

# Congenital defects of the pericardium: a review

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Pericardial defects are a rare disorder that can be characterized as acquired or congenital. Congenital defects can be further characterized by location and size of the defect, e.g. left- or right-sided and partial or complete absence of the pericardium. While physical examination and electrocardiogram are not diagnostic, chest radiographs and echocardiography have findings that should alert the clinician to the absence of the pericardium as a possible diagnosis. Despite its limitations with visualizing the normal pericardium in areas of minimal adipose, cardiac magnetic resonance is currently the gold standard for diagnosing the congenital absence of the pericardium. Patients have a similar life expectancy to those without pericardial defects; however in certain cases, herniation and strangulation of cardiac chambers can be life threatening and lead to sudden cardiac death. Treatment is tailored to the patient's symptoms, presentation, and the size and location of the defect.

**Keywords** pericardium • congenital • imaging

## Introduction

Pericardial defects are a rare disorder that can be characterized as acquired or congenital. Acquired absence of the pericardium is found after pericardiectomy to treat constrictive or recurrent pericarditis.<sup>1</sup> The congenital variant is exceedingly rare with a reported incidence of <1 in 10 000.<sup>2,3</sup> This comes from two large series in which only two cases were found in a study of 13 000 autopsies and only one case in a second study of 14 000 autopsies.<sup>3,4</sup> However, the true prevalence may be underestimated as most patients are asymptomatic, and diagnosis is generally incidental. The disease is found predominantly in males with a male to female ratio of 3:1.<sup>3,5</sup> Familial occurrence is rare.<sup>6</sup>

Congenital defects can be further characterized by location and whether there is partial or complete absence of the pericardium. Complete left-sided defects are most common with a reported prevalence of 70% of all pericardial defects.<sup>4</sup> Complete bilateral absence of the pericardium accounts for 9%, and right-sided defects comprise 17% of all defects.<sup>4,7</sup>

## Embryology

By the fourth week of embryonic life, a wedge of mesoderm has inserted itself between the primitive pericardial and peritoneal cavities, this is the septum transversum.<sup>8</sup> Lung buds grow and protrude into pericardio-peritoneal canals, which fold about them and form the pleural sacs.<sup>8</sup> By the fifth week, two-folds begin to grow out from the common cardinal veins (ducts of Cuvier) on each side; the ventral fold forms the pleuropericardial membrane, while the dorsal fold gives origin to the pleuroperitoneal membrane.<sup>9</sup>

These membranes fuse with the medial wall of the pleuroperitoneal canal on each side of the lung bud, resulting in separation of the pericardial and pleural cavities from the peritoneal cavity. Complete separation of pericardial and pleural cavities takes place when the pleuropericardial membrane unites with the root of lung.<sup>8,10</sup> Pericardial deficiencies are a result of failure of pleuropericardial membranes to fuse completely on one or both sides.

## Normal pericardium

The normal pericardium is an avascular fibrous sac composed of a double-layered inner serosal portion and an outer fibrous pericardium. The inner serosal layer has both visceral and parietal components. The visceral serous membrane has a single layer of mesothelial cells adherent to the epicardial surface of the heart.<sup>11</sup>

Serous fluid, ~15–50 mL, normally occupies the space between the visceral and parietal layers and acts as a lubricant.<sup>12</sup> The parietal pericardium lines the fibrous pericardium and the two are inseparable.<sup>13</sup> The fibrous pericardium is made of collagenous connective tissue and has ligamentous attachments to the diaphragm and sternum.<sup>11</sup>

In normal conditions, the pericardium is not visualized directly by echocardiography. Instead the thin, linear structure is outlined as a bright, echogenic line. Visualization by echocardiography becomes possible in cases when there is significant thickening of either pericardial layer.<sup>14</sup>

The computed tomography (CT) findings of the complete absence of the pericardium were first described in 1980, with the prominence of the main pulmonary artery and interposition of the left lung between the great vessels.<sup>15</sup> On cardiac CT (CCT), the normal

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pericardium is best seen in systole and appears as a line with average thickness of 1.3–2.5 mm. The pericardium is seen as a bright, linear structure that is seen in both contrast and non-contrast enhanced images because of its visibility against the low attenuation of surrounding adipose.<sup>14</sup> The thickness of epicardial fat increases in obesity and is typically asymmetric, with significantly more present along the right ventricle than the left ventricle.<sup>16</sup> Visualization of the pericardium can be difficult at the lateral, posterior, and inferior left ventricular wall due to the paucity of pericardial fat.

