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ORIGINAL ARTICLE
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# Hepatic Hydatid Cyst in Children: An Experience from East of Turkey

## Çocuklarda Karaciğer Hidatik Kisti: Türkiye'nin Doğusundan bir Deneyim

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## ABSTRACT

**Aim**: Hydatid cyst is a parasitic disease caused by the infection of Echinococcus granulosus and rarely Echinococcus alveolaris eggs, which are endemic in our country. Current treatment options for liver hydatid cysts; They can be listed as medical treatment, surgical treatment, percutaneous drainage and only clinical follow-up.

**Material and Method**: Child patients between the ages of 0-16 who were diagnosed with KcKH and followed-up and treated were evaluated retrospectively. In pre-operative evaluation, lung radiography, abdominal ultrasonography, serological tests, hemogram and liver function tests were routinely performed.

**Results**: 250 pediatric patients between the ages of 0 and 16, whose data were fully available, were included in the study. 118 of the cases were girls and 132 were boys. The average age was 11.2 years. Most of the KcKH patients were admitted to the hospital for another reason and the cyst was detected incidentally. Of the 65 patients requiring surgery, 17 were patients who initially underwent primary intervention with PAIR. Patients who underwent PAIR were observed for one night and discharged in an average of 18 hours. Patients with free drainage were discharged in 2-4 days. The average length of stay for patients who underwent open surgery was found to be 9 days.

**Conclusion**: The Gharbi classification updated by WHO is effective and reliable in determining the KcKH treatment strategy. In cases who are receiving chemotherapy and an intervention decision is made, a final USG is performed just before the procedure; It can provide both a change in treatment management and more patients benefiting from medical treatment.

**Keywords**: Liver, cyst hydatic, child

## ÖZ

**Amaç**: Hidatik kist, ülkemizde endemik olan Echinococcus granosus ve nadiren Echinococcus alveolaris yumurtalarının enfeksiyonunun neden olduğu paraziter bir hastalıktır. Karaciğer hidatik kistlerinde güncel tedavi seçenekleri; Bunları medikal tedavi, cerrahi tedavi, perkütan drenaj ve sadece klinik takip olarak sıralayabiliriz.

**Gereç ve Yöntem**: Karaciğer kist hidatik tanısı konularak takip ve tedavi edilen 0-16 yaş arası çocuk hastalar geriye dönük olarak değerlendirildi. Ameliyat öncesi değerlendirmede rutin olarak akciğer grafisi, batın ultrasonografisi, serolojik testler, hemogram ve karaciğer fonksiyon testleri yapıldı.

**Bulgular**: Verileri tam olarak mevcut olan 0-16 yaş arası 250 pediatrik hasta çalışmaya dahil edildi. Vakaların 118'i kız, 132'si erkekti. Ortalama yaş 11,2 idi. KcKH hastalarının çoğu başka bir nedenden dolayı hastaneye başvurmuş ve kist tesadüfen tespit edilmişti. Ameliyat gerektiren 65 hastanın 17'si başlangıçta PAIR ile birincil müdahale uygulanan hastalardı. PAIR uygulanan hastalar bir gece gözlem altında tutuldu ve ortalama 18 saatte taburcu edildi. Serbest drenajı olan hastalar 2-4 gün içinde taburcu edildi. Açık ameliyat olan hastaların ortalama kalış süresi 9 gün olarak belirlendi.

**Sonuç**: DSÖ tarafından güncellenen Gharbi sınıflaması Karaciğer kist hidatik tedavi stratejisinin belirlenmesinde etkili ve güvenilirdir. Kemoterapi alan ve müdahale kararı verilen vakalarda işlemden hemen önce son ultrasonografi yapılır; Hem tedavi yönetiminde değişiklik yapılmasını hem de daha fazla hastanın tıbbi tedaviden faydalanmasını sağlayabilir.

**Anahtar Kelimeler**: Karaciğer, kist hidatik, çocuk

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## INTRODUCTION

Hydatid cyst is a parasitic disease caused by the transmission of Echinococcus granulosus and rarely Echinococcus alveolaris eggs, which is endemic in Turkey. The orally ingested eggs enter the portal venous system through the mucosa of the upper gastrointestinal tract and evolve into the larval stage in the last organ to which they attach. Cysts are localized in the liver with a rate of 50-70%, in the lung with a rate of 20-30% and rarely in the spleen, kidney, heart, bone, central nervous system and other organs(1- 3). Current treatment options for liver hydatid cysts include medical treatment, surgical treatment, percutaneous drainage and clinical monitoring only (2,3).

