# Diagnosis of AFM and Emergency Department Management

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AFM Symposium, June 5, 2020

Children's Hospital of Philadelphia™

# Disclosures

I receive salary support from the US Centers for Disease Control and Prevention for activities related to AFM surveillance.

I am the site PI for the NIAID/NIH AFM Natural History Study

# Objectives

- Review typical presentation
- Document key exam findings
- Summarize initial monitoring, consult, and testing recommendation
- Review initial steps in diagnosis and basics of triage for admission.

# Initial Presentation - Review

- Suspect AFM:
  - Acute onset of flaccid weakness +/- bulbar dysfunction, +/- cranial nerve abnormalities often associated with
    - Recent illness (often febrile URI)
    - Pain head/neck/extremity
    - Weakness with asymmetric onset
    - Absence of significant sensory abnormalities

# AFM Initial Presentation - Exam

- Cranial nerves
  - Eye movements/facial paralysis
  - Palatal elevation/gag/neck strength
- Muscle tone
  - The most involved extremity is flaccid
- Muscle strength
  - Pattern (usually proximal/distal)
  - Strength (against gravity/against resistance)
- Deep tendon reflexes
  - low or absent in the most involved extremity

# What is muscle tone and how do you check it?

- The low level contraction in a muscle at rest, how a muscle resists passive stretching
  - Test it by distracting the patient and moving the body
- Strength is the force that the muscle generates against resistance
  - Tested by how well an extremity moves against gravity or pressure

# Consider Other Diagnoses (not an exhaustive list)

Encephalopathy – Consider ADEM

Seizures – Consider infectious encephalomyelitis/ADEM

Widespread sensory abnormalities: Transverse myelitis/GBS/ cord compression

Ascending paralysis with sensory loss: GBS

No CSF pleocytosis: GBS, cord infarction

Others: neuroinflammatory conditions (neuromyelitis optica, anti-myelin oligodendrocyte glycoprotein antibody associated demyelination (anti-MOG))

