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Acute Flaccid Myelitis: Overview of Treatment and Case Studies

(Part 2)
Kaitlin Hagen, MOT, OTR/L

Disclosures

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

At the conclusion of this webinar, participants will be able to:

- Define AFM and review current research
- Describe Activity-Based Rehabilitation (ABRT)
- Identify therapeutic techniques central to the practice of ABRT for AFM

What is Acute Flaccid Myelitis (AFM)?
Acute Flaccid Paralysis

- National surveillance for acute flaccid paralysis stopped in the USA after the eradication of Polio.
- California continued with passive surveillance and noted an increase in reports of acute flaccid paralysis in 2012 without an etiology. CA implemented increased surveillance.
- Canada and Europe continued with surveillance through the 1990s into the 2000s.

Acute Flaccid Myelitis Definition

Clinical Criteria: An illness with onset of acute flaccid limb weakness

Laboratory Criteria:
Confirmatory: a magnetic resonance image (MRI) showing spinal cord lesion largely restricted to gray matter and spanning one or more vertebral segments
Supportive: cerebrospinal fluid (CSF) with pleocytosis (white blood cell count >5 cells/mm3)

To confirm a case of AFM:
Confirmed:
Clinically compatible case AND Confirmatory laboratory evidence
Probable:
Clinically compatible case AND Supportive laboratory evidence
Acute Flaccid Myelitis Definition

- Spinal cord lesions may not be present on initial MRI.
- A negative or normal MRI performed within the first 72 hours after onset of limb weakness does not rule out AFM. MRI studies performed 72 hours or more after onset should also be reviewed if available.
- Terms in the spinal cord MRI report such as “affecting mostly gray matter,” “affecting the anterior horn or anterior horn cells,” “affecting the central cord,” “anterior myelitis,” or “poliomyelitis” would all be consistent with this terminology.
Presentation of AFM

- Patient’s with AFM range from presenting with monoplegia to tetraplegia on a vent (and everything in between)
- 5% acutely make full recovery
- 18% make a full recovery within 4 months
- We see the other 80% on rehab and will be focusing on those cases today
- To our knowledge there have been no relapses and it is not progressive

Ventilatory Support at KKI

- Extreme flaccidity, poor chest wall recoil, increased chest wall compliance, extremely prone to atelectasis.
- Out of 19 inpatient admissions (before 2018), 10 were requiring some degree of respiratory support on admission, and 9 were at discharge.
- 8 of the initial 10 had decreased levels of support by time of discharge.
- Looking closer at diaphragmatic pacers during the acute phase.
Ventilator Support at KKI

<table>
<thead>
<tr>
<th>Type of Respiratory Support</th>
<th>Admission #</th>
<th>Discharge #</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventilator 24 hours/day</td>
<td>7</td>
<td>4 (3 of these with decreased settings)</td>
</tr>
<tr>
<td>CPAP/BiPAP 24 hours/day</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Vent at night / PAP during day</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Vent at night / No support during day</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>PAP at night / No support during day</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>No support 24 hours/day</td>
<td>9</td>
<td>11</td>
</tr>
<tr>
<td>Total number with decreased respiratory support by time discharge</td>
<td></td>
<td>8 (80% of those with any respiratory support at time of admission).</td>
</tr>
</tbody>
</table>

What have we seen in 2018

- Nearly 100% of our cases this year have required pharmacologic intervention for hypertension
  - Why the increase? Possibly younger average age; seeing sooner in course; different patterns of viruses??
- Increasing numbers of pathologic fractures being noted (often present on arrival to our hospital, so not all therapy-related)
- Have started routine referral to endocrinologist, DEXA scan, labs to evaluate for pamidronate therapy.
Rehabilitation of AFM using Activity Based Restorative Therapy (ABRT)

<table>
<thead>
<tr>
<th>Habilitation</th>
<th>Plasticity</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Guiding children to an anticipated potential</td>
<td>• Very young children have immature brain and spine</td>
</tr>
<tr>
<td>• Attain function and health as we know it, but at a level unknown to them</td>
<td>• Show better recovery</td>
</tr>
<tr>
<td>• Children don’t always understand the end goal as they have not completed the task before</td>
<td>• Continue recovery over many years</td>
</tr>
<tr>
<td>• Don’t have prior learning and skills to fall back on</td>
<td>• May have undamaged areas of the nervous system masked by immaturity</td>
</tr>
<tr>
<td>• Goals always changing</td>
<td></td>
</tr>
<tr>
<td>• Easy for them to fall behind their peers</td>
<td></td>
</tr>
</tbody>
</table>
Why Activity?

