Diaphragmatic Paralysis

Background

58th Tri-State Consecutive Case Conference

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Causes of Diaphragmatic Paralysis

Neurologic Causes

- ◊ Spinal cord transection
- ◊ ALS Multiple sclerosis
- ◊ Polio & 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 dis

Roussos & Macklem. N Engl J Med 1982; 307:786-792
Garcia-Morato & DeVito. Clin Pulm Med 2004;11:25-32

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)

Pdi: Surg Clin North Am 2010;90:955

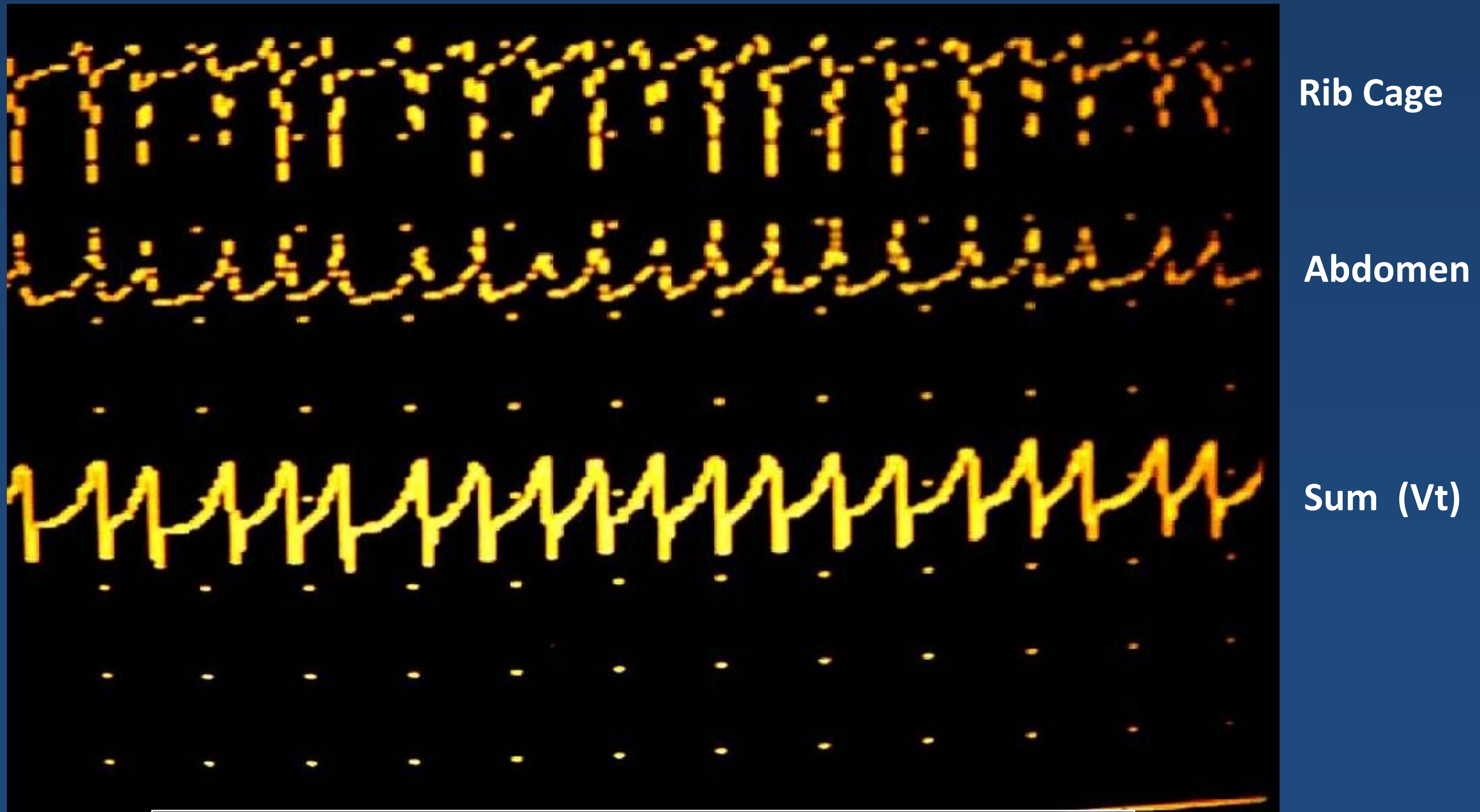
Ultrasound: Chest 2008; 133:737

RIP: Krieger. Chest 1988; 94: 254-261.

