Hydatidosis: Clinical-imaging classification according to Gharbi and the World Health Organization

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Abstract
Hydatidosis is a zoonosis produced by Echinococcus granulosus, being endemic and having a higher prevalence in rural zones. The clinical manifestations of this disease depend on the organ affected and on the parasite growth rate. The most frequent location is the liver parenchyma, manifesting as an incidental finding when it is asymptomatic, or with symptoms such as biliary colic. The Pan American Health Organization (PAHO) and the World Health Organization (WHO) support the Gharbi and the WHO classifications, which allow estimating the stage of hydatid cysts. We propose the study and estadification of hydatic cysts lesions by ultrasonography,computed tomography and magnetic resonance since they are the most important tool for its caracterization.

Keywords
Echinococcosis. Ultrasonography. Computed tomography. Magnetic resonance imaging. Liver.

Introduction

Hydatidosis is caused by Echinococcus granulosus, and humans become intermediate hosts after accidental ingestion of parasite eggs (Fig. 1)[1,2].
The risk factors for infection include the lack of drinking water, having a family member with hydatidosis and contact with dogs.[1]
Argentina has a high incidence of hydatidosis, mainly in the Patagonia region (Chubut and Neuquén), with the highest rates of new cases, the Northwest region (Catamarca, Santiago del Estero and Salta) and the east of the province of Entre Ríos.[1]
The aim of this pictorial essay is to review the most appropriate current classification systems for hydatidosis based on clinical and imaging findings.

Signs and symptoms

In 90% of cases, the parasitic infection is located in the liver parenchyma, followed by the lung parenchyma in order of frequency. Less common sites include, but are not limited to, the spleen, kidneys, brain, mediastinum and peritoneal cavity.[1,5,11]
This infection is usually asymptomatic and incidentally diagnosed. Signs and symptoms may not manifest for years and will depend on the expansive nature of the organ involved.
Clinical manifestations of hydatid cysts may include abdominal pain, fever, palpable mass, nausea, vomiting or non-specific gastrointestinal disorders, even in non-complicated cases.
Complications may include cyst rupture, infection, transdiaphragmatic thoracic involvement, cyst growth beyond the liver, peritoneal seeding, perforation into a hollow viscera, portal vein involvement and invasion of the thoraco-abdominal wall.[3,9]
Cyst rupture is a complication of the natural course of the disease in 50-90% of cases that also may occur by trauma. There are three different types of rupture:

- Contained rupture: this is the natural course of the cyst (type 2 cyst according to the World Health Organization [WHO] classification), and it occurs when the endocyst ruptures but the pericyst remains intact.
- Communicating rupture: the hydatid cyst ruptures into the biliary ducts, manifesting as obstructive jaundice, with or without cholangitis.

• Direct rupture: both the endocyst and the pericyst rupture, allowing passage of the cyst contents into the abdominal cavity and manifesting as acute abdominal pain or anaphylaxis.

Cyst infection occurs in 25% of cases after direct or communicating rupture of the cyst, manifesting as fever and malaise.[2,5,9]

Lung involvement most frequently occurs in the lower segments of the right lung, with cysts ranging in size from 1 to 20 cm in diameter because of the compressibility of the lungs, with a high prevalence during childhood. These cysts remain asymptomatic for a long period of time because of the low lung tissue resistance to cyst growth in this stage, and are incidentally diagnosed in adulthood.

It should be noted that, when hydatid cysts are located in the lung, formation of daughter vesicles and calcifications are rare (0.7%).[9]

During its natural course, the pericyst may erode the walls of the bronchioles, which determined the presence of air into the cyst, showing the following signs:

- Air introduced between the pericyst and the ectocyst:
• Crescent sign or meniscus sign: thin air collection in the upper part of the cyst.
• Inverse crescent sign: air collection at the posterior aspect of the cyst.
• Signet ring sign: focal air accumulation.
- Onion peel appearance: complete separation of the pericyst and the ectocyst, allowing the passage of air into the endocyst and generating air-fluid levels.
- Serpent sign: partial expectoration of the cyst content, leading to the collapse of membranes within the cyst.
- Water-lily sign or camalote sign: the endocyst has completely collapsed and floats freely in the cyst fluid.
- Mass within the cyst: when the fluid is entirely evacuated, the remaining solid component falls to the dependent part of the cyst.

