部修正して，『未熟児網膜症厚生省新分類』[7]となり，現在広く使われている．これと前後して，わが国を含む未熟児網膜症の研究者が集まって国際分類作成を行い，1984年[8]と1987年[9]に発表された．厚生省分類と国際分類はstage 1とstage 2の扱いが異なる．わが国では厚生省分類が広く定着しているが，国際分類への書き換えは容易である．両分類の比較を図6に示す．

国際分類では検査結果をコンピュータに入力可能にできるようにするため，stageを5期に分け，眼底を三つのzoneに分けて病変の局在と範囲を記載するようにした（図7）．一方，厚生省分類では急速に進行して網膜剝離に至る重症網膜症をⅡ型としているが，国際分類ではこの概念がない．後極部静脈の怒張，動脈の蛇行，虹彩血管の充血や瞳孔強直が高度な場合は網膜症の進行が早く重篤なので，これを“plus” diseaseとして，『＋』の記号をつけるようにしているが，厚生省分類Ⅱ型とは異なる．最近，欧米でもこのⅡ型が認識されるようになってきた．さらに，わが国では網膜症が寛解し瘢痕を残した場合の瘢痕期分類が作成されているが，国際分類では記載する瘢痕病変の項目のみにとどめている．

未熟児網膜症の発生に関与する因子

網膜血管は周産期に眼底周辺部に達するが，未熟な血管形成部は，数カ月にわたって原始的な毛細血管網から成人の形態に作り変えられ，通常生後2～3カ月に完成する．未熟児で出生した場合，出生と以後の環境の変化に伴うストレスによって，網膜内の発達過程にある毛細血管床が傷害され消失し，そこから新生血管がおこる．したがって，網膜症の発生にもっとも大きく関与する因子は網膜血管の未熟性であり，在胎週数が早いほど，出生時体重が少ないほど重篤である[10,11]．

網膜症では，vascular endothelial growth factorなどの血管新生因子が網膜無血管領域から放出されて血管新生をおこすと考えられている[12,13]．活動期に行われる光凝固や冷凍凝固治療は，この血管新生因子の産生と放出を抑えることが目的である．

酸素投与は網膜症発生の直接の原因ではないが，悪化させる要因である．初期の酸素投与に関する研究で，4週間高濃度酸素にさらされると網膜症の発生率が非常に増加することが示され，以後は酸素投与の厳重な管理や，抗酸化薬の外用，酸素フリーラジカル形成を促す光の曝露からの遮蔽などが行われてきた．しかし，これらの予防法十分な解決策にはならなかった．経皮膚的に連続計測して酸素をコントロールしても網膜症の発生率や重症化を抑えることはできず，ビタミンE投与などによる酸化予防の試みでも，効果に一致した意見はみられない[10,11]．

その他に，呼吸窮迫症候群，交換輸血，敗血症，脳室内出血，栄養や水分投与のアンバランスなど，呼吸や全身環境の異常に関与して網膜症を悪化させる因子として指摘されている[10,11]．

眼底検査

眼底検査の開始時期については，米国で行われた冷凍凝固の多施設共同研究（CRYO-ROP Study）では出生体重1,300 g以下，あるいは1,800 g以下で補助的に酸素投与を行った低出生体重児には，すべてスクリーニング検査を行うことをすすめている．そして出生後7～9週に最初の検査を行えば活動性を有するものの，まだ重症に至っていない網膜症の大部分を発見することができると考えられている[11]．普通は出生後4～9週に初回検査が行われていることが多い．

われわれは，在胎36週未満，出生体重が1,800 g以下，あるいは高濃度酸素使用，手術を

図8　新生児病棟での眼底検査

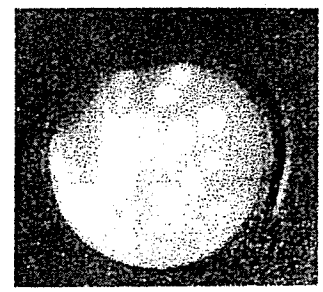

図9　倒像鏡アルゴンレーザーによる凝固斑

行った場合をすべて検査対象としている．これは軽度の網膜血管成長不全をも把握するためで，米国の基準より対象を広めにとっている．検査開始時期は，超未熟児の出生が増加していることからも，全身状態が安定したら，ただちに，遅くとも出生後3週あるいは修正在胎30週前には初回検査を行う．

眼底検査は新生児病棟で行う．眼科医のほかに，患児を抑制する者と，全身状態を観察する新生児科医師の2名の介助が必要である（図8）．

治　療

1．光凝固と冷凍凝固

網膜症が発症しても，厚生省新分類3期初期，あるいは国際分類stage 2までならば自然寛解し，視力予後もよい．しかし，さらに進行すれば網膜凝固を行う．これは無血管領域に汎凝固を行って血管新生因子の産生を抑制し，あわせて新生血管の増殖の場をなくして，網膜剝離発生の可能性を減少させることが目的である．わが国では早くから光凝固が行われており，良好な結果が得られている[12]．米国では，はるかに遅れて，まずCRYO-ROP Studyによって冷凍凝固の有用性が証明され[11]，最近になって光凝固が行われるようになってきた[15]．しかも，米国のCRYO-ROP Studyでは失明予防を目的としているのに比べて，日本ではわずかな瘢痕すらもおこさず，有用な視力を確保することを目的としており，凝固治療の時期が早い傾向にある．

光凝固はキセノンあるいはアルゴン/半導体レーザーによって行う．前者に比して後者のほうが効果は弱いが，古いキセノン光凝固装置をもっている病院はごくわずかとなっている．治療後はできるだけ頻回に眼底検査を行い，不足であれば凝固を追加する（図9）．冷凍凝固は術中の眼球障害のみならず，無呼吸発作や徐脈，血圧低下などの全身合併症をおこす危険性が高い．しかも凝固能が強いので，進行例では後に凝固縁に網膜裂孔を形成し，後の硝子体手術の予後を悪くする．

2．網膜剝離に対する治療（バックリングと硝子体手術）

網膜症がさらに進行して網膜剝離に至った場合，恒久的な視力障害をおこす．これに対しては，まず強膜バックリング手術[16][17]，ついで硝子体手術[18][19]が行われる．バックリングは眼球の外にシリコンスポンジを縫いつけて眼球壁に陥入させ，牽引を軽減させて網膜剝離を治す方法である．しかし，おもに部分網膜剝離に対して行われ，全剝離に向かえば硝子体手術が必要となる．これは，眼内に小さい器具を挿入して網膜を牽引している増殖膜（瘢痕化した新生血管由来の膜組織）を除去し，網膜剝離を治す方法である（図10，11）．

しかし，成人の網膜剝離と比べて非常に重篤なので治癒率は十分とはいえない．しかも，網膜の障害が非常に強いので，剝離が治っても視

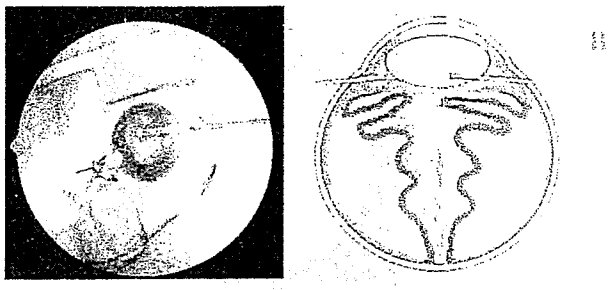

図10　硝子体手術
A：眼球前方の写真，B：眼球シェーマ

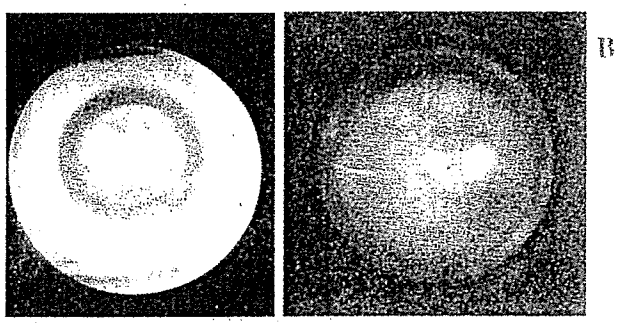

図11　硝子体手術前後
A：術前白色瞳孔の眼球前方写真
B：術後網膜が複位した眼底写真

力は光覚，手動弁程度しか得られないことも多い．硝子体手術は原則として，両眼に高度の剝離がある場合に，片眼ずつ行う．他眼が良好の視力が期待できる場合は，手術はすすめられない．たとえ光覚が得られてもその眼を使わないし，眼球の発育が悪ければ，将来は整容上から義眼を装用することになるからである．

手術時期は，網膜を復位させて視力発達を促すためには早期のほうが望ましいが，本症は手術を急ぐことはむしろ危険である．増殖膜内の血管の活動性が高く，術中に大出血をおこすと止血は不可能なので，瘢痕化が進んで増殖膜中の血管が十分に退縮するのを待ってから手術を行う．通常は網膜剝離がおこってから1～2カ月は待つことが多い．全身麻酔をかけられるか否かも大きな問題である．呼吸器も未熟で，麻酔はかけられても術後に抜管できず長期に呼吸管理をしなければならないこともある．新生児科や麻酔科と十分に相談して手術適応を決める．

