

# The Challenge and Untold Burden of Periodic Fever Syndromes

Periodic Fever Syndromes comprise a cluster of diseases recognized as part of the expanding group of autoinflammatory disorders<sup>1,2</sup>

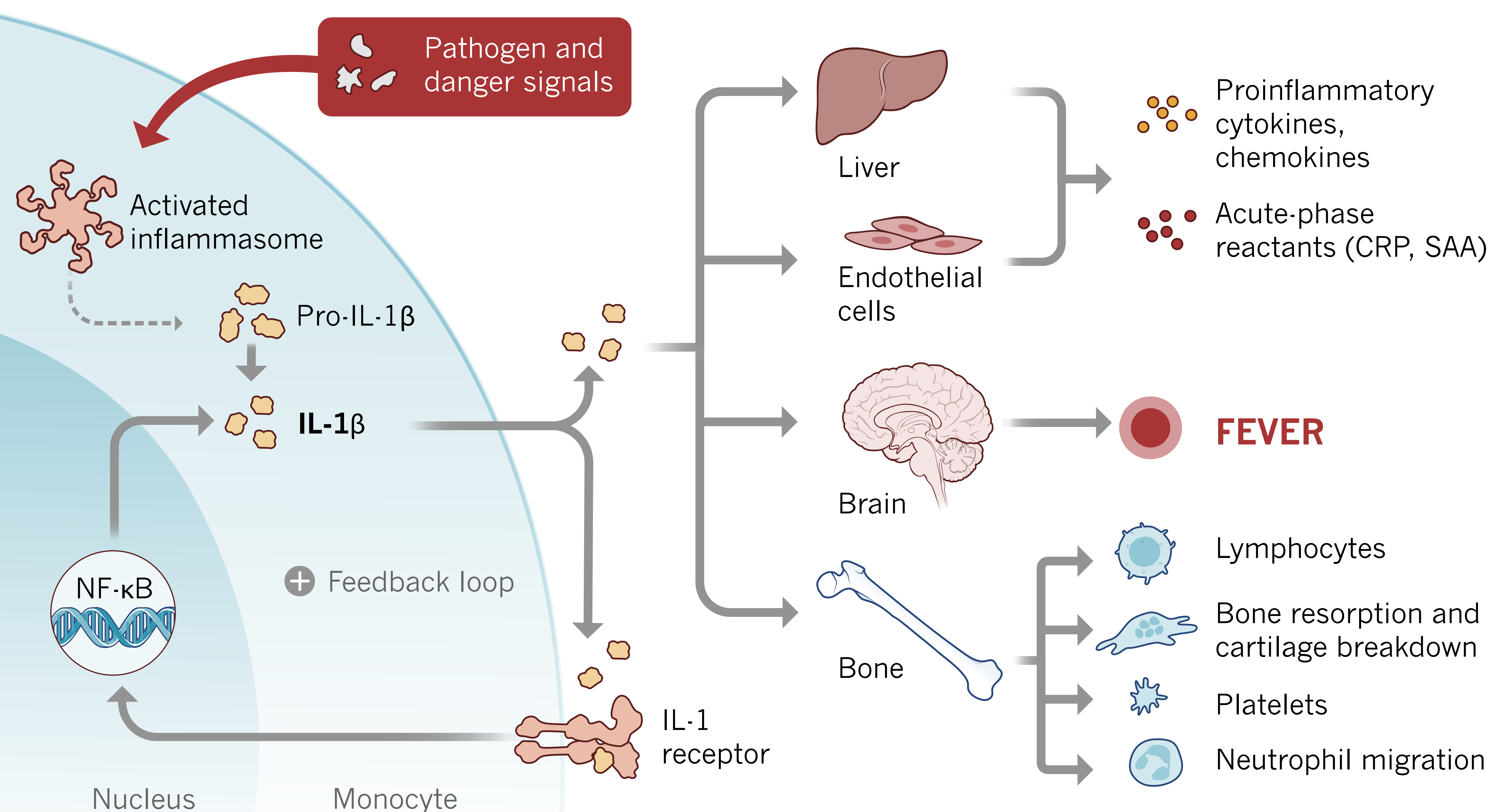


Figure adapted from Lachmann 2011

The 3 classic syndromes are **familial Mediterranean fever (FMF)**, **tumor necrosis factor receptor–associated periodic syndrome (TRAPS)**, and **hyperimmunoglobulinemia D with periodic fever syndrome (HIDS/MKD\*)**<sup>1,2</sup>

Hyperactivation of the innate immune system leads to **recurrent episodes of systemic inflammation** with hallmark features of high fever, rash, and joint symptoms; amyloid A amyloidosis is a serious long-term complication, often leading to renal failure<sup>3,4</sup>

**Interleukin-1β (IL-1β)** is a potent proinflammatory cytokine produced by innate immune cells that plays a prominent role in Periodic Fever Syndromes pathogenesis.<sup>1,2</sup> It elicits a range of inflammatory responses, including production and release of acute phase proteins, induction of fever, stimulation of bone resorption and cartilage breakdown, and roduction/activation of lymphocytes, neutrophils, and platelets.<sup>1</sup>

