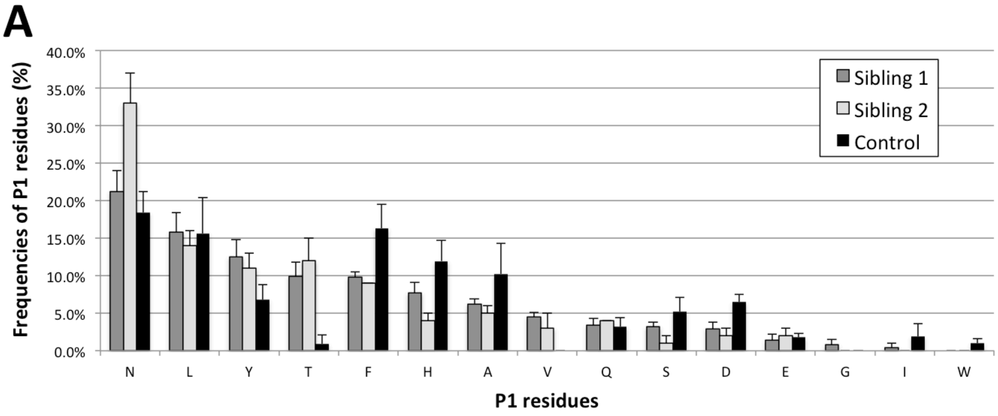

Supplementary material 1. Amino acid P1 residue cleavage distribution in TGFBIp. Frequencies of P1 cleavage sites observed in semi-tryptic TGFBIp peptides of amyloid deposits and control tissue. Tissues from both siblings display higher cleavage frequencies after threonine and valine residues and to a less extent after tyrosine residues. Numbers of counted P1 sites were 626 (sibling 1), 527 (sibling), and 282 (control).

Supplementary material 2. MS protein and peptide information

Supplementary material 3. emPAI quantification and total protein identification list

Supplementary material 4. MS/MS spectra of protein hits identified by one peptide

Table 1: The 30 most abundant proteins according to emPAI-based molar % identified in corneal amyloid deposits and healthy corneal tissue

	A546D/P551Q (Sibling 1)				**A546D/P551Q (Sibling 2)**				**Healthy corneal stroma (3 individuals)**			
#	**Name**	**Molar %**			**Name**	**Molar %**			**Name**	**Molar %**		
1	TGFBIp	69.5	±	11.3	TGFBIp	47.0	±	7.9	Collagen alpha-1(I) chain	36.5	±	5.8
2	Serum amyloid P-component	3.0	±	1.2	Collagen alpha-1(I) chain	4.1	±	0.5	Collagen alpha-2(I) chain	17.3	±	1.5
3	Apolipoprotein A-I	2.5	±	0.6	Serum amyloid P-component	3.8	±	1.0	TGFBIp	7.8	±	2.0
4	Collagen alpha-1(I) chain	2.2	±	0.4	Lysozyme C	3.4	±	1.5	Ig kappa chain C region	5.0	±	3.0
5	Clusterin	2.2	±	0.7	Dermcidin	2.9	±	1.9	Serum albumin	3.4	±	0.3
6	Collagen alpha-2(I) chain	2.0	±	1.0	Collagen alpha-2(I) chain	2.6	±	0.3	Keratocan	1.8	±	0.2
7	Serine protease HTRA1	1.4	±	0.8	Clusterin	2.5	±	0.7	Decorin	1.6	±	0.5
8	Apolipoprotein A-IV	1.3	±	0.5	Lactotransferrin	2.0	±	0.6	Lumican	1.4	±	0.3
9	Dermcidin	1.1	±	0.6	Apolipoprotein A-IV	1.7	±	0.5	Ig kappa chain V-III region SIE	1.3	±	0.4
10	Apolipoprotein D	0.8	±	0.4	Serine protease HTRA1	1.5	±	0.6	Collagen alpha-3(VI) chain	1.2	±	0.2
11	Apolipoprotein E	0.8	±	0.1	Thioredoxin	1.1	±	0.2	Ig gamma-1 chain C region	1.1	±	0.2
12	Protein S100-A8	0.7	±	0.5	Apolipoprotein E	1.0	±	0.3	Ig gamma-3 chain C region	1.1	±	0.3
13	Serum albumin	0.6	±	0.2	Complement component C9	1.0	±	0.2	Collagen alpha-1(VI) chain	0.9	±	0.1
14	Lysozyme C	0.6	±	0.4	Apolipoprotein A-I	1.0	±	0.3	Ig lambda-2 chain C regions	0.8	±	0.4
15	Prostaglandin-H2 D-isomerase	0.5	±	0.3	Apolipoprotein D	0.9	±	0.2	Ig kappa chain V-III region NG9	0.8	±	0.4
16	Lumican	0.5	±	0.2	Fatty acid-binding protein, epidermal	0.8	±	0.2	Aldehyde dehydrogenase, dimeric NADP-preferring	0.7	±	0.1
17	Protein S100-A9	0.4	±	0.2	Actin, cytoplasmic 1	0.8	±	0.2	Collagen alpha-2(VI) chain	0.7	±	0.2
18	Ig gamma-1 chain C region	0.4	±	0.1	Vimentin	0.7	±	0.2	Vimentin	0.7	±	0.1
19	Histone H4	0.4	±	0.1	Cystatin-A	0.7	±	0.4	Collagen alpha-1(XII) chain	0.6	±	0.1
20	Ubiquitin-40S ribosomal protein S27a	0.4	±	0.3	Ig gamma-1 chain C region	0.6	±	0.2	Collagen alpha-2(V) chain	0.6	±	0.2
21	Complement component C9	0.4	±	0.1	Olfactomedin-like protein 3	0.6	±	0.2	Ig gamma-4 chain C region	0.5	±	0.1
22	Olfactomedin-like protein 3	0.3	±	0.1	Protein S100-A6	0.6	±	0.1	Prostaglandin-H2 D-isomerase	0.5	±	0.2
23	Angiopoietin-related protein 7	0.3	±	0.2	Cytokine-like protein 1	0.6	±	0.2	MAM domain-containing protein 2	0.4	±	0.1
24	Keratocan	0.3	±	0.1	Ubiquitin-40S ribosomal protein S27a	0.6	±	0.3	Histone H4	0.4	±	0.1
25	Decorin	0.3	±	0.2	Histone H4	0.5	±	0.1	Dermcidin	0.4	±	0.1
26	Ig alpha-1 chain C region	0.3	±	0.1	Protein S100-A8	0.5	±	0.1	Collagen alpha-1(V) chain	0.4	±	0.1
27	Phospholipase A2, membrane associated	0.2	±	0.1	Ig kappa chain V-III region VG	0.5	±	0.1	Protein S100-A6	0.4	±	0.2
28	Cytokine-like protein 1	0.2	±	0.1	Glyceraldehyde-3-phosphate dehydrogenase	0.5	±	0.3	Ig alpha-1 chain C region	0.4	±	0.2
29	Fatty acid-binding protein, epidermal	0.2	±	0.1	Prostaglandin-H2 D-isomerase	0.5	±	0.2	Alpha-enolase	0.3	±	0.1
30	Actin, cytoplasmic 1	0.2	±	0.1	Metalloproteinase inhibitor 3	0.5	±	0.2	Collagen alpha-1(III) chain	0.3	±	0.1