Echocardiographic detection of intimo-intimal intussusception in a patient with acute Stanford type A aortic dissection

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ABSTRACT
Intimo-intimal intussusception is a very rare and unusual complication of type A dissections, typically noted on TEE exam. It has been reported in a few cases in the cardiothoracic surgical and radiology literature, and even more rarely in the cardiac anesthesia/TEE literature. This uncommon variation occurs in severe, acute, type A dissections when the ascending aortic intima circumferentially strips and detaches from the media and forms a tube-like structure which may either prolapse antegrade into the ascending aortic lumen or retrograde into the left ventricular (LV) outflow tract and LV cavity. Antegrade intussusceptions may be severe enough to partially or completely occlude the ostia of the innominate, left common carotid, and left subclavian arteries producing acute neurologic symptoms. Retrograde intussusceptions may severely impair LV filling in diastole, can worsen aortic insufficiency, mitral regurgitation, as well as produce occlusion of the coronary ostia and acute coronary ischemia. Here, we describe the incidental finding of a retrograde intussusception that was not visualized on computed tomography scan but by intraoperative TEE examination, in a patient with a severe, extensive type A dissection.

Key words: Aortic dissection; ascending; intimo-intimal intussusception; Stanford type A

INTRODUCTION
An intimo-intimal intussusception (III) is a rare and unusual manifestation of Stanford type A aortic dissection and involves a circular detachment of the intima to form an intimal “cylinder” inside the aorta. In severe cases, the detached intima may prolapse into the LV cavity. We present a patient with acute type A dissection in whom this was an incidental finding during intraoperative transesophageal echocardiography examination.

CASE REPORT
A 58-year-old male with no known medical history developed sudden onset chest pain, dyspnea, and a sensation of fullness in his throat. He was transported to the emergency room where he was found to be hypotensive (82/54 mmHg), bradycardic (40 beats/min), and tachypneic (26 breaths/min). His oxygen saturation was normal, and he was awake and alert and exhibited no neurologic deficits. His electrocardiogram showed normal sinus rhythm without acute ST-T wave changes, and his troponin T was normal. Given the concern for pulmonary embolism, a computed tomography (CT) scan with contrast was obtained. The CT scan revealed acute type A dissection arising from the aortic root and extending to the left iliac system. The left coronary ostium, all three great vessels, the celiac trunk, superior mesenteric artery, and left renal artery appeared to be involved. He was scheduled for emergent aortic reconstruction with deep hypothermic circulatory arrest.
The patient underwent induction of general anesthesia with tracheal intubation. Transesophageal echocardiography (TOE) was subsequently performed, which confirmed the presence of a dissection flap in the ascending aorta [Figure 1]. The flap was observed to prolapse across the aortic valve into the LV outflow tract during diastole [Figure 2 and Online Video 1]. Remarkably, there was no aortic valve insufficiency [Figure 3], the LV wall motion was normal, and there was no hemopericardium. Following sternotomy and opening of the pericardium, heavy bleeding through the aortic wall was encountered. Cardiopulmonary bypass (CPB) via femoral cannulation was initiated, and the patient was cooled to 18°C. During 37 min of circulatory arrest, the ascending aorta and inside curve of the aortic arch were excised and replaced with vascular graft (hemiarch). The aortic valve and root were replaced with a valved conduit, and the coronary buttons were implanted into the conduit. After the anastomosis of the valved conduit to the hemiarch graft, the patient was separated from CPB, coagulopathy was corrected, the chest was closed, and the patient transported to the intensive care unit intubated and sedated. The patient had a prolonged recovery that was complicated by cognitive impairment and critical illness neuromyopathy.

**DISCUSSION**

The term III refers to a circumferential detachment of the intima from the media, forming a loose cylinder inside the aorta. The cylinder typically inverts in an antegrade direction due to forward blood flow and causes vascular complications by obstructing perfusion through the aorta and its branching arteries.

In this case report, we describe the incidental finding by TOE of intimo-intimal intussusception with prolapse into the left ventricle, which has been rarely reported in the literature. In contrast to what one might expect, our case did not exhibit aortic regurgitation. A possible explanation is that the cylinder fell into valvular...
orifice lopsided, so that the orifice was obstructed during diastole. Although not present in our patient, severe cases of intussusception into the ventricle may be complicated by impaired ventricular filling, acute cardiogenic shock, acute myocardial infarction, mitral regurgitation as well as aortic regurgitation of varying severity. Extensive antegrade III’s may present with neurologic events of varying severity - ranging from confusion and vertigo to acute stroke symptomatology.

Given the extent of our patient’s dissection, hemiarch reconstruction with a valved conduit during deep hypothermic circulatory arrest was required. III may not be readily apparent on CT or MRI given the nature of the pathophysiology, especially if predominantly retrograde, as seen in our patient.

What must be emphasized is that circumferential dissection of the ascending aorta is a rare phenomenon - one that has been described over 120 years ago in the German literature by Bostroem in 1887.[4,9] Chiari in 1909 also referenced it in autopsy specimens[10] as an “inversion of the internal cylinder”. The phrase intimo-intimal intussusception was first described by Hufnagel et al in 1962, in the surgical literature, where they introduced the concept of aortic luminal obstruction as well as concurrent acute occlusion of the ostia of the supra-aortic vessels.[11] Virtually all of the cases of III have been reported in the surgical and vascular literature with the key presenting symptoms of these patients being chest pain in conjunction with acute neurologic symptoms such as confusion, vertigo and sudden loss of consciousness.[12,13] III has also been reported in the surgical literature in patients with descending thoracic aortic dissections, where patients have presented with clinical features of “pseudocoarctation” - referring to asymmetric peripheral pulses and asymmetric blood pressures but complete absence of neurologic symptoms.[14,15] It is important to recognize that patients with III of the aorta may not exhibit the classical imaging findings of the intimal flap in the lumen of the ascending aorta, in fact there may be no CT or MRI features suggestive of aortic dissection-until the patient is placed on cardiopulmonary bypass, or prior to that by aortography as has been described on the literature.[16] Appearance of the flap in III may also be uniquely different, as Touati et al[4] suggest, with a thick and sinuous picture that intussuscepts into the aortic lumen. It is important for the echo cardiographer to also remember that, in cases where aortic dissection is suspected, if TEE does not visualize a flap in the ascending aorta, the aortic arch must be thoroughly imaged in multiple orthogonal views for a circumferential flap. It has been suggested that this combination - of a circumferential flap in the arch in conjunction with the absence of any flap in the ascending aorta is highly predictive of III.

Surgical management can vary based on the location and severity of the III - as Touati et al suggest.[4] Less complex III’s can be managed by surgical, manual reduction of the III by returning the prolapsed intimal cylinder into the ascending aorta under deep hypothermic circulatory arrest. Supra-aortic vessel involvement will require partial or greater reconstruction of the innominate, left common carotid and left subclavian ostia and trunks.

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