Diagnosis

The diagnosis is based in physical examination, epidemiological history, imaging and serology.

Any asymptomatic person with a cystic mass located in different organs and associated with epidemiological aspects

WHO Classification	Gharbi's Classification
Type CL	----------
Type CE1	Type I
Type CE2	Type III
Type CE3	Type II
Type CE4	Type IV
Type CE5	Type V

Table 1: Comparison of both ultrasound-based classifications.

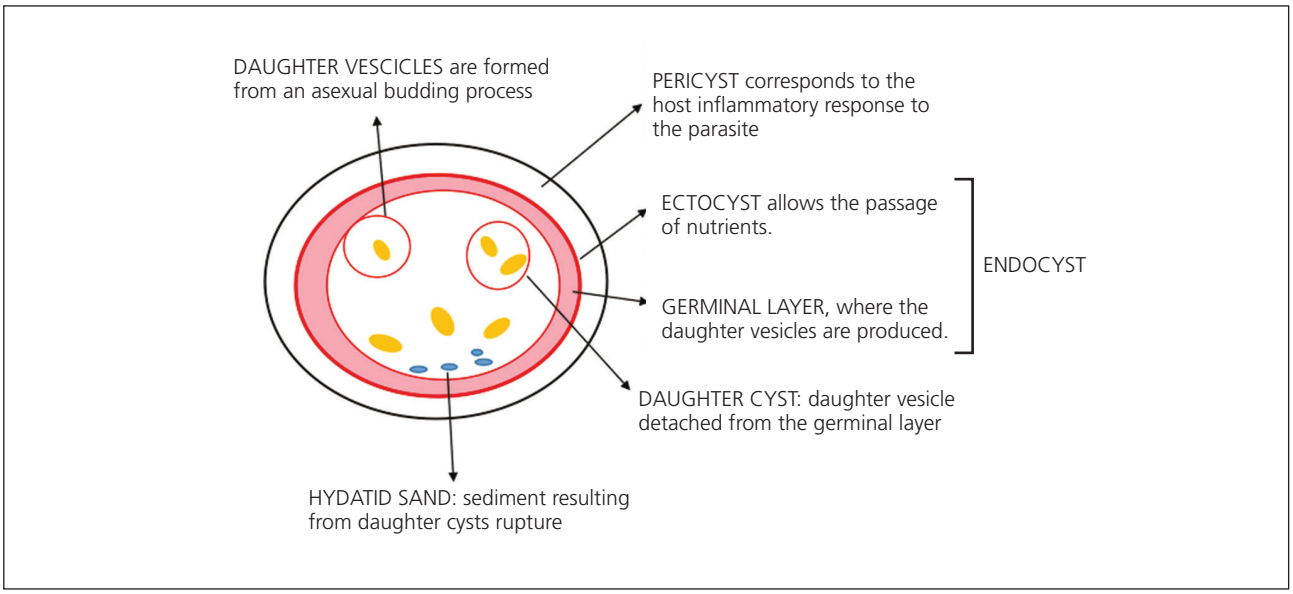

Fig. 1 *Hydatid cyst structure*

of the disease is considered as a suspected case. In this setting, radiologists play an important role, since one of the approaches to confirm diagnosis is through radiography, ultrasound, computed tomography (CT) or magnetic resonance imaging (MRI).

It should be noted that ultrasound is the gold standard method for the diagnosis of abdominal hydatid cysts (most commonly located in the liver), since it has 100% sensitivity and 97% specificity. Furthermore, it is a low-cost method with higher accessibility that allows characterization of the cyst and determination of its stage and follow-up of the course of disease in patients on medical treatment, while CT and

