

# CHARCOT'S CONTRIBUTION TO THE STUDY OF TOURETTE'S SYNDROME

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**Abstract** – We review the history of Tourette syndrome, emphasizing the contribution of Jean-Martin Charcot.

**KEY WORDS:** Tourette syndrome, Gilles de la Tourette, Jean-Martin Charcot, tics, coprolalia.

## A Contribuição de Charcot para o estudo da síndrome de Tourette

**Resumo** – Revisamos a história da síndrome de Tourette, com ênfase a contribuição de Jean-Martin Charcot.

**PALAVRAS-CHAVE:** Síndrome de Tourette, Gilles de la Tourette, Jean-Martin Charcot, tiques, coprolalia.

Tourette's syndrome (TS) is a chronic neuropsychiatric disorder characterized by multiple motor tics associated with the presence of one or more phonic tics, with waxing and waning course and severity<sup>1,2</sup>. The estimated prevalence of the disease ranges from 1 to 10 per 1000 children and adolescents, being associated with various comorbidities such as attention deficit and obsessive-compulsive disorders<sup>1,2</sup>. These comorbidities have been reported in various series of Brazilian patients in the literature<sup>3-5</sup>. It is believed nowadays that TS has significant genetic determinant, and several candidate genes have been described. In addition, countless neuroimaging, neurophysiological and even neuropathological studies have characterized abnormalities of the corticostriatal-thalamocortical circuit, including recent evidences of dopaminergic neurochemical abnormalities of the prefrontal region<sup>1</sup>.

To the 19<sup>th</sup> century French neurologist Gilles de la Tourette (1857–1904), one of the disciples of professor Jean-Martin Charcot (regarded as the father of French neurology and one of the world's pioneers of neurology), it is attributed to have made a major contribution for the understanding of TS by characterizing the disease and distinguishing it from other neurological disorders<sup>6-9</sup>.

We analyze some historical aspects of TS, particularly Charcot's contribution to the definition of this syndrome.

## TOURETTE'S SYNDROME BEFORE ITS CLASSICAL DESCRIPTION

The first report of patients with TS is speculated to

have been presented to the public in the book "Malleus maleficarum", or "Witch's hammer", by Jakob Sprenger and Heinrich Kraemer, published in 1489. The book includes a description of a priest who had motor and vocal tics, which at that time was believed to be related to possession by the devil, witchcraft and exorcism, as well as with the inquisition<sup>10</sup>.

Other important contributions to the description of TS include that by Armand Trousseau (1801–1867), a famous 19<sup>th</sup> century French physician. In 1861 Trousseau published a description of several cases of patients with motor and vocal tics in his book on clinical medicine<sup>10</sup>.

In 1825, the head clinician at the traditional "L'Institution Royale des sourds-muets", Jean-Marc Gaspard Itard (1775–1838), published in Paris an article entitled "Mémoire sur quelques fonctions involontaires des appareils de la locomotion, de la préhension et de la voix"<sup>11</sup>. In this article Itard describes some cases (a total of 10, 7 of which were men), including that of a 26-year-old female patient known as Madame de Dampierre, who was in fact a member of the Parisian aristocracy<sup>11</sup>. Madame de Dampierre suffered from a disorder that was characterized by involuntary movements associated with sudden vocalizations and obscenities; the condition, rarely described at the time, was later defined as coprolalia. These episodes of coprolalia were obviously in stark contrast to the lady's background, intellect, and refined manners<sup>11</sup>.

Seventy years after Itard exhaustive description of the case of Madame de Dampierre, it fell to Gilles de la Tourette the task to characterize this condition in which he

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included the case of this French aristocrat to illustrate what would later come to be known as TS.

Gilles de la Tourette was at that time 28 years old and staff neurologist in professor Jean-Martin Charcot's service at the "Hôpital de la Salpêtrière" (derived from the saltpeter, the principal ingredient of gunpowder) in Paris. This hospital was previously the Louis XIII's gunpowder arsenal on the Left Bank of Seine and was built in the 16<sup>th</sup> century. In 1656, it was used as asylum and in 1680, with the construction of new buildings, it became the largest asylum in Europe. In the 18<sup>th</sup> century, it was transformed into a hospital with an officially appointed staff<sup>11-13</sup>.

