

Balloon Valvuloplasty and Angioplasty in Pediatric Practice

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Congenital heart defect (CHD) is the most common congenital anomaly in children and constitutes approximately 1% of live births. This will result in nearly 35,000 babies born with heart defects in the USA alone. Nearly 50% of these babies can be managed with simple medications, observation and follow-up without any major therapeutic intervention. However, the remaining 50%, in the past, required surgical, some under cardiopulmonary bypass, intervention. Since the advent of transcatheter techniques, 50% of these babies can be managed with less invasive, percutaneous, transcatheter techniques. In this editorial the role of balloon valvuloplasty and angioplasty in the management of children with CHDs will be reviewed.

In early 1950s, Rubio-Alvarez and Limon-Lason used a modified ureteral catheter to perform transcatheter pulmonary and tricuspid valvotomy. A decade later, Dotter and Judkins utilized a gradational transcatheter dilatation technique to open stenotic and occluded peripheral arteries. In mid to late 1970s, Gruntzig and his colleagues extended Dotter and Judkins' technique and successfully dilated stenotic lesions of the iliac, femoral, popliteal, renal and coronary arteries; they utilized a double-lumen catheter with a non-elastic balloon that they developed. Balloon dilatation techniques were then extended to relieve obstructive lesions of the aorta, pulmonary valve, aortic valve, mitral valve, subaortic membrane, branch pulmonary artery stenosis, stenotic bioprosthetic valves and other obstructive vascular lesions. Extensive review of the techniques and results may be found in our previous publications on this subject [1-8].

Pulmonary stenosis

Valvar pulmonary stenosis, initially described by Kan and her associates [9], is the first CHD for which balloon valvuloplasty is acknowledged as a therapeutic procedure of choice. The indications for catheter intervention are the same as those used for surgical valvotomy: a moderate degree of pulmonary valve stenosis with a peak-to-peak gradient >50 mmHg with normal cardiac index. Initially recommended size of the balloon is 1.2 to 1.4 times that of pulmonary valve annulus. Relief of pulmonary valve obstruction by balloon valvuloplasty is documented in neonates, infants, children and adults. Immediate and short-term results are available but, there are limited long-term follow-up data [7]. Doppler echocardiographic evaluation at follow-up is reflective of the results without the necessity for repeat cardiac catheterization. Recent studies suggested pulmonary insufficiency as potential problem [7,10,11]. It appears that use of large non-compliant balloons may be associated with pulmonary insufficiency [11]. Consequently the revised recommendations are to achieve balloon/annulus ratios of 1.2 to 1.25 [12,13]. Balloon pulmonary valvuloplasty is an excellent and preferred alternative to open or closed heart surgery in the treatment of pulmonary stenosis in patients of all age groups. Long-term follow-up evaluation of pulmonary insufficiency is required.

Aortic stenosis

Lababidi [14] was the first to apply balloon valvuloplasty technique to relieve congenital valvar aortic stenosis. Subsequently several others groups of workers have used this technique. The indications for balloon valvuloplasty are also the same as those used for surgical valvotomy; peak-to-peak gradient >70 mmHg irrespective of symptoms or gradient >50 mmHg with associated symptoms and/or ST-T wave changes on the electrocardiogram. The size of the balloon used should be 80%

100% of the aortic valve annulus. Immediate and midterm results have generally been favorable producing reduction of aortic valve gradients by 60% of pre-valvuloplasty values. Complications, such as arterial occlusion, especially in young children, and aortic insufficiency may occur. Restenosis at intermediate-term follow-up has been reported and could be minimized by reducing the gradients to <30 mmHg and by use of adequate size balloons [15]. There is scanty data on long-term follow-up [7]. The available data indicate that aortic insufficiency may develop in significant number of patients, requiring aortic valve replacement or Ross procedure. While immediate and short-term results of balloon aortic valvuloplasty in children are generally encouraging, long-term follow-up studies (10 years and beyond) are necessary to evaluate long-term effectiveness, especially in view of late development of aortic insufficiency.

Native aortic coarctation

Since the description of surgical correction by Crafoord and Nylin and Gross and Hubbard in mid 1940s, surgery has become a standard therapy of choice. More recently, balloon angioplasty techniques have been utilized in the management of aortic coarctation. Gruntzig's technique of balloon angioplasty was adopted by Sos, Singer, and Sperling and their associates, to enlarge coarcted aortic segments in a postmortem specimen, post-surgical recoarctation and native coarctation respectively. This is followed by a report of successful use of this technique in a sizeable population of patients by Lababidi. However, skepticism was expressed: "Now that we can dilate, should we?"; this is presumably based on poor results in a dog model and unsuccessful attempts in less than a handful of patients. Reports of aneurysms and flawed interpretation of VACA (Valvuloplasty and Angioplasty of Congenital Anomalies) Registry data have had an additional negative impact [16,17] on the use of balloon therapy of native coarctation of the aorta.

