Graefe's Arch Clin Exp Ophthalmol (1990) 228:432–437

Graefe's Archive
for Clinical and Experimental
Ophthalmology

X-linked congenital retinoschisis

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Received September 7, 1989 / Accepted March 7, 1990

Abstract. The natural history and electrophysiological findings of 52 patients with X-linked congenital retinoschisis with a follow-up of up to 26 years are described. The mean visual acuity was reduced to 0.24 ± 0.2 and remained unchanged in most patients during this time. If visual loss occurred, it usually happened in the first decennium. The complications were retinal detachments in 11% and vitreous hemorrhages in 4% of the eyes. In general, the vitreous hemorrhages resolved spontaneously. Retinal detachments were treated successfully with conventional buckling procedures. Redetachments occurred in about 40%. Prophylactic laser coagulation was of no use because it was complicated by detachment in 43% of our series. The electro-oculogram was usually normal. In addition to the known electroretinographic findings of normal a-wave and reduced b-wave amplitudes, we found prolonged b-wave latencies and implicit times, as well as a reduced 30 Hz flicker response.

Introduction

Since Haas' [8] first description of X-linked congenital retinoschisis in 1898, several reports have been published. Most have been casuistic and only three larger series of patients have been reported from The Netherlands [5], Finland [7], and Canada [6]. The disease had been described in all races [3, 14]. It is known as a bilateral disorder with characteristic ophthalmoscopical and electrophysiological features. Visual acuity is more or less impaired because of a central retinoschisis. Peripheral retinoschisis is present in about half of the patients [5]. Histology shows a schisis within the nerve fiber layer in contrast to senile retinoschisis [2, 12, 20] where the outer plexiform layer is involved.

We evaluated the natural history and electrophysiological findings of patients with X-linked congenital retinoschisis who attended in the Essen Eye Hospital during the last 27 years.

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Materials and methods

Since 1962, 52 patients with X-linked congenital retinoschisis have been examined. Twenty-one patients were seen only once and 31 twice or more with a follow-up of between 1 and 26 years. The mean follow-up of these 31 patients was 5.8 ± 4.7 years. The mean age at first examination was 18.4 ± 14.5 years. Four older patients of about 50 years were diagnosed when the whole family examined. The mean age at the first examination for the remaining 48 patients was 15.7 ± 10.4 years. The case histories, fundus photographies, and electrophysiological findings of all patients were evaluated. The diagnostic criteria included fundus findings, electroretinographic results and/or family history, especially in older patients. Twenty patients were from several families and another 7 had a positive family history.

Color vision tests included pseudoisochromatic plates, the desaturated Panel-D-15 test and Nagel's anomaloscope test. Electrooculograms (EOG) were recorded according to Rhode and Täumer [16]. For ERG recording we used a modified Henkes' contact lens as a corneal electrode, an indifferent electrode on the forehead, and the preauricular skin served as the reference ground. All electrodes were connected wireless via electrolyte bridges (0.9% NaCl in 1.5% agar agar) to three non-polarizing calomel half-cells [10]. The signals were fed into an amplifier working either in the AC or DC mode (Toennies DA II with DC preamplifier). The upper frequency limit was at 1000 Hz; the lower frequency limit was 0.5 Hz in the AC mode.

After 40 min of dark adaptation and with maximum dilation of the pupil (phenylephrine 2.5% tropicamide 1.0%), the ERGs were recorded in the dark. Stimulus duration was 10 ms. Six different light intensities (1–6), increasing by one logarithmic unit in steps from the b-wave threshold of the normal eye, were used for the dark-adapted recordings. The maximum light intensity was 780 cd/m^2. The light-adapted recordings were performed under white light adaptation with 4.5 cd/m^2 and with the light stimuli 4–6. The 30 Hz flicker stimulus had the light intensity 5. White light from a filtered xenon light source served as stimulus in all examinations. The potentials were displayed on an oscilloscope, recorded on a paper graph and stored digitally for later work-up on a computer disc. No computer averaging was done. The normal range is considered to be the mean of 40 normal eyes $\pm$ two standard deviations. For statistical evaluation we used Student's *t*-test.

