



# Baseline Microperimetry and SD-OCT measures in the RUSH2A study

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PRESENTED ON BEHALF OF THE FOUNDATION FIGHTING BLINDNESS CONSORTIUM INVESTIGATOR GROUP



# Financial Disclosures

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# Summary

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**Baseline RUSH2A data revealed similar MP and SD-OCT metrics between the Usher syndrome type 2A (USH2A) and the non-syndromic autosomal recessive retinitis pigmentosa (ARRP) participants with mutations in the *USH2A* gene.**

**Longer disease duration was associated with more severe abnormalities of retinal structure and function, adjusted for clinical diagnosis.**

# Funding

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# Outline

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- Objectives
- Methods
- MP Results
  - Summary Data
  - Correlation with visual function/structure measures
- OCT Results
  - Summary Data
  - Correlation with Visual function/structure measures

# Objectives

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- To describe MP and OCT data of RUSH2A patients by clinical diagnosis at study baseline
- To evaluate baseline patient characteristics associated with MP mean sensitivity and OCT EZ
- To evaluate correlations among different visual functional and structural measures

# Methods

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## ➤ MP

- Optional for sites with MAIA
- Primary cohort only
- Study eye only

## ➤ OCT

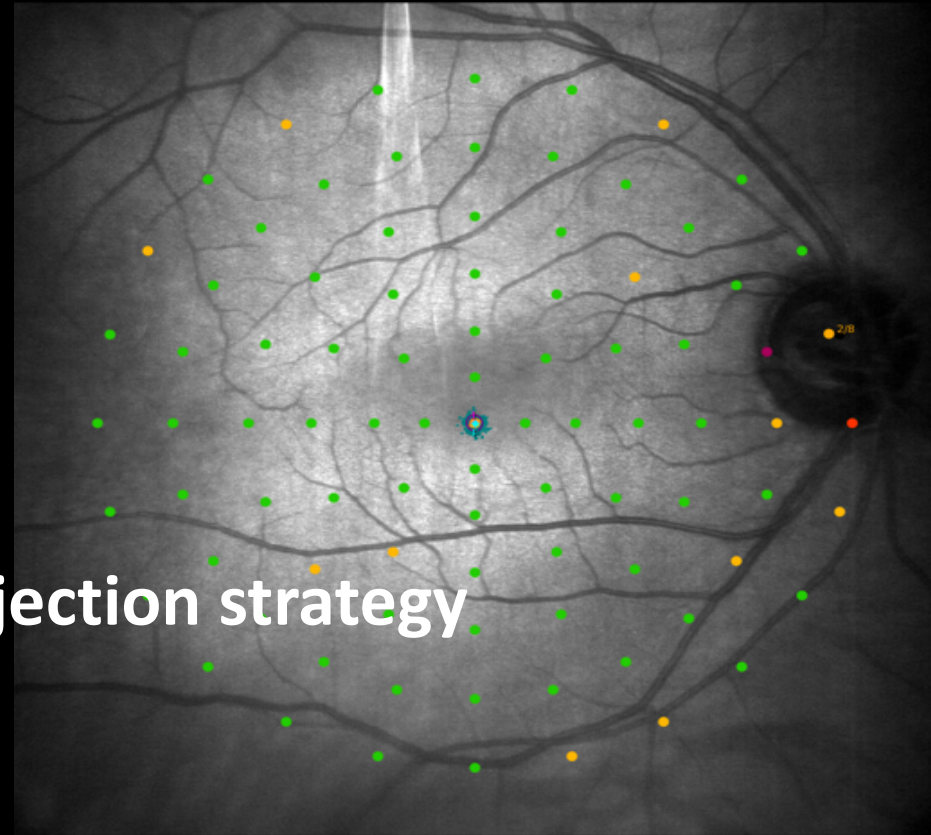
- All RUSH2A subjects
- Performed on both eyes

# Microperimetry (MP) in RUSH2A



MAIA (CenterVue)

- Mesopic/**standard** MAIA, 4-2 projection strategy
- Size-III Stimulus
- **Custom** 30 deg wide grid

