Echinococcus Disease of the Heart

By Jorge Dighiero, M.D., Eduardo Joaquin Canabal, M.D., Cesar V. Aguirre, M.D., Jacobo Hazan, M.D., and José O. Horjales, M.D.

In Uruguay, and in other sheep-grazing countries such as Argentina, Australia, New Zealand, and in the Mediterranean area, echinococcus disease is frequent. Publications relative to cardiac localization of the disease have been few, however.

A careful search of the literature reveals approximately 300 reported cases of echinococcus disease of the heart. We believe, however, that this condition is more frequent than supposed, because cysts located in the territory of distribution of the systemic circulation (spleen, kidneys, brain, muscle), are not uncommon.

Etiology, Pathogenesis, and Incidence

Echinococcus disease in man is caused by the development of the larval stage of the Taenia echinococcus, a cestode tapeworm whose definitive host is the dog in most instances. The dog is infested when fed with cyst-bearing organs (lungs, liver, etc.) of intermediate hosts, usually sheep. These cysts, "water vesicles," contain tapeworm heads (scolex) by the thousands, which develop in the intestine of the dog into the adult form of the cestode. The taenia, of rather small size (4 to 5 mm.), has a head, a neck, and 2 body segments or rings. A double line of hooklets and 4 suckers in the head of each worm help it to get anchored to the intestinal mucosa of the host. The last proglottid containing the sexual organs, when distended by the ova, is shed in the feces of the dog, thus contaminating grass and water that may be ingested by the intermediate host, i.e., sheep. Owing to close contact with infested dogs in rural areas, man may become readily contaminated.

The chitinous shell of the ova of Taenia echinococcus is dissolved in the gastrointestinal tract of the intermediate host (man, sheep), thus liberating a hexacanth (6 hooklets) embryo, which then passes through the gastric or intestinal wall to reach the portal circulation and the liver, the organ most frequently involved by echinococcus disease. Occasionally, the liver may be spared because of the rather small size of the hexacanth embryo and the distensibility of hepatic capillaries. If so, the embryo goes on its way down the pulmonary capillary circulation, where it may be blocked. Pulmonary cysts are second to hepatic ones in incidence. This second capillary barrier may also be passed by the parasite, however.

Most classic authors believed that echinococcus disease of the heart was predominantly or exclusively secondary to rupture of a primary hydatid cyst elsewhere in the body and grafting of the hexacanth embryo on the endocardial surface of the right heart chambers, which were more commonly involved than the left. From its original endocardial graft the embryo would be able to reach any other region of the heart by active movements.

Based on a study of over 100 cases and on laboratory experiments, Dévé2-10 stated that the hydatid cyst of the heart is always primary and single,11,12 even in those cases with other than cardiac localizations.13 According to him and to most modern authors14-18 the hexacanth embryo, after passing through the capillary networks of the liver and lungs, arrives in the left heart chambers, reaches the coronary circulation and becomes lodged in the interstitial tissue of the myocardium of any 1 of the 4 heart chambers or cardiæ septa. Dévé8 also pointed out that the primary hydatid cyst is more frequently located in the

From the Laboratorio Cardiológico, Hospital de Clínicas, Faculty of Medicine, Montevideo, Uruguay.
wall of the left rather than the right heart chambers, particularly the ventricle, because of the richer coronary circulation. Our own experience, based on the observation of 16 cases confirms this conclusion (in 11 cases the cysts were located in the left ventricle).

Although Dévé's views on the pathogenesis of echinococcus disease of the heart have had world-wide acceptance, some authors recently proposed the theory that the hexacanth embryo can reach the heart via the lymphatic system (chyliferous thoracic duct subclavian vein right heart chambers pulmonary capillary network left heart chambers coronary network), based on the observation of many cases of cardiac echinoccosis without concomitant hepatic cysts, on the bigger size of the hexacanth embryo as compared with that of the hepatic and pulmonary capillaries and on the finding of ganglionic echinococosis in some animals (sheep). They also accept the possibility of transcendocardial migration of the embryo once it has arrived in the right heart chambers.

Several authors state that cardiac cysts are seen in about 0.5 to 2 per cent of all cases of human hydatidosis.

Analysis of 30 cases of hydatid cardiac cysts in our country shows that the incidence is higher in men than in women (2:1 ratio). Other statistics are in accordance with this proportion.

Most cases occur in the second to fifth decades in patients living in cattle-raising areas.

Pathology

In a few weeks after its arrival in the interstitial tissue of the myocardium of any part of the heart, the hexacanth embryo becomes vesicular, grows slowly but steadily into a unilocular hydatid vesicle with an outer elastic membrane doubled by an inner germinative layer containing a colorless "rock water" fluid called hydatid fluid.