Results

Refractive state

The refractive state was similar in both eyes in a given patient. Only in two cases there was a difference of more than 2 D. Seventy-two of 91 eyes (79%) had refractive errors, excluding 13 eyes with a visual acuity of less than 0.02 or lack of cooperation. The mean refractive error was $+2.3 \pm 3.1$ D with a range of -5.0 to $+10.0$ D (Fig. 1). Only one patient was more myopic than -1.5 D. However, 23 eyes (25.2%) of 13 patients were more hyperopic than 3 D with a mean of $+6.0 \pm 2.2$ D. Seventeen eyes (19%) of 9 patients were more hyperopic than 4 D with a mean of 6.9 ± 1.7 D and 14 eyes (15.4%) of 7 patients were more hyperopic than 6 D with a mean of $+7.5 \pm 1.3$ D. Fifty eyes (55%) had astigmatism; the mean was -1.5 ± 0.7 D with a maximum of -3.25 D. Astigmatism more than 2.0 D was present in 17 eyes (19%).

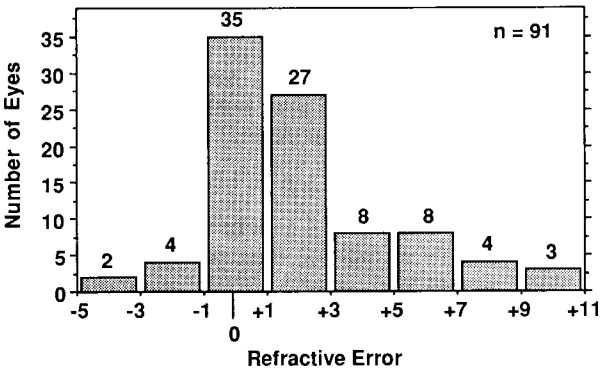

Fig. 1. Distribution of refractive error in 91 eyes in X-linked retinoschisis

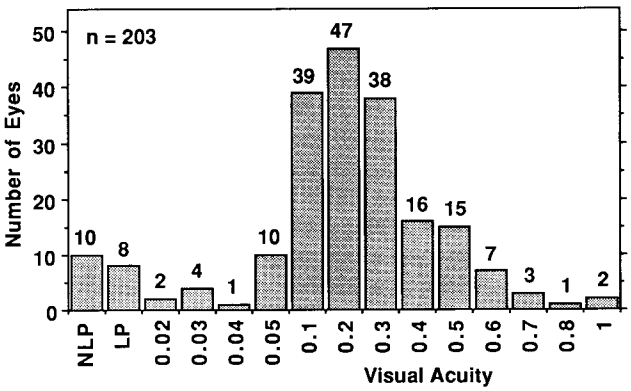

Fig. 2. Distribution of visual acuity in 203 examinations of 98 eyes in X-linked retinoschisis

Visual acuity

In three infants the visual acuity could not be evaluated. In the remaining 98 eyes 203 measurements of visual acuity at different examinations were obtained. The mean visual acuity was 0.24 ± 0.2 (Fig. 2). The range was from no light perception to 1.0. In eyes without complications there was no tendency towards visual loss with increasing age (Fig. 3). On the contrary, visual acuity was better in older patients. The older patients with minor loss of visual acuity and less severe signs of disease were found during familial examinations. Some of them had not even noticed any disturbances in visual function before.

In 21 cases with a follow-up of 5 years or more, the development of visual acuity is known. In 26 eyes without complications like vitreous hemorrhage or retinal detachment the visual acuity remained unchanged over time. Only in 4 eyes (13%) of 2 patients did the visual acuity decrease two or more lines (0.7 to 0.4, 0.5 to 0.3, 0.5 to 0.2 twice). Visual loss occurred before puberty. None of the remaining patients without complications complained about visual loss at any time. The development of degenerative macular changes in older patients was never associated with further visual loss.

