Pulmonary arterial hypertension (PAH) is a hemodynamic and pathophysiologic condition defined as an increase in mean pulmonary artery pressure (MPAP) of ≥25 mm Hg at rest measured at right heart catheterization (RHC). Patients with PAH associated with congenital heart disease (PAH-CHD) are a growing population consisting of an anatomically and phenotypically heterogeneous group, where differences among specific cardiac defects, along with their varied clinical course and prognosis, influence treatment choices for the individual patient.

**ROLE OF IMAGING**
Multimodality imaging plays a key role in assessing and managing patients with PAH-CHD. It is important to recognize the strengths/weaknesses and the complementary nature of different imaging modalities as well as the complex nature of the diagnostic questions that need to be addressed.

In this review, we will focus on the imaging tools commonly employed to evaluate PAH in CHD patients and their relative contribution to diagnostic assessment, evaluation of the functional and hemodynamic impairment, and longer-term prognostication.

**ECHOCARDIOGRAPHY**
Trans thoracic echocardiography (TTE) is the first-line cardiovascular imaging modality in the assessment of patients with various types of PAH because it is easy to apply, relatively inexpensive, and provides accurate information on cardiac anatomy and physiology.

In the setting of PAH-CHD, TTE is particularly suitable for the real-time interrogation of structural abnormalities as well as hemodynamic disturbances. In the majority of these patients, TTE allows the evaluation of cardiac anatomy (i.e., orientation and veno-atrial, atrioventricular, and ventriculoo-arterial connections), the morphology of cardiac structures, ventric-
Figure 1: The Fontan operation and its various modifications. (a) Classic Fontan operation. (b) Lateral tunnel with fenestration. (c) Extracardiac Fontan. (f) Fenestration; IVC, inferior vena cava; PA, pulmonary artery; RA, right atrium; SVC, superior vena cava. (Reprint from Gatouilis MA, Webb GD, Daubeny PEF. Diagnosis and Management of Adult Congenital Heart Disease. Oxford, UK: Churchill-Livingstone. 2003; page 85.).

Table 1: Proposed classification of PAH in the setting of congenital malformed hearts as based on circulatory pathophysiology (modified from 6). Abbreviations: ASD, interatrial communication; AVSD, atrioventricular septal defect; CHD, Tetralogy of Fallot without major anatomical obstructions of the pulmonary vascular system, and PAH; iPAH, (idiopathic) pulmonary arterial hypertension; PVR, pulmonary vascular resistance; PDA, patent arterial duct; POF, patent oval foramen; PVH, pulmonary venous hypertension; VSD, ventricular septal defect.

<table>
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<th>Significant shunting lesions</th>
<th>iPAH-like lesions</th>
<th>PAH due to past or present PVH</th>
<th>Eisenmenger physiology</th>
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<tr>
<td>a) For corrective surgery, PVR is low and presents no problem</td>
<td>a) Small unoperated lesion (eg, POF, ASD, VSD, PAD) not hemodynamically related to PAH</td>
<td>a) After corrective surgery of pulmonary venous stenosis or aortic/mitral valvar disease or coarctation, with normal wedge pressure and left ventricular function</td>
<td>a) Classical Eisenmenger physiology: no subpulmonary outflow obstruction; predominantly right to left shunting at atrial, ventricular, or arterial level, no intraventricular mixing</td>
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<td>a) Due to a surgical shunt previously created to increase pulmonary blood flow, which has led to significant PAH on that side</td>
<td>a) After corrective surgery of hypoplastic PA system</td>
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<tr>
<td>b) For corrective surgery, PVR elevated, risk increased but accepted</td>
<td>b) Small residue after corrective surgery of a shunting lesion, not hemodynamically related to PAH</td>
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<tr>
<td>c) For corrective surgery, PVR elevated, risk too high, not operable</td>
<td></td>
<td></td>
<td>c) Anatomy as above</td>
<td>c) With fenestration</td>
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Table 1: Proposed classification of PAH in the setting of congenitally malformed hearts as based on circulatory pathophysiology (modified from 6). Abbreviations: ASD, interatrial communication; AVSD, atrioventricular septal defect; CHD, Tetralogy of Fallot without major anatomical obstructions of the pulmonary vascular system, and PAH; iPAH, (idiopathic) pulmonary arterial hypertension; PVR, pulmonary vascular resistance; PDA, patent arterial duct; POF, patent oval foramen; PVH, pulmonary venous hypertension; VSD, ventricular septal defect.

