

Supplemental Data for

Phylogenetic and functional analysis of ADAMTS13 identifies highly conserved domains essential for allosteric regulation

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Supplemental Table 1. ADAMTS13 domain structure for selected vertebrate species

Supplemental Table 2. Inhibition of vertebrate plasma ADAMTS13 by monoclonal antibody 3H9

Supplemental Figure 1. Phylogram of vertebrate ADAMTS13

Supplemental Figure 2. Vertebrate ADAMTS13 gene structure

Supplemental Table 1. ADAMTS13 domain structure for selected vertebrate species

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No.	Tax ID	Organism name	Common name	Domains absent ^a	Furin site sequence ^b	GenBank accession number
1	57068	<i>Acanthisitta chloris</i>	Rifleman	T3, T6a, T6	RLQKR	BK009996
2	32536	<i>Acinonyx jubatus</i>	cheetah	T3	RLRR	BK009997
3	10068	<i>Acomys cahirinus</i>	Egyptian spiny mouse	Long propeptide	RWRR	BK009998
4	8715	<i>Agkistrodon piscivorus</i>	cottonmouth	T3, T6a	RIRR	BK009999
5	9646	<i>Ailuropoda melanoleuca</i>	Giant panda	T6	RVRR	BK010000
6	9079	<i>Alectoris rufa</i>	red-legged partridge	T3, T6a	RLQKR	BK010001
7	8496	<i>Alligator mississippiensis</i>	American alligator	None	RHWKR	BK010002
8	38654	<i>Alligator sinensis</i>	Chinese alligator	None	RHWKR	BK010003
9	12930	<i>Amazona aestiva</i>	blue-fronted amazon	T3, T6a, T6	RLQKR	BK010004
10	241585	<i>Amazona vittata</i>	Puerto Rican parrot	T3, T6a, T6	RLQKR	BK010005
11	61819	<i>Amphilophus citrinellus</i>	Midas cichlid	Long propeptide, T6a	RLRR	BK010006
12	8839	<i>Anas platyrhynchos</i>	Mallard	T3, T6a	RLQKR	XM_005013264.2
13	7936	<i>Anguilla anguilla</i>	European eel	T6a	GPRRVLR	BK010007
14	7937	<i>Anguilla japonica</i>	Japanese eel	T6a	GPRRVLR	BK010008
15	229290	<i>Anoplopoma fimbria</i>	sablefish	Long propeptide, T6a	RRLRR	BK010009
16	8845	<i>Anser cygnoides domesticus</i>	Swan goose	T3, T6a	RLQKR	BK010010
17	37293	<i>Aotus nancymaae</i>	Ma's night monkey	Long propeptide, T6a	RQRR	BK010011
18	57397	<i>Apaloderma vittatum</i>	Bar-tailed trogon	T3, T6a, T6	RLQKR	BK010012
19	55534	<i>Apalone spinifera</i>	Spiny softshell turtle	None	RLWKR	BK010013
20	10129	<i>Apodemus sylvaticus</i>	European woodmouse	Long propeptide, T6	RLRRRR	BK010014
21	9233	<i>Aptenodytes forsteri</i>	Emperor penguin	T3, T6a, T6	RLQKR	XM_009286970.1
22	8822	<i>Apteryx australis mantelli</i>	brown kiwi	T3, T6a	RLQKR	XM_013941757.1
23	216574	<i>Aquila chrysaetos canadensis</i>	Canadian golden eagle	T3, T6a, T6	RLQKR	BK010015
24	7994	<i>Astyanax mexicanus</i>	Mexican tetra	T6a	HHRIQR	BK010016
25	9368	<i>Atelerix albiventris</i>	middle-African hedgehog	Long propeptide, T3, T6a	TPSSGLQSPA	BK010017

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26	52670	<i>Austrofundulus limnaeus</i>	Venezuelan mosquito fish	Long propeptide, T6a	RLRR	XM_014007162.1
27	310752	<i>Balaenoptera acutorostrata scammoni</i>	Minke whale	Long propeptide	RLRRR	BK010018
28	100784	<i>Balearica regulorum gibbericeps</i>	East African grey crowned-crane	T3, T6a, T6	RLQKR	BK010019
29	43346	<i>Bison bison bison</i>	American bison	Long propeptide	RLRRR	BK010020
30	39328	<i>Boaedon fuliginosus</i>	(African) brown house snake	T3, T6a	RIRR	BK010021
31	150288	<i>Boleophthalmus pectinirostris</i>	great blue-spotted mudskipper	Long propeptide, T6a	RQYRS	BK010022
32	9915	<i>Bos indicus</i>	Zebu	Long propeptide	RLRRR	BK010023
33	9913	<i>Bos taurus</i>	Cattle	Long propeptide	RLRRR	BK010024
34	89462	<i>Bubalus bubalis</i>	Water buffalo	Long propeptide	RLRRQ	BK010025
35	30397	<i>Buteo buteo</i>	Common buzzard	T3, T6a, T6	RLQKR	BK010026
36	198806	<i>Calidris pugnax</i>	Ruff	T3, T6a, T6	RLRKR	XM_014943048.1
37	9483	<i>Callithrix jacchus</i>	White-tufted-ear marmoset	Long propeptide	RQRR	BK010027
38	7868	<i>Callorhynchus milii</i>	Elephant shark	Long propeptide, T6a	HVRAR	XM_007902869.1
39	9244	<i>Calypte anna</i>	Anna's hummingbird	T3, T6a, T6	RLQKR	BK010028
40	9837	<i>Camelus bactrianus</i>	Bactrian camel	T6	RLWRR	BK010029
41	9838	<i>Camelus dromedarius</i>	Arabian camel	T6	RLWRR	BK010030
42	51853	<i>Candoia aspera</i>	Viper boa	T3, T6a	RLRR	BK010031
43	9615	<i>Canis lupus familiaris</i>	Domestic dog	T3, T6a	RLRR	NM_001242712.1
44	9925	<i>Capra hircus</i>	Goat	Long propeptide	RLRR	XM_018056149.1
45	279965	<i>Caprimulgus carolinensis</i>	Chuck-will's-widow	T3, T6a, T6	RLQKR	BK010032
46	54380	<i>Cariama cristata</i>	Red-legged seriema	T3, T6a, T6	RLQKR	BK010033
47	9478	<i>Carlito syrichta</i>	Philippine tarsier	Long propeptide, T6a, T6	RQRR	BK010034
48	43455	<i>Cathartes aura</i>	turkey vulture	T3, T6a	RLQKR	BK010035
49	10141	<i>Cavia porcellus</i>	Guinea pig	T3, T6a, T6	RRRRR	XM_013155744.1
50	73337	<i>Ceratotherium simum simum</i>	Southern white rhinoceros	Long propeptide	RRLQR	BK010036

