The Canadian Journal of Neurological Sciences (2024), **51**, 333–335
doi:10.1017/cjn.2023.9

Canadian Journal of Neurological Sciences
Journal Canadien des Sciences Neurologiques

Letter to the Editor: New Observation

Typical Absence Status Epilepticus in Later Life Precipitated by Spirometry

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Keywords: Absence status epilepticus; Typical absence status epilepticus; Older people; Generalized nonconvulsive status epilepticus; Video-electroencephalography

Typical absence status epilepticus (ASE) is a rare cause of confusion in the elderly population requiring urgent video-electroencephalogram (VEEG) evaluation.[1] The diagnosis and classification may be a challenge. It most frequently represents reactivation of a previous genetic generalized epilepsy (GGE), although it may also represent a form of presentation in elderly people with epilepsy that begins in childhood or adolescence and that has not been diagnosed or treated.[1] Most rarely, it may be the presentation *de novo* of late-onset GGE.[1–3]

A 65-year-old man with chronic pulmonary obstructive disease undergoing evaluation for lung transplantation came to our hospital for pulmonary function tests. During the spirometry, the patient had a sudden episode of loss of consciousness with a fall to the ground and tonic contraction of the body and the four extremities without myoclonic jerks or other abnormal movements. The code for intrahospital cardiac arrest was activated. The intensivist in charge found a sinus tachycardia at 135 bpm. Subsequently, the patient regained consciousness but remained severely agitated and confused and was immediately transferred to the intensive care unit. An urgent VEEG showed recurrent rhythmic generalized polyspike-and-wave complexes (PSWC) and spike-and-wave complexes (SWC) at 3–4 Hz with preserved alpha activity over the posterior regions compatible with the diagnosis of typical ASE (Figure 1). Slowing of the background was not seen. During the recording, 5 mg of intravenous midazolam were administrated with normalization of mental status in the following hours. In addition, treatment with levetiracetam (1000 mg/12 h) was started. Computed tomography scan of the brain was normal. When the case was discussed in the EEG meeting, the need to perform serial EEGs was suggested to determine the patient's diagnosis of likely undiagnosed epilepsy. A second VEEG revealed generalized paroxysms of SWC at 3–4 Hz and a generalized photoparoxysmal response (generalized spike-wave activity limited to the stimulus train) at 30 Hz. Given the normal baseline cognition, photosensitivity, an EEG with normal background activity and typical paroxysms of PSWC and SWC on VEEG with an episode of typical ASE, we proposed the diagnosis of GGE. The patient confirmed having had "thought blocks" and attention deficit at the age of 9 years, but a diagnosis of epilepsy had never been made.

This case represents a clear example of typical ASE in the course of an undiagnosed benign GGE and helps to further delineate the spectrum of clinical presentation of typical ASE in older people. Although our patient had no prior diagnosis of GGE, a detailed clinical history and subsequent VEEG confirmed the existence of a benign form of GGE complicated by an episode of typical ASE. Recently, a single-center retrospective cohort study has provided a detailed electroclinical profile of patients with *de novo* extreme late-onset GGE in older adults and the elderly.[4] The authors also did a systematic review of the literature and found that late-onset GGE is rare and usually has a good prognosis. Of note, five patients (one in this cohort and four in the literature) had an initial presentation of typical ASE.[1,4]

Most frequently, in older people, the occurrence of typical ASE represents an aggravation or reactivation of a known pre-existing GGE.[1] Often, these patients have been seizure-free for years and may suffer from episodes of altered mental status secondary to typical ASE when they reach an older age. Later life could represent a period of vulnerability associated with predisposing factors including chronic ischemia, white matter disease, cerebral atrophy, metabolic derangements, hormonal changes, and polypharmacy in patients with GGE. In a previous study, four of six patients had small vessel brain disease and/or cortical–subcortical atrophy on neuroimaging studies.[1] Indeed, some authors have suggested that the increase in cerebrovascular lesion burden can alter cortical excitability and lower seizure threshold in patients with a genetic predisposition.[3]

In the case of an elderly patient with no history of epilepsy who has new onset of an ASE episode, the first option would be to consider *de novo* ASE of late onset (dnASLO).[5] This entity is considered a provoked or situation-related type of generalized

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Cite this article: Fernández-Torre JL, Paramio-Paz A, Campos-Fernández S, Rivera-Sánchez M, Orizaola P, and Hernández-Hernández MA. (2024) Typical Absence Status Epilepticus in Later Life Precipitated by Spirometry. *The Canadian Journal of Neurological Sciences* **51**: 333–335, https://doi.org/10.1017/cjn.2023.9

