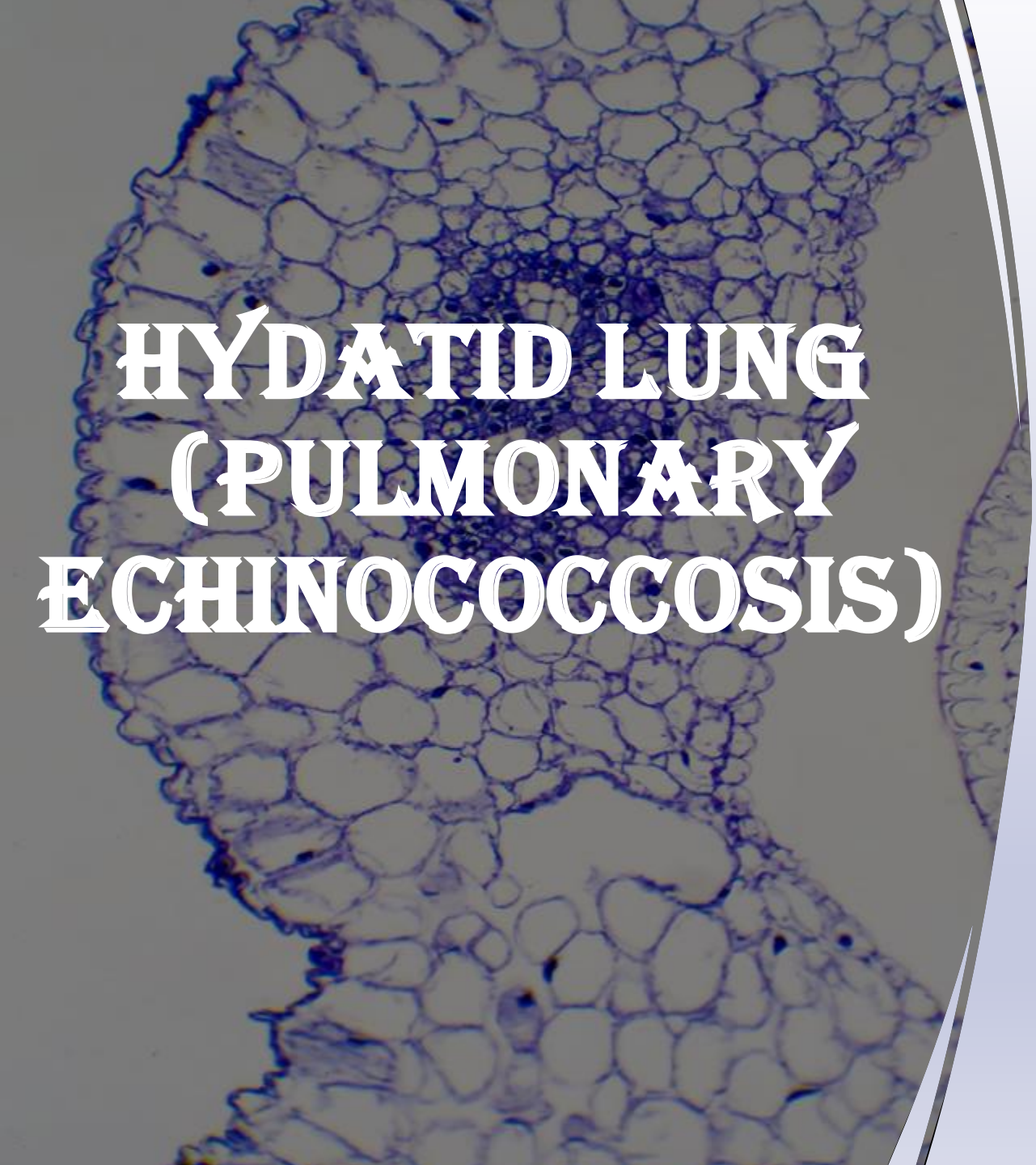

HYDATID LUNG (PULMONARY ECHINOCOCCOSIS)

Introduction and Microbiology:

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Diagnosis and Investigation:

Ayman

Management:

Baker

Supervised by: Dr. Osamah AL-Salaq

ECHINOCOCCOSIS (HYDATIDOSIS)

- It is a parasitic infection of both humans and other mammals such as sheep, cattle and pigs with hydatid cyst, the larval stage (metacestode) of different Echinococcus species.

Epidemiological distribution

❖ Globally

Hydatid disease is a cosmopolitan disease; considered endemic in certain areas including the Mediterranean countries, Asia, North Africa, South and Central America.

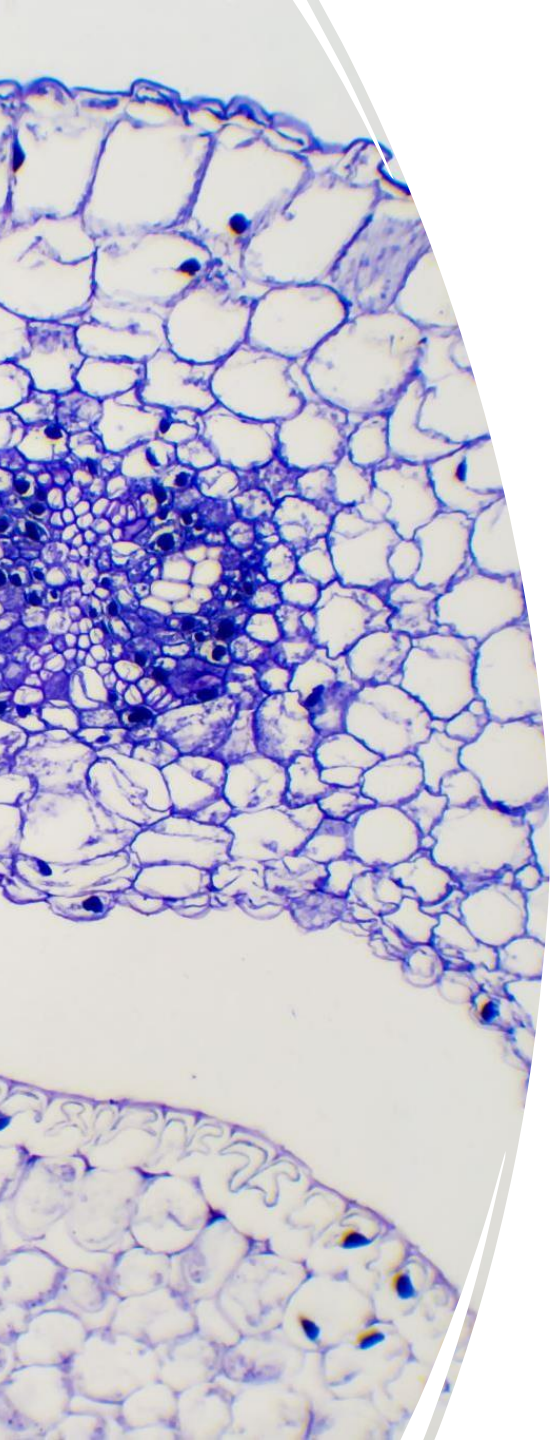
It's is more prevalent in rural areas where there is a closer contact between people and canines and various domestic animals.

❖ In Jordan

The highest number of HD cases was recorded from the central provinces of Jordan; however, most cases were reported from Al-Mafraq governorate.

❖ Age and gender

Hydatid disease is seen in subjects of any age and sex, although it is more common in those aged 20–40 years old. however, Children At high risk to be infected due to poor hygiene.

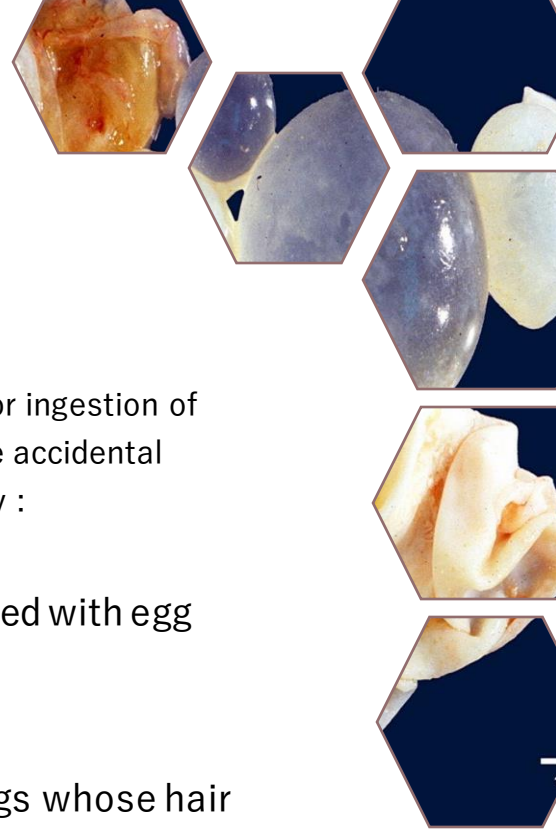


Types of Echinococcus Genus

- **Four specieses of the E. Genus exist:**
- 1. E. granulosus: Common and cause Cystic echinococcosis dog tapeworm. most common site is liver.
- 2. E. Multilocularis : less common but more dangerous cause Alveolar echinococcosis. Small fox tapeworm most common site is lung. Multilocularis infestation usually occurs in colder areas and is the most virulent specie.
- 3. E. vogeli : Rare and cause Polycystic echinococcosis.
- 4. E.oligarthus : rare and cause Unicystic echinococcosis.

Species	Cause
E. Granulosus	Cystic hydatid disease
E. Multilocularis	Alveor hydatid disease
E. Vogeli	Polcystic hydatid disease
E. oligarthrus	Unicystic hydatid disease

ECHINOCOCCUS GRANULOSUS



Habitat: Small intestine of the D.H.

D.H: Dogs, foxes and other canines.

I.H: Sheep, cattle, pigs

incidental intermediate host: also usually end stage host: human

Infective stage: Embryonated eggs " it is also the diagnostic stage "

incubation Period : variable – months to years (depends on number of cysts and growth rate)

Mode of transmission:

Human infection does not occur by the handling or ingestion of meat or viscera from infected sheep ! humans are accidental intermediate hosts that become infected either by :

* Ingesting water, food, or soil contaminated with egg bearing dog feces.

