Ventricular diverticulum: Definition, pathophysiology, clinical manifestations and treatment

George D. Giannoglou, Stefanos H. Delioglanis, Stelios Paraskevaidis, Yiannis S. Chatzizisis, George E. Parcharidis

1st Cardiology Department, AHEPA University General Hospital, Medical School, Aristotle University of Thessaloniki, Greece

ABSTRACT: Ventricular diverticula are rare abnormalities of the heart, whose origin is not fully understood yet. They are mostly congenital, either isolated or associated with other cardiac and extracardiac defects (Cantrell's pentalogy). Although their etiology is not clear, an embryologic developmental defect has been proposed. Yet, some of them are associated with cardiomyopathies, inflammation or trauma. We discuss the case of a patient with hypertrophic cardiomyopathy and an apical diverticulum. The hypothesis made was that, an obstructing hypertrophic mass, localized in the midportion of the left ventricle, creates a pressure gradient between the apical and basal portions which finally leads to the creation of the diverticulum. Acute rupture, ventricular arrhythmias, peripheral arterial embolism and infective endocarditis are few of their complications. Besides transthoracic 2-D echocardiography and left ventriculography, magnetic resonance imaging (MRI) is the gold standard means of diagnosis. The opinions in the literature, as far as the pharmacological treatment or surgical resection, are controversial.

Key Words: Ventricular diverticulum, Hypertrophic cardiomyopathy.

INTRODUCTION

Ventricular diverticula are very rare abnormalities of the heart, only a few of which have been described in the literature for the past 200 years[1] and whose origin is not fully understood yet. They are mostly congenital, either isolated or associated with other cardiac and extracardiac defects. Yet, some of them are not congenital and only seven cases[2,3] have been described to associate hypertrophic cardiomyopathy (HCM) with them. We present the case of a man with HCM and a left ventricular apical diverticulum.

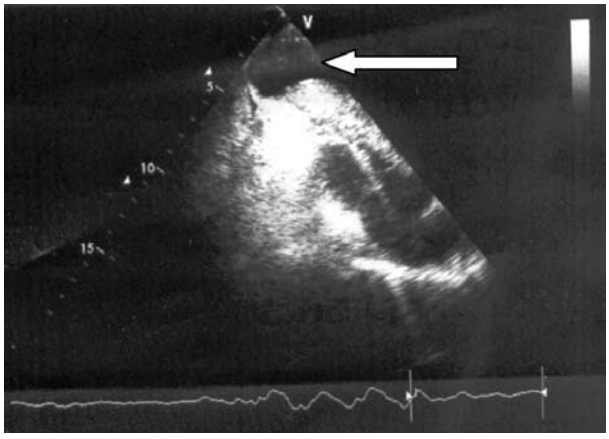
CASE REPORT

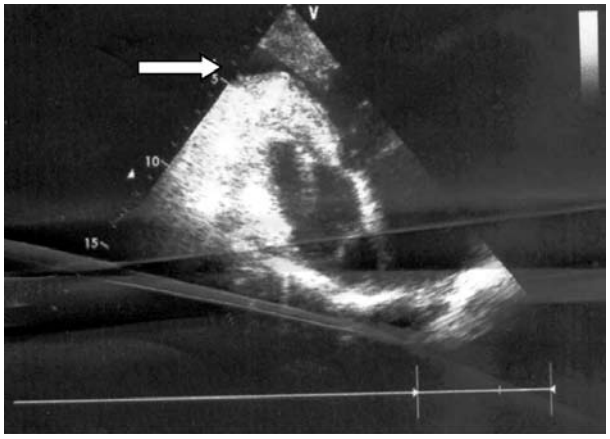
Our patient, a 69-year-old man was admitted to our hospital with symptoms of dyspnea and tachycardia. Hypertrophic cardiomyopathy (for 30 years), NYHA III heart failure and a DDDR pacemaker (for 2 years due to sick sinus syndrome) were the most important recordings of his medical history.

