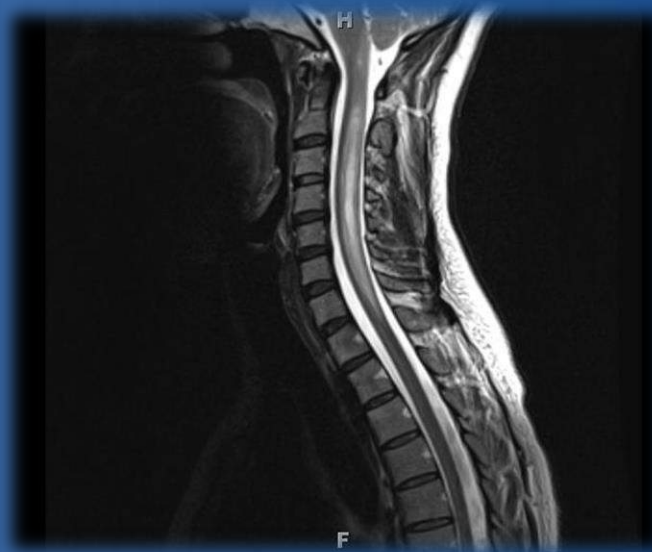




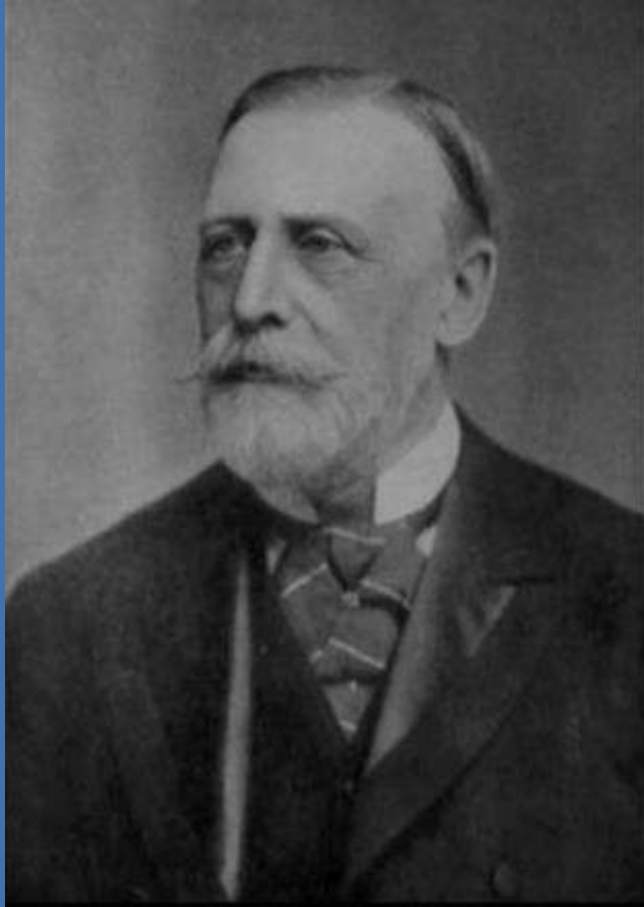
JOHNS HOPKINS
M E D I C I N E

Neuromyelitis Optica 101

Presented by: Maureen A. Mealy, RN, BSN, MSCN
