



Overview of Autoinflammatory Diseases

Your immune system is the network of cells and tissues throughout your body that work together to defend you from viruses, bacteria, and infection. It tries to identify, kill, and eliminate the invaders that might hurt you.

Parts of the immune system include:

- **Acquired (or adaptive) immune system**, which develops as you grow. Invaders provoke your body into producing antibodies so that your body “remembers” those invaders. Your body can then fight them if they come back.
- **Innate (or inborn) immune system**, which uses white blood cells instead of antibodies to destroy invaders.

What happens in autoinflammatory diseases?

Autoinflammatory diseases refer to problems with the innate immune system’s reactions. Immune cells target the body’s own healthy tissues by mistake, signaling the body to attack them. This can cause intense episodes of inflammation that result in such symptoms as fever, rash, or joint swelling. These diseases also carry the risk of amyloidosis, a potentially fatal buildup of a blood protein in vital organs.

Types of Autoinflammatory Diseases

Some examples of autoinflammatory disease and their symptoms include:

- **Familial Mediterranean Fever (FMF)**, which causes recurring bouts of fever. Other symptoms include:
 - Severe abdominal pain due to inflammation of the stomach cavity (peritonitis).
 - [Arthritis](#) (painful, swollen joints).
 - Chest pain from inflammation of the lung cavity (pleurisy).
 - Skin rashes.
- **Neonatal Onset Multisystem Inflammatory Disease (NOMID)** affects the skin, joints, eyes,

and central nervous system. For most children, the first sign of the disease is a rash that develops within the first six weeks of life. Other problems can follow, including:

- Fever.
- Meningitis.
- Joint damage.
- Vision loss.
- Hearing loss.
- Mental retardation.

- **Tumor Necrosis Factor Receptor-Associated Periodic Syndrome (TRAPS)** is associated with:

- Long, dramatic, episodes of high fever.
- Severe pain in the abdomen, chest, or joints.
- Skin rash.
- Inflammation in or around the eyes.

- **Deficiency of the Interleukin-1 Receptor Antagonist (DIRA)** can cause the following serious conditions in children:

- Swelling of bone tissue.
- Bone pain and deformity.
- Inflammation of the layer of connective tissue around bone.
- Skin rash that can cover most of the body.

- **Behçet's disease** can cause the following symptoms:

- Mouth or genital sores.
- Redness and swelling in the eyes.
- Arthritis.
- Skin problems.
- Swelling of the digestive system, brain, and spinal cord.

- **Chronic Atypical Neutrophilic Dermatositis With Lipodystrophy and Elevated Temperature (CANDLE)** can include the following symptoms, which generally develop during the first year of life:

- Recurrent fevers.
- Purpura.
- Joint pain.
- Contractures.
- Developmental delay.

- Facial changes, including a loss of fat on the face, and swollen lips and eyelids.

Causes of Autoinflammatory Diseases

Autoinflammatory diseases are typically caused by changes in certain genes. This causes problems with proteins that are important in specific body functions. For some diseases, the cause is unknown.

Treatment of Autoinflammatory Diseases

Treatments for autoinflammatory diseases can involve medications that:

- Reduce inflammation.
- Suppress the immune system.

Research Progress Related to Autoinflammatory Diseases

Researchers continue to explore the genetics and causes of autoinflammatory diseases. Understanding the causes of these diseases will improve both diagnosis and treatment.

For More Info

U.S. Food and Drug Administration

Toll free: 888-INFO-FDA (888-463-6332)

Website: <https://www.fda.gov>

Drugs@FDA at <https://www.accessdata.fda.gov/scripts/cder/daf>. Drugs@FDA is a searchable catalog of FDA-approved drug products.

Centers for Disease Control and Prevention, National Center for Health Statistics

Website: <https://www.cdc.gov/nchs>

National Institute of Allergy and Infectious Diseases

Website: <https://www.niaid.nih.gov/diseases-conditions/autoinflammatory-diseases>

American Autoimmune Related Diseases Association (AARDA)

Website: <https://www.aarda.org>

If you need more information about available resources in your language or other languages,

please visit our webpages below or contact the NIAMS Information Clearinghouse at NIAMSInfo@mail.nih.gov.

- [Asian Language Health Information](#)
- [Spanish Language Health Information](#)