- “Regular Physical Activity throughout life is important for maintaining a healthy body. Nevertheless, 60% of the global population fails to achieve the minimum physical activity recommendations.”

- “Persons with chronic physical condition are at greater risk due to inactivity than able-bodied persons because they are often restricted in performing normal everyday activity such as walking, housekeeping, gardening, shopping, and participating in sports.”


Activity Based Restorative Therapy (ABRT)

- 5 key components:
  - Functional Electrical Stimulation
  - Locomotor Training
  - Weight Bearing/Loading
  - Patterned Activity
  - Task-Specific Practice

- Plus
  - Aquatic Therapy
  - Vibration
  - Home based/community integration
ABRT in Very Young Children

- ABRT can be applied to children of all ages
- Key Factors:
  - Creativity
  - Family centered
  - Individualized treatment plans evolve as the child grows and achieves new milestones
    - There is more than one recipe for success
  - Ultimate goal should be increased independence in mobility and function.

Functional Electrical Stimulation

<table>
<thead>
<tr>
<th>TES</th>
<th>NMES</th>
<th>FES</th>
<th>TENS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Therapeutic Electrical</td>
<td>Neuromuscular Electrical Stimulation</td>
<td>Functional Electrical Stimulation</td>
<td>Transcutaneous Electrical Nerve Stimulation</td>
</tr>
<tr>
<td>Stimulation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Use of electricity to</td>
<td>Electricity applied across the surface</td>
<td>Application of electrical stimulus to a</td>
<td>Pain modulation by exciting peripheral</td>
</tr>
<tr>
<td>drive a desired nerve</td>
<td>of the skin over intact peripheral nerve</td>
<td>paralyzed nerve or muscle to restore or</td>
<td>nerves.</td>
</tr>
<tr>
<td>response for therapy.</td>
<td>evokes an action potential in the nerve</td>
<td>achieve function. Also refers to orthotic</td>
<td>Common Types:</td>
</tr>
<tr>
<td></td>
<td>fiber which causes an exchange of ions</td>
<td>substitution (Bioness L300).</td>
<td>- Sensory</td>
</tr>
<tr>
<td></td>
<td>to drive the muscle to contract.</td>
<td></td>
<td>- Motor</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>- Noxious</td>
</tr>
</tbody>
</table>
Benefits of FES

- Increased bone mineral density
- Reduction of tone
- Orthotic substitution
- Improving blood flow and muscle health
  - Improve and maintain muscle mass during or following periods of inactivity
  - Maintain/Increase ROM
  - Re-educate/facilitate voluntary contraction
  - Reduce effects of spasticity

Clinical Applications of FES

- Combine with functional activities using portable e-stim unit (EMPI, Intellect)
- Orthotic substitution (Bioness L300 & H200, Walk Aide)
- FES cycling (upper and lower extremities- RT 300, MotoMed)
- Biofeedback (Neuromove, Myotrack Infiniti, Otto bock STIWELL med4)
- Swiss stimulation or 1ms program
Locomotor Training

- An activity-based rehabilitative strategy designed to improve sensory, motor and autonomic function, health and quality of life

- Provides sensory cues to re-train neural patterns that will result in effective locomotion

- Emphasizes recovery of motor function using the intrinsic mechanisms of the nervous system, rather than compensatory strategies

Traditional Locomotor Training

4 Principles of LT:
1. Maximize weight bearing on the legs
2. Optimize sensory cues
3. Optimize kinematics for each motor task
4. Maximize recovery; minimize compensation

