

# Comprehensive echocardiographic imaging of atrioventricular valves in children with atrioventricular septal defect: Accuracy of 2D and 3D imaging and reasons for disagreement

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## ABSTRACT

**Objective:** To compare the accuracy and reasons for disagreement of two-dimensional (2D) and three-dimensional (3D) echocardiography findings in the assessment of the atrioventricular valve complex in patients with atrioventricular septal defect.

**Methods:** A total of 20 children (mean age 8 months) with atrioventricular septal defect were enrolled prospectively into this study. The accuracy of and the reasons for disagreement in the assessment of the atrioventricular valve features were analyzed between 2D and 3D echocardiography and surgical findings.

**Results:** We found that in assessing the Rastelli type and the extension of the inferior leaflet into the right ventricle, 3D echocardiography was more accurate compared to 2D echocardiography. In all other features, 2D and 3D echocardiography showed similar accuracy. A significant reason for inaccuracy by both echo modalities was that the technique itself could not visualize the feature, although the image quality was considered to be adequate. In most cases, where it was not possible to visualize the atrioventricular feature by 2D, it was possible by 3D, and vice versa.

**Conclusion:** The accuracy of 2D and 3D echocardiography and understanding the potential reasons for disagreements in assessing the atrioventricular valve complex with 2D and 3D can guide the use of those two techniques when combining them in the clinical practice. (*Anatol J Cardiol* 2019; 21: 00-00)

**Keywords:** 3D echocardiography, 2D echocardiography, atrioventricular septal defect, accuracy, congenital heart disease

## Introduction

Precise visualization of the anatomical and functional abnormalities of the atrioventricular (AV) valves is a key determinant of appropriate preoperative preparation. Incomplete visualization of the valve structures may lead to impaired patient outcomes (1). Most children with atrioventricular septal defect (AVSD) undergo surgery of the AV-valve. The evaluation of the AV valves in children with AVSD is based on the echocardiography, which is currently the only imaging modality that can visualize valves in real time.

It has been suggested that when using two-dimensional (2D) echocardiography, the need of subjective geometric assumptions limits the accuracy and increases the variability of 2D echocardiography because of the complex nature of the AV valve (2). The complex interactions of the AV-valve structures

are often difficult to grasp by the 2D echocardiography only (2). Three-dimensional (3D) echocardiography can on the other hand miss smaller anatomic structures, mostly because of the lower resolution (2), but it has shown the potential to give a comprehensive insight into the anatomical relationships between various structures in the heart (3-7).

There are limited data on the accuracy of 3D echocardiography in imaging of the AV valves in patients with AVSD. It was shown that the attachments of the superior and inferior bridging leaflet of the AV-valve to the septum can be precisely visualized with 3D echocardiography (8). Other features, such as the size, defects, and function of the leaflets, apposition zone between the leaflets, quality of coaptation between the leaflets, and anatomy of the papillary muscles were not studied. It was suggested that the lower resolution of the 3D images compared to the 2D images may be challenging and may influence the provided information

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(8). Causes of possible misleading information, such as misunderstandings between the interpreter of the images and surgeon and/or types of artifacts were not accounted for nor evaluated.

Understanding how 2D and 3D echocardiography can combine each other for the most accurate visualization of the AV valves in children with AVSD is necessary for the patient management and further development of this technique such as in computer modeling and simulators (9). To understand where the inaccuracies occur and to differentiate between those that are technique dependent (such as artifacts, high noise, and a restricted field of view) and those that are technique independent (such as subjective assumptions or subjective interpretations), leads to the opportunities to develop the method based on the understanding of its strengths and limitations (10).

The aim was to compare the accuracy of the 2D and 3D echocardiography in the assessment of the AV-valve complex in children with AVSD and to evaluate the reasons for inaccuracies. We aim to define those specific AV-valve complex features where 3D echocardiography will be more accurate compared to 2D, and vice versa.

