Critical Care Management of AFM

Acute Flaccid Myelitis:
What we have learned in order to be prepared

Symposium for Health Care Providers and Scientists

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Jessica L Carpenter MD
Associate Professor of Neurology and Pediatrics
George Washington University School of Medicine
Children's National Medical Center

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NO DISCLOSURES

OBJECTIVES

- Describe the frequency with which AFM patients are admitted to the ICU
- Define which AFM patients would benefit from ICU admission
- Delineate the symptomatology of AFM and how to monitor for deterioration
- Establish the importance of supportive care

Critical care for AFM

Current literature: > 50 % of AFM patients are admitted to an ICU at some point in their disease process.

Cohort (n)	% ICU admission	% Mortality
Kane et al, California 2011-2016 (28)	74% (20)	3.5% (1)
Knoester, Europe 2016 (29)	NR	7% (2)
Chong, Japan 2015 (59)	NR	0
Sejvar, US- CDC 2014 (120)	52% (58)	0
Lopez, US – CDC 2018 (288)	60% (127)	0.01% (2)
CNH, Washington DC 2014-2019 (20)	50% (10)	0

Acute mortality is rare

ICU need is primarily for respiratory support.

Goals of ICU Care

- Goal #1 → Limit complications through rigorous monitoring and supportive care
- Goal #2 → Facilitate a rapid and accurate diagnosis
- Goal #3 → Provide disease specific treatment

Criteria for ICU Admission

Variable across institutions:

Model 1

- Suspected AFM diagnosis = ICU admission for first 24 hours after presentation

Model 2

- High risk patients
  - bilateral upper extremity weakness
  - bulbar dysfunction
  - MRI with cervical cord lesion

Model 3

- Patients with respiratory insufficiency/failure

AFM Symptomatology

- Weakness
- Respiratory distress
- Feeding dysfunction
- Pain/parasthesias
- Neurogenic bowel/bladder
- Autonomic dysfunction

Supplemental Table: Categorization of patients based on extremity involvement at presentation

Extremity paresis/paralysis on presentation	Other associated symptoms on presentation	Neurologic Exam at 12 months
four limbs (11)	truncal weakness (2), respiratory failure (7), paraspinal pain, headache, photophobia, dysphagia (3), vocal cord paralysis, altered mental status (2), 6th nerve palsy, neurogenic bowel/bladder, seizure, extraocular deficits	full recovery (2) residual deficits (9)
bilateral legs (10)	urinary retention, constipation, truncal weakness, headache, paraspinal pain	full recovery (7) residual deficits (3)
bilateral arms (2)	neck/arm pain, neck weakness, truncal weakness	full recovery (1) residual deficits (1)
1 upper limb, 1 lower limb (1)	dysphagia, urinary/bowel incontinence	full recovery (0) residual deficits (1)
only 1 extremity involved (4)	neck pain, facial asymmetry, headache	full recovery (1) residual deficits (2) lost to follow up (1)

Supplemental table categorizes patients based on most extensive extremity involvement around presentation (within 3 days of diagnosis), other body system involvement including possible supratentorial/cranial nerve involvement,

Kane, Miranda S., et al. "Incidence, risk factors and outcomes among children with acute flaccid myelitis: a population-based cohort study in a California Health Network between 2011 and 2016." *The Pediatric infectious disease journal* 38.7 (2019): 667-672

Progression of Symptoms

- Onset to nadir typically evolves in hrs to days.
  - 8/16 patients → hyperacute pattern (<6h)
  - 7/16 patients → acute pattern (<72h)
  - 1/16 nadir reached during sedation owing to asthmatic respiratory failure (time to nadir unknown.

- Usually no second phase of worsening in the acute period

Gordon-Lipkin, Eliza, et al. "Comparative quantitative clinical, neuroimaging, and functional profiles in children with acute flaccid myelitis at acute and convalescent stages of disease." *Dev Med & Child Neuro* 61.3 (2019): 366-375.

