Case Presentation

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1st year fellow

Respiratory Medicine - SickKids

Case presentation

- 3 y/o previously healthy female
- 2.5 years of age – admitted to periphery hospital with fever and cough
- Found to have pneumonia of LLL (CXR N/A)
- Admitted and treated with IV antibiotics
- Improved and discharged after 3 days
- During her hospital stay she was found to be **influenza positive**
- She made a complete recovery and has not had any subsequent respiratory complaints, signs or symptoms

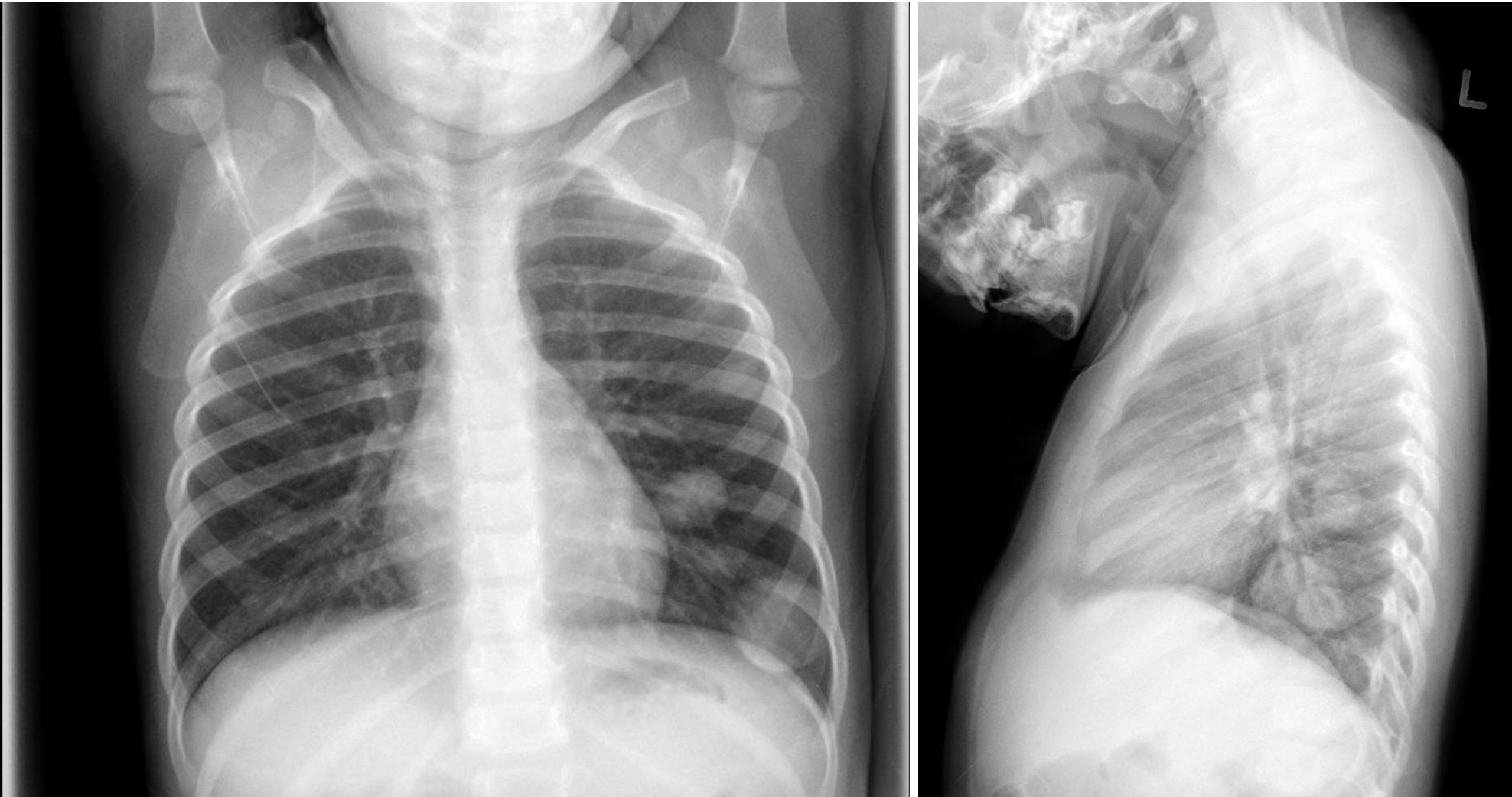
Case presentation

- Normal pregnancy, no abnormalities of the fetus were reported on screening ultrasound
- Delivery at 37 weeks and 4 days, no respiratory complications at birth
- No allergies/ wheezing/ use of puffers
- No history of foreign body aspiration
- No sick contacts
- No recent trips, from Sudbury
- Exposed to animals (farm)

Case presentation

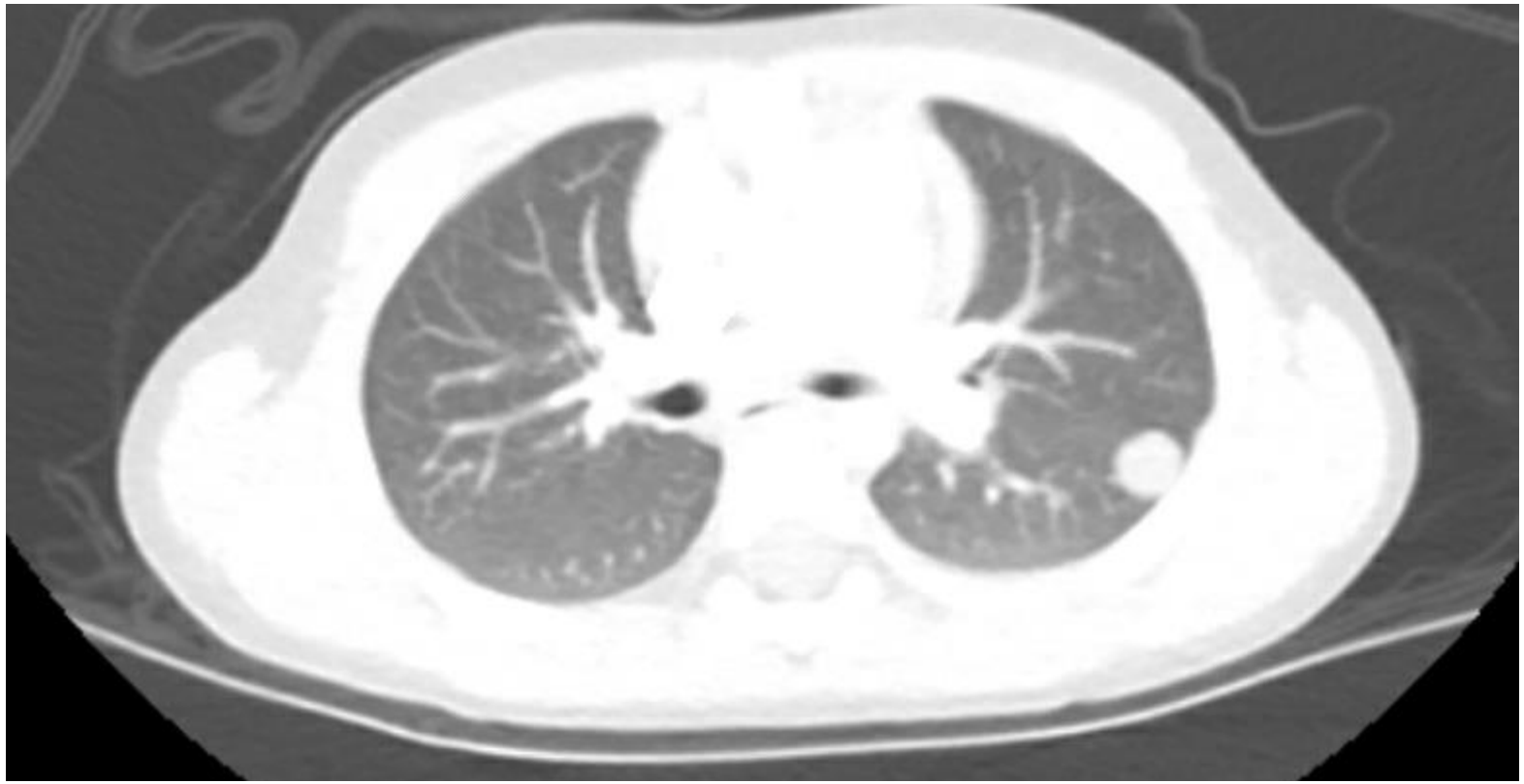
- On mother's side - family history positive for diabetes, strokes, various cancers including bone cancer, leukemia, lung cancer, kidney cancer, liver cancer and breast cancer
- Mother had GI polyps found when she was 24 years of age, removed with endoscopy and found to be benign
- She is not aware if this is a familial condition

