

# Cystic echinococcosis in Central Saudi Arabia: A 5-year experience

Sudi Arabistan'da kistik ekinokokosis: 5 yıllık deneyim

Fraz FAHIM, Saleh Mohammad AL SALAMAH

Department of Surgery, King Saud University, College of Medicine, University Unit, RMC, Riyadh, Kingdom of Saudi Arabia

**Background/aims:** In this series of patients we aim to describe aspects of presentation, diagnosis and management of patients suffering from Echinococcosis in an endemic region. **Methods:** This is an observational study done at Riyadh Medical Complex, Saudi Arabia from 1999 to 2004. All adult patients admitted with the primary or incidental diagnosis of echinococcosis were included. These patients were followed up for 6 months. Data relating to patients' demographic characteristics, mode and duration of presentation, investigations, complications and treatment offered was collected. This data was then analyzed using SPSS 11.0. **Result:** 117 patients with a mean age of 40.9±20.7 years were admitted, male to female ratio being 1.7:1. 114 (97.4%) originated from Middle East which is an endemic area. Pain right upper quadrant (RUQ), followed by cough were the commonest symptoms. Nine patients presented with jaundice ± choleangitis: 3 patients had intrabiliary rupture of the hydatid cyst, while 6 had extrinsic compression. Six patients had infected cyst. Fourteen patients had intrabronchial rupture diagnosed on bronchoscopy. Twenty five (21.4%) patients presented with recurrent disease. The Haemagglutination Inhibition test gave a sensitivity of 78.6. All cysts were visualized using USG, CXR and CT scan. Endocystectomy was the most frequent procedure. Post op 8 patients had biliary leakage and 3 had bronchopleural fistula a majority of whom settled conservatively while two required ERCP and one patient with bronchopleural fistula required surgery. Fifteen patients had infection related complications. During our follow up period no recurrences were recorded. All patients undergoing surgery also received medical treatment. Twenty five patients (21.4%) were unable to undergo operative treatment due to multiple reasons. They were medically treated. **Conclusion:** Cystic Echinococcosis is a disease of the middle aged. Ultrasonogram combined with a serological assay is the best diagnostic tool available for abdominal echinococcosis allowing diagnosis and staging, while chest X ray is the best screen for the pulmonary disease. The recommended treatment is endocystectomy with antihelminthic therapy. But the problem of early detection of echinococcosis in endemic areas needs attention by workers as this approach can potentially prevent the devastating complications due to this disease.

