Epilepsia partialis continua: semiology and differential diagnoses

Christian G. Bien, Christian E. Elger
Dept. of Epileptology, University of Bonn, Germany
Received October 12, 2007; Accepted December 20, 2007

ABSTRACT – Epilepsia partialis continua (EPC) is a rare form of focal status epilepticus. It may have vascular, immune-mediated, neoplastic or metabolic-toxic causes. The origin of EPC has been linked with the motor cortex. This has been solidly supported by sophisticated electrophysiological studies. Here, a series of video sequences from patients with EPC (due to Rasmussen encephalitis, early-stage multiple sclerosis, and steroid responsive encephalopathy with autoimmune thyroiditis), and other cases with repetitive myoclonic jerks or movement disorders (myoclonic epilepsy associated with ragged-red fibers, Jacksonian march, myoclonic seizures in other types of frontal lobe or idiopathic generalized epilepsies, and different types of tremor) is presented.

Key words: epilepsia partialis continua, semiology, diagnosis

In 1894-1895, Koževnikov (or Koshewnikow) reported in Russian and in German on four patients who suffered from mild to moderate hemiparesis and from what he called “epilepsia corticalis sive partialis continua” in the paretic parts of the body (Koževnikov 1894, Koshewnikow 1895). Today, only the term epilepsy partialis continua (EPC) is still in use. The Russian neurologist conceptualized EPC as a particular form of epilepsy and distinguished it from “common epilepsy” because it did not have the tendency to spread and was often not self-limiting. It was clear to Koževnikov that in the four patients he described, the EPC originated in the hemisphere contralateral to the affected side of the body, close to the primary motor area. He was, however, unable to clarify the nature of the organic brain lesion, which in his opinion was underlying the continuous myoclonic jerks. Based on the slowly progressive nature of the disease in his patients, he hypothesized that a chronic encephalitic process was at work. He was probably right because it can be assumed that the majority of his patients suffered from tick-borne, viral Russian spring summer meningoencephalitis, which was detected only some decades later (Omorokow 1927, Smorodintsev 1958).

In contrast to Koževnikov’s terminology, EPC is today conceptualized as a seizure type rather than a form of epilepsy, and is subsumed under the group of focal status epilepticus (Engel 2001). Following Koževnikov’s publications, subsequent research on EPC has mainly dealt with three – in part interrelated – questions: how should EPC be defined? Which pathological processes cause EPC? Where in the nervous system does this phenomenon arise?

The current, broadly accepted answers to these questions are very briefly...
summarized in the following paragraphs. Thereafter, a
description of the semiological features of EPC and possi-
ble differential diagnoses are given, with a video sequence
illustrating them (see video sequence).

Definition of EPC

Basically, two types of definitions have been suggested:
some authors have confined themselves to defining EPC
only on clinical (semiological) grounds, whereas others
have demanded additional electrophysiological evidence
regarding the cortical origin of the phenomena to be
considered. Combining the two most frequently used
clinical definitions (Thomas et al. 1977, Obeso et al. 1985)
results in the following: “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”.
The neurophysiological conceptualization of EPC is com-
licated – or rather: enriched – by the fact that it has been
studied from the viewpoint of both epilepsy and move-
ment disorder research. Depending on their main field
of interest, authors may – in addition to the aforementioned
clinical features – require the demonstration of epilepti-
form EEG abnormalities (ideally in a fixed temporal cou-
pling to the muscle jerks), or abnormalities as shown by
other electrophysiological studies such as giant somatosensory-evoked potentials [SSEPs]) in order to
demonstrate the cortical origin of the muscle jerks
(Cockerell et al. 1996).

Pathological processes underlying EPC

The three largest EPC series available – 32 patients (Tho-
mas et al. 1977), 40 patients (Cockerell et al. 1996), and 76
patients (Sinha and Satishchandra 2007) – came to
surprisingly homogenous results regarding the causes of
EPC: vascular disorders (stroke, intracranial bleeding, ce-
rebral venous thrombosis, vasculitis: 24-28%), encephal-
itides (Rasmussen encephalitis the most common cause
of EPC in childhood), or infectious encephalitides: 15-19%),
neoplasms (glioma, haemangioblastoma, menin-
gioma, lymphoma: 5-16%), and metabolic disorders
(diabetic non-ketotic hyperosmolar coma, mitochondrial-
opathy, Alpers syndrome, intoxications: 6-14%). In 19-
28% of cases, the authors could not determine the cause of
the EPC.

