Pak Heart J

CHALLENGES AND OUTCOMES OF BALLOON AORTIC VALVULOPLAST (BAV) IN CHILDREN

Maad Ullah¹,Amjad Mehmood ², Sajid Ali Shah³, Nadeem Sadiq⁴, Mehboob Sultan⁵, Hajra Akbar⁶

^{1,2,4-6}Paediatric Cardiology Department, AFIC/NIHD, Rawalpindi-Pakistan

³Paediatric Department CMH, Quetta-Pakistan

Address for Correspondence:

Maad Ullah,

Paediatric Cardiology Department, AFIC/NIHD, Rawalpindi-Pakistan.

Email: maad107@yahoo.uk.com

Date Received: December 22,2016 Date Revised: January 15,2017 Date Accepted: January 19,2017

Contribution

MU, AM conceived the idea, designed and conducted the study and analyzed the data. SAS, NS, MS helped in acquisition of data and did statical analysis. HA did critical review. All authors contributed significantly to the submitted manuscript.

All authors declare no conflict of interest.

This article may be cited as: Ullah M, Sadiq N, Shah SA, Mehmood A, Sultan M, Akbar H. Challenges and outcomes of balloon aortic valvuloplast (BAV) in children . Pak Heart J 2017; 50 (02): 79 - 83.

ABSTRACT

Objective: This study was aimed to determine acute success and complications, in pediatric patients undergoing balloon aortic valvuloplasty (BAV).

Methodology: This cross sectional study consisted of patients with severe aortic valve stenosis underwent aortic valve ballooning from August 2010 to April 2015. All patients had single procedure. Clinical, hemodynamic, and follow-up morbidity and mortality data was collected.

Results: A total of 81 patients were included with cohort mean age was 90.5 ± 9.8 months. Male to female ratio was 3:1.BAV was performed for symptomatic relief and to gain time for surgical aortic valve replacement. The mean pressure gradient decreased from 80 ± 6 mm of Hg to 25 ± 7 mm of Hg. The mean aortic valve annulus increased from 12 ± 2 mm to 15 ± 3 mm. Serious adverse events occurred in 5 patients (6.17%), death in one child (1.2%)during procedure ,ventricular premature contractions in 3 (3.7%), bradycardia in 1 (1.2%) and mortality rate 1 (1.2%).

Conclusion: Balloon aortic valvotomy is a rewarding procedure in pediatric age group with good results. This procedure relieves symptoms as well as preserves left ventricular function for future aortic valve replacement.

Key Words: Aortic Valve Stenosis, Cardiac Catheterization, Valvuloplasty

INTRODUCTION

The aortic annulus is of varying anatomy when considered for valvuloplasty, the leaflets may be thickened and the commissures are fused to varying degrees. Dysplastic or unicuspid valves are often seen in newborns, are present in about 10% of infants and 3% of older children in whom the treatment is indicated. Tricuspid valves are seen in 25% of infants and in 40% of older patients who require treatment. In most of the cases the stenosed aortic valve has two cusps. The bicuspid aortic valve is of two type's i.e. functional or unbalanced and anatomic or balanced bicuspid aortic valve. The difference between the two forms is that the functional one has three sinuses and unequal sized cusps hence called the unbalanced and the anatomic one has two sinus of Valsalva and equal size two cusps.² This anatomic concept is important in regard to the prognosis of the valvuloplasty.³ In the balanced bicuspid valves as well as in tricuspid stenotic valves, the orifices are usually enlarged by a splitting of the functioning commissures, whereas in the unbalanced bicuspid valves, the fused cusp is often torn aside from the rudimental commissure.presumably due to unequal rigidity of the different sized cusps.

