UT Southwestern Medical Center

Clinical analysis of children presenting with ADEM with or without MOG-IgG

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2024 Rare Neuroimmune Disorders Symposium

What is ADEM?

- Acute disseminated encephalomyelitis
- Rapid onset and widespread inflammation of the brain and spinal cord
- Occurs throughout the entire lifespan, though >90% of all cases happen in children
- Typically presents as monophasic disorder associated with multifocal neurological symptoms and encephalopathy
- A subgroup of patients are positive for MOG-IgG

Study aim

- To characterize in children presenting with ADEM, a) clinical and paraclinical features and b) outcomes associated with or without MOG-IgG

Participants

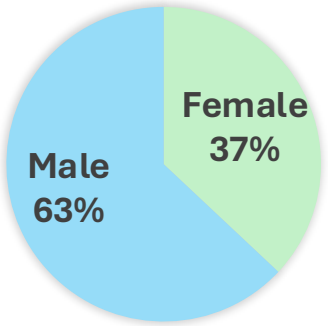
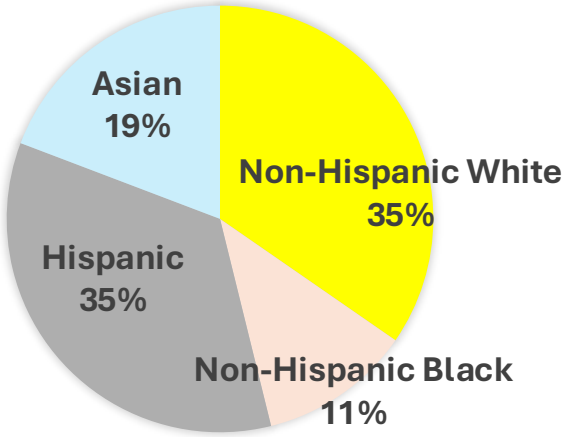
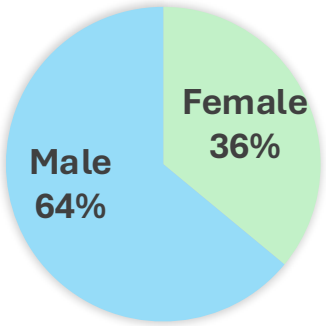
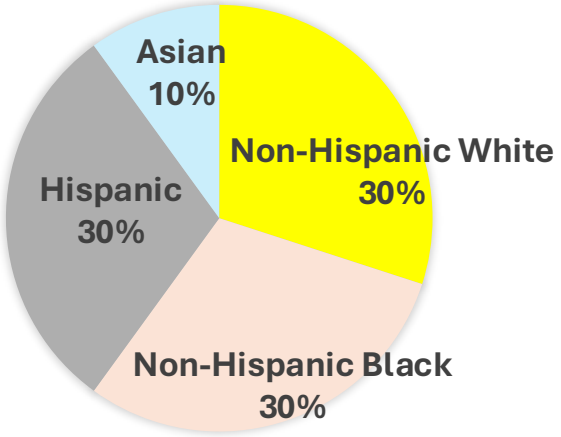
- Patients up to 18 years of age seen between 2017 and 2024 in the demyelinating disease clinic at Children's Health Dallas
- Diagnosed with ADEM
- Had serum MOG-IgG testing at disease onset

Demographics

- 38 children: 27 (71%) MOG positive, 11 (29%) MOG negative

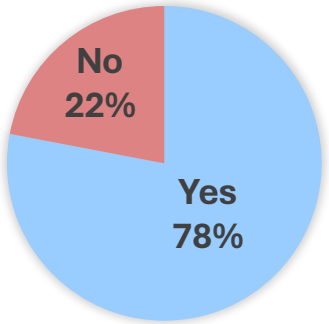
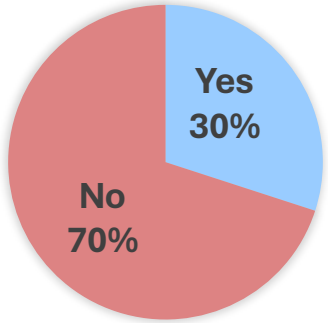
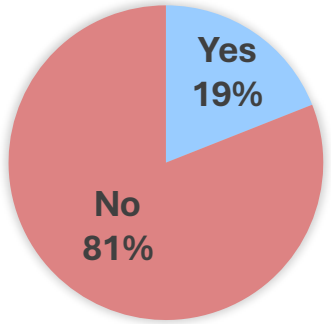
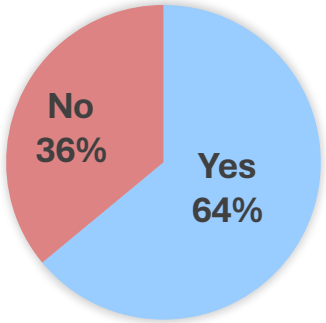
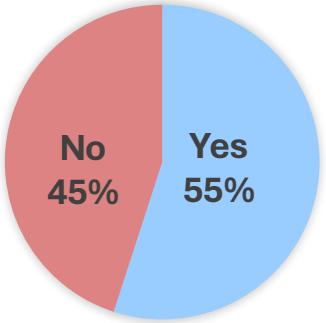
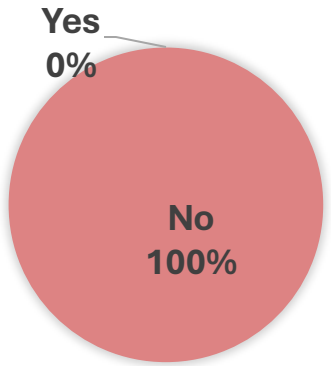
Demographics

- 38 children: 27 (71%) MOG positive, 11 (29%) MOG negative

	★ Age at onset median (IQR), years	Sex	Race/ethnicity
MOG+	5.0 (3.8-8.4)		
MOG-	8.3 (7.1-9.9)		

Acute phase

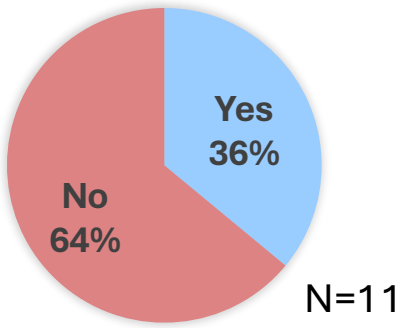
Presenting symptoms

	Preceding infectious symptoms	Seizure	★ Vision changes
MOG+			
MOG-			

MRI

MOG+

Optic nerve involvement



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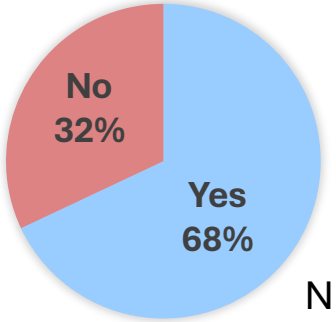
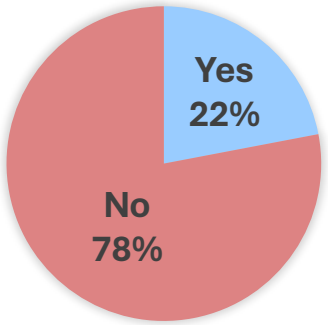
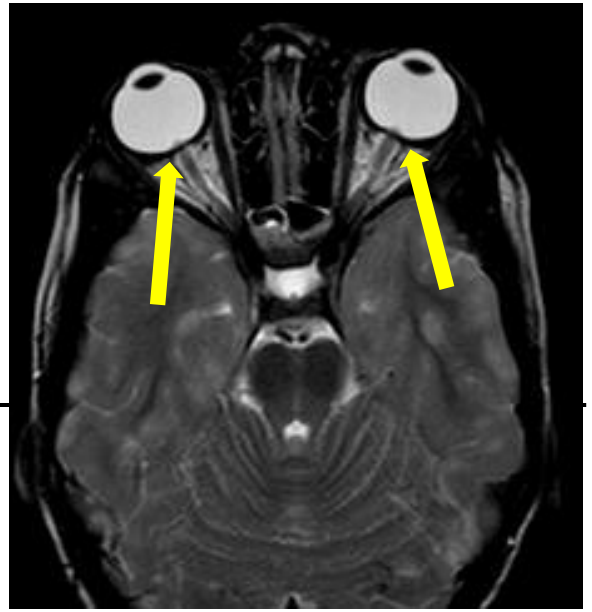
Spinal cord involvement



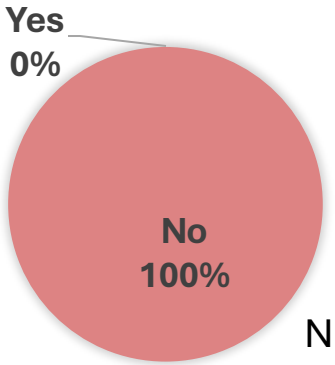
N=25

Posterior globe indentation





MOG-



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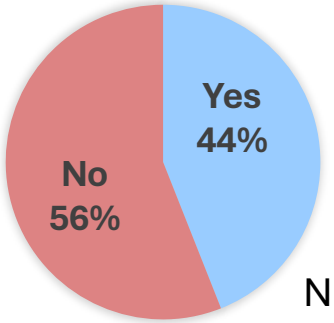

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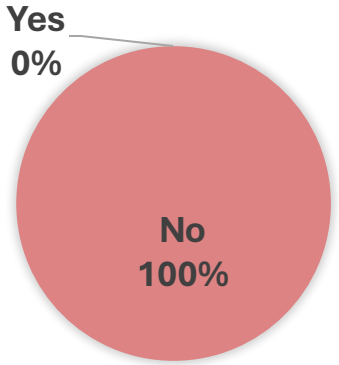


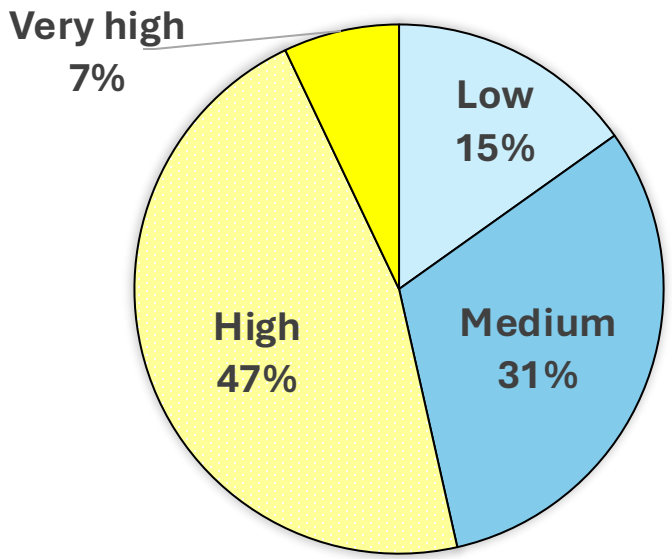
MRI lesion burden score

Area of abnormality	Score
Supratentorial white matter	1 for 25% involvement 2 for 50% involvement 3 for 75% involvement 4 for near complete involvement (max 4 points)
Cortex	1 for unilateral 2 for bilateral (max 2 points)
Deep gray matter	1
Brainstem	1
Cerebellum	1
Optic nerve	1
Spinal cord	1 for each region of involvement (cervical, thoracic, lumbar/conus) (max 3 points)
Total (max)	13

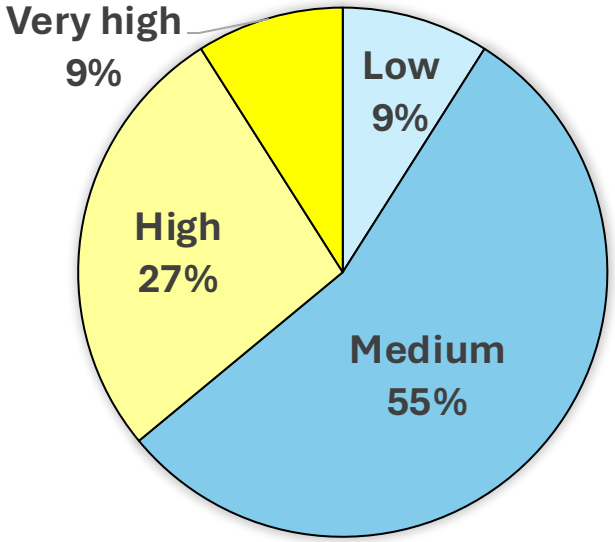
Cumulative score	Burden level
1-3	Low
4-6	Medium
7-9	High
10-13	Very High