November 11, 2013



Neuromyelitis Optica: *In the Beginning*

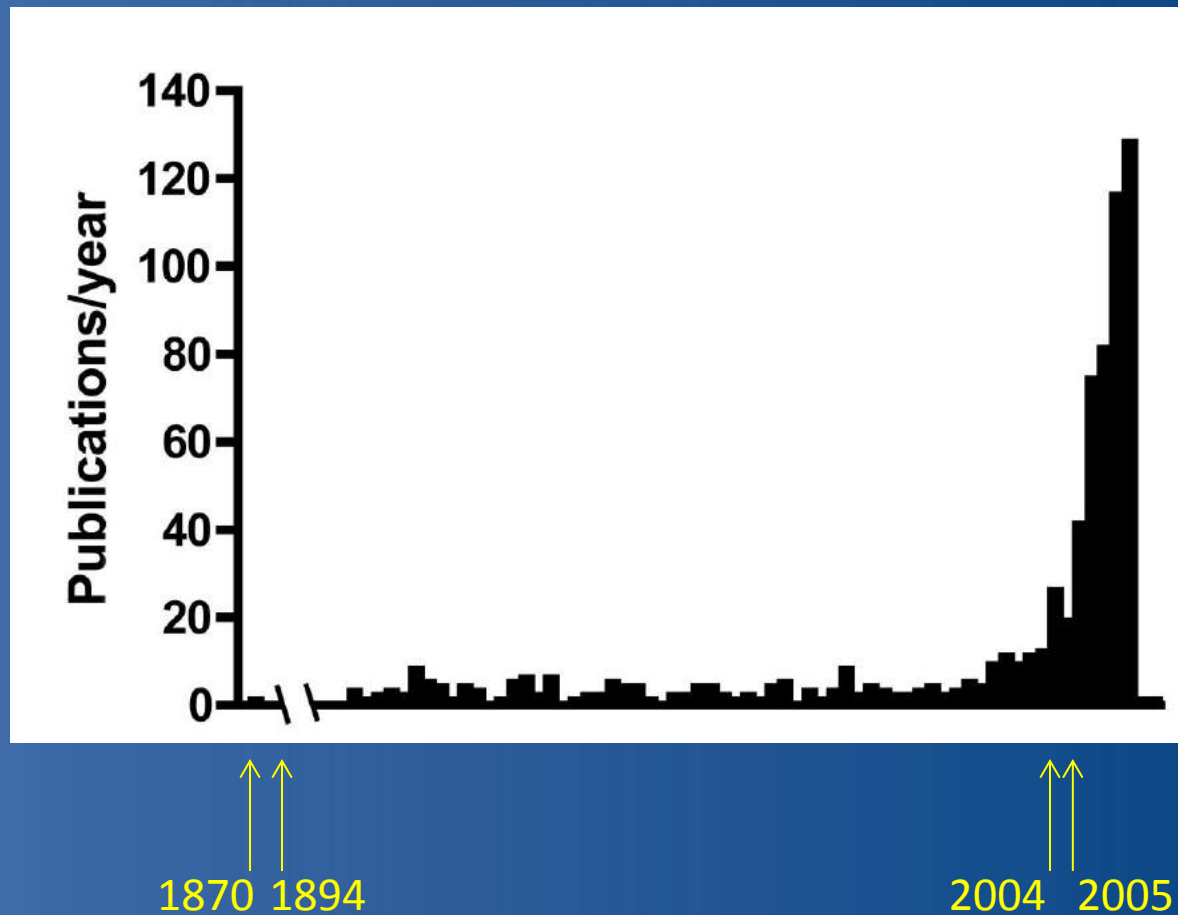


Alfred Allbutt (1836-1925)



Eugene Devic (1858-1930)

