

## COMPREHENSIVE STUDY OF HYDATID DISEASE OF LIVER AND MANAGEMENT AT PERIPHERAL TERTIARY CARE HOSPITAL

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### ABSTRACT

#### BACKGROUND

Hydatid Disease (HD) is a zoonotic disease caused by the larvae of Echinococcus granulosus, now become a rare clinical entity in teaching medical college hospitals. This is due to the public education about the disease, mode of spread and treatment available. The aim of the study is to review the epidemiology, clinical presentation, diagnostic methods available, organs affected and treatment available in the tertiary hospitals and outcome.

#### MATERIALS AND METHODS

Data Collection- Patients with upper right abdominal pain with radiological and ultrasound findings are included in the study. About twenty patients are taken for this study. Medical managements, surgical procedure done and outcomes are recorded, tabulated and analysed. Research Design- Prospective Study, Research Setting- KAPV Govt. Medical College and Mahatma Gandhi Memorial Govt. Hospital, Trichy, Tamilnadu. Duration- 7 yrs. (2009 to 2016) Sample Size- Twenty. Inclusion Criteria- Patients between 12 to 70 years of age of both sexes. Patient having right upper abdomen or epigastric pain with positive radiological and ultrasound findings. Patient willing to participate in the study. Exclusion Criteria- Patients more than 70 years not willing to participate in the study patients absconded in between the management.

#### RESULTS

Liver is a commonest solid organ affected by the hydatid disease. Most of the diagnosis are made accidentally when the patients are investigated for some other diseases. The commonest clinical presentation is right abdominal or epigastric pain with hepatomegaly. The average age group is 45 years. X-ray abdomen, ultrasound abdomen are the most useful investigations. Asymptomatic uncomplicated small cyst less than 5 cms are managed with medical treatment. Symptomatic large cysts are submitted for surgical intervention.

#### CONCLUSION

The main source of income in majority of rural population is agriculture and sheep and cattle grazing. WHO is working towards the validation of effective cystic echinococcosis control strategies by 2020. At this juncture, we have to thank the Public Health Department and Veterinary Department for their effect in educating the public about the disease, especially about washing hands, mode of spread and treatment available in all peripheral hospitals and regularly deworming their dogs.

#### KEYWORDS

Echinococcosis Granulosus, Epigastrium, Hepatomegaly, X-ray, Ultrasound, Surgery.

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#### BACKGROUND

The word Echinococcosis is Greek in origin and means Hedgehog Berry. It was first described in Talmud bladders full of water. Hippocrates described it as liver is filled with water.<sup>1</sup> Hydatid disease is caused by a parasite that belongs

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to phylum platyhelminthes, class cestoda, tape worm, E.granulosus.<sup>2</sup> There are 4 forms of echinococcosis.<sup>1</sup>

1. Cystic echinococcosis also known as hydatid disease or hydatidosis caused by infection with E. granulosus.
2. Alveolar echinococcosis caused by infection with E.multilocularis.
3. Polycystic echinococcosis caused by infection with E.vogeli.
4. Unicystic echinococcosis caused by infection with infection with E. oligarthus.

Two most important forms, which are of medical and public health relevance in humans are cystic and alveolar echinococcosis. Cystic echinococcosis is principally

