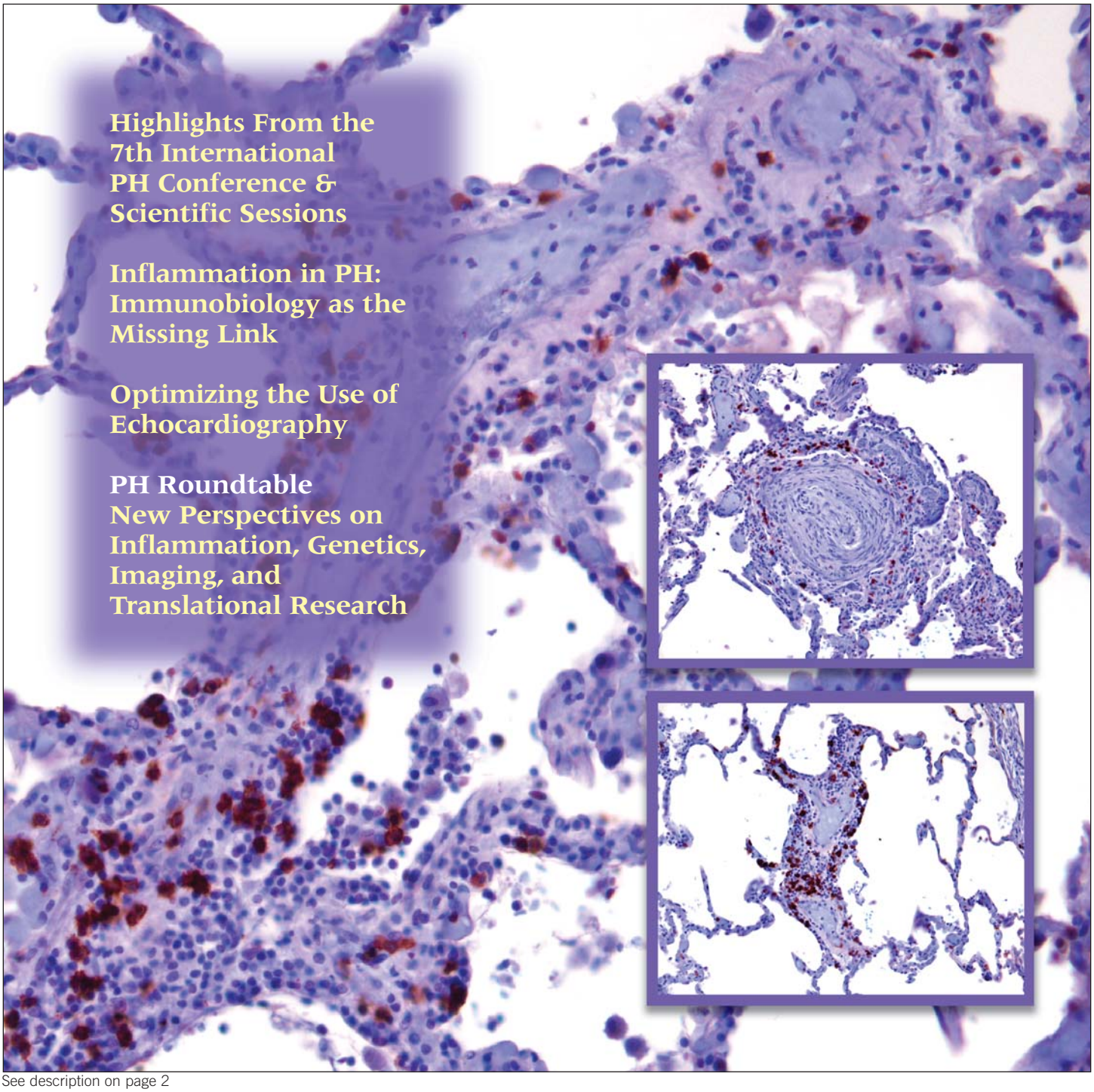
Advances in
Pulmonary Hypertension
Official Journal of the Pulmonary Hypertension Association

Autumn 2006
Vol 5, No 3



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A Stepwise and Practical Approach to Optimizing Echocardiography in Pulmonary Hypertension


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Echocardiography is noninvasive and has high sensitivity and specificity for diagnosing pulmonary hypertension (PH). It allows the assessment of right and left valve integrity and hemodynamic parameters. In patients with suspected or known PH, echocardiography can delineate left ventricular (LV) systolic and diastolic dysfunction, and detect congenital heart defects or valvular heart disease. Whereas the echocardiographic assessment of LV function is well established, internationally standardized methods to quantitate right ventricular (RV) function are less well known and reproducible. This is due to the complex, crescentic nature of RV geometry.[1] This article summarizes a practical approach using echocardiography for evaluating patients with PH.