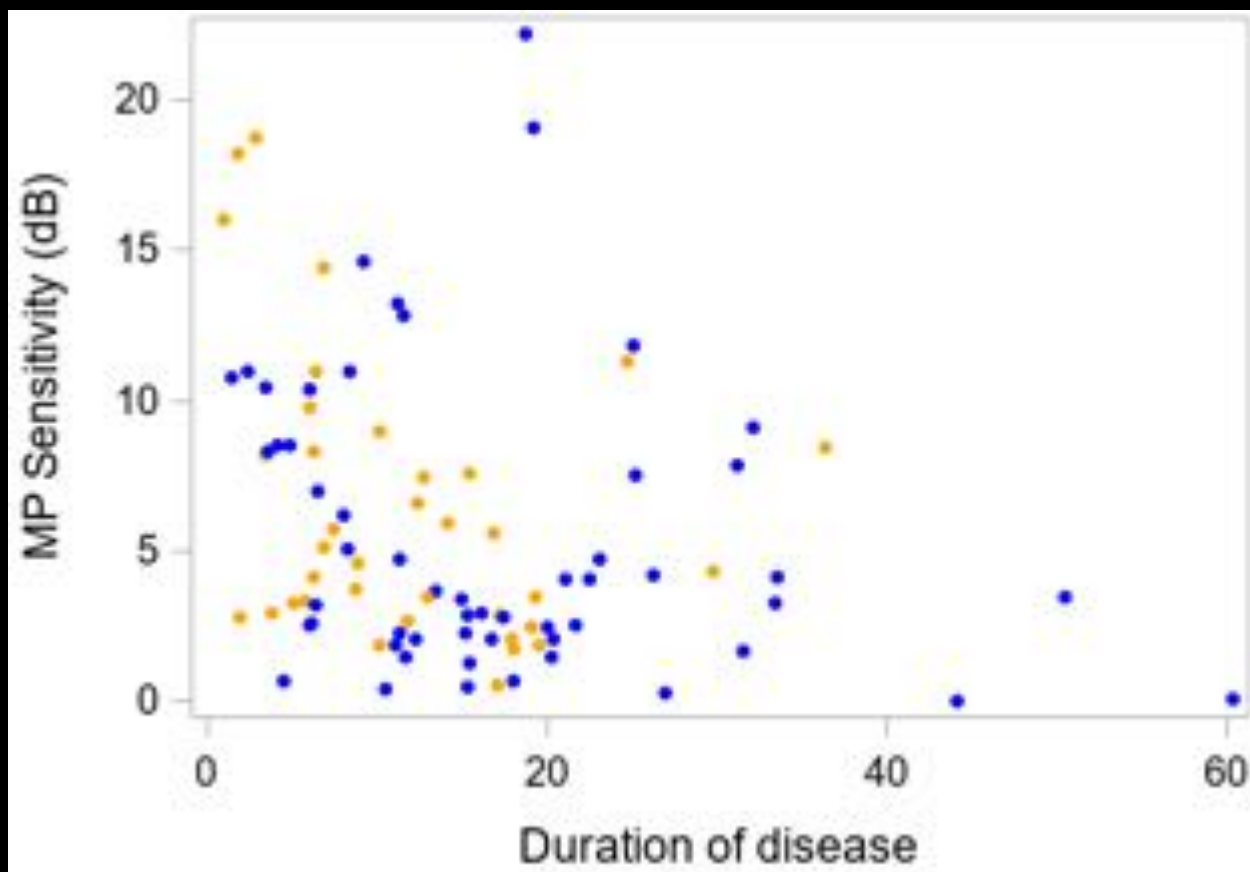




# MP Data

	All	Clinical Diagnosis		P-value
	N = 93	Usher syndrome N= 56	Non-syndromic RP N= 37	
MP Sensitivity (dB)				0.12
Median	4.1	3.5	5.1	
(IQR)	(2.5, 8.5)	(2.1, 8.5)	(3.0, 8.5)	
[Min, Max]	[0.0, 22.2]	[0.2, 22.2]	[0.6, 19.5]	
95% BCEA area (deg <sup>2</sup> )				0.28
Median	1.5	1.4	1.7	
(IQR)	(0.8, 2.8)	(0.8, 2.7)	(0.8, 2.8)	
[Min, Max]	[0.2, 57.2]	[0.2, 57.2]	[0.2, 15.5]	

