Journal of Bangladesh College of Physicians and Surgeons
Vol. 31, No. 1, January 2013

A Rare Case of Aortico Left Ventricular Tunnel (ALVT)

NNF BEGUM

Summary:

Aortico-Left ventricular tunnel (ALVT) is a rare cardiac malformation with a good post operative long term outcome. The embryological basis for the disease is still unknown. Aotico-left ventricular tunnel can be diagnosed by transthoracic, transoesophageal and fetal echocardiography and by magnetic resonance imaging. A significant amount of aortic regurgitation should raise the possibility of this lesion. Here we report one of such case, which is first ever reported case of AVLT in Bangladesh.

(J Bangladesh Coll Phys Surg 2013; 31: 47-50)

Case Report:

M, a 06 months old baby girl was admitted to Combined Military Hospital Dhaka (CMH Dhaka) in May 2011 with the complaints of feeding difficulty, recurrent respiratory tract infection and failure to thrive. She was 2nd in birth order and born normally to a non-consanguineous parent. She was thoroughly evaluated, CXR showed Cardiomegaly, ECG showed left ventricular hypertrophy and Echocardiography showed an echo drop out in upper end of IVS with severe Aortic regurgitation (AR). Her finding and clinical feature were like ventricular septal defect (VSD), but no VSD was seen in echocardiography, severe AR was noticed and LVEF was 35%. Later cardiac catheterization was performed on 27th September 2011, interventricular septum (IVS) was found intact, ascending aorta was dilated. Severe aortic regurgitation (AR) was noticed and narrowing seen at commencement of ascending aorta. So there was dilemma in diagnosis. We initially diagnosed the case as aneurysm of ascending aorta and recommended Bental procedure. We discussed and reviewed this case with Prof Navin C Nanda of Alabama University,USA during his visit to Bangladesh. Later we referred her to cardiac surgeons of home and abroad. She was accepted by Dr K S Iyer of Escort heart research institute New Delhi, India. She was operated there. Her operative findings were: Aorta to left ventricular tunnel arising between the right coronary cusp and aortic wall. Right coronary ostium was not seen. A tunnel was seen opening in left ventricle below the aortic annulus. Valve was competent. Surgeons had performed Aortico-left

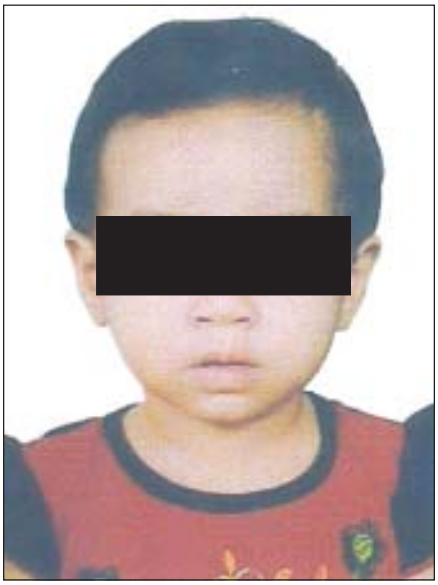

Fig.-1: *Baby M after surgery*

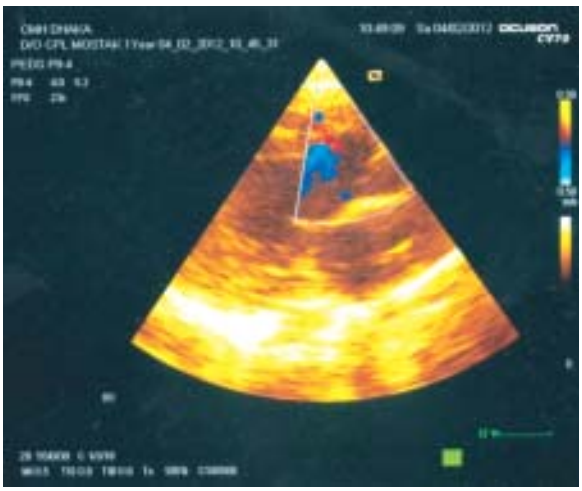

Fig.-2: *Echocardiography of the patient before surgery.*

Address of Correspondence: Colonel Nurun Nahar Fatema Begum, FCPS, Head of Pediatric Cardiology, CMH Dhaka, Cantt.

