The Congenital Heart Surgeon's Society Complete Atrioventricular Septal Defect Cohort: Baseline, Preintervention Echocardiographic Characteristics

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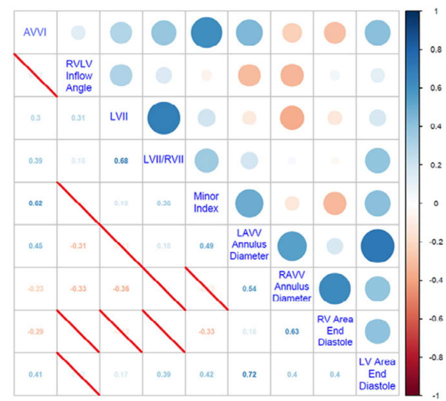
Quantifying unbalance, the threshold for single ventricle palliation vs biventricular repair in patients with unbalanced complete atrioventricular septal defect (AVSD), is challenging. Using a core laboratory review of baseline echocardiograms, we sought to assess the correlations among commonly used measures of unbalance and common atrioventricular valve (AVV) and ventricular sizes. A single reviewer evaluated baseline echocardiograms from an inception cohort of babies age <1 year with complete AVSD admitted to 1 of 25 Congenital Heart Surgeon's Society institutions. A standardized echo review protocol of 111 quantitative and qualitative measures was used. Descriptive statistics were computed and Pearson correlation coefficients were calculated to assess correlation among unbalance indices with valvar and ventricular dimensions. Two-hundred fifty-seven baseline echocardiograms of infants with complete AVSD were included. Median age at baseline echocardiogram was 11 days (interquartile range 1–79) and mean atrioventricular valve index was 0.45 ± 0.1. Mean right ventricle/left ventricle inflow angle was 90.2 ± 15.6° and median left ventricular inflow index was 0.46 (interquartile range 0.4–0.5). There are weak or moderate correlations between the measures of unbalance. Correlations between the measures of unbalance with common AVV leaflet or ventricular sizes are also weak to moderate, when statistically significant. Measures of unbalance in common clinical use correlate poorly, or not at all, with one another, common AVV, and ventricular dimensions. The concept of "unbalance" is difficult to define using baseline echocardiographic indices. These findings suggest that the indices may describe different morphologic and functional characteristics. Further analysis is necessary to quantify the contributions of unbalance indices to patient outcome.



Atrioventricular valve index correlates poorly with common valvar and ventricular sizes.

Central Message

Current echocardiographic measures of unbalance do not correlate well with common atrioventricular valve or ventricular dimensions.

Perspective Statement

Defining the degree of unbalance in patients with complete atrioventricular septal defect is crucial for surgical decision-making. In a core laboratory review of baseline echocardiograms, limited correlation was seen between echocardiographic measures of unbalance with valvar and ventricular measurements. Future study will involve analyzing these measures with regard to surgical triage and outcomes.

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Funding: Funding for Dr. James M. Meza and Dr. Paul J. Devlin was provided by the Congenital Heart Surgeons' Society's John W. Kirklin/David Ashburn Fellowship, the Hospital for Sick Children Division of Cardiovascular Surgery, and the Ann & Robert H. Lurie Children's Hospital of Chicago Division of Cardiovascular-Thoracic Surgery.

Presented at AATS Centennial, May 2, 2017, Boston, MA.

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https://doi.org/10.1053/j.semtcvs.2018.02.004

Semin Thoracic Surg 31:80–86

Keywords: Echocardiography, Atrioventricular septal defect, Surgical decision-making

INTRODUCTION

The echocardiographic definition of unbalanced, complete atrioventricular septal defect (AVSD) remains controversial, as the anatomic and physiologic features that predict successful biventricular repair vs univentricular palliation are still poorly characterized. Different echocardiographic measures of unbalance have been proposed, focusing on the size and function of the left or right atrioventricular valve (AVV) components, as well as left and right ventricular size and function.[1] Echocardiographic indices in clinical use for defining unbalance include:

(1) atrioventricular valve index (AVVI): the ratio between the left AVV area over the total AVV area,[2]
(2) left ventricle inflow index (LVII): the ratio between the widths of the color Doppler inflow at it is narrowest point to the corresponding AVV anulus[3]
(3) RV/LV inflow angle: the angle between the hinge points of the right and left AVVs with the vertex at the crest of the interventricular septum measured in an apical 4-chamber view.[4]

The interrelationships among these indices and their correlations with measurements of ventricular size and function are not well established. The Congenital Heart Surgeon's Society (CHSS) Data Center's prospective multicenter inception cohort study of patients undergoing surgery at the participating CHSS institutions collects echocardiographic data for analysis in its imaging core laboratory (ICL). We sought to describe the echocardiographic features defining unbalance in a large patient cohort. Our primary aim was to assess the correlations among the measures of unbalance and common AVV and ventricular dimensions. We hypothesized that the currently used indices of unbalance would correlate strongly with each other and with common AVV and ventricular dimensions.

METHODS

Patients

The CHSS Complete AVSD prospective inception cohort enrolls infants age ≤1 year when admitted to a CHSS institution and diagnosed with complete AVSD with concordant atrioventricular and ventriculoarterial connections. Enrollment started in January 2012 and is ongoing. This study analyzed patients enrolled between January 2012 and March 2017 for whom a baseline echocardiogram was available. Those with partial or transitional AVSD, anomalous pulmonary venous return (total or partial), aortic atresia, or heterotaxy syndrome were excluded. Consent for enrollment was obtained from the patients' parents or legal representatives. Ethical approval was obtained at each participating institution.

Echocardiographic Analysis

Each participating CHSS institution followed a designated echo protocol to obtain specified views. Training of institutional echo staff was provided through webinars. All echocardiograms were sent to the CHSS Data Center for analysis by the ICL. Data were uploaded into EchoPac software (GE Ultrasound, Horten, Norway). Reviews were performed by a single experienced observer (CTF) according to a predefined standardized protocol of 111 quantitative and qualitative measures. All quantitative measurements were repeated 3 times and the results were averaged. All standard echocardiographic measures were performed according to the guidelines published by the American Society of Echocardiography.[5] A complete list of all measures obtained is available in Supplement 1.

AVVI was measured according to Cohen et al. from a subcostal anterior oblique view of the AVV.[6] The orifice of the common AVV is traced in diastole. Using a line corresponding to the plane of the interventricular septum, the common AVV is subdivided into left and right AVV components, and the left and right AVV areas are measured (Fig. 1A). The AVVI was first described as the left AVV area divided by the right AVV area.[6] It has been modified for clarity and is now calculated by dividing the left AV valve area by the total common AVV area.[2] An AVVI within 0.4–0.6 is considered balanced, ≥0.6 is classified as left dominant, and ≤0.4 classified as right dominant.

The *RV/LV inflow angle* is the angle between the hinge points of the right and left AVVs measured from apical 4-chamber view. For this study, it was measured at end diastole (Fig. 1B).

LVII is based on a color Doppler image obtained from the apical 4-chamber view.[3] The left AVV anulus is measured in end diastole between the medial and lateral hinge points of the left AVV (anatomic orifice). The narrowest point of the color inflow jet is measured (secondary orifice). The LVII is the ratio of secondary orifice diameter to the anatomic anulus (Fig. 1C and D). The RVII is the corresponding right-sided measurement. We also calculated the ratio of LVII/RVII. The *minor index* is the ratio of LV to RV width at end diastole from the apical 4-chamber view (Fig. 1E).

Statistical Analysis

Descriptive statistics were computed. Categorical variables are presented as frequencies or percentages. The normality of the distribution of continuous variables was investigated using the Shapiro-Wilk test. Continuous variables are presented as means with standard deviations or medians with interquartile ranges (IQR). Pearson correlation coefficients were calculated, using pairwise deletion, among indices of unbalance and other echocardiographic measurements. A Pearson correlation coefficient of 0–0.3 indicates a weak correlation, 0.3–0.7 indicates moderate correlation, and >0.7 indicates strong correlation. All analyses were performed using SAS version 9.2 (SAS Institute, Cary,

