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## **History of Neurology**

## The Marquise de Dampierre identified at last, the first described clinical case of Gilles de la Tourette syndrome



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#### ABSTRACT

Among the observations of patients suffering from abnormal movements, Jean-Gaspard Itard (1775–1838) published the case of Madame D. in 1825. It was republished in 1885 as the first clinical case characteristic of the disease described by Georges Gilles de la Tourette in the seminal article leading to his eponym, still in use today. However, the actual identity of Madame D., known throughout the 19th century as the Marquise de Dampierre, has remained a mystery, until now. The 17 July 1884 edition of the literary periodical Gil Blas provided an important lead by detailing the behavioural disturbances in society of the "Countess Picot de Dampierre". Information from diarists at that time make it possible to confirm that this patient, known for her involuntary verbal outbursts, typical of coprolalia, in salons frequented by the 19th-century Parisian aristocracy was in fact Ernestine Émilie Prondre de Guermantes, her maiden name. She was born on 22 August 1800, and her married name was Countess Picot de Dampierre. She died on 08 July 1884. This article examines the life of this woman, her disease, her identification and the connection with the Duchesse de Guermantes, heroine of La Recherche du temps perdu written by Marcel Proust.

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The term "Gilles de la Tourette syndrome" currently refers to an association of motor and phonic tics with variable psychiatric comorbidity. Prevalence is estimated at 0.4–1.0% of the general population [1]. The name of this syndrome was suggested by Jean-Martin Charcot (1825–1893), after he asked his 1884 resident, Georges Gilles de la Tourette (1857–1904), to observe a young boy hospitalised at the Clinic of Nervous System Diseases, part of La Salpêtrière hospital. At the end of the article he wrote in 1884 on "Jumping, Latah, and Myriachit", Gilles de la Tourette noted: "In the department of our Master,

Professor Charcot, there is currently a case in many ways similar to this singular state [see jumping]. The subject is a fifteen-year-old boy, intelligent, perfectly capable of reasoning, of sound constitution, who suffers from extreme hyperexcitability, specific tics, jolting movements of the head and trunk, after which he almost invariably and loudly pronounces the word of Cambronne [shit]. Moreover, if we speak in front of him, he faithfully echoes the last two or three words of the sentence just pronounced". Gilles de la Tourette added this note: "Since the writing of this article, we have had the good

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Fig. 1 – Portrait of Jean-Gaspard Itard.
With permission by the library of the French National
Institute of Deaf Children in Paris (photo by OW).

fortune to come across another subject; the corresponding observation and three others from Mr. Charcot will be described in a forthcoming text on this matter" [2].

In volume IX of Les Archives de Neurologie, on 25 January 1885, Gilles de la Tourette's "Étude sur une affection nerveuse caractérisée par de l'incoordination motrice accompagnée d'écholalie et de coprolalie (Jumping, Latah, Myriachit)" (Study of a nervous condition characterised by motor incoordination accompanied by echolalia and coprolalia (Jumping, Latah, Myriachit)) was published and recognised as the seminal article describing the eponymous syndrome [3]. Gilles de la Tourette noted: "In 1825, Itard published an observation reported in its entirety by Roth in 1850, and by Sandras in 1851. This observation, the first of those we have brought together, is extremely conclusive. What makes it even more interesting is that the patient lived until 1884 and had been seen by Professor Charcot, who checked the retrospective diagnosis". After a brief biography of Itard, we will review the various medical writings referring to this observation of Madame D., providing little-known details. In addition, the accounts of diarists, journalists, and writers of her time have enabled us to formally identify this patient and establish her aristocratic genealogy.