The hydatid vesicle thins out the myocardial wall during its process of growth and exerts pressure on the surrounding muscle fibers, which become more or less ischemic depending on the degree of pressure caused by the para-site and the resistance of the cardiac tissues. Due to mechanical, toxic, allergic, and inflammatory phenomena caused by the hydatid vesicle, the tissue reaction in the host leads to the formation of a fibrous capsule called adventitia (adventitious cyst, according to Dévé) showing cellular infiltration which becomes gradually thicker with time.

The internal germinative layer of the primitive hydatid vesicle, by a process of proliferation, gives rise to multiple vesiculated structures called "brood capsules" (proligerous capsules), which may float in the fluid. From these capsules scolex may develop. If hydatid fluid of a primary unilocular cyst is centrifuged, the so-called "hydatid sand" containing numerous brood capsules and free scolex can be obtained. These 2 elements are of utmost importance in the spread of echinococcus disease in the same individual.

"Due to the particular density of the myocardium, the development of the primary hydatid cyst is restrained. This phenomenon is more obvious when the cyst is located in either ventricle. This is the reason why in most cases there is a reactive daughter cyst formation (multivesicular cyst) and why the primary cyst undergoes other changes such as degeneration or suppuration." Daughter cysts are smaller cysts contained within the primitive or "mother" cyst. They also show an external laminated capsule, an internal germinative layer, hydatid fluid, brood capsules, and scolex.

"When one or more of these changes take place, the adventitia becomes thicker, denser, and even partially calcifie d and causes comparatively more harm to the surrounding myocardial structures." Other histologic changes like perihydatid granulomata, hydatid pseudotuberculosis, etc. have been described in this condition. In some cases of profound alterations of the primitive hydatid cyst one may find an amorphous substance called "hydatid putty," but none of its normal contents.

According to postmortem and surgical reports, the size of the primitive uncomplicated hydatid cyst of the heart varies from that of a pea to a grapefruit. During the period of
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intramural development of the cyst, its presence may be completely overlooked, but later it may produce a localized bulge in the cardiac silhouette that permits roentgenologic recognition of the disease.

The primary hydatid cyst of the heart has a marked tendency to rupture either into the lumen of a cardiac chamber or into the pericardial sac, depending on its primary location and on the direction of least resistance. The primary cyst may also rupture into the myocardium itself (local secondary echinococcosis). While the pericardium usually reacts in front of a hydatid cyst by developing adhesions, the endocardium does not react and is therefore easily invaded by the cyst.

The accident of rupture (single or multiple) of a hydatid cyst of the heart may give rise to different complications.

**Uncomplicated Echinococcus Disease of the Heart**

According to most authors, "uncomplicated echinococcosis of the heart remains often silent and latent." Our own experience and that of others demonstrates that uncomplicated hydatid disease of the heart has no particular clinical picture. However, in some instances, thoracic pains, palpititation, paroxysmal tachycardia, various murmurs, congestive heart failure, angina pectoris, etc., may occur. But most cases of this disease have been suspected initially by roentgenologic examination. Roentgenology (fluoroscopy, roentgenograms, tomography, kymography) represents a diagnostic means of highest importance in the detection of the disease, because the deformities of the cardiac silhouette and the calcifications may be easily discovered and studied in all projections. Information regarding the size, shape, and location of the abnormal mass, its contour, its movements, and the characteristics of the calcification may be obtained. Every shadow of calcific density projected in the periphery of the heart should raise the possibility of a hydatid cyst, if signs of constrictive pericarditis are not present. "When spotty areas of calcification are seen in the cardiac shadow, one may also think of echinococcus disease, if calcification of the heart valves can be ruled out. Intramural fibromas may also calcify, but this disease is extremely rare."!

Angiocardiography gives valuable information on the deformities caused by the intramural development of a hydatid cyst in the internal outline of the cardiac chambers and on the thickness of the myocardial wall underlying the cyst.

Electrocardiography, according to our own experience, is important. Compression ischemia of myocardial fibers and the thinning out of the myocardial wall produced by the cyst cause various electrocardiographic abnormalities. The standard, the unipolar limb, and multiple precordial, back, and esophageal leads must be used in every case of ventricular location of the disease in a search for a pattern of circumscribed ischemia and changes in the QRS complexes. Atrial cysts may or may not cause changes in the P waves. Abnormalities in the P-R interval and QRS complexes (notching, slurring, widening) have been described in cases of septal cysts.

In every case of possible echinococcal disease of the heart various laboratory examinations are indicated. Eosinophilia (7 per cent or more) may be of diagnostic significance, but it is often absent in old, altered cysts. Other conditions also are accompanied by eosinophilia. The intradermal (Casoni) test is only of diagnostic value when positive, particularly, late positive. Several well-established cases of hydatid cardiac disease have repeatedly shown negative intradermal tests. Complement-fixation tests (Weinberg, Ghedini, Imaz-Appathie, and Lorentz) have proved to be not entirely specific.

