Review Article

# Hydatid Cyst of Spleen: A Diagnostic Challenge

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## Abstract

Although splenic involvement alone in hydatid disease is very rare, spleen is the third most common organ involved in hydatid disease. The rarity of splenic hydatid disease poses a diagnostic challenge for clinicians, particularly in non-endemic areas. As the hydatid cyst can present as a simple cyst without having the classic serological and imaging features, and later can lead to life-threatening complications like anaphylaxis, hydatid disease of spleen should be considered in differential in every patient in endemic areas with cystic lesion of spleen until proved otherwise. The author used the keyword "splenic hydatid cyst" in PubMed and reviewed the scientific literatures published from January 1965 to June 2012. The present review is to accentuate the incidence, classification, clinical and pathophysiological features, differential diagnosis, diagnostic modalities, and treatment choices of hydatid cyst of spleen along with follow-up strategy and newer treatment approaches.

**Keywords:** Anaphylactic shock, Laparoscopy, Spleenic hydatid

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## Introduction

Splenic hydatidosis has been reported since ancient times. Berlot in 1790 was the first to describe splenic hydatidosis as an autopsy finding.[1] It may be detected incidentally or present with non-specific complaints. It is endemic in cattle-rearing areas of South America, Africa, Middle East, South Europe, India, and Australia. Worldwide incidence of splenic hydatid is 0.5-4%.[2] Highest incidence is in Iran (4%).[3] Spleen is the third commonest site for *Echinoccocus*.[4,5] The most commonly involved organ is the liver (75%), followed by the lung (15.4%), and the spleen (5.1%).[4,6] Parasitic cysts of the spleen are almost exclusively hydatid cysts. In endemic areas, 50-80% of splenic cysts are echinoccocal.[7]

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## Pathophysiology

Four species of Echinococcus cause infection in humans; *Echinococcus granulosus* and *Echinococcus multilocularis* are the most common, causing cystic echinococcosis (CE) and alveolar echinococcosis (AE) respectively. The two other species, *E. vogeli* and *E. oligarthrus* cause polycystic echinococcosis and are less frequently associated with human infection.

The life cycle of Echinococcus includes a definitive host (usually dogs or related species) and an intermediate host (such as sheep, goats, or swine). Humans are incidental hosts; they do not play a role in the transmission cycle. *E. granulosus* adult tapeworms are usually found in dogs or other canids. *E. multilocularis* adult tapeworms are usually found in foxes, other canids, or occasionally in cats. The adult tapeworm inhabits the small intestine of the definitive host. The tapeworm is composed of proglottid segments which produce parasite eggs containing embryos (oncospheres). The eggs are expelled in the feces of the definitive host and released into the environment, where they are infective to susceptible intermediate hosts and human incidental hosts. Following egg ingestion by the intermediate or incidental host, the oncospheres hatch from the eggs,

penetrate the intestinal mucosa, enter the blood and/or lymphatic system, and migrate to the liver or other visceral organs. A few days later, a fluid-filled cyst begins to develop, with subsequent development of multiple layers to become a metacestode (hydatid cyst) which grows at a rate of 0.3 cm-1 cm/year.[4,8] Subsequently, protoscolices develop within the hydatid cyst. In definitive hosts who ingest the intermediate host visceral organs containing hydatid cysts with protoscolices, the protoscolices evaginate, attach to the intestinal mucosa, and develop into adult worms and the cycle repeats. Transmission frequently occurs in settings where dogs eat the viscera of slaughtered animals. The dogs then excrete infectious eggs in their feces, which are passed on to other animals or humans via fecal-oral transmission. Direct transmission of echinococcosis from human to human does not occur since two mammalian species are required for completion of the life cycle.

The life cycle of *E. multilocularis* involves wild canids (usually foxes) as the definitive hosts and rodents as the natural intermediate hosts. Domestic dogs or cats may also become infected and can transmit the infection to humans directly or can contaminate food with parasite eggs. The definitive hosts for *E. vogeli* are dogs and other canids; the principal intermediate hosts are pacas and other rodents. *E. vogeli* causes polycystic hydatid disease.

