Rehabilitation during the acute phase of Acute Flaccid Myelitis

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LEARNING OBJECTIVES

At the conclusion of this activity, the participant will be able to:

1. Recognize features of autonomic dysfunction in children with AFM
2. Identify potential interventions to maximize the health, participation and well being in this population

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DISCLOSURES

Presenter has no financial interest to disclose.

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ACUTE REHABILITATIVE CARE

- Pediatric Rehabilitation Medicine consultation for spinal cord injury care is recommended
- Early initiation of physical, occupational and speech therapy services to address issues related to positioning, progressive mobilization, splinting/bracing, communication, and swallowing is recommended
- Early initiation of electrical stimulation therapy may be helpful to reduce disuse muscular atrophy Augmentative communication devices and techniques might be needed for relaying pain and discomfort
- Children with bulbar involvement and inability to wean from the ventilator will require a tracheotomy and demonstrate stability on a home ventilator for transfer to inpatient rehabilitation.
- Psychological support should be initiated to assist the child and family through the process of coping and adjustment
- Medically stable children with significant residual neurologic deficit should transfer to an inpatient rehabilitation program

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FACTORS AFFECTING ACUTE REHABILITATIVE CARE

- Autonomic Nervous System Instability and dysfunction has been documented in the acute stage of AFM
- Cardio-vascular: wide blood pressure and heart rate variability
- Pulmonary: respiratory insufficiency/failure; troubles with secretion management
- Bladder: incontinence
- Bowel: constipation

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Case Presentations

AJ: 3 year and 11 months old girl at time of onset, otherwise healthy

Just started preschool

10/6 Symptoms of URI along with twin sibling

10/9 Neck and arm pain,

Parents note that not walking, needing to be carried

Progressed over course of the day

Sibling with same URI symptoms without developing weakness

Saw PCP, treated with amoxicillin for possible pneumonia, watch overnight

JH: 2 year and 9 months old boy at time of onset, otherwise healthy

9/30: URI symptoms

Saw PCP and treated for Pneumonia

10/4: Presented to ED for difficulty breathing and weakness of RUE

Midline back pain

Chest XR clear

EXAM: Flaccid RUE with only 1/5 in right deltoid

Normal appearing LUE and BLE

No trunk or respiratory involvement

Broad Differential, admitted to PICU

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Case Presentations

AJ

10/10 Presents to ED early AM and transferred University Hospital

EXAM: 0/5 BLE, 5/5 BUE, good head and neck control

Good respiratory and vocal control

Neck Pain with ROM

Neurology consulted

Immediate AFM or other neuromuscular disorder suspected

Admitted to PICU for close observation

JH

10/5 (HD2)

Progressive weakness

BUE > BLE

Increased respiratory effort leading to intubation

CSF studies (Pleocytosis)

Nasal swab positive for enterovirus

MRI completed

Involvement of entire spinal cord gray matter to T9

Involvement of pons and medulla

IV Steroids started (x5 days)

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Case Presentations

10/11 (HD2)

- Progressive weakness
  - Loss of BUE strength
  - Minimal head and trunk control
  - Decreased vocalization
  - Facial asymmetry
- Urinary Retention
- Rising CO2
- Intubated for impending respiratory failure

10/10 (HD7)

- Persistent Weakness
  - Flaccid arms
  - Trace movement in BLE
- PLEX x4 followed by IVIG x2

10/11 Diagnosis of AFM by infectious disease

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Case Presentations

10/11 (HD2)

- CSF studies (Pleocytosis)
- Nasal swab positive for enterovirus
- MRI completed
  - Involvement of entire spinal cord gray matter
  - Involvement of pons and medulla
- IVIG started (Daily x5)
- PM&R Consult

ADDITIONAL COMPLICATIONS

- Anxiety
  - Present throughout, mild prior to admission
  - Psychiatry Consulted
  - Treated with sedation early, then Seroquel and fluoxetine
- Lip Smacking and Tongue Thrusting
  - Negative seizure work up
  - Improved over 2 weeks
  - Attributed to dysautonomia
- Pain in RUE
  - Some improvement with Gabapentin

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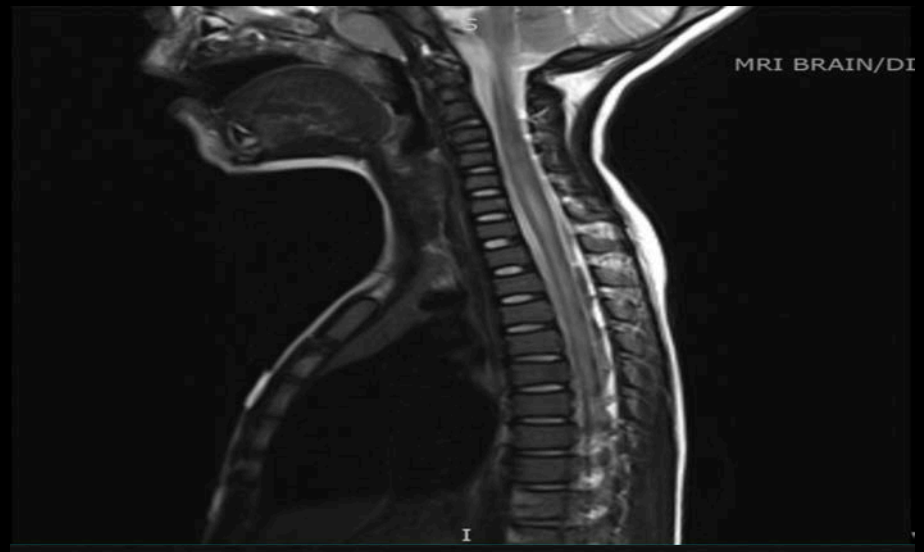