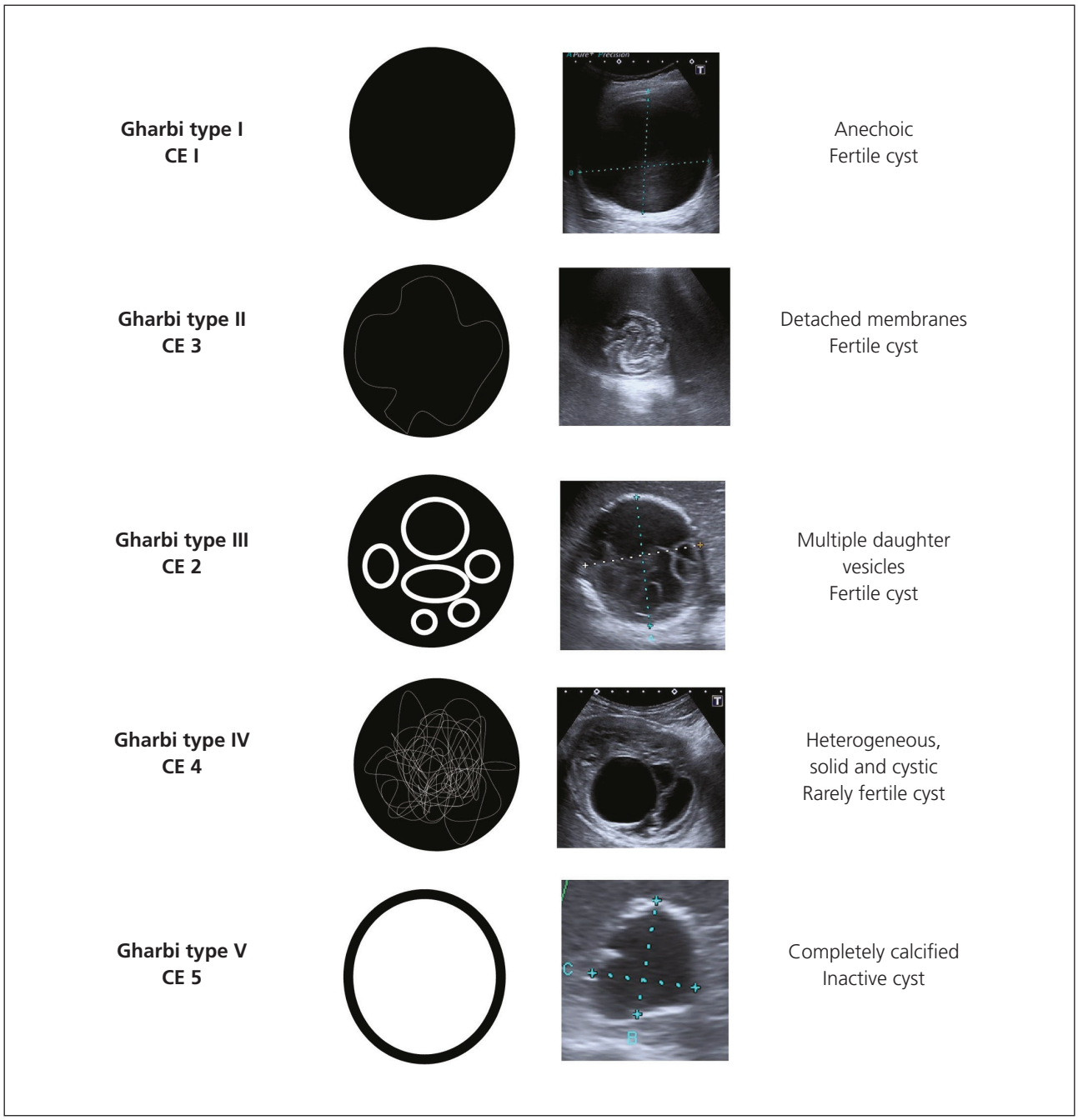

Fig. 2 Ultrasound-based classification of hydatid cysts.