3 Components to LT:
1. Treadmill training
2. Overground training
3. Community training
Weight Bearing/Loading

- UE weight bearing can be achieved through:
  - Seated prop
  - Quadruped
  - Prone positioning

- LE weight bearing can be achieved through:
  - Quadruped or tall kneel
  - Standing:
    - With or without assistance
    - With or without bracing - No or minimal bracing preferred
  - Supported standing in standing frame
    - Static stander
    - Dynamic stander
    - Stander with glider component

Clinical Applications of Weight Bearing
Patterned Activity

- Repetitive task specific and non-task specific activities
- Promote cortical reorganization
  - In CIMT, benefits result from frequency of use of involved side, not constraint of uninvolved side
- Repeated multiple times for multiple hours/days
- Improve strength and ROM
- Perfect practice makes perfect
- Incorporate other components
  - Principles of LT
  - FES

Unweight the arm
Task Specific Practice

- Practice of context specific motor tasks
- Training functional task rather than impairment
- Paired with feedback
- Goal directed
- Repetition
- Incorporate other components
  - Stand at sink to brush teeth
  - FES to ankle dorsiflexion during gait
  - High repetitions of elbow flexion followed by self-feeding

Application of Task-Specific Training

- Relevant: meaningful to patient, context specific
- Random: facilitates retention, transfer, generalizability
- Repetitive: practice assists in skill mastery
- Reconstruction: breakdown the task to identify component weaknesses and areas for improvement
- Reinforced: timely and positive feedback
Don’t Let Bad Habits Persist

- Use it or lose it: Abhorrent patterns and compensatory strategies have to be overcome by rehabilitation
- Patients will figure out how to get things done (ex: tenodesis, one hand, toes, fingers)
- Cortical reorganization responds to non-use as much as therapy
- The body learns what we teach it
- Constraint Therapy

Vibration

- Side-alternating repetitive motion that simulates a (physiologically meaningful) movement pattern similar gait pattern
- Creates motor and neural learning effects by stimulating an involuntarily controlled “stretch-reflex” reaction that quickly and effectively improves existing residual functions and specific movement patterns
- Different settings help to decrease muscle tone or to help recruit nerves to fire
Aquatics

- ABRT principles can be applied in the aquatic environment to facilitate weaker muscle groups and provide resistance against stronger muscle groups.
- Properties of water utilized:
  - Buoyancy
  - Viscosity
  - Turbulence
  - Hydrostatic Pressure
  - Warmth
- Examples of ABRT and aquatics
  - Locomotor training & partial body weight supported gait training
  - High repetition of reciprocal, patterned movement
  - Standing

Secondary Complications: Shoulder Subluxation

- Bracing
  - Goal: to keep the hand free and usable
- Electrical Stimulation
- Kinesiotape or leukotape
- Weight bearing
- Wheelchair set up
Secondary Complications: Scoliosis

- Electrical Stimulation
- Bracing (but strengthen)
- Weight bearing
- Alignment in wheelchair

The distal return can make positioning difficult as patient’s can wiggle out of ideal positions, but not always get themselves back into them.

Secondary Complications Hip Subluxation/Dislocation

- Weight bearing!!
- Ensure good alignment during activities
- Wheelchair set up
Secondary Complications:
Fractures

- Be cautious of how you are moving the patient
- Our job to teach families to be careful, but to try not to scare them as it is important for them to keep their kids moving and active
- We know activity and weight bearing are important

Home Rehabilitation Program

- SCI/AFM requires a lifetime of care
- Incorporating the ABRT principles is key
- Prioritize the patient's impairments and functional limitations
- Consider building challenges into the environment

Common recommendations for equipment for home:
- Electrical stimulation unit
- Standing frame (mobile or glider)
- Wheelchair
- Bath equipment
Outcomes using ABRT on Inpatient Rehabilitation

- PT 2-3 hours a day
- OT 1-2 hours a day
- SLP up to 1 hour a day
- BP, NP, SW, CL, TR, Respiratory Therapy, Nursing, Medical Team
- The children reviewed came to inpatient acutely or within a year of diagnosis
- Most were admitted for 6-12 week admission dependent on medical and rehabilitation needs
- Outcome measures utilized if age appropriate

