■ Clinical commentary

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Epileptic Disorders

Chapeau de gendarme in a toddler points to focal epilepsy originating from the subcentral gyrus

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ABSTRACT – The *chapeau de gendarme* sign or ictal pouting has been associated with focal epilepsy of frontal origin. The identification and characterization of this semiologic feature derive mainly from adult epilepsy surgery series, whereas paediatric cases have rarely been reported. Here, we present a 14-month-old girl with a *chapeau de gendarme* sign with eyes closed as the sole initial manifestation of left frontal lobe epilepsy. Brain MRI revealed an area suspicious for focal cortical dysplasia in the anterior aspect of the left subcentral gyrus, and ictal scalp EEG supported a seizure onset correlating with the lesion. This is the youngest reported patient with a *chapeau de gendarme* sign and the first to be associated with a lesion in the subcentral gyrus. Our observation extends the age range of patients presenting with this intriguing semiology and the range of localizations where it may originate. We propose considering the *chapeau de gendarme* sign as a hallmark of focal epilepsy in all age groups, including early life, and that presurgical evaluation should be timely initiated in patients with refractory seizures. [*Published with video sequence*].

Key words: *chapeau de gendarme* sign; focal cortical dysplasia; focal epilepsy; ictal pouting

VIDEO ONLINE

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The *chapeau de gendarme* sign or ictal pouting has been described as a characteristic ictal sign, defined by a turned-down mouth, also described as an inverted smile, with a symmetrical contraction of the lip and chin, resembling the shape of a gendarme's hat at the time of the French Revolution.

The neuronal network underlying the *chapeau de gendarme* has been localized to the frontal lobe, more specifically involving the anterior cingulate cortex [1, 2], the superior frontal gyrus [3] as well as the frontal operculum [4] and the adjacent insular regions [1, 5], whereas only single cases have been reported in association with a temporal lesion [6, 7]. The *chapeau de gendarme* sign has been attributed a high-localizing value in frontal lobe epilepsy, since it emerges at an early stage during the seizure, thus facilitating the delineation of its onset [1]. To date, focal cortical dysplasia (FCD) is the predominant substrate in reported cases, typically showing a poor response to anti-seizure drugs but responding to surgical treatment.

The identification and characterization of this semiologic feature derive mainly from adult epilepsy surgery series, whereas few paediatric cases have been reported so far, most of them

concerning adolescents (*table 1*). However, seizure semiology in focal epilepsy, including that arising in the frontal lobe, changes with brain maturation, leading to considerable diversity between children and adults [8]. Young children may present with seizure semiology that is age-specific rather than localization-specific [9]. In addition, healthy infants and young children may temporarily engage in repetitive behaviours, such as facial grimacing, as part of their normal movement repertoire.

Here, we present a 14-month-old girl with *chapeau de gendarme* sign with eyes closed as the sole initial manifestation of left frontal lobe epilepsy. Brain MRI revealed an area suspicious for FCD in the anterior aspect of the left subcentral gyrus, and scalp EEG supported seizure onset correlating with the lesion.

Case study

A previously healthy 15-month-old girl presented to the emergency department with brief paroxysmal and stereotypical episodes of pouting with eyes closed or open, lasting several seconds and occurring several times a day, without any other motor or behavioural changes. In the last three weeks, the facial grimacing had increased in frequency, whereas the motor and cognitive development of the child had remained unaffected. The girl reacted to her parents during these brief pouting episodes, she could hold and transfer objects, and continue walking when eyes were open. Exemplary recordings of some of these initial episodes are shown in the accompanying video. Her past medical history, including her perinatal history, was unremarkable, and she had attained age-appropriate cognitive and motor developmental milestones. Her family history was negative for seizure disorders. No neurological deficits were evident at clinical examination.

Over the following week, the episodes of ictal pouting occurred during both wakefulness and sleep, with eyes closed or open, and increased in severity, now including additional features such as behavioural arrest, downward gaze or searching gaze, blinking, tonic posturing of the right arm with a bent elbow and clenched fist, and left-hand dystonia or left manual automatisms (*figure 1A*). These additional features raised suspicion for epileptic seizures, and the girl had a routine video-EEG in wakefulness that showed an intermittent, sharply configured, theta slow discharge with sporadically superimposed spikes and sharp waves over the left fronto-centro-parietal region seen maximally at C3 (*figure 1C*).

