Intracardiac Rupture of Hydatid Cysts of the Heart

A Study Based on Three Personal Observations and 101 Cases in the World Literature

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INTRACARDIAC RUPTURE of a hydatid cyst of the heart is of the greatest importance due to the serious immediate or delayed consequences. It is our purpose to make an exhaustive study of this type of cyst rupture and its consequences.

Material

The material used as the basis of this work consists of 269 cases of hydatid cyst of the heart reported in the world literature and demonstrated at autopsy or operation, 11 being personal observations, which make up nearly the whole of the cases published in the world.

We have diagnosed intracardiac rupture of the cyst when at autopsy or operation we have found the communicating orifice into the heart cavity, when metastatic visceral echinococcosis is present, or when hydatid embolism has occurred.

Frequency

Intracardiac rupture occurred in 104 of the 269 observations of cardiac echinococcosis (38.66 per cent).

The Ruptured Cyst

In 70 cases it was a primary cyst of the heart, in seven cases a secondary cyst of the pericardium, and in two cases a local secondary cyst of the heart. In 14 cases it was not possible to decide whether the ruptured cyst was a primary or a secondary cyst of the pericardium and in seven cases the cyst that gave rise to the intracardiac rupture could not be identified.

Situation of Intracardiac Rupture

The locations of the rupture were as follows: Left ventricle, 35 cases; right ventricle, 30 cases; left atrium, 18 cases; right atrium, 17 cases; aorta, one case; pulmonary artery, three cases. Three cases of rupture of two cysts, one in the left heart cavities and the other in the right side, have been reported.

Secondary cysts of the pericardium open into the atriæ, aorta, or pulmonary artery. Until now it has not been proved that these cysts can rupture into the ventricles.

Intracardiac rupture occurred in 30 cases (88 per cent) of the 34 cysts located in the right ventricle and in only 35 cases (37 per cent) of the 94 cysts situated in the left ventricle. This may be due to the comparatively thin wall of the right ventricle coupled with a higher pressure within the left ventricle.

Multiple Rupture of Cysts

Hydatid cysts of the heart frequently undergo multiple rupture.

Pericardio-Intracardial Rupture of a Primary Cyst

In six cases we are certain that the primary cyst first ruptured into the pericardium and then into a chamber of the heart, in view of the autopsy findings of secondary cysts in the pericardial sac. In one case there was simultaneous rupture into the pericardium and a heart chamber, with hemopericardium and death.

Multiple Intracardiac Rupture of Cysts

We are certain that this type of rupture has occurred when secondary visceral echinococcosis exists in the corresponding vascular area, thus pointing to a previous rupture, together with a recent rupture of a cyst; or when multiple hydatid embolism can be found.
INTRACARDIAC RUPTURE OF HYDATID CYSTS

Figure 1
Hydatid cyst of the lateral wall of the right ventricle. The zone where the cyst ruptured into the ventricular cavity can be clearly identified, being obstructed by hydatid membranes. (Reproduced from Amargós et al.12 with the permission of the journal El Día Médico Argentino.)

We have found that it occurred in 24 cases,8, 12, 14, 27, 29, 33, 40, 42, 45, 48, 50, 52-54, 57, 63, 67, 80-82, 85, 94, 95, 104 in four of which the cyst had first opened into the pericardium.27, 29, 54

Consequences of the Intracardiac Rupture of Cysts

The immediate or delayed consequences of the intracardiac rupture of a cyst depends, above all, upon the contents of the cyst.1 A healthy, intact cyst is unilocular and contains liquid and microscopic elements only; whereas a ruptured cyst, with its vitality threatened, degenerates or dies or gives rise to daughter cysts.1 This explains why the first rupture of a cyst does not generally cause mechanical disorders, but, by the subsequent growth of the germs sown, may give rise to metastatic visceral echinococcosis. On the other hand, if the cyst has previously opened into the pericardium or into the heart cavities, subsequent intracardiac rupture of the cyst may cause serious embolism.

