This study describes the spectrum of CMR findings in a cohort of ARVD/C-associated mutation carriers, including first-degree relatives of ARVD/C patients who were "at risk" of disease. Our study has several important findings. First, in ARVD/C mutation carriers with structural disease on CMR, biventricular abnormalities were seen with similar frequency to isolated RV disease. Involvement of the LV in subjects with ARVD/C has been increasingly recognized, with prevalence reported in up to 80% of ARVD/C patients in several European populations (1,2) and 40% in a cohort of North American and Dutch patients (4). Our study confirms the high prevalence of biventricular disease in North American patients with structurally abnormal hearts and genetic predisposition to ARVD/C. Interestingly, though, LV abnormalities were not detrimental to cardiac function with no significant differences between groups with and without LV involvement. Furthermore, demographic and RV findings were similar between individuals with isolated RV disease and those with biventricular disease, suggesting that LV abnormalities may occur at any stage of RV disease and are not limited only to subjects with advanced RV disease. This finding argues against the traditional notion that LV abnormalities are a late complication of the disease (5). More likely, there is a spectrum of the disease process that varies from patient to patient, extending across both ventricles. Finally, the only finding significantly different between subjects with isolated RV disease and those with LV abnormalities was genotype, suggesting an association between genotype and structural phenotype in ARVD/C mutation carriers.

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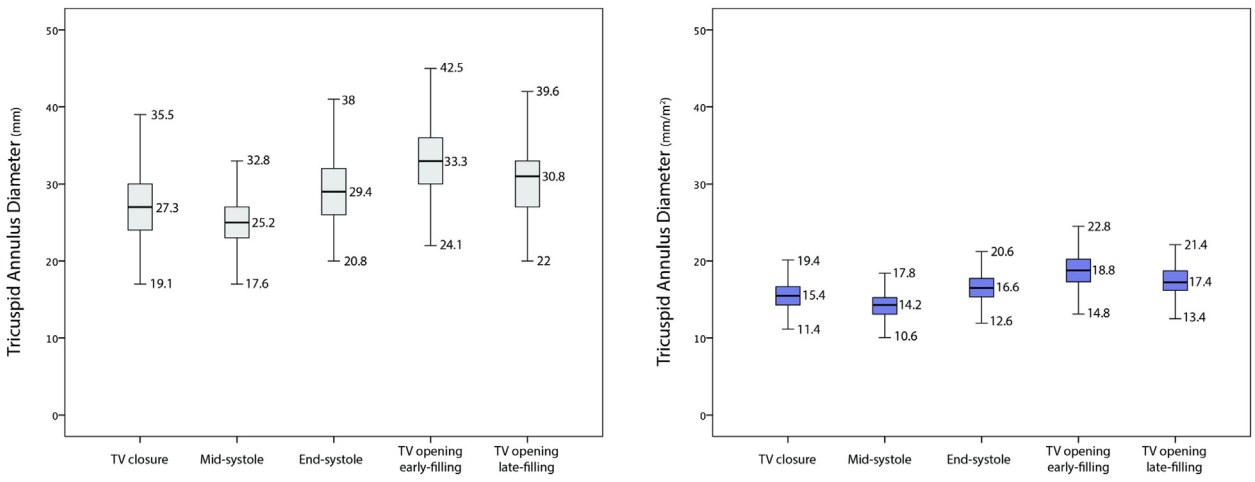
Variability of Tricuspid Annulus Diameter Measurement in Healthy Volunteers

Tricuspid valve (TV) anatomy and function play an important prognostic role in several heart diseases and in the development of functional tricuspid regurgitation. According to current guidelines for management of heart valve disease, the tricuspid annulus (TA) diameter measured by 2-dimensional transthoracic echocardiography (2DE) should be used to define the need of an associated TA annuloplasty in patients undergoing cardiac surgery for left-sided heart valve diseases (1,2). However, the timing during the cardiac cycle when the TA should be measured remains to be established. Moreover, normative data about TA diameter and function are limited (3–5).

Therefore, we designed a prospective cross-sectional study of 219 normal volunteers (age 43 ± 15 years; 57% female; body mass index <30 kg/m^2) to assess the variability of TA diameter measurement in 2DE 4-chamber view (4CH) in relation to timing during the cardiac cycle.

The TA diameter was obtained from the apical right ventricular (RV)-focused 4CH using a Vivid E9 (GE Vingmed, Horten, Norway) equipped with M5S probe and measured as the distance between the insertion points of the TV leaflets (inner edge to inner edge) at 5 time points during the cardiac cycle. The times during the cardiac cycle were determined using both electrocardiogram and valve dynamic visualization: TV closure (end-diastole—the first frame after the TV closure), mid-systole (beginning of T-wave), end-systole (end of T-wave), TV opening early filling (as the frame with the TV wide open during passive flow), TV opening late filling (as the frame with the TV wide open during active flow—after P-wave).

FIGURE 1 Tricuspid Annulus Diameter Variability in Healthy Volunteers Obtained From Apical Right Ventricular–Focused 4-Chamber View



(Left) absolute values. **(Right)** values indexed to body surface area. Whisker lengths ±2 SD from the mean value. TV = tricuspid valve.

Fractional shortening (FS) of the TA diameter was calculated using the largest and smallest diameters and expressed as a percentage. Right atrial (RA) and RV volumes were obtained using 3DE full-volume datasets acquired with a 4V matrix-array transducer (GE Vingmed) and measured using dedicated software designed for volumetric analysis of the left atrium (LA and RV analysis, Tomtec Imaging Systems, Unterschleissheim, Germany).

