ISSN: 2230-9926

Available online at http://www.journalijdr.com

IJDR

International Journal of Development Research
Vol. 08, Issue, 01, pp.18253-18256, January, 2018

ORIGINAL RESEARCH ARTICLE OPEN ACCESS

SURGICAL TREATMENT OF AORTIC-LEFT VENTRICULAR FISTULA

[1,*]Zeraatian, S., [1]Jalilifar, [3]Sezavar, [3]Hajsadeghi, [1,2]Pazooki, D.

[1]Iran University, Hazrat Rasol Akrm Hospital, Department of Cardiothoracic and vascular Surgery Tehran Iran
[2]Sahlgrenska University Hospital Department of Surgery, S-413 45 Gothenburg Sweden
[3]Iran University, Hazrat Rasol Akrm Hospital, Department of cardiology, Tehran Iran

ARTICLE INFO

Article History:
Received 28th October, 2017
Received in revised form
17th November, 2017
Accepted 20th December, 2017
Published online 31st January, 2018

Key Words:

Aortic-left ventricular fistula,
Aortico Left Ventricular Tunnel (ALVT),
Cardiac malformation.

ABSTRACT

Aortico Left Ventricular Tunnel (ALVT) is a rare congenital anomaly. It presents as an asymptomatic murmur to severe Aortic Regurgitation (AR) and heart failure. Most of the patients have heart failure in first year of life.

Method: We present a review of the relevant literature and 20 recent articles with case presentations. Patient rarely present beyond second decade of life. Here we report a 40-year male with previous MI, angina pectoris, he presented with dyspnea on minimal exertion and chest pain for 1 month, and unusual type of ALVT, who presented with heart failure and conduction disturbances.

Discussion: The differential diagnosis of aortico-Left ventricular tunnel includes lesions which produce prominent to-and from murmurs. In the neonatal period, the first three of these conditions are difficult to differentiate by noninvasive means.

Conclusions: Aortico–left ventricular tunnel is a rare cardiac malformation with a good long-term outcome after surgery. Echocardiography can identify the ALVT and associated lesions and is the diagnostic investigation of choice. Diagnosis of ALVT is an indication for operation. Conservative management may be considered in rare, asymptomatic, small AVLT as observed spontaneous closure in one such patient.

Citation: Zeraatian, S., Jalilifar, Sezavar, Hajsadeghi, Pazooki, D., 2018. "Surgical treatment of aortic-left ventricular fistula", *International Journal of Development Research*, 8, (01), 18253-18256.

INTRODUCTION

The aorta-left ventricle tunnel (Ao-LVT) is considered to be a rare congenital disease (Aortico-Left Ventricular Tunnel, 1986; Levy, 1982; Ho, 1998), in which there is an abnormal connection between the left ventricle and the aorta, that is, a connection bypassing the aortic valve the anomaly is a communicating channel between the ascending aorta and the left ventricle; it was first described in 1961, by Edwards1 who considered it an acquired lesion. In 1963, Levy and associates" recognized the congenital origin of the defect. Forty-seven cases of this rare lesion have been reported in the literature, and many were initially misdiagnosed when standard invasive (catheterization) and noninvasive techniques were used (Aortico-Left Ventricular Tunnel, 1986). Generally, this condition results in severe aortic regurgitation, rapidly evolving to cardiac decompensation and death (Levy, 1982; Ho, 1998).

***Corresponding author:* Zeraatian, S.,**
Iran University, Hazrat Rasol Akrm Hospital, Department of Cardiothoracic and vascular Surgery Tehran Iran.

This congenital heart lesion in which the left ventricle communicates directly with the aorta by means of a paravalvular "tunnel and results in severe aortic insufficiency in infancy. The tunnel is usually aneurysmal in form, leaves the left ventricular cavity via a slit-like orifice, and ascends in the ventricular septumto open into the aortic root above or between the coronary ostia. Flow through the tunnel is forward in systole and retrograde from aorta to left ventricle in diastole (Michael Nichols, 1976).

