Pediatric Radiology
https://doi.org/10.1007/s00247-024-06080-7

MINISYMPOSIUM: PEDIATRIC INFECTIOUS DISEASE IN LATIN AMERICA

The many faces of pediatric hydatid disease: a pictorial review

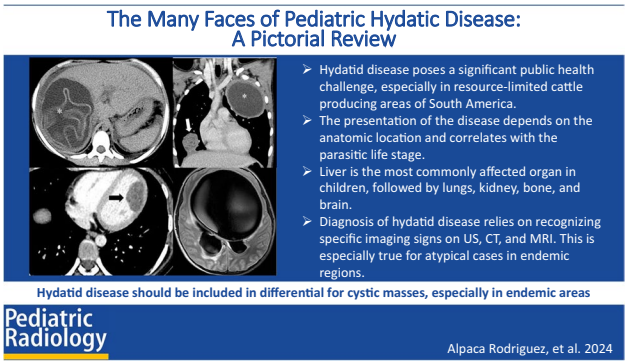
Larry R Alpaca Rodriguez[1] · Joel P. Kirschbaum-Chrem[2] · Gustavo Romero[3] · Edilberto Villanueva[4] · Carlos F. Ugas Charcape[2]

Received: 2 December 2023 / Revised: 8 October 2024 / Accepted: 12 October 2024

Abstract

Hydatid disease, caused by the larval stages of *Echinococcus* species, poses a significant public health challenge, especially in resource-limited cattle-producing areas of South America. The number of cases in children under the age of 15 is nearly 16% of the total cases in South America according to the latest report of the Pan American Health Organization (PAHO). The presentation of the disease depends on the anatomic location and correlates with the parasitic life stage. The liver is the most commonly affected organ in children, followed by the lungs, kidney, bone, and brain. The classification of hydatid cysts varies based on the parasite's stage, from purely cystic lesions to solid masses. The radiological approach varies by cyst location. Clinically, hydatid disease symptoms are nonspecific and organ-dependent, with imaging playing a crucial role in diagnosis. Complications include cyst rupture and superinfection, with potential severe consequences. This pictorial essay aims to illustrate the manifestations of hydatid cysts in an endemic population and highlight atypical signs for radiologists evaluating pediatric cysts in endemic regions.

Graphical Abstract



Keywords Children · *Echinococcus* · Hydatid disease · Parasites · Latin America · Superinfection

✉ Joel P. Kirschbaum-Chrem
jkirschbaum@insnsb.gob.pe

Larry R Alpaca Rodriguez
larrymr1204@gmail.com

Gustavo Romero
garb1990garb@gmail.com

Edilberto Villanueva
edvillanueva.md@hotmail.com

Carlos F. Ugas Charcape
drugas2801@gmail.com

[1] Hospital Sabará, São Paulo, Brasil

[2] Instituto Nacional de Salud del Niño San Borja, Av. Javier Prado Este 3101, Lima 15037, Peru

[3] Universidad Nacional de Trujillo, Trujillo, Perú

[4] Hospital Alberto Barton, Callao, Perú

Published online: 26 November 2024

Introduction

Hydatid disease continues to pose a significant challenge to public health. It is distributed worldwide, but is particularly prevalent in South America, Africa, and Asia, and is endemic in the cattle-producing areas of Argentina, Brazil, Chile, Peru, and Uruguay, which have limited resources [1]. This parasitic infection is caused by the larval stage of the cestodes *Echinococcus granulosus* (cystic) and less frequently by *Echinococcus multilocularis* (alveolaris), *Echinococcus oligarthus* (polycystic), and *Echinococcus vogeli* (extremely rare) [2]. Humans, as accidental hosts, contract the infection by ingesting eggs from contaminated water or vegetables or via direct contact with dogs, leading to the development of hydatid cysts in various organs. Hydatid cysts are mainly acquired during childhood and often go underdiagnosed until adulthood, due to the slow growth of the parasite. Diagnosis typically occurs in the third or fourth decade of life, or when symptoms arise [1, 3].

The Pan American Health Organization (PAHO) reported a total of 54,525 cases of hydatid disease in South America from 2009 to 2021 [4]. According to the latest PAHO epidemiological report for the South American region, 15.8% of the new cases occurred in children under the age of 15 (2019–2021), indicating ongoing transmission of the disease [4, 5]. This highlights the need for continued efforts to control the spread of this parasitic infection.

The varying prevalence of hydatid disease by anatomic location is related to the parasite's life cycle [6]. The parasite can be in an active (fluid-filled sac), transitional (cyst degeneration), or inactive (calcified cyst) stage. Hydatid disease primarily affects the liver, which serves as the first barrier to the parasitic invasion from the bowel through the portal circulation. Secondary involvement can occur in almost any anatomical site via hematogenous dissemination [7].

Hydatid disease predominantly affects the liver (43–89%), followed in frequency by the lungs (10–35%), kidneys (2–3%), bones (1–4%), and brain (1–2%), and more unusual, the heart, spleen, pancreas, muscles, and orbit [1, 4, 8, 9].

This pictorial essay aims to illustrate the manifestations of hydatid disease in an endemic pediatric population and provides guidance for radiologists on identifying imaging signs when evaluating cysts in children from endemic regions.

Classification

The imaging appearance of the hydatid cyst varies depending on the parasite's stage, ranging from purely cystic lesions to solid-appearing masses [10]. Different classifications have been proposed for hydatid cysts. In 1981, Gharbi et al. first described five different sonographic appearances [11]. In 2003, the World Health Organization (WHO) and its working group on echinococcosis proposed a new classification based on sonographic characteristics that reflect the current understanding of the disease and its natural evolution. It is a simple three-group classification that aims to unify the diagnosis and treatment of hydatidosis [12].

According to the WHO classification, a young active/viable cyst manifests as a simple, fluid-filled structure with a "double-line sign" (CE1) or as a multiseptated, "rosette-like/honeycomb" (CE2) cyst (group 1, WHO classification). These may progress to a transitional stage (group 2, WHO classification) where the integrity of the cyst is compromised, leading to detached membranes (CE3a) or daughter cysts in a solid matrix (CE3b). Finally, due to starvation, the cyst dies and degenerates into a heterogenous (CE4) and/or calcified (CE5), inactive/degenerative cyst (group 3, WHO classification) [12, 13]. This classification is comparable to that of Gharbi et al., as seen in Supplementary Material 1.

Clinical presentation

The clinical features of hydatid disease are usually nonspecific and dependent on the involved site. Imaging plays a crucial role in diagnosis, often found incidentally when asymptomatic. Symptomatic cysts typically arise from increased size and complications. Systemic manifestations may include appetite loss, fever, fatigue, and hypersensitivity signs. In cases where the abdomen is involved, particularly the liver, common symptoms are right upper quadrant pain, abdominal distention, and the presence of a palpable mass. If the respiratory system is involved, common symptoms include cough, hemoptysis, and hydatid vomica.

Complications

Complications associated with hydatid disease include cyst rupture, which can occur as a result of aging, chemical reactions, or host defense mechanisms. Parasitic membrane degeneration may lead to cyst rupture in 50–90% of cases [14, 15]. Superinfection is another potential complication of hydatid disease.

Lesions classified as group 1 and group 2 according to the WHO classification may experience three types of rupture: contained, communicating, and direct. A hydatid cyst is composed of three layers. The outermost pericyst is composed of modified host cells, the acellular laminated

membrane in the middle, and the innermost endocyst (germinal epithelium), which produces the laminated membrane and infectious scolices (larval stage) [3, 4].

Contained rupture occurs when the endocyst becomes detached from the pericyst and is clinically silent. Communicating rupture occurs when the cyst ruptures into an anatomical structure incorporated in the pericyst, such as the biliary tract or bronchial tree. This may cause biliary obstruction and cyst evacuation or infection.

