Double-Orifice Left Atrioventricular Valve: The Case for Preoperative Three-Dimensional Echocardiography

Felina Mille, MD, Michelle Kaplinski, MD, Yan Wang, RDCS, Hannah H. Nam, BA, David J. Goldberg, MD, and Matthew A. Jolley, MD, *Philadelphia, Pennsylvania*

INTRODUCTION

We describe the case of a 4-month-old girl with a transitional common atrioventricular (AV) canal who underwent initial surgical repair at 3 months of age. A double-orifice left AV valve was noted on visual inspection by the surgeon. This was previously undiagnosed by routine two-dimensional (2D) echocardiography. The patient was left with moderate left AV valve regurgitation and was discharged home on diuretic therapy. She presented again within a few weeks in low cardiac output in the setting of severe residual left AV valve regurgitation. She was brought back to the operating room, where preoperative three-dimensional (3D) transthoracic echocardiography and pre- and intraoperative transesophageal echocardiography were used to delineate the mechanism of regurgitation through the double-orifice left AV valve and inform the surgical approach. Preoperative 3D transthoracic echocardiography may be of particular benefit in diagnosing double-orifice valves and aid in conveying the anatomy to the cardiothoracic surgeon.

CASE PRESENTATION

Our patient was a 4-month-old girl with trisomy 21 and a transitional AV canal defect. She was diagnosed prenatally. Postnatal echocardiography confirmed the diagnosis and showed trace AV valve regurgitation, two additional small muscular ventricular septal defects, and mild aortic arch hypoplasia. She was observed in the cardiac intensive care unit postnatally, did not develop evidence of aortic coarctation, and was discharged home on no medications. She underwent elective pericardial patch closure of the primum atrial septal defect, suture closure of the muscular ventricular septal defects, and suture closure of a left AV valve cleft at 3 months of age. A double-orifice left AV valve was noted intraoperatively on visual inspection by the surgeon but had not been appreciated on preoperative 2D imaging. Postoperative 2D echocardiography showed moderate left AV valve regurgitation with two jets that were not well characterized.

The patient was discharged on enteral furosemide and presented again 1 month later with lethargy, pallor, poor feeding, and tachypnea. Echocardiography showed severe left AV valve regurgitation (Figure 1, Video 1) with elevated, systemic right ventricular systolic pressure. She was stabilized on milrinone infusion, and referred for surgical left AV valvuloplasty or valve replacement. Further 2D transthoracic imaging was performed, but the full anatomy of the valve and location of regurgitation were difficult to determine.

In the operating room, preoperative 2D transesophageal imaging under anesthesia suggested that the valve anatomy was complex. Valve insufficiency was thought to originate from two sites, but 2D imaging did not clearly convey the anatomic location of the regurgitation or which valve segments were involved (Figure 2). Transthoracic 3D imaging was then used to better delineate valvular anatomy and the cause of the severe left AV regurgitation. On 2D and 3D imaging, a double-orifice valve was clearly visualized with a prominent tissue bridge between the mural leaflet and inferior bridging leaflet. There were two well-developed papillary muscles, which were rotated counterclockwise. The more anterolateral papillary muscle appeared to be located between the anterior bridging and mural leaflets, and the more posteromedial papillary muscles appeared to supply the inferior bridging and mural leaflets. There was the suggestion of an additional accessory papillary muscle, but this was not well visualized and may represent accessory valve tissue. There was severe regurgitation, mainly through the superior orifice, suggesting dehiscence of the previously closed "cleft" between the superior and inferior bridging leaflets in the superior orifice of this this highly atypical valve (Figures 3-6, Videos 2 and 3). Intraoperative visualization confirmed dehiscence of the left AV valve cleft within the superior orifice. This defect was suture-closed completely, and a cold saline test suggested that valve competence was improved. The patient was weaned from bypass, and unfortunately, transesophageal echocardiography showed continued severe regurgitation. Cardiopulmonary bypass was again initiated, and two pledgeted annuloplasty sutures were placed at a commissure near the free wall of the left ventricle. Postoperative echocardiography showed mild to moderate left AV valve regurgitation and mild stenosis with a 4 mm Hg inflow gradient, so valve replacement was not pursued. Unfortunately, the patient continued to have severe left AV valve regurgitation (Figure 7) and respiratory failure postoperatively and ultimately returned to the operating room for prosthetic left AV valve replacement 1 month later.