A painstaking compilation of case reports and small case
series (n = 162) in an excellent review article from the
1970s resulted in the following figures: inflammatory dis-
orders 32%, neoplastic disorders 19%, head trauma 16%,
vascular disorders 14%, others 3%, unknown 16% (Löhler
and Peters 1974).

The site of origin of EPC

Virtually all authors today agree that a contribution from the
primary motor area is indispensable for the generation of EPC,
not to say that the cortex is its main or even single generator.
(Some decades ago, an influential publication had suggested
that subcortical structures play the main role in generation of
EPC [Juul-Jensen and Denny-Brown, 1966]). Evidence for a
cortical dysfunction underlying EPC comes from studies
showing epileptiform EEG potentials recorded by surface
or even subdural electrodes time-locked to the muscle
jerks, giant SSEP over the suspicious cortex, or enhanced C
reflexes. Further corroborating evidence comes from neu-
oradiological or neuropathological studies (Thomas et al.
1977, Wieser et al. 1978, Obeso et al. 1985, Cockerell et
al. 1996). The duration of the muscle twitches, as docu-
mented by EMG, has been reported to be brief in cortically-generated myoclonic jerks including EPC
(< 100 ms) compared to subcortically-arising jerks
(> 100 ms). The muscle jerks affect agonistic and antago-
nistic muscles simultaneously (Obeso et al. 1985, Cocker-
ell et al. 1996).

Semiology of EPC

Typical clinical signs of EPC are: combination of the repetitive
myoclonic jerks with hemiparesis or – less frequently – with
other cortically-generated deficits; monomorphistic, simple,
brief excursions of the affected limb; regular or irregular
occurrence of the jerks; involvement of distal rather than
proximal muscle groups; physical exercise, sensitive stimula-
tion or psychic exertion may increase the amplitude and
frequency of the myoclonic jerks; more frequent involvement
of the upper than the lower half of the body (in 151 patients,
the distribution was as follows: head: 16%; head and upper
extremity: 14%; upper extremity: 40%; trunk: 5%; lower
extremity: 14%; whole side of body: 11% - the sum is larger
than 100% because multiple sites may have been affected
during the disease course [Löhler and Peters 1974]); EPC
more often continues during sleep – sometimes in a milder
form – rather than stops (25 versus 5 patients from the
literature according to [Löhler and Peters 1974]). The
same authors calculated a mean frequency of 90 jerks per
minute (i.e. 1.5 Hz; range 3-360 per minute, equaling
0.06-6 Hz) from the literature. There is no preference for
one side of the body or the other.

Differential diagnoses

In the second part of the video sequence and the accom-
panying table (table 1), differential diagnoses for EPC are
presented. Tremor has a characteristic alternating agonist-
antagonist innervation pattern (patients 13, 15). The corti-
cal myoclonic jerks of patients with myoclonus epilepsies
affect the limbs diffusely (patient 14). Sometimes, even extensive investigations do not clarify the origin of the repetitive myoclonic jerks as in patient 12; the long duration of the bursts mitigates against a typical cortical origin. The Jacksonian march (patient 16) is clearly distinguishable because of the sequential involvement of unilateral body parts.

**Summary**

EPC is a rare type of focal status epilepticus. Characteristic semiological features enable it to be diagnosed and distinguished from other movement disorders or myoclonic symptoms. Additional electrophysiological studies can contribute to an enhanced degree of certainty regarding the cortical origin of the myoclonic jerks. In childhood, the most frequent cause of EPC is Rasmussen encephalitis (Schomer 1993). At older ages, vascular and neoplastic diseases are the most frequent causes. Metabolic, toxic and other immune-mediated causes account for most of the remaining cases. The search for the underlying disease is therefore of paramount importance.

**Acknowledgements**

The authors would like to thank Mrs. Sandra Parke and Mr. Theo Kapp for expert technical assistance.

