

## **Case Report**

# **Left Main Coronary Artery Embolization in an 11-Year-Old Girl Due to Inflammatory Myofibroblastic Tumor of the Mitral Valve**

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This report describes a rare case of a subtotal left main coronary artery occlusion from mitral valve tumor embolization in an 11-year-old African American girl. This case is the first to report isolated ST segment elevation in lead aVR as a sign of a subtotal left main coronary artery occlusion in the pediatric population. In our case, we report a rare case of inflammatory myofibroblastic tumor of the mitral valve presenting with acute myocardial infarction due to embolization into the left main coronary artery. Coronary intervention was successfully performed using an aspiration catheter. Inflammatory myofibroblastic tumor usually presents as a solitary pulmonary nodule. Intracardiac involvement has been rarely reported. Despite the benign nature of the tumor, fatal presentations can occur. Early recognition and rapid intervention can be lifesaving in these patients. © 2015 Wiley Periodicals, Inc.

**Key words:** acute coronary syndrome; cardiac tumor; PCI; percutaneous coronary intervention

## **INTRODUCTION**

Acute myocardial infarction (AMI) from left main coronary artery occlusion is an atypical cause of chest pain among the pediatric population in the absence of identified risk factors. Primary cardiac tumors are rare in pediatric population, with an incidence of 0.002%–0.03% [1,2]. Most primary cardiac tumors in the pediatric age group are benign, with rhabdomyomas, myxomas, and fibromas being the most common [3,4]. Cardiac inflammatory myofibroblastic tumor (IMT) is a rare tumor consisting of inflammatory cells and myofibroblastic spindle cells with a benign potential. Although benign in nature, these intracardiac tumors can compromise blood flow, causing variable degrees of obstruction and can potentially have serious presentations: cyanosis, respiratory distress, valvular insufficiency, or give rise to emboli large enough to cause coronary artery occlusion and sudden cardiac death.

## **CASE REPORT**

An 11-year-old African American girl with history of bronchial asthma was presented to the emergency department with acute onset of chest pain. She woke up to

severe retrosternal pressure-like chest pain; the pain was radiating to the back and was unrelated to change in position. The pain was associated with mild shortness of breath, with no other associated symptoms. There was no reported family history of any congenital heart disease, coronary artery disease, sudden cardiac death, or familial hyperlipidemia.

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The management of the case was done collaboratively between the Children's Hospital of Michigan and Harper University Hospital, Detroit Medical Center.

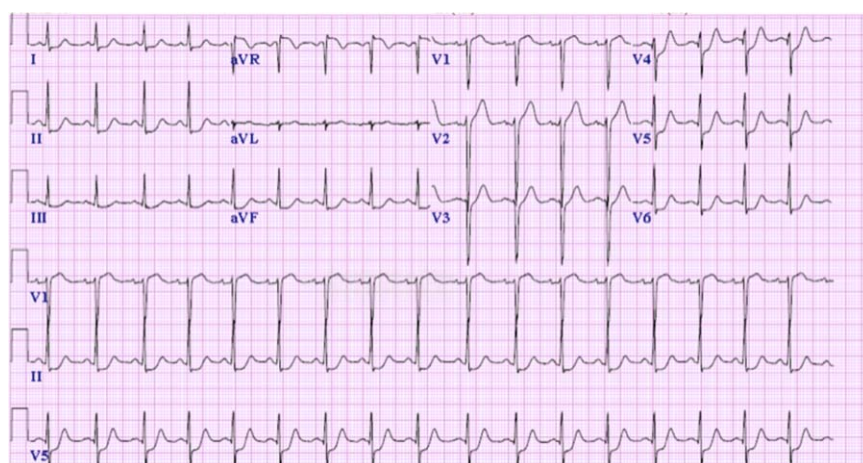
Conflict of interest: Nothing to report.

