

# Teaching NeuroImage: Claustrum Sign in Febrile Infection–Related Epilepsy Syndrome

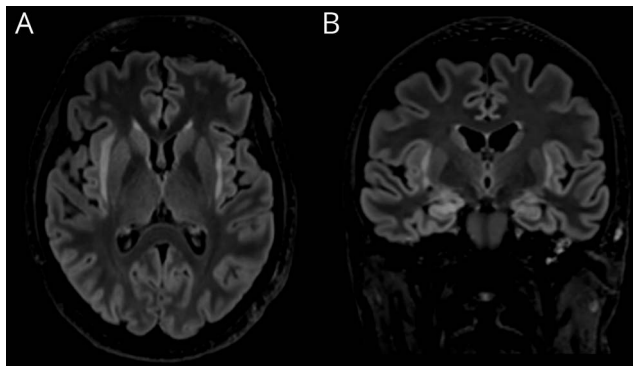
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**Figure 1** MRI (Fluid-attenuated inversion recovery; A, Axial; B, Coronal) Performed 30 days After the Onset of Status Epilepticus Showed Right-Predominant Hyperintensity and Swelling of the Claustrum, Pulvinar, and Hippocampus



A 40-year-old woman presented with acute encephalopathy and super-refractory status epilepticus 6 days after a febrile illness. An extensive diagnostic workup was negative. EEG and brain MRI showed right-predominant abnormalities, including claustrum T2/FLAIR hyperintensity, the so-called claustrum sign (Figures 1 and 2). This finding has been described in patients with

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**Figure 2** EEG Showed Bilateral Asymmetric Lateralized Periodic Discharges, Predominant in the Right Fronto-Temporal Region



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febrile infection–related epilepsy syndrome,<sup>1</sup> a subcategory of new-onset refractory status epilepticus triggered by cytokine storm.<sup>2</sup> Because the claustrum sign has been reported also in other cytokine storm-associated disorders, including acute necrotizing encephalopathy, COVID-19-related encephalopathy, and immune effector cell-associated neurotoxicity syndrome,<sup>3</sup> it may represent a specific marker of cytokine-mediated neuroinflammation.

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## Disclosure

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

## Appendix Authors

Name	Location	Contribution
<b>Lorenzo Muccioli, MD</b>	Department of Biomedical and Neuromotor Sciences, University of Bologna	Drafting/revision of the manuscript for content, including medical writing for content, major role in the acquisition of data, study concept or design, and analysis or interpretation of data

## Appendix (continued)

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## References

1. Meletti S, Slonkova J, Mareckova I, et al. Claustrum damage and refractory status epilepticus following febrile illness. *Neurology*. 2015;85(14):1224-1232.
2. Gaspard N, Hirsch LJ, Sculier C, et al. New-onset refractory status epilepticus (NORSE) and febrile infection–related epilepsy syndrome (FIRES): state of the art and perspectives. *Epilepsia*. 2018;59(4):745-752.
3. Pensato U, Muccioli L, Cani I, et al. Brain dysfunction in COVID-19 and CAR-T therapy: cytokine storm-associated encephalopathy. *Ann Clin Transl Neurol*. 2021; 8(4):968-979.

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