Hydatid cyst consists of three layers – the outermost adventitia (pseudocyst) of fibrous tissue, middle layer of laminated membrane (ectocyst), and innermost layer of germinal epithelium (endocyst) which secretes the hydatid fluid internally and laminated membrane externally producing new generation of parasites. Possible routes of primary hydatid of spleen include arterial route after passing through liver and lung. Another route is the venous route through portal circulation bypassing liver and lung. Secondary hydatid spleen usually follows systemic dissemination or intraperitoneal spread following ruptured hepatic hydatid cyst.[4,9] Associated liver and/or lung involvement may be seen in 20-30% of cases.[9,10] They are usually solitary cysts with multiple being found in one-fourth to one-third of cases.[11] The largest cyst ever reported was in Australia which contained 57 l of fluid.[9]

## Clinical Features

The first clinical indication of the presence of spleenic hydatid disease is usually an accidentally discovered mass in the abdomen mostly in left hypochondrium[11,12] and less frequently in epigastrium. Pain, usually a dull dragging ache, is often the first clinical sign. There can be dyspepsia, constipation due to pressure on colon, dyspnea due to pushing up of the left diaphragm,[13] fistula of the colon[12,13] or perforation into the diaphragm or bronchial tree[10,14] However, up to 30% of the cases are incidental findings in asymptomatic individuals.[15]

Harefuah in 1992 described a 20-year-old soldier who presented with anaphylactic shock due to rupture of the splenic hydatid cyst induced by blunt trauma to abdomen. Case reports of acute abdomen caused by spontaneous rupture into abdominal cavity causing massive hemoperitonium requiring splenectomy are also present in the literature.[16] There are a few case reports of the association between hydatid disease of the spleen and hypersplenism.[17] However, this seems to be a coincidental finding. Hematemesis as a presenting sign has been described in literature.[18]

## Differential Diagnosis

The main problem in the diagnosis of splenic hydatidosis is in differentiating it from other splenic cystic lesions that have similar appearances on sonography and computed tomography (CT). The differential diagnosis of such lesions includes epidermoid cyst, pseudocyst, large solitary abscess or hematoma, intrasplenic pancreatic pseudocyst, and cystic neoplasm of the spleen.[19] Hydatidosis should be suspected in patients with splenic cystic lesions, particularly in endemic areas until proved otherwise.[20] The diagnosis of splenic hydatidosis should be favored if daughter cysts are present within a large cystic lesion or if cystic lesions are observed in other organs such as the liver. Imaging combined with immunological tests solves the diagnostic problem most of the times.

## Diagnosis

At present, sonography and CT are the most valuable imaging techniques for the diagnosis and evaluation of focal splenic diseases.[21] The radiographic appearance of splenic hydatidosis varies and is influenced mainly by the location of the cyst, age of the cyst, and associated complications, such as secondary infection, and rupture.[22] Findings range from purely cystic lesions to a completely solid appearance and are classified on the basis of appearance. Simple cyst with no internal architecture, cyst with daughter cysts and matrix, calcified cyst, or complicated cyst can be observed.

Plain radiograph may demonstrate a marginal/crumpled egg-shell-like calcification in the splenic area suggestive of splenic hydatidosis. Other findings are an elevated left hemi-diaphragm, displacement of stomach, and/or left colonic flexure.[18]

Ultrasound, CT, and magnetic resonance imaging (MRI) are all appropriate for imaging of the Echinococcal liver

disease. Imaging early in the disease process often reveals one or more simple-appearing cysts of varying size that are surrounded by either a thin well circumscribed wall or less often a by a slightly thickened irregular wall, with the wall thickness ranging from 1 to 10 mm. The next stage of the life cycle involves the development of "daughter cysts."[23] At this stage, the cyst will typically demonstrate low echogenicity on ultrasound, hypodensity on CT, and low to intermediate signal intensity on T1- and high signal intensity on T2-weighted MR images.