MRI lesion burden score

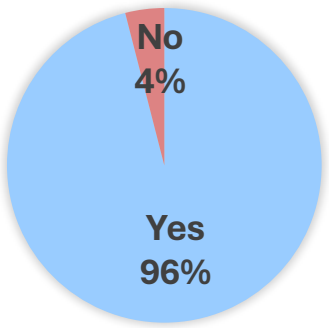
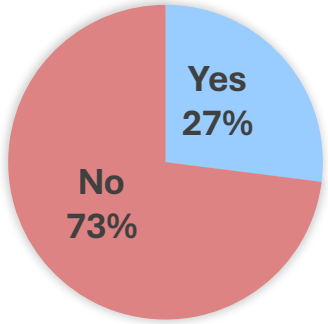
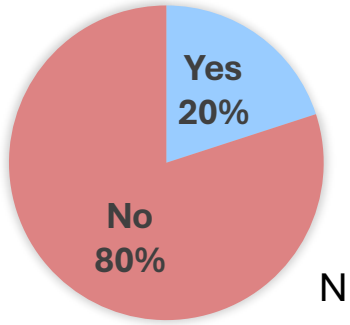
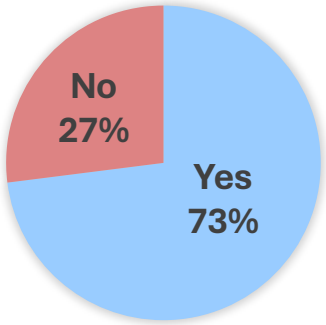
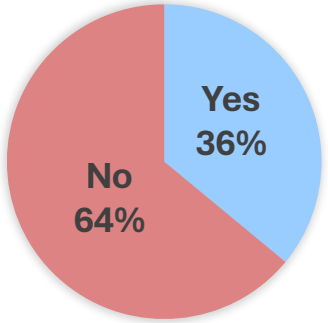
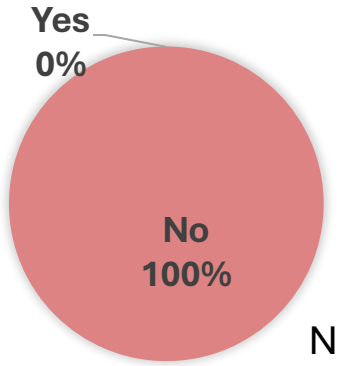
MOG+



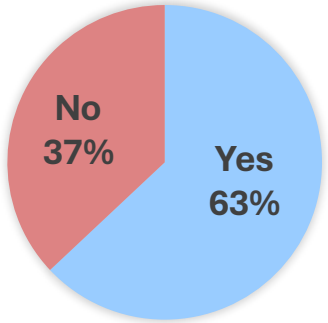
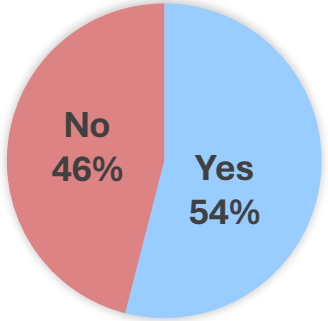
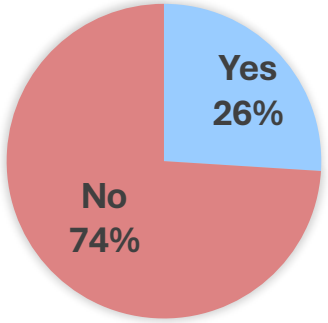
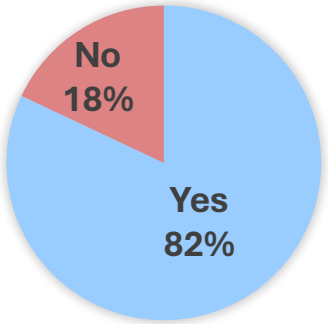
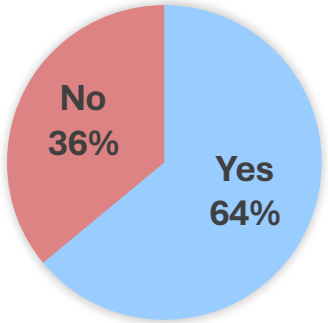
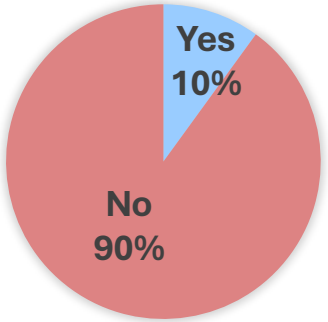
MOG-



Cerebral spinal fluid analysis

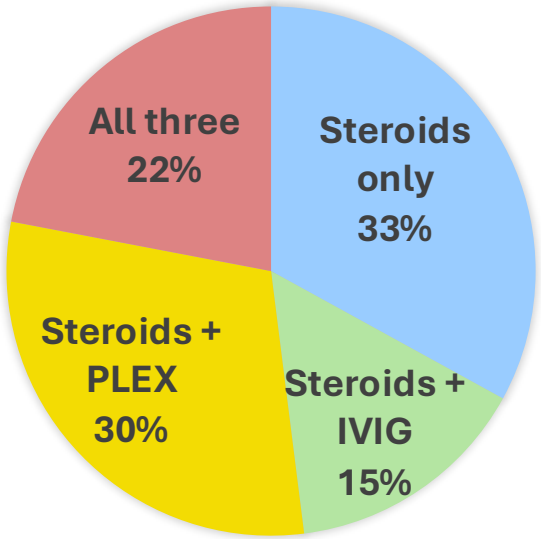
	★ Total nucleated cells >5/mm³	Protein >45 mg/dL	★ Opening pressure >28 mmH₂O
MOG+			 N=10
MOG-			 N=4

