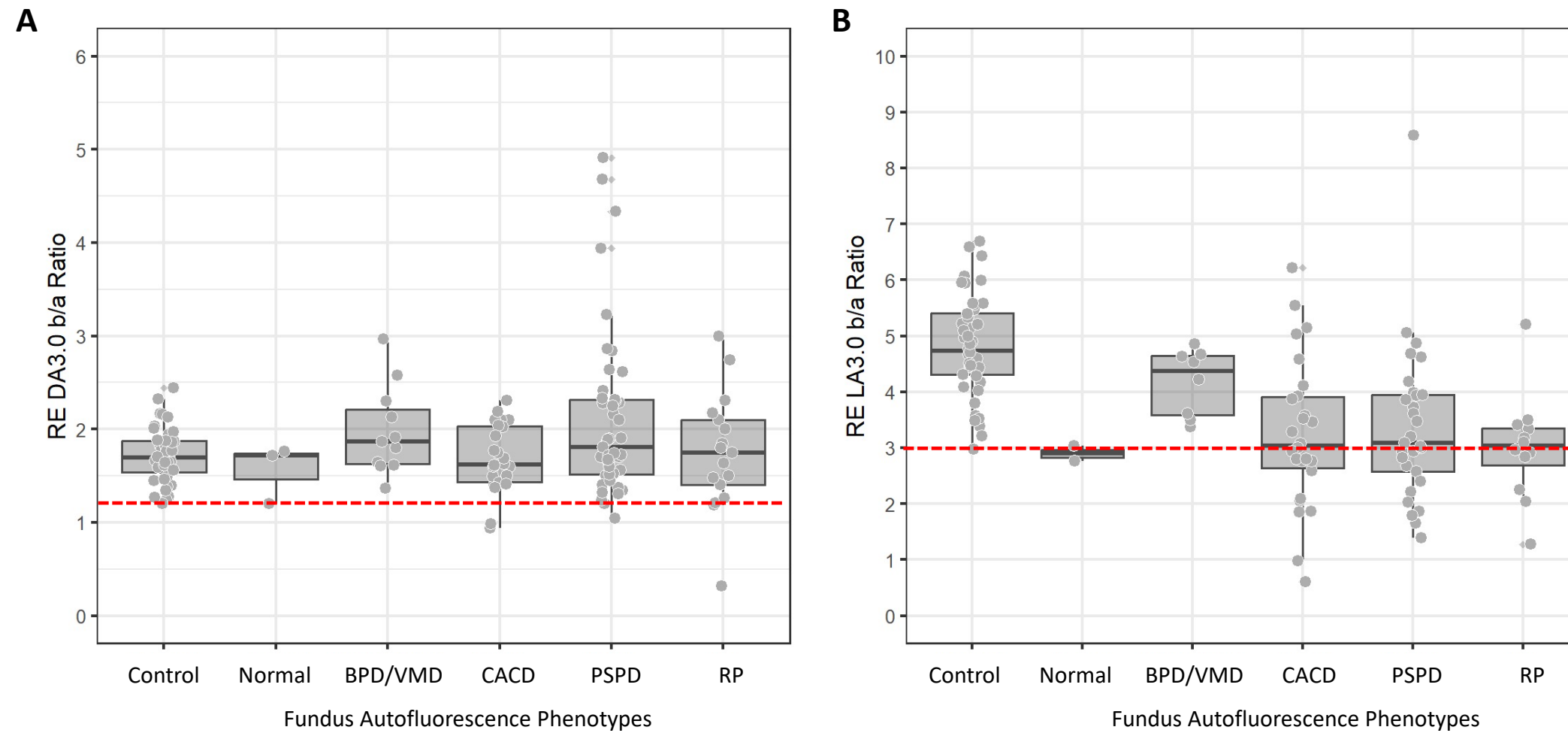


Supplementary Material S3



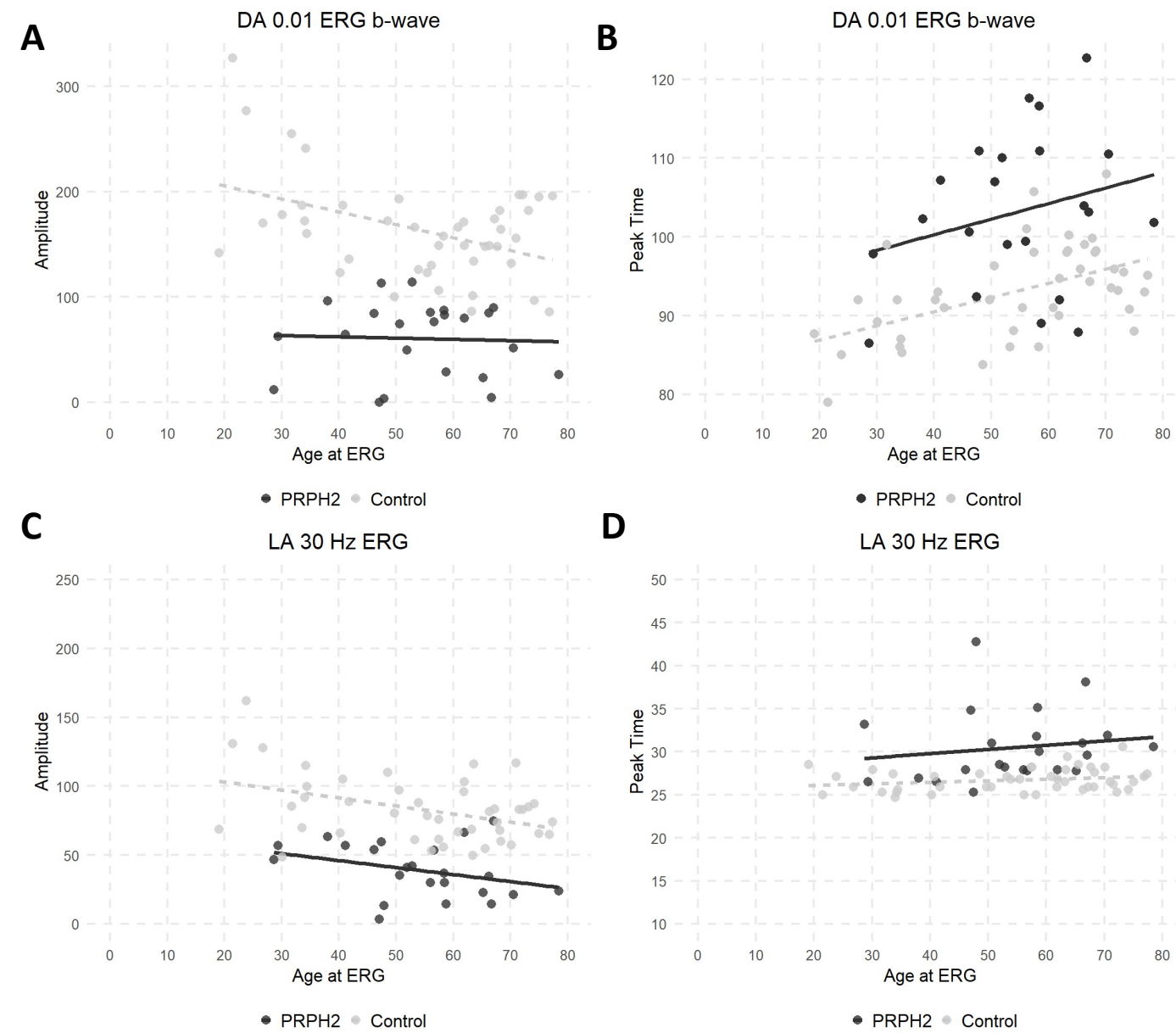
Dark adapted (DA) 3.0 b:a ratio box plot comparing fundus autofluorescence (FAF) phenotypes against healthy subjects showing most have a b:a ratio above 1.2 (red dashed line) (A). Light