

# A patient with two episodes of epilepsy partialis continua of the abdominal muscles caused by cortical dysplasia

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**ABSTRACT** – Epilepsia partialis continua (EPC) is a rare form of focal motor status epilepticus. There is typically a predilection for facial and distal limb involvement, but rarely trunk or abdomen muscles may be affected. Rarely, EPC may also present in association with cortical dysplasia. In this report, we describe the clinical, neuroimaging and ictal electroencephalographic findings of a young woman presenting with persistent myoclonic twitches of the abdominal muscles that were considered to represent a rare manifestation of EPC due to cortical dysplasia. To the best of our knowledge, this is the first report of malformation of cortical development causing abdominal myoclonus. *[Published with video sequences]*

**Key words:** epilepsy partialis continua (EPC), dysplasia, abdomen, trunk, status

Epilepsia partialis continua (EPC) is a rare form of focal motor status epilepticus that is characterized by continuous jerking of a defined part of the body lasting at least one hour and recurring with less than 10 seconds intervals (Thomas *et al.* 1977). There is typically a predilection for face and distal limb involvement, but rarely the trunk or the abdomen may also be affected. Although the anatomic localization of the epileptogenic zone has been debated, circumscribed frontal parasagittal or parietal lesions around the central lobe have been documented in a few cases with EPC involving the abdominal or truncal muscles (Rosenbaum and Rowan 1990, Matsuo 1984, Chalk *et al.* 1991, Gurer *et al.* 2001, Fernandez Torre *et al.* 2004, Dafotakis *et al.* 2006).

The causes of EPC in adults include a variety of conditions such as tumors, vascular lesions, trauma, infections and metabolic disorders (Gurer *et al.* 2001). Rarely, EPC may also present in association with cortical dysplasia (Fusco *et al.* 1992, Kuzniecky and Powers 1993, Gurer *et al.* 2001, Nakken *et al.* 2005).

In this report, we describe the clinical, neuroimaging and ictal electroencephalographic findings of a young woman presenting with two episodes of persistent myoclonic twitches of the abdominal muscles. To the best of our knowledge, this is the first report of abdominal EPC caused by abnormal cortical development.



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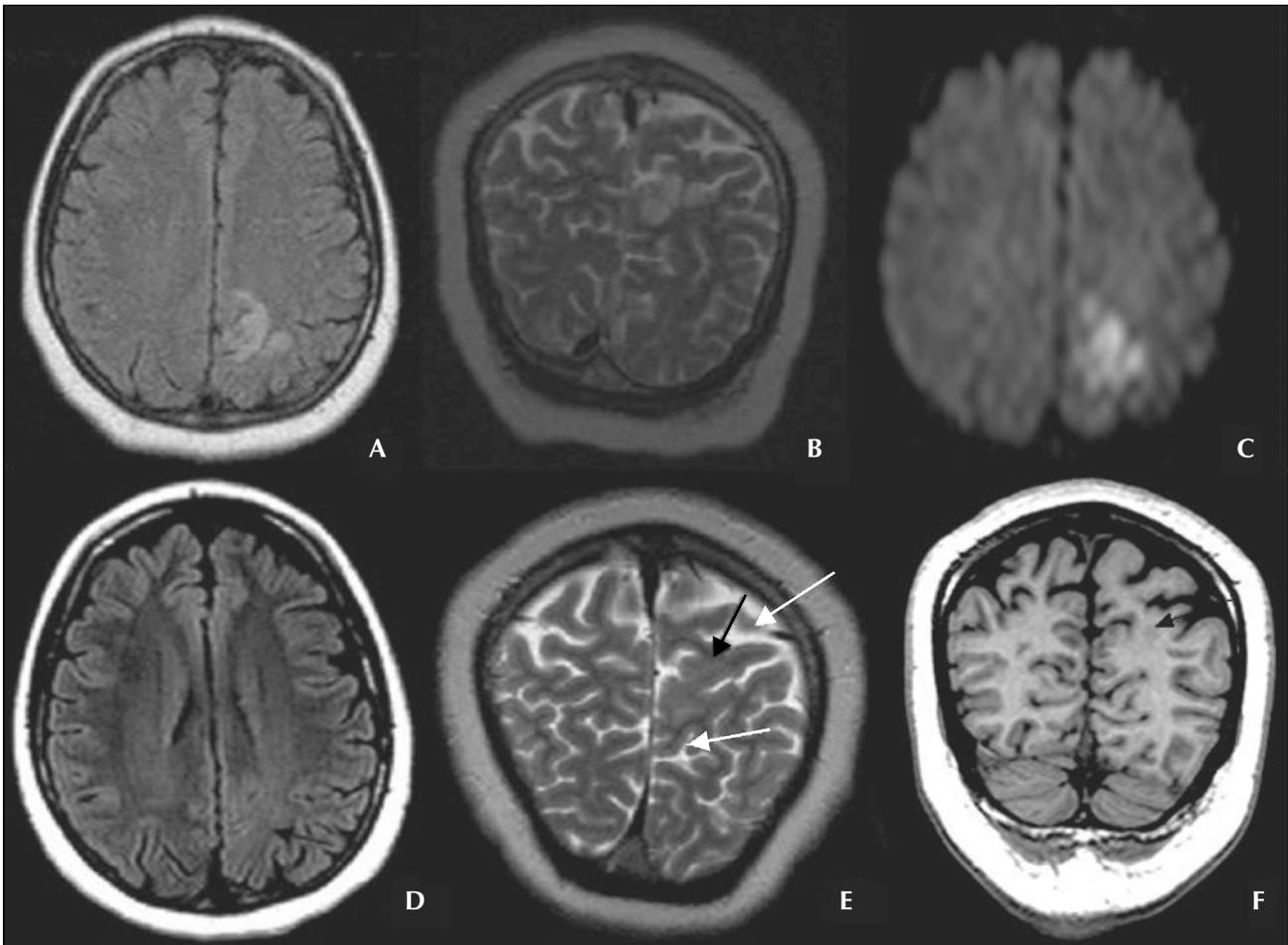
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## Case report

