■ Clinical commentary

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Successful treatment of epilepsia partialis continua with perampanel: two pediatric cases

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ABSTRACT – Epilepsia partialis continua (EPC) is a form of focal motor status epilepticus, associated with multiple etiologies. Etiology-specific treatments, such as hemispherotomy for Rasmussen encephalitis, lesionectomy for focal cortical dysplasia, and metabolic correction for non-ketotic hyperglycemia, have proven to be efficacious in treating EPC, but, in general, EPC is difficult to treat and often drug-resistant, and there is little evidence to guide therapy. We report the successful treatment of EPC with perampanel in two pediatric patients. The first patient was a 12-year-old boy with neuronal ceroid lipofuscinosis (NCL) who started to have EPC around the age of 10 years, characterized by left hemifacial myoclonic twitches and hemibody jerks that were almost continuous throughout the day and disappeared during sleep. He had failed several antiepileptic drugs (AEDs). The EPC stopped within three days of initiating perampanel. The second patient was a six-year-old boy with *POLG*-related mitochondrial disease who presented to the emergency room with continuous jerky movements of the right arm and face after a trivial head injury. After failing several AEDs, including a midazolam drip, the EPC was controlled with perampanel. Both patients showed dramatic improvement and continue to show sustained efficacy after around five months of follow-up. Based on our observations, perampanel, which has a unique mechanism of action, appears to be a promising therapeutic option for treating EPC. [*Published with video sequence*].

Key words: perampanel; epilepsia partialis continua; EPC; children

VIDEO ONLINE

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Epilepsia partialis continua (EPC) is defined as "spontaneous regular or irregular clonic muscular twitching affecting a limited part of the body, sometimes aggravated by action or sensory stimuli, occurring for a minimum of one hour, and recurring at intervals of no more than ten seconds" [1-3]. The International League Against Epilepsy task force for classification of status epilepticus classifies it as focal motor status epilepticus [4]. The pathophysiology of EPC is still only partially understood, and it seems to have more than one mechanism; corticothalamic loops may be involved, and subcortical structures may play a role as well [5]. It is associated with multiple etiologies, including Rasmussen encephalitis, tick-borne encephalitis, stroke, focal cortical dysplasia, brain tumors, non-ketotic hyperglycemia, and mitochondrial diseases [6-8]. A comprehensive list of etiologies was presented in the review of Mameniskiene *et al.* [9]. An electroencephalogram (EEG) can show focal epileptiform abnormalities or generalized

background disturbances; it may even be normal [6, 8]. Etiology-specific treatments may be successful, but, in general, EPC is difficult to treat and often drug-resistant, and there is little evidence to guide therapy. Benzodiazepines, topiramate, and levetiracetam were found to be successful in a European survey [10]. Here, we report the findings of retrospective chart review of two pediatric cases in which EPC was successfully treated with perampanel.

Case studies

Case 1

The patient was a 12-year-old boy with drug-resistant epilepsy, global developmental delay, developmental regression, blindness, and a genetic diagnosis of neuronal ceroid lipofuscinosis (NCL) with a probable pathogenic variant, c.1367C>A (p.Ser456Tyr), detected in exon 11 of the *TPP1* gene in a homozygous state.

The patient was born at term, after an uneventful pregnancy and delivery, to second-degree consanguineous Saudi Arabian parents; his neonatal period was uneventful. There was no history of similar disease in the family. The onset of his symptoms occurred at around three and a half years of age. His initial symptoms included clumsiness and unsteady gait; these were soon followed by his first seizure at the age of four years. He experienced focal unaware behavioral arrest seizures that frequently evolved into bilateral tonic-clonic seizures. He started to present with developmental regression. His epilepsy soon became drug-resistant, and he developed a new type of seizure: frequent focal myoclonic jerks involving different parts of the body and occurring either on the right or left side. Around the age of 10 years, the patient started to experience left hemifacial myoclonic twitches and hemi-body jerks involving the abdomen and upper and lower extremities. These seizures initially occurred in clusters lasting up to 15 minutes but later increased in duration, becoming almost continuous throughout the day and disappearing during sleep. His EEG showed a severely suppressed background, intermittent generalized spikes and polyspike discharges, and periodic lateralized epileptiform discharges in the left frontal region. The clinical seizures did not have an abnormal ictal EEG correlate. These seizures did not respond to several antiepileptic drugs (AEDs), and the patient's mother reported that "they wake up when my child wakes up, and they sleep when he goes to sleep". During these seizures, there was no apparent change in baseline mental status, no cyanosis, and no change to the patient's vital signs.

