

## PAPER DETAILS

TITLE: Pulmonary Hydatid Cyst: Pathophysiology, Etiopathogenesis, Diagnosis and Treatment

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PAGES: 105-109

ORIGINAL PDF URL: <https://dergipark.org.tr/tr/download/article-file/3338352>

# Pulmonary Hydatid Cyst: Pathophysiology, etiopathogenesis, diagnosis and treatment

## Pulmoner Hidatik Kist: Patofizyoloji, etiopatogenez, tanı ve tedavi

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**Submitted Date:** 17 August 2023, **Accepted Date:** 31 December 2023

### SUMMARY

**Aim:** Echinococcosis is a parasitic disease caused by cestodes of the genus Echinococcus, which often affects the liver and lungs. Pulmonary hydatid cysts are often transmitted in childhood and remain asymptomatic for a long time. As a result, the diagnosis is often incidental in noncomplicated cases. In some cases hydatid cysts may rupture and in this occurrence it is then called a complicated cyst. When a complicated pulmonary hydatid cyst occurs, various symptoms such as chest pain, shortness of breath, cough, and hemoptysis may be observed. For diagnosis radiologic methods and serologic tests may be used. The most commonly used radiological method for pulmonary hydatid cysts is chest radiography. Echinococcosis can be treated pharmacologically with medications under certain situations; nonetheless, surgical treatment is the gold standard. Pharmacological treatment includes benzimidazole group drugs such as albendazole and in surgical treatment, various techniques such as enucleation, pericystectomy, capitoned cystotomy are applied.

**Keywords:** Cyst, Echinococcosis, hydatid disease, hydatidosis, lung, pulmonary

### ÖZET

Echinococcosis, genellikle karaciğer ve akciğerleri etkileyen Echinococcus cinsinin cestodlarının neden olduğu parazitik bir hastalıktır. Pulmoner hidatik kistler sıklıkla çocukluk çağında bulaşır ve uzun süre asemptomatik kalır. Sonuç olarak, komplike olmayan vakalarda tanı genellikle tesadüfen konur. Bazı durumlarda kist hidatik patlayabilir ve bu durumda komplike kist olarak adlandırılır. Komplike bir pulmoner kist hidatik oluştuğunda göğüs ağrısı, nefes darlığı, öksürük ve hemoptizi gibi çeşitli semptomlar görülebilir. Tanı için radyolojik yöntemler ve serolojik testler kullanılabilir. Akciğer kist hidatigi için en sık kullanılan radyolojik yöntem akciğer grafisidir. Ekinokokkoz, belirli durumlarda ilaçlarla farmakolojik olarak tedavi edilebilir; bununla birlikte, cerrahi tedavi altın standarttır. Farmakolojik tedavi albendazol gibi benzimidazol grubu ilaçları içerir ve cerrahi tedavide enükleasyon, perisistektomi, kapitonlu sistotomi gibi çeşitli teknikler uygulanır.

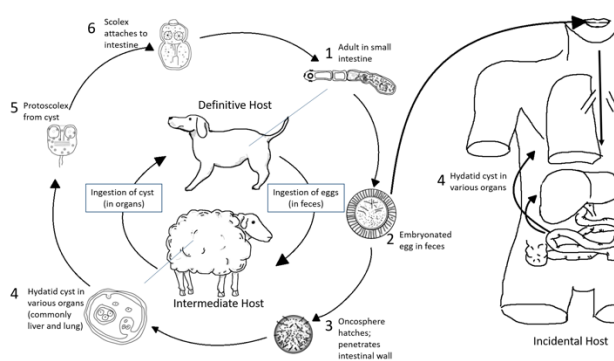
**Anahtar Kelimeler:** Akciğer, Ekinokokkoz, hidatik kist, kist, pulmoner hidatik kist

## Background

Echinococcosis is an infectious disease caused by cestodes of the genus *Echinococcus*. To date, 6 species of *Echinococcus* have been described. From these six species four are pathogenic 2 being pathogenic to humans. These pathological species are: *Echinococcus granulosus* (causing cystic echinococcosis.), *Echinococcus multilocularis* (causing alveolar echinococcosis.), *Echinococcus shiquicus* (non pathogenic for humans) and *Echinococcus felidis* (non pathogenic for humans) (1,2). *Echinococcus granulosus* (cystic echinococcosis) is the most common echinococcosis species, accounting for 95% of echinococcosis cases worldwide (3). Despite being an endemic disease in many regions, including the Mediterranean, South America, and Australia (4,5), echinococcosis is a global disease. This presents to us the importance of the diagnosis and treatment even in non endemic countries since cystic echinococcosis can remain asymptomatic for a long time and is generally found incidentally. The purpose of this review is to highlight the significance of pulmonary echinococcosis diagnosis and therapy in humans.

