

## SUPPLEMENTARY MATERIAL

**Supplementary Table 1.** Demographical and clinical data of 20 CVID patients with SNHL compared to 92 CVID patients without SNHL

	CVID patients w/ SNHL (n=20)	CVID patients w/out SNHL (n=92)	Total CVID patients (n=112)	<i>p</i> value
Female; n (%)	13 (65.0%)	52 (56.5%)	65 (58.0%)	0.6190
Current age median (median; IQR)*	48.5 (43.1 – 57.1)	35.8 (25.2 – 47.3)	40.1 (26.1 – 49.2)	<b>0.0002</b>
Age at CVID onset (median; IQR)*	8.4 (3.0 – 19.9)	5.4 (2.3 – 18.0)	6.2 (2.4 – 18.0)	0.4933
Age at CVID diagnosis (median; IQR)*	27.9 (17.0 – 38.9)	19.6 (10.6 – 27.0)	20.5 (11.4 – 32.1)	<b>0.0110</b>
CVID clinical manifestations				
LRTI	14 (70.0%)	47 (51.1%)	61 (54.5%)	0.1437
URTI	13 (65.0%)	52 (56.5%)	65 (58.0%)	0.6190
Sinusitis	9 (45%)	27 (29.3%)	36 (32.1%)	0.1937
Otitis	5 (25%)	22 (23.9%)	27 (24.1%)	>0.999 9
Meningitis	1 (5%)	5 (5.4%)	6 (5.4%)	>0.999 9
Encephalitis	1 (5%)	1 (1.1%)	2 (1.8%)	0.3266
Sepsis	2 (10%)	3 (3.3%)	5 (4.5%)	0.2169
CMV infection	1 (5%)	4 (4.3%)	5 (4.5%)	>0.999 9
EBV infection	0 (0%)	2 (2.2%)	2 (1.8%)	>0.999 9
VZV infection	1 (5%)	4 (4.3%)	5 (4.5%)	>0.999 9
Hematologic AI	5 (25%)	23 (25%)	28 (25%)	>0.999 9
Rheumatologic AI	2 (10%)	8 (8.7%)	10 (8.9%)	>0.999 9

Endocrine AI	2 (10%)	5 (5.4%)	7 (6.3%)	0.6063
Gastroenterological AI	4 (20%)	22 (23.9%)	26 (23.2%)	>0.999 9
Nephrological AI	1 (5%)	0 (0%)	1 (0.9%)	0.1786
Neurological AI	0 (0%)	1 (1.1%)	1 (0.9%)	>0.999 9
Ophthalmological AI	1 (5%)	1 (1.1%)	2 (1.8%)	0.3266
Dermatological AI	0 (0%)	10 (10.9%)	10 (8.9%)	0.2040
Lymphadenopathies	3 (15%)	14 (15.2%)	17 (15.2%)	>0.999 9
Hepatomegaly	2 (10%)	15 (16.3%)	17 (15.2%)	0.7327
Splenomegaly	12 (60%)	47 (51.1%)	59 (52.7%)	0.6222
Asymptomatic	1 (5.0%)	12 (13.1%)	13 (11.6%)	0.4575
Professional/environmental acoustic trauma	0 (0%)	0 (0%)	0 (%)	>0.999 9
Ototoxic drug exposure*	0 (%)	0 (0%)	0 (0%)	>0.999 9

AI, autoimmunity; CMV, cytomegalovirus; CVID, common variable immunodeficiency; EBV, Epstein-Barr virus; IQR, interquartile range; LRTI, lower respiratory tract infection; SNHL, sensorineural hearing loss; URTI, upper respiratory tract infection; VZV, varicella zoster virus

\*Aminoglycosides, platinum-derived chemotherapy