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AN APPROACH TO IMAGING ADULT CONGENITAL HEART DISEASE: PITFALLS AND PEARLS

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Introduction

Noninvasive cardiovascular evaluation of an adult congenital heart disease (ACHD) patient is a multimodality endeavor that can include chest roentgenograms, electrocardiograms, echocardiography (including stress, three-dimensional, intravascular and intracardiac ultrasound, and transesophageal), cardiac magnetic resonance imaging (CMR), computed tomography angiography (CTA), single photon emission computed tomographic (SPECT) perfusion imaging, and positron emission tomography (PET). The cost-benefit ratio and limitations of each modality should always be considered (Table 1). Adults with ACHD often need repetitive imaging, making them vulnerable to radiation-induced cancer;[1, 2] hence, modalities using ionizing radiation should be minimized. A transthoracic echocardiogram remains the initial noninvasive modality of choice due to its real-time imaging capability with excellent temporal resolution and its ability to quickly assess hemodynamics along with anatomic delineation. A transesophageal echocardiogram is extremely helpful in further defining intracardiac anatomy and guiding surgical and interventional procedures. Cardiac magnetic resonance imaging is an adjunctive modality to an echocardiogram because ACHD patients frequently have intra-thoracic abnormalities (e.g., adhesions from multiple cardiac operations and lung disease) that can limit the resolution of the ultrasound beam. A CMR is especially helpful in periodic evaluation of the right ventricle, visceral situs, extracardiac anatomy, lesions of the pericardium, pulmonary veins, systemic veins, pulmonary arteries, and aorta.[3] For real-time CMR, special techniques such as echo planar can be used; for hemodynamic evaluation by CMR, techniques like velocity-vector mapping, myocardial tagging, echo planar, and fast gradient echo can be used. Besides excellent spatial resolution, the advantage of CTA over other techniques is an extremely fast acquisition time.[4] Furthermore, CTA can replace an invasive procedure such as catheterization and angiography for anatomical diagnosis where hemodynamics is not required, such as the diagnosis of arch or coronary artery anomalies. Nuclear scans such as lung perfusion scans and splenic scans are very helpful in determining differential pulmonary blood flow and splenic function, respectively. Both SPECT and PET are valuable in assessing tissue perfusion and metabolism and help to determine viable versus nonviable myocardium after surgery for hypoplastic left heart syndrome and coronary artery anomaly, after arterial switch operation for d-transposition of great arteries, and in acquired illness such as Kawasaki disease.

When considering imaging a patient with ACHD, an overall evaluation strategy should include the aspects found in Table 2.

	Ionizing Radiation	Time (hour)	Avoid in the Presence of Metals	Extra-cardiac Structures Spatial Resolution	Intra-cardiac Structures Spatial	Contrast Resolution	Temporal Resolution	Hemo-dynamic Assessment	Safe Contrast Agent
ECHO	No	<1	No	Good	Superior**	Good	Superior	Superior	Yes
CMR	No	>1	Yes	Good	Superior	Superior	Good	Good	Yes
CTA	Yes	<1	No	Superior	Superior	Superior	Fair	Fair	Yes
PET	Yes	<1	No	Poor	Poor	NA	Fair	Poor	NA
PET/CT	Yes	<1	No	Poor	Poor	Fair	Fair	Poor	No
SPECT	Yes	>1	No	Poor	Poor	NA	Fair	Poor	NA

Table 1. A comparison of noninvasive modalities for evaluation of congenital heart disease.
ECHO: echocardiogram; CMR: cardiac magnetic resonance imaging; CTA: computed tomography angiography; PET: Positron Emission Tomography; SPECT: single photon emission computed tomography; *Graded on the ease of a routine assessment; **Transesophageal study can be done to increase spatial resolution, but it will add to the time; NA, not applicable.

Cardiovascular Evaluation of Adults with Congenital Heart Disease
• Hemodynamics, myocardial perfusion, metabolism, and tissue characterization
• Undiagnosed congenital cardiac malformations
• Progressive native lesions
• Post-interventional and postoperative residual lesions
• Surgical anastomosis site
• Surgical procedures, shunts, and conduits
• Devices and complications
• Cardiac resynchronization therapy
• Congenital heart disease and pregnancy

Table 2. An approach to imaging adult congenital heart disease.

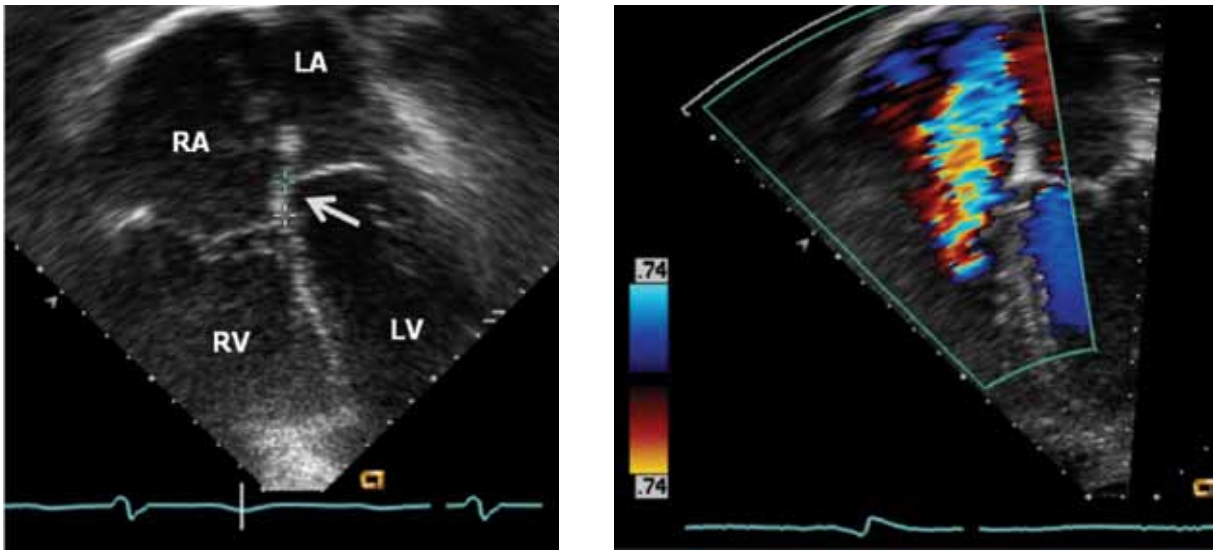

Figure 1 Transthoracic echocardiography with apical 4-chamber view showing apical displacement of the septal leaflet of the tricuspid valve in Ebstein's anomaly (arrow shows the distance displaced). Note the significant tricuspid valve regurgitation. (LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle)

