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Unmasking the entity of 'drug-resistant' perioral myoclonia with absences: the twitches, darts and domes!

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ABSTRACT

Objective. Perioral myoclonia with absences (POMA) is not recognized as a unique electro-clinical syndrome and studies suggest its inclusion under the genetic generalized epilepsy (GGE) spectrum. The aim of this study was to explore the prevalence and electro-clinical homogeneity of this disorder in an epilepsy monitoring unit.

Methods. Between 2013 and 2019, among drug-resistant epilepsy patients who were referred for video-telemetry, those diagnosed with POMA based on the presence of documented absences with prominently observed peri-oral muscular contractions accompanied by generalized EEG features were included.

Results. Among 62 patients who were diagnosed with absence epilepsy, five finally met the criteria for POMA (8.1%) with late childhood or adolescent onset of epilepsy. Four (80%) had a referral diagnosis of focal epilepsy based on historical focal features with exacerbation of seizures on oxcarbazepine. All five patients demonstrated brief absences with orbicularis oris muscle contractions accompanied by subtle focal phenomenology. One patient had concurrent axial-appendicular myoclonic jerks precipitated by hyperventilation. While four patients had strikingly identical interictal and ictal characteristics of typical absence epilepsy, one patient demonstrated additional atypical generalized polyspike discharges without a "dart-dome" morphology. Therapeutic response to introduction of sodium valproate was noted in all five patients. Features that were not consistent with the diagnosis were apparent in one patient who was re-classified with combined focal and generalized epilepsy. Differentiating aspects in this patient included multifocal discharges, background slowing, fast-recruiting ictal rhythms and valproate resistance.

Significance. This is one of the largest case series of POMA. This entity, which falls under the spectrum of GGE, remains under-diagnosed and its distinctive electro-clinical features need to be recorded in order to prevent misclassification as focal epilepsy of probable opercular origin. Background-slowing, atypical ictal rhythms and valproate unresponsiveness are not consistent with the diagnosis of this unique absence epilepsy. [*Published with video sequences*].

VIDEOS ONLINE

Key words: perioral myoclonia with absences; genetic generalized epilepsy; video-EEG; diagnosis

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Peri-oral myoclonia with absences (POMA) is an epilepsy syndrome characterized by absence seizures with specific features. Following its initial description in 1994, the entity is presently not recognized within the International League Against Epilepsy (ILAE) classification schema [1-3]. The presence of short absences makes POMA similar to other genetic generalized epilepsy (GGE) with absences such

as juvenile absence epilepsy (JAE), but the distinctive feature is the associated pronounced rhythmic contraction of the perioral muscles, mainly involving the orbicularis oris. This is reported to be associated with generalized tonic–clonic seizures (GTCS) which may be noted independently or may immediately follow the absence spells. Absence status epilepticus, with unsatisfactory response to antiseizure medications (ASM), and persistence of seizures into adulthood are other notable characteristics. Despite its association with generalized discharges which have a GGE-like morphology, the presence of clinical features, which point towards focal-onset motor seizures with impaired awareness or EEG asymmetry, frequently lead to misdiagnosis as focal epilepsy [4]. The actual prevalence of this entity in epilepsy care centres is sparsely reported in the literature and few case reports with POMA exist; more studies with video-telemetry recordings with documentation of distinctive facial myoclonia are necessary in order to recognize this syndrome [5, 6]. Problems and pitfalls in the diagnosis of this syndrome are also rarely discussed. This study aimed to determine the prevalence of this subtype among patients with other refractory absence epilepsy syndromes who underwent video telemetry for syndromic diagnosis and management in view of drug-resistant epilepsy.

Methods

This was a descriptive study based on results from a prospectively maintained database from a comprehensive epilepsy care centre situated in the south Indian city of Thiruvanathapuram. The video telemetry and discharge summaries were screened for an exit diagnosis of POMA. The demographic data included age of patients, sex, antecedents or initial precipitating injuries, developmental milestones, age at onset of seizures, predominant seizure type based on historical interpretation, associated seizure subtypes, frequency of seizures, duration of epilepsy, and family history. The video-electroencephalography (VEEG) data included recordings of seizures; ictal onset, ictal patterns, duration of events recorded, phenomenological aspects of the seizures with the corresponding ictal rhythm, and interictal epileptiform discharges (IED). VEEG was performed using 32-channel EEG systems with in-built cameras, with a standard arrangement of the 10–20 international electrode placement. Indications for overnight VEEG recording included apparent epilepsy pharmaco-resistance or for exclusion of non-epileptic events. The electroclinical data were interpreted by two certified epileptologists (RNM & AR). The MRI scans were evaluated for presence of any intracranial pathology in order to rule out symptomatic causes. Medications of the patients were reviewed, and details of the ASM used and the resulting changes in seizure types or seizure frequency were recorded.

Absences were considered to be typical if the ictal rhythms showed the classic 3-4-Hz spike-wave (SW) discharges of abrupt onset, commencing without any consistent pre-ictal focal EEG disturbances. Although the frequency may be 3-4 Hz at onset, it may slow down to 2.5 to 3.0 Hz towards the end of the discharge. The characteristic electrical field should be apparent in the form of maximum negativity at F_7/F_8 or F_3/F_4 and brief (<1s), otherwise apparent shifting asymmetry at onset was not considered exclusionary. Morphology was considered to be "dart and dome" if a single spike was followed by a single wave, however, during sleep or in older patients, a double spike may be apparent. Towards the end of the discharge, offset may not be as abrupt as onset, and may be fragmented and gradual. The discharges may be provoked by hyperventilation. The study was conducted with the approval of the Institution Ethics Committee.