Cardiac magnetic resonance (CMR) of the heart uses electrocardiogram gating and acquires images during fixed segments of the cardiac cycle.<sup>17</sup> The pericardium appears dark and is seen as a curvilinear line of low signal intensity between the myocardium (medium intensity) and pericardial fat (high intensity), on both  $T_1$  (longitudinal relaxation time) and  $T_2$  (transverse relaxation time) weighted images. The low signal intensity is explained by the fibrous component of pericardium in combination with the small amount of pericardial fluid and is best seen in systole.<sup>17,18</sup> Improved systolic delineation of the pericardium in front of the right ventricle is probably due to inability of the parietal pericardium to move as much as the visceral pericardium during cardiac ejection, which is caused by its anchorage to the sternum anteriorly.<sup>17</sup> Only the parietal fibrous part of the normal pericardium is sufficiently thick to be potentially visualized by CMR and pericardial thickness can be defined at specific phases of the cardiac cycle.<sup>17</sup> The normal pericardium is 1–2 mm with upper limits of normal of 4 mm.<sup>18</sup>

## Normal pericardial function

The pericardium has been shown to have many physiologic and protective functions. By virtue of its ligamentous attachments, the pericardium helps stabilize and maintain the heart's position within the thoracic cavity.<sup>19</sup> Pericardial fluid functions as a lubricant and decreases friction of the cardiac surface during systole and diastole.<sup>20</sup> The pericardium acts as physical barrier and protects the heart from mechanical trauma and from the spread of infection from the lung and pleura.<sup>9</sup> The inelastic fibrous pericardium is also thought to impede pathologic overdistension of the heart.<sup>21</sup>

## Congenital absence of the pericardium

### Clinical presentation

Due to vague and atypical symptoms, the congenital absence of the pericardium can mimic other conditions such as acute coronary syndromes, cardiac aneurysms, tumours of the lung or heart, mitral valve disease, atrial septal defects, pulmonic stenosis, idiopathic dilation of pulmonary artery, and hilar lymphadenopathy.<sup>4</sup> The physical examination in patients with the congenital absence of the pericardium, whether complete or partial, is not characteristic or helpful in making the diagnosis.<sup>22</sup> In general, the absence of the pericardium is of little clinical significance as most patients are asymptomatic and defects are incidentally found on imaging, in the operating room or at autopsy.

The electrocardiogram is not specific for this condition but typical findings include right-axis deviation with incomplete right bundle branch block and bradycardia from vagal stimulation.<sup>6,23</sup> Historically, a

diagnostic pneumothorax was used to diagnose the absence of the pericardium by seeing a resultant pneumopericardium.<sup>24</sup> However, with introduction of less invasive and more sensitive techniques, this has fallen out of favour. Many of the echocardiographic findings seen in patients with the absence of the pericardium are also found in other pathologic conditions such as right ventricular volume overload, atrial septal defects, tricuspid and pulmonic regurgitation, and anomalous pulmonary venous drainage.<sup>25</sup> Echocardiography may not be diagnostic but certain findings should make clinicians consider this as a diagnosis.

