

# Case Report



## Bentall surgery for acute aortic dissection (type-a) in a patient with marfan syndrome

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### Abstract

**Background:** Marfan syndrome is an autosomal-dominant hereditary connective tissue disorder with the clinical manifestations involving the ocular, skeletal, and cardiovascular systems. The cardiovascular manifestations include aortic root dilatation, aortic valvular insufficiency, mitral valve prolapse, mitral regurgitation, aortic dissection and aortic rupture. Acute aortic dissection is one of the most common catastrophes involving the aorta. A high index of suspicion is important in patients who have predisposing risk factors. Classification is based on the location of dissection and its duration. Stanford type A (De Bakey type I /type II) dissection should be treated surgically in essentially all cases. **Objective:** To report our experience in Bentall surgery in Acute aortic dissection (type A ). The efficacy of right axillary artery cannulation was investigated. **Materials & Methods:** Patient with acute type A aortic dissection involving coronary sinuses with 3 vessels of the arch free of lesions underwent aortic valve with ascending aorta and hemiarch replacement with composite valve graft (Bentall procedure) and reimplantation of coronary arteries under moderate hypothermia. The axillary artery was used for arterial cannulation. **Results:** Weaning from CPB was smooth. Perioperative period was eventless. Follow-up Echo revealed normal cardiac parameters. **Conclusion:** Prompt establishment of the diagnosis, through focused physical examination and noninvasive imaging, followed by rapid medical and surgical therapy, are the only effective methods to alter survival in patients with acute aortic dissection.

**Keywords:** Aortic Dissection, Bentall surgery, Marfan syndrome.

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### Introduction

Marfan syndrome is an autosomal-dominant hereditary connective tissue disorder with the clinical manifestations involving the ocular, skeletal, and cardiovascular systems.<sup>1</sup> It is characterized by Mutations of the FBN1 gene encoding the glycoprotein fibrillin1, which is a major component of elastic fibers of the extracellular matrix in various organs. The

diagnosis is made according to the revised 2010 Ghent<sup>2</sup> criteria (major and minor), which characterize the involvement of different organ systems, as well as genetic testing identifying a FBN1 mutation. The cardiovascular manifestations include aortic root dilatation, aortic valvular insufficiency, mitral valve prolapse, mitral regurgitation, aortic dissection and aortic rupture.<sup>3</sup>

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The prevalence of Marfan syndrome in patients presenting with acute aortic dissection ranges between 5% and 12%.<sup>4</sup> Rupture of an ascending aortic aneurysm is the most common cause of death in patients with Marfan syndrome, while type-A aortic dissection is the second most common fatal lesion.<sup>3</sup> We report our results of the surgical management of the acute aortic dissection with severe aortic regurgitation and moderate mitral regurgitation in a patient of Marfan syndrome.

## Clinical Manifestations

The most common presenting symptom is sudden onset of severe chest pain, sharp or tearing in nature, usually originating in the anterior chest or in the interscapular region, which correlates with the location of the dissection. Persistence of pain suggests continuing expansion within the chest or distal propagation. Pain is due to stretching of the aortic adventitial nerve fibers by the dissection. Other symptoms are hypertension because of preexisting arterial hypertension, high circulating levels of catecholamines, pain, or renal artery compromise by the dissection; Hypotension due to cardiac tamponade or impending rupture; Shock due to rupture or acute left ventricular failure secondary to myocardial ischemia or acute severe aortic valve regurgitation. Heart block due to involvement of the membranous and interatrial septum by a pressurized dissection related hematoma. Symptoms related to acute peripheral arterial compromise: coma, stroke, paraplegia secondary to spinal cord ischemia, upper or lower extremity ischemia, and anuria or abdominal pain due to renal or mesenteric ischemia.<sup>8</sup>

