Acute Flaccid Myelitis (AFM):
The what, the why, and the role of occupational therapy

Rebecca Martin, OTR/L, OTD, CKTP, CPAM
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Rebecca Martin, OTR/L, OTD
• Manager of Clinical Education and Training at the International Center for Spinal Cord Injury (ICSCI) at Kennedy Krieger Institute
• Assistant Professor at The Johns Hopkins University School of Medicine in the Department of Physical Medicine and Rehabilitation
• Speaks internationally on topics related to Activity-Based Rehabilitation
• Research interests are in novel applications of electrical stimulation for the restoration of function lost to spinal cord injury

Disclosures
I, nor any member of my family, receive compensation, financial or otherwise, for any services or products discussed herein.

Objectives
At the conclusion of this webinar, participants will:
• Understand the history of AFM
• List the diagnostic criteria, epidemiology, and clinical characteristics
• Understand the medical management of children with AFM
• Describe occupational therapy’s distinct role and contribution to interdisciplinary management of children with AFM
• Identify improvements for patient with AFM associated with intensive rehabilitation
WHAT IS ACUTE FLACCID MYELITIS

History of AFM

- Acute Flaccid Myelitis (AFM) is not new
  - Polio is the most common cause

- Acute Flaccid Paralysis in monitored worldwide.
  - Includes polio, non-polio, GBS, toxic neuropathies
  - Expected to occur 1/100,000 children worldwide

- AFP not reportable in USA
  - Understanding of incidence in USA is limited

Emergence of AFM in US

- California, 2012
  - Calls to the health department for testing in two adults for polio
  - Between 2012-2015 with 59 cases identified in CA

- Colorado, 2014
  - Unusual cluster of neurologic illness presenting as acute limb weakness seen in hospitals in Denver area.¹
  - Limb and cranial nerve weakness, fever, spinal MRI showing signal abnormalities in gray matter…polio?

- Between August and December of 2014, 120 children from 34 states²

- At the same time, there was a nationwide outbreak of a severe respiratory illness among children due to enterovirus-D68 (EV-D68)¹

AFM Cases in US per CDC¹

- From August to December 2014, 120 confirmed cases of AFM in 34 states.
- In 2015, 22 confirmed cases in 17 states.
- In 2016, 149 confirmed cases in 39 states and DC.
- In 2017, 35 confirmed cases of AFM in 16 states.
- In 2018 YTD, 134 confirmed cases in 33 states.
  - 165 additional patients under investigation
Confirmed cases of AFM in the US by month, as reported to the CDC

Reported enterovirus D68 circulation

No reported enterovirus D68 circulation

Month of onset

Diagnostic Criteria

- Confirmed case
  1. Acute onset of focal limb weakness, AND
  2. An MRI showing a spinal cord lesion largely restricted to gray matter* and spanning one or more spinal segments

- Probable case
  1. Acute onset of focal limb weakness, AND
  2. Cerebrospinal fluid with pleocytosis

- 2017 update
  - Added in the word “flaccid” to better describe
  - Spinal cord lesions may not be present on initial MRI (<72hrs)
  - Removed age restriction

Clinical Characteristics

- Prodromal, often febrile, illness preceded onset of neurological symptoms by 5 days in most patients
- Most patients reported clinical improvement of illness before return of the fever along with headaches, stiff neck or pain in neck, back or affected limb around the time of neurological deficit onset
- Timing, quality, and pattern of limb weakness in AFM consistent with acute lower motor neuron disease
- Rapid progression from full strength to weakness over hours to a few days

Signs and Symptoms

- Severity ranged from complete paralysis to mild weakness, flaccid in nature
  - Typically asymmetric
  - UES more commonly affected
- Cranial nerve dysfunction, may include hypophonia, dysarthria, dysphagia, diplopia, facial weakness
- Pulmonary insufficiency to ventilator dependent
- Sensory deficits (varying reports)
- Varying reports of altered mental status
- Seizures very rare (4%)
- Varying bowel and bladder dysfunction
- Recovery progresses distal to proximal
Demographics and Clinical Findings Among Acute Flaccid Myelitis Cases, 2014 (N = 120)³