The Sonographic findings of splenic hydatid cysts are not specific, even if the typical findings of solitary, anechoic lesions are demonstrated. Sonography is helpful, especially in the early stages, when the lesion is cystic, in detecting daughter cysts, hydatid membranes, and hydatid sand.[24,25] Ultrasound has a sensitivity of approximately 90-95%.[26,27] The most common appearance on ultrasound is an anechoic smooth, round cyst, which can be difficult to distinguish from a benign cyst. When the cyst contains membranes, mixed echoes will appear that can be confused with an abscess or neoplasm. When daughter cysts are present, characteristic internal septations result. Hydatid sand reflects a complex image which consists predominantly of hooklets and scolexes from the protoscolices. This finding may be visible when shifting the patient's position during imaging. When ultrasound reveals infoldings of the inner cyst wall, separation of the hydatid membrane from the wall of the cyst, or hydatid sand, a diagnosis of hydatid disease is probable.[28] When the fluid pressure in cyst becomes high, dissections may occur, resulting in the detachment of parasitic membranes, and these undulating pathognomic membranes seen on ultrasonography (USG) and CT are known as snake/serpent signs.[25] In the more advanced stage of collapse; the membranes appear twisted on imaging known as spin/whirl sign.[29] The "Water-Lily" sign refers to a collapse of the endocyst layer which results in the inner cyst lining falling into the fluid in the dependent aspect of the cystic lesion. This gives the appearance of debris floating on a layer of fluid within the cyst. On axial sonogram, parallel stripes, called double-line signs that delineate the space between parasite's ectocyst and host's pericyst are seen. When seen, this sign helps to differentiate hydatid cysts from simple cysts, cystic tumors, pseudocysts, and metastases. Sonography is particularly useful for detection of the double-line sign. US images in case of *E. multilocularis* infection show the typical Hailstorm pattern, characterized by multiple echogenic nodules with irregular and indistinct margins. After medical therapy, detached membranes within the degenerated cyst – Snake or serpent sign indicates that the parasite is responding to medical therapy.[30]

Ultrasound also allows for the classification of the cyst(s) by biologic activity, which may influence the choice of treatment; these categories are: Active, transitional, or inactive. Characteristics on ultrasound that are suggestive of an inactive lesion include a collapsing, flattened, elliptical cyst (corresponds to low pressure within the cyst), detachment of the germinal layer from the cyst wall ("water lily sign"), coarse echoes within the cyst, and calcification of the cyst wall.[31]

CT has a higher overall high sensitivity than ultrasound, with sensitivity rates of 95-100%.[26,27] CT is the best mode for determining the number, size, and anatomic location of the cysts, and is also better than ultrasound in detecting extrahepatic cysts. CT may also be used for monitoring lesions during therapy and to detect recurrences. Lesions do not enhance after administration of I/V contrast material.[32] The CT attenuation in hydatidosis depends on the intracystic content. Hydatid cysts usually have a homogeneous fluid content showing water attenuation values on CT.[32] However, hydatid cysts may show high CT values on unenhanced CT scans. The presence of intracystic debris, hydatid sand, and inflammatory cells are presumed to cause the high CT values in these cases.[33] Calcification may occur in the wall of the cyst after the death of the parasite and may be observed on plain abdominal radiographs. However, CT is more sensitive than plain films or sonograms in depicting subtle cyst wall calcification. When there is calcification, it will be shown better on unenhanced CT because IV contrast material may cause the calcification to be masked by the surrounding enhanced splenic parenchyma. Calcification is usually curvilinear or ring-like and involves the pericyst.[34] Presence of mural calcification and/or daughter cysts rules out other cystic lesions of spleen. CT scan shows a multivesicular hydatid cyst containing peripheral daughter cysts attached to the membrane of mother cyst in the center. Density of the mother cyst is higher than that of the daughter cyst because of advanced degenerative changes resulting in intracystic debris, "hydatid sand," and inflammatory cells. Difference of density gives characteristic CT picture that is diagnostic for the presence of multivesicular *E. granulosus* cysts. Daughter cyst densities vary between 0 and 15 H, in contrast to higher densities of mother cyst fluid (30-40 H). Exogenously proliferating cysts which are only rarely observed are seen in *E. multilocularis* along with large, irregularly shaped, hypoattenuating lesion with diffuse punctate calcifications, which are more clearly seen on the unenhanced image.[35]

MRI may be an important imaging technique in the diagnosis and evaluation of hydatid disease according to recently published reports.[36,37] In patients with

negative serology and inderminant USG and CT investigations, MRI has been described as helpful. MRI can differentiate parasitic, nonparasitic, or traumatic cysts by demonstrating "low-signal intensity rim" – so-called RIM SIGN and has been described as characteristic of hydatidosis. The hypo intense rim sign, characteristic of hydatid cyst, is best seen in the T2-weighted sequence.