Hemodynamics, myocardial perfusion, myocardial metabolism, and tissue characterization

In ACHD, besides evaluation of the left ventricle, it is important to focus on the right ventricle and estimate the right ventricular pressures. Both three-dimensional (3-D) echocardiogram and CMR are useful in evaluating the right ventricular size, morphology, and function. It is imperative that gradients and pressures be interpreted in conjunction with the ventricular function. The use of intravenous contrast agents should be avoided when pulmonary hypertension is present and should be used cautiously in the presence of intracardiac shunts. The hemodynamic and morphological assessment of complex heart diseases with the physiology of a single ventricle also needs specialized knowledge and protocol.

Exercise or pharmacologic stress echocardiography is an effective method for determining latent tissue dysfunction in ACHD. Contrast echocardiography, perfusion CMR, PET, and SPECT can be used for assessing qualitative and quantitative myocardial perfusion. CMR is invaluable in evaluating for myocardial fibrosis and quantifying ventricular volumes and mass, especially of the right ventricle.

Undiagnosed congenital cardiac malformations

The routine use of pediatric echocardiography has made it possible to find previously undiagnosed lesions, thus increasing the overall prevalence of congenital heart defects to between 3 and 4%.[5] Lesions such as bicuspid aortic valves, atrial septal defects, patent ductus arteriosus, coronary artery anomalies, Ebstein's anomaly (Figure 1), sinus of Valsalva aneurysm, and L-transposition of great arteries continue to be diagnosed for the first time in older populations.[5, 6] Knowledge precedes imaging, and unless the sonographer is trained in a thorough congenital heart protocol that focuses on acquisition of detailed anatomy, lesions can still be missed by a regular adult echocardiogram. For example, a patent ductus arteriosus with purely right-to-left shunt can be completely missed on a routine adult echocardiogram unless one delineates the ductus on a 2-D echocardiogram by a "ductal cut" view.

Besides complex anatomical lesions, even simple congenital heart defects, such as an atrial septal defect (ASD) and coarctation of aorta, need specialized knowledge and effort. For example, the subcostal sagittal plane is invaluable in detecting a sinus venosus or a secundum atrial septal defect. A primum atrial septal defect, however, is best seen from the apical 4-chamber view and a subcostal coronal plane. An acoustic "drop-out" at the septal level must be confirmed by spectral and color Doppler. Doppler investigation of the abdominal aorta is also essential in detecting a thoracic coarctation of the aorta. A transesophageal echocardiogram and CMR should be used as additional aids in complete delineation of the intra- and extracardiac anatomy, respectively. For example, delineation of an anomalous pulmonary venous drainage in association with a sinus venosus ASD is usually best done by a CMR.

Progressive native lesions

Congenitally deformed valves frequently continue to deteriorate throughout life. Subaortic and subpulmonary stenosis can worsen with age; in a single ventricle physiology, the bulboventricular foramen needs to be monitored for such an obstruction (Figure 2). Left ventricular outflow tract can also have a tunnel-like narrowing with obstruction in repaired atrioventricular canal defects and after atrial switch operations for d-transposition of the great arteries (d-TGA). Many defects are often associated with other lesions as well. For example, an unrepaired perimembranous ventricular septal defect (VSD) can lead to formation of a subaortic membrane, a double-chambered right ventricle, a left ventricular to right-atrial shunt through the tricuspid valve leaflet, and aortic valve prolapse with progressive aortic insufficiency.

It is well known that certain genetic syndromes are also associated with specific congenital heart defects. Supravalvular aortic stenosis, mid-aortic syndrome, supravalvular pulmonary stenosis, and peripheral pulmonary stenosis can all occur in diseases such as Williams syndrome, whereas diseases of the main and branch pulmonary artery can occur in Alagille syndrome, congenital rubella syndrome, Noonan syndrome, and Keutel syndrome. The aortic root and aortic arch ectasia can occur in diseases such as Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, familial aortic aneurysm, and in MASS syndrome. In congenital aortic valve disorders and in tetralogy of Fallot (ToF), a progressive dilatation of the ascending aorta can also occur. In this case, a CMR can be very useful in investigating the extracardiac vessels in their entirety.

Post-interventional and post-surgical residual lesions

Since repair of congenital cyanotic heart defects are complex and there is a much higher association for re-intervention with age, the

cardiac imaging needs to be tailored to the underlying disease.

Residual repaired and unrepaired septal defects need to be evaluated for hemodynamic significance. For example, a residual ventricular septal defect after patch repair or multiple unrepaired small muscular ventricular septal defects (often called "Swiss cheese VSDs") can lead to cardiomegaly with left atrial and ventricular dilatation and dilation of the main pulmonary arteries along with increase in blood flow velocities across the pulmonary, mitral, and aortic valves. Residual shunts can also be seen at various atrial baffles sites separating the pulmonary and systemic venous atria after Senning and Mustard operations for d-TGA (Figure 3). Contrast echocardiography can be used to determine residual atrial level shunts after atrial septal defect repair/device closure (Figure 4) or after atrial switch operation.