* Direct contact with a dog or handling dogs whose hair are contaminated by eggs

Flies help to spread the eggs, as does the wind,

The eggs can also be inhaled, causing primary lung disease

❖ Direct transmission of echinococcosis from human to human does not occur

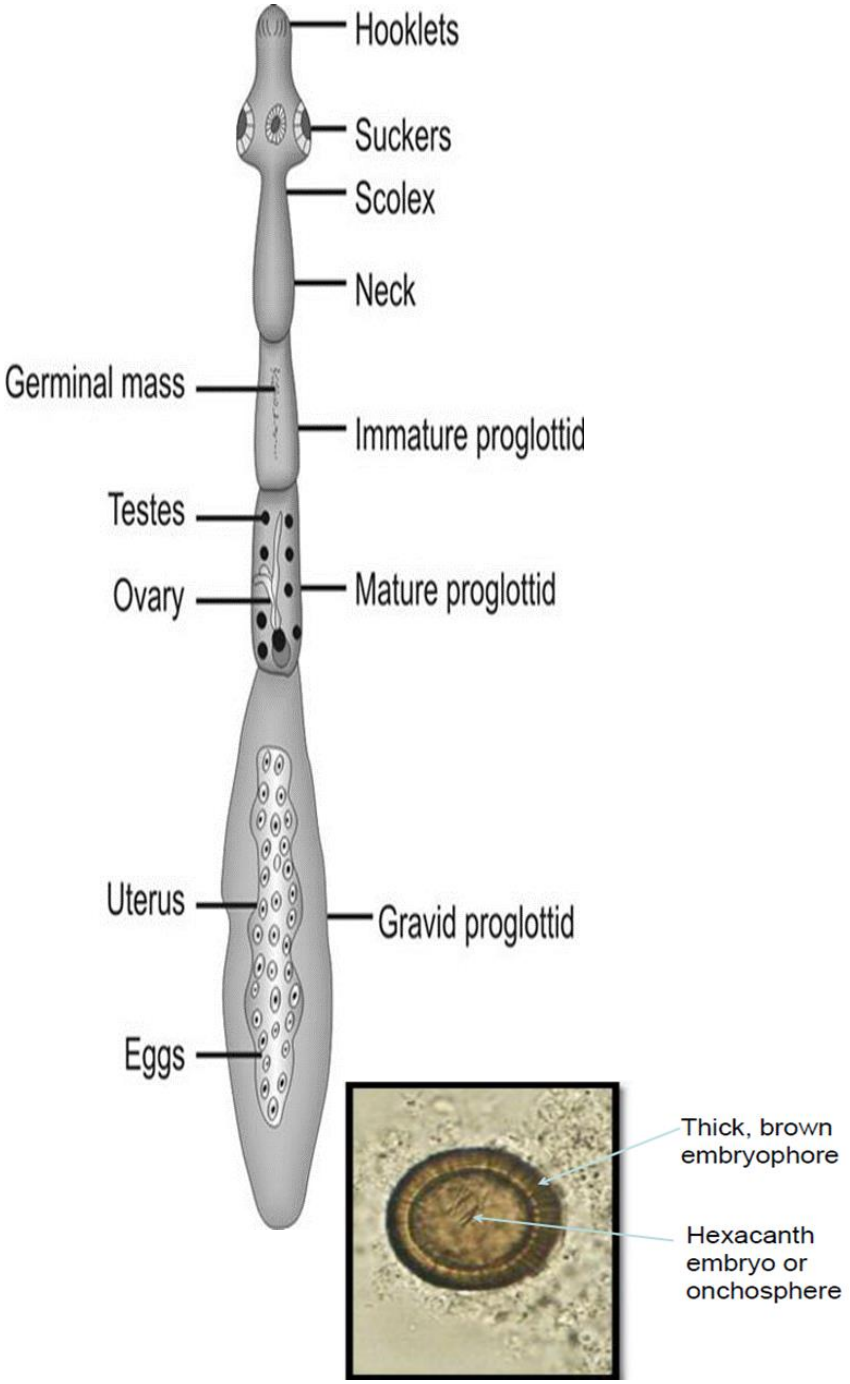
MORPHOLOGY

There are **three** morphological forms:

Adult form

Egg

Larval form (hydatid cyst)



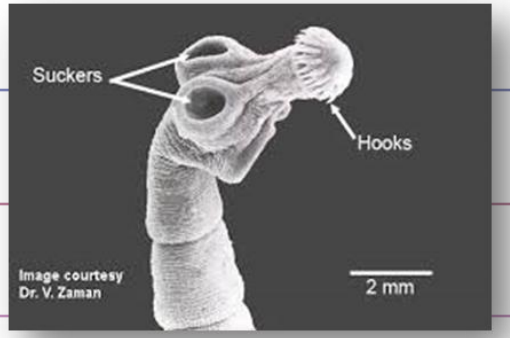
*Adult worm

length 3-6 mm Comprises of

1.Scolex: rostellum with hooks and about four suckers

2.Neck: short and thick

3. strobila/proglottids a : 3 or 4 segments.



1st segment (Immature)

2nd segment (Mature)

3rd and 4th (Gravid)

* Egg

●Spherical in shape, brown color ,have 2 layers outer and inner

● Hexacanth embryo-oncosphere

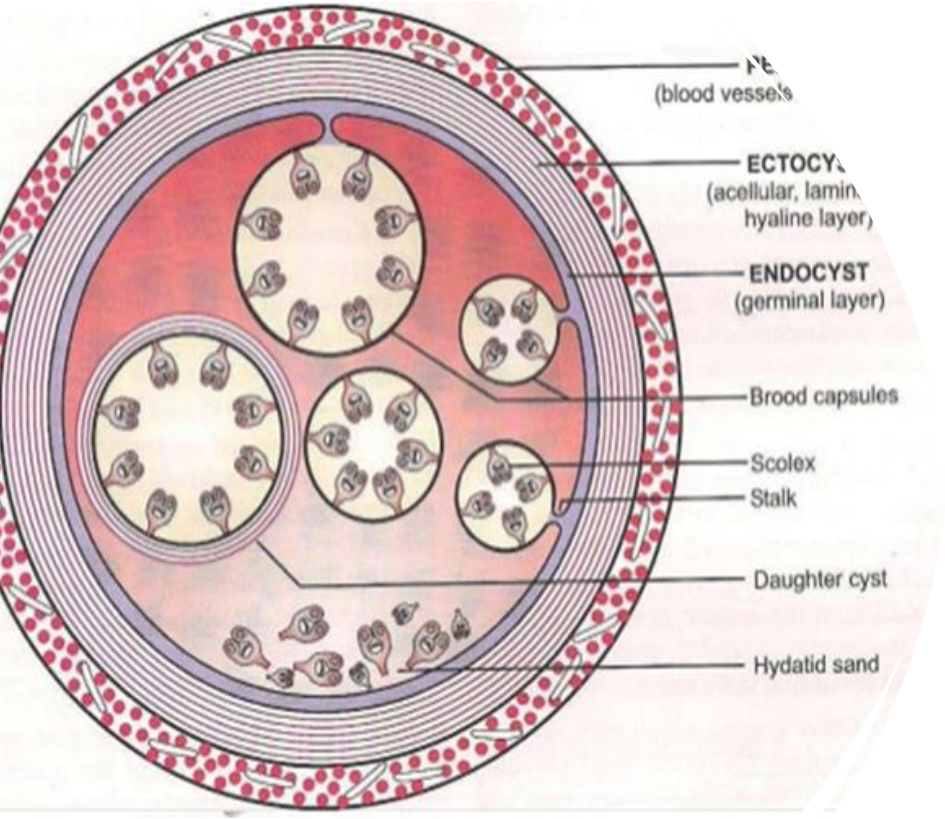
●Passed through the faeces of definitive host and it's ingestion leads to infection in the intermediate host.

❖ Cyst structure

(larval form)

➢ **Consist of :**

1. wall
2. brood capsule
3. Hydatid fluid



- A thick **wall** that is consisted of three layers:

1.Adventitia (pericyst): Host origin granulation tissue represents the host immune response.

2.Laminated membrane (ectocyst): parasite Origin. Elastic fibrous layer functions to maintain the physical integrity of the hydatid cyst.

3.Germinal epithelium (endocyst): is a single layer of cells lining the inner aspects of the cyst and is **the only living component**

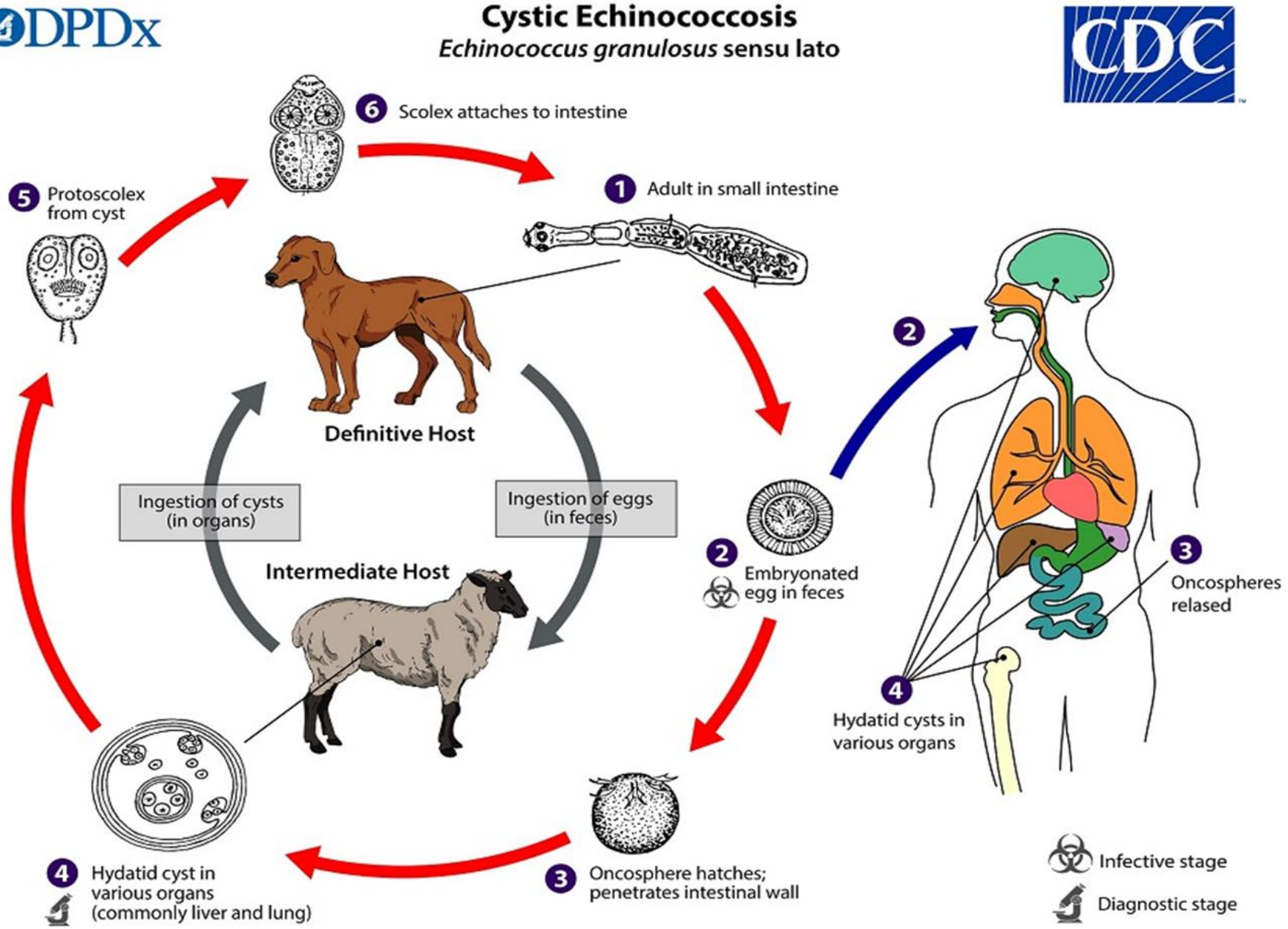
**responsible for formation of :

