



## Acute Type B Dissection in a Bicuspid Aortic Valve Patient During Cardiopulmonary Bypass Associated with Arch Cannulation and the use of an End-Hole Cannula

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### Abstract

The incidence of acute type B aortic dissection is 2.9 per 100,000. Bicuspid aortic valve disease is known to increase the risk to 3.1 per 10,000. An age adjusted relative risk increase of over 8-fold. Aortic dissection occurs at an earlier age along with more rapid development of aortic aneurysms. Cardiac surgery with cardiopulmonary bypass is associated with mechanical manipulation of the ascending aorta that occasionally leads to type A aortic dissection. This occurs in 0.12-0.16% of patients with cardiac surgery. The incidence of type B dissection post cardiac surgery is much lower. The incidence and management of this condition is not well defined in the literature.

We report the case of a 48-year-old man with bicuspid mixed aortic valve disease and aortopathy presenting with a 50mm ascending aortic aneurysm who underwent minimally invasive aortic valve and ascending aortic replacement that led to an acute Stanford type B/DeBakey IIIb aortic dissection. The patient was successfully treated with thoracic endografting and remains well at 2-year follow-up. We describe the management of the case and focus on sites of operative trauma at surgery. We discuss aortic morphology during primary surgery and consider the aortic histopathology at the site of primary entry tear.

Current guidelines do not provide conclusive information on characteristic features and management in these patients. We discuss emergent thoracic aortic endografting as a treatment option for these high-risk patients and discuss all the management strategies that would improve outcomes for this patient cohort.

**Keywords:** Bicuspid aortopathy; Aortic valve replacement; Ascending aortic replacement; Complication; Aortic dissection; Thoracic endovascular aortic repair (TEVAR)

### Abbreviations

ATBAD: Acute Type B Aortic Dissection; BAV: Bicuspid Aortic Valve; CTA: Computer Tomographic Angiography; TEVAR: Thoracic Endovascular Stent Grafting

### Introduction

The incidence of acute type B aortic dissection is 2.9 per 100,000 in the population [1]. Bicuspid aortic valves (BAV) are the commonest congenital cardiac defect with a prevalence estimated between 0.5 to 2% [2]. Aortic aneurysms associated with BAV are a manifestation of multiple single defects, for instance mutations in signalling and transcriptional regulators of NOTCH 1 (9q34.3) that lead to abnormal valve development and de-repression of calcium signalling [3]. Mutations in GATA 5 have also been reported [4]. There are numerous other gene defects that are described in extracellular matrix proteins such as FBN1, FN1, FGF8, HOXA1 and TGFBR1/2 [5]. Histological abnormalities are observed in bicuspid aortopathy as the result of abnormal regulatory pathways involving smooth muscle cells within the aortic media affecting the structural integrity and flexibility of the aorta [6]. The resulting aortopathy develops early and both children and adults with bicuspid aortic valve disease have larger aortic annuli, sinuses and proximal ascending aortae than those with tri-leaflet valves. The reported increased incidence of aortic dissection varies from 0 to 10-fold compared with patients with aortic stenosis [7].

Iatrogenic aortic dissection is a rare but major complication in invasive vascular interventions [6]. Whereas the ascending thoracic aorta is usually affected in cardiac surgery [8,9] dissection of the descending thoracic aorta is commonly caused by manipulation of intraluminal tools introduced *via*.

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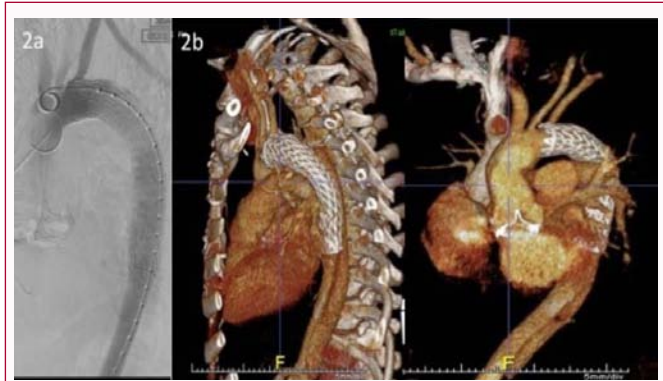
**Figure 1:** Stanford type B/ Debakey Type IIIb aortic dissection extending below the diaphragm. A) CT-Angiography sagittal view, B) 3D reconstruction (flap seen red arrow; occlusive thrombus and flap extending into the region of the visceral vessels seen blue arrow).

the femoral artery for catheter-based interventions. Acute dissection has been described to complicate 0.12-0.16% of cardiac surgery and 0.6% of aortic valve replacements [10,11]. In these cases a low-risk elective operation is converted to an emergent to high-risk case with serious morbidity and mortality.

Guidance on management of this case is difficult as current guidelines do not provide conclusive information on characteristic features and management of acute aortic dissections with endovascular grafts. The operative mortality is between 6-66% suggesting substantial differences in management strategies and thus potential for improvements [12]. This case describes acute type B aortic dissection (ATBAD), in the setting of post-operative BAV aortopathy. We discuss the aortic morphology during primary surgery and consider the aortic histopathology at the site of primary entry tear, the importance of timely diagnosis and multi-disciplinary management including endovascular grafting as treatment options in these high-risk patients.

