Chapter

# Cardiac Cysticercosis: Current Trends in Diagnostic and Therapeutic Approaches

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## Abstract

Cardiac cysticercosis is a rare infection and its diagnosis is usually incidental, as most patients are asymptomatic. Laboratory and imaging tests, such as echocardiogram and cardiac nuclear magnetic resonance, can also be used in the diagnostic approach. The clinical manifestations are broad and patients can present with symptoms that range from heart failure to arrhythmias. Treatment of this condition has been scarcely studied and no protocols have been well established to date. One can choose not to treat the asymptomatic cases or to use cestocides, in the case of symptomatic individuals. Patient monitoring through cardiac enzymes and electrocardiogram during treatment is recommended, as well as performing imaging tests after treatment. This chapter aims to discuss cardiac cysticercosis, divided into sessions that will cover everything from its epidemiology and clinical aspects to diagnostic methods, therapeutics and treatment monitoring, with emphasis on the most current aspects.

**Keywords:** Helminthiasis, Taenia Infections, Cysticercosis, Neglected Diseases, Cysticercus, Cardiovascular Infections, Cardiac Diseases

## 1. Introduction

Endemic in Asia, Africa and Latin America, cysticercosis is caused by the ingestion of *Taenia solium* eggs. Contamination is caused by autoinfection in individuals with taeniasis, due to poor hand hygiene, or by heteroinfection, from contaminated foods, especially raw vegetables and water [1, 2].

The incubation period after the ingestion of the egg lasts for 3–8 weeks, with symptom onset occurring in up to 3–5 years. It is believed that the cysticercus is able to survive for a period of 3 to 6 years, after which it begins to degenerate, causing fibrosis or necrosis in the affected tissues due to the triggered inflammatory process. Nonetheless, the infection usually remains asymptomatic. Morphologically, the cysticercus exhibits two forms: the cystic form that contains the scolex and the racemous form, which corresponds to a set of vesicles, without scolex and a configuration similar to grape clusters [1].

The most affected tissues in cysticercosis are muscle and eye tissues, and the most severe manifestations are, overall, those related to the central nervous system [1].

Cardiac cysticercosis is rare, although autopsy studies have shown a 20–27% prevalence of cardiac cysticercosis occurring concomitantly with neurocysticercosis [3, 4]. In cases of cardiac involvement, pericardial effusion, signs of edema and myocardial inflammation or even myocardial infarction can occur [5].

The diagnosis can be attained by conclusively demonstrating the presence of the cysticercus through histopathological techniques in biopsy material, by visualizing the scolex in computed tomography or magnetic resonance imaging tests or by fundus examination, in cases of intraocular cysticercosis. In the absence of direct demonstration of the parasite presence, serological tests allow the diagnosis of the disease, although these tests are not widely commercially available [6].

There is no consensus on the treatment of cysticercosis with cardiac involvement. Patients with extraneural cysticercosis should be evaluated and high-risk situations, such as disseminated infections, intraventricular cysts and ocular involvement, should be excluded. Cases of asymptomatic individuals may not require surgical or anthelmintic therapy [3, 7].

The role of anthelmintics, such as Albendazole and Praziquantel, in the treatment of cardiac cysticercosis has not been directly investigated. However, it seems that the use is valid due to their effectiveness in the treatment of cysticercosis in other sites, such as neurocysticercosis. The role of cardiac surgery in the treatment of this condition also remains unclear [2].

As it is a rare condition and, therefore, still little discussed, this chapter aims to discuss the existing evidence in the literature on the diagnosis and treatment of cysticercosis with cardiac involvement, emphasizing the most current trends.

## 2. Epidemiology and clinical presentation of cysticercosis with cardiac involvement

Cysticercosis with cardiac involvement, especially myocardial impairment, is considered rare, and has been scarcely studied, being more frequently asymptomatic, so its diagnosis is often incidental, usually attained during cardiac surgery or at autopsy [1, 2]. Retrospective studies with autopsies have shown a variable prevalence of cardiac involvement, between 22.6% and 26.8% [3, 4, 8, 9] of the cases identified with cysticercosis.

