## HYDATIDOSIS: RADIOLOGICAL AND CLINICAL ASPECTS

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Hydatidosis is a parasitic disease (of the liver, lungs, kidneys, bones and other organs) caused by larva cestodes of the species Echinococcus granulosus (dog tapeworm). The aim of the paper was to present the X-ray aspects of echinococcal (hydatid) cysts. We will illustrate the radiological findings and pictures of hydatigenih cysts in the liver, lungs, kidneys and bones. The authors conclude that radiographic image of the hydatid cyst is typical, while the radiological examination methods are relevant to the diagnostics of hydatidosis. Acta Medica Medianae 2017;56(1):82-92.

Key words: radiology, hydatidosis, Echinococcus

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a (larval stage). Echinococcus (hvdatid) cvst is the larval form that develops in the interim host. Mature metacestode produces many parasites that can develop into adult cestodes if it enters a suitable host (1-5).

The man is an accidental (aberrant), transient host of larval forms of *E. granulosus* (Figure 2) (6). The disease is referred to as Echinococcosis/hydatid disease - HD or cystic echinococ-



Figure 1. Echinococcus granulosus, an adult (4)

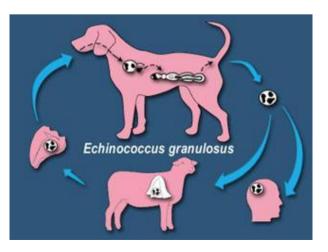


Figure 2. Life cycle of E. granulosus (6)

## Introduction

Hydatidosis is a parasitic disease caused by the larval form of the parasite of the genus Echinococcus. This genus consists of four morphologically and biologically different species. In humans, usually two types can parasitize: Echinococcus granulosus (E. granulosus) and E. Multilocularis (1). In recent years, there is a number of available molecular techniques that allow identification of strains of Echinococcus spp. which allows consideration of epidemiological scenario of echinococcosis in a particular field (2, 3).

The development cycle of parasites (Figure 1) includes two hosts - permanent and intermediate. In the small intestine of carnivores (final/ definitive host, usually a dog, rarely wolf, coyote, jackal, fox) live adult cestodes which produce infectious eggs (containing an embryo heksakant that with/without proglottides throws the faeces into the environment. In the internal organs of the intermediate host (ox, pig, sheep, goats, deer, horses, sometimes a man), after the per os intake of infected eggs/proglottides metacestode develop

cosis (CE). This form of echinococcosis is designated as the primary CE, while the secondary CE (mainly localized in the abdominal cavity) is the result of a spontaneous or trauma-induced rupture of the cyst and the liberation of parasites and / or small cysts that can grow into large cysts (2, 7). On average, 40-80% of patients with primary CE have usually only one organ affected by a cyst. In humans, the cyst is most commonly located in the liver (60%), lungs (20%), less often in the spleen, brain and bones (7-10).

A man is infected by eggs through contaminated food or water, but also by dirty hands due to inadequate hygiene habits. Irresponsible attitudes of dog owners, disrespect of the rules for keeping animals and maintaining their health, avoiding dehelminisation are certainly an important link in the transmission of echinococcosis. When an egg enters the human body, embryo heksakant is released in the small intestine membrane and actively penetrates the mucosa and bowel wall until it reaches the lumen of blood or lymphatic vessel. Circulation reaches the liver, where in most cases it is retained and transformed into a hydatid cyst (hydatid cyst). The transformation is slow (increases by 1 cm in the diameter within a few months). There have been registered cases of cysts old 10-20 years. The embryo, if it is not retained in the liver, reaches the lungs. If it passes the pulmonary barrier and enters the systemic circulation, it can settle in any organ and tissue (1-12).

