ELECTRORETINOGRAPHIC FINDINGS IN PATIENTS WITH POSTERIOR MICROPHTHALMOS

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INTRODUCTION

Posterior microphthalmos is a rare type of microphthalmos that disproportionately affects the posterior ocular segment with normal external appearance of the eyes. High hyperopia and an elevated papillomacular retinal fold are the main findings in such patients.[1,2] Other abnormalities associated with this syndrome have been described, including absence of the capillary-free zone, retinal striae, chorioretinal folds, uveal effusion, pigmentary retinal dystrophy, pseudopapilledema, and a macular hole. No associated systemic abnormalities have been described, and an autosomal recessive mode of inheritance has been postulated.[1-3]

The aim of this study was to analyse electroretinographic changes in the eyes of patients with posterior microphthalmos.

MATERIAL AND METHODS

Thirty-four eyes of 17 patients with posterior microphthalmos were included in this study. The mean patient age was 20.3 ± 12.3 years (range, 5-43 years). There were 3 sporadic cases and siblings from 7 different families. Nine of 17 patients (52.9%) were males and 8 (47.1%) were females (sex ratio: 1.1).

Inclusion criteria were:

- Bilateral foreshortening of the posterior ocular segment (range, 7-11.2 mm).
- Normal or slightly smaller than normal anterior segment dimensions
- High hyperopia (range, + 12.00 - +19.00 diopters).
- No associated ocular or systemic abnormalities.

All patients underwent ophthalmic evaluation that included measurement of best-corrected visual acuity, slit-lamp examination, applanation tonometry, cycloplegic refraction, ophthalmoscopy, fundus photography, fluorescein angiography, and A-mode and B-mode ultrasonographic examination.

All patients underwent a standard flash electroretinography according to a modified protocol of the International Society for Clinical Electrophysiology of Vision (ISCEV)[4] on a 7000B Metrovision instrument. Electroretinographic values were compared with a group of 200 healthy subjects.

Table 1. Funduscopic, fluorescein angiographic and ultrasonographic findings.

Findings	Eyes (n=34)	(%)
Elevated papillomacular retinal fold	30	88.2
Absence or reduction of the capillary-free zone	34	100
Chorioretinal folds	20	58.9
Retinal folds	12	35.3
Crowded optic disc	34	100
Pigmentary retinal changes: ▪ Retinitis pigmentosa ▪ Pseudoretinitis pigmentosa related to uveal effusion	16 12 4	47 35.2 11.8
Uveal effusion	4	11.8
Sclerochoroidal thickening	34	100

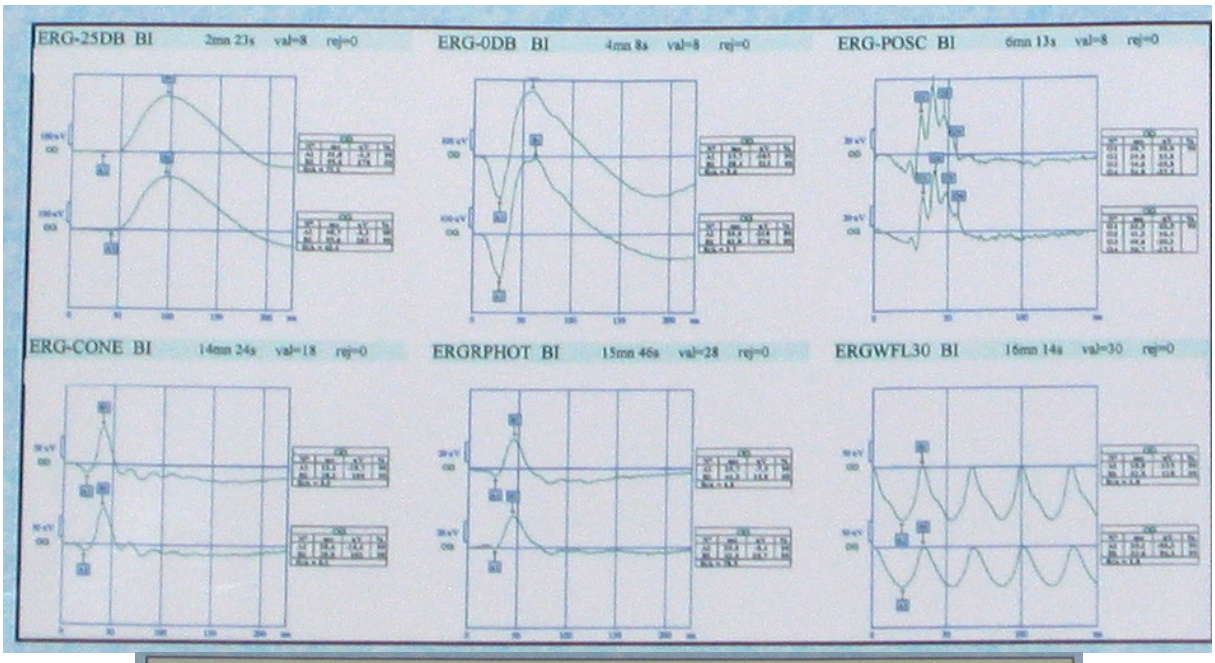

Figure 1. Normal electroretinogram.

