

## Partial Pericardial Defect in an Asymptomatic Patient and Brief Review of Literature

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### Abstract

Congenital absence of pericardium is rare. Pericardial defects can be complete or partial. Partial pericardial defect may be symptomatic. We report a rare case of partial pericardial defect in an asymptomatic patient which was suggested by chest radiograph and confirmed by computed tomography and brief review of literature is also presented.

**Keywords:** Asymptomatic patient, Partial pericardial defect

### Introduction

Congenital absence of pericardium is a rare entity.<sup>1</sup> Pericardial defects can be complete or partial. Most of them are left sided and asymptomatic. The diagnosis of absent pericardium is usually made at surgery or autopsy.<sup>1, 2, 3</sup> We report a rare case of partial pericardial defect in asymptomatic patient which was suggested by chest radiograph and confirmed by computed tomography and review literature.

### Case description

A 80 years old female patient reported in Radiology Department for routine chest X-ray examination for some other reason. She did not have any chest complain. General and systemic examinations were

unremarkable. Chest radiograph postero-anterior view (Fig. 1) revealed a focal bulge in the middle of the left heart border. One of the differential diagnosis as focal pericardial defect was suspected and Echocardiography was done which revealed defect of pericardium in left heart border. Rest of Echocardiographic findings was unremarkable. Non-contrast Computed Tomography (CT) of the thorax (Fig.2) revealed incomplete pericardial defect on left side. The patient is on regular follow up for last 1 year and is doing well without any complication.

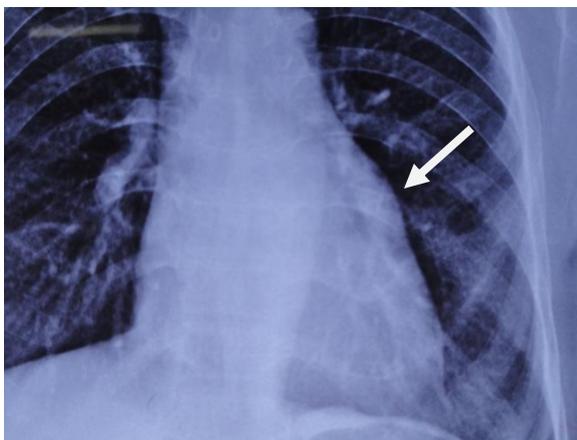
### Discussion

Congenital absence of pericardium is rare. Southworth H and Stevenson CS found only 1 case of pericardial defect in over 14000 autopsy performed at the John Hopkins Hospital.<sup>1</sup> It occurs more commonly in men than women, however our reported patient is a female. Pericardial defects may be complete or partial. Most pericardial defects

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are partial and occur on the left side. Occasionally, defects may occur on the right side or at the diaphragmatic surface.<sup>4</sup>



**Fig 1: Chest X-ray PA view showing prominence of mid left heart border.**

Defective closure of the pleuropericardial membranes lead to the congenital absence or partial defect of the pericardium. Different theories have been proposed to explain why the pericardial defect is common on left side. The most accepted theory is the one proposed by Perna and by Plaut. The left common cardinal vein tends to atrophy and is represented by coronary sinus in adult. The right common cardinal vein finally develops into the superior vena cava. Premature atrophy of left common cardinal vein results in deficient blood supply to pleuropericardial membrane, thereby, causing its incomplete development.<sup>5, 6, 7</sup>

