

Original Article

Clinical Characteristics of Childhood Hydatid Disease: A Single Tertiary Centre Experience from Turkey

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Abstract

Purpose: Hydatid disease is a parasitic infection and it is a major health problem in some areas. We aimed to evaluate the demographic and clinical findings of patients with hydatid disease in our hospital. **Methods:** Between January 2009 and December 2015, patients with hydatid disease were included retrospectively in this study. Demographic characteristics, clinical findings, laboratory and imaging results, treatment modalities and complications were collected. **Findings:** Twenty-eight patients were involved in our study. The median age of patients' was 134 (55-197) months. Most frequently affected organs were liver (71.4%) and lungs (57.1%). In addition cysts were detected in atypical locations. Medical treatment was given in all patients. **Conclusions:** Hydatid disease is an important problem in Turkey. Multiple organ involvement may occur at the same time. Therefore, advanced imaging methods should be used for the detection of localised atypical cysts. Long term outcomes are satisfactory with adequate treatment.

Key words

Children; *Echinococcus granulosus*; Hydatid disease; Treatment outcomes

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Introduction

Hydatid disease (HD) is a zoonotic infection which is caused by larval forms of *Echinococcus* species. The adult form is located in intestinal lumen of dogs. Infected eggs are dispersed into the environment with animal feces. Intermediate host (sheep, goats, etc.) eats contaminated food and parasite eggs drop to the intestine of animals. It may cause cysts formation in organs by absorption into the bloodstream. Then carnivores eat the animals' organs which contain cysts, and the parasite becomes adult form by reaching the animal's intestine again. Humans may be infected by ingestion of parasitic eggs, occasionally may become an intermediate host.¹

Hydatid disease is a major problem in large sheep raising areas like Mediterranean, South America, Asia, Europe and Kenya.² The incidence is reported <1 to 220 per 100000 in endemic areas.³ Although it is seen in all regions of Turkey, most patients are reported from Eastern Anatolia, Southeastern Anatolia and Central Anatolia. The incidence is approximately 2-6/100000 in Turkey.⁴ Hydatid disease is more frequently reported in children compared to adults. Liver, lungs, spleen, kidney, heart, bones and central

nervous system may be affected.⁵

We aimed to examine epidemiologic characteristics, clinical and laboratory findings of patients with HD and to compare our data with previous studies.

Methods

Patients admitted to our hospital between January 2009 and December 2015 with diagnosis of HD, were retrospectively included in this study. Patients who did not come to routine examinations were excluded. Hydatid disease was diagnosed with clinical, radiological and serological tests. Pathological examination was performed in all patients who underwent surgical procedures. Demographic characteristics like patients' age, gender, contact with an animal, symptoms, clinical and laboratory findings, echinococcal indirect hemagglutination test (IHA) results, radiological imaging, treatment modalities and complications were collected from data system by using ICD-10 codes. Chest X-ray, abdominal ultrasonography, echocardiography and brain magnetic resonance imaging (MRI) were performed in all patients. Thorax and abdomen computed tomography (CT) was used in some cases that required advanced imaging. Echinococcal IHA result $\geq 1/32$ was accepted as positive. All patients received albendazole (15 mg/kg/day in two divided doses) treatment after diagnosis. Radiologic imaging was periodically performed to non-operated patients. Disappearance of cysts, calcified cysts and collapse were accepted as a complete healing and treatment was discontinued. Puncture-aspiration-injection-reaspiration (PAIR) was performed in some patients with hepatic hydatid cysts. While albendazole treatment was given to operated patients for one month after the surgery, non-operated patients received treatment at least 6 months.

Results

Thirty-one patients were diagnosed as a HD between January 2009 and December 2015. Three of them were excluded because they did not come to routine examination. Twenty (71.4%) patients were male. The median age of patients' was 134 (55-197) months. A history of animal contact was found in 35% of boys (7/20) and in 25% of girls (2/8).