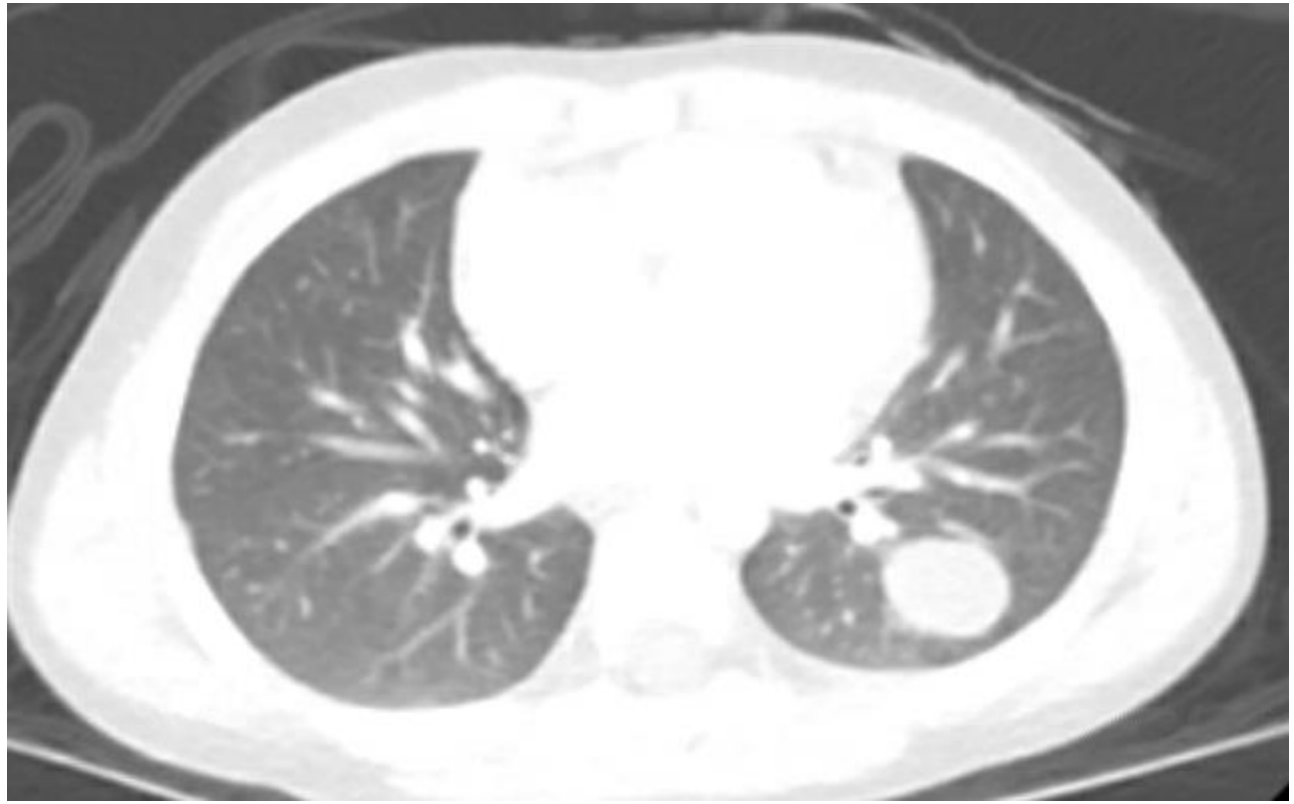
Two months post admission

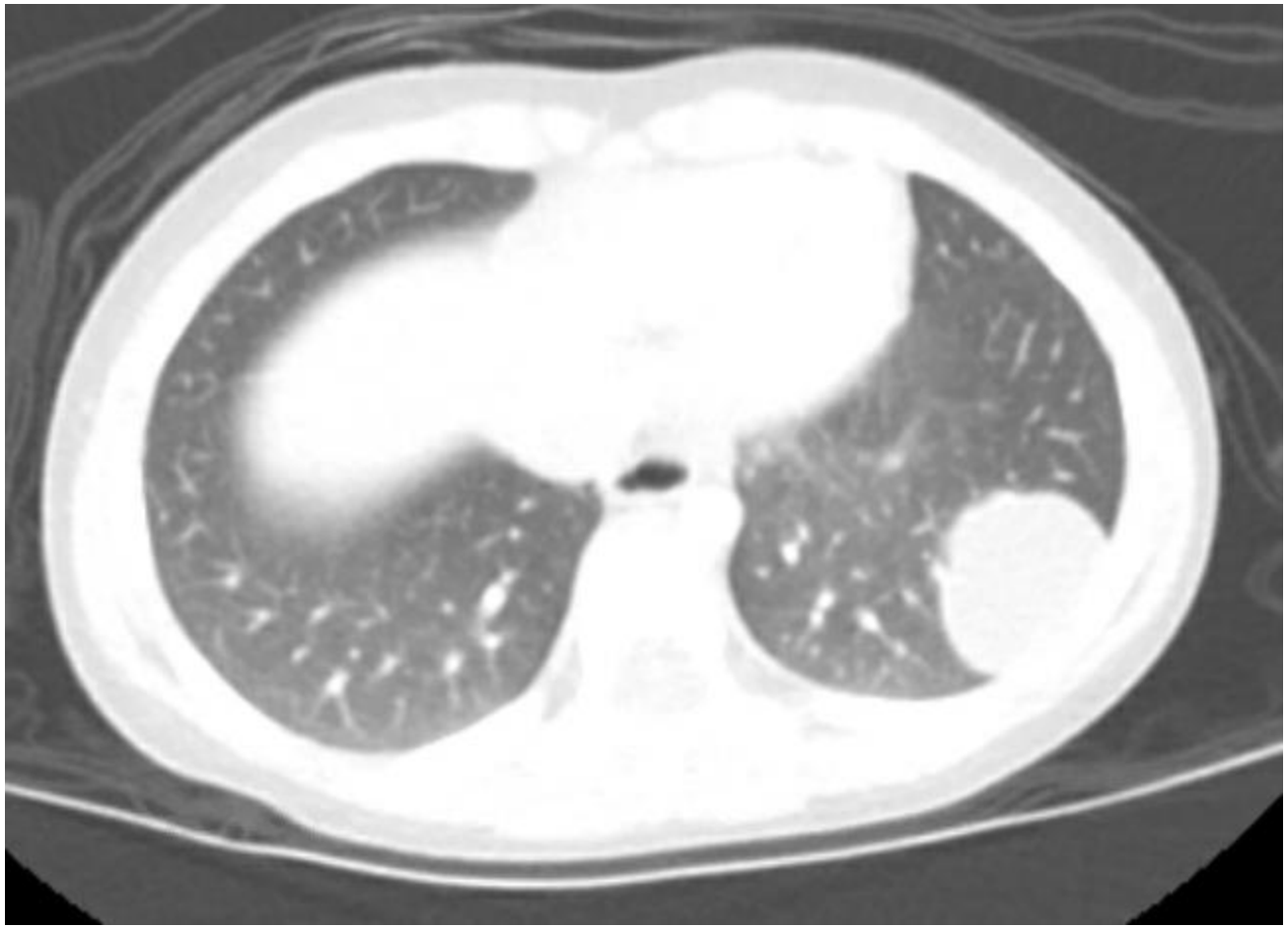


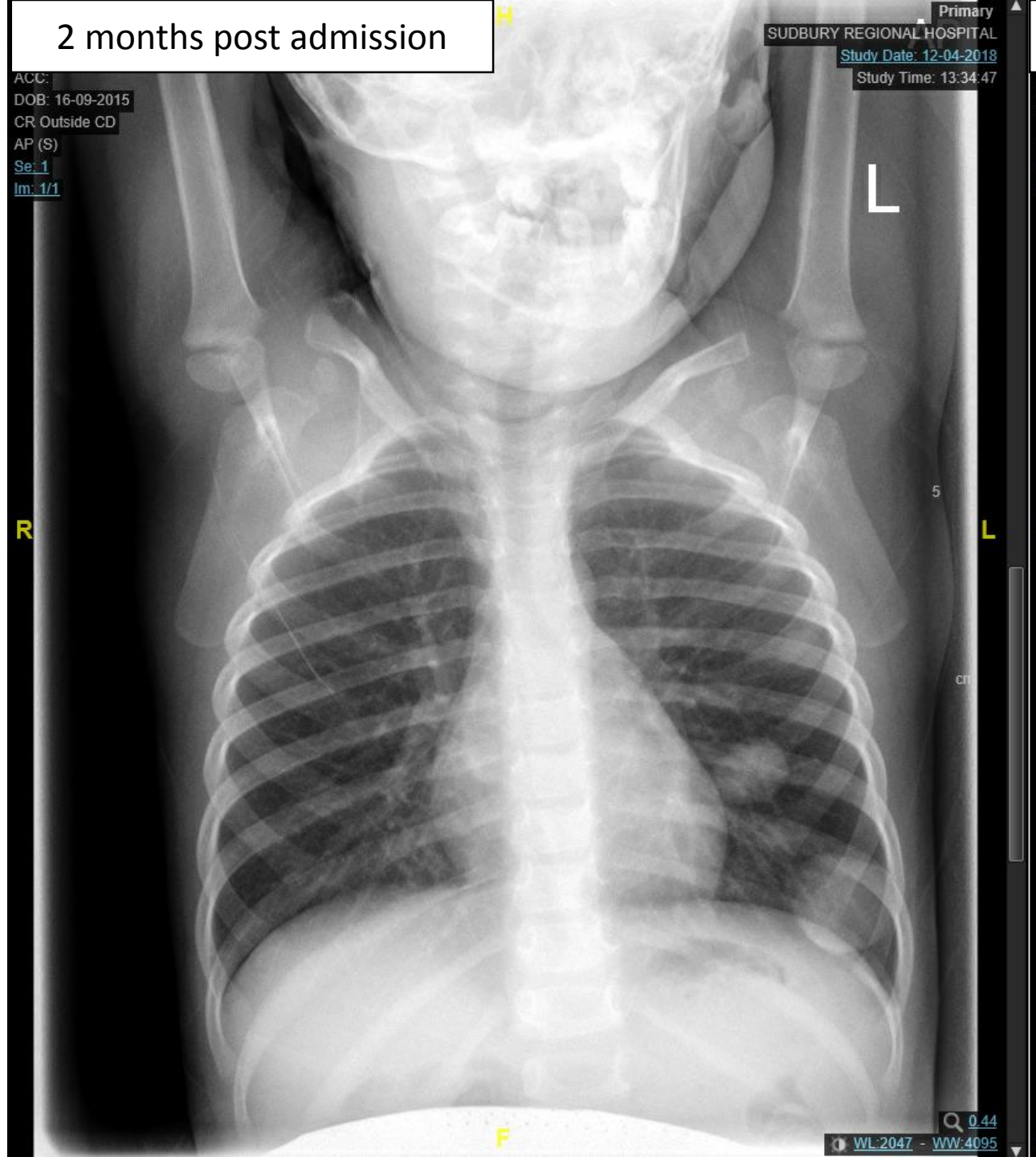
Additional studies

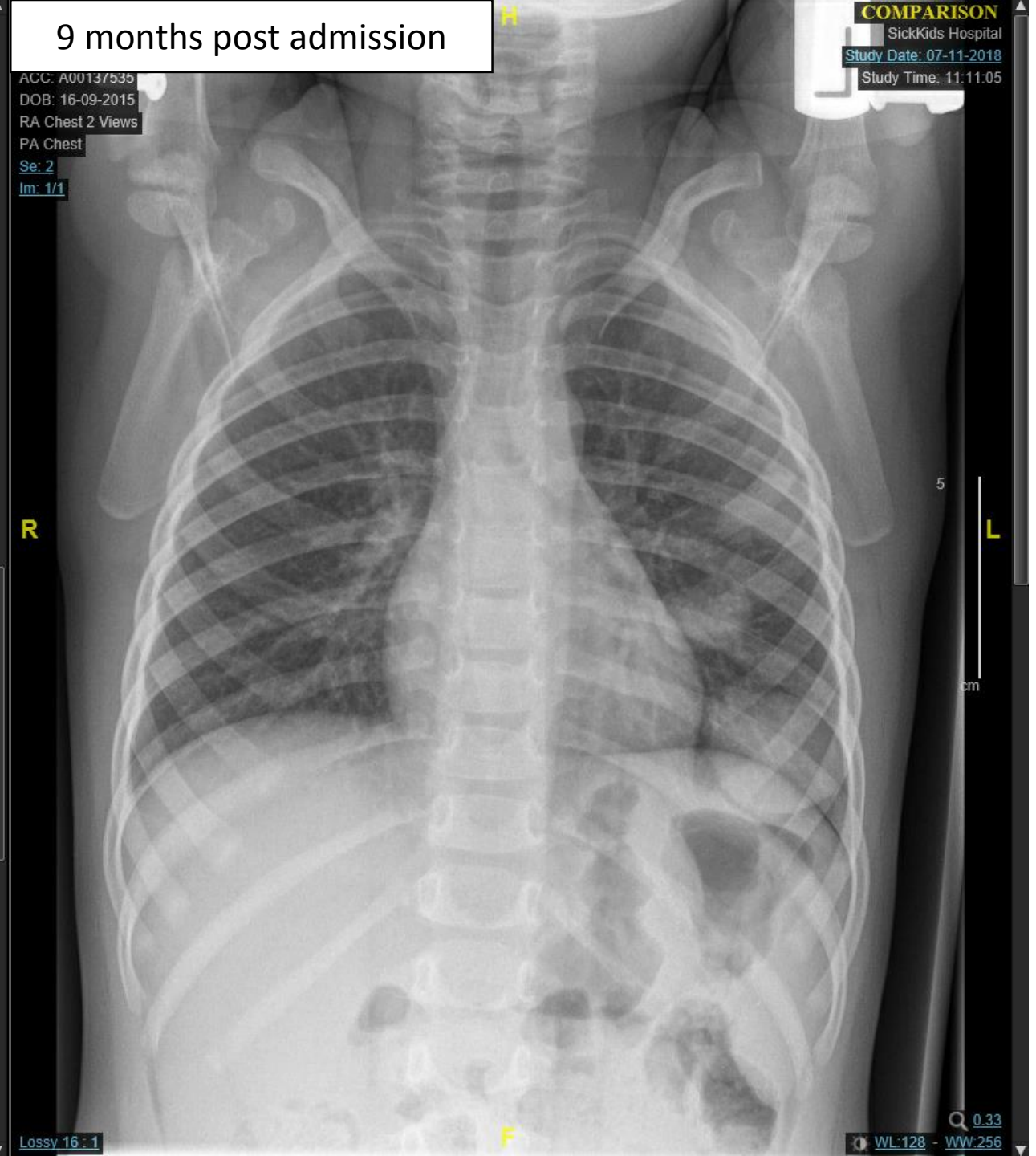
- CBC - normal (WBC 7.05, Hgb 134, plt 315),
- Diff – normal (Neu 2.08, Lym 4.22, Eos 0.16)
- Blood gas normal, elec – normal, Cr – normal
- ALT – 151, GGT – 20, AST – 96, CRP – normal
- Cardiac Echo – normal
- Abdominal US – normal
- Tuberculin skin test (Mantoux) – negative
- Serology for *E. granulosus* and *E. multilocularis* – negative








2 months post admission
Primary
SUDBURY REGIONAL HOSPITAL
Study Date: 12-04-2018
Study Time: 13:34:47
ACC:
DOB: 16-09-2015
CR Outside CD
AP (S)
Se: 1
Im: 1/1
L
R
L
0.44
WL:2047 - WW:4095


9 months post admission
COMPARISON
SickKids Hospital
Study Date: 07-11-2018
Study Time: 11:11:05
ACC: A00137535
DOB: 16-09-2015
RA Chest 2 Views
PA Chest
Se: 2
Im: 1/1
R
L
Lossy 16 : 1
0.33
WL:128 - WW:256

Interpretation

- Three relatively well-defined nodules in the left lower lobe
- Two of the nodules appear to be marginally larger compared to previous suggesting slow growth, the other nodule is stable
- The lungs appear otherwise clear
- No signs of hilar or mediastinal lymphadenopathy
- **Appearances are nonspecific and although the slow growth argues against metastatic disease, this cannot be excluded**
- **Appearances are not typical for infection or congenital pulmonary lesions**

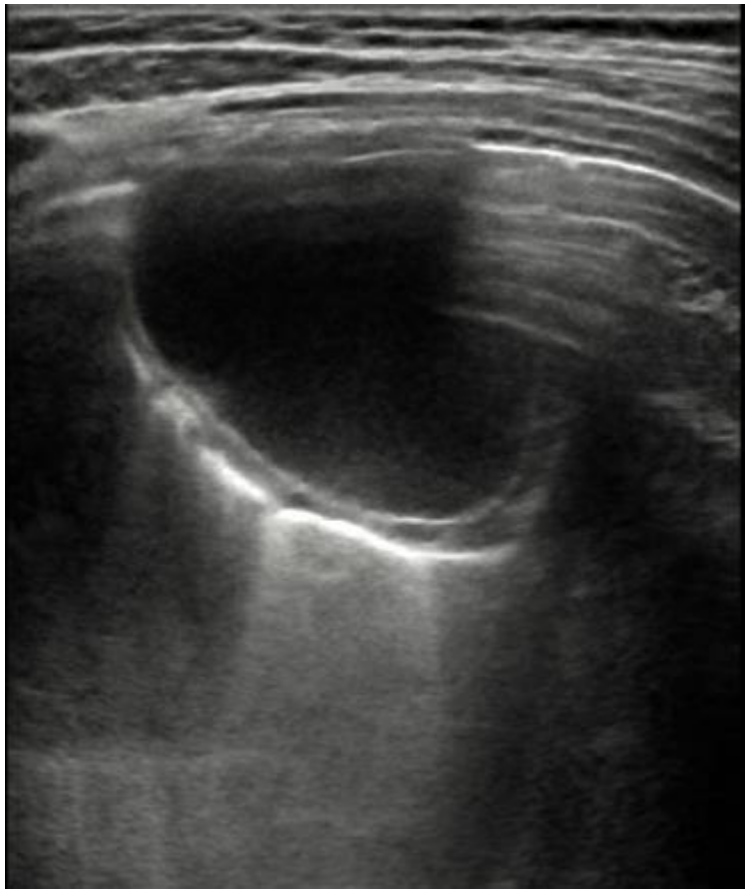
DD – pulmonary nodules/cysts

- Infectious
  - Lung abscess
  - TB
  - Hydatid cysts
  - Aspergilloma
- Congenital
  - Bronchogenic cysts
  - CPAM
- Tumor
  - Primary or secondary, benign or malignant

Next step? (~6 months after initial CT)

- US
- Bronchoscopy + BAL
- Repeat CT
- MRI
- Biopsy
- Other testing? (specify)

Chest US



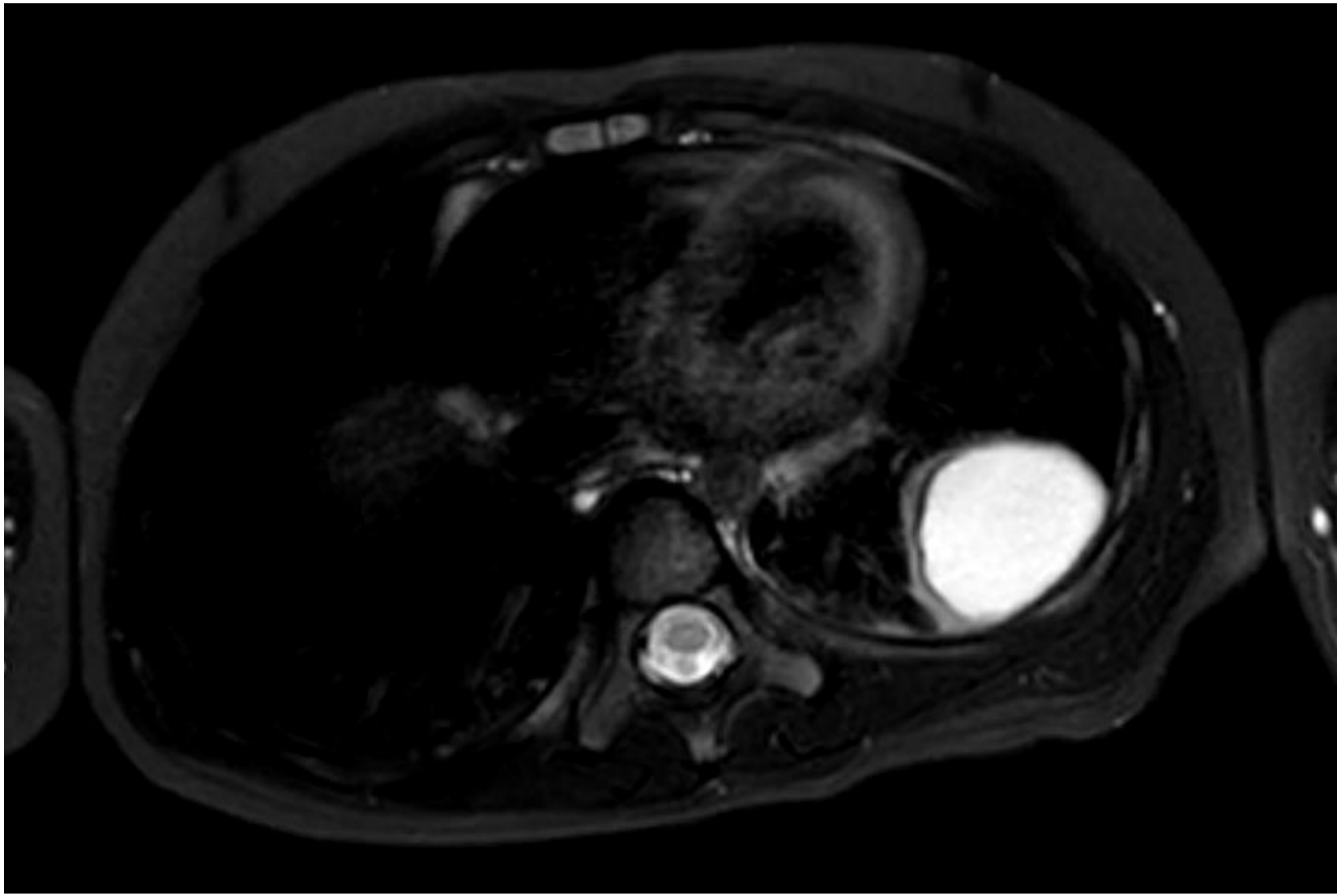
Chest US

- **Unilocular cystic structure is seen in the left posterolateral lower lung** measuring 3.5 x 2.6 x 2.5 cm, **exhibiting a thin smooth** (1 - 2 mm) **wall**. This likely corresponds to the largest lesion on the previous CT scan which abuts the left posterolateral chest wall
- It **exhibits predominantly anechoic fluid**, with minimal internal low-level echoes. **No septations present**. **No solid nodular components or vascular flow demonstrated**
- The other two smaller cysts seen on the previous CT were not demonstrated on this examination because of intervening lung

Chest US

- IMPRESSION:
  - **Unilocular mostly simple cyst involving the left posterolateral lower lung, without septations, solid components, or internal daughter cysts, corresponding to the largest cyst on the previous CT scan.**
  - **Differential diagnoses as discussed on the previous CT scan are unchanged.**

Total body MRI



Total body MRI

- INTERPRETATION:
  - Allowing for cross modality comparison, there has been **mild interval growth of the 3 known pulmonary cystic lesions** (most notably the most superior lesion)
  - **No additional lesions** identified on this whole-body scan

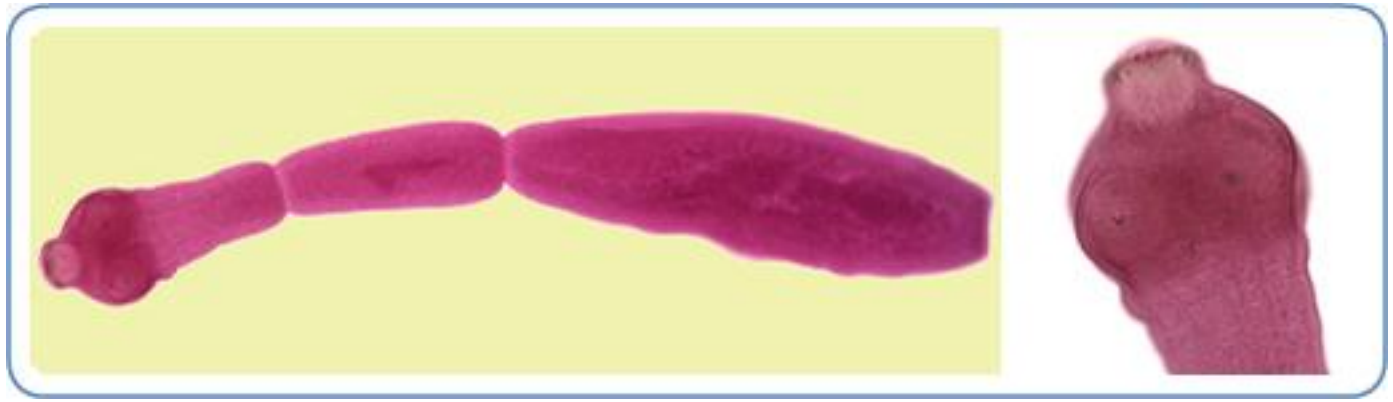
Next step?

