Article

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CLINICAL AND ELECTROENCEPHALOGRAPHIC CHARACTERISTICS OF A COHORT OF PATIENTS WITH EPILEPSY AND ABSENCE SEIZURES

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Abstract – **Background:** Epileptic syndromes with absence seizures (AS) possess unique clinical and electroencephalographic (EEG) characteristics. In typical or atypical AS, ictal phenomenology may include various characteristics. Vídeo-EEG monitoring enables findings to be correlated with ictal phenomenology. **Objective:** To evaluate the different AS in a cohort of patients with drug-resistant epilepsy (DRE) based on the International League against Epilepsy (ILAE)'s 2006 classification, to correlate with ictal phenomenology recorded and to apply the Panayiotopoulos criteria. **Method:** This study included patients with criteria of AS followed up at the Epilepsy Clinic. A dual, cross-sectional cohort study was carried out between 2005 and 2008. Patients receiving care in the Epilepsy Program of the HUCFF-UFRJ, who had been investigated by video-EEG and who presented clinical and EEG criteria for absence seizures, typical or atypical, according to the criteria defined by the ILAE, were included in the study, independent of age onset, the review of clinical history, age onset, family history, epilepsy onset and evolution, seizures phenomenology, antiepileptic drugs response and neuroimaging studies were used to classify the patients among the different epileptic syndrome associated to absence seizures. **Results:** Typical absences were more frequent (71.4%) than atypical absences. Cases of juvenile absence epilepsy were the most frequent (19%) in this series, followed by childhood absence epilepsy (14.4%) and juvenile myoclonic epilepsy (4.8%). In 14 patients (66.67%), diagnosis was modified from focal epilepsy to primary generalized epilepsy. Clinical and EEG diagnosis of absence epilepsy resulted in a dramatic improvement in the control of seizures following modification of diagnosis and indication of an appropriate antiepileptic drug. **Conclusion:** Our results show that typical AS are more frequent than atypical. AS was successfully defined in 10 patients following application of Panayiotopoulos' criteria. The consequent change in diagnosis and therapy resulted in resolution of refractoriness in 9 patients. We concluded that in DRE, AS associated to unusual ictal phenomenology improve dramatically when diagnosed by video-EEG, permitting seizures to be controlled. Clinical and EEG evaluation confirm that myoclonus, automatisms and autonomic disorders are involved and that the consciousness may be affected to different degrees.

KEY WORDS: absence seizures, epilepsy, Panayiotopoulos.

Características clínicas e eletrencefalográficas de uma coorte de pacientes com epilepsia com crises de ausência

Resumo – Síndromes epilépticas com crises de ausência (CA) possuem características clínicas e eletroencefalográficas (EEG) únicas. Nas crises de ausência típica ou atípica, a fenomenologia ictal pode incluir características que podem levar ao erro diagnóstico e à indicação de drogas antiepilépticas que pioraram o quadro. Quando esses pacientes são referidos a um Programa de Epilepsias para investigação, a monitorização

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por vídeo-EEG permite correlacionar os achados eletrográficos com a fenomenologia ictal. **Objetivo:** Identificar em uma coorte de pacientes com epilepsia fármaco-resistente (EFR), pacientes com CA segundo critérios propostos pela Liga Internacional contra a Epilepsia (ILAE) de 2006, correlacionar a fenomenologia ictal ao EEG e aplicar os de critérios Panayiotopoulos neste grupo. **Método:** Estudo de corte transversal incluiu doentes encaminhados ao Programa de Epilepsia do HUCFF-UFRJ entre 2005 e 2008, investigados por vídeo-EEG e que apresentavam os critérios clínicos e EEG para CA típicas ou atípica; a revisão da história clínica, idade início, história familiar de epilepsia, evolução, a fenomenologia ictal, resposta a drogas antiepilépticas e estudos de neuroimagem foram utilizados para classificar os pacientes entre as diferentes síndromes epilépticas associadas a CA. **Resultados:** As CA típicas foram mais freqüentes (71,4%) do que as atípicas. Casos de epilepsia ausência juvenil ocorreram em 19% desta série, seguido por epilepsia ausência infantil (14,4%) e epilepsia mioclônica juvenil (4,8%). Em 14 pacientes (66,67%), o diagnóstico de epilepsia focal epilepsia foi modificado para epilepsia generalizada primária. A mudança do diagnóstico de epilepsia focal para epilepsia com CA, seguido da troca para DAE adequadas, resultou em melhoria no controle de crises. **Conclusão:** Nossos resultados mostram que as CA típicas são mais freqüentes do que as atípicas. Em 10 pacientes, a aplicação dos critérios de Panayiotopoulos foi possível. A conseqüente mudança no diagnóstico e terapêutica resultou na resolução de refratariedade em 9 pacientes. Concluímos que a fenomenologia ictal incomun em pacientes com CA contribui para o diagnóstico errôneo de epilepsia de difícil controle, e que o diagnostico por vídeo-EEG, permitiu a mudança do diagnóstico e a melhora dramática no controle das crises. As avaliações clínica e eletrencefalográfica confirmam que mioclonias, automatismos psicomotores e desordens autonômicas podem fazer parte da fenomenologia ictal de pacientes com CA e que a consciência pode ser afetada em diferentes graus.