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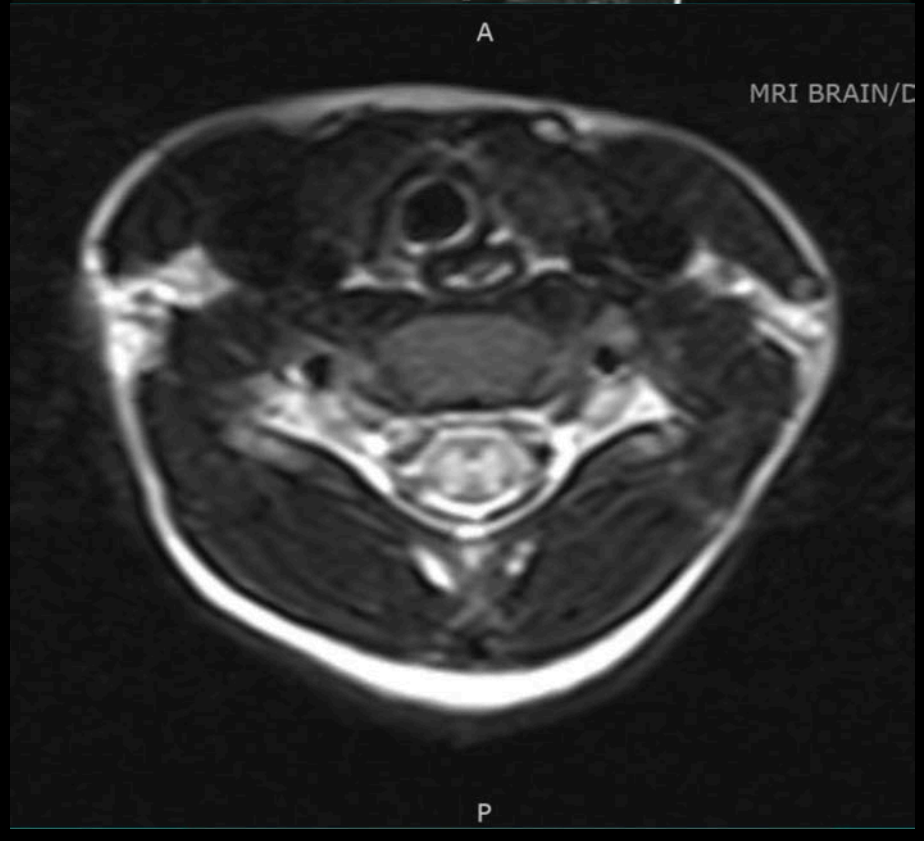

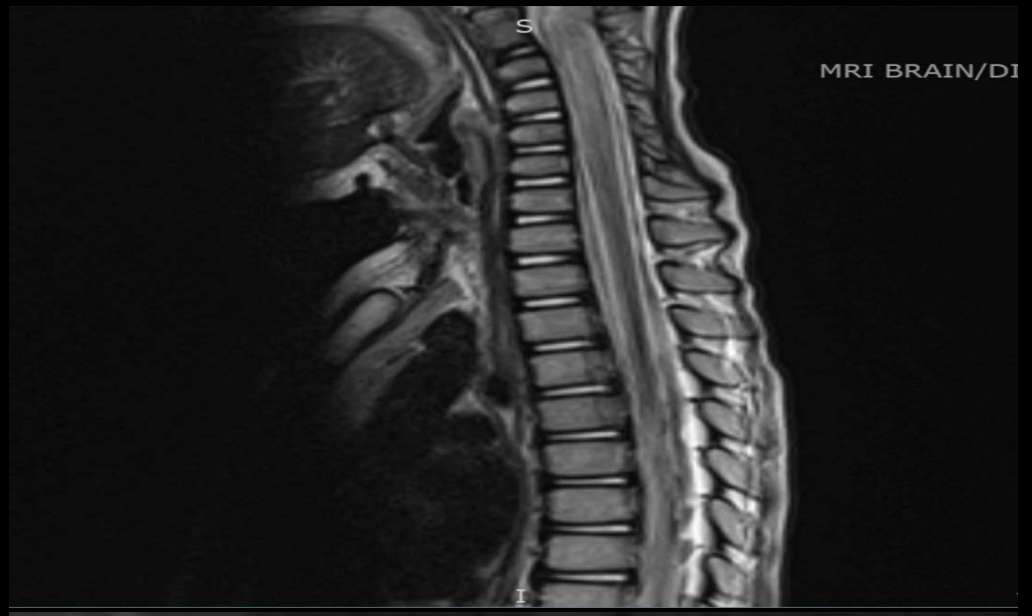

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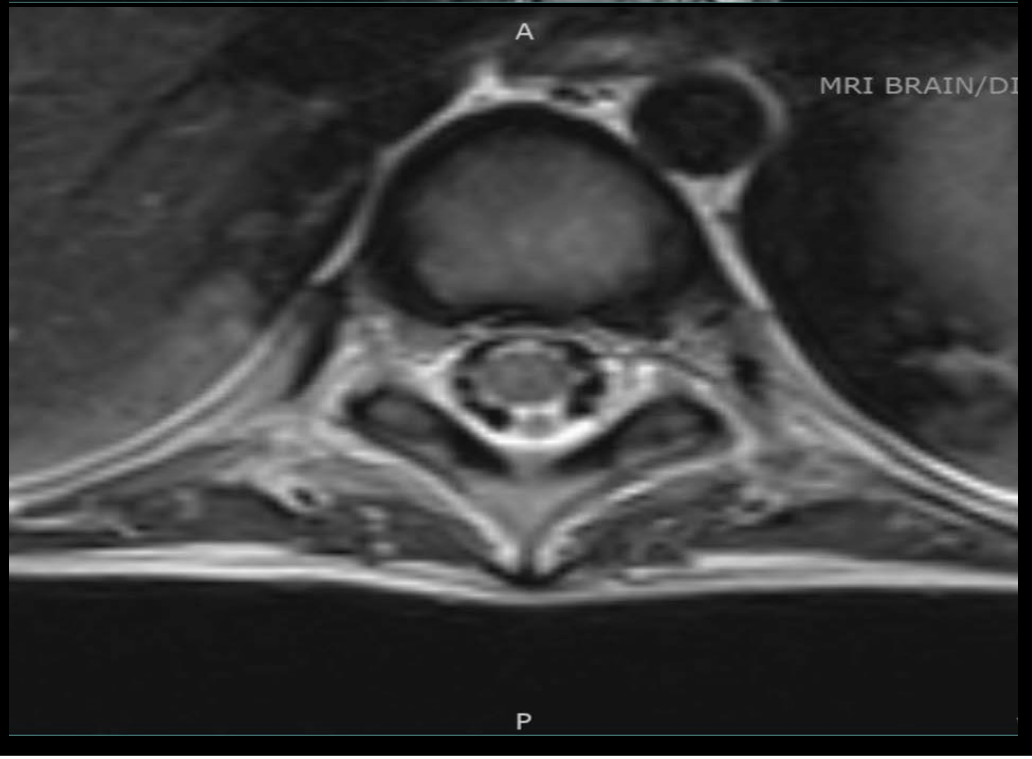