A twenty five-year-old female with a diagnosis of epilepsy was admitted to our hospital because of an increased seizure frequency for a week. She had had multiple episodes of palpitations in her chest, twitching of the right side of her abdomen, and occasionally, mild contractions on the right side of her face and bilateral blinking accompanied the abdominal contractions. Her neurological and physical examinations were normal. The myoclonic twitches primarily involved the abdominal muscles on the right and were usually semi-regular with a frequency of approximately 1-2 Hz. Consciousness was not altered. Her family history was unremarkable. The seizures had appeared when she was two years old. She had suffered from increased seizure frequency and myoclonic twitches of abdomen and face for a week during her pregnancy

(five years prior to this admission). During that time magnetic resonance (MR) imaging of the brain, revealed cortical thickening with an increased T2 signal at the medial aspect of the left cerebral hemisphere, centered around the parietooccipital sulcus with restricted diffusion (figure 1A-C). The findings were then thought to represent acute ischemic changes.

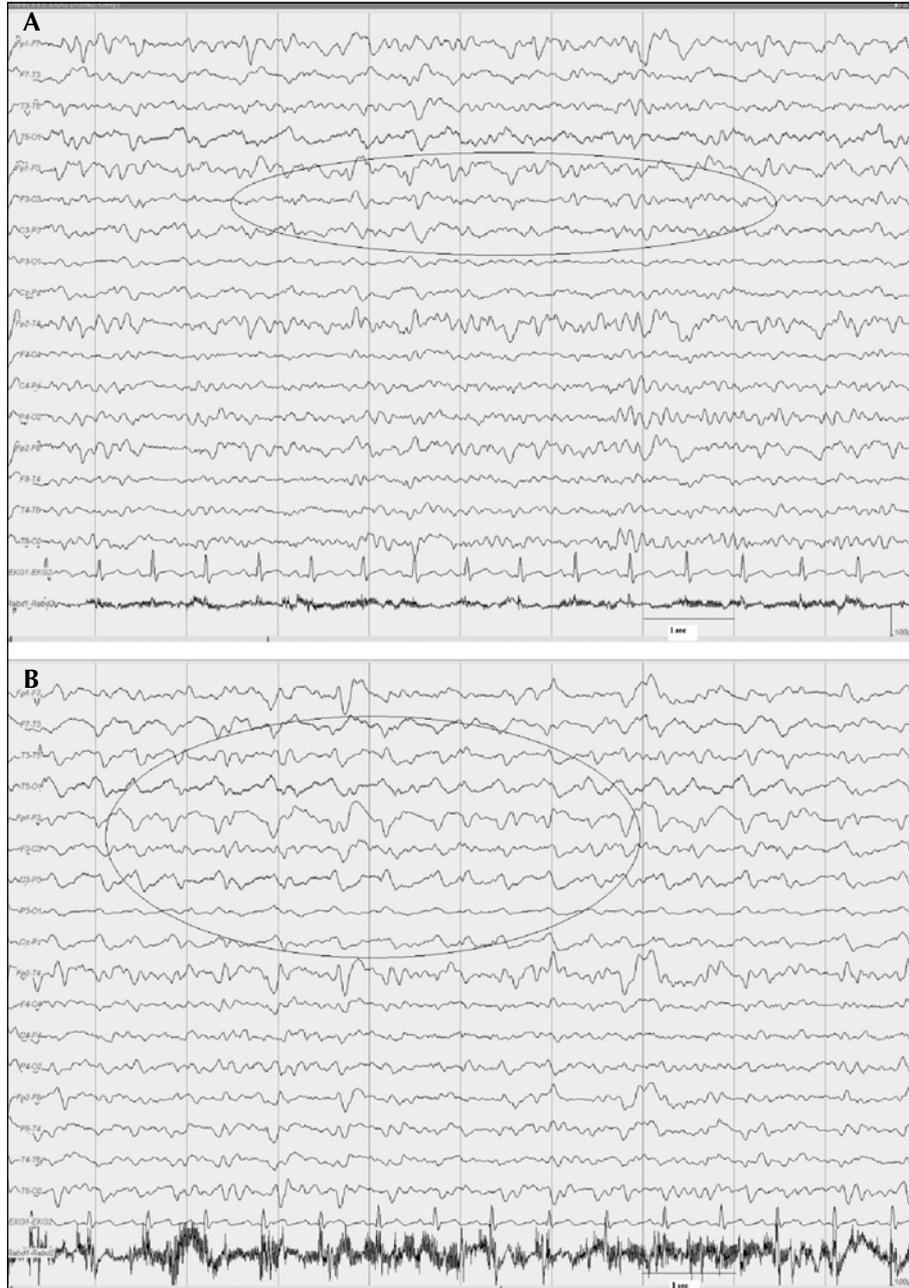
The EEG recordings during the previous admission were obtained on a digital EEG machine with a high frequency filter at 70 Hz and low frequency filter at 0.3 Hz; 10mm/sec in rate. Recordings were performed, while the abdominal EPC was video-taped using a hand-camera. Some samples of the EEG and video were not time-related. In order to include the best example of the the abdominal EPC, we selected the video that was recorded just prior to EEG recording (see video sequence). The interictal EEG showed PLEDs consisting of burst of sharp slow waves



