

Extensive Aortic Dissection from Aortic Root to Iliac Artery



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INTRODUCTION

Aortic dissection is a life-threatening emergency if undiagnosed or not managed promptly. Dissection of the aorta is caused by intimal-medial tear and ripping, with the formation of a false lumen.^{1,2} Common predisposing factors for aortic dissection are hypertension (72.1%), atherosclerosis (31%), previous cardiac surgery (17.9%), and Marfan syndrome (MFS; 4.9%).² The Stanford classification, which is most commonly used, describes type A dissection, with the tear in the ascending aorta, and type B dissection, with the tear distal to the left subclavian artery in the descending aorta.¹ MFS is an autosomal-dominant connective tissue disorder characterized by mutations in fibrillin-1 protein, a glycoprotein that forms sheaths around elastin.³ Similarly to the general population, approximately two thirds of cases of acute aortic dissection in MFS are Stanford type A, and the rest are Stanford type B.⁴ We report a successful case of aortic root and valve surgical repair in a patient with MFS with severe aortic dissection from the ascending aorta to the bifurcation of the common iliac arteries. In this patient, Stanford type A and abdominal dissection evolved from a retrograde Stanford type B dissection over a period of 6 years.

CASE PRESENTATION

A 44-year-old female patient presented to the emergency department with refractory back and chest pain. Physical examination was significant for pectus excavatum, severe right scoliosis of the upper thoracic spine, and long fingers but with normal joint mobility. On admission, the patient had a heart rate of 58 beats/min and blood pressure of 111/51 mm Hg, symmetric in both arms. Medical history was significant for ectopia lentis treated with an intraocular lens 4 years earlier. Frontal chest radiography revealed a prominent widened mediastinum and left pleural effusion (Figure 1). Contrast-enhanced chest computed tomography demonstrated a Stanford type B and abdominal aortic dissection extending to the bifurcation of the common iliac arteries and a dilated ascending aorta (38 mm). The dissection originated at the level of the left subclavian artery, without hemodynamic compromise of the vessel. Despite iliac involvement, the patient presented with normal creatinine levels and urine output. Interestingly, the patient had two previous unremarkable pregnancies with no pregnancy-associated MFS complications (e.g., sudden aortic dissection due to increasing cardiac output, blood volume, or hormonal

changes). On the basis of clinical signs, symptoms, and the Ghent criteria (Z score = 2.77),⁵ MFS was diagnosed. Conservative pharmacologic treatment with carvedilol (6.25 mg twice daily), ramipril (2.5 mg/day), and avoidance of emotional stress was indicated upon discharge.

The patient returned to the emergency department 6 years later reporting shortness of breath, severe chest pain, and fever. No follow-up history or outpatient visits were recorded. Cardiac auscultation revealed a grade 2/6 holosystolic murmur loudest at the aortic area. Asymmetric blood pressure, 65/63 mm Hg in the right arm and 139/53 mm Hg in the left arm, was noted. Urgent transthoracic echocardiography revealed a left ventricular ejection fraction of 35%, severe left ventricular dilation, moderate systolic dysfunction, severe aortic valve insufficiency (Videos 1 and 2), moderate tricuspid insufficiency, mild mitral insufficiency without prolapse (Video 3), mild portal hypertension (47 mm Hg), and localized pericardial effusion (Figure 2A, Table 1). The ascending aorta was 1.65 times larger (62.8 mm) than 6 years earlier, representing a growth rate of 4 mm/y (Figure 2B). Computed tomography revealed that the previously diagnosed chronic Stanford type B aortic dissection had progressed to a retrograde Stanford type A dissection, with extensive intimal flap extending to the entire aorta. The aortic dissection had originated at the level of the sinotubular junction segment of the ascending aorta. In addition, the dilated ascending aorta displayed a tear with false lumen flow detected by color Doppler (Figures 2B and 2D, Video 4). Some areas of the false lumen also presented with thrombosis. Particularly, computed tomographic imaging demonstrated a distorted aorta with a spiral true lumen (Figures 3A and 3B, Video 5) and a large false lumen (Figure 3C, Video 6). The patient was immediately transferred to the cardiothoracic surgery department of a referral hospital for an aortic root and valve

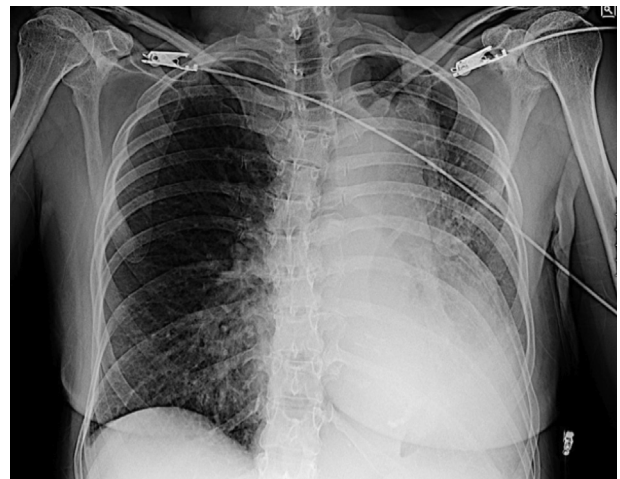


Figure 1 Frontal chest radiograph demonstrates mediastinal widening deviated to the left, diffuse cardiac silhouette with left deviation trachea, and left pleural effusion.

