

Opis choroby *

Definicja

A rare acute encephalopathy with inflammation-mediated status epilepticus characterized by infancy-onset of refractory unilateral, mainly clonic status epilepticus during or shortly after a febrile episode without evidence of central nervous system infection, followed by permanent or transient hemiplegia with a minimum duration of one week. The majority of children develop pharmaco-resistant epilepsy a few months later. Brain imaging shows edematous swelling of the affected hemisphere at the time of the initial status, followed by hemiatrophy that does not correlate with any vascular territory.

Dane

Klasyfikacja

Choroba

Synonimy

HHE syndrome

IHHS

Zespół HHE

Zespół padaczkowy z drgawkami połowicznymi i porażeniem połowicznym

Hemiconvulsion-hemiplegia-epilepsy syndrome

IHHS

Kod ORPHA

86908

Kod OMIM

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Kod ICD10

G40.4

Kod ICD11

8A62.Y

*Źródło

orphanet