Case Presentations

Over the next couple of weeks

Minimal improvement of strength

Autonomic instability

EMG / NCS completed (HD 7)

Diaphragmatic Pacer trial (HD20)

Admit to Rehab Unit (11/19 HD 47)

EXAM: Antigravity of RLE at hip, else minimal to no activation

AG of LLE at ankle/foot, 1/5 KE, else no activation

Flaccid RUE, LUE with 1/5 with finger flexion

Improving urinary retention

Pain improved with increased gabapentin (face and arms)

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Case Presentations

Both AJ and JH: Autonomic Involvement Noted HD2

- Cardiac rhythm dysregulation
- Blood pressure dysregulation
- Temperature dysregulation

+ Bowel and Bladder retention

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Case Presentations

CARDIAC RYTHM DYSREGULATION

Tachycardia *alternating with*

Junctional rhythm events with severe brady cardia

Usually related to stress

Junctional Rhythm

Cardiology Consult, not dangerous rhythm

PICU education and plan

Instability improved over time

Use of sedation weaned (midazolam and fentanyl)

CARDIAC RHYTHM DYSREGULATION

Sinus Tachycardia up to 160s With PVCs

PE noted 10/11, heparin drip started

Tachycardia persisted, requiring pharmacologic intervention

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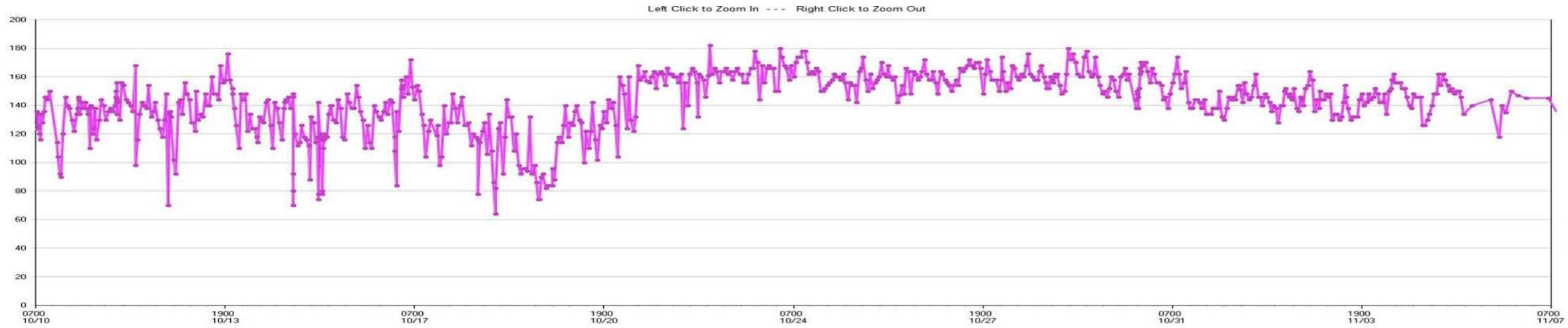
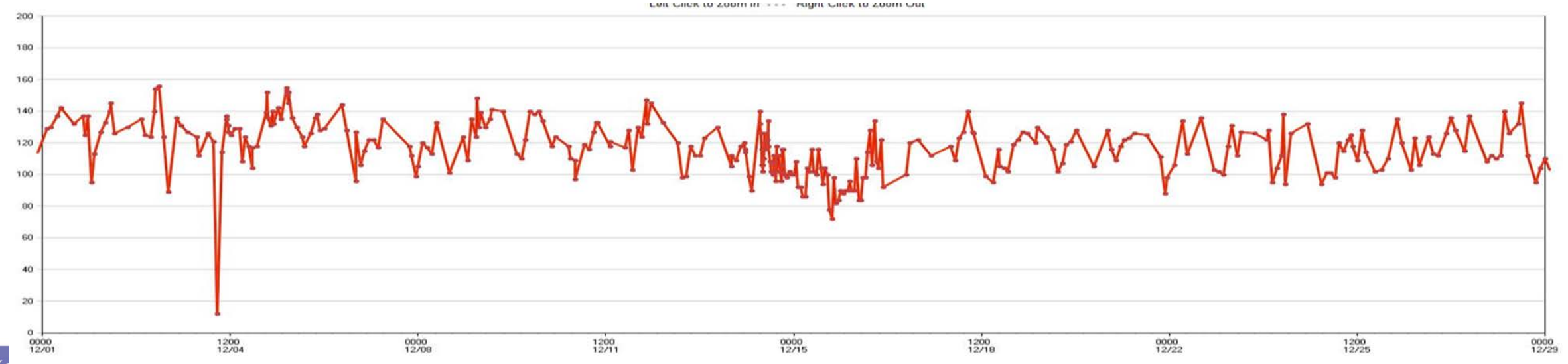
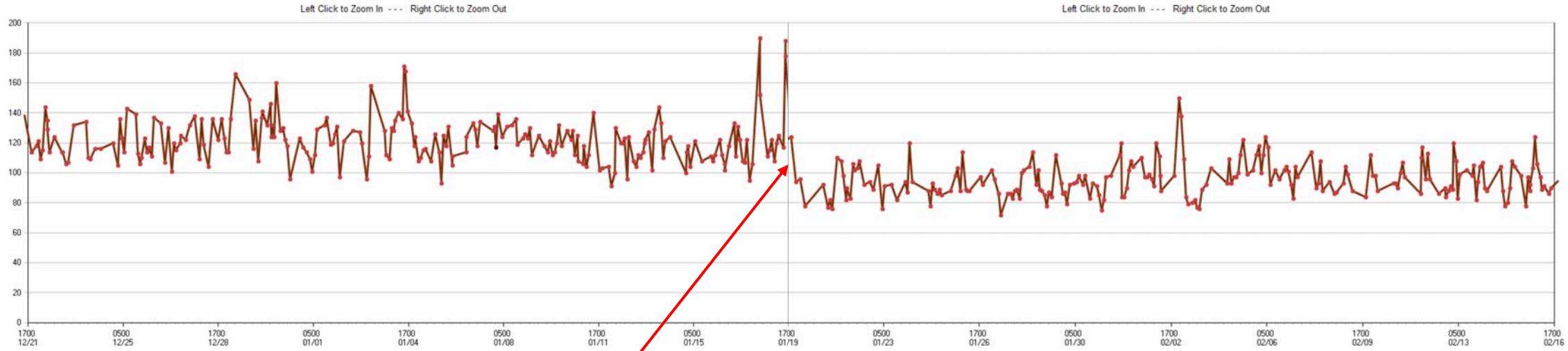
AJ: Heart Rate over first 30 days of admission



