Looking at things from this perspective, he maintained that the underdeveloped will of the Marquise was no different to the language deficit of the “crazy child” Victor - both could be countered by moral treatment.

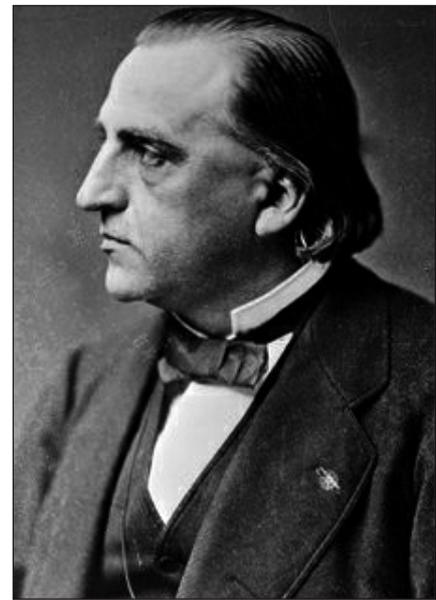
Within the concept of health as a balance, intrinsically linked to Descartes’ mechanical rationalism (the body-machine becomes ill due to a ‘mechanical’ failure, and can therefore only be repaired by repairing the fault), the brain was considered, in essence, a container of organic, physical, mental and moral

functions and faculties. Behavioural disorders were seen as expressions of an imbalance in said functions and faculties brought about by damage or irritation to the brain, and a proper balance could only be restored through external re-education, mediated by the senses - the goal of moral treatment.

#### □ THE MARQUISE AS A METAPHOR

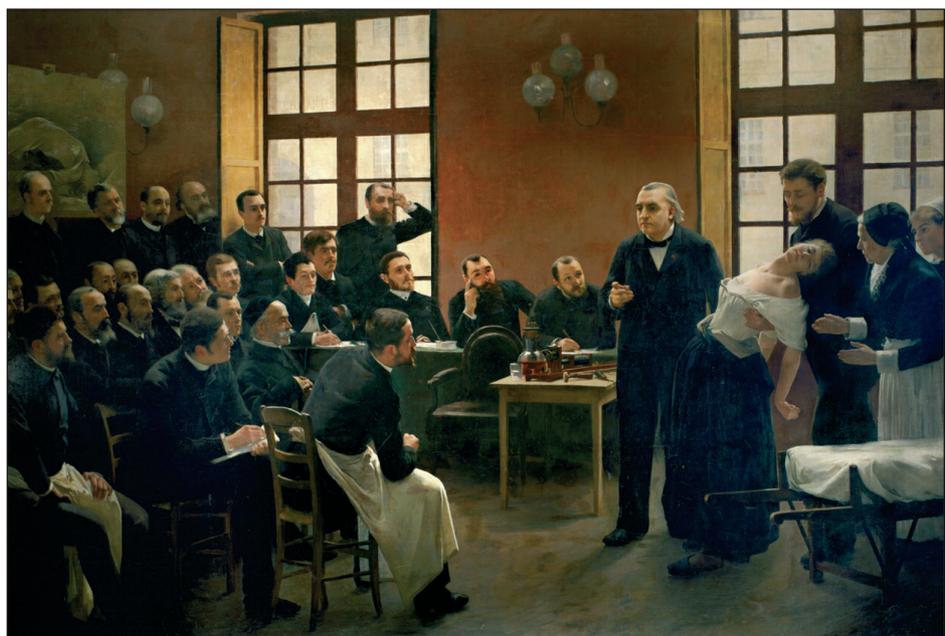
From the mid 1800s onwards, the bizarre behaviour of the Marquise became widely known in Parisian aristocratic circles and beyond. A great many physicians, none of whom had examined the patient in person, or even conducted an interview with her, mentioned her name as a prime example of the validity of their theories on the subject of a vast range of behavioural disorders that spanned from “maladies of will” to “variable palsies”. After Itard came Ernest Billot, who, in his 1847 book *Maladies de la volonté*, described the Marquise as “well known in one of the Parisian quarters, afflicted for many years by a type of palsy that struck the intelligence and the nucleus of discourse,” - to him a clear example of how a diseased will could cause an increase in the expression of thoughts that would normally be suppressed. Influenced by the eclectic philosopher Victor Cousin (1792-1867) Billot, unlike Itard, interpreted “will” as an organ or “faculty” of the mind which, when diseased, could no longer be modified by environmental

