Two-Dimensional Echocardiographic Features of Echinococcosis of the Heart and Great Blood Vessels
Clinical and Surgical Implications

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Echocardiographic findings in cardiac echinococcosis have been previously described in isolated cases, but no detailed account of the various echocardiographic aspects and their clinical and surgical implications have ever been reported. We present a retrospective analysis of the two-dimensional echocardiograms of 15 patients with hydatid cysts affecting the heart or great blood vessels. Two-dimensional echocardiogram features were compared with the main clinical syndromes. Surgical confirmation of the echocardiogram findings was available in 12 patients. The cyst sizes ranged from 0.5 to 12 cm in diameter. Eleven patients had single cysts, three patients had two cysts, and one patient had multiple cysts. Cysts were located in the intramyocardial region in nine patients, the pericardial in three, and the paracardial in another three. All intramyocardial cysts protruded into the adjacent cardiac chamber, but in only two patients was there significant tricuspid valvular dysfunction. Pericardial and mediastinal cysts showed compression of cardiac chambers or great blood vessels, and two cysts had ruptured into the descendent thoracic aorta or inferior vena cava. In most patients, two-dimensional echocardiographic images of hydatid cysts were those of a cystic mass having well-defined edges and internal trabeculations corresponding to daughter membranes. However, in four patients, two-dimensional echocardiographic images showed a "solid" mass instead of a cystic mass, and in one patient with the multivesicular variety of echinococcosis, the images showed a large mass with poorly defined edges having a honeycombed appearance causing lysis of the anterior arch of the second and third left ribs. The two-dimensional echocardiographic finding showed good correlation with main clinical syndromes, but anaphylactic reactions occurred in every cyst location. In one patient, postoperative two-dimensional echocardiography showed two small intramyocardial cysts that had not been noticed during preoperative two-dimensional echocardiography or during surgical examination. Pathological examination in the four patients with a solid mass showed replacement of the hydatid liquid by necrotic matter containing membrane residues with a foreign-body inflammatory reaction of a granulomatous type. In conclusion, two-dimensional echocardiography is a very useful tool for diagnosis and management of patients with cardiac echinococcosis, but the great diversity of findings regarding number, size, location, and appearance of cysts must be borne in mind to interpret correctly the two-dimensional echocardiograms. (Circulation 1988;78:327–337)

Echinococcosis is a human parasitical condition caused by the larval stage of *Echinococcus granulosus*. Cardiac echinococ-
detailed accounts of various echocardiographic aspects and clinical implications from a relatively large series of patients have never been published. The present study correlates the main echocardiographic features with clinical data and surgical findings relating to 15 patients affected with echinococosis of the heart or great blood vessels.

Patients and Methods

From 1981 to 1987, a diagnosis through two-dimensional echocardiography of hydatid cysts affecting the heart or great vessels has been carried out in 16 patients in two Spanish hospitals. The two-dimensional echocardiographic study was performed in one patient with a 30° mechanical sector scanner (Ekosektor I, SK1), while in the remaining 15 patients, either a Hewlett-Packard or a General Electric phased-array ultrasound system was used. Standard tomographic views as previously described from para-sternal, apical, and subcostal positions were taken in all patients. However, in most of them, it was necessary to take atypical tomographic views to better establish the echocardiographic cyst images from various positions. All the echocardiograms were originally read by one of the authors, and the diagnosis of cardiac echinococcosis was made before surgery in every patient in whom surgical examination was carried out. The echocardiographic data relating to two of the patients have been previously published in isolated form.\textsuperscript{10,18}

A retrospective analysis of the two-dimensional echocardiograms regarding number, location, and appearance of the cysts and their relation with adjacent structures was carried out. The cysts were measured on the maximal diameter averaged from two orthogonal views, corresponding to two or more tomographic echocardiographic views. Patients were included in this study when the cardiac echinococcosis had been confirmed after surgery (12 patients) or when there were other confirmed locations involving extracardiac hydatid cysts together with a typical echocardiogram revealing a cystic, echolucent mass, having internal trabeculations produced by daughter vesicles (three patients). This study was carried out in 15 patients since there was another patient that was excluded because he did not show any extracardiac location of hydatid cyst and because cardiac surgery was not carried out. We also compared the echocardiographic features with the clinical and surgical charts. During this period, no other patient was found to have hydatid cysts of the heart or great vessels during cardiac surgery in either of the hospitals.