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**Fig. 1. ECG on presentation showing ST-segment elevation in lead aVR with widespread ST-segment depression in leads II, III, and aVF and leads V4, V5, and V6. [Color figure can be viewed in the online issue, which is available at [wileyonlinelibrary.com](http://wileyonlinelibrary.com).]**

Upon evaluation in the emergency room, she seemed to be in mild to moderate distress with blood pressure of 122/52 mm Hg, heart rate of 120 beats per minute, respiratory rate of 20 breaths per minute, and temperature of 37.2°C. Oxygen saturation was 97% in the room air. There was no jugular venous distention, and the lung examination was normal. Cardiac examination revealed regular rhythm at rate of 120 beats per minute, no murmur, clicks, or gallops. The chest pain was not reproducible on chest palpation. The rest of her physical examination was normal.

The 12-lead electrocardiogram showed ST segment elevation in lead aVR with associated ST segment depression in leads II, III, and aVF (Fig. 1). The chest radiograph was normal. The initial set of cardiac enzymes showed elevated troponins I of 2.5 ng/ml, peaked at >40 ng/ml. Echocardiography was urgently performed in the emergency department and revealed moderately depressed left ventricular systolic function with hypokinesis of the anterior, anteroapical left ventricular wall from base to apex. A small hyperechoic mass 6 × 12 mm was noted on the anterior leaflet of the mitral valve. No mitral valve insufficiency or stenosis was noted (Fig. 2). Cardiac catheterization was urgently performed and revealed 80% obstructive lesion in the left main coronary artery extending to the left anterior descending artery. Two attempts of mechanical embolectomy using 6-F Export aspiration catheter failed followed by a successful embolectomy using 7-F Priority One aspiration catheter with 0% residual stenosis after the procedure. Distal embolic protection technique was used to prevent distal embolization using a Spider filter distal protection device

(Fig. 3). The removed tissue was sent for pathology, and the initial analysis revealed undifferentiated malignant neoplasm with epithelioid features, suggestive of sarcoma (Fig. 4). A computed tomographic scan of the thorax, abdomen, and pelvis revealed no evidence of metastatic disease. Magnetic resonance angiography of the brain was performed and revealed unremarkable intracranial circulation pattern with no evidence of infarction. Positron emission tomography was performed from the skull base to the mid thighs and revealed no abnormal fluorodeoxyglucose uptake suggestive of metastasis.

Cardiac surgery was performed under cardiopulmonary bypass for resection of the tumor. In the operating room, the tumor appeared as a fimbriated mass involving most of the anterior leaflet at the posteromedial commissure; it also appeared to involve the free edge and extended to some of the chordae. The posterolateral leaflet appeared unaffected. The initial impression of the operating surgeon was papillary fibroelastoma based on the gross morphology of the tumor; however, frozen section evaluation in the operating room did not support that diagnosis. The initial consideration was to remove the tumor and save the mitral valve, but the reconstruction was not surgically feasible because the tumor involved the entire medial half of the anterior leaflet with the free edge (Fig. 5). The native mitral valve was replaced with a 27-mm St. Jude prosthetic valve (St. Jude Medical, Minneapolis, MN). Her postoperative course was uncomplicated. The tumor specimen was sent for histopathologic examination. Macroscopic examination showed a villous rubbery mass measuring 0.7 × 0.5 × 0.4 cm. Histologic examination revealed spindle-shaped cells arranged in