Heart Rate Day 60+



JH: Heart Rate Day 60+



Clonidine Patch

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Case Presentations

Hypertension

Worsened slowly over first 2 weeks, then plateaued

Persistent and preventing stepdown/transfer to Rehab Unit

PICU discussion with PM&R (likely autonomic dysregulation)

- Propranolol trial (HTN associated with tachycardia) (clearance with cardiology); well tolerated, improved parameters

- Propranolol weaned after transfer to Rehab Unit

Hypertension
Initially slowly worsened over course
Hypertension up to SBP of 200

Negative renal work up
Developed HTN associated nephropathy
Initially nicardipine then moved to Clonidine, Enalapril
By 10/13 (HD10) fluctuating HTN
Propranolol not effective
Improved later (possibly) with Clonidine patch
Worst events treated with sedation
Hyperhidrosis during events

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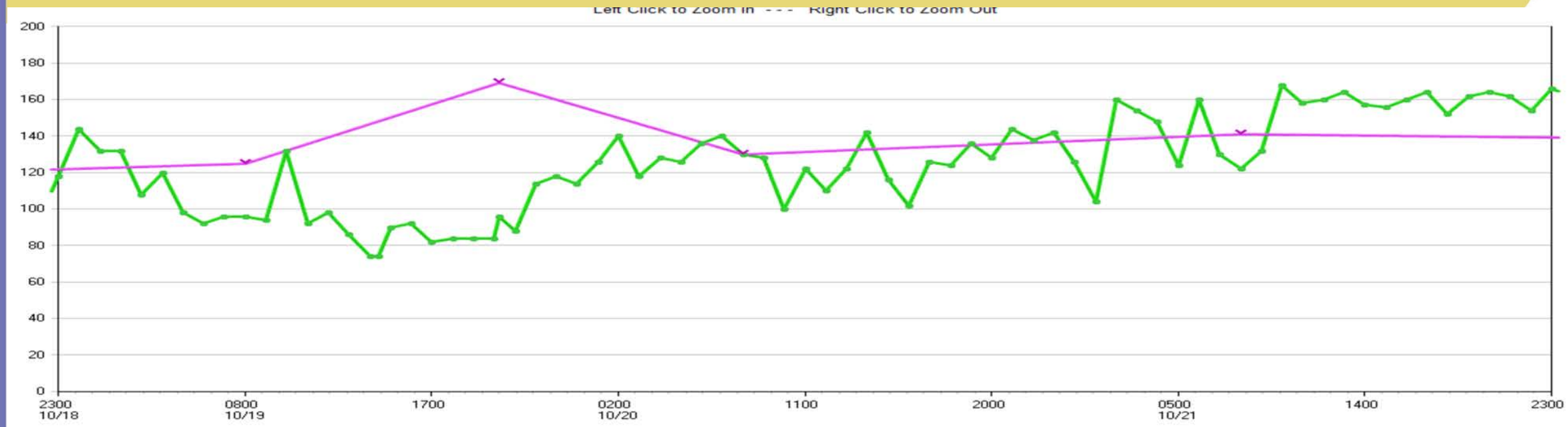
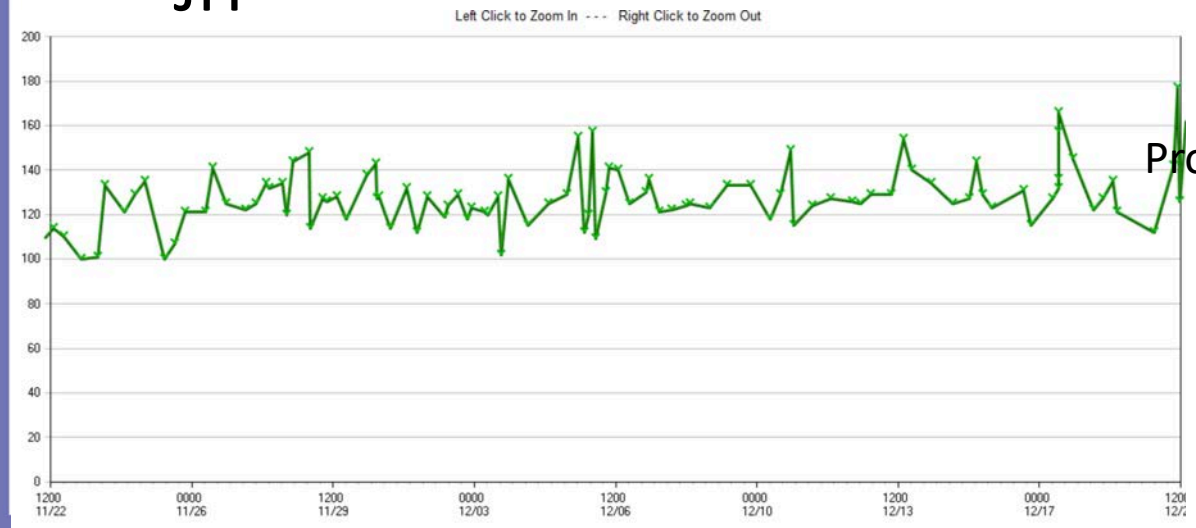
Hypertension

