

Case Report

David procedure for aortic root aneurysm in the background of Marfan syndrome patient with severe kyphoscoliosis and previous mitral valve replacement

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ABSTRACT

Cardiac complication is one of the diseases that usually develop in Marfan syndrome patient such as mitral valve regurgitation and aortic root aneurysm. Apart from that, other congenital anomalies also tend to occur in this group of patients such as pectus excavatum and scoliosis. We report a case of successful high-risk surgery involving a Marfan syndrome patient who had underlying severe kyphoscoliosis and previous mitral valve replacement that underwent redo sternotomy and valve sparing aortic root repair (David procedure). The surgery was challenging due to the need to perform redo-sternotomy on severe pectus patient, performing valve sparing aortic root repair, and complicated with injury to right coronary artery require modified Cabrol modification for implantation of the coronary artery. The available literature about redo sternotomy on severe pectus excavatum for aortic root repair is limited, hence this paper aims to highlight the successful of performing the procedure, and the complication that might occur during the procedure.

Keywords: Aneurysm, Aorta, Marfan, Pectus, Redo-sternotomy, Cabrol

INTRODUCTION

Cardiovascular complications in patients with Marfan syndrome like dilatation and dissection of the aortic root and other segments of the aorta have been the leading cause of mortality and morbidity. Such complications can lead to life threatening conditions like aortic rupture, congestive cardiac failure and aortic valve regurgitation if it is untreated.¹ Aortic disease in Marfan syndrome patient involves impaired synthesis, secretion, and deposition of the fibrillin-1 protein, resulting from various FBN1 gene mutations. The aortic root is especially prone to dilatation and dissection.²

The life expectancy of most patients with Marfan syndrome did not exceed 40 years. However, with the introduction of aortic valve and ascending aorta replacement by Bentall and DeBono in 1968, subsequently the advent of aortic valve-sparing aortic root replacement by remodelling technique of Yacoub and colleagues in 1979 and the reimplantation procedure of David and Feindel in 1988, their life expectancy improved.^{3,4} Numerous modifications were then introduced, including the original Cabrol procedure and its modifications which were performed in cases of reoperation, severe calcification of the aorta, difficult mobilization of the coronary arteries, and large aneurysms.⁵

This case report shows the successful procedure of redo sternotomy with David procedure for aortic root aneurysm repair and modified Cabrol technique in Marfan syndrome patient with previous mitral valve replacement and severe kyphoscoliosis.

CASE REPORT

A 33-years-old male who was diagnosed with Marfan syndrome and has underlying severe kyphoscoliosis developed severe mitral regurgitation and underwent mitral valve replacement about 4 years prior. His routine echocardiography follow-up post mitral valve replacement showed good mitral valve function but progressively dilated aortic root diameter with trivial aortic valve insufficiency. His aortic root diameter increased from 4.2 cm to 5.5 cm within 6 months. Computed tomography angiography (CTA) of thoracic aorta showed dilated aortic root with measurement of 3.3 cm at the level of aortic annulus, 5.7 cm at sinus of valsalva, 3.4 cm at sinotubular junction, and 2.6 cm at ascending aorta with the rest of the thoracic aorta was normal in calibre.

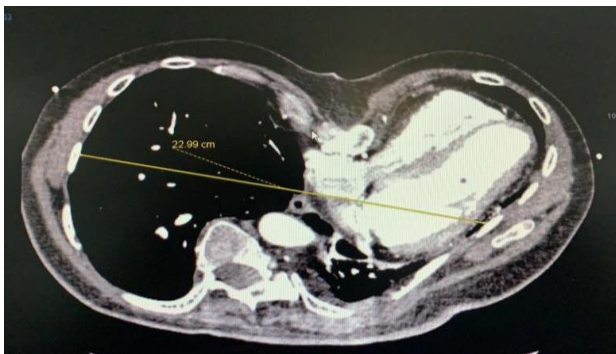


Figure 1: CT scan showing severe pectus excavatum, scoliosis of vertebra and lateral displacement of the heart.

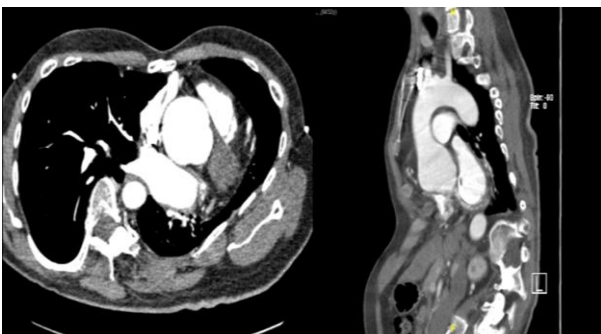


Figure 2: CT scan showing aortic root dilatation with background of kyphoscoliosis.

There were no aortic dissection or peri-aortic collection detected. We performed redo sternotomy, valve sparing aortic root repair (David procedure) with modified Cabrol technique. Intraoperatively, sternotomy was successfully performed with caution, there were extensive adhesions in the pericardial cavity and his heart size was enlarged with

dilated aortic root. Cardiopulmonary bypass was established by cannulating at distal ascending aorta and right atrium.

