Historical underpinnings of the term essential tremor in the late 19th century

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ABSTRACT

Background: The term *essential tremor* has been in regular use since the second half of the 20th century. To modern neurologists, the word "essential" may seem cryptic. The historical underpinnings of this term have not been examined.

Objectives: To bring to attention early medical reports using the term essential tremor and examine the characteristics of the disorder that contributed to the proposed use of the term.

Methods: Review of 19th and early 20th century medical literature on essential tremor.

Results: The term *tremore semplice essenziale* (simple essential tremor) was first used by Burresi (Italy, 1874) to describe an 18-year-old man with severe, isolated action tremor. Several years later, Maragliano (Italy, 1879), Nagy (Austria, 1890), and Raymond (France, 1892) described similar cases and proposed the terms *tremore essenziale congenito* (essential congenital tremor), *essentieller Tremor* (essential tremor), and *tremblement essentiel héréditaire* (hereditary essential tremor) to define the illness. Mirroring contemporaneous views of constitutional and inherited disease, the key ingredients of the disorder were viewed as the constant presence of tremor in the absence of other neurologic signs and its heritable nature. By the early 20th century, the term began to appear in the medical literature with greater frequency.

Conclusions: Toward the end of the 19th century, several clinicians attempted to provide a nosologic separation for a tremor diathesis that was often familial and occurred in isolation of other neurologic signs. This disorder, which was termed *essential tremor*, was later recognized as one of the most common neurologic disorders. ***Neurology*® 2008;71:856–859**

Essential tremor (ET) is one of the most prevalent neurologic disorders. The term itself began to gain regular and widespread use among neurologists during the second half of the 20th century.[1] To current day neurologists, the word "essential" may be cryptic and its original meaning, obscure. To patients with ET, the word "essential" can be irksome because it suggests that the disorder is in some way necessary and desirable. A small number of other medical (e.g., essential hypertension, essential thrombocytopenia) and neurologic conditions (essential anisocoria, essential myoclonus) have received the label "essential." Scholarly reports have been written on the history of tremor,[1,2] yet, to our knowledge, there has been no formal study of the historical roots of this term, *essential tremor*.

The goals of the current study were to 1) bring to attention what seem to be the earliest medical reports proposing the term essential tremor and 2) examine the characteristics of the disorder that contributed to the proposed use of the term essential tremor. An overarching goal is to demonstrate, among other things, how the development of the concept of essential tremor mirrored contemporaneous concepts of hereditary and constitutional disease.

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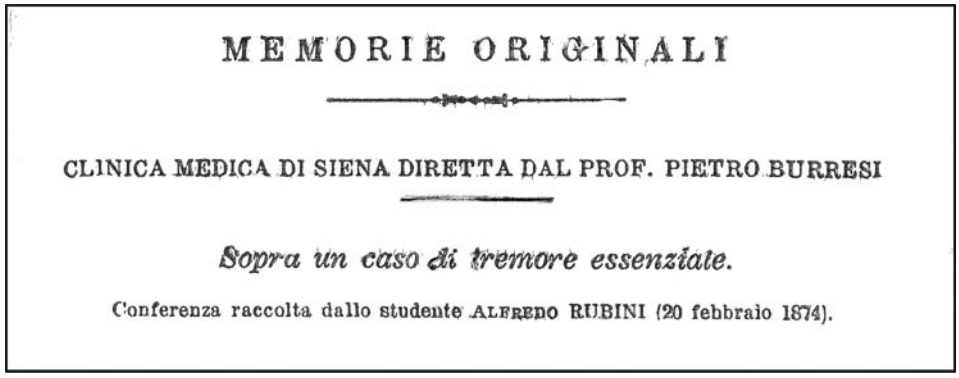
Figure Photograph of the first page of Pietro Burresi's 1874 publication[7]



MEMORIE ORIGINALI

CLINICA MEDICA DI SIENA DIRETTA DAL PROF. PIETRO BURRESI

Sopra un caso di tremore essenziale.

Conferenza raccolta dallo studente ALFREDO RUBINI (20 febbraio 1874).

METHODS We reviewed approximately 100 scientific articles written in the late 19th and early 20th centuries that referred to essential, hereditary, familial, congenital, or benign tremors, citing those that were most relevant to our aims.[3-25]

RESULTS During the 19th century, a number of investigators described single cases or familial forms of action tremor, including Most (in 1836),[3] and then in later decades (1860s–1890s), Sanders,[4] Eulenburg,[5,8] Fernet,[6] Burresi,[7] Maragliano,[9] Liégey,[10] West,[11] Dana,[12] Haebler,[13] Charcot,[14] Nagy,[15] Debove and Renault,[16] Rubens,[17] and Raymond.[18] As detailed below, several of these, namely Burresi,[7] Maragliano,[9] Nagy,[15] and Raymond,[18] proposed the term essential tremor to better define this motor disorder.

