

Periodic Fever in MVK Deficiency: A Patient Initially Diagnosed With Incomplete Kawasaki Disease

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KEY WORDS

auto inflammatory disease, fever of unknown origin, hyperimmunoglobulin D syndrome (HIDS), mevalonate kinase, mevalonic acid, Kawasaki syndrome

ABBREVIATIONS

HIDS—hyperimmunoglobulin D syndrome

IgD—immunoglobulin D

IVIg—intravenous immunoglobulin

MKD—mevalonate kinase deficiency

MVK—mevalonate kinase

NSAID—nonsteroidal antiinflammatory drug

Dr Thors gathered the relevant clinical information, and drafted the initial manuscript and subsequent revisions of the manuscript; Dr Vastert conceptualized the report, provided crucial information on the clinical presentation and follow-up, and reviewed and revised the manuscript; Drs Wulffraat and de Koning conceptualized the report and reviewed and revised the manuscript; Drs van Royen and Frenkel reviewed and revised the manuscript; and Mrs de Sain van der Velden conceptualized the report, provided the data from and was responsible for the metabolic evaluation, and reviewed and revised the manuscript. All authors approved the final manuscript as submitted.

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abstract

Mevalonate kinase deficiency (MKD) is a rare autosomal recessive disorder causing 1 of 2 phenotypes, hyperimmunoglobulin D syndrome and mevalonic aciduria, presenting with recurrent fever episodes, often starting in infancy, and sometimes evoked by stress or vaccinations. This autoinflammatory disease is caused by mutations encoding the mevalonate kinase (*MVK*) gene and is classified in the group of periodic fever syndromes. There is often a considerable delay in the diagnosis among pediatric patients with recurrent episodes of fever. We present a case of an 8-week-old girl with fever of unknown origin and a marked systemic inflammatory response. After excluding infections, a tentative diagnosis of incomplete Kawasaki syndrome was made, based on the finding of dilated coronary arteries on cardiac ultrasound and fever, and she was treated accordingly. However, the episodes of fever recurred, and alternative diagnoses were considered, which eventually led to the finding of increased excretion of mevalonic acid in urine. The diagnosis of MKD was confirmed by mutation analysis of the *MVK* gene. This case shows that the initial presentation of MKD can be indistinguishable from incomplete Kawasaki syndrome. When fever recurs in Kawasaki syndrome, other (auto-)inflammatory diseases must be ruled out to avoid inappropriate diagnostic procedures, ineffective interventions, and treatment delay. *Pediatrics* 2014;133:e461–e465

Mevalonate kinase deficiency (MKD) is an autosomal recessive disease resulting from mutations in the *MVK* gene, which encodes the enzyme mevalonate kinase (MVK). This enzyme is involved in the synthesis of isoprenoids and cholesterol, and mutations in the gene are the underlying cause of both the milder phenotype known as hyperimmunoglobulin D syndrome (HIDS; OMIM #260920) and the more severe phenotype, mevalonic aciduria (OMIM #610377).^{1–3} HIDS is 1 of the hereditary autoinflammatory syndromes, characterized by inflammatory attacks generally starting in infancy with fever as the primary symptom. In HIDS, symptoms typically start at a median age of 6 months, and patients experience repeated febrile attacks into adulthood. In childhood, the febrile attacks usually occur every 4 to 6 weeks, but as the patients grow older, the intervals between attacks tend to be prolonged.⁴ Fever can be accompanied by a variety of symptoms such as lymphadenopathy, skin lesions, arthralgia, and arthritis, as well as abdominal pain and diarrhea.^{4–6} In patients with HIDS, the fever attacks typically last 3 to 6 days and can be provoked by vaccinations, minor trauma, or emotional stress. Sporadically, episodes can last longer, even up to several weeks. Incidentally, oral or vaginal ulcers are reported.⁷ In general, a strong acute-phase response is seen with significantly elevated C-reactive protein, sometimes combined with elevated levels of interleukin-1, interleukin-6, and tumor necrosis factor in serum.^{4,8}

