Hyperimmunoglobulin D Syndrome (HIDS)

What is HIDS?
Hyperimmunoglobulin (hi-PER-im-MU-no-glob-U-lin) D Syndrome, also known as HIDS, is a genetic condition. The most serious type of this condition starts at birth and is called mevalonic aciduria (MEV-a-lon-ic AS-ed-UR-ia).

What are the symptoms of HIDS?
Symptoms usually begin in the first year of life. Symptoms come and go in episodes or attacks of:
• Swollen lymph nodes in the neck and other parts of the body
• High fever
• Skin rash
• Mouth sores
• Stomach pain
• Joint pain and swelling
• Throwing up
• Diarrhea

Babies with HIDS may develop:
• Life-threateningly high fevers
• Poor growth
• Developmental delay
• Trouble with vision
• Kidney damage

What causes HIDS?
HIDS is not contagious. It is caused by a genetic defect in the gene that tells the body to make a protein called mevalonate kinase (MVK). MVK speeds up chemical reactions in the body. It is used in making cholesterol.

The more changes that are made to the MVK gene, the more serious the condition tends to be. Doctors do not know why the gene defect causes fever. Infection, stress, trauma, surgery and vaccination tend to trigger a fever. The weather does not affect symptoms of HIDS.

HIDS can be passed down from parents (inherited). Parents can be carriers without having the condition.
Who gets HIDS?

HIDS is very rare. HIDS is mostly found in Western Europe, especially in the Netherlands and France. Males and females are both affected, as well as all ethnic groups.

How is HIDS diagnosed?

It can take a long time to make a correct diagnosis. At first, your doctor will use your child’s symptoms and a physical exam. HIDS is then diagnosed with tests, including:

- Urine and skin tests: Abnormally high mevalonic acid can be found in your child’s urine when they have symptoms. Special laboratories can measure the amount of mevalonate kinase enzyme (MVK) in blood or skin cells.
- Genetic tests: A test that looks at genes in your child’s DNA. This will show if there is damage to the MVK genes (mutations).
- Blood tests: Signs of inflammation can often be found when your child has symptoms. Inflammation is a physical condition in which part of the body becomes reddened, swollen, hot, and often painful, especially as a reaction to injury or infection.
- Most children have high amounts of proteins called immunoglobulins (especially type D), which are used by the immune system to fight infections. Testing for this is done at the same time as the urine and skin tests for MVK.

How is HIDS treated?

There is no cure for HIDS, but we use medicine to treat the health problems that often affect people with HIDS. These medicines may help while your child is having symptoms:

- Anti-inflammatory drugs like ibuprofen, naproxen or steroids
- Statins (a type of medicine that will lower cholesterol), such as simvastatin
- Biologic treatments that work against proteins that cause inflammation, such as tumor-necrosis factor (etanercept) or interleukin-1 (anakinra). Anakinra is often used at the start of symptoms.

These medicines are not always completely successful at treating symptoms, but all of them seem to help. There is not enough evidence to know if other treatments, such as a change in diet, probiotics, or acupuncture are effective.

Does HIDS go away?

HIDS does not go away. It is a lifelong condition. Symptoms usually get milder and less frequent over time, sometimes ending in adulthood. Episodes of symptoms can upset normal family life, like work and school. Successful treatment of symptoms can make this is less of a problem.
Talk to your child’s teachers about HIDS. Let them know that it is not contagious and your plan if an episode of symptoms starts at school. Your child will still be able to play sports if they would like.

Your child can develop a problem from inflammation called amyloidosis (am-uh-l oid-O-sis). Amyloid is a protein that harms organs in children with chronic inflammatory disease, most commonly the kidney, but also the intestines, liver, skin and heart. Children being treated should have blood and urine tests at least twice a year to monitor for amyloid.

**Can my child be vaccinated?**

Yes, your child can and should be vaccinated, even though this may cause fever attacks.

**How will HIDS affect my child later in life?**

Children with HIDS can grow up to enjoy healthy, productive lives. They can have normal sexual activity and have children of their own. During pregnancy, symptoms of HIDS tend to decrease. HIDS is rare, so it is not likely that your child’s partner will also carry HIDS. When your child’s partner is not a carrier of HIDS, their children will most likely not develop HIDS.

**Where can I learn more?**

If you would like more information, these websites are a place to start. We do not endorse or maintain these websites, and we are 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.

The Rheumatology Research Foundation  
www.rheumatology.org/Foundation

The Centers for Disease Control  
www.cdc.gov

The National Institutes of Health  
ghr.nlm.nih.gov/condition/mevalonate-kinase-deficiency