CMR is currently the gold standard for evaluation of the pericardium as it has the best contrast resolution.<sup>9</sup> It provides a large field of view and spatial resolution that allows for examination of the entire chest and detection of associated abnormalities in the mediastinum and lungs. CMR is highly sensitive and can demonstrate the absence of the preaortic pericardial recess that is present in normal hearts.<sup>26</sup> While CMR can generally identify the pericardium, visualization of the pericardium varies with location as it does with CCT.<sup>14</sup> As a result, lack of visibility of the pericardium on CMR does not prove the absence of the pericardium and can lead to an erroneous diagnosis in 10% of patients.<sup>6</sup>

## Associations

Thirty to fifty per cent of patients with the congenital absence of the pericardium have associated congenital anomalies. Cardiac anomalies include atrial septal defects, patent ductus arteriosus, mitral valve disease, tetralogy of fallot, and sinus venosus defects with partial anomalous pulmonary venous drainage.<sup>18</sup> Pericardial defects have also been seen in patients with aortic connective tissue disorders and characteristics suggestive of Marfan syndrome.<sup>6,27</sup> In addition, several cases of type A aortic dissection have been seen in patients with the absence of the pericardium. These patients may not present with obstructive shock from cardiac tamponade, but instead, they can present with a large left haemothorax, no pericardial effusion and may be haemodynamically unstable from hypovolemic shock.<sup>28–30</sup>

Early atrophy of the left or right duct of Cuvier may lead to abnormal budding of the lung, resulting in bronchogenic cysts, sequestered lungs, and aberrant lobes.<sup>31</sup> Additional non-cardiac anomalies include pectus excavatum and diaphragmatic hernia.<sup>4</sup> Pericardial defects have also been noted in patients with VATER syndrome (vertebral defects, anal atresia, tracheoesophageal fistula, and radial and renal dysplasia),<sup>32</sup> and Pallister–Killian syndrome, the latter caused by tetrasomy of chromosome 12p and characterized by mental retardation, hypo- or hyper-pigmentation, and facial anomalies, including a high rounded forehead, short nose, and neck.<sup>33</sup>

## Complete bilateral and complete left-sided absence of the pericardium

The complete absence of the pericardium is the least likely to cause complications. Patients who experience symptoms generally have atypical chest pain secondary to tension from pleuropericardial adhesions, lack of pericardial cushioning, and undue torsion or strain on the great vessels as without a pericardium, they serve as the only anchor for the heart.<sup>4,22,34</sup> Resolution of pain after surgical

correction that leads to immobilization of the heart help support that the chest pain patients experience is related to heart mobility.<sup>35</sup>

In patients with the complete absence of left pericardium, apical displacement of the point of maximal intensity may be noted on physical exam. Many case reports have noted a systolic ejection murmur at the left sternal border which is thought to be due to turbulence set-up by various mechanical deformities at the base of an unusually mobile heart.<sup>4</sup>

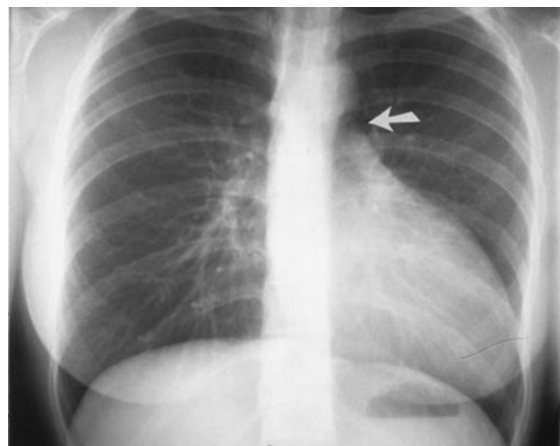
On electrocardiogram, poor R wave progression can be seen and is due to the leftward displacement of the precordial transitional zone. Prior reports have noted electrical alternans, alteration of both P and QRS vectors, which is likely due to increased mobility and unusual intrathoracic position of the heart.<sup>20</sup>

Chest radiographs have more specific findings that can alert the physician to consider the diagnosis of the absence of the pericardium. It was not until 1959 that the characteristic findings on plain chest radiograph were first recognized and described.<sup>36</sup> Patients with the complete absence of the left pericardium will have a leftward and posterior shift of the cardiac silhouette, straightening and elongated left heart border (Snoopy sign),<sup>6</sup> loss of the right heart border (obscured by the spine), and radiolucent bands (lung tissue) between the aortic knob and main pulmonary artery and between the left hemidiaphragm and the base of the heart (Figure 1).<sup>11,35</sup> Despite all the cardiac rotation seen, this condition does not cause deviation of the trachea so cardiac changes as noted above with a midline trachea should prompt the physician to consider this condition.

