

# INFERIOR WALL DIVERTICULUM OF LEFT VENTRICLE COEXISTING WITH MENTAL RETARDATION AND ATRIAL SEPTAL DEFECT

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## Abstract

We report a case of congenital inferior wall left ventricular diverticulum (LVD), atrial septal defect and mental retardation detected by intraoperative transesophageal echocardiography. The combination of three features strongly suggests that genetic factors play important role in the pathogenesis of the disorder. Most LVDs are asymptomatic. Echocardiographers and cardiac anesthesiologists should be aware of this anomaly, and include it in the differential diagnosis of abnormally shaped ventricular wall and seek other congenital abnormalities if LVD is detected.

Key words: Cardiovascular, Left ventricular diverticulum, Atrial septal defect, Mental retardation

Ventricular diverticula are rare congenital cardiac malformations<sup>1,2,3</sup>, especially in the adult population, even though there are significantly more reports in the literatures from the last two decades mainly due to improved technology in establishing the diagnosis. The majorities of these diverticula arise from the apex of the left ventricle and are usually diagnosed in early childhood<sup>4</sup>. Seventy percent of patients with left ventricular diverticulum (LVD) have associated midline thoracoabdominal defects<sup>5</sup>. LVD coexisting with atrial septal defect (ASD) has been reported<sup>6,7</sup>. However LVD coexisting with mental retardation plus ASD has never been reported. Here we report a case that LVD is located in the inferior wall of the left ventricle in association with mental retardation and ASD in an adult patient. The combination of LVD, ASD and mental retardation implies genetic factor(s) contribute to the pathogenesis of this condition.

## Case Report

A fifty-one year old white female presented with shortness of breath and poor exercise tolerance. She is 61 inches tall and weighs 65 kilograms. She has history of mental retardation. No chest wall or abdominal wall defects were found. Evaluation by transthoracic echocardiography (TTE) revealed an atrial septal defect (ASD) with left-to-right shunt. The patient was referred for ASD repair. Preoperatively chest X-ray revealed mild cardiac enlargement with prominence of both left and right pulmonary arteries and clear lung fields. The electrocardiogram showed normal sinus rhythm and heart rate of 90/minute with occasional premature ventricular beats. After

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induction and endotracheal intubation, intraoperative transesophageal echocardiography (TEE) confirmed the presence of a large ostium secundum ASD. A LVD located at the base of the inferior wall, measuring approximately 1.5 cm by 2 cm was also found. The diverticulum was seen to get smaller during systole and larger during diastole, indicating that LCD has synchronized contraction with the left ventricle (Fig. 1). The patient had a successful closure of the ASD and postoperative recovery was uneventful. She was discharged three days after surgery.

## Discussion

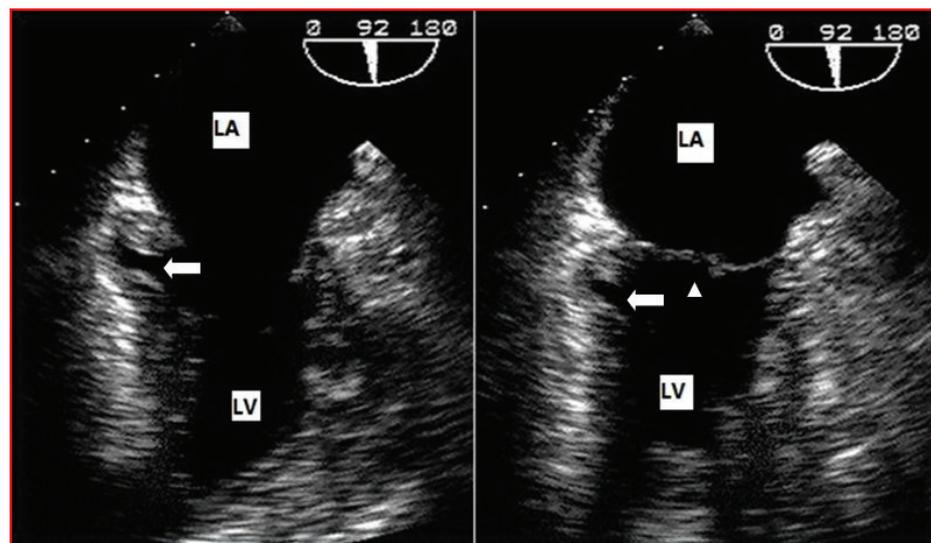
Ventricular diverticula are rarely seen in the adult population. The actual prevalence is unknown. There were only 10 cases in more than 13000 congenital heart disease operations at the Texas Heart Institute over a 20-year period<sup>7</sup>. The majority of ventricular diverticula are congenital, but a small percentage of ventricular diverticula can be caused by other pathological conditions, such as ischemic heart disease<sup>8</sup>. There are two types of ventricular diverticula: muscular and fibrous diverticulum. Muscular diverticulum is characterized by having all three layers of cardiac tissue including endocardium, myocardium and pericardium in the wall. Fibrous diverticulum arises in the subvalvular areas and histologically has predominantly fibrous tissue in the wall. Although the location of the ventricular diverticulum in the majority of patients is in the apex of left ventricle<sup>4</sup>, ventricular diverticula can

also occur in other locations, as in our case. The etiology and mechanism of LVD are unknown. LVD appears to be a developmental abnormality characterized by a local embryological development failure of the ventricular muscle<sup>3,9</sup>. The coexistence of LVD in our patient with mental retardation and ASD implies that genetic factor(s) contribute to the pathogenesis. There has been one report of familial occurrence of LVD involving siblings<sup>10</sup>.

Ventricular diverticula can be isolated or coexist with other clinical conditions. Seventy percent of patients are found to be associated with congenital midline thoracoabdominal defects<sup>5</sup>. Ventricular diverticula has also be reported with other cardiac anomalies, such as ventricular septal defect, ASD, dextrocardia, Tetralogy of Fallot, endocardial cushion defect, persistent truncus arteriosus, tricuspid atresia, anomalous pulmonary or systemic venous return and hypertrophic cardiomyopathy<sup>11</sup>.

The morbidity and mortality associated with LVD varies. Most ventricular diverticula are asymptomatic for many years, but some may have complications including acute rupture and cardiac tamponade<sup>12</sup>, peripheral embolism, chest pain syndrome, dysrhythmia<sup>13</sup> and endocarditis<sup>11</sup>. LVD may present a dilemma. The current asymptomatic status must be weighed against the potential fatal risk of ventricular rupture. Some advocates surgery and there are reports of successful surgical resection of diverticula in order to prevent acute rupture and cardiac tamponade<sup>14</sup>.

*Fig. 1  
Mid-esophageal two  
chamber transesophageal  
echocardiographic view of the  
left ventricular diverticulum  
(arrow) during diastole  
(left) and systole (right). The  
diverticulum has synchronized  
contraction with the left  
ventricle. The mitral valve is  
indicated by arrowheads. (LA =  
left atrium, LV = left ventricle)*



LVD can be diagnosed by various imaging modalities. Most diverticula are detected by echocardiography, left ventricular angiography, magnetic resonance imaging, or computed tomography<sup>15</sup>. It is possible to establish a fetal diagnosis<sup>16</sup>. With echocardiography ventricular diverticulum generally appears as an echo-free space with a narrowed neck communicating with left ventricle and should be visible in multiple views to exclude the possibility of imaging artifacts. Ventricular diverticula should be differentiated from ventricular aneurysms and ventricular pseudoaneurysms. Generally, a

ventricular aneurysm has a broad-based connection with left ventricle and has paradoxical movement with each cardiac cycle, while a ventricular diverticulum has synchronized contraction with ventricle. Ventricular pseudoaneurysm is a chronic contained rupture of the left ventricle, its wall consisting only of pericardium and thrombus<sup>3</sup>. Echocardiographers and cardiac anesthesiologists should be aware of this anomaly, and include it in the differential diagnosis of abnormally shaped ventricular wall and seek other congenital abnormalities if LVD is detected.