**Figure 1.** Imaging of the brain at the time of the first admission, during her pregnancy (A-C). Increased T2 signal and cortical thickening can be seen on the axial FLAIR (A) and coronal T2-weighted images (B). There is also increased signal on the TRACE images of the diffusion sequence (C) with accompanying decreased signal of the ADC map (not shown). MR imaging of the brain performed at last admission (five years after her first MRI) (D-F). Increased T2 signal and loss of white-gray matter differentiation can be seen (arrow) at the left parietal region on the axial FLAIR sequence (D) and on the coronal T2 sequence (E) as well as on the volumetric T1-weighted images (F). The cortical dysplasia is located just above the parieto-occipital sulcus (straight white arrow) and inferomedial to the interparietal sulcus (dotted white arrow).

involving the left frontocentral and parietal areas (figure 2A). These discharges were, at times, greater than at other times, and showed rhythmicity on the left

frontocentro-parietal region. During the period of increasing frequency of the abdominal myoclonus and mild facial myoclonic twitchings, periodic discharges were recorded



**Figure 2.** The EEG was recorded, while the abdominal EPC was video-taped using a hand-camera. Thus, the samples of EEG and video were not time-related. In order to include the best example of the recorded video, we had to select the attached video sequence that was recorded just prior to EEG recording. EEG recording was then performed after placement of EEG electrodes. **A)** Interictal EEG showed PLEDs consisting of burst of sharp slow waves (1-2 Hz) primarily involving the left frontocentral and parietal areas. **B)** EEG example of the ictal period with an increase in the frequent abdominal myoclonus and mild myoclonic twitchings on the right side of face. The periodic discharges consisting of high amplitude sharp waves and slow waves (prominent on the frontocentral and parietal areas) over the whole left hemisphere. (HF: 70 Hz, LF: 0.3 Hz; rate: 10 mm/sec).