## Diagnostic Modalities

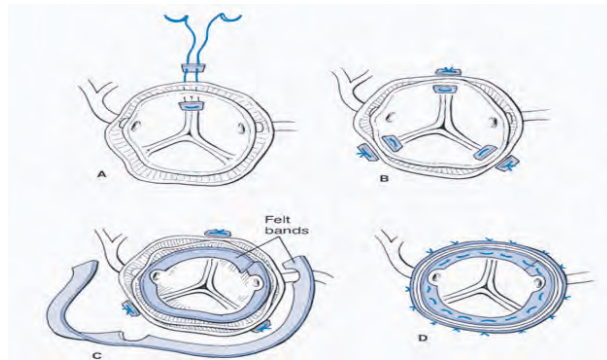
Prompt and accurate diagnosis is crucial to determine the optimal treatment strategy. The initial diagnostic step is exercising a high degree of clinical suspicion. As a general rule, the best initial diagnostic imaging study is the one that can be performed most rapidly in any particular hospital. **Chest X-ray:** widened mediastinum, (sen: 44-80%), Calcium sign -Displaced intimal calcification (>10mm) from outer aortic wall useful in older patients, A localized hump on the ascending aorta or arch, widening of the aortic knob, a double aortic shadow. **ECG:** ST-segment or T-wave changes, are non specific. **Trans thoracic Echo:** detect intimal flap and AR, Limitation: No information beyond aortic root and early part of proximal aorta. **Trans esophageal Echo:** High sensitivity (98%) and specificity (97%). Provides information about aortic valve function, flow characteristics within the true and false lumens, LV size and systolic function, and other associated valvular problems. **CT scan with contrast:** Sensitivity 98-100% & Specificity 98-100%; Noninvasive, easy, and rapid to perform, Accurate measurement of the aortic lumen diameters, Extent of dissection, arch involvement, and perfusion status of all major aortic branches; Blood flow in two distinct lumens separated by an intimal flap, Compression of the true lumen by the false lumen, Displaced intimal calcification, thrombosed false lumen or IMH. **MRA** good alternative to TEE or CT, if readily

available; High sensitivity (98%) and specificity (98%); Provides three dimensional reconstruction, Can detect site of intimal tear and involvement of branch vessels, **Aortography** considered gold standard in olden days. Sensitivity (88%) & Specificity (94%). **Fibrin degradation product (FDP) elevation:** In symptomatic patients, a plasma FDP of 12.6 µg/mL or higher suggests possible aortic dissection with a patent false lumen; An FDP level of 5.6µg/mL or higher suggests the possibility of dissection with complete thrombosis of the false lumen. **Smooth muscle myosin heavy-chain assay:** Increased levels in the first 24 hours are 90% sensitive and 97% specific for aortic dissection.<sup>9,10</sup>

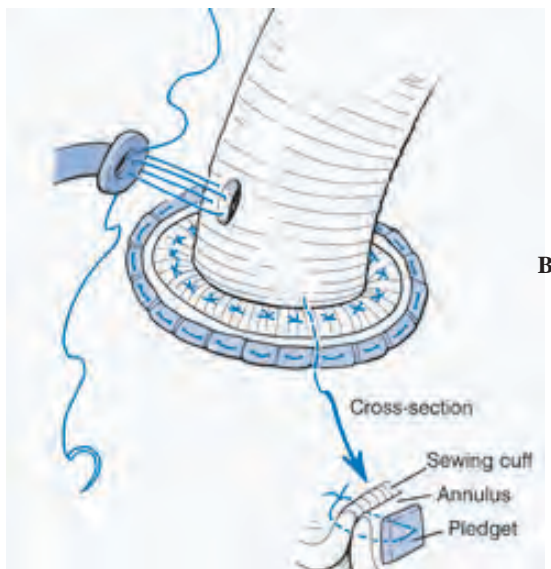
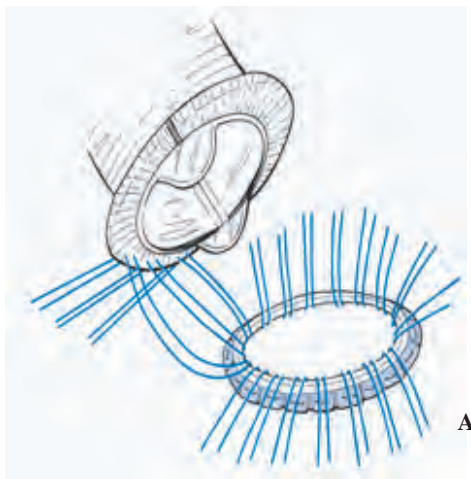
## Management

**Acute Type A Dissection:** The aim of therapy is to prevent death and irreversible end-organ damage. Emergency surgical repair of the ascending aorta to prevent life-threatening complications such as aortic rupture or tamponade. It should precede percutaneous vascular interventions, if needed. An invasive monitoring should be started. Intensive anti-impulse or negative inotropic therapy to minimize propagation of the dissection, to decrease the risk of aortic rupture, and to control pain. Hemodynamic instability suggests free aortic rupture, intrapericardial rupture with tamponade, or acute LVF secondary to severe AR or coronary compromise. In severe hypotension with evidence of tamponade, pericardiocentesis should be attempted to rise BP to the lowest acceptable level to minimize the risk of frank aortic rupture, if immediate surgical intervention is not practical.<sup>11,12</sup>

