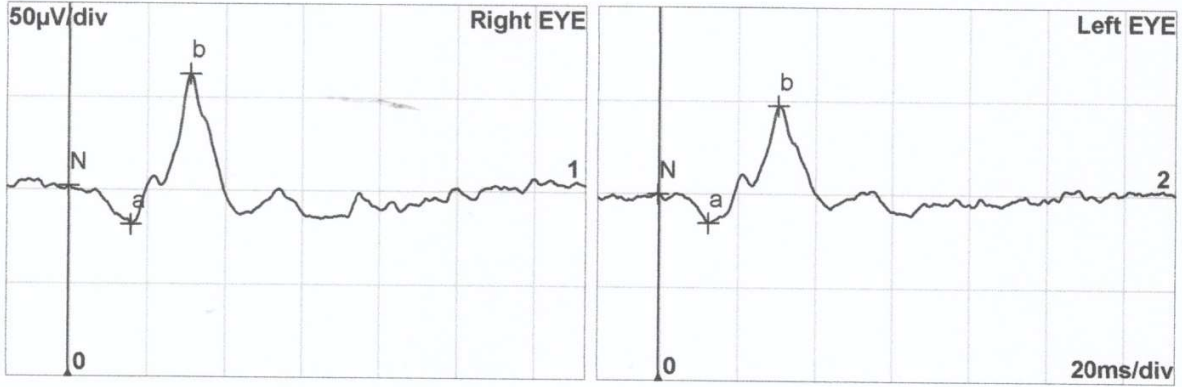


SUPPLEMENTARY FIGURES II.
Electrophysiology

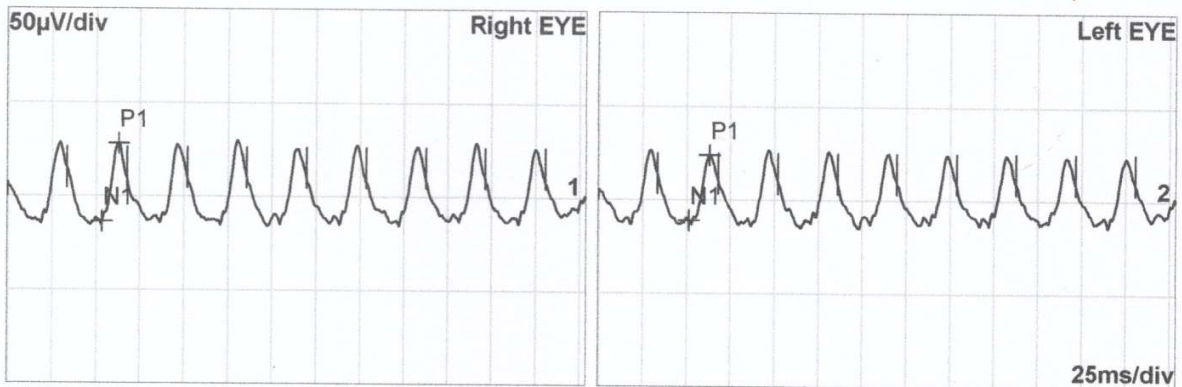
photopic 3.0



Channel	a [ms]	b [ms]	a-wave	b-wave
1 R-1	16	31	20,6µV	81µV
2 L-2	13	31	15,5µV	63,2µV

Normals - -

30Hz Flicker 3.0



Channel	N1 [ms]	P1 [ms]	N1-P1
1 R-1	54	64	41,5µV
2 L-2	52	64	35µV

Figure S22. Standard full-field ERG recording of patient III/3.

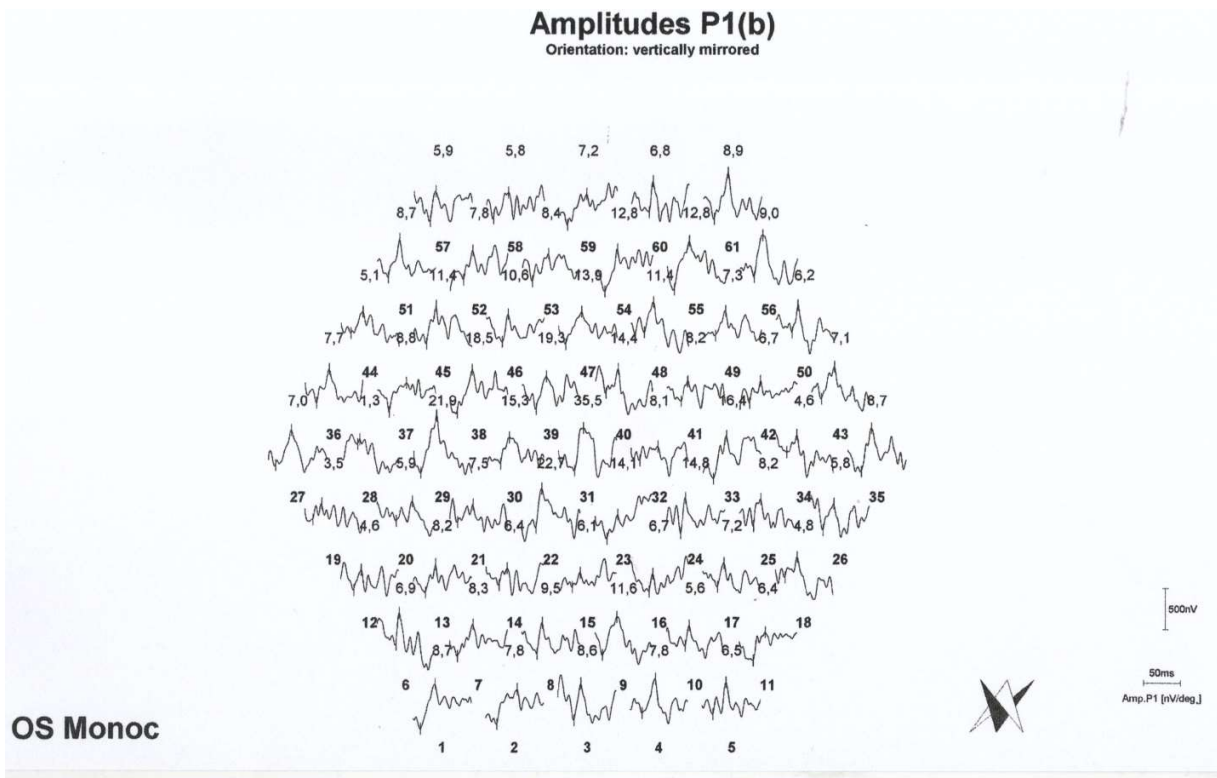


Figure S23. mfERG recording of patient III/3, OS.

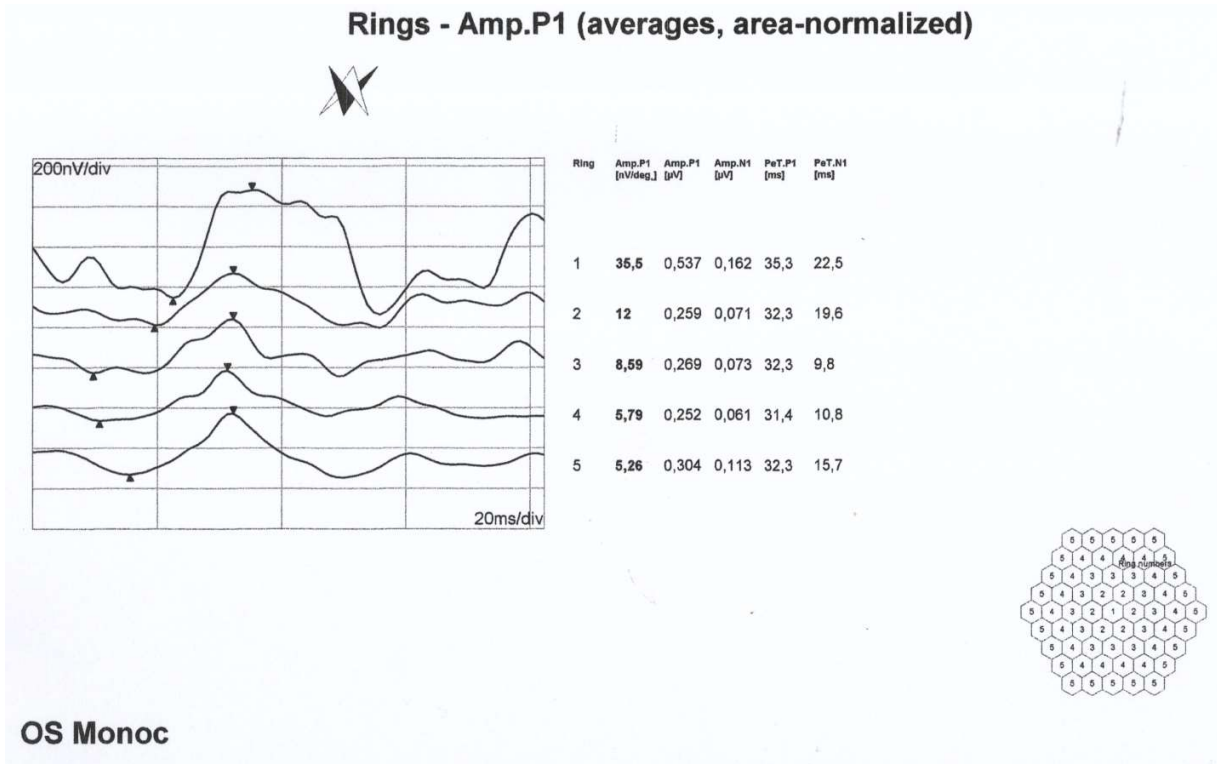
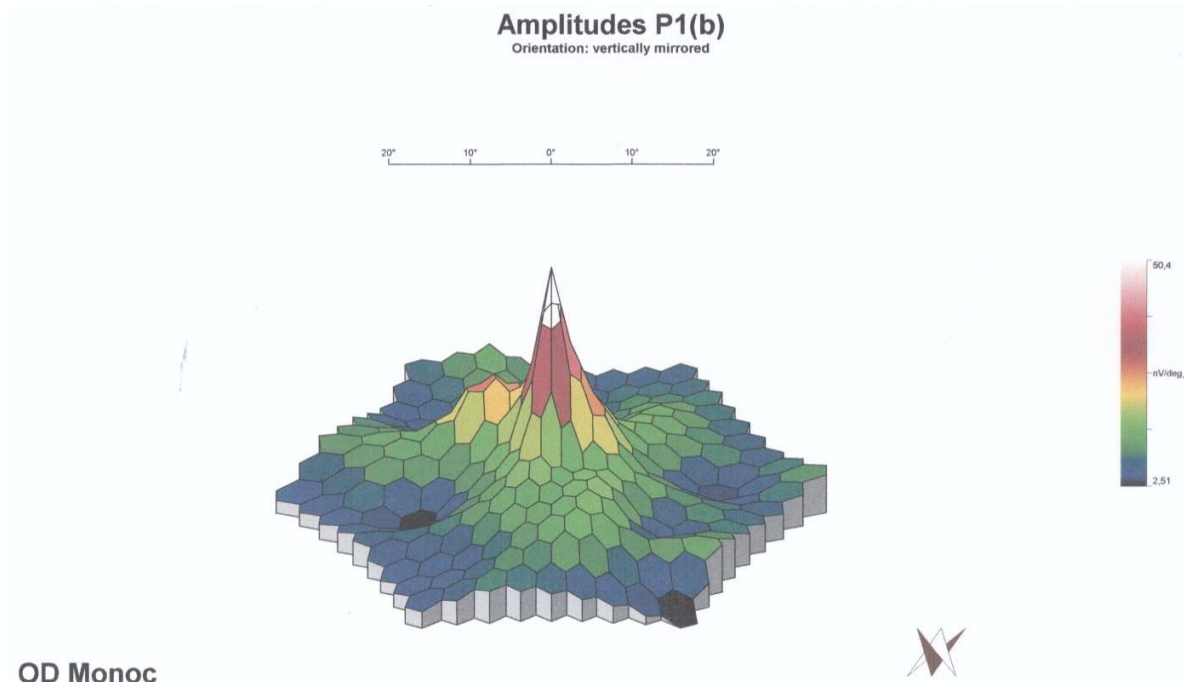
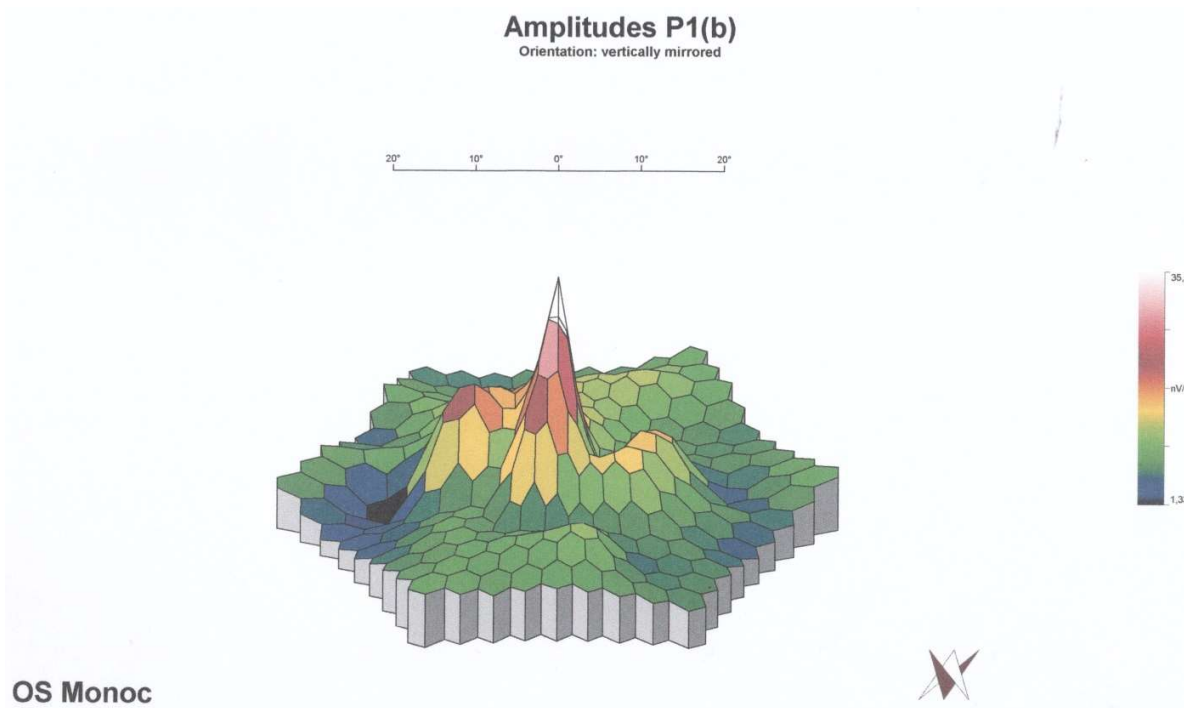


