Isolated Anterior Mitral Valve Leaflet Cleft: 3D Transthoracic Echocardiography-Guided Surgical Strategy

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Introduction

Isolated cleft of the anterior mitral leaflet (not associated with atrioventricular septal defect) is a rare cause of congenital mitral regurgitation. When feasible, mitral valve repair (direct suturing of the cleft with or without prosthetic annular ring insertion) is preferable to valve replacement. We report a clinical case in which we describe the usefulness of three-dimensional (3D) transthoracic echocardiography (TTE) for diagnosis and morphological assessment of the defect to assist in planning the surgical procedure.

Case Report

An 18-year-old asymptomatic man with a history of systolic murmur from childhood presented for a cardiac evaluation. Cardiac examination detected an apical holosystolic murmur radiating to the axilla. His 12-lead rest electrocardiogram was normal. A two-dimensional (2D) TTE showed the presence of a severe eccentric mitral regurgitation jet directed towards the lateral wall of the enlarged let atrium (Figures A and B and Video 1). The mitral annulus was normally sized. The left ventricle showed normal size and function. No other cardiac abnormalities were detected by 2D TTE. To better define the anatomy of the mitral valve, 3D TTE was performed. "En face" views of the mitral valve were obtained by cropping 3D data sets acquired from both the apical and the parasternal acoustic windows. A defect was visualized in the anterior leaflet of the mitral valve at the level of the A3 scallop (Figures D–I and Video 2). At mid-systole, the defect was 0.8-cm large and 1.2-cm deep with a planimetric anatomic regurgitant area of 0.7 cm^2, while the effective regurgitant orifice was 0.61 cm^2.

Potential acquired causes of this morphological finding such as previous trauma, surgery, and infective endocarditis were also excluded, and the final diagnosis was isolated cleft of the anterior mitral leaflet (ICAML). Considering the large size of the defect, the severity of the regurgitation, and its location near the posteromedial commissural, neither a direct suture nor an autologous pericardium patch implant was considered feasible surgical options (Videos 3 and 4).

Intraoperative assessment confirmed the 3D TTE findings. A cleft was identified in the medial third of the anterior leaflet of the mitral valve. No other abnormalities of the mitral apparatus were found. A 31-mm St. Jude Medical Biocor® prosthesis was then successfully implanted according to the patient's choice. The post-operative course was uneventful, and at 6-month follow-up, the patient remained asymptomatic with a normally functioning mitral prosthesis.

Discussion

ICAML is a rare congenital cardiac disease characterized by a cleft on the anterior mitral valve leaflet that is not associated with an ostium primum atrial septal defect or other features of atrioventricular septal defect[1,2]. ICAML has been hypothesized to be the result of an incomplete expression of an endocardial cushion defect[3]. It usually involves the anterior leaflet, dividing it in its entirety and pointing towards the left ventricular outflow tract without endocardial cushion defect[1]. The mitral annulus is usually in a normal position.

ICAML may cause mitral regurgitation of varying severity. Previous reports have suggested that surgical management should be indicated in the presence of more than mild mitral regurgitation, even in asymptomatic patients[4]. When feasible, surgical repair is the intervention of choice; it consists of a direct suture of the cleft or insertion of an autologous pericardial patch (when the cleft is extremely wide due to a retraction of the cleft's edge), with or without insertion of a prosthetic ring[4,5].

Echocardiography is the technique of choice for evaluating suspected or known mitral valve congenital abnormalities. It provides useful information on the topography and morphology of the defect, as well as the mechanism and severity of the regurgitation. However, due to its tomographic nature, 2D echocardiography, both TTE and transoesophageal (TEE), has limited capability in defining the complex 3D anatomic characteristics of the cleft such as position, size, and morphology. The added value of 3D echocardiography in assessing mitral valve morphology and function has been extensively documented in acquired mitral valve disease[6]. 3D echocardiography allows the display of the non-planar geometry of the valve leaflets and annulus, as well as the complex subvalvular apparatus and its spatial relationships with the

Keywords

Mitral Valve Insufficiency; Echocardiography; Echocardiography, Three-Dimensional; Heart Defects, Congenital.

