



3^{ème} Colloque

Echinococcoses Kystique Méditerranée

Besançon, 18/19 novembre, 2025

Treatment options for cystic echinococcosis of the liver

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Cystic Echinococcosis is the name given to the disease caused by the parasite *Echinococcus granulosus sensu lato*

It is a disease common to humans and animals (Zoonosis) and classified by the WHO among the neglected zoonoses

The parasite vital cycle consists of an **adult form**, which lives in the intestines of dogs and other carnivores (**definitive hosts**), and a **larval form** that infects herbivores and, incidentally, humans (**intermediate hosts**).

This larval form of the parasite is the hydatid cyst

The hydatid cyst has been known in humans for thousands of years. It was first described by Hippocrates as a "water-filled vesicle"... But only in the 20th century **Félix Dévé**, of Rouen, gave the definition



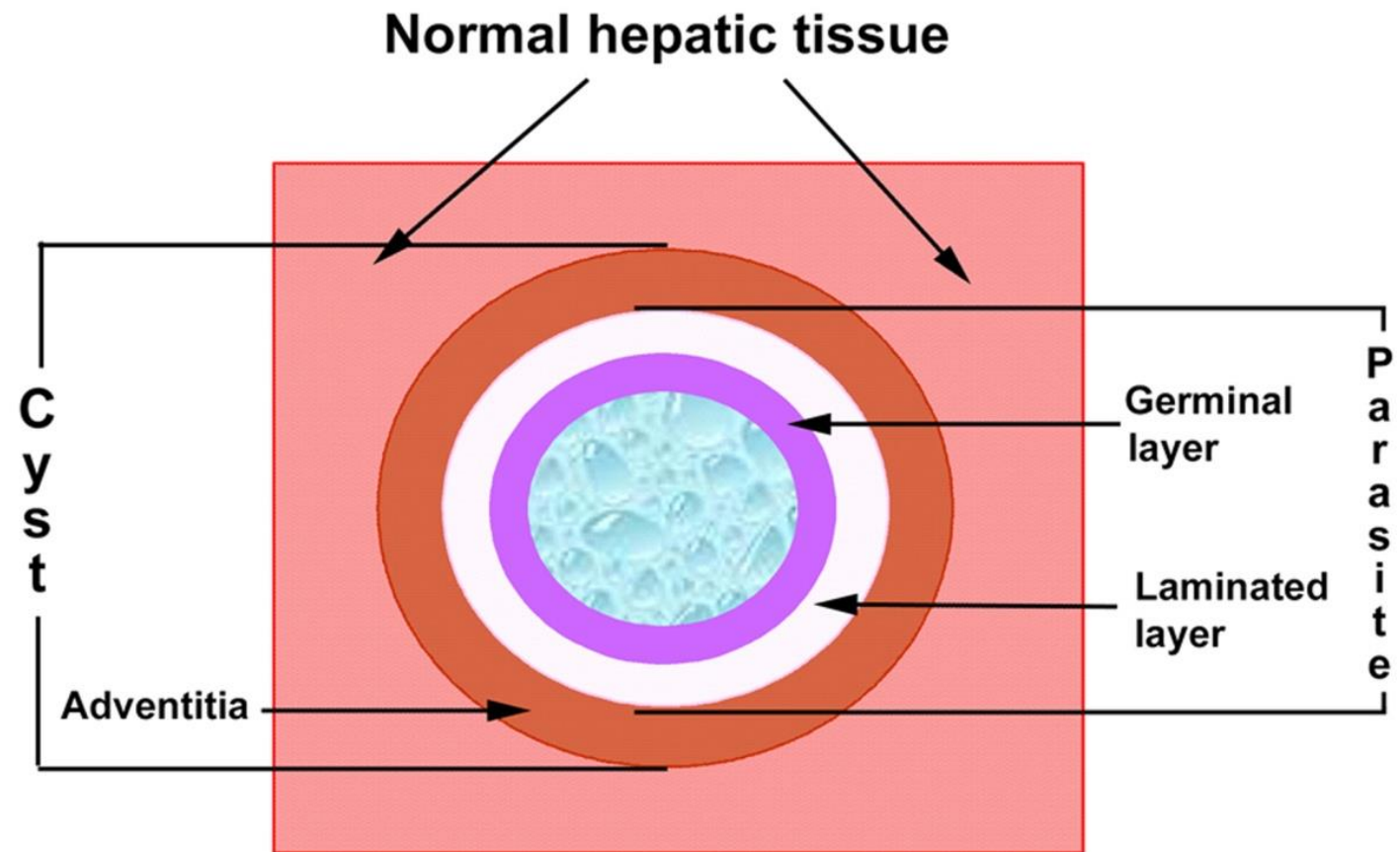
FÉLIX DÉVÉ

A **hydatid cyst** consists of two parts:

- The **echinococcal parasite**
- and
- the **adventitia** that surrounds it

The **adventitia** is a layer of inert tissue, resulting from the **reaction of the host organ** to a foreign body which is the parasite. It is this fibrous "capsule", whose thickness is variable, **which gives the term CYST to this entity**