A.The outer layer **B.**the hydatid fluid **C.**brood capsules.

- **Brood capsule**: it is an encyst of the germinal layer, has protoscolex.
- **Hydatid fluid**: Clear, yellowish fluid with slightly acidic media. Composed of many chemical substances such as Sodium sulfate , Sodium gluconate, Succinate and other nutritional components for the protoscolex .
- It is highly toxic and antigenic thus when the cyst rupture and the fluid leak out it may induce anaphylactic reaction

○ **Hydatid sand**

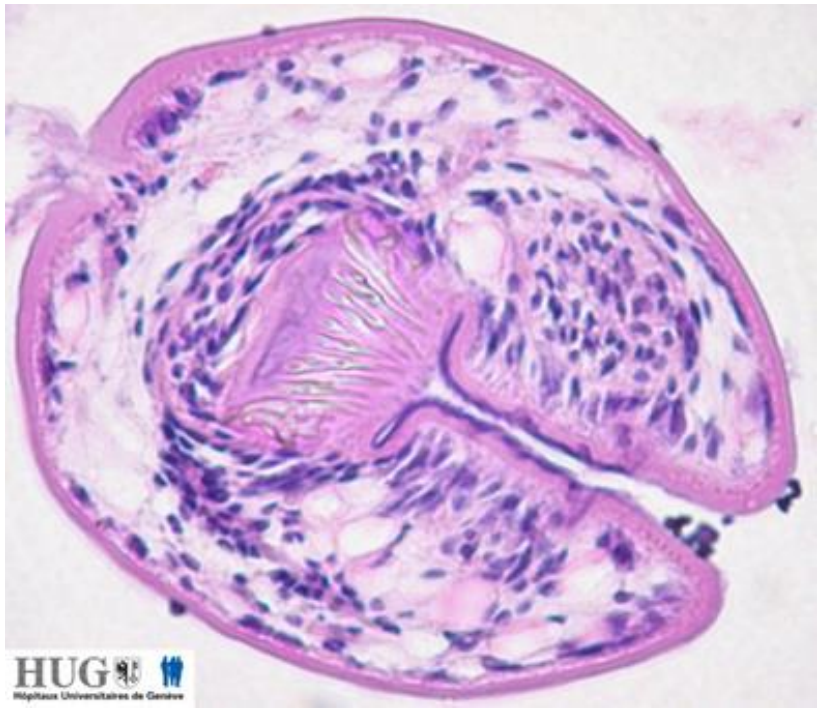
formed when the brood capsules and daughter cyst break off and their contents of scolex deposit down at the primary cyst.

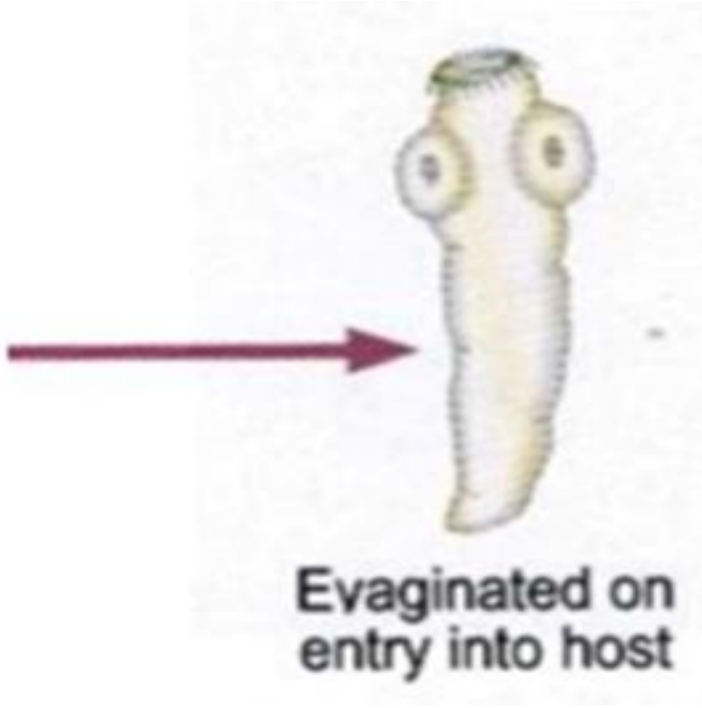


Life cycle

The lifecycle of cestodes involves 2 or more hosts.

- intermediate host where immature parasite lives and
- definitive host which harbors the mature parasite.

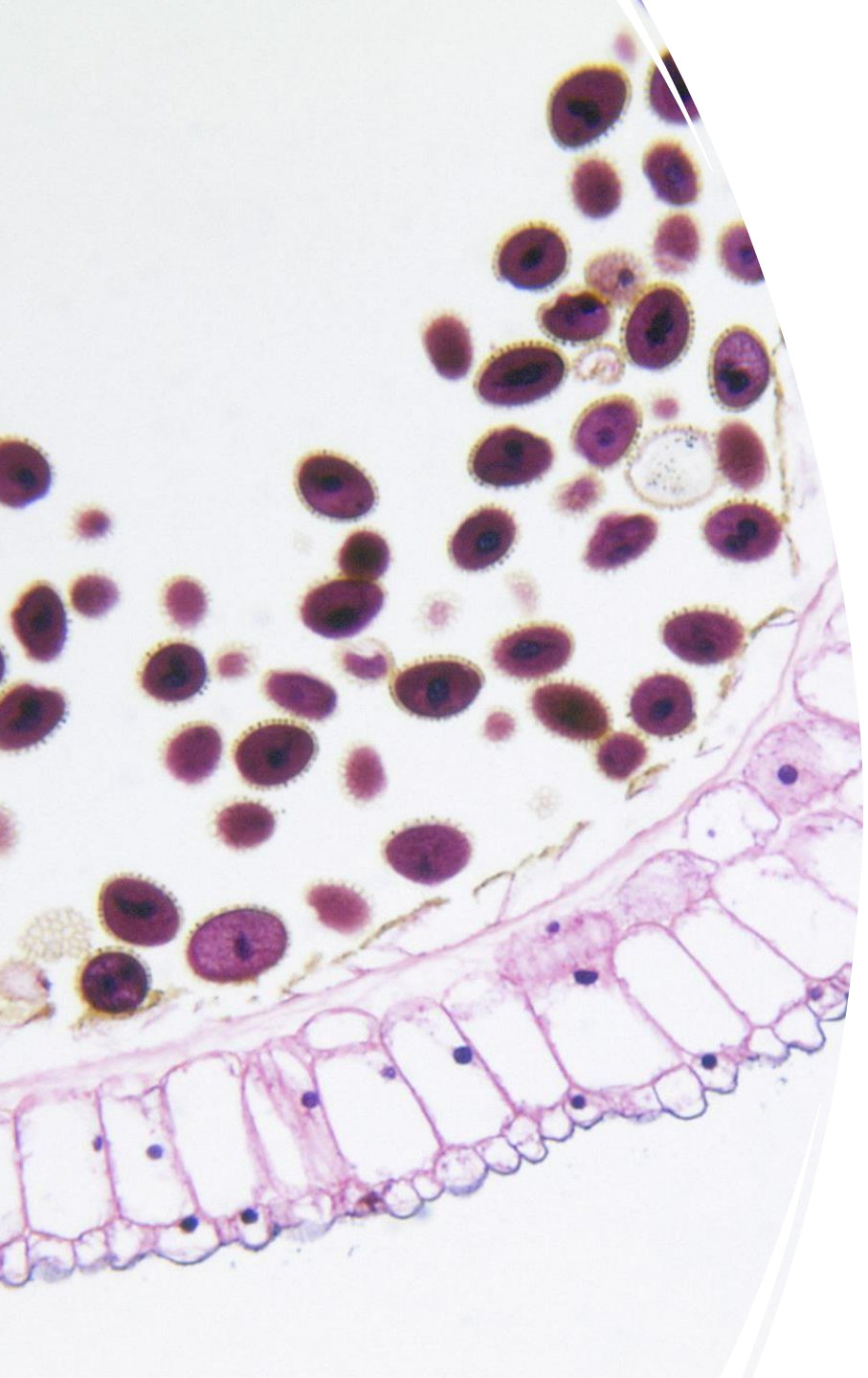




Protoscolex evagination into scolex in the D.H

The cycle

- The adult Echinococcus granulosus (1) resides in the small intestine of the definitive host.
- Gravid proglottids release eggs (2) that are passed in the feces, and are immediately infectious. After ingestion by a suitable intermediate host
- eggs hatch in the small intestine and release six-hooked oncospheres (3) that penetrate the intestinal wall and migrate through the circulatory system into various organs, especially the liver and lungs.
- In these organs, the oncosphere develops into a thick-walled hydatid cyst (4) that enlarges gradually, producing protoscolices and daughter cysts that fill the cyst interior.Protoscoleces are the infective form and will grow to the adult stage after ingestion by a definitive host which becomes infected by ingesting the cyst-containing organs of the infected intermediate host.
- After ingestion, the protoscolices (5) evaginate into Scolics and attach to the intestinal mucosa (6) , and develop into adult stages (1) **in 32 to 80 days**.
- **Humans** are accidental intermediate hosts, and become infected by ingesting eggs (2) Oncospheres are released in the intestine (3) , and hydatid cysts develop in a variety of organs (primary echinococcosis) (4) . If cysts rupture, the liberated protoscolices may create secondary cysts in other sites within the body (secondary echinococcosis)

