

Teaching NeuroImages: Neurovascular features of suspected antenatal-onset Sturge-Weber syndrome without skin involvement

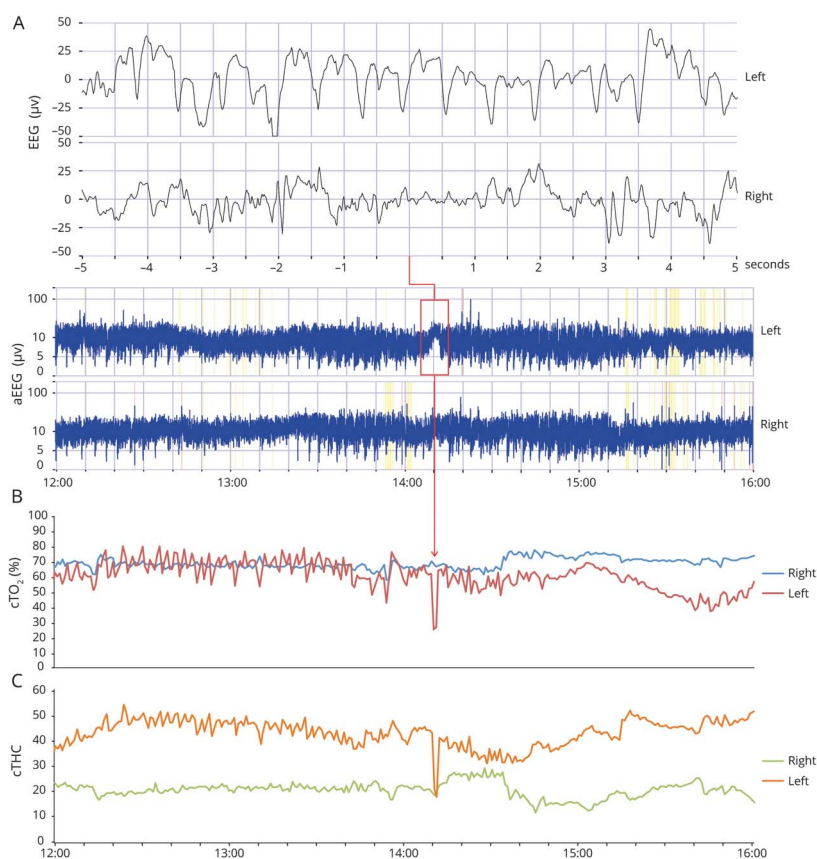
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Figure 1 Bilateral electroencephalography/amplitude-integrated electroencephalography and near-infrared spectroscopy patterns



Electroencephalography/amplitude-integrated electroencephalography (A), cerebral oxygenation (cTO₂, B), and total hemoglobin concentration (cTHC, C) patterns. The electrical seizure in the left hemisphere is accompanied by simultaneous ipsilateral acute changes in cTO₂ and cTHC in the left frontoparietal lobe (red arrow), detected using a NIRO-200NX oximeter (Hamamatsu, Japan).

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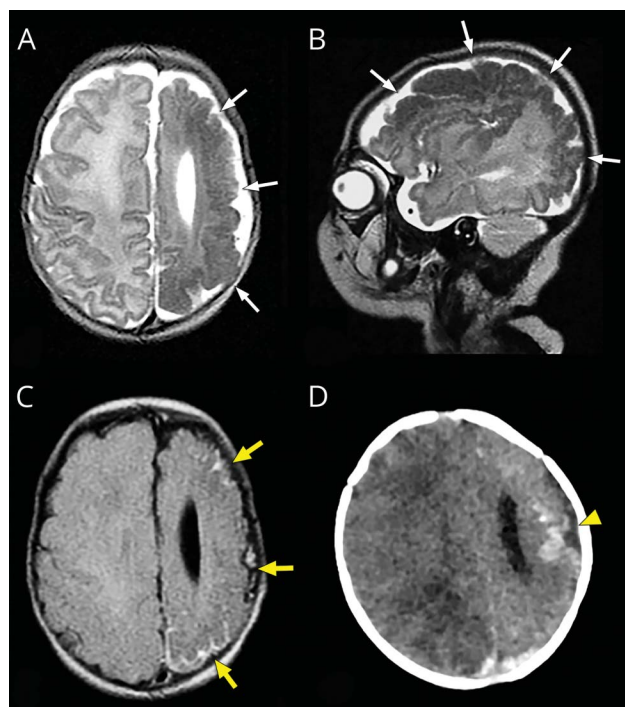
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A female infant was born at 35-week gestation by emergency C-section because of abnormal cardiotocography. No birthmarks were evident. Cord pH was 6.87; therefore, an amplitude-integrated electroencephalography monitoring was performed soon after birth, showing left-sided electrical seizures. Cranial ultrasound revealed left hemisphere atrophy, with subcortical

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Figure 2 Neuroradiologic findings



Left hemispheric atrophy with perisylvian polymicrogyria on T2-weighted axial (A) and sagittal (B) views (white arrows), ipsilateral frontoparietal leptomeningeal angiomas (postgadolinium axial FLAIR T2-weighted scan, yellow arrows) (C), and focal calcifications (axial CT scan, yellow arrowhead) (D). Left hemispheric T2 hypointensity (A and B) may be related to venous congestion or accelerated myelination.

hyperechoic areas in the frontoparietal lobe. Frontoparietal NIRS monitoring showed interhemispheric asymmetry of cerebral oxygenation and total hemoglobin concentration (figure 1) that serves as a proxy for cerebral blood volume.¹ Brain CT and MRI (figure 2) revealed characteristic features consistent with Sturge-Weber syndrome. Based on the evidence of polymicrogyria, the insult was dated back to the second trimester of pregnancy.² The ophthalmologic examination results were

unremarkable. The infant was started on phenobarbital,³ with seizure remission.

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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.

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Name	Location	Contribution
Silvia Martini, MD	S. Orsola-Malpighi University Hospital, Bologna, Italy	Acquired and analyzed NIRS and aEEG data and drafted the manuscript for intellectual content.
Francesco Toni, MD	IRCCS Istituto delle Scienze Neurologiche, Bologna, Italy	Acquired and analyzed neuroimaging data and revised the manuscript for intellectual content.
Vittoria Paoletti, PhD	S. Orsola-Malpighi University Hospital, Bologna, Italy	Contributed to NIRS data acquisition and revised the manuscript for intellectual content.
Luigi Corvaglia, MD	S. Orsola-Malpighi University Hospital, Bologna, Italy	Interpreted the data and revised the manuscript for intellectual content.
Duccio Maria Cordelli, MD	S. Orsola-Malpighi University Hospital, Bologna, Italy	Interpreted the data and revised the manuscript for intellectual content.

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