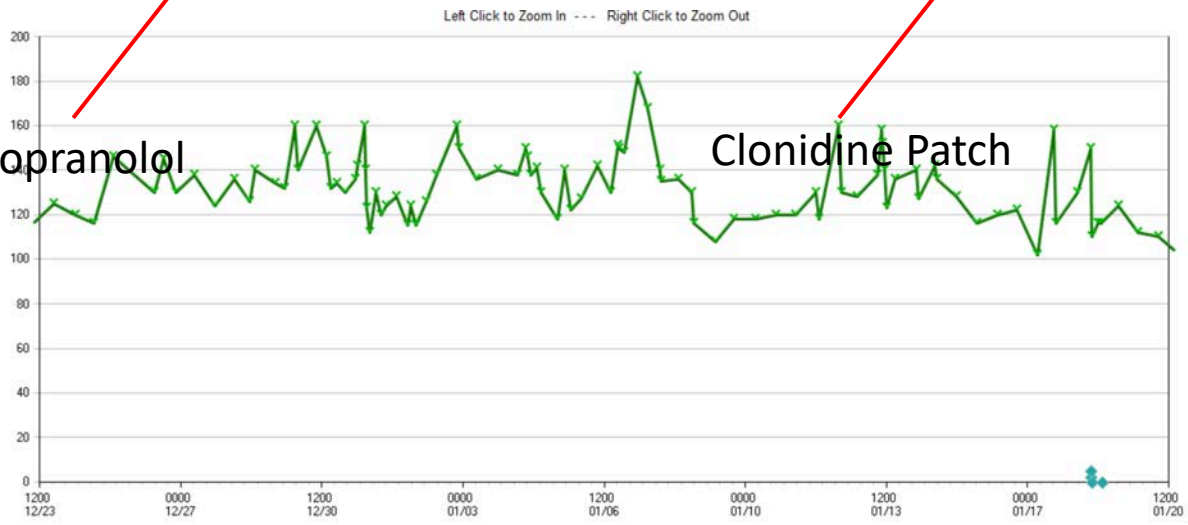
AJ



JH

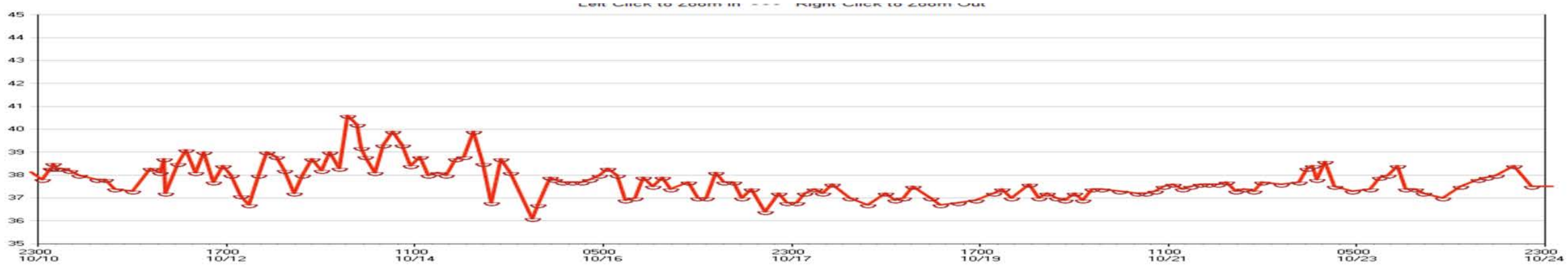


Propranolol



Clonidine Patch

Temperature



Elevated Temperature despite multiple antibiotics

Normalized HD7

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Case Presentations

AJ's Case and takeaways

Present on admission

- Bradycardia
  - Cardiology consult for junctional rhythm
  - Education to PICU
  - Fewer events over time
  - Improved control allowed for sedation wean and improved communication
- Tachycardia
  - Persisted for weeks
  - Appeared to respond to Propranolol (caution with bradycardia)
- Temperature Dysregulation
  - Infectious work up negative
  - Resolved relatively quickly

Not present at admission

- Hypertension
  - Presented second week
  - Slowly increased
  - Persistent for weeks
  - Appears to have responded (modestly) to propranolol

JH's Case and takeaways

Present on admission

- Tachycardia

- PE identified

- AD or similar autonomic disruption
- PVC

- Fewer events over time
  - Persisted for many weeks (longer than AJ)
  - Did not seem to correlate with anxiety or pain

Responded to clonidine patch

Unclear if responded to propranolol

- Temperature Dysregulation

Despite antibiotic coverage from admit

Resolved relatively quickly

Not present at admission

- Hypertension

Presented in first week

Slowly increased

Persistent for MANY weeks

Appears to have responded to Clonidine Patch

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Case Presentations

SUMMARY

- Autonomic Dysregulation is present in AFM
- Tachycardia / Bradycardia
  - Seems to present very early with weakness
  - Persists for weeks, but appears to improve
  - Investigation of alternative causes critical
- Hypertension
  - Seems to present later after weakness progresses
  - Persists for weeks, but appears to improve
  - Multiple agents may need to be trialed

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Case Presentations

SUMMARY

- Temperature Dysregulation
  - Seems to present very early with onset of weakness
  - Persists despite negative infectious work up or treatment
  - Appears to be self-limited after weakness
- Orthostasis
  - Not observed

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AFM and Respiratory Failure

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Ventilatory Support – Literature

Small case series:

California, Dec 2015 – 2/20 still on vent after 1 year

Argentina, 2017 -2/6 still on ventilator at 6 months after diagnosis

Seattle, Jan 2017 – 1/10 patients still on ventilator after 6 months

Scotland, Nov 2018 – 2/5 on ventilator after 18 months

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Respiratory Concerns in AFM

- Increased chest wall compliance
- Impaired ability to produce effective coughing, leading to the subsequent accumulation of secretions.
- Increased production of secretions secondary to autonomic dysfunction.
- Ventilator Induced Diaphragmatic Dysfunction; diaphragmatic atrophy increases over time on vent.
- Goals: prevention atelectasis, management secretions, and activation of diaphragm

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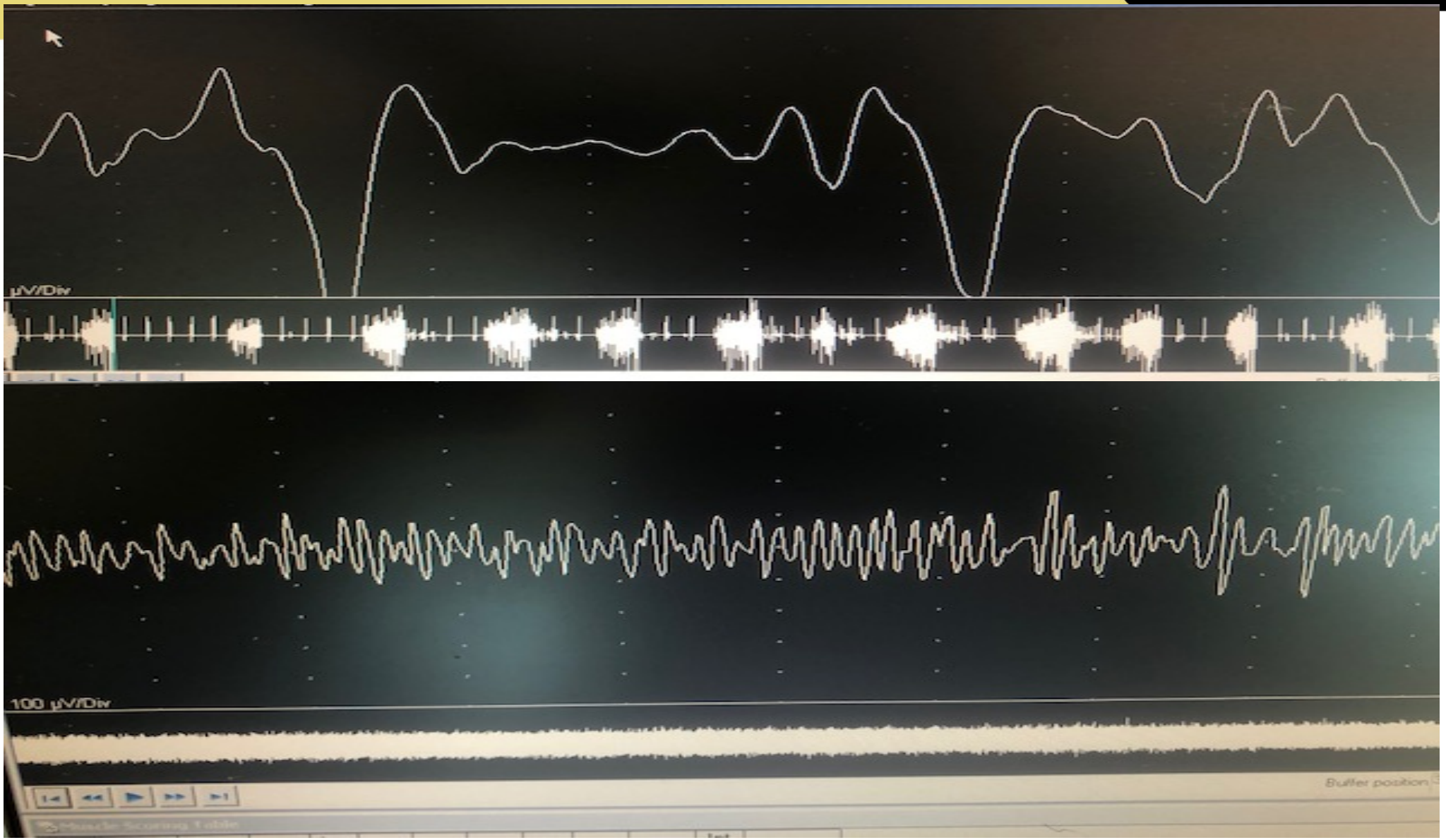
Diaphragmatic Pacers

- Phrenic nerve pacing in kids since the 1970's.
- 2011, Onders et al. published first intramuscular diaphragmatic pacers in children: laparoscopic mapping to find points of maximum contraction with implantation 4 percutaneous intramuscular electrodes. Very successful in traumatic SCI patients.

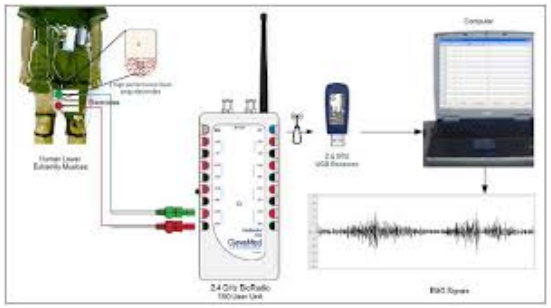
*Require intact phrenic nerve/innervated diaphragm.

	AJ	JH
Age	4	3
Paralysis onset	10/10/2018	10/05/2018
Respiratory failure onset	10/12/2018	10/05/2018
EMG	No phrenic nerve activity on surface NCS pre-DPS implant	N/A
Diaphragm US	had 1 cm excursion on left hemidiaphragm but only 1 mm on right	diaphragm thickening and excursion with and without assisted ventilation on left/none on right
DPS insertion date	10/30/2018 (day 20)	03/01/2019 (day 155)
Intra-op	Min diaphragm contraction on right with twitch stim, adequate contraction on left with twitch stim	Min diaphragm contraction on the right with train stim, adequate contraction on the left with twitch stim
Post DPS insertion evaluation time	1 day – 4 months	2 weeks - 7 months



Edmiston T, Elrick M, Kovler M, Jelin E, Onders R, Sadowsky C. Early Use of an Implantable Diaphragm Pacing Stimulator for a Child with Severe Acute Flaccid Myelitis – A Case Report. Spinal Cord Series and Cases July 2019.