Hospital course

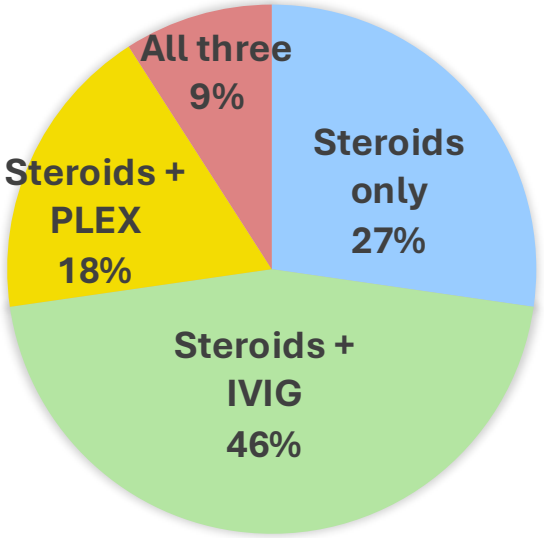
	Hospital duration median (IQR), days	ICU stay	Unable to walk without assistance	★ Increased intracranial pressure management
MOG+	13.0 (6.0-26.1)			
MOG-	13.0 (11.5-22.0)			

Acute treatments

MOG+

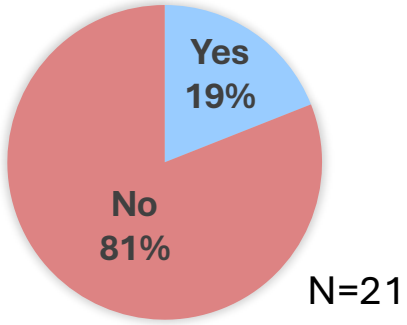
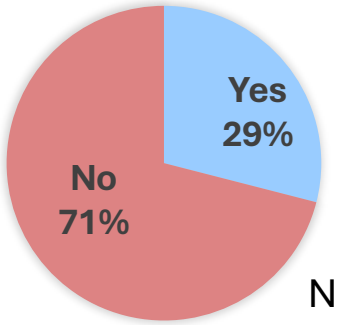
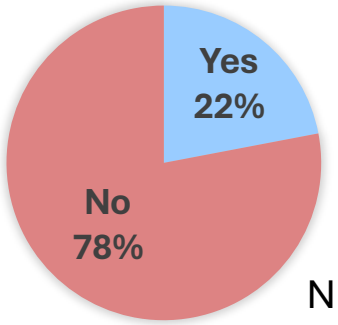
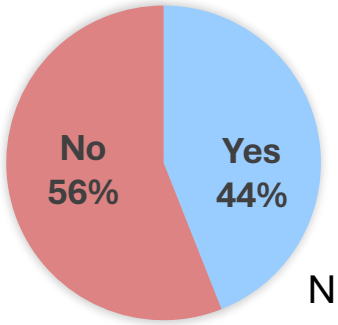


MOG-



Recovery/follow-up phase

Outcomes

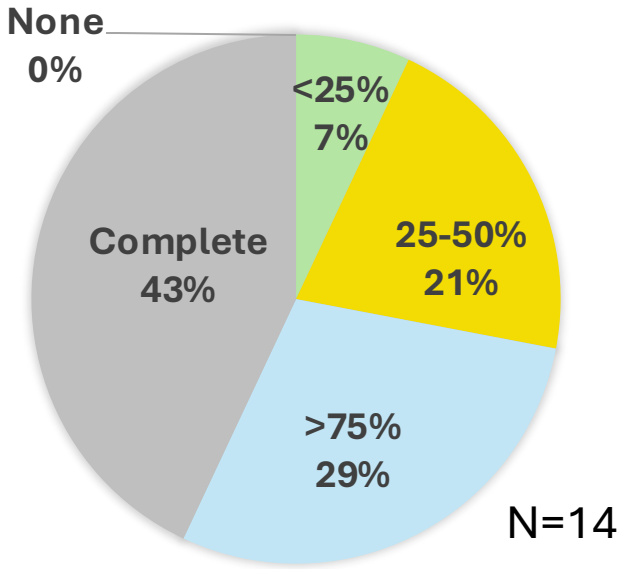
	Length of follow up median (IQR), years	Relapse	Ongoing symptoms affecting function
MOG+	23.0 (8.5-46.5)	 N=21	 N=21
MOG-	12.2 (7.1-54.6)	 N=9	 N=9

MRI recovery

Degree of recovery
No/minimal improvement (<25% improved)
Significant remaining T2 lesions (25-50% improved)
Moderate remaining T2 lesions (50-75% improved)
Few remaining T2 lesions (>75% improved)
Complete resolution

MRI recovery ★

MOG+



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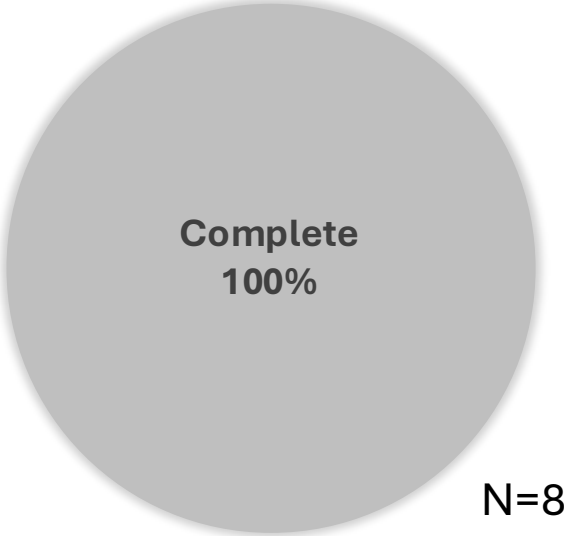
MOG-