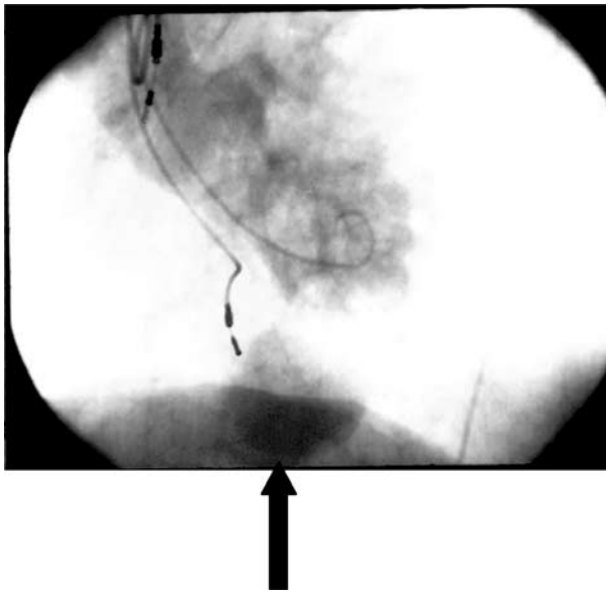
The physical examination revealed a heart rate of approximately 80 beats/min with a regular heart rhythm while his blood pressure was 105/75 mmHg with 20 breaths/min. An olosystolic murmur 2/6 was heard indicating a possible mitral regurgitation. The pulmonary and abdominal examination was unremarkable. There was a mild edema at lower extremities. The ECG showed an atrial flutter and a pacemaker rhythm. The results of laboratory tests were normal and the chest X-ray revealed a cardiothoracic index of approximately 70%.

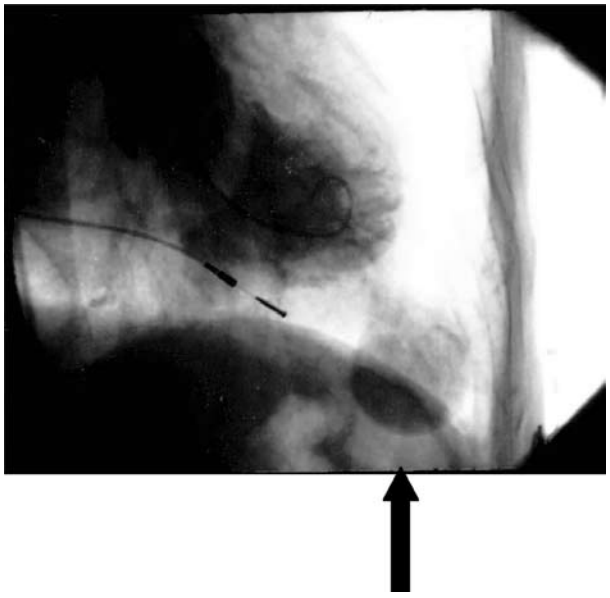
The 24-hour Holter ECG did not record anything significant. The Transthoracic Echocardiography (TTE) indicated hypertrophy and dilation of the left ventricle, ejection fraction approximately 50%, mitral and tricuspid regurgitation 2/4, (Right Ventricular Systolic Pressure) RVSP=40 mmHg and, finally, a bulging of the apex of the left ventricle, with thin walls and a narrow neck, which extended over the

Corresponding author: George D. Giannoglou, MD, PhD, 1st Cardiology Department, AHEPA University General Hospital, Medical School, Aristotle University of Thessaloniki, 1 St. Kyriakidi Street, 54636, Thessaloniki, Greece, Tel. - Fax: +30 2310 994837, e-mail:van@med.auth.gr









right ventricle.

No other significant intracardiac anomalies were found during TTE. The patient was scheduled for an electrophysiologic study and a coronary angiography.

The coronary angiography revealed no atherosclerotic lesions at the arteries and the left ventriculography showed a possible pseudoaneurysm just below the apex (outpouching with a narrow neck and no contractility).

An electrophysiologic study was performed but the attempt for a radiofrequency ablation of the right isthmus had no results. There was a successful electrical cardioversion of the atrial flutter.

The patient was referred to the Thoracic department of our hospital, which proceeded to the resection of the possible pseudoaneurysm. The patient was operated and admitted to the ICU. Thirteen days later he died due to acute renal failure and septic shock. The histological examination of the formation resected, revealed an intense fibrosis with several hypertrophic myocardial fibers and connective tissue. We presume that this was a case of a fibrous diverticulum and its association with HCM is being discussed below.