Results

Demographics

In the study period between 2013 and 2019, 62 patients had a diagnosis of drug-resistant primary absence epilepsy after video telemetry. These included six (9.7%) patients with refractory childhood absence epilepsy, 24 (38.7%) with JAE which also included four with neck myoclonia, seven (11.3%) with eyelid myoclonia with absences, 19 (30.7%) with a diagnosis of epilepsy with myoclonic absences (EMA), and six (9.7%) with a diagnosis of POMA. Age at onset of epilepsy ranged from 5.5 to 12 years and age at presentation to the telemetry unit ranged between 10 and 15 years. No gender predisposition was apparent, with three females and two males in the series. Two patients had a positive family history of febrile seizures; one patient who had atypical interictal and ictal EEG features in the form of generalized polyspikes, also suffered with complex febrile seizures which were associated with similar facial myoclonia and no generalization. Details of five patients with POMA (8.1% of the cohort; Patients 1-5) and one patient initially suspected to have POMA based on electroclinical criteria, but re-diagnosed with combined focal and generalized epilepsy (Patient 6), are listed in *tables 1, 2*.

▼ **Table 1.** Clinical features of patients along with treatment and seizure outcomes.

Patient	Gender/age at onset/age at presentation; relevant history	Seizure subtypes based on history in chronological order	Initial referral diagnosis/final diagnosis after VEEG	Initial AED	Final AED and seizure outcome
1	F/11yrs/12yrs	a) GTCS with head adversion b) Focal motor unaware (2/day-10-15 seconds)- right facial twitch c) Secondary GTCS	Secondary generalized epilepsy + MRI negative/ POMA	OXC+LEV® +PER®+ VAL	Cessation of OXC® VAL+LEV ® seizure-free
2	M/11 yrs/17yrs	a) Focal motor unaware (4-5/day)- right facial twitch b) Associated right-sided asymmetric tonic upper limb posturing	Focal epilepsy-insular + MRI negative/ POMA	OXC+LEV+ CLB	Cessation of OXC® VAL+LEV® seizure-free
3	F/12yrs/15yrs; complex febrile seizures; family history of typical febrile seizures	Absences (3-4/month) Peri-oral twitch Focal non-motor with eye deviation Nocturnal GTCS (3-4/month)	JAE® Focal epilepsy (operculo-insular) + MRI negative/ JME with POMA	LEV® OXC® LEV + CLB	VAL+LEV® seizure-free
4	F/5.5yrs/10yrs-learning disability; family history of typical febrile seizures	GTCS Facial and proximal upper limb myoclonia Absences Myoclonic-absences (10-14/day)	GGE® Epilepsy with myoclonic absences/ POMA	VAL® remission® VAL stopped due to weight gain® LTG+LEV+CLB	VAL+LEV® seizure-free
5	M/11yrs/16 yrs	Focal motor unaware-facial twitch to either side (2-3/day) GTCS; 3-4/month	Focal epilepsy + MRI negative/ POMA	OXC® OXC+CLB® OXC+CLB+ LCSM	OXC+LCSM cessation® VAL+LEV® LTG+VAL® seizure-free
6	M/10 yrs/21 yrs; complex febrile seizures; scholastic decline; karyotyping normal	Focal motor aware (facial twitch to either side) Absences GTCS with nocturnal preponderance Myoclonic jerks Secondary generalized seizures; 1-5/month	Focal epilepsy + negative MRI, PET ® atypical GGE with POMA/ resistant, combined focal and generalized epilepsy of unknown aetiology	Withdrawal seizures on CBZ taper; CBZ+CLB+ VAL® CBZ+VAL+ LTG	TOP+VAL+CLB® drug-resistant epilepsy

M: male; F: female; yrs: years; GTCS: generalized tonic-clonic seizures; GGE: genetic generalized epilepsy; JAE: juvenile absence epilepsy; JME: juvenile myoclonic epilepsy; POMA: perioral myoclonia with absences; OXC: oxcarbazepine; VAL: sodium valproate; LEV: levetiracetam; CLB: clobazam; LTG: lamotrigine; TOP: topiramate; CBZ: carbamazepine; LCSM: lacosamide; PER: perampanel.

▼ **Table 2.** Electrophysiological findings in patients.

Patient	Seizure semiology recorded (video files available for patients 1-4 & patient 6)	Background activity	Interictal epileptiform discharges– topography & morphology	Ictal rhythm-onset and evolution
1 (VEEG on OXC) *(figure 1, video 1)*	Behavior arrest-perioral myoclonia-right facial twitches-eye deviation to left	8.5-9 Hz; no focal slow activity	Bifrontal dominant GSW; "dart and dome" PPR at 24 Hz	Typical abrupt 3-Hz GSW; evolves as 2-2.5 Hz; GPSW; left frontal rhythmic theta; bifrontal slow offset
2 (VEEG on OXC) *(figure 2, video 2)*	Stare-facial myoclonia- bilateral upper limb myoclonia (left> right)	9-9.5 Hz; no focal slow activity	Bifrontal dominant GSW; "dart and dome"	Typical bifrontal leading GSW 4-Hz (asymmetric right); abrupt onset & offset
3 (VEEG on LEV) *(figure 3, video 3)*	Stare-facial myoclonia	8.5-9 Hz; no focal slow activity	Sleep activation of bifrontal dominant GPSW	Atypical GPSW onset at 4.5-5 Hz; abrupt onset-offset
4 (VEEG on LEV) *(figure 4, video 4)*	During HV; behavior arrest-facial myoclonia-neck & bilateral upper limb myoclonia	9-9.5 Hz; no focal slow activity	Rt frontal; bifrontal dominant GSW; GPSW; "dart & dome"	Typical abrupt bifrontal (right fontal asymmetry) onset and evolution at 3.5-4 Hz; fragmented right frontal offset
5 (VEEG on OXC)	Behavior arrest- facial myoclonia- head deviation to right side	8.5-9.5 Hz; no focal slow activity	Independent frontal; bifrontal SW; GSW; "dart and dome"	Typical bifrontal onset GSW at 3.5-4 Hz; abrupt onset and offset
6 (VEEG on CBZ + VALP) (Ictal onset: *figure 5*, video 5A) (ictal evolution & termination: video 5B)	Behavior arrest-right facial jerks-facial myoclonia-termination with initial head jerks to the right side & facial myoclonia	7.5-8 Hz; generalized slow activity (bilateral parieto-occipital maximum)	Multifocal & GSW	GPSW burst® bifrontal, generalized metamorphic 9-10-12-Hz recruiting rhythm; slow offset as seen in video 5B with generalized sharp/ slow SW (1 Hz)

VEEG: video electroencephalography; GSW: generalized spike-wave; GPSW: generalized polyspike-wave; HV: hyperventilation; Hz: hertz; PPR: photoparoxysmal response.