**Figure 2.** Jean Martin Charcot (1825-1896), father of modern neurology and mentor to Gilles de la Tourette.



interventions by re-education of the senses. Billot dismissed Itard’s theory that educating and curing the Marquise of her embarrassment would have cured her of her symptoms, because, in his opinion, it was her will itself that was (irreparably) damaged. According to Billot, the Marquise had suffered “a *lesion* to the association of ideas, memory, reminiscence and imagination”<sup>(1)</sup>. The refined education and undisputed intelligence of the Marquise were insufficient to enable her to resist her secret urges, which drove her to emit

**Figure 3.** Jean-Martin Charcot presents a case of hysteria to his patients. From “A clinical lesson at the Salpêtrière” tableau by Pierre-André Brouillet (1890) (*Museum of the History of Medicine, Descartes University, Paris*). Georges A.E.B. Gilles de la Tourette, whin a white apron, is sitting in front of Charcot<sup>(4)</sup>.



scatological utterances and behave strangely in public. His opinion was that the difference between the Marquise and ‘normal’ people was the fact that “we have the power not to express all the ideas that come into our heads; [we have] the intelligence to make choices and express only ideas whose suitability has been assessed”. This power “of the intellect of expression has been altered in this woman”, and, as a result, her “will is subjugated by other forces”, whether contrary to the Marquise’s wishes or not.

Three years after the publication of Billot’s conclusions, David Dieder Roth re-published Itard’s description of the Marquise de Dampierre in his own book *Histoire de la muscolation irresistible ou de la chorée anormale*. He grouped her case with six others already in the literature, including that of a certain Mademoiselle de C., a patient of Itard’s whom he categorised as suffering from “muscular tics of speech and the larynx”. According to Roth, what these seven cases had in common was an inability to control certain sounds, in particular baying and grunts. He rejected both Itard and Billot’s interpretations, and argued that vocal tics were identifiable as an anomalous and uncontrollable palsy of physiological (muscular) origin. He interpreted Itard’s description of the Marquise’s behaviour as a demonstration of the organic aetiology of many of her symptoms, which he associated with choreas.

In 1851, the Parisian doctor C.M.S. Sandras compared the case of the Marquise with Itard’s disquisition on the subject of a man who was unable to refrain from walking, classifying both as “*corées partielles*”. Although Sandras, like Roth, put a more organic slant on the Marquise’s affliction, it is interesting to note that all four of her would-be diagnosticians made use of her symptoms for their own ends, as an example of a wider problem, often without reference to the observations and conclusions of the preceding Authors. In this way, the Marquise’s malady became a *medical metaphor*, ripe for service in support of a variety of different hypothesis and theories.

Itard used her as evidence of a difference between *cerebral irritations*, which could be cured by moral treatments, and *cerebral lesions*, which would benefit from more traditional medical interventions. For Billot, the problem was a deficit of will, while Roth took her case as demonstration of muscular pathology - a partial form of palsy, different from Sydenham’s chorea. In Sandras’ opinion, on the other hand, the Marquise was a shining example of the variability of chorea.

The final ‘appropriation’ of the Marquise de Dampierre’s symptoms prior to the publication of Gilles de la Tourette’s seminal 1885 article can be found in the work *Les maladies de la volonté* (1883) by Théodule Ribot (1839-1916). Ribot was the main proponent of the psychological theory of degeneration. This theory linked hereditary factors, in turn associated with poor moral behaviour, to a large number of disorders and social ills (from mental retardation and depression to deprivation and sterility). According to Ribot, ‘bad habits’ such as alcoholism, poor diet, and immoral behaviour had a destructive impact on the nervous system, and such cerebral alterations were passed on to the following generations.

“By adopting a hereditary approach,” wrote the psychiatry historian Ian Dowbiggin on the subject, “psychiatrists could base their diagnoses on an integrated, biological relationship between body and mind, without appearing to be less medical for recording psychological symptoms”<sup>(3)</sup>. One can almost see them busily constructing elaborate family trees to explain the behavioural alterations and disorders of their patients.

The degeneration theory of Ribot distinguished between two categories of the so-called “deficits of will”. The first, based on a revisitation of Billot’s idea, comprised those afflicted by “an absence or lessening of will”, and the second, inspired by the theories of Itard, those who “had a subjugated will that only re-emerged with the aid of external assistance”. Agreeing with Billot, he classed the Marquise as belonging to the former group, as, “from the point of view of physiology and psychology, the human being who is in these conditions is comparable with an animal that has been decapitated and deprived of its cerebral lobes”<sup>(9)</sup>.