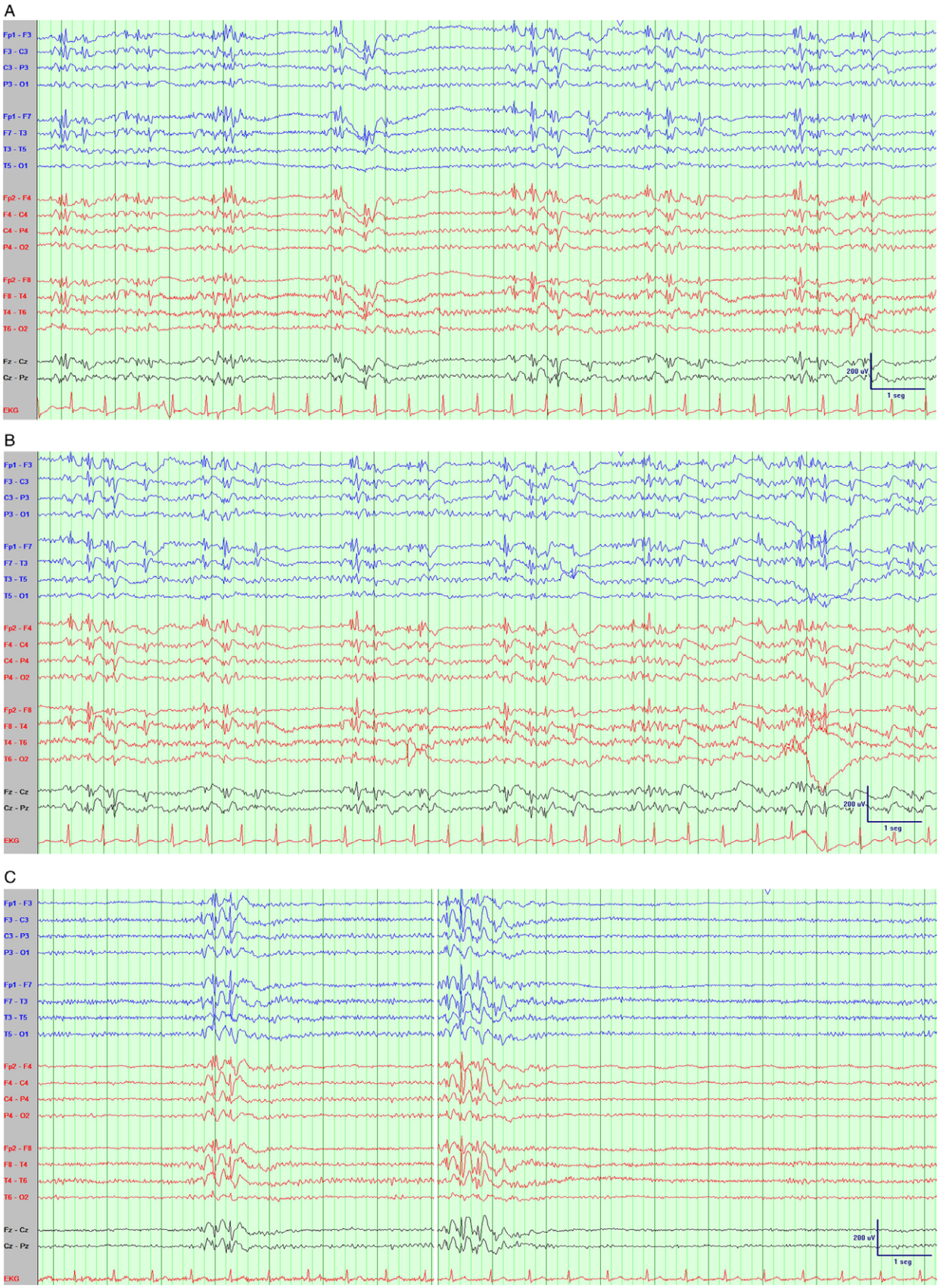

Figure 1: A) and B) First VEEG revealing continuous recurrent generalized paroxysms of polyspikes and spike-wave complexes at 3–4 Hz with preserved alpha activity over posterior regions in keeping with the diagnosis of typical absence status epilepticus. Low filter: 0.53 Hz; High filter: 70 Hz; Notch filter: 50 Hz. Speed: 30 mm/second. C) Second VEEG study showing brief generalized paroxysms of spike-wave complexes supporting the diagnosis of generalised genetic epilepsy. Low filter: 0.53 Hz; High filter: 70 Hz; Notch filter: 50 Hz. Speed: 30 mm/second.

