Physical Examination in Pulmonary Arterial Hypertension

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The physical examination is an integral to the evaluation of any patient with suspected cardiopulmonary disease. This is particularly true of patients with pulmonary arterial hypertension (PAH) as careful attention to physical signs will not only alert the clinician to the presence of PAH, but guide assessment of disease severity and provide important clues regarding underlying pathogenesis. This paper will focus on the physical findings most characteristic of patients with PAH, with a brief discussion of normal physical findings where appropriate. Descriptions in the text link directly to the audio and video examples of the relevant physical findings on the CD-ROM Pulmonary Hypertension: An Interactive Guide to Diagnosis. A complimentary copy is available through the Pulmonary Hypertension Association at (see page 5).

Jugular Venous Pulse

Important information regarding the condition of the right heart can be obtained from observation of the jugular venous pulse.[1] Discriminating the internal jugular vein from the external jugular vein and the carotid pulsation is an important part of this evaluation. The internal jugular vein is located deep in the neck, covered by the sternocleidomastoid muscle, but its pulsations are transmitted to the skin of the neck. It is both easier and more clinically useful to evaluate the internal jugular pulse on the right side of the neck. It is a relatively straight path from the right atrium to the right internal jugular via the innominate vein, and therefore assessment of the right internal jugular gives a more accurate account of right atrial pressures. By convention, most patients are examined at a 45º angle; however, in patients with very high venous pressures a more acute (60º to 90º) angle may be required to adequately discriminate the jugular pulsations. It is helpful to make a note of the height of the jugular venous pulsations and the angle at which they were measured during each evaluation to track changes in this physical finding during subsequent visits.

The internal jugular vein is usually not visible as a discrete structure except in the presence of significant right heart pressure elevation. However, transmitted pulsations of the internal jugular are visible at the surface of the neck in many patients. The first important step in evaluating the internal jugular wave forms is discriminating them from the carotid pulsations. There are a number of clues to assist with this discrimination. The venous wave can often be dampened or suppressed by firm placement of a palpating finger below the pulsation at the base of the neck (this may not be true if severe elevation of right heart pressures exists), while arterial pulsations continue despite firm palpation. Arterial pulsations do not change location in relation to patient position while venous waves increase and decrease in height depending on the angle of incline of the patient. Lastly, the venous pulse has two peaks and two troughs, while the arterial pulse has a single upstroke.[2]

Two aspects of the jugular venous pulse should be assessed in the evaluation of the patient with PAH: 1) the height of the jugular venous distension above the sternal angle (angle of Louis), and 2) the quality of the venous wave pattern.[3]

Height of jugular venous distension (Case 7 2L). PAH results in an elevation in jugular venous pressure, indicating an increase in right atrial pressure. To measure jugular venous distention, identify the top of the oscillating jugular vein waveforms with the patient in a semirecumbent position (usually a 45º angle). The distance from the sternal angle to the top of the waveform is measured in centimeters. By convention, 5 cm is then added to this measurement, as the right atrium is approximately 5 cm below the sternal angle. Four centimeters above the sternal angle is the upper limit of normal for jugular venous distension and this corresponds to a jugular venous pressure of 9 cm H_20 (4 cm + 5 cm).

Quality of jugular venous waves. The jugular venous wave is composed of three peaks or positive deflections (*a, c,* and *v*) and two descents (*x* and y) (**Figure 1**). The morphology of these waves can be altered by a number of cardiovascular diseases, including PAH. The *a* wave results from venous distension during right atrial contraction. The *x* descent occurs during right atrial relaxation. The *c* wave is generated by the bowing of the tricuspid valve into the right atrium during ventricular systole. The *v* wave occurs due to increased right atrial pressure as venous blood fills the right atrium during ventricular contraction when the tricuspid