Some groups of workers and we continued to advocate balloon angioplasty as a reasonable alternative to surgical therapy. We have examined the effectiveness and safety of balloon angioplasty [17] and carefully compared balloon with surgical results [18] and concluded that balloon angioplasty is safe and effective and compares favorably with surgery.

The indications for intervention are a peak-to-peak systolic pressure gradient >20 mmHg across aortic coarctation with hypertension and/or congestive heart failure. Initial balloon dilatation is performed with a balloon whose diameter is an average of aortic isthmus or transverse aortic arch and descending aorta at the level of diaphragm. If there

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is no adequate relief of obstruction i.e. gradient < 20 mmHg and angiographic improvement, repeat dilatation with a balloon as large as the diameter of the descending aorta at the level of diaphragm should be undertaken. It is extremely rare that the balloon size should ever need to exceed the descending aortic size. Immediate and intermediate-term follow-up results are extensively documented and are generally good with a small probability of recoarctation and an aneurismal formation at the site of balloon dilatation [17]. The rate of recoarctation is higher in neonates and infants than in children. However, the recoarctation can be successfully treated with repeat balloon or surgical therapy. Echo-Doppler studies and nuclear magnetic resonance imaging techniques are useful adjuncts to cardiac catheterization and selective cineangiography in the evaluation of follow-up results.

To the best of my knowledge, there are few studies [7] that examined long-term (beyond 5 years) results. Despite problems of recoarctation and aneurysm formation at the intermediate-term follow-up, some requiring repeat intervention, late follow-up results seem encouraging in that there was minimal (2%) incidence of late recoarctation and no late aneurismal formation. Twenty-three percent of patients had blood pressure above 95th percentile at late follow-up compared to hypertension in 89% of subjects prior to angioplasty. Overall, the results appear promising, although longer-term (15 to 20 year) follow-up may be necessary to reaffirm these good results.

Based on an extensive review of the literature and personal experience with the procedure for a period in excess of 20 years, some generalizations with regard to balloon therapy may be made:

1. Children, older than 1 year, and adults with discrete native coarctation are candidates for balloon dilatation. There is a reasonable consensus on this issue among most cardiologists. Long-segment coarctations or those associated with significant isthmic hypoplasia may be candidates for stent placement, especially in adolescents and adults.
2. Recurrent coarctation following previous balloon angioplasty may be treated with repeat balloon angioplasty; others prefer surgery. If the recoarcted segment is long, surgical treatment in younger children and stents in adolescents and adults would seem appropriate.
3. Treatment of neonatal and infant (< 3 months) coarctation is perhaps the most controversial issue. Many cardiologists prefer surgical intervention whereas a few cardiologists, including myself, opt for balloon angioplasty.
4. For long-segment narrowing, stent deployment is evolving to be a major therapeutic option, especially in adolescents and adults.

Other uses

There are a large number of other congenital, acquired or post-surgical stenotic lesions which can be effectively treated with balloon therapy [2,3,5,6,19,20] and include stenotic pulmonary valve in tetralogy of Fallot or other single ventricle lesions, post-surgical aortic recoarctation, branch pulmonary artery stenosis, mitral valve stenosis, some forms of Takayasu's arteritis, heterograft or homograft valve/conduit stenosis, supraaortic stenosis following arterial switch procedure, baffle stenosis following Mustard or Senning procedure for transposition of the great arteries, vena caval obstructive lesions, stenosed Blalock-Taussig shunts, obstructed collateral vessels in pulmonary atresia patients. Discussion of these lesions is beyond the scope of this presentation.

Conclusion

Balloon dilation of stenotic lesions of the cardiovascular system in children is feasible, safe and effective and should be first line therapeutic option. While the causes of restenosis during initial studies appear to be related to technical issues, it is now increasingly recognized that the substrate, the pathology of the stenotic lesion, is important determinant of the ineffectiveness of dilatation. While there are a large number of studies of short-term and intermediate-term results, there are only few studies investigating long-term results. In stenotic long-segment vascular lesions, especially in the adolescent, stent therapy may serve as a good option.

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