

Figure 1A

Wildtype ADAMTS13 – Top (SEQ ID NO: 1)

Q⁹⁷R Vatriant of ADAMTS13 – Bottom (SEQ ID NO: 2)

1	mhqrhprarcpplcvgilacgfligcwgpsfhqqsclqalepqavssyl	50
1	mhqrhprarcpplcvgilacgfligcwgpsfhqqsclqalepqavssyl	50
51	spgaplkgrppspgfgqrqrqrnaaggilhllellvavgpdvfqahqedt	100
51	spgaplkgrppspgfgqrqrqrnaaggilhllellvavgpdvfqahredt	100
101	eryvltlnigaeallrdpslgagfrvhlvkmviltpegapnitanltss	150
101	eryvltlnigaeallrdpslgagfrvhlvkmviltpegapnitanltss	150
151	llsvcgwsqtinpedtdpghadlvlyitrfdlelpgdnrqvrgvtqlgg	200
151	llsvcgwsqtinpedtdpghadlvlyitrfdlelpgdnrqvrgvtqlgg	200
201	acsptwscilitdgtgfdlgvtiaheighsfglehdgapgsgcgpsghvma	250
201	acsptwscilitdgtgfdlgvtiaheighsfglehdgapgsgcgpsghvma	250
251	sdgaapraglawspcsrrqlisllsagrarcvwdpprpqpgsaghppdaq	300
251	sdgaapraglawspcsrrqlisllsagrarcvwdpprpqpgsaghppdaq	300
301	pglyysaneqcrvafgpkavactfarehldmcqalschtdpldqsscsrl	350
301	pglyysaneqcrvafgpkavactfarehldmcqalschtdpldqsscsrl	350
351	lvplldgtecgvekwcskgncrslveltpisavhgrwsswgprspcsrsc	400
351	lvplldgtecgvekwcskgncrslveltpisavhgrwsswgprspcsrsc	400
401	gggvvtrrrrqcnnprpafgggracvgadlqaewcntqacektqlfmsqqc	450
401	gggvvtrrrrqcnnprpafgggracvgadlqaewcntqacektqlfmsqqc	450
451	artdgqplrsspggasfyhwgaavphsqqgdalcrhmcraigesfimkrqd	500
451	artdgqplrsspggasfyhwgaavphsqqgdalcrhmcraigesfimkrqd	500

Figure 1B

501	sflldgtrcmpsgpredgtlslcwsgscrtfgcdgrmdsqqvwdrcqvcgg	550
501	sflldgtrcmpsgpredgtlslcwsgscrtfgcdgrmdsqqvwdrcqvcgg	550
551	dnstcsprkgsftagrareyvtflvtvpnltsvyianhrplfthlavrig	600
551	dnstcsprkgsftagrareyvtflvtvpnltsvyianhrplfthlavrig	600
601	gryvvagkmsispnttypsilledgrveyrvaltedrlprleeiriwgplq	650
601	gryvvagkmsispnttypsilledgrveyrvaltedrlprleeiriwgplq	650
651	edadiqvrryrygeeygnltrpdtitftyfqpkprqawwaaavgpcsvscg	700
651	edadiqvrryrygeeygnltrpdtitftyfqpkprqawwaaavgpcsvscg	700
701	aglrwnynscldqarkelvetvqcqgsgqppawpeacvlepccpywavgd	750
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751	fgpcsasccggglrerpvrvcveaagsgliktlpparcragaaqqpavaletcn	800
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901	agscsvscgrglmelrflcmddsalrvpvqeelcglaskpgsrrevcqavp	950
901	agscsvscgrglmelrflcmddsalrvpvqeelcglaskpgsrrevcqavp	950
951	cparwqyklaacsvsccgrgvvrrilvcarahgeddgeeilldtccqglpr	1000
951	cparwqyklaacsvsccgrgvvrrilvcarahgeddgeeilldtccqglpr	1000

Figure 2A

Wildtype human ADAMTS13 – Top (SEQ ID NO: 1)

Wildtype gorilla ADAMTS13 – Bottom (SEQ ID NO: 3)

