Quantitative Real-Time Three-Dimensional Echocardiography Provides New Insight into the Mechanisms of Mitral Valve Regurgitation Post-Repair of Atrioventricular Septal Defect

Ken Takahashi, MD, Andrew S. Mackie, MD, SM, Richard Thompson, PhD, Ghassan Al-Naami, MD, Akio Inage, MD, Ivan M. Rebeyka, MD, David B. Ross, MD, Nee S. Khoo, MBChB, Timothy Colen, MBBS, and Jeffrey F. Smallhorn, MBBS, *Edmonton, Alberta, Canada*

Background: Mechanisms of mitral valve regurgitation after atrioventricular septal defect repair are unclear.

Methods: To gain further insight into mitral valve regurgitation, real-time three-dimensional echocardiography was performed in 53 patients after atrioventricular septal defect repair (30 partial and 23 complete) and 40 controls. Mitral valve {x, y, z} coordinates from the annulus, leaflet surface, papillary muscle, and chordal attachments were recorded. Vena contracta area of the regurgitant jet(s) and volume of leaflet prolapse and tethering were measured.

Results: Twenty-three patients had mild (group 1) and 30 moderate (group 2) mitral valve regurgitation. Patients in both groups 1 and 2 had more circular annuli than controls. Annular area was greater in group 2 than in group 1 and controls ($P < .01$). Group 2 had more frequent segmental prolapse in the superior–mural leaflet segment. The anterolateral papillary muscle was more laterally displaced in group 2 than in controls and group 1 at end-diastole ($P = .01$ and $P = .05$) and formed a more acute angle with the mitral valve annulus than in controls or group 1 ($P = .01$).

Conclusions: In patients with atrioventricular septal defects, significant mitral valve regurgitation is associated with leaflet prolapse, larger annular area, and lateral papillary muscle displacement. (J Am Soc Echocardiogr 2012;25:1231-44.)

Keywords: Three-dimensional echocardiography, Endocardial cushion defect, Congenital cardiac defects, Mitral valve

Although current surgical strategies have led to an improvement in survival after atrioventricular septal defect (AVSD) repair, progressive mitral valve regurgitation represents an ongoing challenge.[1,2] Several studies have addressed risk factors for the development of postoperative mitral valve regurgitation, but precise mechanisms remain unclear.[3-5] Anomalies of the papillary muscles, the mitral valve, and an open cleft have been believed to accentuate the degree of regurgitation.[6-8] Despite this, the main focus has been on follow-up of surgical results and qualitative assessment.

The term *left atrioventricular valve* is commonly used in the setting of an AVSD because the left-sided inflow valve in no way represents a normal mitral valve. The normal mitral valve is bileaflet and consists of a triangular anterior and a broader posterior leaflet. In "all hearts" with AVSDs, the more posterior leaflet is much narrower and smaller and is called the mural leaflet (ML). The remaining two other left-sided leaflets are referred to as the superior bridging leaflet (SBL) and inferior bridging leaflet (IBL), with a cleft separating them as they bridge the interventricular septum (Figure 1). For the sake of clarity and to avoid confusion, the left-sided atrioventricular valve in patients with AVSDs is referred to here as a "mitral valve." The papillary muscle location in an AVSD is different from the normal mitral valve, with counterclockwise rotation of the posteromedial papillary muscle when viewed in short axis from the apex, a feature linked to the narrower ML (Figure 1).

Real-time three-dimensional (RT3D) echocardiography permits a qualitative and quantitative evaluation of atrioventricular valve function in both the acquired and congenital populations.[9-19] We have previously applied RT3D echocardiography to evaluate the mitral valve in AVSD but in a more qualitative fashion.[13,16-18] The purpose of this study was to apply quantitative RT3D echocardiography to gain further insight into the mechanisms of mitral valve regurgitation in patients after AVSD repair.

METHODS

Patient Characteristics

Between January 2006 and November 2007, we prospectively acquired two-dimensional (2D) transthoracic echocardiographic and

From the Department of Pediatrics (K.T., A.S.M., G.A., A.I., N.S.K., T.C., J.F.S.), the Department of Surgery (I.M.R., D.B.R.), and the Department of Biomedical Engineering (R.T.), University of Alberta and Stollery Children's Hospital, Edmonton, Alberta, Canada.

Reprint requests: Jeffrey F. Smallhorn, MBBS, Stollery Children's Hospital, Room 4C2, Walter C. Mackenzie Health Sciences Centre, Edmonton, AB T6G 2B7, Canada (E-mail: *jeffrey.smallhorn@albertahealthservices.ca*).

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Abbreviations

AP = Anterior-posterior

AVSD = Atrioventricular septal defect

BSA = Body surface area

CC = Commissure-commissure

IBL = Inferior bridging leaflet

ML = Mural leaflet

RT3D = Real-time three-dimensional

SBL = Superior bridging leaflet

3D = Three-dimensional

2D = Two-dimensional

VC = Vena contracta

transthoracic RT3D echocardiographic images from patients with AVSDs who had undergone complete repair and were >3 years of age. Patients were sequentially recruited from the cardiac outpatient clinic at the Stollery Children's Hospital. The inclusion criteria were ability to cooperate for the study, technically adequate RT3D echocardiographic images of the "mitral valve" and its subvalvar apparatus, laminar flow across the left ventricular outflow tract, and sinus rhythm. Exclusion criteria were any form of left ventricular outflow tract obstruction, unbalanced AVSD, and right ventricular hypertension (right ventricular pressure greater than half systemic). We collected mitral valve data from normal subjects of similar age for comparison. The study was approved by the University of Alberta ethics committee.

