Echocardiography in Congenital and Acquired Absence of the Pericardium

An Echocardiographic Mimic of Right Ventricular Volume Overload

MOHAMMAD N. PAYVANDI, M.D., AND RICHARD E. KERBER, M.D.

SUMMARY The purpose of this study was to investigate the echocardiographic effects of absence of the pericardium. Five patients with congenital complete absence of the left pericardium were studied. All had typical chest X-rays, four had cardiac catheterization which excluded any intracardiac shunts, and one had diagnostic pneumothorax. All five had an enlarged right ventricular dimension (RVD): 1.9 ± 0.1 cm/m² (normal: <1.3 cm/m²) and abnormal interventricular septal (IVS) motion (three Type A, two Type B). Sixteen additional patients were studied after pericardial stripping for a variety of conditions. In none was cardiac catheterization; one had diagnostic pneumothorax. After a cardiac history and physical examination, an electrocardiogram and cardiac X-rays in four views with barium and (in four patients) right and left lateral decubitus X-rays were obtained. Echocardiograms were performed in the supine position, using a Smith-Kline Ekoline 20A ultrasonoscope and 2.25 MHz transducer focussed at 7.5 cm and recorded on a Honeywell 1856 fiberoptic strip chart recorder. Interventricular septal motion was recorded at the choral level and classified as normal (posterior motion during systolic ejection), Type A (anterior or "paradoxic" motion during systolic ejection) or Type B (flat motion). Right ventricular dimension (RVD) was measured from the chest wall to the right side of the interventricular septum; 0.5 cm was subtracted to estimate the position of the right ventricular epicardium.

Group II

Sixteen patients were studied after pericardiotomy for a variety of indications: eight for recurrent pericardial effusion, five for constrictive pericarditis and three for subacute and chronic pericarditis, with persistent severe pain. Only two patients had calcification in the pericardium. No patient had continued symptoms referable to pericardial disease after surgery; and no patient had clinical, X-ray or electrocardiographic evidence of valvular regurgitation, intracardiac shunt, pulmonary hypertension or tricuspid insufficiency. These patients underwent the same evaluation as the group I patients, although not all had full cardiac series X-rays. All had postoperative echocardiograms; in eight, preoperative echocardiograms were available which were of quality sufficient to permit accurate assessment of septal motion and right ventricular dimension.

Results

Group I. Congenital Absence of the Pericardium

The findings in these patients are summarized in table 1. Two of the patients complained of left-sided nonexertional chest pain; one was aware of his heart beating against the

□ CONGENITAL ABSENCE of the pericardium is a well described but uncommon condition. In 1959, Ellis et al.1 noted that in the vast majority of cases partial or complete left-sided pericardial defects were present; right-sided or total pericardial absence is extremely rare. Although characteristic roentgenographic findings are usually present in congenital absence of the pericardium, clinically it may be mistaken for a variety of cardiac and noncardiac conditions. These include atrial septal defect, idiopathic dilatation of pulmonary artery, pulmonic stenosis, cardiac tumor, mitral valve disease, left ventricular aneurysm, corrected transposition of the great vessels, left hilar adenopathy, mediastinal tumor and bronchogenic carcinoma. Associated abnormalities of the heart and lungs may also occur. The diagnosis can be definitively established by inducing a left pneumothorax; air appears abnormally under the right pericardium when the patient is turned to the left lateral decubitus position.4

The purpose of this study was to evaluate the echocardiographic appearance of pericardial absence in order to determine whether the ultrasound technique could be used reliably to differentiate this condition from those which it clinically resembles, especially atrial septal defect. Two groups of patients were included: 1) patients with congenital absence of the pericardium and 2) patients with acquired absence of the pericardium.

Methods

Group I

Five patients with the previously established diagnosis of congenital absence of the pericardium were recalled for this study. Four of these five patients had previously undergone

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1. From the Cardiovascular Division, Department of Medicine and The Cardiovascular Center, University of Iowa College of Medicine and Veterans Administration Hospitals, Iowa City, Iowa.