Hydatid cyst (CH) (Figure 3) is enveloped by the cuticle of 1-2 mm in size, which is milky white, impermeable to bacteria, but misses the necessary nutrients (13). It is resistant to the effects of phagocytes, bile and enzymes. Beneath is the membrane proligera (germinative) which contains glycogen, fat and lime granules, it is clear and smooth. By the production of the inner membrane,



Figure 3. Hydatid cyst removed by surgery (13)

hydatid liquid is formed and it is sterile, with alkaline pH, colorless, odorless, rich in minerals, fatty acids, glycogen, inorganic salts. CH-toxalbumine contains toxic substances, and does not coagulate when heated. Budding from proligera memrane creates proligera vesicles towards the inside of the cyst and vesicles are connected by penducles to the membrane proligera. The vesicles can break off and thousands of them can swim freely into the hydatid liquid. The vesicles contain invaginated protoscolex future tapeworm. When vesicles burst in the hydatid fluid, protoscolex freely swim in the form of cystic liquid sand. Daughter cells can be found in the hydatid cysts and have two membranes. The growth of cysts can turn into pericyst when the membrane gets adventitia (fibrous tissue) due to the reaction of the organism to toxic and mechanical effects of cysts. Cysts daughters have no cuticle. Besides being fertile, cysts can become sterile due to immunity (5).

*E. multilocularis*, as an adult species, is difficult to distinguish from species *E. granulosus*. Between them, there is a difference in larval forming, *E. granulosus* forms one large hydatid cyst, which can create a secondary dissemination cysts, while *E. multilocularis* produces a spongy cystic mass that is constantly growing (1-13).

The clinical picture of hydatitosis depends on the localization of hydatid cysts (1, 2, 5, 7, 8).

If the hydatid cyst is localized in the liver, the patient complains about the pain under the right rib cage, which "hits" his right shoulder. There is an aversion to fatty foods. If it is big, it can be detected by palpation as a smooth, firm, elastic, round formation, and by auscultation as "guided trembling," only if the cyst has a large number of vesicles proligera (14-19).

If the hydatid cyst is localized in the lungs, the clinical picture may be asymptomatic or dominated by cough, hemoptysis, shortness of breath, chest pains, and under different circumstances (shot in the chest, fatigue, etc.) the cyst can burst which is manifested by coughing and yellow expectoration, salty, sometimes a bit bloody liquids (7-12, 14-19).

If the hydatid cyst is localized in the brain, the clinical picture indicates a tumor, while the development in bones leads to spontaneous fractures of the stem bone (7-12, 14-19).

Due to chronic intoxication by parasites, weight loss as well as severe allergic phenomena occur in patients due to a response to the parasite. Usually, there is a marked eosinophilia (7-12, 14-19).

Radiological images of hydatidosis are pathognomonic. For a definitive diagnosis, other radiological methods need to be done, primarily immunodiagnostics (5, 7-12, 14-20).

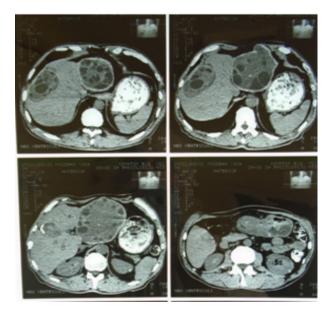
The aim is to show the X-ray aspects of the echinococcal (hydatid) cysts, while the material consists of radiographs belonging to PhD Rade R. Babic from which he selected and processed radiographs of the lungs, abdomen, kidneys, urograms and computerized tomograms with the findings of hydatid cysts in the liver (21).

Our results illustrate the radiographic findings and x-ray images of hydatid cysts in the liver, lungs, kidneys and bones.

## Hydatid cyst of the liver

The hydatid cyst in the liver becomes radiologically visible if the pericyst of the hydatid deposits calcium salts which are seen on native radiograph of the abdomen as oval or round shadows of calcium intensity with sharp contours with around 10 cm in diameter. Differential diagnosis should include porcelain gall bladder, calcified cyst of other etiology, etc. Diascopy of the abdomen can be used to diagnose hydatid cysts of the liver with non-calcified pericysts but only if they are in the liver parenchyma located subphrenically. In that case, the hydatid cyst is observed as hemispheric diaphragm-hepatic shadow with bulged contour of the diaphragm cranially, and its size depends on the size of the hydatid cyst with sharp contours. Differential diagnosis is partial relaxation of the diaphragm, subphrenic abscess, etc.