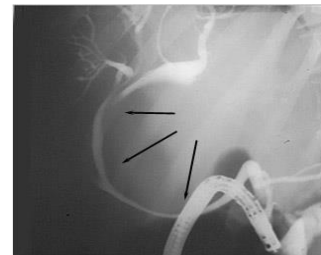
Diagram of the structure of a hydatid cyst



In treatment, it is necessary to distinguish between **complicated** and **uncomplicated** cysts

Complicated cysts

- Rupture in the bile ducts (biliary fistula)
- Rupture in the gastrointestinal tract
- Rupture in the thoracic cavity
- Rupture in the peritoneal cavity
- Compression of vessels and other organs
- Rupture in major vessels
- Primary infection
- Hemorrhage



Complicated cysts

They always have indication for treatment, whether surgical or not!
Surgery is indicated in most of these situations, but in some, particularly biliary fistulas, the **minimally invasive approach** is already the **method of choice**



Internal drainage
(endoscopic)



+



External drainage
(percutaneous)



Treatment of **uncomplicated** cysts depends on the **type, stage, location, and number**

Ultrasound classification plays an important role in **choosing the treatment method**

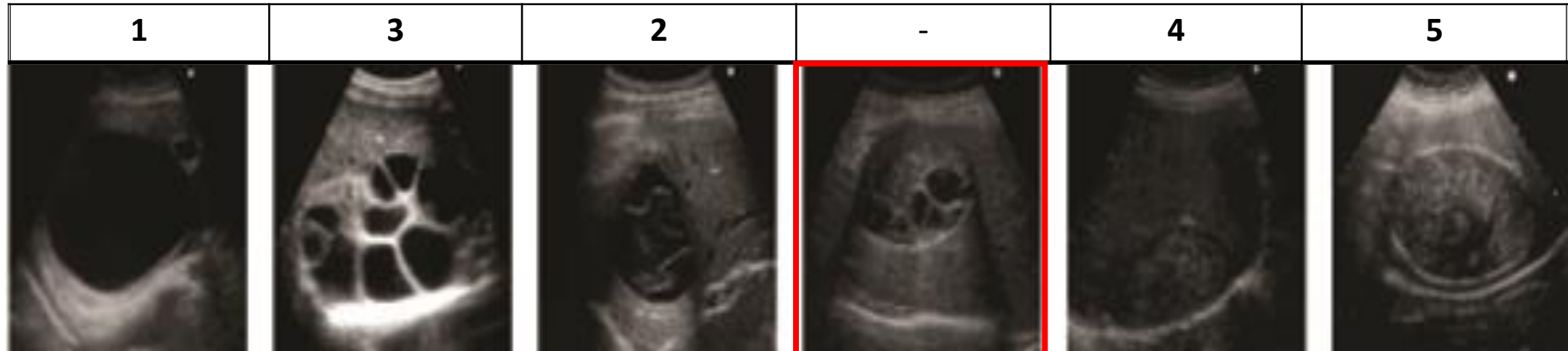
In 1980, Gharbi (Tunisia) published the **first ultrasound classification** of hydatid cysts in the liver [*Gharbi HA, Hassine W, Brauner MW, Dupuch K: Ultrasound examination of the hydatid liver. Radiology 1981*]