- Although PASP measured by echocardiography correlates relatively well with PASP measured invasively, Bland-Altman analysis in the clinical setting demonstrates that large (10-20 mm Hg) differences between invasive and non-invasive PASP are common. The most common causes of inaccurate estimation of PASP include an incomplete Doppler envelope, resulting in underestimation of pressure or an overestimate of RA pressure from inferior vena cava diameter and collapsibility.  

- PASP regurgitation (TR) velocity (V) by applying the Bernoulli equation [PASP = 4V2 + estimated right atrial (RA) pressure, where V is the average peak TR velocity]. In patients with CHD, PASP can also be calculated using maximum flow velocity across a VSD or an aortopulmonary shunt (PDA, Blalock-Taussig shunt) (PASP = systolic blood pressure – 4V2).  

A few aspects must be kept in mind to ensure accurate estimates of PASP.

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• The right ventricular systolic pressure (RVSP) calculated from TR velocity may only be taken as the PASP in the absence of RV outflow obstruction. In CHD, more care should be taken to exclude any obstruction along the pulmonary pathway, especially after pulmonic valve surgery or in patients with previous systemic to pulmonary shunts. In some cases, peripheral or segmental pulmonary stenosis may also be present; this will require complementary imaging such as cardiovascular MRI to delineate.

• Velocity measurements are angle dependent. Tricuspid regurgitant jets should be taken from multiple acoustic windows (apical 4-chamber views, RV inflow, and off axis if necessary) with accurate transducer angulation in order to obtain a parallel intercept angle between the ultrasound beam and jet to avoid underestimation. In some cases of trivial regurgitant jet and suboptimal continuous-wave Doppler spectrum, the injection of contrast agents (agitated saline, sonicated albumin, or air-blood-saline mixture) may be required to achieve clear delineation of the jet envelope.11

• Close relationship between PASP and RV cardiac output exists. In cases of “end-stage” PAH, where both advanced RV dysfunction and increased PVR cause a significant reduction in stroke volume, PASP may appear “pseudonormalized” as a consequence of the low driving pressure generated by the failing RV.12 Underestimation of RV pressure may also occur with the development of diastolic RV dysfunction, characterized by high RA pressure and a stiff RV.

• Furthermore, in cases of severe TR the peak velocity may underestimate the trans-tricuspid pressure gradient because of early equalization of pressure between RA and RV, leading to truncation of the Doppler envelope.12

Pulmonary artery mean and end-diastolic pressure. Mean pulmonary artery pressure and pulmonary end-diastolic pressure (PAPD) are especially useful when TR velocity cannot be obtained or when further information is required.

A jet of pulmonary regurgitation, present in the majority of patients with PAH-CHD, permits the measurement of the end-diastolic pulmonary pressure using the modified Bernoulli equation: [PAPD = 4 × (end-diastolic pulmonary regurgitant velocity)² + RA pressure].

Similarly, MPAP can be determined from early peak pulmonic regurgitation velocity using the modified Bernoulli equation and adding the estimated RA pressure.13 Mean pulmonary artery pressure can also be estimated by using pulmonary acceleration time (AT) measured from the onset of RV ejection to peak pulmonary flow velocity (Figure 2). Generally, the shorter the AT, the higher the PVR and hence the pulmonary artery pressure. A value <105 ms is suggestive of PH.14 Mean pulmonary artery pressure can also be derived by regression formulas where: MPAP = 79 − (0.45 × AT). The same authors also found that in patients with AT <120 ms, the formula [MPAP = 90 − (0.62 × AT)] performed better.15

In addition to AT, the shape of the flow wave is of interest, as PH is associated with a deceleration of flow in mid systole (notching). In the presence of increased PVR and low arterial compliance, pulse wave reflection has greater magnitude and propagates more rapidly, arriving at the right ventricular outflow tract (RVOT) during systole.16

In patients with a Fontan circulation, as previously discussed, even a minor increase in pulmonary artery pressure may have significant hemodynamic consequences on the Fontan circulation. Conventional diagnostic criteria for PAH cannot be applied in this type of circulation. Information about MPAP in this setting can be derived from mean flow velocity across a fenestration between the Fontan or total cavopulmonary connection pathway and the atria; when such a fenestration is
present and can be interrogated by echo Doppler \(\text{MPAP} = 4 \, V^2 + \text{left atrial (LA) mean pressure}\). If this value is more than 17 mm Hg, it is highly suggestive of PAH.

