

Antistoffen Huid

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SKML-HIM 31 januari 2019

Nabespreking rondzendingen 2016-2018

	A	B	C	D	E	F	G
1		var	expertwaarde	IIF score	afw	opm	overall
2	2016 1A	BP	IIF positief	7/7			7/7
3	2016 1B	geen	IIF negatief	5/7	1x BMZ pos, 1x BMZ neg		7/7
4	2016 1C	PF	IIF positief	5/7	2x IIF neg	1 alsnog pos via ELISA	6/7
5	2016 1D	BP	IIF negatief	6/7	1x pos	via mozaiek	6/7
6	2016 1E	geen	IIF negatief	7/7			7/7
7	2016 1F	PV	IIF positief	7/7		discrepantie Dsg1 ELISA vs mozaiek	7/7
8							
9	2017 1A	PF	IIF positief	7/8	1x dub		7/8
10	2017 1B	BP	IIF positief	3/8	4x neg, 1x dub	1 alsnog pos via ELISA, uit ronde gehaald	
11	2017 1C	geen	IIF negatief	8/8			8/8
12	2017 1D	BP	IIF positief	8/8			8/8
13	2017 1E	PF	IIF positief	7/8	1x dub		7/8
14	2017 1F	PF	IIF positief	6/8	2x neg		6/8
15							
16	2018 1A	PV	IIF positief	5/7	1x neg, 1x dub		5/7
17	2018 1B	PF	IIF positief	6/7	1x neg (expert)		6/7
18	2018 1C	PF	IIF positief	7/7			7/7
19	2018 1D	BP	IIF negatief	5/7	1x dub, 1x neg	theoretisch positief, uit ronde gehaald, 1x ELISA pos	
20	2018 1E	BP	IIF positief	7/7			7/7
21	2018 1F	geen	IIF negatief	6/7	1x dub BMZ		6/7

Diagnostics of autoimmune blistering diseases

When your body attacks yourself

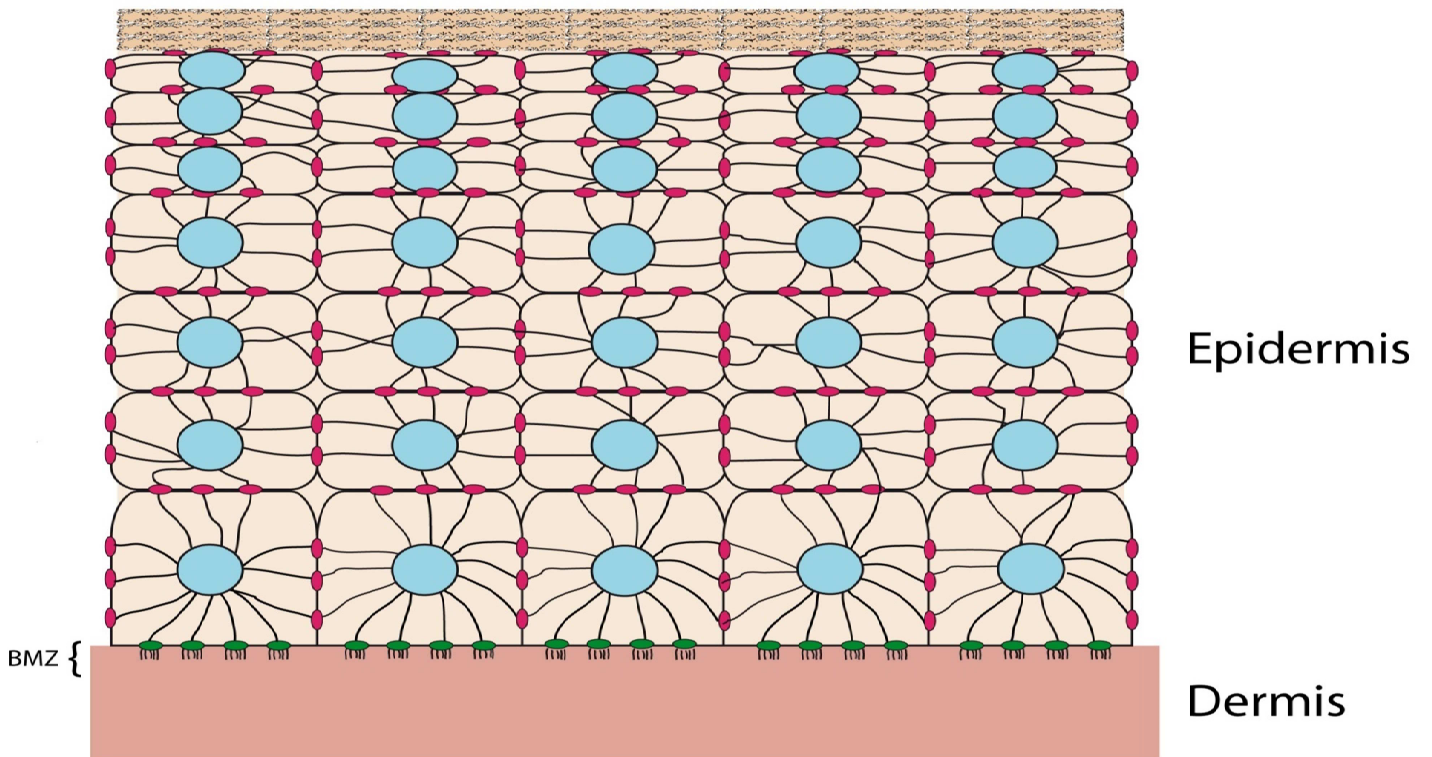


Disclosure belangen spreker

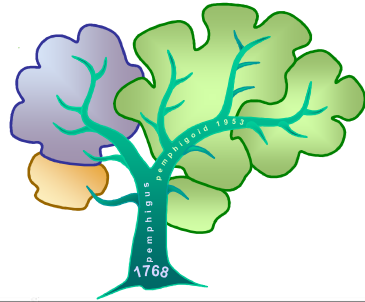
(potentiele) belangenverstrengeling	Geen
Voor deze bijeenkomst mogelijk relevante relaties met bedrijven	Bedrijfsnamen
<ul style="list-style-type: none">• Sponsoring of onderzoeksgeld• Honorarium of andere (financiële) vergoeding• Aandeelhouder• Andere relatie, namelijk:	<ul style="list-style-type: none">••••

Keratinocyte in vitro binding assay
Keratinocyte footprint assay

Diagnostics of autoimmune blistering diseases
based on cell behaviour



- Desmosoom
- Hemidesmosoom



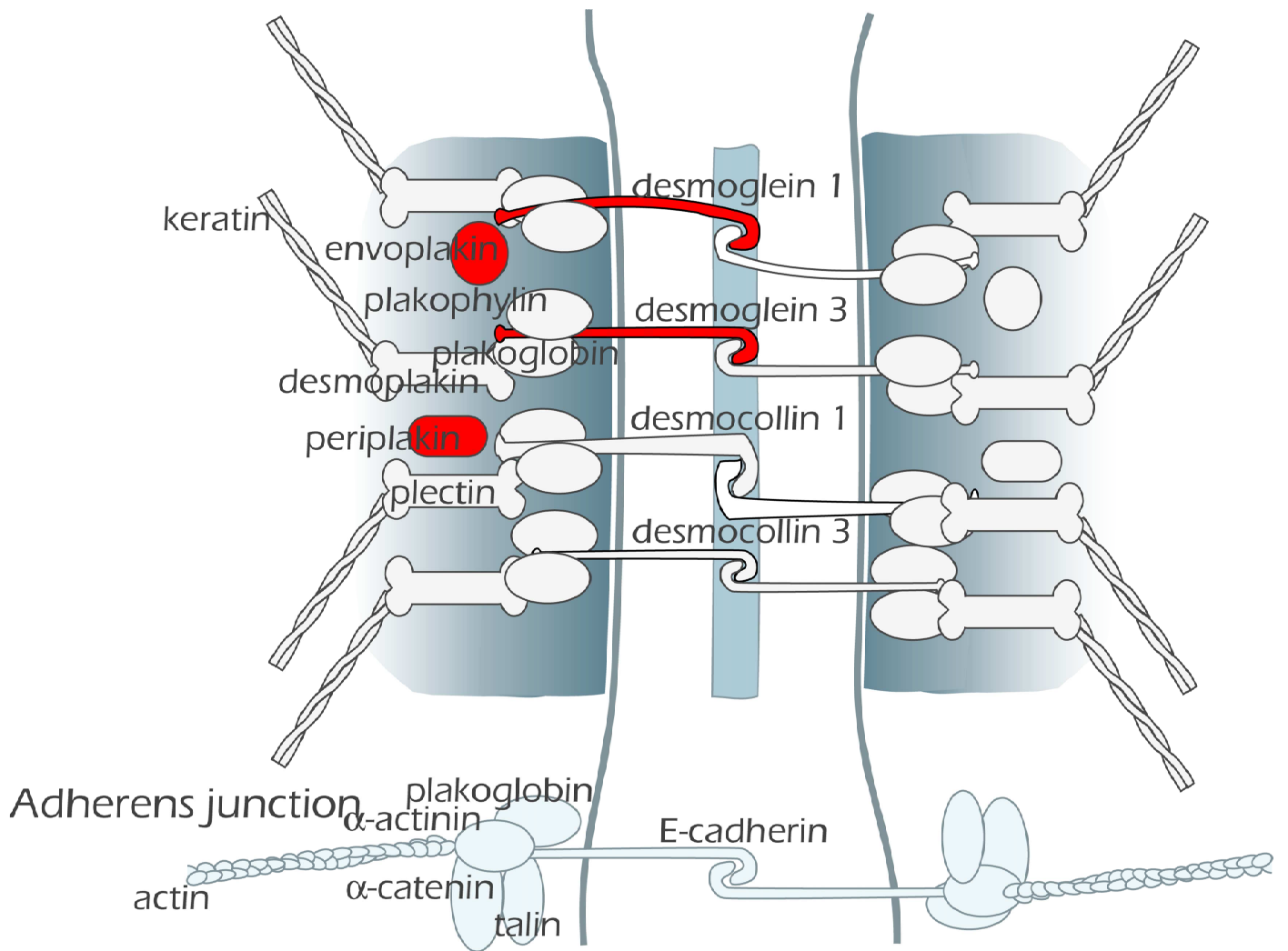
		BP230	BP180	LAD-1	plectine	lam-332	lam-γ1	col7	
	pemfigoïd spectrum								
90%	bulleus pemfigoïd	+	+	+	(+)				
	pemfigoïd gestationis		+	+					
	slijmvliespemfigoïd		+	+					
	lichen planus pemphigoides		+						
	oculair pemfigoïd		+						
	lineaire IgA dermatose		+	+					
10%	epidermolysis bullosa acquisita							+	
	anti-laminine 332 pemfigoïd					+			
	anti-p200 pemfigoïd						+		
	IgA epidermolysis bullosa aquisita							+	
	pemphigus spectrum	dsg1	dsg3	dsc1	dsc3	dp	ep	pp	a2ml1
	pemphigus foliaceus	+							
	mucosale pemphigus vulgaris		+		(+)	(+)			
	mucocutane pemphigus vulgaris	+	+		(+)				
	pemphigus erythematous	+							
	pemphigus herpetiformis	+	+	(+)	(+)				
	pemphigus vegetans	+	+	(+)					
	paraneoplastische pemphigus	+	+			+	+	+	+
	subcorneale pustuleuze dermatose			+					
	intra-epidermale neutrofiele IgA dermatose								

dermatitis herpetiformis

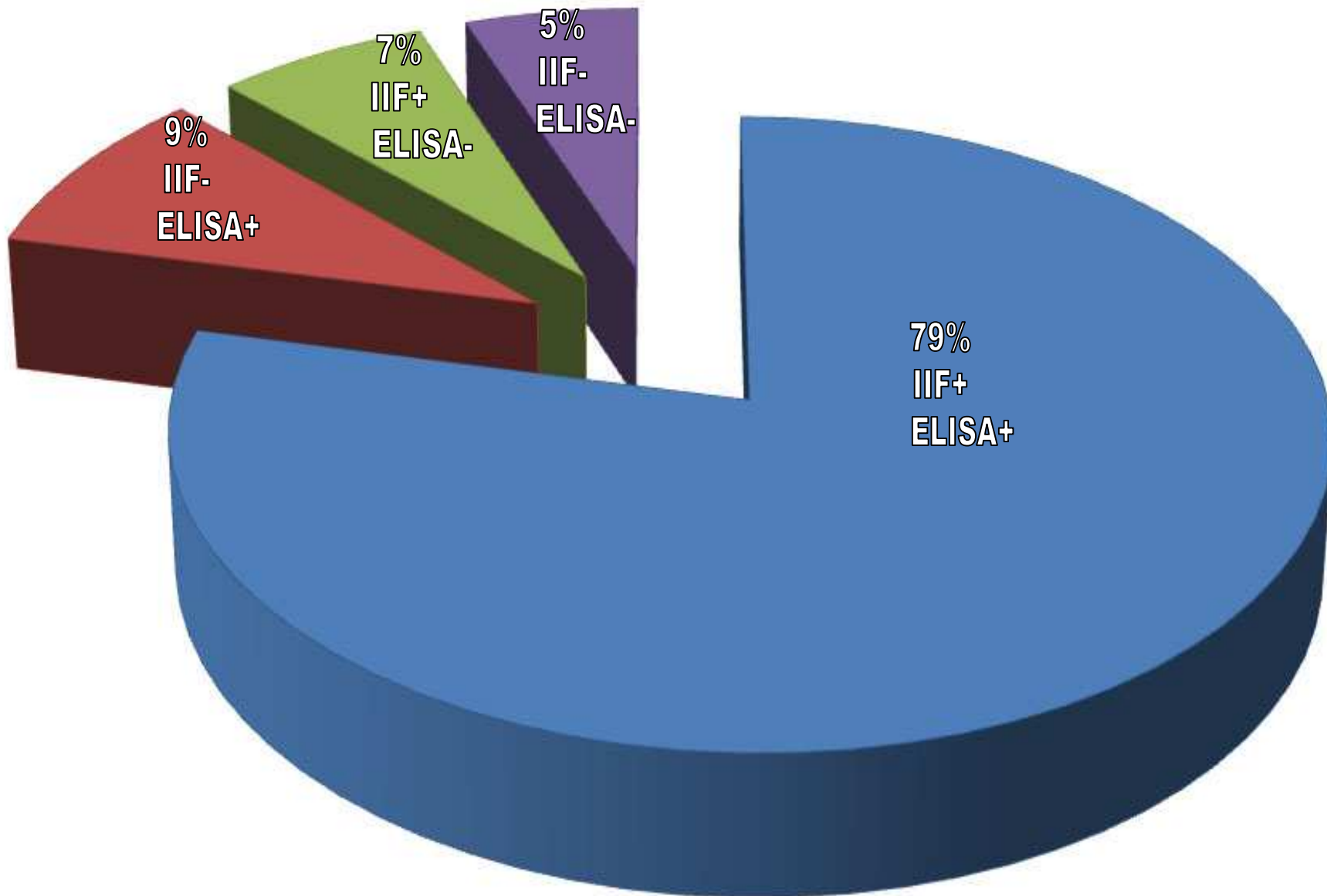
epidermale transglutaminase, eTG, TGase 3