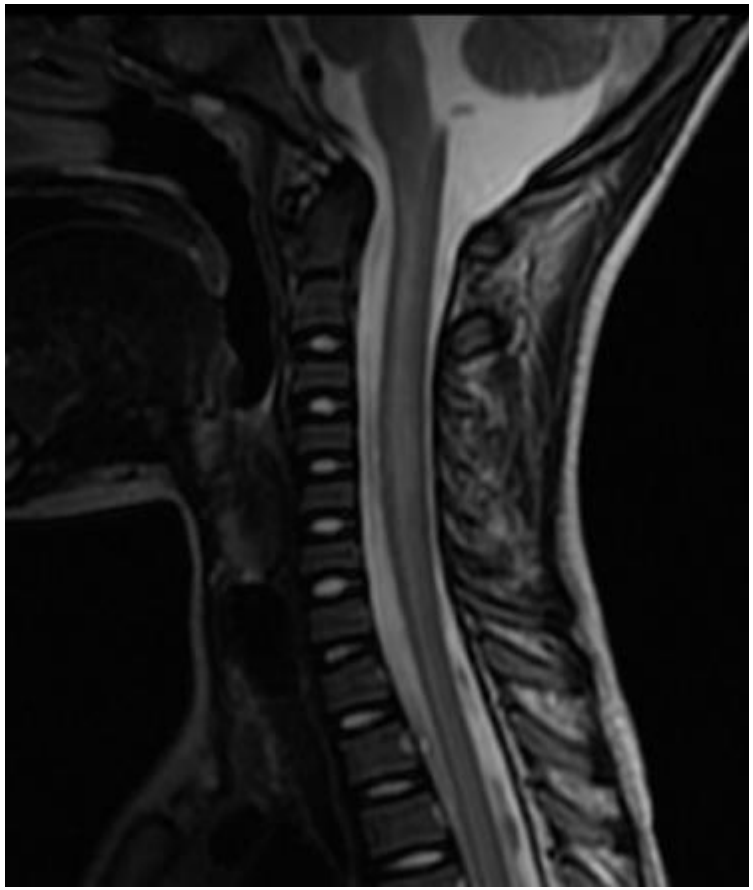
Risk Factors for Respiratory Failure

Currently no good data

Weakness leading to hypoventilation
Weakness of the neck/trunk and/or bilateral UE
Cervical cord lesion → specific but not sensitive

Aspiration
Bulbar weakness
Encephalopathy

Apnea
Brainstem lesion (central)
Bulbar weakness (obstructive)
Weakness of the neck/trunk (obstructive)