Direct rupture involves the rupture of both the endocyst and the pericyst (perforation), causing cyst contents to spill into the pleural and peritoneal cavities (seeding) or a hollow viscus. Direct rupture is associated with the most clinically significant consequences, including anaphylaxis, dissemination within the host (secondary hydatosis), and bacterial infection of the pericyst cavity. In cases of communicating and direct ruptures, the cyst loses its spherical shape and becomes significantly smaller. The consequences of rupture generally outweigh the mass effect of hydatid cysts, except in the brain, where mass effects can be severe. Other potential complications include exophytic growth, trans-diaphragmatic thoracic involvement, portal vein involvement, and abdominal wall invasion [7, 9, 13]. Up to 25% of ruptured hydatid cysts may become infected (bacterial superinfection), resulting in the abscess formation. From an imaging perspective, poorly defined lesions are often considered indicative of infected cysts, typically characterized by the presence of gas or air–fluid levels. However, the presence of air within the cyst does not necessarily indicate infection when establishing the diagnosis of communicating or direct rupture [14, 15].

Radiological approach by location

The choice of diagnostic modality primarily depends on the location of the parasitic infection. Cystic formations may exhibit distinctive features that assist clinicians in detecting complications [16]. Similarly, certain atypical signs can aid in the differential diagnosis of cyst types, making the choice of imaging modality crucial, as each technique offers unique insights into the specific complications associated with hydatid cysts.

Liver

The liver, being the most common site of infection, functions as a filter for parasitic larvae [9, 16, 17]. These larvae enter the bloodstream through the intestinal wall and can potentially develop into cysts in the liver. The liver receives blood directly from the gastrointestinal tract via the portal vein, making it a favorable site for cyst growth, particularly in the right lobe. The size of the cysts can vary significantly, and they may remain clinically silent for years due to the liver's considerable reserve and ability to function despite the presence of cysts. When symptoms do occur, they usually include abdominal discomfort or pain and a palpable mass, and less commonly jaundice, if the cysts obstruct the biliary system [3, 14]. The most common complication of these cysts is intrabiliary rupture, primarily affecting the right hepatic duct (communicating rupture) [18].

Abdominal radiography may show hepatomegaly or right hemidiaphragm elevation due to large or multiple cysts [14]. Calcification, seen in 20–30% of patients, may occur internally or in the cyst wall, and manifests as

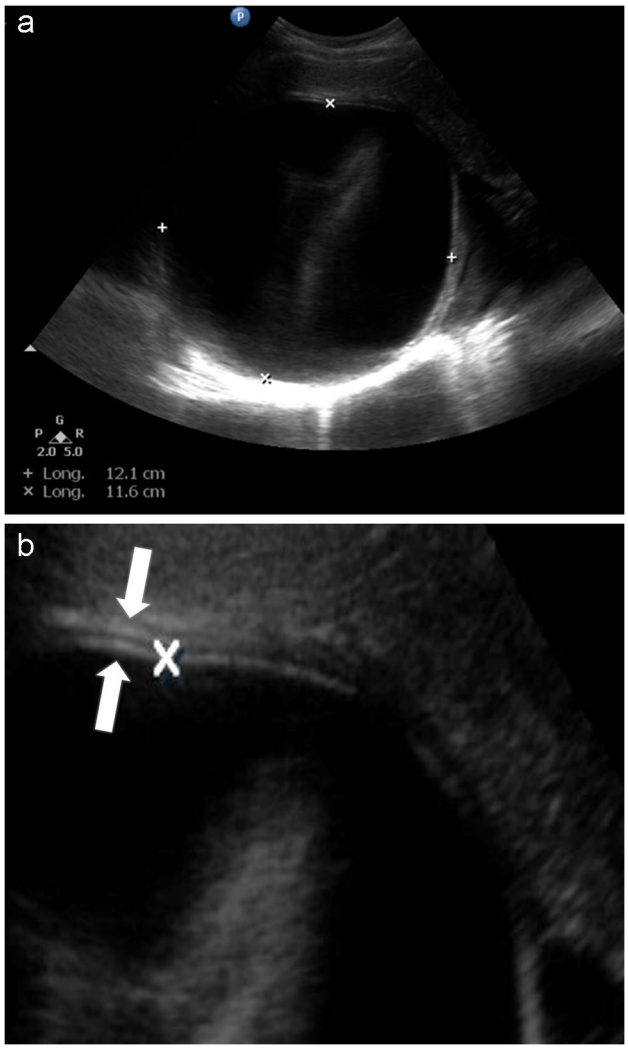


Fig. 1 Hepatic hydatid disease in an 11-year-old girl from an endemic region. **a** Transverse abdominal ultrasound image shows a rounded, well-defined, unilocular cystic lesion (between calipers), classified as CE1—group 1 of WHO classification, Gharbi I. **b** Magnified ultrasound image to emphasize the components of the cyst wall, seen as "the double-line sign" (*arrows*)

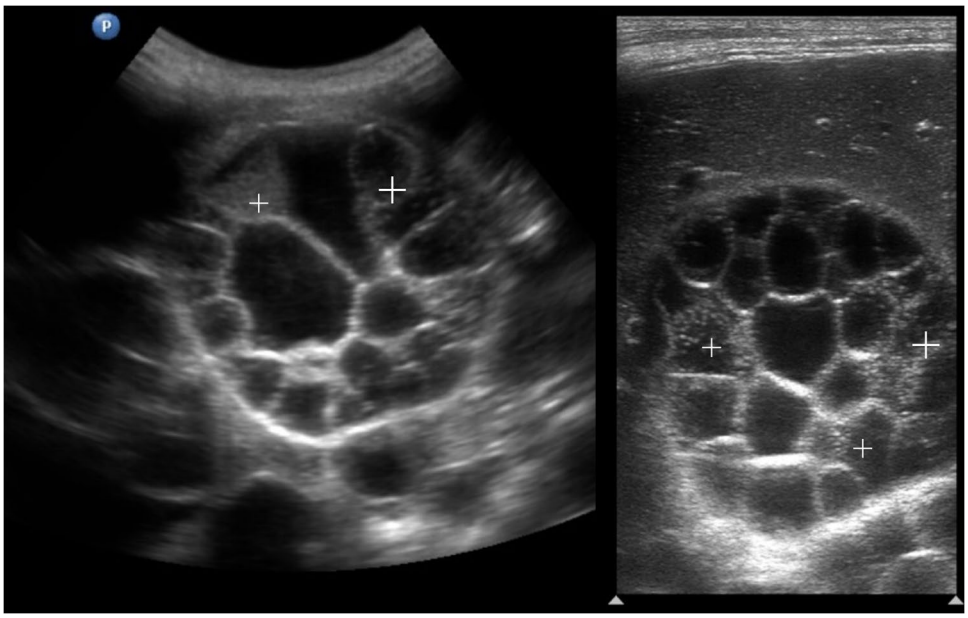


Fig. 2 Disseminated hydatid disease in a 12-year-old girl. US shows a multiseptated "honeycomb cyst" (CE2—group 1 of WHO classification, Gharbi III) in the right hepatic lobe, with some daughter cysts with hydatid sand (+)

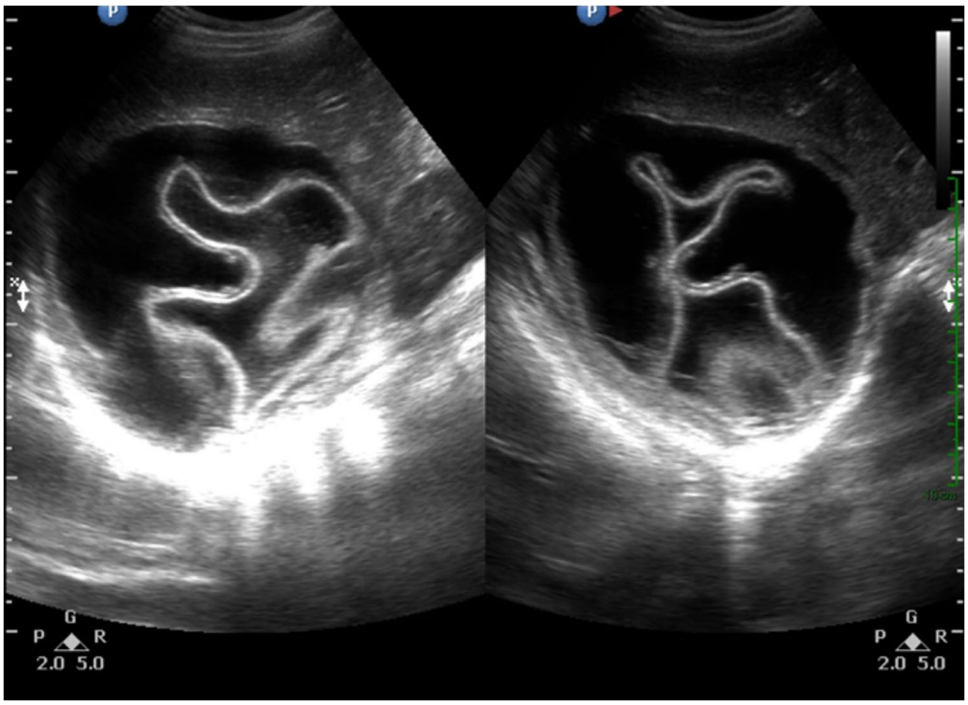


Fig. 3 Hepatic hydatid disease in a 7-year-old girl from an endemic region. Transverse and longitudinal ultrasound images show a cyst with detached membranes ("water-lily sign"), classified as CE3a—group 2 of WHO classification, Gharbi II

curvilinear or ringlike densities lying in the pericyst on radiographs. However, the presence of calcification does not necessarily indicate parasite death [3, 14].