N=8

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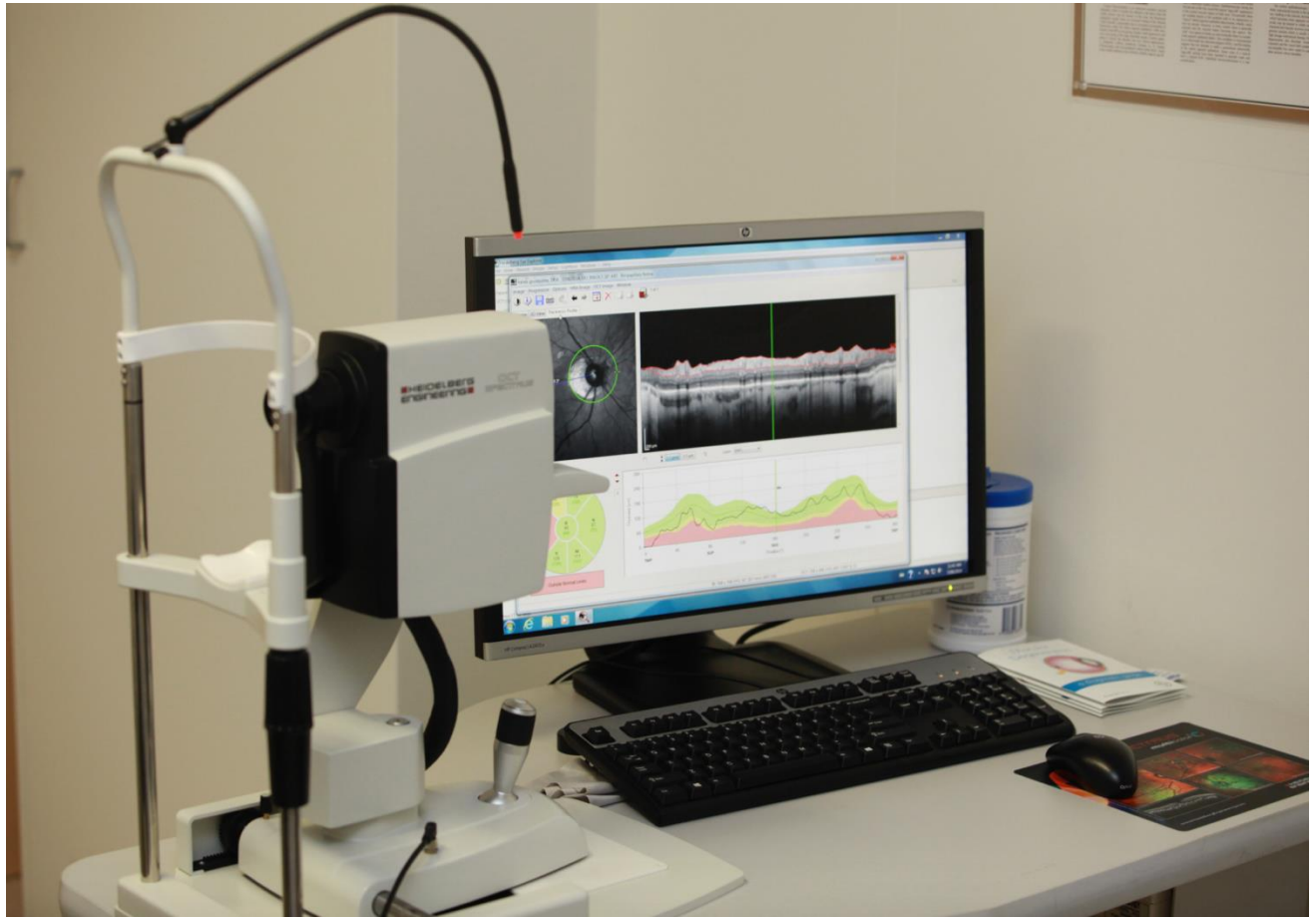
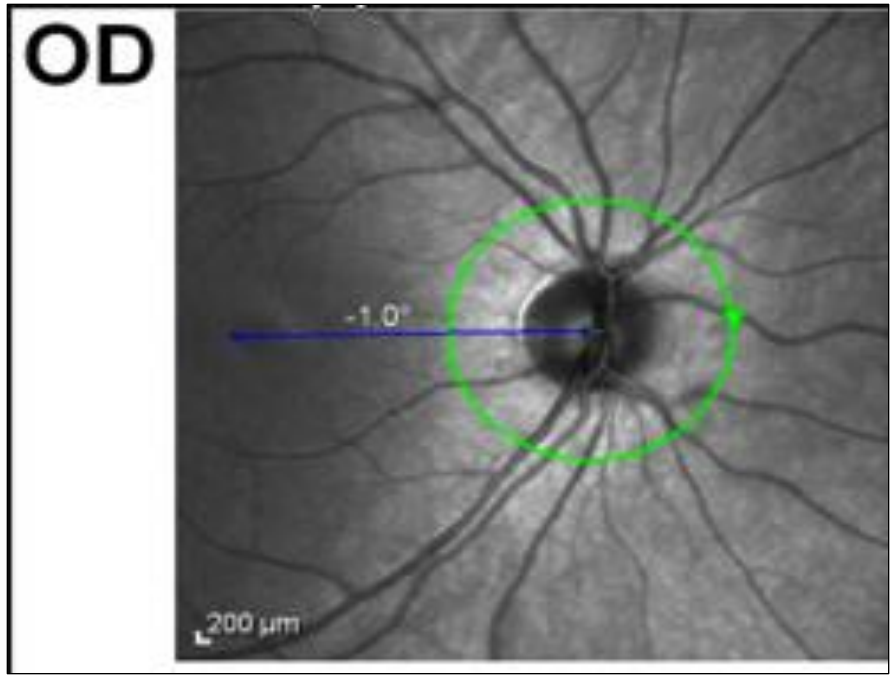
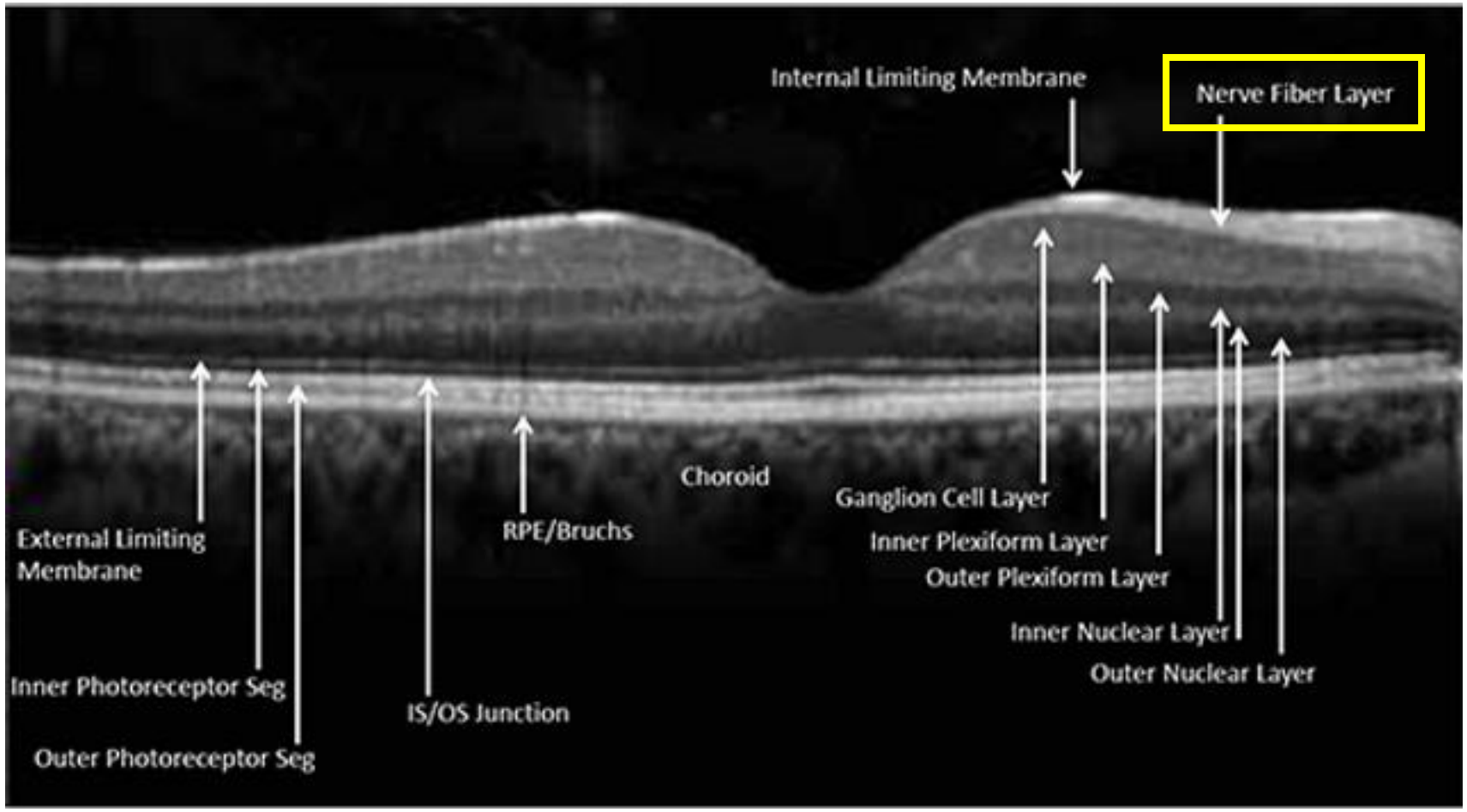
OCT





