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## SHORT COMMUNICATION

### Frontal Absence Seizures: Clinical and EEG Analysis of Four Cases

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**A**CCORDING to the commission of International League Against Epilepsy (ILAE) in 1989, typical absence seizures are generalized seizures, characterized by transient impairment of consciousness (TIC) and generalized >2.5 Hz spike and slow wave discharges in ictal EEG.<sup>1</sup> Absence seizures with focal onset have been reported occasionally, such as frontal, temporal or parieto-occipital origin.<sup>2-3</sup> Lagae *et al*<sup>4</sup> identified a subgroup of children with absence epilepsy as frontal absence, whose ictal electroencephalograms (EEGs) showed generalized spike and slow wave preceded by frontal discharge. We presented four cases of frontal absence, with one case of frontal absence status epilepticus in them. The differences between typical and frontal absence seizures in clinical context and EEG were discussed.

From July 2009 to Jan 2015, four patients were diagnosed with frontal absences in our department. The diagnosis was made according to the following: 1) clinically characterized by recurrent transient impairment of consciousness; 2) the ictal EEG showed generalized >2.5 Hz spike and slow wave preceded by frontal discharges. Three were females and one was male, with age ranged from 5 to

42 (mean  $18.0 \pm 16.4$ ) years. The age of onset ranged from 1 month to 18 years (mean  $7.5 \pm 7.7$  years).

All patients underwent history taking, physical examination, brain MRI and continuous video-EEG monitoring (VEEG). After administration of antiepileptic drugs (AEDs), they were all followed up for one year. The electro-clinical features were analyzed and compared with typical absence seizures.

The clinical information and EEG characteristics are summarized in Table 1. The first three cases were characterized by recurrent, brief episodes of staring, behavior and speech arrest. Besides, the first and the third case had some motor signs during the ictus. The second case could walk at 18 months old and speak at 5 years old. The first two cases had retardation in intellectual development.

The fourth case was a prolonged absence seizure. Seizure semiology usually started with palpitation. She complained of limb weakness, but she could lift her arms and legs on command. Soon after, she showed a slowness of behavior, accompanied by staring, blinking, smacking, fumbling and involuntary movements of limbs. During the ictus, she could not speak most of the time, but she could follow simple instructions on repeated requests. The episode could last several days and recurred several times a month. If diazepam was given, the episode could be terminated in half an hour. Afterwards, she could only

recollect what had happened in the first a few minutes. The interictal and ictal EEG of the fourth case are described in (Fig. 1).

All the four cases were resistant to AEDs treatment. In case 1, there was no improvement after the administration of Sodium Valproate, Clonazepam and Levetiracetam. As for case 2 and case 3, after oral administration of daily 1000 mg Sodium Valproate together with 2 mg Clonazepam, and daily 12 ml Sodium Valproate, respectively, remarkable reductions in seizure frequency were achieved after two or three months. The fourth case benefited from daily 800 mg Carbamazepine and 1000 mg Sodium Valproate. But none of them had attained a complete remission during 12 months of follow-up.

Lagae *et al*<sup>4</sup> has concluded some differences between typical and frontal absence group. The frontal absence group has more complex absences, which means staring associated with motor phenomena. Besides, high incidence of learning and behavioral problems, poor response to standard treatment and worse prognosis have been observed in frontal absence group. In EEG, more patients of frontal absence group have background slowing and inter-

ictal isolated frontal spikes than those in typical absence group, who usually have normal background and interictal single or short-range generalized 2.5 Hz-4 Hz spike and slow wave.<sup>4-6</sup>

In this study, the delayed intellectual development in first two cases and the resistance to standard AEDs treatment in all the four cases were consistent with Lagae's findings.<sup>4</sup> In addition, compared with childhood absence epilepsy which occurs in otherwise normal children of school age (peak at 6-7 years),<sup>1</sup> both case 1 and case 3 had relatively earlier age of onset, and the episodes occurred as frequent as up to 30 events a day, which was in accordance with the clustering feature of frontal lobe epilepsy.

The fourth case was diagnosed as frontal absence status epilepticus (SE). Compared with typical absence SE which usually occurs in patients with idiopathic generalized epilepsy as reported,<sup>7-8</sup> she has mild impairment of consciousness and long duration of episode. Aphasia is prominent during the ictus, which is common in frontal lobe epilepsy. Carbamazepine combined with Sodium Valproate was effective in seizure control in this case, which supported the diagnosis of focal onset absence SE.

