Supporting Information

Xiang et al. 10.1073/pnas.1018559108

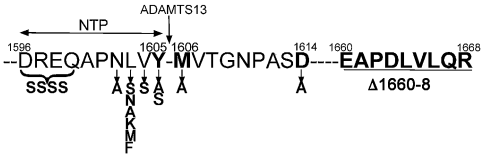

Fig. S1. Partial primary sequence of the ADAMTS13 substrate VWF115. VWF amino acids are shown as single letter, the ADAMTS13 cleavage site is indicated, substitutions/deletions referred to in the text are annotated below the sequence, and the N-terminal peptide (NTP) is represented by the horizontal arrow. VWF115 spans residues 1554Glu–1668Arg, but residues shown are only for VWF73, spanning1596Asp–1668Arg.

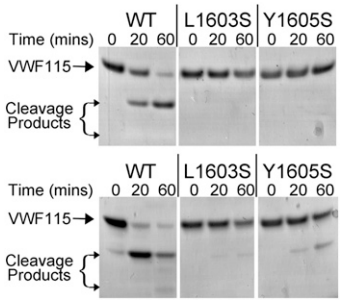

Fig. S2. Cleavage of WT VWF115, VWF115 Leu1603Ser, and VWF115 Tyr1605Ser by ADAMTS13. Cleavage of 6 μM WT VWF115 and variants was performed with 1 nM (*Upper*) and 10 nM (*Lower*) WT ADAMTS13 and reactions visualized by SDS/PAGE.

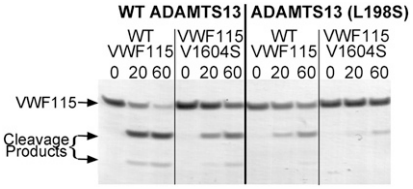

Fig. S3. Studies with VWF115 Val1604Ser, the P2 residue, and ADAMTS13 Leu198Ser. Cleavage of 6 μM WT VWF115 and VWF115 Val1604Ser by 1 nM WT ADAMTS13 and ADAMTS13 Leu198Ser visualized by SDS/PAGE.

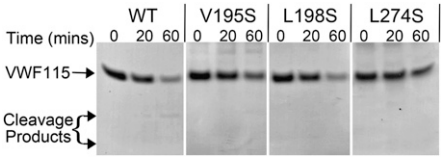

Fig. S4. Cleavage of VWF115 Leu1603Ser by ADAMTS13 Val195Ser, Leu198Ser, and Leu274Asn. Cleavage of 6 μM VWF115 Leu1603Ser by 30 nM WT ADAMTS13 and ADAMTS13 Val195Ser, Leu198Ser, and Leu274Ser are visualized by SDS/PAGE.

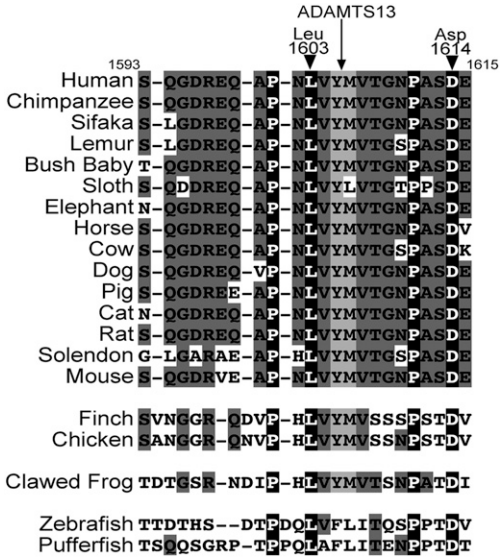

Fig. S5. Regional alignment of the VWF sequence in different species. The amino acid residues within VWF1593–1615 were aligned to identify highly conserved residues. The position of the scissile bond, Leu1603, and Asp1614 are indicated at the top of the figure.

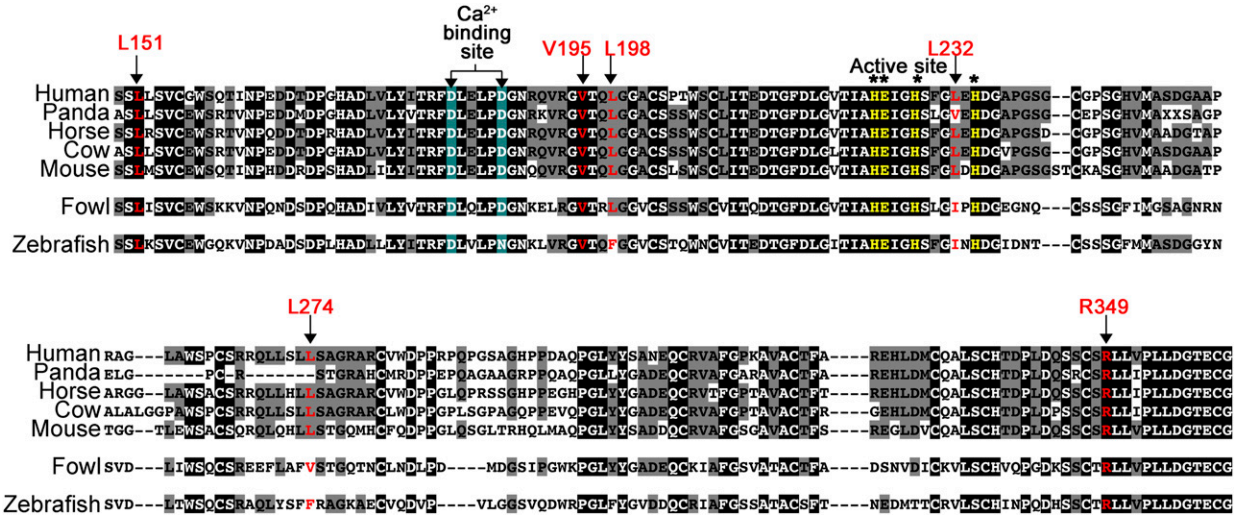

Fig. S6. Regional alignment surrounding the active site of the ADAMTS13 sequence in different species. The active site, conserved Ca^{2+} binding site and residue Arg349 are indicated, along with hydrophobic residues implicated in ADAMTS13 function, Leu151, Val195, Leu198, Leu232, and Leu274.