Infants and young children with fever of unknown origin are frequently referred to hospitals for diagnostic evaluation. The differential diagnosis is broad, and the diagnosis of MKD can be challenging. Diagnosis of MKD is therefore often delayed, with some series reporting a median delay of up to 10 years.⁹ In children, an elevated level of serum immunoglobulin D (IgD) is neither sensitive nor specific for the diagnosis

of MKD.^{3,4} The diagnosis of MKD can be confirmed by measuring increased excretion of mevalonic acid reflecting the reduced activity of the MVK enzyme^{1–4} and by the finding of mutations in the *MVK* gene. Many mutations have been recognized, 4 of which are responsible for >70% of all cases.^{1,2} These gene mutations result in an 85% to 95% reduction of MVK enzyme activity.

We report a case of a young female patient admitted to our hospital for evaluation of fever of unknown origin. She was initially diagnosed and treated for incomplete Kawasaki syndrome, but at a later stage, after recurrence of fever attacks, it turned out that the correct diagnosis was HIDS.

CASE REPORT

An 8-week-old white girl, first child of non-consanguineous parents of Dutch origin with an unremarkable medical history, presented at our hospital with fever for a total of 6 days without a clear origin. Fever persisted despite administration of broad-spectrum antibiotics (amoxicillin + clavulanic acid and gentamicin). The fever was accompanied with signs of mild purulent conjunctivitis and redness around the umbilical area, resembling omphalitis, and she seemed to develop subtle changes in consciousness during fever peaks, which led to referral to our hospital. The child had not yet had the first immunization when the fever commenced.

Physical examination on admission revealed an irritable infant with a temperature of 38.8°C. Other findings were a purulent conjunctivitis and redness surrounding the umbilical area. There was desquamation of the lips and a slightly enlarged liver. A grade 2/6 cardiac murmur was detected through auscultation of the heart. There were no remarkable changes in the extremities or skin, and no apparent lymphadenopathy. Laboratory evaluation showed strongly elevated inflammatory

parameters (C-reactive protein: 252 mg/L; erythrocyte sedimentation rate: >140 mm/h), anemia (hemoglobin: 4.4 mmol/L or 6.6 mg/dL), and an elevated leukocyte count ($23.7 \times 10^9/L$) with a predominance of neutrophils and thrombocytes of $543 \times 10^9/L$. Serum transaminases, creatinine, ferritin, and standard coagulation test results were normal. Using culture, polymerase chain reaction, and serology samples, no evidence of viral or bacterial infections were found in blood, cerebrospinal fluid, or urine.

Because of the persistent unexplained fever with evidence of severe systemic inflammation, treatment of incomplete Kawasaki syndrome was initiated according to international guidelines with intravenous immunoglobulin (IVIg; 2 g/kg in 24 hours) and acetylsalicylic acid (80 mg/kg daily)¹⁰ while awaiting cardiac ultrasound. Cardiac ultrasound performed on the fourth day of admission (day 10 after the onset of fever) revealed mild dilation of the right (maximum diameter: 2.0 mm; z score: 2.6) (Fig 1) and left (maximum diameter: 2.1 mm; z score: 2.2) coronary artery.

Initially, 36 hours after completion of the IVIg, the fever resolved and there were clear signs of clinical recovery. However, 2 days later, the fever reappeared and a second dose of IVIg was given while the patient was still on a high dose of acetylsalicylic acid. She again responded well, with normalization of inflammatory parameters, and the fever subsided. She was discharged in good clinical condition. During follow-up, a repeat ultrasound of the heart was performed showing normalization of the z score of the coronary arteries; this finding was confirmed a few months later (Table 1).