**Key words:** Hydatid cyst, echinococcosis, Echinococcus granulosus, hydatidosis

**Amaç:** Bu hasta serisinde, ekinokokozisin endemik olduğu bir bölgede karşılaşılan hastaların bulguları, tanıları ve tedavileri hakkında bilgi verilmesi amaçlanmıştır. **Yöntem:** Riyad Tıp Merkezi'nde, 1999 – 2004 yılları arasında gerçekleştirilmiş gözlemsel bir çalışmadır. Primer veya insidental olarak ekinokokoz tanısı alan tüm erişkin hastalar çalışmaya dahil edilmişlerdir. Bu hastalar 6 ay süreyle takip edilmişlerdir. Çalışma parametreleri, hastaların demografik özelliklerini, şikâyetlerini, şikâyet sürelerini, yapılan incelemeleri, komplikasyonları ve önerilen tedavi yöntemlerini kapsamaktadır. Bu veriler SPSS 11.0 programı ile değerlendirilmiştir. **Bulgular:** Çalışmaya, 117 (ortalama yaş: 40,9±20,7; erkek:kadın oranı 1,7:1) hasta dahil edildi. 114 hasta (%97,4) bu hastalığın endemik olduğu Ortadoğu kökenliydi. Sırasıyla, sağ üst kadran (SÜK) ağrısı ve öksürük en sık karşılaşılan semptomlardı. 9 hasta sarılık ± kolanjit tablosunda başvurdu: 3 hastada kistin intrabilier boşalması, 6 hastada bilier sisteme dışarıdan bası söz konusuydu. 6 hastada enfekte kist tespit edildi. 14 hastada kistin intrabronşial açılması bronkoskopi ile tespit edildi. 25 (%21,4) hasta tek-rarlayan hastalık nedeniyle başvurdu. Hemagglütinasyon inhibisyon testinin duyarlılığı %78,6 bulundu. Kistlerin görüntülenmesinde direk grafi, USG veya tomografi kullanıldı. Hastaların tedavisinde en sık endosistektomi kullanıldı. Operasyon ertesinde, 8 hastada bilier kaçak, 3 hastada bronkoplevral fistül gelişti. Hastaların çoğu konservatif olarak tedavi edilirken 2 hastanın ERCP'ye, bronkoplevral fistüllü bir hastanın cerrahi girişime ihtiyacı oldu. 15 hastada enfeksiyonlara bağlı komplikasyon görüldü. Takip dönemimizde rekürens ile hiç karşılaşılmadı. Cerrahi tedavi uygulanan tüm hastalara medikal tedavi de verildi ancak 25 (%21,4) hastaya çeşitli kontraendikasyonlar nedeniyle cerrahi tedavi verilemedi ve sadece medikal tedavi uygulandı. **Sonuç:** Kistik ekinokokozis orta yaş döneminin hastalığıdır. Serolojik testler ile kombine edilen USG abdominal hastalığın tanısının konulmasında en iyi yöntemdir. Buna karşılık torakal hastalığın taramasında en uygun yöntem direk grafidir. Önerilen tedavi endosistektomi ile beraber antihelminetik tedavidir. Endemik bölgelerdeki tanı konulmasındaki gecikmelerin önlenmesi ve ağır komplikasyonların önüne geçilmesi için sağlık çalışanlarının konuya daha iyi eğilmeleri gerekmektedir.

**Anahtar kelimeler:** Hidatid kist, ekinokokozis, ekinokokus granulozus, hidatidoz

**Address for correspondence:** Saleh Mohammad AL SALAMAH  
Department of Surgery, College of Medicine, King Saud University,  
University Unit, RMC, Riyadh, PO Box 261283, Riyadh 11342,  
Kingdom of Saudi Arabia  
Phone: +00966-1-4671585 • Fax: +00966-1-4679394  
E-mail: smsalamah@hotmail.com

**Manuscript received:** 03.06.2006 **Accepted:** 01.02.2007

## INTRODUCTION

Cystic Echinococcosis (CE) has been recognized as the most important and widespread helminth zoonosis caused by tapeworm *Echinococcus granulosus* (1). It is endemic in the Middle East and areas around the Mediterranean (2, 3). But with increasing foreign travel and immigration, practitioners all over the world need to be aware of this condition (4). The infestation has a significant economic impact due to loss of work and healthcare costs, resulting in heavy financial burden; especially in the developing countries (1). The cattle and sheep rearing regions with inadequately dewormed canine population leads to a higher prevalence of infestation (1). The growth of the cyst is often insidious and becomes symptomatic only late in the course of the disease, and precise knowledge of natural history of cystic echinococcosis in humans is somewhat arcane (5, 6). Liver and lungs are the first and second most frequently involved organs respectively but occasionally CE can be found in unusual sites (7-9). The CE can present with complications such as compression effects, rupture into adjacent structures and secondary infections, in addition to the presentation as uncomplicated cysts (7, 10). In this series we aim to describe aspects of presentation, diagnosis and management of patients suffering from Echinococcosis in an endemic region.

## MATERIAL AND METHODS

This is an observational study carried out at Riyadh Medical Complex, Saudi Arabia from December 1999 to December 2004, spanning a period of five years. All cases admitted with the primary diagnosis or incidental discovery of echinococcosis were included. The cases comprise both elective and emergency admissions. Pediatric patients were excluded since they are dealt in the Department of Pediatric Surgery. These patients were followed up in the OPD for at least 6 months to look for short term complications.

Data relating to patients' demographic characteristics, mode and duration of presentation, investigations, complications and treatment offered was collected. This data was then analyzed using SPSS 11.0.

## RESULTS

A total of 117 patients were admitted with the diagnosis of Echinococcosis during the five year

study period. The mean age was  $40.9 \pm 20.7$  years ranging from 13 years to 95 years. Seventy four (63.24%) were males giving a male to female ratio of 1.7:1. Among the patients 114 (97.4%) originated from Middle East or had been living in the region for more than 10 years.