In CT scans, the hydatid cyst of the liver is seen as solitary, rarely multiple, oval or round, sharply delineated formation of different size, depending on the size of the hydatid cyst. The wall of the hydatid cyst is hypodense, sometimes more or less calcified due to the calcification of the pericyst. The contents of the hydatid cyst are watery and do not change after IV application of contrast agents (21-23). Endoluminally, mostly along the wall, there are oval or round, often multiple and rarely solitary daughter cysts of the first line, with sharply delineated hypodense wall whose content is highly absorptive as in the mother cyst. Identical CT aspects are encountered in hydatid cysts located in other places.



**Figure 4**. CT examination of the two liver hydatid cysts which contained numerous hydatid daughter cysts

From our documentation, we present CT examination of the liver with two hydatid cysts

which contained numerous hydatid daughter cysts (Figure 4). Hydatid mother cysts are oval, with sharp contrours, hypodense wall, apart from the smaller one which has a partially calcified wall. The content of the cyst has absorptive value 34 HU. Endoluminally, mainly along the wall of the hydatid mother cysts, there are numerous, oval, sharply delineated first-line daughter cysts with hypodense wall whose contents have absorptive value of watery fluid.

## Hydatid cyst of the lungs

Lung hydatidosis is less frequent than liver hydatidosis. It is more common in young people, often in the right lower lobe of the lung. It is diagnosed by radiography and/or roentgenoscopy of the lung. CT and MRI are very significant.

Radiographs show the hydatid cyst as ovoid or spherical, usually solitary, soft, homogenous, sharply delineated formation, with the diameter depending on the size of the hydatid cyst. In differential diagnosis, hydatid cysts are differentiated from solitary ones by Escudero-Nemenov sign. The Escudero-Nemenov sign in diascopy refers to the cyst which is round during expiration and ovoid with longer cranio-caudal diameter during inspiration phase (9, 10).

In addition to the described X-ray picture, lung hydatidosis can also show some pathognomonic varieties:

- Zhebe's sign which appears due to a pericyst burst and air penetrates into the space between pericysts and cyst. The air appears in the lung radiograph when the patient is in the standing position and it resembles a sickle-shaped transparency with downward concavity and cranial convexity.

- The penetration of air into the cyst lumen is seen in radiographs as a ring-shaped shadow with levels. The air is cranially positioned and the cyst fluid is caudally positioned. The border fluid level is horizontal and it changes when the patient changes the position, and it is always horizontal and sharply delineated.

- Ivanissevich sign (double arch symptom) develops due to the separation of membrane from the pericyst and penetration of air into the hydatid cyst and the capillary space between the pericyst and hydatid membrane. Radiographs show a double ring-shaped shadow with two cranially positioned sickle-like transparencies (the air in the capillary space between the pericyst and cyst and within the cyst), separated by linear, soft-tissue shadow (hydatid membrane). In the caudal half, there is a soft-tissue shadow (cyst fluid) with horizontal sharp level which changes with a change in the patient's position.

- Camelot sign is characterized by the ringshaped shadow (pericyst) with uneven level of the shed and shriveled hydatid membrane which floats on the surface of the fluid. The condition is that the central ray is parallel to the level of the cyst fluid. - Escudero-Tobias sign is characterized by the ring-shaped shadow with uneven level caused by the shed and shriveled hydatid membrane which overlies the hydatid daughter cysts of the first line, within which there is some air. The condition is the same as in the Camelot sign as well as other listed signs.

Cub's sign is seen as a ring-shaped shadow (pericyst) without fluid content and with shriveled hydatid membrane on the bottom which moves when the patient changes the position. It develops by bursting and coughing out of the liquid cyst content.