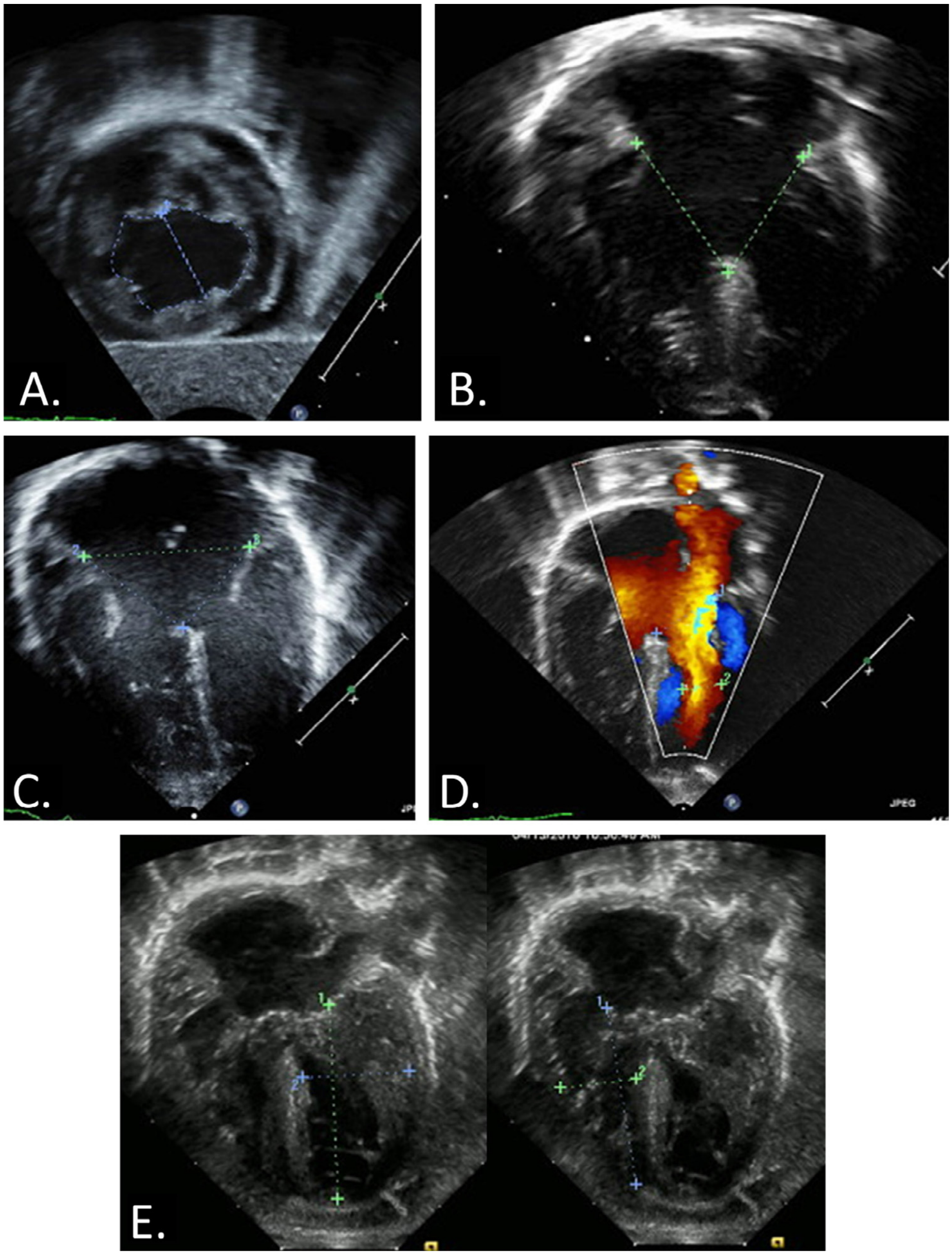

Figure 1. Measures of unbalance. (A) Atrioventricular valve index. (B) Right ventricular/left ventricular inflow angle. (C) Left ventricular inflow index, left atrioventricular valve anulus diameter. (D) Left ventricular inflow index, color inflow diameter at the papillary muscles. (E) Minor index, left ventricular width/right ventricular width. (Color version of figure is available online.)