Most common complaints were cough, fever, abdominal pain, nausea and vomiting. In addition, 3 (10.7%) patients

were asymptomatic and they were diagnosed incidentally. Two of these patients who were admitted to hospital because of chest pain, were diagnosed during transthoracic echocardiographic examination. Another patient was diagnosed with abdominal ultrasonography which was performed after trauma. Most common physical finding was decreased breath sounds. Pulmonary crackle, hepatomegaly, hepatosplenomegaly, convulsions and abdominal mass were detected as other findings. Physical examination was found completely normal in 9 (32.1%) patients. But history and radiographic images of these patient support HD. Patients' clinical symptoms and physical examination findings are summarised in Table 1.

According to localisation; HD was found in as follows: 20 (71.4%) liver, 16 (57.1%) lungs, 2 (10.7%) spleen, 2 (7.1%) heart, 2 (10.7%) brain, 1 (3.6%) pancreas, 1 (3.6%) kidney and 1 (3.6%) pelvis. While 5 (17.9%) patients had only lung involvement; in 10 (35.7%) patients cysts were

Table 1 Clinical symptoms and physical examination findings of patients

	n (%)
Symptoms	
Cough	11 (39.3)
Fever	10 (35.7)
Abdominal pain	8 (28.6)
Nausea	7 (25.0)
Vomiting	7 (25.0)
Haemoptysis	4 (14.3)
Chest pain	3 (10.7)
Asymptomatic	3 (10.7)
Shortness of breath	5 (17.9)
Purulent sputum	3 (10.7)
Weight loss	2 (7.1)
Urticaria	2 (7.1)
Physical examination findings	
Decreased breath sounds	11 (39.3)
Pulmonary crackle	4 (14.3)
Hepatomegaly	2 (7.1)
Hepatosplenomegaly	1 (3.6)
Convulsions	2 (7.1)
Abdominal mass	1 (3.6)
Normal	9 (32.1)

seen in only liver. Hydatid disease was detected in 37.5% (6/16) right lobe, 37.5% (6/16) left lobe and 25% (4/16) both lobes of lungs. In addition it was found in 45% (9/20) right lobe, 40% (8/20) left lobe and 15% (3/20) both lobes of liver (Table 2). Some radiographic imaging of patients with hydatid cyst are presented in Figures 1-4.

Echinococcus IHA was not performed in 2 patients who were previously diagnosed at another hospital. This test was found positive in 69.2% (18/26) of the patients.

Treatment modalities were as follows: 60.7% (17/28) surgical treatment, 14.3% (4/28) interventional radiologic drainage, 21.4% (6/28) only medical treatment and 3.6%

(1/28) interventional radiology drainage and surgery treatment together (Table 3). All patients received albendazole treatment in various times and any drug side effects were not observed.

Post-operative complications were seen in 4 (14.3%)

Table 2 Localisation of cyst hydatid lesions

Localisation	n (%)
Liver	10 (35.7)
Lung	5 (17.9)
Liver+Lung	7 (25.0)
Liver+Lung+Spleen	1 (3.6)
Liver+Lung+Heart+Spleen	1 (3.6)
Lung+Renal	1 (3.6)
Liver+Lung+Pelvis	1 (3.6)
Heart+Brain+Pancreas	1 (3.6)
Brain	1 (3.6)

