Cystic tumor in liver

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Outline

- Diagnosis
  - Liver cyst
  - PCLD
  - MCN
  - Hydatid disease
- Management

Clinical Practice Guidelines

JOURNAL OF HEPATOLOGY

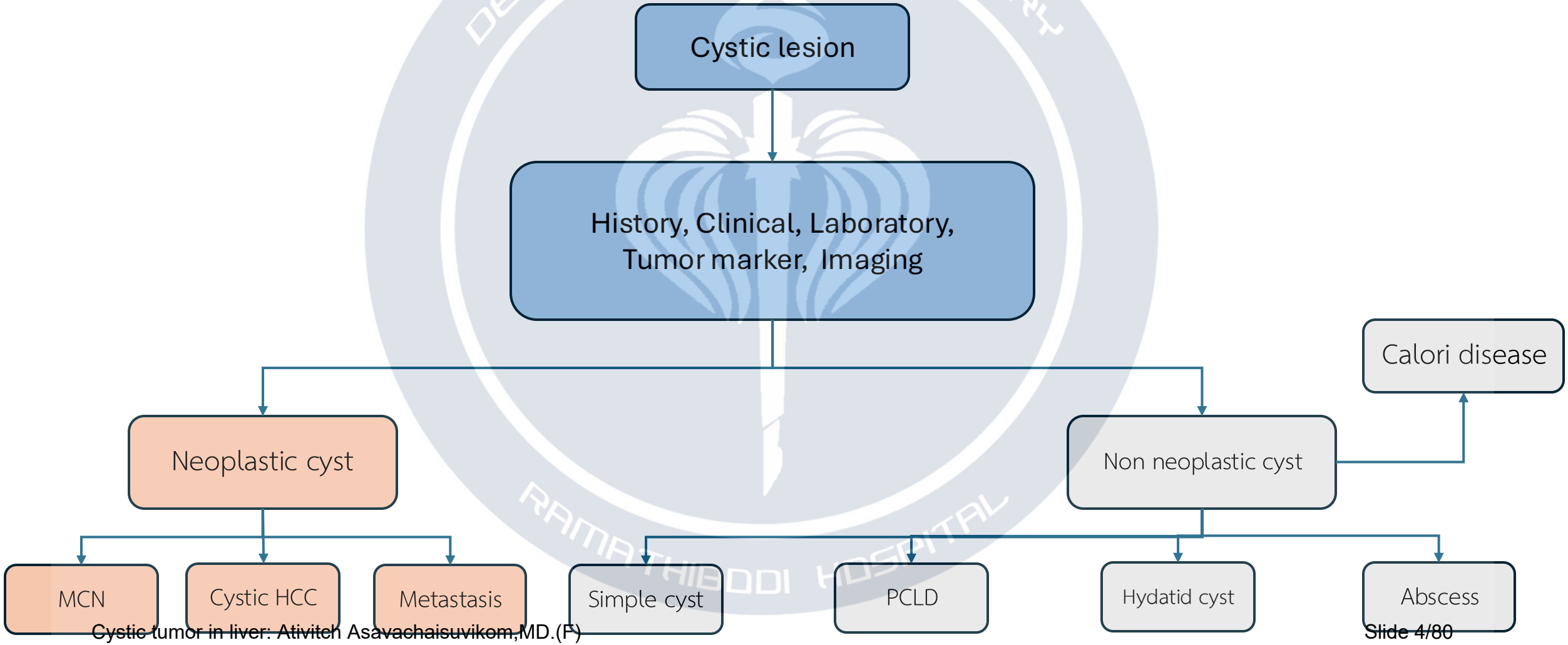
EASL Clinical Practice Guidelines on the management of cystic liver diseases☆

European Association for the Study of the Liver*

Ultrasonography should be the first imaging modality when demonstrating complex features (e.g.atypical cyst wall or content) required further evaluation using additional imaging

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

Differential diagnosis



Hepatic cyst

- Diagnosis
  - US : anechoic content with posterior enhancement
  - CT : homogeneous and hypoattenuating lesions in NC, no enhancement
  - MRI : A strong signal on T2-weighted sequences, similar to other fluids (cerebrospinal fluid) and a low T1-weighted signal
- Characteristic of cyst
  - Number (solitary vs. multiple) and architecture (simple vs. complex cyst).
  - The presence of complex features within a lesion
    - calcifications, septations, mural thickening or nodularity, debris-containing fluid, haemorrhagic or proteinaceous contents, fluid levels, wall enhancement, and associated bile duct dilatation.

Mavilia MG, Pakala T, Molina M, Wu GY. Differentiating cystic liver le-sions: a review of imaging modalities, diagnosis and management. J Clin Transl Hepatol 2018;6:208–216.

Simple liver cyst

- Ultrasound is the modality of choice to diagnose a simple hepatic cyst.
- Simple cysts are round or oval-shaped, anechoic with sharp and smooth borders with thin walls, and strong acoustic posterior enhancement
- CT and MRI are not indicated to further characterize simple hepatic cysts.

Mavilia MG, Pakala T, Molina M, Wu GY. Differentiating cystic liver lesions: a review of imaging modalities, diagnosis and management. J Clin Transl Hepatol 2018;6:208–216.

Hepatic cyst

- Worrisome feature for differentiate from MCN

Major worrisome features	Minor worrisome features
Thick septation	Upstream biliary dilatation
Nodularity	Thin septations
	Internal haemorrhage
	Perfusional change
	<3 coexistent hepatic cysts

MRI should be used to characterize hepatic cyst with worrisome features

A combination of >= 1 major and >= 1 minor feature may be consider for MCN

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Simple liver cyst

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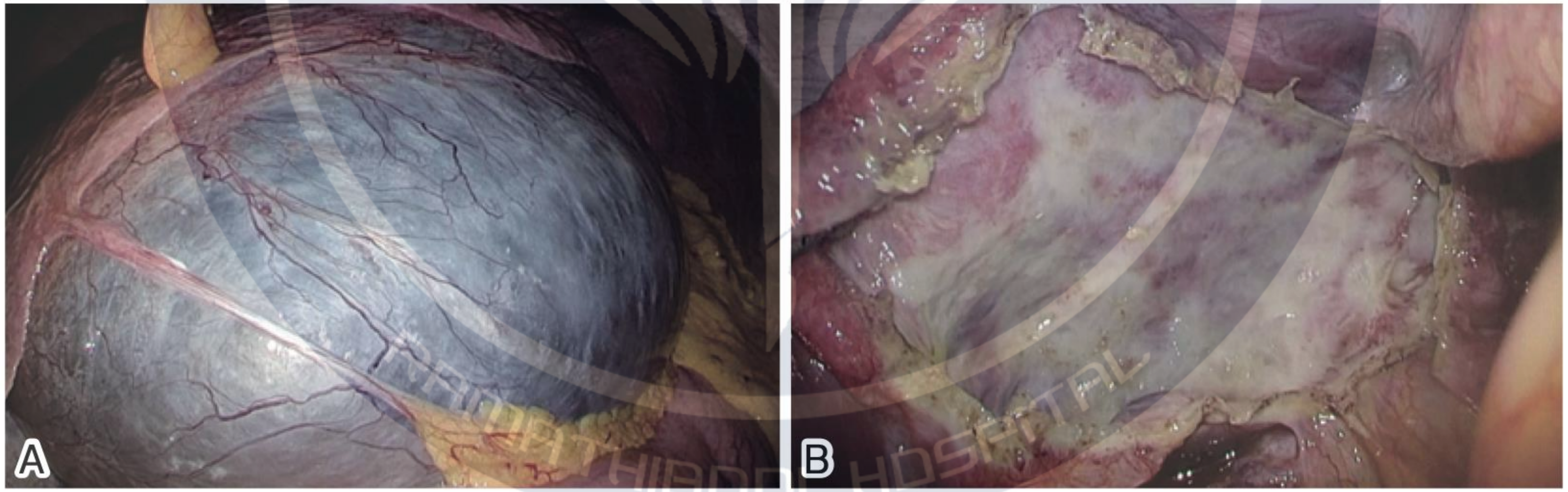
Simple liver cyst

- Management
  - Asymptomatic
    - Not required treatment or follow-up
  - Symptomatic
    - Percutaneous aspiration and sclerotherapy
      - symptoms persisting in less than 4% of patients, and complication and recurrence rates were each <1%
      - The most commonly used sclerosing agent is ethanol. However, ethanolamine oleate, polidocanol, minocycline hydrochloride, and bleomycin have also been used.
      - Maximum response at 6 months

Furumaya A, van Rosmalen BV, de Graeff JJ et al. Dutch Benign Liver Tumor Group. Systematic review on percutaneous aspiration and sclerotherapy versus surgery in symptomatic simple hepatic cysts. HPB (Oxford). 2021 Jan;23(1):11-24

Simple liver cyst

- Management
  - Symptomatic
    - Surgical options(open and laparoscopic)
      - provide long-term relief in up to 90% of patients with symptomatic hepatic cysts



Furumaya A, van Rosmalen BV, de Graeff JJ et al. Dutch Benign Liver Tumor Group. Systematic review on percutaneous aspiration and sclerotherapy versus surgery in symptomatic simple hepatic cysts. HPB (Oxford). 2021 Jan;23(1):11-24

Yoshida H, Onda M, Tajiri T, et al. Long-term results of multiple minocycline hydrochloride injections for the treatment of symptomatic solitary hepatic cyst. J Gastro- enterol Hepatol. 2003;18:595—8.

Simple liver cyst

- Management
  - Symptomatic
    - Percutaneous aspiration and sclerotherapy vs drainage
      - EASL 2022, Recommended for percutaneous aspiration and sclerotherapy and laparoscopic fenestration
      - ACG 2024, There is a lack of robust randomized controlled trials (RCTs) and long-term outcome data comparing these methods. Both modality are effectively.

REVIEW ARTICLE

Systematic review on percutaneous aspiration and sclerotherapy versus surgery in symptomatic simple hepatic cysts

Alicia Furumaya[1,*], Belle V. van Rosmalen[1,*], Jan Jaap de Graeff[1,*], Martijn P.D. Haring[2], Vincent E. de Meijer[2], Thomas M. van Gulik[1], Joanne Verheij[3], Marc G. Besselink[1], Otto M. van Delden[4,**], Joris I. Erdmann[1**] on behalf of the Dutch Benign Liver Tumor Group

[1]Department of Surgery, Amsterdam Gastroenterology Endocrinology Metabolism, Amsterdam UMC, University of Amsterdam, [2]Department of Surgery, University Medical Center Groningen, University of Groningen, [3]Department of Pathology, and [4]Department of Interventional Radiology, Amsterdam UMC, University of Amsterdam, Netherlands

In total, 736 patients from 34 studies

- 265 (36%) underwent PAS
- 348 (47%) laparoscopic cyst deroofing
- 123 (17%) open surgical management.

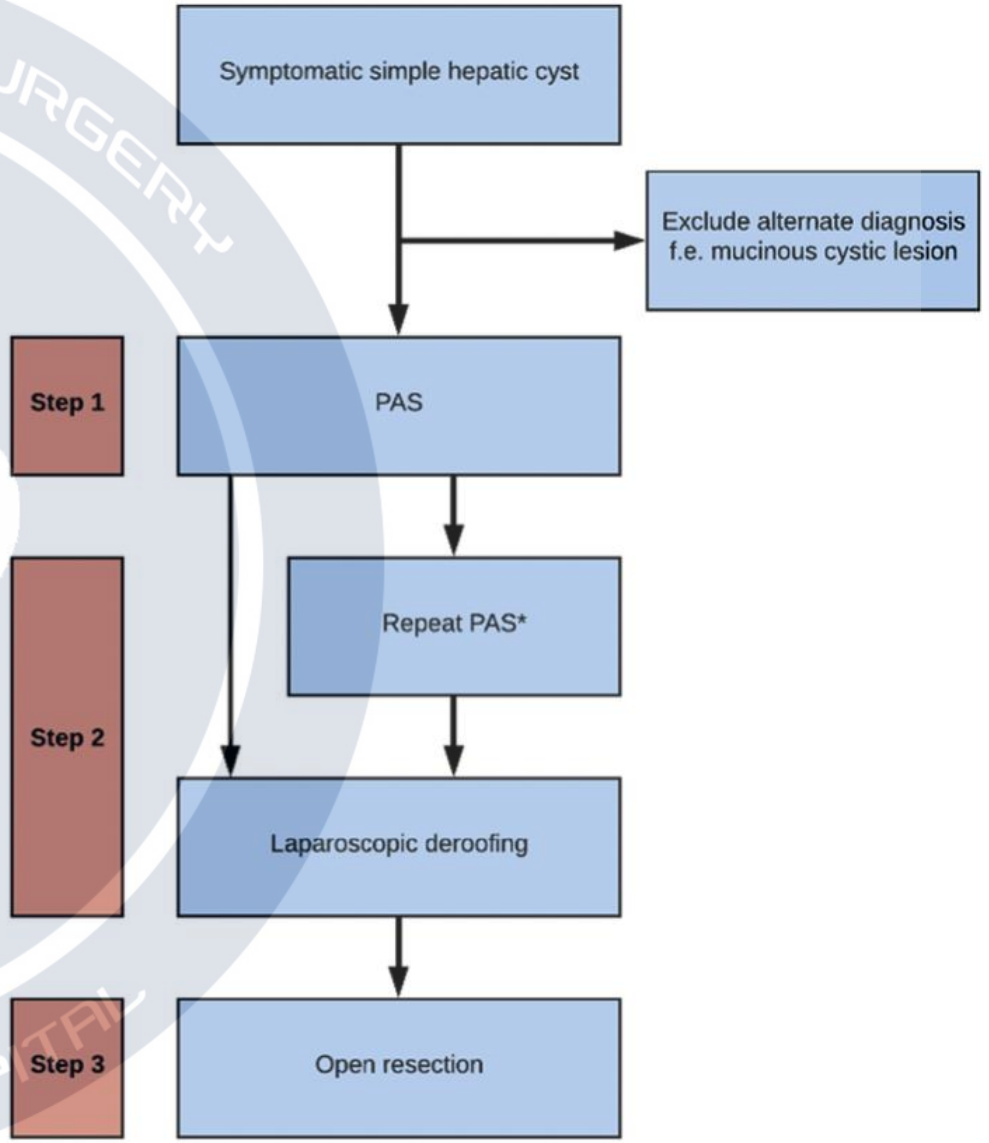
During weighted mean follow-up of 26.1, 38.2 and 21.3 months

Symptoms persisted in

- 3.5% for PAS
- 2.1% for laparoscopic cyst deroofing
- 4.2% for open

Major complication rates were 0.8%, 1.7%, and 2.4%, respectively.

Cyst recurrence rates were 0.0%, 5.6%, and 7.7%, respectively.



Alicia Furumaya, Belle V. van Rosmalen, Jan Jaap de Graeff et al. Systematic review on percutaneous aspiration and sclerotherapy versus surgery in symptomatic simple hepatic cysts, HPB, Volume 23, Issue 1, 2021

Complicated liver cyst

- Complication of liver cyst
  - Infection
    - Incidence 3-5%
  - Haemorrhage
    - Incidence unknown

Lantinga MA, Geudens A, Gevers TJ, Drenth JP. Systematic review: the management of hepatic cyst infection., *Aliment Pharmacol Ther* 2015; **41:** 253–61.

Renée Duijzer, Lucas H P Bernts, Anja Geerts, Clinical management of liver cyst infections: an international, modified Delphi-based clinical decision framework,. The Lancet Gastroenterology & Hepatology,, Volume 9, Issue 9, 2024,

Complicated liver cyst

Table 3. Criteria for hepatic cyst infection and radiological findings suggestive of hepatic cyst infection.

Criteria definite hepatic cyst infection	Criteria likely hepatic cyst infection (after exclusion of other sources)
• Cyst aspiration showing evidence of infection (neutrophil debris and/or microorganism)	• Fever (temperature >38.5°C for >3 days) with no other source of fever detectable • CT or MRI detecting gas in a cyst • ^{18}FDG PET-CT showing increased FDG activity lining a cyst compared to normal parenchyma • Tenderness in the liver area • Increased C-reactive protein • Increased leukocyte count (>11,000/L) • Positive blood culture
Radiological findings suggestive of hepatic cyst infection	
• Liver ultrasound: debris with a thick wall and/or a distal acoustic enhancement in at least one cyst • Liver CT/MRI: enhanced wall thickening and/or perilesional inflammation in at least one cyst • MRI: high signal intensity on diffusion-weighted images, fluid-fluid level, wall thickening, or gas in at least one cyst • Positron emission tomography scan (^{18}FDG PET-CT): increased FDG activity lining a cyst compared to normal parenchyma	

^{18}FDG PET-CT, 18fluorodeoxyglucose positron emission tomography-CT.

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

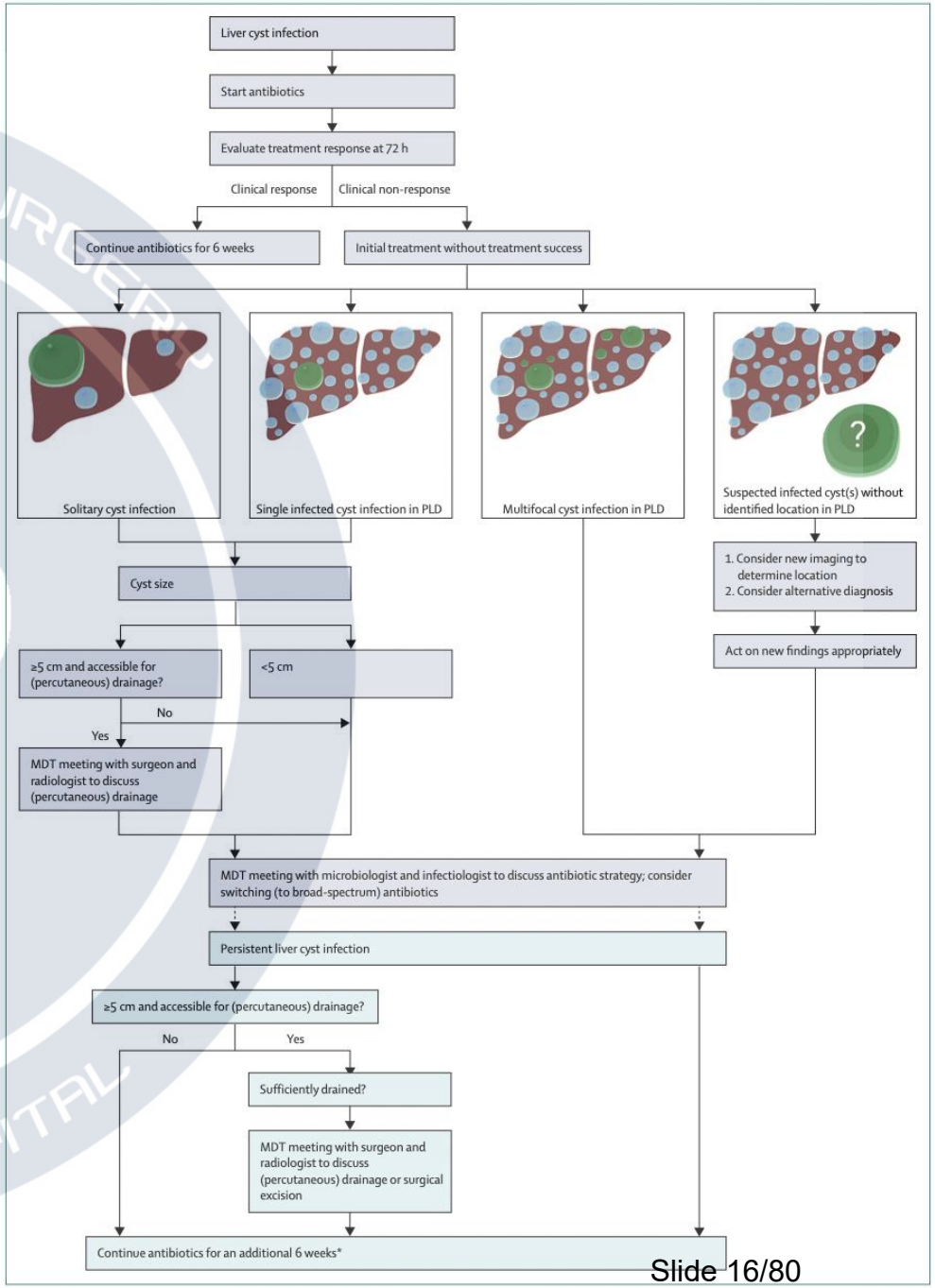
Complicated liver cyst

- Management
  - Antibiotic therapy
    - Fluoroquinolones and third-generation cephalosporins are recommended as empirical first-line antibiotics for hepatic cyst infection
    - Duration 4-6 weeks
  - Secondary prophylaxis for hepatic cyst infection is not recommended
  - Role for drainage
    - Persistence of temperature >38.5° C after 48 hours on empirical antibiotic therapy
    - Isolation of pathogens unresponsive to antibiotic therapy from a cyst aspirate
    - Severely compromised immune system
    - CT or MRI detecting gas in a cyst
    - Large infected hepatic cysts(>= 5 cm)

