HYDATID CYST (ECHINOCOCCOSIS)

Ashraf F. Al-Faouri, MD, MRCS, FACS

General & HBPS & Liver transplantation

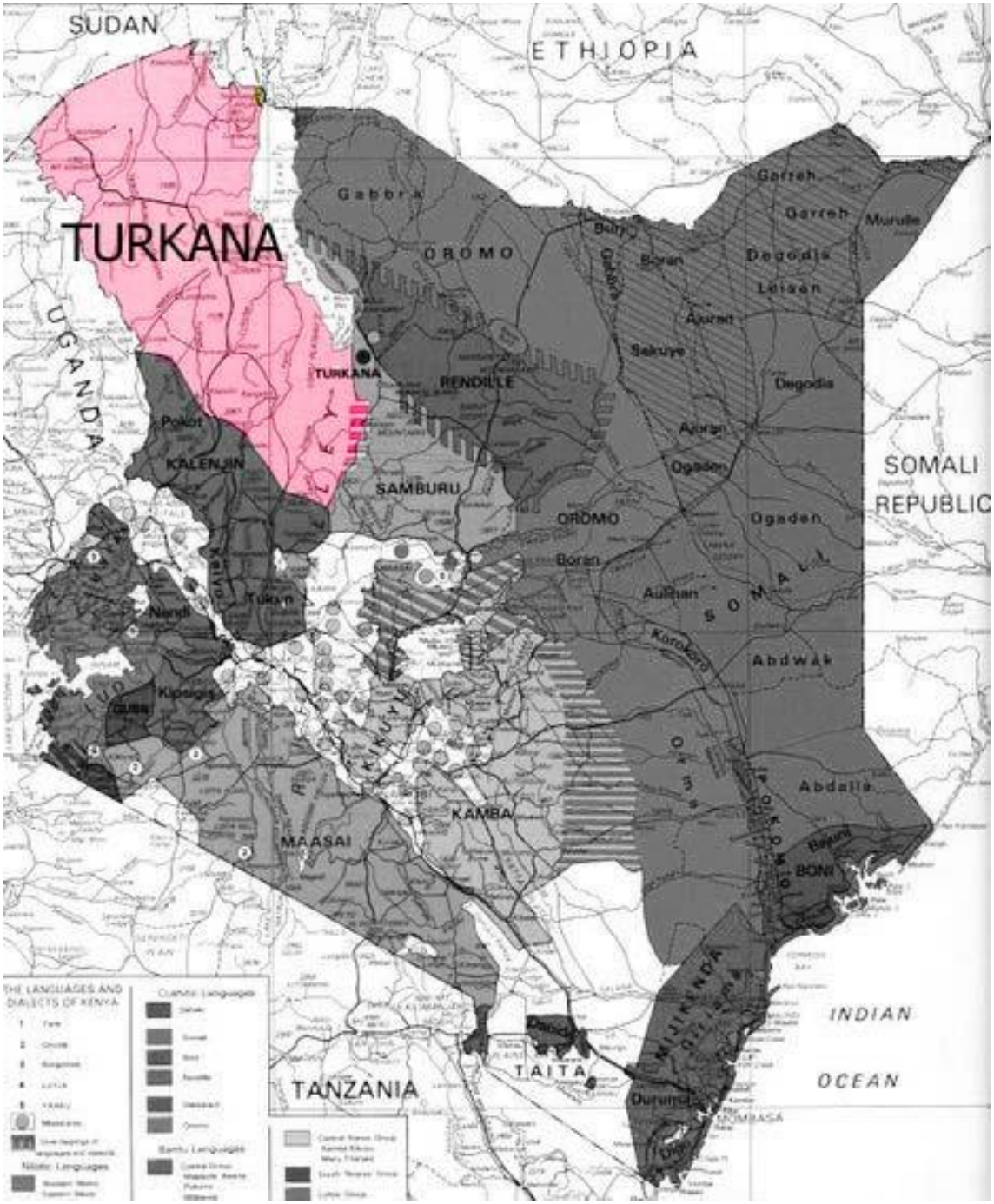
Dressed In A Surgeon Gown A Physician Might Make Progress & Should Succeed In Management

Definition

- Zoonotic infestation caused by the genus Ecchinococcus (tape worm)
- 16 species & 13 subspecies have been described
- Only 4 are clinically relevant

Incidence & Epidemiology

- 0.4/100,000 in Switzerland and Wales
- 196 in Turkana in Kenya


SUDAN
ETHIOPIA
TURKANA
UGANDA
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RENDILLE
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SAMBURU
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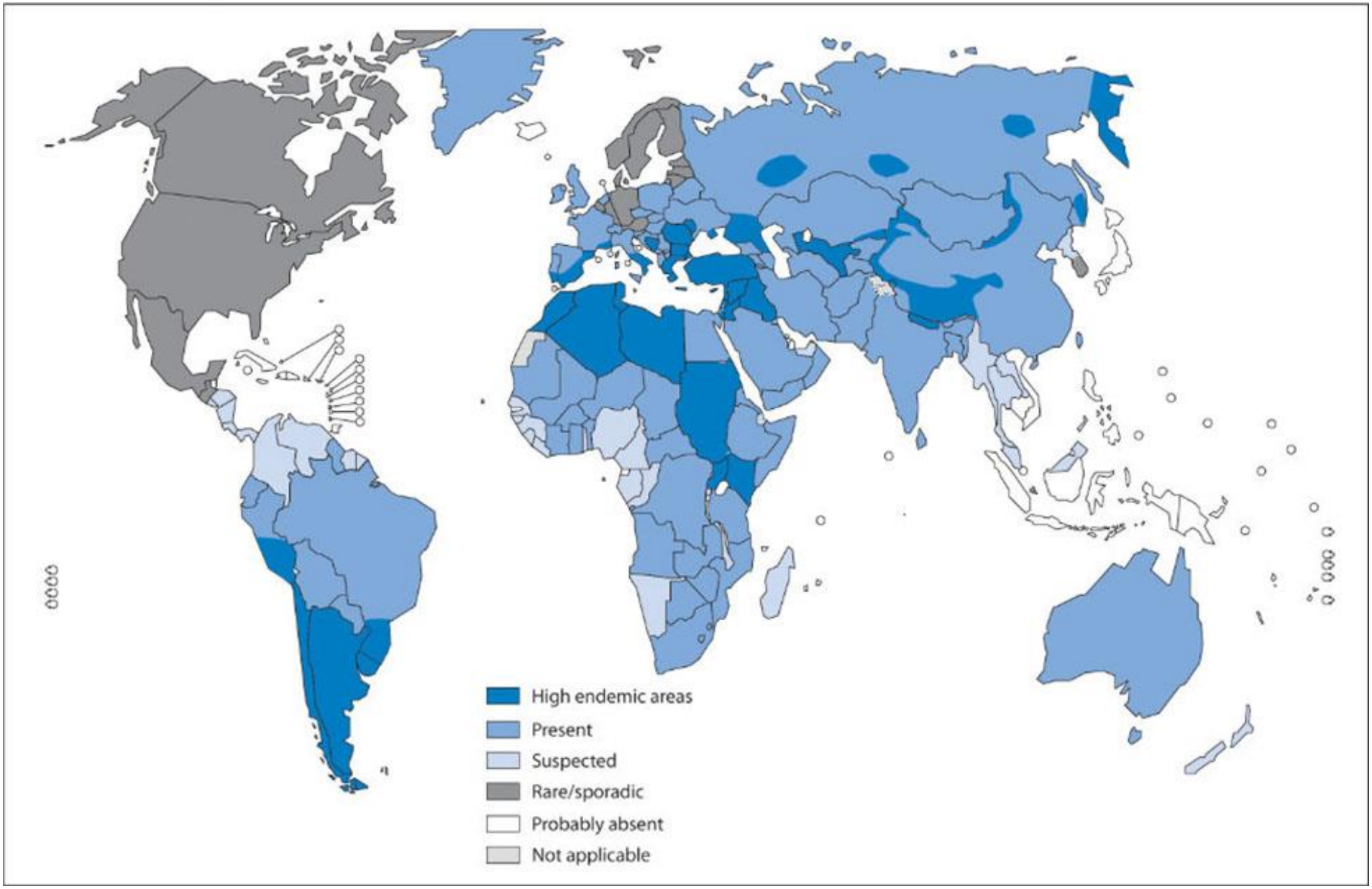
Boran
Garreh
Murulle
Degodia
Leisan
Ajuran
Sakuye
Degodia
Ajuran
Ogaden
SOMALI REPUBLIC
Ogaden
Boran
Auliahan
SOMALI
Korokoro
Abdwak
Nandi
Tukan
Kipsigis
GUSII
KIKUYU
KAMBA
MAASAI
Abdalla
Bajuni
BONI
OROMO
MIJIKENDA
TAITA
Dawida
Duruma
MOMBASA
INDIAN
OCEAN
TANZANIA
THE LANGUAGES AND DIALECTS OF KENYA
Cushitic Languages
Bantu Languages
Nilotic Languages

