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The Prevalence of Macular Telangiectasia Type 2 (MT2) in the Beaver Dam Eye Study

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Abstract

Purpose—To examine the prevalence of macular telangiectasia type 2 and lesions characterizing it.

Design—Population-based cohort study.

Setting: City and township of Beaver Dam, 1988–1990.

Study Population: 4790 people 43–86 years of age.

Observation Procedure(s): Grading from stereoscopic fundus photographs to measure macular telangiectasia type 2.

Main Outcome Measure: Prevalent macular telangiectasia type 2.

Results—Macular telangiectasia type 2 was present at baseline in 0.1% of the population (95% Confidence Interval [CI] 0.09, 0.1). The frequencies of loss of retinal transparency, crystals in the inner retinal layers, blunted retinal vessels, localized intraretinal pigment migration in the juxtafoveolar region, and presence of yellow deposits and lamellar holes in the foveal area in those

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without macular telangiectasia type 2 varied from 0.06% for retinal telangiectatic vessels to 1.2% for lamellar holes. Smoking was associated with pigment clumping (Odds Ratio [OR] per pack year 1.02; 95% CI 1.00, 1.03; $P=0.02$), RPE depigmentation (OR 1.02 per pack year; 95% CI 1.00, 1.04; $P=0.02$), loss of transparency (OR 1.02 per pack year; 95% CI 1.00, 1.03; $P=0.008$) and the presence of a yellow spot in the fovea (OR 2.24 current vs. never smoker; 95% CI 1.29, 3.89; $P=0.004$) but not with presence of macular telangiectasia type 2 (OR 2.72; 95% CI 0.45, 16.28; $P=0.27$).

Conclusions—The prevalence of macular telangiectasia type 2 (0.1%) is higher than previously thought. These data are useful in estimating the burden of this condition in the population. The role of smoking in the development of macular telangiectasia type 2 requires further study.

Macular telangiectasia type 2 is characterized by abnormal juxtafoveolar retinal capillaries and is usually accompanied by one or more of the following lesions on ophthalmoscopic examination: a loss of retinal transparency, crystals in the inner retinal layers, blunted retinal vessels, and localized intraretinal pigment migration in the juxtafoveolar region and the presence of yellow deposits and lamellar holes in the foveal area (Figure 1).^{1–4} In the earlier stages there is a change in color (gray) and apparent loss of retinal transparency in the juxtafoveolar retina. In its later stages, retinal pigment epithelial (RPE) depigmentation, RPE atrophy and subretinal new vessels may develop. The pathogenesis of macular telangiectasia type 2 is poorly understood.

Macular telangiectasia type 2 is usually asymptomatic in its earliest stage and is often not discovered until persons, usually in their fifth through eighth decades of life, present with decreased visual acuity. Diagnostically, fluorescein angiography and more recently, spectral domain optical coherence tomography (OCT), fundus autofluorescence and microperimetry testing have been used to identify and characterize macular telangiectasia type 2.^{5–10}

The disease is said to be infrequent. However, there has been only one population-based study which described its prevalence using gradings of non-stereoscopic fundus images.¹¹ The purpose of this paper is to examine the prevalence of macular telangiectasia type 2 and lesions characterizing it in the large population-based Beaver Dam Eye Study.

METHODS AND MATERIALS

Population

Methods used to identify and describe the population have appeared in previous reports.^{12–16} In brief, a private census of the population of Beaver Dam, Wisconsin, was performed from September 15, 1987, to May 4, 1988, to identify all residents in the city or township of Beaver Dam who were 43 to 84 years of age. Of the 5,924 eligible individuals, 4,926 participated in the baseline examination between March 1, 1988, and September 14, 1990.^{12,13} Ninety-nine percent of the population was white. Year 5 examinations occurred between 1993 and 1995 in 3,722 participants,¹⁴ year 10 in 1998 to 2000 in 2,962 participants,¹⁵ and year 15 in 2003 to 2005 in 2,375 participants.¹⁶ The fourth follow-up examination began in 2008 and is still in progress. Comparisons between participants and non-participants at the time of the baseline, 5-, 10-, and 15-year follow-up examinations have appeared elsewhere.^{13–16}

