BANGLADESH J CHILD HEALTH 2018; VOL 43 (2): 117-121

Case Reports

Atypical Presentation of Unicuspid Unicommissural Aortic Valve in Infant

MD. TARIQUL ISLAM[1], MOHD ZAHID HUSSAIN[2], TAHMINA KARIM[3], MD. MOSTAFIZUR RAHMAN BHUIYAN[4]

Abstract:
Unicuspid aortic valve (UAV) is a very rare congenital heart defect with an ill-defined natural history. It's very important to diagnose early as UAV has a risk of sudden cardiac death and association of other congenital anomalies. Here we present a case of unicommissural UAV with severe AS, severe LV dysfunction diagnosed by colour doppler TT echo, successfully treated with balloon valvoplasty (BAV).

Keywords: *Atypical Presentation, Aortic Valve, Unicuspid,Unicommissural*

Introduction:
This rare congenital anomaly Unicuspid aortic valve has two subtypes, acommissural and unicommissural UAV.[1] The acommissural UAV usually presents early in infancy with severe aortic stenosis and the unicommissural UAV usually presents later in adults with a less aggressive course.[2] Here we present an infant of 2 months age with unicommissural UAV who has an atypical early presentation and treated successfully by BAV.

Case report:
Momtasin a 2 months old male infant delivered by LUCS at term in a rural clinic, with birth wt of 2 kg, has history of delayed cry and resuscitation, admitted to Bangabandhu Sheikh Mujib Medical University on 15th November, 2016. He suffered from repeated attack of respiratory tract infection since birth. He was then referred to us for further evaluation. He presented to us with fever, cough and shortness of breath. On examination the baby was febrile, wheezing, and chest indrawing was present. On auscultation there was bilateral rhonchi and crepitation, there was a 5/6 systolic murmur at right upper sternal border. Examination of other system was normal. Chest X- ray showed cardiomegaly with pneumonic consolidation. Transthoracic echocardiogram showed severe AS (P max 83.3 mmHg, P mean 52.6mmHg) & severe LV dysfunction (Fig: 1). Aortic valve was unicuspid, unicommissural with a slit shape eccentric opening. LA and LV was hugely dilated. No other associated congenital heart defect was seen. Aortic annulus size was 8.11 mm (+1.36z) and aorta at the level of diaphragm was 10 mm.

After conservative management of respiratory tract infection and congestive cardiac failure we planned for balloon valvoplasty for this case. In the cath lab under general anaesthesia a root aortogram was performed showing a very narrow opening of the aortic orifice at 3 O'clock position and partially covered by the aortic valve leaflet at systole (Fig: 2).

With difficulty and multiple attempts to cross the aortic orifice we finally crossed it with a 4Fr cut pigtail with 0.018" straight exchange length terumo wire. Then we dilated the aortic orifice with a Tyshak 6 mm x 2 cm balloon catheter to its full diameter (fig:3). During this intervention we didn't paced the heart as left ventricular ejection was already compromised in this pt.

1. Associate Professor, Department of Paediatric Cardiology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Shahbag, Dhaka, Bangladesh
2. Professor, Department of Paediatric Cardiology, BSMMU, Shahbag, Dhaka, Bangladesh
3. Assistant Professor, Department of Paediatric Cardiology, BSMMU, Shahbag, Dhaka, Bangladesh
4. Medical officer, Department of Paediatric Cardiology, BSMMU, Shahbag, Dhaka, Bangladesh

Correspondence: Dr. Md. Tariqul Islam, Associate Professor, Department of Paediatric Cardiology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Shahbag, Dhaka, Bangladesh, Mobile: 01711969736; Email: tariqul125@yahoo.com
Received: 28-08-2018 **Accepted:** 24-06-2019

