

# Adult absence semiology misinterpreted as mesial temporal lobe epilepsy

Rebecca Hurst<sup>1,2</sup>, Nicole Chiota-McCollum<sup>1,3</sup>, William Tatum<sup>1</sup>

<sup>1</sup> Department of Neurology, Mayo Clinic, Jacksonville, FL

<sup>2</sup> Department of Neurology, Duke University, Durham, NC

<sup>3</sup> Department of Neurology, Keesler Air Force Base, Biloxi, MS, USA

Received February 27, 2014; Accepted September 16, 2014

**ABSTRACT** – Correct diagnosis of seizure type and epilepsy syndrome is the foundation for appropriate antiepileptic drug selection. Inappropriate medication choices occur in the treatment of generalized epilepsy and may aggravate some seizure types, including absence seizures, potentially leading to pseudo-drug resistance. Fortunately, a correct diagnosis of absence seizures is usually not difficult, though rarely demonstrates electroclinical overlap with focal seizures. EEG can be especially misleading when secondary bilateral synchronous discharges occur in patients with focal seizures. However, the semiology of focal seizures associated with mesial temporal lobe epilepsy has a characteristic and consistent semiology that is the mark of this common epilepsy syndrome in adulthood. We recently encountered a 53-year-old female with refractory seizures and a semiology strongly suggesting mesial temporal lobe epilepsy. Instead of focal seizures, prolonged absence seizures were validated by video-EEG monitoring and she became seizure-free after a change to broad-spectrum antiepileptic drugs. This case further expands our understanding of the complexity of semiology in electroclinical classification and the spectrum that may occur in adult absence seizures. It serves to underscore the need for ictal EEG recordings and the importance of concordance with the clinical course during the pre-surgical evaluation of patients with lesions and drug-resistant epilepsy. [*Published with video sequences*]

**Key words:** absence seizures, focal seizures, antiepileptic drugs, temporal lobe epilepsy, video-EEG monitoring



**Correspondence:**

William O. Tatum  
Department of Neurology,  
Mayo Clinic,  
4500 San Pablo Road,  
Jacksonville, FL 32224, USA  
<tatum.william@mayo.edu>

Historical reporting of witnessed seizures is the basis for diagnosis, classification, and treatment of epilepsy syndromes, but may be misleading (Baykan *et al.*, 2011). Routine clinical diagnosis relies upon “astute observations and expert opinions” obtained from the clinical history and course (Berg

*et al.*, 2010), though this is subject to error (Benbadis *et al.*, 2003). In clinical practice, focal seizures with dyscognitive features associated with staring in adulthood are commonly referred to by patients as “petit mal” seizures (to distinguish them from convulsions). Overall, a third of patients are

uncontrolled, despite antiepileptic drugs (AEDs) (Tatum, 2012), yet misdiagnosis is common (Baykan *et al.*, 2011). Video-EEG monitoring (VEM) with ictal recording provides definitive diagnosis and seizure classification (Baykan *et al.*, 2011, Benbadis *et al.*, 2003). We report an adult with drug-resistant seizures, a left temporal meningioma in the lateral wall of the cavernous sinus on MRI, and a semiology characteristic of mesial temporal lobe epilepsy (MTLE) identified during a presurgical evaluation, reflecting juvenile absence epilepsy (JAE).

## Case study

A 53-year-old female had an initial generalized tonic-clonic seizure at 17 years of age. Rare seizures, which she described as “petit mal”, were treated with primidone for 10 years, though she remained seizure-free for 23 years. Seizure onset (recurrence) at age 40 led to a diagnosis of refractory focal epilepsy. She reported monthly convulsions with tonic stiffening and subsequent clonic jerking as “grand mal” seizures, and described “small seizures” that occurred with a “glazed look”, unresponsiveness, and non-lateralizing automatisms of repetitive lip licking and fidgeting with her hands for 1-2 minutes. Shorter daily “phase outs” occurred in which she “froze” and was unresponsive for 5-10 seconds, which were sometimes accompanied by repetitive movements.