Six weeks after discharge, the patient again developed a fever, this time after vaccination against diphtheria-tetanus-pertussis-poliomyelitis, *Haemophilus influenzae*, and pneumococci. In the

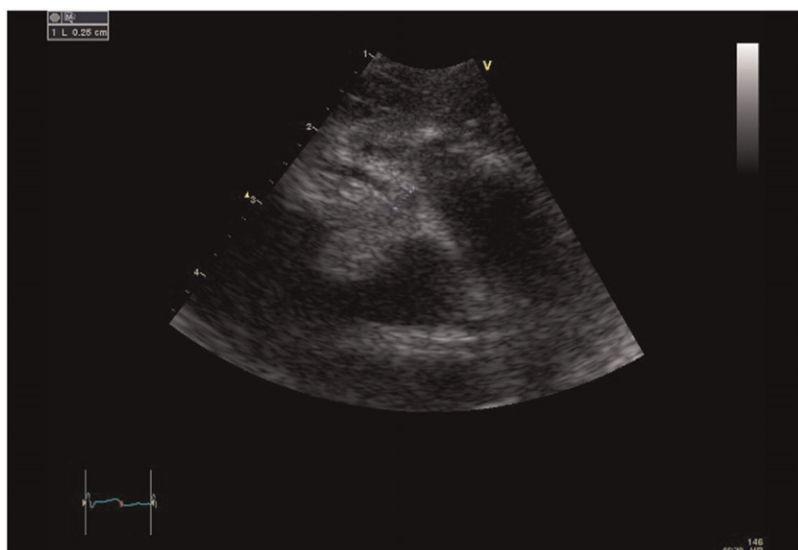


FIGURE 1

Echocardiogram of the patient showing the dilated right coronary artery measuring a maximum of 2.6 mm (average of 2.0 mm) and a z score of 2.6 based on international standards.^{24,25}

TABLE 1 Coronary Artery Measurements

Time Point	BSA	RCA, mm	z Score	LCA, mm	z Score
Presentation	0.29	2	2.6	2.1	2.2
6-wk follow-up	0.30	1.8	1.9	1.9	1.6
6-mo follow-up	0.33	2	1.6	2	1

BSA, body surface area; LCA, left coronary artery; RCA, right coronary artery.

subsequent months, she continued to present regularly with episodes of fever accompanied by various symptoms such as bloody diarrhea, malaise, and erythematous skin lesions. The occasional bloody stools were judged to be a part of an inflammatory response or nonsteroidal antiinflammatory drug (NSAID) treatment, and no colonoscopy was performed. Interestingly, at cardiologic follow-up, the dilation of the proximal coronary arteries had spontaneously resolved within 5 months after the initial presentation. Episodes of fever persisted, characterized by abdominal pain and fatigue, recurring every 4 to 6 weeks and lasting for 3 to 5 days. This finding led to the suspicion of a periodic fever syndrome such as cryopyrin-associated periodic syndrome or MKD, warranting further investigations. The urinary mevalonic acid excretion was strongly elevated (13.1

$\mu\text{mol}/\text{mmol}$ creatinine [normal: $<0.8 \mu\text{mol}/\text{mmol}$ creatinine]) as was plasma mevalonic acid (1.12 $\mu\text{mol}/\text{L}$ [normal: $<0.034 \mu\text{mol}/\text{L}$]). Mutation analysis of the *MVK* gene revealed that the girl was compound heterozygote for 2 mutations, c.803T>C (p.Ile268Thr) and c.1129G>A (p.Val377Ile), known to be associated with the HIDS phenotype. Genotyping of both parents revealed that both parents carried a heterozygous mutation of the *MVK* gene: mother, c.803T>C (p.Ile268Thr); father, c.1129G>A (p.Val377Ile).

The patient was initially treated for HIDS by using supportive therapy, NSAIDs, and acetaminophen, but because of frequent episodes of fever in combination with debilitating symptoms of severe fatigue and discomfort during the fever episodes, we started treatment with a recombinant interleukin-1 receptor antagonist (anakinra [Kineret, Swedish Orphan Biovitrum, Stockholm,

Sweden]) in a prophylactic regimen. Unfortunately, this regimen did not significantly affect the severity or the frequency of the fever attacks. Anakinra was stopped within 12 weeks. A 6-month period of prophylactic treatment with anti-tumor necrosis factor (etanercept) did not result in less frequent or less severe attacks. Therefore, she was switched again to treatment with NSAIDs and acetaminophen. The child has had no considerable infectious episodes since the first admission.