- Bronchoscopy + BAL?
- OR (biopsy or resection)?
- Other testing? (specify)

- Hydatid disease highly suspected
- Surgery booked for full removal of cysts
- Echinococcus (hooklets) seen in resected cysts microscopically
- **Hydatid disease confirmed**

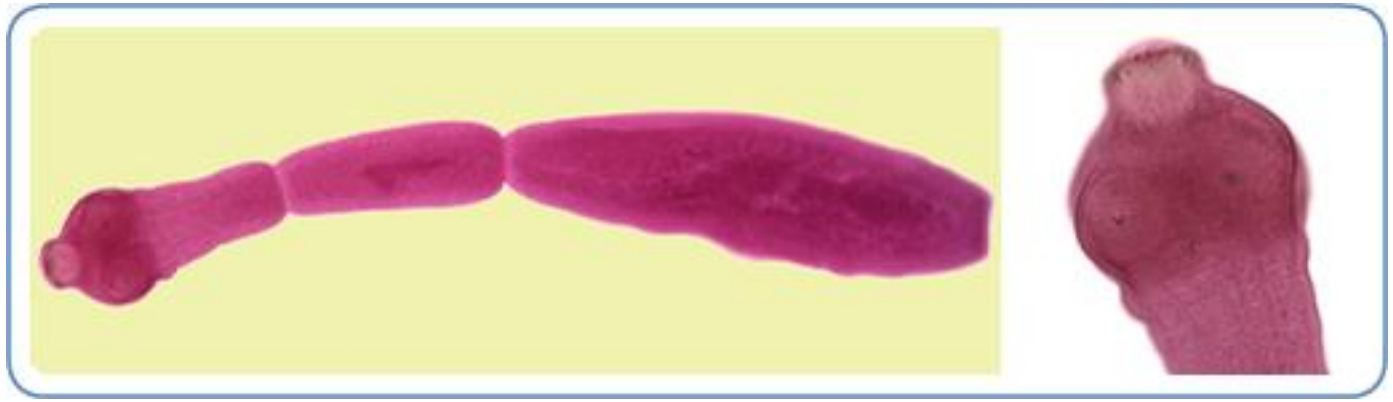
Hydatid Disease

- Pathophysiology
- Epidemiology
- Clinical manifestations
- Diagnosis and Imaging
- Management



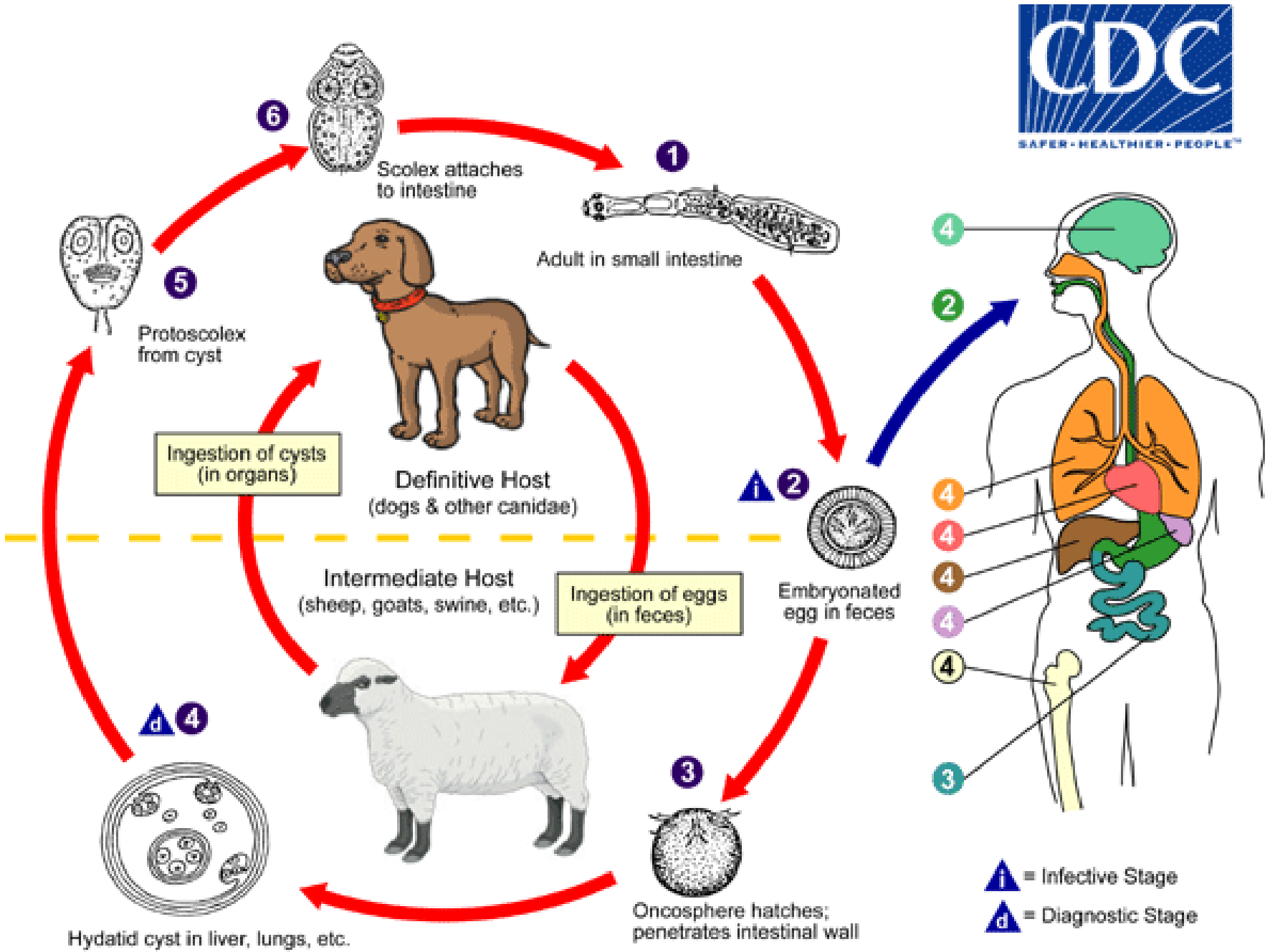
Hydatid Disease

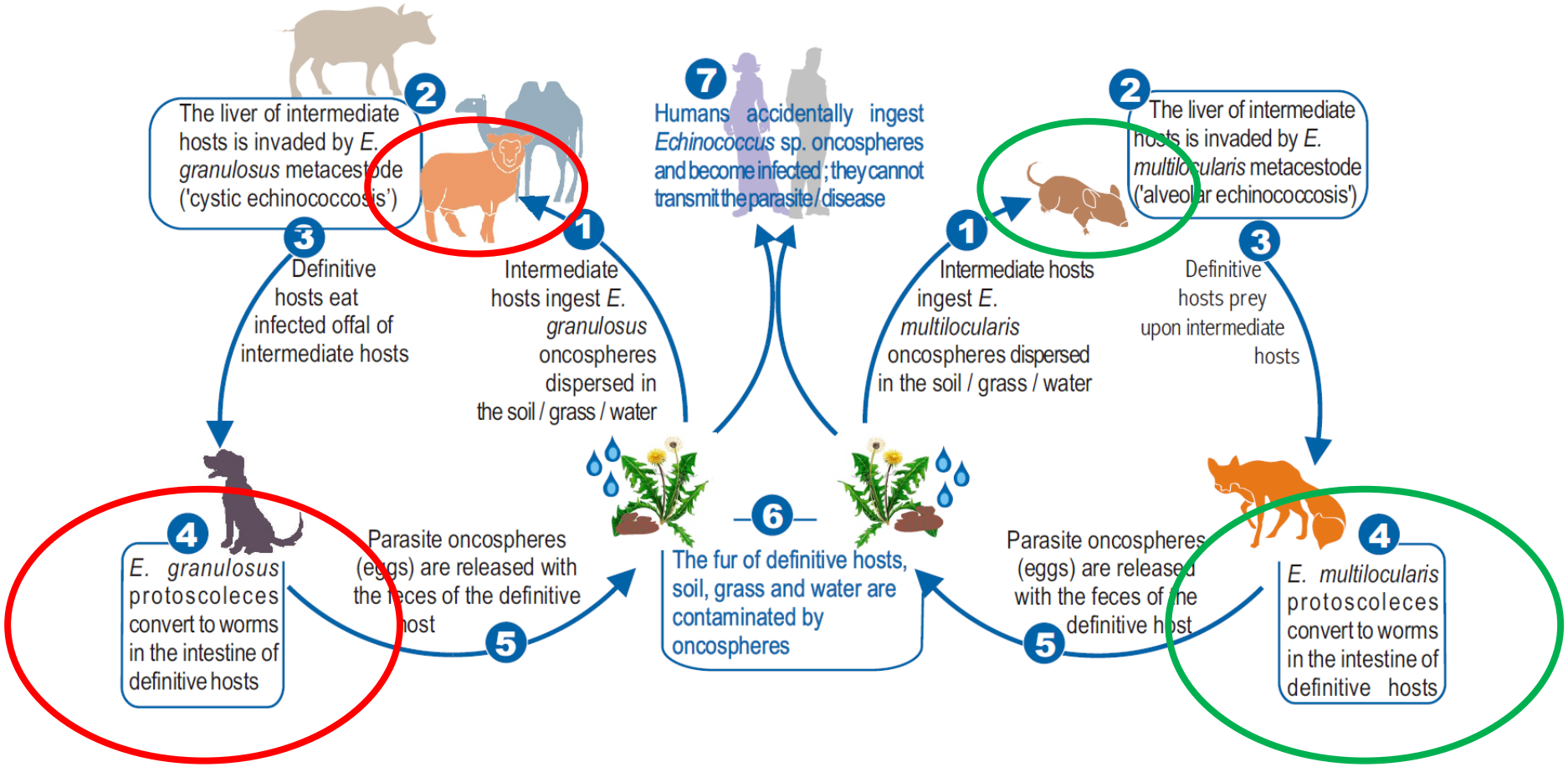
- Echinococcal disease is caused by infection with the metacestode stage of the **tapeworm *Echinococcus***, which belongs to the family *Taeniidae*
- Several species of *Echinococcus* produce infection in humans
- ***E. granulosus*** and ***E. multilocularis*** are the most common, causing **cystic echinococcosis** and **alveolar echinococcosis** respectively



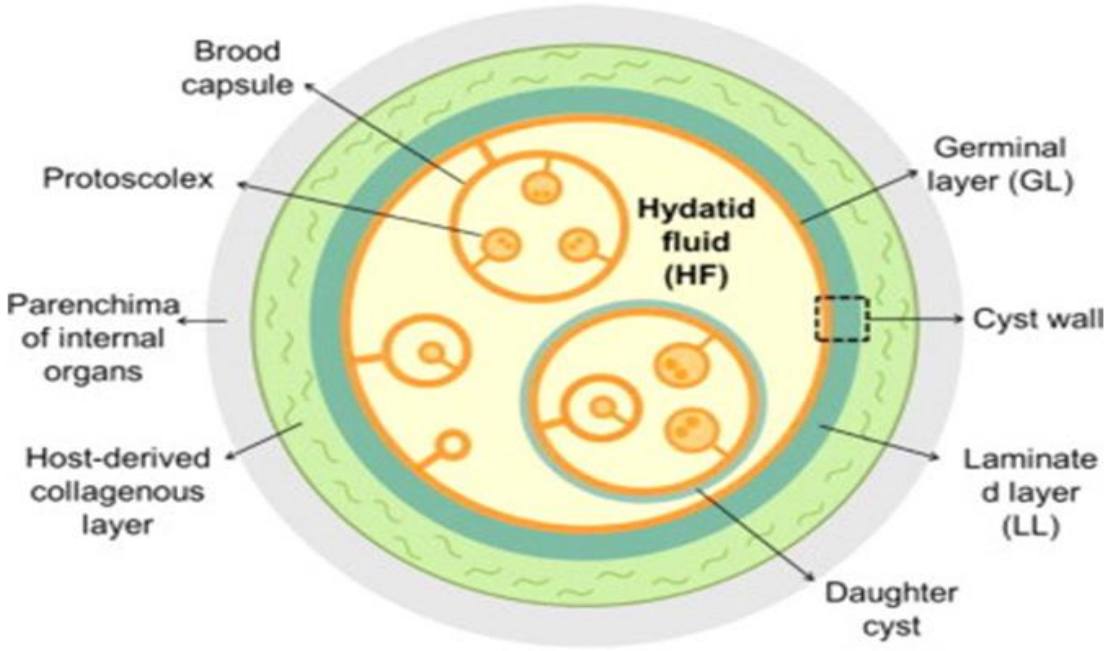
Hydatid disease

- ***E. granulosus* = Cystic echinococcosis**
- ***E. multilocularis* = Alveolar echinococcosis**


CDC
SAFER·HEALTHIER·PEOPLE™
6
Scolex attaches to intestine
1
Adult in small intestine
5
Protoscolex from cyst
Ingestion of cysts (in organs)
Definitive Host
(dogs & other canidae)
i 2
Embryonated egg in feces
Intermediate Host
(sheep, goats, swine, etc.)
Ingestion of eggs (in feces)
d 4
Hydatid cyst in liver, lungs, etc.
3
Oncosphere hatches; penetrates intestinal wall
4
2
4
4
4
4
4
3
i = Infective Stage
d = Diagnostic Stage


2
The liver of intermediate hosts is invaded by E. granulosus metacestode ('cystic echinococcosis')
3
Definitive hosts eat infected offal of intermediate hosts
1
Intermediate hosts ingest E. granulosus oncospheres dispersed in the soil / grass / water
7
Humans accidentally ingest Echinococcus sp. oncospheres and become infected; they cannot transmit the parasite / disease
2
The liver of intermediate hosts is invaded by E. multilocularis metacestode ('alveolar echinococcosis')
1
Intermediate hosts ingest E. multilocularis oncospheres dispersed in the soil / grass / water
3
Definitive hosts prey upon intermediate hosts
4
E. granulosus protoscoleces convert to worms in the intestine of definitive hosts
Parasite oncospheres (eggs) are released with the feces of the definitive host
5
6
The fur of definitive hosts, soil, grass and water are contaminated by oncospheres
Parasite oncospheres (eggs) are released with the feces of the definitive host
5
4
E. multilocularis protoscoleces convert to worms in the intestine of definitive hosts

- The hydatid cyst wall is composed of three layers:
  - The outermost layer – “pericyst” formed by the protective response of the **host tissue**
  - Acellular middle **laminated layer** – “ectocyst” allows for the passage of nutrients
  - Innermost **germinal layer** – “endocyst” produces scolices toward the inner side and laminated membrane on the outer



Ikram C, Ahsan C, Hadi M, Miral M, Alaa A S. et al. Pulmonary Hydatid Cysts Causing Massive Hemoptysis and Hydatoptysis Treated Surgically: A Case Report and Review of Literature. Int J Pul & Res Sci. 2018