Table 2. Scotopic electroretinographic findings

		Rod response (– 25 dB) 95% confidence interval	Maximal response (0 dB) a-wave 95% confidence interval	Maximal response (0 dB) b-wave 95% confidence interval
Normal fundus (n=14)	Amplitude (µV)	120.6 - 156.4	143.8 - 191	334.8 - 428.6
	Implicit time (msec)	93.5 - 102.1	21.5 - 23.3	44.8 - 51.4
Retinitis pigmentosa (n=12)	Amplitude (µV)	16.6 - 31.6	42.6 - 64.6	57.1 - 114.5
	Implicit time (msec)	76.5 - 97.3	23 - 25.6	49.2 - 53.8
Pseudoretinitis pigmentosa related to uveal effusion (n=4)	Amplitude (µV)	66.9 - 79.5	97.6 - 115.8	129.1 - 191.3
	Implicit time (msec)	78.7 - 95.7	21.1 - 26.3	54.2 - 56.4
Uveal effusion without retinal pigmentary changes (n=4)	Amplitude (µV)	20.6 - 131.4	52.5 - 177.1	138.7 - 377.7
	Implicit time (msec)	95.7 - 116.3	25 - 29	53.4 - 59

Tableau 3. Photopic electroretinographic findings

		Cone response (White) 95% confidence interval		Cone response (Red) 95% confidence interval		Cone response 30-Hz flicker b-wave 95% confidence interval
		a-wave	b-wave	a-wave	b-wave	
Normal fundus (n=14)	Amplitude (µV)	13.7 - 20.1	53.4 - 78.4	8.9 - 12.5	17.3 - 21.9	54.1 - 82.5
	Implicit time (msec)	23.1 - 24.7	40.2 - 41	22.2 - 25.6	40.5 - 42.5	35.2 - 36.6
Retinitis pigmentosa (n=12)	Amplitude (µV)	10.4 - 14.4	45.2 - 62.8	2.7 - 4.3	13 - 19	37.7 - 73.7
	Implicit time (msec)	22.8 - 24.2	39.8 - 44.8	24.4 - 28	42.4 - 47.2	34.1 - 37.3
Pseudoretinitis pigmentosa related to uveal effusion (n=4)	Amplitude (µV)	8.9 - 14.7	53.5 - 63.4	1.4 - 4.8	16.9 - 22.5	78.7 - 104.9
	Implicit time (msec)	22.3 - 23.3	38.8 - 39.6	22.3 - 24.5	41.1 - 44.3	33 - 34.3
Uveal effusion without retinal pigmentary changes (n=4)	Amplitude (µV)	10.5 - 19.5	34.8 - 75.4	3.4 - 5	8.7 - 26.7	34.6 - 111
	Implicit time (msec)	22.3 - 24.5	40.2 - 46	25.5 - 31.8	45.5 - 54.5	34.5 - 41.6