His baseline examination indicated an encephalopathic child with no response to visual threat, an expressionless face, positive pyramidal signs, and near-continuous hemi-facial or hemi-body jerks. He showed no response to visual evoked potentials. Magnetic resonance imaging (MRI) of the brain showed generalized atrophy and diffuse white matter abnormalities with associated low-signal intensity in the thalamus on T2-weighted images, suggestive of NCL.

During this time, multiple AEDs were tried at the maximum tolerated dosages without any significant improvement, although lacosamide initially decreased the intensity of the seizures. Valproic acid, levetiracetam, topiramate, clonazepam, and phenytoin were inefficacious. A ketogenic diet was also tried but showed no benefits; vagus nerve stimulation (VNS) therapy was offered, but the patient's mother declined it. Recently, perampanel was introduced in our hospital, and we started the patient on 2 mg every night at bedtime for one week, increasing the dosage by increments of 2 mg every week until he was taking 6 mg every night at bedtime. The seizures, which were previously continuous, stopped within three days of initiating perampanel. The patient's mother described it as "magic". As per our instructions, the mother reached the goal dose of 6 mg although the patient became seizure-free at 2 mg. We did realize that we should have instructed the mother to stop escalating the dose if the seizures stop. The mother is now very hesitant to decrease the dose of perampanel since he is doing so well. The patient has now been seizure-free for almost five months except for one breakthrough EPC, two months ago, lasting for four days, which was attributed to sleep deprivation after long travel. After initiation of perampanel, valproic acid was successfully weaned.

Case 2

The patient was a six-year-old boy with drug-resistant epilepsy, mild global developmental delay, hypospadias, and a genetic diagnosis of mitochondrial disease with a pathogenic variant, c.3286C>T p.(Arg1096Cys), in the *POLG* gene in a homozygous state.

The patient was born at term, after an uneventful pregnancy and delivery, to non-consanguineous Saudi Arabian parents; he experienced an uneventful neonatal period. There was no history of similar disease in the family. He has seven healthy brothers and one healthy sister. The onset of his epilepsy occurred at the age of 14 months. He had two types of seizures: generalized atonic seizures presenting as drop attacks that occurred up to 30 times per day, and focal motor tonic-clonic status epilepticus lasting 30 to 90 minutes and occurring twice a month.

Long-term video-EEG monitoring suggested severe epileptic encephalopathy with recurrent generalized atonic and atypical absence seizures that correlated with bursts of diffuse spike/polyspike-wave discharges at 2-3 Hz. He failed to respond to multiple AEDs (phenobarbital, topiramate, levetiracetam) until clobazam was initiated, which made him completely seizure-free. A physical examination revealed dysmorphic features in the form of almond eyes, a fish-like mouth, generalized hypotonia, and absent reflexes, but otherwise non-focal.

The patient remained seizure-free for around four months until he presented in the emergency room with continuous jerky movements of the right arm and face; the seizures started after a trivial head injury. The patient was treated as a case of focal motor aware status epilepticus. After failing to respond to first- and second-line AEDs (lorazepam, levetiracetam, phenobarbital), he was intubated and started on a midazolam drip that was gradually increased to 24 micrograms/kg/min. The patient continued to experience seizures in the form of head nodding and right shoulder jerking for three days. A computed tomography (CT) head scan was normal, and an EEG showed baseline interictal discharges with no abnormal ictal electrographic correlate. Clobazam was continued, and phenobarbitone was added. He was also started on L-carnitine and vitamin B6. At this stage, the seizures were characterized as EPC; he was subsequently extubated and weaned off the midazolam drip. The patient was discharged home on clobazam, phenobarbitone, and vitamins. Based on telephone follow-up, two weeks later, his mother reported that his symptoms had continued after discharge from the hospital and were present continuously during wakefulness and sleep to a degree that limited the patient's ambulation (*see video*). The patient was started on 2 mg of perampanel every night at bedtime for a week, which was increased by increments of 2 mg per week until he was taking 6 mg every night at bedtime. One week after the initiation of perampanel, the mother reported that his EPC had completely disappeared. He has now been on perampanel for five months. During a follow-up telephone call, three months after initiating perampanel, his mother reported that he was doing very well but was experiencing some twitching of the right arm and face, similar to the EPC. These seizures lasted three to five seconds and occurred every 30 minutes during sleep. When he was awake, they occurred when he tried to use his right arm, such as holding a glass. They lasted for a few seconds and then stopped. During the latest follow-up visit, five months after initiating perampanel, the mother reported that the EPC had completely stopped.