## 1. Jean-Gaspard Itard

Jean-Gaspard Itard (1775–1838) was born in Oraison (Southern French Alpes) on 24 April 1774 (Fig. 1). During his studies at the Riez seminary, he learned Italian and English, which was rare at that time. In 1794, he was a surgeon's assistant at the hospital in the southern city of Toulon, under the orders of Dominique Larrey (1766-1842), the chief military surgeon for the army of Italy, from whom he learned anatomy. After spending the summer of 1795 in Oraison with his friend, Gaspard Laurent Jessé Bayle (1774-1816), the two of them accompanied Larrey to Paris in 1796, to begin their medical studies. On 08 May 1798 ("21 Floréal an VI" according to the Revolutionary calendar), Itard was appointed junior military surgeon ("chirurgien de troisième classe") at the Val-de-Grâce hospital. In 1797, he met Jean-Étienne Dominique Esquirol (1772-1840), who would remain a life-long friend. Both were favourite students of Philippe Pinel (1745-1826). Also in 1797, Pinel published his Nosographie philosophique. In 1800, Itard succeeded in being appointed physician at the Institution des Sourds-Muets (institution for deaf-mutes), in a newly created position which included room and board. He was also right next to Val-de-Grâce. It was at this institution that, starting in 06 August 1800, he treated "the wild boy of Aveyron", who would make him as famous as his teacher, Pinel. He named the boy Victor (Victor of Aveyron, called "the wild boy"). For five years, Itard tried to educate and socialise Victor, but was unsuccessful. Spending half his time treating young deaf patients, and the other half with his private Parisian clientele, Itard worked relentlessly, publishing his "Traité des maladies de l'oreille et de l'audition" (Treatise of diseases of the ear and hearing) in 1821, one of the very first otolaryngology treatises. Like Pinel, he belonged to the first contingent of nominations to the French Royal Academy of Medicine, in 1822. It was around this time that his health faltered and he had to reduce his activities, finding himself in pain and bent over by what was probably ankylosing spondylitis. He died bedridden on 05 July 1838 [4].

## 2. The case of Madame D.

In the July 1825 edition of a journal founded in 1823, "Les Archives générales de Médecine", Itard published observations of abnormal movement: "When the will loses control over the muscles of expression due to a morbid stimulus provoking contractions and convulsions, these violent and repeated movements are characterised by tumultuous disorder and cannot ensure their functions" [3]. Itard believed that in these cases, the brain is suddenly "dispossessed of its control over some of the muscular systems entirely subject to it". After these probable cases of hysteria, epilepsy, or dystonia, Itard added a tenth observation, that of Madame D., then 26 years old and presenting since the age of seven "continual spasmodic contractions" and "extraordinary contortions and grimaces. The disease had continued to progress and, since the spasm had affected the organs of the voice and speech, this young woman would utter bizarre cries and words devoid of meaning, but without any disturbance of her mental faculties and never in a state of delirium". A stay in Switzerland helped her temporarily, but marriage, "rather than consolidating and completing her cure as had been hoped, caused her disease to return quite rapidly". When Itard examined her, the tics were predominant in an upper limb. He was especially impressed by her vocalisations: "A most rare phenomenon, as well as a most disagreeable hindrance, which deprives the sufferer from all the joys of society [...]. For example, in the middle of a conversation in which the subject is most interested, she suddenly, without being able to restrain herself, interrupts what she is saying or hearing with bizarre cries and words that are even more extraordinary and contrast deplorably with her intellect and her distinguished manners. These words are mostly vulgar curses, obscene epithets and, what is no less embarrassing for her and others listening, blunt expressions of judgment or unfavourable opinions on those present in society" [5]. In his analysis, Itard eliminated the aetiology of hysteria without offering any other explanation for "her clonic convulsions".

## 3. David-Didier Roth, Stanislas Sandras, Pierre Briquet, Eugène Billod, Armand Trousseau, and Charcot

While Gilles de la Tourette chose to copy this observation as the first demonstrative clinical case of the new pathology he hoped to distinguish from the various forms of chorea, he noted that his predecessor David-Didier Roth (1800–1888), a Hungarian physician and disciple of Samuel-Christian Hahnemann (1755–1843), had copied it in 1850, including it among various descriptions of abnormal movement that he named "abnormal chorea", using the word "tic" [6] in his discussions. Stanislas Sandras (1802–1856), a hospital physician at Hotel-Dieu in Paris, published his Traité pratique des Maladies Nerveuses in 1851, one of the very first of its kind and in which he listed Madame D.'s case as an example of partial chorea [7].

