Acute Flaccid Myelitis

Student Inservice: Spring 2019
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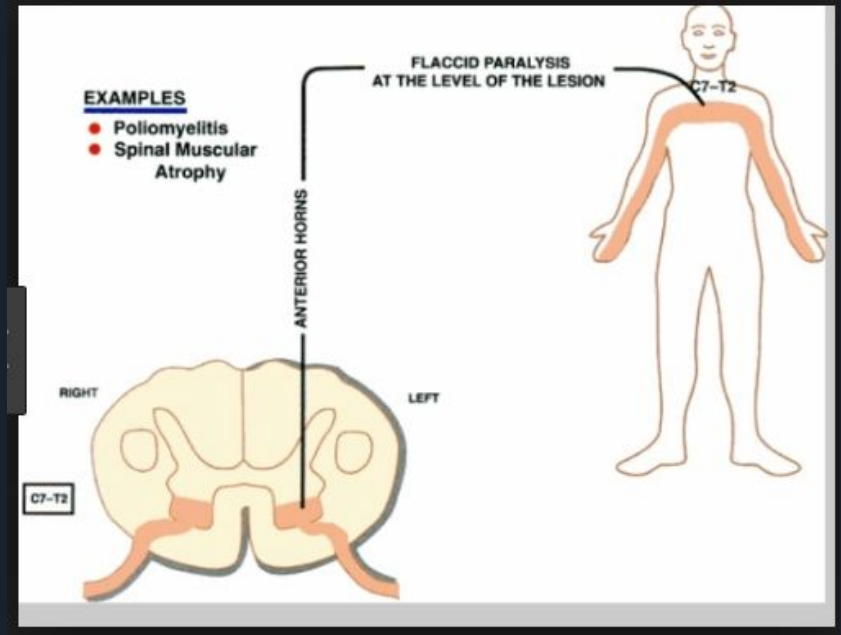
Objectives

By the end of this presentation the learner will be able to…

- Understand the physiological mechanism of Acute Flaccid Myelitis
- Explore studies to further analyze the presentation of children with AFM
- Establish general treatment parameters to utilize when managing a patient with Acute Flaccid Myelitis

What is Acute Flaccid Myelitis (AFM)?

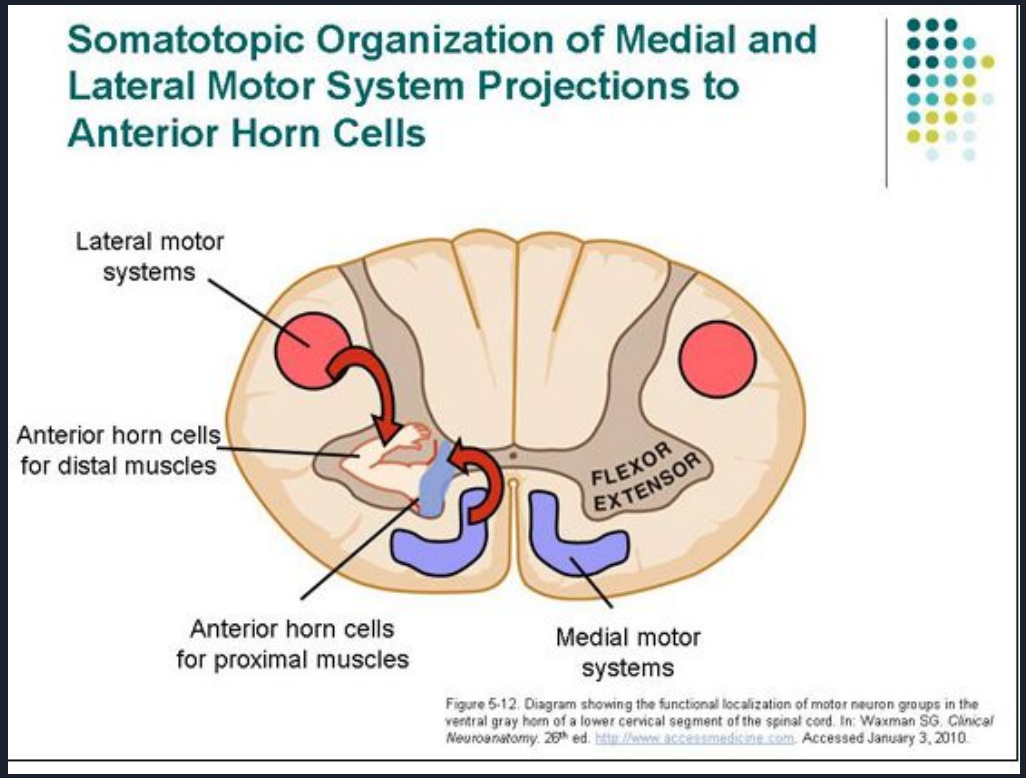
- Acute onset of flaccid limb weakness with spinal cord grey matter lesions, primarily of the ventral horn
  - Motor pathways terminate in ventral horn of spinal segments
- Characteristics: Asymmetric lower motor neuron deficits
  - Prominent motor deficits, limited sensory involvement, high probability of residual motor deficits[1,2]
- Causal relationship between enterovirus D68 infection and manifestation of AFM, however, AFM is a rare result of EV-D68 and several other pathogens have the capacity to cause the condition[1]