In 2011, a new treatment strategy was proposed by WHO-IWGE based on Gharbi's radiologic classification(4,5). In this study, we evaluated the efficacy of the WHO-IWGE (**Table 1**) protocol in terms of diagnosis and treatment strategies in our pediatric patients with hepatic hydatid cyst in a tertiary care institution in eastern Turkey in the light of the literature.

**Table 1: WHO-IWGE classification of the hydatid cyst**

| Stage | Ultrasonographically aspect according to WHO-IWGE Classification |
|---|---|
| CL | Anechogenic uniloculated cyst, with no echoes or internal sepsis |
| CE 1 | Anechogenic cyst, with fine echoes inside, representing the hydatic sand - active cyst |
| CE 2 | Cyst with multiple septums at the interior, giving it a multivesicular aspect or "honeycomb" aspect,with a uniloculated primary cyst - active cyst |
| CE 3 | Uniloculated cyst with decolated proligere membrane ("waterlily sign") (CE3a) or daughter vesicles associating hypo/hyperechogene images (CE3b) - cyst in transition phase |
| CE 4 | Cyst with mixed content, hypo/hyperechogenic, without daughter vesicles - "wool clew" aspect-cyst in the degenerative phase |
| CE 5 | Cyst with partial or totally calcified wall - inactive cyst |

## MATERIAL AND METHOD

Ethical approval for this study was received from Van Training and Research Hospital, dated 03.10.2019 and numbered 2019/18.

We retrospectively evaluated pediatric patients aged 0-16 years who were diagnosed and followed-up and treated for hepatic hydatid cyst between 2015 and 2024. Pre-operative chest radiography, abdominal ultrasonography, serologic tests, hemogram and liver function tests were routinely performed. All patients were started on Albendazole 10mg/kg in 2 doses at least 3-4 weeks before surgery or percutaneous intervention. All hepatic hydatid cysts were radiologically classified according to Gharbi. The Gharbi classification was updated by the World Health Organization (WHO) Echinococcus Working Group(5).

According to the WHO classification, CE1 and CE3a cysts larger than 5 cm were treated with PAIR (puncture - aspiration - scolocidal injection - reaspiration) or PAIDS (puncture - aspiration - scolocidal injection - drainage catheter placement - sclerosing agent injection) and those smaller than 5 cm were treated medically. CE2 and CE3b cysts larger than 5 cm were treated surgically due to the risk of perforation, and cysts smaller than 5 cm were treated medically. All cysts that were found to be associated with the biliary tract underwent surgical intervention regardless of size. CE4 and CE5 cysts were followed up without treatment regardless of size.

In patients eligible for percutaneous intervention, US-guided PAIR technique (for cysts smaller than 6 cm) and PAIDS catheterization technique (for cysts larger than 6 cm) were used. We used 20% hypertonic saline solution as a scolocidal agent and 95% pure alcohol as a sclerosing agent. Ul

The results were statistically assessed by using SPSS version 24. Normality controls were done using Shapiro-Wilk Test. Groups were compared in terms of mortality using independent sample t-test. Statistical significance level was set as $p<0.05$.

## RESULTS

250 child patients between the ages of 0 and 16, whose data were fully available, were included in the study. 118 of the cases were girls and 132 were boys. The average age was 11.2 years.

Most of the hepatic hydatid cysts detected at the time of initial admission were type 1 hydatid cysts (**Figure 1**). No statistically significant difference was found in the ratio of girls to boys ($p>0.05$). No significant relationship was found between the radiological type of the cyst and the average age. The cyst size of patients with type 1 and type 3 hydatid cysts was found to be significantly larger. This was attributed to the growth tendency of active cysts. Additional organ involvement was more common in active cysts (20%).