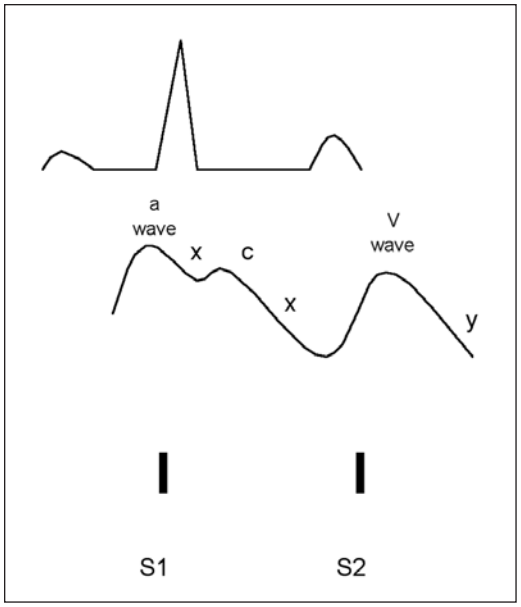

Fig 1a. Jugular venous pulse waves and descents and their relation to the ECG and heart sounds.

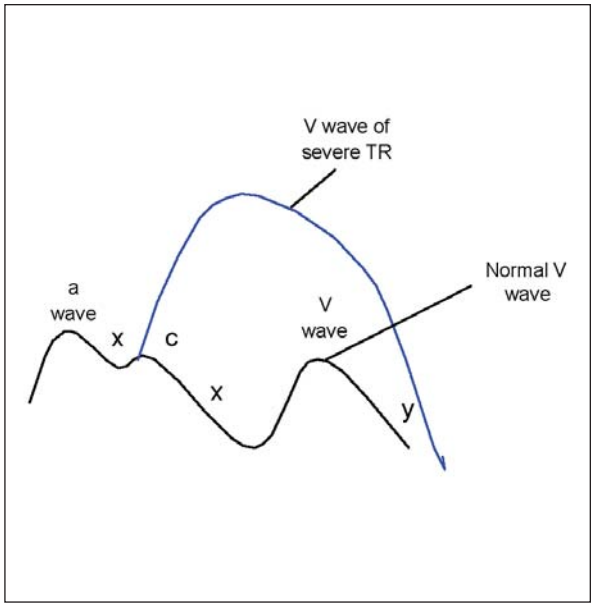

Fig 1b. Jugular venous wave forms in a normal individual and in a patient with severe tricuspid regurgitation (TR). Note the significant increase in size of the "c-v" wave.

valve is closed. The *y* wave is related to the decrease in right atrial pressure as the tricuspid valve opens.

Pulmonary hypertension can affect the *a* wave and the *v* wave as well as their associated descents. Contraction of the right atria against increased downstream resistance (such as occurs in PAH) results in an increased *a* wave (**Case 2: 3L**). Significant tricuspid regurgitation (common in PAH) increases the amplitude of the v wave (**Case 7: 2L**). Typically in severe PAH, the *v* wave is the more prominent of the two waveforms.

Cardiac Sounds

Normal Findings

S1 and S2. The first heart sound (S1) is generated by the closing of the mitral (M1) and tricuspid (T1) valve, while the second heart sound (S2) is produced by the closure of the aortic (A2) and pulmonic (P2) valves, respectively. Normally, S1 is heard as a single sound as the mitral and tricuspid valves close essentially simultaneously. The second heart sound (S2) varies within the respiratory cycle. During expiration, S2 is heard as a single sound as the aortic and pulmonic valve close together. However, during inspiration, negative intrathoracic pressure generated by the diaphragm causes increased blood flow to the right heart. This increased flow delays the closure of the pulmonic valve and a "split" second heart sound, which is an audible differentiation of A2 and P2, can be detected (**Figure 2a**).

Findings in PAH

***S2* (Case 6 or 7: 3L).** The second heart sound is frequently accentuated in patients with pulmonary hypertension. This is because the intensity of P2 is dependent on the velocity of blood coursing back toward the right ventricle after ventricular contraction and the suddenness in which that motion is arrested by the closing valve. In patients with PAH, the diastolic pressure within the pulmonary artery is high and therefore the velocity of blood moving toward the tricuspid valve is increased, resulting in an accentuated P2.

Widened splitting. This refers to a longer than expected interval between S1 and S2 and can be caused by pulmonic valve stenosis (this is not a true form of PAH but can present with right heart failure and is easy to misdiagnose by history as well as by echocardiography). A widened split S2 can be an important clue to pulmonic stenosis (**Figure 2b**).

