# Cardiac Hydatid Disease and Extracardiac Organ Involvement: A Tertiary Single-Center Experience

Kardiyak Hidatik Kist Hastalığı ve Ekstrakardiyak Organ Tutulumu: Üçüncül Tek Merkezli Deneyim

İsa Çam<sup>1</sup>, Kurtuluş Karaüzüm<sup>2</sup>, İrem Karaüzüm<sup>2</sup>, Törehan Özer<sup>1</sup>, Ahmet Yalnız<sup>3</sup>, Öözgür Çakır<sup>1</sup>,
Hakan Cebeci<sup>4</sup>, Başak Erdemli Gürsel<sup>5</sup>, Oğuz Omay<sup>6</sup>, Ercüment Çiftçi<sup>1</sup>, Tayfun Şahin<sup>2</sup>, Yonca Anık<sup>1</sup>

<sup>1</sup>Kocaeli University Faculty of Medicine, Department of Radiology, Kocaeli, Turkey

<sup>2</sup>Kocaeli University Faculty of Medicine, Department of Cardiology, Kocaeli, Turkey

<sup>3</sup>Gebze Fatih State Hospital, Clinic of Radiology, Kocaeli, Turkey

<sup>4</sup>Selçuk University Faculty of Medicine, Department of Radiology, Konya, Turkey

<sup>5</sup>Uludağ University Faculty of Medicine, Department of Radiology, Bursa, Turkey

<sup>6</sup>Kocaeli University Faculty of Medicine, Department of Cardiovascular Surgery, Kocaeli, Turkey

# ABSTRACT

**Introduction:** Cardiac involvement is rare in hydatid cyst disease, which accounted for 0.5%-2% of all hydatidosis cases. Cardiac cysts usually occur as part of a wider infestation with extracardiac involvement. This study aimed to describe the general characteristics, imaging findings, and range of organ involvement in cases of cardiac hydatid disease.

**Methods:** Retrospective assessment of cardiac hydatid disease records, between 2007 and 2019, was undertaken to identify patients with cardiac hydatidosis. Hydatid cysts were assessed by transthoracic echocardiography, magnetic resonance imaging, and computed tomography. Clinical symptoms, treatment modalities, and follow-up were also evaluated.

**Results:** Over the study period, 22 (13 males: 59.1%; mean age: 43.1 years; range: 12-63 years) patients with 24 cardiac hydatid cysts were identified. The most common symptom was chest pain, which occurred in 13 (59.1%) patients. Cardiac cysts were located in the left ventricle (n=10, 41.7%), right ventricle (n=5, 20.8%), interventricular septum (n=5, 20.8%), and pericardium (n=4, 16.7%). Extracardiac involvement was present in 14 (63.6%) patients, most commonly affecting the liver (n=10, 45.4%), but in 8 (36.3%) patients, there was no extracardiac organ involvement. Cardiac hydatid cysts were surgically removed in 20 (90.9%) patients.

**Conclusion:** Cardiac hydatid disease is very uncommon, with the left ventricle as the most commonly affected structure in this series. Chest pain was the most common presenting symptom. Extracardiac involvement is common, so patients with cardiac hydatid cysts should be investigated for involvement of other organs.

**Keywords:** Hydatid disease, cardiac hydatid cyst, cardiac echinococcosis, multiple organ involvement, cystectomy, cardiac MRI, transthoracic echocardiography

# ÖΖ

**Amaç:** Hidatik kist hastalarında kardiyak tutulum nadirdir ve tüm hidatidoz olgularının sadece %0,5-2'sinde bulunur. Kardiyak hidatik kistler genellikle ekstrakardiyak tutulum ile beraber ortaya çıkar. Bu makalenin amacı, kardiyak hidatik hastalığın genel özellikleri, görüntüleme bulgularını ve ekstrakardiyak organ tutulum oranlarını tanımlamaktır.

Yöntemler: Kardiyak hidatidoz'lu hastaları belirlemek için 2007 ile 2019 yılları arasında kardiyak hidatik hastalık kayıtlarının retrospektif değerlendirmesi yapılmıştır. Hidatik kist değerlendirmesi, transtorasik ekokardiyografi, manyetik rezonans görüntüleme ve bilgisayarlı tomografi kullanılarak yapıldı. Klinik semptomlar, tedavi yöntemleri ve takipleri değerlendirildi.