In 1847, Eugène Billod (1818–1886), in his Des maladies de la volonté ou étude des lésions de cette faculté dans l'aliénation mentale (Diseases of the will or study of lesions of this faculty in mental alienation) [8], spoke of "the Marquise", rather than Mme D., as if he had personally known her, using her to illustrate the involuntary inability to inhibit certain more or less conscious behaviours. He contradicted Itard concerning the potential of rehabilitation to eliminate these behaviours, arguing for the probable existence of a cerebral lesion, due to chorea, that had damaged the inhibition of certain behaviours.

In 1859, Pierre Biquet (1796–1881), in his Traité clinique et thérapeutique de l'hystérie, mentioned "a remarkable disturbance of phonation in hysterics", referring to Itard's observation which he likened to fits of involuntary laughter [9].

Armand Trousseau (1801–1867) told the story of Madame D. in 1862 in his Medical Clinic at Hôtel-Dieu hospital, considering her condition a form of epilepsy: "Everyone here has heard of a society lady who, when out in the world – at the theatre, at church, or while taking a walk – would suddenly utter the most serious insults, or the most obscene words, of which she was supposedly unaware. She was a respectable woman in every way and of great intelligence". He saw this as a case of the

sudden and irresistible impulses that epileptics seemed to experience: "The society lady I spoke of just now, and who, under the control of singular, irresistible influences would utter the strangest words, of which she was supposedly unaware, would express out loud during her attack witty ideas that, according to convention, were usually kept silent. Even though in this case the impulse was irresistible, the veracity of the response or statement could make it seem, to those unaccustomed to the phenomena of epilepsy, that these words were intentionally uttered" [10].

A young Italian, Giulio Melotti (1857-19?), who graduated from the University of Bologna in July 1882, spent two years in Paris from 1884 to 1886. Melotti transcribed one of Charcot's lessons, on tics and jumping, the "Lezione quattordicesima", published in Milan in 1887 [11]. There is no trace of this lesson in Charcot's complete works, and it is likely that Melotti, rather than transcribing an actual lesson, compiled several he had heard into one, using information from Gilles de la Tourette himself to give a detailed clinical picture of the tic disease to his Italian readers. He wrote, among other things, that Charcot had examined the Marquise de Dampierre, giving credence to this oft-repeated error. Here is what Charcot did say, on Tuesday, 13 December 1887: "In Parisian high society, there was a person of the most aristocratic circles who was known for uttering filthy words. I did not have the honour of knowing her: I met her one day on my way up the stairs from the Salon and I was surprised to hear her suddenly say 'shit' and 'fucking pig'".

## 4. Who, in fact, was the Marquise de Dampierre?

Aside from Billod, none of the authors mentioned above explicitly named this patient, although she had been known as the Marquise de Dampierre since the 19th century. As for men of letters and journalists, they had no scruples about divesting her of her anonymity. In their journal dated 27 January 1860, Jules de Goncourt (1830–1870) and Edmond de Goncourt (1822–1896) were apparently familiar with this aristocrat's singularities of speech, citing the fine interpretation of the painter Paul Gavarni, a pseudonym of Sulpice-Guillaume Chevalier (1804–1866), who seems as if he had read Billod: "As to the way in which Mme de Dampierre barks and spits out her thoughts, he is frightened by the nearness of thought with its expression, by these utterances that toss thought out before the mediation of the will: 'It is like a fearsome leakage'" [12].

In his authoritative work on the history of this pathology, A Cursing Brain?, published in 1999, Howard I. Kushner devoted an entire chapter to "Itard's Marquise de Dampierre". However, his investigation, and the ones of others, of the few descendants of the Dampierre family did not enable him to positively identify her [13].