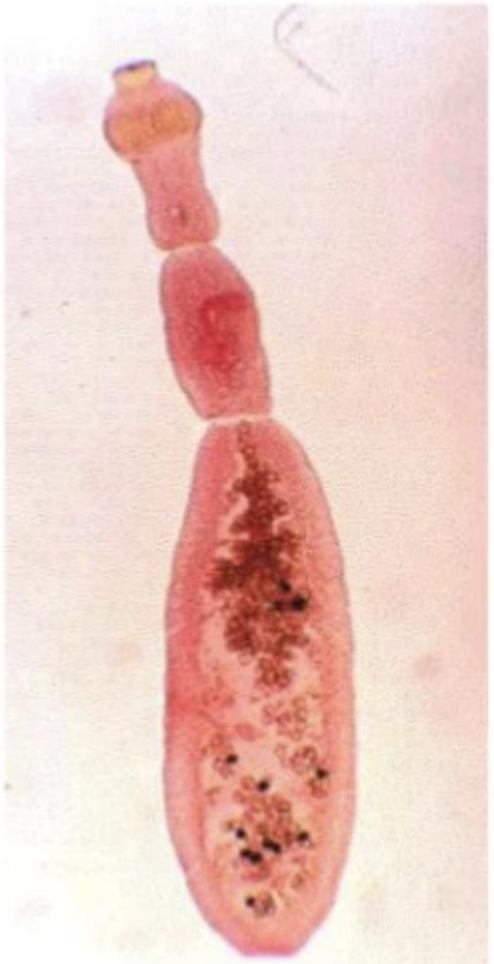
Man's Best Friend, the Turkana Tribe & a Gruesome Parasite

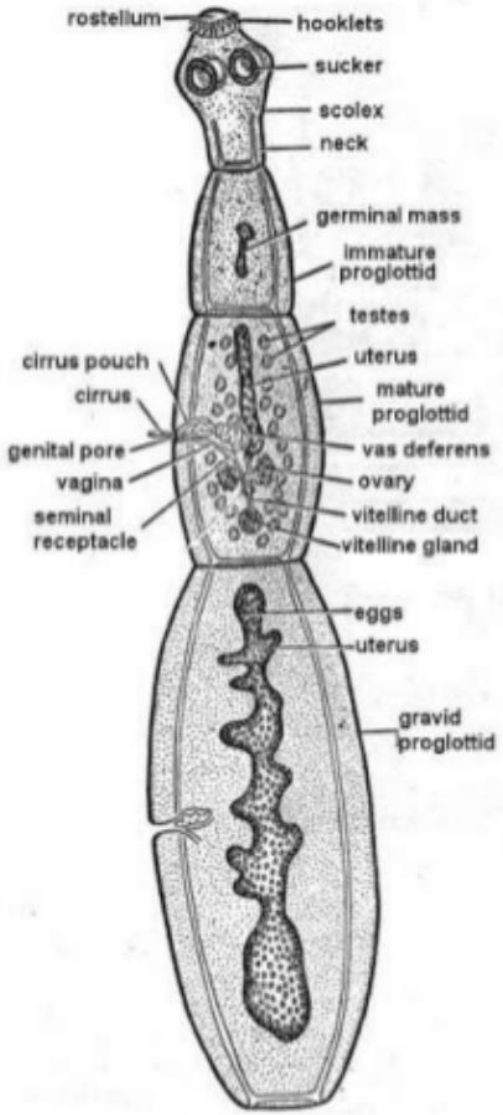
By Rebecca Kreston | January 26, 2012


High endemic areas
Present
Suspected
Rare/sporadic
Probably absent
Not applicable