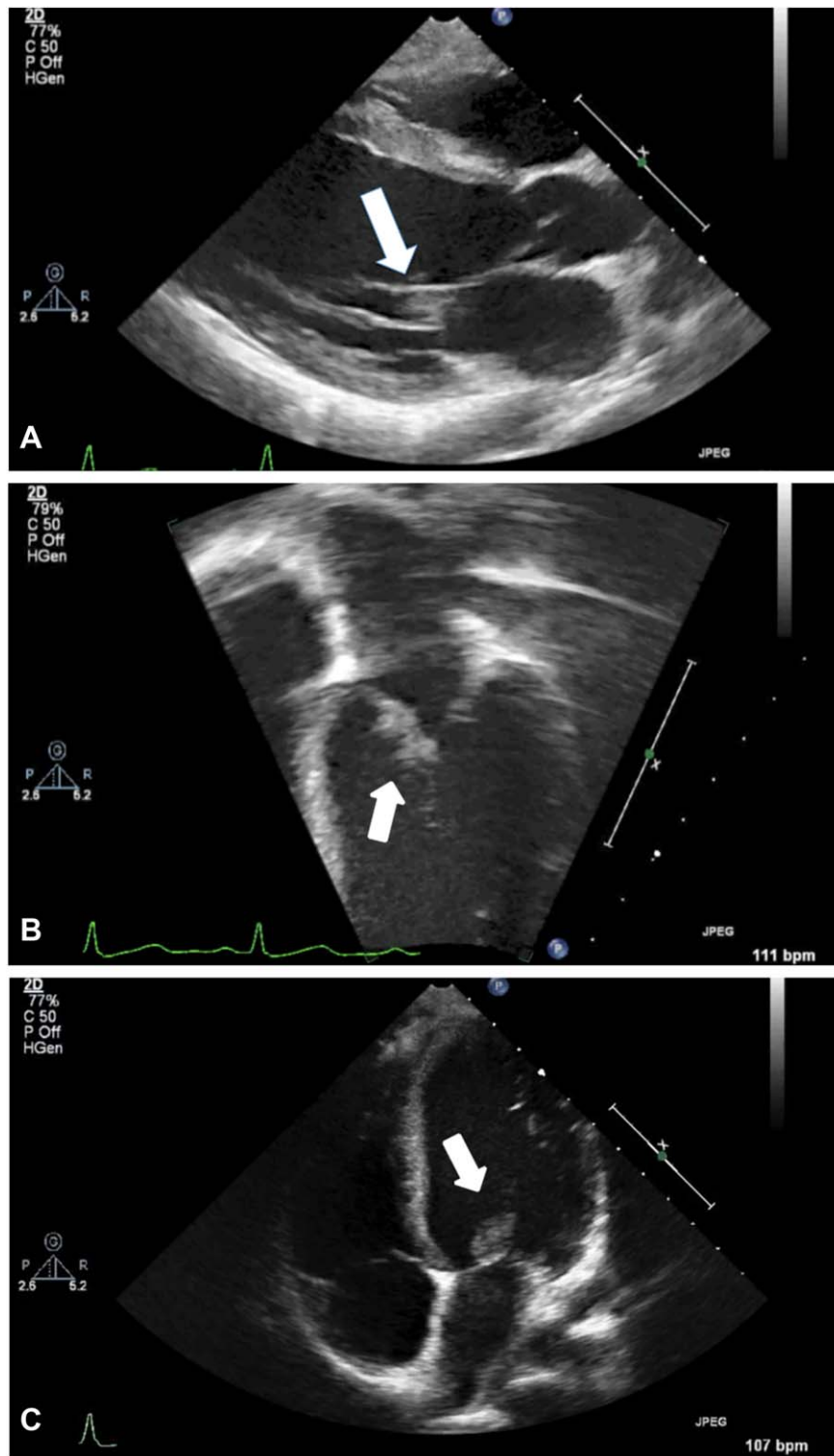