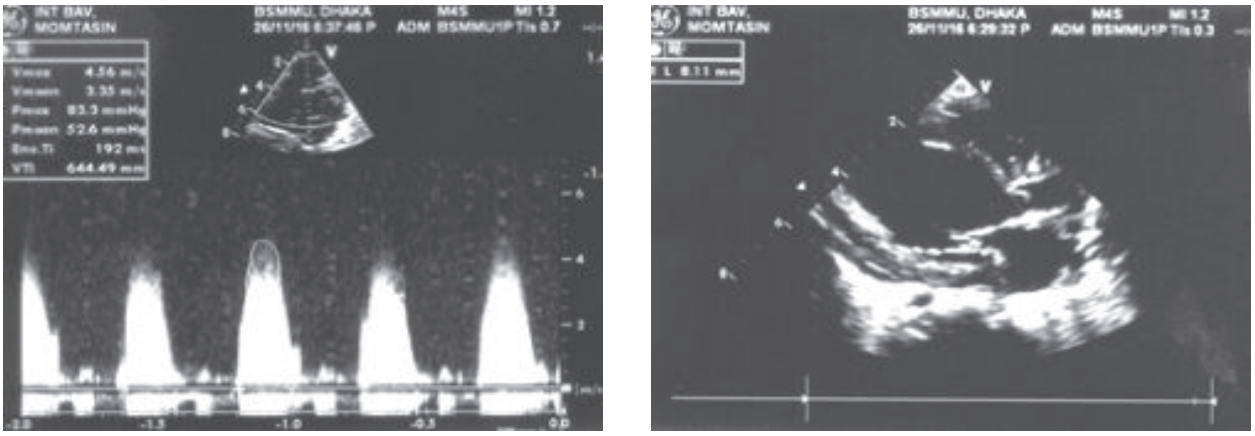

Fig.-1: *Transthoracic echocardiogram showed severe AS (P max 83.3 mmHg, P mean 52.6mmHg) & severe LV dysfunction.*

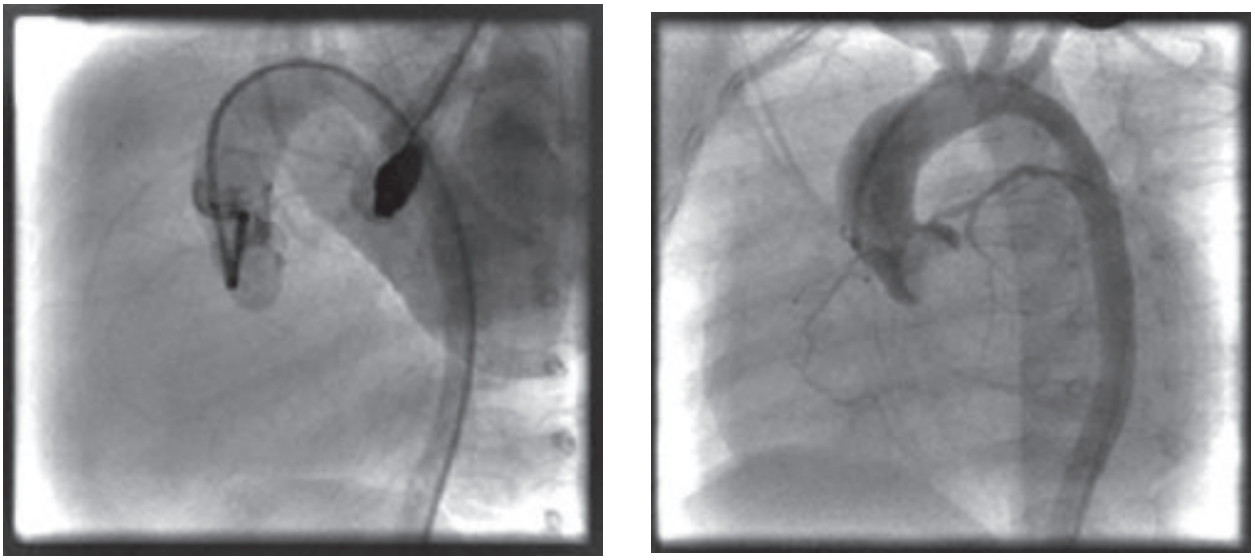

Fig.-2: *Root aortogram shows a very narrow opening of the aortic orifice at 3 O'clock position and partially covered by the aortic valve leaflet at systole.*