晩期合併症に対する検査

光凝固後であれ自然寛解であれ，活動期を乗り切って網膜症が瘢痕化しても，眼底検査を定期的に行わなければならない．晩期合併症として，裂孔原性網膜剝離がおこる危険性がある．瘢痕が軽度であれば10歳代後半におこりやすいが，網膜ひだなど高度な牽引があれば，学童期でも裂孔が生ずる．ことに年少では片眼の視力低下に気づかないので，3～4カ月ごとに眼底検査を行い，眼球を打撲した場合は早期に受診するよう家族にすすめておく．

家族に対する説明とインフォームド・コンセント，ハビリテーション

未熟児網膜症は軽度であれば寛解するが，進行すれば失明につながることもあり，発生初期には予後がわからないことも多い．したがって家族に十分な説明を行っておくことが必要である．網膜症による視覚障害では，米国はもとより，わが国でも多数の訴訟がおこされており，医師は患児の治療のみならず，社会的な問題にも配慮しなければならない．通常，初回の眼底検査の際に，家族に未熟児網膜症の一般について説明し，現在の患児がどの状態にあるかを告げておくべきである．急に光凝固が必要になっても，すでに十分な説明がされていれば家族の納得がただちに得られる．硝子体手術のような予後が十分でない治療を行う場合は，ことにインフォームド・コンセントが重要である．

また，網膜の状態に応じて，できる限り視力を発達させるように努力するべきである．比較的視力が望めるのであれば屈折矯正や訓練などを積極的に行う．不幸にして視覚障害が重篤な場合には，日常生活や就学指導など種々の社会的問題が生ずる．発達遅滞などの重複障害も多

いので，ハビリテーションは専門家との連携のもとにできるだけを早期から行ったほうがよい．

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Early Vitreous Surgery for Aggressive Posterior Retinopathy of Prematurity

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• PURPOSE: To assess the efficacy of early vitrectomy for aggressive posterior retinopathy of prematurity (ROP) to stop progression of retinal detachment.

• DESIGN: Retrospective, noncomparative, consecutive case series.

• METHODS: Twenty-two eyes (15 patients) with aggressive posterior ROP underwent vitrectomy with or without lens sparing, because retinal photocoagulation failed to stop progression of fibrovascular proliferation, despite being performed early, densely, and with early retreatment. We assessed the status of retinal attachment and foveal formation ophthalmoscopically and the presence or absence of fixation of visual behavior.

• RESULTS: Follow-up ranged from six to 12 months (mean, 9 months). Six eyes (100%) in which a lens-sparing vitrectomy was performed developed a large tractional retinal detachment. In contrast, the retinas were completely reattached in 16 eyes (100%) in which vitrectomy with lensectomy was performed, nine eyes (56%) had foveal configuration, and 14 eyes (88%) had steady fixation.

• CONCLUSIONS: These results suggest that early vitrectomy is effective for preventing retinal detachment in aggressive posterior ROP. (Am J Ophthalmol 2006; 142:636–643. © 2006 by Elsevier Inc. All rights reserved.)

IN EYES WITH RETINOPATHY OF PREMATURITY (ROP), visual outcomes are generally poor[1,2] when the retina begins to detach and progresses to Stage 4B or 5. Surgical interventions for retinal detachment associated with progressive ROP, that is, scleral buckling,[3–6] or vitrectomy for Stages 4 and 5,[7–9] usually fail to obtain foveal formation, resulting in poor vision. Recent studies of lens-sparing vitrectomy for Stage 4 ROP have reported retinal reattachment and foveal formation.[10–14]

In contrast to the classical course described by the Committee for the International Classification of ROP,[15] an unusual form of ROP rapidly progresses to a closed funnel of tractional retinal detachment within one to two weeks if left untreated. This severe form is referred to as type II ROP by the Japanese Diagnostic and Therapeutic Criteria for ROP[16,17] or aggressive posterior ROP by the revised International Classification of ROP.[18] Aggressive posterior ROP commonly occurs in zone I and sometimes posterior zone II, with substantial dilation and tortuosity of the vessels of the posterior pole. The flat network of neovascularization on the retinal surface at the deceptively featureless demarcation between the vascularized and nonvascularized area arises circumferentially, usually extends for 12 clock hours, and rapidly extends toward the posterior lens surface. Another characteristic of aggressive posterior ROP is that it may progress to Stage 5 without exhibiting the classical course that includes Stages 1 to 3.

Early intervention with photocoagulation or cryopexy is necessary but often fails to stop ROP progression to Stage 5.[19,20] We report anatomic success after early treatment with vitrectomy in eyes with aggressive posterior ROP.

METHODS

• PATIENTS: All aspects of this study were approved by the institutional ethics committee, and the parents of the patients provided informed consent before the infants were enrollment in the study. We retrospectively reviewed the clinical charts and surgical outcomes of 15 infants (30 eyes) with aggressive posterior ROP. All eyes had previously undergone peripheral laser ablation at our clinic or elsewhere; however, the primary treatment stopped the progression of fibrovascular proliferation in eight eyes. Twenty-two eyes underwent early vitrectomy as a secondary treatment performed by one surgeon (N.A.) in our clinic between July 2004 and August 2005.

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• OCULAR EXAMINATION AND LASER PHOTOCOAGULATION: In our clinic and attendant clinics, the initial ocular screening examinations using binocular indirect ophthalmoscopy were performed as early as possible when systemic conditions stabilized, at the latest at three weeks of chronologic age or 29 weeks of postmenstrual age, in all premature infants less than 34 weeks of gestational age or weighing 1800 g or less at birth, and for all infants who underwent supplemental high-concentration oxygen treatment or surgical intervention. Aggressive posterior ROP, which is designated as type II ROP in Japan, especially should be treated as early as possible at the time of identification.[16–18] The criteria that characterizes the early phase of aggressive posterior ROP by the Committee for the International Classification of ROP[18] and type II ROP by the Japanese Diagnostic and Therapeutic Criteria for ROP[16,17] are the same: (1) posterior location (usually zone I, and sometimes posterior zone II); (2) prominently increased dilation and tortuosity of the posterior pole arteries and veins in all four quadrants; and (3) shunting from vessel to vessel within the retina and not solely at the junction between the vascularized and nonvascularized retina, a flat network of neovascularization at the deceptively featureless junction between the vascularized and nonvascularized retina, or both. Although we usually perform laser ablation of prethreshold retinopathy based on the criteria of the Early Treatment for Retinopathy of Prematurity Study,[21–23] neovascularization in aggressive posterior ROP often progresses rapidly from the intraretinal to the extraretinal areas, circumventing the typical formation of the ridge. Thus vascular shunting sometimes associated with the presence of hemorrhage widely observed within the retina is an important initial sign.[16,17]

All eyes of all patients were treated immediately with argon green laser photocoagulation through an indirect ophthalmoscope (Lumenis, Santa Clara, California, USA) when the initial signs of aggressive posterior ROP were identified. The treatment was applied densely to the nonvascularized retina (duration 200 to 400 ms; power 300 to 400 mW). The adjacent vascularized retinal area posterior to the ridge, which contained marked vessel shunting, also was coagulated, because capillary nonperfusion seemed to be present in the already vascularized retina.[24] Laser photocoagulation often was applied repeatedly to the skip areas, because a remnant of the hyaloid vascular system including the tunica vasculosa lentis interrupts penetration of the laser light. Near-infrared diode laser that sometimes produces excessive coagulation with hardly distinguishable laser spots is difficult to use, when the density of the laser application should be varied on the nonvascularized and vascularized retina. By using argon green laser that is absorbed more by the hemoglobin in the tunica vasculosa lentis, subcapsular cataract does not develop after photocoagulation.[25,26] When fibrovascular proliferation progressed circumferentially for six or more continuous clock hours and tractional retinal detachment occurred simultaneously (Stage 4), the eye underwent vitreous surgery as a secondary treatment.

• VITREOUS SURGERY: In the patients who required secondary treatment, vitreous surgery was performed in each eye separately a few days to one week apart, or sometimes in both eyes on the same day, when ROP was suspected of having progressed rapidly to Stage 5 simultaneously in both eyes,[16–18] or when a lengthy period of anesthesia was more acceptable in the presence of a systemic condition, such as a respiratory disorder, than a repeated period of anesthesia. Four infants (six eyes) underwent a lens-sparing vitrectomy that did not stop the progression of retinal detachment. Using a small contact lens designed for premature eyes, a core vitrectomy was performed in the six eyes.

In the other 11 patients (16 eyes), the lens was removed to perform vitrectomy in the periphery. In these eyes, a three-port vitrectomy was performed using the Accurus 25-gauge surgical system (Alcon, Fort Worth, Texas, USA) that includes cannulas, an infusion pipe, an illumination pipe, an endophotocoagulation probe, a vitreous cutter, scissors, and forceps. After performing conjunctival peritomy, 25-gauge sclerotomies were made 1.0 mm posterior to the limbus through the pars plicata. A wide-field vitrectomy was performed from the posterior pole to the vitreous base in the aphakic eyes. Dissection or removal of the fibrovascular tissues was minimized to avoid bleeding. A fluid-air exchange and endophotocoagulation were performed in three eyes in which an iatrogenic break developed. With the exception of these patients, no specific positioning was used postoperatively. No additional surgical intervention was performed in any of the 16 eyes.

