Aortic valve repair in patients with Marfan syndrome—the "Brussels approach"

Stefano Mastrobuoni, Saadallah Tamer, Emiliano Navarra, Laurent de Kerchove, Gebrine El Khoury

Department of Cardiovascular and Thoracic Surgery, St. Luc's Hospital, Catholic University of Louvain, Brussels, Belgium

Correspondence to: Stefano Mastrobuoni. Cardiovascular and Thoracic Surgery, St. Luc's Hospital, Avnue Hippocrate 10, 1200 Brussels, Belgium. Email: stefano.mastrobuoni@uclouvain.be.

Submitted Aug 23, 2017. Accepted for publication Oct 02, 2017.
doi: 10.21037/acs.2017.10.01
View this article at: http://dx.doi.org/10.21037/acs.2017.10.01

Clinical vignette

A 27-year-old man with Marfan syndrome was referred to our department for dilatation of the aortic root with mild aortic regurgitation. His past medical history was unremarkable. Physical examination was normal except for a pectus excavatum. Transthoracic ultrasounds scan showed normal left ventricular function, mild aortic and mitral regurgitation, and dilatation of the aortic root with maximum diameter of 50 mm. An magnetic resonance imaging (MRI) further confirmed a dilatation of the aortic root up to 50 mm with mild aortic insufficiency (AI). The patient was scheduled for elective replacement of the aortic root.

Surgical technique

Preparation

Standard approach with patient under general anesthesia in the supine position, mechanical ventilation, internal jugular central venous line, radial artery cannulation (bilaterally if arch replacement is considered), trans-esophageal echo, axillary temperature and Foley catheter.

Exposition

Our standard approach is through median sternotomy. However, if the patient presents with severe pectus excavatum, a clamp-shell incision with transverse sternotomy is considered. After incision of the pericardium, conventional cannulation for cardiopulmonary bypass (CPB) of right atrium and distal ascending aorta is performed unless the patient is presenting with acute type A aortic dissection or requires a concomitant arch replacement, in which case, we prefer cannulation of the right axillary artery in order to ensure antegrade cerebral perfusion during the open distal aorta replacement. Further, in patients undergoing concomitant mitral or tricuspid valve repair, we would ensure venous drainage through bicaval cannulation.

Operation

In standard elective procedures, after clamping of the distal ascending aorta, warm blood cardioplegia is given in the ascending aorta. If there is significant aortic regurgitation, the aorta is opened transversely 1 cm above the sino-tubular junction and the cardioplegia given directly into the coronary ostia. The aortic root and valve are then assessed to confirm the indication for valve-sparing aortic root replacement (VSRR). At this point, it is indeed particularly important to carefully and systematically assess the quality of leaflets tissue, mobility, presence of fibrosis, calcification or fenestration on the cusps, in order to figure out the probability of preserving the valve and ensuring long-term durability at the same time. Small fenestrations as well as little calcifications or fibrosis can be addressed during the repair and do not necessarily represent a contra-indication to the valve sparing procedure. However, severe calcifications, extensive fibrosis or large fenestrations compromise the durability of the operation. Moreover, it is important to evaluate the height of the free margin of each cusp in order to identify concomitant cusp prolapse. The measurement of geometric and effective heights can be also helpful in this step. At this stage, the sizing of the Valsalva

© AME Publishing Company.

graft used for the reimplantation technique can be carried out (see below). The distal portion of the same graft can be eventually used for aortic arch replacement. If the patient is presenting with acute type A aortic dissection or requires arch replacement, reconstruction of the distal aorta or arch replacement are performed first; otherwise we proceed to the VSRR.

The surgical steps towards the reimplantation technique are described below and presented in the video. Overall, in elective cases, the reimplantation technique in Marfan patients is very similar to the technique described in cases of degenerative aortic root aneurism.

Root exposure and preparation

The aortic root is prepared for VSRR procedure. First, three commissural traction stitches are placed at the tip of each commissure.

Root dissection

The root is separated from the surrounding structure down to the level of ventriculo-aortic junction (VAJ). While it is quite easy to dissect the root at the level of the non-coronary sinus from the surrounding structures (left and right atrium), it is more challenging but particularly important the deep dissection of the root at the level of the left and right coronary sinus and the R-L commissure from the right ventricular outflow tract (RVOT). In order to ensure an appropriate circumferential annuloplasty at the level of the basal ring, the external dissection should indeed reach this same level. This step can be carried out with both scissors and diathermy. Diathermy is usually helpful to coagulate small vessels in the adventitia and adipose tissue around the aorta that can otherwise be a source of bleeding at the end of the procedure. Once the root is free, the sinuses of Valsalva are resected, leaving approximately 5 to 8 mm of aortic rim. The right and left coronary buttons are created leaving a generous patch of aortic wall in order to facilitate further manipulations of the same area (e.g., cannulation for cardioplegia). During the course of the reimplantation technique, warm blood cardioplegia is given intermittently through the coronary ostia at 15 minutes intervals. The cannulas are fixed to the coronary buttons in order to give the cardioplegia without disrupting the procedure.

