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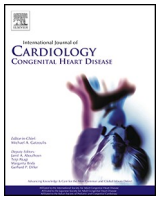

Advances in the imaging of pulmonary hypertension

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A B S T R A C T

Pulmonary hypertension (PH) is a complex and progressive disorder characterized by elevated pulmonary arterial pressures leading to right ventricular dysfunction and increased morbidity and mortality. Non-invasive imaging, including echocardiography, computed tomography (CT) and cardiovascular magnetic resonance (CMR), plays a crucial role in the diagnosis, risk stratification, and management of PH. The integration of these imaging modalities facilitates a multimodal approach to PH evaluation, enabling more precise diagnosis, improved phenotyping, and better-guided therapeutic decision-making. Echocardiography remains the first-line modality, offering valuable insights into pulmonary artery pressures, right ventricular size and function, and associated cardiac anomalies. Recent developments in speckle-tracking echocardiography and 3D imaging have enhanced its diagnostic and prognostic utility. CT imaging provides detailed evaluation of the pulmonary vasculature, parenchyma, and perfusion, which is essential in distinguishing PH subtypes. CMR is non-invasive, radiation free, and highly sensitive to changes in anatomy and function making it ideal for the long-term follow up of patients with PH. It offers in-depth evaluation of all cardiac chambers as well as pulmonary blood flow assessment and tissue characterisation. In this work we discuss current strengths, limitations, and future directions in these key imaging modalities used for the comprehensive assessment of PH.

1. Introduction

Pulmonary hypertension (PH) represents a serious progressive condition which can be associated with various cardiovascular and respiratory disorders. It is considered that approximately 1 % of the global population is affected by PH, rising to 10 % among individuals aged over 65 years [1]. According to recent epidemiological data, its prevalence has doubled over the last 10 years [2]. It is associated with a significant decrease in exercise tolerance and can lead to progressive right ventricular (RV) dysfunction, development of heart failure (HF), life-threatening arrhythmias, and premature death [3,4].

Both treatment and prognosis are highly dependent on the underlying cause of PH; therefore, careful assessment of patients with suspected PH is critical for accurate classification. Although right heart catheterization (RHC) is considered the gold standard for PH diagnosis [5], non-invasive imaging is essential for identifying and classifying PH, allowing for earlier diagnosis and management. Echocardiography remains the first-line diagnostic tool for patients with suspected PH due to its wide availability and accuracy [6]. In recent years, major advances in imaging techniques have highlighted the value of a multi-modality imaging approach including computed tomography (CT) and cardiovascular magnetic resonance (CMR) in the classification, risk stratification, and follow-up of patients with PH [7,8]. The aim of this review is to provide an overview of current imaging modalities applied in the evaluation and monitoring of patients with PH.

2. Echocardiography

Transthoracic echocardiography is the first-line diagnostic modality when evaluating patients with suspected PH. It can provide information regarding RV and left ventricular (LV) morphology and function, valvular abnormalities, intra-cardiac shunts, provide an estimate of hemodynamic parameters, as well as allowing for assessment of potential underlying causes of PH, including congenital heart disease (CHD) and left-heart disease. Based on echocardiographic findings the probability of the PH and the need for further investigation, including RHC,

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can be estimated. Echocardiography provides both diagnostic value and prognostic information, and has a pivotal role in monitoring the response to therapy and monitoring during long-term follow-up [9,10]. RV dysfunction leading to progressive RV failure is the major cause of morbidity and mortality in PH; therefore, the precise evaluation of its morphology and function is crucial [11,12]. This is often challenging as the shape of the RV is complex and multiple echocardiographic parameters should be considered when assessing the RV. In Table 1 and Fig. 1 we provide commonly used echocardiographic parameters in PH assessment.

2.1. Hemodynamic assessment

Systolic pulmonary artery pressure (sPAP) may be estimated based on the peak tricuspid regurgitation velocity (TRV) and the estimation of the right atrial pressure (RAP). However, this is only true in the absence of right ventricular outflow tract (RVOT) obstruction. TRV is obtained by continuous wave (CW) Doppler of the tricuspid valve (TV) regurgitant jet, and the modified Bernoulli equation is then used automatically on the machine to calculate and provide the pressure gradient across the TV. The estimation of RAP is added to the pressure gradient across the TV to obtain estimated sPAP. While RAP can be estimated by measuring the dimension of inferior vena cava (IVC) obtained from the subcostal view combined by evaluation of its collapsibility during the inspiratory sniff [13] this can lead to numerous inaccuracies, hence, the most recent ESC guidelines recommend using the peak TRV as the key variable for assessing the probability of PH rather than derived sPAP. A peak TRV of >2.8 m/s may suggest the presence of PH [14]. Correlations between Doppler derived sPAP and invasive measurements are high; however, discordance may occur and the presence or absence of PH cannot be confirmed by TRV alone [15,16]. Inaccuracies may be caused by insufficient TV regurgitation jet, Doppler misalignment with the jet, poor acoustic windows, underestimation of pressure gradients in case of severe TV regurgitation or overestimation in patients with high cardiac output (e.g. patients with liver disease) [17–19]. Therefore, additional parameters should be considered when assessing the PH probability.

With pulsed wave (PW) Doppler interrogation of the RVOT we can measure RVOT acceleration time (RVOT AT) as well as evaluate the signal for mid-systolic notching which reflects an increase in pulmonary vascular resistance [20,21]. A RVOT AT < 105 ms is suggestive of PH [22,23] with high sensitivity and specificity for PH detection [24]. Another important parameter suggestive of PH presence includes early pulmonary regurgitation velocity >2.2 m/s as obtained from CW Doppler across pulmonary valve [25]. Hence, it is recommended to attempt to measure pulmonary regurgitation jet velocity whenever possible.

RV – pulmonary arterial (RV-PA) coupling describes the adaptation of RV to afterload, and it can be assessed by tricuspid annular plane systolic excursion (TAPSE) divided by sPAP, TAPSE/sPAP ratio [26,27]. More recently RV free wall longitudinal strain/sPAP has been proposed as a new echocardiographic index of RV-PA coupling in patients with pre-capillary PH as it was shown that it predicts all-cause mortality and heart-lung transplantation in these patients [28].

Table 1
Commonly used echocardiographic parameters used at diagnosis and follow-up in PH.