The mean duration of complaints was  $14.1 \pm 17.4$  months. Their commonest presenting complaint was pain right upper quadrant (RUQ). (Table 1). Pain in the RUQ and upper abdomen with or without jaundice usually correlates with hepatic or abdominal echinococcosis. And cough with or without sputum points towards a pulmonary disease. Most of these patients had hepatic Echinococcosis, but 4 of them also had extra hepatic disease. The second most common presentation was dry or productive cough followed by non localized upper abdominal pain.

**Table 1.** Commonest presenting complaints

	Symptom	N	%-age
1	Pain RUQ	51	43.6
2	Cough $\pm$ Sputum	22	18.8
3	Pain Upper Abdomen	21	17.9
4	Incidental	10	8.5
5	Jaundice	9	7.7
6	Fever	3	2.6
7	Lump L Thigh	1	0.9

Twenty nine (24.8%) patients had Echinococcosis related complications. Amongst the patients with hepatic Echinococcosis infection (fever/leucocytosis) was present in 6 (5.1%), while 9 (7.7%) had jaundice. Six of these 9 patients had compression of major biliary ducts while 3 had intrabiliary rupture of hydatid cyst, diagnosed and cleared endoscopically. And 14 out of 22 patients (63.6%) with pulmonary Echinococcosis had intrabronchial rupture as diagnosed on bronchoscopy.

Twenty five (21.4%) patients presented with recurrent Hydatid Cyst. Regarding the site of previous Echinococcosis 14 patients had hepatic disease, 4 had pulmonary disease, 2 hepatopulmonary disease while 1 had cyst in the thigh. Four patients who had disseminated disease had received medical treatment successfully.

The initial hematological profile revealed a mean hemoglobin level of  $12.7 \pm 17.4$  G/dl and a mean WBC count of  $9.3 \pm 4.9$  /cc. Eosinophilia, defined as

more than 6% eosinophils on differential count (11) was present in 53%, with a mean of  $11.3\% \pm 13.1$  (Range 0-51%). The Haemagglutination Inhibition test was the serological assay used. It was positive in 92 cases giving a sensitivity of 78.6. USG was the main imaging modality used for all cases of extrapulmonary Echinococcosis coupled with contrast enhanced CT scan in selected cases. For pulmonary disease CXR was the initial investigation for imaging. All the patients had active echinococcal cysts and according to WHO-IWGE classification 34 (29.1%) were Type CE 1, 61 (52.1%) were Type CE 2 and 22 (18.8 %) were categorized as Type CE 3.

In a majority of cases i.e. 103 (88%) Echinococcosis was present in one organ while 14 (12%) patients had two involved organs. Liver was the primary site in 93 (79.5%) patients out of whom 11 also had other organs involved; while 24 (20.5%) patients had primary pulmonary hydatid cyst amongst whom 3 had asymptomatic hepatic cysts as well. Surgery was the primary treatment offered. (Table 2). The most frequent procedure was endocystectomy in both the liver and lung.

**Table 2.** Operative procedures

Procedure	N	%
<b>Liver</b>		
Endocystectomy	54	58.7
Pericystectomy	7	7.6
Lap Endocystectomy	5	5.4
Liver & Extrahepatic abdominal	4	4.3
Liver & Thigh Compartmentectomy	1	1.1
<b>Lungs</b>		
Endocystectomy	17	18.5
<b>Liver &amp; Lung</b>		
Endocystectomy	4	4.3
	<b>92</b>	<b>100</b>

Major post operative complications occurred in 11 patients. Eight patients had biliary leakage. Six settled on conservative treatment while two required papillotomy and stenting of CBD. Post op bronchopleural fistula was seen in 3 patients amongst whom one had to be re-operated. Thirteen (14.1%) patients developed superficial wound infection.

