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## Left mediastinal pleural and left pericardial agenesis, diagnosis, complications and management

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### Abstract

We present a case of 69 yrs old man with history of coronary artery disease who required coronary artery bypass graft surgery and is noted to have incidental detection of complete absence of left mediastinal pleura and left pericardium with levocardia and multiple adhesions which were dissected out and adhesions cleared. Although complete pericardial absence is rare with reported incidence of 1 in 14,000 patients, left mediastinal pleural and pericardial combined agenesis is even rare and very few cases have been reported in literature. We describe the technical difficulties, operative diagnosis and complications.

**Keywords:** Pleural pericardial agenesis, adhesions of heart, case report, adult, surgical

### Introduction

Left pleuropericardial agenesis is a defective developmental abnormality due to failed development of left pleuropericardial membrane and closure of left pleuropericardial canal [2].

Most patients are asymptomatic and they are generally diagnosed during surgical procedures and may present with technical difficulties during surgery. We present a case of incidental detection of combined left pleural pericardial agenesis with adhesions of heart and lung in a patient who presented with coronary artery disease requiring coronary bypass surgery.

### Case Report

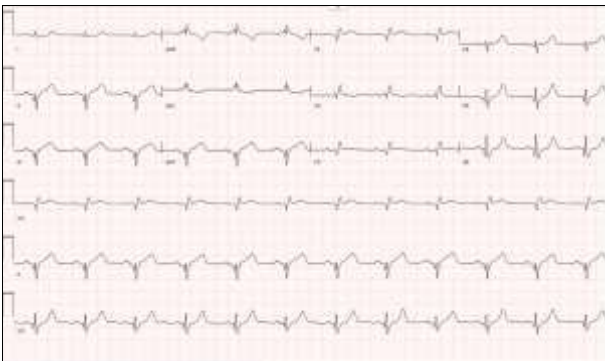
69 yr old gentleman with a past medical history of hypertension, tobacco use who presented with stable angina. Patient had an abnormal myocardial perfusion study showing inferolateral ischemia and left heart catheterization showing critical ischemic heart disease requiring surgical vascularization. Patient denied any history of previous trauma or chest surgery or chest infections. Electrocardiogram showed the right bundle branch block with right axis deviation. Chest x ray showed left heart displacement. CT scan of chest confirms left heart deviation. During surgery following sternotomy and placement on cardiopulmonary bypass multiple adhesions were noted between pericardium and heart and between lung and heart and noted complete absence of left mediastinal pleura and left pericardium. Right mediastinal and pericardium were noted to be intact. Subsequent bypass surgery times two with left internal mammary artery was performed uneventfully and the patient recovered successfully without any perioperative complications.



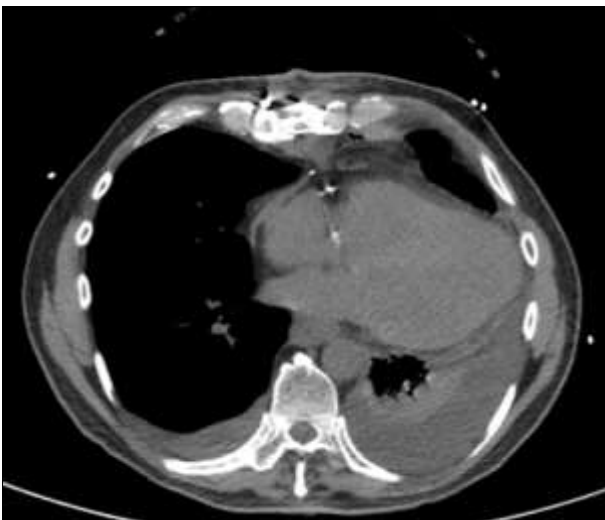
**Fig 1:** Chest x-ray showing levorotation of the heart



