

Importance of Echocardiography in the Differential Diagnosis of Rheumatic Mitral Regurgitation in Children and Adolescents

Camila Magalhães Silva,¹ Maik Arantes,¹ Fátima Derlene da Rocha Araújo,¹ Adriana Furletti Machado Guimarães,¹ Zilda Maria Alves Meira¹

Hospital das Clínicas da Universidade Federal de Minas Gerais,¹ Belo Horizonte, MG – Brazil

Introduction

Mitral regurgitation (MR) can be a result of abnormalities in different locations of the valve apparatus (cusps, annulus, tendinous cords and papillary muscles), being of acquired or congenital etiology.¹ MR can be classified as primary, when resulting from a deformity in the valve structure, or secondary, when related to another heart or systemic disease. In our field, the main cause of MR in students and adolescents is the acquired one. Rheumatic fever (RF) is among the causes, as well as bacterial endocarditis, which should be considered in the differential diagnosis.^{2,3} However, congenital anomalies, such as mitral valve (MV) prolapse, mitral cleft (MC), parachute MV and, rarely, isolated cleft MV (ICMV) should be ruled out.^{1,2} The etiological diagnosis of MR is very important to determine the proper treatment, once it can be clinical or surgical, with definitive correction of the lesion.

We present the report of a student whose clinical and echocardiographic scenario was first diagnosed as acute RF, showing the importance of echocardiography in the posterior differential diagnosis of MR.

Clinical case description

Male, 8-year-old student, was admitted in an intensive care unit presenting with rapid onset exhaustion, low fever and paleness. He denied articular involvement or abnormal movements. He reported previous episodes of pharyngotonsillitis unrelated to the current disease and denied previous hospitalization or regular pediatric follow-up. Negative family history of RF. At examination, his general status was compromised. He was feverish, acyanotic, anicteric, with no edema in the lower limbs, full and symmetrical peripheral pulses, moderate tachypnea, inspiratory crackles in lung bases and painful hepatomegaly. Cardiac auscultation showed three heart sounds, second

hyper phonetic sounds, holosystolic murmur grade III-IV/VI, in a mitral area irradiating to the axilla. Laboratory examinations showed leukocytosis, increased C-reactive protein, increased velocity of hemossedimentation and high levels of anti-streptolysin O; blood cultures were negative. Thoracic X-ray showed moderate cardiomegaly in the left chambers, besides lung congestion. Electrocardiography showed sinus rhythm and overload of the left atrium. The first echocardiography diagnosed major MR, increased left chambers and pulmonary hypertension, which raised the possibility of possible rheumatic etiology. The patient was treated for rheumatic carditis with corticotherapy, as well as clinical treatment for heart failure. After discharge, he was referred to the pediatric cardiology outpatient clinic using penicillin G benzathine 1.200.000 UI applied every 21 days, furosemide, captopril and prednisone, remaining stable.

Echocardiographic control confirmed the diagnosis of major MR (Figure 1), increased left chambers, pulmonary hypertension (systolic pressure of the pulmonary artery estimated in 80 mmHg) and left ventricular systolic dysfunction (LVSD 67%). However, morphological evaluation of the MV did not show thickening nor reduction of the cuspid mobility (video); a discontinuity was visualized in the middle third of the anterior cuspid, measuring about 4 mm (Figure 2). There was no change in the aortic valve.

Mitral valvuloplasty was indicated and, during the surgical act, a cleft was identified in the anterior cuspid of the MV and enlargement of the valve ring. Cleft grafting was performed, as well as valve annulus plication, without interurrences. In the follow-up, the child was asymptomatic, with no need for medications, normal cardiac auscultation and normalization of heart chamber dimensions, pulmonary pressure and presence of minimal MR jet.

