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Review

A brief history of typical absence seizures — Petit mal revisited

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ABSTRACT

In this article, we have traced back the history of typical absence seizures, from their initial clinical description to the more recent nosological position. The first description of absence seizures was made by Poupart in 1705 and Tissot in 1770. In 1824, Calmeil introduced the term "*absences*", and in 1838, Esquirol for the first time used the term *petit mal*. Reynolds instead used the term "*epilepsia mitior*" (milder epilepsy) and provided a comprehensive description of absence seizures (1861). In 1854, Delasiauve ranked absences as the seizure type with lower severity and introduced the concept of idiopathic epilepsy. Otto Binswanger (1899) discussed the role of cortex in the pathophysiology of "abortive seizures", whereas William Gowers (1901) emphasized the importance of a detailed clinical history to identify nonmotor seizures or very mild motor phenomena which otherwise may go unnoticed or considered not epileptic. At the beginning of the 20th Century, the term *pyknolepsy* was introduced, but initially was not universally considered as a type of epilepsy; it was definitely recognized as an epileptic entity only in 1945, based on electroencephalogram (EEG) recordings. Hans Berger, the inventor of the EEG, made also the first EEG recording of an atypical absence (his results were published only in 1933), whereas the characteristic EEG pattern was reported by neurophysiologists of the Harvard Medical School in 1935. The discovery of EEG made it also possible to differentiate absence seizures from so called "psychomotor" seizures occurring in temporal lobe epilepsy. Penfield and Jasper (1938) considered absences as expression of "*centrencephalic epilepsy*". Typical absences seizures are now classified by the International League Against Epilepsy among generalized nonmotor (absence) seizures.

1. Introduction

In 1981, the ILAE defined absence seizures as impairment of consciousness with mild clonic, atonic, tonic, or autonomic components [1]. The clinical hallmark of absence seizures is a "blank stare". In the new ILAE classification, absences are defined ad generalized nonmotor seizures [2], which does not take the full spectrum of clinical phenomena into consideration. Typical absences are associated with generalized spike–wave discharges on electroencephalogram (EEG), and develop in childhood, typically between 4 and 10 years of age, but later ages have been reported [3], and are encountered almost exclusively in idiopathic generalized epileptic syndromes [4]. These seizures usually last 9 to 10 s, and often occur tens or hundreds of times per day. They are easily provoked by hyperventilation [5] and less commonly by photic stimulation, and may be associated with eye opening, eyelid movements, and oral automatisms [6,7].

Brain magnetic resonance imaging of patients with typical absence seizures is normal, and may not be required for the diagnosis in cases with typical electroclinical features.

The neurophysiological correlate of typical absence seizures is generalized 2.5 to 5 Hertz (Hz) spike–wave discharges (classically, 3 Hz) with abrupt onset and termination [8], sometimes with higher amplitude over frontal regions [9] on the EEG. The interictal EEG has normal background activity although sometimes short bursts of spike–wave discharges may occur. These characteristic electroclinical features permit to differentiate typical absence seizures from atypical absence seizures or other seizures with impaired consciousness.

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Depending on the epilepsy in the context of which they appear, typical absence seizures may occur as the only seizure type or together with generalized tonic–clonic seizures or myoclonic seizures.

In this article, we have traced back the history of typical absence seizures, from their initial clinical description to the more recent nosological positioning. However, one should be extremely cautious when dealing with historic records on epilepsy. The neurophysiological basis of the disease was discovered only toward the end of the 19th Century, and one had to wait until the 1920's for the possibility of recording the brain electrical activity of patients with epilepsy by means of the EEG. As reported above, absences are seizure types which can be adequately diagnosed only with an EEG recording showing the characteristic 3-Hz generalized pattern. Consequently, it is possible that historical records referring to "petit mal" or "absences" in the pre-EEG era may have actually referred to nonmotor seizures other than true typical absences or even to nonepileptic events. With this caveat in mind, we have searched the extant scientific literature to report any relevant reference to absence seizures with the final aim of sketching out a short, but comprehensive history of this seizure type. Being aware of the importance of analyzing historical documents as directly as possible, we have reported the most relevant excerpts found in the literature in English translation (made by F. Brigo and E. Trinka; the original French and German texts are reported as supplementary material).

2. The first descriptions

At the first London-Innsbruck Colloquium on Status epilepticus a historical case was presented, which represents most likely the first historical description of absences in the form of status. It is documented on ex-voto table of 1501 in the parish church of Gmünd in Austria (Fig. 1). The text says:

> *"Oswalt…, a citizen of Gmünd, went 6 years ago to the old Ötting with his son who has the falling sickness, was little improved and he resolved never again to take to old Ötting. Now, in the week after Oculi in the year 1501, fell into great illness that he lay with open eyes speechless until the third day and many people came to him, did not recognize or see anybody, and they lost all hope for his life. So, his wife engaged him here with a priest and a waxen head, and he recovered. Say honor, praise and thanks to the Virgin Mary in eternity. And this happened here on the Sunday before Ascension Day in the year 1501".* – Altötting or "the old Ötting" is to this day a very popular pilgrimage in Bavaria [10].