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No.	Tax ID	Organism name	Common name	Domains absent ^a	Furin site sequence ^b	GenBank accession number
51	9531	<i>Cercocebus atys</i>	Sooty mangabey	Long propeptide, T6a	RQRR	BK010037
52	8897	<i>Chaetura pelagica</i>	Chimney swift	T3, T6a, T6	RLQKR	BK010038
53	50402	<i>Charadrius vociferus</i>	Killdeer	T3, T6a, T6	RLQKR	BK010039
54	8469	<i>Chelonia mydas</i>	Green sea turtle	None	RLWKR	BK010040
55	8475	<i>Chelydra serpentina</i>	snapping turtle	None	RLWKR	BK010041
56	586796	<i>Chiloscyllium griseum</i>	Grey bambooshark	None	RRSVR	BK010042
57	34839	<i>Chinchilla lanigera</i>	Long-tailed chinchilla	T3, T6	RQRRRRRWGR	XM_013506558.1
58	187382	<i>Chlamydotis macqueenii</i>	Macqueen's bustard	T3, T6a, T6	RLQKR	BK010043
59	60711	<i>Chlorocebus sabaeus</i>	Green monkey	Long propeptide, T6a	RQRR	XM_008005775.1
60	9358	<i>Choloepus hoffmanni</i>	Hoffmann's two-fingered sloth	None	RLPR	Incomplete exon 7, exon 8
61	8479	<i>Chrysemys picta</i>	Western painted turtle	None	RLWKR	BK010044
62	185453	<i>Chrysochloris asiatica</i>	Cape golden mole	Long propeptide, T3, T4, T6	RLPRRAR	BK010045
63	7950	<i>Clupea harengus</i>	Atlantic herring	T6a	RRQRR	BK010046
64	30724	<i>Clupea pallasii</i>	Pacific herring	T6a	RRQRR	BK010047
65	9014	<i>Colinus virginianus</i>	northern bobwhite	T3, T6a	RLQKR	BK010048
66	57412	<i>Colius striatus</i>	Speckled mousebird	T3, T6a, T6	RLQKR	BK010049
67	336983	<i>Colobus angolensis</i>	Angolan colobus	Long propeptide, T6a	RQRR	BK010050
68	8932	<i>Columba livia</i>	Rock pigeon	T2 (4 Cys), T3, T4, T5 (2 Cys), T6a, T6	RLQKR	BK010051
69	85066	<i>Corvus brachyrhynchos</i>	American crow	T3, T6a, T6	RLQKR	BK010052
70	446433	<i>Cottus rhenanus</i>	Rheingroppe	Long propeptide, T6a	RLRR	BK010053
71	9091	<i>Coturnix coturnix</i>	Common quail	T3, T6a	RLQKR	BK010054
72	10029	<i>Cricetulus griseus</i>	Chinese hamster	Long propeptide	RLRWRR	BK010055
73	8501	<i>Crocodylus niloticus</i>	Nile crocodile	None	KEKKFHPRHWKR	BK010056
74	8502	<i>Crocodylus porosus</i>	Australian saltwater crocodile	None	RHWKR	XM_019540879.1