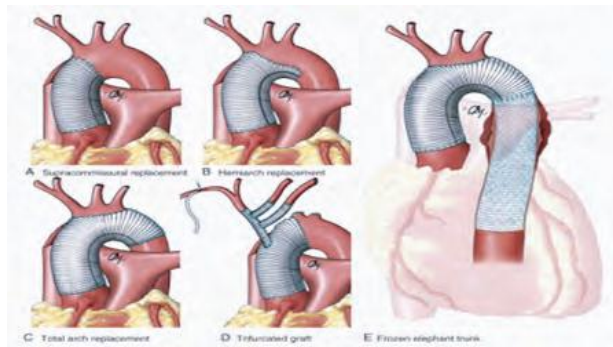
**Surgical Principles:** The primary goal is to replace the ascending aorta and proximal arch to prevent aortic rupture or proximal extension of the process with resultant tamponade. The primary intimal tear should be completely resected if it is located in the ascending aorta or the arch. The dissected aortic layers are reconstituted proximally and distally with fine continuous sutures (5-0 Prolene on C1 needle) with or without reinforcement to obliterate the false lumen. When AR is present, aortic valve competence is achieved by reconstruction of the sinuses of Valsalva and aortic root with resuspension of the commissures (Figure 4). If one or more of the sinuses of Valsalva is severely damaged, Marfan syndrome or other connective tissue disorder, a large root aneurysm, severe annuloaortic ectasia, or severe aortic stenosis then complete aortic root replacement with reimplantation of the coronary ostia is indicated by use of either a composite valve graft (CVG) (Figure 5) or a valve-sparing aortic root replacement technique, as advocated by Yacoub and David. Valve-sparing aortic root replacement using David's reimplantation method might be the ideal technique in the setting of acute aortic dissection in young patients with normal valve leaflets. Separate aortic valve replacement and supra coronary aortic graft replacement is appropriate in older patients in whom the sinuses can be salvaged but aortic valve competence is not achievable otherwise.<sup>13-15</sup>



**Figure 4:** Resuspension and preservation of the native aortic valve in a type A dissection



**Figure 5:** A. Everting 2-0 pledgeted mattress sutures are placed shoulder to shoulder around the aortic annulus to anchor a composite graft containing a St. Jude prosthesis. B. The coronary ostia are attached to the graft by the button technique using continuous 5-0 polypropylene suture.



**Figure 6:** Surgical treatment options for acute ascending aortic dissection. (A) Supra-commissural replacement of the ascending aorta. (B) Hemi-arch replacement. (C) Total arch replacement. (D) Trifurcated graft technique. (E) Frozen elephant trunk procedure.

**Technical Considerations:** Satisfactory hemostasis remains one of the chief technical challenges because of the friable dissected aortic tissue and the coagulopathy that may be present preoperatively. Replacement of the dissected aorta includes complete transection of the aorta both proximally and distally and use of full-thickness aorta-to-graft anastomoses to minimize the risks of late complications. A precise anastomotic technique is critical; deep suture bites with a continuous 5-0 or 4-0 polypropylene (3/8 circle needles) The needle must be advanced carefully on its full curve through the aortic tissue paying particular attention to the needle follow-through to avoid needle hole tears, which can cause troublesome bleeding or anastomotic disruption. If the aortic tissue is highly friable, reinforcement of the dissected aortic layers can be facilitated by re approximation of the dissection flap to the aortic wall with strips of autologous or bovine pericardium or Teflon felt. A tissue adhesive (Bio Glue, Cryo Life) is currently approved in the United States and can be used if necessary. PHCA with selective antegrade cerebral perfusion (SACP) using axillary or innominate artery CPB cannulation, has become routine and safe in patients with acute type A dissection. This allows careful inspection of the aortic arch and performance of an open distal aortic anastomosis replacing the bulk of the transverse arch. Careful inspection of the arch minimizes the possibility of leaving unrecognized intimal tears in the arch, which are present in up to 20% to 30% of patients and increase the risk of late distal aortic reoperation. Radical "hemiarch" replacement is preferred, sewing obliquely from the ligamentum arteriosum on the lesser curve of the arch to the innominate artery take-off on the greater curve, which eliminates as much dissected aorta as possible. Careful construction of a sound, completely hemostatic distal anastomosis is technically easier in the absence of an aortic cross-clamp, which itself can also traumatize the fragile dissected aortic tissue and tear the intima. The open arch replacement approach enables one to add a downstream surgical elephant trunk graft or a FET stent-graft in the true lumen of the descending aorta, which theoretically will reduce the rate of late false lumen aneurysmal degeneration (Figure 6).<sup>16,17</sup>

