Patent Ductus Arteriosus with Severe Aortic Stenosis with Moderate Aortic Regurgitation: Whom to Treat First

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Abstract

Patent Ductus Arteriosus (PDA) is a common Acyanotic congenital Heart disease but its association with Aortic Stenosis (AS) is rare beyond the neonatal period. Here we present a rare case of PDA associated with Aortic Stenosis with Aortic regurgitation (AR) and its treatment plan based on hemodynamic.

Keywords
Aortic Stenosis; Aortic Regurgitation; Cardiovascular Examination; Hemodynamic

Introduction

PDA is one of the common Acyanotic congenital Heart disease found in children but its association with the aortic valve disease is less frequent beyond the neonatal period [1-3]. This finding is important so that while planning for the surgical management of LVOT obstruction, PDA should not be missed. We here present a case of a patient who presented with cardiac failure and was diagnosed with PDA with severe AS and moderate AR.

Case Report

A 5 yr old male child presented with failure to thrive with congestive heart failure. On examination no e/o dysmorphism present and Heart rate was 134/min with collapsing pulse in all four limbs. Saturation in all four limbs was 95% with pulse oxymetry. Anthropometric examination showed weight below third percentile with height percentile between 25th-50th centile. Cardiovascular examination showed Hyper dynamic precordium in left 2nd intercostal area below clavicle where as in right second intercostal space ejection systolic (IV/VI grade), a grade II/IV diastolic murmur present in the right 3rd intercostal space. Respiratory examination revealed b/l wheeze in lower zone. Other systemic examination was within normal limit. On investigation Complete Blood count revealed Hemoglobin was 11.8 with total leucocytes count 11,000 and platelet count 1.6lakhs. X-ray chest s/o cardiomegaly (CT ratio >0.55) with bulging at Aortopulmonary conus suggestive of left atrial enlargement along with features of Left ventricular hypertrophy and cephalisation of pulmonary venous system. Echocardiography examination showed dilated left ventricle with turbulence across aortic valve with peak by mean gradient of 72/40mmhg across AV with AVmax of 4.2m/s with AR PHT 385. Basal short axis view showed normal aortic valve (3 Cusps) and 6mm of PDA with Left to right shunt (Figure 1-3). This was a challenging case to decide which lesion to operate first, as patient was Haemodynamically unstable. After discussion with parents about dual lesion we have decided to go ahead with staged procedure with PDA to be closed with the device as Left ventricle was much dilated as compared to hypertrophied and during cardiac catheterization there was a significant reduction of the AV gradient during the procedure itself suggesting that the increased gradient which we were getting was mostly flow related. Hence we had closed PDA with PDA device Cocoon 8/10mm (Figure 4,5). Gradually patient Haemodynamically improved. 1 week after PDA Device closure repeat echo showed significant reduction in gradient across aortic valve with peak by mean gradient of 30/15mmhg with AR PHT of 534 s/o mild AS with mild AR, with no shunt across
uncommon entity [2,4] and is cited very scarcely in literature especially after neonatal period. In one of the study reported out of 146 patient sent for PDA device closure only 3 patient had LVOT obstruction 2 being subaortic membrane and 1 with bicuspid Aortic valve [2] In aortic stenosis patients the gradient across Aortic valve is dependent on aortic valve Area and amount of flow across the aortic valve [5-7]. For treatment of severe aortic stenosis, an outstanding balloon dilation procedure may leave the patient with only mild aortic stenosis, but it may result in regurgitation. An outstanding valve replacement operation will relieve essentially all outflow obstruction, but leaves the child with an artificial valve, with need of medical management and closed follow up for aortic lesion. Follow-up echo after one month of PDA closure shows regression of LV size with gradient of 26/11mmHg across AV with trivial Aortic regurgitation (Figure 6,7).

**Discussion**

The Combination of Aortic stenosis with PDA is distinctly uncommon entity [2,4] and is cited very scarcely in literature especially after neonatal period. In one of the study reported out of 146 patient sent for PDA device closure only 3 patient had LVOT obstruction 2 being subaortic membrane and 1 with bicuspid Aortic valve [2] In aortic stenosis patients the gradient across Aortic valve is dependent on aortic valve Area and amount of flow across the aortic valve [5-7]. For treatment of severe aortic stenosis, an outstanding balloon dilation procedure may leave the patient with only mild aortic stenosis, but it may result in regurgitation. An outstanding valve replacement operation will relieve essentially all outflow obstruction, but leaves the child with an artificial valve, with need of medical management and closed follow up for aortic lesion. Follow-up echo after one month of PDA closure shows regression of LV size with gradient of 26/11mmHg across AV with trivial Aortic regurgitation (Figure 6,7).

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anticoagulation. The long-term concern is that the child will outgrow the size of the artificial valve and will require a repeat surgical valve replacement in later years. And Ross procedure consisting of replacing aortic valve with the pulmonary valve carries its own disadvantage. All these aortic valvular intervention requires a good expertise and also carries morbidity and mortality risks which adds to the hemodynamic instability in patients with dual lesion. So by correcting the lesion causing High Output state, patient gets stabilized and there is possibility of reduction in severity of Aortic Stenosis which can be medically managed as in our case after PDA device closure, gradient across aortic valve decreased significantly. Thus the severity of aortic obstruction may actually be overestimated in a large Left to right shunt cases like PDA. Thus surgical management of Aortic valvular disease can be done at a later stage if required on follow up.

Conclusion

Based on our experience in this case and through observation of previous reported studies it is advised that before planning for surgical/transcatheter guided management of LVOT obstruction treatment of anomalies leading to High output state (like PDA) should be done first.

References