The literature is in fact contradictory regarding the issue of whether the lady in question, who was subsequently awarded the title of Marquise of Dampierre, was also examined by Charcot. However, the scientific evidence suggests that in his lectures Charcot referred to having met this lady in social events and witnessed the symptoms of coprolalia but not in fact having examined her<sup>10-12</sup>. In his famous "Leçons du mardi a la Salpêtrière", Charcot also mentioned having met the Marquise of Dampierre in public and hearing her using the expression "Sacré nom de Dieu", and there are references of other obscenities uttered involuntarily by the marquise, of which the most common were "merde" and "foutu cochon" (which translates literally as filthy pig but the truer colloquial meaning is "fucking pig")<sup>10-12</sup>.

### GILLES DE LA TOURETTE

Gilles de la Tourette, whose complete name was George Albert Édouard Brutus Gilles de la Tourette (Fig 1), was born on October 30<sup>th</sup>, 1857, in the small town of Saint-Gervais-les-Trois-Clochers, in the district of Châtellerault near the city of Loudun, France, and died on May 22<sup>nd</sup>, 1904, in Lausanne, Switzerland<sup>9-12,14</sup>.

From a family of doctors, Gilles de la Tourette graduated from medical school in Poitiers in western France in 1881, and then moved to Paris to continue his medical training<sup>9,12,14</sup>. In the course of his career, three professors had particularly influence on him, namely: Théophraste Renaudot, a doctor in the town of Loudun; Paul Camille Hyppolyte Brouardel, professor of forensic medicine in Paris; and last, but most importantly, Professor Jean-Martin Charcot<sup>9,12</sup>. Gilles de la Tourette started his activities as a clinician at the Salpêtrière hospital in 1884 under the supervision of Charcot. He was later promoted to the post of Charcot's registrar and finally to head of clinic<sup>7,9,12,14</sup>.

Gilles de la Tourette's tremendous scientific interest and dynamic, restless, talkative behavior was the target of criticism by his colleagues, who accused him of lacking discipline and being impulsive, inattentive, and even hypomaniac<sup>9,12,14</sup>. One of his most hostile critics was Léon Daudet, a personal friend of Charcot, who defined Gilles de la To-

urette as "ugly like a Papuan idol with bundles of hair stuck on it. He was neither good nor bad, neither studious nor lazy, neither intelligent nor foolish, and he vacillated with his confused and malicious mind between a multitude of faults without lingering. He had a husky and worn-out voice, abrupt gestures, and a strange gait. He passed for an eccentric starting an interesting subject but leaving it for another, disconcerting his masters by his queer ways which got worse and became less amusing..."<sup>9</sup> Gilles de la Tourette, however, described himself as "ugly as a louse, but intelligent"<sup>9</sup>.

Gilles de la Tourette's academic career will always be remembered for his publications in the area of movement disorders and hysteria, as well as forensic medicine and hypnotism<sup>9,12,14</sup>. However, his two most important works were the articles entitled "Jumping, latah, myriachit" published in 1884, and "Étude sur une affection nerveuse caractérisée par de l'incoordination motrice, accompagnée d'écholalie et de coprolalie (jumping, latah, myriachit)" published in 1885. The last is considered the watershed in the description of TS<sup>6,9,12,14</sup>.

However, after Charcot's death in 1893 and of his own son from meningitis in the same year, Gilles de la Tourette's scientific career became very irregular, with a variety of often unfinished projects, ranging from neurology to theater. During this period he published only one article of interest, in 1899, entitled "La maladie des tics convulsifs"<sup>9,12,14</sup>.

There are quotations testifying to his precarious mental state during this disturbed period of his life, particularly by the writer Alphonse Daudet<sup>7,9,12</sup>. During an exam for medical students, Gilles de la Tourette asked one of them which disease started with bleeding in the left nostril. After hearing the student's answer – typhoid fever – Gilles de la Tourette remained silent for about 5 minutes and then said, "Typhoid fever, you are wrong!" On another occasion Gilles de la Tourette asked another student, "Who are the three greatest doctors of the 19<sup>th</sup> century?" The student replied, "Laennec, Duchenne de Boulogne and Charcot". Gilles de la Tourette then replied, "No, you are wrong! They were my grandfather, my father and me!"<sup>9,12</sup>.