1	mhqrrhprarcpplcvagilacgflgwcgshfqqsclqalepqavssyl	50
1	mhqrrhprarcpplcvagilacgflgwcgshfqqsclqalepqavssyl	50
51	spgaplkgrppspgfv---qrqrqrqrraaggilhlellvavgpdvfqah	96
51	spgaplkgrppspgfvqrqrqrqrraaggilhlellvavgpdvfqah	100
97	qedterylvltlnlignaeilrdpslgaqfrvhlvkmviltepegapnitan	146
101	qedterylvltlnlignaeilrdpslgaqfrvhlvkmviltepegapnitan	150
147	ltssllsvcgwsqtinpeddtdpghadlvlyitrfdlelpgdnrqvrgvt	196
151	ltssllsvcgwsrtinpeddtdpghadlvlyitrfdlelpgdnrqvrgvt	200
197	qlggacspwscclitedtgfdlgvtiaheighsfglehdgapgsgcgpsg	246
201	qlggacspwscclitedtgfdlgvtiaheighsfglehdgapgsgcgpsg	250
247	hvmadgaapraglawspcsrrqlslsagrarcvwdpprpqpgsaghp	296
251	hvmadgaapraglawspcsrrrllpcrsagrarcvwdpprpqpgsaghp	300
297	pdaqpglyysaneqcrvafgpkavactfarehldmcqalschtdpldqss	346
301	pdaqpglyysaneqcrvafgpkavactfarehldmcqalschtdpldqss	350
347	csrllvplldgtecgvekwcskgrcrslveltpiaavhgrwsswgprspc	396
351	csrllvplldgtecgvekwcskgrcrslveltpiaavhgrwsswgphspc	400
397	srscgggvvtrrrqcnnprpafggracvgadlqaemcntqacektqlfem	446
401	srscgggvvtrrrqcnnprpafggracvgadlqaemcntqacektqlfem	450
447	sqqcartdgqplrsspggasfyhwgaavphsqqgdalcrhmcraigesfim	496
451	seqcartdgqplrsspggasfyhwgaavphsqqgdalcrhmcraigesfim	500

Figure 2B

497	krqdsfldgtrcmpsgpredgtlsicvsgscrtfgcdgrmdsqvwdrcq	546
501	krqdsfldgtrcmpsgpredgtlsicvsgscrtfgcdgrmdsqvwdrcq	550
547	vccgdnstcsprkgsftagrareyvtflvtvtpnltsvyianhrplfthla	596
551	vccgdnstcsprkgsftagrareyvtflvtvtpnltsvyianhrplfthla	600
597	vriggryvvagkmsispnttyslledgrveyrvaltedrlprleeiriw	646
601	vriggryvvagkmsispnttyslledgrveyrvaltedrlprleeiriw	650
647	gplqedadiqvrrygeeygnltrpditftyfqpkrqawvwaavrgpcs	696
651	gplqedadiqvrrygeeygnltrpditftyfqpkrqawvwaavrgpcs	700
697	vscgaglrwnyscldqarkelvetvqcqgsqppawpeacvlepccpyw	746
701	vscgaglrwnyscldqarkelvetvqcqgsqppawpeacvlepccpyw	750
747	avgdfgpcasascggglrerpvrvcveaagsllktlpparcragaaqpaval	796
751	avgdfgpcasascggglrerpvrvcveaagsllktlpparcragaaqpavvl	800
797	etcnpqpcparwevsepssctsaggaglalenetcvpgadgleapvtegp	846
801	etcnpqpcparwevsepssctsaggaglalenetcvpgadgleapvtegp	850
847	gsvdeklpapepcvgmscppgwhldatsagekapspwgsirtgaqaahv	896
851	gsvdeklpapepcvgmscppgwhldatsagekapspwgsirtgaqaahv	900
897	wtpaagscsvscgrglmelrflcmdsalrvpvqeelcglaskpgsrrevc	946
901	wtpaagscsvscgrglmelrflcmdsalrvpvqeelcglaskpgsrqevc	950
947	qavpcparwqyklaacsvscgrgvrrilycarahgeddgeeilldtqcq	996
951	qavpcparwryklaacsvscgggvrrilycarahgeddgeeilldtqcq	1000

