

A proposal for new taxonomy for AIDs

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Taxonomy is the practice and science of classification of things or concepts. Diseases are given names mainly for communication purposes. Names often give some data about the disease, its clinical features or its cause/origin (etiology), as well as possible clues for the therapeutic approach.

He showed how diseases are usually named. This can be by using the name of the physician who first described the disease (i.e. Sjogren's syndrome), the geographical area affected reflecting the spread of the disease (i.e. West Nile Fever) or the name of a major symptom or sign characteristic of the disease (i.e. foot & mouth disease). Some names describe the clinical features of a disease (i.e. PFAPA: **P**eriodic **F**ever, **A**phthosis, **P**haryngitis, **A**denitis). Others are named according to the geographical area of the disease's spread (i.e. Familial Mediterranean Fever); or named according to the name of the physician who first described them (i.e. Muckle-Wells syndrome).

In 2016, Prof. Ben Chetrit decided to start a project for defining SAIDs and giving them more appropriate names with the support and infrastructure of PRiNTO (**P**ediatric **R**heumatology **I**nter**N**ational **T**rials **O**rganization). They used the Delphi technique to reach a consensus among a group of experts. Delphi can be defined as a series of questionnaires involving several steps, each of which is based on the results of the previous step. After 6 rounds of questionnaires, in which suggestions were improved by taking into account the proposals and comments made previously by the 6 members, 100% consensus on all issues was reached. They then shared their suggestions with an additional 27 experts in the field of SAIDs, in order to gain larger support for the proposals or to make further comments for improvements.

The first step was to establish rules for naming diseases:

1. Try not to change the name whenever it is appropriate.
2. Avoid names of persons or geographical spread of the disease (eponyms)
3. Include the genetic basis (name of the gene) of the disease, where it is known (prefer the name of the gene over the name of the encoded protein unless the name of the gene is not accurate or meaningful)
4. Include key clinical features where appropriate
5. Shorten the name as much as possible
6. Choose a name that is as clear as possible
7. In diseases where our knowledge about the pathogenesis is still limited, leave the current name (i.e. PFAPA)
8. In diseases with different phenotypes but mutations in the same gene, use a general "roof" name with subtypes (i.e. PAAD, NOD2).
9. When the clinical features appear to be "continuous", give a general "roof" name and classify the various presentations according to their phenotypic severity (i.e. NLRP3- AID, MKD).
10. Keep the term "periodic" for attacks which have a regular pattern; otherwise, use the term "recurrent".

Prof. Ben-Chetrit then showed a summary of the above and showed some specific examples of the new naming convention and concluded that, the new recommended rules for nomenclatures of SAIDs should allow a better organization of these groups of immune diseases, help to understand their etiology or nature and improve communication with physicians from other disciplines.