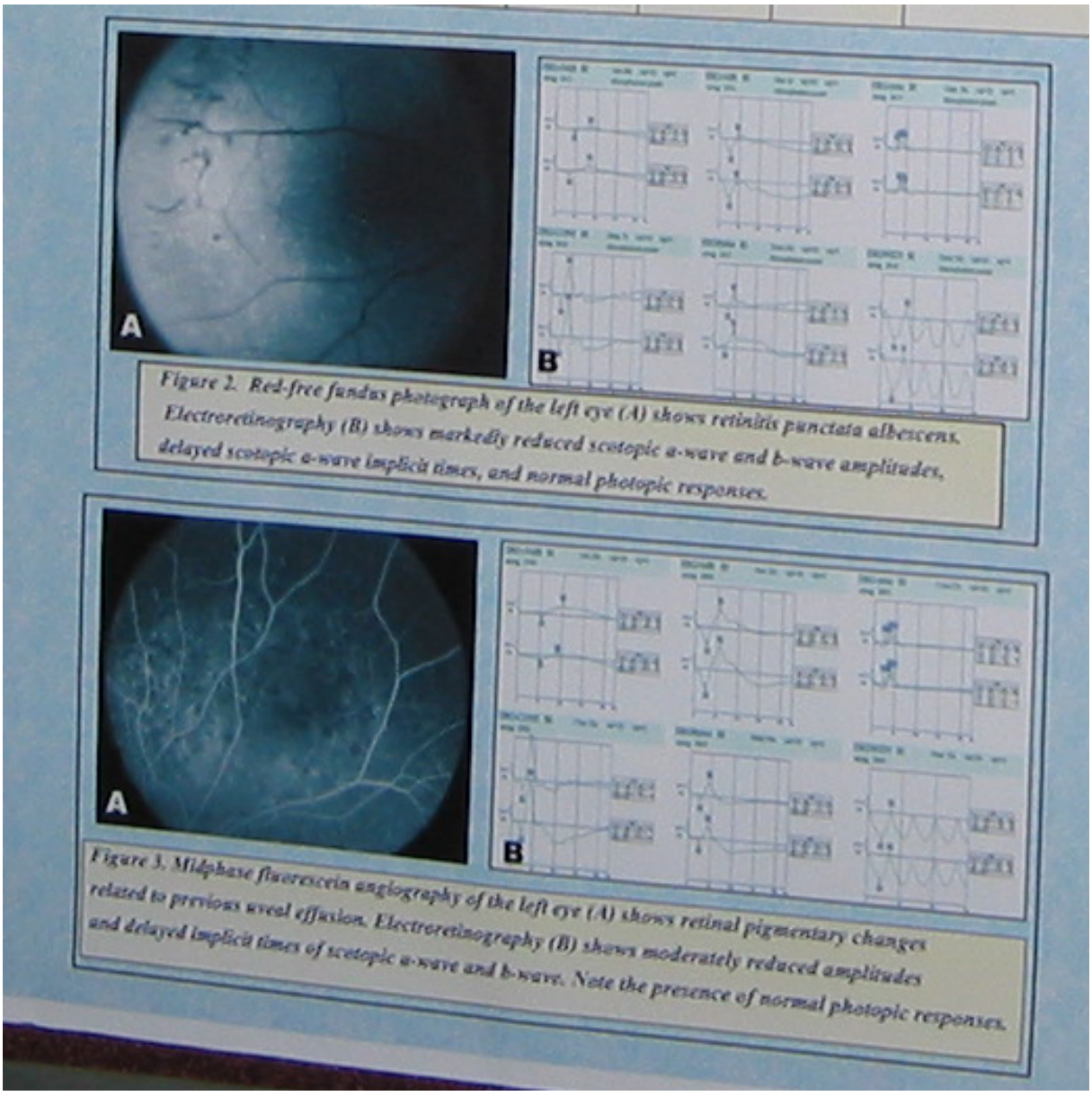

Figure 2. Red-free fundus photograph of the left eye (A) shows retinitis punctata albescens. Electroretinography (B) shows markedly reduced scotopic a-wave and b-wave amplitudes, delayed scotopic a-wave implicit times, and normal photopic responses.

Figure 3. Midphase fluorescein angiography of the left eye (A) shows retinal pigmentary changes related to previous uveal effusion. Electroretinography (B) shows moderately reduced amplitudes and delayed implicit times of scotopic a-wave and b-wave. Note the presence of normal photopic responses.