Results

Clinical Data

The main clinical data relating to the 15 patients are shown in Table 1. There were 10 men and five women. Ages ranged from 20–63 years. In 13 patients, hydatid cysts were also present in other locations; the lungs (10 patients) were the most frequent extravascular location. Hepatic hydatid cysts were present in three patients; and in two patients, cysts were located in either the brain or the kidneys. Three patients had progressive dyspnea on exertion and severe pulmonary hypertension caused by a spread of countless lung cysts.
TABLE 2. Main Echocardiographic Findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Cyst (n)</th>
<th>Maximal cyst diameter (cm)</th>
<th>Location</th>
<th>Appearance</th>
<th>Other findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1</td>
<td>3.5</td>
<td>IVS (apex)</td>
<td>Cystic mass with inner trabeculations</td>
<td>LV, RV protrusion</td>
</tr>
<tr>
<td>2</td>
<td>2</td>
<td>3</td>
<td>IVS (right side)</td>
<td>Solid mass</td>
<td>RV protrusion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0.5</td>
<td>LA (post wall)</td>
<td>Cystic mass</td>
<td>LA protrusion</td>
</tr>
<tr>
<td>3</td>
<td>1</td>
<td>4.5</td>
<td>Pericardium</td>
<td>Cystic mass with inner trabeculations</td>
<td>LA compression</td>
</tr>
<tr>
<td>4</td>
<td>1</td>
<td>3</td>
<td>IVS (right side)</td>
<td>Solid mass</td>
<td>RV protrusion</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>1</td>
<td>LV (papillary muscle)</td>
<td>Cystic mass</td>
<td>LV protrusion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0.8</td>
<td>IVS (left side)</td>
<td>Solid mass</td>
<td>LV protrusion</td>
</tr>
<tr>
<td>6</td>
<td>Multiple</td>
<td>1–3</td>
<td>Pericardium</td>
<td>Cystic mass</td>
<td>SVC compression</td>
</tr>
<tr>
<td>7</td>
<td>1</td>
<td>10</td>
<td>RV (free wall)</td>
<td>Cystic mass with inner trabeculations</td>
<td>RV inlet obstruction</td>
</tr>
<tr>
<td>8</td>
<td>1</td>
<td>12</td>
<td>Liver</td>
<td>Cystic mass with inner trabeculations</td>
<td>Open within IVC</td>
</tr>
<tr>
<td>9</td>
<td>1</td>
<td>9</td>
<td>Mediastinum</td>
<td>Multivesicular</td>
<td>RA protrusion</td>
</tr>
<tr>
<td>10</td>
<td>1</td>
<td>3</td>
<td>Pericardium</td>
<td>Cystic mass with inner trabeculations</td>
<td>None</td>
</tr>
<tr>
<td>11</td>
<td>1</td>
<td>6</td>
<td>Mediastinum</td>
<td>Cystic mass with inner trabeculations</td>
<td>Open to DTA</td>
</tr>
<tr>
<td>12</td>
<td>1</td>
<td>5</td>
<td>LV (post wall)</td>
<td>Cystic mass</td>
<td>LV protrusion</td>
</tr>
<tr>
<td>13</td>
<td>1</td>
<td>5</td>
<td>LV (post wall)</td>
<td>Cystic mass with inner trabeculations</td>
<td>LV protrusion</td>
</tr>
<tr>
<td>14</td>
<td>2</td>
<td>1–2</td>
<td>RV (free wall)</td>
<td>Cystic mass</td>
<td>RV protrusion</td>
</tr>
<tr>
<td>15</td>
<td>1</td>
<td>4</td>
<td>IVS (middle)</td>
<td>Solid mass</td>
<td>LV, RV protrusion</td>
</tr>
</tbody>
</table>

