

Supplementary table 1. Patients with IEI baseline clinical characteristics and SARS-CoV-2 infection description.

Patient	Gender	Age (years)	IEI ^a	COVID-19 risk factors	Baseline treatment			COVID-19				Publication
					IgRT	Antibiotic	Immunosuppressant	Severity	Symptoms	Treatment	Outcome	
P1	M	9	MHCII def. (<i>RFXANK</i>)	No	Yes	Cotrimoxazole, posaconazole	Systemic corticosteroids	Moderate	Fever, non-hypoxemic pneumonia	Remdesivir, empiric antibiotic	Recovered	No
P2	M	9	MHCII def. (<i>RFXANK</i>)	No	Yes	Cotrimoxazole, posaconazole	Systemic corticosteroids	Mild	Asymptomatic	Remdesivir	Recovered	No
P3	F	17	CD4 ⁺ idiopathic lymphopenia	No	No	No	No	Mild	Low-grade fever, myalgia, asthenia, smell and taste loss	No	Dysgeusia, dysosmia (for 2 years)	No
P4	F	12	PGM3 def.	No	No	No	Dupilumab	Moderate	Cough, low-grade fever Ground-glass opacity on chest scan	No	Recovered	Deyà-Martínez <i>et al.</i> , 2021; García-García <i>et al.</i> , 2021
P5	M	19	A-T (<i>ATM</i>)	No	Yes	No	No	Mild	Fever No pneumonia (chest X-ray)	Empiric antibiotic	Recovered	Deyà-Martínez <i>et al.</i> , 2021
P6	F	13	Jacobsen sd (11q del.)	No	No	No	No	Mild	Cough	No	Recovered	
P7	M	17	CATCH22 (22q11del.)	No	No	No	No	Mild	Asymptomatic	No	Recovered	
P8	F	16	Down sd. (47,XX+21)	Down syndrome, mitral regurgitation	No	No	No	Mild	Asymptomatic	No	Recovered	
P9	M	10	Jacobsen sd. (11q del.)	No	No	No	No	Mild	Asymptomatic	No	Recovered	
P10	M	5	CATCH22 (22q11 del.)	Cyanotic congenital heart disease	No	No	No	Mild	Fever	No	Recovered	
P11	M	10	CATCH22 (22q11del.)	No	No	No	No	Mild	Asymptomatic	No	Recovered	No

COVID-19 immune responses in pediatric IEI

Patient	Gender	Age (years)	IEI ^a		COVID-19 risk factors	Baseline treatment			COVID-19				Publication
						IgRT	Antibiotic	Immunosuppressant	Severity	Symptoms	Treatment	Outcome	
P12	F	4	Charge	sd.	Mild SAHS	No	No	No	Mild	Asymptomatic		Recovered	No
P13	M	13	Kabuki	sd.	No	No	No	No	Mild	Odynophagia, fever		Recovered	No
P14	M	9	THE	sd.	No	Yes	No	No	Mild	Cough, fever, runny nose, diarrhea	No	Recovered	No
P15	F	14	Sotos	sd	No	No	No	No	Mild	Headache, asthenia	No	Recovered	No
P16	M	8	Bruton	sd.	No	Yes	No	No	Mild	Asymptomatic	No	Recovered	Deyà-Martínez <i>et al.</i> , 2021
P17	F	17	CVID		No	No	No	No	Mild	Asymptomatic	No	Recovered	
P18	M	16	CVID		No	No	No	No	Mild	Low-grade fever, odynophagia	No	Recovered	No
P19	F	14	CVID		No	No	No	No	Mild	Asymptomatic	No	Recovered	
P20	M	18	ALPS		No	No	No	No	Mild	Odynophagia, retroocular pain, runny nose, loss of smell and taste	No	Recovered	Deyà-Martínez <i>et al.</i> , 2021
P21	M	15	STAT1 GOF		No	No	Fluconazole	No	Mild	Asymptomatic	No	Guillain-Barré ^b	
P22	F	7	STAT1 GOF		No	Yes	Fluconazole, acyclovir	Ruxolitinib	Mild	Asymptomatic	No	Recovered	No
P23	M	23	CGD		No	No	Cotrimoxazole, itraconazole	Corticosteroids	Mild	Asymptomatic	No	Recovered	
P24	M	19	MyD88 def.		Obesity	No	No	No	Severe	Fever, cough Hypoxemic pneumonia (no MV)	Corticoids Remdesivir Antibiotic Heparin	Recovered	Von Bernuth <i>et al.</i> , 2018 Deyà-Martínez <i>et al.</i> , 2021
P25	F	17	MyD88 def.		No	No	No	No	Moderate	Fever, cough Non-hypoxemic pneumonia	Corticoids Remdesivir Antibiotic	Recovered	

^aThe genetic defect is described if known; otherwise, we only provide the immunodeficiency according to the ESID clinical criteria classification (2019)(46) ^bGuillain-Barré after 9 months of testing positive for SARS-CoV-2, with no other infectious trigger detected in the routine screening. ALPS: autoimmune lymphoproliferative syndrome; A-T: ataxia-telangiectasia; CGD: chronic granulomatous disease; def.: deficiency; GOF: gain-of-function; IEI: Inborn errors of immunity; MHCII: major histocompatibility complex II; MMF: mycophenolate mofetil; MV: mechanical ventilation; SAHS: sleep apnea-hypopnea syndrome; sd.: syndrome; THE: trichohepatoenteric; 22q11del: 22q11 deletion.