One of the first findings appreciated on echocardiography in patients with the complete absence of the pericardium is that the standard views may not apply. On the traditional left parasternal view, more of the right ventricle is seen due to the left shift of the heart and, as a result, the patient may be falsely diagnosed with right ventricular dilatation. In the apical window, there is marked lateral displacement of the heart and the appearance of compressed atria with the patient in the left lateral decubitus position.<sup>14,37</sup> As a result, some find that images are better obtained in supine position rather than left lateral ducubitus.<sup>37</sup> Exaggerated movement of the heart may also be seen during stress echocardiography examination.

The typical findings with the complete absence of the pericardium are a 'teardrop' shape of the heart, a bulbous left ventricle, and elongated atria in the apical four-chamber view, because of the absence of normal tethering applied by the normal pericardium.<sup>6,38</sup> There is exaggerated cardiac motion, especially of the left ventricular posterior wall.<sup>39</sup> The characteristic findings on m-mode include right ventricular enlargement and abnormal septal motion. The paradoxical motion of the interventricular septum is due to the exaggerated posterior left ventricular wall motion which produces anterior displacement of the interventricular septum.<sup>19</sup> Evaluation of speckle tracking shows decreased left ventricular torsion in patients with the absence of the pericardium. However, longitudinal, radial, and circumferential strain and strain rates showed no significant abnormalities in patients with pericardial defects compared with controls, suggesting that the absence of the pericardium does not impact regional left ventricular myocardial function.<sup>40</sup>

While CCT can generally identify the pericardium, the most common sites of pericardial defects, left-sided, happen to correlate with the locations that are not ideally visualized on CCT.<sup>15,21</sup> Interposition of lung tissue between the aorta and pulmonary artery or



**Figure 1** Characteristic posteroanterior radiograph from a patient with the complete congenital absence of left pericardium. Note the marked levoposition of the cardiac silhouette, loss of right heart border, prominent main pulmonary artery, with borderline irregularities of the upper left heart border, and a 'tongue' of lung tissue interposing between the main pulmonary artery and aorta (arrow). Reprinted from *Annals of Thoracic Surgery*, Volume 69, Gatzoulis MA, Munk MD, Merchant N, Van Arsdell GS, McCrindle BW, Webb GD, Isolated congenital absence of the pericardium: clinical presentation, diagnosis, and management, 1209–15, 2000, with permission from Elsevier.

between the diaphragm and base of heart is a specific and diagnostic sign that is seen in patients with complete left pericardial defects or partial defects overlying these structures.<sup>35</sup> Leftward cardiac displacement usually seen with complete left pericardial absence.<sup>41</sup>

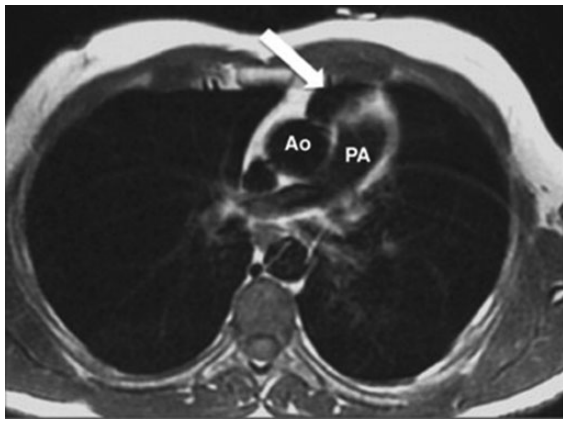
With the absence of the pericardium, CMR demonstrates leftward protrusion of left atrial appendage or main pulmonary artery, and lung insinuates between aorta and pulmonary artery and between base of heart and hemidiaphragm (Figure 2).<sup>42</sup> In addition, with the complete absence of the pericardium, one will see leftward and posterior rotation of the heart (Figure 3).<sup>9,35</sup>