Eventually, the girl was admitted to our video-EEG monitoring unit, where she had 12 stereotypic seizures within 24 hours, all in wakefulness. Each of these seizures featured *chapeau de gendarme* with eyes mainly closed as the very first and most prominent semiologic sign (*figure 1A*), lasting up to the end of the seizure in all cases and even persisting for up to 10-20 seconds after the seizure end on EEG in some cases. Seizure semiology, specifically ictal pouting, presented for 1-3 seconds following seizure onset on EEG and included asymmetric tonic posturing of the legs in the lying position (more pronounced on the right side), chin stiffening with a downward gaze, as well as mild body rocking movements in the sitting position. The girl was partly responsive during the seizures and fully responsive immediately after the end of the seizure. Seizures were occasionally followed by a brief period of postictal agitation and tearfulness. Ictal EEG recordings showed a rhythmic alpha activity in the left fronto-centro-parietal regions, seen maximally at C3, with propagation within 1-3 seconds to the contralateral homologous regions and evolution into a repetitive sharp wave pattern over bilateral fronto-centro-parietal regions, dominant over the left hemisphere (*figure 1D*). Seizures had a duration of 15-60 seconds and remitted spontaneously. The postictal EEG was characterized by a left fronto-centro-parietal delta slow discharge, lasting for 20-30 seconds, following the end of the ictal discharge. 3T MRI of the brain revealed an area of cortical thickening in the anterior aspect of the left subcentral gyrus with a blurring of the grey-white junction and a transmantle sign, suggesting FCD type II (*figure 1B*). The neuropsychological evaluation verified motor and cognitive development within normal range.

Despite initial treatment with levetiracetam at a daily dosage that was gradually increased to 54 mg/kg, the girl continued to have several seizures per day. Oxcarbazepin was introduced as add-on therapy and titrated to a daily dosage of 22 mg/kg, leading to seizure freedom that was corroborated by video-EEG monitoring. Since seizures are controlled with a well-tolerated anti-seizure drug regime, further presurgical evaluation (PET, SPECT, source imaging, *etc.*) and surgical treatment are currently not indicated. Thus, the FCD remains to be confirmed by histopathology in case of deterioration, with ultimate surgical resection.

Discussion

FCDs have been identified as the underlying substrate in over two thirds of children with pharmacoresistant focal, lesion-associated epilepsy undergoing presurgical evaluation and, eventually, epilepsy surgery in the first three years of life [10, 11]. These cortical malformations, particularly FCD type II, are intrinsically