PALAVRAS-CHAVE: crises de ausência, epilepsia, Panayiotopoulos.

Absence seizures (AS) are classified under the category of generalized epilepsies; however, unlike other seizures, absence seizures possess clinical characteristics and electroencephalographic (EEG) patterns that are specific for this diagnosis[1-3] and occur in different epileptic syndromes. Absence seizures may be divided into typical absences and atypical absences, according to EEG pattern. Absence seizures are present in a heterogenous group classified by the International League against Epilepsy (ILAE) into four epileptic syndromes: childhood absence epilepsy, juvenile absence epilepsy, juvenile myoclonic epilepsy and myoclonic absence epilepsy[4]. Typical absence seizures are most frequent and are characterized by loss of consciousness which is time-locked with bursts of bilaterally synchronous spike-and-wave discharges of 3 to 4 cycles per second, and, in general, present good response to pharmacological treatment. Conversely, atypical absence seizures are less frequent, the seizures are associated with 1 to 2 cycles per second spike-wave discharges and slow background rhythms for aging; they are often associated with severe neurologic impairment and poor response to treatment. The criteria proposed by Panayiotopoulos in 1997 for the classification of patients with typical absence seizures include a greater number of absence syndromes, some of which have not yet been recognized by the ILAE, such as phantom absences, absence epilepsy with eyelid myoclonus and early-onset absence epilepsy, among others[5].

Epilepsies with typical absence seizures are common forms of generalized epilepsy. Cases of childhood absence epilepsy constitute 2–10% of all cases of epilepsy in children. Juvenile absence epilepsy constitutes 10% of all cases of idiopathic generalized epilepsy, whereas absence epilepsy with eyelid myoclonus makes up 2% of all epilepsies and 11% of all cases of idiopathic generalized epilepsy. It is difficult to determine the frequency of the rarer syndromes[6]. Epilepsy was classified into focal and generalized forms rather more for didactic purposes than from any intention to define the localization of the disease, bearing in mind that various authors have questioned the essentially bilateral onset of the generalized forms of epilepsy, believing all to have a focal onset[7,8]. Interictal EEG may result in an incorrect diagnosis of the type of seizure and/or epileptic syndrome, following which therapy may consequently fail[7], particularly in cases of patients with generalized epilepsy in whom clinical characteristics are suggestive of focal epilepsy during seizures or as a result of focal patterns revealed by interictal EEG.

The objective of this study was to evaluate with what frequency patients referred to the Epilepsy Program of the *Clementino Fraga Filho* Teaching Hospital, Federal University of Rio de Janeiro (HUCFF-UFRJ) with an initial diagnosis of drug-resistant epilepsy present absence seizures, analyze what clinical and electroencephalic variables were associated with drug-resistance, and reclassified this patients according to Panayiotopoulus criteria.

METHOD

A dual, cross-sectional cohort study was carried out between 2005 and 2008. Patients receiving care in the Epilepsy Program of the HUCFF-UFRJ, who had been investigated by vid-

eo-EEG and who presented clinical and EEG criteria for absence seizures, typical or atypical, according to the criteria defined by the ILAE, were included in the study, independent of age onset; the review of clinical history, age onset, family history, epilepsy onset and evolution, seizures phenomenology, antiepileptic drugs response and neuroimaging studies were used to classify the patients among the different epileptic syndrome associated to absence seizures. Patients included could present other seizures type, besides absence seizures. All the seizures were registered and the concomitance of different seizures phenomenology and EEG pattern were analyzed. The video-EEG monitoring was done for at least 24 h, with a Neurotec® machine of 32 channels, and the scalp electrodes were distributed according to the 10–20 system. Patients with focal epilepsy and other generalized epilepsy without the presence of absence seizures were excluded. The clinical, ictal phenomenology and EEG variables were used to reclassify the patients according to Panayiotopoulus criteria analyzing the different epileptic syndromes with absence seizures.

This study was approved by the Institutional Review Board and all patients signed an informed consent form prior to admission. The clinical data and correlated variables recorded during video-EEG were stored on an Excel spreadsheet (Excel 1997–2003) and transformed into a DBF file to enable them to be read in the generally available Epi Info program, version 6.0.