<table>
<thead>
<tr>
<th>Variable/No. With Information</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, median (range; IQR) (n = 119)</td>
<td>7.1 y (0.4–20.8 y; 4.8–12.1 y)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>71 (59)</td>
</tr>
<tr>
<td>Female</td>
<td>49 (41)</td>
</tr>
<tr>
<td>Respiratory or febrile illness preceding limb weakness (n = 117)</td>
<td>105 (90)</td>
</tr>
<tr>
<td>Limb involvement (n = 119)</td>
<td></td>
</tr>
<tr>
<td>Upper extremity(ies) only</td>
<td>41 (34)</td>
</tr>
<tr>
<td>Lower extremity(ies) only</td>
<td>37 (31)</td>
</tr>
<tr>
<td>Upper and lower extremities, but not all extremities</td>
<td>21 (18)</td>
</tr>
<tr>
<td>All 4 extremities involved</td>
<td>30 (25)</td>
</tr>
<tr>
<td>Cranial nerve findings (n = 120)</td>
<td>34 (28)</td>
</tr>
<tr>
<td>Altered mental status (n = 109)</td>
<td>12 (11)</td>
</tr>
<tr>
<td>Seizures during illness (n = 116)</td>
<td>5 (4)</td>
</tr>
<tr>
<td>Required mechanical ventilation (n = 109)</td>
<td>26 (20)</td>
</tr>
</tbody>
</table>

Function

<table>
<thead>
<tr>
<th>Level of Function</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete dependence on caretakers</td>
<td>8 (14)</td>
</tr>
<tr>
<td>Somewhat functionally impaired</td>
<td>38 (68)</td>
</tr>
<tr>
<td>Fully functional</td>
<td>10 (18)</td>
</tr>
</tbody>
</table>

Strength (compared to initial presentation)

<table>
<thead>
<tr>
<th>Level of Strength</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>As weak</td>
<td>11 (20)</td>
</tr>
<tr>
<td>Some improvement</td>
<td>41 (73)</td>
</tr>
<tr>
<td>Full recovery</td>
<td>3 (5)</td>
</tr>
<tr>
<td>More weak</td>
<td>1 (2)</td>
</tr>
</tbody>
</table>

What we know from CDC…

- Most patients with AFM (more than 90%) had a mild respiratory illness or fever consistent with a viral infection before they developed AFM.
- AFM is not caused by poliovirus.
- Some patients with AFM have tested positive for coxsackievirus A16, EV-D71, and EV-D68 but in MANY of the patients no pathogen has been detected.
- Most patients have onset between August and October, with increases in AFM cases every two years since 2014.
- Most AFM cases are children (over 90%) and have occurred in 46 states and DC.

Why do some kids develop AFM?

- Not a clear definitive link between virus strains and AFM. CDC working to understand the circulation of enteroviruses.
- Possible causes:
  - Direct infection of a virus on the motor neurons
  - Post-viral inflammatory or immune response directed toward motor neurons
  - Host genetic factors making certain children more susceptible than others

ACUTE INTERDISCIPLINARY MANAGEMENT
Medical Treatment

- No clinical trial data available to guide therapeutic recommendations
- Majority of patients receive IVIG, high-dose intravenous steroids, plasmapheresis and some received antiviral therapy
  - No significant clinical improvement or deterioration was noted with any of these therapies
- IVIG may be safest therapy available
- Fluoxetine is effective against EV-D68 and other enteroviruses
- Vigilant supportive care is central component
  - Mechanical ventilation
  - PT and OT to prevent atrophy and contractures and maximize functional outcomes.
  - Support for psychological as well as physical effects of disability

Acute Hospitalization

- No randomized controlled data, management based upon expert opinion
- Early aggressive rehabilitation to prevent muscular atrophy, joint contractures, and other sequelae of severe and persistent limb weakness and may improve functional outcomes
- 5% of AFM reported complete recovery of strength in acute phase and 18% reported being fully functional 4 months after onset

OT in Acute Care

- Prevention of atrophy, active exercise or FES
  - Recruitment of surrounding, functional muscle groups, even in cases with no objective improvement in strength
- Positional tolerance, ex: upright, head control
- Prevention of contracture or joint deformity
  - Splinting
  - Wheelchair fitting
- Promote engagement in meaningful activity, as appropriate
  - Leisure tasks
  - Bedside self-care
- Assist family with coping, educate on expectations
Rehabilitation vs. Habilitation

- Habilitation for the developing nervous system, guiding children to an anticipated potential.
- Attain function and health as we know it, but at a level unknown to them.
- Children don’t always understand the end goal as they have not completed the task before.
- Don’t have prior learning and skills to fall back on.
- Goals always changing.
- Easy for them to fall behind their peers.