DISCUSSION

Double-orifice left AV valve was thought to be a rare finding in common AV valve defects. However, more recent case series have shown

From the Division of Cardiac Critical Care Medicine, Department of Anesthesiology and Critical Care Medicine (F.M.), Division of Pediatric Cardiology, Department of Pediatrics (M.K., Y.W., D.J.G., M.A.J.), and Department of Anesthesiology and Critical Care Medicine (H.H.N., M.A.J.), The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania.

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VIDEO HIGHLIGHTS

Video 1: Apical four-chamber sweep with color obtained upon readmission shows severe left AV valve regurgitation through three jets. The mechanism of the regurgitation is not clear.

Video 2: Subcostal frontal view in color-compare showing the double-orifice left AV valve. The larger, more superior orifice represents the residual left AV valve "cleft" between the superior and inferior bridging leaflets, with visible echo-bright suture material.

Video 3: Three-dimensional view of the left AV valve from the ventricle shows the two valve orifices. There is a tissue bridge between the interventricular septum and the intersection of the inferior and mural leaflets. The small mural leaflet is partially obscured beneath the tissue bridge.

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that it occurs in as many as 8% of AV canal lesions and is a risk factor for reoperation in these patients.[1-3] Two-dimensional echocardiography is frequently insufficient to diagnose or fully profile double-orifice AV valve and may overestimate the number of regurgitant jets.[4,5] Three-dimensional echocardiography has the advantage of showing both orifices and their relationship to each other in one image, even if they are not in the same plane. In adults, 3D images are obtained using breath-held transesophageal echocardiography, but there are currently no 3D transesophageal probes for small children. Pediatric-sized transthoracic probes are available, but it is often difficult to obtain 3D images with high temporal and spatial resolution in unsedated or even sedated but free-breathing children. Single-beat acquisitions lack artifacts but are constrained by relatively low frame rates, field of view, or resolution. Attempts at multibeat electrocardiographically gated acquisitions are often confounded by significant stitch artifact due to respiratory motion. As such, the immediate preoperative period, when the patient has been intubated and paralyzed in preparation, provides a unique opportunity to gather gated, breath-held acquisitions with optimal temporal and spatial resolution.

We described a case in which 3D echocardiography was used to delineate left AV valve anatomy and guide valvuloplasty in a patient with a common AV canal defect and double-orifice left AV valve. Durable repair of AV canal defect remains challenging, with up to 30% of children requiring reintervention for left AV valve regurgitation.[6] Double-orifice left AV valve represents a more complex variant, and our experience suggests more challenging repairs and worse outcomes. Although double-orifice left AV valve may be recognized using careful 2D imaging (Figure 3), we suggest that preoperative transthoracic 3D valve imaging be considered for all patients with AV canal defects to better understand the 3D anatomy of these complex valves. Currently, most children undergoing initial repair will be too small (<20 kg) for a 3D transesophageal examination, but breath-held transthoracic imaging before repair can result in informative images. Improved preoperative imaging will allow improved understanding and classification of common AV canal defect, and complex variants such as these, which may require unique and individualized repair strategies. Improved visualization could allow collaborative discussion among surgeons to harness the experience of many rather than an individual surgeon visualizing the anatomy in a flaccid heart at the time of repair. It will also allow the application of quantification tools to move toward the identification of 3D structural correlates of valve dysfunction and difficult repair. It remains to be seen whether superior imaging will positively affect surgical outcomes, but without further image-informed delineation of anatomic variants, and the creation and description of successful repairs, it is unlikely that significant progress will be made in this challenging population.