Ultrasound is the most sensitive imaging modality for detecting purely cystic lesions showing the "double-line sign" (Fig. 1), daughter cysts, and vesicles, as well as floating membranes, septa, and hydatid sand. The honeycomb pattern (Fig. 2) of the multivesicular cysts is characterized by well-defined fluid collections with multiple septa (daughter cyst walls). The "wheel spoke" pattern features multiple daughter cysts separated by a mixed echogenicity material (hydatid matrix). The "water-lily sign" indicates complete membrane detachment within the cyst (Fig. 3) [7, 11]. In our experience, the detection of hydatid sand is a pathognomonic sign of the disease, representing sediment/debris resulting from daughter cyst rupture (Supplementary Material 2). Cyst calcification is demonstrated by US as hyperechoic areas with a strong posterior acoustic shadow [9].

CT imaging reveals purely cystic lesions as well-defined, round, or oval hypoattenuating masses (3–30 UH) (Fig. 4),

or multiseptated, "rosette-like/honeycomb" cysts (Fig. 5). When daughter cysts are present, these show lower attenuation values compared to the mother cysts, indicating viability. Detached membranes appear as isoattenuating serpentine structures within the cystic lesion (Figs. 6 and 7). Cyst calcification can be seen as round, hyperattenuating areas on CT [9].

MRI shows hydatid cysts with a typical low signal rim, on T2-weighted images. Daughter cysts and detached membranes have varying signal intensities; calcification is hypointense.

CT and MRI can reveal cyst wall defects and the passage of contents through them. Ruptures, especially those involving wide communication, are often detectable on US and CT. Biliary communication can be detected by US and MRI and all imaging modalities can identify trans-diaphragmatic migration (Fig. 8) [7]. Differential diagnosis includes simple epithelial cysts, which lack internal structures and polycystic disease, which may resemble multiple unilocular cysts [9].

Lungs

The lungs are the second most common site of infection with some authors indicating that the lungs are the most common site of infection in children [9, 16, 17, 19]. This is mainly attributed to the lungs' great elasticity and compressible nature, high vascularization, and negative pressure, which facilitate for cyst growth. Studies suggest that cysts in the lungs can grow three times faster than those in the liver [16, 19].

After traversing the hepatic filter, the larvae may reach the pulmonary circulation, where they can form hydatid cysts in the lungs (hematogenous dissemination). Lymphatic dissemination is also possible, bypassing the liver, via lymphatics of the small intestine to the chylous cistern, and through the thoracic duct to the venous circulation (jugular vein) [20].

Direct pulmonary exposure by inhaling air contaminated with *Echinococcus* eggs is also a risk [20]. In pediatric populations, these cysts represent a significant manifestation of hydatid disease [9, 16, 17, 19]. Statistically, pulmonary hydatid

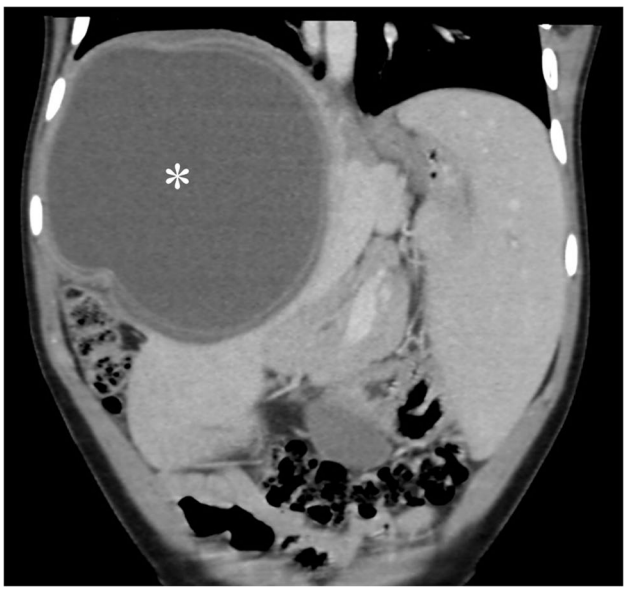


Fig. 4 Hepatic hydatid disease of the liver in an 11-year-old girl from an endemic region. Coronal contrast-enhanced CT image shows a typical unilocular hydatid cyst (20 UH), CE1—group 1 of WHO classification, Gharbi I, with an enhanced, noncalcified wall in the right hepatic lobe (*asterisk*)

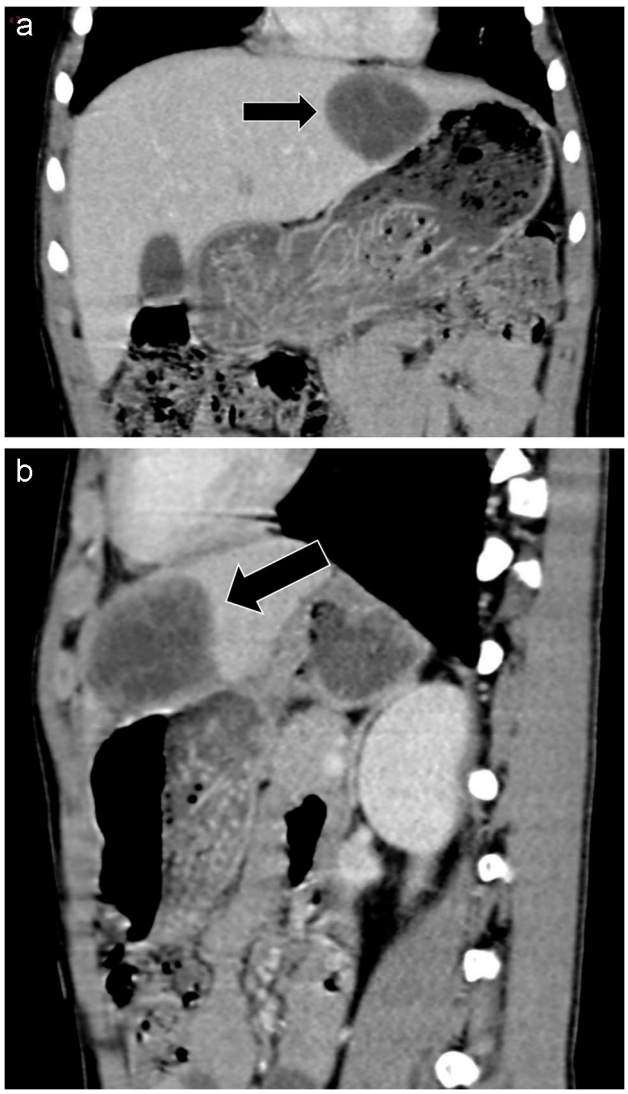


Fig. 5 Hepatic hydatic disease in a 13-year-old girl from an endemic region. **a** Coronal and (**b**) sagittal contrast-enhanced CT images show a multiloculated hydatid cyst (*arrows*) with mixed density and enhancing septations in the left hepatic lobe (CE2—group 1 of WHO classification, Gharbi III)