As for the presentation according to the age group, another autopsy study showed that, of patients with cysticercosis, 27.8% were elderly and 72.2% were non-elderly, and that among the first, 20% had cardiac involvement due to cysticercosis, whereas of the latter, 25% showed cardiac cysticercosis [10]. Research [8] has shown a higher prevalence of cysticercosis in male individuals in all age groups, except for those between 30 and 39 years old, in which there was a greater number of affected women.

Cysticerci appear as oval cystic structures with thin, semitransparent and serous walls, containing liquid and measuring up to 30 mm in diameter, which contain a characteristic scolex [6]. Cysticercal involvement and distribution are variable in cardiac tissues, including the pericardium, subendocardium and myocardium [1, 6]. Cardiac cysticerci are usually multiple and, rarely, a single cardiac cyst may be present [1].

The immune system of the infected individual may not recognize the cysts for many years. However, when cysticerci age, their cystic structures can rupture, which will result in an inflammatory response [9] with variable expression and the possibility of granulomas and also fibrosis [2]. Although most cases are asymptomatic, clinical manifestations may occur as a result of inflammation, precisely at

the time of spontaneous cysticercus degeneration or during treatment, which may result in different degrees of cardiac involvement [1, 2].

One study [11] showed that non-elderly individuals had significantly more cardiac inflammation than the elderly and that the inflammatory infiltrate decreases with age and depends on the evolutionary stage of the cysticercosis. Moreover, the study showed there are gender differences regarding the intensity of the inflammatory response triggered by the presence of cysticerci in the heart, with women (elderly and non-elderly) showing a more intense response to the parasitosis than men.

Therefore, in cases of cardiac involvement, myocarditis with transient left ventricular dysfunction, pericardial effusion of variable extension, restrictive cardiomyopathy due to fibrosis formation [6], ischemic heart disease [12], in addition to valve pathologies [1, 2, 6] and conduction system defects, such as bradyarrhythmia and advanced atrioventricular block [13, 14] can occur. Dilated cardiomyopathy [6] and even severe ventricular dysfunction and cardiogenic shock have also been reported, in cases with severe cardiac or cardiopulmonary infestation [15].

The reasons why some patients have multiple cysticerci, while others have a single lesion remain uncertain. In a prospective follow-up study in India with 60 patients with disseminated cysticercosis, it was observed that changes in the Toll-like receptor-4 of *Asp299Gly* and *Thr399Ile* genes increased the risk (6.63 and 4.61-fold in the presence of polymorphisms, respectively) of disseminated cysticercosis [7].

The relationship between cysticercosis and immunosuppression remains uncertain, although post-chemotherapy cases of cysticercosis have been documented in Brazil and Mexico [15, 16]. Animal experiments using chemotherapeutic drugs have suggested that innate resistance contributes to the outcome of primary infection and there is a high degree of resistance to reinfection, both in the humoral and cellular mechanisms. This resistance to reinfection is altered by immunosuppression, probably due to the delay in antibody synthesis onset [17].

**Table 1** shows a summary of several cases of patients with cysticercosis and cardiac involvement reported in the literature, with patients' general characteristics, clinical manifestations and sites of disease presentation (cardiac and extracardiac).

| Reference | Case report location | Patient | Presentation | Site of disease |
|---|---|---|---|---|
| Kalra et al. [18]. | Chicago, United States of America. | 35-year-old man. | Palpitations and mild shortness of breath. | Heart. |
| Kochanowski et al. [19]. | Warsaw, Poland. | 39-year-old man. | Asymptomatic. | Heart and liver. |
| Nery et al. [12]. | Belo Horizonte, Brazil. | 59-year-old man. | Angina. | Heart. |
| Thomas et al. [13]. | Mthatha, South Africa. | 42-year-old man. | Bradycardia. | Heart and brain. |
| Bastos et al. [20]. | Belo Horizonte, Brazil. | 39-year-old man. | Dyspnea, progressive low visual acuity at the left side and multiple subcutaneous nodules. | Heart, lung and brain. |
| Dsilva et al. [21]. | Mumbai, India. | 62-year-old man. | Episodes of generalized tonic–clonic seizures and multiple subcutaneous nodules over both calves, arms and nape of the neck. | Heart, brain, subcutaneous tissue, liver, and muscles. |