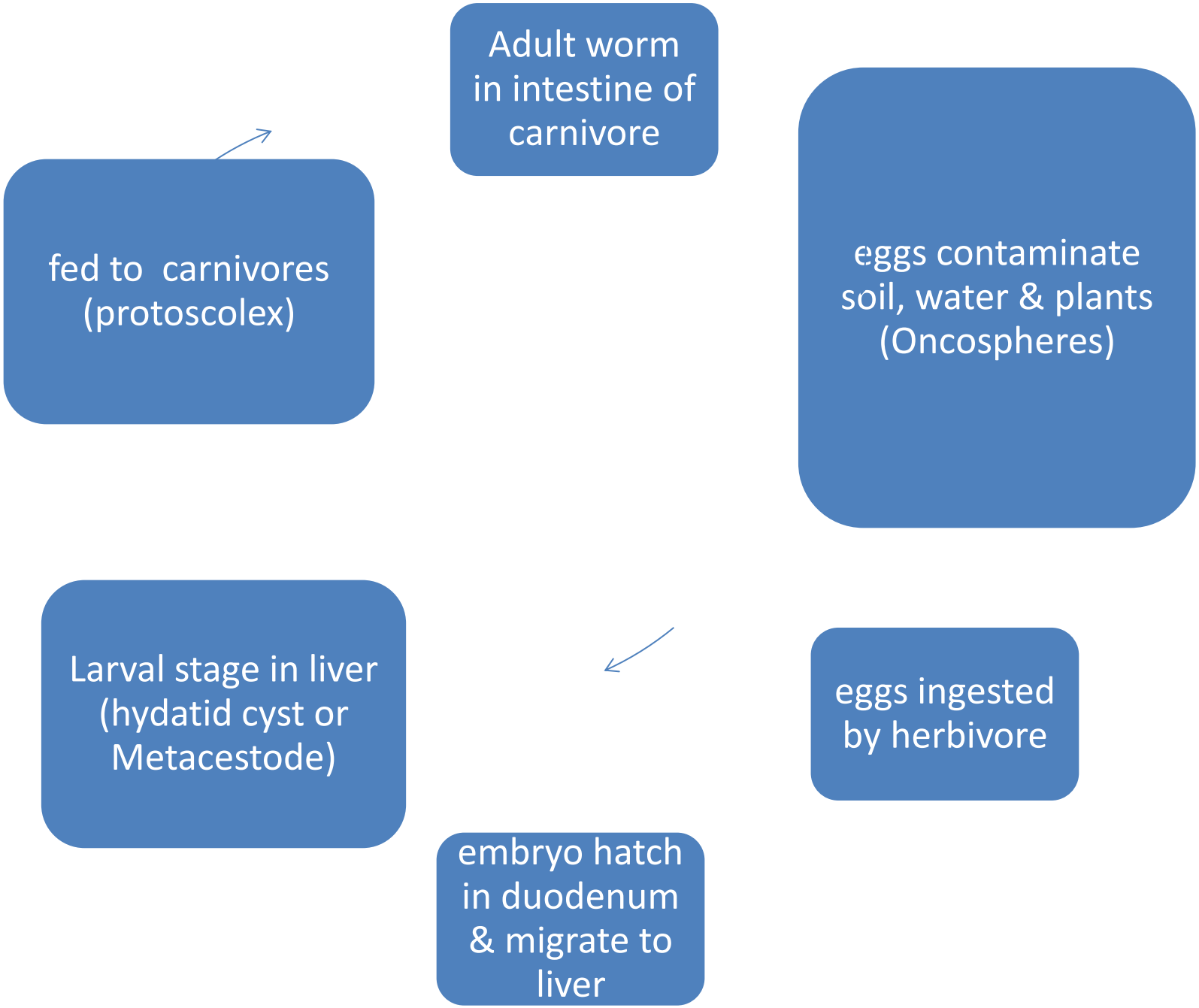
Etiology & Life-cycle

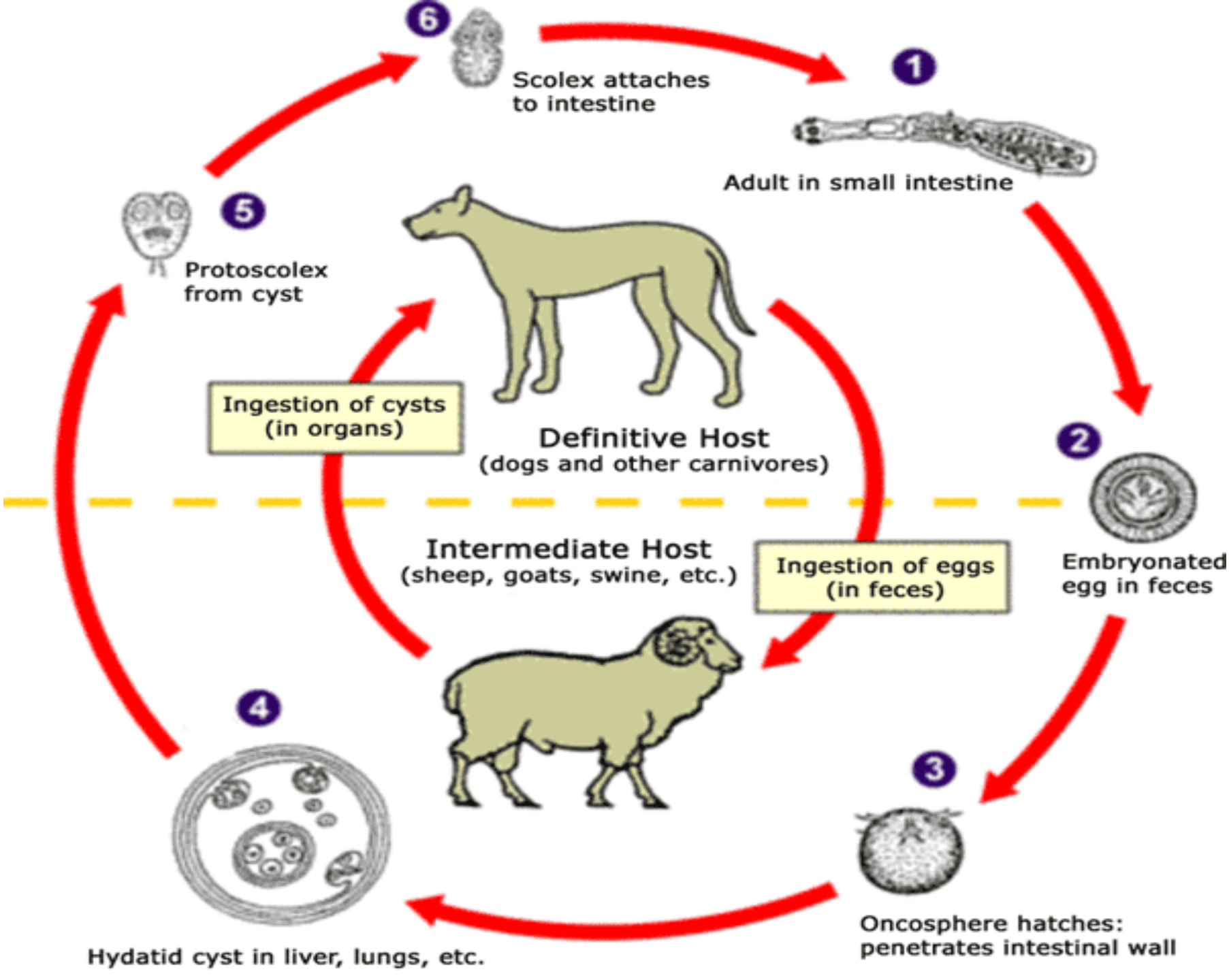
E. granulosus

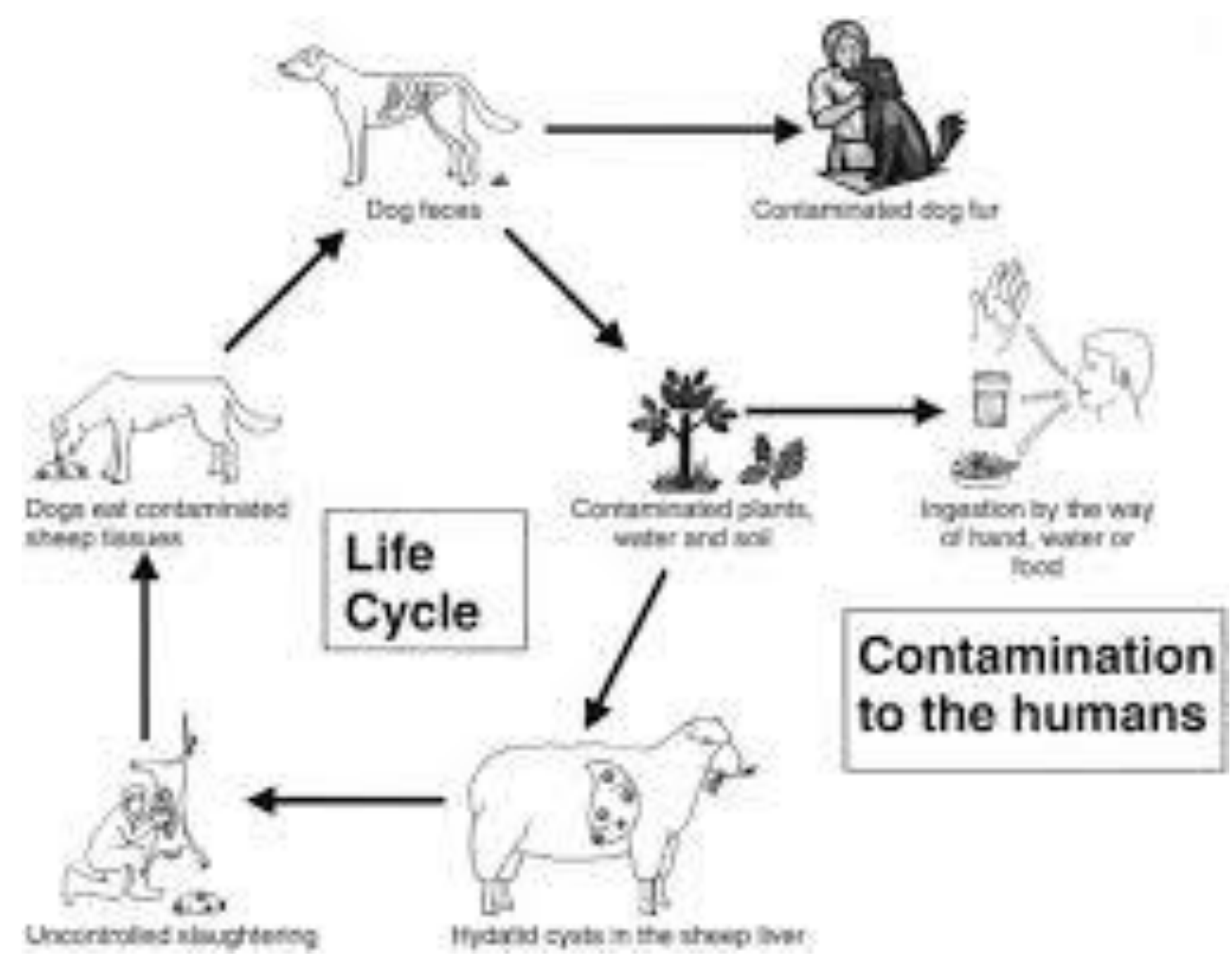




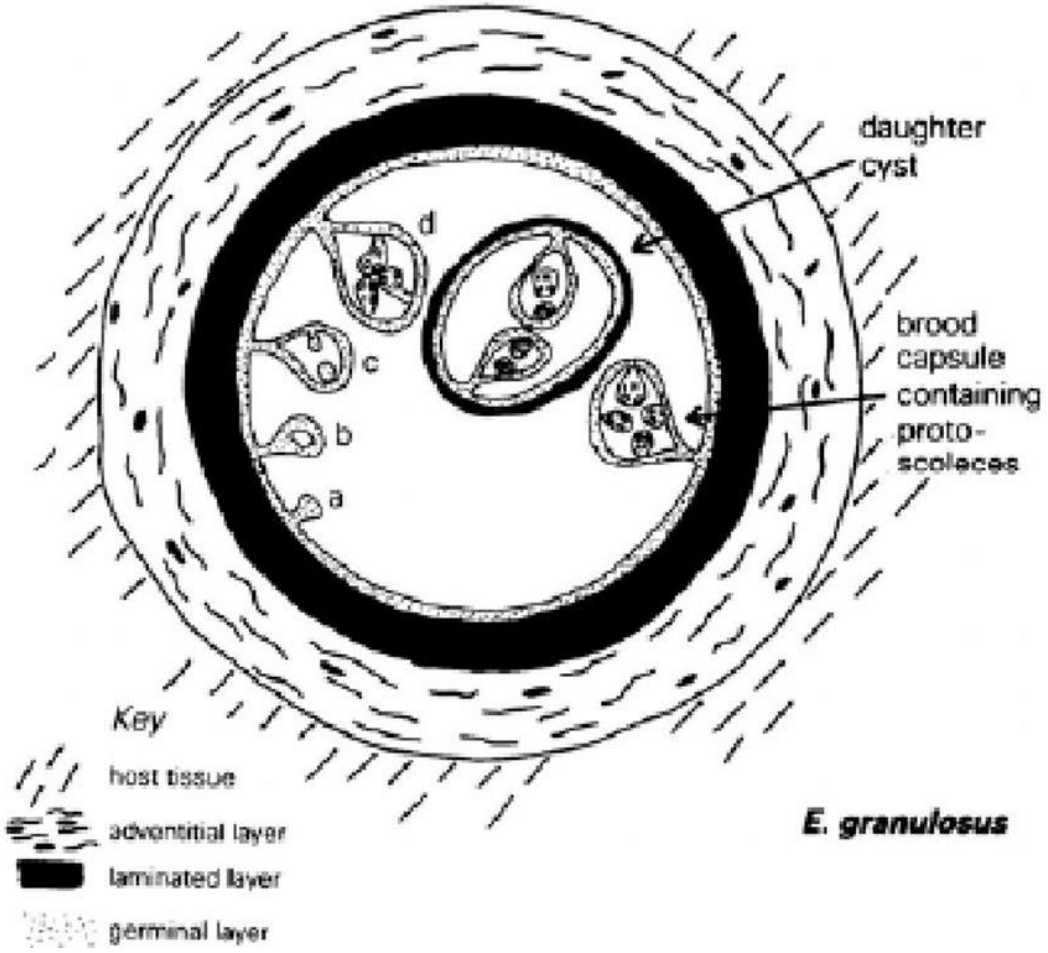
Characteristics	E. granulosus	E. multilocularis	E. oligarthus	E. vogelli
Geography	Worldwide	Northern hemisphere	Central & south America	Central & south America
Definitive hosts	Dogs, fox, wolf	Fox, dog, cat	Jaguar, puma	Bush dogs
Intermediate	Sheep, Cattles, pig, human	Rodent, pig, horse, human	Rodent, human	Rodents, human
Cyst	Unilocular	Multivesicular	Polycystic	Polycystic
Involvement	Visceral liver & lung	Visceral liver	Peripheral muscles	Visceral liver