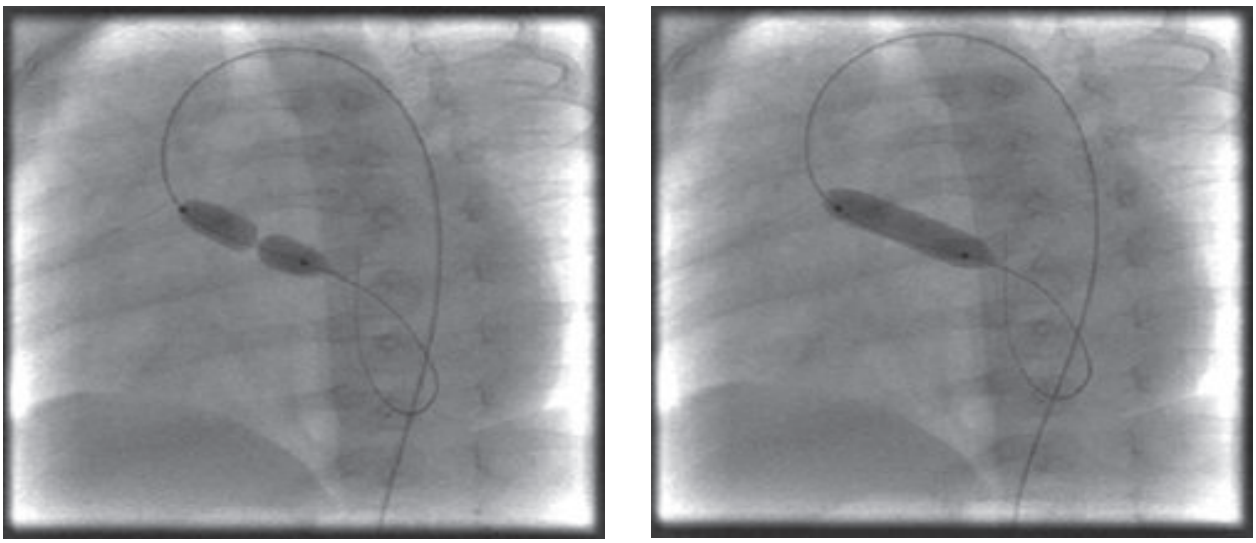

Fig.-3: *Aortic balloon valvuloplasty with a Tyshak 6 mm x 2 cm balloon catheter.*

During the process the baby's heart rate decreased from 140 to 60 beat/minute and IBP fall to 40/20mmHg. We gave i.v atropine and adrenaline. But as there was no improvement we intubated the baby with a 3.5 non-cuffed ET tube, started CPR and Adrenalin and dopamine infusion. After 5 minutes of CPR the baby's HR was stable and BP was 90/50 with adrenalin and dopamine Infusion. We put the baby on mechanical ventilator in A/C mode. He had a single episode of convulsion at 3 pm which was managed with i.v phenobarbitone only. At night we started to taper off the ionotropes infusion and the next morning we stopped all ionotropes. We successfully weaned off the baby from mechanical ventilator and extubated him at 12 noon of 2nd day of post BAV. The follow up echocardiogram showed mild AS (Pmax 38 mmHg, Pmean 24 mmHg) & mild LV dysfunction. After 7 days we discharged the baby from hospital.

Discussion:

Congenital heart disease accounts for 0.8% live birth.[3,4] Aortic stenosis accounting for 3% to 8% of these lesions making it one of the common congenital malformation.[4,5,6] The normal aortic valve consists of three valve cusps, sinuses and coaptations.[7] During cardiogenesis the bulbous cordis elongates and forms the proximal conus and distal truncus arteriosus. Two spiral truncoconal ridges, derived from neural crest mesoderm divides the bulbus cordis and truncus arteriosus into aortic and pulmonary trunks. At the same time cardiac mesoderm and cranial neural crest derivatives grow into the aortic trunk forming 3 tubercles of subendocardial tissue.[8] Hollowing out of these tubercles forms 3 equal sized valve leaflets and the sinuses of Valsalva.

Unicuspid unicommissural aortic stenosis is a very rare congenital valvular heart defect and was first reported by Edwards in 1958.[9] There are two sub-types of UAV: acommissural & unicommissural.[10] When the leaflet mesoderm grows circumferentially from annulus, aortic valve has no commissures and appears as a pinhole opening on Echocardiogram; and if it grows in with two coaptation points, there is one lateral commissural attachment to the aorta at the level of the orifice, it results in the slit-shaped unicommisural UAV[2] (fig : 4).

Developmental abnormalities of aortic valve cusps in decreasing order of frequency include bicuspid (0.9%-1.3%) then unicuspid (0.02%).[11,12] Decreasing cusp number in the congenitally abnormal aortic valve has shown increasing male predilection (3:1 to 5:1), earlier valve failure and aggressive pathological changes compared to normal tricuspid aortic valve.[13,14,15]

