

**E-TABLE 51-1 CLASSIFICATION OF VON WILLEBRAND DISEASE**

FACTOR	TYPE 1	TYPE 2A	TYPE 2B	TYPE 2M	TYPE 2N	TYPE 3	PSEUDO-vWD	BSS
Inheritance	AD	AD, AR	AD, AR	AD	AR	AR, AD	AD	AR
Platelet count	NL	NL	NL, ↓	NL	NL	NL	↓, NL	↓, NL
Bleeding time	NL, ↑	↑	↑	↑	NL, ↑	↑↑	↑	↑
PTT	NL, ↑	↑, NL	↑, NL	↑	↑↑	↑↑	↑, NL	NL
VIII	NL, ↓	NL, ↓	↓, NL	NL, ↓	↓↓	↓↓	↓, NL	NL
vWF:Ag	NL, ↓	NL, ↓	↓, NL	NL	NL	Absent	↓, NL	NL
vWF:Rcof	NL, ↓	↓↓	↓, NL	↓↓	NL	Absent	↓, NL	NL
Multimers	NL, ↓	↓ H/I	↓↓ H	NL	NL	Absent	↓↓ H	NL
RIPA	NL, ↓	↓↓	↑*	↓	NL	↓↓	↑*	↓↓

AD, Autosomal dominant; AR, autosomal recessive; BSS, Bernard-Soulier syndrome; H, high-molecular-weight multimers; I, intermediate-molecular-weight multimers; NL, normal; PTT, partial thromboplastin time; RIPA, ristocetin-induced platelet agglutination; vWD, von Willebrand disease; vWF:Ag, von Willebrand factor antigen level; vWF:Rcof, von Willebrand factor:ristocetin cofactor activity; ↑, increased; ↓, decreased; ↑\*, increased agglutination in response to low-dose ristocetin.