Partial Congenital Absence of the Pericardium

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A 45-year-old male was admitted to the hospital with recurrent, stabbing, nonexertional chest pain. Chest x-ray showed an increased cardiothoracic ratio (Figure 1A), and ECG demonstrated sinus rhythm with incomplete right bundle-branch block and poor R-wave progression (Figure 1B). Echocardiography showed normal left ventricular dimensions and function; however, the right ventricle appeared grossly dilated with moderate to severe tricuspid regurgitation with no evidence of intracardiac shunts. These findings were confirmed by right heart catheterization, which also showed normal pulmonary and right atrial pressures without a pressure gradient over the pulmonic valve. Catheterization of the left heart showed normal ventricular pressures and function as well as normal coronary arteries. The patient was referred for cardiac magnetic resonance imaging (CMR) with suspected right ventricular cardiomyopathy. CMR showed cardiac displacement into the left hemithorax (Figure 2A). There was severe tricuspid regurgitation with marked dilatation of both the right atrium (6×8 cm) and ventricle (end-diastolic volume, 343 mL). Right ventricular function was mildly reduced (ejection fraction, 46%) without localized wall motion abnormalities. Left ventricular volumes and function were normal; however, there was marked indentation of the mid-lateral wall that resulted in herniation of the apical portion of the left ventricle without associated wall-motion abnormalities (Figure 2B). In addition, there was an extension of lung tissue between the inferior surface of the heart and the diaphragm as a result of the unusually posterior displacement of the left ventricle (Figure 3). Late enhancement images were normal, which made a diagnosis of right ventricular cardiomyopathy less likely.

Both displacement of the heart and interposition of lung parenchyma between heart and diaphragm are typical features of congenital absence of the pericardium, and herniation of the left ventricular apex suggests *partial* absence of the pericardium distal to the indentation of the left ventricular wall. Because of the severity of tricuspid regurgitation, the patient underwent tricuspid valve surgery, and at operation the partial absence of the pericardium was confirmed. The pericardial defect was repaired with a Goretex patch, and the tricuspid valve was replaced with a 33-mm biological (Perimount) prosthesis because a large defect in the anterior leaflet rendered it unsuitable for repair. Postoperative CMR (Figure 4) demonstrated successful restoration of normal left ventricular anatomy and normalization of right ventricular volumes (end-diastolic volume, 176 mL) with only mild tricuspid regurgitation. The patient recovered quickly from surgery and remains well.

Lateral and posterior displacement of the heart and interposition of lung parenchyma between heart and diaphragm are typical features of congenital absence of the pericardium.[1–3] Although CMR is useful in determination of anatomic abnormalities, the lack of visibility of the pericardium on CMR images does not prove its congenital absence and may lead to an erroneous diagnosis of pericardial absence in up to 10% of patients.[4] In the present case, the features of left ventricular indentation, cardiac displacement, and lung interposition between heart and diaphragm rather than direct visualization of the pericardium suggested the diagnosis, which was confirmed by surgery. In patients with absence of the pericardium, ECG often shows incomplete right bundle-branch block with poor R-wave progression as a result of the leftward displacement of the heart,[4,5] as seen in the present case. Congenital absence of the pericardium is a rare condition, and patients may either be asymptomatic or present with nonexertional stabbing chest pains. Approximately half the cases of congenital absence of the pericardium are associated with dilatation of the right ventricle, and other congenital cardiovascular and pulmonary abnormalities may be present.[5] The male to female ratio has been reported as 3:1.[5] Prognosis may be benign,[1] particularly in complete absence of the pericardium, but has not been adequately clarified. Surgical pericardioplasty (Goretex mesh) may only be considered for highly symptomatic patients[1] or in those patients who present with partial absence that leads to herniation or strangulation of cardiac and vascular structures.

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The online-only Data Supplement, consisting of a movie, can be found at http://circ.ahajournals.org/cgi/content/full/116/6/e126/DC1.

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Disclosures

None.