Parameter	Abnormal Threshold	Comment
Hemodynamic assessment		
TR peak velocity	>2.8 m/s	Capturing the TR jet velocity may be technically challenging.
Early diastolic PR velocity	>2.2 m/s	
RVOT AT	<105 ms	High sensitivity and specificity for PH detection [24].
RV – PA coupling (TAPSE/sPAP)[a]	>0.32 mm/mmHg predicted low 1-year mortality <0.19 mm/mmHg predicted high 1-year mortality	
RV morphology and function		
RV hypertrophy	>0.5 cm	
LV eccentricity index	>1.1	
RV size	>42 mm base >35 mm mid-cavity	Poor correlation with CMR which is gold standard for RV volume assessment [31].
Basal RV/LV ratio	>1.0	
TAPSE[b]	<18 mm	One of the three strongest predictors of mortality or transplant in patients with PAH [41].
FAC[b]	<35 %	Independent predictor of mortality in PH [36].
RV S'[b]	<9.5 cm/s	Independent predictor of mortality in PH [37].
RIMP	>0.54 by tissue Doppler	Independent predictor of mortality in PH [38].
RV FWS[b]	> −20 % (or <|20 %|)	
3D RVEF	<45 %	Patients with PAH and RVEF <38 % had significantly shorter event-free survival [52].
Other		
RAA[a]	>18 cm^2	
Pericardial effusion[a]		Associated with poor prognosis [38,41].
PA dilation	PA diameter > AR diameter or PA diameter >25 mm	

Abbreviations: TR, tricuspid regurgitation; PR, pulmonary regurgitation; RVOT AT, right ventricular outflow tract acceleration time; PH, pulmonary hypertension; RV, right ventricle; PA, pulmonary artery; TAPSE, tricuspid annular plane systolic excursion; sPAP, systolic pulmonary artery pressure; LV, left ventricle; CMR, cardiovascular magnetic resonance; PAH, pulmonary arterial hypertension; FAC, fractional area change; RV S', tricuspid annulus velocity; RIMP, right ventricular index of myocardial performance; FWS, free wall strain; RVEF, right ventricular ejection fraction; RAA, right atrial area; AR, aortic root.

[a] Parameters used for risk stratification in PAH patients [5].

[b] Parameters with established prognostic value in patients with PAH [138–140].

2.2. Assessment of right heart morphology and function

The features of pressure overload of the RV include RV free wall hypertrophy (>0.5 cm) which can be assessed from the subcostal view, bowing of the interatrial septum into the left atrium (LA) during diastole, and flattening of the interventricular septum (IVS) at end-systole which can be objectively assessed by the use of LV eccentricity index (ratio of the diameter perpendicular to the IVS and the diameter perpendicular to this line) and is abnormal if > 1.1 in systole [29].

Progression of PH may lead to RV dilatation; RV dimensions are most commonly measured from RV focused apical 4-chamber view at base and mid-levels with abnormal cut-offs being >42 mm and 35 mm respectively; although, this only weakly correlates with CMR which is the gold standard for RV size and volume assessment [30,31]. Moreover, a basal RV/LV ratio >1 assessed from apical 4-chamber view is suggestive of RV dilatation. Increase in PA pressure also leads to right atrial (RA) pressure-overload and increased RA pressure is associated with poor prognosis in patients with PAH [32]. Recent meta-analysis showed that increased RA area (RAA)/RA area index is associated with increased risk of poor prognosis [33]. ESC guidelines recognize RAA >18 cm2 as one of the echocardiographic parameters used in risk stratification of these patients [5].

RV systolic function is evaluated using multiple parameters including TAPSE, RV fractional area change (FAC), tricuspid annulus velocity (RV S'), and RV myocardial performance (RIMP) index. All these measures have also been found to be independent predictors of mortality in PH [34–38]. In clinical practice at least two of the parameters should

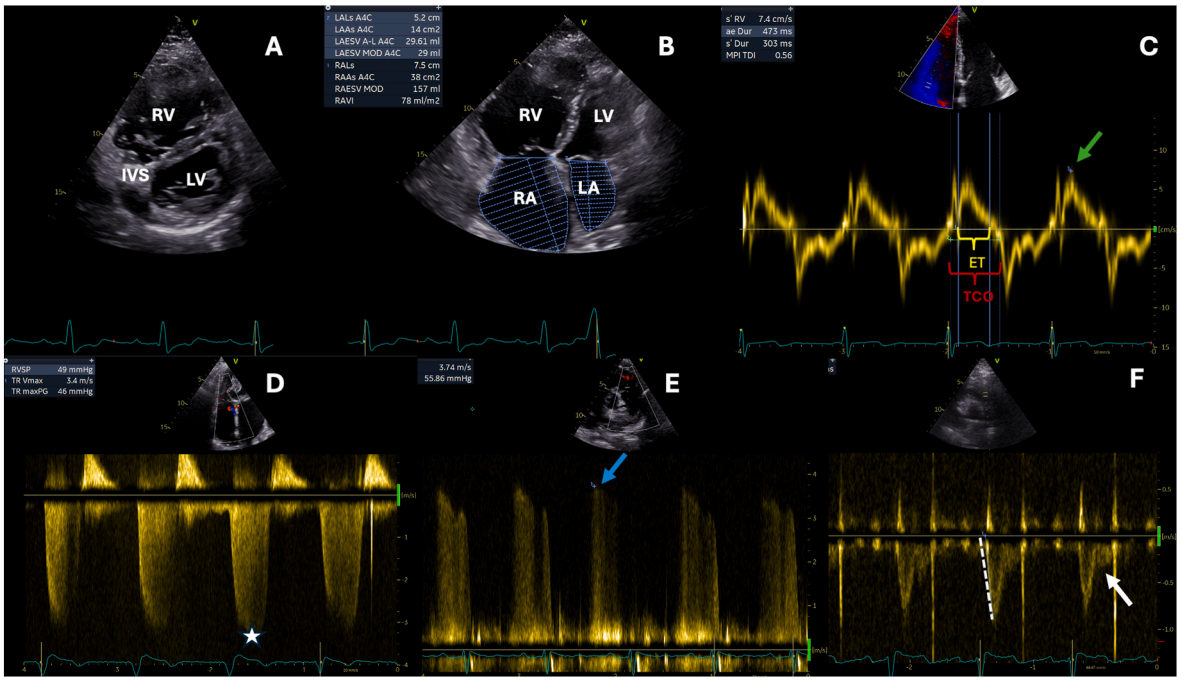

Fig. 1. Echocardiographic Parameters for the Assessment of Right Heart Morphology and Function **A.** Parasternal short axis view showing a dilated right ventricle (RV) and flattened interventricular septum (IVS) due to RV pressure overload. **B.** Four chamber apical view showing dilated right atrium (RA) and right ventricle (RV). **C.** Tissue Doppler imaging (TDI) profile of the tricuspid annulus with the measurement of systolic velocities (RV S′, green arrow) and measurement of ejection time (ET) and tricuspid closure opening time (TCO) used for calculation of the right ventricular index of myocardial performance (RIMP). **D.** Peak tricuspid regurgitation velocity obtained by continuous wave Doppler of the tricuspid valve (asterisk). **E.** Early diastolic pulmonary regurgitation velocity obtained by continuous wave Doppler of the pulmonary regurgitation (blue arrow). **F.** Mid-systolic notching (white arrow) and short pulmonary artery acceleration time (dashed line) of the pulmonary valve pulsed wave Doppler signal. LV, left ventricle; LA, left atrium.

be performed to provide reproducible assessment of the RV function [13]. More novel techniques include estimation of RV ejection fraction (RV EF) by 3D echocardiography and RV strain assessment discussed later. TAPSE is obtained by utilization of M-mode across the lateral tricuspid annulus. It is a reproducible measurement of RV function along the longitudinal axis of contraction and is abnormal if < 18 mm. It correlates well with RVEF measurement on CMR and RHC but must be interpreted with caution in presence of severe TR as it was shown to be less accurate in this setting [39,40]. TAPSE was found to be one of the three strongest predictors of mortality or transplantation in patients with PAH, along with RAA and the presence of pericardial effusion [41]. RV S' is a tissue Doppler derived method which quantifies the velocity of systolic longitudinal movement of the lateral tricuspid annulus. RV FAC measures the difference between end diastolic and end systolic RV area divided by end diastolic area obtained from RV focused apical 4-chamber view. Right ventricular index of myocardial performance (RIMP) is an index of global right ventricular performance, reflecting both RV diastolic and systolic function. It can be acquired by two different methods, either from the PW Doppler imaging of the TV inflow and RVOT or tissue Doppler imaging of the lateral tricuspid annulus. The latter is the preferred method, as it allows the measurement to be performed on a single beat. Right ventricular dysfunction is suggested by a tissue Doppler RIMP >0.54 [13].