Review: Semin Respir Crit Care Med 2009;30:315

Respiratrory Inductive Plethysmography

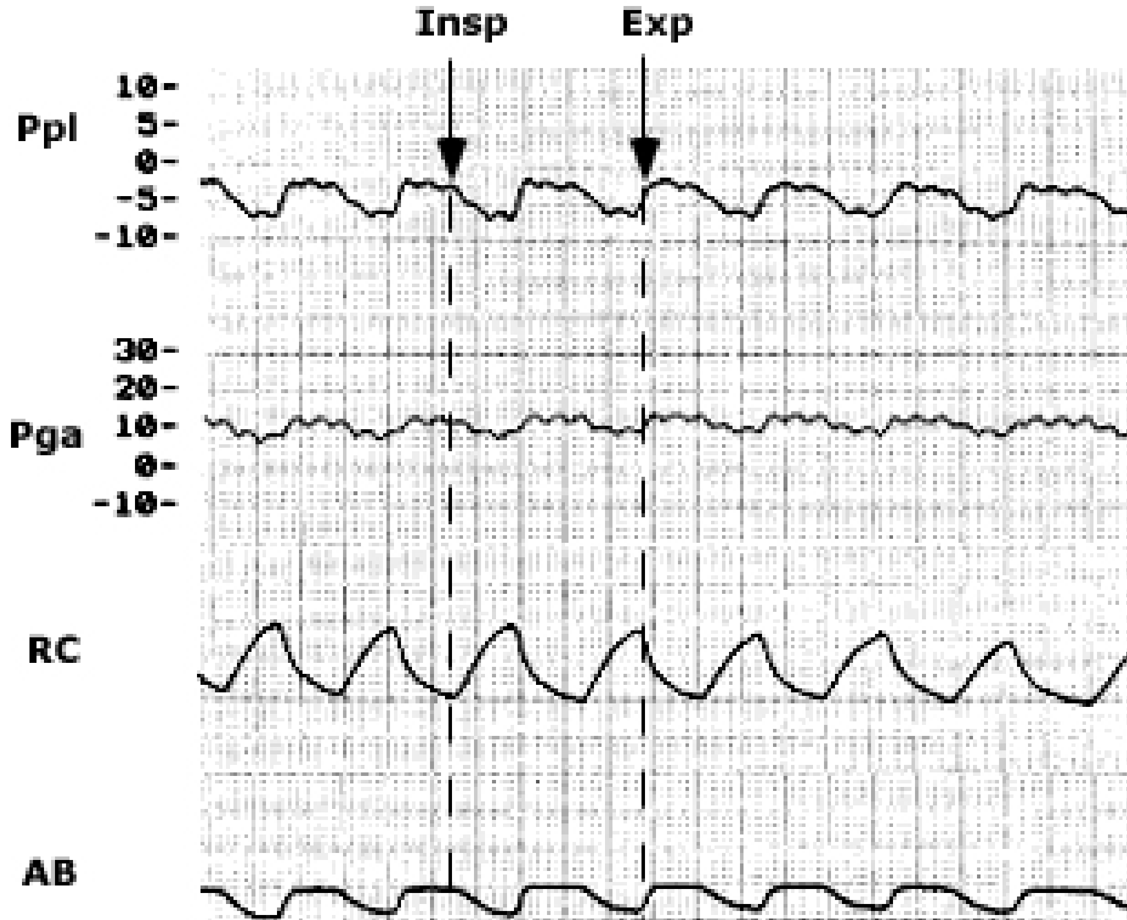
Diaphragmatic Flutter Post-CABG
3 Weeks After Phrenic Nerve Frostbite