## Treatment

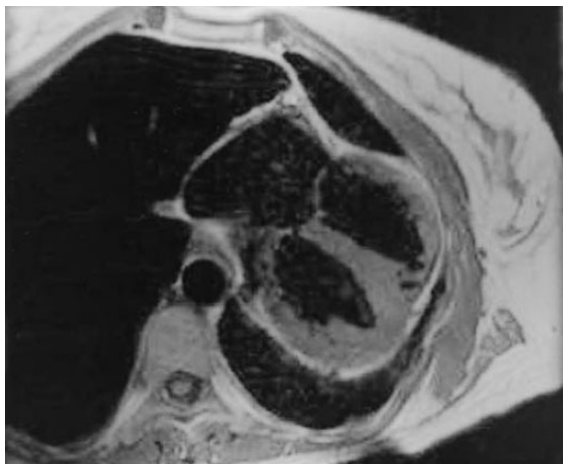
For patients with the complete bilateral or complete left-sided absence of the pericardium, no treatment is indicated. Studies of cardiac function have shown that patients with the complete absence of pericardium have similar ejection fractions to controls.<sup>4,40</sup> Patients with the congenital absence of the pericardium have the same life expectancy as those with a normal pericardium.<sup>3</sup> Although, some authors suggest that the cardiac displacement and increased mobility impose an increased risk for traumatic type A aortic dissections.<sup>43,44</sup>

## Partial absence of the pericardium

Patients with partial defects can have more symptoms and are at higher risk for complications than those with the complete bilateral or complete left-sided absence of the pericardium. They can have atypical chest pain described as stabbing, paroxysmal, non-exertional, and positional.<sup>22</sup> Vague chest pain is noted in one-third of patients.<sup>45</sup>



**Figure 2** Chest magnetic resonance imaging showing the interposition of lung tissue between the aorta and pulmonary artery (arrow). Ao, aorta; PA, pulmonary artery. Reprinted with open access from Santos JLA, Greguolo C, Junior JFF, Paiva AN, Trad HS, Andrade TTd. *Partial Congenital Absence of Pericardium*. *Rev Bras Cardiol Invasiva* 2012;20:435–7.



**Figure 3** Axial  $T_1$ -weighted spin-echo image demonstrates marked displacement of the heart into the left hemithorax with the cardiac apex pointing posteriorly. Reprinted from *Annals of Thoracic Surgery*, Volume 69, Gatzoulis MA, Munk MD, Merchant N, Van Arsdell GS, McCrindle BW, Webb GD, *Isolated congenital absence of the pericardium: clinical presentation, diagnosis, and management*, 1209–15, 2000, with permission from Elsevier.

### Partial left-sided defects

The left duct of Cuvier normally atrophies and in the adult is only represented by the coronary sinus.<sup>3</sup> Thus, it is proposed that left-sided defects are due to premature obliteration of the embryonic left duct of Cuvier. This leads to malnourishment of the left pleuropericardial membrane and incomplete formation of the parietal pericardium.<sup>18</sup>



