An Option for Concomitant Management of Moderate Marfan Root Aneurysm at the Time of Mitral Valve Repair: A Role for Personalized External Aortic Root Support

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Two patients had mitral valve repair for severe regurgitation in the presence of a Marfan aortic root aneurysm. Concomitant personalized external aortic root support was used at the same operation to halt aneurysm progression and to correct mild aortic regurgitation.

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PEARS is a distinct procedure from wrapping the aortic root with stiff vascular graft material. The fabric used is soft, pliable, and macroporous, and the use of the patient’s digital imaging ensures an intimate fit. The mesh covers the aorta from its junction with the left ventricle to beyond the brachiocephalic trunk, and it is incorporated to form a neoaortic wall (Fig 2). Thereafter, the size and shape of the sinuses remains unchanged. External support has been shown to correct aortic regurgitation [2], which can also be achieved with PEARS.

The histologic changes that stabilize the aortic wall take time to develop. If other surgery is performed at the same time with the use of cardiopulmonary bypass, the perioperative hazard of instrumentation of the Marfan aorta is not averted. For patients with mitral valve and aortic root manifestations, the trade-off of risks and benefits in the timing and nature of surgery has to be carefully considered, by an experienced surgeon. We report just one of several possible personalized operative strategies.

Case Reports

Patient 1
A 17-year-old man with severe mitral regurgitation owing to posterior mitral leaflet prolapse also had a characteristic Marfan root aneurysm. The diameter at the level of apposition of the valve leaflets was 45 mm, and there was mild aortic regurgitation. He underwent operation in January 2015 (Fig 3).

Patient 2
A 55-year-old woman with severe mitral regurgitation owing to posterior mitral leaflet prolapse also had a characteristic Marfan root aneurysm. The aortic root diameter was also 45 mm, and she had mild aortic regurgitation. She underwent operation in March 2015 (Fig 3).

Cardiopulmonary bypass was established between bivacal cannulae (inserted through the right atrium) and the ascending aorta. Cardioplegia was delivered through the aortic root. Intraoperative examination confirmed prolapse of P2 posterior mitral leaflet without chordal...
rupture in both cases. The mitral valve surgery and PEARs are standardized procedures that were performed similarly in both patients. P2 resection and mitral annuloplasty were performed in each patient. The aortic cross-clamp was then released, and a normal sinus rhythm was restored. An external mesh support [4] was placed around the ascending aorta and root, positioned proximal to the origin of both left and right coronary...
arteries and secured to the left ventricle with interrupted sutures of 4-0 Ethibond (Fig 1C). The mesh was closed anteriorly and secured distally around the origin of the brachiocephalic artery. In view of the aortic regurgitation, the transverse dimensions of the external mesh support mesh were scaled down during manufacture to be 95% of the preoperative aortic root dimensions. M.P. performed both operations. Postoperative echocardiograms confirmed successful mitral valve repair and the absence of aortic regurgitation. Follow-up at 11 and 13 months after the surgery was satisfactory.

**Comment**

The indications and the management of mitral regurgitation were the standard of care for both of these patients with Marfan syndrome. Concomitant root replacement in this scenario is concerning because it exposes patients to an additional operative risk. In addition, there are the postoperative consequences of either a mechanical valve with a combined risk from bleeding or thrombosis of 7% per decade or reoperation for failure of the repaired aortic valve, presenting a risk of 13% per decade [5]. By halting dilatation while minimizing the additional operative risk, concomitant PEARs implantation represents an attractive option in this challenging setting.

The addition of any surgery requiring cardiopulmonary bypass compromises some of the advantages of PEARs. The desired no-touch approach to the ascending the aorta is sacrificed if the aorta is cannulated and cross-clamped, and cardioplegia is administered through the aortic root as in both these cases. Because the native aorta remains in situ rather than being replaced with a tube graft, the cross-clamp sites and other sites of instrumentation remain with an accompanying risk of medial dissection. Surgeons with experience operating on the Marfan aorta have a repertoire of techniques to ameliorate these risks. The candidates for the PEARs approach have less dilatation, and the ensleeving mesh undoubtedly reduces the strain on the aortic wall, which can mitigate the risks to some extent. However, the perioperative and subsequent risks would have been greater if surgery had been limited to the proximate problem of the regurgitant mitral valve.

On the positive side, PEARs avoids the uncertainty of a valve-sparing root replacement or life-long anticoagulation mandated by mechanical aortic valve replacement. By preserving both the aortic valve and the endothelium of the aorta, while supporting the aortic wall, the patient arguably receives a better remedy for the enlarged aorta. Should the aortic valve require replacement at some future date, the PEARs sleeve does not impede that procedure. It is clear that there is no easy solution. Whereas PEARs “personalizes” the aortic root support, it behooves the aortic surgeon to personalize the whole strategy for patients with both mitral and aortic manifestations of Marfan syndrome.

PEARs has been available at a limited number of cardiac surgical centers [1]. In these two patients, PEARs provided a solution to the clinical problem of a combination of clinically important mitral regurgitation and a root aneurysm that was not immediately life threatening by current convention, but likely to represent a hazard in the not too distant future.

**References**