

Predicting HLH diagnosis and mortality in cancer

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Zoref-Lorenz A, Murakami J, Hofstetter L, Iyer S, Alotaibi AS, Mohammed SF, Miller PG, Guber E, Weinstein S, Yacobovich J, Nikiforow S, Ebert BL, Lane A, Pasvolsky O, Raanani P, Nagler A, Berliner N, Daver N, Ellis M, Jorgan MB. An improved index for diagnosis and mortality prediction in malignancy-associated hemophagocytic lymphohistiocytosis. *Blood*. 2022;139(7):1098-1110.

1. Your patient is a 62-year-old man with B-cell lymphoma and suspected hemophagocytic lymphohistiocytosis (HLH). According to the international cohort study by Zoref-Lorenz and colleagues, which of the following statements about clinical, diagnostic, and prognostic parameters and optimized laboratory cutoff values in patients with hematologic malignancy (HM)-associated hemophagocytic lymphohistiocytosis (HM-HLH) compared with patients with only HMs is correct?

- Patients with or without HLH differed significantly in distribution of underlying HMs
- Patients with HM-HLH were significantly older than patients with HMs
- Most HLH-2004 parameters differed between patients with HMs and patients with HM-HLH
- The only significantly different laboratory parameters between groups that were above diagnostic thresholds were the inflammatory markers soluble CD25 (sCD25) and ferritin

2. According to the international cohort study by Zoref-Lorenz and colleagues, which of the following statements about component features and diagnostic performance of the optimized HLH inflammatory (OHI) index is correct?

- OHI, or combined elevation of sCD25 (>3900 U/mL) and ferritin (>1000 ng/mL), best identified HLH-2004 defining features (sensitivity 84%, specificity 81%)
- The OHI index was not predictive of mortality
- The OHI index was not useful for routine surveillance of patients with newly diagnosed HMs
- Among patients discordantly classified by OHI vs other indices, the OHI index was less effective in recognizing high-risk patients

3. According to the international cohort study by Zoref-Lorenz and colleagues, which of the following statements about clinical implications of the diagnostic performance of the OHI index is correct?

- Using the OHI index is unlikely to affect patient outcomes
- Applying optimized thresholds for sCD25 and ferritin to the HLH-2004 resulted in a better diagnostic and prognostic tool than the OHI index
- The OHI index identifies patients with HM with an inflammatory state and is associated with high-mortality risk, warranting further prospective validation
- sCD25 and ferritin should be measured at diagnosis of various HMs only if HLH is clinically suspected