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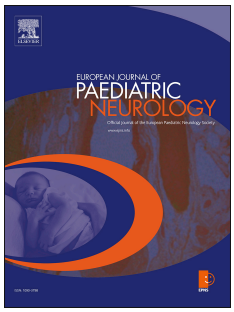

Transient regional cerebral hypoperfusion during a paroxysmal hemiplegic event in GLUT1 deficiency syndrome

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Abstract:
GLUT1 deficiency syndrome (GLUT1DS) is a well described neurometabolic disorder that results from impaired glucose transport into the central nervous system. GLUT1DS classically presents with infantile-onset epilepsy, progressive microcephaly, developmental delay, ataxia, dystonia, and spasticity, but a minority of patients may manifest with paroxysmal non-epileptic phenomena including hemiparesis (Wang et al., 2002). We report for the first time cerebral perfusion changes during an acute episode of hemiparesis in a 9 year old child with GLUT1DS. The patient presented as a code stroke with her second episode of acute-onset left hemiparesis and altered mental status. Emergency MRI of brain demonstrated normal diffusion-weighted imaging, but arterial spin label perfusion weighted imaging (ASL-PWI) showed regional hypoperfusion of the right cerebral hemisphere and magnetic resonance angiography (MRA) revealed distally restricted flow related enhancement in the right MCA. The patient's deficits resolved entirely within several hours from onset. Repeat MRI one month later was normal. Our