## Methods

The study was conducted in compliance with the Declaration of Helsinki, and the research protocol was approved by the locally appointed Ethics Committee. Informed consent was obtained from the legally authorized representative of all patients.

### Study population

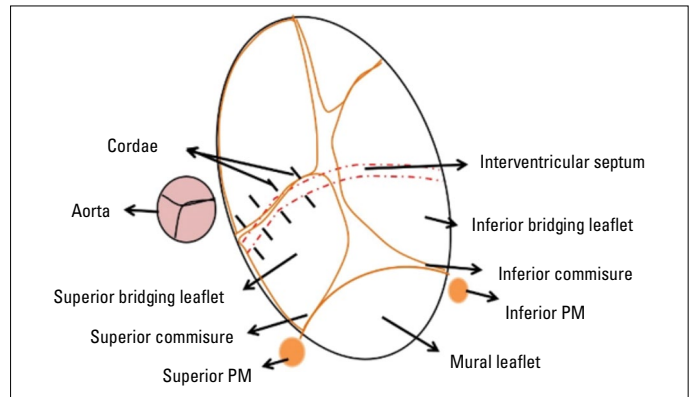
The study was designed as a prospective single-center study. We consecutively enrolled 20 children prior to AVSD surgery at the time of their routine transthoracic echocardiography (TTE). Specific criteria for inclusion into the study were diagnosis of AVSD, the age 0–18 years, and both 2D and 3D ultrasound available before surgery. The exclusion criterion was previous AV-valve surgery. In each patient, 26 AV-valve features were involved in the assessment, as described further in text.

### Definitions

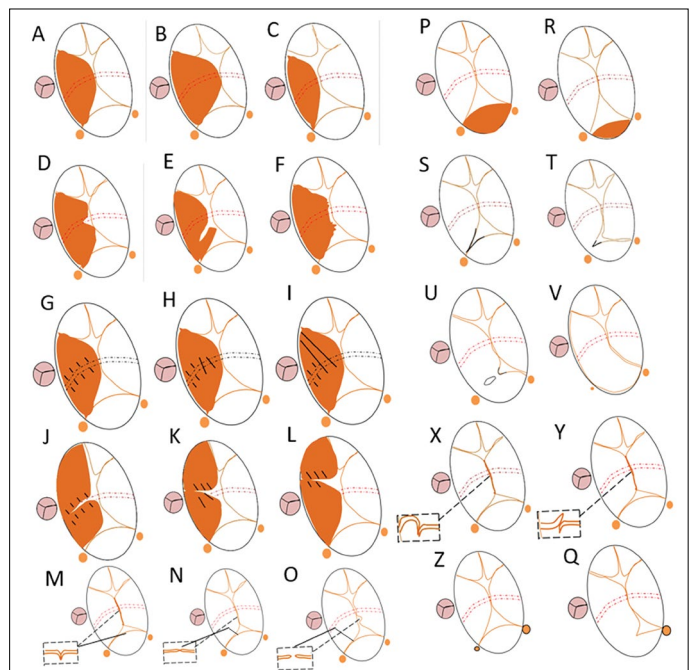
Figure 1 provides a schematic picture of the AV-valve (11). Abnormal features of the AV-valve complex were defined as the features that had impaired morphology or function. Anatomical and functional characteristics of the AV-valve, including leaflets, commissures, chordae, and/or papillary muscles, were involved in the assessment. All assessed features are summarized and presented in Figure 2 and Table 1.

Reasons for disagreement were defined as the following:

- Being unable to visualize a particular feature: The technique itself could not visualize the feature although the image quality was considered to be adequate.
- Inadequate image quality: Impossible to recognize the feature because of the low image quality.



**Figure 1.** Schematic image of the AV-valve and its components



**Figure 2.** Anatomical and functional characteristics of the AV-valve complex features, including leaflets, commissures, chordae, and/or papillary muscles

- Artifacts not accounted for: Artifacts that were not recognized before surgery and were understood after re-analysis.
- Different description of pathology although the technique showed the feature adequately: Difference in the judgment itself, not the technique.

