Double whammy: A case of bilateral bicuspid arterial valves in transposition, with a review of the literature

Pushpa Shivaram, Kavisha Shah, Amy Dossey, Brian Reemtsen, Robert H Anderson[1]

Department of Pediatrics, Division of Cardiology, The University of Arkansas for Medical Sciences, Little Rock, AR, USA, [1]Cardiovascular Research Center, Institute of Genetic Medicine, Newcastle University, Newcastle-Upon-Tyne, United Kingdom

ABSTRACT

Bicuspidity of both the semilunar valves is rarely reported. We report the first ever case of bilateral bicuspid semilunar valves in a case of transposition.

Keywords: Bicuspid aortic valve, bicuspid pulmonary valve, transposition

INTRODUCTION

The bicuspid aortic valve is a common congenital anomaly, having an incidence of from 1% to 2% in the general population.[1] Bicuspid pulmonary valves, in contrast, are very rare, having an incidence of only 0.1%.[2] The bicuspid aortic valve is commonly seen in association with left-sided heart disease but has rarely been reported in the setting of transposition,[3] defined on the basis of concordant atrioventricular and discordant ventriculoarterial connections. The presence of both arterial valves with two leaflets is exceedingly rare. To our knowledge, such a finding has been reported only in postmortem studies and in few isolated case reports in the setting of concordant ventriculoarterial connections. We report here a neonate with bicuspidity of both arterial valves in the setting of transposition.

CASE REPORT

A male neonate was transferred on prostaglandin E1 infusion on the 1st day of life to our facility after he failed the critical congenital heart screen. He had been born at term and weighed 2.68 kg. Physical examination revealed a patient small for gestational age, with mild dysmorphism. There was posterior sloping of forehead, a mildly depressed nasal bridge, and low-set ears. Cardiac examination revealed normal rate and rhythm. A systolic ejection murmur graded at 2 from 6 was heard at the cardiac base. All pulses were palpable and strong. Cross-sectional echocardiography revealed usual atrial arrangement with concordant atrioventricular connections, right-handed ventricular topology, and discordant ventriculoarterial connections, with the aorta positioned anterior and rightward relative to the pulmonary trunk. He was found to have both aortic and pulmonary valves with two leaflets. The pulmonary valve annulus, measured at the level of the virtual basal plane, had a diameter of 12 mm, with a *Z*-score of 2.32. The comparable value for the aortic valve was 9 mm, equating to a *Z*-score of 2.83 (Boston *Z*-score) [Figure 1 and Video 1]. The pulmonary valve was mildly thickened and domed in systole, permitting mild regurgitation but producing no stenosis. The zone of apposition between the two leaflets of the aortic valve ran from front to back, with one coronary artery arising from each of the valvar sinuses. The left coronary artery arose from the left-sided sinus, while the right coronary artery arose from the right-sided sinus. The left coronary artery then branched into the circumflex and anterior interventricular arteries. He had a tiny perimembranous ventricular septal defect,

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Address for correspondence: Dr. Pushpa Shivaram, Department of Pediatrics, Division of Cardiology, The University of Arkansas for Medical Sciences, 1 Children's Way, Mail Slot 512-3, Little Rock, AR 72202, USA. E-mail: drpushpashivaram@gmail.com

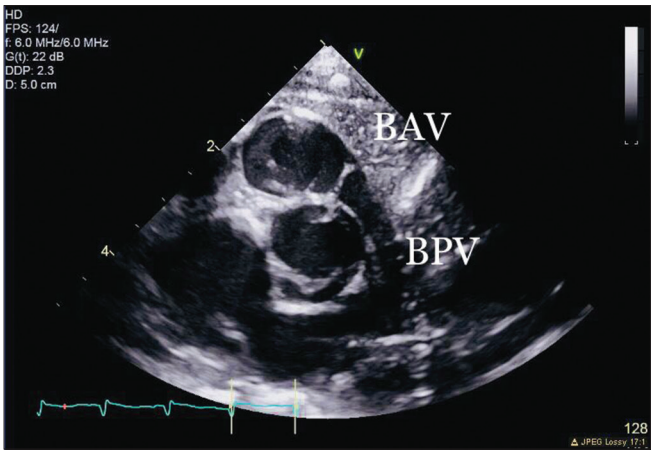

Figure 1: Bicuspid aortic and pulmonary valve: Aortic is anterior and to the right of pulmonary trunk. True bicuspidity of both the semilunar valves noted

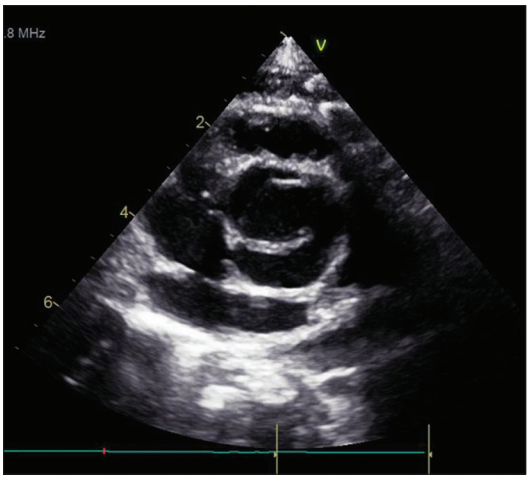

Figure 2: Postoperatively, bicuspid dilated neoaortic valve

an oval foramen of moderate size, and a large persistently patent arterial duct. He underwent emergent balloon atrial septostomy and subsequently, on the 13th day of life, underwent an arterial switch operation, with ligation of the duct and closure of atrial septal defect on day 13 of life. Postoperatively, he has developed moderate insufficiency of the new aortic valve, with dilation of the sinuses and moderate dilation of the left ventricle [Figure 2]. The bicuspid nature of both valves was confirmed intraoperatively. The zones of apposition of the valves, however, were aligned, making it possible to transfer the coronary arteries to the adjacent pulmonary valvar sinuses during the arterial switch operation.