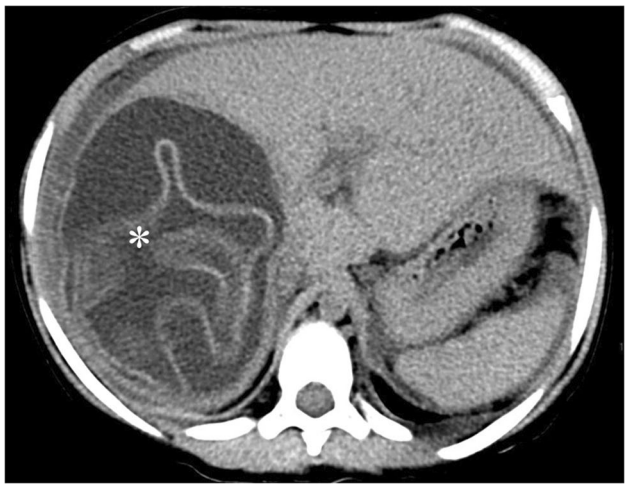


Fig. 6 Hepatic hydatid disease in a 7-year-old girl from an endemic region. Axial, contrast-enhanced CT image shows a hydatid cyst with enhancement of the detached laminated membrane (*asterisk*), classified as CE3a—group 2 of WHO classification, Gharbi II, with no communication to the biliary system or to adjacent structures (contained rupture)

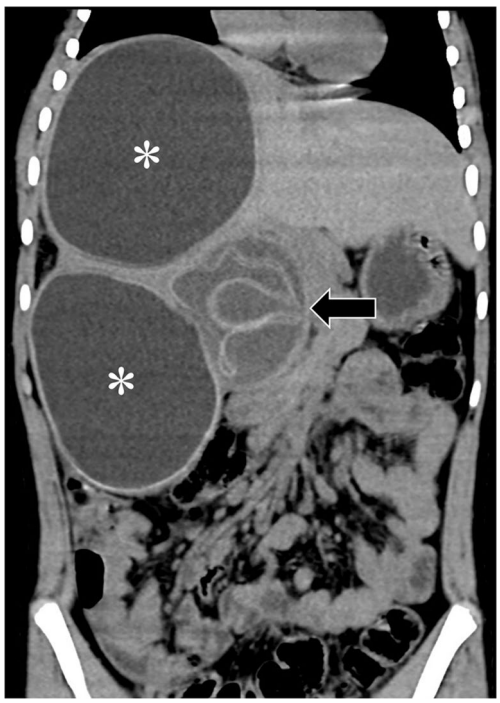


Fig. 7 Hepatic hydatid disease in a 12-year-old girl. Coronal contrast-enhanced CT image shows two unilocular hydatid cysts (*asterisks*) in the right hepatic lobe (CE1—group 1 of WHO classification, Gharbi I) as well as another hydatid cyst with detachment of the laminated membrane (CE3a—group 2 of WHO classification, Gharbi II) in the right lobe as well (*arrow*)

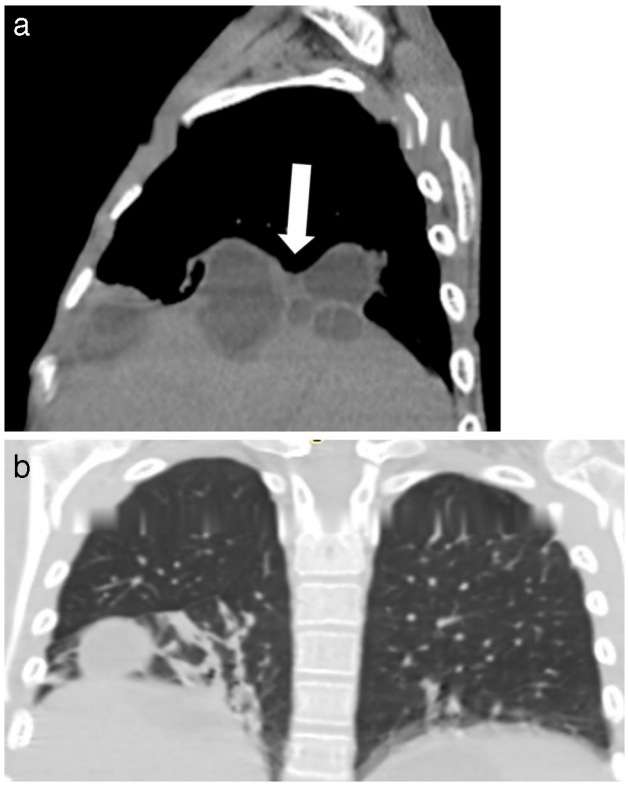


Fig. 8 Disseminated hydatid disease in a 12-year-old girl. **a** Sagittal and (**b**) coronal contrast-enhanced CT reformatted images show trans-diaphragmatic migration (*white arrow*)

cysts manifest as multiple cysts in 30% of cases, with 20% affecting both lungs, and 60% located in the lower lobes [3, 14].

Chest radiography effectively diagnoses uncomplicated hydatid disease, revealing well-defined, rounded opacities resembling "cannon balls" or "rugby balls" (Fig. 9). Calcification is extremely rare (less than 1%) [3, 14]. Cysts can sometimes exceed 20 cm in diameter, potentially displacing the mediastinum or diaphragm. Cysts located at the lung periphery are influenced by the pressure of surrounding tissues, while those situated in interlobar fissures may mimic pleural effusions. Enlarging cysts increase the risk of rupture, which can lead to lung collapse and/or might obscure the cyst borders (Fig. 10) [3, 14].

Ultrasound can be helpful in detecting peripherically located hydatid cysts along the chest wall, demonstrating the cyst wall as double echogenic lines separated by a hypoechoic layer ("double-line sign") in unilocular hydatid cysts and a double-layered septum in multivesicular ones, with a specificity of nearly 100%, aiding differentiation from simple cysts, cystic tumors, pseudocysts, or metastases [17].

CT demonstrates cystic lesions as well-defined, round, or oval hypoattenuating masses (3–30 UH) (Figs. 11 and 12).

Daughter cysts, if present, show lower attenuation values than that of mother cysts, indicating viability [8]. Cyst calcification can be seen as round, hyperattenuating areas on CT [9]. CT is the imaging modality of choice, mainly when the classical radiographic signs for complicated cysts are absent and an accurate diagnosis is required (Fig. 10). CT can show specific signs such as the "crescent sign" and "inverse crescent sign" (air infiltrating the posterior aspect of membranes without anterior extension), "air bubble sign" (small intracystic air foci between membranes), and "water-lily sign" or "camalote sign" (Figs. 9 and 13) [9, 21]. Complicated cases may show bronchial communication to the pericyst (Fig. 14), with air pockets or bubbles within the cyst [22].

MRI without contrast material can be compared to contrast-enhanced multidetector CT for detecting pulmonary hydatid disease, without using radiation. MRI has the advantage of being able to depict internal cyst membranes, thus providing additional diagnostic value [23].

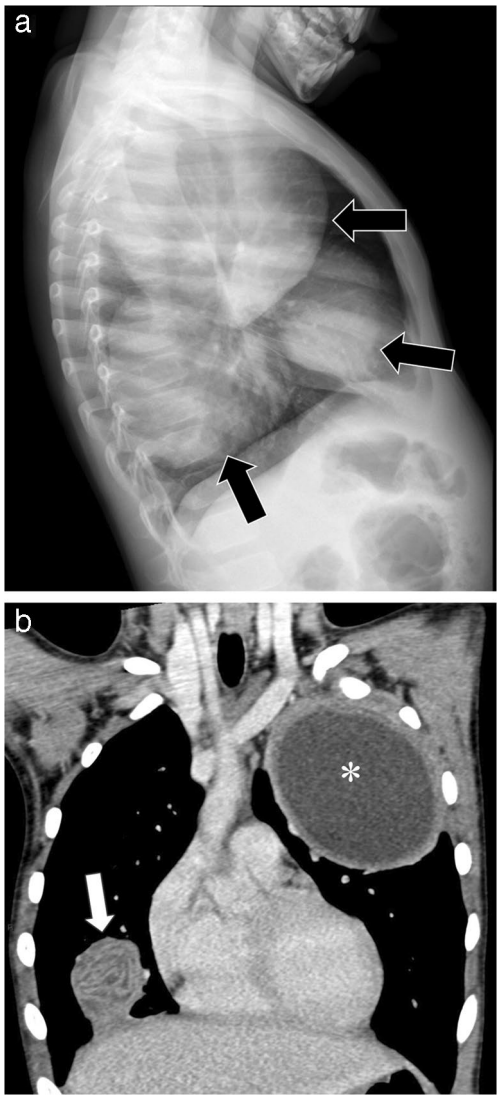


Fig. 9 Pulmonary hydatid in a 6-year-old boy from an endemic region. **a** Lateral chest radiograph shows several rounded opacities projecting over the upper and lower lung lobes in a "cannon ball pattern" (*black arrows*). **b** Coronal contrast-enhanced CT image shows two hydatid cysts. One in the left upper lobe (*asterisk*) with a thickened, enhancing wall, classified as CE1—group 1 of WHO classification, Gharbi I, and the other in the right lower lobe (*white arrow*) with detachment of the laminated membrane ("water-lily sign"), classified as CE3a—group 2 of WHO classification, Gharbi II, consistent with contained rupture