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4. Address for reprints: Richard E. Kerber, M.D., Cardiovascular Division, Department of Medicine, University of Iowa Hospitals, Iowa City, Iowa 52242.

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**Table 1. Congenital Absence of the Left Pericardium**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Symptoms</th>
<th>Physical exam</th>
<th>Chest X-ray</th>
<th>ECG</th>
<th>Catherization</th>
<th>IVS motion</th>
<th>RVD (cm²/m²)</th>
<th>LVD (cm²/m²)</th>
<th>Echo</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>None</td>
<td>Axillary cardiac impulse; wide split S2, not fixed; grade II/V1 systolic ejection murmur in pulmonic area</td>
<td>Levoposition of heart; prominent pulmonary artery; radiolucent zones between aorta and PA and over diaphragm; excessive leftward cardiac mobility on decubitus</td>
<td>RAD, RBBB, clockwise rotation</td>
<td>Not performed</td>
<td>A</td>
<td>2.1</td>
<td>2.6</td>
<td>Echo</td>
</tr>
<tr>
<td>2</td>
<td>None</td>
<td>Axillary impulse; grade II/V1 systolic ejection murmur in pulmonic area</td>
<td>Same as in patient 1</td>
<td>Axis +90°, rSr' in V1, clockwise rotation</td>
<td>Normal — no shunt</td>
<td>A</td>
<td>1.6</td>
<td>3.2</td>
<td>Echo</td>
</tr>
<tr>
<td>3</td>
<td>Left chest pain, awareness of heart beat lying on left side</td>
<td>Axillary impulse; wide split S2, not fixed; grade I/V1 systolic ejection murmur left sternal border</td>
<td>Prominent pulmonary artery; radiolucent zones between aorta and PA and over diaphragm; excessive leftward cardiac mobility on decubitus</td>
<td>RBBB</td>
<td>Normal — no shunt</td>
<td>A</td>
<td>1.8</td>
<td>1.9</td>
<td>Echo</td>
</tr>
<tr>
<td>4</td>
<td>Left chest pain</td>
<td>Axillary impulse; grade II/V1 systolic ejection murmur in pulmonic area</td>
<td>Same as in patient 1</td>
<td>RAD, clockwise rotation</td>
<td>Normal — no shunt; induced pneumothorax; air under right pericardium</td>
<td>B</td>
<td>2.1</td>
<td>3.3</td>
<td>Echo</td>
</tr>
<tr>
<td>5</td>
<td>None</td>
<td>Axillary impulse; grade II/V1 systolic ejection murmur pulmonic area</td>
<td>Levoposition of heart; prominent pulmonary artery; radiolucent zones between aorta and PA and over diaphragm</td>
<td>RAD, clockwise rotation</td>
<td>Normal — no shunt</td>
<td>B</td>
<td>1.9</td>
<td>2.8</td>
<td>Echo</td>
</tr>
</tbody>
</table>

Abbreviations: RAD = right axis deviation; RBBB = right bundle branch block; PA = pulmonary artery; IVS = interventricular septum; RVD = right ventricular dimension; LVD = left ventricular diameter.
point of maximum cardiac impulse was in the axilla. The chest X-ray in each patient showed all or most of the findings characteristic of complete left pericardial absence\(^7\) (fig. 1): levoposition of the heart without tracheal deviation, prominence of the main pulmonary artery, interposition of lung tissue causing a radiolucent zone between the aorta and pulmonary artery and between the inferior border of the heart and the left hemidiaphragm. The right and left lateral decubitus X-rays obtained in four of the patients showed unusual cardiac mobility in the leftward direction. The lateral border of the heart rested against the chest wall in the left lateral decubitus position, but the heart showed little or no movement to the right (as compared with the frontal projection) in the right lateral decubitus projection (fig. 2). The electrocardiograms were also characteristic\(^8\), showing right axis deviation (+90\(^\circ\) or more) in four patients, incomplete or complete right bundle branch block in three patients and clockwise rotation (leftward displacement of the transition zone in the precordial leads) in four patients. Cardiac catheterization had been previously performed in four cases: no intracardiac shunts, valvular regurgitation or stenosis or other cardiac abnormalities were demonstrated in any patient. One patient had previously undergone diagnostic pneumothorax which showed air between the right border of the heart and the right pericardium.