Clinical features *(table 1)*

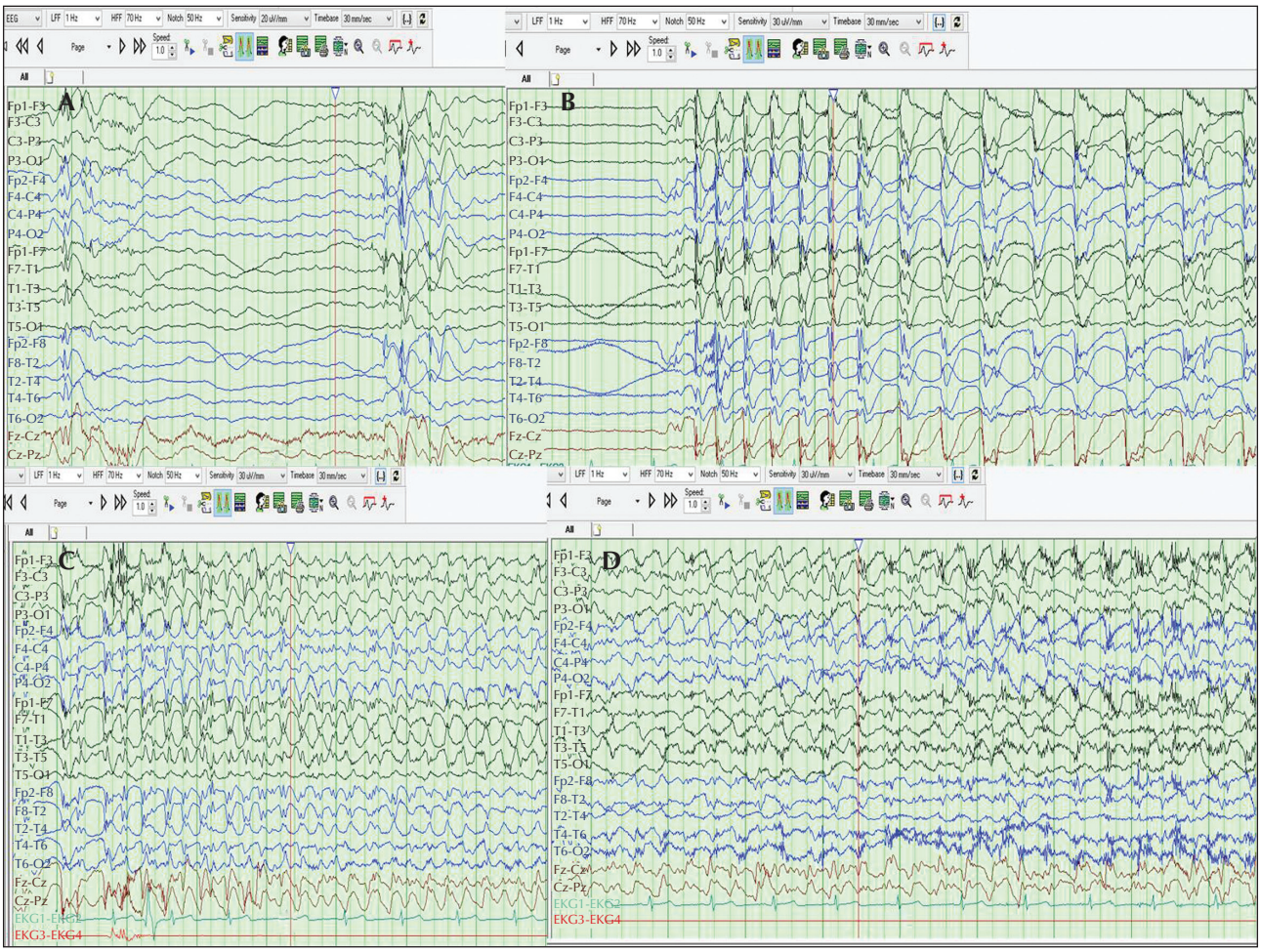
Out of five patients, four had a referral diagnosis of focal epilepsy based on the clinical interpretation of seizure subtypes. Exposure to ASM not suited for absence seizures was also noted with four patients on oxcarbazepine, each one on lacosamide and perampanel at the time of referral, in combination with sodium valproate, clobazam or levetiracetam. These patients showed an increase in the frequency of absence seizures and facial jerks. One patient was referred due to uncertainty regarding juvenile myoclonic epilepsy versus epilepsy with myoclonic absences. Phenomenologically, based on video EEG records (*video sequences 1-4*), seizures were consistent with brief absences with orbicularis oris myoclonia along with focal phenomenological features noted in three patients in the form of asymmetric facial

twitches, upper limb jerks and eye deviation. Two patients demonstrated symmetric perioral and upper limb myoclonias. In one patient, hyperventilation precipitated facial myoclonia with absences which evolved into generalized myoclonic jerks.

