QUADRICUSPID AORTIC VALVE ASSOCIATED WITH AGENESIS OF CIRCUMFLEX CORONARY: A RARE CONGENITAL CARDIAC ENTITY TO SEE AT LEAST ONCE IN LIFE

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Introduction

Quadricuspid aortic valve is a rare malformation. Its incidence is estimated of 0.003 to 0.043% of all congenital heart disease (1, 2). It may be associated with other malformations (the most common being coronary artery anomalies found in around 10% of cases (3, 4)).

The case

We report the case of a 52 years old male patient presenting with new onset heart failure symptoms in the context of a dilated cardiomyopathy, in which transthoracic echocardiography (TTE) revealed a quadricuspid aortic valve (QAV), with no signs of stenosis and only mild to moderate centrovalvular regurgitation.
2D TTE raise the suspicion of a QAV and 2D and 3D transoesophageal echocardiography confirmed the presence of 4 equal-sized aortic cusps (***figure 1***). There was no hemodynamically significant impairment of leaflet opening (***figure 2***).

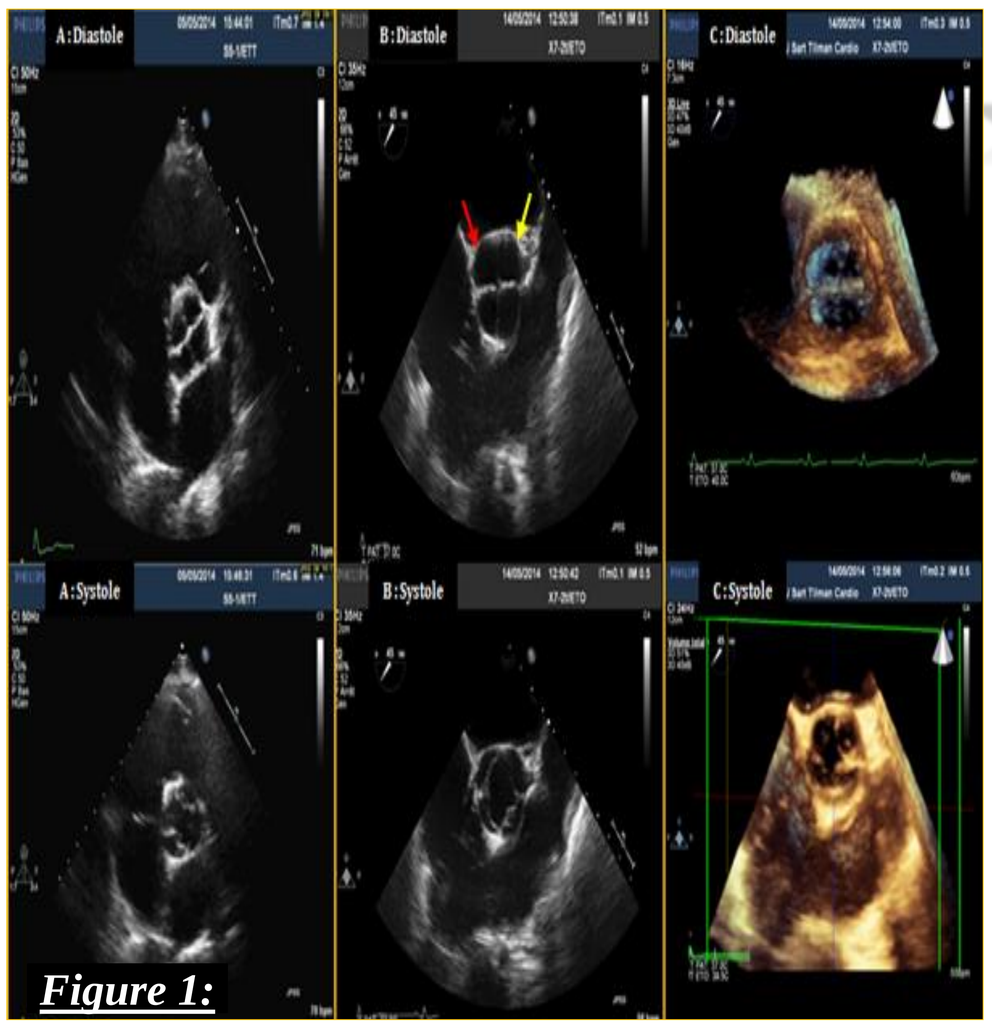

Figure 1:

