

Evaluation and Management of Diaphragm paralysis

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Diaphragm: Chief inspiratory muscle

- With quiet breathing, the diaphragm accounts about 75 to 80% of ventilation.
- The vertical movement of the diaphragm is 1 to 2 cm during quiet breathing and 6 to 7 cm during deep breathing.
- Each cm of vertical movement contributes 300 to 400 ml of air during normal breathing.

Diaphragm: Anatomy

- Dome shaped
- Musculotendinous structure
- Muscular portion
 - 2 parts: costal and crural portions
 - Costal portion is thinner and the crural portion is thicker.
- Aponeurosis of central tendon
- Both hemidiaphragms are innervated by the phrenic nerve.
- The costal portion flatten the diaphragm and lift the rib.
- The crural portion causes downward placement of the diaphragm(less effective in breathing.)

Normal effects of the diaphragm on the rib cage

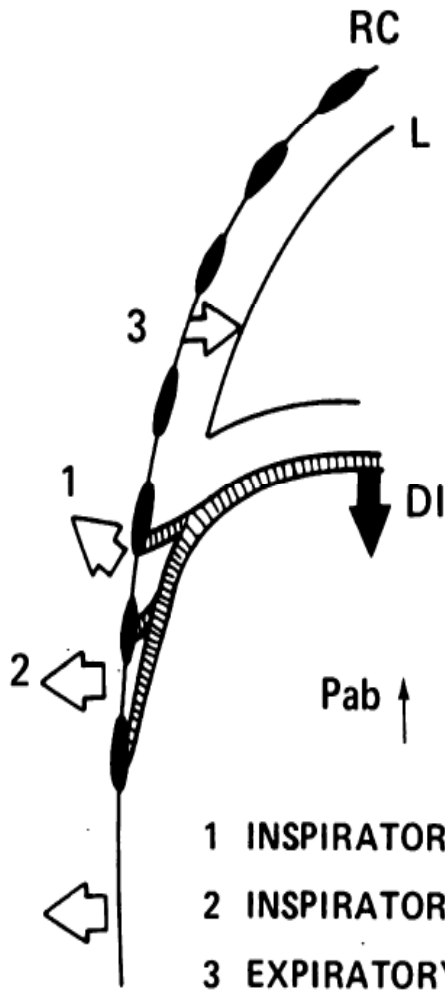
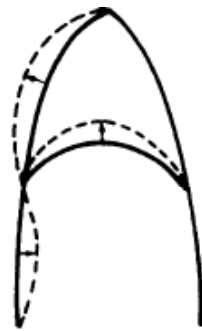
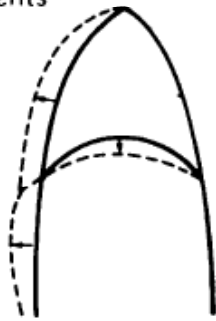


Fig 2 Normal effects of the diaphragm on the rib cage (RC). The lower rib cage is expanded (1) by direct insertions, the diaphragm (DI) using the abdominal contents as a fulcrum, and (2) by the effect of increasing abdominal pressure (P_{ab}) via the zone of apposition; these effects are countered (3) by the deflating effect of pleural pressure (P_{pl}) as it becomes more negative. The net effect is inspiratory but mechanisms (1) and (2) become progressively less effective as lung volume increases.

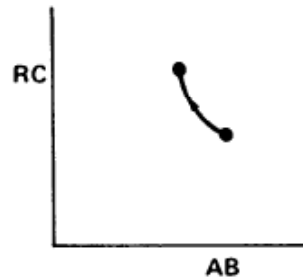
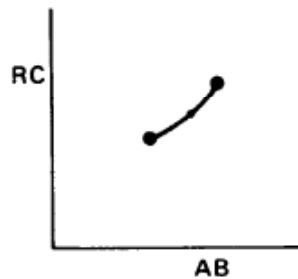
INSPIRATION WITH
DIAPHRAGM ALONE