**Bulgular:** Çalışma süresi boyunca ortalama yaş 43,1 (dağılım: 12-63) olan 24 kardiyak hidatik kisti olan 22 (13 erkek: %59,1) hasta belirlendi. En sık görülen semptom 13 hastada (%59,1) göğüs ağrısı idi. Kalp hidatik kistlerinin lokalizasyonları şu şekildeydi; sol ventrikül n=10 (%41,7); sağ ventrikül n=5 (%20,8); interventriküler septum n=5 (%20,8); ve perikardiyum n=4 (%16,7). Ekstra kardiyak tutulum 14 (%63,6) olguda mevcuttu, en sık karaciğerde saptandı (n=10, %45,4), ancak sekiz (%36,3) olguda kalp dışı organ tutulumu yoktu. Kalp kist hidatiğinin cerrahi olarak çıkarılması 20/22'de (%90,9) hastada yapıldı.

Sonuç: Kardiyak hidatik hastalığı çok nadirdir ve en sık tutulan yapı bu çalışmada sol ventriküldür. Göğüs ağrısı en sık görülen semptomdu. Ekstra kardiyak tutulum yaygındır, bu nedenle kardiyak hidatik kistleri olan hastalar başka organ tutulumu açısından araştırılmalıdır.

Anahtar Kelimeler: Hidatik kist hastalığı, kardiyak hidatik kist, kardiyak ekinokokus, çoklu organ tutulumu, kistektomi, kardiyak MRG, kardiyak BT, transtorasik ekokardiyografi



Address for Correspondence/Yazışma Adresi: İsa Çam MD, Kocaeli University Faculty of Medicine, Department of Radiology, Kocaeli, Turkey

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Phone: +90 554 333 77 88 E-mail: dr.isa.cam@gmail.com ORCID ID: orcid.org/0000-0001-9551-2364

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## Introduction

Hydatid cyst disease results from a parasitic infection with Echinococcus granulosus. Hydatidosis is still endemic in some countries, including Turkey. Although domestic dogs and cats are the most commonly affected species, humans may become infected after ingestion of viable E. granulosus ova because of poor food hygiene (1). Hydatid cysts may occur in many organ systems; however, in humans, they are most commonly found in the liver (55%-70%) and lungs (20%-30%) (1). Cardiac involvement is rare, presenting in only 0.5%-2% of all cases of hydatidosis, and can occur either as a part of a broader systemic infection or more rarely as the only organ system affected. The most commonly affected cardiac structures are the left ventricle (LV) in 50%-60%, interventricular septum in 10%-20%, and right ventricle (RV) in 5%-15% of patients with cardiac hydatidosis. Less commonly, cysts have been reported in the pericardium (7%), pulmonary artery (6%), and right or left atrium (5%-8%) (2-5). Most patients with cardiac hydatid cyst disease develop symptoms very late, because cardiac cysts grow very slowly. Symptoms of cardiac hydatid cyst include chest pain, dyspnea, syncope, and palpitation, and symptom severity correlates with the size of the cyst and the affected cardiac structures (2-6). On diagnosis of a cardiac hydatid disease cyst, because of the likely involvement of other organ systems, whole body screening should be performed. The main treatment of hydatid disease is surgery with complete removal of the cyst (7). Medical therapies may include albendazole or mebendazole, given before and after surgery (2). This study aimed to present a single tertiary center experience of cardiac hydatid cyst disease and describe the general characteristics, management, imaging findings, range of organ involvement, and follow-up in cases of cardiac hydatid disease.

### Methods

Patients diagnosed with cardiac hydatid cyst with extracardiac organ involvement and presented to the cardiology and radiology departments between December 2007 and December 2019 were enrolled retrospectively in this study. Patient data were extracted from a detailed clinical database of all patients diagnosed with hydatid cyst disease. Details extracted included demographic information, such as age and sex, and other relevant medical history. Clinical symptoms on admission including chest pain, dyspnea, palpitation, syncope, and headache were also evaluated. From each patient, other information obtained included cardiac structures affected by the hydatid cysts, all extracardiac organ system involvement, surgical and medical treatment, preoperative complications, and postoperative follow-up of at least 12 months.

