Supplementary Table 1. Components and weights of the genotype-based M694V prediction score, as defined in the derivation study [Tunce et al. (11)].

Predictor variable	Assigned points
Splenomegaly	10
Erysipelas-like erythema	9
Arthritis	8
Disease onset ⩽3 years	3
Myalgia	2
Protracted febrile myalgia syndrome	6
Family history of FMF	2
Total possible score	**40**

Each variable is scored dichotomously (0 = absent, 1 = present); the weighted total ranges from 0 to 40 points. FMF, Familial Mediterranean Fever.

Supplementary Table 2. Genotype distribution of group 2 (other biallelic exon 10 variants; n = 192).

Genotype	n (%)
M694V/M680I	66 (34.4)
M694V/V726A	43 (22.4)
M680I/V726A	21 (10.9)
M694V/R761H	17 (8.9)
M680I/M680I	17 (8.9)
V726A/V726A	6 (3.1)
M680I/R761H	5 (2.6)
M694V/M694I	4 (2.1)
V726A/R761H	4 (2.1)
M694V/K695R	3 (1.6)
M680I/M694I	1 (0.5)
M694I/M694I	1 (0.5)
M680I/A744S	1 (0.5)
V726A/K695R	1 (0.5)
A744S/A744S	1 (0.5)
M694V/A744S	1 (0.5)
Total	192 (100)

Group 2 comprised 16 distinct biallelic exon 10 genotypes; the three most frequent were M694V/M680I (34.4%), M694V/V726A (22.4%), and M680I/V726A (10.9%). The gene name MEFV is italicized in the main text; variant designations are shown in regular font.

Supplementary Table 3. Score performance in the full cohort vs. the non-overlapping subset.

Cohort	n	AUC (95% CI)	⩾9 Se/Sp	⩾12 Se/Sp	⩾15 Se/Sp
Full cohort	347	0.69 (0.64–0.75)	54.2/78.6	43.2/87.0	27.7/91.7
Non-overlapping	225	0.71 (0.64–0.77)	54.4/77.9	43.7/85.2	27.2/91.0

Se, sensitivity (%); Sp, specificity (%). The non-overlapping subset excludes the 122 patients who were included in the Tunce et al. (11) derivation submission, identified by matching patient initials and dates of birth. AUC: area under the curve; CI: confidence interval.