Opis choroby *

Definicja

A rare metabolic liver disease characterized by progressive liver disease and early cirrhosis due to accumulation of toxic cholesterol metabolites, which are detectable in bile, plasma, and urine, in association with dental abnormalities such as general hypomineralization and enamel hypoplasia, as well as occurrence of supernumerary teeth. There have been no further descriptions in the literature since 1996.

Dane

Klasyfikacja
Choroba

Synonimy
Lyngstadaas syndrome
Zespół Lyngstadaasa

Kod ORPHA
3196

Kod OMIM
-

Kod ICD10
K76.8

Kod ICD11
-

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*Źródło

orphanet