Diaphragmatic Paralysis

Background

58th Tri-State Consecutive Case Conference

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Causes of Diaphragmatic Paralysis

**Neurologic Causes**
- Spinal cord transection
- ALS (amyotrophic lateral sclerosis) and Multiple sclerosis
- Polio and post-polio syndrome
- Guillain-Barre syndrome
- Phrenic nerve dysfunction
  - Cardiac surgery "frostbite"
  - Tumor compression
  - Idiopathic phrenic neuropathy
  - Post viral neuropathy
  - Radiation
  - Chiropractic manipulation

**Myopathic Causes**
- Hyper- or hypothyroidism
- Limb-girdle dystrophy
- Malnutrition (hypo-PO4)
- Acid maltase deficiency
- Amyloidosis
- Idiopathic myopathy
- Connective tissue diseases
  - SLE
  - Dermatomyositis
  - Mixed connective tissue disease

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Dx of Bilateral Diaphragmatic Paralysis

◊ **Clinical manifestations**
  - Rapid onset of dyspnea when supine

◊ **PFTs and ABGs**
  - VC falls > 50% from upright to supine position
    - Normal decline only ~10%

◊ **Static Imaging Studies**
  - CXR- Bilateral smooth elevations of HDs
    - nonspecific low lung volumes
    - DDx includes subpulmonic effusions
  - Ultrasound to measure HD thickness
  - CT and MRI of little value
Dx of Bilateral Diaphragmatic Paralysis

Dynamic Studies

- Transdiaphragmatic pressure (Pdi) – gold standard
  - Requires esophageal and gastric balloon monitors
    - Pdi = Pga – Pdi
    - Twitch Pdi with stimulation of phrenic nerve improves reliability

- Fluoroscopy or ultrasonography with sniff test
  - Can be misleading in bilateral HD paralysis

- PSG – often discloses concomitant sleep disorders

- EMG or Respiratory Inductive Plethysmography (RIP)

Ultrasound: Chest 2008; 133:737
Review: Semin Respir Crit Care Med 2009;30:315
Respiratory Inductive Plethysmography
Diaphragmatic Flutter Post-CABG
3 Weeks After Phrenic Nerve Frostbite

Diaphragmatic Paralysis
Pdi & Resp Inductive Plethysmography
Diaphragmatic Paralysis - Treatment

◊ Bilateral
- MVS with NIPPV or via trache
- Diaphragmatic pacing (via phrenic or directly) for patients with CNS etiology
  - High (C1 & C2) spine injury (C 3,4,5 keeps the HD alive!)
  - Primary central sleep apnea, incl children (Ondine’s curse)
  - Secondary causes: brainstem infarcts, tumors, infection

◊ Unilateral
- Tx often not required
- Surgical plication (open or VATS)
  - Stabilizes the affected HD, limiting paradoxical motion
  - Limits wasted work of breathing

Qureshi. Semin Respir Crit Care Med 2009;30:315
Hx of Diaphragmatic Pacing

◊ 1777-1818
  - Cavallo & Hufeland proposed using electricity and then Ure used electricity to produce diaphragmatic contractions in a criminal (after death by hanging)

◊ 19th Century
  - Duchenne & Remak stimulated phrenic nerves

◊ 1948- Sarnof coined “electrophrenic stimulation”
  - Achieved normal gas exchange in animals & patients

◊ 1959- Glenn, et al
  - Employed radio frequency stimulation of phrenic nerve

Moxham. ARRD 1993;148:533-566
Plication of the Diaphragm to Minimize Its Paradoxical Motion During Inspiration
...When you can’t breathe, nothing else matters...”

“So you can’t breathe. I can’t play the piano but I don’t go on about it!”