RV Pump Function

Manifest PH leads to an enlargement of the RV and atrial areas and to eventual RV dysfunction. The evaluation of the RV size and function in patients with PH may be more prognostically important than the simple estimation of the tricuspid regurgitation velocity.[2] As a general rule the right ventricle, as seen from the apical 4-chamber view, should normally not exceed LV dimensions (**Figure 1**). If the right heart and/or right atrium are enlarged, further evaluation is usually warranted, particularly in patients with resumed PH. However, the crescent-shaped form of the right ventricle, particularly as it wraps around the left ventricle makes it difficult for direct calculation of RV dimensions and ejection fraction.[1] Whereas the action of the LV with its circular myocardial fibers is similar to a syringe, the functional geometry of the RV makes its action more like a bellow, with little excursions of the RV wall moving a large volume. In the longitudinal view, the right ventricle is similar to a triangle, while in the cross-section it is tall and thin.[3] It is not possible to completely visualize the right ventricle in one single, 2-dimensional echocardiographic view. Thus, accurate

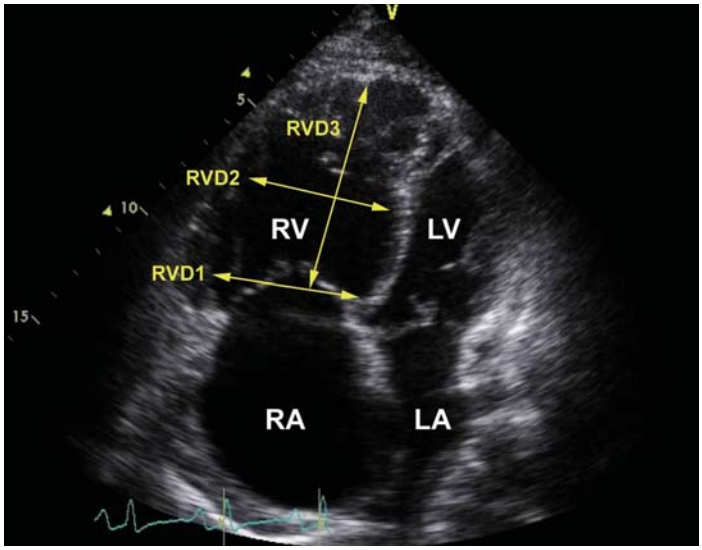

Figure 1. Right ventricular diameters in the apical 4-chamber view. Right ventricular size can be quantified by obtaining the basal (RVD1), midcavity (RVD2), and longitudinal (RVD3) diameters in the apical 4-chamber view at end diastole.[1] Normal diameters for the right ventricle are: RVD1: 2.0-2.8 cm, RVD2: 2.7-3.3 cm and RVD3: 7.1-7.9 cm.[1] In this patient with pulmonary hypertension, the right ventricle is hypertrophied and the pump function is severely impaired with marked enlargement of the right ventricle and right atrium and impression of the left ventricle and left atrium.

assessment of the right heart requires multiple echocardiographic views, including parasternal long- and short-axis, RV inflow, apical 4-chamber, and subcostal views.[1] Assessment of ventricular ejection fraction with the modified Simpson rule is not accurate in defining RV systolic function. Various approaches, including real time 3-dimensional transthoracic[4] or transesophageal 3-dimensional echocardiography[5] are proposed to assess RV volumes and function more accu-

