A 38-year-old man was admitted to our institute for chronic hypoxemia with cyanosis, secondary polycythemia (hematocrit, 67.5%; hemoglobin, 232 g/l; red cell count, 7.14 × 10^{12}/l), clubbing of the fingers and toes, decreased oxygen saturation of the peripheral arterial blood (86%), and gradually aggravating exertional dyspnea. Trans-thoracic 2- and 3-dimensional echocardiography demonstrated the presence of mitral valve cleft (A, arrow, Online Video 1), a primum atrial septal defect (B and C, asterisks, Online Videos 2 and 3), and tricuspid anterior leaflet enlargement and prolapse (B and C, arrow, Online Videos 2 and 3). Doppler echocardiography showed a left-to-right shunt at the atrial level during diastole, and the pulmonary pressure estimated by tricuspid regurgitation was 45 mm Hg. However, both transthoracic and transesophageal color Doppler imaging revealed that during systole, the lengthy and prolapsed tricuspid anterior leaflet directed the transtricuspid regurgitant blood flow not to the right atrium but to the left atrium across the atrial septal defect (D and E, double arrow, Online Videos 4 and 5), causing derangement of the desaturated venous blood flow to the left heart and thus to the systemic circulation. The surgical procedure was performed later and confirmed the described anatomic morphology. Cyanosis disappeared and arterial oxygen saturation was 99% after surgical correction. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.