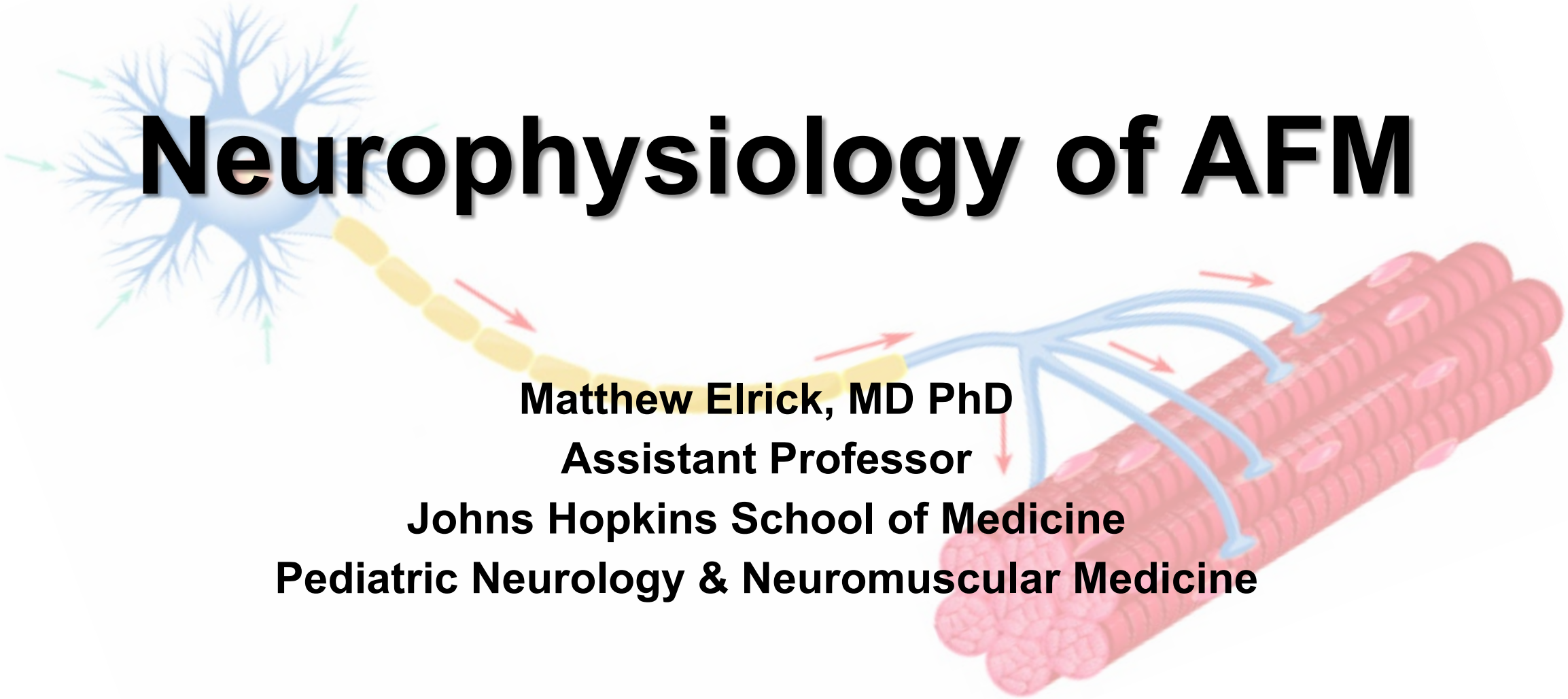
Neurophysiology of AFM



Matthew Elrick, MD PhD
Assistant Professor
Johns Hopkins School of Medicine
Pediatric Neurology & Neuromuscular Medicine

