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History of Neurology

Jules Sottas (1866–1945) forgotten despite the eponym: “Dejerine-Sottas syndrome”

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ABSTRACT

The eponym “Dejerine Sottas” makes 21st-century neurologists think of a form of heredity peripheral neuropathy leading to amyotrophy and secondary to a mutation of one of the many genes responsible for the formation of myelin. The seminal description was the work of Sottas and Jules Dejerine (1849–1917); Dejerine was the prestigious second successor of Jean-Martin Charcot at the Clinic of Nervous System Diseases at the La Salpêtrière hospital. Jules Sottas (1866–1945) has almost been forgotten, but as a young man he was a brilliant resident under Dejerine. However, Sottas eventually gave up medicine, even though he could have had a successful career as a neurologist, to devote himself to his passion for history, especially the history of navigation. But during his time as a physician he published several original works, always supported by detailed neuro-pathological studies, the result of his very close collaboration with Dejerine at Bicêtre then at La Salpêtrière. After a brief biography of Sottas, we will analyse his neurological work and then highlight the quality of his publications on naval and maritime history, which are still relevant today.

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During the conference on Jules and Augusta Dejerine held at La Salpêtrière on 10 February 2017, on the occasion of the 100th anniversary of the death of Jules Dejerine, the name of Jules Sottas came up several times, but was unknown to many of the participants, despite their extensive knowledge of the history of neurology [1]. They were curious about the life and times of this nearly unknown figure. Sottas accomplished much in his life. For example, his name was associated with one of the major French neurologists, Dejerine, with whom he established the clinical picture for Dejerine-Sottas Syndrome, identified at the end the 19th-century when Sottas was still a resident in the Paris hospitals. Our biography will be brief due to limited resources, but we will use them to present a sketch of Sottas’s medical work. We will then look at another facet of Sottas’s activities: his work as a historian and astronomer,

which must not be overlooked even though most neurologists are unaware of it.

Jules Sottas (Fig. 1) was born on 22 May 1866 in Paris. His parents were Louise Saluce (1846–1923) and Eugène Sottas (1838–1920), a physician. Eugène Sottas created the photographic collection for Louis Brocq’s (1856–1928) department at Hôpital Broca, a collection Sottas maintained at Hôpital Saint-Louis until 1912. This “Brocq” collection that Eugène Sottas assembled includes several thousand photographs and was an important addition to the collection known as the “photographic museum of Hôpital Saint-Louis”, mainly on dermatology.

After his classical studies, Jules Sottas enrolled in the Faculté de Médecine and ranked 144th in the competitive exam for *externes* (medical students living and working in the

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Figure 1 – Jules Sottas, seated, in a white apron and wearing a forage cap. Physician at Hospital no. 31 in Deauville in May 1915. (Private collection, jmb/14910.net, with kind permission).

Paris hospitals) in 1887. His teachers were Paul Blachez (1827–1890) at Hôpital La Charité in 1888, then Léon Labbé (1832–1916) at Hôpital Beaujon in 1889. Blachez's evaluation was flattering: "Intelligent and hard-working student, good at preparing observations, will make a good resident". Labbé's evaluation was also very favourable: "Exceptional student, knowledgeable, devoted to patients". At his first resident exam in 1889, he ranked 20th [2]. Sottas married Gilberte Jamet (1875–?) on 1 February 1897. They would have five children.

Sottas began his residency in surgery in 1890 at Hôpital Tenon. There he worked with Albert Blum (1844–1914). In 1891, he was a resident under Jules Dejerine (1849–1917) at Hôpital Bicêtre and in 1892, under Georges-Sainfort Dujardin-Beaumez (1833–1895). He finished his residency under a former pupil of Jean-Martin Charcot (1825–1893): Georges Debove (1845–1920) at hôpital Andral. All of his teachers spoke highly of him. For example, Dejerine complimented him for being "an excellent resident" and added: "Very good resident, curious and hard-working" [2]. Sottas went on to dedicate his thesis to his teachers: "Learned masters who allowed me to complete my residency in their departments. I was very fortunate to learn from their vast experience and remarkable benevolence".