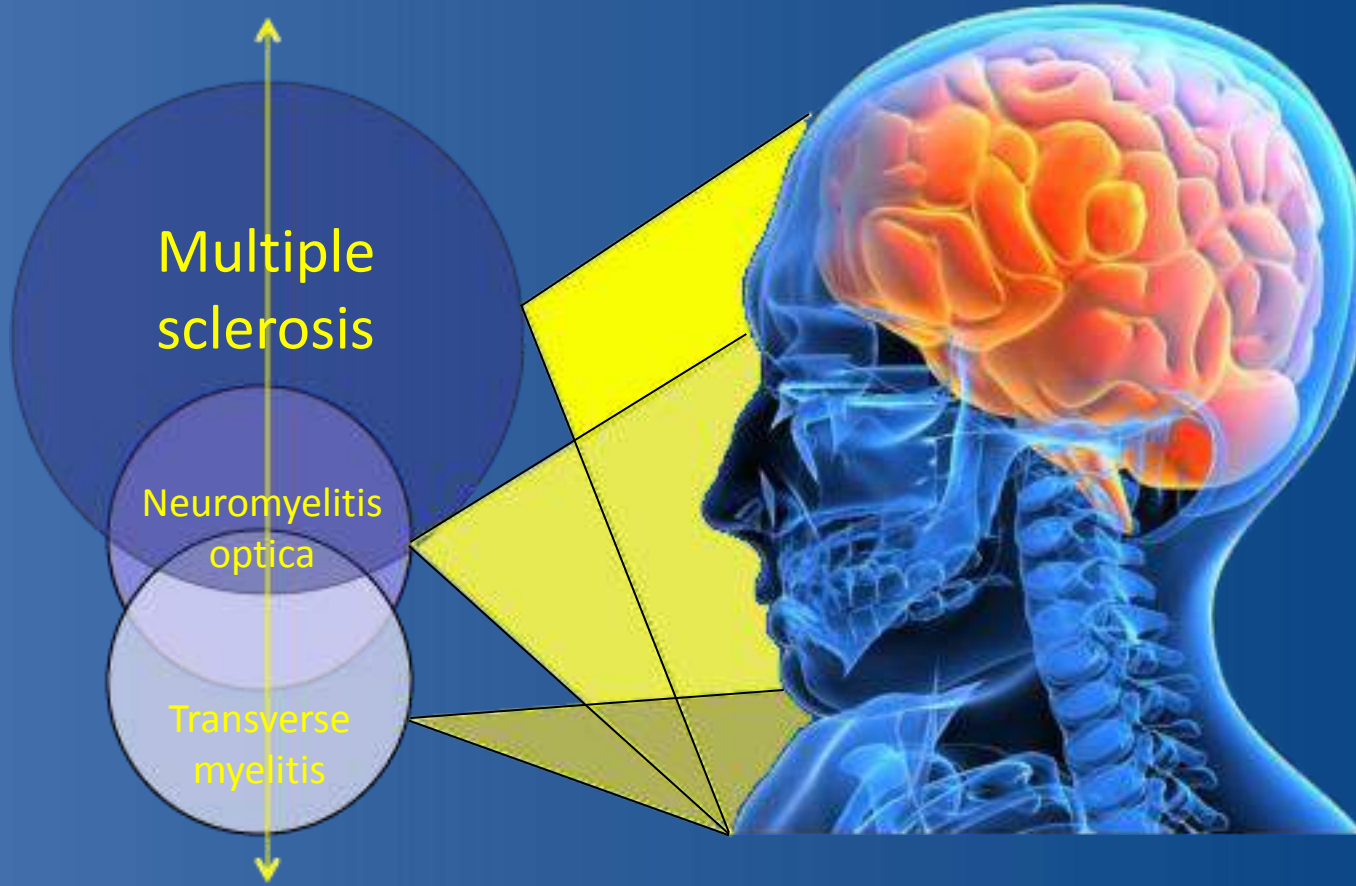
PubMed search: Devic OR neuromyelitis



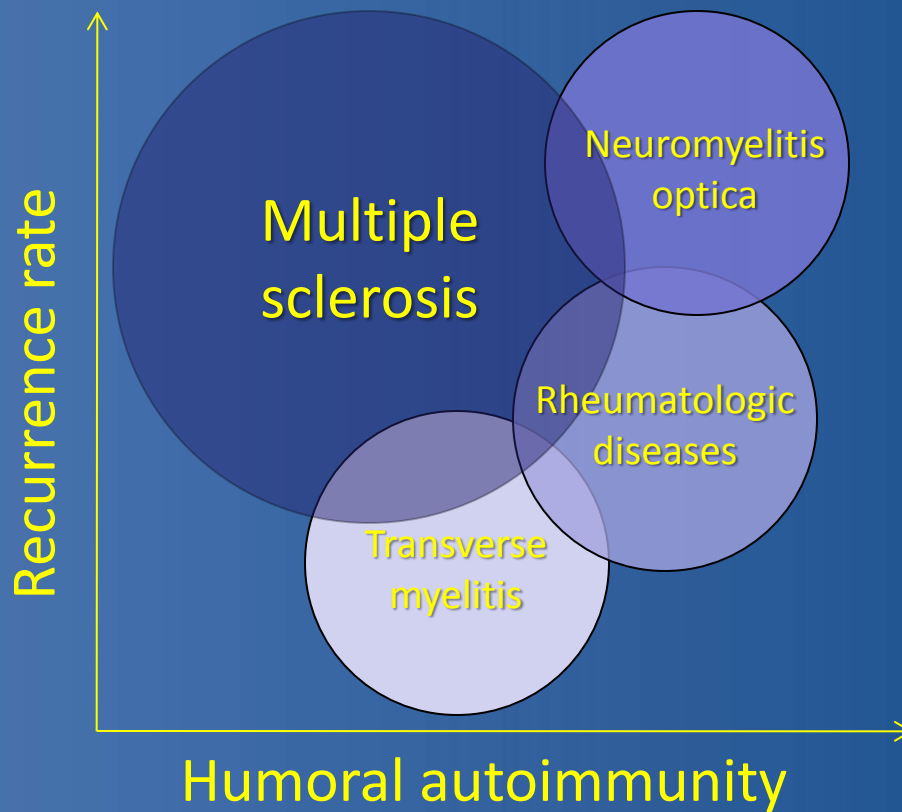
Outline: NMO 101

- Diagnosis
- Epidemiology
- Treatment

What is NMO?



What is NMO?



