Interventional Procedure Guidance 394 from NICE concerns external aortic root support. Interventions are performed without the need to open the aorta or to use bypass or myocardial ischaemia, the pliable support is placed around the aorta from the aortoventricular junction, extending beyond the brachiocephalic artery. Appropriately sized openings are fashioned to allow for the exit of the coronary arteries. The technical details have been described along with postoperative MRI confirmation of unaltered morphology and function of the aortic valve and blood/endothelial interface. Images of the patient’s own aorta are the source of data for computer aided design. A replica of the aorta is used as a former on which an external support is manufactured prior to surgery (figure 1). At operation, performed without the need to open the aorta or to use bypass or myocardial ischaemia, the pliable support is placed around the aorta from the aortoventricular junction, extending beyond the brachiocephalic artery. Appropriately sized openings are fashioned to allow for the exit of the coronary arteries. The technical details have been described along with postoperative MRI confirmation of unaltered morphology and function of the aortic valve (figure 2).

Where does external aortic root support, and the guidance concerning it, fit into present practice? For people with known or suspected Marfan syndrome, echo imaging of the aortic root is routine. The aortic root in people with the Marfan phenotype has a characteristic morphology. Once that appearance is seen, current advice is that the patient should be monitored with interval echo measurements, with a view eventually to intervening with surgery. The purpose of surgery is to prevent the more common (type A) form of aortic dissection, prevalent in those with Marfan syndrome, in which a transverse intimal tear within the sinuses of Valsalva is associated with propagation of dissection in the aortic media. This has devastating consequences: the aorta may rupture into the pericardium; or the dissection may obstruct blood flow in one or a series of arterial branches resulting in cerebral, spinal cord, renal and/or other visceral ischaemia. Although familiar to cardiac specialists, dissection is easily missed in the emergency situation. Given its devastating consequences, there are compelling reasons to prevent it. Bentall, of the Hammersmith Hospital, is credited with performing the first operation replacing the aortic root and reconnecting the coronary arteries.

Total root replacement was at the outset a high risk operation. During the 1980s, surgery was progressively made safer by refinement of technique and materials, the availability of factory made valved aortic grafts—and practice, practice, practice. The most reliable form of surgery included insertion of a mechanical valve, with subsequent lifelong anticoagulation, starting in the teens or twenties in many cases. Conserving the native aortic valve to avoid these hazards re-set the learning curve, and once more surgery became technically challenging. With more practice in the hands of highly skilled superspecialist surgeons, the perioperative risks have again become low. Re-operation for failure of the native valve is a legacy. The trade-off of the hazards of anticoagulation and bleeding versus tissue valve failure, true of all aortic valve surgery, is a particular concern in patients with Marfan syndrome. Their inherited fibrillar deficiency remains: further life-saving aortic surgery, and surgery on the spine and other parts of the body, may be called for during the hoped-for normal lifetime with the disease.
As preventive surgery has become safer and more confidently recommended, the threshold at which it is offered has been lowered, specifically in terms of the size of the aortic root, defined as the diameter of the aorta at the level of apposition of the valve leaflets. Other factors include family history of dissection, and the rate of change of the aortic diameter. The diameter at which the aortic root should be replaced came down from 6 cm to 5 cm. After formal decision analysis, a new threshold as low as 3.5 cm was proposed, albeit that the likelihood of dissection was based on five individuals’ opinions rather than being derived from data. An unbiased estimate of the ‘unpredictable natural history’ will now not be available due to the lack of contemporary observational data. Nonetheless, clinicians familiar with both the disaster of dissection and the relative safety of contemporary surgery, may broach the subject of root replacement as soon as the characteristic aortic morphology is demonstrated. Even though the natural history data are well over 40 years old, the risks of continued monitoring are sufficiently alarming to bring up the very scary ‘sitting on a time bomb’ analogy.