Early on, during his first year of residency, he presented three papers at the Société Anatomique. The first described death by internal haemorrhaging of a patient aged 58 who had oesophageal cancer that had eroded the aortic wall [3]. He concluded that without knowing it, he had taken a risk by introducing a catheter into the patient's oesophagus. During the same session, he presented the case of a 38-year old man who died following an accident. He was struck by a carriage and died of peritonitis a few days later. At the autopsy, a fracture of the pubis was found that had pierced the bladder and a small bowel loop [4]. At the end of 1890, it was Sottas who performed the anatomo-pathological examination of the surgical specimen removed by Gabriel Walther

(1855–1935), the assistant surgeon working under Blum. Sottas identified the formation of a cyst on the anterior wall of the vagina the development of which "destroyed what remained of the Wolffian structures (embryological vestiges) or Gartner's duct" [5].

Sottas continued with his presentations at the Société Anatomique during his residency under Dejerine. His presentations always included detailed data from histological examinations, undoubtedly the result of the influence of his teacher, Augusta Klumpke-Dejerine (1859–1927). Examples from 1891 include observations of bronchial dilation, a foreign body in the peritoneal cavity, and diabetes during calcific pancreatitis in a tabes patient [6]. Of his eight publications in 1892, three concerned neurological subjects. When he was a resident under Dujardin-Beaumez, "a case of multiple sclerosis during convalescence from small pox" led him to question the causal link between multiple sclerosis and a recently resolved infectious disease [7]. He referred readers to the *agrégation* thesis defended in 1882 by Louis Landouzy (1845–1917) [8]. On 23 July 1892, on behalf of Dejerine, Sottas presented an observation to the Société de biologie that involved syringomyelia, occurring at age 53, in which the asymmetric cavity led to a typical clinical picture of sensitive dissociation of only the right upper limb (Fig. 2) [9]. Sottas and Dejerine had performed the histological examination, the subject of more than half of their presentation.

A little-known fact is worth highlighting here: in October 1892, Sottas wrote an article that he had prepared with his father, Eugène Sottas, "on a case of generalised puerperal paralysis or puerperal polyneuritis". This article referred to the thesis of Augusta Dejerine-Klumpke. A tuberculous woman, treated by Sottas's father, with a number of antecedents, came down with a fever after giving birth to her sixth child. She then developed an ascending paralysis that did not affect her face, together with severe diffuse pain. They noted the disappearance of the plantar reflex, i.e. flexing of the big toe. Four years later, Joseph Babiński (1857–1932) would describe its extension in a case of damage to the pyramidal system. They rejected the possibility of Landry syndrome because of the existence of pain. This led them to opt for the diagnosis of diffuse central myelitis or polyneuritis. It is probable that Eugène Sottas drew on his son's neurological knowledge. The patient died shortly thereafter of tuberculosis without having recovered from paralysis. At the end of the article, they expressed regret, Jules especially, that an autopsy had been impossible [10].

On 4 March 1893, at the Société de Biologie, Sottas described two cases of compression of posterior roots prepared in the Dujardin-Beaumez department. Dejerine commented on the work of his former resident, with praise he hardly concealed: "In the first case of Mr. Sottas, involving the compression of the posterior roots at the level of the cauda equina, the author gave a detailed description, previously unavailable, positing that the nerve roots had been compressed. In terms of the anatomy of the spinal cord, this point is very important, because it is now clear where the posterior sacral roots terminate in the posterior tracts" [11].

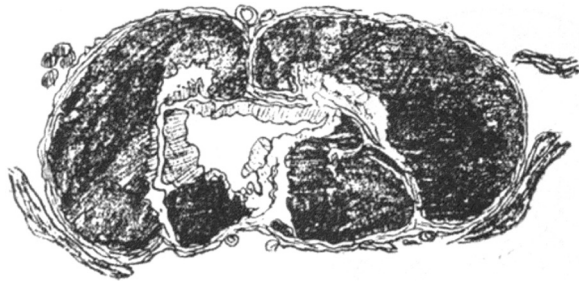


Figure 2 – One of the drawings of a syringomyelic spinal cord illustrating the 1893 article by Dejerine and Sottas. (Collection of the author).

