

**Table 1. The clinical spectrum of patients with PKC $\delta$  deficiency**

Kindred	Patient	Mutation (predicted protein variant)	Origin	Sex	Follow-up	Age at onset of symptoms	Autoimmune manifestations	BCG status Clinical infectious phenotype (infections, microbiology, pathology results) Treatment, Prophylaxis	Reference
A	P1	c.1840C>T/ c.1840C>T (p.R614W/p.R614W)	Mexico, living in USA	M	Alive	3 years	Autoimmune hemolytic anemia SLE ANA(+) Anti-RNP(+) Anti-smith(+) Anti-SSA(+)	BCG-vaccine: no AE Recurrent pneumonia, EBV infection, recurrent otitis media Corticosteroids, rapamycin Prophylaxis: NR	(Kuehn et al., 2013)
B	P2	c.742G>A/ c.742G>A (p.G248S/p.G248S)	Turkey	M	Alive	1 year	SLE ANA(+)	BCG-vaccine: no AE Recurrent pneumonia, CMV infection, recurrent gastroenteritis Hydroxychloroquine Prophylaxis: IVIG, antibiotics.	(Kiykim et al., 2015)
C	P3	c.1528G>A/ c.1528G>A (p.G510S/p. G510S)	France	F	Alive	10 years	SLE ANA(+) Anti-dsDNA(+)	BCG-vaccine: no AE Severe <i>Legionella</i> pneumonia, recurrent pneumonia, severe papillomatosis Corticosteroids, hydroxychloroquine Prophylaxis: NR	(Belot et al., 2013)
	P4	c.1528G>A/ c.1528G>A (p.G510S/p. G510S)		F	Dead	3 years	Autoimmune hemolytic anemia SLE ANA(+) Anti-dsDNA(+)	BCG-vaccine: no AE Died at 13 years, from septic shock due to <i>Pseudomonas aeruginosa</i> Corticosteroids, MMF, anti-CD20 (rituximab) Prophylaxis: NR	
	P5	c.1528G>A/ c.1528G>A (p.G510S/p. G510S)		M	Alive	6 years	SLE ANA(+) Anti-dsDNA(+)	BCG-vaccine: no AE Pleuritis and pneumonia Corticosteroids, MMF Prophylaxis: IVIG	
D	P6	c.1352+1G>A/c.1352+1G>A (?/?)	Turkey	M	Alive	1 year	SLE ANA(+) Anti-dsDNA(+) Anti-cardiolipin(+)	BCG-vaccine: no AE Recurrent pneumonia, urinary tract infections, gastroenteritis, otitis media Corticosteroids, anti-CD20 (rituximab) Prophylaxis: IVIG, antibiotics	(Salzer et al., 2013)
E	P7	c.1294G>T/ c.1294G>T (p.G432W/p.G432W)	Pakistan, living in UK	F	Alive	1 year	Autoimmune hemolytic anemia SLE ANA(+) Anti-dsDNA(+)	BCG-vaccine: no AE Corticosteroids, anti-CD20 (rituximab), azathioprine Prophylaxis: IVIG	(Lei et al., 2018; Nanthapisa l et al., 2017)
	P8	c.1294G>T/ c.1294G>T (p.G432W/p.G432W)		F	Alive	1 year	Autoimmune hemolytic anemia SLE ANA(+) Anti-dsDNA(+)	BCG-vaccine: no AE Cervical and inguinal lymphadenopathy Corticosteroids, anti-CD20 (rituximab), azathioprine	