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75	384069	<i>Crotalus mitchellii pyrrhus</i>	speckled rattlesnake	T3, T6a	RIRR	BK010057
76	55661	<i>Cuculus canorus</i>	Common cuckoo	T3, T6a, T6	RLQKR	BK010058
77	156563	<i>Cyanistes caeruleus</i>	Blue tit	T3, T6a, T6	RLHKR	BK010059
78	193059	<i>Cyanopica cyanus</i>	azure-winged magpie	T3, T6a, T6	RLQKR	BK010060
79	69242	<i>Cyprinodon nevadensis pectoralis</i>	Amargosa pupfish	Long propeptide, T6a	RLRR	BK010061
80	28743	<i>Cyprinodon variegatus</i>	sheepshead minnow	Long propeptide, T6a	RLRR	BK010062
81	7962	<i>Cyprinus carpio</i>	common carp	T6a	RLRR	BK010063
82	7955	<i>Danio rerio</i>	Zebrafish	T3, T6a	RLRR	XM_002663221.5
83	9361	<i>Dasyus novemcinctus</i>	Nine-banded armadillo	Long propeptide, T3, T6, ½ CUB1, CUB2	RPLSR	BK010064
84	31869	<i>Daubentonia madagascariensis</i>	Aye aye	Long propeptide, T6a	RRQRR	BK010065
85	13489	<i>Dicentrarchus labrax</i>	European sea bass	Long propeptide, T6a	RLRR	BK010066
86	10020	<i>Dipodomys ordii</i>	Ord's kangaroo rat	Long propeptide, T4 (part), T6a, T6, CUB1, CUB2	QRRR	BK010067
87	9371	<i>Echinops telfairi</i>	Madagascar hedgehog	Long propeptide, T3, T6a, T6	LLGQTR	BK010068
88	187118	<i>Ectopistes migratorius</i>	Passenger pigeon	T2 (4 Cys), T3, T4, T5 (2 Cys), T6a, T6	RLQKR	BK010069
89	188379	<i>Egretta garzetta</i>	Little egret	T3, T6a, T6	RLQKR	BK010070
90	28737	<i>Elephantulus edwardii</i>	Cape elephant shrew	Long propeptide, T3, T6	FPRNR	BK010071
91	9783	<i>Elephas maximus</i>	Asiatic elephant	Long propeptide, T3, T6	LQRQTR	BK010072
92	71008	<i>Elgaria multicarinata</i>	California alligator lizard	T3	RIRR	BK010073
93	29078	<i>Eptesicus fuscus</i>	Big brown bat	Long propeptide	LRSR	BK010074
94	9793	<i>Equus asinus</i>	Ass	None	RLQRR	BK010075
95	9796	<i>Equus caballus</i>	Horse	None	RLQRR	BK010076
96	9365	<i>Erinaceus europaeus</i>	Western European hedgehog	Long propeptide, T3, T6a	TPSSGLQSPA	BK010077
97	8010	<i>Esox lucius</i>	Northern pike	T6a	HMKRLPRSR	XM_010895705.2
98	481883	<i>Eublepharis macularius</i>	leopard gecko	T3, T6a	RVKR	BK010078

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99	87288	<i>Eulemur flavifrons</i>	Sclater's lemur	Long propeptide, T6a	RRQRR	BK010079
100	54383	<i>Eurypyga helias</i>	Sunbittern	T3, T6a, T6	RLQKR	BK010080
101	345164	<i>Falco cherrug</i>	Saker falcon	T3, T6a, T6	RLQKR	BK010081
102	9685	<i>Felis catus</i>	Domestic cat	T3	RLRR	BK010082
103	59894	<i>Ficedula albicollis</i>	Collared flycatcher	T3, T6a, T6	RLHKR	XM_005055623.2
104	885580	<i>Fukomys damarensis</i>	Damara mole-rat	Long propeptide, T3, T6	RRTRRR	XM_010608913.1
105	30455	<i>Fulmarus glacialis</i>	Northern fulmar	T3, T6a, T6	RLQKR	BK010083
106	8078	<i>Fundulus heteroclitus</i>	Mummichog, Atlantic killifish	Long propeptide, T6a	RLRR	XM_012866268.1
107	8049	<i>Gadus morhua</i>	Atlantic cod	Long propeptide, T6a	PGQRRWPR	BK010084
108	482537	<i>Galeopterus variegatus</i>	Sunda flying lemur	None	RLRLRR	BK010085
109	9031	<i>Gallus gallus</i>	Chicken	T3, T6a	RLRR	XM_004945941.2
110	69293	<i>Gasterosteus aculeatus</i>	Three-spined stickleback	Long propeptide, T6a	RLQKR	BK010086
111	37040	<i>Gavia stellata</i>	Red-throated loon	T3, T6a, T6	RLQKR	BK010087
112	94835	<i>Gavialis gangeticus</i>	gharial	None	RLWKR	BK010088
113	48883	<i>Geospiza fortis</i>	Medium ground finch	T3, T6a, T6	RLHKR	BK010089
114	9593	<i>Gorilla gorilla</i>	Gorilla	Long propeptide, T6a	RQRR	BK010090
115	8969	<i>Haliaeetus albicilla</i>	White-tailed eagle	T3, T6a, T6	RLQKR	BK010091
116	8153	<i>Haplochromis burtoni</i>	Burton's mouthbreeder	Long propeptide, T6a	RLRR	XM_005926926.2
117	10181	<i>Heterocephalus glaber</i>	Naked mole-rat	T3, T6	RRTRRR	XM_013073846.1
118	9606	<i>Homo sapiens</i>	Human	Long propeptide, T6a	RQRR	AY055376
119	7998	<i>Ictalurus punctatus</i>	Channel catfish	T6a	RRLQR	XM_017467535.1
120	43179	<i>Ictidomys tridecemlineatus</i>	Thirteen-lined ground squirrel	Long propeptide, T6	RLQRRRR	XM_005337003.1
121	51337	<i>Jaculus jaculus</i>	Lesser Egyptian jerboa	Long propeptide	RLRKTR	BK010092
122	215358	<i>Larimichthys crocea</i>	Large yellow croaker	Long propeptide, T6a	RLRR	XM_019274637.1
123	8187	<i>Lates calcarifer</i>	barramundi perch	Long propeptide, T6a	RRLRR	XM_018669486.1
124	7897	<i>Latimeria chalumnae</i>	Coelacanth	Long propeptide	KDRKPALQVHRR	XM_005990607.1

