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External Aortic Root Support to Prevent Aortic Dilatation in Patients With Marfan Syndrome

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ABSTRACT

BACKGROUND Personalized external aortic root support (PEARS) was introduced in 2004 for prevention of aortic root dilatation in Marfan patients. The individual's aortic root is replicated by 3-dimensional printing. A polymer mesh sleeve is manufactured, which is implanted with the aim to support and stabilize the aortic wall.

OBJECTIVES The aim of this study was to assess effectiveness of PEARS for prevention of aortic root dilatation in Marfan patients.

METHODS A total of 24 consecutive Marfan patients operated 2004 to 2012 were prospectively monitored with magnetic resonance imaging. Following a pre-defined protocol, baseline and follow-up aorta measurements were made in a blinded random sequence.

RESULTS The mean age of the patients was 33 ± 13.3 years (range: 16 to 58 years), and the mean aortic root diameter was 45 ± 2.8 mm (range: 41 to 52 mm). Follow-up was 6.3 ± 2.6 years. There was no increase in the aortic root and ascending aorta diameters, but there was a tendency toward reduction: annulus diameter 28.9 ± 2.3 mm to 28.5 ± 2.4 mm (change −0.39 mm, 95% confidence interval [CI]: −1.05 to 0.27 mm), sinus of Valsalva diameter 44.9 ± 2.9 mm to 44.5 ± 3.0 mm (change −0.37 mm, 95% CI: −1.23 to 0.51 mm), and ascending aorta diameter 32.4 ± 3.6 mm to 32.3 ± 3.7 mm (change −0.10 mm, 95% CI: −0.92 to 0.74 mm). In the same period, the descending aorta diameter increased from 22.9 ± 2.4 mm to 24.2 ± 3.0 mm (change 1.32 mm, 95% CI: 0.70 to 1.94 mm; $p < 0.001$) with a tendency toward increase in aortic arch diameter 24.1 ± 2.0 mm to 24.5 ± 2.8 mm (change 0.41 mm, 95% CI: −0.56 to 1.37 mm).

CONCLUSIONS PEARS is effective in stabilizing the aortic root and preventing its dilatation. It is a viable alternative for prevention of aortic root dissection in Marfan patients. (J Am Coll Cardiol 2018;72:1095–105)

Dissection and rupture of the ascending aorta is the main cause of mortality in Marfan syndrome patients (1). The risk of dissection progressively increases with increasing aortic root size (1,2). Current approaches for prevention of aortic dissection in Marfan patients have been focused on prevention of aortic root dilatation with drugs and prophylactic surgery to replace the aortic root (1,2). The drug therapy with beta-blockers and angiotensin receptor blockers is aimed at slowing the rate of aortic dilatation. Although the drugs might slow the rate of dilatation, the aorta still dilates, and Marfan patients eventually undergo root replacement when a guideline-directed diameter threshold is reached (1).

Prophylactic replacement of the aortic root aims at replacing the vulnerable aortic root prone to dissection with a prosthetic graft. The techniques include replacement of the aortic root and the aortic valve with a composite valve conduit (Bentall operation)

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ABBREVIATIONS AND ACRONYMS

CMR = cardiovascular magnetic resonance

CT = computed tomography

PEARS = personalized external aortic root support

VSRR = valve-sparing root replacement

and the valve-sparing root replacement (VSRR) surgery (3). Bentall operation is now a straightforward technique with a low operative mortality; however, it involves replacement of the aortic valve, which is competent in most of the patients. This introduces mandatory lifelong anticoagulation if a mechanical valve is used or risk of more frequent reoperation if a bioprosthetic valve is used. The cumulative lifetime risk of prosthetic valve complications is substantial for younger patients having these operations. The VSRR techniques allow replacement of only the root with preservation of the native aortic valve. Therefore, the risk of thromboembolism and endocarditis are reduced; however, there are other concerns with VSRR. These techniques are technically more challenging with less standardization among centers. Apart from intraoperative challenges, there is a risk of significant aortic regurgitation and reoperation with VSRR (4). A recent multicenter study involving patients from centers with substantial expertise reported a 7% rate of significant aortic regurgitation at 1-year follow-up after VSRR in Marfan patients, suggesting that real-life concerns about durability of these procedures are valid (5). Moreover, sparing of the aortic valve may not always be possible, and there is probability of intraoperative conversion to aortic valve replacement.

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Personalized external aortic root support (PEARS) surgery has been developed as an alternative surgical method to prevent dilatation of the aortic root in Marfan patients (6,7). It involves surgical implantation of an individualized mesh support around the aortic root and the ascending aorta. With 3-dimensional printing utilizing the imaging data, a plastic model of the aortic root unique to each patient is produced. This plastic model is then used as a form upon which the mesh is produced, which fits perfectly to each individual patient's aortic root shape. It is essential that this is placed proximal to the coronary arteries and reaches and is secured to the aortoventricular junction. Cardiopulmonary bypass is typically not needed during the surgery (8). The native aortic valve and the blood endothelium interface are preserved, and therefore, the prosthetic valve-related complications of the Bentall procedure and technical challenges and the risk of aortic regurgitation of VSRR are ameliorated. These advantages would allow operating early in the natural history of the disease when the aortic root diameter lies between 40 and 50 mm. This would alleviate the significant anxiety that Marfan patients are facing during the long duration of watchful monitoring until their aorta reaches the current size threshold recommended in the guidelines for aortic root replacement. The critical question is whether PEARS can be reproducibly implanted to restrain all the proximal aorta segments and is really effective in preventing aortic root dilatation with the ultimate target of eliminating the risk of dissection.

We have previously shown perioperative and procedural advantages of PEARS compared with root replacement (8) and the favorable clinical outcome of the patients during follow-up (9). We also showed in the preliminary reports of the technique that it keeps the aortic root size stable (10). The target in PEARS as a prophylactic operation is to stabilize the aortic root and prevent its dilatation, because the size of the aortic root is currently regarded as the main factor determining the risk of dissection. The aim of the present study is to assess the medium term effectiveness of the PEARS on prevention of aortic root dilatation.

METHODS

PATIENTS AND THE PEARS SURGERY. Following the conceptual and technical development period that was reported previously (6,7), PEARS surgery was first performed in 2004 following approval by the Royal Brompton Hospital Research and Ethics Committee (6). To date, >100 patients with Marfan syndrome have undergone this surgery, which is now offered by several other centers across Europe where the PEARS mesh support is commercially available (ExoVasc Personalized External Aortic Root Support, Exstent Limited, Tewkesbury, United Kingdom). The present study involves prospective data from the series of the first 27 consecutive Marfan patients who had PEARS operation for prevention of proximal aorta dilatation and dissection between May 2004 and July 2012 during the evaluation phase of this new surgical technique at the Royal Brompton Hospital. These patients had close follow-up of their aorta size to monitor the effectiveness of PEARS.

All patients were diagnosed with Marfan syndrome according to Ghent criteria. They were recruited from the aortopathy clinic of the hospital, which has a well-established aortic surgery program, with both the Bentall and the VSRR surgeries routinely being performed. Eligibility criteria for PEARS were an aortic root size of 40 to 55 mm and no or only mild aortic regurgitation (8). PEARS was developed as a prophylactic surgery to prevent dilatation of the aortic root at an early point in the natural history of