Echocardiography was abnormal in each case (figs. 3 and 4). An enlarged right ventricular dimension (RVD) was present in all. The mean RVD for these five cases was 1.9 ± 0.1 (se) cm/m\(^2\) (range 1.6-2.1 cm/m\(^2\)). The upper limit of normal in our laboratory is less than 1.3 cm/m\(^2\). Left ventricular diameter at end-diastole was 4.8 ± 0.3 cm (2.7 ± 0.1 cm/m\(^2\)); diastolic left ventricular volume, obtained by cubing the left ventricular diameter, was 114 ± 18 cc, which is normal. The interventricular septal motion during systole was abnormal in all five patients. In each case

![Figure 1](http://circ.ahajournals.org/content.figures/1976/CIRCULATION-1976-0000088/v1-fig1a.png)

**Figure 1** Chest X-ray of patient 1 showing the characteristic abnormalities of congenital absence of the left pericardium: the heart is displaced to the left without tracheal deviation and the pulmonary artery is prominent. Interposed lung tissue, causing radiolucent zones (arrows) is seen between the aorta and pulmonary artery and between the inferior cardiac border and the left hemidiaphragm.

![Figure 2](http://circ.ahajournals.org/content.figures/1976/CIRCULATION-1976-0000088/v1-fig2.png)

**Figure 2** Right and left lateral decubitus chest X-rays of patient 1 demonstrating abnormal cardiac mobility to the left. In the right lateral decubitus position (A) the right cardiac border remains hidden behind the vertebral column, since the right pericardium is present and prevents excessive motion to the right. In the left lateral decubitus position (B) the left heart border lies against the chest wall, due to the absent left pericardium and loss of normal left-sided pericardial restraint. Compare with figure 1.
considerable variability in septal motion was noted so that at times the interventricular septum appeared to be moving anteriorly in systole ("paradoxical" or Type A), and at other times the systolic motion was essentially flat (Type B). This could usually be related to minor alterations in transducer position (always caudal to the mitral valve leaflets, however) or to respiration. The motion was classified in each case according to the pattern which appeared in the majority of the cycles which were technically satisfactorily recorded.

Group II. Acquired Absence of the Pericardium (Pericardiectomy)

Pre and postoperative echocardiographic measurements of right ventricular dimension and left ventricular diameter are presented in graphic form in figure 5. The right ventricular dimension increased after surgery in six of eight patients with adequate preoperative studies; and was enlarged in all of the eight patients in whom only postoperative studies were done. The mean right ventricular dimension increased from 1.0 ± 0.2 cm/m² to 1.7 ± 0.1 cm/m², P < 0.01. Left ventricular diameter, however, remained unchanged at 2.5 ± 0.1 cm/m². The interventricular septum moved normally in all eight patients in whom adequate preoperative echocardiograms were available (fig. 6). Fourteen of the 16 patients showed abnormal systolic motion of the interventricular septum after pericardiectomy (figs. 6 and 7), nine showing Type A motion and five Type B. Both of the patients with normal septal motion postoperatively had right ventricular dimensions of 1.5 cm/m²; the preoperative right ventricular dimension measured in one of the two was 1.3 cm/m².

FIGURE 3  Echocardiogram of patient 1, showing abnormalities which were seen in all five patients with congenital pericardial absence: abnormal interventricular septal motion in systole and enlarged right ventricular dimension. Abbreviations: RVD = right ventricular dimension, LVD = left ventricular diameter, IVS = interventricular septum.

