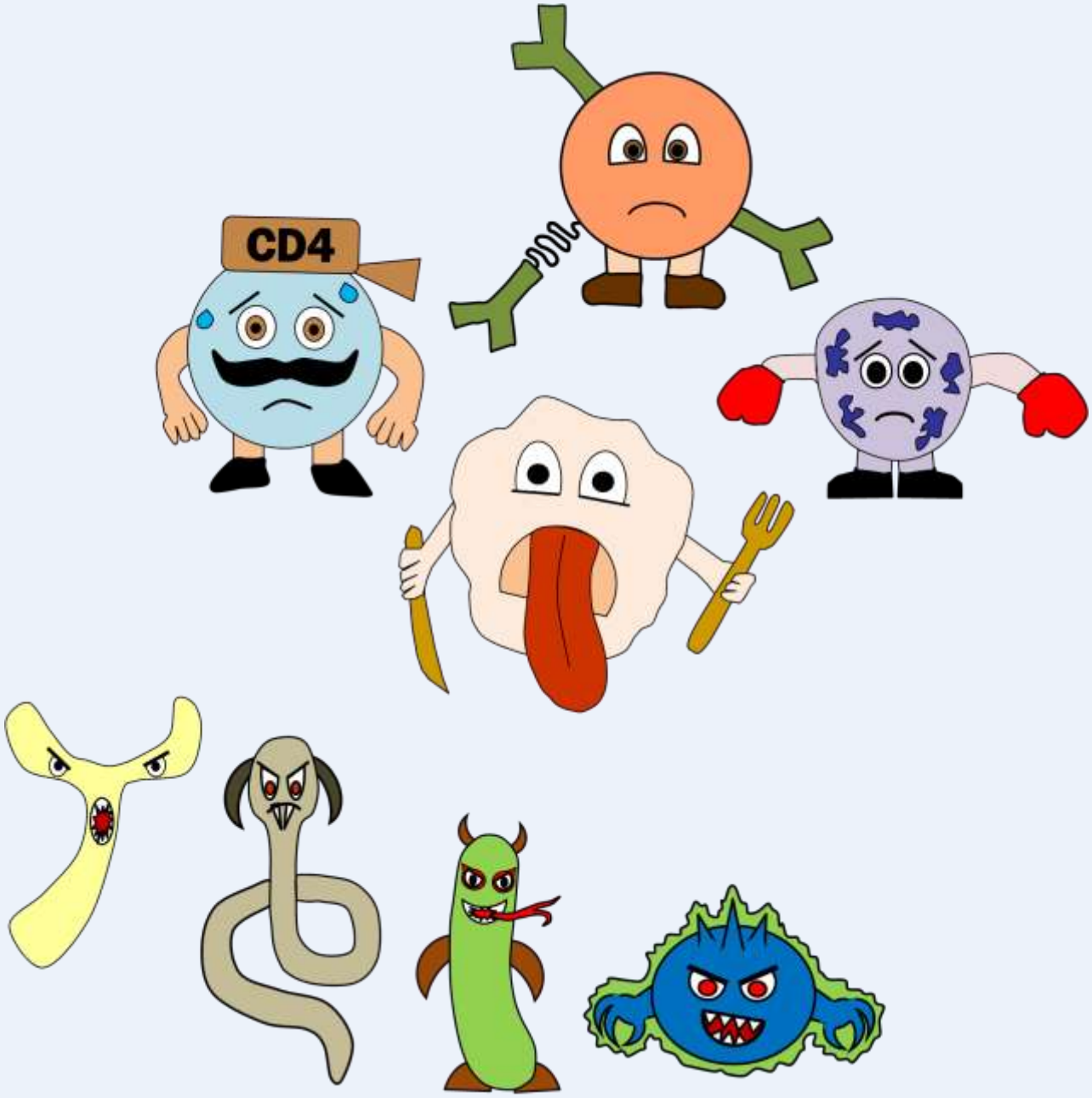
When the Immunocytes get sick...

Primary Immunodeficiencies



Juan Carlos Aldave, MD

Allergy and Clinical Immunology

When the Immunocytes get sick

Primary Immunodeficiencies

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Around us, in the environment, there are many microbes that can harm us, causing illness or death.

There are 4 major groups of germs: viruses, bacteria, fungi and parasites. We are exposed to these threats since birth, so we need to have many cells and molecules to defend our bodies.

We will call "immune system" to our body defenses, and "immunocytes" to the immune cells that protect us.

In this little book I will show the danger we face when our immunocytes get sick and stop working.

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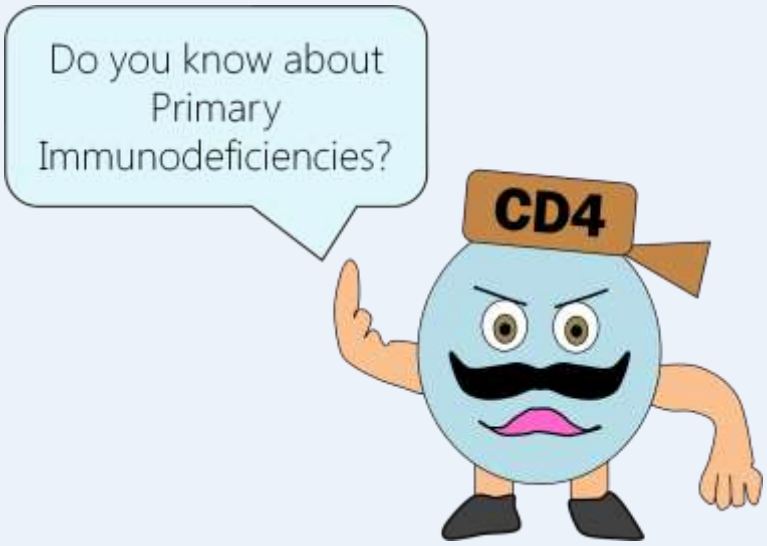
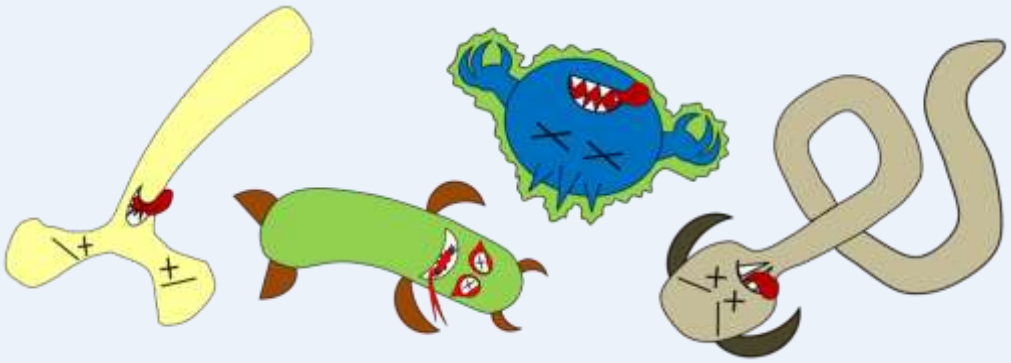


