ELSEVIER

Late-onset Retinal Detachment Associated with Regressed Retinopathy of Prematurity

Hiroko Terasaki*,† and Tatsuo Hirose*

*Schepens Retina Associates, Schepens Eye Research Institute, Harvard Medical School, Boston, MA, USA;
†Department of Ophthalmology, Nagoya University School of Medicine, Nagoya, Japan

Purpose: To study the characteristics of late-onset retinal detachments in patients with regressed retinopathy of prematurity (ROP) and the condition of their fellow eyes.

Methods: We carried out a retrospective review of 29 patients (38 eyes) who had been treated at two institutions, one in the US and the other in Japan, between 1986 and 1997. The age at the time of treatment ranged from 6 to 51 years (mean = 23.1). Five of the 38 eyes with tractional detachment were treated with either open-sky vitrectomy, closed vitrectomy, or scleral buckling; 27 of the 38 eyes with rhegmatogenous retinal detachment underwent scleral buckling or closed vitrectomy or both. The remaining 6 of the 38 eyes had subclinical rhegmatogenous detachment and were treated with photocoagulation or cryopexy, or followed without treatment. The most characteristic retinal breaks were multiple holes with a prevalence of equator and posterior types.

Results: Overall, anatomical reattachment was accomplished in 27/32 eyes (84%) that underwent surgery. Two thirds of the patients who underwent vitrectomy either initially or at a later time had poor postoperative visual acuity. More than half of the fellow eyes had retinal detachment and others had various characteristic fundus changes of regressed ROP.

Conclusions: Long-term, probably life-long follow-up of high-risk patients is necessary so that diagnosis and treatment can be instituted at an early stage of retinal detachment. **Jpn J Ophthalmol 2003;47:492–497**

Key Words: Retinal detachment, retinopathy of prematurity, surgery.

Introduction

Recent advances in neonatal intensive care have saved many of the extra-small premature infants. This has resulted in an increase in the number of small children with potential visual problems. Cryotherapy or laser photocoagulation has stopped the progression of threshold retinopathy of prematurity (ROP), thus preventing retinal detachment in many children. In others that have not reached the threshold, ROP has regressed spontaneously to the cicatricial stage.

Retinal detachment is a less frequent complication of the cicatricial stages of ROP than myopia, macular displacement, amblyopia, and nystagmus. Nevertheless, retinal detachment is a vision-threatening complication.[1–6] Before the development of vitrectomy, rhegmatogenous retinal detachment in cicatricial ROP was treated with scleral buckling procedures with reasonable success.[3] With the development of vitrectomy, surgeons are operating in more severe ROP retinal detachment cases that could not have been treated by scleral buckling. Literature on this topic is scanty. Sneed et al[7] reported that retinal detachment with significant traction and/or posterior retinal breaks may be treated by closed vitrectomy in conjunction with scleral buckling.

This retrospective analysis was prompted by the authors' initial impression of rather unfavorable visual results with vitrectomy in ROP related to retinal detachment as compared with retinal detachment not associated with ROP.

Materials and Methods

Twenty-nine patients (38 eyes) 6 years or older with late-onset retinal detachment associated with regressed ROP and the condition of their fellow eyes were reviewed retrospectively from the patient charts at the

Received: August 15, 2002
Correspondence and reprint requests to: Hiroko TERASAKI, MD, Department of Ophthalmology, Nagoya University School of Medicine, 65 Tsuruma-cho, Showa-ku, Nagoya 466-8550, Japan. Tel.: +81-52-744-2277; fax: +81-52-744-2279; E-mail: terasaki@med.nagoya-u.ac.jp

0021-5155/03/$–see front matter
doi:10.1016/S0021-5155(03)00088-1

Schepens Retina Associates in Boston, MA, USA (26 patients, 33 eyes), and from records at the Department of Ophthalmology of the Nagoya University, Nagoya, Japan (3 patients, 5 eyes). These patients had been treated at these institutions between 1986 and 1997. All cases with a clinical diagnosis of retinal detachment and a birth history consistent with ROP were included.