Congenitally deformed valves that were addressed in childhood need to be evaluated carefully for morphology and hemodynamics. Congenital pulmonary valve stenosis, seen either in an isolated form or with more complex diseases such as TOF, can be addressed in childhood by balloon valvuloplasty or surgical valvotomy with or without transannular patch augmentation. Transannular patch augmentation can lead to varying degrees of residual pulmonary valve stenosis and a significant insufficiency that can cause chronic pressure and volume overload of the right ventricle (Figure 5). CMR is useful in following right ventricular volumes and function to determine the optimal timing of valve replacement; it is also used to image the right ventricular outflow aneurysm, which can occur after patch augmentation (Figure 5).

In Ebstein's anomaly, the tricuspid valve (Figure 1) also needs accurate assessment to determine candidacy for operative intervention since surgical management is quite challenging and controversial. A detailed evaluation of the right ventricular function in an Ebstein's anomaly is crucial prior to any intervention. Atrioventricular (AV) canal defects can have AV insufficiency from residual clefts (Figure 6). Of note, repair of the AV can lead to residual mitral or tricuspid valve stenosis. The multiple left-sided obstructive lesions in Shone's complex (i.e., mitral valve stenosis, subaortic stenosis, aortic valve stenosis, and coarctation of aorta) can all recur and need to be assessed carefully.

Surgical anastomosis site

Comprehension of post-operative anatomy is paramount for imaging ACHD patients, since the majority of these patients have had at least one prior cardiac operation. For example, coronary ostia after arterial switch operation for d-TGA can become stenotic (Figure 7), and periodic assessment of the coronary arteries is imperative. Transthoracic echocardiography should be routine, but other options to evaluate for coronary compression and myocardial ischemia include stress echocardiography, perfusion imaging via SPECT, and CTA. If noninvasive

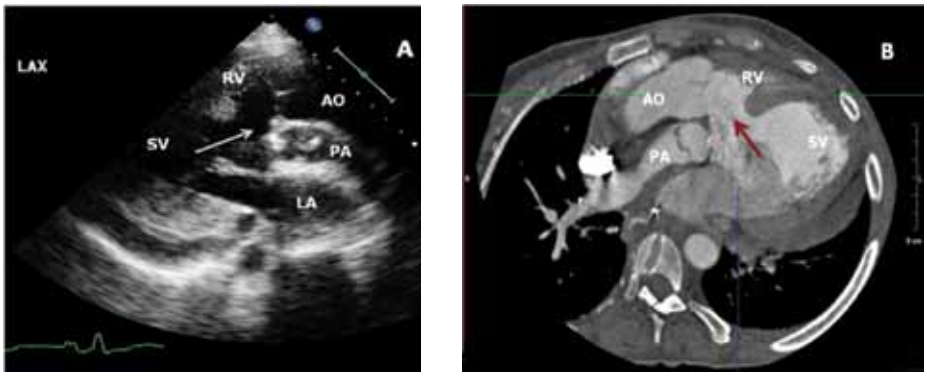

Figure 2 (A) Transthoracic echocardiography with parasternal long-axis view showing patent subaortic bulboventricular foramen (arrow). Note the hypoplastic right ventricle beneath the anteriorly placed aorta in this heart with single ventricular physiology. Also note the pericardial effusion and the blind pouch above the pulmonary valve in the oversewn pulmonary artery. (B) Computed tomography angiography showing a patent subaortic bulboventricular foramen (arrow) in a similar anatomy of a single ventricle (courtesy: Dr. Michael W. Bungo, Children's Memorial Hermann Hospital, Houston, Texas). (AO: aorta; LA: left atrium; PA: pulmonary artery; RV: right ventricle; SV: single ventricle)

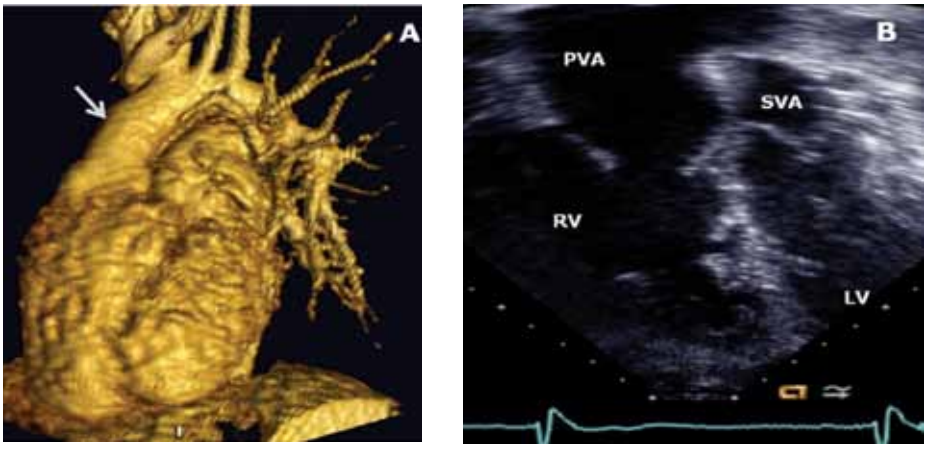

Figure 3. (A) Volume rendered reconstruction of gadolinium-enhanced 3-D magnetic resonance angiogram showing the D-transposition of the great arteries after Mustard operation. The arrow shows the anteriorly placed aorta arising from the right ventricle (courtesy: Dr. Timothy C. Slesnick, Texas Children's Hospital, Houston, Texas). (B) Transthoracic echocardiography with apical 4-chamber view showing an atrial switch procedure for D-transposition of the great arteries. Note the baffling within the atria. (LV: left ventricle; RV: right ventricle; PVA: pulmonary venous atrium; SVA: systemic venous atrium)