Figure S24. mfERG ring analysis of patient III/3, OS.



OD Monoc

Figure S25. Multifocal ERG 3 D colour map of III/3 carrier male right eye displays overall normal central macular function with scarce subnormal responses only.



OS Monoc

Figure S26. Multifocal ERG 3 D colour map of III/3 carrier male, left eye displays overall normal central macular function with scarce subnormal responses only.

Pattern-ERG

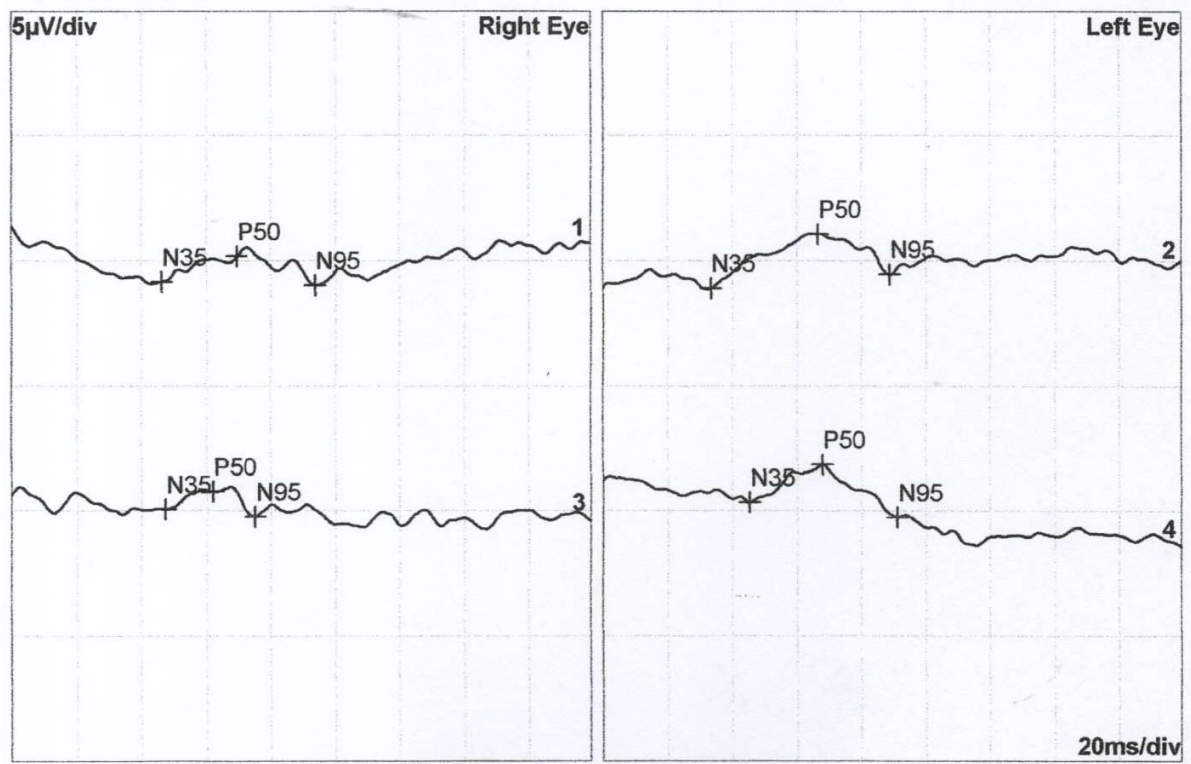


Figure S27. PERG recording of patient III/8. Lines 1 and 3 and lines 2 and 4 represent pairs of replicate measurements.

Pattern-VEP

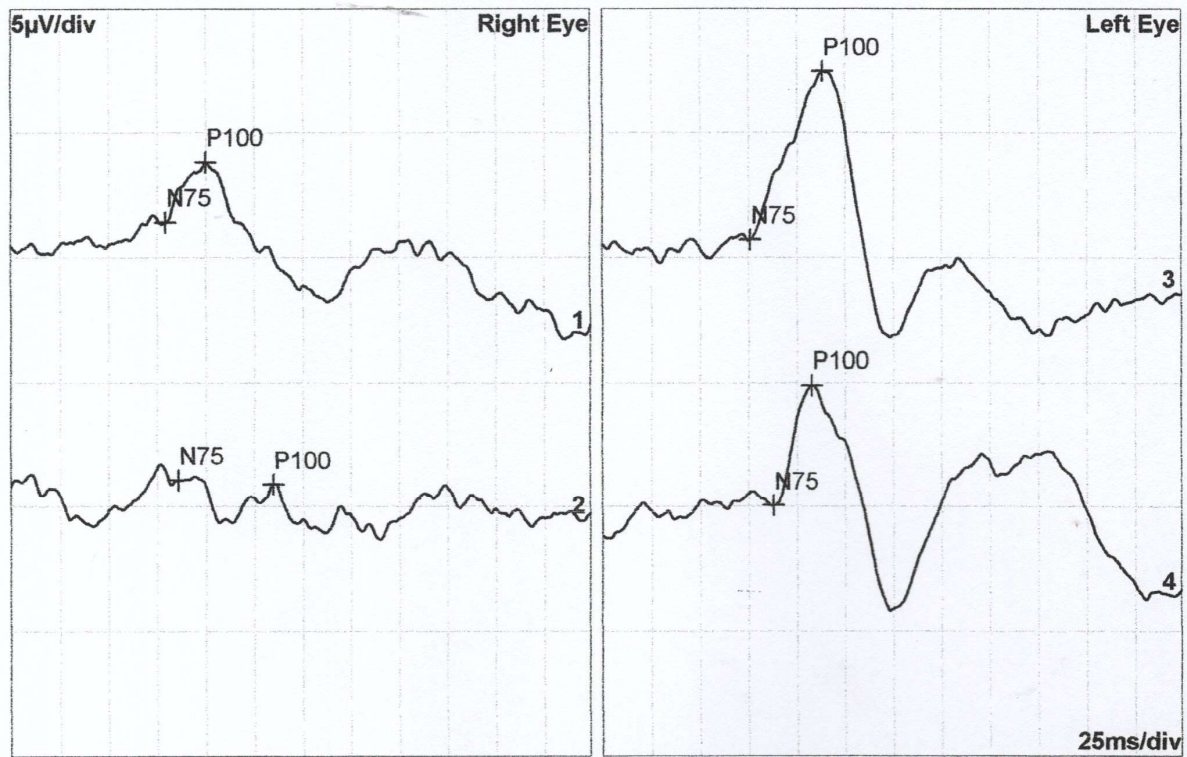
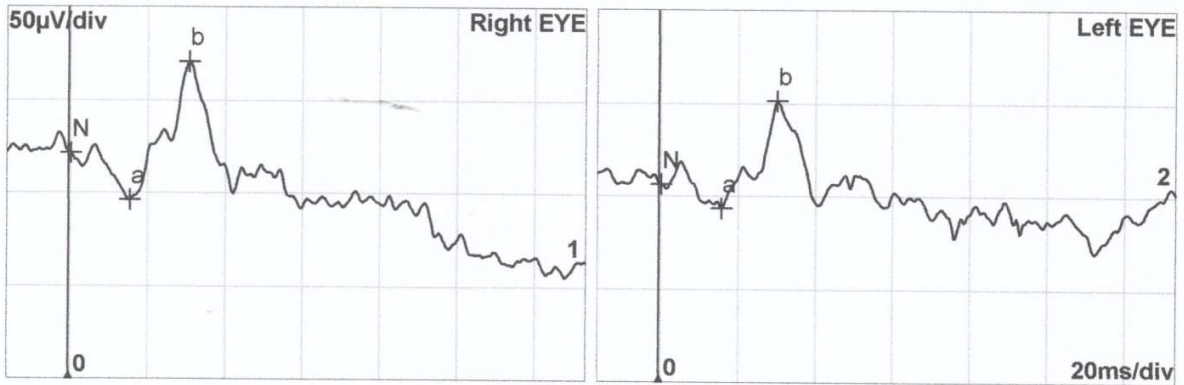


Figure S28. pVEP recording of patient III/8. Lines 1 and 3 display responses to 60' stimuli and lines 2 and 4 represent responses to 15' stimuli.

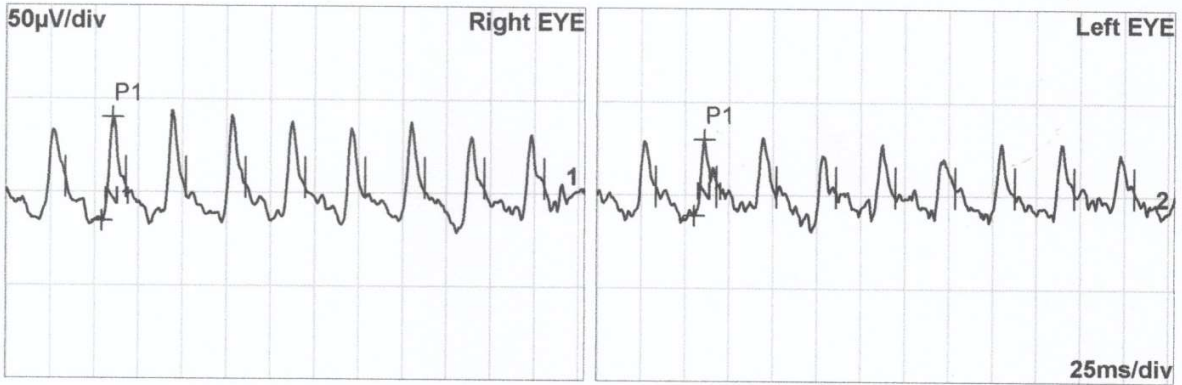
photopic 3.0



Channel	a [ms]	b [ms]	a-wave	b-wave
1 R-1	16	31	25,6µV	74,5µV
2 L-2	16	31	13µV	58,1µV

Normals - -

30Hz Flicker 3.0



Channel	N1 [ms]	P1 [ms]	N1-P1
1 R-1	55	61	55,6µV
2 L-2	56	62	40,9µV

Figure S29. Standard full-field ERG recording of patient IV/1.

