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### Authors

Yang, Eric H  
Kwon, Murray H  
Mahajan, Aman  
et al.

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# Circumferential Type A Aortic Dissection and Intimal Intussusception of the Aorta Causing Severe Aortic Regurgitation and Obstruction of the Left Main Coronary Artery

Eric H. Yang, M.D.,\* Murray H. Kwon, M.D.,† Aman Mahajan, M.D., Ph.D.,‡ John S. Child, M.D.,\* Jonathan M. Tobis, M.D.,\* Gopi Manthripragada, M.D.,\* Cheri A. Silverstein, M.D.,\* and Kamran Shamsa, M.D.\*

\*Division of Cardiology, Department of Medicine, University of California at Los Angeles Medical Center, Los Angeles, California; †Division of Cardiothoracic Surgery, Department of Surgery, University of California at Los Angeles Medical Center, Los Angeles, California; and ‡Division of Cardiothoracic Anesthesiology, Department of Anesthesiology, University of California at Los Angeles Medical Center, Los Angeles, California

(Echocardiography 2012;0:E1-E5)

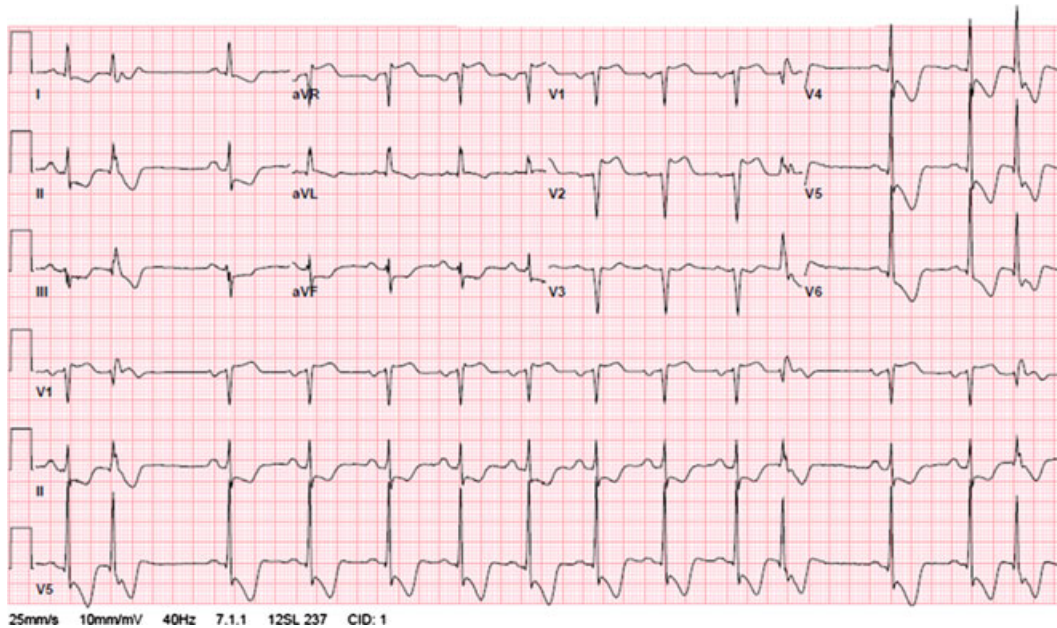
**Key words:** angiography, aortic dissection, aortic regurgitation, ST-segment elevation myocardial infarction, transesophageal echocardiography

A 58-year-old man with a history of repaired pectus excavatum presented with acute onset of severe substernal chest pain, 10/10 in severity. A Grade III/VI holosystolic murmur was heard throughout the precordium but was loudest at the apex. No diastolic murmur was audible. Bibasilar inspiratory rales were present without peripheral edema. An electrocardiogram (ECG) (GE MAC 5500, General Electric, Waukesha, WI, USA) demonstrated evidence of an anteroseptal ST segment elevation myocardial infarction (Fig. 1). An aortogram revealed significant aortic regurgitation without opacification of the coronary arteries, and an intimal tear in the ascending aorta was noted (Fig. 2, movie clip S1). Cannulating the coronary arteries was unsuccessful, which raised concern that an aortic dissection flap was obstructing the coronary ostia. The patient underwent emergent surgery. A transesophageal echocardiogram (TEE) (Philips iE33, Philips Healthcare, Andover, MA, USA) demonstrated a Stanford type A aortic dissection with a significant flap that prolapsed into the left ventricle causing severe aortic regurgitation (Fig. 3A–D, movie clips S2–S4), and obstruction of the left main coronary artery in diastole (Fig. 3E,F, movie clip S5). Exposure of the ascending aorta revealed a circumferential intimal tear which extended into