IVS, interventricular septum; LV, left ventricle; RV, right ventricle; LA, left atrium; TV, tricuspid valve; SVC, superior vena cava; IVC, inferior vena cava; and DTA, descending thoracic aorta.

visible through chest x-ray films. The main symptom shown by four patients was precordial pain; in one patient, the pain was of a pericardial type, and in the other three patients, it was interpreted as atypical angina. Sudden anaphylactic shock was the first symptom in three patients, one of whom also presented bilateral femoro-popliteal embolism. One patient had severe cardiomegaly and distension of the neck veins simulating severe tricuspid stenosis. Two patients presented a clinical picture of evolving stupor and intracranial hypertension caused by multiple brain hydatid cysts, which were surgically removed. None had any cardiovascular symptoms, but two-dimensional echocardiography was carried out because of the finding of nonspecific T-wave alterations on the electrocardiogram. One patient showed a superior vena cava obstructive syndrome and mediastinal widening on chest x-ray film, while another patient had a large anterior thoracic wall tumor without cardiac symptoms.

Echocardiographic Findings

Table 2 presents the main echocardiographic findings relating to the 15 patients.

Size and number of cysts. In 11 patients, the two-dimensional echocardiogram proved the existence of a single hydatid mass. Three patients had two cysts each, and in one patient, the echocardiogram showed multiple cystic masses (Figure 1). The cysts ranged in size from 0.5 to 12 cm in diameter (Figure 2).

Cyst locations. The cysts were located in the intramyocardial region in nine patients, the pericardial in three, and the paracardial in the other three. When the cysts were intramyocardial, the most common location was the interventricular septum (five patients) (Figure 3), followed by the posterior wall of the left ventricle and the right ventricular free wall (two patients each) (Figures 2 and 4). One patient with a cystic mass in the interventricular septum also had a 1-cm diameter cyst on the anterolateral left ventricular papillary muscle. Three patients had pericardial cysts showing multiple hydatid masses (one patient) or a single mass (two patients) (Figure 1). Two patients had cysts located on the posterior or anterior mediastinum, respectively. One patient had a large hepatic cyst that protruded into the inferior vena cava (Figure 5).

Echocardiographic appearance. The echocardiographic appearance of the cysts in 10 patients was that of one or several spheroidal masses of liquid content with a well-contrasted capsule. Seven of 10 patients showed multiple intracystic trabeculations probably produced by the daughter membranes (Figures 2 and 5). On the other hand, the echocardiogram in four patients revealed a mass having a solid content (Figures 3 and 6), two of which also had a cystic mass with liquid content in a different
location (Figure 3). The remaining patient’s echocardiogram showed a large anterior mediastinal mass having the echocardiographic appearance of honeycomb, which was formed by countless vesicles (Figure 7).

Relations with adjacent structures. All intramyocardial cysts protruded inside the adjacent cardiac chamber: right ventricle in four patients, left ventricle in four, both ventricles in two, and left atrium in one. One patient showed an echocardiographic obstruction of the inlet tract of the right ventricle caused by a very large right ventricular cyst (Figure 2). In another patient, a “solid” hydatid mass located on the right side of the interventricular septum showed a free intracavitary portion that adhered to the tricuspid valve, causing retraction of the septal leaflet (Figure 6). Three patients had extramyocardial compression of one of the cardiac chambers: left atrium in one patient, right atrium and the superior vena cava in one patient (Figure 1), and the anterior wall of the right ventricle in one patient (Figure 7). Finally, in two patients, the cysts were directly related to the great blood vessels. In one patient, a large hepatic cyst that had opened into the inferior vena cava had prolapsed inside the right atrium, while the other patient showed a posterior mediastinal cyst that had opened into the descending thoracic aorta (Figures 5 and 8).

