

Discussion

Ictal and interictal ocular motor abnormalities in EAs, including DBN, have been mostly described in EA2/PxMD-CACNA1A and are believed to be rare or absent in EA1/PxMD-KCNA1.¹ Previously, only interictal gaze-evoked nystagmus has been described in a single EA1/PxMD-KCNA1 patient.² Here, we expand interictal ocular motor impairment in EA1/PxMD-KCNA1 by showing the presence of central positional DBN, suggesting involvement of cerebellar nodulus, uvula, and/or tonsil.³ The midline cerebellar atrophy found in our patients is consistent with this finding.^{1,3} Ictally, there was spontaneous DBN in two of our patients, further suggesting transient floccular/parafloccular dysfunction.³ Genetic assessment showed a novel disease-causing *KCNA1* variant. Most pathogenic variants in EA1/PxMD-KCNA1 are missense changes, which ultimately disturb Kv1.1 channel opening, leading to excessive GABA release into Purkinje cells and reduced cerebellar inhibitory output.⁴ In sum, interictal positional DBN, ictal spontaneous DBN, and cerebellar atrophy can be present in EA1/PxMD-KCNA1. The use of self-recording of eye movements, occlusive ophthalmoscopy, and positional testing with the help of video-oculography in the dark might further increase the detection of interictal and ictal forms of nystagmus in EAs. ●

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Authors Contributions

A.J. and D.M.: writing – original draft preparation (lead), investigation (equal), and methodology (equal); C.F. and O.G.: investigation (equal), methodology (equal), and resources (equal); J.O. and A.I.M.: investigation (equal), methodology (equal), supervision (equal), and writing – review & editing (supporting); J.L.: conceptualization (lead), investigation (equal), methodology (equal), supervision (lead), and writing – review & editing (lead).

Informed Consent and Ethics Committee Approval

Written informed consent was obtained from the patients for the anonymized information to be published in this article. Ethics committee approval was also obtained.

DATA AVAILABILITY STATEMENT

The data that supports the findings of this study are available in the supplementary material of this article.

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Supporting Data

Additional Supporting Information may be found in the online version of this article at the publisher's web-site.

Functional Movement Disorders During the COVID-19 Pandemic: Back to Charcot's Era at the Salpêtrière

The coronavirus disease 2019 (COVID-19) pandemic led the French government, like others throughout the world, to introduce measures such as lockdowns, bans on public events,

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TABLE 1 Patients characteristics and phenomenology of functional disorders

	Patients (n = 39) Clinical manifestations (%)
Age (years; mean; SD)	46 (15)
Female/Male (n)	25/14
Depression (%)	41.0
Reported past physical or psychological trauma (%)	41.0
Dystonic features (%)	43.6
- Paroxysmal hyperkinetic movements of four limbs [#]	25.6
- Fixed hand dystonic posture	18.0
Gait disorders [*]	41.0
Tremor [§]	10.3
Others (non-epileptic seizures)	5.1

[#]One patient with associated recurrent opisthotonos attacks;

^{*}Cautious gait, unsteadiness, forced slowness, slow pace with hesitations;

[§]Postural / action tremor with variability and distractibility. Various types of movement disorders were associated in the same patient.

and social distancing.¹ Healthcare structures were forced to reorganize their services to adjust to mutating needs.

When the number of patients with COVID-19 who required hospitalization exceeded the capacity of the health

system, hospitals reacted by reallocating resources for patients with COVID-19 and reducing the number of beds for other disorders. This resulted in restricted hospital admissions for patients without a life-threatening condition, such as those that can be found in a movement disorders unit.²

The various restrictive measures introduced to control the spread of COVID-19 have had a profound psychological impact on society in general, resulting in increased levels of stress, anxiety, and depression.³ Although patients with movement disorders tended to experience a gradual worsening during this period,² the consequences of lockdown measures on patients with functional movement disorders (FMDs) are largely unknown. Nevertheless, it is known that anxiety and depression can be strong triggers for the development or aggravation of FMDs. Furthermore, patients with FMDs often present sudden onset of symptoms with rapid progression, contrary to the slow, progressive course of most movement disorders.⁴ Consequently, given the general acuity of worsening or newly presenting symptoms, FMDs are a common presentation in emergency and acute medical settings.

We thus investigated whether referrals and hospital admissions of patients with FMDs have been more frequent during the COVID-19 pandemic, especially when COVID-19 hospital admissions peaked, and we studied the characteristics of FMD patient admissions in the context of the COVID-19 pandemic.