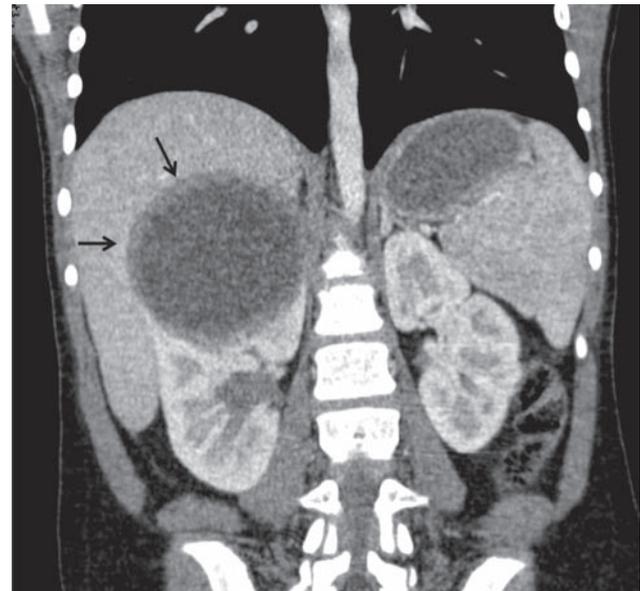


Figure 2 A 9-year-old girl, coronal reformatted contrast-enhanced CT reveals a thick-walled right renal hydatid cyst (arrows).

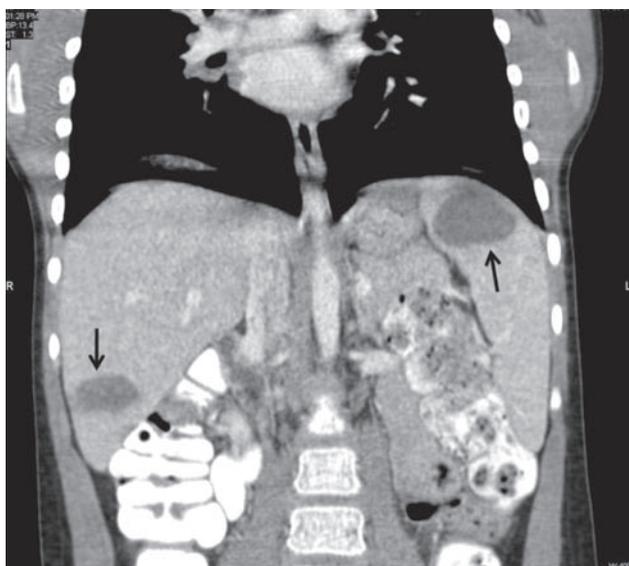


Figure 1 Contrast enhanced CT coronal reformatted image shows hepatic and splenic hydatid cysts (arrows).

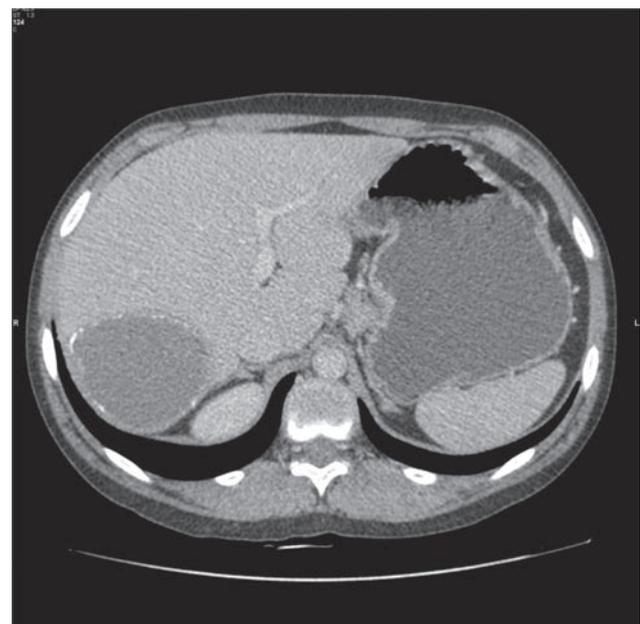


Figure 3 Axial contrast-enhanced CT; liver hydatid cyst, wall calcifications are noted.

patients. While bile leakage and cholangitis occurred in 2 patients with hepatic hydatid cysts, pneumothorax and lung fistula were seen in 1 patient with pulmonary hydatid cysts. Post-operatively infected cyst was detected in 1 (3.6%) patient. Additionally during preoperative period, cyst was infected in 5 (17.9%) patients with lung hydatid cyst.