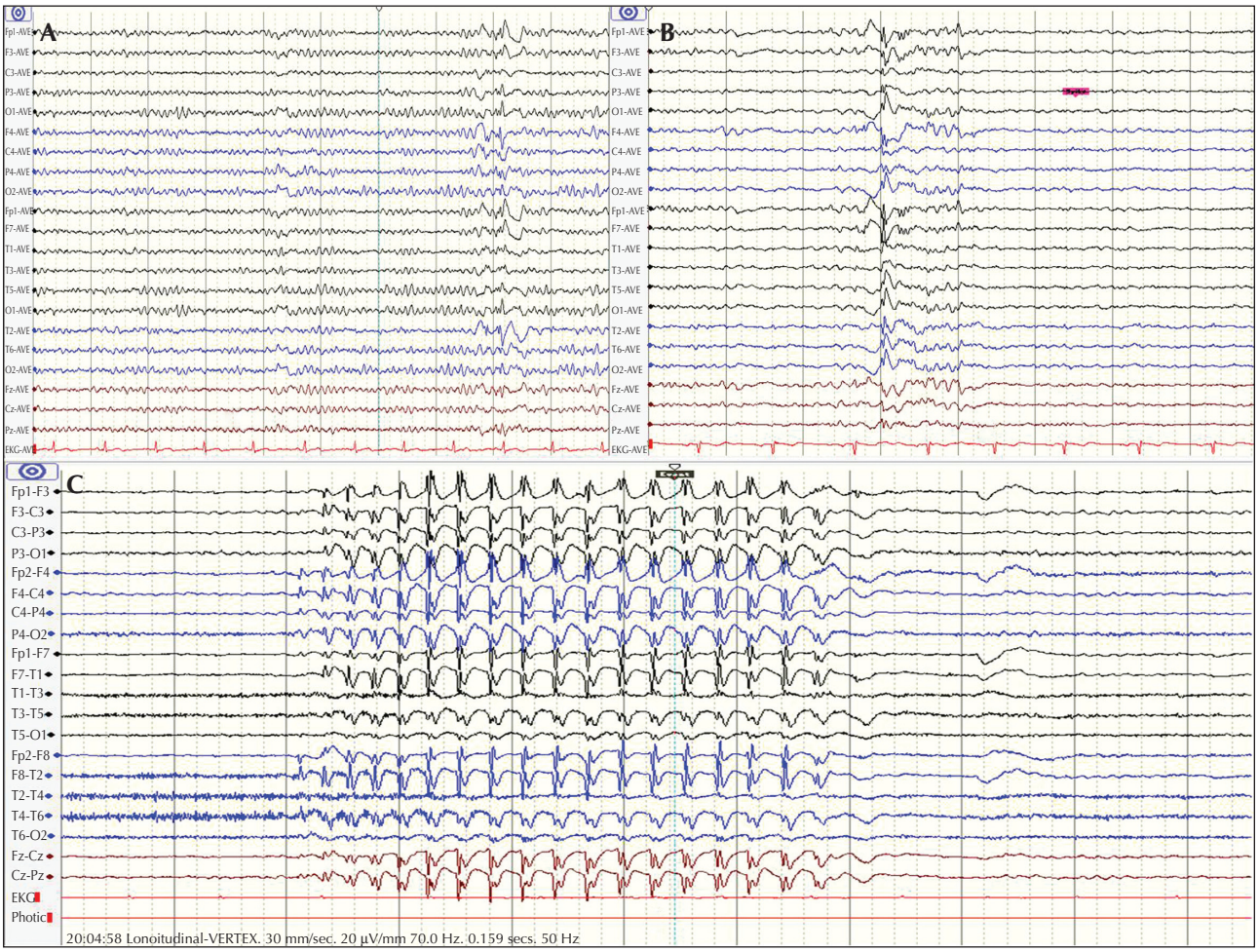
EEG features *(table 2)*

Interictal EEG features favoured GGE in all five patients with typical generalized 3.5-4-Hz SW discharges with a "dart-dome" morphology noted in four patients during the ictal rhythm, although borderline asymmetry was noted in two. A focal pattern of ictal evolution was noted in Patient 1. Patient 3 had atypical generalized polyspikes based on the interictal and ictal records, favouring the possibility of juvenile myoclonic epilepsy, however, the recorded phenomenology was suggestive of POMA (*figures 1-4 and video sequences 1-4*). All five patients had fairly abrupt ictal onsets. None of the patients exhibited slowing of the awake background posterior dominant rhythm, clear focal ictal onset, or post-ictal focal slowing.

Patient 6, detailed in this study to demonstrate the problems in diagnosing POMA, had a distinctive course with a clinical history showing a later appearance of myoclonic jerks, interictal and ictal EEG abnormalities that favoured a combined focal and generalized epilepsy. The ictal semiology, consisting of focal features and demonstrable asymmetric facial myoclonia, along with a bifrontal and generalized 10-12-Hz ictal rhythm without a 'dart-dome'



■ **Figure 1.** (A) Interictal EEG of Patient 1 showing a burst of frontal dominant generalized spike-and-wave discharges. (B) Ictal onset commencing abruptly as a 3-Hz generalized spike wave (with typical dart-dome appearance) which slows down to 2 Hz. (C) Asymmetric evolution noted over the left hemisphere coinciding with asymmetric right facial jerks in video sequence 1. (D) Gradual offset over bilateral frontal regions.

