

Malignancy-Associated Hemophagocytic Lymphohistiocytosis

in a Low-Middle Income Country, Egypt

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BACKGROUND

Hemophagocytic lymphohistiocytosis (HLH) is a severe hyperinflammatory syndrome that may complicate malignancy.

Diagnosing malignancy-associated HLH (MA-HLH) is difficult because some HLH 2016 criteria overlap with manifestations of underlying malignancy. Early recognition is essential, especially in rapidly progressive cases.



AIM

To describe the clinical presentation, laboratory profile, treatment strategies, and outcomes of children with malignancy-associated HLH treated at a single tertiary center in Egypt.



METHODS

- Prospective single-center study.
- Included children younger than 15 years with HLH and underlying malignancy treated between 2010 and 2023.
- Forty-seven patients were identified, 5 with incomplete records were excluded.
- Minimal follow-up was six months.
- Collected data was included demographic, underlying malignancy, HLH diagnostic features, timing, solid tumor, BCS, triglyceride, fibrinogen, treatment, response, and survival.



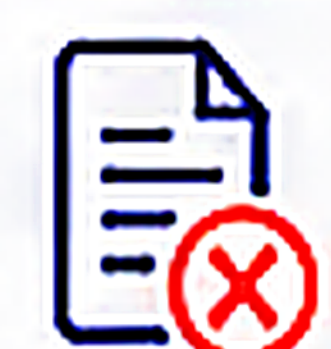
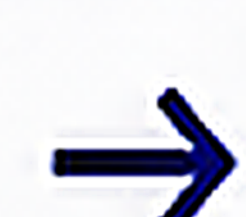
ACKNOWLEDGMENTS

MA-HLH in Oncology team, Ain Shams University and all family members of children with HLH and their caregivers.