Procedures and Definitions

Similar procedures, used at both the baseline and follow up examinations, have been described in detail elsewhere.^{16–24} Pertinent parts of the examination included measuring the refractive error, best corrected visual acuity for distance, weight, height, blood pressure and administering a standardized questionnaire with questions about medications taken and chronic conditions (e.g., hypertension, diabetes, emphysema, cardiovascular disease, physical activity). Blood was drawn for measurements of serum lipid levels, white blood cells, platelets and glycosylated

hemoglobin. Stereoscopic 30° color fundus photographs centered on the fovea (Diabetic Retinopathy Study [DRS] standard field 1) and the macula (DRS standard field 2) and a single image temporal to the macula (DRS standard field 3) were taken. The presence and severity of lesions associated with macular telangiectasia type 2 (Figure 1) were graded using a standard protocol that evaluates the individual lesions of macular telangiectasia type 2 (loss of retinal transparency, crystalline deposits, dilated tortuous capillaries, blunted retinal vessels, localized intraretinal pigment migration, yellow foveal deposits and lamellar holes in the foveal area, RPE depigmentation and RPE atrophy) were developed prior to beginning grading. Procedures included grading all eyes at baseline for the presence of macular telangiectasia type 2 lesions. Macular lesions consistent with small branch vein occlusion or macular telangiectasia type 1, including lipid exudates, cystoid macular edema and visible aneurysms, were excluded. If any one of the macular telangiectasia type 2 lesions was graded as questionable or definitely present, a second grading was done by a different grader. If the two graders agreed as to the status of the lesions the grading was complete. If the second grader disagreed with the first grader, adjudication was performed by a retinal specialist (RK). Additionally, 229 eyes from the same population with no, questionable, or definite macular telangiectasia type 2 lesions were regraded by a different grader at the same time. Twenty-three of these eyes had a lesion regraded from absent to present. Three eyes had a lesion grade changed from present to cannot grade during the adjudication process. The most common lesion changed during the regrade was lamellar hole. All fundus photographs of eyes suspicious for having definite macular telangiectasia type 2 (two or more different types of macular telangiectasia type 2 lesions) were examined by three retinal specialists (BB, EC, RK). Definite macular telangiectasia type 2 was determined to be present in eyes with two or more different types of macular telangiectasia type 2 lesions was agreed upon by the three retinal specialists.

Current age was defined as the age at the time of the examination. Hypertension was defined as a mean systolic blood pressure of 140 mmHg and/or a mean diastolic blood pressure and/or a history of hypertension using antihypertensive medication at the time of the examination. Body mass index was defined as body weight (kg)/height (m²). Cigarette smoking, pack years smoked, and current heavy drinking have been previously defined elsewhere.²⁵

Statistical Methods

For these analyses, we examined the prevalence of macular telangiectasia type 2 and the lesions defining it by age and sex. For the purposes of this report, only the 4780 people (97.0%) with at least one eye with gradable fundus photographs for macular telangiectasia type 2 without an excluded lesion are included in the analyses. Eyes with epiretinal membranes with traction on the retina or eyes with signs of age-related macular degeneration (AMD) resulting in increased retinal pigment or RPE depigmentation or atrophy were excluded. Also examined were the relationships of risk factors to the prevalence of the lesions. SAS v.9 (SAS Institute, Cary, NC) was used to analyze the data. Multivariate odds ratios (OR) and 95% confidence intervals (CI) were calculated from discrete logistic hazard models (logistic regression for prevalence).²¹

RESULTS

Agreement among graders (absent vs present) for specific macular telangiectasia type 2 lesions varied from 100% (for yellowish spots) to 77% (for lamellar holes). Macular telangiectasia type 2 was present at baseline in 0.1% of the population (95% CI 0.09, 0.1). It was present in one woman and four men (Table 1). The mean age at baseline of those with macular telangiectasia type 2 was 63.4 (range 52–68) years; the mean age of those without macular telangiectasia type 2 was similar (62.3 [range 43–86] years). None of those with macular telangiectasia type 2 were related. Macular telangiectasia type 2 was bilateral in 2 of the 5

persons. Visual impairment (best corrected visual acuity of 20/40 or less) was present in 2 (29%) of affected eyes.

The frequencies of specific macular telangiectasia type 2 lesions in those without macular telangiectasia type 2 were low, varying from 0.06% for retinal telangiectatic vessels to 1.2% for lamellar holes (Table 2). When present, retinal blunted vessels, retinal crystalline deposits, and lamellar holes were more likely to be bilateral (Table 2). Of the 160 eyes with a lesion characterizing macular telangiectasia type 2, 131 (81.9%) had only one type of lesion, 26 (16.2%) had two types of lesions, 3 (1.9%) had three types of lesions, and none had 4 or more types of lesions. When two types of lesions characterizing macular telangiectasia type 2 were present, the most common pairs were lamellar holes and yellowish spots in right eyes and blunted vessels and lamellar holes in left eyes. Lesions characterizing macular telangiectasia type 2, when present, were most common in the inner temporal subfield and least frequent in the inner inferior subfield. The presence of any of the lesions in the absence of macular telangiectasia type 2 was unrelated to visual impairment.

Smoking was associated with pigment clumping, RPE depigmentation, loss of transparency and the presence of a yellowish spot in the fovea (Table 3) but not to macular telangiectasia type 2 (OR 2.72; 95% CI 0.45, 16.28; $P=0.27$). Persons with higher hematocrit were less likely and those who were physically active were more likely to have retinal blunted vessels present. White blood cell count was inversely associated with retinal telangiectatic vessels. Platelet count was associated with loss of retinal transparency and inversely associated with macular telangiectasia type 2. Cataract surgery was associated with presence of retinal blunted vessels ($P=.05$), retinal telangiectatic vessels ($P=.02$), and loss of retinal transparency ($P=.003$). No systemic or ocular characteristics were found to be associated with retinal crystalline deposits or lamellar holes. There were no associations of diabetes, hypertension, history of cardiovascular disease, presence of chronic kidney disease, history of aspirin use, or intraocular pressure with any of the lesions characterizing macular telangiectasia type 2 (data not shown).

Of the five persons with macular telangiectasia type 2 at baseline, follow-up varied from 0 to 20 years (Table 1, Figures 2 and 3). At baseline, only one of the uninvolved eyes had a sign of macular telangiectasia type 2 (blunted retinal vessels temporal to the macula [Figure 2]). Of the 3 persons with unilateral involvement at baseline and followed for 10 or more years, one eye with retinal blunted vessels had progressed to macular telangiectasia type 2 after 10 years of follow-up, developing retinal crystals, pigment clumping, and RPE depigmentation temporal to the macula (Figure 2).

A sample of 30 study participants without MT2 but having one of more MT2 lesions present at baseline were randomly selected for grading at the 5, 10, 15 and 20 year visits. None of these or their fellow eyes went on to develop MT2 at follow-up.

DISCUSSION

Macular telangiectasia type 2 was present in 0.1% of the predominantly white Beaver Dam Eye Study cohort 43–86 years of age. Macular telangiectasia type 2 is considered by many retinal specialists to be a rare condition. Our data suggest that it may be more common than previously thought.^{11,26} This may be due, in part, to its being asymptomatic in its early stages and to possibly being misclassified as age-related macular degeneration or a pattern dystrophy.

Based on the United States white population this age, we estimate there are approximately 70,000 persons with this condition. Because none of the participants in this population-based study had fluorescein angiography or OCT, the prevalence of macular telangiectasia type 2 as determined solely by grading color fundus photographs may have led to an underestimate of its prevalence. It is possible that fine retinal telangiectatic vessels were missed in eyes with

other signs characterizing macular telangiectasia type 2, such as lamellar holes or loss of retinal transparency. The use of spectral domain OCT and other new noninvasive imaging techniques in future population based studies may provide more precise estimates of macular telangiectasia type 2.¹⁰

The prevalence of macular telangiectasia type 2 in the Beaver Dam cohort was higher than the prevalence of macular telangiectasia type 2 in the Melbourne Collaborative Cohort Study, where it was estimated to range from 0.004 to 0.022%.¹¹ The lower prevalence in the Melbourne study may have been due to the fact that only 17% of images that had one or more findings (e.g., diabetic retinopathy, epiretinal membrane, increased pigment) that might indicate the presence of macular telangiectasia type 2 were selected for grading. This may have led to missing eyes with retinal signs of macular telangiectasia type 2 in the 83% of the cohort not graded for macular telangiectasia type 2 in that study.

Data from some studies suggest a possible genetic predisposition to macular telangiectasia type 2.^{26–29} In the Beaver Dam cohort, none of the persons with macular telangiectasia type 2 were related. There were too few persons in our cohort with this condition or lesions characterizing it to reliably examine their heritability.

Clinical lesions characterizing macular telangiectasia type 2 identified by grading of fundus photographs were infrequent in the Beaver Dam population. These lesions were examined because they have been found at different stages in eyes with macular telangiectasia type 2. Some of the lesions, including lamellar holes and yellow foveal deposits, are non-specific in nature. Data from case series have suggested associations of hypertension, diabetes, coronary artery disease and smoking with macular telangiectasia type 2.^{1,3,4,29–32} Aside from smoking, few systemic factors were consistently associated with macular telangiectasia type 2 lesions in the absence of the disease in the Beaver Dam cohort. Green et al. have speculated that the telangiectatic appearance of the retinal capillaries may be a result of focal endothelial defects.³³ It is possible that smoking, through its effect on endothelial function, may exacerbate this process. Smoking, through its inflammatory and hypoxic effects, may also exacerbate processes such as chronic leakage from hyperpermeable capillaries and ischemia, hypothesized to be involved in the progressive degeneration of the neuronal and glial cellular elements leading to lamellar holes in eyes with macular telangiectasia type 2.³⁴

While there are many strengths to this study, there are also limitations aside from the inherent low prevalence of macular telangiectasia type 2. First, misclassification may have led to an underestimate of the frequency of macular telangiectasia type 2. Second, the concomitant low frequency of some risk factors (e.g., diabetes) and of the prevalence of the macular telangiectasia type 2 lesions likely limited our ability to detect (or reject) meaningful relations. Third, some findings which may be of potential biological significance may be entirely due to chance given the number of associations examined. Fourth, the findings cannot be generalized to blacks, Hispanics and other racial ethnic groups.

In summary, we have provided descriptive epidemiology of macular telangiectasia type 2 showing a prevalence of 0.1% in persons 43–86 years of age. Use of additional noninvasive diagnostic approaches such as spectral domain OCT in epidemiological studies should provide more precise estimates of the prevalence and incidence of this condition. The role of one modifiable factor, smoking, in the development of macular telangiectasia type 2 lesions requires further study.

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D. *Statement about Conformity with Author Information*: Institutional Review Board approval was granted by the Health Sciences Institutional Review Board at the University of Wisconsin, Madison. Written informed consent for the use and disclosure of protected health information, compliant with the provisions of HIPAA, was obtained from all subjects before being enrolled in the study.

E. *Other Acknowledgments*: None.

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Figure 1.

Lesions characterizing macular telangiectasia type 2 from patients not participating in the Beaver Dam Eye Study. A. Loss of transparency; B. Retinal crystals (black arrow) and pigment clumping (white arrow); C. Lamellar hole (black arrow), yellowish spot, and retinal vessel blunting (white arrow); D. Telangiectatic retinal vessels (black arrow), pigment clumping, and RPE depigmentation.

Figure 2.

Development of macular telangiectasia type 2 in right eye of participant categorized as having macular telangiectasia type 2 in left eye. L. Baseline - left eye with blunted retinal vessels, retinal pigment clumping, retinal crystalline deposits, RPE depigmentation, and retinal telangiectatic vessels (black arrow); 5 year follow-up visit - a temporal lesion of pigment clumping and depigmentation has appeared with increasing size at the 10 year and 15 year follow-up visits (white arrow); R. Baseline and 5 year follow-up visit - right eye with beginnings of blunted retinal vessels and retinal crystals; 10 year follow-up visit - now labeled as having macular telangiectasia type 2 with retinal crystals, pigment clumping, and RPE depigmentation temporal to the macula (black arrow); 15 year follow-up visit - a temporal lesion of retinal pigment clumping and depigmentation (white arrow) similar to the left eye has developed.

Figure 3.

Four other participants with macular telangiectasia type 2 at the Beaver Dam Eye Study baseline examination in 1988–90. A. Right eye with pigment clumping (black arrow), retinal crystals, retinal vessel blunting, and RPE depigmentation; B. Left eye of same person with lamellar hole (green arrow), pigment clumping, RPE depigmentation, and retinal crystals; C. Left eye with RPE depigmentation (black/white arrow), pigment clumping, retinal crystals, and a loss of retinal transparency; other eye with no signs of macular telangiectasia type 2 is not shown; D. Left eye with retinal crystals (yellow arrow), pigment clumping, and RPE depigmentation; other eye with no signs of macular telangiectasia type 2 is not shown; E. Right eye with loss of retinal transparency (gray arrows), retinal vessel blunting, retinal crystals, and RPE depigmentation; other eye with no signs of macular telangiectasia type 2 is not shown; F. Left eye with retinal crystals, pigment clumping, and RPE depigmentation; other eye with no signs of macular telangiectasia type 2 is not shown.

pigment clumping and RPE depigmentation; F. Left eye of same person with a lamellar hole, loss of retinal transparency, vessel blunting, retinal crystals, pigment clumping and RPE depigmentation.

Characteristics of Persons with Macular Telangiectasia Type 2 in the Beaver Dam Eye Study, 1988–1990.

Table 1

Participant	Age	Sex	Eye Involved	Baseline VA		Years of Follow-up	Last VA		Lesions Present at Baseline	
				Right	Left		Right	Left	Right	Left
1	67	M	Both	20/25	20/32	10	20/63	20/63	Blunted retinal vessels Loss of retinal transparency Retinal crystalline deposits Retinal pigment clumping RPE depigmentation	Blunted retinal vessels Loss of retinal transparency Lamellar hole Retinal crystalline deposits Retinal pigment clumping RPE depigmentation
2	66	M	Left	20/25	20/50	10	20/32	20/32	None	Retinal crystalline deposits Retinal pigment clumping RPE depigmentation
3	64	M	Both	20/25	20/50	0	N/A	N/A	Blunted retinal vessels Retinal crystalline deposits Retinal pigment clumping RPE depigmentation	Loss of retinal transparency Lamellar hole Retinal pigment clumping Retinal crystalline deposits RPE depigmentation
4 ^a	52	M	Left	20/15	20/15	20	20/25	20/25	Blunted retinal vessels	Blunted retinal vessels Retinal pigment clumping Retinal crystalline deposits RPE depigmentation Retinal telangiectatic vessels
5	68	F	Left	20/25	20/25	20	20/40	20/40	None	Loss of retinal transparency Retinal pigment clumping Retinal crystalline deposits RPE depigmentation

Abbreviations: M = male; F = female; VA = visual acuity; N/A = not applicable; RPE = retinal pigment epithelial

^aDeveloped macular telangiectasia type 2 by 10 year follow-up in right eye.

Table 2

Prevalence and Bilaterality of Retinal Lesions Characterizing Macular Telangiectasia Type 2 in the Beaver Dam Eye Study, 1988–1990.^a

Lesion	# at risk	# affected	% affected	% bilateral
Retinal blunted vessels	4747	56	1.18	0.00
Retinal pigment clumping	4738	7	0.15	0.00
Retinal crystalline deposits	4770	10	0.21	0.00
RPE depigmentation	4737	5	0.11	0.02
Lamellar hole	4770	56	1.17	0.02
Retinal telangiectatic vessels	4759	3	0.06	0.00
Yellowish spot	4773	56	1.17	0.04
Loss of retinal transparency	4741	10	0.23	0.00

Abbreviations: RPE = retinal pigment epithelium.

^aEstimates in persons without macular telangiectasia type 2. Number at risk varies due to exclusion criteria or inability to grade for a type of lesion.

Association of Systemic Characteristics to Macular Telangiectasia Type 2 Lesions and Macular Telangiectasia Type 2 in the Beaver Dam Eye Study, 1988–1990.

Table 3

Lesion	Characteristic	OR ^a	95% CI	P value
Blunted vessels	Hematocrit, per %	1.09	1.02, 1.17	0.009
	Sedentary lifestyle, exercise ≤ 3x/week vs >3x/week	0.45	0.30, 0.78	0.003
Pigment clumping	Pack years smoked, per pack year	1.02	1.00, 1.03	0.02
RPE depigmentation	Pack years smoked, per pack year	1.02	1.00, 1.04	0.02
Telangiectatic vessels	White blood cell count, per k/μL	0.41	0.17, 1.00	0.05
Loss of transparency	Pack years smoked, per pack year	1.02	1.00, 1.03	0.008
	Platelets, per k/μL	1.004	1.00, 1.01	0.04
Yellowish spot	History of emphysema, present vs absent	13.22	3.39, 51.63	0.0002
	Smoking status, current vs never smoker	2.24	1.29, 3.89	0.004
	History of heavy drinking, current vs never heavy drinker	4.06	1.59, 10.36	0.0034
Macular telangiectasia type 2	Platelet count, per k/μL	0.99	0.97, 1.00	0.04

Abbreviations: OR = odds ratio; CI = confidence interval, RPE = retinal pigment epithelium.

^aOdds ratios are based on comparisons to persons with macular telangiectasia type 2 lesions, except for comparisons for macular telangiectasia type 2; individuals with macular telangiectasia type 2 were excluded from the individual lesion analyses.