There are 17 cases in which we found secondary visceral echinococcosis, absence of hydatid emboli, and absence of anatomic signs of rupture of the cyst into the pericardium.4, 16, 18, 19-23, 46, 47, 51, 61, 62, 65, 66, 76, 86 In these 17 cases we believe the initial rupture of the cyst did not give rise to emboli. There are four cases35, 55, 83, 89 in which the cyst had ruptured previously into the pericardium, and in which the subsequent intracardiac rupture gave rise to voluminous hydatid emboli. There are four other cases40, 50, 52, 57 with metastatic visceral echinococcosis, which certified intracardiac rupture a long time previously, and in which a second intra-
cardiac rupture, at a later date, gave rise to large mortal emboli. Nevertheless the following exceptions were found: 1. One case in which, apparently without a previous rupture into the pericardium, the cyst ruptured into the heart chambers, giving rise to embolism and with no secondary visceral echinococcosis. In that case, however, the rupture with embolism described at autopsy may not have been the first intracardiac rupture of the parasite; it is not certain that a previous rupture without embolism did not occur, but with scolices that did not develop in the viscera in which they were sown, so that the accident was not detected. 2. In three cases signs of a previous rupture into the pericardium existed, and yet when the cyst opened into the heart the only indication was the presence of secondary visceral echinococcosis without any signs of hydatid embolism.

Secondary cysts of the pericardium, because of the precarious conditions under which they develop, are frequently degenerated or multivesicular, for which reason, although in a different way from the primitive cyst of the heart, they generally give rise to large emboli from the moment of their first rupture.

Rupture of the Cystic Adventitia with Intracardiac Migration of the Whole Mother Cyst

This type of cystic rupture has been observed in seven cases.\textsuperscript{25, 26, 38, 68, 70, 73, 96}

**Intracardiac Heterotopic Echinococcosis\textsuperscript{117, p. 42}**

In five cases the mother cyst, which migrated from its original site through the ruptured adventitia into a chamber of the heart, continued to develop there for some time.\textsuperscript{25, 26, 38, 73, 96}

**Intracardiac Rupture Causing Obstruction of Cardiac Orifice**

Hydatid vesicles set free within the heart chambers through rupture of a cyst may obstruct the cardiac orifices, giving rise to serious circulatory accidents.\textsuperscript{25, 26, 38, 72, 92, 96}

**Intracardiac Rupture of Cysts Causing Hydatid Embolism of the Arteries**

A macroscopic hydatid embolus in an artery may consist of a mother cyst,\textsuperscript{68, 70} of curled fragments of membranes of a mother cyst or, more often, of daughter cysts of varying size, either alive or collapsed.\textsuperscript{1}

Embolic obstruction of a blood vessel may lead to a visceral infarct, to softening of the brain, or to peripheral gangrene. This is not always so, however, for the particles of the laminated layer forming the emboli are sometimes so small as to give rise to almost microscopic specific lesions.\textsuperscript{91} Besides, parasitic embolism does not necessarily lead to complete obliteration of the affected artery, and this may be due (1) to the absence of subsequent arterial thrombosis, in spite of prolonged permanence of a parasitic embolus\textsuperscript{64, 68}—this is unusual, however;\textsuperscript{1} and (2) when the fragments of the laminated layer are not voluminous, they may become marginated against a bifurcation or at the orifice of a collateral
vessel—these soft and essentially ductile foreign bodies spread out along the corresponding wall of the artery and only produce a partial or, so to say, unilateral, parietal thrombosis.118,119

Hydatid Aneurysm.120 Arterial hydatid embolism may give rise to an aneurysmal dilatation of the embolized vessel arising at the point where the hydatid membranes or vesicles are held up.44, 52, 74, 82, 84, 91, 121

Periarterial Hydatid Embolism.122 Fragments of hydatid membranes carried along within the lumen of an artery may be found, some time later, encysted outside the artery or, rather, in the adventitia.

