

Opis choroby *

Definicja

A severe form of alpha-thalassemia that is mostly lethal, and associated with severe long-term outcome and lifelong transfusions in survivors. It is characterized by fetal onset of generalized edema, pleural and pericardial effusions, and severe hypochromic anemia.

Dane

Klasyfikacja	Synonimy
Podtyp kliniczny	Alpha-thalassemia hydrops fetalis Alfa-talassemia ciężka Alfa-talassemia homozygotyczna Alfa-talassemia obrzęk płodu Hemoglobin Barta obrzęk płodu Alpha-thalassemia major Hemoglobin Bart's hydrops fetalis Homozygous alpha0-thalassemia

Kod ORPHA	Kod OMIM	Kod ICD10
163596	236750	D56.0

Kod ICD11
3A50.03

*Źródło

orphanet