# Perspectives on the management of the complications of senile retinoschisis

#### Abstract

Purpose To evaluate various published views regarding the management of complications of senile retinoschisis. Methods To analyze and compare various reports of studies of natural history and prognosis, and comparative risks of nonintervention vs prophylactic surgery. Results Although a number of recent techniques for treatment or prevention of complications of senile retinoschisis have been reported with generally satisfactory results, the numbers of cases have been small and no consensus can be said to have been achieved as to superiority of method. When compared to the extremely low likelihood of the natural occurrence of complications such as posterior progression to involve the macula, or of the development of progressive clinical retinal detachment, as well as the occurrence of risks of treatment, it appears preferable to postpone treatment until these complications actually make their appearance.

*Conclusions* It is recommended that in cases of retinoschisis either without or with breaks, or retinoschisis with localized 'schisis-detachment', that prophylactic treatment be withheld except in a very few exceptional cases. In all cases of progressive, symptomatic rhegmatogenous retinal detachment however, prompt surgical treatment should be carried out. *Eye* (2002) **16**, 359–364. doi:10.1038/ sj.eye.6700191

*Keywords:* senile retinoschisis; retinal breaks; posterior progression; 'schisis-detachment'; rhegmatogenous retinal detachment; prophylactic treatment; natural history

#### Introduction

Although first reported 68 years ago<sup>1</sup> and by now a well-known and commonly recognized

#### NE Byer

disease, senile retinoschisis is still, in some aspects, a poorly understood and somewhat mystifying entity. The fact that it is almost always without symptoms explains why most cases go without detection for the lifetime of the patient. This absence of symptoms has effectively prevented very many unnecessary operations by surgeons hoping to eradicate or control the disease.

#### Materials and methods

My purpose is to discuss some overall perspectives that should guide our ideas of the management of its different forms, rather than to evaluate details of surgical techniques which may be useful in the relatively rare complications of progressive retinal detachment or posterior extension of the retinoschisis or of subretinal fluid. Four complications will be discussed including: enlargement, posterior layer retinal breaks, schisis-detachment and progressive, frank, symptomatic retinal detachment.

## Results

#### (I) Enlargement

Posterior extension of retinoschisis to endanger the macula has always been one of the chief concerns in this disease and one of the most urgent indications for treatment. Retinoschisis in a small minority of cases has a tendency to gradually enlarge. In a large series, over a period of 9 years, this was observed, in order of frequency, in the following directions: laterally (6%); height (5%); and posteriorly (3%).<sup>2</sup> In the large majority of cases enlargement does not occur. This progression, when present, is usually slow and almost never reaches the macula. In the same long-term study of 218 consecutive eyes with retinoschisis followed for an

Clinical Professor (Emeritus) of Ophthalmology University of California School of Medicine Los Angeles, CA, USA

Correspondence: NE Byer, MD PO Box 1036 Torrance CA 90505, USA Tel: 310 378 5540 Fax 310 326 2266 E-mail: nebyer@ earthlink.net average of 9 years, no case of macular involvement occurred.<sup>2</sup> There is controversy regarding the incidence of this complication. A poll taken in the Retina Society, reported in 1996,<sup>3</sup> resulted in 87 cases having been observed. However the American ophthalmic literature includes no more than seven published cases of proven macular involvement.<sup>4–7</sup> The well known report by Okun and Cibis in 1964<sup>4</sup> of eight cases was later corrected to only three cases which were caused by senile retinoschisis.<sup>8</sup>

Some think that treatment should be given to far posterior cases, but it is not possible to know which cases should be treated. Treatment of cases which are close to the macula is also attended with a risk of extension, or of new breaks or retinal detachment or maculopathy caused by the treatment itself. It is also undoubted that the very serious complication of irreversible proliferative vitreoretinopathy (PVR) has occasionally occurred as the result of electing to treat asymptomatic retinoschisis, and that this complication has been under-reported in the literature.

It appears that no meaningful justification for treatment has been established. Retinoschisis in a posterior location should only be observed every 2 or 3 years unless the patient notices symptoms of extension caused by an associated detachment.

#### (II) Retinal breaks in one or both layers

It is generally agreed that breaks in the outer layer are the ones that are of primary significance in relation to later schisis-detachment and progressive, frank retinal detachment. The published prevalence of such breaks in clinical studies is as follows: Shea *et al*<sup>9</sup>—17%; Hirose *et al*<sup>10</sup>—24%; and Byer<sup>2</sup>—11%. Somewhat higher figures have been reported in autopsy studies: Foos<sup>11</sup>— 30%; and Göttinger<sup>12</sup>—56%.