## References

1. HAEGELI LM, ERCIN E, WOLBER T, BRUNCKHORST C, TANNER FC, JENNI R, LÜSCHER TF, DURU F: Arrhythmic manifestations in patients with congenital left ventricular aneurysms and diverticula. *Am J Cardiol*; 2011 Dec 15, 108(12):1826-30.
2. CHEN HW, CHEN ML, YANG B, JU WZ, ZHANG FX, HOU XF, CHEN C, SHAN QJ, ZOU JG, XU DJ, CAO KJ: Clinical characteristics of congenital ventricular aneurysm and diverticula in inland China. *Zhonghua Xin Xue Guan Bing Za Zhi*. 2011 Sep, 39(9):865-8.
3. OHLOW MA: Congenital left ventricular aneurysms and diverticula: definition, pathophysiology, clinical relevance and treatment. *Cardiology*; 2006, 106(2):63-72.
4. YAMASHITA H, MOMOMURA S, MOCHIZUKI T, SUZUKI J, TAKENAKA K, SERIZAWA T: Congenital left ventricular diverticulum with septal defect, mitral and tricuspid regurgitation, and coronary disease. *Am Heart J*; 1993, 126:461-463.
5. CANTRELL JR, HALLER JA, RAVITCH MM: A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. *Surg. Gynecol. Obstet*; 1958, 107:602-614.
6. VICTOR S, RAVINDRAN P, MARAGATHAM, ET AL: Congenital diverticulum of muscular interventricular septum in association of atrial septal defect. *Indian Heart J*; 1984, 36(6):401-2.
7. OKEREKE OUJ, COOLEY DA, FRAZIER OH: Congenital diverticulum of the left ventricle. *J. Thoracic & Cardiovascular Surgery*; 1986, 91:208-14.
8. BASHOUR TT, ET AL: Left ventricular narrow-neck large diverticulum caused by ischemic heart disease. *Am. Heart J*; 1997, 133(4):481-483.
9. CIANCIULLI TF, DEL CARMEN GONZALEZ COLASO P, ET AL: Left ventricular diverticulum, a rare echocardiographic finding: two adult patients and review of the literature. *Cardiology J*; 2009, 16(1):76-81.
10. SHIZUKUDA Y, MURAKAMI H, IWAKURA M, ET AL: Siblings with left ventricular diverticulum and hypertrophic cardiomyopathy. *J. cardiology (Japan)*; 1988, 18:867-974.
11. WALTON-SHORLEY M, SMITH SM, TALLEY JD: Left ventricular diverticulum: case report and review of the literature. *Catheterization and Cardiovascular Diagnosis*; 1992, 26(1):31-33.
12. GOWITT GT, ZAKI SA: Rupture of a cardiac diverticulum resulting in sudden death. *Am. J. Forensic med Pathology*; 1988, 9:155-58.
13. SHEN EN, FUKUYAMA O, HERRE JM, ET AL: Ventricular tachycardia with congenital ventricular diverticulum. *Chest*; 1991, 100:283-285.
14. SUILEN C, BEAT F, RUTISHAUSER W: Congenital intrathoracic left ventricular diverticulum in an adult. *Chest*; 1990, 98:750-751.
15. GOBAL F, SACHDEVA R, URETSKY BF: Computed tomography and angiography of left ventricular diverticulum. *J Am Coll Cardiol*; 2011 Aug 23, 58(9):e17.
16. BRACHLOW A, SABLE C, SMITH S, SLACK M, MARTIN G: Fetal diagnosis and postnatal follow-up of an asymptomatic congenital left ventricular diverticulum. *Pediatr Cardiol*; 2002, 23:658-660.