

Table 3. Risk factors for anatomical progression of ocular VHL disease during follow-up: logistic regression analyses by eye.

Types of anatomical progression of VHL disease during follow-up						Risk Factors (p-value)						
Progression description	Subtypes of progression	Baseline status	# eyes	Follow-up condition	# eyes	Sex	Genotypic category of germline VHL mutation**	Younger baseline age	Younger age of onset	Fellow eye involved	Ever-smoker	Decreased BMI
New ocular involvement	None to any retinal location (O, P, or O&P)	No ocular VHL disease	262	Presence of ocular VHL disease	58	0.79	0.44	0.10	0.89	0.01*	0.90	0.14
	None to O	No ocular VHL disease	262	Juxtapapillary RCH only	6	0.47	0.79	0.88	0.57	0.75	CNE	0.74
	None to P	No ocular VHL disease	262	Peripheral RCH only	48	0.55	0.29	0.08	0.65	0.009*	0.53	0.05
	None to O&P	No ocular VHL disease	262	Juxtapapillary and peripheral RCH	4	0.93	CNE	0.23	0.73	0.91	CNE	0.53
Increase in affected retinal locations	All progressions to a new retinal location	Juxtapapillary only or Peripheral only involvement	196	Juxtapapillary and peripheral involvement	11	0.69	0.58	0.94	0.71	0.75	0.21	0.24
	O to O&P	Juxtapapillary only involvement	20	Juxtapapillary and peripheral involvement	2	0.77	CNE	0.25	0.25	0.89	CNE	0.37
	P to O&P	Peripheral only involvement	176	Juxtapapillary and peripheral involvement	9	0.69	0.75	0.62	0.41	0.63	0.32	0.12
Increased peripheral number		Peripheral involvement (1-2 RCH(s))	116	Peripheral involvement (≥ 3 RCHs)	34	0.75	CNE	0.10	0.07	0.06	0.99	0.48
Increased peripheral extent		Peripheral involvement (< 1 quadrant)	133	Peripheral involvement (≥ 1 quadrant)	28	0.18	CNE	0.001*	0.007*	0.53	0.48	0.07
New severe involvement		Ocular involvement, not severely affected	208	Severely affected	14	0.27	0.66	0.01*	0.0007*	0.24	0.59	0.84

Key: VHL = von Hippel-Lindau; RCH = retinal capillary hemangioblastoma; O = optic nerve/juxtapapillary RCH; P = peripheral RCH; BMI = body mass index; CNE = could not estimate p-value in logistic regression analysis due to small n in subgroups. ** indicates p-value for adjusted type 3 analysis of effects / Chi-square test for difference among multiple categorical strata