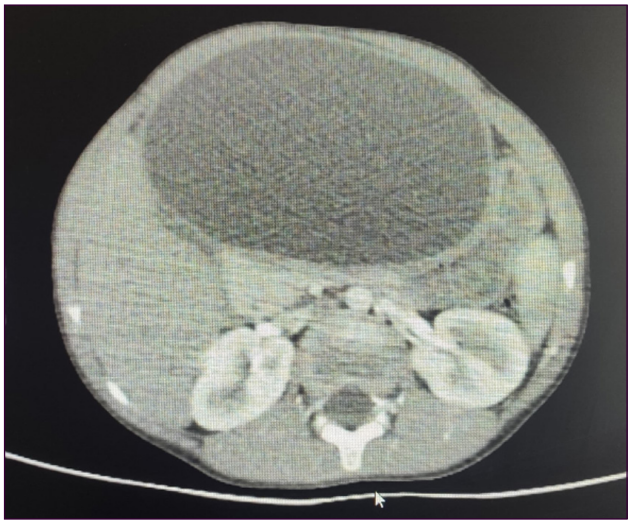

**Figure 1.** CT of Type 1 Cyst Hydatid of left hepatic lobe

Most of the hepatic hydatid cyst patients were admitted to the hospital for another reason and the cyst was detected incidentally. Symptomatic patients often presented for reasons such as abdominal pain and a feeling of abdominal fullness. These symptoms were frequently detected in children over the age of 10 who could express themselves easily. The patients diagnosed as a result of our screening were siblings or other family members who shared the same environmental environment.

Of the 65 patients requiring surgery, 17 were patients who initially underwent primary intervention with PAIR. The remaining 48 patients underwent primary surgical intervention. Infection developed in the cyst site in 4 of the patients who underwent surgery. In 10 of these patients, the bile ducts were sutured due to bile leakage. Bile drainage lasting more than 6 weeks developed in 4 of them. Of the 6 patients who were found to have bleeding from the drain in the first week after surgery, bleeding from the abdominal drain stopped spontaneously on the 3rd day on average. Surgical intervention was required in 17 of the patients who underwent PAIR because of bile leakage or lack of cyst resolution. In 4 patients, PAIR was preferred again. All cysts that were smaller than 5 cm, did not show exophytic extension, were unlikely to rupture due to trauma, and had a change in type with medical treatment were followed up with medical treatment.

Hydatid cyst was positive in 62% of our patients. Almost all of the patients were diagnosed with ultrasound (97%) and treatment was planned, in line with the literature. In patients with additional complications whose relationship with the vascular structures and bile ducts could not be clearly determined, the surgical decision was made according to the CT (3%) result.

Patients who underwent PAIR were observed for one night and discharged in an average of 18 hours. Patients with free drainage were discharged in 2-4 days. The average length of stay for patients who underwent open surgery was found to be 9 days.

Cure was achieved after 3-6 months of chemotherapy in 21 patients (8%) whose type change was detected according to the Gharbi classification in the pre-procedural ultrasonography, although the decision for PAIR, PAIDS or surgery was made at the first application.

## DISCUSSION

Eggs excreted in the feces of dogs, the main host of Echinococcus granulosus, are the main source of this type of infection in both livestock and humans. Hepatic hydatid cyst disease is widespread globally, especially in endemic regions. In our country, a high prevalence of parasites is observed in the Eastern and Southeastern regions, where livestock farming is the main livelihood (6, 7, 8). The hepatic hydatid cyst disease mostly manifests itself as a solitary and single cystic lesion in the right lobe (9). Hepatic hydatid cyst disease has a growth rate of approximately 1-3 cm in diameter per year (8). Clinical findings vary depending on the mass compression effect of the cyst and its complications. Physical examination findings are not guiding in the diagnosis, since hepatic hydatid cyst disease usually does not give a prominent clinical sign. However, if the cyst is complicated or ruptured, it produces a distinct clinical presentation.

In a study by Cıgşar et al. in which abdominal ultrasonography was performed in 2138 patients who presented to the emergency department in city of Turkey for any reason, incidental detection of hepatic hydatid cyst disease was found in 96 of the patients (1/22)(8). The prevalence of the disease was found to be 1.05% in our country (3).

Although the diagnosis can be made with anamnesis, clinical findings, laboratory tests and radiologic examinations, surgical excision and histopathologic examination are required for definitive diagnosis (1,7,8).

Among laboratory tests, ELISA basic immunodiagnostic tests are frequently used but their contribution to the diagnosis is limited(5). Its sensitivity is reported between 85-98% (5,7). As organ involvement increases, sensitivity may increase to 90-100% (7). There may be cases with negative serology results despite having hydatid cysts as well as cases with false positive results despite the absence of cysts(5). In our cases, the most common mode of presentation was incidental, and it was learned that these patients were diagnosed by ultrasound performed in an external center for another reason. The most common clinical finding was nonspecific abdominal pain.