We retrospectively reviewed the medical records of patients with FMDs who were admitted in the Movement Disorders Unit (MD-Unit), Pitié-Salpêtrière University Hospital (Paris France) during the COVID-19 pandemic (from February 2020 to May 2021). The definition of FMDs was based on recent diagnostic criteria.⁴ We studied the kinetics of

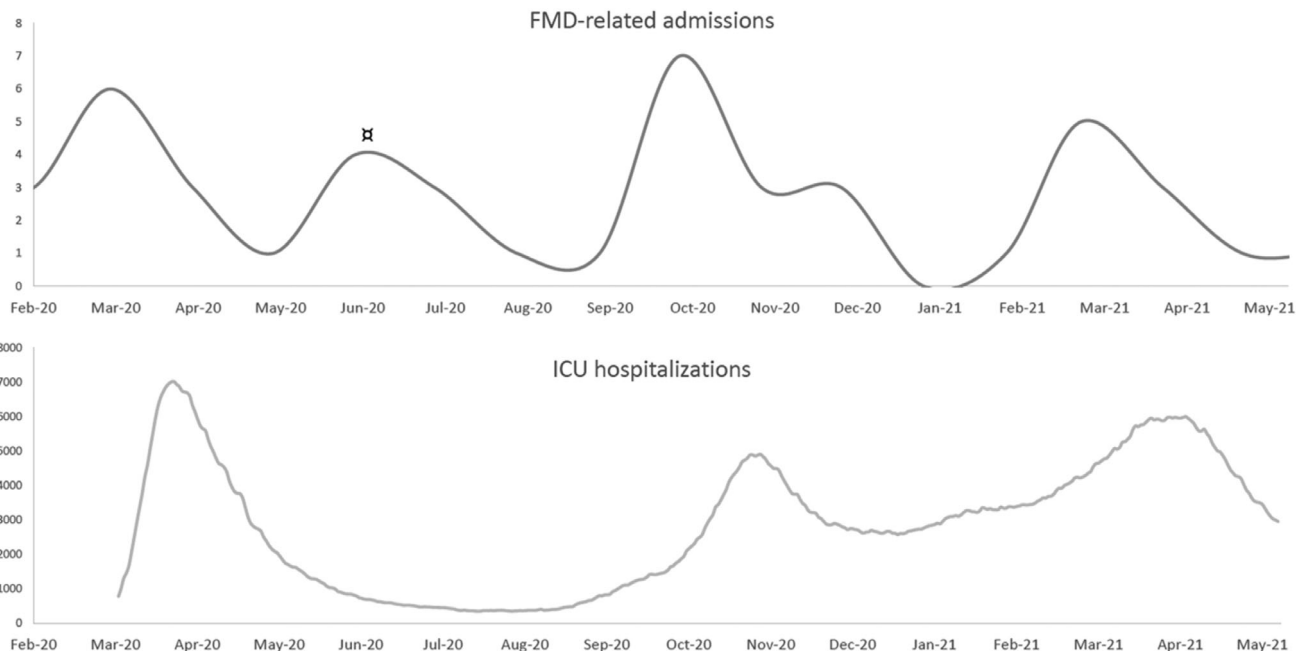


FIG. 1. Time course of patient admissions for functional movement disorders (FMDs) and intensive care unit (ICU) hospitalizations for coronavirus disease 2019 (COVID-19). Top graph: number of FMD patient admissions in our Movement Disorders Unit; Period when the unit was closed for reallocation of human resources to COVID-19 units, with delayed admissions of patients with FMD (□). Bottom graph: number of hospitalizations of patients with COVID-19 to ICUs in France (<https://www.data.gouv.fr/fr/datasets/r/f335f9ea-86e3-4ffa-9684-93c009d5e617>).

emergency referrals for FMDs within the context of the burden on intensive care units in France during the same time frame.


Thirty-nine patients presented with FMDs (mean age, 46 ± 15 years; 64.1% women; one with a history of COVID-19). Demographic and clinical characteristics are shown in Table 1. Patients had a combination of several types of movement disorder: opisthotonos attacks, intense hyperkinetic movements, and spasms of four limbs. A total of 53% of the patients had to stop work and required caregiver's assistance because of symptom severity. FMDs represented 9% of all patients hospitalized for movement disorder in the MD-Unit during this period versus the usual 4% observed before COVID-19. Waves of hospitalizations for FMDs followed the epidemic epochs and COVID-19 intensive care unit hospitalizations as shown in Fig. 1.

Our data showed an increase in patients presenting with FMDs who required urgent admission in the MD-Unit despite the COVID-19 pandemic context (Fig. 1). FMDs were severe in most patients (Table 1), and some presentations, such as opisthotonos crisis, were similar to those described in Charcot's era (19th century) illustrated by Richer's drawings and photographs from the "Nouvelle Iconographie de La Salpêtrière" (1904). These were referred to at the time as episodes of "major hysteria." Our results are consistent with other studies showing an increase in functional tics⁵ or nonepileptic seizures during the COVID-19 pandemic.⁶ In a time when bed availability was limited because the number of patients with COVID-19 who required hospitalization exceeded the capacity of the healthcare system, this increase in patients presenting with FMDs was an issue because 78.9% of them required urgent hospitalization.

This increase in FMDs could be seen as an additional "silent epidemic" alongside the COVID-19 pandemic. ●

Data Availability Statement

No data are available

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