

Table V: Subjects with severe Protein S deficiency: Genotype, biological, and clinical features

Pedigree	Mutation	Total PS	Free PS	PS activity	Clinical data (Age of the first thrombotic episode)	Ref
HOMOZYGOUS SUBJECTS						
PS-1-unnamed	43, AA (del A)	8	1.2		(birth) PF, DIC, SK	133
PS-221-276	340, G D	37	7	8	DVT (20)	130
PS-34-29	460, S P	50	34	40	DVT (38), PE	119
PS-34-214	520, R G	36	16	14	DVT, PE (12)	118, 120
PS-33-107	624, S L	50	33	19		156
COMPOUND HETEROZYGOUS SUBJECTS						
PS-33-108	intron e, G A, exon 5 +5 460, S P	70	50	30	DVT, PE, MI (23)	137
PS-81-07	46, V L	20	ND	ND	DVT, PE, SN (10)	97
PS-81-08	intron n, G T, exon 15 -1					
PS-66-1	146-147 ins A 410 R Stop		8		(10 days) Purpura, SK 7 year-old	138 139

PF, Purpura fulminans. DVT, Deep vein thrombosis. MI, Myocardial infarction. PE, Pulmonary thromboembolism. SK, Skin necrosis. MVT, Mesenteric vein thrombosis.