- Lessentisseur's signis seen as a ringshaped shadow with cranially positioned two sickle-like transparencies (gas in the cyst and in the capillary space between the pericyst and hydatid membrane), separed by curved, linear, soft-tissue, sharply delineated shadow (hydatid membrane) and uneven, sharply delineated hydro-gas level (first line daughter cysts which float on the surface).

- Ring-shaped shadow develops when the cyst membrane and its content is coughed out. Empty pericyst remains, which is radiologically visualized as a ring-shaped shadow.

The varieties of the radiographic image of lung hydatidosis are shown in Figure 5.

From our documentation, we present the standard X-ray of the lungs and heart in standard projections, with hydatidosis of the left lung with Zhebe sign (Figure 6). Hydatid cyst located in the

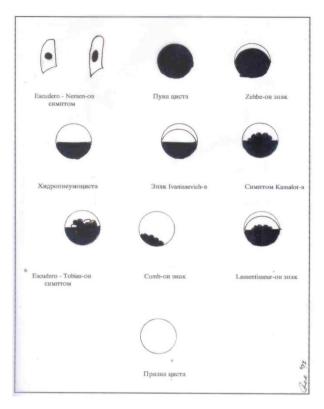


Figure 5. Varieties of radiographic images of lung hydatidosis

middle left lung area is visualized as round, sharply delineated, soft-tissue shadow, with 8 cm in the diameter and with pathognomonic Zhebe sign.

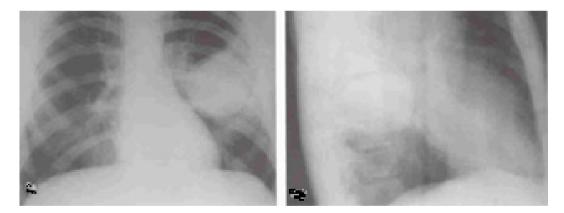


Figure 6. Radiograph of the lungs and heart in the standard projections on which the left pulmonary hydatid cust with Zhebe sing can be seen

The thickness of the pericyst is about 4 mm.

## Hydatid cyst of the kidney

Hydatid cyst in the kidney is rare. It becomes visible on the native radiogram of the urinary tract if calcium salts are deposited in the pericyst and it is then seen as oval or a round shadow of calcium intensity, with sharp contours, different diameters, depending on the size of the hydatid cyst. In urograms, larger hydatid cyst deforms the kidney and its pyelocaliceal system and causes astonishing irregularities. Sometimes, the pyelocaliceal system resembles a spider or wheel spokes. The calices are elongated, distorted, stocky, stubby or cut off at the periphery. Pressed by the hydatid cyst, infidibulum cups are bent like branches or tentacles, deformed, elongated or shortened (9, 10, 15). If the hydatid cyst communicated with the pyelocaliceal kidney system, the following

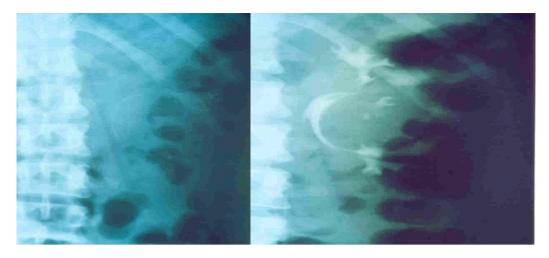


Figure 7. Hydatid cyst of the left kidney

pathognomonic signs can be diagnosed:

 champagne glass sign – the hydatid cyst communicated with the drainage cup and takes on the form of the champagne glass;

 crescent sign – occurs when contrast agent penetrates between the cyst and pericyst;

- grape cluster sign – occurs when iodine contrast agent penetrates the cyst and stains it, while endoluminally, daughter cysts of the first line, presented in the form of multiple, sharply delineated, mutually grouped defects, form grapelike cluster formation;

- liquid pneumocyst sign can be found in a partially emptied hydatid cyst (identical to the pulmonary hydatidosis).We present the native radiogram of the urinary tract and intravenous urography (IVU). The requirement is to make the urogram of the patient in the standing position.