**References**


---

**Legend for video sequence**

In the attached video sequence, we present video EEG recordings of patients with repetitive involuntary jerks:

Eleven patients with EPC, defined according to clinical criteria (all patients with the diagnosis of EPC and an adequate video EEG recording from this center during the period September 2000 until March 2007), and 5 patients with other types of myoclonia or movement disorders are included.

Ten of the EPC patients have been diagnosed with immune-mediated disorders; one patient’s EPC was of unknown cause.

In some patients, additional electrophysiological studies were performed. For details, see table 1.

The apparently long duration of the EMG jerks on our recordings are probably best explained by the peculiarities of recordings with an EEG system with a lower sampling rate (here: 200 Hz), and filter settings different from those usually applied in pure EMG recordings.

SSEPs on stimulation of the median nerves were considered as giant potentials if the amplitude of P25-N33 was > 12 μV (Obeso et al. 1985). The overrepresentation of inflammatory conditions and the lack of vascular diseases in this series is due to the patient population from this tertiary center, with its particular interest in immune-mediated syndromes and a (surprisingly) low number of referrals of elderly patients. For recording montages, see figures 1 and 2; in the split-screen videos, red curves represent left hemispheric leads, and blue curves represent right hemisphere leads. EEG channels with epileptiform activity, and EMG channels are marked by red colour.
Figure 1. 23 EEG channels.

Figure 2. 16 EEG channels.

ADM: musculus abductor digiti minimi
AH: musculus abductor hallucis (foot)
APB: musculus abductor pollicis brevis (hand)
centr: central
EPC: epilepsia partialis continua
F: female
FCU: musculus flexor carpi ulnaris
FLE: frontal lobe epilepsy
fr: frontal
hem: hemisphere
Hypoth: hypothenar
IGE: idiopathic generalized epilepsy
L: left
LTC: lamotrigine
M. add. long.: musculus adductor longus
M. grac.: musculus gracilis
M. orb. oc.: musculus orbicularis oculi
M. vast. lat.: musculus vastus lateralis
M: male
MERRF: myoclonic epilepsy-associated with ragged-red fibers
mo: months
MS: multiple sclerosis
Nd: not done
R: right
RE: Rasmussen encephalitis
SREAT: steroid responsive encephalopathy with autoimmune thyroiditis (formerly Hashimoto’s encephalopathy)
SSEPs: somatosensory-evoked potentials
sync: synchronous jerks in the recorded muscles
temp: temporal
VPA: valproic acid
wks: weeks
yrs: years
# Table 1. Patients shown in video with epilepsia partialis continua and with other conditions.

