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Authors: Agata Krawczyk-Ożóg, Bernadeta Chyrchel, Mateusz K Hołda, Jakub Batko,

Barbara Zdzierak, Andrzej Surdacki, Stanisław Bartuś

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Biannular atrioventricular valve disjunction as a potential cause of ventricular remodeling and subsequent cardiac arrest

Agata Krawczyk-Ożóg<sup>1, 2</sup>, Bernadeta Chyrchel<sup>2, 3</sup>, Mateusz K Hołda<sup>1, 4</sup>, Jakub Batko<sup>1</sup>, Barbara Zdzierak<sup>2</sup>, Andrzej Surdacki<sup>2, 3</sup>, Stanisław Bartuś<sup>2, 3</sup>

<sup>1</sup>HEART — Heart Embryology and Anatomy Research Team, Department of Anatomy,

Jagiellonian University Medical College, Kraków, Poland

<sup>2</sup>Clinical Department of Cardiology and Cardiovascular Interventions, University Hospital,

Kraków, Poland

<sup>3</sup>2<sup>nd</sup> Department of Cardiology, Jagiellonian University Medical College, Kraków, Poland

<sup>4</sup>Division of Cardiovascular Sciences, The University of Manchester, Manchester, United

Kingdom

### **Correspondence to:**

Agata Krawczyk-Ożóg, MD, PhD,

Department of Anatomy,

Jagiellonian University Medical College,

Kopernika 12, 31–034 Kraków, Poland,

phone: +48 12 422 95 11,

e-mail: krawczyk.ozog@gmail.com

Atrial mitral annular disjunction (MAD) is defined as a spatial displacement (≥2 mm) of the leaflet hinge line towards the left atrium within any part of the mitral mural leaflet or mitral commissures [1]. Similar disjunction may be observed for right atrioventricular valve - tricuspid annular disjunction (TAD) and may occur along whole tricuspid valve circumference [2].

We present the case of a 30-year-old, previously untreated female admitted to the Clinical Department of Cardiology and Cardiovascular Interventions, University Hospital Krakow, Poland after a prehospital cardiac arrest in the mechanism of ventricular fibrillation, to expand cardiac diagnostics. The patient has no known comorbidities and no significant cardiac diseases in family history. Transthoracic echocardiography showed normal left ventricular ejection fraction (60%), a small mitral valve prolapse (displacement of the mitral leaflets 2–3 mm into the left atrium during systole) with mild mitral regurgitation and atrial MAD located in posterior mitral leaflet (disjunction height of 6 mm) (Figure 1A;

Supplementary material, *Videos S1* and *S2*). Moreover, the suspicion of atrial-shifted TAD in mural leaflet has been raised with disjunction height of 4 mm (Figure 1B; Supplementary material, *Video S3*). Cardiac magnetic resonance confirmed well ejection fraction and the presence of MAD as well as TAD (Figure 1C–D). Moreover, late gadolinium enhancement (LGE) was present within the tricuspid, mitral annulus and subvalvular apparatus of both atrioventricular valves (Figure 1E). Linear subepicardial areas of LGE were found in the basal segments of the inferior and lateral wall of the left ventricle, additionally LGE was found in the free wall of the right ventricle. Contrast-enhanced electrocardiogram-gated computed tomography showed no changes in the coronary arteries and confirmed the presence of disjunctions (Figure 1F). In 24-hour electrocardiogram monitoring, numerous ventricular extrasystoles were found (15% of the recording) (Supplementary material, *Figure S1*). During the hospital stay, a cardioverter-defibrillator was implanted without complications and the dose of the beta-blocker gradually increased. After antiarrhythmic drug escalation, the amount of ventricular ectopic beats was reduced to 6% of recordings.

Annulus disjunction could involve both the tricuspid and the mitral annulus. In our patient MAD, TAD, mitral valve prolapse and myocardial fibrosis could be potential causes of ventricular arrhythmias and cardiac arrest. Previous research related to MAD focuses on the association between MAD and mitral valve prolapse, arrhythmias and sudden cardiac death [3, 4]. The prevalence of MAD varies depending on the patient population, imaging method and MAD's definition [1]. Recent studies have shown that in 50% of patients with MAD the TAD is also detectable [2]. The clinical significance of TAD is still unknown; however, we can find some case reports showing that presence of isolated TAD may be associated with ventricular arrhythmias [5]. Annular disjunctions could lead to excessive local contraction and stretching of cardiomyocytes, resulting in potential remodeling and fibrosis of the myocardium, and may predispose individuals to premature depolarization and trigger ventricular arrhythmias [4]. The presence of LGE in both the right and left ventricles of our patient suggests that the impact of both disjunctions on ventricular damage could be a possible cause of cardiac arrest.

### **Supplementary material**

Supplementary material is available at https://journals.viamedica.pl/polish\_heart\_journal.

#### **Article information**

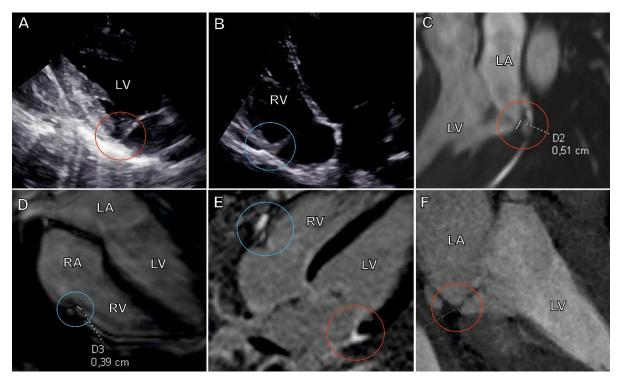
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**Figure 1.** Biannular atrioventricular valve disjunction. Transthoracic echocardiography images showing the mitral annular disjunction ( $\mathbf{A}$ ) and tricuspid annular disjunction ( $\mathbf{B}$ ). Cardiac magnetic resonance examination with visible mitral annulus disjunction ( $\mathbf{C}$ ) and tricuspid annulus disjunction ( $\mathbf{D}$ ). Cardiac magnetic resonance examination with late gadolinium enhancement visible within the tricuspid and mitral annuluses ( $\mathbf{E}$ ). Contrast computed tomography with visible mitral annulus disjunction ( $\mathbf{F}$ )

Abbreviations: LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle