Circumscribed choroidal hemangioma: Visual outcome in the pre-PDT vs PDT eras in 458 cases

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Financial Disclosure

I have no financial interests or relationships to disclose.

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In summary, Over 5 decades in 458 cases of circumscribed choroidal hemangioma, we have made headway with PDT providing significantly better visual outcome

Circumscribed choroidal hemangioma

- Benign vascular tumor
- Post-equatorial region
- Ultrasound dense
- Median basal diameter 6 mm
- Median thickness
 3 mm
- Lead to profound visual loss
 - Subretinal fluid
 - Cystoid macular edema

What did we know 20 years back?

Circumscribed Choroidal Hemangioma

Clinical Manifestations and Factors Predictive of Visual Outcome in 200 Consecutive Cases

Carol L. Shields, MD, Santosh G. Honavar, MD, Jerry A. Shields, MD, Jacqueline Cater, PhD, Hakan Demirci, MD

Purpose: To review the clinical features and management of circumscribed choroidal hemangioma and determine factors predictive of poor visual outcome.

Design: Retrospective consecutive noncomparative interventional case series.

Participants: Two hundred consecutive patients with circumscribed choroidal hemangioma.

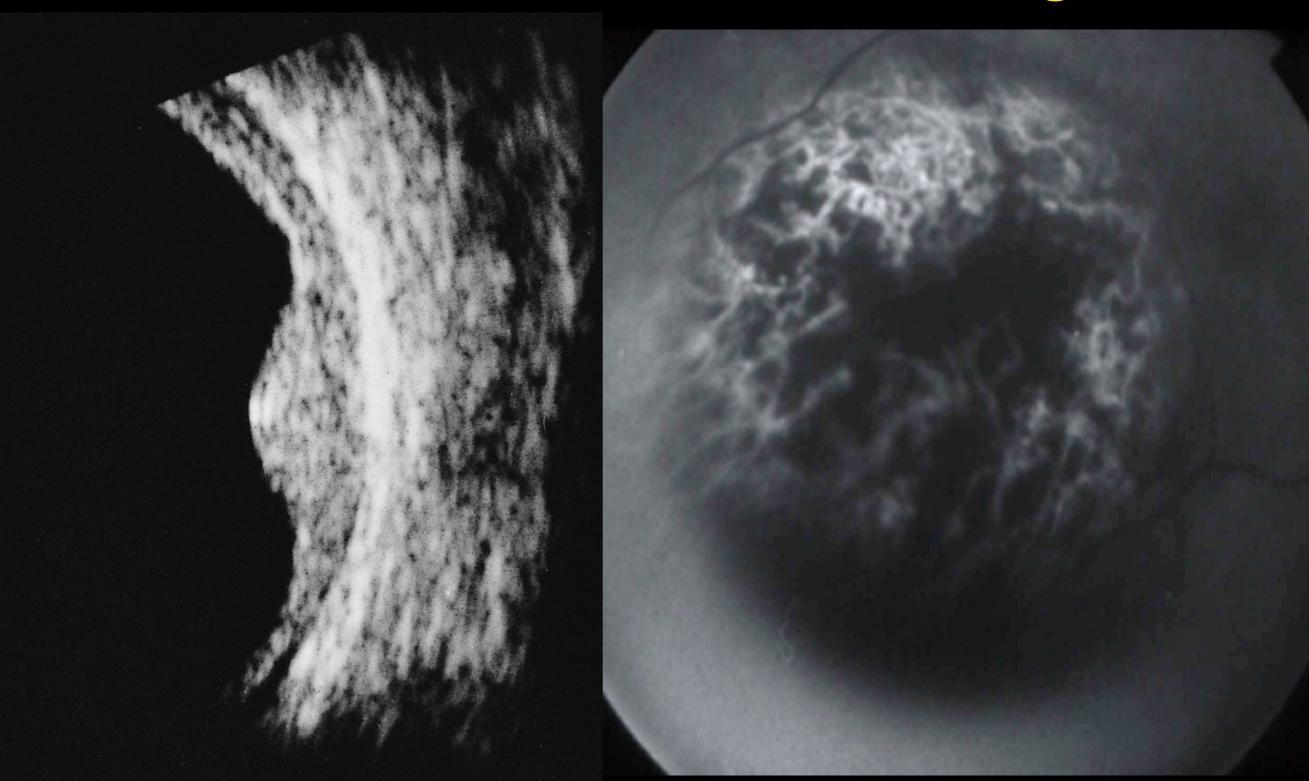
Main Outcome Measures: The main outcome measures were analyzed in 155 patients with follow-up of at least 3 months and included complete resolution of subretinal fluid, worsening of visual acuity (more than 2 Snellen lines), and poor final visual acuity (20/200 or worse).

n=200 cases laser era misdiagnosis in 38% poor vision (\leq 20/200) in 60% @ 10 yrs, despite control of SRF

before referral (P = 0.01), and tumor management with observation after referral (P = 0.02). Worsening of visual acuity (by more than 2 Snellen lines) was observed in 8% at 5 years and 28% at 10 years of those 82 patients who were initially seen with poor vision. Worsening of visual acuity was found in 10% at 5 years and 30% at 10 years of those 73 patients who initially were seen with good to moderate vision.

Conclusions: Circumscribed choroidal hemangioma is a rare intraocular tumor. In 38% of cases, this tumor is initially misinterpreted before referral as choroidal melanoma or metastasis. Visual acuity is poor in more than 60% of patients at 10 years, despite successful control of associated subretinal fluid in 76% patients. *Ophthalmology 2001;108:2237–2248* © 2001 by the American Academy of Ophthalmology.

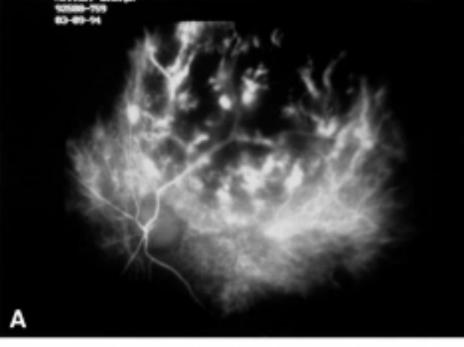
Circumscribed choroidal hemangioma



Ultrasound echodense

IVFA early hyperFA

Circumscribed Choroidal Her **Characteristic Features with** Green Videoangiography



J. Fernando Arevalo, MD,^{1,2} Carol L. Shields, MD,¹ Jerry A. Shields, MI Patrick De Potter, MD¹

Objective: To determine the characteristic features of indocyanin

circumscribed choroidal h Design: Prospective, Participants: Twenty-Intervention: Indocya prospectively performed a

Main Outcome Measures: The behavior of circumscribed choroidal ICG-V and IVFA.

Results: On ICG-V, earliest hyperfluorescence of circumscribed characteristics mean of 27.6 seconds (range, 13–62 seconds), whereas maximum hype (range, 33–707 seconds). In the late frames, all eyes demonstrated a relation 18 eyes (72%) that demonstrated "washout" of the dye. Other findings o eyes (76%), a late hyperfluorescent rim in 19 eyes (76%), and late frame the earliest hyperfluorescence was achieved at a mean of 24 secon onds (rang washout @ 10

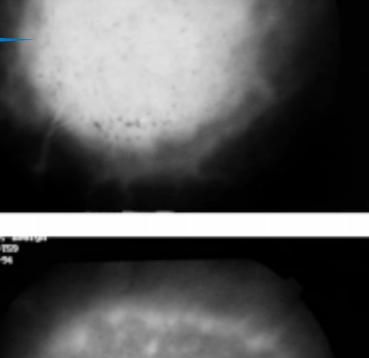
maximum hyperfluorescer fluorescence in the late fra and hot spots in the late f

Conclusions: Circum

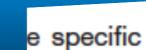
hyperICG @ I

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visualized with IVFA. We believe that ICG-v may become an importan choroidal hemangioma. Ophthalmology 2000;107:344-350 © 2000 by t oav.

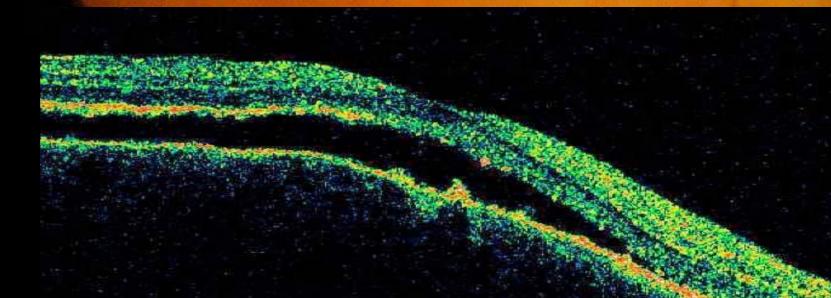
Choroidal Hemangioma



Choroidal Hemangioma

Treatment Observation

Caser
Transpupillary thermotherapy
Photodynamic therapy
Plaque radiotherapy
Propranolol



Choroidal Hemangioma

Photodynamic therapy

Before Va 20/60

After Va 20/20

Photodynamic Therapy for Symptomatic Choroidal Hemangioma 2002

Visual and Anatomic R

Ursula M. Schmidt-Erfurth, MD,¹ Stephan M Albert J. Augustin, MD³

Objective: To document the anatomic a atic choroidal hemangioma

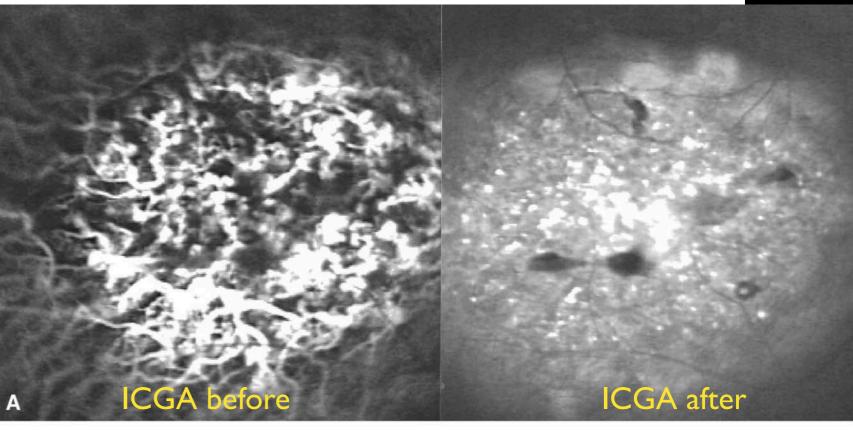
Design: Prospective, noncomparative, in **Participants:** Fifteen patients with circu with progressive vision loss caused by exud

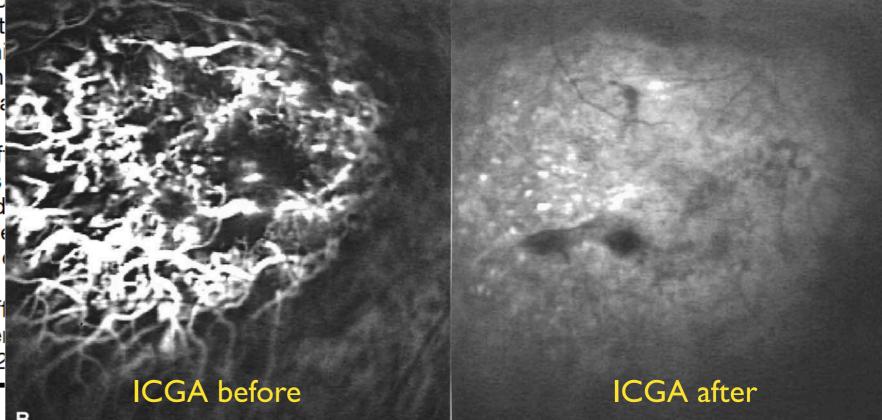
Intervention: PDT using 6 mg/m² body s performed. One to four treatments with a s evaluation was provided before and at 6-wee follow-up 19 months after the last application

Main Outcome Measures: Functional t betic Retinopathy Study criteria) and scanni thalmoscopy, fluorescein/indocyanine green

Results: A complete regression of the value four consecutive treatments. Tumors (mean each treatment, with the most intensive effangiomatous net without recanalization was resolution of metamorphopsia; 13 patients d ment of an average of 3 lines was documented therapy. Visual fields showed withdrawal of a follow-up for up to 50 months.

Conclusions: PDT using verteporfin off Complete anatomic regression with persister in vision. *Ophthalmology* 2002;109:2284–22





Photodynamic therapy using verteporfin in circumscribed choroidal haemangioma

B Jurklies, G Anastassiou, S Ortmans, A Schüler, H Schilling, U Schmidt-Erfurth, N Bornfeld

Br J Ophthalmol 2003;87:84-89

Aim: To investigate the safety and efficacy of photodynamic therapy with verteporfin in patients with choroidal haemangioma.

Methods: A non-randomised, prospective clinical investigation of 19 patients with symptomatic circumscribed choroidal haemangioma was performed. Unsuccessful pretreatment (external beam irradiation, laser photocoagulation) was performed in four patients. Patients were included when (1) sub-retinal exudation involving the fovea, (2) a decrease in visual function, and (3) additional symptoms (for example, metamorphopsia) were present. Photodynamic therapy (PDT) was performed with verteporfin

at a concentration of 6 mg/m² bod **Results:** The mean follow up time v sions was 2.15 (range 1–5). Visual in 42.1%, was stable in 21.1%, a completely resolved in 94.8% of t tumours. Patients receiving any pre more than 30 months, and no sign cant improvement. Cox regression inversely associated with the impro local or systemic side effects were **Conclusion:** PDT using verteporfit choroidal haemangioma even in tu

n=19 cases
tumor reduction in 100%
visual acuity
improved ≥ 1 line 73%
improved ≥ 2 lines 42%
PDT sessions inverse to VA
no recurrence

See end of article for authors' affiliations

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Photodynamic therapy of circumscribed choroidal

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RESEARCH ARTICLE

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Clinical outcomes and predictors of response

to photodynamic therapy in symptomatic

Aim: To eva therapy (PD1 circumscribe Design: Pros series and c Methods: 10 transpupillar beam radio oidal haema given as an laser (690) 83 seconds tion of infusi were applie Periodic foll and angiogr Results: All flattening o reduction o visual acuity (80%) patien was seen in Conclusions: ment for mo oidal haemo lead to visue Analysis of Long-term Outcomes of Radiotherapy and Verteporfin Photodynamic Therapy for Circumscribed Choroidal Hemangioma

Vasilios P. Papas Marie Restori, P Victoria M.L. C

Purpose: T choroidal hema Design: Re Subjects: Methods: radiotherapy (LS Main Outcor dimensions, and Results: Th 42 (70%) receiv [27%]; mean fol location, mottle group. In the LS deviation, 1.65 Snellen lines in at 5 years. In t 1.0±0.8; range, in 30% of cases versus single du central retinal t difference in de brachytherapy, cases at the en group and 2 of Conclusions

Photodynamic Therapy with Verteporfin for Symptomatic Circumscribed Choroidal Hemangioma: Five-Year Outcomes

Maria Antonietta Blasi, MD,^{1,2} Alessandra C. Tiberti, MD, PhD,¹ Andrea Scupola, MD,¹ Angelo Balestrazzi, MD,³ Egle Colangelo, MD,¹ Paola Valente, MD,¹ Emilio Balestrazzi, MD¹

Objective: To evaluate the long-term efficacy of verteporfin photodynamic therapy (PDT) as the primary treatment for symptomatic circumscribed choroidal hemangioma (CCH).

Design: Prospective consecutive, 2-centered, noncomparative, interventional case series.

Participants: Twenty-five subjects with symptomatic CCH. All patients had recent onset of visual symptoms and evidence of exudative macular changes on fluorescein angiography (FA) and optical coherence tomography (OCT).

Methods: Verteporfin 6 mg/m² body surface area was administered intravenously over a 10-minute interval. Five minutes after infusion, a 689 nm laser was applied with a light dose of 50 J/cm² for the first 3 patients and a light dose of 100 J/cm² for all the other patients. Retreatments were performed in case of persistent exudation found on OCT. Evaluation of best-corrected visual acuity (BCVA) using Early Treatment of Diabetic Retinopathy Study (ETDRS) criteria, FA, indocyanine green angiography (ICGA), OCT, and ultrasound were performed before PDT and on follow-up examinations. All patients were followed for at least 5 years.

Main Outcome Measures (FCT) between baseline and n on FA, and adverse events.

Results: Twenty-two par Three eyes, treated with 50 J

Main Outcome Measures Primary outcome measures were changes in RCVA and foveal center thickness

all with ≤ 25 cases

Deeu

Collected data on <u>458 eyes</u> with circumscribed choroidal hemangioma over 5 decades

Evaluate Outcomes based on age Outcomes based on era •pre-PDT era vs the PDT era

Carol L. Shields, MD,¹ Lauren A. Dalvin, MD,² Li-Anne S. Lim, MD,¹ Michael Chang, MD,¹
 Sanika Udyaver, BS,¹ Mehdi Mazloumi, MD, MPH,¹ Pornpattana Vichitvejpaisal, MD,³ Grace L. Su, BS,¹
 Eleni Florakis, BS,¹ Arman Mashayekhi, MD,¹ Jerry A. Shields, MD¹

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Pre-PDT vs PDT 1967-2001 2002-2018 n=220 n=238

vs. PDT [n = 238 cases]) revealed PDT era patients were of older mean age (48.9 vs. 53.8 years, P = 0.002) and were more likely to have systemic hypertension (17.7% vs. 33.8%, P < 0.001), tumor location in the macula (57.4% vs. 67.5%, P = 0.01), subretinal fluid on OCT (33.3% vs. 70.7%, P = 0.01), and greater extent of overlying lipofuscin (P = 0.001). Findings of tumor basal diameter and thickness and fluorescein and indocyanine green angiography were no different in the 2 eras. Treatment (pre-PDT vs. PDT) included argon laser photocoagulation (42.1% vs. 0.4%), PDT (0% vs. 43.8%), transpupillary thermotherapy (0% vs. 0.4%), plaque radiotherapy (7.0% vs. 5.2%), external beam radiotherapy (1.4% vs. 1.3%), enucleation (0.9% vs. 0.4%), and observation (48.6% vs. 47.6%). After treatment, patients in the PDT era demonstrated better mean logarithm of the minimum angle of resolution visual acuity (1.28 vs. 0.51, P < 0.001) (Snellen equivalent 20/400 vs. 20/63, P < 0.001). Final visual acuity was \geq 20/40 for those with entering vision of \geq 20/40 (59.6% vs. 74.7%, P = 0.001) and for those with entering vision of 20/50–20/200 (25.4% vs. 47.3%, P < 0.001).

Conclusions: Management of choroidal hemangioma in the PDT era has allowed for significantly better vi-

Table 1. Visual Outcome of Circumscribed Choroidal Hemangioma in the Pre–Photodynamic Therapy Era (n = 220) versus Photodynamic Therapy Era (n = 238) in 458 Tumors of 457 Patients: Patient Demographics

Demographic Features	Pre-PDT Era (1967–2001) n = 220 Tumors in 220 Patients (%)	PDT Era (2002–2018) n = 238 Tumors in 237 Patients (%)	P Value	Total N = 458 Tumors in 457 Patients (%)
Age at presentation (yrs) ($n = 457$ patients)				
Mean (median, range)	48.9 (49.5, 3–93)	53.8 (56, 4–89)	0.002	51.4 (52, 3–93)
Sex $(n = 457 \text{ patients})$	126 (57.2)	142 (60.2)	0.51	260 (59 0)
Male	126 (57.3)	143 (60.3)	0.51	269 (58.9)
Female	94 (42.7)	94 (39.7)		188 (41.1)
Race $(n = 457 \text{ patients})$				
White	190 (86.4)	203 (85.6)		393 (86.0)
African American	7 (3.2)	5 (2.1)		12 (2.6)
Asian	4 (1.8)	6 (2.5)	0.22	10 (2.2)
Hispanic	7 (3.2)	12 (5.1)		19 (4.2)
Middle Eastern	1 (0.5)	1 (0.4)		2 (0.4)
Indian	0 (0.0)	4 (1.7)		4 (0.8)
Unknown	10 (4.5)	5 (2.1)		15 (3.3)
Study eye ($n = 458$ tumors)				
Right eye	107 (48.6)	122 (51.3)		229 (50.0)
Left eye	113 (51.4)	115 (48.3)	0.51	228 (49.8)
Both eyes	0 (0.0)	0 (0.0)		0 (0.0)

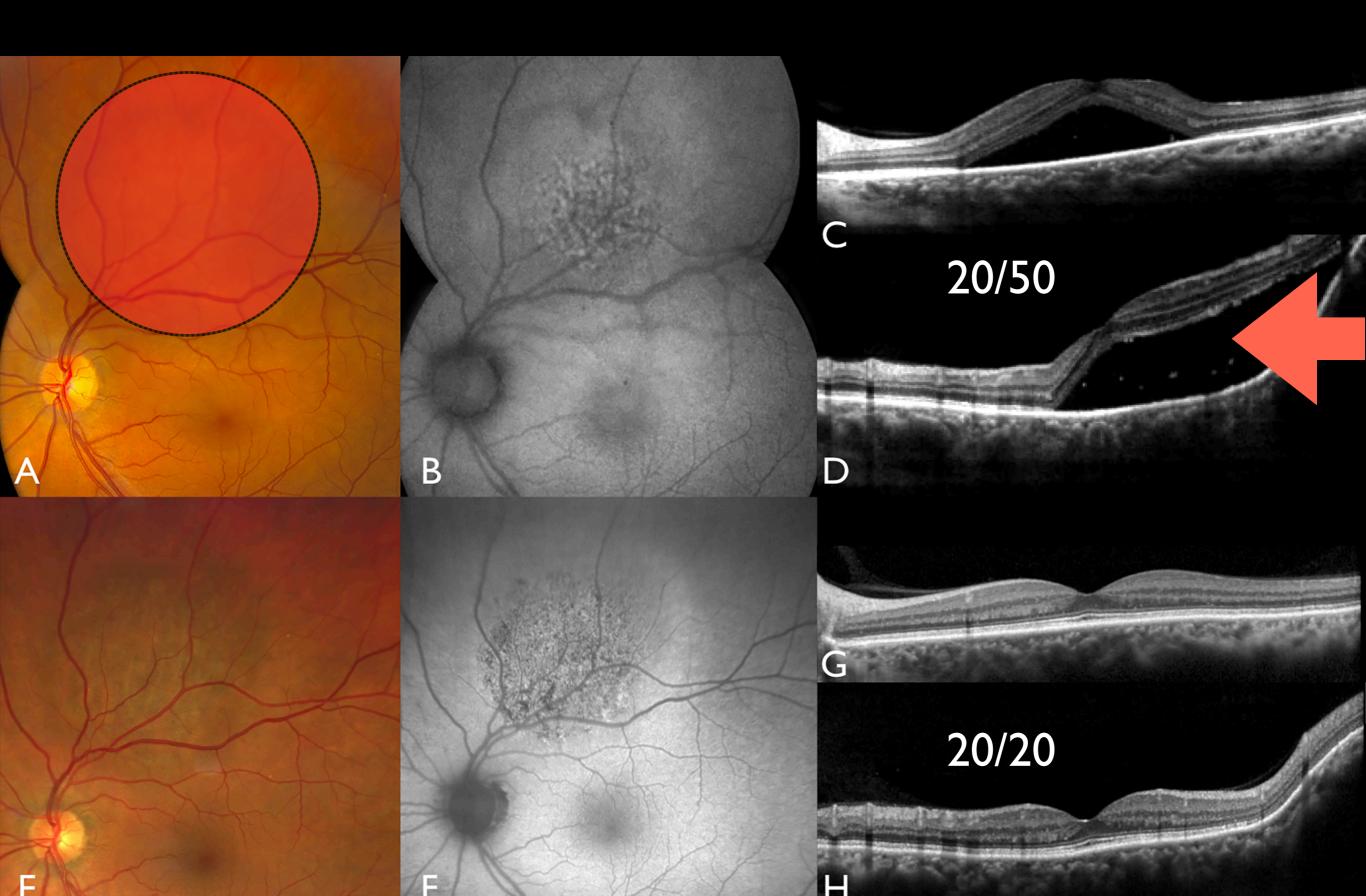
Table 2. Visual Outcome of Circumscribed Choroidal Hemangioma in the Pre–Photodynamic Therapy Era (n = 220) versus Photodynamic Therapy Era (n = 238) in 458 Tumors of 457 Patients: Clinical Features at Presentation

Clinical Tumor Features	Pre-PDT Era (1967–2001) n = 220 Tumors in 220 Patients (%)	PDT Era (2002–2018) n = 238 Tumors in 237 Patients (%)	P Value	Total N = 458 Tumors in 457 Patients (%)
No. of tumor(s)	n = 220	n = 238		N = 458
No. of tumors per patient, mean (median, range)	1.00 (1, 1–1)	1.00 (1, 1-2)	0.34	1.00 (1, 1-2)
No. of tumors per eye, mean (median, range)	1.00 (1, 1–1)	1.01 (1, 1-2)	0.17	1.01 (1, 1-2)
Size tumor	n = 220	n = 238		N = 458
Tumor diameter (mm), mean (median, range)	6.84 (6.5, 1–16)	6.77 (6, 2-24)	0.80	6.80 (6, 1-24)
Tumor thickness (mm), mean (median, range)	3.05 (3, 1-8)	3.24 (3, 1–11)	0.12	3.15 (3, 1–11)
Location proximity disc and foveola	n = 220	n = 238		N = 458
Distance to optic disc (mm), mean (median, range)	1.65 (0.5, 0–18)	1.75 (1.0, 0-8)	0.62	1.71 (1.0, 0–18)
Distance to foveola (mm), mean (median, range)	1.24 (0.5, 0–15)	1.22 (0.5, 0-7)	0.91	1.23 (0.5, 0–15)
Location quadrant	n = 216	n = 237		N = 453
Macula	124 (57.4)	160 (67.5)		284 (62.7)
Inferior	18 (8.3)	14 (5.9)		32 (7.1)
Temporal	33 (15.3)	14 (5.9)	0.01	47 (10.4)
Superior	22 (10.2)	27 (11.4)		49 (10.8)
Nasal	19 (8.8)	22 (9.3)		41 (9.1)
Subretinal fluid surrounding tumor	n = 215	n = 236		N = 451
None	52 (24.2)	74 (31.4)		126 (27.9)
Subretinal fluid cap	36 (16.7)	48 (20.3)		84 (18.6)
Subretinal fluid <3 mm from tumor	47 (21.9)	50 (21.2)	0.10	97 (21.5)
Subretinal fluid 3–6 mm from tumor	24 (11.2)	25 (10.6)		49 (10.8)
Subretinal fluid >6 mm from tumor	56 (26.0)	39 (16.5)		95 (21.1)
Subretinal fluid under macula	n = 213	n = 235		N = 448
Submacular fluid	56 (26.3)	52 (22.1)	0.06	108 (24.1)