The semiology of her seizures was reported to be stable over the years, despite increasing frequency, and was characteristic of the events captured during VEM. A family history of epilepsy was absent. Her general and neurological examination was normal. Current AEDs include lacosamide at 200 mg twice daily, primidone at 250 mg twice daily, and carbamazepine at 200 mg daily after failing phenytoin, valproate (terminated due to side effects), ethosuximide, zonisamide, and oxcarbazepine. 3-T brain MRI identified an 8 mm x 4 mm meningioma in the lateral wall of the left cavernous sinus adjacent to the uncus, and a routine awake EEG was normal.

Previous EEGs from her adolescence were reported by the patient as “abnormal” with “spikes for 14-15 seconds”. Recent routine EEGs on two occasions were reported to be normal, though prolonged recording was not performed prior to VEM. Laboratory studies were unrevealing and a presumptive diagnosis of symptomatic drug-resistant TLE prompted VEM as part of a pre-surgical evaluation.

Several “small seizures” consisting of a stare, impaired responsiveness, motion arrest, and oral and bimanual-bipedal automatisms without focal or lateralizing signs occurred for 1-2 minutes (see *video sequence*) during a two-day VEM session. However, the ictal EEG

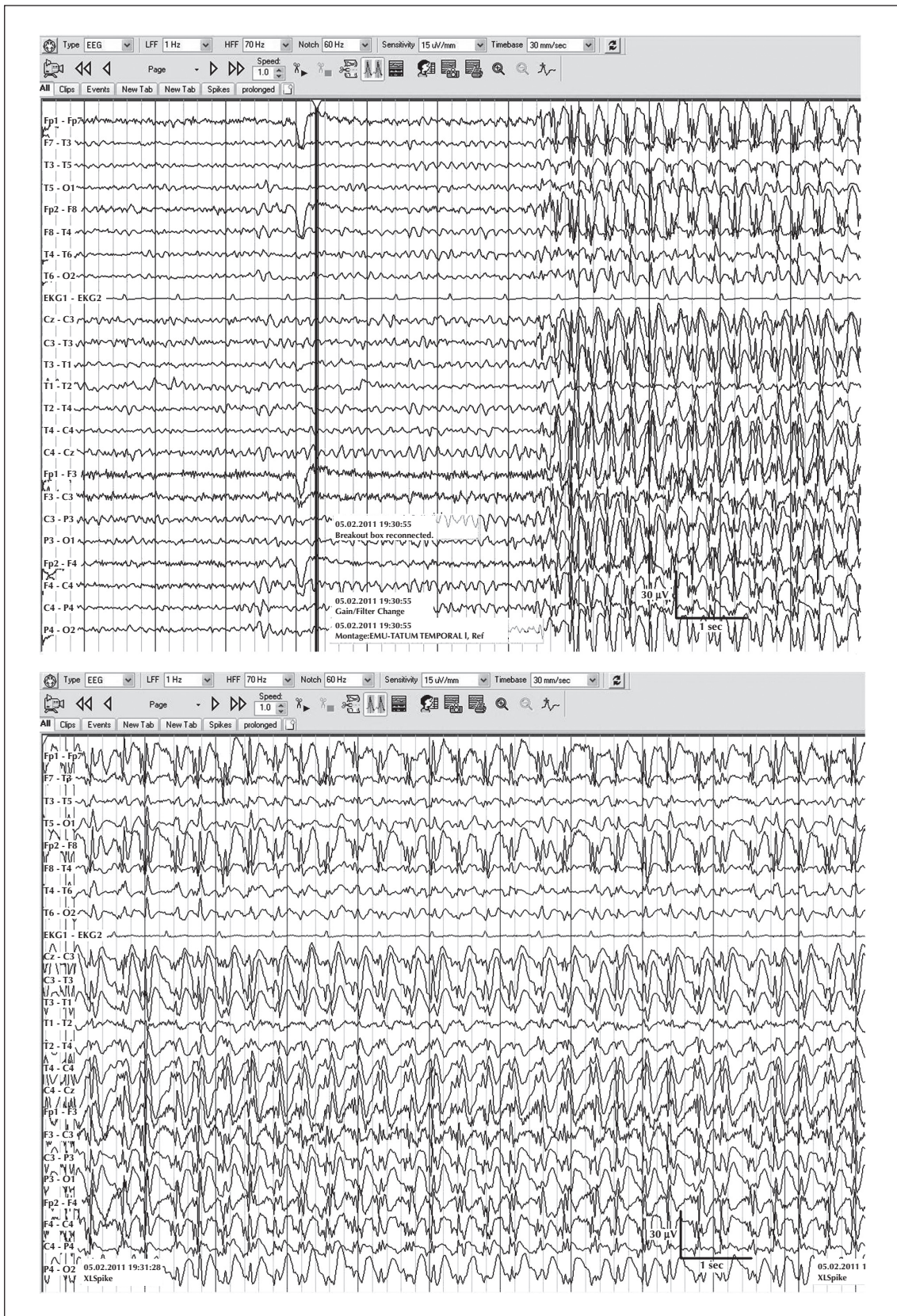
revealed a 3.5-Hz synchronous, symmetric, bifrontally-predominant generalized spike-and-wave (GSW) and polyspike-and-wave discharges. These were spontaneously recorded on six occasions, on average lasting 18 seconds, though with a range that included absence seizures of up to 2 minutes and 18 seconds (*figure 1*). Hyperventilation produced clear activation of bursts of “fast” GSW. Photic stimulation did not produce a photoparoxysmal response. Lamotrigine was begun and carbamazepine and lacosamide were subsequently discontinued. She has remained seizure-free for two years on lamotrigine at 150 mg twice daily.

