

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.

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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 as 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

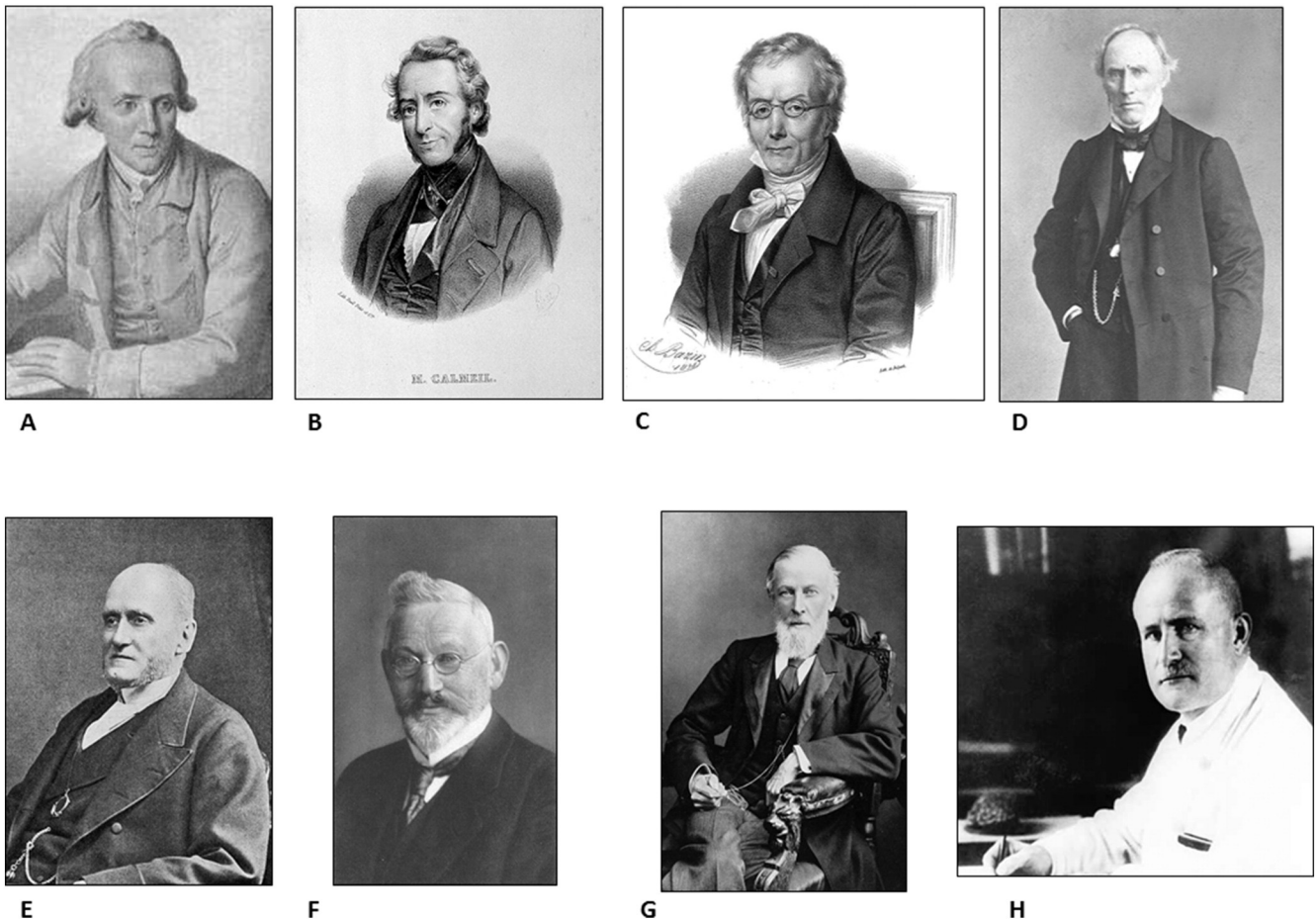


Fig. 1. A. Simon-Auguste Tissot (1728–1797); B. Louis-Florentin Calmeil (1798–1895); C. Jean-Étienne Dominique Esquirol (1772–1840); D. Louis Jean François Delasiauve (1804–1893); E. John Russell Reynolds (1828–1896); F. Otto Binswanger (1852–1929); G. William Richard Gowers (1845–1915); H. Hans Berger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

talked without interruption.”