Brain

Cerebral involvement in hydatid disease is rare in children, but when it occurs, cyst growth is rapid due to the compressible nature of the brain tissue [3, 9]. In cerebral

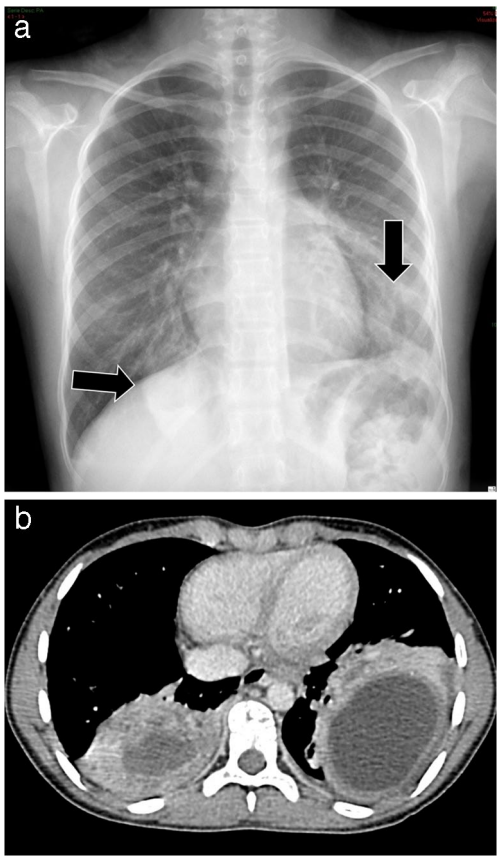


Fig. 10 Pulmonary hydatid disease in a 13-year-old girl from an endemic region. **a** Frontal chest radiograph shows two rounded opacities (*black arrows*): one projects over the right upper abdomen, and the second rounded opacity projects over the left lower hemithorax. **b** Axial contrast-enhanced CT image shows two hydatid cysts (*white arrows*) in the lower lobes; the right one reveals a thickened, enhancing, irregular wall, with heterogenous internal density consistent with contained rupture (CE4—group 3 of WHO classification, Gharbi IV). The left lesion shows a thickened enhancing wall (CE1—group 1 of WHO classification, Gharbi I), without signs of complication. Both lesions result in mass effect and compressive atelectasis

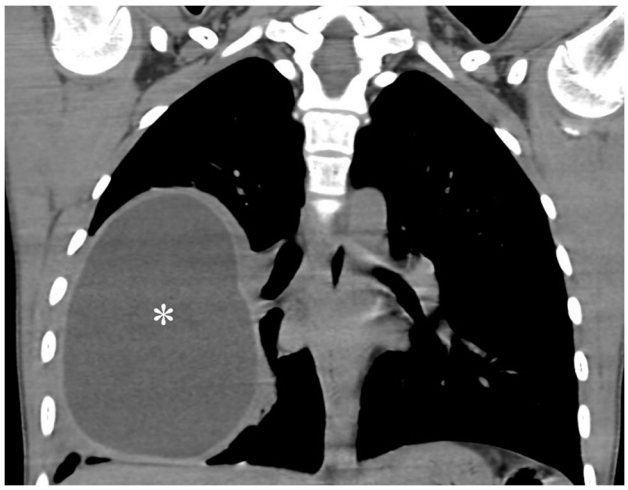


Fig. 11 Pulmonary hydatid disease in a 15-year-old girl from an endemic region. Coronal contrast-enhanced CT image shows a typical unilocular hydatid cyst (CE1—group 1 of WHO classification, Gharbi I) with enhancing walls in the right lower lobe (*asterisk*)

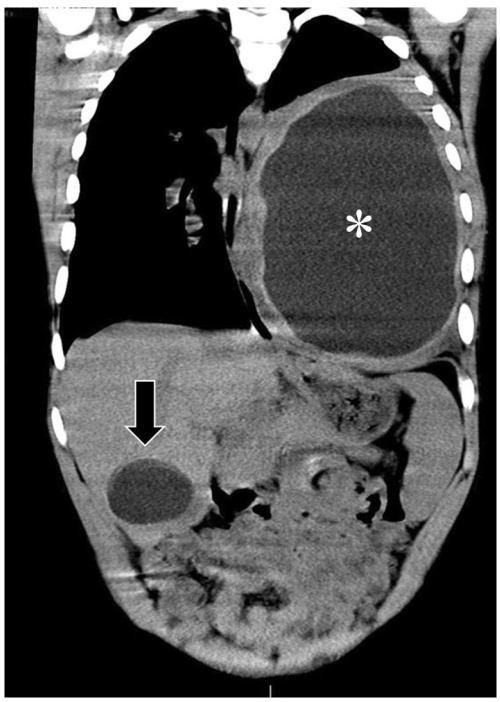


Fig. 12 Pulmonary and hepatic hydatid disease in a 15-year-old girl. Coronal contrast-enhanced CT image shows a typical unilocular hydatid cyst (CE1—group 1 of WHO classification, Gharbi I) with a noncalcified, enhancing wall, in the left lung (*asterisk*). An additional lesion is present in the right hepatic lobe (*arrow*)

hydatid disease, the cysts are typically solitary and predominantly located within the parietal lobes, corresponding to the territory supplied by the middle cerebral artery. Although this is the typical location, hydatid cysts can also

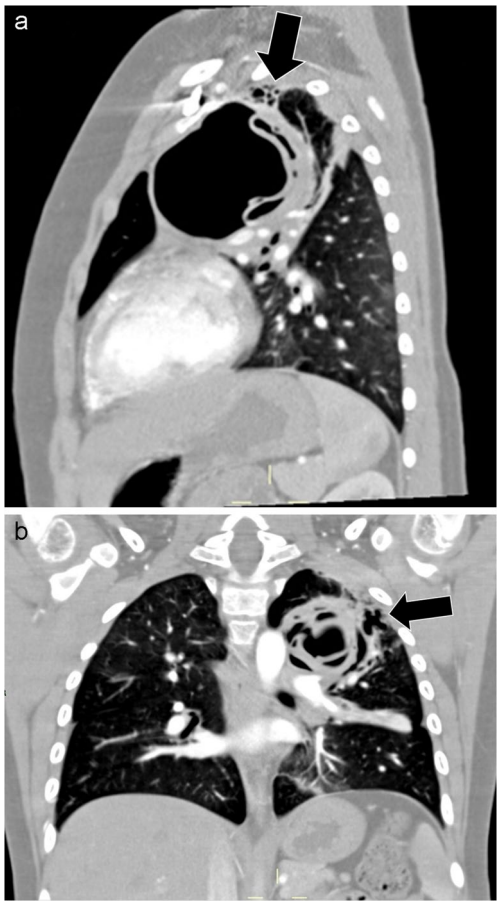


Fig. 13 Complicated pulmonary hydatid disease in an 8-year-old boy. **a** Sagittal and (**b**) coronal contrast-enhanced CT reformatted images show a hydatid cyst in the left upper lobe with a thickened wall and a detached laminated membrane (CE3a—group 2 of WHO classification, Gharbi II). There is direct rupture into the adjacent pleura (*arrow*)

affect the infratentorial region and even the intraventricular spaces [24]. Multiple cerebral cysts often result from spontaneous rupture, trauma, or iatrogenic causes during surgery, with an intraoperative rupture rate exceeding 25% [14, 15, 24].

Skull radiographs are generally insensitive and less preferred as an imaging modality, but may reveal bone erosion, particularly of the posterior clinoid processes. In younger patients, unilateral skull enlargement and suture diastasis may also be evident.