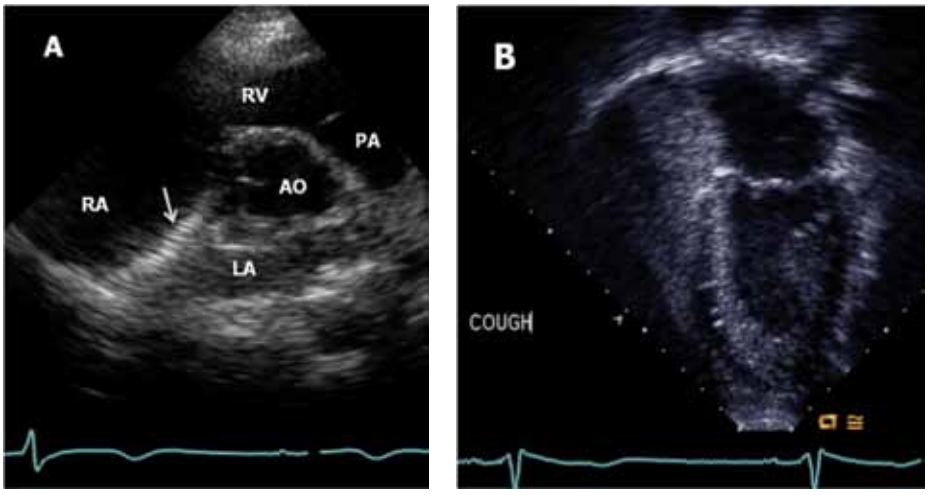

Figure 4. Transthoracic echocardiography with (A) parasternal short axis view showing an Amplatzer device (arrow) in place for a secundum atrial septal defect. (B) Apical 4-chamber view with saline contrast injection during Valsalva maneuver showing no residual shunt. (AO: aorta; LA: left atrium; PA: pulmonary artery; RA: right atrium; RV: right ventricle)

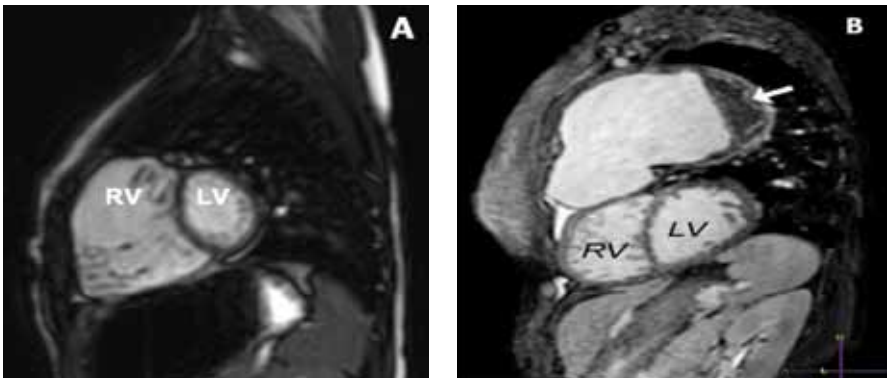

Figure 5. (A) Cardiac magnetic resonance imaging showing tetralogy of Fallot status post right ventricular outflow augmentation. Note the significantly enlarged right ventricle (courtesy: Dr. Timothy C. Slesnick, Texas Children's Hospital, Houston). (B) Cardiac magnetic resonance imaging showing tetralogy of Fallot status post right ventricular outflow augmentation with a large aneurysm of the right ventricular outflow. Note the thrombus in the aneurysm marked by an arrow (courtesy: Dr. Tal Geva, Children's Hospital Boston). (LV: left ventricle; RV: right ventricle)

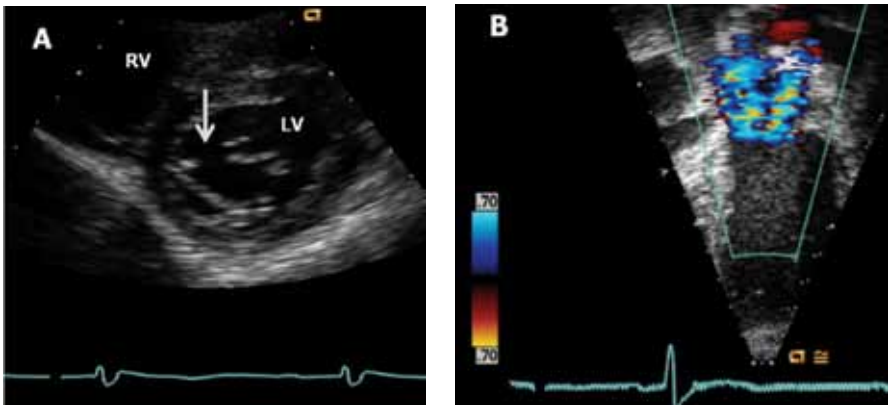

Figure 6. Transthoracic echocardiography with (A) parasternal short axis view showing a cleft in the mitral valve (arrow) of a repaired complete atrioventricular canal defect. (B) Apical 4-chamber view showing mitral regurgitation from the coaptation of the leaflets in addition to the mitral regurgitation from the residual cleft. (LV: left ventricle; RV: right ventricle)

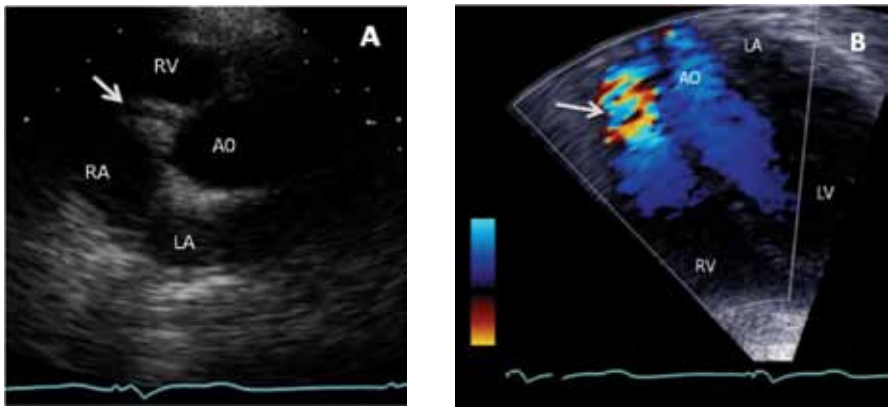

Figure 7. Transthoracic echocardiography with (A) parasternal short axis view showing the transplanted right coronary artery (arrow) after arterial switch operation for d-transposition of the great arteries. (B) Apical 4-chamber view showing the great arteries after arterial switch operation for d-transposition of the great arteries. Note the disturbed flow in the supravalvular pulmonary region at the suture site indicating supravalvular stenosis (arrow). (AO: aorta; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle)

imaging suggests a high likelihood for coronary artery compression, then cardiac catheterization for conventional angiography and angioplasty should be considered. Other modalities such as intravascular ultrasound (IVUS) may also play a role. Other sites of surgical anastomosis, such as at the supravalvular level (Figure 7), need to be followed for obstruction after arterial switch operation and Ross procedure. Several potential sites of obstruction after atrial switch procedures (Mustard or Senning) for d-TGA are noteworthy, especially at the pulmonary and systemic venous baffle sites and venoatrial junctions (Figure 3). Dilatation of the azygos veins may occur in the presence of superior vena caval obstruction, and this should also be investigated. Contrast echocardiography can be used to determine superior or inferior baffle obstruction after an atrial switch operation.