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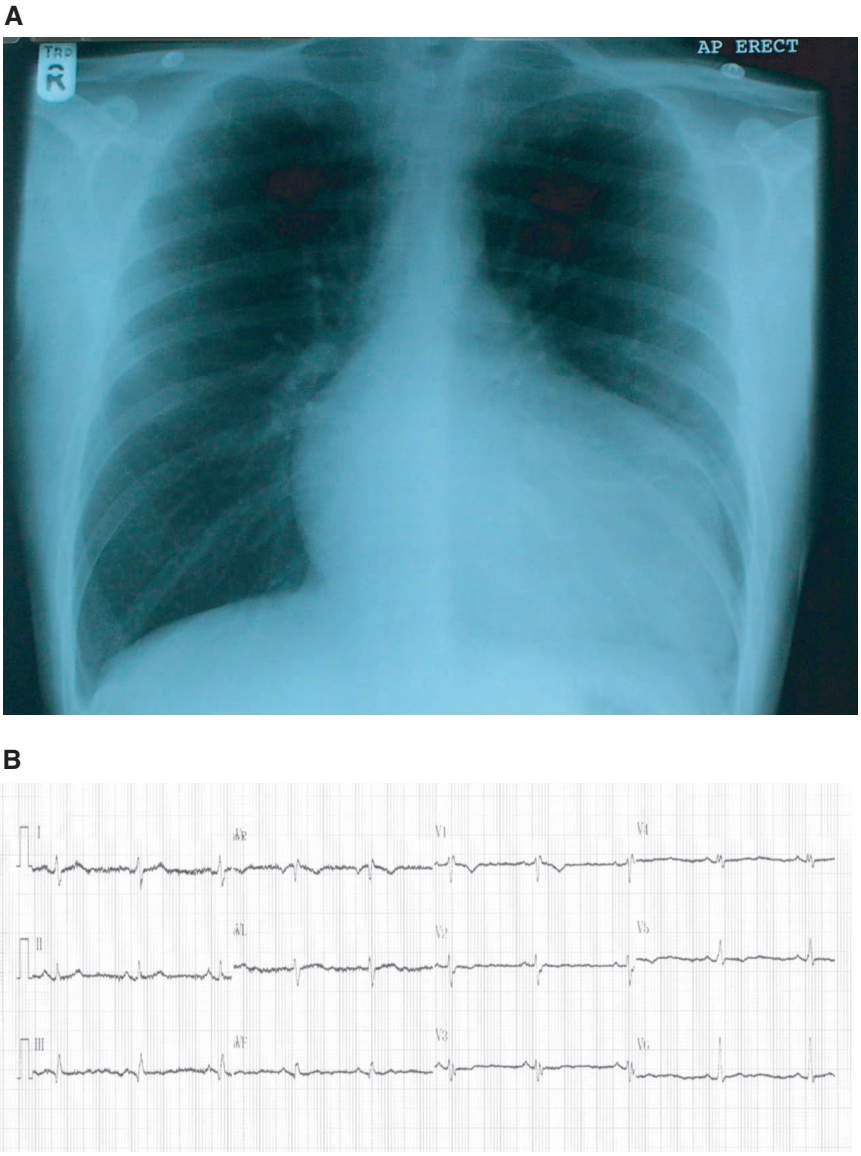

Figure 1. A, chest x-ray shows increased cardiothoracic ratio. B, Twelve-lead ECG shows incomplete right bundle-branch block and poor R-wave progression.