No mortality occurred, but recurrence was seen in 1 (3.6%) patient with brain hydatid cyst. This patient was operated and received albendazole treatment for 6 months. After the surgery, the cyst had completely disappeared. However, one year after operation, cyst occurred in the brain again. The patient was operated on a second time and received albendazole treatment for 6 months. The cyst disappeared at the end of therapy.

Treatment failure was detected in 1 patient with kidney hydatid cyst who received only medical treatment. Surgical features, postoperative complications and recurrence are summarised in Table 4.

Discussion

Hydatid disease is a parasitic infection which is caused by larval form of the genus *Echinococcus*. Most responsible species are *Echinococcus granulosus* and *Echinococcus multilocularis*. It is a major health problem in some areas where farming is widespread.³ This disease may be acquired in childhood, however diagnosis may be delayed due to long incubation period. In literature many centers report

their experience of childhood HD.⁶⁻¹³ In this study, we aimed to evaluate our experience of childhood HD and to compare our results with previous studies.

Hydatid disease is more frequently reported in boys than girls. This difference was considered due to more contact with animals in boys.¹⁴ We reported a history of animal contact was found in 35% of boys and 25% of girls. Our study supports the hypothesis above.

Although HD may be seen in many organs; liver and lungs are commonly involved. Lungs are affected more often than liver in children.^{11,15} Hepatic involvement is most frequently seen in adults.³ In addition cysts may be detected atypical in locations such as heart, brain, spleen, pancreas, kidney, eye and pelvis. Çakır et al analysed 41 pediatric patients with HD which was detected in 37% lungs, 35% liver, 17% both lungs and liver, 5% spleen, 2% both spleen and heart, 2% brain, 2% both brain and heart. Hydatid cysts were identified in 29% right lobe, 15% left lobe and 10% both lobes of lungs. In addition 34% right lobe, 12% left lobe and 7% both lobes of liver involvement were seen in children.¹¹ However there were some studies showing opposite results.^{6,9} In Tiryaki et al's study 101 children who were operated because of hydatid cysts were examined retrospectively. They found HD in 32% lungs, 48% liver, 16% both lungs and liver, 2% spleen, 1% retrovesical and 1% retroperitoneal region.⁹ Djuricic et al's study included 149 children with 272 hydatid cysts. They reported HD was detected in 60.7% liver, 30.1% lungs and 9.2% other localisations (omentum, peritoneum, intestine, spleen,