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VIDEO HIGHLIGHTS

Video 1: Echocardiographic apical long-axis view of dissected aorta showing severe aortic regurgitation.

Video 2: Echocardiographic apical long-axis view showing no damaged leaflets but severe dilatation causing severe aortic regurgitation.

Video 3: Echocardiographic parasternal long-axis view at the level of the mitral valve showing mild mitral regurgitation.

Video 4: Echocardiographic parasternal long-axis view of the ascending aorta. Color Doppler flow signal at the entry point of the tear at the level of the sinotubular junction.

Video 5: Extent of the aortic dissection by computed tomographic angiography. Coronal computed tomography showing dilated and distorted descending and abdominal aorta.

Video 6: Extent of the aortic dissection by computed tomographic (CT) angiography. Transverse CT view showing the intimal flap with a large false lumen in the aorta.

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replacement. Aortic root replacement was performed with a 23-mm ATS Medical composite mechanical valved graft (Figure 3D). After the operation, the patient presented symmetric blood pressures in both arms and sinus rhythm. Upon discharge, pharmacologic treatment included bisoprolol (2.5 mg/day), furosemide (40 mg/day), warfarin (3.75 mg/day), dipyron (1 g three times/day), omeprazole (20 mg/day), and potassium chloride (600 mg/day). The international normalized ratio at discharge was 2.75. The patient was safely discharged home with close cardiovascular follow-up.

DISCUSSION

Aortic dissection is a life-threatening medical emergency with elevated short-term mortality in patients with MFS. The varied symptomatology of MFS (ocular, musculoskeletal, and cardiovascular anomalies) over an extended period of time, including diffuse signs of chest pain or shortness of breath, renders aortic dissection easily disregarded.^{2,6} In the case presented, the patient was initially diagnosed with Stanford type B dissection, observed in only one third of patients with MFS and aortic dissection.⁶ Over time, pharmacologic therapy was unsuccessful, and the patient developed a retrograde Stanford A dissection associated with compromised cardiac function. In addition to Stanford type B dissection, the patient presented an abdominal dissection extending to

