

## Echinococcophobia

Farrokh Habibzadeh,<sup>1</sup> Farrokh Saidi<sup>2</sup>

Caused by *Echinococcus granulosus*, cystic echinococcosis (CE) involves the liver in three-quarters of infested patients<sup>1</sup>. Most of the surgeons practicing round the globe, influenced by the academic training they had, believe that the mainstay of treatment for CE is pre-emptive surgery of the cyst. For instance, in Maingot's Abdominal Operations, a well-known English-American textbook of surgery, it is mentioned that "most echinococcal cysts are asymptomatic on presentation, but potential complications such as pulmonary infection, cholangitis, rupture, and anaphylaxis give good reason to consider treatment for all" and that "surgery remains the treatment of choice for uncomplicated hydatid disease of the liver"<sup>2</sup>.

In fact, there is no solid evidence in support of the pre-emptive operation of patients with asymptomatic liver CE. The current treatment policy might probably be formulated based on the adage of the Australian pioneer of hydatid disease, Harold R. Dew, who in 1928 asserted that "sooner or later, the pressure of the growing hydatid cyst causes a quiet necrosis of the wall of fairly large bile ducts and, if a large enough orifice results, some of the hydatid elements are forced into its lumen...emphasizing the necessity for operative interference"<sup>3</sup>. The Dew's dictum has been followed unconditionally by almost all surgeons ever since and no one has dared to question it<sup>4</sup>. Dew was clearly terrified of the imminent fatal complications of the release of the cyst elements into the biliary tree, and considering the limited technology available then, he might be right; however, in light of the tremendous advancements made in operative surgical techniques over the past century, what that might be considered a nightmare for Dew would be a piece of cake in the hands of the present-day surgeons.

The mortality rate associated with the pre-emptive surgery of liver hydatid is estimated at 6.5%; the overall morbidity (e.g., recurrence) rate is about 54%<sup>5</sup>. Given this unacceptable high risk of mortality and morbidity, is it wise to pre-emptively operate every person with liver CE?

<sup>1</sup>Global Virus Network, Middle East Region, Shiraz, <sup>2</sup>Department of Surgery, Modarres Hospital, Beheshti University of Medical Sciences, Tehran, Iran

**Correspondence:** Farrokh Habibzadeh.

Email: Farrokh.Habibzadeh@gmail.com **ORCID ID:** 0000-0001-5360-2900

**DOI:**10.47391/JPMA.16-23

This would be of paramount importance in regions where the disease is more prevalent. Even if no complication occurs, if the surgery is really not necessary, this common practice would impose a heavy burden on the health care system, especially in low- and middle-income countries (where the CE typically has a higher frequency). What if the cyst is not a ticking bomb? What if the widespread fear of impending fatal complications is baseless — just a phobia, *echinococcophobia*?

Echinococcosis is rare in Western Europe and North America but prevalent in South America, Asia, India, and many other regions<sup>6</sup>. A recent systematic review reveals that the overall prevalence of CE in slaughtered livestock in Iran is 14%<sup>7</sup>. In a recent study, around 10% of butchers in Pakistan were found anti-*E. granulosus* IgG seropositive<sup>8</sup>. The number of patients with liver CE visited by a surgeon practicing in the most developed countries during their whole career would typically be less than that visited annually by a surgeon practicing in the Middle East. On account of their experience, it seems that the surgeons practicing in our region should examine the best available data and try to arrive at a consensus on how to approach and treat liver CE, instead of unconditionally following a mere century-old conjecture<sup>4</sup>.

In a 20-year-long study, we prospectively followed around 300 patients with incidentally discovered asymptomatic univesicular liver CE; 127 patients with 137 cysts did accept to participate in a watch-and-wait programme<sup>9</sup>; 80% of the participants had a favourable outcome after a median follow-up of 6 years; 6% developed complications necessitating surgical operation (development of fistula and secondary infection); no mortality<sup>9</sup>. We also identified two highly diagnostic radiological markers for predicting a favourable patient outcome<sup>10</sup>. Almost 70% of the incidentally discovered liver CEs have either "laminated membrane detachment" or "pericyst degenerative changes" on unenhanced computed tomography at diagnosis<sup>9,10</sup>; presence of either of the markers is associated with a high probability of a favourable outcome without surgery (specificity of 88% and positive predictive value of 96%). These markers are highly specific but not sensitive, and their absence does not rule out a favourable outcome<sup>10</sup>.