FIGURE 4  Echocardiograms of patients 2, 3, 4 and 5 with absent left pericardium. Systolic septal motion is anterior or paradoxical (Type A) in patients 2 and 3 and flat (Type B) in 4 and 5. Right ventricular dimension was enlarged in all.
Discussion

This study indicates that such echocardiographic abnormalities as an enlarged right ventricular dimension and abnormal systolic septal motion are common when there is pericardial absence, either congenital or as a result of surgical removal of the pericardium. A possible explanation of the cause of these abnormalities is shown in figure 8. Due to the levoposition of the heart in patients with a congenitally absent left pericardium, as well as the posterior displacement of the heart in the supine position due to absence of the normal pericardial support, the ultrasound beam from a transducer placed in the usual parasternal position would tend to traverse the right ventricle tangentially and more in the mid-portion of the chamber. This would tend to yield a larger measured right ventricular dimension. The abnormality of septal motion may also be explained by the cardiac malposition and unusual mobility characteristic of congenital pericardial absence, which has been considered the cause of the chest pain commonly found in this condition (torsion of the great vessels). In the normal patient the apex of the left ventricle lies anteriorly. During systole there is a twisting motion of the heart, with a movement of the apex away from the anterior chest wall in the majority of cases, so that the LV chamber comes to lie more parallel with the A-P (i.e., frontal) plane at end-systole than at end-diastole. This systolic twisting motion is shown diagrammatically in figure 8A by the curved arrows pointing posteriorly. This motion tends to carry the septum posteriorly during systole. On the other hand, if the pericardium is absent the heart lies posteriorly when the patient lies in the supine position. The heart is freely mobile, and the apex probably twists in an anterior direction during systole since it is positioned well back in the thorax at the onset of ventricular contraction. As indicated by the arrows in figure 8B, this directionally abnormal twisting motion would tend to carry the septum forward in systole, resulting

FIGURE 5 Echocardiographic measurements of right ventricular dimension (RVD) and left ventricular diameter (LVD). After pericardiectomy the mean right ventricular dimension increased significantly, but the left ventricular diameter remained unchanged. In the patients with congenital left pericardial absence, the right ventricular dimension is large and similar to the postpericardiectomy patients; the left ventricular diameter is normal. Brackets indicate mean and standard error.

![Figure 5](http://circ.ahajournals.org/)

FIGURE 6 Pre and postpericardiectomy echocardiograms in a patient with constrictive pericarditis. The interventricular septal motion is normal preoperatively and flat (Type B) postoperatively, and the right ventricular dimension is enlarged after surgery.

![Figure 6](http://circ.ahajournals.org/)
in the observed abnormality of septal motion on echocardiography. Since the left ventricle has a basically spherical shape, its echocardiographic dimensions would be minimally affected by these abnormalities of position and motion.

Does this explanation also apply to the pericardiectomy patients? The general approach to pericardiectomy at the University of Iowa Hospital, as at most institutions, is to remove the entire left pericardium to the pulmonary veins, and the entire anterior pericardium as far as reasonably accessible on the right side, often just anterior to the phrenic nerve. This leaves pericardium posterior to the phrenic nerve on the right — i.e., over at least part of the right atrium as well as posteriorly over the left atrium. Thus some right-sided pericardial restraint remains while restraining forces on the left are essentially completely removed. This produces a situation analogous to congenital pericardial absence, since the latter always involves the left pericardium (partial or complete) but only very rarely the right. These statements may not apply, however, to the patient with a thick and/or calcified visceral pericardium where incomplete surgical decortication may result. (In only two of our 16 patients was calcified pericardium noted.)

Can the absence of the pericardial echo be observed directly? In the normal individual, reducing the sensitivity of the ultrasonoscope produces a single strong, moving posterior echo. Although commonly referred to as a pericardial echo, it is actually an amalgam of reflections from the interface of the pericardium and lung. In the absence of the pericardium, echoes similar to the pericardial echo were obtained, probably from the epicardium-lung interface (fig. 9). Although these signals appeared more vigorously pulsatile than usual, again reflecting the unusual cardiac mobility, this alone did not appear to be specific enough to permit diagnosis of pericardial absence.