CT typically shows well-demarcated, round, or oval lesions with cerebrospinal fluid density [3]. Although rim enhancement is rare, intravenous contrast administration can slightly increase the cyst fluid's attenuation. Unlike abscesses or neoplastic cystic lesions, cerebral hydatid cysts characteristically lack perilesional edema; however, the significant mass effect can lead to hydrocephalus. The occurrence of multivesicular cysts in the brain is relatively uncommon [3, 14].

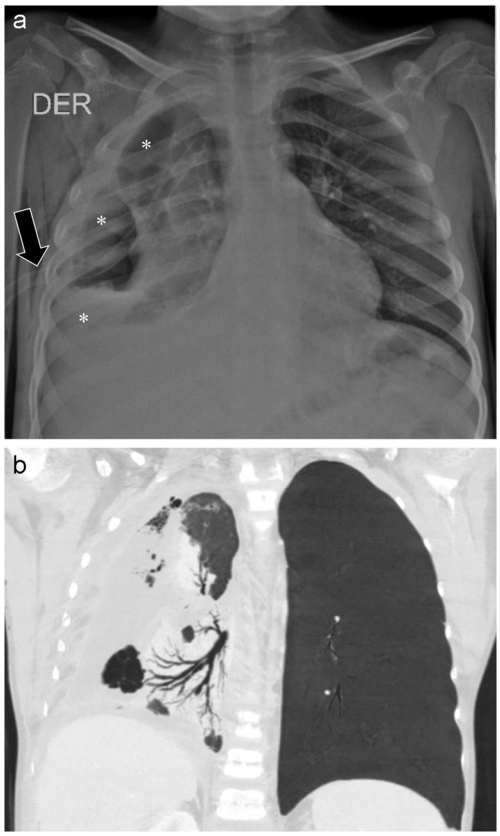


Fig. 14 A 9-year-old boy presented with fever and cough was misdiagnosed and treated for complicated pneumonia with pulmonary abscess and right empyema. **a** Chest radiograph reveals a right hydropneumothorax (*asterisks*) with a chest tube (*black arrow*) in place. **b** Coronal CT reformatted image demonstrates a complicated hydatid cyst connected to bronchi (*white arrow*), which was confirmed during the surgical procedure

MRI is the imaging modality of choice for brain hydatid cysts, which tend to be unilocular with cerebrospinal fluid signal. Secondary mass effect typically occurs, leading to hydrocephalus [25]. A key feature distinguishing cerebral hydatid cysts from abscesses and other cystic masses is the lack of significant surrounding edema. In T2-weighted images, a hypointense rim along the walls of the cyst is a classic sign (Fig. 15) [27]. Complications, such as superinfection and rupture, may present with rim enhancement and significant surrounding edema. Differential diagnosis includes arachnoid cyst, epidermoid cyst, porencephalic cyst, neurocysticercosis, other infectious etiologies, and cystic malignancies [25].

Heart

Cardiac involvement in hydatid disease is rare in children [26, 27]. It typically arises from one of two pathways: via the coronary arteries, leading to the direct invasion of the myocardium, or through the pulmonary veins following the rupture of pulmonary echinococcal cysts into the venous system [23]. Cysts in the myocardium are most commonly found in the left ventricular wall, accounting for 60% of cases, and within the interventricular septum in approximately 7% of the cases. Often, cardiac hydatid disease may be asymptomatic or present with vague symptoms such as chest pain, exertional dyspnea, and fever [3]. When a cyst ruptures into the heart chambers, it most frequently occurs in the right ventricle, causing severe reactions such as anaphylaxis and resulting in the embolization of daughter cysts to the lungs or systemic circulation [3]. Moreover, the heart may suffer secondary effects due to proximity to infected organs, such as the liver or lungs, when these primary sites harbor hydatid cysts [23].

Radiography may show localized deformity (bulge) in the cardiac silhouette, and changes of shape and size on serial exams. Calcifications may be seen.

Angiography demonstrates abnormal pulsation, resembling a ventricular aneurysm with interventricular septal thickening or localized bulges into the cardiac chambers [3, 26].

Echocardiography is highly sensitive in detecting intracardiac hydatid cysts. Echocardiography is noninvasive, easily performed, and widely available, which makes it the first-line imaging modality [26]. It can show characteristic multiloculated cystic lesions and the presence of daughter cysts. Hydatidosis must always be considered when an intracardiac cystic structure is detected [27].

CT and MRI provide better visualization of cardiac involvement and assess for possible extension beyond the heart (Fig. 16). CT has the advantage of detecting wall calcification, while MRI depicts the internal morphology of the cyst. Both modalities can help to rule out arterial and pericardiac extension.

Spleen

Primary splenic involvement is quite rare [8–10, 13]. Splenic hydatid disease is typically secondary, resulting from systemic dissemination or intraperitoneal spread from a ruptured hepatic hydatid cyst [28]. Splenic hydatid cysts are usually solitary, with imaging features similar to those of hepatic hydatid cysts, ultrasound being the most sensitive imaging modality (Fig. 17) [6, 13].

However, the diagnosis can be challenging, as the cyst may present as a simple cyst without typical serologic and imaging findings. Differentiating it from other splenic cystic lesions, such as epidermoid cysts, pseudocysts, abscesses, and cystic neoplasms, can be difficult [10, 13, 28, 29]. Diagnostic accuracy improves when daughter cysts and detached floating membranes are observed [28]. CT is the most useful imaging modality for

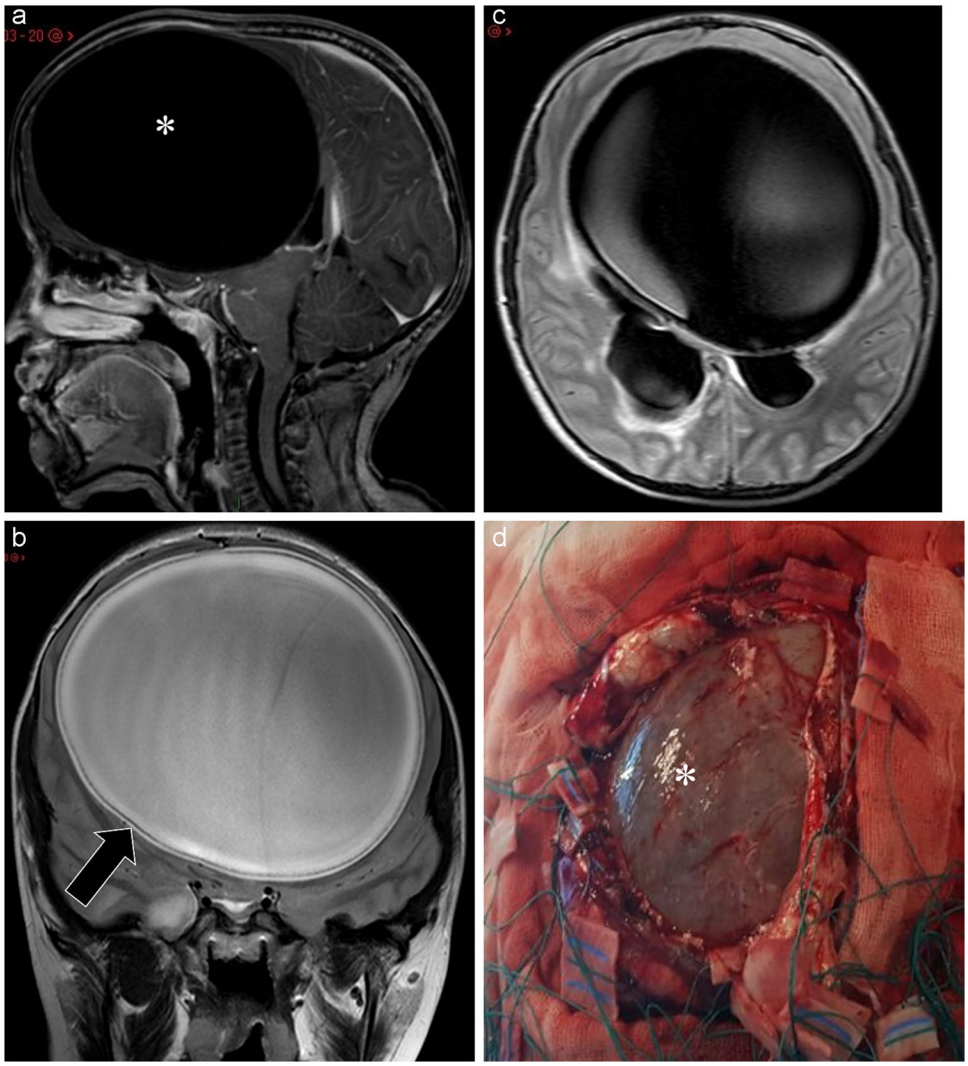


Fig. 15 Cerebral hydatid disease in a 13-year-old girl. **a** Sagittal T1-weighted MR image shows a rounded cystic structure (CE1—group 1 of WHO classification, Gharbi I) with cerebrospinal fluid signal (*asterisk*). **b** Coronal T2-weighted MR image reveals a hypointense rim (*arrow*) delineating the hydatid cyst. **c** Axial FLAIR-weighted MR image. **d** Intraoperative image of the hydatid cyst (*asterisk*)