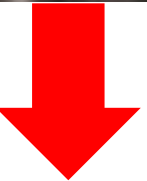

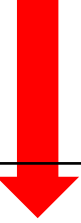
In 2001, Calum McPherson of the WHO Informal Working Group on Echinococcosis (WHO-IWGE), **under the leadership of Dominique Vuitton**, standardized the ultrasound classification [*Macpherson C. Standardization of the US classification of the Hydatid cysts of the Liver. Acta Tropica 85*].

In **2008**, it was modified to consider **two subtypes 3 (a and b)**

Ultrasound classification

Gharbi (1980)



		 			
CE 1	CE 2	CE 3		CE 4	CE 5
		3 a	(2008)	3 b	
ACTIVE		TRANSITIONAL		INACTIVE	

OMS (2001)

In **1986** Ben Amor et al. (same Tunisian team) published the results of the first trials of percutaneous puncture of hydatid cysts of the liver (PAIR), a new treatment modality: *Ben Amor N, Gargouri M, Gharbi HA, Golvan YJ, Ayachi K, Kchouck H. [Essai de traitement des kystes hydatiques abdominaux inopérables par ponction. Annales de Parasitologie Humaine et Comparée, 1986]*

The WHO-IWGE **under the leadership of Dominique Vuitton**, created a Working Group, led by Carlo Filice and Enrico Brunetti, to evaluate the efficacy and safety of transcutaneous puncture of hydatid cysts.[*Filice C, Brunetti E, Crippa F, Bruno R. Treatment of Echinococcal Abdominal Cysts. Ultrasound quarterly 1999*]. The results were presented in 1997, at the WAE Congress in Lisbon

Percutaneous cyst puncture (PAIR) was approved as a safe treatment method for liver CE

Non-invasive therapeutic approach (medical treatment) Albendazole (alone)

INDICATIONS:

- Inoperable patients
- Multiple unresectable sites
- Small cysts (less than 5 cm) stage CE1 and CE3a

THERAPEUTIC REGIMEN (400mg tablet)

- **ABZ should be administered continuously**, at an average dosage of 0-15 mg/kg/day (adjusted to the patient's weight)
- **Administration in 2 doses**, morning and evening: generally 1 tablet at 400 mg, twice a day, for an adult of average weight.

DURATION OF TREATMENT: Depends on the treatment method and the type of cyst.

Multidisciplinary care is essential for the indication, treatment monitoring, and the search for recurrences. It is important that any doctor (general practitioners and/or specialists) can manage this disease.

Invasive approaches

1. Percutaneous

Surgery is increasingly being replaced by other treatment options, **particularly percutaneous approaches**, which offer advantages and benefits for patients.

2. Surgical treatment

- ❖ Non-radical surgery (partial and subtotal cystectomy)
- ❖ Radical surgery
 - a) Total cystectomy: removal of the entire cyst (open / closed method)
 - b) Organ resection:
 - Segmentectomy
 - Lobectomy

SURGERY

In 1887, **Samuel Pozzi** performed the first cystectomy at the Lourcine-Pascal Hospital (now Broca Hospital) in Paris.



After the work of Pothrat and Baraduc in 1895, cystectomy became the preferred treatment method among Russian surgeons, where Napalkoff excelled, performing it without opening the cyst.