| Reference | Case report location | Patient | Presentation | Site of disease |
|---|---|---|---|---|
| Khandpur et al. [22]. | New Delhi, India. | 48-year-old man. | Innumerable soft to firm, deep-seated asymptomatic nodular swellings over the trunk and extremities. | Heart, skin, central nervous system, skeletal muscles, eye and lungs. |
| Vaidya et al. [23]. | New Delhi, India. | 27-year-old man. | Multiple subcutaneous nodules all over the patient's body. | Heart, brain, face, orbit, lungs, pancreas and spleen. |
| Spina et al. [6]. | Sydney, Australia. | 24-year-old woman. | Frontal headaches, high fever, sweating, arthralgia, nausea, vomiting and weight loss of about 6 kg in one month. | Heart, brain, pancreas, liver, pleura and skeletal muscles. |
| Jain et al. [24]. | Mumbai, India. | 19-year-old man. | Headache and vomiting, seizures, decreased vision and bilateral proptosis. | Heart, brain, extradural spinal space, muscles, lungs, pancreas, and eyes. |
| Eberly et al. [25]. | De Bilt, The Netherlands. | 17-year-old boy. | Asymptomatic. | Heart. |
| Sousa et al. [26]. | Fortaleza, Brazil. | 26-year-old man. | Headache and generalized seizures. | Heart, brain and subcutaneous tissue. |
| Melo et al. [27]. | Salvador, Brazil. | 46-year-old woman. | Dyspnea on exertion and palpitations. | Heart, brain and muscles. |
| Mauad et al. [15]. | São Paulo, Brazil. | 53-year-old woman. | Mental confusion, incoherent speech and hypoactive behavior. | Heart, lungs, pleura, subcutaneous tissue and brain. |
| Robinson et al. [28]. | Montreal, Canada. | 33-year-old man. | Skin nodules, polyarthritis, hemolytic anemia, and malnutrition. | Heart, skin, muscles, brain, larynx, pleura and liver. |
| Sun et al. [14]. | Beijing, People's Republic of China. | 33-year-old man. | Headache, nausea, vomiting, and bradycardia. | Heart, brain and subcutaneous tissue. |

**Table 1.**
*Clinical characteristics, sites of disease presentation (cardiac and extracardiac), and clinical manifestations in patients reported in the literature.*

## 3. Current diagnostic and therapeutic approaches to cardiac cysticercosis

### 3.1 Diagnostic methods used in the analysis of cardiac involvement due to cysticercosis

The diagnosis of cardiac cysticercosis can be attained by conclusively demonstrating the presence of the cysticercus through histopathological techniques in biopsy material or by visualizing the scolex, either by computed tomography or

nuclear magnetic resonance imaging tests. Some authors consider these tests to be the gold standard in the diagnosis of cysticercosis, as they allow the visualization of the parasite and the host's reaction process [29].

The computed tomography shows greater sensitivity in the detection of calcified cysticerci, whereas the magnetic resonance imaging has greater resolution power, which may show the scolex with better accuracy [6, 30]. The echocardiogram may play a role in identifying cardiac cysts and occasionally identifies cysts consistent with cysticercosis during routine screening for other purposes [1, 6].

In the absence of direct visualization of the parasite, serological tests allow the disease diagnosis [1, 6, 30, 31]. The oldest tests, which used unfractionated antigens, including the enzyme-linked immunosorbent assay (ELISA), have been associated with high rates of false-positive and false-negative reactions [21, 30]. Currently, the Enzyme-linked immunoelectrotransfer blot (EITB) is considered one of the most reliable immunological tests for the diagnosis of cysticercosis and neurocysticercosis [29, 31]. The initial evaluation of this test indicates a 98% sensitivity and 100% specificity in serum and cerebrospinal fluid (CSF) samples. Other studies have reported an 86–100% variation in sensitivity in serum and 81–100% in CSF, while the variation in specificity ranged from 93–100% in both sample types [29]. Unfortunately, the availability of these tests is experimental, not being commercially available and rarely available at most medical centers, except for research centers working in this area [31].

Some laboratory alterations can be identified in cardiac cysticercosis, such as marked peripheral eosinophilia disclosed in the blood count, albeit only in the case of a ruptured cyst [1, 32]. Most individuals affected by this pathology do not have viable *Taenia solium* in the intestine, making the stool parasitological test ineffective [1, 31, 32].