Continuous EMG recording with CleveMed device



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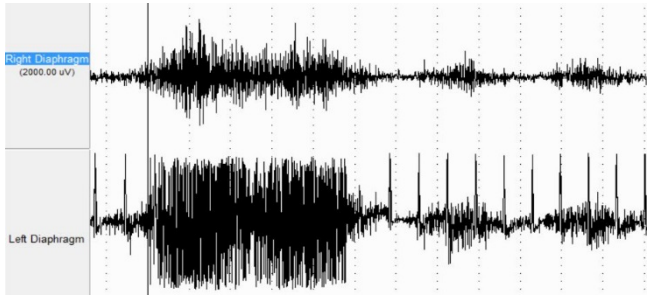
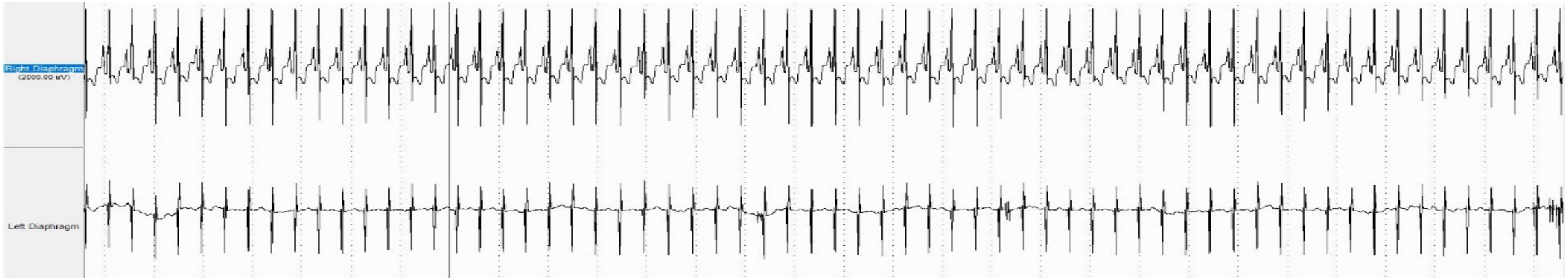
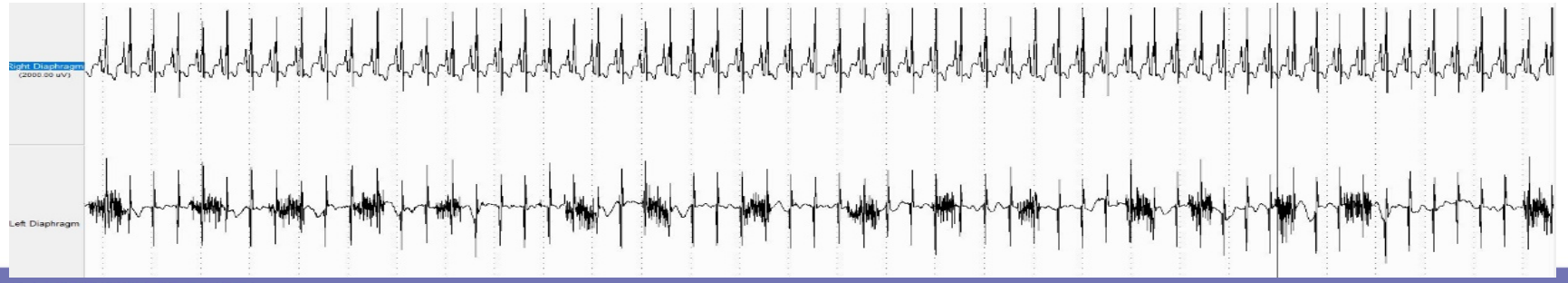


On ventilator – set respiratory rate: suppression of EMG activity



On ventilator – no set respiratory rate: no active EMG on the right diaphragm and diminished, but voluntary activity on the left.