maintained in a dog-sheep-dog cycle. Yet several other domestic animals may be involved including goats, swine, horse, cattle, camel and yaks. Alveolar echinococcosis usually occurs in wildlife cycle between foxes, other carnivores and small rodents (mostly rodents). Hydatid disease is endemic mainly in Mediterranean countries (particularly Greece), the Middle East, the Baltic areas, South America, India, Northern China and other sheep raising areas. However, owing to increased travel and tourism all over the world it can be found anywhere even in developed countries.<sup>3</sup> In India hydatid disease is common in most of the states, of which Andhra Pradesh and Tamil Nadu predominate.<sup>4</sup> Infection with *E. granulosus* is a commonest cause for liver cyst in the world.<sup>5,1,6</sup> 52% - 70% occurs in the right lobe of the liver; 25% in the lungs, 5% - 10% distributed along the arterial system.<sup>7</sup> Human beings become accidental intermediate hosts through contact with definitely host dog or by ingestion of contaminated water and vegetables, but there is no human-to-human transmission liver.<sup>6</sup> The first filter and thus most frequently involved. The right lobe is involved in 85% patients.<sup>8,9</sup> The small (5 mm long) adult *E. granulosus* complex worms, which live for 5-20 months in the jejunum of dogs have only 3 proglottids: one immature, one mature and one gravid. The gravid segment splits to release eggs that are morphologically similar to *Taenia* eggs and are extremely hard. After humans ingest the eggs, embryos escape from the eggs, penetrate the intestinal mucosa, enter the portal circulation and are carried to various organs, most commonly liver and lungs. Larvae develop into fluid-filled unilocular hydatid cysts that consist of an external membrane and an inner germinal layer. Daughter cysts develop from the inner aspect of the germinal layer, as do germinating cystic structures called brood capsules. New larvae called protoscolices, develop in large numbers within the brood capsule. The cysts expand slowly over a period of years. The larval form of *E. multilocularis* however is quite different, in that it remains in the proliferative phase, the parasite is always multilocular and vesicles without brood capsule or protoscolices progressively invade the host tissue by peripheral extension of processes from the germinal layer.

The clinical features depend upon the sites, size, stage of development of the cyst whether alive or dead or infected and calcified.<sup>10</sup> There are studies to show that depending upon the pressure in the cyst, we can say the nature of the cyst. When the pressure is 0 cm of H<sub>2</sub>O, the cyst is dead one. When the pressure is 35 cms of water, the cyst is viable. When the pressure increases more than 70 cms of water, it is more prone for spontaneous traumatic rupture into the peritoneal cavity.<sup>11,12</sup> Pain in the right upper abdomen or epigastrium is the commonest symptom. The classical physical sign of hydatid thrill is rarely elicitable.

Sl. No.	Symptoms	Percentage of Hydatid Cyst
1	Asymptomatic	75
2	Abdominal pain	20

3	Dyspepsia	13
4	Fever and Chills	8
5	Jaundice	6
<b>Signs</b>		
1	Hepatomegaly	70
2	Right upper abdominal tenderness	20

**Table 1. Signs and Symptoms of Hydatid Disease of Liver<sup>13,6,14</sup>**

### Signs and Symptoms of Hydatid Disease of Liver<sup>1,10,15</sup>

The diagnosis is based on contact history, geography, imaging and serology. Parasitology of the cyst content confirms the diagnosis.<sup>1</sup> The sensitivity and specificity of the test depends upon the quality of the antigen. The well-known Casoni test has been largely abandoned because some cysts never leak and tests are never positive.<sup>16</sup> Intradermal Casoni test, human basophil degradation test and complement fixation test have only historical relevance.<sup>17</sup> Sbihi and Associates reported that purified fraction enhance the antigen 5 and B and glycol proteins from hydatid fluid yielded sensitivity of 95% and specificity of 100%.<sup>18</sup> ELISA is used for epidemiological study.<sup>19</sup> Immunoelectrophoresis is used for post-treatment followup. RAST (Radioallergosorbent Test) is most diagnostic of active disease. Normally, hydatid cyst will not alter the liver function. When the cyst is large more than 10 cms in diameter may alter the liver function test. Increase bilirubin and increased alkaline phosphatase indicates cystobiliary fistula. X-ray abdomen and ultrasound abdomen are the cost wise low investigation and easily available at any peripheral hospital. Advance investigation like CT, MRI, ERCP and MRCP will help to identify the early complication like cystobiliary fistula. When the segment seven and eight are involved the incidence of cystobiliary fistula is most common.<sup>20,21</sup> Mostly, the minor biliary fistula are asymptomatic. When patient presented with obstructive jaundice and cholangitis think of major biliary fistula. There are studies to show that when the diameter of the cyst is more than 10 cms, we have to think of intrabiliary fistula.

