1. Introduction

The gold standard for diagnosis and confirmation of PH is right heart catheterization. But right heart catheterization is an invasive procedure, and it associated with morbidity (1.1%) and mortality (0.55%)(Hoeper et al. 2006). Although right heart catheterization is the method to measure pulmonary arterial pressures, it is not suitable in daily practice to screen the possible candidates of PH. Echocardiography is an widely available non-invasive tool, and it can offer the information on the structures of hearts and the estimated pressure profiles of cardiovascular system. Therefore, echocardiography plays an important role in diagnosis and monitoring treatment effects.

The classification of pulmonary hypertension (PH) includes 5 groups: 1) pulmonary arterial hypertension (PAH); 2) pulmonary hypertension owing to left heart disease; 3) pulmonary hypertension due to lung disease; 4) chronic thromboembolic pulmonary hypertension; 5) miscellaneous.(Simonneau et al. 2009) In the clinical situation, while suspecting a patient with PH, it plays an important role in screening the patients with suspected PH, ruling in or out PH due to left heart disease, and evaluating the presence of congenital heart disease. If the patient is diagnosed as PAH, echocardiography can afford the semi-quantitative methods to evaluate the effects of treatment.

2. Echocardiography in diagnosis of PH

2.1 Structures of heart

Conventional echocardiography can provide the two-dimensional images to evaluate the structural abnormalities of heart. Group 2 PH account for the most cases of PH (Oudiz 2007), therefore examination of the function and structure of left heart is the first thing to do in diagnosis of PH. Congenital heart disease is another common cause of PH, and it usually can be corrected by surgery. Therefore, it is important in evaluating the gross anatomy of left heart, the systolic function of left ventricle, mitral valve and aortic valve disorders, the diastolic dysfunction of left heart, and the congenital heart defects. The detail evaluation of structural evaluation of heart is beyond the scope of this book. In the beginner of PH, who is not the cardiologist, cardiovascular consultant is necessary. But diastolic dysfunction is usually overlooked in the clinical practice. The possible clue of isolated diastolic dysfunction of left heart is dilated left atrium. If this sign is present and the pulmonary capillary wedge
pressure is less than 15 mmHg during right heart catheterization, saline replacement during right heart catheterization may uncover the occult diastolic dysfunction. (Hoeper et al. 2009).

It not uncommon that congenital heart disease cannot be diagnosed by transthoracic echocardiography due to limited windows, transesophageal echocardiography is a necessary tool in this situation, especially in atrial septal defect and pulmonary vein stenosis.

Because of chronic right ventricular (RV) pressure overload, most patients present with enlarged right-side chambers, RV hypertrophy, and reduced global RV systolic function. (Bossone et al. 1999) The hallmark of RV pressure overload is systolic flattening of the interventricular septum with D-shape of left ventricle (Figure 1A), with reduced diastolic and systolic left ventricular volumes, but preserved global left ventricular systolic function. The eccentric index of left ventricle, which measured at papillary muscle level of mitral valve in paraternal short-axis view, is less than 1. (Figure 1A) Pericardial effusions have also been described in PAH patients, and this sign represents higher right atrial pressures, higher pulmonary arterial pressures and poor prognosis. (Park et al. 1989) (Figure 1B)

Fig. 1. Echocardiographic parasternal short-axis view. The interventricular septum is flattened due to pressure overload of the right ventricle.
2.2 Hemodynamics

As mentioned before, right heart catheterization is the gold standard in diagnosis of PH, but non-invasive Doppler echocardiography can also afford estimates of hemodynamic parameters, such as pulmonary arterial pressure, cardiac output, pulmonary vascular resistance and pulmonary capillary wedge pressures. Therefore, it is a good screening tool in patients with PH.

2.2.1 Pulmonary arterial pressures

The hemodynamic criterion of pulmonary hypertension is mean pulmonary arterial pressure greater than 25 mmHg by right heart catheterization. Pulmonary arterial pressures can also be measured semi-quantitatively by echocardiography. For calculating pulmonary arterial systolic pressures, the first step is to estimate right atrial pressure (RAP), which is evaluate by examining the diameter of inferior vena cava. IVC diameter < 2.1 cm that collapses >50% with a sniff suggests normal RA pressure of 3 mm Hg (range, 0-5 mm Hg), whereas IVC diameter > 2.1 cm that collapses < 50% with a sniff suggests high RA pressure of 15 mm Hg (range, 10-20 mm Hg). In scenarios in which IVC diameter and collapse do not fit this paradigm, an intermediate value of 8 mm Hg (range, 5-10 mm Hg) may be used. (Table 1) (Rudski et al.)
2010) The second step is search the presence of tricuspid regurgitation (TR). When TR is present, measurement of the peak TR velocity ($V_{TR}$) should be performed. Because velocity measurements are angle dependent, it is recommended to obtain TR signals from several windows and to use the signal with the highest velocity. The third step is calculating pulmonary arterial systolic pressure (PASP) by the following equation:

$$PASP = RAP + V_{TR}^2$$ (Figure 3)