2.3. Assessment of LV function

LV diastolic dysfunction is the most common cause of PH (group 2 PH) [42]; therefore, a thorough assessment of the LV is crucial in every patient with suspected PH and should include LV and left atrium (LA) size, LV systolic and diastolic function, and left-sided valve assessment. Atrial and ventricular septal defects should also be excluded. The prevalence of PH increases with advanced valvular disease and it is present in up to 50 % of patients with symptomatic aortic stenosis and up to 70 % of those with severe and symptomatic mitral valve disease [43,44].

2.4. Congenital heart disease

It is not uncommon for patients with CHD to develop PH. [45,46]. It is estimated that approximately 10 % of adults with CHD have PH, which impacts on their quality of life and outcome [46,47]. The frequency of screening of CHD patients for PH depends on the underlying anatomy and it should occur at each echocardiographic evaluation of the patient [48]. However, guideline recommended echocardiographic parameters for PH assessment might not apply in patients with CHD due to complex underlying anatomic and hemodynamic conditions. Good understanding of underlying anatomy is crucial when evaluating CHD patient for the presence of PH. Limitations of specific echocardiographic parameters in CHD are presented in Table 2. Current cut-off values used in general population are also applied in patients with CHD; however, further studies are needed to investigate their applicability to this unique population.

2.5. Advances in echocardiography

With continuous technological progress a better evaluation of myocardial structure and function in terms of motion and deformation is now possible, resulting in a more comprehensive RV assessment. Due to complex RV geometry, three-dimensional echocardiography (3D) can more accurately evaluate RV volumes and EF compared to two-dimensional (2D) echocardiography as it can provide RV evaluation without the geometric assumptions which are integral to 2D echocardiography [49]. Studies have shown that 3D derived EF could be a useful parameter in predicting the clinical outcomes in PH patients[50–52]. Although it was shown that women have slightly higher EF compared to men due to smaller volumes, RV EF <45 % normally reflects RV systolic

Table 2
Limitations of specific echocardiographic parameters in CHD.

Parameter	Limitation in CHD
TR peak velocity	Not useful if: • RV is not directly communicating with pulmonary circulation Not appropriate for PAP assessment if: • RVOTO or peripheral pulmonary stenosis is present
Basal RV/LV ratio	Not useful in patients with: • SRV • Left-to-right shunt • RVOTO or peripheral pulmonary stenosis
LV eccentricity index	Not useful in patients with: • SRV • Left-to-right shunt • RVOTO or peripheral pulmonary stenosis
PA diameter	Not useful in patients with: • ASD • Severe pulmonary regurgitation
RA area	Not useful in patients with: • ASD • TV stenosis or severe regurgitation • Restrictive RV in patients with TOF

Abbreviations: TR, tricuspid regurgitation; RV, right ventricle; RVOTO, right ventricle outflow tract obstruction; LV, left ventricle; SRV, systemic right ventricle; PA, pulmonary artery; ASD, atrial septal defect; RA, right atrium; TV, tricuspid valve; TOF, tetralogy of Fallot.

dysfunction [53]. Although, 3D derived measurements of RV volumes and EF have been validated against MRI, there are several challenges that limit the use of 3D echocardiography, mainly load dependency, and limited accuracy in presence of irregular rhythm and interventricular changes affecting septal motion [54,55]. 3D reconstruction also cannot be adequately performed in cases of poor acoustic windows or significantly enlarged RV [56].

By employing speckle tracking technology, RV myocardial injury due to PH can be evaluated by strain (myocardial deformation) and strain rate (speed at which deformation occurs) [6]. RV free wall strain is preferred over global longitudinal strain (GLS), as it does not include the interventricular septum which may be affected by LV disease [57]. RV strain might be a marker of occult RV dysfunction when other indices of RV dysfunction are still within normal limits [58,59]. It has been shown that RV strain correlates well with the degree of RV dysfunction, PH severity, and is associated with all-cause mortality[60–62]. It is also useful for monitoring the response to therapy [63,64]. Currently, there is no universally agreed reference range; however, RV free wall strain higher than −20 % (lower than 20 % in absolute value) can be regarded as abnormal [65]. The major limitation of strain techniques is the lack of standardization of the method and of the used software, as well as load-dependency [66].

Similarly, RA longitudinal strain is a feasible and reproducible method for assessing RA function. In PH patients, RA longitudinal strains is associated with survival and of prognostic importance, making this another promising parameter to be used in every day clinical practice[67–69].

Echocardiographic pulmonary to left atrial ratio (ePLAR) can be used to non-invasively distinguish between pre-capillary and post-capillary PH [70]. It is estimated by measuring the maximal TVR divided by the tissue Doppler derived E/e' velocity of the mitral annulus. ePLAR of 0.3 ± 0.09 m/s was considered as reference value for normal population; pre-capillary PH patients had significantly higher ePLAR values (0.44 ± 0.22 m/s) compared to post-capillary PH patients (0.20 ± 0.11 m/s, $p < 0.001$). Patients with combined pre- and post-capillary PH had intermediate ePLAR (0.28 ± 0.18 m/s) but still significantly higher compared to patients with isolated post-capillary PH.

2.6. Stress echocardiography

A physiologic response of pulmonary circulation to exercise includes mild increase of PAP that is linear with cardiac output together with decrease in pulmonary vascular resistance secondary to the vasodilation of compliant vessels and/or recruitment of vessels in the upper portion of lungs. Exercise PH is defined by mPAP/cardiac output (CO) slope >3 mmHg/L/min between rest and exercise measured by RHC [71]. Exercise RHC is an invasive diagnostic method, therefore, non-invasive tests, including stress echocardiography are being investigated as potential alternative techniques.

Stress echocardiography is valuable in assessing patients with exertional dyspnoea of unknown aetiology and normal resting echocardiography as it distinguishes between cardiac and noncardiac causes of unexplained dyspnoea [72,73]. Its value in identifying patients with high risk for developing PH has also been studied, including asymptomatic relatives of patients with familial or idiopathic PAH and patients with connective tissue diseases [74–76].

Multiple exercise derived echocardiographic parameters have been described over the years for detection of exercise PH; however, with variable diagnostic accuracy. Stress echocardiography is considered abnormal if the average E/e' ratio at peak stress increase to ≥15, with or without a peak TR velocity >3.4 m/s [72,77,78].

