Hydatid disease in children: Our experience

Rahul Gupta a,*, Shyam Bihari Sharma b, Girish Prabhakar c, Praveen Mathur a

a Department of Pediatric Surgery, Sawai Man Singh Medical College, Jaipur, Rajasthan, India
b Department of Pediatric Surgery, National Institute of Medical Sciences, University Medical College, Shobha Nagar, Jaipur, Rajasthan, India
c Department of Pediatric Surgery, Sardar Patel Medical College, Bikaner, Rajasthan, India

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Summary Background/Introduction: Hydatid disease is relatively uncommon in children. The liver and lungs are most commonly involved, while involvement of other sites in the body is unusual.

Purpose(s)/Aim(s): To study the presentation of hydatid disease in children, evaluate the risk factors, and derive appropriate management recommendations for prevention of recurrence and disease control.

Methods: This was a retrospective study performed from April 2006 to April 2014 in three pediatric institutes.

Results: There were 18 cases of hydatid disease in children; their age ranged from 7 to 16 years, and the male to female ratio was 8:1. All 18 patients were from a rural background or a farming community and of low socioeconomic status. Cattle rearing was common to the households of all patients. Nine (50%) patients had isolated liver hydatid cyst and three (16.7%) had isolated primary lung hydatid cyst. Two (11.1%) patients had multiple cysts with lung and liver involvement, while four (22.2%) had unusual presentation, i.e., primary hydatid cyst of the spleen in three (16.7%) and primary hydatid cyst of the brain in one (5.5%). All cases were managed surgically and there was no recurrence in any of our patients.

Conclusion: Hydatid disease is not rare in the pediatric age group. The liver and lung are commonly involved, but it may also present as primary disease in unusual sites like the spleen and brain. Proposed risk factors are rural background, farming community, low socioeconomic status, cattle rearing, lack of supply of potable water, and male sex. Intrinsic defects in the filter mechanism of the liver and lungs, dissemination through enteric lymphatic channels, and patency of the ductus arteriosus in early infancy are the proposed causes of unusual presentation of hydatid disease in children. During cyst removal, spillage of the contents must be...
1. Introduction

Hydatid disease usually presents in adults (19–64 years) and is relatively uncommon in children. It is characterized by cystic lesions occurring in different parts of the body, most commonly the liver (70%) and lungs (20%). Unusual sites of involvement include muscle (5%), bones (3%), kidneys (2%), heart (1%), pancreas (1%), central nervous system (1%), and spleen (1%). The peritoneal cavity, thyroid, breast, gallbladder, thigh, supraclavicular region, soft tissue of the face, pericardium, mediastinum and pleural cavity are rarely involved. In about 5–13% of cases, two organs are affected simultaneously. No site in the body is completely immune from it, except for the hair, nails, and teeth. Although hydatid cyst is uncommon in the pediatric age group, there have been isolated cases of unusual or atypical presentations reported in the literature, particularly from the Mediterranean region and North Africa. Here, we present a series of pediatric cases with unusual and unusual presentations from India in an effort to raise awareness among surgeons of this entity in children.

Our aims were to study the presentation of hydatid disease in children, evaluate the risk factors, and derive appropriate management recommendations for prevention of recurrence and disease control.

2. Methods

The records of patients admitted to three pediatric institutes between April 2006 and April 2014 were retrospectively reviewed. The inclusion criteria were age less than 17 years and a diagnosis of hydatid disease. The clinical and operative records of included patients were analyzed. Charts were reviewed according to: age, sex, chief complaints and duration of illness, background and socioeconomic status of the patient, presence of cattle and availability of potable water supply in the household. All possible radiologic investigations, including chest X-ray, ultrasonography (USG) scans and computed tomography (CT) scans were carefully reviewed. Operative intervention was carried out in all patients. Operative records with establishment of preoperative diagnosis, site and number of cysts, intraoperative findings, operative procedures and complications including spillage of contents, postoperative fate and recurrence were studied.

