

International Prognostic Scoring System for Mastocytosis

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Abstract

The International Prognostic Scoring System for Mastocytosis (IPSM) risk score is prognostic of survival outcomes for patients with non-advanced and advanced systemic mastocytosis. The IPSM score for non-advanced systemic mastocytosis is based on age and alkaline phosphatase level while the IPSM score for advanced systemic mastocytosis is based on age, skin involvement, and tryptase, leukocyte, hemoglobin, and platelet levels.

I. HOW TO USE

When to Use

This prognostic tool is relevant for patients with both non-advanced and advanced systemic mastocytosis.

Pearls / Pitfalls

The validation study excluded patients with mast cell sarcoma and children (< 17 years) with cutaneous mastocytosis. The score did not evaluate nonroutine laboratory values such as β 2-microglobulin given the retrospective nature.

Why Use

This simple tool has strong prognostic implications based on age, laboratory values, and skin involvement. It can be incorporated into clinic discussions to potentially assist in treatment decisions.

II. NEXT STEPS

Advice

This tool should not be the sole basis for care, and clinicians should use judgment and a full clinical evaluation. The score could be used to contextualize WHO classification and aid in treatment discussions.

Management

The validation study did not lead to any specific management recommendations but instead to consider the risk

stratification when balancing tradeoffs between curative potential and side effects of treatment options.

Critical Actions

Management of systemic mastocytosis should be guided by clinician judgment, regardless of the IPSM score.

III. EVIDENCE

Evidence Appraisal

The [validation study](#) that created the IPSM confirmed the prognostic value of the WHO classification of mastocytosis into distinct variants. A total of 1639 patients were analyzed to identify variables predictive of survival that were included in the score and 462 patients to validate the IPSM. Prognostic variables were included if the hazard ratio was 1.50 or higher (+1) or 0.50 or lower (-1). IPSM scores were predictive of overall survival, progression-free survival, and event-free survival (all $p < 0.0001$).

A [follow-up study](#) further stratified the advanced systemic mastocytosis (AdvSM) IPSM risk score by using serum tryptase, alkaline phosphatase, β 2 microglobulin, lactate dehydrogenase, and albumin.

Formula

Non-Advanced Systemic Mastocytosis

Age: 60+ yo (+1) vs < 60 yo (0)

Alkaline Phosphatase: 100+ IU/L (+1) vs < 100 IU/L (0)

Risk Level = low (0), intermediate-1 (1), and intermediate-2 (2)

Advanced Systemic Mastocytosis (AdvSM)

Age: 60+ yo (+1) vs < 60 yo (0)

Tryptase: 125+ ng/mL (+1) vs < 125 (0)

Leukocyte: $16 \times 10^9/L$ (+1) vs $< 16 \times 10^9/L$ (0)Hemoglobin: ≤ 11 g/dL (+1) vs > 11 g/dL (0)Platelet: $\leq 100 \times 10^9/L$ (+1) vs $> 100 \times 10^9/L$ (0)

Skin Involvement: yes (-1) vs no (0)

Risk Level = AdvSM-1 (-1 to 0), AdvSM-2 (1), AdvSM-3 (2-3), AdvSM-4 (4-5)

Facts & Figures

10-year overall survival was 98.1% for low-risk non-advanced systemic mastocytosis, 87.1% for intermediate-1 risk, and 52.1% for intermediate-2 risk level, and 22.0% for advanced systemic mastocytosis (AdvSM). 10-year progression-free survival was 96.3%, 86.7%, 76.3%, and 61.1%, respectively. AdvSM risk groupings were prognos-

tic for overall survival, progression-free survival, and event-free survival ($p < 0.0001$).

Literature*Original/Primary & Validation*

Sperr WR et al. International prognostic scoring system for mastocytosis (IPSM): a retrospective cohort study. *Lancet Haematol.* 2019;6(12):e638-e649. PMID 31676322.

Other References (including meta-analyses, CPGs, and impact analyses)

Lübke J et al. Serum chemistry profiling and prognostication in systemic mastocytosis: a registry-based study of the ECNM and GREM. *Blood Adv.* 2024;8(11):2890-2900. PMID 38593217.