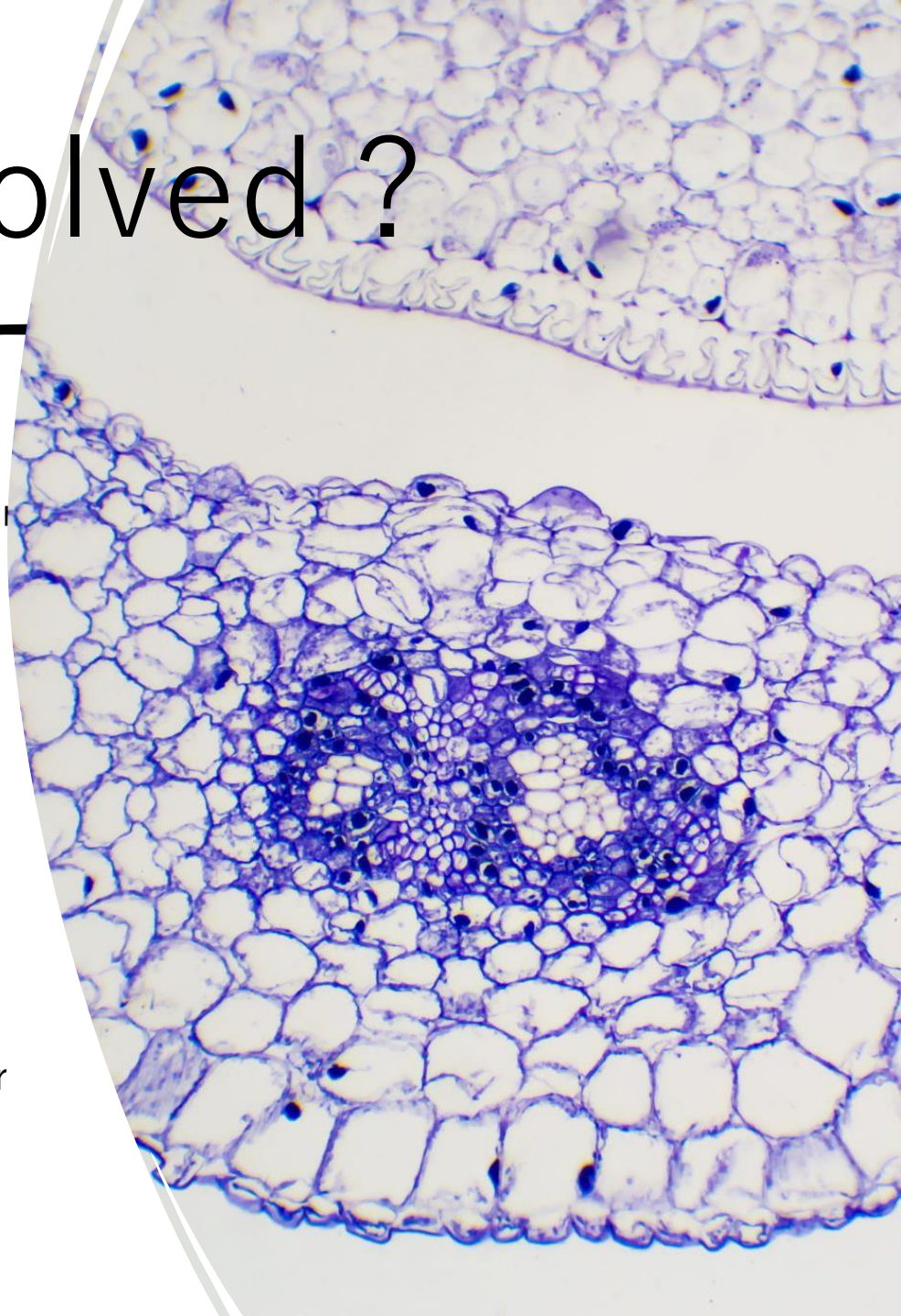


Organ involvement

- The liver is the most common organ infested followed by lungs which accounts for 10-30% of cases. Less commonly the spleen, the kidney, the bones, and the brain. Where they account for about 10% of cases. Within the chest, echinococcosis can primarily involve the pleural cavity , mediastinum and chest wall. In children, the lungs may be the commonest site of cyst formation with frequencies of 64% ,because it allows faster growth of the cyst due to its compressible nature, vascularization, and negative pressure.
- Lower lobe of the lungs is the most common site of pulmonary involvement accounts for 60% of cases.
- Pulmonary hydatid disease affects the right lung in ~60% of cases. Cysts in the lungs are usually solitary and mostly unilateral, 30% exhibit multiple pulmonary cysts, 20% bilateral cysts.

How do lungs get involved ?

- Larvae can enter the lungs through multiple routes for example via **circulatory system**, which is after ingestion of eggs they reach the liver via the portal circulation. Most of the embryos are stuck in the liver sinusoids but embryos with **diameters <0.3 mm** may pass through the hepatic sinusoids and, through the hepatic vein and the inferior vena cava (IVC) enter the right heart and, finally, settle in the lungs.
- Embryos can also reach the lungs via another route. They enter the **thoracic duct via lymphatics of the small intestine** and then through an internal jugular vein, and right side of the heart, and they enter the lungs.
- Another possibility is **direct pulmonary exposure through the inhalation** of air contaminated with Echinococcus eggs.
- Lungs may also become the site of **secondary hydatidosis** due to the rupture of a primary cyst resulting in dissemination of multiple daughter cysts and scolices.



Hepatopulmonary involvement

- In patients with lung cysts, about ~20–40% also have liver cysts **but most patients with pulmonary hydatid disease do not show liver involvement**. In cases with hepatopulmonary involvement usually is due to Dissemination via a bronchobiliary fistula due to transdiaphragmatic rupture of liver hydatid, and may also follow intrathoracic rupture of a cyst of the liver explaining the lung and liver involvement stimutanously
- Liver cysts should be searched in all cases of pulmonary cysts for two reasons. First, the co-existence of pulmonary and hepatic hydatidosis is high, especially if patients of older age. Second, hepatic cysts can be asymptomatic.

Clinical presentation

- The initial phase of primary infection is asymptomatic and may remain so for many years as **Most intact lung cysts are discovered incidentally on chest radiographs.**
- **Symptomatic hydatid disease** of the lung and complications more often follows rupture of the cyst or compression on adjacent structures.
- So clinical features of E. granulosus infection depend upon the cyst site and size , The growth rate of the hydatid cyst depends on the softness of the organ and surrounding tissue elasticity. as pulmonary cysts typically increase in diameter at a rate of 1–5 cm in a year and some reported a doubling in size in time of 16–20 weeks.
- The majority of children and adolescents with lung lesions are asymptomatic despite having lesions of impressive size, assumedly because of a weaker immune response and the relatively higher elasticity of the lung parenchyma relative to older patients.
- Occasionally, an unruptured cyst results in cough, dyspnoea, haemoptysis, or chest pain. Less frequent symptoms include malaise, nausea and vomiting, and thoracic deformations

Cyst rupture

The cyst may rupture spontaneously or as a result of trauma or secondary infection.

** TYPES OF RUPTURE :

A contained rupture, only the endocyst is torn and the contents of the cyst are contained by the pericyst.

A communicating rupture, the contents of the cyst escape into the tracheobronchial tree through bronchioles that have been incorporated into the pericyst.

A Direct rupture, follows tearing of both the endocyst and the pericyst, with discharge of the contents of the cyst directly into the pleural cavity .

RESULTS OF RUPTURE

- Rupture may be associated with the **sudden onset of cough and fever**. If the contents of the cyst are expelled into the airway, **expectoration** of a clear salty or peppery tasting fluid containing fragments of hydatid membrane and scolices may occur **(hydatoptysis)**
- Also Symptoms can result from the release of antigenic material and secondary immunological reactions that develop following cyst rupture. **Fever and acute hypersensitivity reactions** ranging from urticaria and wheezing to life-threatening anaphylaxis may be the principal manifestations.

Conclusion of Symptoms

Cough either due to bronchial irritation by the growing cyst or by direct rupture of the cyst into the bronchus.

Shortness of breath

Hemoptysis in up to 70% of adult patients, but it is rare in pediatric patients.