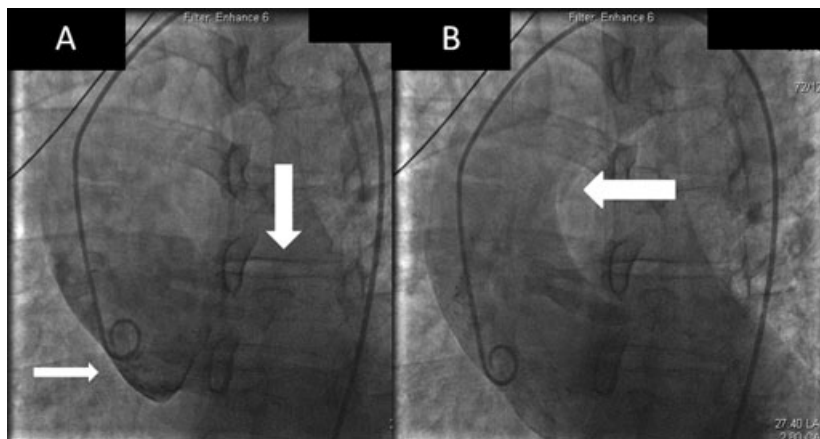
the arch and encircled the subclavian artery. The patient underwent replacement of the ascending aorta, aortic arch, and the aortic valve. Histopathological analysis of the aorta revealed evidence of acute dissection (Fig. 4). The aortic valve had 3 leaflets with myxoid change, fibrosis and focal nodular calcification. The patient's ECG changes resolved and he was discharged 11 days later in good condition without further complications.

First described by Hufnagel and Conrad,<sup>1</sup> circumferential intimal disruption of the ascending aorta with intussusception is a rare variant of aortic dissection which occurs when the intima undergoes a circumferential tear and is separated from the aortic wall forming an intimal tube which causes a "windsock" appearance. An antegrade intussusception prolapses into the aortic arch which can occlude its side branches, causing neurologic symptoms. A retrograde intussusception can prolapse into the left ventricle in diastole—preventing aortic leaflet closure causing severe aortic regurgitation and coronary ostial occlusion—and into the ascending aorta in systole. Noninvasive imaging modalities include TEE imaging, computed tomography, or magnetic resonance angiography, although the latter two can be difficult if the patient is hemodynamically unstable. Aortography may be suboptimal given that a circumferential dissection flap may not be clearly seen, as demonstrated in this case. There are 29 reported cases of intimal intussusception (17 antegrade and 12 retrograde).<sup>2</sup> Overall, prognosis is difficult to

Address for correspondence and reprint requests: Eric Yang, M.D., University of California, Los Angeles, UCLA Heart Center, 100 Medical Plaza, Suite 630, Los Angeles, CA 90095. Fax: +1-310-825-9012; E-mail: datsunian@gmail.com



**Figure 1.** Twelve-lead electrocardiogram showing sinus rhythm with premature ventricular complexes, left atrial enlargement and left ventricular hypertrophy, with ST segment elevations seen in leads V<sub>1</sub>, V<sub>2</sub>, and aVR concerning for an anterior injury current consistent with ST elevation myocardial infarction, and ST-T segment downsloping depressions concerning for subendocardial injury in the inferolateral leads.



**Figure 2.** Aortogram, left anterior oblique view, 27°. **A.** Aortography during diastole, showing complete opacification of the left ventricle (large arrow) consistent with severe aortic regurgitation. There is no opacification of the coronary arteries (small arrow) suggesting evidence of aortic dissection. **B.** Aortography showing opacification of intimal tear (large arrow) in the ascending aorta.

accurately estimate given the small number of cases reported in the medical literature.

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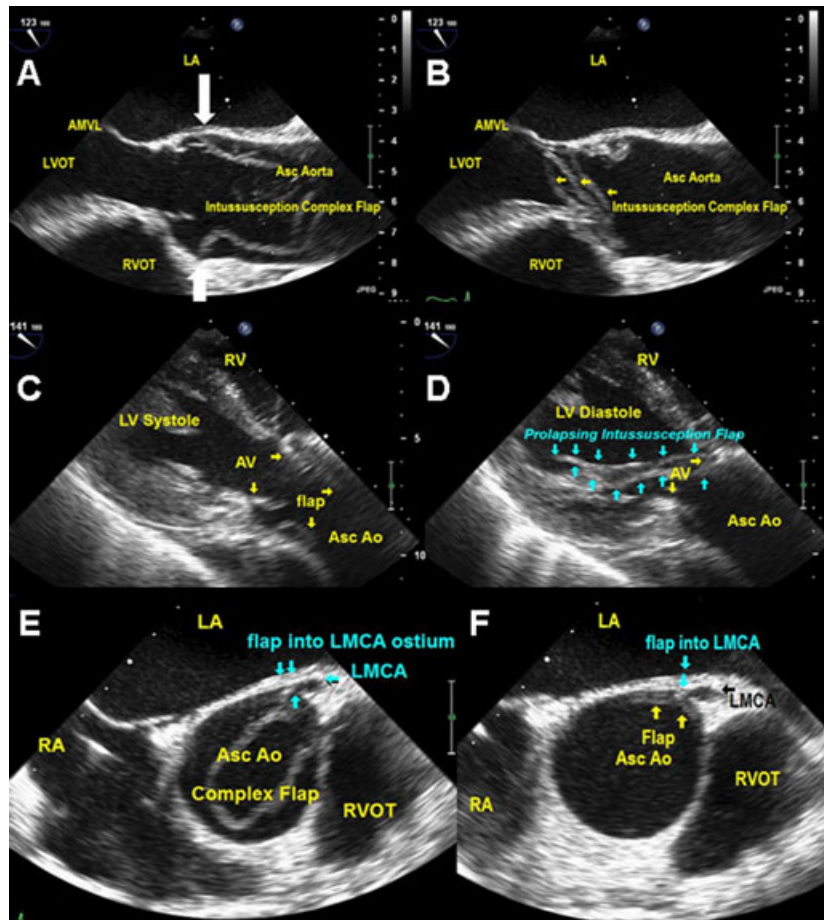
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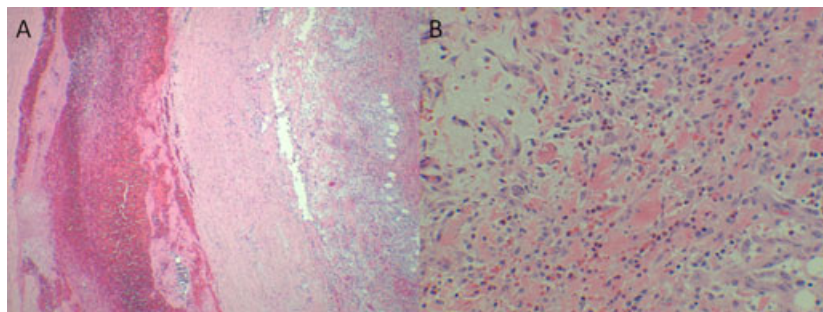
**Supporting Information**