DISCUSSION

Cardiac diverticulum is a very rare malformation of the heart, which is mostly congenital. Since its first description in 1816, 411 cases have been reported[1], 70% of which involved other extracardiac anomalies while the rest of them were reported as isolated. They

usually arise from the left ventricle but many cases have been presented with right ventricular[4-7], biventricular[8,9] or right atrial[10] origin.

Although its etiology is not clear, an embryologic developmental defect has been proposed. A failure of normal midline fusion of the paired primitive mesoderm, at the 4th embryonic week, in combination with abnormal fusion of the cardiac loop to the yolk sac before its descent, is believed to result in the development of ventricular diverticula[11]. Histopathologically, they are classified as fibrous and muscular. As far as the former is concerned, the cardiac wall is constituted mostly of fibrous tissue with few or no muscular fibers. It is non-contractile and most commonly originates from the apical or subvalvular position; it occurs predominantly in black people and Africans. The subvalvular type may be accompanied by aortic or mitral regurgitation and systemic embolism. They are associated with neither defects in the median line nor congenital cardiac malformations[12]. The muscular type includes all the three layers of the heart and usually emerges from the apex but rarely from the right ventricle or both chambers. It has a mechanical activity, synchronous with the activity of the ventricles. Its connection to the ventricular chamber is narrow[13,14]. It is frequently associated with other congenital anomalies, including those of the abdominal wall, diaphragm, sternum, pericardium, and the heart itself. Right atrial septal defect[6,15],ventricular septal defect[16-18], tetralogy of Fallot[19-21], pericardial effusion[6], hypoplastic right ventricle[22], pulmonary stenosis[23], absence of the sternum[24], asplenia[25], disseminated neonatal hemangiomatosis[7], bilateral obstruction of the ureteropelvic junction and agenesis of corpus callosus[26] are the malformations described in the bibliography. The syndrome, which consists of omphalocele, ventricular diverticulum, anterior diaphragmatic herniation, thoracic ectopia cordis (in its most severe form), along with other cardiac deformities (atrial septal defect, anomalous venous pulmonary return, dextrorotation of the heart), is known as the Cantrell's syndrome[27-29]. The latter has been identified in many cases involving ventricular diverticula and, even though a cause has yet to be found, genes located on the X-chromosome may be involved[30].

However, few cases of diverticula which are not congenital but associated with hypertrophic cardiomyopathy (HCM) have been presented in the literature[2,3]. The hypothesis made was that an obstructing hypertrophic mass localized in the midportion of the ventricle creates a pressure gradient between the apical and basal portions. The ventricle chronically exposed to high intracavitary pressure gradually changes and a dilated muscular chamber is being created, as the midventricular obstruction worsens. In addition, transmural infarction may occur to HCM due to imbalance between oxygen supply and demand, in the absence of atherosclerotic lesions of the coronary arteries. As a result, subendocardial and apical infarction may eventually create a diverticulum which mostly consists of fibrous tissue[3].

Patients with cardiac diverticula which are not associated with other malformations are usually asymptomatic. ECG abnormalities such as T wave inversion[31], ST segment depression and repolarization abnormalities[32] can be found accidentally during a medical check-up. Chest pain, usually at rest, mostly atypical[33], is a symptom that may appear to those patients[32,34-37]. Supraventricular[38] or ventricular arrhythmias (ventricular fibrillation[39], focal ventricular tachycardia[40,41], multiple premature ventricular complexes[42]) which may lead to syncope or sudden death can be the first clinical manifestation. Patients with diverticula, especially the fibrous ones (because of their non-contractility), may also experience peripheral arterial embolism[43]. When they are associated with valve impairments, infective endocarditis[44] may be the first clinical manifestation. Congestive heart failure[45] and acute rupture[46,47] have also been described in the literature.