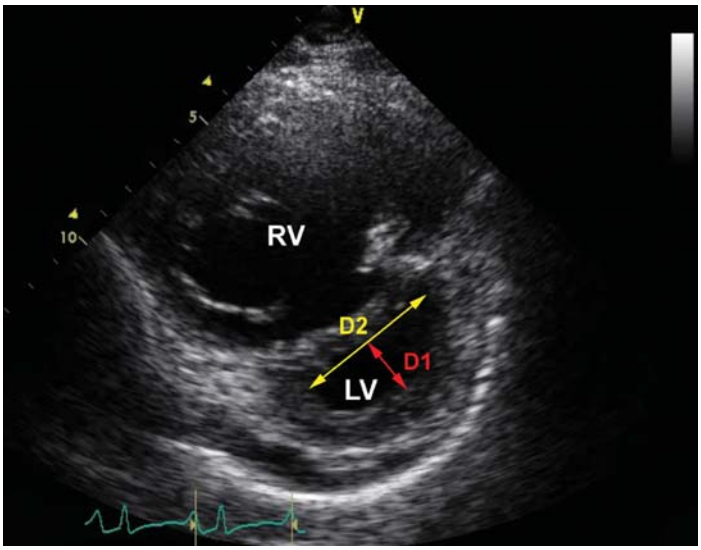

Figure 2. Left ventricular eccentricity index, as assessed from the parasternal short axis, at the level of the papillary muscles. As pressure overload and hypertrophy increase, the right ventricle (RV) tends to dominate chamber interaction and the interventricular septum flattens or bulges into the left ventricle (LV) with impairment of left ventricular function. Left ventricular eccentricity index (LV-EI) can be calculated by the formula LV-EI = D2/D1, where D2 is the diameter parallel and D1 is perpendicular to the interventricular septum; abnormal values are > 1.2, normal values LV-EI sys = 1.00 ± 0.06, LV-EI dia = 1.01 ± 0.04

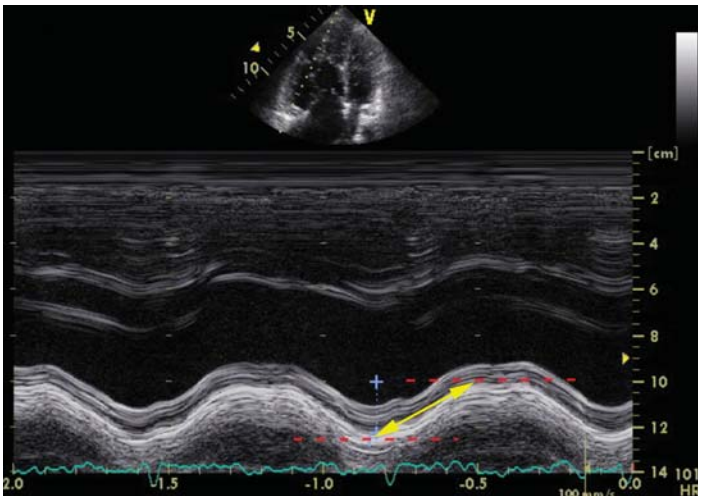

Figure 3. Measurement of tricuspid annular excursion by M-mode in the apical 4-chamber view. M-mode will be positioned in the lateral tricuspid valve annulus. The systolic excursion of the annulus toward the apex will be measured (+-+). In systole the tricuspid annulus will normally move toward the apex 1.5-2.0 cm.

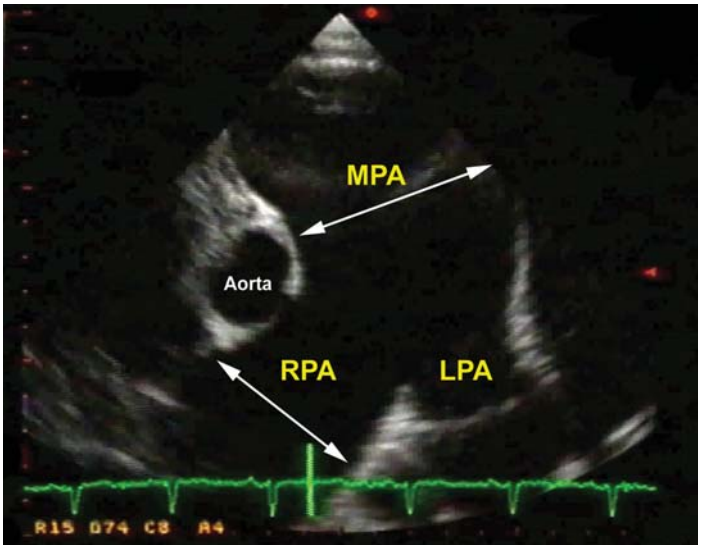

Figure 3. Right ventricular outflow track and main pulmonary artery dimensions from the parasternal short axis view. The diameter of the main (MPA), the right (RPA), or the left (LPA) pulmonary artery should not exceed 2.5 cm. In this view, an aneurysm of the central pulmonary arteries could be detected as well.

rately but are less applicable in routine examinations.