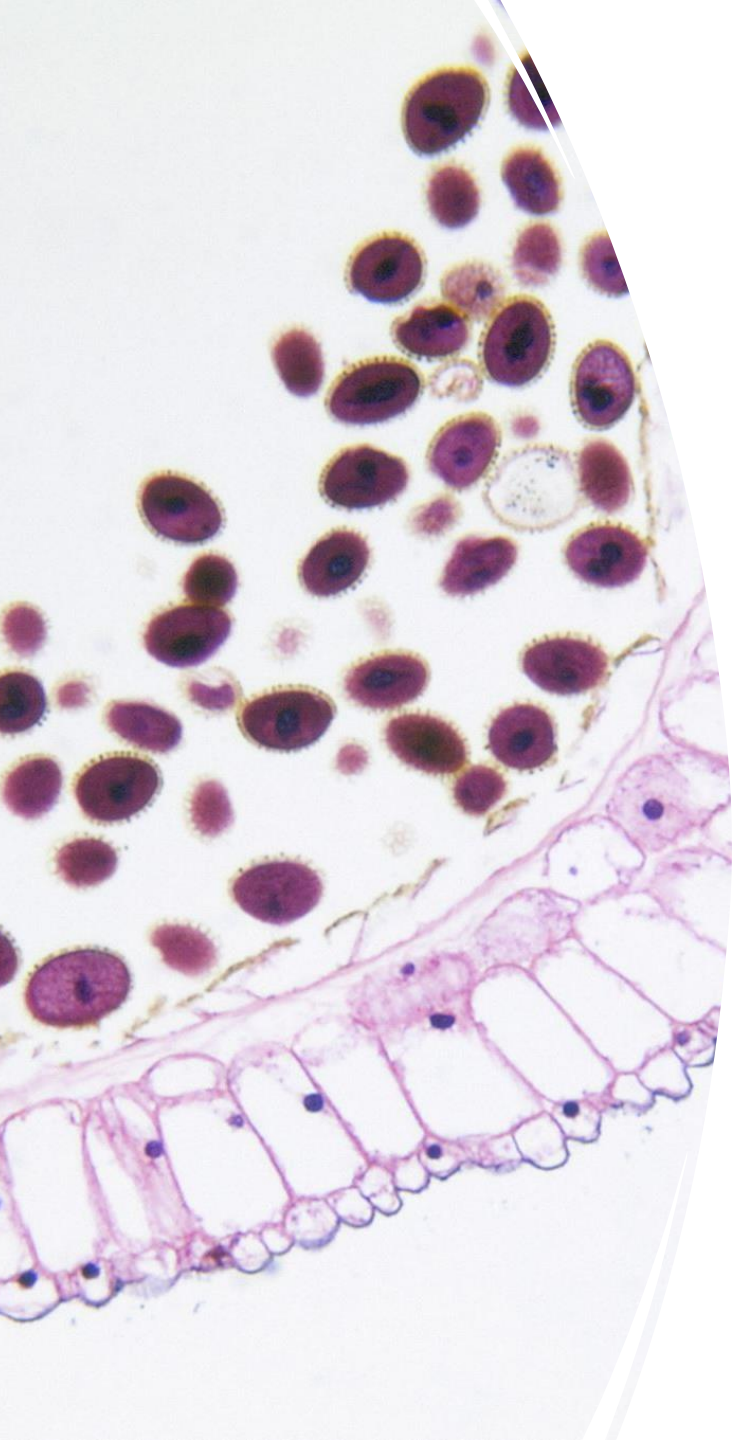
Expectorations(hydatoptysis)

Chest pain

Fever

anaphylactic phenomena

Mediastinal cysts may erode into adjacent structures causing **bone pain, hemorrhage or airflow limitation.**



Complications

- Simple or tension pneumothorax, pleural effusion, or empyema because of cyst rupture into plural cavity.
- Infection of the cyst, manifesting as a pulmonary abscess with poorly defined margins.
- Atelectasis if Lung tissue adjacent to a cyst is compressed.
- Acute pulmonary embolism (rare) This complication may develop after invasion of the cardiovascular system or direct invasion of the inferior vena cava.
- Calcification, which usually requires 5–10 years for development, occurs quite commonly with hepatic cysts but rarely with pulmonary cysts.

Diagnosis

- Contact with cattle and dogs in an endemic area.

- Radiology (chest X-ray ,CT scan, MRI, PET CT) and serology are the principal diagnostic modality used to confirm the diagnosis.

- Peripheral blood eosinophilia is present in <25% of infected persons. Leukocytosis and increased erythrocyte sedimentation rate are also observed. But all are nonspecific.

- Bronchoscopy

Radiological feature

- The classical chest radiography of an intact cyst is a sharply defined, round-to-oval homogenous opacity of variable size .The diameter of the thoracic hydatid cyst varies from 1 to 20 cm.

- Centrally located cyst is smaller compared to peripherally located cyst.

- The cyst may become bizarre shaped by the pressures from adjacent broncho-vascular structures, mediastinum, pleura, and adjacent cysts.

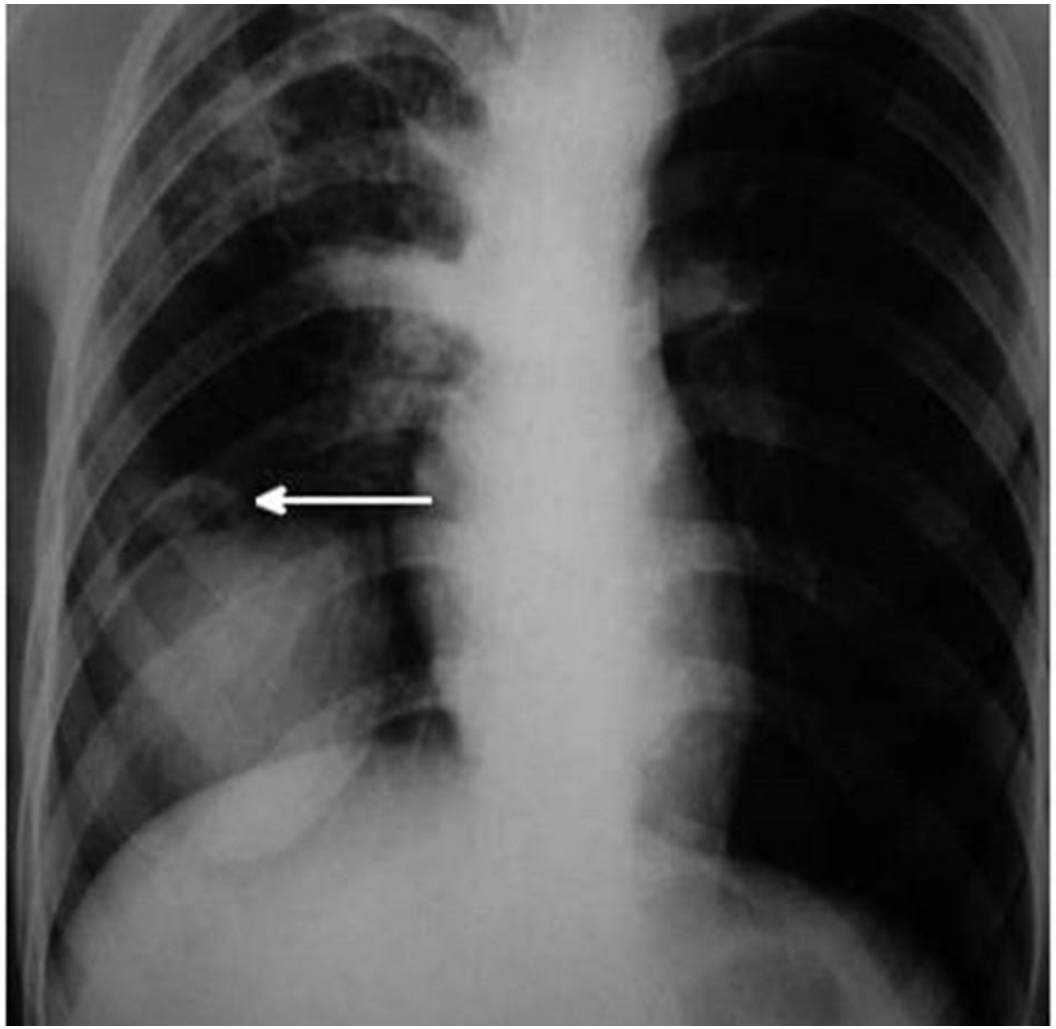
- The shape of the cyst may change radiologically during maximal inspiration and expiration known as the Escudero-Nimerov sign.

Radiological signs of rupture

❖ Air-crescent or meniscus sign

❖ It results in air entry between the pericyst and exocyst layer producing a radiolucent crescent in the upper part of the cyst. It signifies detachment, imminent rupture, and death of the parasite.

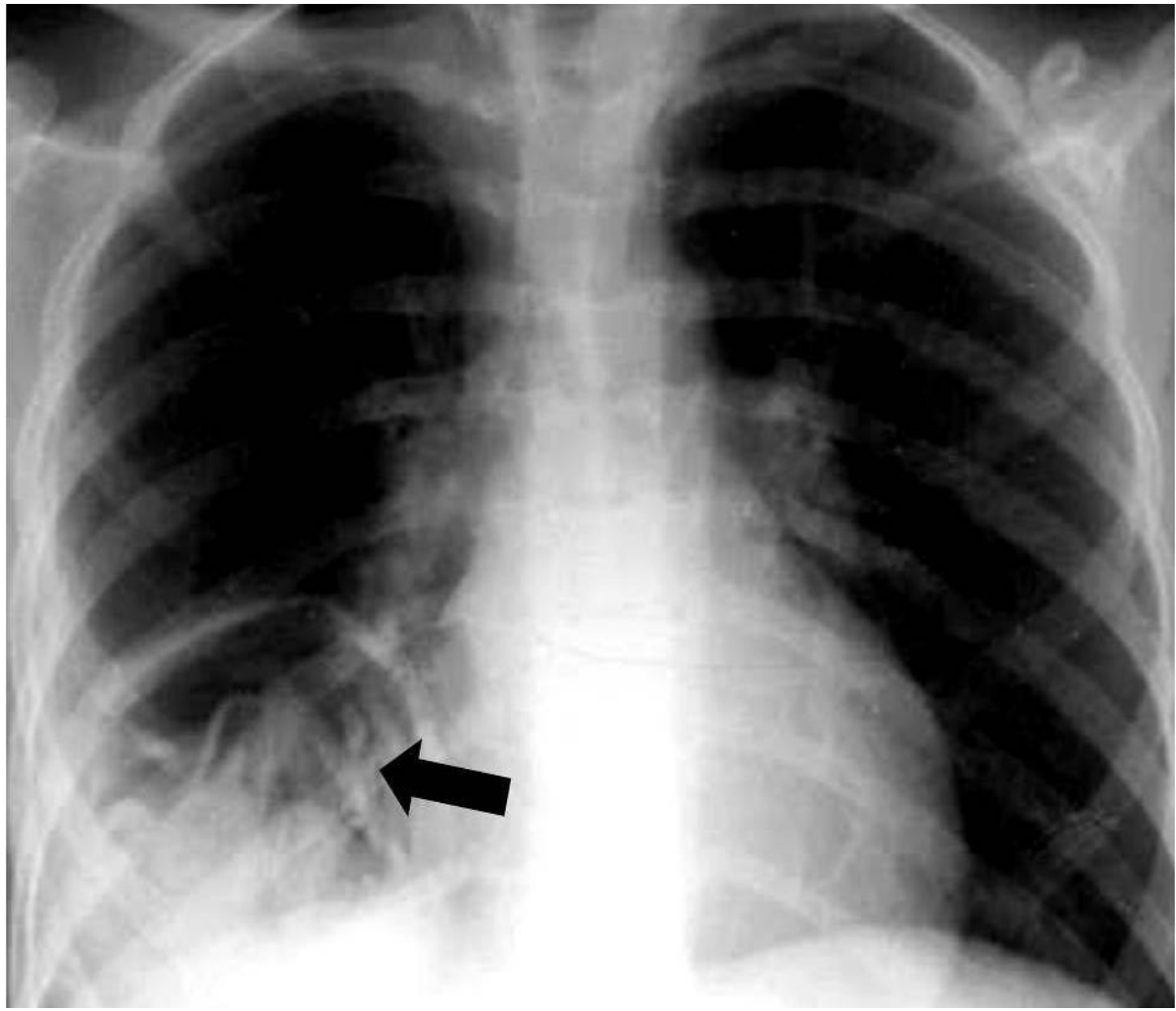
❖ The **"inverse crescent sign"** is opposite to classic crescent sign. Here, the crescent shaped rim of air is present at the lower end of the cyst. It signifies separation of the membranes from the posterior aspect of the cyst without any anterior extension.