Exotische combinaties

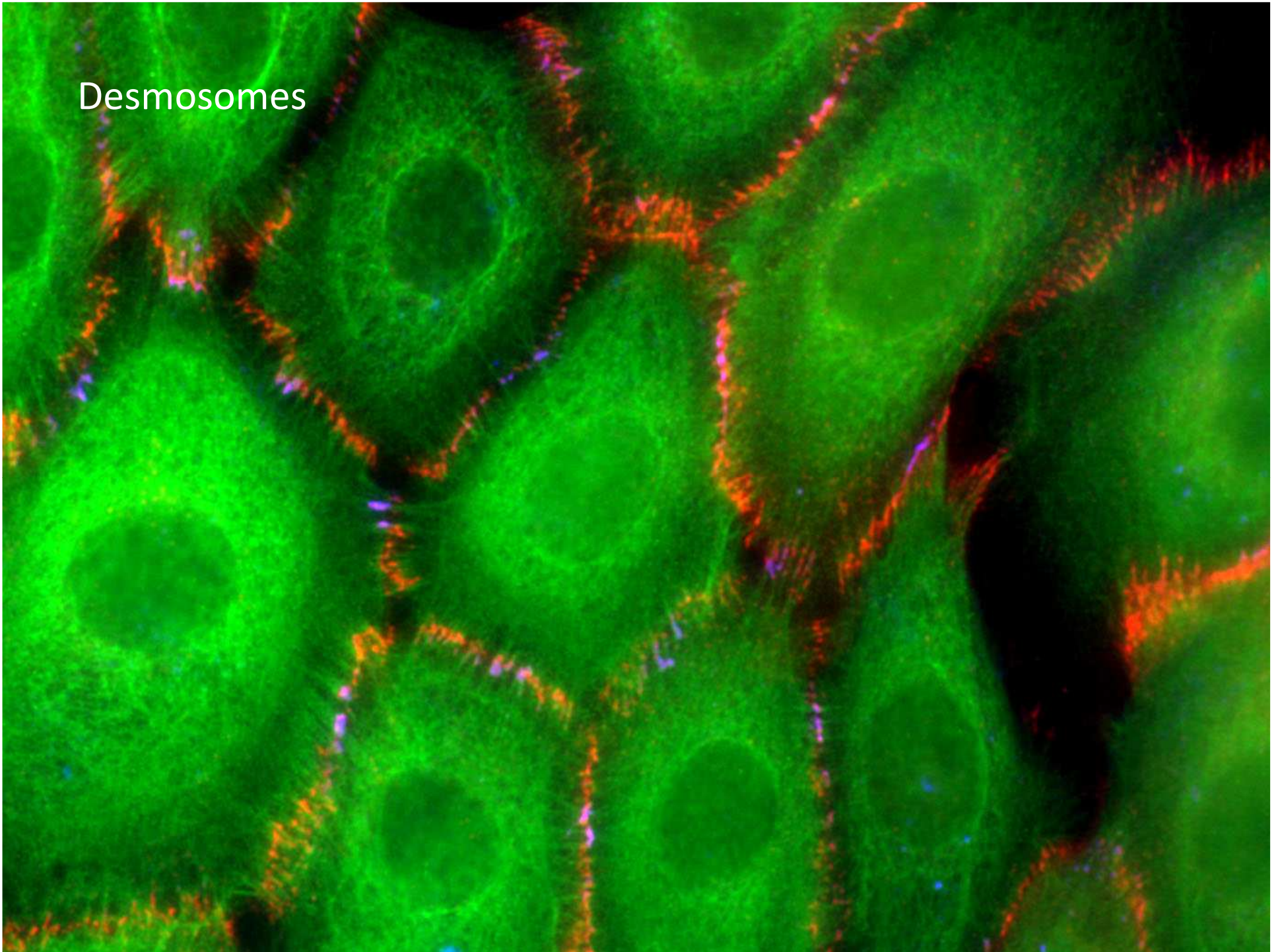
pemfigoïd/pemfigoïd pemphigus/pemfigoïd



Serologie van 234 patiënten met positieve DIF

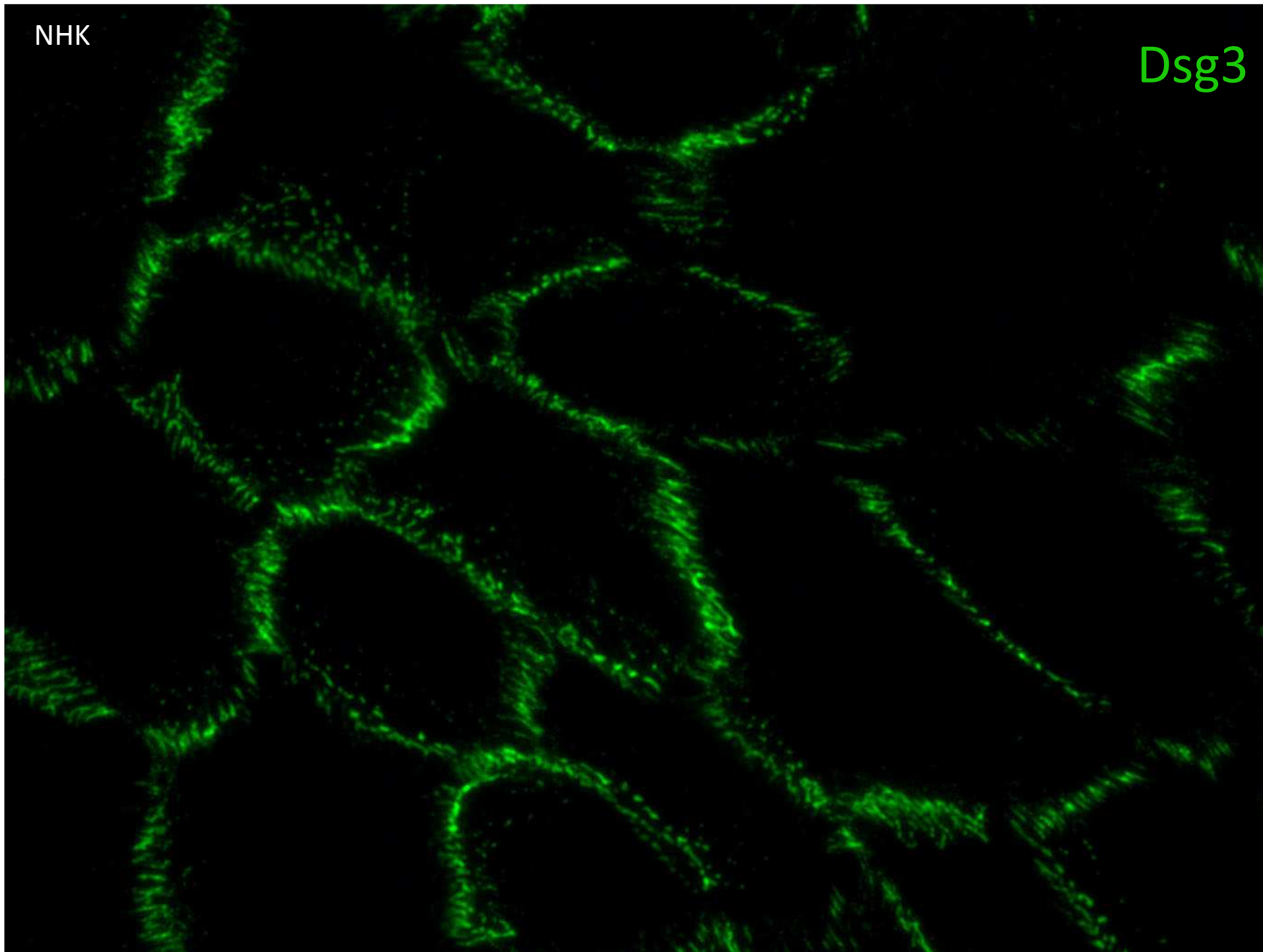


Desmosomes



NHK

Dsg3



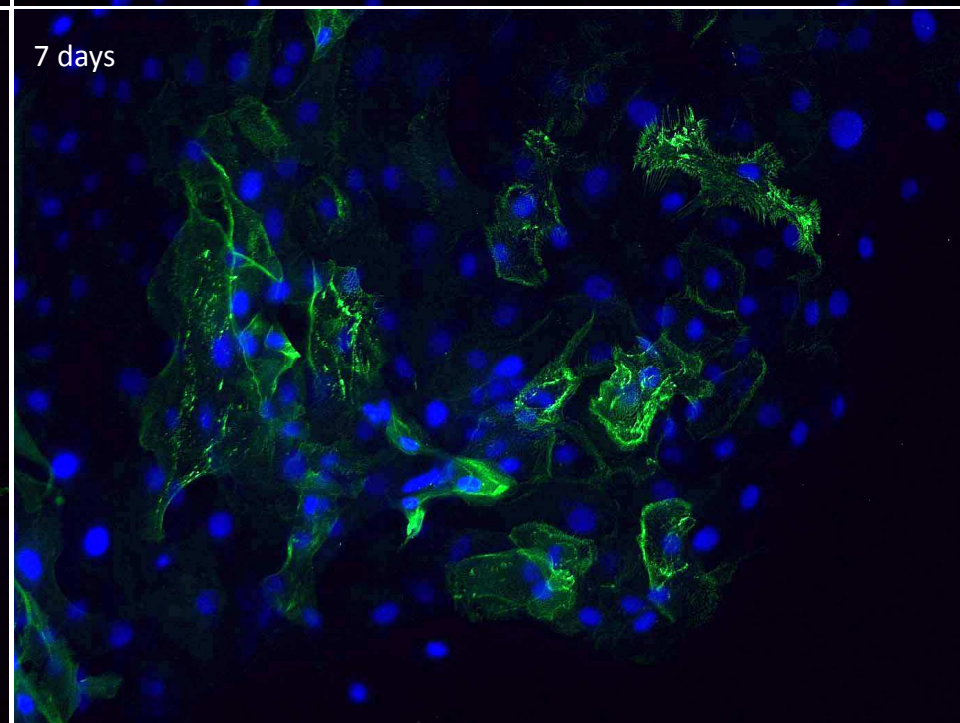
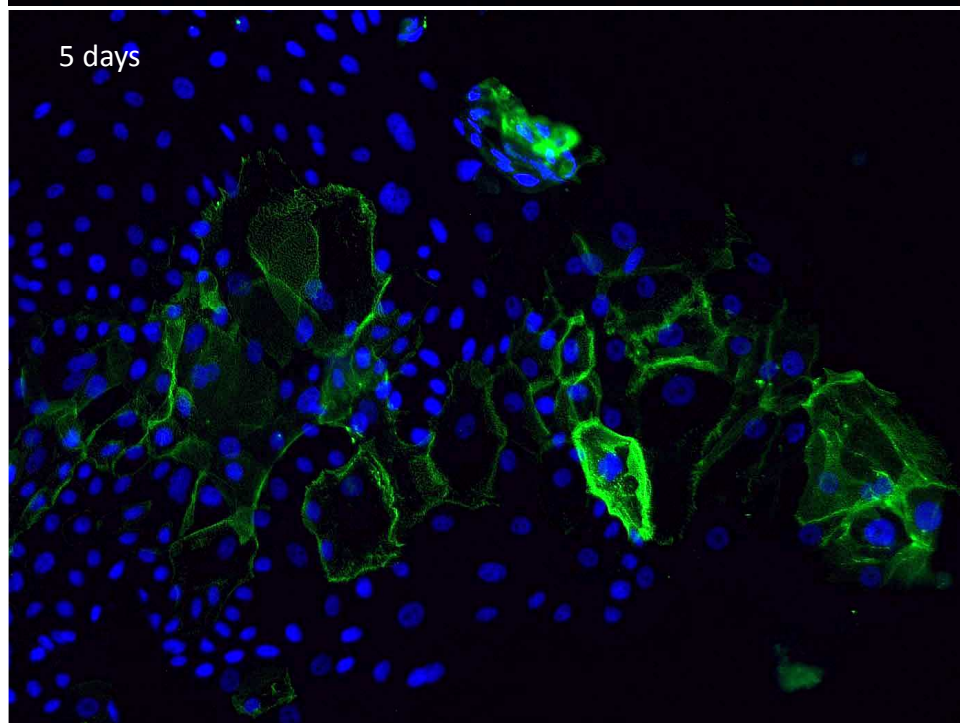
1 day after Ca

Dsg1

3 days

5 days

7 days



In vitro binding assay

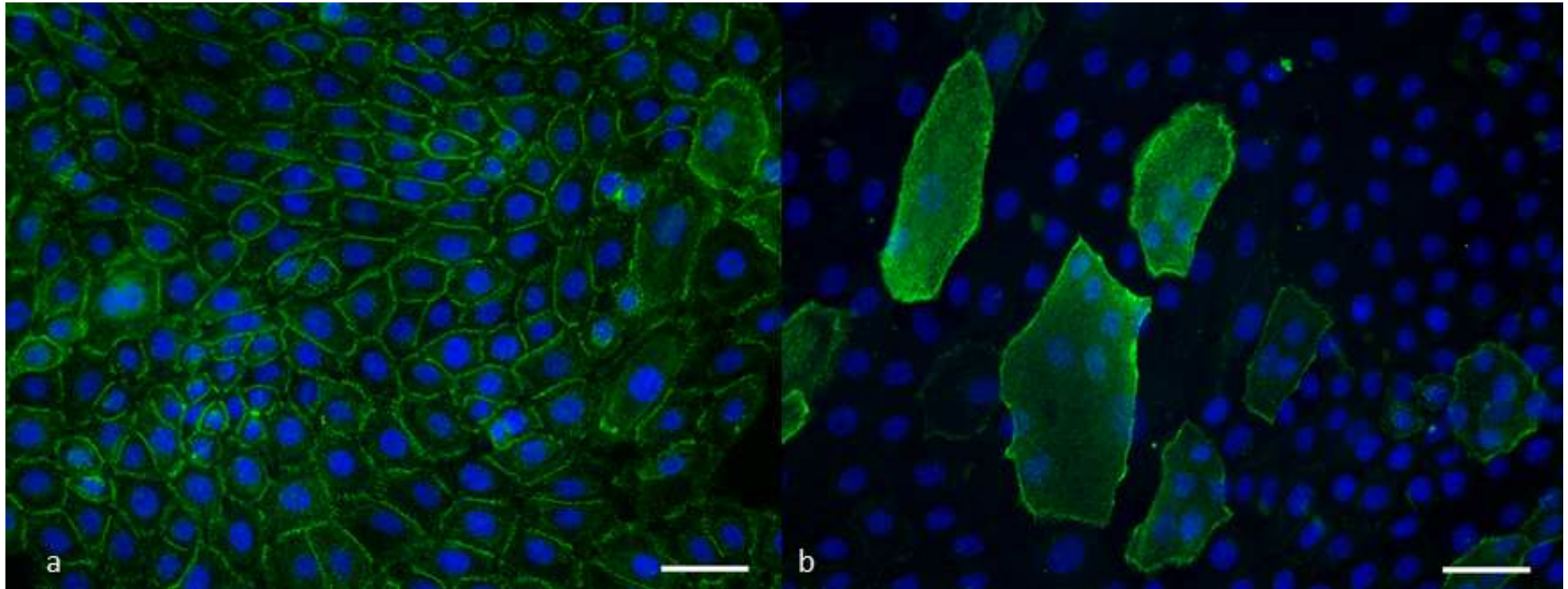


Table S1 - Comprehensive results for the DIF positive pemphigus sera. ELISA values are expressed in U/mL.

IIF-MO: indirect immunofluorescence on monkey esophagus. Positive results are in bold.

KBA: keratinocyte binding assay. nd: not determinable.

Sample	ELISA Dsg1	ELISA Dsg3	IIF-MO	KBA Dsg1	KBA Dsg3	
1	1	3	+	nd	+	10/12 positief
2	3	2	+	nd	+	
3	1	4	+	+	-	
4	2	17	+	nd	+	
5	19	13	+	nd	+	
6	9	5	+	nd	+	
7	8	14	+	nd	+	
8	3	16	+	nd	+	
9	4	2	+	-	-	
10	6	5	+	nd	+	
11	1	1	+	-	-	
12	4	1	+	nd	+	
13	96	1	-	+	-	26/26 positief
14	52	2	-	+	-	
15	>150	2	-	+	-	
16	138	15	-	+	-	
17	123	1	-	+	-	
18	124	2	-	+	-	
19	>150	5	-	+	-	
20	>150	4	-	+	-	
21	113	1	-	+	-	
22	71	1	-	+	-	
23	110	2	-	+	-	
24	72	3	-	+	-	
25	6	84	-	nd	+	
26	6	52	-	nd	+	
27	4	55	-	nd	+	
28	2	37	-	nd	+	
29	134	101	-	nd	+	
30	95	22	-	+	-	
31	115	95	-	nd	+	
32	86	26	-	+	-	
33	74	79	-	nd	+	
34	24	3	-	-	+	
35	44	15	-	+	-	
36	8	33	-	+	-	
37	2	21	-	nd	+	
38	2	37	-	nd	+	
39	17	1	-	+	-	3/5 positief
40	8	2	-	-	-	
41	1	1	-	-	-	
42	1	0	-	+	-	
43	6	2	-	nd	+	

Table S2 - Comprehensive results for the DIF negative pemphigus sera. ELISA values are expressed in U/mL. IIF-MO: indirect immunofluorescence on monkey esophagus. Positive results are in bold. KBA: keratinocyte binding assay.

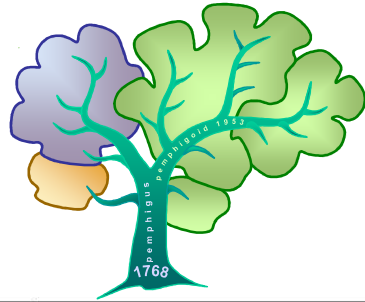
Sample	ELISA Dsg1	ELISA Dsg3	IIF-MO	KBA Dsg1	KBA Dsg3
44	1	2	+	-	-
45	3	2	+	-	-
46	4	5	+	-	-
47	0	8	+	-	-
48	3	3	+	-	-
49	16	12	+	-	-
50	1	3	+	-	-
51	1	1	+	-	-
52	1	9	+	-	-
53	4	1	+	-	-
54	5	2	+	-	-
55	11	15	+	-	-
56	3	1	+	-	-
57	5	1	+	-	-
58	1	1	+	-	-
59	2	0	+	-	-
60	2	2	+	-	-
61	4	1	+	-	-
62	2	1	+	-	-
63	8	1	+	-	-
64	5	1	+	-	-
65	1	1	+	-	-
66	5	3	+	-	-
67	2	4	+	-	-
68	1	1	+	-	-
69	1	0	+	-	-
70	2	3	+	-	-
71	1	3	+	-	-
72	43	1	-	-	-
73	57	2	-	-	-
74	61	1	-	-	-
75	25	2	-	-	-
76	55	2	-	-	-
77	30	6	-	-	-
78	36	5	-	-	-
79	38	15	-	-	-
80	32	1	-	-	-
81	37	0	-	-	-
82	27	12	-	-	-
83	22	8	-	-	-
84	36	1	-	-	-
85	36	1	-	-	-
86	26	9	-	-	-
87	39	19	-	-	-
88	70	2	-	-	-
89	42	1	-	-	-
90	2	22	-	-	-
91	4	42	-	-	-
92	1	33	-	-	-
93	4	45	-	-	-
94	2	84	-	-	-
95	14	45	-	-	-
96	12	53	-	-	-
97	1	27	-	-	-
98	11	29	-	-	-
99	3	29	-	-	-
100	129	30	-	-	-
101	33	35	-	-	-
102	43	34	-	-	-
103	63	35	-	-	-

26 sera IIF positief

32 sera ELISA positief

234 biopten positief

- Keratinocyte Footprint Assay



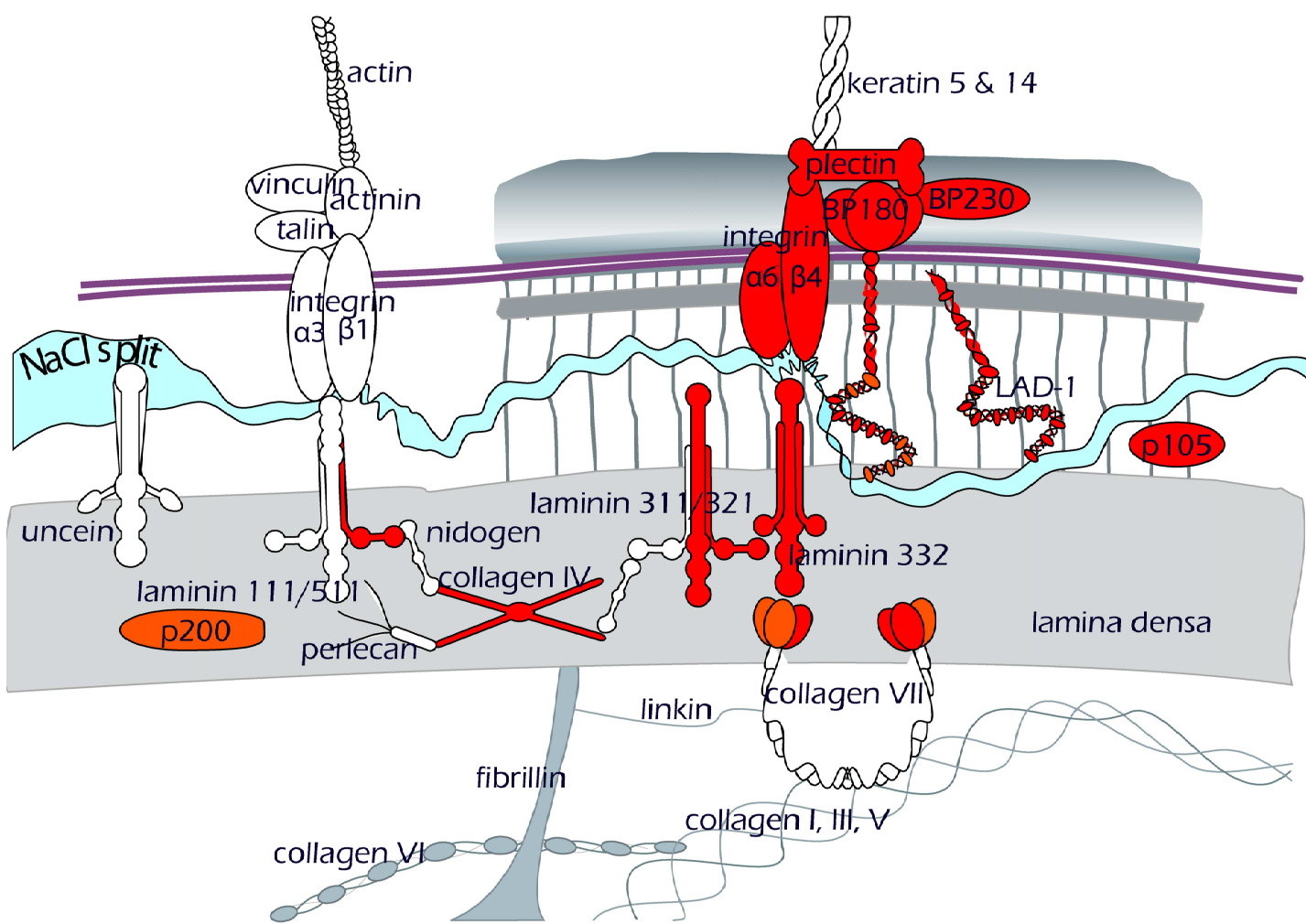
		BP230	BP180	LAD-1	plectine	lam-332	lam-γ1	col7	
	pemfigoïd spectrum								
90%	bulleus pemfigoïd	+	+	+	(+)				
	pemfigoïd gestationis		+	+					
	slijmvliespemfigoïd		+	+					
	lichen planus pemphigoides		+						
	oculair pemfigoïd		+						
	lineaire IgA dermatose		+	+					
10%	epidermolysis bullosa acquisita							+	
	anti-laminine 332 pemfigoïd					+			
	anti-p200 pemfigoïd						+		
	IgA epidermolysis bullosa aquisita							+	
	pemphigus spectrum	dsg1	dsg3	dsc1	dsc3	dp	ep	pp	a2ml1
	pemphigus foliaceus	+							
	mucosale pemphigus vulgaris		+		(+)	(+)			
	mucocutane pemphigus vulgaris	+	+		(+)				
	pemphigus erythematous	+							
	pemphigus herpetiformis	+	+	(+)	(+)				
	pemphigus vegetans	+	+	(+)					
	paraneoplastische pemphigus	+	+			+	+	+	+
	subcorneale pustuleuze dermatose			+					
	intra-epidermale neutrofiele IgA dermatose								