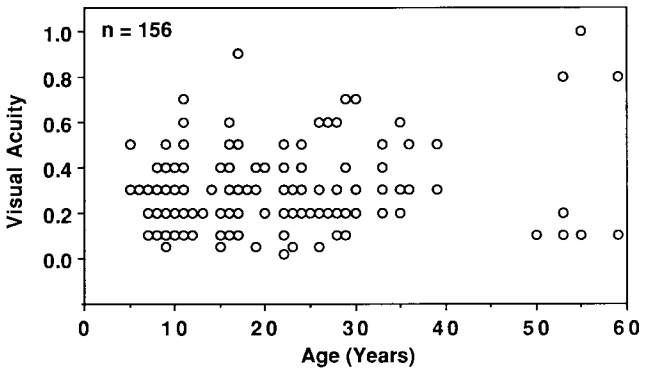

Fig. 3. Distribution of visual acuity compared to age in X-linked retinoschisis in 156 examinations of 79 eyes without complications. The range is from 0.02 to 1.0. In two patients older than 50 years the visual acuity was better than 0.7. These patients were diagnosed during familial examinations

Binocular vision

Strabism was found in 15 patients (29%). Seven times there was a convergent and 8 times a divergent squint. The causes were retinal detachment, media opacities and peripheral retinoschisis extending over the posterior pole with loss of central fixation.

Visual fields and color vision

Visual fields obtained in 50 eyes of 26 patients were normal when no peripheral retinoschisis was present. Eyes with peripheral retinoschisis had visual field defects corresponding to the retinoschisis without progression in all the patients. Because of the temporal location of the retinoschisis they were detected with difficulties in the nasal part of the visual fields.

Color vision was tested in only 14 patients and was normal in 8. Three patients had a protanomaly. In three patients the Nagel anomaloscope revealed red-green dyschromatopsia.

Anterior segments

The anterior segments of the eyes were normal except for two older patients and 10 eyes with a cataract complication after development of retinal detachment. One of the two older patients was aphakic because of senile cataract in one eye and the other had a corneal transplant and aphakia because of a herpetic corneal ulcer. The intraocular tension was normal, with the exception of one older patient with simple chronic glaucoma and 3 eyes with secondary glaucoma.

Posterior segments

In 102 of 104 eyes we were able to perform ophthalmoscopic examinations. In all eyes macular pathology was present. Seventy-two eyes suffered from macular retinoschisis. Seven eyes had a peripheral retinoschisis overriding the posterior pole. In 11 the eyes of patients older than 33 years, including all patients older than 40, we were not able to detect macular retinoschisis ophthalmoscopically, but these patients had distinct pigment irregularities in the macular area. Eight eyes had a retinal detachment, seven eyes a reattached retina.

Of 87 eyes without a history of retinal detachment, 46 eyes (52.9%) had peripheral retinoschisis. In seven eyes (8%), the peripheral retinoschisis was overriding the posterior pole. In 44 of 46 eyes (96%) with peripheral retinoschisis the lower temporal quadrant was affected (Fig. 4). In two eyes (4%) the peripheral retinoschisis was limited to the upper temporal quadrant. In 22 eyes the peripheral retinoschisis was limited to the lower temporal quadrant, in 9 eyes to both lower quadrants, and in 12 eyes limited to both temporal quadrants. In 1 eye the peripheral retinoschisis extended from the upper temporal to the lower nasal quadrant. There was no significant difference in the incidence of peripheral retinoschisis between young and elderly patients. In the course of our study the peripheral retinoschisis was constant except for two patients, who developed peripheral outer retinal breaks.

In most cases without complications the fundus pathology was similar in both eyes. However, of 22 patients with peripheral retinoschisis and no history of retinal detachment, in 6 patients (27%) the peripheral retinoschisis was unilateral.

In 9 eyes of 8 patients fluorescein angiography was done. The macular area showed focal hyperfluorescence in the early phase. This was more distinct in older patients, which was in parallel to the pigment irregularities found on ophthalmoscopy. We did not see fluorescein leakage or staining of the retina.