Received: 21 April, 2012 **Accepted:** 22 October, 2012

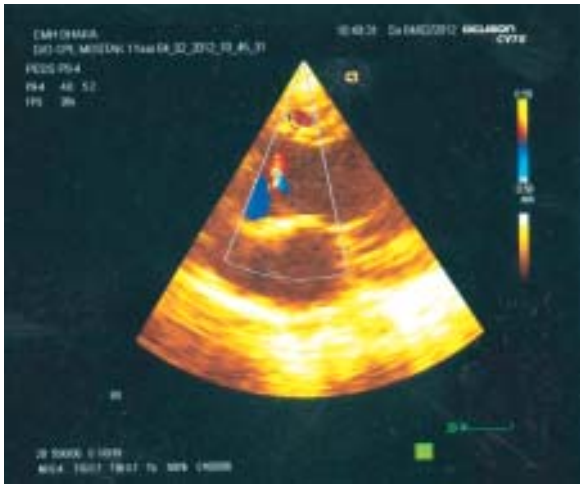

Fig.-3: *Echocardiography showing aortic regurgitation.*

ventricle tunnel repair with gluteraldehyde treated pericardial strip on 9th January 2012. They closed the opening near the valve directly.

Her post operative course was smooth but prolonged due to left ventricular dysfunction. She was electively supported with dobutamine (0-5th POD) in view of LV dysfunction (EF 35%). She had nitroglycerine infusion for high systemic pressure and milrinone infusion (0-6th POD) for persistent tachycardia (HR 160/min).

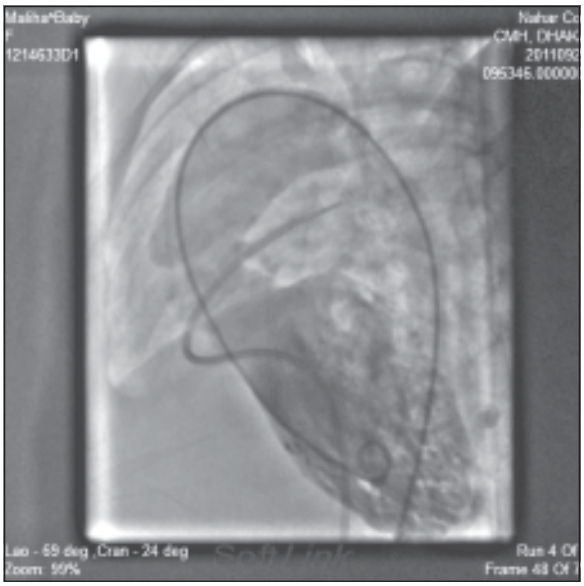

Fig.-4: *LV Graphy showing no VSD.*

She was discharged with Enalapril, Lasix and spironolactone. Digoxin was not given as there was history of ventricular ectopics on Ist and 2nd post operative day. She had fever in post operative period and managed accordingly. She was referred back to Bangladesh and got readmission to CMH Dhaka on 28.02.2012 for re-evaluation. Her echocardiography this time showed no AR, LVEF 50%, no pericardial/pleural effusion and no residual shunt seen through LV to AO tunnel. She was discharged after two days with advice to continue Enalapril, Lasix and Spironolactone till next follow up 6 weeks later.

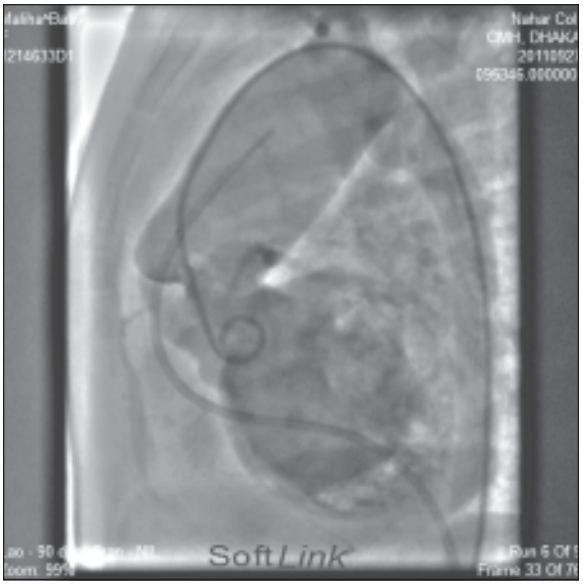

Fig.-5: *LV Graphy showing aortico left ventricular tunnel*

Discussion:

Aortico Left Ventricular Tunnel (ALVT) is a congenital, extra-cardiac channel which connects the ascending aorta above the sinutubular-junction to the cavity of the left or less commonly right ventricle. This condition presents in early childhood as aortic regurgitation and cardiac failure [1,2].

This disease was first described in an adult by hart in 1902 [3]. He described a delayed rupture of a congenital aneurysm of right sinus of valsalva to left ventricle. Levy and his associates first described the entity of the aortico-left ventricular tunnel (AVLT) in three patients [2-4]. The exact incidence of the disease is unknown, but it may range from 0.5% to less than 0.1% of congenitally malformed hearts in clinicopathological series [5]. The

embryological basis of ALVT remains uncertain. Speculations has included an anomalous coronary artery, possibly the conal vessel opening in the LV and rupture of a sinus of valsalva [6,7]. Another group said, it appears to result from a combination of maldevelopment of the cushion which give rise to the pulmonary and aortic root and abnormal septation of the structures[5].

Among 130 cases reported in the literature, more than 90% of ALVT communicated with the left ventricle. It differs from a ruptured sinus of valsalva aneurysm in having its vascular orifice in the tubular aorta rather than to a sinus of aortic valve [8,9,10]. The ostium of a coronary artery may be within an ALVT and absence of origin of both left or right coronary have been observed [10,11]. Associated lesion of the aortic valve present in 20% of the cases .[11.] Some patient develop aortic incompetence (AR)[12]. Stenosis of the pulmonary valve occur less frequently[13]. Male are predominantly affected by ALVT as found in most of the study though our case was a female [14,15]. A loud "to and fro" murmur with thrill and bounding pulse indicate rapid run off of blood. It resembles both aortic stenosis and regurgitation but in ALVT 2nd heart sound is normal[11]. Most patient develop symptom of heart failure in first year of life like our patient. The onset, severity and progression of heart failure is quite variable and ranges from many years of asymptomatic compensation to rapid decompensation and sudden death or death in utero even[16]. Prognosis of heart failure depends on association with pulmonary stenosis (PS) or aortic valve obstruction. In PS heart failure is delayed but the later group congestive heart failure with or without low cardiac out put supervenes early [17]. Echocardiography is the diagnostic investigation of choice. MRI can detect tunnels to left or right ventricle also. Cardiac catheterization is indicated only to see associated lesions or to visualize coronary artery origin [17]. Treatment is always surgical correction. Surgical closure has been recommended at the time of diagnosis, including asymptomatic patients [18,19] due to inadequacy of medical management, [20,] risk of developing severe AR [21,] and satisfactory result in neonates and infants from surgery [22]. Small ALVT in asymptomatic patients may closely be followed up because spontaneous closure may occur in some cases.

Conclusion:

AVLT has a good long term out come after surgery. The diagnosis should be considered in infants with clinical sign of AR. Echocardiography can identify it with associated lesions. Surgery should be done immediately after diagnosis in symptomatic patients. All patients require life long follow up for recurrence of the tunnel, aortic valve incompetence, left ventricular function and aneurismal enlargement of the ascending aorta.