Etiology

The embryological basis for ALVT remains unknown. Speculation has included an anomalous coronary artery, possibly the conal vessel, opening in the LV (Levy, 1963) and rupture of a sinus of Valsalva aneurysm (Turley, 1982). However, we identified a normal conal vessel in one patient, and sinus rupture generally occurs inferior to the RCA ostium (Levy, 1963; Ho, 1998).

Other theories included an anterior aortic wall abnormality with communication into the LV (Hovaguimian, 1988), defective incorporation of the distal end of the bulbus cordis (Somerville, 1974), and persistence of embryonic crests of the fifth aortic arch (Palacio, 1964). The etiology of aorto-ventricular tunnel is uncertain. It appears to result from a combination of mal development of the cushions which give rise to the pulmonary and aortic roots, and abnormal separation of these structures (Bernanke, 2002). Such patients have severe AR and present with heart failure. Symptoms can present at any time but most patients became symptomatic in the first year of life. Patient rarely present beyond the second decade of life. Our patient had AR and conduction disturbances most probably due to compression of conducting system by the tunnel. Echo is the diagnostic investigation of choice (Sousa-Ova et al., 1996). Echo is the diagnostic investigation of choice. This can demonstrate aortic and ventricular openings and the tunnel. Color Doppler can show the flow pattern. Cardiac catheterization is required for associated lesions and to know coronary artery anatomy. Surgical correction should be undertaken without delay, even in asymptomatic patients to prevent left ventricular dysfunction. If possible both ends of the tunnel should be closed. The ventricular end can be closed by pledgeted sutures or with a pericardial or polyester patch. The aortic end can be closed with a composite or pericardial patch (Horvath et al., 1991). Associated lesions of aortic valve should also be treated as indicated.

Clinical/diagnosis

Aortico–left ventricular tunnel is extremely rare (0.001% patients with congenital heart disease (Okoroma et al., 1976). Jose D. Martins et al, find9 patients with AVLT among 17,381 patients (0.0005%) catheterized in 35 years at his institution in Boston, Massachusetts 2004. He found a male predominance often early presentation with CHF (Hageman et al., 1988; Levy, 1982; Sousa-Uva, 1996), and a significant incidence (45%) of associated cardiac defects ((Hageman et al., 1988; Levy, 1982; Chen et al., 1994; Reitz et al., 2002). All Jose D. Martins patients with associated defects had AoV (27%) and/or coronary artery (27%) anomalies.

Differential diagnosis

Aorto-ventricular tunnel must be distinguished from other lesions which cause rapid run-off of blood from the aorta and produce cardiac failure. These include sinus of Valsalva fistula, common arterial trunk with valvar regurgitation, aorto-pulmonary window, ventricular septal defect with aortic regurgitation, persistent patency of the arterial duct, coronary-cameral fistula, valvar aortic stenosis and regurgitation, and cerebral arterio-venous malformation. Because of its "to-and-fro murmur", tetralogy of Fallot with absent pulmonary valve can also mimic aorto-ventricular tunnel with associated right ventricular outflow obstruction.

Case report

Clinical and laboratory presentation

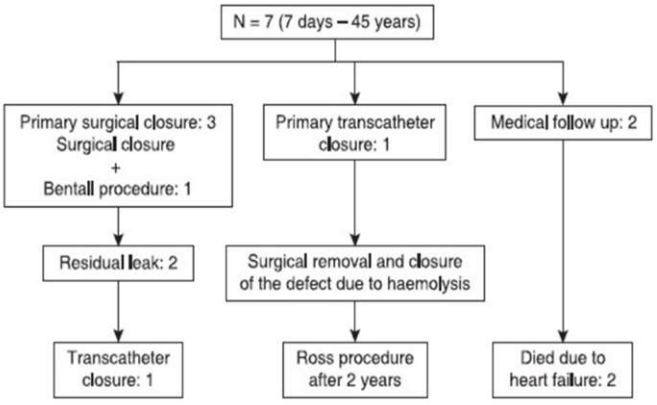
A 40 year male with previous MI, angina pectoris, he presented with dyspnea on minimal exertion and chest pain since 1 month, andunusual type of ALVT, who presented with heart failure and conduction disturbances.