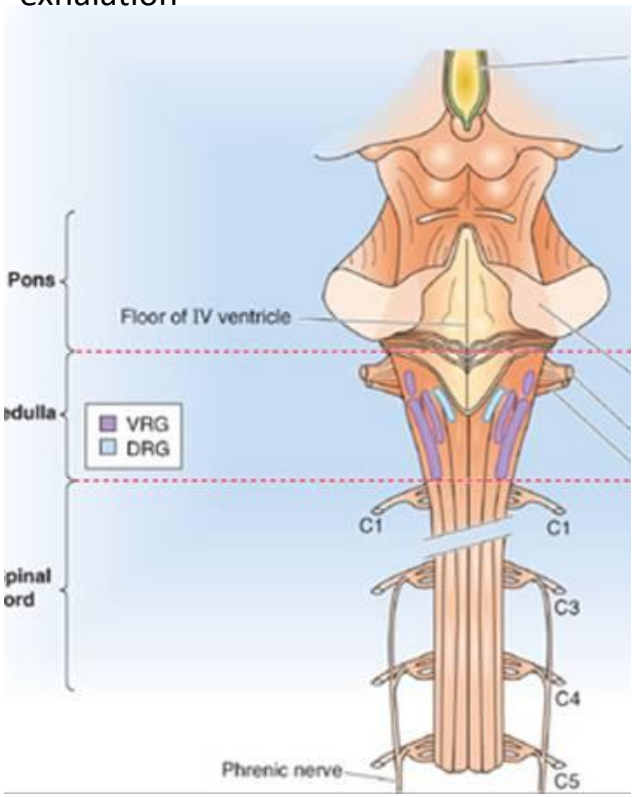


Anatomy and Physiology

Innervation of the Primary and Some Secondary Muscles of Respiration

Muscles	Nerve	Location of Cell Body of Motor Neuron
Primary Muscles of Inspiration		
Diaphragm	Phrenic nerve	Phrenic motor nuclei in ventral horn of spinal cord, C3-C5
Intercostal muscles	Intercostal nerves	Ventral horn of thoracic spinal cord
Secondary Muscles of Inspiration		
Larynx and pharynx	Vagus (CN X) and glossopharyngeal (CN IX) nerves	Primarily within the nucleus ambiguus medulla
Tongue	Hypoglossal nerve (CN XII)	Hypoglossal motor nucleus medulla
Sternocleidomastoid and trapezius muscles	Accessory nerve (CN XI)	Spinal accessory nucleus, C1-C5
Nares	Facial nerve (CN VII)	Facial motor nucleus pons
Secondary Muscles of Expiration		
Intercostal muscles	Intercostal nerves	Ventral horn of thoracic spinal cord
Abdominal muscles	Spinal nerves	Ventral horn of lumbar spinal cord

Phrenic motor neurons (C3, 4, 5) control muscles of inhalation & exhalation



Richerson G & Boron W. Medical Physiology A Cellular and Molecular Approach, CONTROL OF VENTILATION

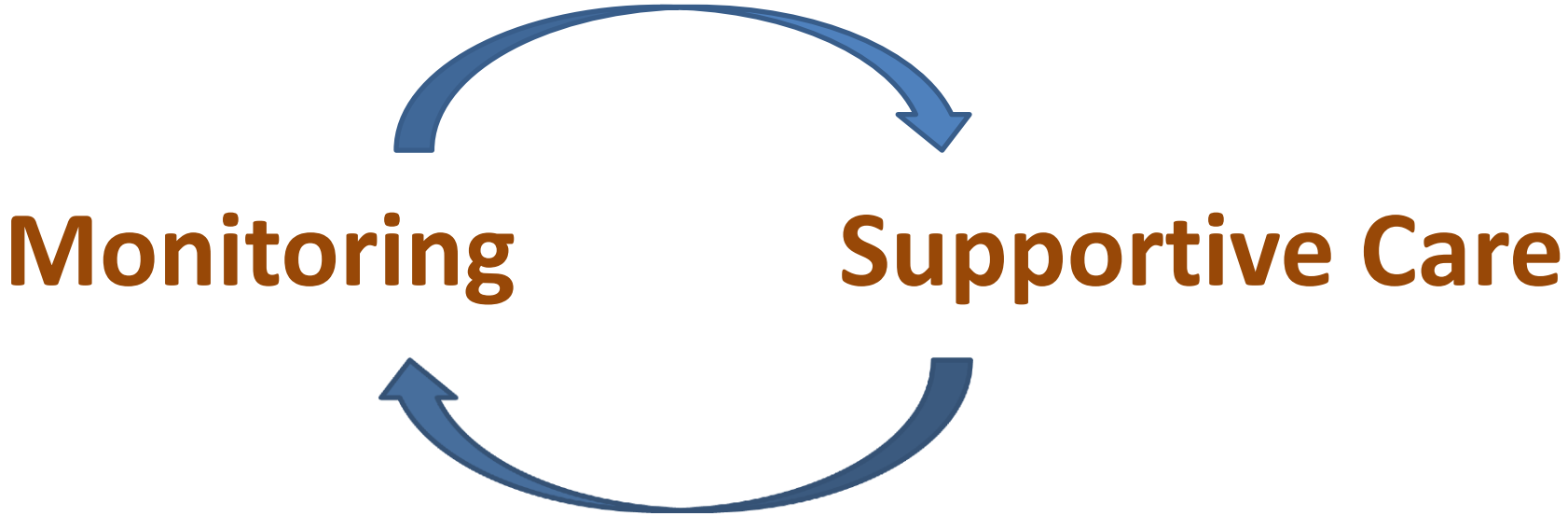
Incidence of Respiratory Failure

While respiratory support is the most common reason for ICU admission, it is unclear what percent require invasive mechanical ventilation (reports are highly variable).