The diverticulum can be easily suspected when it is associated with malformations of the median line as described above. In such a case a pulsatile epigastric mass is very characteristic[48]. Clinical findings such as heart murmur[49] and chest pain[33] or arrhythmias and repolarization abnormalities[32] found at an ECG must be carefully evaluated. Cardiomegaly[50] or an abnormal protrusion of the cardiac silhouette[51] on chest X-ray may provide the earliest clinical suspicion. Transthoracic 2-D and Doppler echocardiography can show the origin, the narrow neck, the walls and the contractility of the diverticula[35,52-54]. Left ventriculography[36,55] with phase analysis can precisely describe the location of the diverticulum, its shape and contractility or other malformations of the heart. Magnetic resonance imaging (MRI) has the poten-

tial not only to identify but also to categorize diverticula non-invasively and differentiate them between muscular contracting and non-contracting fibrous ones[56,57]. Histological examination, once the resection of the formation is decided, is the one that will verify the synthesis of the diverticulum's wall and may give information about its cause[37,49].

Once the diverticula are associated with other malformations, their resection together with the repairing of the deformities is the most common treatment. Specifically, in Cantrell's pentalogy, the strategy of repair is divided in two stages: (1) urgent soft tissue coverage and hemodynamic palliation; and (2) intracardiac repair with concomitant chest wall reconstruction and reduction of the heart into the thoracic cavity[27]. If the patient is asymptomatic and the diverticulum is not associated with other malformations, the opinions, as far as the resection is concerned, are controversial. Because of their life-threatening complications, such as sudden death due to arrhythmias, acute rupture and the possibility of systemic embolization, mostly in the fibrous ones, the surgical resection seems to be the most usual form of treatment seen in the literature. Yet, in asymptomatic patients, many authors suggest long-term anticoagulation therapy[43], antiarrhythmic drugs class I and III or beta-adrenergic block agents[1,33], radiofrequency ablation[33,58] and implantation of cardioverter defibrillators[1].

In our case the diverticulum had no contractility and we believed that it was a case of pseudoaneurysm. We decided that the surgical resection was the best treatment for our patient due to symptoms of heart failure (NYHA III) and potential association with systemic thromboembolism and acute rupture, as described in the literature. The histological examination of the resected formation, revealed its true origin afterwards. All diverticula described in the literature which are associated with HCM are fibrous and, as a result, long term anticoagulation therapy or surgical resection seems to be the preferred treatment. The clinical outcome depends on the co-morbidities of the patient.

Κοιλιακό εκκόλπωμα: Ορισμός, παθοφυσιολογία, κλινικές εκδηλώσεις και θεραπεία

Γεώργιος Δ. Γιαννόγλου, Στέφανος Χ. Δεληογλάνης, Στέλιος Παρασκευαΐδης, Ιωάννης Σ. Χατζηζήσης, Γεώργιος Ε. Παρχαρίδης

1η Καρδιολογική Κλινική, Πανεπιστημιακό Γενικό Νοσοκομείο ΑΧΕΠΑ, Αριστοτέλειο Πανεπιστήμιο, Θεσσαλονίκη