Figure 2C

997	glprpepqeacslepcpprwkvmslgpcsascglgtarrsvacvqldqgq	1046
1001	glprpepqeacslepcpprwkvmslgpcsascglgtarrsvacvqldqgq	1050
1047	dvevdeaacaalvrpeasvpcliadctyrwhvgtwmecsvscgdgiqrrr	1096
	. :	
1051	dvevdeaacvalvrpqasvpcliadctyrwhvgtwmecsvscgdgiqrrr	1100
1097	dtclgpqaqapvpadfcqhlpkpvtrgchwagpcvgqgtpslvpheaaaa	1146
	.	
1101	dtclgpqaqapvpadfcqhlpkpvtrgchwagpcvgqgmplvpheaaaa	1150
1147	pgrttatpagaslewsqangllfspapqprllpgpqensvqssacgrqh	1196
	. : .	
1151	pgrttatpagaslewsqarallfspapqqrllpgpqensaqsacgrqh	1200
1197	leptgtidmrpggqadcavaigrplgevvtlrvlesslncsagdmlllw	1246
1201	leptgtidmrpggqadcavaigrplgevvtlrvlesslncsagdmlllw	1250
1247	rltwrkmcrklldmfssktntlvvrqrcgrpgggvllrygsqlapetfy	1296
1251	rltwrkmcrklldmfssktntlvvrqrcgrpgggvllrygsqlapetfy	1300
1297	recdmqlfgpwgeivspslspatsnaggcrlfinvaphariaihalatnm	1346
1301	recdmqlfgpwgeivspslspatsnaggcrlfinvaphariaihalatnm	1350
1347	gagteganasyilirdthslrttafhgqqvlywesessqaemefsegflk	1396
	.	
1351	gagteganasyisirdthslrttafhgqqvlywesessqaemefsegflk	1400
1397	aqaslrqywtlqswvpemqdpqswkgkegt	1427
	:	
1401	aqaslrqywtlqswvpevqdpqswkgkegt	1431

