QUESTION 28 – respiratory diaphragmatic paralysis

A 45 yo man presents with increasing breathlessness on exertion and poor sleep quality. Physical examination shows paradoxical breathing. Which of the following is the most appropriate investigation to confirm a diagnosis of bilateral diaphragm paralysis?

A. Erect and supine vital capacity
B. Arterial blood gas analysis
C. Sleep study
D. Diaphragm fluoroscopy (sniff test)
E. Maximum inspiratory pressure

DIAPHRAGMATIC PARALYSIS

Normal Diaphragm Function

- 2 components
  o Non-contractile central tendon
  o Contractile muscle fibres which radiate circumferentially and insert into lower 6 ribs and costal cartilages and L1 to L3

- Innervated by C3 to C5 via ipsilateral phrenic nerve
- Contraction has the effect of:
  o Decreasing intrapleural pressure
  o Raising the rib cage using the abdomen as a fulcrum
  o Expanding the rib cage by generating positive intraabdominal pressure

- Diaphragm performs most of the work of normal ventilation
- Assisted by some of the accessory muscle of respiration

Paralysis

- Accessory muscles assume some or all of the work of breathing
- Weakening of accessory muscles can lead to ventilatory failure (due to fatigue)

Bilateral Paralysis

Aetiology:

- Usually seen with severe generalised muscle weakness
- Sometimes can be the first or only muscle involved
- Motor neuron disease (eg: post-polio syndrome, GBS)
- Myopathy (eg: muscular dystrophy, hyperthyroidism, hypothyroidism)
- Idiopathic less common

Clinical Manifestations:

- Dyspnoea that worsens in supine position
- Occurs within minutes of lying down
- Associated tachypnoea and rapid shallow breathing
Daytime fatigue
- Paradoxical abdominal wall retraction during inspiration
- Hypoxaemia, hypercapnia, atelectasis
- If severe: ventilatory failure, pulmonary HT, erythrocytosis

**Diagnosis:**

1) CXR
   - Elevated hemidiaphragms, small lung volumes, atelectasis
2) Vital Capacity
   - Vital capacity in upright and supine positions (50% decrease in VC when supine compared to 10% in normal pts)
3) EMG
   - EMG – can help differential neuropathy from myopathy but requires expertise to perform and interpret
4) Maximal Inspiratory Pressure
   - Detects diaphragm dysfunction and paralysis, but can sometimes be normal due to preserved strength of accessory muscles
5) Transdiaphragmatic Pressure
   - Gold standard
   - Transnasal placement of balloon at lower third of oesophagus to detect changing pleural pressures, and another balloon in stomach to detect changes in abdominal pressure
   - Expertise required
6) Fluoroscopy
   - Can be misleading as movement of ribs and accessory muscles can give false appearance of diaphragmatic displacement
   - This limitation is not an issue in unilateral paralysis where sniff fluoroscopy is positive in over 90% of cases

**Unilateral Paralysis**

**Aetiology:**
- Phrenic nerve injury
- Herpes zoster, cervical spondylosis, polio, compressive tumours, pneumonia
- Idiopathic

**Clinical Manifestations:**
- Often asymptomatic at rest
Dyspnoea with exercise
- Orthopnoea, but less intense than with bilateral paralysis

Diagnosis:
- Radiological tests usually enough to diagnose (ie. Fluoroscopy).
  1) CXR
  2) Fluoroscopic sniff test – paradoxical elevation of the paralysed hemidiaphragm with inspiration is positive in over 90% of pts
  3) Reduced vital capacity (less so than with bilateral paralysis), VC reduced by 15 to 25% in supine position
  4) EMG has limited role in unilateral paralysis

Blood gas analysis and sleep study have no role in the investigation of diaphragmatic paralysis.

Fluoroscopy is useful (and often sufficient for diagnosis) in unilateral paralysis but not bilateral paralysis where it can be misleading.

Maximum inspiratory pressures can be affected by accessory muscle use so will not assist in the diagnosis when the diaphragm is the only muscle affected.

Erect and supine vital capacities are easy and will show a significant reduction in the supine position in bilateral paralysis of the diaphragm so this is the most useful investigation to confirm the diagnosis.