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125	7918	<i>Lepisosteus oculatus</i>	Spotted gar	Long propeptide, T3, T6a	RRLRR	BK010093
126	9713	<i>Leptonychotes weddellii</i>	Weddell seal	T3, T6	RVRR	BK010094
127	188344	<i>Leptosomus discolor</i>	Cuckoo roller	T3, T6a	RLRKR	BK010095
128	7782	<i>Leucoraja erinacea</i>	little skate	Long propeptide	HIRRR	BK010096
129	9785	<i>Loxodonta africana</i>	African elephant	Long propeptide, T3, T6	QRQTR	BK010097
130	162951	<i>Lyrurus tetrix tetrix</i>	black grouse	T3, T6a	RLQKR	BK010098
131	9541	<i>Macaca fascicularis</i>	Crab-eating macaque, cynomolgus	Long propeptide, T6a	RQRR	BK010099
132	9544	<i>Macaca mulatta</i>	Rhesus macaque	Long propeptide, T6a	RQRR	BK010100
133	1356389	<i>Machlolophus spilonotus</i>	yellow-cheeked tit	T3, T6a, T6	RLHKR	BK010101
134	9315	<i>Macropus eugenii</i>	Tammar Wallaby	None	RISKR	BK010102
135	328815	<i>Manacus vitellinus</i>	Golden-collared manakin	T3, T6a, T6	RLQKR	XM_018074289.1
136	9568	<i>Mandrillus leucophaeus</i>	Drill	Long propeptide, T6a	RQRR	BK010103
137	143292	<i>Manis pentadactyla</i>	Chinese pangolin	Long propeptide	RLRR	BK010104
138	9995	<i>Marmota monax</i>	woodchuck	Long propeptide (Exons 2-2a), T6	RLQRRRR	BK010105
139	9103	<i>Meleagris gallopavo</i>	Turkey	T3, T6a	RLQKR	XM_010720964.2
140	13146	<i>Melopsittacus undulatus</i>	Budgerigar	T3, T6a, T6	RLQKR	BK010106
141	10047	<i>Meriones unguiculatus</i>	Mongolian gerbil	Long propeptide	RLRWRR	BK010107
142	57421	<i>Merops nubicus</i>	Carmine bee-eater	T3, T6a, T6	RLQKR	BK010108
143	54374	<i>Mesitornis unicolor</i>	Brown roatelo	T3, T6a, T6	RLQKR	BK010109
144	10036	<i>Mesocricetus auratus</i>	Golden hamster	Long propeptide, T6a	RLRWRR	XM_013124700.1
145	30608	<i>Microcebus murinus</i>	Grey mouse lemur	Long propeptide, T6a	RRQRR	XM_012781995.1
146	79684	<i>Microtus ochrogaster</i>	Prairie vole	Long propeptide, T6a	RLRWRR	BK010110
147	47230	<i>Microtus arvalis</i>	Common vole	Long propeptide, T6a	RLRWRR	BK010111
148	13616	<i>Monodelphis domestica</i>	Gray short-tailed opossum	None	RISKR	XM_001371685.4

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149	10090	<i>Mus musculus</i>	House mouse	Long propeptide, T6	RLRRRR	AB112362.1
150	9669	<i>Mustela putorius furo</i>	Domestic ferret	None	RVRRR	XM_013058572.1
151	447135	<i>Myodes glareolus</i>	Bank vole	Long propeptide, T6a	RLRWRR	BK010112
152	109478	<i>Myotis brandtii</i>	Brandt's bat	Long propeptide, T6a	RLRR	BK010113
153	225400	<i>Myotis davidii</i>	David's myotis	Long propeptide, T6a	RLRR	BK010114
154	59463	<i>Myotis lucifugus</i>	Little brown bat	Long propeptide, T6a	RLRR	BK010115
155	1026970	<i>Nannospalax galili</i>	Upper Galilee mountains blind mole rat	Long propeptide, T6a, ½ CUB1, CUB2	HLKRLRWGR	BK010116
156	125878	<i>Nanorana parkeri</i>	high Himalaya frog	None	HLSKR	XM_018563708.1
157	32507	<i>Neolamprologus brichardi</i>	lyretail cichlid	Long propeptide, T6a	RLRR	BK010117
158	176057	<i>Nestor notabilis</i>	Kea	T3, T6a, T6	RLQKR	BK010118
159	128390	<i>Nipponia nippon</i>	Crested ibis	T3, T6a, T6	RLQKR	BK010119
160	61853	<i>Nomascus leucogenys</i>	Northern white cheeked gibbon	Long propeptide, T6a	RQRRRQRR	BK010120
161	105023	<i>Nothobranchius furzeri</i>	turquoise killifish	Long propeptide, T6a	RLRR	XM_015972136.1
162	8208	<i>Notothenia coriiceps</i>	black rockcod	Long propeptide, T6a	RLPR	BK010121
163	8996	<i>Numida meleagris</i>	helmeted guineafowl	T3, T6a	RLRKR	BK010122
164	9978	<i>Ochotona princeps</i>	American pike	Long propeptide	PPRR	BK010123
165	10160	<i>Octodon degus</i>	Degu	T3, T6	RGTRRWRR	BK010124
166	9708	<i>Odobenus rosmarus divergens</i>	Pacific walrus	T3, T6	RIRR	BK010125
167	8022	<i>Oncorhynchus mykiss</i>	Rainbow trout	T6a	KRLRRRR	BK010126
168	8665	<i>Ophiophagus hannah</i>	king cobra	T3, T6a	RIRR	BK010127
169	30419	<i>Opisthocomus hoazin</i>	Hoatzin	T3, T6a, T6	RLQKR	BK010128
170	9733	<i>Orcinus orca</i>	killer whale	Long propeptide	RPRR	BK010129
171	8128	<i>Oreochromis niloticus</i>	Nile tilapia	Long propeptide, T6a	RLRR	XM_013271591.2
172	9258	<i>Ornithorhynchus anatinus</i>	Platypus	T6a	RIWKR	BK010130
173	1230840	<i>Orycteropus afer afer</i>	Aardvark	Long propeptide, T3, T6	HLRRWIR	BK010131