MRI may be reserved for cases of complications and of hydatid cysts located in sites outside the abdominal cavity (lung, brain, bone, etc.).[2,7]

The Pan American Health Organization (PAHO) and the WHO support Gharbi's and the WHO classifications for diagnostic imaging, which include the following types (Tables 1 and 2, and Fig. 2)[1,2,9-11]:
- CL: cyst of nonspecific characteristics. Unilocular with homogeneous anechoic fluid content and non visible wall. This type represents a hydatid cyst of early diagnosis and cannot be differentiated from the simple cyst. It is a fertile cyst. There are no pathognomonic sonographic signs; therefore, diagnosis is made by other complementary techniques. Twenty-five percent of hydatid cysts have this appearance.
- Type I (CE1): hyaline cyst with anechoic fluid content, with laminar membrane clearly visible, with or without snowflake sign. Fertile cyst (Figs. 3 and 4).
- Type II (CE3): hyaline anechoic cyst with "detached" or "folded" laminar membrane; serpent sign. Usually fertile cyst. (Figs. 5 and 6).
- Type III (CE2): multivesicular; multiple cystic images within a cyst (wheel-spoke or honeycomb pattern). Usually fertile cyst. (Figs. 7 and 8).
- Type IV (CE4): heterogeneous lesion with mainly solid content, with or without partial calcifications. Rarely fertile cyst (Figs. 9 and 10).
- Type V (CE5). Completely calcified lesion. Inactive cyst (Figs. 11 to 13).

When calcifications occur, they are located in the cyst wall, appearing in curved or ring shapes in the pericystic layer. They can be seen in 20-30% of cases. Complete calcification of the hydatid cyst is of paramount importance because it indicates the death of the parasite, with the cyst becoming clinically inactive.

CT has a high sensitivity for detecting calcium and the content of hydatid cyst, as well as signs of peritoneal seeding and superinfection, with imaging findings of infection being similar to those in other hepatic abscesses. The most representative findings are[4-9]:
- Solid or mixed (solid-cystic) appearance.
- Intravenous contrast-enhanced CT: hyperdense ring surrounding the abscess, associated with patchy areas of contrast enhancement due to inflammatory changes.
- Poorly defined margins.
- Air-fluid or fluid-fluid levels (the presence of gas does not necessarily imply infection when located in the lungs or hollow viscera in the case of direct rupture).

On MRI, the signet ring sign is visualized on T2-weighted images. This sign is characterized as a ring of low intensity signal peripheral to the cyst, representing the pericystic collagen. However, this finding has also been reported in other conditions, such as hepatocellular carcinoma, amebic abscesses and hematomas. MRI also allows characterization of the cyst content.[8-11]

Despite the high diagnostic sensitivity and specificity of serology (Western blot and ELISA), false negative results may be seen in liver cysts or lung cysts in patients with no release of immunogens into the bloodstream or presence of antigen-antibody complexes. For this reason, radiologists play an essential role in the detection of these cysts.[1,2,8]

Conclusions

At present, Gharbi's and the WHO classification are very helpful for clinical and imaging correlation, allowing proper characterization of hydatid cysts by the various imaging methods and correlation with their clinical stage. Abdominal ultrasound

Table 2: Description by computed tomography and magnetic resonance imaging according to the type of hydatid cyst

	Computed Tomography	Magnetic Resonance Imaging
Gharbi type I-CE1	They are well-defined and show fluid density (0-20 Hounsfield Units)	T1: Low intensity signal T2: - High intensity signal - Signet ring sign (ring of low intensity signal peripheral to the cyst)
Gharbi type II-CE3	Cysts with fluid density; hyperdense detached membranes may be seen	Detached membranes within the cyst of low intensity signal in all sequences
Gharbi type III-CE2	Greater attenuation in the mother cysts in relation to the daughter cysts	T1 and T2: daughter cysts are iso- and hypo-intense with respect to the matrix
Gharbi type IV-CE4	Round masses of high density, patchy calcifications and occasional daughter vesicles	Heterogeneous masses, with calcified areas that have low intensity signal (hypointense)
Gharbi type V-CE5	Cysts with high density, completely calcified	T1 and T2: cyst with completely calcified walls (hypointense)

