

[Review article]

Practical recommendations on the use of echocardiography to assess pulmonary arterial hypertension – a Belgian expert consensus endorsed by the Working Group on Non-Invasive Cardiac Imaging

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Abstract Pulmonary hypertension (PH) is defined by a sustained increase in mean pulmonary arterial pressure > 25 mmHg. Due to its widespread availability, echocardiography (ECHO) is used as the first-line imaging modality to detect pulmonary PH and assess right ventricular (RV) function in daily routine. As such, ECHO is the key examination to detect the presence of PH, to provide valuable prognostic information and to give an orientation to therapeutic strategies. In addition to detection and screening, ECHO also provides clues for the differential diagnosis of PH. The present document, based on a consensus of experts, provides practical recommendations for the use of ECHO in the evaluation of PH and of its consequences on the right ventricle.

Keywords Pulmonary hypertension – Doppler echocardiography – right ventricle.

INTRODUCTION

Although not diagnostic of the condition, echocardiography (ECHO) is the best non-invasive screening tool currently available to detect pulmonary hypertension (PH) and assess right ventricular (RV) function in daily practice. ECHO helps in the detection and screen-

ing of PH, but also provides clues for the differential diagnosis, such as confirmation of valvular heart disease, of an intracardiac shunt and assessment of left atrial and ventricular function^{1,2,3}. In addition, several ECHO indices are important in risk stratification and markers of outcome of the most severe forms of PH. Current guidelines recommend ECHO in all stages of PH⁴, from diagnosis to follow-up assessment. This document provides practical recommendations for the use of ECHO in the evaluation of PH and its consequences on the RV.

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Definition and classification of pulmonary hypertension

PH is defined by a sustained increase in mean pulmonary arterial pressure (mPAP) > 25 mm Hg at rest

as assessed by right heart catheterization (RHC). The haemodynamic classification of PH is also based on the invasive measurement of pulmonary capillary wedge pressure (PCWP) and distinguishes two main groups: (A) pre-capillary PH defined by a PCWP \leq 15 mm Hg, and (B) post-capillary PH with PCWP $>$ 15 mm Hg. PH can be found in more than 30 medical conditions.

The current clinical classification⁵ established in Dana Point identifies five different groups: *group 1*, pulmonary arterial hypertension (PAH); *group 2*, pulmonary hypertension owing to left heart disease; *group 3*, pulmonary hypertension owing to lung diseases and/or hypoxaemia; *group 4*, chronic thromboembolic pulmonary hypertension; and *group 5*, pulmonary hypertension with unclear and/or multifactorial mechanisms (table 1). Precapillary PH is characteristic of groups 1,

3, 4 and most cases of group 5, whereas postcapillary PH is only found in group 2.

Group 1, Pulmonary arterial hypertension

Pulmonary arterial hypertension (PAH) is characterised by a progressive increase of the pulmonary vascular resistance of small pulmonary vessels, rapidly leading to right ventricular failure and premature death. PAH comprises various conditions sharing similar clinical/haemodynamic manifestations and pathological changes of the arterial lung circulation.

Half of the patients included in registries present with idiopathic (the most frequent aetiology), heritable and/or anorectic-drug induced PAH⁶. The other half consists in PAH associated with other conditions, such as con-

Table 1 Updated clinical classification of pulmonary hypertension (adapted from reference 5)

Group 1. Pulmonary arterial hypertension (PAH); category pre-capillary PH.
1.1. Idiopathic
1.2. Heritable
1.3. Induced by drugs and toxins
1.4. Associated with PAH (connective tissue diseases, HIV infection, portal hypertension, congenital heart disease, schistosomiasis, chronic haemolytic anaemia)
1.5. Persistent pulmonary hypertension of the newborn
Group 1. Pulmonary veno-occlusive disease and pulmonary capillary haemangiomatosis; category pre-capillary PH.
Group 2. Pulmonary hypertension due to left heart disease; category post-capillary PH.
2.1. Systolic dysfunction
2.2. Diastolic dysfunction
2.3. Valvular disease
Group 3. Pulmonary hypertension due to lung diseases and hypoxaemia; category pre-capillary PH.
3.1. Chronic obstructive pulmonary disease
3.2. Interstitial lung disease
3.3. Other pulmonary diseases with mixed restrictive and obstructive patterns
3.4. Sleep-related breathing disorder
3.5. Alveolar hypoventilation disorders
3.6. Chronic exposure to high altitudes
3.7. Developmental abnormalities
Group 4. Chronic thromboembolic pulmonary hypertension; category pre-capillary PH.
Group 5. PH with unclear or multifactorial mechanisms; category pre-capillary PH.
5.1. Haematologic disorders: myeloproliferative disorders, splenectomy
5.2. Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangiomyomatosis, neurofibromatosis, vasculitis
5.3. Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
5.4. Others: tumoural obstruction, fibrosing mediastinitis, chronic kidney failure on dialysis

nective tissue diseases (in 70% due to systemic sclerosis), HIV infection, portal hypertension, congenital heart disease, schistosomiasis, or chronic haemolytic anaemia⁶. Finally, a subcategory (group 1') includes pulmonary veno-occlusive disease and pulmonary capillary haemangiomas.