The guidance states: ‘Further research should report on long-term outcomes, particularly the occurrence of dissection and aortic dilatation, and the need for further procedures.’ This, of course, applies equally to ‘conventional’ procedures in which the rest of the aorta remains at risk, and further procedures are not infrequently required. Bentall’s innovation was regarded as demonstrably worthwhile on the basis of a single case: the signal could be seen from the noise. For many forms of intervention that is a good enough test of efficacy. Bentall’s operation and the subsequent iterations that led to present management were never subjected to the rigours of NICE technology appraisal. As the threshold for intervention has changed, and there is now a new procedure available, any process of evaluation should realistically include a comparison of all contenders.

Is a patient with external support more at risk of future events than those who have undergone other forms of surgery? The only substantive difference is that there remains a theoretical risk of dissection of the aortic tissues within the support. There is a well established consensus that size and change in size are the important variables determining the likelihood of dissection, if the size remains constant, the risk is likely to be reduced. Anticoagulation is avoided with valve sparing surgery, and other hazards are shared to varying degrees by all the available forms of surgery. Given the perfect maintenance of aortic valve morphology (figure 3), which has proved difficult to achieve with valve sparing surgery, external support may be a more durable solution in the long term.

How are the relative merits of these operations best put to the patient? The version of the guidance offered to the patient states: ‘There have been occasional serious safety problems.’ Are we to infer that ‘conventional’ aortic root replacement does not have ‘occasional serious safety problems’? The guidance very reasonably states that ‘further research should report on long-term outcomes’, but long-term results take a long time to accrue. Meanwhile, we should use all the data now available in rigorous comparisons. There is a complex trade-off of relative risks and benefits which calls for more imaginative research, and much sooner than can be provided by traditional surgical follow-up studies.

The timing of the decision to intervene is not addressed in this guidance. As the threshold size for surgery reduces, and milder patients are referred for surgery, it is inevitable that the ‘number needed to treat’ (NNT) to prevent a dissection will increase. Consider carotid endarterectomy to prevent stroke. The NNT is 6 for 70–99% carotid stenosis, and rises to 24 for lesser stenosis of 50–69%. These data are from randomised trials, where like is compared with like, and outcomes are collected fastidiously in both arms. In the absence of such data, there is uncertainty. For Bentall’s classic case, the NNT may have approached unity, but as the size criterion is lowered for preventive root surgery, the benefit attributable to dissections prevented may be exceeded by the harm done to the lives of those who might have lived without ever dissecting. If we expand the number of patients referred for operation to prevent fewer dissections, at what stage is it no longer cost effective? Health economic evaluation is central to the NICE approach, and is applicable in this instance.
It would quantify ‘the potential for more rapid recovery than aortic replacement surgery’, ‘psychological benefits, and improved quality of life’ in comparison with ‘conventional’ approaches.

The aortic root in Marfan phenotype has a characteristic morphology that is readily recognisable. The dilation specifically involves the sinuses of Valsalva. Of particular note is that the distance between the attachment of the valve leaflets and the coronary orifices is increased. This is in contrast to the form of dilation of the proximal aorta seen in bicuspid aortic valve disease, where the aorta, also at risk of dissection, widens out progressively, with loss of the characteristic tubular appearance due to dilation extending beyond the sinutubular junction (figure 4). Future studies might include this group of patients, for whom a lesser intervention to protect the aorta for as long as their bicuspid valve is haemodynamically satisfactory might be an important option.

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Ethics approval Ethics approval was provided by Royal Brompton Hospital.

Contributors All authors are core members of the External Aortic Root Support project team. JP developed the surgical aspects of the procedure and was surgeon on all the patients operated on during evaluation at Royal Brompton Hospital. TG is responsible for the concept and manufacture of the devices. RM provided all the imaging services for the project and the images in this article. TT drafted and edited the paper to which all authors have contributed. RA provided advice with regard to anatomical aspects, and made suggestions concerning grammar and style. TG was the first recipient of the external aortic root device.
Figure 4  The appearance of the aortic root with bicuspid valve disease. These patients are also at risk of dissection. It is likely that the aetiology is related to the eccentric jet and that this represents a form of post-stenotic dilatation. These patients may also be candidates for external aortic root support.

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Data sharing statement  We are happy to share any data referred to.

REFERENCES