This method, **closed total cystectomy**, remains the ideal procedure for radical cyst treatment today, because it removes the parasite and the cyst, cures the disease and prevents recurrences

Uncomplicated Cysts

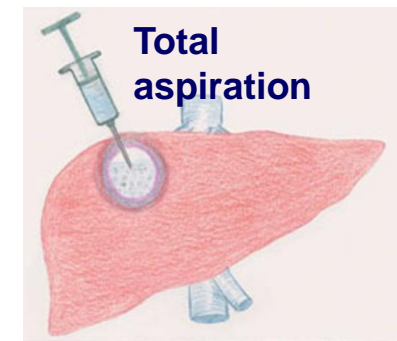
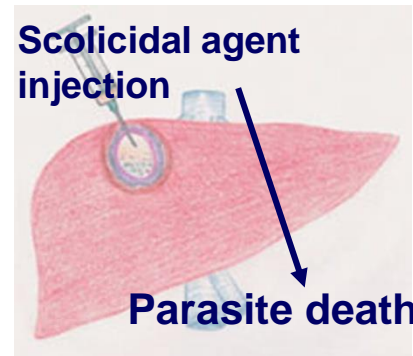
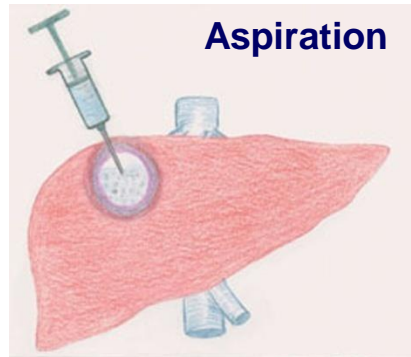
Image-based therapeutic approach criteria



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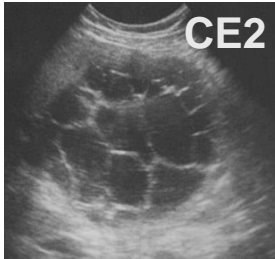


- Albendazole - Small cysts (less than 5 cm)
- Percutaneous puncture (PAIR), scolicide injection and aspiration (**do nothing other than kill the parasite**)



Uncomplicated Cysts

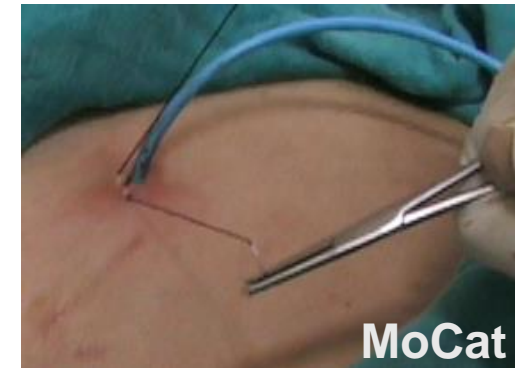
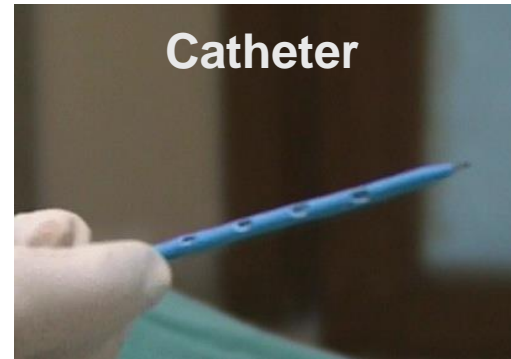
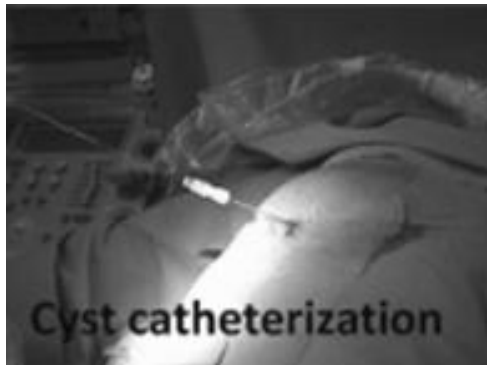
Image-based therapeutic approach criteria



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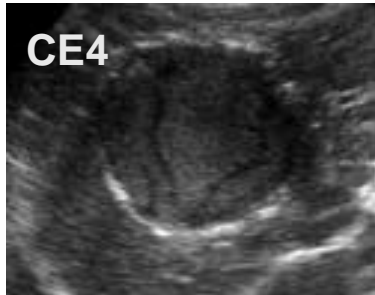


- Percutaneous puncture, scolicide injection, and **evacuation (PEvac)** / **catheterization (MoCat)**
[kills the parasite and removes the cyst contents]



Uncomplicated Cysts

Image-based therapeutic approach criteria



Non-therapeutic approach
Monitoring and waiting (watch & wait)



Albendazole (as neoadjuvant and adjuvant)

Treatment with Albendazole or another, may be necessary if there is a change in the content of the cysts.