This first description of a patient with absences already contains the main clinical features of this seizure type, which are as follows: the sudden onset and abrupt end, the cessation of ongoing motor activity without loss of muscular tone, and with the resume of previous activity as if nothing had happened at the end of the seizure.

A few years later (1770), the Swiss neurologist Samuel August David Tissot (1728–1797) in his book on epilepsy (*Traité de l'épilepsie*) gave the following description of a patient with absence seizures [13]:

“They brought me in September 1765 a foreign 14-year-old female, whose disease has remarkable features.

She had been in good health until the age of seven; at that time, she had a trip on the water with other young people; a violent storm terrified them, but she was the only person who did not vomit. After a few days, one noticed a movement of the eyelids initially appearing as a tic, but which was soon recognized as convulsive. She was entrusted to two experienced physicians who could not prevent the occurrence within the next four months of attacks of epilepsy, which were very severe and frequent, and lasted several months. During part of this time, the young patient frequently had — in the intervals between the great attacks — very short attacks, which were accompanied only by a sudden loss of consciousness which interrupted her speech, together with a very slight movement in the eyes; Often, after recovering, she completed the sentence in the middle of which she had been interrupted; other times she had forgotten it.

During another part of the same time, these sudden attacks seized her only when she was walking: she stopped senseless for a few seconds, and there was always a slight convulsive movement in the leg which was in front. Nonetheless, the great attacks faded away, but the small ones were frequent so that one day, after having had several of them, the patient had a bath in a river with a housemaid, and thereafter that she remained twenty-one months without having either great or small attacks. The great attacks reappeared concomitantly with the grief due to the unexpected death of her father, and afterwards they continued, and are very frequent from the first cold periods of autumn until the first warm days of summer; however, during all this last season the patient has not had any attack and enjoyed good health, despite the fact that she is extremely sensitive: it often becomes sad and gets scared with great ease; despite the efficacy of the previous one, cold baths have been ineffective during this relapse.”

This report is interesting because for the first time it emphasizes the possibility that in some epilepsies, absence seizures may occur together with other seizure types characterized by motor phenomena. It also describes abnormal eyelid movements (“one noticed a movement of the eyelids initially appearing as a tic, but which was soon recognized as convulsive”), clinical phenomena which may be associated with absences. Furthermore, this excerpt is relevant as for the first time in the medical literature two different terms are used to indicate different attacks (*grands accès* and *petits accès*) [11].

In Tissot's view, it is the insensibility and impairment of consciousness that characterize the epileptic attacks, and which may distinguish between epilepsy and other convulsions. It was this feature that “facilitated the recognition of the epileptic character of what is now called absence.” [11].

3. Terminology

In 1824, the French physician Louis Florentin Calmeil (1798–1895), who had experiences of patients with epilepsy in the asylums of the Salpêtrière and Charenton in Paris, introduced the term “absences” in his doctoral thesis for the University of Paris [14]:

“Absences. The absences are not rare among epileptic patients; they do not seem to be dangerous, but constitute a curious phenomenon. The

patient let drop the object he held on his hand; then, without any other bizarre symptoms, he becomes unaware of what happens around him; although he is awake, his senses are close to any [external] impression; it is a true ecstasy. The functions [of the body] are not impaired during these episodes; if at the beginning one questions the patient, the absence interrupts; if one observes it without saying anything, it interrupts within some seconds. I tend to believe that the absence is nothing but an abortive vertigo, just as vertiginous attacks can be incomplete attacks of grand mal.”

The French psychiatrist Jean-Étienne Dominique Esquirol (1772–1840) was the first to use the term *petit mal* to describe this type of epilepsy [15]:

“Sometimes the attacks have different intensity: some of them are strong and others weak; that's what in hospitals is called 'le grand mal' and 'le petit mal'.”

