A recessive form of hyper-IgE syndrome by disruption of ZNF341-dependent STAT3 transcription and activity.

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Résumé en anglais	Heterozygosity for human () dominant-negative (DN) mutations underlies an autosomal dominant form of hyper-immunoglobulin E syndrome (HIES). We describe patients with an autosomal recessive form of HIES due to loss-of-function mutations of a previously uncharacterized gene, ZNF341 is a transcription factor that resides in the nucleus, where it binds a specific DNA motif present in various genes, including the promoter. The patients' cells have low basal levels of STAT3 mRNA and protein. The autoinduction of STAT3 production, activation, and function by STAT3-activating cytokines is strongly impaired. Like patients with DN mutations, ZNF341-deficient patients lack T helper 17 (T17) cells, have an excess of T2 cells, and have low memory B cells due to the tight dependence of STAT3 activity on ZNF341 in lymphocytes. Their milder extra-hematopoietic manifestations and stronger inflammatory responses reflect the lower ZNF341 dependence of STAT3 activity in other cell types. Human ZNF341 is essential for the transcription-dependent autoinduction and sustained activity of STAT3.

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