Premature Pulmonic Valve Opening Following Sinus of Valsalva Aneurysm Rupture into the Right Atrium

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SUMMARY
Echocardiographic features of a patient with sinus of Valsalva aneurysm rupture into the right atrium are described. The aneurysm presented as a dense echo-producing mass in the right atrium which descended into the tricuspid orifice during diastole and withdrew back into the atrium during ventricular systole. Pulmonic valve echo motion demonstrated early diastolic pulmonic valve opening indicating an early right ventricular diastolic pressure rise exceeding simultaneous pulmonary artery pressure. Since the aorta is the only source of early diastolic pressure in excess of pulmonary artery pressure available to the right heart, this finding of early diastolic pulmonic valve opening indicated the presence of a fistula between the aorta and right heart. Other interesting echocardiographic features of this case are also presented.

Additional Indexing Words:
Ultrasound Echocardiography Bacterial endocarditis

SEVERAL RECENT REPORTS have suggested that pulmonic valve echo motion could be of diagnostic value in patients with valvular pulmonic stenosis and pulmonary hypertension. A relationship between the magnitude of pulmonic leaflet motion produced by atrial systole and the simultaneous right ventricular and pulmonary artery pressures was noted in each of these studies. A prominent opening or doming motion of the pulmonic leaflets was observed in patients with valvular pulmonic stenosis when atrial contraction elevated right ventricular pressure above simultaneous pulmonary artery pressure. Conversely, no leaflet motion was produced by atrial systole in patients with pulmonary hypertension, in whom right ventricular end-diastolic pressure failed to approach simultaneous pulmonary artery pressure.

This report describes a patient who presented with clinical features of bacterial endocarditis complicated by acute aortic insufficiency and biventricular failure. Pulmonic valve echo motion suggested a shunt from the aorta to right atrium or ventricle. A right sinus of Valsalva aneurysm with rupture into the right atrium was later demonstrated at catheterization and confirmed at surgery. A number of other echocardiographic features of this case are presented which may prove valuable in the noninvasive diagnosis of sinus of Valsalva aneurysm.

Case Report
A 24-year-old male college senior was admitted to the Indiana University Medical Center on May 21, 1974, because of persistent fever and pulmonary edema. The patient had been well until three weeks prior to admission, when the onset of fever and rapid heart rate were first noted. During the next two weeks, fever and tachycardia persisted along with dyspnea on exertion which progressed to dyspnea at rest. During graduation ceremonies, he nearly fainted on several occasions. Because of the progression of symptoms, he was admitted to another hospital. At no time did he have any pain.

Initial examination revealed fever, dyspnea, tachycardia and a pericardial friction rub. Chest X-ray demonstrated an enlarged cardiac silhouette and pulmonary edema. A diagnosis of bacterial endocarditis was made, multiple blood cultures were obtained, and large doses of penicillin and streptomycin were begun. Pulmonary edema was treated with digitalis and diuretics. On the third hospital day, the patient became obtunded and the blood pressure fell to 100/70. A pericardiocentesis was performed and 500 cc of clear, straw colored fluid was withdrawn. Because of continued clinical deterioration, decreased urine output, and an increase in blood urea nitrogen, he was transferred to the Indiana University Medical Center. Six blood cultures along
with cultures of the pericardial fluid had been negative. Prior to transfer, no murmurs were appreciated. There was no prior history of congenital or rheumatic heart disease and no history of drug abuse.

Admission physical examination revealed an asthenic Negro male in severe respiratory distress. The patient was forced to sit upright in bed. Respirations were 36/min and labored. The apical pulse was 120/min and regular. The blood pressure was 140/40 mm Hg and the temperature was 102.6°. Examination of the head, ears, eyes, nose and throat was negative. The jugular veins were distended to the angle of the jaw at 90° and pulsatile. The carotid pulsations were bounding. Moist rales were heard throughout both lung fields. The apical impulse was bounding. There was a grade III/VI systolic ejection murmur heard at the base and a grade IV/VI harsh, diastolic decrescendo murmur along the left sternal border. The liver was pulsatile. The peripheral pulses were bounding. There was no evidence of peripheral embolization. The chest X-ray showed cardiac enlargement and massive pulmonary edema. The electrocardiogram revealed sinus tachycardia with left bundle branch block.