• ASSESSMENT OF SURGICAL RESULTS: The infants were followed for six to 12 months (average, nine months) postoperatively. The preoperative and postoperative clinical charts and wide-field fundus photographs obtained using RetCam (Nidek, Gamagohri, Japan) were reviewed retrospectively. The six-month postoperative anatomic outcomes were determined by binocular ophthalmoscopy and photography with the patients under general anesthesia. Pediatric ophthalmologists assessed the visual behavior using suitable refractive correction. Central fixation was assessed by the corneal light reflex, and steady fixation was assessed using still and moving targets with the fellow eye occluded.

RESULTS

• PATIENT CHARACTERISTICS: Seven of the infants were girls and eight were boys. The gestational ages at birth ranged from 25 to 30 weeks (average, 25 weeks), and the birth weights ranged from 466 to 1676 g (average, 773 g). All but two patients weighed less than 1251 g at

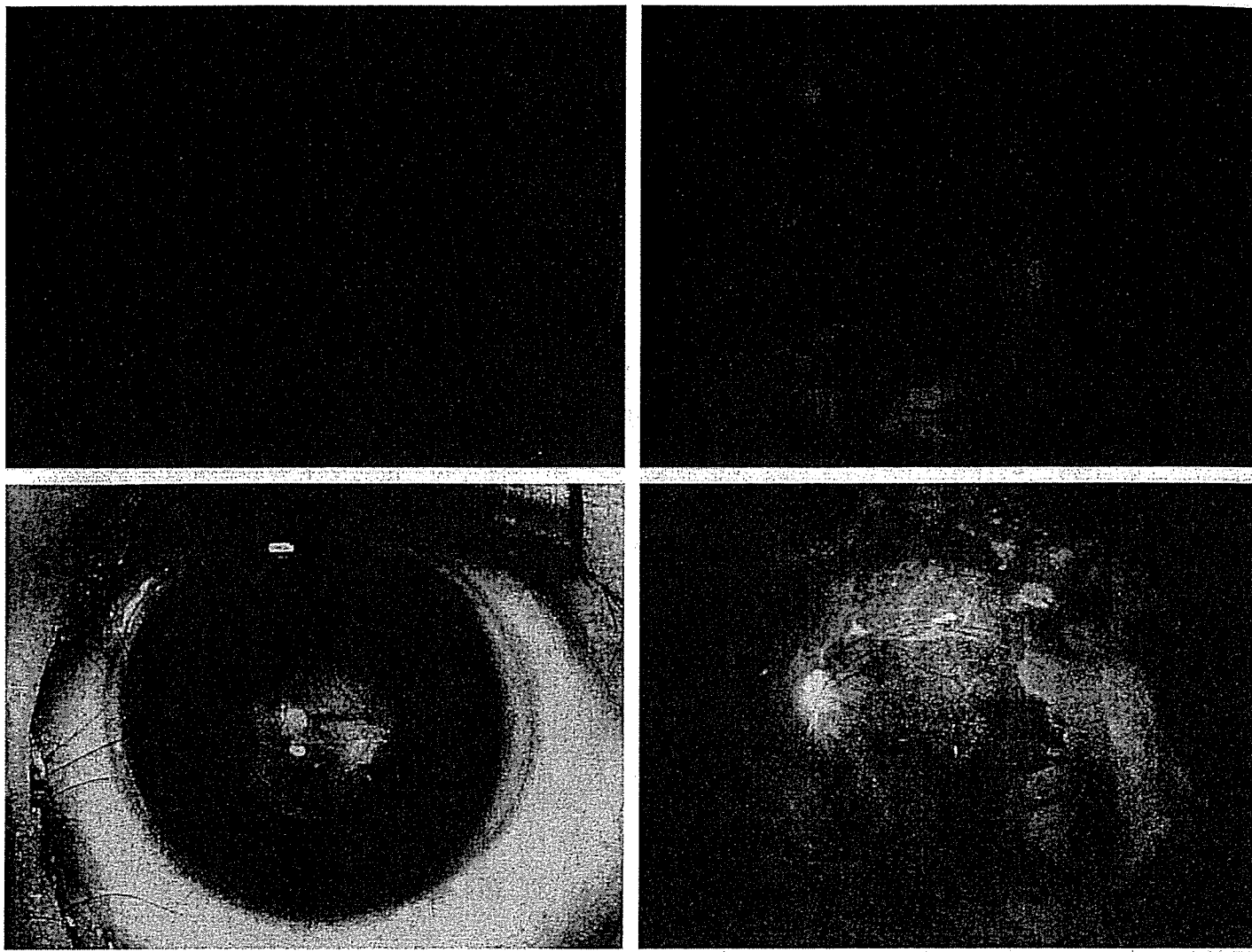

FIGURE 1. Fundus images of fibrovascular tissue (FT) growth and preoperative and postoperative lens-sparing vitrectomy in the left eye of Patient 2 with aggressive posterior retinopathy of prematurity (ROP) (gestational age 22 weeks; birth weight 479 g). (Top left) Ten weeks after birth, when the initial signs of aggressive posterior ROP were identified, the first photocoagulation was performed. (Top right) FT grew from the photocoagulation scars at 18 weeks, despite repeated applications of photocoagulation. (Bottom left) Although an early lens-sparing vitrectomy was performed at 18 weeks, the retinopathy progressed to Stage 5. (Bottom right) The secondary vitrectomy was performed one month after the initial vitreous surgery; however, retinal folds and significant degeneration persist.

birth; however, posterior aggressive ROP also developed in Patient 14 whose birth weight was 1280 g with multiple malformation syndrome and Patient 15 whose birth weight was 1676 g with hydrops fetalis. The initial ocular examination was conducted between one to four weeks (average, three weeks) of chronologic age (27 to 31 weeks of postmenstrual age; average, 28 weeks). At this time, the retina was vascularized within zone I in 22 eyes and in posterior zone II in eight eyes. In all eyes, the ROP extended for 12 clock hours in zone I or posterior zone II and was characterized by prominent dilation and tortuosity of the vessels in the four quadrants of the posterior pole at four to 11 weeks (average, eight weeks) of chronologic age (29 to 36 weeks of postmenstrual age; average, 33 weeks). Argon laser photocoagulation was applied densely to the nonvascularized retina and the adjacent vascularized area that contained prominent vessel shunting two to five times (average, three times) repeatedly during a period of two to seven weeks (average, four weeks) in all 30 eyes, because insufficient regression of the hyaloid vascular system interrupted penetration of the laser light. Photocoagulation stabilized the ROP in eight eyes. In the other 22 eyes, the dilation and tortuosity of the retinal vessels and elevation of the ridge transiently decreased to some extent; however, they increased again, and the fibrovascular proliferation present circumferentially for nearly 12 clock hours then progressed and extended toward the posterior lens surface, and tractional retinal detachment occurred (Figure 1, Top row). Thus the vascularity of ROP remained active at the time of vitreous surgery in all 22 eyes; the fovea was not involved with the retinal detachment (Stage 4A) in 15 of the 22 eyes. The fovea was involved (Stage 4B) in seven

TABLE. Characteristics of Eyes Undergoing Surgery for Aggressive Posterior Retinopathy of Prematurity

Patient No.	Gender	GA (wks)	BW (gr)	Eye	Zone	PMA at PHC (weeks)	PMA at Vitrectomy (weeks)	Stage	FT & VB Adhesion (clock hours)	Surgical Procedures	IOC	POC	Additional Surgical Procedures	Retinal Attachment (Final)	Foveal Formation	SCF	Systemic Complications
1	M	22	466	OD	I	32–39	40*	4A	N	LSV	H	RRD	V	Y	N	N	Hydrocephalus
				OS	I	32–39	40*	4A	N	LSV	H	RRD	V	Y	N	N	
2	M	22	479	OD	I	33–38	39*	4A	N	LSV	H	RRD	V	Y	N	N	Hydrocephalus
				OS	I	33–38	39*	4A	N	LSV	H	RRD	V	Y	N	N	
3	F	26	857	OS	I	34–37	39	4B	N	LSV	H	RRD	V	Y	N	N	
4	F	27	946	OS	II	34–39	40	4A	N	LSV	H	RRD	V	Y	N	N	
5	M	22	470	OD	I	32–38	40*	4B	Y, <3	V	H			Y, DR	Hypoplastic	Y	Hydrocephalus
				OS	I	32–38	40*	4A	N	V	H			Y	Y	Y	
6	M	23	510	OD	I	32–36	37*	4A	N	V	H			Y	Y	Y	
				OS	I	32–36	37*	4A	N	V	H			Y	Y	Y	
7	F	24	526	OS	I	34–38	41	4B	Y, ≥3	V	H			Y, DR	N	Poor	
8	M	23	610	OS	I	36–38	40	4B	Y, ≥3	V, FE, PHC	H, IB			Y, DR	N	Poor	
9	F	23	612	OD	I	32–35	36	4A	N	V	H	OH		Y	Y	Y	
10	M	25	678	OD	I	33–37	39	4A	N	V, FE, PHC	H, IB			Y, DR	Hypoplastic	Y	
11	F	25	798	OD	II	32–35	37	4A	N	V	H			Y	Y	Y	
				OS	I	32–35	36	4A	N	V	H			Y	Y	Y	
12	M	24	803	OD	I	29–33	35	4B	Y, <3	V	H			Y, DR	Hypoplastic	Y	
13	M	26	897	OD	I	34–38	40	4A	N	V	H	OH		Y	Y	Y	Hydrocephalus
				OS	I	34–38	40	4B	N	V, FE, PHC	H, IB			Y, DR	Hypoplastic	Y	
14	F	28	1280	OS	II	36–38	39	4B	Y, <3	V	H			Y, DR	Hypoplastic	Y	Prosoposchisis
																	Hydrocephalus
15	F	30	1676	OD	II	34–36	37	4A	N	V	H			Y	Y	Y	Hydrops fetalis
				OS	II	34–36	38	4A	N	V	H			Y	Y	Y	

GA = gestational age; BW = birthweight; OD = right eye; OS = left eye; M = male; F = female; PMA = postmenstrual age; PHC = photocoagulation; * = vitrectomy was performed in both eyes on the same day; FT = fibrovascular tissue; VB = the ciliary body and peripheral retina at vitreous base; LSV = lens-sparing vitrectomy; V = lensectomy-vitrectomy; FE = fluid-gas exchange; Y = yes; N = no; IOC = intraoperative complications; H = slight hemorrhage; IB = iatrogenic break; POC = postoperative complications; RRD = regrowth of the fibrovascular tissue and retinal detachment; OH = transient ocular hypertension; DR = dragging or folds of the retina; SCF = steady central fixation; retinal attachment (final) = results of initial vitreous surgery; in Patients 5 to 15 and results of the additional vitreous surgery in Patients 1 to 4.

