Cystic diseases of Liver constitute a large percentage of the liver space occupying lesion. Of them Hydatid disease is not uncommon. Though there is no exact data about the incidence and prevalence of Hydatid disease in Bangladesh but Hydatid disease is not uncommon in our day to day practice. Most of the patients come to the Physician with pain or dull ache in the upper abdomen for prolonged period without any weight loss or fever. Some times patients may present with lump in the upper abdomen or as incidental finding for investigations for other diseases.

This disease is due to the larval or cystic stage of infection with the tapeworm, Echinococcus granulosus. Echinococcus granulosus is a small tape worm of 5mm in length with a head region known as scolex and 3 segments.

Dogs are definitive host. Sheep, cattle, camel and horses are usually intermediate host. Dogs become infected with this worm by eating the viscera of sheep. Man is an accidental intermediate host in whom usually there occur a dead cycle.

E. granulosus usually produces unicellular cystic lesions and is prevalent where livestock is raised in association with dogs. This tape worm species is found in Australia, Chile, Argentina, Africa, Eastern Europe, the middle east, New Zealand and Mediterranean region particularly Lebanon and Greece. Other Ecchinococcus like E. multilocularis is found in alpine, Arctic or Subarctic regions including Canada, USA, Asia , Central and Northern Europe. This Echinococcus causes multilocular alveolar lesions that are locally invasive which usually use mice and other rodents as their intermediate host. The life cycle of E. multilocularis is similar except that small rodents serve as the intermediate hosts. The cyst of E. multilocularis however is quite different in that larval form remain in proliferative phase and the cyst is always multilocular and the vesicles invade the host tissues by peripheral extension of process from the germinal layer.

E. Vogeli causes polycystic hydatid disease and is found only in Central and South America. Hydatid cyst contains the protoscolices, proto scolices are released from the cyst into the gut of dog. Scolices get adhere to the wall of the small intestine (Jejunum) and become adult worm (5-20 months). The last segment of the adult worm is gravid. Each adult worm sheds about 500 ova into the small intestine of dog which excretes out along with feaces. The infected feaces contaminate the grass and firm land and also the surface of its own body. Ingesting the grass or contaminated water infects sheep, pig, camel and cattle. Man becomes infected by eating contaminated water, green leaves or, vegetables or accidentally eating ova from the surface of the contaminated pet dogs but never from eating meat.

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**Fig:** Viable Protoscolies (microscopic examination of cyst fluid)-Courtesy by WHO bulletin
Development of Hydatid cyst
The ova adhere to the surface of the dogs or present in contaminated water; green vegetables are the source of infections in man. This ova has chitinous coats which are dissolved by gastric juice. The released ovum burrows through the intestinal mucosa and enters into the portal circulation to reach the liver and other organs. Most (65%) of the cyst developed in the liver, some in the lungs, few in other organs like spleen, brain and bones.

Hydatid cyst in man occur from eating ova of the worm never from raw meat containing the cyst. The adult cyst in the tissue can initiate a cellular response. Wall of the cyst consist of three zones 1) A peripheral zone of fibroblasts which derive from the host tissues and become the adventitia or Ectocyst, that may ultimately undergo calcification. 2) The intermediate zone consist of endothelial cells. 3) Inner zones consist of round cells and eosinophils. The intermediate and inner zones become hyalinized (laminated layer). Finally the inner surface of the cyst become lined with germinal layer that has the capability to multiply and give rise to the development of brood capsules. This brood capsules are invaginations from the germinal layer into the cyst. Scolices lie within these invaginations. The attachment of the brood capsules to the germinal layer becomes progressively thinner. At times the capsules burst and release the Scolices in the cyst fluid, that give rise to the term hydatid sand which change its positions with change of posture of the patients.

Daughter and grand daughter cyst may develop from germinal layer. Cyst fluid is a transudate of serum. Protein of cyst is antigenic and leakage of the fluid cause production of anti-Echinococcal antibody. Cystic fluid released into the circulations causes eosinophilia, urticaria and anaphylactic shock.

Sometimes the cyst may present with complications:
1. Rupture may lead to multiple cyst formations, intestinal obstructions and gross abdominal distensions. Cyst rupture into the bile ducts may lead to cure or cholestatic jaundice with recurrent cholangitis. Rupture in the colon leads to elimination per rectum and secondary infection. Rupture into the lungs may lead to expectoration of scolices. Rupture into or pressure over the hepatic veins may give rise to Budd-Chiari Syndrome.
2. Infections of the cyst by pyogenic organisms may occur from rupture into the biliary passage or colon. This will transform the cyst into a pyogenic abscess. The parasite dies within the abscess. Occasionally the cyst undergoes aseptic necrosis and the parasite dies. Here the cystic fluid becomes amorphous yellowish, which should be distinguished from pus of liver abscess.
3. Other organ involvement: Cyst may also occur in lung, kidney, spleen, brain or bone. If hydatid cyst is found elsewhere, there is always concomitant infestations of the liver.
4. Allergy: Cystic fluid contains specific antigen which is a foreign protein that sensitizes the hosts and may lead to anaphylactic shock but more commonly urticaria or hives. This fluid also sensitizes the patient with the productions of antibody. This antibodies are the basis of serological test.
5. Membranous glomerulonephritis may occur from Echinococcal antigen.
6. Bone involvement may cause pathological fracture.
7. CNS involvement may give rise to SOL like lesion
8. Heart involvement cause conducting defect and pericarditis

Clinical features depend upon the site, size and stage of the cyst whether alive or dead. The liver may become enlarged not only due to the presence of cyst but the rest of the liver also hypertrophies. Uncomplicated hydatid cyst may remain silent till death. The only complaints may be a dull ache in the right upper quadrant and sometimes a feeling of abdominal swelling.