## Etiopathogenesis

The definitive host for *Echinococcus granulosus* is the dog. These dogs that are carriers of adult cestodes in their small intestines also excrete the eggs produced in their feces (6,7). These eggs laid in the environment are often ingested by an intermediate host (sheep are the most common intermediate host for *Echinococcus granulosus*). Eggs ingested by the intermediate host develop into embryos in the small intestine and reach the liver via the portal circulation where they turn into cysts (Figure 1) (6).



**Figure 1.** Echinococcus life cycle

Humans are incidental hosts for *Echinococcus*. Echinococcal infection is transmitted to humans by contact with the definitive host or by ingestion of water or vegetables contaminated with echinococcal eggs (4,8).

Hydatid cyst eggs hatch once they reach the human stomach releasing their embryo (4). Embryos invade into the small intestine surpassing the mucosa into the blood

flow and reaching the liver where they transform into hydatid cysts. Once they reach the liver they can move past into the lung where they also can transform into hydatid cysts. The most common site for infection is the right inferior lobe due to its relatively high blood flow (8).

The wall of hydatid cysts have three layers. These layers are pericyst, ectocyst (acellular middle lamina layer) and endocyst (germinal layer); pericyst being the outermost and endocyst being the innermost layer (8). Pericyst is formed by the defence mechanisms of the host restricting the invasion. Ectocyst is a layer which allows necessary nutrients to pass. Endocyst is the layer which germinates many daughter vesicles inside the cystic cavity.

Pulmonary hydatid cysts have certain characteristics compared to hydatid cysts in other parts of the body. Lungs facilitate the growth of hydatid cysts due to their negative pressure and compressible structure (9,10). As a result of this characteristic of the lung, pulmonary hydatid cysts grow faster than hydatid cysts in other regions of the body (11). Also, unlike other hydatid cysts in the other regions, calcification and juvenile cyst formation are very rare in pulmonary hydatid cysts (7,12).

## Pathophysiology

In the pathophysiology of echinococcosis, canines play an important role. *Echinococcus* species reside inside the small intestine of their definitive hosts and transmit its eggs by the hosts' feces. Then *Echinococcus* eggs are ingested orally and pass through the gastrointestinal tract until they reach the small intestine (4). After hatching and moving to the small intestine the embryos invade into the mucosa gaining access to the blood flow and the organs such as the liver and lungs (Figure 1) (6). Primary infections mostly consist of solitary cyst (13). Echinococcal cysts are most frequently seen in the liver (> 65%), lungs (25%), spleen, kidneys, and heart in adults (8,13,14). However, in the pediatric age group the most common site of infection is the lungs (11).

## Clinical Signs and Symptoms

Echinococcosis can present with various signs and symptoms. However one attribute of the echinococcal cysts is slow growth which in turn causes a long asymptomatic period after its acquisition (15). In pulmonary hydatid cyst infection, cysts larger than 5 cm usually are the cause of bronchial compression. This state of compression causes patients several symptoms or complications. Common symptoms in pulmonary hydatid cyst include cough, followed by chest pain, dyspnea, expectoration (sputum), fever, hemoptysis (4,8,11). Common complications of pulmonary hydatid cysts are cyst rupture, secondary infection, suppuration, and pneumothorax (14). Rupture of the cyst is caused by degeneration of the cyst membranes and the risk increases

proportionally as the number of cysts increase. Rupture can occur in the bronchus or pleural cavity. Cyst rupture in the pleural cavity may present as pneumothorax, effusion, and emphysema (16). The most common complication of hydatid cyst rupture is infection, which clinically shows features of lung abscess (12). Rupture of the cyst may cause symptoms such as: sudden onset of chest pain, hemoptysis, cough and fever, or rarely a salty taste in the mouth (17). In addition cyst rupture may also cause a hypersensitivity reaction and result in an anaphylaxis, threatening the patients life. Symptoms and complications of pulmonary hydatid cyst are defined; they are nondiagnostic by themselves. Therefore, radiological imaging plays an important role in the diagnosis of the disease.