INSPIRATION WITHOUT
DIAPHRAGM

Movements



Magnetometry



Pressures

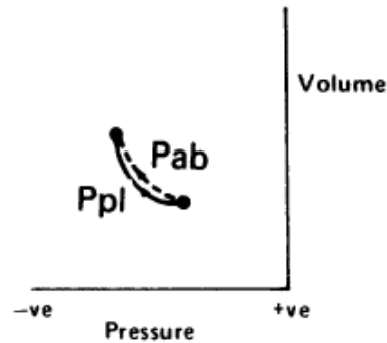
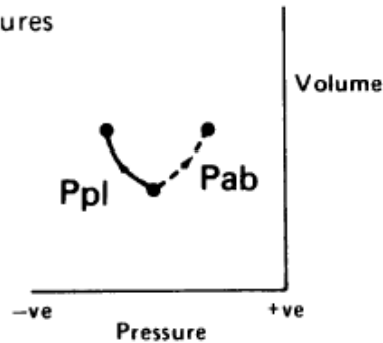
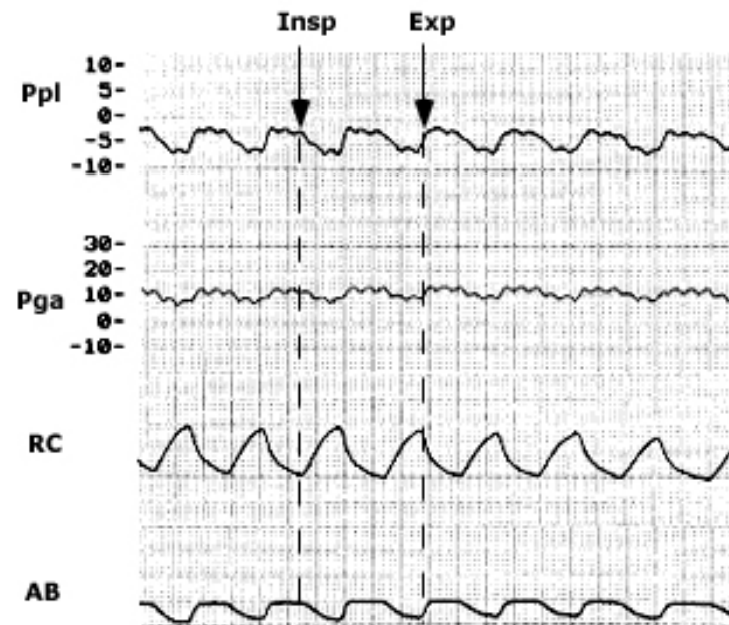


Fig 3 Comparison of chest wall motion and pleural and abdominal pressure changes when inspiration is achieved by the diaphragm (left) and by other inspiratory muscles in complete diaphragmatic paralysis (right); with diaphragmatic contraction both rib cage (RC) and abdominal (AB) dimensions increase and abdominal pressure (Pab) becomes more positive. With bilateral diaphragmatic paralysis the rib cage and abdomen move out of phase with each other—that is, abdominal paradox occurs and abdominal pressure passively follows pleural pressure (Ppl).