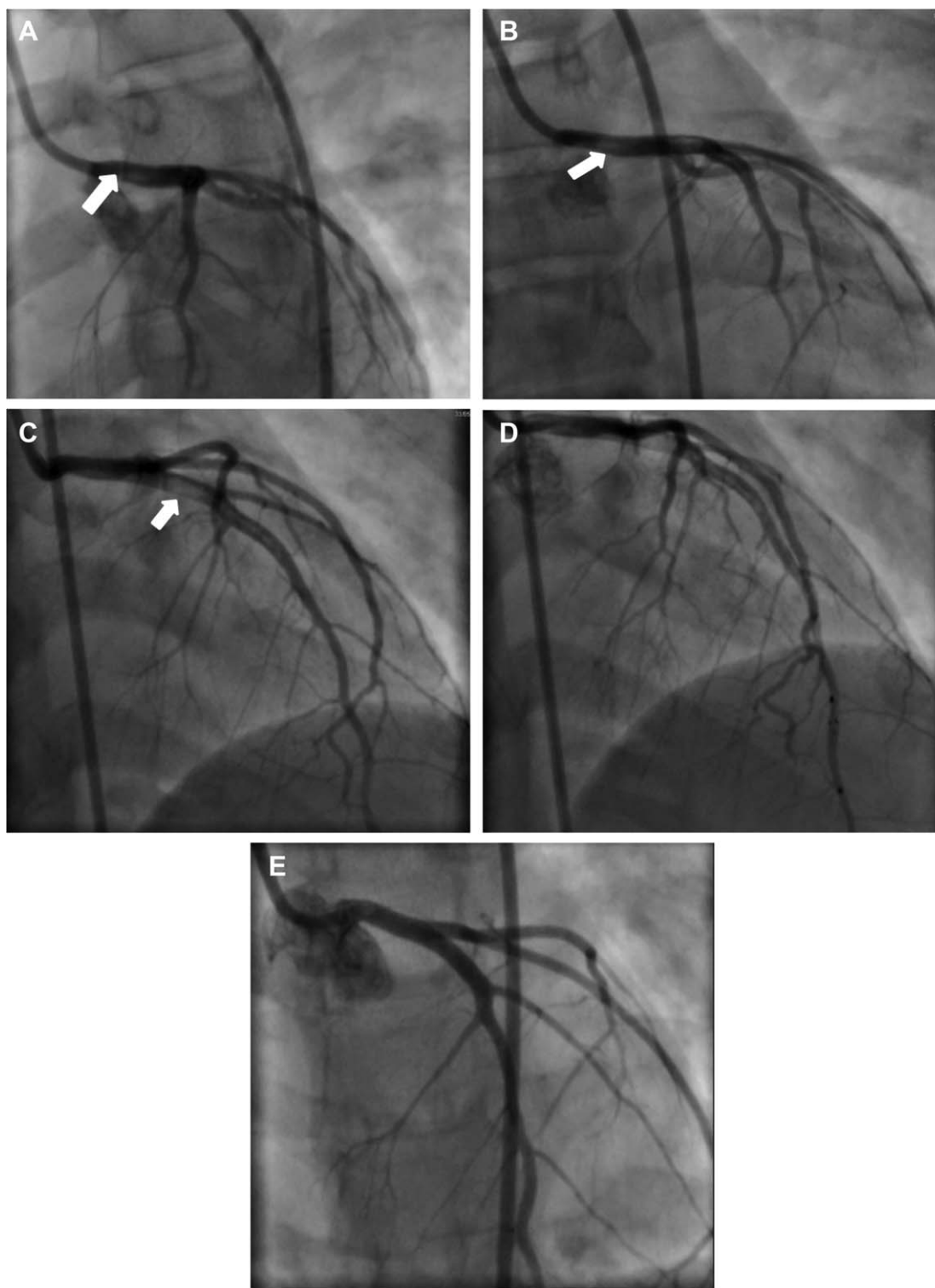