Electrocardiogram showed bradycardia (Rate of around 40/min) and Complete heart block with wide QRS Complex with some ectopics and LV hypertrophy. X-ray chest revealed Cardiomegaly. X-ray chest revealed Cardiomegaly. In the operating room, after institution of routine invasive and non-invasive monitoring and induction of anesthesia, midline sternotomy was performed. After systemic heparinisation, aortic and two stage venous cannulation was achieved. Following establishment of cardiopulmonary bypass, aorta was cross clamped and cardiac arrest was achieved via retrograde and direct antegrade cardioplegia. Moderate hypothermia was maintained on bypass.

Pallavi Kathare et al, had a retrospective study from a single center from June 2004 to August 2013. They Records of all cases with a diagnosis of ALVT were reviewed and various parameters were analyzed. All patients with the diagnosis of ALVT were included. Complete clinical examination, electrocardiogram (ECG), chest X-ray and transthoracic echocardiogram (TTE) were available for all patients. Various parameters including

With a report of diagnosis and management of aorto-left ventricular tunnel (ALVT) over a period of 11 years from a single institution. Seven patients (age range: 7 days-45 years) presented with heart failure.by Pallavi Kathare et al, The diagnosis of ALVT was made by transthoracic echocardiogram in all cases. As in our case report ,Pallavi Kathare et al had one patient in 45 year of age (Michael Nichols, 1976)

Surgical management of PallaviKathare et al The surgery was performed under standard cardiopulmonary bypass (CPB) technique using right atrial cannulation and moderate hypothermia. The tunnel was visualized as an external bulge on the aorta at right sinus of Valsalva. The aorta was opened and the entire tunnel was inspected. Aorta was dilated and a varying degree of aortopathy was noted in most of the cases.
Both the aortic and LV openings of the tunnel were identified. The oval-shaped aortic opening was seen extending from the right side of sino-tubular junction to LV opening through interventricular septum. Some degree of aortic valve deformation and sagging of right coronary cusp was observed in all cases (Michael Nichols, 1976).



Pallavi Kathare et al, Flow chart of patients with aortico-left ventricular tunnel showing the various modalities of treatment 20 Ann Pediatr Cardiol. 2015 May-Aug; 8(2): 103–107.

Fig. 1. Patients with aortico-left ventricular tunnel showing the various modalities of treatment

Case no.	Age	Sex	Weight (kgs)	Morphological type	Tunnel size/ BSA (mm)	Relation to coronary	LVIDd 'Z'	EF (%)	Asc. Ao 'Z'	AR grade
1	7 days	M	3	4	7	Above RCA	4.1	25	5	Mild
2	10 days	M	2.8	2	11	Above RCA	3.8	30	4.5	Mild
3	10 yrs	F	28	2	14	Above NCC	5.5	25	4	Mild
4	11 yrs	M	30	3	20	Above RCA	5.9	30	3.5	Severe
5	12 yrs	M	35	2	16	Above RCA	5.0	34	3	Mild
6	16 yrs	M	46	2	20	Above RCA	5.1	20	6	Moderate
7	45 yrs	F	61	2	6	Above RCA	3.0	40	4	Severe

Pallavi Kathare et al Baseline characteristics of the study population (RCA: right coronary artery, NCC: Non-coronary cusp, AR: Aortic regurgitation)21Ann Pediatr Cardiol. 2015 May-Aug; 8(2): 103–107.

