

External aortic root support for the Marfan aorta: anatomically normal coronary orifices imaged seven years after surgery[†]

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Abstract

The occurrence of angina necessitated investigation of a patient seven years after an operation to protect his dilated Marfan aorta. The customized support, manufactured by a process of computer-aided design, had been fitted in May 2004 when the aortic root measured 49 mm. The magnetic resonance imaging appearances of the aorta remained unchanged over a postoperative period of 7 years and he remained completely well until he began to experience exercise-related angina in 2011. Coronary angiography showed the cause of angina to be an atherosclerotic left anterior descending coronary artery stenosis which was successfully stented. Aortography and coronary angiography performed at that time showed widely patent coronary orifices with no sign of impingement of the external support on the smooth lumen of his coronary arteries. The soft pliant nature of the textile from which the support was made, its intimate fit to the aorta and porous nature allowing incorporation into the aortic adventitia were deliberately built into the design. Nevertheless it was affirming to see these features realized on imaging seven years later. The patient is again completely well and angina free. He is one of a consecutive series of 30 patients who have had this device. There have been no device-related events in over 100 patient/years of follow-up, and all the patients remain alive and well.

Keywords: Marfan syndrome • Aortic root • Coronary anatomy

INTRODUCTION

External aortic root support has completed initial evaluation at the Royal Brompton Hospital and has Health Technology Appraisal from the National Institute for Health and Clinical Excellence (NICE) [1]. The support is manufactured on a replica of the patient's aorta, made with computer-aided design (CAD modelling) [2]. The operation is performed without cardiopulmonary bypass and without opening the arterial circulation, clamping the aorta or sewing fragile Marfan tissue [3].

The support is manufactured from thread made of polyethylene terephthalate which is the chemical material from which standard vascular Dacron grafts are made. It shares Dacron's favourable characteristics, confirmed over many years of use, in very many patients: it is chemically inert, biologically well tolerated and strong beyond any possible aortic pressure. However, unlike familiar vascular grafts, the textile (or cloth) is a soft, pliant mesh with pores of 0.7 mm, very different from a familiar vascular graft. It protects the whole ascending aorta and is anchored by sutures to the aortoventricular junction. At operation, the mesh is incised in an asterisk fashioned to allow exit of the coronary artery so that no edge is presented to the coronary artery.

[†]This work was done within the Cardiovascular Biological Research Unit (BRU) at Royal Brompton Hospital.

Anxiety has been expressed concerning the hazard to the coronary arteries as they exit the support. We have now had the opportunity to image the coronary arteries in one patient >7 years after surgery.

CASE REPORT

In 2004, a 47-year old man with Marfan syndrome, inherited from his father, reached an aortic root of 49 mm diameter. Elective aortic root replacement was discussed during the phase of monitoring with interval echocardiography. He chose to have a customized external support. This operation was performed in May 2004. He was subsequently well and physically very active.

During 2011, he began to experience exercise-induced constricting chest pain. On exercise testing, he developed 4-mm ST depression at low threshold. A recent magnetic resonance imaging had shown no change in his ascending aorta (Fig. 1). Coronary angiography revealed that the right and left coronary orifices and proximal arteries were normal (Fig. 2; [Supplementary Videos 1 and 2](#)). A typically atherosclerotic narrowing of this left anterior descending coronary artery was identified and stented. He returned to full exercise capacity in the gym and remains well.

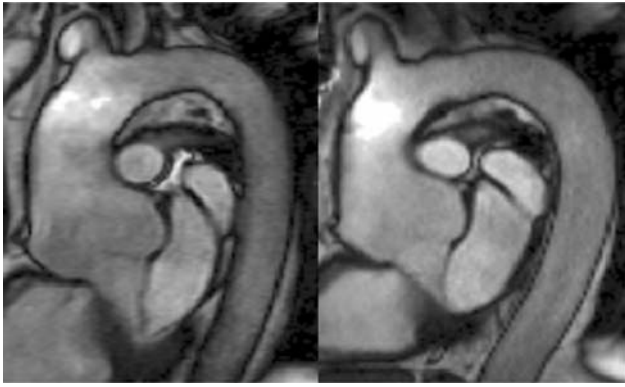


Figure 1: On the left is a pseudosagittal magnetic resonance image of the aorta prior to operation in May 2004; on the right is a similar image in October 2010. The anatomy of the aortic root, believed to be the major predominant determinant of aortic valve competence in Marfan syndrome, is unchanged during this period.