Pulmonary Hydatid Embolism (figs. 3 and 4). There are 27 cases with positive autopsy findings.12, 33, 42, 44, 45, 49, 52, 53, 63, 64, 68, 70, 78, 80-85, 88, 90, 97, 103, 105, 107, 109, 114 in six of which the embolus occluded the trunk of the artery. The emboli generally affect several arterial branches and both lungs, but there are cases presenting embolic obstruction of only one branch64, 68, 70, 103 and with multiple embolism in only one lung.53, 80, 83, 85 Pulmonary embolism was the cause of death in eight cases.52, 68, 70, 78, 81, 83, 88, 103 In 11 instances cardiac echinococcosis was revealed only by the symptoms of pulmonary embolism.14, 39, 43, 55, 68, 69, 70, 78, 83, 88, 103

Chronic Hydatid Cor Pulmonale.12, 123 The obstruction of the branches of the pulmonary artery by hydatid emboli may cause pulmonary hypertension with consequent hypertrophy and insufficiency of the right ventricle.44

Arterial Hydatid Embolism of the Greater Circulation. Cerebral hydatid embolism (eight cases)8, 27, 56, 57, 71, 74, 91, 102 may affect the internal carotid artery,56, 71, 102 the middle cerebral artery,27, 57, 102 the anterior cerebral artery,91 or peripheral arterial branches.27, 91 It is generally bilateral, but there are exceptions.56, 71

Hydatid embolism of the abdominal aorta and its branches (eight cases)8, 15, 28, 34, 36, 36, 74, 99 may affect only the aorta15 or the branches into which it divides. Gangrene of the lower limb may result.8, 36, 74, 99

Renal hydatid embolism (six cases)34, 74, 91,

97, 102, 114 may be unilateral97, 102, 114 or bilateral.34, 91

In Kähler’s case121 hydatid embolism of the upper limbs almost certainly occurred; in six other cases there were clinical manifestations suggesting such embolism but without positive autopsy findings.8, 56, 60, 74, 100, 102

Less frequently the hydatid embolism occurred in other arteries: the splenic,91, 102 the coronary,91, 102 the mesenteric, the celiac trunk,102 etc.

In seven cases the hydatid embolism was multivisceral.8, 34, 56, 74, 91, 94, 102

Metastatic Visceral Echinococcosis124

When rupture of a cardiac cyst spills fertile microscopic elements into the blood stream, they are swept along until held up in the smallest vascular ramifications in the viscera. There they become definitely implanted and may give rise to new cysts, which are called metastatic because they are situated in organs far distant from the mother cyst.

Pulmonary Metastatic Echinococcosis.125-128 It may result when a cardiac cyst ruptures into the pulmonary circulation (27 cases).12, 14, 33, 40, 42, 45-52, 54, 63, 65-67, 76, 77, 80, 81, 85, 90, 95, 107, 109

Systemic Metastatic Echinococcosis. It may take place when rupture of a cardiac cyst occurs into the systemic circulation.126, 129-133

Metastatic echinococcosis of the brain was found in 21 cases,4, 8, 16-19, 22, 25, 28, 36, 33, 36, 37, 57, 60-62, 96, 98, 99, 110 of the spleen in 13 cases.4,
of the kidney in 12 cases; 4, 29-22, 38, 55, 78, 80, 90, 97, 102
of the cerebral in one case, 37 of the thyroid gland in one case, 102 and of the subcutaneous, lax connective tissue in one case. 8 This metastatic echinococcosis is generally multivisceral, but there are exceptions. 17, 23, 27, 38, 61, 68, 110

The discovery of multiple cysts in the brain, spleen, or kidney indicates their metastatic origin, granted that one can exclude massive infestation, always accompanied by multiple cysts in the liver and in the lungs.