Transdiaphragmatic pressure

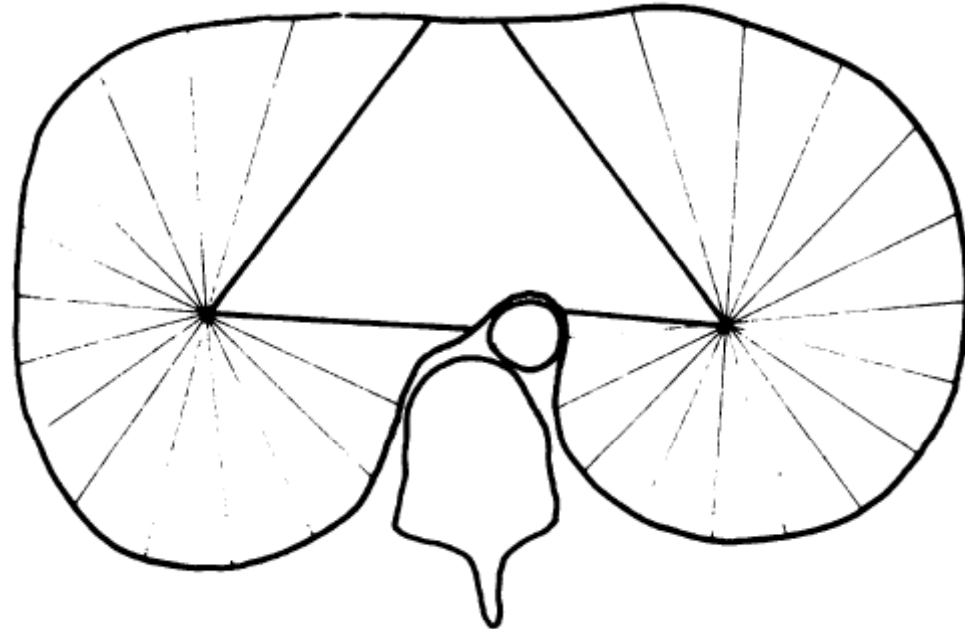


Real-time tracing of esophageal pressure as a reflection of pleural pressure (Ppl) and abdominal pressure as a reflection of gastric pressure (Pga) in a patient with bilateral diaphragmatic paralysis. Decreased intrathoracic pressure (more negative Ppl) on inspiration (Insp) is associated with an abnormal decrease in gastric pressure (more negative Pga). At the same time, there is outward displacement of the rib cage (upward deflection of RC) with a paradoxical decrease in abdominal volume (downward deflection of AB).

Adaptation to diaphragmatic paralysis

- Inspiration is achieved by inspiratory intercostal and accessory muscles, which lower pleural pressure and expand the rib cage
- Muscles of abdominal wall act as accessory muscles of inspiration
 - Ascent of paralyzed diaphragm at end of expiration, so that when abdominal contraction ceases the diaphragm descends passively at the onset of inspiration by pull of gravity at upright position
- Glottal closure at end inspiration, which allows maintenance of lung volume without continuing contraction of the inspiratory muscles.

Independence of right & left hemidiaphragms



Direction of lines of tension from the centre of each hemidiaphragmatic dome to fixed attachments, based on the computed tomography reconstruction

Because lines of tension project to the opposite hemidiaphragm only via the narrow isthmus between the spine and the xiphoid process tension on one side is not well transmitted to the other.

Whitelaw WĀ. Shape and size of the human diaphragm in-vivo. *J Appl Physiol* 1987;62:180-6.

Costal & Crural Portions: Concept of "TWO DIAPHRAGMS"

- Stimulation of the costal and crural portions individually produces very different effects:
 - In supine - contraction of the costal fibres at resting lung volume increased rib cage dimensions but contraction of the crural fibres did not
 - At higher lung volumes, or when the abdomen was opened to the atmosphere, stimulation of crural diaphragm led to diminution of rib cage dimensions.
- Embryological, costal diaphragm develops from the lateral body wall whereas the crural portion develops in the dorsal mesentery of the oesophagus.

Methods of assessment of diaphragmatic function

CxR

- An unusually high position of one or both hemidiaphragms is less specific.
 - Felson showed raised diaphragm on left in 9% and on right in 2% of 500 normal chest films*
- On full inspiration, bilateral elevation of the diaphragms implies one of the following:
 - reduction in pulmonary compliance
 - reduction in abdominal compliance
 - pleural adhesions, or
 - weakness of the diaphragmatic muscle
- Unilateral elevation is seen with
 - local lesions above or below the diaphragm,
 - hemidiaphragmatic weakness or paralysis, or
 - Diaphragmatic eventration

* Felson B. *Chest Roentgenology*. Philadelphia: WB Saunders, 1973.

Fluoroscopy:

- Best established method
- Value in recognition of hemidiaphragmatic paralysis
- Unequal excursion of the two hemidiaphragms is normal
 - In one series the hemidiaphragms moved asynchronously in 77%, although the difference in excursion was usually <1 cm
 - The excursion in this group of normal subjects ranged from 3 to 6 cm in 75%, with 23% showing excursion of less than 3cm and 2% more than 6 cm.

Clin Radiol 1969; 20: 231.

Fluoroscopy: Sniff test

- Unilateral paradox on sniffing has been seen in up to 6% of normal subjects on a deep inspiratory effort against a closed airway (sniff test) and is more common on the right

Clin Radiol 1966; 17: 79

- Results to be interpreted with some caution
 - The sniff test result is considered positive if a ≥ 2 cm excursion is present and the whole leaf of the hemidiaphragm, in the oblique view, is involved.
- Positive in 90% of cases of U/L diaphragmatic paralysis.
- In bilateral paralysis, the sniff test result may be misleading
 - cephalad movement of the ribs and accessory muscle contraction gives the false appearance of caudal displacement of the diaphragm

Diaphragmatic ultrasonography

- New method of assessing diaphragmatic function
 - Diaphragmatic motion during inspiration and
 - Diaphragmatic thickness in the zone of apposition
- Diaphragmatic thickness at FRC of <2 mm combined with <20% increase in thickness during inspiration can provide discrimination between paralyzed and normal diaphragm

Am J Respir Crit Care Med 1997;155:1570-1574.