Diagnosis
Diagnosis is done by history, clinical examination routine investigation, serological test and imaging. Routine blood test shows eosinophilia in 30% cases of Echinococcosis.

Serology
Cystic fluid of Echinococcosis is antigenic and leak of this cyst induce the production of specific antibodies that are the basis of
serological test. A cyst that never leaks will always give negative result.
1. IHA test for Echinococcus 2. Echinococcal antibody detection by ELISA are positive in 85% cases. Both tests may give false (+) ve or false (-) ve result. Cason's test and CFT for Echinococcus are not well practice now a day.

Imaging
1. Radiology may shows calcification of the cyst, raised poorly moving right dome, hepatomegaly 2. Ultra sonogram and CT scan are very sensitive test for diagnosis. By USG, WHO classify Hydatid cyst into active, transitional and inactive cyst.

WHO-IWGE classifications of USG images of cystic Echinococcosis cysts
1. CL (Cystic lesion) : Unilocular cystic lesions with anechoic (Echo free) content without any limiting membrane that is cyst wall is not visible. Usually indicate active cyst but are not fertile in the initial stage. USG does not detect any pathognomic sign. Differential diagnosis require further diagnostic techniques.

![Cystic Lesion (CL) -Courtesy by WHO bulletin](image)

2. CE1 (Cystic Echinococcosis 1) : Unilocular simple cyst with uniform anechoic content with hydatid sand at the floor or may shows fine echoes due to shifting of brood capsule/Hydatid sand in the floor (snow flake sign). Cyst wall is visible. Active cyst usually fertile. Ultrasonographically pathognomic signs are visible cyst wall and snow flake sign.

![Cystic Echinococcosis 1 (CE1-active)-Courtesy by WHO bulletin](image)

3. CE2 (Cystic Echinococcosis2) : Multivesicular, multisepatated cysts. Cystic septations produce ‘Wheel like’ structures and presence of daughter cysts are indicated by’ rosette like’ or’ honey-comb like’ structure. Daughter cysts may partly or completely fill the unilocular mother cyst. Cyst wall normally visible. Cyst is active and fertile. USG features are pathognomic.

![Cystic Echinococcosis 2 (CE2-active)-Courtesy by WHO bulletin](image)

4. CE3 (Cystic Echinococcosis 3) : Unilocular cyst which contains daughter cysts. Echo free content with detached laminated membrane from cyst wall give rise to the’ water lily' sign, which indicates many detached membrane floating in the cystic fluid. In the earlier stages the cyst is round in appearance but at this stage the cyst is less round due to decrease in the intra-cystic fluid pressure. It is the transitional stage of the cyst which may degenerate or give rise to further daughter cyst. USG features are pathognomic.

![Cystic Echinococcosis 3 (CE3-Transitional)-Courtesy by WHO bulletin](image)

5. CE4 (Cystic Echinococcosis 4) : Cyst contains mixed hypo and hyperechoic degenerative contents. No daughter cysts. Contents may give' ball of wool' sign which is indicative of degenerating membranes. The cyst is inactive and infertile. USG features are not diagnostic and diagnosis need further investigations.
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6. CE5 (Cystic Echinococcosis 5) : Cyst is characterized by Arch like thick calcified wall producing cone shaped shadow. Calcifications may occur partially or completely. Cyst is inactive and not fertile in most cases. USG features are not pathognomic but highly suggestive for E. granulosus. All the cyst type can be divided as small (less than 5 cm) medium (5-10 cm) and large (more than 10 cm).

Treatment and prognosis
Uncomplicated hydatid cyst has good prognosis but risk is always present. Complications like rupture into intrapleural and intraperitoneal space. On the other hand rupture into biliary tree cause spontaneous cure following biliary colic. Abscess formation or infection is controlled by antibiotics. Medical treatment consist of drugs and minor invasive therapy. Drugs like Albendazole is the treatment of choice. 15 mg/ kg/ day in two divided doses for 12 weeks to 6 months. Medical therapy with Albendazole results in cure 30% of cases and improvement is 50%. Many of the failures are subsequently treated with additional courses of medical therapy. Minimum invasive therapy include- PAIR/ PAIRD/ PPDC PAIR-Puncture Aspiration Injection Re-aspiration PAIRD-Puncture Aspiration Injection Re-aspiration Drainage PPDC-Puncture Aspiration Drainage Curettage These therapy can be used in primary case or failed medical therapy case or an alternate to surgery. Scolicidal agent used are hypertonic saline (20%) or absolute alcohol. Response to treatment is best assessed by serial imaging.

Surgical treatment
Cysts are excised and washed with Silver Nitrate solution

References