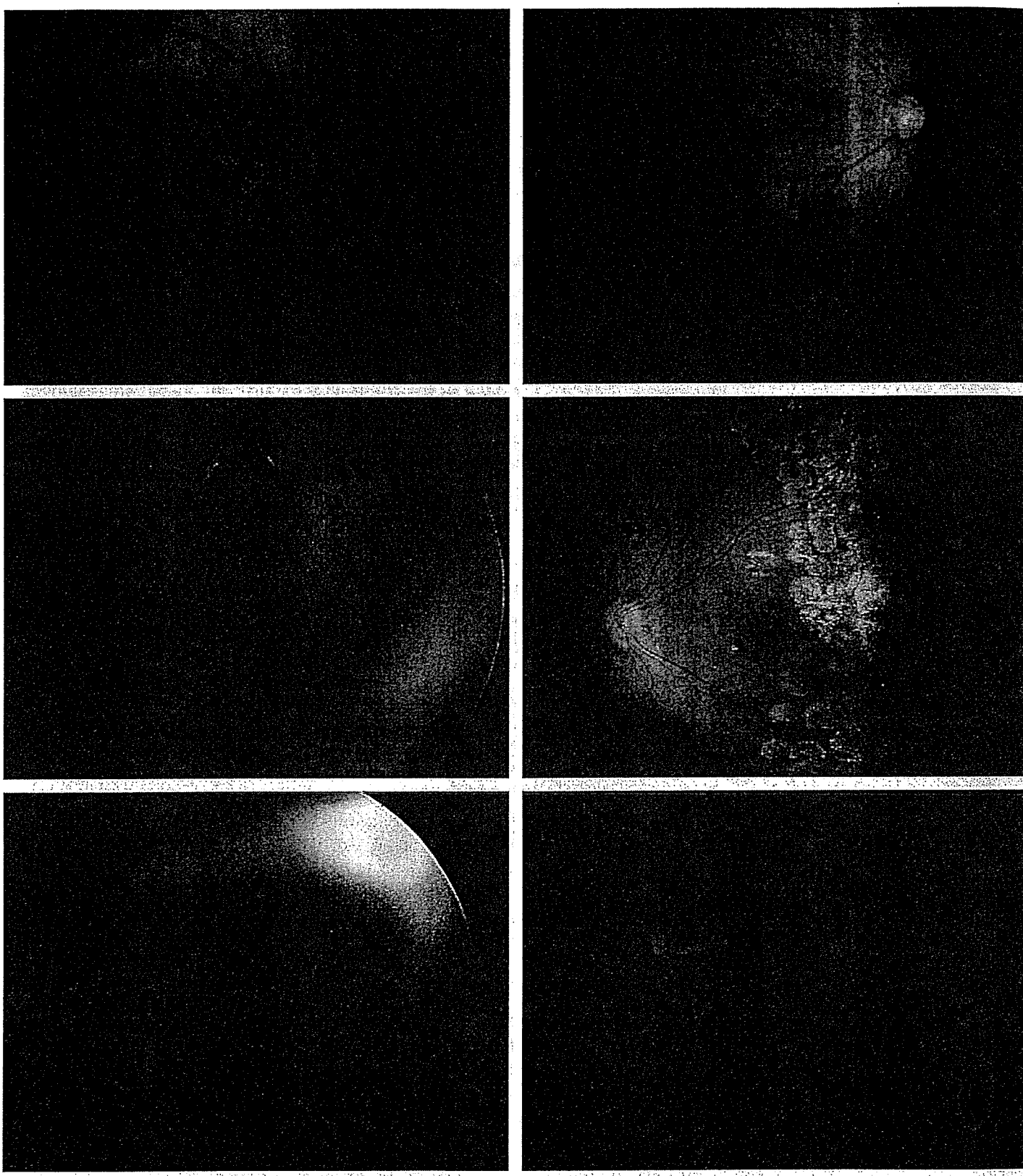

FIGURE 2. Preoperative and postoperative fundus images of aggressive posterior retinopathy of prematurity (ROP) in which early vitrectomy and lensectomy were performed. (Top row) The right eye of Patient 11 (gestational age 25 weeks; birth weigh, 798 g). (Middle row) The left eye of Patient 13 (gestational age 26 weeks; birth weight 897 g). (Bottom row) The left eye of Patient 7 (gestational age 24 weeks; birth weight 526 g). (Left column) Preoperative fundus images. (Right column) Postoperative fundus images. Various configurations of retinal detachment and fibrovascular tissue (FT) in Stage 4 ROP may reflect the outcomes of vitreous surgery. Preoperatively, FT arises in the photocoagulation scars, reaches the posterior lens surface (Top left), extends toward the vitreous base (Middle left), and is attached to the ciliary body and peripheral retina at the vitreous base (Bottom left), under which a regional traction retinal detachment does (Stage 4B) (Middle left, Bottom left) or does not (Stage 4A) (Top left) affect the fovea. The retina has reattached with (Middle right, Bottom right) or without (Top right) residual retinal folds and dragging under the residual fibrous tissue. The dilation and tortuosity of the retinal vessels have decreased postoperatively, and there is little intraoperative and postoperative bleeding in each eye.

eyes; fibrovascular tissue did not extend to the vitreous base in two eyes, was attached to the ciliary body and peripheral retina at the vitreous base for less than 3 clock hours in three eyes and 3 clock hours or more in two eyes (Patients 7 and 8 in Table).

• SURGICAL RESPONSE: In six eyes in which a lens-sparing vitrectomy was performed, fibrovascular tissue (FT) continued to grow along the residual peripheral vitreous, resulting in substantial retinal detachments (Stage 5) (Figure 1, Bottom left). Another vitrectomy with lensectomy was performed one month later when the vascularization became quiet. Retinal reattachment was achieved in all six eyes, but there was substantial retinal degeneration (Figure 1, Bottom right). The visual function of the six eyes was light perception or hand motions.

In contrast, in the 16 eyes in which vitrectomy was easily performed to the vitreous base after lensectomy, complete retinal reattachment was achieved at the last follow-up examination. The retina was dragged around the scarring of the residual FTs in one (10%) of 10 eyes in which ROP had progressed to Stage 4A preoperatively, but in all six eyes in which ROP had progressed to Stage 4B, because the surgery failed to release the traction of the wide circumferential FT that was not dissected or removed (Figure 2, Middle and Bottom row). In three eyes in which an iatrogenic break developed, the retina also was reattached by fluid-air exchange and endophotocoagulation.

Slight vitreous bleeding occurred from the FT during vitrectomy in all eyes but was spontaneously absorbed within two to three weeks postoperatively. The fovea was well formed at the correct retinal position in nine (90%) of 10 eyes with preoperative Stage 4A ROP (Figure 2, Top row) and was mildly hypoplastic in one (10%) of 10 eyes with Stage 4A ROP and four (75%) of six eyes with Stage 4B (Figure 2, Middle row). All 14 eyes had steady central fixation (SCF). However, in two eyes with stage 4B ROP, in which wide circumferential FT was attached to the ciliary body and the peripheral retina at the vitreous base preoperatively, the retina had been dragged extensively, the fovea failed to form, and there was no SCF (Figure 2, Bottom row; Patients 7 and 8 in Table).

No eyes had additional vitreous bleeding, endophthalmitis, rhegmatogenous retinal detachment, or neovascular glaucoma. Two eyes had a transiently high intraocular pressure level that was managed with hypotensive medication. The characteristics of the 22 eyes are summarized in the Table.

