The Pediatric Patient with Acute Weakness: When to Think about AFM

Elizabeth Wells, MD, MHS
Medical Director, Inpatient Neurology
Children's National Hospital
Associate Professor of Pediatrics & Neurology
The George Washington University School of Medicine
Washington, DC

Children's National

Clinicians: Timing is Key for AFM

Recognize AFM early

Be alert for onset of acute flaccid limb weakness and consider AFM on your differential diagnosis

Collect specimens & then get MRI

Collect cerebrospinal fluid (CSF), serum, stool, and nasopharyngeal (NP) swab as soon as possible, and handle and store specimens properly

Rapidly report to health department

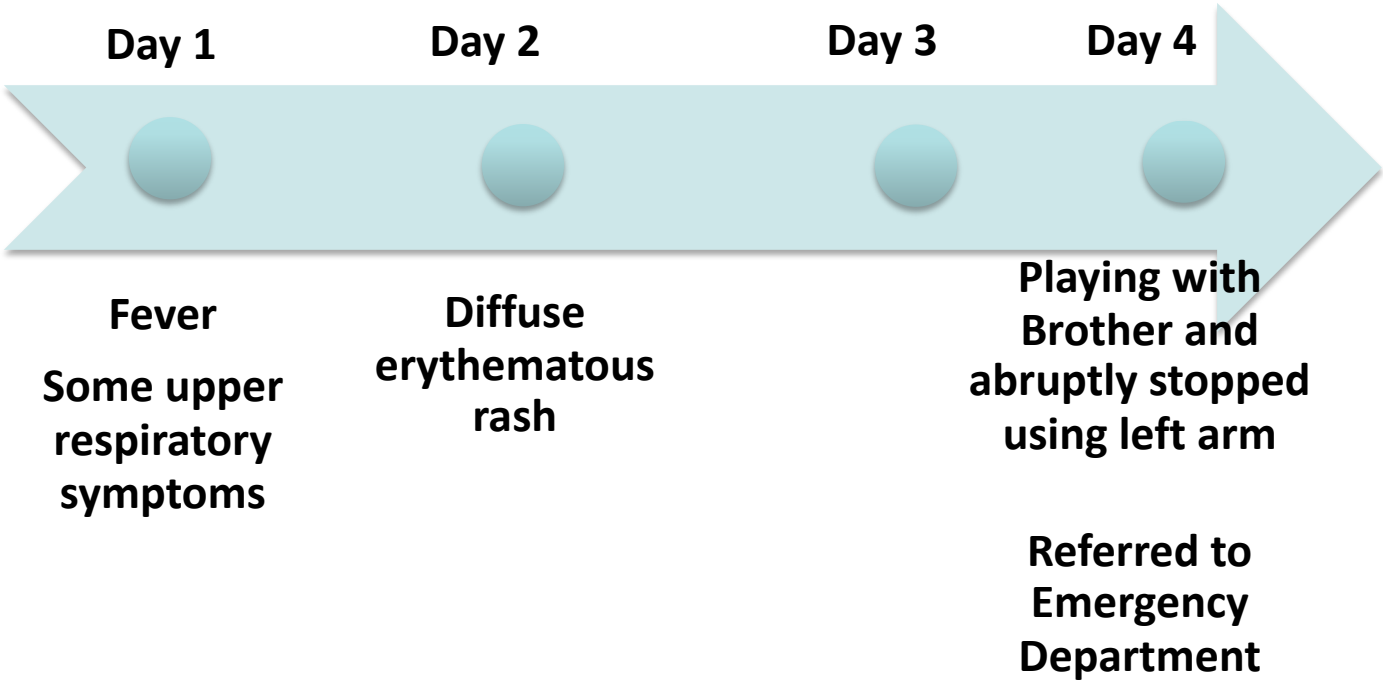
If the MRI shows a spinal lesion with some gray matter involvement, alert the health department and send specimens and medical records

Diagnosis & medical management

Refer to specialists, monitor for signs of worsening symptoms, hospitalize if indicated, and begin treatment and rehabilitation

Meet “Antonio”

6 year-old boy presenting to the Emergency Department



Past Medical and Social History

- 6 year-old, normal development
- No medications, allergies or surgical history
- Lives with parents, 4 year old brother and maternal grandparents in Maryland-all well
- 2 healthy dogs
- No travel or known insect exposures
- Immunizations UTD
- No family hx of autoimmune disease

Physical Exam

T 36.9C HR 118 RR 32 BP 91/57 O_2 99%

Gen: non-toxic appearing 6 month old boy

Heart/Lungs/Abdomen: normal

Skin: fading papular rash on trunk

MSK: no erythema, tenderness or swelling of joints

Neuro: attentive, smiles in mothers lap; tracks appropriately; CN II-XII intact, discs flat, symmetric facies. Occasional head drop. RUE and BLE full ROM and strength. L arm limp, hanging at his side with decreased tone, does not reach for toy; decreased muscle tone, diminished DTRs left bilateral brachioradialis, biceps, triceps; LE DTRs intact

Laboratory and Diagnostic Testing

- L Shoulder X-ray: normal
- Head CT: negative for acute findings

	12.6	
8.1		382
	37.4	

CRP: 0.27
ESR: 32

CK: 92

CSF:
WBC: 84
RBC: 405
Segs: 35%
Lymphs: 20%
Monos: 22%
Eos: 1%
Plasmacytoid: 22%

CSF glucose: 59
CSF protein: 51

CDC Acute Flaccid Myelitis (AFM) Case Definition

Acute onset flaccid limb weakness

Confirmed AFM Case

MRI findings of spinal cord lesion largely restricted to gray matter and spanning one or more spinal segments*

OR

Probable AFM Case

CSF with pleocytosis (>5 cells/mm3)

www.cdc.gov/acute-flaccid-myelitis

When to think about AFM?

