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Nonconvulsive status epilepticus in non-progressive encephalopathy: Presentation of two cases

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Nonconvulsive status epilepticus (NCSE) is described in children affected by non-progressive encephalopathy, mainly presenting as long-lasting episodes of alteration of contact, associated with myoclonic jerks or other movement disorder. Most children are affected by genetic syndromes (i.e. Angelman syndrome, Wolf Hirschhorn syndrome), developmental cortical malformation, or fetal/neonatal anoxic injury. Recognition of NCSE in such disabled children can be very difficult and often tardive. Treatment of NCSE is notoriously difficult, as it is usually resistant to antiepileptic drug. We present 2 children with non-progressive encephalopathy and epilepsy, who developed NCSE. First patient is a 5 year-old girl affected by Rett syndrome, due to MCP2 mutation and symptomatic generalized epilepsy. At age 4 year 6 months, she gradually developed a NCSE characterized by worsening of eye contact and deambulation in a period of several weeks. EEG showed continuous and diffuse slow spike-and-wave discharges. She was treated for her epilepsy with a combination of drug including carbamazepine (CBZ). Rapid CBZ withdrawn and levetiracetam introduction was followed by resolution of NCSE in a week. Second patient is 10 year-old girl affected by Down syndrome, left temporal desmoplastic infantile ganglioglioma, surgically treated, and Lennox-Gastaut syndrome. During a lung infection, she gradually developed a tonic status epilepticus characterized by recurrent, subtle axial tonic seizures for several hours, which were recognized with video-EEG recording. NCSE proved resistant to benzodiazepines and phenobarbital, but remitted with phenytoin iv.

Biography

Lucio Parmeggiani graduated in Medicine in Bologna in 1989. He was Resident in Neurology at Bellaria Hospital in Bologna, (1989-1993), and in Child Neurology at Stella Maris Hospital in Pisa (1997-2002). He completed his PhD at Bologna University in 1997. He was Researcher at Loyola University in Chicago (1991-1992), worked as a Neurology Consultant at King's College Hospital in London (1999-2001), and as Clinical Neurophysiologist at Great Ormond Street Hospital in London (2001-2003). Since 2006, he has been working as a Child Neurologist at Bozen Regional Hospital in Bolzano, Italy. He has published more than 30 papers in reputed journals.

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