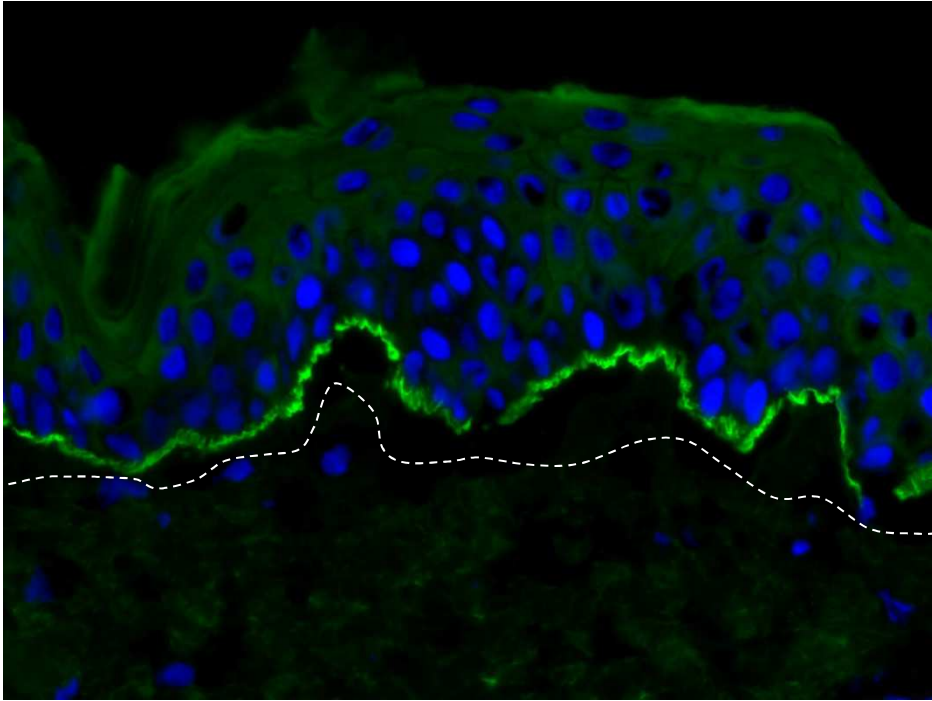
dermatitis herpetiformis

epidermale transglutaminase, eTG, TGase 3

Exotische combinaties

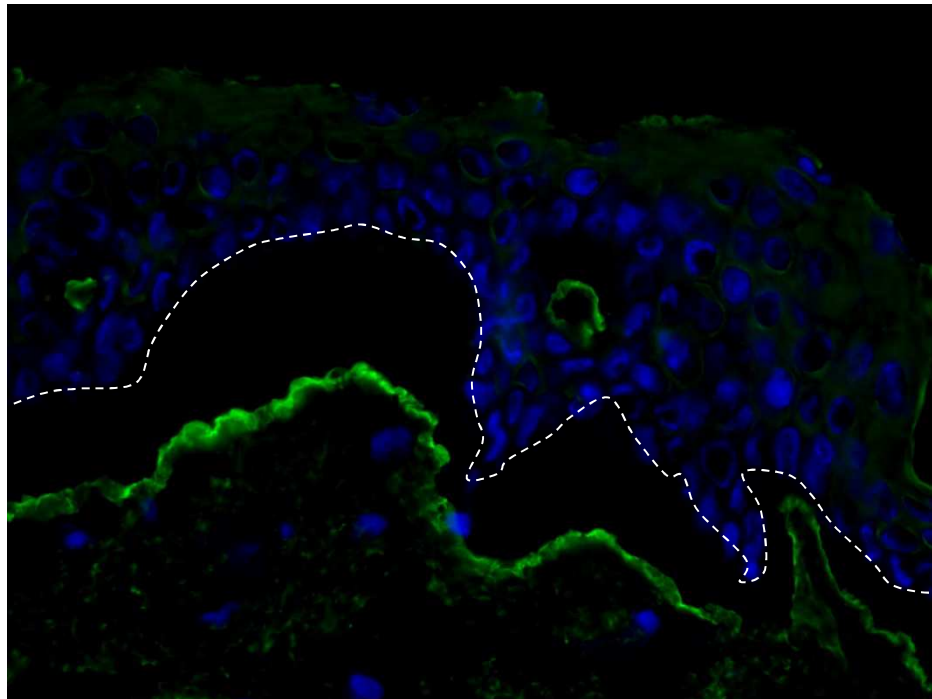
pemfigoïd/pemfigoïd pemphigus/pemfigoïd





BP180 and other
hemidesmosomal antigens

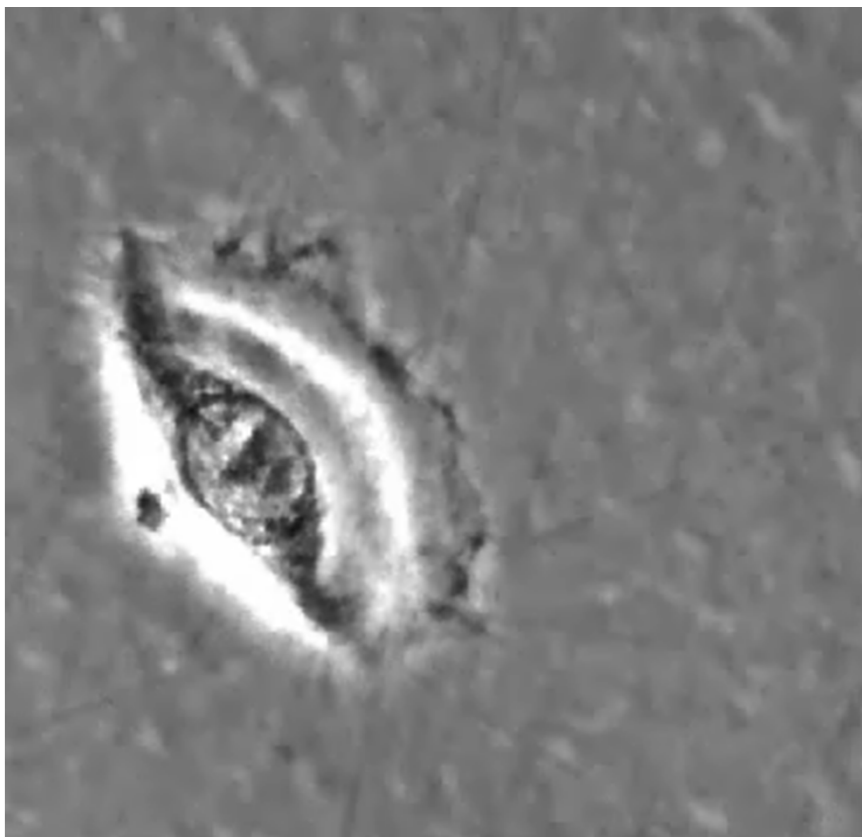
90%

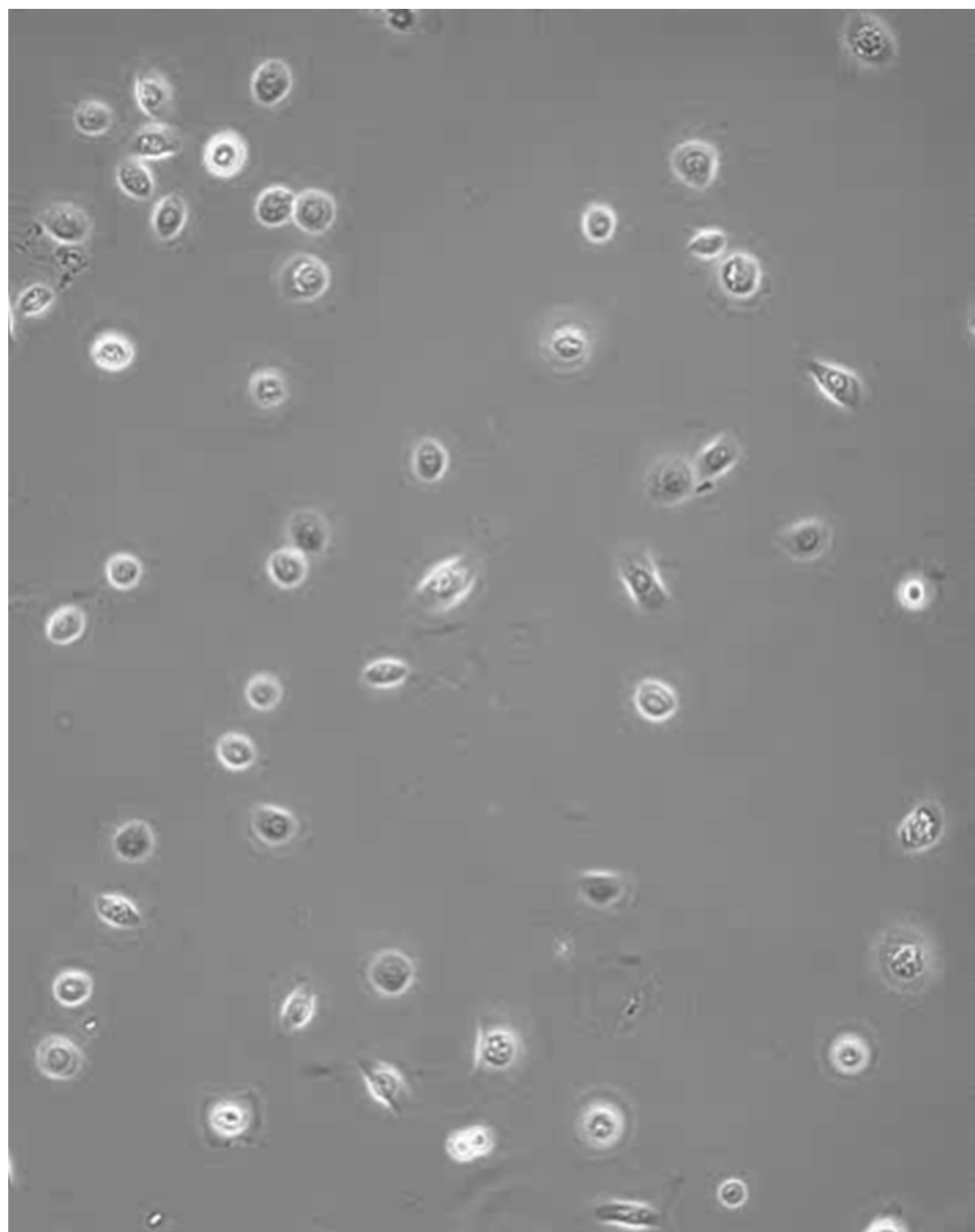


Collagen 7 6%

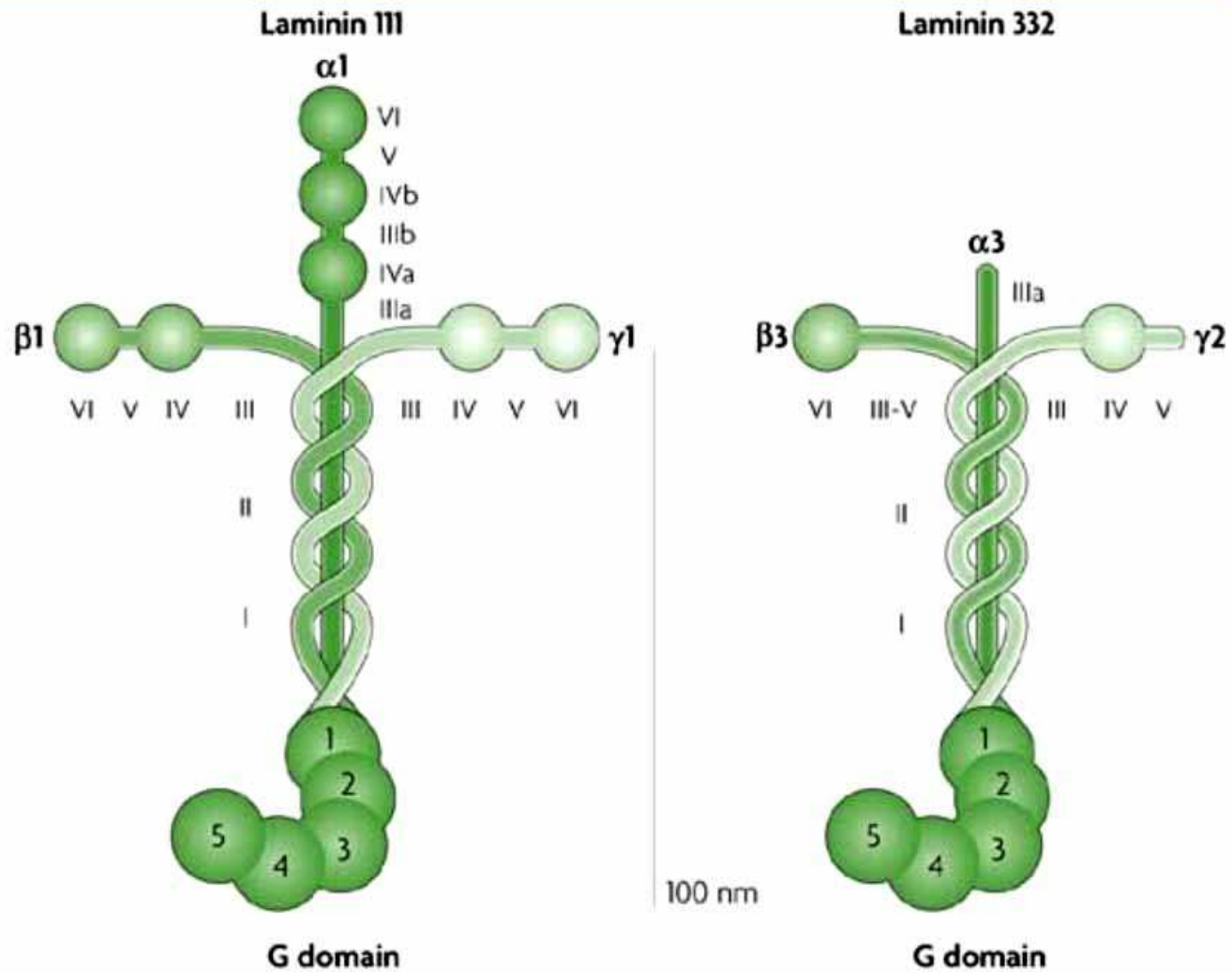
Laminin-332 2%

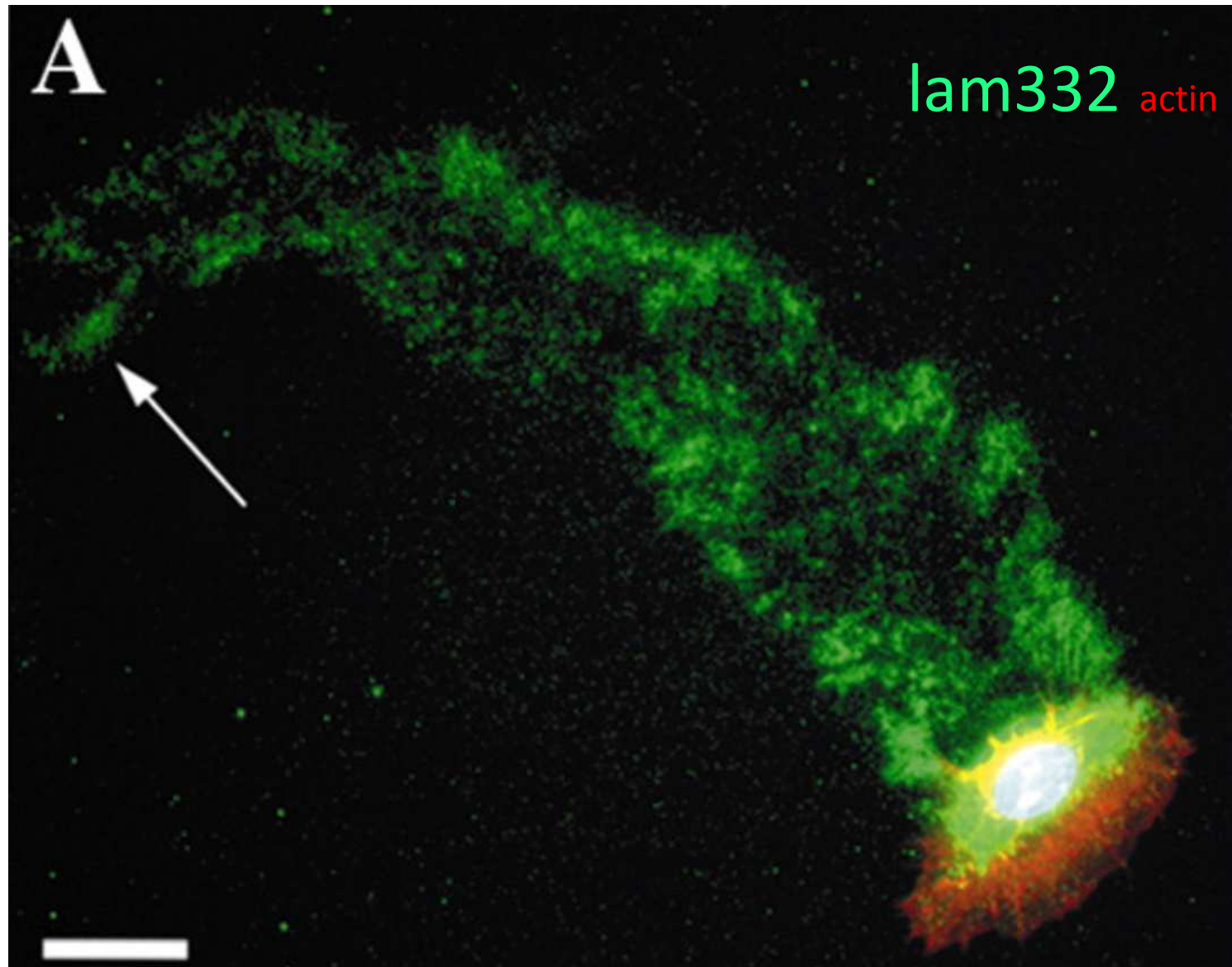
p200 2%