**Fig 2:** Post operative chest x-ray again confirming levorotation of the heart attached to the chest wall



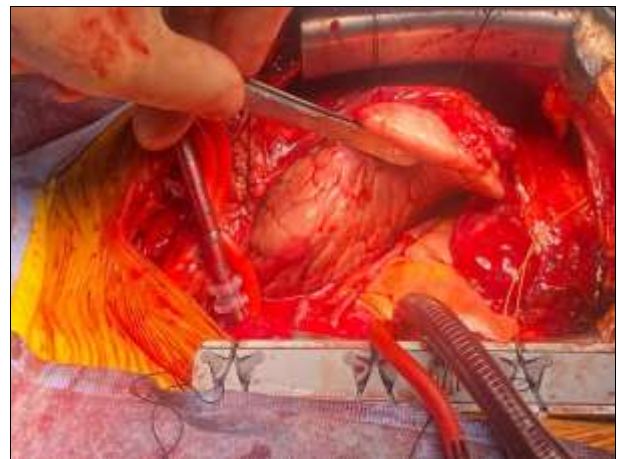
**Fig 3:** EKG showing right bundle branch block



**Fig 4:** Axial CT image showing left heart attached to the wall



**Fig 5:** Coronal image showing left heart attached to the wall again



**Fig 6:** Demonstrating intraoperatively Pleuro pericardial agenesis



**Fig 7:** Demonstrating intraoperatively Pleuro pericardial agenesis

### Discussion

Left pleural pericardial agenesis is a rare embryonic developmental abnormality. Complete absence of pericardium is reported to have incidence of about 1 in 10,000 to 14,000. Out of 364 cases isolated pericardial agenesis was found in 70% of cases on left side, 17% inferiorly and 4% on the right side [4, 5]. Although most of the cases are asymptomatic there have been rare reports of left atrial appendage incarceration and sudden death.

During normal embryonic development, the left duct of Cuvier is smaller than the right duct and is manifest usually later in adult life as Vestigial oblique vein of left atrium and ligament of left vena cava and therefore failure of left pleuropericardial canal is more common where left lung and heart share a common coelomic cavity due to failure of the closure of left pleuropericardial opening <sup>[15]</sup>.

Preoperative diagnosis of pleuropericardial agenesis is rarely made. On physical exam on supine position the apical impulse is more medial and shifts more lateral than usual on left lateral position and the exaggerated positional displacement maybe due to swinging motion of heart in systole. A systolic left 2<sup>nd</sup> space murmur can be audible as well.

Radiologic imaging including Chest X ray and CT scan suggests levocardia and becomes worse on the with patient on left lateral decubitus position <sup>[9-11]</sup>. Visualization of ligamentum arteriosum on CT scan is considered by some to be a characteristic sign for left pericardium congenital absence. MRI scan is also confirmatory.

### Complications

The rare recognized complications of left pericardial agenesis include incarceration of heart tissues and coronary artery disease <sup>[12, 13]</sup>. Patients with small or completed defects of pericardium do not need usually any intervention. However, patients with partial pericardial agenesis are at risk of herniation leading to fatal myocardial strangulation and death <sup>[1]</sup>.

### Management

Surgery is recommended in rare instances of preoperative diagnosis of partial pericardial agenesis to avoid future risk of strangulation. Coronary artery bypass graft surgery may be difficult due to the increased distance between LIMA and LAD due to cardiac malposition <sup>[11, 14]</sup>.

### Conclusion

Left combined pleuropericardial agenesis is a rare condition which is almost always diagnosed intraoperatively but increased use of Cardiac CT for coronary artery calcification detection, may lead to increased preoperative detection of possible pleuropericardial defects. This may require approaching the mediastinum with caution, in view of difficult anatomy and modifying the surgical technique appropriately to avoid any tears of the organs and complications.

The implications of the absence of pleuropericardial may be somewhat limited in coronary artery bypass grafting because of a controlled situation in which the surgery is taking place. The presence of cardio pulmonary bypass gives a safety net in conduction of procedures related to heart. However more caution is advised during the surgeries of the lung and the esophagus as they are not done under cardio pulmonary bypass. Anticipation and proper planning is much more important in these procedures because of the risk of catastrophic bleeding from poorly defined tissue planes. Also staging of lung cancer can present challenges from lack of parietal and visceral pleura, which leads to chest wall invasion from earlier stages.

### Conflict of Interest

Not available

### Financial Support

Not available

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