Clinical and Echocardiographic Correlations

Severe pulmonary hypertension. Severe pulmonary hypertension caused by multiple pericardial hydatid cysts was seen in three patients, and all showed hydatid cysts that opened inside the right cardiac chambers. One patient had a large hepatic cyst opening into the inferior vena cava (Figure 5).

In the other two patients, the echocardiogram showed a cyst on the right side of the interventricular septum that protruded inside the cavity of the right ventricle. It was interesting that in both patients the echocardiographic image was of a solid mass, suggesting a thrombotic degeneration of the cyst after rupturing (Figures 3 and 6).

Systemic hydatid embolism. Systemic hydatid embolism occurred in four patients. Three suffered brain or kidney embolism, and in each patient, the echocardiogram showed an intramyocardial cyst that protruded inside the cavity of the left ventricle. The fourth patient showed a bilateral femoropopliteal embolism corresponding to the cyst that ruptured into the descending thoracic aorta (Figure 8).

Tricuspid valve dysfunction. Tricuspid valve dysfunction was present in two patients. In one, the echocardiogram showed a large cyst occupying the whole inlet and the apical portion of the right ventricle and produced right ventricular inflow obstruction, simulating a tricuspid stenosis (Figure 2). In the second patient, a cyst located on the interventricular septum adhered to the septal leaflet of the tricuspid valve, causing severe tricuspid regurgitation (Figure 6).

Chest pain. Chest pain was present in four patients. Pain was pericardial in origin in one patient who showed a pericardial cyst on the echocardiogram. In the other three patients, precordial pain was anginous, and one patient also had symptomatic sustained ventricular tachycardia; in all four patients, the echocardiogram showed intramyocardial cysts.

Sudden anaphylactic shock. Sudden anaphylactic shock was the first symptom in three patients. One patient had a left ventricular intramyocardial cyst,
another patient had a pericardial cyst, and one patient had a posterior mediastinal cyst that had ruptured into the descending thoracic aorta (Figure 8).

Atypical clinical pictures. Atypical clinical pictures were present in two patients. One patient had a radiological mediastinic mass that was producing superior vena cava obstructive syndrome, and the echocardiogram showed multiple pericardial cysts. One patient showed an anterior thoracic wall tumor that had grown rapidly. The echocardiogram showed a large, infiltrative, multivesicular, mediastinal mass located anterior to the right ventricle, which perforated the thoracic wall through the complete erosion of the anterior arch of the second and third left ribs (Figure 7).

Surgical Findings

The main surgical findings involving the 12 patients in whom surgery was performed are presented in Table 1. There were single hydatid cysts in nine patients and multiple cysts in three. Seven patients had nine intramyocardial cysts; four were located on the interventricular septum, three were located on the left ventricular free wall, one was located on the right ventricular free wall, and the ninth one was located on the left atrial posterior wall. Intramyocardial cysts always protruded into the adjacent cardiac chamber; right ventricle in three patients, left ventricle in four patients, both ventricles in one patient, and left atrium in one patient.

In the other five patients, the cysts had extra-myocardial locations. Surgical examination proved the presence of multiple cysts in one patient, and a single cyst that had an intrapericardial location in another patient. In one patient, a cyst was in the posterior mediastinum and opened into the descending thoracic aorta, causing multiple peripheral

**Figure 2.** Preoperative apical view on two-dimensional echocardiograms showing a large hydatid cyst within the right ventricle (Panels A and B), and postoperative apical two-dimensional echocardiograms of the same patient showing two small cysts on the right ventricular wall (Panels C and D) (patient 7). HC, hydatid cyst; LV, left ventricle; LA, left atrium; RA, right atrium; RV, right ventricle.
embolism of the hydatid membranes, which were removed by the Fogarty technique. One patient had a large anterior mediastinic multivesicular cyst that compressed the anterior wall of the right ventricle and perforated the costal plane through lysis of the anterior arch of the second and third left ribs and protruded in hourglass fashion on the external thoracic wall. Finally, in another patient, there was a large hepatic hydatid cyst that opened into the inferior vena cava and protruded into the right atrium.