## □ THE ROLE OF GILLES DE LA TOURETTE

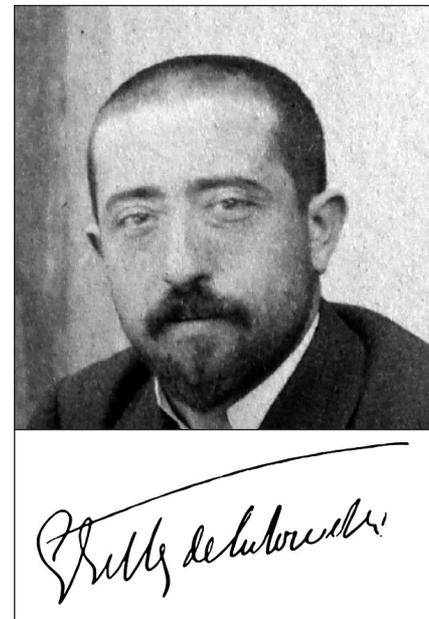
Despite the fact that Gilles de la Tourette appropriated Itard’s description of the Marquise - his primary prototype - he subscribed to Ribot’s degenerationist view of her behaviour. “What can be said with regard to the more internal *nature* of the affliction,” Tourette wondered in the concluding part of his 1885 treatise, “in the absence of any anatomopathological exhibit?”. He found his answer in Ribot’s *Les maladies de la volonté*, tracing an

inevitable evolution that began in infancy with involuntary motor and phonic tics, and which would eventually lead to unrestrainable episodes of cursing. According to de la Tourette there was no hope “of a cure”, because “once a ticqueur, always a ticqueur”. However, this assertion was not based on any clinical experience, it only sprang from the literature on the case of the Marquise de Dampierre, a woman that neither he nor Charcot had ever examined. Despite his phrase stating “the famous Marquise [was] one of Charcot’s patients”, de la Tourette later goes on to say, “In 1825 Itard published a study [...] which [...] is extremely definitive and even more interesting because the afflicted woman, our subject, lived until 1884 and was *seen* by Prof. Charcot, who retroactively verified the diagnosis,” followed by, a few lines below, “professor Charcot frequently *saw* this afflicted person, who, in her advanced age, still manifested her awkwardness, and continued, despite herself, to use bawdy words even in public places, as witnessed by professor Charcot”<sup>(5)</sup>. The fact that Charcot only *saw* the Marquise’s behaviour, rather than having occasion to examine her in a medical setting, is backed up by his own words. He mentioned her case five times during his famous public lectures, each time recounting a slightly different story - as per the oratorical fashion of the day - but never once stated that he had had any direct contact with her, or taken her on as a patient. Charcot was even more explicit in December 1887, when he stated that, “there was in Parisian high society one who was known throughout the most aristocratic part of society because she uttered vulgar words. *I have never had the honour of meeting her*; I recognized her one day when she was ascending the stairs at a salon, and I was surprised to hear her suddenly exclaim the sainted name of our Lord”<sup>(2)</sup>.

Thus, Gilles de la Tourette’s assertion that “Charcot saw this afflicted person frequently” is revealed to be nothing more than a rhetorical expedient, with the aim of justifying the Marquise’s case as the most convenient example of the type, and his description was not, in fact, based on any real clinical observations or insider knowledge. Claiming that the Marquise was under Charcot’s care conferred greater ‘scientific’ authority on what was, in fact, merely anecdotal evidence.

There are numerous references to the Marquise’s case in the literature, but it will not escape an astute reader that each clinical aspect of it can be referred back to Itard’s initial, and unique, observations. The only

**Figure 3.** Georges Albert Édouard Brutus Gilles de la Tourette (1857-1904), author of the scientific article which in 1884 systematically described for the first time the symptoms of the malady that today bears his name, and his signature (*Charcot Library at the Salpêtrière hospital, Université Pierre et Marie Curie, Paris*)



sources for the claim that she was still suffering from her unfortunate condition at her demise were the death notices, not the record of any clinical examination conducted in her later adulthood.

Charcot, who habitually used his pupils to elaborate his new theories, charged Gilles de la Tourette to deal with these issues, presumably due to his clinical dealings with other patients afflicted by tics, and because of his a wider interest in similar phenomena. He was “fascinated” by the relationship between ticcing disorders and the reports of uncontrollable jumping and jerking coming out of Malaysia, Siberia and Maine. By all accounts, these bizarre behaviours were often accompanied by imitations, blasphemy and explicit sexual gestures and manifestations. It was precisely by considering these behaviours together with the clinical presentations of ticqueurs treated at Paris’ Salpêtrière hospital that Gilles de la Tourette, prompted and heavily influenced by his mentor, came to describe the “*maladie des tics multiples*”. The distinct *habitus mentale* that Charcot’s school imprinted on attendees of his lessons and clinic, united with the particular epistemological moment that the approach to “nervous and mental maladies” was going through towards the end of the 19th century, made fertile ground for the germination of the nosographical characterization of the new syndrome.

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