Diagnosis of congenital pericardial defects, including a pathognomonic sign for dangerous apical ventricular herniation, on magnetic resonance imaging

Ingmar Gassner, Werner Judmaier, Christoph Fink, Manfred Lener, Ferdinand Waldenberger, Helmut Scharfetter, Ignaz Hammerer

Abstract
Objective—To establish criteria for the accurate diagnosis of different forms of left sided pericardial defects on magnetic resonance imaging. Early detection of a partial apical defect is essential as it is potentially fatal.

Design—Examination of four children with congenital pericardial defects by magnetic resonance imaging, the results being compared with the features on conventional chest radiography and echocardiography and with published data.

Results—Magnetic resonance imaging improved the ability to diagnose and distinguish between complete and partial left sided pericardial defects. A deep myocardial crease was visualised in a patient with apical pericardial defect, indicating the risk of a life threatening ventricular strangulation. A prominent left atrial appendage was, in contrast to many reports, not a reliable sign for partial left sided pericardial defect.

Conclusions—The various forms of congenital left sided pericardial defects cannot reliably be diagnosed in plain chest radiographs or on echocardiography. Their diagnosis and the distinction between partial and complete defects, however, is of clinical importance and can be accomplished more confidently by magnetic resonance imaging.

Keywords: congenital pericardial defects; apical ventricular herniation; magnetic resonance imaging

The radiological features in chest radiographs of complete and partial pericardial defects have been described.1-3 Whereas a complete left sided pericardial defect has little importance for the wellbeing of the patient, a partial left sided pericardial defect can indicate a life threatening condition. The features in chest radiographs have, however, proved to be sometimes absent or misleading.2

The purpose of this study was to determine whether magnetic resonance imaging can be used to diagnose left sided pericardial defects and to identify the different types.

Patients and methods

We examined four children with congenital pericardial defects by magnetic resonance imaging, comparing the results with those on conventional chest radiography and echocardiography and with published data.

Magnetic resonance imaging was performed on a 1.5 T superconducting magnet (Magnetom, Siemens, Erlangen, Germany). Imaging variables included an acquisition matrix of 128 × 256 to 192 × 512, a field of view of 385 cm, four excitations, and a slice thickness of 5–8 mm with a gap of 0–2 mm between sections. Electrocardiographically triggered coronal and axial T1 weighted spin echo images were obtained with an echo time of 15 ms or 22 ms and a variable repetition time between 580 ms and 1100 ms, depending on the child’s RR interval.

Results

CASE 1

A boy presented at the age of 3 years with a murmur characteristic of persistent ductus arteriosus. A conventional chest radiograph showed a somewhat enlarged heart but no displacement to the left. Pulmonary vascularity was increased. A slight spherical bulge over the left hilum was interpreted as ductal diverticulum. At thoracotomy the duct had a diameter of about 5 mm without evidence of a ductal diverticulum. The main pulmonary artery and left pulmonary artery were dilated. A previously unsuspected complete left sided pericardial defect was discovered.

Only at the age of 13 years did a chest x ray film show all the criteria of a complete left sided pericardial defect, including leftward displacement of the heart. At this age echocardiography and magnetic resonance imaging were performed. The findings are summarised in the table.

CASE 2

A boy had a chest radiography at the age of 6 months because of a respiratory tract infection. At fluoroscopy a remarkable bulge in the left hilar region was identified as a strongly pulsating left atrial appendage. We assumed that herniation had occurred through a partial left sided pericardial defect.
### Summary of radiological and surgical findings in four patients studied