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

Clinical management of liver cyst infections: an international, modified Delphi-based clinical decision framework

Renée Duijzer, Lucas H P Bernts, Anja Geerts, Bart van Hoek, Minneke J Coenraad, Chantal Rovers, Domenico Alvaro, Ed J Kuijper, Frederik Nevens, Jan Halbritter, Jordi Colmenero, Juozas Kupcinskas, Mahdi Salih, Marie C Hogan, Maxime Ronot, Valerie Vilgrain, Nicolien M Hanemaaijer, Patrick S Kamath, Pavel Strnad, Richard Taubert, Ron T Gansevoort, Roser Torra, Silvio Nadalin, Tatsuya Suwabe, Tom J G Gevers, Vincenzo Cardinale, Joost P H Drenth, Marten A Lantinga



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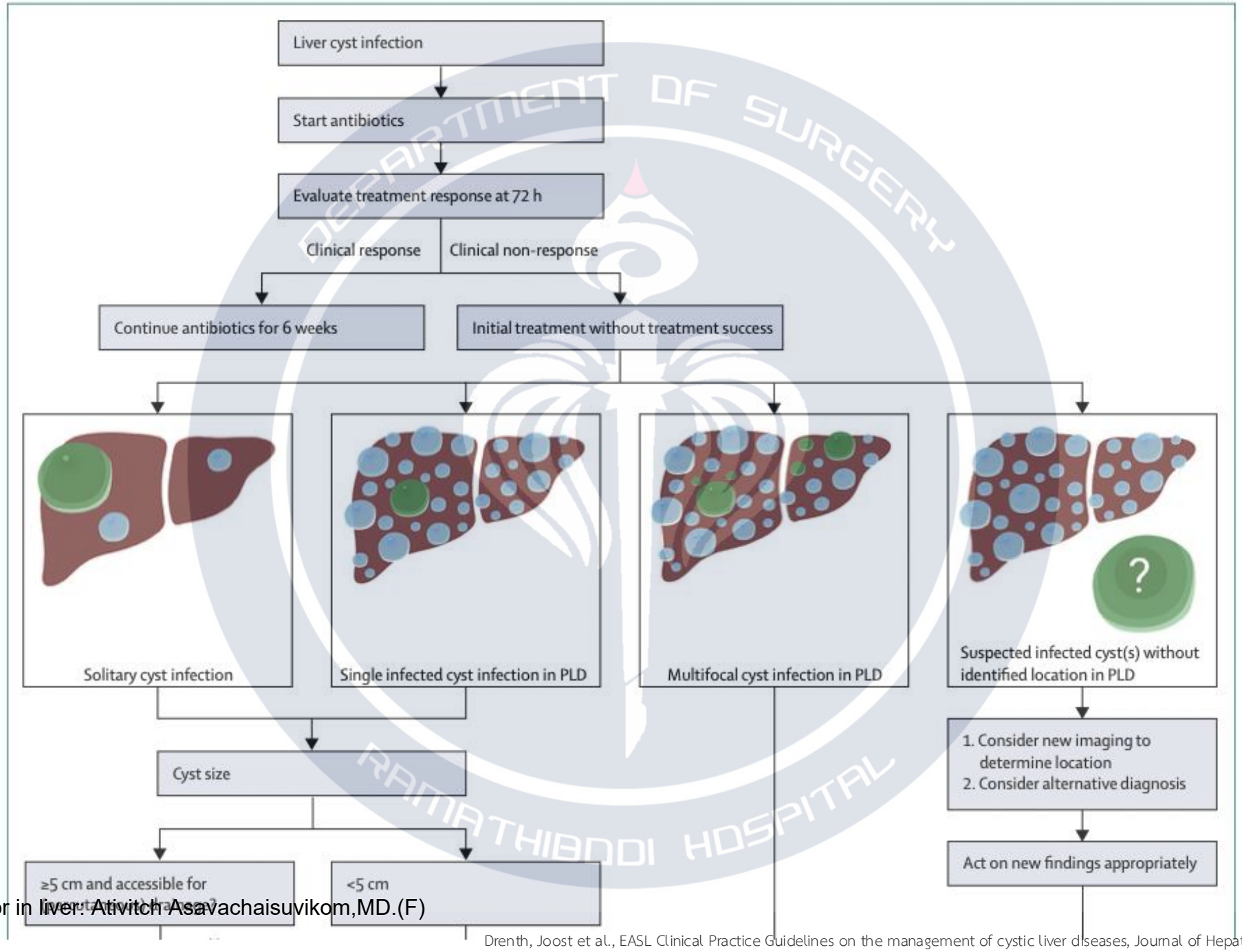

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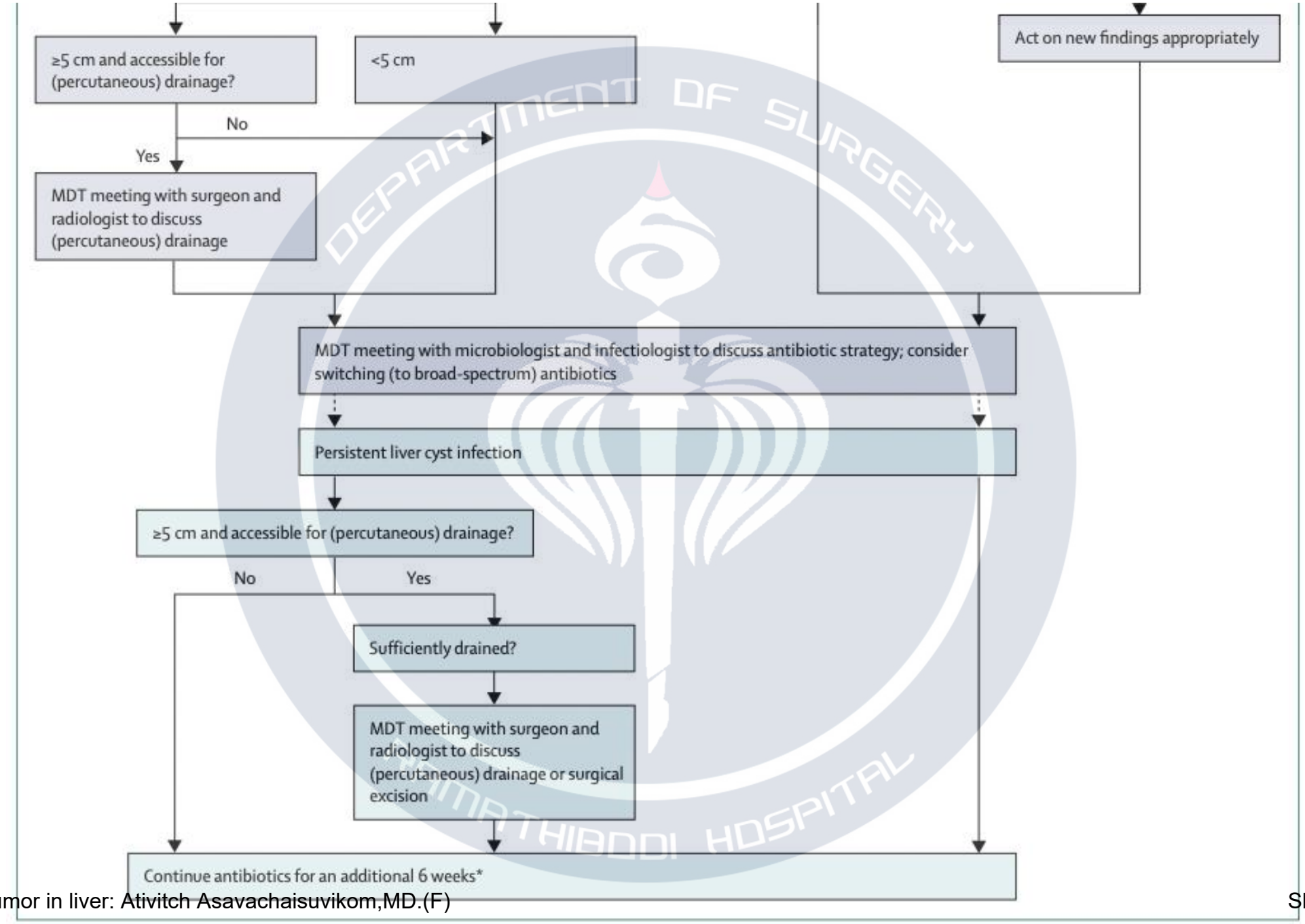

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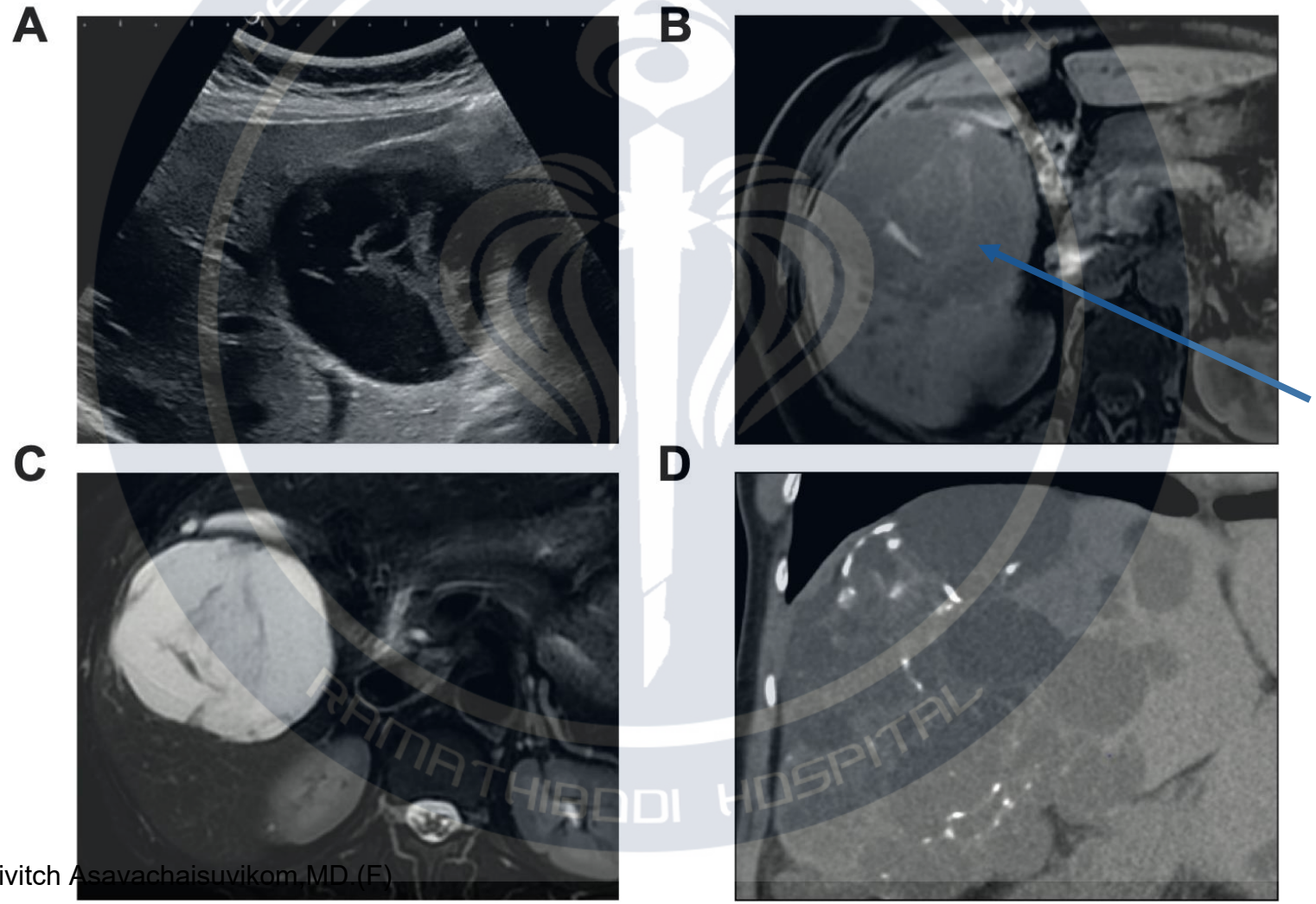
Complicated liver cyst

- Complication of liver cyst
  - Haemorrhage
    - Clinical : sudden and severe abdominal pain, Hct drop
    - Diagnostic : US or MRI
    - Management :
      - Conservative is preferred
      - Avoid to aspirate, deroof in active hemorrhage

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

Hemorrhage liver cyst



haemorrhagic septation

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

Sclerotherapy

- Aims to destroy the epithelium lining the inner surface of the wall to stop intracystic fluid secretion
- The most frequently used sclerosing agent is 95% ethanol(beware alcohol intoxication)
- Alternative : Minocycline hydrochloride, ethanolamine oleate and hypertonic saline and bleomycin40 have been proposed as alternatives.
- Contraindication : connect with biliary tract
- Complication : painful, Transient neuropsychic disorders, Intracystic bleeding(rare)

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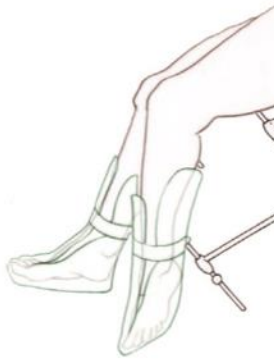
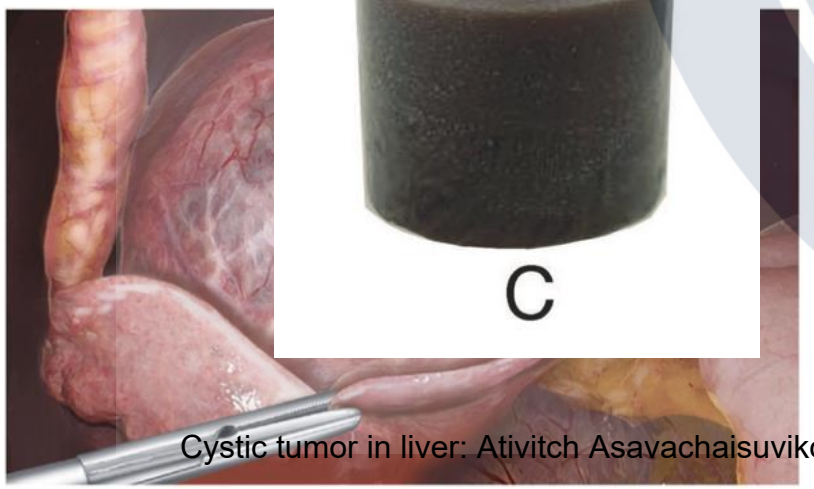
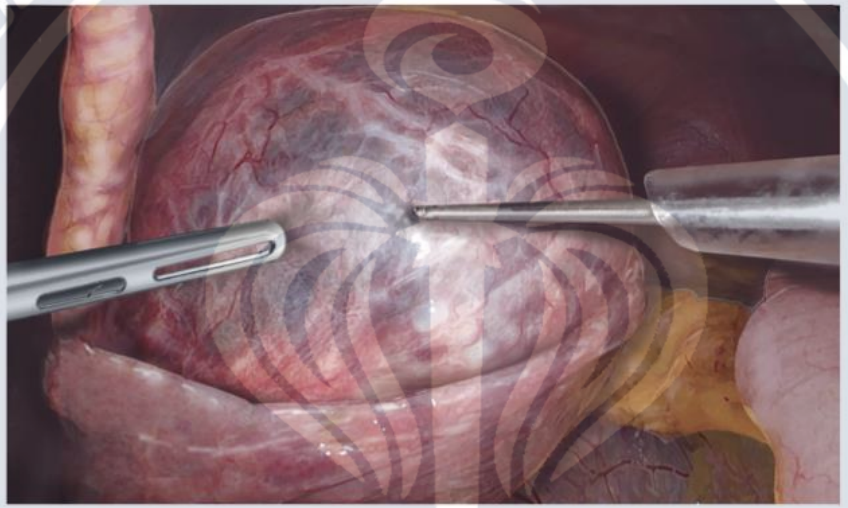
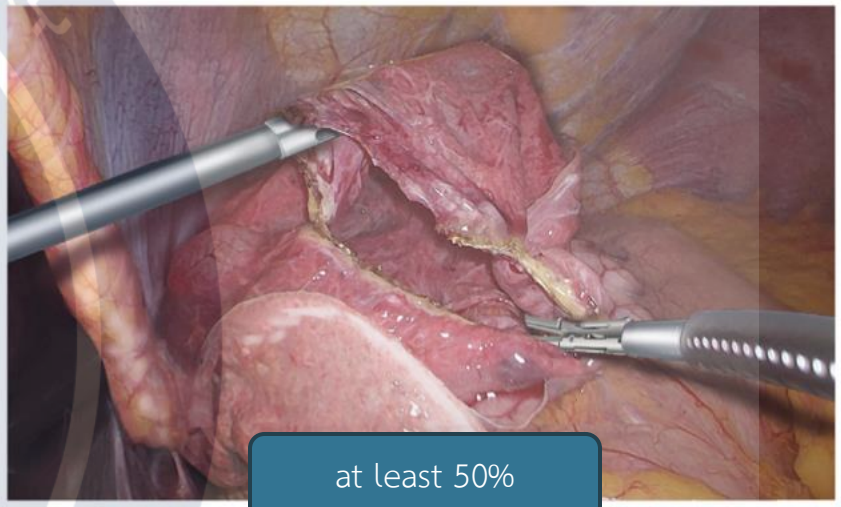
Laparoscopic deroof cyst

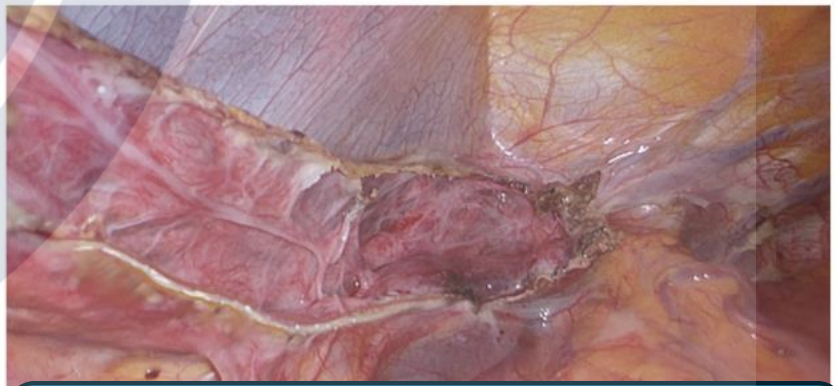








at least 50%



For internal epithelial : electrocoagulated or destroyed either using bipolar forceps or an argon laser