NC) and R version 3.4.1 (R Foundation for Statistical Computing, Vienna, Austria).

Patient Characteristics

In total, 375 patients from 25 institutions are enrolled in the CHSS Complete AVSD cohort, among whom 257 (69%) had echocardiograms available for analysis. Of the study population, 48% were male. Eighty percent were diagnosed with trisomy 21. The median age at baseline echocardiogram was 11 days (IQR 1−79). Initial biventricular repair was performed in 80%, whereas initial single ventricle palliation was performed in 9%. The remaining 11% of the infants underwent initial pulmonary artery banding. Of the babies who received pulmonary artery banding, 40% went on to biventricular repairs and 60% have had no further intervention as of March 2017. Overall, mortality at last follow-up was 9% (Table 1).

RESULTS

Echocardiographic Characteristics

All infants were confirmed to have complete AVSD at core laboratory review. Mean AVVI was 0.45 ± 0.09. Based on

Table 1. Demographic and Clinical Characteristics

	Study Population (N = 257)
Age at echo (d)	11 (1–79)
Weight (kg)	3.4 (2.9–3.9)
Male sex	48% (124)
Trisomy 21	80% (205)
Tetralogy of Fallot	6% (15)
Systemic venous anomalies	3% (7)
Additional ventricular septal defects	5% (12)
Surgical pathway	
Biventricular repair	80% (206)
Pulmonary artery banding	11% (27)
Single ventricle palliation	9% (22)
Overall mortality	9% (24)
No surgery	1% (2)
Pulmonary artery banding	2% (4)
Single ventricle palliation	2% (4)
Biventricular repair	5% (14)

AVVI, right dominance was present in 22%, left dominance in 4%, balanced ventricles in 52%, and could not be determined in the remaining 22% because of missing or inadequate imaging views. Mean RV/LV inflow angle was 89.7 ± 16.3°. Median LVII was 0.46 (IQR 0.38–0.54), median RVII was 0.46 (IQR 0.40–0.54), median LVII/RVII ratio was 1.04 (IQR 0.84–1.19), and median minor index was 1.0 (IQR 0.9–1.2).

Moderate or severe common AVV regurgitation was present in 26%. Papillary muscle abnormalities were found in 30%. All remaining echocardiographic measures are summarized in Table 2.

Correlation of Measures of Unbalance With One Another

The correlation of the 5 measures of unbalance with one another was assessed (Table 3 and Fig. 2). AVVI correlates moderately with LVII (0.30, $P < 0.0001$), the minor index (0.62, $P < 0.0001$), and LVII/RVII (0.39, $P < 0.0001$). There is no significant correlation between AVVI and the RV/LV inflow angle. The RV/LV inflow angle correlates moderately with LVII (0.31, $P < 0.0001$) and weakly with the LVII/RVII ratio (0.16 $P = 0.042$). The inflow angle does not significantly correlate with the minor index. LVII weakly correlates with the minor index (0.19, $P = 0.0062$). The LVII/RVII ratio moderately correlates with the minor index (0.36, $P < 0.0001$).

Correlation of Measures of Unbalance With Ventricular Size and Common AVV Size

The correlation of the 5 measures of unbalance with AVV annular diameters and with ventricular end-diastolic area (EDA) were evaluated (Table 4 and Fig. 2). AVVI moderately correlates with LAVV annular diameter (0.45, $P < 0.0001$) and weakly negatively correlates with right atrioventricular valve (RAVV) annular diameter (−0.23, $P = 0.0013$). AVVI moderately correlates with LV EDA (0.41, $P < 0.0001$) and weakly negatively correlates with RV EDA (−0.29, $P < 0.0001$). The RV/LV inflow angle moderately negatively correlates with LAVV and RAVV annular diameter (−0.31, $P < 0.0001$, −0.33, $P < 0.0001$, respectively). The RV/LV inflow angle does not significantly correlate with LV EDA or RV EDA. LVII has moderate negative correlation with RAVV annular diameter (−0.36, $P < 0.0001$) and does not significantly correlate with LAVV annular diameter or RV EDA.