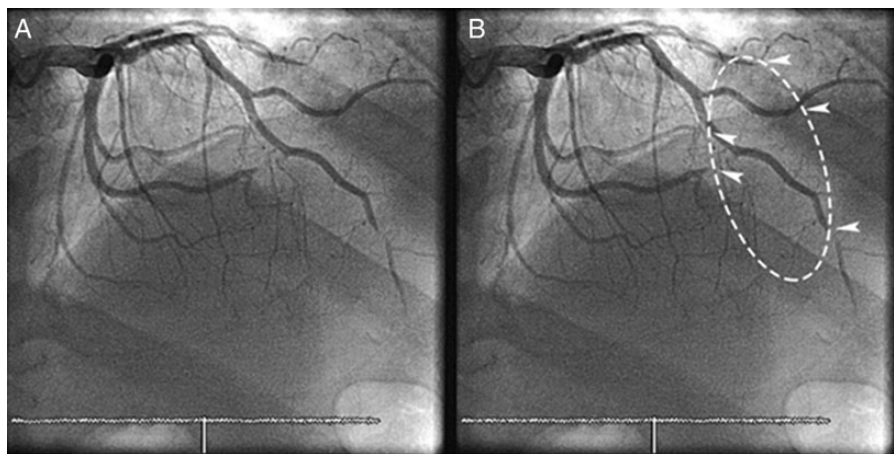
**Figure 4** Posteroanterior radiograph from an 8-year-old patient presenting with acute stabbing chest pain. Herniated left atrial appendage (arrow) is seen through a foramen-type partial absence of the left pericardium. Reprinted from *Annals of Thoracic Surgery*, Volume 69, Gatzoulis MA, Munk MD, Merchant N, Van Arsdell GS, McCrindle BW, Webb GD, *Isolated congenital absence of the pericardium: clinical presentation, diagnosis, and management*, 1209–15, 2000, with permission from Elsevier.

A subgroup of partial defects, foramen-type defects, increase the risk for herniation of the left atrial appendage, the entire left atrium, or both ventricles leading to cardiac strangulation or compression of coronary vessels.<sup>24,46–48</sup> These defects can be fatal as they may allow strangulation and necrosis of the heart.<sup>14,22</sup> Sudden death has been attributed to cardiac strangulation.

However, as in the case of complete absence of the pericardium, patients with partial left-sided defects are generally asymptomatic and do not experience complications. When symptoms do occur, they can be similar to those in patients with complete defects. Trepopnea, the presence of dyspnoea when lying on one side but not the other, is a unique finding that can be seen in patients with partial defects. Symptoms may be worse in the left lateral position because of volume loading of the ventricles (especially the right).<sup>6,49</sup> Chest pain in these patients can be due to external compression of coronary arteries by the rim of remaining pericardium.<sup>4,22</sup>

On chest radiography, a bulge of the left upper heart border or prominent pulmonary artery may be noted and may signify a herniation of the left atrial appendage through the defect (*Figure 4*).<sup>24,35</sup> In these cases, angiogram can be diagnostic as the prominence opacifies with contrast at the same time as the left atrium suggesting that the left atrial appendage herniated through a pericardial defect.<sup>50</sup> Thus, cardiac catheterization can be diagnostic in the presence of partial pericardial defects with herniation but generally is of no benefit with the complete absence or partial absence without cardiac herniation.<sup>50</sup>

In patients with partial defects and acute chest pain, ST segment elevation may be seen on electrocardiography. Cardiac catheterization in these cases can show a unique angiographic finding of normal caliber coronaries with good flow followed by an abrupt focal kink or



**Figure 5** Cardiac left heart catheterization showing marked stenosis (A, arrows in B) of the left anterior descending artery, diagonal arteries, and obtuse marginal arteries. The coronary arteries are otherwise of normal caliber and appearance. An ellipse (B) has been created demonstrating the appearance of what appeared to be either intrinsic or extrinsic compression of the coronary arteries. Figure reproduced with permission from Wilson SR, Kronzon I, Machnicki SC, Ruiz CE. A constrained heart: a case of sudden onset unrelenting chest pain. *Circulation* 2014;130:1626.

angulation in the arteries which is secondary to external compression of coronary arteries by the pericardial rim (Figure 5).<sup>22</sup>