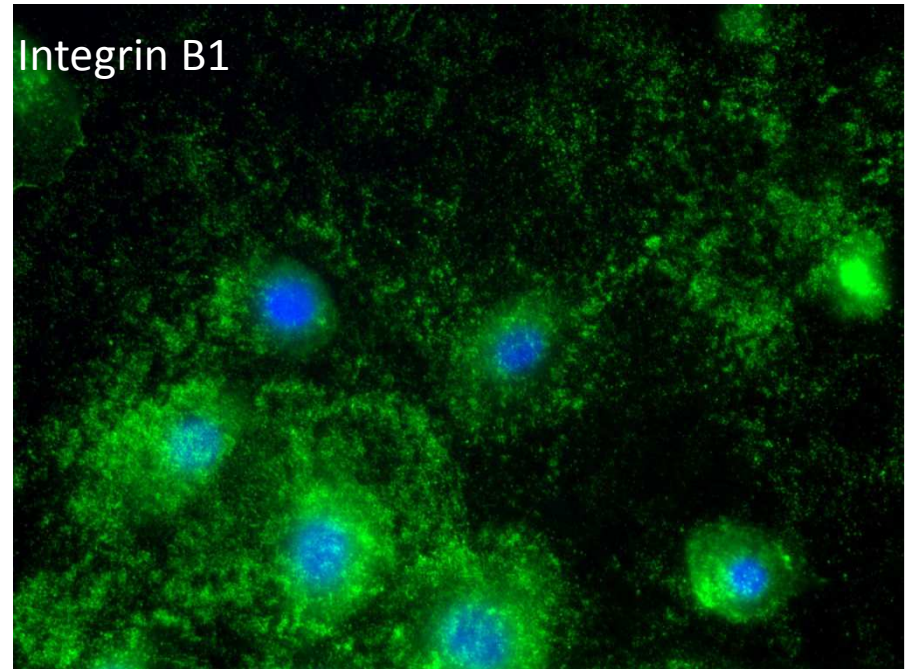
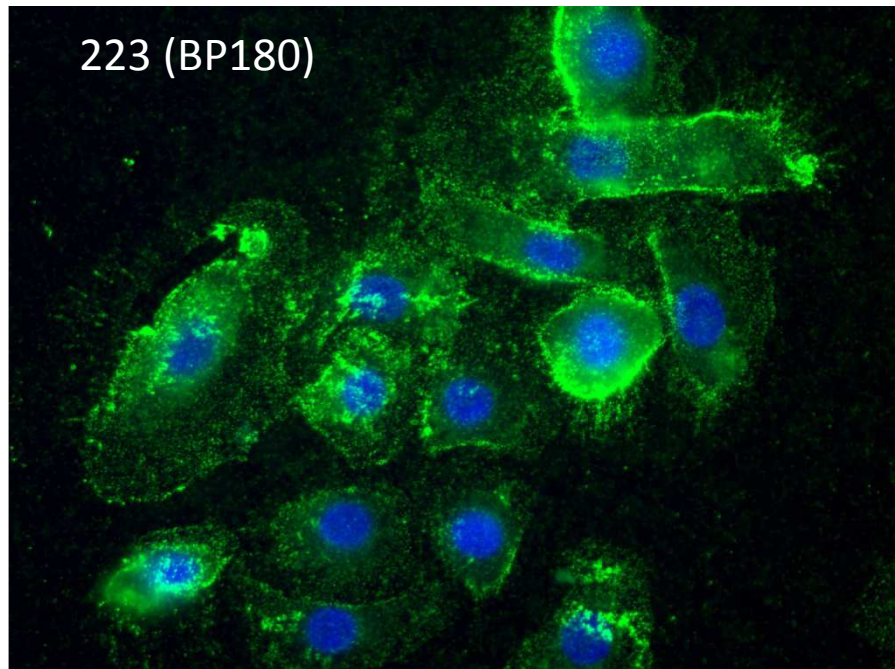
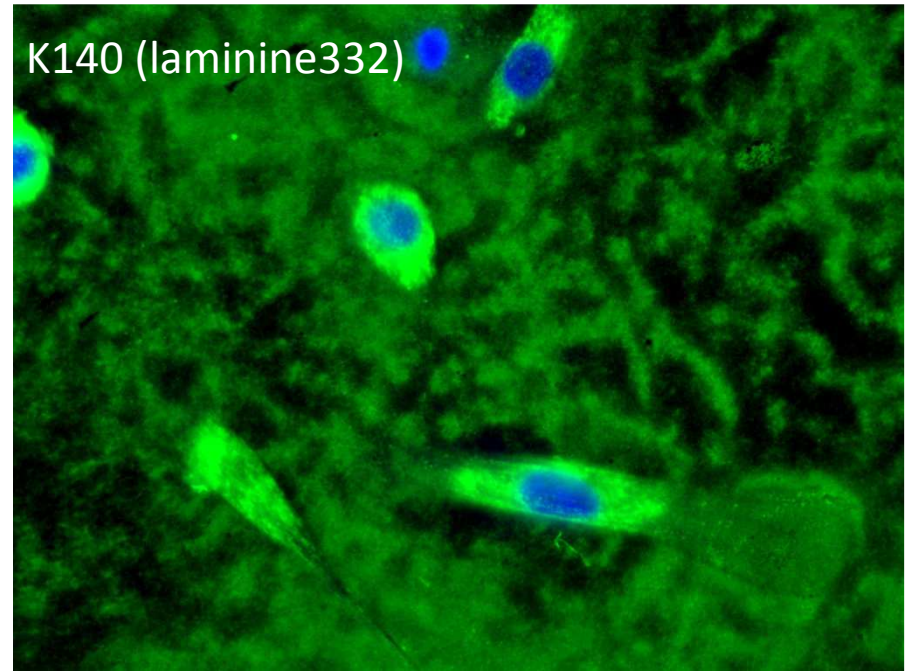
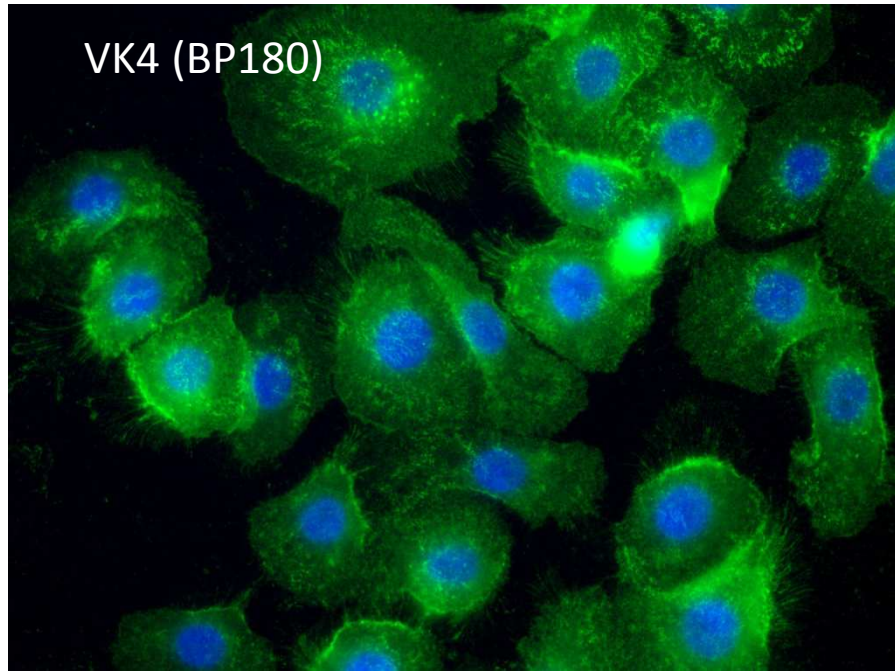


Laminin

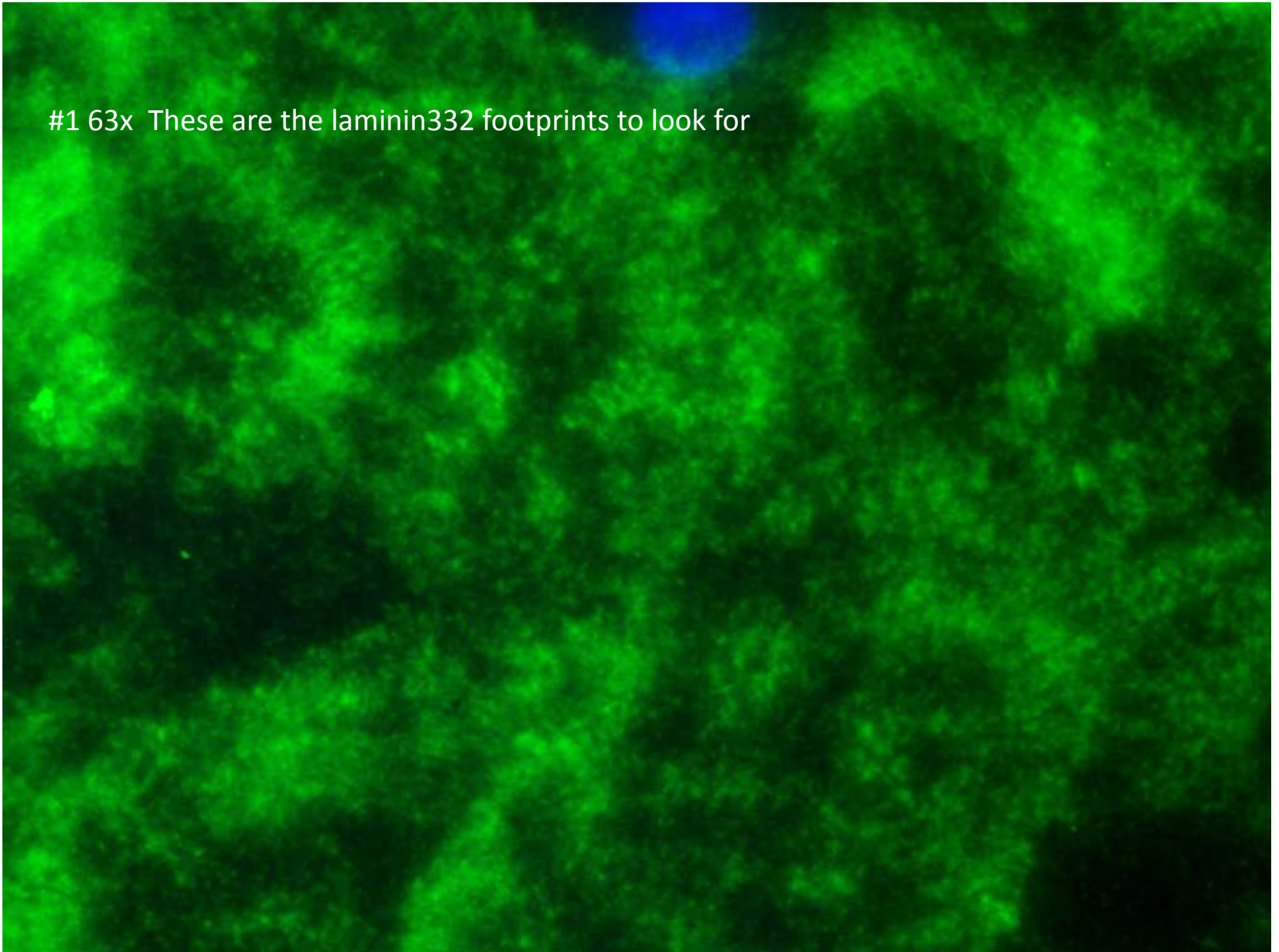




Diane E. Frank, William G. Carter
Journal of Cell Science 2004 117: 1351-1363;
doi: 10.1242/jcs.01003

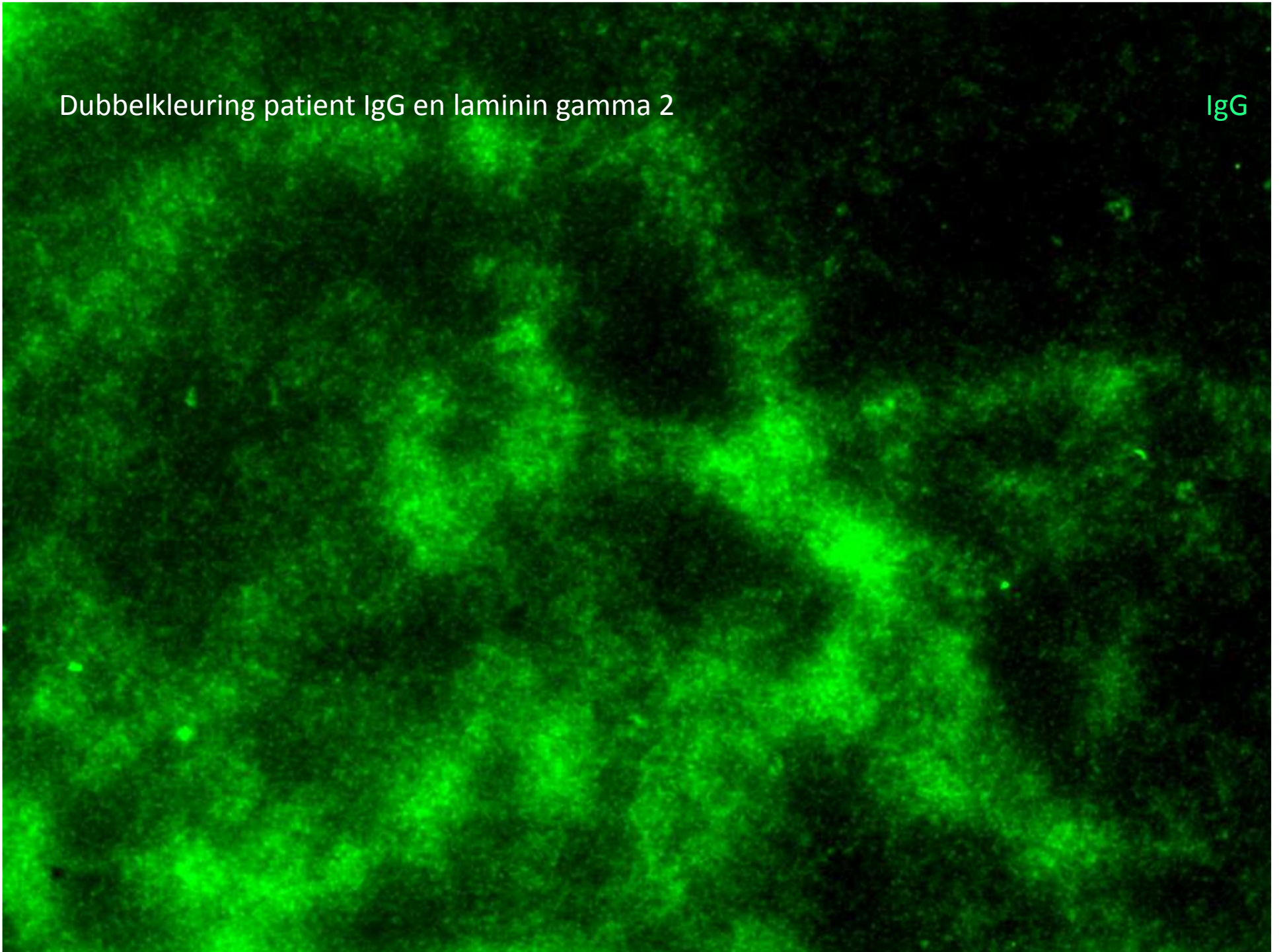


#1 63x These are the laminin332 footprints to look for



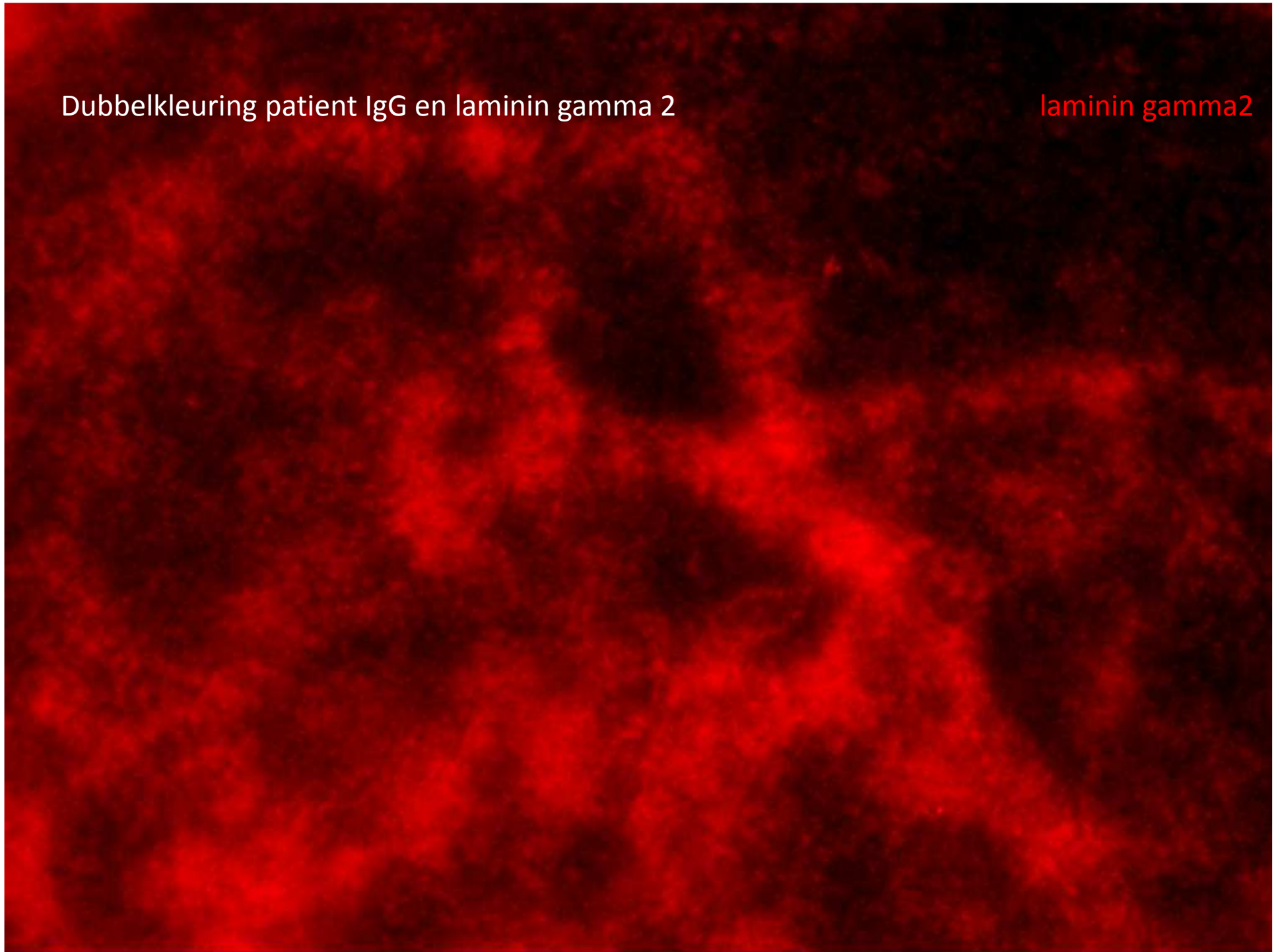
Dubbelkleuring patient IgG en laminin gamma 2