The ages of the patients at the time of surgery or cryo/laser treatment for retinal detachment ranged from 6 to 51 years (mean ± SD, 23.1 ± 12.6 years). The age at their initial visit ranged from 10 months to 50 years. The gestational age at birth of the 26 patients documented on the charts, ranged from 23 to 32 weeks, and their birth weight ranged between 694 to 2250 g (mean ± SD = 1199 ± 385 g). A 2250-g baby was an ''outlier'' who was born prematurely at 32 weeks of gestation. Patients were followed for 1–26 years (9.6 ± 7.6 years) after treatment. The preoperative mean refractive error noted in their charts for 23 eyes was 6.75 ± 3.60 diopters (spherical equivalent). Four eyes had significant cataract before retinal detachment surgery and 26 eyes had clear lenses.

The choice of operative procedure—buckle versus vitrectomy, closed versus open-sky vitrectomy—was determined by the surgeons. Generally, scleral buckling was chosen first. When the buckle was considered ineffective, closed vitrectomy was performed. When the closed vitrectomy was thought less effective, open-sky vitrectomy was chosen. The latter cases were those in which the traction was particularly severe with dense white membranes or even fibrous mass in the extreme periphery. The retina was pulled much forward toward the lens and the stretched, detached retina was adherent to the retrolental fibrous membranes.

Of the 38 eyes with tractional detachment, 5 eyes (13%) were diagnosed with tractional detachment with no retinal breaks, and the other 33 eyes (87%) had rhegmatogenous detachment. The initial procedure for the five tractional detachment cases was open-sky vitrectomy in 2 eyes, closed vitrectomy in 1, and scleral buckling in 2. Of the 33 eyes with rhegmatogenous detachment, 27 eyes underwent major surgery (Table 1). Four eyes underwent cryotherapy and/or laser photocoagulation for subclinical retinal detachment. Very localized retinal detachments with holes in 2 eyes were observed without treatment. In 5 eyes that had closed vitrectomy, 4 were associated with posterior and/or anterior proliferative vitreoretinopathy (PVR); Grade CA, CP-12 (according to the classification for PVR[8]) in 3 eyes, and Grade CP-2 in 1 eye. The remaining 22 eyes including 2 eyes that had PVR Grade B and 1 eye that had Grade CP-1, underwent scleral buckling as the initial procedure.

Table 1. Type of Retinal Detachment and Initial Surgical Procedure

	Initial Surgical Procedure*			
Type of Detachment	OSV	CV	SB	Total Eyes
Tractional	2	1	2	5
Rhegmatogenous	0	5(4)	22(3)	27(7)
Total	2	6	24	32

*Values in parentheses indicate proliferative vitreoretinopathy.
OSV: open-sky vitrectomy, CV: closed vitrectomy, SB: scleral buckling.

Twenty-one eyes (55%) had had previous surgery (Table 2). Eight eyes had cryotherapy or laser photocoagulation or both for threshold ROP. Two eyes had laser iridotomy for glaucoma, and 8 eyes had cataract surgery (3 eyes had intraocular lens implantation followed by YAG laser capsulotomy). The time from the cataract surgery to the retinal detachment surgery ranged from 8 months to 24 years (median = 3 years). Five eyes had a history of scleral buckling and 1 of them had closed vitrectomy later after scleral buckling before the patients came to see us.

Results

The characteristics of the retinal breaks are listed in Table 3.

Number of Retinal Breaks

Twelve of the 33 eyes (36%) with rhegmatogenous detachment had a single retinal break, including one giant tear, and 17 (52%) had multiple retinal breaks. The retinal break could not be detected in 4 eyes (12%); however, the detachment was considered rhegmatogenous from the clinical features and surgical history.

Types of Retinal Breaks

The most characteristic retinal breaks were retinal holes in 20 of the 29 eyes (69%) and retinal tears with traction in

Table 2. Previous Surgeries (21 of 38 Eyes)

Cataract surgery	8*
Cryo and/or laser photocoagulation	8
Laser iridotomy	2
Scleral buckling (SB)	4
SB followed by closed vitrectomy	1

*One eye underwent lens aspiration followed by scleral buckling after 5 months. Another eye underwent lensectomy combined with closed vitrectomy.

Jpn J Ophthalmol
Vol 47: 492–497, 2003

Table 3. Characteristics of Retinal Breaks in Patients with Possible Rhegmatogenous Retinal Detachment

No.	Eyes	Type	Eyes	Location	Breaks	Depth*	Breaks
1	12	Hole	20	Temporal	27	Post	13
2	7	Tear	8	Nasal	9	EQ	18
More than 3	10	Both	1	12 o'clock	2	Ora	8
Undetected	4			6 o'clock	1		
Total	33		29		39†		39†

*Post: more posterior to equator, EQ: around equator, Ora: ora zone.
†The eyes with 2 breaks or more are counted twice or more.