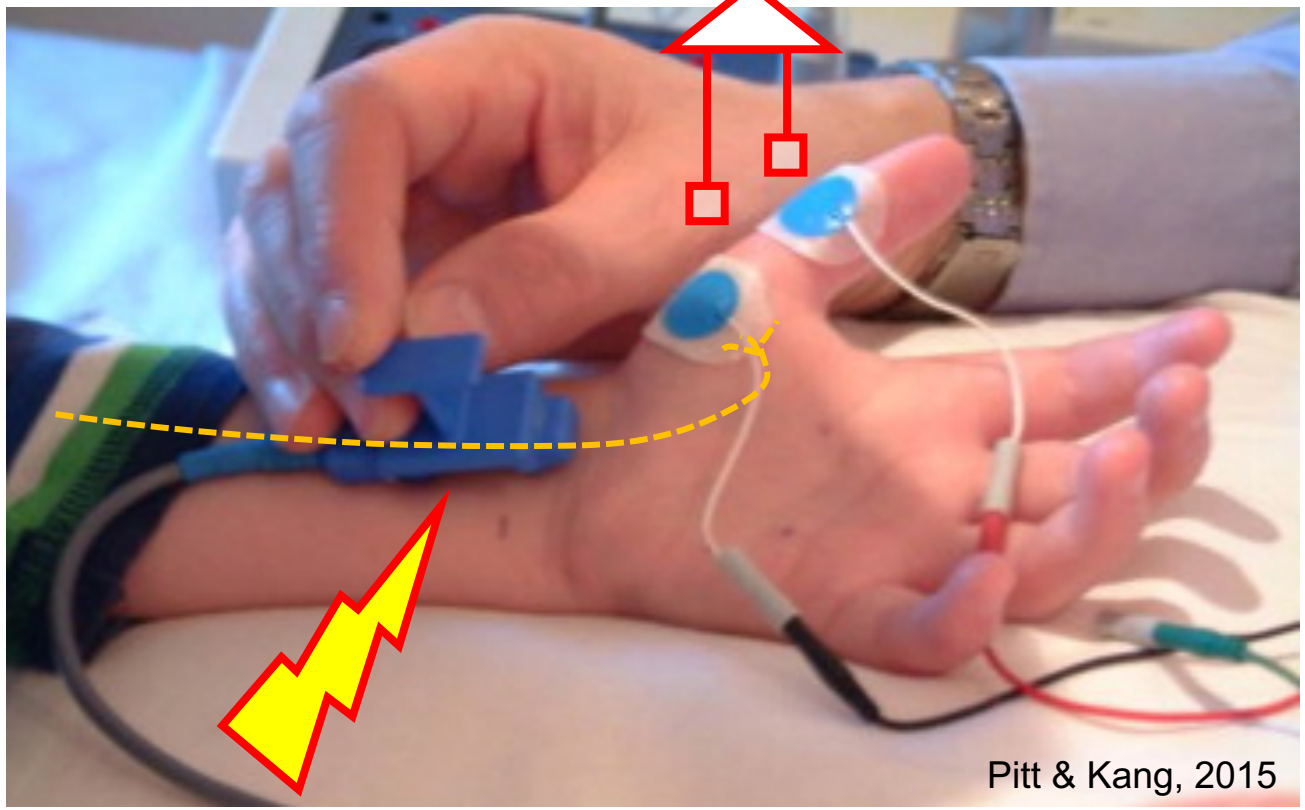
JOHNS HOPKINS
CHILDREN'S CENTER

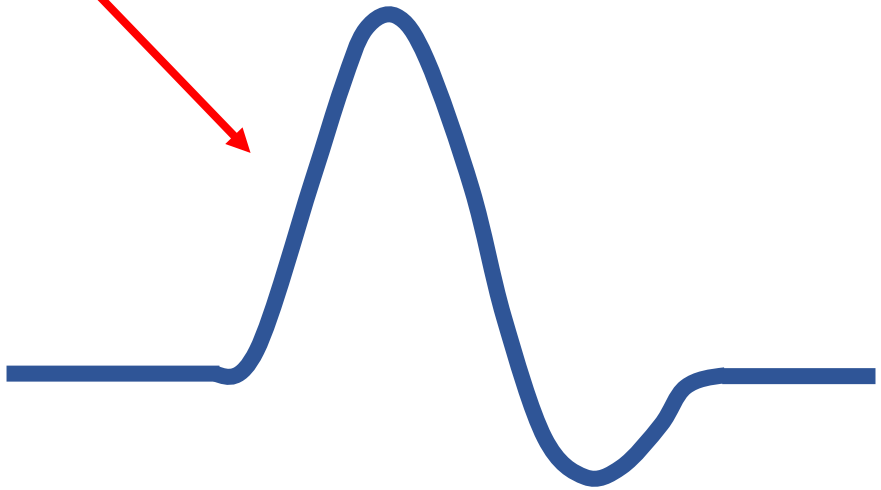
Outline

- Introduction to Electromyography and Nerve Conduction Studies (EMG)
- EMG findings in AFM
- Challenges for EMG in children with AFM
- Uses for EMG in AFM and what it can teach us