DISCUSSION

RETINAL PHOTOCOAGULATION OR CRYOPEXY IS SOMETIMES effective for stabilizing aggressive posterior ROP; however, it sometimes cannot stop the progression to retinal detachment, despite being performed early, densely, and with early retreatment.[19,20] When FT and traction retinal detachment occur 360° circumferentially in zone I, where aggressive posterior ROP usually occurs, scleral buckling is not only difficult to perform but also does not effectively release the traction, because the summit of the buckle cannot be positioned to counteract the direction of the fibrovascular growth.[9,19] Another surgical procedure to remove or divide the encircling buckle then is needed to facilitate ocular growth.[27]

When vitreous surgery is performed for ROP that progresses to Stage 5, postoperative visual outcomes are poor despite successful retinal reattachment.[7–9] Furthermore, vitreous surgery sometimes cannot be performed when aggressive posterior ROP results in the development of corneal opacity, glaucoma, and phthisis soon after progression to Stage 5.[16–18,20]

In the current study, we successfully treated aggressive posterior ROP by early surgical intervention with photocoagulation and vitreous surgery. The fovea was well formed in 56% of the treated eyes, and a good visual outcome was achieved in all but two eyes. These results indicate the great benefit of early surgery for aggressive posterior ROP, in comparison to the poor visual outcomes after vitreous surgery for Stage 5 ROP, despite surgical and anesthetic intervention on very small infants. Even though the vascularity was still active, it was suppressed considerably by retinal photocoagulation that was performed early, densely, and with early retreatment. We did not completely remove the FT but did remove the surrounding vitreous gel, even though there was still retinal traction. Thus, there was little intraoperative and postoperative bleeding.

Surgical removal of the vitreous framework might contribute not only to reduced tractional force of the fibrovascular tissue but also to suppressed growth of new vessels, which is activated by the traction.[3–6] This is similar to outcomes of early vitreous surgery for diabetic retinopathy.[28,29] In patients who are young and have more severe proliferative diabetic retinopathy, there is an obvious advantage to performing early vitrectomy, while no such advantage exists for patients with retinopathy consisting of minimal severe new vessels. Early vitrectomy for diabetic retinopathy is most suitable for eyes in which both fibrous proliferation and at least moderately severe new vessels are present and in those eyes in which extensive scatter photocoagulation has already been applied.[29]

Lens preservation is important to prevent deprivation amblyopia and promote visual development.[30,31] However, our experience with lens-sparing vitrectomy to treat aggressive posterior ROP did not stop the progression of retinal detachment compared with the group in which lensectomy was performed. Fibrovascular tissue arises from the posterior retina, reaches the posterior lens surface, then extends toward the vitreous base, where the vitreous framework is most condensed, and contracts.[1] The space in which the vitreous gel is removed by lens-sparing vitrec-

tomy is limited to the posterior portion of the vitreous cavity.[10,31] Thus lensectomy is necessary to remove the vitreous gel from a wide area including the vitreous base.

There are various configurations of retinal detachment and fibrovascular tissue in Stage 4 ROP, which may predict the outcomes of vitreous surgery. When the fovea is involved in the retinal detachment, early vitreous surgery fails to obtain good foveal formation, probably because the fovea continues to develop after birth.[32] When fibrovascular tissue reaches the posterior lens surface and does not extend toward the vitreous base, the vitreous gel around the FT is easily removed surgically, resulting in good outcomes. In contrast, when the FT extends and attaches to the ciliary body and the peripheral retina at the vitreous base, the retinal detachment usually progresses more and involves the fovea. In this case, cutting the FT and removing the vitreous gel from the vitreous base region are difficult and result in residual retinal folds or a dragged retina and no foveal formation. Thus vitreous surgery may be performed during the period when the FT grows to the posterior lens surface and has not extended toward the vitreous base.

The current study had some limitations in that it was not randomized, controlled, or prospective, the patients had various systemic conditions, and there were various types of retinal vasculature and retinal detachments. In eyes with much poorer retinal vasculature, in which vessels are present only near the optic disk, FTs likely grow on the retinal surface and along the trunk of the hyaloid vessels that arise from the optic disk. Retinopathy that has progressed further with more extensive retinal detachment also may be a contraindication to early vitrectomy, because new vessels secondarily invade and mature in the FT at an early phase of the regression of retinopathy.

Certain logistical problems also accompany early surgery for aggressive posterior ROP. Because aggressive posterior ROP rapidly progresses to Stage 5,[16–18] and there are few surgeons who specialize in vitreous surgery for ROP, prompt transport of infants and preoperative systemic examination and management for anesthesia are necessary. If the surgery is delayed as little as a few days, ROP might progress to Stage 4B or 5, resulting in poor surgical outcomes. The surgery should be completed within a short time, because small premature babies cannot tolerate systemic anesthesia for a long period.[33] Additional anesthesia after a short period sometimes causes edema of the vocal cords or trachea or apnea after extubation that might result in respiratory insufficiency.[33,34] Thus surgery that is performed on both eyes with simultaneous aggressive posterior ROP on the same day may be unavoidable in premature babies who cannot tolerate anesthesia again during the course of a few days. Lengthy surgical procedures also should be avoided, including application of additional photocoagulation, hemostasis, and repairing of iatrogenic breaks. Thus aggressive removal of the FT on the retina also might be contraindicated to avoid bleeding and iatrogenic break formation. Early, wide, and dense application of photocoagulation before vitrectomy is important not only to delay progression of retinal detachment but also to stabilize vascularity of the retinopathy.

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TRANS-TENON RETROBULBAR TRIAMCINOLONE INFUSION FOR CHRONIC MACULAR EDEMA IN CENTRAL AND BRANCH RETINAL VEIN OCCLUSION

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Chronic macular edema associated with central retinal vein occlusion (CRVO) and branch retinal vein occlusion (BRVO) is often refractory to treatment including topical and oral corticosteroids, oral acetazolamide, and grid pattern laser photocoagulation.[1,2] Recently, more invasive therapies, including vitrectomy and intravitreal corticosteroid injections, have been investigated as a means of treating macular edema,[3,4] although these carry risks of severe complications, including vitreous hemorrhage, retinal detachment,

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and endophthalmitis. We previously reported the use of trans-Tenon retrobulbar infusion of triamcinolone acetonide to treat macular edema and other manifestations of uveitis.[5] Here we report on use of the same to treat chronic macular edema in association with CRVO and BRVO.

Methods

Five eyes of five patients with CRVO and 11 eyes of 11 patients with BRVO were studied. The median patient age was 71 years (range, 30–80 years); three patients had a history of diabetes mellitus and seven had hypertension. Four of the CRVO eyes were ischemic, all having received panretinal photocoagulation, and one eye was nonischemic. One CRVO eye had optic disk neovascularization and an additional CRVO eye had retinal neovascularization elsewhere. One BRVO eye had received scatter photocoagulation to nonperfused areas of retina. All eyes had macular edema of greater than 3 months' duration that was unresponsive to topical corticosteroid therapy. Two CRVO eyes had undergone vitrectomy for the macular edema, one with epiretinal membrane peeling in addition, but both eyes were without improvement. No eyes had received grid pattern laser photocoagulation to the macula.

Informed consent was obtained before each procedure. The patient's eye was prepped and draped, with topical instillation of 4% Xylocaine. The conjunctiva and Tenon capsule were incised in the inferotemporal quadrant, followed by insertion to the hub of a 23-gauge curved blunt cannula approximately 2.1 cm in length (#HS-2764; Handaya Co., Tokyo, Japan) into the sub-Tenon space and infusion of 20 mg triamcinolone acetonide (Bristol Pharmaceutical, K.K., Tokyo, Japan). The wound was left unsutured, and 0.5% levofloxacin was instilled three times daily for 1 week.

Patients were examined after the procedure at 2 to 4 weeks, 2 months, 3 months, and 6 months. Eyes were evaluated by measurement of best corrected decimal visual acuity (VA) and intraocular pressure (IOP), slit-lamp examination, dilated funduscopy, fluorescein angiography, and optical coherence tomography (OCT). VA improvement or worsening was defined as a change of 0.2 logMAR or greater. Foveal thickness was defined as retinal thickness at the center of the fovea by OCT, taking the mean of two cross-hair images. Efficacy of treatment was defined as a foveal thickness decreased by 30% or more within 3 months from the pretreatment foveal thickness. Recurrence of macular edema was defined as a 20% or more increase in foveal thickness over the thinnest documented previously. Repeat triamcinolone infusion was considered for recurrence of macular edema or for an inadequate response to the initial infusion, and only in the absence of elevated IOP. Clinical records were reviewed retrospectively.

Case Report

A 73-year-old woman (Table 1, patient no. 2) presented 14 months after the onset of CRVO in her right eye. She had received scatter peripheral retinal photocoagulation at 1 month after onset. At presentation, the VA was 0.2, and dilated funduscopy revealed retinal venous sheathing consistent with an old CRVO, peripheral laser scars, and severe cystoid macular edema (Figure 1A, B). The foveal thickness was 368 μm. Two weeks after triamcinolone infusion, the visual acuity had improved to 0.4 with a foveal thickness of 57 μm (Figure 1C). At approximately 13 months, the visual acuity was unchanged at 0.4 and the foveal thickness was 140 μm. No complications were observed.

Results

Visual acuity and OCT results of each patient are presented in Table 1. The median time between onset of vein occlusion and triamcinolone infusion was 13.9 months (range, 3.8 months to 10 years), and the median post-triamcinolone infusion follow-up period was 7.0 months (range, 3.0–22 months). Before treatment, one eye (6.3%) had a VA of 0.6 or better; 14 eyes (87.5%) had a visual acuity of worse than 0.6 but better than 0.1, and one eye (6.3%) had a visual acuity of 0.1 or worse. After treatment, four eyes (25.0%) had a VA of 0.6 or better; 10 eyes (62.5%) had a VA of worse than 0.6 but better than 0.1, and two eyes (12.5%) had a VA of 0.1 or worse. At the time of best VA and at final examination, VA improvement was documented in 8 of 16 eyes (50.0%). At final examination, two eyes (12.5%) had VA worsening, presumably due to persistent macular edema.