Discussion

EPC is a form of focal motor status epilepticus characterized by recurrent focal myoclonic jerks that occur over a prolonged period of time and are typically resistant to pharmacotherapy. Etiology-specific treatments, such as hemispherotomy for Rasmussen encephalitis, lesionectomy for FCD, and metabolic correction for non-ketotic hyperglycemia, have proven to be efficacious in treating EPC [6, 8, 11, 12]. Various other therapies have been reported to successfully stop EPC, but there are too few cases to reach any definite conclusions [13-17] (*table 1*). Neurostimulation techniques, such as vagus nerve stimulation [18, 19], transcranial magnetic stimulation [20] (Rotenberg *et al*., 2009) and transcranial direct current stimulation [21], have also been used to successfully treat EPC. Immunomodulatory treatments have also been used in cases of Rasmussen encephalitis when early surgery was not feasible and in late-onset patients with slower disease progression [13]. Various AEDs, including levetiracetam, topiramate, lacosamide, and primidone, have been reported to successfully treat EPC [10, 22-26], but so far there is little evidence for a drug of choice. Overall, the results of treatment are discouraging, and the outcome is often poor [27].

It has been postulated that an increase in the rate of intracellular accumulation of GABAA receptors may decrease GABA inhibition during prolonged seizures in status epilepticus [28]. Activation of NMDA receptors can enhance AMPA receptor-mediated transmission during prolonged seizures [29, 30]. Perampanel is a highly selective, non-competitive α-amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid (AMPA) receptor antagonist that reduces the neuronal hyperexcitation associated with seizures by targeting glutamate activity at AMPA receptors on postsynaptic membranes [31]. Randomized double-blind placebo-controlled trials have demonstrated the efficacy of perampanel for treating focal seizures [32, 33] and tonic-clonic seizures in idiopathic generalized epilepsy in patients 12 years and older [32]. Perampanel has recently been approved for focal seizures in patients four years and older [34] (https://www.fycompa.com/hcp/efficacy/poshttps://www.fycompa.com/hcp/efficacy/pos). Retrospective observational studies and case series have also demonstrated the efficacy of perampanel for treating myoclonic and absence seizures in patients with idiopathic generalized epilepsy [35], drug-resistant myoclonic seizures [36], and progressive myoclonic epilepsy [37-39]. The two cases presented are the only EPC patients we treated with perampanel. To our knowledge, there are as yet only two reports in the literature demonstrating successful treatment of EPC with perampanel in adult patients (*table 2*).

▼ **Table 1.** Successful treatment options for EPC reported in the literature.

Type of treatment	Treatment	Etiology	Reference
Surgery	Hemispherotomy	Rasmussen encephalitis	[6, 7, 11]
	Focal resection	Focal cortical dysplasia	[12]
		Sturge-Weber syndrome	[40]
	Subpial dissection	Rasmussen encephalitis	[40]
Neurostimulation	Chronic cortical stimulation	Unknown Old ischemic insult	[19]
	rTMS	Non-specific	[20]
	tDCS	POLG-related mitochondrial disease	[21]
		Autoimmune epilepsy	[41]
	Vagus nerve stimulation	Rasmussen encephalitis Chronic inflammatory encephalitis	[18]
AEDs	Perampanel	Meningioma	[42]
		Rasmussen encephalitis	[43]
	Levetiracetam	Oligodendroglioma	[22]
		ICH	[23]
		Non-specific	[10]
	Topiramate	Non-specific	[24]
	Lacosamide	Subdural hematoma	[25]
	Primidone	Unknown	[26]
Immunotherapy	Immunomodulatory treatments	Rasmussen encephalitis	[13]
Others	Botulinum toxin	Rasmussen encephalitis	[14]
		Anaplastic astrocytoma	[16]
	Metabolic correction	Non-ketotic hyperglycemia	[15] [17]

AEDs: antiepileptic drugs; ICH: intracerebral hemorrhage; rTMS: transcranial magnetic stimulation; tDCS: transcranial direct current stimulation.

