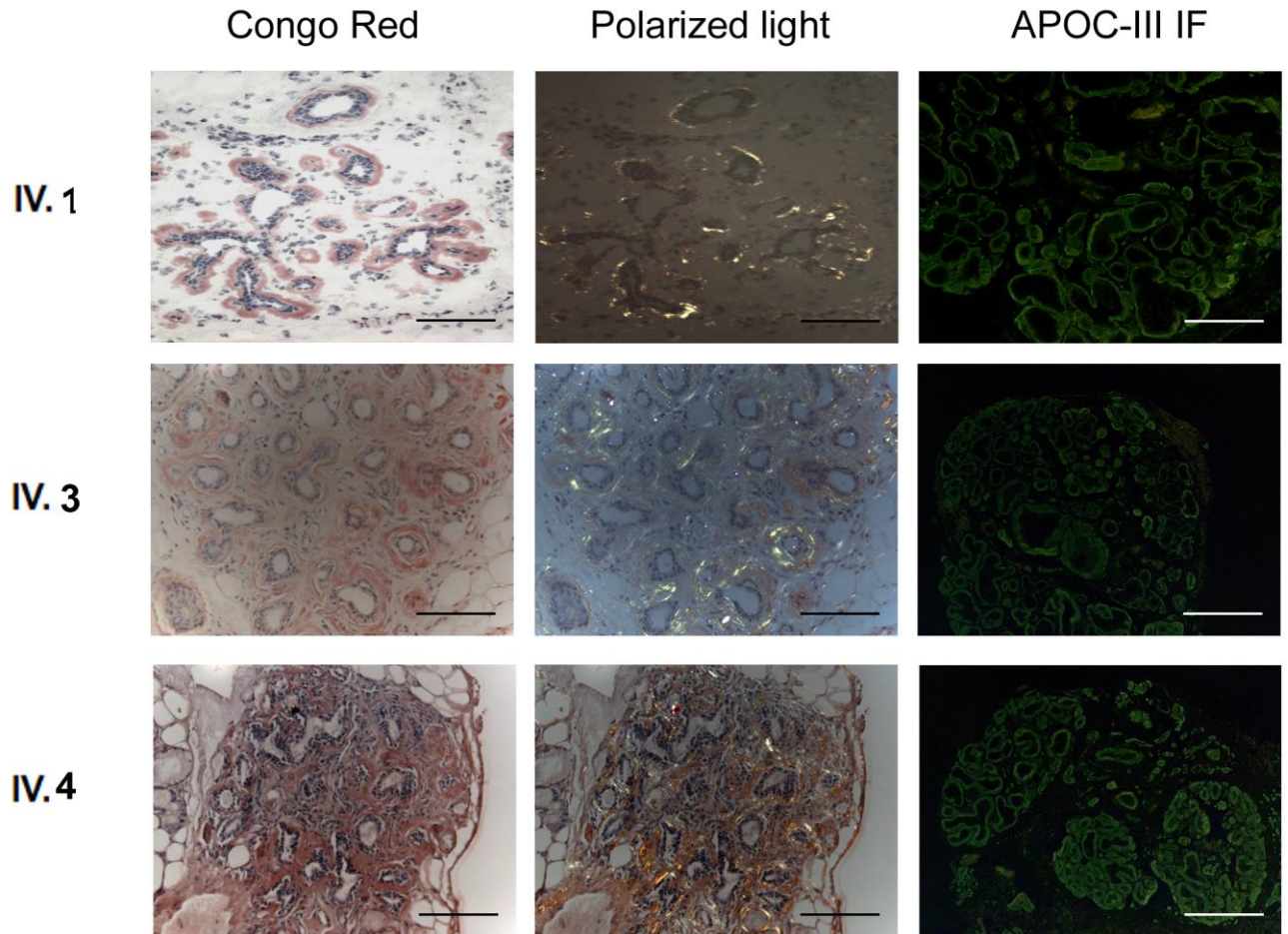
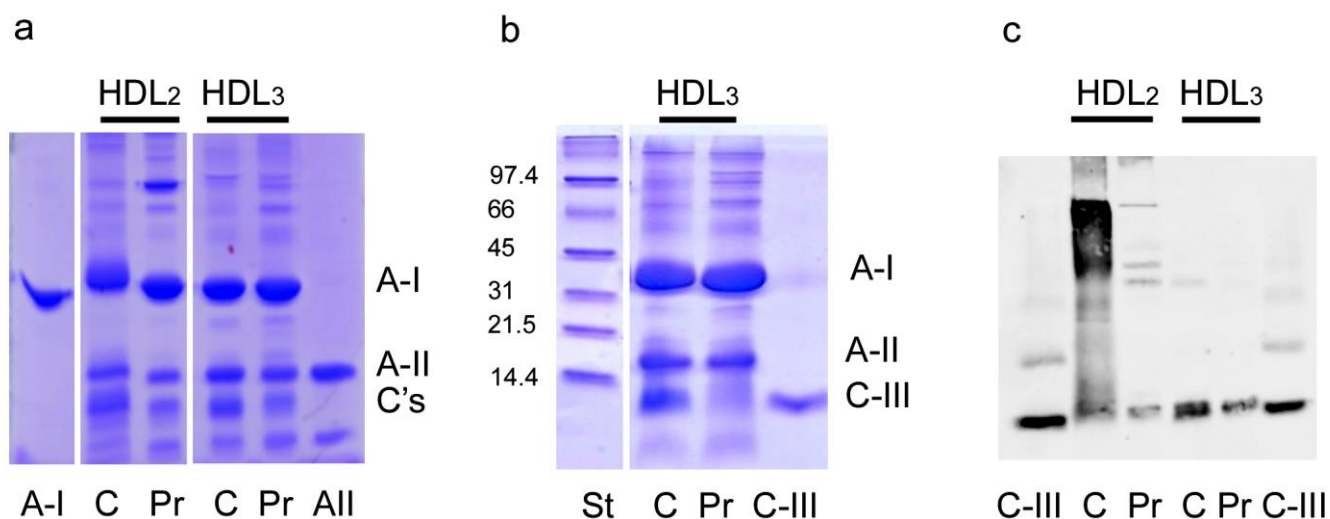