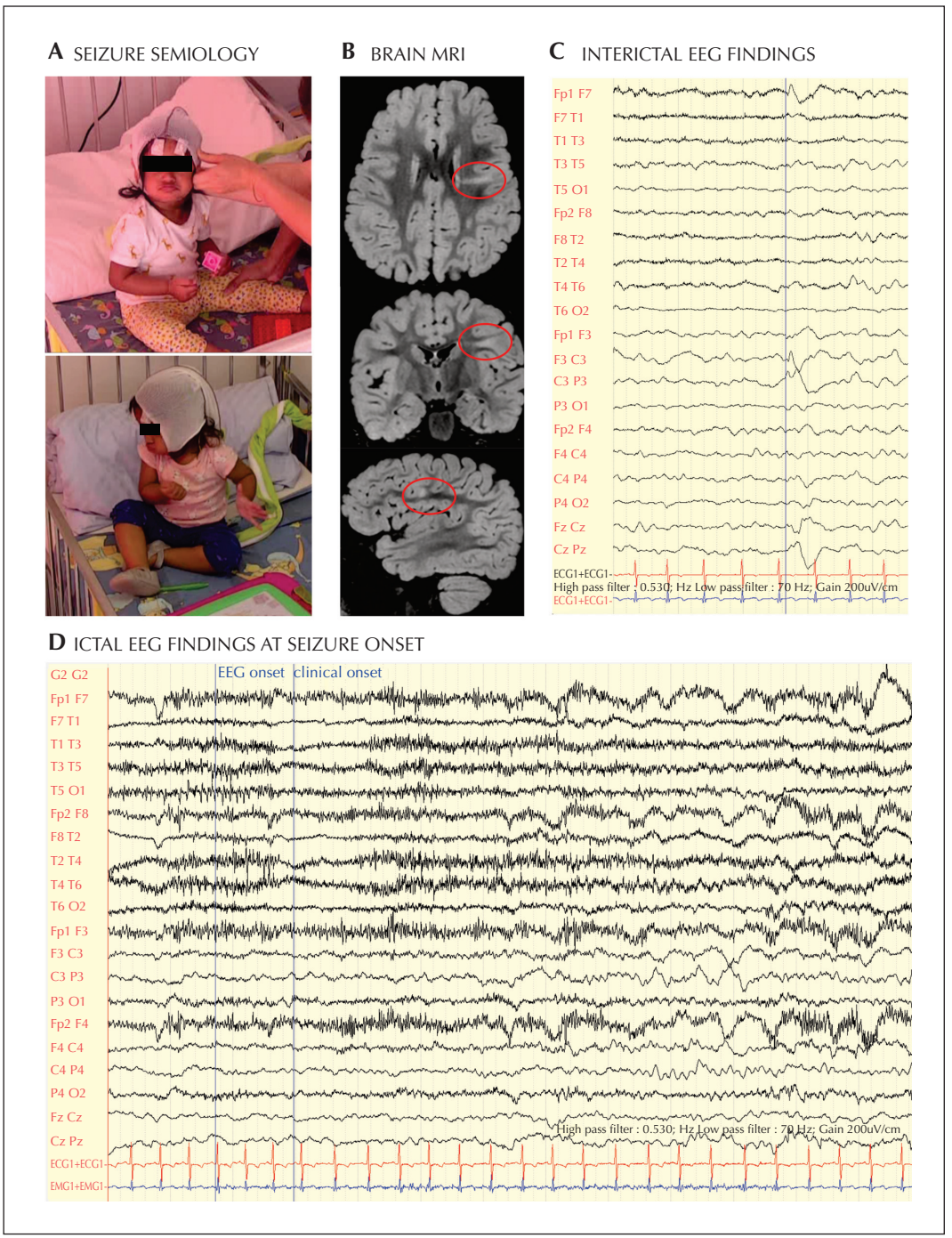
▼ **Table 1.** Reported cases of *chapeau de gendarme* first presenting in the pediatric age group.

Pt	Reference	Gender	Age at epilepsy onset (years)	Age at presurgical evaluation (years)	Ictal semiology	Presumed epileptogenic zone: based on scalp EEG, iEEG, SEEG	MRI findings	PET findings	Treatment	Histo-pathology	Seizure outcome at last follow-up visit
1	Souirti *et al.*, 2014 [1]	F	3	18	Fear, staring, CdG, vocalization, ipsilateral head deviation, contralateral tonic posturing, confusion, neurovegetative signs	SEEG: R orbito-frontal-ACC/ inferior frontal gyrus	MRI-negative	R orbitofrontal, ACC, temporal hypometabolism	Lesionectomy	FCD type 2	Seizure-free
2	Souirti *et al.*, 2014 [1]	M	12	16	Fear, staring, CdG, neurovegetative signs, vocalization, turning, waddling, pedaling, grasping, laughter, disinhibition	SEEG: R ACC / L ACC, R insula	MRI-negative	R ACC, insular hypometabolism	Lesionectomy	FCD type 2	Seizure-free
3	Souirti *et al.*, 2014 [1]	F	8	16	Sensation in the contralateral arm, neurovegetative signs, CdG, expression of disgust, ipsilateral head and eye deviation	n.a.	MRI-negative	L inferior frontal, insula, ACC hypometabolism	Lesionectomy	FCD type 2	Seizure-free
4	Souirti *et al.*, 2014 [1]	F	2	18	Psychic aura, grimacing, CdG, closed eyes, neurovegetative signs, contralateral head deviation, face and arm tonic, pedaling, disinhibition, confusion, laughter	SEEG: L mesial prefrontal - ACC	Susp. FCD L mesial prefrontal	L mesial prefrontal hypometabolism	Lesionectomy	FCD type 2	Seizure-free
5	Hayakawa *et al.*, 2018 [6]	M	15	16	CdG, no signs of fear, neurovegetative signs	Scalp EEG: R fronto-temporal:	Susp. glioblastoma R temporo-polar	Not performed	Anterior temporal lobectomy; radiation	Glioblastoma	Seizure-free
6	Yu *et al.*, 2018 [14]	M	6	15	Vegetative nervous sensations, CdG, automatism	iEEG: R operculo-insular	MRI-negative	Bilateral temporal hypometabolism	Lesionectomy	FCD	Seizure-free
7	Yu *et al.*, 2018 [14]	M	1	14	Viscera and vegetative nervous sensations, smiling, CdG, hypermotor	iEEG: R operculo-insular	MRI-negative	Not performed	Lesionectomy	FCD	Seizure-free