Since their first appearance the terms “absences” and “petit mal” had been accepted rapidly by the medical community; the terms were initially used to indicate a great variety of attacks lacking the characteristics of grand mal. According to the Oxford English Dictionary the term “absence” denotes a state of being absent or away from any place. In the 18th Century, the term was also used in a sense of absence of mind, such as “inattention to what is going on or failure to receive impression of what is present” [16]. As Owsei Temkin has brilliantly pointed out “from a logical point of view this terminology looseness might appear unsatisfactory, but from the point of view of a progressive science it proved adequate.” [11].

In 1854, Louis Jean François Delasiauve (1804–1893), who was based at the Bicêtre, differentiated four types of seizures, which were ranked in order of increased severity: *absences* for petit mal, *vertiges* for vertiginous attacks, *accès intermédiaires* or *chutes*, and *chutes* or *accès complets* for generalized convulsive seizures. In his book “*Traité de l'épilepsie; histoire, traitement, médecine légale*”, he also classified epilepsies into the following: 1. idiopathic (*épilepsie essentielle ou idiopathique*; characterized by absence of physical lesions; the seizures are by themselves the disease and a neurotic disorder); 2. symptomatic (*épilepsie symptomatique*; cerebral lesions are present; convulsions are a symptom of a certain disease and not the disease itself); and 3. sympathetic epilepsy (*épilepsie sympathique*; produced by the irradiation of abnormal impressions which can have their seat outside the central nervous system) [17].

In his monography, Delasiauve describes *absences* as follows [17]:

“Absences. – They correspond to the lower degree of epilepsy. The symptoms characterizing the absences have been perfectly described by Mr. Calmeil. On the other hand, they were not completely unknown. In particular Mercuriali and, later on, Tissot and Portal have reported some cases where the convulsions were lacking or too mild to be recognized. Within one or some seconds, the consciousness of the patient is suddenly impaired. If the patient speaks, he stops speaking; if he listens, he stops paying attention to the person who is talking; if he walks, he stops. This interruption can go unnoticed by the bystanders; the patient himself is unaware of it, and sometimes takes up again the talk exactly at the same point at which he stopped talking, he adds the lacking sentences and completes the words he had started. It also happens, during this sort of eclipse of the mind, something more severe: the epileptics not only let drop or break the objects they have in their hands, but they may fall from heights, are burned or drown accidentally. One of our patients called his interruptions “blackouts”, which occur frequently, sometimes with violent spasms, which luckily are very rare. For those who observe them very accurately, the absences have their own external correlate, manifesting with staring gaze, astonishment, and even unusual pallor of the face. With this regard, one could say that although aware, the senses are close to any [external] impression.

Mr. Herpin even thinks, like other authors, that it is not impossible to notice mild partial convulsions. Mr. Calmeil, conversely, regards the absences as abortive vertiginous attacks.

This transitory state can sometimes be interrupted by calling the patient with violence. In many cases it is preceded or followed by a painful cerebral numbness, which indicates the attack, and persists long time after it. The memory remains confused, whereas the character of the patient is taciturn, impatient, susceptible and annoyed. This complication becomes worrisome with the increasing number of absences."

Sir John Russell Reynolds (1828–1896) used the term "*epilepsia mitior*" (milder epilepsy) and provided a comprehensive description in his book "Epilepsy; its symptoms, treatment, and relation to other chronic convulsive diseases" [18]:

"A. EPILEPSIA MITIOR; OR, LE PETIT MAL

There are, as already stated, two forms of 'le petit mal': in the one there is no evident spasm; in the other there is.

1. *Epilepsia mitior*, without evident spasm. — Although M. Delasiauve states that epileptic attacks always present 'leur traduction extérieure'; and Portal, Hasse, and others speak of spasm as constantly present, we are bound by experience to say, that in some instances such spasm is not to be observed; and, by pathological inference, to assert that visible spasm is by no means necessary.