Cystic tumor in liver: Ativitch Asavachaisuvikom,MD.(F)

Marichez A, Fernandez B, Adam JP, Chiche L. Laparoscopic cyst fenestration for simple hepatic cyst. J Visc Surg. 2022 D 82.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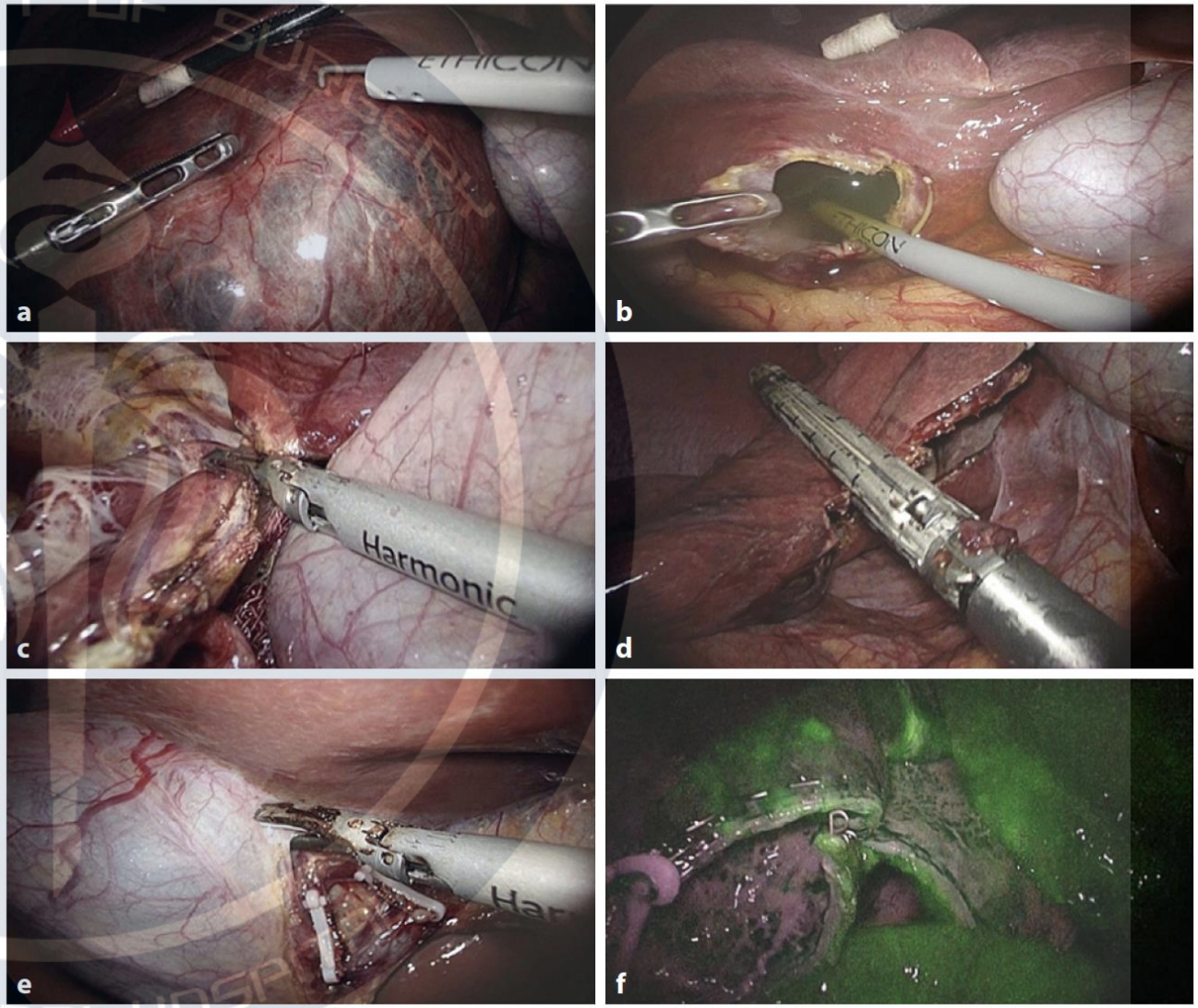
Eur Surg (2023) 55:100–106
https://doi.org/10.1007/s10353-023-00798-8

european surgery
ACA Acta Chirurgica Austriaca

Laparoscopic deroofing of simple liver cysts: do ancillary techniques, surgical devices, and indocyanine green improve outcomes?

Alessia Kersik · Luca Galassi · Giulia Colombo · Luigi Bonavina

Received: 8 March 2023 / Accepted: 15 March 2023 / Published online: 18 April 2023



Ancillary techniques

omentopexy (n= 8),

argon plasma coagulation (n=4),

ethanol sclerotherapy (n=4).

No clear consensus

Kersik, A., Galassi, L., Colombo, G. *et al.* Laparoscopic deroofing of simple liver cysts: do ancillary techniques, surgical devices, and indocyanine green improve outcomes?. *Eur Surg* **55**, 100–106 (2023). https://doi.org/10.1007/s10353-023-00798-8

Polycystic liver disease

- Related genetic
  - ADPKD Related :
    - PKD1 or PKD2 gene mutation
    - Incidence 1 in every 800 live birth
  - Isolated PCLD :
    - heterozygous mutation in either the protein kinase C substrate 80K-H(PRKCSH), SEC63, LRP5 or, very rarely, GANAB genes
    - Incidence less than 0.01%

Olivier Farges and Béatrice Aussilhou, Simple cysts and polycystic liver disease : Clinical and radiographic features, surgical and nonsurgical management. Blumgart's Surgery of the Liver, Biliary Tract and Pancreas, Chapter 75, 1122-1147.e3,

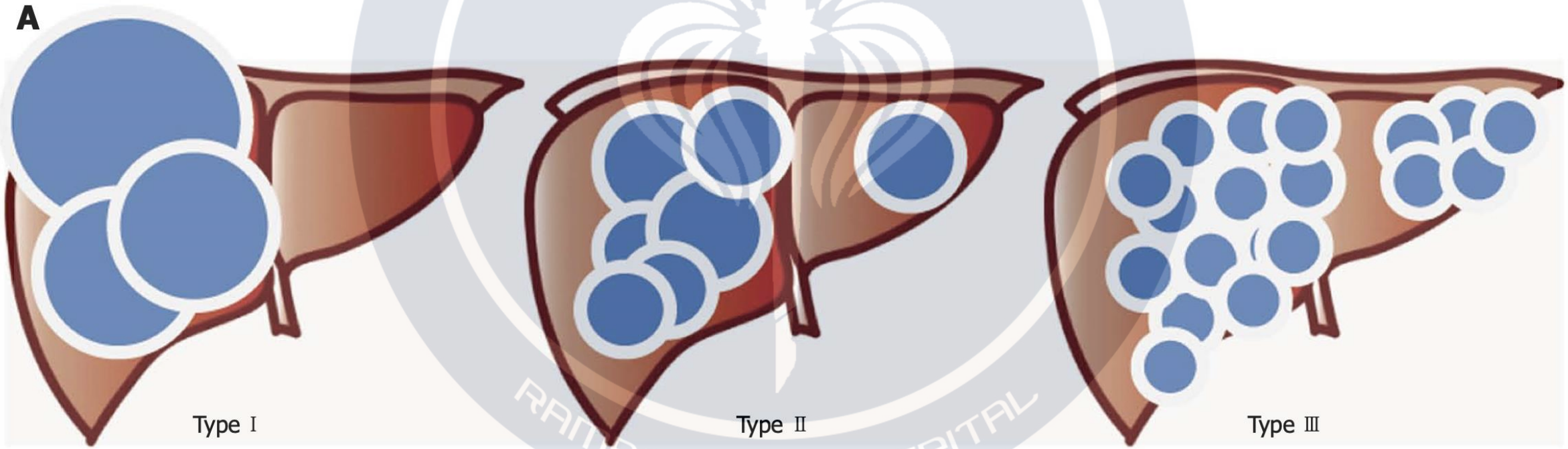
Polycystic liver disease

- Diagnosis
  - No previous family history
    - More than 15 to 20 cysts
  - History PCLD or ADPKD
    - 4 cysts

Olivier Farges and Béatrice Aussilhou, Simple cysts and polycystic liver disease : Clinical and radiographic features, surgical and nonsurgical management. Blumgart's Surgery of the Liver, Biliary Tract and Pancreas, Chapter 75, 1122-1147.e3,

Polycystic liver disease

Gigot's classification



Number : less than 10 large hepatic cysts
Size : measuring more than 10 cm

Number : diffuse involvement of liver parenchyma
Remaining large normal liver parenchyma

Small and medium size
with only few area of normal liver parenchyma

B. Aussilhou, S. Dokmak, F. Donderoet al., Treatment of polycystic liver disease. Update on the management, Journal of Visceral Surgery, Volume 155, Issue 6, 2018

Polycystic liver disease

Qian's classification

Grade	Number of cysts	Hepatomegaly
1	0	Asymptomatic
2	1 to 10	Asymptomatic
3	11 to 20	Asymptomatic
4	>20	Symptomatic

Jung, Donghwan Hwang, Shin Ahn et al., Fenestration of liver cysts in polycystic liver disease to improve quality of life: a case report and literature review. Korean Journal of Hepato-Biliary-Pancreatic Surgery.

Polycystic liver disease

Schnelldorfer's classification

Table 1 Schnelldorfer lesional classification of polycystic liver disease [44].

	Type A	Type B	Type C	Type D
Symptoms	Absent or moderate	Moderate or severe	Severe	Severe
Size and number of cysts	Few	Few but large in size	Few and small in size	Few to numerous
Number of spared liver segments	> 3 segments	≥ 2 segments	≥ 1 segment	< 1 segment
Presence of collateral venous circulation in the spared segment	Moderate	Absent	Absent	Present
Recommended treatment	Therapeutic abstention or medical treatment	Fenestration	Partial hepatectomy with fenestration of contralateral cysts	Hepatic transplantation

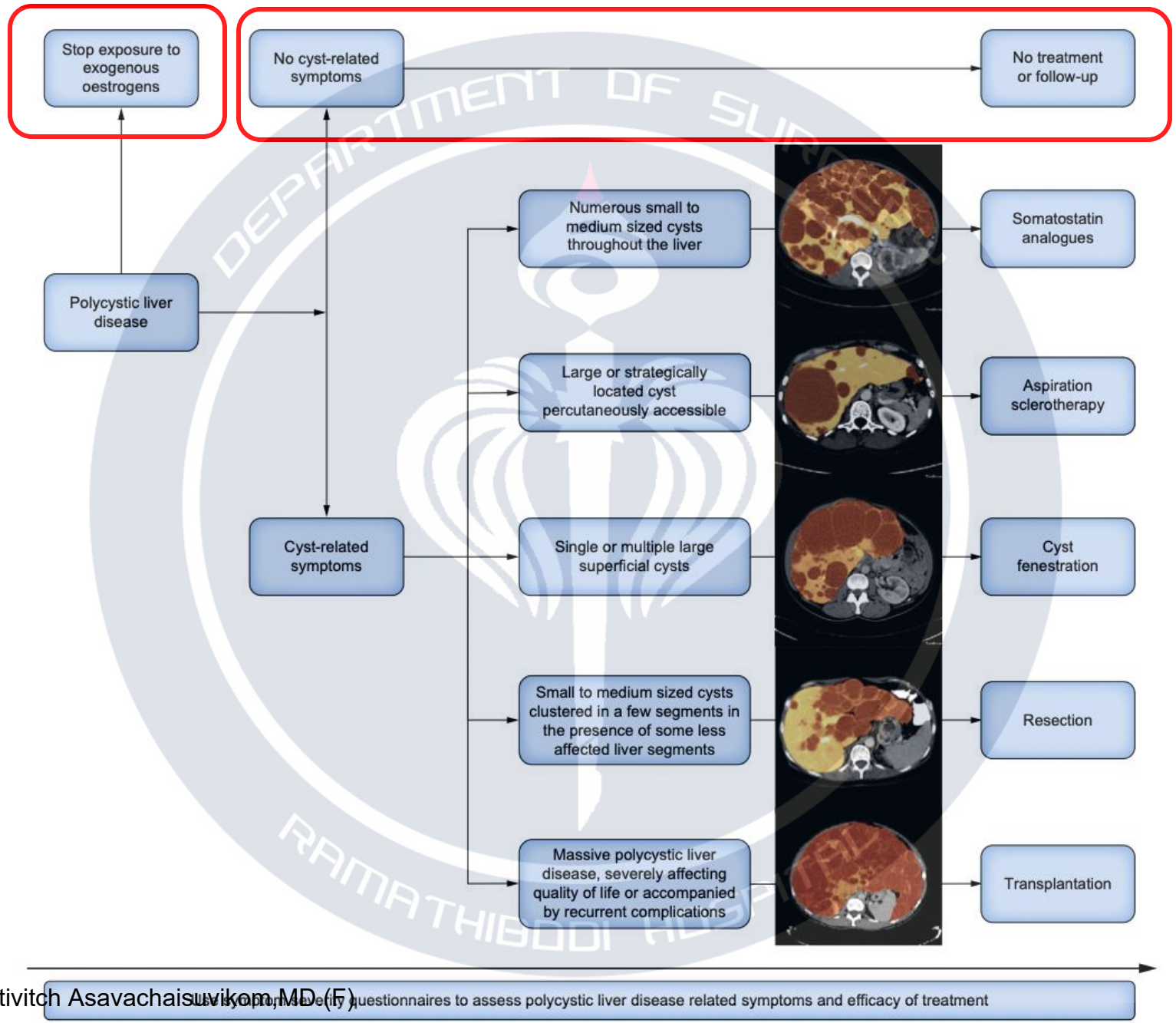
B. Aussilhou, S. Dokmak, F. Donderoet al., Treatment of polycystic liver disease. Update on the management, Journal of Visceral Surgery, Volume 155, Issue 6, 2018

Polycystic liver disease

Table 5. Polycystic liver disease-related symptoms and complications.

Polycystic liver disease-related symptoms	Polycystic liver disease-related complications
Abdominal fullness	Jaundice
Lack of appetite or early satiety	Hepatic venous outflow obstruction
Acid reflux	Portal hypertension
Nausea and vomiting	Recurrent cyst infection
Pain in rib cage, sides, abdomen or back	Recurrent cyst haemorrhage
Shortness of breath	
Limited mobility	
Fatigue	
Anxiety about the future	
Concern or dissatisfaction with abdomen size	
Problems with intercourse	
Involuntary weight loss	

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108



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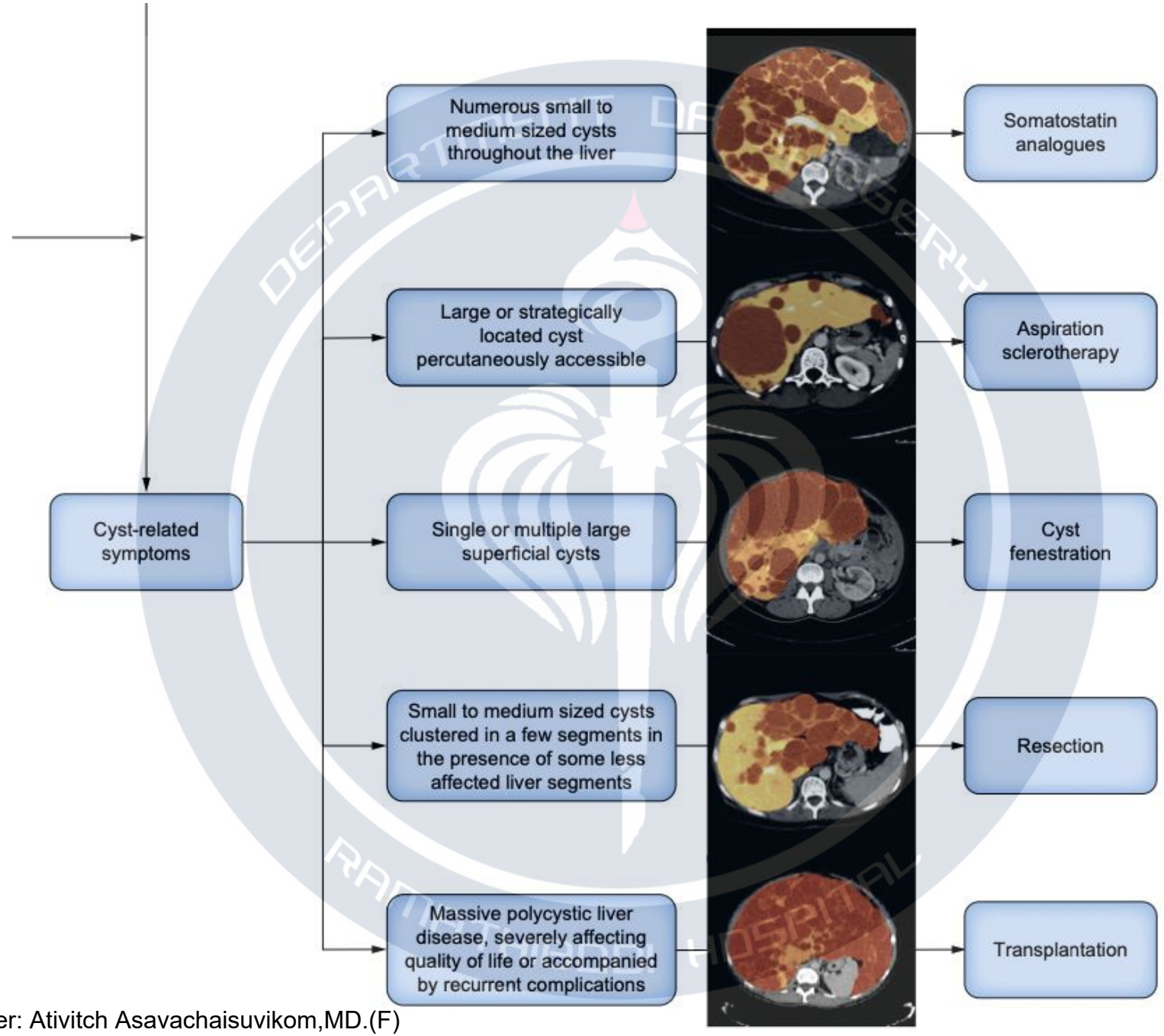


Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

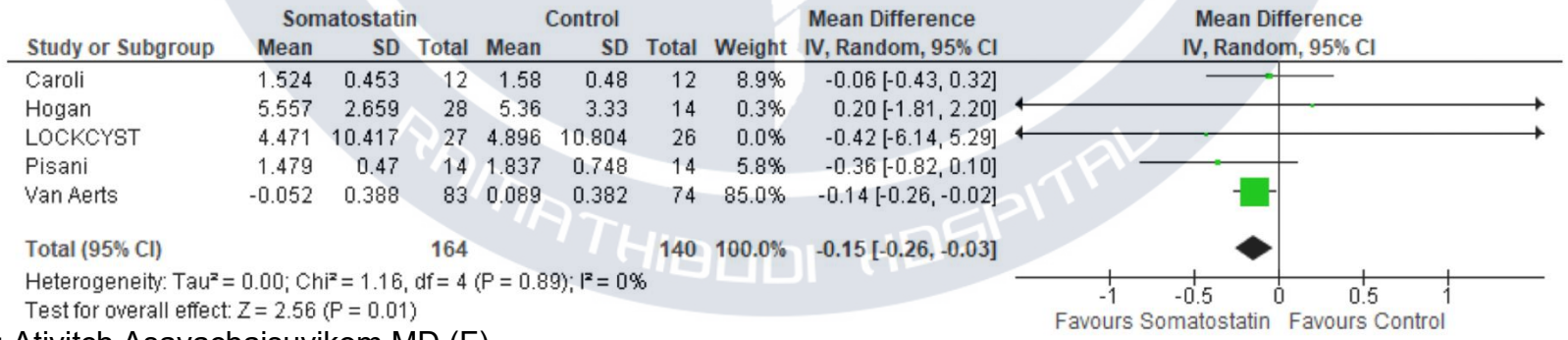
Polycystic liver disease

- Management and evaluation
  - Nutrition support
    - CT is suggested to assess sarcopenia in patients
      - Sarcopenia ; skeletal muscle index <38.5 cm2/h2 in females and <52.4 cm2/h2 in males
    - Intensive nutrition care and rehabilitation
  - Avoidance of estrogen replacement therapy

Griffiths J, Mills MT, Ong AC. Long-acting somatostatin analogue treatments in autosomal dominant polycystic kidney disease and polycystic liver disease: a systematic review and meta-analysis. BMJ Open. 2020 Jan 9;10(1):e032620.

