

# Pearls & Oysters: The chapeau de gendarme sign and other localizing gems in frontal lobe epilepsy



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## PEARLS

- Unlike temporal lobe epilepsy (TLE), the many semiologic features of frontal lobe epilepsy (FLE) are protean, and often epileptiform discharges or ictal patterns on EEG are not localized.
- We highlight 2 lesser-known signs associated with FLE:
  1. Ictal pouting, also known as the chapeau de gendarme sign, is an easily recognizable facial expression with its origins in the frontal lobe, seen early on in frontal lobe seizures.
  2. Interictal rhythmic midline theta (RMT) on scalp EEG occurs with a greater preponderance in FLE compared to TLE, and may be a useful localization clue if not seen during drowsiness or mental activation.

## OY-STERS

- Interictal FDG-PET imaging is often used in the identification of epileptogenic cortex. Subtle areas of hypometabolism on FDG-PET may be missed; FDG-PET/MRI coregistration can improve its sensitivity.

**CASE REPORT** A 28-year-old right-handed woman with a history of medically intractable localization-related epilepsy was admitted to the epilepsy monitoring unit (EMU) for continuous video-EEG, with the aim of characterizing her seizures and evaluating her surgical eligibility. Her seizures began when she was 18 months of age. There were no perinatal complications, and she attained normal cognitive and motor developmental milestones. Her history was negative for febrile seizures, significant head trauma, or CNS infections. No family history of a seizure disorder was present. She had failed several antiepileptic medications that achieved therapeutic dosing levels, including but not limited to carbamazepine, clonazepam, levetiracetam, and eslicarbazepine.

Her epilepsy was characterized by partial-onset seizures arising predominantly during sleep, and only rarely secondarily generalizing into tonic-clonic seizures. Her family described a downward turning of the mouth (“as if she was really sad and about to cry”), followed by tonic posturing of the right arm. When awake at the onset, she recounted an antecedent aura of a pressure-like sensation in her chest and inability to speak. The duration of her habitual seizures was less than a minute; they were usually followed by a short period of postictal agitation and tearfulness. They also had the tendency to cluster, and on average she had at least 3 seizures per night. Her neurology examination yielded normal findings.

She was monitored in the EMU for 11 days. During this period, more than 50 brief focal seizures were captured, averaging 10–30 seconds in duration. Both the semiology and EEG findings were stereotyped. She would rouse, open her eyes, and develop a sustained ictal pout (figure 1). Longer seizures were followed by right gaze deviation, and a tonic extension of first the right, then left arm. The seizures occurred predominantly out of stage 2 sleep, and had the propensity to occur in the early morning between 4:30 and 6:00 AM. One secondarily generalized tonic-clonic seizure was also recorded. She was not able to activate the push button or alert anyone during the seizures, despite remaining conscious. A video-EEG recording of one of her habitual seizures is shown in the video at [Neurology.org](http://Neurology.org).

The EEG background in wakefulness consisted of a low-voltage diffuse beta activity, with a poorly sustained 8 Hz posterior dominant rhythm. No interictal epileptiform abnormalities were seen, but there were intermittent rhythmic runs of 3.5–4 Hz central theta slow waves seen maximally at Cz, without accompanying clinical signs (figure e-1). The ictal onset was neither lateralizing nor localizing, and was characterized by widespread paroxysmal fast frequencies seen maximally over the parasagittal electrodes, sometimes preceded by a diffuse attenuation/desynchronization.

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The chapeau de gendarme or ictal pout sign is shown here during one of the patient's habitual seizures, characterized by a turned-down mouth.

During the same admission, the patient underwent 3T MRI of the brain, which revealed an area of cortical thickening deep in the left frontal operculum extending into the insula more posteriorly. This thickening was associated with blurring of the gray-white junction and T2 prolongation in subadjacent white matter that formed a transmantle sign (figure e-2). An interictal FDG-PET scan was acquired subsequently a few months later during presurgical planning. Initially read as negative, a further step in image analysis using FDG-PET/MRI coregistration later revealed a focal area of hypometabolism concordant with the lesional site on MRI (figure e-3).

