FMC Sleep and Respiration Rounds

November 1, 2017

Karen Rimmer
Contribution of diaphragmatic paralysis to respiratory failure

SLEEP AND RESPIRATION ROUNDS
NOVEMBER 1 2017
Disclosures

- **Financial**: None relevant to this talk
- **Other**: I AM NOT A SLEEP DOC!
Objectives

- Structure and innervation of the diaphragm
- Uncertainty and/or variability in respiratory muscle use in sleep
- Causes of diaphragm dysfunction/paralysis
- Symptoms associated with diaphragm paralysis
- Workup of suspected diaphragm paralysis
- How does diaphragm weakness cause respiratory failure?
- Management options for respiratory failure in the setting of diaphragm paralysis
- Introduce the concept of diaphragm pacing and phrenic nerve surgeries.
Hard to find these cases? Nope!

Yesterday in clinic:
- 2 central cord syndromes
  - One on treatment with normal tCO2
  - One with ABG 7.41/56/53 untreated
- One idiopathic bilateral phrenic neuropathy
  - Happens to be one of the cases in for follow up
CASE ONE

- 61 year old man - previously healthy
- August 2015, driving and shoulder checked and felt right neck pain
- Subsequently noticed some dyspnea which progressively worsened over 1 year until presentation
- MRC 3 dyspnea
- Can’t lay flat on his back → SOB. Sleeping in a chair.
- No clear history to suggest SDB
- Patient referred for dyspnea
Exam

- Sitting - normal RR without accessory muscle use
- Supine – RR 24 with frank abdominal paradox
- Clinically reduced lung volumes
- BMI 29
CXR in inspiration
<table>
<thead>
<tr>
<th>Units</th>
<th>Predicted (Favor Mean)</th>
<th>Sitting</th>
<th>Sitting</th>
<th>Supine</th>
<th>Supine</th>
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Other Testing

- ABG was normal (including a normal HCO3)
- SNIP -21 cm H2O
The Diaphragm
Innervation of the diaphragm
How does it work?
Physiology of abdominal paradox
Other muscles
Symptoms and signs

- Dyspnea - postural “tell”
- ± Symptoms of SDB

- Community folks:
  - May not be the typical OSA pt
  - History of orthopnea
  - Paradox when observed
### Causes of Diaphragmatic Paralysis

<table>
<thead>
<tr>
<th>Neurologic causes</th>
<th>Myopathic causes</th>
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<tbody>
<tr>
<td>Spinal cord transection</td>
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<tr>
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<td>Postviral phrenic neuropathy</td>
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<tr>
<td>Radiation therapy</td>
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<tr>
<td>Cervical chiropractic manipulation</td>
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Note: Copyrights apply.
Confirmation -not all elevated diaphragm are paralyzed

- Symptoms
- PFTs – what is a significant drop in VC when supine?
- SNIP, MIP, MEP
- Imaging
- ENG/EMG – In this case
  - Lt: abnormal insertional activity with spontaneous fibrillations. No voluntary activity recorded
  - Rt: No abnormal insertional activity or activity with voluntary activity
Wakefulness

- Behavioral - avoid the supine posture
- Recruitment or increased activity of other inspiratory muscles - sometimes VERY obvious
- IF no additional load on the respiratory muscles such as obesity or underlying lung pathology, respiratory failure may or may not occur

Remember this case has a VC of 65%
Breathing in sleep???

- Minute ventilation in sleep - decreases
- Postural muscles activity in REM - inhibited
- Diaphragm activity in NREM and REM - maintained or increased
- Intercostals in REM
  - Parasternal - Maintained (though likely reduced)-animal data 2016
  - External intercostals- Less info
  - Scalenae - less info
- Accessory muscles in REM
  - SCM - Often absent
  - Abdominal - usually not used in expiration
  - Other accessory muscles - little known
Case one

- SOB
- Can’t sleep in bed
- Not in respiratory failure
- Neurology opinion:
  - Likely idiopathic bilateral phrenic neuropathy
So……

- What should I do with this man?
- What will happen if I do nothing?
  - Will he go into respiratory failure given time?
Evidence? What happens in sleep in patients with diaphragm paralysis?