Figure 3

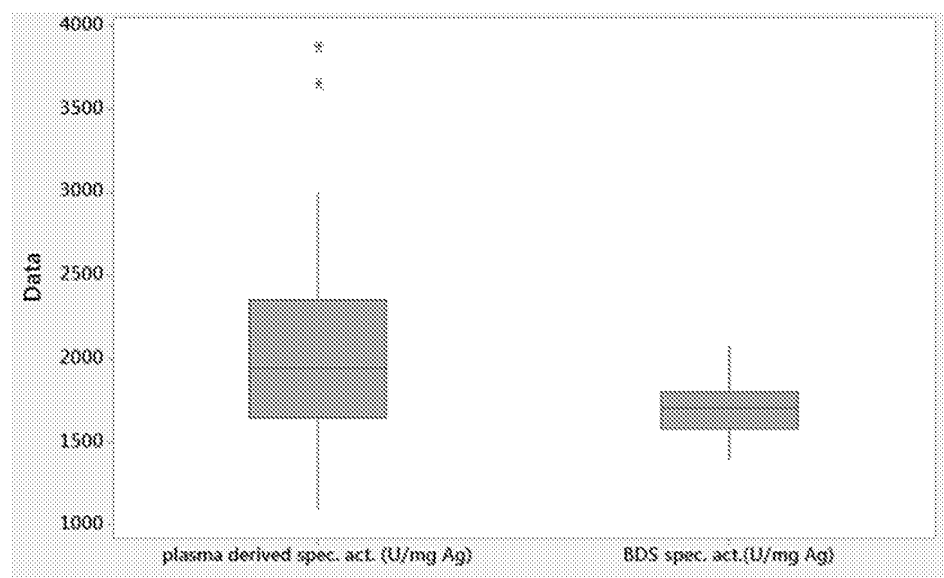


Figure 4

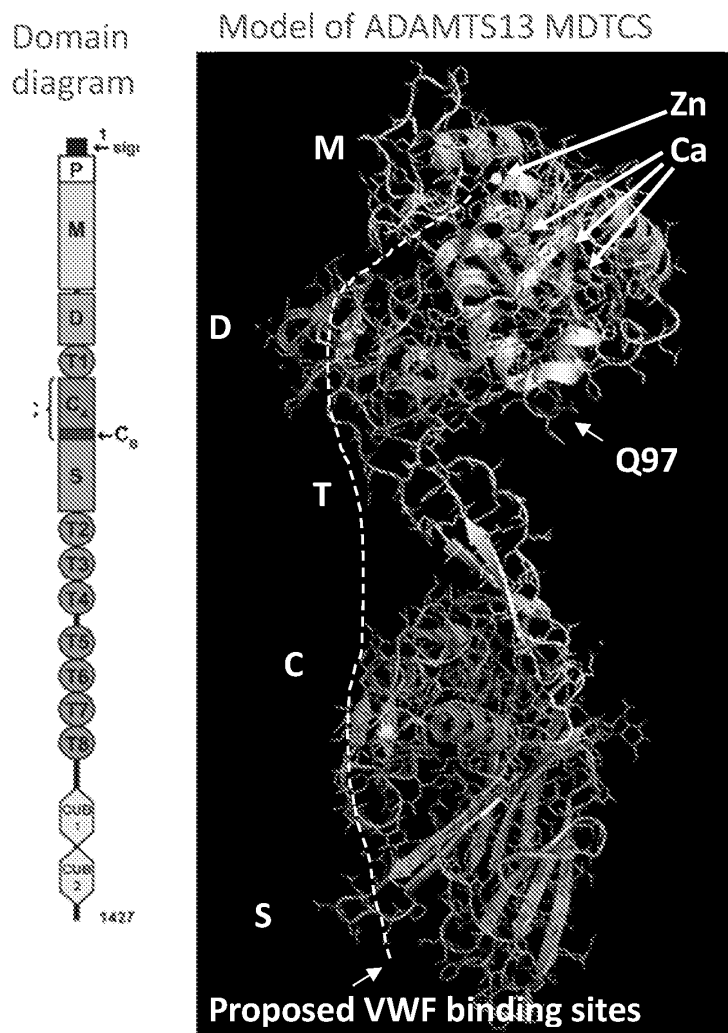


Figure 5

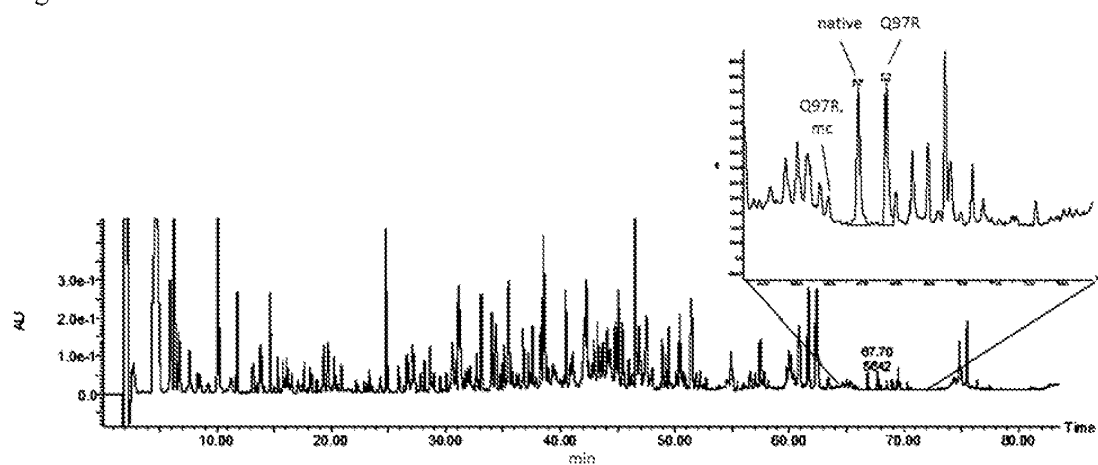


Figure 6

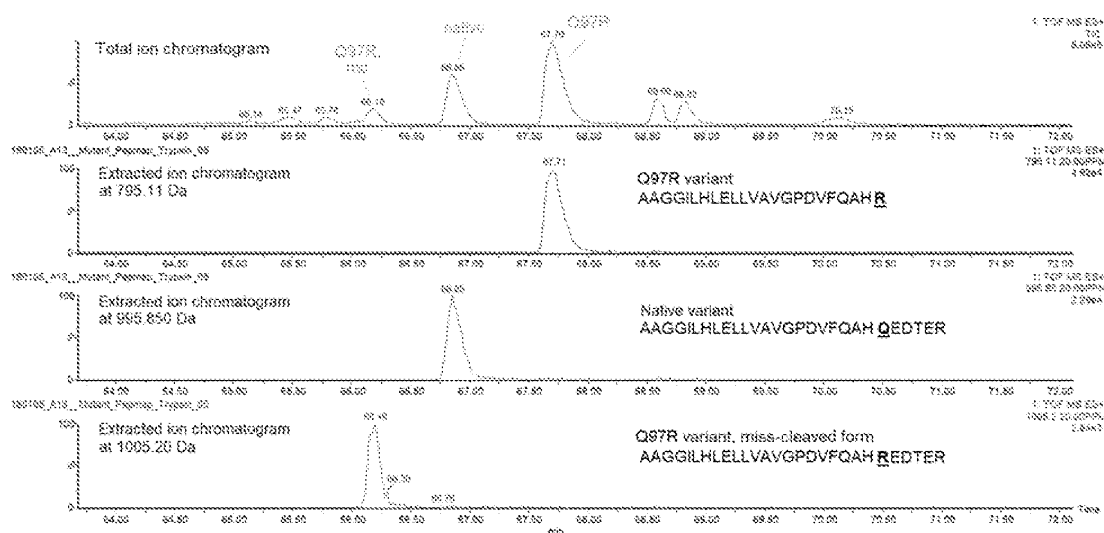


Figure 7

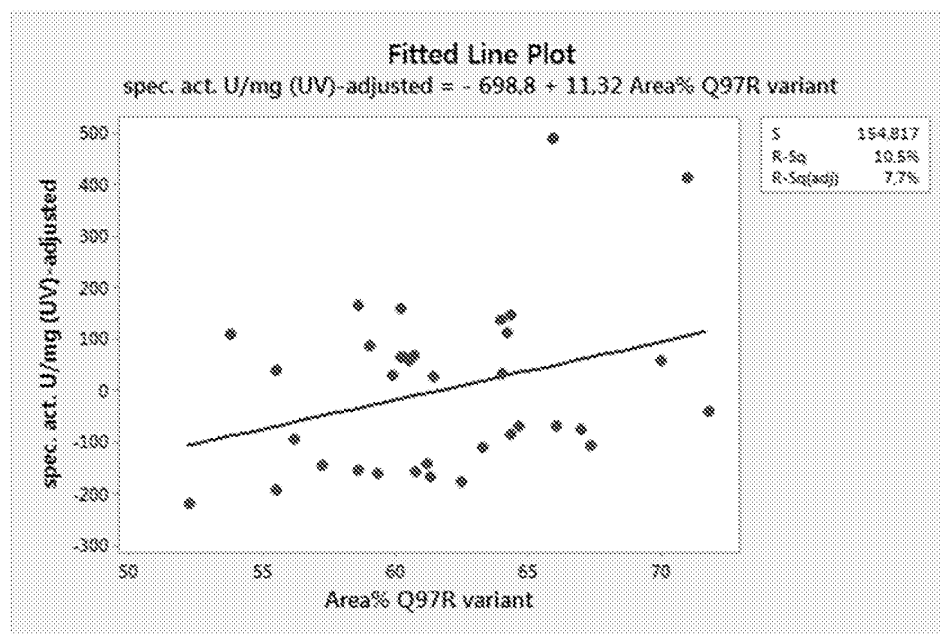


Figure 8

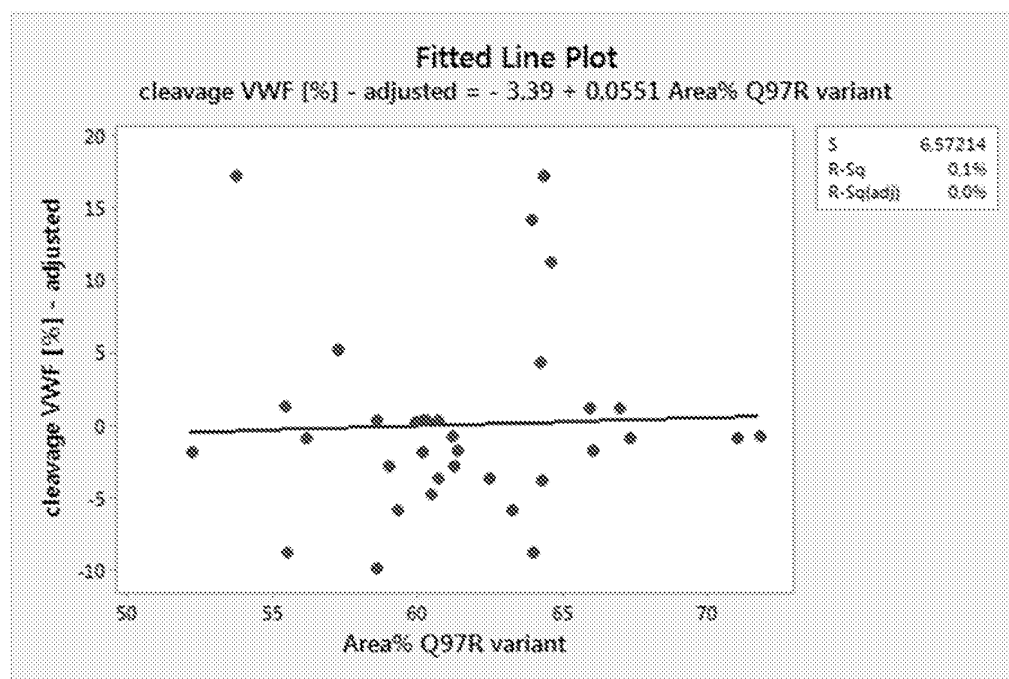


Figure 9

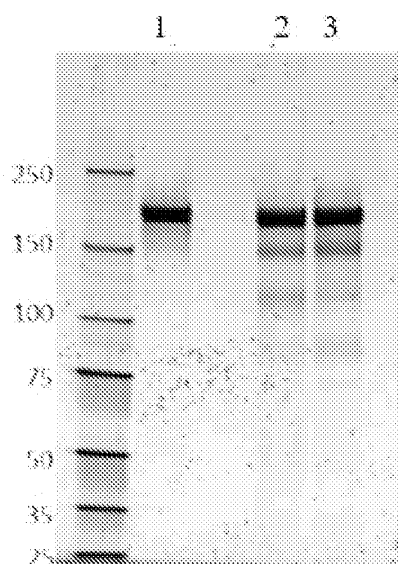


Figure 10

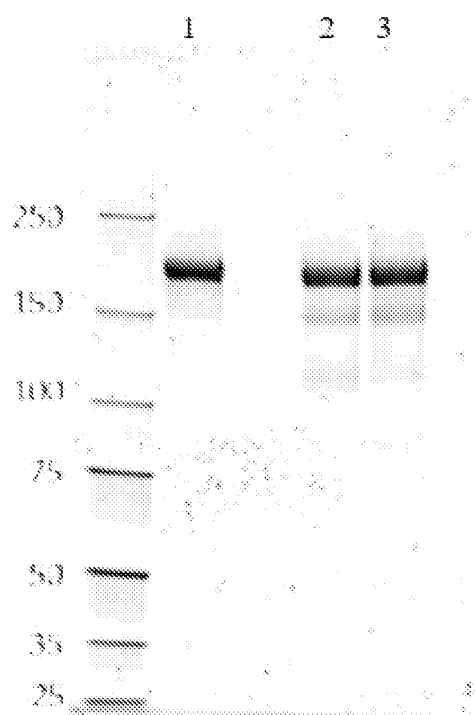


Figure 11

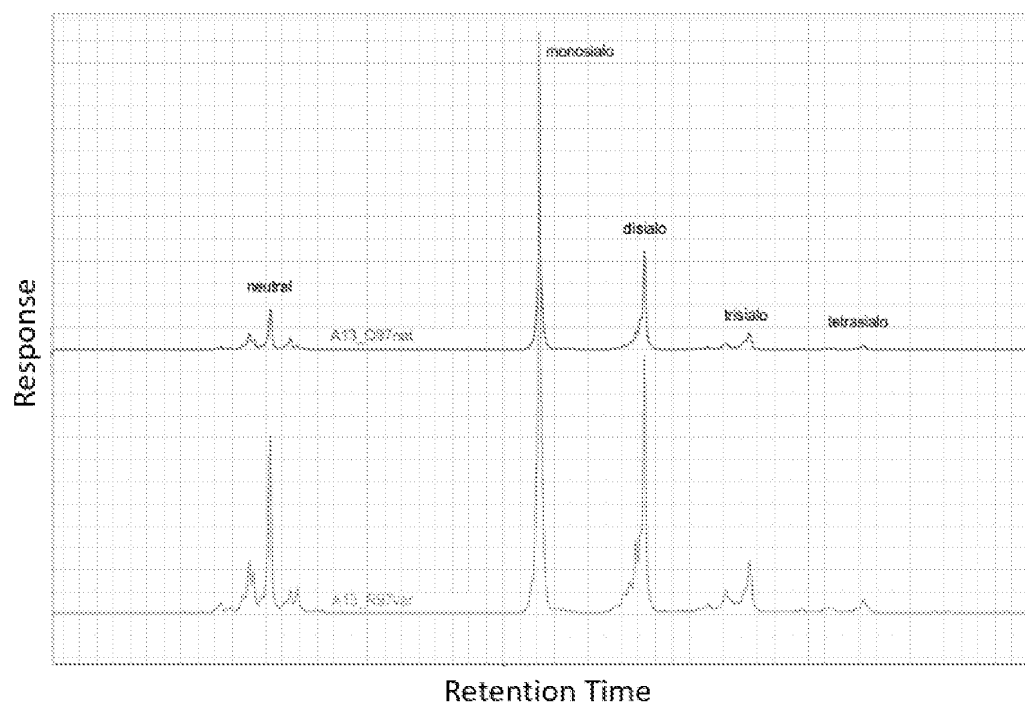


Figure 1A

Wildtype ADAMTS13 – Top (SEQ ID NO: 1)
Q⁹⁷R Variant of ADAMTS13 – Bottom (SEQ ID NO: 2)

```

1  mhqrhprancppicvagilacgfilgcwgsphfqqscilaqlepqavssyl  50
1  mhqrhprancppicvagilacgfilgcwgsphfqqscilaqlepqavssyl  50