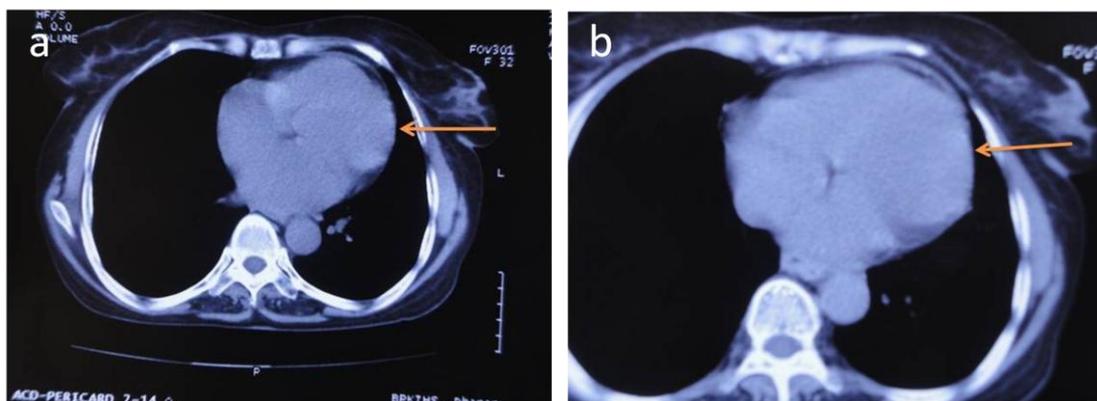
Congenital pericardial defects are frequently asymptomatic and discovered only during autopsy or thoracotomy.<sup>8</sup> The most common presenting symptom is chest pain. The exact mechanism of chest pain has not been well defined. Myocardial ischemia due to impingement of fibrous pericardial rim on coronary arteries is one of the cause of the chest pain.<sup>9</sup> Dyspnoea, syncope are other

symptoms that have been reported.<sup>10</sup> Partial pericardial defect may also cause sudden death of the patient.<sup>11</sup> Patients who have a pericardial defect with associated congenital abnormalities are often symptomatic and have one or more abnormalities, including atrial septal defect, patent ductus arteriosus, mitral valve stenosis, or tetralogy of Fallot.<sup>4</sup> Pericardial defect may be associated with bronchogenic cyst or diaphragmatic hernia.<sup>12,13, 14</sup>

In the past, the diagnosis of pericardial defect was often established on chest radiography by iatrogenic induction of left pneumothorax which has become obsolete now days. Spontaneous pneumothorax, if present on left side, aid in its diagnosis.<sup>15</sup>

Chest radiograph often raises suspicion as in our case. The findings of pericardial defect on chest radiograph include rotation of the heart into the left chest with the right cardiac border projected over the midline, prominent contour of the left cardiac border with accentuation of the convexities of the aortic knob, the main pulmonary artery and the left ventricle. There may be an area of hypotransparency between the aortic arch and the pulmonary artery, which is the result of the interposition of the pulmonary tissue at this level.<sup>6, 10</sup>

Differential diagnosis on plain film include atrial dilatation, mitral stenosis, and causes of enlargement of the pulmonary artery like idiopathic dilatation of the pulmonary artery, hypertension from any cause, pulmonary valvular stenosis, large left-to-right shunts (especially atrial septal defect) and rarely pulmonary arteriovenous aneurysm or ventricular aneurysm. Clinical features and echocardiography will be useful to exclude these conditions.<sup>16</sup>



**Fig 2: CT scan of the chest showing localized focal partial absence of pericardium on left side (a). Magnified view (b).**

Although radiographs may show evidence of this condition, a definitive diagnosis can be obtained with CT or MR imaging.<sup>17, 18, 19</sup>

Excessive myocardial displacement and a large difference in total heart volume in the end-systolic and end-diastolic phases on functional MRI have been proposed for diagnosis of congenital absence of pericardium.<sup>20</sup>

Complications of congenital pericardial defect may include herniation and entrapment of a cardiac chamber, especially the left atrial appendage. Therefore, prophylactic closure of partial pericardial defects has been recommended.<sup>21</sup>

Surgical closure or enlargement of the defect is sometimes necessary to alleviate herniation. Small defects require treatment whereas large and total absence of the left pericardium can be left untreated.<sup>7, 13</sup>

### Conclusion

Focal pericardial defect might impose diagnostic challenge on chest radiography. MDCT is useful modality to confirm its diagnosis. Early detection of partial pericardial defect is essential to avoid further complications.

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