RESULTS



47 screened

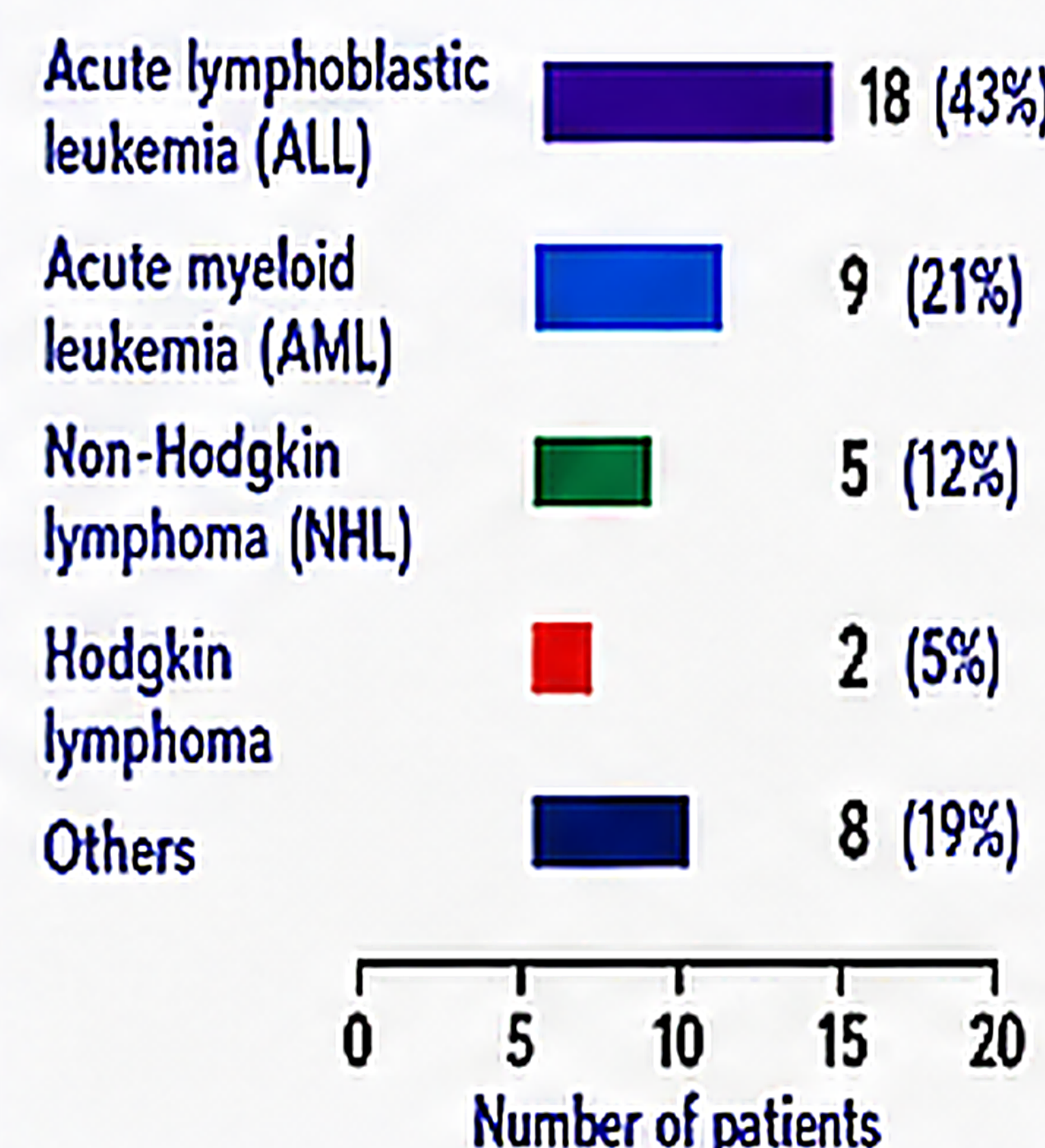


5 excluded
(incomplete clinical data)