CT scan

- Estimate the thickness of the diaphragmatic leaves and the height of the dome; both show large ranges in normal subjects
- Reconstructions of images in the sagittal or coronal planes give more accurate information on the thickness and composition of the diaphragmatic dome and have been used in detailed reconstruction of diaphragmatic topography
- No clinical use appears to have been made of this approach

Respiratory Muscle Assessment

Chest wall motion

- Measured by magnetometers or inductance Plethysmography
- Ratio of rib cage and abdominal displacement
 - Index of diaphragmatic function
 - Ratio is negative with complete bilateral paralysis
 - Not specific as displacement influenced by contraction of the other groups of respiratory muscles too

LUNG VOLUME S

- Difference in erect and supine vital capacity
 - Fall by < 10% in normal subjects
 - Fall by 20 – 50 % in diaphragmatic weakness
- TLC is virtually unchanged in unilateral paralysis
- Maximal voluntary ventilation, VC, and FEV1 are each decreased by about 50% with bilateral paralysis and by about 25% with unilateral paralysis

- Wide variation in decrease in lung volumes among patients with unilateral & bilateral paralysis
 - Some pts with b/l paralysis have normal or near normal values.
 - About 1/2 of pts with b/l or u/l paralysis have decreased respiratory muscle endurance
 - Decrease is associated with a shorter duration of disease and less output of the expiratory muscles
- The findings suggest that patients compensate by recruiting their expiratory muscles over time

Ratio of tidal swing in $\Delta P_{ab}:\Delta P_{pl}$.

- Semiquantitative test
- Measure pressure change in P_{ga} & P_{eo}
- Assess relative contribution of the diaphragm, rib cage, and expiratory muscles to tidal breathing
- In healthy subjects, ratio is ≥ -1
- A less negative ratio indicates an ever-increasing contribution of the rib cage and expiratory muscles, as compared with the diaphragm, to tidal breathing.
- With complete diaphragmatic paralysis, the ratio becomes equal to 1.
 - Ratios more positive than 1 occur in 20% -25% of patients with bilateral diaphragmatic paralysis

Maximal inspiratory pressure (P_{imax})

- Measured at the airway opening during a maximal, static inspiratory effort (Muller maneuver) initiated at either FRC or RV
- U/L paralysis,
 - P_{imax} reduced to 60% predicted
- B/L paralysis,
 - P_{imax} reduced to < 30% predicted

Transdiaphragmatic Pressure (Pdi)

- Diaphragm contraction lowers intrathoracic pressure while increasing intra-abdominal pressure.
- Pressure developed specifically by the diaphragm can be measured as the difference between abdominal pressure, as assessed with a gastric catheter (Pga), and the pleural pressure, as assessed with an esophageal catheter (Pes).
- Transdiaphragmatic pressure (Pdi) is then calculated as $Pdi = Pga - Pes$.
 - Normal quiet inspiration, $Pdi = Pga(+5) - Pes(-5) = 10$ cm H₂O
 - With inhalation to TLC, $Pdi > 30$ cm H₂O

TABLE 3. TRANSDIAPHRAGMATIC PRESSURES DURING MAXIMAL STATIC RESPIRATORY EFFORTS AND MAXIMAL SNIFFS

	n	P _{i,di,max} (cm H ₂ O)			P _{di,sn} (cm H ₂ O)		
		Mean	SD	Range	Mean	SD	Range
Men	37	108	30	52–164	148	24	112–204
Women	27	65	31	16–40	121	25	82–182
All	64	90	37	16–164	137	28	82–204

Definition of abbreviations: P_{i,di,max} = maximum static transdiaphragmatic pressure; P_{di,sn} = transdiaphragmatic pressure during sniff.

