

EDITORIAL COMMENT

# Aortic Root Support in Marfan Patients

## Time for a Closer Look?\*



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**A**ortic and cardiovascular complications are an important cause of mortality in patients with Marfan syndrome, and cardiovascular surgery is an important reason why life expectancy for Marfan patients has increased over time (1). The most commonly performed cardiovascular procedure in Marfan patients is replacement of the proximal aortic root and ascending aorta, that is, the Bentall procedure (2). Although the early and long-term results of the Bentall procedure are good, patients must live with the lifetime consequences of a prosthetic aortic valve.

Valve-sparing root replacement (VSRR) surgery is an attractive option for patients with aortic root aneurysms. First described by David and Feindel (3), the aortic valve reimplantation (i.e., David) procedure is the most commonly performed VSRR operation. The David operation has become the surgical treatment of choice in many reference centers for patients with Marfan syndrome and other connective tissue disorders (4,5). Such operations prevent future complications of aortic root aneurysms (i.e., dissection, rupture, or sudden death), while avoiding known complications of aortic valve prostheses. However, the David operation is technically more complicated than the Bentall procedure and may be associated with increased rates of recurrent aortic regurgitation (6). Technical challenges associated with the David procedure are a concern, particularly since the median number of aortic root operations performed annually in cardiac surgery centers within the United States is only 2 (7).

In this issue of the *Journal*, Izgi et al. (8) have described a novel surgical technique—personalized external aortic root support, or PEARS—for prophylactic treatment of aortic root aneurysms in Marfan patients. The procedure is intended to be a relatively simple method of stabilizing the aortic root, leading to decreased risk of catastrophic aortic complications or progression of aneurysm growth and subsequent aortic regurgitation. The PEARS procedure involves wrapping a polymer resin support mesh—whose dimensions are determined by pre-operative cardiac magnetic resonance imaging (CMR)—around the aortic root and ascending aorta. The operation is performed via a full sternotomy, but without cardiopulmonary bypass (CPB) or cardioplegic arrest. Interestingly, the polymer resin support mesh was developed by an engineer with Marfan syndrome, who was also the first patient to undergo the PEARS procedure (9).

SEE PAGE 1095

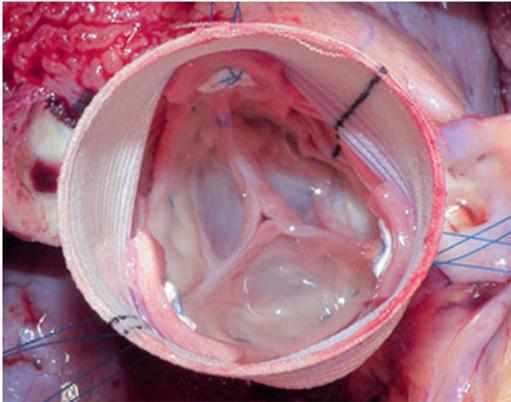
The current study is a retrospective review of the first 27 patients who underwent the PEARS procedure at the Royal Brompton Hospital from 2004 to 2012. There were no perioperative deaths. Follow-up CMR imaging was not available in 3 patients, and data from 2 other patients were excluded because of technical issues at the time of PEARS mesh implantation. During a follow-up period of over 6 years, there were no aortic complications, no significant dilation of any segment of the aortic root or ascending aorta, and no progression of aortic regurgitation. By contrast, the diameter of the unsupported descending aorta increased significantly over time. On the basis of these encouraging data, the PEARS procedure is now being performed in several cardiac surgery centers with >100 implants worldwide. However, the procedure has not yet gained wide acceptance within the cardiac surgery community.

The idea of external ascending aortic support is not new. Robicsek and Thubrikar (10) described external

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**FIGURE 1** Positioning of the Native Aortic Valve Within a Tubular Dacron Graft During the David Operation



Reproduced with permission from Masri et al. (15).

wrapping of the thoracic aorta with a Dacron graft in 1994, with acceptable early and mid-term results. However, the procedure was mostly limited to those patients with dilation of the supracoronary ascending aorta, because of anatomic challenges of wrapping the aortic root. Although some cardiac surgery centers continued to perform aortic wrapping (11), the majority opted for aortic replacement procedures in patients with thoracic aortic aneurysmal disease (2). Izgi et al. (8) have leveraged advancements made in CMR imaging and 3-dimensional printing technology in order to develop a personalized inert polymer resin mesh. With these technological developments and some creative thinking, the investigators have revived external aortic support as a potentially viable procedure for patients with aortic root aneurysms.

One possible criticism of the PEARS procedure is whether it has the ability to fully stabilize the aortic annulus. The aortic annulus is a complex 3-dimensional structure whose inferolateral border is surrounded by right ventricular outflow tract tissue. It is difficult to access this part of the annulus without CPB and cardioplegic arrest, which are not used

during the PEARS procedure. By contrast, the aortic annulus is completely encased within a tubular graft during the David operation (Figure 1). This prevents future dilation of the aortic annulus and subsequent aortic regurgitation, which is one of the advantages that this procedure has over another commonly performed VSRR: the aortic remodeling or Yacoub procedure (12). Izgi et al. (8) demonstrated that the aortic annulus did not dilate during follow-up in patients undergoing the PEARS procedure. Such a finding is encouraging, but also requires confirmation from other investigators.

The current investigators performed the PEARS procedure in Ghent criteria-confirmed Marfan patients with aortic root aneurysms ranging from 40 to 55 mm, with a mean diameter of just under 45 mm. Although current aortic guidelines identify a threshold of 50 mm for prophylactic aortic repair in Marfan patients (13), previous guidelines recommended surgical intervention starting at 40 mm (14). Operating on Marfan patients with smaller aortic root aneurysms may be desirable because the degree of aortic regurgitation increases as the aortic annulus and sinotubular junction dilate. If larger studies confirm that the PEARS procedure is safe, an argument could be made for a lower threshold for surgery in Marfan patients, particularly because the small, but definite, risks of CPB and cardioplegic arrest are avoided.

Although the data from Izgi et al. (8) are encouraging, it is important to remember that it is single-center and noncontrolled. One can nonetheless argue that the safety and possibly efficacy of the PEARS procedure have been demonstrated, and that the time is nigh for a closer look at this provocative operation. More data from other centers with an appropriate control group—or possibly a randomized clinical trial comparing the PEARS procedure to more established methods of prophylactic aortic root repair—is required.

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- KEY WORDS** aortic aneurysm, aortic wrapping, Marfan