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Title	Extracellular cleavage of collagen XVII is essential for correct cutaneous basement membrane formation
Author(s)	Nishimura, Machiko; Nishie, Wataru; Shirafuji, Yoshinori et al.
Citation	Human molecular genetics, 25(2), 328-339 https://doi.org/10.1093/hmg/ddv478
Issue Date	2016-01-15
Doc URL	https://hdl.handle.net/2115/63665
Rights	This is a pre-copyedited, author-produced PDF of an article accepted for publication in Human Molecular Genetics following peer review. The version of record Hum. Mol. Genet. (2016) 25 (2): 328-339. is available online at: http://hmg.oxfordjournals.org/content/25/2/328
Type	journal article
File Information	SupplementaryFigures_HMG-2015-TWB-00765R1Nishimura.pdf, Supplementary material

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Extracellular cleavage of collagen XVII is essential for correct cutaneous basement membrane formation

Authors:

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Supplementary material
Supplementary Figures

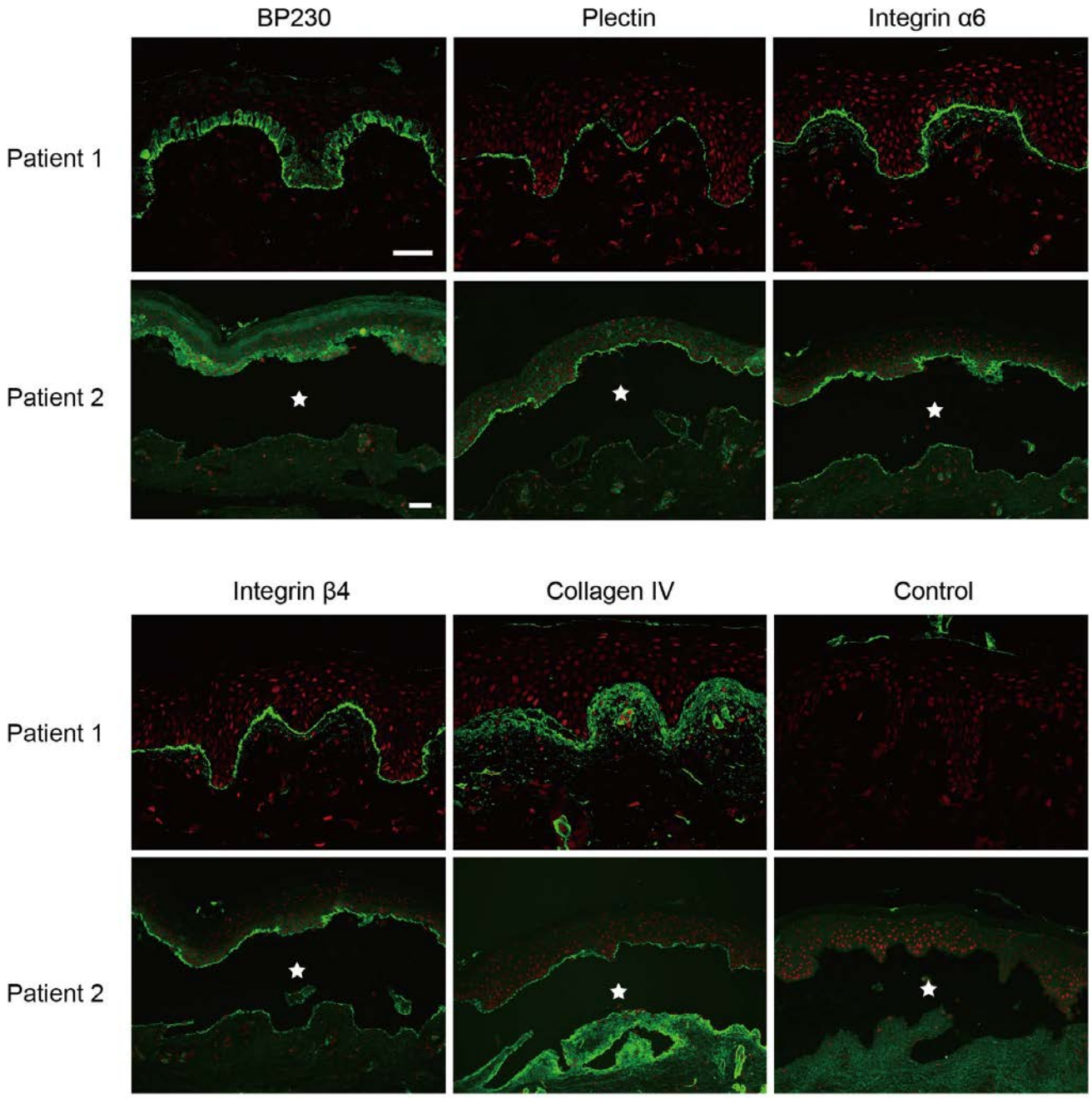


Figure S1. Immunofluorescence results for skin samples from case 1 and 2. Note that there is duplication of collagen IV, whereas expression of plectin and integrin α6 and β4 are normal. BP230 is distributed in the cytoplasm. Stars: blisters. Scale bar: 50μm.