Nerve Conduction Studies

Record



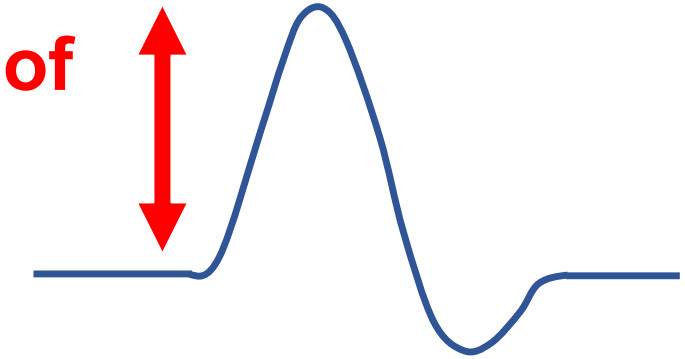


Stimulate

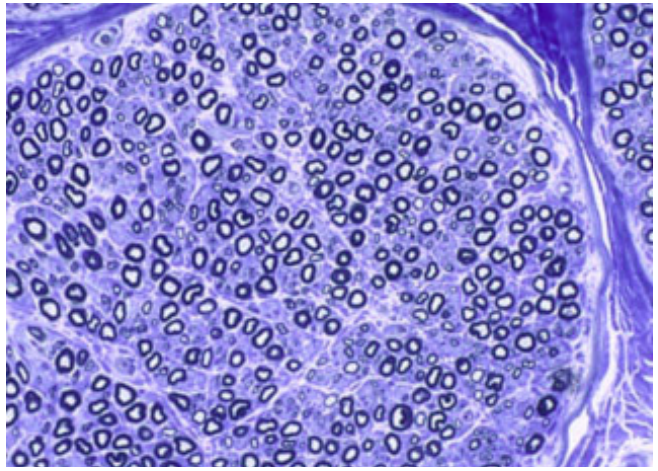
Nerve Conduction Studies

Amplitude =

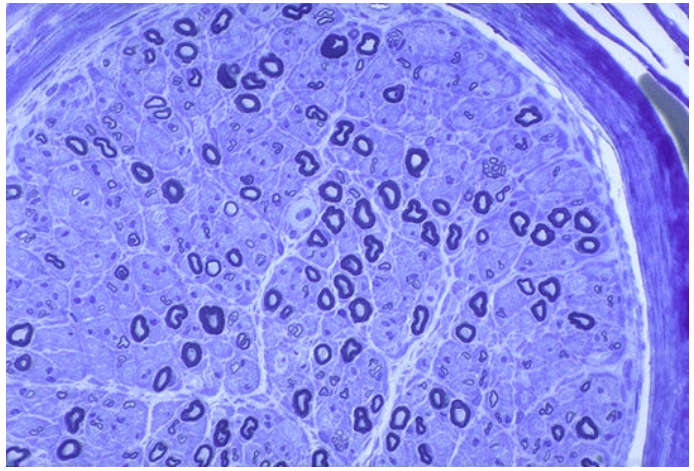
number of axons



Normal

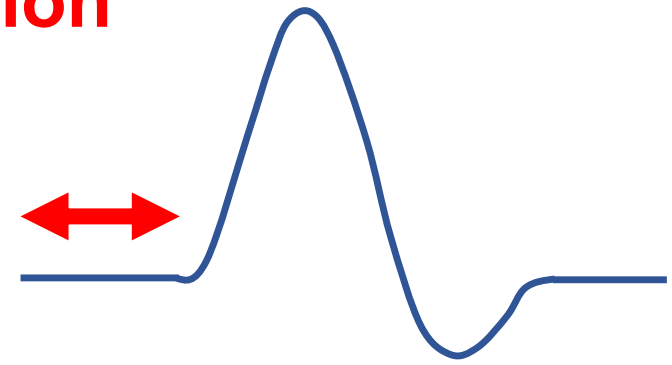


Axon loss



Velocity =

myelination



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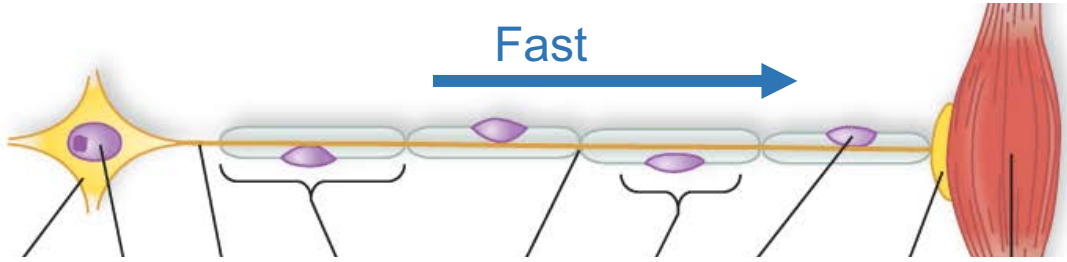
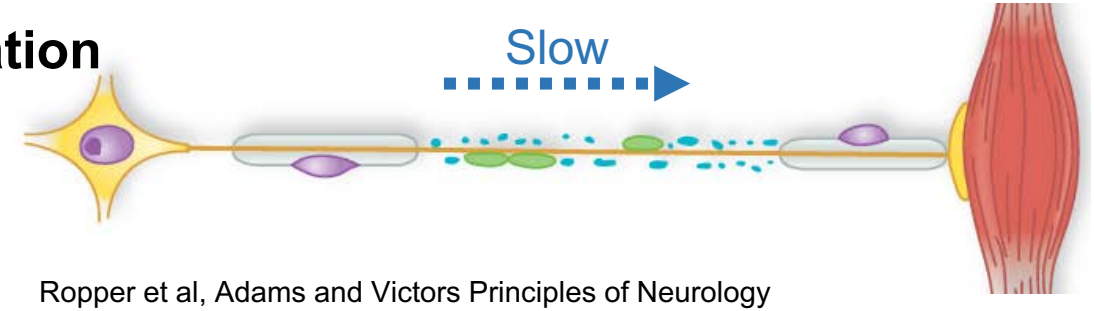
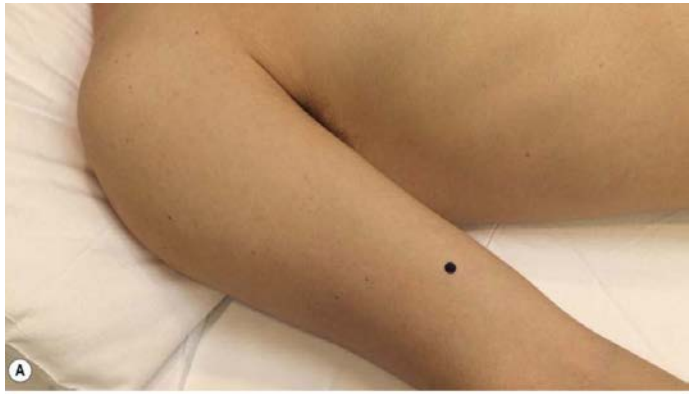


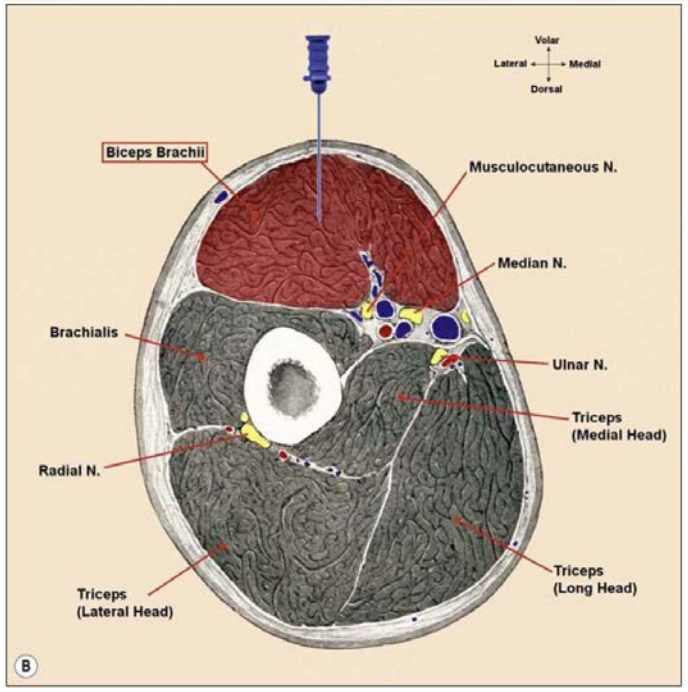
Demyelination



Ropper et al, Adams and Victors Principles of Neurology

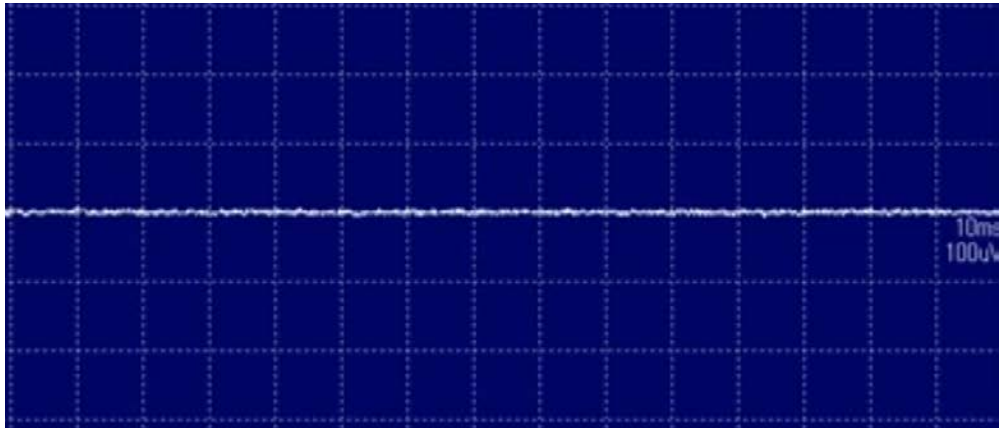
Electromyography (EMG)