Comprehensive diagnosis of PAH-CHD should combine Doppler pressure measurements with other accompanying echocardiographic features such as ventricular size and systolic function, as it is the RV that plays the key role in determining clinical presentation and prognosis in PAH-CHD.

**Assessment of RV Morphology and Function**

**Right ventricular morphology.** Normally the RV is a thin-walled chamber. In most forms of PAH, as a result of chronic progressive pressure loading, progressive RV remodelling occurs, initially in the form of hypertrophy and later as dilatation, along with progressive contractile impairment and, eventually, RV failure.

Compared to the patients with other forms of PAH, Eisenmenger syndrome hemodynamics and resulting RV remodelling are distinctly different. In adults with Eisenmenger syndrome with post-tricuspid defects and 2 ventricles, the RV often appears greatly hypertrophied with no significant dilatation. This unique physiopathologic adaptive model is explained by the preservation of a “fetal-like” phenotype without loss of RV hypertrophy and the presence of a ventricular communication, allowing both ventricles to function as a single entity.

In contrast, adults with PH and a pre-tricuspid shunt (ie, ASD) show greater LA, RA, and RV dilatation. It can therefore be postulated that loss of RV hypertrophy during infancy, lack of a training effect on the RV during childhood, and the absence of a ventricular communication that pairs the 2 ventricles functionally likely explain the differing RV response.

**Eccentricity index.** In patients with PAH, the high RV pressure may reduce the trans-septal pressure gradient between the 2 ventricles and leads to the frequently observed flattening of the intraventricular septum (IVS). M-mode analysis, with its high temporal resolution, can accurately estimate differences in the timing of leftward IVS shift during the cardiac cycle. Two-dimensional echo permits the quantification of the septal deformation using the eccentricity index, measured from a parasternal short axis view at the level of the chordae tendineae as the ratio of the left ventricle (LV) dimension parallel and perpendicular to the IVS respectively. It is usually measured both at end diastole and end systole with a normal value of 1.0, which occurs when the LV cavity maintains a round and symmetrical configuration on short-axis imaging. Mild, moderate, and severe septal bowing is represented by values of 1.1–1.4, 1.5–1.8, and >1.8.

**LV filling abnormalities.** Intraventricular septum deformation also alters LV shape, size, and diastolic filling. Thus, a common echocardiographic finding in these patients is blunted early diastolic filling of the LV, which in this scenario is not indicative of LA hypertension, but instead represents a marker of abnormal ventriculo-ventricular interaction. In fact, increased RV pressure and prolonged RV systole cause early diastolic reversal of the IVS. As a result, early diastolic transmural filling is reduced and redistributed to late diastole.

**Right ventricular function.** Assessment of RV function is the single most important aspect of the echocardiographic examination in patients with PAH, because symptoms and outcome both depend on the ability of the RV to adapt to an increased pulmonary vascular load.

Right ventricular dysfunction is challenging to quantify on echocardiography. All available acoustic windows and views should be used to provide complementary information and allow for a comprehensive assessment. Qualitative assessment of function based on visual inspection is commonly used in practice, but is limited by a significant interobserver variability, which is especially problematic when assessing relative changes in RV function in the same patient.

**Tricuspid annular plane systolic excursion.** Differences in muscle fiber orientation of the RV suggest that longitudinal shortening plays a greater role in RV emptying than in the LV. This predominantly longitudinal contractile pattern of the RV can be easily obtained.

Tricuspid annular plane systolic excursion (TAPSE) is the longitudinal systolic displacement of the RV base toward the RV apex and has been shown to correlate strongly with RV ejection fraction (EF).

Tricuspid annular plane systolic excursion can be derived using 2D guided M-mode (Figure 3a), is simple and highly reproducible, and has been recommended by American Society of Echocardiography (ASE) guidelines as part of routine echocardiographic evaluation. Normal values vary between 2.3 cm–2.6 cm, with a TAPSE of 2.0 cm likely representing the lowest acceptable normal value. Values in the range of 1.8 cm–2.0 cm, 1.6 cm–1.8 cm, and <1.6 cm are consistent with mild, moderate, and severe RV systolic dysfunction.