Fig 1. Georges A.E.B. Gilles de la Tourette (1857–1904).

Gilles de la Tourette's health worsened steadily, and in 1902 he was forced to resign his clinical practice. He was admitted to a psychiatric hospital with the diagnosis of neurosyphilis and died in 1904 with advanced dementia<sup>7,9,12,14</sup>.

### THE CLASSIC DESCRIPTION OF TOURETTE'S SYNDROME

In 1884 Charcot gave a new assignment to Gilles de la Tourette, namely, to reclassify all movement disorders, or to "sort out the chaos of the choreas"<sup>7,9,12,14</sup>. In the same year Gilles de la Tourette published his first important article on three strange movement disorders, in which he discussed the cases known as "Jumping Frenchmen of Maine", Latah (described in Malaysia) and Myriachit (first described in Russia)<sup>7,9,12,14</sup>. These diseases, which are now classified as startle diseases, were considered at the time to be variants of chorea (as opposed to what was called true chorea, or Sydenham's chorea) and to have an hysterical etiology. Gilles de la Tourette described the three diseases and related them to the case of a boy who was examined at the Salpêtrière hospital by Charcot and who may have been the first case of the disease Gilles de la Tourette ever studied<sup>9,12,13</sup>.

In 1885 Gilles de la Tourette published his masterpiece, which he entitled "Étude sur une affection nerveuse caractérisée par de l'incoordination motrice, accompagnée d'écholalie et de coprolalie" with the subtitle "Jumping, Latah, Myriachit". This work characterized in great detail nine cases of patients with tics (although this term was not used by Gilles de la Tourette), of which the first one to be described was the Marquise of Dampierre, and the second a case previously described by Trousseau<sup>9,10,12-14</sup>.

In this report, which made him famous, Gilles de la Tourette gives a masterly description of the clinical characteristics of the syndrome that bears his name – involuntary movements (motor tics) and uncontrollable sounds (vocal tics) associated with echolalia, coprolalia and echokinesis (echopraxia)<sup>9,12-14</sup>. According to Gilles de la Tourette's definition, the disease began in infancy, had a waxing and waning course, and was inherited<sup>9,12-14</sup>.

After this article was published, Charcot named the disease "Gilles de la Tourette syndrome" in recognition of Gilles de la Tourette's work<sup>6,7,9,10,12-14</sup>.

However, another disciple of Charcot, Georges Guinon, who was also on close terms with him, as well as having carried out studies in this field and having extensive clinical experience with patients with tics, published two studies that directly conflicted with the ideas in Gilles de la Tourette's publication<sup>10,12-14</sup>. Although there was the possibility of conflict between the egos of Charcot's two favorite disciples, Guinon's ideas were extremely valuable as he raised important questions that had been overlooked

by Gilles de la Tourette in his classic article<sup>7,10,12-14</sup>. The first problem was the definition of lack of motor coordination in the very title of the article, as for Guinon it was not a question of lack of motor coordination but rather one of systematic involuntary movements, which were never uncoordinated. Other questions raised by Guinon related to the progression of the disease throughout the individual's life (despite fluctuations in the symptoms, Gilles de la Tourette was of the opinion that the disease was progressive and lifelong), as he had described cases in which there had been a permanent improvement. In addition, Guinon raised the possibility of other symptoms being present, such as the so-called "idées fixes", which are now considered to fall under the category of obsessive-compulsive disorders. Lastly, Guinon suggested that the name of the syndrome be changed from Gilles de la Tourette syndrome to the term previously advocated by Charcot – "Convulsive tics" or "Illness of convulsive tics". Nevertheless, Guinon also suggested that the disease was associated with hysteria, a hypothesis not shared by Charcot<sup>7,9,12,14</sup>.

### CHARCOT'S CONTRIBUTION TO THE STUDY OF TOURETTE'S SYNDROME

Professor Jean-Martin Charcot, the brilliant French neurologist considered the father of modern neurology and the first professor of diseases of the nervous system, is regarded as one of the most important researchers in the field of clinical neurology in the 19<sup>th</sup> century (Fig 2)<sup>7,8,15-17</sup>.