The value of stress echocardiography in prognostic assessment of patients with established PH has also been evaluated. Multiple parameters, including change in sPAP during exercise, change in TAPSE, RV FAC, and S' have been investigated as potential measures of the RV contractile reserve during stress echocardiography[79–82]. Nevertheless, due to the lack of validated criteria and prospective confirmatory data the current guidelines question the clinical value of stress echocardiography for recognition of exercise PH [5]. Reaching universally adopted exercise protocol during echocardiographic assessment is one of the prerequisites for prospective data to be collected in the future [83].

3. Computed tomography

Excellent spatial and temporal resolution as well as ability to perform multi planar reconstruction and expanded field of view make computed tomography (CT) one of the standard investigations during workup of patients with suspected/confirmed PH. Non-contrast chest CT provides information regarding lung parenchyma, while CT pulmonary angiography (CTPA) may detect filling defects and other cardiovascular abnormalities; both helpful to determine the underlying cause of PH. There are also numerous CT findings which may raise the suspicion of PH in patients being evaluated for unexplained dyspnoea.

3.1. CT findings suggestive of PH

Dilatation of the main pulmonary artery (MPA) is one of the best-known CT features of PH (Fig. 2). An MPA diameter >29 mm has 84 % sensitivity and 75 % specificity of PH [84]. Another sensitive predictor of PH is also diameter ratio between MPA and ascending aorta (Ao), with ratio >1.0 correlating with elevated mPAP with 71 % sensitivity and 77 % specificity [85]. The latter was shown to be a stronger predictor of elevated mPAP compared to MPA diameter alone [86]. Nevertheless, diseases unrelated to PH can cause dilatation of aorta, therefore clinically significant disease can still be present despite normal MPA:Ao ratio in the setting of dilated aorta. Although echocardiography and CMR are the methods of choice for evaluation of cardiac structural abnormalities in patients with PH, many cardiac abnormalities associated with PH, such as right ventricular hypertrophy, enlargement of right cardiac chambers and leftward deviation of the interventricular septum can be reliably detected by very fast scanners designed for cardiac CT and ECG-gated acquisitions[87–89].

CT is also an invaluable diagnostic tool for identifying potential

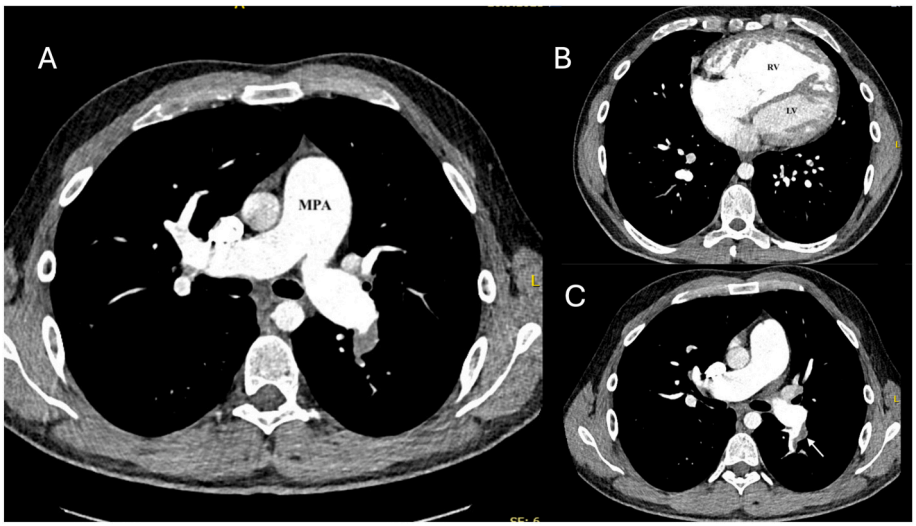

Fig. 2. Computed tomography pulmonary angiography (CTPA) in a patient with Eisenmenger syndrome. **A.** Transaxial view showing main pulmonary artery dilatation. **B.** Transaxial view showing enlarged right heart chambers. **C.** Transaxial view showing thrombus (arrow) in the interlobar portion of the left pulmonary artery.

complications of long-standing pulmonary hypertension, such as compression of mediastinal structures—including the left main coronary artery, left bronchus, and left recurrent laryngeal nerve—by markedly dilated pulmonary arteries. Severe PH, commonly in the context of Eisenmenger syndrome, can be associated with pulmonary arterial calcification, the formation of 'in situ' mural thrombus, and centrilobular nodules thought to represent a combination of the plexiform lesions and cholesterol granulomata that can be appreciated with CT [90].

3.2. Assessment of the underlying cause of PH

Identifying the underlying aetiology should be a key objective in the evaluation of any patient with suspected PH, as it directly influences subsequent management and treatment strategies. When left heart disease is excluded or if present but another cause of PH cannot be excluded then cross-sectional imaging with CT should be performed. Non-contrast CT can reliably evaluate the extent and severity of interstitial lung disease and chronic obstructive pulmonary disease (COPD)[91–93]. Features of pulmonary veno-occlusive disease (PVOD) and pulmonary capillary hemangiomatosis (PCH), including centrilobular ground-glass opacities, septal lines, and lymphadenopathy can be well visualised on CT, making it an invaluable diagnostic method for diagnosing these rare, high-risk disorders [94]. Distinction of PVOD/PCH from other causes of PH is crucial as these patients have a poor prognosis and limited response to medical therapy [95,96]; they should be fast-tracked for transplantation referral where appropriate.

Mediastinal structures are also well visualised on CT and its evaluation can provide important information regarding aetiology and severity of PH. A dilated oesophagus in the setting of PH should raise a suspicion of underlying systemic sclerosis [87]. Although non-specific, findings such as pericardial effusion, lymphadenopathy and reflux of contrast into the hepatic veins may suggest poor prognosis in patients with established PH [97].

CTPA involves intravenous administration of iodinated contrast agent for visualization of pulmonary arteries down to the subsegmental level. It is the gold standard for diagnosing acute pulmonary embolism. Although, the diagnostic accuracy of CTPA for CTEPH is limited, it can detect direct and indirect signs of CTEPH, including filling defects, webs or bands in the pulmonary arteries, pulmonary artery retraction/dilatation, mosaic perfusion and enlarged bronchial arteries[98–100]. Diagnostic accuracy is higher if modern, high-quality multi-detector CT scanners are used and when interpreted by experienced readers [101,102]. It also plays important role in the assessment of CTEPH patients being considered for surgical candidacy [103]. Pulmonary arteriovenous malformations (AVMs) seen in hereditary haemorrhagic telangiectasia can be visualised on CTPA, appearing as nodular opacity with a feeding artery and a draining vein [104].

CTPA can also accurately detect intracardiac shunts, abnormal pulmonary venous return, and patent ductus arteriosus, making it invaluable diagnostic method when evaluating patients with suspected PH associated with CHD [105].

3.3. Advances in CT

Dual-energy CT (DECT) is increasingly used for evaluation of patients with PH. It works by acquiring images at two different energy levels—typically using high- and low-kilovoltage X-ray beams—during a single scan [106]. Different materials in the body, such as iodine, calcium, and soft tissues, absorb X-rays differently at varying energy levels. By analyzing how tissues attenuate these two energy spectra, DECT can differentiate and characterize materials more precisely than conventional CT. In PH, DECT can generate quantitative lung perfusion and pulmonary blood volume (PBV) maps using a standard CTPA protocol. It provides comprehensive morphological information on the vasculature and functional information on perfusion. Although PBV maps are not true perfusion images, since they represent iodine distribution at a single time point during image acquisition, they serve as a reliable surrogate marker for assessing lung perfusion [107]. Studies have shown good correlation between these maps and V/Q scans [108]. In cases of CTEPH, PBV maps have been utilized to evaluate a the suitability for surgery and to identify the presence of bronchial collateral circulation, which is considered a favourable prognostic indicator in these patients [109,110].