## Discussion

Accurate seizure classification is required to correctly define an epilepsy syndrome, select appropriate AEDs, and anticipate long-term prognoses (Benbadis *et al.*, 2003; Baykan *et al.*, 2011). Our patient had absence seizures, yet her semiology and clinical diagnosis and course were characteristic of drug-resistant TLE. Even in today’s world of “smart” phone videos that can record outpatient seizures for later physician review, the presence of automatisms and repetitive movements in our patient would have suggested MTLE with the atypical semiology and course. The use of incorrect AEDs (*i.e.* carbamazepine and lacosamide) in her case may have produced seizure aggravation and created pseudo-drug resistance (Benbadis *et al.*, 2003). It is unclear if the incorrect use of AEDs induced a prolonged duration of absence seizures in our patient with JAE since the duration of the pre-treatment period could not be determined (using reports or tracings) prior to AED re-initiation in adulthood. The use of inappropriate AEDs in generalized epilepsy is not uncommon and in one study was present in 70% of patients (Benbadis *et al.*, 2003).

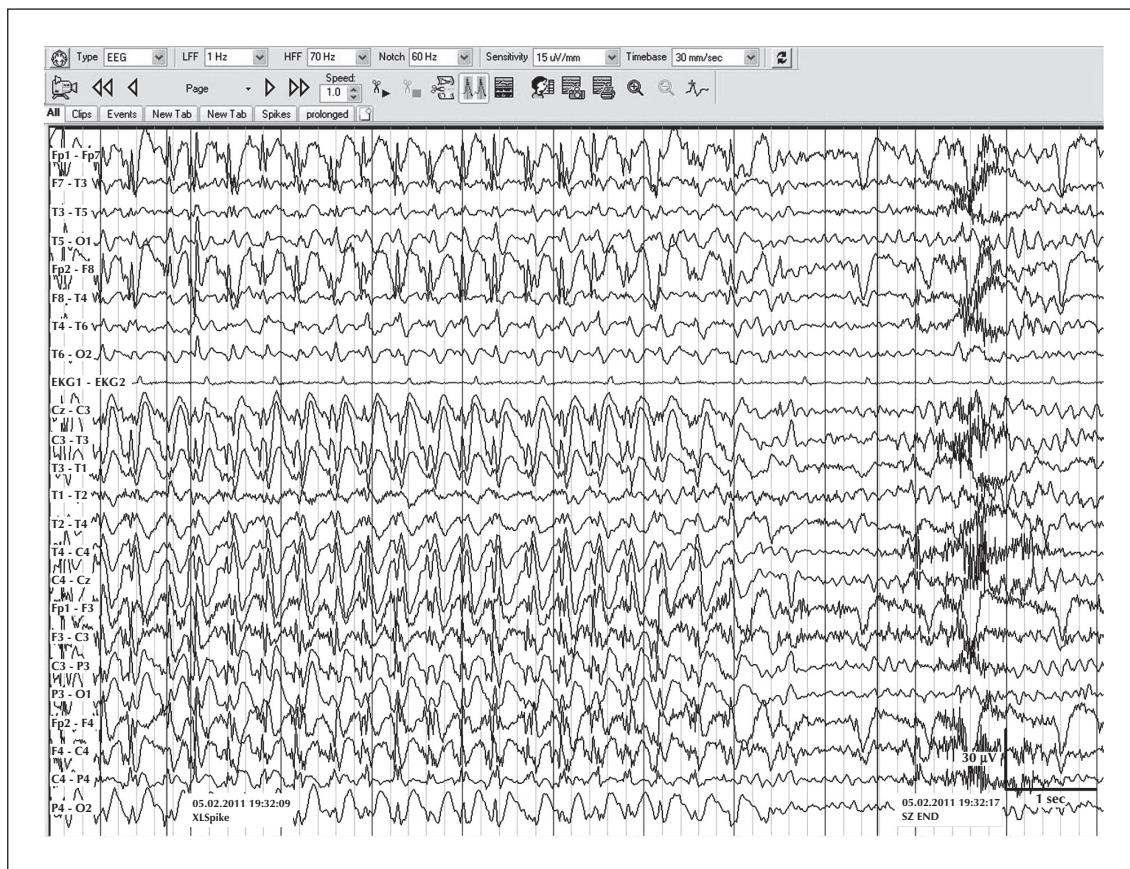
Electroclinical overlap between focal and generalized seizures occurs, though the typical differences are much greater than their similarities (Usui *et al.*, 2005). EEG may produce diagnostic pitfalls in genetic generalized epilepsy due to frequent focal and lateralizing features in up to 50% of patients, falsely suggesting focal seizures (Usui *et al.*, 2005). The prolonged duration and presence of automatisms in an adult are characteristic of TLE (Tatum, 2012). However, automatisms are also common in absences (Sadleir *et al.*, 2009). In a series of 405 absence seizures occurring in 70 children, 53 children experienced automatisms (76%) in 163 seizures (40%) and they were more likely to occur with prolonged seizures and hyperventilation (Sadleir *et al.*, 2009).

JAE is relatively common, with seizures beginning between 10 and 17 years of age in young people with normal intelligence, neurological function, and neuroimaging. While the distinction between focal