The Damus-Kaye-Stansel anastomosis site can become stenotic as well and should be evaluated carefully. The suture sites in the aortic arch after Norwood procedure or after repair of interrupted aortic arch and other arch procedures should be monitored for progressive narrowing and obstruction. The suture sites between the pulmonary arteries (PAs) and a conduit should be monitored for progressive obstruction as this can affect pulmonary blood flow and the growth of the distal PAs. A nuclear lung perfusion scan can be performed to demonstrate the differential pulmonary blood flow between the right and left lungs due to PA stenosis. Similarly, the suture sites in cavopulmonary shunts — between the superior vena cava (SVC) and branch PA (Glenn anastomosis) and inferior vena cava (IVC) and atrial baffle (intracardiac Fontan anastomosis) or conduit (extracardiac Fontan anastomosis) — should be evaluated for narrowing (Figures 8, 9). Physicians should specifically look for pulmonary venous obstruction of the right pulmonary veins, which can occur after intracardiac Fontan operation due to a dilated baffle. Furthermore, CMR and CTA can help delineate major arteriovenous and venovenous collateral vessels that can occur with cavopulmonary shunts. However, this is often best done during cardiac catheterization where these abnormal vessels can be device occluded.

Following repairs for pulmonary venous anomalies such as total anomalous pulmonary venous return, obstruction of the anastomosis sites and individual pulmonary veins can be followed by an echocardiogram (Figure 10) and by CMR. Surgical repair of lesions such as supravalvular mitral stenosis and cor triatriatum also need to be evaluated for residual obstruction to antegrade flow into the left atrium.

Surgical Procedures, Shunts, and Conduits

Obstruction at the pulmonary arteries after Lecompte maneuver during arterial switch operation for d-TGA can also occur with time (Figure 11). Evaluation of prosthetic valves should be done similar to protocol.

Conduits are routinely used to channel the right ventricular blood flow to the branch PAs in defects

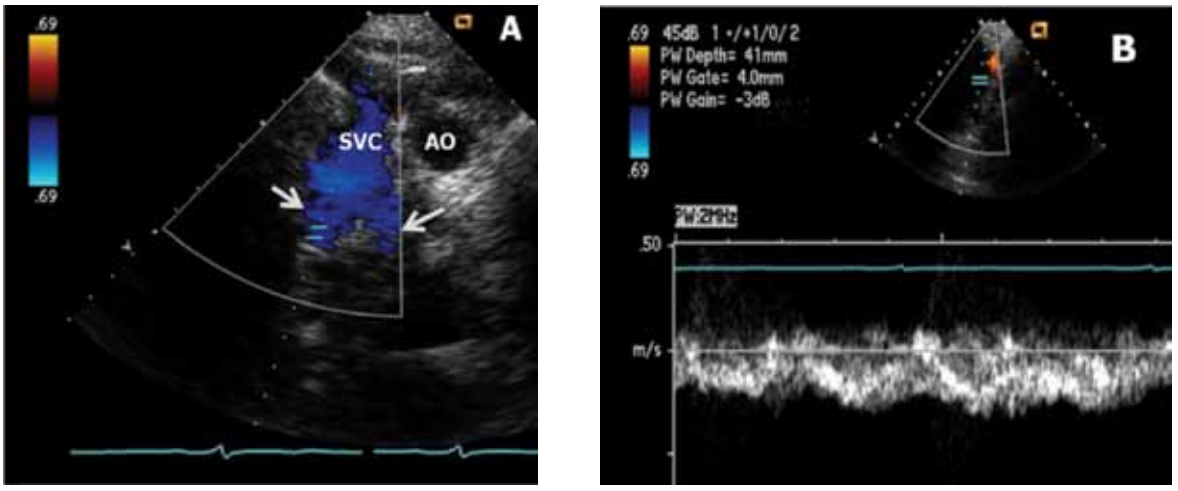

Figure 8. Transthoracic echocardiography with (A) suprasternal view showing color flow Doppler mapping of the superior vena cava after bilateral bidirectional Glenn anastomosis. Note the laminar antegrade flow in the branch pulmonary arteries (arrows). (B) Spectral Doppler interrogation in the right pulmonary artery shows low velocity biphasic flow. (AO: aorta; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle)