On echo T2-weighted image, septated hyperintense splenic mass is seen along with smaller peripheral cysts which represent the daughter cysts. Axial gradient-echo unenhanced and delayed contrast-enhanced 3D T1-weighted images show cystic nature of the mass. These are typical features of hydatid cysts, although similar appearances can be found in epithelial cysts or lymphangiomas. Axial gradient-echo T1-weighted MR image shows a hydatid cyst with a hypointense fibrous pericyst. The hydatid matrix has intermediate signal intensity, and peripheral daughter cysts are hypointense relative to the matrix. On an axial T2-weighted MR image, the matrix is hyperintense and the daughter cysts again are relatively hypointense. After medical therapy T2-weighted-echo MR image shows degenerated cyst having features of detached membranes are seen within original cyst as "snake (or serpent) sign." T2-weighted echo MR image shows multivesicular hydatid cyst with daughter cysts and intervening septa. Daughter cysts may show high or low signal intensity, depending on factors such as composition and volume of fluid, infection, degeneration of parasitic material, and presence of scolices. Lesion is surrounded by low-intensity rim.[38] However, we believe that MRI should not be used as the first imaging method in the study of patients with proved or suspected hydatid disease.

Among the serological methods, immunoelectrophoresis has 90-95% sensitivity. It remains positive for 1 year after the organism has been eradicated. So it has a role in screening in endemic areas. Indirect hemagglutination has a sensitivity of 85% and it remains positive for many years so it has no role in screening. Capron, *et al*.[39] reported the presence of an antigen specific for *E. granulosus* that appeared immunoelectrophoretically as a band of characteristic morphology and location when tested against sera from human patients. They named this band arc 5 because of its relative position in the immunoelectrophoretic pattern. However, in residual or highly calcified hydatidosis, the arc 5 may be absent. In calcified hydatid cysts, it is postulated that the physical status of the hydatid cyst membranes influences the degree of antigen stimulation of the immune system of the host, and therefore, affects the success of the immunodiagnostic test.[40] False-positive serologic test results have been reported in patients with malignant disease.[41] Several immunologic tests may also help in the diagnosis, although laboratory data are sometimes uncertain; hepatic hydatidosis is reported to be confirmed in 80-94%, but extrahepatic hydatidosis only in 65% of cases, even when the immunologic tests are based on multiple methods.[42,43] Serological tests for echinococcosis show high sensitivity and specificity (in ranges of about 75-100%) and offer a good differentiation rate between *E. granulosus* and *E. multilocularis* (up to 95%).[43] Elevated levels of Immunoglobulin E (IgE) are non-specific indicator of prior sensitization/active infection. Although elevated levels of Immunoglobulin M (IgM) class-specific to echinococcal organisms may be sensitive indicators of recurrent disease.

Imaging when combined with serological tests such as ELISA, immunoelectrophoresis, or indirect Hemagglutination test can lead to successful diagnosis of spleenic hydatid in 90% of the cases.[44] In spite of all the above investigations, diagnosis is always challenging. Willinki, *et al*. described personal history, presence of calcification, daughter cysts, or concomitant cystic lesions in liver and other organs that are helpful for diagnosis of splenic hydatidosis.

## Treatment

Although currently antihelminthic medications are used in addition to surgery, these drugs are not expected to replace surgery in the near future. Most antihelminthic drugs are absorbed poorly from the gastrointestinal tract and they do not reach adequate concentrations within the cyst cavity to kill the parasites. As oral drug administration is not effective, the standard treatment for hydatid cyst disease remains surgery (open or laparoscopic).[45]

## Splenectomy

Currently, splenectomy is the conventional treatment of choice as it has a low morbidity and mortality rate.[7,8] Due to constant dreadful complications of rupture, cysts, especially larger ones should be treated surgically. Literature favors total splenectomy in larger cysts, because the splenic parenchyma is significantly reduced due to pressure atrophy and thick fibrous membrane, as seen in hepatic hydatid cysts, is quite thin and fragile in splenic hydatid cyst, so risk of intraoperative rupture is high in such cases if conservative approach is adapted during surgery.[46] So splenectomy must be carried out in large and giant hydatids of the spleen localized in the organ or in its hilum and in irreversible derangement of the organ function.[34] The mainstay of treatment in AE is surgical resection. Due to the invasive growth of AE, the resection has to follow oncological principles and should provide free margins of at least 1 cm.[47]