# MP Sensitivity

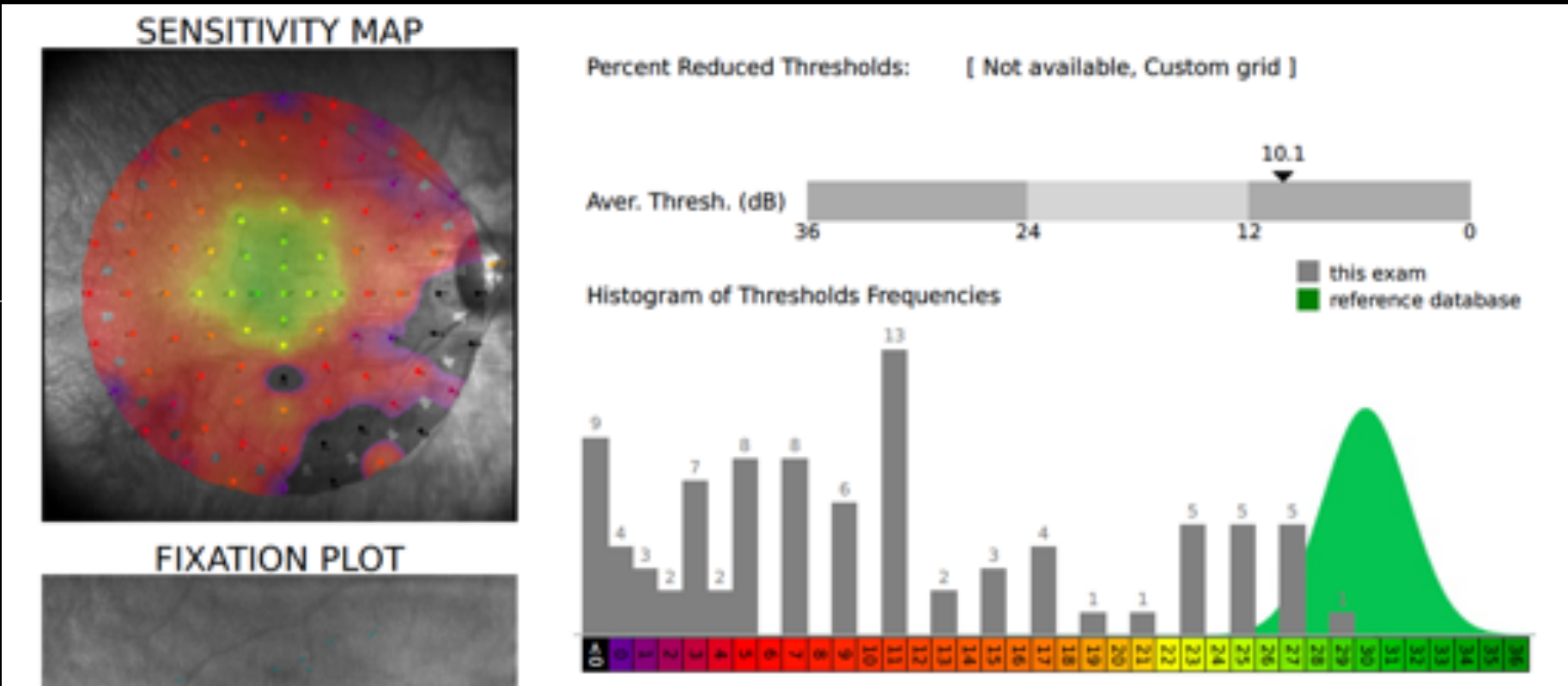


Usher 2: **blue**

Non-syndromic RP: **orange**

Duration of disease:  $P < 0.001$

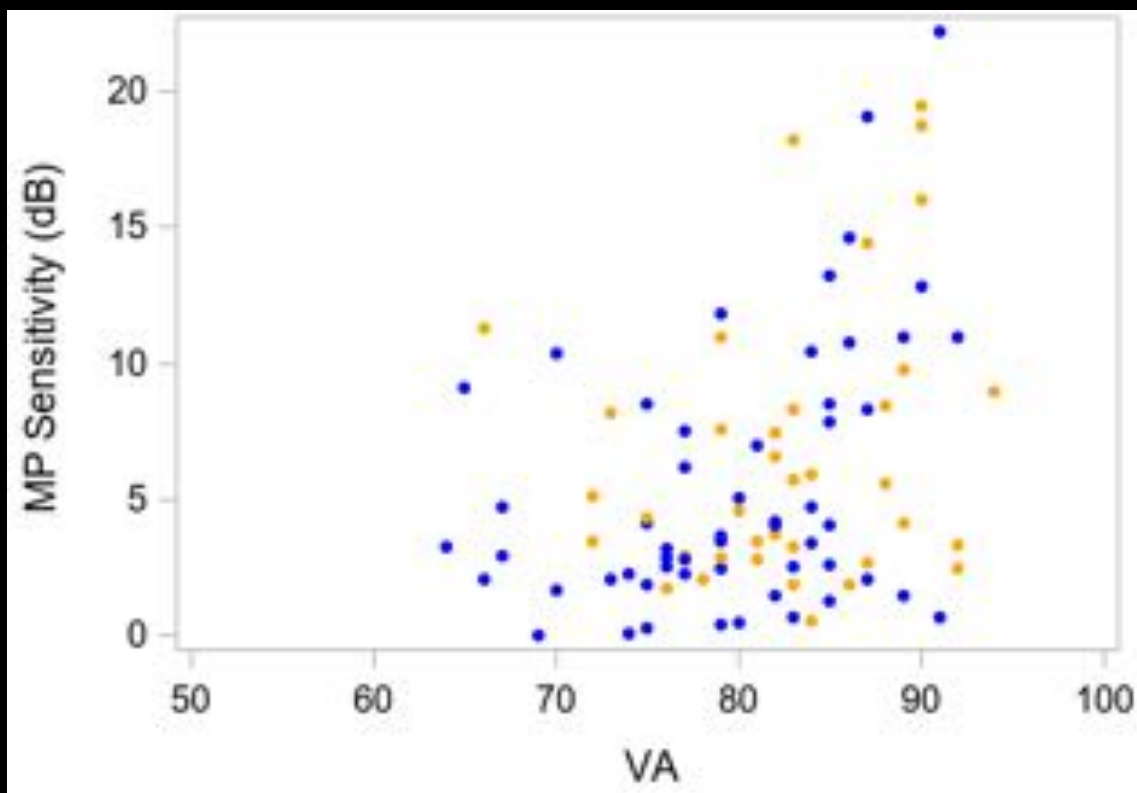
# MP Data (Cont.)



	All	Clinical Diagnosis		P-value
Number of loci with absolute scotoma <0 dB black (not seen)	N = 93	Usher syndrome N= 56	Non-syndromic RP N= 37	
Median	43	43	37	0.33
(IQR)	(21, 58)	(20, 64)	(24, 53)	
[Min, Max]	[1, 83]	[1, 83]	[1, 82]	

# VA correlated with MP sensitivity

Better VA -> Higher MP sensitivity



Usher 2: **blue**

Non-syndromic RP: **orange**

Spearman correlation

$r = 0.31$  (95% CI: 0.12, 0.49)

