Opis choroby *

Definicja

A rare, congenital, non-syndromic heart malformation characterized by a single fibrous annulus with two orifices opening into the left ventricle. Clinical presentation is variable and related to the degree of resulting mitral insufficiency and/or stenosis, and depending on the associated heart disease, most commonly atrioventricular septal defect, obstructive left-sided lesions, and cyanotic heart disease. Rare cases of isolated disease have been reported.

Dane

Klasyfikacja
Podtyp kliniczny

Kod ORPHA
95474

Kod OMIM
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Kod ICD10
Q23.8

Kod ICD11
LA87.1Y

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*Źródło

orphanet