The surgical examination confirmed the echocardiographic findings with regard to size, number, and location of the cysts in all patients. However, in one patient, the repeated echocardiogram taken after surgical removal of a large hydatid cyst from the right ventricle showed two small intramyocardial cysts close to the apex of the right ventricle that had not been noticed during the preoperative echocardiographic study or during surgical examination (Figure 2). In another patient, the surgical examination only showed a hydatid cyst on the interventricular septum. A second operation was scheduled when an intraoperative echocardiogram was carried out whereby a small cyst was located at the antero-
FIGURE 5. Subcostal two-dimensional (Panels A–C) and M-mode (Panel D) echocardiograms showing a very large hepatic hydatid cyst ruptured into the inferior vena cava and prolapsing inside the right atrium (patient 8). L, liver; HC, hydatid cyst; RA, right atrium; IVC, inferior vena cava.

lateral papillary muscle of the left ventricle that could then be surgically removed.

The pathological examination of the cysts in seven patients demonstrated the characteristic findings of the unilocular hydatid cysts. However, in four patients who had a solid-mass echocardiographic image, the pathological examination showed that the cyst contents, which usually consisted of hydatid liquid and the daughter membranes, had been replaced by necrotic matter containing membrane residues together with a foreign-body inflammatory reaction of granulomatous type with giant cells. Finally, in the patient whose echocardiogram showed a multivesicular mass with a honeycombed pattern, the pathological examination revealed a large multilocular cyst consisting of countless vesicles with gelatinous content.

Discussion

The findings shown in this retrospective study represent the first comprehensive description of the echocardiographic patterns in a relatively large series of patients with echinococcosis affecting the heart or great blood vessels.

In most of the patients with cardiac echinococcosis, the two-dimensional echocardiogram with images of spheroidal cystic masses showed well-defined edges and a liquid content that was often contained within internal trabeculae corresponding to the daughter membranes. These are usually single, intramyocardially located cysts affecting mostly the interventricular septum and the free wall of both ventricles. The average size of these cysts is 3–5 cm in diameter, and they usually protrude into one or
both ventricular chambers without producing any symptoms of mechanical interference. When these typical findings relate to the proven existence of hydatid cysts located in other organs or tissues, it is possible to establish with certainty an echocardiographic diagnosis of cardiac echinococcosis.

Nevertheless, this study shows that in many patients the echocardiographic findings may widely differ from this typical pattern. Cysts may not be single, but could be multiple, necessitating a detailed echocardiographic examination of all the cardiac structures in the search of other cysts. Cysts may be as small as 0.5 cm in diameter or even smaller, making them rather difficult to locate and differentiate from adjacent structures. In other patients, these cysts are so large that they distort the normal cardiac structures to such a degree that they are difficult to recognize from the usual transducer positions. Although these cysts are typically located on the interventricular septum or the ventricular free wall, they can also be found on the atrial wall or on the pericardium, or they may affect the heart or the great blood vessels from a paracardial location.