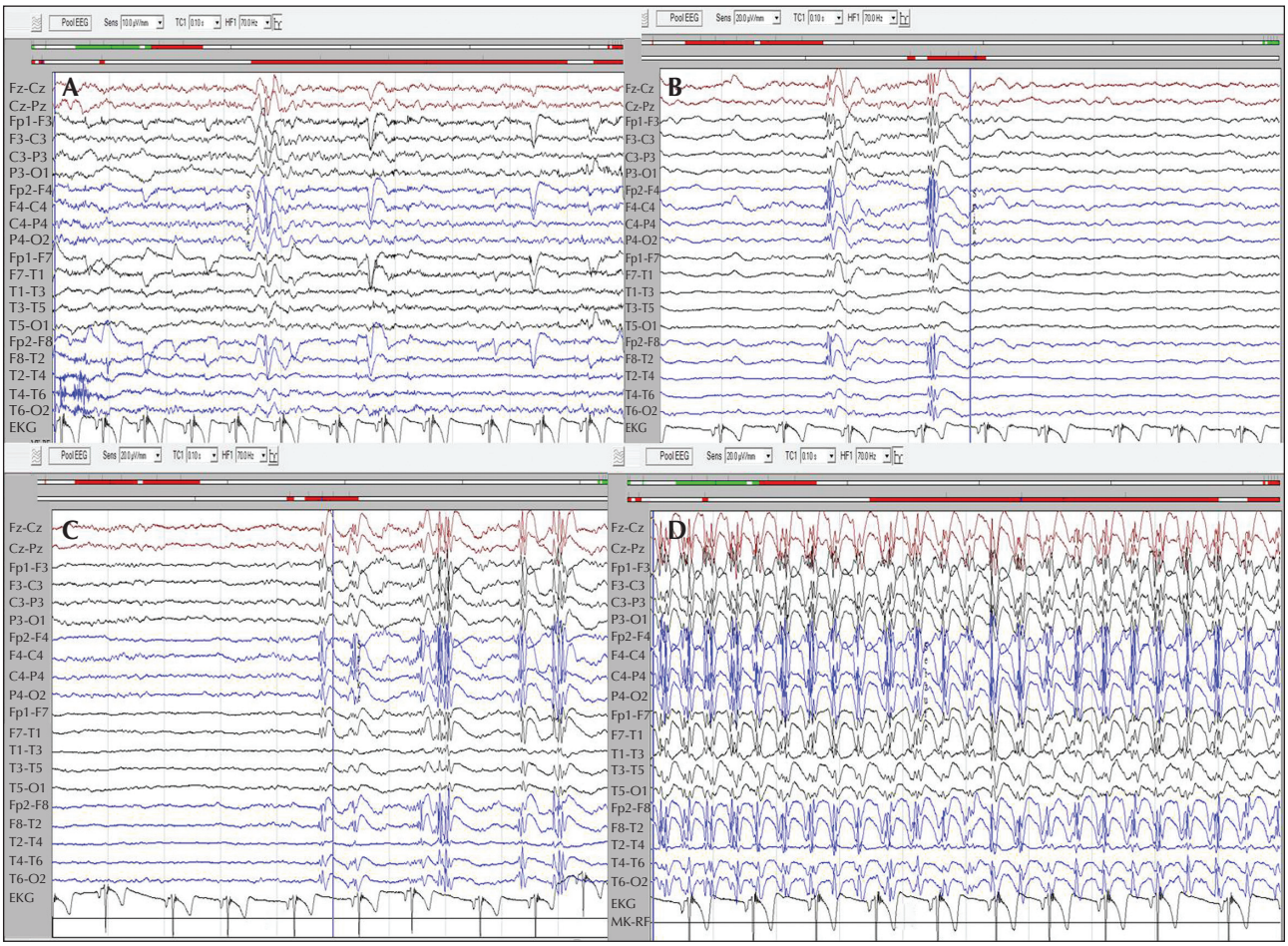


■ **Figure 2.** (A-B) Interictal EEG in Patient 2 during wakefulness and sleep showing bursts of frontal dominant generalized spike-waves. (C) Ictal EEG during perioral myoclonia with absence, as noted in video sequence 2, associated with a typical burst of asymmetric generalized 3-Hz spike-wave and dart-dome morphology with abrupt onset and offset.

pattern, (figure 5, *video sequence 5A, B*), as has previously been reported in patients with POMA [6]. With an initial diagnosis of resistant GGE and POMA, the patient failed to respond to trials of valproate and withdrawal of carbamazepine. His karyotyping, which was performed to exclude the possibility of ring 20 chromosome-related epilepsy (due to the refractory phenotype and an electro-clinical course favouring frontal lobe epilepsy with atypical absences), was normal. He continued to remain drug-resistant at the last available follow-up visit along with scholastic decline. He underwent pre-surgical evaluation with serial VEEG, high resolution 3T MRI, and positron emission tomography (PET), which failed to generate a clear hypothesis for focal epilepsy.

Discussion

This is the largest case series to date on epilepsy with POMA, among a cohort of patients diagnosed with distinctive absence epilepsy syndromes after referral for video telemetry, as a consequence of drug-resistant epilepsy. It was apparent that drug resistance in this syndrome was a 'pseudo-resistance'; a result of inappropriate medication due to incorrect diagnosis (oxcarbazepine), as noted in the form of seizure freedom in five patients with the introduction of optimal dose of valproate in combination with either levetiracetam or lamotrigine. Absence of response to valproate, multifocal interictal epileptiform abnormalities, background slowing