Adult worm in intestine of carnivore
eggs contaminate soil, water & plants (Oncospheres)
eggs ingested by herbivore
embryo hatch in duodenum & migrate to liver
Larval stage in liver (hydatid cyst or Metacestode)
fed to carnivores (protoscolex)


6
Scolex attaches to intestine
1
Adult in small intestine
5
Protoscolex from cyst
Ingestion of cysts (in organs)
Definitive Host
(dogs and other carnivores)
2
Embryonated egg in feces
Intermediate Host
(sheep, goats, swine, etc.)
Ingestion of eggs (in feces)
4
3
Hydatid cyst in liver, lungs, etc.
Oncosphere hatches: penetrates intestinal wall


Dog feces
Contaminated dog fur
Dogs eat contaminated sheep tissues
Contaminated plants, water and soil
Ingestion by the way of hand, water or food
Life Cycle
Contamination to the humans
Uncontrolled slaughtering
Hydatid cysts in the sheep liver

Pathology & pathophysiology



Mature cyst consists of

a. Endocyst

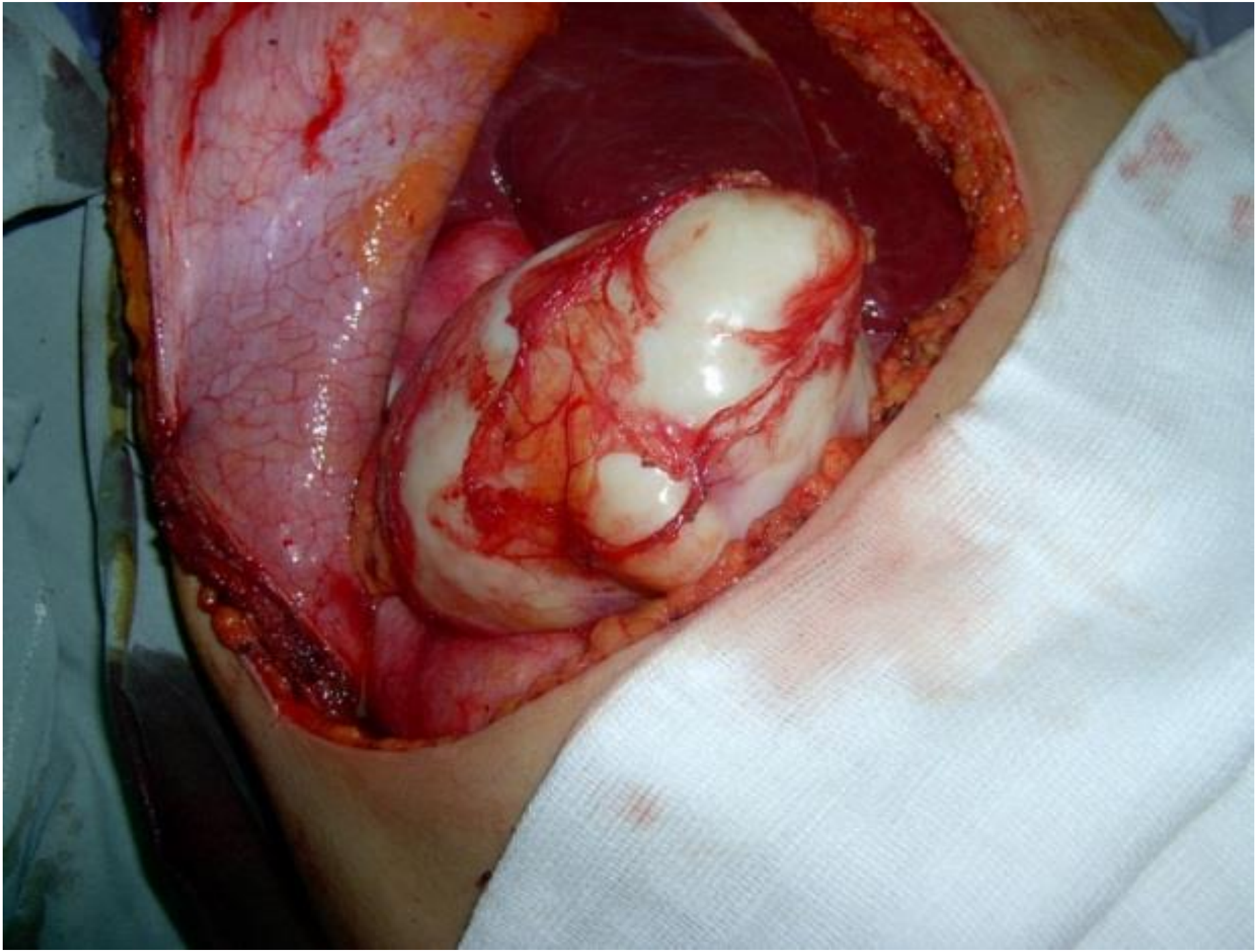
1.Germinative membrane (germinal layer, GM)→ broad capsules or daughter cysts (containing protoscolices) by endogenic vesiculation or satellite lesions by ectogenic vesiculation through small defects in LM

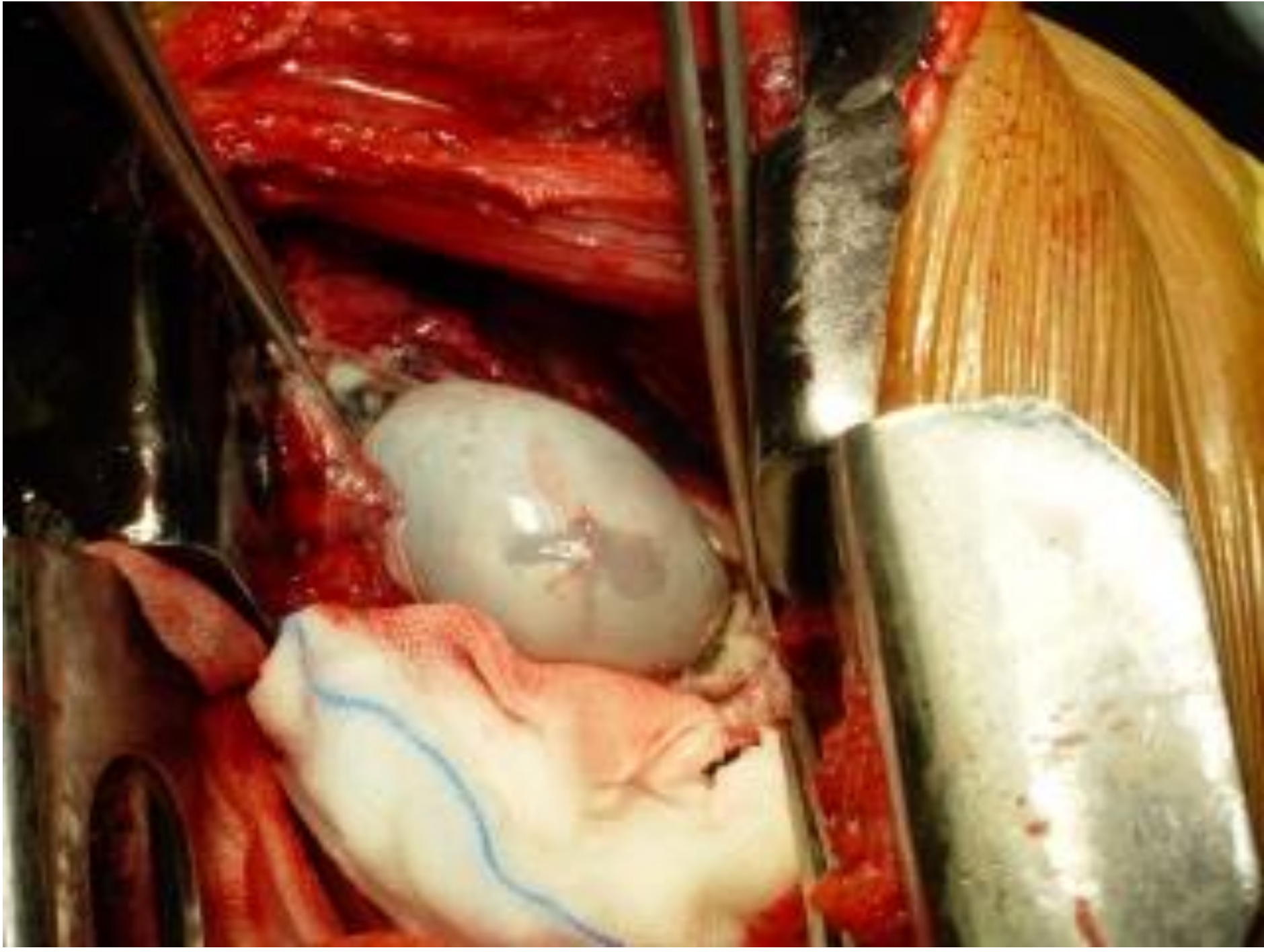
2.Laminated membrane(LM): 1-2mm thick acellular membrane, separating the Germinative membrane from pericyst. Permeable to water, electrolytes, urea & nutrients but not to bile, enzymes & bacteria. Calcification reduce permeability of the membrane reducing nutrient supply to cyst