The minimal estimated prevalence of PAH is 15 cases/million but may be as high as 26 to 54 cases per million^{6,7}. The minimal incidence of PAH is 2.4 cases/million/year⁶ but has been found as high as 7 cases/million/year⁷. PAH appears to be slightly more prevalent in women, although this has recently been questioned.

Group 2, Pulmonary hypertension due to left heart disease

Left heart diseases (LHD), including systolic and/or diastolic left ventricular (LV) dysfunction, represent the most frequent causes of PH. In most cases, a pure passive post-capillary PH is observed due to increased PCWP. However, a small subset of patients may present with significant PH that is not accounted for by the increase in PCWP, determined by a transpulmonary pressure gradient (TPG: mPAP – PWP) > 12 mm Hg⁸. Such an increase in TPG is explained by an increased vasomotor tone of the pulmonary arteries and/or structural obstructive remodelling of the pulmonary resistance vessels process^{8,9}. The presence of severe PH in LHD is associated with a poor prognosis, irrespective of the underlying condition¹⁰. Its detection is also an important step that must be addressed in the heart transplant candidates' assessment.

Group 3, Pulmonary hypertension due to lung diseases and hypoxaemia

PH group 3 can be found in any respiratory disorder although the underlying pathophysiology is largely unknown. However, mechanisms involved in this form of PH include among others hypoxic vasoconstriction, mechanical stress of hyperinflated lungs, loss of capillaries, inflammation, or endothelium-derived vasoconstrictor–vasodilator imbalance processes^{4,8}.

PH may be present in patients with idiopathic pulmonary fibrosis, chronic hypoxaemia or chronic obstructive pulmonary disease (COPD). However, severe PH is uncommon in COPD patients (< 1%) and the majority of patients with severe PH have an additional cause of pulmonary arterial pressure increase, such as LV disease or sleep apnoea syndrome¹¹. In COPD patients, presence of PH is linked with shorter survival and more frequent episodes of exacerbation^{4,8}.

Group 4, Chronic thromboembolic pulmonary hypertension (CTEPH)

Chronic and mechanical obstruction of central and/or distal pulmonary arteries by thromboembolic material leads to PH. Most patients with CTEPH experienced previous acute pulmonary thromboembolism (74.8%) and deep vein thrombosis (56.1%)¹². In addition, a further potential cause of PH was detected in 20.9% of patients, COPD being the most frequently associated condition (9.5%)¹².

It has been hypothesised that group 4 PH can be considered as a primary pulmonary vascular arteriopathy with endothelial dysfunction, triggered by overt or occult pulmonary embolism, leading to secondary in situ thrombosis¹³.

CTEPH occurs within 2 years after acute pulmonary embolism in 0.5–3.8% of cases^{14,15}.

Group 5, Pulmonary hypertension with unclear or multifactorial mechanisms

This group includes several remaining forms of PH with unclear aetiology or multifactorial causes. PH can be caused by chronic haematologic disorders (i.e. chronic myeloid leukaemia), systemic disorders (i.e. sarcoidosis), metabolic disorders (i.e. type Ia glycogen storage disease), or miscellaneous conditions (i.e. mediastinal fibrosis).

A roadmap to the diagnosis of PH

Clinical presentation /When to suspect PH?

Although PH may be asymptomatic in its initial stages, exercise-related dyspnoea is the initial presenting symptom in almost all patients, which can be accompanied by fatigue and dizziness. Signs of RV failure, such as peripheral oedema and ascites, syncope and decompensated RV failure are only present at a later stage of the disease.

The signs and symptoms of PAH are, however, non-specific and may be subtle, which explains why the disease is often diagnosed too late; a delay between symptom onset and diagnosis of 27 months has been reported⁶. PAH should be considered early in patients with connective tissue diseases, especially in systemic sclerosis. The severity of the disease and the availability of specific therapies justify screening of PH even in asymptomatic cases in patients with this condition¹⁶. Detection of PH should also be performed in symptomatic individuals with another condition or risk factor associated with PAH, such as liver diseases, HIV infection and congenital heart diseases.