ΠΕΡΙΛΗΨΗ: Περιγράφεται σπάνια περίπτωση κοιλιακού εκκολπώματος (ventricular diverticulum). Ασθενής ηλικίας 69 ετών προσήλθε στο νοσοκομείο με συμπτώματα δύπνοιας και ταχυκαρδιών. Ο ασθενής έπασχε από υπερτροφική καρδιομυοπάθεια (επί 30 χρόνια), έφερε μόνιμο καρδιακό βηματοδότη από 2ετίας λόγω συνδρόμου νοσούντος φλεβοκόμβου και παρουσίαζε καρδιακή ανεπάρκεια σταδίου ΙΙΙ ΝΥΗΑ. Ο ασθενής ήταν συμπτωματικός και κρίθηκε υψηλού κινδύνου. Υποβλήθηκε σε καρδιακό καθετηριασμό, ηχωκαρδιογράφημα, ηλεκτροφυσιολογική μελέτη και καρδιοχειρουργική επέμβαση αφαίρεσης εκκολπώματος. Ο ασθενής παρουσίαζε αύξηση του καρδιοθωρακικού δείκτη, υπετροφία και διάταση της αριστερής κοιλίας, κλάσμα εξωθήσεως περίπου 50%, ανεπάρκεια μιτροειδούς και τριγλώχινας 2/4, υπερτροφία και διάταση αριστερής κοιλίας, συστολική πίεση δεξιάς κοιλίας 40 mmHg και προβολή κορυφής αριστερής κοιλίας με λεπτό τοίχωμα και στενό μίσχο. Η στεφανιογραφία δεν έδειξε αθηροσκληρωτικές βλάβες στις στεφανιαίες αρτηρίες και η αριστερή κοιλιογραφία έδειξε πιθανό ψευδοανεύρυσμα ακριβώς κάτω από την κορυφή (προβολή προς τα έξω, με στενό αυχένα και χωρίς συσπαστικότητα). Έγινε ηλεκτροφυσιολογική μελέτη, αλλά η προσπάθεια για κατάλυση με υψίσυχνορευμα του δεξιού ισθμού ήταν χωρίς αποτέλεσμα. Η προσπάθεια ηλεκτρικής ανάταξης της κολπικής μαρμαρυγής ήταν ανεπιτυχής. Ο ασθενής υποβλήθηκε σε χειρουργική επέμβαση και εισήχθη στη Μονάδα Εντατικής Θεραπείας. Μετά 13 ημέρες ο ασθενής απεβίωσε λόγω οξείας νεφρικής ανεπάρκειας και σηπτικού shock. Η ιστολογική εξέταση του μορφώματος που αφαιρέθηκε, έδειξε έντονη ίνωση, με πολλές υπερτροφικές μυοκαρδιακές ίνες και συνδετικό ιστό. Συμπεραίνεται ότι η σπάνια περίπτωση εκκολπώματος της αριστερής κοιλίας μπορεί να εμφανίζεται με τη μορφή δύσπνοιας και αρρυθμιών, σε έδαφος υπερτροφικής μυοκαρδιοπάθειας.

Λέξεις Κλειδιά: Κοιλιακό εκκόλπωμα, Υπερτροφική καρδιομυοπάθεια.

