A nearly 4-year-old Ethiopian girl with a 1-year history of weight loss and rapid breathing who had just immigrated to the United States presented to the emergency department with respiratory distress and increasing lethargy. Over the preceding year, she had been found to have increasing tachypnea, anorexia, a 5-kilogram weight loss, fatigue, intermittent emesis, and decreased urine output.

There were multiple notable findings on the physical examination. Abnormalities of the vital signs included a respiratory rate of 52 breaths per minute, blood pressure of 70/53, and an oxygen saturation of 85% on room air. In general, she was cachectic and lethargic. There was jugular venous distension with accentuated a waves. There were diffuse fine inspiratory crackles across the lung fields. There was asymmetrical protrusion of the left anterior chest with a visible, dynamic cardiac impulse. On auscultation, there was a regular rhythm with an S3 present. There was a III/VI, holosystolic, midfrequency, blowing murmur in the 5th intercostal space at the midclavicular line with radiation to the apex and the left axilla. A diastolic rumble was not appreciated. The liver edge was 4 centimeters below the right costal margin. Pulses were normal at the brachial and femoral arteries, bilaterally. There was no peripheral edema or clubbing.

Blood work revealed a negative troponin and a B natriuretic peptide of 2761 picograms per milliliter. A chest radiograph was performed, which showed cardiomegaly with enlargement of the left superior cardiac border, bilateral pulmonary edema, and superior deviation of the left mainstem bronchus suggestive of left atrial dilatation (Figure 1). A 15-lead electrocardiogram demonstrated sinus rhythm with left atrial enlargement and ST segment elevation in the anterior precordial leads (Figure 2).

An echocardiogram demonstrated intact atrial and ventricular septae. The left atrium was massively dilated. The annulus of the mitral valve was dilated, measuring 30 mm (z-score 7.37). There was severe mitral regurgitation via a large coaptation defect between the leaflets of the mitral valve; the defect measured 5 mm (Figure 3A). Further imaging demonstrated a large cleft in the anterior leaflet of the mitral valve that was directed toward the ventricular septum. (Figure 3B) Transesophageal echocardiography confirmed the presence of the cleft in the anterior leaflet of the mitral valve and demonstrated that a significant amount of the regurgitation arose through the cleft (Figures 3C and 3D).

The atrial septum was noted to bow severely into the right atrium to the point that the tricuspid valve orifice was effectively occluded by the atrial septum. (Figure 4A) At the onset of diastole, the mitral valve was demonstrated to open while the tricuspid valve remained closed. (Figure 4B) As the left atrium began to empty during diastole, the atrial septum moved away from the tricuspid valve orifice. (Figure 4C) As the left atrial volume continued to decrease and the atrial septum moved further away from the tricuspid valve orifice,
the tricuspid valve opened, allowing for right ventricular filling (Figure 4D). Pulsed wave Doppler interrogation of mitral and tricuspid inflows confirmed the visual findings by demonstrating a delay of 220 milliseconds from the onset of diastolic flow across the mitral valve to the onset of flow across the tricuspid valve (Figure 5). Color Doppler interrogation of the superior and inferior venae cavae demonstrated normal flow toward the right atrium during systole. (Figures 6A and 6B). There was abnormal late diastolic color flow reversal demonstrated in the venae cavae and hepatic veins immediately at the time of atrial contraction, immediately after atrial depolarization, consistent with obstructed antegrade egress across the tricuspid valve (Figure 6C and 6D). Pulsed wave Doppler interrogation in the descending aorta demonstrated evidence of pulsus alternans (Figure 7).

Due to the patient’s clinical status, the patient proceeded to surgery without further diagnostic imaging studies such as cardiac catheterization, magnetic resonance imaging, or computed tomography. At the time of operation, the cleft in the anterior leaflet of the mitral valve was noted. A ring mitral valvuloplasty was performed and the cleft in the anterior leaflet was closed primarily. Thereafter, the patient’s clinical status was markedly improved.