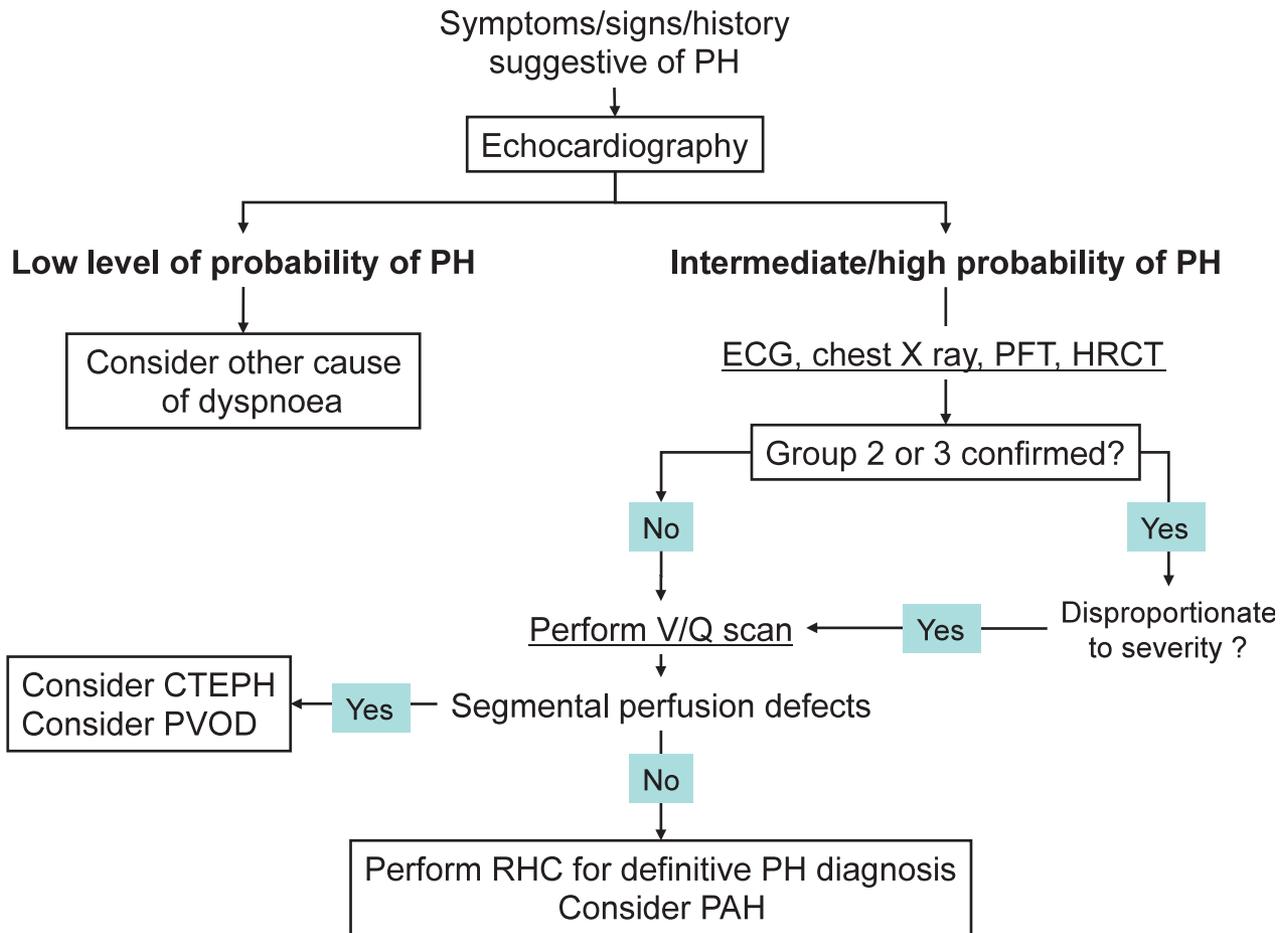


Fig. 1 Diagnostic algorithm of pulmonary hypertension (PH)

Diagnostic algorithm /How to detect PH in clinical practice

PH should be suspected in any case of unexplained dyspnoea. Simple non-invasive tests should be performed because they allow to quickly identify the most common causes of PH. They include an electrocardiogram (ECG), a chest X-ray, and pulmonary function tests (including measurement of the diffusion capacity for carbon monoxide – DLCO). Arterial blood gases may be performed at this stage to identify hypoxaemia and/or hypercapnia that are typical features of lung disorders leading to group 3 PH. The diagnostic algorithm of PH is presented in figure 1.

Review of echocardiographic variables

Transthoracic ECHO is the “gate-keeper” to the differential diagnosis of PH and the assessment of its impact

on the RV. ECHO allows the following: (1) detection of increased right chambers pressures; (2) evaluation of RV changes as a consequence of increased afterload; (3) assessment of LV morphology, function and detailed analysis of cardiac valves; (4) detection of intracardiac shunts and congenital cardiac defects^{1-3,17}. The diagnosis of PH is based on a probability combining ECHO criteria, other signs suggestive of PH or RV strain, signs/symptoms and the presence of risk factors for PAH. In routine, it is advocated to perform a comprehensive ECHO in all patients. As a first approach, recommended basic ECHO parameters to be measured in patients with suspected PH are listed in table 2. A suspicion of PH can reasonably be excluded when these variables fall within the normal range. However, when at least one of these parameters is found abnormal, the present consensus of experts recommends performing an advanced ECHO examination, which includes more quantitative assess-

Table 2 Basic ECHO features for standard examination useful for exclusion of PH.