51  spgaplkgrppspgfgqrqqrqrrnaaggilhllellvavgpdvfqahgedt  100
51  spgaplkgrppspgfgqrqqrqrrnaaggilhllellvavgpdvfqahredt  100
101  eryvitnlngiaellrdpslgaqfrvhlvkmviltpegapnitanitss  150
101  eryvitnlngiaellrdpslgaqfrvhlvkmviltpegapnitanitss  150
151  llsvcgwsqtinpedddtpghadlvlyitrfdlelpgdnrqnrvgtqlgg  200
151  llsvcgwsqtinpedddtpghadlvlyitrfdlelpgdnrqnrvgtqlgg  200
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201  acsptwscilitetgfdlgtviahhighsfglehdgapgsgcgpsghvma  250
251  sdgaapraglewspcsnrqlislsisagracvwpprpqpsaghpdaq  300
251  sdgaapraglewspcsnrqlislsisagracvwpprpqpsaghpdaq  300
301  pgllyसानेकरवाफेगकवाक्टफारेहलमकालसचतदपलदसससल  350
301  pgllyसानेकरवाफेगकवाक्टफारेहलमकालसचतदपलदसससल  350
351  lvpildgtetgvekwcsgkncrsilveltpiaavhgrwsswgrpspcsrsc  400
351  lvpildgtetgvekwcsgkncrsilveltpiaavhgrwsswgrpspcsrsc  400
401  gggvvttrrrqcnnpafggracvgaadiqaeemcntqacektqlfmsqqc  450
401  gggvvttrrrqcnnpafggracvgaadiqaeemcntqacektqlfmsqqc  450
451  artdgqplrsspggasfyhwaavphsqgdalcrhmraigesfimkrqd  500
451  artdgqplrsspggasfyhwaavphsqgdalcrhmraigesfimkrqd  500

```

Figure 1B

```

501  sflldgtrcmppsgpredgtlsicvsgscrtfpcdgrmdsqqvwdrcqvcgg  550
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551  dnstcspnkgsftagrareyvtflvtvtpnltsvyianhrplfthlavrig  600
551  dnstcspnkgsftagrareyvtflvtvtpnltsvyianhrplfthlavrig  600
601  gryvvagkmsispnttyslledgrveyrvaltedrlpleeiriwglp  650
601  gryvvagkmsispnttyslledgrveyrvaltedrlpleeiriwglp  650
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701  aglrwnvysclldqarkelvetvqcqsgsqppawpescvlepcppywavgd  750
751  fgpcsasccggglrerpvrveaagsllktlpparcragaaqpavaletcn  800
751  fgpcsasccggglrerpvrveaagsllktlpparcragaaqpavaletcn  800
801  pqpcparwevsepssctsaggaglalenetcvpgadgleapvteggpsvd  850
801  pqpcparwevsepssctsaggaglalenetcvpgadgleapvteggpsvd  850
851  ekilpapepcvgmscpggwhldatsagekapspwgsirtgaqaahwtpa  900
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901  agscsvscgrglmelrflcmdsalrvpqeclgaskpgsrrevcqavp  950
901  agscsvscgrglmelrflcmdsalrvpqeclgaskpgsrrevcqavp  950
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951  cparwqyklaacsvscgrgvvrrilycarahgeddgeeilldtacqglpr  1000