However, splenectomy is associated with hemorrhage, pancreatitis, gastric injuries, and overwhelming post-splenectomy infection (OPSI) which are all reviewed in literature.[48,49] The sepsis-related deaths occurred in 1.9% of the adults and 4% of the children who underwent splenectomy.[49] The clinical syndrome of OPSI comprises culminant bacteremia, disseminated intravascular coagulation, multiple organ failure, severe hypoglycemia, and often rapid death. Its reported incidence after splenectomy varies from 0.9 to 60% with mortality exceeding 50%. The Hib and meningococcus C conjugate vaccines should be given at least 2 weeks before or 2 weeks post-splenectomy. The splenectomized patient should be vaccinated to decrease the risk of overwhelming post-splenectomy sepsis (OPSS) due to organisms such as *Streptococcus pneumoniae*, *Haemophilus influenzae* type B, and *Neisseria meningitidis*. Patients should be educated prior to discharge on the risk of OPSS and their immunocompromised state.

Relative contraindications to surgery include the presence of multiple cysts, cysts that are difficult to access, dead cysts, and small cysts that are asymptomatic.[50] The benefit of surgery is immediate and total cure. The surgery does, however, carry risks in excess of those generally associated with surgery. These include secondary echinococcosis due to spillage (in 2-21% of cases[51] and a reported operative mortality of 0.5-4%.[52,53] In addition, recurrence is possible if other cysts are present but not resected. Incidence of recurrence has not been defined yet.

## Laparoscopy Approach

Since it was first reported in 1991, laparoscopic splenectomy has gained increased acceptance and is the standard procedure for the surgical treatment of hematological splenic disorders.[54] Technological progress has allowed partial laparoscopic splenectomy to be performed with lower risks and good hematological results by preserving the immunological function of the spleen.

The laparoscopic technique is an easy-to-apply, safe, and effective method to conduct spleen hydatid cyst surgery. Laparoscopic surgery, for patients with unique, small-sized, superficially located cysts, will yield a surgical success similar to the conventional open surgery, with the advantage of minimally invasive procedures and will also help prevent intraperitoneal spillage of cyst contents.[55] One of the retrospective study compared open and laparoscopic hydatid cystectomy to evaluate the reliability and feasibility of the laparoscopic approach. Recurrence of the cyst was not seen in any patient during the post-operative serologic and sonographic follow up period, which was maintained at least 24 months after operation. But the morbidity rates for laparoscopic and open approaches were 14.2% and 33.3% respectively. The most frequent post-operative complications with laparoscopic approach were wound infections.[56]

Khoury, *et al*. recommended the laparoscopic approach for uncomplicated hydatid cyst of liver and spleen. In their study, 108 hydatid cysts were approached laparoscopically, and there was no mortality.[51] Nowadays, laparoscopic splenectomy has been done in some centers and is shown to be feasible and safe, offering the advantages of open surgery.[51,57-59] The reluctance to perform laparoscopic surgery for hydatid disease is probably because of the concern of spillage of the fluid into the peritoneal cavity, with the possibility of anaphylactic reaction and recurrence, secondary to intraperitoneal dissemination subsequent to uncontrolled puncturing of cyst.[48,34,60] In one study, spillage of the protoscolex-rich fluid during surgery occurred in 5-10% of the cases, despite all the precautions taken to prevent it, but this does not necessarily lead to dissemination.[61] Recurrence after surgery was reported in up to 18% of the cases, which may be due to incomplete removal, spillage, or growth of small occult cysts that were missed initially.[62] Laparoscopic surgery can be preferred for patients with unique, small-sized, superficially located cysts, but if there is more than 1 cyst, large cysts, cysts in deep organ locations, infected cysts for the second time, open surgery should be done.[63]