The patient was eager to pursue surgery, and was willing to accept the risk of language deficits posed by the lesion's close proximity to the Broca area (from the anatomical standpoint). Her case was discussed at a multidisciplinary presurgical conference, and a surgical strategy was planned to include intracranial EEG (ICEEG) with both subdural and depth electrodes. ICEEG recorded several of her habitual seizures, confirming an intralesional and perilesional ictal onset (figure e-4). Near-total en bloc resection of the lesion was performed (part of the lesion deep in the insula was left behind due to accessibility limits), and tissue histopathology confirmed FCD type IIB. Due to seizure recurrence, she had further extension of the lesional resection along the inferior aspect of the remaining insula. Postresection MRI did not reveal any residual dysplasia. No neurologic deficits were seen following either of the surgeries. At her last (6 months) postoperative follow-up, she was free of her habitual seizures. Eslicarbazepine and clonazepam were discontinued, and she was maintained on a stable dose of carbamazepine.

**DISCUSSION** FCDs are intrinsically epileptogenic, and stereo-EEG studies have demonstrated both ictal discharges and intralesional rhythmic spike discharges to originate from dysplastic cortex.<sup>1</sup> Type II FCDs are more common in extratemporal regions, especially in the frontal lobe.<sup>2</sup> This case vignette highlights 2 uncommon but useful features seen in frontal lobe epilepsy, with corroborative anatomical localization from MRI and ICEEG.

The chapeau de gendarme or ictal pout sign is characterized by a turned-down mouth produced by bilateral lip and chin contraction. This term gives reference to the shape of the gendarme's hat seen during Napoléon I's time. If seen early, it confers a high localization value to the frontal lobe. In an analysis of ICEEG recordings belonging to 11 patients with this sign,<sup>3</sup> the epileptogenic zone was localized to the anterior cingulate (n = 4), orbito-frontal region (n = 2), mesial prefrontal/premotor cortex (n = 3), supplementary motor area (n = 1), and inferior frontal gyrus (n = 1). The presence of the ictal pout sign in our patient, together with the stereotyped, brief, and nocturnal nature of her seizures with preserved consciousness, were all highly suggestive of FLE. Although zones of frontal lobe networks underlying semiologic production exist,<sup>4</sup> it remains difficult to pinpoint sublobar anatomical origins of frontal lobe seizures based on semiology alone, due to extensive interregional connectivity within the frontal lobe and rapid seizure propagation.

The RMT on EEG, another feature present in our patient, was first described by Cigánek<sup>5</sup> in 1961 as an abnormal finding associated with epilepsy. Subsequently, RMT was also observed in normal participants

who were drowsy or engaged in cognitive tasks. More recently, a study conducted on 162 patients who underwent at least 3 days of continuous video-EEG monitoring demonstrated that interictal RMT in wakefulness was found significantly more frequently in FLE (26/54; 48.1%) compared to TLE (2/54; 3.7%) patients, and was not observed in healthy controls.<sup>6</sup> Therefore, when care is taken to exclude a drowsy state or mental activation, RMT serves as a useful EEG correlate of dysfunction, seen more commonly in FLE compared to TLE. It may be the only interictal abnormality seen in FLE patients, since many FLE patients lack any form of interictal epileptiform discharges.

MRI is the primary imaging modality used in epilepsy evaluation; however, there is an additional role performed by interictal FDG-PET. FDG-PET is particularly useful for detecting FCDs that are MRI-negative. MRI also often underestimates the extent of the FCD, and FDG-PET can help to identify the perilesional epileptogenic region, or it may reveal a seizure focus at some distance away from the lesion. This is especially pertinent if the scalp EEG findings are nonlateralizing or nonlocalizing, and these additional areas of hypometabolism should be investigated further with ICEEG to delineate the epileptogenic zone. FDG-PET/MRI coregistration has been shown to increase the sensitivity of FCD detection compared to visual analysis of FDG-PET alone.<sup>7</sup> It also gives more precise anatomical information due to better spatial resolution of the MRI. In an observational study,<sup>8</sup> the use of FDG-PET/MRI coregistration aided lesion localization in 48% of patients with type I FCD and 11% of patients with type II FCD with nonconcordant MRI and EEG findings. False-positives were rare (2%). Care should be taken to ensure that FDG-PET scans are performed in the interictal state, as seizures occurring during the FDG uptake period may result in false normalization of an interictal hypometabolic region.

This case is a reminder that complete resection of the FCD lesion remains the most important predictor overall of seizure freedom.<sup>9</sup> In type IIB FCDs, complete cortical resection of the lesion may lead to seizure freedom in as many as 92% of patients.<sup>10</sup>

## AUTHOR CONTRIBUTIONS

Dr. Tan: study concept, acquisition of data, data analysis and interpretation, drafting of manuscript. Dr. Muhlhofer: study concept, acquisition of data, data analysis and interpretation, drafting of manuscript. Dr. Knowlton: study supervision, revision of manuscript for intellectual content.

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