adapted (LA) 3.0 b:a ratio box plot showing significant reduction in b:a ratio in those with CACD, PSPD and RP (B).

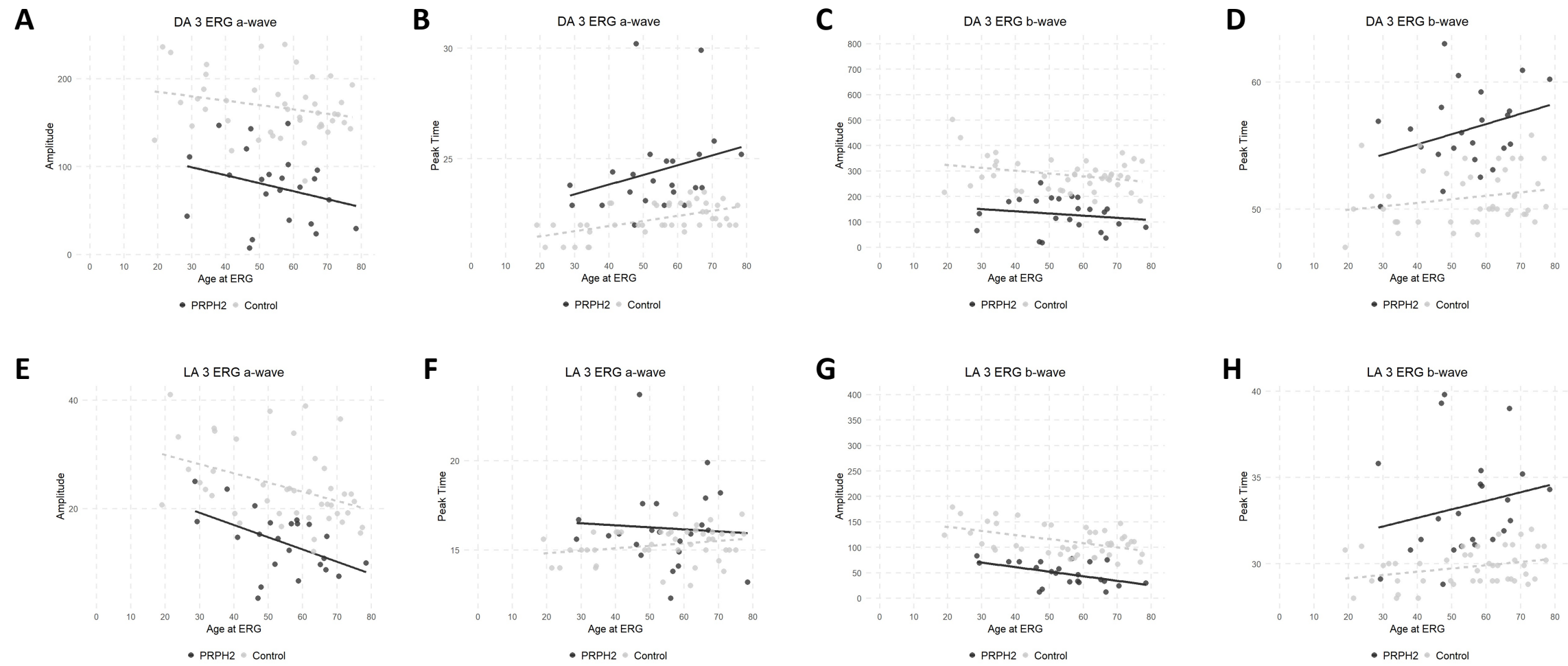
Full-field electrophysiology b-wave amplitude and peak time comparison with normal controls at the Lions Eye Institute, Perth, Australia.

