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Repairing the aortic valve is best for children

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In this review, I will address the problem as to whether children with disease of the aortic valve should undergo repair, or be put forward for the Ross procedure. So as to put this debate in the correct framework, we need to evaluate all the options for aortic valvar disease. These include not only the Ross procedure, involving the insertion of a pulmonary autograft, and valvar repair, but also the option of replacing the valve with a biological or mechanical prosthesis.

Valvar replacement

The options when choosing to replace the aortic valve are mechanical prostheses, homograft valves, porcine valves, and pericardial valves. There are multiple problems with inserting mechanical valves. Such valves do not grow with the patient. Patients with such valves require anticoagulation, putting them at risk of complications from warfarin therapy. Although mechanical valves will not degenerate, they do not offer a permanent solution. Patients often develop pannus on the valve, which eventually can lead to failure and the need for replacement. The haemodynamics of mechanical valves are not as good as those of natural valves. The smallest mechanical valve available, furthermore, is one of 17 millimetres. Despite all these potential problems, sometimes a mechanical valve is the only option available.

An aortic homograft valve also does not grow with the patient. These valves are prone to early calcification. Some valves will fail early on, developing both stenosis and insufficiency. We have had the experience of having to reoperate on a patient within several months of inserting a homograft valve in a neonate. These valves are difficult to replace, because the coronary arteries require reimplantation each time a homograft root is replaced.

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The Ross procedure

The Ross operation, although thought by some to be the ideal solution for these patients, has its own set of potential complications, which must be carefully analyzed. There is a risk of dilation of the muscular pulmonary infundibulum supporting the valvar leaflets, which if occurring will lead to aortic valvar insufficiency. All these patients will require eventual replacement of the pulmonary valvar homograft inserted at the time of the Ross operation. These patients, therefore, have the potential to have disease of both arterial valves.

Repairing the aortic valve

In comparison to replacing the valve, or the Ross procedure, there are many advantages to repairing the aortic valve. The native aortic valve will grow with the patient. There is no need for anticoagulation following repair. Repair of the aortic valve means the pulmonary valve is not at risk for failure. Furthermore, the techniques used during repair do not preclude either a Ross operation in the future, or insertion of a mechanical valve when the patient is an adult.

Assessing the evidence

A review from the University of California at San Francisco evaluated the results of valvar repair as opposed to the Ross procedure.[1] In that review, the surgeons repaired the valve in 22 patients, 9 with aortic stenosis, 7 with regurgitation, and 6 with both. Of these patients, 5 required early replacement of the aortic valve, or a Ross operation, because

3 manifested residual aortic stenosis, while 2 developed aortic insufficiency. Late reoperations for aortic stenosis were needed in 3 patients. Their best results were obtained in patients with trifoliate aortic valves. No patients in their series died either early or late. These authors specifically noted that attempted valvoplasty did not preclude a late Ross procedure, or replacement of the aortic valve.

The group at Toronto[2] has reported their results with the Trussler repair for patients with ventricular septal defect and aortic valvar insufficiency. Between 1968 and 1988, an insufficient aortic valve was repaired in 70 patients in association with a ventricular septal defect, which was perimembranous in 20, and doubly committed and subarterial in 20. In two-thirds of the cases, there was prolapse of the right coronary aortic leaflet. There were no early deaths, and two late deaths in this series. The freedom from reoperation was 85 percent at 10 years. Repair of the aortic valve, therefore, especially in patients in whom the valvar insufficiency is caused by prolapse of a leaflet into a ventricular septal defect, is quite successful. Walters et al.[3] from Wayne State University have also reported excellent results in this subgroup of patients.

It should be noted that many patients who have undergone repair of a doubly committed and subarterial ventricular septal defect, where the patch abuts immediately beneath the leaflets of the pulmonary valve, will develop fibrosis and mild pulmonary valvar insufficiency following this operation. These patients are probably not candidates for a Ross operation because of this fibrotic tissue, along with the haemodynamic changes induced in the pulmonary valve secondary to both the ventricular septal defect and the later repair using a patch.