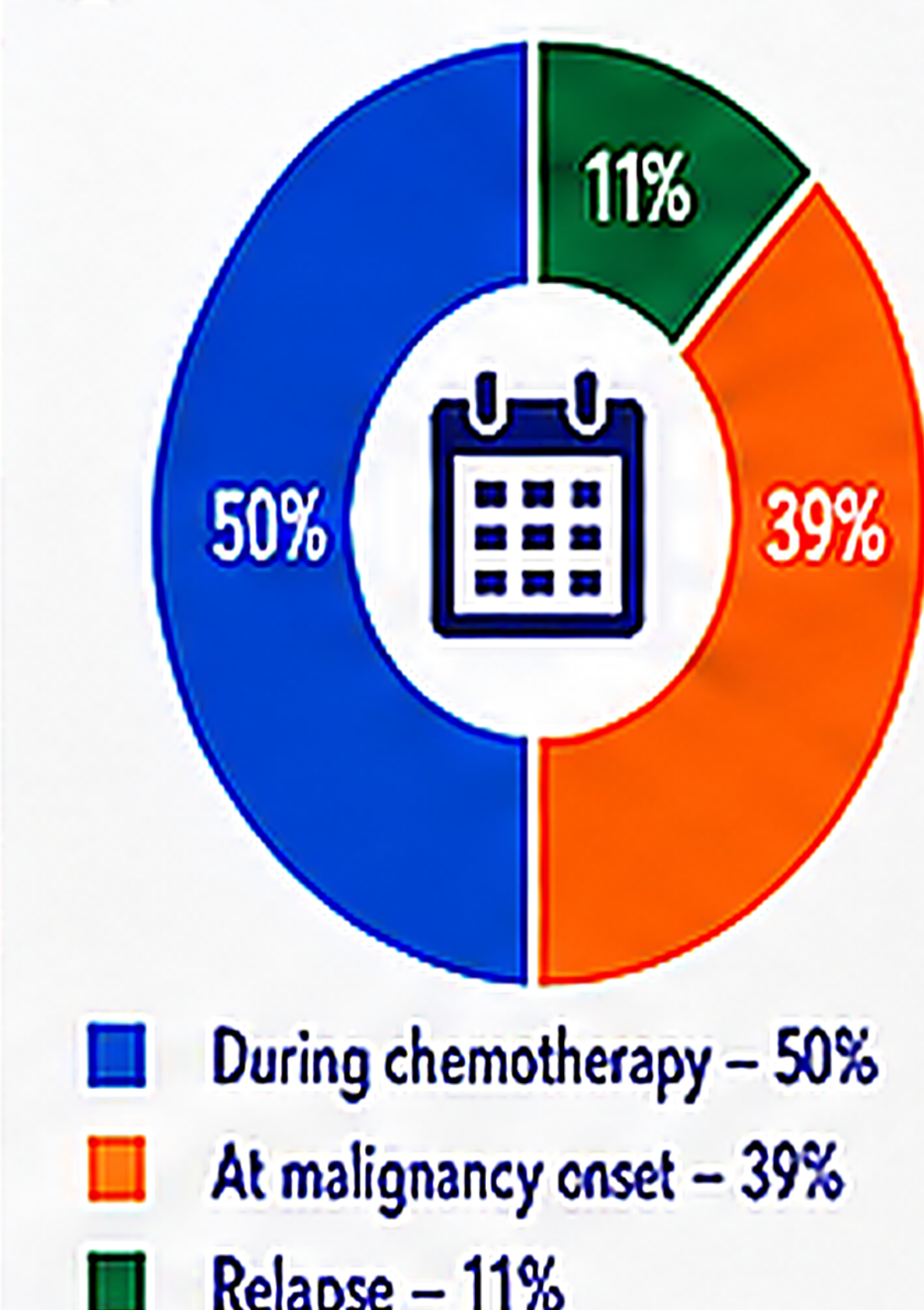


42 analyzed

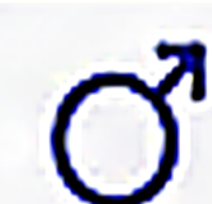