**Figure 1** summarizes the main diagnostic methods that can be used for the diagnostic definition of cysticercosis with cardiac involvement.

### 3.2 Treatment and monitoring of the patient with cysticercosis and cardiac involvement

There is no consensus regarding the treatment of cardiac cysticercosis. In patients with extra-neural cysticercosis, the existence of high-risk conditions, such as disseminated infection, intraventricular cysts and ocular involvement, should be evaluated [1, 31]. Asymptomatic cases might not require more specific

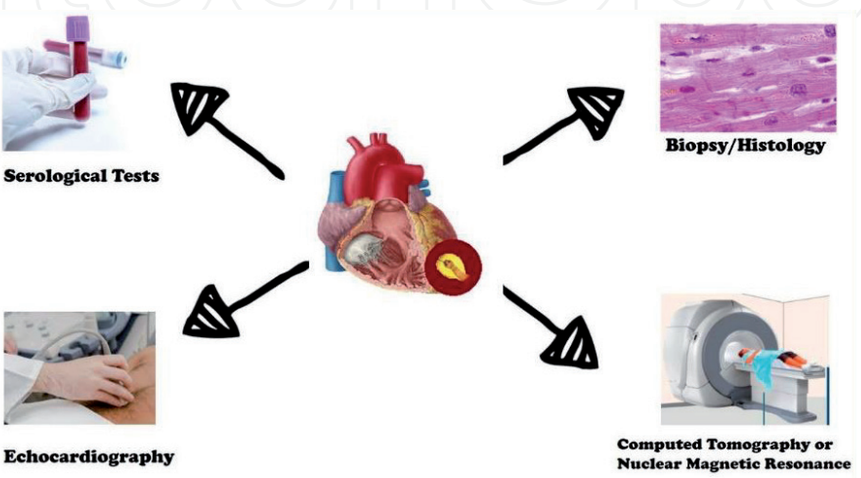


**Figure 1.**
*Diagnostic methods that can be used for the diagnostic definition of cysticercosis with cardiac involvement.*

| Reference | Cardiac manifestations | Diagnostic methods | Cardiac structures involved | Treatments performed | Course of disease |
|---|---|---|---|---|---|
| Kalra et al. [18]. | Palpitations and mild shortness of breath. | Magnetic resonance and serological tests by Western blot. | Anterior wall of the left ventricle. | Oral Albendazole 400 mg twice a day for 2 weeks. | One week after completing the treatment a repeat cardiac magnetic resonance showed that the lesion had decreased considerably in size. |
| Kochanowski et al. [19]. | None. | Magnetic resonance, computed tomography and specific antibodies in serum. | Apical segment of the anterior left ventricular wall. | No specific treatment was performed. | The patient has been followed for 16 years and has had no cardiac symptoms. |
| Nery et al. [12]. | Angina, new 1-mm ST-segment elevation of the inferior and lateral walls, and elevated cardiac troponin level. | Echocardiography, magnetic resonance and biopsy of the cardiac mass. | Intramural lesion at the left ventricular apex with extension through the free wall into the pericardial space. | Cardiac surgery. Specific drugs for the treatment of cysticercosis were not prescribed. | The postoperative period was uneventful, and the patient was discharged on day 6 postoperatively. The patient was symptom free at follow-up. |
| Thomas et al. [13]. | Bradycardia and complete heart block. | Computed tomography and echocardiography. | The exact location of the lesions was not described, although it was reported that there were multiple myocardial calcified and active cysts. | Treatment was started only with Prednisolone (there is no description on dose and treatment duration with this drug), and five days later with the anti-helminthic drug (Praziquantel 50 mg/kg/day for 14 days). | Repeat electrocardiogram after a week of treatment showed sinus rhythm with a heart rate of 70 beats per minute. |
| Bastos et al. [20]. | Dyspnea. | Computed tomography and an excisional biopsy of a subcutaneous nodule. | Myocardium (the exact location of the cysticercus was not described). | Albendazole (there is no description regarding the dose, route of administration and treatment duration). | Two months later the chest radiographies were normal and the patient was asymptomatic. |
| Dsilva et al. [21]. | None. | Nuclear magnetic resonance. | Ventricular myocardium and right pericardial fat pad. | Patient was started on Albendazole 15 mg/kg/day, divided in two doses and Prednisolone 1 mg/kg in tapering doses along with Phenytoin for 28 days. | There is no description of the evolution of cardiac lesions. A review brain nuclear magnetic resonance with whole body screening was performed after treatment completion, which showed a reduction in the number of active lesions. |