<table>
<thead>
<tr>
<th>Case No</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Chest radiography and fluoroscopy</th>
<th>Echocardiography</th>
<th>Angiography</th>
<th>Magnetic resonance imaging</th>
<th>Intra-operative findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Male</td>
<td>3</td>
<td>Slightly enlarged heart and increased pulmonary vascularity. Spherical bulging of the left hilum. No cardiac shift to the left. No fluoroscopy</td>
<td>Normal septal movement. Prominent left atrial appendage and main pulmonary artery. Horizontal position of the heart</td>
<td>Heart completely fallen into the left hemithorax. Left atrium and descending aorta side by side. Main pulmonary artery and left atrial appendage extending far beyond the normal margins of the mediastinum. Lung between aorta and main pulmonary artery and between inferior surface of the heart and the diaphragm</td>
<td>Persistent ductus arteriosus, dilated main pulmonary artery and left pulmonary artery, complete left sided pericardial defect. Straddling remnant of the right sided pericardium</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Male</td>
<td>10</td>
<td>Very prominent bulge of left atrial appendage and main pulmonary artery. Moderate cardiac shift to the left.</td>
<td>Normal septal movement. Prominent main pulmonary artery, marked enlargement of left atrial appendage. Horizontal position of the heart</td>
<td>Equivalent to case 1 at 13 yr. Left atrial appendage even more prominent</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Female</td>
<td>8½</td>
<td>Slight leftward shift of the heart. Bulging left atrial appendage. Fluoroscopy: abnormal cardiac mobility</td>
<td>Flat septal movement. Prominent left atrial appendage and main pulmonary artery. Horizontal position of the heart. Mitral valve prolapse</td>
<td>Equivalent to case 1 at 13 yr</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Male</td>
<td>12½</td>
<td>Spherical, normal sized heart. Obvious interposition of lung between diaphragm and heart. Fluoroscopy: no abnormal mobility of the heart</td>
<td>Dilated left atrium. Left ventricle: rounded with diminished contractility. Wedge shaped echogenic indentation of left ventricular myocardium</td>
<td>Left ventricle: outpouching in diastole, normal shape in systole. Left coronary artery diastole: obvious narrowing only during diastole</td>
<td>Left ventricular: myocardial crease. Bulging and elevated apex. Lung interposition between heart and diaphragm. Apical pericardial defect with deep myocardial crease</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td></td>
<td></td>
<td>Chest radiograph unchanged</td>
<td>Normal cardiac dimensions. No myocardial groove</td>
<td>No myocardial crease. Lung interposition unchanged</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

An angiocardiogram showed further evidence for this assumption: in atrial diastole the enlarged atrial appendage extended far laterally and ventrally. Because of the reported risks of herniation with subsequent strangulation of the left ventricle he was sent for thoracotomy. Intraoperatively a complete left sided pericardial defect was found, but this needed no correction.

A follow up chest x ray film 10 years later showed an extremely prominent left atrial appendage and a moderate leftward shift of the heart (figure 1). Echocardiography and magnetic resonance imaging were performed (figures 2–4; table).

**CASE 3**
A girl aged 8 years and 6 months who had been free of symptoms presented with a slight pectus excavatum and a heart murmur caused by an echocardiographically proved mitral valve prolapse. In a chest x ray film the middle shadow was somewhat dissociated to the left, and immediately below the pulmonary artery there was a bulge corresponding to the left atrial appendage on fluoroscopy. When she was moved to the left lateral decubitus position the heart was displaced and abutted the left thoracic wall, which was strongly suggestive of a complete left sided pericardial defect. This finding was even more apparent on magnetic resonance imaging.

**CASE 4**
A boy aged 12 years and 3 months had a history of decreasing physical fitness over nine months. After swimming and drinking a large
amount of a carbonated soft drink he suffered severe retrosternal stinging pain, which extended to the left shoulder and the left arm. In addition, he sweated profusely, felt nauseous, and repeatedly vomited. The pain worsened on deep inspiration.

An electrocardiogram showed signs of an anterolateral myocardial infarction. Activities of the heart muscle specific isoenzymes of creatine kinase and lactate dehydrogenase were raised.

In a chest x-ray film the normal sized heart was almost spherical, with interposition of lung between the diaphragm and the heart (figure 5). Fluoroscopy did not show an abnormal postural mobility of the heart. On echocardiography the left ventricle was remarkably round, with an apparent hypokinesia of the anterior wall. Apical to the sulcus atrioventricularis there was a wedge shaped area—with increased echogenicity indenting the left ventricular myocardium (figure 6).