In 1972 Cox and Gutow<sup>13</sup> reported a series of 41 eyes of referred patients in which there were 45 separate areas of retinoschisis, each of which also had outer layer breaks. Of this group 27 (60%) also had a schisis-detachment. In 1978 in an autopsy series, Göttinger<sup>12</sup> reported 17 eyes with retinoschisis and outer layer breaks, 10 (58%) of which had schisisdetachment. In 1986 Byer<sup>2</sup> also reported in a clinical study a prevalence of schisis-detachment of 58% among 24 eyes with outer layer breaks. Several reports of large outer layer breaks involving up to 135 arcuate degrees suggest that such eyes may be more susceptible to secondary progressive retinal detachment.<sup>14–16</sup> However in 1995, in attempting to diminish this danger, Clemens et al<sup>17</sup> reported that in retinoschisis with outer layer breaks larger than one hour in size, treatment with cryotherapy actually

caused progression to a schisis-detachment in 11% of 52 eyes.

The proper management of eyes with outer layer breaks depends upon the degree to which such breaks predispose the eye to later loss of vision as the result of other complications such as localized schisis-detachment or progressive retinal detachment. In 1972 Hirose *et al*<sup>10</sup> reported 35 eyes with outer-layer breaks which did not lead to retinal detachment for follow-up periods up to 13 years.

There has not been a consensus about whether outer layer breaks should be treated or observed. The best basis for management must come from natural history and documentation of the actual outcome of these breaks, left untreated, over an extended duration of time.<sup>2</sup> Such studies indicate the relative safety of simply observing outer layer breaks without treatment. However some are again advocating treatment of the outer-layers in such patients with mild laser applications while avoiding treating the borders of the breaks themselves.<sup>18,19</sup> There are still insufficient data to substantiate the safety of this procedure.

Even the high potentiality of 58% to develop a schisis-detachment from untreated outer layer breaks does not, of itself, mandate urgent, or even eventual, treatment of outer layer breaks. This decision must be predicated on further information about the prognosis of schisis-detachment for causing subsequent progression and damage to vision.

### (III) Schisis-detachment

The transition from retinoschisis with outer layer breaks to the stage of schisis-detachment is the result of transfer of some of the less viscous intracystic fluid to a location beneath the outer layer via an outer layer break, thus creating an area of true retinal detachment. Even this change does not of itself merit urgent treatment. I was able to find only eight documented cases contained in two reports in the American literature<sup>14,20</sup> of eyes with definite extension of 'schisisdetachment' posteriorly, beyond the border of the retinoschisis. However in two of these the extension did not reach the macula.

There are four clinical clues which point to the presence of a 'schisis-detachment'.

- First an outer layer retinal break must be present.
- The *appearance of the schisis may not be uniform*, with one part being more elevated than another part, or the texture of the schisis may appear different in one part, appearing more transparent in one area than in another.
- There may be a yellowish line deep to the inner layer,

and usually in contact with an outer-layer break, and which in some way is related to its being elevated. It is produced by the outer layer, and indicates its location.

• Finally there may be a *pigmentation line*. This is produced only when there is an actual detachment of the outer layer which has remained unchanged for a period of time. All of these findings do not need to be present, and two or three are sufficient to make the diagnosis. The presence of a 'schisisdetachment' often is subtle and may be difficult to identify. Its recognition is enhanced by the use of stereoscopic photography.

Ambler *et al*<sup>21</sup> prefer the term 'retinoschisisdetachment' to 'schisis-detachment', and reported four eyes in which this form developed a rare complication characterized by a shallow, relatively small extension of subretinal fluid posterior to the posterior border of the retinoschisis and which progressed in area extremely slowly. These cases involved the macula, becoming symptomatic.

While it is very unusual to be able to identify a case of 'schisis-detachment' which shows posterior extension beyond the posterior schisis border, it is relatively easy to find examples of cases which are situated quite far posterior, some with quite large posterior layer breaks, but which then have become stationary in that position.

Here are some clinical examples of 'schisis-detachment':  $^{\rm 22}$ 

A 43-year-old woman (Figure 1) had this asymptomatic 'schisis-detachment', with retinal breaks in both layers in the superior-temporal quadrant, and was only observed with no change, and without treatment for 2 years.

A 44-year-old woman (Figure 2) had this asymptomatic mid-peripheral 'schisis-detachment' in the superior temporal quadrant, showing a yellow line which marks the outer-layer, which has come forward to lie in apposition to the inner layer as it has detached from the pigment epithelial layer. This was observed with no change in appearance or treatment for 9 and a half years.

A 57-year-old man (Figure 3) had three separate asymptomatic 'schisis-detachments', involving both eyes. This one in the superior temporal quadrant shows a prominent yellow line, marking the position of the outer layer which is also detached. It was observed with no change in appearance or any treatment for 6 years.

A 57-year-old man (Figure 4) had this asymptomatic posterior 'schisis-detachment' which extended to 3 DD from the macula. It also showed retinal breaks with a rolled yellow border and a pigment line. It was observed for 5 years without treatment, during which time it became much flatter. 361

A 49-year-old man (Figure 5) had a bullous 'schisisdetachment' temporally in the right eye, extending posteriorly to 3 DD from the macula. The large outerlayer breaks had a rolled border and later a pigment line appeared. Over the next 12 years the bullous appearance flattened from a height of >2 DD to almost completely flat, and the posterior border receded from a point 3 DD to about 8 DD from the macula. It was observed without treatment for 18 years.

