Pulmonary Arterial Pulsus Alternans Secondary to Primary Pulmonary Hypertension*


Pulsus alternans of the pulmonary artery without systemic pulsus alternans is uncommon and is associated with multiple diseases. Two cases of pulmonary hypertension with pulmonary arterial pulsus alternans and right-sided heart failure are described. Primary pulmonary hypertension was demonstrated at autopsy in both cases. These two case reports constitute another previously unreported cause for pulsus alternans in the pulmonary circuit.

Pulsus alternans was originally described by Ludwig Traube1 in 1872 and is defined as a regular alternation in the amplitude of the pulse pressure. This pulse pattern may occur independently in either the systemic or pulmonary circulations, or it may occur simultaneously in both circulations.2-5 Pulsus alternans in the systemic circuit has been most frequently observed with coronary arterial disease, aortic stenosis, and primary myocardial disease;6-8 and frequently connotes left ventricular failure. It has also been noted with paroxysmal atrial tachycardia.9 Isolated pulmonary arterial alternans has been reported with mitral stenosis,2 mitral stenosis with atrial fibrillation,10 pulmonary embolism,11 chronic obstructive pulmonary disease with cor pulmonale,2 arteriosclerotic and hypertensive heart disease with left ventricular failure.2 To our knowledge, this phenomenon has not previously been reported with primary pulmonary hypertension. This report describes two patients with primary pulmonary hypertension and right-sided heart failure who had pulmonary arterial alternans without systemic pressure alternans.

Case Reports

Case 1

A 20-year-old black woman, gravida 3, para 2, in her 24th week of gestation, was admitted on July 19, 1975, to the Kansas University Medical Center because of dyspnea, pedal edema, two-pillow orthopnea, and the recognition of a new heart murmur. These symptoms were first noted three weeks before admission. No history of hemoptysis, syncope, or leg or chest pain was elicited. At the time of admission, the patient also complained of right upper quadrant pain and easy fatigability. She was taking a multivitamin preparation, was not a drug abuser, and denied cigarette smoking.

The patient was a 160 cm (5 ft 3 in) and 53.1 kg (117 lb) black woman in mild respiratory distress with tachypnea of 24/min. The blood pressure was 110/60 mm Hg, and the pulse rate was 85 beats per minute. Examination of the lungs revealed bilateral basilar rales which cleared with coughing. On cardiac examination, there was a systolic left parasternal heave, a systolic thrust over the pulmonary artery with a palpable pulmonic closure, and an increased pulmonic component of the second heart sound. A grade 4/6 tricuspid holosystolic murmur was audible at the left lower sternal border and increased in intensity with inspiration. A grade 2/6 early pulmonic regurgitant murmur was present. Prominent third and fourth heart sounds were audible. The liver was tender and pulsatile and was 12 cm in breadth in the right midclavicular line. The lower extremities revealed pitting pedal edema (1+).

A chest x-ray film showed cardiomegaly with right ventricular prominence and an enlarged pulmonary artery. Three years prior to this admission, a chest x-ray film had demonstrated a slightly enlarged main pulmonary artery without right ventricular enlargement or cardiomegaly. The electrocardiogram showed sinus tachycardia, right axis deviation of +120°, right atrial hypertrophy, and moderate to severe right ventricular hypertrophy.

The phonocardiogram confirmed the physical findings and, in addition, demonstrated a pulmonary ejection click. On the jugular venous pulse tracing, there was a prominent "a" and "c-v" wave. The carotid pulse tracing was within normal limits, without evidence of systemic pulsus alternans (Fig 1).

The patient was treated with digitalis, diuretics, and strict bed rest, resulting in symptomatic improvement. On July 24, 1975, right cardiac catheterization was performed. The mean right atrial pressure was 11 mm Hg, with prominent "a" and "c-v" waves. The right ventricular pressure was 125/0 mm Hg with obvious pulsus alternans and an elevated right ventricular end-diastolic pressure of 20 mm Hg. The
PULMONARY ARTERIAL PULSUS ALTERNANS

Figure 1. Carotid pulse tracing was within normal limits. There is no evidence of pulsus alternans (case 1, July 24, 1975).

Pulmonary arterial pressure was 130/60 mm Hg and showed pulsus alternans (Fig. 2). The pulmonary arterial wedge pressure was 10 mm Hg. There was no intracardiac shunt. On July 25, 1975, a dose of 80 gm of urea was infused intraamniotically, and the aborted fetus was delivered, followed by a normal placenta. After the abortion, the systolic blood pressure fell to 50 mm Hg, and 500 ml of 5 percent human plasma protein fraction (Plasmanate) was given intravenously, with prompt return of the blood pressure to 110/60 mm Hg. Six hours after the abortion, respiratory arrest occurred, which was followed by shock and an unsuccessful resuscitative attempt.

At autopsy, a markedly enlarged right ventricle was noted, with a thickness of 0.5 cm for the right ventricular free wall. There were no pulmonary thromboemboli or amniotic emboli. The walls of the pulmonary arteries were twice the normal thickness, and the intima contained atherosclerotic plaques. Microscopically, the small arteries showed concentric laminar intimal fibrosis of an onionskin character. Necrotizing arteritis and plexiform lesions were present throughout both lungs.

