

# Niedobór dehydrogenazy steroidowej - wady zębów

## Kod Orpha: 3196 Kod OMIM:

### 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

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#### Kod ICD10

K76.8

#### Kod ICD11

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#### [\\*Źródło](#)

orphanet

### Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.