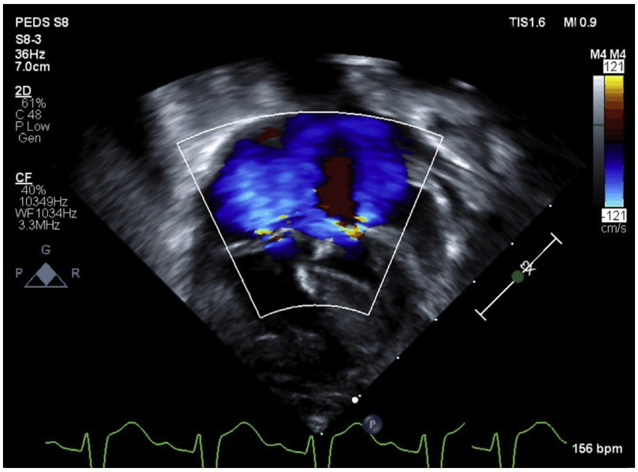

Figure 1 Apical four-chamber sweep with color obtained upon readmission shows severe left AV valve regurgitation through three jets. The mechanism of the regurgitation is not clear (Video 1).

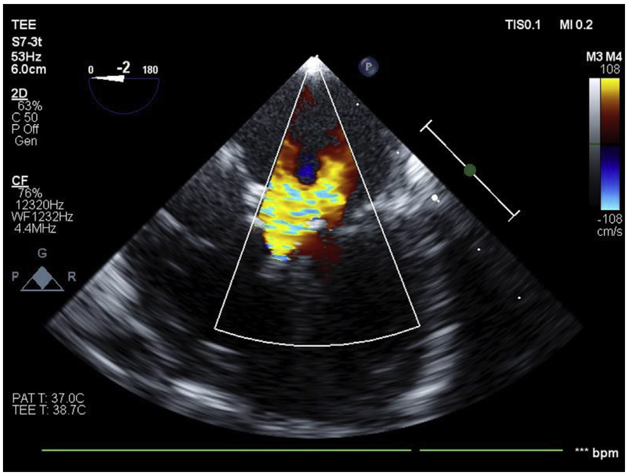

Figure 2 Four-chamber view from preoperative transesophageal echocardiography showing severe left AV valve regurgitation through multiple jets. The mechanism of the regurgitation is not clear.

There are limitations to transthoracic imaging, however. Three-dimensional transthoracic imaging is not an option once the chest is open, which, in this case, actually made describing the location of the initial postoperative residual defect to the surgeon more difficult. A pediatric-sized 3D transesophageal probe would be ideal for this situation but is not yet commercially available. Three-dimensional epicardial imaging has been reported to provide more complete information and have a lower false-negative rate when used during aortic valve repair.[7] This modality has been applied routinely during mitral valve surgery at some large surgical centers, but the optimal beam