All that is positively known to occur is a sudden and temporary, but absolute arrest of both perception and volition. The individual so attacked loses consciousness, for two or three or more seconds; and may, after that period, resume his sentence, or employment, perfectly unaware that anything abnormal has transpired. Thus, Georget mentions the case of a lady who was frequently attacked while performing on the pianoforte, and who would resume the phrase of music which was broken by the seizure.

If such attacks take place when the individual is standing, he may exhibit no loss of equilibrium; or he may lean slightly forward, or to one side. I have known these seizures to occur in a gentleman while hunting, and yet he has maintained his seat. Sometimes there is pallor of countenance; at other times there is pallor succeeded by slight flushing; again, there may be flushing, or a dusky hue without previous pallor; but in certain cases, there is no evident change of colour; — the face appearing precisely the same, in this respect, before, during, and after the suspension of consciousness.

It often happens, however, that the attack — although unaccompanied by apparent spasm—is of more than this momentary duration; and that some after-effects are observed, which continue for a variable time. Thus, although perception and volition return, after a few moments' suspension, they do not return to their normal state; but the patient is confused, and may exhibit some delusion; i.e. he may not only fail to comprehend where he is, or to recognise a well-known face, but he may be under the impression that something has transpired which is not the fact, or that a person near him is somebody else. He is dull of apprehension, depressed in spirits, suspicious; and, for an hour or two, may remain in this condition, and be quite forgetful of what has occurred immediately before the attack, or even for several days beforehand. Gradually, however, he recovers, and is in the same state as before.

In other instances, the attack is preceded and accompanied, for the first few moments of its occurrence, by vertigo; the individual thinking that he is about to fall, and perhaps exhibiting some tendency to do so. He staggers, and grasps some object for support; or his legs sink under him; or if sitting, the muscles which maintain the erect posture, yield; and he slips forwards or to the side. It rarely, however, happens that the patient falls, — he ceases to perform any act requiring volition; — i.e. he ceases to speak; to use his fingers, if writing or otherwise engaged; but those movements which are primarily automatic continue, such as those which combine to produce the erect position, either of sitting or standing; and those which are secondarily automatic are only

disturbed, but not arrested, so that the individual may continue riding or walking, and may still hold in his hand the pen, riding-whip, or walking-stick, which he grasps by an unconscious effort.

He may not stand symmetrically; he may walk doubtfully may grasp an object less firmly while the attack continues, than he did before; but the coordinated movement persists, and differs only in the force with which it is exhibited."

The term *epilepsia mitior* without evident spasm was used by Reynolds to refer to seizures without major motor signs, such as myoclonic jerks or convulsions. It might therefore have been used to indicate absence seizures. Some of the clinical features here reported (sudden onset and abrupt end, short duration, cessation of ongoing motor activity without loss of muscular tone, resume of previous activity at the end, vegetative symptoms such as pallor) are characteristic of typical absence seizures. Conversely, other symptoms (particularly the confusion and delusion lasting some time "after a few moments' suspension" of "perception and volition") appear more typical of seizures currently classified as "focal seizures with impaired awareness" [2].

4. Otto Binswanger and William Richard Gowers: the first modern descriptions of absence seizures

In 1899, the Swiss neurologist and psychiatrist Otto Binswanger (1852–1929) published a book on epilepsy (*Die Epilepsie*), which became a reference in the field. An entire chapter of this textbook is devoted to the description of "petit mal" seizures, which Binswanger calls abortive seizures (*Abortive Anfälle*; transl.: abortive seizures) [19]:

"**Abortive seizures (petit mal)**

Under this name we summarize all those seizures, in which one of the two main factors of the epileptic insult either is completely absent or is present only to a very limited extent. The epileptic seizures with the highest frequency and variability of clinical forms are a) the abortive seizures in which the motor-convulsive component is absent or minimal, while the impairment of consciousness captivates almost exclusively the attention of the observer. Much rarer are b) the seizures, in which the disturbance of consciousness is absent and only short motor excitation or inhibition discharges constitute the attack."