## Outcome

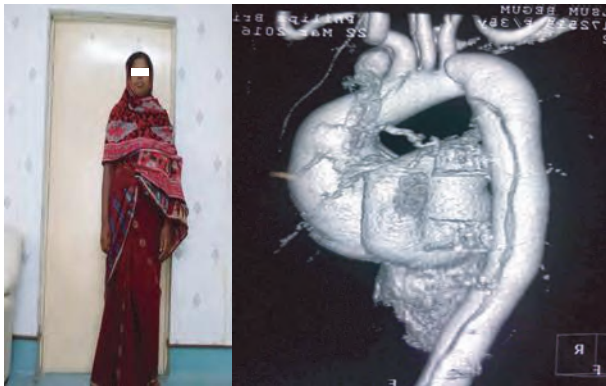
The operative mortality for repair of acute aortic dissection has fallen since DeBakey's original 40% mortality was reported in 1965. In the last two decades, most centers consistently report an operative mortality 10 to 20%. The published results for long-term survival following acute type A dissection surgically treated over the last decade are roughly 71 to 89% at 5 years and between 54 and 66% at 10 years.<sup>18,19</sup>

## Follow-Up

CTA or MRI of the thoracic and abdominal aorta at 3 to 6 month intervals during the first year and then every 12 months indefinitely. A transthoracic echocardiogram should also be performed annually to evaluate the aortic root and aortic valve function. Strict long-term blood pressure control with a combination of conventional antihypertensive agents and negative inotropic medications must be continued indefinitely. ACE inhibitors and ARB, which paradoxically increase aortic dp/dt, should be avoided.<sup>20</sup>

## Case report

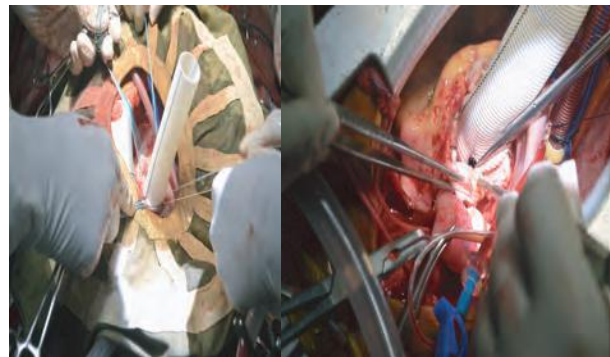
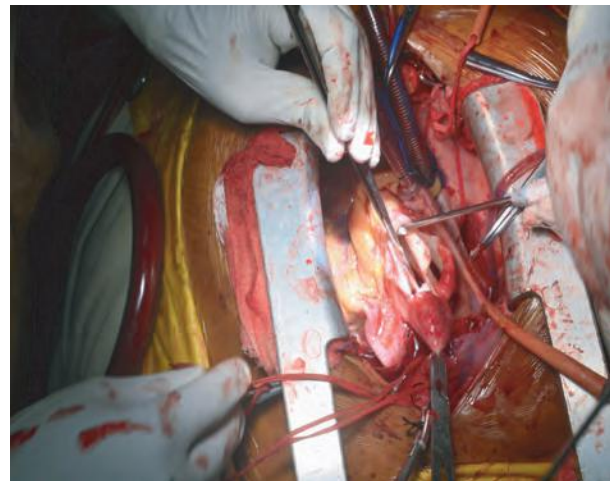
A woman of 35 yrs came to us from Chuadanga with symptoms of sudden onset of chest pain, tearing in nature and breathlessness for last 10 days. In addition to positive family history of Marfan syndrome, she had the clinical findings of long slender body stature, arachnodactyly and high arched palate (Figure 1). Her blood pressure was 150/20 mm of Hg. A systolic murmur was heard in the mitral area and an early Diastolic murmur was detected at the left sternal border.