Albendazole (as neoadjuvant and adjuvant)

A – MINIMALLY INVASIVE APPROACHES

- 1 - Duration of treatment before the intervention: **1 week**
- 2 - Duration of treatment after the intervention:

Even if there is no fluid leakage from the cyst, it is recommended to undergo therapy with Albendazole for, at least 4 weeks.

B – SURGERY

1 – SINGLE OPERABLE LESION

- Duration of treatment before the intervention: **1 week**
- Duration of treatment after the intervention:
 - a) **If the cyst could be completely removed without opening it [total cystectomy closed method], it is not recommended to continue the administration after the procedure;**
 - b) **If the cyst was opened during surgery (subtotal or partial cystectomy) or cyst ruptur occurs: 4 months of treatment is recommended**

2 – MULTIPLE LESIONS OF THE SAME ORGAN

- **Resectable lesions**

- Duration of treatment before the intervention: 1 week
- Duration of treatment after the intervention:

- **Unresectable lesions**

- a) **Cysts removed entirely: identical to the single operable lesion 1-a)**
- b) **If the cyst opens: identical to the single operable lesion 1-b)**

3 – MASSIVE INFESTATIONS (cystic lesions in multiple organs)

If the patient is operable: palliative surgery and prolonged medical treatment should be undertaken in conjunction with possible surgery

Surgery (the only method that removes the cyst)

Surgical treatment options

A O R C

p p e o
p e s m
r n e p
o i c l
a n t e
c g i e
h o n
n e
s
s

D.A. Vuitton et al.
Parasite 2020

Total cystectomy – complete resection of all (three) layers of the cyst;

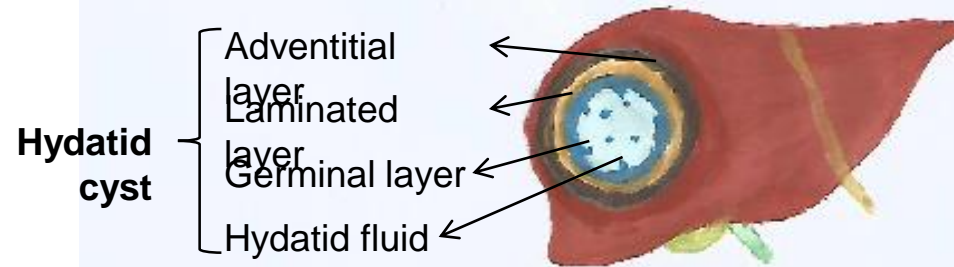
Subtotal cystectomy – almost total resection of the adventitial layer (only a fragment of the adventitia remains, for reasons of surgical safety);

Partial cystectomy – incomplete resection of the adventitial layer (part of the adventitia remains for surgical safety reasons);

Hepatectomy – en bloc resection of a part of the liver parenchyma, following the rules of hepatic resection.

AORC

Approach; **O**pening; **R**esection; **C**ompleteness



Non radical procedures

Partial cystectomy
(Removal of part of the adventitial layer)



Subtotal cystectomy
(Almost total removal of the adventitial layer)



Radical procedures

Total cystectomy
(no adventitial layer)



Organ resection
(hepatectomy)



