Management of valvar aortic stenosis in children

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What is the best treatment for the child with valvar aortic stenosis—balloon or surgical valvotomy?

In this issue of Heart two sets of authors report their separate experience of balloon dilation for valvar aortic stenosis in a combined total of 339 consecutive children of all ages. Balloon valvoplasty in this setting was first described in 1983 and has gradually become the therapeutic procedure of choice in most units for the treatment of isolated congenital aortic stenosis. With any new treatment data takes time to emerge but 20 years down the line are we now able to put balloon aortic valvoplasty in its rightful context?

BALLOON OR SURGICAL VALVOTOMY

Surgical valvotomy for congenital aortic stenosis was the treatment for childhood aortic stenosis for over 30 years before balloon aortic valvoplasty. Unfortunately there are no trials of appropriate design to allow a valid comparison of surgery and balloon valvotomy. When considering the best treatment for childhood aortic stenosis by necessity we must rely on data which is fraught with problems. Existing studies are retrospective and non-randomised, reporting different age groups using varied techniques across a number of eras. A major problem in the literature is the significant variation in indication for intervention and reintervention.

Even allowing for such inconsistencies both techniques seem broadly comparable in terms of immediate gradient relief, procedural mortality, and longer term survival. There are relatively recent reports of encouraging survival rates with both surgery and balloon valvotomy even in the troublesome neonatal group in which triage for a Norwood approach based on an improved understanding of the limits of a biventricular repair must have played a significant part.

Valvotomy of any kind is a palliative procedure and reintervention remains frequent. As longer term data emerges reintervention rates after balloon valvotomy become of some concern. Reich and colleagues report overall intervention-free survival of 39% to 14.4 years. In the neonatal group the results are worse with 29% of patients in this study surviving to 14.4 years without reintervention and just 35% surviving without a second procedure to three years in the study by Balmer and colleagues. A mixture of recurrent aortic stenosis and important regurgitation were the indications for further procedures. These data are not out of context with the existing balloon valvoplastic literature in which other authors report 50% of children requiring reintervention by eight years after the initial valvotomy.

Are surgical results any better?

In historical surgical series 15–40% of patients have required reintervention between 5–10 years after the initial valvotomy. Undoubtedly over the 20 year story of balloon valvotomy, surgery, particularly of the left ventricular outflow tract, has improved significantly. There are a scattering of contemporary surgical reports describing excellent results with low reintervention rates (up to 90% free from reintervention at 10 years post-surgery). However, absolute numbers reported in such studies are small (possibly as a result of the increasing prevalence of balloon valvotomy as the primary treatment modality) and the same concerns about the comparability of data exist.

Studies that report non-randomised comparisons of balloon and surgical valvotomy generally report similar results (including reintervention rates) for both approaches, but important demographic differences between groups are always present. Are there particular anatomical variants that should be addressed surgically?

A particular problem for the interventional cardiologist is the severely abnormal valve, usually bicuspid or unicuspid, in which there is a high risk for procedural failure and subsequent reintervention. The application of a circumferential force to an “unbalanced” valve, often with a rudimentary third cusp, is more likely to induce cusp avulsion through the uneven distribution of force. Reich and colleagues identified “functionally bicuspid” valves as a risk factor for both aortic regurgitation and subsequent reintervention. It has been suggested that this sort of valve is better dealt with surgically. Although there are reports of skilful valve repair in this setting often with valve augmentation (it would be unfair to call this sort of procedure a simple valvoplasty), numbers are small and as yet there is no longer term follow up data. There are also surgical data suggesting that patients with this sort of anatomy come to valve replacement early whatever the approach.

Aortic regurgitation

Although reintervention after valvotomy occurs for recurrent stenosis, the major concern after any sort of valvotomy is aortic regurgitation. Quantifying aortic regurgitation after valvotomy
is difficult. Echocardiography is the mainstay of assessment but it is a far from perfect tool.22 23 In addition many (retrospective) studies fail to apply consistent and robust echocardiographic assessments during follow up. Given the occurrence of late deaths, particularly in some of the older studies, we cannot assume that lack of subsequent valve replacement means that important aortic valve regurgitation was not present.

Reich and Balmer are to be commended on their echocardiographic follow up.2 4 Using consistent criteria both sets of authors report what others have previously suggested after both forms of aortic valvotomy—that is, that aortic regurgitation is progressive. We should not be surprised by this; normal aortic valves leak progressively with age and it is possible that in anatomically abnormal valves, the process is accelerated,24 25 Unrecognised cusp avulsion and perforation after balloon valvotomy is relatively common26 27 and perhaps this further increases the rate of the degenerative process in the balloon group leading to earlier reintervention.

Complications

A key stimulus behind the introduction of balloon aortic valvoplasty was the avoidance of a median sternotomy in a patient group likely to require further surgery. The growing evidence of the neurodevelopmental consequences of cardiopulmonary bypass in the developing brain add further justification to this rationale.27

However, of serious concern after balloon aortic valvoplasty is the high incidence of procedural femoral artery damage which is a consistent feature in reported series, particularly in infants.14 Balmer and colleagues report femoral artery occlusion requiring treatment in 57% of infants less than 3 months of age.22 Magnetic resonance imaging studies of iliofemoral vessels after balloon angioplasty show that the majority of children have evidence of obstructive lesions.28 Other procedural approaches are reported in small numbers, but as yet we have little or no evidence of the long term implications for these vessels.29

WHAT CAN WE CONCLUDE

Both balloon and surgical valvotomy are firmly established as effective initial treatments in childhood aortic stenosis; as such, it is exceedingly unlikely that we will ever have robust randomised data to really say what the best treatment is for the child with aortic stenosis. We cannot conclude with any certainty that reintervention comes any earlier after balloon valvotomy, but it seems clear that aortic regurgitation is progressive and reintervention almost inevitable. As in many aspects of the management of the patient with congenital heart disease difficult anatomy is a challenge for everyone concerned with treatment. Further consideration to the approach for balloon angioplasty, particularly in infants and neonates given the high incidence of long term femoral artery damage, seems appropriate.

REFERENCES