Treatment was judged to be effective in 13 of 16 eyes (81.3%) by 3 months after a single infusion, 5 of 5 CRVO eyes (100%) including the 2 eyes status-post vitrectomy, and 8 of 11 BRVO eyes (72.7%). Of the three eyes in which initial treatment was judged not to be effective, two eyes (both with BRVO) underwent a second triamcinolone infusion at 3 months and at 6 months, but without improvement in the macular edema. Of the 13 eyes in which initial treatment was judged to be effective, 8 eyes (61.5%) had recurrences at 2 weeks to 9 months after treatment. Five of these recurrence eyes received a second triamcinolone infusion, four eyes at 3 months and one eye at 9 months after the initial infusion. One recurrence eye eventually received a total of four infusions over a 1-year, 4-month period. After repeat triamcinolone infusion for recurrence of macular edema, treatment was

Table 1. Visual Acuity and Foveal Thickness after Triamcinolone Infusion

Patient No.	Diagnosis	Duration* (months)	Initial VA	Best VA	Time of best VA† (months)	Final VA	Time to recurrence§ (months)	Number of infusions	Follow-up (months)	Initial FT (μm)	Best FT (μm)	Final FT (μm)
1	CRVO	5.2	0.4	1.0	12	1.0‡	3	4	22	708	249	249
2	CRVO	14	0.2	0.4	0.5	0.4	9	2	13	368	57	140
3	CRVO	8.9	0.2	0.4	1	0.1	1	2	10	509	227	481
4	CRVO	6.1	0.2	0.5	1	0.5	NR	1	6.9	340	57	124
5	CRVO	10.6	0.2	0.4	3	0.4	NR	1	7.0	396	113	150
6	BRVO	23	0.5	0.8	12	0.8	1	2	13	396	170	516
7	BRVO	120	0.7	0.8	0.5	0.7	1	1	4.5	395	226	340
8	BRVO	3.8	0.2	0.5	3	0.5‡	NR	2	12	368	198	556
9	BRVO	14	0.4	0.4	NA	0.4	NR	2	10	340	283	526
10	BRVO	17	0.4	0.5	6	0.4	NR	1	7.0	283	NA	375
11	BRVO	20	0.4	0.6	3	0.5	NR	1	6.2	509	425	828
12	BRVO	23	0.5	0.5	NA	0.5	NR	1	3.1	283	113	150
13	BRVO	5.0	0.2	0.2	NA	0.1	3	2	4.7	509	283	573
14	BRVO	27	0.2	0.2	NA	0.2	1	1	3.3	255	170	210
15	BRVO	16	0.1	0.4	1	0.4	1	1	3.5	379	182	263
16	BRVO	8.8	0.5	0.9	1	0.8	NR	1	3.0	521	131	131
	median	13.9	0.3	0.5	2	0.5			7.0	387	182	302

BRVO, branch retinal vein occlusion; CRVO, central retinal vein occlusion; FT, foveal thickness; NA, not applicable; NR, no recurrence; VA, visual acuity.

* Duration between time of onset and triamcinolone infusion.

† Earliest time at which best VA or best FT was achieved after triamcinolone infusion.

‡ Visual acuity partially affected by cataract progression.

§ Time of first recurrence measured from time of initial triamcinolone infusion.

judged to be effective in two eyes and not effective in three eyes.

Cataract progression was documented in three eyes (18.8%), one eye having received four triamcinolone infusions. IOP elevation, infection, and other complications were not noted.

Discussion

Chronic macular edema remains one of the most difficult to treat sequelae of CRVO and BRVO. Although some eyes improve spontaneously, 58% of eyes with macular edema due to CRVO will have a visual acuity worse than 20/100 after 3 years and less than 20% of eyes will gain two or more lines of visual acuity.[1] The Central Vein Occlusion Study demonstrated that grid pattern laser photocoagulation had no significant impact on the final visual acuity of eyes with CRVO and macular edema.[1]

Triamcinolone acetonide is a potent, relatively insoluble corticosteroid used for the treatment of intraocular inflammation by periocular injections. We have recently shown efficacy of trans-Tenon retrobulbar infusion of this drug for the treatment of vitritis and cystoid macular edema in association with uveitis.[5] The mechanism by which triamcinolone infusion can decrease chronic macular edema in eyes with CRVO and BRVO is unclear, but stabilization of the blood-retinal barrier via reduction of inflammatory cytokines may play a role. Chronic macular edema with CRVO or BRVO may also be due to traction on the retina by posterior vitreous cortex or epiretinal membrane. However, the fact that two CRVO eyes in our study, which did not improve after vitrectomy, responded to triamcinolone infusion suggests that vitreoretinal traction alone was not the cause of the macular edema.

Treatment efficacy by OCT was obtained in 13 of 16 eyes (81.3%) after a single triamcinolone infusion, but 8 of these initially responding eyes had recurrence of macular edema. Two of five recurrence eyes that underwent repeat triamcinolone infusions subsequently responded. Therefore, overall efficacy was obtained in 7 of 16 eyes (43.8%) in this study. Recurrences are a clear limitation of this treatment, and longer follow-up is required to investigate this issue. However, since 50% of eyes experienced visual acuity improvement, the clinical course after triamcinolone infusion appears to represent an improvement over natural history.[1]

Conclusion

Trans-Tenon retrobulbar infusion of triamcinolone acetonide may be effective in decreasing foveal thick-

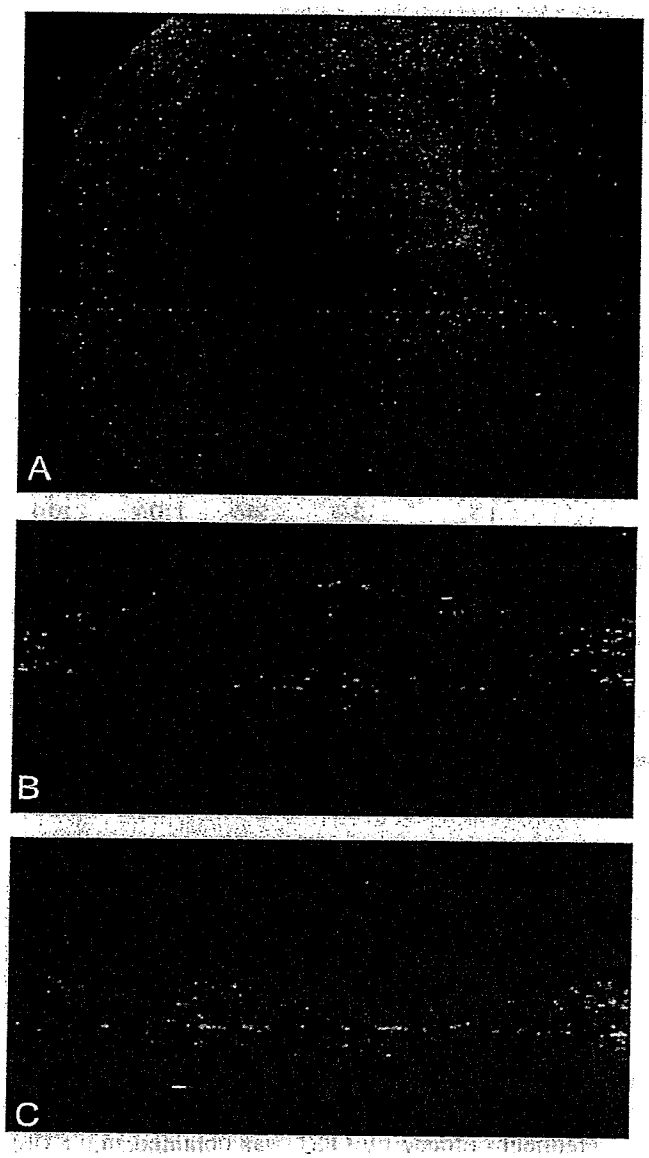

Fig. 1. Fundus photograph (A) of a 73-year-old woman at 14 months after onset of central retinal vein occlusion in her right eye. Optical coherence tomography examination revealed a foveal thickness of 368 μm before (B) and 57 μm at 2 weeks after (C) trans-Tenon retrobulbar triamcinolone infusion.

ness and improving visual acuity in eyes with chronic macular edema due to BRVO or CRVO.

Key words: branch retinal vein occlusion, central retinal vein occlusion, macular edema, triamcinolone acetonide.

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Long-term Results of Vitrectomy without Laser Treatment for Macular Detachment Associated with an Optic Disc Pit

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Purpose: To evaluate the efficacy of vitrectomy and gas tamponade, without laser photocoagulation to the margin of the optic nerve, for the treatment of macular detachment associated with an optic disc pit.

Design: Noncomparative interventional case series.

Participants: Eleven consecutive patients (8–47 years of age) who presented with unilateral macular detachment associated with an optic disc pit.