Aortic stenosis causes left ventricular pressure overload. Despite the high left ventricular pressure, the wall stress throughout the systole is usually not higher than normal because of an increased left ventricular wall thickness. Also in these patients there is shortening of diastolic phase with increase in ejection time.⁴ Due to the adaptation, the systolic left ventricular function is usually well maintained over a long period of time. Diastolic function varies according to the severity of the left ventricular hypertrophy. In a pronounced hypertrophy, the compliance of the left ventricular wall is markedly decreased and the left ventricular end diastolic pressure is elevated. Elevated end diastolic pressure and a shortened diastole duration contribute to a limitation of the subendocardial coronary flow. In a severe stenosis, the restriction of the coronary flow causes subendocardial ischemia during exercise or even at rest. Acute myocardial ischemia during exercise may cause ventricular arrhythmias and a syncope or sudden death.⁵ Another sign of severe stenosis is the inability of the left ventricle to adequately increase cardiac output with exercise, which is reflected by an insufficient systolic pressure rise or even by a systolic pressure drop on exertion and can sometime lead to cardiac arrest.6

Some infants present with congestive heart failure due to the left ventricular dysfunction. In some, subendocardial ischemia causes left ventricular endocardial fibroelastosis and fibrosis of the papillary muscles with mitral insufficiency.^{7,8} The majority of patients are asymptomatic and the disease is usually diagnosed by a murmur. Other most common signs are fatigue, exertional dyspnea, poor ejection fraction, chest pain and syncope. In a natural

course, most of the patients with severe a ortic stenosis die suddenly. $\ensuremath{^{9}}$

This study was aimed to determine the success, complications, trouble shooting, and survival of patients after balloon aortic valvuloplasty (BAV).

METHODOLOGY

This cross sectional study was performed at AFIC from August 2010 to April 2015 .Starting in 1999, aortic valvuloplasty has been used at Armed Forces Institute of Cardiology (AFIC)/ National Institute of Heart Diseases (NIHD) as the only method of treatment of pediatric aortic valve stenosis. AFIC/NIHD is the best paediatrics cardiac institute of Pakistan Army and one of the best Institute of Pakistan. It is well equipped with latest machinery and has state of the art peadiatric ICU. The details of all consecutive patients who received an aortic valvuloplasty at our institution were reviewed. Patients with other associated cardiac defects except coarctation of the aorta, patients less than one year of age and those who had previous cardiac surgery were excluded from study.

The indication for aortic valvuloplasty was based on a modification of the method of Hossacket al^{10} . Peak instantaneous gradient was measured by Doppler method and patients having values of ≥ 75 mm Hg or a smaller values but with signs of cardiogenic shock, syncope, left ventricular strain on the ECG, angina pectoris, fatigue on exercise, duct-dependent circulation,low cardiac output or severe left ventricular dysfunction were included in the study.

Informed consent was taken from parents before procedure. The procedure was carried out with sedation and local anesthesia. Right femoral artery was used for percutaneous cardiac catheterization. After insertion of catheter the artery was heparinized. A systolic frame of the aortography was used to locate the position of effective orifice by a jet of noncontrast blood as a road map. Right coronary catheter was used to guide a hydrophilic polymer coated wire through the valve orifice. The angle of attack was adjusted by two different kinds of movements in the sagittal plane by rotating the catheter and by pulling or pushing the catheter in the frontal plane. The wire was slided into the left ventricle by guiding the wire into the orifice by rapid but gentle stabs. In some patients we used an angled tip wire. To get fine angle changes in certain situations we had to cut off the pigtail catheter tip. The pressure gradient was measured once the wire reached the left ventricle. A J-shaped exchange wire was then inserted into the left ventricular apex and an appropriate balloon catheter was advanced over the wire. The exchange wire was aim to the apex rather than to the inflow portion of the left ventricle so that the mitral valve was not exposed to risk by the balloon inflation. In two patients prograde approach using patent foramen ovale was used. This way the catheter was introduced into the left ventricular inflow, and a J-shaped hydrophilic polymer coated guidewire was turned upward in the left ventricular apex into the aortic valve and advanced to the descending aorta. The catheter was then pushed over the wire to the descending aorta, the wire was exchanged, and an appropriate balloon catheter was advanced into the valve, where a waist was produced by the stenotic valve as shown in figure 1.

Figure 1: Balloon Catheter Positioned at Appropriate Site, Waist Showing the Stenosed Aortic Valve



During the dilation, care was taken to protect the anterior mitral leaflet and its chordae by keeping the loop in the left ventricle wide open and extended as far as possible into the apex.