### Diagnosis

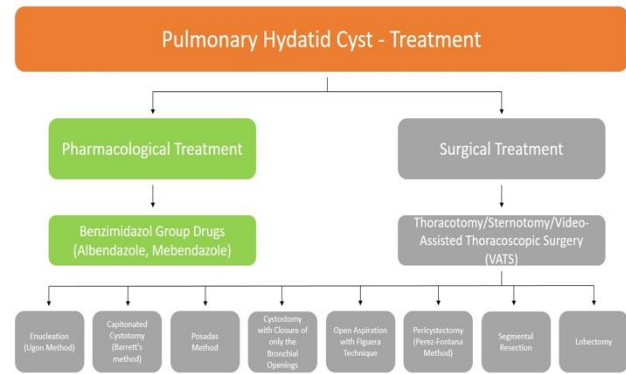
The most commonly used radiological method in the diagnosis of hydatid cyst is chest radiography (8). Chest radiography findings are divided into complicated and noncomplicated findings. In a noncomplicated hydatid cyst radiography; a well circumscribed round radio opacity, polycystic and lobulated appearance, Slot sign can be observed. In a complicated hydatid cyst radiography; Crescent sign, Escudero-Nemerow sign, Cumbo or Double Arch sign, Water Lily or Camelotte sign, Rising Sun sign, Dry Cyst sign can be observed (15).

Computed tomography (CT) can also be used for diagnosis. CT findings may show; signs of contained rupture, Crescent sign, Inverse Crescent sign, Air Bubble sign, sign of cyst rupture, Cumbo sign, Serpent sign, Swirl sign, Water Lily sign, mass within a cavity sign, Incarcerated Membranes sign, Dry Cyst sign, sing of cyst infection, Air Bubble sign, Ring Enhancement sign, air level fluid level (15).

Apart from these methods (Chest radiography and CT); USG and MRI methods, can be but are rarely used in the diagnosis of hydatid cysts. In addition to radiological methods, serological tests are used in diagnosis. These tests are; latex agglutination, passive hemagglutination, immunoelectrophoresis and specific IgE, IgM, IgG(the most selective serological test) enzyme-linked immunosorbent assay (ELISA) (18).

### Treatment

The main treatment of pulmonary cyst hydatid is surgical treatment. However, pharmacological therapy can also be applied in the treatment (Figure 2) (8,13).



**Figure 2:** Treatment of pulmonary hydatid cyst

Pharmacological treatment is applied in cases, where surgery is contraindicated, in cases of diffuse disease, multiple cysts or recurrent cysts. In such cases benzimidazole group drugs such as mebendazole or albendazole are used for pharmacological treatment (4,19). Albendazole is relatively preferred in

pharmacological treatment because albendazole has a high bioavailability and a minimum contact time

requirement of approximately 11 days with the cyst (20). The usual recommended dose is 10-15 mg/kg/day, twice a day, and should be continued for at least 3-6 months for pulmonary hydatid cysts (13). Continuous dosing has been found to be more effective than the previous method of monthly dosing interrupted 2 weeks apart to avoid hepatotoxicity (13).

Complications such as pulmonary abscess, pleural empyema may occur during pharmacological treatment in pulmonary hydatid cysts (21). These generally arise within 2 months of the treatment. Accordingly, patients who are taking pharmacological treatment should be monitored for this period.

Pharmacological treatment should be avoided due to these contraindications such as, cysts that are larger than 6 cm in diameter, inactive or calcified cysts, patients prone to bone marrow depression and pregnancy (especially in the first trimester).

### Surgical Treatment

The gold standard treatment of pulmonary hydatid cyst is surgery (13). Surgical treatment is applied to superficial cysts, ruptured large cysts, infected cysts, cysts close to vital anatomical structures and cysts with significant mass effect (22). During thoracotomy in the case of multiple cysts, cysts that are intact should be prioritized over ruptured ones due to the risk of rupture (23). In the case of cysts rupture during operation, a gauze soaked in 20% hypertonic saline or 10% povidone-iodine solution can be

used to avoid further complications.