### 2D imaging and analysis of the AV-valve complex

A commercially available ultrasound system (Philips Medical Systems, Andover, MA, USA) with a 7 MHz and 5 MHz probe was used for the TTE imaging of the AV-valve features. One of two experienced TTE operators (>1000 procedures each) obtained standard views from subcostal, parasternal, and apical windows for the 2D assessment of all specific features of the AV-valve complex (12, 13). The AV-valve complex features were analyzed before the surgery, directly after the image acquisition by the same operator who acquired the images.

**Table 1. Definitions and details of assessment of anatomical and functional characteristics of the AV-valve complex features**

Variations/Abnormality	Specification how measured/Assessed	Depicted in figure 2 as the letter
<b>Superior/inferior bridging leaflet</b>		
Size: normal, smaller, bigger	Eyeballing assessed ratio between SBL and IBL (normal: ratio 1; lower SBL: SBL-to-IBL ratio<1; greater SBL: SBL-to-IBL ratio>1)	A. Normal SBL B: Bigger SBL C: Smaller SBL
Missing material (yes/no)	Local defect at the edge of the leaflet that is bigger than the erosion and appears as a "hole"	D: Missing material of IBL
Cleft in the leaflet (yes/no)	Division of the one of the leaflets appearing as a slit-like hole	E: Cleft in SBL
Erosions of the edges (yes/no)	Abrasion of the edges of the valve tissue echocardiographically appearing as fine defects	F: Erosions of SBL
Rastelli type A/B/C	Assessed based on the insertion of the superior bridging leaflet to the right ventricle, as well as its grade of overhanging to the right ventricle (14)	J: Type A* K: Type B* L: Type C*
<b>Chordae of superior/inferior commissure</b>		
Straddling of chordae	Chordae crossing the ventricular septum from one side of the ventricular septum to the other (15)	G: No straddling H, I: Straddling of the SBL chordae
Prolapse or flail (yes/no)	Prolapse: Extension of the leaflet more than 2 mm above the plane of the AV annulus during ventricular systole. Flail: leaflet tip turning outward, becoming concave toward the left atrium	X: Prolapse of SBL Y: Flail of SBL
<b>Mural leaflet</b>		
Size: normal, smaller, bigger	Eyeballing assessed ratio between the mural leaflet and the left-sided part of the SBL and IBL	P: Normal mural leaflet R: Smaller mural leaflet
Cleft/defect in the leaflet (yes/no)	Division of the one of the leaflets appearing as a slit-like hole	
<b>Superior/inferior commissure</b>		
Size: normal, shorter, missing	Eyeballing assessed ratio between sup. commissure and inf. commissure	S: Normal T: Shorter V: Missing
Double orifice	Appearing as a tissue connecting the anterior and posterior leaflets at the leaflet edge level, causing two orifices opening into the left ventricle	U: Double orifice
<b>Apposition zone (16)</b>		
Size of area: smaller area, diastasis	Eyeballing assessment of the space between the facing surfaces of the left ventricular components of the bridging leaflets of the common AV-valve	Dashed line M: Normal N: Smaller O: Diastasis
<b>Coaptation between mural and bridging leaflet</b>		
Size of area: smaller area, diastasis	Eyeballing assessment of the space between the facing surfaces of the mural leaflet and respective bridging leaflet	Full line M: Normal N: Smaller O: Diastasis
<b>Superior/inferior papillary muscle</b>		
Underdeveloped, missing	Eyeballing assessment of the ratio of the size between papillary muscles	Z: Underdeveloped SPM Q: Missing SPM

