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Severity scoring system for paediatric FMF

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CORRESPONDENCE

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In a recent commentary in *Nature Reviews Rheumatology*, Livneh[1] discussed paediatric scoring systems for familial Mediterranean fever (FMF; Severity score system for paediatric FMF, *Nat. Rev. Rheumatol.* 8, 309-310; 2012) in relation to our recent manuscript on the topic.[2] We consider sharing our opinion with the scientific community as an ethical obligation and, therefore, would like to respond to the criticisms in the Livneh commentary.[1]

One of the key messages of our study was the lack of consistency between the current severity scoring tools for paediatric FMF. However, this finding would not necessarily make studies of severity scoring systems questionable. In this regard, Ozen *et al.*[3] examined the environmental effects on the severity of childhood FMF by adjusting Mor[4] and Pras[5] scoring systems to children, and did not compare the consistency of different scoring systems, but compared the results of each scoring system according to the country of residence. Ozen *et al.*[3] also demonstrated disagreement between the two scoring systems, although they did not specifically mention such finding. For example, the percentage of patients with severe disease in Turkey was 78.2% based on modified scoring system by Mor and colleagues. However, the percentage of patients with severe disease was only 34.5% according to modified Pras scoring system.

Another area of criticism in the Livneh commentary was the modification of the colchicine dose, in that it might have biased our results. The clinician's decision regarding the dose of colchicine administered for adults is primarily related to disease severity. However, age and weight are the main determinants of colchicine dosing in children. Therefore, it is evident that the amount of dosing might not be a direct indicator of disease severity for an infant as compared with an adult. Moreover, colchicine dose by itself as a measure of disease severity might not be the best choice in light of current knowledge of pharmacogenetics; increase in colchicine dose used can be due to the pharmacogenetic profile of an individual, rather than an increase in disease severity. In addition, there is a subgroup of patients who do not respond to colchicine but do respond to other therapies (such as as interleukin-receptor blockers). In this regard, severity scoring systems, irrespective of their modification status, will probably miss a patient with severe disease.

Lastly, we are pleased to hear Dr Livneh's support for an international consortium to establish a severity scoring system specifically for children with FMF. The distinct course of the disease in the paediatric age group decreases the utility of the scoring systems of Mor *et al.*[4] and Pras *et al.*,[5] which were originally developed for adults with FMF. There are many qualitative FMF researchers across the Mediterranean basin. We will be supportive of any initiative that aims to bring expert experiences together.

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Competing interests
The authors declare no competing interests.

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