A significant limitation of TAPSE in PAH-CHD is that it is highly load dependent, such that it may become pseudonormalized in the presence of significant ventricular volume loading, such as with left to right shunting or severe TR.

**Tissue Doppler imaging.** Analogous to TAPSE, systolic wave velocity by tissue Doppler imaging (TDI) is a measure of longitudinal myocardial contraction. Tissue Doppler imaging like TAPSE is load dependent and may become pseudonormal under conditions of increased ventricular volume loading. Mean value in normal controls is approximately 15 cm/s at the annulus, with a lower accepted reference limit of normal of 10 cm/s.

**Fractional area change.** A more quantitative approach of assessing RV function is to measure the RV functional area change (FAC), \[[(\text{end-diastolic area} - \text{end-systolic area})/\text{end-diastolic area}] \times 100\], which has demonstrated a close correlation with RVEF by MRI. It is obtained by tracing areas of the RV at end diastole and end systole from the apical 4-chamber view beneath the trabeculations. Unfortunately, incomplete visualization of the RV cavity, especially when the RV is dilated, as well as difficulties in endocardial definition lead to relatively poor reproducibility, thus making it an unreliable tool for serial assessment.

**Myocardial performance index.** The myocardial performance index (MPI), also known as the Tei index, provides a
global assessment of both RV systolic and diastolic function. It can be calculated either from Doppler imaging (apical 4-chamber view for the tricuspid inflow pattern and the parasternal short-axis RVOT view for the determination of ejection time) or from TDI (single image from the lateral annulus of the tricuspid valve), according to the formula: MPI = (isovolumic contraction time + isovolumic relaxation time)/RV ejection time.24

Values greater than 0.40 by pulsed-wave Doppler or greater than 0.55 by tissue Doppler signify RV dysfunction.8 It has a good reproducibility, does not rely on geometric assumptions, and can be applied even in the presence of a suboptimal acoustic window. On the other hand, it is relatively load dependent and unreliable when RA pressure is elevated. Right ventricular ejection time, a component of MPI, has been shown to increase on targeted therapy for PAH.

Total isovolumic time. The total isovolumic time (t-IVT), which represents the sum of both isovolumic relaxation time (IVRT) and isovolumic contraction time (IVCT), can be calculated by subtracting filling time and ejection time from RR interval. It can be expressed as seconds per minute when calculated using the following formula: t-IVT=60 – [(ejection time × heart rate/1000) + (total filling time × heart rate/1000)].

Total isovolumic time is the time during the cardiac cycle heart neither ejecting nor filling. It is the total wasted time. In patients with increased pulmonary artery pressure, reduced pulmonary artery compliance will limit RV ejection time and prolonged tricuspid regurgitation duration will result in shortened filling time. Therefore, t-IVT will be significantly prolonged, and as a consequence stroke volume and hence cardiac output decreases.18,25 Total isovolumic time can be used for monitoring disease progression and assessing prognosis.

Advanced right ventricular imaging. Speckle tracking—strain and strain rate examine the deformation and rate of deformation of the myocardial segments. These represent a method of assessing intrinsic RV myocardial contractility that is less load dependent, but currently remain outside the standard echocardiographic protocols due to the lack of normative data and high interobserver variability.26

Real time 3-dimensional echocardiography can overcome the limitations of 2D echo in the assessment of RV volumes and EF. Three-dimensional echocardiographic RV volumes are comparable to those derived by MRI, although little data exist for significantly dilated or dysfunctional ventricles.27 Technologies, able to create a 3D model of the RV by a post-processing analysis of anatomical landmarks identified in any 2D view, are currently under investigation in an ongoing study in PAH.28

Echocardiographic Predictors of Clinical Outcome

Different echocardiographic variables have been demonstrated to yield prognostic information that may guide clinical management. From the current literature, and according to the European Society of Cardiology (ESC) guidelines, the echo-