3. Results

The results of the study are shown in Tables 1 and 2. There were 18 cases of hydatid disease in children, whose mean age was 11.2 years (range, 7–16 years). The male to female ratio was 8:1 (16 boys, 2 girls). All patients were from a rural background or a farming community and of low socioeconomic status. Cattle rearing was common to the household of all the patients. Nine (50%) patients had isolated involvement of the liver: seven (38.9%) had cyst(s) smaller than 10 cm in diameter and two (11.1%) had isolated giant hydatid cyst (>10 cm in diameter) of the liver. Three (16.7%) patients had isolated primary involvement of the lung. Two (11.1%) patients had multiple cysts with involvement of the lungs and liver (Figs. 1–5). Four (22.2%) patients had hydatid disease with unusual presentation: primary hydatid cyst of the spleen (Fig. 6) in three (16.7%) and primary hydatid cyst of the brain (Fig. 7) in one (5.6%).

The duration of illness in our series ranged from 3 months to 2.5 years. The most common presentation in hydatid cyst of the liver was abdominal pain, while it was productive cough in patients with hydatid lung disease and those with both liver and lung involvement. Children with primary hydatid cyst of the spleen presented with a dragging sensation and/or pain in the left hypochondrium. Primary hydatid cyst of the brain presented with the features of raised intracranial pressure. In two patients with hydatid cyst of the liver, although the site was common, its presentation was not. One child who had a history of jaundice and who presented with fever had a giant tense cyst mimicking choledochal cyst; abdominal CT confirmed the diagnosis. The other patient presented with fever and...
## Table 2: Clinical features and outcome of cases with multiorgan involvement, disease in unusual sites, and disease in usual sites but presenting in an unusual manner.

<table>
<thead>
<tr>
<th>Case</th>
<th>Site</th>
<th>Cysts, n</th>
<th>Age, y</th>
<th>Presentation</th>
<th>Duration of illness</th>
<th>Procedures</th>
<th>Major complications</th>
<th>Spillage of cyst content</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Liver</td>
<td>4 + 5 = 9</td>
<td>11</td>
<td>Initially dry cough → productive cough</td>
<td>2 y</td>
<td>Pericystectomy, capitonnage &amp; omentopexy for hydatid liver, plain-meidan sternotomy for bilateral hydatid lung</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>2</td>
<td>Spleen (primary)</td>
<td>1</td>
<td>9</td>
<td>Dragging sensation, pain in left hypochondrium to left arm, irritability</td>
<td>1.5 y</td>
<td>Excision</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>3</td>
<td>Brain (primary)</td>
<td>1</td>
<td>7</td>
<td>Headache, vomiting, irritability</td>
<td>2.5 y</td>
<td>Splenectomy</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>4</td>
<td>Spleen (primary)</td>
<td>1</td>
<td>11</td>
<td>Pain in left hypochondrium → productive cough</td>
<td>1.5 y</td>
<td>Splenectomy</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>Right lung</td>
<td>1 + 2 = 3</td>
<td>10</td>
<td>Abdominal mass, jaundice, anorexia, fever</td>
<td>6 mo</td>
<td>Pericystectomy, capitonnage, omentopexy</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>6</td>
<td>Liver</td>
<td>1 (giant): 12 × 13 cm</td>
<td>9</td>
<td>Massive hepatomegaly, anorexia, fever</td>
<td>6 mo</td>
<td>Pericystectomy, capitonnage</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>7</td>
<td>Liver</td>
<td>1 (giant): 12 × 10 × 11 cm</td>
<td>14</td>
<td>Dragging sensation</td>
<td>1 y</td>
<td>Splenectomy</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>8</td>
<td>Spleen (primary)</td>
<td>1</td>
<td>9</td>
<td>Massively enlarged, dragging sensation</td>
<td>1.5 y</td>
<td>Right posterolateral thoracotomy, cyst excision, pericystectomy, capitonnage</td>
<td>No</td>
<td>No</td>
</tr>
</tbody>
</table>

