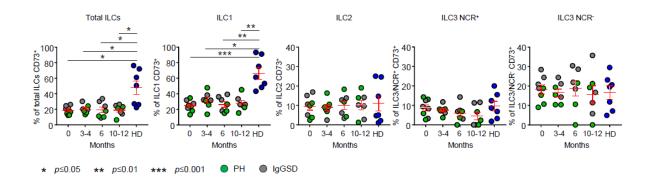
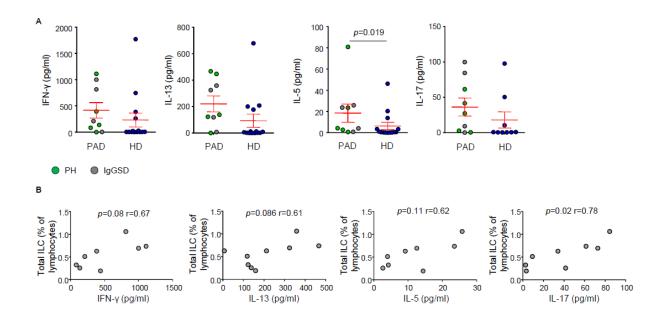
Supplementary Figures.

Supplementary Figure 1. CD73 expression by ILCs in PH/IgGSD patients and HDs. CD73 expression evaluated on total ILCs and ILC subsets in PH/IgGSD patients over 12 months of IVIG treatment and in HDs. Each dot represents one patient or healthy donor and mean±SEM are superposed. Green dots represent PH patients, grey dots represent IgGSD patients and blue dots represent healthy donors.

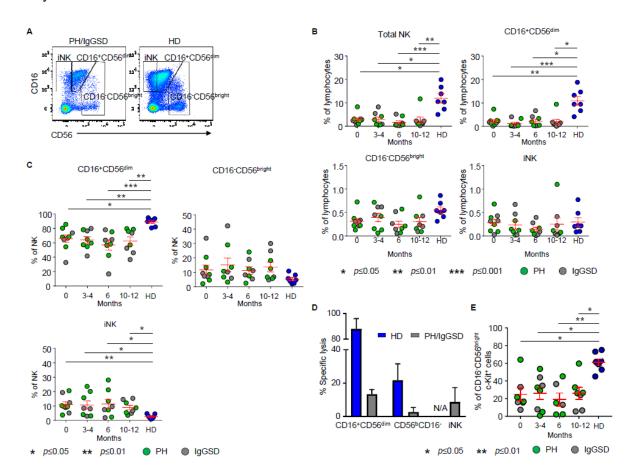


Supplementary Figure 2. Cytokine quantification in PH/IgGSD patients and HDs serum and correlation with ILCs frequency. A. Quantification of IFN-γ, IL-13, IL-5 and IL-17 concentration (pg/ml) measured in the serum of treatment-naive PH/IgGSD patients and in HDs by MSD multiplex assay. Each dot represents one patient or healthy donor and mean+/-SEM are superposed. Green dots represent PH patients, grey dots represent IgGSD patients and blue dots represent healthy donors. B. Correlation between the IFN-γ, IL-13, IL-5 and IL-17 concentration (pg/ml) and the frequency of total ILCs among lymphocytes measured in treatment-naive PH/IgGSD patients.

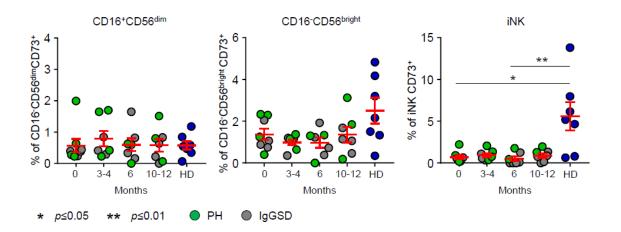


J Investig Allergol Clin Immunol 2017; Vol. 27(6) doi: 10.18176/jiaci.0200

Supplementary Figure 3. Immature and hypofunctional NK cells are increased in PH/IgGSD patients. A. Representative flow cytometry profile and gating strategy used for monitoring total NK and NK subsets in PH/IgGSD patients over 12 months of IVIG treatment and in HDs. **B.** Frequency of total NK and of CD16⁺CD56^{dim}, CD56^bCD16 or iNK cells among total alive lymphocytes in PBMC from patients and in HDs. **C.** Proportion of CD16⁺CD56^{dim}, CD16⁺CD56^{bright} or iNK cells among total NK cells in PBMC from patients and HDs. **D.** Cumulative data representing the cytotoxic capacity of NK subsets in HDs, (blue bars) and in PH/IgGSD patients (grey bars). iNK sorted from healthy donors were not enough in number to perform this assay (N/A: not applicable). **E.** cKit expression by CD16⁺CD56^{bright} NK cells in PH/IgGSD patients. Each dot represents one patient or healthy donor and mean±SEM are superposed. Green dots represent PH patients, grey dots represent IgGSD patients and blue dots represent healthy donors.



Supplementary Figure 4. CD73 expression by NK cells in PH/IgGSD patients and HDs. CD73 expression evaluated on NK cell subsets in PH/IgGSD patients over 12 months of IVIG treatment and in HDs. Each dot represents one patient or healthy donor and mean±SEM are superposed. Green dots represent PH patients, grey dots represent IgGSD patients and blue dots represent healthy donors.



Supplementary Table 1. Clinical description.

PH was diagnosed as a reduction of IgG (at least 2 SD below the mean of age-matched controls), with normal IgA and IgM, onset of the immunodeficiency after four years of age and lack of defined causes of hypogammaglobulinemia. Inclusion criteria for IgGSD were selective or combined IgG subclass deficiency. Patients were given human IVIG (Privigen, CSL-Behring) at monthly doses of 0.4g/kg body weight.

Suppl. Table 1 Clinical description

Patient Number	Sex	Age	Tot IgG 8.54- 12.6	Tot IgA 0.98- 2.03	Tot IgM 0.76- 1.48	IgG1 4.9-11.4	IgG2 1.5-6.4	IgG3 0.2-1.11	IgG4 0.08-1.4	Diagnosis	Resp Infect	ΑI	Atopic/Allergic Asthma	Malignancies	Poor PneumoVac Resp
1	f	22	6.88	0.68	1.31	4.61	3.02	0.35	0.17	Hypogamma	1	1	0	0	1
2	f	53	6.44	1.07	0.81	4.62	1.65	0.15	uld	Hypogamma	1	0	0	0	0
3	f	23	6.51	1.47	1.22	nd	nd	nd	nd	Hypogamma	1	0	0	0	0
4	f	39	6.55	2.89	1.44	nd	nd	nd	nd	Hypogamma	1	0	0	0	1
5	f	44	6.03	0.69	0.23	3.9	2.25	0.37	0.36	Hypogamma	1	0	0	0	0
6	f	34	9.51	1.37	0.64	4.33	3.53	0.3	2.93	Subclass	1	0	0	0	0
7	f	32	8.53	1.29	1.89	3.5	5.52	0.15	0.36	Subclass	1	0	1	0	0
8	f	62	8.86	1.17	0.95	6.25	0.73	0.26	0.25	Subclass	1	0	0	0	1
9	f	47	7.44	1.08	2.48	4.86	2.5	0.25	0.35	Subclass	1	0	1	0	1

nd: not determined

AI: autoimmunity

1: present

0: not present