AFM

RESPIRATORY CARE AND REHABILITATION

Respiratory Failure in AFM

In early series of AFM cases (2014-16), apx 25% of patients required ventilatory support at some point during their illness; in 2018-2019 reviews this rose to 40-65%.

Overall, estimate 10-20% will have a persistent requirement for support:

Facebook parent group survey (pub Pediatric Neurology, V102, Jan 2020) - 15% at 24 months.

California, Dec 2015 – 10% still on vent after 1 year

Argentina, 2017 – 33% still on ventilator at 6 months after diagnosis

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

Scotland, Nov 2018 – 40% on ventilator after 18 months

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Respiratory Issues in AFM

- While UMN SCIs demonstrate decreased chest wall compliance, AFM results in **extreme flaccidity** with increased chest wall compliance, poor chest wall recoil, and heightened tendency to develop atelectasis.
- Impaired ability to produce effective coughing, leading to the subsequent accumulation of secretions.
- Increased production of secretions secondary to autonomic dysfunction.
- VIDD = Ventilator Induced Diaphragmatic Dysfunction, caused by diaphragmatic atrophy which increases with time spent on ventilator. Appears to occur rapidly in AFM (c/w clinically observed muscle atrophy).

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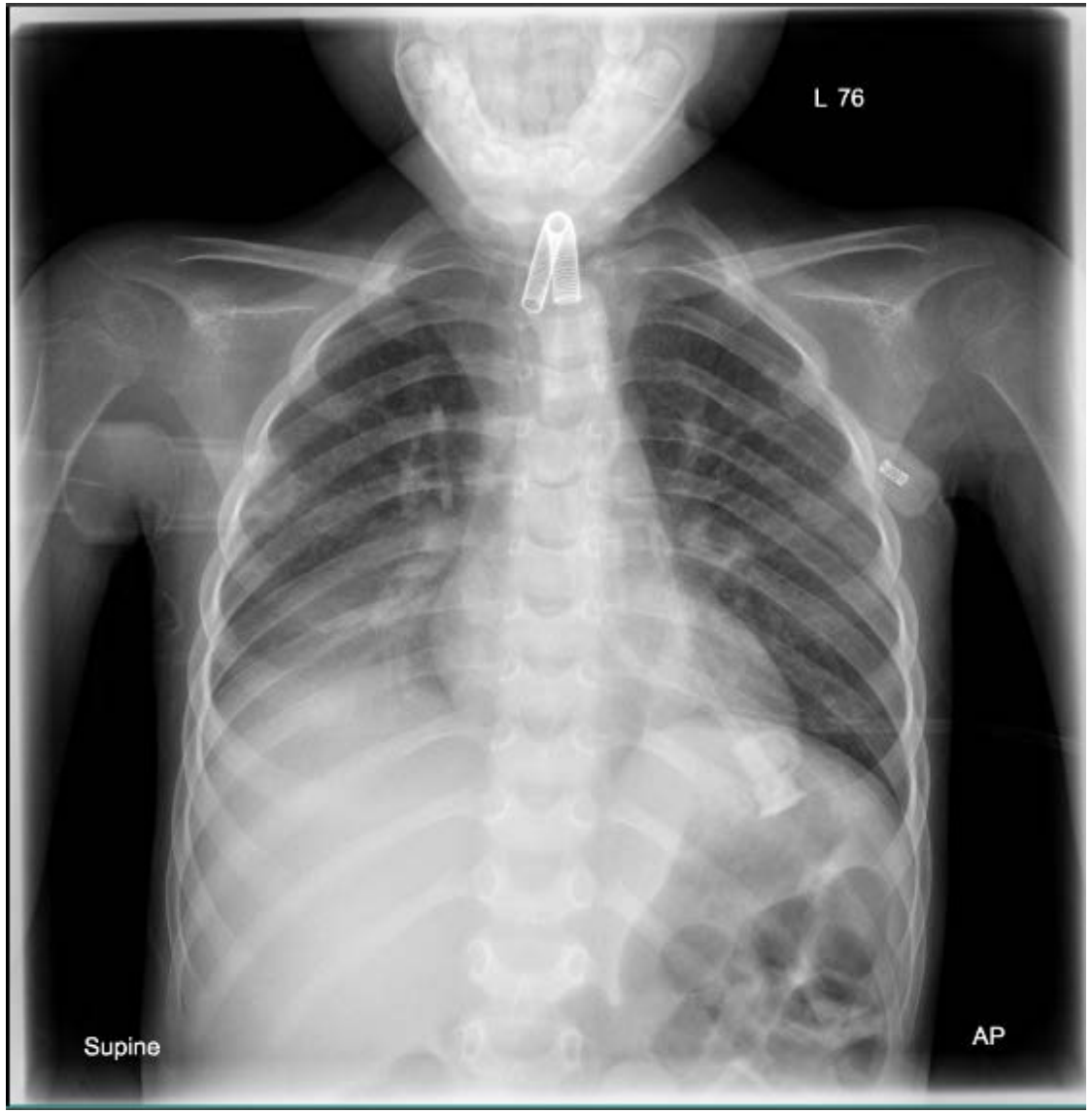
3 yr old with AFM, CXR at presentation to KKI.