**Table 1. Cont.**

Variations/Abnormality	Specification how measured/Assessed	Depicted in figure 2 as the letter
Size: normal, small	<p><b>Opening of the left-sided AV-valve</b></p> <p>Assessed by measuring the area of the valve opening as it will appear after surgery (after separation of the common valve in the left and right orifice) and then categorized into normal/small by using z scoring (normal: z score&gt;2, small: z score&lt;2)**</p>	
<p>*Rastelli type A: The SBL does not overhang the right ventricle, and attachments of the left-sided part are on the left side of the ventricular septum. Rastelli type B: The SBL overhangs partly to the right ventricle with some attachments of the chordae in the right ventricle. Rastelli type C: The SBL overhangs to the right ventricle, and its attachments are in the right ventricle only. **The annular levels of the valve were identified from short axis cut planes, and their areas were measured by planimetry. The diameters of mitral annulus were measured by 2 DE from the apical and parasternal long-axis views (17). SBL - superior bridging leaflet, IBL - inferior bridging leaflet</p>		

### 3D imaging of the AV-valve complex

3D images of the atrioventricular valve complex were acquired by one of two experienced TTE operators (>500 procedures each) using Philips iE33 (Philips Medical Systems, Andover, MA, USA). An X7-2 MHz probe was used in all patients for the 3D-TTE imaging of the AV-valve features. The most appropriate views (subcostal, parasternal, and apical) were chosen to acquire 3D real time images based on the bedside evaluation of the best image quality. The QLab software (Philips Medical Systems) was then used for the offline analysis. Reconstruction of the AV-valve complex was carried out by an experienced operator directly after the image acquisition, before the AV-valve surgery. The surgical en-face view of the valve, view of the AV-valve complex from the apex in the 3D mode, as well as cropping of the AV-valve complex using the multiplane reconstruction technique were used to analyze the specific AV-valve complex features.

### Surgical assessment of the AV-valve complex

The valve was assessed by the surgeon on the non-beating heart. The anatomical features of the valves and subvalvular complex were assessed under the surgery, and a drawing of the anatomy of the AV-valve was made after the surgery. The site of regurgitation was performed using saline injection. The accuracy of the echo assessment was determined using the surgical description as the reference method.

### Image quality quantification

Both 2D and 3D images were assessed regarding the image quality for all AV-valve complex features. Images were scored as 0, if the image quality was inadequate, and as 1, if the image quality was adequate for each particular AV-valve complex feature.

### Accuracy of morphological and functional assessment

Anatomical and functional characteristics of the AV-valve were analyzed by two independent observers who agreed on expert consensus before the surgery. The receiver (surgeon) documented AV-valve complex features as seen during the surgery. The surgeon was aware of the 2D findings and 3D findings for patient-safety purposes. A 2-point scoring protocol was used

to evaluate the accuracy of the 2D and 3D-echocardiography in the assessment of the AV-valve complex features. Specifically, features were graded 0 if the preoperative description disagreed with the surgical findings. Features were graded 1 if the preoperative description agreed with the surgical findings.

### Evaluation of the reasons for disagreement

Both the cardiologist and the surgeon re-evaluated 2D images and 3D images, retrospectively, from the database (Xcellera) in cases where disagreement was observed. A three-letter scoring system was used to evaluate the reason for disagreement. Specifically, the feature was scored as (a) if the reason for disagreement was because the technique was unable to visualize that particular feature; (b) if the reason was an inadequate image quality or artifacts; or (c) if the opinion was different although the technique could show the feature adequately and during the reassessment, an agreement was reached. In cases where several factors were causing the disagreement, all factors for disagreements were included in the results, respectively.

### Inter-observer variability in 2D and 3D assessment

To calculate the inter-observer variability in 2D and 3D assessment, a second operator analyzed the reconstructions in all patients.

### Statistical analysis

Statistical analysis was performed using the Statistical Package for the Social Sciences Version 10.1.0 for Windows. Continuous data are presented as the mean value±standard deviation (SD). Nominal data were presented as percentages. The chi-squared test was used to compare differences between groups. An inter-observer correlation was made using the Pearson r correlation. A probability of <0.05 was considered significant.