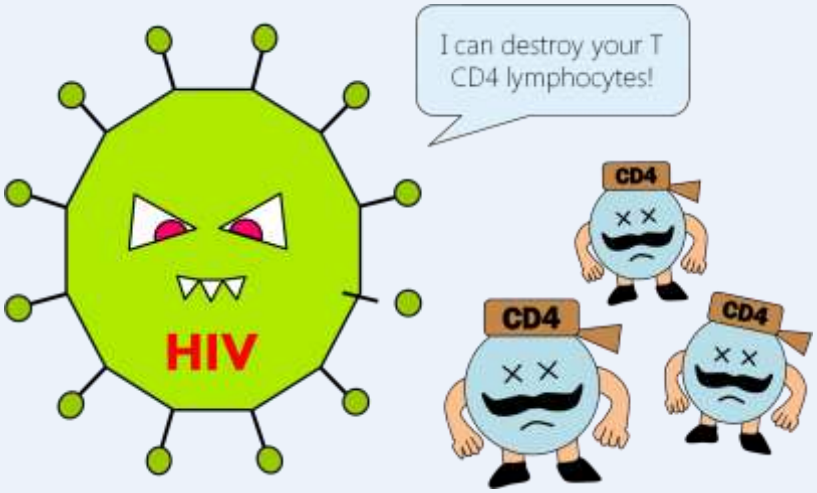
Chapter 1: Why do we lose the ability to defend?

The main function of our immunocytes is to protect us from dangerous microbes that threaten our life. In the previous books we learned how our immunocytes kill the bugs Candida, Mycobacterium, Ascaris and Pneumococcus.



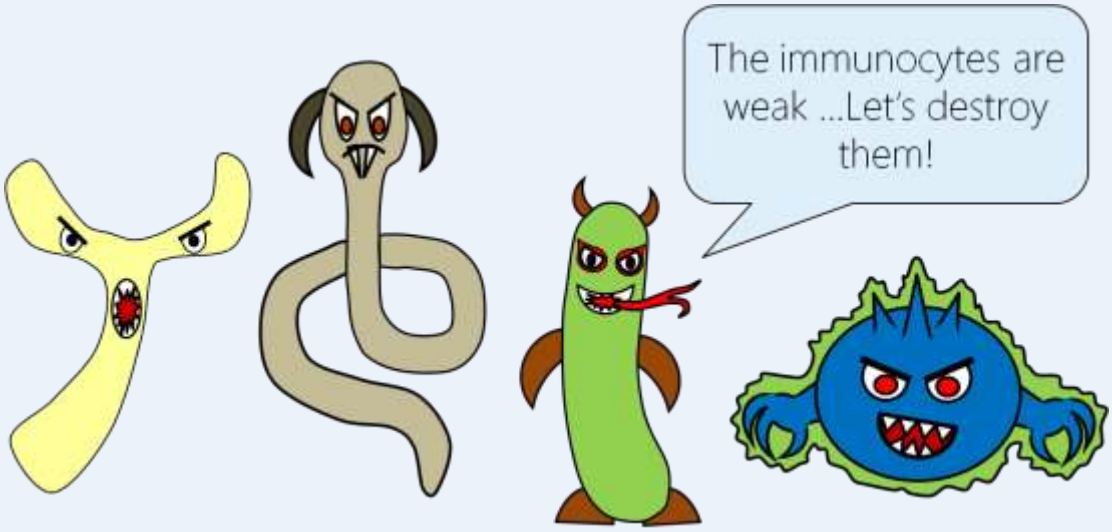
Sadly, there are conditions that cause a decrease in the number or function of our immunocytes. For instance:

- Acquired immunodeficiency syndrome (AIDS), caused by human immunodeficiency virus (HIV).



- The use of chemotherapy for the treatment of cancer or during transplantation procedures.
- The use of high doses of corticosteroids, like prednisone or dexamethasone.
- Poorly controlled chronic debilitating diseases, such as cancer, diabetes mellitus, chronic renal failure or liver cirrhosis.
- Genetic defects of the immune system, known as primary immunodeficiencies.

Children and adults whose immune cells are sick become highly susceptible to the attack of surrounding microbes.



Please answer the following questions:

1. What is the main function of our immunocytes?

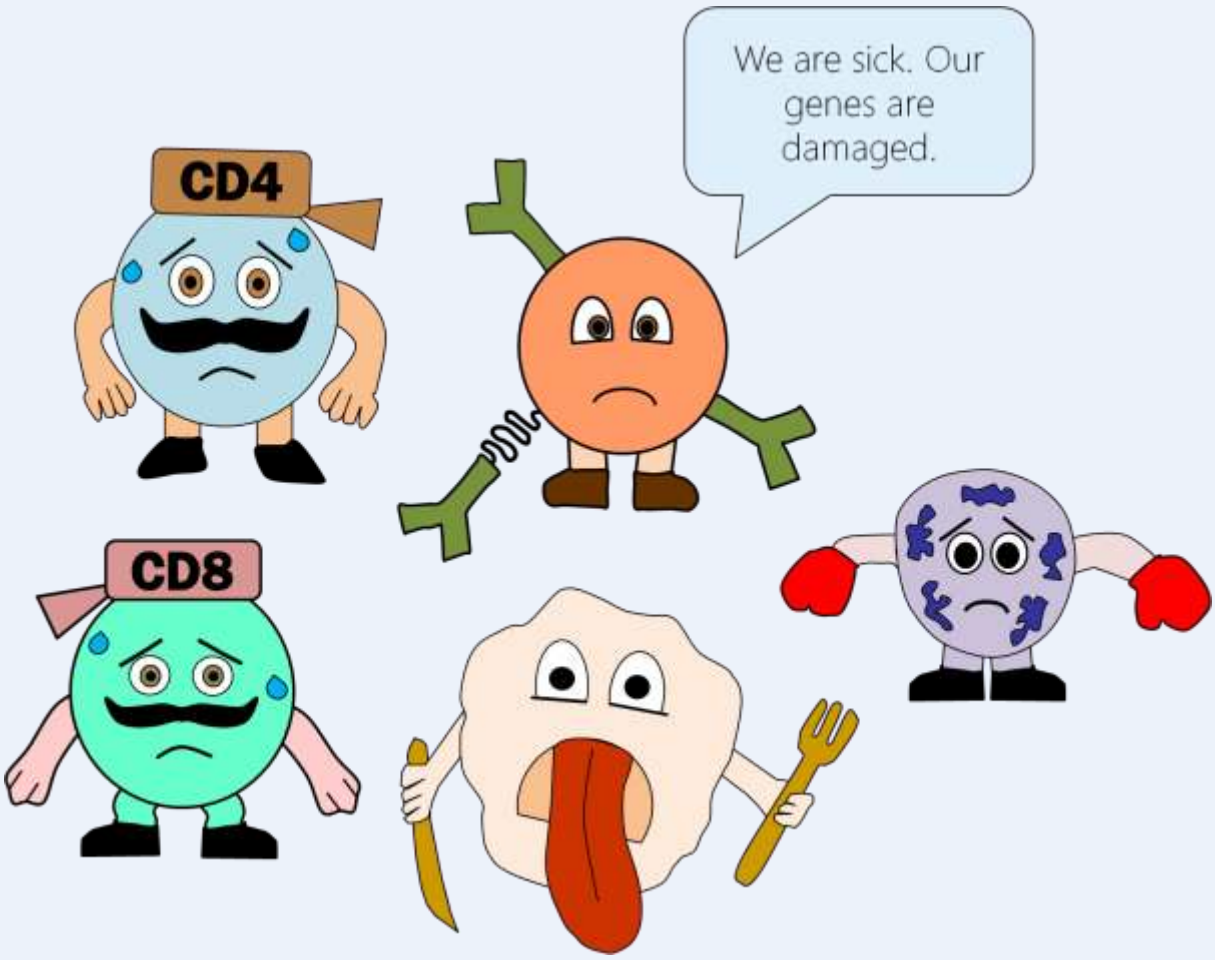
   _____

2. Write some conditions that decrease the number or function of our immunocytes:

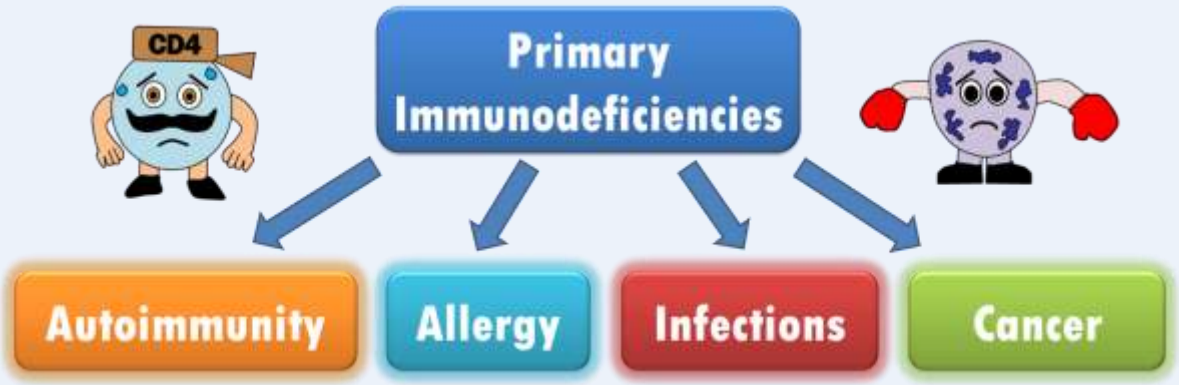
   _____

Chapter 2:
Primary immunodeficiencies

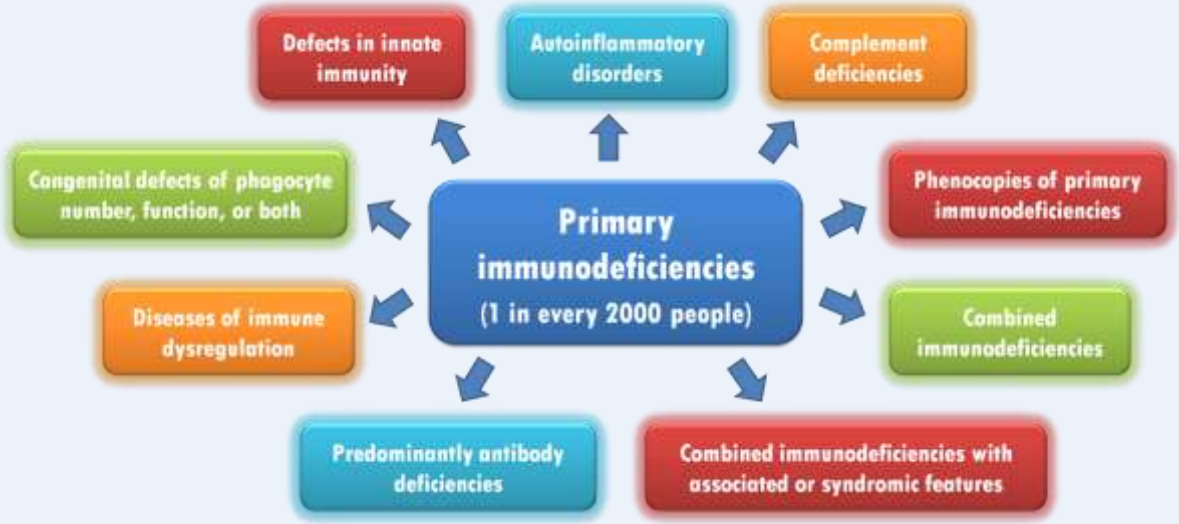
Primary Immunodeficiencies are diseases that occur because of genetic defects of the immune system.



When immune cells are sick, the microbes take advantage and cause frequent and severe, even fatal, infections. People with immune deficiency may also be more susceptible to autoimmune diseases, allergies and cancer.



Primary immunodeficiencies are frequent diseases. They affect one in every 2000 people, mostly children. The types of infections that affect the patient depend on which immunocytes are sick. There are 9 large groups of primary immunodeficiencies.



In the following chapters we will see what happens when four of our major immune cells get sick. Please answer these questions:

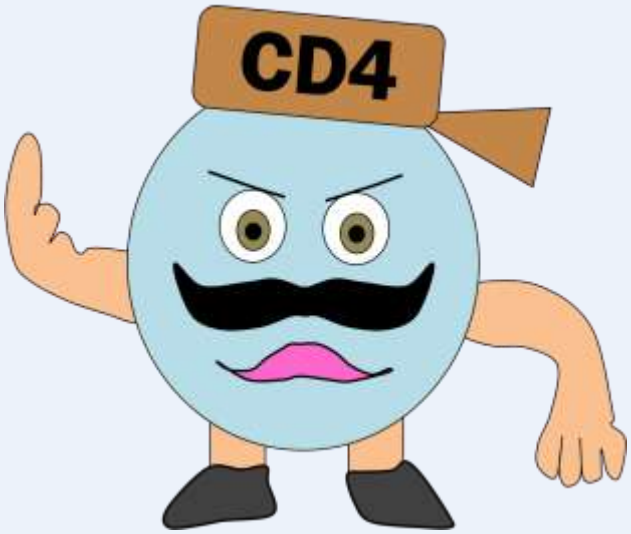
1. What are primary immunodeficiencies?