Functional Respiratory Imaging (FRI) combines high-resolution CT with computational modelling to deliver size-resolved, region-specific views of the pulmonary vasculature and airways. In PAH, it helps assess disease severity by quantifying distal vessel pruning (i.e., loss of small distal vessels) and proximal dilation, reflecting hallmark vascular remodelling. FRI also supports monitoring of disease progression through serial small-vessel measurements, offering potential prognostic benefit, and aids treatment evaluation by capturing early regional vascular responses that correlate with hemodynamic improvements [141].

4. Cardiovascular magnetic resonance

CMR is non-invasive, radiation free, and sensitive to changes in anatomy and function making it ideal for the long-term follow up of patients with PH. It offers comprehensive and multi-metric evaluation of the RV and other cardiac chambers as well as pulmonary blood flow assessment and tissue characterisation. Not only can it reveal the underlying aetiology for PH, including accurate determination of underlying CHD (Fig. 3), but CMR derived RV morphology and function has a proven role in determining prognosis. It is also used to monitor response to medical therapies [5]. In Table 3 we provide main advantages and disadvantages of imaging modalities including CMR.

4.1. CMR assessment of ventricular morphology and function

Although PH is a disease of the pulmonary vasculature, it is the RV function that determines clinical outcomes. CMR provides gold standard measurements of RV mass, volume, and EF that are not limited by poor acoustic windows, an important advantage in this cohort of patients in whom a significant proportion may have advanced lung disease and musculoskeletal abnormalities. In the early disease phase of PH, RV hypertrophy occurs as an appropriate adaptation to the increase in afterload to maintain RV function. As the disease advances, the increase in RV mass is outstripped by the increase in RV volume as the RV fails to adequately adapt to the elevated afterload. Low RV mass/RV volume ratio is associated with increased risk of mortality [111,112].

RV EF is a powerful predictor of outcomes in patients with PAH [113–115]. Together with RV volumes and stroke volume index (SVI), it is one of the three essential cMRI measurements with prognostic value in PAH that are included in current risk scores [5]. RVEF also closely tracked symptoms reported by the patients on Em-PHasis-10 score questionnaires [114]. In patients on PH medical therapy, a reduction in RVEF was significantly associated with mortality irrespective of PVR [116]. This suggests that RV dysfunction prevails over PVR as a prognostic marker in PAH supporting the use of RVEF to monitor treatment response. [117]. Furthermore, RVEF is also used as a surrogate endpoint for mortality for PH drug trials given its prognostic value. Four-dimensional (4D) flow CMR has the advantage of being able to potentially quantify RV diastolic dysfunction [118,119]. Reduced biventricular circumferential and longitudinal strain measured by feature tracking CMR was found to be associated with reduced RVEF and predicted mortality [120,121].

4.2. Exercise CMR

As PH progresses, the RV maladapts by increasing in size which can lead to a reduction in stroke volume (SV). Exercise CMR can help determine whether patients have adequate RV inotropic reserve to meet the increased cardiac output requirements by measuring the increase in RV SV during exercise when compared to rest [122].

4.3. The left ventricle, septal motion, and fibrosis

Although predominantly thought of as a disease that affects the right heart, direct ventricle-ventricular interaction through the shared interventricular septum, has consequential deleterious effects on the left heart. The left-ward shift of the septum caused by raised RV pressures, impairs LV filling, and can result in a small LV cavity. Small CMR derived LV volumes and lower LV SV have been shown to predict mortality in PAH although not independent of RVEF [115]. Patients with Eisenmenger syndrome and biventricular impairment measured from CMR had the worst survival when compared to patients with either reduced RVEF or LVEF alone. When tested against echocardiographic, blood markers including natriuretic peptides, and exercise, only CMR derived impaired RVEF and LVEF, and resting oxygen saturations were predictive of outcome suggesting that CMR derived EFs may be more sensitive than other markers [123].

The degree of ventricular septal shift has been quantified using a septal curvature ratio, measured from mid-ventricular short axis cines at peak systole [124]. This was shown to correlate closely with indexed PVR (iPVR) measured using hybrid MR and RHC technique and reflected acute changes in iPVR secondary to pulmonary vasodilator reversibility

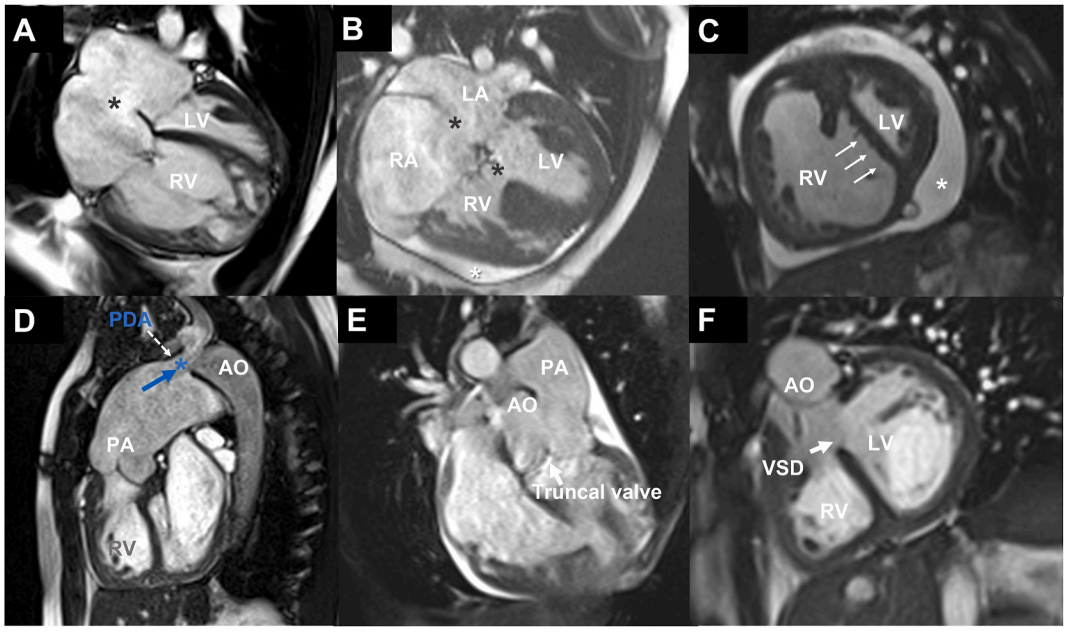

Fig. 3. Pulmonary hypertension secondary to unrepaired congenital heart disease in adults **A.** Pulmonary hypertension secondary to a large secundum atrial septal defect (asterisk). The right ventricle (RV) is markedly dilated and there is moderate tricuspid regurgitation. **B.** Unrepaired atrioventricular septal defect (asterisk) with Eisenmenger syndrome. **C.** Severely dilated RV with systolic flattening of the interventricular septum in keeping with RV pressure load in a patient with a large atrial septal defect. A moderate size pericardial effusion (asterisk) is present, which is a marker of poor prognosis. **D.** Large patent ductus arteriosus (PDA) with Eisenmenger syndrome (blue arrow). **E.** Unrepaired type I truncus arteriosus in a 16-year-old male with Eisenmenger syndrome. **F.** Large non-restrictive outlet ventricular septal defect (VSD) with Eisenmenger syndrome.