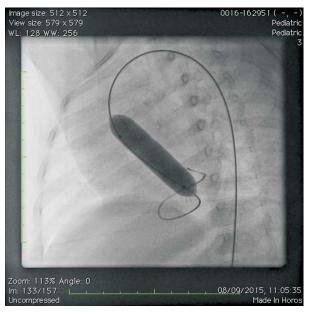
To prevent air embolism due to a balloon rupture, the balloon was gently flushed with CO_2 before the insertion. It was further deflated after the insertion by fluoroscopically controlled flushes with diluted contrast medium in the thoracic descending aorta. Using the balloon radiopaque markers and the road map, the balloon (VACS II) was then positioned to straddle the aortic valve and gradually inflated. Before the full inflation was reached, tiny position adjustments were made if necessary. Once the waist caused by the stenotic valve was abolished, the balloon was quickly deflated. (Figure 2)

During inflation, the balloon in the valve should remain as steady as possible. Displacement of the balloon prior to its full inflation prevents the effective valve dilation and to-andfro movement of the inflated balloon may cause injury to the valve leaflets or surrounding tissues.

Post procedure patients were monitored using ECG and Echocardiography to look for any immediate complications. Aortic regurgitation before and after the aortic valvuloplasty

Pak Heart J 2017 Vol.50 (02) : 79 - 83

Figure 2: Waist Abolished Due to Dilation of Balloon



was evaluated. Aortic regurgitation was assessed echocardiographically and was classified as severe, moderate, mild and trivial.¹¹

Patients were discharged once stable with femoral pulses returned. Any complication or intervention required during or after procedure was recorded.

Procedure was regarded successful if pressure gradient reduced to less than 50 mm of Hg after the procedure and failed if it remained more than 50 mm of Hg after the procedure.

All the collected data was analyzed in SPSS version 23. Mean \pm SD were calculated for numerical variables like age (in months) of the children. Frequencies and percentages were calculated for categorical variables.

RESULTS

Up to April 2015, 81 consecutive patients were treated by aortic valvuloplasty without having a previous cardiac intervention or surgery. All 81 patients were included in this study. The cohort mean age was 90.5 ± 9.8 months. Male to female ratio was 3:1.BAV was performed for symptomatic relief and to gain time for surgical aortic valve replacement. Retrograde approach was used in two patients only and in the remaining antegrade approach was used. The mean pressure gradient decreased from 80 ± 6 mm of Hg to $25 \pm$ 7 mm of Hg. The mean aortic valve annulus increased from 12 ± 2 mm to 15 ± 3 mm as given in table 1. The procedure was successful in 77 patients (95%) in first attempt and 4 patients (5%) required repeated ballooning. Serious adverse events occurred in 5 patients (6.17%), death in one child (1.2 %) during procedure, ventricular premature contractions in 3 (3.7%), bradycardia in 1 (1.2%), mild aortic regurgitation in 10 (12%) and mortality rate 1 (1.2%). Of total, 5 (6%) patients required temporary pacemaker and 8 (10%) patients required adenosine during the procedure.

Table 1: Pre and Post Procedure Changes in Pressure Gradient and Aortic Valve Annulus (n=81)

	$\begin{array}{l} \text{Pre procedure} \\ \text{(Mean } \pm \text{SD)} \end{array}$	$\begin{array}{l} \text{Post procedure} \\ \text{(Mean } \pm \text{SD)} \end{array}$	P value
Pressure Gradient (mmHg)	80 ± 6	25 ± 7	< 0.001
Aortic valve annulus(mm)	12 ± 2	15 ± 3	<0.001

DISCUSSION

The normal aortic valve area is approximately 2.0 cm². Quantification of Aortic stenosis is done on the basis of mean gradient across the aortic valve, peak aortic jet velocity, aortic valve area. Aortic stenosis may be mild (mean gradient less than 20 mmHg, peak aortic jet velocity 2.5-2.9m/s or aortic valve area more than 1.5cm²), moderate (mean gradient 20-39 mmHg, peak aortic jet velocity 3-3.9m/s and aortic valve area1.0-1.5cm²) and severe (mean gradient, aortic iet velocity greater than 4m/s and aortic valve area less than 1 cm²) ¹². In association with normal cardiac output, severe stenosis causes a peak systolic gradient over 75 mmHg. Severe aortic stenosis is an indication for treatment. In our cohort children with mild aortic stenosis were, those with moderate stenosis were and the rest were with sever aortic stenosis. Treatment options include percutaneous balloon valvuloplasty and surgical aortic valvotomy. Each procedure has its merit and demerits and it is difficult to decide that which one is the best treatment option for congenital aortic stenosis. Its individual option to adopt either one or the other intervention¹³. In our institution the primary intervention for congenital aortic stenosis used is balloon aortic valvuloplasty.