RESULTS

Of a cohort of 643 patients receiving care between 2005 and 2008, 68 (10.57%) were found to have clinical and EEG criteria for generalized epilepsy and, of these, 21 (30.09%) had absence seizures. In these 21 patients, 2 presented clinical suspicion of generalized epilepsy according to seizures semiology and triggering factors described. The mean age of patients at the time of inclusion into the study was 25.5 years (range 3–58 years). The mean age at onset of symptoms was 11.8 years (range 1–39 years).

Of these 21 patients, the onset of symptoms occurred before the patient reached 10 years of age in 10 patients (47.6%), between 11 and 20 years of age in another 10 (4.76%) and after 20 years of age in 1 patient (4.76%). Eight of these patients (38.09%) were male. Twelve (57.1%) were white, while 9 (42.85%) were of African descent. The mean duration of the disease was 14.4 years (range 1–42 years). Six patients (28.6%) had a family history of epilepsy, while 13 (61.9%) did not and 2 (9.5%) were unable to provide any information on the subject.

Of the 20 patients in use of antiepileptic drugs, 8 (40%) were in use of specific first-line antiepileptic drugs for absence seizures and in 4 of these cases (50%), these drugs were associated with other antiepileptics that are contraindicated for absence seizures, including: carbamazepine (CBZ) (n=2), phenytoin (PHT) (n=2) and oxcarbazepine (OCX) (n=2). Of the 4 remaining patients (50%) using specific first-line drugs for absence seizures, 1 patient (25%) was using the drug as monotherapy, while the remaining 3 (75%) were using the drug in association with another antiepileptic drug that was not contraindicated for absence seizures. A total of 14 patients (70%) were using antiepileptic drug that were contraindicated for absence seizures. In 4 of these cases (28.58%), the contraindicated antiepileptic drug was associated with a specific first-line drug for absence seizures, while in 5 patients (35.71%) the drug was associated with a second contraindicated drug, in another 5 (35.71%) the drug was associated with a second-line antiepileptic drug and in 2 patients (14.28%) the contraindicated medication was being used as monotherapy. One patient without AED in the moment of the video-EEG monitoring fulfilled criteria to pharmacoresistant epilepsy but had interrupted the treatment by himself after worsening with carbamazepine and phenytoin.

Of the 14 patients in use of antiepileptic drugs that are contraindicated for absence epilepsy, the drug could not be withdrawn in 3 cases (21.42%). However, in 7 patients (50%), complete control of the seizures was achieved following withdrawal of the deleterious drugs, while in four cases (28.56%) partial control was achieved, although occasional absence seizures persisted.

Of the four patients who were in use of specific first-line antiepileptic drugs for absence seizures, adjustment of the dose was effective in 1 patient (25%). In another patient (25%), in addition to adjustment of the dose, another antiepileptic drug had to be associated, a strategy that led to a reduction in the frequency of seizures. Despite all the relevant measures taken, seizures persisted in two patients.

Neuropsychological and motor development was normal in 18 patients (85.7%) and abnormal in 3 (14.3%). In 17 patients (81.0%), there was no loss of function following the onset of seizures, whereas in 4 (19%) there was a decline in neuropsychological and motor development.

Two patients (9.52%) had only absence seizures, while 16 (76.19%) had generalized tonic-clonic seizures, 6 (28.57%) had appendicular myoclonus, 5 (23.80%) had eyelid myoclonus with absences and 4 (19.04%) had astatic seizures. Of the 21 patients, 2 (9.52%) had one single type of seizure, 11 (52.38%) had two associated types of seizure, 5 (23.80%) had three associated types of seizure, 2 (9.52%) had four associated types of seizure and 1 patient (4.76%) had five different types of seizure. Four patients (19.04%) had a previous history of *status epilepticus*, 3 of which (75%) consisted of absence epilepsy and 1 case (25%) of tonic clonic seizures (Table 1).

During the absence seizures, 5 patients (23.80%) had no motor phenomena, 10 (47.61%) had eyelid myoclonus, 7 (33.33%) had appendicular myoclonus, 3 (14.28%) had associated automatisms (Fig 1) and 2 (9.52%) had autonom-

Table 1. Seizures phenomenology and antiepileptic drug response.