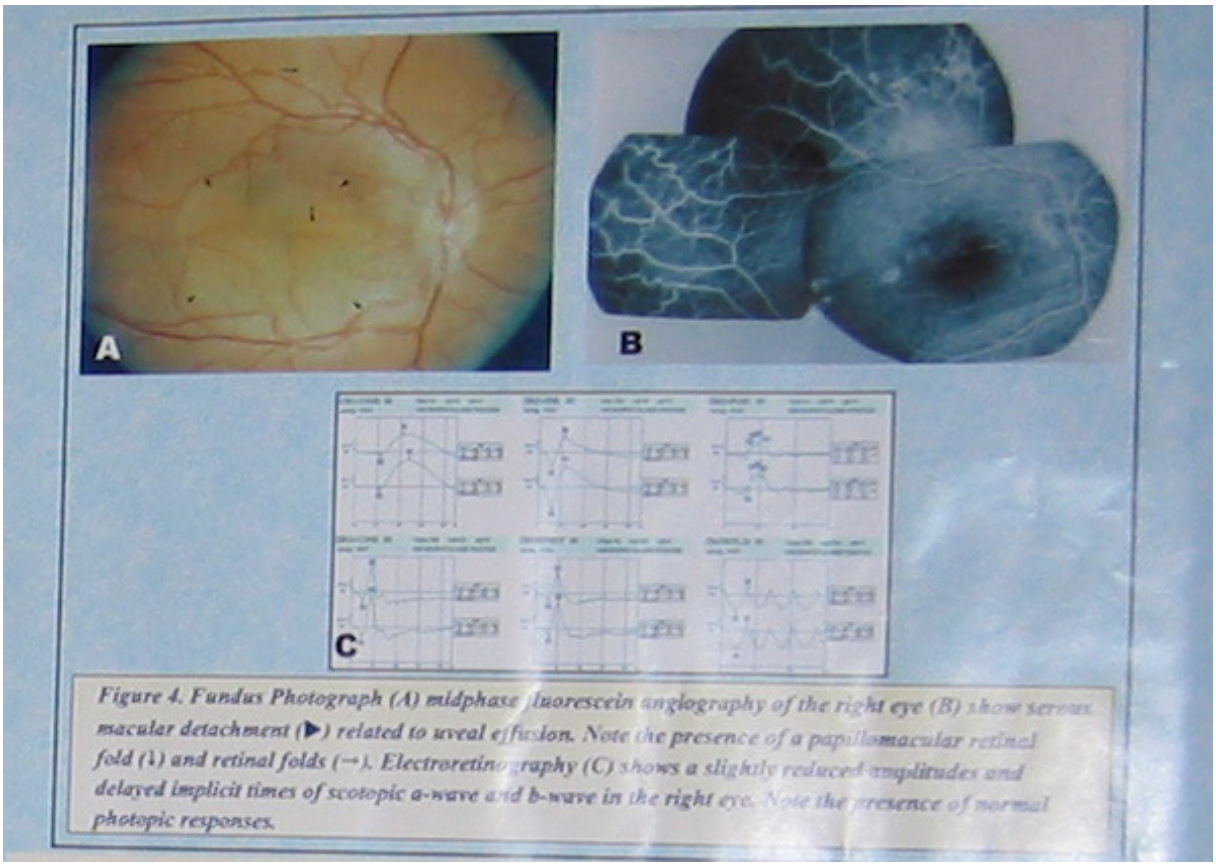

Figure 4. Fundus Photograph (A) midphase fluorescein angiography of the right eye (B) show serous macular detachment (▶) related to uveal effusion. Note the presence of a papillomacular retinal fold (↓) and retinal folds (→). Electroretinography (C) shows a slightly reduced amplitudes and delayed implicit times of scotopic a-wave and b-wave in the right eye. Note the presence of normal photopic responses.

RESULTS

The best-corrected visual acuity ranged from 20/800 to 20/40. The posterior segment changes associated with posterior microphthalmos are listed in Table 1.

Twenty of 34 eyes (58.9%) showed abnormal electroretinographic values (Table 2 and 3) :

- Retinitis pigmentosa was observed in 12 eyes (35.3%) with a history of nyctalopia and gradual visual loss. Electroretinography disclosed markedly reduced scotopic a-wave and b-wave amplitudes, delayed scotopic a-wave implicit times, and slightly reduced amplitudes and delayed implicit times of photopic a-wave and 30-Hz flicker b-wave (Figure 2).
- Pseudoretinitis pigmentosa secondary to prior uveal effusion, without history of nyctalopia was noted in 4 eyes (11.8%). Electroretinography showed moderately reduced amplitudes and delayed implicit times of scotopic a-wave and b-wave, with slightly reduced photopic a-wave amplitudes (Figure 3).
- Uveal effusion without retinal pigmentary changes was found in 4 eyes (11.8%). Electroretinography showed variably reduced amplitudes and delayed implicit times of scotopic a-wave and b-wave, with slightly reduced amplitudes and delayed implicit times of photopic a-wave and 30-Hz flicker b-wave (Figure 4).

CONCLUSIONS

- Our findings, as well as those from a previous report[1], showed that retinitis pigmentosa may be associated with posterior microphthalmos.
- Findings from previous reports, showed that high refractive amblyopia and elevated papillomacular retinal fold were the main causes of visual loss in patients with posterior microphthalmos.[1,2,5-7] Data from our study indicate that other chorioretinal changes, such as retinitis pigmentosa, pseudoretinitis pigmentosa secondary to prior uveal effusion, and active uveal effusion, should be considered as causes of significant visual impairment in patients with posterior microphthalmos.
- Electroretinography in conjunction with clinical examination is helpful in differentiating between retinitis pigmentosa and pseudoretinitis pigmentosa, and in monitoring the visual function in patients with posterior microphthalmos.

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