Fig. 3. A 51-year-old female, with a history of systemic lupus erythematosus and pulmonary arterial hypertension, had an estimated tricuspid regurgitation pressure gradient of 54mmHg. After adding estimated right atrial pressure, pulmonary arterial systolic pressure would be obtained.

But the accuracy of the estimated PASP depends on recording a clear envelope of the TR velocity by continuous-wave Doppler tracing. If the signal of TR is not clear or incomplete, underestimation of peak velocity of TR will occurs.

Estimation of pulmonary arterial diastolic pressure (PADP) is similar to that of PASP, and the velocity used in estimated PADP is the pulmonary regurgitation (PR) velocity at end-diastole. The formula to calculate PADP is as following:
PADP = RAP + $V_{\text{end-diastole}} P_{R}^{2}$ (Figure 4)

Fig. 4. A 51-year-old female, with a history of systemic lupus erythematosus and pulmonary arterial hypertension, had an estimated pulmonary regurgitation pressure gradient of 32mmHg. After adding estimated right atrial pressure, pulmonary arterial diastolic pressure would be obtained.

After obtaining PASP and PADP, the mean PA pressure (MPAP) can be obtained as following:

$$\text{MPAP} = \text{PADP} + \frac{1}{3} (\text{PASP} - \text{PADP}) \quad (\text{Figure 3})$$

<table>
<thead>
<tr>
<th>RAP (mmHg)</th>
<th>Normal (0-5)</th>
<th>Intermediate (5-10)</th>
<th>High (15)</th>
</tr>
</thead>
<tbody>
<tr>
<td>IVC diameter (mm)</td>
<td>21</td>
<td>&lt;21</td>
<td>&gt;21</td>
</tr>
<tr>
<td>Collapse with sniff</td>
<td>&gt;50%</td>
<td>&lt;50%</td>
<td>&gt;50%</td>
</tr>
<tr>
<td>Secondary indices of elevated RAP</td>
<td>Restrictive filling</td>
<td>Tricuspid E/E’ &gt; 6</td>
<td>Diastolic flow predominance in hepatic vein (systolic filling fraction &lt; 55%)</td>
</tr>
</tbody>
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Table 1. Estimation of right atrial pressure (RAP) on the basis of IVC diameter and collapse (Rudski et al. 2010)
2.2.2 Cardiac output

Cardiac output is related to the symptoms and prognosis of PH. Cardiac output can also be calculated by echocardiography. Before calculating cardiac output, the presumption is absence of intra-cardiac or great vessel shunt, and the cardiac output can be measured from left ventricular outflow tract (LVOT) or right ventricular outflow tract. In daily practice, measurements of diameters and velocity time integral of left ventricular outflow tract is a routine practice in our echocardiographic laboratory. Therefore, application of pulse-wave Doppler over LVOT, a clear envelope of LVOT flow (VTI\textsubscript{LVOT}) can be acquired. The diameter of LVOT (D\textsubscript{LVOT}) can be measured at parasternal long-axis view. The cardiac output can be calculated as the following equation:

\[
\text{Cardiac output} = 0.785 \times D^{2} \times \text{VTI}_{\text{LVOT}} \times \text{heart rate} / 1000
\]

*1000 is for conversion of units

Fig. 5. A 34-year-old male with thalassemia and chronic hemolytic anemia related pulmonary hypertension. (VTI\textsubscript{LVOT}: 16.3 cm/s, D\textsubscript{LVOT}: 2.3 cm, Cardiac output= 0.785*2.3\(^2\)*16.3*66/1000 = 4.4L/min)

3. Echocardiography in evaluation of right heart function

Right ventricular function is a prognostic factor of PH. However, because of complex geometry of right ventricle, it is difficulty to evaluate right ventricular function. In current era, more studies demonstrated the clinical utility and value of right ventricular myocardial performance index, tricuspid annular plane systolic excursion (TAPSE), right ventricular fractional area changes and Sm of the tricuspid annulus.