8 eyes (28%). In one eye (3%), both types of break existed. Six of the 9 eyes with retinal tears had cryotherapy during the active stage of ROP. All 6 eyes showed a visible vitreous membrane attached near the periphery of the tear.

Location of the Retinal Breaks

In the 29 eyes, there were 39 retinal breaks with single or multiple retinal breaks in one quadrant or in separate quadrants. Retinal breaks were located in all quadrants of the eye but most commonly in the temporal quadrants (27/39, 69%) (Table 3). Retinal breaks were located around the equator in 18 of the 39 breaks and more posterior to the equator in 13 of the 39 breaks. Eight breaks were located in the oral zone.

Surgical Results

The type of the initial surgery performed by us, including reoperation (the primary surgery had been done elsewhere) consisted of scleral buckling (24 eyes), closed vitrectomy (6 eyes) and open-sky vitrectomy (2 eyes) (Table 4).

The retina was reattached after the first operation in 19 of 32 (59%) eyes, including those eyes with tractional (5 eyes) and rhegmatogenous detachment (27 eyes) and, with additional surgery, anatomical reattachment was accomplished in 27 of the 32 eyes (84%).

Retinal reattachment was achieved with open-sky vitrectomy in 2 of 2 eyes with tractional detachment (100%). Two of 6 eyes (1 with tractional and 5 with rhegmatogenous detachment) were successfully reattached after single closed vitrectomy, while the other 4 eyes were followed by a second vitrectomy that resulted in a final reattachment in these 4 of 6 eyes (67%). With the scleral buckling procedures, 15 (2 eyes with tractional and 13 eyes with rhegmatogenous detachment) of 24 eyes (62%) had successful reattachment with a single operation, 2 of the 24 required a revision of the buckling, and 7 were followed by vitrectomy. In these 7 eyes, 3 had abnormal leaky retinal vessels with yellow exudate before and after scleral buckling procedure. In the end, successful anatomical reattachment was attained in 21 of the 24 eyes (88%). Of all 38 eyes, 15 eyes were treated by vitrectomy either initially or as a reoperation.

Table 4. Initial Surgical Procedure and Anatomical Results*

Initial Procedure	OSV	CV	SB	Total Eyes
Attached after initial surgery	2/2	2/6	15/24	19/32(59)
Final attachment	2/2(100)	4/6(67)	21/24(88)	27/32(84)

OSV: open-sky vitrectomy, CV: closed vitrectomy, SB: scleral buckling.
*Values in parentheses are percentages.

Visual Results

The preoperative and postoperative corrected visual acuities are summarized in Figure 1. Four eyes treated with laser and/or cryopexy and 2 untreated eyes retained essentially the initial visual acuity during the follow-up period (6 months to 9 years). One eye treated with open-sky vitrectomy had only light perception before surgery and showed no significant change after surgery because of the preexisting optic atrophy and diffuse retinal degeneration. Another eye that underwent open-sky vitrectomy retained a visual acuity of 20/400.

All 6 eyes treated with closed vitrectomy initially had extremely low preoperative vision, except for 1 eye with 20/200, which remained so after surgery. In these 5 eyes, 2 were surgical failures; 1 eye had suffered long-standing retinal detachment with PVR, and the remaining 2 eyes developed tractional macular fold or retinoschisis of the macula that resulted in a macular hole 19 years after surgery. In the 24 eyes treated with scleral buckling followed by revision of the buckle or vitrectomy, 3 eyes were surgical failures. In the remaining 21 eyes, 20 essentially retained or had improved visual acuity. One eye that underwent scleral buckling followed by vitrectomy developed a tractional macular fold and decreased vision postoperatively in spite of reattachment of the retina. Five eyes of 4 patients without a history of cryotherapy had the well-demarcated macular coloboma-like scar. These 5 eyes were treated with open-sky vitrectomy in 1 eye, scleral buckling in 3 eyes, and laser photocoagulation in 1 eye. Their postoperative visual acuity ranged from light perception to 20/70.