In his seminal book on "The Falling Sickness – A History of Epilepsy from the Greeks to the beginnings of modern Neurology" [11], Owsei Temkin writes that the first description of absence seizures was made by a certain Monsieur Poupart (possibly François Poupart, 1661–1709, French physician and anatomist) and reported in the proceedings of the *Académie Royale des Sciences* [12]:

> *"In that occasion Mr. Poupart added that he was aware of a case of a young female child with epilepsy, who at the onset of the seizure sits down in a chair, and there remains immobile, without speaking and senseless. Her eyes are open, and after the episode she does not absolutely remember having fallen into this state. If she had previously begun a talk that the seizure interrupted, she takes it up again exactly at the same point at which she stopped talking, and she thinks she has*

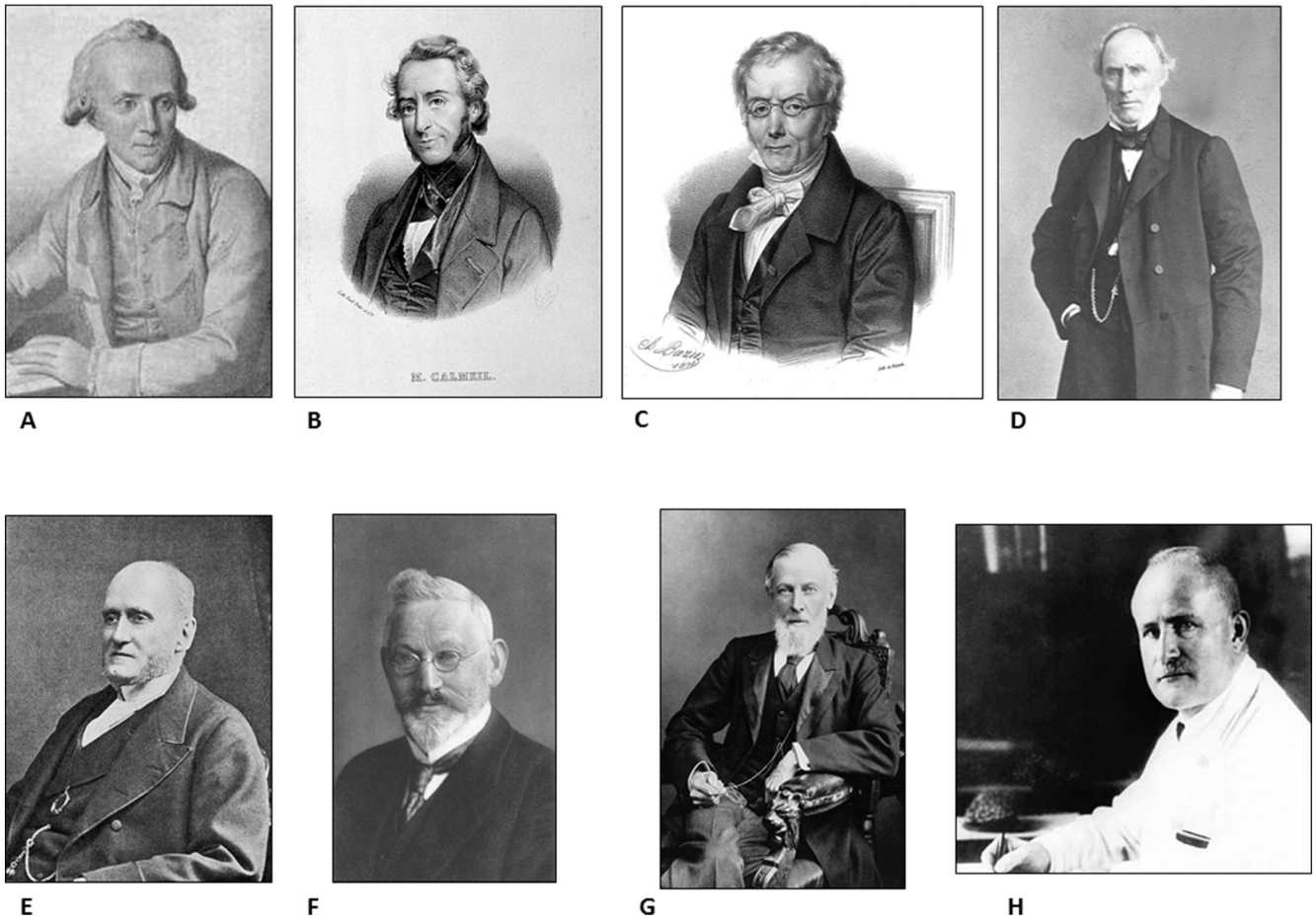

Fig. 1. A. Simon-Auguste Tissot (1728–1797); **B.** Louis-Florentin Calmeil (1798–1895); **C.** Jean-Étienne Dominique Esquirol (1772–1840); **D.** Louis Jean François Delasiauve (1804–1893); **E.** John Russell Reynolds (1828–1896); **F.** Otto Binswanger (1852–1929); **G.** William Richard Gowers (1845–1915); **H.** Hans Berger (1873–1941).