

Appendix:

Additional Table: Multivariate analysis of the different clinical and imaging parameters related to the severity of the disease after adjustment using 4 covariates: age, hypertension, other vascular risk factors and location of the NOTCH3 mutation inside or outside the EGFR domains 1 to 6. The results obtained using in the whole fovea are close and nearly overlapping.

		Superficial VD				Deep VD			
		Estimates	Std Error	t value	Pr(> t)	Estimates	Std Error	t value	Pr(> t)
Stroke	(Intercept)	53.4621	2.8662	18.65	0.0000	0.3966	0.0484	8.19	0.0000
	stroke	0.3505	0.6903	0.51	0.6132	0.0149	0.0119	1.25	0.2140
	Age	-0.1248	0.0295	-4.23	0.0001	-0.0006	0.0005	-1.17	0.2452
	<i>HT</i>	0.2770	0.8443	0.33	0.7438	-0.0302	0.0146	-2.07	0.0406
	Other VR factors	0.4062	1.6718	0.24	0.8087	-0.0140	0.0279	-0.50	0.6173
	Location EGFr 1-6	-0.3034	0.7184	-0.42	0.6741	0.0172	0.0128	1.35	0.1814
mRS	(Intercept)	51.0034	3.5682	14.29	0.0000	0.4005	0.0500	8.01	0.0000
	mRS score	0.4112	0.4940	0.83	0.4080	-0.0002	0.0065	-0.04	0.9715
	Age	-0.1018	0.0349	-2.92	0.0047	-0.0005	0.0005	-0.96	0.3399
	<i>HT</i>	0.4884	1.0369	0.47	0.6391	-0.0321	0.0148	-2.16	0.0332
	Other VR factors	2.4347	2.0488	1.19	0.2387	-0.0147	0.0281	-0.52	0.6026
	Location EGFr 1-6	-0.0805	0.8930	-0.09	0.9284	0.0184	0.0128	1.43	0.1556
MMSE	(Intercept)	54.3136	7.8709	6.90	0.0000	44.7517	9.2894	4.82	0.0000
	MMSE score	-0.0381	0.2290	-0.17	0.8685	0.2653	0.2702	0.98	0.3299
	Age	-0.1185	0.0302	-3.93	0.0002	-0.0890	0.0356	-2.50	0.0151
	<i>HT</i>	0.1548	0.9093	0.17	0.8654	0.2382	1.0732	0.22	0.8251
	Other VR factors	0.4408	1.8292	0.24	0.8103	3.4429	2.1588	1.59	0.1158
	Location EGFr 1-6	-0.1566	0.7968	-0.20	0.8449	-1.4157	0.9404	-1.51	0.1372
nb lacunes	(Intercept)	53.4375	2.8943	18.46	0.0000	53.9742	3.5128	15.37	0.0000
	nb lacunes	-0.0010	0.0363	-0.03	0.9776	lacunes	-0.0637	0.0440	-1.45
	Age	-0.1208	0.0285	-4.24	0.0001	-0.0955	0.0346	-2.76	0.0074
	<i>HT</i>	0.2803	0.8463	0.33	0.7415	0.0668	1.0272	0.07	0.9484
	Other VR factors	0.3786	1.6800	0.23	0.8224	2.4688	2.0390	1.21	0.2300
	Location EGFr 1-6	-0.3464	0.7162	-0.48	0.6302	-0.6789	0.8693	-0.78	0.4375
nb microbleeds	(Intercept)	53.4854	2.9023	18.43	0.0000	53.1908	3.5304	15.07	0.0000
	nb microbleeds	-0.0046	0.0095	-0.48	0.6293	-0.0036	0.0115	-0.31	0.7567
	Age	-0.1190	0.0294	-4.06	0.0001	-0.0912	0.0357	-2.55	0.0129
	<i>HT</i>	0.2640	0.8580	0.31	0.7592	0.0740	1.0437	0.07	0.9437
	Other VR factors	0.3029	1.6943	0.18	0.8586	2.7392	2.0610	1.33	0.1883
	Location EGFr 1-6	-0.2888	0.7262	-0.40	0.6921	-0.8171	0.8834	-0.92	0.35
nWMH	(Intercept)	53.7997	2.9761	18.08	0.0000	52.6789	3.6600	14.39	0.0000
	nWMH	-0.0311	0.0668	-0.47	0.6433	0.0540	0.0822	0.66	0.5129
	Age	-0.1235	0.0290	-4.25	0.0001	-0.0903	0.0357	-2.53	0.0137
	<i>HT</i>	0.3469	0.8563	0.41	0.6866	0.0053	1.0531	0.01	0.9960
	Other VR factors	0.2685	1.6895	0.16	0.8742	2.9109	2.0778	1.40	0.1656
	Location EGFr 1-6	-0.4475	0.7448	-0.60	0.5499	-0.5952	0.9160	-0.65	0.5180
BPF	(Intercept)	50.6604	7.7781	6.51	0.0000	47.4650	9.5606	4.96	0.0000
	BPF	BPF	0.0338	0.0884	0.38	BPF	0.0717	0.1087	0.66
	Age	-0.1177	0.0296	-3.98	0.0002	-0.0884	0.0364	-2.43	0.0176
	<i>HT</i>	0.2054	0.8678	0.24	0.8136	-0.0406	1.0666	-0.04	0.9698
	Other VR factors	0.3711	1.6727	0.22	0.8251	2.6886	2.0561	1.31	0.1953
	Location EGFr 1-6	161-6	-0.3945	0.7240	-0.54	-0.8674	0.8899	-0.97	0.3331