Fixed splitting. This refers to an audible separation between A2 and P2 that persists through both inspiration and expiration (**Figure 2c**). This finding frequently indicates the presence of an atrial septal defect. The fixed splitting is due to the continuous delay of P2 closure throughout the cardiac cycle related to increased blood flow to the right ventricle. The blood flow to the right ventricle is increased in inspiration for the reasons discussed above and increased in expiration from the volume of blood shunted from the left to the right heart through the septal defect.[4]

Extra Cardiac Sounds in PAH

S3. An S3 occurs in early diastole during the ventricular rapid filling stage, following the opening of the atrioventricular valves. While an S3 heard in children or young adults is often a normal finding, in older individuals, patients with a depressed left ventricular ejection fraction, and patients with PAH an S3 is a sign of increased diastolic ventricular filling pressure and ventricular failure.[5] The sound is generated by the tensing of the chordae tendineae. A right-sided S3 (more typical of patients with PAH) is best appreciated with the bell of the stethoscope placed over the tricuspid region with the patient in the supine position and during inspiration.[3]

S4. An S4 occurs late in diastole and is caused by contraction of the atria as they force the last of the blood from the atria into a stiffened ventricle just prior to ventricular systole. The presence of an S4 indicates decreased ventricular compliance, often due to hypertrophy. An S4 is heard best with the bell of the stethoscope with the patient lying in the left lateral decubitus position.

Cardiac Murmurs

Heart murmurs are caused by turbulent blood flow. Pathologic changes in patients with PAH can result in a variety of murmurs detectable and distinguishable by careful auscultation.

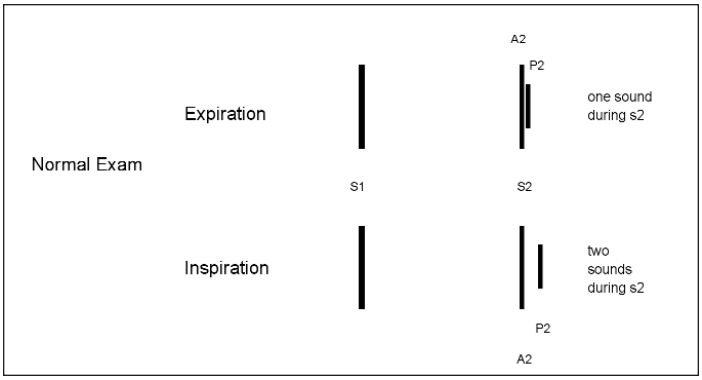

Fig 2a. Graphic representation of the normal cardiac ausculatory findings on physical examination. During inspiration P2 separates from A2 due to increased blood flow to the right heart and can be detected as a separate sound.

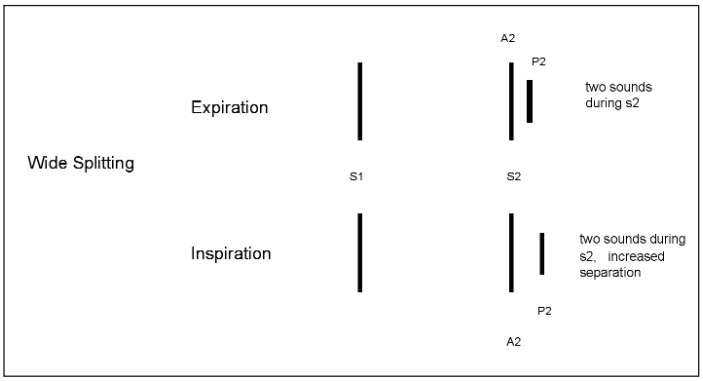

Fig 2b. Graphic representation of widened splitting of the second heart sound (S2). This can be heard in pulmonic stenosis.