Patient	Other associated seizures	Absence seizures concurrent phenomenology	AED response
1		Absence and eyelid myoclonus masticatory automatism	
2	Generalized tonic clonic	Absence and appendicular myclonus	Worsening after CBZ
3	Eyelid myoclonus Generalized tonic clonic and appendicular myoclonus	Absence and eyelid myoclonus	Worsening after CBZ
4		Absence and eyelid myoclonia, oral and tongue automatism, upper and lower limbs automatism	Worsening after CBZ
5	Generalized tonic clonic, tonic and astatic	Absence with upper and lower limbs automatism	
6	Absence epilepsy, Generalized tonic clonic and eyelid myoclonus	Absence and eyelid myoclonus, sometimes with epigastric discomfort	Worsening after CBZ
7	Generalized tonic clonic	Brief absences	
8	Astatic seizures and focal seizures	Absence and eyelid myoclonus, and astat seizures	CBZ and no worsening
9	Absence and upper and lower limbs myoclonus	Absence and upper and lower limbs myoclonus	Worsening after CBZ
10	Generalized tonic clonic	Absences and eyelid myoclonus	Worsening after CBZ
11	Generalized tonic clonic	Brief absences	
12	Generalized tonic clonic, astatic seizures	Absences and eyelid myoclonus, appendicular myoclonus, sometimes astatic seizures	
13	Generalized tonic clonic, release sphincteric	Absences and episodic confusional states	Worsening after CBZ
14	Generalized tonic clonic, episodic confusional states and aggression	Brief absences	Worsening after CBZ
15	Generalized tonic clonic preceded by upper and lower limbs myoclonus	Absences with appendicular myoclonus	Worsening after CBZ
16	Generalized tonic clonic, confusional sates episodic, eyelid, upper and lower limbs myoclonus	Absence associated with eyelid and appendicular myoclonus	
17	Generalized tonic clonic, upper and lower limbs myoclonus	Absence, sometimes associated to masticatory movments	Worsening after OXCBZ
18	Generalized tonic clonic	Brief absences	
19	Eyelid myoclonus	Absences and eyelid myoclonus	Worsening after CBZ
20	Generalized tonic clonic, eyelid, upper and lower limbs myoclonus, astat seizures	Absences and eyelid myoclonus	
21	Generalized tonic clonic	Brief absences	

AED: antiepileptic drug; CBZ: carbamazepine; OXCBZ: oxcarbazepine.

ic abnormalities, while in 1 patient (4.76%) there was a report of sphincter release and in another (4.76%) a report of epigastric discomfort.

Ictal EEG findings were characteristic of generalized seizures, fulfilling the criteria for typical or atypical absence seizures in all patients. In 15 patients (71.4%), video-EEG findings fulfilled the criteria for typical absence seizures (Fig 1) and in 6 patients (28.6%) findings were characteristic of atypical absence seizures (Fig 2). Interictal EEG findings showed focal electroencephalographic elements in 14 patients (66.66%), in combination with diffuse slowing of the brain electrical activity in one case (7.14%) and generalized spike wave patterns in another (7.14%). Five patients (23.80%) had fragments of electroencephalographic

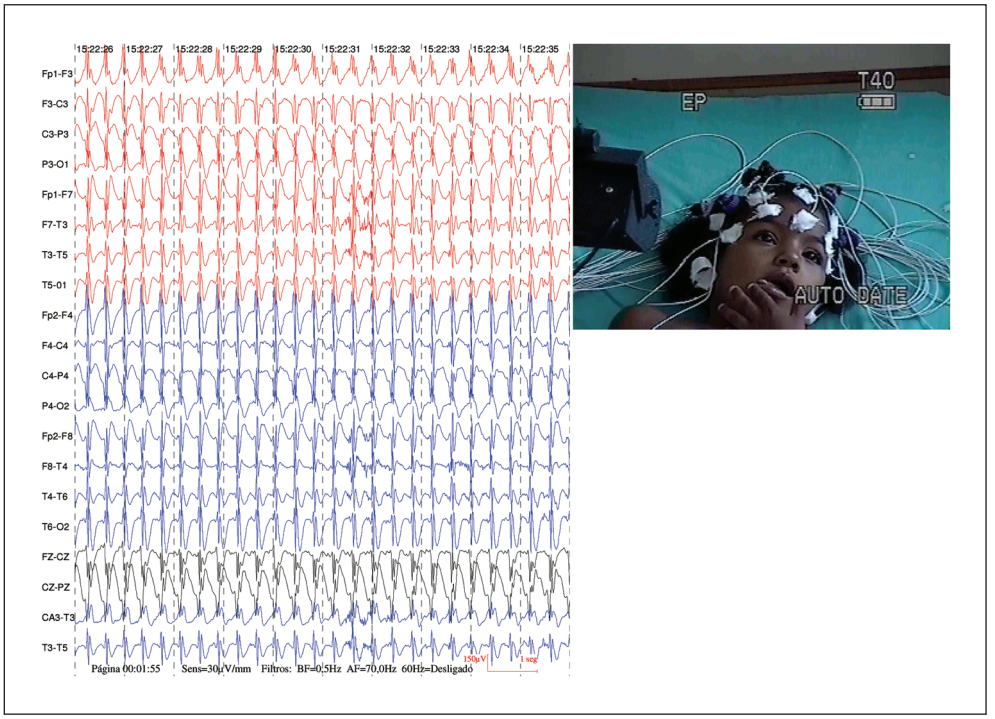

Fig 1. Registered ictal video-electroencephalographic showing complex of spike slow waves rhythmic of 3 Hz, high amplitude, with projection widespread, synchronous and symmetrical, with duration of 18 seconds. Clinically, seizures of absence with eyelid myoclonia, perioral and language automatism.