   _____

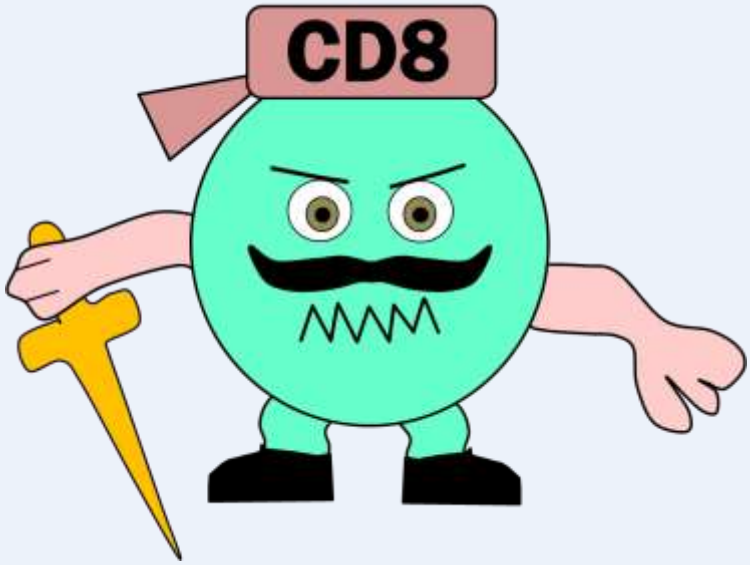
2. True or false? People with immunodeficiencies can be more susceptible to infections, autoimmune diseases, allergies and cancer _____

Chapter 3: Felix and Pacco, the T lymphocytes, got sick

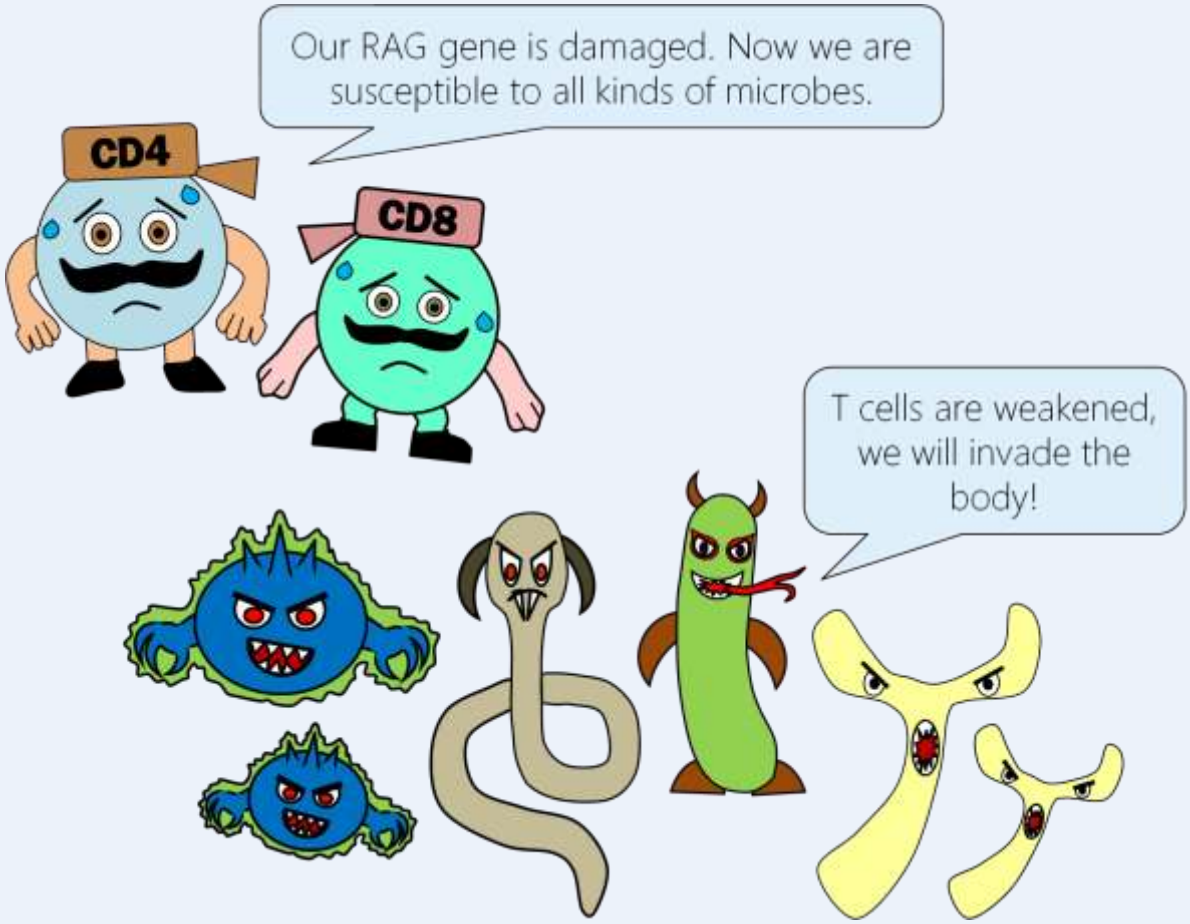
Felix is our T CD4 lymphocyte. His major ability is to help the other immunocytes for an optimal activation and function.



Pacco is our T CD8 lymphocyte. His main work is to destroy cells that are infected by viruses and malignant cells that cause cancer.



T lymphocytes are the most important cells in our immune system. When Felix and Pacco get sick, our body becomes vulnerable to all kinds of infections by fungi, bacteria, viruses and parasites.



Children with T-cell deficiencies who do not receive prompt treatment die at an early age. The most widely used curative treatment is stem cell transplantation, also known as bone marrow transplant.