Proximal suture line

Nine to twelve 2-0 Tycron stitches with pledget are generally used for the proximal suture line. They are distributed along the circumference of the VAJ. Sutures are passed from inside the aorta to outside with the pledgets on the inside, starting from the NC/LC commissure and moving clockwise. Along the fibrous portion of the aortic annulus, these sutures are inserted along the horizontal plane formed by the base of the inter-leaflet triangles. Notably, however, along the muscular portions of the annulus, these sutures are inserted along the lowest portion of the freely dissected aortic root, making the proximal suture line slightly higher at the LC/RC commissures compared to the LC/NC commissure. Furthermore, these stitches are placed higher than the basal ring line at the level of the RC-NC commissure in order to avoid iatrogenic lesion to the membranous interventricular septum or the atrio-ventricular node and conductive tissue. Although some surgeons use only three stitches to anchor the graft, we prefer a complete circumferential annuloplasty that permanently fix the VAJ and avoid later dilatation with the risk of recurrence of aortic insufficiency.

Graft preparation and fixation

A Dacron prosthesis with built-in neo-aortic sinuses (GelweaveValsalva^{TM} graft, Vascutek Ltd., a Terumo company, Renfrewshire, Scotland) is generally used. The graft sizing is based on the height of the commissure between the non- and the left coronary sinuses. The height is measured with a ruler from the base of interleaflet triangle to the top of the commissure. This measurement corresponds to the size of the graft that will be used. When this measurement does not correspond to a labelled graft size, the next larger size graft is chosen. The most common graft sizes range between 28 and 32 mm.

Due to the anatomy of the base of the aortic root, the proximal sutures are not in a linear plane and the proximal end of the graft need to be trimmed accordingly, particularly for the portion of the graft corresponding to the RC-NC commissure for the reasons seen above. The pledgeted sutures are then passed through the base of the prosthesis, respecting the spacing and the curvilinear contour of the proximal suture line at the RC-NC commissure. Finally, the Dacron graft is pulled down and the stitches are tied to ensure appropriate seating of the graft around the aortic annulus.

Valve reimplantation

The three commissures are re-implanted first using 4-0 polypropylene sutures. To prevent valve distortion, the

© AME Publishing Company.

three commissures must be reattached at the same level of the neo-sinotubular junction in the Valsalva graft. Radial traction is then applied on two adjacent commissural sutures to help unfold the valve within the graft and delineate the crescent-shape line of implantation. A running suture starting from the RC-NC commissure towards the RC-LC commissure and aimed to fix the rest of the valve to the graft is then performed in small regular steps passing the suture from outside the prosthesis to inside and through the aortic wall, staying close to the annulus, and then back out of the prosthesis. Particular care should be paid during this step of the operation as the correct orientation of the valve therefore its function and also the haemostasis of the reimplantation technique is dependent on this suture.

Valve testing and leaflet management

After valve reimplantation, it is critical to re-examine the leaflets for any residual prolapse, symmetry, and the height and depth of coaptation. After placing the leaflets into their closure position, a sealing test is carried out using a syringe of saline solution flushed with pressure in the neo-aortic root. The valve is then re-observed after suction of the same solution. As the Marfan syndrome is a congenital disorder of the connective tissue, it is not infrequent to observe some degree of disease also in the aortic cusps (particularly thinning, elongation of the free margin and fenestration) that should be address at this point in order to ensure durability of the repair. Cusp prolapse can be usually treated with central plication or triangular resection. As Marfan patients nowadays arrive early to surgery large fenestrations are rarely seen. Nevertheless large fenestrations, particularly those next to the commissure that can be repaired with a small pericardial patch, are not a contraindication to the procedure. Large fenestrations of the body that require larger patch compromise the durability of the repair and may contraindicate the procedure. Smaller fenestrations that do not produce significant regurgitation can be left untouched, otherwise can be corrected with free margin resuspension.

Coronary reimplantation

The left coronary button is re-implanted first. A hole of approximately 6 to 8 mm of diameter is made in the graft in face of the coronary button. The aortic tissue around the ostia is trimmed in order to leave as less tissue as possible to avoid potential late complications such as pseudo-aneurysm formation. The anastomosis is performed with a running suture of 5-0 polypropylene. A strip of autologous pericardium can be used to reinforce the anastomosis and improve the hemostasis. A similar technique is used for the anastomosis of the right coronary button. Afterwards, warm blood cardioplegia is given through the distal end of the graft with partial clamping in order to distend the new aortic root. This maneuver allows assessing hemostasis and valve competence by indirect signs such as good pressure in the root and left ventricular dilatation. A limited echocardiographic view of the aortic valve (AV) may also be obtained at this time. The cardioplegia solution is then slowly aspirated out of the prosthesis without distorting the leaflets for a last check of valve configuration.

Distal closure

Finally, the distal end of the graft is anastomosed to the distal aorta with a 4-0 polypropylene running suture.

Completion

After weaning from CPB, eventual bleeding is carefully controlled and finally the chest is closed in the standard fashion with steel wires.

