

Opis choroby *

Definicja

A rare neoplastic disease characterized by occurrence of atypical and aggressive gastric type 1 neuroendocrine tumors (NET) in early adulthood. The tumors often show nodal infiltration requiring total gastrectomy. Synchronous gastric adenocarcinoma has also been reported. Patients present high serum gastrin concentrations and iron-deficiency anemia (rather than megaloblastic anemia, which is a typical feature in patients with sporadic gastric type 1 NET, where the tumor usually arises on the background of autoimmune atrophic gastritis).

Dane

Klasyfikacja

Choroba

Kod ORPHA

464756

Kod OMIM

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Kod ICD10

C16.9

Kod ICD11

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*Źródło

orphanet