Transdiaphragmatic Pressure (Pdi)

- Greatest values of Pdi max are not obtained during Muller maneuvers or sniffs but rather during a more complex maneuver where the individual attempts to lower Pes by contracting the diaphragm while raising Pga by contracting the abdominal muscles ().
- Values of Pdi max during maximal combined maneuvers may be 60% greater than those obtained during maximal sniffs or Muller efforts

Disadvantages

- Requires placement of esophageal and gastric catheters that may be uncomfortable or even hazardous in pts with swallowing impairment
- Pts with diaphragm paralysis or profound respiratory muscle weakness, proper placement of the gastric catheter may be difficult because the changes in Pga and Pes may parallel one another
- Maneuver needed to generate Pdi max is complex and difficult to perform
- Does not discriminate which side, or both, of the diaphragm is dysfunctional

Phrenic Nerve Stimulation

- Stimulated transcutaneously either by electric or magnetic impulses
- 2 parameters:
 - Phrenic nerve conduction time
 - Assesses the integrity of the phrenic nerve
 - Twitch Pdi
 - Assess mechanical output of the diaphragm

Phrenic nerve conduction time

- time from the onset of the stimulus to the onset of the diaphragm action potential
- measured using a surface electromyogram placed on the lower rib cage
- conduction time less than 9 msec is considered normal

Twitch Pdi

- Diaphragm mechanical output is measured as the magnitude of twitch Pdi.
- In normals, Pdi following bilateral electric phrenic nerve stimulation is generally 25 - 35 cm H₂O.
 - Values are lower with unilateral stimulation
- Values are greater with magnetic stimulation.
 - reflect activation of the inspiratory muscles of the rib cage.
 - These muscles may directly increase Pdi by lowering Pes or indirectly by decreasing chest wall compliance

Pressure tracing to PNST

Twitch Pdi

Advantages

- No patient effort
 - attractive for patients with neuromuscular disease
- Successfully and reproducibly measured in various neuromuscular disorders using either electric or magnetic stimulation

Disadvantages

- Magnitude of twitch Pdi depends on the impedance of the abdomen and rib cage.
- “Contraction history” of the diaphragm must be considered.
 - Twitch potentiation is the phenomenon whereby twitch pressures are increased if there has been a preceding maximal contraction of the diaphragm.
 - Individuals with neuromuscular disorders may have a high degree of diaphragm activity at baseline and this may predispose to twitch potentiation.
- Not readily available to most clinicians

Diaphragm paralysis

Clinical presentation

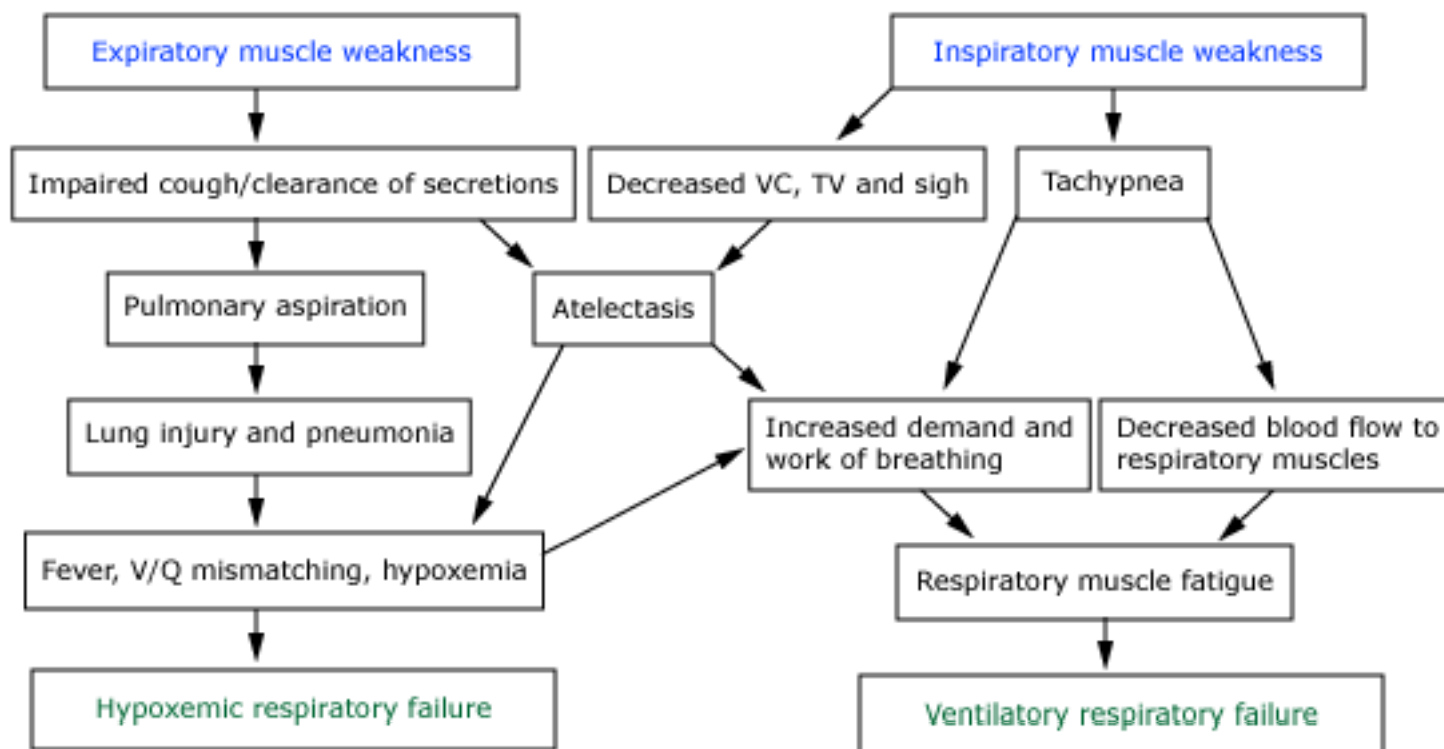
- Depends on
 - Unilateral / Bilateral
 - Isolated or part of neuromuscular weakness
 - Underlying pulmonary disease
- Dyspnea on exertion
- Dyspnea with supination, bending, or immersion in water (i.e., entering a swimming pool)
- Altered sleep cycles
- Recruitment of accessory muscles
- Abdominal paradox during resting breathing
 - 10-100% in B/L paralysis on standing/sitting
 - 50-100% on supine
 - maximal transdiaphragmatic pressure < 30 cm H₂O