1. On interstitial, hypertrophic, and progressive neuritis in children

At the Société de Biologie, on 18 March 1893, Sottas and Dejerine read the dissertation leading to the eponym that associated their two names. Following a history of the publication of observations of progressive muscular atrophy by Albert Eulenburg (1840–1917), Nikolaus Friedreich (1825–1882), William Hammond (1828–1900), Howard Tooth (1856–1925), Charcot and Pierre Marie (1853–1940), amongst others, Sottas and Dejerine highlighted the thesis defended in 1886 by Jules Brossard (1855–1911), who would become a professor of internal pathology at the medical school in Poitiers [12]. The dramatic life of their first patient is related in detail. The description of the autopsy is now famous: “The patient died outside of the hospital and was buried, but we were allowed to perform an exhumation, which occurred on 22 January at 8 o’clock in the morning. The extreme thinness of the subject together with the extreme weather conditions (temperatures from -8 to -10° below 0 lasting several days) were such that the cadaver was perfectly conserved”. They removed the limbs, brain, and spinal cord: “What is striking is the considerable development of the nerves of the cauda equina; each of the constituent tracts had almost doubled in volume”. The histological conclusion, based on more than four pages, provided detailed information: “This is a primitive, hypertrophic, interstitial neuritis which is monotubular”.

Damage was maximal at the peripheral extremity of the nerves of the limbs, but was also found in the anterior and posterior spinal roots. In the discussion Dejerine and Sottas noted that Albert Gombault (1844–1904) and his resident Henry Mallet (1864–1918) had presented a similar observation in 1889, in which tabes was assumed to be the cause. There were significant clinical and histological similarities with the condition of their first patient [13]. Dejerine and Sottas gave this summary: “The clinical picture of the two patients combines atrophy and ataxia. This disease is generally found in families”. For them, there could be no doubt: “It is certain that the cases we describe in our study are examples of a new disease which has not yet been classified and which is clearly distinct in clinical and anatomic-pathological terms from the cases studied [by the authors cited above]”. This new disease is caused by “a lesion specific to the peripheral nerves; this is an ascending hypertrophic interstitial neuritis along the

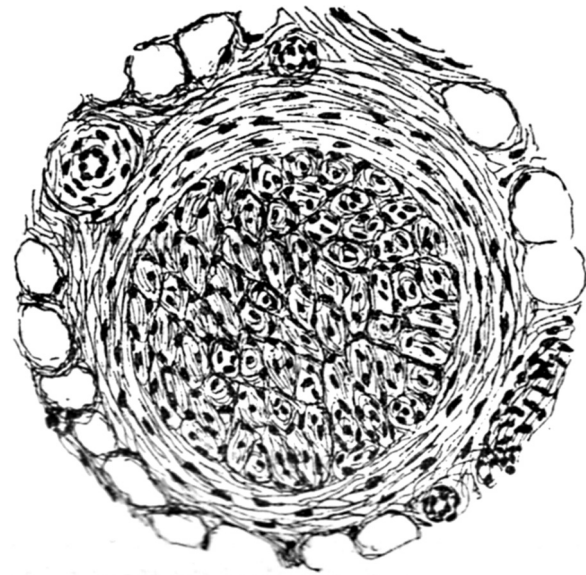


Figure 3 – One of the illustrations of the seminal article on the Dejerine-Sottas syndrome, 1893. (Collection of the author).

posterior tracts” (Fig. 3). The use of electron microscopes has since shown that nervous hypertrophy is more directly related to “swirling” Schwann cell proliferation, described as onion bulb-like, and is around the remaining axons, rather than a simple augmentation of interstitial conjunctive tissue [14]. In 1896, Dejerine published a new case [15] and a second case for which the autopsy was performed in 1906, in collaboration with André-Thomas (1867–1963) [16].

Whereas Dejerine refuted any link between the new entity and Charcot-Marie-Tooth disease, the former is currently referred to as “Hereditary motor and sensory polyneuropathy type III” or “Type 3 Charcot-Marie-Tooth disease.” With an onset in early childhood, the progression of the disease is slow until adolescence, after which it accelerates, leading to a marked handicap. The progression of this type 3 condition is more severe than that of type 1 Charcot-Marie-Tooth disease. It often leads to confinement to a wheel chair starting at age 10. Muscular atrophy, sensory deficiencies especially in the limbs, pain, scoliosis, and deformation of the hands and feet are characteristic symptoms. During the clinical examination, severe amyotrophy, ataxia, lack of tendon reflexes, late motor acquisition, and sometimes nystagmus, are found. The cause is attributed to an anomaly in the synthesis of a myelin protein secondary to mutations of genes MPZ, PMP22, PRX, EGR2, and others possible. The genetic spectrum spans more than 70 genes, likely to explain inherited peripheral neuropathies [17,18]. The heredity is autosomal dominant or recessive. Therapeutic tests using lonaprisan, curcumin or histone deacetylase 6 inhibitors are currently being conducted in animal models of peripheral neuropathies and seem to improve animal’s condition by slowing progression [18–20].