The Fellow Eye

The condition of the fellow eyes was also analyzed. Of the 29 patients, 9 patients had bilateral late-onset

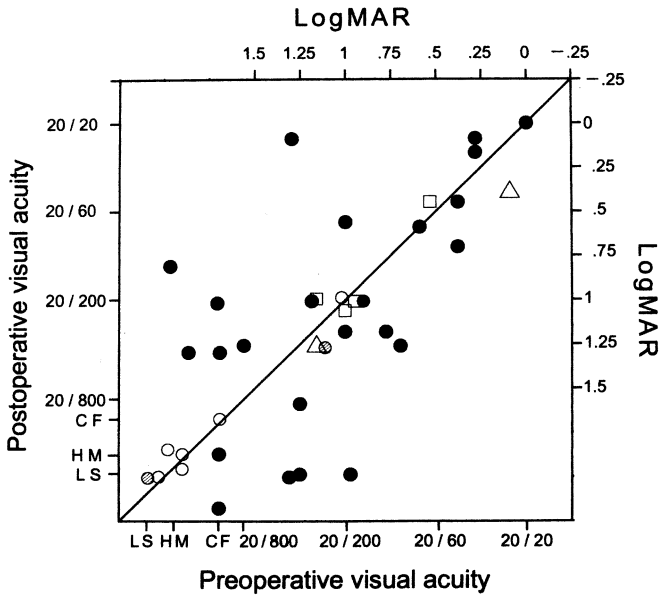

Figure 1. Pre- and postoperative corrected visual acuity classified by the initial procedure. Striped circle indicates open-sky vitrectomy, open circle indicates closed vitrectomy, closed circle indicates scleral buckling, square indicates laser and/or cryopexy, and triangle indicates observation only. LS: sensation of light, HM: hand motion, CF: counting fingers.

retinal detachment, 7 fellow eyes were phthisical, enucleated, or had tractional detachment without light perception. Other findings in the remaining 13 fellow eyes included dragged retina (11 eyes), vitreous membrane (11 eyes), equatorial degeneration such as lattice (7 eyes), retinoschisis secondary to ROP (2 eyes), and macular atrophy (1 eye) (Table 5).

Discussion

One of the main causes of retinal detachment under the age of 16 years is ROP.[1,9] In older patients who have had ROP, rhegmatogenous detachment[1–5] and tractional detachment sometimes occur.[5,6,10] Useful vision has been found in these patients, even with marked posterior changes such as macular displacement and folds.[11] However, a later complication of retinal detachment threatens their vision.

Table 5. Condition of Fellow Eyes (29 Patients)

	No. of Eyes
Phthisis or enucleated	7
Retinal detachment (postop)	9
Other	13
Dense vitreous membrane	11*
Dragged retina	11*
Lattice degeneration	7*
Retinoschisis	2
Macular atrophy	1*

*These categories include same patients.

Diagnosis of retinal detachment that comes acutely with significant subjective symptoms is usually no problem, but there are at least 2 cases in this study in which diagnosis of shallow retinal detachment was delayed in children. Because of heavy pigmentation due to ROP, it was considered that progressive retinal degeneration was the cause of visual failure rather than retinal detachment. They showed limited vision to start with and, because of nystagmus and young age, it must have been difficult to diagnose shallow retinal detachment in the fundus examination. The retinal breaks can be small and may be hidden under the peripheral vitreous membrane. Careful and thorough examination in these patients whose vision continued to decline disclosed retinal detachment. Examination under general anesthesia may be needed in order to detect or rule out retinal detachment in such cases.

Faris and Brockhurst[3] reported that most retinal detachments occurred between the ages of 6 and 15 years, and retinal breaks were round or operculated. Tasman[5] described the characteristics of 53 retinal detachments in the cicatricial stage; 39 of the detachments were rhegmatogenous and 14 were tractional exudative detachments which developed at the mean age of 5.7 years, that is, much younger than the age of the patients with rhegmatogenous detachments. He reported that the mean age of the patients with rhegmatogenous retinal detachment was 13

Jpn J Ophthalmol
Vol 47: 492–497, 2003

years (range from 6 months to 31 years) at the time of diagnosis. In our series, the age of the patients who had tractional detachment ranged from 11 to 24 years, no younger than those with rhegmatogenous detachments, probably because we excluded from the study a large population of stage 5 ROP infants in our practice.

