**Mevalonate Kinase Deficiency/ Hyperimmunoglobulin D Syndrome MKD/HIDS**

**CONDITION DESCRIPTION**
Mevalonate kinase deficiency (MKD), also known as hyperimmunoglobulin-D syndrome (HIDS), is a very rare, genetic disease that causes recurrent episodes of fever and other symptoms. It is caused by an abnormal gene, mevalonate kinase (MVK), and is mostly found in children in Western Europe. MKD affects both boys and girls, and symptoms usually appear in infancy.

**SIGNS/SYMPTOMS**
Recurrent episodes of fever are the main symptom of MKD. They may last for 3 to 7 days and recur every 2-12 weeks. Episodes begin suddenly, usually with chills. Children often have headaches, abdominal pain, appetite loss, flu-like symptoms, nausea, vomiting and diarrhea. Skin rashes all over the body, including painful mouth sores, may occur. Children may have joint pain and swelling, or muscle aches. Swollen lymph nodes are a striking feature. Diagnosis of MKD must be made by a physician with expertise in this type of disease. During an episode, blood tests may show high levels of inflammation. Most children will test for high levels of immunoglobulin-D. During episodes, urine tests will show high levels of mevalonic acid. A genetic test to show the abnormal MVK gene will confirm the diagnosis.

**COMMON TREATMENTS**
There is no cure for MKD, but treatments may help control inflammation and symptoms during episodes. Children may take nonsteroidal anti-inflammatory drugs (NSAIDs) like ibuprofen (Advil, Motrin) or naproxen (Aleve, Naprosyn), or corticosteroids during an episode. Biologic medications that block tumor necrosis factor or interleukin-1 have been found to be effective treatments to reduce the frequency of attacks. Anakinra (Kineret) may be used “on-demand” when an episode starts. Canakinumab (Ilaris) and etanercept (Enbrel) may be used regularly to prevent attacks in patients with more severe disease.

**CARE/ MANAGEMENT TIPS**
MKD episodes usually get milder and less frequent as children grow up. However, children may miss many days of school due to frequent episodes. It is important to stress to other children and classmates that the fevers related to MKD are not contagious. Patients rarely may develop a complication of inflammation called amyloidosis, but this is extremely rare in the United States, and it is usually prevented by appropriate treatment. Regular medical check-ups to monitor for amyloidosis and other potential problems are important.