Complications

In 5 eyes vitreous hemorrhages occurred before the age of 10 years. One eye became amaurotic because of a retinal detachment. In 4 eyes the hemorrhage resolved

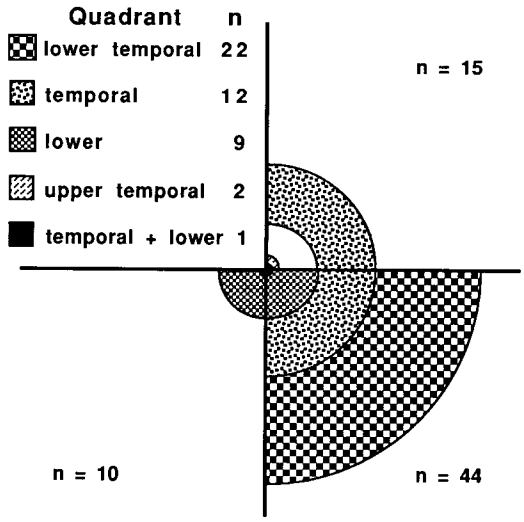

Fig. 4. Distribution of peripheral retinoschisis in 46 eyes in X-linked retinoschisis; $n = 46$ eyes

spontaneously without visual loss. In one case the hemorrhage occurred following contusion. After absorption of the blood, peripheral retinoschisis was detected that was not seen earlier.

In 15 eyes (14.4%) of 12 patients retinal detachment occurred. In 11 eyes (10.7%) detachment occurred spontaneously and in two cases, bilaterally. In one eye retinal detachment developed following a contusion and in 3 eyes after prophylactic laser treatment. In 6 of 8 cases with spontaneous unilateral retinal detachment, the other eye had a peripheral retinoschisis.

In most cases the retinal detachment occurred before the age of 11. In 2 cases detachment developed at age of 2 years, in 1 at 5 years, in 3 at 8 years, and in 1 at 11 years. In 2 eyes a long-standing retinal detachment was diagnosed at the age of 16 years and in 2 eyes at the age of 50 years. In the last 2 cases the visual acuity of the detached eye had been low since childhood. One patient presented with spontaneous reattachment in one eye at the age of 20 years with a visual acuity of 0.03.

Four eyes with retinal detachment were not treated because of long-standing detachment or secondary glaucoma. Ten eyes were operated on successfully with encircling bands. Four eyes developed redetachments even after secondary buckling procedures or vitrectomies. Three of those eyes had a secondary glaucoma that could be treated with medication in two cases. One eye had to be enucleated.

In the treated eyes with long-term successful follow-up, visual acuity was light perception in 1 case, 0.05 in another case and 0.1 in the other 4 cases. In the other eyes with retinal detachment, visual acuity was light perception in 4 eyes and no light perception in 4 eyes.

In 7 eyes prophylactic laser treatment had been done before 1975. Three eyes (43%) developed retinal detachment, 2 of which became amaurotic. In 1 eye the retinal detachment was treated successfully. Four eyes were unchanged after laser treatment.

