International Journal of Cardiology 197 (2015) 154–160

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Effect of personalized external aortic root support on aortic root motion and distension in Marfan syndrome patients☆

Cemil Izgi [a], Evangelia Nyktari [a], Francisco Alpendurada [a], Annina Studer Bruengger [a], John Pepper [b,c], Tom Treasure [d], Raad Mohiaddin [a,c,e,*]

[a] Cardiovascular Magnetic Resonance Unit, Royal Brompton Hospital, London, UK
[b] Department of Cardiac Surgery, Royal Brompton Hospital, London, UK
[c] NIHR Cardiovascular Biomedical Research Unit, Royal Brompton Hospital, London, UK
[d] Clinical Operational Research Unit, University College London, London, UK
[e] National Heart and Lung Institute, Imperial College London, London, UK

ARTICLE INFO

Article history:
Received 28 February 2015
Received in revised form 27 May 2015
Accepted 12 June 2015
Available online 14 June 2015

Keywords:
Marfan syndrome
Aorta
Aneurysm
Prevention
Elasticity
Magnetic resonance imaging

ABSTRACT

Objective: Personalized external aortic root support (PEARS) is a novel surgical approach with the aim of stabilizing the aortic root size and decreasing risk of dissection in Marfan syndrome patients. A bespoke polymer mesh tailored to each patient's individual aorta shape is produced by modeling and then surgically implanted. The aim of this study is to assess the mechanical effects of PEARS on the aortic root systolic downward motion (an important determinant of aortic wall stress), aortic root distension and on the left ventricle (LV).
Methods/results: A cohort of 27 Marfan patients had a prophylactic PEARS surgery between 2004 and 2012 with 24 having preoperative and follow-up cardiovascular magnetic resonance imaging studies. Systolic downward aortic root motion, aortic root distension, LV volumes/mass and mitral annular systolic excursion before the operation and in the latest follow-up were measured randomly and blinded. After a median follow-up of 50.5 (IQR 25.5–72) months following implantation of PEARS, systolic downward motion of aortic root was significantly decreased (12.6 ± 3.6 mm pre-operation vs 7.9 ± 2.9 mm latest follow-up, $p < 0.00001$). There was a tendency for a decrease in systolic aortic root distension but this was not significant (median 4.5% vs 2%, $p = 0.35$). There was no significant change in LV volumes, ejection fraction, mass and mitral annular systolic excursion in follow-up.
Conclusions: PEARS surgery decreases systolic downward aortic root motion which is an important determinant of longitudinal aortic wall stress. Aortic wall distension and Windkessel function are not significantly impaired in the follow-up after implantation of the mesh which is also supported by the lack of deterioration of LV volumes or mass.

1. Introduction

Acute aortic dissection is the most feared complication of Marfan syndrome [1,2]. Increased aortic wall stress is a major predisposing factor for dissection. According to the Laplace law, diameter of the aorta, aortic wall thickness and the luminal pressure are major determinants of aortic wall stress. These factors fully explain aortic wall stress in a circumferential direction [3,4]. Besides the circumferential wall stress, longitudinal wall stress, in the long axis of aorta, has also been proposed as a risk factor for dissection [4,5]. Indeed, longitudinal aortic wall stress has been shown to concentrate just above the sinotubular junction where most of the dissections occur as a transverse tear [5]. Other than the factors included in Laplace law, the systolic axial downward displacement of the aortic root also affects the longitudinal wall stress [6]. Marfan syndrome patients are rarely hypertensive and therefore high blood pressure does not seem to be a common predisposing factor for either circumferential or longitudinal increased wall stress [7]. This leaves the aortic root diameter, aortic wall thickness and systolic axial displacement of the aortic root as important factors associated with increased wall tension and risk of dissection in Marfan patients.

Personalized external aortic root support (PEARS) surgery is a novel surgical method for prevention of aortic root dilatation and dissection in Marfan patients [8]. A replica of the patient's aortic root and ascending aorta is constructed from the imaging data by computer modeling and then a bespoke porous medical grade fabric mesh sleeve is manufactured. The personalized external support fits intimately to patient's aorta shape, thanks to the exact modeling of the aorta by a computer aided design (Fig. 1). This mesh is then surgically implanted around the aorta extending from the annulus to the proximal aortic arch [9]. The aim of this procedure is to reinforce the aortic wall for prevention of dilatation. The

☆ All the authors take responsibility for all aspects of the reliability and freedom from bias of the data presented and their discussed interpretation.

* Corresponding author at: Cardiovascular Magnetic Resonance Unit, Royal Brompton Hospital, National Heart and Lung Institute, Imperial College, SW3 6NP London, UK.
E-mail address: R.Mohiaddin@rbht.nhs.uk (R. Mohiaddin).