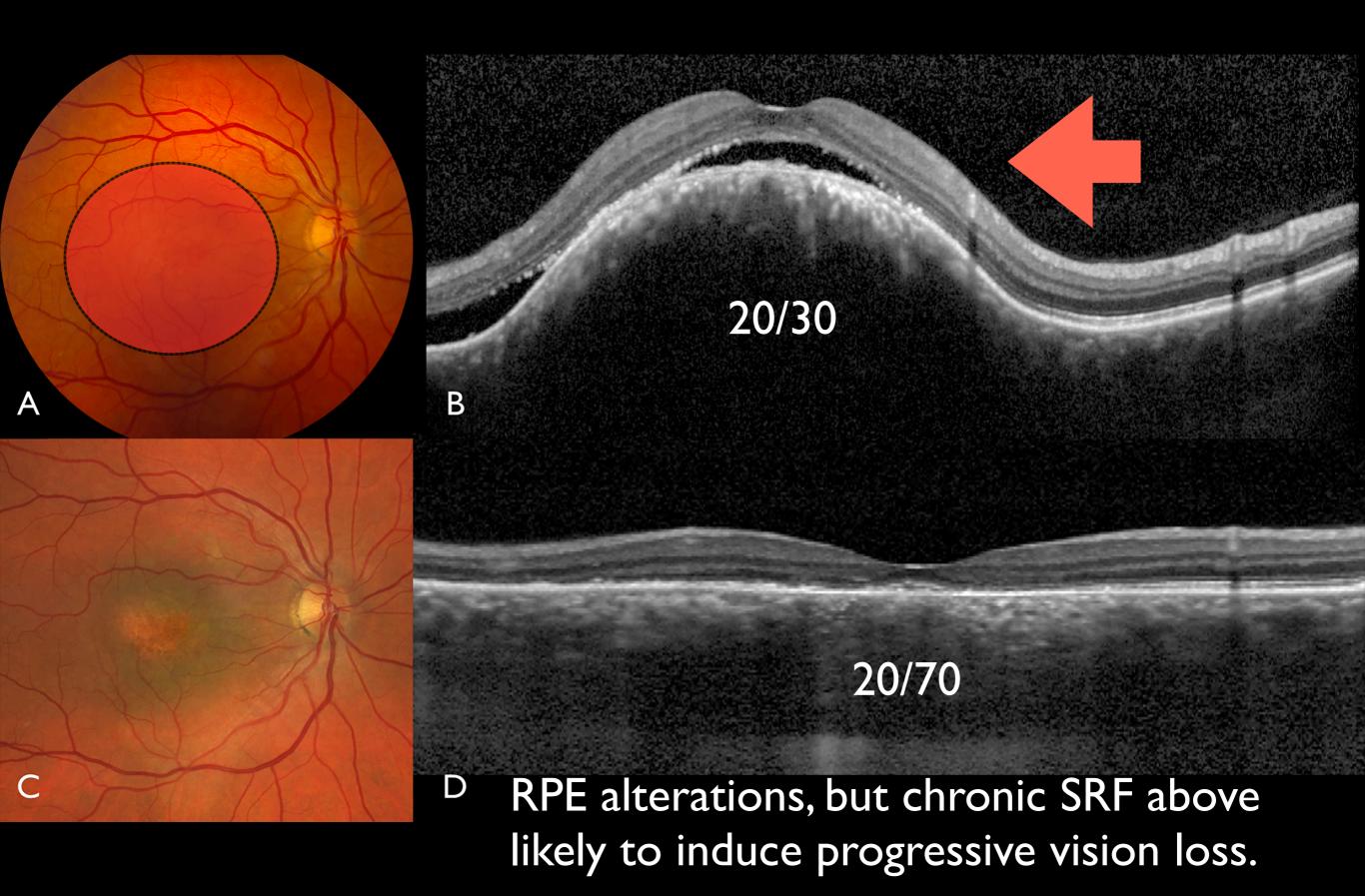
Table 4. Visual Outcome of Circumscribed Choroidal Hemangioma in the Pre–Photodynamic Therapy Era (n = 220) versus Photodynamic Therapy Era (n = 238) in 458 Tumors of 457 Patients: Treatment Features

Treatment Features	Pre-PDT Era (1967–2001) n = 220 Tumors in 220 Patients (%)	PDT era (2002–2018) n = 238 Tumors in 237 Patients (%)	P Value	Total N = 458 Tumors in 457 Patients (%)
Reason for treatment	n = 106	n = 119		N = 225
Subretinal fluid progression	36 (34.0)	27 (22.7)		63 (28.0)
Subretinal fluid at fovea	65 (61.3)	80 (67.2)		145 (64.4)
Retinal exudation	2 (1.9)	2 (18.2)		4 (1.8)
Hemorrhage	0 (0.0)	0 (0.0)	0.04	0 (0.0)
Macular edema	2 (1.9)	10 (8.4)		12 (0.9)
Choroidal neovascular membrane	0 (0.0)	0 (0.0)		0 (0.0)
Blind painful eye	1 (0.9)	0 (0.0)		1 (0.4)
Primary treatment modality	n = 214	n = 233		N = 447
Observation	104 (48.6)	111 (47.6)		215 (48.1)
Transscleral diathermy	0 (0.0)	0 (0.0)		0 (0.0)
Argon laser photocoagulation	90 (42.1)	1 (0.4)		91 (20.4)
Transpupillary thermotherapy	0 (0.0)	1 (0.4)		1 (0.2)
PDT	0 (0.0)	102 (43.8)		102 (22.8)
Plaque radiotherapy	15 (7.0)	12 (5.2)	< 0.001	27 (6.0)
External beam radiotherapy	3 (1.4)	3 (1.3)		6 (1.3)
Enucleation	2 (0.9)	1 (0.4)		3 (0.7)
Subtenon triamcinolone	0 (0.0)	0 (0.0)		0 (0.0)
Intravitreal triamcinolone	0 (0.0)	0 (0.0)		0 (0.0)
Intravitreal anti-VEGF	0 (0.0)	2 (0.9)		2 (0.4)
C 1 1 1 1 1	140	170		NJ 210