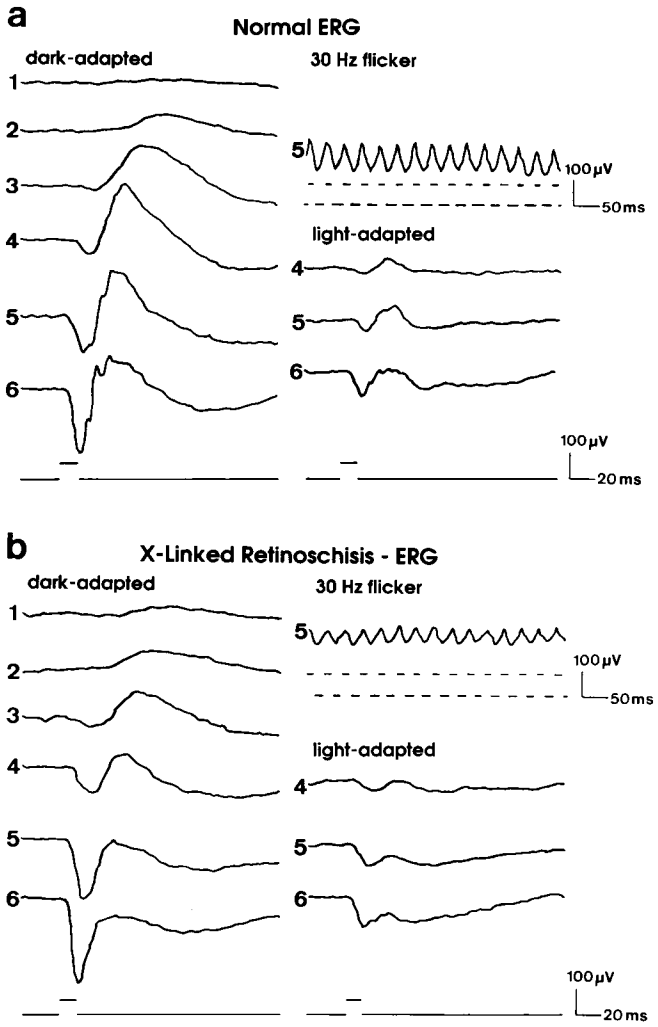

Fig. 5. a Normal electroretinogram: the dark-adapted recordings are on the left side; the light-adapted recordings are on the lower right side and the 30 Hz flicker response is on the upper right side. Below each group of recordings is a trace of the light stimuli. The numbers in front of each trace indicate the light intensity. **b** Electroretinogram of a 17-year-old patient suffering from X-linked retinoschisis. Compared with the normal electroretinogram, the b-wave amplitudes, the b/a-ratios and the 30 Hz flicker response are reduced

Electro-oculography

Electro-oculograms were obtained of 17 patients. Three eyes of 34 had largely diminished base values and light rises due to retinal detachments. In all other cases the electro-oculogram was normal. The mean light rise/base value ratio was 215±41% in 31 eyes (normal 187± 36%).

Electroretinography

Electroretinograms were obtained in 37 eyes of 22 patients. Figure 5 shows a typical electroretinogram of a patient with X-linked retinoschisis. In all patients the a-wave amplitudes were in the lower normal range when dark adapted and quite normal when light adapted. The b-wave amplitudes were reduced to 40–50% when dark adapted. When light adapted, the b-wave amplitudes were reduced at light intensity 4, borderline at light intensity 5, and normal at maximum light intensity. Accordingly, the mean b/a-ratio at maximum light intensity was reduced to 0.86±0.1 (normal 1.28±0.1, $P<0.0001$) when dark adapted and 0.74±0.2 (normal 0.95±0.18, $P<0.0002$) when light adapted.

The a-wave latencies were in the upper normal range at all stimulus conditions. The b-wave latencies and implicit times were in the upper normal range or longer than normal at all stimulus conditions. Compared to 20 normal eyes in the same age range, the b-wave latencies were prolonged at dark and light adapted conditions at all stimulus intensities ($P<0.0001$, Table 1). The b-wave implicit times were prolonged when light adapted at all stimulus intensities ($P<0.0001$) and when dark adapted at all stimulus intensities except at maximum intensity ($P<0.02$–0.0001, Table 2).

The mean 30 Hz flicker response was reduced (60± 21 μV, normal 136±26 μV, $P<0.0001$). It was normal in 4 eyes. Oscillatory potentials were missing in 28 eyes and were present in 9 eyes.

Table 1. B-wave latencies in 20 normal eyes and 37 eyes with X-linked congenital retinoschisis

Stimulus	Normal		Retinoschisis		$P<$
	mean (ms)	±SD	mean (ms)	±SD	
Dark ad. 3	28	±3.2	36	±5.0	0.0001
Dark ad. 4	23	±3.2	31	±5.0	0.0001
Dark ad. 5	20	±2.7	23	±2.9	0.0001
Dark ad. 6	14	±1.0	17	±2.0	0.0001
Light ad. 4	20	±3.2	24	±2.0	0.0001
Light ad. 5	17	±1.4	21	±1.9	0.0001
Light ad. 6	15	±0.8	18	±2.1	0.0001

ad., Adapted; SD, standard deviation

Table 2. B-wave implicit times in 20 normal eyes and 37 eyes with X-linked congenital retinoschisis