Discussion

Rheumatic carditis corresponds to the most important manifestation of acute RF, occurring in about 40 to 70% of the cases. MV is the first to be affected in practically 100% of the cases of carditis, manifesting as MR in different grades.² Due to the epidemiology of the disease in our field, it should always be included in the differential diagnosis of acute heart failure and MR, especially in the pediatric age group.² For the diagnosis of the first RF outbreak two major manifestations, or one major and two minor manifestations, are necessary, besides the evidence of previous infection by group A streptococcus.^{3,4} The child in

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Mailing Address: Camila Magalhães Silva •

Avenida Alfredo Balena, 100, quinto andar, Postal code: 30130-100, Santa Efigênia, Belo Horizonte, MG – Brazil

E-mail: camila.magalhaes@live.com

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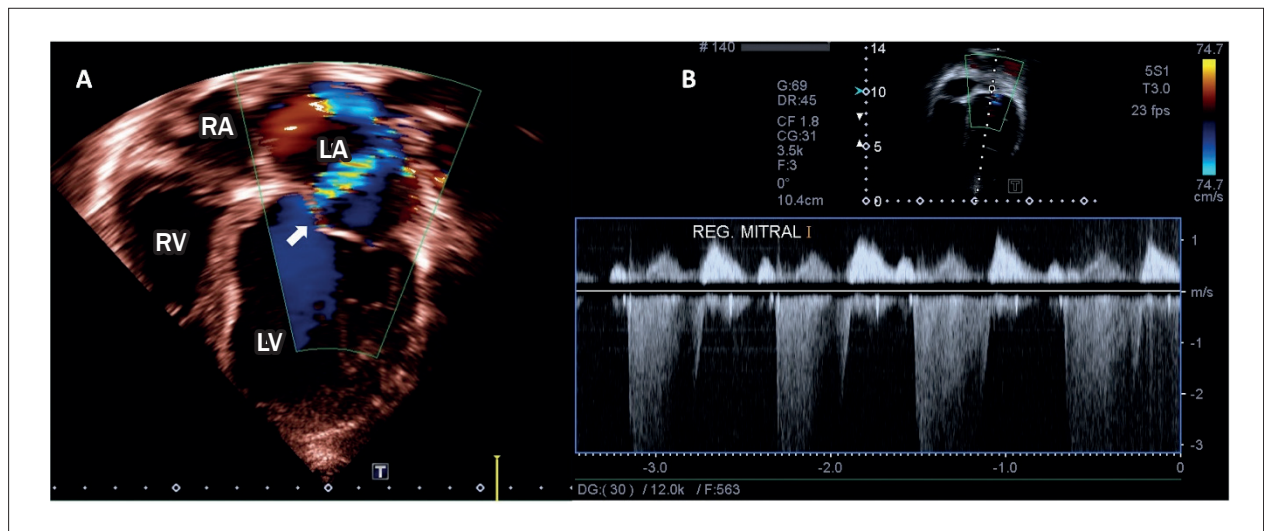


Figure 1 – RA: right atrium; RV: right ventricle; LA: left atrium; LV: left ventricle; Arrow points to the location of the cleft with ColorDoppler image compatible with major MR.

the report presented with a major manifestation (carditis) and two minor manifestations (fever and increased evidence of the inflammatory activity phase); previous streptococia was proven by dosing the antistreptolysin O. Therefore, he was treated for heart failure due to acute rheumatic carditis. However, a detailed analysis of mitral morphology showed that the regurgitating jet originated from a flaw along the anterior cuspid and, for that, the most likely diagnosis would be the perforation of the secondary cuspid to bacterial endocarditis, fact that was not proven by the surgical finding that showed an aspect compatible with congenital cleft isolated from the MV.

ICMV is a rare congenital cause of MR with an incidence of 1:1340 in the pediatric population; it may occur both in the anterior and the posterior MV.⁵ It can be isolated or in association with other congenital heart lesions, and the most common ones are interventricular communication, accessory chordae in the left ventricle outflow, without obstruction, the ostium secundum interatrial communication and the persistence of the arterial channel.^{6,7} When associated with other heart lesions, they can be more asymptomatic, and have an earlier diagnosis when compared to isolated cleft cases. However, after the advent of high resolution bidimensional echocardiography and the tridimensional echocardiography, the ICMV diagnosis has been earlier and more recurrent than in the past.⁸

This case shows how important the echocardiography is in the differential diagnosis of mitral valvulopathy, even in those patients who met the clinical criteria for RF, thus

preventing a mistaken diagnosis and all of the implications related to secondary prophylaxis and late diagnosis of the true etiology.

