

# Congenital Bicuspid Aortic Valve with Severe Aortic Stenosis and Patent Ductus Arteriosus - An Uncommon Association - Treated in Unison

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**Abstract:** *Bicuspid aortic valve, the most common congenital cardiac malformation, may occur in isolation or in association with other congenital cardiovascular malformations. Although occurrence of PDA [Patent ductus arteriosus] with bicuspid aortic valve in pediatric population is known, especially as a component of Heart hand syndrome, association of a PDA with left ventricular outflow tract obstruction is uncommon beyond neonatal period. Further, in patients presenting with this combination, either no intervention was performed for aortic valve or they were referred directly for open surgery. We report a unique case of adolescent female with bicuspid aortic valve with severe valvular aortic stenosis with PDA in which both lesions were tackled in same setting by percutaneous approach rather than referring patient directly to open surgery.*

**Keywords:** bicuspid aortic valve, valvular aortic stenosis, patent ductus arteriosus, balloon aortic valvuloplasty, PDA device closure

## 1. Introduction

Bicuspid aortic valve may occur in isolation or in association with other congenital cardiovascular malformations.[1]The Patent ductus arteriosus is usually present in pediatric patients with bicuspid aortic valve, in particular as a component of hand heart syndrome.[2]The PDA with critical left ventricular outflow tract obstruction has been observed in neonates.[3],[4] However, the association of a PDA with aortic stenosis beyond the neonatal period is infrequent.[3],[4],[5] Till date, only few cases with such a combination have been reported and were managed with either no intervention for aortic valve or surgically with aortic valve replacement and PDA ligation.[4],[6]Herewith, we report a case of adolescent female with this rare combination which we treated by percutaneous approach with balloon aortic valvuloplasty and PDA device closure, rather than directly referring patient for open surgical repair.

## 2. Case Report

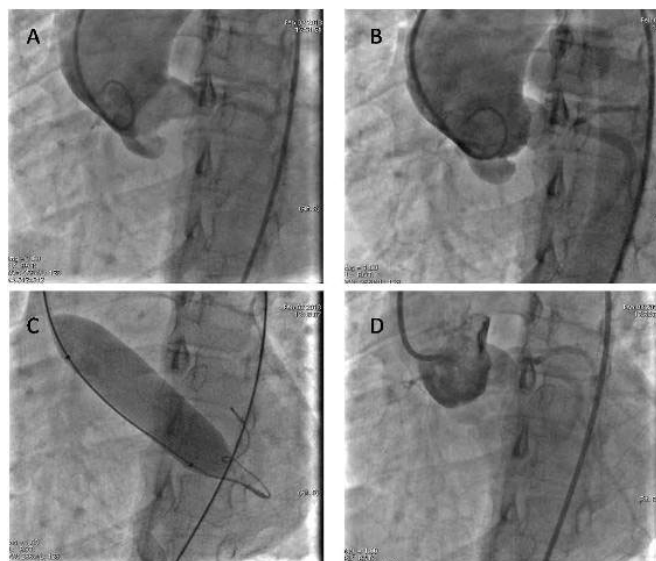
An adolescent female presented with chief complaints of recurrent episodes of angina and dyspnea on exertion with history of presyncope for a period of 6 months. No abnormal findings were noted on general examination. The apical impulse was in left fifth intercostal space at mid clavicular line and heaving in character, with systolic thrill in second right intercostal space radiating to both carotids. The auscultatory findings included normal first and second heart sounds, aortic ejection click, crescendo decrescendo ejection systolic murmur grade IV/VI in intensity, peaking in late systole at second right intercostal space and additional continuous murmur in Left second intercostal space. The chest x ray was normal. There was left ventricular hypertrophy on ECG. The 2D-echo findings included concentric left ventricular hypertrophy, bicuspid aortic valve with commissures at four and eleven o'clock