- Serological factors
- Innate immune system
- Responsive to plasma exchange

Outline: NMO 101

- Diagnosis
- Epidemiology
- Treatment

Definition of NMO: 2006 Revised criteria

1. Optic neuritis
2. Acute myelitis
3. At least 2 of the following 3 supportive criteria:
 - a. Longitudinally extensive spinal cord lesion
 - b. Non-MS (multiple sclerosis) brain MRI
 - c. NMO-IgG seropositive

Definition of NMO: NMO Spectrum

1. Optic neuritis *or* myelitis
2. NMO-IgG seropositive

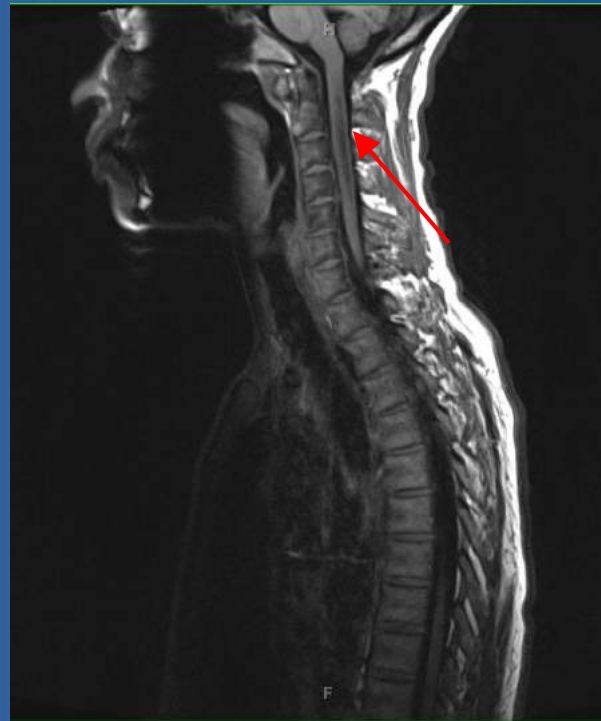
Which NMO-IgG assay?

- Indirect immunofluorescence + immunoprecipitation (Mayo Clinic)
- ELISA (Labcorp/Quest) – false positives
- Cell-based assay (Mayo/Oxford)

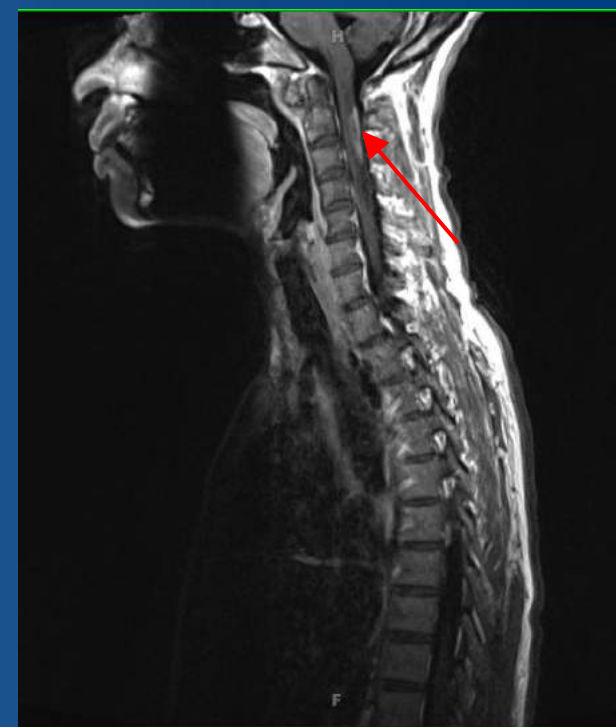
MRI spine in Neuromyelitis optica



T2-weighted



T1-pre-contrast



T1-post-contrast

Diagnostic workup: Make sure it's not MS

Different responses to interferon beta-1b treatment in patients with neuromyelitis optica and multiple sclerosis

A. Uzawa, M. Mori, S. Hayakawa, S. Masuda and S. Kuwabara
Department of Neurology, Graduate School of Medicine, Chiba University, Chiba, Japan

Interferon- β_{1b} Treatment in Neuromyelitis Optica

Masami Tanaka^a Keiko Tanaka^{b,c} Mika Komori^a

^aMS Center, Utano National Hospital, Kyoto, ^bDepartment of Neurology, Brain Research Institute, Niigata University, Niigata, and ^cDepartment of Neurology, Kanazawa Medical University, Uchinada, Japan

Interferon beta-1b exacerbates multiple sclerosis with severe optic nerve and spinal cord demyelination[☆]

Yoko Warabi^{a,b,*}, Yoh Matsumoto^b, Hideaki Hayashi^a

Immunosuppressive therapy is more effective than interferon in neuromyelitis optica

C Papeix¹, J-S Vidal², J de Seze³, C Pierrot-Deseilligny¹, A Tourbah^{1,4}, B Stankoff^{1,5}, C Lebrun⁶, T Moreau⁷, P Vermersch³, B Fontaine¹, O Lyon-Caen¹ and O Gout⁸

Development of extensive brain lesions following interferon beta therapy in relapsing neuromyelitis optica and longitudinally extensive myelitis

Received: 23 June 2007

Received in revised form: 15 August 2007

Accepted: 5 September 2007

Published online: 21 November 2007

Look for clues...

