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

Umberto Benedetto, PhD, Xu Yu Jin, FRCP, FRCS, Elaine Hill, FRCA, FFICM, Tom Treasure, MD, FRCS, and Mario Petrou, PhD, FRCS

John Radcliffe Hospital, Oxford; University College London, London; and Bristol Royal Infirmary, Bristol, United Kingdom

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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If patients with Marfan syndrome have mitral valve prolapse at the time of aortic root replacement, correction of mitral valve regurgitation may be performed at the same time if warranted by the severity of the regurgitation. Conversely if there is severe mitral regurgitation and an aortic root aneurysm where root replacement is not mandated, root surgery may be postponed to avoid additional perioperative risk leaving the patient prone to further dilatation and rupture. Since 2004, personalized external aortic root support (PEARS), has been under evaluation as an alternative to valve sparing root replacement (Fig 1). The first 67 patients, a median of over 6 years since surgery, provide 270 patient years of follow-up data over a 12-year period [1].

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Address correspondence to Dr Treasure, Clinical Operational Research Unit, University College London, Taviton St, London WC1H 0BT, United Kingdom; email: tom.treasure@gmail.com.

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 bicaval 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

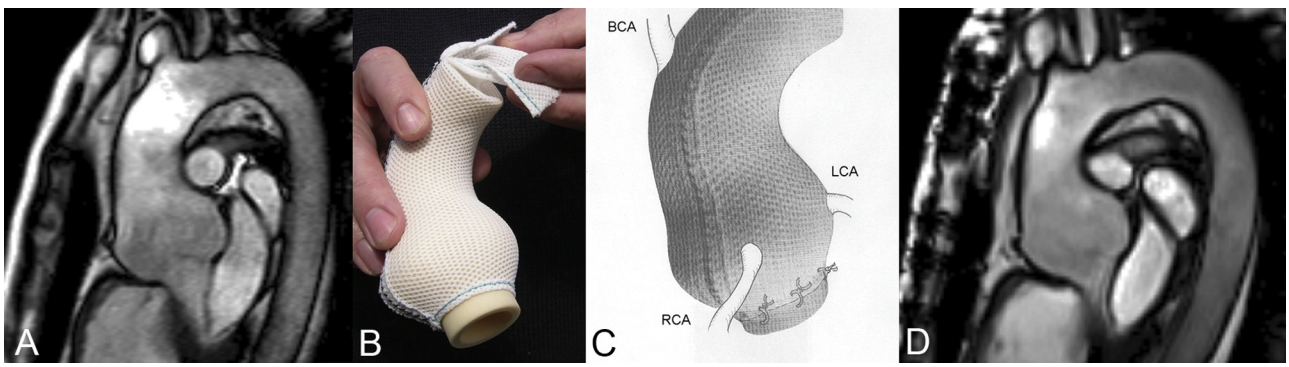

Fig 1. The essential features of personalized external aortic root support (PEARS). (A) Preoperative magnetic resonance imaging (MRI). (B) The mesh support on the thermoplastic model of the patient's aorta. (C) Depiction of mesh in position. (D) MRI in 2014, 10 years after PEARS placement. (BCA = brachiocephalic artery; LCA = left coronary artery; RCA = right coronary artery.)

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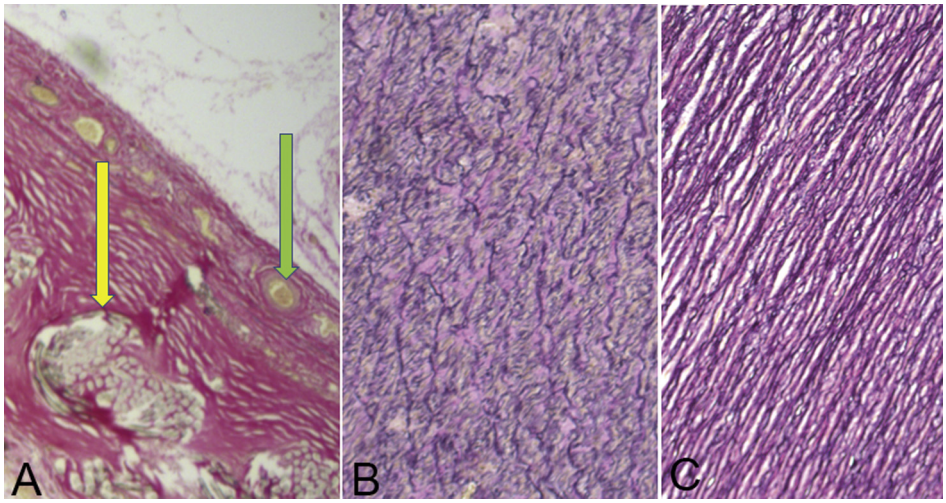

Fig 2. Histology of the mesh–aorta composite. These histologic preparations are from the only patient to have died with a mesh in place, more than 4 years after operation for a presumed arrhythmia without dissection and with a competent aortic valve [3]. (A) The mesh fibers (yellow arrow) are completely incorporated with collagen fibers running through and around the porous mesh (left panel) and new adventitial blood vessels outside the mesh (green arrow). (B) The aortic media in the unsupported arch has the appearances of Marfan syndrome. (C) The cardiac pathologists reported the histologic appearances in the proximal aorta within the mesh support to be normal with apparent healing in the supported segment.