### 4. Discussion

Hydatid disease has been known since the time of Hippocrates. It is endemic in areas with tropical or subtropical climates, particularly in cattle-grazing areas where the sheep–dog cycle is dominant. Most cases have been seen in the Mediterranean region, North Africa, Middle East, Australia, South America and Indian subcontinent. It has an annual incidence of 0.03–1.2 per 100,000 inhabitants in the northern hemisphere. The close association of people with sheep and dogs, compounded with non-availability of clean potable water supplies make the disease endemic to the rural areas of the Indian subcontinent.

Hydatid disease (echinococcosis or hydatidosis) is a severe cyclozoonotic parasitic infection caused by the larval form of *Echinococcus*. *Echinococcus* is a cestode tapeworm, a platyhelminth. Of the known species of *Echinococcus*, only three are of medical significance in humans: *E. granulosus* is the most common, followed by *E. multilocularis*, and finally *E. vogeli* which is rare. *E. oligarthrus* has been found in Central America. There are two main clinical categories of hydatid disease based on the species of *Echinococcus*: cystic hydatidosis caused by *E. granulosus* (which is the most common in India) and alveolar hydatidosis caused by *E. multilocularis*.

*E. granulosus* is a small, 5-mm-long tapeworm that requires two mammalian hosts for completion of its life cycle. Dogs, wild canines and other domestic carnivorous animals are definitive hosts. Sheep, cattle and horses are the intermediate hosts. Humans can accidentally become the intermediate hosts through the feco-oral route either by direct contact with the definitive host or by consuming vegetables and water contaminated with *Taenia* eggs. The eggs hatch in the duodenum and the larvae penetrate the intestinal mucosa, invading the venules. The usual destination is the liver (first filter) via the portal vein, where they either die or develop into hydatid cysts. Sometimes, the larvae pass through this sieve and reach the right side of the heart and lungs (second filter) and then all the other viscera, where they transform into small cysts leading to massive hepatomegaly. He was being evaluated for malaria when he was diagnosed, using USG, with giant hydatid cyst of the liver involving the right lobe.

All of the patients were managed surgically. Pericystectomy, capitonnage and omentopexy were performed for those who had hydatid liver disease. Posterolateral thoracotomy and cyst excision were carried out for those with hydatid lung disease. All patients with splenic hydatid disease underwent splenectomy. Left frontotemporal craniotomy was performed for removal of intracranial hydatid cyst. The details of the procedures undertaken for multiple hydatid cysts in patients with involvement of both lobes of the liver and bilateral lung disease are summarized in Table 3. All were given albendazole, corticosteroids and antihistamines perioperatively. Albendazole was continued postoperatively for 6 weeks. Histopathologic examination confirmed hydatid cyst in all (100%) patients. There was no recurrence in our series, owing to the preoperative planning and meticulous perioperative precautions to prevent spillage.
multiorgan involvement. Apart from the portal vein, dissemination via enteric lymphatic channels appears to be the most likely mechanism to account for primary hydatid disease of the spleen as seen in our study. This method of dissemination has been elucidated in other studies also.\(^3\) Direct spread from adjacent sites may be another way of involvement.\(^3\) The explanation for primary intracranial hydatid disease in our patient could be the patency of the ductus arteriosus in early infancy.\(^{11}\) We propose that intrinsic defects in the filter mechanism of the liver and lungs could be one cause of primary hydatid disease of the spleen, brain and other organs of the body.