Intervention: Pars plana vitrectomy, induction of posterior vitreous detachment (PVD), and gas tamponade were performed, with postoperative facedown positioning for 1 week. The presence of a double-layer detachment consisting of an inner layer separation and an outer layer detachment was observed in 10 of 11 eyes either preoperatively or postoperatively. Patients were observed for 10 to 98 months (mean, 47) after surgery.

Main Outcome Measures: Anatomic outcome and visual acuity were retrospectively analyzed for all eyes. Optical coherence tomography was used to observe anatomic changes in the macula in some eyes.

Results: Complete retinal reattachment was achieved in 10 of 11 eyes, although these eyes required nearly 1 year to reach this state. The one eye with persistent retinal detachment was observed to have a marked reduction of the detachment by 10 months postoperatively. No recurrences were observed. Visual acuity improvement was documented in 7 of 11 eyes.

Conclusions: These results suggest that vitrectomy with induction of PVD and gas tamponade, without additional laser treatment, is successful in reattaching the macula and improving central vision in most patients with optic disc pit maculopathy. *Ophthalmology 2005;112:1430–1435 © 2005 by the American Academy of Ophthalmology.*

Congenital pit of the optic nerve head is a rare anomaly first described by Wiethe in 1882.[1] Approximately two thirds of patients have a concurrent or previous associated serous retinal detachment (RD) of the macula.[2–4] The age at onset of the RD is variable, with the mean being 30 years. The pathogenesis of optic disc pit maculopathy is unknown. In 1988, based on a study of stereoscopic transparencies and visual fields (VFs), Lincoff et al proposed that fluid from the optic disc pit creates a schisislike inner layer separation of the retina.[5] The outer layer detachment centered over the macula was suggested to be a secondary phenomenon. More recently, several authors have confirmed the 2-layer structure of optic disc pit maculopathy using optical coherence tomography (OCT).[6–8]

The treatment of serous RD associated with an optic disc pit is still controversial. The use of laser therapy to produce a barrier of chorioretinal adhesions at the optic disc border is often unsuccessful, and repeated treatments are needed.[9–11] Several reports suggest that vitrectomy combined with laser photocoagulation and gas tamponade may be more effective than external laser therapy alone, particularly in eyes with severe visual loss.[12–15] Theodossiadis reported that macular scleral buckling can yield favorable anatomical and functional results.[16] Lincoff et al reported that intravitreal gas injection alone can induce pneumatic displacement of the outer layer detachment and improve central vision.[17] However, the effect may only be temporary, because recurrence caused by fluid movement from the remaining inner layer separation was found by OCT.[8]

Bonnet reported that none of 25 eyes with macular detachment associated with optic disc pit had a posterior vitreous detachment (PVD), and that 2 of the eyes had spontaneous reattachment of the macula after development of PVD.[18] Gordon and Chatfield[19] and Gass[10] found no evidence of PVD in their cases, and suggested that vitreous traction on the macula may cause passive migration of fluid into the submacular space via the pit. Recent experience with the surgical treatment of macular holes and macular edema has shown the importance of vitreous tangential traction in thepathogenesis of these diseases.[20–22] Similar tangential vitreous traction at the pit, an area of abnormal

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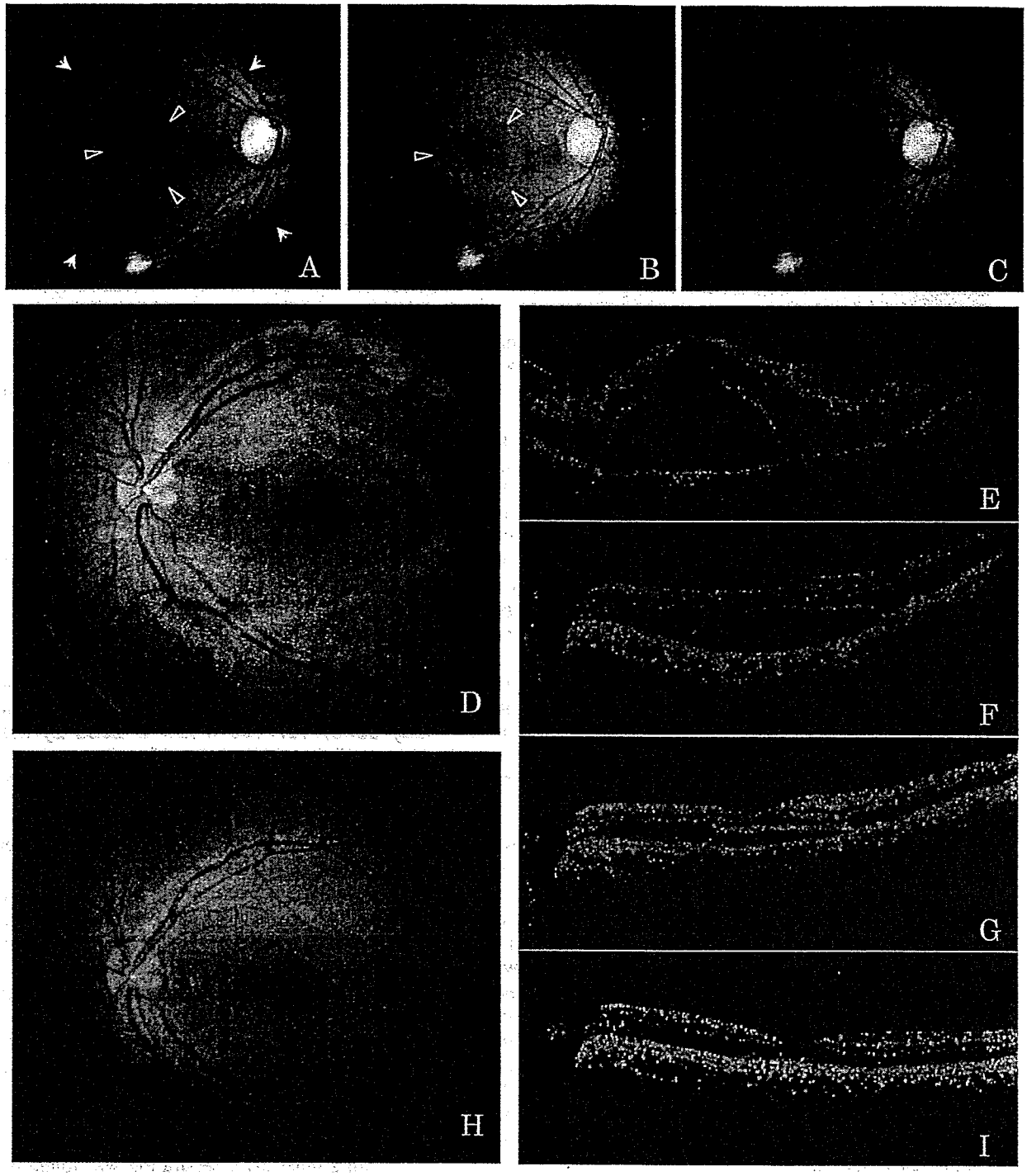

Figure 1. Composite of representative clinical findings from patients 1 (A–C) and 10 (D–I). All optical coherence tomography (OCT) scan lengths were 10 mm. A, Photograph of patient 1's right fundus preoperatively, showing an inferotemporal optic disc pit associated with a large oval-shaped area of macular detachment (large arrows), a nerve fiber layer defect leading from the optic disc pit (green arrow), and an area of myelinated nerve fibers in the inferior macula. The outer layer hole (blue arrow) and the round 1.5–disc diameter outer layer detachment (white open arrowheads) also shown were not present initially but developed after 4 months of follow-up. Preoperatively, the best-corrected visual acuity (VA) was 0.2. B, Two months after vitrectomy and gas tamponade, an irregularly shaped break was noted in the outer layer (blue arrow), with the outer layer detachment (white open arrowheads) being slightly enlarged. A new nerve fiber layer defect was also observed leading from the superotemporal edge of the optic disc (green arrows). C, One year postoperatively, the macula was observed to be completely reattached, with a VA of 1.0. The nerve fiber layer defects remained unchanged. D, Fundus photograph of patient 10's left eye at presentation, showing a shallow retinal elevation extending from the superotemporal to inferotemporal arcades. A round 2–disc diameter outer layer detachment was also observed in the center of the macula, and preoperatively, VA was 0.08. E, Optical coherence tomography at presentation revealed an inner layer separation as well as a hole in the outer layer detachment at the macula and an outer layer detachment surrounding the hole. F, Three months after vitrectomy and gas tamponade, OCT showed a decrease in the inner layer separation and outer layer detachment. G, At 9 months, OCT showed resolution of the inner layer separation but a residual outer layer detachment. H, I, Fundus photograph and OCT at 12 months postoperatively showing the macula completely reattached and the optic disc pit appearing grayer and deeper than preoperatively. Visual acuity was 1.2.