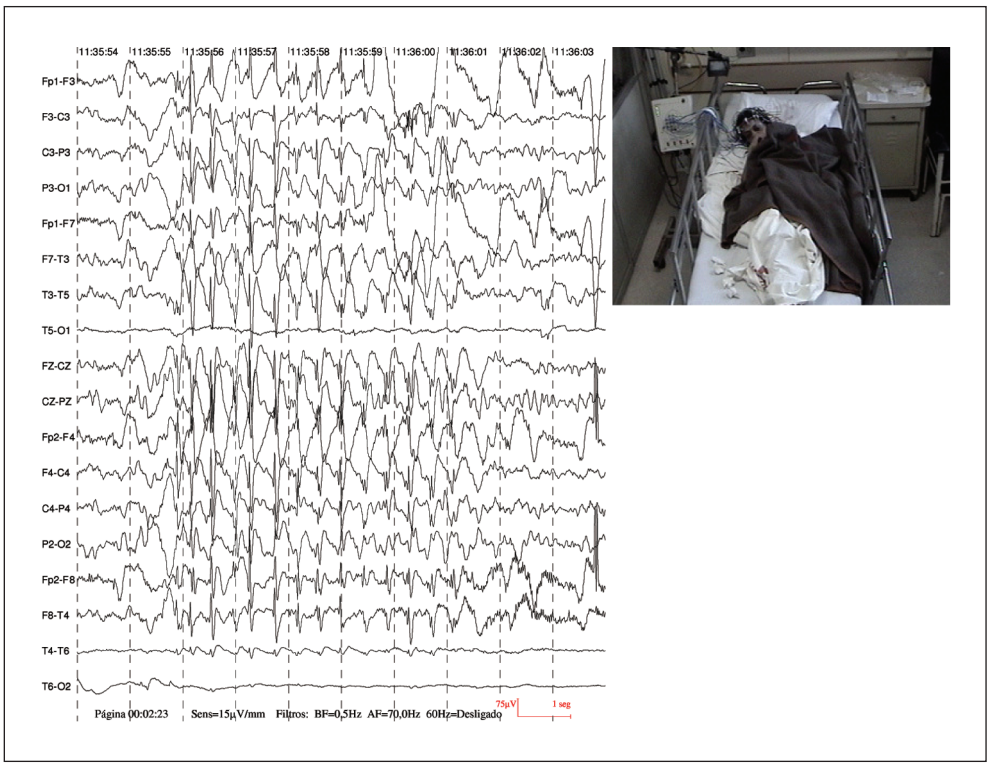

Fig 2. Record video-electroencephalographic evidence outbreak of spike wave complex and poly spike wave complex, about 2.5 Hz, high amplitude, with widespread projection.

Table 2. Results of ictal and interictal video-EEG.