More common in NMO	More common in MS
Longitudinally extensive TM	Small, sensory, white matter TM
Permanent CNS destruction	Recovers quickly
CSF WBC > 50/cc	Oligoclonal bands
Other autoantibodies (e.g., SS-A)	Normal labs
NMO-IgG seropositive	NMO-IgG seronegative
MRI brain “non-symptomatic”	Typical MS pattern

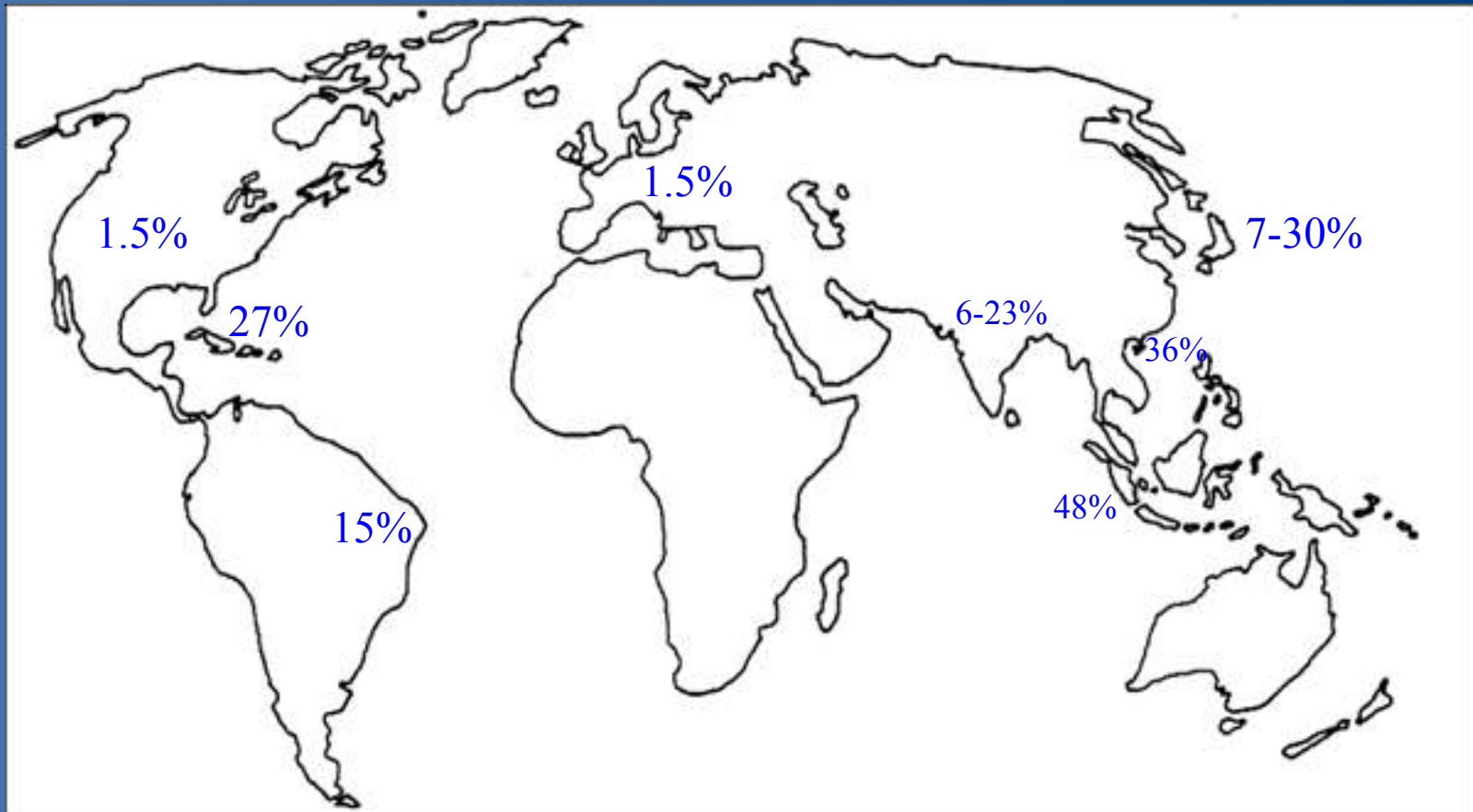
NMO Consortium: MRI brain

Normal brain MRI	Total patients	61 (40%)
	NMO	45
	NMO spectrum	16
Non-specific white matter lesions	Total patients	70 (46%)
	NMO	49
	NMO spectrum	21
Multiple sclerosis-typical	Total patients	19 (13%)
	NMO	14
	NMO spectrum	5
Brainstem lesions	Total patients	30 ¹