The left atrium or left atrial appendage may herniate through the foramen-type defect which may be seen as an enlarged left atrial appendage<sup>51</sup> or left atrial aneurysm on echocardiography.<sup>52</sup> Occasionally, chambers of the heart may herniate when patients are in left lateral decubitus position but not when in right lateral decubitus position because of the intact right-sided pericardium and this can be seen as paradoxical septal motion on echocardiography. There also may be regional bulging of the left ventricle with partial defects.<sup>7</sup> In cases of left ventricular herniation, marked thickening of the apical left ventricle may be seen. Wall thickening is secondary to oedema from venous coronary obstruction.<sup>22</sup>

CMR can show prominent convexity along the left side of the cardiac silhouette in the area of the left atrial appendage, if the extension of the pericardial defect crosses the basal portion of the heart.<sup>17</sup>

## Treatment

Symptomatic patients with partial defects and cardiac chamber herniation should undergo surgery.<sup>6,35,48</sup> Treatment options include patch closure of the defect, enlargement of defect to prevent incarceration of herniated tissue, pericardiectomy, or pericardioplasty.<sup>13,35,48</sup> In cases of herniation of the left atrial appendage, the appendage can be excised and the pericardial defect can be closed with a patch.<sup>48</sup>

Total unilateral defects do not require treatment.<sup>48</sup> Many suggest surgery should be reserved for only highly symptomatic patients with partial defects.<sup>4,35</sup> Others suggest prophylactic closure of small partial pericardial defects to prevent the risk of herniation and strangulation.<sup>47,53,54</sup> This may be due in part to studies showing that pericardioplasty for symptomatic patients is safe, with a low morbidity.<sup>35</sup> If a thoracotomy is being done for other reasons, some suggest fixing a partial defect as the heart and great vessels tend to move within the

mediastinum.<sup>55</sup> For patients undergoing surgery, the phrenic nerve should be visualized as its course can be altered on the side with pericardial defect. We suggest that asymptomatic patients incidentally found to have defects of the pericardium be aware of the possible complications but be observed rather than have prophylactic cardiac surgery.

## Right-sided defects

The right duct of Cuvier normally persists as the superior vena cava, which tends to ensure closure of the right pleuropericardial membrane.<sup>56</sup> As a result, right-sided defects are much more rare.

On chest radiograph, patients may have a prominent bulge of the right heart border which may represent a right atrial appendage herniation.<sup>54</sup> On echocardiography, these patients can have an enlarged right ventricle and hypertrophied right atrium with severe tricuspid regurgitation due to traction of the chordal structures.<sup>57</sup>

## Other defects

Other defects are exceedingly rare. There has been one report of the absence of the anterior pericardium incidentally found during cardiac surgery.<sup>58</sup> In addition, the absence of the inferior pericardium can be seen and is associated with a rare type of diaphragmatic hernia, in which abdominal contents herniate into the pericardial cavity.<sup>20,59</sup> This defect is due to aberrant development of the septum transversum and is associated with a defect in the central tendon of the diaphragm. Diaphragmatic pericardial defects should be surgically corrected as these defects are associated with permanent risk of diaphragmatic hernia.<sup>60</sup>

## Conclusion

The congenital absence of the pericardium is a rare disorder generally of little clinical significance but in certain cases can be life threatening. The unique findings for each type of pericardial defect based on

location have been discussed. Clinicians should be aware of the specific findings on chest radiographs, echocardiograms, and angiograms as early recognition can be lifesaving. CCT or CMR can be obtained to aid in diagnosis and CMR stands as the gold standard to identify the congenital absence of the pericardium.

**Conflict of interest:** None declared.

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## IMAGE FOCUS

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# Two balls around aortic root: multiple huge unruptured aneurysms of the Valsalva sinus

