Hydatid means cyst full of water

Epidemiology:
Domestic hygiene, standard of living & human behavior play important role in epidemiology.

Life cycle of parasite:
- Hydatid disease is caused by larval stage of Echinococcus granulosus.
- The adult worm is a cestode (tape worm) 5-10 cm in length with a scolex bearing 4 suckers with the body containing 2-6 proglottids
- The larval stage or meta-cestode is a cyst with an acellular laminated outer layer and an inner nucleated germinial layer which gives rise to broad capsule and then scolices.
- The adult worm lives in dogs which is the primary host
- The cystic part of the cycle occur in sheep, pigs, camel, or man which act intermediate host
- The adult worm live in the intestine of dogs for months. The terminal segment proglottidis detach from the worm and release thousands of ova with the stool. These are ovoid in shape and can remain infective for long time.
- Ingestion of the ova by sheep in contaminated water or grass or vegetables lead to infection and hydatid cyst subsequently develop in the viscera
- The parasite goes back into the dog when the sheep is slaughtered and its viscera containing the hydatid cyst are eaten by another dog.
- In each of these cysts, thousands of scolices are found which will grow into new tape worms in the dog's intestine and so the cycle repeats itself indefinitely.
- If a worm gets into a man instead of sheep, the life cycle of the parasite will be broken (because entrails of man are not eaten by dog)
- Contamination of the food is the common source of infection in man, more frequently he contaminates his fingers by fondling a dog and then takes his food without washing his hands
- The ova hatches in the stomach → hexacanth embryo or oncospheres are released in the stomach and duodenum → pass through the intestinal wall → travel via he portal vein → liver where the majority settle and develop hydatid cyst.
- Only those embryo which successfully pass through the capillaries of the liver, reach the lung and develop into hydatid cyst and few which get through the lung are then carried to the viscera and tissues supplied by the systemic circulation
- Sometimes the hatched embryo pass through the wall of the stomach or duodenum into the lymphatic channel → via thoracic duct → lung (bypass the liver)
- Once implanted in the interstitial tissue of the lung, the embryo grows into a vesicle and then into a hydatid cyst.
Pathology:
Hydatid cyst in the lung is surrounded by a capsule formed of atelectatic alveoli & fibrous tissue (pericyst). This adventitia is part of the host & becomes thicker with age thus separating the cyst from the lung parenchyma.

- **Hydatid cyst itself is composed of:** (layers)
  1. **Ectocyst:** (the laminated membrane) formed by the germinal layer. It is an acellular white laminated membrane and completely separated from the adventitia.
  2. **Endocyst:** (the germinal layer) it is the only living part of the cyst which secretes:
     a. Internally: hydatid fluid
     b. Externally: it forms the laminated membrane
  3. **Broad capsule:** develops from the germinal layer by the process of asexual budding & vacuolation and are attached by pedicles to the inner most wall of the germinal layer
    - With each broad capsule 2-3 scolices (head of future worm) develop
    - The attachment of the broad capsule to the germinal layer becomes loose and the capsule bursts releasing the scolices into the fluid of the cyst
    - These fall to the bottom by gravity and sometimes are called hydatid sand
    - The main bulk of the cyst is made of hydatid fluid which is highly antigenic
    - Scolecis may form daughter cysts by retrograde metamorphosis.

- **Simple hydatid cyst:** is an intact cyst
- **Complicated cyst:** is an injured, ruptured or infected.
  Rupture into the bronchial tree is common while into the pleura is very rare.