The chapter devoted by Binswanger to abortive seizures opens with a description of the underlying pathophysiology: these seizures are considered the expression of abnormal discharges in the cortex. The cortical origin of epilepsy had been discovered few years before by the German anatomist physiologist Gustav Theodor Fritsch (1838–1927) and the German neurologist and neuropsychiatrist Eduard Hitzig (1838–1907), who reported in the article "On the Electric Excitability of the Cerebrum" the results of seizures provoked by electric stimulation of the brain cortex of dogs [20].

"Pathophysiologically, the abortive seizures are the closest to the aura, as they also arise from quite limited discharges. As the examples below show, they may be so difficult to distinguish from sensory, motor and vasomotor aura symptoms that it is hardly possible to separate these two phases of the abortive seizure. Only cases of the first category are considered here. We are dealing with initially isolated inhibitory or excitatory discharges, which have led to the diffuse discharge of the cerebral cortex (unconsciousness). We must assume that there are minor charge processes which have already been compensated for by these short-lasting discharges limited to the cortex.

On the other hand, the abortive attacks of category a) almost never lack the vasomotor symptoms. In the abortive seizures without a pronounced aura, so in the cases sub a) the insult starts with a diffuse inhibition discharge of the Cortex, to which isolated sporadic motor irritation phenomena can add. In the seizures sub b) even the initial diffuse

inhibition discharge of the cortex is absent."

Binswanger then discusses the appropriateness of the term *Vertigo epileptica* (the concept of epileptic vertigo was first proposed by Calmeil and then extensively adopted and further developed by Delasiauve); Binswanger reports the objection to this term previously made by the German neurologist Carl Wilhelm Hermann Nothnagel (1841–1905), and questions its use to indicate epileptic interruptions of consciousness (*epileptische Bewusstseinslücken*), suggesting to limit this terminology to seizures with incomplete impairment of consciousness, of which the patient has "a certain, albeit very summary and incomplete memory of the insult and very often refers to it with the term dizziness or fainting". It is likely that these seizures correspond to what are currently classified as "focal seizures impaired awareness" [2].

"The seizures classified under a) are generally referred to epileptic vertiginous seizures, which the French very appropriately call 'absence'. The objection made by Nothnagel to the term 'epileptic dizziness' is somewhat justified, as in a great number of cases dizziness in the narrow sense does not appear, but transient interruptions of consciousness, which do not even reach the patient's perception. Only in the cases in which the impairment of consciousness is not complete, but only distortion of consciousness, aggravation of mental operations, instantaneous inability to organize thoughts, obscuration of visual fields and the like occur, the patient has a certain, albeit very summary and incomplete memory of the insult and very often refers to it with the term dizziness or fainting. To do justice to the doubts of Nothnagel, the term Vertigo epileptica can be limited to the cases just described; these are in contrast to the cases of epileptic interruptions of consciousness reported above."

Binswanger subsequently discusses the need for a careful clinical observation to detect abortive seizures and claims that this kind of seizures is often misunderstood or underrecognized even by physicians. Probably, he was right in his concerns, if one considers that in some neurology textbook of the time, such as the "Sémiologie des Affections du Système Nerveux" by Joseph Jules Déjerine (1849–1917) [21], petit mal was not even mentioned.

"These abortive seizures have already been attributed to epilepsy by previous physicians (here we refer only to Tissot and Portal), because the observation of patients has taught that in many patients these seizures occur besides fully developed seizures, and sometimes they are even more frequent. It took an in-depth understanding of the nature and symptoms of epileptic seizures to properly recognize these conditions. A knowledge of the nature and the symptoms of epileptic seizures is required to correctly recognize these conditions. The clinical picture here is silent and quiet, barely noticeable by the distant observer. The diametrical contrast to the noisy, stormy and frightening mode of appearance of the fully developed epileptic seizure is well suited to diverting the theory that two affections apparently so different actually belong to the same disease.

Even today the importance of abortive seizures is often misunderstood and underestimated in medical and lay circles. Every experienced neurologist will therefore be acquainted with a greater number of observations in which the diagnosis of epilepsy has not been made correctly, as long as nocturnal or abortive seizures occur."

Binswanger then provides a detailed description of the clinical features of abortive seizures, and refers to a personal case and previous observations made by Jean Étienne Georget (1795–1828), a French psychiatrist who had been a student of Jean-Étienne Dominique Esquirol (1772–1840) and by the French physician Armand Trousseau (1801–1867).

