

Aortic root surgery in Marfan syndrome

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Elective root replacement of the ascending aorta in people with Marfan syndrome has been established practice for more than 25 years; it greatly reduces the risk they face from fatal aortic dissection.¹ Now that low perioperative risk is assured and long-term survival is expected, long-term consequences and the durability of the surgery have become important considerations. The question for people with Marfan syndrome, their families and their surgeons is whether to choose the valve-sparing or valve-replacement forms of surgery. This question is at the core of the systematic review and meta-analysis published in this issue of *Heart* (see page 955).² The decision as to which option to prefer is dominated by a trade-off between these competing risks; the analysis presented informs the choice but does not make it any easier. How would an individual patient weigh the risk of stroke versus a given possibility of the need for reoperation? The calculated estimates of the thromboembolic hazards associated with a mechanical valve at 0.7% per year, and the risk of native valve failure mandating reoperation at 1.3% a year, are the central conclusions of this report.

The review incorporates 12 publications from 1995 to 2009, comprising reports of 35–675 patients, with a total of 1434. The average age of the patients at the time of surgery was 34 years. The systematic reviewers could not find data on the degree of aortic regurgitation, or the aortic root dimensions before surgery, which are two of the important criteria used in decision-making¹ and would have informed readers of the severity of disease in patients in whom surgery was undertaken. But the large majority of patients with Marfan syndrome develop aortic root dilatation and we know that if this is allowed to continue unchecked then

about two-thirds of them will eventually present with aortic dissection, which is fatal in 21% of patients in registry data.³ We also know from data acquired before pre-emptive aortic root surgery was established that fewer than a third of individuals at this average age would have been alive 26 years later.⁴ Emergency surgery for dissection is hazardous in the best hands but in view of the urgency of the situation many of these patients are taken to the nearest hospital, not necessarily a cardiac surgical centre, and there may be many more deaths associated with aortic dissection than enter the registry.

The history of root replacement undertaken to modify the risk of aortic dissection has been one of stepwise progress since first reported by Bentall at Hammersmith Hospital in 1967.⁵ The original operation required preclotting of unsealed graft material and incorporation of a Starr–Edwards mechanical valve hand sewn into the tube graft. There were very real anxieties about fatal bleeding such that an inclusion technique was used in early practice: the coronary orifices were sutured *en face* into the graft and the native aorta wrapped around the root replacement. This changed as presealed material became available, followed by factory-made composite grafts during the 1980s. Refinement of surgical technique and precise anastomosis of the coronary arteries with an aortic button obviated the need for an inclusion technique and the operation became standardised and highly reproducible.⁶ Criteria for elective root replacement were established and could be recommended with confidence.¹

While excellent reproducible results are obtained with the ‘modern Bentall’ operation, these young patients are committed to a lifelong risk of valve-related thromboembolism and an accompanying fear of bleeding from the anticoagulation required to minimise that risk. This led Yacoub *et al*⁷ and David *et al*⁸ to pioneer means of conserving the aortic valve. These operations have been through numerous iterations, each seeking to correct the failings of an earlier version. Such experience is often described as being a ‘steep learning curve’, but if the intention of this well-worn

phrase is to describe the trajectory towards reliability, plotted against repetition, such learning curves would be better described as flat, and sometimes faltering. Compared with the modern Bentall operation, valve-sparing surgery is a much more time-consuming operation and relies heavily on skill and repeated practice. Drawing an analogy from the writing of the wood worker and professor of furniture design, David Pye, the modern Bentall operation comes close to “the workmanship of certainty” in which the quality of the result is mechanically predetermined and less in the control of the operator. Valve-sparing surgery reverts to the “workmanship of risk” where any lapse in concentration or judgement may irretrievably spoil the result.⁹ This analogy was used by Tirone David in his presidential address to the American Association for Thoracic Surgery¹⁰ and it is a concern for many excellent surgeons who recognise that for the patient the certainty of the result is more important than the surgeon’s creative flair.

In the review, the valve failure rate among the 413 patients having valve-sparing surgery was 1.3% a year which, within the limits of the meta-analysis, appeared to be a rate not dependent on follow-up duration. This means that by 20 years more than a quarter of patients who have had valve-sparing surgery might need further aortic valve surgery and at best only half of them are likely to complete their lifespan without another aortic root operation.

The competing haematological risks faced by those with the more certain mechanical valve are by no means trivial either. The apparently small 0.7% risk of thromboembolic complications reported in this study (no data are provided for bleeding complications) might result in one in seven patients having a thromboembolic event in the next 20 years and more than a quarter might have one or more events in their expected lifespan. Since lesser events may not be captured in the retrospective reports used in the meta-analysis, this is likely to be a considerable underestimate and will not capture the impact of repeated blood testing and drug treatment on the patient’s perception of quality of life.

There are other means of avoiding anticoagulation, including tissue valve replacement, which is more expeditious than a valve-sparing operation.¹¹ Continuing experience with non-surgical placement of an aortic valve (transcatheter aortic valve implantation) may offer a bale-out option

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in the future for failing tissue valves, whether native or bioprostheses. These may be unattractive compromises but merit consideration in what, despite outstanding achievements, have proved to be imperfect solutions for the patient with Marfan syndrome.² It is in the nature of Marfan syndrome that people with the disease face the prospect of other aortic events, further cardiovascular surgery on the aorta and operations elsewhere on a body affected by fibrillin deficiency. And that is in addition to living through a background of life events such as pregnancy and trauma, where anticoagulation undoubtedly adds complications. In considering the big picture we need to consider all competing risks and their trade-offs over the many years, which must be catered for in this often very young group of patients. The solution cannot be perfect; it need only be better than the best alternative to merit our serious consideration.

The proposal made by Golesworthy *et al* is to manufacture an exactly fitting external support derived by computer-aided design (CAD), in engineering terms CAD modelling.¹² A soft and pliant but more than adequately strong support is manufactured to fit precisely the individual patient's aorta. Without the need for cardiopulmonary bypass the device is positioned encircling the vulnerable segment of the aorta from the aorto-ventricular junction, proximal to the coronary arteries and extending beyond the brachiocephalic artery.¹³ The valve, its architecture, and the blood/endothelial interface are all completely preserved in an operation which can be performed to a well-regulated protocol.¹⁴ The whole

process has the advantage that it reinstates important elements of "the workmanship of certainty". What Golesworthy's operation does not include is resection of the Marfan aortic wall. There is animal evidence of reduction in non-collagenous, non-elastin components of extracellular matrix when arteries are wrapped in polytetrafluoroethylene,¹⁵ but does this matter? Given that the external support is intimately in contact with the aortic adventitia we expect that the combined strength will be adequate but it must be said that the exact future of the supported aortic wall is uncertain. That apart, the Golesworthy procedure achieves no less than valve-preserving surgery while, better for the patient, it avoids bypass and myocardial ischaemia.¹⁴ It is likely to greatly reduce, even if it does not obviate, the risk of dissection which is a size-related event. Once incorporated into the aortic adventitia the external support is likely to greatly lessen the hazards of dissection should it occur. Revision surgery remains an inevitable consideration for these patients and is likely to be facilitated by this reinforcement and by the total avoidance of anticoagulation.

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