IgG



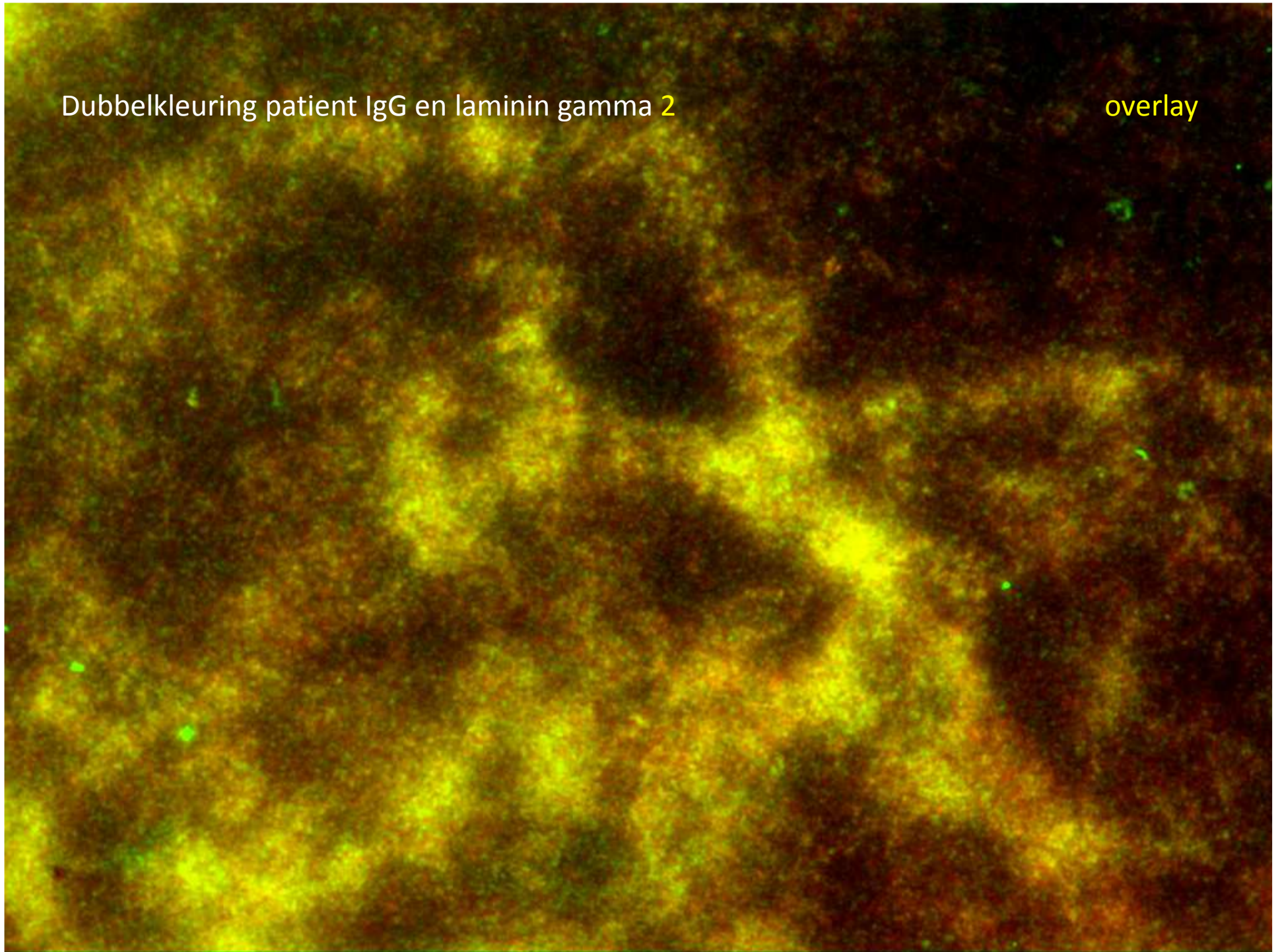
Dubbelkleuring patient IgG en laminin gamma 2

