

Supplementary Online Content

Schönbach EM, Wolfson Y, Strauss RW, et al; ProgStar Study Group. Macular sensitivity measured with microperimetry in Stargardt disease in the Progression of Atrophy Secondary to Stargardt Disease (ProgStar) Study: Report No. 7. *JAMA Ophthalmol*. Published May 25, 2017. doi:10.1001/jamaophthalmol.2017.1162

eFigure 1: Modified Humphrey 10-2 Pattern (68 Points) for Sensitivity Testing of the Macula.

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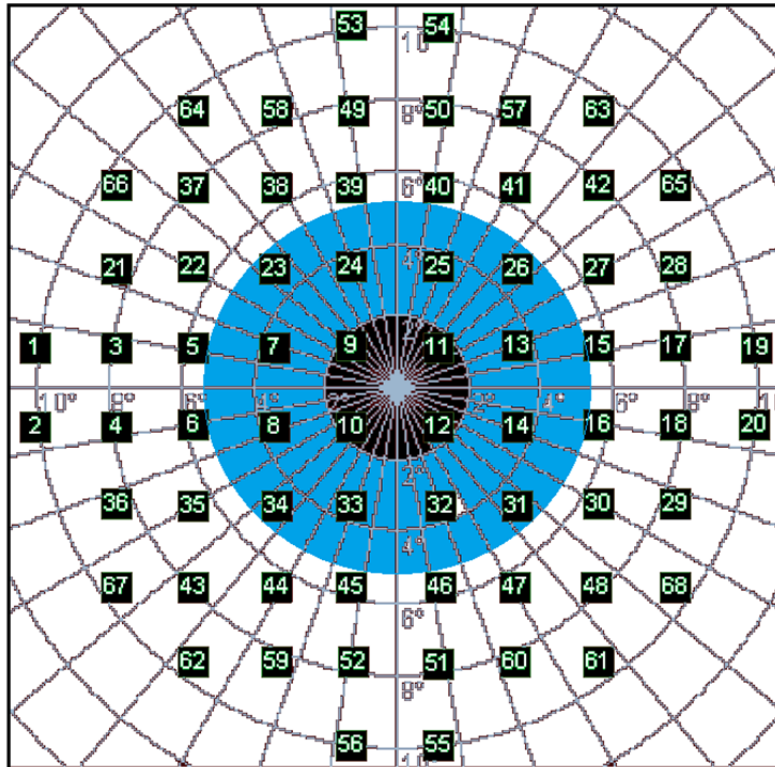
eTable 5: Macular Sensitivity in Stargardt Disease and Its Association with Visual Acuity including Patients with Poor Pattern Placement.

This supplementary material has been provided by the authors to give readers additional information about their work.

eFigure 1.

Title: Modified Humphrey 10-2 Pattern (68 Points) for Sensitivity Testing of the Macula.

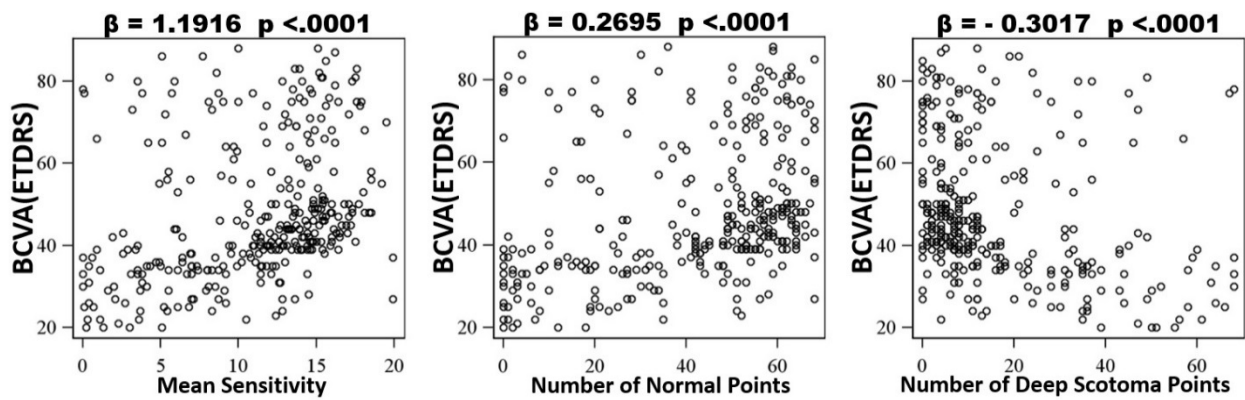
Legend: The central 4 points were grouped as foveal (black background), the 12 perifoveal points of the blue ring were grouped as the inner ring (blue background) and the remaining 52 test locations fell into the outer ring (white background).



eFigure 2.