- Conflicting evidence
- Why?
  - Methods of recording EMG
  - Methods of diagnosis of paralysis
  - Finding may not be consistent between individuals
For example: REM sleep

- Again conflicting
  - Reduced REM (and Stage N3) VS
  - Normal total REM (some suggest increase phasic REM)

- Presence of hypoventilation, but not in all especially if unilateral only

Not surprising then: Some report pts in respiratory failure, but some don’t
Respiratory muscles activity during REM sleep in patients with diaphragm paralysis
J R Bennet 2004 Neurology

- 5 Pt and 4 controls
- Symptom duration - mean 19 months
- Mean VC 52% predicted
- BMI mean 26
- PSG with EMG of SCM, parasternal, Pec minor, Rectus abdominus
Results

- AHI Mean 7.2, REM AHI 10.2
- Nadir saturation: 85% (lowest recorded pt 71%)
- SCM all pts in REM
- Parasternals 3/5 pts in REM
- Pec minor 1/5
- Rectus abdominus 4/5 in NREM and 2/5 in REM In expiration
- Pt with highest REM AHI did NOT recruit parasternals
Our Case one...

- He was not happy sleeping in a chair
- He wanted help with breathing so he could get back to bed and his “nagging but loving wife”

- No sleep study done
- Supine titration performed
- Follow up at 2 months
  - Back in bed
  - tCO2 remains normal. Level 3 on Rx - no events, no desats
  - Less SOB in the day
  - Happy patient!
One year follow up...
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**Spirometry**

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CASE two

- 75 year old man
- History of trauma (hit head on bottom of pool AND also hit in the face with a golf club)
- Known Left hemi diaphragm elevation since at least 2011
- 10 year history of mild dyspnea
- April 2017: swimming in deep pool and “couldn’t breath”
- Subsequent severe orthopnea - sleeping over a table
- Admitted to SHC and extensively worked up causes of dyspnea
EXAM

- Sitting – RR 20 and subtle use of accessory muscles
- Frank paradox and will not lay down for more than 15 seconds
- Saturation 88%
CXR 2011
Old to the left
New on the right
# Workup

<table>
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<tr>
<th>Arterial Blood Gas</th>
<th>pH</th>
<th>PaCO2</th>
<th>PaO2</th>
<th>HCO3</th>
<th>FiO2</th>
<th>BE</th>
<th>Hgb</th>
<th>HbO2</th>
<th>COHb</th>
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<td>mmHg</td>
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<td>g/dl</td>
<td>%</td>
<td>%</td>
<td>%</td>
<td>mmHg</td>
<td>°C</td>
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|           |       |           |           | Sitting Reported | Sitting % Predicted | Supine Reported | Supine % Predicted |%
| FVC       | L,btps | 4.12      | 3.21 - 5.04 | 2.38 < | 58 < | 0.99 < | 24 < | -58
| FEV1      | L,btps | 2.98      | 2.21 - 3.75 | 1.43 < | 48 < | 0.61 < | 20 < | -57
| FEV1/FVC  | %      | 73        | 63 - 82    | 60 <   | 83    | 62 <   | 85    | 2
| FEF25-75% | L/s    | 2.16      | > 0.60     | 0.59 < | 27 < | 0.30 < | 14 < | -49
| FEF25%    | L/s    | 7.25      | 7.25       | 2.48   | 34 < | 0.59 | 8 < | -76
| FEF50%    | L/s    | 3.76      | 3.76       | 0.83   | 22 < | 0.34 | 9 < | -59
| FEF75%    | L/s    | 1.25      | 1.25       | 0.25   | 20 < | 0.20 | 16 < | -20
| FEFmax    | L/s    | 7.68      | > 5.41     | 6.41   | 83   | 2.79 < | 36 < | -56
| FIVC      | L,btps | 2.40      |           |        |       | 0.90  |       | -63
| FIFmax    | L/s    | 3.64      |           |        |       | 1.97  |       | -46
What to do?