## Results

### Patient characteristics

Twenty patients were included to the study, 11 patients with atrial and ventricular shunting and nine with only atrial shunt-

ing. Eight patients were male with the mean age 8 months (range 3–72 months) and mean weight 5.6 kg (range 3.5–21 kg). Nine patients (45%) had Down’s syndrome, one (5%) had Noonan’s syndrome. Twenty-five percent of patients had associated abnormalities such as persisting arterial duct (2 patients), ostium secundum atrial septal defect (2 patients), and pulmonary valve stenosis (1 patient).

### Baseline AV-valve characteristics

Twenty-one features of the AV-valve were described in each patient, as summarized in Figure 2 and Table 1. In total, 520 AV-valve features were described in all 20 patients. Abnormalities were found in 57 (11%) out of 520 AV-valve features. Most often, abnormalities of the superior commissure (double orifice or

short in 45% patients) and coaptation abnormalities between the bridging leaflets (diastasis or short coaptation in 45% patients) were found. Left-sided AV-valve regurgitation was present in 85% of patients (mild: 75%, moderate 25%, severe 0%).

### Technical details

For 2D imaging, a probe S8-3 was used in all 20 patients. The frame rate of the 2D images was 79±17 mHz (mean±SD). For 3D imaging, a probe X7-1 was used in all patients, in one patient, in addition to the X7-1 probe, an X5-1 probe was used. The frame rate of the 3D images was 32±2 mHz (mean±SD). The heart rate of the patients was 132±8 bpm and had normal distribution. The images were acquired when patients were calm (feed and sleep method). None got sedation nor anesthesia.

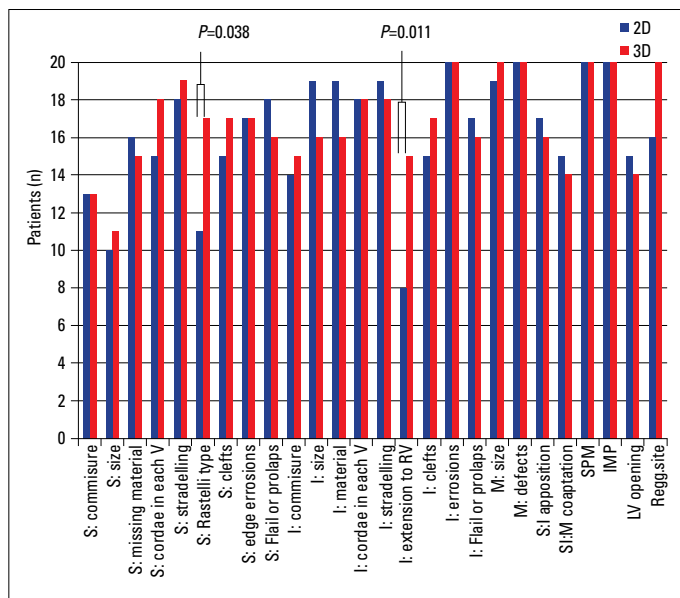
**Table 2. Comparison of accuracy in the assessment of the AV-valve features between 2D and 3D echocardiography**

Valve feature name (assessed in each of 20 patients)	2D Agreement	3D agreement	P-value*
S: Commissure	13	13	1.000
S: Leaflet size	10	11	0.750
S: Missing material	16	15	0.700
S: Cordae attachments	14	18	0.110
S: Cordae straddling	18	19	0.550
Rastelli A/B/C	11	17	<b>0.038</b>
S: Clefts	13	17	0.140
S: Edge erosions	17	17	1.000
S: Flail of prolaps	18	16	0.380
I: Commissure	14	15	0.720
I: Leaflet size	18	15	0.210
I: Missing material	18	18	1.000
I: Cordae attachments	18	18	1.000
I: Cordae straddling	19	18	0.550
I: Extension to the RV	7	15	<b>0.011</b>
I: Clefts	15	17	0.430
I: Edge erosions	20	18	0.150
I: Flail of prolaps	17	16	0.680
M: Size	18	20	0.150
M: Defects	17	18	0.630
Apposition between S and I	17	16	0.680
Coaptation between SI and M	15	14	0.720
SPM	20	20	1.000
IPM	19	19	1.000
LV opening	15	14	0.720
Regurgitation site	16	18	0.310
All	413 (79%)	432 (83%)	0.130