over the whole left hemisphere (*figure 2B*). These consisted of high amplitude sharp waves and slow waves, prominent on the frontocentral and parietal areas. Cranial MR imaging obtained on the first day of the frequent seizures showed increased T2 signal, cortical thickening and loss of gray-white matter differentiation at the left mesial parietal region, just above the parieto-occipital sulcus (*figure 1D-F*), corresponding to the area of restricted diffusion on the previous MR imaging of the brain. The radiological findings were consistent with cortical dysplasia.

She was treated initially with parenteral valproic acid and consequently with a combination of valproic acid, levetiracetam and carbamazepine. The seizures were completely controlled within two days.

## Discussion

Our patient had two specific seizure episodes, with a five-year interval, that could be defined as EPC with primarily, involvement of the right side of the abdominal muscles. Occasionally, the right side of the facial muscles and bilateral eyelid muscles were also involved. EPC consisting of truncal-abdominal muscle contraction without involvement of distal limbs or face has been rarely reported (Matsuo 1984, Rosenbaum and Rowan 1990, Chalk *et al.* 1991, Gurer *et al.* 2001, Fernandez Torre *et al.* 2004, Dafotakis *et al.* 2006). The area of cortical representation of the truncal and abdominal muscles is much smaller than the representation of distal limb and face muscles (Penfield and Rasmussen 1950). Circumscribed frontal parasagittal or parietal lesions, either neoplastic (Matsuo 1984, Rosenbaum and Rowan 1990, Gurer *et al.* 2001, Fernandez Torre *et al.* 2004), cystic (Gurer *et al.* 2001) or infectious (Matsuo 1984, Chalk *et al.* 1991) have been well-documented as causes of abdominal-truncal EPC (table 1). Thus the MR imaging findings of parietal cortical thickening and subcortical lesions were compatible with the diagnosis of cortical dysplasia.

MR imaging findings in cortical dysplasia can be variable, ranging from obvious abnormalities to normal appearance, and most of the time the findings are not specific. Therefore, low grade cortical neoplasias and cortical infarct sequelae may be included in the differential diagnosis. However, the lack of expansion and mass effect makes cortical neoplasia such as a neuroglial tumor unlikely in our patient. Similarly, focal cortical infarct with resultant encephalomalacia is not likely due to the absence of loss of volume at the site of restricted diffusion on the follow-up imaging. Although the first neuroimaging changes had not been followed up, the diffusion restriction at the site of the cortical dysplasia was probably a transient change, representing cytotoxic edema due to the recent, frequent seizure activity. Similarly Dafotakis *et al.* reported a case defined as idiopathic EPC of the abdominal muscles that

had transient MRI abnormalities around the precentral gyrus (Dafotakis *et al.* 2006).

Although cortical dysplasia is an increasingly recognized cause of chronic epilepsy involving focal seizures, there are only a few reports describing patients with cortical dysplasia presenting as EPC on mostly distal parts of body (Fusco *et al.* 1992, Kuzniecky and Powers 1993, Gurer *et al.* 2001, Nakken *et al.* 2005). To the best of our knowledge, our patient is the first case of malformation of cortical development resulting in abdominal myoclonus. With the underlying structural malformation of the brain, any acute metabolic abnormality may contribute to decrease the seizure threshold. Our patient was pregnant at the time of her first episode of EPC when her antiepileptic drug level might have been lower, but we could find no precipitating factor for the second round of EPC. However, focal cortical dysplasia may be highly epileptogenic in itself, or may lead to deafferentation of the overlying cortex (Schomer *et al.* 1993).