Title: Correlation of Visual Acuity with Microperimetric Sensitivity

Legend: Scatterplots showing the correlation between best-corrected visual acuity (BCVA) in ETDRS letter score with microperimetric sensitivity measured as the number of normal test locations, relatively scotomatous test locations, deeply scotomatous test locations, and mean sensitivity. Univariate linear regression analyses.



Characteristic at baseline	Adequate and fair pattern placement in at least one eye N= 199 (63.8 % bilateral)	Poor or un-gradable pattern placement in both eyes N= 37	All participants N=236	p-value*
Person level				
Age in years (mean (SD))	34.2 (14.7)	30.7 (17.5)	33.6 (15.2)	0.20
% Female	57.3	48.7	55.9	0.33
Race				
% Asian/South Asian	3.5	5.4	3.8	0.02
% African American	7.5	13.5	8.5	
% White	87.9	73.0	85.6	
% Other	1.0	8.1	2.1	
Age of disease onset** in years				
(Mean(SD))	22.7 (12.6)	21.2 (14.4)	22.5 (12.8)	0.53
% ≤18 years	41.7	43.2	42.0	0.10
% >18	52.8	40.5	50.8	
% Unknown	5.0	13.5	6.4	
% Asymptomatic	0.5	2.7	0.8	
Duration of Disease** (mean(SD))	12.0 (9.5)	10.1 (9.2)	11.7 (9.5)	0.30
Eye level				
	Eyes with adequate and fair N=326	Eyes with poor or un-gradable N=114	All eyes N=440	
Best-corrected visual acuity (ETDRS letters) (Mean(SD))	47.8 (16.9)	41.1 (13.1)	46.1 (16.3)	<0.0001

eTable 1: Demographic Characteristics by Inclusion in the Analyses.

* Chi-Square/t-test were performed as appropriate.

** Duration & age of disease onset are missing for 15 participants.

SD= Standard Deviation

Variable	Foveal		Inner ring		Outer ring		Overall	
	Median	(Mean± SD; Range)	Median	(Mean± SD; Range)	Median	(Mean± SD; Range)	Median	(Mean± SD; Range)
Mean Sensitivity (dB)	0	(2.69± 4.43; 0 to 20)	6.17	(6.77± 5.81; 0 to 20)	14.46	(12.66± 5.3; 0 to 19.92)	12.4*	(11.02± 4.97; 0 to 19.90)
Count								
Deep scotoma points	4	(2.94± 1.39; 0 to 4)	5	(5.55± 4.39; 0 to 12)	0	(7.67± 12.94; 0 to 52)	10	(16.15± 16.88; 0 to 68)
Relative scotoma points	0	(0.58± 0.87; 0 to 4)	2	(2.29± 2.02; 0 to 12)	4	(7.57± 8.55; 0 to 37)	8	(10.43± 9.08; 0 to 43)
Normal points	0	(0.49± 1.03; 0 to 4)	3	(4.16± 4.16; 0 to 12)	46	(36.57± 17.36; 0 to 52)	49	(41.25± 20.8; 0 to 68)
Percentage of								
Deep scotoma points	100%	(73 %± 35 %; 0 %-100 %)	42 %	(46 %± 37 %; 0 %-100 %)	0%	(15 %± 25 %; 0 %-100 %)	15%	(24% ± 25%; 0% to 100%)
Relative scotoma points	0%	(15 %± 22 %; 0 %-100 %)	17 %	(19 %± 18 %; 0 %-100 %)	8%	(15 %± 17 %; 0 %-71 %)	12%	(15% ± 13%; 0% to 63%)
Normal points	0%	(12 %± 26 %; 0 %-100 %)	25 %	(35 %± 35 %; 0 %-100 %)	89%	(71 %± 33 %; 0 %-100 %)	72%	(61%± 31%; 0% to 100%)

eTable 2. Macular Sensitivity in Stargardt Disease.

