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Tips and Tricks of the Trade

Approach to Right-Sided Chamber Dilatation in Cardiac Shunts: Part 1 of a 2-Part Series

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Congenital cardiac shunts redirect blood flow, with each shunt creating a unique remodelling pattern. Transthoracic echocardiogram (TTE) commonly is the first step in shunt assessment, given its relatively low cost, high accessibility, and lack of radiation. We present a 2-part series reviewing cardiac shunts and their associated pattern of remodelling using TTE, with a focus on shunts that result in right-sided chamber dilatation. Image optimization and interpretation in ultrasound are challenging, even for cardiology graduates.[1] This guide was developed to assist clinicians in approaching chamber dilatation when a shunt is suspected but is not visualized directly.

Echocardiogram and Cardiac Shunts

In the context of cardiac shunts, trainees must understand how an image is generated and how it can be optimized to interrogate shunts, even if the shunts are not seen directly. An ultrasound image is generated based on the time required for waves to reflect to the transducer. Important to note is that if an object of interest, such as a shunt, is parallel to the waves, a lack of reflection may occur, and thus, a lack of image generation. This phenomenon is called tissue dropout, and it can explain false-positive findings of a septal defect. Thus, interrogation of a shunt should be done with the suspected defect positioned perpendicular to the ultrasound waves.

Colour Doppler is an integral tool in shunt assessment. Here, flow across a defect can be visualized by aligning the shunt direction parallel to the ultrasound waves. The colour generated is based on the Doppler effect, which is the change in detected frequency when the ultrasound waves are reflected. The frequency shift is represented in a colour scale of blue to red, with blue representing flow away from the transducer, and red representing flow toward the transducer. If the direction of flow is perpendicular to the waves, no Doppler shift will occur. Thus, in interrogation of a shunt, the optimal TTE view has the shunt perpendicular to the ultrasound waves, aligning the shunt flow parallel for visualization with Doppler.

In this guide, we highlight the subcostal 4-chamber, parasternal short axis at the base of the heart, and the off-axis apical 4-chamber view as particularly relevant to use to scrutinize cases of right-heart dilatation with a suspected shunt (Fig. 1).

Atrial septal defects (ASDs)

ASD refers to the 4 types of interatrial communication. Ostium secundum, the most common type of ASD,[2] is a defect in the atrial septum at the fossa ovalis. Ostium primum, the second-most-frequent phenotype, accounting for 10% of ASDs,[2] is a defect in the atrioventricular canal of the atrial septum and inherently will have associated abnormalities in the atrioventricular valves. Ostium primum and ostium secundum are true defects of the atrial septum.

Sinus venosus and coronary sinus defects are the other forms of ASD. Sinus venosus defect is an abnormality of the vena caval—atrial junction and the accompanying right pulmonary vein. Coronary sinus defects allow atrial communication through a hole at the coronary sinus leading into the left atrium (LA). They are the rarest form of ASD.

The anatomic origin of the defect, if visualized, is helpful in delineating the phenotype of interatrial communication (Videos 1-4, view videos online). The location of the defect also will influence the vector of shunt flow between atria.

In general, an ASD allows for continuous left-to-right shunting, with larger defects shunting more blood. With time, the right atrium (RA), right ventricle (RV), and pulmonary vasculature (including the main pulmonary artery [MPA]) are all expected to dilate due to the increased blood