The term essential tremor was initially used in 1874 by Pietro Burresi,[7] Professor of Medicine at the University of Siena, Italy (figure). During a case conference that was later published,[7] Burresi described an 18-year-old man with severe, isolated action tremor. Tremor of the arms was present during voluntary movements. It was also present while walking yet disappeared during sleep. Head tremor was present. Neither parents nor siblings had tremor; hence, the tremor did not seem to be familial. In Burresi's discussion of the differential diagnosis, mercury and lead intoxication were easily excluded. Multiple sclerosis (MS) was excluded because there were no other motor or sensory abnormalities, and paralysis agitans, because there was no festination or gait and balance changes. Burresi proposed the term *tremore semplice essenziale* (i.e., simple essential tremor).[7] Important in his discussion was the notable absence of paralysis or other CNS signs.

Five years later, Edoardo Maragliano, Professor at University Hospital, Genova, Italy, reported a 62-year-old man with isolated action tremor.[9] The patient's mother had also developed a generalized limb and trunk tremor in advanced age. The tremor, which was severe and involved all limbs, was noted almost at birth, and worsened thereafter during adolescence and adulthood. It was professionally disabling and by age 50, he was no longer able to work as a gilder. Aside from tremor and mild gait unsteadiness, the neurologic examination was normal. Maragliano noted two similar patients who had been presented to him shortly after he had examined the reported case, and he discussed the differential diagnosis. Paralysis agitans, MS, and tremor secondary to toxic exposure were judged unlikely. The case was considered to be an idiopathic action tremor. Viewing its early (congenital) onset and heritable nature to be important, he proposed the term *tremore essenziale congenito* (i.e., essential congenital tremor).[9] He was apparently unaware of the 1874 report of Burresi,[7] not citing the earlier publication.

Anton Nagy (1863–1935) was an assistant in the Clinic of Nervous and Mental Disease in Graz, Austria, when, in 1890, he reported a 26-year-old woman with severe familial tremor with onset during childhood.[15] While attending school, she noticed tremor while writing. Her tremor worsened with time, increasing especially in the 2 years prior to Nagy's examination. Initially the arms had been affected but the tremor had gradually spread to her head and legs. When examined, there was severe action tremor of the arms; indeed, the patient could not use her hands for most daily activities (e.g., she could not use a knife or a fork to eat and she could not write). She also had severe head tremor as well as tremor in the legs and trunk. The neurologic examination was otherwise normal. Her brother, also examined by Nagy, had the same type of isolated action tremor. Drinking small amounts of ethanol made his tremor stop. Nagy further investigated the family and found, among 41 members over 6 generations, that 19 had tremor.[15] In each case, onset had occurred at a very early age, prior to puberty, and had progressed so that tremor was most prominent in the hands but also occasionally involved the head. Three affected family members also had a gait disorder but none exhibited other signs of CNS involvement.

Nagy discussed the differential diagnosis, excluding MS. He proposed the term *essentieller Tremor* (i.e., essential tremor).[15]

Shortly after Nagy's report, Fulgence Raymond (1842–1910), at the Salpêtrière Hospital in Paris, reported a particular form of tremor that he termed *tremblement essentiel héréditaire*.[18] The patient, a 52-year-old cabinet maker, had been hospitalized for influenza. The tremor had a frequency of 4–5 Hz, was sometimes responsive to ethanol, and occurred mainly in his arms. While primarily a postural and kinetic tremor, it also overflowed into resting conditions. Raymond noted that tremor occurred in isolation; muscle strength, reflexes, and sensation were all normal. The tremor had begun at an early age. Indeed, at age 9, "he realized that he shook when he started to learn how to write."[18] The tremor had increased in severity so that by age 13, he could not write any longer. Importantly, the patient's father, mother, and brother each had tremor. Raymond discussed the differential diagnosis. He reasoned that paralysis agitans, brain tumor, and MS, which were diseases in which tremor might be a major symptom, could be excluded since other symptoms and signs were usually present. The particular clinical presentation of this patient, as Raymond wrote, was "tremor . . . by itself . . . that is tremor without any other injuries or diseases of which it could be a symptom (*sine materia*)."[18] Raymond also excluded tremor due to alcoholism and hysterical tremor on the account that the patient did not resemble patients with these conditions and that the patient's history had not been one of excessive ethanol use. He considered senile tremor to be unlikely because the tremor had begun at an early age and there was no head tremor, which in Raymond's opinion,[18] was "one of the main features of senile tremor." Raymond proposed the term *tremblement essentiel héréditaire* (i.e., hereditary essential tremor).[18] He cited earlier reports of Dana[12] and Nagy,[15] reproducing large pedigrees from those reports, which illustrated the hereditary nature of the tremor. At the end of his report, Raymond wrote: "In summary, what we can conclude from the facts I just presented in this conference is that there is a variety of tremor that has hereditary component, which should be named *essential tremor* [Raymond's italics], because it occurs independently from any other symptom which would make us think of brain injury or intoxication."[18]