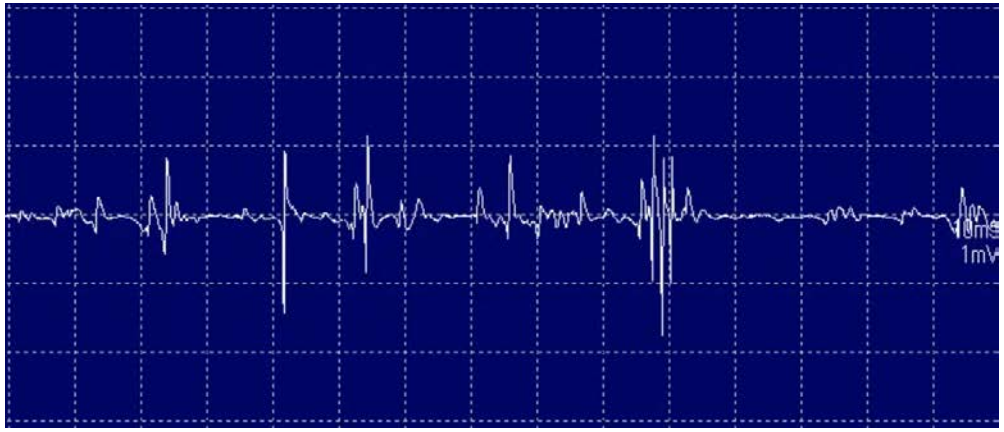


Preston and Shapiro

Rest

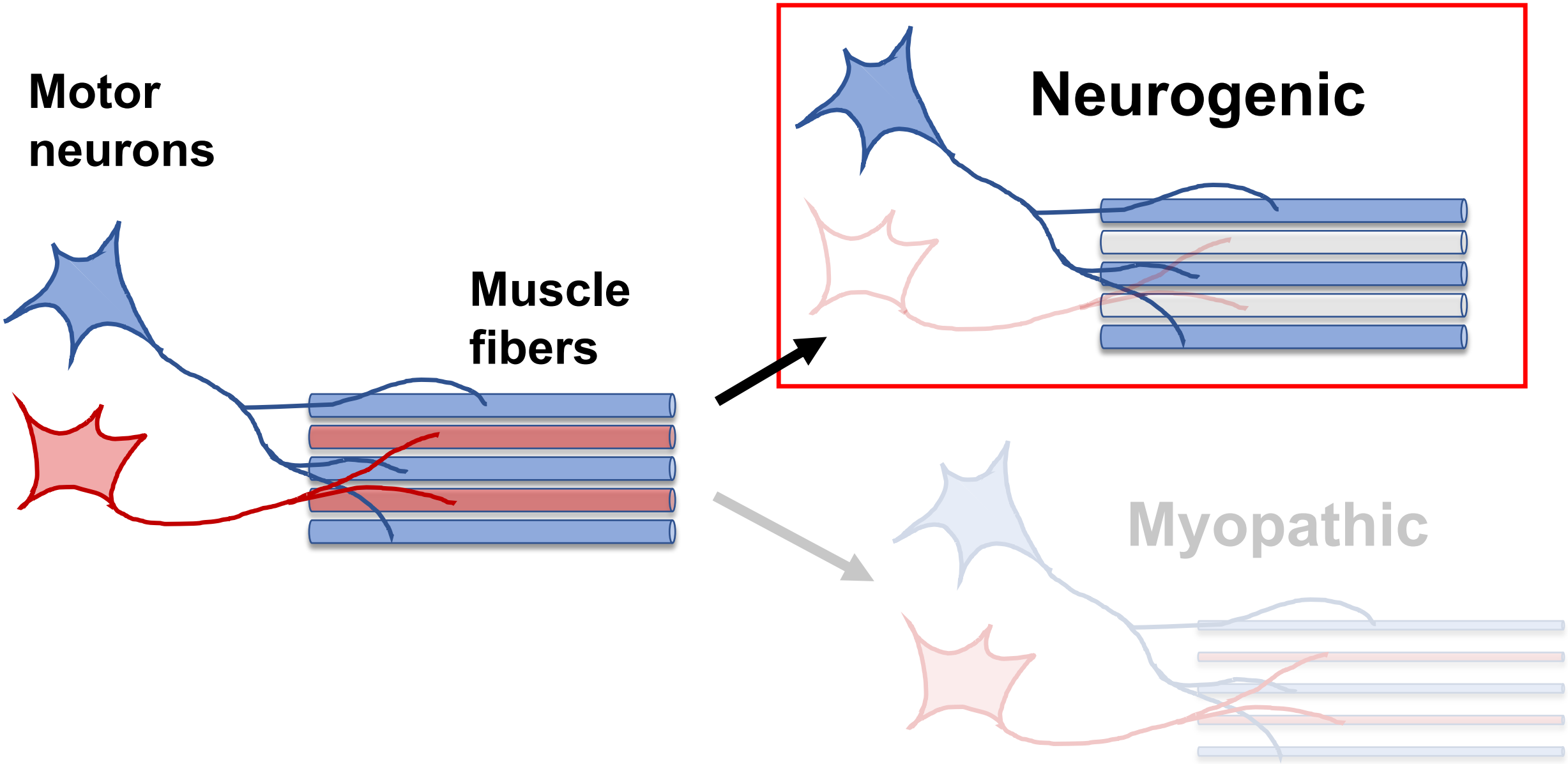


Activation

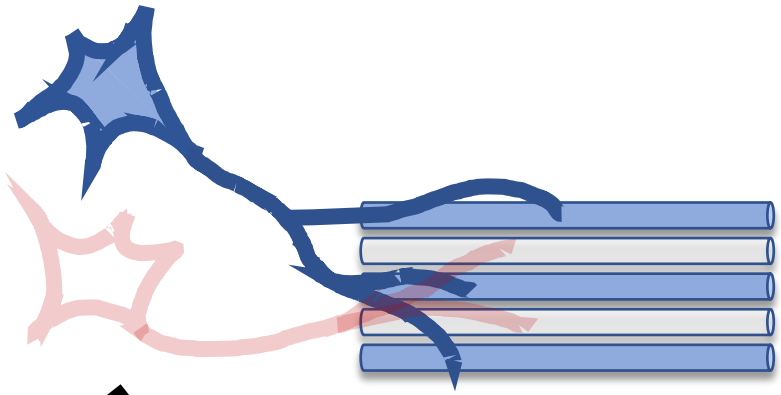


Kimura & Kohara, youtube.com

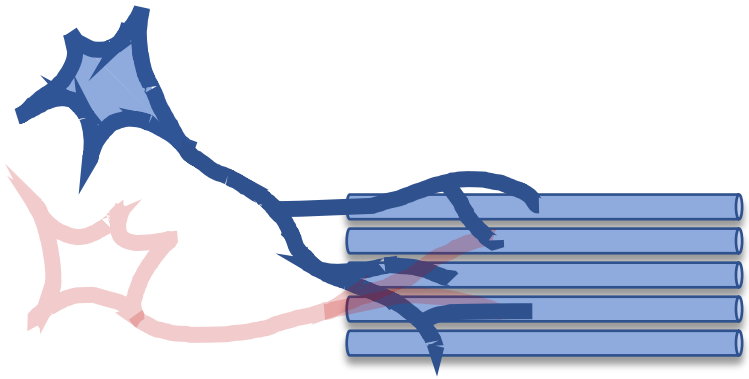
EMG: Patterns of Injury



EMG: Recovery from Neurogenic Injury

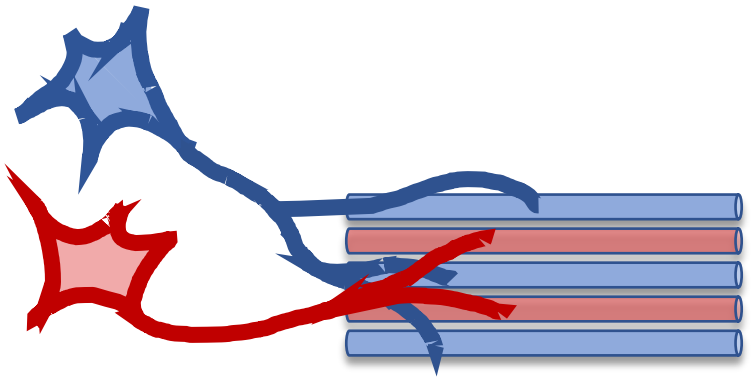


Collateral sprouting



Regeneration

Reversal



EMG/NCS Findings in AFM

Finding	Significance
NERVE CONDUCTION STUDIES	
Compound Muscle Action Potential (CMAP) Amplitude Decreased	Motor neuron / axon loss
F wave a	
EMG: ACUT	
Fibrillatic	
Reduced Recruitment of motor units	Neurogenic / Motor neuron
EMG: CHRONIC	