The same collection of reports from the Société de Biologie in 1893 included three other presentations by Sottas. On 15 April 1893, Sottas showed that the anatomic-pathological

lesions found in cases of syphilitic myelitis were secondary to “a vascular lesion producing softening of the spinal cord by ischemia”; this was the cause of the cases of paraplegia that he observed [21]. On 25 June 1893, Dejerine and Sottas described the histological appearance of the muscles in a case of Thomsen’s disease; very few histological studies of this pathology had been published before their study [22]. On 25 November 1893, Sottas began to study a phenomenon which, according to Waller’s law, seemed paradoxical: retrograde degeneration of the pyramidal tract in medullary syphilis. Before defending his doctoral thesis, Sottas carried out numerous studies and published his findings, often neurological, and involving in-depth anatomic-pathological investigations. This was also true of his teachers Jules and Augusta Dejerine, who pursued their work steadfastly and with devotion.

2. Syphilitic spinal paralysis

Jules Sottas defended his thesis entitled *Contribution à l'étude anatomique et clinique des paralysies spinales syphilitiques* (contribution to the anatomical and clinical study of syphilitic spinal paralysis) on 10 March 1894. The jury was presided by Debove in the presence of Alexandre Laboulbène (1825–1903), Dejerine, and d'André Chantemesse (1851–1919) [23]. Sottas introduced his work this way: “The syphilitic nature of an affection of the nervous system can only be affirmed through reasoning”. The pathogenic agent had not yet been identified and the serology was still unknown. Sottas attempted to elucidate “the types of lesions produced in the spinal cord by this affection and their consequences.” However, from the start he decided to the set limits “to be attributed to the syphilitic influences on the progression of spinal diseases”; that is, he did not take sides in the discussion between neurologists at that time with regard to the origin of syphilis, whether it was exclusive or not in cases of tabes.

After gathering observations from French and foreign literatures to support his arguments and to make clinical and histological comparisons, Sottas presented the results of his own anatomic-pathological examination, of seven syphilitic spinal cords from deceased patients, in the departments of Dejerine and Dujardin-Beaumetz. Sottas then presented the conclusion that syphilitic paraplegia resulted from “an inflammation of the vascular, lymphatic, and conjunctive element of the nervous tissue. The damage to the parenchyma is secondary to these primary lesions [...]. Inflammation begins with the vascular walls and the perivascular parts; it particularly affects the small vessels around the periphery of the spinal cord”. The spinal cord is thus affected by acute or subacute ischemia ending “either suddenly in a confined transversal softening that can be found at several locations of the spinal cord, or slowly, such that the destruction of the elements is accompanied by a process of substitutive reaction of neuralgia that gradually replaces the degenerated elements”.

Henri Lamy (1864–1909) wrote a thesis on a similar topic one year prior; his teacher Charcot was one of the members of his defence jury. His thesis associated meningitis with vascular lesions in order to explain myelitis. He noted: “the

lesions of the spinal veins are the first to appear and thereafter they generally remain more severe than those in arteries” [24]. Lamy made a report on Sottas’s thesis in the *Revue Neurologique* in 1894. He noted that “in this work, the anatomical part is covered in the most depth because of the personal views that the author puts forward in it”. He clearly described the pathophysiology proposed by Sottas, but without stating approval or refutation, and without mentioning the absence of description of the venous damage by Sottas. The *Gazette des Hôpitaux civils et militaires* invited Sottas to write columns so he could write a summary of the clinical and histo-pathological arguments he had developed in his thesis [25]. There was thus interest in his work and his innovative approach.

