

An option for concomitant management of moderate Marfan root aneurysm at the time of mitral valve repair: a role for Personalised External Aortic Root Support.

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Key words: Marfan syndrome, mitral regurgitation, aneurysm

ABSTRACT

Two patients had mitral valve repair for severe regurgitation in the presence of a Marfan aortic root aneurysm. Concomitant Personalised External Aortic Root Support (PEARS) was used at the same operation to halt aneurysm progression and to correct mild aortic regurgitation.

Introduction

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 it may be postponed to avoid additional perioperative risk leaving the patients prone to further dilatation and rupture. Since 2004 personalised 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 six years since surgery, provide 270 patients years of follow-up data over a twelve year period. [1]

PEARS is a quite 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 is incorporated to form a neo-aortic 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 histological changes, which stabilise the aortic wall, take time to develop and 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. Here we report just one of several possible personalised operative strategies.

Cases

Case 1. A 17 year old man with severe mitral regurgitation due 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 was operated on in January 2015. (Fig.3)

Case 2. A 55 year old woman with severe mitral regurgitation due to posterior mitral leaflet prolapse also had a characteristic Marfan root aneurysm. The aortic root diameter was also 45 mm and she too had mild aortic regurgitation. She was operated on in March 2015. (Fig.3)

Cardiopulmonary bypass was established between bicaval cannulae (inserted via the right atrium) and the ascending aorta. Cardioplegia was delivered via 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 standardised procedures and were performed similarly in both patients. P2 resection and mitral annuloplasty was 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 aortic root. 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.

Discussion

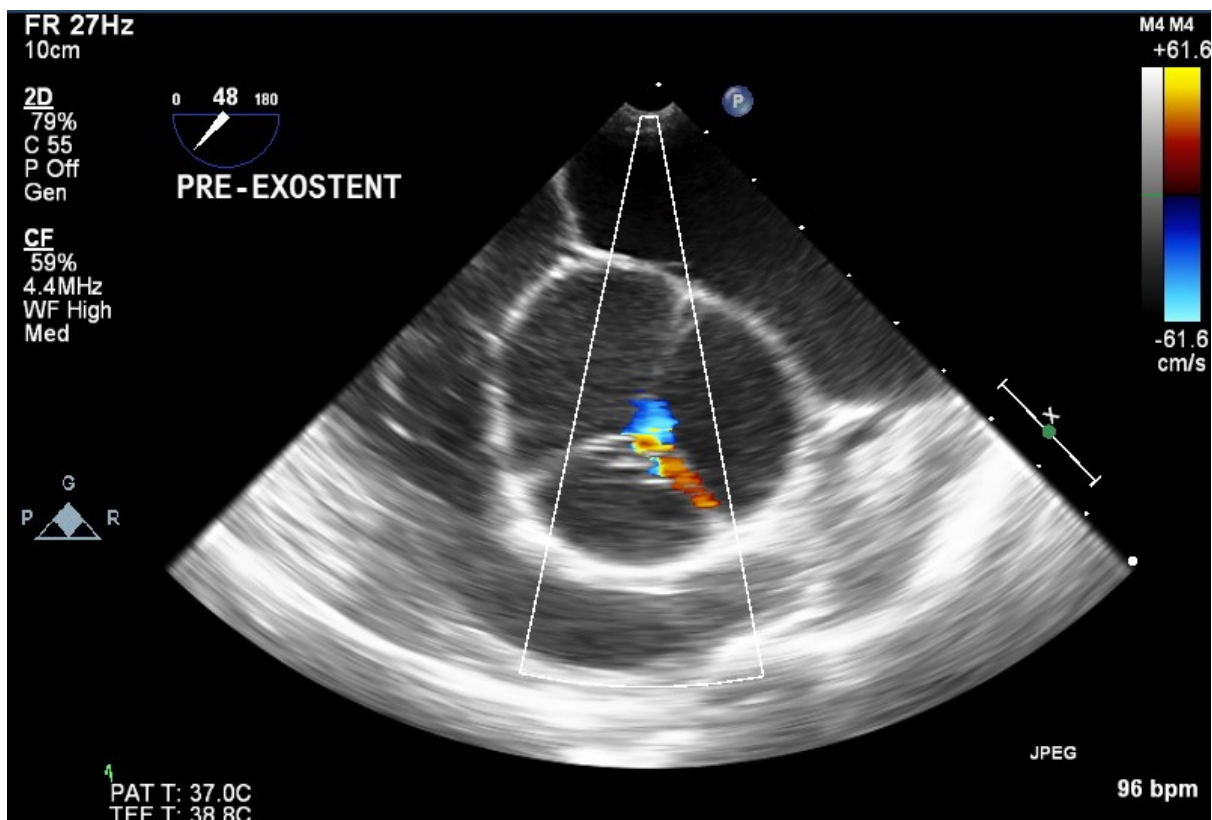
The indications and the management of mitral regurgitation was standard of care for both of these patients with Marfan syndrome. Concomitant root replacement in this scenario is concerning because it exposes patients to an additional operative risk. In addition there are the postoperative consequences 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 minimising the additional operative risk, concomitant PEARS implantation represents an attractive option in this challenging setting.

