

## Mini Review

# Cardiac Hydatid Disease

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**Abstract**

Human Echinococcosis is a parasitic disease caused by ingestion of parasites egg, tapeworms of the genus *Echinococcus* present in food, water or contaminated soil or by direct contact with host animals. Is present on every continent, with a higher prevalence in rural regions of Africa, Central and South America, Asia, Middle East and southern Europe. The World Health Organization (WHO) estimates that there are about 1 million people affected by echinococcosis in the world. In Latin America hydatid disease affects mainly agricultural and pastoral regions and is often associated with low-income populations and high rurality, with an average fatality rate of 2.9%, responsible for more than 300,000 hospital days.

Although cardiac involvement in hydatid disease is rare, it represents only 0.5-3% of all hydatid cysts in humans, cardiac cysts are extremely dangerous because they often break and can produce pericardial tamponade, anaphylaxis, embolisms and dissemination of scolex, so this clinic presentation represents an interesting surgical challenge.

**INTRODUCTION**

Human echinococcosis is a parasitic disease caused by ingestion of parasites egg, tapeworms of the genus *Echinococcus* present in food, water or contaminated soil or by direct contact with host animals.

Human echinococcosis occurs in four clinically well-established forms, the Cystic echinococcosis (CE) and Alveolar Echinococcosis (EA) is the most frequent and disturbing.

- 1) Cystic Echinococcosis: is produced by infection with *Echinococcus granulosus*.
- 2) Alveolar Echinococcosis: caused by infection with *E. multilocularis*.
- 3) Polycystic Echinococcosis is caused by infection with *E. Vogeli*.
- 4) Unicystic Echinococcosis is caused by infection with *E. oligarthrus*.

**Epidemiology**

Cystic Echinococcosis or hydatid disease is caused by the infection of the larvae of *E. granulosus*, which is present on every continent, with a higher prevalence in rural regions of Africa [1], Central and South America, Asia, Middle East and southern Europe (Figure 1).

Worldwide incidence is estimated at approximately 0.4 per 100,000 people [2]. The World Health Organization (WHO) estimates that there are about 1 million people affected by echinococcosis in the world [3]. In Latin America hydatid disease

affects mainly agricultural and pastoral regions and is often associated with low-income populations and high rurality.

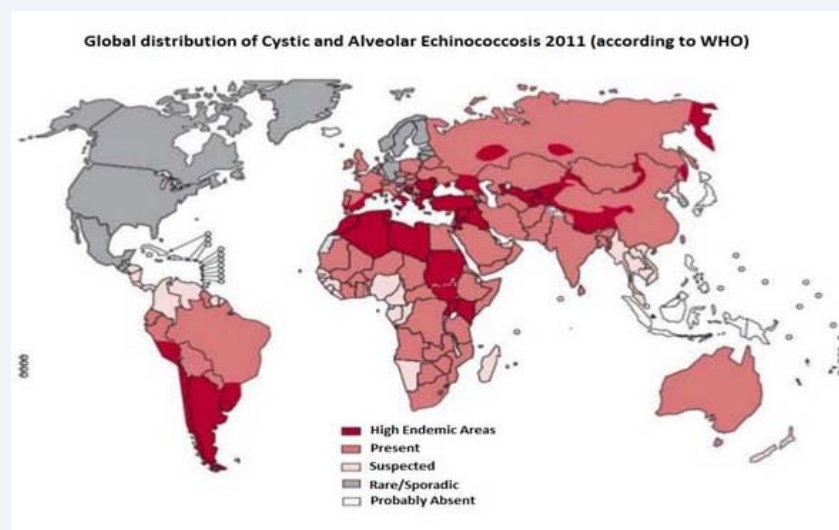
The higher prevalence countries are Argentina [4], Uruguay, Brazil, Peru and Chile [5]. In the period January 2009-December 2014, the five countries that make up the initiative to control cystic echinococcosis (EQ) in South America reported 29,556 human cases; Peru is the country with the highest number of human cases in this period with 20,785 cases, with an average fatality rate of 2.9%, responsible for more than 300,000 hospital days [6].