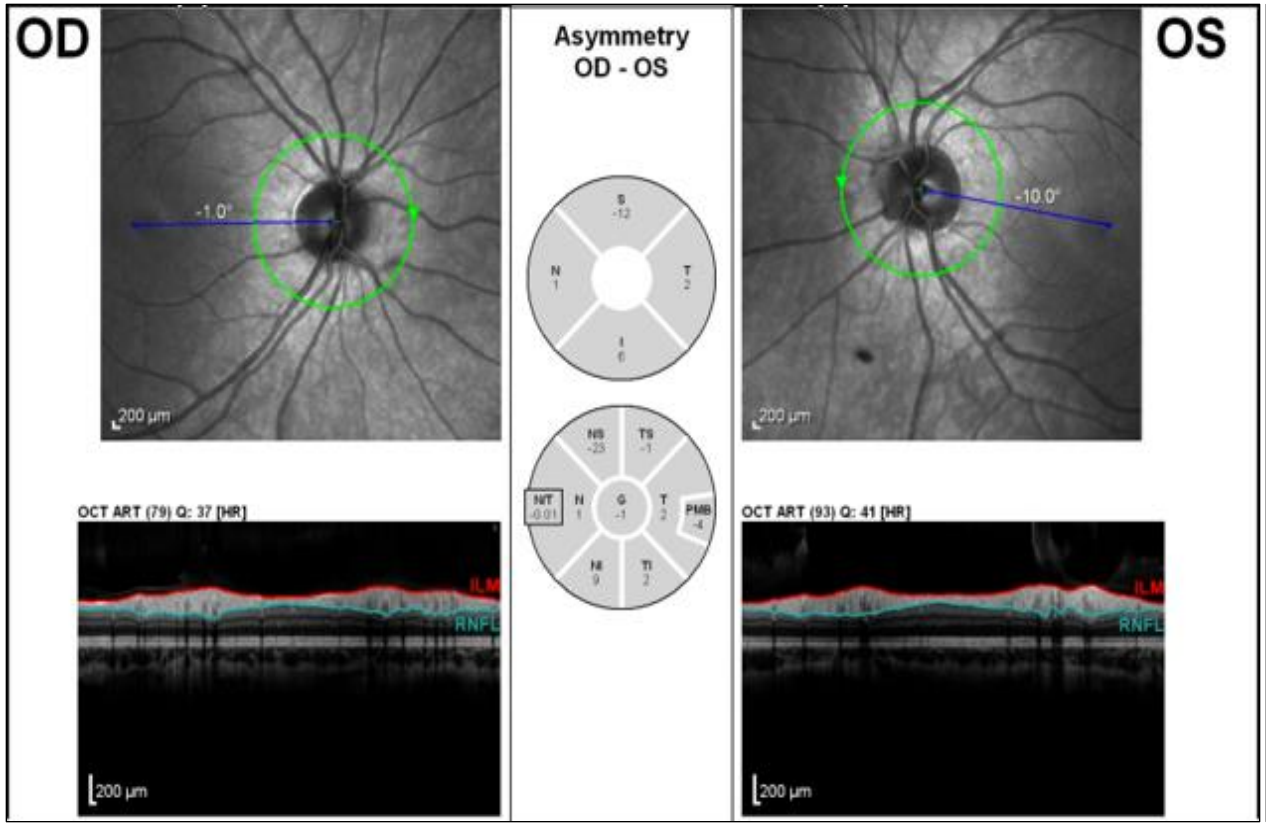
Optical Coherence Tomography (OCT)