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174	9986	<i>Oryctolagus cuniculus</i>	Rabbit	Long propeptide	SRRRR	BK010132
175	8090	<i>Oryzias latipes</i>	Japanese medaka, rice fish	Long propeptide, T6a	RLHR	BK010133
176	109271	<i>Osteoglossum bicirrhosum</i>	Silver arawana	T6a	RLKR	BK010134
177	30611	<i>Otolemur garnettii</i>	Small-eared galago	Long propeptide, T3, T6a	RWRR	BK010135
178	9940	<i>Ovis aries</i>	Sheep	Long propeptide	RLRR	XM_012118015.1
179	9598	<i>Pan troglodytes</i>	Chimpanzee	Long propeptide, T6a	RQRR	BK010136
180	9694	<i>Panthera tigris</i>	Tiger (all subspecies)	T3	RLRR	BK010137
181	94885	<i>Pantherophis guttatus</i>	Red corn snake	T3, T6a	RIRR	BK010138
182	9555	<i>Papio anubis</i>	olive baboon	Long propeptide, T6a	RQRR	BK010139
183	1071634	<i>Paradoxornis webbiana</i> <i>bulomachus</i>	Crow tit	T3, T6a, T6	RLHKR	BK010140
184	9157	<i>Parus major</i>	Great tit	T3, T6a, T6	RLHKR	XM_015644841.2
185	9049	<i>Pavo cristatus</i>	Indian peafowl	T3, T6a	RLQKR	BK010141
186	36300	<i>Pelecanus crispus</i>	Dalmatian pelican	T3, T6a, T6	PRQKR	BK010142
187	13735	<i>Pelodiscus sinensis</i>	Chinese soft-shelled turtle	None	RLWKR	BK010143
188	367368	<i>Pelusios castaneus</i>	West African mud turtle, sideneck turtle	None	RLRKR	BK010144
189	9052	<i>Perdix perdix</i>	Grey partridge	T3, T6a	RLQKR	BK010145
190	1365757	<i>Periophthalmodon schlosseri</i>	giant mudskipper	Long propeptide, T6a	RQYRSVK	BK010146
191	409849	<i>Periophthalmus magnuspinnatus</i>	Common mudskipper	Long propeptide, T6a	RQYRSVK	BK010147
192	10041	<i>Peromyscus leucopus</i>	White footed mouse	Long propeptide	RLRWRR	BK010148
193	230844	<i>Peromyscus maniculatus bairdii</i>	Prairie deer mouse	Long propeptide	RLRWRR	XM_015986629.1
194	97097	<i>Phaethon lepturus</i>	White-tailed tropicbird	T3, T6a	RLQKR	BK010149
195	9209	<i>Phalacrocorax carbo</i>	Great cormorant	T3, T6a, T6	RLQKR	BK010150
196	9054	<i>Phasianus colchicus</i>	Ring-necked pheasant	T3, T6a	RLQKR	BK010151
197	9218	<i>Phoenicopterus ruber ruber</i>	American flamingo	T3, T6a, T6	RLQKR	BK010152

Supplemental Table 1. ADAMTS13 domain structure for selected vertebrate species
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No.	Tax ID	Organism name	Common name	Domains absent ^a	Furin site sequence ^b	GenBank accession number
198	9755	Physeter catodon	Sperm whale	Long propeptide	RLRRR	BK010153
199	118200	Picoides pubescens	Downy woodpecker	T3, T6a, T6	RRRRR	BK010154
200	90988	Pimephales promelas	fathead minnow	T6a	RLRR	BK010155
201	345573	Podiceps cristatus	great crested grebe	T3, T6a, T6	RLQKR	BK010156
202	257846	Podoces hendersoni	Mongolian ground-jay	T3, T6a, T6	RLQKR	BK010157
203	48698	Poecilia formosa	Amazon molly	Long propeptide, T6a	RLRR	XM_007565190.2
204	48699	Poecilia latipinna	sailfin molly	Long propeptide, T6a	RLRR	XM_015021450.1
205	48701	Poecilia mexicana	shortfin molly	Long propeptide, T6a	RLRR	XM_014986798.1
206	8081	Poecilia reticulata	Guppy	Long propeptide, T6a	RLRR	XM_008419233.2
207	103695	Pogona vitticeps,	central bearded dragon	Long propeptide	RVRRK	BK010158
208	9601	Pongo abelii	Orangutan	Long propeptide, T6a	RQRR	BK010159
209	9813	Procavia capensis	Cape rock hyrax	Long propeptide, T3, T6a, T6	KSRPASPGPQTQ TWV	BK010160
210	379532	Propithecus coquereli	Coquerel's sifaka	Long propeptide, T6a	KGLRRESR	XM_012664714.1
211	103944	Protobothrops mucrosquamatus	Chinese Habu snake	T3, T6a	RIRR	BK010161
212	245875	Pseudopleuronectes yokohamae	marbled flounder	Long propeptide, T6a	GVRLPRSAA	BK010162
213	181119	Pseudopodoces humilis	Tibetan ground-tit	T3, T6a, T6	RLHKR	XM_014258024.1
214	240206	Pterocles gutturalis	Yellow-throated sandgrouse	T3, T6a, T6	RLQKR	BK010163
215	303518	Pundamilia nyererei	(a Lake Victoria cichlid)	Long propeptide, T6a	RLRR	BK010164
216	42514	Pygocentrus nattereri	red-bellied piranha	T6a	RLQR	XM_017696426.1
217	9238	Pygoscelis adeliae	Adelie penguin	T3, T6a, T6	RLQKR	BK010165
218	176946	Python bivittatus	Burmese python	T3, T6a	RIRR	BK010166
219	10116	Rattus norvegicus	Norway rat	Long propeptide, T6	RQRR	XM_006233879.2
220	163638	Rhamphochromis esox	Tigerfish (cichlid)	Long propeptide, T6a	RLRR	BK010167
221	259920	Rhincodon typus	Whale shark	None	RSRSSLVTRR	BK010168
222	61622	Rhinopithecus roxellana	Golden snub-nosed monkey	Long propeptide, T6a	RQRR	BK010169

