Case report - Congenital

Three-dimensional transesophageal echocardiography in Ebstein’s anomaly

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Received 30 November 2009; received in revised form 28 January 2010; accepted 29 January 2010

Abstract

Three-dimensional (3D) transthoracic echocardiography has advantages over traditional two-dimensional (2D) echocardiography in visualizing tricuspid valve morphology in Ebstein’s anomaly. We describe the application of intra-operative 3D transesophageal echocardiography during a tricuspid valve repair procedure in a patient with Ebstein’s anomaly. Intra-operatively three-dimensional transesophageal echocardiographic (3D TEE) data sets revealed morphology and function of the tricuspid valve, right ventricle outflow tract (RVOT) and pulmonary valve before and after repair. Tricuspid valve leaflet morphology and coaptation as visualized with 3D TEE proved to be consistent with intra-operative findings. Analysis of the tricuspid valve, RVOT and pulmonary valve in the multi-planar review (MPR) mode revealed a bicuspid pulmonary valve, which had not been noticed on the preoperative 2D echocardiographic work-up. In this patient with Ebstein’s anomaly, 3D TEE provided additional information on morphology and function of tricuspid valve, RVOT and pulmonary valve.

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Keywords: 3D TEE; Tricuspid regurgitation; Ebstein’s anomaly

1. Introduction

Ebstein’s anomaly, occurring in about 1–5 per 200,000 births accounts for <1% of all cases of congenital heart disease [1]. It is characterized by adherence of the septal and posterior leaflets of the tricuspid valve to the underlying myocardium. This results in an apical displacement of the valvular annulus into the right ventricle. The tricuspid orifice rotates around the aortic root, below the anatomic atrioventricular junction [2], making it difficult to visualize by two-dimensional (2D) echocardiography. Three-dimensional (3D) transthoracic echocardiography, however, has proven to be capable of showing the morphology and coaptation of the tricuspid leaflets [3–5].

Until today no information is available on 3D TEE and Ebstein’s anomaly.

2. Materials and methods

A 17-year-old woman, diagnosed with Ebstein’s anomaly shortly after birth, was suffering from progressive exercise intolerance, and had undergone several transvenous catheter ablations for recurrent atrial arrhythmias. She was admitted for longitudinal reconstructive surgery of the right ventricle and tricuspid valve.

2D transthoracic echocardiography revealed no abnormalities of the aortic and mitral valve. An enlarged and dilated right atrium was visualized, resulting in dyssynchronous contractions of the intraventricular septum. The tricuspid valve was displaced apically. Coaptation of the leaflets was non-existent, resulting in a severe tricuspid regurgitation. Across the pulmonary valve a pressure gradient of 13 mmHg was measured (v=1.8 m/s). 3D echocardiography had not been possible in the preoperative work-up because of an irregular cardiac rhythm, making it impossible to obtain a full-volume data set at that time.

After induction of anesthesia a 3D TEE was performed with an x7-2t MATRIX-array transducer connected to an iE33 ultrasound system (Philips, Andover, MA, USA). A full volume data set was made up of seven sub volumes and acquired during a 10 s mechanical ventilation hold to avoid motion artifacts during acquisition. Images were analysed on the online workstation Philips Qlab 3D quantification (3DQ advanced) (Philips, Andover, MA, USA).

In the multi-planar review (MPR)-mode three orthogonal cutting planes can be moved independently of each other through the data set [6]. This ability to ‘walk through’ the 3D data set in any 2D image plane and during any time in the cardiac cycle allows a much better appreciation of the complex anatomy and function, allowing visualization of the valvular orifice during systole, revealing a double orifice of malcoaptation, responsible for the severe tricuspid regurgitation. Such a precise description of the tricuspid
valve anatomy by conventional 2D echocardiography remains difficult [3].

With the MPR-mode the location of malcoaptation relative to the separate leaflets and its surface could be assessed, showing two separate zones. These findings were confirmed by surgery. Scanning through the 3D color data set revealed multiple insufficiency jets through the tricuspid valve.

In addition, the right ventricle outflow tract (RVOT) and pulmonary valve were analysed in the MPR-mode, showing how the tricuspid orifice rotates around the aortic root [2] and its extent of displacement towards the RVOT (Video 1). This revealed a bicuspid pulmonary valve (Video 2), which is virtually impossible to image with 2D echocardiography, explaining the increased velocity ($v = 1.8 \text{ m/s}$) over the pulmonary valve.

Surgical correction was performed by mobilizing the anterior and posterior leaflets from the annulus and right ventricle wall and a longitudinal plication of the atrialized portion of the right ventricle which is atrialized may impinge upon the pulmonary valve in a patient with Ebstein’s anomaly.

The patient had an uncomplicated recovery from surgery. Postoperative 2D transthoracic echocardiography four days after surgery showed much improved tricuspid valve function with only mild residual regurgitation.

3. Conclusion

3D TEE with MPR-mode provided additional information on morphology and function of tricuspid valve, RVOT and pulmonary valve in a patient with Ebstein’s anomaly.

References


eComment: Re: Three-dimensional transesophageal echocardiography in Ebstein’s anomaly

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doi:10.1510/icvts.2009.229476A

Ebstein’s malformation is a rare congenital heart disease. Detailed information on the condition of tricuspid leaflets in patients with Ebstein’s anomaly has great value in planning tricuspid valve repair. It is necessary to visualize precisely the location of malcoaptation of each leaflet causing the expressed insufficiency. Actually, two-dimensional (2D) echocardiography cannot provide accurate information about the anatomy of this congenital heart defect. Three dimensional (3D) echocardiography gives better information concerning leaflets and subvalvaral structures [1–3].

Three-dimensional data sets may be analyzed in two main ways: cropping (3D reconstruction) and multplanar review mode. We believe the optimal technique is described in this article [1]. The operator may position the planes through the structures under study, and for example, may study each leaflet individually, and in their relationship to each of the other two leaflets. Thus, the morphology of the individual leaflet and the coaptation planes may be visualized without losing the sight of their anatomical relationship to the rest of the intracardiac structures [3]. The cases in whom the right atrium and the inlet component become grossly dilated due to regurgitation across and the ventricular wall is paper-thin, have a particularly poor prognosis.

Recent developments in 3D echocardiographic analysis software allow to quantify the effectiveness of the function of the right ventricle [3]. Large portion of the right ventricle which is atrialized may impinge upon the function of the left ventricle. The left ventricle in Ebstein’s malformation has been shown to have abnormal shape, as well as regional function, including the lateral and posterior walls [4].

A special problem is tricuspid valve repair in infants and in young patients with right ventricular dysfunction (thinning ventricular wall, low ejection fraction). Prosthetic valves in this group of patients are characterized by their dysfunction within 2–3 years after surgery. When planning surgery in children it makes sense to perform transthoracic 3D echocardiography.
Three-dimensional TEE before surgery should be used for adult patients with poor transthoracic ultrasonic window. Three-dimensional echocardiography can estimate adequate heart valve repair.

References