| Reference | Cardiac manifestations | Diagnostic methods | Cardiac structures involved | Treatments performed | Course of disease |
|---|---|---|---|---|---|
| Khandpur et al. [22]. | None. | Computed tomography, echocardiography and skin biopsy. | Anterior cardiac wall. | Specific drugs for the treatment of cysticercosis were not prescribed. Carbamazepine 200 mg twice daily was used for seizure prevention. | The patient is still being followed by the medical team and is well. |
| Vaidya et al. [23]. | None. | Computed tomography and anticysticercal antibodies by enzyme-linked immunosorbent assay (ELISA). | Interventricular septum musculature. | Patient was treated with oral Albendazole and antiepileptic medication. There is no description of the administered doses or treatment duration | The patient showed good evolution and was discharged with instructions to maintain regular follow-ups. |
| Spina et al. [6]. | Tachycardia, hypotension, respiratory distress, pleuritic chest discomfort and large pericardial effusion. | Magnetic resonance and serological test for the presence of cysticerci in serum and cerebrospinal fluid. | Multiple myocardial and epicardial cysts in the left ventricular anterior, septal, posterior and lateral walls. | Prednisone (1 mg/kg daily), followed by 15 days of Albendazole (400 mg twice daily). | A repeat cardiac magnetic resonance after 9 months showed resolution of the myocardial cysts, pericardial effusion, and myocardial inflammation. On follow-up one year later, the patient was well. |
| Jain et al. [24]. | None. | Magnetic resonance, echocardiography, muscle biopsy. | Cardiac muscle. | Specific drugs used for the treatment of Cysticercosis were not prescribed. The patient was treated symptomatically with antiepileptic drugs, steroids, and diuretics. | There is no description regarding patient evolution and follow-up. |
| Eberly et al. [25]. | None. | Computed tomography, nuclear magnetic resonance and biopsy. | Left ventricle along the endocardial surface of the anterior wall near the anteroseptal basal region. | Cardiac surgery and one-month course of Albendazole (daily dose and route of administration were not reported). | The patient's postoperative course was uneventful. A follow-up echocardiography one month later was normal. |
| Sousa et al. [26]. | Acute heart failure and shock. | Subcutaneous nodule biopsy, computed tomography and echocardiography. | The location of the cardiac cysticerci was not described. | Patient was treated with Phenytoin, Diazepam, Dexamethasone and Thiabendazole. There was no information on doses, administration routes or treatment duration. | One year after discharge, the patient was doing well taking only Phenytoin. |

| Reference | Cardiac manifestations | Diagnostic methods | Cardiac structures involved | Treatments performed | Course of disease |
|---|---|---|---|---|---|
| Melo et al. [27]. | Left ventricular (LV) overload, secondary alterations in ventricular repolarization, supraventricular ectopic activity and LV diastolic dysfunction. | Positive hemagglutination reaction up to 1: 4 for cerebrospinal cysticercosis and ELISA reaction for positive anti-*Cysticercus cellulosae* antibody detection. Microcalcifications were also shown on the transthoracic echocardiography, a pattern similar to that demonstrated in plain soft tissue radiography. | Diffuse myocardial microcalcifications in both ventricles. | No treatment was carried out with antiparasitic agents. | The patient showed evolution with significant clinical improvement after specific treatment for restrictive cardiomyopathy, being referred for outpatient treatment with a cardiologist. |
| Mauad et al. [15] . | Acute heart failure and shock. | Patient died before diagnosis was attained. | Subendocardium, subpericardium and in-tramyocardium. | No specific treatment was implemented. | Patient died during hospitalization. |
| Robinson et al. [28]. | Pericardial effusion, large left ventricular apical filling defect and severe tricuspid stenosis. | There is no description of the diagnostic methods. | Epicardium, myocardium (left ventricular apex), inferior vena cava, tricuspid valve and pulmonary artery. | Cardiac surgery and pharmacological treatment (drugs used in the treatment were not described). | Unstable postoperative course, requiring inotropic support and intra-aortic balloon pump. Patient died eleven weeks postoperatively. |
| Sun et al. [14]. | Complete intra-Hisian block. | Subcutaneous nodule biopsy and complement fixation test for *Cysticercus cellulosae*. | The cardiac location of the cysticercus was not described. | The patient was treated with Mannitol and Furosemide and then with Praziquantel. There is no description regarding the dose, route of administration and treatment duration with these drugs. | The subcutaneous nodules decreased in size and the symptoms due to increased intracranial pressure disappeared, but complete AV block persisted. The patient refused a pacemaker implantation. |