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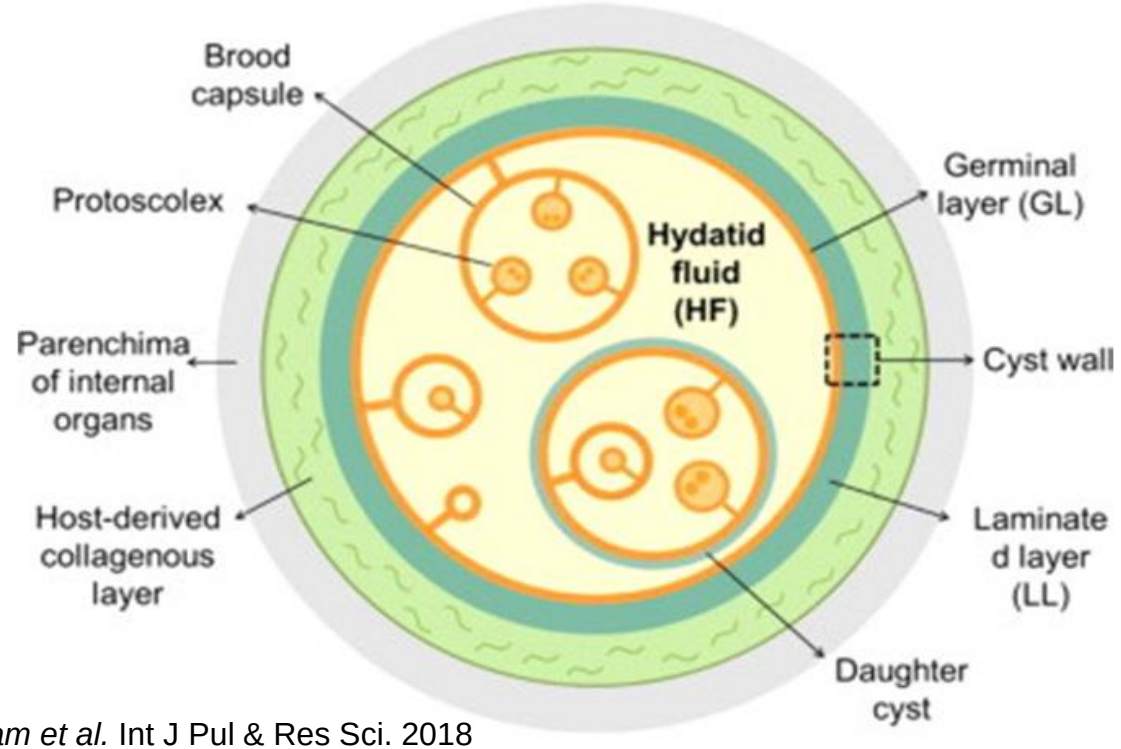
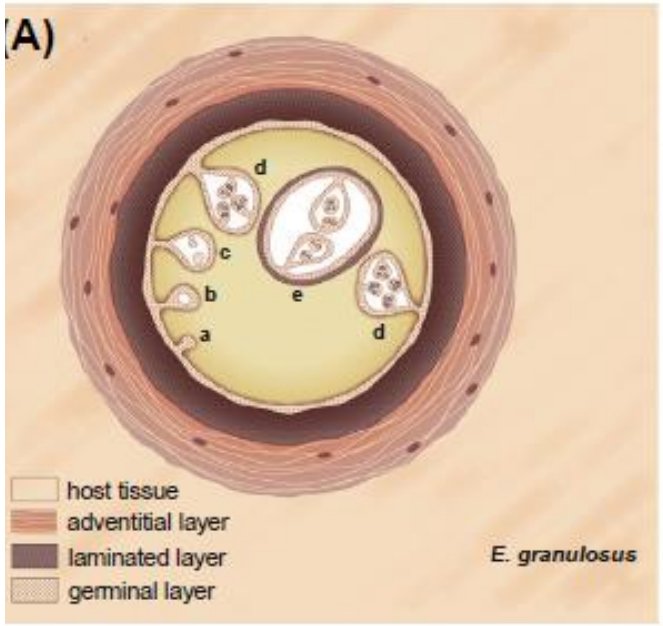
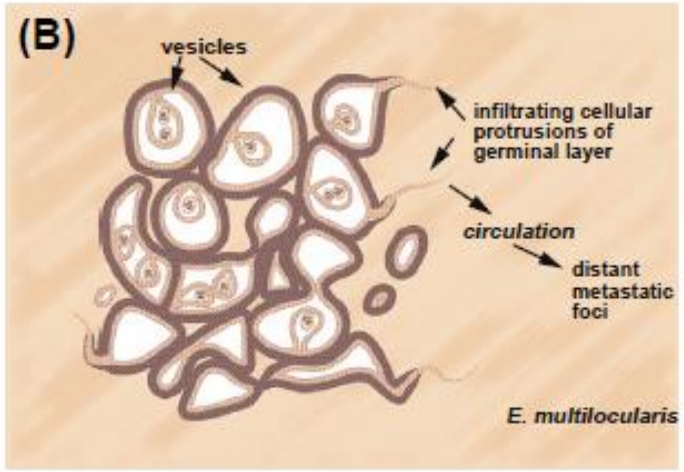
Cyst characteristics of the four echinococcal species

Species	Larval form in humans	Cyst components	Cyst growth
E. granulosus	Cystic, unilocular, expansile	Metacestode has an internal germinative layer (endocyst) surrounded by a parasite-derived acellular laminated layer (exocyst), which in turn is surrounded by a host-derived adventitial layer (pericyst).	Cells bud internally within the cystic cavity, then vacuolate and become "brood" capsules. Protoscolices develop within the brood capsules.
E. multilocularis	Multilocular, infiltrative	Very thin laminated layer only and no pericyst, which enables tissue invasion.	Germinative layer of the metacestode proliferates within cyst and exogenously to infiltrate host tissue. Cells from the germinative layer can detach and metastasize to other organs.



Ikram et al. Int J Pul & Res Sci. 2018

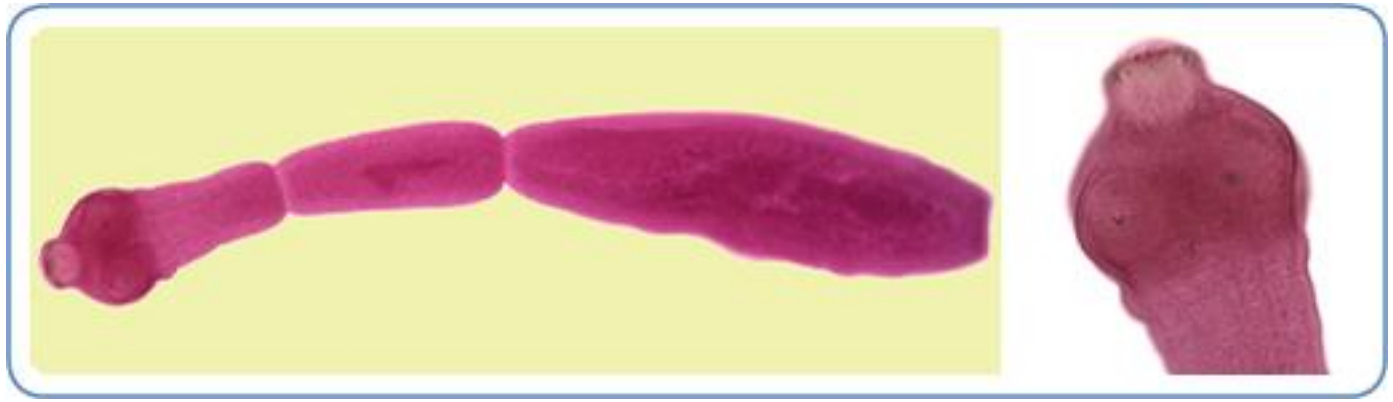




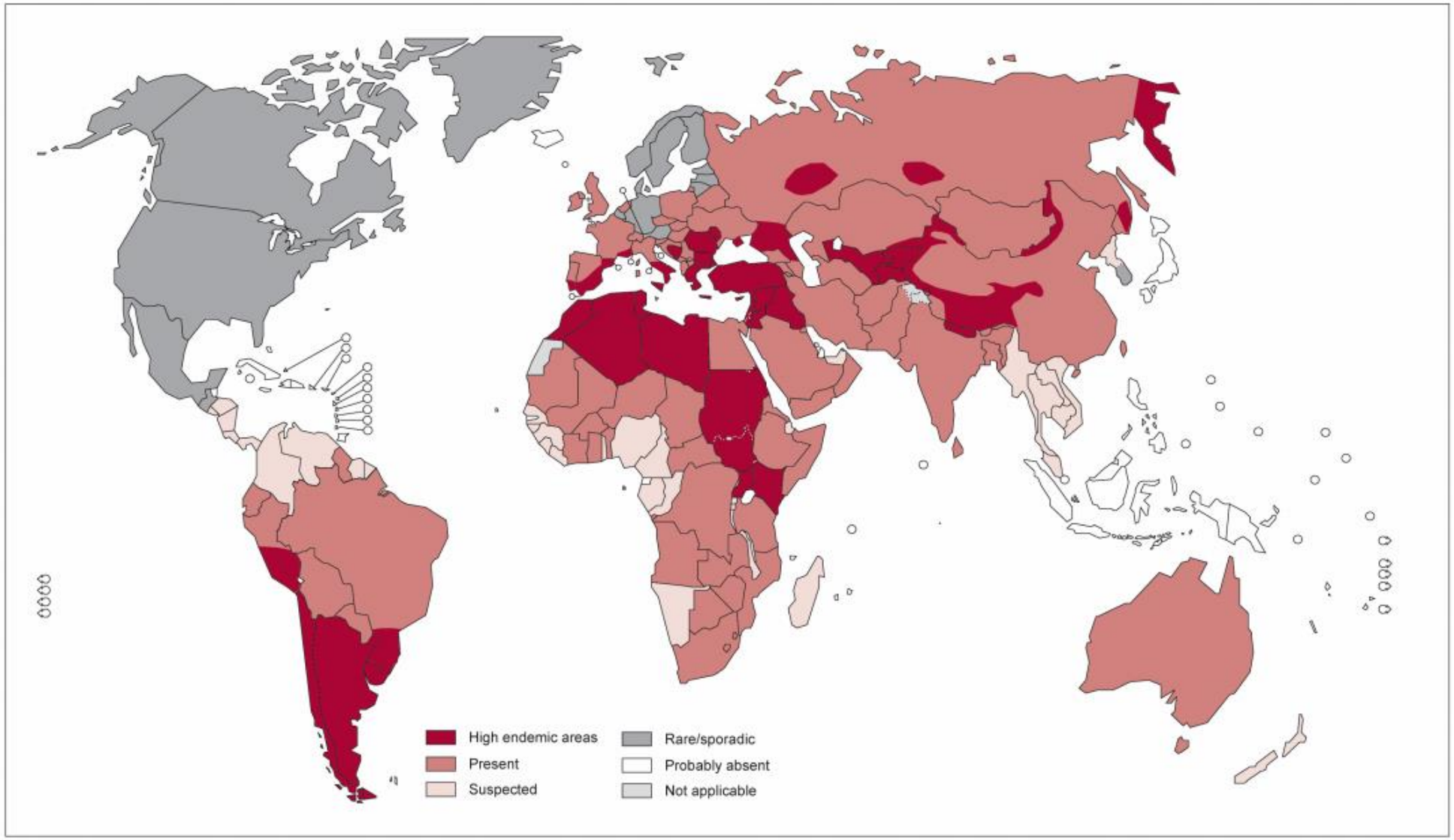
Kern et al. Advances in Parasitology 2017

Hydatid Disease

- Pathophysiology
- Epidemiology
- Clinical manifestations
- Diagnosis and Imaging
- Management



Distribution of *Echinococcus granulosus* and cystic echinococcosis, worldwide,2011

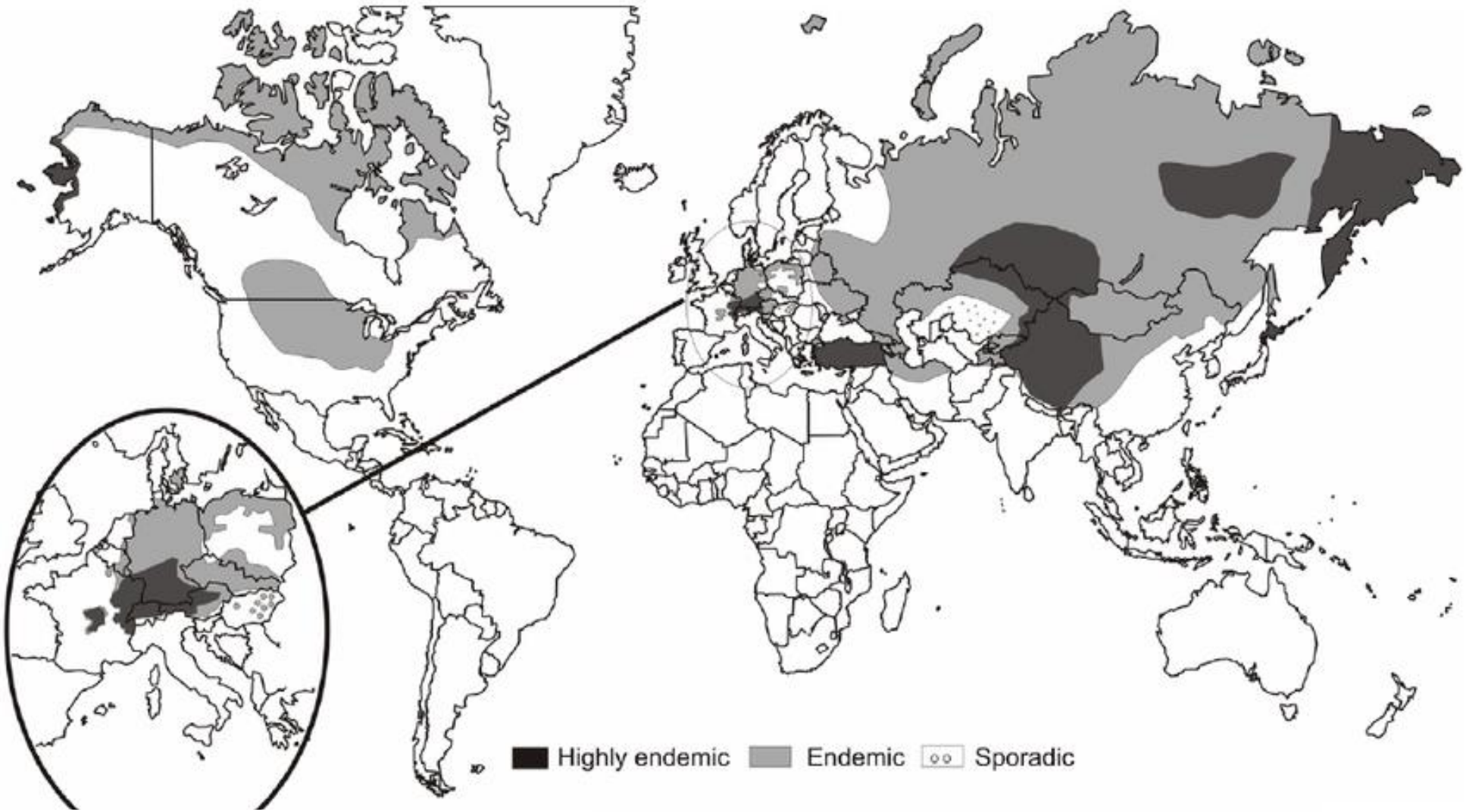


The boundaries and names shown and the designations used on this map do not imply the expression of any opinion whatsoever on the part of the World Health Organization concerning the legal status of any country, territory, city or area or of its authorities, or concerning the delimitation of its frontiers or boundaries. Dotted lines on maps represent approximate border lines for which there may not yet be full agreement. © WHO 2012. All rights reserved

Data Source: World Health Organization
Map Production: Control of Neglected Tropical Diseases (NTD)
World Health Organization

World Health Organization

World distribution of alveolar Echinococcus



The Global Burden of Alveolar Echinococcosis

Paul R. Torgerson[1,2*], Krista Keller[1], Mellissa Magnotta[1], Natalie Ragland[1]

June 2010

PLoS NEGLECTED TROPICAL DISEASES

- **Cystic echinococcosis** (*Echinococcus granulosus)* Africa, Europe, Asia, the Middle East, Central and South America, and in rare cases, North America
- **Alveolar echinococcosis (***Echinococcus multilocularis)* found across the globe and is especially prevalent in the northern latitudes of Europe, Asia, and North America

EMERGING RISK:
ECHINOCOCCUS MULTILOCULARIS IN ONTARIO

***Echinococcus multilocularis* (EM)** is also known as the fox tapeworm. Foxes, coyotes and other canids (including domestic dogs), and rarely cats, can carry adult worms in their intestinal tracts and shed the tapeworm eggs in their feces.

The eggs are usually ingested by rodents or other small mammals. They hatch in the intestine, and the larvae then migrate primarily to the liver and form budding cysts that behave like a malignant tumour. This is called **alveolar echinococcosis (AE)**. Sometimes AE occurs in dogs too.

Humans are an accidental intermediate host for EM. If a person ingests the eggs from the feces of an infected dog or wild canid, then AE can develop. The cysts grow slowly, so the clinical incubation period can be 5-15 years. **AE can be very difficult to treat** due to the invasive growth of the parasitic cysts.

FACT:

EM was known to exist in parts of central and northern Canada for decades, but until 2012 locally-acquired cases had never been detected in Ontario (people or animals)

FACT:

Since 2012, a total of 5 dogs, 2 lemurs and a chipmunk have been diagnosed with AE in Ontario. Only one dog had a history of travel outside the province

FACT:

Dogs are thought to develop AE primarily by ingesting large numbers of EM eggs in the environment, which suggests that EM is likely now present in Ontario wildlife

THROUGH A UNIVERSITY OF GUELPH RESEARCH STUDY CO-SPONSORED BY OAHN AND BAYER ANIMAL HEALTH,

FECAL SHEDDING OF EM WAS CONFIRMED IN FOXES AND COYOTES IN PARTS OF SOUTHERN AND EASTERN ONTARIO IN 2016.

ADDITIONAL TESTING WILL BE DONE IN 2017.