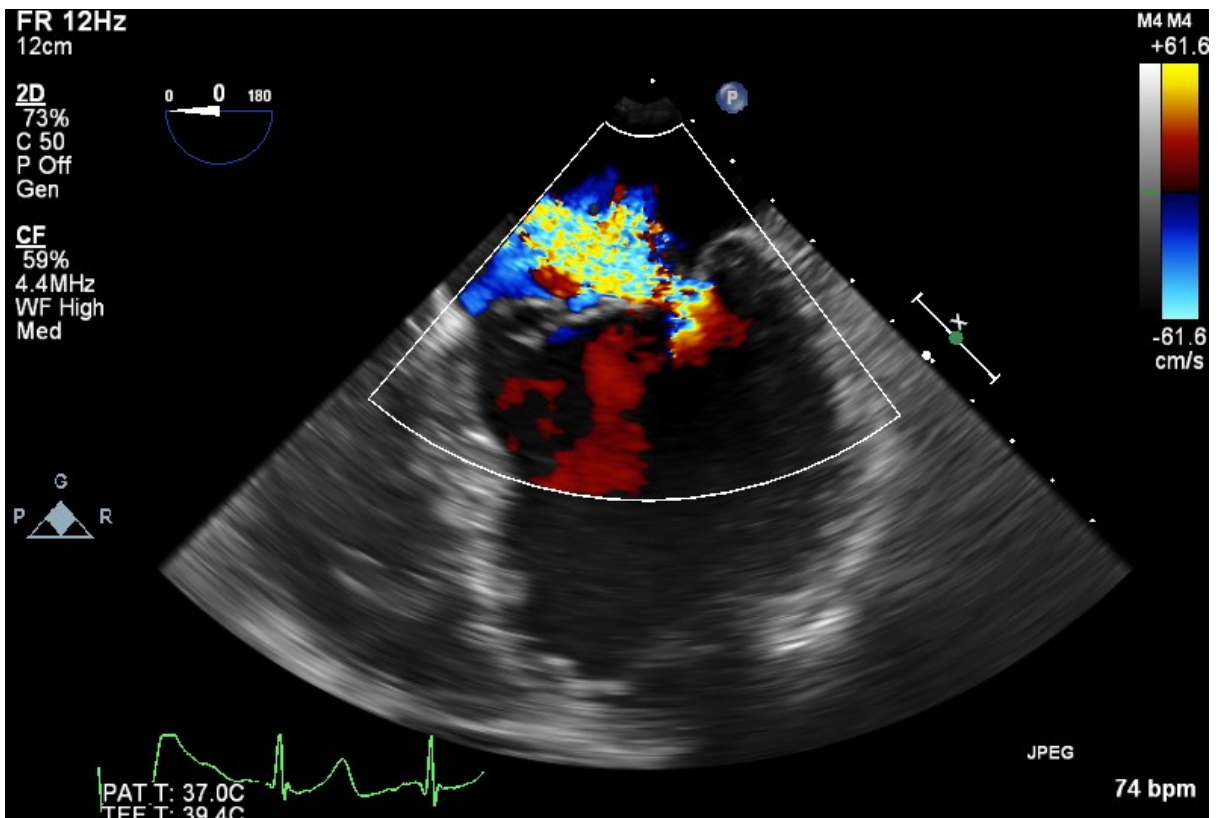
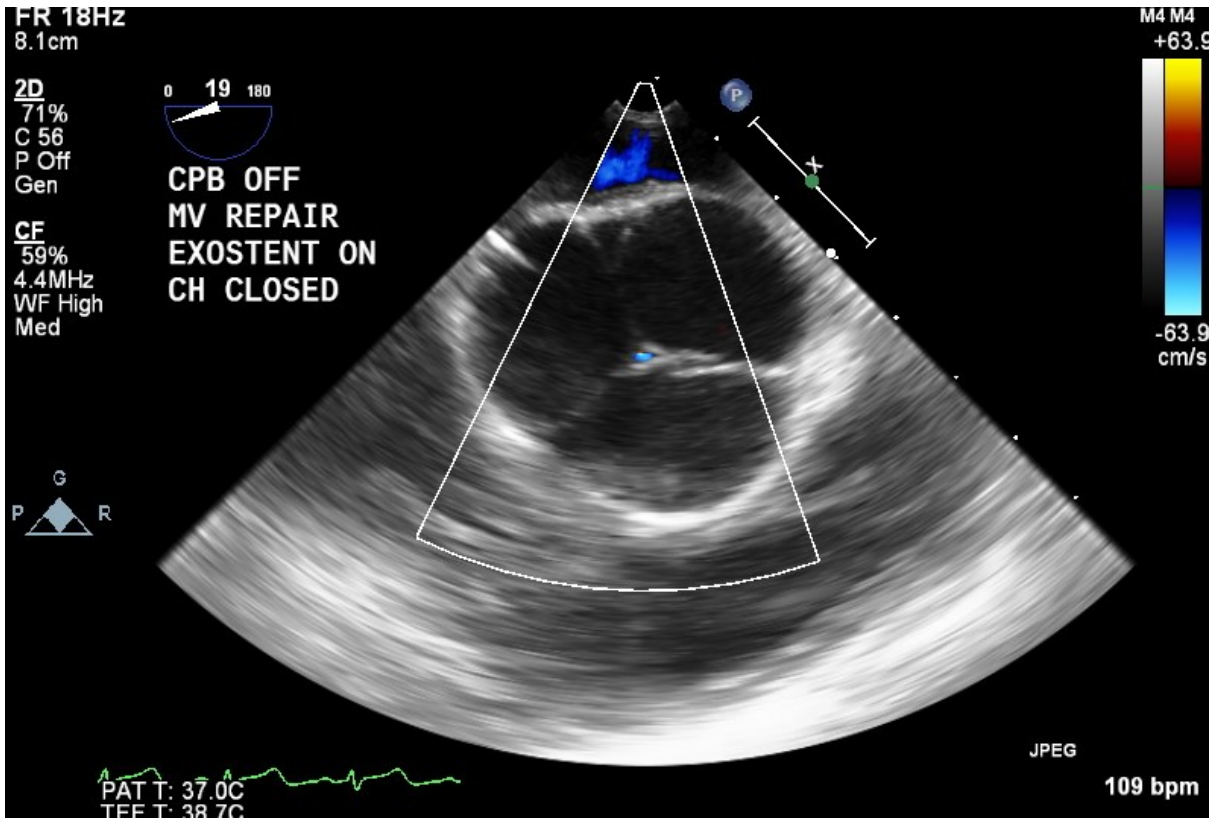
The addition of any surgery requiring cardiopulmonary bypass compromises some of the advantages of PEARS. The desired no-touch approach to the ascending the aorta is sacrificed if the aorta is cannulated, cross clamped, and cardioplegia is administered through the aortic root as in both these cases. 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 of 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 may 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 and while PEARS ‘personalises’ the aortic root support, it behoves the aortic surgeon to ‘personalise’ 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 centres[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 not immediately life-threatening by current convention, but likely to represent a hazard in the not too distant future.





