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CASE REPORT

# Atrioventricular septal defect in an adult patient: There are 'clefts' and clefts☆

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**KEYWORDS**
Mitral cleft;
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**Abstract** In this report, we present the case of an adult male with severe mitral regurgitation due to an atrioventricular septal defect. Anatomical assessment by two- and three-dimensional transesophageal echocardiography was essential for detailed morphological characterization and surgical planning. The different features of a 'cleft' in an atrioventricular septal defect compared to an anterior leaflet cleft in an otherwise normal mitral valve are here discussed.


**PALAVRAS-CHAVE**
Fenda mitral;
Ecocardiografia tridimensional;
Defeito do septo auriculoventricular

**Defeito do septo auriculoventricular num doente adulto. Há «fendas» e fendas**

**Resumo** Os autores apresentam o caso de um doente adulto com regurgitação mitral severa no contexto de um defeito do septo auriculoventricular. A avaliação conjunta por ETE 2 D e 3 D foi fundamental para a caracterização anátomo-funcional da válvula e orientação do procedimento cirúrgico. As particularidades anatómicas de uma «fenda» em contexto de defeito do septo auriculoventricular e de uma fenda numa válvula mitral são aqui discutidas.


## Introduction

Atrioventricular septal defect (AVSD) covers a range of congenital cardiac malformations that have in common a single atrioventricular (AV) junction. This results in significant morphological alterations, partial AVSD being characterized by a common AV junction but two separate AV valves due to the

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fusion of the superior and inferior leaflets (bridging leaflets), which gives the left AV valve a trifoliate configuration. This is different from mitral valve clefts not associated with endocardial cushion defects.[1]

## Case report

A 52-year-old man was referred for valve surgery due to severe mitral regurgitation secondary to a mitral cleft and moderate aortic regurgitation through a bicuspid aortic valve. Two-dimensional transesophageal echocardiography (2D TEE) showed a single AV junction with a trifoliate left valve: one mural, one anterosuperior and one posteroinferior, with a 'cleft' oriented towards the interventricular septum in the zone of apposition (Figure 1A and B). The papillary muscles were displaced laterally, next to the mural leaflet commissures with bridging leaflets. Three-dimensional (3D) TEE provided detailed anatomical information (Figure 1C and D). No evidence of interatrial or interventricular communication, usually characteristic of AVSD, was observed.

Both valves were reconstructed. The mitral valve was repaired by direct suturing of the 'cleft' using separate 6-0 polypropylene sutures, enlarging the posterior leaflet with an autologous pericardial patch, and annuloplasty using a 28 mm semi-rigid Physio ring (Carpentier-Edwards, Edwards Lifesciences). Aortic regurgitation was corrected by plicating the noncoronary leaflet and reducing the annulus by annuloplasty of the aortoventricular junction. Postoperative TEE showed minimal regurgitation and a mean left atrial/left ventricular gradient of 5 mmHg.

## Discussion

Unlike the normal heart that has separate left and right AV junctions, the distinguishing feature of AVSD is a common AV junction – there is no AV septum and the AV annuli are in the same anatomical plane,[2] and there is no morphologically normal AV valve. In total AVSD there is a single common AV valve consisting of five leaflets, while in partial AVSD the valve is usually trifoliate, arising from the fusion of the superior and inferior leaflets and consisting of the anterosuperior, posteroinferior and mural leaflets, with three commissures designated anterolateral, posterolateral and septal according to their respective positions. This fusion results in a 'cleft' that causes varying degrees of valvular regurgitation, although valve stenosis is rare. Chordal abnormalities are

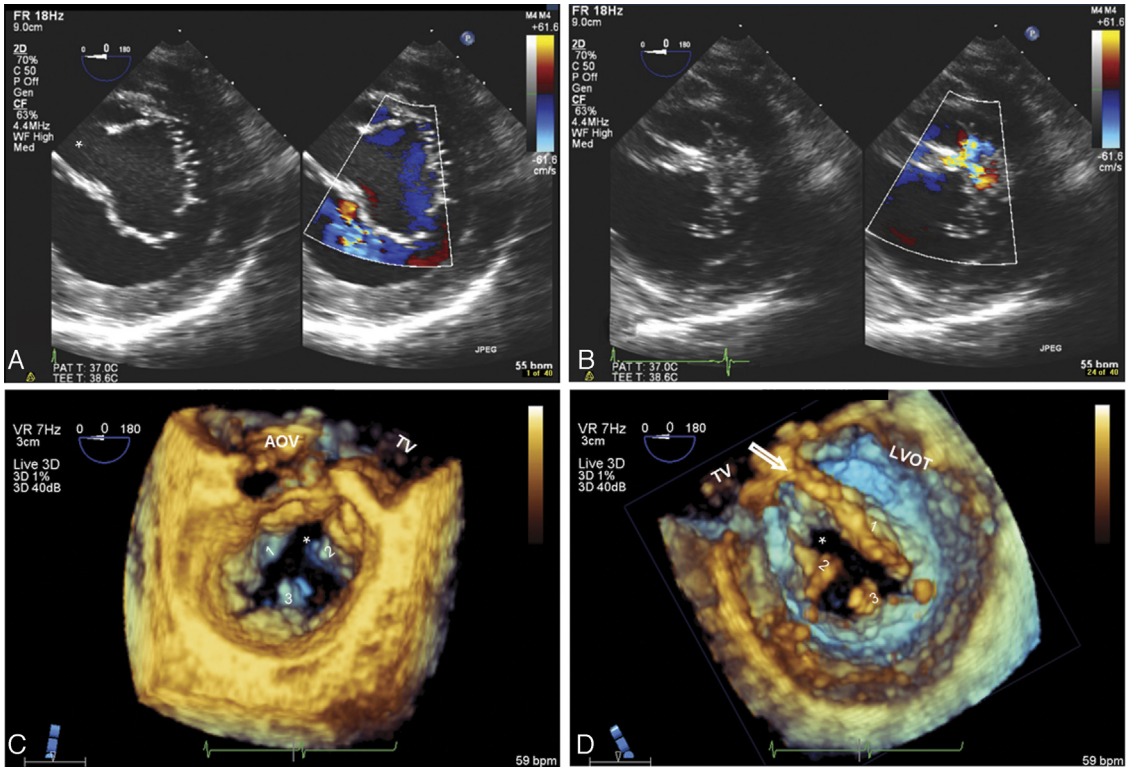


**Figure 1** (A and B) Two-dimensional transesophageal echocardiography, transgastric view, showing trifoliate configuration of the left AV valve and the 'cleft' (*) in diastole (A); mitral regurgitation from the 'cleft' in systole (B); (C and D) real-time three-dimensional zoom, cropped images: in diastole, atrial view (C) and ventricular view (D), the morphology of the three leaflets of the left AV valve can be seen in detail, as well as the septal orientation (towards the right ventricle) of the 'cleft'. Abnormal chordal attachments linking the edges of the 'cleft' to the interventricular septum can be clearly seen in ventricular view (arrow). Other features of a left atrioventricular valve can be seen – elliptical annulus and small mural leaflet. 1: anterosuperior leaflet; 2: posteroinferior leaflet; 3: mural leaflet; AOV: aortic valve; LVOT: left ventricular outflow tract; TV: tricuspid valve.