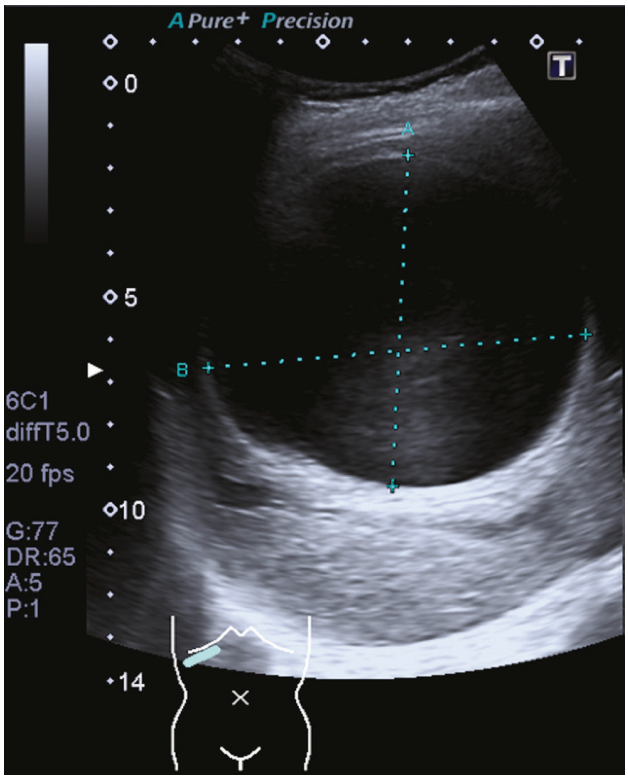

Fig. 3 *Abdominal ultrasound in a 55-year-old woman with diffuse abdominal pain shows a large cystic lesion with hyperechogenic margins in the right lobe of the liver and posterior acoustic enhancement. Gharbi I-CE1.*

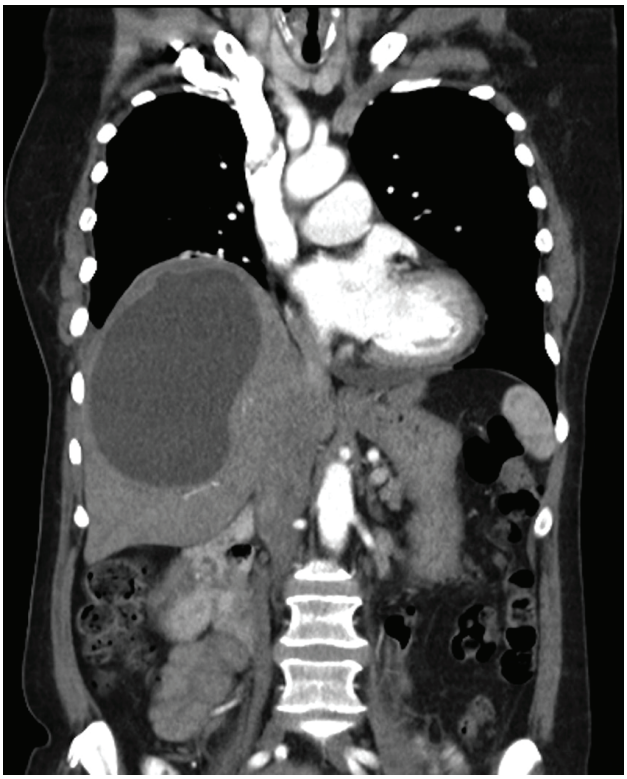

Fig. 4 *Coronal contrast-enhanced abdominal CT scan in the same patient as in Figure 3 shows a cystic lesion of simple appearance in the liver parenchyma. Microscopic examination: liver tissue with dense capsule of hydatid cyst. Gharbi I-CE1.*

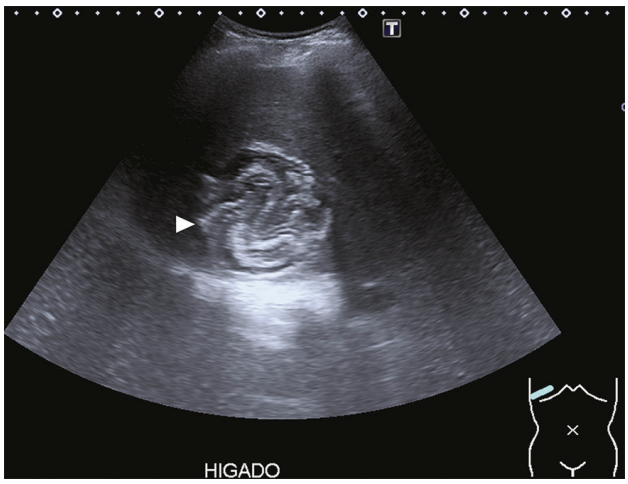

Fig. 5 *Abdominal ultrasound in a 48-year-old patient with right upper quadrant pain shows, in the liver parenchyma, a heterogeneous lesion with echogenic linear structures inside (arrowhead) and posterior acoustic enhancement. Gharbi type II-CE3.*

