Congenital isolated hyperinsulinism

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Congenital isolated hyperinsulinism. ORPHA:657

Congenital isolated hyperinsulinism (CHI), a rare endocrine disease is the most frequent cause of severe and persistent hypoglycemia in the neonatal period and early infancy and is characterized by an excessive or uncontrolled insulin secretion (inappropriate for the level of glycemia) and recurrent episodes of profound hypoglycemia requiring rapid and intensive treatment to prevent neurological sequelae. CHI comprises 2 different forms: diazoxide-sensitive diffuse hyperinsulinism and diazoxide-resistant hyperinsulinism (see these terms).