Pattern-ERG

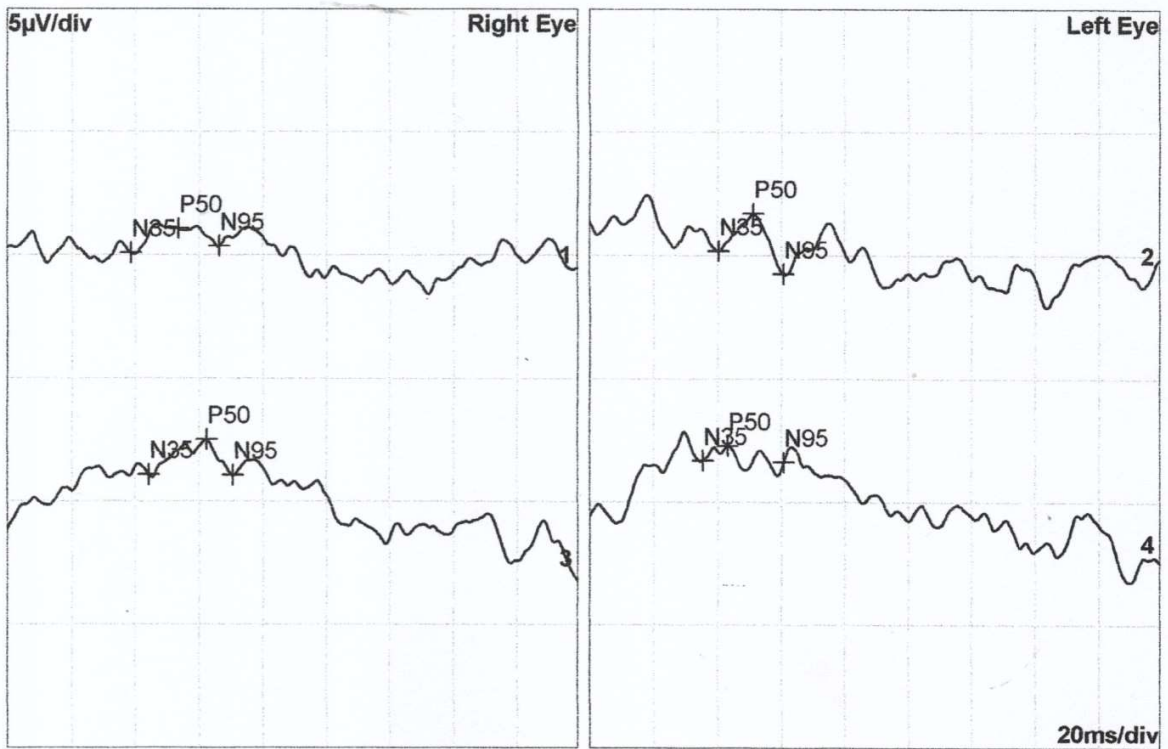


Figure S30. PERG recording of patient IV/1. Lines 1 and 3 and lines 2 and 4 represent pairs of replicate measurements.

Pattern-VEP

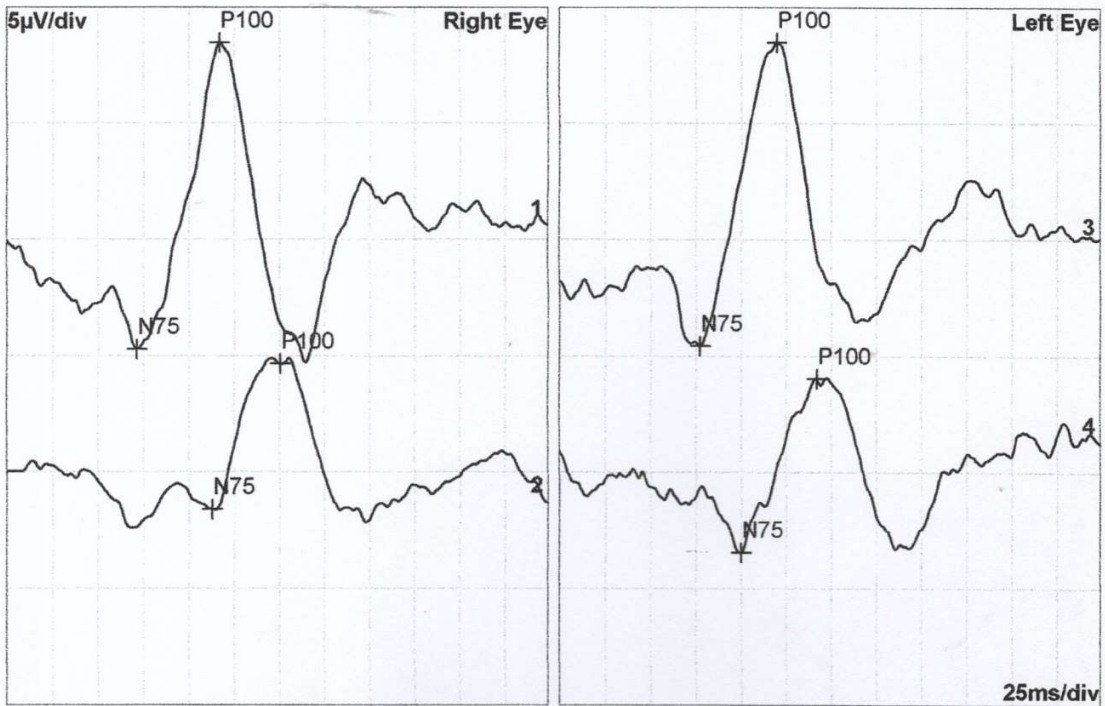


Figure S31. pVEP recording of patient IV/1. Lines 1 and 3 display responses to 60' stimuli and lines 2 and 4 represent responses to 15' stimuli.

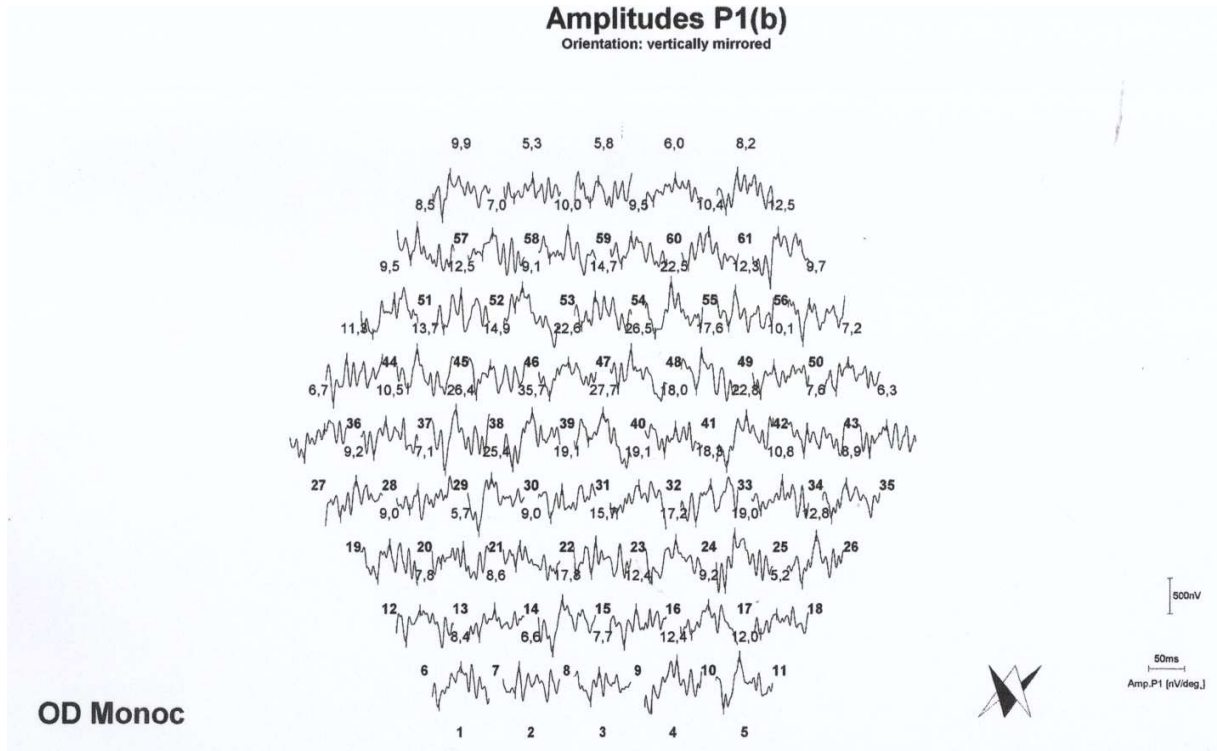


Figure S32. mfERG recording of patient IV/1, OD.

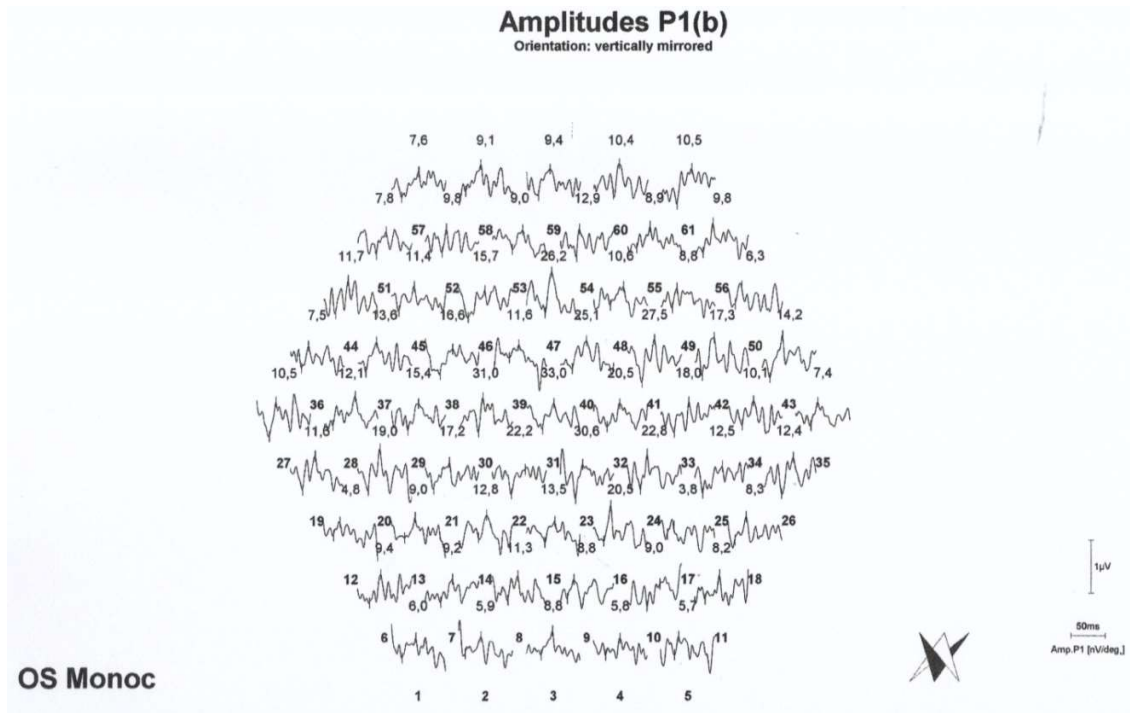


Figure S33. mfERG recording of patient IV/1,, OS.

