

# Acute treatments at onset and relapse

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## Relevant Disclosure

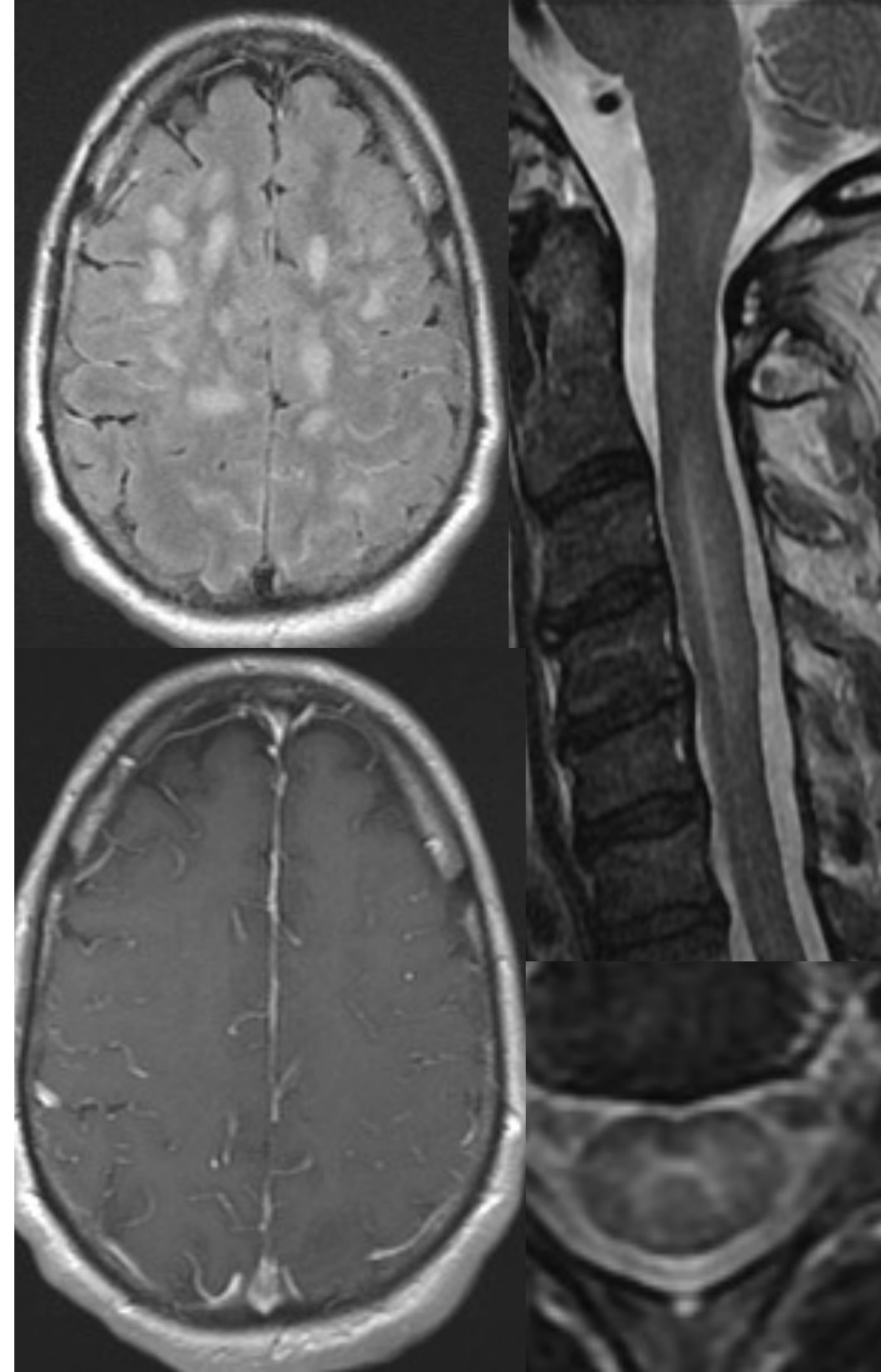
Dr Flanagan has served on advisory boards for Alexion, Genentech and Horizon Therapeutics. He has received speaker honoraria from Pharmacy Times. He received royalties from UpToDate. Dr Flanagan was a site primary investigator in a randomized clinical trial on Inebilizumab in neuromyelitis optica spectrum disorder run by Medimmune/Viela-Bio/Horizon Therapeutics. Dr Flanagan has received funding from the NIH (R01NS113828). Dr Flanagan is a member of the medical advisory board of the MOG project. Dr Flanagan is an editorial board member of the Journal of the Neurological Sciences and Neuroimmunology Reports.