Fig. 2. (A–C) Transthoracic echocardiography illustrating a  $6 \times 12$  mm mass on the anterior mitral valve. (A) Parasternal long-axis. (B, C) Apical four-chamber views. [Color figure can be viewed in the online issue, which is available at [wileyonlinelibrary.com](http://wileyonlinelibrary.com).]

storiform pattern in a myxoid background. The cells showed hyperchromatism with frequent mitotic figures. The final histopathology analysis revealed IMT. The

tumor has a benign potential. No further intervention was indicated. She was discharged home the 11th post-operative day on warfarin and carvedilol.



**Fig. 3.** (A–E) Percutaneous coronary intervention. (A–C) Embolized tumor in the left main coronary artery extending to the left anterior descending coronary artery. (D) Aspiration catheter with the Spider filter distal protection device. (E) Left main coronary artery and long anterior descending coronary artery with 0% residual occlusion after successful aspiration of the tumor.



Fig. 4. The tumor embolus aspirated from the left main coronary artery. [Color figure can be viewed in the online issue, which is available at [wileyonlinelibrary.com](http://wileyonlinelibrary.com).]

## DISCUSSION

IMT is a rare benign tumor characterized by proliferation of myofibroblast and inflammatory cell infiltration. There have been few case reports of IMT with the exact incidence unknown [5–8].

It occurs most commonly in the lungs and usually presents as a solitary pulmonary nodule [9]. IMT has been rarely reported in the cardiac tissue [10–12]. The most accepted pathogenesis of IMT is thought to be related to up-regulation of the immune system with resulting symptoms of fever, weight loss, and laboratory abnormalities, including elevated sedimentation rate, iron-deficiency anemia, thrombocytosis, and hypergammaglobulinemia. Patients with cardiac involvement usually present with syncope, chest pain, or shortness of breath. Krishna et al. [8] reported a presentation with aortic valve involvement resulting in aortic regurgitation.

The differential diagnosis include cardiac myxoma, papillary fibroelastoma, and low-grade sarcoma. Histological differences can easily differentiate IMT from myxoma and papillary fibroelastoma. The distinction between IMT and low-grade sarcoma, however, can be challenging with lack of significant pleomorphism and atypical mitotic figures favoring IMT diagnosis. Cardiac IMTs are biologically benign, but because of their location inside the heart, they could be fatal. Burke et al. reported a case series of 10 patients, with one case showing coronary artery embolization resulting in sudden cardiac death. In our case, we are reporting AMI as a presentation of IMT of the mitral valve due to embolic occlusion of the left main coronary artery. Our patient was successfully rescued with percutaneous coronary intervention. Cardiac IMT seems to lack metastatic potential. Burke et al. reported no metastatic complication in their case series.

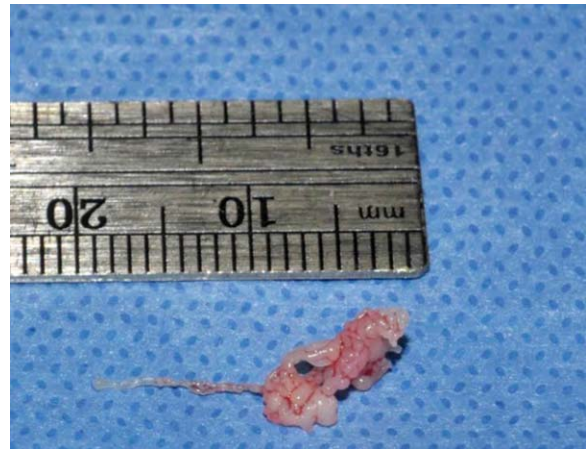


Fig. 5. Macroscopic examination of the excised tumor revealing a gelatinous mass 9 × 5 mm. [Color figure can be viewed in the online issue, which is available at [wileyonlinelibrary.com](http://wileyonlinelibrary.com).]

Chest pain is a common presenting complaint to the pediatric cardiology clinic and pediatric emergency room. The most common source of chest pain in children and adolescents is musculoskeletal in nature. The cardiac origin of chest pain is extremely low in children with no risk factors. The common identified cardiac etiologies include Kawasaki disease, pericarditis, aortic stenosis, hypertrophic cardiomyopathy, anomalous origin of coronary arteries, coronary cameral fistulae, or collagen vascular disease-induced coronary arteritis. On the other hand, acute occlusion of coronary arteries related to thrombus formation on top of atherosclerotic plaque disruption is the main pathophysiology of AMI in adult population. AMI is a rare etiology of cardiac chest pain in the pediatric population. To the best of our knowledge, this is the first reported case in the pediatric population of AMI due to left main coronary artery occlusion in the absence of identifiable risk factors that was successfully rescued by percutaneous coronary intervention.