WHAT CAN VETERINARIANS DO?

- ✓ Emphasize the importance of routine fecal exams for dogs at high risk of exposure (e.g. dogs that hunt small mammals, or dogs imported from endemic areas) – but remember that the eggs can be hard to detect
- ✓ Pets shedding tapeworm eggs or at high risk of exposure should be dewormed monthly with praziquantel
- ✓ Unusual masses in the liver or elsewhere in the body should be tested to confirm they are not AE

WHAT CAN OWNERS DO?

- ✓ Don't allow pets to hunt or scavenge other animals, and don't allow hunting dogs to eat raw offal
- ✓ Pick up pet feces promptly to prevent contamination of the environment, and wash hands thoroughly when done
- ✓ If working with soil that may be contaminated with feces from dogs, cats or wild canids, wear gloves and wash hands thoroughly when done

The Ontario Animal Health Network is funded in part through Growing Forward 2, a federal-provincial-territorial initiative.

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For references and to learn more please visit: www.oahn.ca

Table 2 Current taxonomy of *Echinococcus*

Species	Strain/genotype	Known intermediate hosts	Known definitive hosts	Infectivity to humans	Disease
Echinococcus granulosus	Sheep/G1	Sheep (cattle, pigs, camels, goats, macropods)	Dog, fox, dingo, jackal and hyena	Yes	CE
	Tasmanian sheep/G2	Sheep (cattle?)	Dog, fox	Yes	CE
	Buffalo/G3	Buffalo (cattle?)	Dog, fox?	Yes	CE
Echinococcus equinus	Horse/G4	Horses and other equines	Dog	Probably not	CE?
Echinococcus ortleppi	Cattle/G5	Cattle	Dog	Yes	CE
Echinococcus canadensis	Cervids/G8,G10	Cervids	Wolves, dog	Yes	CE
Echinococcus intermedius	Camel/Pig/G6/G7	Camels, pigs, sheep	Dog	Yes	CE
Echinococcus felidis	Lion/?	Warthog, (zebra, wildebeest, bushpig, buffalo, various antelope, giraffe Hippopotamus?)	Lion	?	-
Echinococcus multilocularis	Some isolate variation	Rodents, domestic and wild pig, dog, monkey, (horse?)	Fox, dog, cat, wolf, racoon-dog, coyote	Yes	AE
Echinococcus shiquicus	?	Pika and ?	Tibetan fox and?	?	AE?
Echinococcus vogeli	None reported	Rodents	Bush dog	Yes	PE
Echinococcus oligarthra	None reported	Rodents	Wild felids	Yes	PE

R.C.A. Thompson, Advances in Parasitology, Volume 95, 2017

E.canadensis

- “Northern biotype”, “Alaskan Canadian type”
  - Definitive host – domestic dogs, wolves
  - Intermediate hosts – Caribou, moose, elk
- Most infections in Alaskan native Americans,
- Pathogenicity – fairly benign, with smaller cysts, asymptomatic and sometimes self resolving

Roche Cerda et al. Emerging inf dis 2018

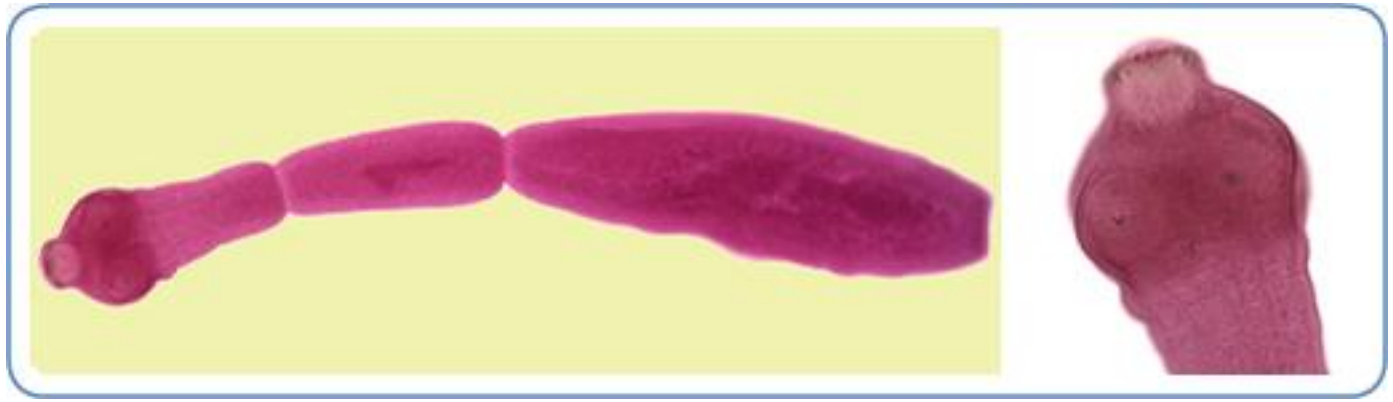
Hydatid disease at SickKids

- 5 patients diagnosed Since 1998
- All were foreigners from endemic areas
- 1/5 with pulmonary disease – 4.5 year old
- 4/5 liver disease, ages 10-17

Data courtesy of Dr Kescha Kazmi

Hydatid Disease

- Pathophysiology
- Epidemiology
- Clinical manifestations
- Diagnosis and Imaging
- Management



Echinococcus Granulosus – Cystic Echinococcosis (CE)

- Initial phase of primary infection is always asymptomatic
- Many infections are acquired in childhood but do not cause clinical manifestations until adulthood
- Approximately **50%** of detected cases occur in **asymptomatic** patients
- Although highly variable, cysts typically increase in diameter at a rate of 1-5 cm/year
- Cysts may be found in almost any site of the body, either from primary inoculation or via secondary spread
- **Liver-** ~ 2/3 of patients, **lungs-** ~25%
- Other organs include - brain, muscle, kidneys, bone, heart and pancreas
- Single-organ involvement occurs in 85 to 90%
- One cyst is observed in more than 70%

Moro et al. Clin Infect Dis. 1999

Clinical Manifestations (CE)

- **The lungs are the most common site in the pediatric population** and the second-most common site in adults
- The lungs facilitate the cyst's growth due to negative pressure and their compressible nature, as a result, pulmonary hydatid cysts grow **three times faster than in the liver**
- Calcification and daughter cyst formation in lung hydatids are rare
- lower lobes are the most common location in the lungs (in 60% of cases) with the right basal lobe being more common
- In 30% of cases, there is more than one cyst, and they can be bilateral in 20% of cases

Moro et al. Clin Infect Dis. 1999

Clinical Manifestations (CE)

- The most common symptoms of pulmonary CE include **cough** (53-62%), chest pain (49-91%), dyspnea (10-70%), hemoptysis (12-21%)
- Less frequent symptoms include malaise, nausea and vomiting, and thoracic deformations
- 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)

Arinc et al. Int J Surg. 2009

Arroud et al. Pediatr Surg Int. 2009

Most commonly reported presenting manifestations among cases of pulmonary cystic echinococcosis treated in hospitals*

Country	Ref	Total pulmonary CE	Cough	Chest pain	Fever	Hemoptysis	Dyspnea	Cyst content expectoration	Asymptomatic
Syria	56	206	112 (54.4%)	75 (36.4%)	–	39 (18.9%)	52 (25.2%)	21 (10.2%)	–
Egypt	57	56	19 (17.8%)	16 (28.6%)	6 (10.7%)	12 (21.4%)	15 (26.8%)	3 (5.4%)	4 (7.1%)
Iran	58	120	75 (62.5%)	10 (8.3%)	20 (16.7%)	10 (8.3%)	–	–	5 (4.2%)
Turkey	59	1032	846 (82.0%)	629 (60.9%)	124 (12.0%)	217 (21.0%)	258 (25.0%)	155 (15.0%)	165 (16.0%)
Turkey	60	405	101 (24.9%)	208 (51.3%)	8 (2.0%)	1 (0.2%)	–	–	65 (16.0%)
Turkey	61	288	155 (53.8%)	142 (49.3%)	40 (13.9%)	61 (21.2%)	29 (10.1%)	14 (4.9%)	30 (10.4%)
Turkey	62	107	43 (40.2%)	30 (28.0%)	16 (14.9%)	2 (1.9%)	–	8 (7.5%)	22 (20.6%)
Turkey	63	70	38 (54.3%)	33 (47.1%)	22 (31.4%)	–	21 (30.0%)	11 (15.7%)	–
Turkey	64	139	68 (48.9%)	48 (34.5%)	39 (28.0%)	11 (7.91%)	–	–	18 (12.9%)
Iran	55	24 (ped)	22 (91.7%)	10 (41.7%)	20 (83.3%)	2 (8.3%)	11 (45.8%)	–	–
Libya	65	43 (ped)	30 (69.8%)	2 (4.7%)	16 (37.2%)	9 (20.9%)	–	–	2 (4.7%)
Turkey	66	112 (ped)	79 (70.5%)	27 (24.1%)	38 (33.9%)	2 (1.8%)	–	2 (1.8%)	–
Turkey	67	102 (ped)	57 (55.9%)	41 (40.2%)	19 (18.6%)	9 (8.8%)	21 (20.6%)	8 (7.8%)	–
Turkey	68	47 (ped)	35 (74.5%)	17 (36.2%)	21 (44.7%)	2 (4.3%)	–	–	–
Turkey	69	33 (ped)	22 (66.7%)	11 (33.3%)	6 (18.2%)	10 (30.3%)	–	–	4 (12.1%)
Morocco	70	23 (ped)	–	20 (86.9%)	–	5 (21.7%)	2 (8.7%)	–	2 (8.7%)
Turkey	71	34† (intact)	9 (26.5%)	17 (50.0%)	4 (11.8%)	2 (5.9%)	6 (17.6%)	–	9 (26.5%)
Turkey	71	33† (ruptured)	15 (45.4%)	16 (48.5%)	12 (36.4%)	11 (3.3%)	14 (42.4%)	5 (15.1%)	1 (3.0%)
Turkey	72	20† (intact-ped)	13 (65.0%)	10 (50.0%)	7 (35.0%)	2 (10.0%)	8 (40.0%)	1 (5.0%)	1 (5.0%)
Turkey	72	14† (ruptured-ped)	14 (100%)	8 (57.1%)	7 (50.0%)	8 (57.1%)	3 (21.4%)	8 (57.1%)	–
Proportion (95% CI) - All ages			51.3% (35.8–66.7%)	39.9% (29.8–50.4%)	14.8% (9.3–21.3%)	12.6% (5.6–21.8%)	23.7% (17.1–31.0%)	8.8% (5.1–13.3%)	12.8% (9.8–16.0%)
Proportion (95% CI) - Pediatric cases only			70.3% (61.2–78.6%)	36.5% (21.4–53.1%)	38.1% (23.4–54.0%)	12.3% (5.5–21.3%)	24.1% (9.5–42.7%)	–	10.2% (4.9–17.3%)

*Other clinical manifestations included tachypnea, sweating, infectious symptoms, pleuritis, pneumothorax, weight loss, purulent sputum, allergic reactions, malaise, nausea and vomiting, abdominal pain, and headache.
†Not included in meta-analysis.

Budke et al. 2013 The American Society of Tropical Medicine and Hygiene

Clinical Manifestations (CE)

- **cyst rupture** - spilling of cyst material containing fragments of larval tissue into the bronchial tree or the pleural cavity
  - Can cause fever and acute hypersensitivity reactions, including **anaphylaxis** d/t release of antigenic material and secondary immunologic reactions
  - **Bronchial tree involvement** can lead to cough, chest pain, hemoptysis, or emesis
  - **Pleural cavity involvement** can cause pneumothorax, pleural effusion, or empyema.
- **Secondary bacterial infection** of the cyst can manifest as a pulmonary abscess with poorly defined margins

Dziri et al. World J Surg. 2009

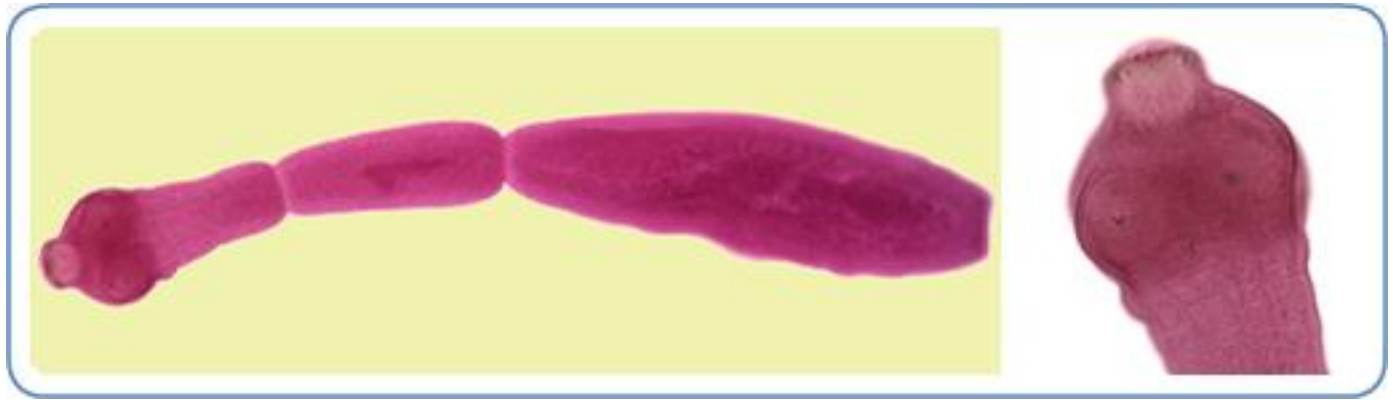
Echinococcus Multilocularis – Alveolar Echinococcosis (AE)

- **Extrahepatic primary disease is very rare (1% of cases)**
- **Usually symptomatic**, although the clinical manifestations are frequently **nonspecific**
- The most common P/S - malaise, weight loss, and RUQ discomfort due to hepatomegaly
- Cholestatic jaundice, cholangitis, portal hypertension, and the Budd-Chiari syndrome can also occur
- If left untreated, **more than 90% of patients will die within 10 years** of the onset of clinical symptoms

Torgerson et al. J Hepatol. 2008

Hydatid Disease

- Pathophysiology
- Epidemiology
- Clinical manifestations
- Diagnosis and Imaging
- Management



Diagnosis (CE)

- Leukopenia, thrombocytopenia, mild eosinophilia, and nonspecific liver function abnormalities may be observed but are not diagnostic
- Eosinophilia is observed in fewer than 15%, generally occurs if there is leakage of antigenic material
- Serologic tests:

A negative serologic test does not rule out echinococcosis!

Sensitivity of serologic tests for echinococcosis at different sites

Site of lesion	Sensitivity of serologic tests
Liver	IgG ELISA: 80 to 90 percent
	IgE ELISA: 82 to 92 percent
	Latex agglutination: 65 to 75 percent
	Hemagglutination: 80 to 90 percent
	Immunoblot (using antigen 5 and/or a B-rich fraction): 80 to 90 percent
	Enzyme-linked immunotransfer blot: 80 percent
Lung	IgG ELISA: 60 to 85 percent
	IgE ELISA: 45 to 70 percent
	Latex agglutination: 50 to 70 percent
	Hemagglutination: 50 to 70 percent
	Immunoblot (using antigen 5 and/or a B-rich fraction): 55 to 70 percent
	Enzyme-linked immunotransfer blot: 55 percent

Ig: immunoglobulin; ELISA: enzyme-linked immunosorbent assay.

UpToDate 2019 (based on studies from 1992 to 2003)

Diagnosis (CE)

- **Antigen assays** — A variety of purified or recombinant diagnostic antigens have been evaluated
- **Up to 50% of patients with echinococcal cysts do not have circulating antigens**
- Latex agglutination or a dot-ELISA to detect echinococcal antigens **from cyst fluid** have excellent sensitivity and specificity
- PCR?