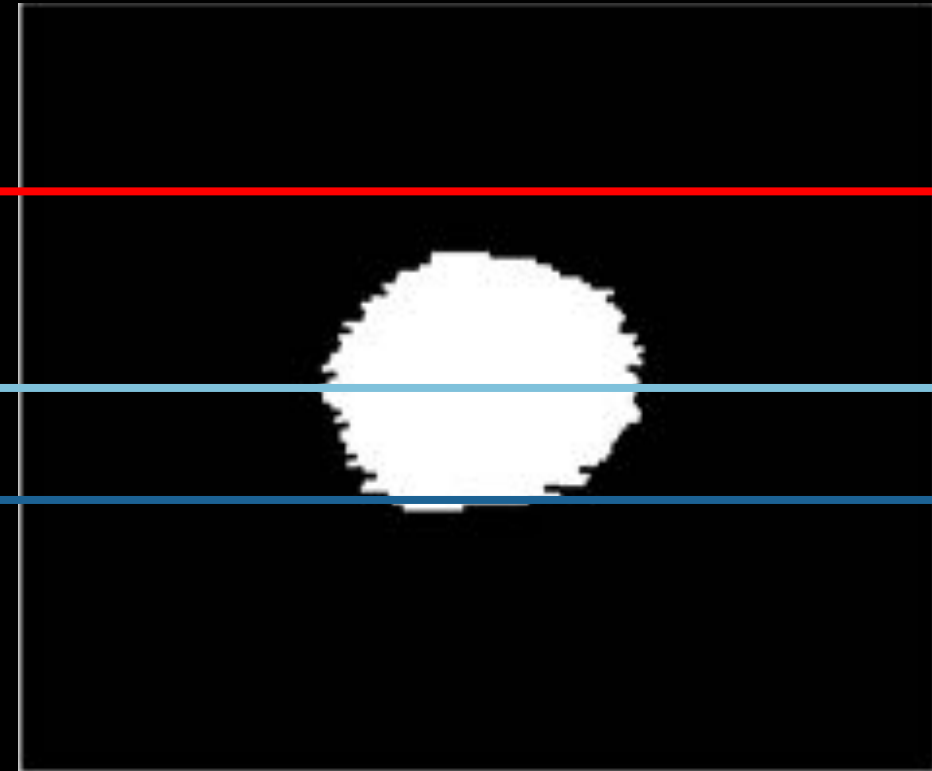
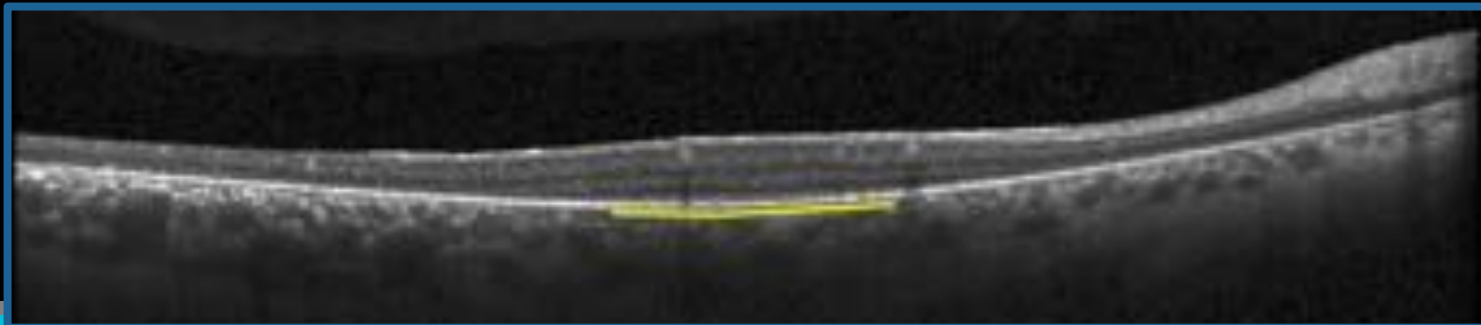
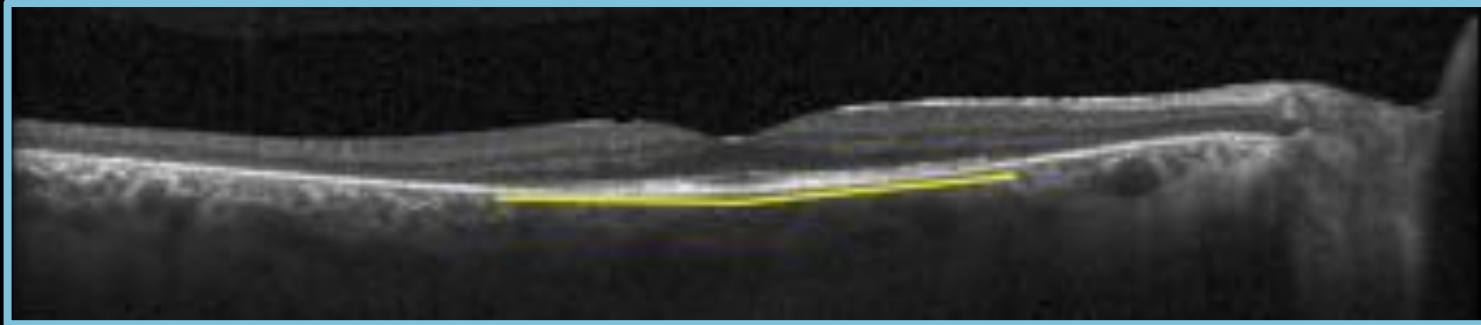
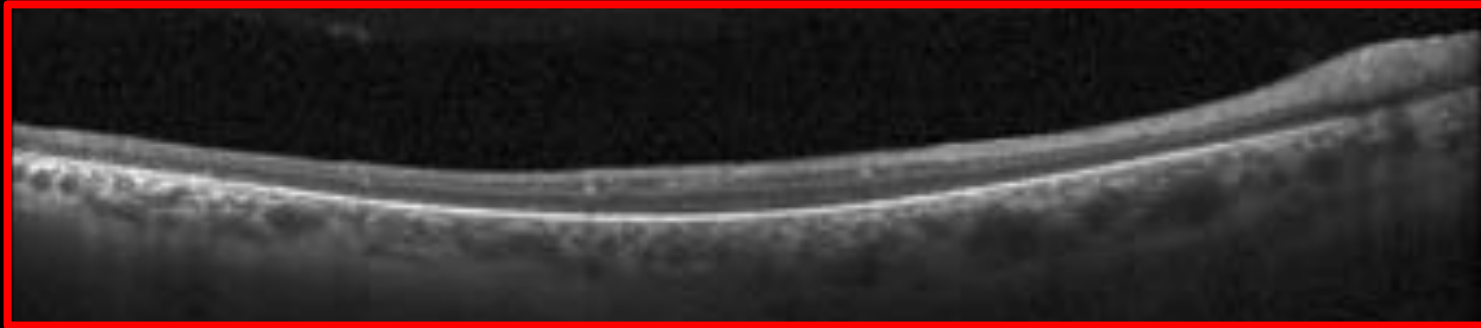
$P = 0.002$