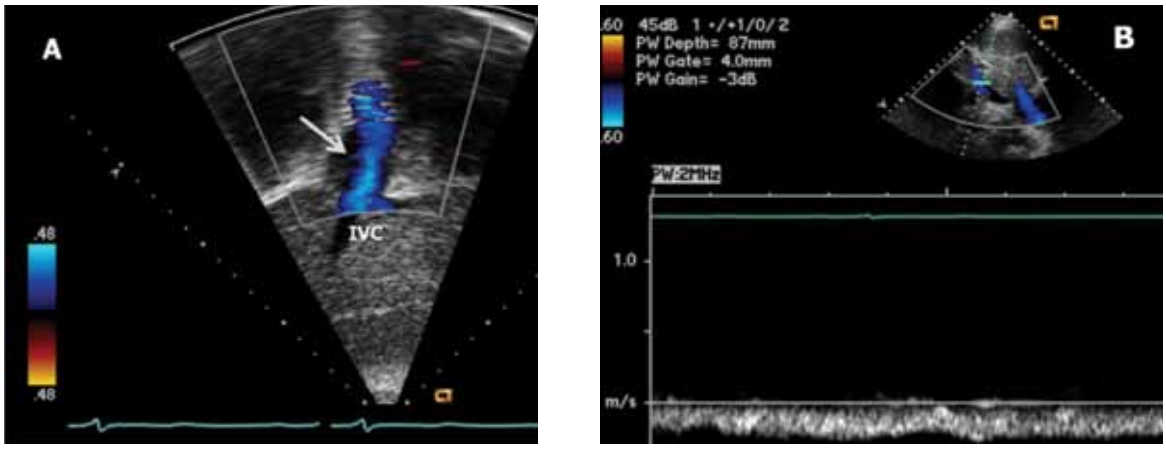

Figure 9. Transthoracic echocardiography with (A) subcostal sagittal view showing color flow Doppler mapping of the IVC along with hepatic vein after extracardiac Fontan anastomosis. Note the laminar antegrade flow in the conduit (arrow). (B) Spectral Doppler interrogation in the inferior vena cava showing low velocity biphasic flow. (IVC: inferior vena cava)

such as tetralogy of Fallot with pulmonary atresia or absent pulmonary valve, L-TGA, and truncus arteriosus and in surgeries such as Rastelli and Ross operations. Homografts, conduits, and shunts need to be closely followed for calcification and obstruction (Figure 12). There can be severe or "wide-open" pulmonary insufficiency due to failure of the valves in homografts or presence of valveless conduits (Figure 11) leading to progressive right ventricular enlargement. A patch is sometimes placed between the conduit and the right ventricular outflow tract, which can then become aneurysmal and is best evaluated from the subcostal views on echocardiogram and by CMR. Also, "jump grafts" used for severe aortic hypoplasia and coarctation of the aorta can be followed by CMR (Figure 13). Intracardiac baffles and conduits placed to divert left ventricular blood into the anteriorly placed aorta can also obstruct with time and need to be evaluated for the same.

Rarely, a palliative systemic-to-pulmonary artery shunt (Blalock-Taussig shunt, Waterston-Cooley shunt, Potts shunt, or a central Davidson shunt) may be present in ACHD, and both echocardiogram and CMR are valuable in assessing the size, gradient, and the patency of the shunt and the distal branch PAs. A PA band may be placed in childhood to palliate a lesion such as Swiss cheese VSD or a lesion with single ventricular physiology without pulmonary stenosis, and the band can be assessed by an echocardiogram and CMR. In the presence of a PA band, the right ventricular size, func-

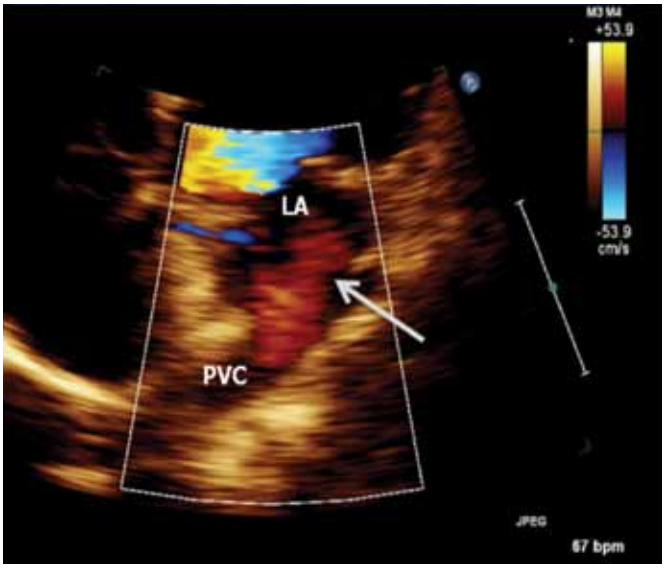

Figure 10. Transthoracic echocardiography apical 4-chamber view showing color flow Doppler mapping of flow across the fenestration between the pulmonary venous confluence and the left atrium after repair of total anomalous pulmonary venous return. Note the laminar antegrade flow across the fenestration (arrow). (LA: left atrium; PVC: pulmonary venous confluence)

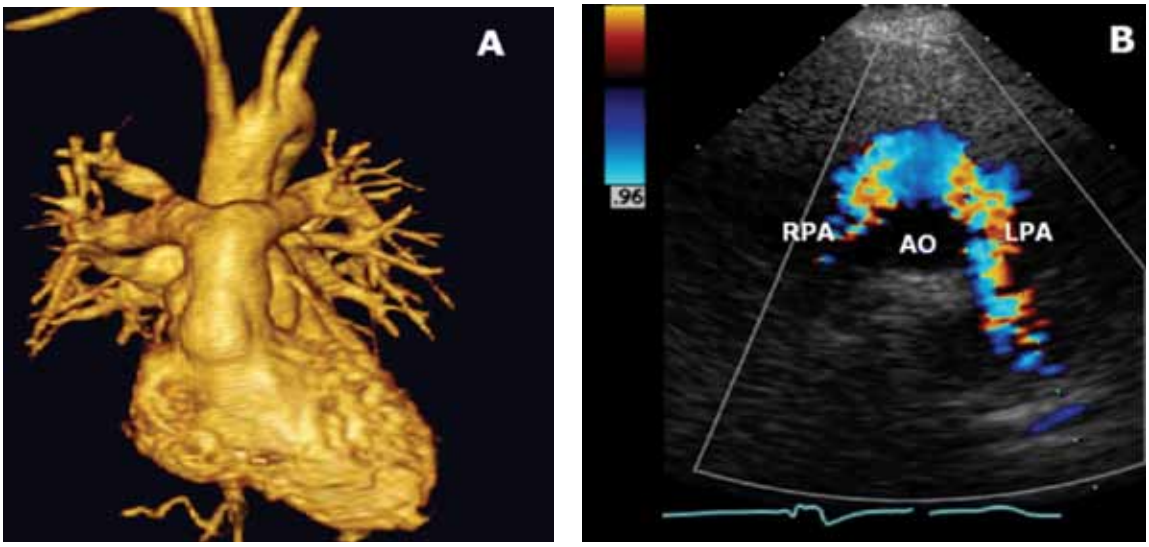

Figure 11. (A) Volume rendered reconstruction of gadolinium-enhanced 3-D magnetic resonance angiogram showing the branch pulmonary arteries after Lecompte maneuver in arterial switch operation for d-transposition of the great arteries (courtesy: Dr. Tal Geva, Children's Hospital Boston). (B) Transthoracic echocardiography with a suprasternal view showing color flow Doppler mapping of branch pulmonary arteries after Lecompte maneuver with disturbed antegrade flow in the branch pulmonary arteries indicating stenosis. (AO: aorta; LPA: left pulmonary artery; RPA: right pulmonary artery)