This zoonosis takes place in a cycle that mainly involves the infected cattle and dogs, with occasional infection of humans. The parasite is transmitted to dogs when they eat animal organs containing hydatid cysts. Once ingested by dogs, these cysts develop into adult tapeworms that remain fixed in the intestine (Figure 2). The mature worm is found exclusively in the intestine of canids where not usually generate pathology.

Subsequently infected dogs eliminate tapeworm eggs in their feces every 30-45 days or so, contaminating soils, crops, water and general environment where dogs pass. Sheep, goats, cattle and pigs ingest the eggs of *Echinococcus granulosus* from contaminated soil. Once eaten, the eggs hatch and develop into cysts in the internal organs of the animal.

In the vast majority of cases, transmission of *E. granulosus* to humans is through ingestion of water or food contaminated with fecal matter from an infected dog food. *Echinococcus* eggs deposited in the soil can remain viable for a year.

Despite the various efforts made in epidemiological and prophylactic aspects, WHO and the Pan American Health



**Figure 1** Global distribution of Cystic and Alveolar Echinococcosis (WHO).

Organization (PAHO) estimate that there are approximately 100 million people in the Region of the Americas, suffering at least one infectious disease unattended, generating a major health and economic development impact, since these diseases are chronic and have lasting effects on health. Affect growth, physical and intellectual development, learning capacity and reduce labor productivity and the opportunity to generate adequate income [7].

In Chile, the annual incidence is about 800 cases. These have a risk of death of 6 to 7%. The disease attacks throughout the ages, being more frequent between 10 and 60 years [8].

### Physiopathology

The life cycle of the tapeworm *Echinococcus*, has intermediate and definitive hosts. Intermediate hosts are cattle, sheep, goats and pigs. The definitive hosts in the vast majority of cases are related to livestock or grazing, but may also be some wild animals. Dogs usually become infected by eating the remains of infected cattle and sheep. Subsequently it develops the adult tapeworm in the dog's intestine and eggs are removed to the environment through feces.

Man is an intermediate host, infected in most cases through accidental exposure to canine feces, but can also become infected by eating contaminated animal viscera. Tapeworm eggs, which hatch in the small intestine and the larvae hatched from these eggs can enter the small intestine come to any part of the human body, subsequently develop into hydatid cysts, which contain many protoscolices, each with ability to become an adult tapeworm in a definitive host (Figure 3).

Organs that tend to be more affected are liver in 50-79% of the cases, lungs at 20% and the rest of the body 10% [9]. Many patients (55-85%) also have multivisceral damage, compromising liver, lung, and other organs [10].

Most human infections are asymptomatic for years, becoming symptomatic by the slow growth of the cyst and the mass effect

that produces less common symptoms are secondary bacterial infection of the cyst or anaphylactic reactions cyst rupture.

### Cardiac Hidatidosis

Although cardiac involvement in hydatid disease is rare, in represents only 0.5-3% of all hydatid cysts in humans [12], cardiac cysts are extremely dangerous because they often break and can produce pericardial tamponade, anaphylaxis, embolisms and dissemination of scolex.

*E. granulosus* can reach the heart through the coronary circulation, a patent foramen ovale or pulmonary circulation. Any part of the heart may be affected, the most common location is left free wall ventricle (50-77%), of which 50% have pericardial involvement [13]. The right ventricle is affected in 10% of cases, pericardium in 7%, 6% pulmonary artery, and the interventricular septum in 4% of the cases [14].