QUIZ QUESTION!

Which portion of the ventral horn houses tracts of proximal musculature?

A. Lateral
B. Medial



Medial motor systems (blue)[3]:

- Anterior corticospinal tract
- Vestibulospinal tract
- Reticulospinal tract
- Tectospinal tract

Lateral motor systems (red):

- Corticospinal
- Rubrospinal tracts

Characteristic	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Case status	Confirmed	Confirmed	Confirmed	Confirmed	Probable
Age at onset (yrs)	3.5	10	4	9	12
Sex	Boy	Girl	Girl	Girl	Girl
Onset of focal limb weakness	August 23, 2016	August 19, 2016	September 15, 2016	September 8, 2016	August 27, 2016
Onset of preceding respiratory or gastrointestinal illness	August 21, 2016 (respiratory)	August 14, 2016 (respiratory)	September 13, 2016 (respiratory)	September 6, 2016 (gastrointestinal)	August 17, 2016 (respiratory)
Presence of fever (tactile† or measured)	Yes	No	Yes	Yes	No
Limbs affected (region)	1 (LUE)	4	3 (BUE, RLE)	4	1 (LUE)
Cranial nerve features and timing	None	Facial droop subsequent to onset of limb weakness	Facial droop subsequent to onset of limb weakness	Facial droop before onset of limb weakness	Diplopia concurrent with limb weakness
Patient and family history of asthma	Asthma and family history of asthma	Asthma	None	Family history of asthma	Mild asthma, seasonal allergies, food allergies, eczema
Corticosteroid history	Maintenance inhaled fluticasone; oral budesonide for asthma exacerbation August 15–19	Maintenance inhaled fluticasone; oral prednisolone for asthma exacerbation beginning August 14	None	Oral prednisolone for treatment of Bell's palsy beginning September 5	Maintenance inhaled fluticasone
Magnetic resonance imaging (MRI) findings	T2 signal abnormalities in anterior and posterior columns of the central gray cervical cord	T2 signal abnormality with anterior and posterior involvement, contiguous through multiple levels of the cord	T2 signal abnormality in the anterior horn of the central gray cord	Anterior horn signal abnormality extending four cervical levels	Normal MRI result
Cerebrospinal fluid/white blood cell/mm^3	50	150	207	115	7
Nasopharyngeal swab polymerase chain reaction results from TGen	Positive for EV-D68	Positive for EV-D68	Positive for EV-D68	Unavailable	Unavailable
Stool specimen testing results	Negative enterovirus/ parechovirus by RT-PCR	Negative viral culture and enterovirus/ parechovirus by RT-PCR	Negative viral culture and enterovirus/ parechovirus by RT-PCR	Positive for coxsackievirus A10 by Sanger sequencing of the VP1 region	Unavailable

Iverson S, Ostdiek S, Prasai S et al. Cluster of acute flaccid myelitis in five pediatric patients. *MMWR*;2017: 66(28).