Fig 2. characteristics of the study population by Pallavi Kathare et al

DISCUSSION

The differential diagnosis of aortico-Left ventricular tunnel includes lesions which produce prominent to-and from murmurs. Thus, ruptured sinus of Valsalva, coronary arteriovenous fistula, ventricular septal defect with aortic insufficiency, and absent pulmonary valve should all be considered. Aorto-ventricular tunnel is a congenital extra cardiac channel that connects the ascending aorta above the sin tubular junction to the cavity of left ventricle or (less commonly) right ventricle. In the neonatal period, the first three of these conditions are difficult to differentiate by noninvasive means. Even at cardiac catheterization the distinction is not readily apparent. The left ventriculogram of aortic left ventricular tunnel closely resembles an aneurysm of the membranous ventricular septum; however, the distinction is clear when the supravalvular injection demonstrates paravalvular continuity of the aorta and left ventricle.

Medical management: is only for control of heart failure in neonates and while awaiting surgery. Without surgical treatment, most patients die early in life due to congestive heart failure as noted in two of our patients. Therefore, medical management should be of limited duration and should be only to prepare the patient for surgery (Michael Nichols, 1976).

Conclusion

Aortico–left ventricular tunnel is a rare cardiac malformation with a good long-term outcome after surgery. ALVT is a very rare congenital anomaly that should be distinguished from other lesions such as aortic regurgitation and sinus of Valsalva fistula. Echocardiography can identify the ALVT and associated lesions and is the diagnostic investigation of choice. Diagnosis of ALVT is an indication for operation. Surgery should be performed as early as possible to minimize damage to LV from chronic volume overload imposed by Aortic Regurgitation. Conservative management may be considered in rare, asymptomatic, small AVLT as observed spontaneous closure in one such patient.

Disclosure: The authors declare no conflicts of interest.

Acknowledgment: Many thanks to Z Hosseini PhD student, for checking and statistics.

The middle figure shows a cross-sectional view at the approximate level of the aortic sinotubular junction. The tunnel passes from the ascending aorta into the tissue plane between the aortic and pulmonary roots. (a') is a longitudinal section across the left ventricular outflow, through the left and right coronary sinuses of Valsalva (plane "a" of the central figure). In this example, the aortic end of the tunnel lies above the ostium of the right coronary artery, while the ventricular end is found within the intercoronary, interleaflet triangle. The position of the aortic opening is variable and may be found anywhere above the left or right coronary sinus, or the intervening commissure. (b') depicts a longitudinal section crossing the noncoronary and right coronary aortic sinuses (line "b" in the central figure). Because the pulmonary valve lies distal to the aortic valve, the tunnel may displace the free-standing, muscular, subpulmonary infundibulum enroute to the left ventricular cavity. It does not, however, pass through any ventricular myocardium.

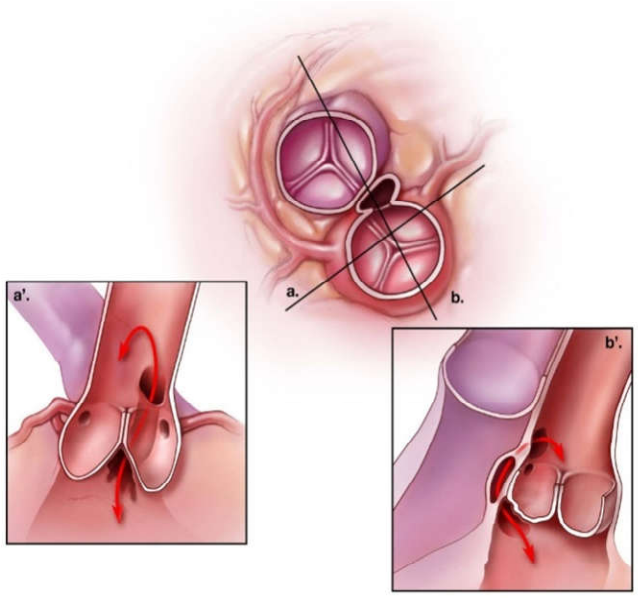


Figure 3

Schematic representation of the most common type of aorto-left ventricular tunnel.