```

Figure 1C

```

1001  pepqeacslepccprwkvmslpgcsasccgigtarrsvacvqldaqdv  1050
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1401  lngqywtlqswvpemadpqswkgegt  1427
1401  lngqywtlqswvpemadpqswkgegt  1427

```

Figure 14A

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1  atgacaccagc  gtacacccccg  ggaagatgc  cctccccctct  gtgtggccgg  aatccttgcc
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121  ttggagccac  aggcogtgtc  ttcttacttg  agccttggtg  ctcccttaaa  aggcogcctc
181  ccttccccctg  gcttccagag  gcagaggcag  aggcagaggc  gggctgcagg  cggcatccta
241  caactggagc  tctgtgtggc  cgtgggccc  gatgtctcc  aggcctacca  ggaggacaca
301  gagcgctatg  tcttcaccaa  cctcaacatc  ggggcagaac  tgcttcggga  cccgtccctg
361  ggggctcagt  ttgggttgca  cctggtgaag  atggtcattc  tgacagagcc  tgagggtgct
421  ccaaatatca  cagccaacct  cactctgtcc  ctgctgagcg  tctgtgggtg  gagccagacc
481  atcaaacctg  aggacgacac  ggaatcctgg  catgctgacc  tggctcctca  tatcactagg
541  tttgacctgg  agttgcctga  tggtaacccg  cagggtgcgg  gcgtcaccca  gctgggcggg
601  gcctgctccc  caacctggag  ctgcctcatt  accgaggaca  ctggcttcca  cctgggagtc
661  accattgccc  atgagattgg  gcacagcttc  ggcctggagg  acgacggcgc  gcccggcagc
721  ggcgtggggc  ccagcggaca  cgtgatggct  tcggacggcg  ccgcgcgccg  cggcggcctc
781  gctgtgtccc  cctgcagccg  ccggcagctg  ctgagcctgc  tcagcgcagg  accggcgcgc
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901  cctggcctct  actacagcgc  caacgagcag  tgcgcgtgg  ccttcggccc  caaggctgtc
961  gctgcacct  togcacggga  gcacctggat  atgtgcacgg  cctctcctct  ccacacagac
1021  ccgctggacc  aaagcagctg  cagccgcctc  ctgcttccct  tcttgatgg  gacagaatgt
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1141  gcagcagtg  atgggcgctg  gtctagctgg  ggtcccccga  gtcccttgct  ccgctcctgc
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1381  tccccctggc  gcgcctcctt  ctaccactgg  ggtgtgtctg  taccacacag  ccaaggggat
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1501  agcttcctcg  atgggacccg  gtgtatgcca  agtggccccc  gggaggacgg  gacccctgagc
1561  ctgtgtgtgt  cgggcagctg  caggacattt  ggctgtgatg  gtaggatgga  ctcccagcag
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1681  tctttcacag  ctggcagagc  gagagaatat  gtcaagtctc  tgacagttac  ccccaacctg
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1861  ctggaggatg  gtctgtgtga  gtaacagatg  gccctcaccc  aggaacgggt  gccccgctg
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2101  gcaggcctgc  gctgggtaaa  ctacagctgc  ctggacccag  ccagggaagg  gttggtggag
2161  actgtccagt  gccaaaggag  ccagcagcca  ccagcgtggc  cagaggcctg  cgtgctcgaa
2221  cctgcccctc  cctactgggc  ggtgggagac  ttccggccat  gcagcgcctc  ctgtgggggt
2281  ggcctgcggg  agcggccagt  gcgtgcgtg  gagggccagg  gcagcctct  gaagacattg
2341  cccccagccc  ggtgcagagc  aggggcccag  cagccagctg  tggcgctgga  aacctgcaac

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Figure 14B

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2521 ccagtgaactg aggggocctgg ctccgtagat gagaagctgc ctgcccctga gccctgtgtc
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2761 gactctgccc tcagggtgcc tgtccaggaa gagctgtgtg gcctggcaag caagcctggg
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