b. Ectocyst (pericyst): fibrous layer formed by compression of surrounding parenchymal tissue (liver & spleen but not in brain & lung). Compressed vascular & biliary structures are present in ectocyst (→hypervascular rim or halo on CECT-scan)

c. Cyst fluid:

- Clear, colorless odorless sterile fluid secreted by GM
- High osmotic pressure
- Turbid & bile stained fluid suggest infection & biliary communication





Presentation

1. Compression of liver parenchyma (atrophy), Glisson capsule (pain), biliary tree(jaundice) or PV/HV (PHTN or BCS)
2. Infection via biliary route or blood stream (PLA)
3. Intrabiliary rupture (Most common)
4. Intrathoracic rupture→ bronchobiliary fistula & coughing “grape-skins”
5. Intraperitoneal rupture
6. Rupture into duodenum, stomach, pericardium & IVC (case reports)

Investigations

- Laboratory
  - ❖ Mild elevation of ALP/GGT & normal transaminases
  - ❖Eosinophilia (>3%) occurs in 25-45% of patients
  - ❖↑Ig in 31%

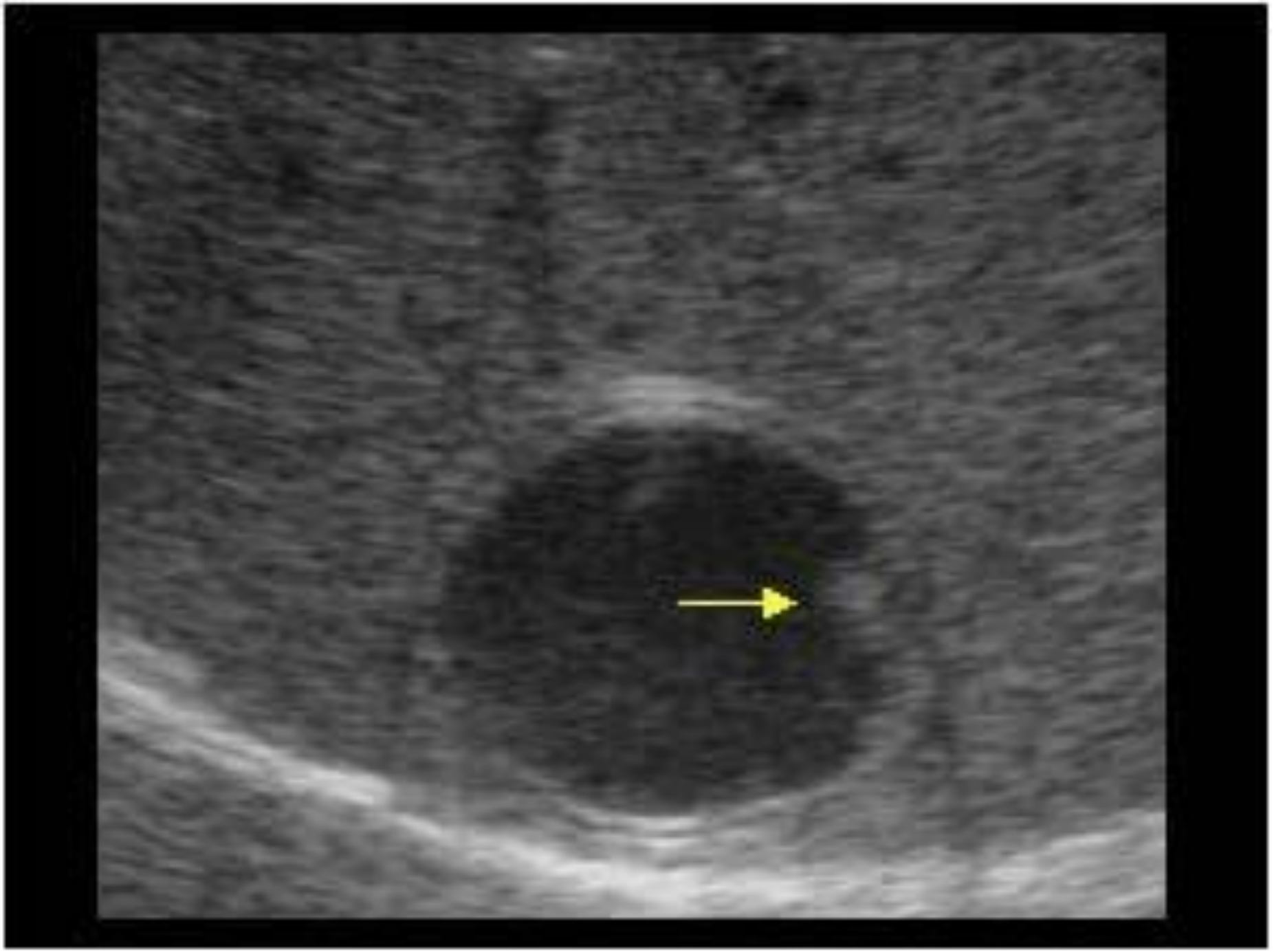
Laboratory

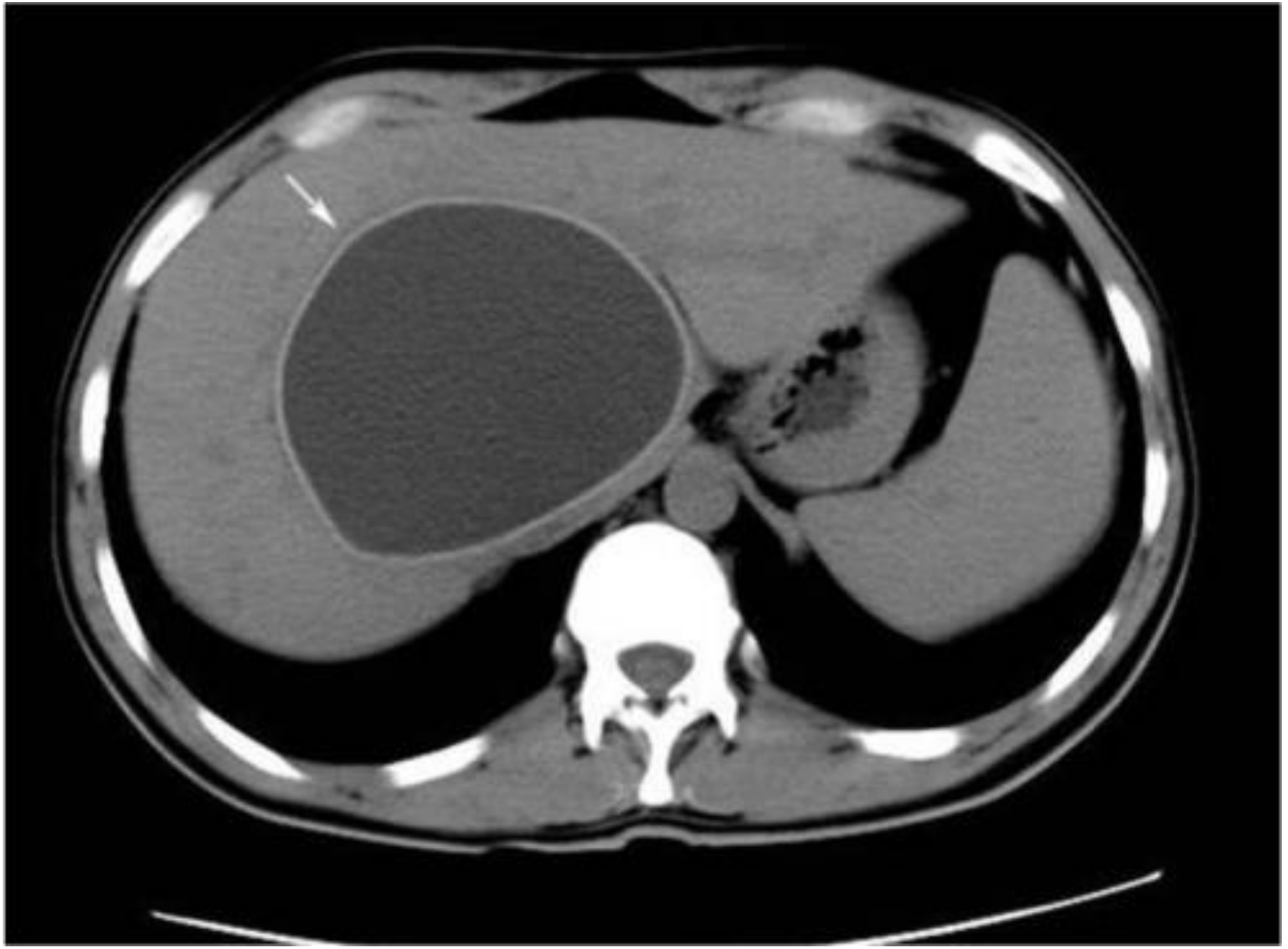
- Serology

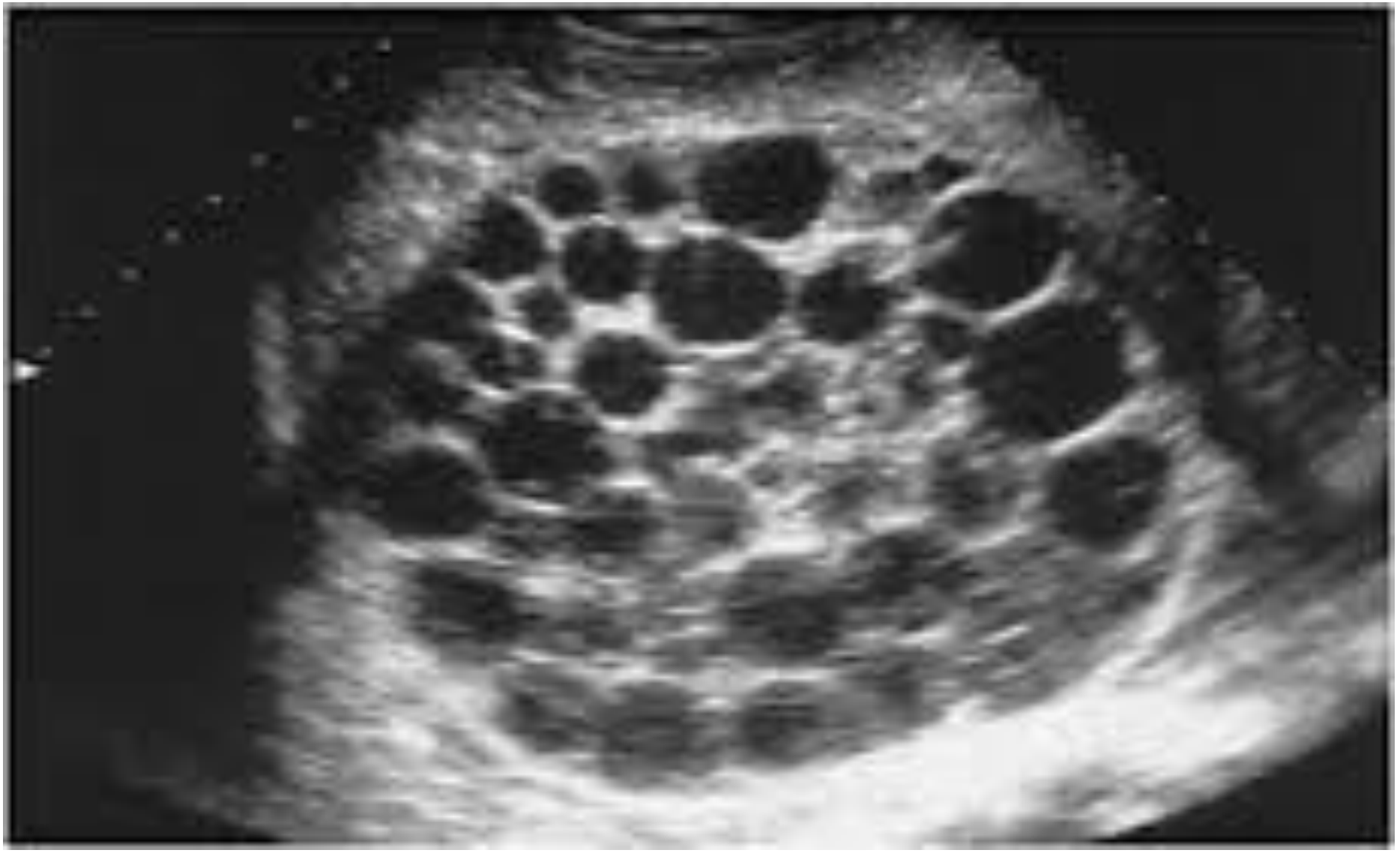
1. Immunoelectrophoresis: posttreatment FU
2. ELISA:
   - ✓Sensitivity 46-100%
   - ✓IgG: epidemiologic surveillance, remain +ve 4years after successful therapy
   - ✓IgM: posttreatment FU, negative 6months after successful therapy
3. Blotting : DDx & posttreatment FU

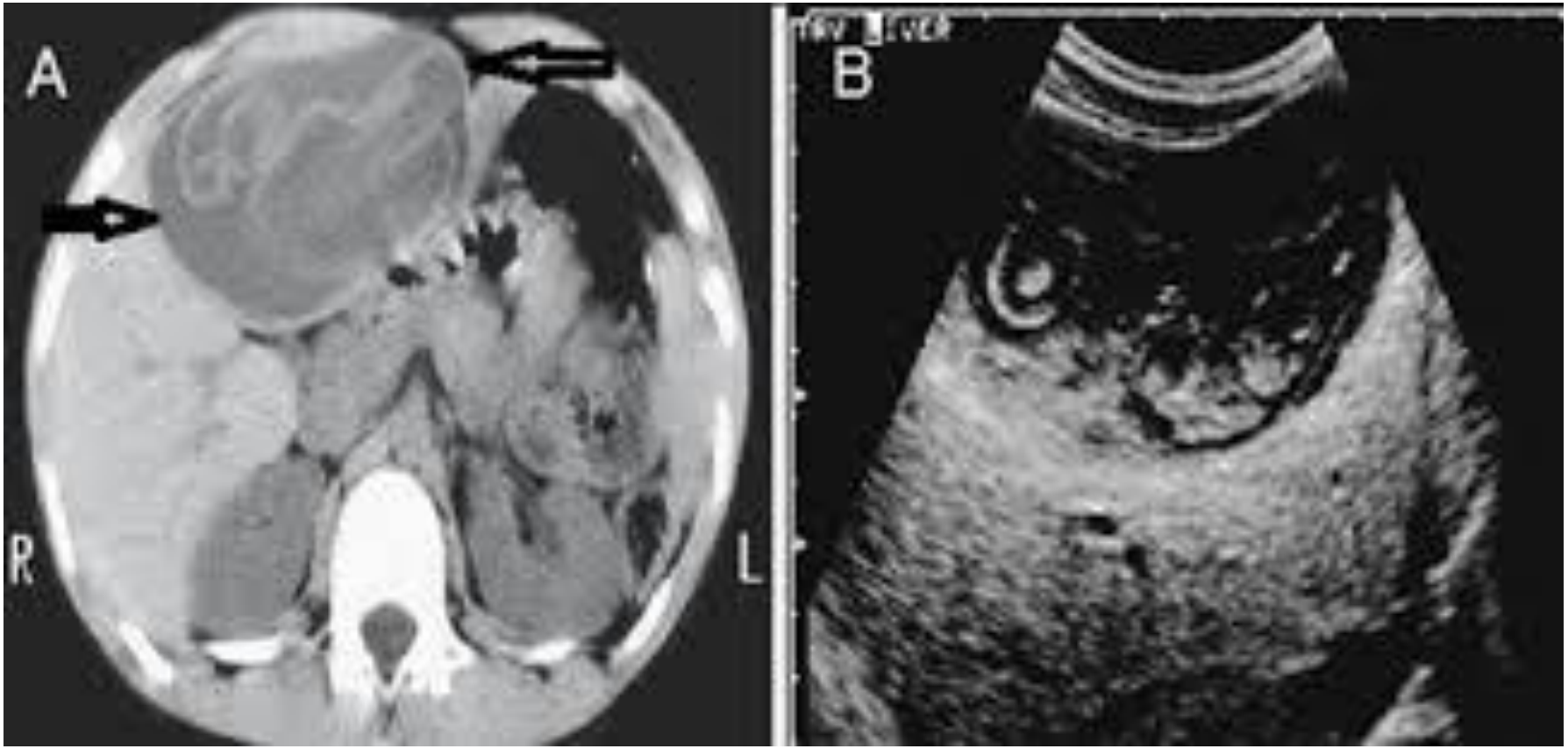
Imaging (US & CAT)