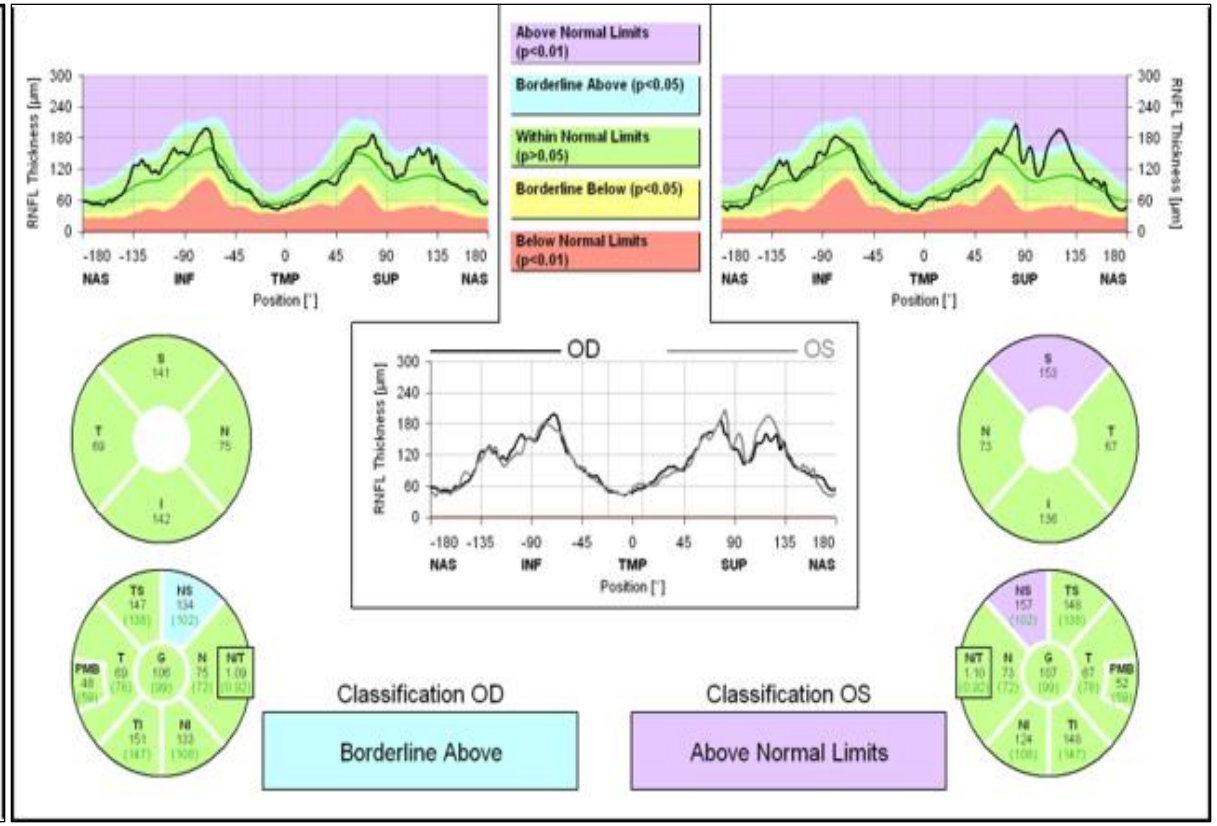
OCT



Retinal layers

Retinal nerve fiber layer thickness

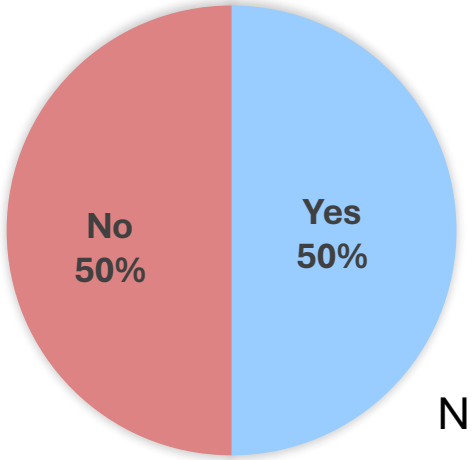




Example report from OCT

OCT with abnormal thinning ★

MOG+



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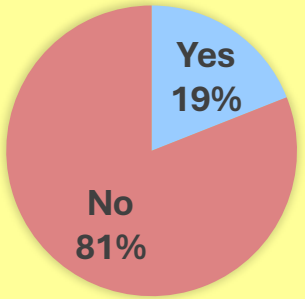
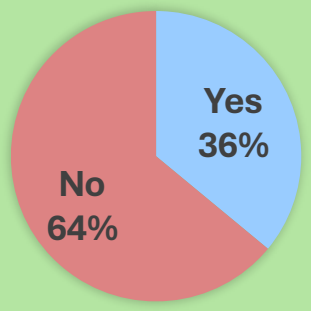
Vision changes