Table 2. Echocardiographic Characteristics

	Study Population (N = 257)
Atrial and ventricular septae	
Malalignment of atrial and ventricular septae	15% (39)
Right (double outlet LA)	8% (3)
Left (double outlet LA)	90% (35)
Cannot determine	2% (1)
Measure of unbalance	
AVVI	0.45 ± 0.09
Right dominant (AVVI < 0.4)	22% (56)
Balanced (0.4 < AVVI < 0.6)	52% (134)
Left dominant (AVVI > 0.6)	4% (11)
Unable to be determined*	22% (56)
RV/LV inflow angle	89.7 ± 16.3°
LVII	0.46 (0.38–0.54)
RVII	0.46 (0.40–0.54)
LVII/RVII ratio	1.04 (0.84–1.19)
Minor index	1.0 (0.9–1.2)
Common AVV	
Common AVV regurgitation, moderate or severe	26% (67)
Papillary muscle abnormalities	30% (78)
Mural leaflet length (cm)	0.9 (0.7–1.0)
Mural leaflet width (cm)	1.4 (1.2–1.7)
Total common AVV diameter (cm)	2.2 (2.0–2.5)
Color inflow diameter at anulus Right AVV (cm)	1.5 (1.3–1.7)
Color inflow diameter at anulus Left AVV (cm)	1.3 (1.1–1.5)
Total area (cm^2)	2.8 (2.2–3.8)
Right AVV area (cm^2)	1.5 (1.2–2.1)
Left AVV area (cm^2)	1.2 (0.9–1.7)
Color inflow diameter at mid cavity, right AVV (cm)	0.8 ± 0.2
Color inflow diameter at mid cavity, left AVV (cm)	0.7 ± 0.2
Left AVV annular diameter (cm)	1.5 ± 0.3
Right AVV annular diameter (cm)	1.7 ± 0.4
Ventricles	
RV end diastolic area (cm^2)	4.1 (3.3–5.3)
RV end systolic area (cm^2)	2.7 (2.1–3.5)
RV width	1.6 ± 0.4
LV end diastolic area (cm^2)	4.0 (3.1–5.5)
LV end systolic area (cm^2)	2.2 (1.7–3.0)
LV width	1.6 ± 0.4

AVVI, atrioventricular valve index; LV, left ventricular; LVII, left ventricular inflow index; RV, right ventricular; RVII, right ventricular inflow index.
*Echocardiographic views or images used to determine AVVI not always available.

Table 3. Correlation of Measures of Unbalance With One Another

	Pearson Coefficient	*P* Value
AVVI		
RV/LV inflow angle	0.12	0.09
LVII	0.30	<0.0001
LVII/RVII ratio	0.39	<0.0001
Minor index	0.62	<0.0001
RV/LV inflow angle		
LVII	0.31	<0.0001
LVII/RVII ratio	0.16	0.042
Minor index	−0.06	0.32
LVII		
LVII/RVII ratio	0.68	<0.0001
Minor index	0.19	0.0062
LVII/RVII ratio		
Minor index	0.36	<0.0001

AVVI, atrioventricular valve index; LV, left ventricular; LVII, left ventricular inflow index; RV, right ventricular; RVII, right ventricular inflow index.

LVII has mild positive correlation with LV EDA (0.17, $P = 0.0151$). The LVII/RVII ratio weakly correlates with LAVV annular diameter (0.18, $P = 0.0158$) and is not significantly correlated with RAVV annular diameter. The LVII/RVII ratio moderately correlates with LV EDA (0.39, $P < 0.0001$) but is not significantly correlated with RV EDA. The minor index moderately correlates with LAVV annular diameter and LV EDA (0.49, $P < 0.0001$ and 0.42, $P < 0.0001$, respectively). The minor index moderately negatively correlates with RV EDA (−0.33, $P < 0.0001$) and does not significantly correlate with RAVV annular diameter. A subset analysis was repeated for these measurements indexed to body surface area with no change in the interpretation of the correlations (Supplement 2).