Table 3
Advantages and disadvantages of imaging modalities in pulmonary hypertension.

Imaging Modality	Advantages	Disadvantages
Echocardiography	- Widely available and inexpensive - Provides detailed info on right/left heart morphology and function - Non-invasive haemodynamic estimates (TRV, sPAP) - Useful for identifying LHD, CHD, and causes of PH - Guides probability assessment of PH and further investigations	- Cannot confirm PH (RHC required) - No single reliable parameter; requires multiparametric assessment - sPAP/RAP estimates subject to inaccuracies - Over/underestimation possible with TR severity or artefacts - Limited reliability of Doppler signs (exercise PH, diastolic dysfunction)
Computed Tomography (CT)	- Detects structural signs of PH (PA dilation, RV enlargement) - Helps determine cause of PH (e.g., lung disease, PVOD/PCH) - CTPA useful for detecting CTEPH (thrombus, webs, bands) - Can identify cardiovascular abnormalities (shunts, PAVMs) - DECT may add perfusion info - DSA helpful for confirming CTEPH and guiding BPA	- Radiation exposure - Requires contrast (risk in renal dysfunction/ allergy) - CTPA diagnostic accuracy not perfect; reader-dependent - DECT value not fully validated - DSA invasive
Cardiac Magnetic Resonance (CMR)	- Accurate and reproducible for atrial/ ventricular size and function - Provides myocardial strain analysis - Quantifies blood flow, SV, shunts, retrograde flow - Comprehensive imaging with contrast MR angiography and perfusion - Sensitive to early PH and CHD detection	- No established method to estimate PAP - Expensive and less available - Time-consuming - Not suitable for all patients (implants, claustrophobia)

Abbreviations: BPA: Balloon Pulmonary Angioplasty, CHD: Congenital Heart Disease, CMR: Cardiac Magnetic Resonance, CO: Cardiac Output, CT: Computed Tomography, CTEPH: Chronic Thromboembolic Pulmonary Hypertension, CTPA: Computed Tomography Pulmonary Angiography, DECT: Dual-Energy Computed Tomography, DSA: Digital Subtraction Angiography, LHD: Left Heart Disease, LV: Left Ventricle, PA: Pulmonary Artery, PAH: Pulmonary Arterial Hypertension, PAP: Pulmonary Arterial Pressure, PAVMs: Pulmonary Arteriovenous Malformations, PCH: Pulmonary Capillary Hemangiomatosis, PE: Pulmonary Embolism, PH: Pulmonary Hypertension, PVOD: Pulmonary Veno-Occlusive Disease, RAP: Right Atrial Pressure, RHC: Right Heart Catheterization, RV: Right Ventricle, RVOT: Right Ventricular Outflow Tract, SV: Stroke Volume, TR: Tricuspid Regurgitation, TRPG: Tricuspid Regurgitation Pressure Gradient, TRV: Tricuspid Regurgitation Velocity, sPAP: Systolic Pulmonary Arterial Pressure.

testing in real time, showing its potential to track response to treatment [124].

Late gadolinium enhancement (LGE) CMR defined focal myocardial fibrosis has been associated with mortality in acquired heart disease although this is less proven in CHD due to the lack of prospective data. In PH, the presence of LGE at the RV/LV septal insertion points is very common, affecting at least 80 % of patients and in our experience, septal insertion point LGE is commonly seen even in normal hearts [125,126]. This is likely to reflect blood pooling at the insertion points rather than pathological fibrosis, caused by myocardial disarray that occurs as the RV and LV fibres cross each other. In PH, insertion point LGE often appears more accentuated than in normal hearts which may be due to increased blood pooling at this location due to RV hypertrophy (Fig. 4). In ~30 % of PH patients, LGE at the insertion points extends into the mid-interventricular septum [125]. Patients with mid-septum extension of LGE were found to have worse survival although not when tested against conventional risk factors in bivariable analysis [125]. LGE confined to the insertion points was not found to be predictive of outcome thereby supporting our view that this finding is non-specific and benign [125]. In patients with Eisenmenger syndrome, LGE was found to be present most commonly in the RV (in ~70 %) and RV trabeculations, but also found in the LV. LGE in Eisenmenger syndrome patients was not associated with function, exercise capacity or survival [127].

Diffuse myocardial fibrosis detected using T1 mapping has been associated with cardiac events in patients with acquired and CHD. In PH, elevated native T1 typically occurs at the insertion points similar to LGE and was not found to be predictive of outcome or useful in distinguishing PH severity. Similar to LGE, elevated native T1 is also likely to reflect blood pooling at the insertion points rather than myocardial fibrosis (Fig. 4). T1 mapping indices have not shown correlation with resting oxygen saturations in patients with Eisenmenger syndrome [128]. Further work is required to investigate the value of LGE and T1 mapping in patients with PH and other CHD lesions. T1 mapping for application in the RV is not yet ready for clinical use and requires further sequence development.

4.4. CMR assessment of the pulmonary vasculature and function

Unlike in echocardiography, PAP cannot be estimated using the peak recorded velocity of the tricuspid regurgitant jet as the core of the jet is dispersed and too small to measure [125]. Indirect CMR markers of reduced pulmonary vascular compliance have been identified. The extent of high signal intensity in the pulmonary arteries on black blood images reflecting slow blood flow correlates positively with the severity of PH [129] (Fig. 5). Two-dimensional phase contrast velocity CMR can be used to identify a typical 'mid-systolic notch' in the pulmonary artery flow curve that represents arterial wave deflections caused by abrupt changes in vessel area and compliance [130]. Given that PH is characterised by widespread pulmonary vascular remodelling, it has been postulated that cumulatively, abnormal reflections may be an additional source of RV afterload. A CMR technique using wave intensity analysis has been shown to non-invasively quantify wave reflections and differentiate between healthy and disease states [130]. Pulmonary artery expansibility can be measured from CMR cines in the transaxial plane by tracing maximum pulmonary artery area change in systole when compared to diastole. Pulmonary artery distensibility measured using this method was shown to be a predictor of outcome in patients with PH [129].

Four-dimensional (4D) flow has been used to identify abnormal flow signatures in the pulmonary arteries in PH patients. The presence of a persistent vortex in the pulmonary artery has been shown to correlate with mPAP and reduced pulmonary artery wall stress correlated with PH [131] (Fig. 6). Although 4D flow sequences have been available for the past decade, it is not yet widely applied in clinical protocols as the acquisition and post-processing is time consuming. Its current role, therefore, is more confined to the research arena.

4.5. MR-augmented cardiac catheterization and MRI guided catheterization

During RHC, calculation of cardiac output using Fick's principle is subject to inaccuracies as the formula requires multiple measures.

