Opis choroby *

Definicja

A form of multiple endocrine neoplasia type 2 (MEN2) syndrome characterized by medullary thyroid carcinoma in association with pheochromocytoma (one or both adrenal glands can be affected) and/or primary hyperparathyroidism (caused by parathyroid adenoma). Onset is typically later than in MEN2B, before 35 years of age. Diarrhea is the most frequent systemic symptom. Patients can develop Hirschsprung disease and, less frequently, cutaneous lichen amyloidosis or excessive production of adrenocorticotropic hormone.

Dane

Klasyfikacja
Podtyp kliniczny

Synonimy
MEN2A
MEN2A
Zespół PTC
Zespół Sipple'a
PTC syndrome
Sipple syndrome

Kod ORPHA
247698

Kod OMIM
171400

Kod ICD10
D44.8

Kod ICD11
2F7A.0

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*Źródło

orphanet