Patient	Interictal vídeo-EEG	Ictal vídeo-EEG	Hiperpnea	Light stimulus	Diagnosis before Pann Criteria	Diagnosis after Pann Criteria applyed
1.	Irregular slow waves, acute frontal waves temporal left	Complex spike slow wave 3 Hz	++	–	JAE	JAE
2.	Spike complex and poli spike slow wave frontal and central bilateral	Spike complex, irregular and generalized poli spike slow wave (2.5–3 Hz)	++	–	JME	JME
3.	Intermittent slow wave frontal right	Spike complex and poli spike slow wave intermittent generalized (±4 Hz)	–	++	NC	ABEEM
4.	Occipital slow wave	Complex spike slow wave generalized 3 Hz	++	–	EAI	NC
5.	Diffuse slow track, intermittent slow waves on frontal left	Complex spike slow wave generalized 2 Hz and generalized slow wave acute wave 2 Hz	NA	–	AA	AA
6.	CE and poli spike slow wave fronto central	CE and intermittent poli spike slow wave generalized 3–4 Hz	–	–	CAE	Phantom absences
7.	CE and poli spike slow wave fronto temporal	CE e intermittent poli spike slow wave generalized 3–4 Hz	++	–	NC	Phantom absences
8.	Irregular slow waves on temporal left	Complex spike slow wave generalized 1.5–2 Hz	+	–	AA	NC
9.	Irregular acute wave and slow wave	Complex spike slow wave generalized 3–4 Hz	++	–	JAE	NC
10.	CE e poli spike slow wave on frontal left	CE intermittent poli spike slow wave generalized 3–4 Hz	++	++	JAE	AEEM
11.	Irregular slow wave generalized	Intermittent complex spike slow wave generalized	–	+	NC	Phantom absences
12.	Diffuse slow track; complex spike slow wave fronto-central	Complex spike slow wave generalized 2–2.5 Hz	++	–	AA	NC
13.	Complex spike slow wave on frontal right	Intermittent complex spike slow wave generalized 3–4 Hz	–	–	NC	Phantom absences
14.	Irregular slow waves on parietal right	CE and poli spike slow wave generalized 2.5 Hz	+	+	NC	NC
15.	EF and slow wave/acute wave on frontal and temporal left	CE and intermittent poli spike slow wave generalized	+	+	JME	NC
16.	Irregular slow waves on fronto-temporal; generalized spikes	Generalized spikes and generalized poli spike slow wave	NA	–	AA	NC
17.	Fronto temporal spikes on right	Intermittent generalized spike slow wave complex 2 Hz	+	–	AA	NC
18.	Acute and slow waves on fronto-temporal left	CE and generalized poli spike slow wave 3 Hz	NA	–	NC	EAIP
19.	Irregular acute and slow wave on temporal left	Generalized spike slow wave complex 3 Hz	–	–	CAI	NC
20.	Irregular slow waves on fronto-temporal	generalized spike slow wave complex 1.5–2.5 Hz	+	+	AA	NC
21.	Irregular acute slow wave fronto-central rigth	Generalized spike slow wave complex 3 Hz	+	+	JAE	JAE

JAE: juvenil absence epilepsy; JME: juvenile myoclonic epilepsy; NC: not classified; EMAE: early myoclonic absence epilepsy; AEEM: absence epilepsy with eyelid myoclonus; CAE: childhood absence epilepsy; AA: atypical absence.

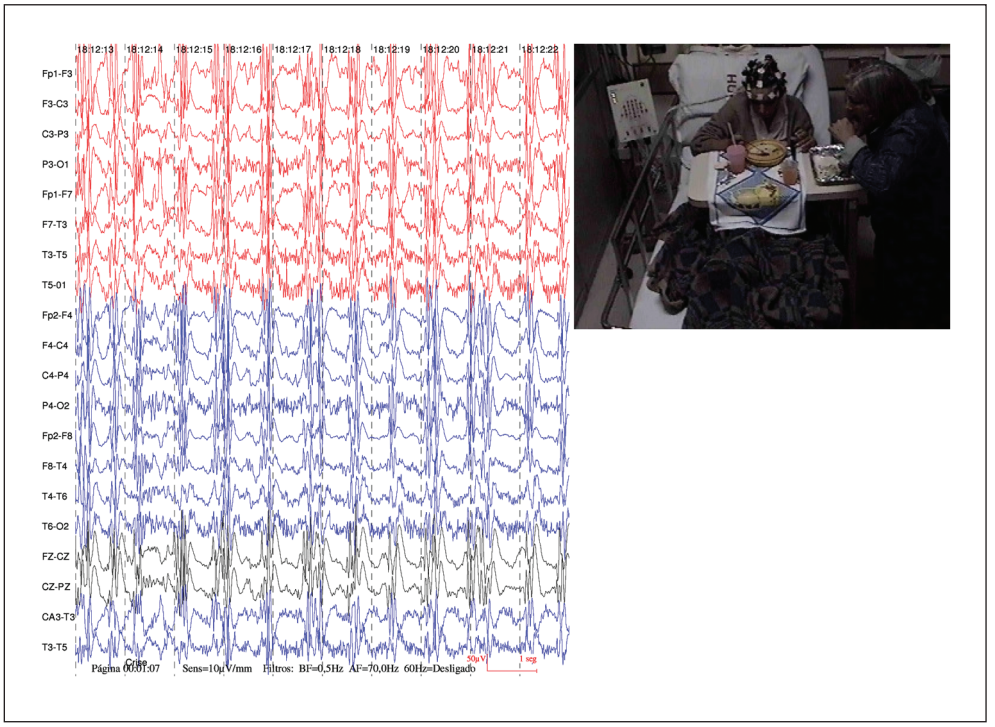

Fig 3. Registration ictal video-electroencephalographic evidence outbreak of poly spike wave complex, approximately 1.5 Hz, high amplitude, with widespread projection. Clinically patient remains lunching with difficult in organizing movements for eating.

elements with a predominance of morphological characteristics similar to focal ictal features in a normal interictal tracing. Of these patients, 4 (80%) had spike complexes and polyspike-slow wave complexes, and 1 patient (20%) had spike-slow wave complex. In one patient (4.76%), interictal recording showed slow wave and intermittent, generalized spike wave activity and in another patient (4.8%), slowing of background activity in the EEG tracings was found in association with spike-slow wave complex (Table 2).