At the Société de Biologie, in 1895, Dejerine and Sottas gave a detailed presentation of the neuroanatomy “of the various radicular tracts [that] are located in the posterior tract and are found successively outside the lowermost tracts which they push inside”. However, the posterior tracts do not amplify their volume in a caudo-rostral way. They explained this as follows: “Each tract in its ascending path is weakened by the release of numerous collateral fibres that enter:

- the anterior horn;
- the posterior horn;
- posterior commissure;
- Clarke’s column” [26].

These data were published when the voluminous first volume of “*L’anatomie des centres nerveux*” was published in 1895 by Jules and Augusta Dejerine.

Until 1898, Sottas continued to publish clinical cases from time to time in the *Revue Neurologique* or *La Médecine Moderne*, but thereafter he stopped publishing new medical works. From this time forward, it was the Dejerine-Sottas eponym that kept Sottas’s name in medical journals, and this remains true today. Sottas did not have a hospital or university career. He worked as a “*médecin consultant*” – today this would be called consulting physician – at 47 avenue Bosquet in Paris.

3. World War I

When war broke out in 1914, Sottas was 48 years old. He was mobilized as a “*médecin major de première classe du troisième corps d’armée*”. Archival family photographs, dating from 1915 and 1916, show him taking care of the wounded at a rear-guard hospital in Deauville (Figs. 1, 4 and 5).

He was made *Chevalier de la Légion d’honneur* in April 1917, as cited in the “*ordre du service de santé du 32^e CH n°98*” of 27 May 1917. He was then made an *Officier de la Légion d’honneur* in 1927 for his military service. Here are some of the remarks that favoured his nomination: “A devoted doctor demonstrating initiative and goodwill. Distinguished himself during the bombardments on 21 and 22 April 1917 by ensuring, despite the particularly critical circumstances, the treatment of the wounded, for whom he worked tirelessly. He was part of a group of ambulance personnel serving members of the forward guard, who were particularly exposed to injury. Doctor with high medical and professional standards. Abso-



Figure 4 – Jules Sottas and Mme Mercadé, nurse at hospital no. 31 in Deauville, May 1915. (Private collection, jmb/14910.net, with kind permission).



Figure 5 – Jules Sottas, on the right, overseeing the treatment of a wounded soldier, May 1915. (Private collection, jmb/14910.net, with kind permission).

lutely devoted to wounded soldiers. His service during the war was much appreciated. 30 June 1927". These testimonies indicate that he started working on the front in 1917 [27].

4. Jules Sottas, historian

Around 1900, while Sottas was walking along the Seine, he found a book at one of the stands: *Journal d'un voyage fait aux Indes orientales par une escadre de six vaisseaux, commandée par M. de Quesne* (travel log relating a journey to the East Indies by a squadron of six vessels, under the command of Mr. de Quesne). It was after having read this book that Sottas was inspired to write a history of the India Company. Sottas set out to write a lively account, alternating between scenes of daily life, taverns, festivals and famines, and detailed technical information about ships at the time of Louis the 14th, or political matters. As an astute historian, he drew on the numerous archives that he consulted (Archives of the Ministry

of the Colonies, National Archives, the Arsenal, Maritime Affairs, etc.) to write a valuable work, one of real substance as well as literary quality, entitled *Histoire de la Compagnie royale des Indes Orientales, 1664–1719* (History of the Royal Oriental India Company, 1664–1719) and published in 1905 [28,29].

In 1907, Sottas, a member of the Société Astronomique de France, used his knowledge of mathematics and astronomy to describe an astrolabe from 1543 and the way in which it was used by sailors [30]. The abby Albert Anthiaume (1855–1931), a specialist in maritime and navigational history, collaborated with Sottas to write about the history of astronomical procedures for navigation that have been in use since antiquity. Their book [29] described the famous astrolabe in the Museum of Rouen and the millennial evolution of cosmographical knowledge that sailors have always relied on [30,31]. One wonders if those who used this instrument in the 16th century had a user's manual as clear and comprehensive as Sottas's book!

Just before being mobilized for World War I, Sottas became interested in the deployment strategies of Espinay Saint-Luc (1554–1597), one of the favourites of King Henri III. Espinay Saint-Luc became governor of Saintonge and was put in charge of naval construction [32,33]. All of Sottas's writings relate to maritime history, notably shipbuilding, in Saintonge and Aunis. In 1925, he wrote of the suffering endured by the cardinal Mazarin (1602–1661) because of his gout, involving incessant fits accompanied by renal colics and ultimately terminal renal failure [34]. But this was also to describe the role of the Prime Minister of King XV with regard to commerce and shipbuilding at La Rochelle and the role of the fortifications at Brouage [35], one of his very lucrative prebendaries (Fig. 6).