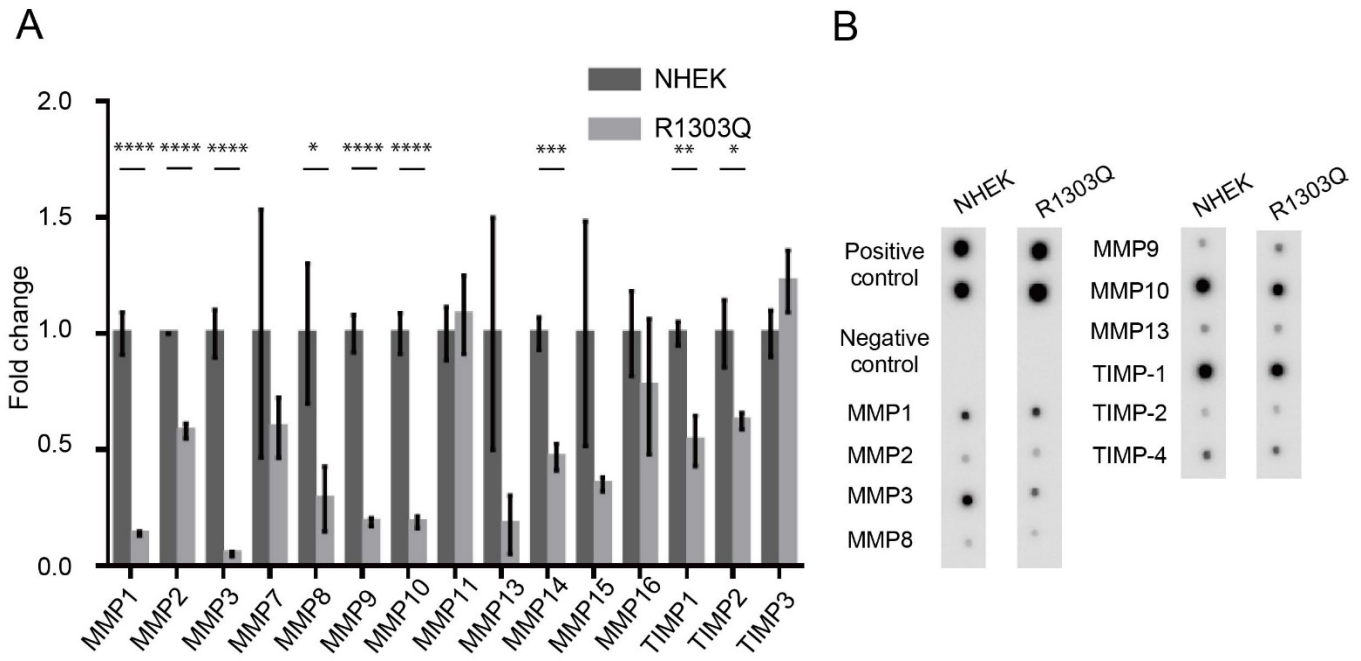


Figure S2. Expression of MMPs in NHEKs and R1303Q keratinocytes. (A) Gene expression of MMPs in R1303Q keratinocytes tends to be reduced compared with normal and R1303Q keratinocytes. **(B)** Protein expression of MMPs in the medium is not significantly different between normal and R1303Q keratinocytes.