Two-Dimensional Echocardiography

Two-dimensional transthoracic echocardiography was performed using an iE33 system (Philips Medical Systems, Andover, MA). Left ventricular global function was assessed by shortening fraction and corrected velocity of circumferential fiber shortening. Left ventricular volume was measured using the biplane Simpson method. Mitral valve inflow mean Doppler velocity was recorded, as was the right ventricular pressure from tricuspid valve regurgitation or interventricular septal shape. The degree of mitral valve regurgitation was assessed by vena contracta (VC) width measured at the orifice in the parasternal long-axis view and characterized on a 0 to 3+ grading scale, defined as follows: 0 = no mitral valve regurgitation, 1 = mild (VC width < 0.3 cm), 2 = moderate (VC width, 0.3–0.69 cm), and 3 = severe (VC width > 0.7 cm), per the published guidelines of the American Society of Echocardiography.[19] In a similar fashion to our previous study,[20] we combined grades 0 and 1 to represent mild (group 1) and grades 2 and 3 (group 2) to represent moderate mitral valve regurgitation.

Acquisition of RT3D Data Sets

Images were obtained using a RT3D ultrasound system (iE33) with a transthoracic matrix X3-1 or X7-2 probe. Full-volume images of the mitral valve and papillary muscles at end-expiration were obtained from a position as close to the apex as possible, including a color Doppler data set from the same position to permit precise anatomic location of any regurgitant jets. During acquisition, the high–frame rate mode was selected to optimize temporal resolution, and high density was selected to optimize image spatial resolution. The apical position was chosen for image acquisition, because this provided an image of the mitral valve in the axial plane, hence again optimizing leaflet resolution.

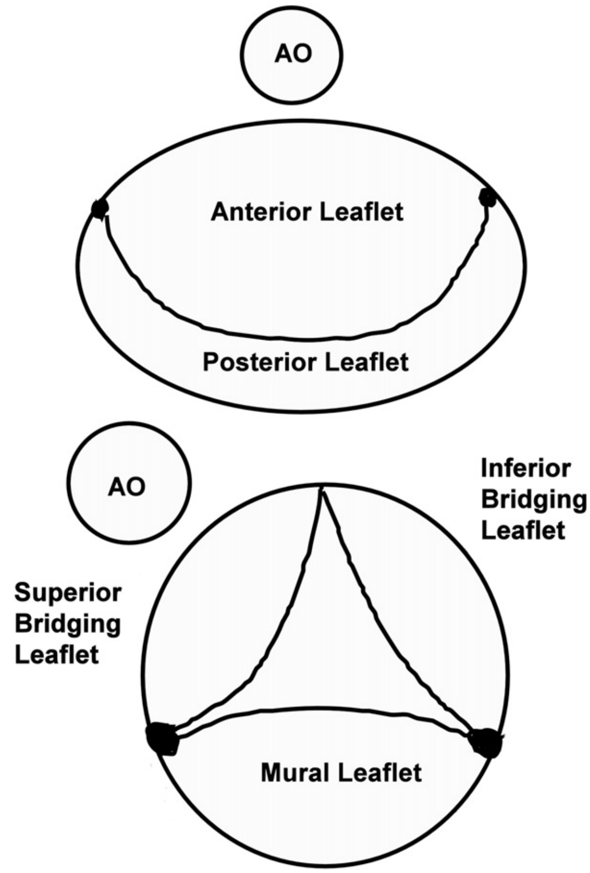

Figure 1 Schematic diagram of the comparison between a normal mitral valve (*top*) and the left-sided inflow valve after repair of an AVSD. Note the smaller ML in the diagram of the AVSD, as well as the different position of the papillary muscles. *AO*, Aorta.

Mitral Valve Three-Dimensional Analysis

Mitral valve assessment software (TomTec Imaging Systems, Munich, Germany) was used to analyze the RT3D data sets. Data points were recorded at end-diastole and midsystole. Exploration of the RT3D data set was performed using multiplanar 2D views. This enabled markers to be placed to delineate the annular hinge points (Figures 2A and 2B). We then obtained 15 vertical radial and equally spaced 2D planes (24°) passing through the center of the mitral valve (Figure 2C). Our reference within each RT3D data set was marked between the aortic and mitral valves. On these 2D planes at midsystole, we placed nine makers on the mitral valve leaflets to delineate its shape in three-dimensional (3D) space for subsequent reconstruction (Figures 2A and 2C). Using the multiplanar navigation tool, we located the tip of the anterolateral and posteromedial papillary muscle (Figures 2A, 2B, and 2D) and marked the subvalvar apparatus position with five points (Figures 3A, 3B, and 3C): (1) the base of the papillary muscle, (2) the tip of the papillary muscle, (3) the chordal attachments on the leaflet that were closest to the tip of the papillary muscle, (4) the annular point closest to the papillary muscle tip, and (5) the annular point furthest from the papillary muscle tip. All 3D marked locations were converted into spatial coordinates {x, y, z} and exported to MATLAB (The MathWorks Inc, Natick, MA) to extract measures of annular, leaflet, and papillary muscle geometry as detailed below.