A further reflection of the excessive cardiac mobility was encountered when we attempted to study the patients with congenital pericardial absence in a partial left lateral decubitus position. Although this position is widely used to enhance the quality of echocardiographic recordings, we found that it caused serious degradation of the ultrasound recordings in our cases; in several patients recordings could only be obtained in the supine position. Presumably the considerable additional leftward cardiac displacement produced by turning the patient (fig. 2) resulted in the beam striking many intracardiac structures tangentially rather than perpendicularly, with a consequent diminution in the strength of the reflected energy reaching the transducer.

The echocardiographic pattern of enlarged right ventricular diameter and abnormal septal motion seen in absent pericardium mimics that seen in conditions of right ventricular volume overload, atrial septal defect, partial anomalous pulmonary venous connection and tricuspid regurgitation. This is important, since atrial septal defect in particular shares many clinical findings in common with congenitally absent pericardium: wide split second sound, systolic ejection murmurs in the pulmonic area, rSR' patterns on ECG and prominent pulmonary artery on chest X-ray. The echocardiogram cannot be used to differentiate the two conditions and may be considered, erroneously, confirmatory of an atrial septal defect. If the correct diagnosis is not suggested by the characteristic roentgenographic features described, cardiac catheterization to exclude an in-

**Figure 7** Postpericardiectomy echocardiogram in a patient with constrictive pericarditis. A large right ventricular dimension and paradoxical (Type A) septal motion are apparent.

**Figure 8** Diagrams illustrating a proposed explanation for the echocardiographic abnormalities of absent pericardium. In the normal patient (A) the ultrasound beam traverses the right ventricular cavity near its apex, yielding a small right ventricular dimension. During systole a twisting motion of the entire heart occurs, with a posterior movement of the cardiac apex (curved arrows); this carries the ventricular septum away from the transducer. In patients with absent left pericardium (B) the leftward and posterior displacement in the supine position causes the beam to traverse the mid portion of the right ventricular cavity, thus yielding a larger measured dimension. From its abnormal posterior position the mobile heart twists anteriorly during systole (curved arrow); this cancels or over-balances the internal movement of the septum toward the left ventricular posterior wall, resulting in a net flat or anterior septal motion on echocardiogram during ventricular ejection.
tracardiac shunt may be necessary. Diagnostic pneumothorax, although definitive, may be hazardous, and many authors now consider it unnecessary in most cases.4,9

Echocardiographic abnormalities of interventricular septal motion have been noted following cardiac surgery.10 The cause of these abnormalities is unclear; possibilities include septal ischemia and right ventricular adhesions. At most institutions performing open-heart surgery the pericardium is not closed at the termination of open-heart surgery, but rather is left open vertically in the mid-line from the aortic reflection to the diaphragmatic surface so as to minimize the chance of postoperative tamponade. The consequence of such an open pericardium may be a reduction of intrapericardial tension and thereby increased cardiac mobility; if so, abnormal septal motion may result by mechanisms analogous to those already discussed. As postoperative adhesions form, however, cardiac mobility would again be reduced, and this may account for the observed reduction in incidence of postoperative septal motion abnormality in patients studied later than two months postoperatively.10 Leaving the pericardiectomy incision open cannot be the sole explanation for these echo abnormalities, however, since Burggraf and Craigie13 did not encounter septal dyssynergy after open or closed mitral commissurotomy whereas it was frequent after aortic and mitral valve replacement. Pericardiectomy is performed in all these procedures.

Addendum

Since the submission of this manuscript we have encountered a sixth patient with pneumothorax-proven congenital absence of the left pericardium. The echocardiographic findings in this patient are similar to the other five patients with this condition: large right ventricular dimension and abnormal (flat) interventricular septal motion.

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References

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M N Payvandi and R E Kerber

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