The technique of extending the leaflets of the aortic valve has been widely used in Europe and Asia. Carpentier et al.[4] have reported their experience in 89 patients undergoing surgery between 1992 and 2000. Their mean age was 16 years. All patients had extensions of all three leaflets using pericardium. Hospital mortality was 2.2 percent, with two early failures and seven late failures. After 7 years, 90 percent of these patients were free of complications related to the valve. The group from South Korea[5] reported on 34 patients undergoing surgery between 1995 and 2001, with a mean age of 31 years. All patients had valvar insufficiency, and none of them died. At a mean follow-up of 4 years, 2 patients had required a reoperation. Of the overall group, 23 patients had no regurgitation, 10 had mild regurgitation, and only one had moderate regurgitation. These authors demonstrated a decrease in both the left ventricular end-systolic and end-diastolic diameters (Fig. 1). The group from Geneva, Switzerland has also reported their

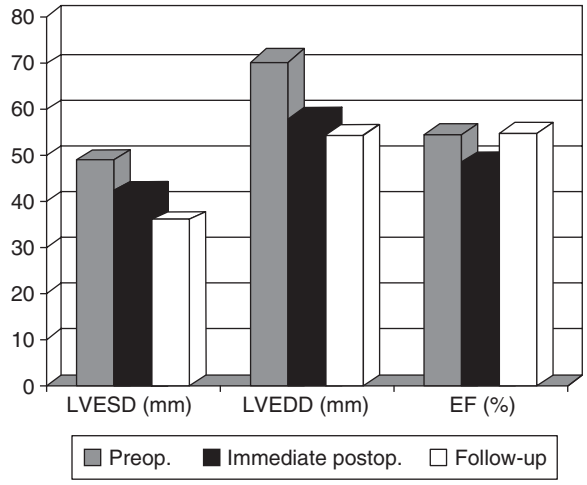

Figure 1.
Left ventricular end-diastolic (LVEDD) and left ventricular end-systolic (LVESD) diameter decrease following repair of the aortic valve. EF, ejection fraction. (From Ahn H, Kim KH, Kim YJ. Midterm result of leaflet extension technique in aortic regurgitation. Eur J Cardiothorac Surg 2002; 21: 465–469. Reproduced with permission.)

results with repair.[6] They used fresh autologous pericardium to extend the leaflets in 41 children, all with insufficient rheumatic valves. Mean age was 11 years. There were no early deaths, and one late death. At discharge, 27 patients had no insufficiency, while mild insufficiency was seen in 14, albeit that none required reoperations.

These excellent results with extension of the aortic valvar leaflets can be compared to the results of the Ross operation in children. It was Elkins et al., from Oklahoma, who pioneered the use of the Ross procedure for children in the United States of America. They reported on 178 patients undergoing surgery between 1986 and 2001.[7] The operative mortality in their series was 4.5 percent, with a late mortality of 0.5 percent. The freedom from reoperation on the autograft was 90 percent at 12 years. One-eighth of the homograft valves placed to restore continuity from the right ventricle to the pulmonary arteries, however, had either been replaced or were close to requiring replacement. Eventually, all patients undergoing the Ross procedure will need replacement of the conduit placed from the right ventricle to the pulmonary arteries.

As already discussed, the Ross operation also places the patient at risk for aortic insufficiency caused by dilation of the pulmonary autograft. The infundibular muscle, the sinuses of Valsalva, and the sinutubular junction are all known to increase in size after the Ross operation. The aetiology of this dilation is multifactorial. There is passive dilation due to exposure to increased blood pressure. A mismatch between the size of the pulmonary autograft and the diameter

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of the aortic root or ascending aorta can also cause dilation. There may also be an intrinsic abnormality of the pulmonary root associated with congenital aortic valvar disease. The group from Ann Arbor, Michigan,[8] reported having to perform valve-sparing replacement of the aortic root because of dilation of the pulmonary autograft leading to severe neo-aortic regurgitation after the Ross operation in 4 patients.

David et al. from Toronto[9] have also noted that the geometric mismatch of the aortic and pulmonary roots can cause valvar insufficiency after the Ross procedure. They reported aortic insufficiency developing in a patient who did not have reduction in the diameter of the aortic root, and replacement was necessary 2 weeks later. In another review, David et al.[10] examined their results with 118 patients who had undergone a Ross procedure. Moderate aortic insufficiency developed in 7 patients, and 3 patients required replacement of the pulmonary autograft. All patients with moderate aortic insufficiency had dilation of the aortic root and/or the sinutubular junction. In the discussion of that paper, David stated: "I believe we should abandon the technique of aortic root replacement in patients with bicuspid aortic valve. I am convinced that most patients with bicuspid aortic valve have advanced degenerative changes in the arterial wall of the pulmonary trunk."

Conclusions

Performing the Ross procedure places patients at risk of developing disease of both arterial valves. Dilation of the pulmonary autograft, with resultant aortic valvar insufficiency, unequivocally occurs in a number of patients.[10] The homograft implanted to restore continuity between the right ventricle and the pulmonary arteries will need to be replaced in all patients. Techniques designed to reconstruct the aortic valve are constantly improving, and should be employed whenever possible. Excellent results for patients with aortic insufficiency by extending the leaflets with pericardium have now been reported by a number of centres.[4–6]

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