In subsequent magnetic resonance tomograms we noted a striking elevation of the apex of the heart, well away from the surface of the diaphragm (figure 7), and, especially on axial images, a well defined indentation of the outer margin of the left ventricle. This pleat was most pronounced at the surface of the heart near the diaphragm and extended cranially along both the posterior and anterior wall. Abutting this sulcus was a collar-like accumulation of pericardial fat (figure 8).

Although these findings suggested ventricular herniation through an apical partial left sided pericardial defect, cineangiography was performed to ascertain the diagnosis. It showed an outpouching of the left ventricle, from its base to the apex, which was only visible in diastole (figure 9). In systole the shape of the left ventricle seemed to be almost normal. The left anterior descending coronary artery showed a subtotal occlusion in ventricular diastole shortly after its origin from the aorta.

Figure 2 Case 2. Coronal magnetic resonance tomogram. Because of the left and dorsal displacement of the heart the descending aorta and the left atrium abut each other (arrowheads). The absence of the pericardial layer allows the left atrial appendage to expand (arrow). The triangular signal loss below the inferior surface of the heart is caused by lung tissue taking the place of the pericardio-diaphragmatic junction (star).

Figure 3 Case 2. Axial magnetic resonance tomogram at the level of the great vessels. Main pulmonary artery (PA) and left atrial appendage (LA) are protruding far to the left. Lung is interposed between the aorta and the pulmonary artery.

Figure 4 Case 2. Axial magnetic resonance tomogram at the level of the ventricles. In the supine position the heart completely fell into the left hemithorax. The left atrium is wrapped over the aorta.

Figure 5 Case 4. Chest radiograph when patient was aged 12 years showing a spherical heart and interposition of lung between inferior cardiac surface and left hemidiaphragm.
Diagnosis of congenital pericardial defects on magnetic resonance imaging

Figure 6  Case 4. Echocardiogram (subxiphoidal approach) showing a pronounced groove (arrows) in the diaphragmatic surface of the myocardium. A, apex of the left ventricle.

Figure 7  Case 4. Coronal magnetic resonance tomogram (T1 weighted spin-echo) showing lung tissue separating the diaphragm and the heart. A groove is noted in the diaphragmatic portion of the left ventricular myocardium (arrow).

Figure 8  Case 4. Axial magnetic resonance tomogram at the apical level of the heart showing section comparable with that in figure 6, and myocardial groove (arrows). RV, right ventricle; A, apex of the left ventricle.

Figure 9  Case 4. Cineangiogram showing the left ventricle bulging from its base to the apex like an aneurysm during diastole.

Figure 10  Case 4. Aortic root injection. Narrowing of the lumen of the left anterior descending coronary artery is seen only in ventricular diastole (arrow).

with a normal diameter in systole (figure 10).

The diagnosis of a partial left sided pericardial defect with herniation of the left ventricle and diastolic compression of the left coronary artery due to the free edge of the pericardium was established. Intraoperatively the left sided pericardium proved to be absent in the apical region, thus the apex and the anterior and posterior wall lay outside the pericardial sac. Along the free rim of the defect there was a deep indentation of the myocardium with subepicardial haemorrhage. The left phrenic nerve was displaced ventrally and lay within the free edge of the pericardium. The pericardial gap was widened surgically and the phrenic nerve transposed dorsally.
Two years after surgery the patient was symptom free and the myocardial indentation was no longer identifiable on magnetic resonance imaging.

Results
The table summarises the radiological, echocardiographic, and operative findings in all our patients.

In all patients with a complete left sided pericardial defect axial MR tomography in supine position showed that the mobile heart had completely fallen into the left hemithorax (fig 4). Thus the left atrium was no longer preaortic but stretched around the descending aorta (figs 2 and 4). Lung tissue interposed between the aorta and the main pulmonary artery replaced the preaortic recess of the pericardial sac (fig 3). The pulmonary artery and the left atrial appendage clearly extended beyond the margins of the normal mediastinum, especially in case 2. The bulging of the left atrial appendage and the side by side position of the aorta and part of the left atrium were evident on coronal slices (fig 2). In addition coronal scans showed an area of signal loss in the triangle between the inferior surface of the heart, the aorta, and the diaphragm caused by interposed lung (fig 2).