Motor Unit Potentials high amplitude, long duration	Collateral sprouting and reinnervation

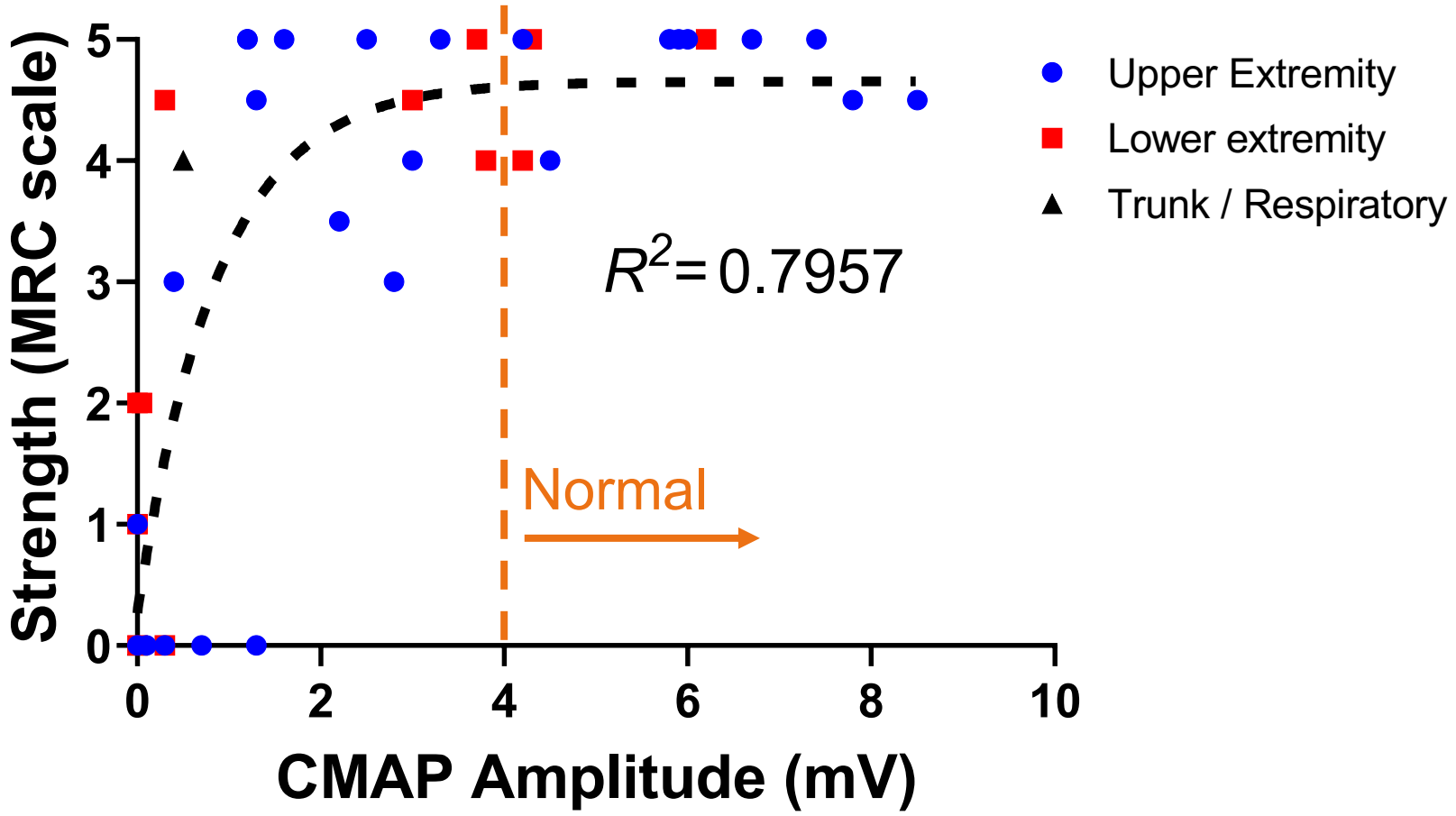
MOTOR NEURON INJURY

1. Van Haren et al, *JAMA*, 2015
2. Hovden et al, *Muscle Nerve*, 2015
3. Messacar et al, *Ann Neurol*, 2016
4. Martin et al, *Neurology*, 2017
5. Andersen et al, *Eur J Neurol*, 2017
6. Ruggieri et al, *Eur J Paediatr Neurol*, 2017
7. Elrick et al, *JAMA Pediatr*, 2018
8. Saltzman et al, *Pediatr Neurol*, 2018
9. Knoester et al, *Pediatr Infect Dis J*, 2018
10. Edmiston et al, *Spinal Cord Ser Cases*, 2019
11. Natera-de Benito et al, *J Clin Neuromuscul Dis*, 2018
12. Sarmast et al, *J Trop Pediatr*, 2019
13. Aubart et al, *Front Neurol*, 2020