\*by Chi-square

S - superior bridging leaflet; I - inferior bridging leaflet; RV - right ventricle; M - mural leaflet; SPM - superior papillary muscle; IPM - inferior papillary muscle; LV - left ventricle





**Figure 3.** Comparison of accuracy in the assessment of the AV-valve complex features of 2D (blue columns) and 3D (red columns). S - superior; I - inferior; M - mural; V - ventricle; RV - right ventricle; LV - left ventricle; SPM - superior papillary muscle; IPM - inferior papillary muscle

### 2D imaging of the AV-valve complex

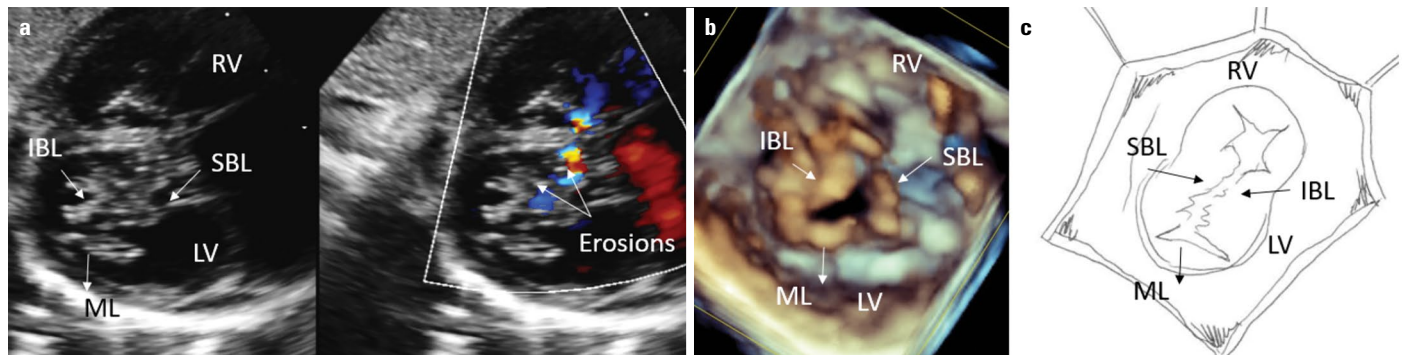
The accuracy of the 2D echocardiography is depicted in Figure 3 (blue columns) and the first column in Table 2. A 2D ultrasound agreed with the surgical description in 413/520 (79%) AV-valve features. Most often, the disagreement was related to the assessment of the Rastelli type (disagreement in 40% patients) and in the assessment of the superior commissure: inadequate recognition of the double orifice type of commissure (5% of patients) or under/overestimation of the commissure size (30% patients).