# OCT VARIABLES

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# Segmentation of EZ on OCT

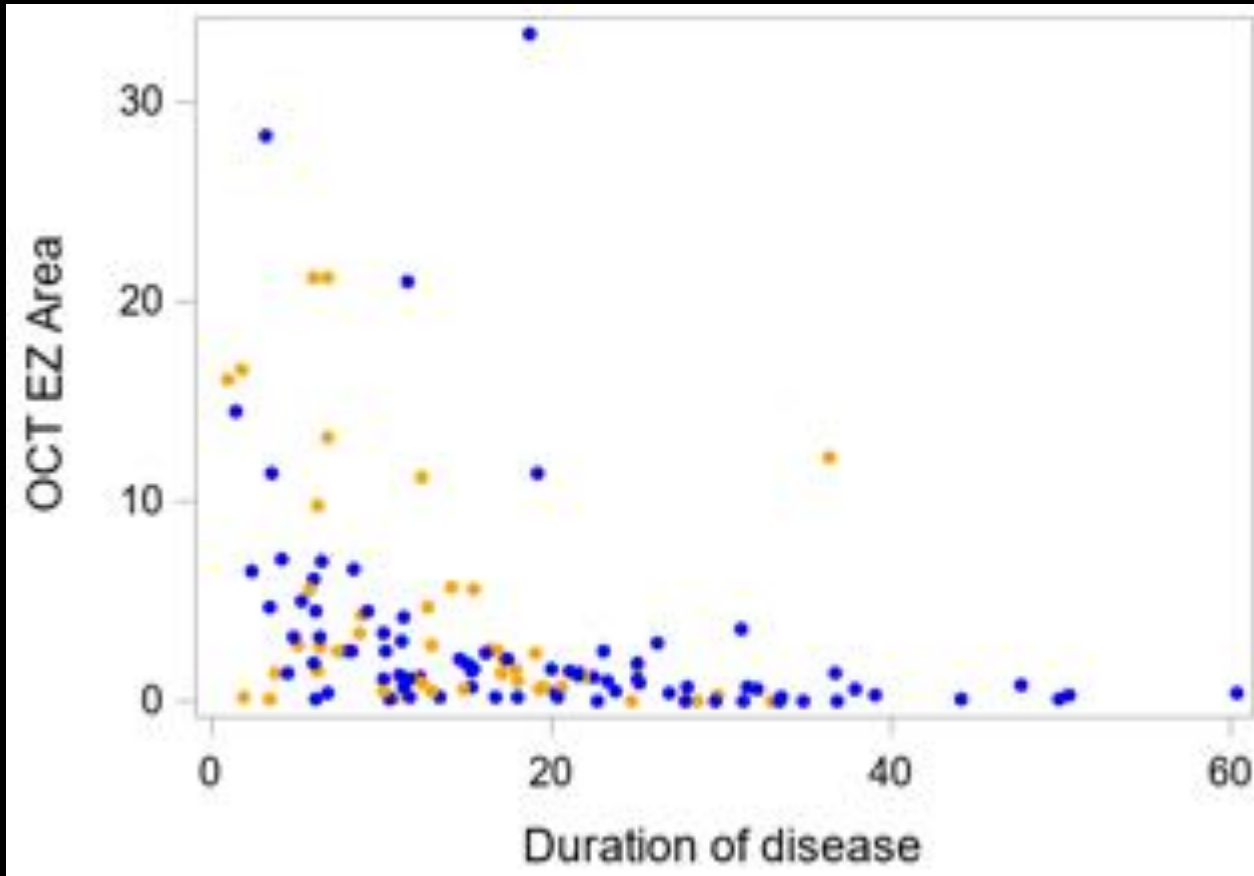


Intact EZ area

# OCT Data

	All	Clinical diagnosis	
	N = 127	Usher syndrome N= 80	Non-syndromic RP N= 47
Central Subfield Thickness ( $\mu\text{m}$ )			
Median	253	247	259
(IQR)	(228, 283)	(223, 280)	(246, 286)
[Min, Max]	[137, 519]	[137, 519]	[175, 323]
EZ Area ( $\text{mm}^2$ )			
Median	1.5	1.4	2.3
(IQR)	(0.5, 3.5)	(0.4, 3.1)	(0.7, 5.7)
[Min, Max]	[0.0, 33.4]	[0.0, 33.4]	[0.0, 21.3]