- **Partial rupture:** → adventitia bursts → small bronchial fistula appear → air enters the space between the laminated membrane & the adventitia → radiological appearance as a crescent shaped translucency in the upper part of the hydatid (called perivesicular pneumocyst or signet ring appearance)

- **Complete rupture:** → cyst itself rupture and small amount of fluid escape between the laminated membrane and the adventitia. And at the same time a little air gets in through the small bronchial fistula → fluid level appear on either side of the shrunken but dome shaped hydatid → radiological appearance of "sun set or rising sun sign". or may give appearance of "double arc sign".
  - Complete rupture may permit expulsion of all the contents through large bronchial fistula and allow air to enter the cyst → laminated membrane collapsed and floats upon the remaining of the fluid → irregular projection over the level of the fluid → radiological appearance of "water lily sign"
  - **2 complication of water lily cavity** (cavity with retained disintegrated membrane):
    o **Daughter cyst develop** from surviving part of germinal membrane
    o **Lung abscess:** parasite die after complete rupture and retained disintegrated fragments of the laminated membrane act as sequestrum and with the associated bronchial fistula lead to infection and lung abscess formation
  - Occasionally all the membranes are coughed out leaving a **pulmonary cavity** and a cured patient
Rarely *calcification* occurs in the adventitia of complicated cyst but this doesn't signify death of the parasite

Simple cyst develop from an ovum

**Secondary hydatid cyst** means that the primary cyst lodged in the lung or liver has ruptured and caused new daughter cysts to develop

**Daughter cyst develop from either :**

- fragmentation of the germinal layer left behind after inadequate surgical or spontaneous evacuation.
- From scolices in the hydatid fluid which has been spelt at surgery or after spontaneous rupture.
- Live hydatid element (scolecis) could revert to new hydatid cyst by retrograde metamorphosis.

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**Clinical presentation:**

- Incidence is increased in children who fondle dogs and in young men who work with dogs in farms.
- There is no significant difference in sex distribution.

1) The lesion may be **asymptomatic** unless complications develop. The patient may present to the thoracic surgical department with incidental CXR finding of well circumscribed opacity.

2) **Patient with intact or mildly complicated cyst** may experience transient symptoms of pain, cough, and hemoptysis.

3) The acute onset of rigor, fever and cough with purulent exudation and pleuritic pain is experienced with all patients with **lung abscess**.

4) Fragmentation of the membrane or daughter cysts may be described by the patient as "**grape skin**" which may be coughed out when the cyst ruptures into the bronchial tree.

5) Severs dyspnea may occur in rare events of **rupture into the pleural space** → hydropneumothorax or pyopneumothorax.

6) Rupture of the cyst may cause signs of **anaphylaxis** (due to its high antigenicity).
Diagnosis:

I- **CXR:**

1. Well defined circular or oblong shaped opacity of homogenous density is diagnostic of simple intact hydatid cyst.
2. A pathognomonic X-ray finding is perivesicular pneumocyst due to presence of air between the adventitia and laminated membrane so appears as a slender crescent "signet ring" in case of adventitial rupture.
3. "Double arc sign" appears as a cyst with fluid level on either sided of the dome shaped cyst.
4. "Water lily sign" or "Camelot sign": cyst with collapsed membrane floating in the fluid seen in cases of rupture of the cyst itself.
5. Daughter cysts: may produce radiological appearance identical to primary cyst. Large daughter cysts with partially or completely drained pericyst → "rising sun sign".
6. If suppuration occurs → huge abscess with air fluid level.
7. Often an empty cavity with remnants of collapsed membrane are seen.
8. Rarely all the membranes are coughed up & a pulmonary cavity is seen surrounded by thin margin.
9. Rarely, the adventitia calcify → thin shell of egg "egg shell sign".
10. Hydropneumothorax is seen (due to rupture into pleural cavity).
11. Bilateral multiple cysts is a common X-ray finding.

II- **Ultrasound of abdomen.**

III- **CT scan:**

IV- **Isotope liver scan**

V- **Immunological tests:**

1) **Casoni intradermal test:** unreliable because of false positive in unaffected individuals in endemic areas and may remain positive long time after the cyst has been removed. Also it is difficult to obtain the test fluid.
2) **Weinberg CFT:** best method and it is accurate.
3) **Latex slide agglutination test:** simple, reliable and specific.
4) **Indirect hemagglutination test:** non-specific.
5) **Immunoelectrophoresis:** detects the specific bands of Echinococcus granulosus.

VI- **Blood tests:**

Eosinophilia is common but of little value because of multiple parasitic infection in the middle east.

VII- **Parasitological tests:**

depend on finding the scolices, broad capsule, and daughter cyst in the sputum or pleural aspirate.