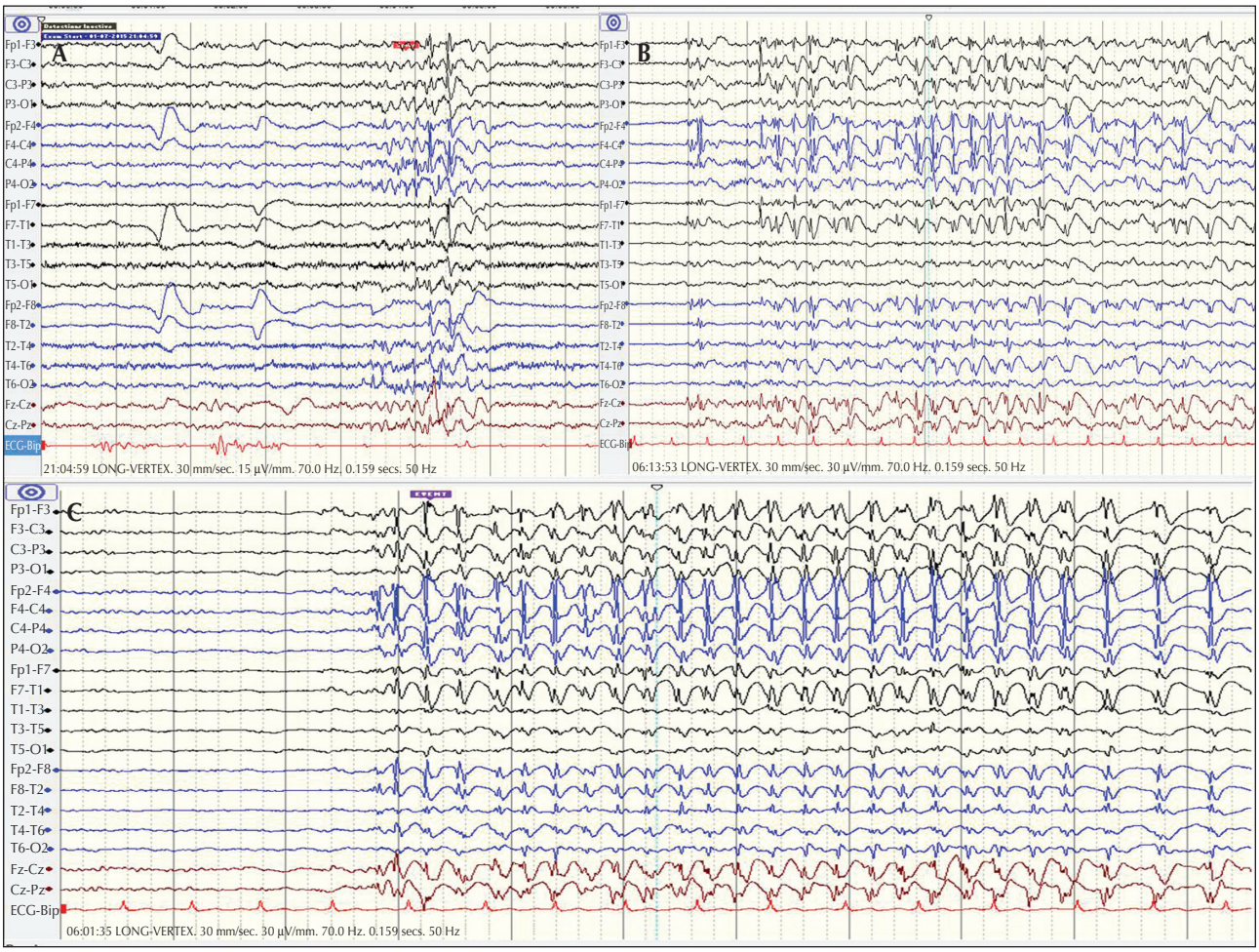
■ **Figure 3.** (A) EEG of Patient 3 during wakefulness showing bifrontal dominant bursts of generalized spike-waves with normal background activity. (B-C) Sleep showing activation of generalized discharges of polyspike (more than three spikes) -wave morphology. (D) Ictal rhythm during perioral myoclonia, as shown in video sequence 3, and runs of rhythmic atypical generalized spike-polyspike-wave dicharges at >4-4.5 Hz.

and absence of typical "dart-dome" 3-4-Hz ictal rhythm, as demonstrated in Patient 6, should direct the clinician away from the diagnosis of POMA.

The electro-clinical data of our patients are compatible with criteria defining the syndrome of POMA, similar to published cases (*see supplementary table*):

- peri-oral myoclonia is the main seizure type along with GTCS, although other areas of myoclonus, such as the neck and upper limbs (as noted in Patient 4, post hyperventilation) have been described [7];
- epilepsy onset occurs during late childhood or adolescence;
- psychomotor development is usually normal;
- seizures are often difficult to control.