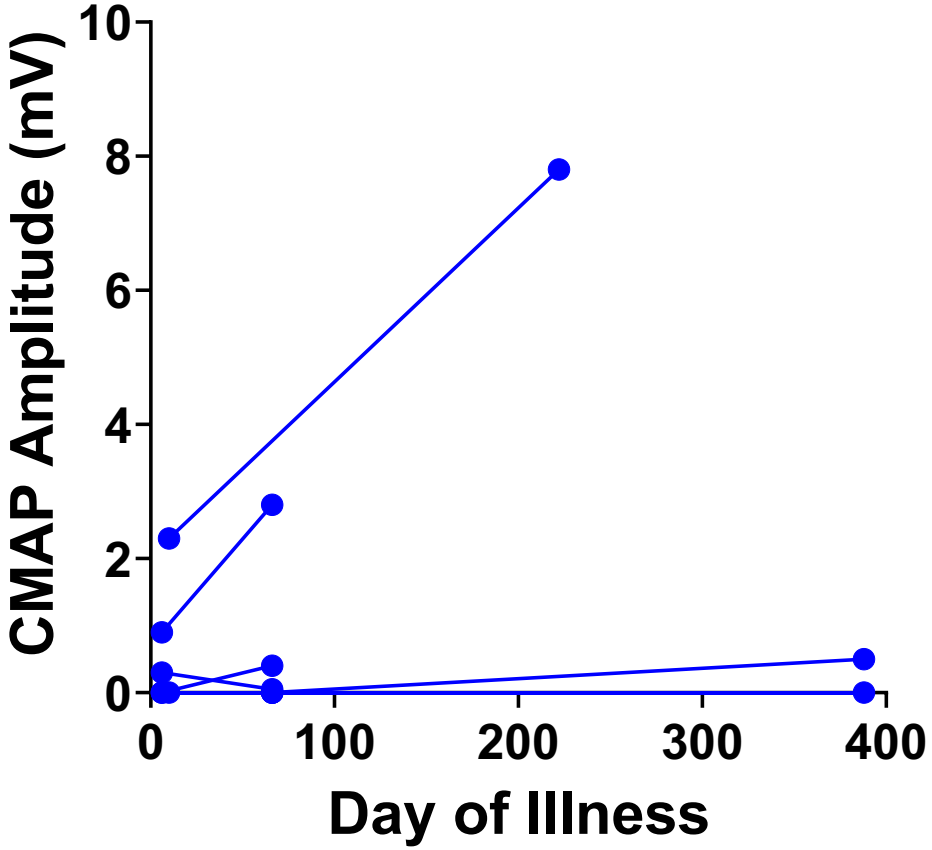
Johns Hopkins EMG Data

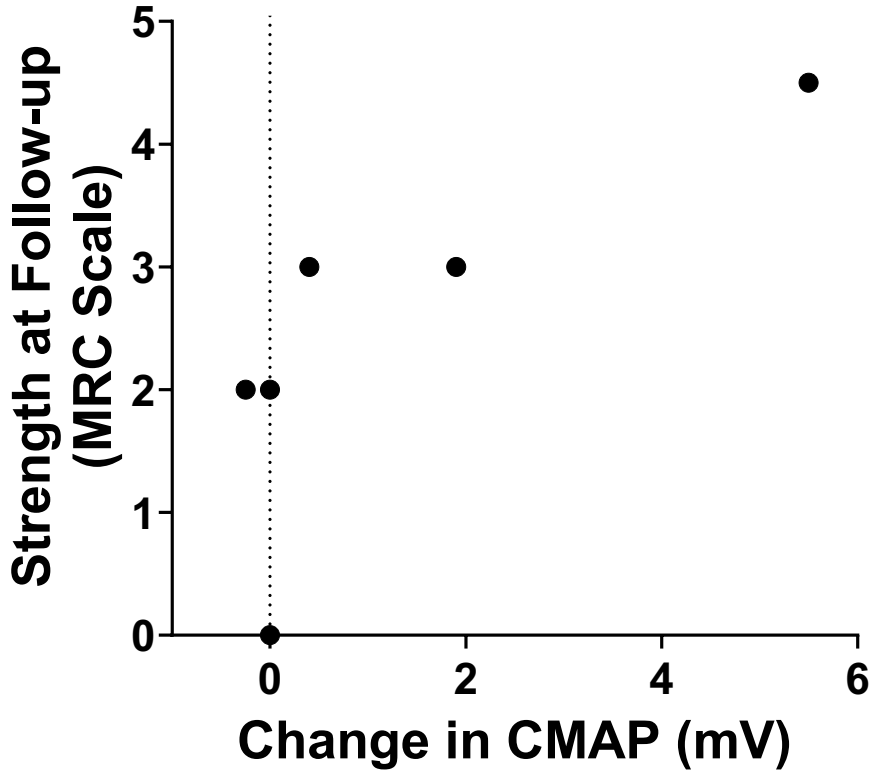
- 22 EMG/NCS studies from 18 patients
  - 16 patients evaluated at Johns Hopkins and Kennedy Krieger
  - 2 patient records extracted from Genetic Susceptibility Study
- Range 6 days to 7 years after onset of weakness

Motor Conduction Amplitude Correlates with Clinical Strength and Recovery



Motor Conduction Amplitude Correlates with Clinical Strength and Recovery





Challenges for EMG in Children With AFM

- Age: ability to understand and cooperate, anxiety
- Technical challenges
- Misplaced angst about EMG among providers

PERCEPTION OF PAIN DURING ELECTROMYOGRAPHY IN CHILDREN: A PROSPECTIVE STUDY

NAHLA M. ALSHAIKH, MD, JEIMMY PINZON MARTINEZ, MD, and MATTHEW C. PITT, MD, FRCP

Neurophysiology Department, Great Ormond Street Hospital, London, WC1N 3JH, UK

MUSCLE & NERVE September 2016

- 66% rated EMG comparable or less painful than blood draw
- Key factors: Location and number of muscles tested

Strategies for EMG in Children With AFM

- Focused testing – only what is absolutely needed!
- Sedation – when excessively painful, or potentially dangerous
  - e.g. Phrenic nerve; Diaphragm and Intercostal muscles
- Child Life approaches
  - Collaboration before, during, and after the test
  - Language matters (“Needle” vs. “Pin with a microphone on the tip”)
  - Elicit and follow patient preferences
  - Encourage parent to stay with child
  - Engage the child when possible
  - Distraction techniques when cooperation not needed
  - Medical play

Preparation for EMG with Child Life Specialists

Courtesy of: Ali Van Eck

Uses of EMG in AFM

- Diagnosis
- Prognosis
- Pre-Surgical Evaluation for Nerve/Tendon Transfers
- Insights into Physiology

Uses of EMG in AFM

- Diagnosis
  - If doubt remains after other investigations
- Prognosis
- Pre-Surgical Evaluation for Nerve/Tendon Transfers
- Insights into Physiology

Uses of EMG in AFM

- Diagnosis
- Prognosis
  - Which muscles are likely to recover, by how much?
- Pre-Surgical Evaluation for Nerve/Tendon Transfers
- Insights into Physiology

Uses of EMG in AFM

- Diagnosis
- Prognosis
- Pre-Surgical Evaluation for Nerve/Tendon Transfers
  - Is the target muscle unlikely to recover without surgery?
  - Is the donor nerve sufficiently intact?
- Insights into Physiology

Uses of EMG in AFM

- Diagnosis
- Prognosis
- Pre-Surgical Evaluation for Nerve/Tendon Transfers
- Insights into Physiology