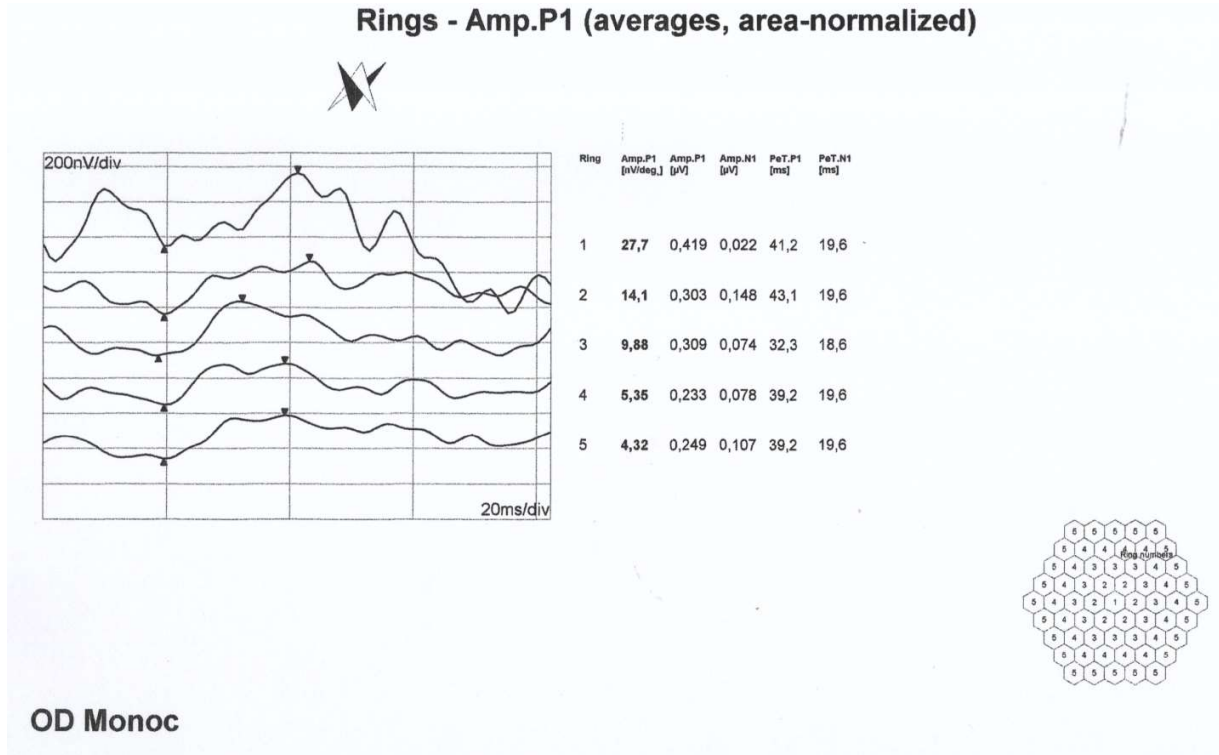
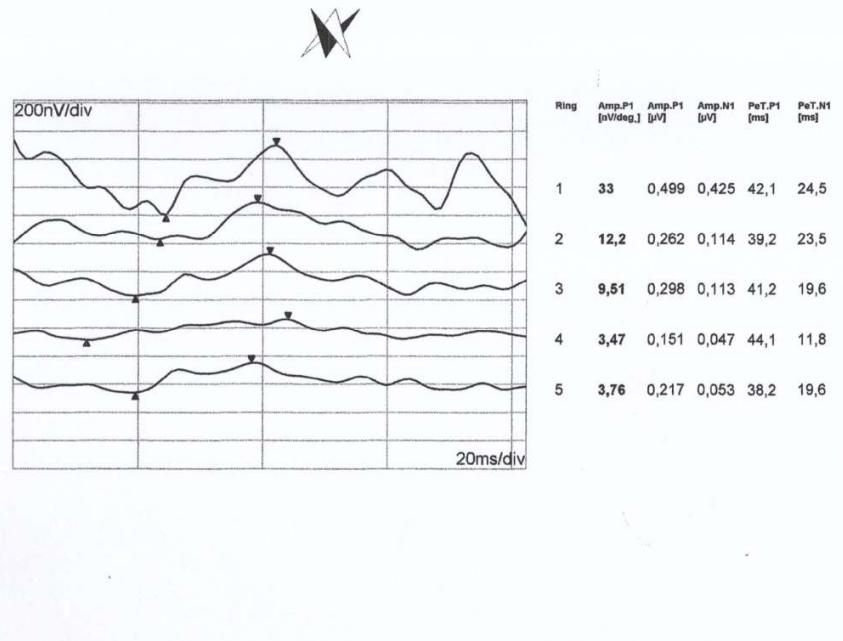


Figure S34. mfERG ring analysis of patient IV/1, OD.

Rings - Amp.P1 (averages, area-normalized)

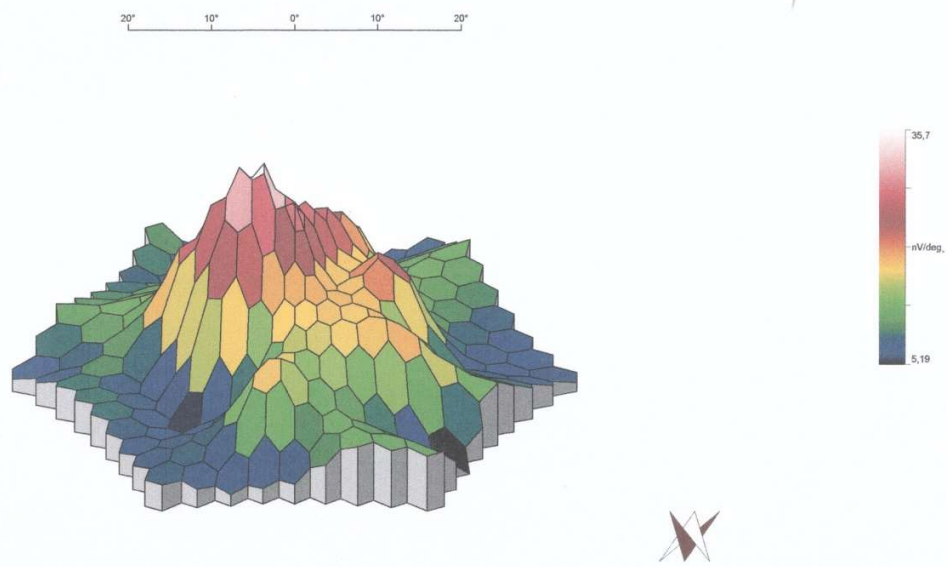


OS Monoc

Figure S35. mfERG ring analysis of patient IV/1, OS.

Amplitudes P1(b)

Orientation: vertically mirrored



OD Monoc

Figure S36. Multifocal ERG 3 D colour map of patient IV/1, OD

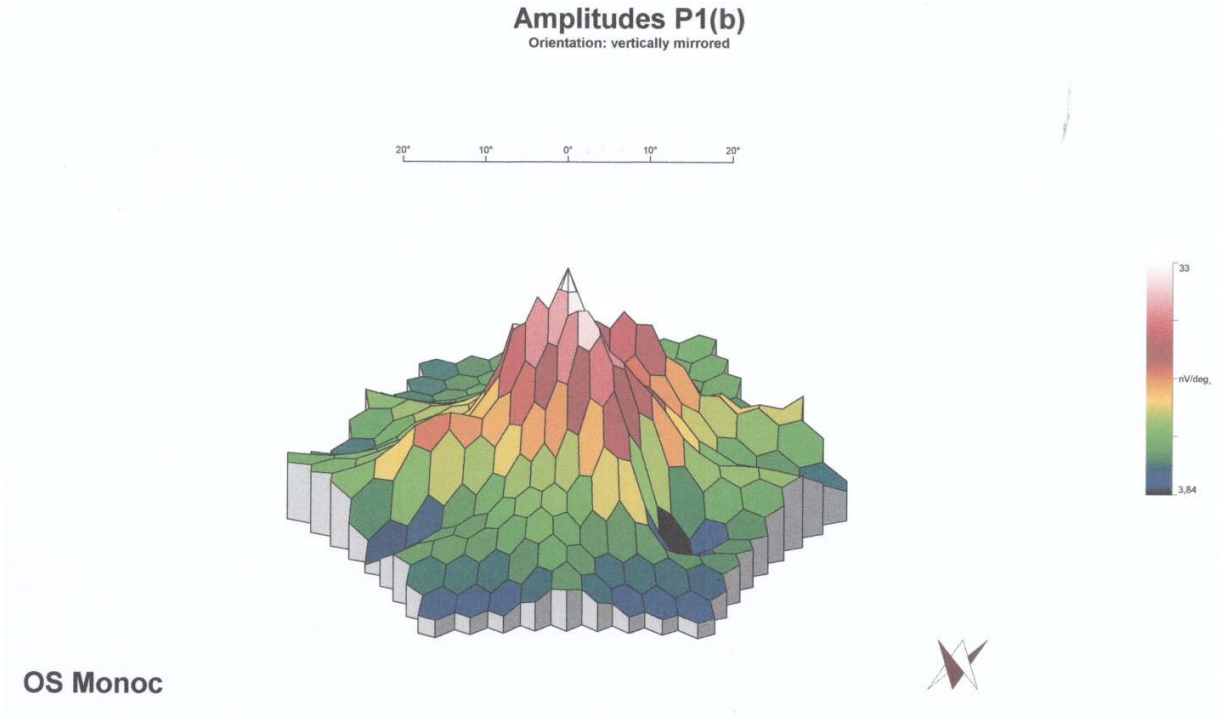
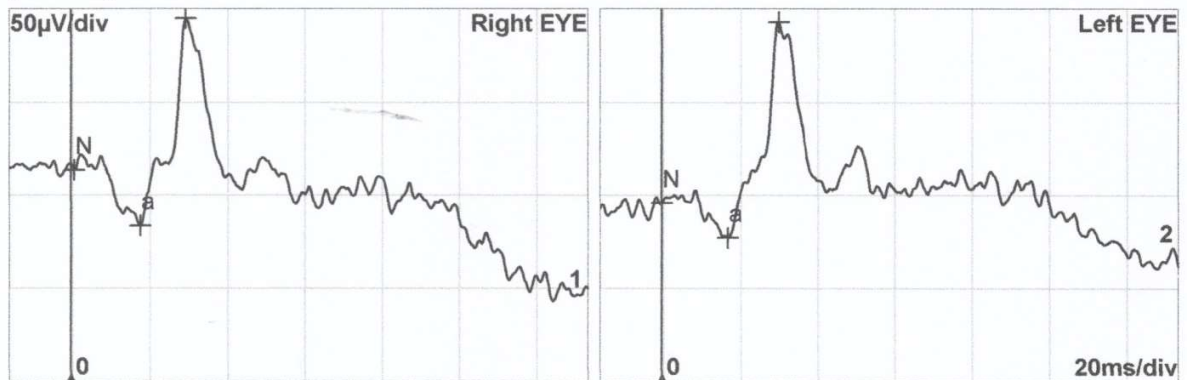


Figure S37. Multifocal ERG 3 D colour map of patient IV/1, OS

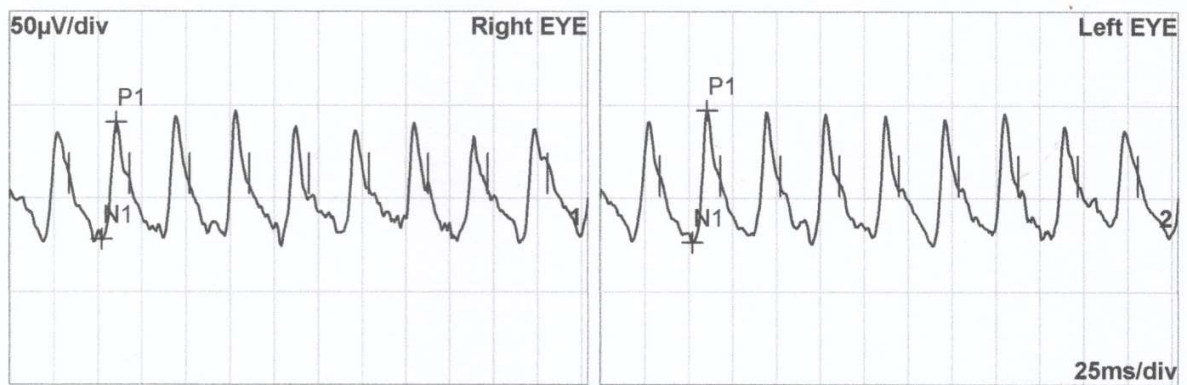
photopic 3.0



Channel	a [ms]	b [ms]	a-wave	b-wave
1 R-1	18	30	30µV	111µV
2 L-2	17	30	18,7µV	116µV

Normals - -

30Hz Flicker 3.0



Channel	N1 [ms]	P1 [ms]	N1-P1
1 R-1	53	61	63,1µV
2 L-2	53	61	70,7µV

Figure S38. Standard full-field ERG recording of patient IV/2.