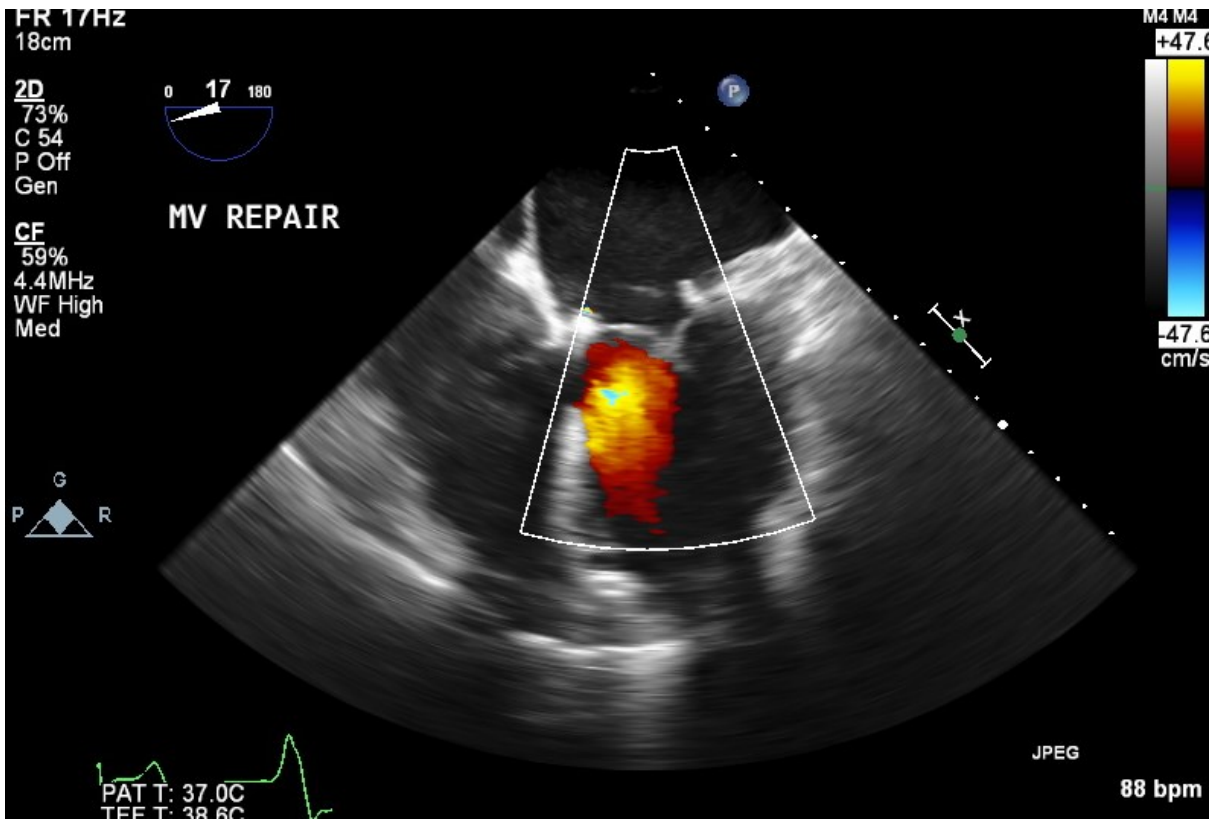


Figure Legends

Figure 1 The essential features of personalised external aortic root support (PEARS).

A. Preoperative MRI. B. The mesh support on its former. C. Depiction of mesh in position. D. MRI in 2014 ten years after PEARS placement.

Figure 2. Histology of the mesh/aorta composite. These histological preparations are from the only patient to have died with a mesh in place, more than four years after operation of presumed arrhythmia without dissection and with a competent aortic valve.[3] A.The mesh fibres (yellow arrow) are completely incorporated with collagen fibres 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 histological appearances in the proximal aorta within the mesh support to be normal with apparent healing in the supported segment.

Figure 3. Trans-oesophageal echo (TOE) images of mid oesophageal four chamber views (A) before and (B) after MV repair in case 2. Doppler colour flow mapping demonstrated severe MR with jet directed anteriorly (A). Mitral regurgitation was fully corrected (B) TOE images of the aortic valve in short axis views (C) before and (D) after PEARS implantation in case 1. The Doppler colour flow mapping demonstrated mild central AR (C). AR was almost completely abolished after PEARS implantation (D).

Reference List

- 1 Treasure T, Petrou M, Rosendahl U, Austin C, Rega F, Pirk J, Pepper J: Personalised external aortic root support: a review of the current status. *European Journal of Cardio-Thoracic Surgery* 2016. DOI 10.1093/ejcts/ezw078
- 2 Plonek T, Dumanski A, Obremaska M, Kustrzycki W: First beating-heart valve-sparing aortic root repair: a "corset" technique. *Ann Thorac Surg* 2015;99:1464-1466.
- 3 Pepper J, Goddard M, Mohiaddin R, Treasure T: Histology of a Marfan aorta 4.5 years after personalized external aortic root support. *Eur J Cardiothorac Surg* 2015;48:502-505.
- 4 Pepper J, Petrou M, Rega F, Rosendahl U, Golesworthy T, Treasure T: Implantation of an individually computer-designed and manufactured external support for the Marfan aortic root. <http://mmcts.oxfordjournals.org/content/2013/mmt004.full.pdf+html>.
- 5 Benedetto U, Melina G, Takkenberg JJ, Roscitano A, Angeloni E, Sinatra R: Surgical management of aortic root disease in Marfan syndrome: a systematic review and meta-analysis. *Heart* 2011;97:955-958.