Variable	Normal findings	Limitation through dependence on	Comment
Full classical echo examination	Normal	--	Normal LV function and shape, normal valves, absence of intracardiac shunt, etc.
Cardiac chambers "eyeballing"	Normal	Image quality Observer experience	Normal wall motion
Right and left ventricular dimensions	RV < LV	LV size and function must be normal	Apical 4-chamber view
Pericardial effusion	Absent	Not reported	Strong prognostic factor: presence is linked to poor prognosis
Tricuspid regurgitation jet velocity (TRV)	< 2.8 m/s	Dependent on age gender, body mass index, massive tricuspid regurgitation	End-expiratory measurement
Inferior vena cava (IVC) diameter	< 2.1 cm	Body position	End-expiratory and inspiratory measurement. Incumbent position
Pulmonary acceleration time (ACT)	> 100 ms	Only when heart rate < 100 beats/min	If heart rate > 100 beats/min, acceleration time is not reliable Low cardiac index may prolong the acceleration time

LV: left ventricle, RV: right ventricle.

Table 3 Advanced ECHO features for comprehensive assessment of PH.

Variable	Normal findings	Limitations through dependence on	Comment
Assessment of haemodynamics			
Mitral E/e'	< 8	Calcification of the annulus Age, mitral valve prosthesis, annular ring	Use the mean between septal and lateral values
Cardiac index	2.2 l/min/m ²	Accurate LVOT measurement Aortic valve regurgitation	--
Inferior vena cava respiratory collapse	> 50%	Body position	Incumbent position Estimation of RA pressure: -Collapse > 50%: 6-10 mm Hg -Collapse < 50%: 10-15 mmHg -No collapse: > 15 mmHg
Pulmonary regurgitation velocity	< 1.2 m/s	Dip in PR jet signal related to RA kick	End-diastolic measurement Evaluate both morphology and velocity Explore further in case of high values
Assessment of chamber size and function			
Septal motion "eyeballing"	Normal	Conduction disorders Volume overload	Visual estimation
Tricuspid annular plane systolic excursion (TAPSE)	> 20 mm	Preload Severe TR LV dysfunction Severely enlarged RV Overall cardiac motion	Surrogate marker of the RV longitudinal function
Peak systolic velocity of tricuspid annulus (s')	> 12 cm/s	Preload Severe TR LV dysfunction Severely enlarged RV Angle dependent	Tissue Doppler imaging
Right ventricular fractional area change (RVFAC)	> 32%	Preload Foreshortening of the RV apex RV rotation RV trabeculation/geometry	--
Left ventricular eccentricity index	1	--	End-systolic and end-diastolic measurements at end expiratory phase Values ≠ 1 are abnormal.

LVOT: left ventricular outflow tract, RAP: right atrial pressure, RV: right ventricle, TR: tricuspid regurgitation.

ment of cardiac haemodynamics, chamber sizes and functions (table 3). It should be reminded that ECHO has several limitations in terms of accuracy of the assess-

ment of left heart filling pressures. As a result, a right heart catheterization is mandatory to establish the diagnosis of PH, especially in the case of PAH. In addition,

it is an essential tool to analyse the vasoreactivity of the pulmonary circulation at diagnosis (in idiopathic PAH) and approach the severity of haemodynamic deterioration associated with disease progression.

Comprehensive echocardiographic evaluation (figures 2-8)

A) Basic standard ECHO variables

The first part of the standard ECHO examination is to scrutinize the presence of any structural heart disease. When the exam is normal, the 2D and colour Doppler evaluation reveals normal LV contractility and function and the absence of any valvular heart disease. The following analyses are recommended as a first step evaluation (table 2).

Right and left ventricle dimensions

RV end-diastolic dimension is best measured from the apical 4-chamber view, approximately 1 cm apically of the tricuspid annulus (QRS onset). Care should be taken to obtain an image demonstrating the maximum area of the RV without foreshortening. In normal conditions, the RV appears smaller than the LV¹⁸. The normal RV/LV ratio is 0.6-0.8, increasing to 0.8-1.0 (mild RV dilatation), 1.1-1.4 (moderate RV dilatation) and ≥ 1.5 (severe RV dilatation). Any value > 1.0 is strongly suggestive for RV dilatation and often corresponds to RV dysfunction¹⁹.

Pericardial effusion

Pericardial effusion is a potential cause of dyspnoea. However, when present in the context of PAH, it is one of the strongest prognostic factors. The presence of a pericardial effusion at diagnosis is associated with a less favourable outcome^{20,21}. It can be present in patients with systemic sclerosis, despite a preserved RV function, as a marker of inflammation.

Tricuspid regurgitation jet velocity (TRV)

Although some of the limitations pertaining to the inability to detect TR jet exist, Doppler ECHO is the most commonly used modality to estimate PASP. It is recommended that Doppler sweep speeds of > 50 mm/s be used for all tracings. If the signal is weak, it may be enhanced with agitated saline. A TRV < 2.5 m/s is considered to be normal, 2.5-2.8 m/s as borderline, and > 2.8 m/s as strongly indicative of manifest PH. The interpretation of TRV-derived PASP carries potential limitations that should be taken into consideration. The resting physiologic range of TRV-derived PASP depends on age, sex, body mass index and level of physical activity. As a result, TRV may exceed 2.8 m/s in the elderly, obese or in highly

trained athletes. The presence of severe tricuspid regurgitation may make the assessment of pulmonary artery systolic pressure (PASP) using TRV less reliable^{2,3,22}.

