Opis choroby *

Definicja

A rare, genetic pituitary disease characterized by infantile-onset, rapid and excessive acceleration of linear growth and body size due to mixed growth hormone (GH)- and prolactin-secreting adenomas and/or pituitary hyperplasia. Patients present with gigantism and may have associated acromegalic features (e.g. coarse facial features, frontal bossing, prognathism, increased interdental space) as well as marked enlargement of hands and feet, soft tissue swelling, increased appetite and acanthosis nigricans.

Dane

Klasyfikacja
Choroba

Synonimy
Familial infantile gigantism
Dziedziczny gigantyzm dziecięcy
Dziedziczny przerost przysadki
Gigantyzm dziecięcy z powodu hiperplazji przysadki
X-LAG (Gigantyzm kończyn sprzężony z chromosomem X)
Hereditary infantile gigantism
Hereditary pituitary hyperplasia
Infantile gigantism due to pituitary hyperplasia
X-LAG

Kod ORPHA
300373

Kod OMIM
300942

Kod ICD10
E22.0

Kod ICD11
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*Źródło

orphanet