

# Zespół niedoboru Rh

Kod Orpha: 71275 Kod OMIM: 268150

## Opis choroby \*

### Definicja

A rare constitutional hemolytic anemia due to a red cell membrane anomaly characterized by lack or severe reduction of Rh blood group antigens, resulting in increased osmotic fragility of red blood cells and chronic hemolytic anemia of varying severity with stomatocytosis and spherocytosis. Two types of the syndrome arising from independent genetic mechanisms have been distinguished: the regulator type is caused by defects of the Rh associated glycoprotein (encoded by the *RHAG* gene), while the amorph type is due to mutations at the *RH* locus itself.

### Dane

#### Klasyfikacja

Choroba

#### Synonimy

Rh-null syndrome

Rh-null syndrome

#### Kod ORPHA

71275

#### Kod OMIM

268150

#### Kod ICD10

D58.8

#### Kod ICD11

3A10.Y

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#### [\\*Źródło](#)

orphanet

## Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.