But articles in the Parisian daily press in July 1884, after her death, provide some clues. On 17 July 1884, Gil Blas reported anecdotes on the unusual personality of the "Comtesse Picot de Dampierre": "During high mass at the Madeleine, it was not uncommon to hear such perfect barking that one might have believed a pack of hounds belonging to the Duchess of Uzès was present in the church. It was, in fact, Madame de

Dampierre, during the Elevation. This good, witty, charitable woman, well-bred by all accounts, could not keep her condition silent at certain times. Having a remarkable talent for producing artificial flowers and decorative frames, cut from cordovan leather, for example, she was awarded a firstclass gold medal at the horticulture exhibit which took place at the Champs Élysées and the Carré Marigny, under the Second Empire. The Emperor presided over the awards ceremony. When she heard her name called, Mme de Dampierre stepped up to receive her medal. But as an old Bourbon royalist, finding herself for the first time in the presence of the nephew of the 'Corsican Ogre', she totally forgot the good Catholic history of Father Lariquet that she had learned as a child. Instead of receiving a large medal from the Marquis de Bonaparte's nephew, Lieutenant-General of the Royal Armies, she saw only the usurper, the despoiler of Orléans properties ['Orléans' suggesting the king and the royalty], and in response to the Emperor's courtesy, stupefying all present, she proffered a volley of insults and barking that none of the spectators were likely to forget, even twenty-five years later" [14].

Mme de Dampierre was also known for having nicknamed Jean Antoine Brutus Menier (1795–1853), the famous founder of the Menier chocolate factory on rue Sainte-Croix de la Bretonnerie in Paris's Marais district, "Baron Cocoa".

## 5. Ernestine Émilie Prondre de Guermantes

Ernestine Émilie Prondre de Guermantes was born on 22 August 1800 ("4 Fructidor an VIII" according to the Revolutionary calendar) at the Guermantes family castle in Seine et Marne, east of Paris. The public registry indicates a name stripped of any link to the French nobility, owing to the French Revolution. On 02 March 1798, her father, Emmanuel Paulin Prondre de Guermantes (1775–1800), married Eulalie de Brisay (1779–1866). The young couple had their first daughter, Albertine Adélaïde (1799-1819), on 02 June 1799. In poor health (tuberculosis?), the father died at only 25 years old, on 03 November 1800, when his daughter Ernestine was only three months old. A widow at age 24, Eulalie de Brizay remarried Jean-Baptiste de Tholozan, Marquis de Vernon, on 21 November 1802 and had two more children, Eulalie de Tholozan (1804-1889) and Ernest René de Tholozan (1808-1890) (Fig. 2).

Ernestine was a source of worry for her mother and stepfather due to her homeliness and her behavioural problems, which Itard described as follows: "At age 7, she was overcome with convulsive contractions in the muscles of her hands and arms, especially when she was practising her writing and would abruptly move her hand away from the letters she was forming. After shifting her hand, its movements became regular and could be controlled voluntarily, until another jolt once again interrupted her work. At first this was considered a sign of vivacity and harmless mischief, but since the behaviour was repeated over and over, adults around the girl began to respond with reprimands and punishment. Soon, however, they became convinced that these movements were involuntary and convulsive, involving the shoulder, neck, and face muscles. The result was extraordinary contortions and grimaces. The disease continued to progress, and with the

spasm spreading to the organs of the voice and speech, this youngster would utter bizarre cries and words devoid of meaning, but never in a state of delirium, without any disturbance of the mental faculties". Physicians offered the hope that her condition might improve with puberty, but this did not happen. After a stay in Switzerland that seemed to bring temporary improvement, "either because of the spa treatments, or because she enjoyed her time there in the mountains", she relapsed after returning home.