Pediatric Patient with Acute Weakness

When to think about AFM?

Pediatric Patient with Acute Weakness

- Usually young child (median 6.3 years)
  - M=F
  - All races/ethnicities
- Usually previously healthy
  - Without a prior acute neurologic (demyelinating) event

When to think about AFM?

Pediatric Patient with **Acute** Weakness

- Most have rapid progressive weakness
- Some have a stuttering start or prolonged plateau rather than fulminant onset
- Hours to days

When to think about AFM?

Pediatric Patient with Acute **Weakness**

- Asymmetric
- One or more limbs
- Upper/proximal > lower/distal
- Range in severity
- Often also weak neck, trunk, diaphragm
- 30% bulbar weakness
- +/- weakness of eyes, face, neck

Neurologic Exam:

- Proximal muscle weakness
  - raise hands over head
  - Sit to stand up
- Axial weakness (neck)
- Decreased muscle tone, DTRs in weak limb
- Cranial nerves

Children's National

How to Spot Symptoms of Acute Flaccid Myelitis in Your Child

Acute flaccid myelitis, or AFM, is a rare but serious condition that affects the nervous system.

Symptoms of AFM

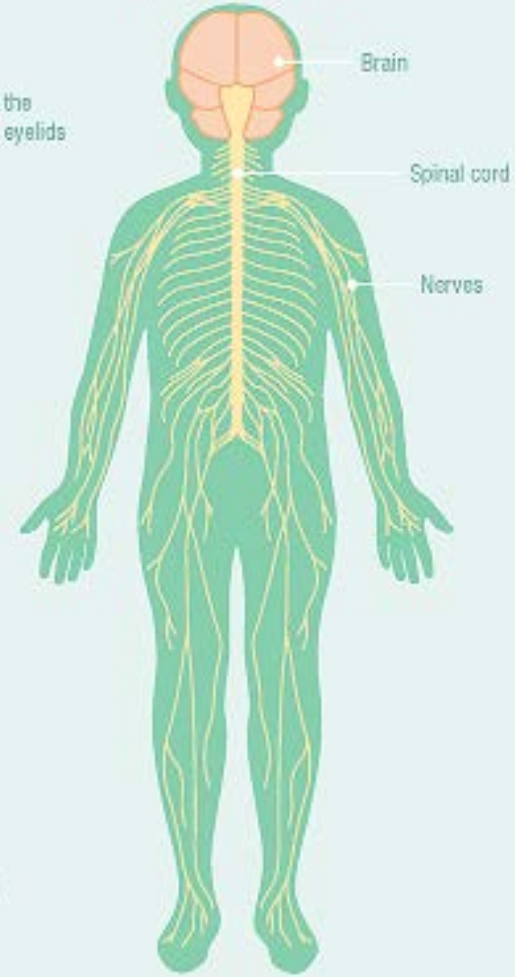
Difficulty moving the eyes or drooping eyelids

Facial droop or weakness

Difficulty with swallowing or slurred speech

Sudden arm or leg weakness

The Nervous System



Seek medical care right away if your child has any of these symptoms.

www.cdc.gov/acute-flaccid-myelitis

Children's National

Associated Symptoms

- Often prodromal fever and respiratory symptoms
  - Usually resolves before weakness
  - Not necessary for diagnosis
- Less frequently GI symptoms
- Sometimes headache, stiff neck, return of fever
- Pain in affected limb, lower back, neck
- Bowel/bladder dysfunction
- Autonomic manifestations
  - labile blood pressure, irregular heart rate and breathing patterns
- Not typical: sensory, encephalopathy, seizures

Differential Diagnosis/Conditions AFM can Mimic

	Polio-myelitis	Acute Flaccid Myelitis	Demyelinating Transverse Myelitis	Guillain-Barre	Stroke
Symptom onset	Days	**Hours-Days**	Hours-Days	Days	Minutes
Weakness	Lower>Upper, Asymmetric	**Usually 1 limb (can be multiple) Asymmetric Upper>Lower**	**Multiple limbs**	Legs>Arm Symmetric	Usually lateralized
Reflexes	Diminished or absent	Diminished/ absent (may be present early)	Diminished/ Absent acutely	Diminished/ absent (may be present early)	Diminished/ absent acutely; then hyperreflexic
CSF	pleocytosis	Often pleocytosis	pleocytosis	Cytoalbumino-dissociation	**Usually normal**
MRI Finding	Gray matter predominant	Gray matter predominent	Gray and white matter	**Nerve root enhancement**	Often anterior with diffusion restriction

Also consider: acute disseminated encephalomyelitis, botulism, tick paralysis

Conclusions

- Strongly consider AFM in a pediatric patient with rapid onset weakness
  - Especially after respiratory illness or fever
  - Especially in August through October
  - At other times too!

- Early recognition -> prompt reporting/investigation, appropriate specimen collection, the best care

Acknowledgements

CNH Acute Flaccid Myelitis Taskforce

Justin Burton, MD
Roberta DiBiasi, MD
Jessica Carpenter, MD
Nada Harik, MD
Jonathan Murnick, MD, PhD
Alexandra Yonts, MD
Alyssa Doslea, MS
Elizabeth Wells, MD, MHS

Neurology & Infectious Disease Divisions

Patients & Families