KKI Inpatient Rehab Program
### Physical Ability and Mobility Scale (PAMs)

*Also known as PAMS: scored 1-5*

1. Tolerance to positioning
2. Tolerance to sitting in chair
3. Tolerance to orthoses or splint
4. Support for seating system
5. Head control
6. Trunk control
7. Rolling supine to/from prone
8. Transitioning from supine to sit
9. Transitioning from sit to stand
10. Standing

---

### PAMs Continued

11. Transitioning from floor to stand
12. Environmental transfers
13. Transfers into and out of a car
14. Walking on level ground – assistive device
15. Walking on level ground – distance
16. Walking on level ground – level of assistance
17. Community skills
18. Wheelchair mobility
19. Standing balance
20. Stairs
### Outcome Measures- Admission to Discharge

<table>
<thead>
<tr>
<th>Outcome Measure</th>
<th>Admission M</th>
<th>SD</th>
<th>Discharge M</th>
<th>SD</th>
<th>t-test</th>
<th>d</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCIM (n=18)</td>
<td>31.1</td>
<td>26.4</td>
<td>51.3</td>
<td>29.0</td>
<td>-3.91***</td>
<td>-0.92</td>
<td></td>
</tr>
<tr>
<td>PAMS (n=27)</td>
<td>49.7</td>
<td>22.4</td>
<td>67.3</td>
<td>20.5</td>
<td>-7.21***</td>
<td>-1.39</td>
<td></td>
</tr>
<tr>
<td>WeeFIM® Self-Care DQ (n=29)</td>
<td>46.3</td>
<td>24.1</td>
<td>62.7</td>
<td>24.7</td>
<td>-5.54***</td>
<td>-1.03</td>
<td></td>
</tr>
<tr>
<td>WeeFIM® Mobility DQ (n=29)</td>
<td>39.7</td>
<td>26.9</td>
<td>57.0</td>
<td>23.6</td>
<td>-5.71***</td>
<td>-1.06</td>
<td></td>
</tr>
<tr>
<td>WeeFIM® Cognitive DQ (n=26)</td>
<td>85.3</td>
<td>36.5</td>
<td>97.6</td>
<td>54.0</td>
<td>-2.18*</td>
<td>-0.43</td>
<td></td>
</tr>
<tr>
<td>WeeFIM® TOTAL DQ (n=16)</td>
<td>54.0</td>
<td>24.3</td>
<td>70.1</td>
<td>25.8</td>
<td>11.87***</td>
<td>-1.54</td>
<td></td>
</tr>
</tbody>
</table>

*p<.05, **p<.01, ***p<.001
Table 3: Change from Inpatient Admission to Discharge on MMT by muscle group

<table>
<thead>
<tr>
<th>Muscle Group</th>
<th>Admission (n=27)</th>
<th>Discharge (n=29)</th>
<th>t-test</th>
<th>df</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ankle Flexion (n=27)</td>
<td>2.37 ± 1.50</td>
<td>2.66 ± 1.65</td>
<td>-2.46*</td>
<td>-0.47</td>
</tr>
<tr>
<td>Ankle Extension (n=27)</td>
<td>2.28 ± 1.53</td>
<td>2.68 ± 1.74</td>
<td>-2.90**</td>
<td>-0.56</td>
</tr>
<tr>
<td>Knee Flexion (n=29)</td>
<td>1.95 ± 1.60</td>
<td>2.30 ± 1.64</td>
<td>-4.10***</td>
<td>-0.76</td>
</tr>
<tr>
<td>Knee Extension (n=29)</td>
<td>1.90 ± 1.60</td>
<td>2.30 ± 1.60</td>
<td>-2.63*</td>
<td>-0.49</td>
</tr>
<tr>
<td>Hip Flexion (n=28)</td>
<td>1.90 ± 1.60</td>
<td>2.30 ± 1.50</td>
<td>-3.14**</td>
<td>-0.59</td>
</tr>
<tr>
<td>Hip Extension (n=28)</td>
<td>1.80 ± 1.53</td>
<td>2.13 ± 1.46</td>
<td>-3.17**</td>
<td>-0.60</td>
</tr>
<tr>
<td>Finger Flexion (n=28)</td>
<td>3.09 ± 1.70</td>
<td>3.41 ± 1.50</td>
<td>-3.17**</td>
<td>-0.60</td>
</tr>
<tr>
<td>Finger Extension (n=27)</td>
<td>3.06 ± 1.69</td>
<td>3.42 ± 1.40</td>
<td>-3.35**</td>
<td>-0.64</td>
</tr>
<tr>
<td>Elbow Flexion (n=28)</td>
<td>2.52 ± 1.69</td>
<td>2.81 ± 1.70</td>
<td>-4.53***</td>
<td>-0.86</td>
</tr>
<tr>
<td>Elbow Extension (n=28)</td>
<td>2.50 ± 1.72</td>
<td>2.85 ± 1.75</td>
<td>-3.76***</td>
<td>-0.71</td>
</tr>
<tr>
<td>Shoulder Flexion (n=29)</td>
<td>2.01 ± 1.56</td>
<td>2.40 ± 1.73</td>
<td>-3.24**</td>
<td>-0.60</td>
</tr>
<tr>
<td>Shoulder Extension (n=28)</td>
<td>2.10 ± 1.66</td>
<td>2.45 ± 1.65</td>
<td>-3.55***</td>
<td>-0.67</td>
</tr>
</tbody>
</table>