A and B: 2D Short axis transthoracic and transoesophageal echocardiographic views (4 equal-sized aortic cusps opened and closed); C: 3D transoesophageal echocardiography (4 equal-sized aortic cusps opened and closed).
: Posterolateral left cusp.
: Anterolateral left cusp.

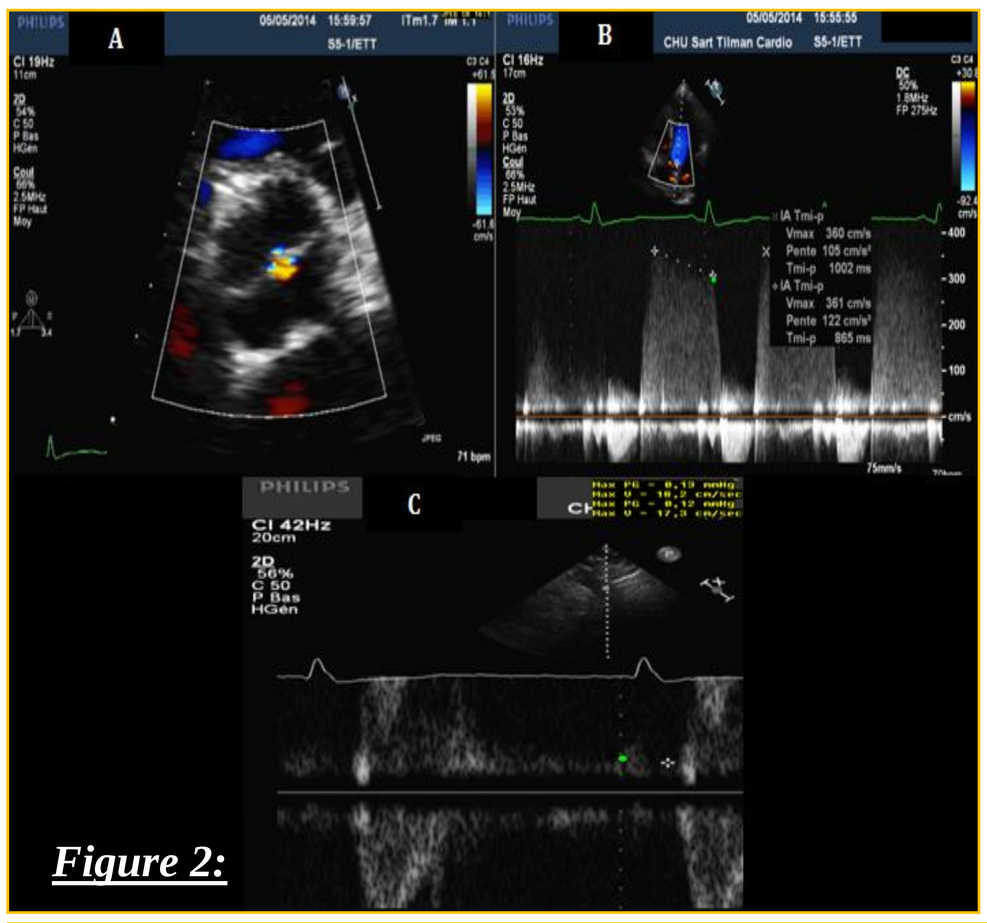

Figure 2:

A: Transthoracic short axis view: color Doppler on the centrovalvular aortic regurgitation.
B: Pressure half-time at 933 ms on the Continuous wave Doppler.
C: Pulsed Doppler within the descending aorta demonstrates mild aortic regurgitation: end-diastolic flow velocity at 16 cm/s.