Considerations for Rehabilitation Goals

- Age
  - Goals will change as the child develops to continue to address age appropriate needs.

- Lifelong process
  - Goals and rehabilitation programs will need to change over time as the person progresses and their priorities change.
  - Frequent updating prevents boredom, increases motivation, adapts to functional changes.

- Parents
  - How do you prioritize the parents’ goals versus the child’s goals?

Role for Occupational Therapy

- Compensatory Training
  - What do they need to do now?
  - What’s the most efficient way to do it?
- Environmental Modifications and Adaptations
  - How do I get them back to where they want to be?
  - How can I set them up for the most long term success?
- Rehabilitation Strategies for Recovery
  - How to optimize the nervous system for return of function?
  - How to maintain physical integrity while we wait?

Treat the Things You See

Impairments

- Decreased range of motion
- Decreased strength
- Decreased coordination
- Decreased fine motor skills
- Decreased sensation
- Increased / Decreased muscle tone
- Joint contracture
- Decreased endurance
- Decreased mobility
- Limited static/ dynamic balance
- Limited cognition
- Decreased vision/ visual disturbance

Activity Limitations and Participation Restrictions

- Decreased independence with self-care activities
- Decreased independence with home management tasks
- Decreased participation in school/ work/ leisure activities
- Decreased community access
- Increased caregiver burden
Compensatory Training

- Engagement in valued role-defining activities
- Intervention aimed at completion of context specific motor tasks in meaningful
- Training functional task rather than impairment
- Paired with feedback, repetitious, and goal directed
- Modifying task, person, environment

Areas of Occupation

- Strength based, family centered, occupation based approach
- Self-Care
  - Sitting balance to reduce caregiver burden
  - Rolling for in bed dressing
- School
  - Modified utensils and mobile arm support for reaching
  - Modified computer access
- Leisure
  - Voice activated electronics for social participation
  - Learn to provide caregiver direction for preferred activities

Environmental Modifications and Adaptations

- Adaptive equipment for self-care and school activities
- Home modifications
  - Ramps
  - Wheel-in shower
- Environmental Control Units and Smart Home devices, ex: Alexa
- Wheelchair access
  - Micro-light switches
  - Proportional joystick with alternative access (ex: chin, toe)

Rehabilitation Strategies for Recovery

- Provide repeated, near-normal input to the nervous system to optimize capacity for recovery
  - Robotics
  - Electrical Stimulation
- Offset chronic complications and maintain physical integrity
Activity-Based Rehabilitation

- Patterned activity intended to restore a task component, focus on impairment
- Use near normal kinematics and conditions
- Often high volume repetition
- Aimed at recovery of motor and sensory function
- Very young children have immature brain and spine
  – Show better recovery
  – Continue recovery over many years
- May have undamaged areas of the nervous system masked by immaturity

Best of Both Worlds

- Plan for near-term function while considering long-term impact, ex: Tenodesis
- Addition of an activity-based approach elevates occupation-based interventions to restorative
  – Add FES to assist with grasp during self-feeding with MAS for proximal assist
  – Prime nervous system with motor-assist UE cycling then use elbow flexion in tooth-brushing activity
- Intensive activity-based interventions are safe for all populations

Cases of AFM at KKI

<table>
<thead>
<tr>
<th>Demographics</th>
<th>Number</th>
<th>%</th>
</tr>
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<tbody>
<tr>
<td>Total number</td>
<td>31</td>
<td></td>
</tr>
<tr>
<td>Age Range</td>
<td>7 months – 18 years</td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>17</td>
<td>54.8</td>
</tr>
<tr>
<td>Female</td>
<td>14</td>
<td>45.2</td>
</tr>
<tr>
<td>Year of Diagnosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prior to 2014</td>
<td>14</td>
<td>45.1</td>
</tr>
<tr>
<td>2014</td>
<td>10</td>
<td>32.3</td>
</tr>
<tr>
<td>2016</td>
<td>7</td>
<td>22.6</td>
</tr>
<tr>
<td>Referral Source</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acute Care Hospital</td>
<td>10</td>
<td>32.3</td>
</tr>
<tr>
<td>Rehabilitation Facility</td>
<td>3</td>
<td>9.7</td>
</tr>
<tr>
<td>Admitted from Home</td>
<td>15</td>
<td>48.4</td>
</tr>
<tr>
<td>Location of Initial Rehabilitation Services</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Outpatient</td>
<td>18</td>
<td>54.8</td>
</tr>
<tr>
<td>Inpatient</td>
<td>12</td>
<td>38.7</td>
</tr>
</tbody>
</table>
Initial Presentation