DISCUSSION

Diagnosing periodic fever syndromes such as HIDS can be a challenge for physicians because of the rarity of the disorders and the considerable overlap with other inflammatory syndromes and common infections. This difficulty is underlined by the fact that considerable delay exists before the diagnosis is made, as was shown in a retrospective analysis of 103 patients with confirmed HIDS.⁹

Kawasaki syndrome is a well-known entity in young children. The hallmark of this disease is prolonged fever without localizing signs as seen in HIDS, as well as the presence of clear-cut diagnostic symptoms that serve as diagnostic criteria. However, some of these symptoms, including rash, lymphadenopathy, and mucosal changes, also occur in HIDS. Moreover, in a minority of patients with Kawasaki syndrome, especially infants,¹¹ not all principal criteria are met. Patients with Kawasaki syndrome are at risk for coronary artery complications.¹² To decrease the risk of cardiac or coronary complications, patients with Kawasaki disease should receive IVIg and high-dose met. Moreover, in a minority of patients with Kawasaki syndrome, especially infants,¹¹ not all principal criteria are met. Patients with Kawasaki are at risk for coronary artery complications.¹² To decrease the risk of cardiac or coronary complications, patients with

Kawasaki disease should receive IVIg and high-dose aspirin before the 10th day of fever.^{13,14}

The finding of dilated coronary arteries in a febrile child is considered to be a good indicator of Kawasaki syndrome, as recommended by the American Heart Association and supported by Bratincsak et al.^{15,16} A comparison was made between 145 children with Kawasaki syndrome and 45 children with fever (36 with infectious illness and 9 with a self-limiting febrile condition); they found no children in the non-Kawasaki syndrome group with a coronary artery z score of ≥ 2.5 and a significant difference in overall z scores between the groups. In our patient with prolonged fever without an identifiable cause, the suspicion of incomplete Kawasaki syndrome was supported by the finding of mildly dilated proximal coronary arteries, which led to the clinical decision to treat with IVIg. It is important to note that coronary artery involvement is not confined to Kawasaki syndrome but has been documented in some other inflammatory syndromes, especially vasculitis (microscopic polyangiitis or polyarteritis nodosa)¹⁷ and systemic

onset juvenile arthritis.¹⁸ One can speculate that the dilated coronaries in our patient were part of a systemic inflammatory response seen in other autoinflammatory syndromes. To our knowledge, this is the first report of a patient with MKD and signs of dilation of the coronary arteries. The pathogenesis behind these findings is unclear.

Although there is no evidence-based therapy for MVD, NSAIDs and short courses of steroids reportedly improve symptoms. Moreover, case reports mention that biologic agents, especially anakinra (interleukin-1 receptor antagonist) and etanercept (tumor necrosis factor- α inhibitor),^{19,20} can be beneficial. Alternatively, in a small trial of adult patients, treatment with simvastatin led to a considerable reduction in febrile days compared with placebo controls.²¹

The term “hyper-IgD syndrome” was originally given to this condition because elevated levels of IgD were observed in patients with periodic fever, frequently associated with elevated immunoglobulin A levels in >80% of cases.⁵ However, a multicenter, international report found that 22% (of

103 patients with HIDS) had normal IgD levels, although IgD levels tended to rise as patients grew older.^{22,23} Indeed, our patient had normal levels of IgD (15 mg/L).

CONCLUSIONS

In young infants, incomplete Kawasaki syndrome can be diagnosed based on clinical symptoms of fever and coronary artery dilation. A first episode of suspected Kawasaki syndrome, regardless of whether it was complete, should be managed according to existing guidelines without delay. However, because Kawasaki syndrome is a clinical diagnosis, and coronary artery dilation can also be present in some other systemic inflammatory diseases, recurrence of symptoms after the diagnosis of Kawasaki syndrome should raise the suspicion of MKD or other autoinflammatory syndromes or vasculitis (ie, polyarteritis nodosa).

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