https://doi.org/10.1017/cjn.2023.9 Published online by Cambridge University Press

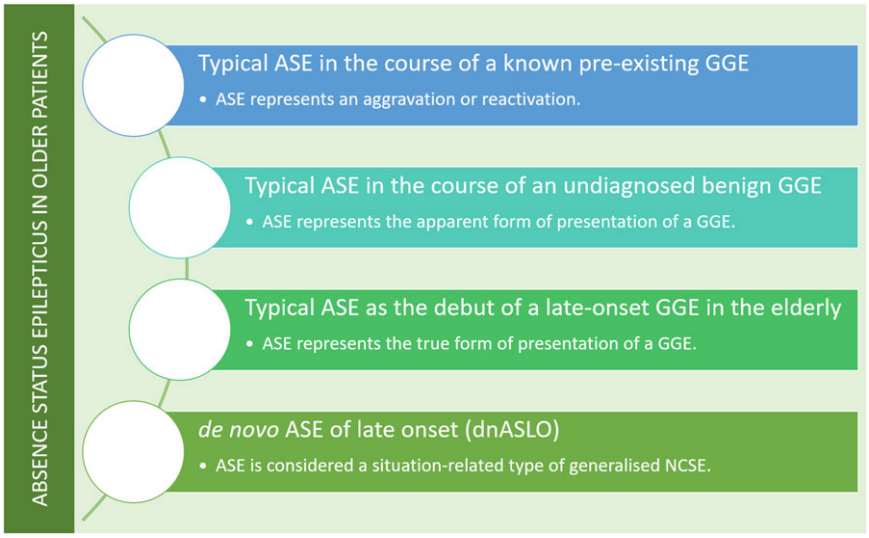

Figure 2: Types of ASE in mentally normal older people. ASE: absence status epilepticus; GGE: generalized genetic epilepsy; NCSE: non-convulsive status epilepticus.

non-convulsive status epilepticus. In these cases, clinical history should focus on possible exogenous triggers such as psychotropic medications, antidepressants, antiepileptic drugs, antibiotics, and antineoplastic agents, alcohol, and metabolic disturbances.[1] However, in our patient, the presence in the first VEEG of well-formed generalized symmetrical and synchronous PSWC and SWC, led us to suspect the existence of an underlying GGE. Typical ASE may be the presentation of late-onset GGE in the elderly[2,3] (Figure 2). In these patients, the episode can be categorized as true *de novo* typical ASE of late onset (*de novo* late-onset typical ASE). Although some authors use the term "*de novo*" only in subjects with dnASLO, when epilepsy starts in old age with an episode of typical ASE, the epilepsy is "new" and hence "*de novo*". To add "typical" would suggest that the patient has GGE. The terminology "idiopathic late-onset ASE" has also been used.[2]

The patient was seizure-free until old age. Although we cannot rule out the existence of subclinical seizures that had gone unnoticed for years, this seems unlikely. Unexpectedly, the spirometry was associated with a typical episode of ASE. We hypothesize that the respiratory effort during spirometry would have played a role similar to that of hyperventilation in triggering the ASE episode. Hyperventilation causes cerebral vasoconstriction secondary to hypocapnia and respiratory alkalosis, leading to a state of thalamo-cortical excitability. It is well-known that this activation procedure is considerably more effective in generalized epilepsy. Our patient had a sinus tachycardia. In a past prospective multicenter study, although hyperventilation was rarely associated with adverse events, one of the patients experienced symptomatic tachycardia.[6]

We emphasize that ASE occurring for the first time in elderly people is not necessarily secondary to external provoking factors but may rarely be the late presentation of an unknown or undiagnosed GGE in the elderly.

In summary, typical ASE may be the beginning and initial presentation of GGE in the elderly.[1] A high level of suspicion and consecutive VEEGs are needed to confirm the diagnosis.

Disclosures. Dr Fernández-Torre reports no disclosures.
Dr Paramio-Paz reports no disclosures.
Dr Sandra Campos-Fernández reports no disclosures.
Dr Pedro Orizaola reports no disclosures.
Dr María Rivera-Sánchez reports no disclosures.
Dr Hernández-Hernández reports no disclosures.

Statement of Authorship. JL Fernández-Torre was a lead author and wrote the paper and performed the literature review with substantive contributions of expertise and editing by MA Hernández-Hernández, Pedro Orizaola, and A Paramio-Paz. JL Fernández-Torre, A Paramio-Paz, Sandra Campos Fernández, and María Rivera-Sánchez identified the case in clinical grounds. JL Fernández-Torre obtained and edited the figures. All authors finalized the last version.

Ethical Position Statement. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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