The first step in the routine evaluation of the right ventricle is the visual qualitative assessment of global systolic function in different views. With this type of examination, the RV systolic function is classified as normal, slightly, moderate, or severely impaired. When the left ventricle is deformed and/or there is a pericardial effusion, frequently more than RV function is severely impaired (**Figure 1**). The degree of LV distortion caused by the enlarged right ventricle can be further quantified using the LV eccentricity index (**Figure 2**).[6] In the short axis, the left ventricle is usually round with equal across and longitudinal diameters. In case of RV enlargement with deformity of the left ventricle, the across diameter is shorter than the longitudinal one, resulting in an eccentricity index above 1.2, which is abnormal.

Further, routine views may be useful in characterizing the patient with PH. In the short-axis view, the RV outflow tract and the central pulmonary artery can be evaluated. The main pulmonary arteries and their major branches are often dilated in patients with severe PH (**Figure 3**). Early recognition of an aneurysm of the central pulmonary artery is important, since dissection or even rupture can occur suddenly with excessive changes in intrathoracic pressure.[7] The tricuspid annular plane systolic excursion estimates RV systolic function by measuring the level of systolic excursion of the lateral tricuspid valve annulus toward the apex. In systole, the tricuspid annulus will normally move toward the apex approximately 1.5 to 2.0 cm. Tricuspid annular excursion of less than 1.5 cm is noted with severe RV dysfunction and is associated with a poor prognosis.[8]

Additional assessments of RV systolic function include tissue Doppler imaging (TDI) of tricuspid annular velocity or RV index of myocardial performance (Tei). The velocity of the lateral tricuspid annulus can be obtained by TDI. Velocities less than 10 cm per second reflect an abnormal RV function.[9] For calculation of the Tei index, RV ejection time can be obtained within pulsed wave (PW)-Doppler in the RV outflow tract. The Tei index can be calculated with the formula: isovolumic contraction time + isovolumic relaxation time/ejection time.[10]

When myocardial function deteriorates, ejection time is shortened and the pre-ejection and isovolumic relaxation periods are lengthened. This index is unaffected by heart rate, loading conditions, or the presence and severity of tricuspid regurgitation.[10] A Tei index greater than 0.40 can be associated with an abnormal global RV function with a sensitivity and specificity of 100% and 35%, respectively.[9]

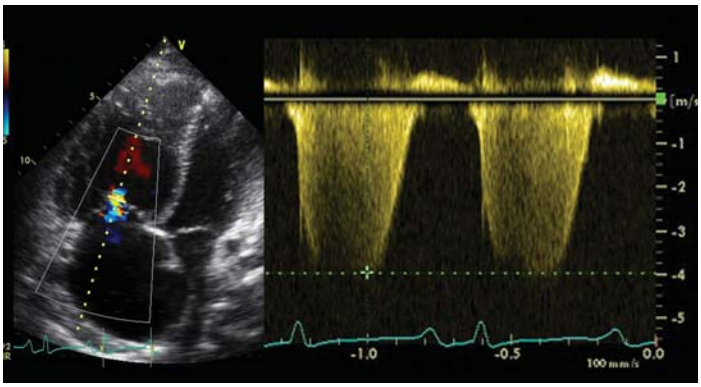

Figure 4. CW-Doppler assessment of the tricuspid regurgitation velocity. The CW-Doppler is guided by the color Doppler to obtain the tricuspid regurgitation jet within the correct angle. The regurgitation velocity profiles (on right) should be obtained by color Doppler spectral imaging and with a sweep velocity of at least 100-200 mm/s in order to measure the maximal velocity at the true edge of the profiles. The systolic pulmonary artery pressure (PASP) will be calculated by a modified Bernoulli equation, where PASP = (Vmax^2 x 4) + estimated right atrial pressure.