- This man – I’m more worried about.
- HCO3 already up
- Hypoxic without other cause found

- Decided to treat with NIV
- Again, supine clinic titration
- Given marked restriction also started LVR
- Follow up and Level 3 planned
CASE three

- 47 year woman well until MVC in Nov 2015
- Severe left brachial plexus injury with loss of function of left arm
- Pain post injury requiring narcotics
- Nerve transfers Feb 2016 - Left phrenic used to reinnervate the left arm
- Dyspnea following surgery with 2 presentations with hypercapneic respiratory failure
- Ongoing dyspnea and assessed in pulmonary clinic
EXAM

- Obese
- Hypoxic
- No accessory muscle use
- No abdominal paradox
PFTs and ABG (note BMI 33)

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<td>FEV1/FVC</td>
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<td>71 - 91</td>
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<td>FEF25%</td>
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Yikes

- Very worried
- Already presented to hospital twice
- Hypercapneic and hypoxic

- Didn’t feel comfortable with empiric therapy given more than one cause of resp failure
- PSG booked - didn’t attend?????
Sleep disordered breathing in unilateral diaphragm paralysis or severe weakness
J Steier 2008 ERJ

- 11 of 39 screened participated/ 11 controls
- Questionnaires, cap gas, PFTs, PSG with EMGs

Note: probable bias given these were referred and more likely to be symptomatic
Results

- 9/11 preferred to sleep good side down
- 8/11 had abdominal paradox
- FVC 74% predicted
- 2/11 had no SDB (not completely paralysed)
- AHI in NREM 8.1/ REM 26
- QOL significantly reduced
- Parasternals active in BOTH pts and controls
- Neck muscles active in pts in REM
So....

- In both Uni and bilateral paralysis some get into trouble and some don’t!!??

- Factors
  - Obesity - increased load
  - Comorbid illness
    - Lung disease - increased load
    - Narcotics/sedatives - decreased drive
  - Neuroplasticity / Neural reorganization
Who needs a PSG?

- You could argue, if symptomatic, just empirically titrated supine and then follow up with LEVEL 3
- If you want to answer the question of who will get into trouble... PSG will definitely help
  - Degree of hypoventilation
  - Muscles compensation if assessed
Questionnaire to predict SDB in n-m disease: ERJ 2011, 32(7): 400 (Moxham’s group)

- Reasoned that PSG may not always be readily available
- Many N-M pts don’t have typical symptoms (ESS often normal with documented SDB)
- Developed a questionnaire to predict SDB in n-m diseases (respiratory muscle weakness)
- Score of $\geq 5$ was predictive
- Discriminates normal and known pts with OSA
**SiNQ-5 (SDB in NM disease questionnaire)**

Dear Patient,

The following questions may help us to decide whether you may have disordered breathing during sleep related to muscle weakness. Please circle the most appropriate answer to each question.

Thank you for your cooperation.

<table>
<thead>
<tr>
<th>Question</th>
<th>Yes (2)</th>
<th>Sometimes (1)</th>
<th>No (0)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Do you feel breathless, if you lie down? (e.g. on your bed)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Do you feel breathless, if you bend forward? (e.g. to tie your shoelaces)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Do you feel breathless, if you swim in water or lay in a bath?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Have you changed your position when in bed?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Have you noticed a change in your sleep (waking more, getting up, poor quality sleep)?</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Numbers in parentheses represent scores.
CASE four

- 57 year old farmer
- FSHD with minimal functional impairment
- Head trauma while working 2008 and 2011
- July 2012 fall down stairs with C4 fracture
- Functionally C2 and failure to wean off ventilation
- Consulted with the question of chronic ventilation
Question in my mind…to ventilate or to pace?
Advantages of pacing over chronic vent

- Distribution to dependent lung (post/basal) is better
- Speech is better
- Reduced noise
- Sense of smell preserved
- Cosmetic
- Mobility
- Reduced costs of care
Evaluation for possible pacing

- Not weanable
- MUST HAVE A FUNCTIONAL PHRENIC
Phrenic stimulation
Direct Diaphragmatic Stimulation

- Laparoscopic approach
- Intraoperative muscle stimulation
- Mapping of diaphragm
  - Points of maximal contraction
  - Qualitative (direct visualization)
  - Quantitative (change in abdominal pressure)
- Once sites identified
  - Two electrodes placed in each diaphragmatic leaflet
  - Wires brought out and tunneled to the outside
  - Direct observation of pacemaker test

Figure 1 - Laparoscopic implantation of an intra-diaphragm phrenic stimulation electrode in the right hemidiaphragm.

Figure 2 - Laparoscopic view of an intra-diaphragm phrenic stimulation electrode successfully implanted in the left hemidiaphragm.