Pattern-ERG

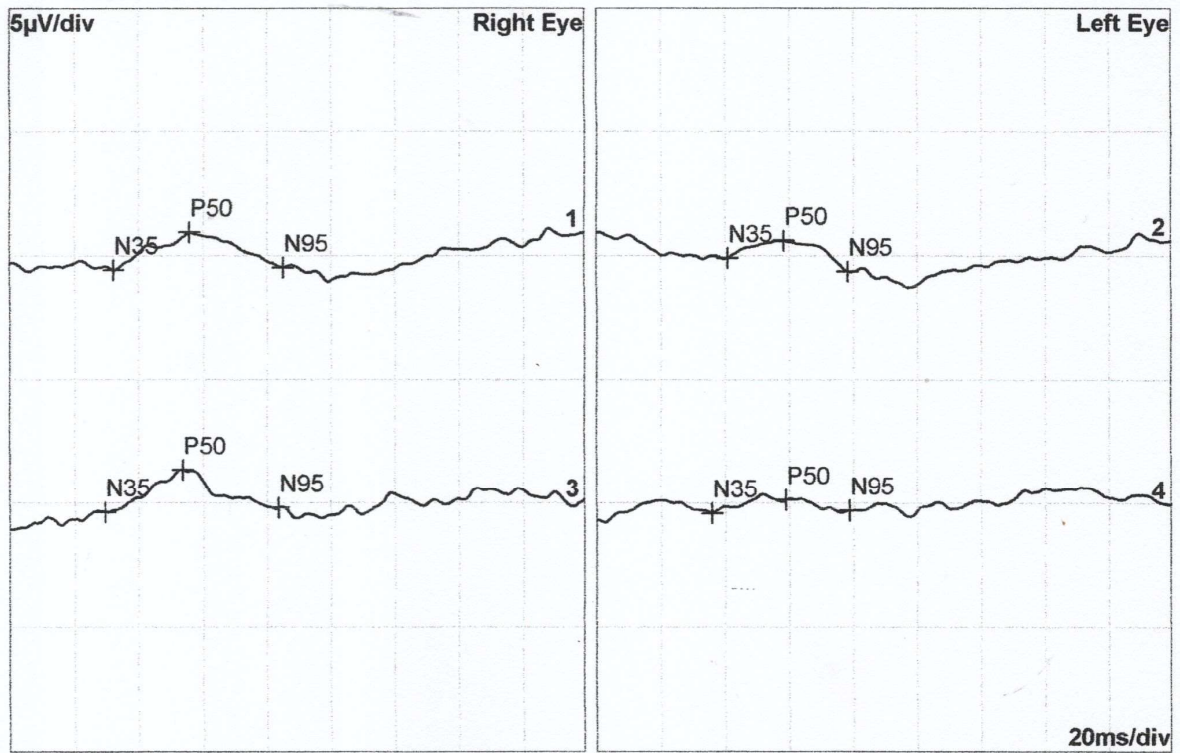


Figure S39. PERG recording of patient IV/2. Lines 1 and 3 and lines 2 and 4 represent pairs of replicate measurements.

Pattern-VEP

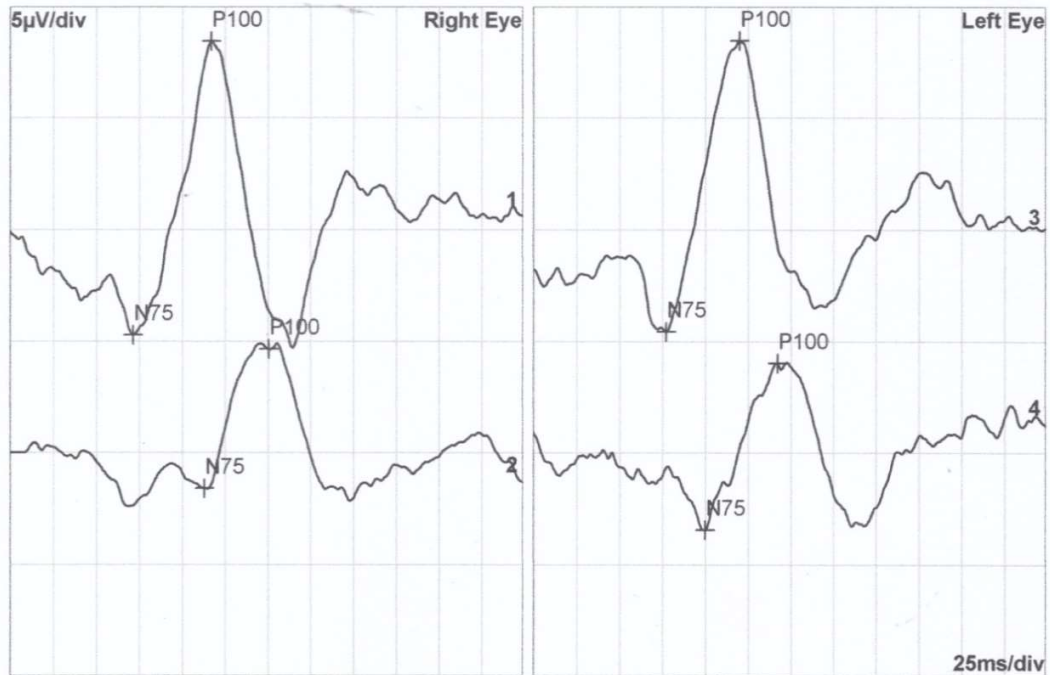


Figure S40. pVEP recording of patient IV/2. Lines 1 and 3 display responses to 60' stimuli and lines 2 and 4 represent responses to 15' stimuli.

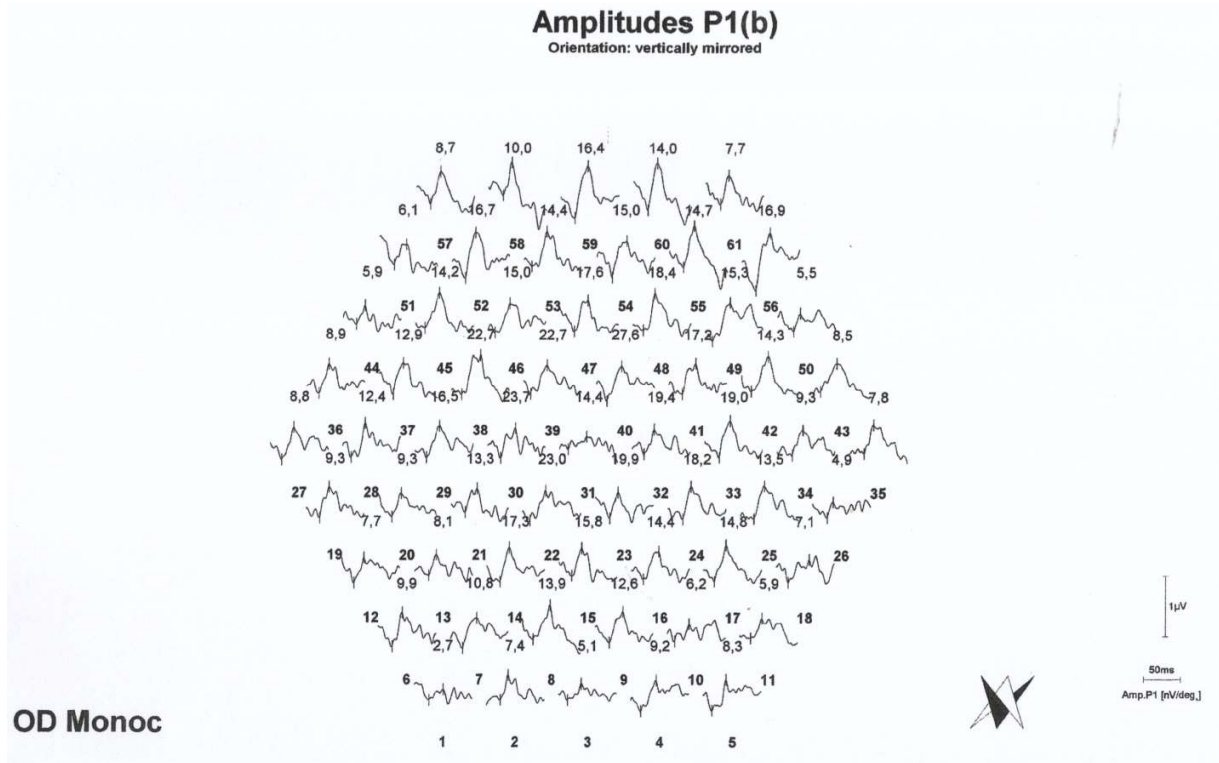


Figure S41. mfERG recording of patient IV/2, OD.

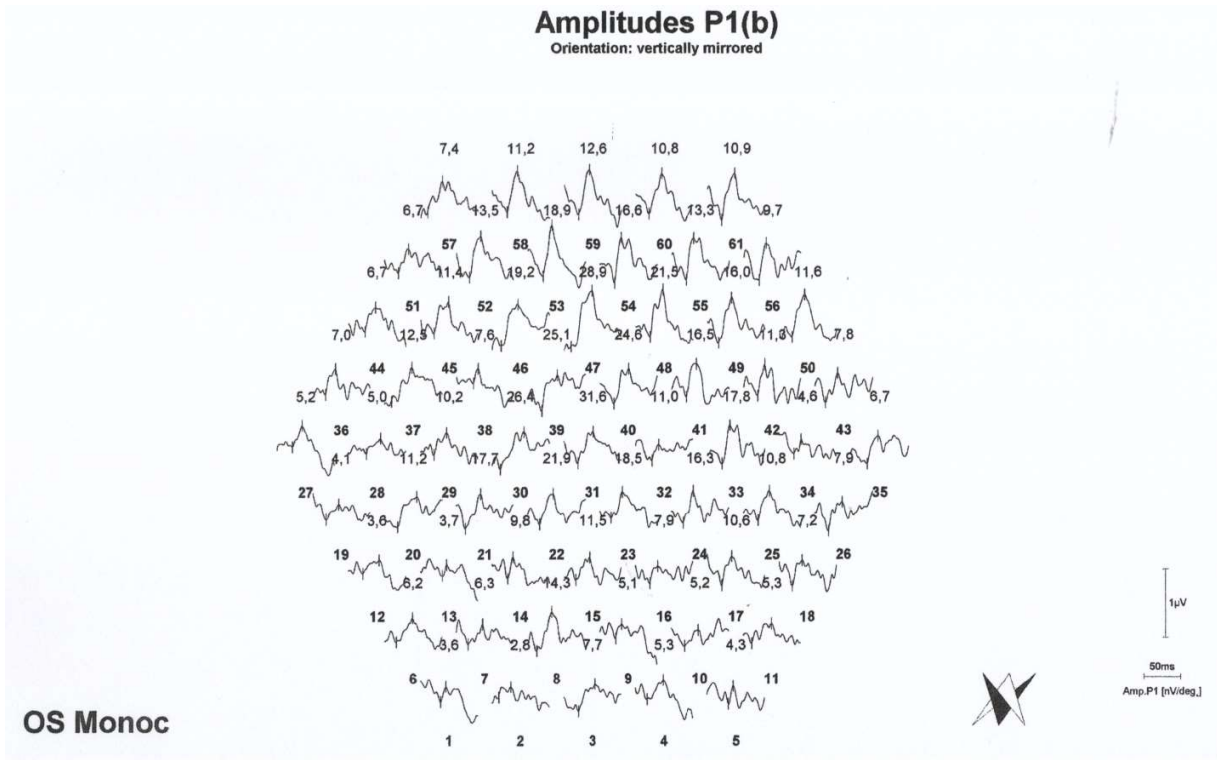


Figure S42. mfERG recording of patient IV/2, OS.