Figure 3: (a) TAPSE by M-mode recording. (b) Kaplan-Meier curve for TAPSE. Patients with TAPSE <15 mm had higher mortality rates than patients with TAPSE ≥15 mm. (Reprinted with permission from Moceri P, Dimopoulos K, Liodakis E, et al. Echocardiographic predictors of outcome in Eisenmenger syndrome. *Circulation*. 2012;126:1461-1468.)
cardiographic indices most closely associated with unfavorable outcome, including RA area index, diastolic eccentricity index, pericardial effusion, MPI, and TAPSE, are all indicators of RV decompensation. However, prognosis is significantly affected by the etiology of PAH. Patients with Eisenmenger syndrome exhibit a better prognosis compared with idiopathic PAH and connective tissue disease–associated PAH. Patients may survive decades after the initial diagnosis of PAH-CHD, even before the advent of advanced targeted PAH therapy. As mentioned before, the difference in outcome is thought to be related to better adaptation of the RV to systemic or high pulmonary artery pressure. In support of this view, we have recently demonstrated that the longitudinal function of the RV is preserved or mildly impaired in the majority of patients with Eisenmenger syndrome, and that even though RV dilation was prevalent, it was less severe than that described in idiopathic PAH and was not related to adverse outcome.29

Right ventricular long axis function (TAPSE). Right ventricular longitudinal contraction in Eisenmenger patients has been shown to be an independent prognostic factor, both in our cohort, where even small reductions in TAPSE were associated with adverse outcome (Figure 3b), and in another recent prospective study from Van De Bruaen and colleagues where TAPSE <15.9 mm was predictive of lower event-free survival and higher all-cause mortality.30

Ratio of RV effective systolic to diastolic duration. The duration of TR, a marker of impaired adaptation to pressure overload and of RV failure, is strongly related to outcome. In fact, in these cir-

Figure 4: (a) Measurement of effective RV systolic and diastolic duration on trans-tricuspid Doppler. (b) Measurement of RA area on 2D echocardiogram. (c) Time-dependent receiver operating characteristic curves at 1.5 years for the echocardiographic composite score (TAPSE <15 mm, ratio of RV effective systolic to diastolic duration ≥1.5, right atrial area ≥25 cm², RA/LA area ratio ≥1.5). (Reprinted with permission from Moceri P, Dimopoulos K, Liodakis E, et al. Echocardiographic predictors of outcome in Eisenmenger syndrome. Circulation. 2012;126:1461-1468.).
cumstances RV filling time is limited by prolongation of TR in presystole and/or early diastole, and cardiac output may consequently decrease. In order to improve the diagnostic power of echo in Eisenmenger patients, a ratio of RV effective systolic to diastolic duration can be calculated (Figure 4a). Durations of systole and diastole can be measured from the clearest Doppler signal of TR from the apical view. Effective systolic duration is measured from the onset to the end of TR. Effective diastolic duration is measured from the end of TR to the onset of the subsequent TR signal. A ratio ≥1.5 is an independent predictor of outcome.29

**Right atrial area and ratio of RA to LA area.** Parameters reflecting high central venous pressure have also been shown to predict mortality in PAH. Right atrial dilation is a reflection of long-standing pressure overload and ensuing heart failure. Quantitative assessment of RA size is performed from the apical 4-chamber view (Figure 4b). Right atrial measurements are obtained at the end of ventricular systole, when chamber size is maximal. Right atrial area is usually measured, as it has been reported to predict adverse outcome in the setting of PAH. Eisenmenger patients with pre-tricuspid shunts, who are thought to have a worse prognosis compared with those with post-tricuspid shunts,31 are expected to have larger atria because of the long-standing shunt at the atrial level. Right atrial dilation, beyond being a marker of right-sided overload and possibly stiffness of a hypertrophied RV, is also a predisposing factor for arrhythmias. Mortality risk is significantly increased when RA area is ≥25 cm² or RA/LA ratio is ≥1.5.29

All the parameters discussed above have their limitations when used in isolation. Comprehensive assessment with a combination of multiple parameters provides the most accurate prognostication.

In our cohort, a composite score based on these strong echocardiographic predictors of outcome (TAPSE <15 mm, ratio of RV effective systolic to diastolic duration ≥1.5, RA area ≥25 cm², and RA/LA area ratio ≥1.5) identified patients with more than 3-fold increased risk of death at 1.5 years, with a very high area under the curve on receiver operating curve (Figure 4c).