In the MR tomograms of the patient with a partial left sided pericardial defect the heart showed no leftward displacement. We noted a striking elevation of the apex of the heart, away from the diaphragm (fig 7). A well defined, deep indentation of the outer margin of the left ventricle was visible, especially on axial images. This pleat was most pronounced on the diaphragmatic surface of the heart and extended cranially on both the posterior and anterior walls (figs 7 and 8). Abutting this sulcus was a collarlike accumulation of pericardial fat. At the level of the big vessels the preaortic recess was preserved.

Discussion
Congenital pericardial defects are rare. They are either complete or partial, occurring more often on the left than the right side. Bilateral complete defects or absence of a defect near the diaphragm is uncommon.

Patients with complete left sided pericardial defect are either asymptomatic or present with non-specific chest pain. No treatment is needed in patients with this diagnosis. Patients with partial left sided pericardial defect, however, are at risk of possibly symptomatic herniation of the atrial appendage or herniation of the left ventricle, eventually leading to fatal myocardial strangulation. Apical defects need to be detected as soon as possible because the time from the onset of symptoms of apical herniation and strangulation of the left ventricle to death varies from one hour to two days.

In cases 1 and 2 the diagnosis of complete left sided pericardial defect was missed in childhood because chest radiographs showed a prominence in the hilar region but no characteristic leftward displacement of the heart. This bulge was identified as a diverticulum of the persistent ductus arteriosus in case 1 and as a dilated and strongly pulsating left atrial appendage in case 2. Thus we suspected a partial left sided pericardial defect since in published reports protrusion of the left atrial appendage is considered pathognomonic for this defect. In both cases complete left sided pericardial defect was diagnosed only intraoperatively. In echocardiograms of patients with complete left sided pericardial defects Payvandi and Kerber and Nicolosi et al described a right ventricular dilatation and a paradoxical or flat movement of the septum, that was simulated due to the abnormal position and mobility of the heart. In our cases 1 and 2, however, M mode echocardiography gave normal results. In case 3 there was only a flat movement of the septum. In all cases the right ventricular dimensions were normal, whereas the cross sectional images showed an abnormal transverse position of the heart. The dilatation of the left atrial appendage was most pronounced in case 2.

Magnetic resonance imaging proved to be helpful in all our cases by showing directly all the criteria allowing differentiation of pericardial defects.

In complete left sided pericardial defect the heart has completely fallen into the left hemithorax in scans taken in the supine position owing to the absence of the supporting pericardial sac (figs 2 and 4). Thus the left atrium is wrapped over the descending aorta (fig 2). The preaortic recess is absent and its place is taken by lung tissue (fig 3). In coronal scans the lung interposed between the inferior surface of the heart, the diaphragm, and the descending aorta is particularly well visualised (figure 2). Both in partial left sided pericardial defects at atrial level and in complete defects the pulmonary artery and the left atrial appendage are uncovered by pericardium and thus bulge (figures 2 and 3). The presence of this bulge combined with a normal position of the heart in the supine position is thus evidence for the presence of a partial left sided pericardial defect in a more cephalad position. Our findings in complete left sided pericardial defects are in concordance with the few reported cases describing signs for this malformation on computed tomography and magnetic resonance imaging. In plain radiography and echocardiography these imaging techniques allow for a clear depiction of the anatomical relations unimpeded by overlying structures. In both computed tomography and magnetic resonance imaging, however, normal pericardium at the level of the atria and ventricles is seen only in the presence of sufficient epicardial and pericardial fat or fluid collection in the pericardial sac. Only the preaortic recess is regularly seen. The higher soft tissue contrast of magnetic resonance imaging offers a clear advantage over computed tomography, but adequate imaging quality is highly dependent.
Diagnosis of congenital pericardial defects on magnetic resonance imaging

on patient cooperation and sufficient electrocardiographic synchronisation of the measurement sequence. Motion induced imaging artefacts will be reduced with the introduction of echo planar imaging, in which the scanners have shorter acquisition times.