Author Contributions

Conception and design of the research and critical revision of the manuscript for intellectual content: Araújo FDR e Meira ZMA; acquisition of data: Silva CM, Guimarães AFM, Araújo FDR, Meira ZMA, Arantes M; analysis and interpretation of the data: Guimarães AFM, Araújo FDR, Meira ZMA; writing of the manuscript: Silva CM, Araújo FDR, Meira ZMA, Arantes M.

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No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

Case Report

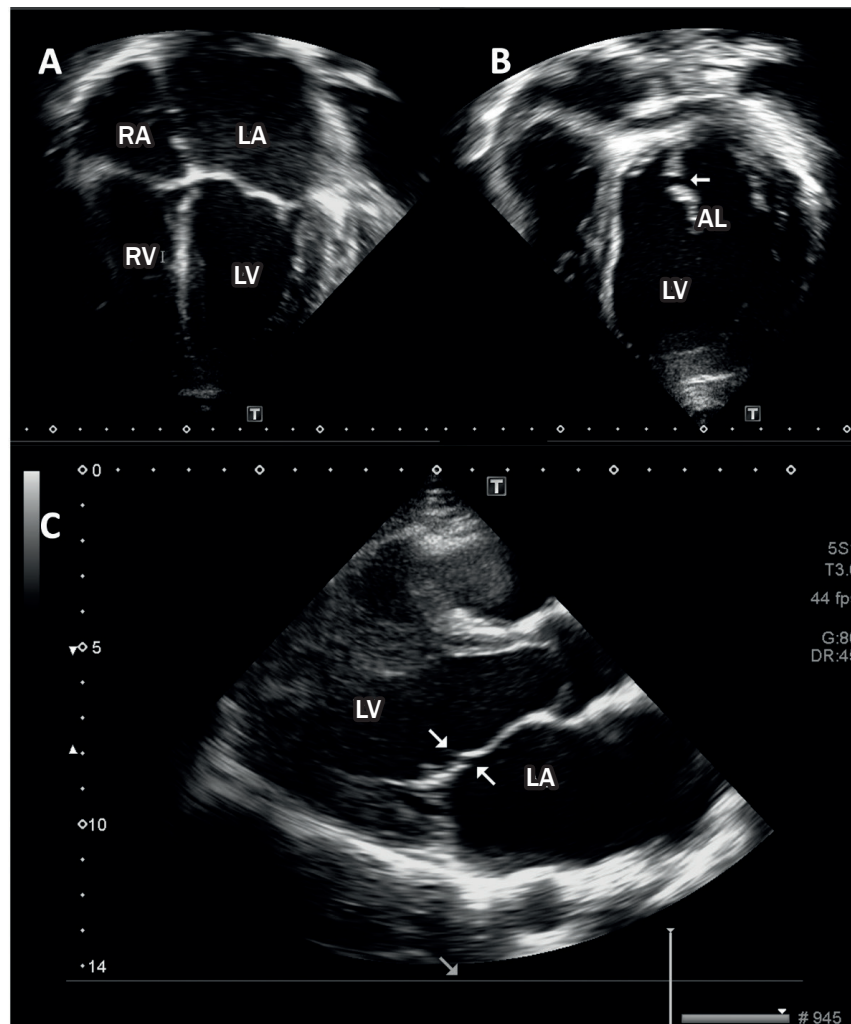


Figure 2 – RA: right atrium; RV: right ventricle; LA: left atrium; LV: left ventricle; AL: mitral valve anterior leaflet. Arrow points to the cleft.

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