The Immune System

The **immune system** contains many types of cells and molecules that work together to defend our bodies against microorganisms (e.g. bacteria, fungi, parasites), viruses, cancer cells, and toxins. A functional system can identify a threat, attack it, eliminate it, and remember it.

Antigens are things that will trigger the immune system to react. Pathogens are disease-causing organisms (bacteria, parasites etc.) and are one type of antigen that will trigger the immune system to react.

Two Branches: Innate and Adaptive Immune Responses

	Innate Immune Response		Adaptive (Acquired) Immune Response
Timing	Fast. Response starts within minutes to hours of encountering a pathogen.		Slow. Takes a few days to weeks after meeting a pathogen to mount a full response.
Target	Non-specific. Responses are similar for a class of pathogens (for example- viruses vs. bacteria)		Highly specific and diverse. Each cell has a receptor that targets a specific antigen.
Memory	Cannot remember the pathogen. Will battle it the same way next time.		Remembers the pathogen and responds more quickly/strongly if the same pathogen invades again.
White Blood Cells	Granulocytes	Antigen-presenting cells (APCs) Dendritic cells Macrophages Monocytes Others Natural killer (NK) cells	B cells matures in the bone marrow T cells matures in the thymus
Non-cell parts	 Chemical barriers (tears, mucus, stomach acid) Physical barriers (skin, gut, hair in airway) Molecules that trigger inflammation (histamine, cytokines, and others) 		Non-applicable
Main Tasks	 Recruits other cells by releasing cytokines Raises the body temperature (starts a fever) Triggers inflammation pain, swelling, discoloration, heat Phagocytosis: Engulfs and kills pathogens Presents the antigen to T-cells to trigger an adaptive immune response. 		Eliminates the specific pathogen B cells – make antibodies (immunoglobulins) that "label" the pathogen to be destroyed CD4+ Helper T-cells releases cytokines to coordinate response CD8+ Cytotoxic T-cells kills specific pathogens Develops a memory of the pathogen.

In some cases, the innate response is enough to clear the pathogen. If this first line of defence is overwhelmed, the adaptive immunity helps fight it. The antigen presentation by APCs to T cells is the connection between the innate and adaptive immune responses.

Immune System Series for Individuals with 22q11.2 Differences

The Immune System (continued)

A Simplified Example of an Immune Response



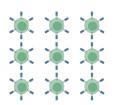
Microphages gobble up invading pathogens and are specialized for fungi and an ancient type of bacteria called mycobacteria



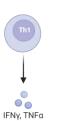
Neutrophils are also "gobblers" and release granules to destroy most common bacteria.



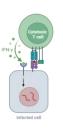
Dendritic cells gobble up the bacteria and show the broken pieces to T cells to say, "This is the enemy."



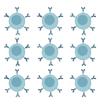
A T cell that has the correct receptor for the pathogen – mostly viruses- makes many copies of itself.



Helper T cells release cytokines (signalling molecules) to call for help from macrophages and B cells.



Cytotoxic T cells kill infected cells. Most T cells die after doing their job, but some stay alive and become memory T cells.



A B cell that has the correct receptor for the bacteria makes many copies of itself.



B cells release antibodies to label the bacteria for destruction. Most B cells die after doing their job, but some stay alive and become memory B cells.

When the immune system does not work properly...

- Hypersensitivity the immune system reacts to something harmless like nuts, dairy, eggs, or seafood. E.g. allergy
- Autoimmunity the immune system reacts to the host's own healthy tissues. E.g. multiple sclerosis, type I diabetes
- Chronic inflammation the features of normal inflammation (heat, redness, swelling, and pain) become uncontrolled. E.g. asthma, rheumatoid arthritis, inflammatory bowel disease
- Immunodeficiency the ability of the immune system to fight infections is weakened or absent.
 Features include having many infections, severe infections, or infections with unusual organisms.
 Autoimmunity can also be part of immunodeficiency.

Immunodeficiency is common in 22q11.2DS and is also seen in people with 22q11.2DupS.

Resources

- · Short videos by Oxford Immunotec:
 - Introduction to the immune system
 - How to we fight infections?
 - How to T cells protect us from infections?
- A more detailed video by Osmosis by Elsevier: Introduction to the immune system
- What is immunology? British Society for Immunology
- <u>Parts of the Immune System</u> Children's Hospital of Philadelphia
- An introduction to immunology and immunopathology – Review article (2018)
- Images created in BioRender.com



The mission of the <u>International 22q11.2 Foundation</u> is to improve the quality of life for individuals affected by chromosome 22q11.2 differences through family and professional partnerships.

This information is brought to you by the Foundation for educational purposes only. It is <u>not</u> intended to be taken as medical advice. If you have concerns, please talk to your healthcare provider.