SUPPLEMENTARY INFORMATION

SUPPLEMENTARY FIGURES



Supplementary Fig. 1. Histology of salivary gland tissue. Amyloid deposits in the salivary gland tissue from three additional D25V-carriers IV.1, IV.3 stained with Congo red, exhibited green birefringence when viewed under polarized light and were immunoreactive for apoC-III antibody in all D25V-carriers. Scale bar: 100 μ m.



Supplementary Fig. 2. Analysis of HDL apolipoproteins. (a) SDS 15% PAGE of HDL₂ and HDL₃ from the proband III.3 and the control subject (12 μ g protein per lane) carried out under reducing conditions. In the first and last lane, respectively, purified human apoA-I (28 kDa) and human dimeric apoA-II (17 kDa), the major HDL apolipoproteins, were run as standards. (b) SDS 15% PAGE of HDL₃ from the proband and the control subject (12 μ g protein per lane) was carried out as in (a). Low Range SDS-PAGE Standards (kDa) (LMW BioRad) were run in the first lane, and purified human apoC-III in the last lane as standard (4 μ g per lane). SDS-PAGE for HDL₂ and HDL₃ were replicated 7 times, using as standards either purified apoA-I, apoA-II and apoC-III or LMW. (c) Western blot developed with a primary rabbit anti-human apoC-III antibody (1 μ g ml⁻¹, Abnova). Isolated apoC-III (8.8 kDa, 4 μ g per lane) was loaded in the first and last lanes as standard controls. Western blots for apoC-III were replicated 3 times. A-I, apolipoprotein A-I; A-II, apolipoprotein A-II; C-III, apolipoprotein C-III; C, control subject; Pr, proband III.3; A-II, apolipoprotein A-II; C-III, apolipoprotein C-III.

Supplementary Table 1. Amyloid proteome of salivary gland.

Entry Name (a)	Accession number (b)	MW (kDa)	Sample 1	Sample 2
Serum albumin	ALBU_HUMAN	69	22	20
Collagen alpha-3(VI) chain	CO6A3_HUMAN	34	17	16
Apolipoprotein E	APOE_HUMAN	36	17	13
Keratin, type I cytoskeletal 10	K1C10_HUMAN	58	15	14
Lactotransferrin	TRFL_HUMAN	78	15	14
Apolipoprotein C-III	APOC3_HUMAN	11	13	8
Keratin, type II cytoskeletal 2 epidermal	K22E_HUMAN	65	12	10
Keratin, type II cytoskeletal 1	K2C1_HUMAN	66	11	10
Vitronectin	VTNC_HUMAN	54	10	9
Serum amyloid P-component	SAMP_HUMAN	25	10	9
Vimentin	VIME_HUMAN	54	9	9
Basement membrane-specific heparan sulfate proteoglycan core protein	PGBM_HUMAN	47	8	8
Keratin, type I cytoskeletal 19	K1C19_HUMAN	44	8	8
Hemoglobin subunit beta	HBB_HUMAN	16	8	7
Keratin, type II cytoskeletal 7	K2C7_HUMAN	51	6	6
Apolipoprotein A-IV	APOA4_HUMAN	45	6	6
Keratin, type II cytoskeletal 5	K2C5_HUMAN	62	6	6
Keratin, type I cytoskeletal 14	K1C14_HUMAN	51	5	5
Collagen alpha-2(VI) chain	CO6A2_HUMAN	11	4	4
Actin, cytoplasmic 1	ACTB_HUMAN	42	4	3
Complement component C9	CO9_HUMAN	65	3	2

Amyloid proteome of salivary gland from individual IV.3 detected apoC-III protein along with apoE and SAP.

Supplementary Table 2. Primer sequences for PCR amplification of *APOC3*.

PRIMER SEQUENCES FOR PCR AMPLIFICATION OF THE <i>APOC3</i> GENE			
	Primer name	Primer sequence (5'->3')	Product size (bp)
Promoter region	<i>APOC3_5PF</i>	gaaggtgaacgagagaatcagtcctg	511
	<i>APOC3_5PR</i>	gcctcgggcccatctcagcctttcacactg	
Exon 1	<i>APOC3_1F</i>	caagccacccacttgttctc	600
	<i>APOC3_1R</i>	tccgaggcttccttagctc	
Exon 2	<i>APOC3_2F</i>	cttctggcagaccagctaa	250
	<i>APOC3_2R</i>	gaccaccattgggactg	
Exon 3	<i>APOC3_3F</i>	atgggtggtcaagcagga	296
	<i>APOC3_3R</i>	gagcacctcattccattgt	
Exon 4	<i>APOC3_4F</i>	ctgactggtgtcgtccagtg	288
	<i>APOC3_4R</i>	ccctggagattgcaggac	
3'UTR	<i>APOC3_3PF</i>	ggagcaccgtaaggacaag	562
	<i>APOC3_3PR</i>	ggggcacacactcaagtcac	