position with peak and mean gradient of 110 and 70 mmHg across aortic valve, respectively and absence of aortic regurgitation. The aortic annulus was 20mm in diameter. In addition, there was a 4 mm PDA with left to right shunt with restriction of 70 mmHg across it. With due consideration to the age of a patient, lifelong need of anticoagulation with mechanical valves, problems associated with anticoagulation and mechanical heart valves especially during pregnancy, risk of early degeneration with bioprosthetic valve in children and adolescents, issue of scar with surgery and lower rate of restenosis in congenital aortic stenosis following balloon aortic valvuloplasty, decision was taken to attempt balloon aortic valvuloplasty followed by PDA device closure, rather than referring patient directly to open surgery. With the written informed consent of the family, first balloon aortic valvuloplasty was performed using Zmed 20mm x6cm balloon. Post procedure peak gradient across aortic valve decreased from 110mmHg to 40mmHg. Grade I aortic regurgitation was noted on post procedure aortic root angiogram. (Figure 1) After successfully performing balloon aortic valvuloplasty, PDA was closed by using 8mm x 10mm amplatz ductal occluder device. The peak gradient across aortic valve further reduced to 20mmHg following PDA device closure. (Figure 2) The post procedure 2D-echo showed peak gradient of 20mmHg across aortic valve with mild aortic regurgitation. The residual shunt was noted across PDA device on immediate post procedure 2D-echo. (Figure 3) The patient was advised infective endocarditis prophylaxis and discharged subsequently.

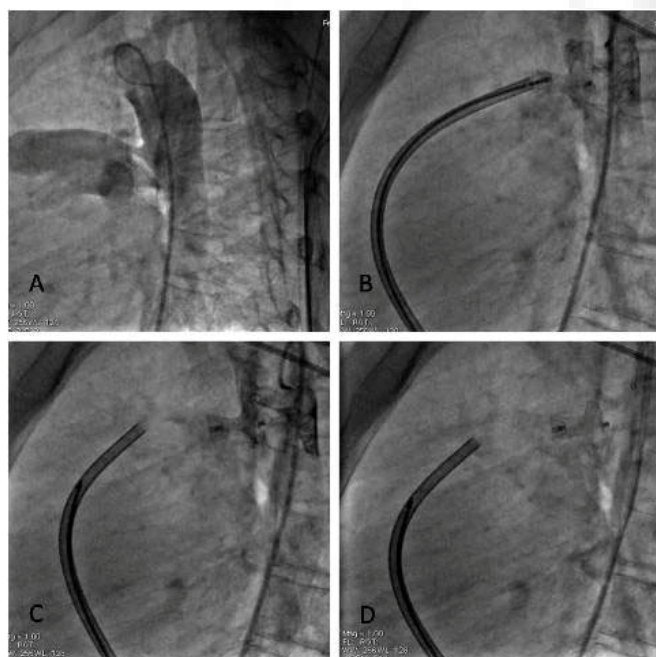
## 3. Discussion

Bicuspid aortic valve is the most common congenital cardiac malformation, affecting 1-2% of the general population with strong male preponderance.[1], [7] It may occur in association with other congenital cardiovascular

malformations such as co-arcuation of the aorta (50–80%), interruption of the aorta (36%) and ventricular septal defect (20%).[1] Duran AC et al found bicuspid aortic valve in association with other congenital heart diseases in 6.7% of total 994 specimens studied, with the aortic arch