Insights into Physiology

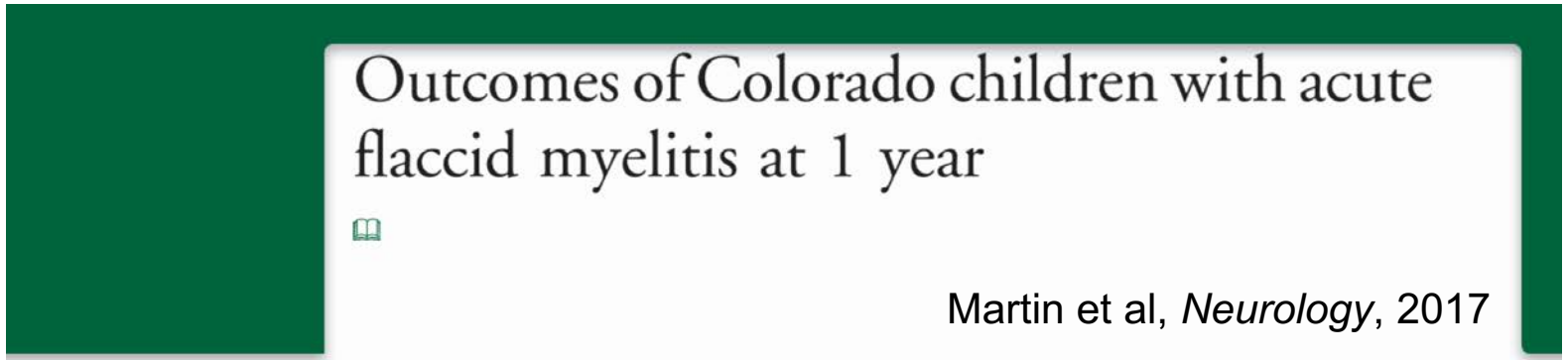
- A child with AFM presented with near complete quadriparesis
- At ~1 month after onset, regains strength in the left arm
- EMG data, left biceps:

Days since onset	Fib/PSW	Recruitment	Motor unit amplitude	Motor unit duration
6	3+	No motor units	-	-
62	Normal	Normal	Normal	Normal

- Spontaneous reversal to normal
- Without evidence of collateral innervation or regeneration

Insights into Physiology

More evidence of “reversibly sick” motor neurons



Martin et al, *Neurology*, 2017

- One patient with similar clinical and EMG data

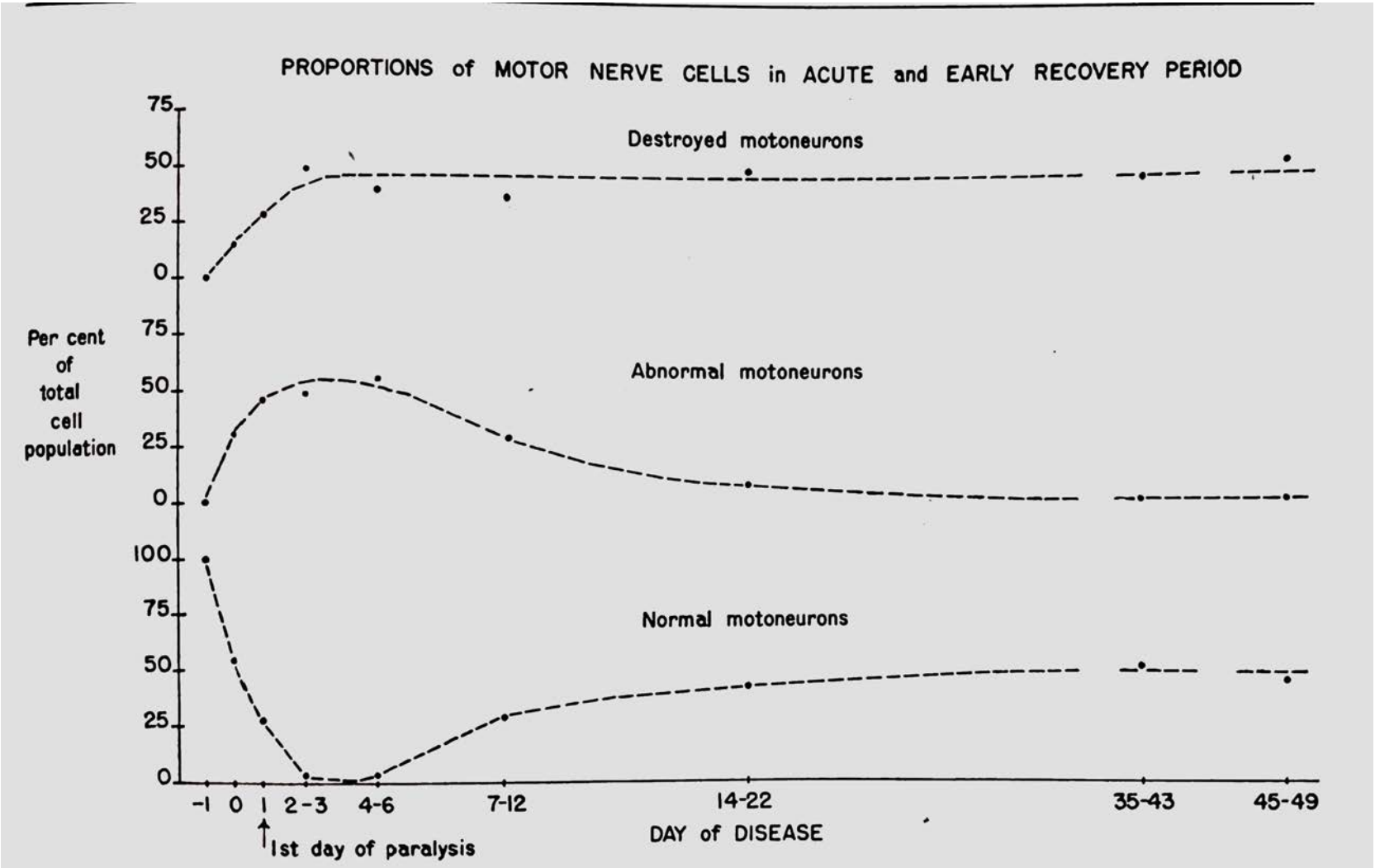
Clinical characteristics of enterovirus A71 neurological disease during an outbreak in children in Colorado, USA, in 2018: an observational cohort study

Kevin Messacar, Emily Spence-Davizon, Christina Osborne, Craig Press, Teri L Schreiner, Jan Martin, Ricka Messer, John Maloney, Alexis Burakoff, Meghan Barnes, Shannon Rogers, Adriana S Lopez, Janell Routh, Susan I Gerber, M Steven Oberste, W Allan Nix, Mark J Abzug, Kenneth L Tyler, Rachel Herlihy, Samuel R Dominguez

