A 28-year-old male with transposition of the great vessels underwent a Mustard operation in infancy. Revision was required for inlet venous pathway obstruction at 3 years of age at which stage a delayed arterial switch operation was performed.

Cardiological surveillance revealed a dilating aortic root. He is a very keen competitive sportsman. Increasing serial size measurements were demonstrated on cardiac Magnetic Resonance Imaging (MRI) scan (Fig. 1) with the root measuring 3.7 cm and the sinuses of Valsalva being 5.0 cm with preserved aortic valve function.

At operation the adhesions were difficult but median sternotomy was performed uneventfully. The great vessels were side by side and the pulmonary artery was lying to the right. To greatly aid visualisation of the right side of the proximal aorta, the main pulmonary artery was transected at its previous transection anastomosis after instigation of cardiopulmonary bypass. Complete mobilisation of the coronaries and aortic root was performed to the ventricular arterial junction. Mobilisation was further complicated as the circum aortic root was performed to the ventricular arterial junction.

The circumflex artery arose from the right coronary and passed in a retro-pulmonary position being 5.0 cm with preserved aortic valve function.

Increasing serial size measurements were demonstrated on cardiac Magnetic Resonance Imaging (MRI) scan following surgery. An 11-year boy who had been diagnosed in the post-natal period with a double outlet right ventricle, VSD and subpulmonary stenosis. A traversing anterior coronary artery precluded a proposed Rastelli operation at 10 months of age. He was palliated at that operation with an open atrial septectomy and right modified Blalock–Taussig shunt. He then had an arterial switch operation at 4 years of age.

Cardiological follow-up demonstrated an enlarging aortic root. He then developed chest pain with exertion and a MRI scan demonstrated that the left anterior descending coronary was stretched by marked dilatation of the anterior sinus of Valsalva from which it arose (Figs. 3 and 4). The circumflex coronary artery arose from the right coronary artery. There was mitral regurgitation consequent to a cleft in the mitral valve.

A PEARs external aortic root support was postulated after excision and reduction plasty of the dilated anterior sinus of Valsalva. The PEARs prosthesis was computer generated from computer graphics of the expected aortic shape after reduction aortoplasty. The surgeon directed reduction plasty of the dilated anterior sinus of Valsalva. The PEARs prosthesis was computer generated from computer graphics of the expected aortic shape after reduction aortoplasty. The surgeon directed the prosthesis exactly over the site of the excised coronary buttons and the coronaries were re-implanted over the PEARs prosthesis to the same location as before with 4/0 Prolene. The heart was de-aired and the aortic cross clamp was released. The longitudinal seam was then re-approximated with interrupted 4/0 Ethibond up to the level of the brachiocephalic artery and beyond the aortic cannulation site after cessation of cardiopulmonary bypass and decannulation.

The cross clamp time was 52 min and the bypass time 140 min. A trans-oesophageal echo was performed which measured a slight reduction of the aortic root diameter, un-impinged coronary blood flow and no aortic valve insufficiency. The patient made an uneventful recovery following surgery.

Annual serial MRI over three years scans have confirmed no change in the aortic root dimensions.
The circumflex artery arose from the right coronary artery posteriorly. Under cross clamping the aorta was transected at the previous suture line, the dilated anterior sinus was reduced in size by excision. The coronaries were fully mobilised and the sinuses dissected out to the ventricular arterial junction. A PEARS calculated at 90% of the original aortic dimension was selected and after 4/0 Prolene reconstruction of the reduction aortoplasty it was applied to the entire aorta beneath the coronaries and secured to the ventricular arterial junction with multiple 4/0 Ethibond sutures. The cleft in the mitral valve was repaired via the aorta. The transected aorta was re-anastomosed and the seam of the PEARS, calculated to lie to the left of the LAD, was closed with multiple 4/0 Ethibond. The clamp was removed after de-airing. With the reduction plasty of the anterior sinus and the application of the prosthesis the left coronary was free and loose over the prosthesis (like a bucket handle) (Fig. 5).

The cardiopulmonary bypass time was 151 min with 112 min of aortic cross clamping (Fig. 6). There was minimal mediastinal blood loss. The patient made an uneventful recovery.

Aortic root dilatation is an increasingly recognised problem associated with the Arterial Switch operation for Transposition of Great Arteries [1,2]. PEARS for Marfan’s aortopathy is a procedure recognised by NICE with 278 patient years of follow-up [3]. Long-term surveillance in 30 patients to 5 years and in one patient to 12 years, has failed to demonstrate any increase in the diameter of the aortic root or ascending aorta. There has been no aortic dissection in the supported aorta and no movement in the external support. There is evidence both experimentally and in one patient that the material of the macroporous polyester sleeve becomes incorporated in the aortic wall [4,5].

The above two cases demonstrate that the PEARS procedure is a versatile suitable external support for complex congenital aortopathies which present late after radical repair in infancy. Cardio-pulmonary bypass is necessary to facilitate coronary mobilisation plus dissection to the ventricular arterial junction to fully support the entire aortic root. Normal blood endothelial interface is maintained. The patient retains their own valve and the judgement involved in valve sparing...
techniques is avoided and as a non-ablative procedure it can be offered to the patient earlier. Furthermore, no bridges are burned as it is quite feasible to return at a later date and replace the aortic valve if required. The first case required the coronary artery ostia to be excised and then re-implanted after application of the prosthesis. Increased experience proved that this was not necessary in the second case.

Clinical MRI monitoring post PEARS procedure over the past 10 years has demonstrated stable aortic size in all Marfan’s patients. We are confident that the same will be observed in MRI follow-up of these patients. Imaginative use of this novel prosthesis may be extended to its use in aortopathies of a non-connective tissue origin.

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

References