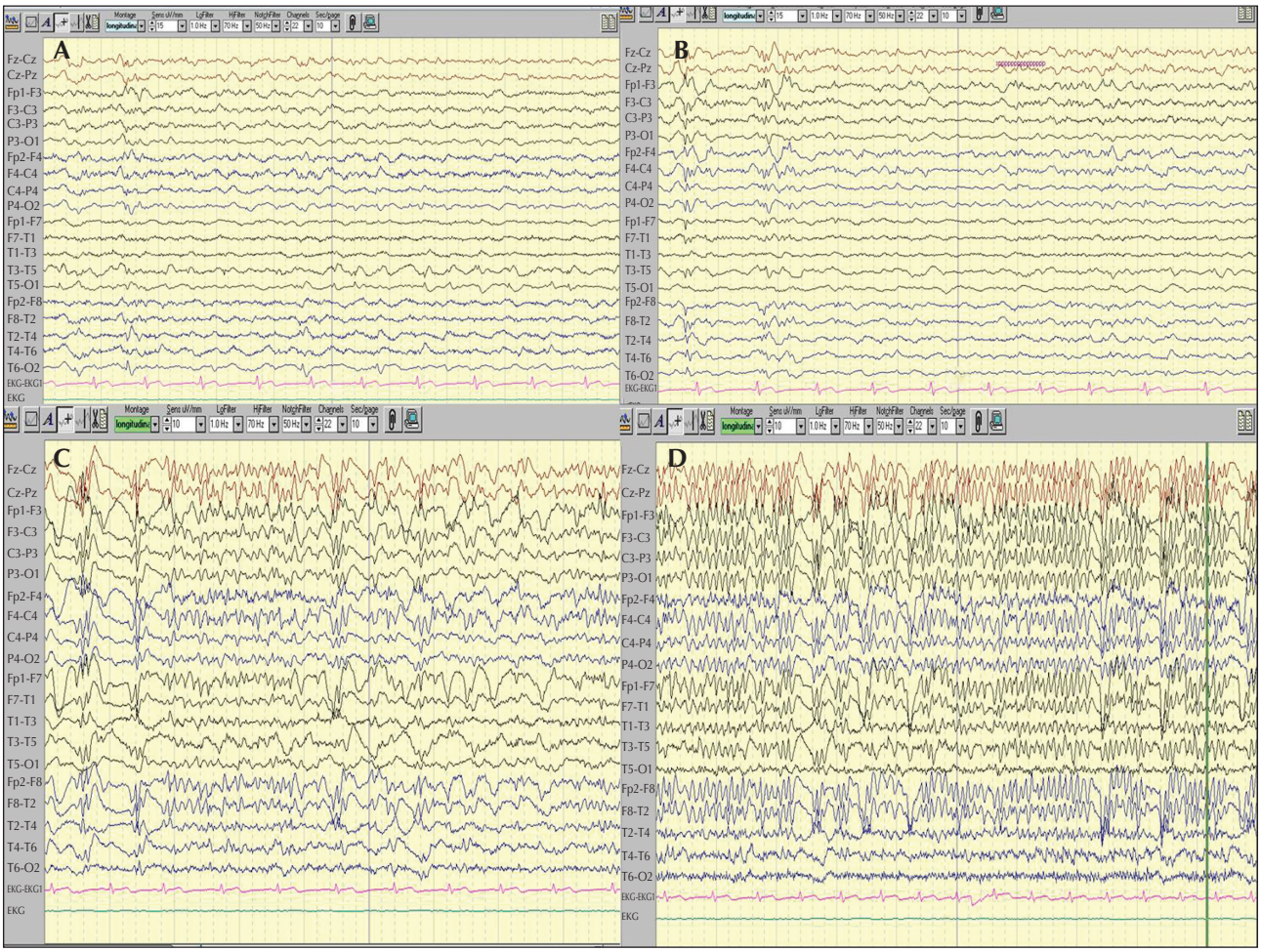
The latter feature is often secondary to introduction or increasing dose of inappropriate ASM and can potentially lead to myoclonic or absence status epilepticus in GGE syndromes, similar to previous reports of eyelid myoclonia with absences [8, 9]. In specific case reports of POMA, seizure exacerbation on carbamazepine and development of myoclonic status by phenytoin were documented [10, 11]. Similarly, in our series, the use of oxcarbazepine, which is a narrow-spectrum ASM, at the time of referral, led to aggravation of not only facial myoclonia (which were interpreted as focal motor seizures involving one side of the face) but also generalized tonic-clonic seizures (GTCS). It is crucial to note that this aggravation of GGE syndromes with various narrow-spectrum ASM often results from misdiagnosis as focal epilepsy [12]. This can be minimized by accurate clinical diagnosis supported by video-EEG. Unless the correct diagnosis



■ **Figure 4.** (A) Interictal EEG of Patient 4 during wakefulness showing normal background activity and bursts of frontal dominant generalized spike-wave discharges. (B) Ictal EEG during peri-oral myoclonia showing asymmetric (fight frontal dominant) bursts of generalized typical 3.5-4-Hz spike-and-wave discharges. (C) Ictal EEG during hyperventilation, as noted in video sequence 4, showing abrupt onset of generalized typical 3.5-4-Hz spike-wave discharges with right frontal asymmetry, slowing down to 2 Hz towards offset with typical dart-dome appearance.

of GGE syndromes such as POMA is sought and treatment is initiated with ASM that have proven efficacy in GGE (such as valproate, lamotrigine, topiramate and levetiracetam); along with prudent avoidance of narrow-spectrum medications (such as carbamazepine, oxcarbazepine, gabapentin, tiagabine, vigabatrin and phenytoin), the issue of seizure aggravation is likely to persist.

The mechanism of seizure aggravation by sodium channel blocking agents such as carbamazepine/oxcarbazepine is largely hypothesized based on animal models of genetic absence epilepsy and this effect is proposed to be due to GABAergic action on the ventrobasal thalamus rather than sodium channel blocking properties [13]. However, recent evidence which can be extrapolated from experiments in mouse models of Dravet syndrome, a complex epilepsy syndrome of childhood with loss of function of the *SCN1A* gene, suggests that reduced sodium currents in GABAergic inhibitory interneurons in *SCN1A*+/– heterozygotes may cause hyperexcitability. This can lead not only to epilepsy in patients with this disorder but also to seizure exacerbation on conventional sodium channel blocking ASM [14]. It is probable that such a mechanism is responsible in GGE, given its now evident complex genetic basis [15]. An overzealous conclusion regarding seizure aggravation may be detrimental, as noted in Patient 6 in whom exacerbation of seizures was reported on carbamazepine withdrawal. However, as discussed above, syndromic diagnosis



■ **Figure 5.** (A) Interictal EEG of Patient 6 during wakefulness showing generalized slowing of background activity, interrupted by multifocal spikes which have a temporo-occipital predominance. (B) Sleep recording showing activation of generalized atypical polyspikes. (C) Ictal onset (corresponding to video sequence 5) showing a burst of generalized polyspikes, following by 10-12-Hz metamorphic bifrontal dominant generalized sharp waves which evolve as generalized monomorphic rhythmic 10-12-Hz sharp waves interrupted by spike-wave discharges (D).

is paramount and the patient evidently met the criteria for combined focal and generalized epilepsy based on EEG findings. This is an entity which has an unknown basis and is increasingly being recognised by the ILAE. While classically exemplified by Dravet syndrome and Lennox Gastaut syndrome, this entity essentially includes a spectrum of complex epilepsies wherein the interictal EEG may show both generalized SW and focal epileptiform discharges [16].

