Aortic balloon valvuloplasty in corrected transposition of the great arteries with severe valvular aortic stenosis of congenital bicuspid aortic valve

Sir,

Valvular aortic stenosis in congenital corrected transposition of great arteries is extremely rare. So far, only two cases have been reported.[1,2] The first was with congenital bicuspid valve[1] and the other with senile aortic stenosis.[2] Both of them were above 50 years of age and had undergone aortic valve replacement. There was no significant systemic atrioventricular (AV) valve regurgitation in both the cases.

We present a 10-year-old boy presented with congenital corrected transposition of great arteries (CCTGA). He also had ebstenoid malformation of morphological right AV valve with 1.9 cm apical displacement with severe tricuspid regurgitation [Figure 1], doming, and pliable severely stenosed congenital bicuspid aortic valve (BAV) with peak gradient of 78 mmHg, trivial aortic regurgitation, and fairly contracting subaortic right ventricle. Morphologically left ventricle and its outflow tract were normal. Aortic annulus measured 21 mm and valve area was 0.7 cm. Aortic root angiogram in anterior posterior view confirmed bicuspid aortic valve without calcification and inverted coronaries [Figure 2]. Two sequential dilations of aortic valve was done in tandem using two balloons of 16 mm (76% annulus diameter) followed by 18 mm (86% of aortic annulus) ATLAS® PTA Dilatation Catheter balloon [Bard Peripheral Vascular, Inc.] respectively. Balloons were dilated manually with final complete disappearance of waist by 2nd balloon. The pullback gradient across the aortic valve reduced to 5 mmHg (Systemic Ventricle pressure of 115/0-7 mmHg and ascending aorta pressure of 110/70 mmHg) without any addition of aortic regurgitation. Follow-up echocardiography at the end of one month follow up, showed reduction of left AV valve regurgitation despite ebstenoid malformation and peak gradient across aortic valve was less than 30 mmHg without any aortic valve regurgitation [Figure 3]. Aortic valve click was still audible without stenotic murmur. This patient was successfully discharged from hospital on 3rd day post procedure with appropriate medical management for left atrioventricular valve regurgitation.

Associated cardiac abnormalities are seen up to 90% or more in cases of CCTGA. The most common cardiac lesions associated with CCTGA are ventricular septal defect (VSD) occurs in 70-80%, left ventricular outflow tract obstruction has been reported in 30-60%, systemic atrioventricular valve abnormalities are observed at autopsy in up to 90% of L-TGA cases. Regurgitation is frequent and generally progressive. An ebstein-like malformation of the tricuspid valve, which is usually accompanied with right ventricular dysfunction and failure, has been reported in 20-53 percent of patients. Mitral valve abnormalities occur up to the extent of 50% of cases. Tricuspid regurgitation is significantly related

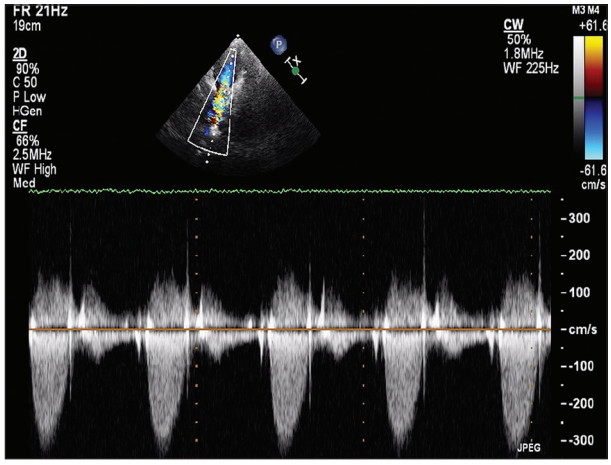

Figure 1: Echocardiography in apical 4-chamber view with color Doppler and continuous wave showed severe left AV valve regurgitation with Tricuspid regurgitation velocity up 6 meter/second

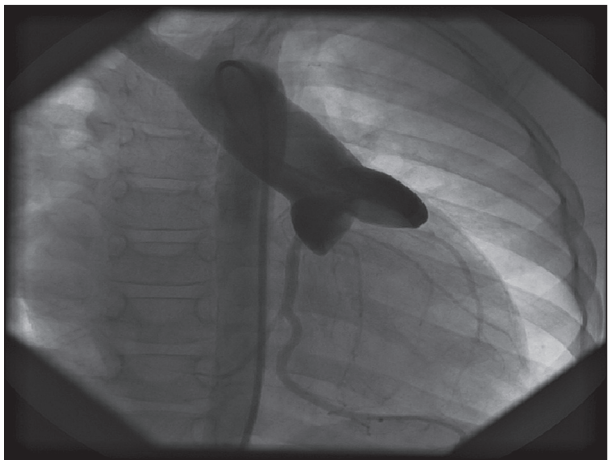

Figure 2: Aortic root angiogram in anterior posterior view showed left anterior aorta, stenosed congenital bicuspid aortic valve, inverted coronaries, and almost no aortic regurgitation

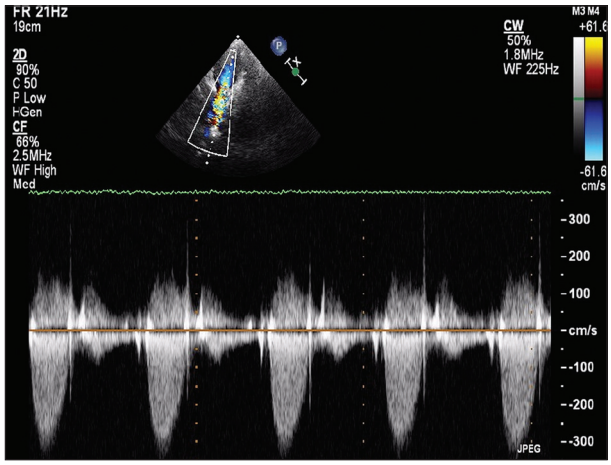

Figure 3: The interrogation of aortic valve using 2D echocardiograph with continuous wave Doppler in long axis view with a swipe to base to focuses aortic valve. The peak gradient across aortic valve is less than 30 mmHg without any aortic regurgitation after 1 month of percutaneous aortic valve dilatation. The peak gradient across aortic valve was 75-80 mmHg prior to the procedure

to mortality in both children and adults.[3] The repair of isolated systemic atrioventricular valve regurgitation with heart failure almost always responds poorly a suboptimal result of anatomic repair or replacement of tricuspid valve.[4] The combined hemodynamic impact of inappropriate ventricle on left side, aortic stenosis, and ebstenoid malformation of left AV valve is lethal as in our case. Relief of aortic valve obstruction improves the AV regurgitation. The replacement of ebstenoid AV valve is preferred over repair.[5] However, this treatment option is not suitable for a teen, considering the long term consequences. Therefore, we staged the management of this case. We hope he can be best helped by only left AV valve replacement later.

CONCLUSION

To our level of knowledge from review of contemporary literature, this case is the first case report that describes PBAV in CCTGA to relieve systemic right ventricular outflow tract obstruction and significant functional improvement in systemic AV valve regurgitation.

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