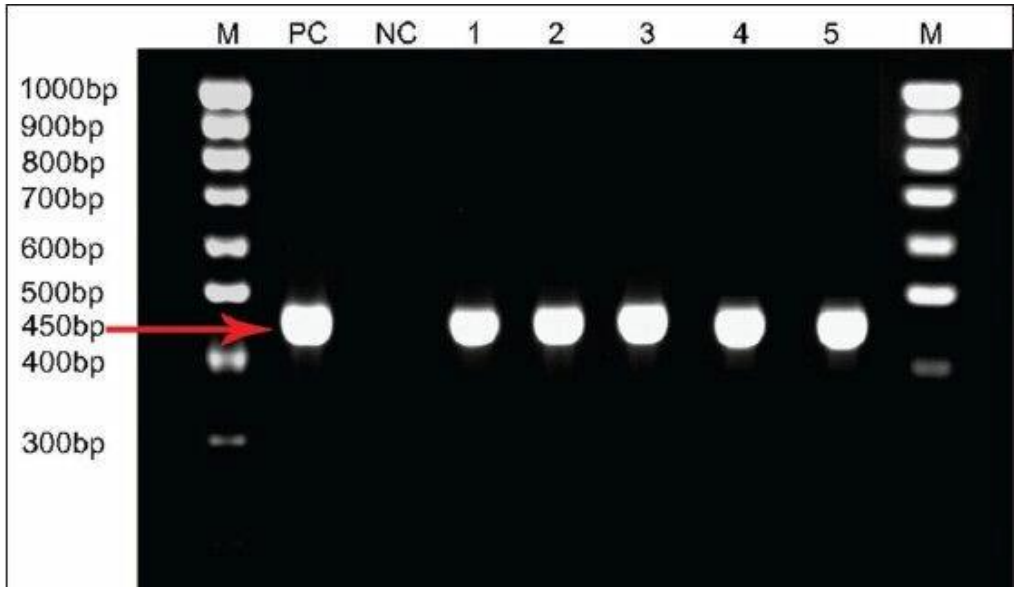
Wang et al. Ann Trop Med Parasitol. 2002

Ravinder, Parija Acta Trop. 1997

Performance of polymerase chain reaction for the diagnosis of cystic echinococcosis using serum, urine, and cyst fluid samples

D. Chaya and Subhash Parija

Tropical Parasitology. 4.1 (January-June 2014): p43.



Polymerase chain reaction amplification of the 450 bp *Echinococcus granulosus* specific NADH1 gene from **five serum samples of surgically confirmed cases with a ruptured cyst**. M: Molecular ladder, PC: Positive control, NC: Negative control, 1-5 depicts the number of cases

Method employed	Surgically and sonographically proven cases of CE (Group I+Group II) (*n*=25)	CE negative control group (Group III+Group IV) (*n*=25)
PCR to detect *Echinococcus granulosus* DNA		
+	5	0
–	20	25
Antibody detection ELISA		
+	23	4
–	2	21
Antibody detection using EITB		
+	23	0
–	2	25

CE: Cystic echinococcosis, PCR: Polymerase chain reaction, DNA: Deoxyribonucleic acid, ELISA: Enzyme-linked immunosorbent assay, EITB: Enzyme immunotransfer blot

Uncomplicated cysts

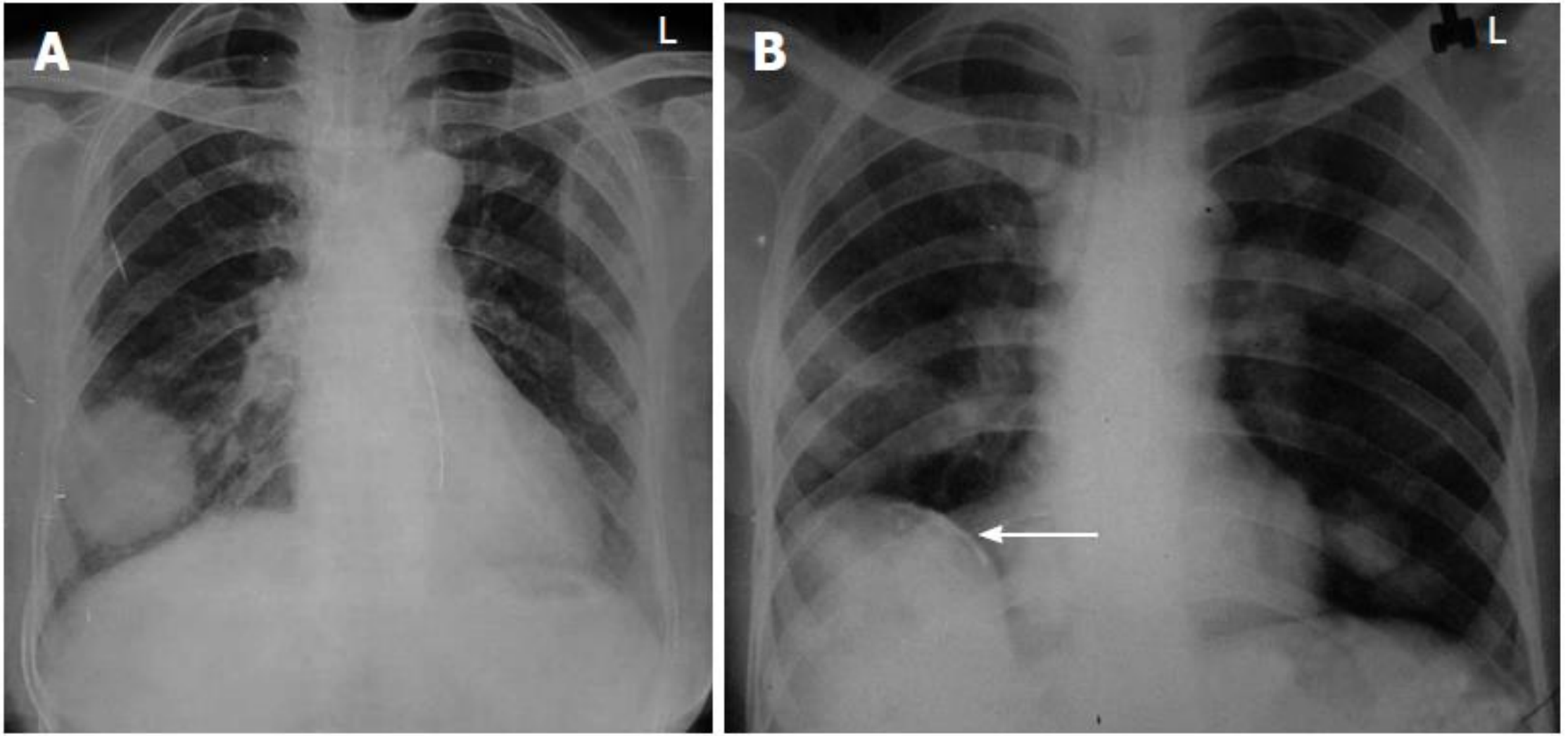
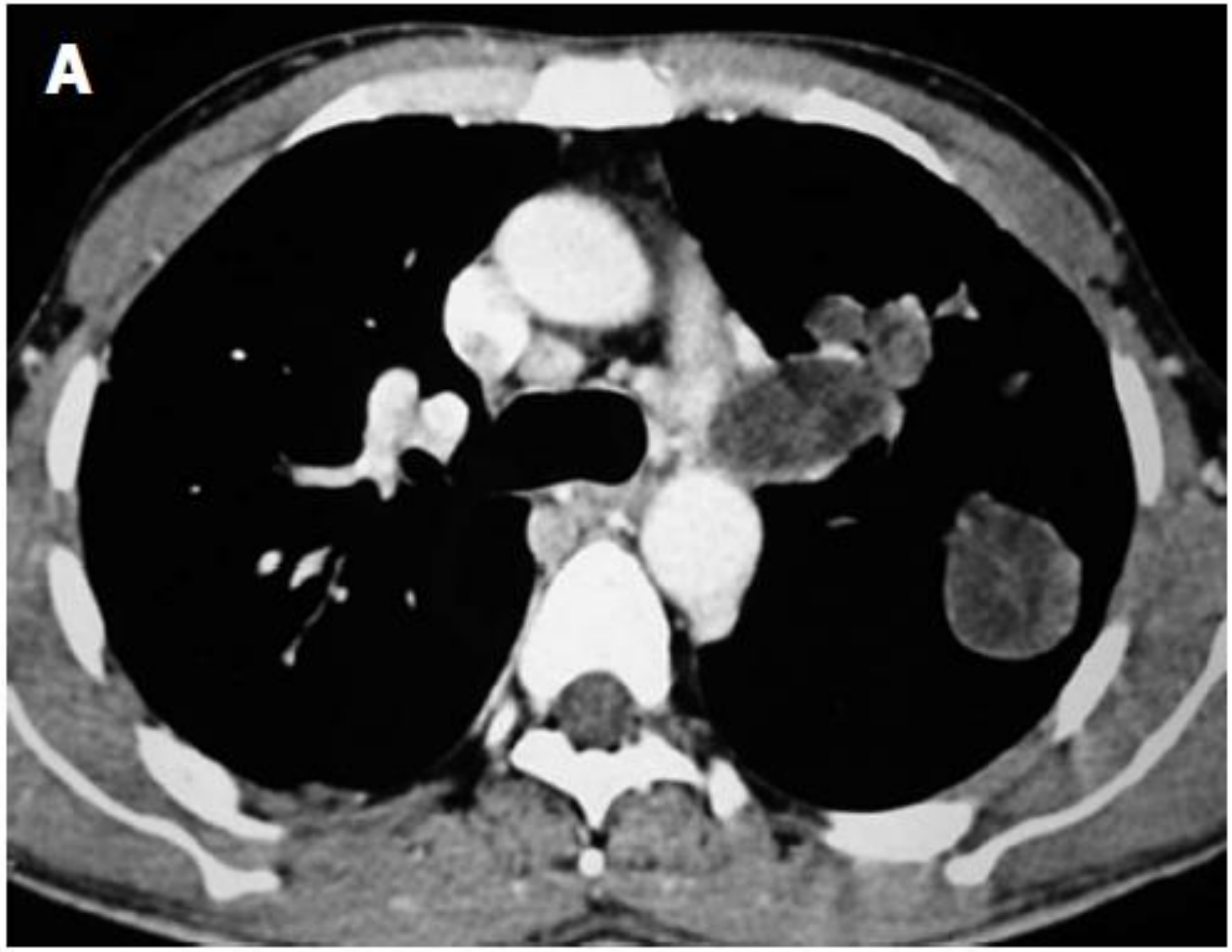


Figure 1 Uncomplicated hydatid cyst. A: Posteroanterior view of chest X-ray showing well defined round radio-opacity in right lower zone; B: Chest X-ray showing multiple well defined round opacities in left lung. Also note presence of calcified cyst in liver (arrow in B), which makes diagnosis of hydatid cyst almost certain.

Garg MK *et al*. *World J Radiol* 2016 June 28

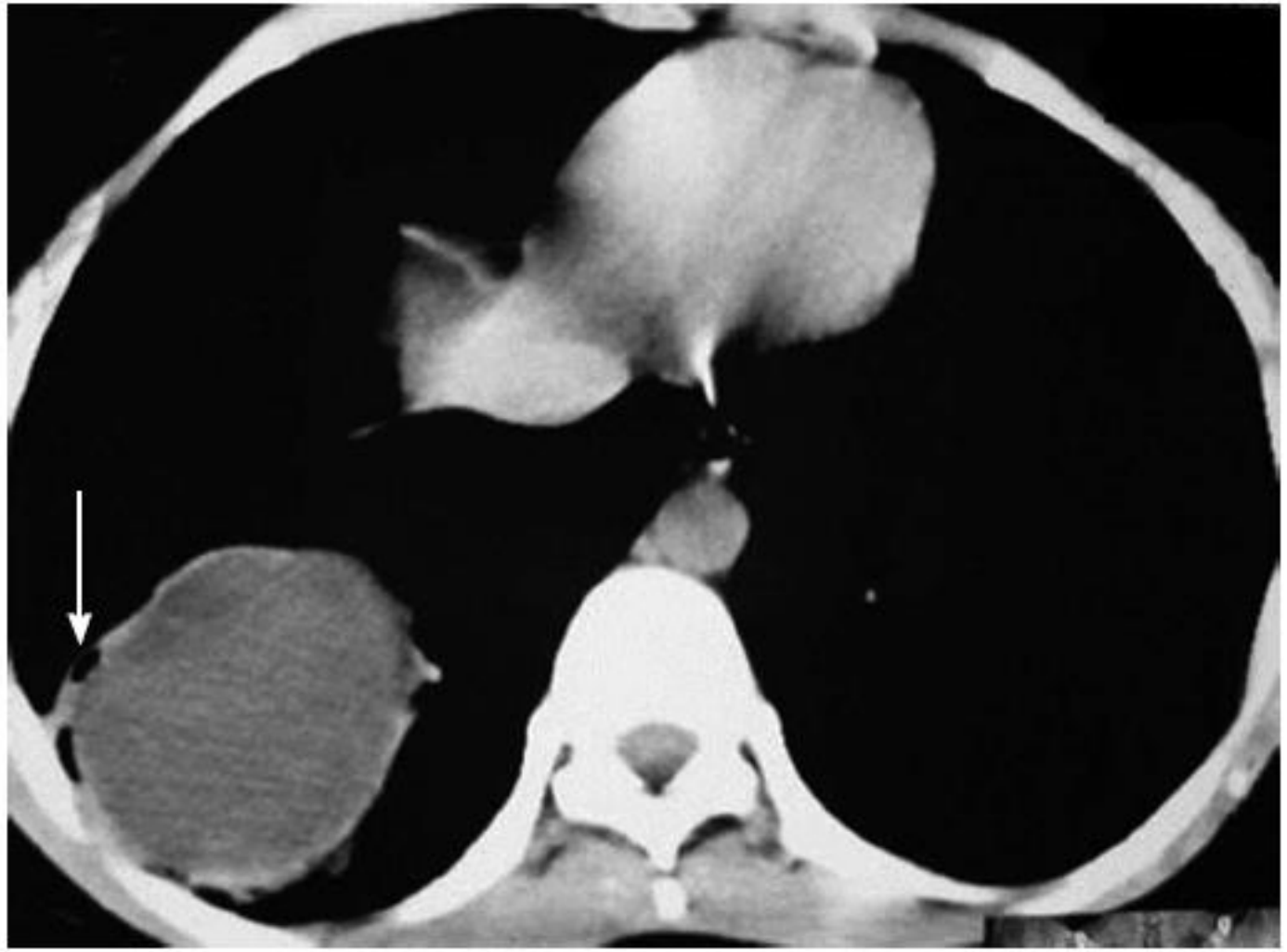
The appearance of cysts
has been compared to **cannon balls** in PA and to **rugby balls** in lateral

Uncomplicated cysts (CT)



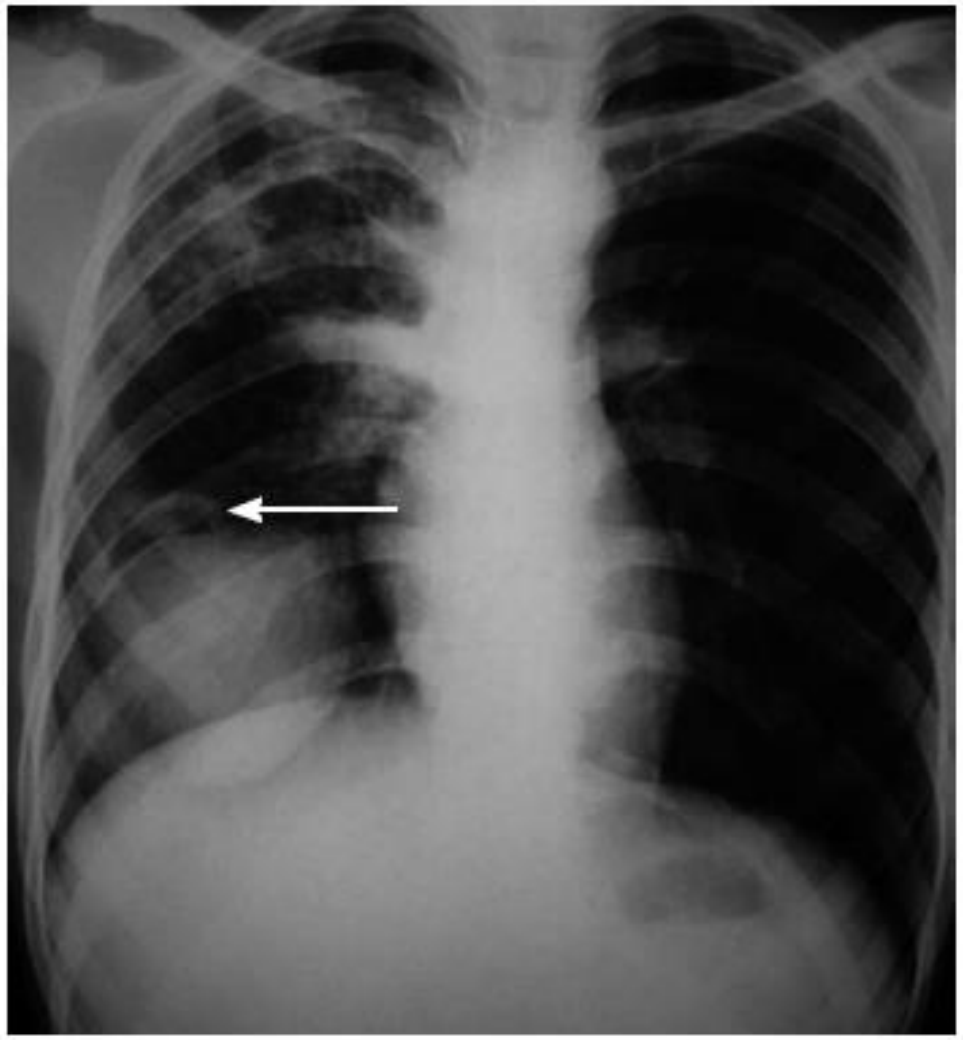
Garg MK *et al*. *World J Radiol* 2016 June 28