Inferior vena cava (IVC) diameter and its respiratory changes

IVC size is significantly influenced by patient position, being largest in the right lateral position, intermediate in the supine position, and smallest in the left lateral position. The diameter of the IVC decreases in response to inspiration with minimal size observed at end inspiration when the negative intrathoracic pressure leads to an increase in RV filling from the systemic veins. The diameter of the IVC, measured with the patient in the left decubitus position at 1.0 to 2.0 cm from the junction with the RA, using the long-axis view, and the percent decrease in its diameter during inspiration (collapsibility index) correlate with right atrium (RA) pressure. Estimation of RA pressure is deduced from the decrease of IVC diameter: inspiratory collapse of $> 50\%$ indicates a mildly elevated RA pressure (6-10 mm Hg), while a collapse of $< 50\%$ indicates high RA pressure (10-15 mm Hg). Dilated IVC (> 2.1 cm) without any collapse suggests a markedly increased RA pressure greater than 15 mm Hg^{19,22}.

Pulmonary acceleration time (AcT)

AcT provides an indirect estimate of mean PAP and represents a useful marker of RV function. AcT varies inversely with increasing pulmonary vascular resistance^{22,23}. AcT is defined as the interval from the onset to maximal velocity of forward flow in the right ventricular outflow tract. Because its measurement is independent of an anatomic defect or valvular regurgitation, this variable can be measured in the vast majority of patients. However, it is influenced by heart rate and prediction of mPAP by AcT are less accurate in patients with HR > 100 -110 beats/min²³. AcT tends to be longer in patients with low cardiac index and with increased pulmonary flow due to pre-tricuspid shunts².

B) Advanced extended ECHO variables

A second set of measurements is recommended when any of the basic ECHO parameters is abnormal (table 3). In the absence of identified LHD (including valvular disorders), any abnormality in RV size, function and haemodynamics^{18,19} represents a further incentive to perform a RHC.

Mitral E/e'

Impaired early diastolic filling of the LV can be the consequence of RV enlargement, septal bowing, and increased diastolic RV-LV interaction. Increased LV

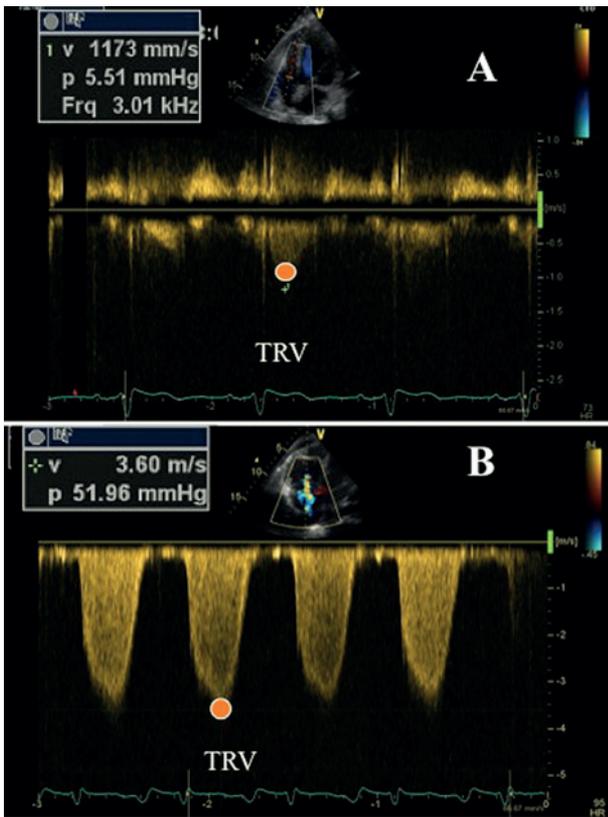


Fig. 2 Doppler echocardiographic determination of tricuspid regurgitation jet velocity (TRV): (A) in a patient without pulmonary hypertension; (B) in a patient with pulmonary hypertension.

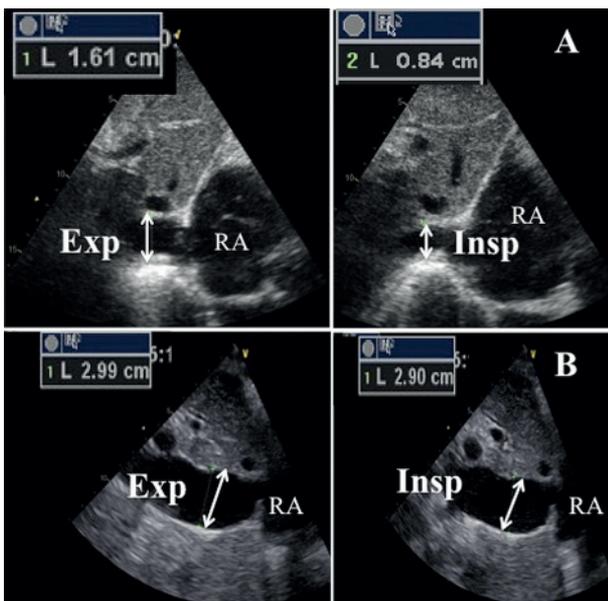


Fig. 4 Evaluation of inferior vena cava (IVC) diameter and its respiratory changes: (A) in a patient without pulmonary hypertension; (B) in a patient with pulmonary hypertension. The diameter (arrows) is measured perpendicular to the long axis of the IVC at end expiration and at end inspiration, just proximal to the junction of the hepatic veins to the ostium of the right atrium (RA).