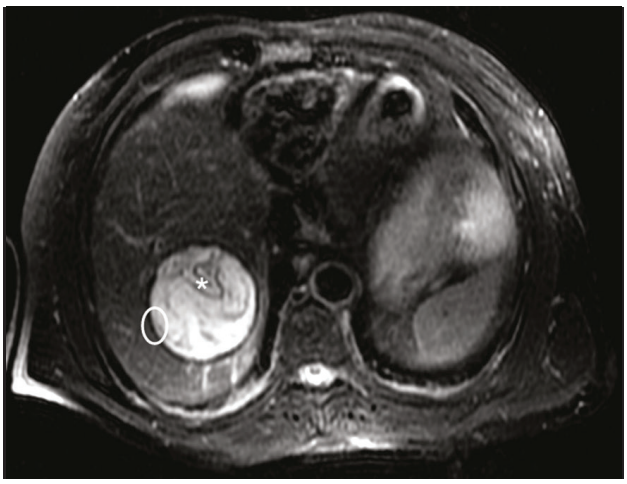

Fig. 6 *Axial fat-suppressed (FS) T2-weighted MRI of the abdomen shows a focal lesion in segment VII of the liver with heterogeneous, mainly high, signal intensity, with linear structures of low signal intensity with an appearance of "detached membranes" (asterisk), associated with ring of low intensity signal corresponding to the signet ring sign (white ellipse). Gharbi type II-CE3.*

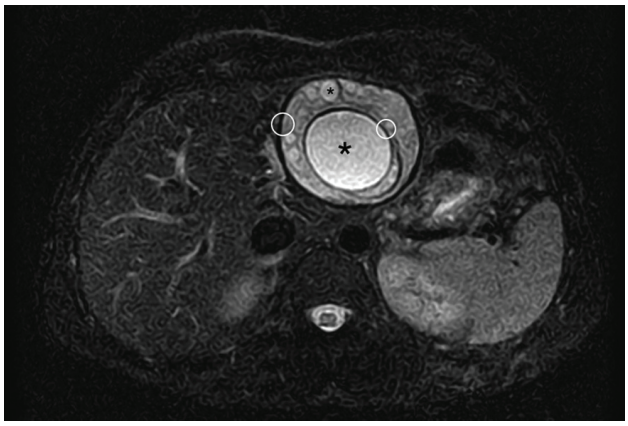

Fig. 7 *Axial T2 Trigger abdominal MRI shows a focal lesion in the left lobe of the liver with multiple rounded hyperintense lesions (asterisks) of cystic appearance, with a subtle hypointense rim corresponding to the signet ring sign (white circumferences). Gharbi type III-CE2.*

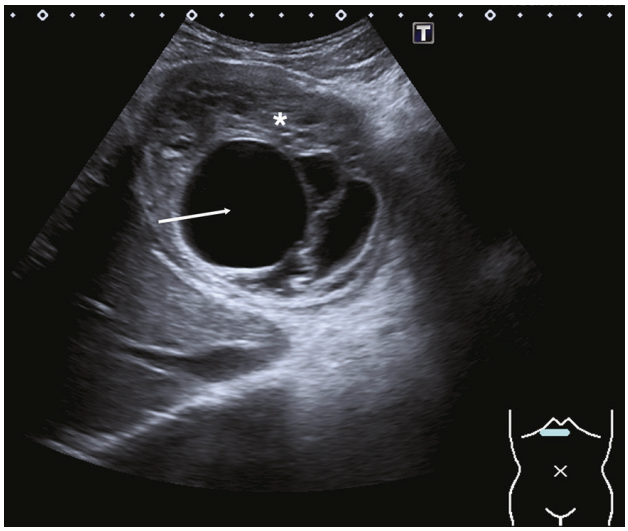

Fig. 9 *Abdominal ultrasound in a 27-year-old patient with pain in the epigastric region of several days' duration shows a focal lesion of heterogeneous appearance in the left lobe of the liver, with echogenic areas of solid (asterisk) and cystic (white arrow) appearance inside. Gharbi type IV-CE4.*