## Case Presentation

48-year-old man with bicuspid aortopathy presented with a 50mm ascending aortic aneurysm with moderate aortic regurgitation in a setting of preserved left ventricular function. He underwent minimally invasive aortic valve and ascending aortic replacement through a mini J-sternotomy. Cardiopulmonary bypass was established cannulating the atrium and aortic arch with an end hole aortic cannula. The aortic cross clamp was applied and the heart arrested with one litre of cold blood cardioplegic solution delivered directly into the coronary ostia. The ascending aorta was excised down to the sino-tubular junction. The aortic valve was excised and replaced with a 27 mm CE Perimount bioprosthesis. The ascending aorta was replaced with a 28 mm Dacron graft. Following routine air drill the aortic cross clamp was released, sinus rhythm regained and at normothermia, bypass was discontinued without inotropic support. Post-operative course was complicated by pericardial tamponade necessitating re-sternotomy. Patient failed to progress with worsening lactatemia and developed acute lower limb ischaemia. Computer tomographic angiography (CTA) demonstrated an acute Stanford type B/Debakey type IIIb dissection complicated by coeliac and superior mesenteric compression due to a spiralling flap (Figure 1). The patient underwent thoracic endovascular stent grafting (TEVAR) to seal the entry tear in the proximal



**Figure 2:** Gore®Tag® thoracic endoprosthesis in the proximal descending aorta. A) Fluoroscopy demonstrating deployment of the endograft B) 3D reconstruction with an demonstrating the endograft in Mitchell and Ishimaru landing zone 3, sealing the entry tear. The frame of the perimount prosthesis is seen at the LVOT.

descending aorta with a GORETAG® endograft (Figure 2). This led to angiographic resolution of the visceral malperfusion. Due to the previous physiological insult to the viscera in the setting of continued clinical deterioration, the patient underwent a laparotomy, extended subtotal colectomy, ileostomy and mucous fistula formation for ischaemic bowel. Recovery was complicated with multi-organ failure and prolonged neuro-rehabilitation. Patient was discharged well with mild residual weakness in the left leg 42-days post-operatively. He remains well at 2-year follow-up.

## Discussion

Aortic dissection can complicate cardiac surgery either intraoperatively, early or late post-operatively. This converts an elective low-risk operation to high-risk surgery with a high morbidity and mortality [12].

Patients with bicuspid aortic disease are at high-risk as the underlying aorta is structurally abnormal. This pertains to the gene defects highlighted [3,4], that led to defective extracellular matrix proteins [5]. As a result matrix disruption, elastin and lamellar fragmentation is observed accompanied with increased apoptosis of vascular smooth-muscle cells. These events may lead to an aorta with weakened structural integrity and reduced elasticity [7]. At these sites aortic manipulation and trauma are the likely causes of entry tears namely cannulation site, cross-clamping injury, cardioplegia site and suture lines. The arterial flow during cardiopulmonary bypass has to be monitored and high line pressures avoided. In cases of high cannulation, vessel bifurcation points need to be avoided and the flow jet directed down the descending thoracic aorta. Use of end hole cannulae may increase wall shear stress and place the patient at risk of intimal injury. Altered aortic haemodynamics associated with BAV have been implicated in development of aortopathy [13]. BAV fusion has been shown on 4D MRI to be associated with changes in systolic flow uniformity, peak systolic velocities and flow angles. Altered flow jet patterns towards aortic wall lead to altered wall shear forces and aortopathy [14]. The structurally abnormal wall may thus be more likely to develop iatrogenic aortic dissection as in our case.

The appropriate treatment strategy for descending aortic dissection continues to be a challenge. This case highlights the importance of high clinical index of suspicion in patients not progressing post-operatively. The only clinical indication of problem was high lactate followed by eventual development of lower limb

ischaemia. Rapid diagnostic confirmation with CTA, intensive care monitoring, and aggressive blood pressure with antipulse therapy involving beta blockers are important [15].

The current indications for intervention in acute type B dissections involve rupture or signs of impending rupture, rapid diameter progression, malperfusion of abdominal or peripheral vessels, persisting pain, and uncontrollable hypertension [16]. Some authors would argue that all type B aortic dissections should be considered for endovascular repair [17].

Endografting has been shown to be an effective and feasible treatment strategy in patients with a complicated ATBAD. It is dependent on closure of the entry tear, re-expansion of the true lumen, and thrombosis of the false lumen [16]. It has been shown that the immediate outcomes are better when compared with open surgical repair, reducing in-hospital mortality and better 1 and 5-year survival compared to medical management alone [18]. In symptomatic patients or patients with visceral or end-organ malperfusion thoracic endografting leads to a favorable aortic remodeling during follow-up after intervention [19]. This is specially the case if there is false lumen thrombosis following the endografting [20].

## Conclusion

Early endovascular stent graft placement should be considered in patients with post-operative ATBAD specially in cases with evidence of end-organ malperfusion and haemodynamic instability. The use of a dispersion tip cannulae to cannulate the aortic arch should be considered to reduced shear stresses on the aorta. Long-term studies are required to delineate the use of endovascular treatment options in patients with connective tissue disease and genetically triggered thoracic aortic disease.

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