TABLE.
Selected Clinical Characteristics, Treatment, and Outcome of Children With Acute Flaccid Myelitis

Patient	Age	Presenting Sign(s)	MRI Abnormality*	Treatment	Outcome
1	14 y	R + L arm weakness	C1-T10	MP, IVIG, PLEX	Ambulatory; ventilator dependent
2	7 y	R facial droop and R arm weakness	Brainstem; C1-T8	MP, IVIG	Walks with AFO
3	9 y	L arm weakness	C2-C7; patchy T4-T7	MP	L shoulder weakness
4	9 y	L arm weakness	C1-C8	MP	Proximal L arm weakness
5	7 y	R arm weakness	Pons, medulla, C1-C8	IVIG	Proximal R arm weakness
6	11 y	R facial droop	Pons, medulla, C1-C8	IVIG, MP	Normal
7	13 m	R + L arm weakness (severe)	C2-T5	MP, IVIG	Minimal weakness L hand
8	7 y	R + L leg weakness	T6-T9	IVIG	Mild R hip weakness; neurogenic bladder
9	11 y	R + L leg weakness	T2-T4	IVIG	Walks with crutches
10	12 y	R + L leg weakness	Patchy T1-T12	IVIG	Requires walker for long distances; neurogenic bladder
11	21 m	Diffuse weakness	Subtle C1-C6	IVIG	No data

Abbreviations:
AFO = Ankle-foot orthosis
C = Cervical
IVIG = Intravenous immunoglobulin
L = Left
m = Months
MP = Methylprednisolone
PLEX = Plasma exchange
R = Right
T = Thoracic
y = Years

* All abnormalities consisted of T2 hyperintensities.



FIGURE 3.
Axial T2-weighted magnetic resonance imaging (Patient #3) demonstrates selective involvement of the gray matter of the spinal cord. Note the asymmetric involvement of the left anterior horn (arrow).



Nelson G, Bonkowsky J, Doll E et al. Recognition and management of acute flaccid myelitis in children. *Pediatr Neurol*. 2016; 55: 17-21.

Treatment parameters

- Minimal high quality evidence on suggested rehabilitation strategies or dosage
- General trends of impairments seen in AFM[1,2,4]:
  - Proximal weakness > distal weakness
  - UE > LE
  - Asymmetric > symmetric
  - Rarely sensory involvement
- Other anterior horn cell disorders: poliomyelitis, transverse myelitis, SMA

Recommendations for LMN disorders, regardless of cause[5,6]:

- Extensive baseline exam
- Flexibility: reduce contractures present secondary to weakness
- Aerobic: simple exercises- walking, stationary bike, swimming at submax levels
- Strength: maintain muscle strength, postpone loss of strength
  - A weak muscle can be damaged if overworked! It is already functioning close to maximal limits
  - Exercise using low (including body weight) to moderate intensity
- Consider energy conservation (intermittent rest)
- Be aware of signs of muscles being overworked (muscle cramping, fatigue, increased twitching)

- Those who have treated kids with AFM here at Gillette, what has been the biggest challenge or roadblock?
- Any successful interventions or management strategies you have discovered?

References

1. Messacar K, Asturias E, Hixon A et al. Enterovirus D68 and acute flaccid myelitis-evaluating the evidence for causality. *Lancet Infect Dis*. 2018;18:e239-47.
2. Nelson G, Bonkowsky J, Doll E et al. Recognition and management of acute flaccid myelitis in children. *Pediatr Neurol*. 2016; 55: 17-21.
3. Burke-Doe A. Motor Pathways. University of St Augustine. https://accessphysiotherapy.mhmedical.com/data/Multimedia/grandRounds/motorpathways/media/motorpathways_print.html
4. Iverson S, Ostdiek S, Prasai S et al. Cluster of acute flaccid myelitis in five pediatric patients. *MMWR*;2017: 66(28).
5. Smith M. Powerpoint presentation: Neuromuscular disorders. *Regis University.* Spring 2018.
6. Tiffreau V, Rapin A, Serafi R et al. Post polio syndrome and rehabilitation. *Ann Phys and Rehab Med*. 2010;53: 42-50.