Radiological sign of rupture

Water-lily sign or Camelotte sign

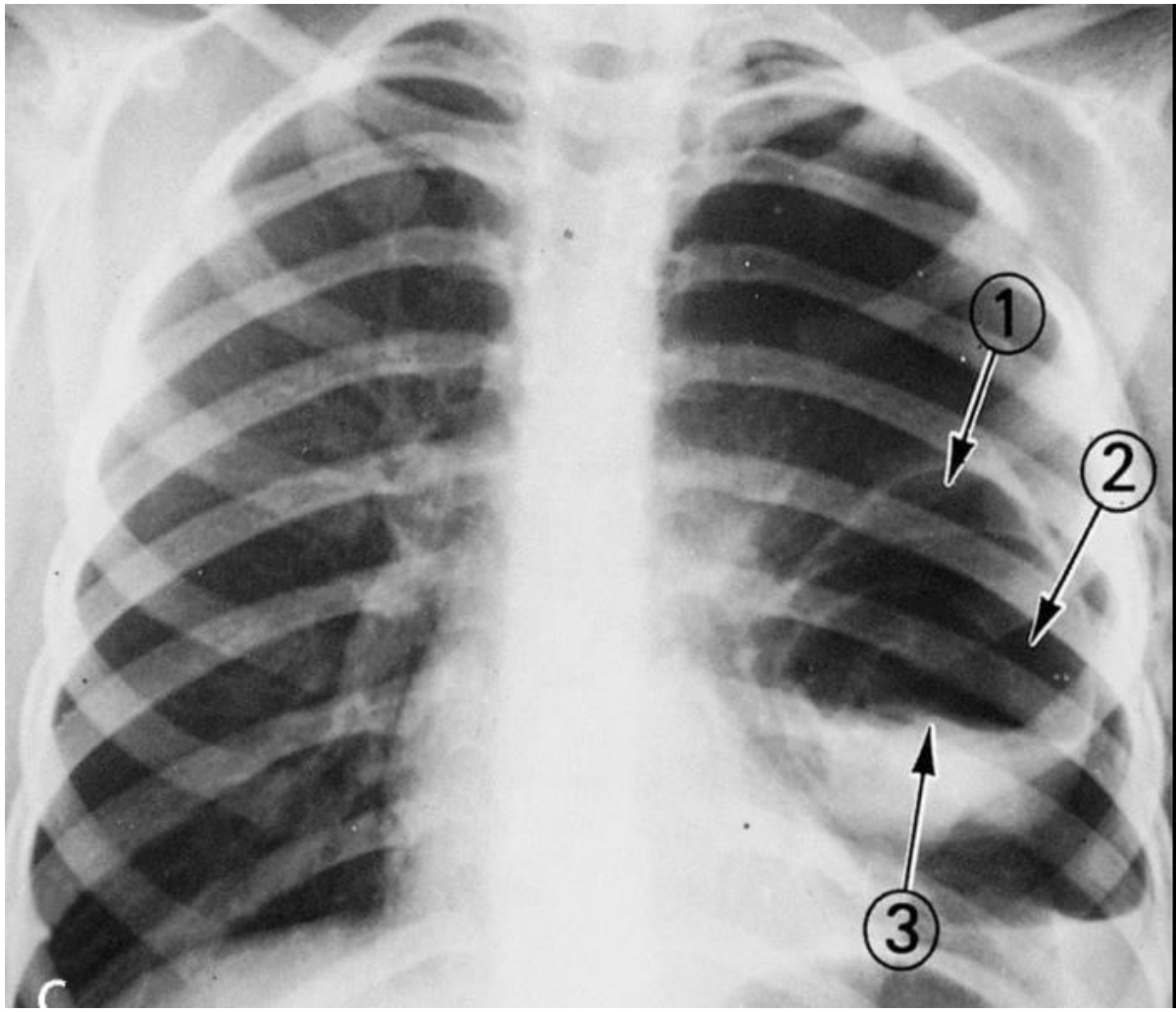
Following ruptures partial evacuation of the cyst fluid occurs, and the collapsed endocyst can be seen floating freely in the cyst fluid. It results in unevenness of the air-fluid interface, the so-called water-lily sign or Camelotte sign. Water-lily sign is seen in only 10-15% of lungs hydatid cysts.



Radiological sign of rupture

Cumbo sign .

This is also known as the onion peel sign or double arch sign. As more air enters between pericyst and endocyst, endocyst rupture. It leads air entry inside endocyst and creation of air-fluid level surrounded by a crescent of air between the pericyst and the endocyst.

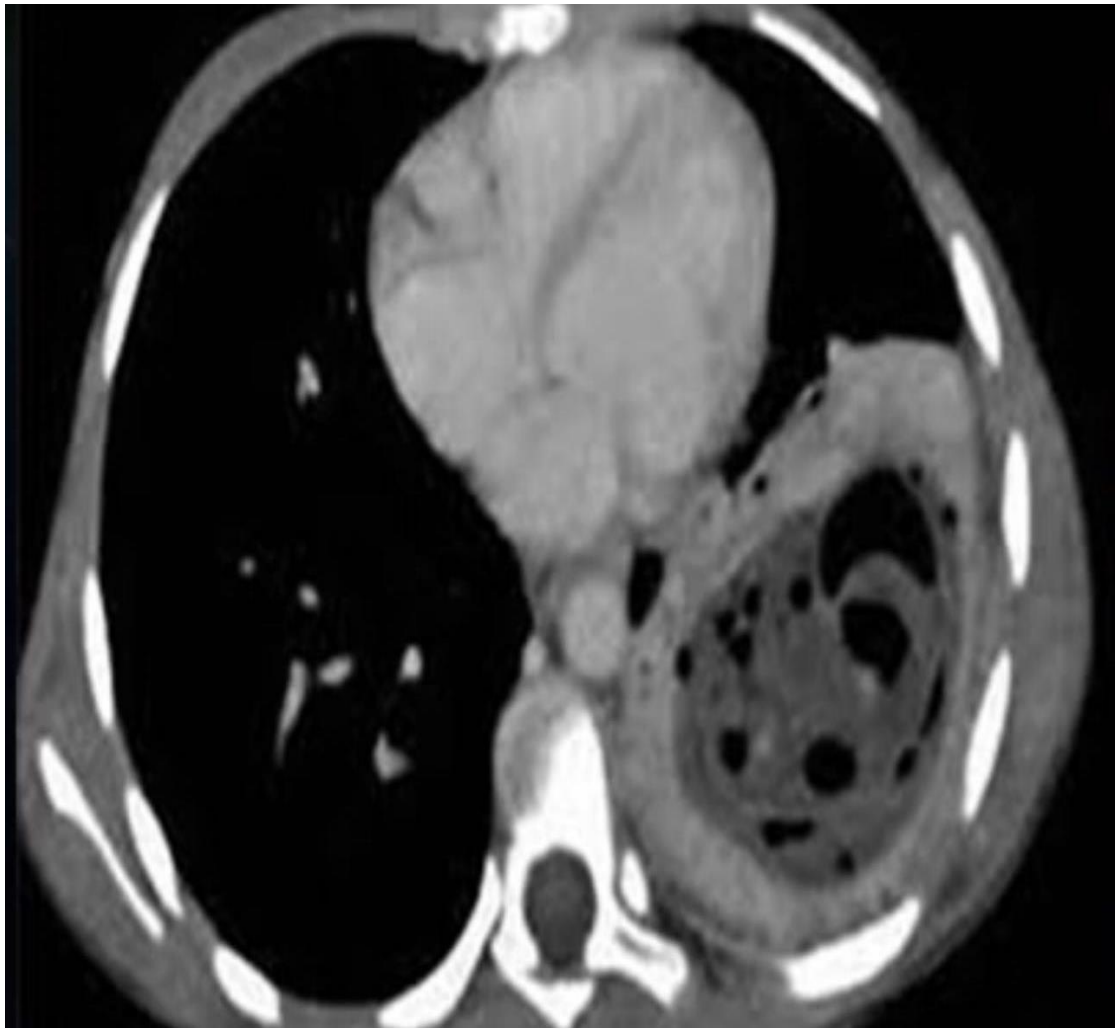


Radiological sign of rupture

Air bubble sign.

- It consists of single or multiple small, rounded radiolucent areas with very sharp margins within solid media or pericystic areas and indicates ruptured infected hydatid cyst. It is best seen in the mediastinal window.