**Figure 1.** Ictal EEG of the seizure shown in the video.





**Figure 1.** (Continued)

EEG onset occurred at 19:30:9 and clinical onset at 19:30:59. The middle part of the seizure corresponds to 19:31:16. End of clinical manifestations at 19:32:21.

and generalized seizures in adulthood is basic, the importance of correct seizure classification reflects their differences with respect to diagnosis, treatment, and prognosis, despite the common link of staring (table 1).

MTLE is the most common adult epilepsy with drug-resistance, commonly leading to pre-surgical evaluation. The semiology, lesional brain MRI, lack of GSW on EEG, and clinical course in our patient suggests MTLE (Tatum, 2012), with true classification only revealed by VEM. A lesion on MRI suggests a symptomatic cause of epilepsy since seizures are often the presenting symptom of an intracranial meningioma (Lieu and Howng, 2000). The 3-Hz GSW (the hallmark of absence on EEG) was absent due to the limited yield encountered in wakefulness (Benbadis et al., 2003, Usui et al., 2005). Secondary bilateral synchronous GSW may occur on EEG with focal seizures due to the close proximity to the corpus callosum (Usui et al., 2005). Frontal lobe seizures are always a consideration in patients with generalized seizures (epilepsy). However, the initial age at onset, atypical hypomotor semiology, temporal lesion, and lack of lateralizing features on interictal and ictal EEG make this less likely.

**Table 1.** The expected differences between the semiology of a temporal lobe seizure and an absence seizure.

Semiology	Focal Seizure	Absence Seizure
Warning	Typical	None
Staring	Yes	Yes
Impaired responsiveness	Yes, but variable degree	+/-
Simple Automatisms	Yes	Maybe
Complex Automatisms	Yes	No*
Postictal confusion	Yes	No

\*The finding of complex automatisms are not expected in absence seizures, however, our patient exhibited them with bi-manual-bipedal automatisms simulating a temporal lobe seizure.