Tricuspid Regurgitation (Case 7: 2L or 3L). Perhaps the most common and recognizable murmur of PAH is tricuspid regurgitation. A tricuspid regurgitation murmur is best heard along the lower left sternal border. It can radiate to the right of the sternum and is high pitched and blowing in quality. The murmur of tricuspid regurgitation is holosystolic and can be augmented by inspiration, increasing return of venous blood to the right ventricle.[6]

Ventricular Septal Defect (Case 3: 2R). The murmur of a ventricular septal defect is holosystolic. It is heard best at the 4th to 6th left intercostal space and is high pitched and may be associated with a palpable thrill. The intensity of a ventricular septal defect murmur does not change with inspiration and it does not radiate. Paradoxically, the smaller the ventricular septal defect, the greater the turbulence of blood flow moving from the high-pressure left ventricle to the low-pressure right ventricle, and thus the louder the murmur.[7,8]

Pulmonic Stenosis (Case 1: 2L). The murmur of pulmonic stenosis is a systolic ejection murmur (crescendo-decrescendo). It is best appreciated at the left upper sternal border.[9]

Pulmonic Regurgitation (Case 2: 2L). This early diastolic murmur can occasionally be detected in patients with severe pulmonary hypertension. It occurs as the pulmonary artery dilates, resulting in pulmonic valve incompetence. It is a high-pitched, blowing murmur best heard using the diaphragm of the stethoscope and with the patient sitting up, leaning forward, and in fixed expiration. This murmur is also referred to as the Graham Steell murmur.[3]

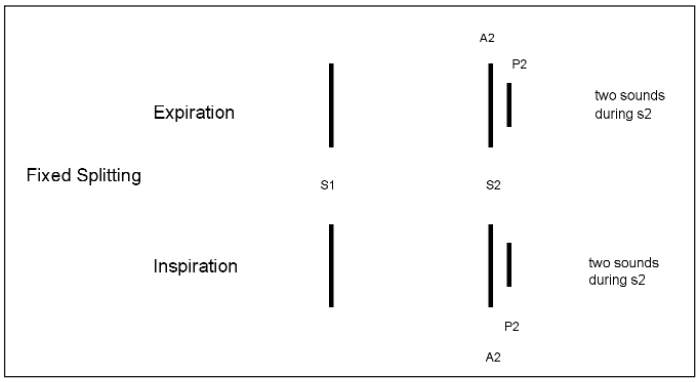

Fig 2c. Graphic representation of fixed splitting of S2 during inspiration and expiration. This is a common finding in patients with atrial septal defects.

A variety of other valvular diseases and intracardiac shunts can result in PAH; however, a discussion of all these physical findings is beyond the scope of this paper.

Other Physical Findings in PAH

Clubbing (Case 4: Insp Hands). Clubbing is a descriptive term, referring to the bulbous, uniform swelling of the soft tissue of the terminal phalanx of a digit resulting in the loss of the normal angle between the nail and the nail bed. Although clubbing is a common physical finding in many pathologic processes, the pathophysiologic mechanism of this finding remains unclear. The earliest forms of clubbing are characterized by increased glossiness of the distal skin of the finger and the root of the nail. There is then obliteration of the normal angle between the base of the nail and the skin. The soft tissue of the pulp becomes hypertrophied and the nail root floats freely. On examination one may note a spongy sensation as the nail is pressed toward the nail plate. The sponginess results from increased fibrovascular tissue between the nail and the phalanx. The skin at the base of the nail may be smooth and shiny.

Clubbing of the digits is common in congenital heart diseases that cause pulmonary hypertension (atrial septal defects, ventricular septal defects). It is an unusual finding in other forms of pulmonary hypertension.

Edema (Case 6: Insp). Edema of the lower extremities is a common finding in advanced pulmonary hypertension resulting in right heart dysfunction. Firm pressure on the pretibial region for 10 to 15 seconds may be necessary for detection of edema in less severe disease.

Hepatojugular reflux. Firm pressure over the liver (or other areas of the abdomen) can cause an increase in jugular venous distension. This is indicative of right heart failure.

Ascites. Abdominal distension with shifting dullness or a fluid wave is a sign of ascites familiar to most clinicians. The presence of ascites tends to be a late finding in patients with PAH and is indicative of severe right ventricular dysfunction and elevated right atrial pressure. Ascites is a common finding in patients with portal hypertension related to hepatic disease. As portal hypertension is recognized as being associated with pulmonary hypertension, this physical finding

may provide a clue toward the etiology of the elevated pulmonary pressures.

Raynaud Phenomenon (? Case 4: Insp Hands). The Raynaud phenomenon is an occasional finding in patients with idiopathic pulmonary arterial hypertension (IPAH) and a common finding in individuals with PAH associated with connective tissue disease, in particular, limited-stage scleroderma or CREST (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, telangiectasia).

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