### 3D imaging of the AV-valve complex

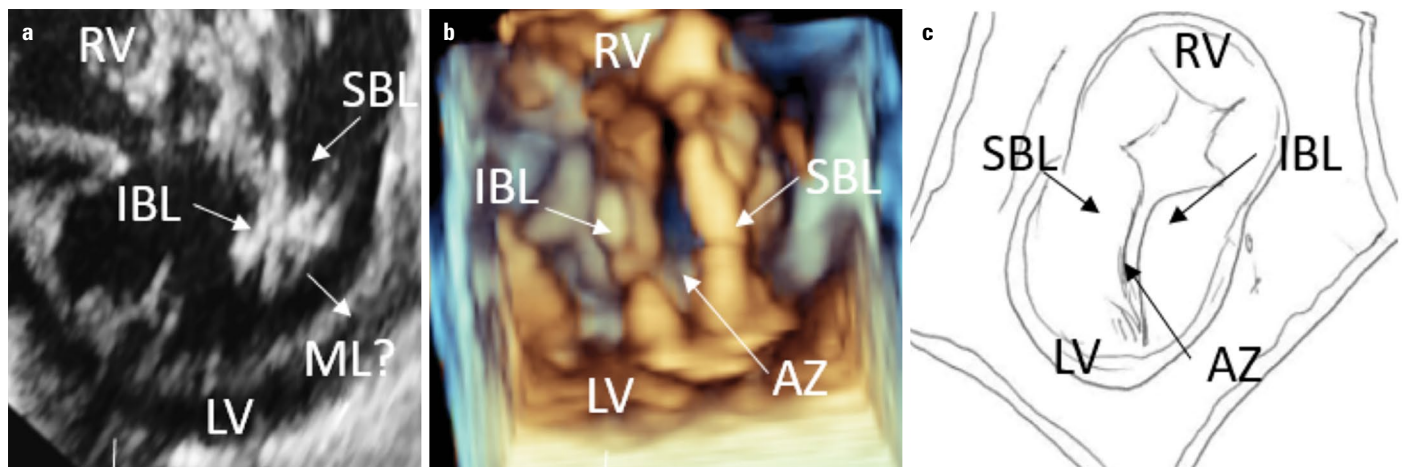
The accuracy of the 3D echocardiography is depicted in Figure 3 (red columns) and the second column in Table 2. A 3D ultrasound agreed with the surgical description in 432/520 (83%) AV-valve features. The disagreement was most often related to the evaluation of the apposition zone: the presence of diastasis/smaller area (disagreement in 30% patients).

### Comparison of the accuracy between 2D and 3D echocardiography

Figure 3 and Table 2 depict the comparison of accuracy in the assessment of the AV-valve features between 2D and 3D. 2D and 3D



**Figure 4.** (a) 2D echocardiography (without and with color Doppler) of the AV-valve in the subcostal short axis view. (b) 3D echocardiography of the AV-valve in the en-face view from the ventricle. (c) Schematic picture of the surgical view. LV - left ventricle; RV - right ventricle; IBL - inferior bridging leaflet; SBL - superior bridging leaflet; AZ - apposition zone



**Figure 5.** (a) 2D echocardiography of the AV-valve in the subcostal short axis view with color-compare image (b) 3D echocardiography of the AV-valve in the en-face view from the ventricle. (c) Schematic picture of the surgical view. LV - left ventricle; RV - right ventricle; IBL - inferior bridging leaflet; SBL - superior bridging leaflet

echocardiography had a similar accuracy in the imaging of almost all AV-valve features, in addition to the assessment of the Rastelli type, where 3D echocardiography was more accurate compared to 2D echocardiography ( $p=0.038$ ) and in the assessment of the extension of the inferior leaflet into the right ventricle ( $p=0.011$ ).

In almost 48% of cases where the 2D was accurate, the 3D was inaccurate, and vice versa. Figure 4 provides an example of the AV-valve when the 2D echocardiography and 3D echocardiography findings disagreed in the assessment of the number of commissures. As it may be observed, 3D echocardiography shows the presence of one commissure only. Figure 5 provides an example when 2D and 3D echocardiography disagreed when assessing the valve erosions. 2D echocardiography shows the presence of erosions in the superior and inferior bridging leaflet.

### Evaluation of the reason for disagreement

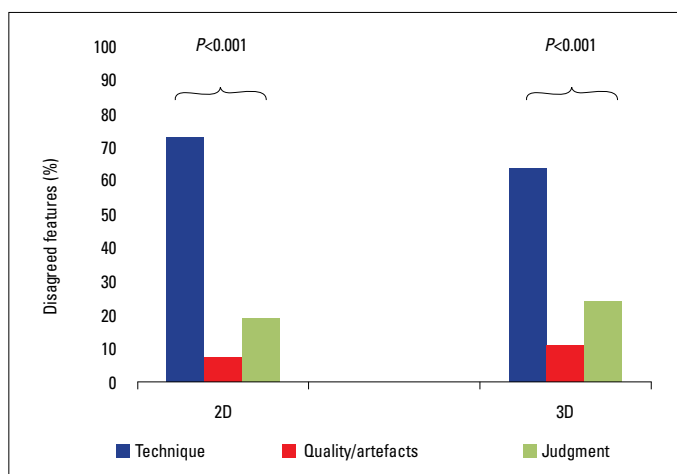
Figure 6 shows the comparison of different reasons for disagreement in each modality. It demonstrates that a significant reason for misdiagnosis by both 2D and 3D is the impossibility of the technique to visualize that particular feature. The judgment of the observer or an inadequate image quality or artifacts was not significant reasons for disagreement.