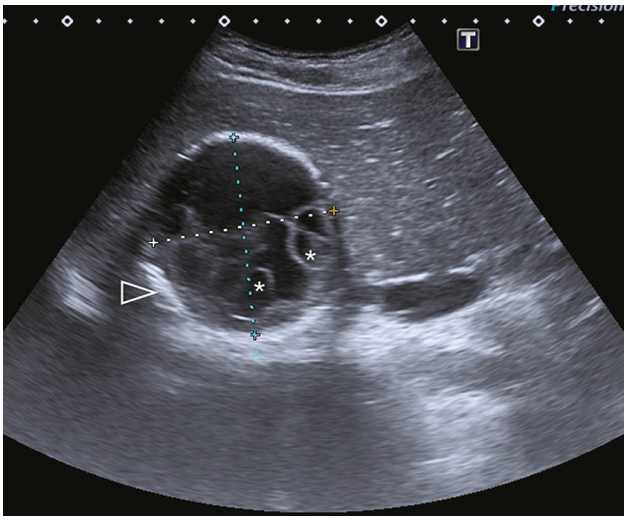

Fig. 8 *Abdominal ultrasound in a 14-year-old patient with generalized abdominal pain associated with vomiting shows a focal anechoic lesion in the right lobe of the liver, with multiple cystic images inside (asterisks) and thickened echogenic wall (arrowhead). Gharbi type III-CE2.*

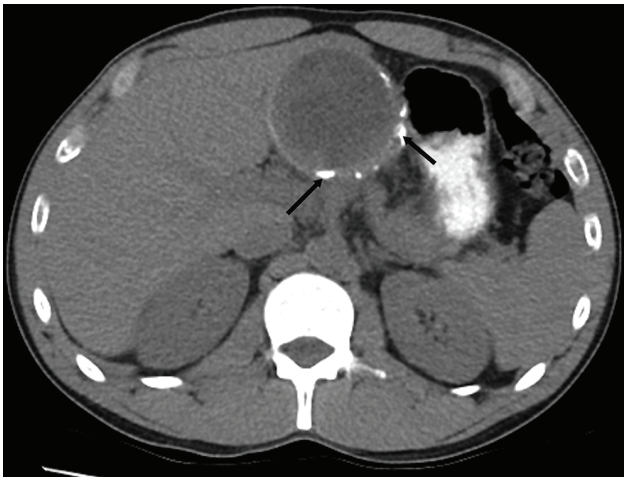

Fig. 10 *Axial, non-contrast enhanced abdominal CT scan shows a focal, hypodense rounded lesion of partially calcified margins (arrows) in segment II of the liver. Gharbi type IV-CE4.*

is the method of choice due to its accessibility, low cost and ability to determine the characteristics of cysts for the diagnosis of hepatic hydatidosis. CT and MRI are reserved for controversial cases or cases with other potential differential diagnoses, as well as complications, location of the disease in sites outside the abdominal cavity or simply for planning the surgical approach. All these imaging methods are considered an essential tool for diagnosis, along with serology and epidemiology.

Financial support
The authors declare no financial support for this article.