Cohort	Cranial Nerve palsy	Quadriplegia(%)	Cervical cord involvement	Mechanical ventilation
Kane, CA 2011-16 (28)	NR	43% (12)	25% (7)	NR
Knoester, Europe 2016 (29)	59% (17)	55% (16)	55% (16)	66% (19)
Chong, Japan 2015 (59)	17% (10)	19% (11)	NR	8% (5)
Sejvar, US 2014 (120)	28% (34)	25 % (30)	87% (103)	20% (26)
Lopez, US 2018 (227)	NR	NR	NR	23 % (53)
CNH, D.C. 2014-19 (20)	45 % (9)	35 % (7)	85% (17)	15% (3)
Total (% range)	**17 -59%**	**7-30**	**25 – 87%**	**8 -66**

Single Center ICU Experience

Symptomatology	Number of ICU patients (n = 10)	Number of all AFM patients (n = 20)
Constipation	80% (8/10)	75% (15/20)
Neurogenic bladder	30% (3/10)	30% (6/20)
Dysautonomia	0 % (0/10)	0 % (0/20)
Pain	100% 10/10 (6/10 on gabapentin)	75% (15/20) (10/20 on gabapentin)


Monitoring
Supportive Care

Recognizing Respiratory Distress from Neurological Causes

Intrinsic Lung Disease

- Acute distress
- Cough
- Wheezing, crackles, decreased breath sounds
- Retractions
- Tachypnea noted on the CR monitor with clear waveform
- Hypoxia

**** The distress is visible and audible**

Neurological Disease

- "No acute distress", frozen
- Sleepy or lethargic in appearance
- Lung sounds may be clear or fine crackles may be present
- CR monitor with unreliable RR
- Oxygen saturation is normal until respiratory compromise is severe

**** The distress is not clearly visible or audible**

Monitoring for Respiratory Dysfunction

Hypoventilation

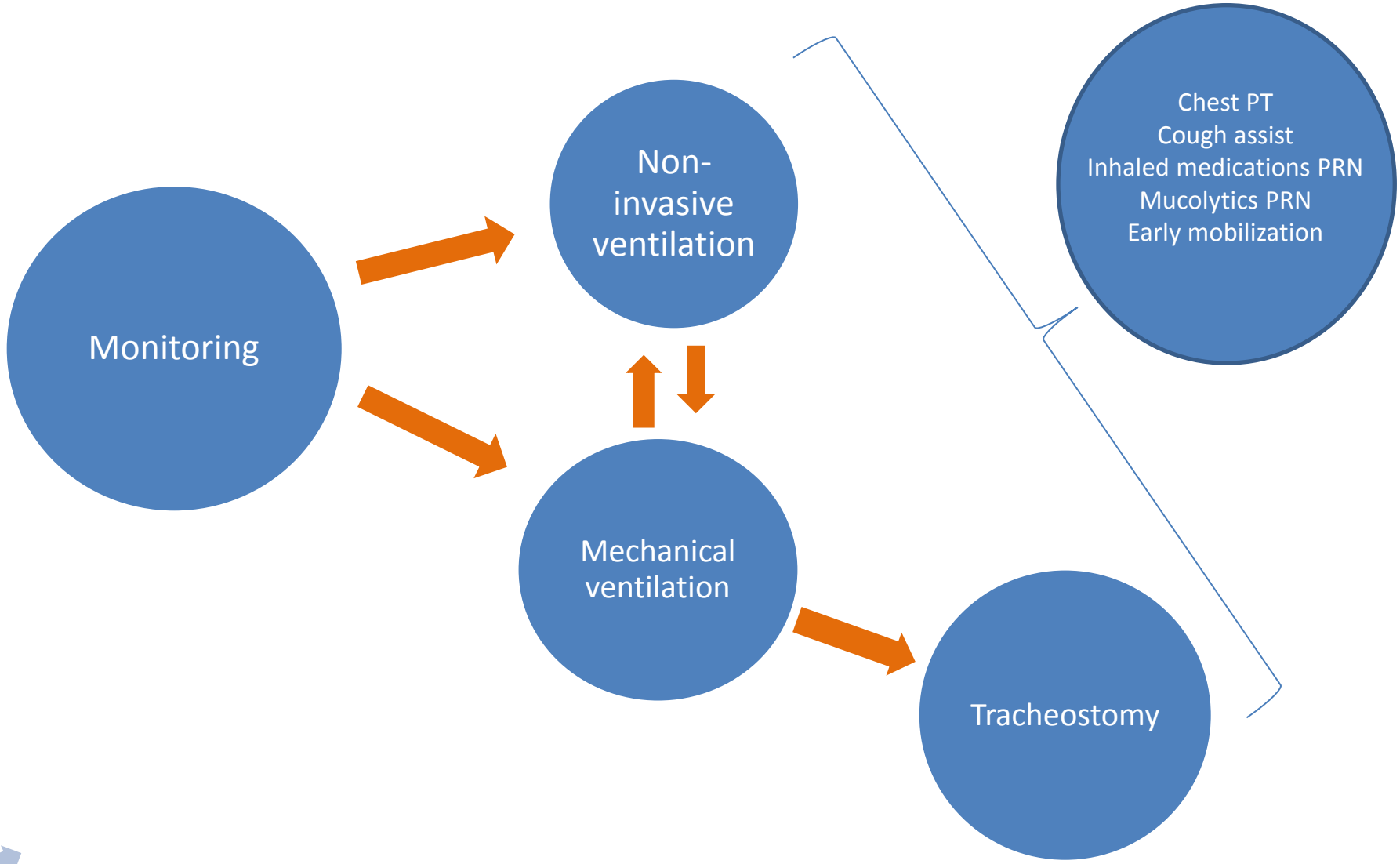
- consider Q4 hr assessment for 1st 48 hours
- NIF/forced vital capacity (age > 6yr, cooperation dependent)
- Counting or singing a song
  (pick a task the child repeat consistently to assess for changes over time)
- Pulse oximetry
  - hypoxia is a late sign or evidence of secondary process