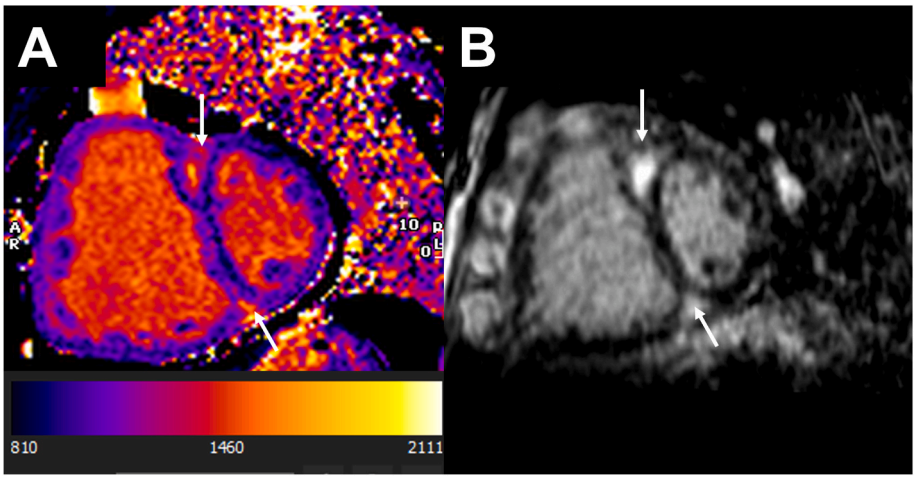

Fig. 4. Tissue characterisation using cardiovascular magnetic resonance (CMR). **A.** A native T1 map at mid-ventricular short axis level used to determine the presence of diffuse myocardial fibrosis in the left ventricle. High T1 signal can be seen in the superior and inferior RV/LV septal insertion points (arrows). **B.**A similar image plane shows prominent LGE at the superior and inferior RV/LV septal insertion points. Elevated native T1 and LGE at the insertion points is common in pulmonary hypertension and likely reflects blood pooling as opposed to pathological fibrosis in pulmonary hypertension.

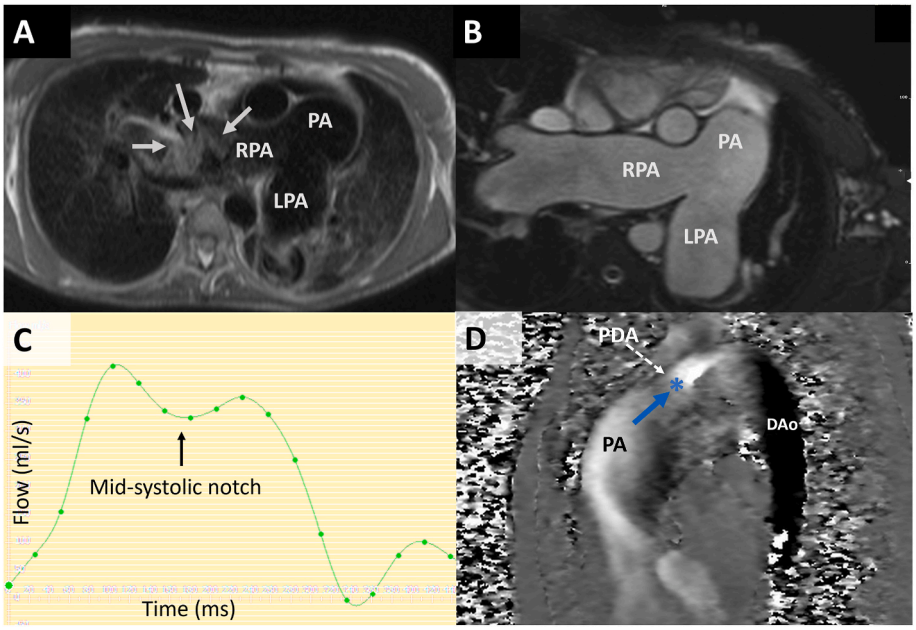

Fig. 5. Abnormalities in pulmonary blood flow identified with cardiovascular magnetic resonance (CMR). **A.** Increased signal in the branch pulmonary artery and subsegmental branches indicating slow blood flow on black blood CMR images seen on HASTE (Half fourier Single-shot Turbo-spin Echo) image. **B.** Corresponding still from a cine image of dilated central and sub-segmental pulmonary arteries. **C.** Mid-systolic notch in the pulmonary arterial wave form derived from through-plane phase encoded velocity mapping. The arterial wave reflection arrowed is caused by abrupt changes in pulmonary vessel area and compliance which contribute to right ventricle afterload. This feature is indicative of pulmonary hypertension. **D.** In-plane 2D phase contrast image of flow showing right to left flow from the main pulmonary artery to the descending aorta (blue arrow) through the patent ductus arteriosus (blue asterisk).

Thermodilution, as an alternative to the Fick principle, may also be less accurate in the presence of a shunt or valvar regurgitation. Magnetic resonance (MR)-augmented cardiac catheterization, whereby a right heart catheter is used to measure mean pulmonary artery and pre-capillary wedge pressure during simultaneous CMR flow acquisitions, may avoid inaccuracies associated with invasive flow quantification by combining gold standard flow measurement from CMR with invasive pressure [132]. This procedure can also allow for vasodilatory testing using inhaled nitric oxide and oxygen to assess pulmonary vascular reactivity and additionally measurement of pressure-volume loops using CMR RV volumes. MR-augmented pressure-volume loop assessment may aid more accurate RV and pulmonary vascular assessment in PH which may be particularly useful in cases where there are multiple sources of blood flow or a combination of shunts.

4.6. MR angiography

Although MR angiography has lower spatial resolution than when compared to CT pulmonary angiography, MR angiography avoids ionising radiation and nephrotoxic contrast agent. Patients with chronic cyanosis such as in Eisenmenger syndrome are predisposed to abnormalities in coagulation and may develop in situ thrombus, typically in at-risk areas such as the pulmonary arteries [128]. MR angiography is highly sensitive and specific to detecting proximal pulmonary artery

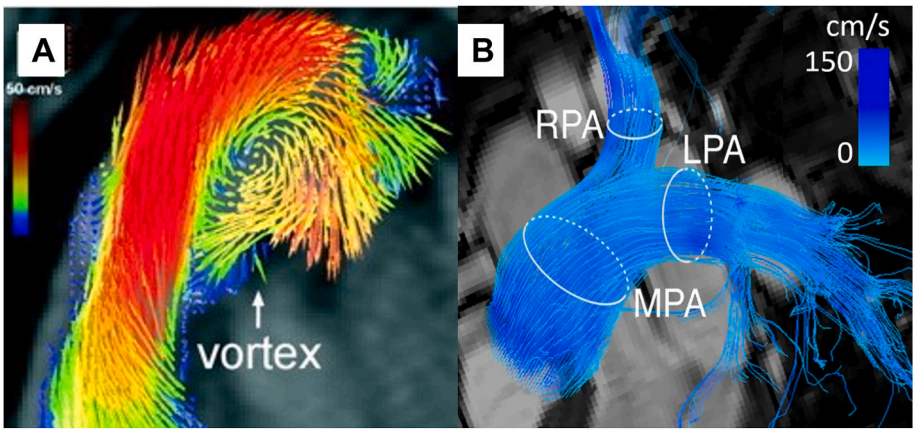

Fig. 6. Four-dimensional (4D) flow patterns in the pulmonary artery and quantification Image adapted from Reiter et al., 2008 (A) and Bissel et al. (2023). **A.** Four dimensional (4D flow map demonstrating a vortex in the forward direction of blood flow in the main pulmonary artery (MPA) in late systole. The presence of vortices in the MPA is associated with pulmonary hypertension, with the duration of the vortex existence relating to mean pulmonary artery pressures. **B.** Examples of position of regions of interest that can be used to quantify blood flow in the MPA, right pulmonary artery (RPA) and left pulmonary artery (LPA).

