

Opis choroby *

Definicja

A rare congenital aortic malformation characterized by an aortic valve with four cusps instead of the usual three. The cusps can be equal-sized or vary in size. The malformation is an isolated finding in the majority of cases but may also be associated with other cardiac anomalies. The most common complication is aortic regurgitation. Aortic stenosis is infrequently observed. Patients usually become symptomatic in the fifth to sixth decade of life and may present with palpitations, chest pain, dyspnea, fatigue, pedal edema, and syncope. In severe cases, congestive heart failure can be the presenting symptom.

Dane

Klasyfikacja

Wada morfologiczna

Kod ORPHA

542568

Kod OMIM

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Kod ICD10

Q23.8

Kod ICD11

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*Źródło

orphanet