**CHEST RADIOGRAPHY AND CARDIAC COMPUTED TOMOGRAPHY**

A plain chest x-ray provides a record of cardiac size, which in the CHD population as a whole carries prognostic significance.32 The typical changes of PAH on the chest x-ray are enlargement of the central pulmonary arteries with relative pruning of the distal vessels. There may also be signs of specific chamber enlargement or other features to suggest a particular underlying defect (Figure 5).

Although disadvantaged by the need for ionizing radiation and contrast, computed tomography (CT) has an important part to play in the investigation of PAH-CHD, as it provides information on cardiac chambers, great arteries, lung vasculature, and parenchyma and mediastinal structures in a single acquisition with high spatial resolution.33 This is particularly the case when acoustic windows have been poor (limiting echocardiography), lung disease is present, or devices such as pacemakers prevent cardiovascular magnetic resonance (CMR) scanning.

Similar features of cardiac physiology associated with PAH described previously can also be identified using cardiac CT. Communications between chambers can be visualized and the direction of shunt inferred by the direction of contrast (Fig-
High-resolution CT scanning provides valuable information on lung parenchyma, which can be abnormal in patients with CHD because of bronchiectasis or hypoplasia. It may also detect parenchymal changes due to PAH, for example, ground glass changes, nodular opacities, and serpiginous intrapulmonary vessels. In those with chronic thromboembolic PH, it may also identify within the lung tissue hemorrhage, infarction, or a mosaic pattern (due to heterogeneous lung perfusion). High-resolution CT has a vital role in identifying patients with pulmonary veno-occlusive disease, where advanced therapies might be harmful. The key features of this rare entity on CT are interlobular septal thickening, ground glass shadowing, and adenopathy.

When contrast is enhanced, this technique is particularly adept in the assessment of extracardiac features, particularly native or surgically fashioned systemic to pulmonary shunts. Collateral vessels are readily identified. These include dilated bronchial arteries (a feature commonly seen in PAH) or bypassing vessels in cases of pulmonary venous occlusion.

An analytic technique called fractal analysis has been studied to determine whether the degree of branching within the pulmonary arteries of children and young adults with PAH, half of whom had CHD, could be used as a noninvasive...
Figure 7: Representative examples of segmented pulmonary artery masks, and below them the derived skeletonized representations for patients with mild, moderate, and severe PH. (Reprinted with permission from Moledina S, de Bruyn A, Schievano S, et al. Fractal branching quantifies vascular changes and predicts survival in pulmonary hypertension: A proof of principle study. *Heart*. 2011;97:1245-1249.).

Figure 8: (A) Cardiovascular magnetic resonance still systolic frame from balanced steady-state free precession cine image demonstrating a large PDA (asterisk) measuring 13 mm diameter. The pulmonary artery is severely dilated with maximum diastolic diameter 50 mm. The RV is severely hypertrophied. (B) A corresponding in-plane phase velocity map is shown, and blood flow is demonstrated to be predominantly from the main pulmonary artery to the aorta in systole.
measure of PH (Figure 7). This index compared well with conventional markers of disease severity such as functional class, 6-minute walk test distance, and indexed PVR, and predicted death among the cohort [HR 5.6 (95% CI 1.2 to 25; \( P = 0.027 \)].

CARDIOVASCULAR MAGNETIC RESONANCE

Cardiovascular magnetic resonance has the major advantage of being able to image in any plane with high spatial and temporal resolution without requiring ionizing radiation. As repeated examinations become common, the latter characteristic is particularly beneficial for patients with PAH-CHD.

In order to optimize image quality, breath holding is required. This may be problematic for some patients with PAH-CHD. In addition, pacemakers/devices currently represent a contraindication to routine CMR.

CMR acquisition and subsequent analysis of a stack of contiguous cine image covering the whole heart from base to apex provides the gold standard assessments of right and left ventricular size and function, especially so for heavily trabeculated RVs. However, analysis remains operator dependent.