Peak instantaneous gradient was measured by Doppler method and patients having values of \geq 75 mm Hg or a smaller values but with signs of cardiogenic shock, syncope, left ventricular strain on the ECG, angina pectoris, fatigue on exercise, duct-dependent circulation, low cardiac output or severe left ventricular dysfunction were included in the study. Almost same criteria was used for BAV by L Wu.¹⁴ The patients included in their study were 14 which is much less than our study. The mean age at the time of intervention in their study was 17.1 ± 10.5 years and most of the patients were female. In our study the mean age at intervention was 90.5 ± 9.8 months but in contrast to study by L Wu most of the patients in our study were male. In an Egyptian study the mean age at time of intervention was 5.6 ± 3.7 years and again in contrast to our study most of the patient were male

Pak Heart J 2017 Vol.50 (02) : 79 - 83

as compared to female. The cohort included in their study was patients with gradient of \geq 50 mmHg.¹⁶ In a study from Thailand the mean age at intervention was 65 months and total number of patients 60 with male predominance. Aortic valve stenosis classification was done on the basis of peak instantaneous systolic gradient and mean pressure gradient.

Significant improvement in aortic valve annulus and mean pressure gradient in our study was observed (The mean pressure gradient decreased from $80 \pm 6 \text{ mm}$ of Hg to $25 \pm 7 \text{ mm}$ of Hg and the mean aortic valve annulus increased from $12 \pm 2 \text{ mm}$ to $15 \pm 3 \text{ mm}$). Our results are comparable to studies done in other part of the world reporting improvement in mean pressure gradient from $66.7 \pm 9.8 \text{ mmHg}$ to $20.65 \pm 2.99 \text{ mmHg}$, from $59 \pm 22 \text{ mmHg}$ to mean $24 \pm 12 \text{ mmHg}$ and from $69 \pm 26 \text{ mmHg}$ to $29 \pm 13 \text{ mmHg}^{14.15,17}$.some studies have reported that certain morphological features of aortic valve respond well to aortic balloon valvuloplasty.^{1, 3} However in our cohort we did not study this relationship and further work up is advised to establish the validity of this relationship in our institution.

Only about 6% of patients suffered from serious adverse event in our study. The mortality in our study was 1.2%. Ventricular premature contraction and bradycardia were observed in 3.7% and 1.2%, respectively. The mortality and morbidity in our study is comparable with other centers. Mortality reported for aortic balloon valvuloplasty by different centers is 7%, 9.5% and 5.9% which is more than our study.¹⁴⁻¹⁶ The morbidity reported by different centers is mostly aortic regurgitation (7%), restenosis requiring repeat valvuloplasty (14%), congestive heart failure (21%) and arrhythmias (17%).¹⁴⁻¹⁶ balloon valvuloplasty has excellent long term results. Risk factors for later aortic valve replacements include post dilation gradient more than 30mmHg and aortic insufficiency. Additional risk factors in which there is increased chance of re intervention include age at intervention less than 30 days, additional cardiac defects and high pre dilation gradient.¹⁸ In our study we only studied short term outcomes of aortic valvuloplasty. Further studies are recommended to look for long term outcomes of aortic vulvoplasty.

CONCLUSION

Balloon aortic valvotomy is quite rewarding in pediatric age group with good results. This procedure relieves symptoms as well as preserves left ventricular function for future aortic valve replacement. Complications of this procedure are minimal in expert hands and if adverse events do occur they are manageable.

REFERENCE

1. Petit CJ, Gao K, Goldstein BH, Lang SM, Gillespie SE, Kim SI, et al. Relation of aortic valve morphologic characteristics to aortic valve insufficiency and residual stenosis in children with congenital aortic stenosis undergoing balloon valvuloplasty. Am J Cardiol 2016; 117(6):972-9.

