Personalised External Aortic Root Support (PEARS) Clinical Results up to Nine Years in Marfan Syndrome

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Data from before the root replacement era indicated that two thirds of people with aortic root manifestations of Marfan syndrome would die of dissection, many of them young. We know that aortic diameter correlates with the risk of aortic dissection (N=113; P<0.005)[1] but size alone is a poor predictor: the there is a large overlap in the size of the aorta in those who have dissected and those who have not. This knowledge, only available once echocardiography was widely available in the 1980s, led to more intense monitoring for change in dimension for fear that preventable fatal dissections would be missed. [2] As surgery became safer, and the need for life-long anticoagulation was avoided by the introduction of valve sparing root replacement (VSRR) the size criteria for intervention was progressively reduced, more so in Europe than in the United States. It should be remembered that for each lowering of the size criterion the specificity is reduced in the process of increasing sensitivity. While the clinical feeling is that every root replaced is a dissection prevented (that is the say the "number needed to treat" is one) the reality is that an increasing number of people with Marfan syndrome, not destined to dissect, are having root replacement surgery.[3]

VSRR is associated with a non-trivial risk of reintervention of 1.3% per annum both in meta-analysis[4] and expert series.[5] This accumulates to a greater than evens chance by the age of 60 years. Personalised external aortic root support (PEARS) offers a non-ablative, measured and engineered approach: the workmanship of certainty.[6] The perioperative burden of care is markedly reduced[7] and case by case follow-up shows the aortic root architecture to be unchanging.[8] Clinical experience by Laks group since 1984 confirms that the macroporous Dacron mesh becomes incorporated in the aortic adventitia[9] as it does the mesh in our animal recovery experiments.[10] The histological appearances make the propagation of dissection within the support unlikely. It presents no obstacle to future surgery and would make it less hazardous.

PEARS has favourable Health Technology Appraisal from the British National Institute for Health and Clinical Excellence (NICE). The surgery has been performed in 33 patients with maximum follow-up approaching nine years (average four) years and 113 patient years of follow-up with no device related events.

The surgeons contributing to this surgical experience are

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Reference List

- 1 Murgatroyd F, Child A, Poloniecki J, DeBelder M, Treasure T, Pumphrey C: Echocardiographic and clinical features are inadequate guides to the management of patients with the Marfan syndrome. British Heart Journal 1993;68:84-85.
- 2 Treasure T, Chow T, Gallivan S: Replacement of the aortic root in Marfan's syndrome. N Engl J Med 1999;341:1473-1474.
- ³ Treasure T, Golesworthy T, Pepper J, Ruiz F, Gallivan S: Prophylactic surgery of the aortic root in Marfan Syndrome: reconsideration of the decision making process in the era of customised external aortic root support. J Vasc Endovasc Surg 2011;18:215-223.
- 4 Benedetto U, Melina G, Takkenberg JJ, Roscitano A, Angeloni E, Sinatra R: Surgical management of aortic root disease in Marfan syndrome: a systematic review and meta-analysis. Heart 2011;97:955-958.
- 5 Shrestha M, Baraki H, Maeding I, Fitzner S, Sarikouch S, Khaladj N, Hagl C, Haverich A: Long-term results after aortic valve-sparing operation (David I). Eur J Cardiothorac Surg 2012;41:56-61.
- 6 Pye D: The Nature and Art of Workmanship. Cambridge University Press, 1968.
- 7 Treasure T, Crowe S, Chan KM, Ranasinghe A, Attia R, Lees B, Utley M, Golesworthy T, Pepper J: A method for early evaluation of a recently introduced technology by deriving a comparative group from existing clinical data: a case study in external support of the Marfan aortic root. BMJ Open 2012;2:e000725.
- 8 Pepper J, Golesworthy T, Utley M, Chan J, Ganeshalingam S, Lamperth M, Mohiaddin R, Treasure T: Manufacturing and placing a bespoke support for the Marfan aortic root: description of the method and technical results and status at one year for the first ten patients. Interact Cardiovasc Thorac Surg 2010;10:360-365.
- 9 Cohen O, Odim J, De la ZD, Ukatu C, Vyas R, Vyas N, Palatnik K, Laks H: Long-term experience of girdling the ascending aorta with Dacron mesh as definitive treatment for aneurysmal dilation. Ann Thorac Surg 2007;83:S780-S784.
- 10 Verbrugghe P: External aortic root support: a histological and mechanical study in sheep. Interact Cardio-Vasc Thorac Surg. In press.