All patients underwent basal electrocardiography, chest X-ray imaging, and echocardiography. Transthoracic echocardiography (TTE) images were obtained for each patient using a Vivid 7 ultrasound system (GE Medical System, Horten, Norway). When hydatid cysts were found in the heart, patients underwent cranial, thoracic, and abdominal computed tomography (CT) and/or magnetic resonance imaging (MRI) to detect the involvement of other organ systems. Cardiac MRI was performed in selected cases using a 3-T Philips MRI unit (Philips Achieva Intera Release, Eindhoven, Netherlands). Non-contrast and contrast-enhanced CT was performed with a 64-channel multidetector CT scanner (Aquilion 64; Toshiba Medical Systems, Tokyo, Japan). Survival and disease status was obtained from the hospital records or, if necessary, by direct communication with the patient or their families.

The study was approved by the Kocaeli University Faculty of Medicine Ethic Committee (approval number: KU-GOKAEK-2020/37). Given the retrospective nature of the study, no specific individual informed consent was required. All patients had given informed consent for the treatment procedure.

#### **Statistical Analysis**

Statistical analysis was performed using SPSS 22.0 software (IBM Corp., Armonk, NY, USA). Continuous data are shown as mean  $\pm$  standard deviation and categorical variables as numbers and percentages.

## Results

In 12 years, 22 (13 males: 59.1%) patients were diagnosed with cardiac hydatid cyst after cardiac examination, and these patients were initially admitted for various cardiac symptoms, including chest pain, dyspnea, palpitation, syncope, and headache. The mean age was 43.1±18.2 years. The baseline demographic, clinical, treatment, follow-up characteristics and imaging findings of patients with hydatid cyst disease are shown in Table 1.

All patients underwent non-contrast and contrast-enhanced CT, while 14 (63.6%) patients underwent cardiac MRI. Twenty (90.9%) patients had a single cardiac hydatid cyst, while the remaining two (9.1%) had two cardiac cysts. Cardiac hydatid cysts were found in the following cardiac regions: LV (n=10, 41.7%), RV (n=5, 20.8%), interventricular septum (n=5, 20.8%), and pericardium (n=4, 16.7%). Common symptoms at admission included chest pain (n=13, 59.1%), dyspnea (n=7, 31.8%), and palpitations (n=5, 22.7%). In addition, one patient (4.5%) presented with syncope due to a complete atrioventricular (AV) block and one (4.5%) was admitted for complaint of headache due to brain hydatid cysts. Finally, one patient presented without symptoms, and the cyst was found incidentally at checkup with chest X-ray imaging.

Lesions were assessed according to Gharbi classification (1). This revealed that 8/24 (33.3%) cases were type 1, 5/24 (20.8%) were type 2, 1/4 (4.2%) was type 3, 3/24 (12.5%) were type 4, and 7/24 (29.2%) were type 5. MRI was performed in 14 patients. In patients who underwent MRI, 4 lesions were hyperintense and 10 lesions were hypointense on T1-weighted images, and 10 lesions were hyperintense and 4 lesions were hypointense in T2-weighted images. CT and TTE were performed in all 22 patients. On CT, 9/24 (37.5%) lesions were mildly hypodense and 14/24 (58.3%) lesions were hypodense. A calcification component was observed on CT examination in seven patients, all with Gharbi type 5 cardiac hydatid cyst. On TTE, 5/24 (20.8%) lesions were hyporechoic, 7/24 (29.1%) lesions were anechoic, and 11/24 (45.8%) lesions were hypoechoic. In patient 2, CT, MRI, and TTE showed LV aneurysmatic dilatation due to a ruptured cardiac hydatid cyst. Imaging findings of cardiac hydatid cysts are shown in Figure 1-7.