Based on what we found, the mortality and morbidity

rates associated with a watch-and-wait approach is much less than that reported for pre-emptive surgical intervention of the cyst. We therefore believe that asymptomatic liver hydatids should no longer be operated upon, preventatively. Further efforts should be made to systematically collect more data on patients with liver hydatids in our region. To better understand the natural history of the disease and plan for a better treatment strategy, cooperation of clinicians and researchers actively involved in this field and sharing our data in a central registry/database is crucial. In the meantime, to change the current maximally radical therapeutic strategy of operating [almost] all of the liver hydatids to an ultra-conservative policy of medically management of asymptomatic liver CEs through a watch-and-wait approach, it is necessary to spread the words and raise the awareness of surgeons practicing in endemic regions. This can be done by discussing the issue and examining the available evidence in various scientific forums, say, annual national and regional surgery congresses, seminars, and meetings. The consequences of switching to such conservative non-operative treatment on reducing the number of overtreated patients and the financial burden imposed on the country health care system are undeniable, particularly in areas hyperendemic for the disease, mostly poor low-income nations with limited access to health resources. We believe that it is time to make a change, to take an action, to end the widespread *echinococophobia*.

**Disclaimer:** None.

**Conflict of Interest:** None

**Source of Funding:** None.

## References

- 1 Saidi F. Surgery of Hydatid Disease. Philadelphia: W.B. Saunders Co. Ltd, London; 1976.
- 2 Chatzizacharias NA, Christians KK, Pitt HA. Chapter 56: Hepatic Abscess and Cystic Disease of the Liver. In: Zinner MJ, Ashley SW, Hines OJ, editors. Maingot's Abdominal Operations. 12 ed. New York: McGraw-Hill; 2019.
- 3 Dew HR. Hydatid disease: its pathology, diagnosis and treatment. Sidney: The Australasian Medical Publishing Co; 1928.
- 4 Saidi F, Habibzadeh F. Unnecessary pre-emptive surgical operation of incidentally discovered liver hydatids. Clin Res Hepatol Gastroenterol .. 2018;42:e100-e1.
- 5 Daradkeh S, El-Muhtaseb H, Farah G, Sroujeh AS, Abu-Khalaf M. Predictors of morbidity and mortality in the surgical management of hydatid cyst of the liver. Langenbeck's Arch. Surg. 2007;392:35-9.
- 6 Shams M, Khazaei S, Naserifar R, Shariatzadeh SA, Anvari D, Montazeri F, et al. Global distribution of Echinococcus granulosus genotypes in domestic and wild canids: a systematic review and meta-analysis. Parasitology. 2022;149:1147-59.
- 7 Vaisi-Raygani A, Mohammadi M, Jalali R, Salari N, Hosseinian-Far M. Prevalence of cystic echinococcosis in slaughtered livestock in Iran: a systematic review and meta-analysis. BMC Infect Dis. 2021;21:429.
- 8 Alvi MA, Li L, Saqib M, Ohiolei JA, Younas MW, Tayyab MH, et al. Serologic evidence of Echinococcus granulosus in slaughterhouses in Pakistan: global alarm for butchers in developing countries. J Infect Dev Ctries. 2021;15:861-9.
- 9 Saidi F, Habibzadeh F. The Non-operative Management of Asymptomatic Liver Hydatids: Ending Echinococophobia. J. Gastrointest. Surg. : official journal of the Society for Surgery of the Alimentary Tract. 2018;22:486-95.
- 10 Habibzadeh F, Habibzadeh P, Shakibafard A, Saidi F. Predicting the outcome of asymptomatic univesicular liver hydatids: diagnostic accuracy of unenhanced CT. Eur. Radiol.. 2021;31(8):5812-7.