Ictal recording of 12 patients (57.4%) with spike-slow wave complexes showed regular ictal discharge in 7 (58.3%) and irregular ictal discharge in 5 (41.7%). Of the 12 patients with spike-slow wave complex, frequency was >2.5 Hz in 7 (58.4%) and ≤ 2.5 Hz in 5 (41.7%) (Table 2).

Of the 9 patients (42.8%) in whom ictal recording registered spike complexes and polyspike-slow wave complexes, ictal activity was regular in 2 cases (22.2%) and irregular in 7 (77.8%). In 3 patients (33.3%), frequency was >2.5 Hz, while in 1 patient (11.1%), frequency was ≤ 2.5 Hz, and in another (11.1%) frequency varied between >2.5 Hz and ≤ 2.5 Hz during seizures. In 2 patients (22.2%) with spike complexes and polyspike-slow wave complexes < 2.5 Hz, there was an association of spike-wave and slow-wave paroxysms (Table 2).

Of the 18 patients (85.7%) in whom hyperpnea was successfully accomplished, 13 (72.2%) had abnormalities; 6 (46.2%) had accentuated electroencephalographic elements and seizures were triggered in another 7 (53.8%). Seven patients (33.3%) had an abnormal EEG recording because of photic stimulation, 5 of these patients (71.4%) having accentuated electroencephalographic elements while 2 (28.6%) developed seizures as a result of photosensitivity. In three patients (14.3%), the presence of eyelid myoclonus was registered following ocular occlusion in an illuminated environment, with absences in one patient.

Of the 21 patients evaluated, 15 (71.42%) fulfilled clinical and EEG criteria for typical absences and 6 patients (28.58%) for atypical absences. Of the 15 patients of the group with clinical and EEG characteristics of typical absences, 9 (60%) were diagnosed with an epileptic syndrome, while 6 (40%) were not classified as having any of the syndromes described by the ILAE. Of the 9 patients with an established epileptic syndrome, 5 (55.6%) were classified as juvenile absence epilepsy, 3 (33.3%) as childhood absence epilepsy and 1 (11.1%) as juvenile myoclonic epilepsy. After applying Panayiotopoulos' criteria, 10 patients (66.7%) were able to be diagnosed with specific syndromes, while 5 (33.3%) failed to fulfill the criteria for any specific syndrome. Of the 10 patients with a specific classification, 2 (20%) retained the diagnosis of juvenile absence epilepsy and 1 (10%) of myoclonic absence epilepsy, while 4 (40%) were classified as phantom absences (Fig 3), 2 (20%) as absence epilepsy with eyelid myoclonus and 1 (10%) as early-onset absence epilepsy.

DISCUSSION

The present results show that some absence seizures may include ictal phenomena with both simple and complex motor automatisms, confounding diagnosis and leading to inappropriate therapy, as shown by Lombroso, Ferrie and Holmes, and Brown and Tucker[7,8,13]. It can justify why patients with primary generalized seizures were referred to our video-EEG unit.

The rate (10.57%) of patients with drug-resistant epilepsy at the HUCFF-UFRJ was lower than figures reported in the literature[6,14] and may be attributable to the low rate of diagnosis or to the age group of the cohorts in those studies. In the present study, this finding may perhaps reflect the large number of patients with focal epilepsy referred to epilepsy programs such as that offered at the HUCFF, which receives a greater percentage of patients with supposedly focal seizures.

The age at onset of absence seizures, which ranged from 1 to 39 years, is justified by the inclusion of patients with different epileptic syndromes in which absence seizures are found. Each one of the syndromes is characterized by a certain age at onset according to the respective age-group. Another factor that may explain this wide range is the inclusion of patients with epileptic syndromes involving absence seizures that have not yet been recognized by the ILAE. These syndromes have, however, been the subject of recent studies and their inclusion into the ILAE classification has been proposed by various investigators[1,5,9]. One example is the patient in whom onset of absence seizures occurred in adulthood (at 39 years of age), a case similar to others previously reported by Trinka[10]. Another example was the patient with early onset at 12 months of age, other investigators also having reported similar cases[9,10]. Despite the disparate variation in age at onset, the mean age of patients at the onset of absence seizures in the present study was 11.04 years, which is in agreement with data reported from other studies on juvenile absence epilepsy[12].