There was no extracardiac hydatid disease organ involvement in 8/22 (36.4%) patients. The remaining 14 patients had hydatid organ system involvement, as follows: Liver in 10/22 (45.4%), lung in 5/22 (22.7%), brain in 2/22 (9.09%), and spleen hydatid cysts in 2/22 (9.09%) patients.

a	phic, clinical, diag Location	size (mm)	herapeutic cha Gharbi classification	aracteristics and in MRI	naging findings CT	in 22 patients w TTE	rith cardiac hy Admission symptom	datid cyst disea Extracardiac involvement	ise Treatment	Preoperative complication	Postoperative complication
LV lateral (intraventricular and extraventricu extension)	lar	59x45	Type 2	T1 heterogeneous and hyperintense; T2 heterogeneous and hyperintense	Heterogeneous and hypodense	Heterogeneous and hypoechoic	Chest pain	° Z	Cardiac surgery		
LV inferolateral		42x34	Type 1	Aneurysmatic dilatation	Aneurysmatic dilatation	Aneurysmatic dilatation	Headache	Brain	Cardiac and brain surgery	Vascular embolism	
1. interventricula septum	_	70x30	Type 4		Hypodense	Hyperechoic	Palpitation,			Pulmonary	
2. RV (intraventricular)		46x28	Type 5		Hypodense + peripheral calcification	Hypoechoic	dyspnea, chest pain	Lung		embolism, exitus	
LV lateral		35x30	Type 5	T1 heterogeneous and hypointense; T2 heterogeneous and hypointense	Mildly hypodense + peripheral calcification	Hyperechoic	Chest pain	° Z	Cardiac surgery		
Interventricular septum		54x35	Type 2	T1 heterogeneous and hyperintense; T2 heterogeneous and hyperintense	Hypodense	Anechoic	Chest pain, palpitation,	° Z	Cardiac surgery		Intraoperative exitus and anaphylaxis
Interventricular septum		44x38	Type 4		Mildly hypodense	Hyperechoic	Dyspnea, chest pain, palpitation,	Lung, liver	Surgery for all	1	
LV apex		49x45	Type 1	T1 hypointense; T2 hyperintense	Hypodense	Heterogeneous and hypoechoic	Dyspnea, palpitation.	No	Cardiac surgery		1
Pericardial		78x65	Type 2	ı	Hypodense	Hypoechoic	Chest pain	No	Cardiac surgery	ı	Pulmonary atelectasis
LV inferior		98x76	Type 3	T1 heterogeneous and hypointense; T2 heterogeneous and hyperintense	Hypodense	Heterogeneous and hypoechoic	Chest pain	° Z	Cardiac surgery		
LV inferior		26x24	Type 1	T1 hypointense; T2 hyperintense	Hypodense	Anechoic	Dyspnea	Liver	Cardiac surgery, PAIR for liver		
LV anterolatera	_	48x34	Type 2	1	Hypodense	Hypoechoic	Chest pain	Liver	Cardiac and liver surgery		