http://dx.doi.org/10.1016/j.ijcard.2015.06.015
0167-5273/

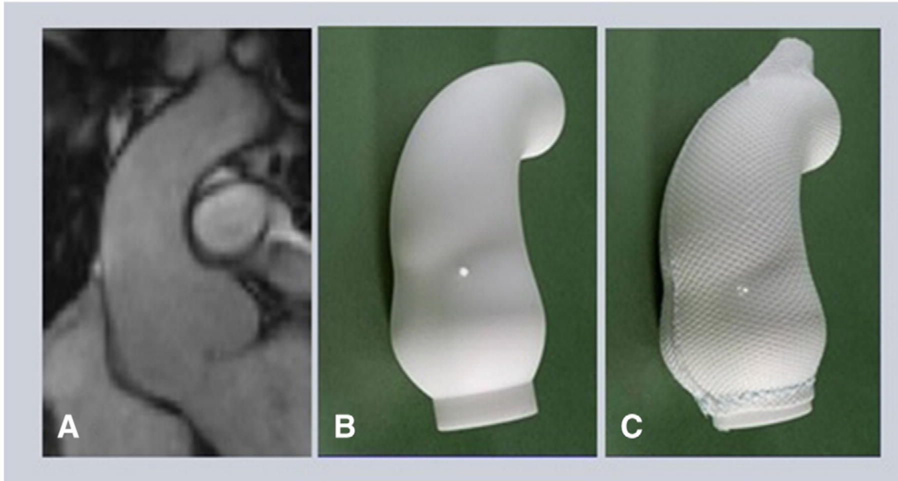

Fig. 1. Imaging data retrieved from the CMR study (A) is used to produce an exact model of each patient's aorta by using computer aided design and rapid prototyping (B) and then a bespoke porous, soft and pliable mesh is produced that is ready for implantation as seen mounted over the aorta model (C).

mesh is incorporated into the aortic wall and contributes to its thickness [10]. We have shown previously on follow-up studies that it holds the aortic shape and size stable, preventing any further increase in aortic dimensions [11,12]. Being incorporated in the aortic wall and extending from the aortic annulus up to the aortic arch, it may also stabilize the aorta in longitudinal direction and provide an additional benefit by limiting the axial downward motion of the aortic root in each systole.

The bespoke mesh sleeve used in PEARS surgery and the tubular Dacron grafts used in Bentall and valve sparing root replacement operations are made from the same basic polymer. However in contrast to the woven inelastic and stiff tubular grafts [13], the mesh used in PEARS is produced as a macroporous, soft and pliable fabric. It is well proven that decreased elasticity of the aortic wall is associated with increased afterload for the left ventricle with possible consequences of decrease in left ventricular ejection fraction, increase in left ventricular mass and possibly increased rates and severity of aortic regurgitation [14,15]. It is unknown whether the PEARS procedure is associated with any of these unwanted consequences.

In this study we investigated the effect of PEARS surgery on systolic downward motion of the aortic root. We also investigated the long term effects of implantation of the mesh on systolic distension of the aortic root with the possible consequences of impaired distensibility on left ventricular ejection fraction, left ventricular mass and aortic regurgitation.

2. Methods

2.1. Patients

To date 47 patients with Marfan syndrome have had prophylactic PEARS surgery for prevention of thoracic aorta dilatation and dissection. This novel surgical technique was developed and first implemented in the Royal Brompton Hospital under strict control of research ethics committee [2] and 27 patients had the operation at the Royal Brompton Hospital between the years 2004 and 2012. Criteria for the operation and clinical characteristics of the patients have been reported previously; briefly a suitable candidate for this preventive surgery would have an aortic root diameter of 40–45 mm and no or little aortic regurgitation [8]. In two of the patients the main imaging modality was computed tomography and in one patient clinical imaging was performed abroad before the patient had PEARS surgery in our center. The remaining 24 patients had cardiovascular magnetic resonance (CMR) examinations in our center before the operation and were followed up by CMR examinations after the operation at six months, 12 months and wherever possible annually thereafter. These patients formed the study group. The study was registered and approved as a clinical audit to assess the effectiveness of PEARS surgery by the Quality and Safety Department of the Royal Brompton Hospital.

2.2. CMR examinations and analysis of LV ejection fraction and mass

All CMR examinations were performed in a 1.5 T scanner (Siemens Avanto, Erlangen, Germany). Briefly, the basic CMR protocol performed in all the patients included localizers, multislice black-blood and bright-blood anatomic images, ventricular long axis and short axis cine images, short axis stack of aortic valve and root cine images [16]. All cine images were acquired with the balanced steady state free precession sequence with retrospective ECG gating to capture full cardiac cycle and at end-expiration.

Left ventricular volumes, ejection fraction and mass were calculated with dedicated software with exclusion of the papillary muscles from blood pool (CMRTools, Cardiovascular Imaging solutions, London, UK). The high reproducibility for these parameters with the software used in this study has been previously reported from our center [17]. Lateral mitral annular plane systolic excursion (MAPSE) was measured from the four chamber cine images by subtracting the distance from a reference point outside of the left ventricular apex to the lateral mitral annulus in end-diastole and end-systole. The measurements were performed blinded to patients' details. Left ventricular mass index was derived by dividing left ventricular mass by body surface area.

2.3. Assessment of aortic root motion, distension and aortic regurgitation

All the preoperative and the latest follow-up CMR study sessions were saved and anonymized in random order for analysis and comparison of aortic root motion, distension and aortic regurgitation.

Aortic root motion was defined as systolic downward motion of the annular plane as previously described [6]. For this measurement the left ventricular outflow tract cross-cut (LVOTxc) CMR cine images were used. This plane fully shows the annulus and the ascending aorta throughout systole and diastole and the systolic downward motion of the aortic root can be fully assessed. The plane of the aortic annulus was drawn at diastole and systole. In most cases these two planes were not parallel to each other due to the three dimensional motion of the aortic root. Therefore, for consistency we took the systolic downward motion of the aortic root as the length of a line drawn perpendicular to the mid-point of diastolic annulus plane to its intersection with the systolic annulus plane (Fig. 2).