1. What happens when Felix and Pacco get sick?

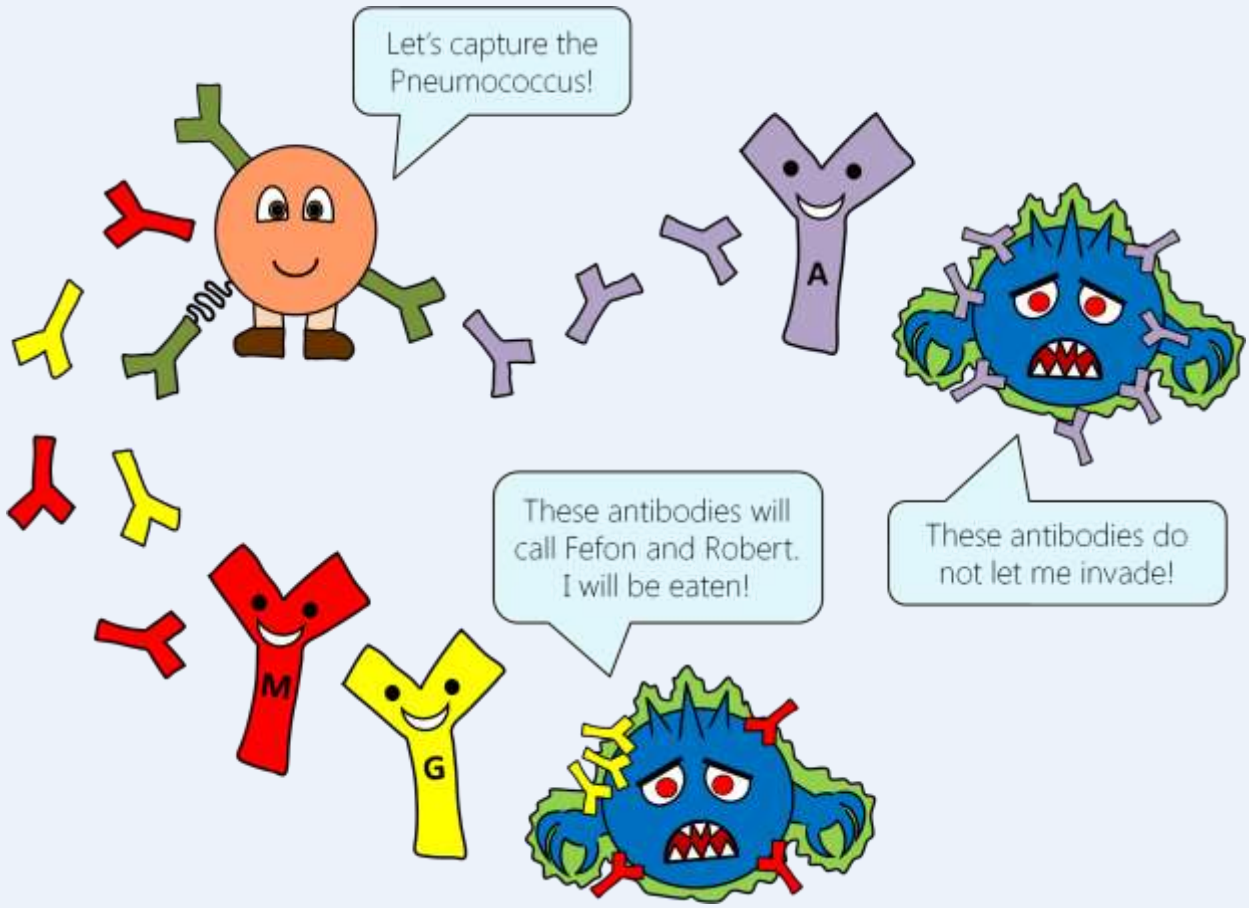
   _____

2. What is the most widely used curative treatment for children with T-cell deficiencies?

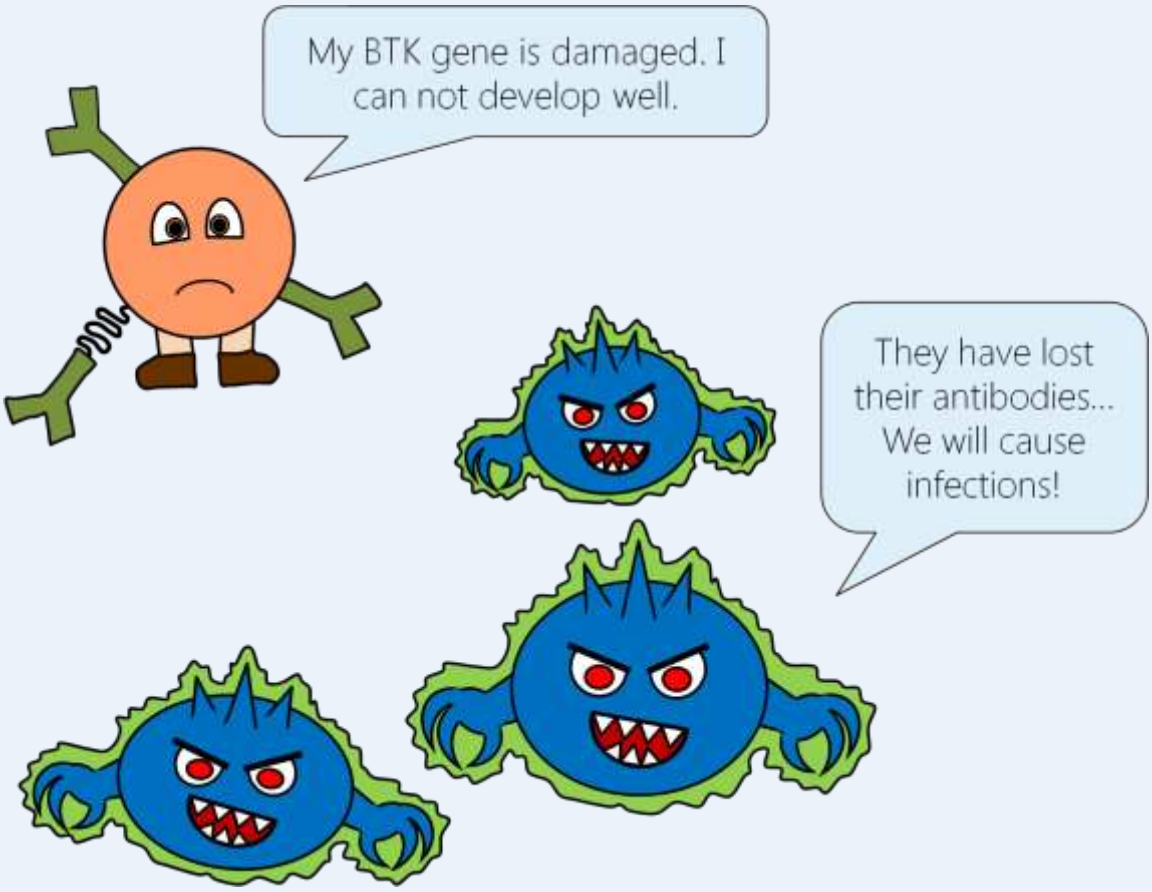
   _____

Chapter 4: Moli, the B lymphocyte, became ill

Moli is our B lymphocyte, the immunocyte that fabricates our 5 classes of antibodies or immunoglobulins (IgG, IgA, IgM, IgD and IgE). Antibodies protect us from many bacteria, viruses and parasites.



The action of B cells is very important for our life. When Moli becomes ill and stops producing antibodies, our body becomes susceptible to various infections by bacteria, viruses and parasites.



Defects of B lymphocytes are the most frequent primary immunodeficiencies, representing about 50% of cases.

Treatment of children with primary immunodeficiencies affecting B cells depends on the severity of the disease. For some children it is only necessary to monitor the patient or to give him antibiotics. In more severe cases it is necessary to use antibody replacement therapy lifelong.

Let's help Moli to solve the following questions:

1. What is the main function of Moli?

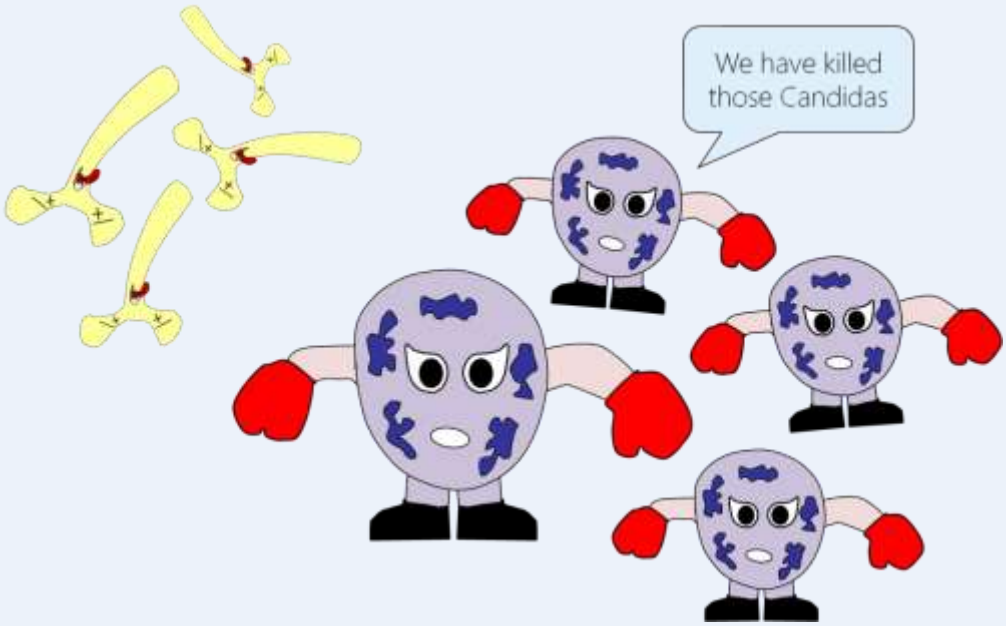
   _____

2. What occurs when Moli becomes ill?

   _____

Chapter 5: When Robert the neutrophil can no longer fight

The principal labor of Robert the neutrophil is to quickly destroy bacteria and fungi by eating them and throwing toxic substances. We have millions of neutrophils in the blood, all of them ready to fight.