Posterolateral thoracotomy is the most commonly performed method in pulmonary hydatid cyst surgery (13). However, for bilateral cysts, median sternotomy or two-stage thoracotomy is preferred also, superficial and small to moderate hydatid cysts can be managed with video-assisted thoracoscopic surgery (24).

Various techniques are available for the surgical treatment of pulmonary hydatid cyst. These techniques are; enucleation (Ugon method), pericystectomy (Perez-Fontana method), capitonated cystotomy (Barrett's method), cystostomy by closing bronchial openings and capitonnage (Posadas method), cystostomy with closure of only the bronchial openings, open aspiration with Figuera technique, segmental resection and lobectomy (Table 1) (25). Enucleation (Ugon method) is the process of removing the hydatid cyst with its germinal membrane (11). This method is applied in small cysts with a low risk of rupture. Positive pressure ventilation is helpful during cyst removal. Pericystectomy (Perez - Fontana method) is the operation to remove the hydatid cyst along with the pericyst (4,11). Quilted cystotomy (Barrett's method) consists of two stages; cystotomy and quilting (4,11). Cystotomy is the removal of the germinative membrane by aspiration of fluid from the cyst (Barrett technique). Quilting helps to reduce the risk of residual cavity infection, airway leak and empyema formation. However, there is a risk of deformity of the lung parenchyma when quilting is performed (26). Cystostomy (Posadas method) by closing the bronchial openings and capitonation is similar to the Barrett method (4).

Unlike the Barrett method, open airways are closed before quilting. In this way; the risk of infection, airway leak and empyema formation remaining in the cystic space is reduced. In the cystostomy technique with the closure of only the bronchial openings, the deformity of the lung parenchyma is less since capitonation is not performed. However, the risk of air leakage and infection increases (27). Open aspiration with Figuera technique is similar to; percutaneous aspiration, instillation of scolicalid agents and reaspiration (PAIR) for hepatic hydatid cysts. In this technique, cyst membranes and daughter cysts are sucked. This technique is less invasive but increases the risk of infection and air leakage within the cavity, followed by empyema (28). Segmental resection is applied in ruptured hydatid cysts. Segmental resection reduces the risk of infection and recurrence. However, it causes a decrease in lung volume. On the other hand lobectomy, involves the anatomical resection of one or more of the lung lobes containing the cysts. Lobectomy is more appropriate in the cases where cyst mass over than 50% of the lobe, infected cysts that do not respond to treatment, multiple unilobar cysts, and consequences of hydatid disease such as bronchiectasis, pulmonary fibrosis, or severe bleeding (29).

In case of multiple cysts, the risk of rupture, cyst size and spread should be evaluated before the type of treatment or surgical method is decided. Care should be taken to close the dead space after the management of large cysts to minimize air leakage and empyema (30).

There are many methods of treatment for pulmonary hydatid cysts; a patient focused approach should be preferred and chosen accordingly.

## Conclusions

Echinococcosis is a disease that is endemic however, it can be encountered worldwide. The genus *Echinococcus* which is a definitive host in the canines incidentally infect humans. If infected the parasite most often settles in the liver and lungs. When a pulmonary infection occurs the disease progression is asymptomatic, until a complication occurs or is found incidentally. Surgery is the gold standard treatment for pulmonary hydatid cysts. Abendazole is recommended farmacological treatment (13,25). In surgical treatment, there are various techniques such as enucleation, pericystectomy, and capitonated cystotomy (13). Among these multiple methods of treatments a method best suited to the patient should be selected, considering the state of the disease and the patient's current status.

## Abbreviations

%: percent; cm: centimeter; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; USG: Ultrasound; IgM: Immunoglobulin M; IgG: Immunoglobulin G; IgE: Immunoglobulin E; ELISA: enzyme-linked immunosorbent assay; mg: milligram; kg: kilogram

**Author Contribution:** Working Concept/Design: DG, Data Collection: ZM, Data Analysis / Interpretation: FU, Text Draft: DG, FU, ZM, Critical Review of Content: DG, FU, ZM, Final Approval and Responsibility: DG, FU, Material and technical support: DG, ZM, Supervision: DG, FU

**Conflict of Interest:** The authors state that there is no conflict of interest regarding this manuscript.

**Financial Disclosure:** The authors of this study stated that they did not receive any financial support.

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