Dark adapted 0.01 (A, B) shows a reduced amplitude and delayed b-wave.

30 Hz flicker (C, D) shows a reduced amplitude and slight delay in peak time.

Overall there is more rod system involvement compared to cone system involvement.





Full-field electrophysiology a- and b-wave comparison with normal controls at the Lions Eye Institute, Perth.

Dark adapted 3.0 (A-D) responses showed a reduction and delay in the a- and b-waves.

Light adapted 3.0 (E-H) responses showed a reduction in the a- and b-wave amplitudes. The b-wave peak time was delayed. There was more significant rod photoreceptor dysfunction.

Slope and significance of the goodness of fit of the regression lines featured in Figures 7 and 8

		PRPH2 - Overall				PRPH2 - LEI				Control			
		N	Slope (SE)	R ²	p-Value	N	Slope (SE)	R ²	p-Value	N	Slope (SE)	R ²	p-Value
DA0.01	Peak Time	81	0.33 (0.10)	0.12	0.002	22	0.20 (0.17)	0.07	0.249	44	0.18 (0.05)	0.26	<0.001
	Amplitude	90	-2.37 (0.78)	0.09	0.003	23	-0.12 (0.61)	0.00	0.848	44	-1.23 (0.40)	0.18	0.004
DA3.0 a-wave	Peak Time	109	0.04 (0.03)	0.01	0.220	23	0.04 (0.03)	0.08	0.198	44	0.02 (0.01)	0.31	<0.001
	Amplitude	118	-1.84 (0.54)	0.09	0.001	23	-0.91 (0.69)	0.08	0.199	44	-0.50 (0.31)	0.06	0.111
DA3.0 b-wave	Peak Time	111	-0.01 (0.05)	0.00	0.850	23	0.08 (0.05)	0.10	0.134	44	0.03 (0.02)	0.04	0.167
	Amplitude	118	-3.79 (0.87)	0.14	<0.001	23	-0.86 (1.10)	0.03	0.443	44	-1.12 (0.56)	0.09	0.053
LA 30 Hz	Peak Time	110	0.05 (0.03)	0.02	0.145	23	0.05 (0.07)	0.02	0.497	44	0.02 (0.01)	0.05	0.128
	Amplitude	115	-0.97 (0.23)	0.14	<0.001	23	-0.51 (0.32)	0.11	0.124	44	-0.58 (0.20)	0.16	0.007
LA3.0 a-wave	Peak Time	85	0.00 (0.03)	0.00	0.934	23	-0.01 (0.04)	0.08	0.781	44	0.01 (0.01)	0.06	0.105
	Amplitude	93	-0.62 (0.12)	0.22	<0.001	23	-0.23 (0.08)	0.25	0.014	44	-0.17 (0.06)	0.16	0.007
LA3.0 b-wave	Peak Time	87	0.01 (0.03)	0.00	0.785	23	0.05 (0.05)	0.04	0.348	44	0.02 (0.01)	0.09	0.044
	Amplitude	93	-1.28 (0.45)	0.08	0.005	23	-0.90 (0.34)	0.25	0.016	44	-0.80 (0.23)	0.23	0.001

SE = Standard Error

ERG tracings and FAF imaging demonstrating the variable effect of CACD phenotype on macular, cone and rod functions