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Manuscript received April 07, 2014; revised manuscript July 16, 2014; accepted July 22, 2014.

DOI: 10.5935/abc.20140191

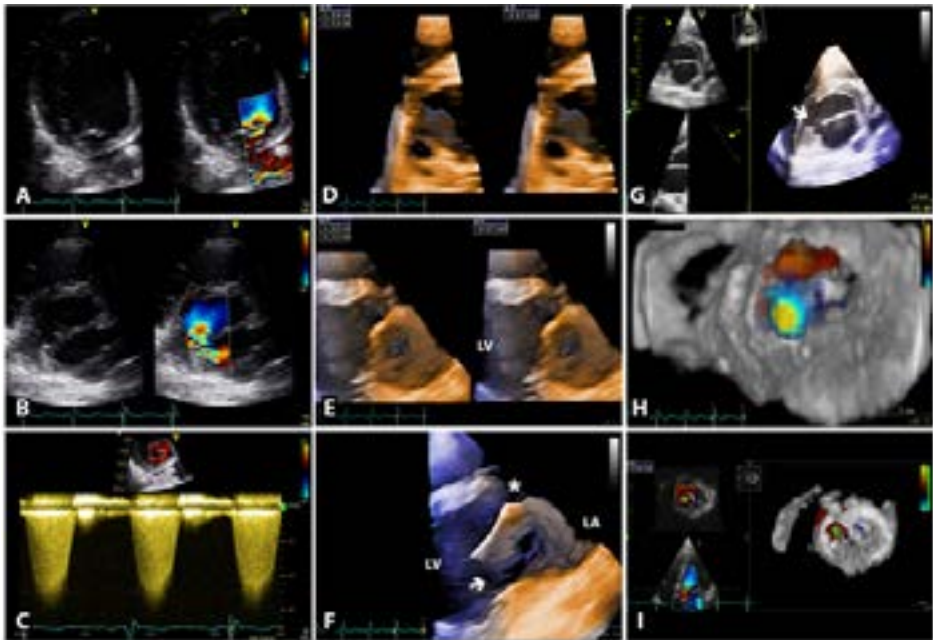


Figure 1 – *A) 2D apical two-chamber view showing a defect in the anterior leaflet, where an eccentric regurgitant flow path is identified by color Doppler; B) 2D short-axis view just below the aortic root, at the level of the aortic to mitral valve fibrous continuity; C) continuous Doppler tracing of the regurgitant flow, showing a dense spectrum suggestive of severe regurgitation; D and E) 3D ventricular and atrial, respectively, "en face" views of the mitral valve at mid-systole showing the anatomic orifice with 3D planimetric area and diameter measurements; F) 3D atrial "en face" view of the mitral valve at diastole demonstrating the cleft in A3; G) 3D TTE acquisition demonstrating the defect in the anterior leaflet echo localized in A3 (note the division in the anterior leaflet as indicated by the arrow); H) 3D ventricular "en face" view of the mitral valve with color Doppler, demonstrating the PISA at the A3 portion; I) 3D effective regurgitant orifice planimetric area.*

surrounding structures. In addition, with 3D echocardiography, there is no need to mentally reconstruct individual tomographic views of the mitral valve because real-time anatomical views of the mitral valve similar to the surgical view could be displayed in the beating heart. Due to its higher image resolution, 3D TEE has been described for the evaluation of ICAML in several case reports[7-12] In our patient, 3D TTE allowed us to visualize ICAML from a surgical perspective, define its exact position, morphology, and size, assist in planning the surgical procedure, and discuss the choices for prosthesis with the patient without the need for semi-invasive procedures such as TEE. Our findings suggest that in patients with good acoustic window, the data provided by 3D TTE allow surgeons to plan the surgical procedure before entering the operating room. A 3D TEE can be performed in the operating room after anesthesia induction to obtain further anatomical details prior to the actual surgery. This strategy will reduce patient discomfort and the corresponding costs for patient workup.

Acknowledgements

Marcelo Miglioranza received a post-graduate grant from CAPES, a Brazilian governmental agency for post-graduate support.

Sorina Mihaila is a recipient of a research grant funded by the European Association of Cardiovascular Imaging.