Apnea

- CR monitor
- +/- sleep study

** assess for concomitant conditions (e.g. approximately 30% of patients with prior dx of asthma)

Supportive Care – A Spectrum of Respiratory Care



Monitoring and Supportive Care

Aspiration

- Identify high risk patients
  - Bulbar weakness (hypophonic voice, drooling/copious secretions, dysarthria, fatigue with talking, coughing with drinking, pocketing food)
  - Encephalopathy
  - Make patient NPO and obtain speech consult for swallowing eval

- Identify aspiration promptly
  - Symptoms of fever, hypoxia, and tachypnea/dyspnea
  - Modify PO status where applicable
  - Assess for infection
  - Increase respiratory support as needed

Monitoring and Supportive Care

Neurological assessments→

- Neuro checks
  - Any interval leads to diminishing returns if prolonged
  - Sleep deprivation and delirium confuse the picture.
- Ask the family to video the motor exam, speech and facial movements.

Hemodynamics →

- CR monitor
- Hypertension is common
  - Etiology is often multifactorial
  - Assess for pain, neurogenic bladder, autonomic dysfunction

Monitoring and Supportive Care

Pain management

- Children may not report pain when asked directly.
- Irritability or poor cooperation may be indicators of poorly controlled pain
- Good pain control can normalize vital signs and improve cooperation with assessments and therapies.
- Encourage use of non-narcotic pain medications (e.g. gabapentin)
- Consult psychology and/or Child Life early to assist with anxiety and behavioral challenges
- Promote sleep and maintenance of day/night cycles

Sedation

- Minimize sedation where possible
- Avoid medications that may exacerbate weakness (e.g. paralytics, steroids)

Monitoring and Supportive Care

Bowel/bladder dysfunction

- Gut dysmotility
  - Constipation is common.
  - Initiate bowel regimen early
- Neurogenic bladder
  - Ask and assess for bladder retention.
  - In a child who is continent, ask them to urinate then perform a bladder scan or post void residual.
  - For neurogenic bladder → in and out cath preferred over continuous Foley catheterization

Supportive Care

Nutrition

- Avoid gaps in feeding
- Modify diet and encourage enteral feeds when appropriate/safe
- Place NG tube early if risk for aspiration

Mobility and rehabilitation in the ICU

- Consult PT/OT and PM&R early for assessment
- Plan for mobilization
  - establish when mobilization is safe
  - Set activity goals

Supportive Care

Skin care (avoidance of pressure palsies)

DVT prophylaxis for immobile patients (post pubertal patients highest risk)