Her physicians advised her mother to find her a husband, since marriage was an ancestral treatment for hysteria. Eulalie de Brisay-Tholozan selected a first cousin, Louis Augustin Picot de Dampierre (1780-1841), as a potential son-in-law capable of accepting this young woman as his wife, despite her natural defects. A brave soldier under Napoleon who was wounded at the Battle of Friedland, survived the retreat from Russia, and was made "Baron d'Empire", at 36 he aspired to a peaceful life. Thus the fortune of the Prondre de Guermantes was nothing if not attractive to him, given his mountain of debt. Since he was Ernestine's third-degree relative, he had to request a dispensation for the marriage from the Bishop of Meaux, in these terms: "The most pressing reason is this young woman's health: for several years, she has suffered from an acute nervous and convulsive disease. All the medical remedies used by the physicians in Paris, Brittany, and Switzerland that her parents took her to see have not improved her constitution. Her doctors agree that only a change in her civil status can bring an end to the accidents she experiences. In this situation, it is improbable that she should rapidly find a husband appropriate to her rank and birth, and the idea of a delay in the consummation of the union proposed to her and approved by her, or of some other obstacle, may worsen her condition" [15] (Fig. 3). Authorisation was granted and the marriage contract approved by King Louis XVIII. The marriage was celebrated at the Guermantes castle on 07 December 1817.

Ernestine's older sister died on 23 March 1819, leaving her as the sole heiress to the Guermantes fortune. Gradually she established her dominion over the castle and its occupants, being the only one to possess a master key to every door, including the gates to the grounds. The Count of Dampierre was only too willing to grant his wife this authority, a role that tied her to the castle and left him free to frequent the Parisian salons, without her and her uncontrollable speech problem (Fig. 4). He died in Paris, on 11 February 1841. Ernestine would survive him by more than forty years. She died in her Paris residence, at 191 Boulevard Saint-Germain (less than 200 m from Charcot's home) on 08 July 1884, at age 83, in the presence of her nephew, the Baron Henri Baillardel de Lareinty (1824–1901), a French senator from 1876 to 1901 (Fig. 5).

## 6. The Duchess of Guermantes and Marcel Proust

Marcel Proust (1871–1922) used the aristocratic family name "Guermantes" for one of the most important characters in À la recherche du temps perdu, the Duchess of Guermantes. One of the models for this character was the Countess Greffuhle (Élisabeth de Riquet de Caraman-Chimay 1860–1852, married

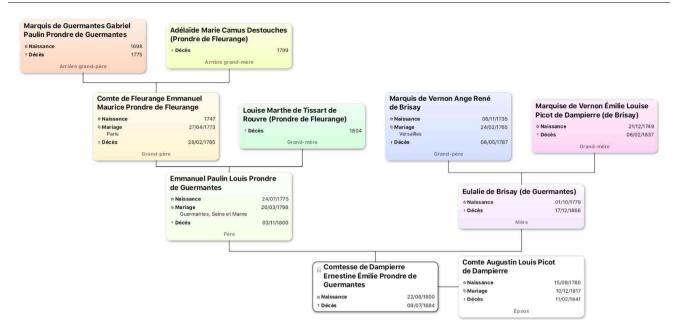


Fig. 2 – Genealogical tree of Ernestine Prondre de Guermantes. Reconstitution by OW.

decette Demoiselle: Depuin plusieurs armées elle est detat de Sante decette Demoiselle: Depuin plusieurs armées elle est attaquée D'une maladie nerveuse, convulsive, aigué de l'our les remèden de l'art employées pav les médecins de Larin, Dela Bretaque, de la suisse où ser parents l'out conduite, n'out pu améliores sa constitution. Ser médecins s'accordent à dire que le seul changement d'état peut faire disparaître les accident qu'elle éprouve. Dans cette situation, il n'est parprobable qu'elle trouve sitôt un établissement couvenable à son rang et à sa naissance: l'idée d'un retaid à la consommation de l'union qui lui a été proposée es qu'elle désire, ou d'un empêchement, peut empires d'on état.

Fig. 3 – Certificate drawn up for Ernestine's marriage. Archives of Seine et Marne.

Exercitive Duondre Guermantes

Fig. 4 – Signature of Ernestine at age 8. Archives of Seine et Marne.

to the Count Henry de Greffulhe), whose daughter married Armand de Gramont, the Duke of Guiche and a friend of Proust's. All the names of the aristocracy included on the Countess of Dampierre's death notice were names familiar to Proust, but his close friends, who bore these names, belonged to the following generation [16] (Figs. 5 and 6).

