CASE REPORT

COMBINED SEVERE AORTIC AND PULMONARY VALVULAR STENOSIS IN A CHILD AND ITS MANAGEMENT: A CASE REPORT

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Combined congenital aortic and pulmonary valvular stenosis is a rare congenital heart defect. Prevalence of severe combined valvular stenosis of aortic and pulmonary valve accounts about 0.01% and also has association with many syndromes. This combination presents unusual diagnostic as well as management problems. Apart from a few case reports, there is little in the literature on the combined stenosis of both semilunar valves and its management. We present this rare combination in a 9 year old boy which was promptly managed with the balloon valvoplasty without any complications in the same setting under local anesthesia with sedation.
Keywords: Valvular aortic stenosis, Valvular pulmonary stenosis, Semilunar valves, Balloon valvoplasty

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INTRODUCTION

Congenital aortic and pulmonary valvular stenosis is not an uncommon defect in pediatric population. Prevalence of isolated pulmonary stenosis in pediatric population in local regional data is 2.6%[1] and around 8-10%[2] in the western world literature. The prevalence for isolated aortic stenosis is 1.4%.[1] The combined stenosis of both valves in the same patient is a rare entity. The combined valvular stenosis is usually associated with other congenital heart defects like Atrial Septal Defect, Ventricular Septal Defect and Patent Ductus Arteriosus. Occasionally, it may have syndromic association like Noonan and William syndrome. But isolated severe left and right outflow tract obstruction is very rare in literature and only 16 cases have been reported.[3-10] The first report was published in 1957 by Horlick[3] and treated with surgical option. Later on, another case was reported by Chandrashekhar Y et al and managed by balloon valvoplasty instead of surgery.[6] The increased importance of combined aortic stenosis (AS) and pulmonary stenosis (PS), in addition to its rarity, is the fact that delayed diagnosis and management may have catastrophic consequences. The procedure was done under local anesthesia with sedation which also has its own implications in developing countries where pediatric anesthetist is not readily available.

CASE REPORT

Nine years old boy with normal growth and developmental parameters, presented with history of mild distress on exertion (NYHA class I), and parental concern about chest deformity and hyper dynamic precordium. He was product of consanguineous marriage. On physical examination, the weight was on 50th centile and height was on 25th centile. He was pink in room air with pulse of 94 beats/min, regular and synchronous and blood pressure (BP) of 108/70 mmHg. Cardiac examination revealed a precordial bulge, pulsatile and hyper dynamic precordium with apex beat in 6th intercostal space which was heaving in character (well sustained heave). Left parasternal heave was also palpable. Systolic thrill was palpable in both left and right 2nd intercostal and supra sternal notch. On auscultation, there was a grade IV ejection systolic murmur in left 2nd intercostal space with an additional grade IV ejection systolic murmur in right 2nd intercostal space with radiation to neck.

Electrocardiogram (ECG) revealed sinus tachycardia with normal left axis deviation. There were also features of bi- ventricular hypertrophy with deep S waves in lead V1 and V6 but left ventricular hypertrophy was more marked. Chest X Ray revealed cardiomegaly with predominant left ventricle (LV) as apex was formed by LV along with no uplifting of apex. But right atrium (RA) and pulmonary artery (PA) shadow was also enlarged suggesting severe pulmonary stenosis as well. Two-dimensional echocardiography showed a thickened, dysplastic, bicuspid aortic valve with maximum instantaneous gradient of 96mmHg and mean gradient of 78mmHg with peak velocity of 4.9m/s across the valve. The aortic annulus was 12.3mm and there was marked hypertrophy of left ventricle. Additionally, there was doming pulmonary valve with maximum instantaneous gradient of 121mmHg and mean gradient was 97mmHg with peak velocity of 5.5m/s across the valve. On M-mode, there is normal systolic LV function with ejection fraction (EF) of 69% and tricuspid annular plane systolic excursion (TAPSE) of 18mm. The pulmonary valve annulus measured 16.1mm.