Tasman[5] also reported that the retinal breaks were usually multiple, round or oval, and located on the temporal side along the equator. In our study, the main characteristic type of retinal breaks was small, multiple holes, and holes located around the equator or more posteriorly were relatively common. This contrasts with the other retinal detachments in general, in which the most common location of the break is anterior to the equator.[12]

Our findings showed that 9 eyes of 8 patients had retinal tears and 6 of these eyes had undergone cryotherapy previously. Of the 7 eyes with cryotherapy during the active stage of ROP, 6 eyes had retinal tears with total retinal detachment with or without PVR. The shape of the tear was a slit-like break at the posterior edge of the cryoscar in 2 eyes, crescent-shaped tear in 2 eyes, and large confluent crescents in 2 eyes. Most of these tears were located on the temporal side of the fundus. The patients developing the cryo-related breaks were relatively young (range, 6–21 years; mean = 12 years) compared to the mean age of our total patients. In a previous report,[13] 3 patients with retinal detachment were reported with late complications of trans-scleral cryotherapy. The retinal area treated with cryotherapy usually stayed attached. The posterior edge of the retina, which lacks strong adhesion to the underlying pigment epithelium other than the natural adhesive force, may be more mobile and, with traction, the tear could form at the posterior edge of the cryotherapy treatment. Sneed et al[7] described 14 patients with retinal detachment associated with regressed ROP, and about half were found to have a horseshoe tear, that contrasts with the previous reports of Faris and Brockhurst[3], Tasman[5] and ours, which frequently describe a round hole. It was not clear if these horseshoe tears developed more commonly in the eyes that had been treated by previous cryotherapy.

In our series, 7 eyes underwent cataract surgery 8 months to 24 years before the retinal detachment developed. The results of cataract extraction in 14 eyes of 10 adults with ROP were reported by Krolicki and Tasman.[14] In their series, 1 patient developed retinal detachment more than 6 years after phacoemulsification and intraocular lens implantation and 13 months after YAG capsulotomy. They were uncertain whether the cataract surgery was related to the retinal detachment.

As for treatment options, Tasman[5] indicated encircling buckling with or without vitrectomy for rhegmatogenous detachment. His choice of treatment for tractional detachment was scleral buckling and/or vitrectomy; however, the results in the treated eyes were disappointing. Direct comparison of the surgical results with those of others is nearly impossible, particularly for those results before vitreous surgery became an option.[2,3] Furthermore, our patient population included the most severe cases: about one quarter had phthisis (7/29), one third had retinal detachment (9/29) in the fellow eye. In these cases, particularly with monocular vision, who had had very limited vision before developing retinal detachment, the operations were performed with the clear understanding by the parents and guardians of unfavorable visual prognosis. Anatomical attachment was obtained in patients in whom vitrectomy was performed, but vision was very limited. One of 2 eyes that underwent open-sky vitrectomy and 5 of 6 eyes that underwent closed vitrectomy as a first surgery had vision less than counting fingers preoperatively. The vision did not improve after the operation. Another 2 eyes, which underwent open-sky or closed vitrectomy essentially maintained their preoperative visual acuity of 20/400 and 20/200. The postoperative vision in the remaining 20 of 21 eyes had improved or was unchanged.

Among the 21 eyes with initial buckling surgery and with final reattachment of the retina, one that required vitrectomy as a second procedure had decreased visual acuity after surgery. The difficulty of separating the vitreous cortex, which was strongly adherent to the detached retina in these youngsters, was noted. Furthermore, very dense white fibrous tissue on the retina near the ora serrata cannot be easily removed without the danger of tearing the thin, stretched, detached peripheral retina. Although anterior-posterior traction may be released by vitrectomy, tangential traction may not be entirely released. As a result, when the retina reattaches, it is much more distorted with more prominent dragging of the posterior pole that may result in poor or limited vision. The surgeon has to caution the patient that even if the retina is anatomically reattached by surgery, vision may be much poorer than the level the patient enjoyed before the retina detached. Another unique feature of retinal detachment in some of our patients is abnormally leaky retinal vessels. The fundus of such an eye showed yellow subretinal exudate particularly along the stretched normal as well as abnormal retinal vessels. Probably stretched normal retinal vessels, along with the abnormal vessels leak, causing yellow exudate similar to tractional retinal detachment from other causes such as those associated with familial exudative vitreoretinopathy or posterior type of persistent hyperplastic primary vitreous. These eyes tended to develop intraocular fibrin membrane after extensive or repeated vitreoretinal surgery leading to ocular hypotony

or to the formation of the new membrane that caused redetachment.