MRI optic nerve involvement



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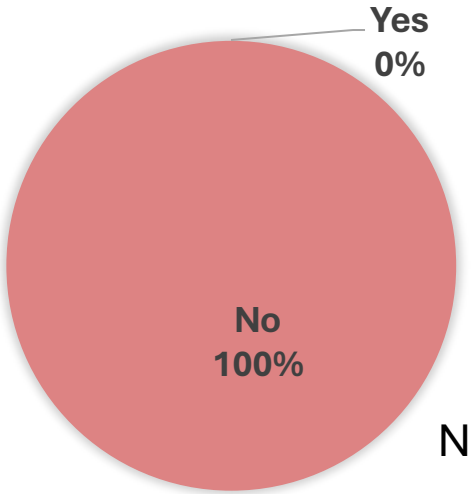

N=4

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Summary

At presentation			
Increased intracranial pressure	MOG+	>	MOG-
Optic nerve involvement	MOG+	>	MOG-
CSF total nucleated cells >5/mm^3	MOG+	>	MOG-
At follow-up			
MRI recovery	MOG+	<	MOG-

Summary

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Increased intracranial pressure	MOG+	>	MOG-
Optic nerve involvement	MOG+	>	MOG-
CSF total nucleated cells >5/mm^3	MOG+	>	MOG-

At follow-up			
MRI recovery	MOG+	<	MOG-
Ongoing symptoms affecting function	MOG+	≤	MOG-

Summary

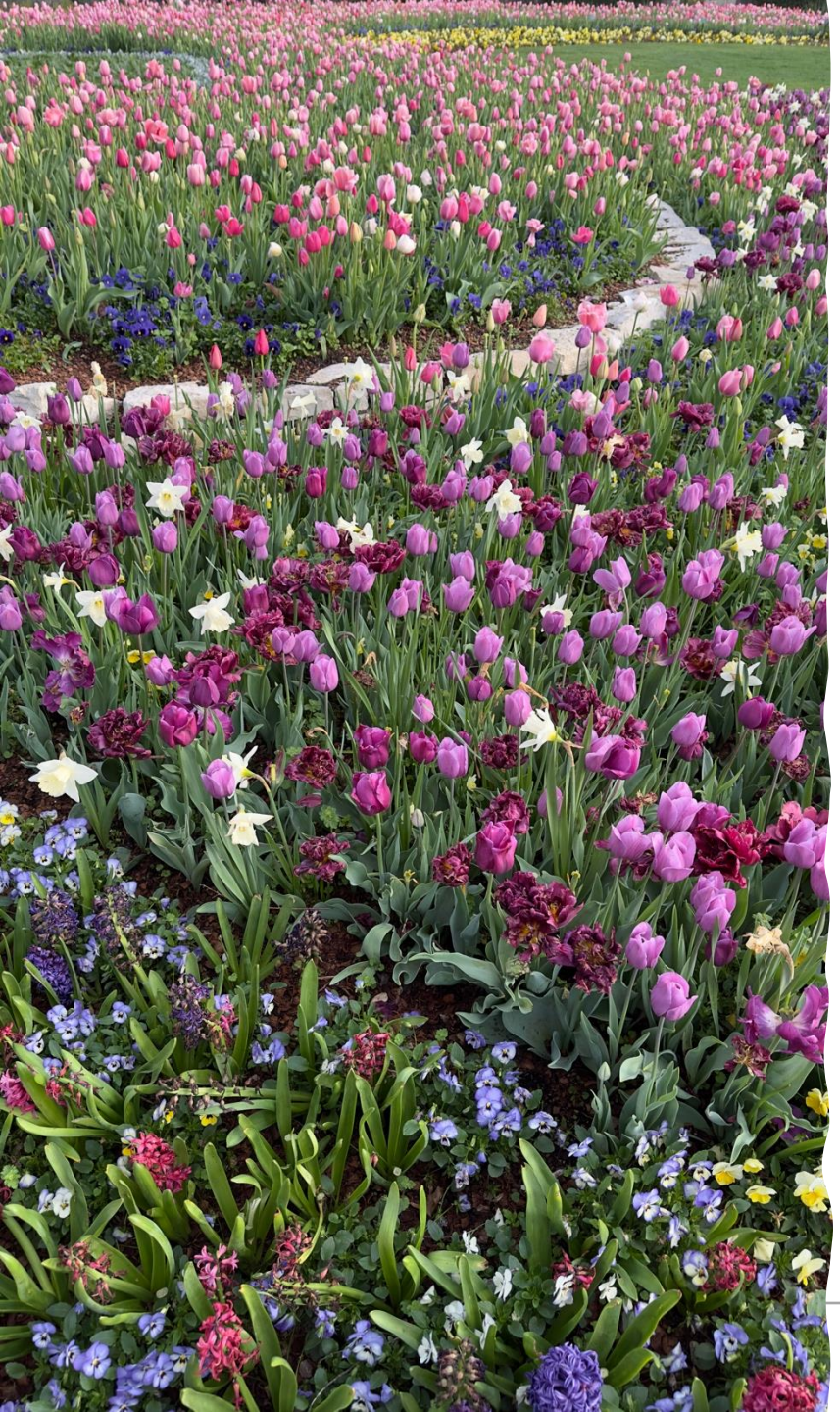
At presentation			
Increased intracranial pressure	MOG+	>	MOG-
Optic nerve involvement	MOG+	>	MOG-
CSF total nucleated cells >5/mm^3	MOG+	>	MOG-

At follow-up			
MRI recovery	MOG+	<	MOG-
Ongoing symptoms affecting function	MOG+	≤	MOG-
Relapse	MOG+	=	MOG-

Summary

At presentation			
Increased intracranial pressure	MOG+	>	MOG-
Optic nerve involvement	MOG+	>	MOG-
CSF total nucleated cells >5/mm^3	MOG+	>	MOG-

At follow-up			
MRI recovery	MOG+	<	MOG-
Ongoing symptoms affecting function	MOG+	≤	MOG-
Relapse	MOG+	=	MOG-
OCT with abnormal thinning	MOG+	>	MOG-



Thank you!

Dr. Benjamin Greenberg
Dr. Cynthia Wang
Dr. Sumit Singh
Darrel Conger


Questions or comments?