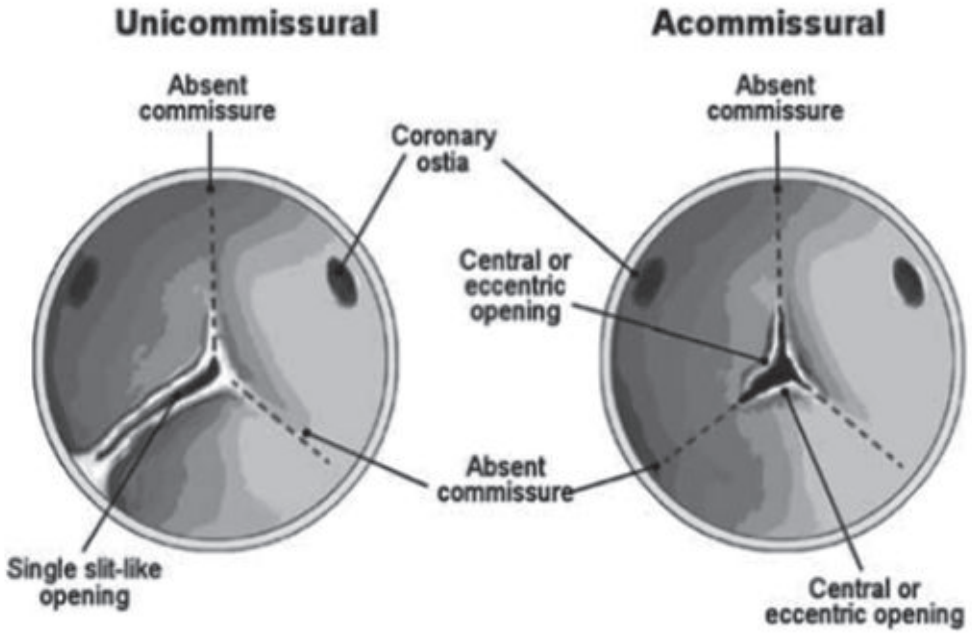

Fig.-4: *Showing unicommissural & acommissural aortic valve.*[14]

Patients with pinhole-shaped UAV present at birth or infancy with severe aortic stenosis in need of correction.[16,17] Patient with slit-shaped UAV has relatively larger orifice thus remain asymptomatic until the 3rd to 5th decade of life.[18,19] The UAV is prone to be associated with dilatation or dissection of the aorta, involving the aortic root, ascending aorta or aortic arch which typically requires surgical intervention.[20,21,22] Other associated disorder include- aortic regurgitation, aortic aneurysm, aortic dissection, patent ductus arteriosus, and the coarctation of aorta.[9,23]

Colour Doppler echocardiogram is a reliable tool for assessment of valve morphology, valve orifice, annular attachment zone and severity and also for detection of associated congenital anomalies.[24] UAV is best imaged during systole, because the absence of cusp separation during aortic valve opening reveals the eccentric "teardrop" opening in a unicommissural UAV.[19,25]

While aortic valve replacement (AVR) is common in adults, among pediatric patients balloon valvoplasty, surgical valvotomy or commissurotomy are the treatments of choice.[26,27] In pediatric patient primary surgical treatment of aortic valve disease should be favoured as balloon dilatation causes non-reversible damage to the good portion of valve leaving in place the thickened dysplastic portion.[28]

According to 2014 AHA / ACC Valvular Heart Disease Guideline our patient is in stage D1 as he has the following: congenital, symptomatic, severe high gradient AS, Vmax 4.56 m/s, Mean AP 52.6 mmHg & LV diastolic dysfunction. As per guideline The prognosis is poor once there is a peak aortic valve velocity of >4.0m per second, corresponding to a mean aortic valve gradient >40 mmHg. In Pt with low forward flow AS is severe if valve area is <1.0 cm^2. The current recommendation for symptomatic AS is IB i.e AVR is recommended with severe high-gradient AS who have symptoms by history or on exercise testing (D1) & IIbC i.e percutaneous aortic balloon dilation as a bridge to surgical or transcatheter AVR in severely symptomatic patient with severe AS.[29]

Conclusion:

Though congenital AS is common, UAV is a very rare disorder. Eccentric UAV though supposed to be of late presentation, this patient presented early as the united valve leaflets partially covering the orifice during systole obstructing the blood flow from LV. We have successfully done balloon valvuloplasty of UAV without any complication.

References:

1. Anderson RH. Understanding the structure of the unicuspid and unicommissural aortic valve. J Heart Valve Dis 2003;12 (6):670--3.
2. Brantley HP, Nekkanti R, Anderson CA, Kypson AP. Three dimensional echocardiographic features of unicuspid aortic valve stenosis correlate with surgical findings. Echocardiography 2012;29(8):E204--.7
3. Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Circulation 1971;43(3):23-32.
4. Hoffman JI, Christianson R. Congenital heart disease in a cohort of 19,502 births with long-term follow-up. Am J Cardiol 1978;42(4):641-7.
5. Serratto M, Hastreiter AR, Miller RA. Management of congenital aortic stenosis in children and young adults. Prog Cardiovasc Dis 1965;8:78-99.
6. Fukida T, Tadavarthy SM, Edwards JE. Dissecting aneurysm of aorta complicating aortic valvular stenosis. Circulation 1976; 53(1):169-75.
7. Angelini A, Ho SY, Anderson RH, et al. The morphology of the normal aortic valve as compared with the aortic valve having two leaflets. *J Thorac Cardiovasc Surg* 1989; 98:362-7.
8. Carlson BM. Human Embryology and Developmental Biology. 2nd edition. St. Louis, MO: Mosby, 1999; pp. 422-424. ISBN-10: 0815114583.
9. Mookadam F, Thota VR, Garcia-Lopez AM, et al. Unicuspid aortic valve in children: A systematic review spanning four decades. *J Heart Valve Dis* 2010; 19:678-83.
10. Lewin MB, Otto CM. The bicuspid aortic valve: Adverse outcomes from infancy to old age. *Circulation* 2005; 111:832-4.
11. Cemri M, Cengel A, Timurkaynak T. Pentacuspid aortic valve diagnosed by transoesophageal echocardiography. *Heart* 2000; 84:E9

12. Hoffman JIE, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002 Jun 19;39(12):1890-900.

13. Collins MJ, Butany J, Borger MA, Strauss BH, David TE. Implications of a congenitally abnormal valve: a study of 1025 consecutively excised aortic valves. *J Clin Pathol*. 2008 Apr; 61(4):530-6

14. Campbell M. The natural history of congenital aortic stenosis. Br Heart J 1968;30:514-26.

15. Frank S, Johnson A, Ross J. Natural history of valvular aortic stenosis. Br Heart J 1973;35:41-46.

16. Moller JH, Nakib A, Eliot RS, Edwards JE. Symptomatic congenital aortic stenosis in the first year of life. J Pediatr 1966;69(5):728--34.

17. Falcone MW, Roberts WC, Morrow AG, Perloff JK. Congenital aortic stenosis resulting from a unicommissural valve. Clinical and anatomic features in twenty-one adult patients. Circulation 1971;44(2):272--80.

18. Roberts WC, Ko JM. Frequency by decades of unicuspid, bicuspid, and tricuspid aortic valves in adults having isolated aortic valve replacement for aortic stenosis, with or without associated aortic regurgitation. Circulation 2005;111(7):920--5.

19. Brantley HP, Nekkanti R, Anderson CA, Kypson AP. Three dimensional echocardiographic features of unicuspid aortic valve stenosis correlate with surgical findings. Echo-cardiography 2012;29(8):E204--7.

20. Singh D, Chee TS. Incidental diagnosis of unicuspid aortic valve in an asymptomatic adult. J Am Soc Echocardiogr 2008;21:876.e5.

21. Roberts WC, Ko JM. Frequency by decades of unicuspid aortic valve in adults having isolated aortic valve replacement for aortic stenosis, with or without associated aortic regurgitation. Circulation 2005;111:920-5.

22. Bansal A, Arora S, Traub D, Haybron D. Unicuspid aortic valve and aortic arch aneurysm in a patient with Turner syndrome. Asian Cardiovasc Thorac Ann 2008;16:266-7.

23. Braunwald E, Goldblatt A, Aygen MM, et al. Congenital aortic stenosis. I. Clinical and hemodynamic findings in 100 patients. II. Surgical and the results of operation. Circulation 1963;27:426-462.

24. Aortic valve. Edited by Ying-Fu Chen and Chwan-Yau Luo, ISBN 978-953-307-561-7, InTech, December 12, 2011. Page 269-74.

25. Novaro GM, Mishra M, Griffin BP. Incidence and echocardiographic features of congenital unicuspid aortic valve in an adult population. J Heart Valve Dis 2003;12 (6):674--8.

26. Gatzoulis MA, Rigby ML, Shinebourne EA, Redington AN. Contemporary results of balloon valvuloplasty and surgical valvotomy for congenital aortic stenosis. Arch Dis Child 1995;73:66-69.

27. Miyamoto T, Sinzobahamvya N, Wetter J, Kallenberg R, Brecher AM, Asfour B. Twenty years experience of surgical aortic valvotomy for critical aortic stenosis in early infancy. Eur J Cardiothorac Surg 2006;30:35-40.

28. Yves d'Udekem. Aortic valve repair in children. Ann Cardiothorac Surg 2013;2(1):100-4.

29. Nishimura, RA et al. 2014 AHA/ACC Valvular Heart Disease Guideline.