The feasibility of measuring the TA diameter in 4CH by 2DE was 99.5%. Absolute TA diameters were significantly larger in men than in women. However, this difference was eliminated after indexation by body surface area (Figure 1). In healthy subjects, the TA diameter increased from mid-systole to early diastole and then decreased (FS 24.1 ± 6.2%; Δ 8.0 ± 2.4 mm; $p < 0.001$), with a consistent correlation with the phasic changes of RA ($r = 0.56$; $p < 0.001$) and RV volumes ($r = 0.53$; $p < 0.001$), and height ($r = 0.49$; $p < 0.001$), weight ($r = 0.54$; $p < 0.001$). At multivariate linear regression analysis, age ($r^2 = 0.2$; $p < 0.001$), sex ($r^2 = 0.2$; $p = 0.002$), and RA ($r^2 = 0.45$; $p < 0.001$) and RV volumes ($r^2 = 0.2$; $p < 0.001$) were independently correlated with TA absolute diameters and accounted for 55% of the variance of mid-systolic TA diameter in the 4CH view.

Applying the cutoff values for TA annuloplasty proposed in the current guidelines and in published reports (TA >40 mm or FS <25%) (1,2,4) to our study population resulted in a significant number of our healthy subjects fulfilling 1 or more criteria for intervention: 19.7% with dilated early diastolic TA in 4CH view (95% confidence interval: 14.4 to 25) and 58% with reduced FS in the 4CH view (95% confidence interval: 51.2 to 64.4).

It is well known that the complex geometry of the TA is difficult to assess by a tomographic imaging technique like 2DE. However, current guidelines recommend 2DE to assess TA size and indicate the need for tricuspid annuloplasty. The present study provided reference values for TA diameter obtained in the 4CH view specific for 2DE and identified the main determinants of TA size. Age, sex, and right chamber sizes, as well as the timing during the cardiac cycle, significantly influence the TA diameter in 4CH in healthy individuals. According to our data, TA evaluation to define the need for TV annuloplasty should include diameter measurements at both mid-systole and early diastole, complemented with right heart chamber volumes. Our data may help to correctly identify TA enlargement by 2DE.

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A Combined Transcatheter Aortic Valve-in-Valve and Pulmonary Valve Implantation

Patients with surgically treated congenital cardiac disease increasingly survive into adulthood and face the need for repeat sternotomies for senescent prosthetic valves and conduits. This poses increased risk of chest re-entry injury, and alternatives to redo median sternotomy are important. Although transcatheter aortic valve-in-valve (ViV) and transcatheter pulmonary valve implantation (TPVI) (Melody valve, Medtronic, Minneapolis, Minnesota) have both been separately described (1–3), we describe a combination of aortic ViV and TPVI utilizing 3-dimensional (3D) computed tomography (CT) fluoroscopic coregistration to aid procedural execution. A 24-year-old man with congenital bicuspid aortic stenosis, prior Ross procedure, re-replacement of the right ventricle-pulmonary artery homograft, ascending aortic aneurysm repair, and aortic valve replacement (25-mm Perimount bioprosthesis, Edwards Lifesciences, Irvine, California) was referred for symptomatic aortic and pulmonary conduit stenoses. Aortic valve mean gradient was 52 mm Hg, and mean pulmonary conduit gradient was 38 mm Hg. CT demonstrated pulmonary conduit stenosis and an aortic annular area of 360 mm^2 (annular dimensions 21.9 mm × 21.5 mm). The distances of the right coronary artery origin and proximal left anterior descending artery from the pulmonary artery were 8.7 mm and 11.9 mm, respectively. Coronary artery relational anatomy was felt to be favorable for intervention with combined transcatheter aortic ViV and TPVI (**Figure 1A**).

Using standard techniques, a 26-mm transcatheter aortic valve replacement (Sapien-XT, Edwards Lifesciences) was successfully undertaken within the aortic prosthesis. Post-deployment echocardiographic assessment demonstrated normal valve function. A noncontrast rotational angiogram of the cardiac and vascular structures was then performed, and images were matched with previously performed CT reconstructions of the pulmonary conduit. Coregistration using known skeletal landmarks enabled overlay of 3D images onto live fluoroscopy to guide transcatheter pulmonary valve placement. An exchange-length stiff guidewire was placed in the left lower lobe pulmonary artery using a balloon wedge catheter, and serial balloon dilations of the pulmonary conduit were performed using 18- and 22-mm balloons with simultaneous left coronary angiography demonstrating no coronary-conduit interaction. A 22-mm diameter balloon-in-balloon "BIB" catheter (NuMed, Hopkinton, New Jersey) with a mounted stent (Palmaz-XL-4010, Cordis, Bridgewater, New Jersey) was delivered via a 14-Fr Mullins sheath into the pulmonary conduit. This was used to expand the homograft conduit and prepare a landing zone to minimize future risk of transcatheter pulmonary valve stent fracture. Fluoroscopic and 3D CT overlay facilitated accurate positioning and deployment of the stent. The stent was then post-dilated using a 22-mm balloon, and no stent recoil was noted. A 22-mm transcatheter pulmonary valve was then successfully deployed using a 22-mm delivery system (Ensemble delivery system, Medtronic) within the landing zone created by the stent (**Figures 1B and 1C**). The peak-to-peak catheter right ventricular outflow tract (RVOT) gradient was reduced to 5 mm Hg. Transesophageal echocardiography showed normal valve function with mild regurgitation. The patient was dismissed from the hospital on day 2 with a mean transaortic gradient of 21 mm Hg, and 11 mm Hg across the RVOT, with no demonstrable regurgitation.

This presentation is unique in that this was a young adult with 3 previous median sternotomies within a period of 6 years, and efforts to avoid a repeated median sternotomy were critical. Multidisciplinary assessment and 3D imaging were essential to procedural planning and execution. Although annular calcification and radiographic signatures of prosthetic valves can often serve as fluoroscopic markers and