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Respiratory Interventions

Goals: prevention of atelectasis, management of secretions, and activation of diaphragm

- Respiratory muscle training (i.e., incentive spirometry, decreased PS, sprints off vent). NIF of -20 vs -30.

- SIMV: Preserves remaining strength respiratory musculature; decreases risk hyperventilation and barotrauma; and facilitates weaning. Pressure-Control vs Volume-Control: traditionally 6-8ml/kg tV. In AFM we see increased atelectasis at these volumes, often require 10ml/kg +

- DPS has been shown to help recover phrenic nerve function and maintain diaphragm muscle mass especially when begun early.

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RESPIRATORY CARE/ REHAB TECHNIQUES

3 main techniques to consider simultaneously

- Mechanical ventilation
- Tracheostomy hygiene
- Bronchial hygiene

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Mechanical Ventilation

Optimal ventilator settings (Nighttime or Full support settings)

Volume/Pressure

- Volume ventilation preferred (10ml/kg +)

Optimal PEEP (5-8cmH_2O)

- Least amount of PEEP to obtain adequate lung expansion and oxygen saturation with minimum FiO_2

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Mechanical Ventilation

Conditioning/Weaning (Daytime settings)

- Diaphragmatic Pacing AND Conditioning
- Lower respiratory rate
- Ideal PSV; adequate ΔP – monitor Vt
- Sprints off vent Follow $EtCO_2$/breathing over vent; then check NIFs

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Tracheostomy Hygiene

Appropriate tracheostomy size; Cuffless/Cuffed

- Nighttime cuff inflation (MOV)
- Lung inflation and alveoli recruitment
- Rest/recuperation

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Tracheostomy Hygiene

Infection prevention

- Tracheostomy care; Daily/PRN (minimum)
- Tracheostomy changes; Bi-weekly (minimum)
- Oral care; BID (minimum)
- Suctioning; Inline catheter (sterile technique)

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Bronchial Hygiene

Goals;

- Secretion mobilization/excretion
- Functional alveoli recruitment

Bronchial Hygiene

- ❑Respiratory treatments;
  - Aerosols , MDIs (Bronchodilators, Mucolytics, Corticosteroids)
- ❑CPT; secretion mobilization
  - Vest therapy, Manual percussion, Pneumatic percussion
- ❑Cough assist/ oscillation; lung inflation, secretion mobilization and removal

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Bronchial Hygiene

- ❑ Intrapulmonary percussion:- lung inflation and secretion mobilization
  - IPV/Metaneb

- ❑ Airway clearance; secretion removal
  - suctioning

Secretion management / Prevention of atelectasis

Aggressive Airway Clearance:

Intrapulmonary Percussive Ventilation (IPV)

- clinical peds trials show more improvement in atelectasis than traditional CPT

- we have used in AFM vent-dependent patients since 2016 (with bronchodilators); better tolerated in older children

Inhaled Steroids - systemic inflammation associated with chronic SCI may contribute to reduced pulmonary function

Bronchodilators – Possibly increase mucociliary clearance

Cough-Assist (mechanical insufflation/ exsufflation)

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Ventilatory Support – The KKI Experience

In a review of 19 inpatient admissions:

- 10 were requiring some degree of respiratory support on admission (full ventilator or CPAP/BiPAP)
- All received respiratory rehabilitation as described above
- 8 patients were able to decrease the amount of support they required by discharge (ie, vent to PAP; weaning daytime support; or decreased vent settings)
- 2 of these eliminated need for respiratory support entirely.

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