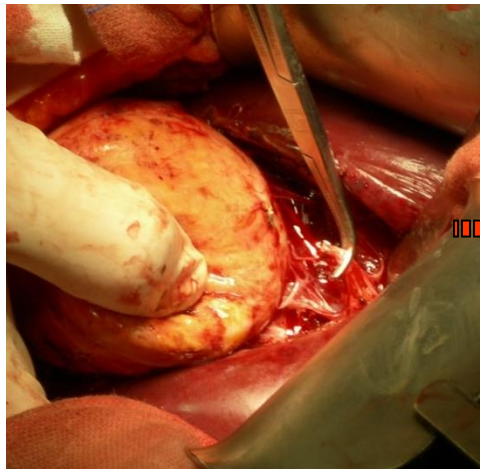
Total cystectomy – open method

(Requires absolute protection of the surgical field, against the dissemination of protoscolices due to the opening of the cyst)

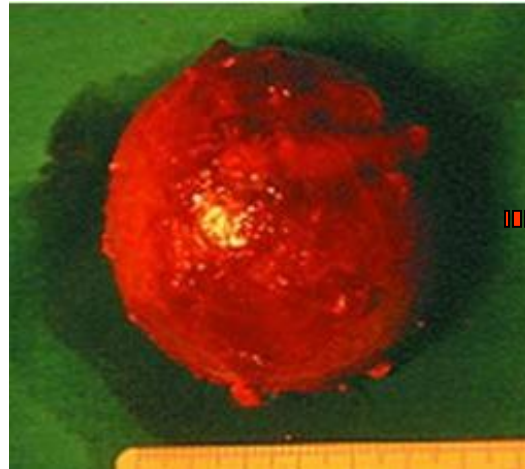


Total cystectomy – closed method (Napalkoff Operation)

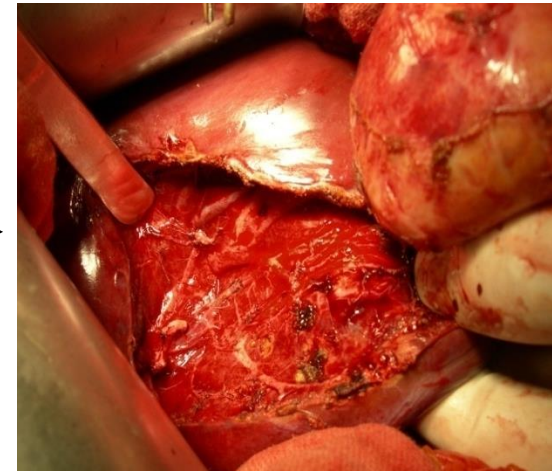
Complete resection of all (three) layers of the cyst
without opening the cyst