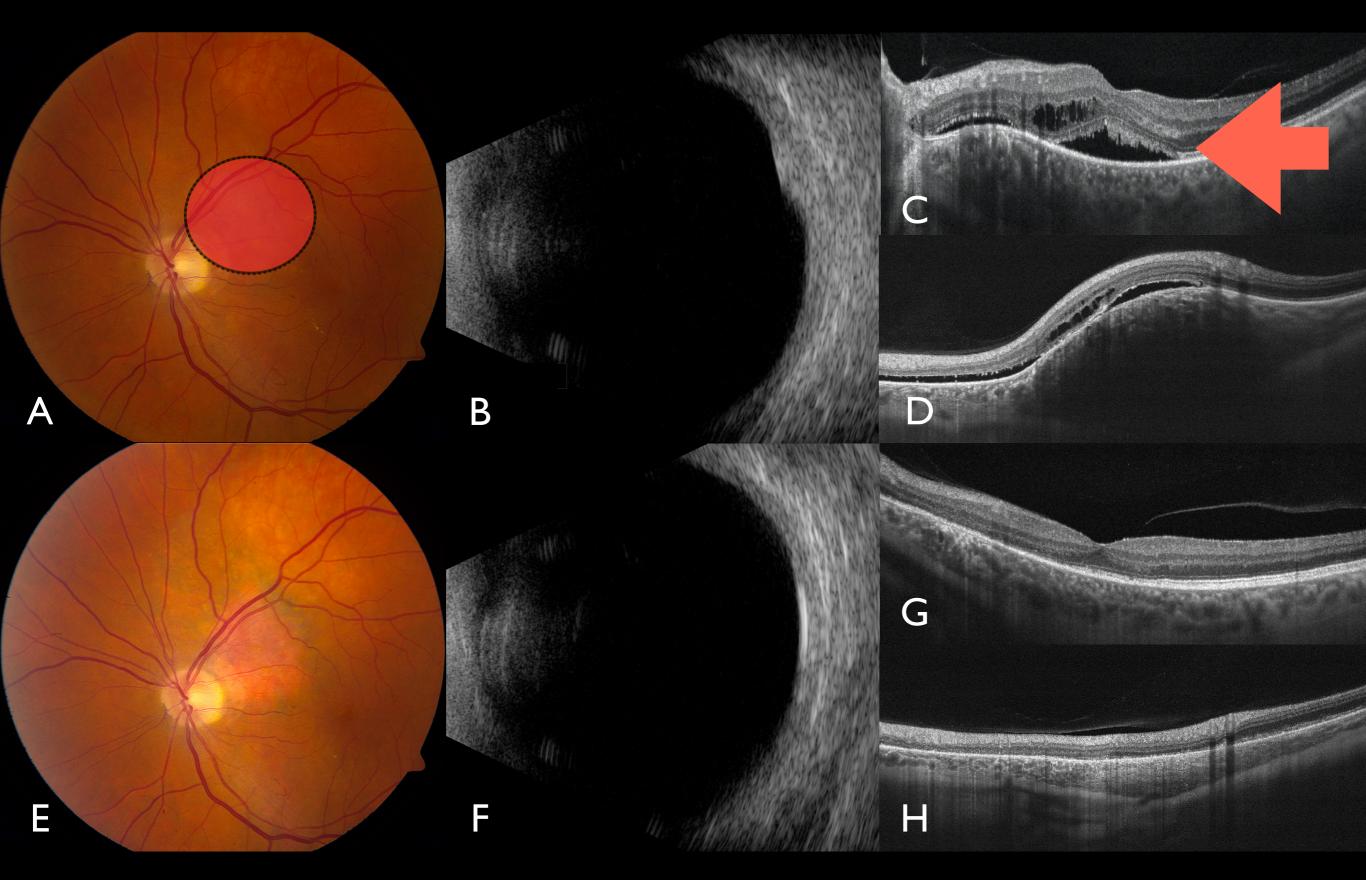
Post-PDT - SRF



Post-PDT - Subfoveal



Post-PDT - CME



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 Sanika Udyaver, BS,¹ Mehdi Mazloumi, MD, MPH,¹ Pornpattana Vichitvejpaisal, MD,³ Grace L. Su, BS,¹
 Eleni Florakis, BS,¹ Arman Mashayekhi, MD,¹ Jerry A. Shields, MD¹

Conclusions: Management of choroidal hemangioma in the PDT era has allowed for significantly better visual outcome compared with the pre-PDT era, with mean final visual acuity of 20/400 (pre-PDT era) versus 20/63 (PDT era). *Ophthalmology Retina* 2019; $\equiv :1-12 \otimes 2019$ by the American Academy of Ophthalmology

Main Outcome Measure: Visual acuity outcome. Results: A total of 458 tumors were treated over this 51-year period. A comparison (pre-PDT [n = 220 cases]

Over 5 decades in 458 cases pre-PDT PDT 20/400 vs 20/63

PDT wins



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In summary, Over 5 decades in 458 cases of circumscribed choroidal hemangioma, we have made headway with PDT providing significantly better visual outcome

Circumscribed choroidal hemangioma: Visual outcome in the pre-PDT vs PDT eras in 458 cases

> Carol L. Shields, M.D. Lauren A. Dalvin, M.D. Li-Anne S. Lim, M.D. Michael Chang, M.D. Sanika Udyaver, B.S. Mehdi Mazloumi, M.D. Arman Mashayekhi, M.D. Jerry A. Shields, M.D.

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