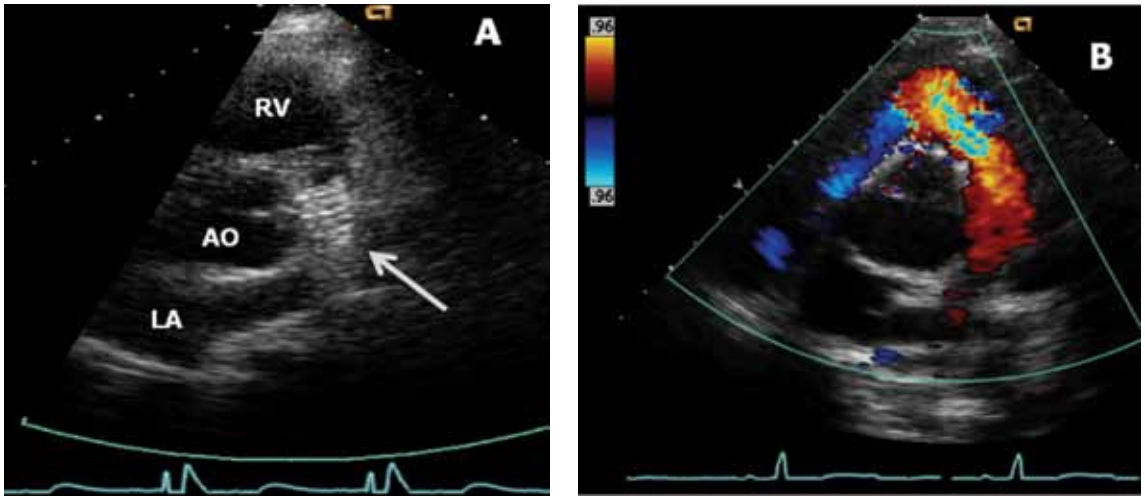

Figure 12. Transthoracic echocardiography with (A) parasternal short axis view showing a stent (arrow) after balloon angioplasty in the obstructed conduit after truncus arteriosus operation. (B) Color flow Doppler mapping of a conduit in tetralogy of Fallot showing severe or "wide-open" pulmonary insufficiency. Note the retrograde flow into the right ventricle in diastole (red) due to the pulmonary insufficiency. (AO: aorta; LA: left atrium; RV: right ventricle)

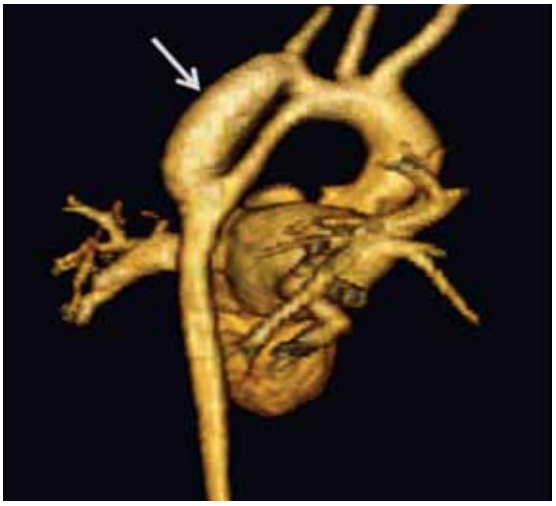

Figure 13. Volume rendered reconstruction of gadolinium-enhanced 3-D magnetic resonance angiogram in a patient with severe long-segment coarctation, status post jump graft (arrow) repair (courtesy: Dr. Tal Geva, Children's Hospital Boston).

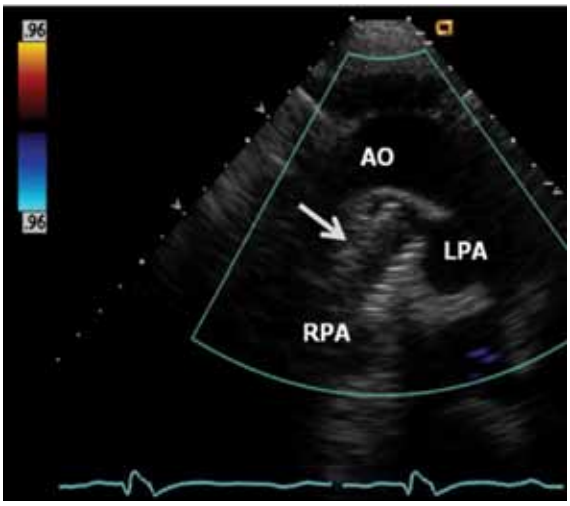

Figure 14. Transthoracic echocardiography with a suprasternal view showing a stent placed in the right pulmonary artery (arrow) after balloon angioplasty for stenosis. (AO: aorta; LPA: left pulmonary artery; RPA: right pulmonary artery)