- Pan X. Aortic valvular stenosis. In: Butera G, Chessa M, Eicken A, Thomson J, editors. Cardiac catheterization for congenital heart disease. New York: Springer: 2015. p. 243-60.
- 3. Maskatia SA, Justino H, Frank F, Crystal MA, Mattamal RJ, Petit CJ. Aortic valve morphology is associated with outcomes following balloon valvuloplasty for congenital aortic stenosis. C a t h e t e r Cardiovasc Interv 2013;81(1):90-5.
- Loudon M, Bissell MM, Dyverfeldt P, Carlhall CJ, Ebbers T, Hess AT, et al. Turbulent kinetic energy in theascendingaorta is greater in bicuspid than tricuspid aortic valve stenosis. J Cardiovasc Magn Reson 2015;17:88.
- 5. Hameed AB, Rahimtoola SH. Congenital aortic stenosis: pregnancy is another dimension. J Am Coll Cardiol 2016;68(16):1738-40.
- 6. Saiki H, Sugimoto M, Senzaki H. Exercise-induced cardiopulmonary arrest in a child with aortic stenosis. Cardiol Young 2016;26(5):1013-6.
- Singh A, Jerosch-Herold M, Greenwood JP, Berry C, Dawson DK, Lang CC, et al. Myocardial perfusion reserve but not fibrosis predicts outcomes in initially asymptomatic patients with moderate to severe aortic stenosis: the PRognostic Importance of Microvascular Dysfunction in AS study-PRIMID AS. J Cardiovasc Magn Reson 2016;18(1):36.
- Dusenbery SM, Jerosch-Herold M, Rickers C, Colan SD, Geva T, Newburger JW, et al. Myocardial extracellular remodeling is associated with ventricular diastolic dysfunction in children and young adults with congenital aortic stenosis. J Am Coll Cardiol 2014; 63(17):1778-85.
- 9. Amano M, Izumi C, Obayashi Y, Fuki M, Imanaka M, Nishimura S, et al. The recent trend of natural history and prognosis in patients with symptomatic severe aortic stenosis. Circulation 2016;134:14319.
- 10. Hossack KF, Neutze JM, Lowe JB, Barratt-Boyes BG. Congenital valvar aortic stenosis: natural history and

assessment for operation. Br Heart J 1980;43(5):561-73.

- 11. Tribouilloy C, Avinee P, Shen WF, Rey JL, Slama M, Lesbre JP. End diastolic flow velocity just beneath the aortic isthmus assessed by pulsed Doppler echocardiography: a new predictor of the aortic regurgitant fraction. Br Heart J 1991;65(1):37-40.
- Baumgartner H, Orwat S, Sade E, Bermejo J. Heart valve disease (aortic valve disease): aortic stenosis. In: Lancellotti P, Zamorano J, Habib G, Badano L, editors. The EACVI textbook of echocardiography. 2nd ed. Oxford: Oxford University press; 2017.
- Hill GD, Ginde S, Rios R, Frommelt PC, Hill KD. Surgical valvotomyversus balloon valvuloplasty for congenital aortic valve stenosis: a systematic review and meta-analysis. J Am Heart Assoc 2016;5(8): 3003-931.
- 14. Wu L, Qi C, He L, Liu F, Lu Y, Huang G. Balloon valvuloplasty for congenital aortic valve stenosis in children. Zhonghua Er Ke Za Zhi 2014;52(9): 699-702.
- 15. Al Marshafawy H, Al Sawah GA, Hafez M, Matter M, El Gamal A, Sheishaa AG, et al. Balloon valvuloplasty of aortic valve stenosis in childhood: midterm results in a children's hospital, Mansoura University, Egypt. Clin Med Insights Cardiol 2012;6:57-64.
- Sangtawesin C, Layangkool T, Kirawitaya T, Promphan W, Jamjureeruk V, Thongthip P. Percutaneous balloon aortic valvuloplasty at the Queen Sirikit National Institute of Child Health: 25 years' experience. J Med Assoc Thai 2014;97:5117-21.
- Soulatges C, Momeni M, Zarrouk N, Moniotte S, Carbonez K, Barrea C, et al. Long-term results of balloon valvuloplasty as primary treatment for congenital aortic valve stenosis: a 20-year review. Pediatr Cardiol 2015;36(6):1145-52.
- Sullivan PM, Rubio AE, Johnston TA, Jones TK. Long-term outcomes and re-interventions following balloon aortic valvuloplasty in pediatric patients with congenital aortic stenosis: a single-center study. Catheter Cardiovasc Interv 2017;89(2):288-96.