Supplemental Table 1. ADAMTS13 domain structure for selected vertebrate species
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No.	Tax ID	Organism name	Common name	Domains absent ^a	Furin site sequence ^b	GenBank accession number
223	9407	Rousettus aegyptiacus	Egyptian rousette (bat)	None	RPSRPPRH	BK010170
224	8030	Salmo salar	Atlantic salmon	T6a	HMKRLRSRR	XM_014160257.1
225	9305	Sarcophilus harrisii	Tasmanian devil	None	RISKR	XM_012553998.1
226	166764	Scartelaos histophorus	walking goby	Long propeptide, T6a	RQHRSVK	BK010171
227	155319	Scincella lateralis	ground skink	Long propeptide, T3	RVRK	BK010172
228	113540	Scleropages formosus	Asian bonytongue	T6a	RLKR	XM_018758492.1
229	7830	Scyliorhinus canicula	Smaller spotted catshark	T6a	RSSLVRR	BK010173
230	72099	Sebastes rubrivinctus	flag rockfish	Long propeptide, T6a	RPRR	BK010174
231	9135	Serinus canaria	Common canary	T3, T6a, T6	RLHKR	BK010175
232	1608454	Sinocyclocheilus anshuiensis	Eyeless golden-line barbel	T6a	RLRR	BK010176
233	42254	Sorex araneus	European shrew	Long propeptide, T3, T6a	PPPPSGQSWA	XM_004613415.1
234	144197	Stegastes partitus	Bicolor damselfish	Long propeptide, T6a	RLRR	BK010177
235	142480	Sternotherus odoratus	stinkpot turtle	None	RLWKR	BK010178
236	441894	Struthio camelus australis	African ostrich	T3, T6a	RLQKR	BK010179
237	9172	Sturnus vulgaris	Common starling	T3, T6a, T6	RLHKR	XM_014873269.1
238	9378	Suncus murinus	House shrew	Long propeptide, T3, T4, T6a	SETPWK	BK010180
239	9823	Sus scrofa	Pig	Long propeptide	RPPARRLWRR	XM_013987089.1
240	59729	Taeniopygia guttata	Zebra finch	T3, T6a, T6	RLHKR	BK010181
241	31033	Takifugu rubripes	Fugu	Long propeptide, T6a	RARR	XM_011615207.1
242	121530	Tauraco erythrolophus	Red crested turaco	T3, T6a, T6	RLRKR	BK010182
243	158814	Terrapene carolina	box turtle	None	RLWKR	BK010183
244	99883	Tetraodon nigroviridis	Spotted green pufferfish	Long propeptide, T6a	RGRLPR	BK010184
245	35003	Thamnophis couchii	Sierra garter snake	T3, T6a	RIRR	BK010185
246	35005	Thamnophis elegans	Western terrestrial garter snake	T3, T6a	RIRR	BK010186

Supplemental Table 1. ADAMTS13 domain structure for selected vertebrate species

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No.	Tax ID	Organism name	Common name	Domains absent ^a	Furin site sequence ^b	GenBank accession number
247	35019	Thamnophis sirtalis	Common garter snake	T3, T6a	RIRR	BK010187
248	8238	Thunnus orientalis	Pacific bluefin tuna	Long propeptide, T6a	RQRR	BK010188
249	94827	Tinamus guttatus	White-throated tinamou	T3, T6a	RLHKR	BK010189
250	127582	Trichechus manatus latirostris	Caribbean manatee	T3, T6	RLRR	BK010190
251	246437	Tupaia chinensis	Chinese tree shrew	T3, T6a, T6	RQRR	BK010191
252	9739	Tursiops truncatus	Bottle-nosed dolphin	Long propeptide	RLRR	BK010192
253	56313	Tyto alba	Barn owl	T3, T6a	RLQKR	BK010193
254	9644	Ursus arctos	brown bear	T6	RVRR	BK010194
255	29073	Ursus maritimus	Polar bear	T6	RVRR	c
256	30538	Vicugna pacos	Alpaca	T6	RLWRR	BK010195
257	31156	Vipera berus berus	common viper	T3, T6a	RVRR	BK010196
258	196253	Xenopeltis unicolor	sunbeam snake	T3, T6a	RIRR	BK010197
259	8364	Xenopus (Silurana) tropicalis	Western clawed frog	None	RISKR	BK010198
260	8355	Xenopus laevis	African clawed frog	None	RIKKR	BK010199
261	32473	Xiphophorus couchianus	Monterrey platyfish	Long propeptide, T6a	RLRR	BK010200
262	8084	Xiphophorus hellerii	green swordtail	Long propeptide, T6a	RLRH	BK010201
263	8083	Xiphophorus maculatus	Southern platyfish	Long propeptide, T6a	RLRR	XM_014471723.1
264	44394	Zonotrichia albicollis	White throated sparrow	T3, T6a, T6	RLHKR	XM_014272103.1

Species for which plasma samples were assayed are indicated in green shading.