▼ **Table 1.** Reported cases of *chapeau de gendarme* first presenting in the pediatric age group (*continued*).

Pt	Reference	Gender	Age at epilepsy onset (years)	Age at presurgical evaluation (years)	Ictal semiology	Presumed epileptogenic zone: based on scalp EEG, iEEG, SEEG	MRI findings	PET findings	Treatment	Histo-pathology	Seizure outcome at last follow-up visit
8	Cebeci *et al.*, 2019 [7]	F	2	15	Loss of consciousness, CdG, ipsilateral head turning, contralateral hand dystonia, bilateral hand and mouth automatisms	iEEG: L fronto-temporal	Susp. L hippocampal sclerosis/ Susp. FCD L temporal lobe	L temporo-mesial/lateral hypometabolism	anti-seizure drugs	n.a.	n.a.
9	Cebeci *et al.*, 2019 [7]	F	14	16	Discomfort, nausea, loss of consciousness, CdG, eye blinking, contralateral hand dystonia, bilateral hand and mouth automatisms	Scalp EEG: L frontal	Susp. FCD L mesio-temporal to L insular	L mesio-temporal hypometabolism	anti-seizure drugs	n.a.	n.a.
10	Cebeci *et al.*, 2019 [7]	M	6	9	Forced expiration, loss of consciousness, CdG, closed eyes, hypermotor	Scalp EEG: Bilateral frontal/parasagittal, L predominance	MRI-negative	L inferior frontal hypometabolism	anti-seizure drugs	n.a.	n.a.
11	Wiwchar *et al.*, 2019 [5]	F	6	17	CdG, contralateral head and eye deviation, raising of the head, generalized stiffening, fear	SEEG: R anterior insular	Susp. FCD R fronto-opercular /	R frontal posterior and anterior insular hypometabolism	Lesionectomy	FCD type 2b	Seizure-free
12	Zhang *et al.*, 2019 [3]	M	5	16	Ictal pouting, R arm rising, moaning	SEEG: L superior frontal sulcus	Susp. FCD L superior frontal sulcus	L superior frontal hypometabolism	Lesionectomy	FCD type 2b	Seizure-free
13	Wang *et al.*, 2019 [15]	n.a.	3	4	Exaggerated and increased respiration, increased heart rate, focal tonic (neck, face), pouting, R arm tonic	SEEG: L insulo-opercular	MRI-negative	L insular hypometabolism	Lesionectomy	n.a.	Seizure-free
14	Rüsch *et al.*, 2020	F	1	1	CdG with eyes closed, contralateral clenched fist/ tonic posturing of the arm, blinking, tonic posturing of the contralateral> ipsilateral leg	Scalp EEG: L central	Susp. FCD L anterior gyrus subcentralis	Not performed	anti-seizure drugs	n.a.	Seizure -free

M: male; F: female; CdG: chapeau de gendarme; R: right; L: left; n.a: not available; susp.: suspected; ACC: anterior cingulate cortex; FCD: focal cortical dysplasia; SEEG: stereo-electroencephalography; iEEG: intracranial EEG.