The aortic valve cusps were normal and preserved, and reimplanted into the Dacron graft size 30 mm. However, there was a tear over right coronary artery (RCA) button during dissection, thus modified Cabrol technique was carried out using polytetrafluorethylene (PTFE) graft size 6 mm by end-to-side anastomosis on the Dacron graft. His chest was not closed after the operation due to oedematous heart. Delayed chest closure was performed two days after initial operation. Post-operatively, he developed chylothorax as a complication.

After 3 weeks post-operative, he was discharged well and on regular follow-up. Post-operative echocardiogram at one week, six weeks, and six months showed good cardiac function with trivial aortic regurgitation only.



Figure 3: Pre-operative echocardiogram showing dilated aortic root.

DISCUSSION

Chest wall and spine deformities are well known to be associated with Marfan syndrome. This will make the sternotomy difficult and it is even harder to perform redo sternotomy and another cardiac operation on the same patient. As in our case of Marfan patient with severe kyphoscoliosis and background of previous mitral valve replacement, performing David procedure is very challenging and technically difficult. Proper planning and vigilant dissection are required to avoid inadvertent injury to another structure. Despite our attentive effort of dissection, we inadvertently injured the RCA button, hence later we did perform Cabrol technique using PTFE graft to anastomose the RCA and Dacron graft on the ascending aorta (end-to-side).

Aortic root involvement is the principal cause of death for patients with Marfan syndrome because of proximal aortic dissection and rupture. Surgical correction by replacing the aortic root and ascending aorta prevents aortic dissection and increases their lifespan.^{6,8} Aortic valve-sparing operation may further improve their longevity, but late survival is shorter compared with general population. This

is largely due to complications of residual dissection and new developments of dissection in the remaining aorta.⁷

It is recommended for patients with Marfan syndrome who have aortic root aneurysm to undergo surgery when the aortic root transverse diameter reaches 50 mm or even smaller such as 45 mm when there is a family history of aortic dissection or intending pregnancy.⁶ They are usually in their thirties or forties when surgery is required. There is a preference for aortic valve-sparing operation in young patients because of their proven durability and not needing anticoagulation. Bioprosthetic aortic valves and aortic homografts are not durable in young adults.⁹ Then, one would argue that mechanical valves are durable, but it requires lifelong anticoagulation with warfarin, which is associated with a constant risk of bleeding and thromboembolism.

Pre-operative aortic insufficiency is not a contraindication for aortic valve-sparing operation provided the cusps are reasonably elastic and the cusp prolapse is addressed during the operation, and had shown a good result.⁵ A systematic review by Burgstaller et al comparing Bentall versus the David procedure in patients with Marfan syndrome showed aortic root reimplantation is clearly associated with improved in-hospital and mid and long-term survival rates compared to composite graft group. Despite the longer cross-clamp time, longer extracorporeal circulation duration and longer circulatory arrest time, aortic root reimplantation has superior in-hospital survival. Evaluating the mortality rate in both groups, the aortic root reimplantation was constantly showing superior outcome.⁴

It has been previously reported that aortic valve-sparing procedures could be performed with acceptable operative mortality in settings associated with higher operative risk, including significant aortic insufficiency, redo cardiac surgery, and acute type A aortic dissection. But when the procedure was performed using appropriate anatomic criteria, it yields more than 90% of acceptable valve durability and freedom from moderate aortic insufficiency at midterm follow-up.¹⁰ Leontyev et al showed both the David and Bentall operations are associated with excellent early and long-term result. The David operation is associated with less bleeding than the Bentall procedure, without an increased risk of reoperation. Because of avoidance of bleeding and other long-term complications associated with prosthetic heart valves, the David procedure is preferred with appropriate pathoanatomy.¹¹

David procedure is a complex surgical procedure that requires specialised surgical training and experience to be performed successfully. However, in cases of redo cardiac surgery, difficult mobilisation of the coronary arteries, severe aortic calcification, and large aneurysms, the Cabrol technique served as pivotal role as a standalone and often as a bailout procedure.⁵ The modification of the Cabrol technique involved the anastomosis of a separate graft to mobilised ostial buttons and subsequently it is anastomosed to the aortic conduit in multiple

configurations (side-to-side, end-to-side, T-fashion). The strengths of the Cabrol technique is due to its ability to provide a safe and tension-free anastomosis of the coronary arteries to the aortic conduit when conventional reimplantation failed. Difficult dissection with friable or torn coronary ostia and poor mobilisation of the coronary arteries can happen with low origin of the coronary ostia, heavily calcified aorta, extreme aortic dilatation and dissection, and in reoperations procedures. Although early and long-term mortality of the Cabrol technique appears to be inferior to the Bentall procedure, it is an invaluable first-line procedure in few selected cases, or as a bailout procedure in complex aortic disease.⁵

CONCLUSION

Performing David procedure is very challenging however it carries a good long-term outcome especially for Marfan patient with aortic root aneurysm. The procedure is more difficult to perform on patient with the background of previous mitral valve replacement and severe chest deformity, hence complication can occur especially during redo sternotomy and dissection. Modified Cabrol technique can serve as bailout procedure for right coronary artery injury during David procedure.

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