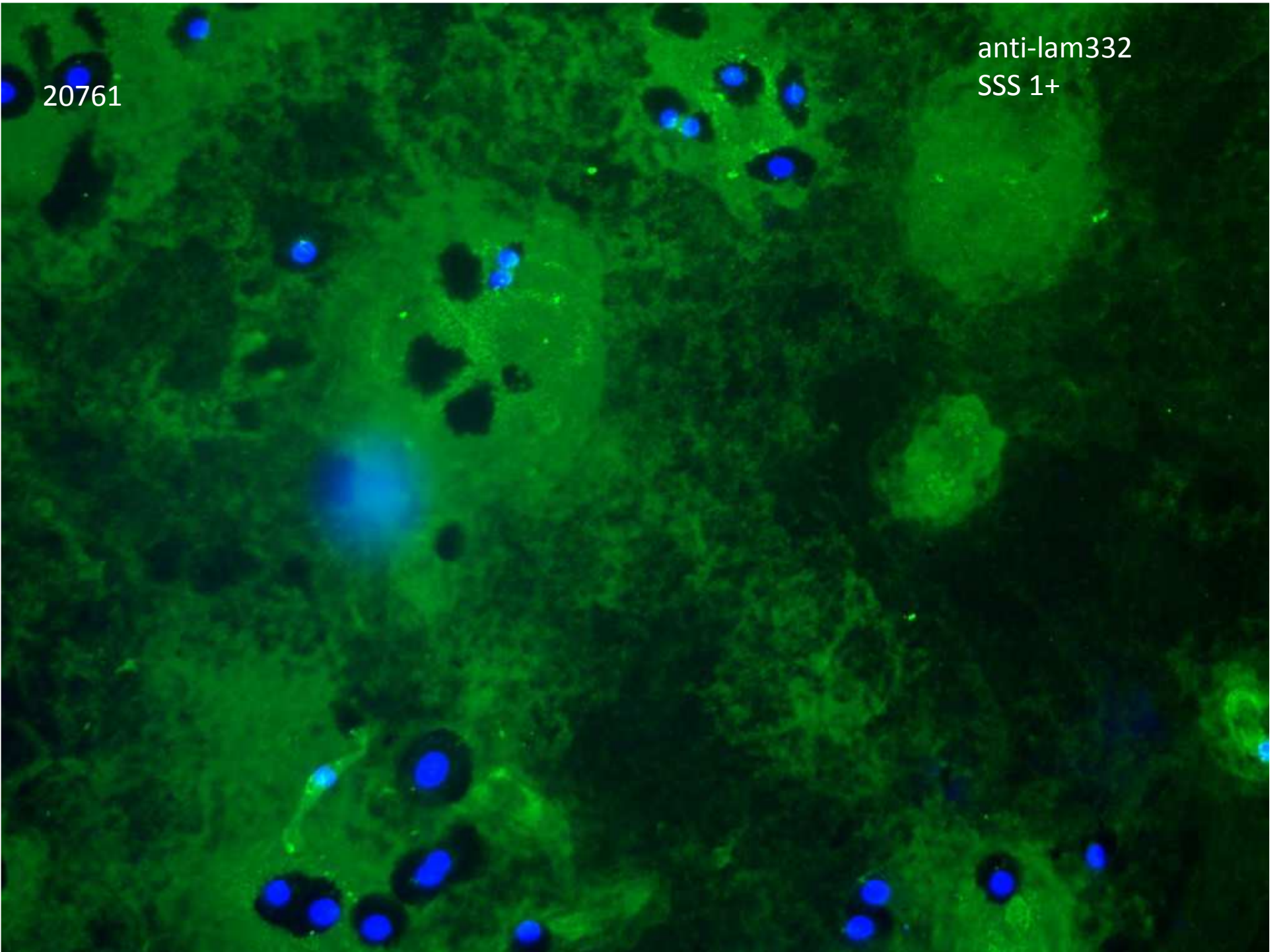
laminin gamma2



Dubbelkleuring patient IgG en laminin gamma 2

overlay



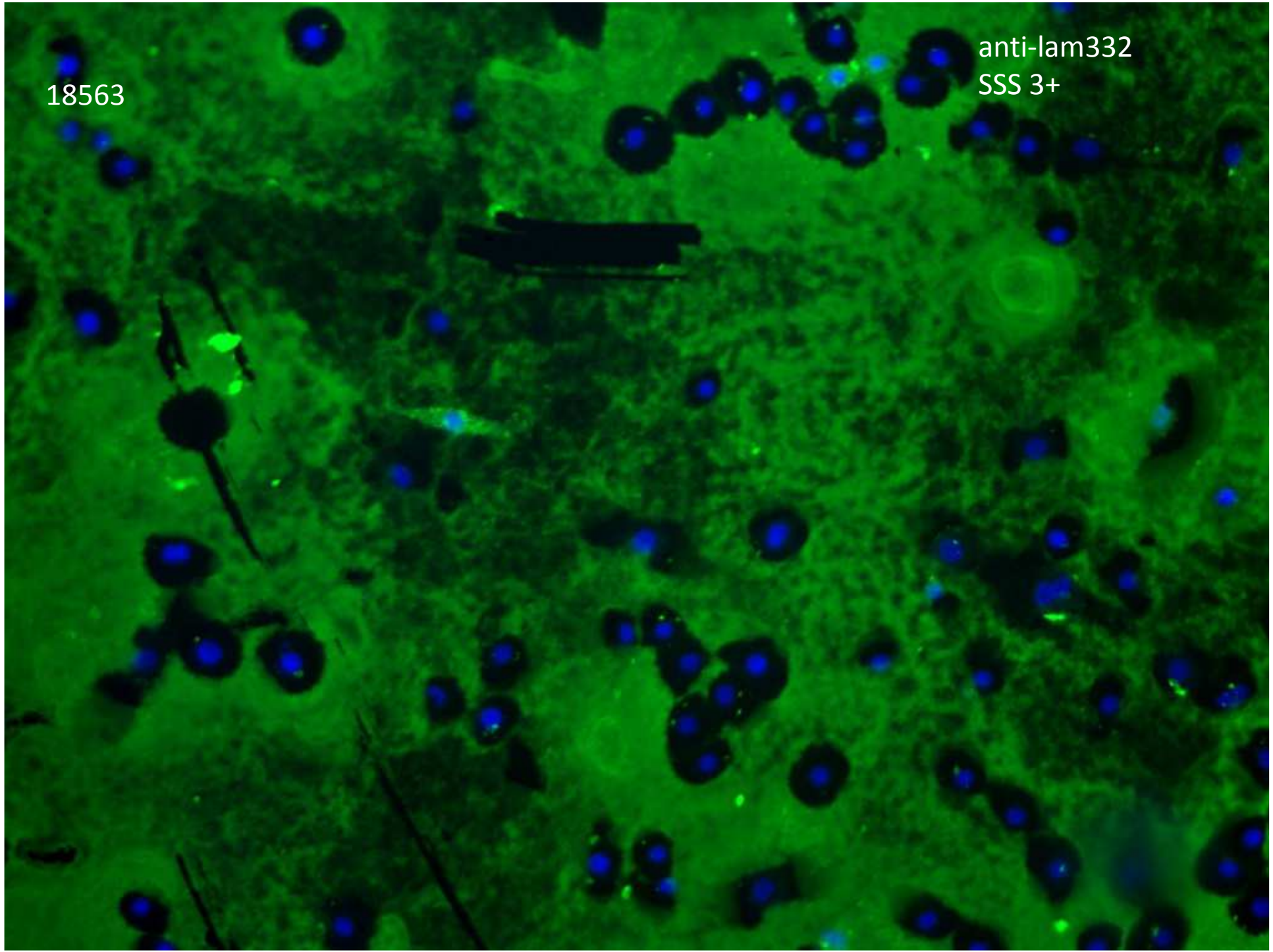


20761

anti-lam332
SSS 1+

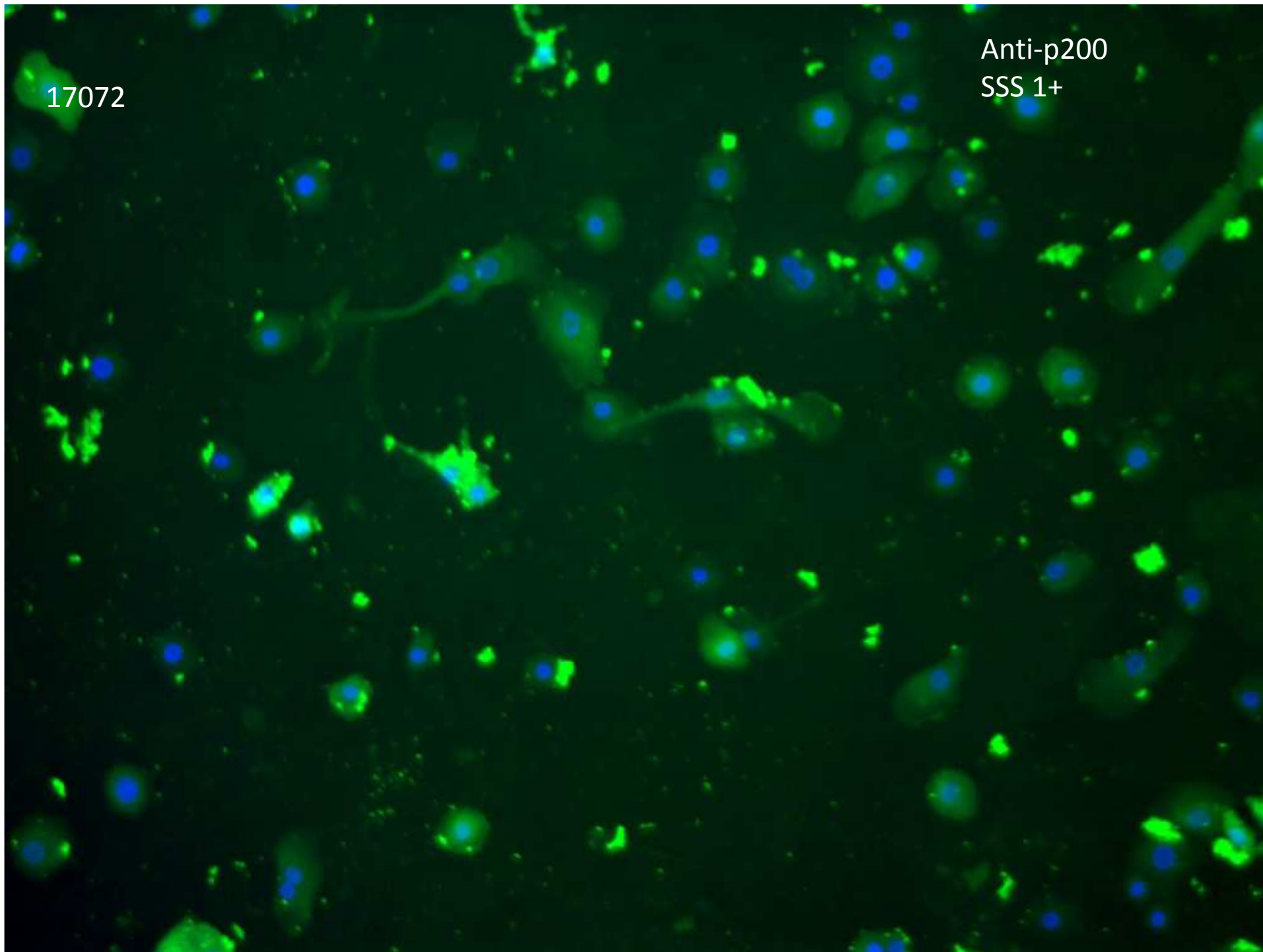
18563

anti-lam332
SSS 3+



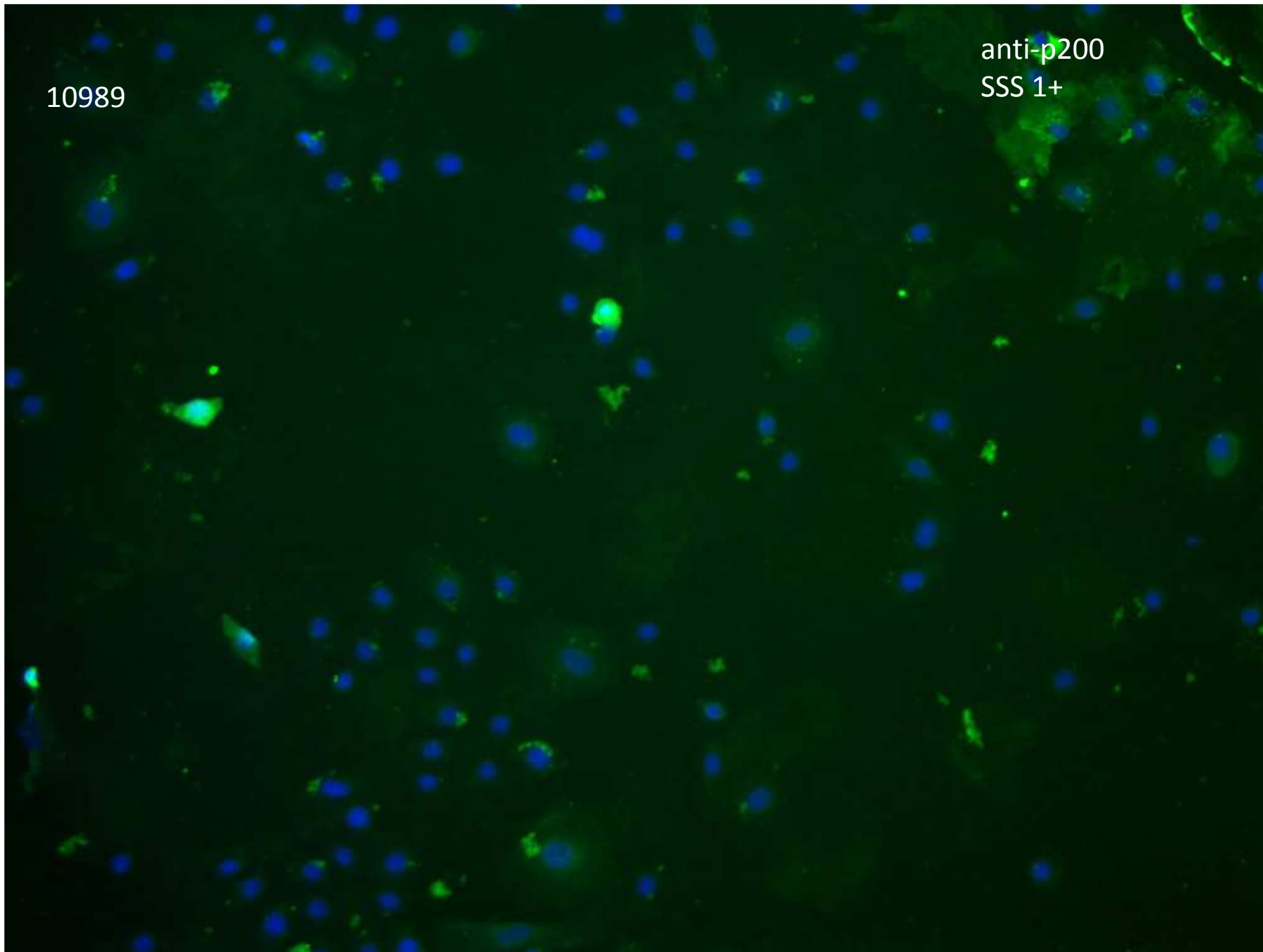
17072

Anti-p200
SSS 1+



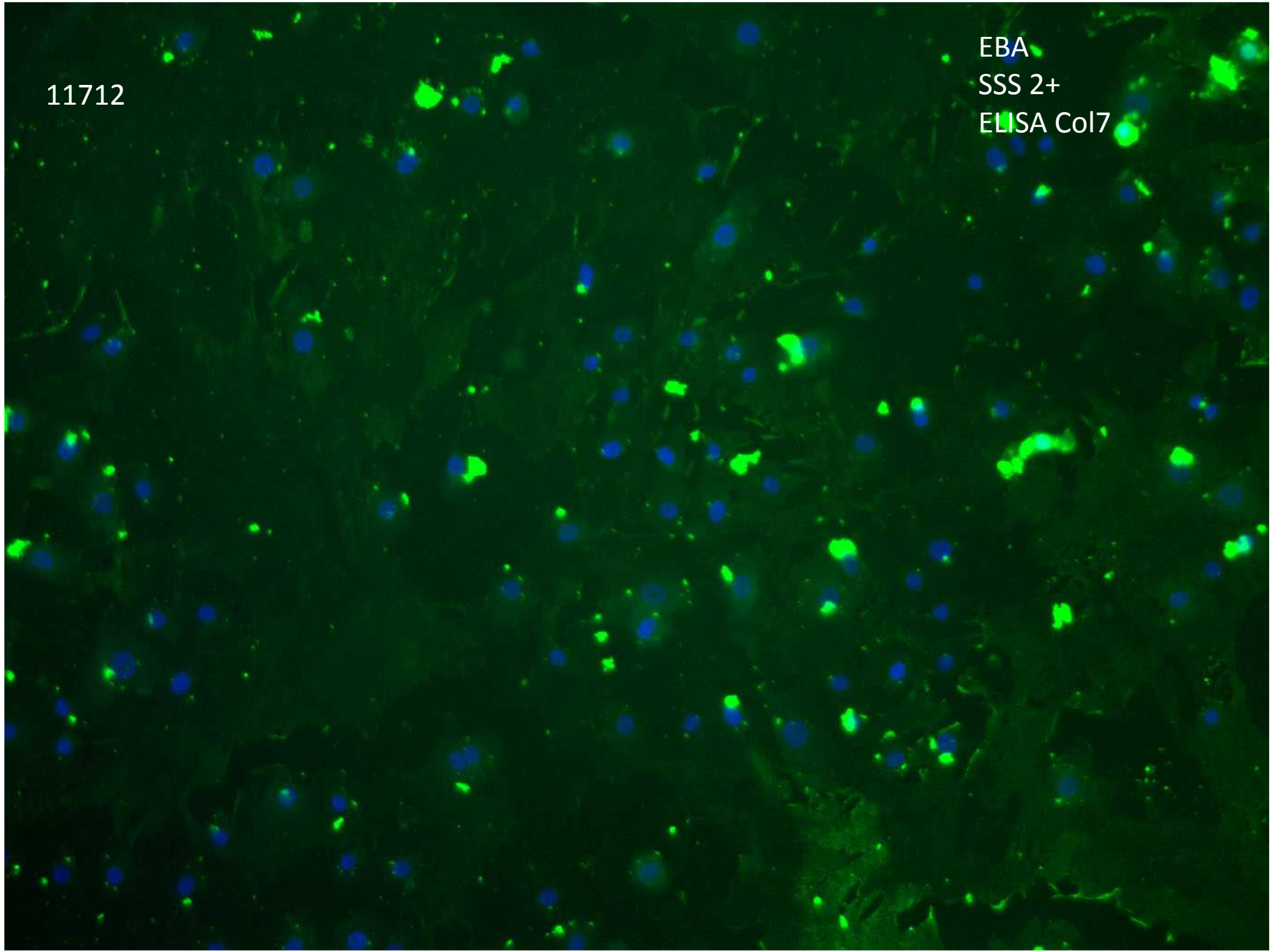
10989

anti-p200
SSS 1+



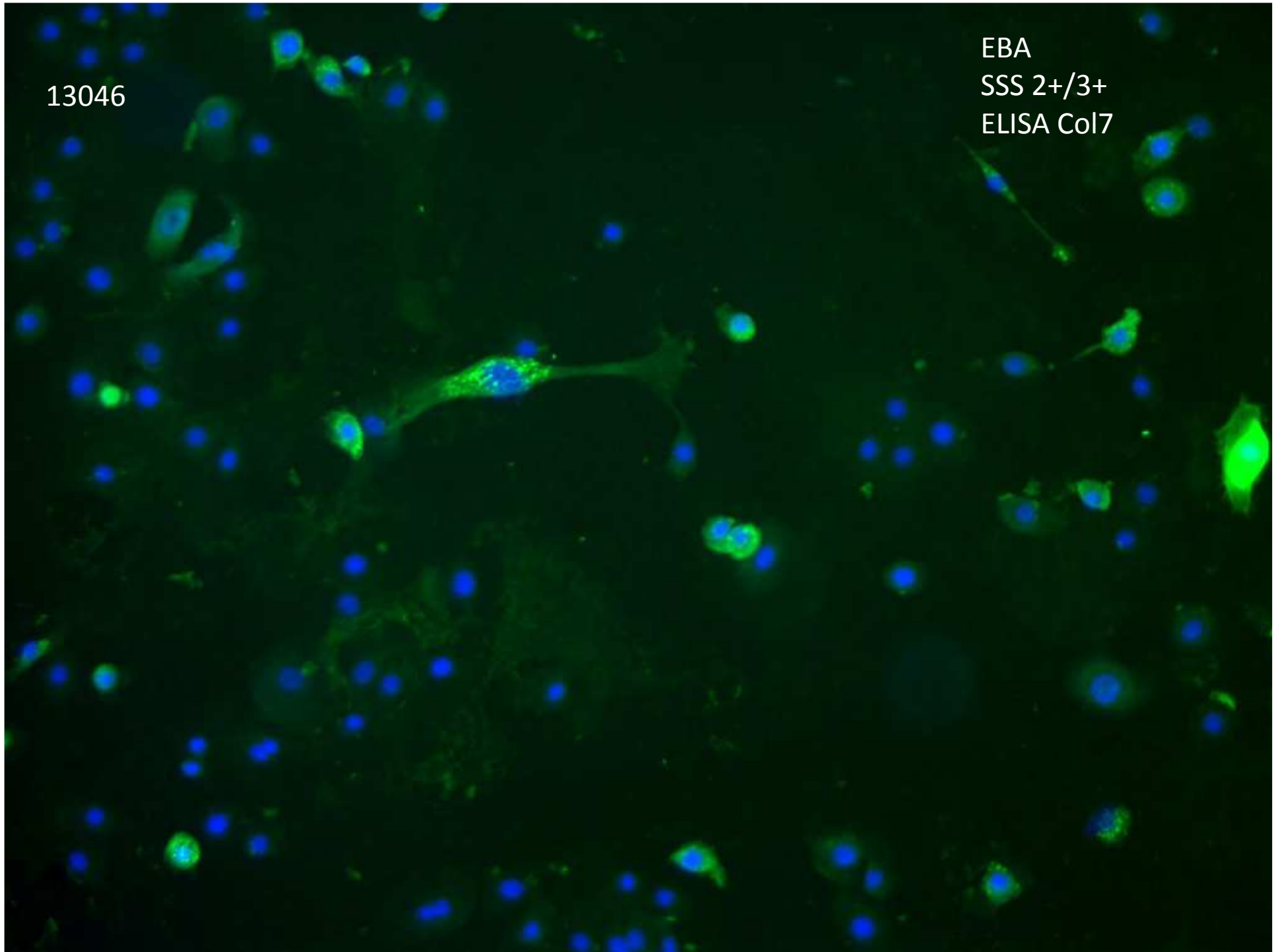
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EBA
SSS 2+
ELISA Col7



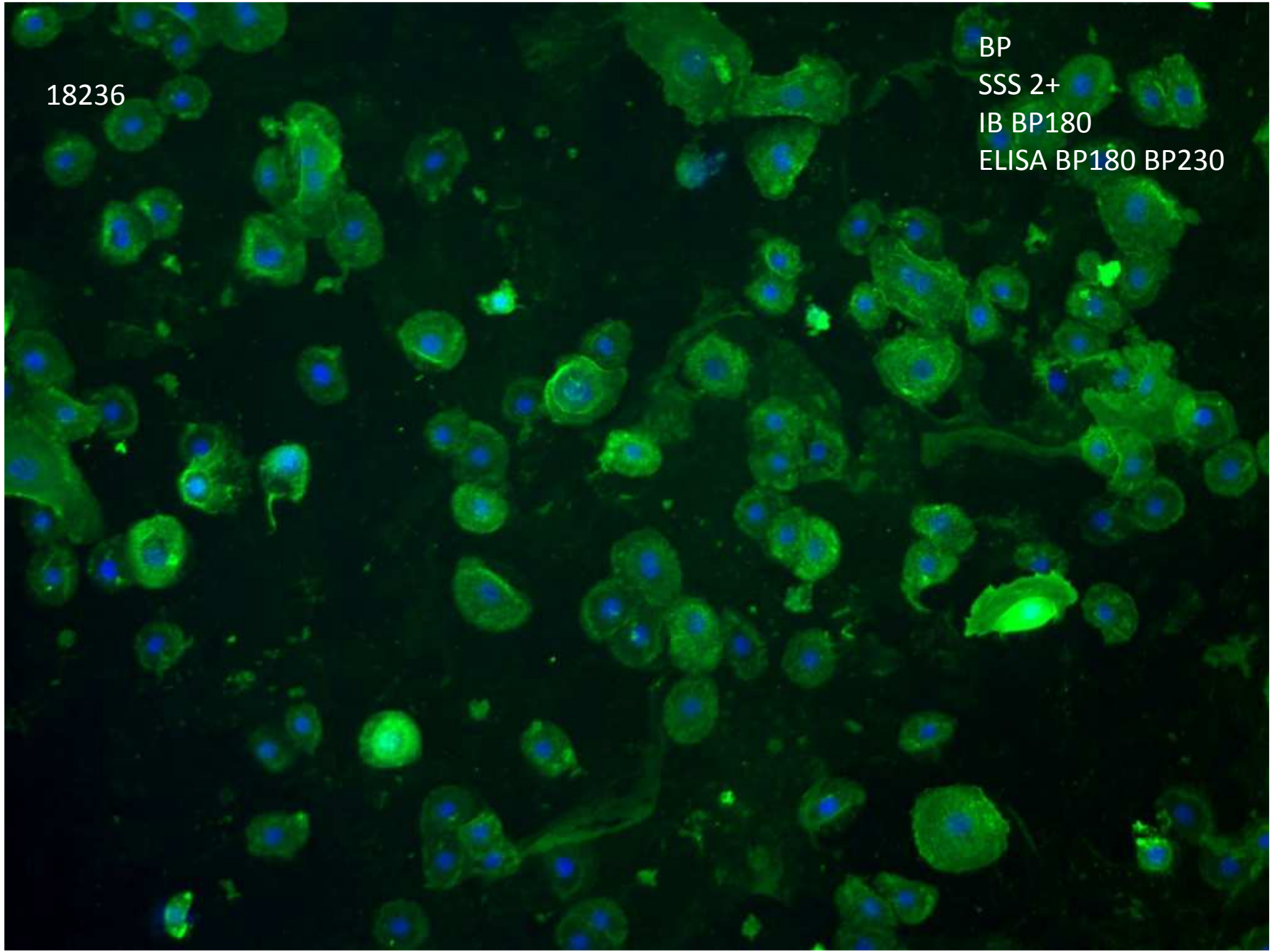
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EBA
SSS 2+/3+
ELISA Col17



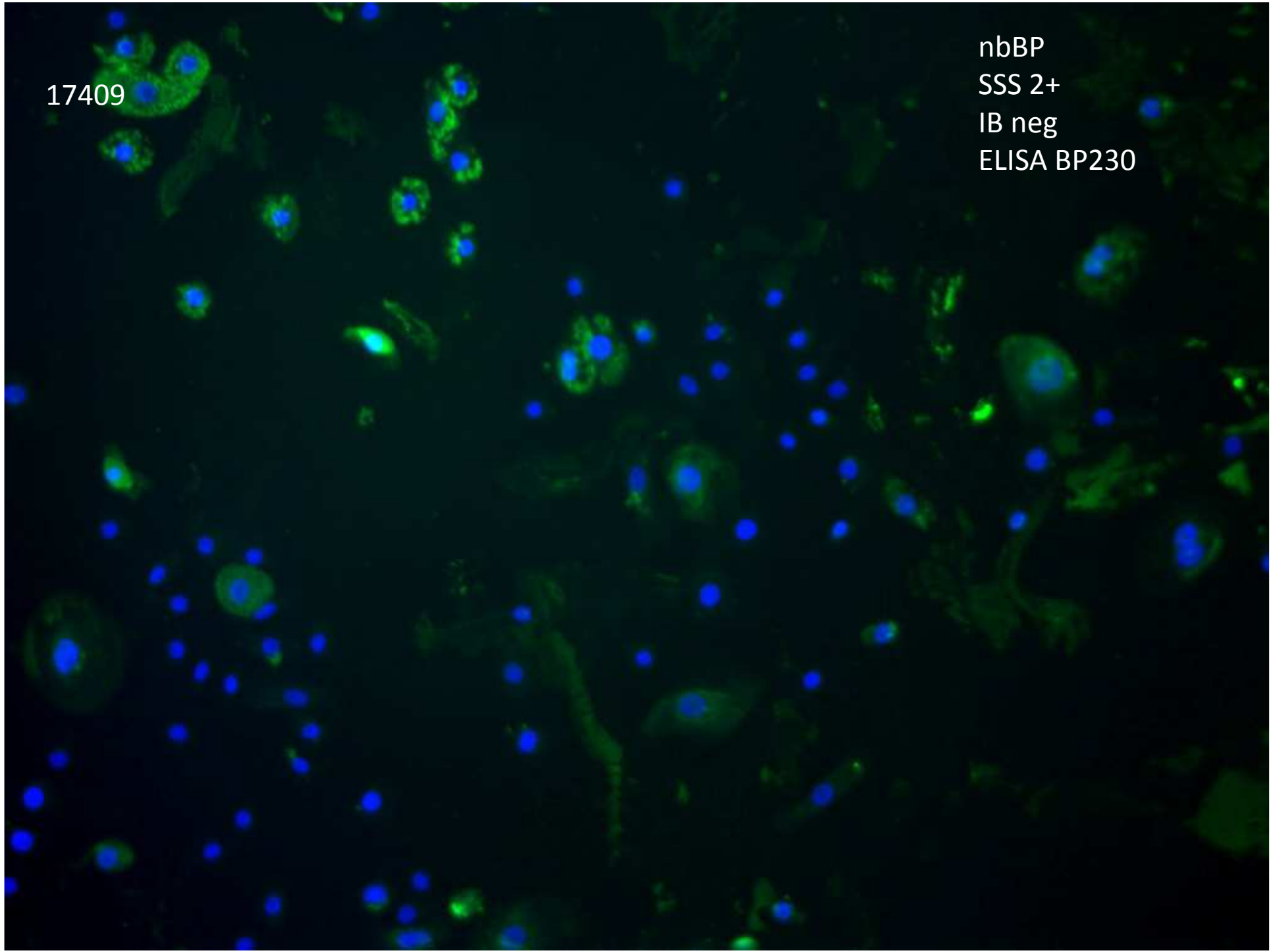
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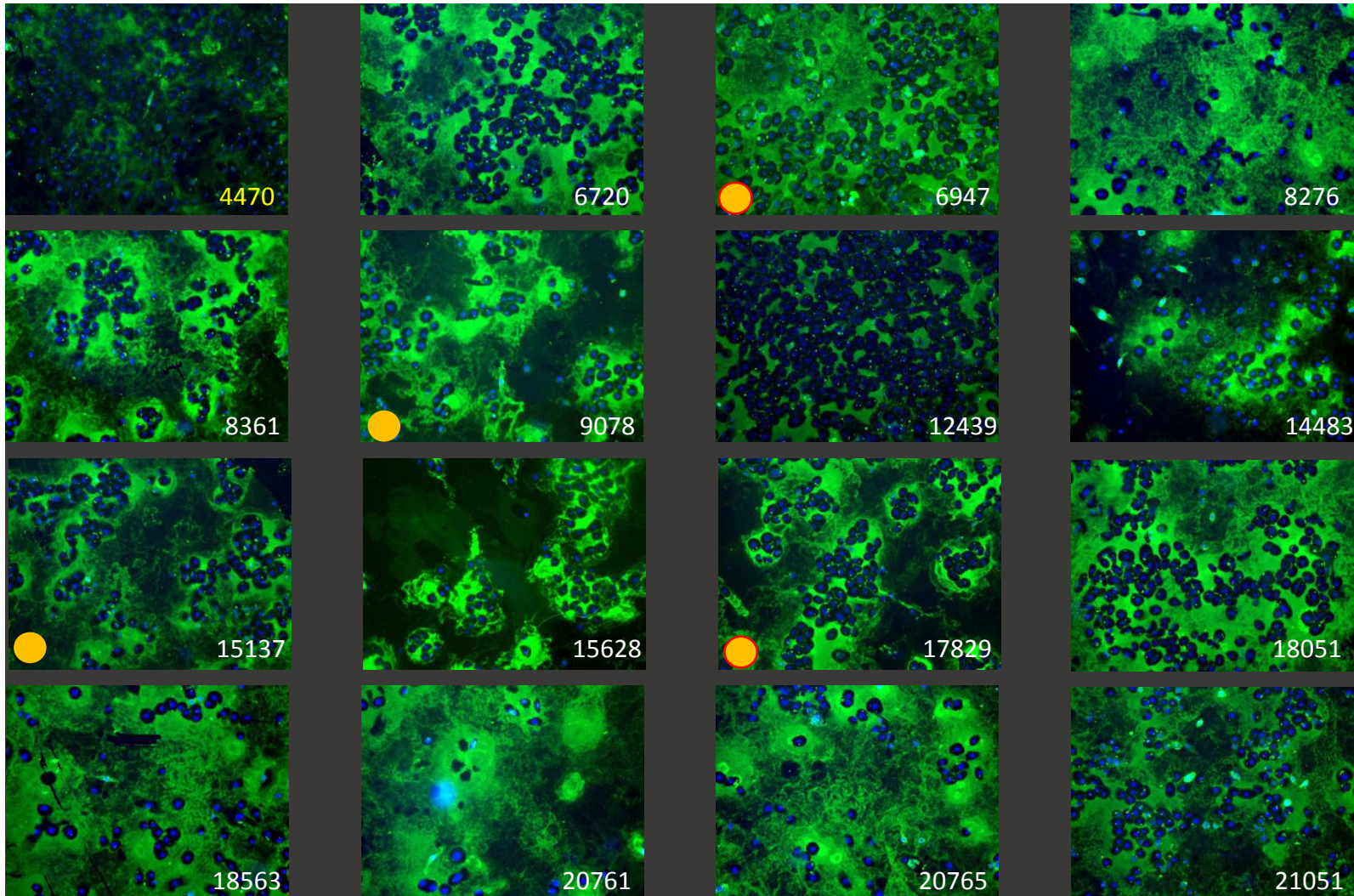
BP
SSS 2+
IB BP180
ELISA BP180 BP230