Polycystic liver disease

- Management
  - Nonsurgical treatment
    - Medication
      - Somatostatin analogue :
        - Block cAMP > decrease secretion from cyst wall
        - Dose : octreotide-long acting release 20 mg IM bid, Lanreotide
        - Decease total liver volume but ADPKA still same



Study or Subgroup	Somatostatin Mean	SD	Total	Control Mean	SD	Total	Weight	Mean Difference IV, Random, 95% CI
Caroli	1.524	0.453	12	1.58	0.48	12	8.9%	-0.06 [-0.43, 0.32]
Hogan	5.557	2.659	28	5.36	3.33	14	0.3%	0.20 [-1.81, 2.20]
LOCKCYST	4.471	10.417	27	4.896	10.804	26	0.0%	-0.42 [-6.14, 5.29]
Pisani	1.479	0.47	14	1.837	0.748	14	5.8%	-0.36 [-0.82, 0.10]
Van Aerts	-0.052	0.388	83	0.089	0.382	74	85.0%	-0.14 [-0.26, -0.02]
Total (95% CI)			**164**			**140**	**100.0%**	**-0.15 [-0.26, -0.03]**

Heterogeneity: Tau² = 0.00; Chi² = 1.16, df = 4 (P = 0.89); I² = 0%
Test for overall effect: Z = 2.56 (P = 0.01)

Griffiths J, Mills MT, Ong AC. Long-acting somatostatin analogue treatments in autosomal dominant polycystic kidney disease and polycystic liver disease: a systematic review and meta-analysis. BMJ Open. 2020 Jan 9;10(1):e032620.

Table 1 Summary of included study characteristics with regards to study details, patient population details, intervention, relevant outcomes, interventions and baseline characteristics

Study	Study design	Country	Follow-up period (months)	Number of patients (males)	Renal function criteria	Imaging modality	Intervention	Baseline characteristics mean (±SD)	Relevant outcomes
Ruggenenti *et al*[28]	Randomised, crossover, placebo-controlled trial	Italy	6	14 (9)	Serum creatinine <3.0 mg/dL, but >1.2 mg/dL (males) or >1.0 mg/dL (females)	CT	Octreotide-LAR, 40 mg intramuscularly every 28 days	eGFR ► (I) 59.5±25.2 ► (C) 57.9±22.4 TKV ► (I) 2551±1053 ► (C) 2461±959	TKV, eGFR, blood pressure, blood glucose and adverse effects
van Keimpema *et al*[24] (LOCKCYST)	Randomised, double-blind, placebo-controlled trial	The Netherlands	6	54 (7)	No eGFR restrictions, Haemodialysis (HD) patients excluded	CT	Lanreotide-LAR, 120 mg intramuscularly every 28 days	eGFR ► (I) 83±198 ► (C) 91±282 TKV ► (I) 1000±1846 ► (C) 1115±3727	TKV, TLV and adverse effects
Caroli *et al*[31]	Secondary analysis of Ruggenenti *et al*[28]	Italy	6	14 (9)	Serum creatinine <3.0 mg/dL, but >1.2 mg/dL (males) or >1.0 mg/dL (females)	CT	Octreotide-LAR, 40 mg intramuscularly every 28 days	TLV ► (I) 1595±478 ► (C) 1580±487	TLV
Hogan *et al*[23]	Randomised, double-blind, placebo-controlled trial	USA	12	42 (6)	Patients with a serum creatinine concentration >3 mg/dL or dialysis-dependant excluded	CT	Octreotide-LAR, 40 mg intramuscularly every 28 days	eGFR ► (I) 70±27 ► (C) 71±27 TKV ► (I) 1142.9±826 ► (C) 803.0±269 TLV ► (I) 5907.7±2915 ► (C) 5373.9±3565	TKV, TLV, adverse effects, eGFR, blood glucose and blood pressure
Caroli *et al*[25] (ALADIN)	Randomised, single-blind placebo-controlled trial	Italy	36	79 (37)	eGFR of 40 mL/min per 1.73 m² or higher	MRI	Octreotide-LAR, 40 mg intramuscularly every 28 days	eGFR ► (I) 90±37 ► (C) 76.1±40 TKV ► (I) 1556±1035 ► (C) 2161±1274	TKV, eGFR, adverse effects, blood glucose and blood pressure
Pisani *et al*[30]	Secondary analysis of ALADIN	Italy	36	27 (10)	eGFR of 40 mL/min per 1.73 m² or higher	MRI	Octreotide-LAR, 40 mg intramuscularly every 28 days	TLV ► (I) 1609±501 ► (C) 1693±470	TLV

Griffiths J, Mills MT, Ong AC. Long-acting somatostatin analogue treatments in autosomal dominant polycystic kidney disease and polycystic liver disease: a systematic review and meta-analysis. BMJ Open. 2020 Jan 9;10(1):e032620.

Table 1 Continued

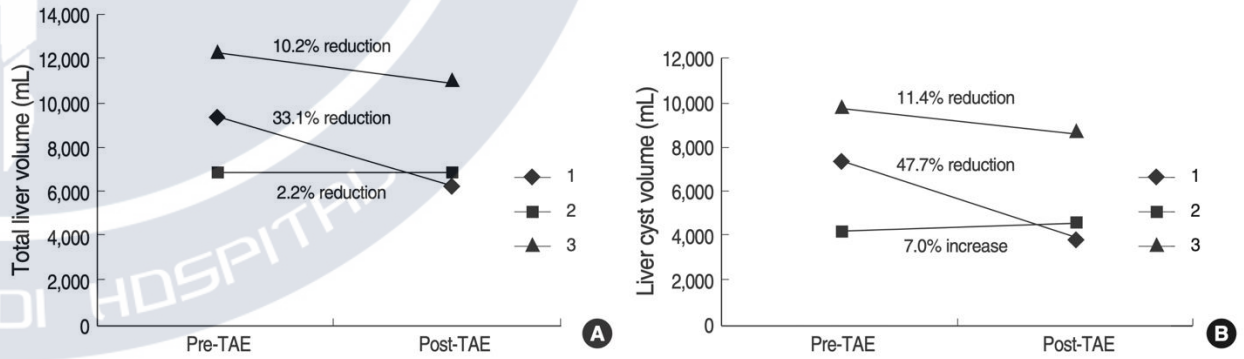
Study	Study design	Country	Follow-up period (months)	Number of patients (males)	Renal function criteria	Imaging modality	Intervention	Baseline characteristics mean (±SD)	Relevant outcomes
Meijer *et al*[26] (DIPAK-1)	Randomised, open-label, 'standard-of-care' controlled trial	The Netherlands	30	309 (142)	eGFR of 30–60 mL/min/1.73 m^2	MRI	Lanreotide-LAR, 120 mg intramuscularly every 28 days	eGFR ► (I) 51.0±11.5 ► (C) 51.4±11.2 TKV ► (I) 2046±1171 ► (C) 1874±1202	TKV, eGFR, blood pressure, blood glucose and adverse effects
Perico *et al*[27] (ALADIN 2)	Randomised, double-blind, placebo-controlled trial	Italy	36	100 (57)	eGFR between 15 and 40 mL/min/1.73 m^2	CT	Octreotide-LAR, 40 mg intramuscularly every 28 days	eGFR ► (I) 27.9±10.15 ► (C) 25.8±6.44 TKV ► (I) 2,338±1362 ► (C) 2,591±1876	TKV, eGFR, progression to ESRF, adverse events, blood glucose and blood pressure
van Aerts *et al*[29]	Secondary analysis of DIPAK-1	The Netherlands	30	175 (74)	eGFR of 30–60 mL/min/1.73 m^2	MRI	Lanreotide-LAR, 120 mg intramuscularly every 28 days	TKV ► (I) 1528±883 ► (C) 1376±347	TLV

(C), control; eGFR, estimated glomerular filtration rate; (I), intervention; LAR, long-acting release;TKV, total kidney volume; TLV, total liver volume.

Griffiths J, Mills MT, Ong AC. Long-acting somatostatin analogue treatments in autosomal dominant polycystic kidney disease and polycystic liver disease: a systematic review and meta-analysis. BMJ Open. 2020 Jan 9;10(1):e032620.

Polycystic liver disease

- Management
  - Nonsurgical treatment
    - Medication
      - Everolimus and mTOR inhibitors
        - it showed a decrease in liver volume of 3.5% in the octreotide group alone compared with a 3.8% decrease in the octreotide plus everolimus group
    - ARTERIAL EMBOLIZATION
      - Case series in Japan, Korea
      - Need further study



Chrispijn M, Gevers TJ, Hol JC, et al. Everolimus does not further reduce polycystic liver volume when added to long-acting octreotide: results from a randomized controlled trial. J Hep- atol 2013;59(1):153—9.

Park HC, Kim CW, Ro H, et al. Transcatheter arterial embolization therapy for a massive polycystic liver in autosomal dominant polycystic kidney disease patients. J Korean Med Sci 2009;24:57—61.

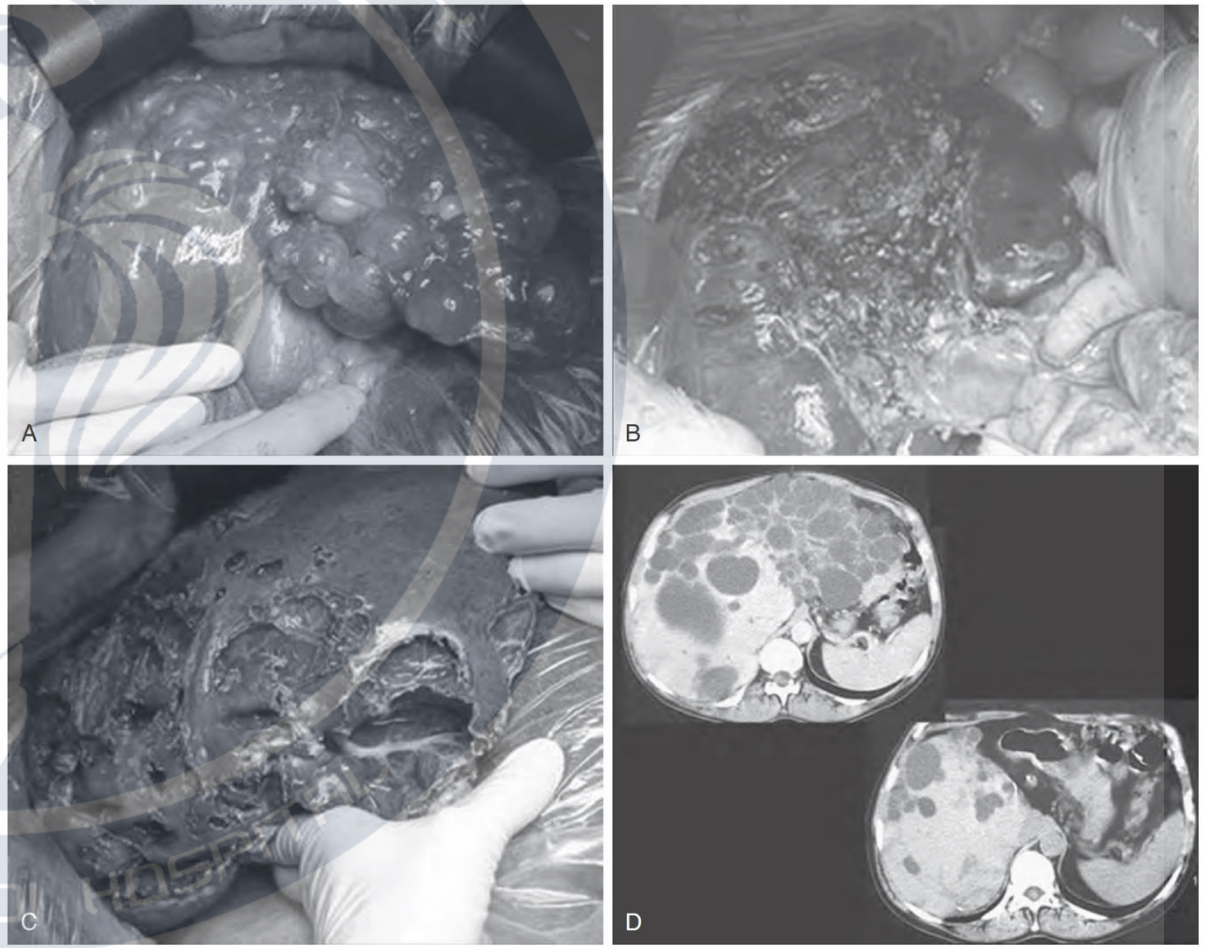
Polycystic liver disease

- Management
  - Fenestration
    - the aim of fenestration is to unroof as many cysts as possible, starting with superficial cysts and proceeding stepwise to the deeper cysts
    - In PCLD
      - Likely to have symptomatic recurrence and reintervention(33.7% vs. 9.6% and 26.4% vs. 7.1%, respectively)
      - Complications were also more frequent in PCLD patients (29.3% vs. 10.8%)
      - Serious complication : Bleeding from hepatic vein, Biliary tract injury
      - Intraoperative cholangiogram should be performed when suspect bile duct injury

Bernts LHP, Echternach SG, Kievit W, Rosman C, Drenth JPH. Clinical response after laparoscopic fenestration of symptomatic hepatic cysts: a systematic review and meta-analysis. Surg Endosc. 2019;33(3):691-704.

Polycystic liver disease

- Management
  - Hepatectomy with fenestration
    - Possible by the frequently asymmetric distribution
    - Very selected patient



Bernts LHP, Echternach SG, Kievit W, Rosman C, Drenth JPH. Clinical response after laparoscopic fenestration of symptomatic hepatic cysts: a systematic review and meta-analysis. Surg Endosc. 2019;33(3):691-704.

TABLE 73.2 Treatment of PCLD by Combined Resection and Fenestration

REFERENCE, YEAR	NO. PATIENTS	MORTALITY	MORBIDITY	RECURRENT SYMPTOMS	FOLLOW-UP
Turnage et al., 1988[163]	3	2 (67%)	2 (67%)	33%	10 mo
Vauthey et al., 1991[176]	5	0	5 (100%)	0	14 mo
Newman et al., 1990[a,177]	9	1	5	1	17 mo
Henne-Bruns et al., 1993[178]	8	0	3 (38%)	50%	15 mo
Madariaga et al., 1993[179]	2	0	NA	0	>96 mo
Que et al., 1995[a,174]	31	1 (3%)	15 (48%)	3%	28 mo
Soravia et al., 1995[180]	10	1 (10%)	2 (20%)	33%	68 mo
Koperna et al., 1997[166]	5	0	NA	0	—
Martin et al., 1998[167]	9	0	6	0	NA
Vons et al., 1998[181]	12	1 (8%)	10 (83%)	2	34 mo
Hansman et al., 2001[61]	2	0	0	0	NA
Yang et al., 2004[182]	7	0	7 (100 %)	0%	20 mo
Li et al., 2008[183]	21	0	16 (76%)	14%	61 mo
Schnelldorfer et al., 2009*,[104]	124	4 (3%)	78 (63%)	—	96 mo
Aussilhou et al., 2010[175]	45	2 (4%)	32 (71%)	30%	41 mo
Bernts et al., 2020[184]	18	0	2 (11%)	5.5%	6 mo
TOTAL	253	10 (4%)	161 (64%)		

[a]Studies held at the same institution.

NA, Not available; *PCLD,* polycystic liver disease.

Olivier Farges and Béatrice Aussilhou, Simple cysts and polycystic liver disease : Clinical and radiographic features, surgical and nonsurgical management. Blumgart's Surgery of the Liver, Biliary Tract and Pancreas, Chapter 75, 1122-1147.e3,

Polycystic liver disease

- Management
  - Liver transplantation
    - Only curative treatment
    - In patients with ADPKD, Simultaneous kidney transplant is most often indicated.

Criteria to refer patients with polycystic liver disease for liver transplantation

1. Clinically apparent liver disease due to massive polycystic liver severely affecting quality of life
2. Massive polycystic liver disease and complication(s), that can exclusively be treated by liver transplantation

Complications include: severe malnutrition, hepatic venous outflow obstruction, ascites, portal hypertension, variceal haemorrhage, recurrent hepatic cyst infections

3. Failure of non-transplant related interventions and contraindications for non-transplant related interventions

Criteria to consider referral for combined liver-kidney transplantation

1. Creatinine clearance <30 ml/min

Martin AP, Bartels M, Kliene A, et al. Polycystic liver and kidney disease: post-transplant kidney function in patients receiving preemptive kidney transplantation. Transpl Int. 2008;21:263-267.

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

REFERENCE, YEAR	NO. PATIENTS	LT	LT + KT	DEATHS
Starzl et al., 1990[199]	4	2	2	1
Uddin et al., 1995[200]	3	3	0	1
Klupp et al., 1996[201]	10	5	5	1
Washburn et al., 1996[202]	5	4	1	1
Lang et al., 1997[197]	17	9	8	5
Swenson et al., 1998[203]	9	6	3	1
Jeyarajah et al., 1998[192]	6	3	3	2
Pirenne et al., 2001[193]	16	15	1	2
Becker et al., 2003[204]	17	0	17	3
Gustafsson et al., 2003[205]	7	4	3	0
Kirchner et al., 2006[206]	36	21	15	5
Ueno et al., 2006[194]	14	9	5	1
Krohn et al., 2008[207]	14	11	3	1
Taner et al., 2009[196]	13	6	7	3
Schnelldorfer et al., 2009[104]	7	4	3	2
Aussilhou et al., 2010[175]	27	4	23	4
Le Roy et al., 2019[208]	15	4	11	0
TOTAL	220	11	110	33 (15%)

KT, Kidney transplantation; *LT,* liver transplantation; *PCLD,* polycystic liver disease.

Olivier Farges and Béatrice Aussilhou, Simple cysts and polycystic liver disease : Clinical and radiographic features, surgical and nonsurgical management. Blumgart's Surgery of the Liver, Biliary Tract and Pancreas, Chapter 75, 1122-1147.e3,

RESEARCH ARTICLE Open Access

Liver transplantation in adult polycystic liver disease: the Ontario experience

Mohammed Alsager[1], Shuet Fong Neong[2], Radhika Gandhi[1], Anouar Teriaky[1], Ephraim Tang[2], Anton Skaro[2], Karim Qumosani[1], Les Lilly[2], Zita Galvin[2], Nazia Selzner[2], Mamatha Pallavi Bhat[2], Klajdi Puka[3] and Mayur Brahmania[1,4*]

Toronto General Hospital and London Health Sciences Center

Retrospective database review

A total of 3560 patients underwent LT, of whom 51 (1.4%) had PCLD and met inclusion criteria

- 33 (65%) had PCLD
- 9 (17.5%) had ADPKD with primary disease
- 9 (17.5%) had both diseases

Mortality 4(7.8%)

Alsager M, Neong SF, Gandhi R, Liver transplantation in adult polycystic liver disease: the Ontario experience. BMC Gastroenterol. 2021 Mar 9;21(1):115.