N= 323 eyes. *N=322. SD= Standard Deviation

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Variable	Foveal		Inner ring		Outer ring		Overall	
	Median	(Mean± SD; Range)	Median	(Mean± SD; Range)	Median	(Mean± SD; Range)	Median	(Mean± SD; Range)
Mean Sensitivity (dB)	0	(2.9± 4.52; 0 to 20)	6.33	(6.7± 5.68; 0 to 20)	14.04	(12.4± 5.27; 0 to 19.92)	12.1*	(10.8± 4.97; 0 to 19.90)
Deep scotoma	4	(2.9± 1.41; 0 to 4)	5	(5.6± 4.3; 0 to 12)	1	(8± 12.7; 0 to 52)	10**	(16.5± 16.7; 0 to 68)
Relative scotoma	0	(0.61± 0.9; 0 to 4)	2	(2.4± 2.1; 0 to 12)	5	(8.3± 8.8; 0 to 41)	8**	(11.1± 9.43; 0 to 46)
Normal points	0	(0.53± 1.1; 0 to 4)	3	(4.1± 4.1; 0 to 12)	44	(35.5± 17.5; 0 to 52)	48**	(40.3± 20.99; 0 to 68)
Deep scotoma	100 %	(72 %± 35 %; 0 %- 100 %)	42 %	(47 %± 36 %; 0 %- 100 %)	2 %	(16 %± 25 %; 0 %- 100 %)	15%	24%±25%; 0% to 100%)
Relative scotoma	0 %	(15 %± 22 %; 0 %- 100 %)	17 %	(19 %± 18 %; 0 %- 100 %)	10 %	(16 %± 17 %; 0 %- 79 %)	12%	(16%±14%; 0% to 68%)
Normal points	0 %	(13 %± 26 %; 0 %- 100 %)	25 %	(35 %± 34 %; 0 %- 100 %)	85 %	(69 %± 34 %; 0 %- 100 %)	71%	(59%±31%; 0% to 100%)

3 eTable 3. Macular Sensitivity in Stargardt Disease including Patients with Poor
 4 Pattern Placement.

5 N= 427 eyes. *N= 405. **N= 428. SD= Standard Deviation

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Outcome	Independent variable	All eyes with graded microperimetric testing					
		Univariate			Multivariate		
		β estimate	p value	95% Confidence Limits	β estimate	p value	95% Confidence Limits
Mean Sensitivity (in dB)*	Current age (per year)	-0.074	0.0005	(-0.11 to -0.03)	-0.02	0.37	(-0.076 to 0.028)
	Current age \geq 50 yrs	-2.88	0.0004	(-4.47 to -1.30)			
	Age of onset > 18 yrs	-0.12	0.87	(-1.48 to 1.25)			
	Duration of Stargardt disease in years	-0.20	<.0001	(-0.27 to -0.14)	-0.18	<.0001	(-0.27 to -0.093)
	Male Gender	0.58	0.39	(-0.74 to 1.90)			
	Non-White Ethnicity	-0.40	0.67	(-2.23 to 1.43)			
Count of Normal Points	Current age (per year)	-0.30	0.0007	(-0.47 to -0.13)	-0.12	0.29	(-0.3 to 0.097)
	Current age \geq 50 yrs	-12.89	0.0001	(-19.43 to -6.35)			
	Age of onset > 18 yrs	-0.70	0.81	(-6.33 to 4.93)			
	Duration of Stargardt disease in years	-0.82	<.0001	(-1.10 to -0.53)	-0.71	0.0001	(-1.08 to -0.35)
	Male Gender	1.13	0.68	(-4.31 to 6.57)			
	Non-White Ethnicity	0.0096	0.998	(-7.22 to 7.24)			
Count of Deep Scotoma Points	Current age (per year)	0.27	0.0002	(0.13 to 0.41)	0.082	0.30	(-0.074 to 0.24)
	Current age \geq 50 yrs	9.42	0.0012	(3.70 to 15.14)			
	Age of onset > 18 yrs	1.24	0.59	(-3.30 to 5.79)			
	Duration of Stargardt disease in years	0.78	<.0001	(-0.06 to 0.17)	0.71	<.0001	(0.40 to 1.01)
	Male Gender	-1.81	0.42	(-6.18 to 2.56)			
	Non-White Ethnicity	1.88	0.55	(-4.29 to 8.05)			

eTable 4. Macular Sensitivity in Stargardt Disease and Its Association with Demographic Features including Patients with Poor Pattern Placement. N=427 eyes. *N=425.

Independent variable	All eyes with graded microperimetric testing (N= 427)					
	Univariate			Multivariate		
	β estimate	p value	95% Confidence Limits	β estimate	p value	95% Confidence Limits
MS all points	1.12	<.0001	(0.75 to 1.50)			
MS Fovea	0.87	<.0001	(0.51 to 1.23)	0.48	0.0224	(0.07 to 0.89)
MS Inner ring	0.98	<.0001	(0.66 to 1.30)	0.48	0.0508	(-0.002 to 0.96)
MS Outer ring	0.90	<.0001	(0.56 to 1.24)	0.39	0.0788	(-0.045 to 0.83)
Count						
Normal Points	0.24	<.0001	(0.15 to 0.32)			
Deep Scotoma	-0.28	<.0001	(-0.38 to -0.17)	-0.26	<.0001	(-0.37 to -0.16)

eTable 5. Macular Sensitivity in Stargardt Disease and Its Association with Visual Acuity including Patients with Poor Pattern Placement.

MS= mean sensitivity