# Learning Objectives

- To highlight the importance of early and effective acute immune treatments in CNS demyelinating diseases
- To highlight the other medication and non-medication treatments utilized

## Case: ADEM

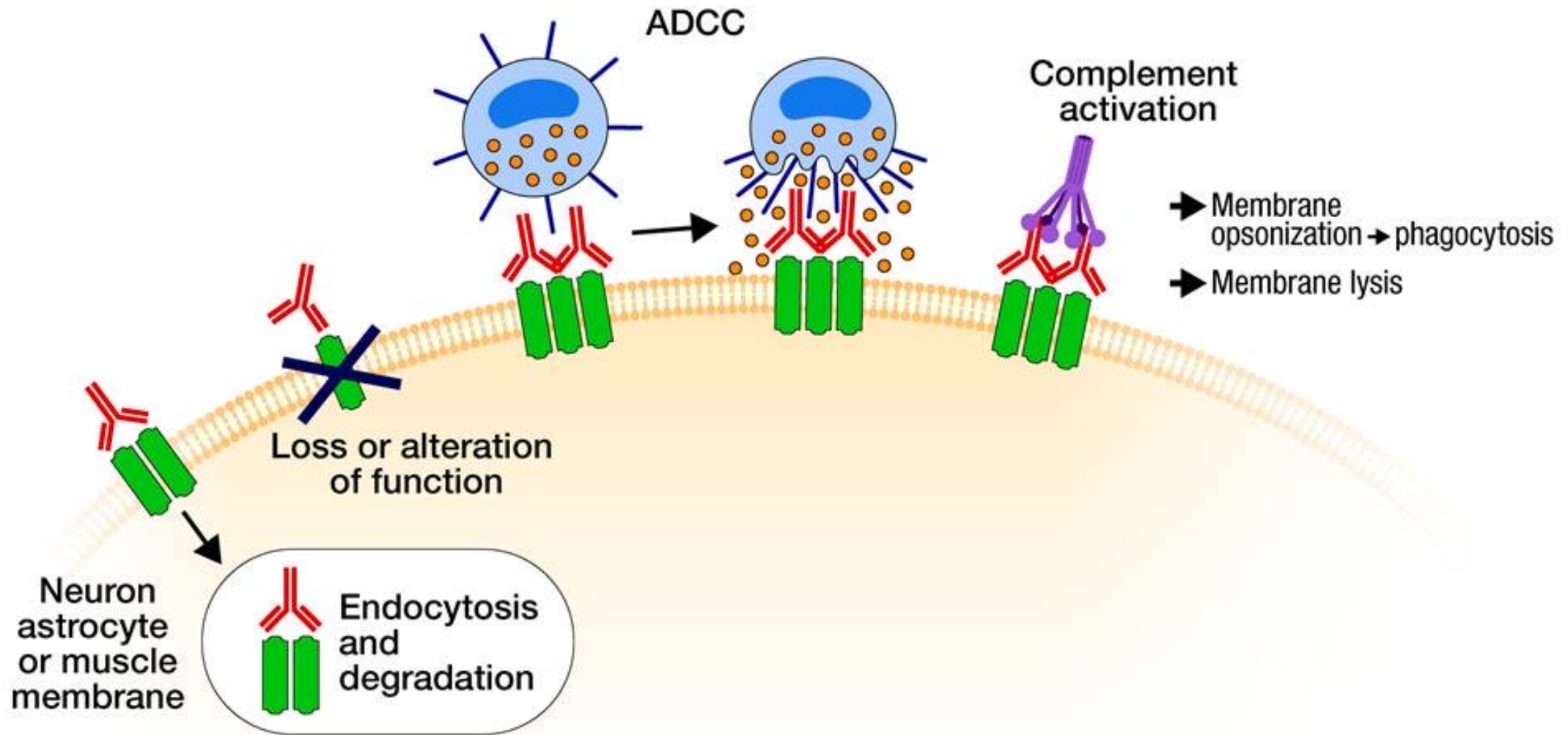
- 47 year old man
- Symptoms:
  - Viral illness
  - Confusion, numbness & weakness
  - Urine difficulty
  - Breathing difficulty
  - Swallow troubles
- Progressed to a coma & paralysis
- Spinal tap & brain biopsy: inflammation