Giant left atrium is a rare phenomenon that has historically been seen primarily in the setting of rheumatic heart disease with mitral regurgitation as the major valvular lesion. Patients usually present in the third or fourth decade with chest pain, hemoptysis, dysphagia, and hoarseness; there has been only 1 reported case of giant left atrium presenting in childhood. Radiographic signs of giant left atrium include splaying of the main bronchi and cardiomegaly with double contours at the heart border, both of which were seen in the present patient. (Figure 1).

The opening of the tricuspid valve precedes that of the mitral valve in normal adults. Conversely, in newborns the mitral valve opens approximately 16 milliseconds prior to the tricuspid valve opening. However, in the present case, the opening of the tricuspid valve was markedly delayed (220 milliseconds) after the opening of the mitral valve. The phenomenon of atrioventricular valve dyssynchrony resulting from atrial septal occlusion of the tricuspid valve orifice has never been reported. The obstruction to flow across the tricuspid valve resulted in decreased left ventricular preload and decreased cardiac output, as demonstrated by pulsus alternans (Figure 7). Further, the obstruction resulted in prominent jugular venous a waves, Doppler evidence of late diastolic flow reversal in the systemic veins, and hepatomegaly.

An isolated cleft in the anterior leaflet of the mitral valve (ICMV), which is not associated with an atrioventricular canal defect, is an uncommon congenital cardiac malformation. In studies of human embryos, Wenink and colleagues...
have shown that complete atrioventricular canal results from a deficiency of the inlet septum, while ICMV arises due to failure of the endocardial cushions to fuse the 2 components of the anterior leaflet of the mitral valve. Early surgical repair of ICMV is recommended for cases of mild or more-than-mild mitral regurgitation, even for asymptomatic patients, due to the relative simplicity of the repair of ICMV, the low risk of reoperation or morbidity, and the increased risk of the cleft edges being more scarred and retracted in those operated later in life, thus requiring more complicated repair techniques.

The present case serves as the first reported case, to our knowledge, of atrioventricular valve dyssynchrony resulting from severe mitral regurgitation and subsequent left atrial dilatation. It also demonstrates clearly the physiological origin and repercussions, as well as several ancillary radiographic and echocardiographic findings, important to the case. Further, it demonstrates echocardiographic findings in an ICMV.

Disclosures
None.

References

Figure 3. Transthoracic (A and B) and transesophageal (C and D) echocardiographic images of the mitral valve demonstrating: A, a massively dilated left atrium (LA) and moderately dilated left ventricle (LV) with severe regurgitation by color Doppler via a large coaptation defect (D) in the valve; B, moderate left ventricular dilatation and a large cleft (C) in the anterior leaflet of the mitral valve (AM) that is directed toward the ventricular septum (S); C, a large cleft in the AM during systole—the cleft is directed toward the S and right ventricle (RV); and D, a significant amount of mitral regurgitation arising, in part, through the C in AM.
A consecutive series of apical 4-chamber echocardiographic images taken throughout 1 cardiac cycle demonstrates: A, a massively dilated left atrium (LA), both atrioventricular valves in a closed position, and a right ventricle (RV) and left ventricle (LV) that appear unremarkable; B, marked bowing of the atrial septum (AS) to the right such that the tricuspid valve (TV) orifice is completely covered—the TV remains closed while the mitral valve (MV) has opened; C, that the mitral valve is open, and the AS has shifted leftward and exposed the TV orifice—the TV remains closed; and D, that the AS is no longer obstructing the TV and the TV is now open along with the MV.

Figure 5. Pulsed wave Doppler tracings obtained at a heart rate of 100 beats per minute, demonstrating that the onset of mitral valve inflow (M) precedes the onset of tricuspid valve inflow (T). The delay (D) from the onset of M to the onset of T is 220 milliseconds.
Figure 6. Color Doppler interrogation images demonstrating: A, normal systolic antegrade flow in the superior vena cava (SVC); B, normal systolic antegrade flow in the inferior vena cava (I) and hepatic veins (H); C, abnormal color flow reversal in the SVC in late diastole after atrial depolarization; and D, abnormal color flow reversal in the I and H in late diastole after atrial depolarization.

Figure 7. Pulsed wave Doppler interrogation in the descending aorta demonstrating cycling variations in the peak velocity of the aortic flow consistent with pulsus alternans.
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