**Table 2.**
*Diagnostic methods and treatments used, evolution and course of the disease in cases of cysticercosis with cardiac involvement reported in the literature.*

therapy, such as surgical procedures or anthelmintic pharmacological therapy [3, 7]. In cases of asymptomatic myocardial involvement, there is usually no justification for any type of intervention, given the benign prognosis associated with this condition [3, 6].

The role of anthelmintic drugs such as Albendazole and Praziquantel in the treatment of cardiac cysticercosis has not been directly investigated in large studies; however, it seems that their use is valid due to their efficacy in the treatment of cysticercosis in other sites, such as neurocysticercosis [6]. Therefore, these drugs are used at the same dose and duration utilized to treat neurocysticercosis, with Praziquantel at a dose of 50 mg/kg/day for 15 days and Albendazole at a dose of 15 mg/kg/day for 8–15 days [6, 30, 31]. In case of a solitary cyst or granuloma, monotherapy with Albendazole may be sufficient [31].

The role of the surgical removal of cysts through cardiac surgery for the treatment of cardiac cysticercosis is also not yet clear and may be indicated when some valvular apparatus is compromised, when there is left ventricular outflow tract obstruction or even when there is epicardial coronary artery compression, with subsequent myocardial blood supply reduction [6, 12, 25, 28].

Corticosteroids are used together with antiparasitic agents in the initial treatment of neurocysticercosis to decrease the pericystic inflammatory reaction that follows larval necrosis, but there is no definition regarding its use in patients with cardiac involvement, although it is theoretically possible [1, 2, 7, 17]. A randomized trial comparing 6 mg/day of Dexamethasone for 10 days with 8 mg/day for 28 days, followed by a gradual reduction over 2 weeks, suggested that increasing the dose of Dexamethasone results in fewer seizures during treatment for neurocysticercosis [30, 31]. However, in some cardiac conditions, such as pericarditis, steroids have been associated with increased relapse and recurrence. Due to the rapid response to anti-helminthic therapy in some cases, Albendazole can be used without steroids, but with adequate monitoring [18].

Cardiac monitoring is recommended, with cardiac enzymes and electrocardiogram, during the early stages of the treatment for cardiac impairment due to cysticercosis [6]. The cases reported in the literature show that the lesions on the MRI disappear 6 to 9 months after treatment [1, 16]. However, there is no recommendation on the most appropriate periodicity for the assessment of myocardial necrosis biomarkers, or imaging tests after treatment [6].

**Table 2** shows a summary of several cases reported in the literature regarding compromised cardiac structures, the several methods used for the diagnosis, the performed treatments and their monitoring, in addition to the disease evolution in each case of cysticercosis with reported cardiac impairment.

## 4. Conclusion

The clinical management of cysticercosis with cardiac involvement is complex, due to the rarity of the pathology and the broad spectrum of clinical presentation, ranging from asymptomatic cases to those with more severe manifestations, such as cardiogenic shock and advanced cardiac blocks. The lack of studies directly investigating the role of diagnostic methods for its detection, as well as drug therapy effectiveness with anthelmintic drugs and corticosteroids, and the role of the surgical approach are also factors that have an impact on the management of these cases. Therefore, we propose that individuals with cardiac cysticercosis should be evaluated individually by a multidisciplinary team, so that the best diagnostic and therapeutic conduct, as well as the best way of monitoring each specific case, can be implemented.

## Acknowledgements

We would like to thank the Brazilian Research Council (Conselho Nacional de Desenvolvimento Científico e Tecnológico – CNPq) for supporting our research (research grant to GBSJ # 310974/2020-8).

## Conflicts of interest

The authors declare no conflicts of interest.

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