	P9	c.1294G>T/ c.1294G>T (p.G432W/p.G432W)		M	Alive	2 years	SLE ANA(+) Anti-dsDNA(+)	Prophylaxis: IVIG  BCG-vaccine: no AE Corticosteroids, anti-CD20 (ofatumumab) Prophylaxis: IVIG	
F	P10	c.788-2A>G /c.571C>T ( ?/p.Q191*)	UK	M	Alive	9 months	SLE ANA(+)	BCG-vaccine: no AE <i>Listeria meningitis</i> , recurrent otitis media, early-onset enterocolitis, systemic <i>Candida</i> infection, sepsis due to <i>Enterococcus faecium</i> , recurrent <i>Achromobacter xyloxidans</i> cervical lymphadenitis, SARS-CoV-2 infection Sirolimus, anti-CD20 (rituximab) Prophylaxis: IVIG	submitted manuscript Roderick <i>et al.</i>  (Meyts et al., 2020)
	P11	c.788-2A>G /c.571C>T ( ?/p.Q191*)		F	Alive	6 months	SLE ANA(+)	BCG-vaccine: no AE <i>Haemophilus influenzae</i> sepsis, recurrent gastroenteritis, early-onset enterocolitis MMF, anti-CD20 (rituximab) Prophylaxis: IVIG	
G	P12	c.1293_1294insA/ c.1293_1294insA (p.G432Rfs*15/ p.G432Rfs*15)	Afghanist an, living in Iran	M	Alive	1 year	SLE ANA(+) Anti-dsDNA (+)	BCG-vaccine: no AE <i>Streptococcus pneumoniae</i> pneumonia, <i>Haemophilus influenzae</i> otitis media, recurrent oral candidiasis, recurrent pneumonia, recurrent gastroenteritis Corticosteroids, NSAID, Immuran Prophylaxis: cyclic rotation of sulfamethoxazole-trimethoprim/amoxicillin and azithromycin	(Sharifinej ad et al., 2020)
H	P13	c.571+2dup/c.571+2dup (?/?)	Turkey	F	Alive	1 year	SLE ANA(+) Anti-dsDNA(+)	BCG-vaccine: at 2 months of age, BCG-osis <i>Salmonella</i> bacteremia <i>S. aureus</i> inguinal lymphadenitis, recurrent gingivitis Antimycobacterial treatment, ceftriaxone Corticosteroids Prophylaxis: amoxicillin, IVIG	This paper
I	P14	c.1384C>T/ c.1384C>T (p.Q462*/p.Q462*)	Turkey	M	Alive	7 months	Autoimmune hemolytic anemia ANA(+)	BCG-vaccine: BCG-itis Gastroenteritis Isoniazid, rifampicin Corticosteroids Prophylaxis: none at present	This paper
	P15	c.1384C>T/ c.1384C>T (p.Q462*/p.Q462*)		M	Alive	7 months	Autoimmune hemolytic anemia ANA(+)	BCG vaccine: BCG-itis Gastroenteritis, shingles Isoniazid, rifampicin Corticosteroids Prophylaxis: IVIG	

J	P16	c.642del/c.642del (p.N214Kfs*26/ p.N214Kfs*26)	Iran	F	Alive	1 year	No SLE, ANA (-) Anti-dsDNA (-)	BCG vaccine: BCG-itis Isoniazid, rifampicin, azithromycin Prophylaxis: none at present	This paper
	P17	c.642del/c.642del (p.N214Kfs*26/ p.N214Kfs*26)		M	Alive	-	No SLE, ANA (-) Anti-dsDNA (-)	Asymptomatic Prophylaxis: none at present	

Family code, patient number, and mutations are as in **Fig. 1**. Predicted protein variations are indicated in parenthesis. “?” indicates unknown prediction of splice site variants. Patient origin, sex, current vital status, age at onset of symptoms, and clinical phenotypes are shown. AE: adverse effect, ANA: antinuclear antibodies, BCG: *Bacillus Calmette-Guérin*, CMV: cytomegalovirus, EBV: Epstein-Barr virus, dsDNA: double-stranded (ds, native) DNA, F: female, IVIG: intravenous immunoglobulin, M: male, MMF: mycophenolate mofetil, NR: not reported, NSAID: non-steroidal anti-inflammatory drug, SARS-COV2: severe acute respiratory syndrome coronavirus 2, SLE: systemic lupus erythematosus, UK: United Kingdom