## Spleen-Preserving Surgery

Spleen-preserving surgery like partial splenectomy, cyst enucleation, deroofing of cyst with omentoplasty or external drainage are in vogue. Authors in favor of total splenectomy argue that there is a lower risk of recurrence and post-operative hemorrhage.[4,8] Those in favor of conservative surgery believes total splenectomy predisposes to sepsis and this should be avoided, especially in children.[64] The strive for salvage of the spleen whenever possible is fully justified based on updated knowledge of the role it plays in promoting protection against infection.[65] Conservative techniques are used for superficial cysts confined to one of the poles of spleen and cysts with extensive adhesions.[60] Preservation of spleen should always be tried in children to avoid OPSI, mortality rate of which exceeds 50%.[66] Atmatzidis, *et al*. compared splenectomy and spleen-preserving surgery. There was no significant difference between the splenectomy and spleen-preserving groups concerning median hospital stay and post-operative complication rate. The median follow-up was 52 months. Recurrence occurred in 12% patients in the splenectomy group and in 14% patients in the spleen-preserving group. The study concluded there

showed no significant increase in recurrence between the two surgical approaches.[67] Most of the other studies that also show recurrence rate after total splenectomy do not differ significantly from spleen-preserving surgery and complication rates are also comparable.[64,68]

Following a hepatic infestation, hydatid cysts are known to recur with a frequency from 8 to 20%. Although reliable data are missing, recurrence rates after splenic hydatidosis seem very low following splenectomy, but might be higher after spleen-sparing operations or when additional cysts were removed from other organs. The goal of partial splenectomy in splenic hydatidosis is to preserve as much splenic parenchyma as possible. But it leads to a bigger splenic remnant with more difficult hemostasis on the splenic surface, eventually leading to post-operative complications like hematomas or abscesses. Last but not least, the local inflammation and perisplenic adhesion as well as splenomegaly in cases of large cysts make the laparoscopic dissection more difficult. This is why a hand-assisted laparoscopic approach is also a good option for surgical treatment.[69]

## Robotic Splenectomy

Laparoscopic partial splenectomy is a challenging procedure. The introduction of the robotic surgical system has revolutionized the field of minimally invasive surgery by improving vision and motion control.[70,71] However, so far, there are only a few reports on partial or even total robotic splenectomy.[72] Although laparoscopic splenectomy became the standard procedure for the surgical treatment of hematological splenic disorders, the role of robotic splenectomy is still a matter of debate because the robotic equipment is expensive, still bulky, and the surgical team needs special training. However, there are some limitations of laparoscopy in difficult splenectomies (massive splenomegaly, portal hypertension, partial/subtotal splenectomy) which can be overcome by the robotic system through better visualization and maneuverability and motion control allowing for a better dissection of the splenic vessels and for precise and time-efficient intracorporal maneuvers. The superior maneuverability of the tips of the instruments within the splenic hilum allows for a more accurate dissection of the splenic vessels and identification of the pedicle of the remnant spleen, increasing the rate of successful partial splenectomies.[72] Furthermore, better visualization and maneuverability and motion control make the lysis of perisplenic adhesions more bloodless and safer than in the laparoscopic approach. The median overall operative time is shorter when compared with the operative times of early minimally invasive splenectomies. With increasing experience using this technique, it is suggested that even more time can be saved. That is why robotic approach is indicated in difficult splenectomies.

## Percutaneous Aspiration Irrigation and Reaspiration

Percutaneous aspiration irrigation and reaspiration (PAIR) is the only method that is diagnostic and therapeutic. PAIR, under CT and USG guidance is advocated in patients who do not give consent for surgery or have greater anesthetic risk. It is contraindicated in uncooperative patients and mature calcified cysts. Anaphylactic shock and spillage are rarely seen.[73,74] Splenic hydatid cyst abscess-formation was seen in cases that underwent percutaneous treatment in some studies which were surgically proved, although their appearance on sonography and CT imaging methods didn't confirm a diagnosis of abscessed splenic hydatidosis.[75] Percutaneous drainage of splenic abscessed lesions must be avoided when hydatid disease is suspected.[76] Long-term results indicate that percutaneuos treatment modality of spleenic hydatidosis can be an effective and a safe method and causes no major complications.[77] However, the duration of treatment follow-up is long and there is uncertainty about the possible recurrence rate. Simple and small cysts must be selected by combined opinion of the surgeon and the interventionist. Formerly, the percutaneous drainage of the hydatid cyst was deemed to be a hazardous procedure due to the potential risk of anaphylaxis and dissemination.[78] Filice and Brunetti, however, observed no anaphylactic reaction or peritoneal dissemination having applied the PAIR technique for 231 cysts in 163 patients. In agreement with this report,[79] Haddad, *et al* experienced only minor complications including pain, mild fever, pleural effusion, and transient hypernatremia in their series. In a 6-9-year follow-up involving USG, CT scans, and serology tests, no local recurrence or spread of the disease was found.[80] Etlik, *et al*. evaluated the effectiveness of percutaneous treatment under sonographic guidance in abdominal hydatid cysts. A decrease in the dimensions of the cysts, solidification of the contents, and irregularity in the walls of cysts, all of which were considered signs of cure, were found in all patients. Recurrence was observed in one case and anaphylaxis in one.[81] Treatment with PAIR is successful less often for cysts, that on sonography, appear predominantly solid and for multiseptated cysts that contain multiple daughter cysts. These cysts may be treated better surgically, although some centers are reporting some short-term success with radiofrequency ablation of these cysts. In addition to routine post-procedural observation, these patients should be followed-up closely with serology and sonography every week for the 1st month, every other month for 6 months, and every year for 5 years.[34]