Cyst Type	Status	Ultrasound Features	Remarks
CL	Active	Not Pathognomonic Unilocular No Cyst Wall	Early Stage Not Fertile Needs D/D
CE I	Active	Cyst wall Hydatid Sand	Fertile
CE 2	Active	Multi-locular cyst Characteristic appearance with daughter cyst as honey comb Rosette image	Fertile
CE 3	Transitional	Detached laminated membrane Water-Lily sign	Starting to degenerate rarely produce daughter cyst
CE 4	Inactive	Hyperechogenic degenerative content No daughter cyst	No living protoscolices

			D/D is necessary
CE 5	Inactive	Thick calcified wall Partial or Complete Not pathognomonic Suggestive of diagnosis	No living protoscolices
<b>Table 2. WHO Classification of Hydatid Cyst</b>			

**WHO Classification of Hydatid Cyst<sup>22</sup>**

Uncomplicated cyst which contains colourless clear fluid with decreased amount of potassium and calcium and bacteriologically sterile size, less than 5 cms are treated medically with tablet albendazole for three months and periodically assessed with ultrasound abdomen. Only albendazole is ovicidal, larvicidal and vermicial. Medical management is the commonest cause for recurrence of the disease in endemic area.<sup>23</sup> Symptomatic cysts are managed surgically<sup>14</sup> either by radical method<sup>24</sup> or conservative method. The conservative methods are PAIR (Puncture Aspiration Instillation of Scolicidal agents and Reaspiration), PEVAC (Percutaneous Evacuation of the Cyst Content),<sup>25,26,27</sup> Simple evacuation of the cyst and inactivation of the protoscolices using scolicidal agents.<sup>28,29</sup> PAIR and PEVAC needs qualified hepatologist to handle the complications following these procedures. Whereas in every general hospitals, a general surgeon can do the simple evacuation of the cyst. Now in the era of minimally invasive procedure you can also do the same procedure<sup>1,30,27</sup> laparoscopically. Scolicides preferably cetrimide 40% concentrate (Cetavlon), which is safer than other scolicides which includes 1% formalin in 0.9% saline, 10% sodium chloride, 70% alcohol, 0.5% silver nitrate or hypochlorite; 10% formalin is more dangerous. During surgical management of cyst, if cyst is high in right lobe under the diaphragm it may need a thoraco-abdominal incision and extrapleural dissection and if the diaphragm is incised it may be repairs or referred to higher centre. For cysts involving the left lobe and more in right lobe make a median, paramedian or rooftop incision. Another way of getting better access is to stuff packs soaked with scolicidal agents into right subphrenic space, so as to push liver down. One can also release the ligaments of liver to have a good access for the procedure.

There are studies showing that the recurrent of the disease following PAIR,<sup>31</sup> the newer method Radiofrequency thermal ablation is under study.<sup>32</sup> The complicated cysts are difficult to manage in the peripheral hospitals, where normally there is no specialised departments. When the patient presented with fever and chills, we have to think in terms of secondary infection of the cyst which may turn into hydatid abscess. Under antibiotic cover, we can aspirate the content with ultrasound guidance. Normally, in the abscess the daughter cysts are dead. Other complications of the hydatid cyst are by direct pressure upon the adjacent anatomical organ, rupture into the physiological channels namely Biliary tract or Bronchial tract; Body cavities like Peritoneal cavity, Pleural cavity or Pericardial cavity.<sup>11</sup>

Among the ruptures, 5% - 10% is rupture into the biliary tract is more common.<sup>28</sup> 1% - 8% into the peritoneal cavity.<sup>11</sup> There are reports to show rupture into the pericardium, IVC, Stomach.<sup>33</sup> There are studies to show that when the intracystic pressure goes more than 70 cms of water, the chances of spontaneous or traumatic rupture is

more. Classically, we can appreciate a rupture into the biliary tree by Camellotte sign where the gas enter into the cyst leaving a partial collapse of the cyst wall. Recent study shows lymphatic spread of the larvae of *E. multilocularis* to regional lymph node and suggest removal of lymph node during the procedure to reduce the risk of persistent infection.