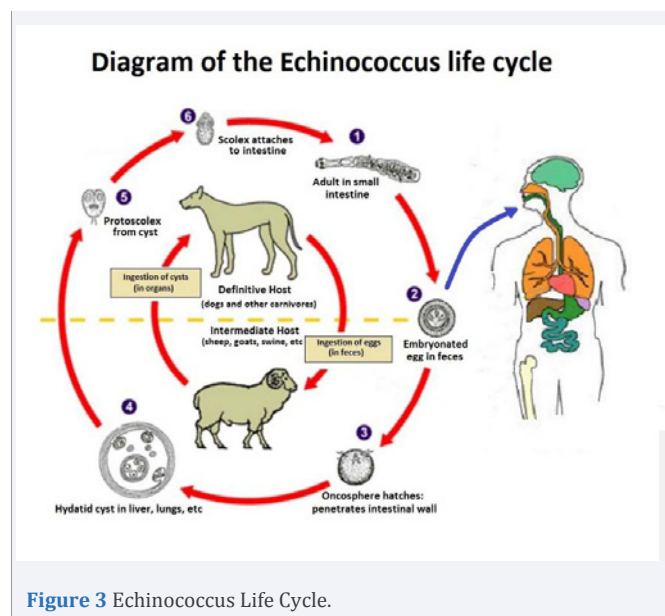
The cyst growth process in the heart is similar to other organic locations, with the difference that the continuous traumatic action of myocardial contractions induces a multivesicular development and increases tension on the cysts facilitating their breakage [15].

The clinical manifestations are extremely variable; from patients remain asymptomatic (90%) [16], to those who manifest complications, often fatal abruptly by its severity. The clinic is directly related to the location and size of the cyst, chest pain being the most common symptom, but we can find ventricular tachyarrhythmia's, atrioventricular or fascicular blocks [17], peripheral embolism and obstruction of the outflow tract of the left ventricle. Involvement of the atrium or right ventricle can produce varying degrees of stenosis or tricuspid valve insufficiency, pulmonary embolism, obstruction of the outflow tract of the right ventricle. If hydatid cysts are intracardiac, they can produce destruction of valvular tissue being need the replacement of the affected valve. When the cyst is complicated and breaks can cause pericardial tamponade, anaphylaxis, and embolisms with dissemination of scolex [18].

Some studies show that left ventricular hydatid cysts are



**Figure 2** EG in the mucosa of Dog intestine.



**Figure 3** Echinococcus Life Cycle.

often located in the subepicardium and unusually break (usually into the pericardium); instead cysts located in the right ventricle usually located in the subendocardial, the breakage can cause (to the ventricular cavity) a pulmonary embolism.

The diagnosis requires a high degree of suspicion, because the symptoms are usually little and too late. In the vast majority of cases it is done with imaging. Serology usually has good specificity, but does not have good sensitivity [19]. The chest radiograph may show some signs like; presence of pulmonary cysts, altered cardiac silhouette or calcifications, which are suggestive of hydatid disease. Echocardiography plays an important role because it is a diagnostic tool easy to use and not expensive [20], and can be supplemented by CT scan or MRI, which primarily help in determining the non-cardiac commitment of cysts.

The differential diagnosis with other cystic formations arises as teratoma, congenital cysts pericardium secondary to intrapericardial hematomas or cysts cystic degeneration of tumors.

In heart echinococcosis surgical treatment is better than

the exclusive medical treatment, so treatment should be a combination of both.

With respect to time of onset of antiparasitic therapy, there is no consensus. Some groups administered benzimidazole before surgery [21], others use it in Intraoperative or post-operative [22], as a measure to reduce the risk of recurrence. In our group, we believe it is an appropriate measure to opt for the pre-operatorion treatment to reduce the risk of spread.

Surgery requires cardiopulmonary bypass and aortic clamping, with mild hypothermia. One aspect to take into account in the removal of heart cysts, is the possibility of rupture and subsequent massive embolism content circulation with consequent hypersensitivity reaction and the potential anaphylactic shock [23], so prevention measures of this potentially fatal complication must be very diligent. This complication can occur during surgery or in the immediate postoperative period. 75% of patients who died of cyst rupture were secondary to shock or embolic complications [24].

As to which is the best strategy to monitor response to therapy, there is no consensus. Some studies suggest patients control these tests repeated image at intervals of 3 months, at least 3 years; as recurrence may appear long after the medical and / or surgical treatment [24].

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