Infection risk

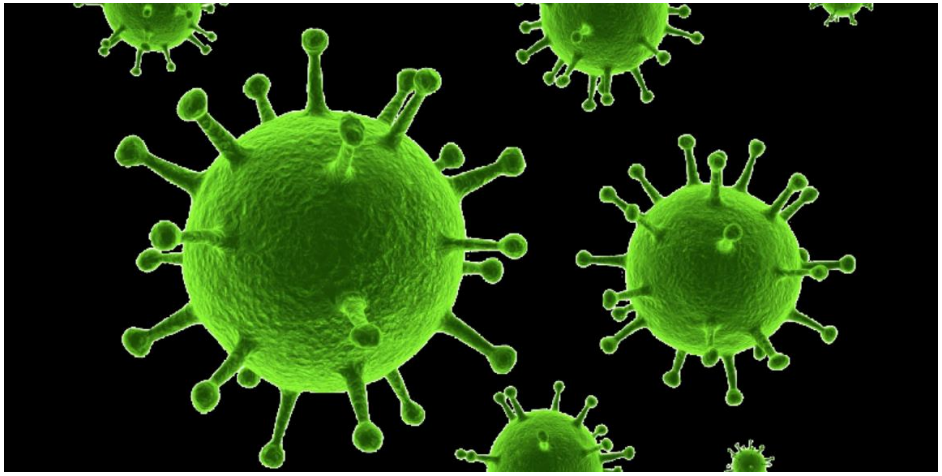
- Aggressive screening for infection & treatment where appropriate
- Central line care
- Avoidance of prolonged use of Foley catheters

Diagnosis

- Refer to AFM diagnostic criteria – labeling is important
- Physical Exam
- LP for CSF
- MRI

Anesthesia – considerations

- Laboratory studies
- EMG – variable, timing



Diagnosis

SUPPLEMENTARY TABLE. Timing from limb weakness onset to medical care, collection of specimens, and reporting to CDC for confirmed cases of acute flaccid myelitis by year — United States, 2016 and 2018 outbreaks

Event	2016 (N = 149) Median days (range, IQR)	2018 (N = 233) Median days (range, IQR)
Any illness	5 (0–28, 2–8)	5 (0–27, 2–8)
Hospitalization	1 (0–31, 0–3)	1 (0–54, 0–2)
Lumbar puncture	2 (0–32, 1–4)	2 (0–31, 1–3)
MRI	3 (0–145, 1–6)	2 (0–164, 1–3)
CSF specimen collection	3 (0–27, 1–5)	2 (0–31, 1–4)
Respiratory specimen collection	4.5 (0–34, 2–12)	3 (0–35, 2–6)
Serum specimen collection	4 (0–46, 2–9)	4 (0–31, 2–7)
Stool specimen collection	7.5 (1–63, 4.5–13.5)	7 (0–44, 4–11)
Completion of patient summary form	10 (0–582, 2–24)	8.5 (1–175, 4–25)
CDC notified	15 (0–344, 8–34)	18 (0–208, 7–35)

Abbreviations: CSF = cerebrospinal fluid; IQR = interquartile range; MRI = magnetic resonance imaging.

MRI brain and spine

- Identification of cervical lesions may help guide the intensity of care needed.
- MR may be normal early in the course, consider repeat in 2-7 days if findings not consistent with AFM initially

Outcomes

- Data is incomplete

- Kane et al, 12 month follow up (27/28)
  - 11 % (3) with tracheostomy (2 placed acutely, 1 later) patients
  - 15 % (4) with G tube dependence.

Kane, Miranda S., et al. "Incidence, risk factors and outcomes among children with acute flaccid myelitis: a population-based cohort study in a California Health Network between 2011 and 2016." *The Ped infect Ds J* 38.7 (2019): 667-672.

Conclusions

- > 50% of AFM patients are admitted to an ICU during the acute phase
  - ~ 1/5 required invasive respiratory support
  - Most are monitored and supported briefly and before transitioning to floor status.

- ICU admission should be considered for patients with weakness of the bilateral upper extremity and/or neck/trunk and/or bulbar dysfunction

Conclusions

- Respiratory failure occurs predominantly in patients with brainstem and cervical involvement.
  - The neurological exam is the best guide
  - Imaging abnormalities may contribute to identifying high risk patients

- Good supportive care can minimize the impact of the primary disease:
  - mobilize earlier
  - suffer fewer ICU complications
  - spend less time in the ICU overall.

Respiratory issues in the acute phase of AFM

Rebecca J. Riggs, MD.

Assistant Professor of Anesthesiology and
Critical Care Medicine
Pediatric Critical Care Medicine
Johns Hopkins University School of Medicine