Outline: NMO 101

- Diagnosis
- Epidemiology
- Treatment

Epidemiology of NMO: of all demyelinating diseases...



NMO Consortium: Diagnosis, age and sex

Diagnosis	NMO	126 (67%)
	Seropositive NMO	86 (68%)
	Seronegative NMO	40 (32%)
	NMO spectrum	61 (33%)
Age at onset	Mean	41.1 years range 3-81
	Median	40.0 years
Sex	Female	162 (87%)
	Male	25 (13%)
	Ratio (F:M)	6.5:1

NMO Consortium: Race

Race	Caucasian	89 (48%)
	African descent	69 (37%)
	Hispanic	15 (8%)
	Asian	10 (5%)
	Native American	4 (2%)

Outline: Updates

- Diagnosis
- Epidemiology
- Treatment

Types of Treatments for NMO



- Treatment of symptoms
 - Symptomatic relief; palliative
- Treatment of the underlying immune/inflammatory disease process
 - Relapse prevention with immunosuppression
 - Treatment of acute exacerbations

Acute Treatment in NMO

- Currently Available:
 - IV Methylprednisolone
 - Plasma exchange
 - Cyclophosphamide
 - ASIA A
 - systemic auto-immune disease
 - IVIG???

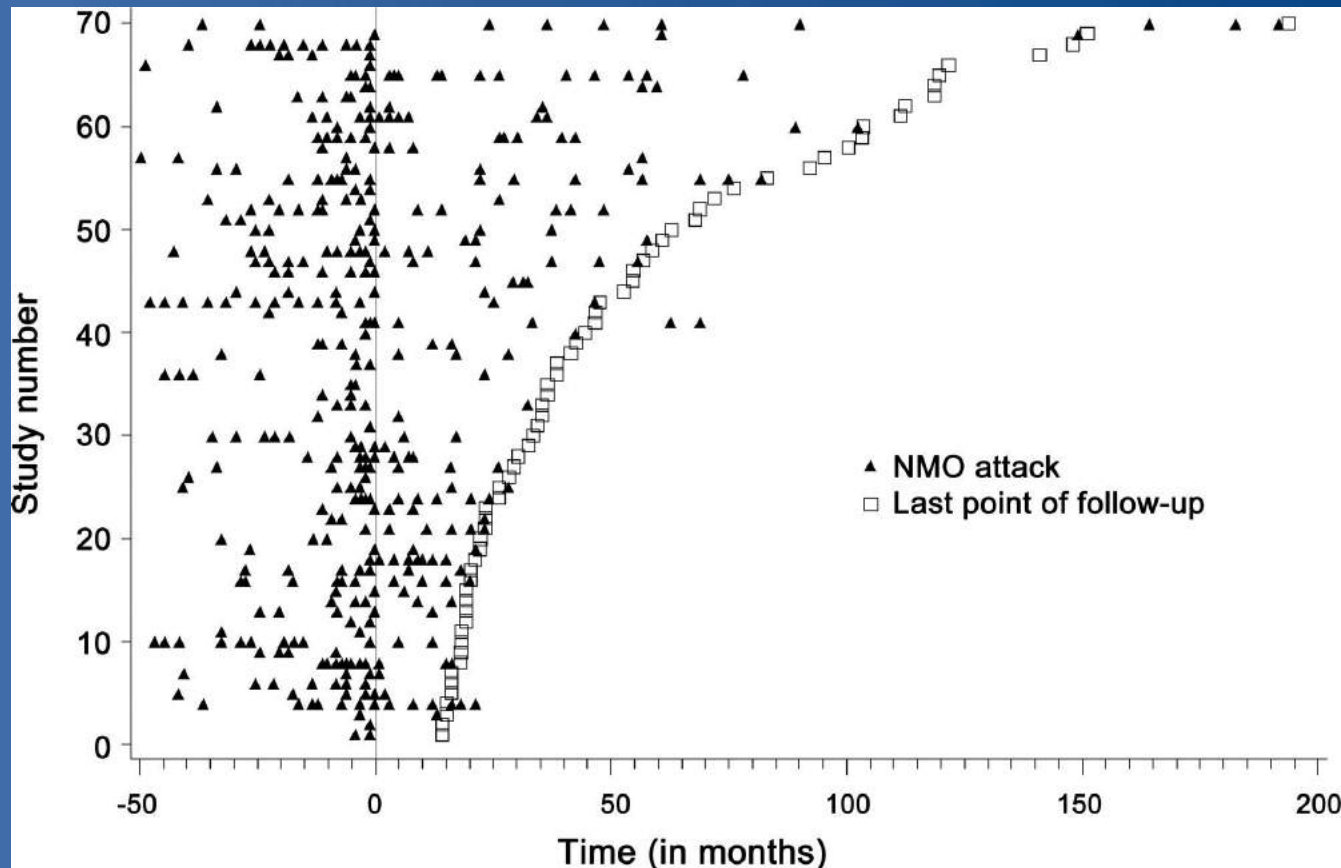
Chronic Immunosuppression in NMO in the US