DISCUSSION

In the early days of the arterial switch operation, a bicuspid pulmonary valve was considered a potential impediment to successful repair. With advancement in operative techniques, the switch procedure has increasingly performed in patients with bicuspid pulmonary valves. Although the bicuspid nature of both the aortic valve and the pulmonary valve have previously been described in the setting of transposition, we are unaware of any previous description of both valves being bicuspid in the same patient with transposition.

Development of arterial valves and Bicuspid semilunar valves

During the 7th week of gestation in man, the arterial valves develop in the middle component of the developing outflow tract. It is the appearance of the intercalated cushions that herald the appearance of the developing arterial roots. The central parts of the major outflow cushions fuse together during this period of development, whereas the parietal parts remain unfused. It is then the interdigitation of the unfused parietal parts with the intercalated cushions that form the primordiums of the two roots. The leaflets of the valves then excavate from the distal ends of the cushions, which become populated by cells derived from the endothelium and neural crest.[4,5] Valves with two leaflets can be the consequence either of the fusion between developing cushions[4] and hypoplasia of the intercalated cushions.[5] It is the latter mechanism that would produce leaflets having aligned zones of apposition. Various mechanisms have been proposed to account for bicuspid valves at a molecular level, such as deficiency of C-type natriuretic peptide receptor 2,[6] loss of function mutation in GATA4 gene,[7] and NOTCH1, GATA5, and SMAD6 gene.[8] Linkage analysis of the genome-wide marker has demonstrated a linkage of bicuspid aortic valve to loci on chromosome 18, 5, and 13q.[9] Bilateral bicuspid arterial valves have been noted in association with trisomy 18 and trisomy 13.[10] Our patient, however, had normal chromosomal microarray.

Review of literature

Table 1 summarizes the current literature on the existence of bicuspidity of both arterial valves. Our extensive search of literature shows this finding to be reported with an incidence of 0.01%–0.03%. Most of the cases have been encountered at postmortem or in anatomical specimens. In the few cases reported preoperatively, all patients had concordant ventriculoarterial connections. Surprisingly, however, two such neonates with concordant ventriculoarterial connections have parallel arterial trunks, as opposed to spiraling trunks. The arrangement with parallel trunks is often described as "anatomically corrected transposition."

Surgical outcomes with bicuspid valves

Bicuspid pulmonary valves are not uncommon in the setting of transposition, being reported at 4% and 7% in 2 large series.[11,12] Such bicuspid valves are known to cause complications postoperatively with the need for replacement of the new aortic root.[10] Others have shown, nonetheless, that in the short to mid-term, the arterial

Table 1: Review of literature of bilateral bicuspid semilunar valve

Authors	Study type	Description	Incidence of BBSV	Great vessel relationship	Other associations
Jashari, Van Hoeck, and Vanderkelen, 2009[13]	Retrospective observational	3861 donor hearts, 1% had abnormality of one or both outflow valve	1 donor heart (0.03%)	NRGV	Patient had dilated cardiomyopathy
Koenraadt *et al.*, 2018[14]	Retrospective observational	83 postmortem heart specimens with BAV	8 patients (10%)	NRGV	3 had trisomy 18, 1 had trisomy 13
Duran *et al.*, 1995[15]	Retrospective observational	1022 specimens of which 95 had BAV	11 patients (0.01%)	NRGV	1 had TOF, 8 had VSD, 2 had aortic arch hypoplasia
Oz *et al.*, 1995[16]	Case report	47 years old with hypertension	1 patient	NRGV	Vertically oriented bicuspid pulmonary valve with mild stenosis
Sughimoto *et al.*, 2006[17]	Case report	63 years old with BAV BPV found intraoperatively	1 patient	NRGV	Patient had ascending aorta aneurysm
Lee *et al.*, 1998[18]	Case report	Neonate 1 – ACM, perimembranous inlet to outlet VSD, pulmonary stenosis Neonate 2 – HLHS	2 patients	ACM	Neonate 1 had Goldenhar syndrome
Posada-Martínez *et al.*, 2018[19]	Case report	32 years old	1 patient	NRGV	Pulmonary artery was enlarged

BBSV: Bilateral bicuspid aortic valve, NGRV: Normally related great vessels, TOF: Tetralogy of Fallot, VSD: Ventricular septal defect, BAV: Bicuspid aortic valve, BPV: Bicuspid pulmonary valve, ACM: Anatomically corrected malposed great vessels, HLHS: Hypoplastic left heart syndrome

switch procedure can be performed with low morbidity and mortality in the setting of well-performing bicuspid neoaortic valves. To the best of our knowledge, there are no long-term data currently available. In our patient, in the short term, there is some dilation of the neoaortic sinuses, with mild-to-moderate valvar insufficiency. The presence of the bicuspid aortic valve does create potential problems in describing the sinusal origin of the coronary arteries, which is the basis of the Leiden classification. In our patient, nonetheless, the sinuses were located in the right- and left-sided locations, with each sinus giving rise to the appropriate coronary artery.

CONCLUSION

We believe that we have reported the first case of bilateral bicuspidity of the arterial valves in a patient with transposition successfully undergoing the arterial switch operation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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