

Malformacja Ebsteina

Kod Orpha: 1880 Kod OMIM: 224700

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

Wada morfologiczna

Synonimy

Ebstein anomaly of the tricuspid valve
Anomalia Ebsteina zastawki trójdzielnej

Kod ORPHA

1880

Kod OMIM

224700

Kod ICD10

Q22.5

Kod ICD11

LA87.03

[*Źródło](#)

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.