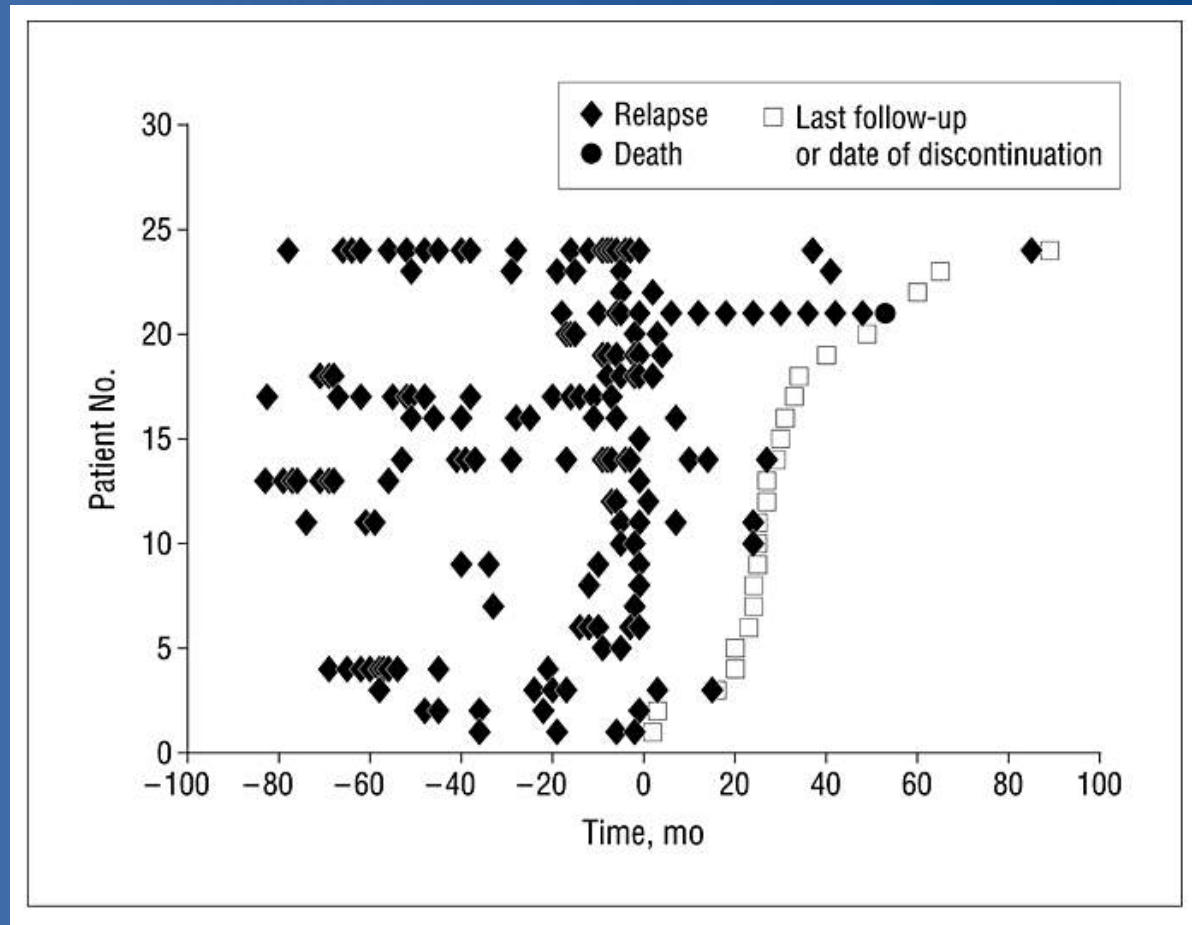
- Currently Available:
 - Azathioprine
 - Mycophenolate mofetil
 - Rituximab