How did Proust come to hear of the Guermantes name? "If only I could delicately remove the wrappings of habit and behold this name, Guermantes, in its initial freshness...". Professor Jean-Yves Tadié showed that Proust had started by initially writing "Guarmantes" to rename the Villebon castle (west of Paris), not far from the town of Illiers. This castle, medieval in appearance with its crenelated towers, had nothing in common with the 17th-century castle where the Countess of Dampierre lived. Proust probably took little interest in the original Guermantes castle, on the other side of Paris, far from the roots of the Proust family in Illiers [17,18]. He knew, however, that his friend François de Pâris (1875–1958), part of the group of young aristocrats he was tied to at

the turn of the century, used this castle as a second home. Proust referred to it in a letter sent to him in July 1903: "What's more, visiting the country would only be possible if you had isolated buildings and I could visit you without your parents' knowing; they would naturally be furious that a stranger should enter Guermantes. I think that's the name of the place that enjoys what I cannot; that is, to see you". It was thus in 1903 that Proust used the name "Guermantes" for the first time. On 23 May 1909, he put this question to his friend Georges de Lauris (1876-1963): "Do you know whether 'Guermantes', which must have been a family name, was already in the Pâris family, or, to speak properly, whether the name of Count or Marquis de Guermantes was a title of Pâris's relatives, and whether it has entirely died out and might thus be used by a man of letters?". In December 1909, in another letter to Lauris, Proust wrote: "But if Guermantes is a name of the Puységur family, this amounts to the same thing as its being in the Pâris family [...]. I want my castle not to belong to the family whose name it bears (e.g. Dampierre belonging to the Luynes) and if the current owner exists, I want at least the name of the castle to have died out, with no ties to the family" [17,18]. It is curious that Proust used Dampierre as an example, a family name that, in our research to identify Madame D, led us down an impasse [19].

Charmed by the sound of "Guermantes", Proust was most likely entirely unaware of the Countess of Dampierre, suffering from this terrible disease, during which words and phrases were not carefully crafted but involuntarily eructed.

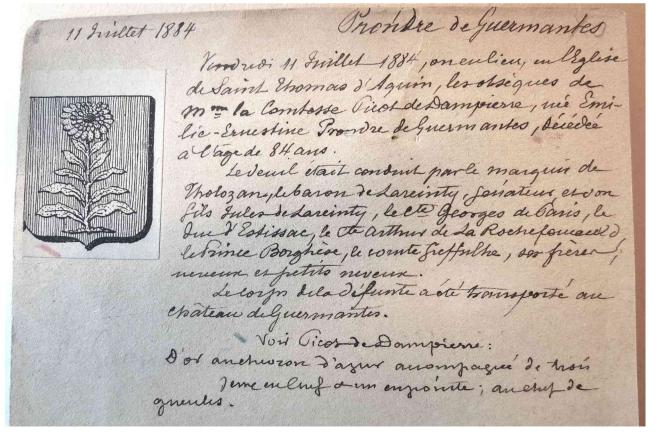


Fig. 5 – Text for Ernestine's funeral procession. Archives of Seine et Marne.