Additional Supporting Information may be found in the online version of this article:

**Movie clip S1.** Aortogram, left anterior oblique, 27°, showing angiographic evidence of +4



**Figure 3.** Transesophageal echocardiographic views of the circumferential dissection flap. **A, B.** Mid-esophageal view, 123°. Motion of the intussusception dissection flap with its hinging point at the sinus of Valsalva (arrows) is seen in systole (**A**), which originates in the mid ascending aorta and extends retrograde circumferentially toward the sinotubular junction. The flap extends into the left ventricle during diastole (**B**), preventing closure of the aortic leaflets and contributing to severe aortic regurgitation. **C, D.** Transgastric view, 141°. The intussusception intimal tube, resembling a “windsock,” is seen in the ascending aorta during systole (**C**) and prolapsing into the left ventricle in diastole (**D**). **E, F.** Mid-esophageal view, short-axis view, 50°. The motion of the complex intussusception dissection flap seen in systole (**E**), which is circumferential in nature. The flap, when seen in diastole (**F**), obstructs the ostium of the left main coronary artery, leading to the ECG changes seen. LA = left atrium, RA = right atrium, AV = aortic valve, AMVL = anterior mitral valve leaflet, LV = left ventricle, LVOT = left ventricular outflow tract, RVOT = right ventricular outflow tract, LMCA = left main coronary artery, Asc Ao = ascending aorta.



**Figure 4.** Hematoxylin and eosin (H&E) histopathological analysis of the aortic wall. **A.** H&E stain at 40× magnification showing intramural hematoma and adventitial fat necrosis with associated chronic lymphoplasmacytic inflammation. **B.** H&E stain at 200× magnification showing prominent eosinophils.

aortic regurgitation, +3–4 mitral regurgitation, and an intimal tear in the ascending aorta. The coronary arteries do not opacify, which suggest the presence of an aortic dissection.

**Movie clip S2.** Transesophageal echocardiography, mid-esophageal view, 123° showing the aortic root and the intussusception of the aortic dissection flap prolapsing into the left ventricle during diastole and into the ascending aorta during systole. LA = left atrium, AMVL = anterior mitral valve leaflet, LVOT = left ventricular outflow tract, RVOT = right ventricular outflow tract, Asc = ascending.

**Movie clip S3.** Transesophageal echocardiography, mid-esophageal view, 123°, showing color Doppler flow across the aortic valve demonstrating severe aortic regurgitation due to the intussusceptions of the aortic dissection flap prolapsing into the left ventricle during diastole. LA = left atrium, AMVL = anterior mitral valve leaflet, LVOT = left

ventricular outflow tract, RVOT = right ventricular outflow tract, Asc = ascending.

**Movie clip S4.** Transesophageal echocardiography, transgastric view, 141°, showing the intussusception intimal tube, resembling a “windsock” prolapsing into the left ventricle in diastole. RV = right ventricle, LV = left ventricle, AV = aortic valve, Asc Ao = ascending aorta.

**Movie clip S5.** Transesophageal echocardiography, mid-esophageal view, 50°, short-axis view. The circumferential aortic dissection flap is seen occluding the ostium of the left main coronary artery in diastole leading to the ECG changes as seen on admission. In systole, the left main coronary artery is unobstructed, but there is significant lack of coronary perfusion during diastole due to obstruction by the dissection flap. LA = left atrium, RA = right atrium, Asc Ao = ascending aorta, RVOT = right ventricular outflow tract, LMCA = left main coronary artery.