The renown of Sottas as a marine historian crossed the English Channel and led to two English publications: in 1928, *A Phoenician ship of the first century, A.D.* [36] and in 1930, *The Corvette l'Aurore, and its model* [37]. Some of his articles were published in journals for the general public, for example *Le bateau à travers les âges* (Ships throughout time) published in February 1928 in *Le Monde colonial illustré*. In 1938, Sottas published a history of a "sea parcel service in Venice during the 14th and 15th centuries" (Figs. 7 and 8) [38], praised by English specialists when it was published, [39] and it remains relevant today [40].

Shortly before he died, Sottas spoke of his admiration for Jean-Baptiste Charcot, who was a resident around the same time as Sottas. Charcot was the son of the Master at La Salpêtrière and was well known for his polar expeditions. Sottas's admiration was evident in the preface he wrote for the book by Marthe Emmanuel (1901–1997) entitled *J.-B. Charcot, le Polar Gentleman*. Sottas's curious mind, his vast knowledge of maritime history, and his multiple publications led to his being elected as a member of the Académie de Marine, of which he became president in 1942. In 2011, a Parisian publisher found and published a manuscript that Sottas was not able to finish before his death: *La marine ancienne de la Méditerranée, Antiquité et Moyen-Âge* (Ancient marine history in the Mediterranean, during antiquity and the middle ages.) The book bears witness to Sottas's encyclopaedic knowledge in the field of marine history [41]. He studied the shipbuilding techniques used by the Egyptians and Aegean peoples to design their oars, the techniques the Athenians used to build

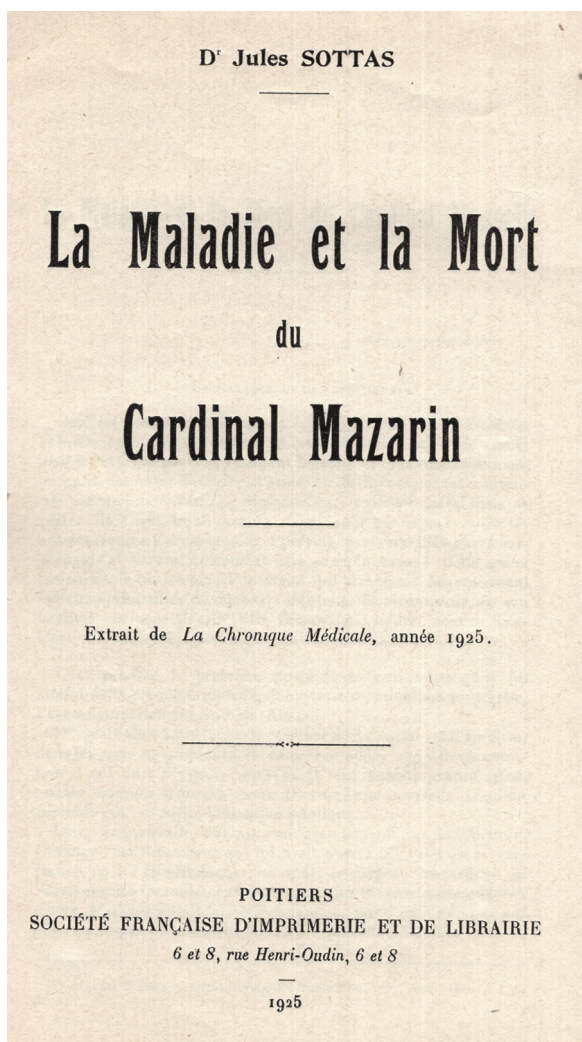


Figure 6 – Cover of a book written by Jules Sottas (Collection of the author).