The patient was taken to cardiac catheterization lab where dilatation of both aortic and pulmonary valves was done with balloon in the same setting. The procedure was performed under local anesthesia with sedation. The catheterization revealed a thickened aortic valve with severe aortic stenosis and trace aortic regurgitation (AR) and gradient across the aortic valve was 153mmHg with peak LV systolic pressure of 300 mmHg (Mean 122 mmHg). The aortic annulus was measured to be 12.1mm on fluoroscopy. The aortic valve was crossed and dilated with a 12x3 mm Osypka balloon (VACS II) under cardiac pacing at 210 bpm (Figure 1). Post balloon dilation, the gradient across the aortic valve dropped to 50mmHg with mild AR and peak LV pressure of 155mmHg (Mean 70 mmHg).

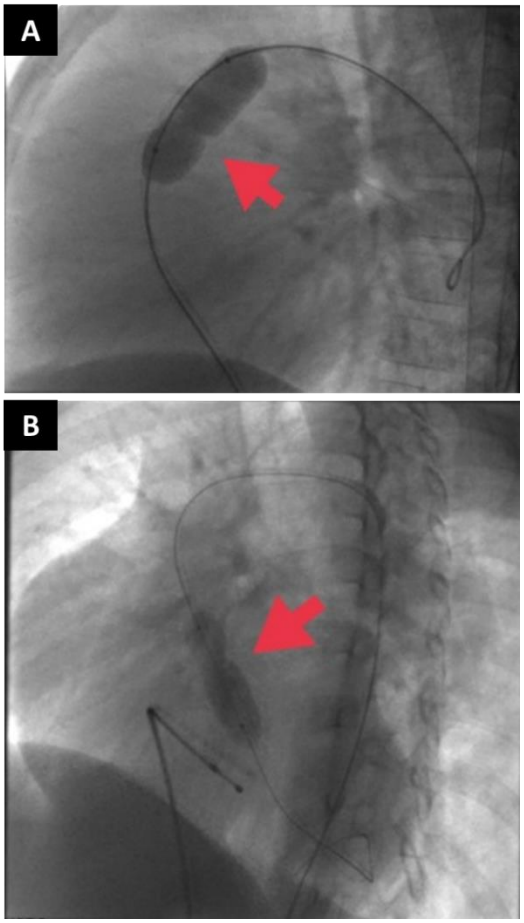

Figure 1: Pulmonary (A) and Aortic (B) balloon valvoplasty by using Osypka balloon (VACS II)

After aortic balloon valvoplasty, balloon dilatation of pulmonary valve was done. Gradient across the pulmonary valve was 96mmHg with mild pulmonary regurgitation. There was also marked post stenotic dilatation of pulmonary artery. The pulmonary valve annulus measured to be 14.1mm and pulmonary balloon dilatation was done with 18x3mm Osypka balloon (VACS II) (Figure 1). The gradient across pulmonary valve reduced to 35 mmHg after dilatation. No complication was observed during procedure and post procedure 2D echocardiography revealed aortic valve maximum instantaneous gradient of 4mmHg and pulmonary valve gradient of 49 mmHg (Table 1).

Table 1: Pressure pre and post balloon pulmonary and aortic valvoplasty

Site	Pre-Balloon Valvoplasty (Peak, Mean mmHg)	Post-Balloon valvoplasty (Peak, Mean mmHg)
RV	181(89)	112(68)
LPA	85(41)	77(44)
LV	300 (122)	155 (70)
AO	147(129)	105(72)

RV=right ventricular, LPA=left pulmonary arteries, LV=left ventricular, AO=aorta

The patient was discharged next day with stable vitals and no complication. The approval from institutional review board was taken for the publication of the case report.