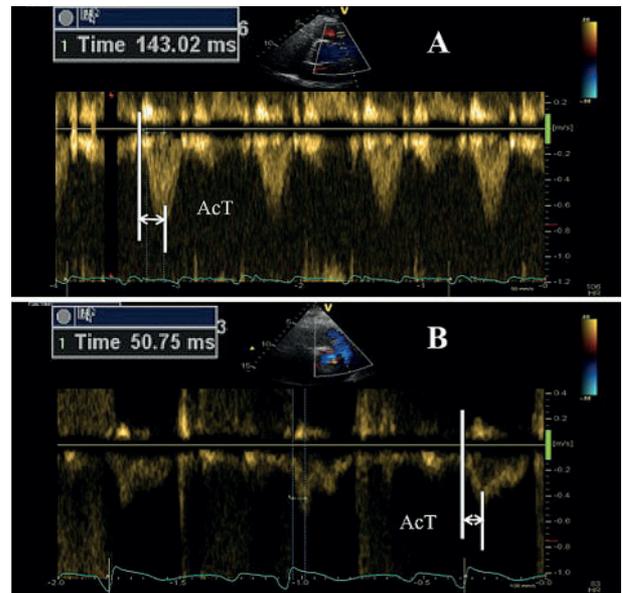


Fig. 3 Pulsed Doppler assessment of pulmonary acceleration time (AcT): (A) in a patient without pulmonary hypertension; (B) in a patient with pulmonary hypertension.

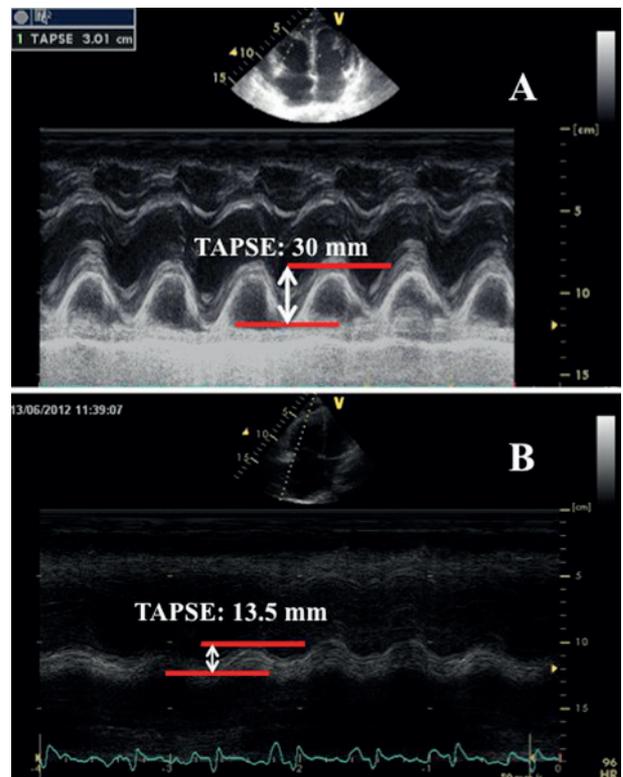


Fig. 5 Measurement of the tricuspid annular plane systolic excursion (TAPSE) by M-mode echocardiography: (A) in a patient without pulmonary hypertension and preserved right ventricular function; (B) in a patient with pulmonary hypertension and depressed right ventricular function.

filling pressure can be estimated by determining the ratio between transmitral E velocity and pulsed tissue Doppler-derived early diastolic velocity (E/e' ratio), as a predictor of PCWP. An E/e' ratio <8 is in favour of a normal LV filling pressure.

Cardiac index

In the absence of significant valvular regurgitation, the LV stroke volume can be calculated by measuring the LV outflow tract area and the amount of blood going through this area. LVOT and the respective flow velocity are more easily obtained than in the RV outflow tract. However, this method is subject to variability mainly due to the estimation of the diameter of the LVOT, which because squared in the final formula amplifies the error made.

End-diastolic pulmonary regurgitation velocity (PRV)

Doppler ECHO estimation of PA diastolic pressure can be obtained by measuring the end-diastolic velocity of pulmonary regurgitation reflecting the end-diastolic pressure gradient between the pulmonary artery and the RV. At end diastole, RV pressure should be equal to RA pressure. PRV reflects small pressure differences between the PA and the RV, usually with a pressure gradient less than 5 mm Hg (PRV < 1.2 m/s). Increases in this pressure gradient may correlate with systolic and diastolic dysfunction. Of note, the morphology of the PR jet is also affected by the degree of PAP. In the absence of PAH, the RA contraction usually does not make a significant change in PA-RV pressure gradients; hence, there is no dip in the PR jet signal. Diastolic PAP is calculated as follows: $4 \times (\text{end-diastolic PRV})^2 + \text{RA pressure}$. Once systolic and diastolic pressures are known, the mean pressure may be estimated by using the following formula: $\text{mean PAP} = 1/3(\text{SPAP}) + 2/3(\text{PADP})$. Mean PAP can also be derived from the measurement of the proto-diastolic PRV. A proto-diastolic PRV < 2 m/s (< 16 mmHg) reflects a small PA-RV pressure gradient.