^aAncestral ADAMTS13 included these domains: a long propeptide, metalloprotease, disintegrin-like, TSP1 (T), Cys-rich, Spacer, 8 additional T domains (numbered T3, T4, T5, T6a, T6, T7, T8), and two CUB domains.

^bAmino acid sequences at the potential furin cleavage site in ADAMTS13 are shaded yellow if the sequence does not contain at least one RxxR or RxxKR motif.

^c*U. maritimus* ADAMTS13 cDNA sequence was not deposited because it is almost identical to *U. arctos*.

Supplemental Table 2. Inhibition of Plasma ADAMTS13 by Monoclonal Antibody 3H9

Species	Activity (percent of control) with MAb 3H9		
	3.2 μ g	6.4 μ g	9.6 μ g
Armadillo	98		
Baboon	0		
Cattle	107	99	103
Chicken*	125	119	131
Dog*	20	7.4	
Chimpanzee	0 (2.3 μ g)		
Ferret	10.2	6.1 (8 μ g)	
Frog	104		
Goat	105	107	
Horse*	72	36	
Human	0		
Macaque	0		
Marmoset	0		
Mouse (FVB)	94		
Pig*	49	21	
Pigeon*	101	102	96
Polar bear	57	42 (8 μ g)	
Rabbit	99		
Rat*	95	97	100
Sheep*	102		

Plasma was assayed with human-VWF71 or cattle-VWF71(*) without or with the indicated amounts of MAb 3H9 in a total assay volume of 200 μ L. Cleavage rates are expressed as a percentage of the rate observed in the absence of MAb 3H9. Standard errors are <10% of the values shown.