## Drug Therapy

With multiple cysts, multiple initial locations, recurrence in multiple organs and especially in the peritoneum represents a good indication of chemotherapy alone which may also be the first step before a hazardous operation in complicated cases.[82,83] Albendazole is significantly more effective than mebendazole in the treatment of whole hydatid cysts (77.9% vs. 50.6% respectively).[84] It is usually preferred at an average daily dosage of 15 mg/kg/day; it must be given continuously, without those treatment interruptions which were recommended in the past.[82,83] Blood count and transaminases must be checked every week for the 1st month and every month thereafter.[82,83,85,86] Combination of chemotherapy and surgery or PAIR is increasingly being used. Presurgery treatment with albendazole may facilitate a complete removal of the germinal layer, as shown by Genetzakis, *et al*.[86] Praziquantel, which is protoscolicidal but has no efficacy on germinal layer cells, may be added, especially after surgery when the risk of spillage is high. But generally to prevent the recurrence of hydatid cysts, various measures include reduction of viability by using drugs, prevention of cystic fluid spillage, and complete removal of their germinative membranes which is achieved by splenectomy.[87] Ideally, preoperative prophylaxis, i.e., albendazole and praziquantal should be started 1 month and 2 weeks prior to surgery respectively or at least 4 days before surgery according to WHO guidelines.[85] It stabilizes cysts, decreases tension inside the cyst, decrease anaphylaxis and recurrence. Patients who undergo PAIR typically take albendazole or mebendazole from 7 days before the procedure until 28 days after the procedure.[88]

In case of AE, to reduce recurrence, patients are placed under treatment of benzimidazoles (Albendazol, Mebendazol) 1-3 months before surgery and for up to 24 months after surgery. In inoperable cases or when only palliative resection can be achieved, chemotherapy should be continued throughout life because benzimidazoles only have parasitostatic action. Nevertheless, as it has been shown that fluoro-D-glucose positron emission tomography (FDG-PET) can discriminate active from inactive lesions and attempts at discontinuation of benzimidazoles have been made. But recurrence rates are high and discontinuation cannot be recommended, in general, at this stage.[89,90]

Medical management with antihelminthic agents alone has yielded some success, with between 30% and 50% of the patients treated showing some improvement in the radiologic appearance of the cysts. However, cure should not be expected with the currently available medications alone. The failure rate for medical management alone is 25%, with most cases of relapse occurring within 2 years of cessation of therapy. Regarding post-operative therapy, studies show that if the surgeon totally excises the cyst without spillage of contents in the operative area, it is possible to avoid use of drug therapy after surgery. It should be noted that drugs should never be used alone as evidence suggests that the drugs principally act parasitostatically and not as parasiticides, hence they cannot cure.[91] However, no controlled long-term studies have ever evaluated the efficacy of chemotherapy to prevent recurrence after surgery, as well as the optimal schedule before and after surgery or PAIR and the risk/benefit of a combined treatment with praziquantel. This clearly indicates the absence of consensus and the need for guidelines.

## Intraoperative Therapy

Intraoperatively, the use of scolicidal agents like cetrimide, hypertonic saline, alcohol, or 0.5% silver nitrate solutions before opening the cavities tends to kill the daughter cysts, and therefore, prevent further spread or anaphylactic reaction. Alcohol sclerosis of hydatid cysts has proven to be a safe and effective therapeutic option. Percent reduction of cyst size has been reported at between 73% and 99%.[50,92,93] In a series of 61 hydatid cysts treated percutaneously, only one cyst showed recurrence at 4 years.[50] This was successfully treated with a second percutaneous procedure. A second large study of 57 hydatid cysts of the liver treated percutaneously had a recurrence rate of 2%,[92] with one cyst recurring after 11 months. This cyst was successfully treated with a second percutaneous procedure.