Although ictal rhythms and interictal activity, as mentioned above, are classic signs of GGE, it is notable that many absence syndromes with distinctive features, which are potentially drug-resistant, such as EMA, may also manifest similarly. However, the clinical profile is more heterogeneous than that of POMA [17]. While the 3-4-Hz generalized SW rhythm is diagnostic and usually bilaterally symmetrical, varying degrees of asymmetry and occasional brief unilateral discharges can be present. On EEG, the asymmetries typically change from side to side, and only by evaluating the whole recording can the generalized basis become certain. Under the influence of narrow-spectrum AEDs, such as oxcarbazepine, focal features may appear more striking, as noted in the ictal evolution in Patient 1. Such asymmetries should not lead to an erroneous diagnosis of focal epilepsy. Polyspikes during sleep, as noted in Patient 3, appear to be associated with a less favourable prognosis with regard to long-term epilepsy remission and can be seen in JAE [18].

Based on the atypical features noted in Patient 6, it is pertinent to note that the peri-oral motor manifestations (*video sequence 5A, B*), similar to opercular-onset

focal epilepsy, have been associated with a metamorphic generalized 10-12-Hz sharp-wave rhythm without a consistent focal onset, as described previously in a case report [6]. The latter was associated with a more sustained tonic nature of facial contraction or 'chapeau de gendarme' which was not apparent in our patient. The atypical ictal rhythm without the 'dart-dome' morphology, multifocal (predominantly over parieto-occipital regions) and generalized IED, background slowing, and evolution as drug-resistant focal-onset bilateral tonic-clonic seizures excluded the possibility of POMA in Patient 6. The asymmetric perioral myoclonia were strikingly similar to the evolution noted in Patient 1 (*video sequence 1*). Patients 1-5 demonstrated the typical 3-4-Hz generalized SW or generalized polyspike rhythms with the "dart-dome" appearance, despite shifting asymmetries. In the absence of the typical ictal rhythm of POMA, this distinction may not be straightforward. A phenomenological aspect which would help to distinguish focal opercular seizures from POMA is the presence of well-defined auras such as visceral sensations and non-specific somatosensory phenomena which could indicate an insulo-opercular origin [19]. This was crucially absent in Patient 6. Focal spikes, as seen in Patient 4, are also evident in typical absence seizures and represent abortive forms of generalized SW discharges. Similarly, many 3-Hz generalized SW discharges demonstrate a subtle lateralized onset, a few milliseconds before the typical generalized rhythm, as illustrated in *figure 2C, 4C*. Our series suggests that these findings do not necessarily indicate a cortical pathology that secondarily leads to generalized discharges. Although one is inclined to complete the evaluation with MRI, the typical morphology of the ictal rhythm and IED should narrow down the diagnosis to POMA. Debate will continue to rage with regard to such classifications, as reports also exist of combinations of surgically remediable syndromes with GGE overlap as well as cases with focal epilepsy of temporal origin which manifest with generalized 3-Hz SW ictal rhythms of typical morphology that have been conclusively shown to be secondary generalized or may point to a coexistent GGE syndrome [20, 21].

Limitations of our descriptive study include a referral bias given the fact that evaluation of patients was undertaken at a stage when they were ASM-resistant and without initial EEG tracings available for comparison. This could account for the high percentage of patients diagnosed with POMA among those with refractory absences and absences misdiagnosed/treated as focal-onset seizures. These patients are more likely to be evaluated by us using video-EEG, which conclusively helps to establish the final electroclinical syndrome. However, this may also highlight the inherent complexity of absences in this group of patients, similar to EMA. Additionally, resting state and spike-triggered EEG-functional MRI studies would throw more light on the pathophysiology of this novel subtype of absence epilepsy.

Conclusion

POMA represents a GGE syndrome with distinctive features that require video telemetry for diagnosis. Unless the diagnosis is suspected and actively confirmed, the patient is likely to continue to be misdiagnosed, as noted in our experience, given the historical interpretations of focal features of peri-oral myoclonia akin to operculo-insular epilepsy. The syndrome is amenable to treatment once seizure aggravating AEDs are withdrawn. Electro-clinical homogeneity is fairly apparent and the presence of consistently focal/multifocal EEG features with background abnormalities, in addition to atypical ictal rhythms, should initiate investigation for a focal epilepsy. ■

Supplementary material.
Summary didactic slides and supplementary table are available on the www.epilepticdisorders.com website.

Disclosures.
None of the authors have any conflict of interest to declare.

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Legends for video sequences

Video sequence 1

POMA in Patient 1 (see *table 2* for a detailed description).

Video sequence 2

POMA in Patient 2 (see *table 2* for a detailed description).

Video sequence 3

POMA in Patient 3 (see *table 2* for a detailed description).

Video sequence 4

POMA in Patient 4 (see *table 2* for a detailed description).

Video sequence 5

Focal dyscognitive seizures in Patient 6: (A) ictal onset; (B) ictal evolution and termination (see *table 2* for a detailed description).

Key words for video research on www.epilepticdisorders.com

Phenomenology: behavior altered (videos 1-5), eye deviation (video 1), head deviation (video 5), myoclonic seizure (videos 2-3), face (videos 1-5)
Localization: generalized (videos 1-5)/operculum (left) (videos 1 and 5)
Syndrome: perioral myoclonia with absences (videos 1-4), focal non-idiopathic frontal (FLE) (video 5)
Aetiology: genetic predisposition (videos 1-4), unknown

TEST YOURSELF

(1) Is perioral myoclonia with absences (POMA) a genetic generalised epilepsy (GGE) syndrome?

(2) What are the clinical and EEG features of POMA?

(3) What are the pitfalls in the diagnosis and management of POMA?

(4) What should be the diagnostic and management approach to POMA?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".