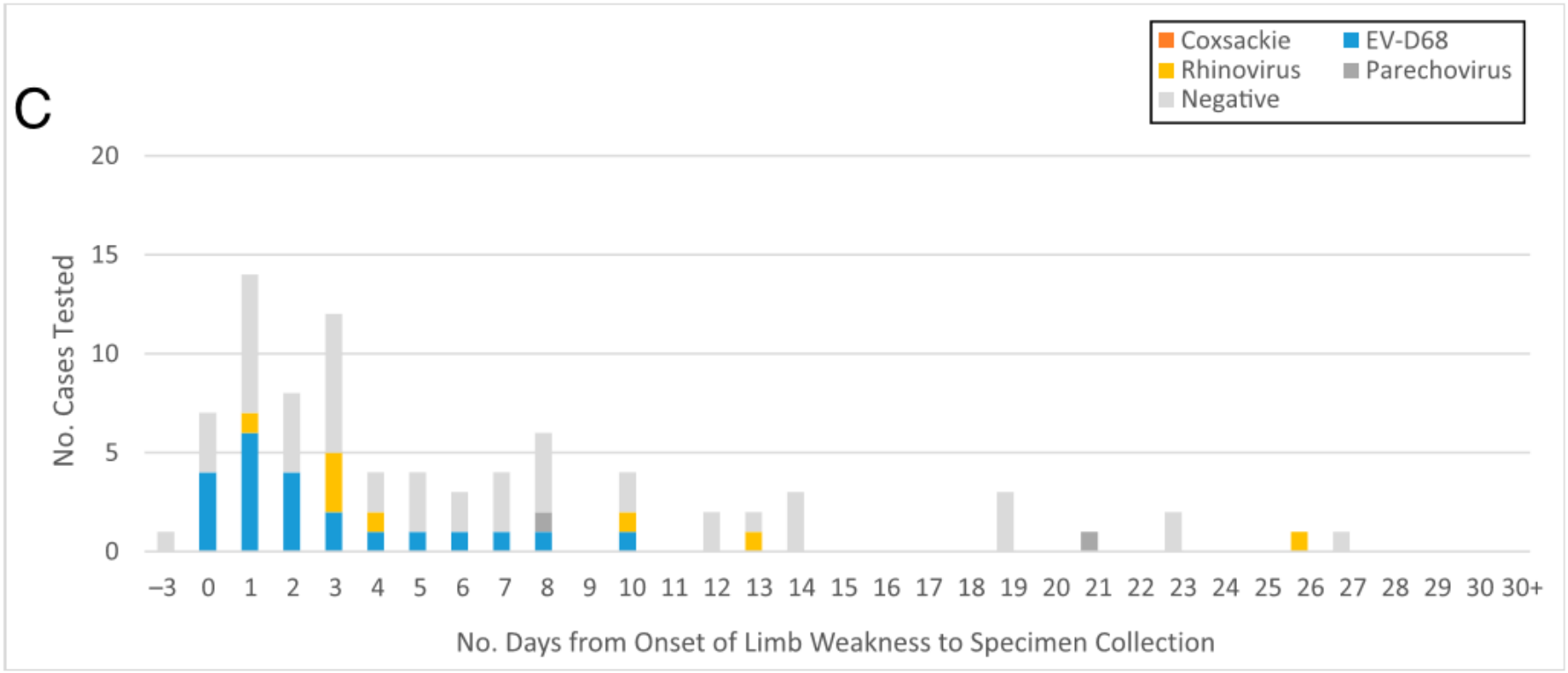
# Suspected AFM - Next Steps

- Assure safety:
  - Autonomic dysfunction
    - Regular vital signs
  - Bulbar dysfunction
    - Consider NPO status
  - Respiratory decline
    - Higher risk if bulbar/cranial nerve/upper extremity weakness
    - Monitor pulmonary function testing
      - Negative inspiratory force every 4 hours
      - Be wary of oxygen supplementation

# Suspected AFM - Next Steps

- Consult key teams – neurology and infectious diseases
- Obtain key specimens
- Diagnostic testing
  - MRI of the spinal cord  and brain w/wo IV contrast
    - May consider a more limited study in certain situations
  - Lumbar puncture
- Treatment

# Obtain Key Specimens Early
## Yield of Respiratory Samples, 2015-2017

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Ayers T, Lopez A, Lee A, Kambhampati A, Nix WA, Henderson E, Rogers S, Weldon WC, Oberste MS, Sejvar J, Hopkins SE, Pallansch MA, Routh JA, Patel M.: Acute Flaccid Myelitis in the United States: 2015-2017. Pediatrics 144(5), November 2019.

# Suspected AFM – Laboratory Studies

| | |
|---|---|
| NP: | Respiratory virus panel, NP swab or aspirate for enterovirus * |
| Blood: | Enterovirus PCR, other microbiological studies depending on season and location |
| Serum: | NMO IgG, anti-MOG IgG |
| CSF: | Cell count, protein, glucose, Enterovirus PCR, other micro studies depending on season and location |
| Stool: | Enterovirus PCR |



*send as soon as AFM is suspected

# Suspected AFM
# Diagnostic Testing

- Diagnostic testing
  - MRI of the spinal cord
    - Typical longitudinal gray matter hyperintensities
  - Lumbar puncture
    - CSF pleocytosis is typical, +/- protein elevation
- Minimize sedation as possible
- MRI/CSF done early may be negative

# Admission

- Recommend admission for close monitoring, and to establish the diagnosis
- Consider ICU if bulbar/cranial nerve/upper extremity weakness, or concerns from pulmonary function testing.
- Regardless of what service the patient is admitted to, close monitoring of vitals and respiratory function is essential – minimum of every 4 hours.

# Neuroimaging as a tool in the diagnosis of AFM

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