Using both cine imaging and in-plane flow mapping, intracardiac shunts can be easily identified and quantified. By comparing the ratio of flow through the pulmonary

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Figure 9: Typical flow patterns in the RVOT at different cardiac phases for a patient with manifest PH (A, D, and G), a patient with latent PH (B, E, and H), and a normal subject (C, F, and I). PA indicates main pulmonary artery; PV, pulmonary valve; and RV, right ventricle. At maximum outflow (A through C), flow profiles were distributed homogenously across the cross sections of the main pulmonary artery in the manifest PH group (B), and the normal group (C). In later systole (D through F), a vortex was formed in the manifest PH group (D). No such vortex could be found in the latent PH group (E) or the normal group (F). After pulmonary valve closure (G through I), the vortex in the PH group persisted for some time. In all cases, continuous diastolic blood flow upward along the anterior wall of the main pulmonary artery could be observed. (Reprinted with permission from Reiter G, Reiter U, Kovacs G, et al. Magnetic resonance-derived 3-dimensional blood flow patterns in the main pulmonary artery as a marker of pulmonary hypertension and a measure of elevated mean pulmonary arterial pressure. Circ Cardiovasc Imaging. 2008;1:23-30.)
artery to that in the aorta using flow mapping. Qp:QS can be determined and aortic flow mapping can be used to determine cardiac output. Tricuspid and pulmonary regurgitation can be assessed with cine imaging, through-plane and in-plane flow.

With the addition of contrast enhanced magnetic resonance angiography, extracardiac shunts (Figures 8a and b) as well as the pulmonary vasculature can be delineated. Septal motion can be qualitatively noted on cine imaging just as in echocardiography.

Late gadolinium enhancement for myocardial tissue characterization has been applied to PAH and typically demonstrates areas of enhancement at the RV-LV insertion points. Histological data, however, support the concept that the myocardial disarray at these sites is a normal feature of insertion-region anatomy exaggerated in PH by the hypertrophy of the RV.

Significant efforts have been given over to noninvasively assessing pulmonary pressures using CMR with indices such as RV mass, septal deviation, pulmonary artery stiffness, and more recently 4-dimensional flow patterns (Figure 9). Of the longer established measures, none has emerged as a reliable measure in patients with PAH. Moreover, none have been routinely tested in a CHD population where the presence of pre-existing RV hypertrophy, septal defects, shunts, and arterial abnormalities may confound many of these parameters.

In the setting of a hybrid CMR interventional lab, PVR derived from the Fick method has been shown to be inaccurate in conditions of high pulmonary blood flow or increased oxygen concentration. With promising results, the same technique has been used to show that PVR can be determined noninvasively in a small cohort with mainly atrial and ventricular septal defects. A pulmonary flow of 6.05 l/min/m² or a Qp/QS ratio <2.5/1 had a specificity of 100% for predicting PVR of ≤3.5 Wood units/m² on receiver-operator characteristic analysis.

CMR Assessment of Treatment and Prognosis

The main measure used in clinical practice and as a trial endpoint in PAH is the 6-minute walk test. However, this has faced considerable criticism given its limitations and failure to demonstrate a relationship with clinical endpoints. This has spurred investigation into alternative surrogates, one of which has been RV mass as assessed by CMR. This measure, when employed after medical and surgical therapies, has been shown to reflect improvements in indices of remodelling.

Cardiovascular magnetic resonance has also been used to prognosticate in patients with PAH. Right ventricular dilatation and impaired systolic RV function as well as increased degrees of pulmonary artery stiffness are predictors of a poor outcome in PAH. Unfortunately, data specific to PAH-CHD have yet to be published.

**CMR in Eisenmenger Syndrome**

Cardiovascular magnetic resonance in Eisenmenger syndrome occasionally correctly defines the precise nature and functional significance of the underlying CHD. In surgically palliated patients, CMR can be used to assess the presence and patency of surgical shunts. It is also useful for assessment of other relevant extracardiac features such as PDA or aortopulmonary collaterals. The central pulmonary arteries should be imaged for presence of aneurysmal dilatation, poor expansibility, sluggish flow, and in situ rather than thromboembolic pulmonary arterial thrombus.

**CONCLUSION**

In summary, multimodality imaging plays a major role in assessing patients with PAH-CHD in terms of anatomy, physiology, presence, extent and progression of PAH, and RV function. Different imaging modalities come with strengths and weaknesses, and physicians and imaging specialists should be aware of their complementary and prognostic role, as to provide the optimal therapy and outcomes for the patient with PAH-CHD.

**References**


