6th International Workshop on Breathing Pacemakers

Indications and Preoperative Evaluation of Patients

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Objectives

- Discuss neurologic control of respiration
- Review phrenic nerve physiology
- Explain diagnostic testing in diaphragm weakness
Motor control

• Generally a two neuron process
  – Cortical upper motor neuron to anterior horn cell
  – Anterior horn cell to muscle

• Interruption anywhere along the pathway can cause weakness
Respiratory muscle control

- Voluntary/Involuntary control
  - Cortical input provides volition
  - Pontine centers
    - Apneustic center (inspir)
    - Pneumotaxic center (expir)
  - Medullary centers
    - VRG
      - Primary driver of breathing
      - Output to the phrenic nerve via spinal integrating center
    - DRG
      - Integrates pontine centers
Phrenic nerve

- Anterior horn cells at C3-5 cervical level
- Solely composed of lower motor neuron axons
Axonal degeneration

A: Neurons, Myelin, Axon, Myocytes
B: Degenerative change
C: Normal state
Normal Phrenic

Injured Phrenic

Paced Injured Phrenic
Pathology Amenable to Pacing

Amenable

• Upper motor neuron lesions
  – High cervical cord injury
• Respiratory center lesions
  – Central sleep apnea
  – Brainstem stroke
  – Pontine tumors
  – MS
• Muscle diseases?
  – Disuse atrophy
  – Pompe disease

Rarely amenable

• Phrenic nerve lesions
  – Brachial neuritis
  – Trauma
  – Head/Neck cancers
• LMN disorders
  – Charcot Marie Tooth
  – ALS
  – Spinal muscular atrophy
Partially Reinnervated Phrenic
Vs.
Injured Phrenic
Partially Reinnervated Phrenic
Phrenic Nerve Conduction Study

• Mimics pacing
  – Stimulate phrenic nerve at neck
  – Record diaphragm muscle response (CMAP)
  – Right and Left performed independently
Phrenic Nerve Conduction Study

![Graph showing phrenic nerve conduction study results]

<table>
<thead>
<tr>
<th>Stimulus site</th>
<th>Beat1 (ms)</th>
<th>Dur (ms)</th>
<th>Amp (mV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>A1: Left phrenic nerve</td>
<td>10.5</td>
<td>24.8</td>
<td>0.5</td>
</tr>
<tr>
<td>B2: Right phrenic nerve</td>
<td>7.9</td>
<td>15.6</td>
<td>0.8</td>
</tr>
</tbody>
</table>

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Phrenic Nerve Conduction Study

Pros

• Ex-vivo test of pacing
• Objective and measurable
• Easily compares sides

Cons

• Focuses on anterior diaphragm
• Not all neurologists comfortable
• Must time stimulation
• Body habitus
• Postioning
Needle Electromyography

Needle Electromyography

Pros

• Distinguishes muscle from nerve pathology
• Detects ongoing denervation versus reinnervation

Cons

• Painful
• Risk of pneumothorax
• Can only study one side
• Positioning
Video Fluoroscopy

Pros
• Real-time assessment
• Readily available
• Easily compares sides

Cons
• Focuses on anterior diaphragm
• No localization information
• Must rely on radiologist
• Paradoxical movement
• Radiation
Ultrasound

• Emerging as a useful technique
  – Parameters still being developed
    • Thickness
    • Movements
• Can be combined with nerve conduction studies
  – Helpful when body habitus is a problem
Conclusions

• Etiology/pathophysiology of diaphragmatic paralysis critical to pacing decision
  – Diseases of lower motor neurons are generally resistant to pacing
  – Central nervous system diseases are most amenable

• Evaluation can be multimodal and utilize objective measures of diaphragm function
  – NCS, fluoroscopy, ultrasound