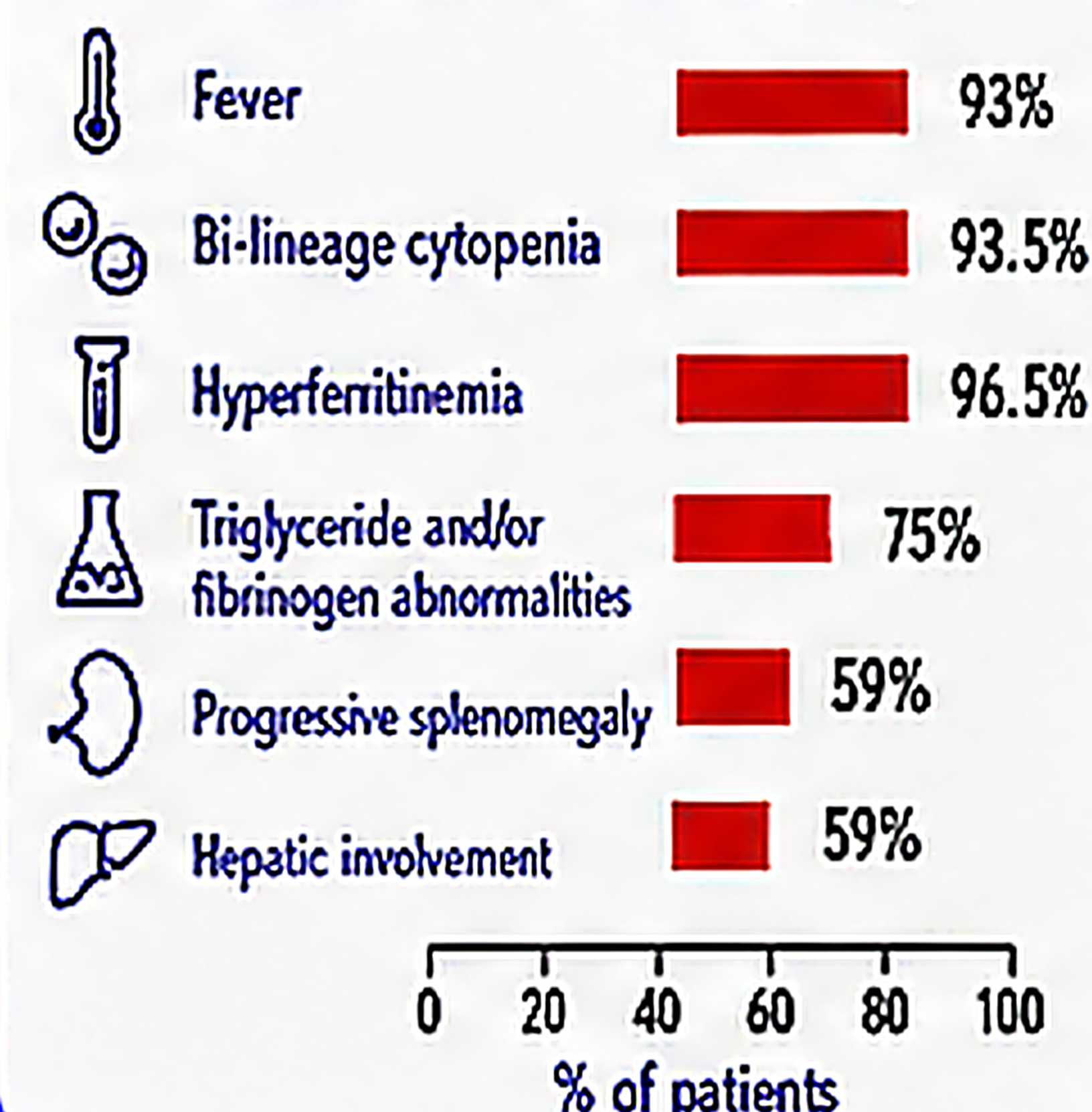
A UNDERLYING MALIGNANCIES (n=42)