By the last decade of the 19th and early years of the 20th century, the term essential tremor began to appear more regularly in the medical literature.[19-25] Authors wrote about this entity, characterizing it as a chronic or lifelong condition,[19,20,23] which was hereditary,[19,20,24,25] and which occurred in relative isolation of other neurologic signs.[19,23,24,25] Several authors recorded the frequency of tremor and some did extensive physiologic recordings of their patients with tremor using sphygmographs,[12,16,18,19,21,22,24] showing that the tremor frequency ranged from 3 to 12 Hz.

DISCUSSION We studied the origins of the term essential tremor, citing the four reports that first seemed to have suggested the precise term or a slight variant of the term. While those of Nagy in Austria[15] and Raymond in France[18] have occasionally been cited, the two earlier reports by the Italian authors, Burresi[7] and Maragliano,[9] have not.

The authors of these reports highlighted a number of characteristics that could help in its delineation from the other varieties of tremor. The first was that the disorder was hereditary and often multigenerational. Physicians of the 19th century viewed diseases in three broad categories: acute infectious diseases (nonheritable), chronic and constitutional diseases (partly heritable), and absolutely heritable conditions.[26,27] Acute infectious diseases, like smallpox, were not under ordinary circumstances considered to be heritable. Chronic and constitutional diseases included a large number of entities like heart disease, cancers, and gout. These disorders were seen as having a substantial hereditary component,[26] although there were clear examples of families in whom "like did not beget like." Patients were seen as inheriting a tendency for the disorder, but lifestyle choices and life events determined whether each person would indeed succumb to illness and what type of illness he or she would develop.[26] Finally, a small number of discrete conditions were viewed as absolutely hereditary.[26,27] The affected families had evidence of many affected persons in the same family and in some diseases, affected individuals in each of several generations. These included families with hemophilia and colorblindness as well as neurologic disorders like Huntington disease and Friedrich ataxia.

The second and perhaps key feature of ET was that the tremor was virtually always present (even sometimes at rest), yet was the "only symptom" detected in patients. In this sense, the use of the word "essential" embodied the notion of a constitutional property. Some individual aspects of appearance, character, and health, regardless of whether they ran in families, were viewed in the 19th century as highly resistant to change over time.[27] Hence, a person was born and died with fixed properties such as hair color, a general body form, a certain mental acuity, and a certain emotionality and temperament.[27] As the tremor of ET, these were often simple, unitary properties, existing independently of other properties. These core elements of individuality were, in many respects, akin to physiologic constants.[27] With

regards to health, some conditions (e.g., infections) were the result of the entry of foreign entities into the body, while other more constitutional conditions were viewed as so firmly fixed within the organized structure of the body that they become an intrinsic part of the individual makeup.[27-29]

The current study suggests that the first use of the term essential tremor was in 1874 by Burresi.[7] To our knowledge, the origins and development of this term have not previously been traced. Of interest is that the 1874 report of Burresi[7] was never properly referenced in the medical literature and has been largely forgotten, which may also be due in part to the fact that Italian was not one of the three mainstream medical-scientific languages (German, English, French).

By chance, we found a reference to this article in an 1899 publication by the German author Graupner.[22] However, Graupner incorrectly attributed the article to Rubini, who was a student who reported the conference given by Burresi (figure). Earlier, in a book chapter published in 1877,[8] the name Rubini and the year 1874 were cited but there was no title and an incomplete journal reference. This may explain why the Burresi report remained without citation during the 20th century. In the mid-20th century, for example, Macdonald Critchley[30] published an exhaustive clinical review on ET, including many early references from German, French, English, and Italian languages, yet the Burresi report was not cited.

Beginning in the late 1800s, a number of clinicians began to provide a nosologic separation for a tremor diathesis that was often familial and occurred in isolation of other neurologic signs. This disorder, which was termed essential tremor, was later recognized as one of the most common neurologic disorders.

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