

Aortic valve repair in children

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Submitted Oct 10, 2012. Accepted for publication Nov 14, 2012.

doi: 10.3978/j.issn.2225-319X.2012.11.08

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Introduction

We are now realizing that aortic valve disease afflicting children is for the majority a life-long condition requiring repetitive interventions. Treatment strategies should be favoured that allow the largest proportion of patients to have the smallest number of interventions over their lifetime. Surgical techniques have been refined in the last two decades making primary surgery a more attractive option for these patients.

Interventional catheterization versus surgery: a fair comparison?

It has long been assumed that in the pediatric population balloon valvuloplasty and aortic valve surgery resulted in similar outcomes, but there is emerging data showing that this assumption may no longer be correct (1). The largest study on neonates with critical aortic stenosis was a North American multicenter prospective study performed by McCrindle and colleagues who observed 82 neonates undergoing dilatation via balloon valvuloplasty versus 28 neonates treated with open aortic valve surgery (2). This study demonstrated equal rates of reoperation and survival in both groups. However, how valid this study is to current times has been questioned as the interventional techniques used are now somewhat obsolete. Specifically, 9 of the 29 open surgical patients underwent a blind transapical dilation of the aortic valve, a procedure closer in similarity to balloon dilatation than directed surgery. Additionally, the remaining surgical procedures were likely limited to simple blade commissurotomy since the mode of failure of these surgical patients was recurrent stenosis (a complication typically seen with inadequate resection of the valve nodular

dysplasia).

For the last 2 decades, pediatric aortic valve stenosis has been primarily treated by interventional catheterization due to its decreased invasiveness, and because the patients were primarily referred to cardiologists who by nature determine the treatment modality. Only a restricted number of centers continued to offer primary surgery for aortic valve disease and it is likely that most surgical teams have little expertise in these techniques for neonates.

In older children, no comparative study between treatment approaches currently exists. Yet interestingly, the ACC/AHA guidelines recommend balloon valvuloplasty for the treatment of adolescents with aortic stenosis, despite being contraindicated in adults (3). One may wonder that this difference in practice is not more dependent on the change in caring physician from pediatric to adult cardiologists than on a sudden change of the patient's innate physiology.

Evolution of open surgical techniques in the pediatric population

Debulking and resuspension of the valvular commissures in younger patients

In the last 2 decades, techniques of aortic valve repair have evolved. In Melbourne, all diseased aortic valves undergo an extensive debridement including resection of all nodular fibrosis, thinning of the leaflets, opening of the fused commissures and importantly, carving of new interleaflet triangles. It has become clear to us that unless the fibrotic material present below the fused commissure is resected, reocclusion of the commissure is likely to occur. In our initial experience, we realized that in neonates and

young infants, the effective orifice area had to be increased to achieve a durable repair. Therefore, we have opted to make bicuspid valves from unicuspid valves and tricuspid valves from stenotic bicuspid valves. As the severity of the commissural fusion increases, it is more likely to have involution of the commissures resulting in the typical dome appearance seen in unicuspid valves. Similarly, largely fused commissures lose their commissural suspension into the sino-tubular junction. Because of the loss of this commissural suspension, the opening of unicuspid valves and of largely fused commissures has to be followed by the resuspension of the edges of the incision by small triangular pericardial patches (4).

Regurgitant valves

Aortic valve disease is almost exclusively a stenotic disease in infants and only in older patients does regurgitation become a predominant feature. Repair of regurgitant valves in younger patients has benefited from the advances made in the field of adult aortic valve repair (5). The exact mechanism leading to regurgitation has to be identified and addressed by a technique still allowing the growth of the aortic structures. Prolapsing leaflets are best addressed by triangular plication if the leaflets are thin. Frequently a thick raphe restricts the motion of the largest leaflet. This raphe is best resected and quite often in children, needs to be replaced by a small pericardial patch. We have been reluctant to use Gore-Tex resuspension of the free edges of prolapsing leaflets in growing children.