Denisa Muraru and Luigi P. Badano have received equipment grants from GE Vingmed (Horten, N) and served on the Speakers' Bureau of this company.

Consent

Written informed consent was obtained from each patient that the findings of this study will be published as a case report, together with various kinds of images.

Author contributions

Conception and design of the research and Analysis and interpretation of the data: Miglioranza MH, Muraru D, Mihaila S, Haertel JCA, Badano LP; Acquisition of data: Miglioranza MH, Muraru D, Mihaila S, Haertel JCA; Writing of the manuscript and Critical revision of the manuscript for intellectual content: Miglioranza MH, Muraru D, Mihaila S, Haertel JCA, Iliceto S, Badano LP.

Potential Conflict of Interest

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

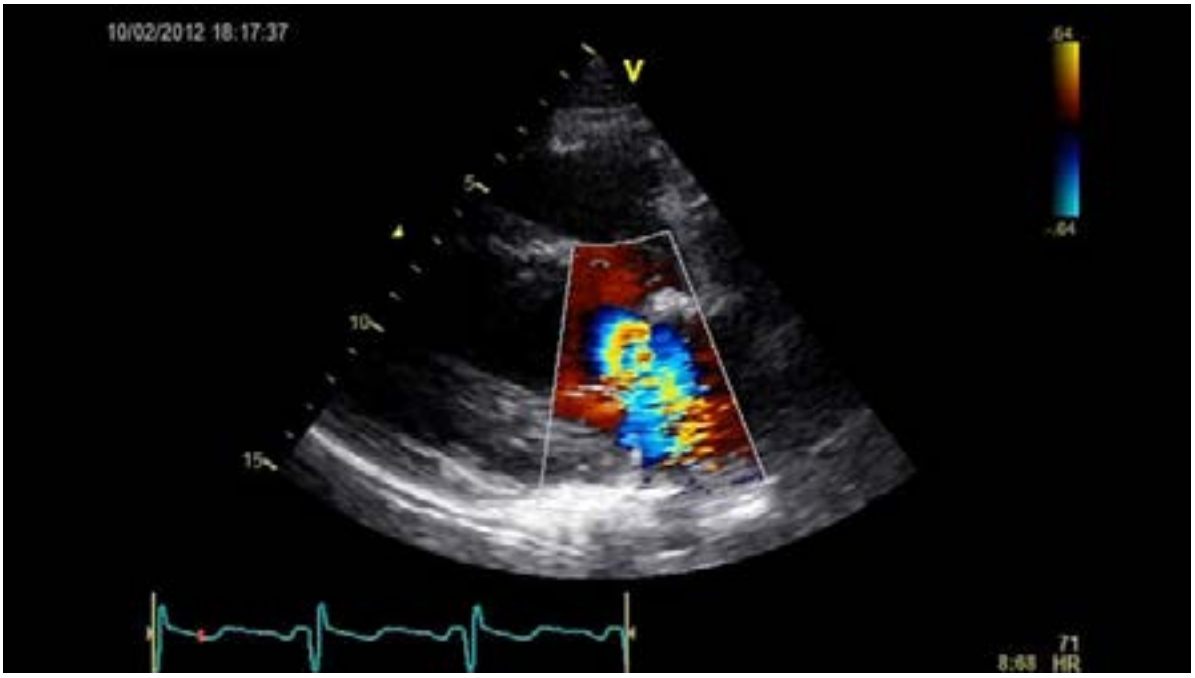
Sources of Funding

There were no external funding sources for this study.

Study Association

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

Case Report



Video – *1) 2D TTE parasternal long-axis view with color Doppler demonstrating an eccentric regurgitant flow through the anterior mitral leaflet and directed towards the left atrium lateral wall; 2) 3D TTE "en face" mitral view in which we could visualize the two portions of the anterior leaflet floating together during systole, whereas during diastole, the edges of the cleft were widely apart; 3) 3D TTE ventricular mitral view in which we could visualize the two portions of the anterior leaflet floating together during systole, whereas during diastole, the edges of the cleft were widely apart; 4) 3D TTE in which an echo-free area is easily visualized in this cut, representing the break in the leaflet.*

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