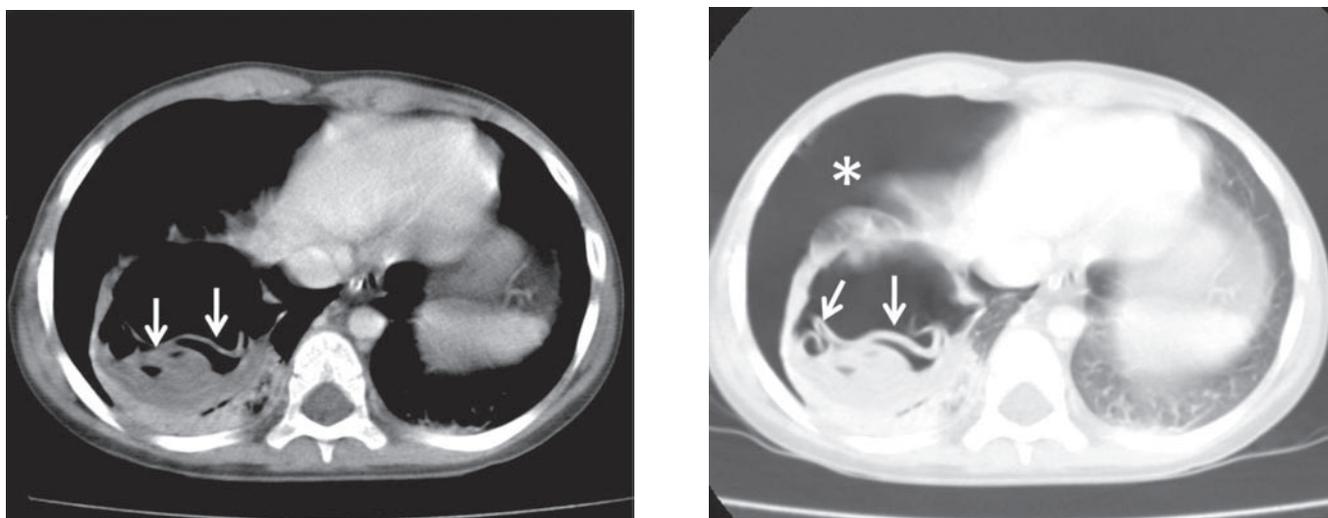


Figure 4 CT of thorax axial images in soft tissue (A) and lung (B) window demonstrate ruptured lung hydatid cyst. Detached membranes (arrows) and fluid collections and right sided pneumothorax (asterisk) is visible.

kidney, abdomen, heart, pancreas and abdominal wall). Both lungs and liver involvement were identified in 9 (6.0%) patients.⁶ We found liver was more frequently affected than lungs and some cysts were detected in atypical localisation.

In our study cardiac involvement was found in 2 patients. In literature cardiac hydatid cysts are rare and it is reported 0.2-3% of all cases. The most common affected areas are the left (75%) and right ventricles (18%) and the interventricular septum.¹⁶ In our patients, left ventricle and interventricular septum involvement were detected. Both patients had also operated and they were completely recovered.

This disease may have different number of cyst. As single cyst may be detected in single organ, multiple cysts may occur in multiple organs. Djuricic et al identified single cyst in 59 patients, multiple organ involvement was seen in 18 children. Additionally in 24 patients, multiple cysts were detected in the same organ.⁹ In our study, 16 patients had one organ involvement and single cyst was found in single organ in 10 patients. Our study supports single cysts are more common in children.

Clinical manifestations of HD may be seen according to

localisation, size, number and condition of cyst. While abdominal pain, vomiting, hepatomegaly, obstructive jaundice may occur in hepatic hydatid cysts; cough, fever, dyspnea, haemoptysis and chest pain may be seen in pulmonary hydatid cysts. Symptoms may appear early in lungs and brain tissue because of weak tissue support.¹⁷ Oral et al found abdominal pain, tenderness, abdominal mass and fever were common symptoms in hepatic hydatid cysts.⁷ Tiryaki et al detected abdominal pain and cough were the most prevalent clinical findings.⁹ In our study, patients were frequently admitted to hospital with abdominal pain, cough, and fever. Although liver was more often affected than lungs, patients were frequently admitted to hospital with pulmonary symptoms. It supports that clinical symptoms occur earlier in lungs. Brain hydatid cysts may cause headache, seizures, nausea, vomiting, increased intracranial pressure syndrome and cranial nerve palsy. Our patients with brain HD had headache and seizures.

Chest X-ray may show pulmonary hydatid cysts and gives information whether the cyst is intact, ruptured and rupturing of cysts indicates specific radiologic findings like "meniscus sign and snake sign". Ultrasonography is useful test for diagnosis and follow up. According to the WHO classification; cysts are seen in 5 stages. Type I cysts are unilocular and simple which may contain uniform anechoic content. Type II cysts are multivesicular and multiseptated. Daughter cysts may fill the unilocular mother cyst. Type III cysts are unilocular and they may contain daughter cysts and echoic areas. Type IV cysts show heterogeneous echo pattern; and type V cysts have a calcified wall.¹⁸ Other imaging techniques such as computed tomography (CT), echocardiography and MRI are used to identify detailed anatomical location and appearance of cysts (daughter cysts, ruptured or calcified cysts).¹⁹ In this study, tests above were sufficient for diagnosis.