Often, only the metastatic visceral echinococcosis gives rise to symptoms, whereas the cardiac cyst remains latent. 11, 16, 18, 19, 22, 23, 28, 30, 37, 61, 86, 98, 110

Intra-arterial Metastatic Echinococcosis.
Hydatid vesicles carried into large arterial branches sometimes adhere and continue to grow and develop within the lumen of the artery. 12, 42, 44, 45, 49, 52, 53, 56, 58, 64, 83, 90, 97, 99

Local Secondary Echinococcosis of the Heart

The intraarterial rupture of a primary hydatid cyst of the myocardium may give rise to a local or neighboring sowing of fertile elements capable of producing further cysts of the heart wall. 16, 41, 51, 66, 73, 86, 111, 112

Hydatid Allergy

Intraarterial rupture may give rise to allergic reactions that are sometimes fatal. 8, 48, 51, 102, 108

Sudden Death

This occurred in 29 per cent of cases, 15, 24-29, 29, 31, 38-40, 42, 43, 47, 52, 55, 59, 64, 69, 70, 72, 78, 81-85, 88, 89, 92, 93, 96, 103, 104, 108, 111 and in 16 instances it was apparently the only evidence of the existing cardiac echinococcosis. It was much more frequent in intraarterial rupture on the right side of the heart than on the left.

Latency

The hydatid cyst of the heart that has ruptured into the cardiac chambers may exceptionally remain latent. 21, 57 It becomes evident either through clinical manifestations connected with the rupture itself or from the symptoms due to the late consequences of this accident. Nevertheless the actual accident of intraarterial rupture may be latent or go unnoticed. 1, 16, 18, 19, 21-23, 25, 26, 28, 30, 33, 37, 40, 41, 46, 47, 50-52, 61, 82, 86, 87, 98, 110, 118

Summary

Intraarterial rupture of hydatid cysts of the heart is very frequent (38.66 per cent) and often causes sudden death (29 per cent). Cysts of the right ventricle rupture into the heart cavities far more often than do those of the left ventricle. Frequently, the cyst undergoes multiple intraarterial or pericardio-intraarterial ruptures.

The consequences of the intraarterial rupture depend above all upon the contents of the cyst. If it contains liquid and microscopic elements, no mechanical disorders occur, but it may give rise to metastatic visceral echinococcosis. If it contains vesicles, or is degenerated, it may give rise to large emboli.

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Knowledge and Ignorance, Pride and Power

Absolute knowledge could, therefore, leave nothing outside itself; and only on condition of knowing everything could man be granted its attainment. Man behaves as if he were destined to reach this absolute knowledge; and the incessant why which he puts to nature proves it. Indeed, this hope, constantly disappointed, constantly reborn, sustains and always will sustain successive generations in the passionate search for truth.

Our feelings lead us at first to believe that absolute truth must lie within our realm; but study takes from us, little by little, these chimerical conceits. Science has just the privilege of teaching us what we do not know, by replacing feeling with reason and experience and clearly showing us the present boundaries of our knowledge. But by a marvellous compensation, science, in humbling our pride, proportionately increases our power. Men of science who carry experimental analysis to the point of relatively determining a phenomenon doubtless see clearly their own ignorance of the phenomenon in its primary cause; but they have become its master; the instrument at work is unknown, but they can use it. This is true of all experimental sciences in which we can reach only relative or partial truths and know phenomena only in their necessary conditions. But this knowledge is enough to broaden our power over nature. Though we do not know the essence of phenomena, we can produce or prevent their appearance, because we can regulate their physico-chemical conditions. We do not know the essence of fire, of electricity, of light, and still we regulate their phenomena to our own advantage. We know absolutely nothing of the essence even of life; but we shall nevertheless regulate vital phenomena as soon as we know enough of their necessary conditions. Only in living bodies these conditions are much more complex and more difficult to grasp than in inorganic bodies; that is the whole difference.—CLAUDE BERNARD, M.D. An Introduction to the Study of Experimental Medicine. New York, The Macmillan Company, 1927, p. 82.
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Circulation. 1963;27:366-374
doi: 10.1161/01.CIR.27.3.366
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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