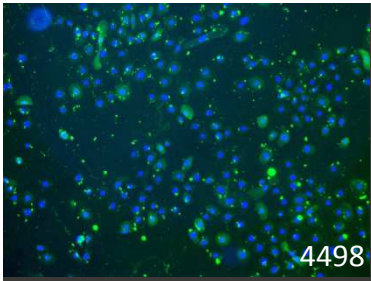
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nbBP
SSS 2+
IB neg
ELISA BP230

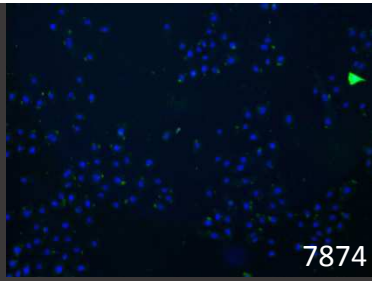




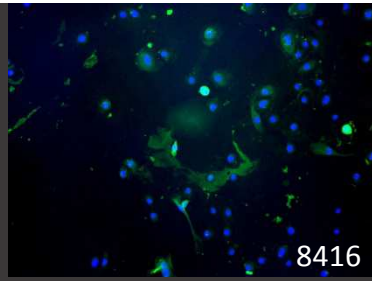
Anti-laminin 332 pemphigoid based on immunoprecipitation, immunoblotting or knock-out IIF analysis



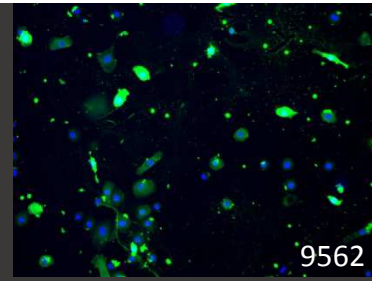
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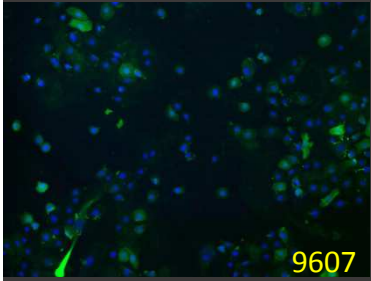
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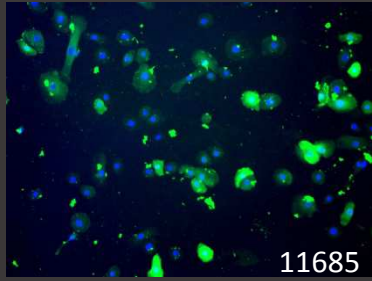
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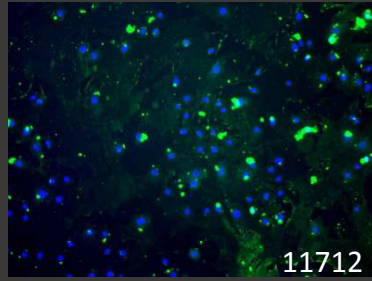
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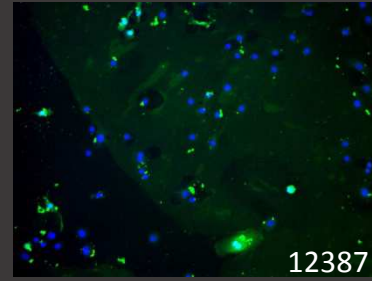
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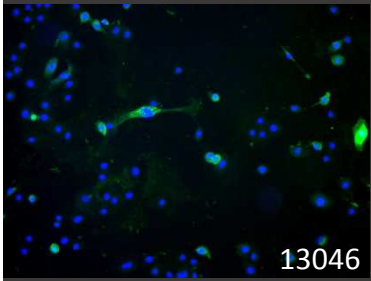
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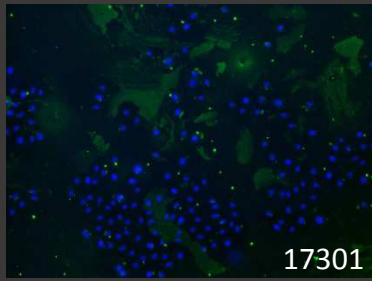
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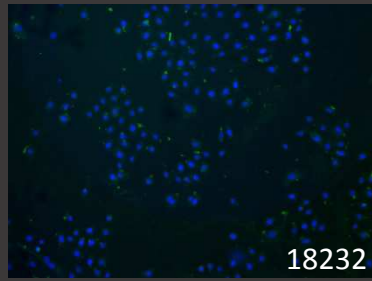
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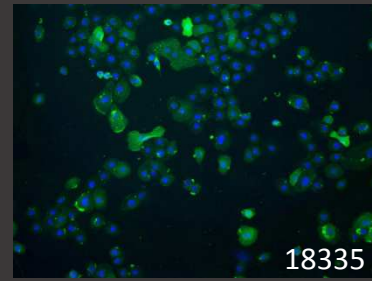
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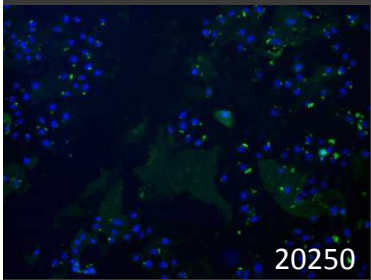
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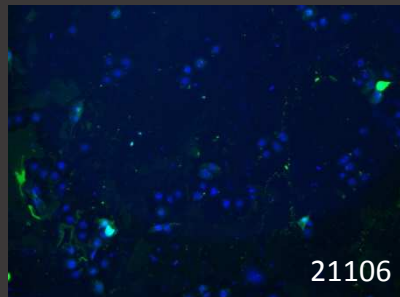
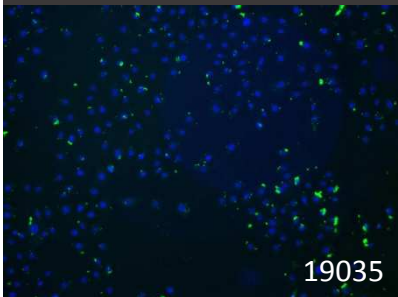
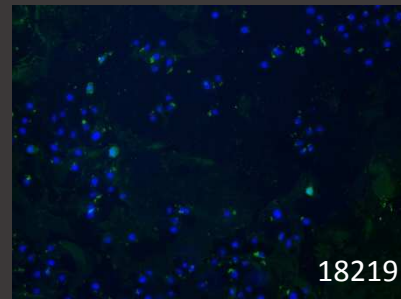
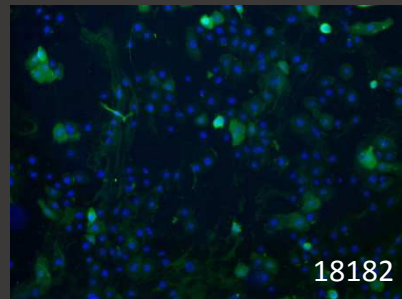
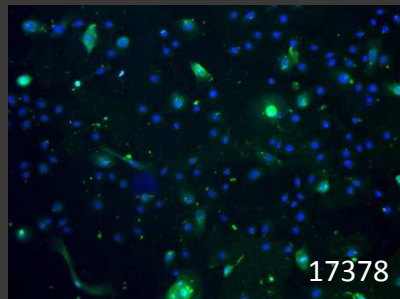
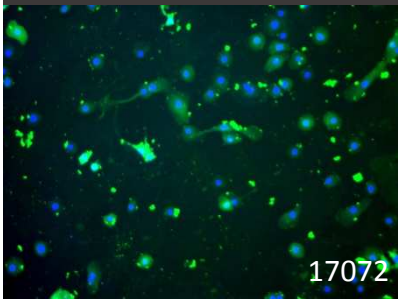
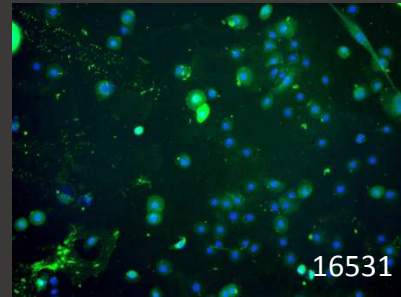
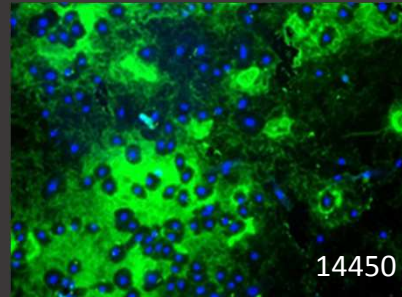
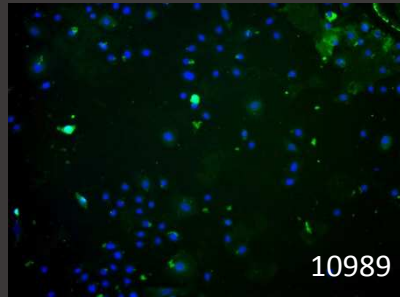
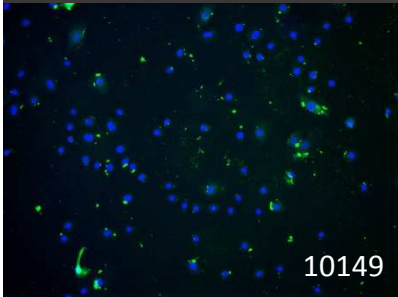
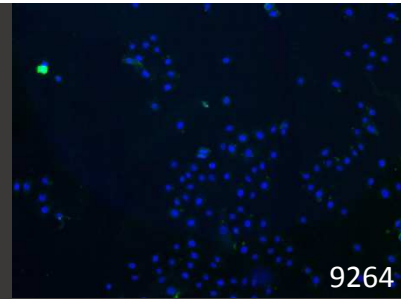
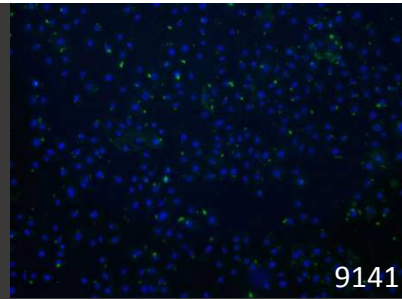
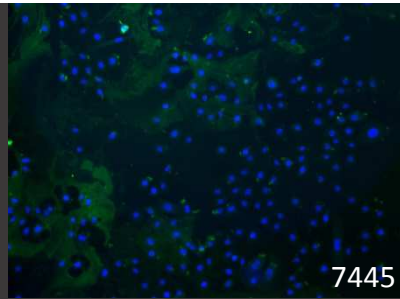
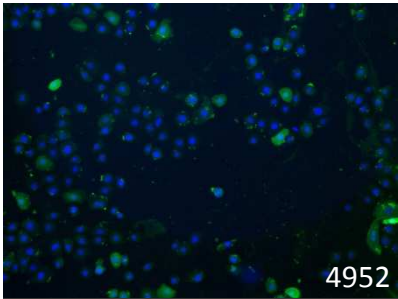


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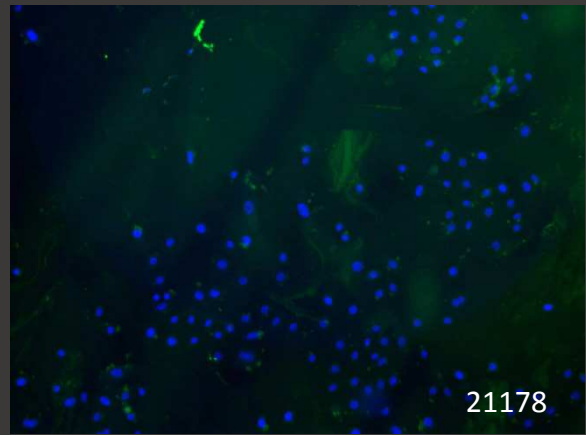
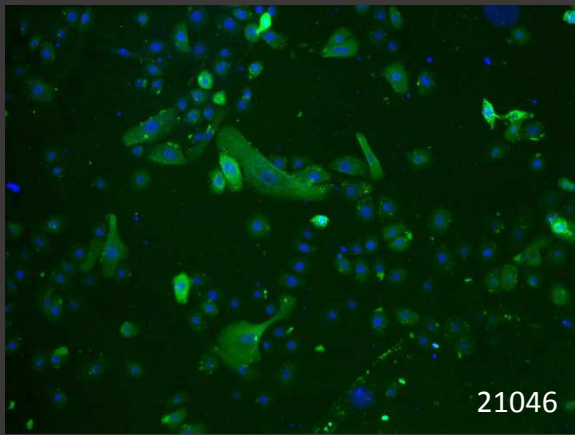
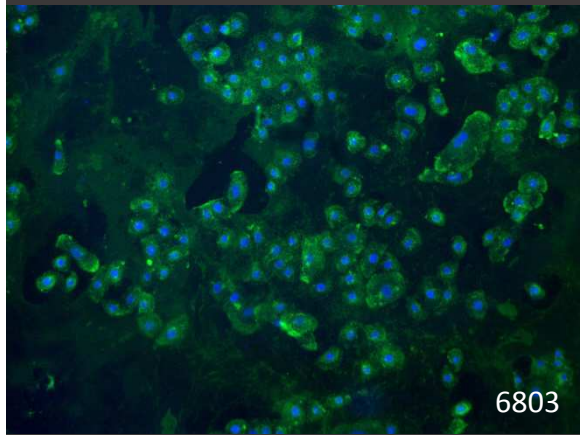
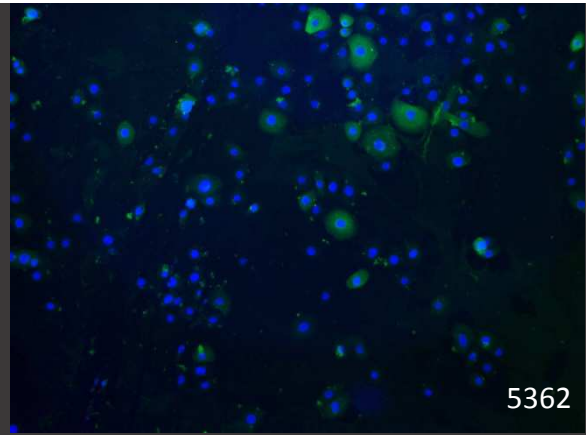
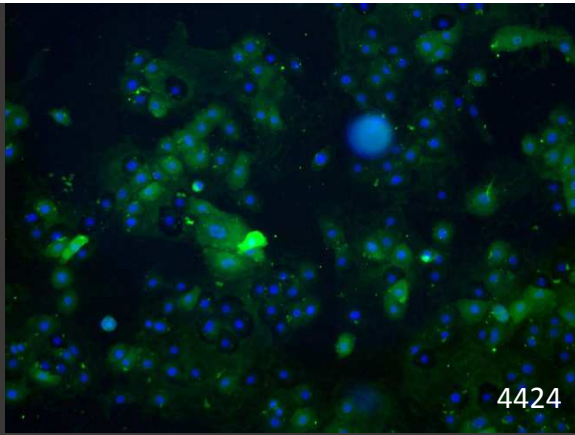
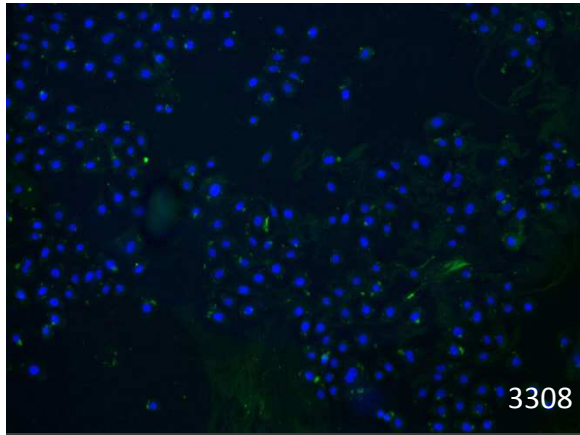