Sneed and colleagues[7] recommended pars plana vitrectomy combined with scleral buckling as a first choice if posterior retinal breaks with significant traction or PVR were present. Because of the diversity of our cases, from subclinical retinal detachment to severe tractional detachment with dense fibrous tissue at the peripheral vitreous and PVR, the surgical technique of buckle versus vitrectomy can be chosen, depending upon the severity of the retinal detachment and the surgeon's choice. Generally, however, authors prefer scleral buckle as a first operation unless the buckle is considered ineffective. Saito and associates[15] reported pigmentary maculopathy as a complication to cryotherapy; however, pigmentary maculopathy can occur as a result of severe ROP without a history of cryotherapy.[16] Five eyes of 4 patients in our series had the well-demarcated macular coloboma-like scar; however, none had been treated by cryotherapy.

Retinal detachment in ROP has the tendency to affect both eyes in a large number of cases. In our series, more than half of the fellow eyes suffered retinal detachment, and the unaffected eyes had various characteristic fundus changes of regressed ROP. This study suggests that patients with regressed ROP with significant vitreoretinal changes are at high risk for retinal detachments and that long-term follow-up is necessary to enable us to treat the patients at an early stage of retinal detachment. Peripheral localized retinal detachment without symptoms may stay as it is for a long time. Careful follow-up is needed. The surgical management of complicated retinal detachment has advanced considerably recently, and surgery should be performed before the retina suffers extensive irreversible damage.

This study was supported by Grant-in-Aid No 14370557 from the Ministry of Education, Science and Culture (H. Terasaki).

References

1. Winslow R, Tasman W. Juvenile retinal detachment (rhegmatogenous retinal detachment). Int Ophthalmol Clin 1976;16:97–105.
2. Tasman W, Annesley W. Retinal detachment in the retinopathy of prematurity. Arch Ophthalmol 1966;75:608–614.
3. Faris BM, Brockhurst RJ. Retrolental fibroplasia in the cicatricial stage. The complication of rhegmatogenous retinal detachment. Arch Ophthalmol 1969;82:60–65.
4. Winslow RL, Tasman W. Juvenile rhegmatogenous retinal detachment. Ophthalmology 1978;85:607–618.
5. Tasman W. Late complications of retrolental fibroplasia. Ophthalmology 1979;86:1724–1740.
6. Machemer R. Late traction detachment in retinopathy of prematurity or ROP-like cases. Graefes Archive Ophthalmol 1993;231: 389–394.
7. Sneed SR, Plido JS, Blodi CF, Clarkson JG, Flynn HW, Mieler WF. Surgical management of late onset retinal detachments associated with regressed retinopathy of prematurity. Ophthalmology 1990;97:179–183.
8. Machemer R, Aaberg TM, Freeman M, Irvine AR, Lean JS, Michels RM. An updated classification of retinal detachment with proliferative vitreoretinopathy. Am J Ophthalmol 1991;112:159–165.
9. Scott IU, Flynn HW Jr, Azen SP, Lai MY, Schwartz S, Trese MT. Silicone oil in the repair of pediatric complex retinal detachments: a prospective, observational, multicenter study. Ophthalmology 1999;106:1399–1407.
10. Bradford JD, Trese MT. Management of advanced retinopathy of prematurity in the older patient. Ophthalmology 1991;98:1105–1108.
11. Ferrone PJ, Trese MT, Williams GA, Cox MS. Good visual acuity in an adult population with marked posterior segment changes secondary to retinopathy of prematurity. Retina 1998;18:335–338.
12. Schepens CL. Retinal detachment and aphakia. Arch Ophthalmol 1951;45:1–17.
13. Greven CM, Tasman W. Rhegmatogenous retinal detachment following cryotherapy in retinopathy of prematurity. Arch Ophthalmol 1989;107:1017–1018.
14. Krolicki TJ, Tasman W. Cataract extraction in adults with retinopathy of prematurity. Arch Ophthalmol 1995;113:173–177.
15. Saito Y, Harukawa Y, Lewis JM, Koike H, Omoto T, Tano Y. Macular coloboma like lesions and pigment abnormalities as complications of cryotherapy for retinopathy of prematurity in very low birthweight infants. Am J Ophthalmol 1996;122:299–308.
16. Palmer EA. The continuing threat of retinopathy of prematurity. Am J Ophthalmol 1996;122:420–423.