Radiologically, the classification system proposed by Gharbi is used in hepatic hydatid cyst disease. Abdominal ultrasonography, which is an inexpensive and easily applicable method, should be preferred for the diagnosis (8,10). CT is the best method to determine the type and location of the cyst, but high dose radiation and cost are the most important disadvantages (10).Even if cross-sectional imaging modalities are used to determine cyst typing and treatment modality in hydatid cysts, each patient should be evaluated by ultrasonography by the same radiologist before the procedure(5).

Diagnosis; although it can be diagnosed by anamnesis, clinical findings, laboratory tests and radiological examinations, surgical excision and histopathological examination are required for a definitive diagnosis (1,7,8). Among laboratory tests, ELISA basic immunodiagnostic tests are frequently used, but their contribution to diagnosis is limited (5). Its sensitivity is reported to be between 85-98% (5,7). As organ involvement increases, sensitivity can increase to 90-100% (7). While there may be cases with negative serology despite having hydatid disease, there may also be cases with false positive results even though they do not have a cyst (5). In

our cases, the most common presentation was incidental; it was learned that these patients were diagnosed with an abdominal ultrasoundperformed for another reason at an external center. The most common clinical finding was nonspecific abdominal pain.

In hepatic hydatid cyst disease, the radiological classification system suggested by Gharbi is used. In diagnosis, abdominal ultrasonography, which is a cheap and easily applicable method, should be preferred first (8,10). CT is the best method to determine the type and location of the cyst, but high dose radiation and cost are its most important disadvantages (10). Even though cross-sectional imaging modalities have been performed to determine cyst typing and treatment in hydatid cysts, each patient must be evaluated with ultrasonography by the same radiologist before the procedure (5). Thus, by detecting the type change, more patients can benefit from medical treatment. In all our cases, the diagnosis was confirmed by abdominal ultrasonography and serological tests.

Complications such as allergic reaction due to spontaneous, traumatic, iatrogenic rupture, secondary infection and cholangitis are seen in 2%-4% of hepatic hydatid cyst disea (5,8,11,12). The most common complication is cystobiliary fistula. The aim of hydatid cyst treatment should be interventions to prevent infection, obstruction, rupture to neighboring organs and anaphylaxis. Since chemotherapy will be added to each patient regardless of the treatment method used, hemogram and liver fonction tests should be monitored before starting the treatment to monitor the side effects of chemotherapy (5,8,10,11).

Treatment methods can be examined under four headings: surgical, medical, percutaneous and follow-up (2,3). Surgical treatment was the traditional method in the treatment of hydatid cyst until recently, but in the last two decades, percutaneous intervention procedures and medical treatment; They have become the preferred methods instead of surgery in selected cases. Surgery is the definitive treatment method in the treatment of hepatic hydatid cyst disease, but its mortality, morbidity and recurrence rates are higher than other methods. While complication rates are reported to be between 1.7-8.6% (8,11) in the literature, mortality rates of up to 25% have also been reported, especially in cases of rupture (8). Major complications of surgery are bleeding, sepsis and fistula formation(5,8,11). Omentoplasty, capitonnage, simple closure, deroofing and external drainage (12,13) are the most commonly used methods in the management of the residual cavity. ERCP can be used effectively in the treatment of biliary fistula in the preoperative and postoperative periods (5,9). In hepatic hydatid cyst disease patients, ERCP is performed when intrabiliary rupture is suspected before the operation or when there is persistent icterus in the postoperative period, common bile duct pathologies in radiological imaging, or laboratory abnormalities; It can be used effectively in the diagnosis and treatment of these undesirable conditions (9).

Although it is reported that the results of medical treatment are not satisfactory, in selected cases, it can be used as a stand-alone treatment method, and in many cases, it is used as a complementary agent before or after surgical or percutaneous intervention (2,5). Preoperative use of chemotherapy reduces the viability of the cyst, and postoperative use reduces the risk of recurrence. Albendazole and mebendazole are most commonly used as chemotherapeutic agents in the treatment of hepatic hydatid cyst disease (2,6). It is recommended that medical treatment be used alone, especially for CE1 - CE3a (iwge) cysts smaller than 5 cm (3,5,12).

