

Zespół hiperinsulinizm-hiperammonemia

Kod Orpha: 35878 Kod OMIM: 606762

Opis choroby *

Definicja

A rare diffuse form of congenital hyperinsulinism characterized by an excessive/ uncontrolled insulin secretion (inappropriate for the level of glycemia), chronic hyperammonemia and recurrent episodes of hypoglycemia induced by fasting and protein rich meals. Epilepsy and cognitive deficit, which are unrelated to hypoglycemia but possibly related to the chronic hyperammonemia, may also occur. This disorder is usually responsive to diazoxide treatment.

Dane

Klasyfikacja

Choroba

Synonimy

HI/HA syndrome

Zespół HI/HA

Kod ORPHA

35878

Kod OMIM

606762

Kod ICD10

E72.8

Kod ICD11

5C50.AY

[*Źródło](#)

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.