42-year-old man presented with congestive heart failure and atrial fibrillation. He had been diagnosed with endomyocardial fibrosis 8 years earlier and was stable on diuretics. He had congested neck veins, pedal edema, and massive ascites and a loud third heart sound. ECG confirmed atrial fibrillation. The chest radiograph showed massive cardiomegaly with a cardiothoracic ratio of 90%. Two-dimensional echocardiography revealed features of endomyocardial fibrosis with predominant right ventricular involvement (online-only Data Supplement Movie I; Figure, A). The right atrium was aneurysmally dilated with dense spontaneous echo contrast. The right ventricular cavity was obliterated. The inferior vena cava measured 28 mm with no respiratory variation, suggesting high systemic venous pressures. Hepatic vein flow showed loss of a wave reversal and increased antegrade flow in inspiration. Low-pressure tricuspid regurgitation was reflected in the inspiratory systolic flow reversal in the hepatic vein Doppler (Figure B). Fibrosis of the right ventricular endocardium converted the chamber into a passive conduit. The forward flow in the pulmonary artery occurred throughout diastole and systole (online-only Data Supplement Movie II) driven by the high systemic venous pressure (Figure, C and D) and was augmented by inspiration, as illustrated in the Doppler and color M mode tracing (Figure, E and F). This report documents the natural evolution of Fontan Physiology in burnt-out endomyocardial fibrosis in the fifth decade. Endomyocardial fibrosis is a rare restrictive cardiomyopathy, with peculiar geographical distribution.1 When the right ventricle is predominantly involved, right atrial systole and elevation of systemic venous pressure maintains the right ventricular filling and forward flow. Although right ventricular systolic function is normal, severe impairment of right ventricular filling converts the right ventricle into a passive conduit. This is one of the rare situations where atrial systolic forward flow into pulmonary artery can be demonstrated.1 Onset of atrial fibrillation resulted in the loss of this atrial pump and further increase in systemic venous congestion and decline in cardiac output. This elevation of systemic venous pressure above the pulmonary arterial pressures to maintain cardiac output is referred to as the Fontan paradox, and pulmonary blood flow reflects the variation in intrathoracic pressures.2,3 The Doppler flow patterns illustrated in this report document forward flow into pulmonary arteries in diastole and systole driven by the high systemic venous pressure and varying intrathoracic pressures, as occurring in a Fontan circulation. In a surgically created Fontan circulation, the systemic venous return is connected to the pulmonary arteries without the interposition of a pumping subpulmonary ventricle. In such a circuit the postcapillary energy is no longer wasted into the systemic venous chambers, but collected and used to push the blood through the lungs by connecting the systemic veins directly to the pulmonary arteries.1 Typically, for this circuit, cardiac output is determined by the transpulmonary flow reflecting the respiratory variation in intrathoracic pressures.2,3 To the best of our knowledge, this is the first report of spontaneous evolution of Fontan physiology in an acquired heart disease reported in the literature.

Disclosures

None.

References


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**Figure.** A, Apical 4-chamber view showing massively dilated right atrium (RA) with obliterated right ventricular apex. The left atrium (LA) is also dilated. The left ventricle (LV) shows features of mild apical obliteration. B, Hepatic vein Doppler confirming atrial fibrillation with loss of a wave reversal, increased antegrade flow in inspiration, and minimal intermittent systolic flow reversal (low-pressure tricuspid regurgitation). C, Frozen frames of color Doppler scanning in systole and diastole showing the antegrade flow into the pulmonary arteries throughout the cardiac cycle. D, Color Doppler M mode echocardiogram showing the forward flow during the entire cardiac cycle into the pulmonary arteries. E and F, Continuous wave Doppler and color M mode echocardiograph of the low-velocity antegrade flow in the pulmonary artery throughout the cardiac cycle, completely dependent on venous pressure head, increasing in inspiration, and decreasing in expiration similar to Fontan physiology.
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