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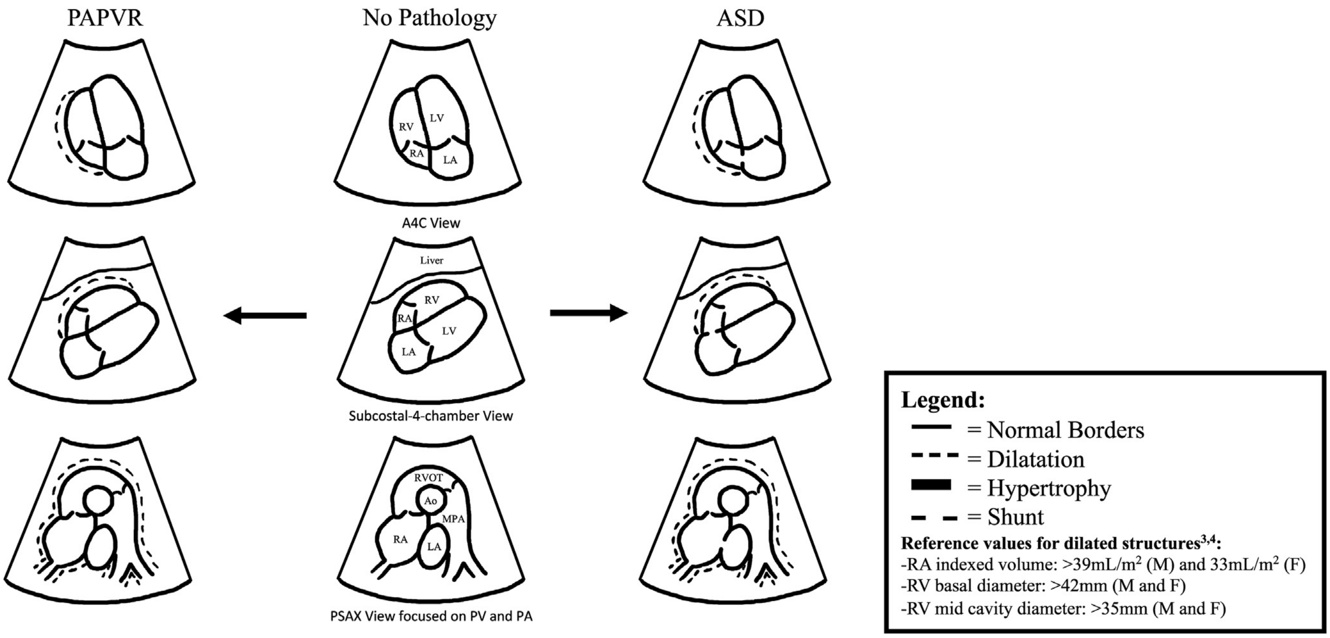

Figure 1. Patterns of remodelling in shunts that dilate the right heart. Schematics of apical 4-chamber (A4C) view, subcostal 4-chamber view, and parasternal short-axis (PSAX) view focused on the pulmonic valve (PV) and the pulmonary artery (PA), with **dotted lines** representing dilatation. Although the chamber enlargement patterns are comparable, notice the septal defect that may be appreciable in the case of an atrial septal defect (ASD). F, female; LA, left atrium; LV, left ventricle; M, male; PAPVR, partial anomalous pulmonary venous return; RA, right atrium; RV, right ventricle,

volume in the pulmonic circuit. The LA will not dilate, because the increased venous return passively flows into the RA. The RA likely is dilating when the tricuspid valve is closed, as the atrium must expand to accommodate the shunt. The RV and MPA will then dilate from the additional volume.

In using TTE to assess for ASD, subcostal windows are particularly useful, as the suspected shunt will be perpendicular to the ultrasound waves, and the shunted flow will be detectable with colour Doppler (Video 5, view video online). Recall that red flow with colour Doppler represents flow toward the transducer. Thus, given that blood is expected to be shunted from the LA to the RA, a red jet should be seen in the subcostal window if an ASD is present. The parasternal short-axis window at the level of the base of the heart is another option that can optimize the defect and shunt direction relative to the ultrasound waves.

If an ASD is suspected but not seen, an RV-focused apical 4-chamber view is useful to assess for dilatation of the RA and RV, using the volumetric cutoffs from the American Society of Echocardiography (ASE).[3,4] The apical 4-chamber view also is helpful, as an off-axis view can be used to assess, visually and with Doppler, for the presence of an ASD. Likewise, the parasternal short-axis window focused on the pulmonary vessels also can help detect dilatation of the MPA.