▼ **Table 2.** Clinical characteristics.

Description	Case 1	Case 2	Patient A [42]	Patient B [43]
Age (years)	12	7	76	44
Etiology of EPC	Neuronal ceroid lipofuscinosis	*POLG*-related mitochondrial disease	Remnants of meningioma and surrounding edema	Rasmussen encephalitis
Onset of EPC	Chronic (2 years ago)	Acute	Acute	Acute
Trigger	None	Trivial head injury	None	None
Semiology	Hemi-body jerks	Right arm and face jerks	Clonic movements of right hand, forearm and hemiface	Left face myoclonic jerks
Duration of EPC	Continuous all day and disappear during sleep	Continuous	Continuous	Recurrent all day long with each episode lasting 20 seconds

▼ **Table 2.** Clinical characteristics (*continued*).

Description	Case 1	Case 2	Patient A [42]	Patient B [43]
EEG	No abnormal electrographic correlation with EPC	No abnormal electrographic correlation with EPC	No abnormal electrographic correlation with EPC	N/A
Other seizure types	Myoclonic GTC Focal tonic	Generalized atonic Atypical absence	None	Tonic Generalized status epilepticus
Failed AEDs	LEV, TOP, CZP, LCM, PHT, VPA	Lorazepam, LEV, PHB, CLB, Midazolam infusion	CZP, LEV, LCM	LTG, LEV, CZP, PHT, VPA and PGB
Efficacy of PER	EPC stopped within three days of initiating PER 2 mg	EPC stopped one week after initiating PER and reaching 4 mg	EPC improved on PER 2 mg and stopped on 4 mg	EPC stopped within three days of initiating PER 6 mg
Follow up	5-month follow up – Seizure-free except one breakthrough EPC lasting for 4 days due to sleep deprivation after long travel	3-month follow up: developed twitching of right arm and face similar to EPC lasting 3-5 seconds and occurring every 30 minutes during sleep 5-month follow up: EPC resolved	6-month follow up: seizure-free but due to irritability, PER was tapered and EPC recurred, so PER was reintroduced slowly to reach 4 mg 1-year follow up: seizure-free	1-year follow up: seizure-free
Brain MRI	Generalized atrophy, diffuse white matter abnormalities with associated low-signal intensity in the thalamus	Few scattered T2 hyper intense foci in the cerebellum	Status post resection of left frontal meningioma	Right hemispheric atrophy pronounced in the frontal and temporal regions

EPC: epilepsia partialis continua; PER: perampanel; EEG: electroencephalogram; GTC: generalized tonic-clonic; AEDs: antiepileptic drugs; LEV: levetiracetam; VPA: valproic acid; CZP: clonazepam; LCM: lacosamide; PHT: phenytoin; PHB: phenobarbital; CLB: clobazam; LTG: lamotrigine; TOP: topamax; PGB: pregabalin; N/A: not available; MRI: magnetic resonance imaging.

In this article, we report the successful treatment of EPC in two pediatric patients, one with NCL and the other with *POLG*-related mitochondrial disease. Both patients showed dramatic improvement, indicating the efficacy of perampanel. Based on our observations, perampanel, which has a unique mechanism of action, appears to be a promising therapeutic option for treating EPC. Larger controlled studies are needed to determine its long-term efficacy. ■

Ethical statement.
The investigation was carried out in accordance with the latest version of the Declaration of Helsinki and was approved by the local Review Board (King Fahad Specialist Hospital Dammam).

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

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Legend for video sequence

EPC in right upper extremity.

Key words for video research on www.epilepticdisorders.com

Phenomenology: myoclonic seizure
Localization: not applicable
Syndrome: epilepsia partialis continua
Aetiology: ceroid-lipofuscinosis, mitochondrial disorder