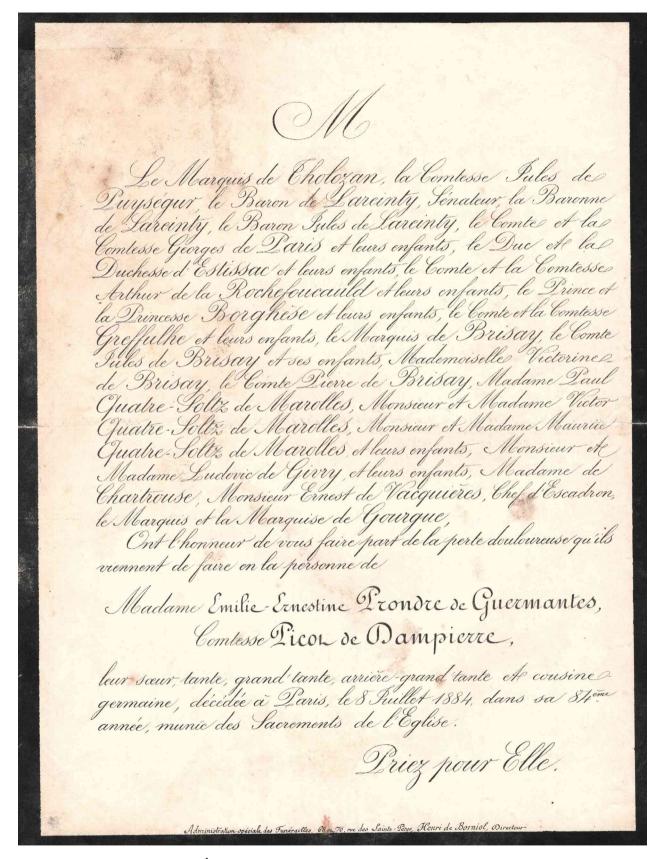


Fig. 6 – Death notice of the Countess Émilie Prondre de Guermantes. Collection of OW.

That said, both Proust's Duchess and our Countess may be honoured by Tadié's description of the Duchess of Guermantes: "She came from the beginnings of History and the summits of society".

## 7. Successful social integration despite an embarrassing disease

Ernestine's coprolalia was perceived as extreme outbursts of language in an aristocrat born during the French Revolution. Her vocabulary was at times filthy, borrowed from popular registers. Within the family, it was seen as a transgression, a comic form of the caustic judgments heard in Parisian salons. Actually, some of her reported behaviors would not be regarded as coprolalic but would correspond to what has been described as non-obscene socially inappropriate symptoms (NOSIS) [20,21]. There are significant social and emotional sequelae to living with tics and NOSIS, which can adversely affect the quality of life. However, these inconveniences do not seem to have hindered Ernestine's social life, in particular after her husband's death. She may have suffered from physical consequences such as discomfort from the repetitive movements and the stigma of her severe, violent, or socially inappropriate movements, vocalizations, or perhaps actions, not reported by contemporaries. Some ticcers have low selfesteem or consequences that may lead to poorer psychosocial functioning. It seems that this was not the case with Ernestine. Unfortunately, we did not find any personal handwritten testimony that could inform us about her psychological state and her own perception of her illness.

Anyway, it may seem surprising that in the accounts of Ernestine de Guermantes' eruptions in high-society circles, there is no mention of a medical diagnosis. This reality is confirmed in the Journal inédit of the Count Alfred of Gramont (1856–1915), a diplomat made famous by the Ems Dispatch: "This morning, General de Vacquières came for breakfast. He is very agreeable and told us many amusing stories, especially about the famous Countess of Dampierre, whom everyone used to know before her death in 1884. Deaf and always spitting, she was high-spirited and couldn't keep herself from saying what she thought out loud". It was Alfred de Gramont who related the exclamation "Cocoa! Cocoa!" cited above, as well as "Tambour! Tambour!", uttered to Madame Santerre, recalling the order of Antoine Joseph Santerre (1752-1809) for a drumroll to drown out the voice of King Louis XVI on the scaffold; and Ernestine's introduction of three priests in her salon to her guests: "Un cu..., deux cu..., trois cu...rés de village!", where "curé" means priest, but "cu" hints at "cul", slang for ass. Other exclamations seem to lay bare an inner thought. To the Baroness of Rothschild, her neighbour who was hosting her along with other guests at her castle in Ferrières, she said: "I have an entire castle; this is merely a boutique!" [22].

### 8. Conclusion

The Madame D. observed by Itard was thus not the Marquise de Dampierre, but a countess. Her real name was Ernestine Émilie Prondre de Guermantes, and she married the Count Augustin Louis Picot of Dampierre. This toponymic addition to her name resulted in the confusion that has surrounded her identity since 1825. It was journalists and writers, notably Edmond de Goncourt and Alfred de Gramont, whose accounts of Paris high society made it possible to definitively lift the veil that, since 1825, had obscured the identity of the real Countess of Dampierre, an emblematic figure of the coprolalia described by Gilles de la Tourette in 1885.

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The authors declare that they have no competing interest.

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