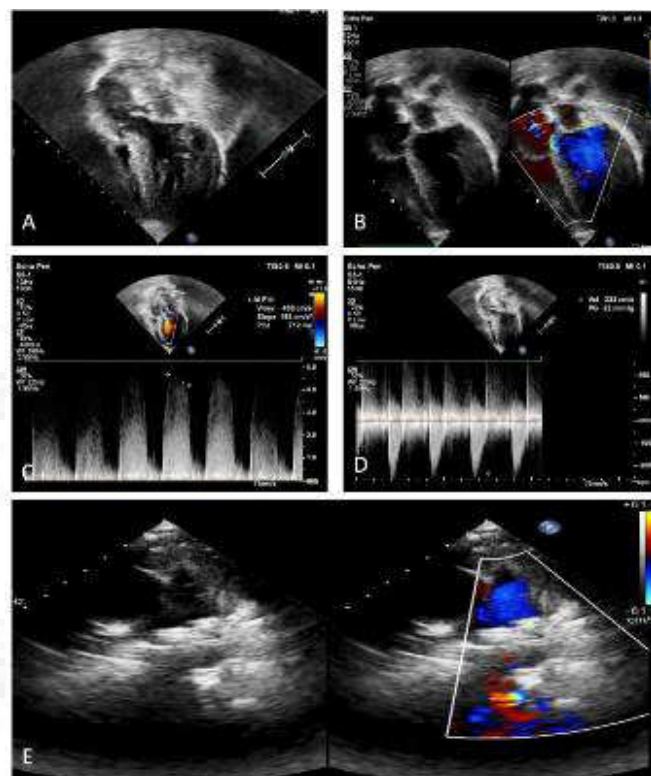
**Figure 1:** Aortic root angiogram in LAO view. A and B - Stenotic aortic valve with no aortic regurgitation, C- Inflation of Z-med balloon across stenotic aortic valve, D- Grade I aortic regurgitation following balloon aortic valvuloplasty.



**Figure 2:** Descending aorta angiogram in lateral view. A – Sizing of PDA, B - Deployment of PDA device across PDA and residual shunt across PDA device, C and D- Release of PDA device

obstruction being most common congenital heart defect(61.2%).[8] In the retrospective analysis of Mayo Clinic echocardiography database, the coexisting congenital heart disease was found in 39% cases of bicuspid aortic valve, with co-arcuation of the aorta (36%) and interrupted aortic arch(36%) being most common.[9] The right-left cusp fusion was more frequently found in patients with bicuspid

aortic valve and left-sided obstructive lesions in comparison to patients with isolated bicuspid aortic valve.[9] Patent ductus arteriosus is usually present in pediatric patients with bicuspid aortic valve and may be associated with hand anomalies. The bicuspid aortic valve with associated patent ductus arteriosus has been found in a syndrome complex, known as hand heart syndrome.[2]The association of a PDA with left ventricular outflow tract obstruction is infrequent



**Figure 3:** 2D –echo and color Doppler images. A and B – Reduction in severity of aortic stenosis and mild aortic regurgitation following balloon aortic valvuloplasty, C– peak gradient of 20mmHg across aortic valve, D - mild aortic regurgitation across aortic valve, E - PDA device and residual shunt across it.

beyond neonatal period.[3],[4],[5] One study of 185 patients referred for transcatheter closure of PDA reported three patients who had stenotic bicuspid aortic valve. All the three patients had a marked reduction in a pressure gradient across aortic valve following transcatheter closure of PDA and all the three patients were managed without intervention for aortic valve.[4] Whitlark JD et al reported additional heart defects in 20 out of 146 patients referred for transcatheter closure of PDA and only three of these 20 patients had left ventricular outflow tract abnormality, two with a sub aortic membrane and one with a bicuspid aortic valve.[6] The combination of bicuspid aortic valve with valvular aortic stenosis with PDA beyond neonatal period is rare and only nine cases of this rare combination were reported till 1994 which were treated concomitantly open surgical repair with aortic valve replacement and surgical ligation of PDA.[3],[6] Furthermore, there are sparse reports of cases which were treated by percutaneous approach and in those cases also, PDA device closure was performed first followed by balloon aortic valvuloplasty. The overestimation of severity of aortic obstruction may occur in presence of a large left to right shunt across the

PDA, as assessment of severity of aortic stenosis is based on measurement of peak to peak systolic pressure gradient, which is further dependent on aortic valve area and flow across it. [4] Thus, when faced with such rare combination of aortic stenosis and PDA, one should first opt for PDA closure. But, in our case, faced with a dilemma of possibility of failure to cross aortic valve following PDA device closure, we decided to attempt balloon aortic valvuloplasty first and if successful, then PDA device closure.

#### 4. Conclusion

Association of aortic stenosis with patent ductus arteriosus is rare beyond neonatal period. The percutaneous approach with balloon aortic valvuloplasty with PDA device closure is a feasible option while treating this rare combination before referring patient directly for open surgical repair.

#### 5. Conflicts of interest

None

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