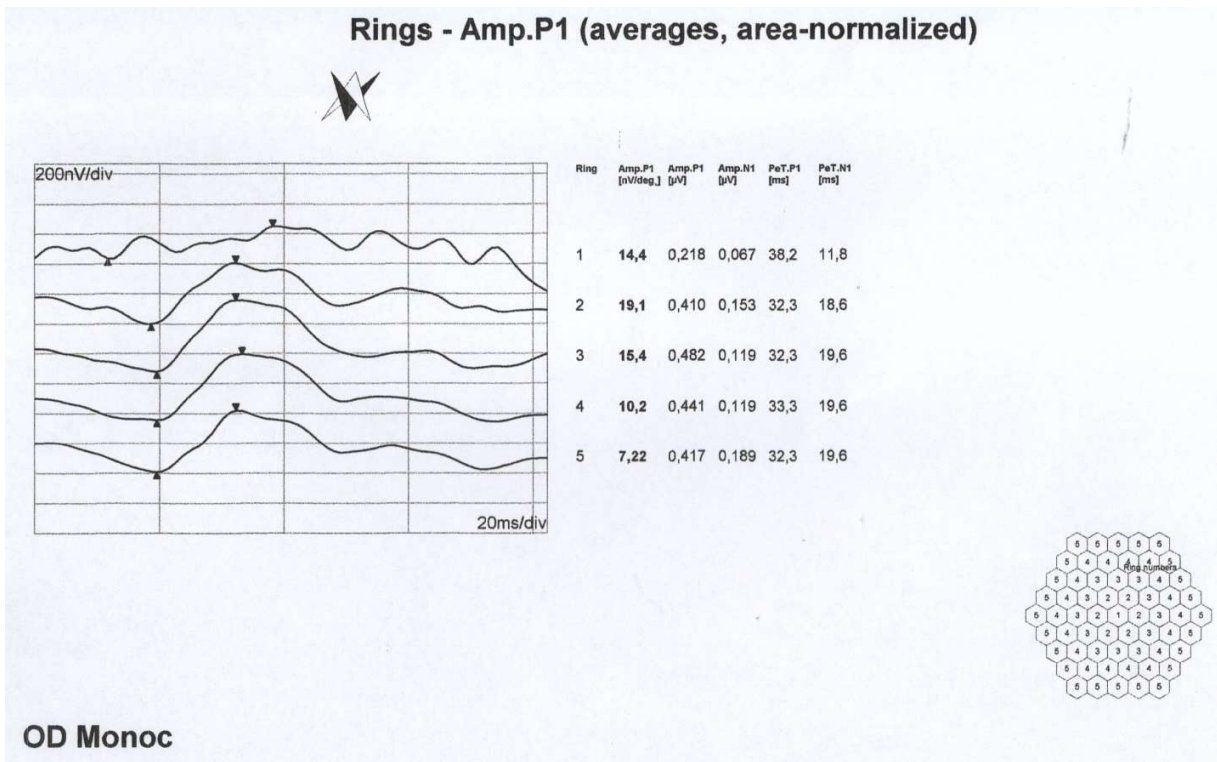
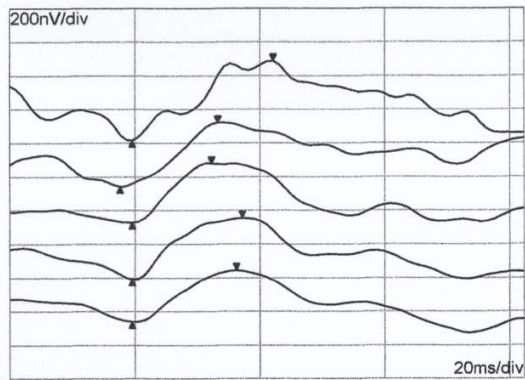


Figure S43. mfERG ring analysis of patient IV/2, OD.

Rings - Amp.P1 (averages, area-normalized)



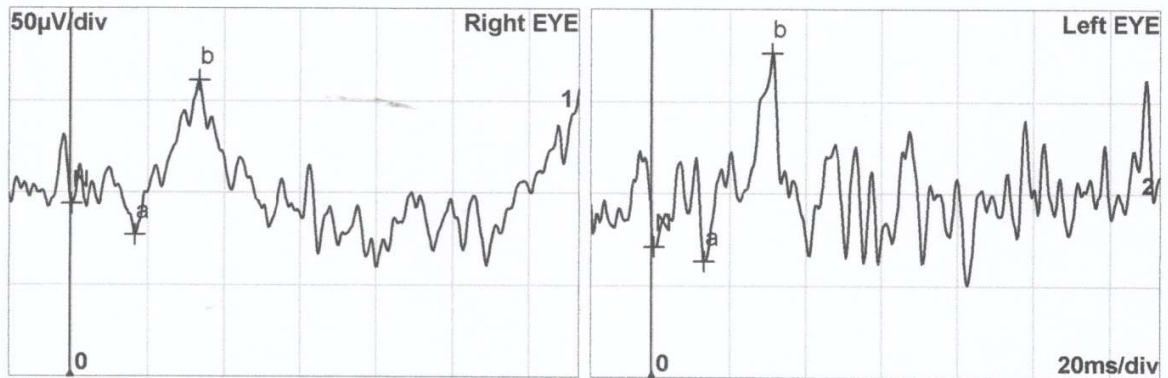
Ring	Amp.P1 [nVdeg.]	Amp.P1 [µV]	Amp.N1 [µV]	PeT.P1 [ms]	PeT.N1 [ms]
1	31,6	0,478	0,174	42,1	19,6
2	17,6	0,379	0,167	33,3	17,6
3	11,3	0,354	0,069	32,3	19,6
4	8,41	0,366	0,167	37,2	19,6
5	5,34	0,308	0,123	36,3	19,6



OS Monoc

Figure S44. mfERG ring analysis of patient IV/2, OS.

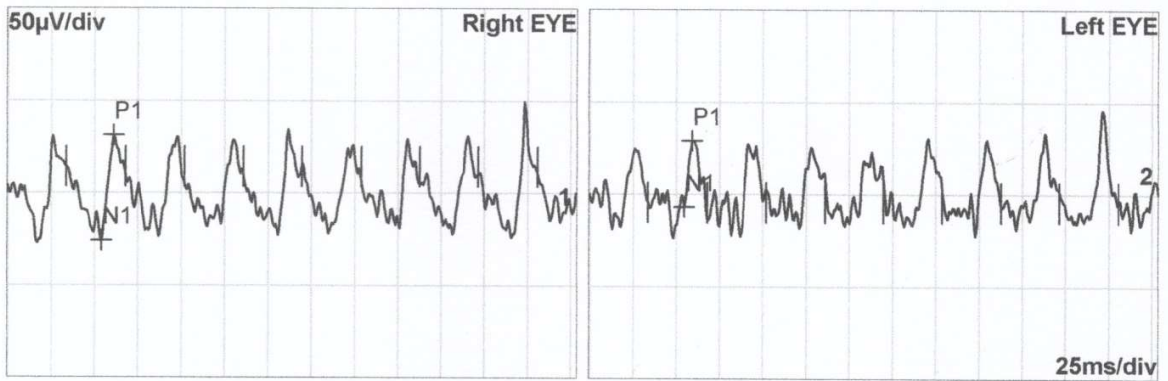
photopic 3.0



Channel	a [ms]	b [ms]	a-wave	b-wave
1 R-1	17	34	16,6µV	83,9µV
2 L-2	14	32	7,71µV	113µV

Normals - -

30Hz Flicker 3.0



Channel	N1 [ms]	P1 [ms]	N1-P1
1 R-1	55	62	57µV
2 L-2	56	60	35,8µV

Figure S45. Standard full-field ERG recording of patient IV/6.

Pattern-ERG

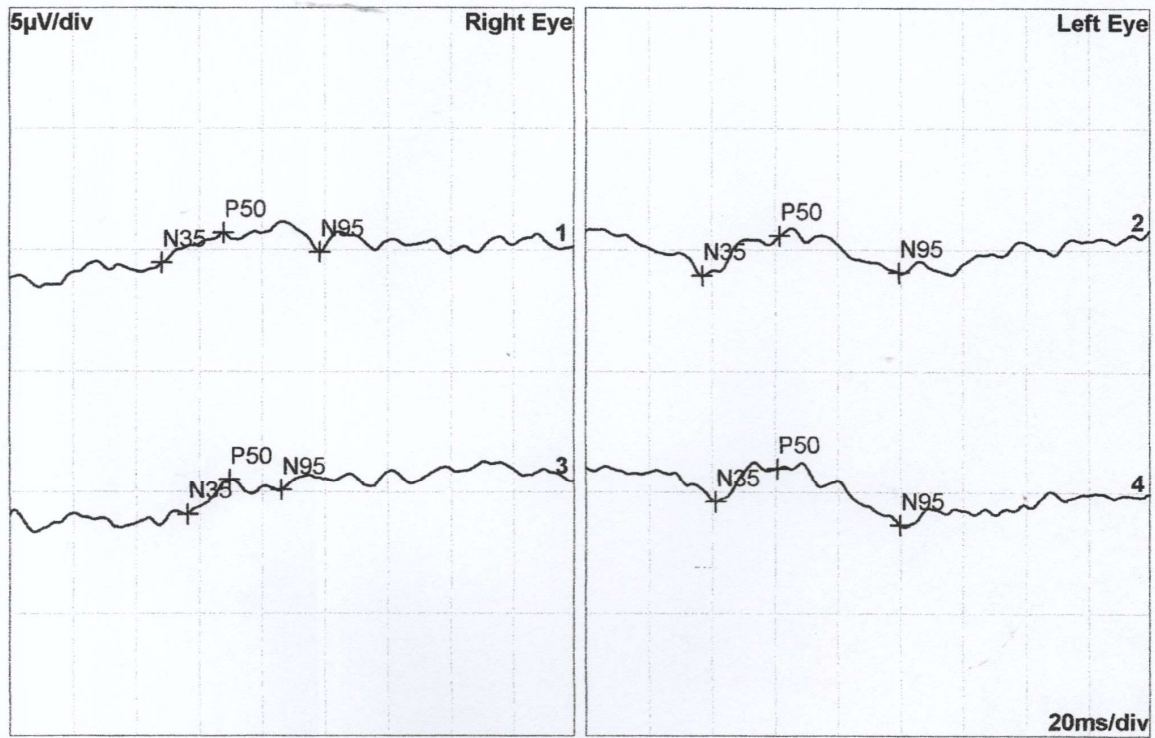


Figure S46. PERG recording of patient IV/6. Lines 1 and 3 and lines 2 and 4 represent pairs of replicate measurements.

Pattern-VEP

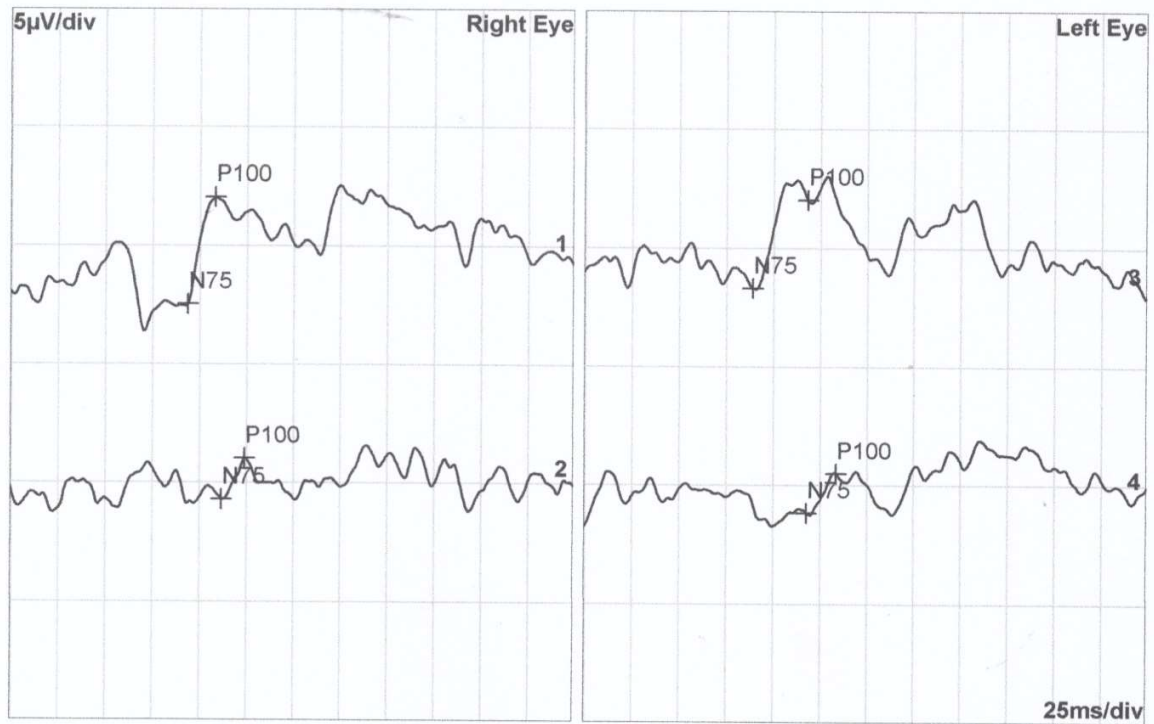


Figure S47. pVEP recording of patient IV/6. Lines 1 and 3 display responses to 60' stimuli and lines 2 and 4 represent responses to 15' stimuli.

