

## Case Report

## Partial Right-Sided Pericardial Defect Associated with Congenital Aortic Valve Disease

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Partial right-sided pericardial defect is an extremely rare congenital anomaly and is often associated with other congenital abnormalities. We describe a unique case of congenital aortic valve disease associated with right-sided pericardial defect. The clinical implications are discussed and a review of the literature is presented.

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**P**ericardial defects are commonly related to structural anomalies of the lung and diaphragm.<sup>1</sup> They are also associated with other congenital anomalies of the heart.<sup>2-6</sup> We present a patient with the incidental finding of right sided partial defect of the pericardium during surgery for congenital aortic valve disease. To our knowledge, this is the first reported case of such an association. The clinical implications are discussed and the literature is reviewed.

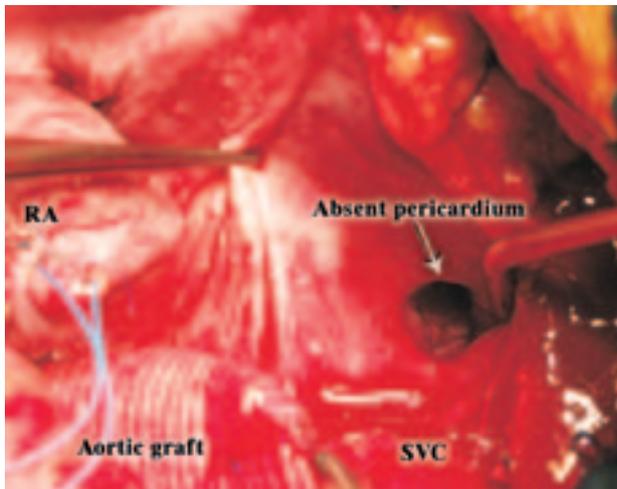
### Case presentation

A 38-year-old man with severe mixed aortic valve disease and ascending aortic aneurysm was admitted to our unit for surgical treatment. He presented with shortness of breath and had been followed from a young age for a bicuspid aortic valve. On auscultation a 3-4/6 murmur was noted at the aortic position. The chest X-ray showed increased heart size (cardiothoracic ratio 19/32). The ECG showed sinus rhythm and left ventricular hypertrophy (LVH). Echocardiography revealed LVH, severe aortic insufficiency (AI +4/ +4), left ventricular end diastolic pressure 70 mmHg, moderate aortic stenosis with peak gradient 55 mmHg (mean 33 mmHg), and dilatation of the as-

ending aorta (55 mm). Atrioventricular valves were normal and left ventricular ejection fraction was 50%. During operation, after the thymic-fat tissue was removed, a moderate-sized (2 × 2 cm) opening was found in the right upper part of the pericardium behind the upper third of the superior *vena cava*, leading to a sizable pocket of the right pleural cavity, outside the parietal pleura (Figure 1). The patient underwent replacement of the aortic valve and the ascending aorta, while the pericardial defect was left untouched as it did not pose any risk to the patient. He made a good overall recovery and was discharged home on the 10th postoperative day. He remains in excellent clinical condition 6 months later.

### Discussion

Congenital pericardial defects are rare and fewer than 200 cases have been reported in the literature.<sup>7</sup> They present as either complete or partial absence of the right or left side of the pericardium. They occur more often on the left side<sup>7,8</sup> and are more frequent in male patients.<sup>6</sup> Isolated absence of the pericardium is extremely rare and usually accompanies some other anomaly of the heart, lung or diaphragm.<sup>9</sup> Right sided pericardial defects are rare, representing



**Figure 1.** The aortic valve and ascending aorta have been replaced. Right sided pericardial defect.

less than 15% of all pericardial defects.<sup>1,2,6-8</sup> As many as one third of all cases present together with other congenital lesions, such as patent *ductus arteriosus*, tetralogy of Fallot, atrial septal defect, ventricular septal defect, absent superior *vena cava*, deficient sternum, bronchogenic cysts, pulmonary sequestration, and tricuspid insufficiency.<sup>1-6</sup> The fact that our patient also had a bicuspid aortic valve suggests a common embryological origin. However, to our knowledge this is the first report of such an association.

The embryogenesis of congenital pericardial defects has not been clearly defined. They are presumed to be the consequence of arrested development of the pleuropericardial membrane that eventually becomes the pericardium, due to atrophy of the duct of Cuvier. Depending on the degree of perfusion deficiency, partial or complete absence of the pericardium may be seen. This also explains the fact that right sided defects are so rare, because on the right side the duct of Cuvier persists as the superior *vena cava*.<sup>1,2,6,7</sup>

Pericardial defects are generally asymptomatic. Chest pain mimicking angina has been attributed to their presence.<sup>7,9,10</sup> For right-sided defects, intermittent right-sided chest pain has been reported. Exacerbation of this pain during deep inspiration should raise suspicion of right atrial herniation.<sup>8</sup> This is the only major complication that has been reportedly associated with these defects. Although lethal, it is an extremely rare complication that involves only partial defects in close relationship with the right atrium or right ventricle. Herniation of lung tissue into the mediastinum may present if the defect includes the parietal layer of the

pleura. Diagnosis is established by computed tomography scan and/or magnetic resonance imaging.<sup>8,10,11</sup>

Asymptomatic and complete defects should not necessarily be treated.<sup>6,7</sup> However, during cardiac operations dissection should be carried out with care in order to avoid injury to the misplaced phrenic nerve.<sup>1,12</sup> Partial defects, depending on their size and their position in relation to the cardiac chambers, should occasionally be treated in order to avoid herniation.<sup>6,7</sup> This applies particularly to the left side, where the heart is mobile. Treatment consists of either closure of the defect with a synthetic patch or enlargement of the opening to ensure that strangulation cannot ensue.<sup>6,7,9</sup> In our case, the patient was asymptomatic, the defect small and the parietal pleura intact. Furthermore, the opening was in a position that posed no threat for herniation of the heart or the lung.

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