In addition, the rare absence seizure exhibiting focal electroclinical transformation also did not occur (Lieu and Howng, 2000). Instead, the ictal EEG in our patient

had bursts of typical prolonged 3.5-Hz GSW, characteristic of JAE in an adult (Sadleir *et al.*, 2009; Berg *et al.*, 2010). Our case underscores the potential for misclassification that is possible with adult absence seizures.

## Conclusion

The clinical signs of our patient, with a temporal lobe lesion, atypical generalized semiology, and an adult clinical course, mimicked those of drug-resistant MTLE, prompting a pre-surgical evaluation. Absence seizures mimicked focal seizures associated with pseudo-drug-resistant JAE caused by misclassification with a favourable outcome. This case emphasizes the importance of appropriate epileptic seizure classification, as well as non-epileptic seizure mimics in the pre-surgical evaluation of patients with epilepsy. It provides further support for a clinical continuum between absence seizures and MTLE which may both exhibit prolonged durations and hypomotor activity in adulthood (Lieu and Howng, 2000). This emphasizes the need for VEM in future classification systems to reflect inherent syndromic heterogeneity. □

## Acknowledgements and disclosures.

This study was not supported by any sponsorship or funding. None of the authors have any conflict of interest to disclose.

## References

- Baykan B, Altindag E, Feddersen D, Ozel S, Noachtar S. Does semiology tell us the origin of seizures consisting mainly of an alteration in consciousness? *Epilepsia* 2011;52: 1459-66.
- Benbadis SR, Tatum 4th. WO, Gieron M. Idiopathic generalized epilepsy and choice of antiepileptic drugs. *Neurology* 2003;61: 1793-5.
- Berg AT, Berkovic SF, Brodie MJ, Buchhalter J, Cross JH, van Emde Boas W, *et al.* Revised terminology and concepts for organization of seizures and epilepsies: report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia* 2010;51: 676-85.
- Lieu AS, Howng SL. Intracranial meningiomas and epilepsy: incidence, prognosis and influencing factors. *Epilepsy Res* 2000;38: 45-52.
- Sadleir LG, Scheffer IE, Smith S, Connolly MB, Farrell K. Automatisms in absence seizures in children with idiopathic generalized epilepsy. *Arch Neurol* 2009;66: 729-34.
- Tatum WO. Mesial temporal lobe epilepsy. *J Clin Neurol* 2012;29: 356-65.
- Usui N, Kotagal P, Matsumoto R, Kellinghaus C, Lüders HO. Focal semiologic and electroencephalographic features in patients with juvenile myoclonic epilepsy. *Epilepsia* 2005;46: 1668-76.

## Legend for video sequence

Video demonstrating a pseudo-temporal lobe seizure in an adult with juvenile absence epilepsy. Note the protracted unresponsiveness with complex non-lateralized automatisms composed of lip smacking, hand fidgeting, and picking at the telemetry cable, as well as automatic, rhythmic circular movements of the feet. The representative video clip begins 21 seconds after electrographic seizure onset. (19:51:16).

## Key words for video research on [www.epilepticdisorders.com](http://www.epilepticdisorders.com)

*Syndrome:* juvenile absence epilepsy (JAE)

*Etiology:* genetic predisposition

*Phenomenology:* staring; automatisms; lip smacking

*Localization:* unknown

## TEST YOURSELF



**(1) Which feature BEST separates temporal lobe seizures from juvenile absences?**

- A. Impaired responsiveness
- B. Staring episodes
- C. Automatisms
- D. Post-ictal state
- E. Abnormal brain MRI

**(2) Which antiepileptic drug would be the best choice for a female patient with unclassified seizures that manifest as a blank stare in addition to generalized tonic-clonic seizures?**

- A. Carbamazepine
- B. Primidone
- C. Phenytoin
- D. Lacosamide
- E. Lamotrigine

*Note: Reading the manuscript provides an answer to all questions. You can check for the correct answer by visiting the Educational Centre section of [www.epilepticdisorders.com](http://www.epilepticdisorders.com)*