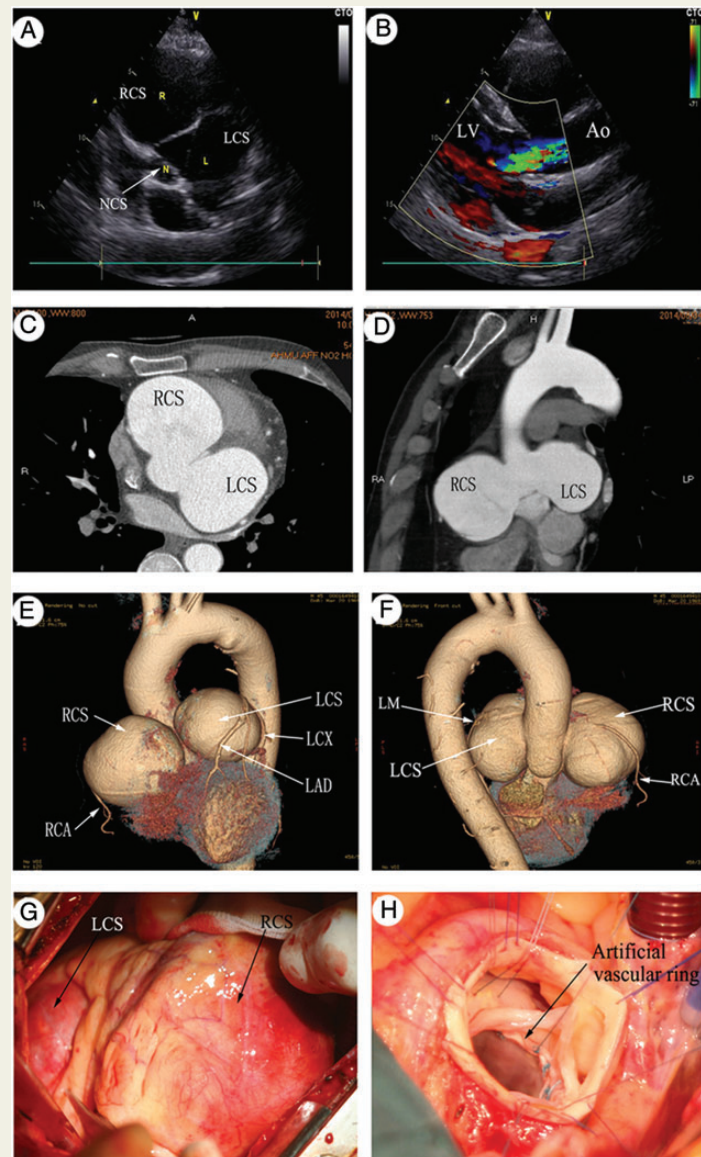
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A 45-year-old man presented with chest tightness and shortness of breath for 3 months, with no chest pain. Auscultation of the heart revealed a moderate diastolic murmur in the second to third left intercostal space. Transthoracic echocardiogram revealed two huge aortic sinus aneurysms [the largest diameter of the left coronary sinus (LCS) and right coronary sinus (RCS) was 56.5 and 56.8 mm, respectively] (Panel A, see Supplementary data online, Figure S1A) with moderate-to-severe aortic valve regurgitation (Panel B), a normal size of aortic root (diameter: 28 mm, see Supplementary data online, Figure S1B), and a 58% ejection fraction. Cardiac computed tomography (CT) and CT angiography (CTA) were performed to clarify the pathology. Horizontal (Panel C), coronal (Panel D), and 3D coloured volume-rendered CTA images displayed huge left and right coronary sinus aneurysms, like two balls around the aortic root (Panels E and F, see Supplementary data online, Figure S1E). The measured values of CT are shown in Supplementary data online, Figure S1C–E. Preoperative transoesophageal echocardiography showed the left and the right aortic sinus dilatation and aortic valve regurgitation (see Supplementary data online, Figure S1F). These findings were confirmed at operation (Panel G). Then, the patient was successfully performed with aortic valve annuloplasty (Panel H), aortic sinus repair, and coronary artery ostia graft.

Multiple huge unruptured SVAs are very rare. The inferred pathological mechanism was a weakness at the junction of the aortic media and the annulus fibrosus. Once huge SVA was diagnosed, it required surgical intervention immediately.



Supplementary data are available at *European Heart Journal – Cardiovascular Imaging* online.