Presenting Complaint

<table>
<thead>
<tr>
<th>Numbers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fever</td>
</tr>
<tr>
<td>URI</td>
</tr>
<tr>
<td>Gi</td>
</tr>
<tr>
<td>Monoplegia</td>
</tr>
</tbody>
</table>

MRI

- All 31 had anterior horn involvement;
- 5 of those had polyradicular extension; in 4 cases, there were additional findings c/w demyelination; 5 presented cervico-medullary junction involvement;

- All showed anterior horn involvement, abnormal or unobtainable late response F waves and/or reflexes, and normal sensory responses;

EMG

- All showed anterior horn involvement, abnormal or unobtainable late response F waves and/or reflexes, and normal sensory responses;

Etiology

- EV D-68 confirmed in 5, rhinovirus in 2, 1 HHV6, 1 Mycoplasma, 1 Epstein-Barr (IgG+, not IgM)

Wee-FIM Scores

<table>
<thead>
<tr>
<th>Admission</th>
<th>Discharge</th>
</tr>
</thead>
<tbody>
<tr>
<td>Self-Care</td>
<td>45.64</td>
</tr>
<tr>
<td>Mobility</td>
<td>40.73</td>
</tr>
<tr>
<td>Cognition</td>
<td>92.0</td>
</tr>
<tr>
<td>Total WeeFIM®</td>
<td>57.0</td>
</tr>
</tbody>
</table>

CASE STUDY

**JM**

<table>
<thead>
<tr>
<th>2016 Admit</th>
<th>2016 Discharge</th>
<th>2017</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head</td>
<td>None</td>
<td>Activities cervical ROM with support</td>
</tr>
<tr>
<td>Upper Extremity</td>
<td>No AROM</td>
<td>Gravity eliminated bilaterally in digits, no AROM otherwise</td>
</tr>
<tr>
<td>Lower Extremity</td>
<td>No AROM</td>
<td>Against gravity at knee and ankle, hip, hamstrings, and glutes (hip, gravity eliminated)</td>
</tr>
<tr>
<td>W/C</td>
<td>Dependent tilt-in-space</td>
<td>Proportional control power mobility using a foot joystick placed on left foot plate</td>
</tr>
<tr>
<td>Ability to eat</td>
<td>NPO</td>
<td>Chose to eat with family, tolerated small trials</td>
</tr>
<tr>
<td>PAMIS</td>
<td>24</td>
<td>38</td>
</tr>
</tbody>
</table>
Resources and Next Steps

- Centers for Disease Control
- Webinar on [www.occupationaltherapy.com](http://www.occupationaltherapy.com) in Spring
- [www.kennedykrieger.org/abrttraining](http://www.kennedykrieger.org/abrttraining)
- Refer to the Children and Youth Evidence Based Resources
  - [www.spinalcordrecovery.org](http://www.spinalcordrecovery.org)
  - [www.kennedykrieger.org](http://www.kennedykrieger.org)
- Start a conversation on AOTA’s forum CommunOT [https://communot.aota.org/home](https://communot.aota.org/home)
- See the AOTA Children and Youth practice resources including FAQs, fact sheets, occupation toolkit and more [http://www.aota.org/Practice/Children-Youth.aspx](http://www.aota.org/Practice/Children-Youth.aspx)
- Get involved in the Children and Youth Special Interest Section [http://www.aota.org/Practice/Manage/SIS.aspx](http://www.aota.org/Practice/Manage/SIS.aspx)
- Join an AOTA pediatric Community of Practice [http://www.aota.org/Practice/Children-Youth/PediatricWorkGroups.aspx](http://www.aota.org/Practice/Children-Youth/PediatricWorkGroups.aspx)
- Share the link to this recording and discuss with your colleagues [http://www.aota.org/Practice/Children-Youth/Early-Intervention/Resources.aspx](http://www.aota.org/Practice/Children-Youth/Early-Intervention/Resources.aspx)

References