Hoffman, Yahr, Krieger. Crit Care Med 1990; 18: 499-501

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

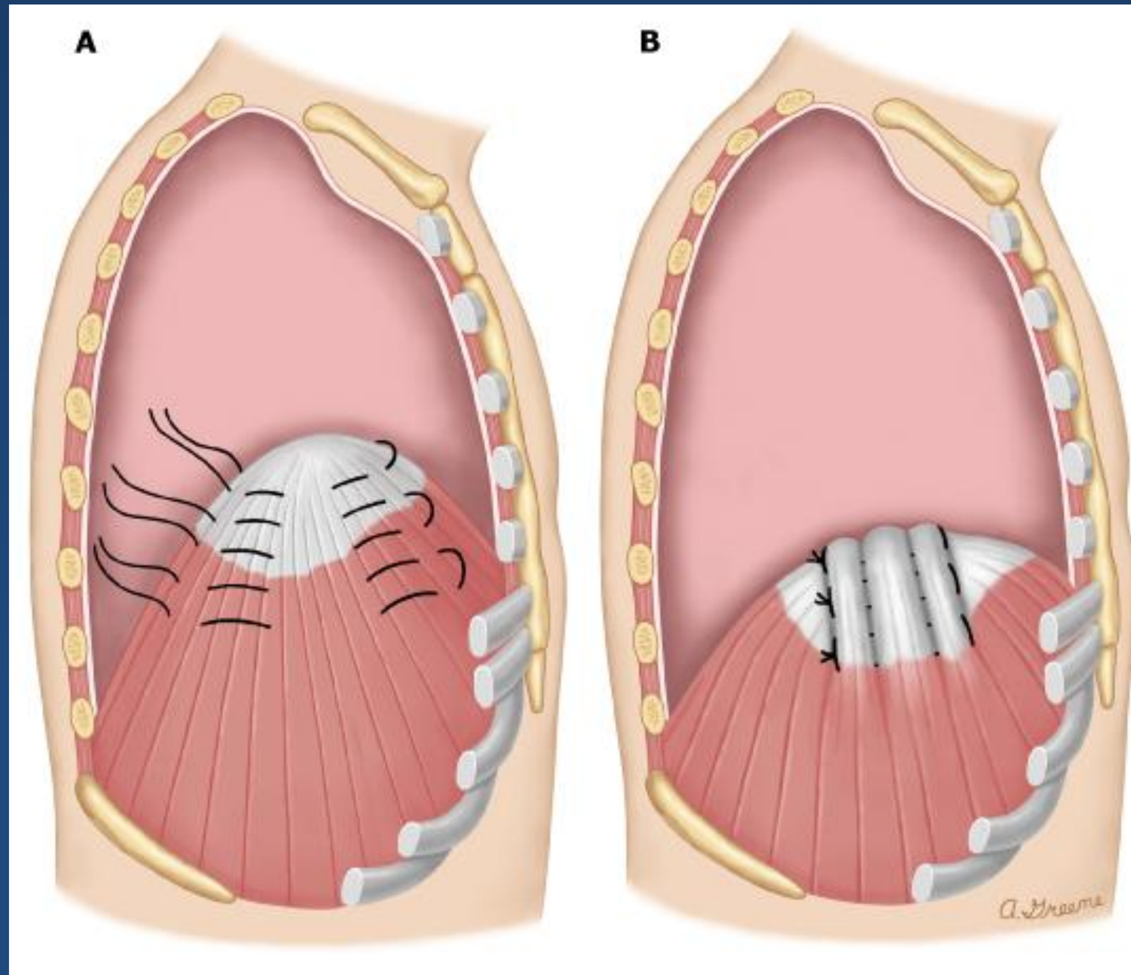
Qureshi. Semin Respir Crit Care Med 2009;30:315

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

Glenn, et al. Ann Surg 1964;160:338-350
Glenn, et al. N Engl J Med1972;286:513-518
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…"