- Most patients with complete recovery at 1-2 months

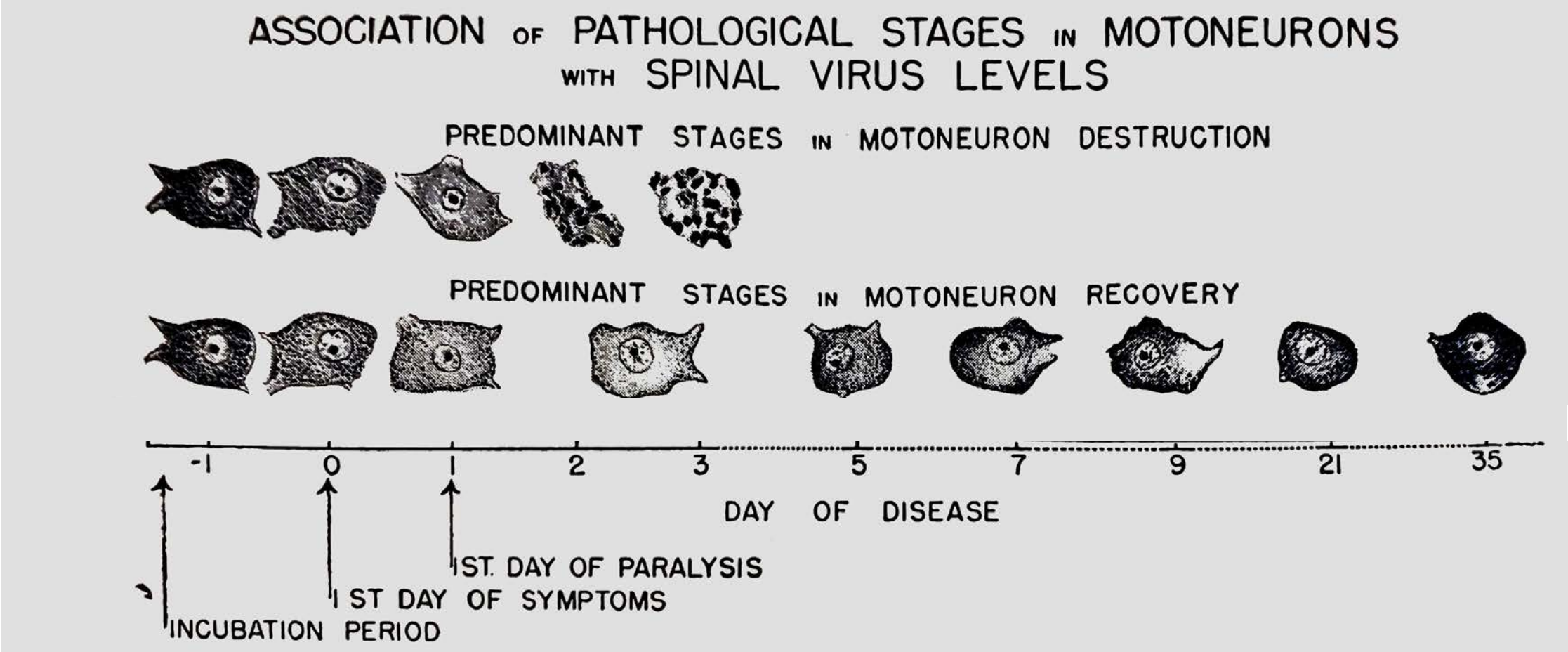
Insights into Physiology

Poliomyelitis: David Bodian, 1940's



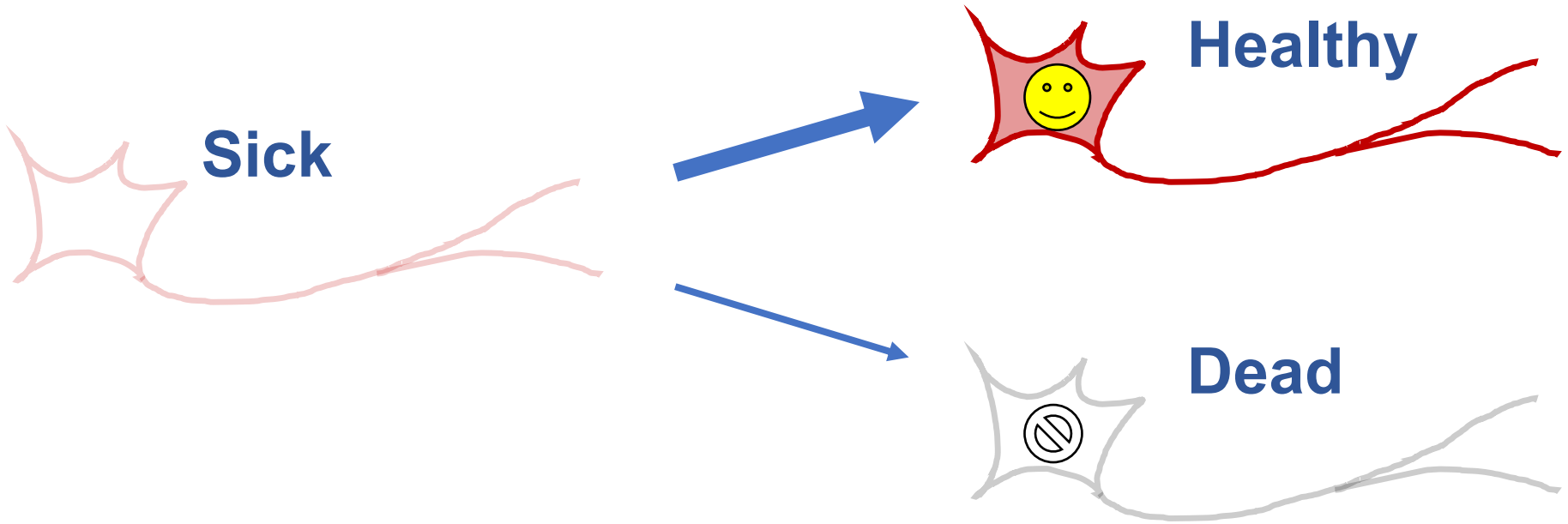
Insights into Physiology

Poliomyelitis: David Bodian, 1940's



Insights into Physiology

- Weakness at the time of presentation may be attributable, in part, to neurons that are “sick” rather than “dead”
- **Therapeutic window for treatments** that target the neuron instead of the virus?



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- Cindy Taylor

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- Jessie Nance

JH Genetic Susceptibility Study
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- Aaron Milstone

AFM Patients and Families!

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AFM: Putting pieces together from admission to discharge

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