Once in the human liver, cysts grow as much as 1 cm during the first 6 months, and then by 2–3 cm every year thereafter (range, 1–5 cm/year) depending on the host’s physical resistance.\(^8\) In most cases, a single cyst develops in one organ. An older cyst tends to form daughter cysts, from the inner germinal layer in the cyst cavity or exogenously, and is an important factor in postoperative recurrence.\(^7\) The other possibility for why there may be multiple cysts in the involved organ is increased load of \textit{Taenia} eggs in these patients. Both theories (formation of daughter cysts and increased load of \textit{Taenia} eggs) may explain the presence of multiple cysts in the liver of two of our patients.

Without treatment, hydatid cysts grow progressively, cause mass effect and may result in life-threatening complications.\(^7,8\) Occasionally, spontaneous collapse or

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Figure 1 Radiography shows: (A) multiple hydatid cysts of the lungs, two on each side; (B) regression in lesion size after treatment with albendazole therapy; (C) hydatid cyst of the middle lobe of the right lung.

Figure 2 Ultrasonography shows: (A) hydatid cysts of the liver in various stages of development; (B–D) liquid collection divided into halves, with daughter cysts corresponding to Type II and Type III of the classification of Gharbi et al.\(^1\).
Figure 3  (A–C) Abdominal computed tomography shows well-circumscribed, sharply demarcated hypodense cystic lesions in the liver. Plain chest computed tomography shows: (D) hypodense ovoid lesion with septations and daughter cysts; (E) contrast enhancement.

Figure 4  Exposure and protection of the operating field: (A, B) hydatid cyst involving segments III and IV of the liver; (C) aspiration of clear liquid; (D) two suction cannulas are seen during cyst removal.
regression has been noted, and a dead cyst becomes calcified. It is important to note that partial calcification of the cyst does not always indicate death of the parasite, whereas densely calcified cysts are considered inactive.9

4.1. Clinical presentation

The clinical features are extremely variable with a long latent period.4 With hepatic involvement, the symptoms

Figure 5  Hydatid cyst with biliary communication: (A) aspiration of bile-tinged hydatid fluid; (B) collapse of the cavity after aspiration; (C–E) extraction of proligerous membrane and dealing with the residual cavity.

Figure 6  Splenic hydatid cyst. (A, C) Intraoperative photograph during splenectomy. Specimen of the spleen: (B) the wall of the exophytic hydatid cyst is visible and (D) not visible.
are often masked because of deep localization at the level of the dome of the diaphragm. Right hypochondrial pain is the most frequent clinical symptom. Other signs and symptoms include fatigue, weight loss, anaphylactic reaction, hepatomegaly, palpable mass, cholestatic jaundice and portal hypertension from portal vein obstruction. Complications occur when the cyst(s) rupture into the biliary tree, peritoneal or pleural cavity or the lungs. Rupture into the biliary tree results in release of daughter cysts, which manifests as biliary colic and jaundice. Rupture may be spontaneous (direct) or iatrogenic. Peritoneal rupture results in acute abdominal pain and systemic anaphylactic reaction. Contamination of the cyst results in the formation of hepatic abscess, when both the pericyst and the endocyst becomes perforated (may be post traumatic).

Pulmonary hydatid disease presents with cough, purulent sputum, fever, and chest pain. It may also present with hemoptysis and allergic reaction. Rarely, the cyst ruptures into the bronchial tree, causing hydatidoptysis. It may be symptom-free, diagnosed from routine chest X-ray. The brain can be involved by metastatic spread or when a cyst ruptures in the heart or lung. Primary intracranial hydatid disease is a rare entity, and its source is most likely to be hematogenous. Distribution via the middle cerebral artery is most frequently implicated, especially in the parietal lobe, and most cerebral hydatid cysts are single and supratentorial. Nearly two-thirds of intracranial hydatid cysts are seen in children. Four of five brain hydatid cysts are associated with hepatic hydatid cysts, while one-fifth is primary. Headache and vomiting are the most common presenting symptoms in children.

Table 3  Characteristics and procedures undertaken for multiple hydatid cysts of liver in patient with involvement of both lobes of liver and bilateral lung disease.