"The most common are the following symptoms: in the midst of some activity the consciousness of the patient fades for a few seconds. He

turns pale, his speech stops suddenly. The patient stares into the void for a moment; unordered movements of the lips and tongues occur, which usually have the character of smacking and snapping movements or are recognizable as incomprehensible murmurings or lolling. After a few moments this condition is over, the patient moves his head slightly, usually with a defensive or shaking movement, looks around in astonishment, passes his hand through his hair, as though something unusual and inconceivable has happened to him, and then continues the started speech. Alternatively, the patient is surprised by the attack in some activities, for example, when eating, playing cards or writing. He pauses for a moment in his movements, the hand shows slight, involuntary tremors or, involuntarily, clumsily gropes on the table or, if raised, remains motionless in the raised position. With the disappearance of the seizure the patient proceeds in the interrupted activity. Not infrequently, the face retains the physiognomic expression that it had when unconsciousness occurred. In other cases he becomes rigid and mask-like. If the patient is surprised by such a seizure while walking, he either falters, stands motionless for a few moments, or goes like nightwalking with a stiff posture, slightly unsteady on his way. The observer usually notices the seizure if there is an interruption of consciousness, forcing the patient to violently 'collapse,' pause for a moment, and look around in astonishment or embarrassment. We mention some well-known examples from the literature, which complete the above description.

Georget tells of a lady who was often plagued by a seizure while playing the pianoforte and after it immediately continued from the interrupted measure. Trousseau tells of a man who had often attacks of petit mal. While holding in his hand the card he is about to throw, he suddenly becomes immobile, closes his eyes or looks straight ahead; with a deep sigh, he soon comes to himself, and then throws, as if nothing had happened, the card on the table.

In one of our patients, we could observe these abortive seizures several times during the midday meal. While he was spooning out the soup, he suddenly stopped moving after lifting his arm and putting his spoon between his lips. The soup came down from his half-opened mouth. The hand shook slightly; after a few seconds the attack was over, and then the patient continued his activity."

William Richard Gowers (1845–1915), in his seminal book "Epilepsy and other chronic convulsive diseases: their causes, symptoms, and treatment" provided accurate and detailed descriptions of absence seizures [22]. Gowers was active at the National Hospital for the Paralysed and the Epileptic, located in Queen Square in London, the first hospital dedicated to the study and treatment of neurologic conditions which was inaugurated in 1860 [23].

He describes absences as follows [22]:

"The attacks or seizures which characterize epilepsy are commonly divided into two classes, — major or severe, and minor or slight. These two forms, although clearly distinguished in their general characters, are not separated by a sharp demarcation. In the major attacks (grand mal) there is loss of consciousness, often prolonged, and severe muscular spasm. In the minor attacks (petit mal) there is commonly brief loss of consciousness without muscular spasm; sometimes there is loss of consciousness and slight muscular spasm; very rarely there is slight muscular spasm without loss of consciousness. The French term 'petit mal' has become widely used, but the slight attacks it designates are termed by their subjects 'sensations,' 'turns,' 'faints,' etc."

In this excerpt, Gowers refers to the widely used terms "petit mal" and "grand mal". In his description of absences as in that made by Reynolds, mild motor symptoms either in the form of automatism or in the form of mild spasms have been clearly described. It has been emphasized, that these mild motor phenomena do not take the form of convulsions, but still they are well observable. Thus, the current term "nonmotor" instead of absence seizures seems to be an oversimplification.

“One of my patients, a girl of twenty-five, had suffered for twenty years from petit mal, with complete loss of consciousness; thirty to forty attacks occurred daily, and yet there was no defect of mental power — not even of memory.”

This passage emphasizes the overall good prognosis of some epilepsy types characterized by typical absences (e.g., childhood absence epilepsy), which may achieve seizure remission spontaneously.