Charcot was born in Paris in November 29<sup>th</sup>, 1825, to a carriage builder of limited means. His father told him and his three brothers that the one with the best grades would pursue a learned profession. Jean-Martin earned the prize and chose the path of medicine<sup>18</sup>.

By the age of 23 he was qualified for a hospital internship and became intern at the Salpêtrière hospital in Paris. In 1853, he became the *Chef de Clinique* soon after defending his original doctoral thesis on the differentiation of gout from chronic rheumatism. In 1856, he was appointed "physician to the hospitals of Paris", and four years later he was promoted, becoming *Professeur Agrégé*<sup>18</sup>.

The turning point of Charcot's medical career came when he was appointed to the Salpêtrière as physician in 1862. At that time the patient population was about 5,000, of whom nearly 3,000 suffered from neurologic disease. Together with his colleague Alfred Vulpian they recorded meticulous description of their patients, classifying their neurological disorders and cataloging them into distinct types<sup>18</sup>.

Charcot's extensive contributions to neurology are exceedingly well-known, particularly those related to multiple sclerosis (or sclerosis in plaques according to Charcot's original definition), lateral amyotrophic sclerosis (or



Fig 2. Professor Jean-Martin Charcot (1825–1893).

Charcot's disease), Charcot-Marie-Tooth disease (or hereditary motor and sensory neuropathy, according to the current definition), motor ataxia, Parkinson's disease, and Tourette syndrome, among other diseases<sup>7,8,15-17</sup>.

Concerning to TS, Charcot's contribution can be broken down into a number of different elements. Firstly, it should be remembered that he often assigned tasks to his residents and assistants and that he suggested to Gilles de la Tourette for carrying out a study to establish a nosological classification of several neurological diseases, particularly movement disorders, such as tics and choreas<sup>7-10,12,14</sup>.

It should be emphasized that after giving his masterly classes on Tuesdays and Fridays, he would give all the material that he had prepared to his assistants, whose task was to publish them. This helps us understand why an article about a case of convulsive tic associated with coprolalia and echolalia that had previously been presented by Charcot was published by Melloti in an Italian journal<sup>6,7,9</sup>.

Compulsive tic syndrome, in its classic definition by Charcot, was subsequently renamed Gilles de la Tourette syndrome, or TS (as it is better known today), by Professor Charcot himself<sup>7,9,12,14</sup>.

In his traditional "Leçons du mardi à la Salpêtrière" in 1887 and 1888, Charcot described nine general neurology cases, which were translated and published in 1987 by Goetz<sup>6</sup>. Among the various cases described by Charcot, one stands out, namely, that of a 12- to 13-year-old child suffering from a disease defined as convulsive tics associated with coprolalia, which was subsequently renamed as Gilles de la Tourette syndrome<sup>6,7,9,10,12-14</sup>.

Charcot also defined tics clearly, separating them from choreas (which he divided into true choreas, such as Sydenham's chorea, and the others, most of which were considered to be of an hysterical nature)<sup>6,7,9,12</sup>.

Charcot characterized TS as being associated with the presence of obsessive-compulsive disorder (which he defined as "des idées fixes") and gave the example of a patient whom he had examined: "not being able to open the door without first turning the door knob three or four times, or saying out loud...one, two, three, four..."<sup>6,7,9,12</sup>.

Another characteristic of TS defined by Charcot was the presence of imitative behavior in patients (called echokinesis and today referred to as echopraxia) as well as echolalia, and coprolalia. With regard to coprolalia, Charcot defined this as being "a compulsion to pronounce the word used by Cambronne in Victor Hugo's account about the battle of Waterloo"<sup>6</sup>. Cambronne was an official of the Napoleonic army who could not help saying "merde" when facing the enemy. Since then it has become common practice to refer to this vulgar word as Cambronne's word, which subsequently became an example of the utterances associated with coprolalia<sup>6,7,10,11</sup>.

Charcot also suggested that TS had a hereditary determinant and defined the course of the disease as one of waxing and waning symptoms. He also differentiated the disease from other diseases known as Jumping, Latah and Myriachit, which had been incorrectly classified by Gilles de la Tourette as clinically identical to TS<sup>6,7,9,10,12</sup>.

In conclusion, we may say that TS is one of the countless neurological diseases in which nosological definition Charcot's genius contributed significantly.

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