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To all honourable
Members and collaborators of
the ESID Registry Working Party
(Physicians and Documentarists)

Graz, January 31st, 2022

Subject: Call to participate in a study on the disease activity and burden of immune dysregulation

Dear Colleagues and friends,

Herewith I would like to invite you to participate in a study titled:

Prospective monitoring of patients with primary immune dysregulation as evaluated by the IDDA2.1 kaleidoscope score

Importantly, the IDDA score has already been available as optional module (tab) in the level 1 at the initial and follow up patient entries and is designed for all IEI with immune dysregulation. Now, I am inviting your active contribution!

We will collect the data you entered of patients with known monogenic IEI with or without immune-modulating medication within a time frame of roughly two years (until end of 2023, prolongation possible). The score is calculated immediately upon entry of all parameters (, which should take <5min for those who know the patient); and there is a physicians' reporting sheet that may be used for settings, in which other people enter the data at a later time point. You may use the calculated score for your own documentation of the disease activity, while we will record all individual parameter scoring and treatment entries in the background of each time point (multiple desired). The only exclusion criteria are prior hematopoietic stem cell transplantation or gene therapy (no exclusion if HSCT or GT is performed during the study period; interesting for before/after comparisons).

We plan to assess potentially predictive values in diagnosis finding, complication monitoring, and to deduct phenotype-driven, "semi-targeted" therapy suggestions for undiagnosed patients. When taking this approach, we will apply unsupervised machine learning algorithms to detect similarities in patterns in training cohorts consisting of patients with known monogenic IEI

In case of a planned publication, entries will qualify the center representatives for a coauthorship in a contributors list of ESID registry working party members. Please do not hesitate to contact me in case of questions or suggestions.

THANK YOU in advance for your participation!
Sincerely yours

Markus Seidel

Attachments: IDDA2.1 physicians empty score sheet (.xls); example figures (see references)

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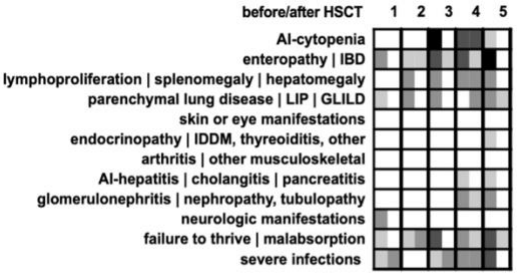
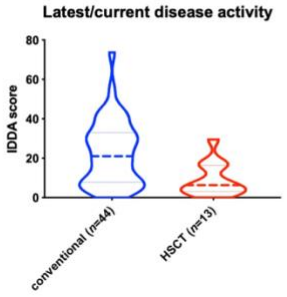
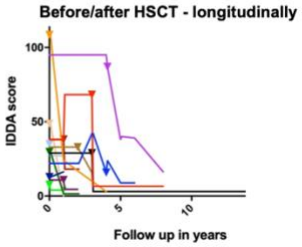
Example applications and figures, taken from:

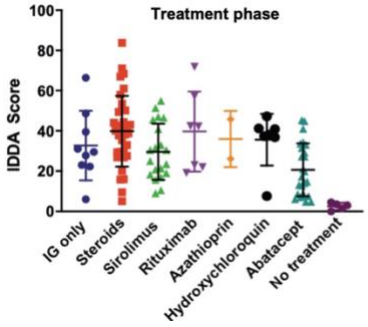
Journal of Clinical Immunology, in press.
https://doi.org/10.1007/s10875-021-01177-2

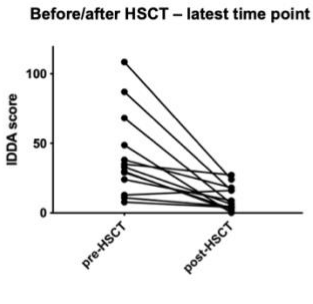
J Allergy Clin Immunol 2020;145:1452-63. (using version 1 of the IDDA score)
https://doi.org/10.1016/j.jaci.2019.12.896

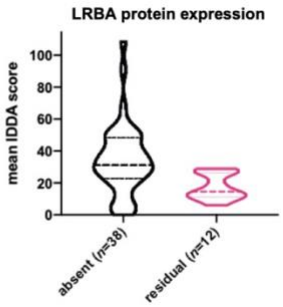


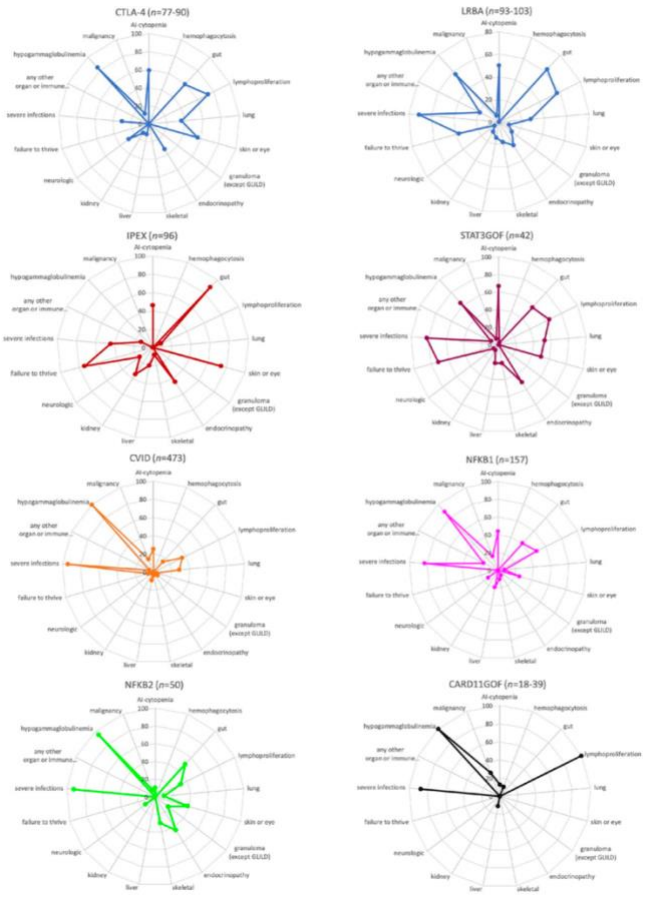


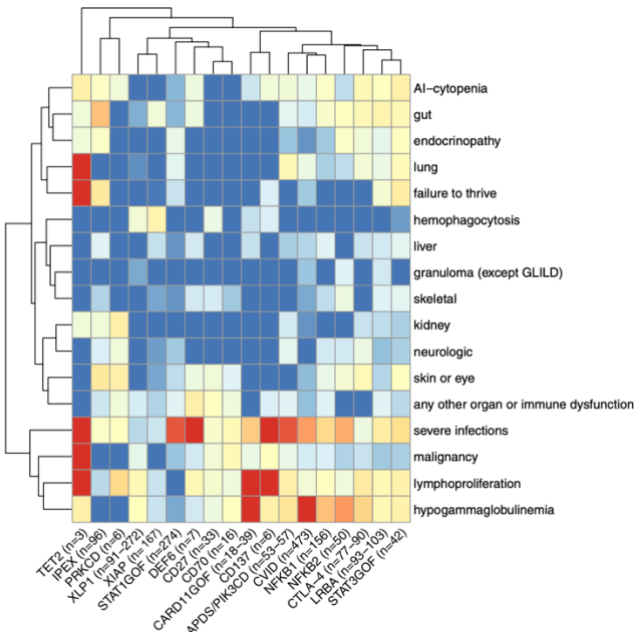












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