GATATCATCGATGCTCACTGTCCCAGGCCCCCCAGGACCTCCTGGAGCCATGGGACCCCCAGGACCTCCAGGTGCCCCAG
EcoRV ClaI

GCCCTGCCGGCCCAGCTGGTCTCCCAGGACATCAAGAAGTTCTTAATTTACAAGGTCCCCCAGGCCCACCCGGCCCACGC

GGGCCACCAGGGCCTTCCATTCCAGGCCCACCAGGACCCCGAGGCCCACCAGGGGAGGGTTTGCCAGGCCCACCAGGCCC

ACCAGGATCGTTCCTGTCCAACTCAGAAACCTTCCTCTCCGGCCCCCCAGGCCCACCTGGCCCCCCAGGTCCCAAGGGAG

ACCAAGGTCCCCCAGGCCCCAGAGGACACCAAGGCGAGCAAGGCCTCCCAGGTTTCTCAACCTCAGGGTCCAGTTCTTTC

GGACTCAACCTTCAGGGACC ACCAGGCCCACCTGGCCCCCAGGGACCCAAAGGTGACAAAGGTGATCCAGGTGTTCCAGG

GGCTCTTGGCATTCCTAGTGGTCCTTCTGAAGGGGGATCATCAAGTACCATGTACGTGTCAGGCCCGCCAGGGCCCCCTG

GGCCCCCTGGGCCTCCGGGCTCTATCAGCAGCTCTGGCCAGGAGATTCAGCAGTACATCTCTGAGTACATGCAGAGTGAC

AGTATTAGATCTTACCTATCCGGAGTTCAGGGTCCCCCAGGCCCACCTGGTCCCCCAGGACCTGTCACCACCATCACAGG

CGAGACTTTCGACTACTCAGAGCTGGCAAGCCACGTTGTGAGCTACTTACGGACTTCGGGGTACGGTGTCAGCTTGTTCT

CGTCCTCCATCTCTTCTGAAGACATTCTGGCTGTGCTGCAGCGGGATGACGTGCGTCAGTACCTACGTCAGTACTTGATG

GGCCCTCGGGGTCCGCCAGG GCCACCAGGAGCCAGTGGAGATGGGTCCCTCCTGTCTTTGGACTATGCAGAGCTGAGTAG

TCGCATTCTCAGCTACATGTCGAGTTCTGGGATCAGCATTGGGCTTCCTGGTCCCCCGGGGCCCCCTGGCTTGCCGGGAA

CCTCCTATGAGGAGCTCCTCTCCTTGCTGCGAGGGTCTGAATTCAGAGGCATCGTTGGACCCCCAGGTCCCCCGGGTCCA

CCAGGGATCCCAGGCAATGT GTGGTCCAGCATCAGCGTGGAGGACCTCTCGTCTTACTTACATACTGCCGGCTTGTCATT

CATCCCAGGCCCTCCAGGACCTCCTGGTCCCCCAGGGCCTCGAGGGCCCCCGGGTGTCTCAGGAGCCCTGGCAACCTATG

CAGCTGAAAACAGCGACAGCTTCCGGAGCGAGCTGATCAGCTACCTCACAAGTCCTGATGTGCGCAGCTTCATTGTTGGC

CCCCCAGGCCCTCCTGGGCCGCAGGGACCCCCTGGGGACAGCCGCCTCCTGTCCACGGATGCCTCCCACAGTCGGGGTAG

CAGCTCCTCCTCACACAGCTCATCTGTCAGGCAGGGCAGCTCCTACAGCTCTTCCATGAGCACAGGAGGAGGTGGTGCAG
c.3908 G>A

GCTCCCTGGGTGCAGGCGGTGCCTTTGGTGAAGCTGCAGGAGACAGGGGTCCCTATGGCACTGACATCGGCCCAGGCGGA

GGCTATGGGGCAGCAGCAGA AGGCGGCATGTATGCTGGCAATGGCGGACTATTGGGAGCTGACTTTGCTGGAGATCTGGA

TTACAATGAGCTGGCTGTGAGGGTGTCAGAGAGCATGCAGCGTCAGGGCCTACTGCAAGGGATGGCCTACACTGTCCAGG

GCCCACCAGGCCAGCCTGGGCCACAGGGGCCACCCGGCATCAGCAAGGTCTTCTCTGCCTACAGCAACGTGACTGCGGAC

CTCATGGACTTCTTCCAAAC TTATGGAGCCATTCAAGGACCCCCTGGGCAAAAAGGAGAGATGGGCACTCCAGGACCCAA

AGGTGACAGGGGCCCTGCTGGGCCACCAGGTCATCCTGGGCCACCTGGCCCTCGAGGACACAAGGGAGAAAAAGGAGACA

AAGGTGACCAAGTCTATGCTGGGCGGAGAAGGAGAAGAAGTATTGCTGTCAAGCCGTGAGCGGCCGCAAGCTT
Stop NotI HindIII

Figure S3. The base sequence of a 2073-bp synthesized DNA fragment with c.3908 G>A mutation in C-terminal region of COL17. The ClaI restriction site is double-underlined, and the stop codon is highlighted in gray. EcoRV and HindIII restriction sites (underlined) are within the multiple cloning sites of pUC57.