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

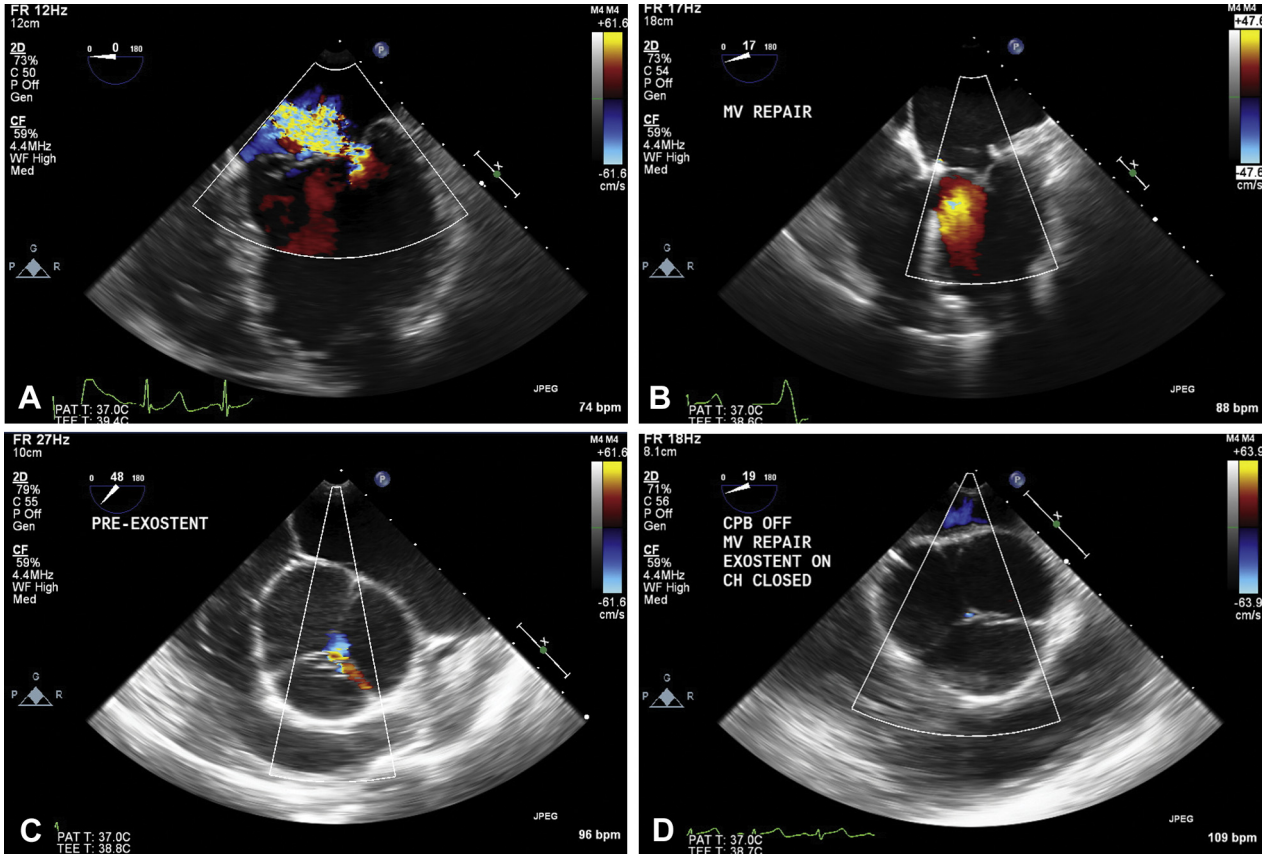

Fig 3. Transesophageal echo images of midesophageal four-chamber views (A) before and (B) after mitral valve repair in patient 2. (A) Doppler color-flow mapping demonstrated severe mitral valve regurgitation with jet directed anteriorly. Mitral regurgitation was fully corrected. (B) TOE images of the aortic valve in short axis views (C) before and (D) after personalized external aortic root support (PEARS) implantation in patient 1. (C) The Doppler color-flow mapping demonstrated mild central aortic regurgitation (AR). (D) AR was almost completely abolished after PEARS implantation.

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.

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