■ **Figure 1.** (A) Seizure semiology. Upper panel: facial expression of our patient with ictal pouting and eyes open; lower panel: ictal pouting with closed eyes, clenching of the right fist, and dystonic posturing of the left hand. (B) MRI scan of the brain in (from top to bottom) axial, coronal, and sagittal sections showing the suspected dysplastic lesion in the anterior part of the left subcentral lobe. (C) Interictal EEG findings showing epileptic discharges in the left central region. (D) Ictal findings revealing initial ictal discharge starting in the left central region.

epileptogenic [12] and often localized in extratemporal regions, mainly in the frontal lobe [13]. This case report highlights a highly uncommon but particularly valuable semiologic feature that may be encountered in association with frontal lobe epilepsy of infancy and early childhood. This feature can serve to discriminate epileptic seizures from normal, though bizarre, behaviours and localize the onset of these epileptic seizures. Establishing an association between the *chapeau de gendarme* sign without any additional semiologic features and predominantly diurnal occurrence with frontal lobe epilepsy in our patient has indeed been particularly challenging. However, the decoding of this sign may prove particularly valuable for early epilepsy diagnosis and initiation of appropriate treatment, including epilepsy surgery in selected cases.

The *chapeau de gendarme* sign or ictal pouting has been described in predominantly adolescent and adult cohorts as a rare feature of seizure semiology with highly localizing value to the frontal lobe if seen at an early stage. In the so far largest study focusing on this bizarre semiology in 11 patients, nine of whom underwent stereoelectroencephalography (SEEG), the epileptogenic zone was localized to the medial frontal regions (anterior cingulate, medial prefrontal/premotor cortex) in eight cases, the fronto-basal (orbito-frontal) cortex in two, and the inferior frontal gyrus in one case [1]. A more recent case report has supported the relation of the *chapeau de gendarme* sign with an epileptogenic zone in the medial frontal regions [2], and three further cases have underlined the association of this sign with the anterior insula and the frontal operculum [4, 5, 14,15], whereas case series have included patients with ictal pouting arising from the superior frontal gyrus [3] (*table 1*). In our case, the presumed epileptogenic zone was localized in the anterior aspect of the subcentral gyrus (Rolandic operculum), in line with the SEEG-corroborated observation that the *chapeau de gendarme* sign is related to a network including the anterior cingulate cortex and the anterior part of the insula [1]. In accordance with a dorsolateral frontal seizure onset with spread to the insula (and the anterior cingulate cortex), our patient presented with tonic posturing and a facial expression resembling disgust. It should be noted, however, that a primary involvement of the insula in our patient cannot be entirely excluded, since invasive exploration or surgical resection have not yet been performed.

This is the youngest reported patient with a *chapeau de gendarme* sign as the initially exclusive and currently predominant manifestation of focal epilepsy and the first to be associated with a lesion in the subcentral gyrus. Our observation extends both the age range of patients presenting with this intriguing semiology and the range of localizations within the frontal lobe where this may originate. We propose considering the *chapeau de gendarme* sign as a hallmark of focal epilepsy in all age groups, including infancy and early childhood. Bearing this in mind, children with this striking semiology should be evaluated by video-EEG monitoring in order to capture and classify these episodes. Also, high-resolution MRI focusing on the frontal lobe should be performed to reveal potential dysplastic lesions. Most importantly, a comprehensive presurgical evaluation should be initiated in infants and young children with refractory seizures, and epilepsy surgery should be offered to appropriate candidates at the earliest possible time point to facilitate optimal outcomes regarding seizure control and cognitive development [16, 17]. ■

Supplementary data.
Summary didactic slides are available on the www.epilepticdisorders.com website.

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

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Legend for video sequence

Brief paroxysmal and stereotypical episodes of pouting with eyes closed or open at initial presentation, lasting several seconds and occurring several times a day, without any other motor or behavioural changes.

Key words for video research on www.epilepticdisorders.com

Phenomenology: face
Localization: frontal lobe (left)
Syndrome: focal non-idiopathic frontal (fle)
Aetiology: focal cortical dysplasia

TEST YOURSELF

(1) Which of the following statements are true? The *chapeau de gendarme* sign can be seen in:

A. temporal lobe epilepsy
B. generalized epilepsy
C. frontal lobe epilepsy
D. occipital lobe epilepsy

(2) Which of the following statements are true?

i) The *chapeau de gendarme* sign has the highest localizing value at seizure onset.
ii) Most patients with a *chapeau de gendarme* sign feature FCDs on brain MRI.

A. i) true, ii) false
B. i) false, ii) true
C. both true
D. both false

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".