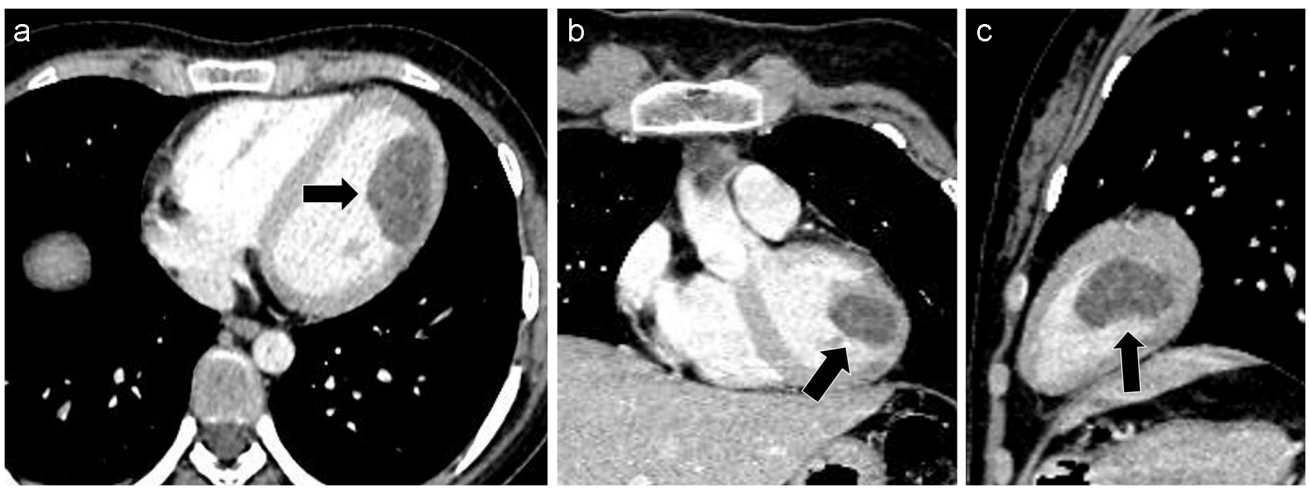


Fig. 16 Cardiac hydatid disease in a 14-year-old girl from an endemic region. **a** Axial contrast-enhanced CT image, (**b**) coronal CT reformatted image, and (**c**) sagittal CT reformatted images show a multiseptated hydatid cyst (CE2—group 1 of WHO classification, Gharbi III) with mixed density and enhancing septations in the left ventricular free wall (*arrow*)

visualizing the cyst walls, determining whether or not calcifications are present, and identifying daughter vesicles [7].

Others

Hydatid cysts can develop in various body locations including atypical sites such as the kidney, bone, muscle, pancreas, diaphragm, adrenal gland, ovary, and peritoneum (Fig. 18) [3, 9, 10]. These cysts may exhibit features similar to typical cysts, potentially leading to diagnostic confusion. Characteristic imaging findings include ring or curvilinear calcifications (a hallmark feature) visible on plain radiographs, as well as the presence of daughter cysts and membrane detachment, which are more easily identifiable on US or MRI scans [3, 10].

Renal hydatid cysts are often unilateral and usually located in polar regions. These cysts can be either unilocular or multiloculated with daughter cysts. In bones, hydatid cysts predominantly occur in vascularized areas, frequently affecting the spine and pelvis. They manifest as well-defined, multiloculated, osteolytic, expansile lesions that extend into adjacent soft tissues and can lead to pathologic fractures or spinal cord compression. In muscle tissue, the imaging presentation is variable and nonspecific, ranging from unilocular or multilocular cysts to complex solid lesions. Hydatid cysts in the pancreas may mimic pseudocysts; however, features such as a thick, laminated wall with a thin layer of calcification can suggest a hydatid cyst, especially if associated with hepatic hydatidosis. When the diaphragm is involved, it appears thickened and lobulated, with either unilocular or multilocular cysts that may split the leaves of the diaphragm [10]. Adrenal gland hydatidosis initially presents as simple cysts that may evolve over time to develop daughter cysts, floating membranes, or calcifications. Ovarian hydatid cysts are important considerations in the differential diagnosis of cystic pelvic masses, presenting as unilocular or multiloculated cysts with daughter cysts. These non-hepatic manifestations of hydatid disease are generally secondary in nature [3, 9, 10]. Peritoneal hydatid disease often results from the rupture of a primary hepatic cyst and can manifest as any type of hydatid cyst within the cavity, frequently appearing as multiple cysts. When single and unilocular, these cysts may be indistinguishable from mesenteric or duplication cysts [9, 10].

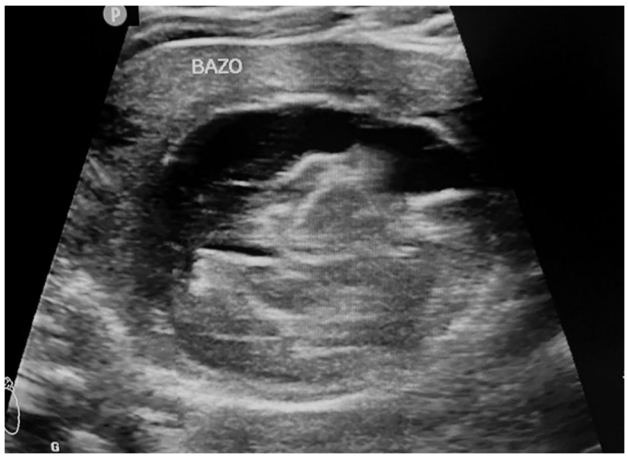


Fig. 17 Splenic hydatid disease of the spleen in a 7-year-old boy from an endemic region in Argentina. Transverse ultrasound image shows a cyst (*asterisk*) with detached membranes ("water-lily sign"), classified as CE3a—group 2 of WHO classification, Gharbi II

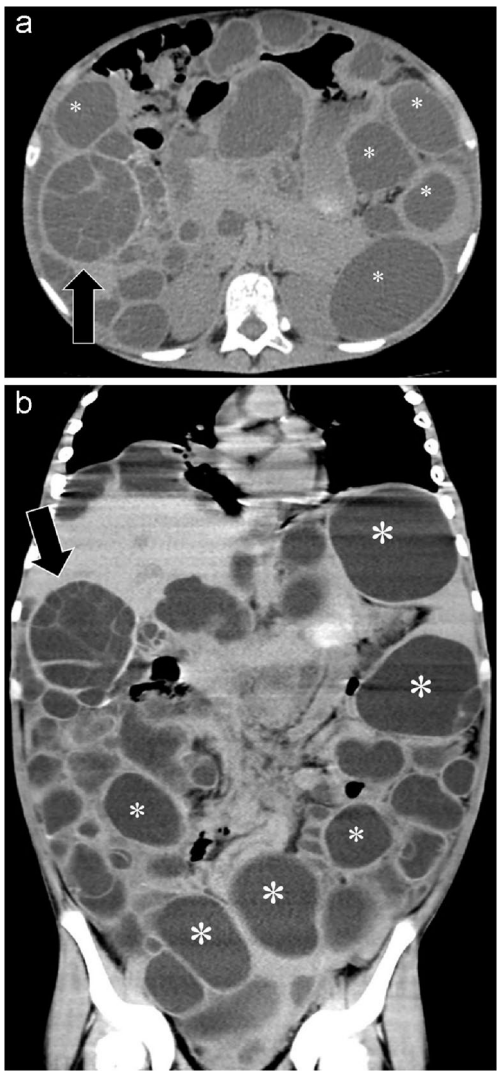


Fig. 18 Disseminated hydatid disease in a 12-year-old girl. **a** Axial and (**b**) coronal contrast-enhanced CT reformatted images show multiple intraperitoneal unilocular hydatid cysts (*asterisks*), classified as CE1—group 1 of WHO classification, Gharbi I, with variable wall thicknesses, due to peritoneal seeding. There is one multiseptated "honeycomb cyst," classified as CE2—group 1 of WHO classification, Gharbi III, in the right hepatic lobe (*black arrow*)

Role of interventional radiology

Interventional radiology plays an important role in the management of hydatid disease, using minimally invasive techniques, particularly in cases involving the liver classified as group 1 and group 2 (WHO classification). Interventional radiologists can perform procedures that effectively reduce cyst size while minimizing the risk of rupture and infection.