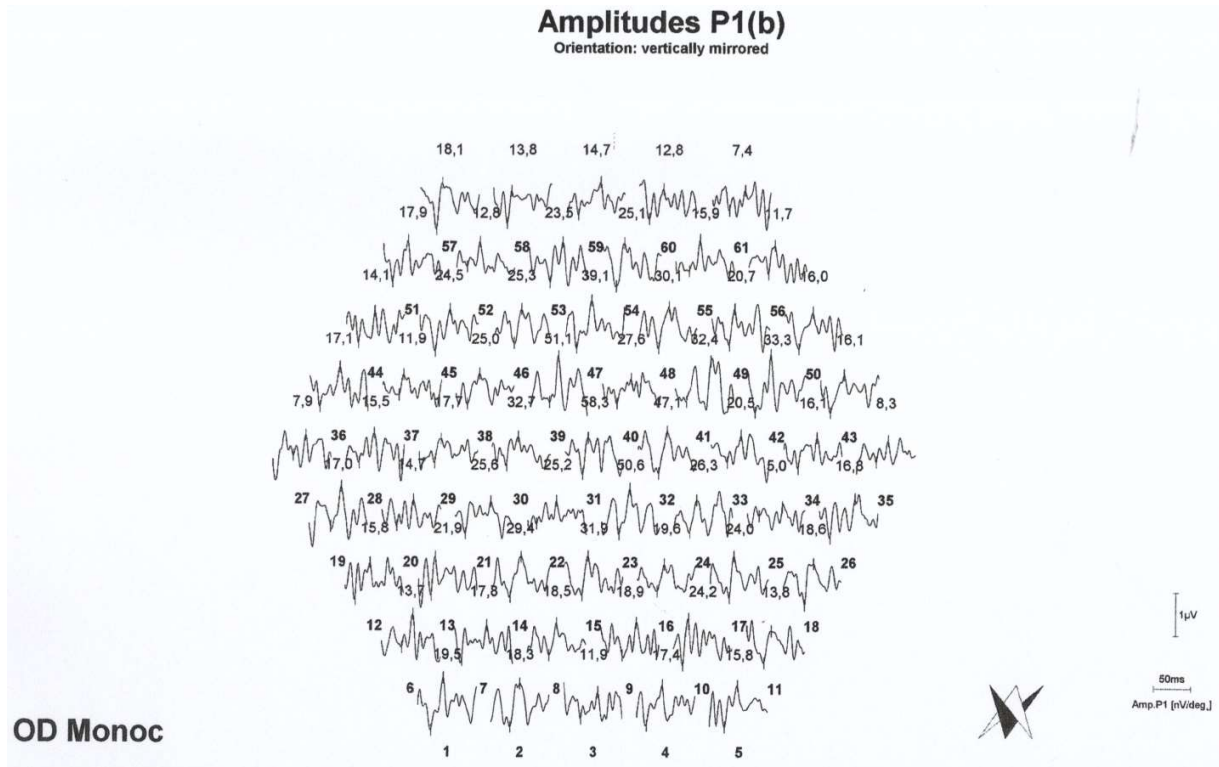


Figure S48. mfERG recording of patient IV/6, OD.

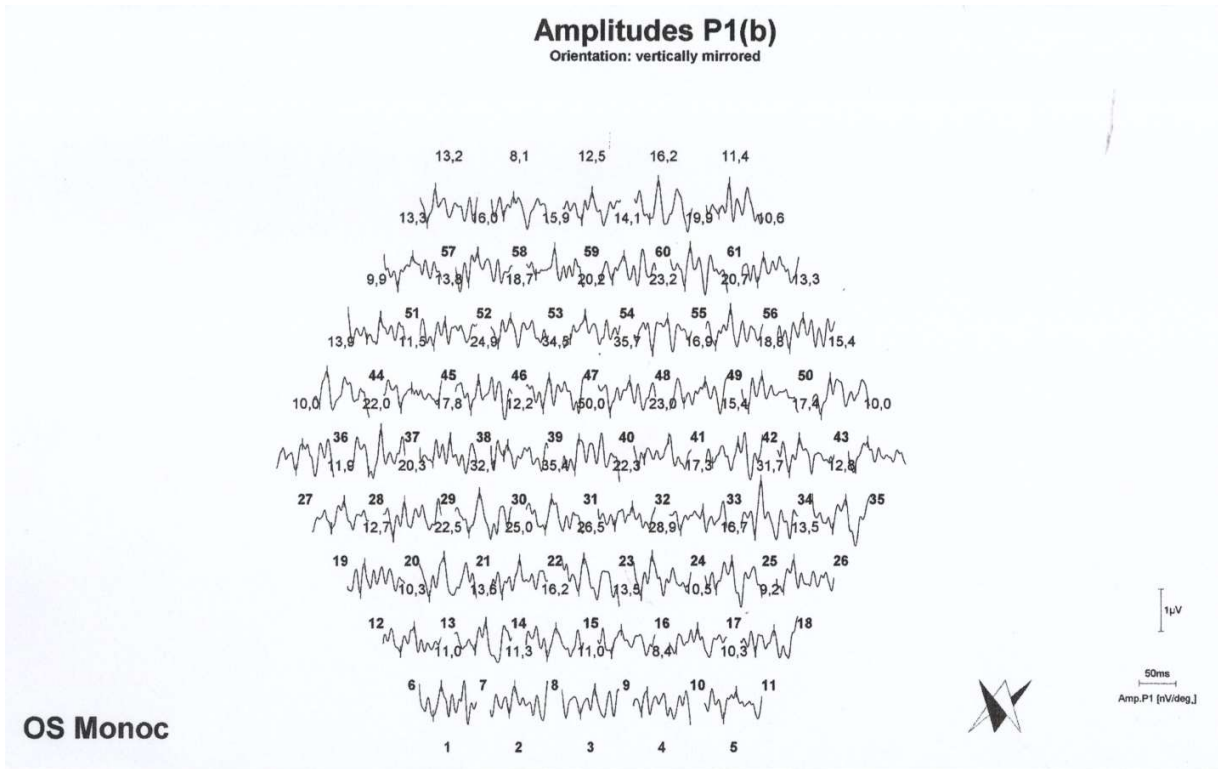


Figure S49. mfERG recording of patient IV/6, OS.

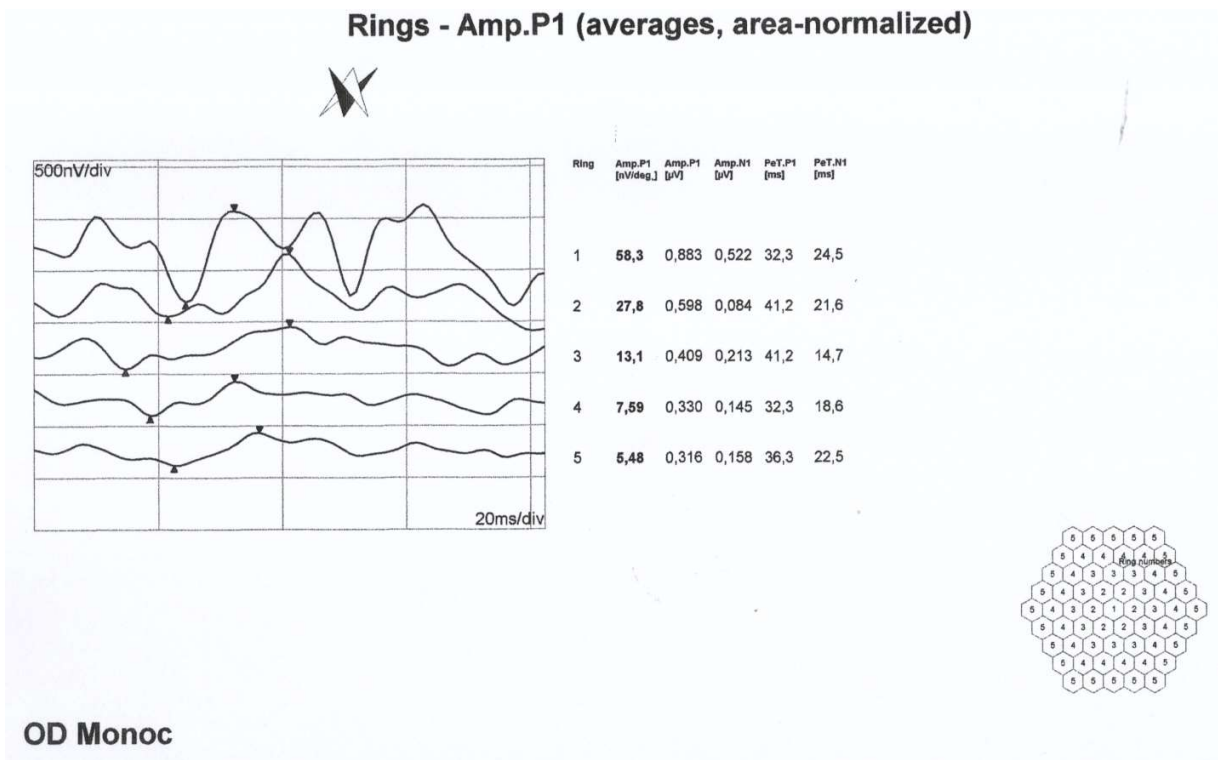


Figure S50. mfERG ring analysis of patient IV/6, OD.

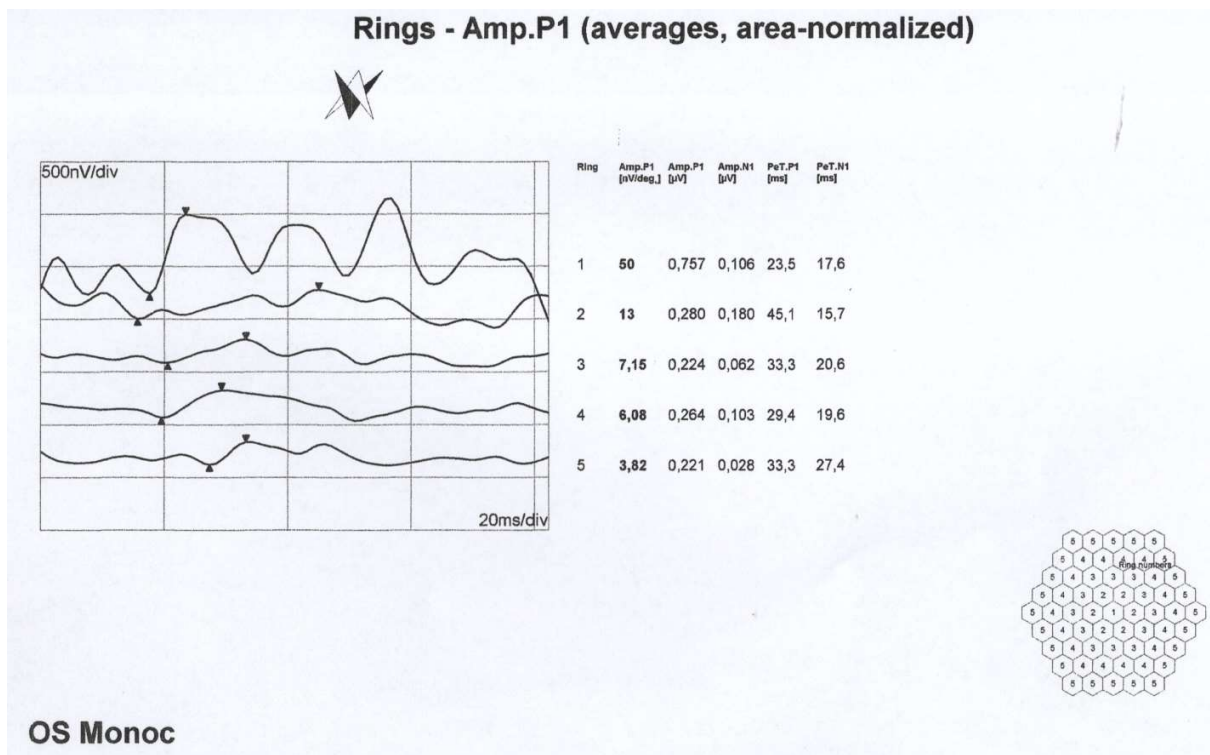


Figure S51. mfERG ring analysis of patient IV/6, OS.