**Currently no approved treatments for TRAPS, HIDS/MKD, and colchicine-resistant (cr)FMF<sup>†</sup> exist**

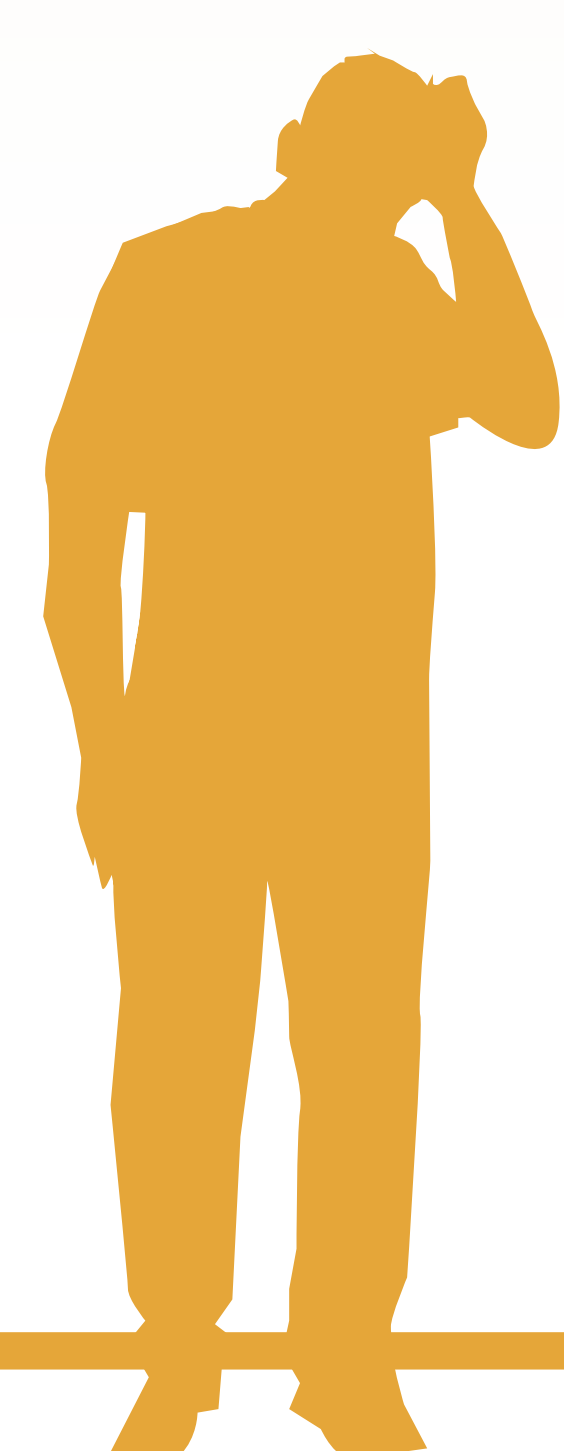
## Understanding the Human Burden of Periodic Fever Syndromes



**FMF**

**ABBAS, 38**

“ I can never plan to do anything with my family because I’m embarrassed to say I am sick again<sup>‡</sup> ”



Patients are more likely to suffer from **depression** and **anxiety**<sup>5</sup>



**TRAPS**

**SOPHIE, 21**

“ I used to be upset a lot because I just didn’t know what it was. Obviously I’d have to take a lot of time out of school. I didn’t understand why I had to go to hospital all the time<sup>‡</sup> ”



Takes a **physical toll** on children and an **emotional toll** on their parents<sup>6</sup>



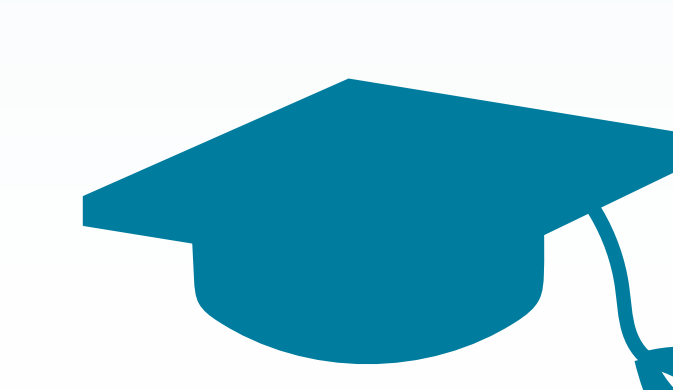
**HIDS/MKD**

**CAREGIVER FOR ASHLEY, 21**

“ I remember breaking down a few times and saying: I can’t deal with this anymore<sup>‡</sup> ”



**27%** of patients are **unemployed**<sup>7</sup>



**46%** of patients **delay their education**<sup>7</sup>

CRP, C-reactive protein; SAA, serum amyloid A. \*Also known as mevalonate kinase deficiency; †A subset of FMF patients are resistant or intolerant to colchicine, the only approved treatment.<sup>8,10</sup> ‡From patient experience recorded during Novartis market research, 2013. NOTE: Patient photos are representative and not actual patients.

**References** 1. Lachmann HJ, et al *Arthritis Rheum.* 2011;63:314-24. 2. Gattorno M, et al *Arthritis Rheum.* 2013;65:1137-47. 3. ter Haar N, et al *Ann Rheum Dis.* 2013;72:678-85. 4. Simon A, et al. In: Firestein G, et al, eds *Kelley’s Textbook of Rheumatology*. 8th ed. Philadelphia, PA: Saunders; 2008: 1863-82. 5. Deger SM, et al *Rheumatol Int.* 2011;31:623-8. 6. Pelagatti MA, et al *Arthritis Rheum.* 2011;63:1141-50. 7. van der Hilst JC, et al *Medicine.* 2008;87:301-10. 8. Özen S, et al *J Rheumatol.* 2011;38:516-8. 9. Colcris® (colchicine) Prescribing Information. Takeda Pharmaceuticals America, Inc., 2012. 10. Ozturk MA, et al *Clin Exp Rheumatol.* 2011;29(suppl 67):S77-86.