## Recurrent Hydatid Cyst

Despite the number of therapies now available, recurrence remains one of the major problems in the management of hydatid disease, ranging from 4.6% to 22.0% in different series.[94-97] The main reasons for recurrence appeared to be microscopic spillage of live parasites, failure to remove all viable cysts at inaccessible or difficult locations, or leaving a residual cyst wall at the initial operation. Recurrence detected during the early post-operative period is indicative of inadequately treated cysts in the first operation.[98] Recurrence is actually never seen following complete resection of an intact cyst with radical surgical interventions when feasible. However, with more conservative procedures, the rate of recurrence reaches 12%.[98] The confirmation of therapeutic efficacy is difficult since recurrences may develop many years later. Thus, the post-operative follow-up period should be at least 3 years and continued as long as possible.[96] Blood titer does not return to normal values soon after the operation, therefore, positive serologic tests are not significant for the diagnosis of recurrence, which had

to be confirmed by USG or CT. Differentiation of the remaining cavities of effectively treated cysts from locally recurrent disease is difficult, therefore, increase in size of the cyst on serial examination, which has proved to be effective is a reliable marker of recurrence.[94] The choice of operation (radical or conservative) is based on cyst location, size, morphology, complications, prior treatment, and presence of technical difficulties owing to adhesions. The fact that appropriate primary treatment failed owing to "vitality" of the parasite, more radical treatment might be indicated. In patients with recurrence after evacuation of cyst contents, complete cyst resection could appear to be reasonable, so long as it would be done safely. However, these radical operations are technically more difficult, and reoperations have higher morbidity and mortality rates.[96] In most recurrent cases, conservative treatment (repeat evacuation with or without partial cystectomy) can be done.

### Follow-up

Assessment of treatment efficacy relies mainly on the morphology and size of the cyst(s), appreciated by repeated imaging. Following treatment, hydatid cysts are best followed sonographically due to the absence of ionizing radiation and ease of access to this modality. The cysts will gradually fragment, collapse, and reduce in size leading to the sonographic appearance of a pseudosolid or echogenic mass. At this stage, the lesions can easily be mistaken for a tumor or abscess. Color Doppler can help differentiate a collapsed hydatid cyst from a tumor as the collapsed cyst will demonstrate lack of internal vascularity, whereas a tumor in the absence of necrosis will usually demonstrate internal blood flow. A major endpoint is the disappearance of the cyst; however, persistence of ultrasound or CT-scan image may well be associated with parasite death and thus response to the treatment. Serology is of little use to assess the cyst viability; specific IgG4 could be good indicators of treatment efficacy but are not widely available.[91,99]

The search for new drugs is ongoing. The parasitocidal effect of nitazoxanide was recently proven *in vitro* but has never been tested in human CE.[91,99] New therapeutic approaches such as radiothermal ablation are currently under investigation.[100] What is most urgently needed is to construct controlled studies aimed at more clearly specifying the treatment guidelines.

## Conclusion

Despite the advances in medical technology, it is still difficult to differentiate between parasitic and non-parasitic splenic cysts. Although serology, sonography, and CT can provide pathgnomonic evidence of splenic hydatid disease but cannot always reach the diagnosis. It is important to underline that it is relevant to maintain hydatid disease and its possible complications as one of the differential diagnosis to consider in endemic areas when the clinical conditions justify. Patient's personal history, the presence of calcification of the cyst wall, and especially the presence of daughter cysts in a large cystic lesion or concomitant cystic lesions in the liver or other organs, are helpful for diagnosing splenic hydatidosis. CT remains the most sensitive investigation for diagnosis. MRI is an emerging imaging technology. Total splenectomy is the treatment of choice in adults because it offers complete cure from the disease with low mortality and morbidity rates, but in children spleen-preserving surgery should be considered to prevent OPSI. Although reliable data are missing, recurrence rates after splenic hydatidosis seem very low following splenectomy, but might be higher after spleen-sparing operations or when additional cysts were removed from other organs. Therefore, preoperative and post-operative medical prophylaxis in form of albendazole and praziquantal should be considered to prevent recurrence.

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