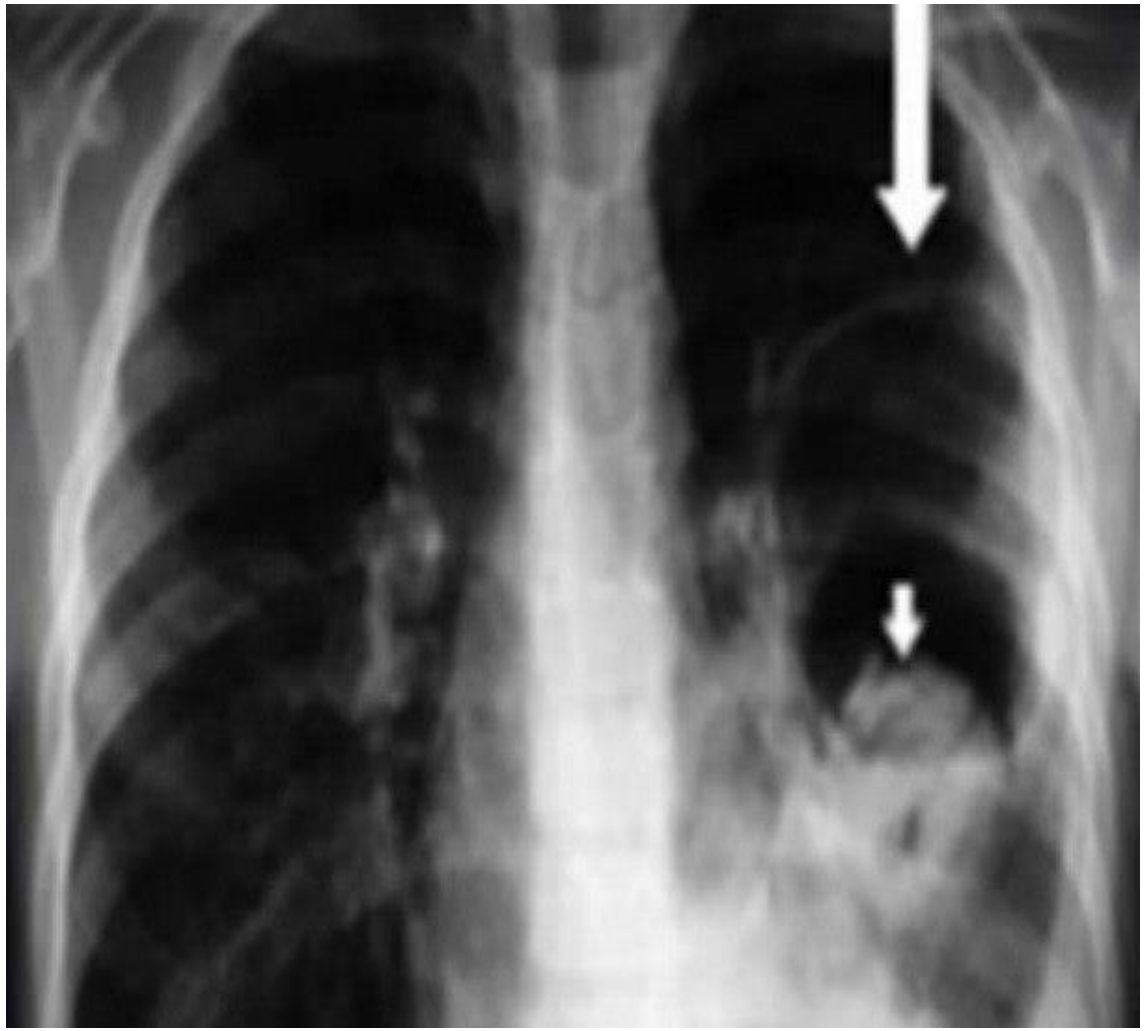
- NB: Thick wall cyst denots infection.



Radiological sign of rupture

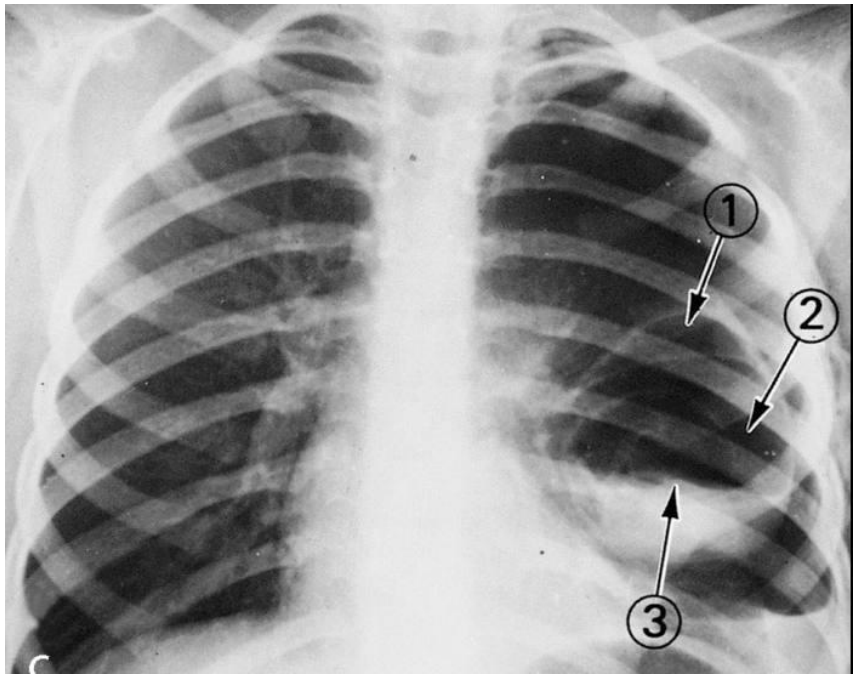
Rising sun sign:

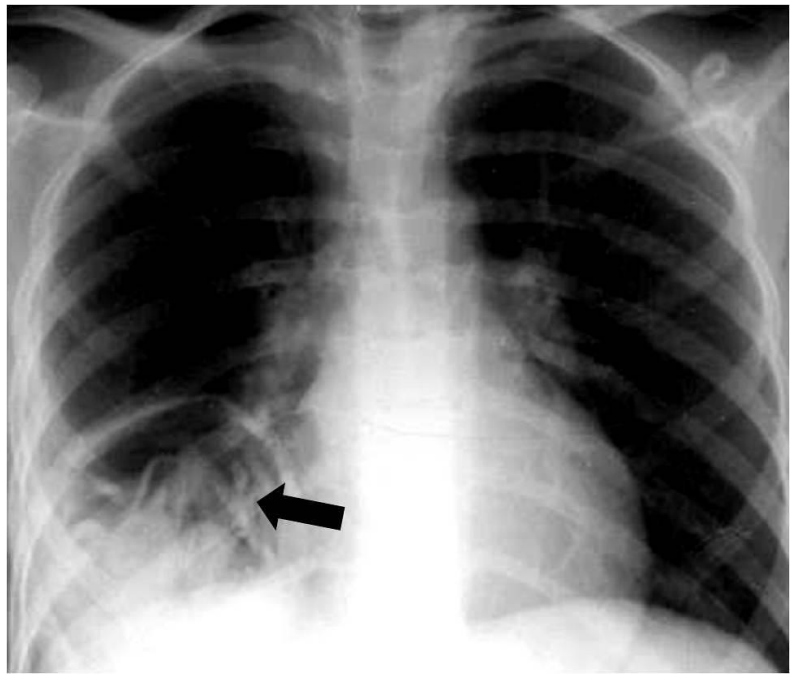
- Daughter cysts are rarely present in the lungs. When present, daughter cysts may appear in the lowest part as circular shadows amidst disintegrated endocyst membrane giving rise to the rising sun sign



The double arch sign(Cumbo)and the sign of the camalote (water-lily)are pathognomonic of a ruptured hydatid cyst.

Unlike extrapulmonary cysts, pulmonary hydatid cysts calcification, and daughter cyst formation are rare.





Serological testing

- **Serological testing is useful in the primary diagnosis of hydatid cysts The majority of the serological tests is based on detecting antibody as it is more sensitive than antigen detection.**

  - **The enzyme-linked immunosorbent assay (ELISA),**
  - **the indirect hemagglutination antibody assay (IHT),**
  - **latex agglutination test, and immunoblot test (IB).**

Other serological based methods include the immunofluorescence antibody test and arc-5 immuno- electrophoresis (IEP).

Casoni test

intradermal skin test had a sensitivity of 70%.

intradermal injection of sterilised fluid from hydatid cysts, a wheal response occurring at the injection site within 20 minutes is considered positive

Management

- 1-pharmacotherapy
- 2- Surgical therapy

Management

Pulmonary hydatid cysts is treated by pharmacotherapy and/or surgery.

Surgical intervention is the treatment of choice though pharmacotherapy may also be useful in selected patients.

pharmacotherapy

Benzimidazol group of drug(anthelmintic drugs):

Albendazol 10-15 mg/kg two times a day

Mebendazol 40-50 mg/kg three times a day

* minimum 3-6 months

Benzimidazoles act by inhibiting microtubular assembly within the parasite, leading to impaired glucose absorption through the wall of the germinative cell layer of the larva. It results in glycogen depletion and degeneration of the endoplasmatic reticulum and mitochondria of the parasite, along with an increase in lysosomes and subsequent autolysis of cells.

When to use pharmacotherapy ?

- Smaller cysts
- recurrent cysts or multiple cyst
- poor surgical risk
- refusal for surgery
- multiorgan disease
- When there is intraoperative spillage of hydatid fluid

Contraindication of pharmacotherapy

- Large cysts that are at risk of rupture
- Inactive or calcified cysts
- Bone marrow depression
- Pregnancy, specifically the first trimester of pregnancy.

Surgical therapy

*It's considered the treatment of choice since the parasite can be completely removed and the patient cured

*The aim of surgery is to remove the cyst completely while preserving the lung tissue as much as possible without allowing intraoperative spillage.

The indications for surgery include large cysts that are likely to rupture, infected cysts, cysts in vital anatomical locations, and cysts exerting substantial mass effect.

Surgical therapy

- The surgical method may be different in the intact (simple) and ruptured(complicated) cysts
- The operation has two steps :
- A) removal of the cyst (germinal layer)
- B)management of the residual pulmonary cavity

Surgical therapy

- During surgery its important to prevent cystic content spillage in order to prevent intraoperative dissemination and eventual recurrence

- Preoperative albendazol is essential

  started 4 days prior the surgery and continue for 1-3 month post.op

The proponent view is that preoperative use of benzimidazoles softens the cysts and reduces the intracystic pressure. It simplifies cysts removal and reduces the risk of recurrences.