Mucinous cystic neoplasm

- Incidence less than 5% of all liver cyst
- Predominate in female(90%), middle age
- May present low-grade dysplasia, high-grade dysplasia, or invasive carcinoma
- CEA and CA 19-9 may be elevated, particularly in patients with invasive carcinoma.(But cannot used for distinguish simple liver cyst or PCLD from MCN)
- Malignant transformation 5 – 10%(Compared with pancrease 10 - 36%)

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

Mucinous cystic neoplasm

- WHO define as

  " Cyst-forming epithelial neoplasm, typically showing no communication with the bile ducts, composed of cuboidal to columnar, variably mucin-producing epithelium, associated with ovarian-type subepithelial stroma"

- Clinical presentation
  - Various clinical manifestations: abdominal pain, fullness, or early satiety due to large size and mass effect
  - Asymptomatic

Soares KC, Arnaoutakis DJ, Kamel I, Anders R, Adams RB, Bauer TW, Pawlik TM. Cystic neoplasms of the liver: biliary cystadenoma and cystadenocarcinoma. J Am Coll Surg 2014;218:119–28

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

Mucinous cystic neoplasm

- Imaging characteristic
  - Ultrasound : single, massive (average size of 11 cm (ranging 5-23 cm)), multilocular, or sporadically unilocular(6–10%) cystic masses with thick uneven walls and echogenic septa
    - Absence of mural nodule is favor benign
  - CT : large, well-circumscribed, multiloculated cystic mass with a clearly defined fibrotic capsule, mural calcification(47-63%)
    - 76% occur in the left hepatic lobe

\

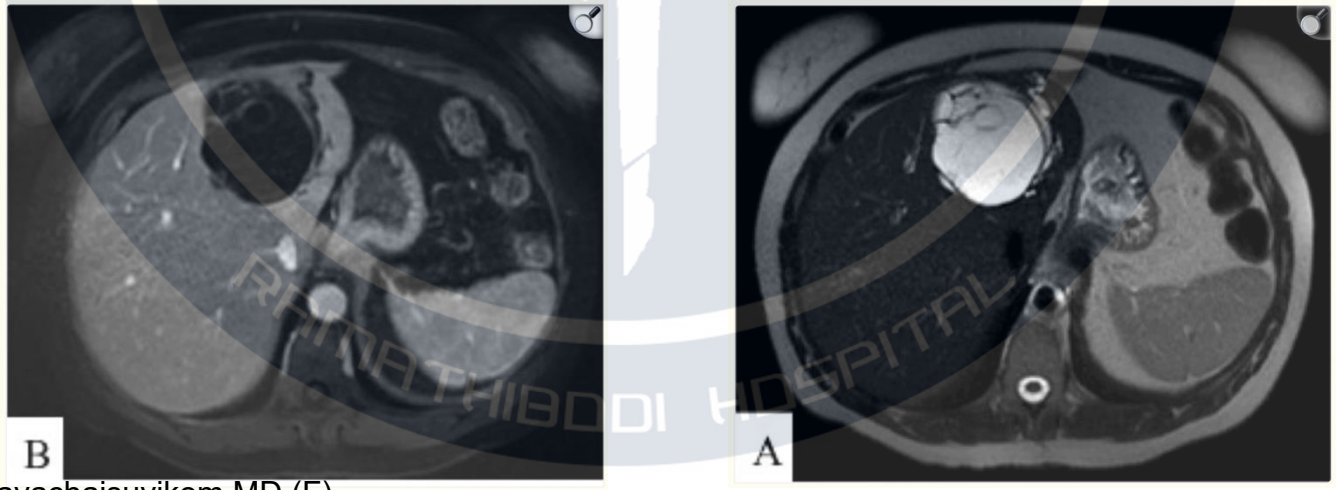
Soares KC, Arnaoutakis DJ, Kamel I, Anders R, Adams RB, Bauer TW, Pawlik TM. Cystic neoplasms of the liver: biliary cystadenoma and cystadenocarcinoma. J Am Coll Surg 2014;218:119–28

Lantinga MA, Gevers TJG, Drenth JPH. Evaluation of hepatic cystic lesions. World J Gastroenterol 2013;19:3543–54

Kim HJ, Yu ES, Byun JH, Hong S-M, Kim KW, Lee JS, Kim SY. CT differentiation of mucin-producing cystic neoplasms of the liver from solitary bile duct cysts. AJR Am J Roentgenol 2014

Mucinous cystic neoplasm

- Imaging characteristic
  - MRI
    - multi-locular with irregular thick walls
    - cyst fluid content by varying signal intensities on T1 weighted images depending on cyst fluid protein content



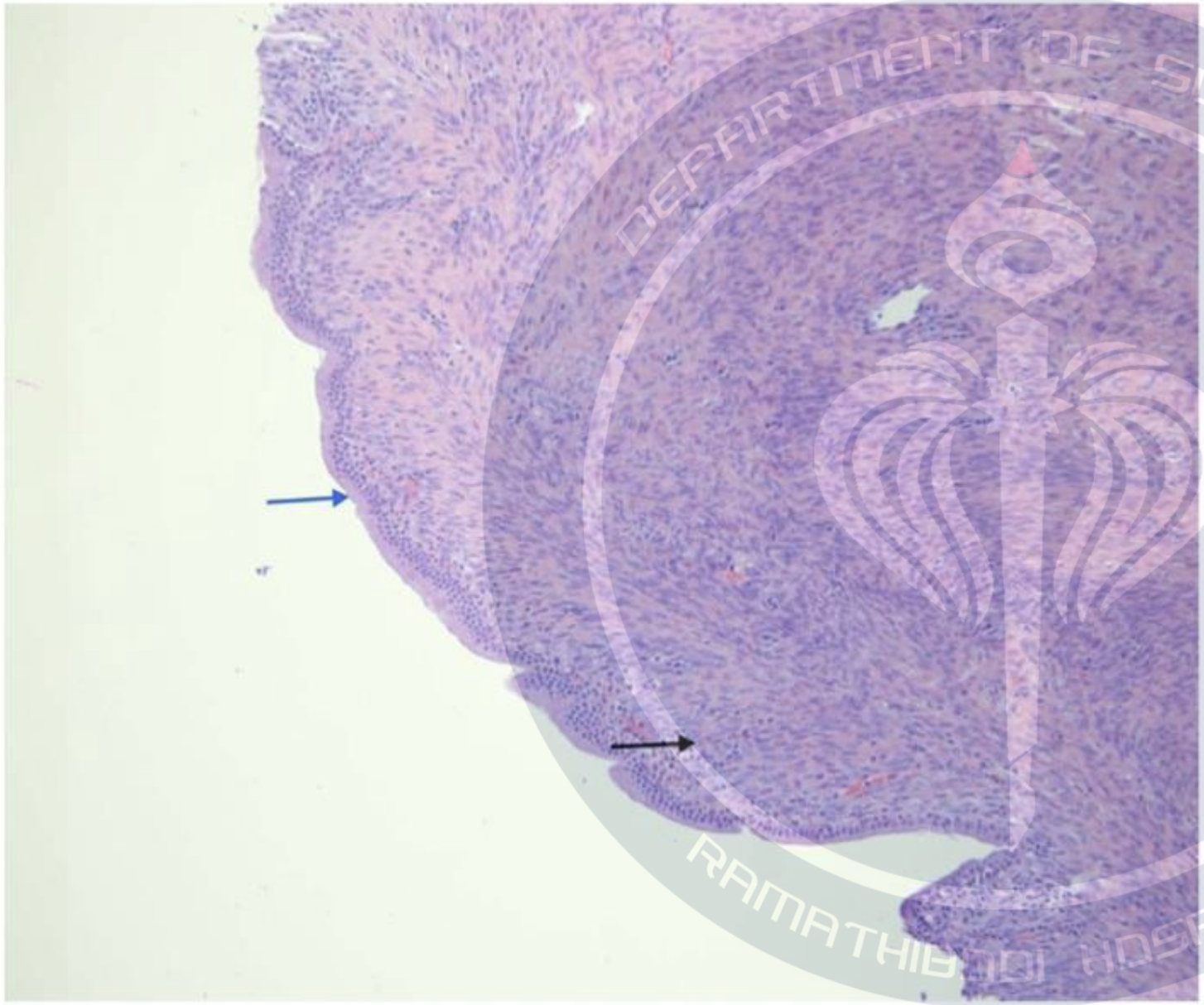
Soares KC, Arnaoutakis DJ, Kamel I, Anders R, Adams RB, Bauer TW, Pawlik TM. Cystic neoplasms of the liver: biliary cystadenoma and cystadenocarcinoma. J Am Coll Surg 2014;218:119–28

Mucinous cystic neoplasm

- Kim HJ et al.
  - solitary bile duct cysts and mucinous cystic neoplasms with a sensitivity of 87% and a specificity of 87%, by using 2-of-5 characteristic selection criteria
    - presence of septa
    - central septa
    - mural nodules
    - bile duct dilation either upstream or downstream

Presence of ovarian-like hypercellular stroma and absence of bile duct communication are the 2 hallmarks that differentiate MCNs from IPNB

Kim HJ, Yu ES, Byun JH, Hong S-M, Kim KW, Lee JS, Kim SY. CT differentiation of mucin-producing cystic neoplasms of the liver from solitary bile duct cysts. AJR Am J Roentgenol 2014

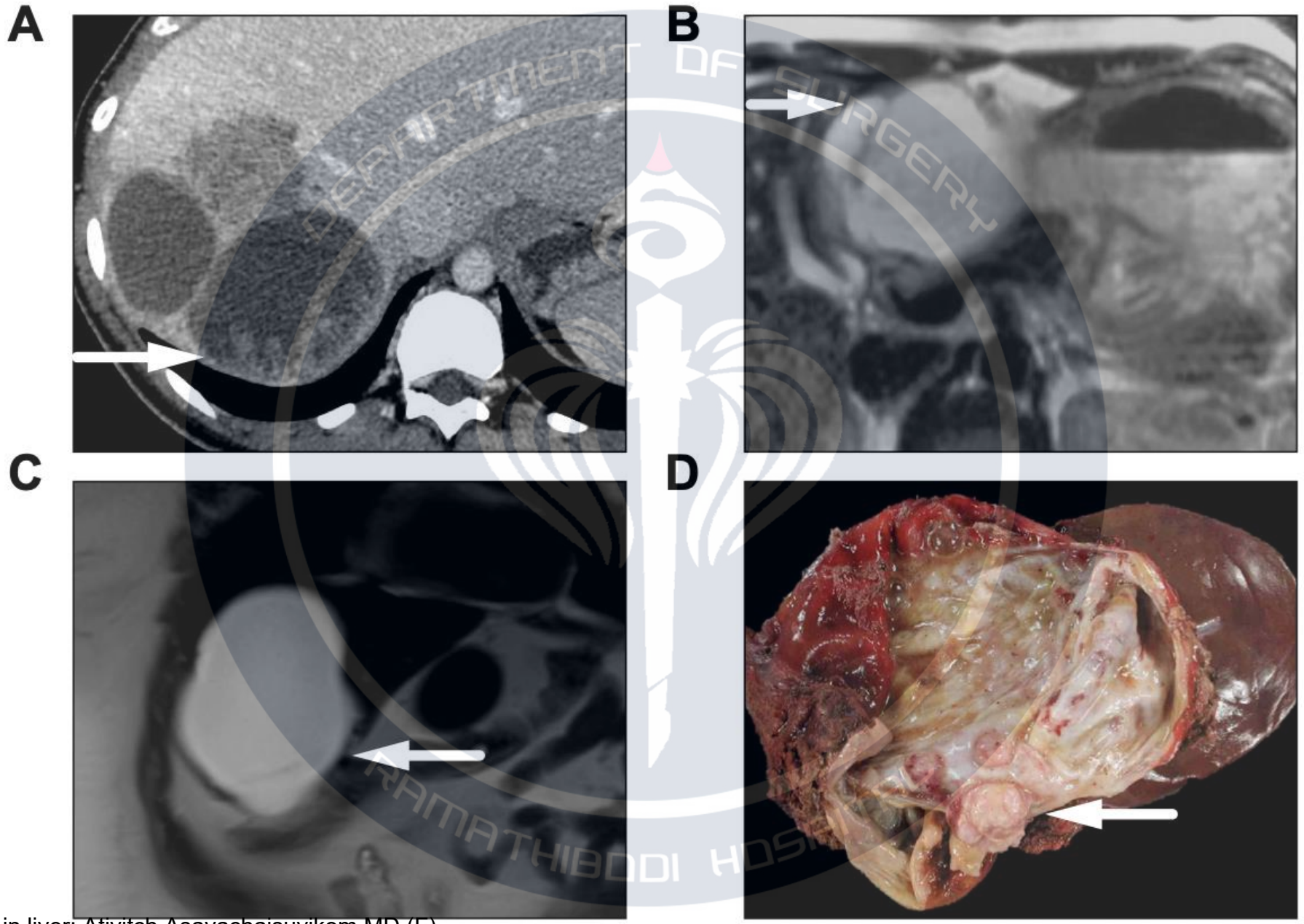


Ovarian like stroma

	Mucinous Cystic Neoplasm of the Liver	Intraductal Papillary Neoplasms of the Bile Duct
Radiographic Findings	Multilocular fluid-filled mass, often with smaller cysts within the cyst wall	Multicystic with a grape-like appearance, papillary nodules, and peripheral bile duct dilation
Ductal Communication	Usually absent	Present
Stroma	Ovarian-like stroma, PR+, ER+	Fibrous
Epithelial Antigens	MUC5AC-, CK7+, CK20-, MUC2-, MUC6-	MUC5AC+, Variable CK7, CK20, MUC2, MUC6
Malignancy Potential	Low	High

Abbreviations: +, positive for expression; -, negative for expression; ER, Estrogen Receptor; PR, Progesterone Receptor; MUC5AC, Mucin 5AC; CK7, Cytokeratin 7; CK20, Cytokeratin 20; MUC2, Mucin 2; MUC6, Mucin 6.

Hutchens JA, Lopez KJ, Ceppa EP. Mucinous Cystic Neoplasms of the Liver: Epidemiology, Diagnosis, and Management. Hepat Med. 2023 Mar 29;15:33-41. doi: 10.2147/HMER.S284842. PMID: 37016682; PMCID: PMC10066895.



Cystic tumor in liver: Ativitch Asavachaisuvikom,MD.(F)

Drenth, Joost et al., EASL Clinical Practice Guidelines on the management of cystic liver diseases, Journal of Hepatology, Volume 77, Issue 4, 1083 - 1108

Mucinous cystic neoplasm

- Management
  - Preoperative tissue biopsy
    - Benefit remain unclear
    - May be indicate in unclear imaging in high risk patients
    - Increase risk of tumor seeding or rupture, low sensitivity

Kim HJ, Yu ES, Byun JH, Hong S-M, Kim KW, Lee JS, Kim SY. CT differentiation of mucin-producing cystic neoplasms of the liver from solitary bile duct cysts. AJR Am J Roentgenol 2014

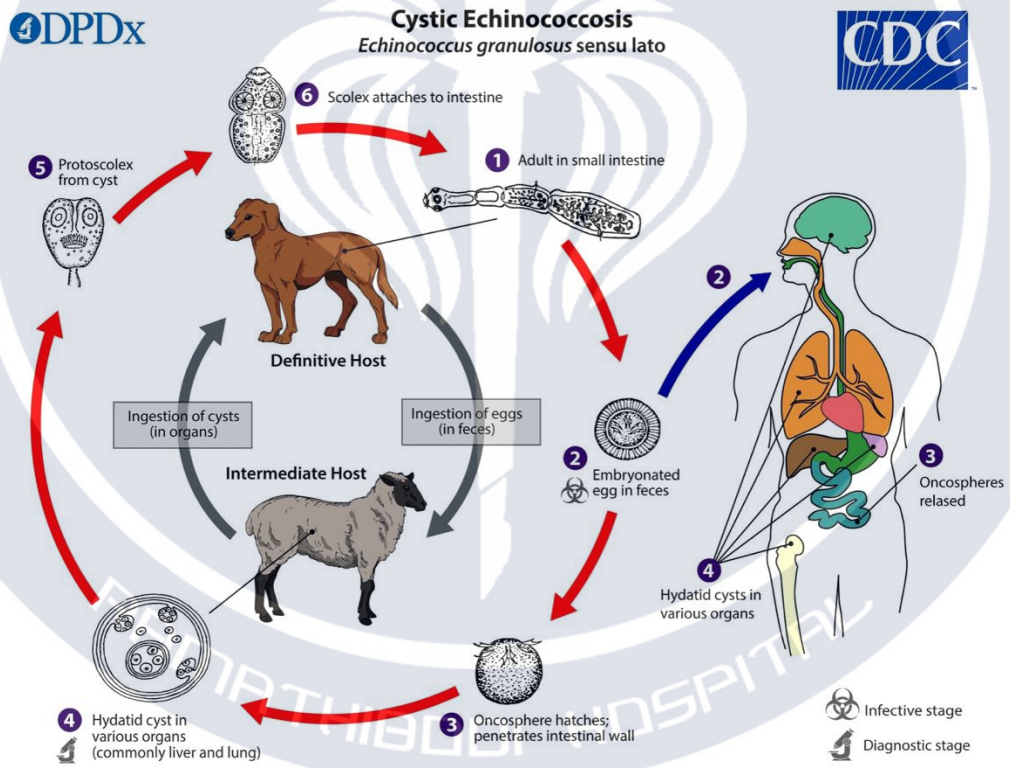
Mucinous cystic neoplasm

- Management
  - Fenestration, Aspiration, Partial resection
    - Not recommend
    - recurrence rate as high as 80-90%
  - Surgical resection with negative margins is recommended

Kim HJ, Yu ES, Byun JH, Hong S-M, Kim KW, Lee JS, Kim SY. CT differentiation of mucin-producing cystic neoplasms of the liver from solitary bile duct cysts. AJR Am J Roentgenol 2014

Hydatid disease

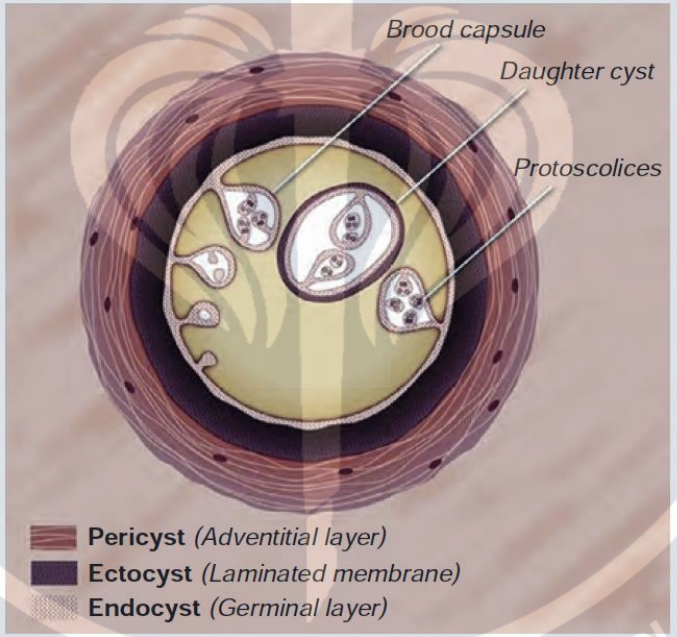
- Caused by the larval Echinococcus sp.