<table>
<thead>
<tr>
<th>Cyst no.</th>
<th>Liver segments involved</th>
<th>Parenchymal relationship</th>
<th>Size (cm)</th>
<th>Volume (ml)</th>
<th>Aspirate</th>
<th>Procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>V, VI</td>
<td>Exophytic (impending rupture)</td>
<td>$11 \times 9 \times 8$</td>
<td>$790$</td>
<td>Clear liquid</td>
<td>Pericystectomy, omentopexy</td>
</tr>
<tr>
<td>2</td>
<td>VI</td>
<td>Exophytic</td>
<td>$7 \times 8 \times 6$</td>
<td>$330$</td>
<td>Clear liquid</td>
<td>Pericystectomy, capitonnage</td>
</tr>
<tr>
<td>3</td>
<td>VI, VII</td>
<td>Intraparenchymal</td>
<td>$7 \times 6 \times 5$</td>
<td>$210$</td>
<td>Clear liquid</td>
<td>Pericystectomy, capitonnage</td>
</tr>
<tr>
<td>4</td>
<td>VII, VIII</td>
<td>Intraparenchymal</td>
<td>$10 \times 8 \times 9$</td>
<td>$720$</td>
<td>Bile stained, biliocystic fistula</td>
<td>Pericystectomy, capitonnage</td>
</tr>
<tr>
<td>5</td>
<td>III, IV</td>
<td>Exophytic</td>
<td>$7 \times 6 \times 6$</td>
<td>$250$</td>
<td>Clear liquid</td>
<td>Pericystectomy, omentopexy</td>
</tr>
</tbody>
</table>
children, while focal neurologic signs are seen in adults. Spinal hydatid cyst is extremely rare in pediatric patients. In renal hydatidosis, flank mass, renal colic, persistent fever, hematuria, dysuria, pyuria, renal stones and/or hypertension are the usual presentations. Cystic rupture into the renal pelvis causes hydaturia (10–20%) and is usually microscopic. Gross hydatiduria is uncommon but has a good diagnostic utility. Painful left upper abdominal mass and dragging sensation are the usual presentations in splenic hydatid cyst. The clinical presentations of cardiac hydatidosis are nonspecific, usually manifesting as chest pain, palpitations, dizziness, dyspnea and syncope, mimicking valvular lesion or heart failure. Hydatid cyst is usually found on the left ventricle in three-quarters of cases, and its rupture may cause arrhythmia, acute arterial occlusion, acute or chronic pulmonary embolism, myocardial infarction, or cardiac tamponade. Intraperitoneal disease following hepatic hydatidosis is usually due to contamination during previous surgery or microperforations in the cyst. Primary omental hydatid cyst may be due to spread via the enteric lymphatic system and present as an abdominal mass or with complications like torsion, infection and rupture. The pathogenesis of mediastinal localization may be due to fissuring of hydatid liver or lung into the systemic circulation, transdiaphragmatic dissemination or via lymphatic abdominal hydatidosis, often presenting with chest pain and signs of mediastinal compression.

4.2. Diagnosis

The diagnostic armamentarium has to be supplemented by clinical examination and a meticulously taken history. History of bathing and swimming in pond water, which is usually contaminated with animal excreta, should be noted. Male children tend to participate in more outdoor activities and games compared to girls in villages and thus have a greater chance of coming into contact with water, soil and feces contaminated with Taenia eggs. Male preponderance has been reported in other studies. In our study, early manifestation could be attributed to the acquisition of infection at a very early age, and the presence of lower immunity in these children resulting in rapid growth of cysts.

