

Opis choroby *

Definicja

A form of diffuse hyperinsulinism due to glucokinase hyperactivity and characterized by an excessive/ uncontrolled insulin secretion (inappropriate for the level of glycemia) and recurrent episodes of hypoglycemia induced by fasting and glucose rich meals. The clinical spectrum can range from mild and intermediate cases that respond well to dietary modifications and medical management with diazoxide to severe cases that are unresponsive to diazoxide. The potential development of type 2 diabetes with age is another notable feature.

Dane

Klasyfikacja

Choroba

Synonimy

Hyperinsulinemic hypoglycemia due to glucokinase deficiency
Hipoglikemia hiperinsulinemiczna z powodu niedoboru glukokinazy
Wrodzony hiperinsulinizm związany z glukokinazą

Kod ORPHA

79299

Kod OMIM

602485

Kod ICD10

E16.1

Kod ICD11

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[*Źródło](#)

orphanet