All patients undergoing surgery also received medical treatment. Thirty two (27.4%) patients rece-

ived 4 weeks of pre op as well as post op treatment while 60 (51.3%) patients received only post operative treatment mainly due to logistic reasons, and 22 (18.8%) patients were treated medically only. The most frequently used drug was Albendazole in 78 (68.4%) at a dose of 15 mg/kg in two divided doses. Mebendazole at a dose of 50 mg/kg in three divided doses was used in the rest of the patients. Both of these drugs were used for 3 months course. Both these drugs have been found efficacious, Albendazole more than Mebendazole (12). However, Mebendazole was used most commonly in earlier patients in our series. These drugs are known to cause hepatic derangements, hence if there was a rise in transaminases, the drug was stopped and resumed only when they returned to normal. Praziquantel at 40 mg/kg weekly was used in 34 (29.8%) patients; always in combination with either of the two drugs mainly because of its potential of preventing relapse (13).

The remaining 25 (21.4%) were unable to undergo operative treatment due to multiple reasons. Ten patients had severe co morbidities due to which 6 of them expired during the same admission due to unrelated causes. Extensive irresectable disease was present in 7 patients. Eight patients refused to give consent for operation. All these patients were medically treated.

The mean hospital stay was 14.4 days (2-187 days). Patients receiving surgical treatment were discharged earlier with a mean stay of 11.9 days, while those receiving medical treatment alone were generally sicker and tended to stay longer with a mean of 23.6 days. We were able to complete follow up of 89 (96.7%) of our operated cases up to 6 months and during that time no recurrences were recorded.

## DISCUSSION

With the increase in foreign travel and migration of people, doctors need to be watchful for diseases common in regions other than their countries. This is especially true of the disease such as cystic echinococcosis as it is endemic in certain areas and chronic in nature, with possible devastating acute complications (4, 14).

The most common sites of lodgment of *Echinococcus granulosus* are liver and lung (15). Growth rate of the cyst depends on the host tissues. The compact structure of the tissues and the patient's immune response influence the size of the cyst and

resulting symptoms (15). This leads to a relatively slow course and delayed presentation making it a disease of middle aged people. Our patients presented in the fourth and fifth decades of life with a mean duration of symptoms of more than one year. Similar results have been reported from other endemic areas (1, 6). The male (63%) to female (37%) ratio in our series is comparable to the 60% males and 40% females in the Jordanian series and also to earlier reports from Saudi Arabia (3, 16).

The commonest abdominal symptom was pain in the upper quadrants, while dry or productive cough was the commonest presentation in pulmonary hydatosis (6). Both these symptom groups are non specific and may be initially ignored by many primary care physicians, which may delay the diagnosis leading to possible complications. Hence these apparently minor symptoms persisting despite treatment need to be fully investigated, and more so in endemic areas. The symptoms generally correlate with the system involved, but occasionally may present with unrelated symptoms or may be diagnosed incidentally when investigating for another disease.

The ideal tool for diagnosis and screening is still debatable. Eosinophilia is of limited value in this condition. In our series only about half of the patients had significant eosinophilia reflecting the inadequacy of this tool. Serological assays have been used in both hospital setting and in-field detection of cases (17). The Moroccan study revealed that 77.4% of ultrasonographically detected cases had a positive serology, which is very close to our figure of 78.6% (6). But the previously reported Saudi study has quoted a figure of 57.7% which is somewhat lower (17). However ultrasonogram has been found to be generally superior for diagnosis of hepatic echinococcosis, allowing diagnosis of the infestation before the immune response is detectable serologically (18, 19). Additionally, using WHO-IWGE classification, ultrasound can detect with confidence the cysts which are or soon may become inactive (20). However, the extra-abdominal manifestations may not be identified using this modality alone. Hence, ultrasonogram needs to be combined with serology to maximize the possibility of reaching the diagnosis (19).

In the hepatic hydatid cyst complications related to the biliary system were most commonly seen (7.7%) in our series. They included biliary compression as well as intrabiliary rupture, diagnosed on ERCP. Other workers have shown similar

trend (21). Infected hydatid cyst (5.1%) was the second most common complication. But unlike the pyogenic liver abscess infected hydatid cyst follows a less dramatic clinical course and seems to be related to the pericystic structure (sometimes calcified), probably representing a non-specific defense mechanism to infection and its dissemination (22). Additionally, the possibility that most clinical features and derangement of liver function tests result from acute cholangitis secondary to cyst structure migration to the biliary tree has been suggested (21). In the present series we were successful in controlling the infection with antibiotic therapy and the patients were then operated as for routine hydatid cyst.