In addition, strain is a new echocardiographic parameter that allows the assessment of regional RV function; however, its clinical value remains to be proven.[11]

Pulmonary Artery Pressures at Rest and During Exercise

Continuous wave (CW)-Doppler guided by color Doppler is the best method to obtain tricuspid regurgitant jet velocity (**Figure 4**).[12] In the absence of pulmonic valve stenosis or outflow tract obstruction, systolic pulmonary artery pressure (PASP) can be estimated using the Bernoulli equation: [tricuspid regurgitant jet velocity (V)^2 x 4] + estimated right atrial pressure (RAP).[12] If the inferior vena cava is less than 20 mm in diameter and collapses with respiration, 5 mmHg should be added for RAP; 10 mmHg if it is greater than 20 mm but with inspiratory collapse, and 15 mmHg when the diameter is above 20 mm without variation with inspiration.[13]

Tricuspid regurgitation jets are detectable in 39% to 86% of patients[14,15] and the derived PASPs correlate well with invasively obtained values at rest[12] and during exercise.[16,23] However, in a significant number of patients, PASP values are over- or underestimated.[17] Invasive[18,19] and noninvasive[16,20] studies note that PASP at sea level in healthy subjects younger than 50 years does not exceed 40 mmHg at rest. In athletes, PASP values can exceed 40 mmHg during higher workloads.[21]

Pulmonary diastolic pressure can also be estimated by Doppler echocardiography and correlates well with invasive measurements.[22] An exaggerated PASP response to exercise can identify persons susceptible to high altitude PH[20] and allow screening of at-risk family members of patients with idiopathic pulmonary arterial hypertension (PAH).[23] Thus, stress echocardiography during supine bicycle exercise may be a useful screening method to identify persons at risk for developing PH. At minimum, echocardiography at rest should be performed in persons with a clinical suspicion of PAH.[24]

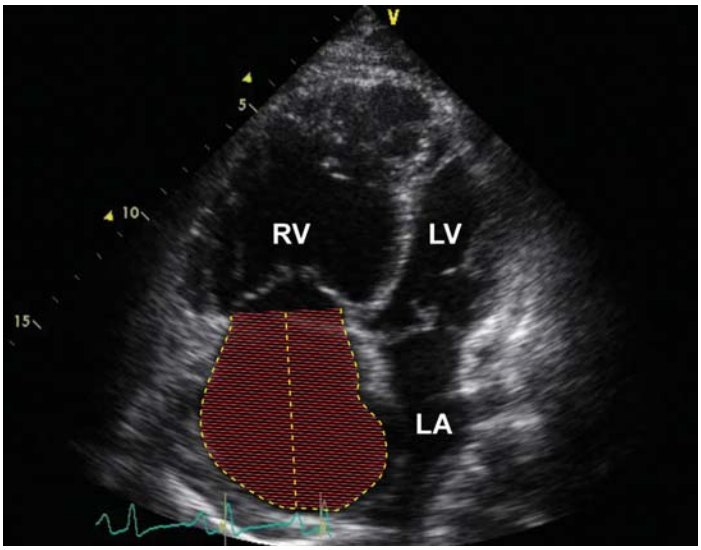

Figure 5. Assessment of right atrial size. Right atrial volume can be assessed from the apical 4-chamber view that is more accurate for measuring the right atrial size than linear dimensions. The single area-length method as shown here (hatched area) can be applied to determine right atrial volume. A right atrial size above 27 cm^2 is pathologic and associated with a poor prognosis.[27] Normal values of indexed right atrial volumes are similar to left atrium (21 m1/m2).[1]

Summary

Further echocardiographic assessments in patients with PH should include the analysis of LV systolic and diastolic function, as well as valvular evaluation to exclude congenital heart disease. Left ventricle diastolic function should be examined by both PW-Doppler and tissue Doppler imaging. An impairment of the early phase of LV diastole is the most common type of dysfunction seen in patients with chronic PH, with reduced early to late diastolic filling ratio and diminished LV end-diastolic volume, which contributes to a reduced LV stroke volume.[25,26]

Echocardiographic predictors of poor prognosis in patients with PH include the occurrence of a pericardial effusion,[27,28] an enlarged right atrium (**Figure 5**),[27] a Tei index of at least 0.83,[29] an RV myocardial performance index greater than 1.4,[30] a dilated inferior vena cava,[31] and an eccentricity index greater than 1.2.[32] Interestingly, pulmonary artery systolic pressures are not independent predictors of prognosis.

Echocardiography is an important tool in terms of both diagnosis and prognosis in PH. Techniques to quantify the RV function should be further evaluated and may include the strain and TDI for determination of regional RV pump function, and stress echocardiography for identification of subjects at risk for PH and as a follow-up tool.

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