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Table 1. G	ontinue	p										
Case	Age, sex	Location	Size (mm)	Gharbi classification	MRI	cı	TTE	Admission symptom	Extracardiac involvement	Treatment	Preoperative complication	Postoperative complication
Patient 12	33, M	RV lateral	37x33	Type 1	T1 hypointense; T2 hyperintense	Hypodense	Anechoic	Chest pain	Lung	Cardiac and lung surgery		Pulmonary atelectasis
Patient 13	48, M	RV lateral	42x24	Type 1		Hypodense	Anechoic	Dyspnea	Liver, spleen	Cardiac and spleen surgery, PAIR for liver		
Patient 14	21, F	LV apex	63x45	Type 1	T1 hypointense; T2 hyperintense	Hypodense	Anechoic	Palpitation	Liver, brain	Cardiac and brain surgery, PAIR for liver		·
Patient 15	58, M	RV lateral	26x32	Type 1		Hypodense	Anechoic	Chest pain	Liver	Cardiac surgery, PAIR for liver		
Patient 16	32, M	RV lateral	35x19	Type 1	T1 hypointense; T2 hyperintense	Hypodense	Anechoic	Chest pain	Lung, spleen	Cardiac, lung, and spleen surgery		
Patient 17	15, M	LV lateral	42x51	Type 2	T1 hypointense; T2 hyperintense	Heterogeneous and hypodense	Heterogeneous and hypoechoic	Chest pain	Lung, liver	Cardiac surgery, lung surgery, PAIR for liver		
Patient 18	69, F	Interventricular septum	21x19	Type 5	T1 hyperintense; T2 heterogeneous hypointense	Mildly hypodense + peripheral calcification	Hyperechoic	Syncope	Liver	Medical treatment		
Patient 19	63, F	L pericardial	53x31	Type 4	T1 hyperintense; T2 heterogeneous and hyperintense	Heterogeneous and hypodense	Heterogeneous and hypoechoic	Chest pain	Liver	Cardiac surgery, liver surgery		
Patient 20	58, M	Interventricular septum	54x42	Type 5	T1 hypodense; T2 heterogeneous and hypointense	Mildly hypodense + peripheral calcification	Hyperechoic	Dyspnea	N	Cardiac Surgery		
	L	L pericardial	93x41	Type 5		Mildly hypodense		c	:	Cardiac and liver		
Patient 21	- 79, F	R pericardial	52x14	Type 5		+ peripheral calcification	Нуроеспоіс	Uyspnea	LIVer	surgery		
Patient 22	60, M	LV apex	35x24	Type 5	T1 hypodense; T2 heterogeneous and hypointense	Mildly hypodense + peripheral calcification	Hypoechoic	Asymptomatic	Q	Medical treatment		
MRI: Magneti cysts	c resonanc	ce imaging, CT: computed	tomograp.	hy, TTE: transthorad	cic echocardiography, F:	female, M: male, L: l	eft, R: right, LV: left ve	entricle, RV: right ve	:ntricle, PAIR: punct	ure, aspiration, injection	and reaspiration fo	or treatment of



**Figure 1.** (Patient 1\*) A 49 year-old female patient with Gharbi type 2 hydatid cyst of the left ventricular lateral wall. The cyst is heterogeneous and hypodense on the post-contrast axial and coronal computed tomography image (arrow, A and B). Extraventricular involvement of cyst is also seen (star, A)



**Figure 2.** (Patient 1\*) The cyst is heterogeneous and hyperintense on the axial and coronal T2-weighted images (arrow, A and C) and heterogeneous and hyperintense on precontrast axial T2-weighted image (curved arrow, B). On coronal T1-weighted, delayed-phase cardiac images after contrast injection, peripheral rim enhancement (curved arrow, D) is seen around the cyst. Extraventricular involvement of the cyst is also seen on the axial T1- and T2-weighted images (arrowheads, A, B)

Of the 8 patients with hydatid disease only affecting the heart, 7/8 (87.5%) underwent open-heart surgery and the eighth patient had medical therapy only. Of those undergoing open-heart surgery, one died intraoperatively because of generalized anaphylaxis (patient 5), while another patient (patient 3) died of pulmonary embolism due to hydatid cyst rupture while being prepared for surgery. A 12 year old male patient (patient 2) had concomitant cardiac and brain surgery for hydatid cyst disease without any postoperative complication. Open-heart surgery and puncture, aspiration, injection, and reaspiration (PAIR) for treatment of cysts in the liver were performed in 5 of 10 patients with cardiac and liver cysts, without any complication, while 4 more of them (18.1%) underwent successful open-heart and liver surgery for cysts. Complete removal of lung hydatid cysts were successfully achieved in four of the five patients with concomitant lung and cardiac involvement, in addition to surgical removal of cardiac cysts. Surgical removal of spleen cysts was



**Figure 3.** (Patient 3\*) A 63 year-old male patient with two cardiac hydatid cysts: one Gharbi type 4 and one type 5. The right intraventricular cyst is hyperechoic on TTE (arrow, A) and hypodense on CT (arrow, B). The interventricular septum cyst is hypoechoic and has hyperechoic peripheral calcification (arrowheads, A). The interventricular septum cyst is hypodense with hyperdense peripheral calcification on axial (arrowheads, B) and coronal (arrowheads, C) CT images. Pulmonary hydatid cysts are also seen on axial (curved arrow, B) and coronal (curved arrow, C) CT images