# Disease duration with OCT EZ area



Usher 2: **blue**

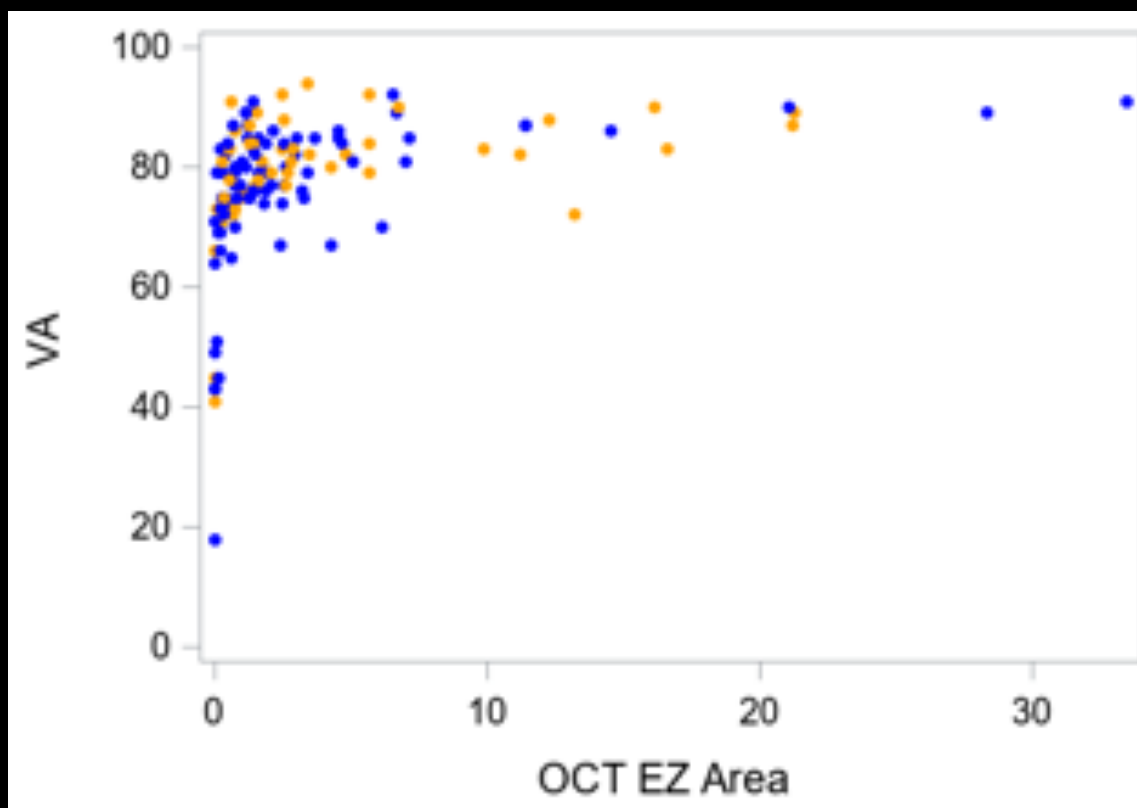
Non-syndromic RP: **orange**

Duration of disease:  $P < 0.001$



# VA correlated with OCT EZ area

Better VA -> Greater OCT EZ area



Usher 2: **blue**

Non-syndromic RP: **orange**

Spearman correlation

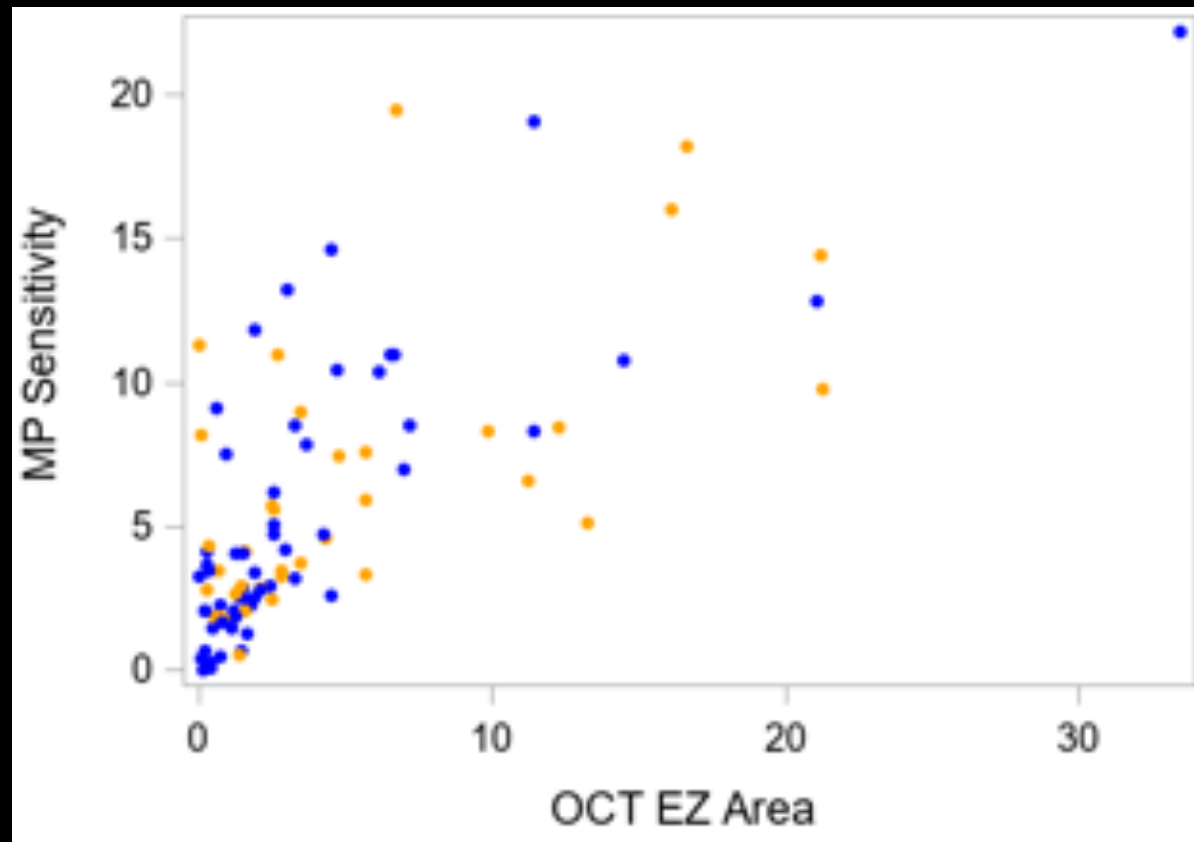
$r = 0.61$  (95% CI: 0.48, 0.71)

$P < 0.001$

# Structure-function correlation: OCT EZ with MP sensitivity



Greater OCT EZ area -> Higher average threshold



Usher 2: **blue**

Non-syndromic RP: **orange**

Spearman correlation

$r = 0.68$  (95%: 0.55, 0.78)

$P < 0.001$



# Conclusions

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**Baseline RUSH2A data revealed similar MP and SD-OCT metrics between the USH2A and non-syndromic ARRP participants.**

**Longer disease duration was associated with more severe abnormalities of retinal structure and function, adjusted for clinical diagnosis.**

**MP and OCT measures may provide useful metrics to monitor during disease progression in studies of *USH2A*-related retinal degeneration.**

*(Manuscript under development)*

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