Table 1. Clinical

Patient	Age (yrs)	Gender	Eye	Refractive Error (Diopters)	Symptom	Preoperative Best-Corrected Visual Acuity	Pit Location
1	40	F	R	−2.0	Central scotoma	0.3	Inferotemporal
2	15	F	L	−1.0	Decreased VA	0.2	Inferotemporal
3	43	F	L	−1.5	Central scotoma	0.08	Temporal
4	30	M	R	−0.5	Central scotoma	0.6	Temporal
5	22	M	L	−1.0	Metamorphopsia	0.5	Inside in coloboma
6	19	M	R	0.0	Central scotoma	0.4	Temporal
7	24	M	L	0.0	Central scotoma	0.6	Inferior
8	42	F	R	−1.0	Metamorphopsia	1.0	Central
9	47	M	L	0.0	Central scotoma	0.2	Inferotemporal
10	8	F	L	−1.0	Decreased VA	0.08	Temporal
11	44	F	R	0.0	Decreased VA	0.4	Temporal

F = female; L = left; M = male; R = right; VA = best-corrected visual acuity; VF = visual field.
+, present; −, absent.
*Duration in months.
†Displacement of subretinal fluid immediately after surgery.

configuration of the optic disc, may cause migration of fluid into intraretinal spaces. We believe that posterior vitreous traction on the margin of the optic disc pit may play an important role in the pathogenesis of this disease. There have been several reports regarding the efficacy of vitrectomy and gas tamponade with conflicting results, some reporting the recurrence of macular detachment with long-term follow-up after surgery.[8,12–14] However, most of these reports did not specify whether PVD induction was performed, and some stated that only core vitrectomy permitting space for a 60% to 70% gas tamponade was performed.

The purpose of this study was to examine long-term clinical outcomes in 11 eyes that underwent vitrectomy with PVD induction and gas tamponade, without laser application, for the treatment of optic disc pit maculopathy.

Patients and Methods

Eleven eyes of 11 consecutive patients who presented to the Kyorin Eye Center with an optic disc pit associated with macular detachment were included in this study. Institutional review board approval was not required, and records were reviewed retrospectively. Best-corrected visual acuity (VA) was recorded, and indirect funduscopy, slit-lamp biomicroscopy using a contact lens, and Goldman VF examinations were performed preoperatively and postoperatively. Scanning laser ophthalmoscopy and fluorescein angiography were done preoperatively to confirm the optic disc pit and macular abnormalities in some patients. Optical coherence tomography (Zeiss-Humphrey, San Leandro, CA) was used to observe posterior retinal changes in cases with follow-up from 1999 on.

Surgery was performed for the indication of worsening VA or for macular detachment persisting for ≥3 months. All surgeries were performed by the same surgeon (AH) between July 1994 and October 2003, and patients were observed postoperatively for 10 to 98 months (mean, 47). Vitrectomy was performed with the intention of releasing vitreous traction at the optic disc pit. Posterior vitreous detachment was initiated by suction over the optic disc or near areas of retinal schisis using the vitreous cutter. To limit retinal damage secondary to surgical manipulation, special attention was given to separating the posterior hyaloid gently over schisis areas. Triamcinolone acetonide[23,24] was used intraoperatively in 2 eyes (patients 10 and 11) and fluorescein dye[25,26] in 1 eye (patient 2) to highlight the posterior hyaloid membrane. After removal of the posterior hyaloid over the posterior pole, fluid–air exchange was performed, followed by gas tamponade with either 15% to 20% sulfur hexafluoride or 14% perfluoropropane and postoperative facedown positioning for approximately 1 week.

Cataract surgery was not performed, except in patient 3, who underwent lens extraction and intraocular lens implantation during a second vitrectomy procedure to close a macular hole.

Results

Clinical Characteristics

The clinical characteristics of all 11 patients are shown in Table 1, and clinical photographs of representative patients are shown in Figure 1. Six of the patients were women and 5 were men, with ages ranging from 8 to 47 years (mean, 30.4). All patients were of Japanese ethnicity, except for patient 7, who was Caucasian. All patients complained of a central scotoma or metamorphopsia in the affected eye for several months. None of the affected eyes had severe refractive errors, and preoperative VA ranged from 0.08 to 1.0 (mean, 0.3). Patient 7 had received previous treatment for the optic disc pit maculopathy, consisting of laser photocoagulation to the edge of the optic disc pit, but no other patients had received any prior treatment. Nine of the 11 patients had no pertinent medical or ocular history. Patient 10 was referred to our hospital for visual disturbance after blunt ocular trauma by a volleyball, at which time macular detachment associated with an optic disc pit was diagnosed in the injured eye. Patient 8 had a history of RD surgery in the fellow eye.

The presence of a double-layer detachment, consisting of both an inner layer separation and an outer layer detachment, was confirmed either before or after surgery in 10 of 11 eyes. The outer layer detachment did not seem to communicate with the optic disc in 8 eyes, but did seem to do so in 2 eyes. An irregularly shaped outer layer break and outer layer detachment were observed to develop after schisislike inner layer separation in patients 1 (Fig 1A) and 4 preoperatively and in patient 8 postoperatively. An outer layer break in the macula was present or developed sometime

Characteristics

Duration of Symptom*	Double Layer Retinal Detachment	Outer Layer Hole at Fovea	Posterior Vitreous Detachment	Duration until Macular Attachment*	Final Best-Corrected Visual Acuity	Complications	Follow-up*
5.0	+	+	−	12	1.2	Peripheral VF defect	80
>36.0	+	+	−	2	0.04	Retinal damage	91
6.0	+	+	−	5	0.1	—	98
6.0	+	+	−	8	0.8	Retinal break	24
2.0	−	−	−	12†	1.2	—	73
9.0	+	+	−	12†	1.0	—	37
24.0	+	−	−	12†	1.2	—	15
2.0	+	+	−	15	1.0	—	57
3.0	+	+	−	10	1.2	—	14
5.0	+	+	−	12	1.2	—	12
24.0	+	+	−	Flatter	1.0	—	10

during the clinical course in a total of 9 of the 11 eyes. One eye (patient 3) had a full-thickness macular hole preoperatively. Neither PVD nor vitreomacular or vitreopapillary traction was observed in any eyes preoperatively by fundus biomicroscopy; OCT performed preoperatively in 5 eyes also did not reveal vitreomacular or vitreopapillary traction.

Anatomic Results

Complete retinal reattachment was achieved in 10 of 11 eyes, although these eyes required nearly 1 year to reach this state (Fig 1). Interestingly, after surgery the outer layer break seemed to enlarge temporarily in most cases (Fig 1B). One eye had persistent RD after surgery, but was documented by OCT to have marked reduction of the detachment by 10 months postoperatively. No recurrences were observed in any eyes.

Observation by OCT showed slow absorption of the inner layer separation and outer layer detachment after surgery, with complete absorption of fluid after 2 to 15 months (Fig 1E–G, I). The macular hole present preoperatively in patient 3 remained open after surgery, but without surrounding RD (fluid cuff) or retinal edema. However, 3 years postoperatively a fluid cuff appeared around the macular hole, with corresponding decrease in vision, and a second vitrectomy procedure with internal limiting membrane peeling and gas tamponade successfully closed the hole.

Two patients (5 and 11) also had an optic disc coloboma, with the optic disc pit present within the area of the coloboma. The pits were not obvious preoperatively; however, postoperatively, as the retina reattached, the pits were easily observed as being darker and deeper relative to the surrounding colobomatous areas.

Visual Acuity Results

Preoperative and final VAs are shown in Table 1. Despite evidence of residual shallow inner layer separation and outer layer detachment, VA started to improve within a few months in most eyes. Improvement in VA of 0.2 logarithms of the minimum angle of resolution or greater was documented in 7 of 11 eyes. Nine of 11 eyes had a postoperative VA of 0.8 or better.

Complications

Intraoperative or postoperative complications were observed in 3 patients. Patient 1 developed a dense scotoma in the inferotemporal quadrant with a nerve fiber layer defect between 12-o'clock and 3-o'clock at 2 weeks postoperatively (Fig 1B). At final examination 1 year postoperatively, VA was 1.0, with the macula reattached and no change in the nerve fiber layer defect (Fig 1C).

Patient 2, a 15-year-old girl, underwent vitrectomy and fluorescein dye–assisted PVD induction followed by 15% sulfur hexafluoride tamponade. The surgeon noted difficulty in inducing the PVD, resorting to the use of a retinal pick and forceps in addition to cutter suction. A few drops of 2% fluorescein dye were introduced to the infusion line to highlight the posterior hyaloid membrane.[25,26] A tiny hemorrhage was noted at the superior margin of the optic disc after complete vitreous separation. On the second postoperative day, the patient complained of darkened vision, and whitening of the retina was observed around the optic disc in a gas-filled eye. Fluorescein angiography showed normal arm-to-retina circulation times. Oral prednisolone at a dose of 40 mg tapered to 30 mg was administered for a total of 9 days for the possibility of inflammation related to the surgical manipulation or phototoxicity, but the retinal findings remained unchanged. Electoretinography performed on the fifth postoperative day showed a negative b-wave and a reduced a-wave. Pigmented atrophy of the peripapillary area developed over 6 months.

Patient 4 developed an iatrogenic peripheral retinal break intraoperatively during PVD induction that was treated successfully with laser photocoagulation.

Discussion

In this study, we were able to confirm Lincoff et al's notion of a double-layer detachment in 10 of 11 eyes with optic disc pit maculopathy.[5] An outer layer detachment was observed either preoperatively or postoperatively as a secondary phenomenon, developing after presence of an inner layer separation in 3 eyes. In the remaining 7 eyes, the outer layer detachment, along with an outer layer break, was observed at presentation.

Lincoff et al reported on the successful use of intravitreal gas tamponade without vitrectomy to induce pneumatic displacement of the outer layer detachment and improve central vision.[17] However, OCT findings have suggested that fluid may continue to flow from the remaining inner layer separation to the outer layer detachment.[8] Because we