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Case Report

Isolated Cleft of the Anterior Mitral Leaflet: A Rare Cause of Congenital Mitral Regurgitation

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Abstract

An isolated mitral valve cleft is a relatively rare anomaly. We report the usefulness of real time three-dimensional transthoracic echocardiography in the assessment of isolated cleft of the anterior mitral leaflet in an 11-year-old asymptomatic female child.

Keywords: *Isolated Mitral Valve Cleft; Two-Dimensional Transthoracic Echocardiography; Mitral Regurgitation; Real Time Three-Dimensional Transthoracic Echocardiography*

Introduction

Isolated mitral valve cleft (IMVC) is a rare cause of congenital mitral regurgitation. Surgical correction might be an effective treatment for patients with IMVC, especially for those with moderate to severe MR (even if asymptomatic) [1]. Therefore, it is important to determine the location, shape and size of the MVC when selecting a surgical procedure for treatment. Real-time 3-dimensional transthoracic echocardiography (RT-3DTTE) is a simple and fast imaging study that can collect real-time images and dynamically show the MVC structure and jet flow [2,3]. We describe the usefulness of RT-3DTTE in the assessment of isolated cleft of the anterior mitral leaflet in an 11-year-old asymptomatic female child.

Case Report

An 11-year-old asymptomatic female child was referred to our echocardiography laboratory for evaluation of systolic murmur. Cardiac examination detected an apical holosystolic murmur radiating to the axilla. The electrocardiogram showed sinus rhythm with signs of left atrial and left ventricular hypertrophy. The heart rate was 100 bpm. The chest radiography showed cardiomegaly. Two-dimensional (2D) TTE showed the presence of a severe eccentric mitral regurgitation jet directed towards the lateral wall of the let atrium. The effective regurgitation orifice and the regurgitant volume evaluated by PISA method were 0,56 cm^2 and 80 ml, respectively. The mitral annulus was normally sized; careful observation of the mitral valve leaflets suggested the presence of a mitral cleft. The left atrium and left ventricle were enlarged, with a left ventricular ejection fraction of 75%. No other cardiac abnormalities were detected. To better define the anatomy of the mitral valve, RT3DTTE was performed. RT3DTTE in zoom mode shows a defect in the anterior leaflet of the mitral valve at the level of the A2 scallop (Figure 1A and 1B, blue arrow), Potential acquired causes of this morphological finding such as previous trauma, surgery, and infective endocarditis were excluded, and the final diagnosis was isolated cleft of the anterior mitral leaflet. Under

extracorporeal circulation the child was submitted to correction of the defect by direct suture. The post-operative course was uneventful, and at 6-month follow-up, the patient remained asymptomatic with a trivial mitral regurgitation. On subsequent RT3DTTE, anterior mitral leaflet was *normal* (Figure 1C and 1D).

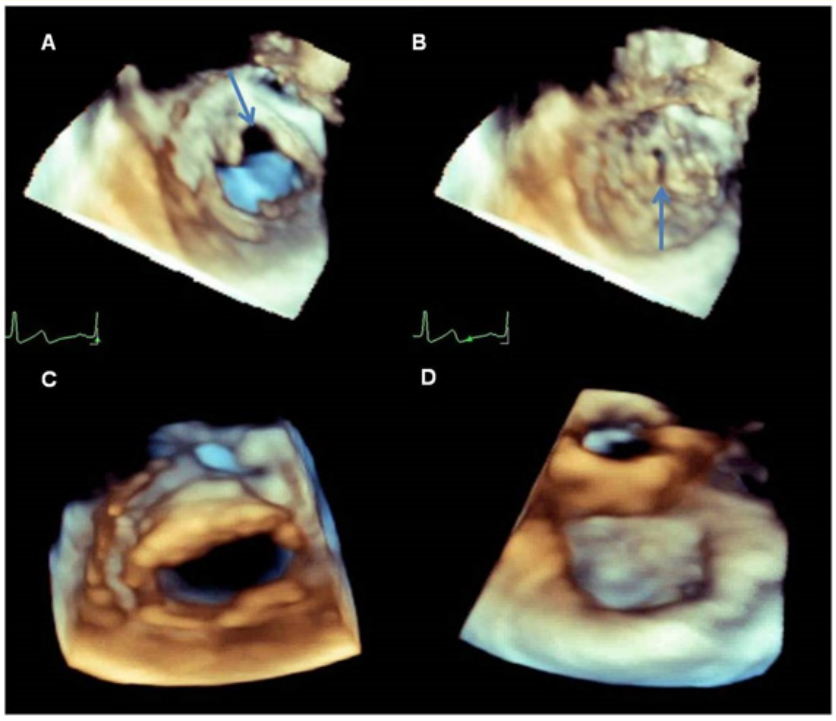

Figure 1: *3DTTE in zoom mode, view from left ventricle in diastole (A); and in systole (B) shows an isolated cleft of anterior mitral leaflet (blue arrow); Postoperative 3DTTE in zoom mode, view from left ventricle in diastole (C); and from left atrium in systole (D) shows a repaired mitral valve.*

Discussion

IMVC is a rare cause of congenital mitral regurgitation. Previous reports have suggested that surgical treatment should be indicated in the presence of more than mild mitral regurgitation, even in asymptomatic patients [1,4]. When feasible, surgical repair is the intervention of choice; consists of direct suturing of the fissure or insertion of an autologous pericardium patch, with or without prosthetic ring insertion [4,5]. Our patient, although asymptomatic, was operated considering the severity of mitral regurgitation. The anatomy and location of the mitral cleft allowed correcting the defect by direct suture with excellent immediate final result and at six months follow-up.

The echocardiogram is the technique of choice to evaluate suspicion or knowledge of congenital anomalies of the mitral valve. However, because of its tomographic nature, 2D echocardiography, both ETT and transesophageal (TEE), has limited ability to define the complex 3D anatomical features of the mitral cleft, such as position, size and morphology. In turn, the RT3DTTE was shown to be superior to 2DTTE in the evaluation of the IMVC [2,3]. In our patient, 3DTTE also allowed us to visualize the IMVC from the surgical point of view, to define its exact position, morphology and size, besides helping in the planning of the surgical procedure.

Conclusion

In conclusion, in patients with good acoustic window, RT3DTTE permit comprehensive 3D viewing of the cleft in the mitral valve and assist in planning the surgical procedure. This type of comprehensive assessment is not possible by 2DTTE, which examines cardiac structures in only two dimensions.

Disclosure

There is no potential conflict of interest for any of the authors.

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