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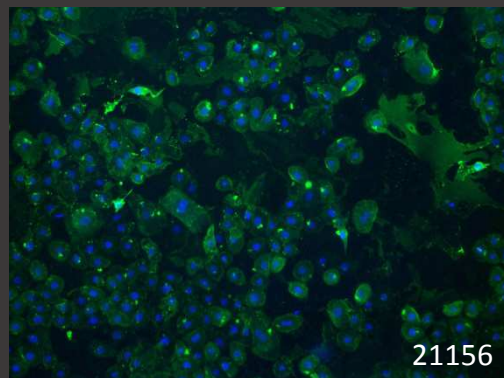
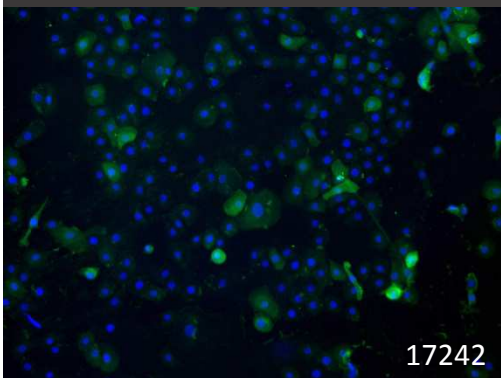
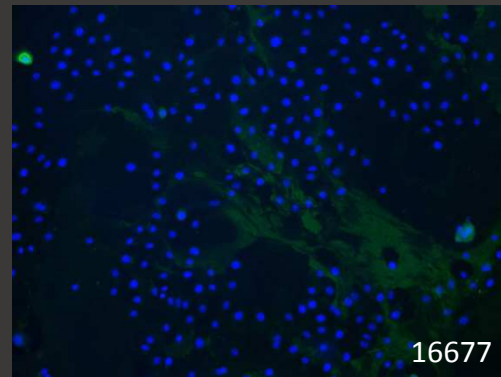
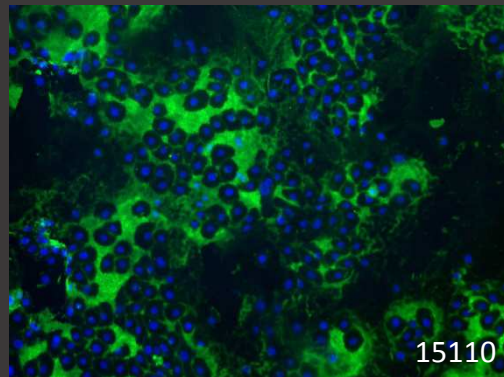
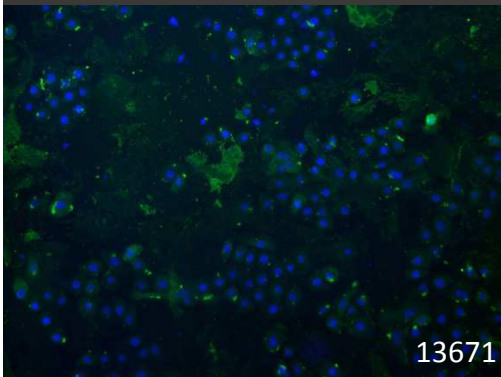
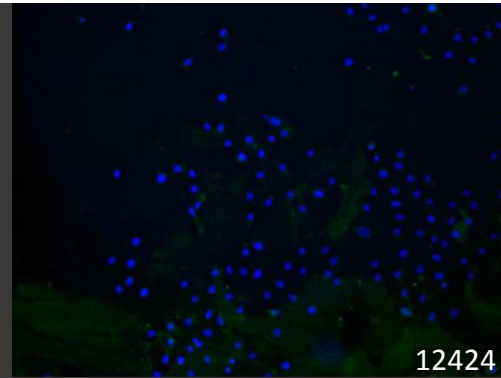
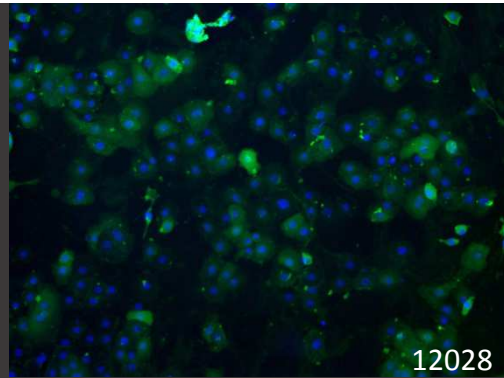
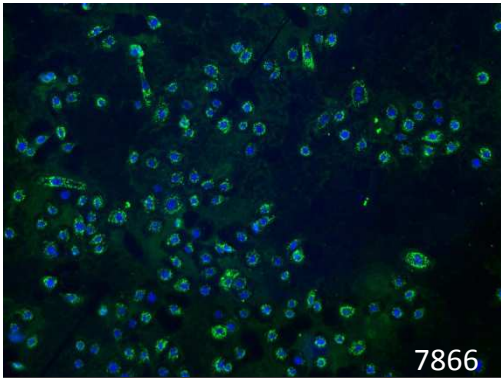
SSS-IIF positive epidermolysis bullosa aquisita



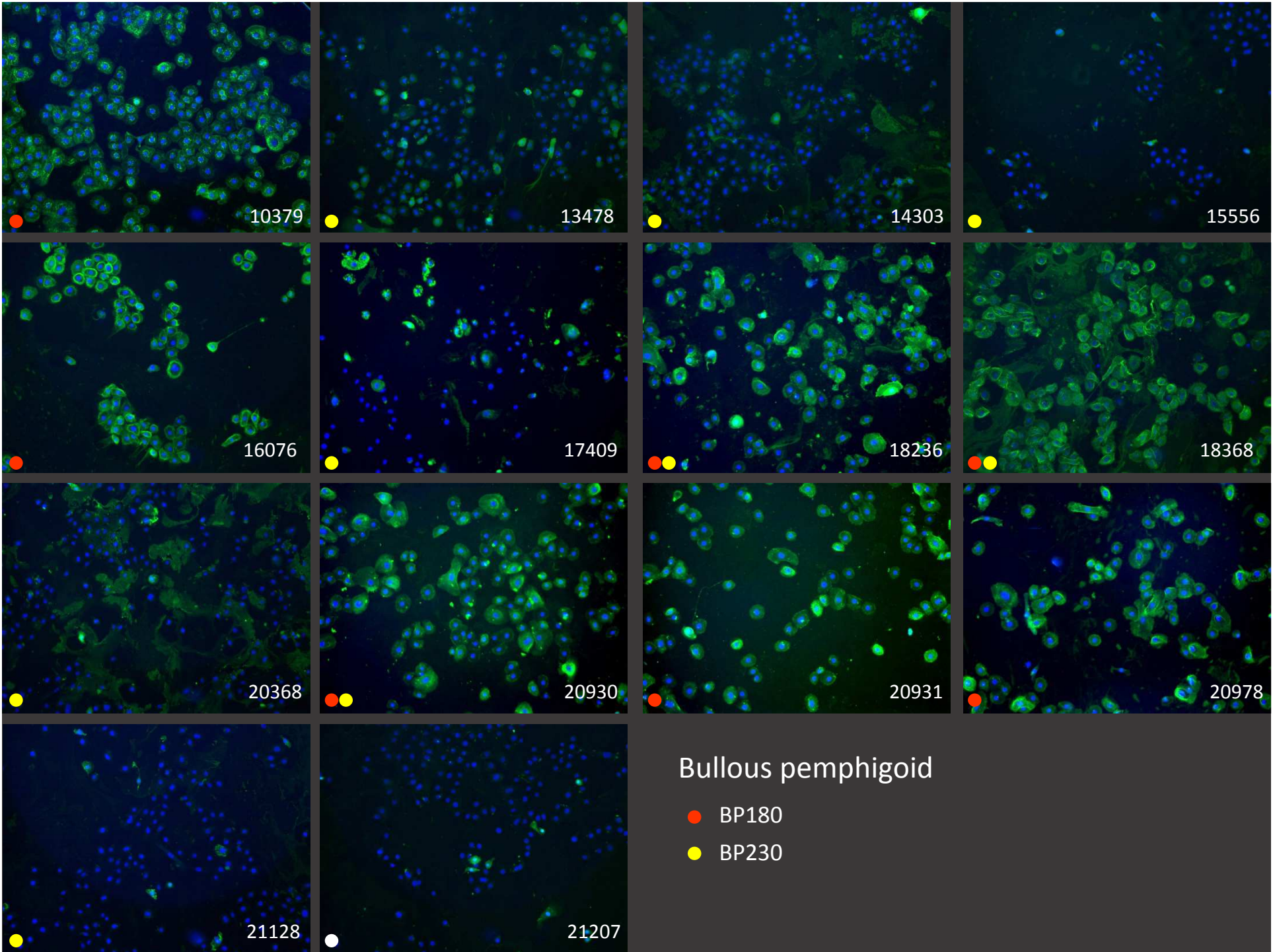
SSS-IIF positive anti-p200 pemphigoid based on both immunoblotting and knock-out IIF analysis



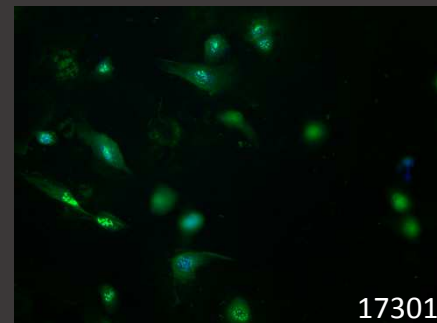
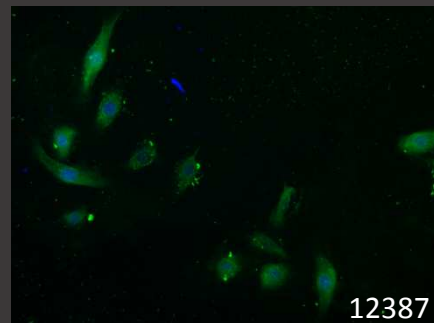
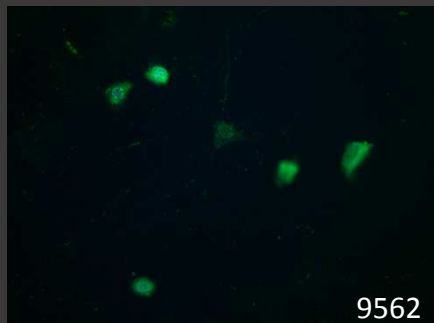
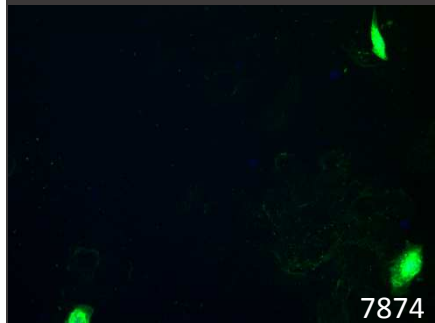
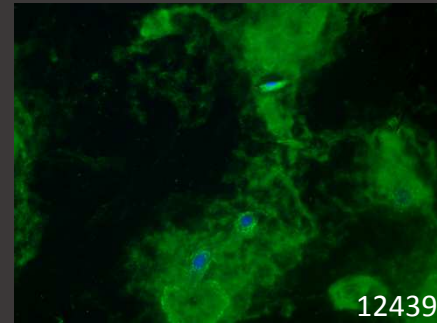
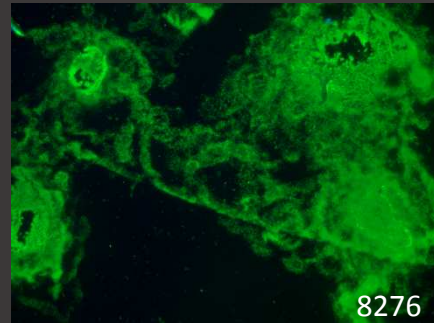
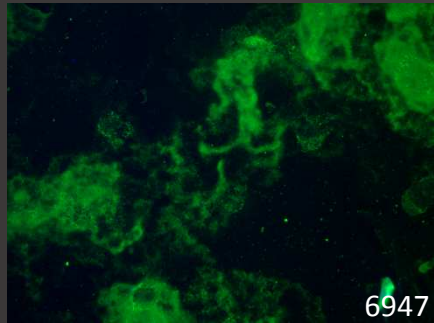
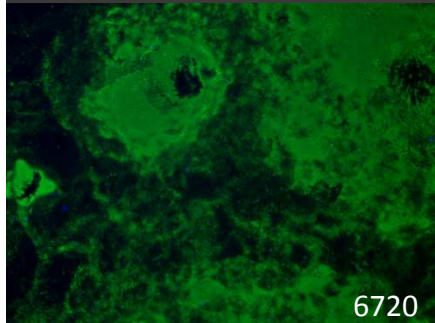
SSS-IIF floorstaining with uncharacterized antigen



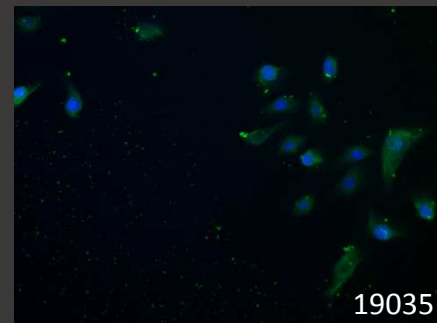
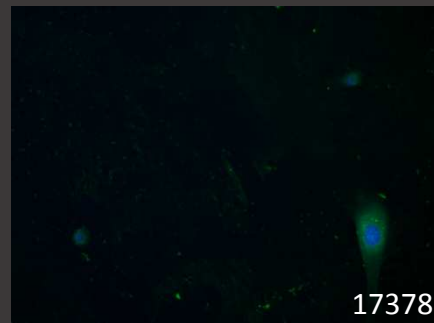
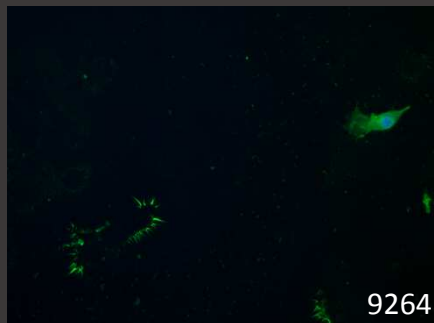
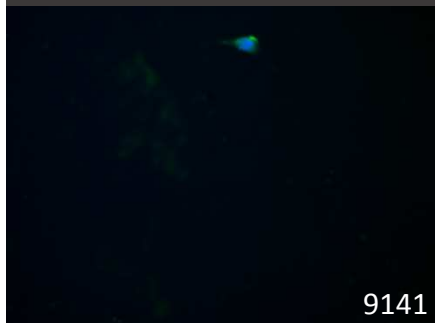
Combined roof/floor SSS-IIF staining
with uncharacterized floor antigen



Serum on air-dried frozen cells

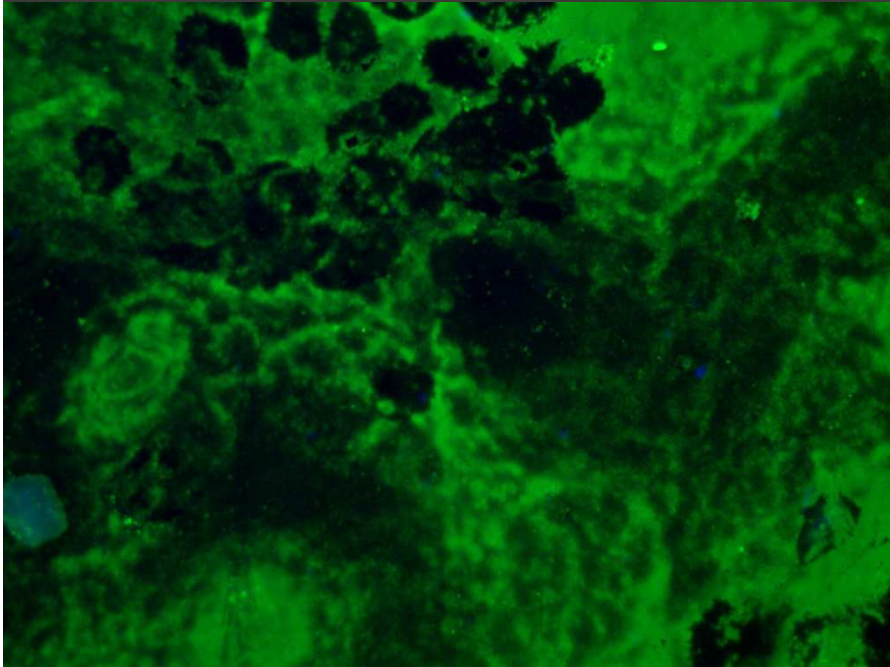


EBA

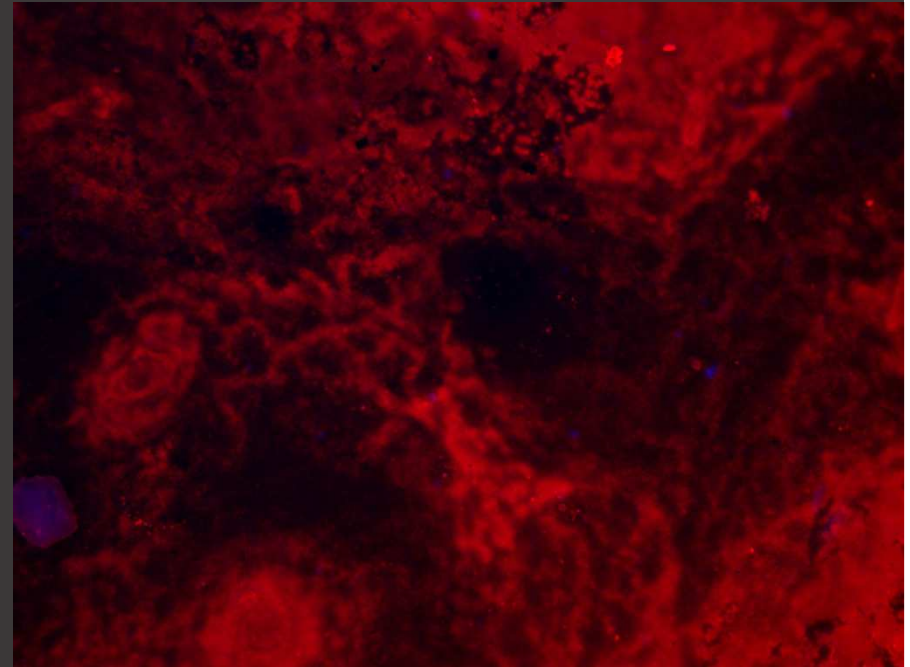


p200

Serum and anti-laminin γ 2 on air-dried cells stored at -30°C for 18 months



Serum 15137



anti-laminin γ 2

