Muckle-Wells Syndrome (MWS): Fact Sheet

TOPICS:

What It Is

Causes and Symptoms

Living with MWS

Diagnosis and Treatment

What It Is

Muckle-Wells Syndrome (MWS) is a subtype of Cryopyrin-Associated Periodic Syndromes (CAPS). Diagnosis usually occurs shortly after birth.

Causes and Symptoms

MWS is generally caused by mutations in a gene called *NLRP3* (nucleotide-binding domain, leucine-rich family [NLR] pyrin domain containing 3). *NLRP3* mutations cause increased activity of cryopyrin, a protein that regulates inflammation in the body. Increased cryopyrin activity causes overproduction of a protein called interleukin-1 beta (IL-1β).

Overproduction of IL-1β, in turn, causes recurrent symptoms of inflammation such as

- Rash
- Fever/chills
- Joint pain
- Eye redness/pain
- Fatigue

MWS symptoms are triggered by unknown, random factors and possibly by stress, exercise, or cold. Episodes generally last 24 to 48 hours. In most cases, patients with MWS develop progressive hearing loss. In some cases, amyloidosis, a disease in which an abnormal accumulation of the protein amyloid occurs in tissues and organs, develops later in life.

Living with MWS

Most patients with MWS experience chronic, daily symptoms of inflammation. Chronic fatigue may severely impact these patients. Symptoms are often worse at night.

Diagnosis and Treatment

Diagnosis of MWS is determined through an evaluation of a patient's symptoms and medical history. Confirmation of the diagnosis is sometimes achieved through genetic testing and the identification of *NLRP3* mutations. Not all patients, however, have a detectable genetic mutation, making accurate symptom evaluation critical.

There are FDA-approved therapies to treat the symptoms of MWS.