Contained rupture of the endocyst (air bubble sign)



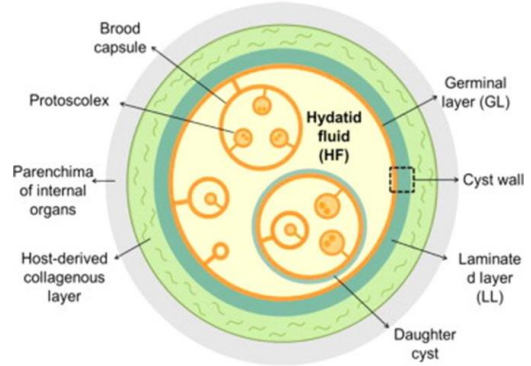
Garg MK *et al*. *World J Radiol* 2016 June 28

Contained rupture of the endocyst (crescent sign)

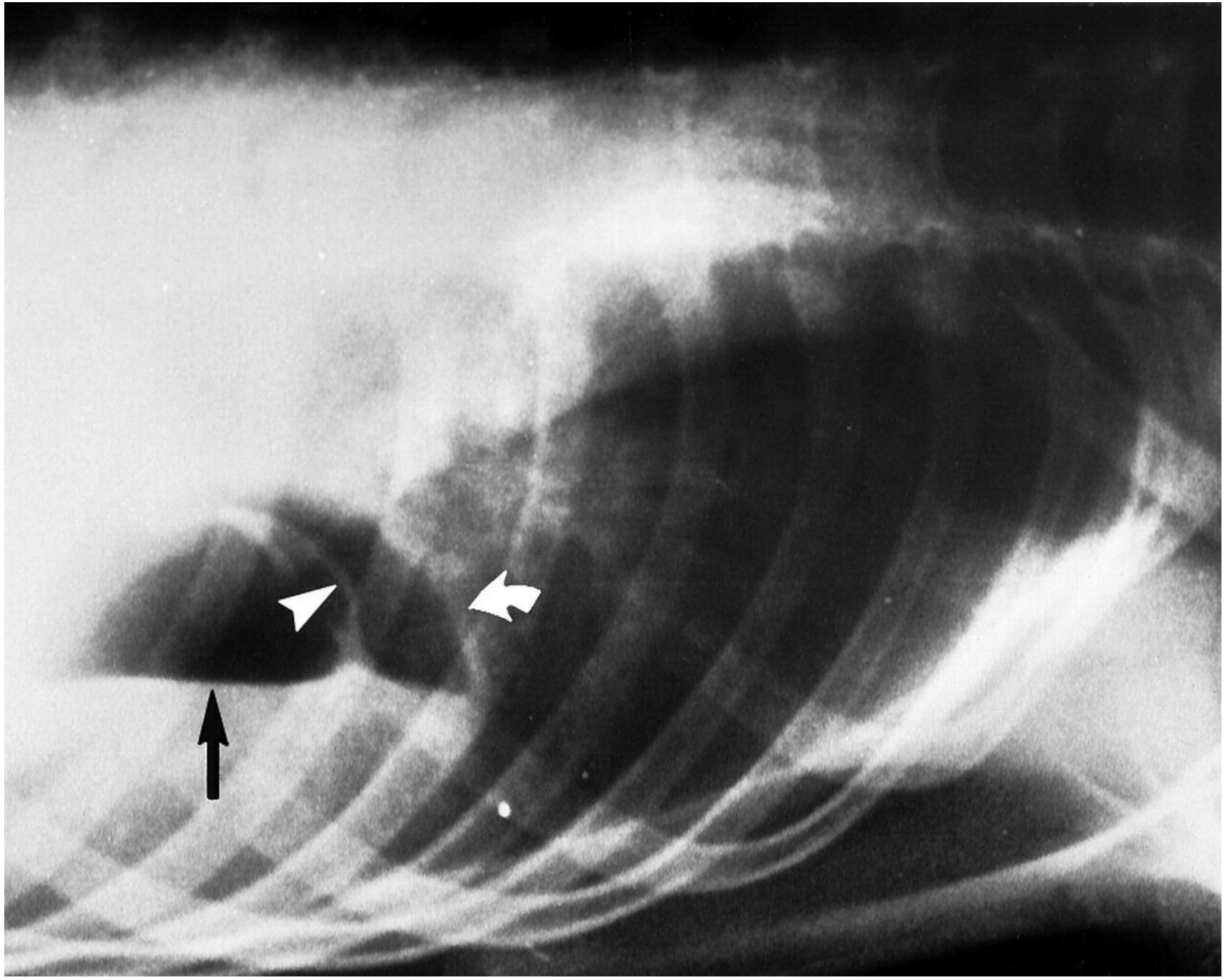


Signs in hydatid disease of lung

Sign	Cause	Radiological representation
Meniscus/ Double arch/ Crescent/ Moon sign	Due to thin crescent of air in the uppermost of the cyst	
Onion peel/ Cumbo sign	Due to air fluid level inside endocyst	
Serpent sign	Collapsed membranes inside the cyst outlined by air	
Water Lilly sign	Completely collapsed cyst floating on the cyst fluid	
Cavity	All contents of cyst breaks out via communicating bronchus	



Garg MK *et al*. *World J Radiol* 2016 June 28



Pedrosa et al. RadioGraphics 2000

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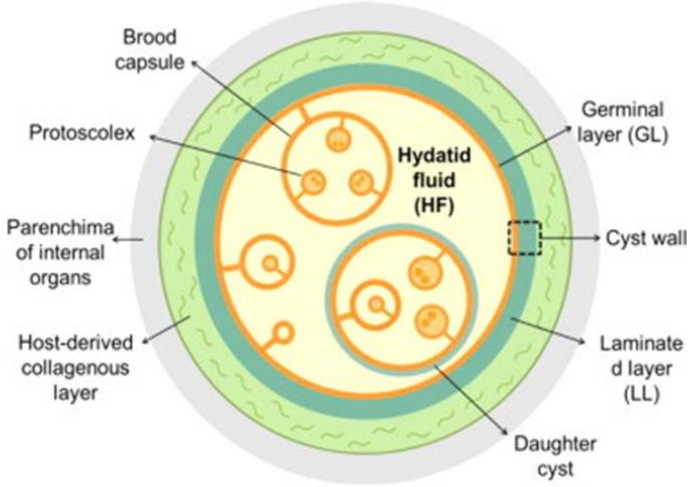
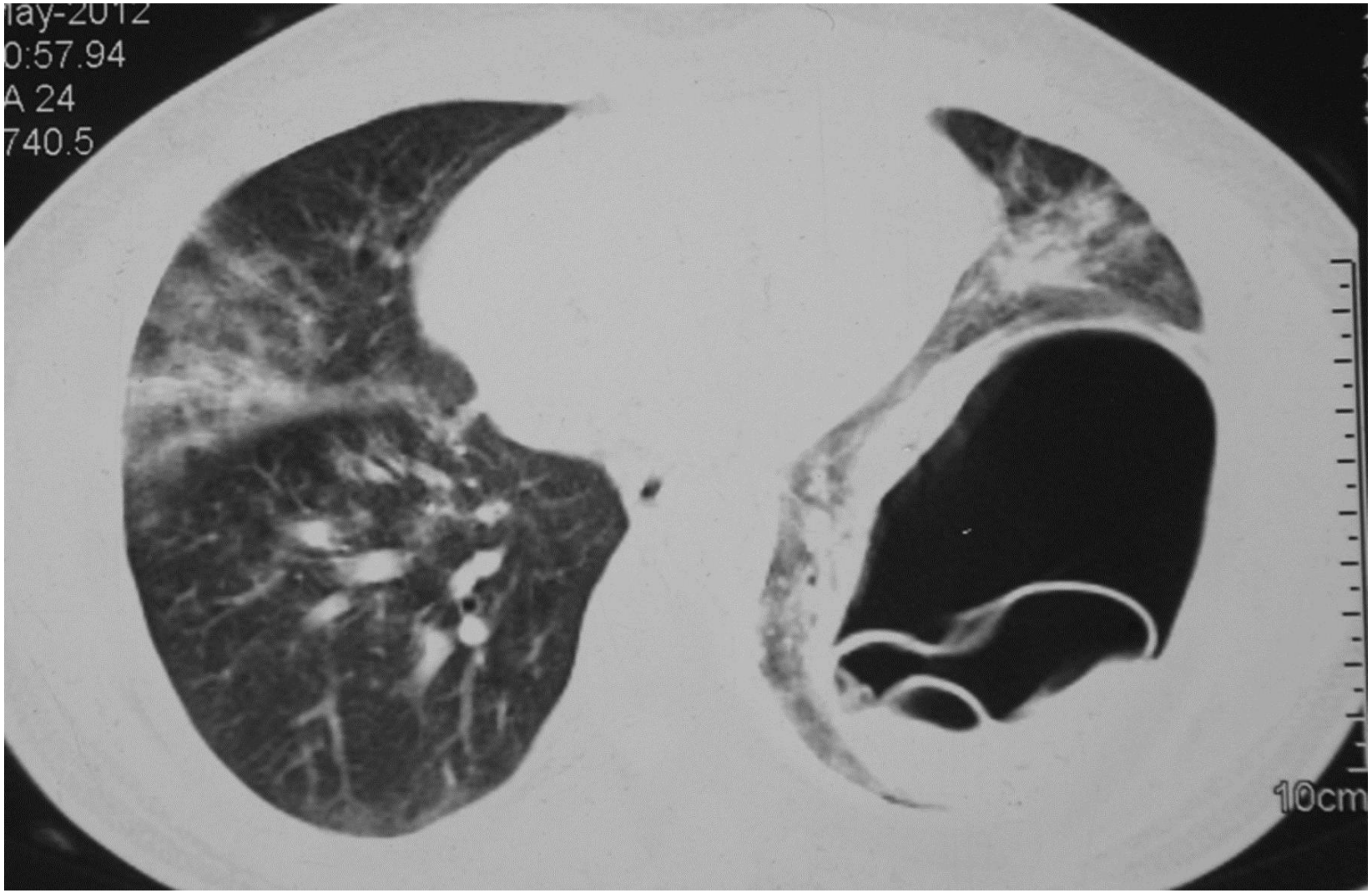


Serpent sign



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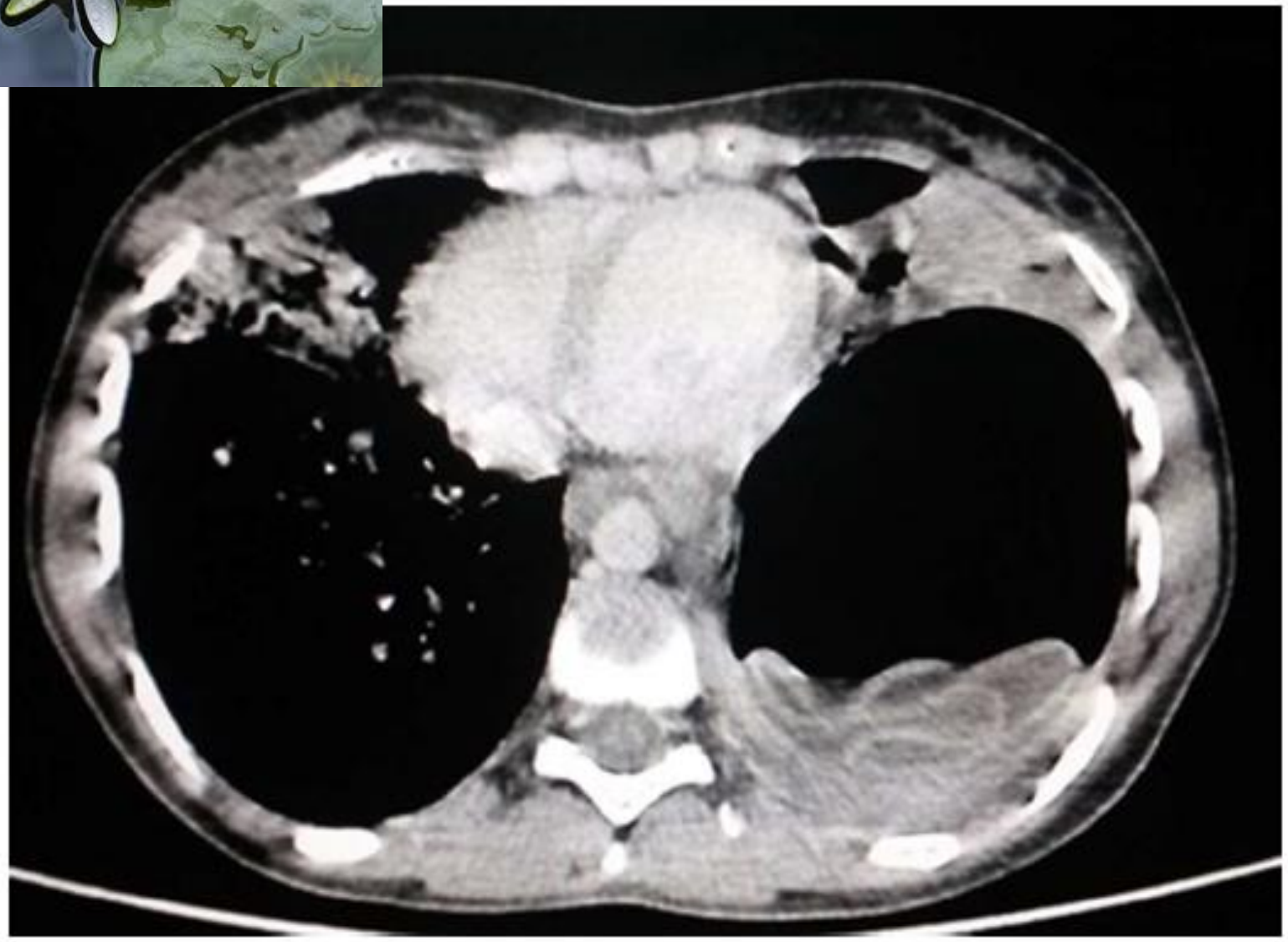
Karan Madan, and Randeep Guleria BMJ Case Reports 2013

Water lily sign



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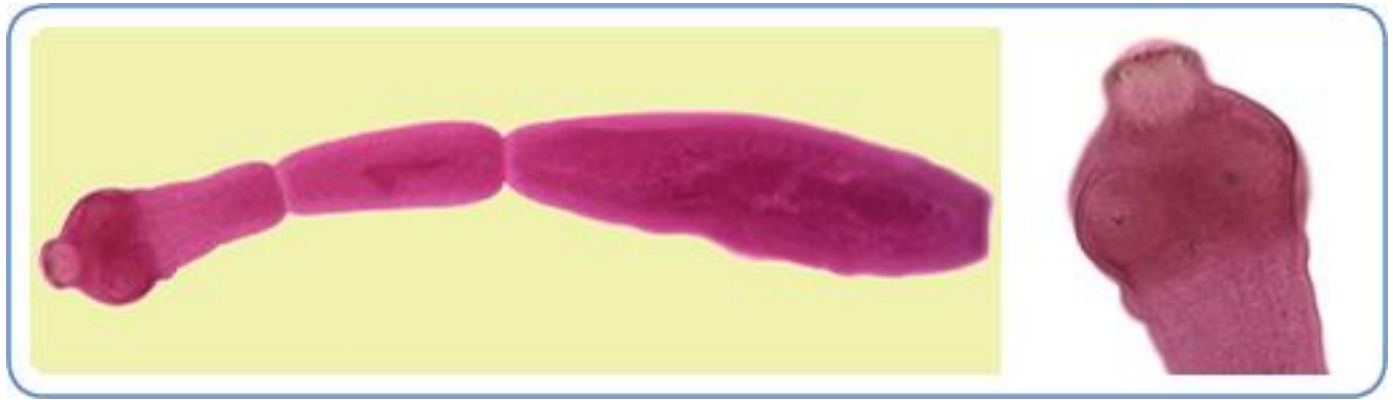

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Acta Tropica

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Review

Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans☆

Enrico Brunetti[a,*,1], Peter Kern[b], Dominique Angèle Vuitton[c], Writing Panel for the WHO-IWGE[2]

[a] Division of Infectious and Tropical Diseases, University of Pavia, IRCCS S.Matteo Hospital Foundation, WHO Collaborating Center for Clinical Management of Cystic Echinococcosis, 27100 Pavia, Italy
[b] Comprehensive Infectious Diseases Centre, University Hospitals, Albert-Einstein-Allee 23, 89081 Ulm, Germany
[c] WHO Collaborating Centre for Prevention and Treatment of Human Echinococcosis, CHU de Besançon/Université de Franche-Comté, 25030 Besançon, France