The Echocardiogram

The initial echocardiographic examination revealed the following: left ventricular dimension at the upper limits of normal (5.6 cm); exaggerated septal and posterior wall motion suggestive of a volume overload of the left ventricle; fluttering of the anterior mitral leaflet consistent with aortic insufficiency; mid-diastolic mitral valve closure indicating rapid elevation of the left ventricular diastolic pressure (fig. 1); a dilated left atrium (4.5 cm) and an abnormal mass of
dense echoes (arrow, fig. 1) beginning immediately beneath the anterior (right) aortic leaflet which appeared to partially obstruct the left ventricular outflow tract. The anterior tricuspid leaflet appeared to move normally. There was a large mass of dense echoes (arrow, fig. 2) in the right atrium just behind the orifice of the tricuspid valve which could be seen to descend through the tricuspid orifice during early diastole and withdraw into the right atrium during late diastole. The pulmonary valve presented a unique pattern of motion (fig. 3A). The pulmonary leaflet closed normally at the end of systole; however, it was observed to reopen early in diastole (arrow) reaching a fully open position well before atrial contraction (dotted line). This observation indicated a rapid increase in early diastolic right ventricular pressure which exceeded simultaneous pulmonary artery pressure and caused valve opening.

Cardiac Catheterization

At cardiac catheterization, the right atrial pressures were markedly elevated. The right atrial a wave was 39 mm Hg, the v wave was 31, and the mean pressure was 29 mm Hg. The right ventricular pressure was 57/26. There was a rapid early diastolic rise in right ventricular pressure to 57 mm Hg (fig. 4). This rise in pressure began before the onset of atrial systole. The pulmonary artery pressure was 56/26 with a mean of 35. The pulmonary capillary wedge mean was 27.

The pressure in the descending aorta was 140/39 with a mean of 69. Because of the patient’s extremely critical condition and the suspicion of bacterial vegetations on the aortic valve, it was elected not to enter the left ventricle. Indicator dilution dye curves demonstrated a shunt from the

Figure 1

Scan from the left ventricle (LV) to the left atrium (LA) and aortic root (AO) demonstrating exaggerated posterior wall motion, fluttering and mid-diastolic closure of the anterior mitral leaflet and a dense mass of echoes beginning beneath the anterior (right) aortic leaflet and partially occluding the left ventricular outflow tract (arrow).

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Surgery

Closure of the aortic-to-right-atrium fistula and aortic valve replacement were performed on the afternoon of admission. At operation, the pericardium was thickened, vascular, and moderately adhesive to the heart. The left and noncoronary aortic cusps appeared normal. The right coronary cusp near its junction with the noncoronary cusp was perforated with approximately one-half the cusp tissue missing. Beneath the cusp was a huge mass of friable, necrotic tissue which appeared to occlude approximately one-half of the subvalvular area. A fistula was demonstrated between the junction of the noncoronary and right coronary cusps and the right atrium. A long pendulous mass of friable tissue was found protruding into the right atrium just above the annulus of the tricuspid valve near the junction of the septal and posterior leaflets. This mass was excised and the fistula closed. The large mass of necrotic tissue below the aortic valve was then excised as completely as possible and the aortic valve itself was replaced by a Bjork-Shiley prosthesis.

Postoperative Echocardiogram

The postoperative echocardiographic examination revealed a decrease in the left ventricular dimension to 4.8 cm; the right ventricular dimension to 1.5 cm; and the left atrial dimension to 3.2 cm. The mass of echoes previously noted behind the tricuspid valve was no longer present. Postoperative septal motion was paradoxical and early systolic posterior motion consistent with left bundle branch block was noted.$^5$ Early diastolic opening of the pulmonic valve was no longer present (fig. 3B). Minimal posterior displacement of the pulmonic leaflet was noted at the time of atrial contraction ($< 2$ mm), a pattern consistent with pulmonary hypertension.

Valve opening following ventricular systole appeared to occur earlier in the preoperative echogram (fig. 3A) than in the postoperative recording (fig. 3B). Preoperatively right ventricular and pulmonary artery end-diastolic pressures were almost identical and partial valve opening was present prior to ventricular systole in the majority of complexes. In this situation a reduction in isovolumic contraction and early

Figure 2

Recording of the anterior tricuspid leaflet (ATV) and right atrium. A dense echo-producing mass can be seen in the right atrium (arrow) which descends into the tricuspid orifice during early diastole and withdraws into the right atrium during late diastole and systole.

aorta into the right atrium. Selective cineangiography showed massive aortic insufficiency and a large sinus of Valsalva aneurysm with rupture into the right atrium (fig. 5).

Figure 3

A) Pulmonic valve recording. The valve can be seen to open in early diastole (solid arrow) well before the onset of atrial systole. The leaflet is in an almost fully open position at the onset of atrial systole (dotted line). There is some late diastolic closure of the valve with full opening occurring again following ventricular systole. Point b = position of the leaflet at the onset of ventricular systole; PA = pulmonary artery. B) Pulmonic valve recording following closure of the aorto-cardiac fistula. Diastolic pulmonic valve opening is no longer present. Minimal diastolic opening motion of the leaflet $< 2$ mm following atrial systole is consistent with the presence of pulmonary hypertension.
block requiring epicardial pacing for two days postoperatively, a persistent increase in QRS duration, and the presence of the echocardiographic pattern of septal motion observed in patients with left bundle branch block which was not present preoperatively.