**MATERIALS AND METHODS**

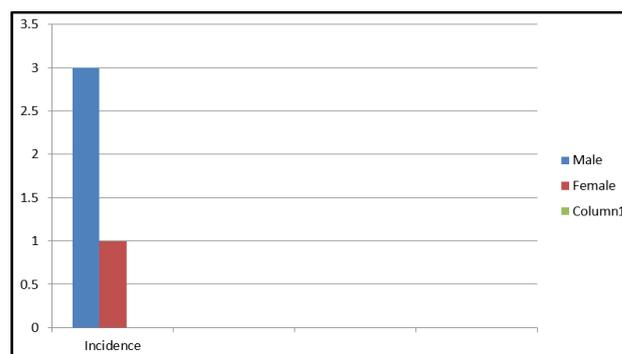
Twenty patients with hydatid disease are admitted in KAPV Govt. Medical College Hospital for the study. Patients between the age group of 12 to 70 of both sexes are included for this study. Though the cases are very limited the study includes clinical features, investigation methods, complication of cyst and surgical techniques available where studied and the outcomes are analysed.

**RESULTS AND OBSERVATION**

In our study, Eighty five percentage of the cases were diagnosed as hydatid disease of the liver. Asymptomatic, uncomplicated smaller cysts less than 5 cms are treated conservatively with medicine with periodical ultrasound abdomen examination. Approximately, forty percentage of the cases were taken up for surgery. We did conservative cyst evacuation followed by omentoplasty for the five patients and capitonnage for two patients because of inadequate omentum, so that it does not leave a sinus or fistula. In some centres, they use specialised cones to prevent spillage of cystic content into the peritoneal cavity, thereby to prevent secondary hydatidosis. We use povidone iodine soaked pads around the cyst to prevent spillage. In Fifteen percentage of cases where lung and kidney are affected, both the cases were diagnosed accidentally when the patients were investigated for some other purpose. Since there is no pericyst formation in the lung, we can see the water-lily sign in case of hydatid cyst of the lung. Among the operated cases one patient had eventful post-operative period, was identified and managed successfully. Other cases were discharged on twelfth postoperative period on advice for regular followup.

Sex	Number of Cases	Age Group				Percentage
		12-20	21-30	31-45	>46	
Male	15	1	0	13	1	75%
Female	5	0	1	4	0	25%

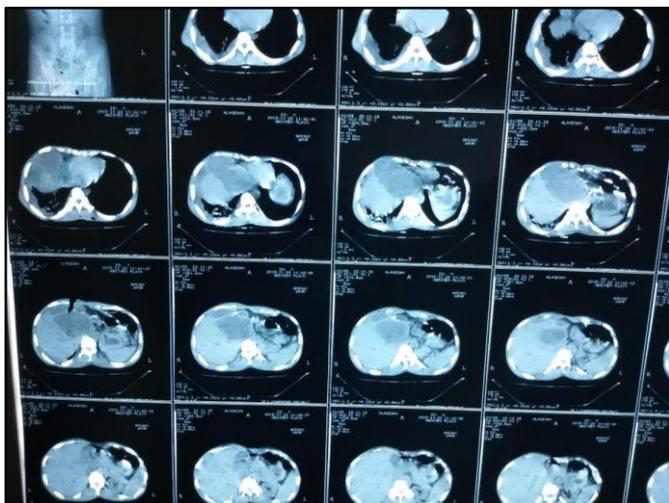
**Table 3. Age Distribution**



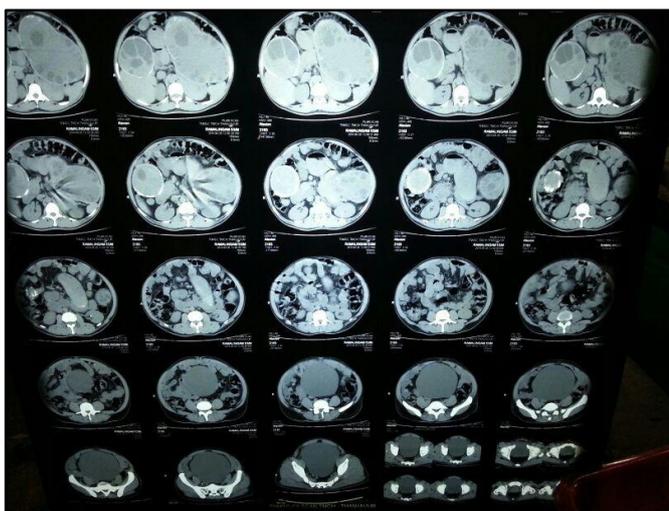
**Graph 1. Age Distribution**

The age of the patients ranges from 12 to 70 years. Male:Female ratio is 3:1. Eighty five percentage of the