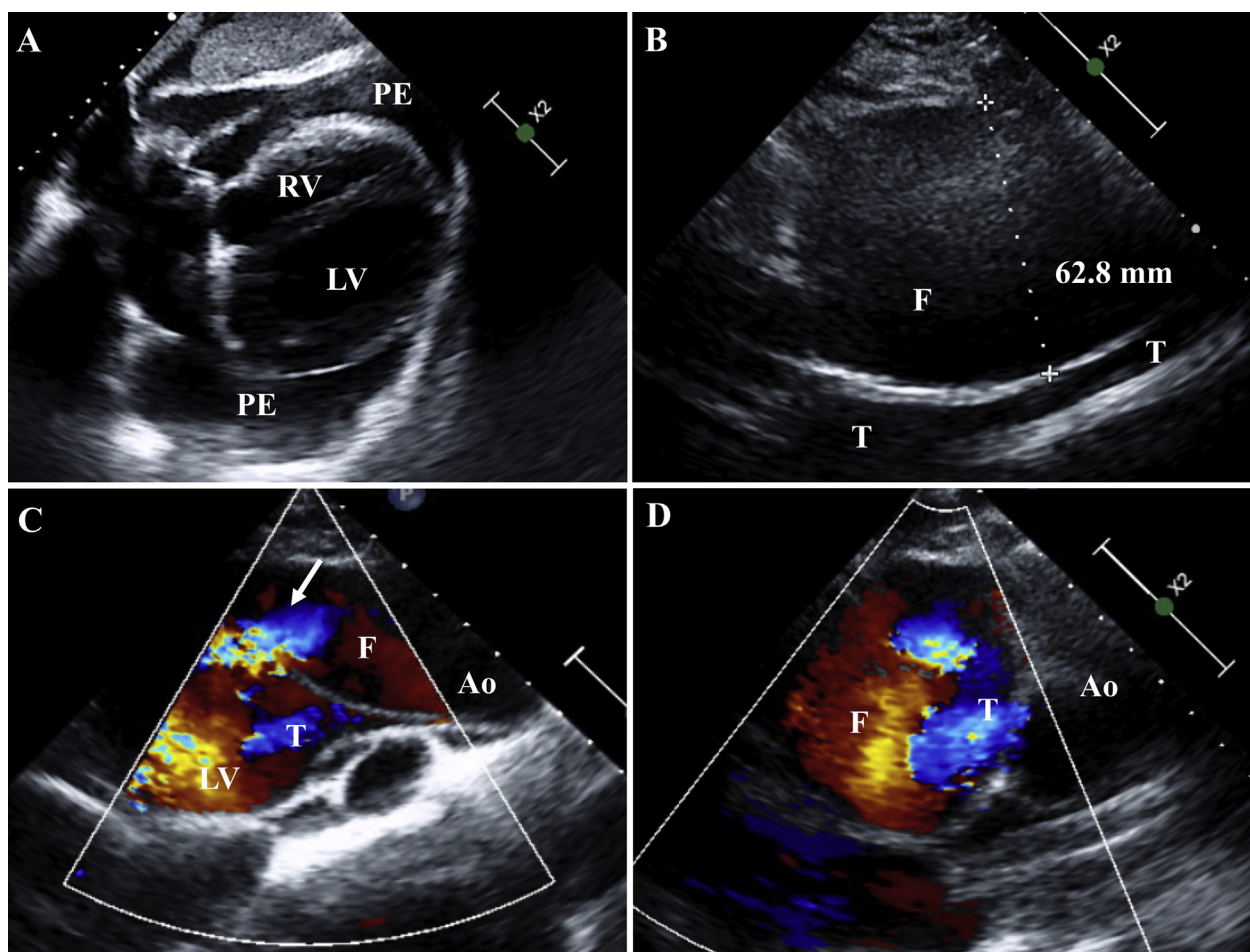


Figure 2 Transthoracic echocardiographic studies. **(A)** Subcostal view shows pericardial effusion. **(B)** Parasternal long-axis view of the dilated aortic root at second admission. **(C)** Parasternal long-axis view and Doppler color-flow imaging reveals entry tear flow into false lumen (arrow). **(D)** Modified parasternal long-axis view of the ascending aorta indicating true (T) and false (F) lumen. Doppler facilitates visualization of the intimal flap and the true and false lumen. Ao, Aorta; LV, left ventricle; PE, pericardial effusion; RV, right ventricle.

Table 1 Echocardiographic measurements on second admission

	Value
LVEDD, mm	72
LVESD, mm	53
Ascending aorta, mm	63
Aortic root, mm	58
LVEF, %	35
LVEDD index, mm/m ²	43.1
LVESD index, mm/m ²	31.7
Mitral flow, cm/sec	63
LA apex, mm	46
LA apex index, mm/m ²	27.5
LA area, cm ²	24
RA area, cm ²	19
Aortic root index, mm/m ²	37.4
LAV, mL/m ²	42

LA, Left atrial; LAV, left atrial volume; LVEDD, left ventricular end-diastolic diameter; LVEF, left ventricular ejection fraction; LVESD, left ventricular end-systolic diameter; RA, right atrial.

the common iliac arteries. Aortic root and descending aortic aneurysm and/or dissection are common findings in patients with MFS, but the presence of abdominal aortic dissection is rarely reported.^{2,7}

The American College of Cardiology recommends screening (class I) to diagnose and determine the diameter of the aortic root and ascending aorta for patients with MFS. Imaging is recommended at 6 months and then annually to assess the rate of enlargement.⁸ Pharmacological management, with β -blockers to lower blood pressure and/or the inotropic state of the heart and angiotensin receptor blockers, is also recommended.^{2,4,9} Nevertheless, inadequate surveillance and poor compliance with medication proved to be ineffective and contributed to worsen our patient's condition over a 6-year period. Notably, if left untreated, mortality in patients with type A and type B dissection can reach 58% and 31.4%, respectively.⁴ In addition, the case presented is particularly interesting because of the rapid dilation of the ascending aorta at a rate of 4.16 mm/y, compared with the average rate of 3 mm/y.² According to the guidelines, surgical repair of the ascending aorta in patients with MFS is performed with an external diameter of ≥ 5.0 cm.⁸ At second admission, the aortic diameter had reached 6.3 cm, potentially increasing the risk for rupture and an evident clinical indication for urgent surgical intervention.