**Complications**

The natural history of the primitive myocardial hydatid cyst is interrupted by its rupture in most instances, although occasionally the primitive cyst may remain alive and quiescent for a long time or death of the para-
site may occur, with subsequent spontaneous cure of the disease.

Rupture of the primitive myocardial cyst into the endocardial cavity, the pericardial sac, or the myocardium itself may produce a series of complications, depending on the way it happens (small fissure in the hydatid membrane and the adventitious cyst, wide open rupture, recurrent ruptures) and on its contents (unilocular cyst, multivesicular cyst, cyst with different alterations). If the opening in a subendocardial fertile hydatid cyst is small (fissure), only hydatid fluid containing free scolex and brood capsules escapes. This accident usually provokes a rapid onset of hyperergic phenomena of varying intensity, i.e., anaphylactic shock, urticaria, fever, and diarrhea. Late complications may occur owing to the evolution of hydatid material (scolex) distant from the primitive cyst (metastatic echinococcosis of the lesser or systemic circulation). If this accident supervenes in a subepicardial cyst, an acute or subacute serofibrinous or purulent pericarditis with a high percentage of eosinophils may ensue. Secondary echinococcosis of the pericardium represents a late complication.9,22

When the primitive myocardial cyst freely ruptures into the cardiac cavity, two different kinds of complications may appear. Immediate ones are represented by the hyperergic phenomena mentioned above, which may be fatal, may allow the patient's survival, or may even be overlooked in some cases. Late complications, besides metastatic echinococcosis of the lesser or systemic circulation17,18,20 due to "the liberation, the dissemination and the distant colonization of living parasitic elements (scolex) proceeding from the primitive lesion,"70 are caused by the embolization of fragments or of the entire membrane of the primitive hydatid cyst, of daughter cysts (living or dead) and of hydatid debris to the lesser or systemic circulation.23,26,28,40,42 These serious complications are not always late. Some cases of sudden death immediately after rupture of the hydatid cyst into the cardiac cavities are due to intracardiac blockage or pulmonary or cerebral embolism by daughter cysts or fragments of hydatid membrane.8,28,43 Similar complications are seen in cases of hydatid cysts of the liver rupturing into the inferior vena cava.25,44,45

Depending on the contents of the primitive cyst rupturing into the pericardial sac, different forms of chronic pericarditis are encountered. Hydatidopericardium is a chronic generalized or localized (due to adhesions) pericarditis with hydatid material in evolution (living daughter cysts) or involution (fragments of membrane pertaining to the "mother" cyst or dead daughter cysts or both). A purulent pericardial effusion, in addition to hydatid material, is usually present,48,49 although cases with no effusion have been described († reabsorption).38 Hydatidopericardium may remain quiescent for a long time, may give symptoms and signs of cardiac compression,50 or be even invasive (pseudoaneurysmatic form).47,51,52 These events are due to the fact that living daughter cysts containing fertile elements (scolex) may continue to grow outside the mother cyst. This phenomenon also takes place in cases of embolization of daughter cysts to the lesser23,40 or systemic6,7 circulation causing, besides subtotal or total arterial obstruction, hydatid arterial aneurysms and a secondary intrarterial echinococcosis. As any living daughter cyst may rupture wherever it be located, further complications of the disease can be expected.23,29,51

Practically all forms of hydatid pericarditis cause thickening of the pericardium. Constrictive pericarditis has been described in cases of protracted evolution of an overlooked hydatidopericardium.50 Secondary echinococcosis of the pericardium is frequently associated with hydatidopericardium.48 Secondary cysts (pericardial grafts), as well as daughter cysts proceeding from the primitive myocardial hydatid cyst, may eventually rupture into the cardiac chambers.41,53

It is to be emphasized that diagnosis of hydatid disease of the heart must be made in the early, uncomplicated stages of the disease prior to the occurrence of any one of its common and dreaded complications. Other-
wise, little or nothing can be done for the benefit of the patients in most cases.

**Summary**

The etiology, pathogenesis, pathology, and the clinical picture of uncomplicated and complicated echinococcosis disease of the heart are discussed.

Cardiac location of the larval stage of *Taenia echinococcus* in human beings is not an uncommon disease in sheep-grazing countries. The authors put special emphasis on the methods helping to establish the diagnosis of the condition prior to the appearance of any one of its dreaded complications, which make treatment much more difficult and prognosis considerably poorer.

**Summario in Interlingua**

Es discutite le etiologia, pathogenese, patologia, e tableau clinic de noncomplicate e complicate morbo cardiae e echinococces.

Le location cardiae de *Taenia echinococcus* in le stadio larval non es un phenomeno incommun in humanos in paises a cultura de oves. Le autores sublinea specialmente le methodos que es de adjuta in establir le diagramo del condition ante le manifesation de ule de su timibile complicationes que rende le tractamento muito plus difficile e le prognose considerabilemente minus favorable.

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