Beginning of the
dissection



Excision of the entire cyst



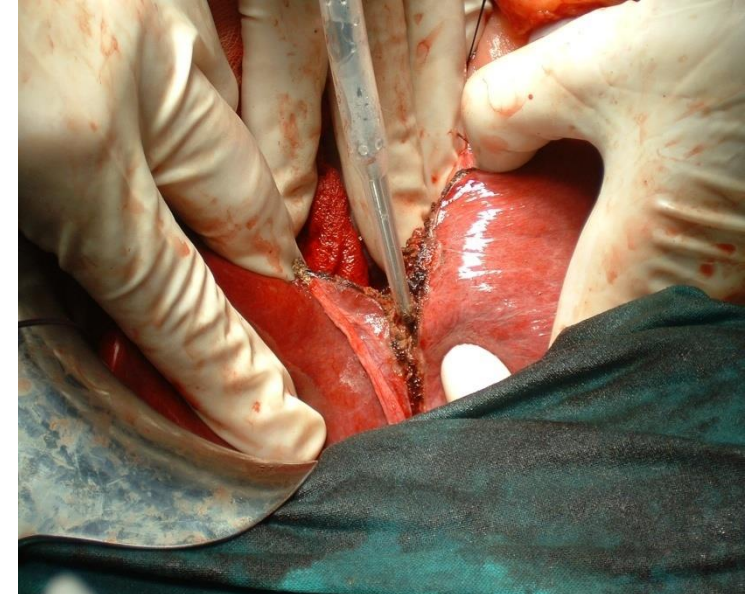
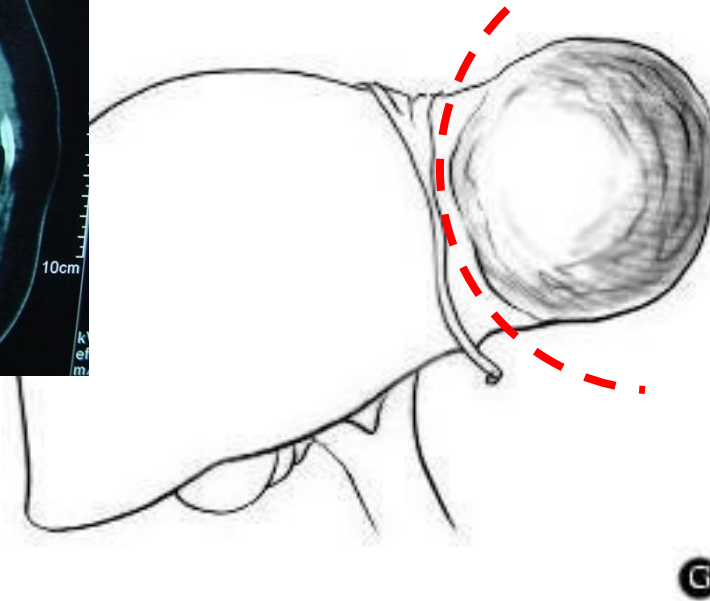
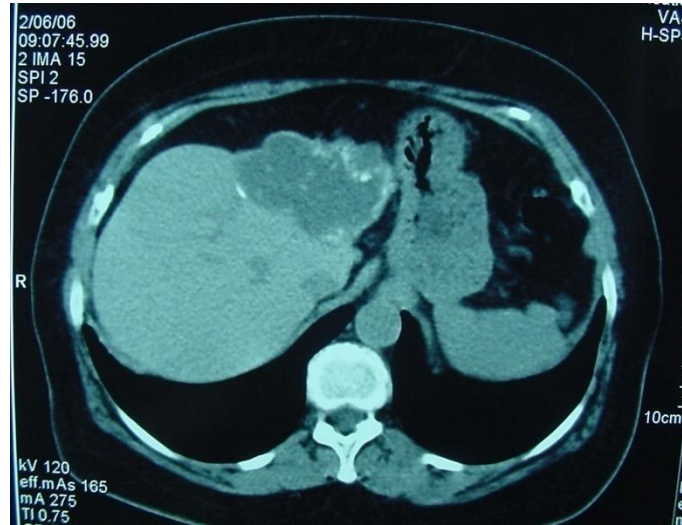
Adventitia removed
(no residual cavity)

Total cystectomy – closed method (Peng Xin Yu's Operation)



Organ Resection

Segmentectomy II/III



Following the rules of liver resection

Therapeutic proposal

Stade/type*		Proposition		Recommendation/ evidence**
Active	Type 1	< 5 cm	ABZ only/PAIR + ABZ	A / I
		> 5 cm	PAIR + ABZ	A / I
	Type 2	Surgery + ABZ		A / II
Transitional	Type 3a	< 5 cm	ABZ only/PAIR + ABZ	A / I
		> 5 cm	PAIR + ABZ	A / II
	Type 3b	PEvac/MoCat/Surgery + ABZ		B / III
Inactive (type 4 / 5)		Don't treat: Monitoring and waiting (Watch & wait)		B / III

* WHO US classification, 2001, modified in 2008, by Junghanns et al.

** Infectious Diseases Society of America grading system (strength of recommendation/quality of evidence)

CONCLUSIONS

- **Surgery is the most effective method** for treating liver CE because it removes the cyst. **Total cystectomy** is therefore the most effective method for CE, even though it can carry risks
- **Percutaneous approaches** are safe and have benefits and advantages for patients, but they only kill the parasite, potentially evacuating the cyst content
- **Albendazole** plays a very important role, not only as a first-line treatment for certain types of cysts, but also in preventing secondary echinococcosis and recurrences

Thank you for your attention



PATRIMÓNIO DA
HUMANIDADE

FADO

HERITAGE OF
HUMANITY



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