WHO classification of HC			
Type	Status	US/CTS features	Remarks
CL	Active	Unilocular, no cyst wall	Early stage nonfertile cyst, DDx necessary
CE 1	Active	Typical	Fertile
CE 2	Active	Rosette or honeycomb appearance	Fertile
CE 3	Transitional	Water lily or water-snake sign	Potentially fertile
CE 4	Inactive	Heterogeneous, degenerative, no daughter cysts	Dead protoscolices, DDx necessary
CE 5	Inactive	Thick, partially or completely calcified wall	Dead , DDx may be necessary with partial calcification








A
R
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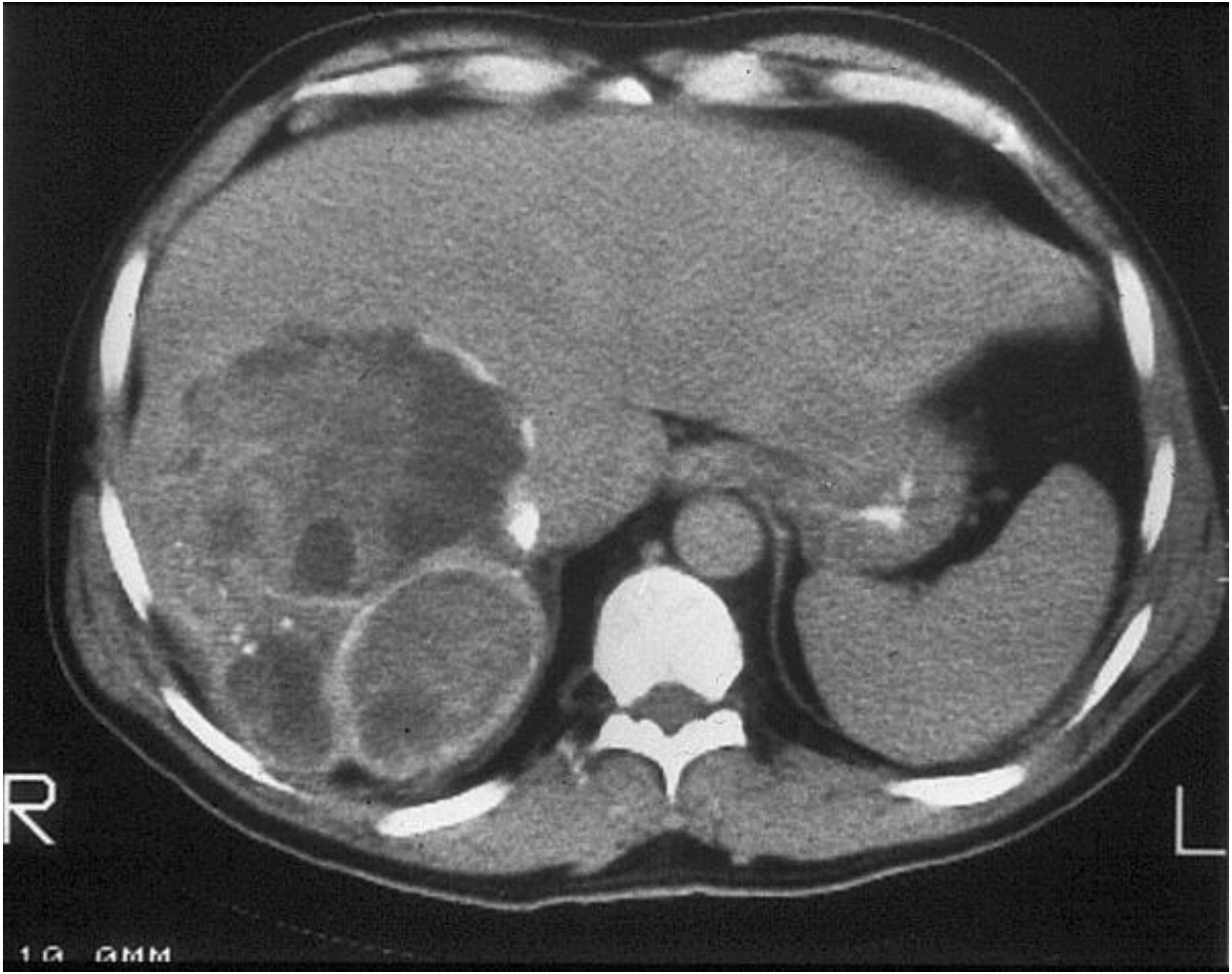
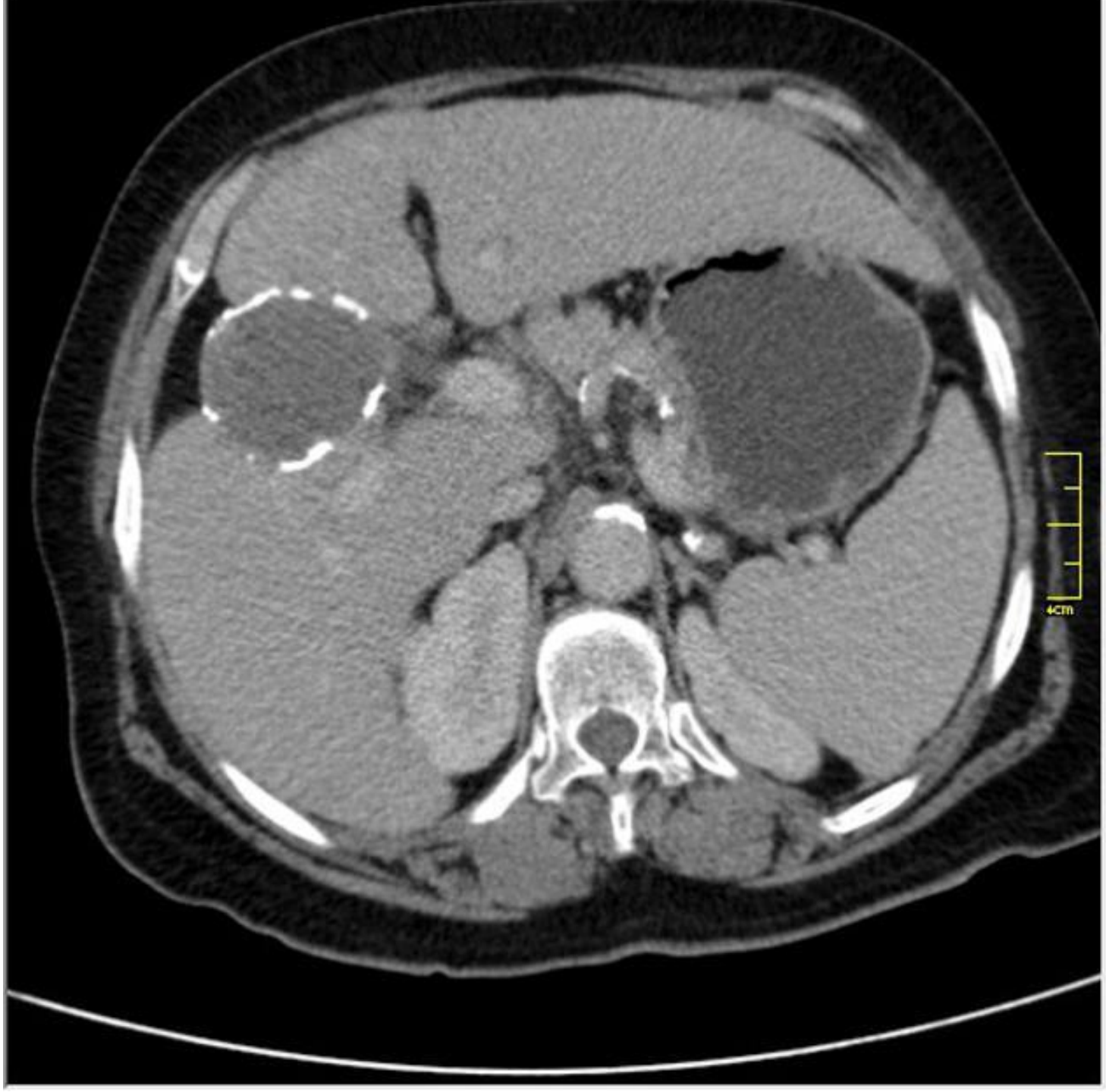

