

Fever study aims to get faster treatment for children

Periodic fever syndromes represent a group of rare, inflammatory disorders that often begin in childhood. In most affected children, they present as a seemingly unprovoked fever and severe inflammatory state. Many symptoms resolve with each event, but certain types of fever syndrome can have damaging effects such as renal amyloidosis, hearing loss, and arthritis. In rare cases, these complications can lead to death.

While the disease is rare, effective treatments are often available that can improve symptoms and at times prevent or even reverse organ damage. A study launched in September under the Canadian Paediatric Surveillance Program intends to gather new data and raise awareness in the medical community, so that patients can get on the path to appropriate treatment sooner.

“The pathway to a diagnosis can understandably be very frustrating for parents, as children will often be seen by multiple physicians before a correct diagnosis is made,” said Dr. Paul Dancy, a paediatric rheumatologist and Associate Professor of Pediatrics at Memorial University of Newfoundland. Two key questions for the study are how these disorders present, and how much time typically elapses from symptom onset to diagnosis.

‘Autoinflammatory’ conditions a new category

Dr. Dancy noted that patients displaying some of the symptoms that accompany the fever—such as rashes, arthritis or cardiac issues—may at first be thought to have an autoimmune disorder.

“We know that many autoimmune conditions, such as juvenile arthritis and lupus, can present with fever,” he explained. “But over the last decade we’ve learned of a new category of illness—autoinflammatory—which causes fever and other disease manifestations but without the autoantibodies normally linked to autoimmune diseases.” The periodic fever syndromes are an example of this kind of process. In many cases, a genetic test is available to help confirm a diagnosis.

Many paediatricians will have seen the more common form known as “PFAPA” (*see sidebar for details*), Dr. Dancy added. When children with fever and inflammation are assessed, he said, it is important that physicians rule out causes, such as infections. While a periodic fever syndrome is “certainly not at the top of the differential, it should be on that list, particularly when a recurrent pattern is noted.”

Is it a periodic fever syndrome?

Patients must have one of the following diagnoses:

- Familial mediterranean fever (FMF)
- Tumor necrosis factor receptor associated periodic syndrome (TRAPS)
- Hyperimmunoglobulin D syndrome (HIDS)
- Cryopyrin associated periodic syndromes (CAPS) including familial cold autoinflammatory syndrome (FCAS), Muckle-Wells syndrome (MWS), and neonatal-onset multisystem inflammatory disease (NOMID)
- Periodic fever, aphthous stomatitis, pharyngitis and adenitis (PFAPA)
- Periodic fever syndrome – undefined

For the exclusion criteria and other details, see the study protocol at: www.cps.ca/English/surveillance/cpsp/Studies/current_concluded.htm.

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