CT: Computed tomography, TTE: transthoracic echocardiography



**Figure 4.** (Patient 7\*) A 22 year-old male patient with Gharbi type 1 cyst of the left ventricular apex. The cyst is heterogeneous and hypoechoic on transthoracic echocardiography (arrows, A: arrowheads, B)

successful in two patients. A 21 year old female patient (patient 14) had successful open cardiac and brain surgery with PAIR for the liver. Two patients did not undergo surgery because of comorbidities (patients 18 and 22), which included pulmonary embolism, pulmonary atelectasis, and vascular embolism that occurred preoperatively. Preoperative treatment of 400 mg albendazole or mebendazole was given to all patients and continued for 12 months after surgery. On follow-up, the remaining 20 patients continued to have good health status, without any further complications. In addition, there was no cardiac reinfestation in 18 patients, while in the two patients who were on medical therapy alone, cardiac cysts were stable at 1 year follow-up.



**Figure 5.** (Patient 17\*) A 15 year-old male patient with Gharbi type 2 cyst of the left ventricular lateral wall. The cyst is heterogeneous and hypodense on post-contrast axial and coronal CT (arrow, A and B) images. Pulmonary (arrowhead, B) and hepatic (curved arrow, B) hydatid cysts are also seen on the coronal CT image

CT: Computed tomography



**Figure 6.** (Patient 22\*) A 60-year-old male patient with Gharbi type 5 cyst of the left ventricular apex. The cyst is hypoechoic on TTE and has hyperechoic peripheral calcification (arrows, A). On the axial CT image, the cyst is hypodense with hyperdense peripheral calcification (curved arrow, B)

CT: Computed tomography, TTE: transthoracic echocardiography



**Figure 7.** (Patient 22\*) The cyst is hypointense on precontrast axial T1-weighted image (arrows, A) and heterogeneous and hypointense on precontrast axial (curved arrow, B) and coronal T2-weighted image (arrowheads, C). On sagittal T1-weighted, delayed-phase cardiac images after contrast injection, peripheral rim enhancement (arrowheads, D) is seen around the cyst

## Discussion

Cardiac hydatid cysts are rare and account for 0.5%-2% of all cases of hydatidosis. When cardiac hydatid cysts are present, extracardiac organ system involvement is reported to range from 38% to 100% of cases (7-9). In hydatid cyst disease with extracardiac involvement, cardiac hydatid cyst may occur as single or multiple cysts and affect a single or multiple organ systems (10). Cardiac involvement has several possible mechanisms: 1) Infection of the myocardium via coronary arterial supply, 2) infestation as a result of echinococcal cyst rupture into a pulmonary vein, and 3) direct tissue-to-tissue contact with cysts in the liver or lungs (11).

The most common symptom in patients with cardiac hydatid cyst disease is chest pain. However, patients may present with dyspnea, palpitations, and/or syncope, depending on the location and size of the cyst. Angina pectoris may occur if the cyst impedes coronary artery function (2-4). Cardiac conduction impairment, such as AV block or nodal rhythm abnormality, may occur if the cysts compress the AV node or bundle of His (6).

Complications of cardiac hydatid cysts can be severe and fatal, so prompt and accurate diagnosis is important. Cyst rupture is the most dangerous complication, which may cause cardiac tamponade, anaphylactic shock, systemic embolism or combined pulmonary and systemic embolisms, and arrhythmias (11-14). In the present series, one patient died from anaphylactic shock and the other from pulmonary embolism.