Picture From:*Orphanet Journal of Rare*

Diseases 2007**2**:41https://doi.org/10.1186/1750-1172-2-41

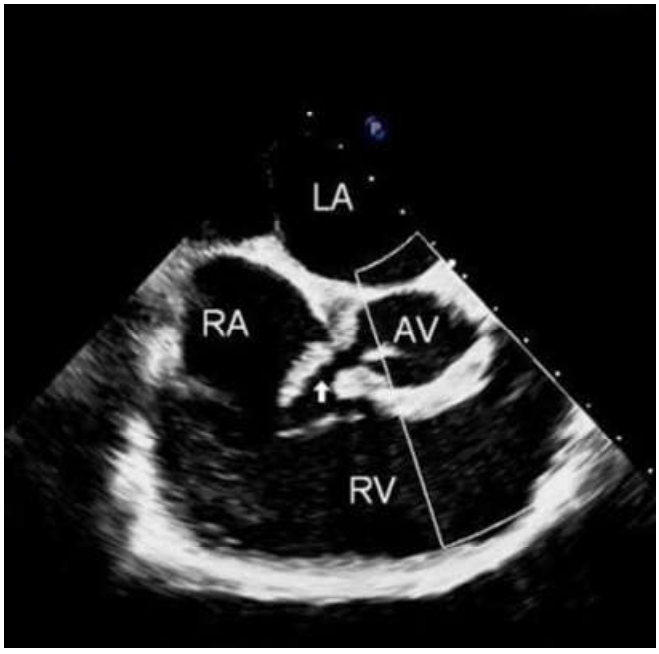


Fig: 4

LA: left atrium, AV: Aortic Valve, RV: Right ventricle, RA: Right atrium; the arrow shows an aorto-RA fistula from the perforation between the aorta and the RA.

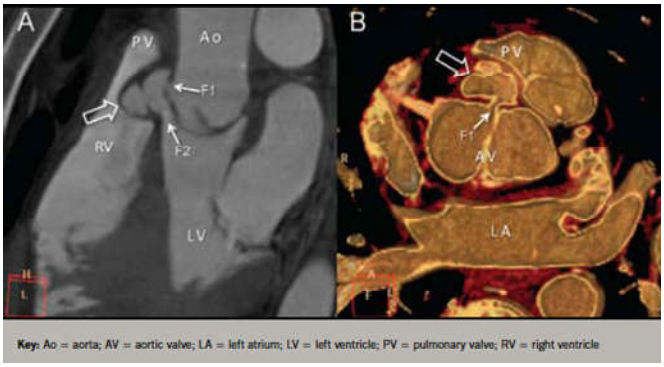


Fig. 5.

Cardiac computed tomography (CT) multi-planar reconstruction. The abscess cavity (hollow arrow) communicates with the aortic root (F1) and left ventricular outflow tract (F2) to form a fistula. The abscess presses forward into the right ventricular outflow tract displacing the pulmonary valve (PV) superiorly. B: Cardiac CT blood pool inversion image. The bicuspid aortic valve (AV) is clearly seen with a fistulous connection (F1) to the abscess cavity (hollow arrow) from: *March 2012Br J Cardiol2012;19:46–7doi:10.5837/bjc.2012.010*

REFERENCES

Aortico-Left Ventricular Tunnel:Richard A. Humes, M.D, Donald J. Hagler, M.D. JAY M. Levy, M.D. Robert H. Feldt, M.D., Hartzell V. Schaff, M.D 1986. *Mayo Clin Proc* 61:901-907.

Bernanke DH, Valley JM. 2002. Development of the coronary blood supply: changing concepts and current ideas. *Anat Rec.,* 269:198–208.

Chen YF, Chiu CC, Wu JR. 1994. Correction of aortico-left ventricular tunnel in a small Oriental infant: a brief clinical review. *J Cardiovasc Surg.,* 35:71–3.