Pulmonary and Ventilatory Function

Variable	Unilateral	Bilateral
VC	75	45
TLC	85	55
FRC	100	60
RV	100	55
FEV ₁	70	50
Pi _{max} (cm H ₂ O)	70	10 to 20
Pdi _{max} (cm H ₂ O)	70	40 to 10

FEV₁, forced expiratory volume in 1 second; FRC, functional residual capacity; Pi_{max} and Pdi_{max}, maximal inspiratory and transdiaphragmatic pressures, respectively; RV, residual volume; TLC, total lung capacity; VC, vital capacity.

Pathophysiology of neuromuscular respiratory failure



Diaphragm weakness and sleep

- At risk of developing hypoventilation during REM sleep
- Central nervous system can adopt two strategies:
 - Phasic recruitment of inspiratory muscles other than the diaphragm during REM sleep, or
 - Suppression of REM sleep
- The failure of all patients to develop these adaptive strategies may explain discrepancies among reports on oxygenation during sleep in patients with isolated diaphragmatic paralysis

Am J Respir Crit Care Med 2000;161:849–856

Thorax 1988;43:75–77

Causes of diaphragm paralysis

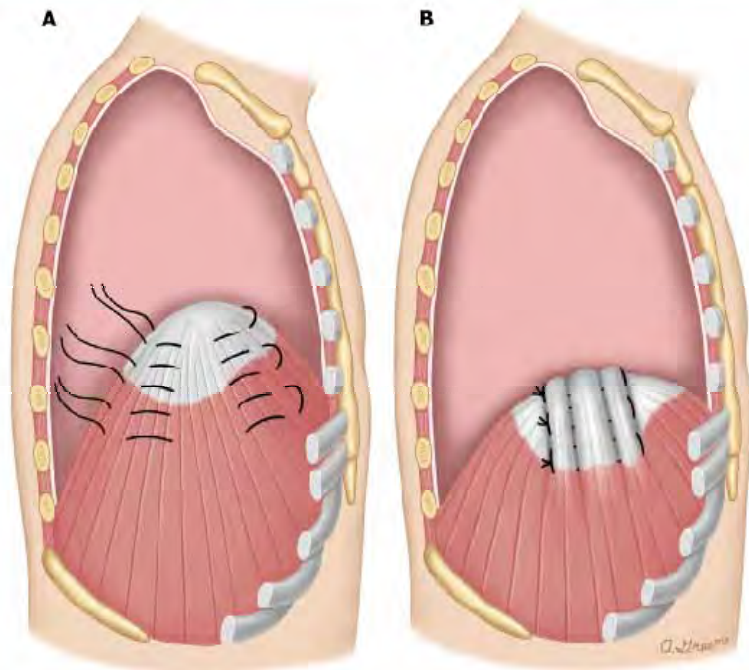
Location	Causes
Upper motor neurons	Cerebral infarction or hemorrhage
Respiratory centers	Infectious processes, Arnold-Chiari II malformation, parkinsonism
Lower motor neurons	Spinal cord injury, poliomyelitis, amyotrophic lateral sclerosis
Phrenic nerve	Trauma, mediastinal malignancies, herpes zoster, diphtheria, Lyme disease, malnutrition, alcoholism, diabetes, lead toxicity, vasculitis, porphyria, neuralgic amyotrophy, and Guillain-Barre' syndrome
Neuromuscular junction	Myasthenia gravis, botulism
Muscle fibers	Myotonic dystrophy, adult onset maltase deficiency, Hypo/hyperthyroidism, CTDs

