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Title of Presentation	Clinical features of hereditary alpha tryptasemia and its role in anaphylaxis
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Abstract of Presentation	Introduction Hereditary alpha-tryptasemia (HAT) is increasingly recognized in patients with elevated baseline serum tryptase, but its clinical significance, particularly regarding mast cell-related symptoms and anaphylaxis, remains uncertain. In this study, we present the clinical features of HAT patients in a tertiary referral cohort. Methods We included 109 patients referred to the Maastricht University Medical Center for evaluation of mast cell related complaints where HAT was suspected and tested for. HAT was identified through DNA-based analysis in blood, while mastocytosis (Masto) was identified according to the diagnostic criteria. Results were classified in two categories: [pos] = disorder present and [neg] disorder absent. Tryptase levels were measured using the ImmunoCAP assay. Clinical data were collected using a standardized form and anaphylaxis graded according to the Müller grading system. Statistical analyses were performed using SPSS. Results Patients were classified in 4 groups: HAT[neg]Masto[neg] (N=47), HAT[neg]Masto[pos] (N=21), HAT[pos]Masto[neg] (N=36), HAT[pos]Masto[pos] (N=5). Mean tryptase levels were 5, 19, 14 and 50 µg/L, respectively. Patients in the HAT[pos]Masto[neg] group were significantly younger (49 vs. 59 overall, p=0.021). HAT[pos]Masto[neg] patients did not

	have anaphylaxis more often than HAT[neg]Masto[neg] patients and the severity of anaphylaxis (Müller grading 3 or 4) was not higher in the HAT[pos]Masto[neg-] group compared to the HAT[neg]Masto[neg] group and lower than in the mastocytosis patients. Conclusion HAT was not associated with a distinct clinical phenotype or an increased frequency or severity of anaphylaxis. These findings suggest that HAT alone has limited value in predicting the clinical presentation of patients with mast cell-related symptoms.