“Epilepsy may commence in one of three ways. First by minor seizures which occur alone for months or years before there are severe attacks. The attacks of petit mal are often at first slight and attract little notice, but become more frequent, until suddenly a severe convulsion occurs. The patient and his friends do not associate the two forms of attack, and it is always necessary to make careful inquiry for the occurrence of minor seizures, antecedent to that which the patient believes to have been the first epileptic fit. It is common, for instance, for a patient to say that he has had fits for a few months only, when attacks of petit mal have been occurring for years. I have known such attacks to occur during eight years in one case and six in another, without any significance being attached to them, although one patient often fell. In one instance attention was only directed to the attacks, which at first simply caused the patient, a boy, to drop his book in class, because, later on, he slowly turned round on each occasion. The meaning of the minor attacks is only recognised, as a rule, when they give place to severe convulsive seizures, and often, even then, their significance is not always discerned.”

“Many patients who suffer from severe attacks, suffer also from minor seizures. The proportion is difficult to ascertain. The occurrence of the slight attacks may elude recognition unless the observer is aware of their common forms, and inquires for them. They are seldom associated with the disease by the patient or friends. Further, attacks are often described as ‘slight’ which present some spasm as well as unconsciousness, because they are less severe than other attacks which the patient has. The attacks of petit mal may alone constitute the disease. I have met with one case in which they occurred, without any convulsions, for forty years.”

These two extracts are particularly remarkable, as they emphasize the importance of a detailed clinical history from the patient and the relatives also with the aim of identifying absence seizures with or without mild motor phenomena (e.g., myoclonic jerks) which otherwise may go unnoticed or considered not epileptic. The second excerpt is interesting as it specifies that in some epilepsies, absences may be the only seizure type.

5. Pyknolepsy: a mysterious entity

In 1906, the Mannheim Neurologist Max Friedmann (1858–1925) reported the long-term favorable prognosis of some absence seizures, which he — contrary to the main view — called *kurze narkoleptische Anfälle* (short narcoleptic attacks), hence denying their epileptic nature [24]. As only 1 out of his 15 reported patients developed convulsive seizures, Friedmann did not consider these seizures to be the expression of epilepsy, but rather of narcolepsy, an entity which had been described toward the end of 19th Century in Germany and France [25], and whose real nature was still debated.

One year later, the German neurologist Karl Heilbronner (1869–1914) described the high frequency of absence seizures occurring in some patients using the term *gehäufte kleine Anfälle* (frequent minor attacks) [26]. The same term was used by Walter Cohn in 1919 [27] and by H. Sauer from the University of Greifswald in 1916 [28]. Sauer further emphasized the high frequency of these seizures in some patients coining the term *pyknolepsy* (from the Greek word πυκνός, *pyknos*, meaning dense, closely packed, aggregated). This term gained popularity and was initially used to indicate a unique entity, different from epilepsy and narcolepsy, and difficult to diagnose reliably in individual cases [29]. The same term was adopted also by William John Adie (1886–1935) in 1924, who described narcolepsy as *“a form of epilepsy*

occurring in children, with a good prognosis”. This epilepsy type was described as *“a disease with an explosive onset between the ages of 4 and 12 years, of frequent short, very slight, monotonous minor epileptiform seizures of uniform severity, which recur almost daily for weeks, months, or years, are uninfluenced by antiepileptic remedies, do not impede normal and psychical development, and ultimately cease spontaneously never to return. At most, the eyeballs may roll upwards, the lids may flicker, and the arms may be raised by a feeble tonic spasm. Clonic movements, however slight, obvious vasomotor disturbances, palpitations, and lassitude or confusion after the attacks are equivocal symptoms strongly suggestive of oncoming grave epilepsy, and for the present they should be considered as foreign to the more favorable disease.”* [30].

It was only in 1945 that, based on EEG recordings, pyknolepsy was recognized as an epileptic entity, with seizures corresponding to the same generalized spike-wave discharge pattern encountered in other typical absences [31].

6. The discovery of provoking factors and the first descriptions of EEG features

In 1924, hyperventilation was recognized as an effective test to provoke absences by Otfried Foerster (1873–1941), a German neurologist and neurosurgeon, who termed the epilepsy type characterized by these seizures as *Hyperventilationsepilepsie* (hyperventilation-epilepsy) [32].