The group with pulmonary Echinococcosis demonstrated a high complication rate (63.6%) in our series, giving the probable explanation that a large majority with minimal symptoms may not be reporting to the hospital, presenting only when complicated by either compression of bronchi or intrabronchial rupture (20). Diagnosis is made by chest X ray and bronchoscopy is usually used in a diagnostic as well as a therapeutic role for clearance of the obstructed bronchial passages (15). The inordinately high rate of complicated pulmonary echinococcosis indicates that there is a pressing need to find means to detect pulmonary disease early.

Our preferred surgical approach was endocystectomy for the hydatid cyst. This is a safe, simple, effective procedure and completely removes the active disease while saving the healthy tissue (23). We were able to achieve relatively good results with endocystectomy since consequent to the high volume of patients the surgeons have been able to develop technical skill in this particular area. The alternative procedure pericystectomy, involves the extirpation of surrounding healthy liver parenchyma. However it carries a significant morbidity and should not be routinely attempted by general surgeons away from the specialized hepatobiliary centers, thus limiting its usefulness in endemic areas (22). We were not able to compare the efficacy of the two surgical techniques with respect to relapse as none was documented in our follow up period. However the long term results when available may be able to shed some light on the matter.

Currently surgery is still the treatment that has the potential to remove *E. granulosus* cysts and lead to complete cure. And it can be performed in upto 90% of the patients successfully (24). Howe-

ver, it may be impractical for the patients in whom the cysts have risky localization, may be located at multiple sites or different organs, the patient may be high risk or the surgical facilities may be inadequate (25). A combination of chemotherapy and PAIR (Puncture-Aspiration-Injection-Reaspiration) technique is being used where surgery may be inappropriate (26).

Interestingly, nearly one fifth of our patients presented with recurrent disease. Perhaps the reason could be inadequate adjuvant therapy offered to these patients. In the present series we used the recommended antihelminthic combination therapy pre and post-operatively combined with meticulous surgical technique to avoid spillage cyst contents. We saw no recurrence in our limited follow up (0%). This however can be considered a pro-

visional figure since a longer follow up may reveal some relapses and also since we were able to follow only 96.7% of our patients.

In conclusion we can surmise that Cystic Echinococcosis is a disease manifesting in the middle aged with a slight male predominance. At present ultrasound combined with a serological assay is the best diagnostic tool available for abdominal echinococcosis allowing both diagnosis and staging, while chest X ray is the best screen for the pulmonary disease. Endocystectomy combined with antihelminthic therapy is the recommended treatment. However the problem of early detection of echinococcosis in endemic areas needs attention by workers as this approach can potentially prevent the distressing complications due to this disease.