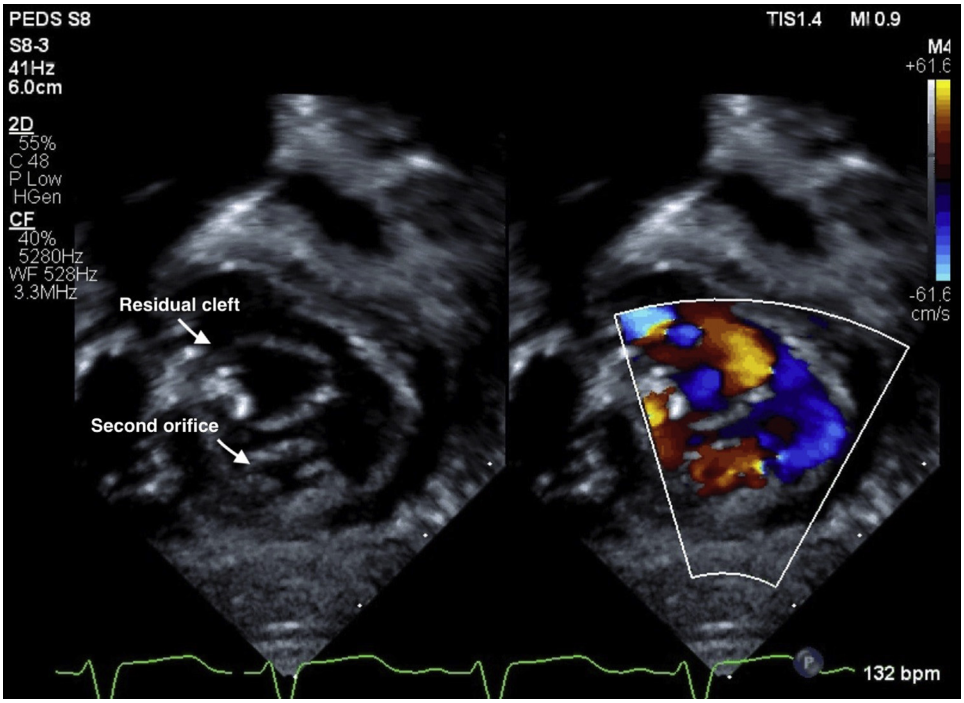

Figure 3 Subcostal frontal view in color-compare showing the double-orifice left AV valve. The larger, more superior orifice represents the residual left AV valve "cleft" between the superior and inferior bridging leaflets, with visible echo-bright suture material (Video 2).

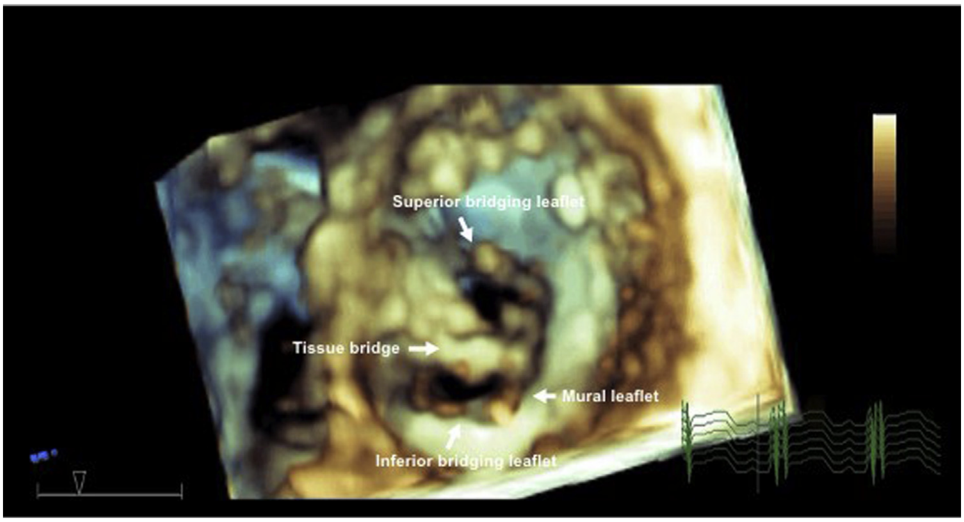

Figure 4 Three-dimensional view of the left AV valve from the ventricle shows the two valve orifices. There is a tissue bridge between the interventricular septum and the intersection of the inferior and mural leaflets. The small mural leaflet is partially obscured beneath the tissue bridge (Video 3).

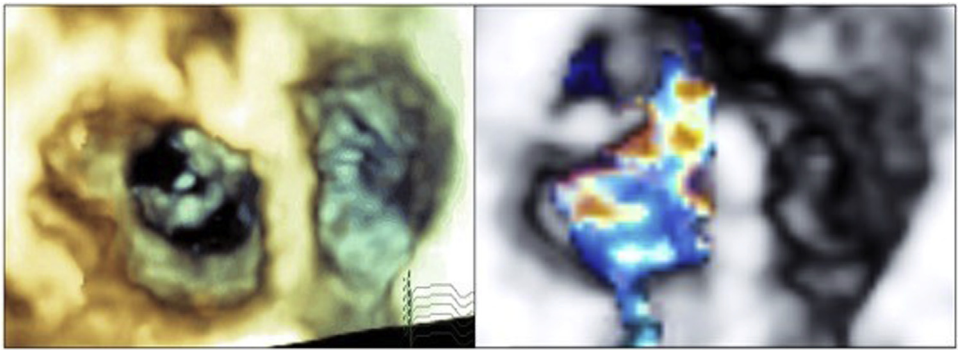

Figure 5 Three-dimensional view of both inflows from the atria (*left*) and with color showing regurgitation (*right*). Again, the double-orifice left AV valve and residual cleft are well profiled, and landmarks such as the atrial septum are easily seen.