**Figure 1:** Marfan patient **Figure 2:** CTA showing aortic dissection.

Chest radiography showed cardiomegaly and a dilated ascending aorta. TTE revealed dilated aorta (AOD-43mm), a mobile structure distal to sinutubular junction is noted which is proximally attached near to aortic valve, distal end could not be traced AR Gr III and MR Gr II+. Normal RV and LV systolic function (LVEF-62%). CT Angiogram of thoracic and abdominal aorta revealed that aortic dissection involving ascending and descending thoracic aorta extending abdominal aorta upto right common iliac artery. Dilatation of ascending aorta, measuring about 6.5 cm in maximum diameter. Descending aorta, measuring about 3 cm and abdominal aorta measuring about 2.2 cm in maximum diameter. No

involvement of renal artery. On thrombus/clot in false lumen, no calcification in intimal flap. Branches arising from aortic arch appear were observed normal (Figure 2). She was diagnosed as a case of Marfan syndrome with Acute Dissecting Aortic Aneurism (Type-A) with AR GrIII with MR GrII+. The patient was underwent Bentall procedure with Mitral valve commissuroplasty with PFO closure. CPB was established by right axillary artery and bicaval venous cannulation. LA vent was placed through RSPV. Hypothermia was done upto 24 degree C. Aortic cross clamp was applied to the arch of aorta obliquely just proximal to the brachiocephalic trunk. Aortotomy was done and Heart was arrested by direct antegrade followed by retrograde cold blood cardioplegia after opening RA. Ascending aorta was double lumened and dissected internally on lesser curvature extending above and below involving both coronary sinus. Ascending aorta was dissected from PA, RA, LA and also resected from the root of the aorta preserving both coronary sinus as coronary buttons. There was PFO in the IAS and was extended. AML was found non coapted and it was shortened by anterolateral commissuroplasty by two gathering pladgeted stiches. Mitral valve was tested by Higginson's syringe and found good co-aptation and no significant MR. The dissected coronary buttons were repaired by multiple pladgeted suture and the dissected proximal arch of the aorta by 2 PTFE felt, inside and outside.



**Figure 3:** Dissected aorta and Bentall procedure with reimplantation of coronary buttons.

Aortic ring size was measured and found 25 mm. A 23 mm SJM aortic valved graft was replaced in to the annulus by interrupted sutures. Two openings were created into the graft as coronary ostia by cautery. The Lt. coronary button anastomosed to the post. ostium. The graft was resected distally measuring the size needed. Distal end of the graft and proximal end of arch of aorta was anastomosed as end to end anastomosis. The Rt coronary button was anastomosed to ant. ostium of the graft (Figure 3). A root vent was placed into the graft. IAS was closed. LA and LV were deaired. Patient was rewarmed. RA was closed. Cross clamp was released. Heart was reverted in to sinus rhythm. Weaned off from CPB smoothly. Decannulation was done. Heparin was reversed. Haemostasis was secured. Total CPB time was 404 min and total cross clamp time was 274 min.

## Results

The post operative period was uneventful and she was discharged on 11 th post operative day. The patient made a good recovery and has been followed up for one and half years. Follow-up echo revealed normal cardiac parameters. She is still alive and no further complications.

## Discussion

Marfan syndrome occurs with a frequency of 1/10,000 in all populations. The prevalence of a positive family history has been reported as 65% to 80%.<sup>5</sup> Aortic dissection, which is common in Marfan syndrome, is usually due to an intimal tear in the proximal ascending aorta involving the sinotubular junction and aortic sinuses, resulting in prolapse of one or more commissures. However, the dissection never crosses or involves the annulus or cusps.<sup>6</sup> It is characterized by the separation of the aortic media by pulsatile blood, with variable extents of proximal and distal extension along the aorta and its branches. The process of dissection creates a false lumen in the aortic wall that parallels the aortic true lumen, allows blood flow communication between the lumens, which are separated by a dissection flap or septum.<sup>7</sup> Stanford Type A dissections involve the ascending aorta irrespective of the site of tear. Acute case diagnosed within 14 days of the onset of presenting symptoms. Mean age is 63 years. Male-to-female ratio: 2:1 to 3:1. Incidence of aortic dissection difficult to determine because many patients die without the correct diagnosis being made antemortem. There is an estimated worldwide prevalence of 0.5 to 2.95 per 100,000 per year. Incidence is higher in Japan and other Asian countries. Early mortality is as high as 1% per hour if untreated. Cause of death are intrapericardial rupture causing cardiac tamponade, acute aortic regurgitation, acute myocardial ischemia, left ventricular failure, occlusion of aortic branches supplying the cerebral or visceral circulation, free aortic rupture and distal end-organ malperfusion.<sup>4</sup>

## Conclusion

Aortic dissection is a medical emergency. many were misdiagnosed or remain undiagnosed, need high clinical suspicion and should consider as a differential diagnosis with all chest pain. Thrombolytic, Anticoagulants and anti platelet therapy may be catastrophic. Early diagnosis and prompt action improve survivality.

## Acknowledgements

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