Septal motion "eyeballing"

The visual estimation of RV function is not an ideal approach due to limitations of RV imaging combined with the subjectivity of the echocardiographer. Clinical decisions should not be made based on this method. LV eccentricity index is the method of choice to assess the RV/LV interactions.

Tricuspid annular plane systolic excursion (TAPSE)

Different muscle fibre orientation of the RV compared to the LV is the reason for the proportionally

greater RV longitudinal shortening of the RV free wall. TAPSE provides a good estimate of the longitudinal systolic displacement of the RV base toward the RV apex, and excellent correlation has been reported with radionuclide angiography derived RV ejection fraction. TAPSE is obtained by placing an M-mode line at the level of the lateral tricuspid annulus on the apical 4-chamber view. In healthy subjects, a TAPSE of 2.0 cm is regarded as the lowest possible normal value. RV dysfunction could be classified according the observed TAPSE values as mild (1.8-2.0 cm), moderate (1.6-1.8 cm), or severe (< 1.5 cm)^{3,24}. However, TAPSE suffers from several limitations, including angle and load dependency. In addition, TAPSE is not a real surrogate of the entire RV function as it focuses only on a small part of the RV myocardium. Finally, TAPSE is influenced by the overall cardiac motion (cardiac translation and rotation) and LV function abnormalities; hence, it might over- or underestimate RV function^{23,25}.

Peak systolic velocity of tricuspid annulus (s')

Tissue Doppler imaging provides information on segmental myocardial motion and displacement during the cardiac cycle and can be used to assess the velocity of RV contraction in the longitudinal axis (s'); this variable correlates with TAPSE. A peak s' < 12 cm/s identifies RV dysfunction with a sensitivity and specificity of around 90 and 85%, respectively¹. To perform this measure, the pulsed Doppler sample volume is placed at the level of the lateral tricuspid annulus on the apical 4-chamber view. Because it is based on the Doppler technique, care must be taken to ensure optimal image orientation (angle dependency) to avoid the underestimation of velocities. Limitations are similar to the assessment of the TAPSE.

Myocardial performance index

The myocardial performance index (MPI, or Tei index) integrates systolic and diastolic function parameters in a single measure. It uses the formula $\text{IVRT} + \text{IVCT} / \text{RVET}$, where IVRT is the RV isovolumic relaxation time, IVCT is the isovolumic contraction time, and RVET is the RV ejection time^{3,19,24}. In PH, the MPI has been employed in a few clinical studies as a non-invasive measure of both systolic and diastolic RV function. The usefulness of the MPI in clinical practice is by large limited by the following: it is highly sensitive to the volume (i.e. preload dependent), less reliable in the presence of tachycardia and seldom used because of its complexity³. Therefore, the expert consensus group attributed to the MPI a lower priority to assess RV function.

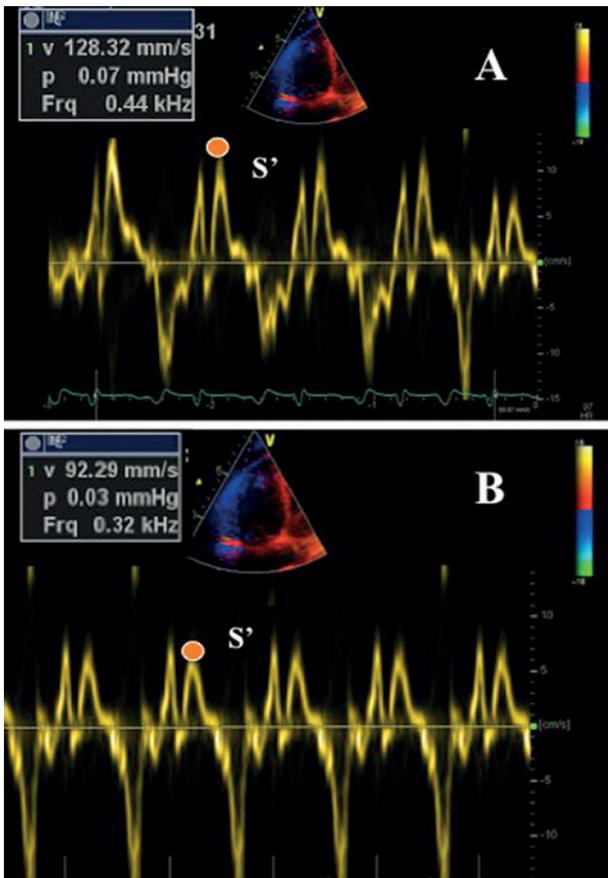


Fig. 6 Pulsed tissue Doppler imaging of the basal part of the right ventricular free wall allowing the measurement of the peak systolic velocity (*s'*): (A) in a patient without pulmonary hypertension and preserved right ventricular function; (B) in a patient with pulmonary hypertension and depressed right ventricular function.