USG and CT have a sensitivity of 85% and 100%, respectively. USG reveals the fluid character (univesicular or multivesicular structure), “falling snowflakes” sign (multiple echogenic foci of hydatid sand), proligerous membrane, and daughter cysts within the larger cyst (characteristic findings of hydatid cyst). Cysts are classified into five types on the basis of their sonographic appearance according to the classification of Gharbi et al. Magnetic resonance imaging is indicated in cases of intolerance to iodine or pregnancy or cyst with atypical CT appearance. Elevated serum alkaline phosphatase levels and eosinophilia (30% of cases) may be seen. Diagnostic sensitivity and specificity of all serologic tests is low. We discourage the use of fine-needle aspiration cytology because of its potential for anaphylactic shock and spread of daughter cysts, and because diagnosis can be made with the help of radiology.

Oval or spherical opacities in the lung fields, usually single or multiple (7.1% of cases), especially in the right lower lobes of the lung, on routine chest X-ray are considered to be suggestive of uncomplicated hydatid cysts. Both lungs (Fig. 1A) are involved in 2% of cases. Other radiographic signs are the water lily sign, meniscus sign, incarcerated membranes, air–fluid levels, hydropneumothorax, pneumothorax, and simple cavity. A single, thin-walled, well-defined spherical cerebrospinal fluid density cyst with mass effect and without perilesional edema is characteristic of brain hydatid, as seen in our case (Fig. 7). Multiple or multilocular lesions, rim enhancement and calcification are rare. Hydrocephalus has been reported. The findings in splenic hydatid disease are similar to those of hepatic hydatid disease. The appearance of soft-tissue mass or ring-shaped calcification in the renal region may be associated with renal hydatid disease. Caliceal distortion, caliectasis, nonfunctioning kidney and multiple round filling defects representing daughter cysts are seen on excretory urography. Echocardiography is the modality of investigation for cardiac hydatid cysts.

4.3. Surgical management

The treatment of hydatid cyst is essentially surgical. The principle is complete excision of the cyst (cystectomy) to confirm the diagnosis and to ensure complete cure.
Hydatid disease in children

Hydatid disease is not rare in the pediatric age group. The liver and lungs are commonly involved, but hydatidosis may also present as primary disease in unusual sites such as the spleen and brain and also in an unusual manner in the usual sites. Having a high index of suspicion is important. The age of the child may be as low as 7 years. The risk factors we found were rural background, low socioeconomic status, cattle rearing, farming community, poor education, lack of a potable water supply and male sex. Intrinsic defects in the filter mechanism of the liver and lungs, dissemination through enteric lymphatic channels, and patency of the ductus arteriosus in early infancy are the proposed causes of unusual presentation of hydatid disease in children. Anti-helminthic treatment in isolation is indicated for inoperable cases, poor surgical candidates or disseminated disease. During surgical removal of hydatid cysts and cysts without a definite preoperative diagnosis, spillage of cyst contents must be avoided (by keeping two suction apparatuses) to prevent anaphylactic reaction, recurrence and multiple hydatidosis. Community-based measures like educating people, and proper hand washing after contact with dogs or accidental contamination with dog feces.

5. Conclusion

Hydatid disease is not rare in the pediatric age group. The liver and lungs are commonly involved, but hydatidosis may also present as primary disease in unusual sites such as the spleen and brain and also in an unusual manner in the usual sites. Having a high index of suspicion is important. The age of the child may be as low as 7 years. The risk factors we found were rural background, low socioeconomic status, cattle rearing, farming community, poor education, lack of a potable water supply and male sex. Intrinsic defects in the filter mechanism of the liver and lungs, dissemination through enteric lymphatic channels, and patency of the ductus arteriosus in early infancy are the proposed causes of unusual presentation of hydatid disease in children. Anti-helminthic treatment in isolation is indicated for inoperable cases, poor surgical candidates or disseminated disease. During surgical removal of hydatid cysts and cysts without a definite preoperative diagnosis, spillage of cyst contents must be avoided (by keeping two suction apparatuses) to prevent anaphylactic reaction, recurrence and multiple hydatidosis. Community-based measures like educating people, treatment and control of all dogs and proper hand washing after contact with dogs or their feces are recommended for disease prevention and control.
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