Thoracic endovascular aortic repair has emerged as an alternative approach to open surgical repair.¹⁰⁻¹² In patients with connective tissue disorders (e.g., MFS and Ehlers-Danlos syndrome), the use of thoracic endovascular aneurysm repair for Stanford type B dissections is limited to exceptional cases and emergency situations as a bridge-to-surgery therapy.^{2,13} Unfortunately, thoracic endovascular aneurysm repair is not recommended in patients with MFS, because the dilation of the aorta increases reintervention rates and vascular complications, as observed in this patient.¹³

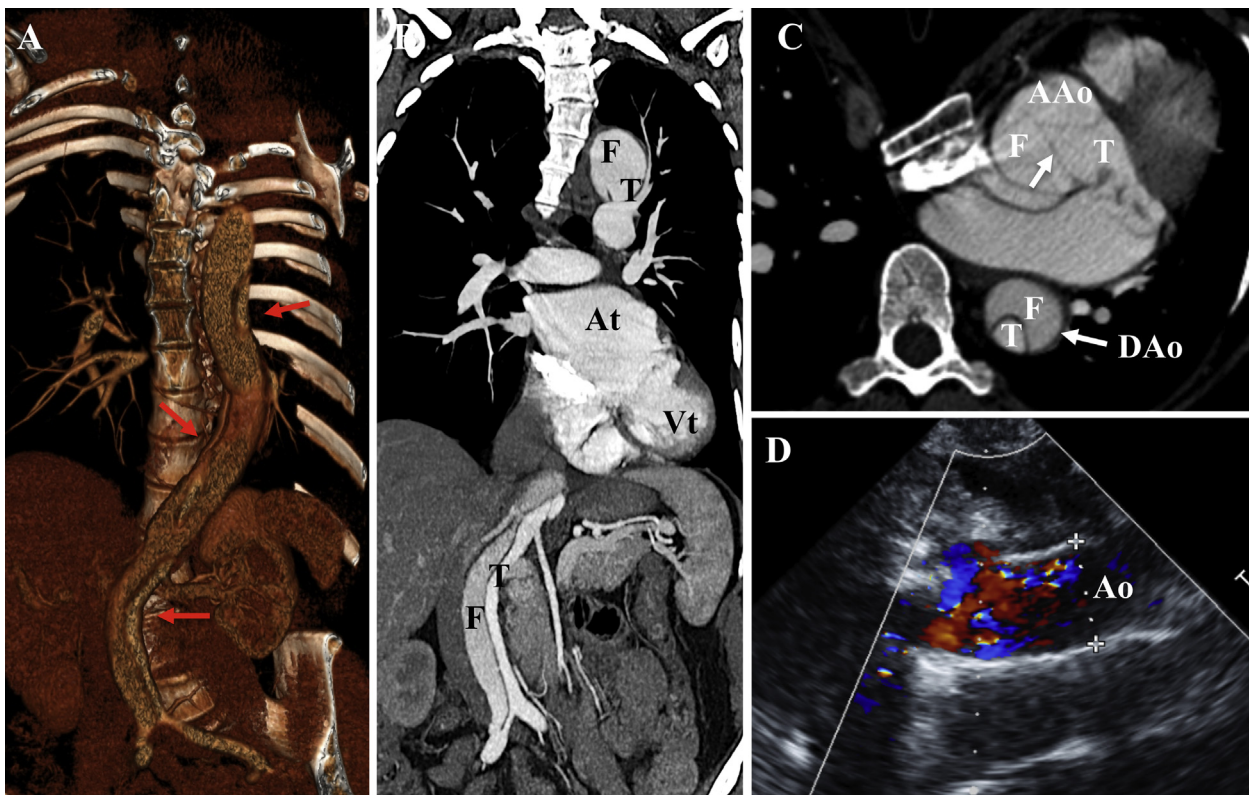


Figure 3 Extent of the aortic dissection by computed tomographic (CT) angiography. (A, B) Coronal CT view of the dilated and distorted descending and abdominal aorta (Ao). Red arrows indicate spiraled true lumen. (C) Transverse CT view of the intimal flap (arrow) separating the true (T) and false (F) lumen at the level of the ascending aorta (AAo). (D) Transthoracic echocardiographic view of the reconstructed aortic vessel with a 23-mm ATS Medical composite mechanical valved graft. At, Atrium; DAo, descending aorta; Vt, ventricle.

CONCLUSION

We report a successful case of Stanford type A dissection repair in a patient with MFS, chronic Stanford type B and abdominal dissection. Early diagnosis of MFS and initiation of prophylactic measures with β -blockers, angiotensin-converting enzyme inhibitors, and angiotensin receptor blockers can potentially reduce progression of the aortic dilation. Clinicians must consider aortic dilation in the differential diagnosis for MFS and ensure compliance with medication despite hemodynamically stability. Furthermore, adequate cardiovascular surveillance can limit MFS risks, complications, and surgery-associated hospital costs.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.04.003>.

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