There was a concordance between the EEG and MR imaging findings for localization of the epileptogenic area. PLEDs were recorded involving the left frontocentroparietal areas during the twitchings of right side of the abdomen corresponding to the location of the cortical dysplasia at the mesial parietal region. The other reported cases in the literature with abdominal-truncal EPC had lesions in different cortical areas such as the frontal (Rosenbaum and Rowan 1990, Fernandez Torre *et al.* 2004, Dafotakis *et al.* 2006), central (Chalk *et al.* 1991, Gurer *et al.* 2001) and parietal regions (Matsuo 1984, Gurer *et al.* 2001). Although all of these patients had similar clinical features (with myoclonic twitchings of abdominal or truncal muscles, that may or may not have propagated to face and limb muscles), their lesions were not identical and did not primarily involve the somatotopic representation of these muscles. This variety of findings may be due to complexity of the homunculus. The classical organization of the homunculus, produced by surface stimulation (Penfield and Rasmussen 1950) has become more complex in recent years with the integration of movements generated by intracortical stimulation. A study in primates showed that representation of orofacial and eyelid muscles are also found in deep layers of cortical representation of the trunk (Dum and Strick 2002). Furthermore, it may be speculated that the typical functional organization of the motor cortex in man could be disrupted or altered during the development of cortical malformation (Andres *et al.* 2005). Also, widespread changes in functional reorganization of adjacent cortical tissue may occur due to direct effects of the hyperexcitable, dysplastic area (Nudo *et al.* 2001).

In conclusion, patients with abdominal seizures or myoclonus must be extensively evaluated and followed up with EEG and neuroimaging studies. Although the abdomen or trunk has a small representation area on the classical cortical homunculus, a malformation of cortical

**Table 1.** Clinical, EEG, neuroimaging findings of reported cases with EPC of abdomen or trunk.

	Age	Localization of EPC	Additional Clinical Features	Duration of EPC	EEG	Cranial CT	Cranial MRI	Pathology
Rosenbaum and Rowan	65 M	L abdomen (± face, limbs)	L hemiparesis for 1 week	1 day	Quasiperiodic delta and sharp wave in the C4-Cz	R parasagittal soft tissue mass destroying parietal bone depressing the brain, frontoparietal edema of white matter	-	R frontoparietal lesion, squamous cell carcinoma
Matsuo F.	27 F	R abdomen	Mild R hemiparesis	9 months	Slow waves over the left posterior quadrant	A mass lesion on L parietal area	-	Necrotic tissue with Aspergillus infection
Matsuo F.	66 F	L abdomen	Mild L hemiparesis	10 years	Low voltage slow waves on R hemisphere	R parietal meningioma	-	-
Matsuo F.	42 M	R abdomen, (± shoulder, face)	-	15 years	Low voltage slow waves over the left parietal area	Normal	-	-
Fernandez-Torre et al.	77 F	L > R trunk (± L hemibody)	Dysphasia, L hemiparesis	6 days	PLEDs on R central, mid-temporal, parietal	R frontal, temporal, cerebellar, L parasagittal frontal multiple lesions	-	Metastatic lung neoplasm ?
Chalk et al.	66 M	L abdomen (± L leg)	-	1 day	Epileptiform discharges on parasagittal area	Mild atrophy	Mild atrophy	Cryptococcal meningitis
Dafotakis et al.	62 M	L abdomen	-	36 hours	-	-	Hyperintensity around precentral gyrus. (disappeared 3 months later)	-
Gurer et al.	28 M	R pectoral m.	R hemiparesis	6 days	Slow wave discharges on left hemisphere	-	Left temporo-parietal cyst (Cyst hydatid)	-
Gurer et al.	43 M	L pectoral m.	Mild L arm weakness	< 24 hours	Normal	A metastatic lesion on the R central region	-	-
Tezer et al.	25 F	R abdomen (± face)	-	7 days	L Frontocentro-parietal PLEDs	-	L parietal cortical dysplasia	-

EPC: epilepsia partialis continua; M: male; F: female; CT: computurized tomography; MRI: magnetic resonance imaging; L: left; R: right; PLED: periodic lateralized epileptiform discharges.

development may be found at that specific location. The involvement of distant parts of cortical homunculus in this case may seem to contradict the classical cortical spreading of discharges through the homunculus, but reorganization of motor cortex due to a dysplastic area, subcortical enlargement of the lesion or propagation of discharges through to the deep layers instead of the surface, must be considered. □

### Legend for video sequence

Intermittent, mild contractions on the right side of the abdominal muscles. Occasionally, very slight facial twitchings of the right corner of the lips, and blinking were seen during the abdominal myoclonus, but there was no involvement of the limbs).

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