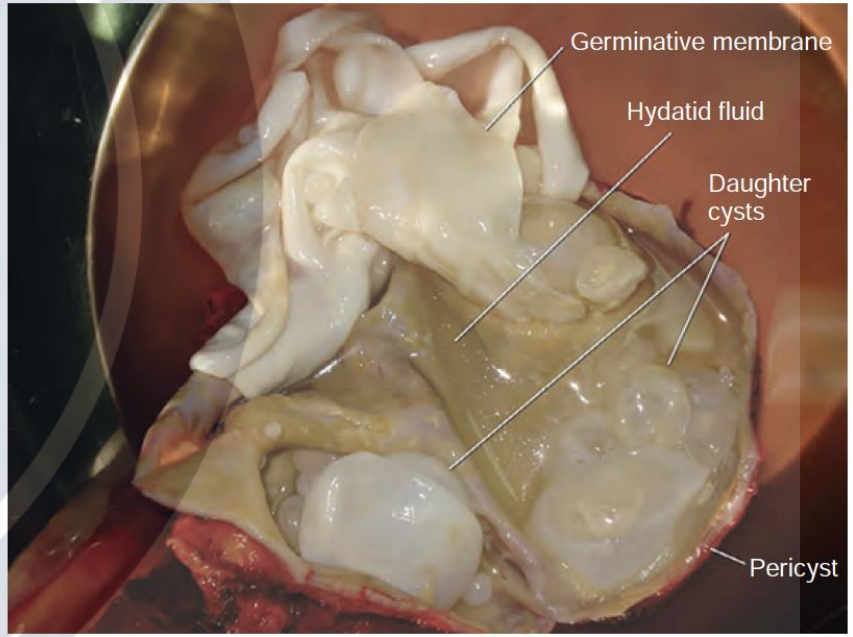


Bakoyiannis A, Delis S, Triantopoulou C, Dervenis C. Rare cystic liver lesions: a diagnostic and managing challenge. World J Gastroenterol. 2013 Nov 21;19(43):7603-19.

Hydatid disease

- Epidemiology
  - Some areas, such as Central Asia
- Diagnosis test
  - Serology : hemagglutination, ELISA
  - Confirmatory test : arc-5 immunoelectrophoresis (positive rate 91.1%)



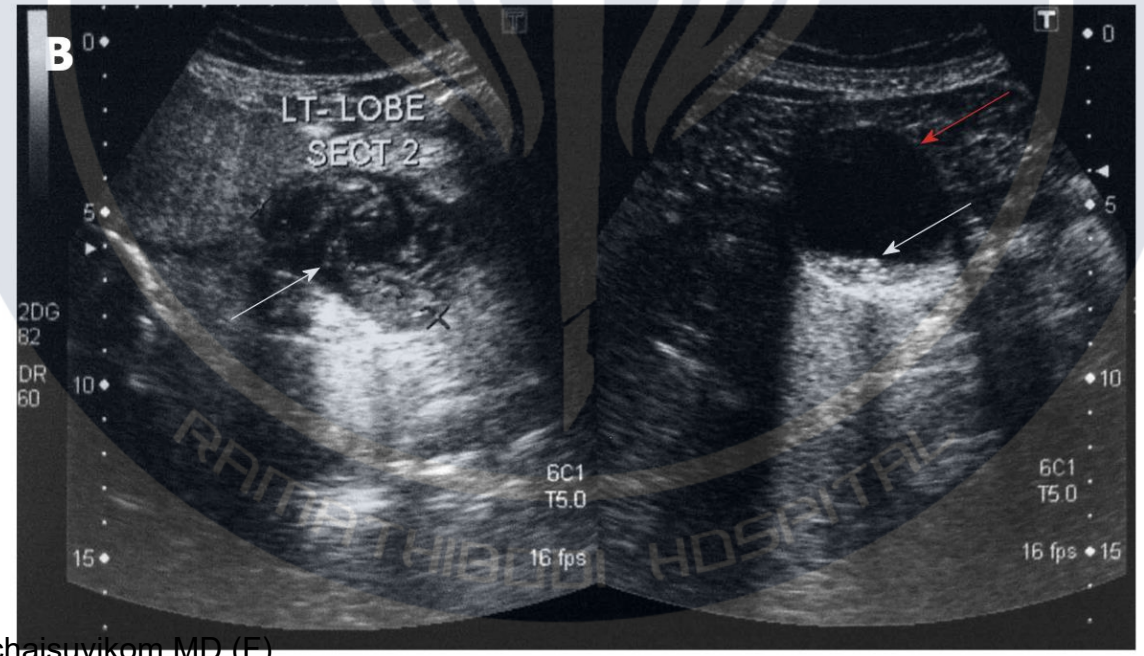


Bakoyiannis A, Delis S, Triantopoulou C, Dervenis C. Rare cystic liver lesions: a diagnostic and managing challenge. World J Gastroenterol. 2013 Nov 21;19(43):7603-19.

Hydatid disease

- Imaging
  - US : internal structure, number, and location of the cysts and the presence of complications
  - The specificity of US is in the range of 90%

an anechoic mass
with hydatid sand



The detached and folded
endocyst membrane

Cystic tumor in liver: Ativitch Asavachaisuvikom,MD.(F)

Bakoyiannis A, Delis S, Triantopoulou C, Dervenis C. Rare cystic liver lesions: a diagnostic and managing challenge. World J Gastroenterol. 2013 Nov 21;19(43):7603-19.

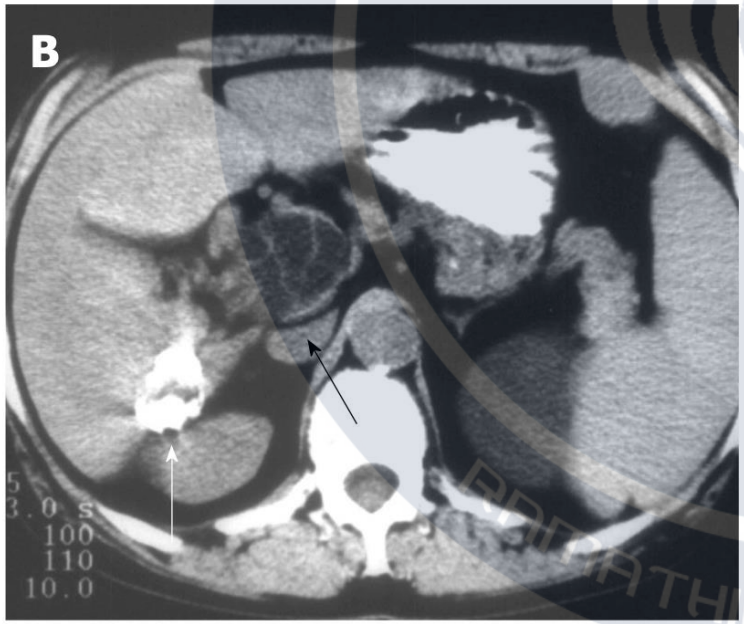
Table 1 Gharbi classification of cystic hydatid disease

Type	Ultrasonographic features and patterns
I	Pure fluid collection
II	Fluid collection with a split wall (water-lily sign)
III	Fluid collection with septa (honeycomb sign)
IV	Heterogeneous echographic patterns
V	Reflecting thick walls

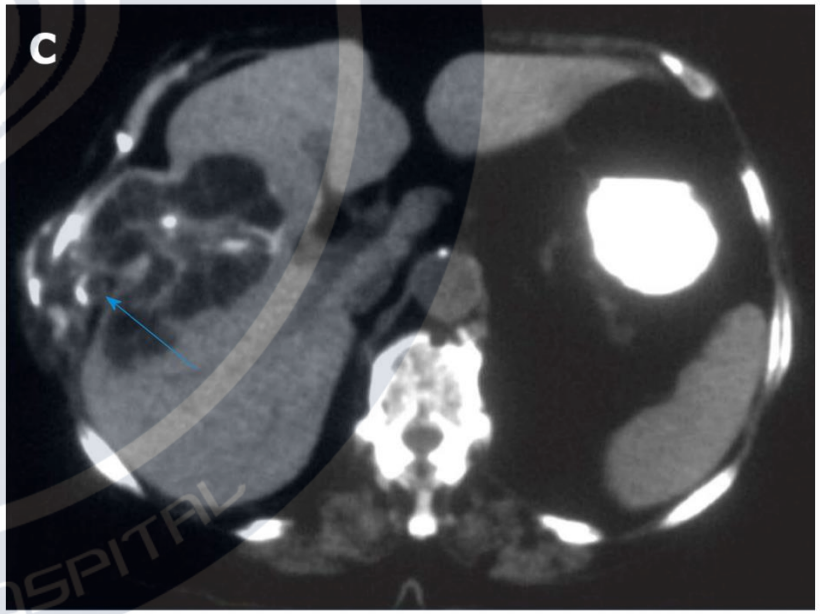
Bakoyiannis A, Delis S, Triantopoulou C, Dervenis C. Rare cystic liver lesions: a diagnostic and managing challenge. World J Gastroenterol. 2013 Nov 21;19(43):7603-19.

Hydatid disease

- Imaging
  - CT : calcified cystic walls, daughter cysts, and exogenous cysts, as well as evaluate their volume and density



multilocular cystic lesion in segment I, calcified mass with irregular margins in segment IV

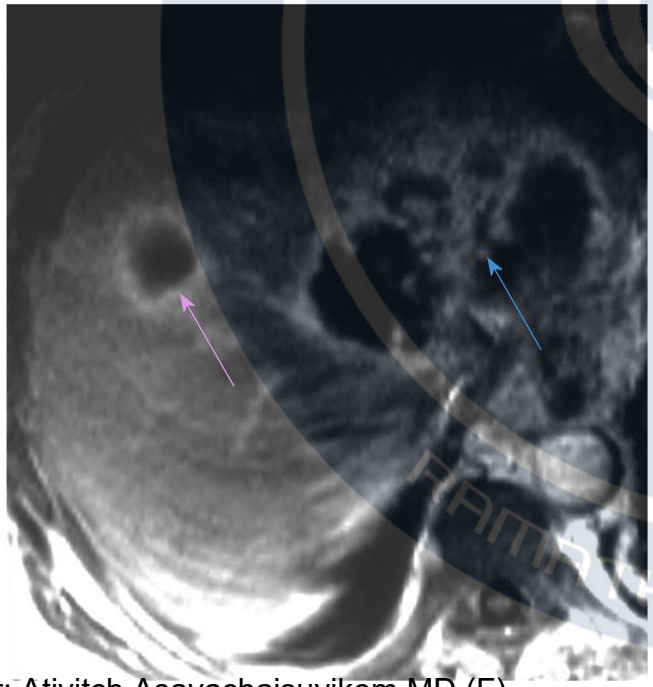


direct infiltration of a liver hydatid cyst in the adjacent peritoneal surface

Bakoyiannis A, Delis S, Triantopoulou C, Dervenis C. Rare cystic liver lesions: a diagnostic and managing challenge. World J Gastroenterol. 2013 Nov 21;19(43):7603-19.

Hydatid disease

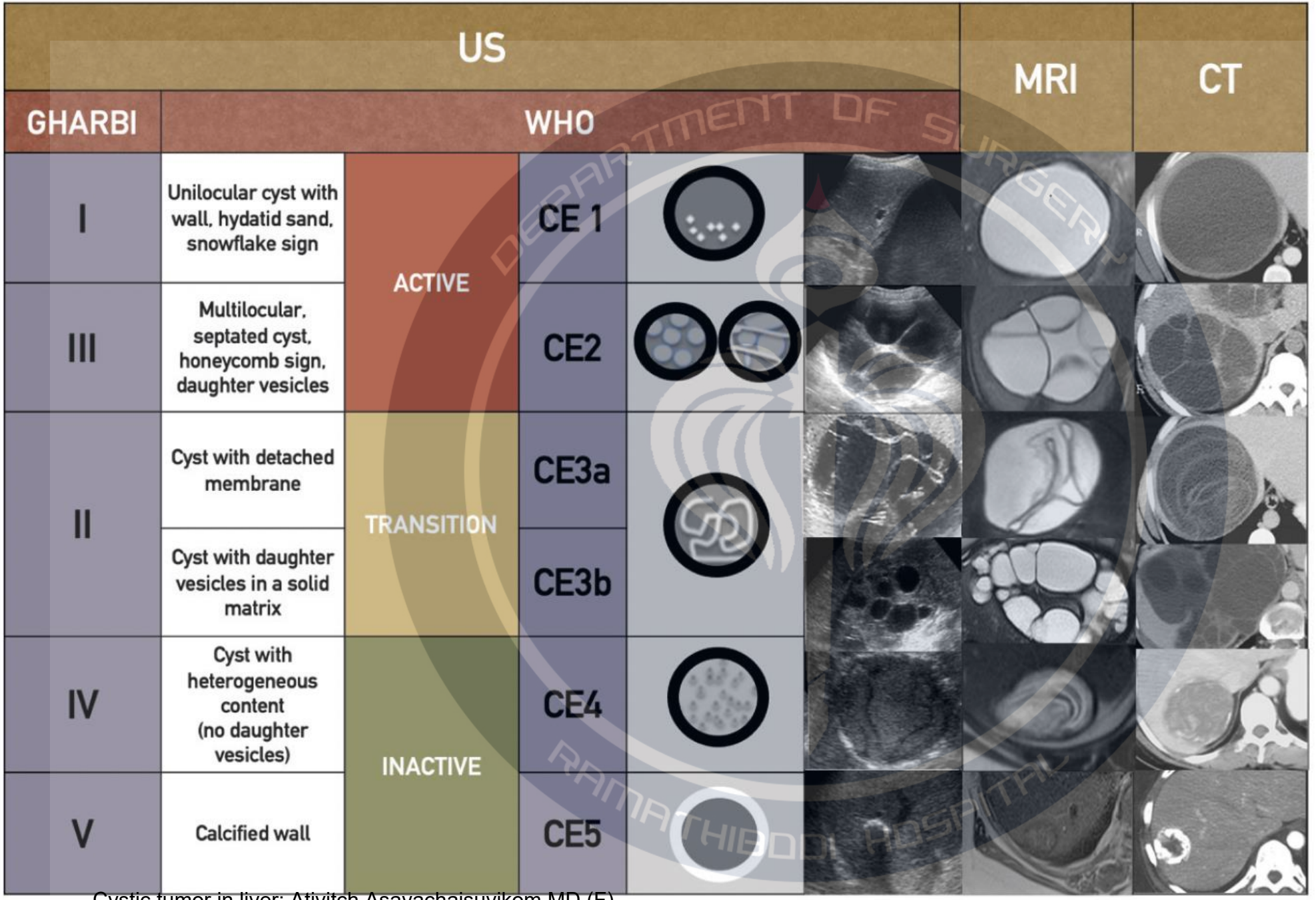
- Imaging
  - MRI : both CT and MRI have high specificity and sensitivity in the detection and differential diagnosis of hepatic cysts and extracapsular (satellite) cysts



multiloculated cystic liver lesion, indicative of the presence of daughter cysts

Cystic tumor in liver: Ativitch Asavachaisuvikom,MD.(F)

Bakoyiannis A, Delis S, Triantopoulou C, Dervenis C. Rare cystic liver lesions: a diagnostic and managing challenge. World J Gastroenterol. 2013 Nov 21;19(43):7603-19.



anechoic cyst with hyperechoic concentric halo

multilocular, multiseptated cysts

CE3a : Detachment membrane
CE3b : Daughter vesicle

heterogeneous, hypoechoic cysts(both cystic-solid)

Simisolid-looking cyst

Ferrer Inaebnit E, Molina Romero FX, Segura Sampedro JJ, González Argenté X, Morón Canis JM. A review of the diagnosis and management of liver hydatid cyst. Rev Esp Enferm Dig. 2022 Jan;114(1):35-41.

Hydatid disease

- Clinical
  - Compression
    - Bile duct compression, PV/HV compression
  - Rupture
    - secondary hydatidosis (incidence of 1-8 %)
    - Severe anaphylactic events occur in 1 % of cases
  - Bacterial overinfection
  - Biliary tree involvement:
    - The most common event, representing 40 %-60 % of complications
  - Respiratory tract involvement
    - Fistula

Ferrer Inaebnit E, Molina Romero FX, Segura Sampedro JJ, González Argenté X, Morón Canis JM. A review of the diagnosis and management of liver hydatid cyst. Rev Esp Enferm Dig. 2022 Jan;114(1):35-41.

Hydatid disease

- Diagnosis
  - Possible case : Clinical or epidemiologic history + imaging findings/serology positive
  - Probable case : Clinical + epidemiologic history + imaging + serology(two test)
  - Confirmed case : The above + demonstration of protoscolices, using direct microscopy/molecular biology, in the cyst contents by PAIR or surgery or changes US appearance

Ferrer Inaebnit E, Molina Romero FX, Segura Sampedro JJ, González Argenté X, Morón Canis JM. A review of the diagnosis and management of liver hydatid cyst. Rev Esp Enferm Dig. 2022 Jan;114(1):35-41.

Hydatid disease

- Management
  - Surgery
    - Relapse rates range from 1-20 %, morbidity rates from 12 - 84 %, and mortality rates from 0.5 - 6.5 %
    - Indication :
      - Size > 10 cm and/or isolated liver cysts superficially located at risk for rupture.
      - Large CE2 or CE3b with multiple daughter cyst
      - Complicated cysts (overinfection, biliary communication, compression of neighboring structure or obstruction).
    - Radical surgery is preferable given its lower risk for postoperative abdominal infection, biliary fistula, and overall mortality.
      - Cystopericystectomy
      - Hepatectomy(uncommon)

Ferrer Inaebnit E, Molina Romero FX, Segura Sampedro JJ, González Argenté X, Morón Canis JM. A review of the diagnosis and management of liver hydatid cyst. Rev Esp Enferm Dig. 2022 Jan;114(1):35-41.