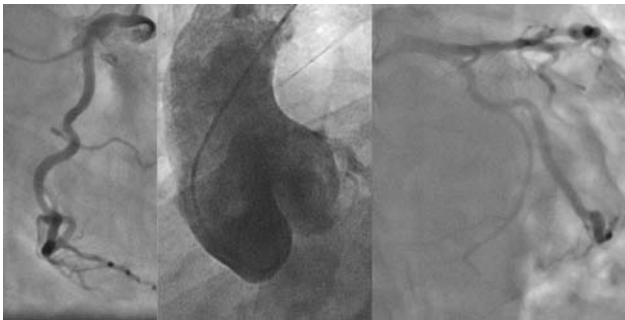
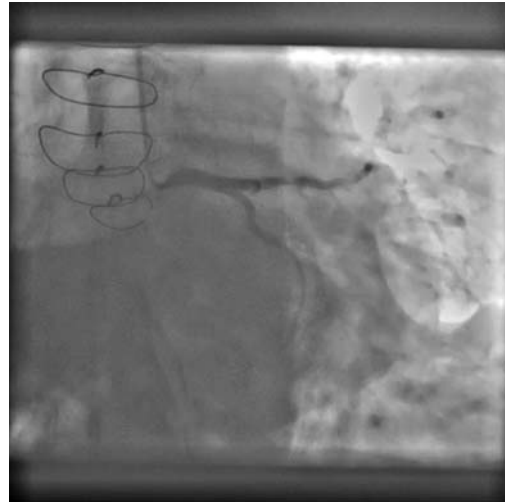


Figure 2: Composite of frames from aortogram and coronary angiograms in 2011. The coronary artery orifices were easily entered and the flow back of contrast indicates that the catheter has not entered beyond a narrowed orifice.



Supplementary Video 1: Right coronary angiogram.



Supplementary Video 2: Left coronary angiogram.

Personalized computer-designed external aortic root support prevents dilatation of the ascending aorta, leaving the blood/endothelial interface totally undisturbed, and conserves perfectly the architecture and function of the aortic valve [1, 2]. There are now over 100 patient/years of event-free follow-up in the first 30 patients receiving a computer-designed external aortic root support since the first operation in May 2004. All operations have been performed with the same device specifications and a standardized surgical procedure [1], mainly in London but recently 4 patients have had this surgery in Leuven, Belgium. Perioperative advantages (avoidance of bypass, myocardial ischaemia and transfusion of blood products) have been confirmed in a carefully conducted comparative analysis [3]. The behaviour of sleeved arteries has been studied in sheep 4–6 months after implantation. Microscopic examination and mechanical stress testing confirm that the mesh becomes incorporated into the adventitia of the artery and is then effectively part of the structure of the vessel wall, increasing its resistance to stress. (Rega, in preparation.) Long-term surveillance continues.

Patients with Marfan syndrome are routinely offered aortic root replacement if they are considered to be at risk of aortic dissection. In the systematic review by Benedetto, the average age at surgery of 1385 such patients was 34.4 years [4]. Based on an expectation of life of perhaps another 35 years, the cumulative risk of a thromboembolic event would be 24.5% (0.7%/year) after total root replacement while those who have valve sparing surgery needed re-interventions at a rate of 1.7%/year which would reach 59.5% by 35 years. Neither complication has been seen in over 100 patient/years follow-up of the aortic root support.

The multiple iterations and variations in valve-sparing surgery, and the admirably frank expositions of the challenging learning curve, suggest that a standardized, reproducible operation may not yet have been attained. In a recently reported series of 101 patients intended for root-sparing surgery, sixteen patients were considered to have repair failure. Of these 3 had intraoperative conversion to Bentall, 3 had reoperations within 3 months and a further 8 patients had reoperations during the follow-up period of 1–10 years [5].

Intraoperative judgements are inevitably prone to error which is minimized by this engineered customized device, made with

DISCUSSION

Follow-up MRI studies have been performed in all patients at regular intervals after surgery [2] and there has been no evidence of migration of the support or impingement on the coronary arteries but this is the only patient in whom we have had clinical indications to perform invasive angiography.

accurate measurements, ahead of time. This exemplifies Tirone David's analogy in his AATS Presidential Address: 'workmanship of certainty' versus 'workmanship of risk'. The customized pre-operative design of the sleeve, the nature of the material, its intimate apposition to the arterial adventitia and its incorporation into the vessel wall, all distinguish this surgery from what is generally envisaged when 'wrapping' aneurysms is considered. The smooth contours and wide-open lumens of the imaged coronary orifices are supportive evidence, albeit in a single patient, for the design of the external support.

With a progressive reduction in the threshold for intervention, the 'number needed to treat' to prevent a dissection has risen: an increasing proportion of these patients are not destined to ever dissect [1]. In advising patients we need to consider lifetime comparisons between strategies, based on Veronesi's principle of maximizing benefit while minimizing harm. Long-term follow-up is needed to confirm that controlling the aortic root shape and size will reduce the risk of dissection, compared with ablative surgery. Dissection arising in other aortic segments remains a risk with both strategies. The competing risks of thromboembolism and valve failure may both be less with external support and early perioperative advantages have already been demonstrated [3]. In advising patients, it is the total overall lifetime outcome that must be considered in the decision-making process [4].

SUPPLEMENTARY MATERIAL

Supplementary material is available at [ICVTS](#) online.

Conflict of interest: Tal Golesworthy is a shareholder and director of Exstent Limited which holds the intellectual property rights in the External Aortic Root Support project. TG was the originator of the concept and the first recipient of the device. Development costs have been met by Exstent Ltd who manufacture the custom made devices for each patient. Costs per device are partly recovered from NHS purchasing. No other author has any pecuniary interests or any other conflict of interests.

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