There is no consensus about ideal serological test for HD. Echinococcus IHA, specific immunoglobulin (Ig)E,

Table 3 Treatment modalities in patients with hydatid cyst

	n (%)
Liver	
Surgery + medical	11 (68.8)
Only medical	5 (31.3)
Liver	
Surgery + medical	5 (25.0)
PAIR+ medical	6 (30.0)
Only medical	9 (45.0)
Pelvis	
Only medical	1 (100.0)
Spleen	
Surgery + medical	1 (50.0)
Only medical	1 (50.0)
Heart	
Surgery + medical	1 (50.0)
Only medical	1 (50.0)
Renal	
Surgery + medical	1 (100.0)
Brain	
Surgery + medical	2 (100.0)

PAIR: puncture-aspiration-injection-reaspiration

Table 4 Surgical features, postoperative complications and recurrence

Surgery features	PAIR	Surgery	Only medical
Postoperative complications			
Bile leakage+cholangitis	1	1	0
Pneumothorax+fistula	0	1	0
Infection	1	0	0
Recurrence	0	1	0

PAIR: puncture-aspiration-injection-reaspiration

IgM, and IgG enzyme-linked immunosorbent assay, latex agglutination or immunoelectrophoresis tests are used for supporting the diagnosis. Serologic titers may be increased due to antigen release within 3 months after surgery. Immunoelectrophoresis, specific IgE and IgM antibody titres reduce and become negative within the postoperative second year. If recurrence occurs, their levels rise again. Serum IgG titer remains positive for a long time after successful operation.^{20,21} Because of all these reasons, use of serological tests for diagnosis and evaluation the response of treatment may lead to false results. In this study, Echinococcus IHA was 78% positive. Specific IgE, IgM and IgG tests were not used routinely for diagnosis and follow-up.

Medical, surgical treatment and PAIR are used for therapy. Surgery is the first choice treatment for cysts which have following criteria: large cysts with multiple daughter cysts, superficial location amenable to rupture, cyst compressing the neighboring organs and cysts in atypical locations such as brain, bones, spleen, kidneys, heart.²²⁻²⁴ In this study, surgery was the most preferred treatment modality. Antimicrobial treatment (albendazole or mebendazole) is given to prevent the spread of protoscolex into the abdominal cavity during surgery and it is started one week before the surgery and continued until at least postoperative four weeks.²⁵

While postoperative complications are reported in less than 1% of cases; recurrence is seen 2-25% in literature.^{26,27} In Ran et al's study 26 children who underwent radical surgery were compared with 86 pediatric patients with conservative surgery. They found that biliary complications and recurrence were more commonly seen in conservative surgery group than in radical surgery group. However, there was no significant difference between these groups.²⁸ In our study post-operative complications after treatment were detected in 14.3% of the patients.

Recurrence was seen in 1 (3.6%) patient with brain HD. The patient was admitted with seizures and vomiting to our hospital. Brain CT showed cyst in the right parietal region of the brain and she was successfully operated. However five months after the operation, she was admitted again with seizures and headache. Radiologic imaging showed hydatid cyst in right occipital region of brain. The patient was operated again and on follow up hydatid cyst did not occur again. Although recurrence is seen very rarely in the literature, some cases with recurrent HD were reported at the postoperative period.^{29,30}

In one patient with kidney hydatid cyst who had received only medical treatment, the size of the cyst was increased

and it was considered treatment failure. The patient was operated and there were no complications during follow-up. Mortality has also been reported rarely. Jordanova et al's study included 2005 children and 7366 adult patients with HD. They detected 3 patients died due to anaphylaxis which occurred after spontaneous rupture of cysts.³¹ In our study, there was no mortality.

In conclusion, HD should be considered in patients with suspicious clinical and radiological findings in endemic areas. According to affected organs, clinical signs may vary. Multi organ involvement should be considered. Advanced imaging methods such as abdominal ultrasonography, echocardiography and brain MRI should be performed in all patients for the detection of atypically located cyst. Serological tests may be used for supporting the diagnosis. Clinical results are satisfactory with adequate treatment.

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