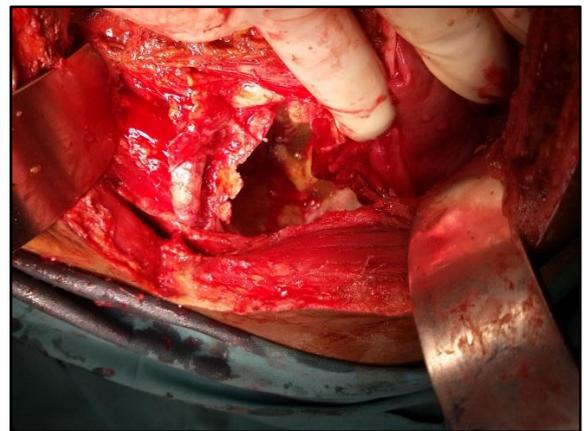
patients were between the age group of 31 to 45 years. There is a history of close association with the dog, was elicited among our patients. There are evidence to show that there is a definite relationship between dog and man in the epidemiology of the disease in the endemic areas. Symptoms and signs depends upon the size of the cyst, stage of development of the cyst, whether the cyst is alive, dead or infected. Pain in the right upper abdomen or epigastrium is the commonest symptom. When the patient is presented with fever and chills you have to think of hydatid abscess. When there are signs of peritonitis, think of rupture of the cyst into the peritoneal cavity. Hepatomegaly is the commonest sign. Ultrasound abdomen is a very useful diagnostic aid in the investigation of hydatid cyst.



**Figure 1. CT Image**



**Figure 2. MRI Image**



**Figure 3. Intra-op Image 1**



**Figure 4. Intra-op Image 2**



**Figure 5. Intra-op Image 3**



**Figure 6. Post-op Specimen- Hydatid Cysts**

**DISCUSSION**

Hydatid disease is caused by *E. granulosus*. Liver is the commonest solid organ affected by hydatid disease. The pericyst formation is absent in lung and brain. There is a close relationship between the dog and the man. Man is the accidental intermediate host. Most of the affected patients are asymptomatic. The cyst size increases averagely by 2 cms per year. In our study, males are most commonly affected between the age group of 30 to 45 years. Pain in the right upper quadrant or epigastrium is the commonest symptom in most of the patients. Hepatomegaly is the commonest sign. The age old clinical sign, hydatid thrill is rarely elicitable and Casoni test is rarely positive. Liver function test is usually normal. When there is increase in bilirubin and alkaline phosphatase, we think of some complications like cystobiliary fistula. RAST is the most diagnostic test to confirm the active disease. In our study, we are not able to use all the sophisticated study like immunology study. X-ray abdomen and ultrasound of the abdomen are the most useful diagnostic tool. Asymptomatic, uncomplicated small cysts approximately sixty percentage of the cases were treated medically with tablet albendazole with periodical ultrasound abdomen. Most of our patients are so irregular. And there are studies to show that the commonest cause for the recurrence is medical management. The symptomatic patients, approximately forty percentage were treated surgically, that is conservative open cyst evacuation. Even though our hospital is a teaching hospital there is no specialised department like Hepatology, so we are not able to perform the procedures like PAIR, PEVAC and Laparoscopic evacuation.

We have operated upon six patients. For four patients whom we did cyst evacuation with omentoplasty and for two patients we did cyst evacuation with capitonnage because of inadequate omentum. Only one patient had eventful post-operative period, which was identified early and treated successfully. All other patients are discharged on twelfth postoperative period and advised for regular followup.

**CONCLUSION**

The main source of income in majority of rural population is agriculture and sheep and cattle grazing. Moreover, there is huge population of stray dogs. The slaughtering of livestock without veterinary control. The widespread practice of feeding dogs with the viscera of home butchered sheep is a common practice. All these factors are highly favourable for transmission of echinococcosis. WHO is working towards the validation of effective cystic echinococcosis control strategies by 2020. At this juncture, we have to thank the Public Health Department and Veterinary Department for their effect in educating the public about the disease, especially about washing hands, mode of spread and treatment available in all peripheral hospitals and regularly deworming their dogs.

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