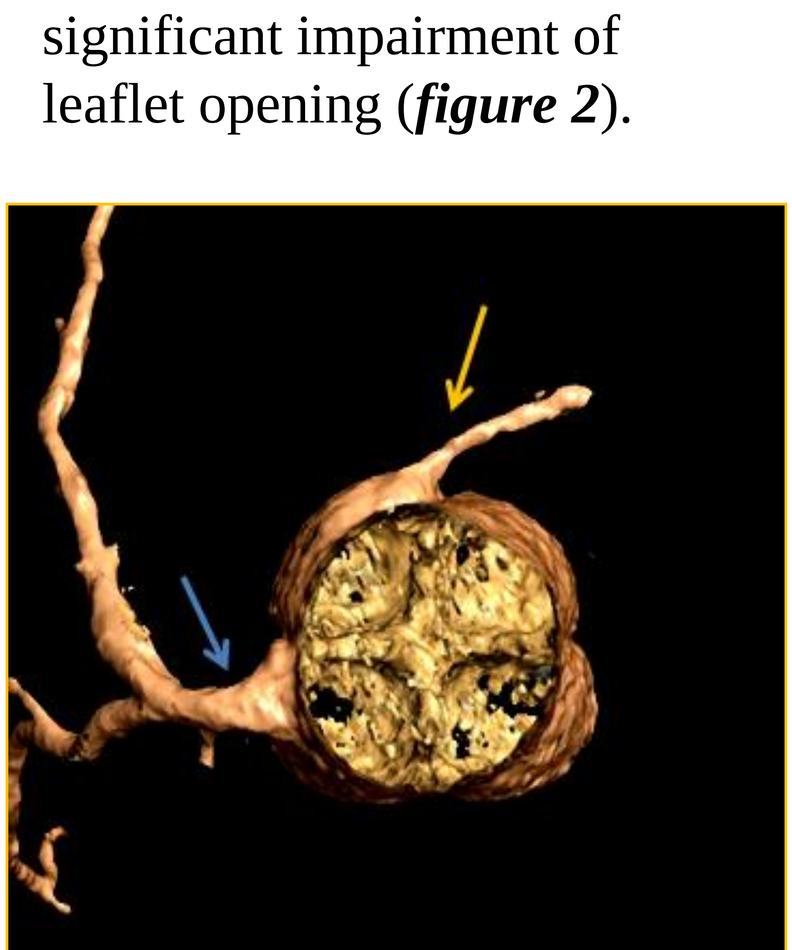

Figure 1:

Topic view of the aortic valve showing emerging of the left and right coronary arteries.
: Left from the posterolateral left coronary sinus.
: Right from the anterolateral left coronary sinus.