Stabilization of the aortic root

The regurgitant valves of children are often slightly different than in adults because they often are associated with a stenotic component of the valve as well. Therefore, the stabilization of the aorto-ventricular junction proven to be necessary in adult cases is performed differently in children (6). Usually, the base of the interleaflet triangle is not enlarged under a fused commissure and no stabilization is necessary at this level. It may however be required to improve the coaptation height of the leaflets and to perform a subcommissural annuloplasty under a non-fused commissure or at the level of the aortic wall where the raphe is inserted. Unfortunately, sub-commissural annuloplasties are renowned to fail in the subsequent years. Undoubtedly the development of techniques to stabilize the aorto-ventricular junction is an area of keen interest, keeping in mind that

circumferential stabilization will not be possible in growing children.

Replacement of diseased sinuses

It is still unclear whether the threshold for replacing the diseased aortic wall should be the same in adults and children (3). In some specific diseases of the aortic root such as Marfan or Loeys-Dietz syndrome, valve-sparing root procedures are indicated as soon as their root can accommodate an adult-size graft (7,8). In the remaining children whose roots have not yet reached adult size, graft replacement of the aortic root should be avoided. It is generally accepted that the benefits of keeping the compliance of the native aortic root outweighs the risk of exposing the patients to a higher rate of reoperation. The occurrence of aortic dissection is exceptionally low in adolescents and young adults and it is likely that this risk does not justify a strategy of root replacement as aggressive as the one adopted in the recent years in adults (9). The risk of recurrent intervention has not yet been ascertained in this patient population and if it is found that they will require increased procedures later in life, the aortic wall may be best addressed at that time. Patients requiring a reduction of the sino-tubular junction might be better served by the plasty of the native aortic wall (10). The limited experience with this practice reported in adults may justify the investigation of this practice in the pediatric population.

Cusp extension techniques

Cusp extension techniques can be used in almost every single case of aortic valve repair as long as the hinge-points of the leaflets are still mobile. It requires the resection of the bulk of the nodular dysplasia and the suturing of patches of glutaraldehyde treated autologous pericardium to recreate new cusps. Some minor technical variations have been described by various teams (11-15). Stenotic bicuspid valves can be made tricuspid by the incision of the raphe, creating a new valve. The ease and reliability of this approach has attracted many centers to favor this technique. However, careful attention must be paid to prevent potential ischemic complications arising with the use of this technique especially in bicuspid valves made tricuspid (11,16-19). The quality of the final result depends on the length of coaptation of the extended cusps. It is therefore a natural trend to perform lengthy cusp extension leading to redundancy of patch material. In rare instances, we

suspect that this redundant patch material can obstruct the coronary orifice and lead to sudden death. Clearly, this patch material will not accommodate growth and it has now been demonstrated that as long as all leaflets are extended with patches, these repairs will last between 5 and 15 years (11).

The Ross procedure: the ultimate operation?

In the 1990's, a great hope was placed in the Ross procedure for children because the translocated autograft was proven to be viable and allowed for growth (20). These great expectations were dampened by the subsequent realization that up to a quarter of these autografts may require replacement in the following two decades because of progressive dilatation of the autograft (21). Alarmist perspectives have reported dilatation of the autograft superior to Z-score of +4 in 97% of the patients within 6 years (22). Dilatation of the aortic root and its deleterious consequences can be prevented by the use of the inclusion technique (23). Unfortunately in the pediatric population, aortic valve disease is predominantly stenotic and the vast majority of younger children have small aortic roots, thus precluding the use of the inclusion technique. Outcomes of the Ross procedure specific to the pediatric population are still unclear because all series in the literature have reported young adults and children at the same time.

Long-term outcomes and what really matter

Parents of babies and children with aortic valve disease have to be warned that this condition is likely to affect their child for the remainder of his/her life. Comparative studies have focused primarily on immediate results of a single intervention rather than analysing more longterm outcomes in a way that would be useful to give lifetime perspectives.