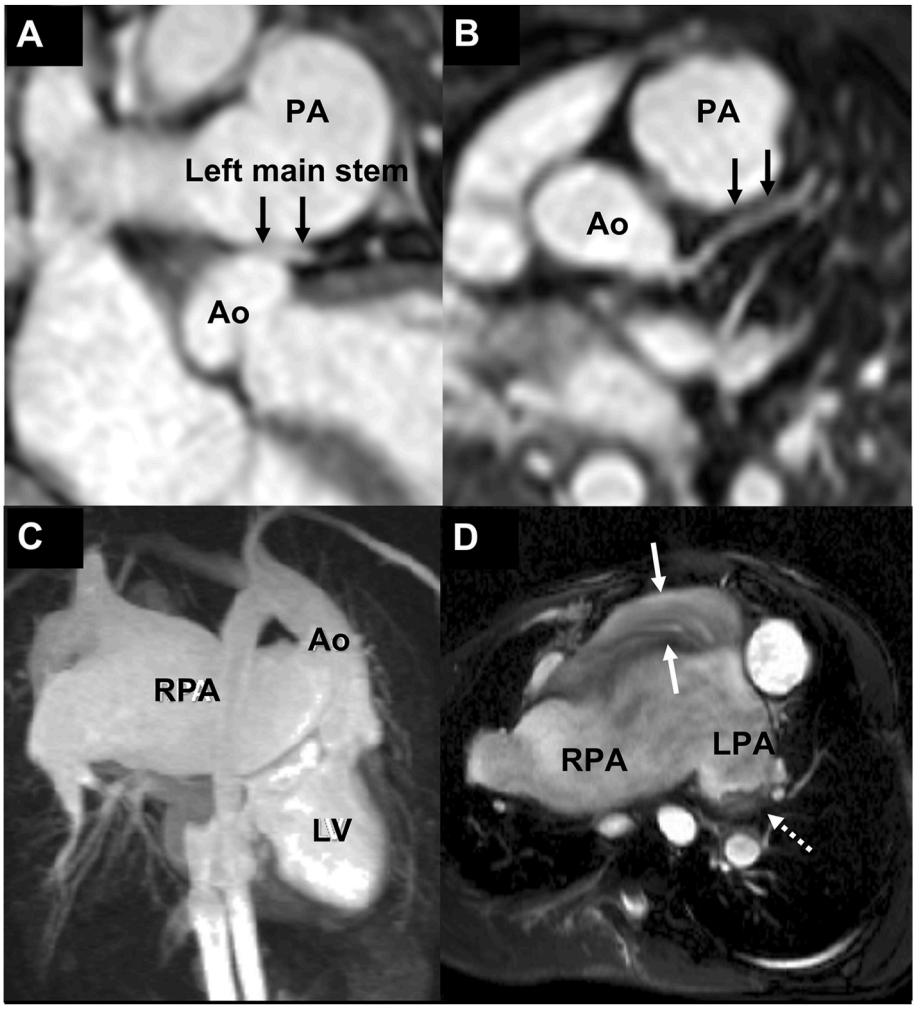

Fig. 7. Three-dimensional non-contrast SSFP angiography and contrast enhance angiography demonstrating complications from PAH. Three-dimensional (3D) non-contrast dimensional balanced Steady State Free Precession (bSSFP) angiography in a patient demonstrating compression of the left main stem coronary artery between a grossly dilated pulmonary artery (PA) and aorta (Ao) as a complication of pulmonary hypertension **(A** and **B**). **C.** Contrast-enhanced magnetic resonance (MR) angiography in a different patient demonstrating the lumen of a grossly dilated right pulmonary artery (RPA) with thrombus in-situ. **D.** Still cine image of corresponding RPA with large thrombus seen proximally (white arrow) with in-situ thrombus in the left pulmonary artery (LPA) (dotted white arrow).

thrombus and can also identify mural thrombus in such patients (Fig. 7). Three-dimensional (3D) balanced Steady State Free Precession (bSSFP) angiography can be performed without contrast administration and correlates well with CT images. Although not first line, it may be useful in situations where there is contraindication to CT contrast or if CT angiography is suboptimal.

4.7. Advances in CMR

Several emerging CMR techniques have shown cross-sectional associations with PH or markers of deterioration, however, their added value to routine clinical practice is yet to be proven. Abnormalities in RV kinetics have been identified using 4D CMR flow and have been found to be associated with adverse RV remodelling and impaired exercise capacity [133]. Diffusion tensor imaging CMR can gain insight into abnormalities in RV myocadial architecture in PH although so far only tested in animal studies [134]. Direct measurement of strain in the plane of tissue displacement is possible with displacement encoding with stimulated echo (DENSE) CMR who ever requires further investigation in patients with PH to understand its clinical value.

We anticipate that artificial intelligence (AI) may have a role in the next frontier of the personalised care delivery to PH patients. AI may help to make CMR acquisition and image analysis faster, which is particularly useful for measurement of RV volumes where the RV can be challenging to contour due to its unique geometry [135]. Different patterns of RV contraction in systole identified using machine learning survival models have been found to predict survival independent of conventional risk factors [136]. Recently, a machine learning workflow was able to differentiate between patients with and without PH from CMR cine images suggesting the potential role of AI in screening for PH [137].

5. Conclusion

Non-invasive imaging plays a central role in the evaluation and management of PH, complementing right heart catheterization by enabling earlier diagnosis, precise classification, risk estimation, and effective monitoring. Echocardiography remains the first-line modality due to its accessibility and diagnostic utility, enhanced by recent advances such as 3D and speckle-tracking techniques. CT offers detailed visualization of pulmonary vasculature and lung parenchyma, aiding in subtype differentiation, while CMR provides comprehensive, radiation-free assessment of cardiac structure, function, and pulmonary flow, making it useful for long-term follow-up. Together, these modalities support a multimodal approach that improves diagnostic accuracy, guides therapy and enhances patient outcomes.

CRediT authorship contribution statement

Polona Kačar: Writing – original draft, Conceptualization. **Katja Prokšelj:** Supervision. **Sarah Ghonim:** Writing – original draft. **Thomas Semple:** Conceptualization. **Sonya V. Babu-Narayan:** Writing – original draft. **Heba Nashat:** Validation, Conceptualization. **Stephen J. Wort:** Writing – review & editing, Validation, Conceptualization. **Michael A. Gatzoulis:** Writing – review & editing, Validation, Supervision, Conceptualization. **Margarita Brida:** Writing – review & editing, Validation, Conceptualization.

Declaration of competing interest

The authors report no relationships that could be construed as a conflict of interest other t.

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