Sir, Touitou et al. [1] suggest that a central registry or database would serve both auto-inflammatory disease (AID) patients and their doctors well. There are three important issues related to what they propose. First, in their scheme, Behçet’s syndrome (BS) is taken as a bone fide auto-inflammatory condition. There are many reasons to say why this is problematic [2]. The prototype disorder for AID is FMF, a disease associated with mutations of the MEFV gene, and almost all conditions characterized as AID have a monogenic inheritance. This is not true for BS. On the other hand, panuveitis, extensive vasculitis, hypercoagulability and a disease course getting milder at later ages are uncommon among AID, but are the main features of BS. Another crucial difference between FMF and BS is the prolonged inflammatory skin response. Pathergy test, a non-specific response to skin trauma, is typically described in BS and in some neutrophilic dermatoses such as pyoderma gangrenosum and Sweet’s syndrome. No pathergy skin response is reported in FMF [3]. Similar to the pathergy test, skin responses to urate crystals are also described in BS, which is again a highly specific inflammatory response not observed in FMF [4]. Epidemiology also shows differences among the two. FMF and BS are common in the Middle East, but FMF is rare in the Far East where BS is common. There is no association of HLA B51 with FMF. Response to colchicine is nearly diagnostic for FMF, whereas it is much less impressive for BS [5]. Apart from every other consideration, 95% of all BS cases are seen in adults. So the inclusion of BS in this database would be rather incongruous.

Secondly, the database they plan to put together for this registry is aimed at highlighting the similarities between the conditions they propose to include in the registry, but what about the differences? It might be a useful exercise for the authors to consider the possible common characteristics of a group of, for example, systemic lupus patients with the other members of the planned registry of auto-inflammatory conditions.

Finally, the whole local approval/patient confidentiality issues requiring the collaboration of many patients/physicians/agents in different countries would make this a very challenging undertaking. This being a web-based database would multiply the problems with the confidentiality issue.

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References


Comment on: Web resources for rare auto-inflammatory diseases: towards a common patient registry: reply

Sir, Yazici and Yazici [1] raised three issues regarding our suggestion of the need for a central registry for patients with auto-inflammatory disease.

First, they assert that considering Behçet’s disease (BD) as an auto-inflammatory disease (AID) is incongruous. We would like to remind the authors of the work carried out in Turkey published in 2005 by Gul strongly advocating BD as an AID [2]. BD has been considered as belonging to the AID group since the first description of AIDs as diseases characterized by seemingly unprovoked inflammation, in which high-titre autoantibodies or antigen-specific T cells do not usually play a major aetiological role [3]. Although the prototype of AID is indeed the monogenic disease FMF, several multi-factorial conditions other than BD exist, which are considered at least partly as AIDs...