Supplemental Data

Supplement Table 1. Overview of all variants found in our cohort and their association with desmopressin response

	Proportion of patients with complete response to desmopressin				
	0%	0-40%	40-60%	60-100%	100%
Type 1	1482M ² (0/1)		D141N ¹ (1/2)	Del exon 4-5 (5/6)	G19R (2/2)
VWD	P812R fs*31 (0/2)		A1716P (1/2)	R854Q (2/3)	R236C (1/1)
	C1149R (0/1)			C1130C (4/6)	C440A *fs17 (1/1)
	R1205H ⁸ (0/8)			Y1584C ¹⁰ (7/8)	C652F (1/1)
	R1374H (0/1)				2442+4A>G (1/1)
	R1379C (0/1)				R924Q ⁵ (4/4)
	S2179F (0/1)				C1060Y (2/2)
					T1156M (2/2)
					P1162L (1/1)
					C1190R (1/1)
					P1266L ⁹ (1/1)
					F1369I (1/1)
					Dup exon 28 (1/1)
					K1794E (2/2)
					Del exon 33-34 (3/3)
					V1934G (1/1)
					T1951A (3/3)
					P2063S (1/1)
					C2304Y (3/3)
					C2360* (1/1)
					A2498D (1/1)
					N2546Y (2/2)
Type 2 VWD	C1130C (0/1)	F1293L (1/5)	C1060Y ⁷ (2/4)	R854Q4 (3/4)	W791M ³ (1/1)
	C1149R (0/2)	R1597W (2/6)		R924Q ⁶ (3/4)	C1190Y (2/2)
	L1282P (0/1)				V1499E (2/2)
	S1285P (0/1)				G1573S (1/1)
	L1288R (0/1)				, , ,
	L1307P (0/1)				
	R1374C (0/3)				
	R1374H (0/8)				
	R1374L (0/1)				
	K1408del (0/1)				
	N1421K (0/1)				
	S1506L (0/2)				
	Y1542C (0/1)				
	I1628W (0/1)				
	G1631D (0/1)				

Of note, some variants, which are known in literature to cause both type 1 and type 2 VWD, are in this table listed as both type 1 and type 2 VWD, depending on the phenotype patents. Between () indicate number of patients responded/total number of patients. ¹both patients also had R2313C. ²also had R960W and 3108+2T>G. ³Also had P812R fs*31. ⁴The patient who did not respond also had 6257-1G>A, whereas two patients who responded also had R2535*. ⁵One patient also had C1169W. ⁶One patient who responded also had G1609R, whereas another patient who respond had R1374H. ⁵In the two patients who did not respond, one had a homozygous form and the other one also had R854Q. One patient who responded also had V1760I. ⁵Two patients also had 5170+10C>T. ⁶Also had V1279I. ¹OThe patient who did not respond had a homozygous form.

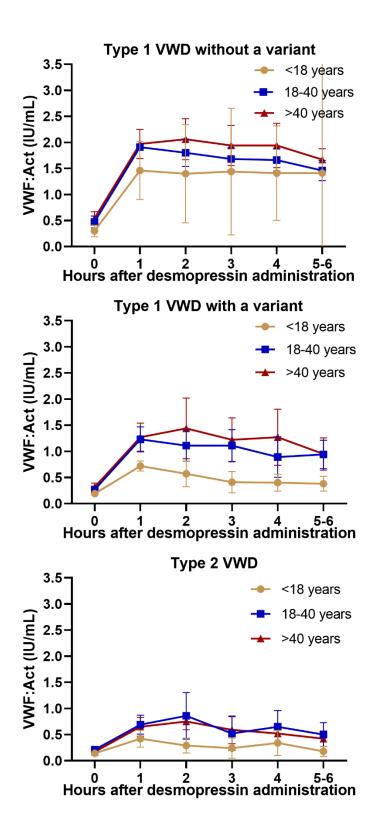


Figure legend: Data are represented as mean and 95% CI.

Supplemental Figure 2. In type 1 VWD without a variant, patients with blood group O seem to have a less sustained desmopressin response, although not statistically significant

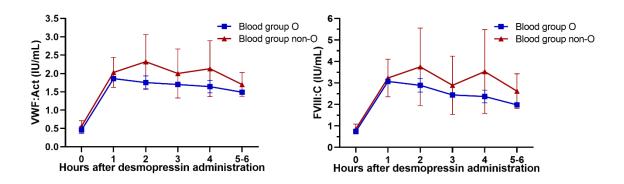


Figure legend: Data are represented as mean and 95% CI. Due to the large 95% confidence interval, none of the differences between blood group O and non-O at 1 to 5-6 hours after desmopressin were statistically significant.