Correlation of Ventricular and Common AVV Sizes

The correlation of the measurements of the AVV anulus diameter and the ventricular EDA were quantified (Fig. 2). The LAVV anulus diameter correlates strongly with the LV end-diastolic volume (0.74, $P < 0.0001$), whereas the RAVV anulus

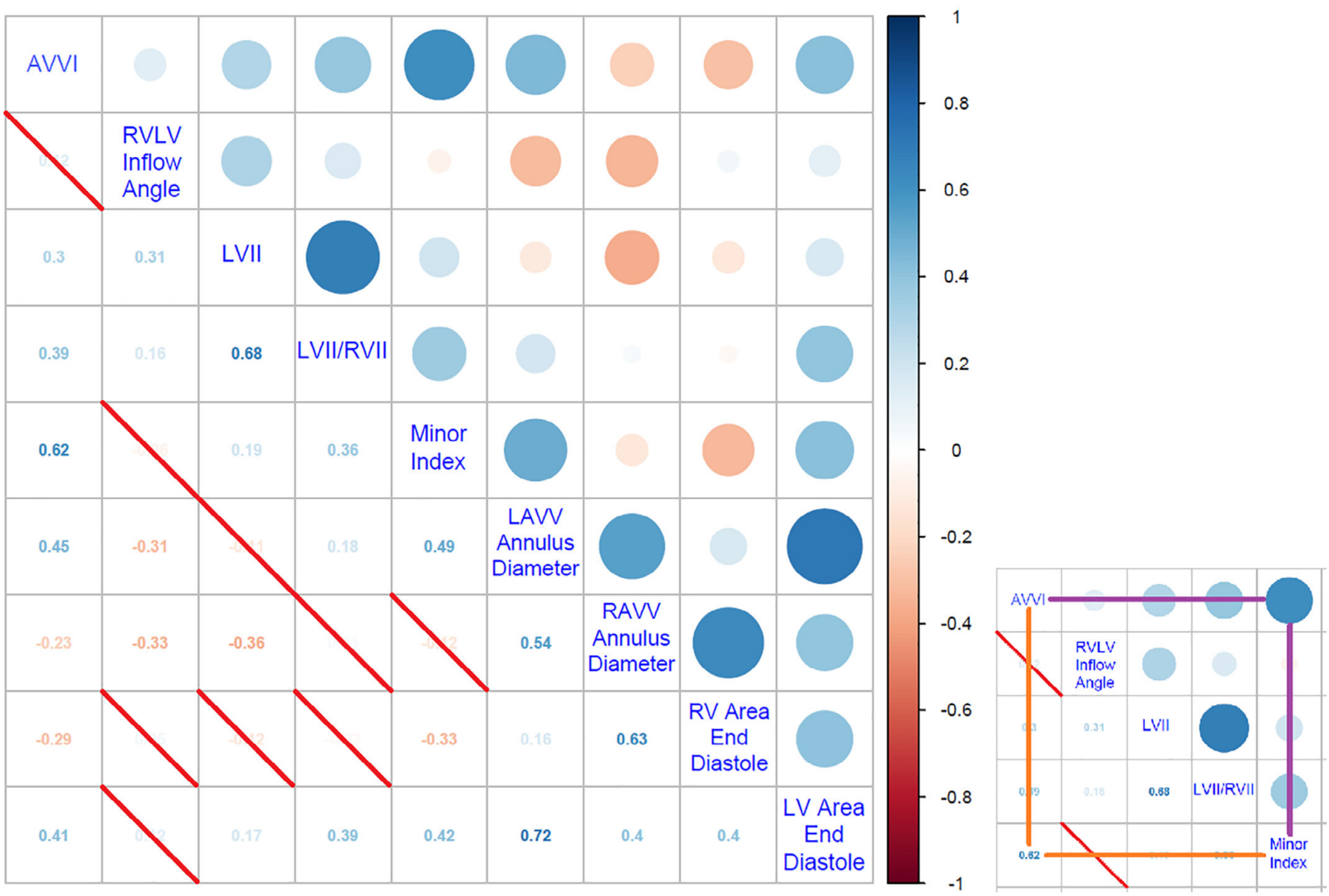

Figure 2. Correlations of AVVI, RV/LV inflow angle, LVII, LVII/RVII ratio, and minor index with each other and measures of AVV and ventricular size displayed in a correlogram. A correlogram is a graph of correlation matrix. Variables are listed on the diagonal axis. The circle at the intersection of 2 variables on the upper right half of the figure (inset: purple lines) is a graphical representation of the correlation coefficient (see legend at right) with color and size representing the degree of correlation. A larger circle size indicates a larger (further from zero) correlation. The box at the intersection of 2 variables on the lower left half of the figure (inset: orange lines) is the Pearson correlation coefficient, shaded and colored according to the direction and strength of the correlation. The boxes with red lines through them were not statistically significant to a *P* value of <0.05. (Color version of figure is available online.)

Table 4. Correlation of Measures of Unbalance With Atrioventricular Valve and Ventricular Sizes

	Pearson Coefficient	*P* Value
AVVI		
Left AVV annular diameter	0.45	<0.0001
Right AVV annular diameter	−0.23	0.0013
RV end diastolic area	−0.29	<0.0001
LV end diastolic area	0.41	<0.0001
RV/LV inflow angle		
Left AVV annular diameter	−0.31	<0.0001
Right AVV annular diameter	−0.33	<0.0001
RV end diastolic area	0.05	0.3993
LV end diastolic area	0.12	0.0619
LVII		
Left AVV annular diameter	−0.11	0.10
Right AVV annular diameter	−0.36	<0.0001
RV end diastolic area	−0.12	0.0829
LV end diastolic area	0.17	0.0151
LVII/RVII ratio		
Left AVV annular diameter	0.18	0.0158
Right AVV annular diameter	0.03	0.6652
RV end diastolic area	−0.03	0.6624
LV end diastolic area	0.39	<0.0001
Minor index		
Left AVV annular diameter	0.49	<0.0001
Right AVV annular diameter	−0.12	0.0571
RV end diastolic area	−0.33	<0.0001