Surgical therapy

- Standard posterolateral thoracotomy is the preferred surgical approach. In patients with bilateral cysts, median sternotomy or two-stage thoracotomy can be used
- Surgery for pulmonary hydatid cysts is a safe procedure with a good outcome, low morbidity, and negligible mortality

- finding the cyst : Once the chest is opened , the lung is palpated gently to avoid rupture of the cyst . Deflate the lung on the affected side to make it easier to find it
- Covering the adjacent lung with saline soaked towels : because there is always risk of spillage of daughter vesicles during the operation , only the area of the cyst-containing lung must be in the operating field

Surgical therapy

- **Various surgical techniques available include :**
- Enucleation (without aspiration) consists of removal of the cyst with its intact germinative membrane by a careful dissection between the pericyst and laminated membrane. It is suitable for the small pulmonary hydatid cysts <5 cm in diameter with little risk of rupture, with enucleation, pericyst remains as such; therefore, there is a risk of postoperative air leak and infection.
- Removal of cyst following aspiration: aspiration is performed with a syringe adapted to 3 valved aspiration catheter + 10% povidone iodine injected to the cyst with the same catheter to kill living vesicles after finishing remove the laminating membrane

Surgical therapy

- Pericystectomy involves removal of hydatid cyst along with the pericyst. Pericyst excision increase risk of airway leakage and tension pneumothorax in the early period or bronchopleural fistula in late period

- If the hydatid cyst occupies more than 50% of a lobe ---- lobectomy

➢Management of residual cavity When the cyst is removed :

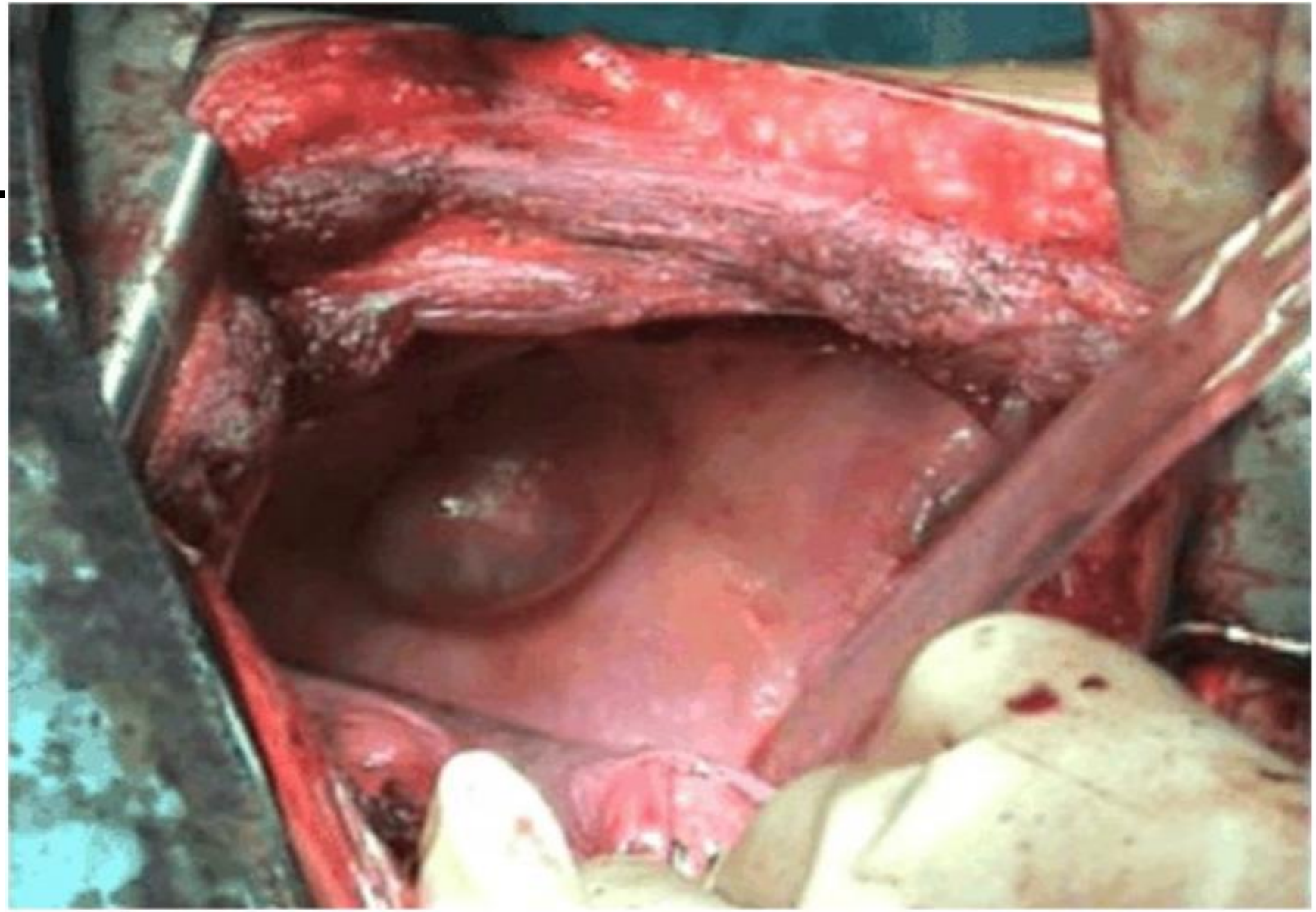
-The pulmonary cavity is cleaned completely with sterile gauze

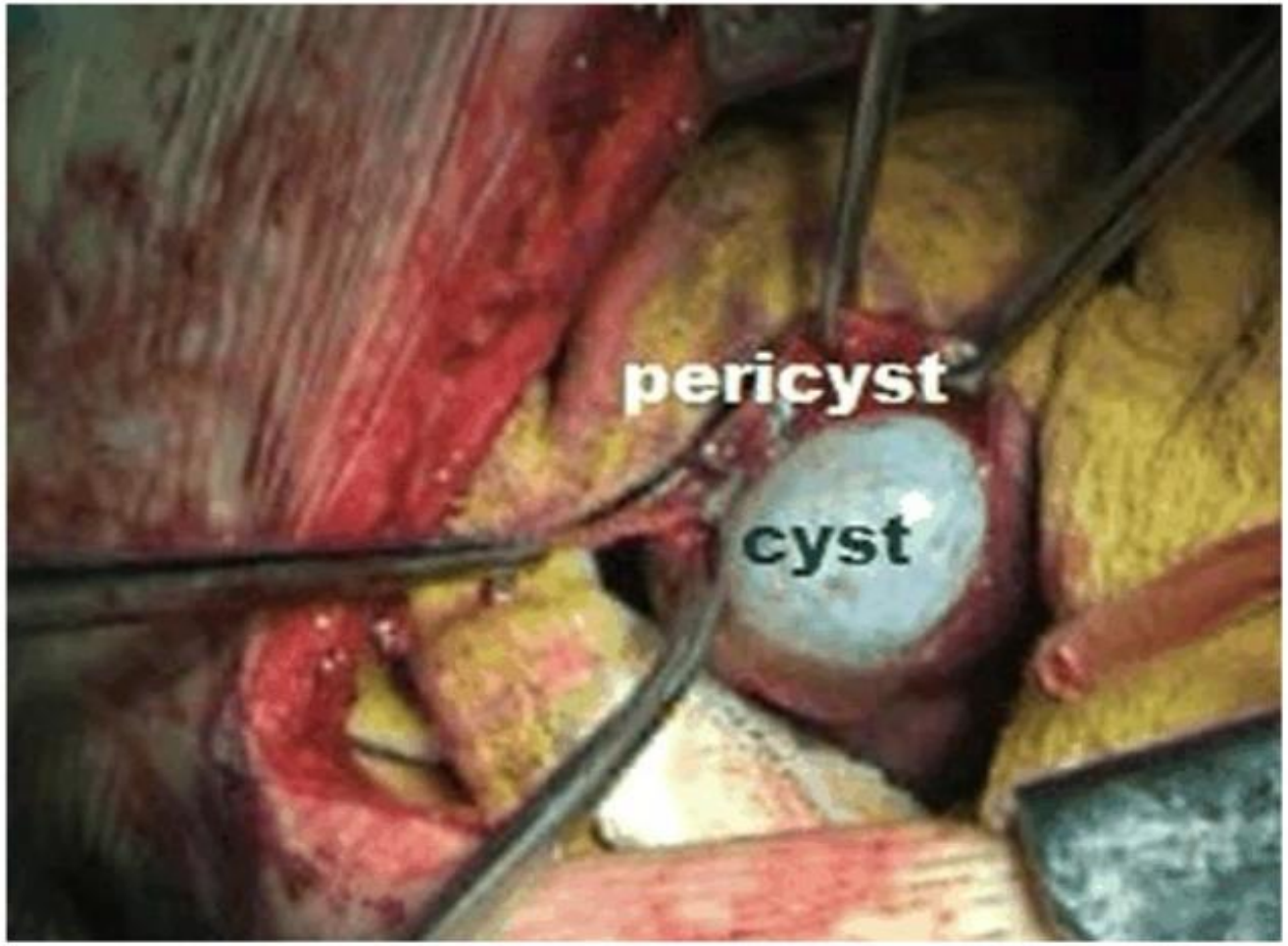
- Observation for presence of air leakage

- Closure of major bronchial opening

- The lung cavity that remains after removal of the cyst may be left as it is or obliterated by sutures (capitonnage is a recognized method, by which the walls of the evacuated pericyst cavity are approximated with multiple sutures, it reduces complications, morbidity and rate of reoperation.) in regard to the size and location of the cyst
- The bronchial openings in the cavity must be closed by sutures in all cases

- Remove the towels by grasping instrument





The macroscopic appearance of the hydatid cyst in the lung

Percutaneous aspiration

percutaneous aspiration is not a routine practice in pulmonary hydatid cyst due to the risk of rupture, dissemination, and anaphylaxis reaction. It is more commonly done in hepatic hydatid cysts.

Postsurgical follow-up

- Percutaneous aspiration of pulmonary hydatid cyst has been reported in literature.

- Postoperative all patients should receive ABZ (10 mg/kg per day) for 6 months to prevent recurrence of the disease. The recurrence rate of pulmonary hydatid cyst without postoperative antihelminthic therapy is as high as 11%.

Postsurgical follow-up

- Liver function tests and chest radiography should be done monthly for the first 3 months. The follow-up then continued every 3 months by chest radiography until the end of postoperative year 1.

Thank You