DISCUSSION

Combined aortic and pulmonary valvular stenosis is a rare entity. Sometimes, this lesion is associated with either sub valvular or supra valvular stenosis which at times is difficult to deal with cardiac catheterization. Either alone or in combination, these lesions may have syndromic association like Noonan and William syndrome. Supra-valvular aortic stenosis and peripheral branch PA stenosis was a common feature in William syndrome. The prevalence of this rare combination is 0.01% of all congenital heart diseases.[7] Literatures has shown the rarity of this combination and there are only 16 case reports of combined aortic and pulmonary stenosis till date.[3-10] This number is even smaller in pediatric population and the children who were dealt with balloon valvoplasty. The first case report had been published by Horlick et al. in 1957[3] and was managed by surgical correction. The first case report with successful balloon valvoplsty in adult was published by Chandrashekhar Y et al.[6] in 1994 and in a neonate by da Cruz E et al.[9] in 2007.

The age of presentation in all 17 cases (including one of ours) ranged from neonate to 55y. Like our case, there is slight male predominance in all the case reports with male: female ratio of 1.5:1. Majority of patients got attention due to hyper-dynamic precordium and a cardiac murmur. Some cases were suspected by dyspnea and fatigue and picked up clinically after detailed precordial examination. In our case, the left ventricle impulse, well sustained heaving apex beat and lateral displacement of apex revealed the predominant LV. But in some cases, RV component was more prominent. Just like in our case, majority of children in case reports[7] had systolic thrill at both

pulmonary and aortic areas. In our case, ECG showed biventricular hypertrophy with predominance of LV. But some case reports[9] showed P-Pulmonale and prominent tall R wave in lead V1. These findings are relative and depends upon the severity of predominant lesion. Cardiomegaly on chest X ray was a universal finding in all the reported cases but with variability of ventricular predominance. The majority[7-9] had right ventricle predominance but our case had left ventricle predominance.

The combined balloon aortic and pulmonary valvoplasty had been the procedure of choice since it had been successfully performed first in an adult patient[6] and then in a neonate.[9] Morbidity and mortality can occur in any catheterization procedure and same is the case with combined valvoplasty. Usually arrhythmia is a common adverse event which happens when the wire is parked in the LV and bradycardia occurs during balloon dilatation[7] particularly in children who are already hemodynamically compromised. Mild residual stenotic lesion is left in some patients just like in our case because of thickened dysplastic valve and high initial gradient across the valve.

There is always a query in combined stenotic lesions regarding management decision that which valve should be repaired first as the procedure has to be performed in the same setting. In our case, we opened the aortic valve first followed by pulmonary valve with a reason that left sided obstruction should be relieved first before the right sided obstruction. It is supported by the fact that opening the pulmonary valve first and relieving the right side obstruction in presence of left sided obstruction may lead to increased preload on LV. This may result in fatal pulmonary edema leading to pulmonary hemorrhage which is a well-documented complication of pulmonary valvoplasty and this complication will be more obvious in presence of compromised LV.[11] This was the case in all the reported literature of combined balloon valvoplasty except a report published by Chandrashekhar Y et al.[6] where they opened the pulmonary valve first to look the hemodynamic effect of relieving right sided obstruction on the left.

Although balloon aortic and pulmonary valvoplasty is a standard of care these days but the objective of the case report in addition to its rarity, is to help in decision making that which valve to be opened first and which type of anesthesia can be used in developing countries where public sector hospitals are handicapped in many ways. The use of local anesthesia with sedation demands a high degree of monitoring during the procedure particularly while pacing at a high rate and also post procedure good ICU care is mandatory.

In conclusion, combined aortic and pulmonary valvular stenosis is a rare disease and simultaneous correction of both valves by balloon valvoplasty is a successful and safe option to deal with these stenotic lesions under the same setting by local anesthesia and strict monitoring. Early recognition, timely correction and age of the child depict the success of the procedure.

AUTHORS' CONTRIBUTION

UR: Concept and design, data acquisition, interpretation, drafting, final approval, and agree to be accountable for all aspects of the work. RN, ZF, SA: Data acquisition, interpretation, drafting, final approval and agree to be accountable for all aspects of the work.

Conflict of interest: Authors declared no conflict of interest.

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