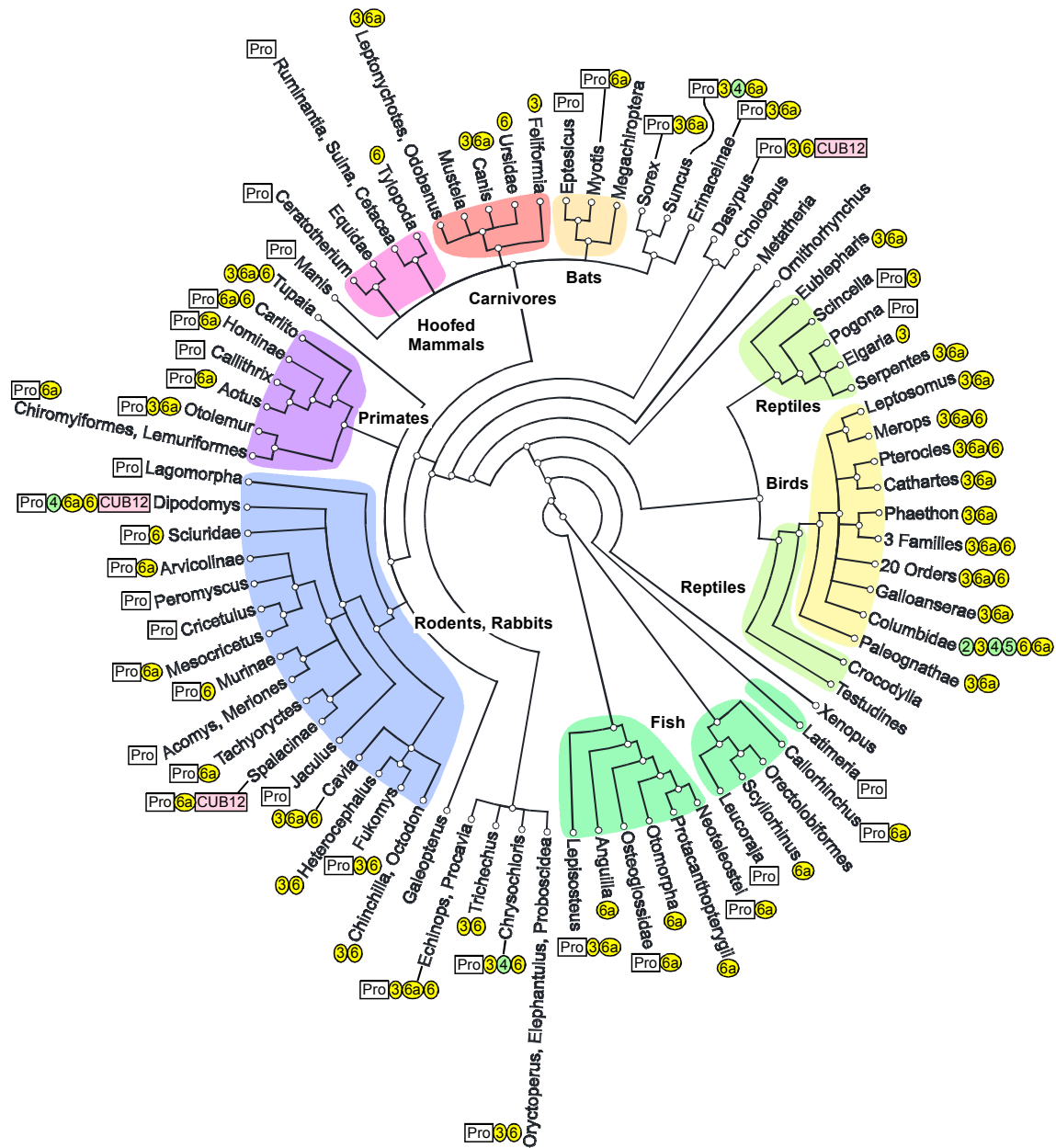


Figure S1. Phylogram of vertebrate ADAMTS13. A phylogenetic tree for the species in Table S1 was generated with Common Tree (www.ncbi.nlm.nih.gov/Taxonomy/CommonTree/) and Dendroscope (version 3.5.7) (Huson DH, Scornavacca C. Dendroscope 3: an interactive tool for rooted phylogenetic trees and networks. *Syst Biol.* 2012;**61**(6):1061-1067). Labels indicate selected major groups of carnivores, bats, reptiles, birds, fish, rodents, primates, and hoofed mammals. Figures at the periphery indicate domains that are absent from ADAMTS13 in the corresponding species, colored as in Figure 1, including the long propeptide (Pro, white), specific T domains (yellow or green), and CUB domains (pink).

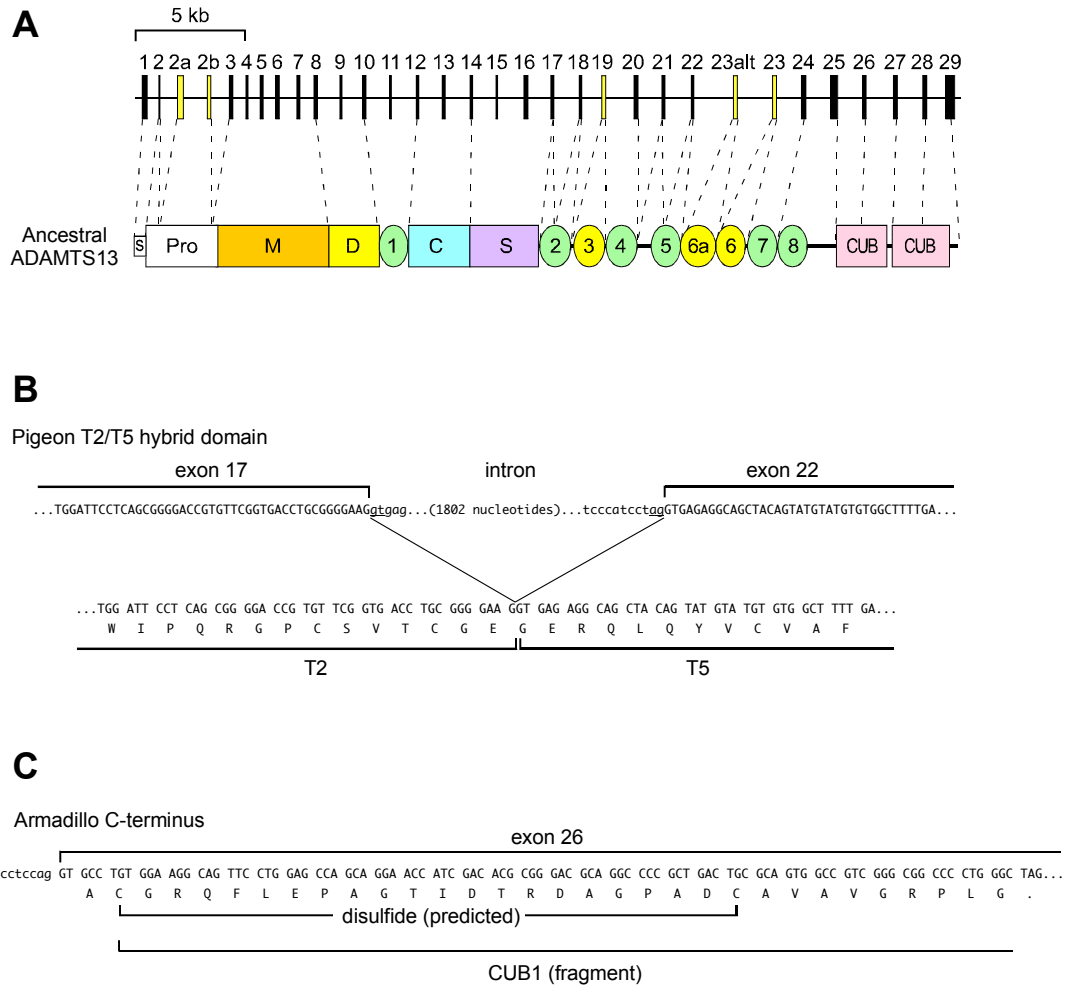


Figure S2. Vertebrate ADAMTS13 gene structure. (A) Human ADAMTS13 is encoded by 29 exons, whereas the ancestral ADAMTS13 gene had 32 exons. The 3 additional exons present in ancestral ADAMTS13 but not human ADAMTS13 are exons 2a, 2b, and 23 alternate (23alt). The correlation between exons and protein domains is indicated. Domain abbreviations and colors are the same as in **Figure 1**. The 5 exons that have been deleted frequently in extant vertebrates, and the domains they encode, are colored *yellow*. (B) The pigeon ADAMTS13 gene lacks exons 18-22. Splicing joins exon 17 and exon 22, which together encode a complete hybrid T domain composed of parts of T2 and T5. (C) Exon 26 of armadillo ADAMTS13 has a nonsense codon after Gly¹¹³⁴, truncating the first CUB domain after 31 residues.