There was a predominance of females in the present study, which is in agreement with findings reported in the literature, principally in studies on generalized epilepsies with absence seizures[13]. There may be a predominance of males in cases of epilepsy with myoclonic absences[13].

The duration of epilepsy found in this study was longer than expected. This finding is justified by the fact that patients with drug-resistant epilepsy require referral to specialized treatment centers as early as possible, as emphasized in the literature[14,15]. Erroneous initial diagnosis and the consequent inefficacy of the prescribed antiepileptic drugs was the most frequent finding. Syndromes such as atypical absences, which have a natural history of refractoriness to treatment, were another cause of drug-resistant epilepsy[16,17].

The present results are in agreement with reports in the literature where in some cases a family history of the condition was common and various different types of seizure were found in the same family[3,18].

In 14 patients (70%) in this series, an increase was found in the frequency of epileptic seizures that was attributed to the use of antiepileptic drugs that are contraindicated for patients with absence seizures, thereby confirming the importance of the new ILAE classification[19], which now includes selective response to antiepileptic drugs as part of the classification criteria[2,20-22]. Incorrect diagnosis based on the presence of apparently focal characteristics[23,24], as well as interictal EEG scans with focal abnormalities[7], were also found in our patients and were confounding factors in the differential diagnosis with focal epilepsy. Indeed, differential diagnosis was only possible with the aid of video-EEG.

In the present series, four patients had limb and orofacial (mouth and tongue) automatisms highly suggestive of temporal epilepsy but which occurred during absence seizures and were correlated with EEG showing 3c/s spike-slow wave complex or spike complex and generalized polyspike-slow wave. In another 9 patients, the presence of eyelid myoclonia was confused with facial automatisms or considered to constitute an event unrelated to epilepsy. In 8 patients, the presence of appendicular myoclonus was confused with temporal lobe automatisms. The diversity in the types of seizures found in patients with absence seizures in the present study is in agreement with reports in the literature[13,23,24]. This heterogeneity in the absence seizures found in patients with generalized epilepsy has been confirmed by other authors[6,10,12,13,23,24].

The finding of absence epilepsy in our patients is in agreement with reports in the literature that patients with absence seizures, albeit apparently asymptomatic, may progress to absence epilepsy or generalized tonic-clonic seizures[5,25-27].

In the present study, a greater association of atypical absence seizure was found in patients with a history of function loss following the onset of seizures, mental retardation and drug-resistant epilepsy. This finding is in agreement with the literature, confirming that atypical absence seizures are associated with a poorer prognosis with respect to the control of seizures, to impaired cognition and neuropsychological and motor development and to a greater variation in the types of seizures[13,16,17]. In the present study, patients with atypical absences had seizures associated with abnormal muscle tone, either hypo or hypertonia, and astatic seizures were more frequent in this group compared to reports from other studies published in the literature[13,16,17,28].

A significant difference was found between the classification of patients in the present series according to the definitions of the ILAE[4,19,29] and their posterior classifica-

tion according to the criteria proposed by Panayiotopoulos et al.[1,5] According to Panayiotopoulos[2], the classification of a specific syndrome is in agreement with the literature when application of the criteria used results in an increase in that classification category and a consequent improvement in the therapeutic options available to the respective patients[1,5,24,30].

We attribute the large rate of cases of phantom absences in this series to the fact that these patients had seizures that were difficult to diagnose, this type of referral being commonplace at specialized centers.

Analysis of this series of patients with absence seizures demanded a meticulous review of the currently proposed diagnostic criteria and showed that a careful evaluation of patients with drug-resistant epilepsy may result in a change in the initial diagnosis to a controllable form of epilepsy that is not drug-resistant. In almost 70% of the cases in the present series, modification and adaptation of the diagnosis of the epileptic syndrome or of the type of seizure altered therapeutical management and the quality of life of these patients and their families, confirming the importance of establishing referral centers for the investigation and treatment of epilepsy.

In conclusion, in patients with epilepsy with typical or atypical absence seizures, ictal phenomenology may include various characteristics, and clinical and EEG evaluation confirms that myoclonus, automatisms and autonomic disorders are involved and that the consciousness of the patient may be affected to different degrees. Typical absences are more frequent (71.4%) than atypical absences. Cases of juvenile absence epilepsy were the most frequent (19%) in this series, followed by childhood absence epilepsy (14.4%) and juvenile myoclonic epilepsy (4.8%). In 14 patients (66.67%), diagnosis was modified from focal epilepsy to primary generalized epilepsy. AS was successfully defined in 10 patients following application of Panayiotopoulos' criteria. Clinical and EEG diagnosis of absence epilepsy resulted in a dramatic improvement in the control of seizures following modification of diagnosis and indication of an appropriate antiepileptic drug.

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