Conflicts of interest
The authors declare no conflict of interests

Ethical responsibilities

Protection of human subjects and animals. The authors

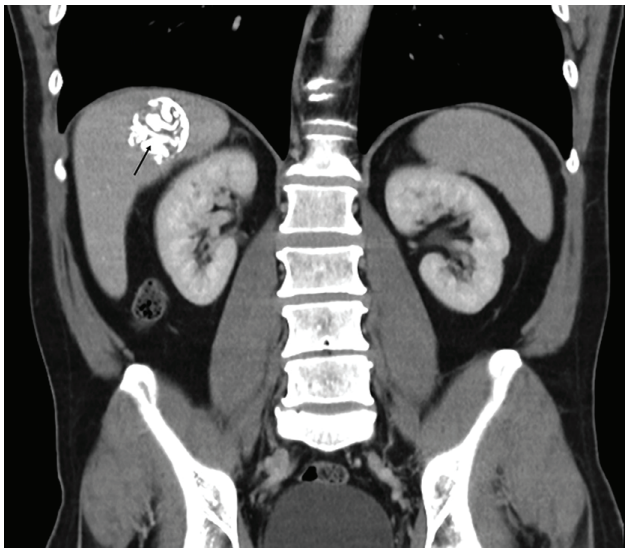

Fig. 11 *Coronal intravenous contrast-enhanced CT scan shows a rounded, entirely calcified lesion (arrow) in the right lobe of the liver. Microscopic examination: calcified, hyalinized/sclerosed capsule and another multilaminated capsule associated with amorphous sand-like material. Diagnosis: dead hydatid cyst. Gharbi V-CE5.*

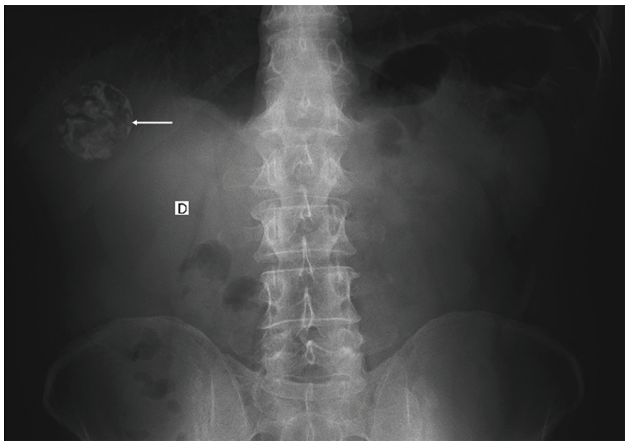

Fig. 13 *Abdominal x-ray taken in a standing position in the same patient as in* Figure 11 *shows a radiopaque oval lesion in the right upper quadrant (arrow). Gharbi V-CE5.*

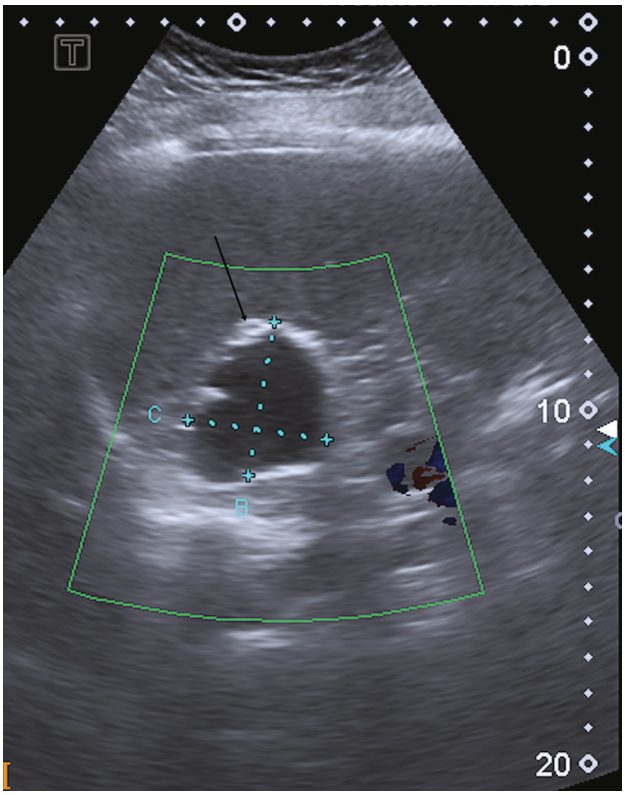

Fig. 12 *Abdominal ultrasound in a 58-year-old asymptomatic woman from the province of Entre Ríos shows a focal anechoic lesion of echogenic margins consistent with calcifications (arrow), located in the liver parenchyma. Gharbi type V-CE5.*

declare that no experiments were performed on humans or animals for this investigation.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects mentioned in the article. The author for correspondence is in possession of this document.

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