Characteristic skin lesions (mastocytosis in the skin)

**INDICATIONS FOR TESTING**
- Symptoms consistent with mast cell activation
- Flushing, loose stools or diarrhea, anaphylaxis
- No identifiable secondary cause of mast cell activation

Proceed to bone marrow aspiration and biopsy

**ORDER**
- Serum tryptase AND KIT D816V Mutational Analysis by PCR in Peripheral Blood

**Adult**
- Proceed to bone marrow aspirate and biopsy to rule out systemic mastocytosis\(^a\)

**Child**
- No further testing, unless symptoms suggest systemic disease

**Tryptase ≥15 ng/mL KIT D816V positive**
- Proceed to bone marrow aspiration and biopsy\(^a\)

**Tryptase <15 ng/mL KIT D816V positive**
- No further testing, unless symptoms suggest systemic disease

**Tryptase ≥15 ng/mL KIT D816V negative**
- Consider alternative diagnosis besides systemic mastocytosis (eg, mast cell activation syndrome)

**Tryptase <15 ng/mL KIT D816V negative**
- Monitor tryptase

\(^a\)Patients may be negative for KIT D816V mutation and still have systemic mastocytosis; consider full KIT gene sequencing

\(^b\)High-sensitivity assay for detection of KIT D816V mutation (eg, allele-specific polymerase chain reaction [PCR]) recommended