Case 2

A 30-year-old black woman was admitted to the University of Kansas Medical Center in September 1970, one week after the delivery of a normal infant. At this time the patient complained of dyspnea on exertion, orthopnea, paroxysmal nocturnal dyspnea, and edema. Her past medical history was pertinent in that in 1966 the patient noted the insidious onset of fever and chills and was found to have sarcoidosis based on biopsy of a cervical lymph node. She was treated with alternate-day therapy with steroids and was essentially asymptomatic until her postpartum presentation at the University of Kansas Medical Center.

The patient was a 168 cm (5 ft 6 in) and 50.4 kg (111 lb) black woman in mild respiratory distress with tachypnea of 32/min. The blood pressure was 100/60 mm Hg, the heart rate was 110 beats per minute, and the patient was afebrile. Other pertinent findings included a loud pulmonic second sound, a grade 3/6 holosystolic murmur of tricuspid insufficiency, and third and fourth heart sound. The liver was tender and pulsatile and was 18 cm in breadth in the right midclavicular line. There was pitting pedal edema (2+)

The chest x-ray film showed cardiomegaly with right ventricular and right atrial prominence and an enlarged pulmonary artery. The ECG showed sinus tachycardia, right axis deviation of +120°, right atrial hypertrophy, and moderate to severe right ventricular hypertrophy. A phonocardiogram was obtained, and it confirmed the physical findings previously described. No carotid arterial alternans was seen.

A cardiac catheterization showed elevated mean right atrial pressure of 12 mm Hg with prominent "a" and "c-v" waves. The right ventricular and pulmonary arterial pressures were 90/0 and 85/35 mm Hg, respectively, and both demonstrated pulsat alternans, as shown in Figures 3 and 4, respectively. The right ventricular end-diastolic pressure was also markedly elevated at 18 mm Hg. Aortic pressure was 90/65 mm Hg and did not demonstrate systemic pulsat alternans (Fig. 5). The left ventricular end-diastolic pressure was 2 mm Hg. There was no intracardiac shunt. There was

Figure 2. Pulmonary arterial pulse tracing shows pulmonary hypertension with pulsat alternans (case 1, July 24, 1975). PA, Pulmonary arterial pressure.
no evidence of pulmonary emboli on pulmonary angiographic studies. The catheterization data and the pulmonary angiograms were consistent with primary pulmonary hypertension and tricuspid insufficiency.

The patient was treated with digitalis, diuretics, and anticoagulation therapy, but over the ensuing five years, she had progressive right-sided heart failure and died in October 1975. At autopsy, there was marked right atrial and ventricular hypertrophy. No pulmonary emboli were present. The pulmonary parenchyma was free of any lesions of sarcoidosis, but the mediastinal lymph nodes did show noncaseating granulomas. Microscopically, the pulmonary arteries showed multiple abnormalities, including atherosclerosis of the large arteries, marked medial hypertrophy, and laminar onionskin intimal fibrosis with plexiform lesions.

**Discussion**

Two cases demonstrating the phenomenon of pulmonary arterial alternans with normal carotid arterial pulse tracings were presented. Both young black women exhibited right-sided congestive heart failure and were shown to have primary pulmonary hypertension by autopsy studies. These are the first case reports of pulmonary arterial alternans associated with autopsy-proven primary pulmonary hypertension.

The physiologic basis of pulmonary or systemic pulsus alternans is unknown. Spodick and St. Pierre accounted for the immediate mechanism of pulsus alternans by two prevailing hypotheses: (1) alternating fiber length at the end of the preceding diastole, or (2) alternating contractility (ie, alternating inotropic state of the myocardium). Guntheroth et al proposed that all cells recover excitability during the refractory period, but only a portion may be restored to normal contractility. This fraction of cells are then excited but do not contract, allowing the energy from excitation to be maintained until the next beat, which then will have an augmented amount of energy of excitation. The alternate beat will be potentiated and produce the phenomenon of pulsus alternans. Katz hypothesized that the weak

**Figure 3.** This right ventricular pressure tracing shows markedly elevated right ventricular pressure (RV) of 90 mm Hg with elevated right ventricular end-diastolic pressure of 18 mm Hg. Systolic pulsus alternans is evident on this tracing (case 2, Oct. 6, 1970).

**Figure 4.** Pulmonary arterial pressure (PA) of 85/35 mm Hg with mean of 55 mm Hg demonstrated pulsus alternans (case 2, Oct. 6, 1970).
contractions seen in pulsus alternans with severe heart failure are partly due to retention of calcium at a site in the sarcoplasmic reticulum where it cannot be released to the contractile protein. Direct evidence for the existence of such an abnormality remains to be demonstrated. Because of the many unproven hypotheses proposed for the mechanism of pulsus alternans, the actual phenomenon requires further elucidation.

Ferrer et al² noted that although pulmonary or systemic hypertension was frequently associated with pulmonary and systemic alternans, hypertension was not invariably present. Pulmonary arterial pulsus alternans without systemic pulsus alternans has been reported to be associated with a variety of clinical disorders. These two cases are associated with autopsy-proven primary pulmonary hypertension. The basic pathophysiologic mechanism is not known but is most likely related to increased pulmonary resistance and right-sided heart failure.

ACKNOWLEDGMENTS: We gratefully acknowledge the secretarial assistance of Ms. Gloria Davison and editorial assistance of Ms. Sharon Stout.

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