3.1 Right ventricular myocardial performance index (RVMPI)

RVMPI is an index of global RV function, and it is calculated by summation of isovolumic contraction time (IVCT) and isovolumic relaxation time (IVRT) and divided by ejection time (ET). (Figure 6)

\[
\text{RVMPI} = \frac{\text{IVCT} + \text{IVRT}}{\text{ET}}
\]
Fig. 6. Myocardial TDI includes all 3 phases. The MPI was calculated as \((a-b)/b\).\( a \): the time interval from the onset of isovolumic contraction to the end of isovolumic relaxation; Am: late diastolic wave; Em: early diastolic wave; EKG: electrocardiogram; IVCT: isovolumic contraction time; IVRT: isovolumic relaxation time; Sm: the wave for the systolic phase.

It can be obtained by flow-Doppler and tissue Doppler technique. By using flow-Doppler technique, pulse-wave Doppler tracings are obtained from tricuspid valve and right ventricular outflow tract. By using tissue-Doppler technique, IVCT, IVRT and ET are obtained from the lateral tricuspid annulus. (Figure 6) RV MPI > 0.40 by flow-Doppler technique and > 0.55 by tissue-Doppler technique indicate RV systolic dysfunction. (Rudski et al. 2010)

3.2 Tricuspid annulus plane systolic excursion (TAPSE)

Although the morphology of RV is complex, contraction of the longitudinal fibers of RV cause most of RV systolic function and measurement of RV longitudinal motion can represent RV systolic function. TAPSE is an easy and convenient way to evaluate the RV longitudinal motion. In apical four-chamber view, the cursor line is placed across the lateral tricuspid annulus, and M-mode imaging of the cursor line reveals the longitudinal motion of lateral tricuspid annulus. TAPSE is the distance, which tricuspid annulus moves from the end-diastole phase to the end-systolic phase, and it represents the RV systolic function. When TAPSE < 16mm, RV systolic dysfunction may be present. (Figure 7)
3.3 Right ventricular fractional area change (RVFAC)

RVFAC, which resembles left ventricular ejectional fraction, provides an estimate of RV systolic function. To obtain RVFAC, clear RV morphology and endocardial border should be clear at apical four-chamber views. RV end-diastolic and end-systolic areas (RVEDA & RVESA) are obtained after tracing RV area.

$$RVFAC = \frac{RVEDA - RVESA}{RVEDA} \times 100\%$$

RVFAC > 40% is normal. RVFAC <35% indicates RV systolic dysfunction. It is important to make sure that the entire right ventricle is in the view, including the apex and the lateral wall in both systole and diastole. Care must be taken to exclude trabeculations while tracing the RV area.

3.4 Peak Sm of lateral tricuspid annulus

Although tissue Doppler imaging detects regional myocardium function, pulmonary hypertension causes global RV change and lateral tricuspid annulus function can represent global RV function. Peak Sm of lateral tricuspid annulus is obtained by tissue-Doppler
technique at end-expiration period. Peak Sm> 10 cm/s is normal, and peak Sm< 10 cm/s represents depressed RV systolic function. (Figure 8)

![Echocardiography Image](image)

Fig. 8. A 45-year-old female with systemic lupus erythematosus and pulmonary arterial hypertension, had a low peak Sm (6.0 cm/s) at lateral tricuspid annulus, which indicates poor RV systolic function.

### 4. Conclusion

Echocardiography is a useful tool in diagnosis of PH, evaluating the left heart disease, and estimation of pulmonary arterial hemodynamics. After diagnosis of PH is established, TAPSE, peak Sm velocity and RVMPI are simple methods to monitor treatment effects.

### 5. References


The textbook "Pulmonary Hypertension - From Bench Research to Clinical Challenges" addresses the following topics: structure and function of the normal pulmonary vasculature; disregulated cellular pathways seen in experimental and human pulmonary hypertension; clinical aspects of pulmonary hypertension in general; presentation of several specific forms of pulmonary hypertension, and management of pulmonary hypertension in special circumstances. The textbook is unique in that it combines pulmonary and cardiac physiology and pathophysiology with clinical aspects of the disease. First two sections are reserved for the basic knowledge and the recent discoveries related to structure and cellular function of the pulmonary vasculature. The chapters also describe disregulated pathways known to be affected in pulmonary hypertension. A special section deals with the effects of hypoxia on the pulmonary vasculature and the myocardium. Other three sections introduce the methods of evaluating pulmonary hypertension to the reader. The chapters present several forms of pulmonary hypertension which are particularly challenging in clinical practice (such as pulmonary arterial hypertension associated with systemic sclerosis), and lastly, they address special considerations regarding management of pulmonary hypertension in certain clinical scenarios such as pulmonary hypertension in the critically ill.

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