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- **There is no "best" treatment option for CE** (no clinical trial has compared all the different treatment modalities, including "Watch and Wait."
- Treatment indications are complex and are based on cyst characteristics, available medical/surgical expertise and equipment, and adherence of patients to long-term monitoring
- patients should be referred to recognized, reference and national/regional CE treatment centres

Anti parasitic drug treatment

- Indications:
  - Inoperable pt's with liver/ pulmonary cysts
  - Small cysts (<5cm) respond favourably to benzimidazoles (Typically **Albendazole**, used to be Mebendazole)
  - BMZ should be used to prevent recurrence after surgery
- Contraindications:
  - Cysts with high risk for rupture
  - Early pregnancy (teratogenic in rats and rabbits, small risk in humans)
  - Not effective in large cysts
- Optimal dose and duration – never formally assessed

E. Brunetti et al. / Acta Tropica 114 (2010)

PAIR- Puncture, Aspiration, Injection, Re-aspiration

- Minimally invasive technique used in the treatment of cysts in the liver and other abdominal locations
- Indicated for inoperable patients and those who refuse surgery, in cases of relapse after surgery or failure to respond to BMZ alone
- **Should not be used for lung cysts**

E. Brunetti et al. / Acta Tropica 114 (2010)

Table 1. Characteristics and procedural information for 11 percutaneously aspirated echinococcal cysts in eight patients

Patient #	Preprocedure cyst			Complication		Cough during procedure	Follow-up (months)	Postprocedure cyst			Volume reduction rate (%)
	Type	Size (mm)	Vol. (cc)	Minor	Major			Type	Size (mm)	Vol. (cc)	
1	I	30 × 30 × 32	14	–	–	+	31	IV	14 × 16 × 11	1	93
2	I	31 × 30 × 33	15	–	–	–	23	IV	27 × 27 × 22	8	47
3	I	85 × 63 × 75	198	Fever	RP effusion	+	20	IV	47 × 26 × 29	17	91
	I	54 × 54 × 50	71		LHPT			IV	31 × 29 × 25	11	85
4	I	59 × 47 × 50	68	–	–	–	18	IV	44 × 19 × 20	8	88
	I	50 × 33 × 35	28					IV	52 × 24 × 20	12	57
5	I	97 × 81 × 100	385	Fever	Pneumothorax Abscess	–	Operated				
6	I	37 × 28 × 25	13	–	–	–	10	IV	28 × 22 × 20	6	54
7	I	100 × 90 × 95	419	Fever Dyspnea	–	–	8	IV	60 × 40 × 50	59	86
8	I	40 × 33 × 35	23	–	–	–	8	IV	30 × 27 × 30	12	48
	I	37 × 32 × 35	20					IV	29 × 25 × 25	9	55

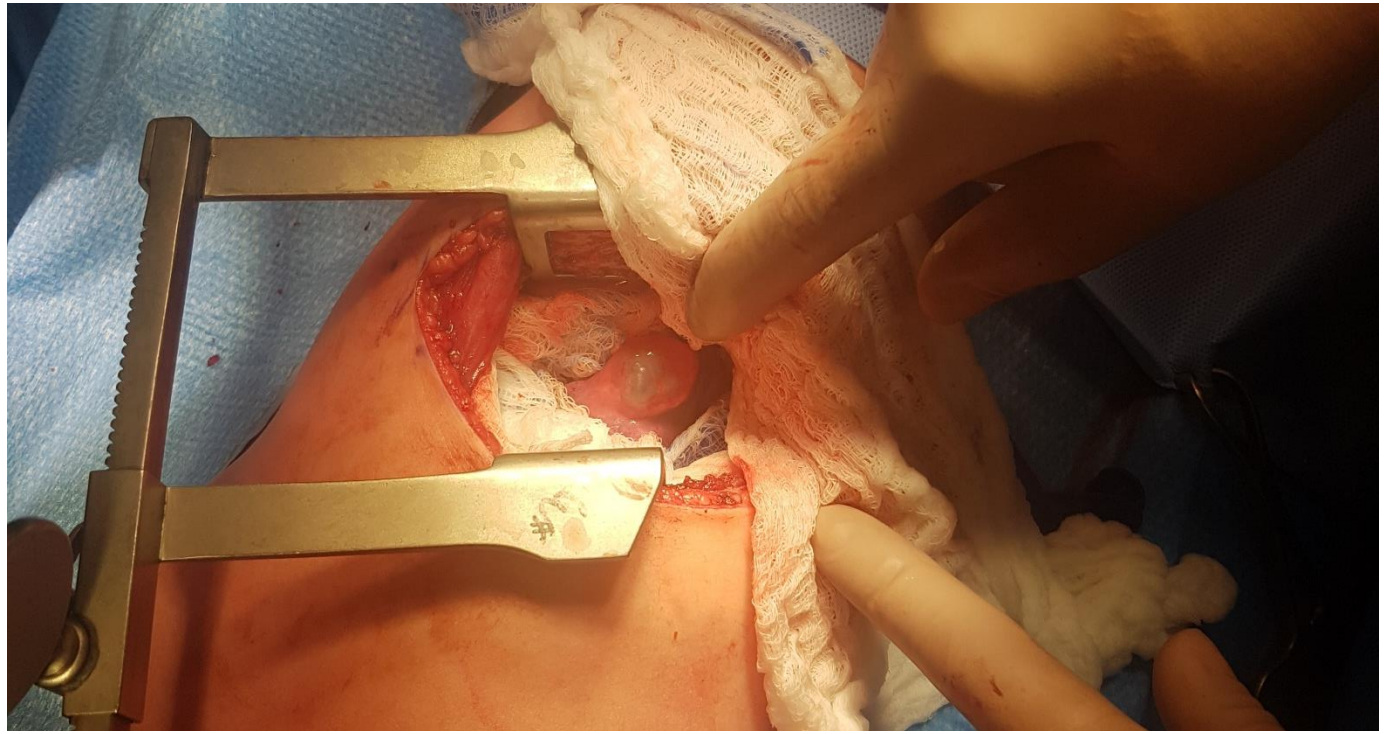
RP: right pleural effusion; LHPT: left hydropneumothorax; Vol.: estimated volume = $A \times B \times C \times 0.49 \times 10^{-3}$ (A, B, C in mm and volume in cubic cm)
Postprocedural measurements are obtained at the last follow-up time indicated in the table

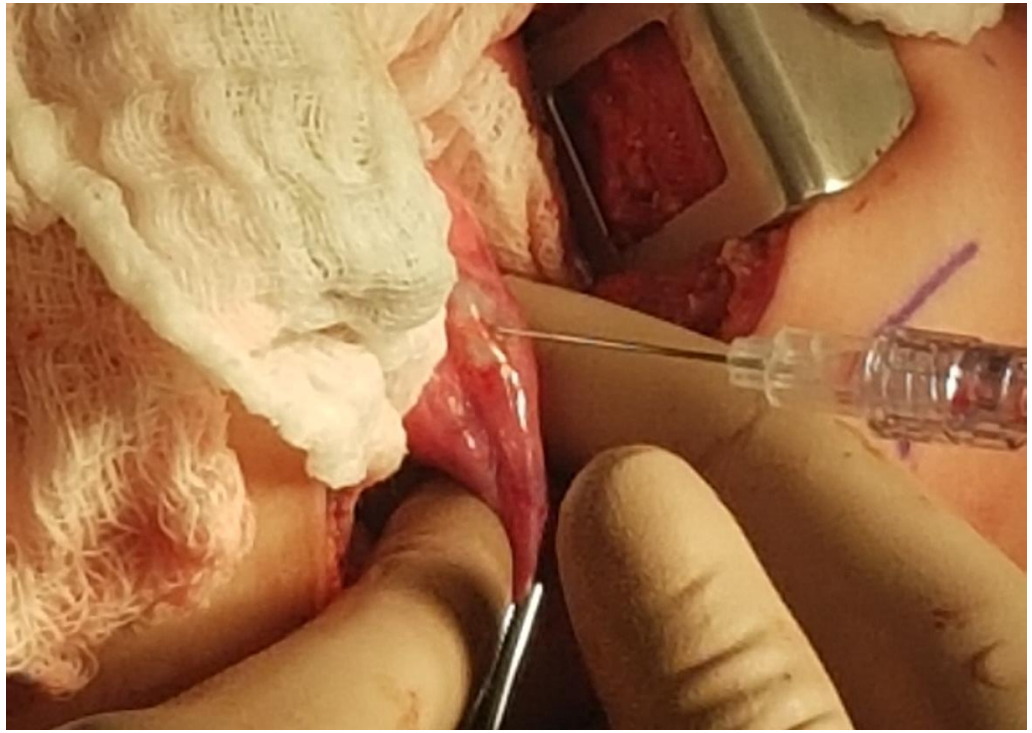
Surgical treatment

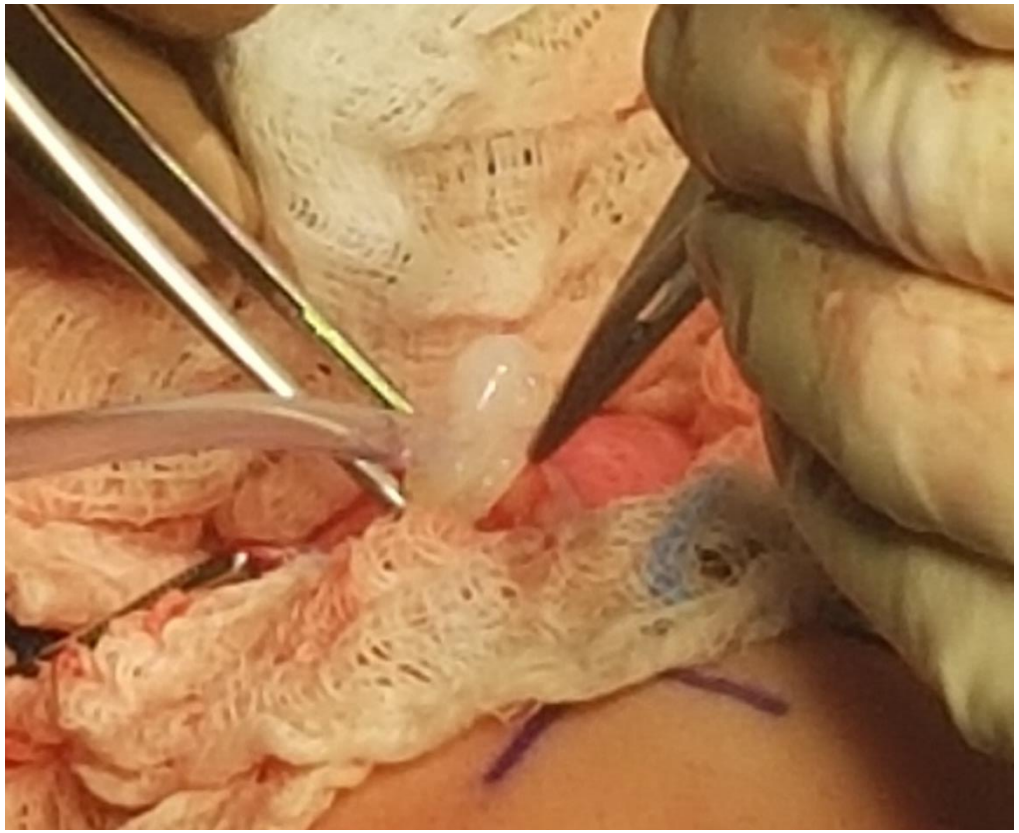
- For patients who are able to undergo surgery, it is considered the treatment of choice
- The Parasite can be completely removed and the patient cured
- The surgical options for lung cysts include:
  - Lobectomy
  - Wedge resection
  - Pericystectomy
  - Endocystectomy

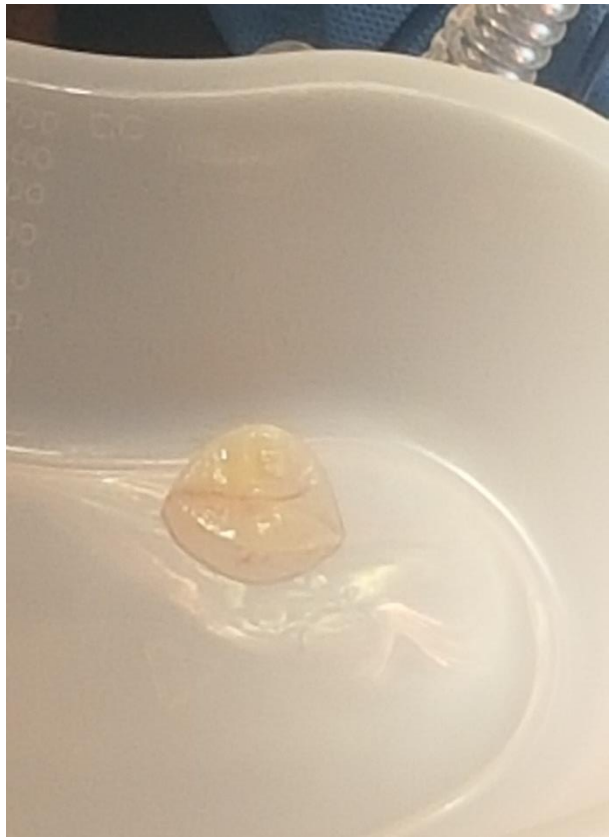
R. Morar, C. Feldman Pulmonary echinococcosis, ERS Journals 2003

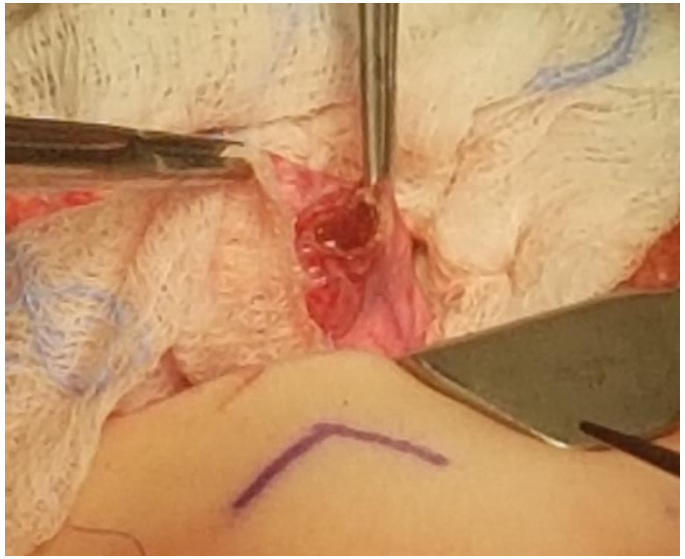
Endocystectomy











Ann Thorac Cardiovasc Surg 2017; 23: 286–290

Online September 5, 2017
doi: 10.5761/atcs.oa.17-00023

Original Article

Is Anatomic Lung Resection Necessary in Surgical Treatment of Giant Lung Hydatid Cysts in Childhood?

Omer Onal, MD and Omer Faruk Demir, MD

- Retrospective study - Kayseri, Turkey
- patients under 16 operated between 2000 and 2017 for pulmonary hydatid cysts (n=200)
- 32 patients who had giant hydatid cyst (>10cm) were included in this study
- Parenchymal saving methods (cystotomy) were preferred and
- No lung resections were applied.

Results:

- The mean age was 11.3 ± 3.2 years
- The total number of giant cysts was 32, The average size of the cysts was 11 cm, 37.5% of the cysts were perforated
- Postoperative complication rate was 31.3%
- **No recurrence and mortality were seen during follow-up period**

Table 1 Postoperative complications

Complications	Frequency (n)	Percent (%)
Atelectasis	5	15.6
Pneumothorax	2	6.3
Bronchopleural fistula	2	6.3
Pneumonia	1	3.1
Total	10	31.3

Ann Thorac Cardiovasc Surg Vol. 23, No. 6 (2017)

Conclusion:

- Considering the high recovery capacity of lung tissue, a chance should be given to recover the existing infection, atelectasis, and parenchymal damage
- Especially in areas where hydatid disease is endemic, children may be re-infected
- Lung resection is not recommended

Ann Thorac Cardiovasc Surg Vol. 23, No. 6 (2017)

Adjunctive anti parasitic therapy

- The optimal duration of chemotherapy before and after surgical procedures is not known
- Therapy generally should begin ~4 days prior to surgery and be continued for ~3 months
- Albendazole is preferred over Mebendazole because it has better bioavailability
- Some add Paraziquantel to Albendazole for better cyst penetration

R. Morar, C. Feldman Pulmonary echinococcosis, ERS Journals 2003

Take home messages:

- Echinococcus in Canada!
- In pediatrics – pulmonary > liver, rapidly growing cysts
- Many times asymptomatic
- Negative serology does not r/o hydatid cysts

Thank you!

- Dr Shaun Morris
- Dr Felix Ratjen