LV end diastolic area	0.42	<0.0001

AVVI, atrioventricular valve index; LV, left ventricular; LVII, left ventricular inflow index; RV, right ventricular; RVII; right ventricular inflow index.

diameter correlates moderately with the RV end-diastolic volume (0.60, $P<0.0001$).

DISCUSSION

In our study, we found that there is only weak to moderate correlation among the 3 commonly used echocardiographic indices (AVVI, LVII, RV/LV angle) and among these indices and measurements of ventricular and AVV size. This dataset provides a robust source of precise morphologic information for echocardiographic analysis of this complex disease. The lack of strong correlation among the indices suggests that they cannot be used interchangeably. Future research in this disease cohort will analyze these data with regard to surgical triage and outcome to determine the importance of these indices.

Ventricular Size and Unbalance

Historically, assessment of ventricular size has been a very important determinant of surgical decision-making. Echocardiographic measurements including ventricular length, length-width ratios, and volumes have been advocated.[7] In babies with extreme ventricular hypoplasia, only single ventricle palliation is appropriate. However, for borderline cases, ventricular dimensions have not proven to be strong predictors for surgical decision-making and outcomes. This is partially related to the difficulty reliably assessing ventricular size by echocardiography, but also by the changes in ventricular size occurring after surgery. Other studies have shown that the LV cavity is compressed by the volume-loaded and pressure-loaded RV resulting in septal shifting into the LV cavity. Removal of the RV loading and increasing LV inflow results in shifting the ventricular septum to its normal position, which increases LV cavity size.[8,9] Ventricular growth may also occur after the procedure, which makes outcome predictions even more difficult.[10] In our data, the LV EDA correlated strongly with the left AVV anulus diameter and moderately with the AVVI, suggesting that inflow does impact the size of the LV and potentially the suitability for a biventricular repair.