B TIMING OF HLH ONSET (n=42)



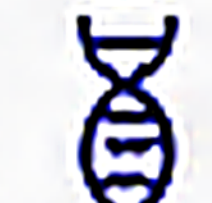
C CLINICAL AND LABORATORY FEATURES AT DIAGNOSIS (n=42)



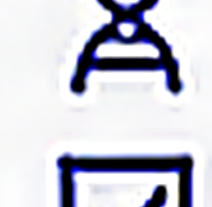
Median age at HLH diagnosis: 87 months.



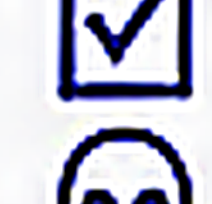
Male sex: 60%.



Median interval between malignancy diagnosis/relapse and HLH onset: 36 days (IQR, 5–101).



Thirty-eight percent underwent genetic testing; half had monogenic variants in HLH-related genes, including one GATA2 mutation.



Eighteen percent fulfilled 3 HLH criteria, while 60% fulfilled 4–5 criteria.



Mortality was higher in younger age groups and in patients with more positive HLH criteria.

KEY FINDINGS

42 analyzed

60% male

60% response rate

38% mortality

60% 2-year overall survival



CONTACT

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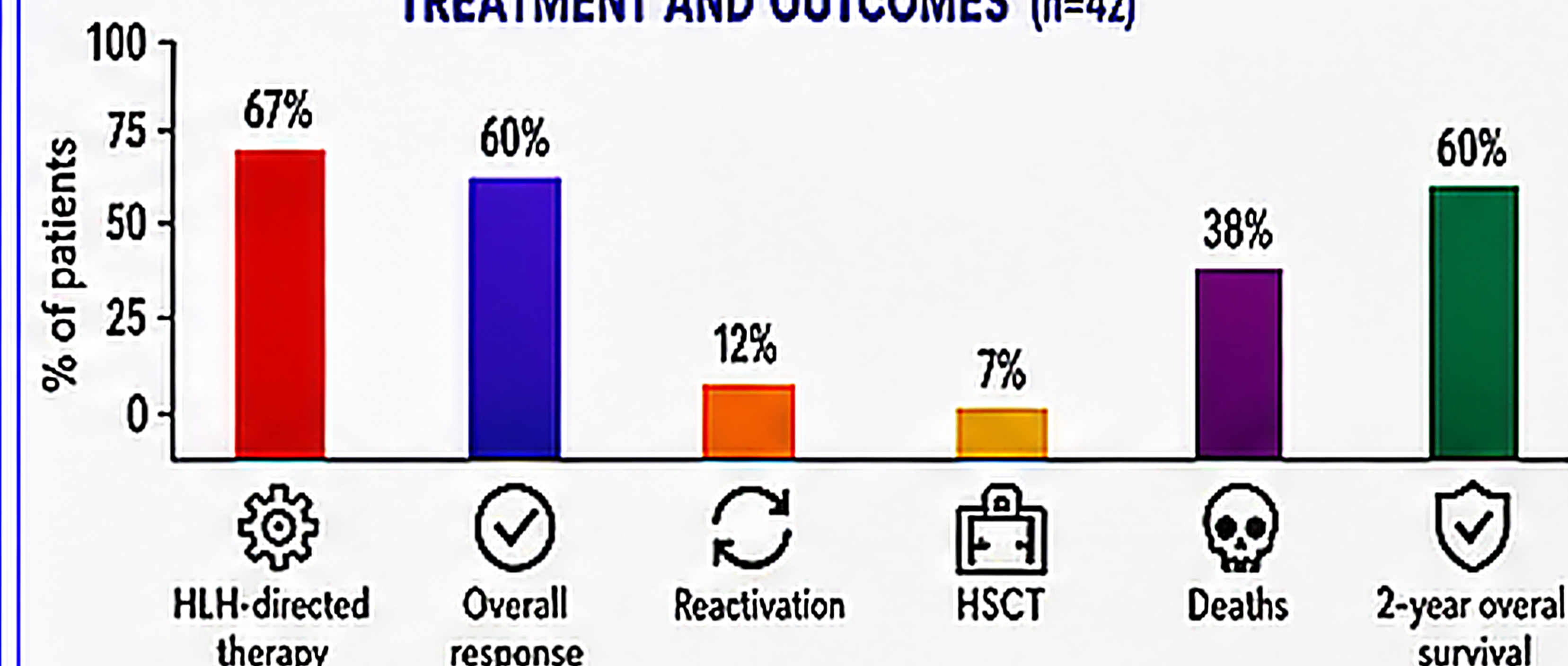
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REFERENCE

Zan-Urbanets E A, et al. *Blood*. 2022;139(7):1088-1111.
doi: 10.1182/blood.2021013140.

TREATMENT AND OUTCOMES (n=42)



- Two-thirds of patients received HLH-directed therapy.
- Etoposide was used in 12 patients, anti-IL-1 α , and corticosteroids in 60%.
- Overall response rate was 60%.
- HLH reactivation occurred in 12.7% of patients with HSCT.
- Median follow-up was 22 months (IQR, 6–84).
- Overall mortality was 38%. 5.7% of deaths were attributed to the underlying malignancy, and 7% attributed to HLH.
- All 8 patients who died from HLH had hepatic involvement and a rapid disease course; median HLH diagnosis to death was 29 days.



CONCLUSIONS

MA-HLH in children carries substantial mortality and often develops during active cancer treatment. Hepatic involvement identified, fulminant subgroup with particularly poor outcomes. Earlier recognition and treatment appear to have improved outcomes in the more recent few years.



CLINICAL MESSAGE

In a child with malignancy, the combination of fever, cytopenia, hyperferritinemia, and hepatic involvement should prompt urgent evaluation for MA-HLH.