# Case: ADEM

- MOG-IgG antibody positive
- **Diagnosis: MOG-IgG Associated Disease**
- **Treatment:**
  - **Immune treatment**
    - IV steroids & plasma exchange (PLEX)
    - Oral steroids x 2 months followed by a taper
  - **Non-medication**
    - Supportive (breathing tube, feeding tube, catheter, bowel regimen)
    - Physical and occupational therapy during a rehab stay
- **Outcome:** Normal 3 months later

# MOG & Aquaporin-4 antibodies target the cell surface



# Inflammatory Demyelinating Diseases of the CNS: Attack Types and Syndromes

- Acute disseminated encephalomyelitis (ADEM)
- Transverse myelitis (TM)
  - Disease-associated (e.g., acute flaccid myelitis)
  - Idiopathic (no known cause)
- Optic neuritis (ON)
- Others (encephalitis, brainstem syndromes)
- Combinations (e.g., TM & ON: neuromyelitis optica)

# Distinct Inflammatory Demyelinating Diseases of the CNS

1. Multiple sclerosis
2. Aquaporin-4-IgG seropositive neuromyelitis optica spectrum disorder (AQP4+NMOSD)
3. Myelin Oligodendrocyte Glycoprotein-IgG associated disease (MOGAD)



# Immune treatments

# Treatment Principles for Demyelinating Attacks

- The earlier the better
- Start with high dose steroids
- Low threshold for additional treatments if still very disabled or concurrently if a severe episode

# High Dose Steroids 1<sup>st</sup> line for Demyelinating Attacks

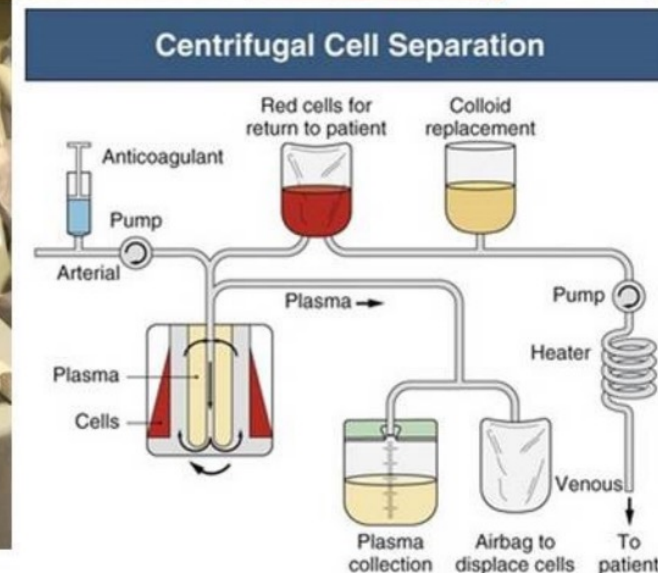
- High dose IV methylprednisolone daily (Solu-medrol) x 3-5 days
- Alternative: High dose oral prednisone daily x 5 days
  - Caveat (25 x 50 mg tablets once a day!!)