R
L
10 6MM


4cm

Treatment

❖Symptoms, size, site & Stage

1. Observation

Asymptomatic small<5cm CL-type cysts

- Densely calcified HC

Treatment

2. Surgery

Conservative: Scolicidal agents (beware of biliary communication→ caustic cholangitis)

1. Hypertonic saline (3-30%), best is 20%/6min contact time, →hypernatremia
2. Cetrimide/ chlorhexidine (Savlon)→ metabolic acidosis & methemoglobinemia
3. Alcohol → flammable & caustic cholangitis
4. Povidone-iodine→ caustic cholangitis, concentration dependency & coloring of cavity
5. Formalin, Silver nitrate & Eusol (sodium hypochlorite) → caustic cholangitis

Treatment-Surgery

- Dealing with cavity
  1. External tube drainage
  2. Capsulorrhaphy
  3. Capitonnage
  4. Omentoplasty
  5. Internal collapse
  6. Introflexion
  7. Myoplasty
  8. Cystoenterostomy
  9. Marsupialization

- Radical (pericystectomy & formal liver resection)
- Laparoscopy
- Complications
  1) Bile leak (1-10%)
  2) Biliary stricture (mostly related to scolicidal agents or bile duct injury)
  3) Recurrence (10%) local or remote

Treatment

3. Percutaneous treatment

Indications: accessible type 1& 3 cysts and some type 2 cysts

- Contraindications

1. Inaccessible cysts
2. Peripheral cysts with thin overlying hepatic tissue
3. Intervening important vascular structures
4. Biliary, intraperitoneal or pleural rupture

Percutaneous treatment

- Techniques

PAIR (puncture, aspiration, injection & reaspiration)

2. PAIR catheterization
   - After puncture of cyst, a catheter is placed using Seldinger technique
   - Injection of scolicidal agent is followed by reaspiration
   - Catheter left in place for 24 hours
   - If drainage is <10cc & free of bile→ alcohol-sclerotherapy after cystogram
   - Recommended for cysts larger than 6cm (>100mL)
3. PEVAC (percutaneous evacuation of cyst contents)
   - 2-step technique
   - 1st, as in PAIR-Catheterization
   - 2nd, 14-18F stiff-sheath is inserted & suction, cutting device & scolicidal-irrigation are used to evacuate the cyst
   - A catheter (same size as sheath) is left in place to monitor for bile leak
   - The catheter is removed when appropriate (±cystography)

Treatment

4. Medical therapy: Albendazole(Albenza) & Mebendazole(Vermox)

Impair parasite glucose uptake

- Albenza is superior
- CI: CLD & pregnancy
- Dose 15mg/kg/day with meals in 2 divided doses (Max 800mg/day), X3 28day courses, separated by 2-weeks
- SE:

a. Headache
b. Nausea, vomiting & anorexia
c. Abdominal pain
d. Itching
e. Elevated liver enzymes in first weeks
f. Hair loss (reversible)
g. Leukopenia (rare)

Medical therapy

Indications & aim of therapy

1. Definitive therapy (3 courses of therapy→ 80% success, 25% relapse rate)
2. Preoperative prophylaxis should start 1 week before the procedure
3. Postoperative prophylaxis:

- ✓ 3-8 weeks for uncomplicated cases with no spillage
- ✓ 3-6 months for complicated cases with spillage

Alveolar hydatid cyst (E. alveolaris & E. multilocularis)

- Malignant Echinococcosis
- Definition: chronically progressive potentially fatal parasitic liver infection (95% MR if untreated)
- The disease is almost restricted to northern hemisphere, and increasing in distribution
- Cell cycle: adult tapeworm in Fox→ eggs ingested by rodents→ oncospheres reach the liver→ protoscolices ingested by fox
- Structure
  - ✓ Very thin laminated membrane & GM (infectious)
  - ✓ Massive infiltrating parasitic mass with no defined borders
  - ✓ Spongy mass with small irregular cavities filled with gelatinous material

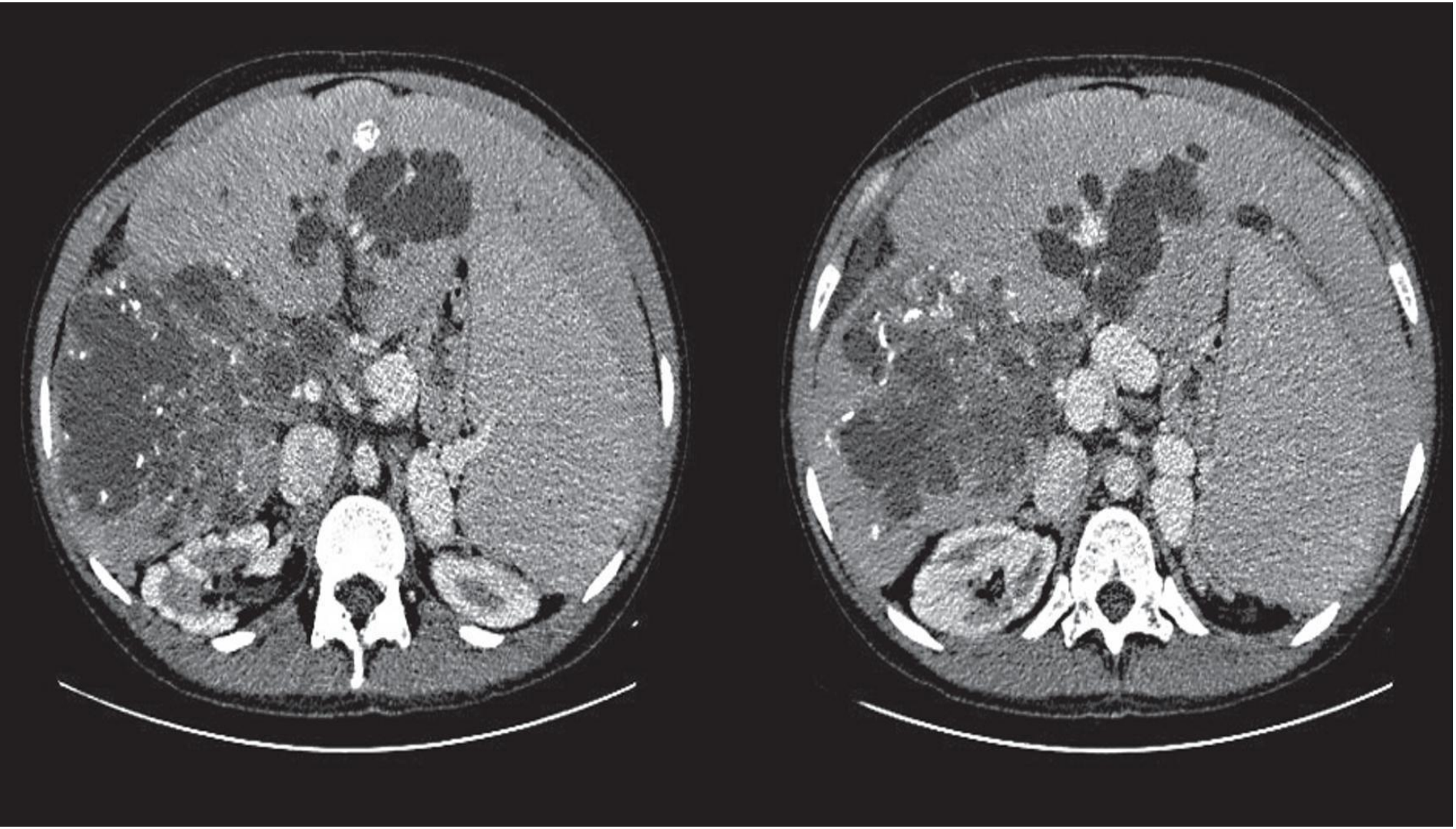
- Presentation : asymptomatic (early)→ hepatomegaly & abdominal mass→ CLD & hepatic failure
- Dx:

➢ CT/US: heterogeneous, hypodense mass with irregular border & central necrosis with clusters of microcalcifications (plaque-like calcifications)

➢ MRI: better than CT in showing central necrosis & margins of the lesion

➢ The large tumorlike lesion in a patient with well general condition suggest the diagnosis in endemic areas

➢ Serology is very useful & effective in diagnosis (better than in HC)



PNM-Classification of alveolar Ecchinococcosis

Primary liver involvement	**P0**	**No detectable liver lesion**
	P1	Peripheral lesion without biliary or vascular involvement
	P2	Central, biliary or proximal vascular involvement of one lobe
	P3	Central, biliary or proximal vascular involvement of both lobes or two hepatic veins
	P4	Any, with involvement of main PV, IVC or hepatic arteries
Neighboring organ	N0	No regional involvement
	N1	Involvement of neighboring organs
Distant metastasis	M0	No Mets
	M1	Lung or brain metastasis

Treatment

- Indicated even when asymptomatic
- Involves resection, OLT & chemotherapy