Diagnostic methods for cardiac cysts include non-invasive imaging techniques such as those used in this series (TTE, CT, and MRI) and serological tests (15). Serologic tests are more sensitive and specific for liver involvement than for cardiac hydatid disease, although these tests are not capable of positively identifying the organ(s) involved (11). Serological testing, using an enzyme-linked immunosorbent assay, is the most sensitive (94%) and specific (90%) test for most hydatid cyst locations (11). TTE is the first-line imaging modality of choice being widely available, easy to use, and with good sensitivity for cardiac hydatid cyst diagnosis (16). CT is a good second-line modality for cardiac cysts and shows cyst wall calcification well in type 5 cysts. However, cardiac motion artifacts are an important limitation of CT. MRI is excellent at identifying the exact anatomic location of cysts and provides information concerning cystic internal and external structures, thus allowing cysts to be differentiated from other masses and tumors. Hydatid cysts have a characteristic appearance on MRI, usually as a hypointense and hyperintense oval lesion on T1- and T2-weighted images, respectively. In addition, T2-weighted images may show a hypointense peripheral ring caused by the formation of a pericyst consisting of host tissues deposited as a result of the inflammatory response to the cyst (5,15). On suspicion of a hydatid cyst, positive identification of the cystic wall and peripheral contrast enhancement are crucial to distinguishing the cyst from other cardiac masses. Cardiac hydatid disease occurs more often in the LV because of its large myocardial mass and abundant blood supply (2,15). Thus, the LV was the most commonly affected cardiac region in our series.

The definite treatment for hydatid cyst disease is surgery, and an openheart surgery for cardiac cysts is known as cystopericystectomy. Rarely, partial pericystectomy can be performed to preserve organ function (11). In our study, majority (19/22) of the patients underwent total cystopericystectomy during open-heart surgery with cardiopulmonary bypass. Different approaches for cystectomy during open-heart surgery have been described. Birincioğlu et al. (17) suggested that subepicardial cysts can be resected without cardiopulmonary bypass. By contrast, Abhishek and Avinash (11) suggested that cardiopulmonary bypass is still required for optimal safety. Gentle handling of the heart during cardiopulmonary bypass minimizes operative risks and complications, such as cyst rupture (18). Unfortunately, cyst rupture occurred in a 17-year-old female patient (patient 5) in our series, resulting in intraoperative anaphylaxis and death.

Previous reports from small series have indicated extracardiac organ involvement in approximately one-third of all patients (7-9). In our series, an incidence of 63.7% falls at the middle of this range. Thus, given the high likelihood of extracardiac involvement in patients initially diagnosed with cardiac hydatid cyst, we suggest that all patients undergo systemic screening to identify other affected organ systems. In patients with hepatic involvement, the gold standard treatment is complete removal with open surgery, such as through percutaneous, minimally invasive surgery and PAIR (19). In our series, PAIR was successfully performed in four patients with liver hydatid cysts. The two remaining patients with liver disease underwent open surgery without any postoperative complication. For all patients with cardiac hydatid disease, medical therapy with albendazole or mebendazole should be implemented to reduce repeated infections and minimize perioperative complications. This treatment should be started 14 days before surgery and continued for at least 1 year following surgery (6,11,14,20).

#### **Study Limitations**

This study has some limitations. First, this is a single-center retrospective study, with inherent biases. Second, the sample size was small; however, cardiac hydatid cyst is a very rare disease, which also led to the long duration of the study period, a further limitation. This led to some missing data and inevitable variation in clinical management and treatment strategy because of the lack of standard guidelines. Prospective, randomized, and multicenter study is the ideal study design for collection of data concerning cardiac hydatid cyst disease, but this is difficult, given the rarity of the condition.

#### Conclusion

Cardiac hydatid cyst is a very rare condition, and majority of the cases have left ventricular involvement. The most common presenting symptom is chest pain. TTE is the first-line imaging modality of choice, while CT and MRI provide valuable additional findings for the diagnosis of cardiac hydatid disease, including the exact anatomic location. In our cohort, more than two-thirds of patients had extracardiac organ system involvement; thus, all patients should be screened for other cyst locations, especially the liver, lung, and brain. In addition, multisystem involvement is common, so patients presenting with non-cardiac hydatid disease should also undergo cardiac imaging. Surgery is the main treatment modality with adjunctive medical therapy using either albendazole or mebendazole therapy. Given the rarity of the condition, there is little data about the clinical course and outcomes in patients with cardiac hydatid cyst, so further studies are necessary.