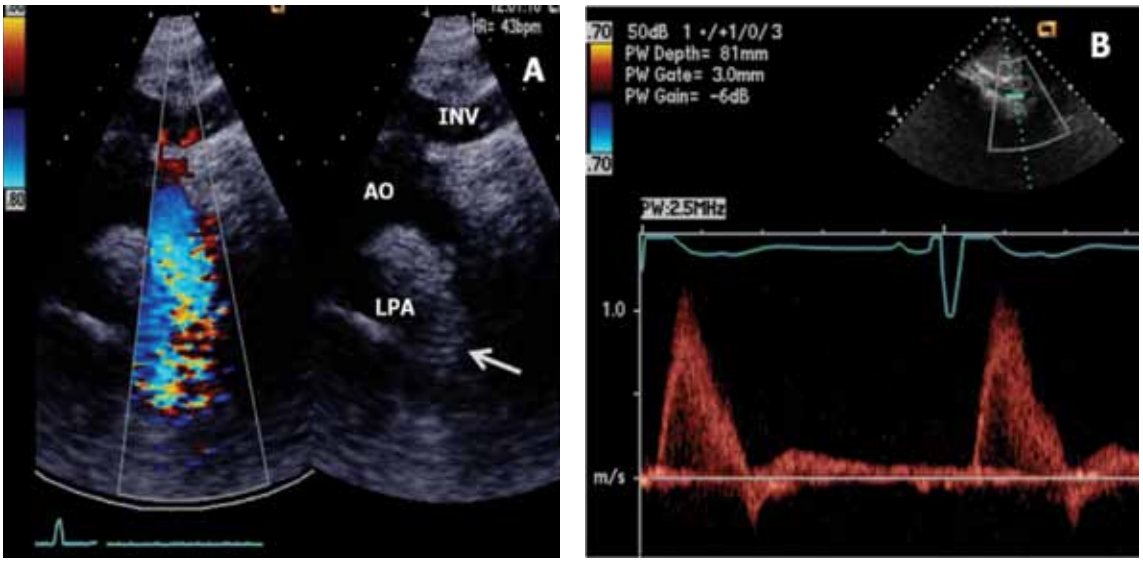

Figure 15. Transthoracic echocardiography with (A) suprasternal view showing antegrade flow in the proximal descending aorta after stent placement (arrow) for native coarctation of aorta. (B) Spectral Doppler mapping of the abdominal aorta below level of diaphragm in the same patient shows a normal pulsatile flow signal without continuous antegrade flow in diastole, indicating no significant residual coarctation. (AO: aorta; INV: innominate vein; LPA: left pulmonary artery)

tion, and mass need to be evaluated. One also needs to determine the degree of tricuspid regurgitation, estimate the right ventricular pressure, determine the gradient across the band and the left ventricular outflow tract, determine the position of band, and look for band migration and impingement over a branch PA.

Cavopulmonary shunts such as Glenn and Fontan anastomosis can develop thrombus, hence the entire circuit including the neighboring systemic venous system should be evaluated by high-resolution 2-D echocardiography followed by color Doppler mapping interrogation. In some cases of single ventricular physiology, the pulmonary valve is oversewn and forms a blind pouch above the valve where a thrombus can form (Figure 2). Therefore, when evaluating for a thrombus in a complex heart disease, this pouch needs to be investigated along with the usual cardiac chambers. Cavopulmonary anastomoses should also be assessed in their entirety by spectral Doppler as abnormal flow signals can portend pressure changes in the circuit.[7,8]

Devices and Complications

Interventional catheterization techniques have revolutionized cardiology, and these procedures have enabled clinicians to effectively treat many congenital cardiac lesions without the need for surgery. Occlusion of a secundum ASD by a percutaneous device is now the recommended first-line therapy for the uncomplicated lesion. These nonferromagnetic devices show minor image artifacts on CMR; however, transthoracic (Figure 4) and transesophageal echocardiograms are routinely used for their evaluation. Evaluation of a device should include determining the device position protrusion, migration, residual shunt, device compression of coronary sinus and/or vena cavae, impingement on AV node or mitral and tricuspid valves, pericardial effusion due to erosion, and presence of an atrial thrombus or vegetation. Percutaneous closure of a patent ductus arteriosus is also done routinely. As with any device, echocardiographic evaluation should include device position (looking for protrusion or embolization), determination of residual shunt, device compression of a branch PA, and impingement on or obstruction of the aorta. In addition, percutaneous device closure of muscular VSD is being performed in selected cases. Inspection for residual shunts, thrombus formation, erosion, and device migration should be done routinely.

Endovascular stents placed in childhood do not grow with age and can cause obstruction to flow in the PAs (Figure 14) and the aorta (Figure 15). The stents in the aorta are placed after balloon angioplasty for coarctation of the aorta, and they need to be carefully evaluated for aneurysm, dissection, and stent fracture and obstruction (Figure 15). CTA is particularly helpful in such cases.

Cardiac Resynchronization Therapy

Biventricular pacing for cardiac dyssynchrony is usually done to improve symptoms, hemodynamics and ventricular function. On echocardiography, the time-to-peak velocity difference between the septal and posterior walls (synchrony) is evaluated. Presence of systemic venous anomalies such as a persistent left SVC and interrupted IVC need to be determined prior to any transvenous pacing. Although biventricular pacing is not routinely utilized during congenital heart operations and some of the early data have not shown an acute benefit, it may serve a future role in patients with severe ventricular failure.

CMR during pregnancy

Gadolinium and other intravenous contrast agents should be avoided during pregnancy, especially if there is a risk of a right-to-left shunt.[9] Medical-legal aspects should be considered. For example, if a CMR is needed and a contrast agent is required, it should be documented in the medical record that (1) the information needed cannot be obtained from ultrasound or another diagnostic test that does not require ionizing radiation, (2) the information that is needed affects the care of the patient and/or the fetus during pregnancy, and (3) the attending referring physician denotes that the scan cannot wait until after the pregnancy and is needed to obtain the necessary information. In addition, informed consent must be obtained from the patient and signed by both the attending radiologist and the attending ordering physician.

Conclusion

Imaging the ACHD patient provides many challenges to clinicians. Intricate knowledge of unrepaired, repaired, and palliated congenital heart anatomy and congenital cardiac operations is vital. With many modalities in the imaging armamentarium, experienced physicians must select the proper diagnostic test and also have the ability to interpret these images in the plethora of ACHD disease states.

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