**Table 1.** Summary of clinical data of the four cases

|        | Age on referral          | Age of onset                                   | Gender                                 | Perinatal history | Seizure semiology                                                 | Duration                                                          | Frequency            |
|--------|--------------------------|------------------------------------------------|----------------------------------------|-------------------|-------------------------------------------------------------------|-------------------------------------------------------------------|----------------------|
| Case 1 | 12 yrs                   | 1 month                                        | Female                                 | Uneventful        | TIC, eye blinking and head bending                                | 10-20 seconds                                                     | 10-15 times a day    |
| Case 2 | 13 yrs                   | 8 yrs                                          | Female                                 | Uneventful        | TIC                                                               | 10-20 seconds                                                     | 20-30 times a day    |
| Case 3 | 5 yrs                    | 4 yrs                                          | Male                                   | Uneventful        | TIC, eye blinking and hands automatism                            | 20-40 seconds                                                     | About 10 times a day |
| Case 4 | 42 yrs                   | 18 yrs                                         | Female                                 | Uneventful        | Slow reaction, automatism, involuntary movements and aphasia      | Several days                                                      | 1-2 times a month    |
|        | Intellectual development | AEDs treatment                                 | Response to AEDs                       | Brain MRI         | Spike and slow wave of interictal EEG                             | Spike and slow wave of ictal EEG*                                 |                      |
| Case 1 | Retarded                 | Sodium valproate, clonazepam and levetiracetam | No improvement                         | Normal            | Intermittent, bilateral frontal leads, prominent on the left side | 3-3.5 Hz, in left frontal leads for 1-2 seconds                   |                      |
| Case 2 | Retarded                 | Sodium valproate and clonazepam                | Episode decreased to 3-5 times a day   | Normal            | Intermittent, left frontal leads                                  | 3 Hz, in left frontal leads for 1-3 seconds                       |                      |
| Case 3 | Normal                   | Sodium valproate                               | Episode decreased to 3-4 times a month | Normal            | Intermittent, bilateral frontal and temporal leads                | 3-3.5 Hz, in bilateral frontal and temporal leads for 2-3 seconds |                      |
| Case 4 | Normal                   | Carbamazepine and sodium valproate             | Episode decreased to 3-5 times a year  | Normal            | Intermittent, predominant in right frontal and central leads      | 3-4 Hz, in bilateral frontal and central leads for 26 minutes     |                      |

yrs: years old; TIC: transient impairment of consciousness; AEDs: antiepileptic drugs; EEGs: electroencephalograms;

\* Preceding bilateral, symmetrical and synchronous discharge.

