Name:

DOB:

Explaining Mast Cell Diseases

Mastocytosis: A condition in which the bone marrow produces too many mast cells. The overproduction of mast cells can lead to their degranulation.

- Patients have chronically elevated tryptase level--tryptase is an enzyme released by mast cells when they are degranulating
- Disease presentation is dependent on the organs impacted

Non-Advanced Subtypes

- Indolent Systemic Mastocytosis
- Smoldering Systemic Mastocytosis

Advanced Subtypes

- Systemic Mastocytosis with Associated Hematologic Neoplasm
- Aggressive Systemic Mastocytosis
- Mast Cell Leukemia
- Mast Cell Sarcoma

Advanced subtypes generally progress more rapidly. Non-advanced subtypes can progress to advanced. Subtype can be dynamic.

Cutaneous mastocytosis only affects the skin

Mast Cell Activation Syndrome (MCAS): A condition in which the body has the right number of mast cells but the mast cells are overly active which can lead to allergic reactions.

- Normal baseline tryptase level

I have mastocytosis/MCAS (circle one)

How were you diagnosed and who diagnosed you?

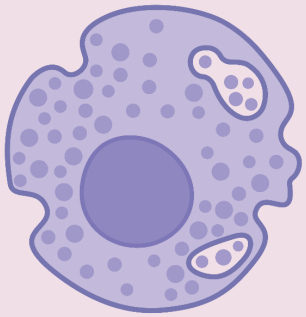
IgE and Non-IgE Anaphylaxis

Anaphylaxis

IgE-Mediated

Effector Cells:

- Mast Cells
- Basophils



Mast cells are universally recognized in all forms of anaphylaxis

Non IgE-Mediated

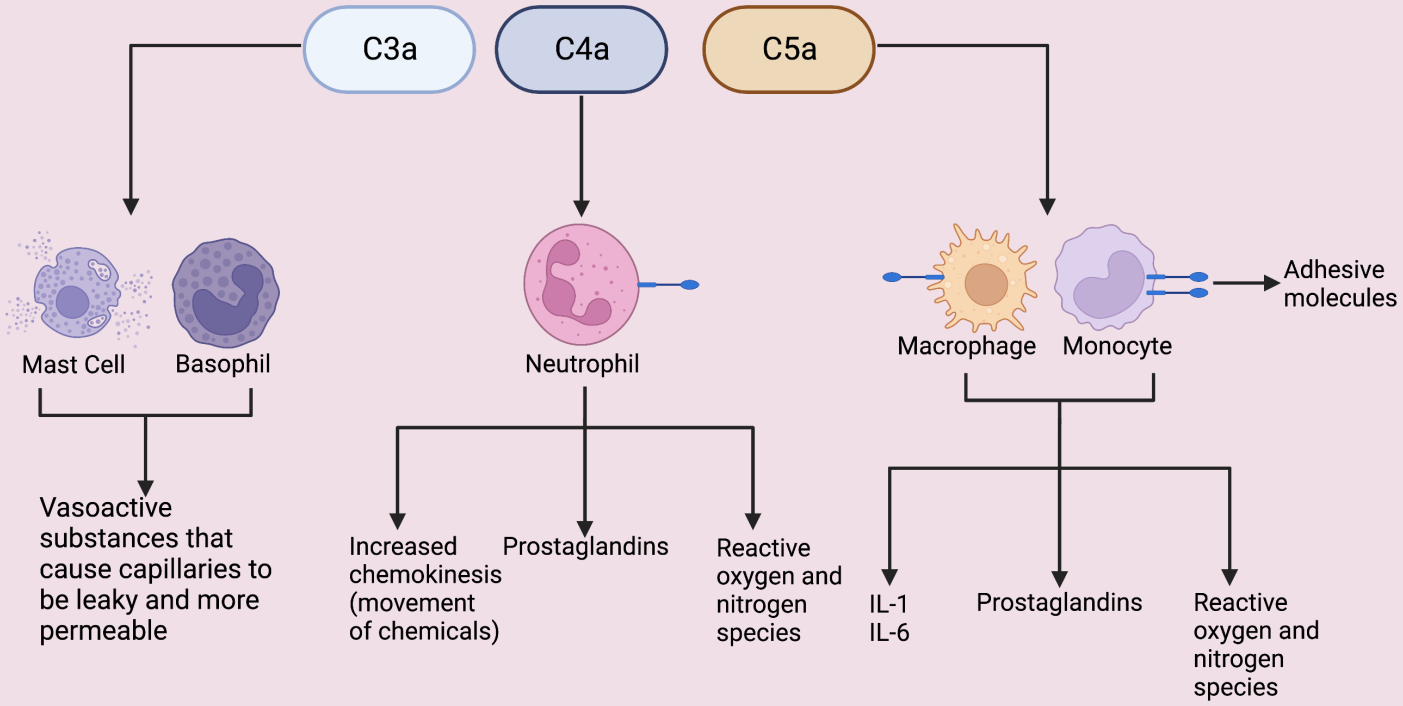
Activation Pathways

- IgG
- Complement
- Bradykinin
- Leukotrienes
- Prostaglandins

Effector Cells

- Neutrophils
- Endothelial Cells
- Smooth Muscle
- Mast Cells
- Basophils
- Eosinophils

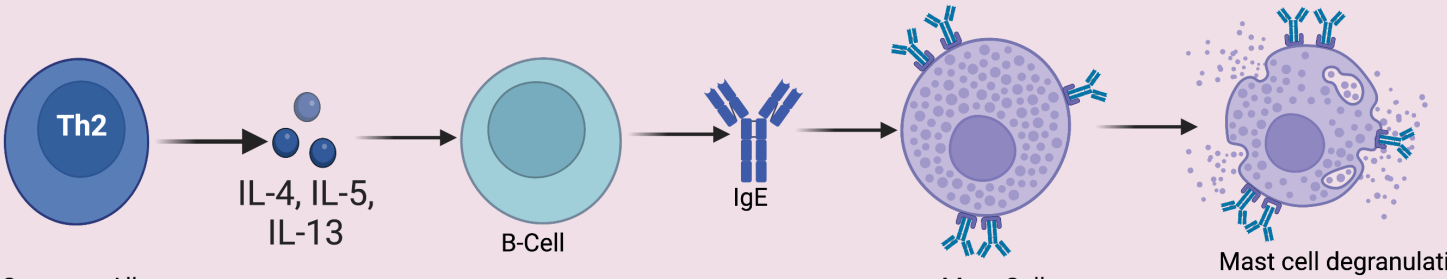
Anaphylatoxin is a substance derived from complement activation that causes smooth muscle contraction, capillary leakage, and, in some cases, anaphylactic shock. C3a, C4a, and C5a are considered anaphylatoxins.



The complement system is an inflammatory pathway that is one of the main contributors to non-IgE anaphylaxis

IgE-Mediated Anaphylaxis

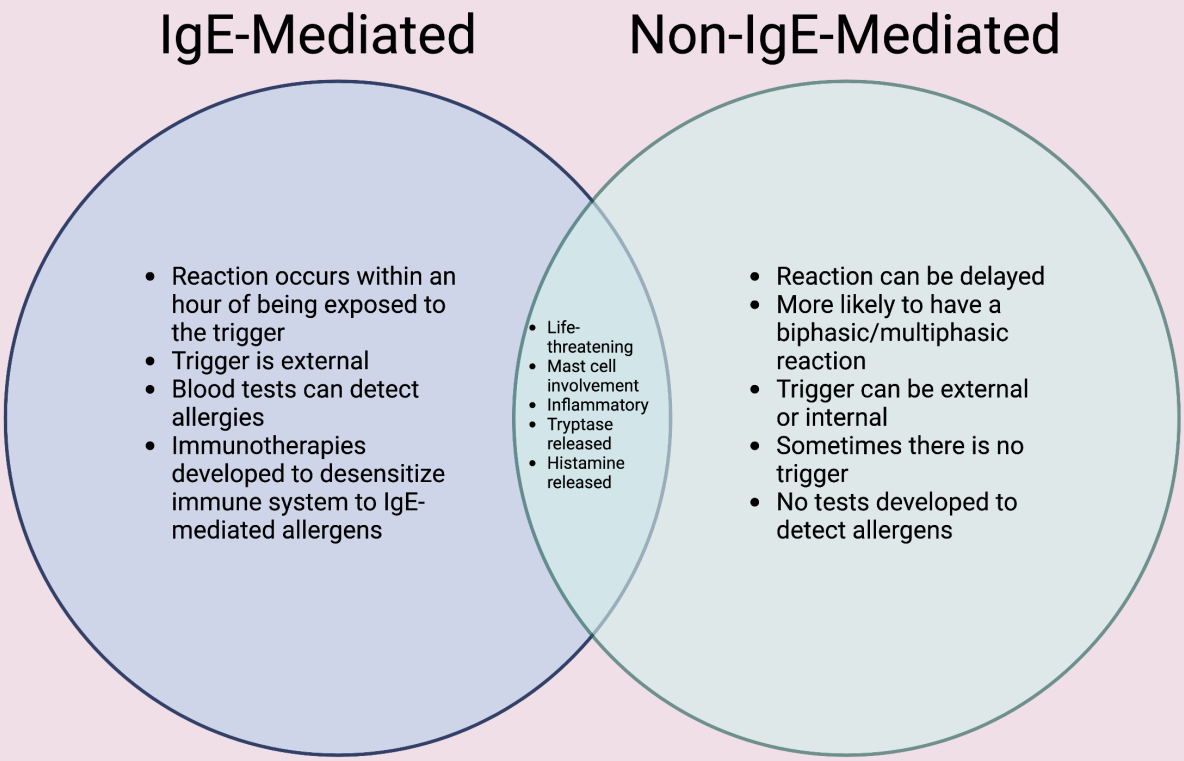
- Classical allergies to foods are IgE mediated
- Getting allergy tests (blood or skin) only tests for IgE-mediated allergies
- IgE-mediated allergies are better understood (as they were discovered first so we have years of knowledge) and there are medications (e.g. omalizumab) that targets the IgE complex to prevent allergic reactions