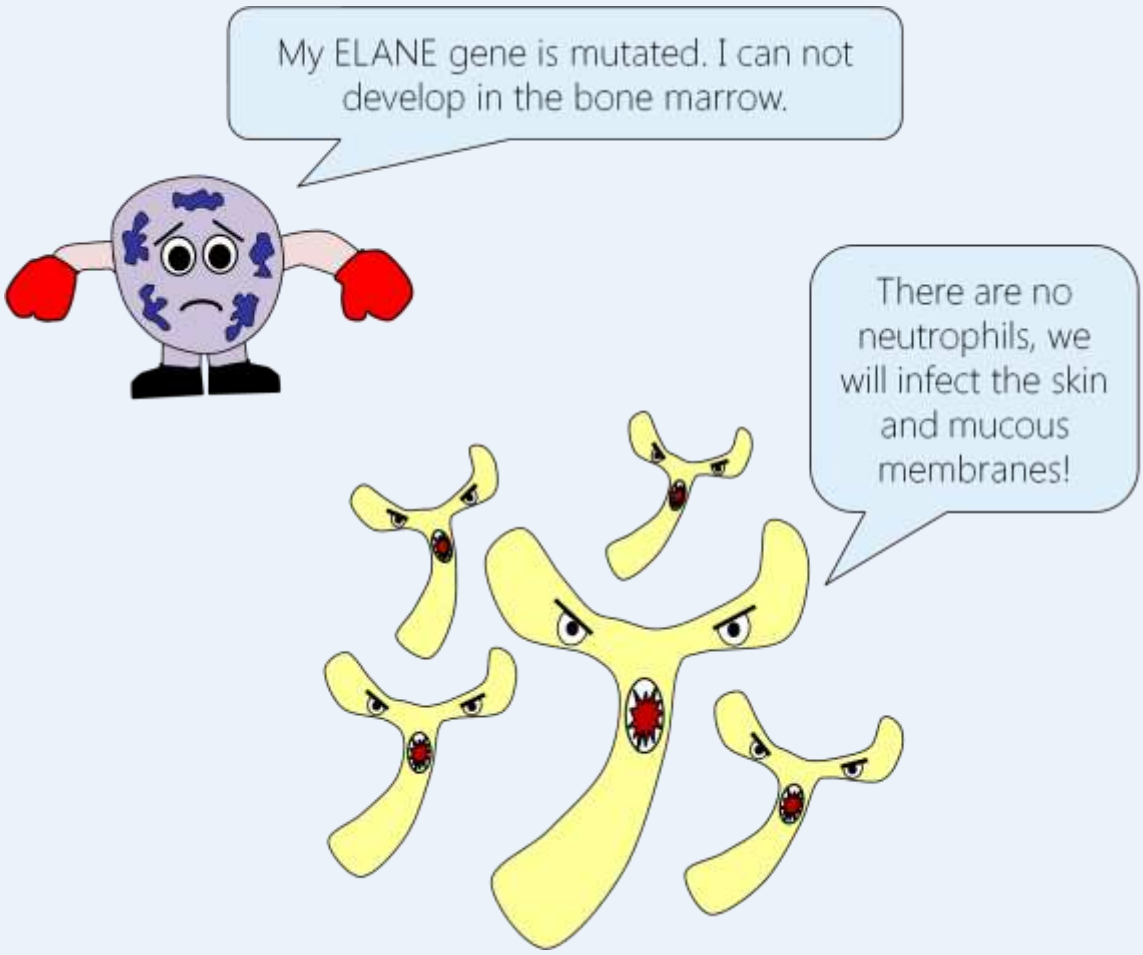


Neutrophils are essential to preserve our life. There are children with genetic defects that cause a detention in the development of neutrophils in the bone marrow. This disease is named 'severe congenital neutropenia'.

In other children, neutrophils are produced normally but they cannot reach infection sites because they lack some important molecules to pass through blood vessels. This group of diseases is called 'leukocyte adhesion deficiency'.

Neutrophil attack is also weakened when TH17 lymphocytes do not develop or do not work properly.

When Robert and his friends are not fabricated, the child becomes prone to severe bacterial and fungal infections.



The only curative treatment for children who lack neutrophils is bone marrow transplantation. The use of antibiotics, antifungal drugs and neutrophil-stimulating factor is also very important for these patients.

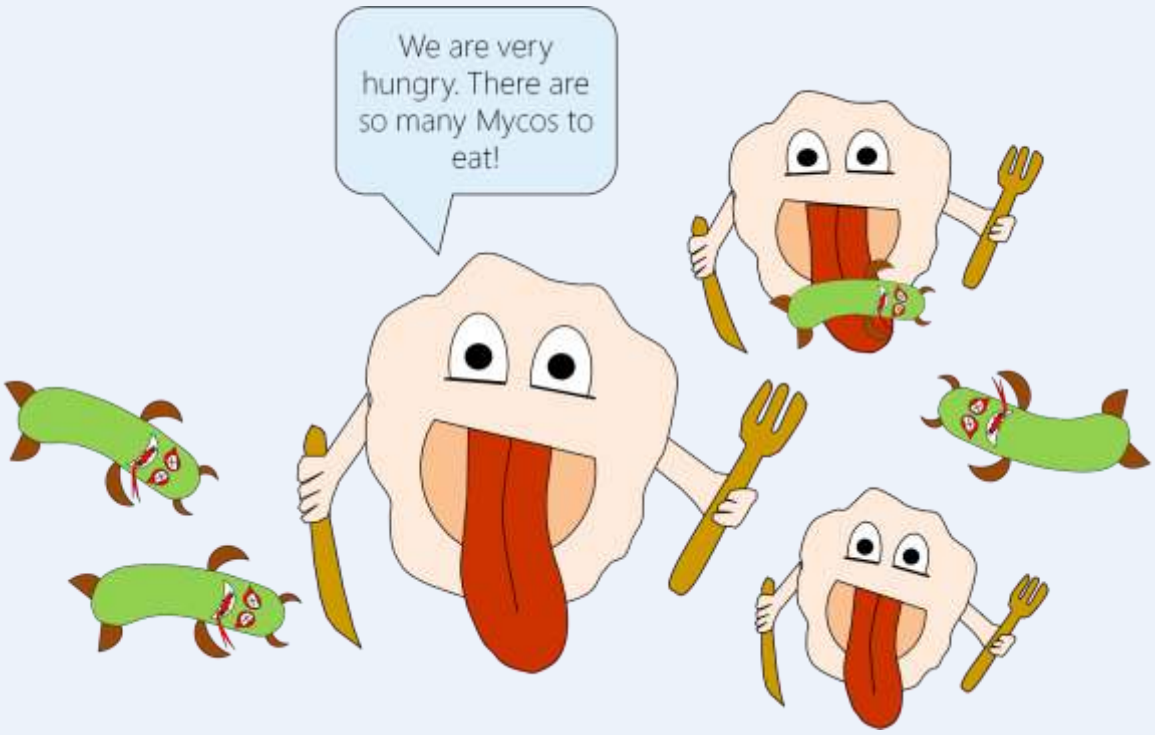
Please help Robert to solve the following question:

1. What happens to children who lack neutrophils?

_____

Chapter 6: When Fefon the macrophage stops eating

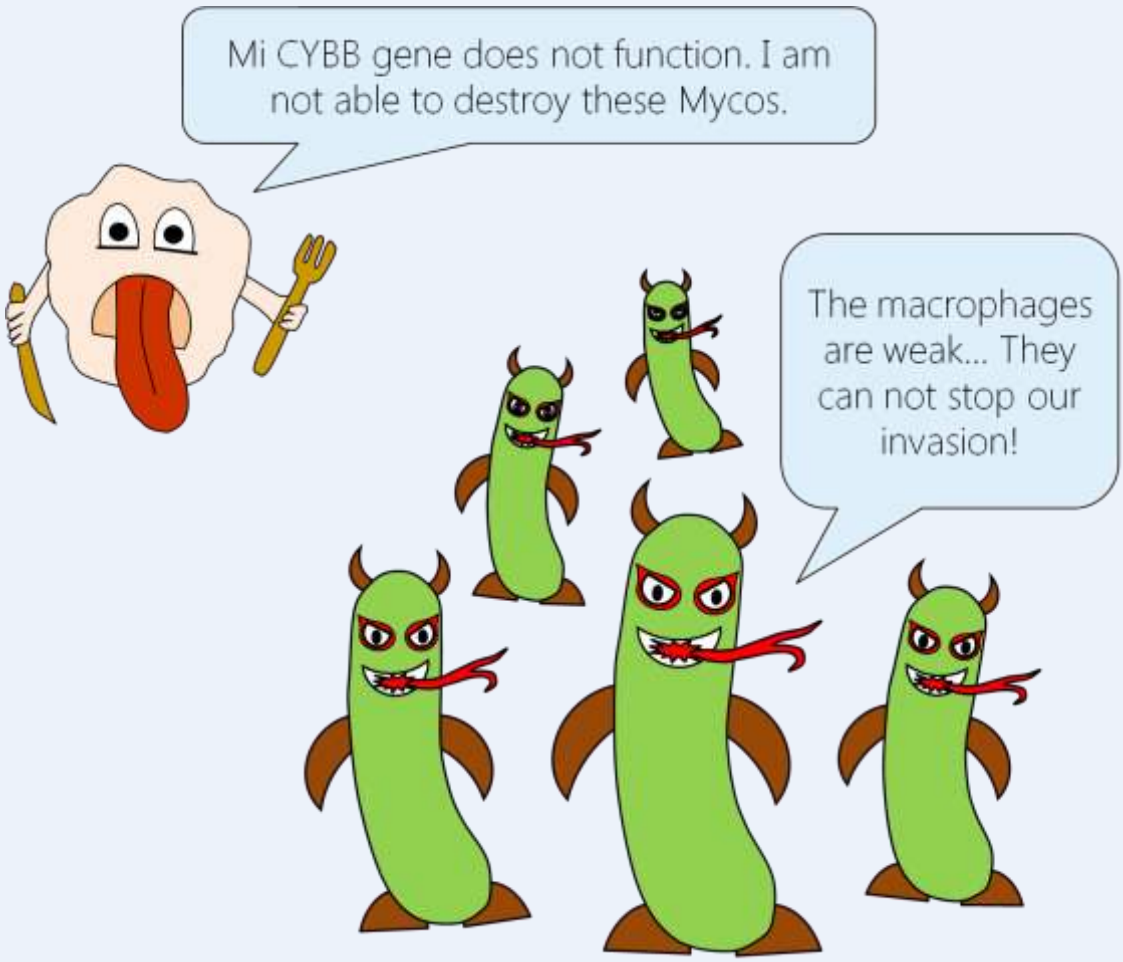
Fefon is our powerful macrophage. His main function is to eat and kill dangerous bacteria and fungi. In addition, Fefon can call other immunocytes to the infection site.



Macrophages are very important cells to preserve our life. When Fefon gets sick and loses the ability to eat and destroy microbes, our body becomes susceptible to severe bacterial and fungal infections.

Chronic granulomatous disease (CGD) is a primary immunodeficiency that affects one in every 250 thousand people. The macrophages of patients with CGD are very ill and cannot kill microbes.

Thus, children with CGD have recurrent severe bacterial and fungal infections. In addition, they are affected by tumors called granulomas. Most children die if they do not receive a bone marrow transplant.



The attack of macrophages is also weakened when TH1 lymphocytes do not develop or do not work properly.

Please help Fefon to solve the following questions:

1. What is the main function of Fefon?

   _____

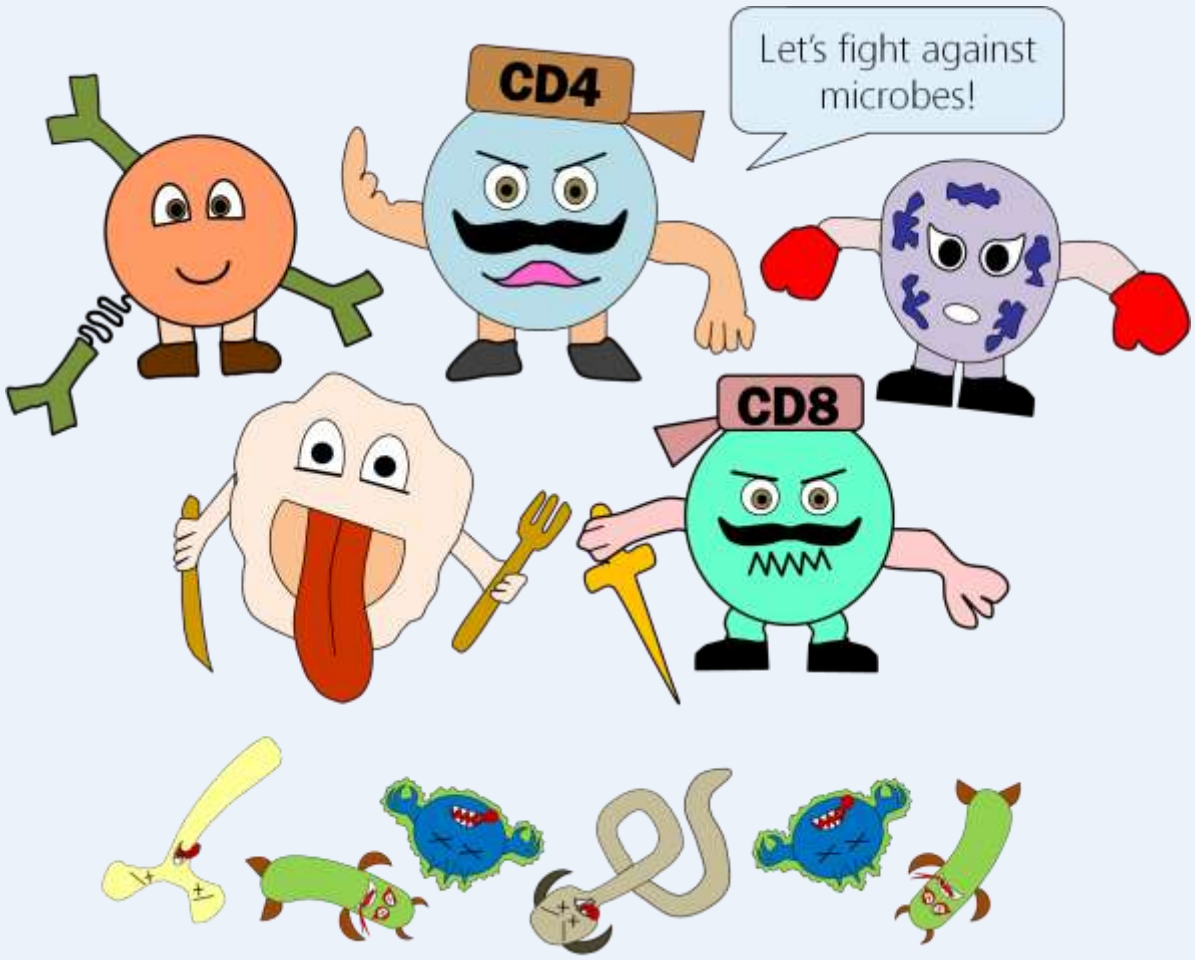
2. What happens when Fefon gets sick?

   _____

Chapter 7: Curing children with primary immunodeficiencies

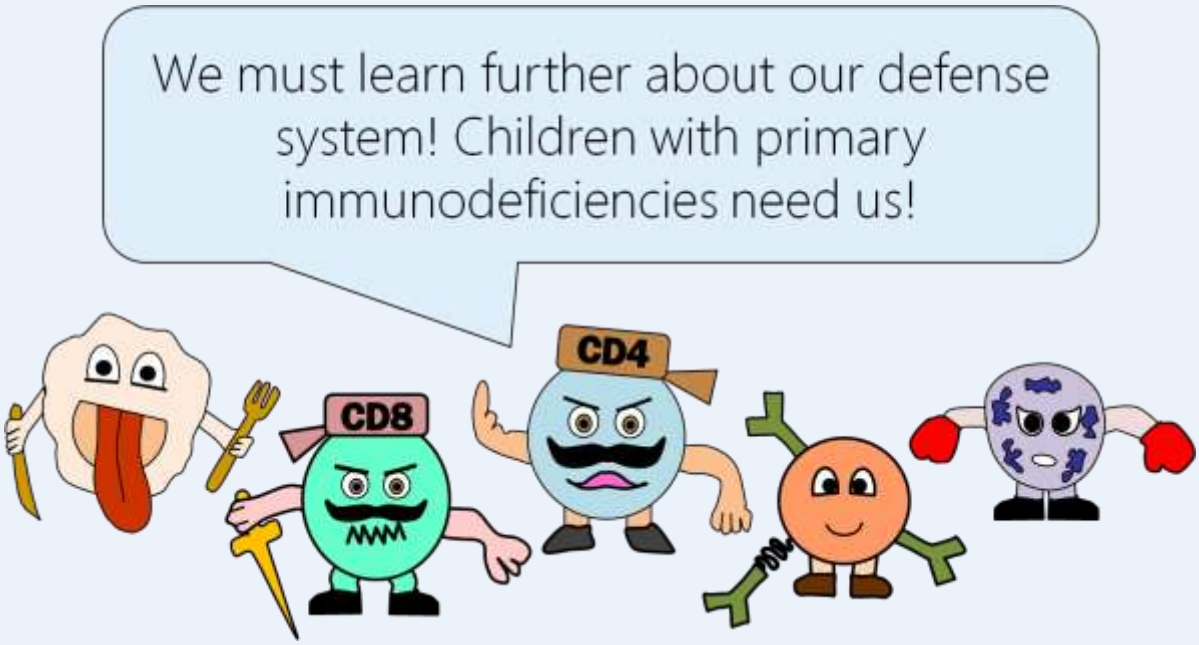
Primary Immunodeficiencies are diseases that occur as a result of genetic defects of the immune system.

When our immune cells get sick, surrounding microbes invade us causing frequent and severe infections. Thus, we need to have healthy and powerful immunocytes. A proper function of the immune system is essential to safeguard our life.



We estimate that in Peru there are 15,000 children with primary immunodeficiencies who have not been diagnosed and, consequently, do not receive adequate treatment. Many of them die in the first year of life.

Therefore, it is important to understand the function of our immunocytes and what happens when they are sick. If we improve our knowledge of primary immunodeficiencies, we will diagnose sick children promptly, give them an appropriate treatment, and hence save their lives.



Let's end this book with the following questions:

1. How many Peruvian children might have a primary immunodeficiency?

   _____

2. Why is it important to know about primary immunodeficiencies?

   _____

In this little book we learned what happens when the immunocytes are sick in children with primary immunodeficiencies.

Do not miss the following book, where we will understand the diseases that occur when our immunocytes lose the ability to develop 'immune tolerance'.

Juan Carlos Aldave, MD

Allergy and Clinical Immunology

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- Dr. Juan Félix Aldave Pita
- Bertha Alicia Becerra Sánchez

"For God so loved the world that he gave his one and only Son, that whoever believes in him shall not perish but have eternal life". John 3:16

10 Warning Signs of Primary Immunodeficiency

Primary Immunodeficiency (PI) causes children and adults to have infections that come back frequently or are unusually hard to cure. 1:500 persons are affected by one of the known Primary Immunodeficiencies. **If you or someone you know is affected by two or more of the following Warning Signs, speak to a physician about the possible presence of an underlying Primary Immunodeficiency.**

1. 
Four or more new ear infections within one year.

2. 

Two or more serious sinus infections within one year.

3. 

Two or more months on antibiotics with little effect.

4. 
Two or more pneumonias within one year.

5. 

Failure of an infant to gain weight or grow normally.

6. 
Recurrent, deep skin or organ abscesses.

7. 
Persistent thrush in mouth or fungal infection on skin.

8. 
Need for intravenous antibiotics to clear infections.

9. 

Two or more deep-seated infections including septicemia.

10. 
A family history of PI.

"These warning signs were developed by the Jeffrey Modell Foundation Medical Advisory Board. Consultation with Primary Immunodeficiency experts is strongly suggested.

www.INFO4PI.org

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Book 11: The armor of the Immunocyte Felix

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