Percutaneous drainage of hepatic hydatid cyst in children under ultrasonography guidance is an effective method. Treatment success in suitable cases with the PAIR/PAIDS method is reported to be over 95% (2). Percutaneous treatment is widely used and minimal morbidity is reported (2,3,5,12). The aim of percutaneous treatment is to drain the endocyst and destroy the germinal membrane. The best results with PAIR treatment are seen in active unilocular cysts (IWGE CE1 or GHARBI Type I) and cysts with a separated membrane (IWGE CE3a and GHARBI Type 2). In the PAIR method, the presence of daughter vesicles (**Figure 2** ) in the cyst content reduces the success of treatment, so it is important to classify the cyst correctly at the beginning. In the presence of a daughter vesicle, cyst drainage is provided by placing a large lumen percutaneous catheter (modified catheterization) instead of the PAIR method. In addition, PAIR is contraindicated in cysts related to the biliary ducts due to the risk of the sclerosing cholangitis caused by the scholicidal agent. After percutaneous intervention, albendazole treatment should be continued for 3 months (2,3,12). The average follow-up period is 24 months (5).

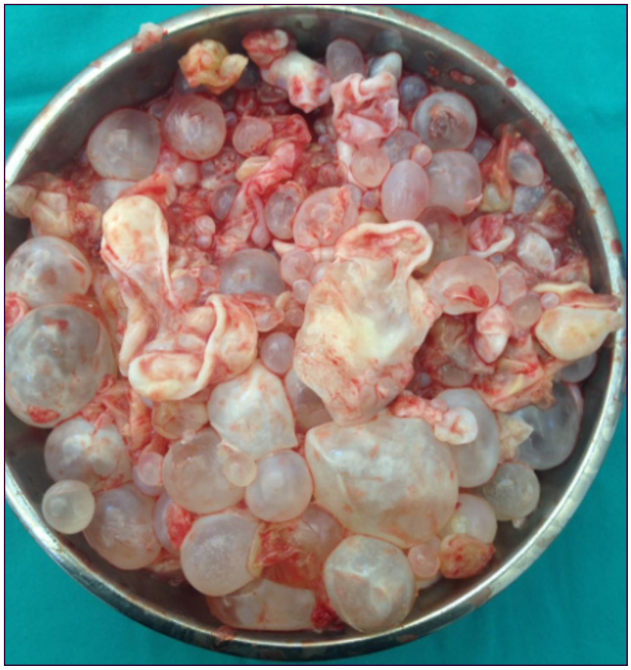

**Figure 2.** Cyst Hydatic constitutive parts

In our study, all patients were started on 10 mg/kg albendezol treatment for 3 weeks before the intervention. Medical treatment was continued in cases where the size of the cyst decreased on ultrasound, its type changed early in the classification, and in patients who were not considered to be at risk of perforation. We think that the decision whether to continue medical treatment should be made by checking the cyst with ultrasound before the procedure and depending on the behavior of the cyst. It is known that newly formed and active cysts with thin walls, smaller than 5 cm, respond better to medical treatment. It is active but smaller than 5 cm and does not show exophytic extension, ii) it is predicted that the possibility of rupture with trauma is low, iii) it does not have symptoms of compression on major vascular structures and/or large bile ducts, iiii) during follow-up with medical treatment (1-3 months). No surgical intervention or drainage was considered for patients with type change (in controls) and medical treatment was continued.

In conclusion, the Gharbi classification updated by the WHO is effective and reliable in determining the treatment strategy for hepatic hydatid cyst disease. In patients who are receiving chemotherapy and in whom intervention is decided, a final ultrasound performed just before the procedure may lead to a change in treatment management and more patients may benefit from medical treatment. We also recommend that family screening should be routinely performed by family physicians with chest radiography and ultrasonography, especially considering the high incidence in endemic areas.

## ETHICAL DECLARATIONS

**Ethics Committee Approval**: This study was approved by the Van Training and Research Hospital Ethics Committee (Date: 03.10.2019, Decision No:2019/18).

**Informed Consent:** Because the study was designed retrospectively, no written informed consent form was obtained from patients.

**Referee Evaluation Process:** Externally peer-reviewed.

**Conflict of Interest Statement:** The authors have no conflicts of interest to declare.

**Financial Disclosure:** The authors declared that this study has received no financial support.

**Author Contributions:** All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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