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (yrs)</th>
<th>Gender</th>
<th>Diagnosis</th>
<th>Time since onset</th>
<th>Body region affected</th>
<th>Epileptiform potentials on EEG</th>
<th>EMG bursts: apparent duration and synchronicity</th>
<th>Amplitude of SSEPs on stimulation of median nerve P25-N33 (µV) Ipsilateral-contralateral</th>
<th>Additional neurological deficit</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Epilepsia Partialis Continua</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1, M</td>
<td>11.3</td>
<td>M</td>
<td>RE, R hem</td>
<td>3.0 yrs</td>
<td>L face</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>L hemiparesis</td>
</tr>
<tr>
<td>2, F</td>
<td>11.4</td>
<td>F</td>
<td>RE, L hem</td>
<td>13 mo</td>
<td>R tongue</td>
<td>Yes</td>
<td>Nd</td>
<td>Normal</td>
<td>R hemiparesis</td>
</tr>
<tr>
<td>3, M</td>
<td>13.2</td>
<td>M</td>
<td>RE, L hem</td>
<td>4.0 yrs</td>
<td>R face</td>
<td>No (artifacts)</td>
<td>&lt; 200 ms (only one muscle)</td>
<td>30-5</td>
<td>R hemiparesis</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Same patient, 1.8 yrs later</td>
<td></td>
<td></td>
<td>R face</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>R hemiparesis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4, M</td>
<td>18.6</td>
<td>M</td>
<td>Unclear</td>
<td>3 yrs</td>
<td>Face, pectoral muscle, L arm</td>
<td>No</td>
<td>&lt; 150 ms, sync</td>
<td>12.5-1.8</td>
<td>None</td>
</tr>
<tr>
<td>5, M</td>
<td>10.5</td>
<td>M</td>
<td>RE, L hem</td>
<td>2.2 yrs</td>
<td>R hand</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>R hemiparesis</td>
</tr>
<tr>
<td>Same patient, 8 mo later</td>
<td></td>
<td></td>
<td>R hand</td>
<td>Yes</td>
<td>&lt; 200 ms, sync</td>
<td>87-7</td>
<td>R hemiparesis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6, F</td>
<td>23.8</td>
<td>F</td>
<td>Early-stage MS</td>
<td>Days</td>
<td>L hand</td>
<td>No</td>
<td>&lt; 250 ms, sync</td>
<td>Normal</td>
<td>None</td>
</tr>
<tr>
<td>7, F</td>
<td>62.8</td>
<td>F</td>
<td>SREAT (Hashimoto’s)</td>
<td>3 wks</td>
<td>L hand</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>Dystonia L hand</td>
</tr>
<tr>
<td>8, F</td>
<td>42.1</td>
<td>F</td>
<td>RE, L hem</td>
<td>26 mo</td>
<td>R hand</td>
<td>No</td>
<td>Nd</td>
<td>Normal</td>
<td>R hemiparesis</td>
</tr>
<tr>
<td>Same patient, 1.8 yrs later</td>
<td></td>
<td></td>
<td>R leg</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>R hemiparesis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9, M</td>
<td>6.3</td>
<td>M</td>
<td>RE, L hem</td>
<td>1 mo</td>
<td>R foot and leg</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>R hemiparesis</td>
</tr>
<tr>
<td>10, F</td>
<td>5.1</td>
<td>F</td>
<td>RE, L hem</td>
<td>10 mo</td>
<td>R hand and foot</td>
<td>Yes</td>
<td>&lt; 150 ms, sync</td>
<td>Normal</td>
<td>R hemiparesis</td>
</tr>
<tr>
<td>11, F</td>
<td>16.6</td>
<td>F</td>
<td>RE, R hem</td>
<td>3.3 yrs</td>
<td>L hand and foot</td>
<td>No</td>
<td>&lt; 150 ms, sync</td>
<td>Normal</td>
<td>L hemiparesis</td>
</tr>
</tbody>
</table>

Differential diagnoses

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (yrs)</th>
<th>Gender</th>
<th>Diagnosis</th>
<th>Time since onset</th>
<th>Body region affected</th>
<th>Epileptiform potentials on EEG</th>
<th>EMG bursts: apparent duration and synchronicity</th>
<th>Amplitude of SSEPs on stimulation of median nerve P25-N33 (µV) Ipsilateral-contralateral</th>
<th>Additional neurological deficit</th>
</tr>
</thead>
<tbody>
<tr>
<td>12, F</td>
<td>18.9</td>
<td>F</td>
<td>Myoclonia of unknown origin. FLE or IGE</td>
<td>5.9 yrs</td>
<td>L leg</td>
<td>No</td>
<td>&lt; 600 ms, sync</td>
<td>Nd</td>
<td>None</td>
</tr>
<tr>
<td>13, F</td>
<td>2.8</td>
<td>F</td>
<td>Tremor ataxia teleangiectasia</td>
<td>2 mo</td>
<td>L arm</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>Ataxia</td>
</tr>
<tr>
<td>14, M</td>
<td>23.3</td>
<td>M</td>
<td>Cortical myoclonia MERRF</td>
<td>14 yrs</td>
<td>Diffuse</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>Ataxia, paraparesis</td>
</tr>
<tr>
<td>15, M</td>
<td>26.8</td>
<td>M</td>
<td>Tremor VPA-LTG overdose</td>
<td>Weeks</td>
<td>Upper extremities</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>None</td>
</tr>
<tr>
<td>16, M</td>
<td>21</td>
<td>M</td>
<td>Jacksonian march. FLE</td>
<td>9 yrs</td>
<td>L hand, spread to rest of hemibody</td>
<td>Late in seizure</td>
<td>Nd</td>
<td>Nd</td>
<td>None</td>
</tr>
</tbody>
</table>