# Oral Steroids as a transitional treatment

- Additional oral steroids:
  - MOGAD: for a few months after attacks prevent a relapse
  - AQP4+NMOSD: awaiting attack-prevention treatment to work
- Side effects
  - Insomnia, confusion/psychosis
  - Blood sugar and blood pressure problems
  - Weight gain and puffiness

# Plasma Exchange, Plasmapheresis (PLEX)

- If still problems after steroids
- Very useful for AQP4+NMOSD attacks
- Side effects
  - Blood clots
  - Complications during IV line placement in neck
  - Cramps



Osmon et al. *Practical Neurol* 2020  
Kleiter et al. *Ann Neurol* 2016  
Weinshenker et al. *Ann Neurol* 1999

# Intravenous Immune Globulin (IVIg)

- 0.4 g/Kg once daily for 3-5 days (sometimes given over 2 days)
- Less commonly used acutely and more often in children (e.g, MOGAD)
- More used as maintenance attack-prevention treatment in MOGAD
- Side effects
  - Allergic reaction - check IgA before using
  - Headache – slowing infusion can help
  - Hard on kidneys – formulations without sucros
  - Blood clots

# Acute flaccid myelitis

- Treatment mostly supportive
- Immunosuppressants: none proven
  - Sometimes IVIg used
  - Steroids and plasma exchange more controversial

## Other Non-immune medications

- Neuropathic pain
- Tonic spasms
- Nausea, vomiting, hiccups



# Supportive treatments: Be your own advocate

- Breathing care
- Management of swallow
- Bladder care
- Bowel care
- Rehab for muscles: Physical & occupational therapy
- Occupational therapy
- Management and Prevention of complications

RESEARCH ARTICLE OPEN ACCESS

## CNS Demyelinating Attacks Requiring Ventilatory Support With Myelin Oligodendrocyte Glycoprotein or Aquaporin-4 Antibodies

Hannah H. Zhao-Fleming, MD, PhD, Cristina Valencia Sanchez, MD, PhD, Elia Sechi, MD, Jery Inbarasu, MD, Eelco F. Wijdicks, MD, PhD, Sean J. Pittock, MD, John J. Chen, MD, PhD, Dean M. Wingerchuk, MD, Brian G. Weinschenker, MD, Sebastian Lopez-Chiriboga, MD, Divyanshu Dubey, MBBS, Jan-Mendelt Tillemans, MD, Michel Toledano, MD, Hemang Yadav, MD, and Eoin P. Flanagan, MD

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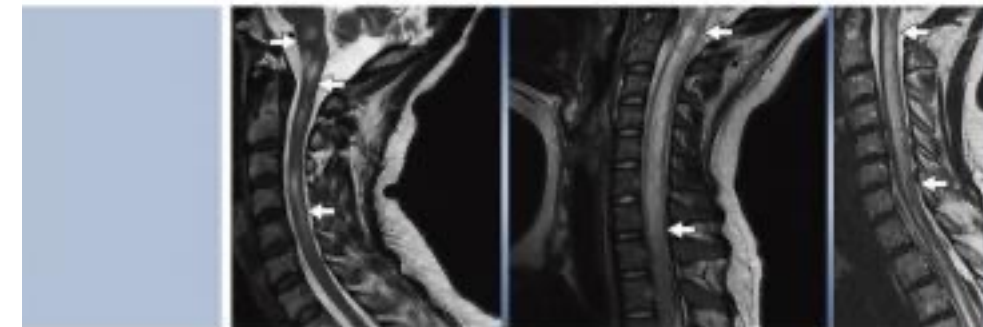
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# Conclusions

- Treat early
- If steroids don't work look for additional treatments (e.g., PLEX)
- Remember other treatments non-medication approaches and be your own advocate for this

**Thank you**