## REFERENCES

- Cetinkaya Z, Ciftci IH, Demirel R, et al. A sero-epidemiological study on cystic echinococcosis in midwestern region of Turkey. *Saudi Med J* 2005; 26: 350-1.
- Tor M, Atasalihi A, Altuntas N, et al. Review of cases with cystic hydatid lung disease in a tertiary referral hospital located in an endemic region: a 10 years' experience. *Respiration*. 2000; 67: 539-4
- El Marsfy YS, Morsy TA. A preliminary study on echinococcosis in Riyadh, Saudi Arabia. *J Pak Med Assoc* 1975; 25: 10-1.
- Cooney RM, Flanagan KP, Zehyle E. Review of surgical management of cystic hydatid disease in a resource limited setting: Turkana, Kenya. *Eur J Gastroenterol Hepatol*. 2004; 16: 1233-6.
- Cobb PJ, Schmeig R, Hunt TK, Mundy LM. Inflammation, infection and antibiotics. In: Way LW, Doherty GM, eds.: *Current surgical diagnosis and treatment*. 11<sup>th</sup> edition, McGraw-Hill Co., 2003; 129.
- MacPherson CN, Kachani M, Lyagoubi M, et al. Cystic echinococcosis in the Berber of the Mid Atlas mountains, Morocco: new insights into the natural history of the disease in humans. *Ann Trop Med Parasitol*. 2004; 98: 481-90.
- Sayek I, Tirnaksiz MB, Dogan R. Cystic Hydatid disease: current trends in diagnosis and management *Surg Today*. 2004; 34: 987-96
- Tiseo D, Borrelli F, Gentile I, et al, Cystic echinococcosis in humans: our clinic experience *Borgia G Parassitologia*. 2004; 46: 45-51.
- Safioleas M, Misiakos EP, Dosios T, et al. Surgical treatment for lung hydatid disease. *World J Surg*. 1999; 23: 1181-5.
- Chautems R, Buhler LH, Gold B, et al. Surgical management and long-term outcome of complicated liver hydatid cysts caused by *Echinococcus granulosus*. *Surgery*. 2005; 137: 312-6.
- Bain B. J. Eosinophilia — idiopathic or not? *N Engl J Med* 1999; 341: 1141-3.
- Smego RA Jr, Bhatti S, Khaliq AA, Beg MA. Percutaneous aspiration-injection-reaspiration drainage plus albendazole or mebendazole for hepatic cystic echinococcosis: a meta-analysis. *Clin Infect Dis* 2003; 37: 1073-83.
- Dautovic-Krkic S, Huskic J, Cengic D, et al. Praziquantel in the prevention of recurrence of human echinococcosis. *Med Arh* 2002; 56: 263-6.
- Laajam MA, Nouh MS. Hydatidosis: clinical significance and morbidity patterns in Saudi Arabia. *East Afr Med J* 1991; 68: 57-63
- Halezeroglu S, Celik M, Uysal A, et al. Giant hydatid cyst of the lung. *J Thoracic Cardiovascular Surg* 1997; 113: 712-17.
- Amr SS, Amr ZZ, Jitawi S, Annab H. Hydatidosis in Jordan: An epidemiological study of 306 cases. *Ann Trop Med Parasitol* 1994; 88: 623-27.
- Hossain A, Bolbol AS, Chowdhury MN. Serodiagnosis of human hydatid disease in Riyadh, Saudi Arabia. *Ann Trop Med Parasitol* 1985; 79: 439-42.
- Shambesh MA, Craig PS, Macpherson CNL, et al. An extensive ultrasound and serologic study to investigate the prevalence of human cystic echinococcosis in northern Libya. *Am J Trop Medicine Hyg* 1999; 60: 462-8.
- Ozkol M, Kilimcioglu AA, Girginkardesler N, et al. A discrepancy between cystic echinococcosis confirmed by ultrasound and seropositivity in Turkish children. *Acta Tropica*. 2005; 93: 213-16.
- Brunetti E, Filice C, MacPherson C, et al. PAIR: Puncture, Aspiration, Injection and Re-aspiration- an option for the treatment of Cystic Echinococcosis. *World Health Organization, Department of Communicable Diseases, Surveillance and Response*. 2001: WHO/CDR/CSR/APH/2001.6
- Paksoy M, Karahasanoglu T, Carkman S. Rupture of the hydatid disease of the liver into biliary tracts. *Dig Surg* 1998; 15: 25-9.
- Manterola C, Barroso M, Vial M, Bustos L, Munoz S, Losada H et al. Liver abscess of hydatid origin: Clinical features and results of aggressive treatment. *ANZ J Surg* 2002; 72: 220-4.
- Buttenschoen K, Schorcht P, Reuter S, et al. Surgical treatment of hepatic infections with *Echinococcus granulosus* *Z Gastroenterol* 2004; 42: 1101-8.
- Eckert J, Gemmell MA, Meslin FX, Pawlowski ZS. WHO/OIE manual Echinococcosis in humans and animals: A public health problem of global concern. *World Health Organization*; 2002.



25. Guidelines for treatment of cystic and alveolar echinococcosis. WHO Informal Working Group on Echinococcosis. Bull WHO 1996; 74: 231-42.
26. Üstünsöz B, Akhan O, Kamiloğlu MA, et al. Percutaneous treatment of hydatid cysts of the liver: long-term results. AJR Am J Roentgenol 1999; 172: 91-6.