One common technique employed is known as PAIR (puncture, aspiration, injection, and reaspiration), which involves puncturing the cyst, aspirating its contents, injecting a scolicidal agent, and then reaspirating any remaining fluid. PAIR is typically utilized for unilocular cysts (CE1) and cysts with detached membranes (CE2a). For multiseptated cysts (CE2) and cysts containing daughter cysts in a solid matrix (CE3b), a modified catheterization technique (MoCaT) can be used. This technique allows for the placement of a catheter into the cyst under imaging guidance, enabling the injection of scolicidal agents and the aspiration of cyst contents [30, 31].

Overall, interventional radiology is essential in the management of hydatid disease, offering minimally invasive techniques to reduce cyst size, prevent complications, and improve patient outcomes [30].

Conclusion

In summary, the diagnostic approach to hydatid disease relies heavily on identifying distinct imaging features that indicate the presence of the condition across various anatomical sites. It is essential to recognize that complex presentations of hydatid cysts can manifest with atypical findings on ultrasonography, computed tomography, and magnetic resonance imaging, particularly in patients from endemic areas. Such recognition is essential to avert severe and potentially fatal complications as well as to prevent disease recurrence. Therefore, in regions where hydatid disease is prevalent, an in-depth understanding of the non-standard radiological signatures associated with complicated cases of hydatid disease is invaluable for ensuring accurate diagnosis and appropriate therapeutic strategies. Clinicians should maintain a high index of suspicion and include hydatidosis in the differential diagnosis when encountering unusual cystic lesions, especially in endemic zones.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s00247-024-06080-7.

Acknowledgements Augustina Martinez from Hospital Schestakow, San Rafael, Mendoza, Argentina, who gave us a beautiful case to depict splenic involvement and Víctor Jesús Trigos Alvarado from Instituto Nacional de Salud del Niño San Borja, Lima, Peru, who designed the visual abstract.

Declarations

Conflicts of interest None

References

1. Salazar-Mesones B, Luna-Vílchez M, Maquera-Afaray J et al (2022) Clinical and epidemiological characteristics of cystic echinococcosis in children from a tertiary center in Peru. Rev Peru Med Exp Salud Publica 39:65–69
2. CDC (2024) DPDx - Case studies- echinococcosis. [cited 2023 november 2023]. Available from: https://www.cdc.gov/dpdx/echinococcosis/index.html
3. Beggs I (1985) The radiology of hydatid disease. AJR Am J Roentgenol 145:639–648
4. Anon P (2022) Equinococosis: informe epidemiológico en la región de América del Sur - 2019–2021, n.5, 2022
5. Pavletic CF, Larrieu E, Guarnera EA et al (2017) Cystic echinococcosis in South America: a call for action. Rev Panam Salud Publica 41:e42
6. Cağlayan K, Celik A, Koç A et al (2010) Unusual locations of hydatid disease: diagnostic and surgical management of a case series. Surg Infect (Larchmt) 11:349–353
7. Pedrosa I, Saíz A, Arrazola J et al (2000) Hydatid disease: radiologic and pathologic features and complications. Radiographics 20:795–817
8. Berberian G, Rosanova T, Inda L et al (2017) Echinococcosis in children: experience in a tertiary care hospital outside the endemic area. Arch Argent Pediatr 115:282–286
9. Yuksel M, Demirpolat G, Sever A et al (2007) Hydatid disease involving some rare locations in the body: a pictorial essay. Korean J Radiol 8:531–540
10. Ilica AT, Kocaoglu M, Zeybek N et al (2007) Extrahepatic abdominal hydatid disease caused by Echinococcus granulosus: imaging findings. AJR Am J Roentgenol 189:337–343
11. Gharbi HA, Hassine W, Brauner MW, Dupuch K (1981) Ultrasound examination of the hydatic liver. Radiology 139:459–463
12. WHO Informal Working Group (2003) International classification of ultrasound images in cystic echinococcosis for application in clinical and field epidemiological settings. Acta Trop 85:253–261
13. Lewall DB (1998) Hydatid disease: biology, pathology, imaging and classification. Clin Radiol 53:863–874
14. Turgut AT, Altin L, Topçu S et al (2007) Unusual imaging characteristics of complicated hydatid disease. Eur J Radiol 63:84–93
15. von Sinner W, te Strake L, Clark D, Sharif H (1991) MR imaging in hydatid disease. AJR Am J Roentgenol 157:741–745
16. Garg MK, Sharma M, Gulati A et al (2016) Imaging in pulmonary hydatid cysts. World J Radiol 8:581–587
17. Pradhan A, Mallick B, Dash A, Nanda D (2022) Clinical presentation and outcome of children with hydatid disease: a retrospective cross-sectional study from a tertiary care hospital in eastern India. J Parasit Dis 46:230–235
18. Alghofaily KA, Saeedan MB, Aljohani IM et al (2017) Hepatic hydatid disease complications: review of imaging findings and clinical implications. Abdom Radiol (NY) 42:199–210
19. Vantankhah A, Taqanaki PB, Rahmanian M et al (2023) A life-threatening encounter: an uncommon case of ruptured hydatid cyst presenting as anaphylactic shock and respiratory distress in a 12-year-old boy. Ann Med Surg (Lond) 85:5742–5747
20. Sarkar M, Pathania R, Jhobta A et al (2016) Cystic pulmonary hydatidosis. Lung India 33:179–191
21. Durhan G, Tan AA, Düzgün SA et al (2020) Radiological manifestations of thoracic hydatid cysts: pulmonary and extrapulmonary findings. Insights Imaging 11:116

22. Dilek E, Kemal Ö, Necdet P, Hasan Emin K (2015) Radiological characteristics of pulmonary hydatid cysts. In: Alfonso JR-M (ed) Current topics in echinococcosis. Rijeka, IntechOpen, p 8
23. Sodhi KS, Bhatia A, Samujh R et al (2019) Prospective comparison of MRI and contrast-enhanced MDCT for evaluation of pediatric pulmonary hydatid disease: added diagnostic value of MRI. AJR Am J Roentgenol 212:982–987
24. Dursun M, Terzibasioglu E, Yilmaz R et al (2008) Cardiac hydatid disease: CT and MRI findings. AJR Am J Roentgenol 190:226–232
25. Abbasi B, Akhavan R, Ghamari Khameneh A et al (2021) Computed tomography and magnetic resonance imaging of hydatid disease: a pictorial review of uncommon imaging presentations. Heliyon 7:e07086
26. Fiengo L, Bucci F, Giannotti D et al (2014) Giant cardiac hydatid cyst in children: case report and review of the literature. Clin Med Insights Case Rep 7:111–116
27. Maroto LC, Carrascal Y, López MJ et al (1998) Hydatid cyst of the interventricular septum in a 3.5-year-old child. Ann Thorac Surg 66:2110–2111
28. Malik A, Chandra R, Prasad R et al (2016) Imaging appearances of atypical hydatid cysts. Indian J Radiol Imaging 26:33–39
29. Rasheed K, Zargar SA, Telwani AA (2013) Hydatid cyst of spleen: a diagnostic challenge. N Am J Med Sci 5:10–20
30. Akhan O (2023) Percutaneous treatment of liver hydatid cysts: to PAIR or not to PAIR. Curr Opin Infect Dis 36:308–317
31. Khuroo MS, Wani NA, Javid G et al (1997) Percutaneous drainage compared with surgery for hepatic hydatid cysts. N Engl J Med 337:881–887