

# Mastocytosis presenting with mast cell mediatory release-associated symptoms elicited by COX inhibitors: prevalence, clinical and laboratory features.

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## BACKGROUND

Nonsteroidal anti-inflammatory drugs (NSAIDs) and other cyclooxygenase inhibitors (COXi) are frequently **avoided in mastocytosis, out of concern for drug hypersensitivity reactions (DHR).**

**Objectives:** to determine the prevalence of DHR to NSAIDs and associated clinical/laboratory findings in mastocytosis patients (pts).

## METHODS

Retrospective review of medical records from **418 ≥ 18 y/o** and **223 < 18 y/o** mastocytosis pts.

**Exclusion criteria:** insufficient clinical data; not having received COXi other than paracetamol following onset (**36 ≥ 18 y/o** and **54 <18 y/o** pts).

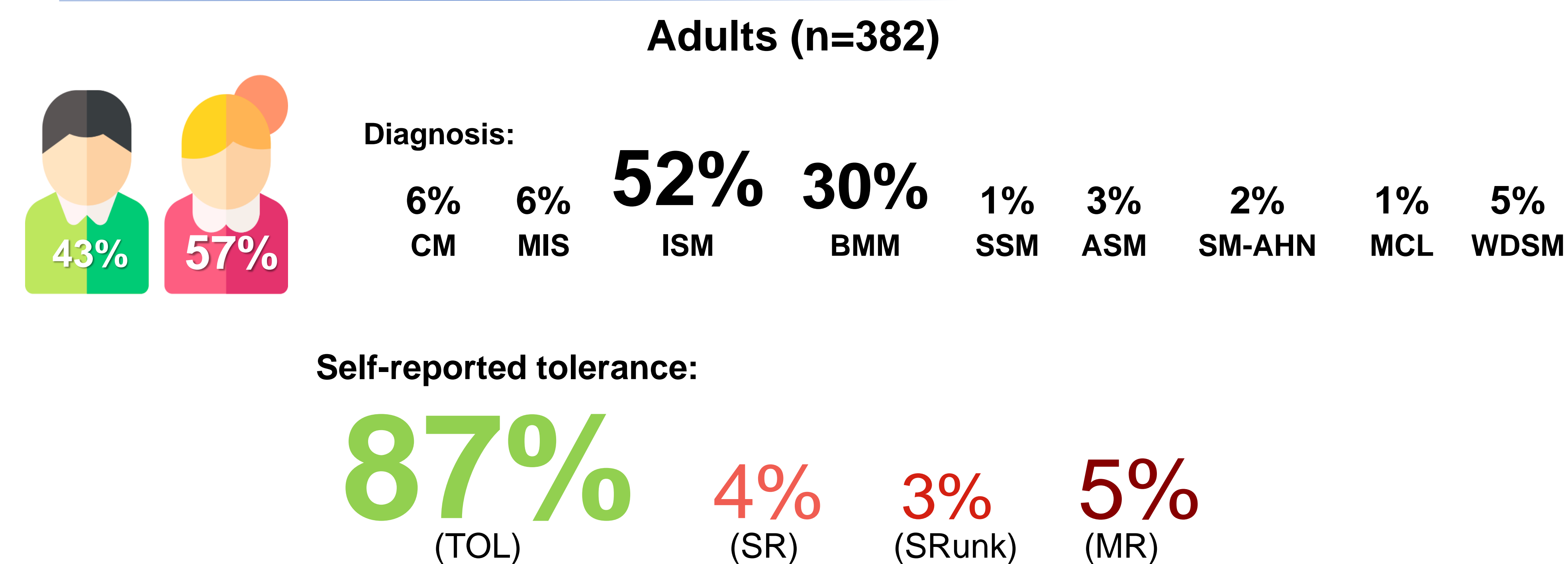
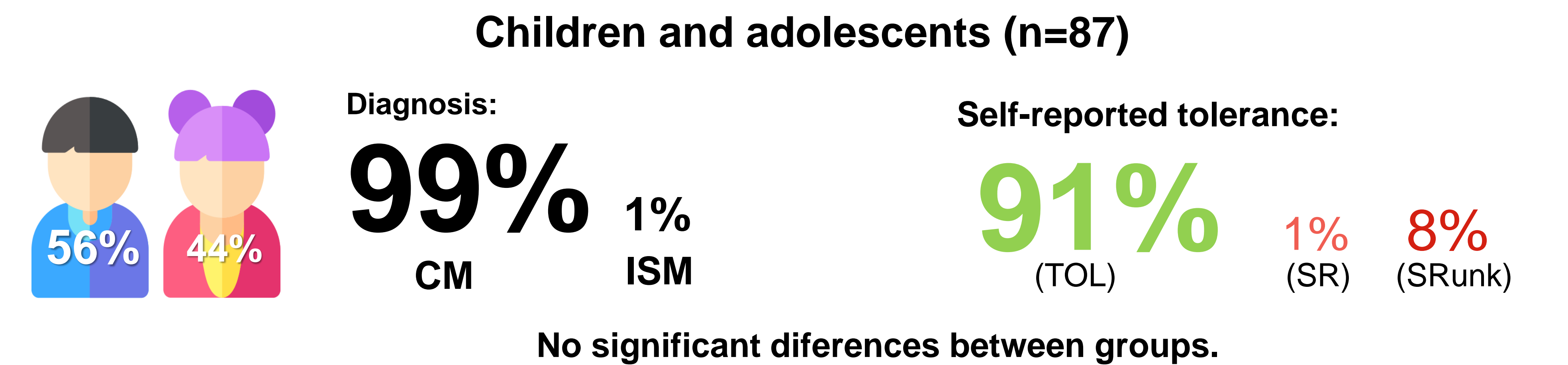
**Grouping:** **tolerants (Tol)**, **single reactors (SR)**, **SR with unknown tolerance to COXi other than paracetamol (SRunk)**, **multiple reactors (MR).**