Sinus venosus and coronary sinus defects may be especially hard to detect with TTE. In such instances, the shunt interrogation can be enhanced using agitated saline contrast. In these forms of ASD, agitated saline can be seen entering both atria simultaneously.[5] Further, it also can delineate the specific sinus defect. For superior sinus venosus defects, a positive saline study should occur when saline contrast is injected into the upper extremities, but not when it is injected into the lower extremities. The inverse is true for inferior sinus venosus defects. Additionally, coronary sinus defects with left-sided superior vena cava also can be inferred if agitated saline is present bi-atrially when it is injected into the left arm, but not when it is injected into the right arm.

Partial Anomalous Pulmonary Venous Return (PAPVR)

In PAPVR, at least one, but not all, pulmonary veins return to the right heart instead of the left heart. PAPVR is rare, with an estimated prevalence of 0.1%.[6] The anomalous return commonly connects to the superior vena cava, but it also may drain into other veins, or directly into the RA. Most cases of PAPVR involve veins from the right lung, with < 10% of cases originating from the left side.[7] The most frequent phenotype described in the literature is the right upper pulmonary vein connecting to the superior vena cava.

PAPVR can be an isolated defect, but it also may be present with other shunts, particularly ASD.[7] In the case of an anomalous vein draining into the superior vena cava, special attention should be given to assess for concurrent presence of a sinus venosus ASD, and vice versa, as the 2 commonly are associated.

The symptomatology and disease severity in PAPVR depend on the degree of shunting and the number of veins involved. Many cases remain asymptomatic throughout life. As in ASD, PAPVR allows left-to-right shunting. The pattern of dilatation in PAPVR includes the RA, RV, and MPA (Fig. 1; Video 6, view video online). The left-heart chambers and vasculature are unaffected. This chamber dilatation pattern is identical to that of ASD. Pulmonary venous confluence also may be detected when additional imaging modalities are used (Video 6, view video online).

An important distinction between PAPVR and ASD is that shunted blood bypasses the LA in cases of PAPVR. Although LA volumes cannot be used to distinguish PAPVR from ASD clinically, indexed LA volumes may inform diagnostic suspicion, as the indexed volume in PAPVR should be lower than it is in ASD.

Shunt detection with TTE is especially challenging in PAPVR, as the location varies and may be hidden behind other structures. Nevertheless, in cases of right-heart dilatation with suspected PAPVR, the subcostal and off-axis apical 4-chamber views are useful for detecting a concurrent shunt. If those views are nondiagnostic for ASD, the level of suspicion for PAPVR should be higher.

Other Imaging Modalities

Although TTE is an excellent first test for investigating a shunt, further testing sometimes is needed. Transesophageal echocardiography is helpful in improving image quality, especially for 3-dimensional reconstruction to characterize the interatrial septum and the surrounding cavae, pulmonary vein, and coronary sinus. Cardiac magnetic resonance imaging is another useful tool, as it uses no radiation and is a highly accurate way to visualize and quantify shunts. Cardiac computed tomography is another option, although it has the associated caveat of radiation exposure.

Discussion and Limitations

This guide was developed as a tool to understand shunts that cause right-heart dilatation. However, the absence of the discussed changes cannot be used to rule out the presence of a shunt. Most importantly, the presence of right-heart dilatation does not confirm the presence of an ASD or a PAPVR shunt. The differential for right-heart dilatation includes cardiac shunts, right-heart valvulopathy, non-shunt congenital heart disease, and pulmonary hypertension and its associated causes. Further, shunt lesions may not present as isolated defects, especially in adults who have other comorbidities that dilate the heart, such as ischemic heart disease, hypertension, arrhythmias, and toxins. Thus, the presence of other cardiac remodelling similarly cannot be used to rule out the presence of a shunt. A holistic patient approach is needed to contextualize TTE findings with the clinical condition.

Ethics Statement

The research is in adherence to the ethical guidelines.

Patient Consent

Patient consent is not applicable to this article, as this article serves as a guide and does not use patient data.

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Disclosures

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Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Open* at https://www.cjcopen.ca/ and at https://doi.org/10.1016/j.cjco.2024.06.005.