Diagnosis and management of aorto-left ventricular tunnel, Pallavi Kathare, Rama G Subramanyam, Tapan Kumar Dash, Kalyana Sundaram Muthuswamy, K Raghu, and Nageswara Rao Koneti

Michael Nichols, G., M.D., O O Martin H. Lees, M.D., Dale P. Henken, M.D., Cecille O. Sunderland, M.D.;§ and Albert Starr, M.D.II CHEST, 70: 1, JUlV, 1976

G. *Michael Nichols,* M.D.; oO*Martin* H. *Lees,* M.D.; t *Dale* P. *Henken, M.D.! Cecille* o. *Sunderland, M.D.;§ and Albert Starr,* M.D.II CHEST, 70: 1, JUlV, 1976

Hageman H, Cobanoglu A, Starr A. 1988. Aortico-left ventricular tunnel: a clinical review and new surgical classification. *Ann Thorac Surg*., 45:106–12.

Ho SY, Muriago M, Cook AC, Thiene G, Anderson RH. 1998. Surgical anatomy of aorto-left ventricular tunnel. *Ann Thorac Surg.,* 65: 509-514.

Ho SY, Muriago M, Cook AC, Thiene G, Anderson RH. 1998. Surgical anatomy of aorto-left ventricular tunnel. *Ann Thorac Surg.,* 65:509–14.

Horvath P, Balaji S, Skoveranek S, Hucin B, de leval MR, Stark J. 1991. Surgical treatment of aortico-left ventricular tunnel. *Eur J Cardiothorac Surg.,* 5:113–16.

Hovaguimian H, Cobanoglu A, Starr A. 1988. Aortico-left ventricular tunnel: a clinical review and new surgical classification. *Ann Thorac Surg*., 45:106–12.

Levy MJ, Lillehei CW, Anderson RC, Amplatz K, Edwards JE. 1963. Aortico-left ventricular tunnel. Circulation., 27:841–53.

Levy MJ, Schachner A, Blieden LC. 1982. Aortico-left ventricular tunnel: collective review. *J Thorac Cardiovasc Surg.,* 84:102–9.

Levy MJ, Schachner A, Blieden LC. 1982. Aortico-left ventricular tunnel: collective review. *Journal of Thoracic and Cardiovascular Surgery,* vol 84, 102-109.

Okoroma EO, Perry LW, Scott LP, McClenathan JE. 1976. Aortico-left ventricular tunnel: clinical profile, diagnostic features and surgical considerations. *J Thorac Cardiovasc Surg.,* 71:238–44.

Palacio J, Peralta A, Sanchez B, Alperovich M. 1964. Intrapericardial congenital supravalvular aortic aneurysm communicating with the outflow tract of the left ventricle. *J Cardiovasc Surg.,* 5:401–4.

Reitz M, Heusch A, Paulus-Koschik A, Schmidt KG. 2002. Aortico-left ventriucular tunnel: a rare differential diagnosis of congenital aortic insufficiency. *Zeitschrift fur Kardiologie.,* 91:269–73.

Somerville J, English T, Ross DN. 1974. Aorto-left ventricular tunnel: clinical features and surgical management. *Br Heart J.,* 36:321–8.

Sousa-Ova M, Touchot A, Fermont L, Piot D, Delezoide AL, Serra A, Lacour-Gayet F, Rosin R, Bruniaux J, Planché C. 1996. Aortico-left ventricular tunnel in fetuses and infants. *Ann Thorac Surg*., 61:1805-1810.

Sousa-Uva M, Touchout A, Fermont L, et al. 1996. Aortico-left ventricular tunnel in fetuses and infants. *Ann Thorac Surg.*, 61:1805–10.

Turley K, Silverman NH, Title D, Mavroudis C, Snider R, Rudolph A. 1982. Repair of aortico-left ventricular tunnel in the neonate: surgical, anatomic and echocardiographic considerations. Circulation, 5: 1015–20.

****