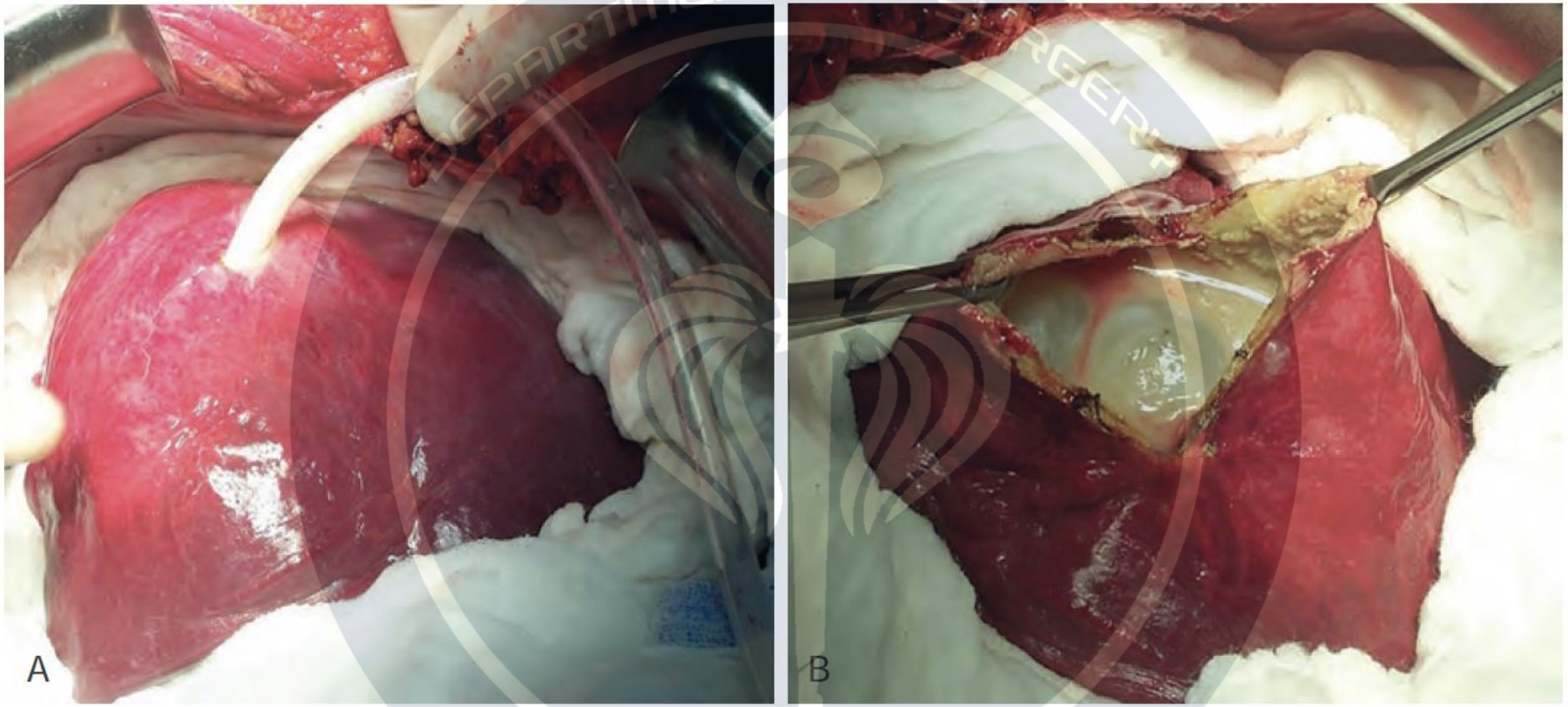
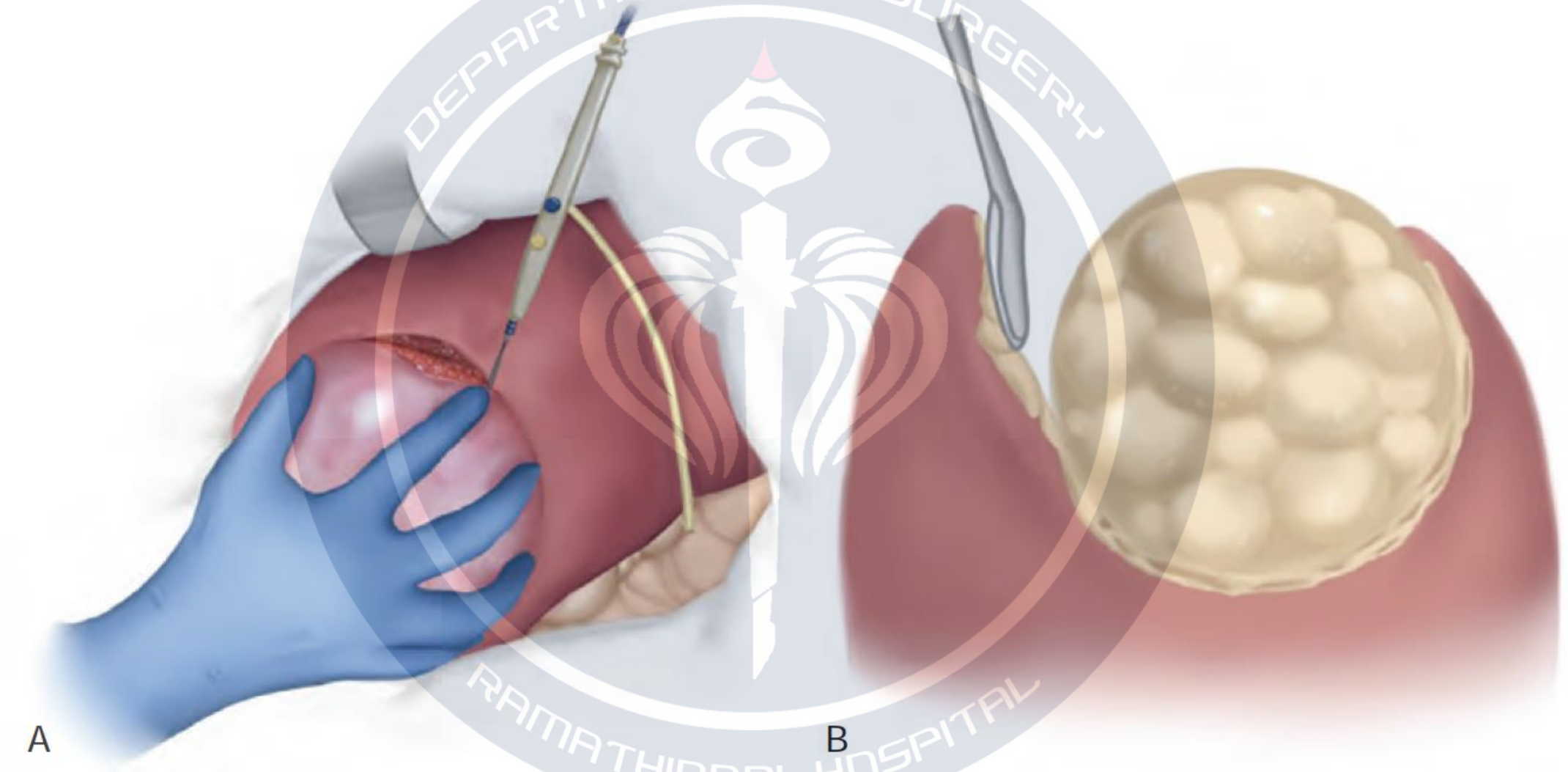


FIGURE 72.13 Operative view of the first surgical steps in the conservative approach. Note that area around the cysts is covered and isolated with packs that can be immersed in a scolicidal agent. Surrounding tissues should be protected from the spread of parasites during the cyst evacuation (a). Then the cyst is punctured and/or incised at its most accessible part (b).

Javier C. Lendoire, Emmanuel Melloul, and Oscar Cesar Imventarza , Hydatid disease of the liver, 7th Blumgart



Javier C. Lendoire, Emmanuel Melloul, and Oscar Cesar Imventarza , Hydatid disease of the liver, 7th Blumgart

Hydatid disease

- Management
  - Puncture, aspiration, injection, and reaspiration (PAIR)
    - Injecting a protoscolicidal agent, and reaspirating after 15-20 minutes
    - indicated for inoperable patients and/or patients who refuse surgery, in case of relapse after surgery, and in the setting of no response to medical therapy
    - contraindication cysts inaccessible to puncture, cysts with non-aspirable thick contents, when at risk of damaging vascular structures, peripheral cysts with inadequate liver tissue for safe trans-hepatic puncturing, inactive and/or calcified cysts, and presence of biliary, peritoneal, or pleural space communications

Pascal G, Azoulay D, Belghiti J, et al. Hydatid disease of the liver. In: Blumbgart's Surgery of the Liver, Biliary Tract, and Pancreas. 6th edition. Philadelphia:Elsevier; 2017. p. 1102-21.

Mihmanli M, Idiz UO, Kaya C, et al. Current status of diagnosis and treatment of hepatic echinococcosis. Rev Esp Enf Dig 2016;8(28):1169-81. DOI: 10.4254/wjh.v8.i28.1169

Hydatid disease

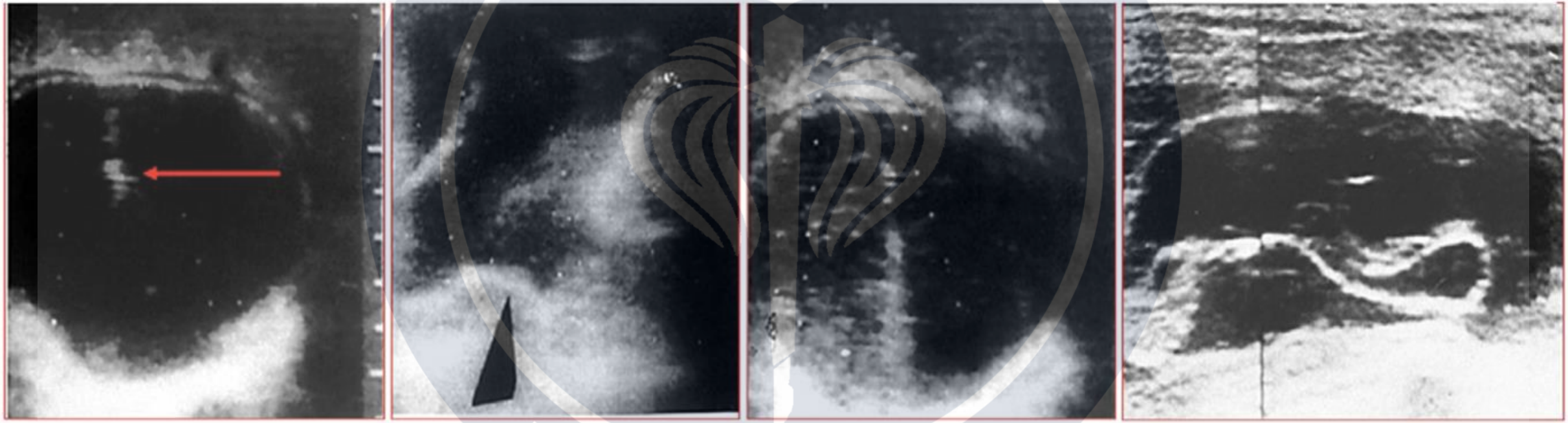
- Management
  - Puncture, aspiration, injection, and reaspiration (PAIR)
    - Unilocular CE1 and CE3a lesions respond well to percutaneous treatment (> 80 %)
    - Multilocular CE2 and CE3b cysts display a lower success rate, inferior to 40 %

      Relapse (1.6 %-5 %), morbidity (0.9 %), and mortality (2.5 %) rates are low, and hospital stays are shorter

Pascal G, Azoulay D, Belghiti J, et al. Hydatid disease of the liver. In: Blumbgart's Surgery of the Liver, Biliary Tract, and Pancreas. 6th edition. Philadelphia: Elsevier; 2017. p. 1102-21.

Mihmanli M, Idiz UO, Kaya C, et al. Current status of diagnosis and treatment of hepatic echinococcosis. Rev Esp Enf Dig 2016;8(28):1169-81. DOI: 10.4254/wjh.v8.i28.1169

P (Puncture) A (Aspiration) I (Injection) R (Re-aspiration)



Cystic tumor in liver: Ativitch Asavachaisuvikom,MD.(F)

Khuroo MS. Percutaneous Drainage in Hepatic Hydatidosis-The PAIR Technique: Concept, Technique, and Results. J Clin Exp Hepatol. 2021 Sep-Oct;11(5):592-602.

Hydatid disease

- Management
  - Medication
    - The most widely used drug is albendazole for 3-6 months
    - Options: Praziquantel and/or in combination with benzimidazoles
    - Medication alone reserved for small cyst, inoperable or multiorgan
    - Relapse has a 9-25 % rate, and mostly occurs at 2-8 years after treatment completion

Keong B, Wilkie B, Sutherland T, et al. Hepatic cystic echinococcosis in Australia: an update on diagnosis and management. ANZ J Surg 2018;88(1-2):26-31. DOI: 10.1111/ans.14117

Hydatid disease

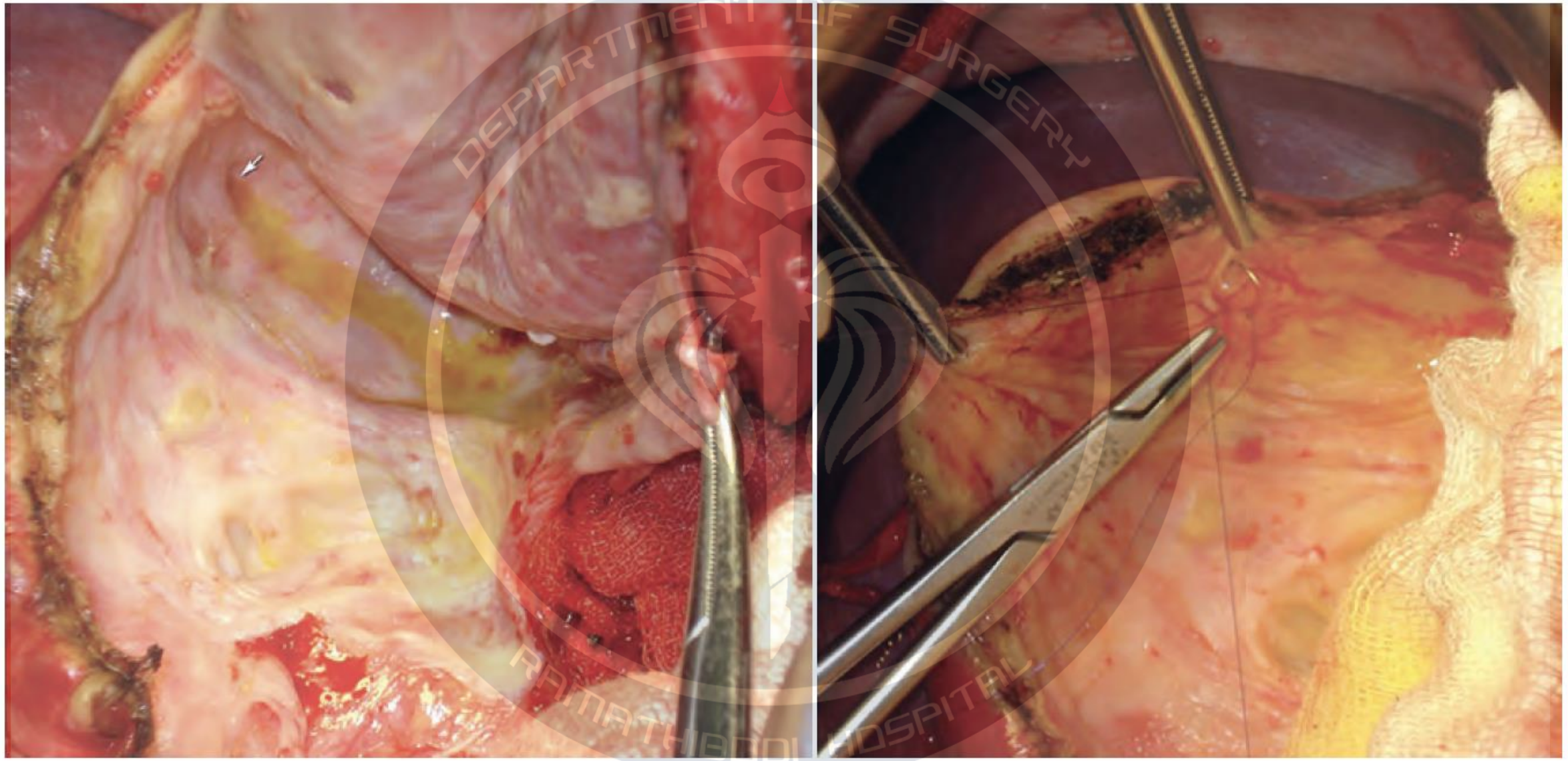
- Management
  - Active surveillance
    - Non-complicated cysts deemed quiescent or inactive by imaging tests (CE4 and CE5) do not require treatment but simply regular follow-up

Pascal G, Azoulay D, Belghiti J, et al. Hydatid disease of the liver. In: Blumbgart's Surgery of the Liver, Biliary Tract, and Pancreas. 6th edition. Philadelphia: Elsevier; 2017. p. 1102-21.

Hydatid disease

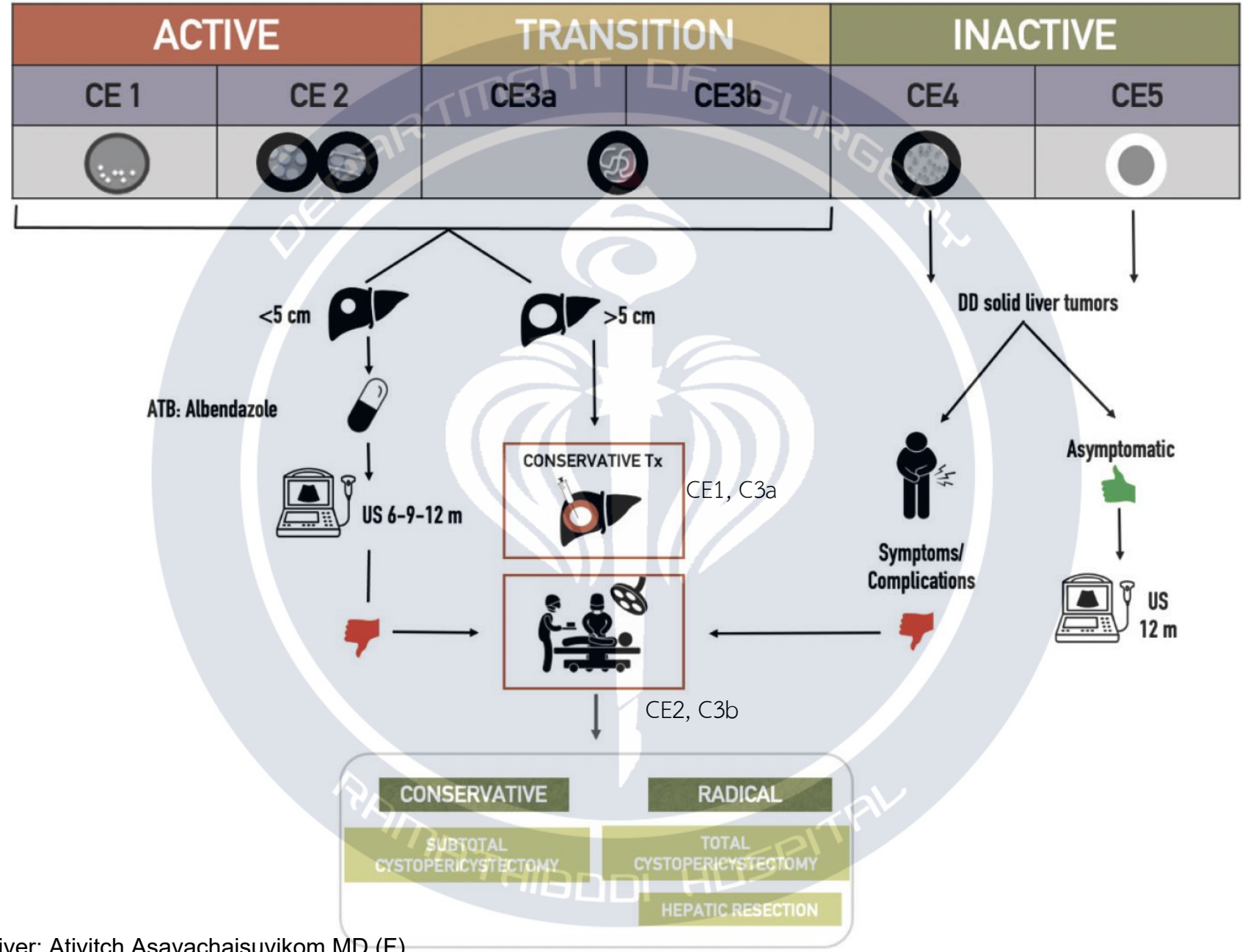
- Management of biliary complication
  - Preoperative
    - Cholangiopancreatography(ERCP) is useful to confirm biliary obstruction secondary to hydatid material, then endoscopic sphincterotomy and removal of hydatid remnants using a balloon or basket
  - Intraoperative
    - Intraoperative cholangiogram
    - Methylene blue test, inject via cystic duct
    - Management
      - obvious biliary orifices must be sutured to prevent postoperative bile leakage, fistula, or cavity infection
      - Dangerous suture : T-tube or Roux-en-Y HJ

Keong B, Wilkie B, Sutherland T, et al. Hepatic cystic echinococcosis in Australia: an update on diagnosis and management. ANZ J Surg 2018;88(1-2):26-31. DOI: 10.1111/ans.14117