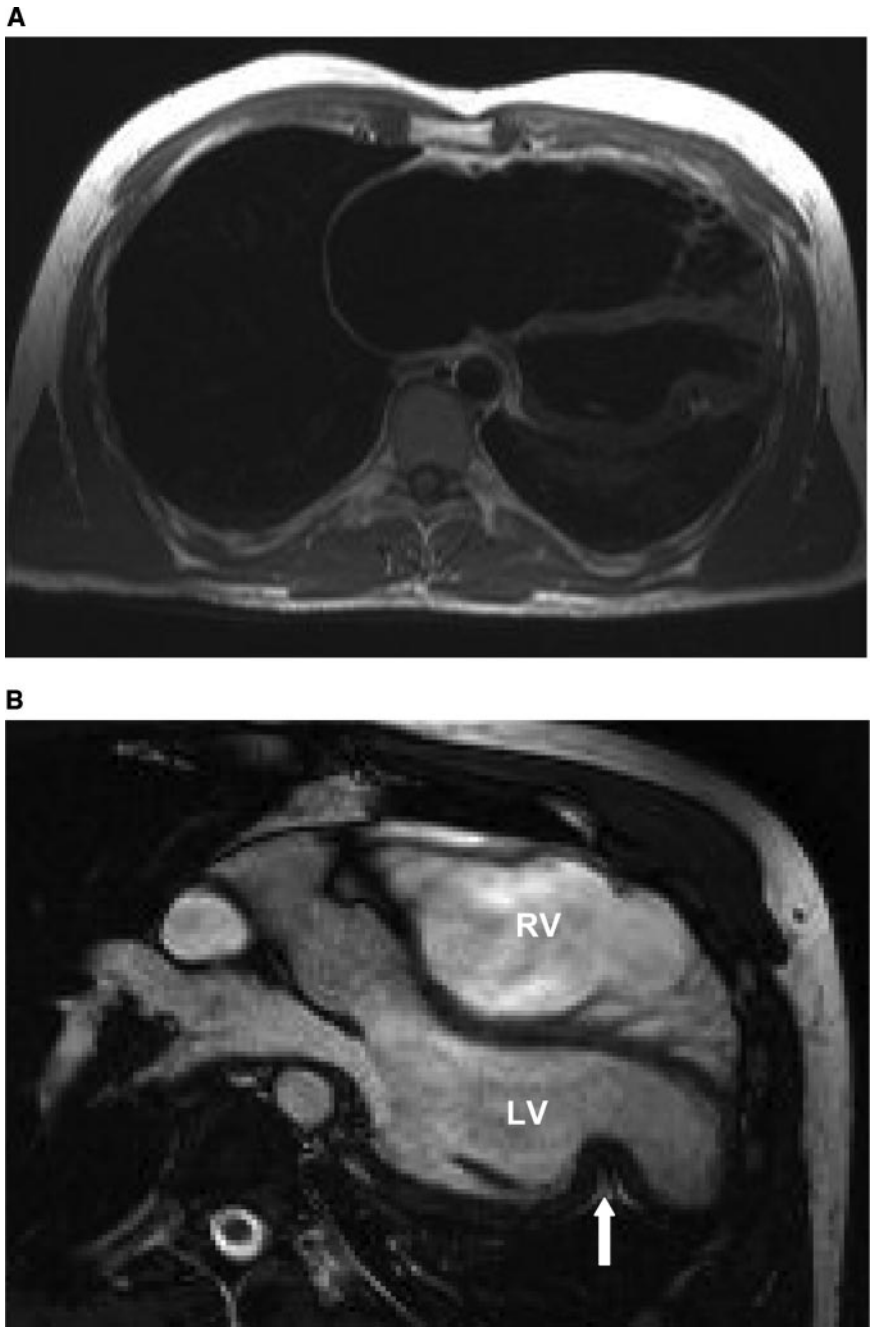

Figure 2. A, marked displacement of the heart into the left hemithorax with increase in right atrial and ventricular size. Image acquired with a cardiac-gated HASTE sequence (nonbreathhold; trigger pulse, 2; slice thickness, 7 mm) with a flip angle of 160°; TR, 800 ms; TE, 24 ms; and a matrix of 256×164. B, herniation (arrow) of the apical left ventricular wall. Image acquired with a cardiac gated True-FISP cine sequence (breathhold; slice thickness, 7 mm) with a flip angle of 60°; TR, 3 ms; TE, 1.51 ms; and a matrix of 256×164. See online Data Supplement for Movie. LV indicates left ventricle; RV, right ventricle.