References:

1. Souse-Uva M, Touchot A, Fermont L, Pet D, Delezoide AL, Serray A et al. Aortico Left ventricular tunnel in fetuses and infants. Ann Thorac Surgery 1996; 61:1805-1810.
2. Okoroma EO, Perry LW, Scott LP, McClenathan JE. Aortico left ventricular tunnel:clinical profile, diagnostic features and surgical considerations. J thorac cardiovasc surg 1976;71: 238-244.
3. Edward JE. Atlas of acquired diseases of the heart and great vessels, volume 3. Philadelphia WB Saunders, 1961:p 1142.
4. Levy Mj, Lillehei CW, Anderson RC, Amplatz K, Edwards JE. Aortico left ventricular tunnel. Circulation 1965; 27:841.
5. Roxane Mckay. Aortico-ventricular tunnel. Orphanet Journal of rare diseases 2007; 2:41.
6. Martins JD, Megan C, Mayer JE, keane JF. Aortico-left ventricular tunnel: 35 years experience. J Am col cardiol 2004:44:446-450.
7. Turley K, Silveman NH, Teitel D, Mavroudis C, Snider , RudolphA. Repair of aortic-left ventricular tunnel in the neonate: Surgical, anatomic and echocardiographic considerations. Circulation 1982;5:1015-1020.
8. McKay R, Anderson RH, Cook AC: The aorto-ventricular tunnels. Cardiol Young 2002, 12:563-80. PubMed Abstract. Publisher Full Text.
9. Cook AC Fagg NKL, Ho SY, Groves AMM, Sharland GK, Anderson RH, Allen LD: Echocardiographic-anotomical correlations in aorto-left ventricular tunnel. Br Heart J 1995, 74:443-8.
10. Saylam A, Tuncail T, Ikizier C, Aytac A: Aorto-right ventricular tunnel. A new concept in congenital cardia malformations. Ann Thorac Surg 1974, 18:634-7.
11. Bonnet D, Bonhoefer P, Sidi D, Kachaner J, Acar P, Villain E, Vouhe PR: Surgical angioplasty of the main coronary arteries in children. J Thorc Cardivasc Surg 1999, 117:352-7.
12. Jureidini SB, de Mello D, Norui S, Kanter K: Aortico-right ventricular tunnel and clinical pulmonary stenosis diagnosis by two-dimensinnal and Donnler echocardiography and angiography. Pediatr Cardiol 1996, 49:921-4.
13. MarUn Jimenez J, Gonzales Diegues CC, Quero Jimenez C, Rico Gomez F, Bermudez Canete R, Quero Jimenez M.

Aortico-left ventricular tunnel associated with pulmonary valve stenosis. Rev esp Cardiol 1992, 49:921-4.

14. Hovaguimian H, Cobanoglu A, Starr A. Aortico-left ventricular tunnel: A clinical review and new surgical classification. Ann Throrac Surg 1988; 45:106-112.

15. Levy MJ, Schachber A, Blieden LC. Aortico-left ventricular tunnel: Collective review. J Thorac Caediovasc Surg 1982;84:102-109.

16. Sousa-Uva M, Touchot A, Fermont l, Piot D, Delezoide AL, Serraf A, Lacour-Gayet F, Roussin R, Bruniauix J, Planche C: Aortico-left ventricular tunnel in fetuses and infants. Ann thorac Surg 1996, 61:1805-10.

17. Hiuda J, Hazekamp MG, Subolka-Plojhar MA, Ollenkamp J, repair of aorto-rihgt ventricular tunnel with pulmonary stenosis and an anomalous origin of the left coronary artery. Eur J Cardiothorac Surg 2002, 21:1123-5.

18. Akalin H, Erol C, Oral D, Corapcioglu T, Ucanok K, Ozyurda o, Ulusoy V: Aortico-left ventricular tunnel: successful diagnostic and surgical approach to the oldest in the literature. J thorac Cardiovasc Surg 1989, 97;804-5.

19. Meldrum-Hanna W, Scohroff R Ross DN. Aortico-left ventricular tunnel: Late follow-up. Ann thorac Surg 1986; 42:304-306.

20. Chen YF, Chiu CC, Wu JR Correction of aortic-left ventricular tunnel: A rare differential diagnosis of congenital aortic insufficiency. Zaitshrift fur Kardiologie 2002;91:269-273.

21. Serino W, Andrade JL, Ross D, de Leval M, Somerville J, Aorto-left ventricular communication after closure: Late post-operative problems. Br Heart J 1983; 49:501-506.

22. Edwards JE. Aortico-left ventricular tunnel: The case for early treatment. Chest 1976;70:5-6.