Treatment of diaphragmatic paralysis

UNILATERAL PARALYSIS

- Asymptomatic and require no treatment
- Good Prognosis unless underlying pulmonary disease
- Treatment is considered when the dyspnea is disproportionate to the degree of physical activity or to severity of the lung disease
- Surgical plication

Diaphragm plication



Redrawn with permission from: Freeman, RK, Wozniak, TC, Fitzgerald, EB. Functional and physiologic results of video-assisted thoracoscopic diaphragm plication in adult patients with unilateral diaphragm paralysis. *Ann Thorac Surg* 2006; 81:1853. Illustration used with the permission of Elsevier Inc. All rights reserved.

- Open or VATS
- Paralyzed hemidiaphragm resists being pulled up
- Adjacent lung segments expand
- Enhances exercise performance, blood gas exchange, and respiratory muscle function

Diaphragm plication in adult patients with diaphragm paralysis leads to long-term improvement of pulmonary function and level of dyspnea[☆]

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Abstract

Objective: There is still controversy about the feasibility and long-term outcome of surgical treatment of acquired diaphragm paralysis. We analyzed the long-term effects on pulmonary function and level of dyspnea after unilateral or bilateral diaphragm plication. **Methods:** Between December 1996 and January 2006, 22 consecutive patients underwent diaphragm plication. Before surgery, spirometry in both seated and supine positions and a Baseline Dyspnea Index were assessed. The uncut diaphragm was plicated as tight as possible through a limited lateral thoracotomy. Patients with a follow-up exceeding 1 year ($n = 17$) were invited for repeat spirometry and assessment of changes in dyspnea level using the Transition Dyspnea Index (TDI). **Results:** Mean follow-up was 4.9 years (range 1.2–8.7). All spirometry variables showed significant improvement. **Mean vital capacity (VC) in seated position** improved from 70% (of predicted value) to 79% ($p < 0.03$), and in **supine position** from 54% to 73% ($p = 0.03$). **Forced expiratory volume in 1 s (FEV₁) in supine position** improved from 45% to 63% ($p = 0.02$). Before surgery the mean decline in VC changing from seated to supine position was 32%. At follow-up this had improved to 9% ($p = 0.004$). For FEV₁ these values were 35% and 17%, respectively ($p < 0.02$). **TDI showed remarkable improvement of dyspnea** (mean + 5.69 points on a scale of –9 to +9). **Conclusion:** Diaphragm plication for single- or double-sided diaphragm paralysis provides excellent long-term results. Most patients were severely disabled before surgery but could return to a more or less normal way of life afterwards.

Bilateral Paralysis

- Surgical Plication
 - Bilateral plication reserved for **irreversible** denervation with substantial paradoxical motion documented by fluoroscopy
- Mechanical ventilation: nasal CPAP, intermittent positive-pressure ventilation by nasal or oral mask
- Diaphragmatic pacing

Ventilatory support

- Noninvasive :
 - Nasal CPAP/BiPAP
 - Negative pressure ventilation- cuirass respirator
- Invasive positive pressure ventilation with tracheostomy
- Continuous/Intermittent

Diaphragm pacing

- Radiofrequency signals to pace phrenic nerve
- Intact phrenic nerve function and no diffuse myopathy
- Ideal patient is a high quadriplegic without intrinsic lung disease
- Retraining the deconditioned diaphragm requires expertise and careful monitoring
- Fast stimulating frequencies and prolonged stimulation times may lead to irreversible muscle dysfunction

Limitations

- (1) diaphragmatic fatigue during long-term diaphragm pacing,
- (2) upper airway obstruction and
- (3) the cost of the diaphragmatic pacemaker system.