Case 4 describes the rare apical location of a partial left sided pericardial defect. Most authors say that in partial left sided pericardial defects the defect occurs in the hilar region behind the ventrally dislocated phrenic nerve, and they warn about the risk of fatal strangulation of the left ventricle.\(^1\) \(^2\) \(^3\) \(^4\) \(^5\) \(^6\) We emphasise, however, that in all lethal strangulations of the left ventricle described so far the partial defect was apical and the apex of the left ventricle was always strangulated; two cases showed additional herniation of the left atrial appendage.\(^7\) \(^8\) \(^9\) Although partial defects confined to the atrium can be symptomatic, no patients have died of them and only one case of strangulation of the left atrial appendage has been reported.\(^10\) Because of the different clinical impact we therefore have to distinguish between the various localisations of partial left sided pericardial defects.

To our knowledge, seven fatal cases of ventricular herniation through an apical pericardial defect have been published; reports at necropsy in six of them mention a deep crescent in the myocardium caused by the thickened, free pericardial rim.\(^1\) \(^2\) \(^3\) \(^4\) \(^5\) \(^6\) \(^7\) In one case the appearance of the heart at necropsy is not described.\(^7\) Our case 4 and three published reports show that this constriction of the myocardium exists even without strangulation. Bruning reported epicardial, pericardial, and myocardial fibrosis and fibrous thickening of the right coronary artery as clear evidence of a long term myocardial constriction before the fatal strangulation. In two other cases of intraoperatively diagnosed partial left sided pericardial defect a myocardial crease was also noted.\(^8\) \(^9\) \(^10\) \(^11\) Therefore this ring-like constriction of the myocardium at the level of the ventricles seems to be the key feature—that is, pathognomonic for life threatening apical pericardial defect.

In our case 4 this constriction ring was obvious on magnetic resonance images as a circumferential indentation of the myocardium along its diaphragmatic surface and the anterior and posterior wall (figures 7 and 8); it was also partially visible in echocardiograms (figure 6). Cineangiography showed in diastole the herniation of the ventricle and the compression of the left anterior descending coronary artery at the level of the free pericardial rim (figures 9 and 10). The diastolic narrowing of the left circumflex artery was previously observed only once by Wolff et al in a 27 year old man.\(^10\)

As in our patient, there is no pathognomonic chest radiographic appearance for the herniation of the left ventricular apex through a partial pericardial defect.\(^11\) \(^12\) \(^13\) \(^14\) \(^15\) Mainly the clear demonstration of the myocardial constriction groove by means of magnetic resonance imaging allowed for ascertaining this diagnosis in our patient. Apart from the myocardial indentation, the displacement of epicardial and pericardial fat along the compression site was easily visible on magnetic resonance imaging. These conditions exist long before the ventricular strangulation occurs as had been shown by histological and necropsy criteria.\(^6\) \(^7\) Thus we believe that magnetic resonance imaging should enable an early diagnosis, possibly in time for successful surgical repair.

Combining the findings in our patients with reports published so far we have five main conclusions.

(1) Magnetic resonance imaging with its multiplanar capability allows criteria for both a secure diagnosis and differentiation of left sided pericardial defects to be assessed in one examination.

(2) The complete laterodorsal displacement of the heart into the left hemithorax in supine position of the patient is pathognomonic for complete left sided pericardial defect.

(3) Contrary to previous reports the noticeable prominence of the left atrial appendage cannot be taken as a reliable sign of a partial left sided pericardial defect. It can be seen in both complete and partial left sided pericardial defects.

(4) The bulging of the left atrial appendage with absence of gross cardiac displacement is indicative of a partial left sided pericardial defect atrially. No fatal outcome has been reported in these defects, in which strangulation of the left atrial appendage can occur.

(5) A life threatening ventricular herniation may occur in a partial defect in apical location. The associated myocardial crease of ventricular herniation is pathognomonic and can be seen with modern cross sectional imaging techniques, especially magnetic resonance imaging.