Regarding the echocardiographic measurements reflecting LV and RV size, we opted to use simple 2-dimensional measurements including LV and RV width and area. These are direct measurements and do not depend on any geometrical assumptions. Previously, Grosse-Wortmann et al. have demonstrated that echocardiographic LV volumes underestimate volumes measured by magnetic resonance imaging.[8] Two-dimensional areas are the best estimations of ventricular volumes in the absence of a good volumetric technique, although these measurements vary with inotropes and volume status. We added the minor index (LV/RV width ratio) as another potential index for estimating size unbalance. When correlating this index with measurements reflecting valve and ventricular size, we found moderate strength correlations, suggesting this may be a useful index. In the subgroup analysis with body surface area-indexed size measurements, the interpretation of the correlation coefficients did not change (Supplement 2).

The Common AV Valve and Unbalance

Over the last few years, interest has shifted from ventricular size to morphologic and functional characteristics of the ventricular inflow. It has become increasingly clear that AVV size, morphology, and function are important factors in determining the feasibility of biventricular repair. Surgical division of the AVV must result in sufficiently large unobstructed inlets without clinically important regurgitation. A sufficient quantity of good-quality AVV tissue at the entrance of both ventricles is necessary. The AVVI represents the relative size of the LAVV to the total AVV area. A previous retrospective study from the CHSS demonstrated that when using AVVI as a predictor for BVR, patients with AVVI between 0.39 and 0.19 could be treated surgically with either UVR or BVR, suggesting that in this zone of uncertainty, other factors should be considered for decision-making.[2] In a subgroup analysis of patients with AVVI in this range, the interpretation of the correlation coefficients did not change (Supplement 3).

A previous study from CHSS using cluster analysis of retrospective data identified the RV/LV angle as strong determinant of unbalance.[4] A steeper angle was associated with unbalance. We adjusted the measurement of the angle by measuring it at end diastole, vs in early diastole in the previous study, which explains the difference in the mean angle values. The adjustment in timing better incorporates VSD size, as the angle will be more influenced by VSD size when the ventricles are full. In our data, the angle did not correlate with LV size, suggesting that it does not entirely capture the degree of inflow across the full spectrum of patients with AVSD if LV size is attributable to the degree of filling of the ventricle during fetal life. The change in the angle over the diastolic period may reflect a dynamic measure of ventricular inflow, which is an area for future study.

LVII reflects the relative size of the color Doppler inflow and represents valve orifice relative to anulus size, providing information on effective orifice. Interestingly, the values we observed are different from previously reported values, which suggests that this may not be the most reliable index to use in clinical decision-making.[3] Further, there is only weak to moderate correlation between AVVI and LVII, suggesting that there is a discrepancy between size and function of the valve that may be important to consider together. Assessing the LVII/RVII ratio does not seem to add additional useful information.

Limitations

The current study looked only at the correlations of different echocardiographic measurements and not at their association with outcomes. This will require further review of postoperative echocardiograms as well as a detailed analysis of surgical outcomes that goes beyond survival. Follow-up of this cohort is in progress. Another significant limitation was the lack of standardized views in the patients' echocardiograms. Fifty-six patients (22%) lacked a proper en face view of the common AVV, which is necessary for assessment of the AVVI. This reduced our sample size with regard to correlation analysis for that index and demonstrates the need for standardized diagnosis-based views in echocardiography. Finally, there is a potential for selection bias given that only 69% of the patients in the cohort had baseline echocardiograms available for evaluation by the ICL at the time of this publication.

CONCLUSIONS

This study provides a detailed morphometric analysis of a large cohort of patients with AVSD. Echocardiographic indices of unbalance poorly correlate with each other and also with measures of valve and ventricular size, indicating that none of the indices should be used in isolation. Standardization of echocardiographic views across centers is imperative for accurate assessment of unbalance in AVSD. This detailed dataset provides a solid basis for future outcome research on this topic, which will allow for continued investigation into the relative importance of these measures for surgical decision-making.

Acknowledgments

The authors thank Sally Cai, Brenda Chow, Kathryn Coulter, Annette Flynn, Kristina Kovach, Susan McIntyre, and Arti Singh from the CHSS Data Center. We acknowledge the invaluable contributions of the data coordinators at CHSS institutions. Finally, we are very grateful to the patients in the Complete AVSD cohort and their parents.

SUPPLEMENTARY MATERIAL

Supplementary materials associated with this article can be found in the online version at https://doi.org/10.1053/j.semtcvs.2018.02.004.

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