The diversity of hydatid cyst appearance on the two-dimensional echocardiograms has even greater implications. In contrast with the typical cystic image, some patients show solid masses on the echocardiogram, which makes these cysts more difficult to differentiate from other primary or secondary heart tumors. Pathological examinations of the four cysts that had a solid-mass echocardiographic appearance showed that they had undergone a degenerative process that replaced the normal contents of the cyst with necrotic material containing membrane residues surrounded by foreign-body reaction inflammatory tissue containing a large number of giant cells. This degenerative process is probably secondary to the opening of the cysts and the emptying of their contents, since a metastatic hematogenic dissemination had affected the lungs in two patients and had affected the brain, kidneys, and probably other organs of the systemic circulation in the other patient. However, the internal degenerative process of the intramyocardial cysts may also take place without any rupturing of the pericystic membrane and dissemination of its contents. This is probably the case with the last patient in the series. The echocardiogram showed a solid mass having a rounded and well-defined outline on the interventricular septum. Surgical draining of the mass contents was performed, but the pericystic capsule was not removed. Pathological examination showed a necrotic material with a large amount of cholesterol, and postoperative echocardiogram showed a cystic type of interventricular septum mass. The most probable mechanism of this degenerative process, without rupture, is the effect of the mechanical compression after the contraction of the cardiac muscle that surrounds the cyst.
Infestation of *Echinococcus granulosus* in humans is characterized by the formation of unilocular cysts consisting of a large vesicle that may contain one or several daughter vesicles. On rare occasions, infestation is characterized by the formation of multilocular cysts consisting of numerous small-sized vesicles. These multilocular cysts have far greater invasive capacity and can destroy adjacent structures. There was a patient in this series who had an anterior mediastinal hydatid cyst of the multilocular variety. Its echocardiographic appearance was very different from the rest of the cysts, its aspect being that of a mass with poorly defined edges, whose internal structure consisted of countless small cells that gave it a honeycomb appearance. The invasive and destructive nature of this type of cyst was demonstrated by the lysis of the anterior arch of the second and third left ribs and the transit to the external thoracic wall. To our knowledge, two-dimensional echocardiographic description of multivesicular hydatid cysts has not been previously reported.

The various forms of clinical presentation involving cardiac echinococcosis was adequately described by Perez-Gomez et al. These authors indicated that cardiovascular manifestations of cardiac echinococcosis could be caused by arrhythmia, angina, valvular dysfunction, pericardial reaction, pulmonary or systemic embolism, pulmonary hypertension, or anaphylactic reactions. This study also shows that there is a close correlation between the various forms of clinical presentation and the echocardiographic findings. Patients suffering from ventricular arrhythmias or angina may have intramyocardial cysts affecting their interventricular septum or the free or both ventricles. Valvular dysfunction may be due to an obstruction at the valvular orifice caused by a large intracavitary growth cyst or by adherence and retraction of the valve leaflets to an adjacent parietal cyst that had undergone a degenerative process accompanied by a pericystic inflammatory reaction. Diagnosis of these two types of valvular dysfunction can be easily established through two-dimensional echocardiography as pre-
Although this is a relatively large series, considering that this disease is extremely infrequent, it is not large enough to make statistical conclusions because of the diversity of locations and other characteristics of the cysts. However, echocardiographic diagnosis of cardiac echinococcosis was confirmed in each patient in whom surgical examination was performed, and in the time interval during which two-dimensional echocardiography was available at both hospitals, cardiac echinococcosis was not found in any other patient having hydatid cysts of the heart or great blood vessels at surgery.

Two-dimensional echocardiographic diagnosis of cardiac hydatid cyst was made in another patient, but he was not included in the present series because pathological examination was not available; therefore, confirmation of echinococcosis was not possible. This patient had rheumatic mitral valve disease with an asymptomatic small-sized, solid mass on the left ventricular posterior wall close to the atrioventricular sulcus. A probably dead hydatid cyst was diagnosed, and surgical replacement of the mitral valve was carried out. The surgeon examined the intramyocardial mass of the left ventricle with his hand but decided not to remove it. Two-dimensional echocardiographic follow-up did not show any change at the left ventricular mass 2 years after surgery.

In conclusion, this study shows that two-dimensional echocardiography is an extremely useful noninvasive tool for diagnosis and handling of patients with cardiac echinococcosis. Although hydatid cysts may also be diagnosed after other noninvasive techniques such as computed axial tomography and nuclear magnetic resonance, two-dimensional echocardiography continues to be the choice for locating hydatid cysts of the heart. This technique provides much information on the relations that exist between hydatid cysts and adjacent cardiac or vascular structure in close association with the various forms of clinical presentation of the disease and the surgical approach. Nonetheless, to correctly interpret the echocardiograms, one must bear in mind the great diversity of the findings in regard to number, size, location, and, above all, appearance of the cysts.

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References


KEY WORDS • echinococcosis • hydatid cysts
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