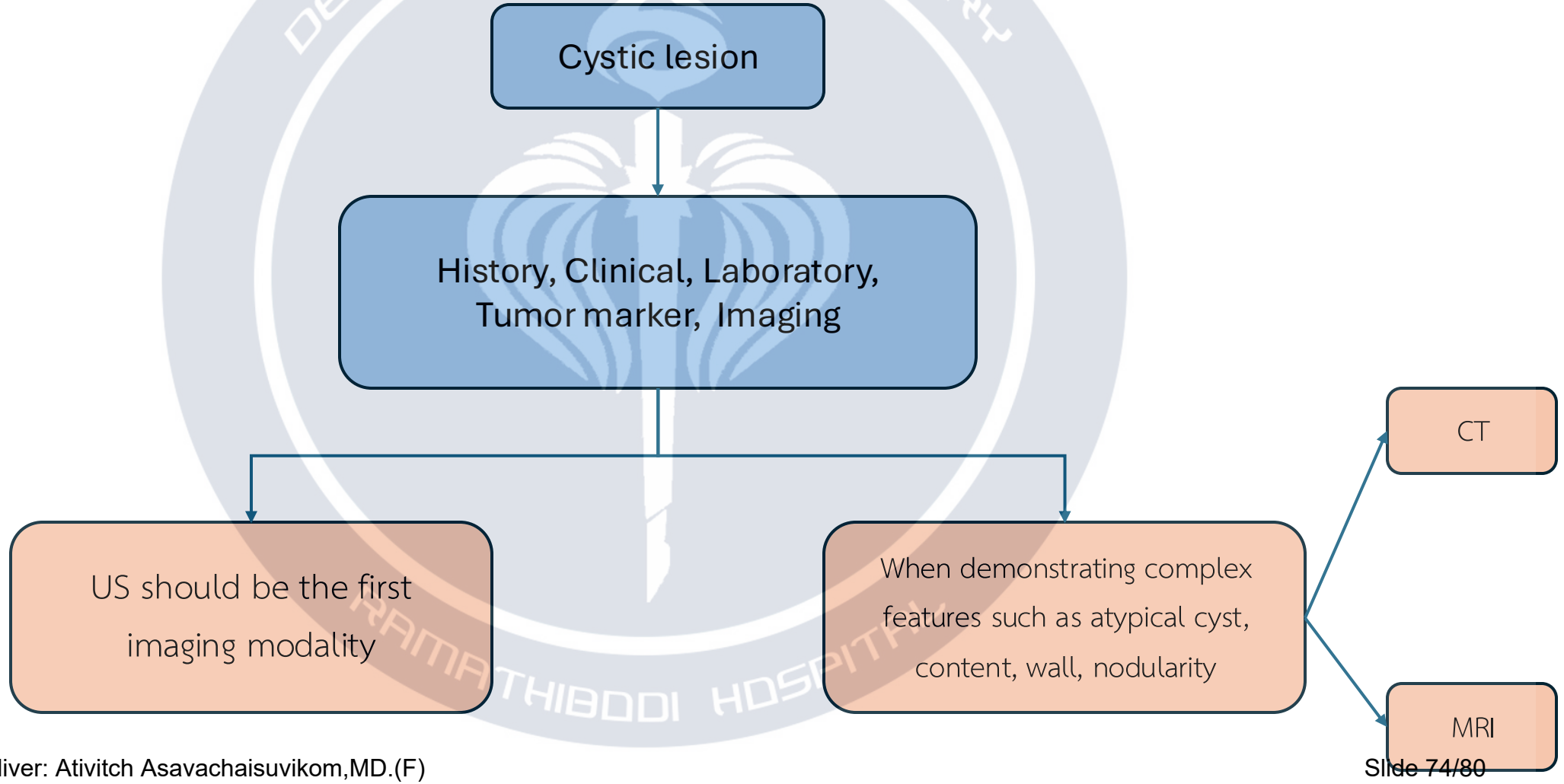
Cystic tumor in liver: Ativitch Asavachaisuvikom,MD.(F)

Javier C. Lendoire, Emmanuel Melloul, and Oscar Cesar Imventarza , Hydatid disease of the liver, 7th Blumgart

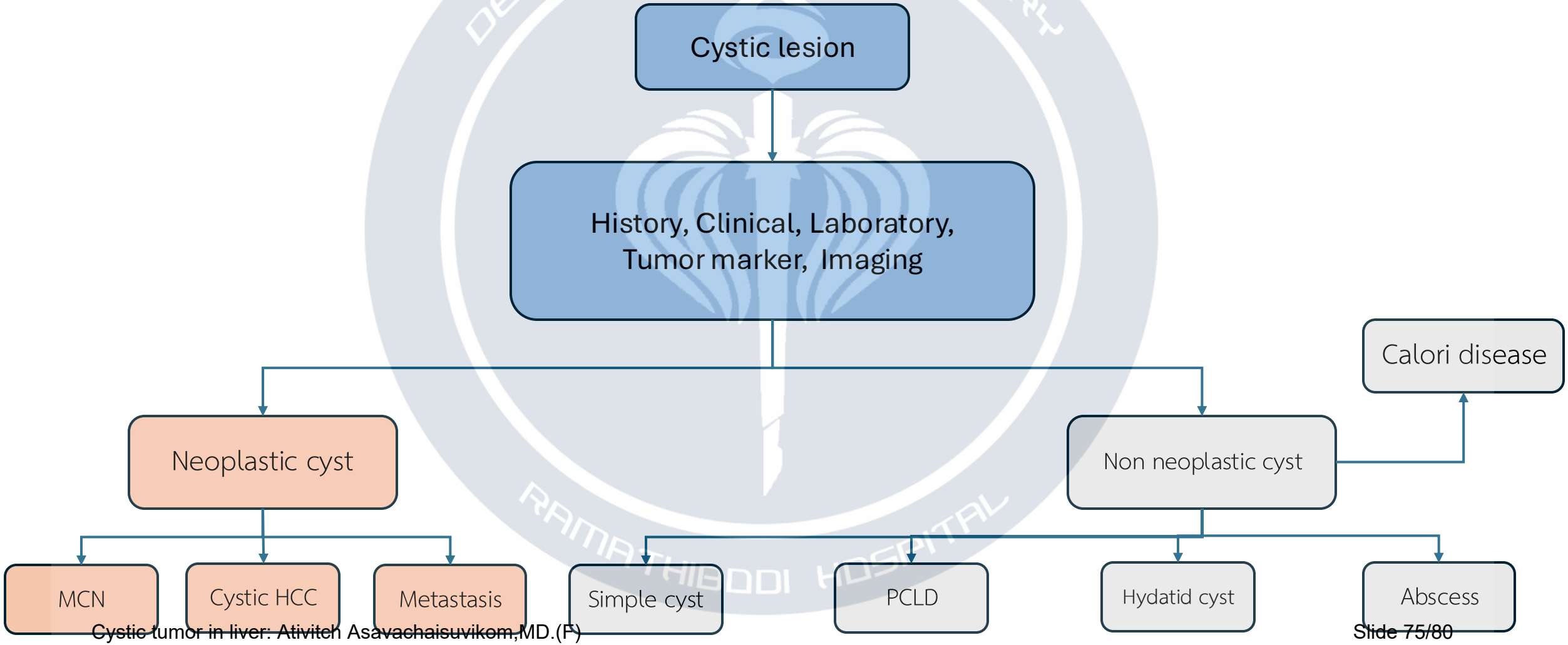


Ferrer Inaebnit E, Molina Romero FX, Segura Sampedro JJ, González Argenté X, Morón Canis JM. A review of the diagnosis and management of liver hydatid cyst. Rev Esp Enferm Dig. 2022 Jan;114(1):35-41.

Take home message



Take home message



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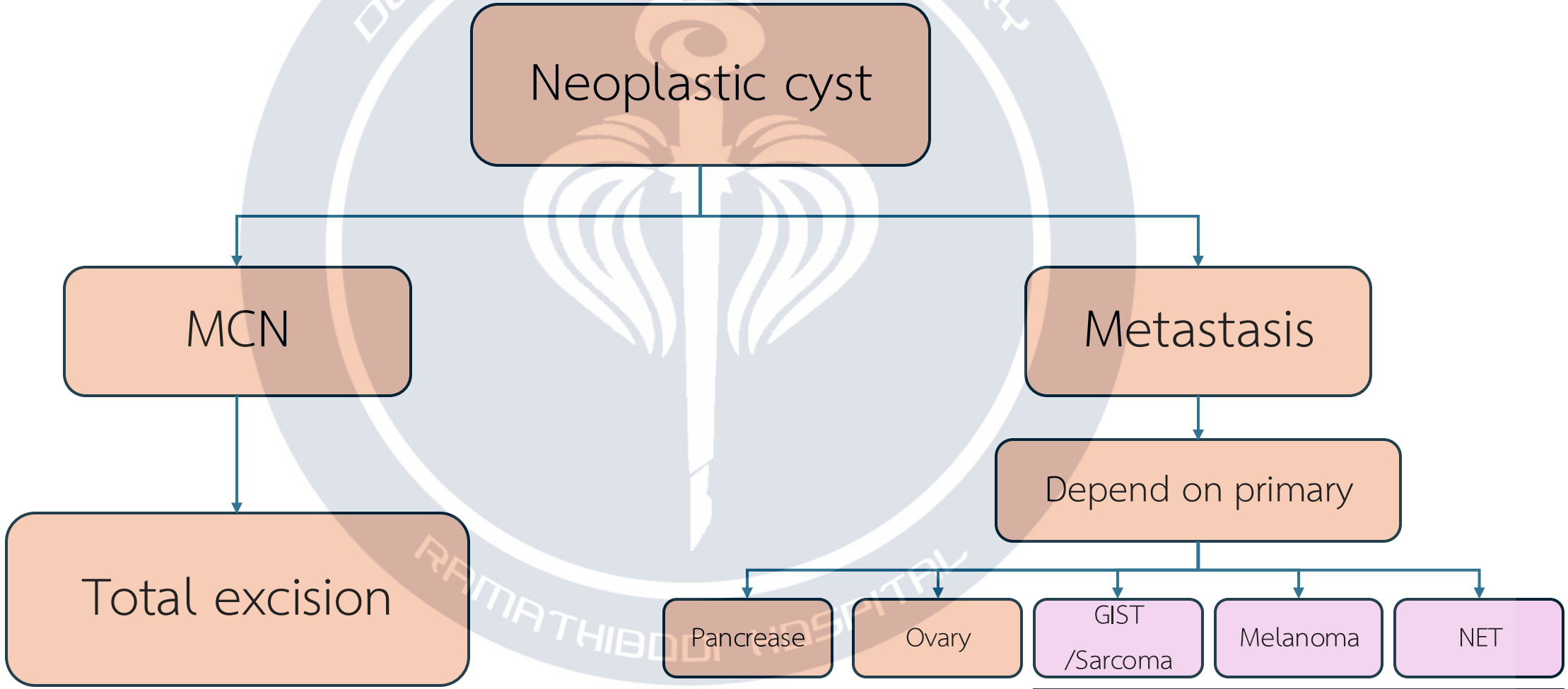

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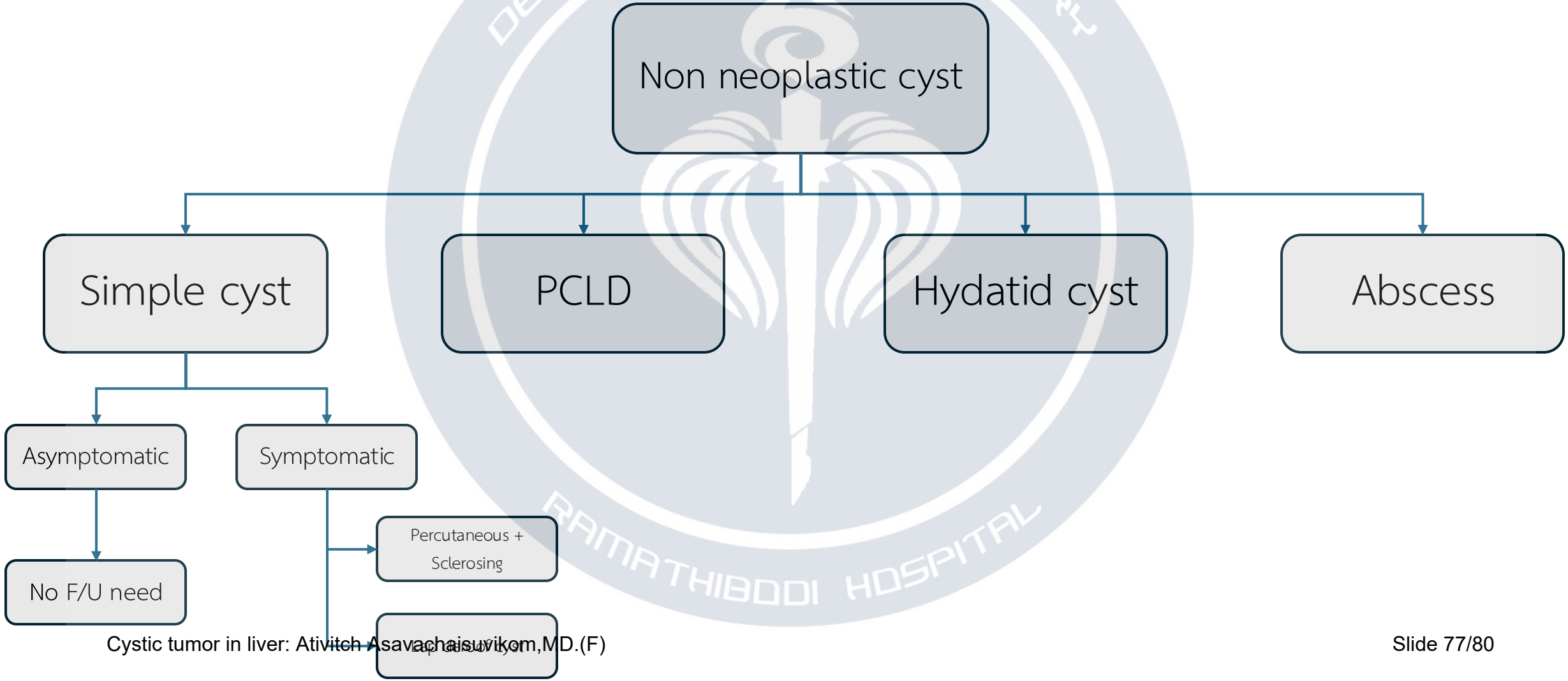

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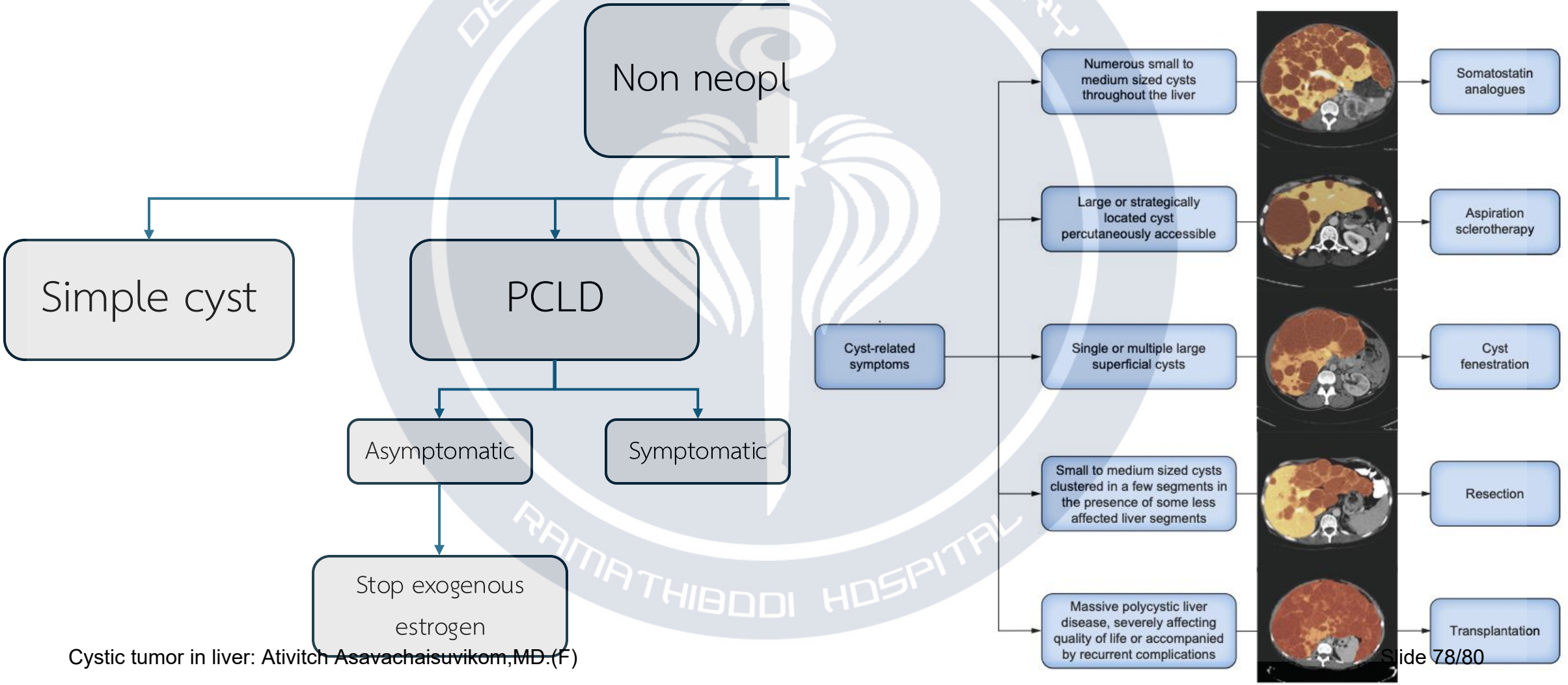

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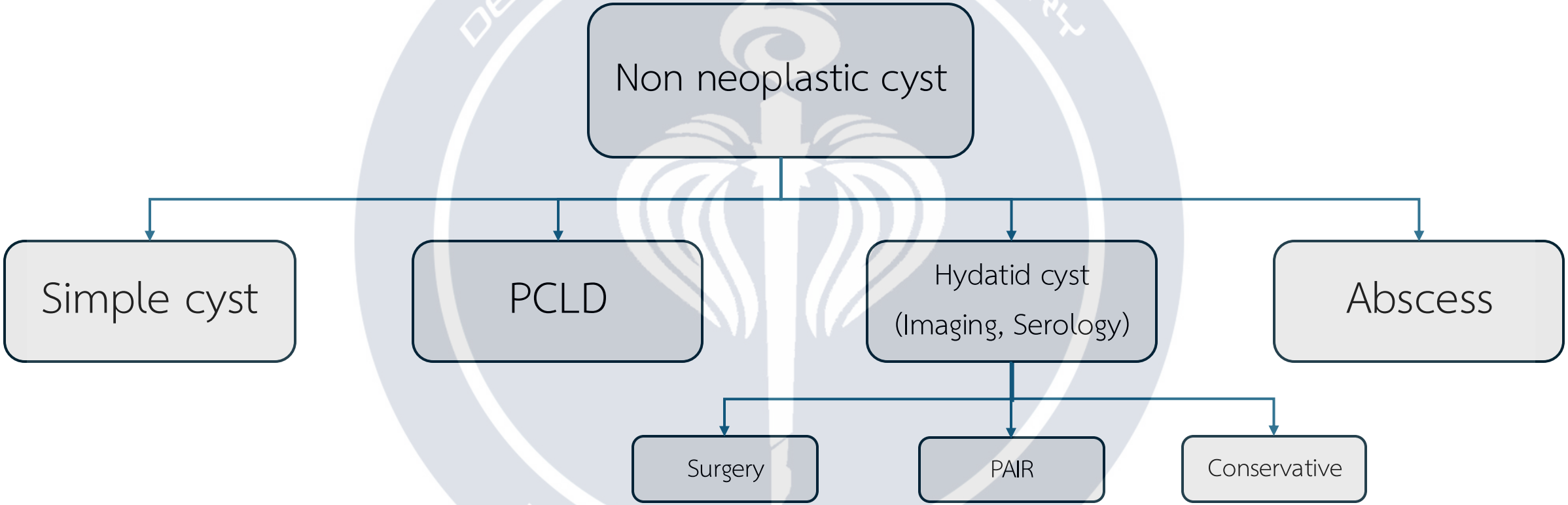

Thank you