Comments

VSRR in Marfan patients is indicated to replace a dilated or damaged aortic root and ascending aorta at further high risk of acute events and to correct AV dysfunction if present. Guidelines (1) recommend surgical intervention for patients with an aortic diameter of 5.0 cm or smaller if a rapid aortic dilatation defined as greater than 0.5 cm/year is proven or if there is a positive family history of aortic dissection or a significant aortic regurgitation. The goal of surgery is both to treat the current aortic disease and to prevent further life-threating complications such as ascending aortic dissection, aortic aneurysm, or false aneurism formation. Therefore, extensive replacement of the root and ascending aorta is suggested.

In Marfan patients, who usually arrive at surgery at a young age, VSRR is particularly attractive compared to composite graft (valve prosthesis + graft) (Bentall operation) in order to avoid the long-term risk of prosthesis-related complications such as thromboembolic and major bleeding events, infective endocarditis on the prosthesis and valve reoperation. However, as the VSRR procedure is technically demanding and increases the cross-clamp time, it should be performed by surgeons experienced with the procedure and in cases of acute aortic dissection it must always be balanced

© AME Publishing Company.

with the Bentall operation that is simpler and quicker to perform.

Valve-sparing root replacement procedures are generally indicated in Marfan patients electively referred for surgery without other severe comorbidities. Although the presence of significant AI is not a contraindication for these procedures however the AV must have leaflets of good quality in order to achieve a durable valve repair. In these patients we specifically use the VSRR-reimplantation technique because of the maximal support it brings to the weakened tissues in the setting of a congenital aortic wall disease and particularly to the VAJ that may later dilate and produce recurrent aortic regurgitation if left without annuloplasty (2).

Several authors have reported on the results of AV-sparing root replacement in the setting of Marfan patients (3-6). Dr. David (3) recently reported on his 20-year experience with the valve-sparing root replacement procedures in Marfan patients. Of 146 patients in this series, 121 received a VSRR. The procedure has shown low perioperative morbidity and mortality and a freedom from reoperation on the AV of around 90% at 15 years. Moreover the reimplantation technique seemed to be associated with a lower rate of recurrence of severe aortic regurgitation and reoperation compared to the remodeling technique. A similar conclusion was drawn by a previous meta-analysis (4). Dr. Svensson and colleagues (5) from the Cleveland Clinic have also reported excellent immediate and mid-term results with the VSRR-reimplantation in a series of 149 Marfan patients. Moreover, Price and colleagues (6) at Hopkins University have reported a freedom from reoperation of over 90% at 10 years and apparently improved results with the reimplantation compared to the remodeling technique. Furthermore, although the VSRR did not yield improved survival, freedom from reoperation or endocarditis, however VSRR was associated with lower thromboembolic and hemorrhagic events than the classic Bentall procedure.

From 1995 to 2017, 41 consecutive adult patients with Marfan syndrome were operated in our Institution. Of these, 31 underwent valve sparing-aortic root replacement with the reimplantation technique and 14 of them (45.2%) also had cusp repair with central plication. Further, 5 patients (16%) had concomitant mitral valve repair. The mean age of this cohort was 34±15 years and 77.4% of patients were male. No patients died at surgery. Median duration of follow-up was 5 years. During follow-up three patients (9.7%) were reoperated for recurrence of aortic regurgitation and none died at reoperation. Further late mortality risk was 9.7% in this cohort.

Conclusions

In conclusion, aortic VSRR with the reimplantation technique in patients with Marfan syndrome can be carried out without increased perioperative risk and is associated with excellent long-term results in terms of recurrence of aortic regurgitation and reoperation on the valve or the aortic root. Nevertheless, experience with the technique as well as careful patient selection are necessary to successfully perform a complex surgery in this young cohort of patients.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

References

1. Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease. A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology,American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons,and Society for Vascular Medicine. J Am Coll Cardiol 2010;55:e27-e129.
2. de Oliveira NC, David TE, Ivanov J, et al. Results of surgery for aortic root aneurysm in patients with Marfan syndrome. J Thorac Cardiovasc Surg 2003;125:789-96.
3. David TE, Feindel CM, David CM, et al. A quarter of a century of experience with aortic valve-sparing operations. J Thorac Cardiovasc Surg 2014;148:872-9; discussion 879-80.
4. Benedetto U, Melina G, Takkenberg JJ, et al. Surgical management of aortic root disease in Marfan syndrome: a

© AME Publishing Company.

systematic review and meta-analysis. Heart 2011;97:955-8.
5. Svensson LG, Blackstone EH, Alsalihi M, et al. Midterm results of David reimplantation in patients with connective tissue disorder. Ann Thorac Surg 2013;95:555-62.
6. Price J, Magruder JT, Young A, et al. Long-term outcomes of aortic root operations for Marfan syndrome: a comparison of Bentall versus aortic valve-sparing procedures. J Thorac Cardiovasc Surg 2016;151:330-6.

Cite this article as: Mastrobuoni S, Tamer S, Navarra E, de Kerchove L, El Khoury G. Aortic valve repair in patients with Marfan syndrome—the "Brussels approach". Ann Cardiothorac Surg 2017;6(6):704-708. doi: 10.21037/acs.2017.10.01

© AME Publishing Company.