ST-segment elevation in lead aVR and widespread horizontal ST segment depression maximally in leads V4–V6 have been recognized as a typical electrocardiographic finding of subtotal left main coronary artery occlusion with preserved flow [13]. ST-segment elevation in Lead aVR is associated with higher 30-days mortality independent of concomitant ST segment changes in other ECG leads [14]. It was also shown that there is a relationship between the amplitude of ST elevation in lead aVR and mortality. Lead aVR reflects the right upper side of the heart; right ventricle outflow tract and the basal part of the septum. Blood supply to the basal part of the anterior septum is provided by the proximal septal branches of left anterior descending artery. Ischemia to the basal part of the interventricular septum is the best explanation for the

occurrence of the ST segment elevation in lead aVR. Changes in aVR are sometimes ignored and considered as reciprocal changes from the left lateral side (aVL, V5, and V6). However, as shown in our patient, changes in aVR can be very useful in identifying acute left main coronary artery occlusion.

In conclusion, this case illustrates the importance of careful clinical examination and thorough evaluation of EKG findings with thorough attention to each ST segment in patients presenting with concerning chest pain symptom.

## REFERENCES

1. McAllister HA Jr. Primary tumours of the heart and pericardium. *Pathol Annu* 1979; 14:325–355.
2. Nadas AS, Ellison RC. Cardiac tumors in infancy. *Am J Cardiol* 1968; 21:363–366.
3. Simcha A, Wells BG, Tynan MJ, Waterston DJ. Primary cardiac tumours in childhood. *Arch Dis Child* 1971; 46:508–514.
4. Chan HS, Sonley MJ, Moes CA, Daneman A, Smith CR, Martin DJ. Primary and secondary tumours of childhood involving the heart, pericardium, and great vessels. A report of 75 cases and review of the literature. *Cancer* 1985; 56:825–836.
5. Chughtai A, Cronin, P, Kelly AM, et al. Cardiac pseudosarcomatous fibromyxoid tumor: A review of the literature. *J Comput Assist Tomogr* 2005; 29:749–751.
6. Dickens P, Lam AK. Sudden death associated with cardiac inflammatory pseudotumour. *Forensic Sci Int* 1991; 49:89–93.
7. Kelly SJ, Lambie NK, Singh HP. Inflammatory myofibroblastic tumor of the left ventricle in an older adult. *Ann Thorac Surg* 2003; 75:1971–1973.
8. Krishna L, Ng WL, Chachlani N. Inflammatory pseudotumor of the heart causing aortic regurgitation. *Ann Thorac Surg* 2001; 71:1361–1363.
9. Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. *Am J Surg Pathol* 1995; 19:859–872.
10. Burke A, Li L, Kling E, Kutys R, Virmani R, Miettinen M. Cardiac inflammatory myofibroblastic tumor: A “benign” neoplasm that may result in syncope, myocardial infarction, and sudden death. *Am J Surg Pathol* 2007; 31:1115–1122.
11. De Montpreville VT, Serraf A, Aznag H, Nashashibi N, Planche C, Dulmet E. Fibroma and inflammatory myofibroblastic tumor of the heart. *Ann Diagn Pathol* 2001; 5:335–342.
12. Arber DA, Kamel OW, van de Rijn M, Davis RE, Medeiros LJ, Jaffe ES, Weiss LM. Frequent presence of the Epstein-Barr virus in inflammatory pseudotumor. *Hum Pathol* 1995; 26:1093–1098.
13. Nikus KC, Eskola MJ. Electrocardiogram patterns in acute left main coronary artery occlusion. *J Electrocardiol* 2008; 41:626–629.
14. Wong C-K, Gao W, Stewart RAH, et al. aVR ST elevation: An important but neglected sign in ST elevation acute myocardial infarction. *Eur Heart J* 2010;31:1845–1853.