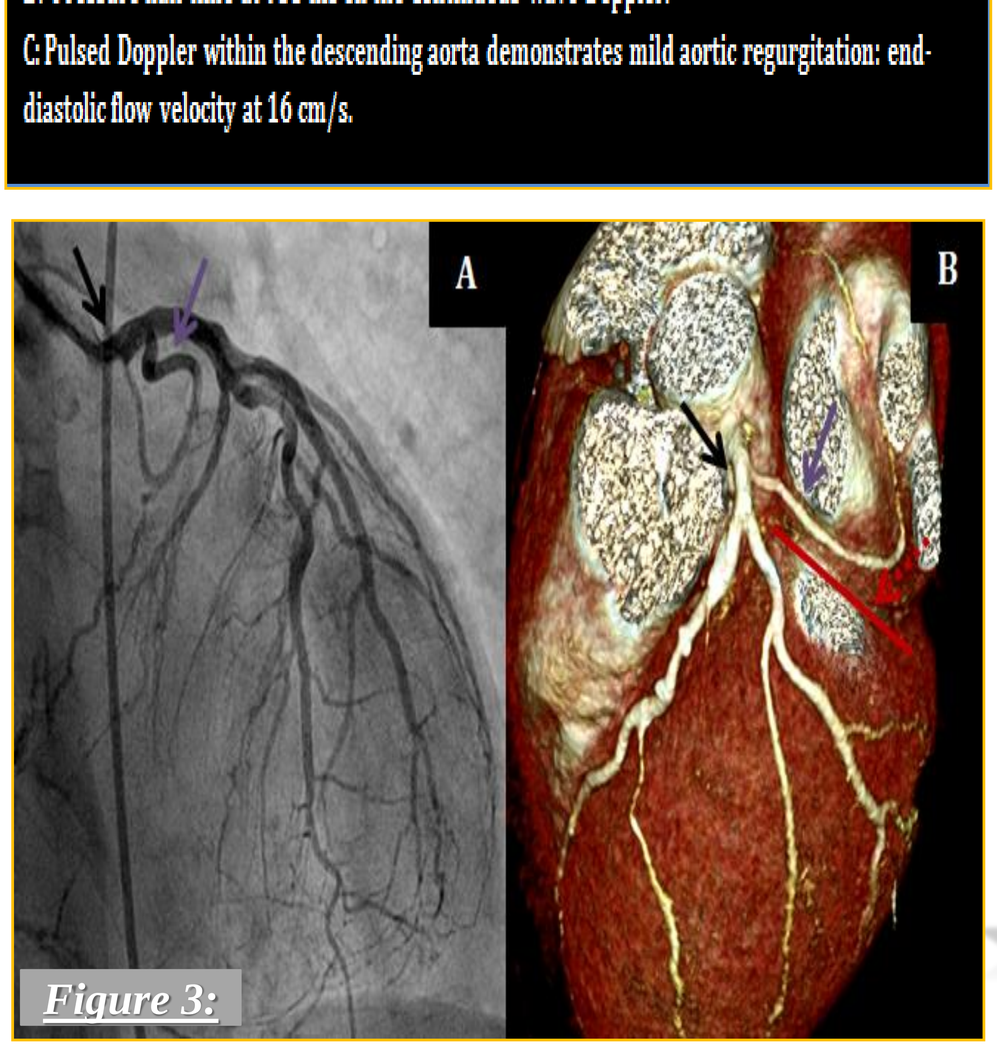

Figure 3:

A: Angiographic view of coronary arteries.
B: Scannographic view of coronary arteries.
: Atrioventricular groove: agenesis of the circumflex coronary.
: Aberrant emerging left atrial branch.
: LAD.

The case

Coronary angiogram showed an occlusion of the distal right coronary artery. In addition, an aberrant coronary branch emerging from the left main was discovered (***figure 3***).
The CT scan with 3D reconstruction provided clear images of the QAV, clarified the origin of each coronary ostia (***figure 4***): the left from the posterolateral left coronary sinus, and the right from the anterolateral left coronary sinus, and described perfectly the peculiar coronary anatomy of this case: **agenesis of the circumflex coronary**, presence of an aberrant branch emerging from the "left main" and heading towards the left atrium, and presence of a big "ramus intermedius" sharing the same origin with the LAD (***figure 3***). Chronic right coronary occlusion was confirmed.

Conclusion

Diagnostic of QAV is rare. Fortuitous discovery of this anatomic entity by TTE should trigger, besides assessment of valve hemodynamic, the search of other cardiac malformations such as coronary artery anomalies. The CT scan can provide a noninvasive accurate assessment of valve morphology and of coronary anatomy.

Refereces

1. Hurwitz LE, Roberts WC. Quadricuspid semilunar valve. Am J Cardiol. 1973; 31:623-6.
2. Feldman BJ, Khandheria BK, Warnes CA, et al. Incidence, description and functional assessment of isolated quadricuspid aortic valves. Am J Cardiol. 1990;65:937-8.
3. Gulyasy B, López-Candales A, Reis SE, et al. Quadricuspid aortic valve: An unusual echocardiographic finding and a review of the literature. Int J Cardiol. 2006;132:68-71.
4. Tutarel O. The quadricuspid aortic valve: a comprehensive review. J Heart Valve Dis. 2004;13:534-7.