**Differential diagnosis:** from other forms of solitary round peripheral lesion

a. Malignancy
b. Other causes of lung abscess
Treatment:

A) Surgical Treatment: Treatment of hydatid cyst is surgical removal

I- Removal of the cyst:
The principles of hydatid cyst removal are:
   a) Evacuation of the contents of the cyst
   b) Avoidance of spillage during the procedure
   c) Closure if the bronchial fistula
   d) Prompt re-expansion of the lung

1. Aspiration / Evacuation technique:
   ▪ Aspiration of the fluid with a fine needle followed by extraction of the cyst through a small adventitial incision
   ▪ It can be used for simple and complicated cyst
   ▪ Aspiration may result in some leakage but this will not be dangerous if adequate precautionary packing of the operative field has been carried out

2. Enucleation:
   ▪ Enucleation of the cyst in a plane between the adventitia and the laminated membrane
   ▪ Technically it is a beautiful procedure, applicable only to simple intact cysts
   ▪ Although the plane of cleavage can readily be found between the adventitia and laminated membrane, there is an appreciable risk of accidental rupture during dissection. If this does occur the risk of contamination is high much greater than aspiration/evacuation technique

3. Excision:
   ▪ Excision of the cyst with its capsule
   ▪ It is applicable to all cases irrespective of whether the cyst is simple or complicated and is often the best chance of avoiding soiling
   ▪ It appears sound, simple and attractive but continuous oozing of blood and persistent leakage of air from the raw surface of the lung may render it unsuitable

After removal of the cyst by whatever method:
   o The edge of the sac must be trimmed and sutured
   o Meticulous search for all bronchial openings which should be sutured individually
   o The residual sac is left open and no need for obliterating it

II- Resection:

Indications:
1) Giant simple cysts that causes permanent & irreversible changes in the affected part of the lung
2) Complicated cyst where the lung infection has caused a the whole lobe to become a huge lung abscess or bronchiectatic
3) Multiple daughter cysts involving one lobe
4) Calcified cyst (?)
5) Patient presenting a diagnostic problem (ca can not be excluded)
6) Massive hemoptysis
7) Recurrent suppuration following removal of the cyst
Resection options include:
A. Segmentectomy: the usual procedure, its aim is to preserve as much lung tissue as possible
B. Lobectomy: procedure of choice when resection is indicated
C. Pneumonectomy: rarely needed

❖ Bilateral disease (bilateral simple intact cyst) → side with the large simple cyst is operated 1st and after few weeks the other side is tackled.
❖ Lung and liver disease → priority is for pulmonary lesion
❖ Resent rupture into pulmonary tree should be treated conservatively by:
  1. Treatment of anaphylactic shock
  2. Antibiotics
  3. Postural drainage
  4. Regular observation for signs of recurrence
❖ Rupture into pleural cavity with hydropneumothorax → thoracotomy.
  Main objectives are:
  • Removal of the cyst which is found floating in the pleural cavity
  &  • Re-expansion of the lung
❖ Even in patient with pyopneumothorax and daughter cyst formation → same treatment indicated.

B ) Medical Treatment :
• It is far from being established. No drugs have yet been found that can effectively reach and destroy the parasite. Even if drugs capable of destroying the live contents of the cyst become available, the retained membrane will ultimately become infected and produce a lung abscess.
• Mebendazole and fluoromebendazole (vermox, fluvermol) & Albendazole are benzinidazole derivatives and have limited role in treatment. They are safe with few side effects

  Indications:
  1) Disseminated cases when the cysts are multiple and inaccessible
  2) Surgery contraindicated for reasons of extreme ill health

Mebendazole 50mg/Kg/day in divided doses
Albendazole is given 100 mg X 2

Control of disease:
❖ Population needs to be informed of the importance of washing their hands after handling dogs and before meals
❖ Dogs should be kept outside the house
❖ Domestic dog must be examined and de-wormed by praziquantel and they should be denied access to raw offal at farms and slaughter houses
❖ All stray dogs in endemic areas should be destroyed