Management strategies in neonates

Excellent results have been reported after balloon dilatation of the aortic valve, but these reports fail to highlight the dramatic outcomes facing the neonates who require urgent surgery as a result of the failure of this strategy. Data from the STS database have recently showed mortality to be 28% for aortic valve replacement in neonates (24). This disastrous statistic is the result of the strategy of primary balloon valvuloplasty. At the Royal Children's Hospital in Melbourne, we recently reported 3% mortality for all neonates undergoing aortic valve intervention, a fact

certainly related to the preference given to primary aortic valve surgery.

Comparative outcomes of balloon valvuloplasty and surgery

The largest series of balloon valvuloplasty comes from the team in Boston (25). Interestingly, their freedom from reintervention over 15 years is quite similar to the reported freedom from reintervention after cusp extension repair of the aortic valve (11). We have recently demonstrated that patients undergoing aortic valve repair without the addition of patch material had higher chances to remain free of reintervention than if their repair had needed addition of patch material. Consequently, we have tried to restrict the amount of patch material and avoid 3 cusps extension repair in all patients where it was deemed feasible. We analysed in our own experience in neonates with aortic valve disease the proportion of patients remaining free of all adverse events (reintervention or stenosis or regurgitation). Like others, we found that patients undergoing surgery had a significantly higher chance to be free of all adverse events than those undergoing balloon valvuloplasty (1). We believe that having a larger population free of all events for the longest possible period is an event that is by far more relevant than actual freedom from first reintervention or the benefit of a less invasive procedure. For some patients, this goal will translate into keeping their native valve possibly beyond early adulthood. For others, it will postpone the need for the Ross procedure to an age where the inclusion of the autograft within the native aortic root or in a Dacron graft becomes possible.

Primary or secondary repair

After a failed balloon valvuloplasty, some patients may still undergo a repair. It has been demonstrated however that the longevity of aortic valve repair is dependent on the quality of the tissues (26). In our experience, the balloon valvuloplasty results in a tear in the thinnest, most pliable portion of the valve. Chronic regurgitation, even if well tolerated clinically, results in the thickening of the edges of the torn valve, and we believe that this damage precludes the possibility of a very long standing result after subsequent surgery. Therefore, we believe strongly that primary surgery provides better results than interventional catheterization followed by surgery. It is possible today in a larger number of patients to sculpt out of a fibrotic

valve a valve that closely resembles a native one. We are now observing that these patients have improved long-term results and we do not know if they will need any reintervention at all. Balloon dilatation is a non-reversible destructive process damaging the best portion of the leaflet and leaving in place the thickened dysplastic portion of the valve. It will prevent those who could have simple repairs to achieve an optimal outcome.

Unfortunately, preoperative investigations do not allow us to identify those who could undergo simpler repairs without addition of patches. We can only hope that progress in imaging will enable us to better delineate preoperatively the type of repair expected, allowing better preoperative selection of patients that would benefit from surgery or catheter interventions.

In conclusion, the belief that balloon valvuloplasty and surgery results in similar outcomes is likely based on wrong assumptions. Today, the techniques of valve repair have evolved and provide better results than twenty years ago. Valve repair without addition of patch material may have results lasting beyond the pediatric age. Additionally, the native stenotic aortic valve is likely to be damaged by balloon dilatation to an extent precluding a long-lasting favourable outcome from subsequent surgery. In the pediatric population, primary surgical treatment of aortic valve disease should be favoured.

Acknowledgements

This research project was supported by the Victorian Government's Operational Infrastructure Support Program. *Disclosure:* Yves d'Udekem is a Career Development Fellow of The National Heart Foundation of Australia (CR 10M 5339).

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Cite this article as: d'Udekem Y. Aortic valve repair in children. *Ann Cardiothorac Surg* 2013;2(1):100-104. DOI: 10.3978/j.issn.2225-319X.2012.11.08