REFERENCES

1. Ohlow, M.A., Congenital left ventricular aneurysms and diverticula: definition, pathophysiology, clinical relevance and treatment. Cardiology, 2006. 106(2): 63-72.
2. Maron, B.J., R.G. Hauser, and W.C. Roberts, Hypertrophic cardiomyopathy with left ventricular apical diverticulum. Am J Cardiol, 1996. 77(14): 1263-5.
3. Yamashita, C., et al., Left ventricular diverticulum with hypertrophy of the left ventricular apex. Ann Thorac Surg, 1992. 54(4): 761-3.
4. Jeserich, M., et al., Congenital left ventricular apical aneurysm or diverticulum mimicking infarct aneurysm and a right ventricular diverticulum in an adult. Clin Res Cardiol, 2006. 95(7): 373-8.
5. Gardiner, H.M., et al., Images in cardiovascular medicine. Pericardiocentesis at 14 weeks: effective treatment of pericardial effusion complicating right ventricular diverticulum. Circulation, 2005. 112(9): e120.
6. Del Rio, M., et al., Prenatal diagnosis of a right ventricular diverticulum complicated by pericardial effusion in the first trimester. Ultrasound Obstet Gynecol, 2005. 25(4): 409-11.
7. Miller, K.K., et al., Congenital right ventricular diverticulum in association with disseminated neonatal hemangiomatosis. Can J Cardiol, 2003. 19(3): 303-5.
8. Ritter, M., E. Oechslin, and R. Jenni, Images in cardiology: biventricular diverticula in a patient with restrictive cardiomyopathy. Heart, 2000. 84(3): 333.
9. Ashraf, M.H., et al., Congenital diverticulum of the heart of biventricular origin. Thorac Cardiovasc Surg, 1984. 32(6): 389-91.
10. Kobza, R., et al., Enlargement of the right atrium--diverticulum or aneurysm? Eur J Echocardiogr, 2003. 4(3): 223-5.
11. Barutcu, I., H. Gullu, and F. Kosar, Isolated true contractile left ventricular diverticulum in an adult patient. Int J Cardiol, 2004. 97(1): 141-2.
12. Shen, E.N., et al., Ventricular tachycardia with congenital ventricular diverticulum. Chest, 1991. 100(1): 283-5.
13. Mady, C., Left ventricular diverticulum: analysis of two operated cases and review of the literature. Angiology, 1982. 33(4): 280-6.
14. Treistman, B., et al., Diverticulum or aneurysm of left ventricle. Am J Cardiol, 1973. 32(1): 119-23.
15. Gengsakul, A., S.S. Sett, and M.C. Hosking, Congenital right atrial diverticulum, atrial septal defect within the oval fossa, and complex pulmonary valvar obstruction in an infant with chromosome 8(p23.1) deletion. Cardiol Young, 2005. 15(3): 306-8.
16. Solomon, H.M., et al., Spontaneous and induced alterations in the cardiac membranous ventricular septum of fetal, weanling, and adult rats. Teratology, 1997. 55(3): 185-94.
17. Gerlis, L.M., S.Y. Ho, and M.L. Rigby, Right ventricular outflow obstruction by anomalies of the tricuspid valve: report of a windsock diverticulum. Pediatr Cardiol, 1992. 13(1): 59-62.
18. Hofbeck, M., et al., Diverticulum of the right ventricle associated with ventricular septal defect. Int J Cardiol, 1989. 24(1): 27-33.
19. Alayunt, A., et al., Left ventricular diverticulum associated with Cantrell's syndrome and tetralogy of Fallot in an adult. Scand Cardiovasc J, 2001. 35(1): 55-7.
20. Villegas Garcia, M., et al., Congenital diverticulum of the right ventricle associated with coarctation of aorta, atrial and ventricular septal defect and ductus. Eur J Echocardiogr, 2001. 2(3): 205-6.
21. Morales, J.M., et al., Ectopia cordis and other midline defects. Ann Thorac Surg, 2000. 70(1): 111-4.
22. Orsmond, G.S., H.S. Joffe, and E. Chesler, Congenital diverticulum of the left ventricle associated with hypoplastic right ventricle and ventricular septal defect. Circulation, 1973. 48(5): 1135-9.
23. Carter, J.B., et al., Congenital diverticulum of the right ventricle. Association with pulmonary stenosis and ventricular septal defect. Am J Cardiol, 1971. 28(4): 478-82.
24. Cottrill, C.M., J. Tamaren, and B. Hall, Sternal defects associated with congenital pericardial and cardiac defects. Cardiol Young, 1998. 8(1): 100-4.
25. Gerlis, L.M., R.H. Anderson, and A.J. Howat, Atrioventricular valve diverticulum--an unusual congenital malformation. Int J Cardiol, 1984. 6(1): 75-8.
26. Massin, M.M. and A. Verloes, A congenital left ventricular diverticulum combined with a complex malformation syndrome. Acta Cardiol, 2003. 58(3): 207-8.
27. Marijon, E., A.O. Hausse-Mocumbi, and B. Ferreira, Cantrell's syndrome. Cardiol Young, 2006. 16(1): 95-6.
28. Di Bernardo, S., N. Sekarski, and E. Meijboom, Left ventricular diverticulum in a neonate with Cantrell syndrome. Heart, 2004. 90(11): 1320.
29. Abinader, E.G. and D. Sharif, Cantrell's Syndrome: Left Ventricular Diverticulum in an Adult Patient. Circulation, 2001. 103(16): E89-E89.
30. Halbertsma, F.J., A. van Oort, and F. van der Staak, Cardiac diverticulum and omphalocele: Cantrell's pentalogy or syndrome. Cardiol Young, 2002. 12(1): 71-4.
31. Barboteu, M., et al., [Giant negative T waves in idiopathic apical diverticulum of the left ventricle in adults]. Arch Mal Coeur Vaiss, 1995. 88(10): 1475-7.