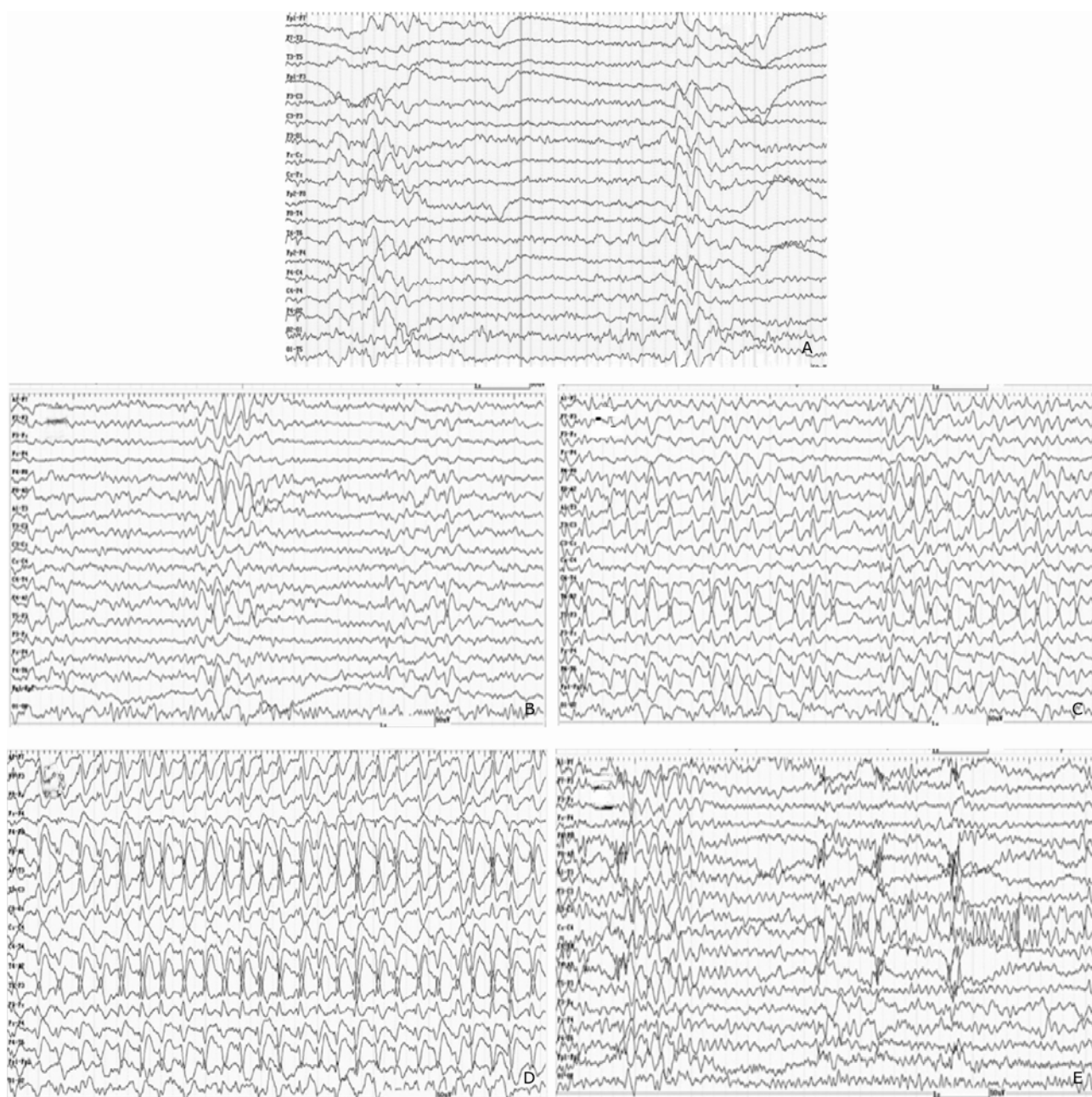


**Figure 1.** EEG of the case 4.

- A. Interictal EEG: Intermittent high amplitude 3-4 Hz spike and slow wave in bilateral frontal and central leads with right-sided predominance, under a background of 8 HZ  $\alpha$  wave.
- B. Ictal EEG when the abnormal epileptic discharge just started, showing frequent high amplitude 3-4 Hz spike and slow wave in bilateral frontal and central regions, which was prevalent in the right side. The patient had no clinical symptoms at this moment.
- C. EEG of 19 minutes after the initiation of abnormal epileptic discharge, showing high amplitude 3-4 Hz spike and slow wave, prominent in bilateral frontal and central regions. The patient began to complain of palpitation and limb weakness, accompanied by automatisms and slow reaction.
- D. EEG of 26 minutes after the initiation of abnormal epileptic discharge, the focal onset abnormal discharge was followed by bilateral, symmetrical and synchronous high amplitude 3Hz spike and slow wave. The patient was not able to recall what had happened during this period.
- E. EEG of 6 hours after complete remission of clinical symptoms, showing intermittent bilateral synchronous high amplitude 5 Hz  $\theta$  wave. The generalized epileptiform discharges gradually subsided. She could move and speak as usual at this time.



The ictal EEGs in these cases showed secondary generalized 3-3.5 Hz spike and slow wave preceded by epileptic discharge in frontal regions; the interictal EEGs showed focal abnormality in frontal leads. The EEG disturbances verified the diagnosis of focal onset absence seizures originating from frontal lobes, instead of generalized absence seizures.

Frontal absence is supposed to be related to secondary bilateral synchrony (SBS), described initially by Tukel and Jasper.<sup>9</sup> The epileptic discharge arises from frontal cortices and rapidly spreads to bilateral hemispheres through corpus callosum. Besides, SBS can also arise from temporal, parietal, and occipital regions. The thalamus may play a role in secondary bilateral synchronous discharges.<sup>9-11</sup>

In conclusion, frontal absences differ from typical absence seizures in clinical context, EEG, response to AEDs and prognosis. To thorough understand differences between typical and focal onset absence seizures might be meaningful in their surgical and pharmacological treatment. Electrode implantation should be considered when it is crucial to identify the source of absence seizures in the treatment.

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