Comparison of groups for epidemiological, clinical, laboratorial and imagiological variables.

**Two models** (using significant variables from the univariate study): 1) identification of pts with **DHR to ≥2 NSAIDs/other COXi vs NSAID tolerants**; 2) identification of **tolerant pts vs those with DHR to ≥1 NSAIDs/other COXi.**

**Decision tree for the prescription of NSAIDs/COXi other than paracetamol in mastocytosis.**

## RESULTS



Variable	Score
<i>Gender</i>	
Female	+6
Male	-2
<i>Manifestations</i>	
Flushing	+4
Absence of pruritus	-3
Skin lesions	-3
Anaphylaxis not caused by NSAIDs/COXi	+5
Anaphylaxis caused by HVA	-4
<i>Laboratory findings</i>	
Multilineal <i>KIT</i> D816V mutation*	+4
sBT ≥ 48 ng/mL	+6

Figure 1. Score model for screening of DHR to multiple NSAIDs/COXi. \*If unknown, count as negative

**Score ≥ 7:**  
High probability of DHR to NSAIDs/COXi

Sensitivity	Specificity
90%	71%
Positive Predictive Value	Negative Predictive Value
14%	99%

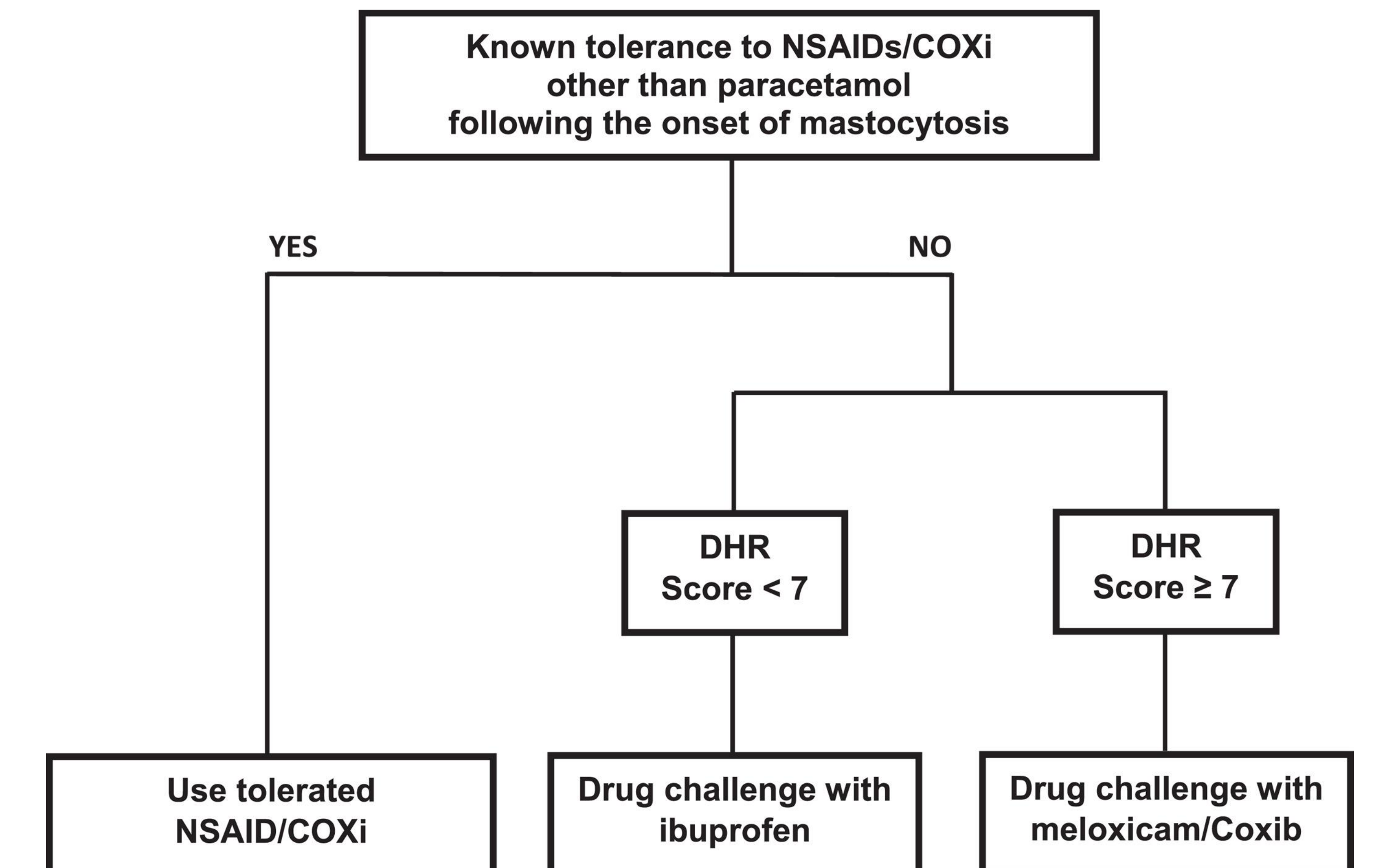


Figure 2. Decision-tree algorithm for adult mastocytosis patients at risk of DHR to NSAIDs/COXi other than paracetamol.

## CONCLUSIONS

DHR to NSAIDs/COXi other than paracetamol are **more frequent in mastocytosis patients** vs. the general population but **less frequent than previously estimated**, being **associated with unique disease features.**

Pts that **tolerated NSAIDs/COXi other than paracetamol following onset of disease should keep using those specifically.** Those with **unknown tolerance and adults with a positive score should be challenged with a preferential/selective COX-2 inhibitor**, while the remaining may be challenged with **ibuprofen.**

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