32. Yazici, M., et al., Left ventricular diverticulum in two adult patients. Int Heart J, 2005. 46(1): 161-5.
33. Mayer, K., et al., [Congenital left ventricular aneurysms and diverticula: clinical findings, diagnosis and course]. Schweiz Med Wochenschr, 1999. 129(35): 1249-56.
34. Kosar, F., I. Sahin, and H. Gullu, Isolated large true contractile left ventricular diverticulum mimicking ischemia in an adult patient: a case report. Heart Vessels, 2005. 20(2): 85-7.
35. Russo, G., et al., Imaging for the diagnosis of an unusual case of left ventricular aneurysm. Cardiovasc Intervent Radiol, 2005. 28(2): 254-5.
36. Takahashi, M., et al., Multiple left ventricular diverticula detected by second harmonic imaging: a case report. Circ J, 2003. 67(11): 972-4.
37. Miura, M., et al., Surgical management of localized left ventricular aneurysm associated with acute myocarditis. Jpn J Thorac Cardiovasc Surg, 2003. 51(6): 249-52.
38. Perez-Fernandez, R., et al., [Congenital apical left ventricular aneurysm]. Rev Esp Cardiol, 2005. 58(11): 1361-3.
39. Speechly-Dick, M.E., R.M. Oliver, and G.I. Slapak, Congenital left ventricular diverticula: a rare cause of sudden cardiac death. Postgrad Med J, 1992. 68(799): 378-80.
40. Intini, A., et al., Electrocardiographic imaging (ECGI), a novel diagnostic modality used for mapping of focal left ventricular tachycardia in a young athlete. Heart Rhythm, 2005. 2(11): 1250-2.
41. Pitol, R., et al., [Congenital ventricular diverticulum associated with ventricular tachycardia]. Arq Bras Cardiol, 2005. 84(2): 173-5.
42. Salazar Gonzalez, J.J., et al., [Isolated non-compaction of the ventricular myocardium]. An Esp Pediatr, 2002. 57(6): 570-3.
43. Parthenakis, F.I., et al., Peripheral arterial embolism due to a left ventricular diverticulum in a young adult. Chest, 2005. 127(4): 1452-4.
44. Kihara, H., et al., [A case of aortic bicuspid valvular endocarditis with congenital left ventricular diverticulum]. J Cardiol, 1995. 26(5): 299-303.
45. Handler, C.E. and J.M. Walker, Congenital diverticulum of the left ventricle presenting as heart failure and diagnosed by magnetic resonance imaging. Int J Cardiol, 1989. 22(1): 115-9.
46. Shauq, A., V. Agarwal, and C. Crawley, Congenital left ventricular diverticulum. Heart Lung Circ, 2006. 15(4): 272-4.
47. Bernasconi, A., et al., Prenatal rupture of a left ventricular diverticulum: a case report and review of the literature. Prenat Diagn, 2004. 24(7): 504-7.
48. Gula, G. and M. Yacoub, Syndrome of congenital ventricular diverticulum and midline thoraco-abdominal defects. Thorax, 1977. 32(3): 365-9.
49. Inoue, T., et al., [A case of isolated congenital left ventricular diverticulum]. Nippon Kyobu Geka Gakkai Zasshi, 1993. 41(3): 467-73.
50. Vaidiyanathan, D., et al., Isolated congenital left ventricular diverticulum in adults. Indian Heart J, 2001. 53(2): 211-3.
51. Hamaoka, K., et al., Congenital ventricular aneurysm and diverticulum in children. Pediatr Cardiol, 1987. 8(3): 169-75.
52. El Kettani, N.E. and R. Dafiri, [Isolated congenital left ventricular diverticulum: report of a paediatric case]. J Radiol, 2006. 87(1): 56-8.
53. Iyisoy, A., et al., A contractile left ventricular diverticulum. Int J Cardiovasc Imaging, 2006. 22(1): 1-3.
54. Gonzales, M.R. and M. Garg, Images in cardiology: A rare finding of left ventricular diverticulum. Clin Cardiol, 2005. 28(6): 308.
55. Pisapia, A., et al., [Criteria of localization of the bundle of Kent]. Arch Mal Coeur Vaiss, 1999. 92 Spec No 1: 57-64.
56. Renzulli, M., L. Lovato, and R. Fattori, MRI findings of small isolated congenital left ventricular diverticulum. Eur Heart J, 2006.
57. Duran, C., M. Kantarci, and K. Karaman, Isolated left ventricular diverticulum in an adult: dynamic contrast-enhanced MRI findings. Anadolu Kardiyol Derg, 2005. 5(1): 85-6.
58. Davidson, N.C., M.J. Cooper, and D.L. Ross, Radiofrequency ablation of a posteroseptal accessory pathway associated with two diverticula of the coronary sinus. Circulation, 2001. 104(2): 240-1.