Stimulus	Normal		Retinoschisis		$P<$
	mean (ms)	±SD	mean (ms)	±SD	
Dark ad. 3	67	±5.7	74	±9.1	0.005
Dark ad. 4	51	±2.3	58	±5.9	0.0001
Dark ad. 5	42	±1.8	45	±5.1	0.02
Dark ad. 6	39	±1.4	38	±4.0	NS
Light ad. 4	38	±2.1	42	±1.9	0.0001
Light ad. 5	38	±1.9	41	±2.0	0.0001
Light ad. 6	35	±0.9	36	±2.9	0.0001

ad., Adapted; SD, standard deviation; NS, not significant

Discussion

The natural course of eyes with congenital retinoschisis showed no marked changes over the years. In our patients the visual acuity was reduced to about 0.25 and remained unchanged with time in most cases. If a loss of visual acuity occurred, it nearly always happened before puberty. After an early decrease in visual acuity it can remain stable for many years. Forsius [7] found a slow deterioration of visual acuity in his patients. The main visual loss occurred in his patients when they were older than 70 years. None of our patients was that old. Other authors with larger series of patients or long follow-up [1, 6, 11] observed minimal or no changes in visual acuity after childhood and considered the X-linked congenital retinoschisis to be slowly progressive [5].

The mean refractive error with a hyperopia of about +2.5 D was in accordance with the findings of other authors [5, 11]. The hyperopia of more than 4 D with a frequency of 19% and of more than 6 D in 15% in our patients has not been described before. In a normal population hyperopia of more than 4 D occurs in 4.5% and of more than 6 D in about 1.5% [17]. In young hyperopic patients with retinal detachment, the fellow eye should be examined carefully, as peripheral retinoschisis is suggested. We did not see myopia of more than 5 D. Accordingly, in patients with myopia and suspected retinoschisis, detachment due to hole formation must be excluded.

In all of our patients with X-linked congenital retinoschisis macular pathology was present. A macular retinoschisis was present in all young patients before the complications developed. When patients grow older than 30 years, macular retinoschisis is only detected with difficulty and central pigment irregularities develop. About 52.9% of eyes without complications had peripheral retinoschisis. Similar fundus findings have been described before [5–7, 11].

The two main complications of X-linked congenital retinoschisis were retinal detachment and vitreous hemorrhage in our series. These complications developed in childhood. The vitreous hemorrhage needed no therapy in our cases and resolved spontaneously. Vitrectomy may be useful when a retinal detachment develops.

Whereas Deutman [5] considered retinal detachment to be a rare complication, retinal detachment occurred spontaneously in about 11% of the eyes in our series. The retinal detachment can be treated successfully with conventional buckling procedures. Redetachments, however, occurred in 40% even after reoperations, including vitrectomies. Peripheral retinoschisis is a risk factor for retinal detachment, because 75% of our patients with spontaneous unilateral retinal detachment had a peripheral retinoschisis in the fellow eye.

We do not consider prophylactic laser coagulations to be useful as most authors [5, 6]. Moreover, prophylactic treatment is a risk factor for retinal detachment because in 43% retinal detachments occurred. In none of our cases did the retinoschisis improve after coagulation.

The range of manifestation of X-linked congenital retinoschisis differs widely between patients. Some have only minimal foveal retinoschisis; others develop retinal detachment in both eyes in early childhood. Deutman [5] states that both eyes of one patient show a similar state of disease. In our series this is only true when considering the refractive status, visual acuity, and macular retinoschisis. However, peripheral retinoschisis appeared in 27% of the cases, spontaneous retinal detachment in 78% of the cases, and vitreous hemorrhage in all cases unilateral.

The electro-oculogram was normal in all eyes without additional pathology. Similar EOG findings have been reported by other authors with normal results in about 80 to 90% [9, 14, 18, 19].

Our electroretinographic findings showed lower normal a-wave amplitudes and reduced b-wave amplitudes with a reduced b/a-ratio as has been described before [4, 9, 15, 18]. The 30 Hz flicker response was also markedly reduced although it can be normal in some patients. Hirose [9] found pathological flicker responses in his patients. Oscillatory potentials were missing in most of our patients, which is understandable because of the alteration in the inner retinal layers. Peachey [15] never found normal oscillatory potentials in his patients.

