

## 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

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#### \*Źródło

orphanet