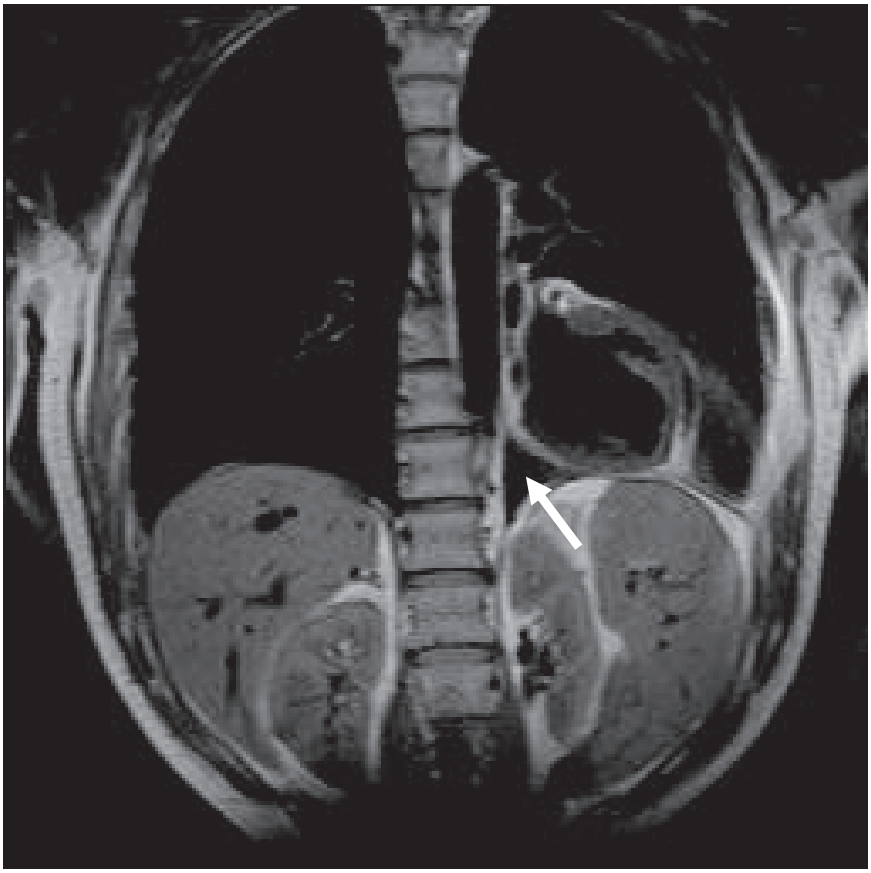

Figure 3. Interposition of lung tissue between the inferior surface of the heart and the diaphragm (arrow), typically seen in congenital absence of the pericardium as a result of the unusually posterior position of the heart. Image acquired with a cardiac-gated HASTE sequence as detailed in Figure 2A.

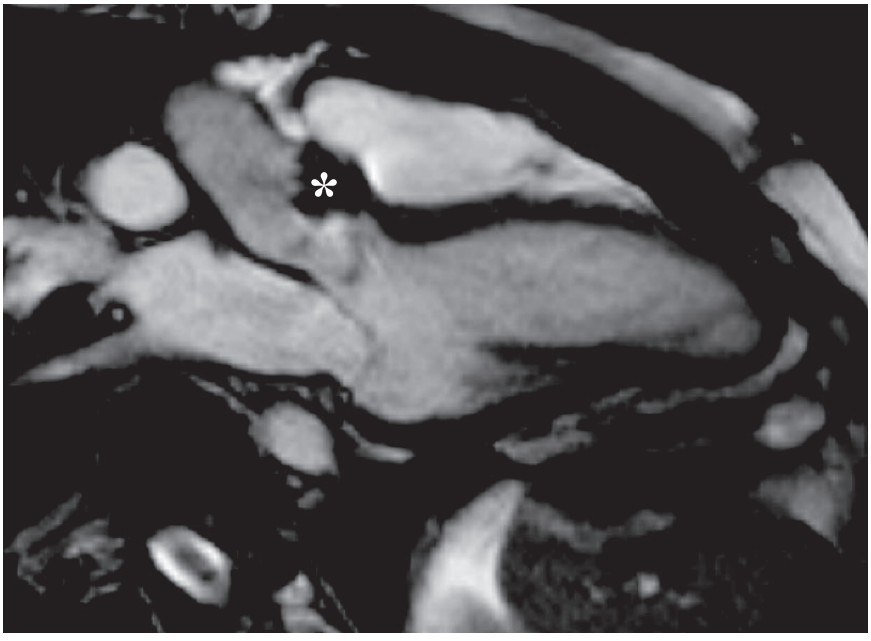

Figure 4. Restoration of the left ventricular anatomy in the same view as in Figure 2B. Note the considerable artifact from the metal frame of the tricuspid valve prosthesis (*). Image acquired with cardiac-gated TrueFISP cine sequence as detailed in Figure 2B.

Correction

In the article by Scheuermann-Freestone et al, "Partial Congenital Absence of the Pericardium," which was published in the August 7, 2007 issue of the journal (*Circulation.* 2007;116:e126–e129), the authors neglected to name a source of funding. Dr. Myerson received support from the Oxford NIHR Biomedical Research Centre programme.

The authors regret the error.

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