A 40-year-old woman (Figure 6) had bilateral asymptomatic 'schisis-detachments'; in the left eye extending to 4.5 DD from the macula, and in this right eye, to a point 2.5 DD from the macula, and showing this prominent rolled border of the large break. This was observed for 21 years without treatment, but showed a moderate lateral enlargement.

In 1986 a consecutive series of 13 eyes with schisisdetachment were followed for up to 21 years (average 6 yrs), and none progressed to clinical retinal detachment, or threatened the macula, and none was treated.<sup>2</sup> The prognosis of schisis detachment appears to be very good, and in almost all cases they remain localized, non-progressive and asymptomatic. In 1972, Cox and Gutow<sup>13</sup> reported 25 cases of 'schisisdetachment' with 100% success in reattaching the outer layer, using a variety of modalities, including scleral buckling, diathermy, cryotherapy and photocoagulation.

It is of basic importance to remember that the typical behavior of schisis-detachments is that only a limited amount of fluid passes into the subretinal space, probably because of the very viscid nature of the intracystic fluid. Therefore the area of the outer layer detachment remains beneath the retinoschisis, and is also asymptomatic and non-progressive. However, especially if very large outer layer breaks occur a frank, progressive and symptomatic retinal detachment may result. But it has been shown that the ratio of the occurrence of asymptomatic 'schisis-detachment' to the progressive symptomatic variety which requires surgery is about 178 to 1.<sup>2</sup> It is a very important clinical responsibility to differentiate between these two entities because of the vastly different management required.<sup>2,20</sup> Even though progressive symptomatic detachments are much less common, they are treated much more often than 'schisisdetachments'. This is simply because they produce symptoms which cause them to be discovered, in great contrast with 'schisis-detachments' which remain hidden and undiagnosed.



Figures 1–6 (1) A 43-year-old woman with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 123). (2) A 44-year-old woman with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 121). (3) A 57-year-old man with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 122). (4) A 57-year-old man with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 122). (4) A 57-year-old man with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 113). (5) A 49-year-old man with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 117). (6) A 40-year-old woman with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 117). (6) A 40-year-old woman with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 117). (6) A 40-year-old woman with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 117). (6) A 40-year-old woman with 'schisis-detachment' (Courtesy of NE Byer, *The Peripheral Retina In Profile—A Stereoscopic Atlas*, Criterion Press, 1982; p 114).

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# (IV) Symptomatic, progressive, frank retinal detachment

This is a rare complication of senile retinoschisis occurring in about 0.05% of cases, or affecting one in 2000 patients.<sup>2</sup> It is also the *only* complication for which surgery is mandatory (with the exception of the still more rare posterior extension of subretinal fluid beyond a 'schisis-detachment' just discussed).

The detachment may be associated with breaks in both layers, or with only outer layer breaks. If these are located in the periphery, they may be treated with scleral buckling, drainage of subretinal fluid, and cryotherapy.

Even in 1973, using primarily scleral buckling, external drainage of subretinal fluid, and cryotherapy, successful reattachment could be reported in 96% of cases.<sup>23</sup> But now this is usually done only for more peripheral breaks. However even with large midperipheral breaks with posterior extension of subretinal fluid, using only demarcation treatment has been reported with success.<sup>16</sup>

Especially if outer-layer holes are large and posterior, newer procedures offer a greater choice of techniques and success can be achieved with various methods. These will not be discussed in this presentation but only summarized. They include vitrectomy, external drainage of the schisis cavity, inner layer retinotomy and internal drainage of schisis fluid and subretinal fluid, endolaser, gas-fluid exchange with long-acting gases with postoperative positioning of the patient, and use of heavy perfluorocarbon liquids to tamponade the schisis and detachment during surgery.14,15,20,24-26 None of these reports record more than a very few cases.

It is obvious that the best method of treating rhegmatogenous retinal detachment associated with senile retinoschisis cannot yet be said to be agreed upon. It is also clear that a variety of surgical techniques have yielded successful results. We may summarize by offering the following general guidelines for treatment:

## Stage of senile retinoschisis

# Management

- without breaks
- no treatment (except rarely) with outer-layer breaks no treatment (except rarely)
- with localized 'schisisno treatment (except rarely) detachment'
- with progressive, prompt surgical repair symptomatic RD

# Conclusion

In surveying the broad spectrum of retinoschisis as it exists in the population as a whole, we may conclude that the best management in summary is to do nothing except in very rare instances. When progressive retinal detachment appears it must be treated promptly. At almost all other times, treatment is a matter of surgical judgment based upon assumptions. An appeal for conservatism is in order because of the propensity not to progress and the frequent tendency actually to regress. No doubt the statement of Claude Bernard is worth our thoughtful reflection, 'True science teaches us to doubt, and in ignorance, to refrain.'

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