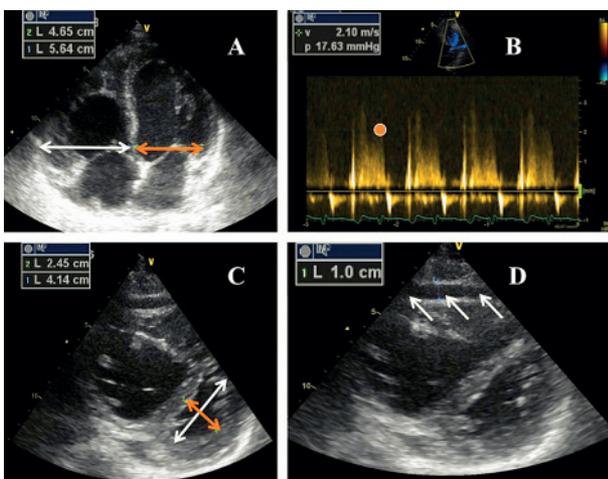


Fig. 8 ECHO imaging in a patient with pulmonary hypertension and right ventricular dilatation. (A) Right and left ventricular dimensions; (B) Doppler echocardiographic determination of end-diastolic pulmonary regurgitation velocity (PRV); (C) left ventricular eccentricity index; (D) pericardial effusion.

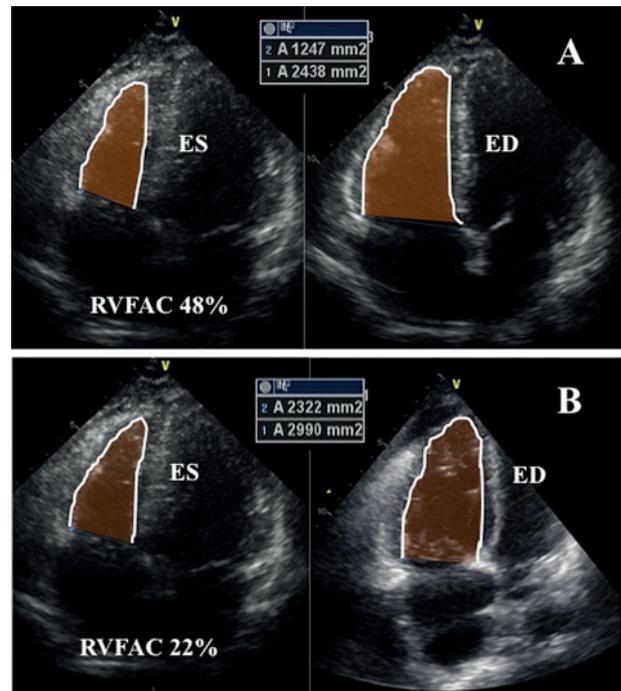


Fig. 7 Assessment of the right ventricular fractional area change (RVFAC): (A) in a patient without pulmonary hypertension and preserved right ventricular function; (B) in a patient with pulmonary hypertension and depressed right ventricular function. Percentage RVFAC = 100 × end-diastolic area (ED) – end-systolic area (ES)/end-diastolic area).

Right ventricular fractional area change (RVFAC)

This variable is derived from planimetered areas of the RV at end diastole and end systole from the apical 4-chamber view. Care must be taken to trace the free wall beneath the trabeculations. RVFAC correlates with RV ejection fraction; normal RVFAC is > 32%.

Left ventricular eccentricity index

As pressure overload and hypertrophy increase, the RV tends to dominate chamber interaction and the inter-ventricular septum flattens or bulges into the LV with impairment of LV function. This relationship between the LV and RV can be quantitated on the basis of the ratio between the LV antero-posterior dimension and the septo-lateral dimension on the LV short-axis view. Measurements are made at both end diastole and end systole. In normal subjects, the ratio between the two is equal to 1.0 at both end diastole and end systole. In patients with RV volume overload, the end-systolic ratio is approximately 1.0 while that at end diastole it is increased. In those with RV pressure overload, the eccentricity index is significantly greater than 1.0 at both end systole and end diastole.

CONCLUSION

PH is a cause of unexplained dyspnoea that can be suspected by a simple and thorough non-invasive approach. Echocardiography plays a critical role in the current diagnostic algorithm. The detection of elevated PAP and estimates of the consequences of an increased afterload on the right heart can be identified during a multiple step process by echocardiography. This also comprises the demonstration of cardiac causes of PH and the identification of the potential cause of PAH such as systemic to pulmonary shunts. Echocardiography should also be used as a screening tool in populations at higher risk of PAH, such as systemic sclerosis, and

should be prompted in patients presenting with dyspnoea in patients with underlying conditions potentially leading to PAH. However, ECHO is not a stand-alone test; it should be placed in a clinical context, together with other non-invasive tests in order to ultimately identify which patient should benefit from a confirmatory right heart catheterization. Finally, ECHO has an important role to play in risk stratification of PAH and, in the future, may be a tool to better assess the complex interplay between the RV and the pulmonary circulation.

CONFLICT OF INTERESTS: none declared.

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