



Fitango Education
Health Topics

Chronic Granulomatous disease



<http://www.fitango.com/categories.php?id=373>

Overview

Chronic Granulomatous Disease (CGD) is an inherited primary immunodeficiency disease (PIDD) which increases the body's susceptibility to infections caused by certain bacteria and fungi.

Overview

Granulomas are masses of immune cells that form at sites of infection or inflammation. People with CGD are unable to fight off common germs and get very sick from infections that would be mild in healthy people.

This is because the presence of CGD makes it difficult for cells called neutrophils to produce hydrogen peroxide. The immune system requires hydrogen peroxide to fight specific kinds of bacteria and fungi.

Overview

These severe infections can include skin or bone infections and abscesses in internal organs (such as the lungs, liver or brain).

Aside from the defective neutrophil function in CGD, the rest of the immune system is normal. People with CGD can be generally healthy until they become infected with one of these germs. The severity of this infection can lead to prolonged hospitalizations for treatment.

Overview

Children with CGD are often healthy at birth, but develop severe infections in infancy or early childhood.

The most common form of CGD is genetically inherited in an X-linked manner, meaning it only affects boys. There are also autosomal recessive forms of CGD that affect both sexes.

Symptoms

People with CGD can easily fight off some infections, but not

those that need the body's natural ability to produce hydrogen

peroxide to control disease. For this reason, symptoms of

Symptoms

recurrent bacterial or fungal infections may be sporadic.

Symptoms

CGD may involve any organ system or tissue of the body, but

infections are usually found in the skin, lungs, lymph nodes,

liver, and bones.

Symptoms

Symptoms

condition known as granuloma may develop.

Symptoms

Pneumonia caused by a fungus such as Aspergillus is a red flag

for CGD and often results in testing.

Symptoms

Some other typical symptoms of CDC are:

Symptoms

-- Bone infections

Symptoms

-- Frequent and difficult-to-clear skin infections
(abscesses,

chronic infection inside the nose, furuncles,
eczema, impetigo,

perianal abscesses)

Symptoms

-- Joint infections

Symptoms

-- Persistent diarrhea

Symptoms

-- Pneumonia

Symptoms

-- Frequent swollen lymph nodes in the neck; those develop

early in life. The lymph nodes may form abscesses that require

surgical drainage.

Symptoms

Causes

Chronic granulomatous disease (CGD) is a genetic disorder in which certain immune system cells are unable to kill some types of bacteria and fungi. The disorder leads to long term (chronic) and repeated (recurrent) infections. The condition is often discovered in very early childhood.

Causes

About half of CGD cases are transmitted as a recessive, sex-linked trait. This means that boys are more likely to inherit the disorder than are girls. Boys have an X and a Y chromosome. Girls have two X chromosomes. The defective gene is carried on the X chromosome. So, if a girl has one X chromosome with the defective gene, the other X chromosome may have a working gene to make up for it. A girl has to inherit the defective gene from both parents in order to have the disease.

Causes

About 1 in a million people have CGD.

Diagnosis

The timing of diagnosing CGD is often dependant upon when the person begins having recurrent bacterial or fungal infections associated with the disease.

Diagnosis

The most accurate test to confirm CDG is done by measuring the amount of hydrogen peroxide produced by the body's cells. Other tests may include a bone scan, chest x-ray, complete blood count, flow cytometry tests, and nitroblue tetrazolium test.

Physical tests may show an enlarged liver, spleen, or a swelling of the lymph nodes.

Diagnosis

<http://www.aaaai.org/conditions-and-treatments/primary-immunodeficiency-disease/chronic-granulomatous-disease.aspx>

Prevention

The best treatment plan for CGD is to prevent infections from occurring. Special preventative antibiotics are a mainstay of treatment for CGD. These greatly reduce the chances of infection.

Prevention

People with CGD should also avoid swimming other than in chlorinated pools. Fresh or salt water may contain organisms that, while safe for healthy people, can cause infection in people with CGD.

Prevention

Exposure to garden mulch can cause a life-threatening form of Aspergillus pneumonia. People with CGD should never use mulch in their own yards and should stay indoors while it is being spread in neighboring yards. Turning compost piles, repotting plants, cleaning cellars and riding on hay rides are other activities that should be avoided.

Prevention

Marijuana can also contain Aspergillus and should not be smoked by CGD patients. When an infection occurs, antibiotic treatment is prescribed but it generally requires a much longer amount of time to be effective.

Bone marrow transplant is another treatment option for some people with severe symptoms of CGD.



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