	Symbol	Fold Change	p value
1	ADAMTS1	0.5986	0.018401
2	ADAMTS13	0.7665	0.05159
3	ADAMTS8	0.3383	0.039734
4	CD44	0.7133	0.002242
5	CDH1	0.7739	0.008541
6	CLEC3B	0.8375	0.526353
7	CNTN1	7.5731	0.00001
8	COL11A1	18.8	0.115165
9	COL12A1	0.5045	0.042252
10	COL14A1	0.3612	0.013834
11	COL15A1	0.2579	0.05926
12	COL16A1	0.6529	0.623838
13	COL1A1	0.2158	0.000005
14	COL4A2	1.6021	0.001767
15	COL5A1	7.4545	0.000502
16	COL6A1	0.529	0.000017
17	COL6A2	0.1153	0.000202
18	COL7A1	0.76	0.024326
19	COL8A1	0.3671	0.000221
20	CTGF	0.3339	0.000065
21	CTNNA1	0.6797	0.000225
22	CTNNB1	1.1089	0.423239
23	CTNND1	0.9537	0.808889
24	CTNND2	1.9773	0.000013
25	ECM1	0.7478	0.007727
26	FN1	0.1232	0
27	HAS1	0.1614	0.000049
28	ICAM1	0.1	0.000002
29	ITGA1	1.0245	0.868199
30	ITGA2	0.6566	0.001607
31	ITGA3	0.9668	0.309538
32	ITGA4	0.9381	0.20631
33	ITGA5	0.9843	0.790541
34	ITGA6	0.7703	0.045357
35	ITGA7	0.5767	0.00372
36	ITGA8	0.6199	0.881667
37	ITGAL	0.3572	0.090007
38	ITGAM	0.2866	0.0095
39	ITGAV	0.5253	0.000027
40	ITGB1	0.8288	0.095152
41	ITGB2	0.2022	0.000413
42	ITGB3	0.2619	0.01014
43	ITGB4	0.8343	0.081267
44	ITGB5	1.7092	0.001087
45	KAL1	1.3146	0.001019
46	LAMA1	1.4527	0.077736
47	LAMA2	0.8331	0.041259
48	LAMA3	0.7167	0.01648
49	LAMB1	0.9575	0.44833
50	LAMB3	0.5609	0.000239
51	LAMC1	0.8146	0.00191
52	MMP1	0.14	0.00008
53	MMP10	0.1879	0.000103
54	MMP11	1.0761	0.530506
55	MMP12	1.6101	0.399273
56	MMP13	0.1654	0.050569
57	MMP14	0.468	0.000496
58	MMP15	0.3768	0.0809
59	MMP16	0.7413	0.313999
60	MMP2	0.5798	0.000021
61	MMP3	0.0521	0.000092
62	MMP7	0.646	0.269731
63	MMP8	0.2742	0.020686
64	MMP9	0.1903	0.000073
65	NCAM1	0.4165	0.029736
66	PECAM1	0.5299	0.039009
67	SELE	0.339	0.074632
68	SELL	0.4192	0.014451
69	SELP	0.2969	0.08379
70	SGCE	1.769	0.002164
71	SPARC	0.6233	0.00022
72	SPG7	0.9024	0.109212
73	SPP1	2.8326	0.000001
74	TGFBI	1.0105	0.895663
75	THBS1	0.3695	0.000156
76	THBS2	2.2957	0.000159
77	THBS3	1.0893	0.61495
78	TIMP1	0.5321	0.002522
79	TIMP2	0.6274	0.011939
80	TIMP3	1.2235	0.079892
81	TNC	0.2883	0.000077
82	VCAM1	0.1818	0.007588
83	VCAN	3.0123	0.000032
84	VTN	0.9751	0.965742
85	ACTB	0.7888	0.020575
86	B2M	0.7982	0.116234
87	GAPDH	0.7048	0.01443
88	HPRT1	1	0
89	RPLP0	0.6902	0.014717
90	HGDC	0.4602	0.201216
91	RTC	1.4254	0.487673
92	RTC	1.5894	0.398412
93	RTC	1.5162	0.388513
94	PPC	0.5579	0.116966
95	PPC	0.5579	0.118136
96	PPC	0.527	0.136876

Supplementary Table

Table S1. Data of RT2 Profiler PCR array-analyzed ECM molecules. Comparison of genes

up-regulated (1.5> -fold, red) and down-regulated (<0.5 –fold, blue) in NHEKs and R1303Q keratinocytes.