Tumor Necrosis Factor Receptor Associated Periodic Syndrome

The severity of TRAPS varies from patient to patient and often depends on the specific gene defect

Fast Facts

- TRAPS is a rare genetic disease with episodes of recurrent fever, abdominal, chest and muscle pain and a typical rash lasting for more than one week.
- The severity of the disease varies from patient to patient and often depends on the specific gene defect.
- Episodes of TRAPS respond to treatment with steroids and other biologic anti-inflammatory medications.

What is TRAPS?

Formerly known as Familial Hibernian fever, Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) is a genetic disease that causes recurrent episodes of fever that typically last 1-3 weeks that are associated with chills and severe muscle pain in the trunk and the arms.

Patients develop a red and painful rash from the trunk to the arms and legs. Abdominal pain with nausea, vomiting and diarrhea are common, as are red, swollen eyes. Other important features include chest pain due to inflammation of the membrane surrounding the lungs or heart.

Who gets TRAPS?

TRAPS is a rare disease, but since the gene defect was discovered, more patients have been diagnosed. The disease affects both males and females, and usually starts before the age of 10 years, but symptoms may start in late childhood, or even in adulthood. The first cases were reported in patients from Irish-Scottish background; the disease has now been seen in almost all ethnic groups.

What causes TRAPS?

TRAPS is a genetic diseases. TRAPS is inherited as an autosomal dominant disease (in which a mutation in only one copy of the gene inherited from one parent is sufficient to cause disease). That means that one of the parents may be ill with TRAPS, or that the gene mutation developed in the sick child. TRAPS is due to a gene defect in a protein called tumor necrosis factor receptor, which leads to an increase of the patient's normal inflammatory response. Infection, trauma, strenuous exercise or psychological stress may trigger episodes.

How is TRAPS diagnosed?

A physician will suspect TRAPS based on the clinical features, the physical examination and the family medical history. Blood tests will show signs of inflammation during, and often even between, episodes. Genetic tests can confirm diagnosis. The physician will probably also test for other types of periodic fever syndromes.

How is TRAPS treated?

There is still no proven treatment to prevent or cure the disease. Non-specific anti-inflammatory agents—including steroids—help relieve symptoms, but long-term use of steroids can lead to serious side effects. Etanercept (*Enbrel*, a medicine used to treat arthritis), which blocks the tumor necrosis factor, has been shown to be an effective treatment in some patients, either when given at the beginning of an attack or as a preventive medicine. Anakinra (*Kineret*) and canakinumab (*Ilaris*) medications that blocks interleukin-1, an important protein involved in inflammation, are effective in most patients, including those who do not respond well to etanercept.

Patients with persistent, uncontrolled inflammation may develop a complication called amyloidosis. Amyloid is a protein that deposits in organs in children with chronic inflammatory disease. The most common organ involved is the kidney, but amyloid can deposit in the intestines, skin and heart. Eventually, amyloid causes a loss of function, especially in the kidneys. This is the main reason to continue life-long treatment, even when the child is feeling better.

Living with TRAPS

Patients with a severe type of TRAPS will suffer from long episodes with severe abdominal and muscle pain as well as side effects of medications, particularly steroids. Many patients will need psychological support or special support to treat the pain during attacks. However, for many, the disease can be controlled with medication, and affected individuals can lead relatively normal lives.

Points to remember

- TRAPS is a rare genetic disease with long (more than 1 week) episodes of fever, rash, red and swollen eyes, abdominal, chest and muscle pain.
- TRAPS may result in late kidney failure if not treated.
- New medications that target tumor necrosis factor or interleukin-1 are effective in most patients and decrease the need to treat with steroids.

To find a rheumatologist

For more information about pediatric rheumatologists, visit www.rheumatology.org.

For a listing of pediatric rheumatologists in your area, click here.

For additional Information

The American College of Rheumatology has compiled this list to give you a starting point for your own additional research. The ACR does not endorse or maintain these websites, and is not responsible for any information or claims provided on them. It is always best to talk with your rheumatologist for more information and before making any decisions about your care.

Familial Mediterranean fever article

www.healthyparent.com/FMF/TRAPS.html

TRAPS syndrome article

www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=GB&Expert=32960

Updated March 2013

Written by Philip Hashkes, MD, MSc, Shaare Zedek Medical Center and the Hebrew University, Jerusalem, and reviewed by the American College of Rheumatology Communications and Marketing Committee.

This patient fact sheet is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.