From our documentation, we present a patient diagnosed with the hydatid cyst of the left kidney (Figure 7) (21). The hydatid cyst is located in the hilus of the left kidney, it is sharply delineated, 6 cm in diameter, with calcified, 3 mm thick wall, which disrupts the appearance of the kidney. The pyelocaliceal system of the kidney is deformed. The pyelon is sickle-shaped with lateral concavities. The large cups include the cyst resembling the eagle's claws. Small cups appear regular.

## Hydatid cyst of the bone

Bone hydatidosis is the least frequent. It develops for a long time, about ten years, and even longer. The X-ray image visualization of the cyst in later stages is not difficult. Radiographic signs are not very characteristic in order for them to be sufficient for the diagnosis. Both clinical picture and disease course are important (9, 10, 18, 19).

The hydatid cyst is most frequently located in the pelvis, followed by the vertebrae, humerus, tibia and femur. It is less frequent in the bones of the head and ribs. Figure 8 shows the most common location of the hydatid cyst in human bones, which is marked with yellow color. Radiological findings depend on the type of parasite, growth, age and volume of the hydatid cyst, particularly on its position in the bones (9, 10, 18, 19).

The hydatic cyst is most often located in the spongiosis of the originally infected bone which it gradually destroys and progresses towards the cortex where it first causes thinning and then inflation of the bone.

Hydatid cyst in the shins is visualized as round or oval, and in flat bones it is seen as usually polygonal or completely irregular transparency with clear thick or thin border towards the surroundings, which represents sclerotic defense pericyst. Sometimes, due to multilocularity, the hydatid cyst has 'honeycomb' appearance. The in-



Figure 8.: The most common localization of hydatid systs in the human bones

fected bone is swollen, has the appearance of a spindle or a gnarled stick. Reaction of the periosteum is sparse. Penetrating into the surroundings, periosteal cysts grow and they are similar to those that begin to develop in the soft tissue, suppress or etch the bone, creating shallow or deeper, regular or irregular, sharply delineated fissures. Spontaneous fractures of the long bones or compressive fractures of the vertebrae are common with large hydatid cysts. Differential diagnosis should include common bone cysts, fibrous dysplasia, osteoclastoma, osteogenic sarcoma, metastases in other rare diseases such as generalized cystic fibrosis, lipid granulomatosis, etc.

### Conclusion

Echinococcosis (hydatid disease - HD; cystic

echinococcosis - CE) is an old disease, unfortunately, still present around the world, which is still a public health problem for many countries. Today, there is a number of direct and indirect methods, invasive and non-invasive diagnostics in HD. Imiging methods are suitable for screening and detection of new cases of patients suffering from these parasites, while tests for the detection of specific antibodies in serum are of special importance in serodiagnostics of HD. We present a radiological picture of a hydatid cysts of the liver, lungs, kidneys and bones. Radiological images of hydatid liver, lungs, kidneys and bone is manifested in various aspects of roentgenology. All presentations of hidatid cysts were confirmed by positive serological findings of specific IgG antibodies to E. granulossus.

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## Novine u medicini

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# HIDATIDOZA: RENDGENOLOŠKI I KLINIČKI ASPEKTI

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Hidatidoza je parazitsko oboljenje (jetre, pluća, bubrega, kosti i drugih organa) uzrokovano larvom cestode vrste *Echinococcus granulosus* (pseća pantljičara). Cilj rada bio je prikaz rendgenskih aspekata ehinokokne (hidatidne) ciste. Radom se ilustruju rendgenološki nalazi i slike hidatigenih cista u jetri, plućima, bubrezima i kostima. Autori zaključuju da je rendgenološka slika hidatidoze tipična, dok su radiološke metode pregleda relevantne u dijagnstici hidatidoze. *Acta Medica Medianae 2017;56(1):86-92.* 

Ključne reči: rendgenologija, hidatidoza, ehinokok