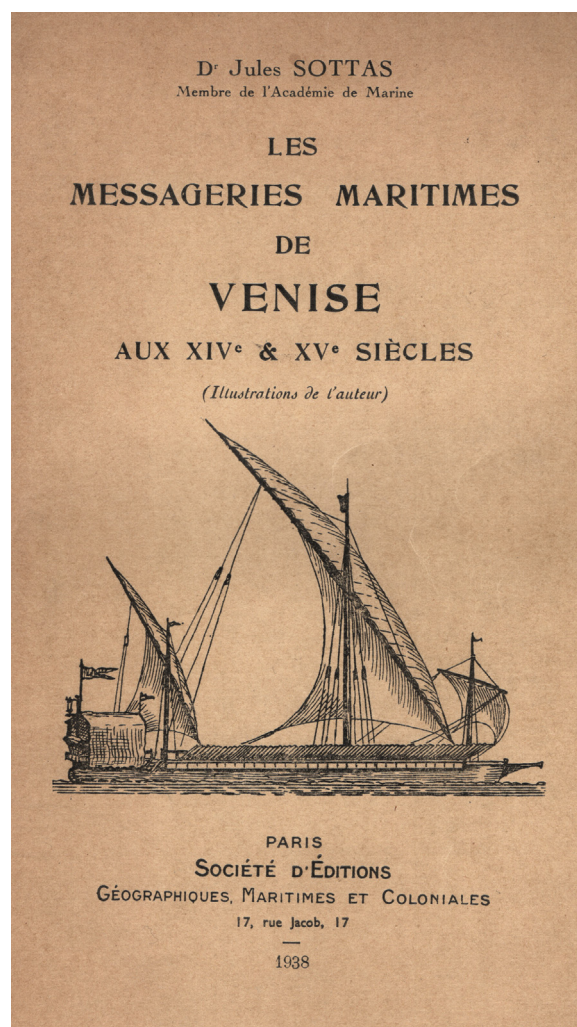


Figure 7 – Cover of the book *Les messageries maritimes de Venise* (Collection of the author).

their galleys, and the techniques used by the Romans to arm their powerful warships. Whether discussing the Spanish, Portuguese, or French ships used during the Crusades, or the ships rich traders from Venice used to sail the Mediterranean, Sottas shared his erudition with his readers, offering them detailed and comprehensive information about the ships, navigation, and the products transported.

5. Anecdotes and epilogue

Yvonne Sorrel-Dejerine (1891–1986), the daughter of Jules and Augusta Klumpke-Dejerine related how, in a family photo album, she found a card Jules Sottas had addressed to her parents to congratulate them when their daughter was born. She said she still remembered how fondly her father spoke of Dr. Sottas. She also related that before World War II, one of Sottas's daughters was the secretary of her husband Etienne Sorrel (1882–1965), an orthopaedic surgeon [42].

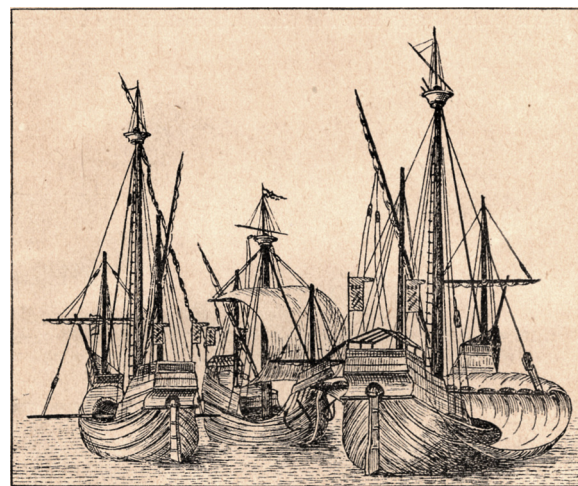


Figure 8 – Drawing of the hand of Jules Sottas in *Les messageries maritimes de Venise aux XIV^e et XV^e siècles* (Collection of the author).

Jules Sottas died on 28 September 1945. Not a single medical journal published so much as a brief obituary. Sottas's name is only remembered by neurologists because of the eponym, at a time when the practice of using eponyms is increasingly criticized. An excellent student who studied neuro-pathology under Jules Dejerine, Sottas decided against working as a clinician or in laboratories, even though a promising career seemed likely. He preferred working as a general practitioner, which made it possible for him to pursue his interests in historical research, especially marine research, a field in which he became a renowned specialist. Neurology undoubtedly lost something in the bargain.

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Disclosure of interest

The author declares that he has no competing interest.

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