Common Allergens:

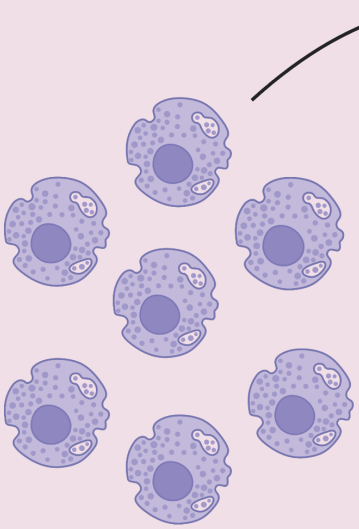
- Peanuts
- Tree nuts
- Sesame
- Shellfish
- Dairy
- Eggs
- Soy
- Fish
- Wheat

- Bee stings
- Venom
- Penicillin
- Sulfa antibiotics
- Latex
- Animal dander

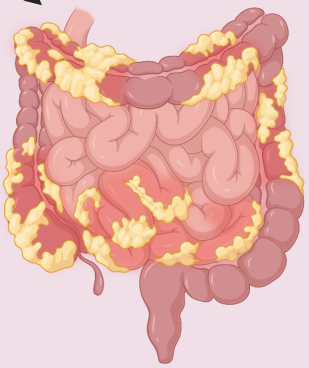


Some conditions like atopic dermatitis/eczema and eosinophilic esophagitis can be both IgE and non-Ige mediated.

Mast Cells and Intestinal Inflammation: A Perfect Storm



Mast cells build up in the gut and cause inflammation, especially upon degranulation



Inflammation in the gut leads to mast cell degranulation

This is why some cases of inflammatory bowel disease can have an anaphylactic component, especially if the patient already has a mast cell disorder

Mast cells (and eosinophils) can mimic strictures (narrowing of the intestine due to inflammation) in terms of symptoms and on imaging

Sources

- https://www.researchgate.net/figure/Biological-functions-of-the-complement-system-Inflammation-the-activation-of-the_fig2_272081321
- https://www.sciencedirect.com/topics/immunology-and-microbiology/anaphylatoxin#:~:text=Anaphylatoxin%20is%20a%20substance%20derived,leakage%2C%20and%20even%20anaphylactic%20shock.
- https://www.rch.org.au/clinicalguide/guideline_index/Food_allergy_-_IgE_mediated_food_allergy/
- https://www.jacionline.org/article/S0091-6749(21)00230-X/fulltext

Much of this information also comes from conversations with doctors specializing in allergy/immunology and/or IBD (gastroenterology) at the University of Pittsburgh Medical Center and Cleveland Clinic

None of the information here is intended to be used as medical advice. I am not a doctor. The purpose of the information presented is to inform people about different types of allergy triggers and reactions. Any and all assumptions made are generalizations. Always talk to your healthcare providers about your specific case!