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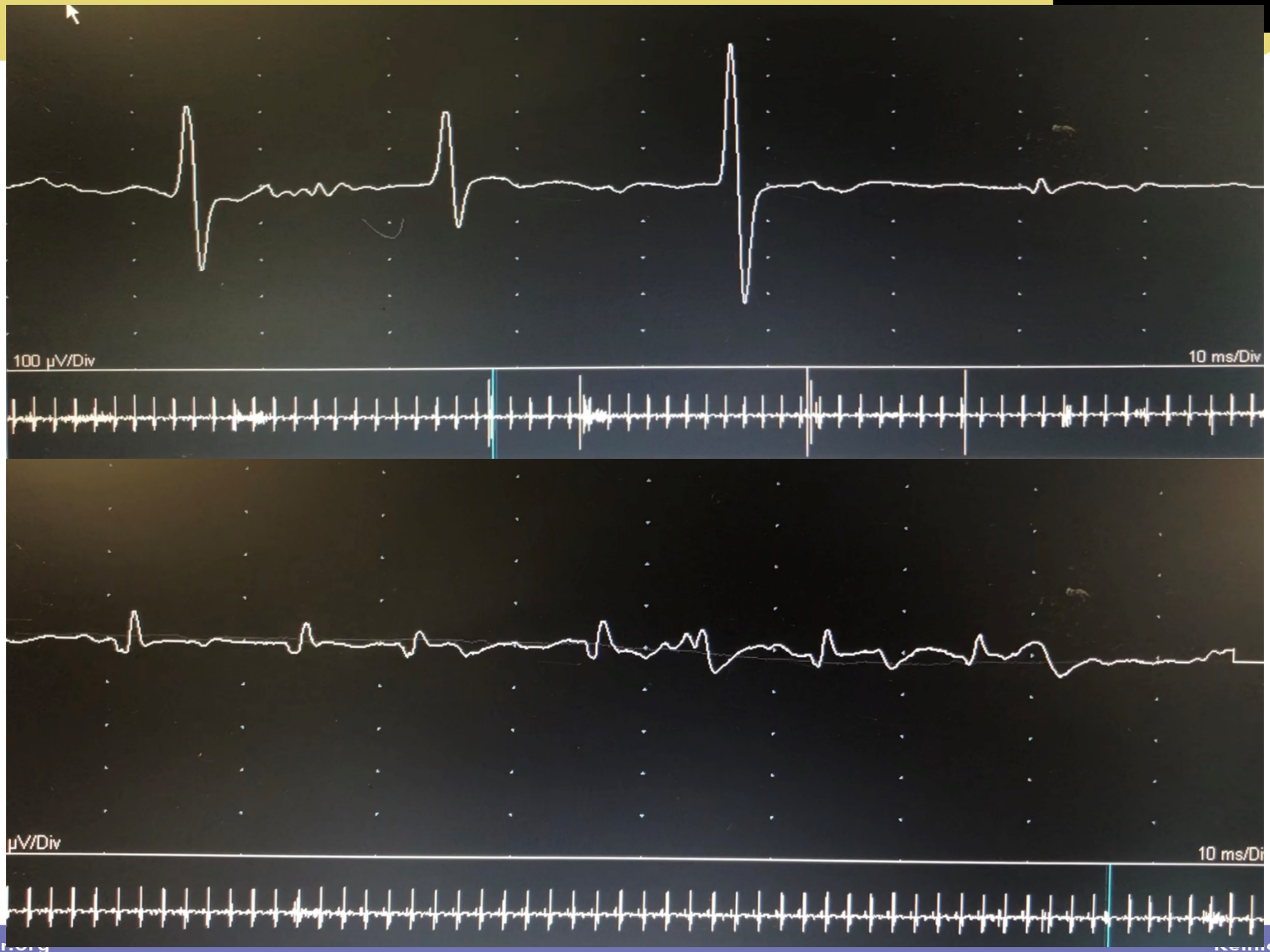
Time Relative to Pacer Placement	Ventilator Setting (Pacing Off)	CPAP/PS (Pacing On)	Pacing Duration (Per Day)	Tidal Volume Information	Additional Info
Prior to DPS	SIMV Volume 140 mL, RR 32 PEEP 5, PS 10 30% FiO2	---	---	Ventilator Set **140** mL	No spontaneous respiration
DPS +1 Day	SIMV Volume 155 mL, RR 25 PEEP 5, PS 10 30% FiO2	PEEP 5, PS 16 30% FiO2	30 min x1	---	Spontaneous RR 18 (Paced)
DPS +8 Weeks	SIMV Volume 155 mL, RR 22 PEEP 6, PS 14 21% FiO2	PEEP 6, PS 14 Paced RR 18	6 hrs x1	CPAP/PS +DPS **120-200 mL** PS + DPS (time to desat <90%) PS 10: **200-240** mL (stopped after 5 minutes) PS 5: **70-105** mL (2 minutes) PS 0: **40-60 mL** (48 seconds)	Breathing over DPS rate of 18 with total RR 25-28
DPS +12 Weeks (Discharged home)	SIMV Volume 155 mL, RR 22 PEEP 6, PS 10 21% FiO2	PEEP 6, PS 14 Paced RR 18	6 hrs x1	---	Breathing over DPS rate of 18 with total RR of 21-23
DPS +19 Weeks	SIMV Volume 155 mL, RR 22 PEEP 6, PS 10 21% FiO2	PEEP 6, PS 14 Paced RR 18	8-12 hrs x1	CPAP/PS alone **100-180** mL CPAP/PS + DPS **170-230** mL	Breathing over DPS rate of 18 with total RR of 30


100 µV/Div
10 ms/Div
µV/Div
10 ms/Di

	03/15/2019	07/24/2019	08/23/2019	10/04/2019
DPS settings RR 18 Inspiration interval 1.5 Pulse frequency 14 Pulse Ramp 10	1: 10 mA, 100µsec (L) 2: 10 mA, 100µsec (L) 3: 10 mA, 100µsec (R) 4**: 5 mA,** 60µsec (R)	1: 10 mA, 100µsec (L) 2: 10 mA, 100µsec (L) 3: 10 mA, 100µsec (R) 4: **10 mA,** 80µsec (R)	1: 17 mA, 100µsec (L) 2: 17 mA, 100µsec (L) 3: 17 mA, 100µsec (R) 4: **17 mA,** 100µsec (R)	1: 23 mA, 100µsec (L) 2: 23 mA, 100µsec (L) 3: 23 mA, 100µsec (R) 4: 23 mA, 110µsec (R)
Ventilator setting	No respiratory rate, FiO2: 21 %, PEEP 6, PS 14	No respiratory rate, FiO2: 21 %, PEEP 6, PS 14	No respiratory rate, FiO2: 21 %, PEEP 6, PS 12	No respiratory rate, FiO2: 21 %, PEEP 6, PS 8
Tidal Volume on ventilator only	TV 60-220 ml	TV 90-140	TV 70-120	TV 60-140
Tidal Volume on ventilator and DPS (measurements taken at different times (rest/therapy)	TV 90-225	TV 148-220	TV 90-165	TV 60-170

Weaning protocol

I. Ventilator FULL support only (set respiratory rate + PS + PEEP)

- Gradually increase time on DPS and full vent support until patient can pace 8-12 hours per day.

II. Ventilator support AND DPS (DPS + PS + PEEP + set ventilator respiratory rate)

- Gradually decrease ventilator rate during time on DPS until the patient reaches a rate of 6; then DC.

III. Pressure Support Ventilation AND DPS (DPS + PS + PEEP)

- Gradually decrease PS while on DPS until level of 5cmH_2O.
- Gradually decrease PEEP until level of 5cmH_2O.

IV. DPS only (no ventilator support) during waking hours.

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THANK YOU

- We have been fortunate to be part of the rehabilitative care of dozens of families with AFM

”We” is actually a very large team!

- Nurses, MD’s
- PT, OT, SLP, TR
- Social workers, Case mangers

Specific Thank You to:
Janet Dean
Carlos Pardo
Matt Elrick
Michelle Melicosta
Kofi Anoh
Eric Jellin
Travis Edmiston
The JHH PICU team

- And, most of all, Our Kiddos and their Parents

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