

# Wrodzony Niedobór siarczanu heparanu enterocytów

**Kod Orpha: 103910 Kod OMIM:**

## Opis choroby \*

### Definicja

A rare, severe, genetic, intestinal disease characterized by congenital absence of heparan sulfate from small intestine epithelium manifesting with secretory diarrhea and massive enteric protein loss. Patients present intolerance to enteral feeds during the first few weeks to months of life. Apart from absence of heparan sulfate from the basolateral surface of small intestine enterocytes, small bowel biopsy is otherwise normal.

Dane

### Klasyfikacja

Choroba

Kod ORPHA

103910

Kod OMIM

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Kod ICD10

P78.3

Kod ICD11

DA90.Y

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[\\*Źródło](#)

orphanet

## Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.