

Opis choroby *

Definicja

Heritable pulmonary arterial hypertension (HPAH) is a form of pulmonary arterial hypertension (PAH, see this term), occurring due to mutations in PAH predisposing genes or in a familial context. HPAH is characterized by elevated pulmonary arterial resistance leading to right heart failure. HPAH is progressive and potentially fatal.

Dane

Klasyfikacja

Podtyp etiologiczny

Synonimy

FPAH

FPAH

Hereditary pulmonary arterial hypertension

Rodzinne tętnicze nadciśnienie płucne

Familial pulmonary arterial hypertension

HPAH

Hereditary pulmonary arterial hypertension

Kod ORPHA

275777

Kod OMIM

178600

Kod ICD10

I27.0

Kod ICD11

BB01.0

*Źródło

orphanet