CASE REPORT

Pulsus Paradoxus in a Patient with Tricuspid Atresia and Hypoplastic Right Heart

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SUMMARY Increased inspiratory right ventricular filling with a consequent shift of the ventricular septum to the left, thereby decreasing left ventricular filling, has been suggested as a mechanism of pulsus paradoxus. We recently saw a patient with tricuspid atresia and hypoplastic right ventricle who developed pulsus paradoxus. His course may help clarify the contribution of this mechanism.

SEVERAL MECHANISMS have been proposed to account for the exaggerated fall in systolic blood pressure that occurs with cardiac tamponade (pulsus paradoxus). These include a fall in venous return to the left atrium caused by a decrease in the pressure gradient between pulmonary veins and left atrium during inspiration; inspiratory traction on the pericardium limiting diastolic filling; increased right ventricular filling with inspiration, raising intrapericardial pressure and limiting left ventricular filling within a limited intrapericardial volume and decreasing systemic blood pressure, possibly via shift of the interventricular septum; and an exaggeration of normal respiratory effects in the face of tachycardia and vasoconstriction. We recently saw a patient whose course clarifies the contributions of these proposed mechanisms.

Clinical Summary

EY is a 10-year-old boy with tricuspid atresia, hypoplastic right ventricle, atrial septal defect, large ventricular septal defect, and a large patent duc tus arteriosus diagnosed in infancy. By 14 months of age, he required a Blalock-Taussig anastomosis for increasing cyanosis after spontaneous closure of his patent ductus arteriosus and the development of severe right ventricular outflow obstruction. He was maintained on digoxin with stable cyanosis and self-imposed exercise limitation. Repeat cardiac catheterization at 5½ years of age demonstrated a nonrestrictive atrial septal defect and a well-functioning Blalock-Taussig shunt; however, the ventricular septal defect had become smaller. At 9½ years of age, he began to experience brief episodes of increased cyanosis and tiring and was scheduled for a Fontan procedure. Because of hypoplastic pulmonary arteries, this was not possible and instead, the rudimentary right ventricular outflow tract was opened, the ventricular septal defect was enlarged, and the Blalock-Taussig anastomosis was ligated. He did well in the immediate postoperative period, but within several days developed tachypnea and episodic mild-to-moderate dyspnea with diffuse expiratory wheezing that was partially responsive to bronchodilator therapy. Since there was a strong family history of asthma and because of his extremely anxious personality, the symptoms were ascribed to hyperactive airways. Because of fever and an increase in cardiac silhouette on chest X-ray, however, an M-mode echocardiogram was done 4 days after surgery. The echocardiogram showed a moderate-sized pericardial effusion. He was begun on aspirin and indomethacin but showed no clinical improvement. Six days after surgery, repeat M-mode and two-dimensional echocardiograms confirmed the presence of increased pericardial fluid with poor ventricular contractility (fig. 1). The next day, an 18-mm Hg pulsus paradoxus was noted, with general worsening of his condition. Central venous pressure was judged to be elevated by distention of the jugular veins to the level of the mandible when he sat up and by the collapse point of the veins of the dorsum of the hand, which was 17 cm H₂O above the mid-right atrial level. He was reoperated for surgical drainage, approximately 500 ml of serosanguinous fluid under pressure were removed. At the time of surgery, an intra-arterial catheter was inserted and a prominent pulsus paradoxus appeared on a monitor. He was extubated immediately after surgery, and while breathing spontaneously was noted to have lost his pulsus paradoxus despite continued mild respiratory distress.

Discussion

We have described a patient with tricuspid atresia and hypoplastic right ventricle who developed pulsus paradoxus associated with pericardial tamponade. Although in addition to his pericardial effusion he had some increased respiratory effort, the immediate disappearance of his pulsus paradoxus upon relief of his pericardial effusion, despite continued wheezing,
Figure 1. Apex echocardiogram taken in diastole (top) and systole (bottom) showing a large pericardial effusion (Eff). There is a diminutive right ventricle (RV) and a mitral valve can be seen between a large left ventricle (LV) and the left atrium (LA). The right atrium (RA) can also be seen and there is an atretic tricuspid valve. A = anterior; P = posterior; R = right; L = left.

strongly suggests that pericardial tamponade, rather than increased respiratory effort, was the cause of this pulsus paradoxus.

The mechanism of pulsus paradoxus during cardiac tamponade in tricuspid atresia is not clear. It cannot be due to an inspiratory increase of systemic venous return dilating the right ventricle within a tense pericardial cavity, thereby compressing the left ventricle, for two reasons. The right ventricle was diminutive and incapable of substantial expansion. Furthermore, in tricuspid atresia, all systemic venous return passes from the right to the left atrium, where it mixes with pulmonary venous return, and this combined venous return enters the large left ventricle; respiratory changes of venous return are thus always sensed by the left ventricle before the right ventricle. Thus, whereas an increase in right ventricular filling may play a role in pulsus paradoxus in the structurally normal heart, it apparently is not required and is, therefore, not the sole cause.

During tamponade, a marked fall in aortic pressure with inspiration is associated with a marked fall in left ventricular stroke output, and this could occur in tricuspid atresia for one of two reasons. Descent of the diaphragm could narrow the transverse diameter of the tense pericardium, thereby preventing adequate filling of the left ventricle. Second, inspiration could decrease pulmonary venous return by decreasing or reversing the pulmonary venous-to-left atrial pressure gradient; this mechanism implies that the inspiratory decrease in pulmonary venous return was greater than the inspiratory increase (if any) in systemic venous return, so that the total venous return to the left ventricle was decreased.

References

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