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#### References

- Pedrosa I, Saíz A, Arrazola J, Ferreirós J, Pedrosa CS. Hydatid disease: Radiologic and pathologic features and complications. Radiographics 2000; 20: 795-817.
- Dikmen Yaman N, Sirlak M. Cardiac hydatid cysts-review of recent literature. J Vet Med Res 2017; 4: 1102.
- von Sinner, WN. CT and MRI findings of cardiac echinococcosis. Eur Radiol 1995; 5: 66-73.
- Fiengo L, Bucci F, Giannotti D, Patrizi G, Redler A, Kucukaksu DS. Giant cardiac hydatid cyst in children: case report and review of the literature. Clin Med Insights Case Rep 2014; 7: 111-6.
- Petik B, Hazirolan T, Uysal G, Erturk SM. Cardiac hydatid cysts: computed tomography and magnetic resonance imaging findings of the 5 cases. J Comput Assist Tomogr 2015; 39:816-9.
- Fennira S, Kamoun S, Besbes B, Ben Mrad I, Zairi I, Ben Moussa F, et al. Cardiac hydatid cyst in the interventricular septum: A literature review. Int J Inf Dis 2019; 88: 120-6.
- Tuncer E, Tas SG, Mataraci I, Tuncer A, Donmez AA, Aksut M, et al. Surgical treatment of cardiac hydatid disease in 13 patients. Tex Heart Inst J 2010; 37: 189-93.
- Oliver JM, Sotillo JF, Domínguez FJ, López de Sá E, Calvo L, Salvador A, et al. Two-dimensional echocardiographic features of echinococcosis of the heart and great blood vessels. Clinical and surgical implications. Circulation 1988; 78: 327-37.
- Shehatha J, Alward M, Saxena P, Konstantinov IE. Surgical management of cardiac hydatidosis. Tex Heart Inst J 2009; 36: 72-3.
- Yılmaz R, Akpınar YE, Bayramoglu Z, Ozyavuz HI, Unal OF, Dursun M. Magnetic resonance imaging characteristics of cardiac hydatid cyst. Clin Imaging 2018; 51: 202-8.

- 11. Abhishek V, Avinash V. Cardiac hydatid disease: literature review. Asian Cardiovasc Thorac Ann 2012; 20 :747-50.
- 12. Lahdhili H, Hachicha S, Ziadi M and Thameur H. Acute pulmonary embolism due to the rupture of a right ventricle hydatic cyst. Eur J Cardiothorac Surg 2002; 22: 462-4.
- 13. Kopp CW, Binder T, Grimm M, Merl O, Thalhammer F, Ullrich R, et al. Images in cardiovascular medicine. Left ventricular echinococcosis with peripheral embolization. Circulation 2002; 106: 1741-2.
- Bogdanovic A, Radojkovic M, Tomasevic RJ, Pesic I, Petkovic TR, Kovacevic P, et al. Presentation of pericardial hydatid cyst as acute cardiac tamponade. Asian J Surg 2017; 40: 175-7.
- Dursun M, Terzibasioglu E, Yilmaz R, Cekrezi B, Olgar S, Nisli K, et al. Cardiac hydatid disease: CT and MRI findings. AJR Am J Roentgenol 2008; 190: 226-32.

- Tufekcioglu O, Birincioglu CL, Arda K, Fansa I, Saritas A, Karahan M. Echocardiography findings in 16 cases of cardiac echinococcosis: proposal for a new classification system. J Am Soc Echocardiogr 2007; 20: 895-904.
- 17. Birincioğlu CL, Tarcan O, Bardakci H, Saritaş A, Taşdemir O. Off-pump technique for the treatment of ventricular myocardial echinococcosis. Ann Thorac Surg 2003; 75: 1232-7.
- 18. Yaliniz H, Tokcan A, Salih OK, Ulus T. Surgical treatment of cardiac hydatid disease: A report of 7 cases. Tex Heart Inst J 2006; 33: 333-9.
- Nunnari G, Pinzone MR, Gruttadauria S, Celesia BM, Madeddu G, Malaguarnera G, et al. Hepatic echinococcosis: clinical and therapeutic aspects. World J Gastroenterol 2012; 18: 1448-58.
- 20. Noaman H, Rawaf S, Majeed A, Salmasi AM. Hydatid cyst of the heart. Angiology 2017; 68: 765-8.