

## Opis choroby \*

### Definicja

Hereditary fructose intolerance (HFI) is an autosomal recessive disorder of fructose metabolism (see this term), resulting from a deficiency of hepatic fructose-1-phosphate aldolase activity and leading to gastrointestinal disorders and postprandial hypoglycemia following fructose ingestion. HFI is a benign condition when treated, but it is life-threatening and potentially fatal if left untreated.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Hereditary fructose-1-phosphate aldolase deficiency  
Dziedziczna fruktozemia  
Dziedziczny Niedobór aldolazy fruktozo-1-fosforanu  
Hereditary fructosemia

#### Kod ORPHA

469

#### Kod OMIM

229600

#### Kod ICD10

E74.1

#### Kod ICD11

5C51.50

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#### \*Źródło

orphanet