Prednisone, methotrexate, mitozantrone

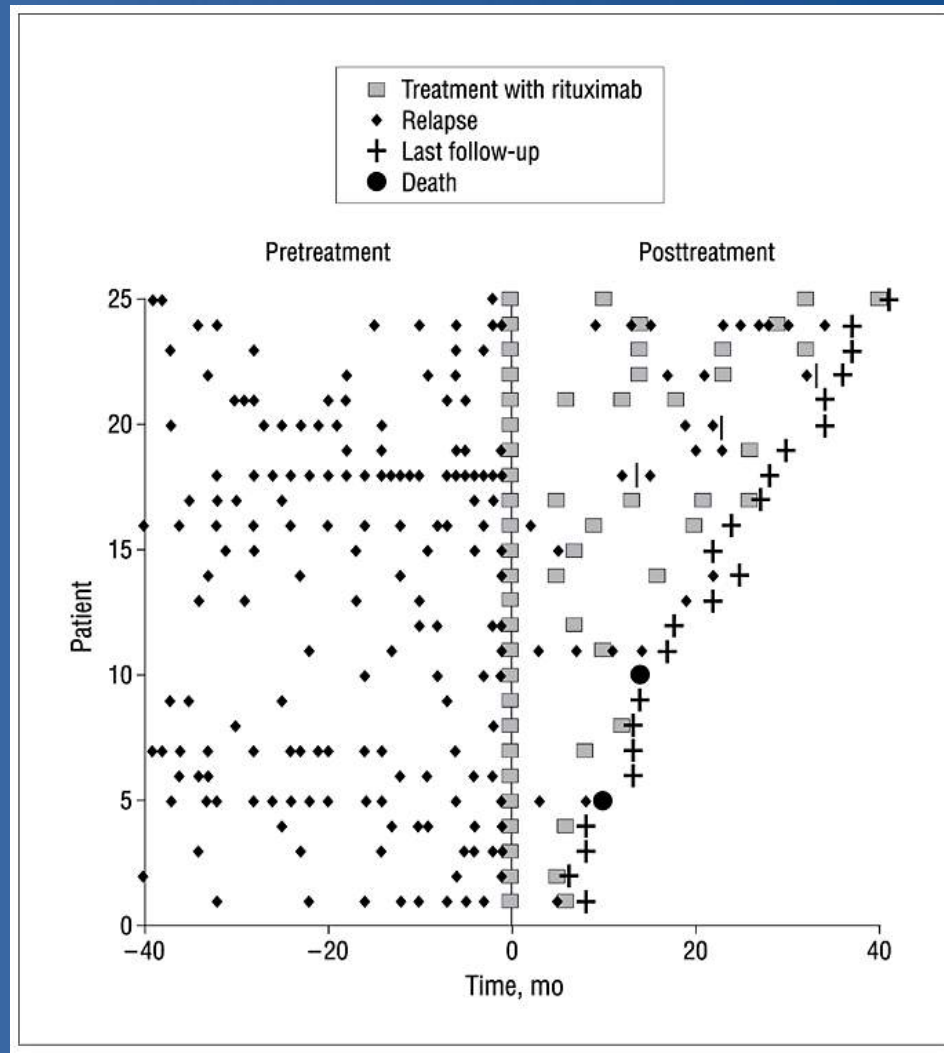
Azathioprine in NMO: retrospective study of 70 patients



CellCept in NMO: retrospective study of 24 patients



Rituximab in NMO: retrospective study of 25 patients



On the Horizon in NMO

- Acute:
 - Human C1 Esterase Inhibitor (Cinryze[®])
 - blocks upstream complement
 - Bevacizumab (Avastin[®])
 - blocks break down of blood brain barrier in neuroinflammation
- Chronic Immunosuppression:
 - Eculizumab
 - blocks downstream complement
 - >4 other agents down the line...

Challenges to Stem Cell Trials

- Cause no harm.
- Access the lesion.
- Prevent stem cell rejection.
- Differentiate in response to local environment.
- Improve neurologic function.

Resources for NMO patients



www.guthyjacksonfoundation.org



<http://www.hopkinsmedicine.org/jhtmc>
e-mail: hopkinsTMcenter@jhmi.edu
www.hopkinsmedicine.org/projectrestore



www.myelitis.org



The Johns Hopkins
PROJECT RESTORE

Restoring hope, function, and lives to MS & TM patients