



PATIENT FACT SHEET

Tumor Necrosis Factor Receptor Associated Periodic Syndrome (Juvenile)



CONDITION DESCRIPTION

Tumor necrosis factor receptor associated periodic syndrome (TRAPS) is a rare, genetic disorder in children. It is caused by a defective gene mutation.

The abnormal gene may be inherited from one parent.

TRAPS affects both males and females. It usually starts before age 10, but can occur in later childhood and even adulthood. The first TRAPS cases were spotted in children

of Irish-Scottish background, but all ethnic groups are affected.

TRAPS is caused by a gene defect in the protein tumor necrosis factor receptor. This increases the inflammatory response. Infection, trauma, stress or strenuous exercise may trigger episodes. Severity varies among patients according to the specific gene defect.



SIGNS/ SYMPTOMS

Children with TRAPS may have episodes of recurrent fever; chills; severe muscle pain in the abdomen, chest or arms; red and swollen eyes; and a rash lasting for more than one week. Red, painful rash may move from the torso to the arms and legs. Abdominal pain with nausea, diarrhea and vomiting is common.

Inflammation of the membrane around the lungs or heart may cause chest pain. In mild cases, symptoms may resemble those of Periodic Fever, Aphthous Stomatitis, Pharyngitis, Cervical Adenitis [PFAPA] Syndrome.

Physicians may suspect TRAPS based on physical exam and family medical history. Blood tests may show signs of inflammation during or between flares. Genetic tests confirm TRAPS diagnosis. Testing for other periodic fever syndromes may rule out those causes.

Persistent, uncontrolled inflammation may lead to amyloidosis. Amyloid proteins may deposit in the kidneys, intestines, skin and heart. This may lead to loss of kidney function without treatment.



COMMON TREATMENTS

TRAPS treatments may control symptoms and prevent complications like kidney failure.

Corticosteroids may help ease symptoms, but long-term steroid use has serious side effects. The biologic drug etanercept (Enbrel), which blocks tumor necrosis factor, has been effective in some patients when given at the beginning of an attack or to prevent attacks. However, its effectiveness may wear off over time.

Medications that block interleukin-1, a protein involved in inflammation, are more effective TRAPS treatments. These include anakinra (Kineret) and canakinumab (Ilaris). In rare cases, the interleukin-6 blocker tocilizumab (Actemra) may be helpful.

Lifelong treatment may prevent amyloidosis and kidney failure. Patients with TRAPS should have regular urine tests to watch levels of the amyloid protein.



CARE/ MANAGEMENT TIPS

Patients with TRAPS should follow their treatment plans for life to prevent episodes and possible amyloidosis.

Treatments should be taken even when children feel well. Long-term TRAPS episodes and some medications, such as steroids, may cause side effects. Patients may need psychological support to cope or to deal with pain during TRAPS episodes.

Patients with TRAPS should be able to lead normal lives with treatment. New medications, such as interleukin-1 blockers, are effective at controlling inflammation and symptoms in these children. While TRAPS is not curable, it is manageable