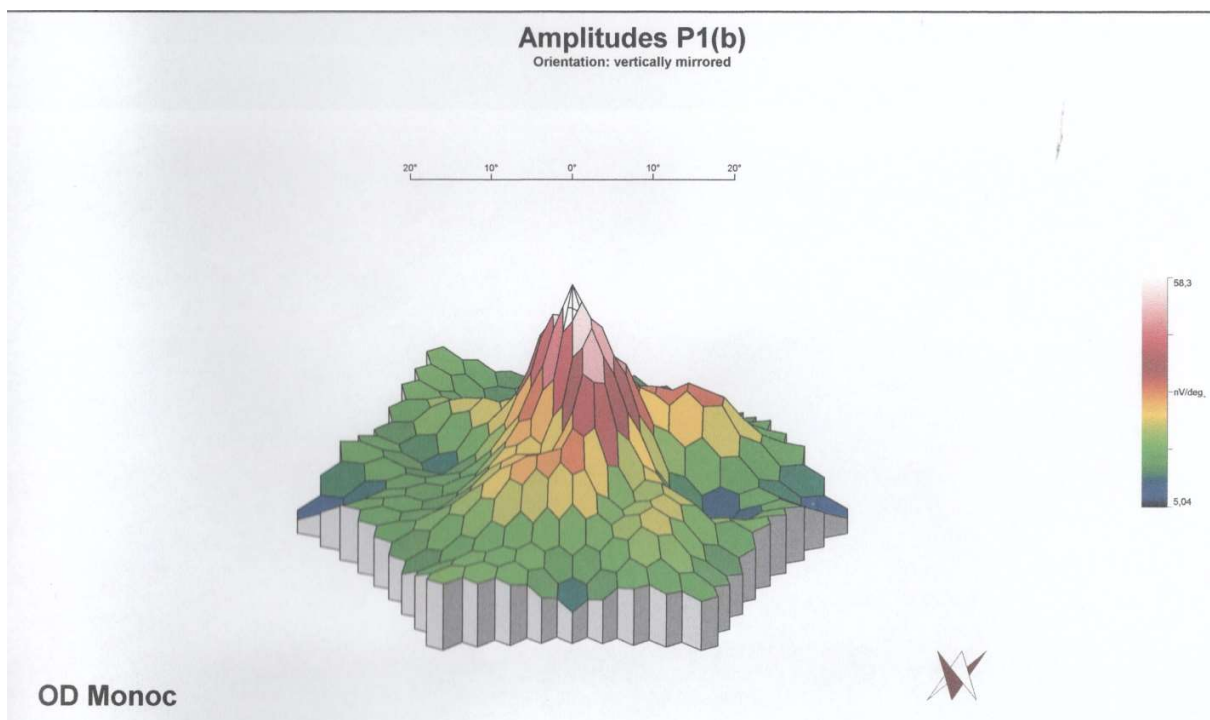


Figure S52. Multifocal ERG 3 D colour map of patient IV/6 severely myopic affected female right eye displays extensive subnormal responses within the whole central 30 degrees.

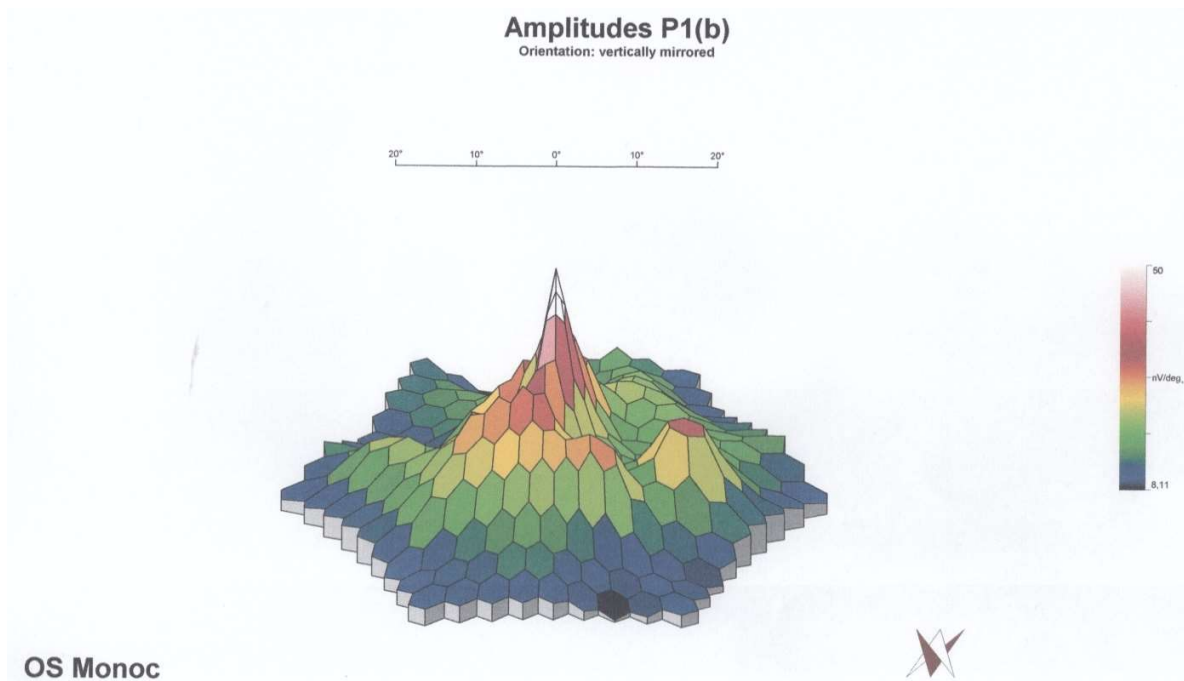


Figure S53. Multifocal ERG 3 D colour map of IV/6 severely myopic affected female left eye displays extensive subnormal responses within the whole central 30 degrees.

Pattern-ERG

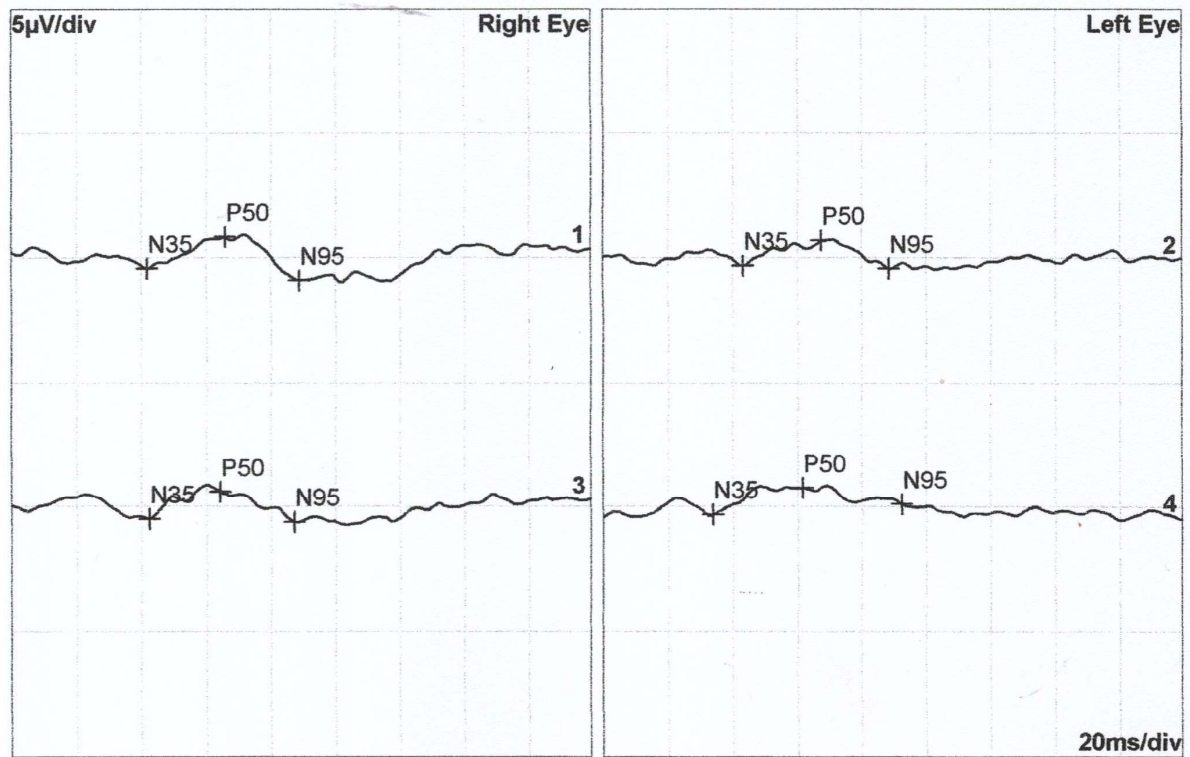


Figure S54. PERG recording of patient IV/10. Lines 1 and 3 and lines 2 and 4 represent pairs of replicate measurements.

Pattern-VEP

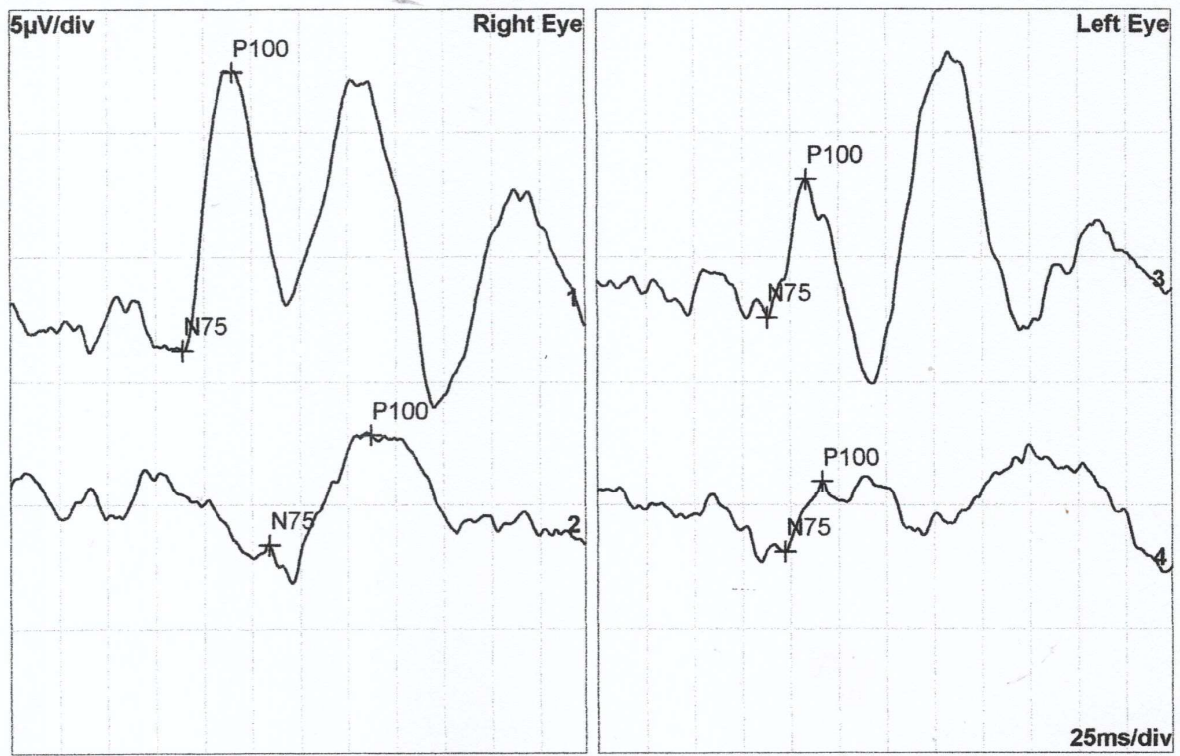


Figure S55. pVEP recording of patient IV/10. Lines 1 and 3 display responses to 60' stimuli and lines 2 and 4 represent responses to 15' stimuli.