*p<.05, **p<.01, ***p<.001

**Upper Extremity Manual Muscle Testing**

![Upper Extremity Movement Tested](chart.png)
Ankle Flexion (n=27)  
Ankle Extension (n=27)  
Knee Flexion (n=29)  
Knee Extension (n=29)  
Hip Flexion (n=28)  
Hip Extension (n=28)

MMT Score

Lower Extremity Manual Muscle Testing

Lower Extremity Movement Tested

Case Study
### Case Study: JG

<table>
<thead>
<tr>
<th></th>
<th>2016 admit</th>
<th>2016 Discharge</th>
<th>2017</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Head Control</strong></td>
<td>None</td>
<td>Activates cervical ROM with support at occiput</td>
<td>Activates cervical ROM with support at occiput (stronger)</td>
</tr>
<tr>
<td><strong>Upper Extremity Status</strong></td>
<td>No AROM</td>
<td>Gravity eliminated bilaterally in digits, no AROM otherwise</td>
<td>Improved digit strength and emerging digit 1 twitches in right hand</td>
</tr>
<tr>
<td><strong>Lower Extremity Status</strong></td>
<td>No AROM</td>
<td>Against gravity at knee and ankle, twitch hamstrings and glutes</td>
<td>Against gravity at knee, ankle, hamstrings, and glutes (hip gravity eliminated)</td>
</tr>
<tr>
<td><strong>W/C</strong></td>
<td>Dependent tilt-in-space</td>
<td>Proportional control power mobility using a foot joystick placed on left foot plate</td>
<td>Proportional control power mobility using a foot joystick placed on left foot plate (community distance)</td>
</tr>
<tr>
<td><strong>Ability to eat</strong></td>
<td>NPO</td>
<td>Trials with therapy only</td>
<td>Cleared to eat with family, tolerates small trials</td>
</tr>
<tr>
<td><strong>PAMS</strong></td>
<td>24</td>
<td>38</td>
<td>44</td>
</tr>
</tbody>
</table>

### Head Control
Head Control Today?!

Digit Progression
Lower Extremity Progression

Sit to Stands
Independent Mobility: return to school

AFM Summary

- AFM is similar to polio in epidemiology, pathophysiology and presentation.
- Presentation can be variable with wide spectrum of impairments.
- Recovery possible but can be slow
- It can be good to take breaks from therapy and let the child be a kid.
AFM Summary

- There is limited knowledge regarding long term outcomes.
- It is important to maximize function and participate in ABRT for secondary health benefits.
- Strong coordination with outpatient plans for these kids to prevent chronic complications such as contractures, scoliosis, anxiety, and decreased community reintegration.

References:


Questions?

- Hagenk@kennedykrieger.org