Letters to the Editor

Familial Mediterranean fever and Behçet's disease

Sirs,

In the editorial on the association between familial Mediterranean fever (FMF) and Behçet's disease (BD) (1), Drs. Ben-Chetrit and Yazici bring to light the prevalence of BD in a healthy Moslem community in Israel (2) to support their notion that the prevalence of BD in Israel is high. The prevalence of 12:10,000 that is quoted, however, is still significantly lower than the 16: 4000 found by us, or the 2: 355 reported by Ben-Chetrit in FMF (3, 4). A recent finding of the Turkish FMF study group confirms the very high prevalence of BD in 2838 FMF patients studied ($p < 0.001$ compared to the general population) (5). I hope that this finding, which brings the population of FMF patients, screened to determine the rate of BD, to more than 7000, will terminate the debate and skepticism on this issue, and allow Drs. Ben-Chetrit and Yazici to face the facts. It should also be noted that the critical remarks on the absence of appropriate controls is just repeating the discussion of our original paper on the association between FMF and BD (6), where all the points raised in the editorial were addressed and answered one by one.

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Reply

Sirs,

In our agonising reappraisal to "face the facts", as Dr. Livneh has kindly asked us to do, we came across the following:

1. The initial report that described the BD-FMF association from Dr. Livneh's group was published in 1997, on the occasion of the first International Conference on FMF (1). The patient recruitment in this report was based on 3 sources:

a. A survey of a computerised database of about 4000 patients with FMF;

b. FMF-BD patients "known to the personnel" who worked in the unit the report originated from;

c. FMF-BD patients seen within 2 years of the manuscript's preparation – new patients or patients from the database who were seen during the same time period.

No numbers were provided for the patients in groups b or c.

2. This same study later became the substance of a peer-reviewed publication (2). At this stage the following can be noted.

a. The FMF-BD patient subgroup "known to our personnel" no longer exists.

b. The authors do not still give the numbers for the new patients with FMF seen during this period. What they do say is that they have seen 60% of their data base of 4000 patients over a time period of 2 years and examination of this group "confirmed that most of the BD patients in our FMF population were identified by the computer search".

c. There are two control groups, not reported in the initial work. One is a group of 100 consecutive FMF patients and the second consists of 29 BD patients; both groups were seen for follow-up during the 2-year period. These patients constitute the comperators for the clinical features of the authors' FMF-BD patients. The circular argument used in the selection of these control groups apparently does not bother the authors. If you pre-select a group of "pure" FMF patients and then compare their clinical findings with those from a group of FMF-BD patients from the same original pool of patients, it will be surprising only if you do not find some clinical differences between the two groups. This point, not strictly relevant to the issue of the frequency of BD among FMF patients, is nevertheless a good indicator of the quality of the data at hand.

3. Dr. Livneh cites a recent abstract (3) from Turkey published in this journal to back up his point that FMF and BD coexist to a high degree in Turkey as well. A statistic of $p < 0.001$ is quoted for the significance of the high prevalence of BD, i.e. 0.5%, among FMF patients in Turkey. It is not stated in the abstract what the comparator group was in the statistical analysis. Personal communication with the senior author reveals that the comparator group was represented by a survey of 46,813 children in Turkey among whom not a single case of Behçet's was found (4). On the other hand, we know from another abstract (5) in the same issue that the mean age of the FMF patients reported in reference 3 was 23 (range: 2-87 years). In other words, in addition to the paediatric cases, there were also hundreds of adult FMF patients in this report, rendering the quoted statistic rather meaningless.

These are some of the facts that make us not only sceptical about the soundness of the data at hand, but also quite surprised at the insistence of our correspondent to stand by them.

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Salivary gland lymphocytic infiltrates and *Helicobacter pylori* serology in anti-SSA/Ro positive patients in Italy

Sirs,

Sjögren's syndrome is a chronic autoimmune disorder involving the salivary and lacrimal glands with lymphocytic infiltration, glandular destruction and an increased