

Cleft mitral valve or levo-transposition of great vessels?

Clinical Case Portal

Date of publication:

13 May 2009

Topics: Non-invasive imaging: Echocardiography, MR/CT, Nuclear
Congenital Heart Disease
Echocardiography (Non-invasive imaging)

Authors:

Purvis, John. MD, FESC

Smyth, Stephen. ASCT

Fitzpatrick, Sinead. ASCT, BSc

Authors details:

Cardiac Unit, Altnagelvin Hospital.
Western HSC Trust
Londonderry BT47 6SB
UK

Contact:

e-mail: john.purvis@btinternet.com

Abstract

A 39-year-old man was referred for echocardiography because of dyspnoea, chest pain and murmur. Main finding was a globular morphology of left ventricle with apical hypertrophy and a seemingly cleft mitral valve with mild mitral regurgitation. Trans-oesophageal echocardiography confirmed the

ventricular hypertrophy and regurgitation but showed the aortic valve to lie anteriorly and to the left of the pulmonary valve in short axis. This finding is typical of levo-transposition of the great cardiac vessels. The "cleft mitral valve" was in fact a morphological tricuspid valve in a systemic position.

Introduction

A 39-year old man attended his primary physician complaining of chest pain and dyspnoea on exertion. The primary physician heard a pan-systolic murmur over the lower left sternal edge and requested an echocardiogram as the initial investigation.

Case Report

Only limited acquisition windows were available on echocardiography. The main findings were an unusual globular morphology of "left" ventricle with very marked apical hypertrophy and a seemingly cleft "mitral" valve (total of 3 valve leaflets identified) with mild "mitral" regurgitation. The "tricuspid" annulus was noted to be higher than the mitral and papillary muscles were not seen (Figs 1 + 2). It proved impossible to adequately image the aortic valve.

In view of this, trans-oesophageal echocardiography was performed. This confirmed the ventricular hypertrophy and mild AV valve regurgitation (Fig 3) but showed the aortic valve to lie anteriorly and to the left of the pulmonary valve in short axis (Fig 4). This finding is typical of levo-transposition of the great cardiac vessels, a rare acyanotic congenital heart disease in which there is both atrio-ventricular and ventriculo-arterial discordant connection. This means that the left atrium is connected to a morphological right ventricle (systemic ventricle; SV) and this in turn connects to the aorta. The right atrium is connected to a morphological left ventricle (pulmonic ventricle; PV) which in turn connects to the pulmonary artery.

Our "cleft mitral valve" was in fact a morphological tricuspid valve and the appearance of the "left" ventricle was due to hypertrophy of the morphological right ventricle in the systemic position. This also explains the reversed annular and papillary muscle findings.

To investigate his complaint of chest pain, he underwent exercise stress test but didn't achieve satisfactory heart rate. He therefore underwent 64-slice multi-detector coronary CT angiogram which showed no atherosclerosis but confirmed an anterior and leftward position of the aorta with the right coronary artery running along the anterior interventricular groove where the LAD is usually located. The LAD itself was displaced towards the lateral aspect of the heart (Fig 5). The CT also demonstrated the severe hypertrophy and trabeculation of the systemic ventricle (Fig 6).

Discussion

This condition is often clinically silent in childhood and becomes manifest in mid adult life as the hypertrophied morphological right ventricle starts to fail. Prognosis depends on the degree of any systemic atrio-ventricular valve regurgitation as well as systemic ventricular function. Conduction abnormalities are frequent, possibly due to abnormal arterial supply. ⁽¹⁾ Approximately 80% of cases have a ventricular septal defect and some have pulmonary outflow tract compression as the pulmonary valve is wedged between the AV valves. ⁽²⁾ These findings were not present in our case.

Conclusion

Our patient received ACE inhibitor therapy to offload his systemic ventricle and is currently doing well. It is worth considering that a left-sided AV valve with 3 cusps could be a levo-transposed tricuspid valve and not a cleft mitral valve.

References

1. Graham TP, Bernard YD, Mellen BG, Celermayer D, Baumgartner H, Cetta F et al. Long term outcome in congenitally corrected transposition of the great arteries. A multi-institutional study. JACC 2000;**36**; 255-61.
 2. Chung JH, Gunn ML, Godwin JD, Takasugi J and Kanne JP Congenital thoracic cardiovascular anomalies presenting in adulthood: a pictorial review. JCCT 2009;**3**; Suppl 1, S35-S46
-

Fig. 1 :
[TTE Apical 4 chamber](#)

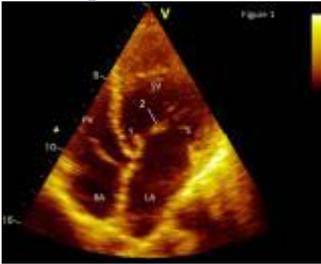


Fig. 2 :
[TTE parasternal long axis](#)



Fig. 3 :
[TOE systemic ventricle and AV regurgitation](#)

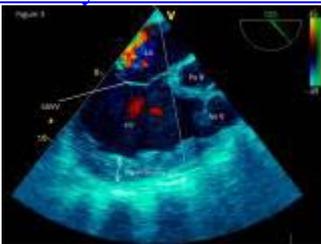


Fig. 4 :
[TOE short axis](#)

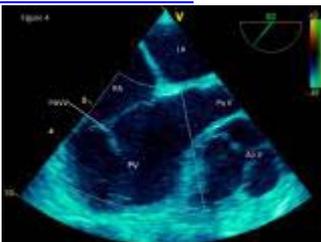


Fig. 5 :

3D volume rendered CT image of coronary anatomy

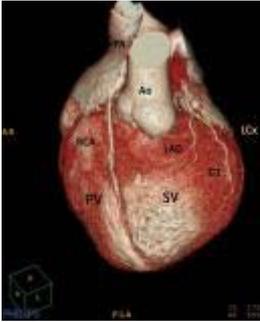


Fig. 6 :
Axial CT image of systemic ventricle