Hans Berger (1873–1941), the inventor of the EEG, made the first EEG recording of an atypical absence in an 18-year-old woman suffering from both generalized convulsive and absence seizures for five years. This finding remained initially unpublished, because Berger suspected that the recorded activity contained artifacts caused by blinks and facial movements. Only in 1933, after other authors had described cortical potentials of large amplitudes, Berger published the EEG recording of this patient [33]. The characteristic pattern of “egg and dart” or “spike and dome” discharges in a patient with typical absence was first recorded during one of the night recording sessions in the EEG laboratory of the American physiologist Hallowell Davis at Harvard Medical School in December 1934. It was described in an article published the following year by the same Davis together with Frederic Gibbs and William Lennox [34].

Based on the EEG findings, in 1936 Frederic Gibbs (1903–1992) and William Lennox (1884–1960) could for the first time differentiate absence seizures characteristic of petit mal from psychomotor seizures occurring within the context of temporal lobe epilepsy [35]; the later introduction of video-EEG allowed a precise electro-clinical correlation of absence seizures [6,36].

7. The classification of typical absence seizures: the (hi)story so far

In 1981, the Commission on Classification and Terminology of the International League Against Epilepsy (ILAE) classified absence seizures among generalized seizures. It also differentiated typical absence seizures of idiopathic generalized epilepsies from atypical absence seizures occurring within the context of symptomatic generalized epilepsies [1]. Epilepsies with typical absences were not further classified, but continued to be grouped under the umbrella term “petit mal”, which was considered a form of “centrencephalic epilepsy” [37,38]. In 1989, the same Commission provided a list of idiopathic epilepsies (i.e., epilepsies without a well-defined etiology except possible genetic cause, precisely reflecting the classic medical term “idiopathic”) grouped according to onset age; epilepsies with typical absence seizures were hence distinguished into childhood absence epilepsy, juvenile absence epilepsy, and juvenile myoclonic epilepsy [39].

In 2017, the ILAE published an updated operational classification of seizure types: typical absence seizures have been listed and classified among generalized nonmotor (absence) seizures, and differentiated from other generalized onset or focal onset nonmotor seizures [2].

While the current and previous classifications of the ILAE is based on electroclinical characteristics, the semiological seizure classification proposed by Lüders and colleagues [40] relies exclusively on ictal semiology. According to this classification, seizures in which the main ictal manifestation is an alteration of consciousness should be termed “dialeptic seizures”. The term dialeptic, which is independent of associated ictal or interictal EEG changes, derives from the Greek word *διαλείπειν* (*dialeipein*, “to interrupt”), and refers to “episodes of unresponsiveness or decreased responsiveness that are not caused by motor alterations” and are inevitably “associated with complete or at least partial amnesia for the episode” [40]. According to this semiological classification, absence seizures should be referred to as “dialeptic seizures with a generalized ictal EEG”, whereas complex partial seizures as “dialeptic seizures with a focal ictal EEG” [40]. In the pre-EEG era only a seizure classification based on semiology alone was possible; the invention of EEG enabled a precise epileptic syndrome classification based on electroclinical features. Intriguingly, these historical aspects are recapitulated in the diagnostic process of any seizure with altered consciousness, when a semiological diagnosis is initially made (“dialeptic seizure”) prompting for an EEG recording for a precise epileptic syndrome classification.

8. Conclusions

From the early descriptions to the electroclinical correlation and classification, the fascinating history of absence seizures/petit mal reflects the history of epilepsy in general. Although the first clinical descriptions of this seizure type date back to the 17th Century, it was only the invention of the EEG in the 1930s that enabled a precise electroclinical evaluation and the differential diagnosis with other seizures with clouding of consciousness and only minor motor symptoms. Absence seizures continue to fascinate. The strong interest toward them has not vanished over time, as the recent research trends in genetic causes and pathogenesis clearly demonstrate [41–43]. The long story of absence seizures has not yet come to an end.

Conflicts of interest

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.yebeh.2018.01.007>.

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