The shape of the b-wave is altered in X-linked congenital retinoschisis not only because of the diminished amplitudes, but also because of changes in the b-wave time characteristic that have not been described before. We found a prolongation of b-wave latencies and implicit times.

It was most interesting that the alteration in the b-wave was more pronounced in the dark-adapted recordings than in the light-adapted recordings. At maximum stimulus intensity the b-wave amplitude was normal when light-adapted. This indicates a more distinct dysfunction of the rod-mediated b-wave than of the cone-mediated b-wave. The electroretinographic findings indicate a general intraretinal disorder even in eyes without peripheral retinoschisis.

Histopathological examinations suggest that the primary lesion in X-linked retinoschisis is in the innermost part of the Müller cells or at the border of the retinal nerve fibers [2, 12, 20]. A defect in the innermost part of the Müller cells is most likely because of electrophysiological findings. Although generation of the b-wave in the electroretinogram is not completely clarified, there is agreement that the Müller cells play an important role in b-wave generation [13]. Our electroretinographic findings show a more pronounced defect in the generation of the rod-mediated b-wave than the cone-mediated b-wave even in eyes without peripheral retinoschisis. This points toward a generalized defect in Müller cells. The foveal pigment irregularities are probably secondary, because they are more distinct with increasing age. In addition, the electro-oculogram shows no evidence of a general disorder in the pigment epithelium. Although the main sign of X-linked retinoschisis is visual impairment due to foveal retinoschisis, a generalized defect of the innermost parts of the Müller cells seems to be responsible for the morphological and functional alterations.

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Communications

Graefe's Archive for Clinical and Experimental Ophthalmology

Graefe's Arch Clin Exp Ophthalmol (1991) 229:497

Letter to the Editor

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Received November 19, 1990 / Accepted February 4, 1991

Sir,

The recent article by Kellner and colleagues [2] constitutes an important report on the follow-up study of X-linked retinoschisis. Nevertheless, it contains a startling omission: in spite of the high number of eyes with peripheral retinoschisis (46 in 52 patients), the authors make no mention of breaks in the inner layer of peripheral retinoschisis. Rather, they report on peripheral outer retinal breaks in 2 of the 46 eyes.

While outer peripheral breaks in X-linked retinoschisis are very rare, many authors [1, 3, 4] confirm that X-linked retinoschisis is usually characterized by a thin, frequently shrunken inner layer of split retina with inner breaks of various size. Ricci's term for this is "retinoschisis with fenestration" [4]. It is not to be confused with rhegmatic retinal detachment.

These inner retinal breaks are occasionally very large, with consequent exposure of retinal vessels. Such vessels are surrounded only by a thin sheet of residual inner layer tissue, protrude into the vitreous, and have the appearance of vascularized vitreous "veils" [3]. Although detached remnants of the inner retinal layer may float in the vitreous, they are not true components of the vitreous.

This aspect of inner retinal breaks has clinical import. The authors of the above-mentioned article found 15 retinal detachments in 12 patients in this group. In my experience, true retinal detachment in X-linked retinoschisis must be carefully distinguished from peripheral retinoschisis with defects of various size in the inner layer.

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Reply

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We acknowledge the comments of W. Lisch, which give us a chance to expand on some of our clinical findings. We believe that a misunderstanding has taken place. We only reported that two outer retinal breaks in peripheral retinoschisis developed within the follow-up time; this is somewhat unusual. Outer retinal breaks are often posterior to the equator and are rarely overlooked because of the changes in light reflectance.

In contrast to the authors cited by Lisch, we found no inner retinal breaks in most of our patients with peripheral retinoschisis. No large holes in the inner retinal layers and vitreous "veils" were seen. Inner retinal breaks are peripheral and their identifications is difficult due to the thinning of the inner retinal layers. For the same reason the retinal vessels seem to be "exposed." When the thinning of the inner retinal layers is pronounced, it may simulate retinal breaks, because it is difficult to identify the thin inner retinal layers between the prominent retinal vessels.

In our series 23 eyes had inner retinal breaks; 15 eyes had retinal detachments and, consequently, inner and outer retinal breaks. The frequency of spontaneous retinal detachment was 11% in our series. In the literature variations are reported between rare [1] and 22% [2].

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