

Opis choroby *

Definicja

A rare congenital cardiac anomaly characterized by downward (apical) displacement of the functional annulus, due to incomplete delamination of the septal and inferior leaflets of the tricuspid valve such that they are hinged within the right ventricle, rather than as expected at the atrioventricular junction. The anterosuperior leaflet is often abnormal (redundancy, fenestrations, tethering with abnormal subvalvar apparatus). The atrioventricular junction and the "atrialized" portion of the right ventricle are dilated, with variable degrees of thinning of the right ventricular wall.

Dane

Klasyfikacja	Synonimy
Wada morfologiczna	Ebstein anomaly of the tricuspid valve Anomalia Ebsteina zastawki trójdzielnej

Kod ORPHA	Kod OMIM	Kod ICD10
1880	224700	Q22.5

Kod ICD11
LA87.03

*Źródło

orphanet