



- Podocytes, negative IgG control
- Podocytes (Itga3^{fl/fl}, Cd151^{fl/fl})
- Podocytes (*Itga3^{-/-}, Cd151^{-/-}*)

Homo sapiens	336	EEVGGAIYVFMNQ	Α	GTSFPAHPSLLLHGPSGSAFG	376
Pongo abelii	336	EEVGGAVYVFMNQ	Α	GTSFPAHPSLLLHGPSGSAFG	376
Macaca mulatta	334	EEVGGAVYVFMNQ	Α	GTSFPAHPSLLLHGPSGSAFG	374
Pan troglodytes	437	EEVGGAIYVFMNQ	А	GTSFPAHPSLLLHGPSGSAFG	477
Bos taurus	335	EEVGGAIYIFMNQ	А	GTSFPDHPSLLLHGPSRSAFG	375
Canis lupus familiaris	336	EEVGGAIYVFMNQ	А	GTSFPAHPSLLLQGPSRSAFG	376
Mus musculus	337	EEVGGAVYVFMNQ	Α	GASFPDQPSLLLHGPSRSAFG	377
Monodelphis domestica	336	EEIGGAVYIFMNQ	G	GI-FPLHPSRHLHGPSGSTFG	376
Anolis carolinensis	461	EEKGGAVYVYMNL	G	GTFHSSANLTLTGPSNSSFG	501
Oreochromis niloticus	422	AEVGGAVYVYMNV	G	GRFNSETS-MVLKGPAGSAFG	462
Ornithorynchus anatinus	422	AEVGGAVYVYMNV	G	GRFNSETS-MVLKGPAGSAFG	462
Xenopus laevis	311	EEIGGAVYVYNNV	R	VFSLIKQQWCSMGPAFLG	351
Danio rerio	331	KEEGGAVYIFMNE	Ν	GSFQKKASLVLKGKKGSGFG	371
Dicentrarchus labrax	335	NDKGGAVYIFMNE	Ν	GSFQKTAS-VVLKGKTDSAFG	475



calnexin \rightarrow



pre- α 3 \rightarrow



WT α3



A349S $\alpha 3$

EXON	FORWARD PRIMERS $5' \rightarrow 3'$	REVERSE PRIMERS $5' \rightarrow 3'$	PRODUCT SIZE (bp)
1a	cgtcacatccatcttgctc	atccgtgggtctatcttcct	460
1b	ctccctcctgtcctccttg	cgaccgagtagccgaagag	539
1c	acgcctgatgctctgtgc	gactaccagcgaggtgctta	605
2	ttttccttgcctgccttac	gcacctcacccatacttcag	583
3	actcactgcccacaaggat	gacacacagccacaggaag	601
4	gtctctcatccttccctgct	aagtcatggtggttgctgat	659
5	ggcaaaatgctcaccaataa	ttccaagtagggcaagaaag	490
6	ctactttcttgccctacttgg	ataaagcetgactgcaaacc	487
7	atattggcatctccatgtcc	acatetgeacatectete	541
8	ctctgtccctgatgctctg	gettetetecatggattace	389
9	cccagcaggtacagagagac	gagacaacagagccagacag	446
10	tettetteatetttgtetgeae	aatgaggttgggtagagagg	372
11+12	cagacctgctttgtggactc	acaccaatagcettecaaac	563
13	agtaggaagtcgcaatttgg	catetgeaagttgeteteae	483
14	atecteaaccaggeacag	gcacctggaggagaaagc	405
15	aggtgggatggtcagaaac	attetecaageageagagae	477
16	ctctggtctgggccttc	ggcctcttctcaccctctac	379
17 + 18	gtggggtgggggggggggg	cagaggagtttgggagatagc	481
19	gctateteceaaacteetetg	aaccacetecatettaccaac	684
20	ctgtggaggatgtaggaagc	gaggaagaattgggagcag	515
21	caacecteteaaceteacte	gceteateaceteateacae	435
22	tgtgcatgagtgaaaggaag	acacatccatgcaaagacac	414
23	ctggctgacagatcctttg	agacaccagaactcctccag	382
24	agcaggacaaacagcaggt	gtgtggtcagaagccagag	457
25	cttctgaccaccaccaa	cttgcccttgaccttgttc	564
26	tctggctttgaggagttctg	gctctttggcttgttttgg	599

Supplemental Table 1: Primers used for *ITGA3* mutation analysis

Supplemental table and figures

Supplemental Table 1. Primers used for *ITGA3* mutation analysis.

Supplemental Figure 1. Abnormalities in patient kidneys. (A) The upper glomerulus shows extensive mesangiolysis with "free-floating" cross-sections of irregularly widened capillaries. The lower glomerulus has a segmental scar (arrow head). (B) The upper segment of the upper glomerulus contains a cellular crescent (fresh extracapillary proliferation without interposition of silver-positive extracellular matrix, single arrow). The middle glomerulus displays a fibro-cellular lesion of extracapillary proliferation with fine lines of silver-stained extracellular matrix between the epithelioid cells (double arrow). The upper segment of the lower glomerulus shows segmental mesangial expansion with increased cellularity, and thickening and splitting of Bowman's capsule (triple arrow).

Supplemental Figure 2. Generation of CD151/ α 3-deficient podocytes. Podocytes were isolated from glomeruli from *Itga3*^{flox/flox};*Cd151*^{flox/flox};*Trp53*^{+/-} mice, and the *Cd151* and *Itga3* genes were deleted by adenoviral delivery of Cre recombinase. Cell-surface expression of CD151 and α 3 was examined by flow cytometry.

Supplemental Figure 3. Alignment of the amino acid sequence of α 3 in different species. The affected residue (alanine 349 in humans) is boxed.

Supplemental Figure 4. The A349S mutation does not prevent interaction of the α 3 precursor with the ER chaperone protein calnexin. Integrin subunits α 3 (with antibody 29A3) or β 1 (using antibodies 9EG7 or MB1.2) were precipitated from podocyte lysates, and α 3, β 1, and calnexin were detected by Western blotting.