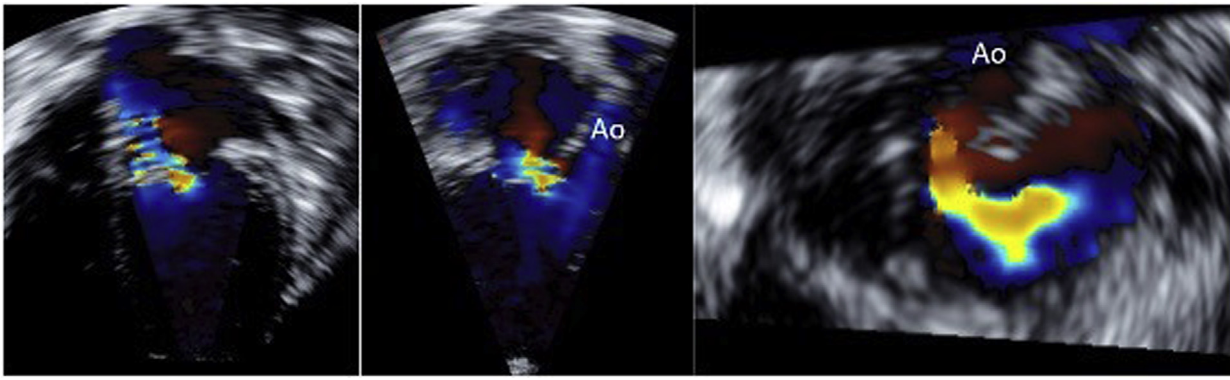

Figure 6 Three-dimensional vena contracta from three different views. There is a large regurgitant jet through the larger orifice at the site of the residual cleft. There is a smaller regurgitant jet through the second valve orifice.

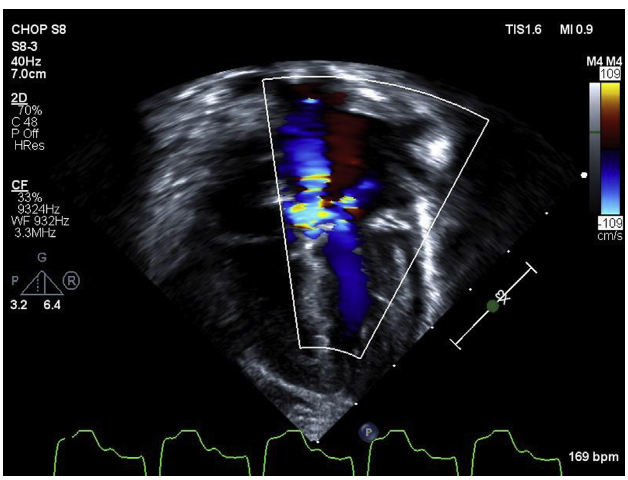

Figure 7 Transthoracic apical view with color obtained approximately 1 week after valvuloplasty shows severe left AV valve regurgitation through multiple jets.

angle for assessing the valve in systole (from apical) is difficult to achieve from epicardial views.[8]

CONCLUSION

Routine preoperative transthoracic 3D echocardiography may improve recognition of double-orifice left AV valves in common AV canal and assist in delineating valve anatomy and mechanism of regurgitation. However, the application of intraoperative 3D imaging remains challenging. Further work is needed to determine whether the detailed delineation of double-orifice left AV valve in common AV canal can lead to the design of improved repairs and better outcomes in this challenging population.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2020.02.005.

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