### Image quality quantification

The image quality was similar for 2D and 3D echocardiography. The number of AV-valve complex features that were visualized with low quality was 27% in 2D and 30% in 3D. There was no significant difference in the image quality of any specific feature ( $p=0.632$ ).

### Inter-observer agreement

3D studies were evaluated by another operator with experience in both 2D and 3D imaging. The opinions of the 2 observers were highly correlated ( $r=0.640$ ;  $p=0.002$ ).



**Figure 6.** Comparison of different reasons for disagreement in each modality

## Discussion

This study evaluated the accuracy of both 2D and 3D imaging in specified features of the AV-valve complex in children with AVSD. To the best of our knowledge, this is the first study that provides a comprehensive evaluation of specified AV-valve features in this patient group by both 3D and 2D echocardiography and that assessed reasons for disagreements between the ultrasound methods.

A previous study showed the feasibility of 3D echocardiography to assess the Rastelli classification and the morphology of the inferior bridging leaflet, as classified by its attachment to the crest of the ventricular septum.<sup>8</sup> Other features of the AV-valve complex, however, were not assessed. Since precise visualization of the whole AV-valve complex is a key determinant of appropriate preoperative preparation, and its underestimation may lead to impaired patient outcomes, it is of a key importance to accurately visualize all AV-valve complex features (1).

Our study shows that, in most cases where it was not possible to visualize the particular feature by 2D, it was possible by 3D, and vice versa. This information is of key importance when applying both 2D and 3D techniques in the clinical use. It may be that knowledge of specific reasons for inaccuracies in assessing the AV-valve complex with 2D and 3D imaging can guide the use of those two techniques when combining them in the clinical practice and also in the valve modeling and simulations.

### Study limitations

Patients were evaluated by the same operator, often first with 2D and then with 3D, so the methods were not blinded to each other. Both modalities were however biased at the same time, which does not favor one of them only. Moreover, reassessment of the images was done by the surgeon after the procedure, so the subjective interpretations and influences from another modality that might have played a role before the surgery were eliminated. The surgical assessment was considered as a reference for all features, although there are features that were better visualized with echocardiography (such as the grade and location of the insufficiency). The number of patients in the study was limited to 20; however, it is not the actual number of patients, but rather 26 AV-valve features in each patient that were studied, which makes a total number of examined AV-valve features 520.

## Conclusion

In conclusion, since 3D echocardiography is more accurate in the assessment of some features compared to 2D, and since in most cases where it was not possible to visualize the particular feature by 3D, it was possible to do so by 2D, and vice versa, we conclude that knowledge of specific reasons for inaccuracies in assessing the AV-valve complex with 2D and 3D echocardiography can guide the use of those two techniques when combining them in the clinical practice.

**Conflict of interest:** None declared.

**Peer-review:** Externally peer-reviewed.

**Authorship contributions:** Concept – N.H., T.H., T.M., J.R.J.; Design – N.H., T.H., T.M., J.R.J.; Supervision – N.H.; Fundings – N.H.; Materials – N.H., J.R.J.; Data collection &/or processing – N.H., T.H., T.M., P.W., J.R.J.; Analysis &/or interpretation – N.H., J.R.J., C.M.; Literature search – N.H., T.H., T.M., P.W., J.R.J., C.M.; Writing – N.H., T.H., T.M., P.W., J.R.J., C.M.; Critical review – N.H., T.H., T.M., P.W., J.R.J., C.M.

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