Discussion

Sinus of Valsalva aneurysm rupture is a relatively rare clinical occurrence. Without surgical correction, the prognosis following rupture is very poor, the majority of patients dying of congestive heart failure within one year. Because of the pattern of involvement of the individual sinuses and their anatomic relationship to other cardiac structures, the majority of these aneurysms rupture into the right heart. Since rupture generally occurs in young patients, has a poor prognosis and is amenable to surgical correction, it is imperative that a rapid and safe diagnosis be established.

Echocardiography has proven valuable in both the direct demonstration of cardiac structural abnormalities and the indirect indication of hemodynamic alterations. In the present case several examples of both types of changes were evident. The echocardiographic pattern produced by a sinus of Valsalva aneurysm will depend on its location within the heart. In the case described by Rothbaum et al. an unruptured right coronary sinus aneurysm dissected into the interventricular septum and presented as an abnormal

Figure 4

Pressure tracing from the right ventricle. A rapid increase in right ventricular diastolic pressure to 37 mm Hg is noted.

valve opening would be expected. In addition, although left bundle branch block was present preoperatively, several factors suggested a further delay in interventricular conduction postoperatively which may have contributed to the delay in valve opening. These factors included the necessity for extensive resection of the septum at surgery, complete heart

Figure 5

Angiogram following injection of contrast into the aortic root (AP projection). Large volume of contrast in the left ventricle is apparent, indicating severe aortic insufficiency. A jet of contrast can also be seen entering the right ventricle from the apex of the right sinus of Valsalva aneurysm. Diagram on the right illustrates diastolic flow of blood into the right heart.
echo-producing structure extending from the aortic root into the left ventricular chamber adjacent to the
interventricular septum.\textsuperscript{11} In our case the aneurysm extended into the right atrium just above the tricuspid valve and presented as a dense mass of echoes in the body of the right atrium which descended into the tricuspid orifice during diastole and withdrew into the atrium during systole. Although right atrial tumors and tricuspid valve vegetations may present as similar
echo-producing masses, the marked alteration in the pattern of pulmonic valve echo motion observed in
this case indicated the correct diagnosis. The pulmonic valve was observed to open in early diastole
almost immediately following end-systolic closure. This indicated that pressure in the right ventricle
exceeded pulmonary artery pressure in early diastole causing the valve opening. This finding was more
significant in a patient with pulmonary edema and left ventricular failure in whom pulmonary hypertension
was expected to be present. The only source of pressure available to the right ventricle during early
diastole which would exceed simultaneous pulmonary artery pressure is from the aorta via an aorta-to-right-
atrium or ventricle fistula. The exact site of entry of the fistula into the right heart is less important since
the atrium and ventricle form a functionally common chamber during diastole. This demonstration of a
rapid rise of right ventricular pressure served to distinguish the nature of the right atrial lesion since the
hemodynamic consequence of an atrial tumor or vegetations would be to restrict diastolic inflow. These
conclusions drawn from the echocardiogram were confirmed at catheterization and surgery with
demonstration of a rapid early diastolic pressure rise in the right ventricle to 37 mm Hg caused by the
aorto-atrial fistula. Postoperatively the early diastolic pulmonic valve opening was no longer present (fig.
3B). In this tracing the elevated pulmonary artery pressure, unopposed by aortic diastolic pressure,
prevented diastolic valve motion.

Other interesting echocardiographic features of this case included: the dense mass of echoes beginning
immediately below the anterior (right) aortic leaflet and extending into the left ventricle which were shown at
surgery to be caused by a mass of necrotic tissue partially obstructing the left ventricular outflow tract; the
exaggerated septal and posterior wall motion which along with fluttering of the anterior mitral leaflet
indicated aortic insufficiency; and the early diastolic closure of the mitral valve caused by elevated left ven-
tricular diastolic pressure and suggesting severe insufficiency, a poor prognosis, and a need for im-
mediate aortic valve replacement.\textsuperscript{12}

This case represents another example of the clinical value of pulmonic valve echo motion. While we have
observed diastolic opening of the pulmonic valve following atrial systole in patients with moderate and
severe pulmonic stenosis and opening throughout diastole in response to atrial contraction in a patient
with pulmonic stenosis, atrial septal defect and complete heart block, this is the only case in which
pulmonic valve opening has been observed to precede atrial contraction. The only possible cause for this
would be an aorto-cardiac fistula and we feel this sign should be specific for this entity.

Addendum

This patient was re-admitted two months following discharge in severe biventricular failure. Pulmonic valve echo motion again
demonstrated early diastolic valve opening similar to that in fig. 3A. Repeat cardiac catheterization and surgery revealed a re-
establishment of the fistula between the aorta and right atrium.

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