EFR dans les maladies musculaires et neuromusculaires

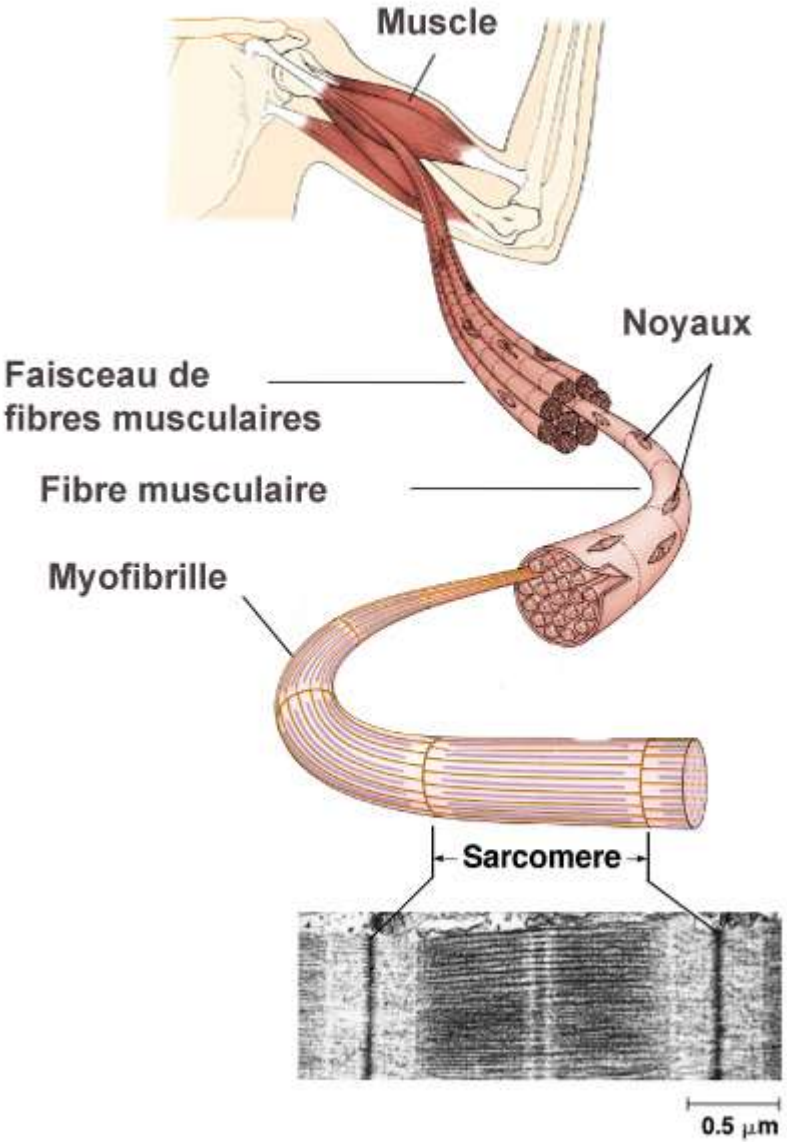
Christian Straus

Explorations Fonctionnelles de la Respiration, de l'Exercice et de la Dyspnée
Hôpitaux Universitaires Pitié-Salpêtrière, Tenon et Saint-Antoine
APHP.Sorbonne Université
Assistance Publique-Hôpitaux de Paris, Paris, France

UMR_S 1158
Inserm et Université Pierre et Marie Curie - ParisVI, Paris, France

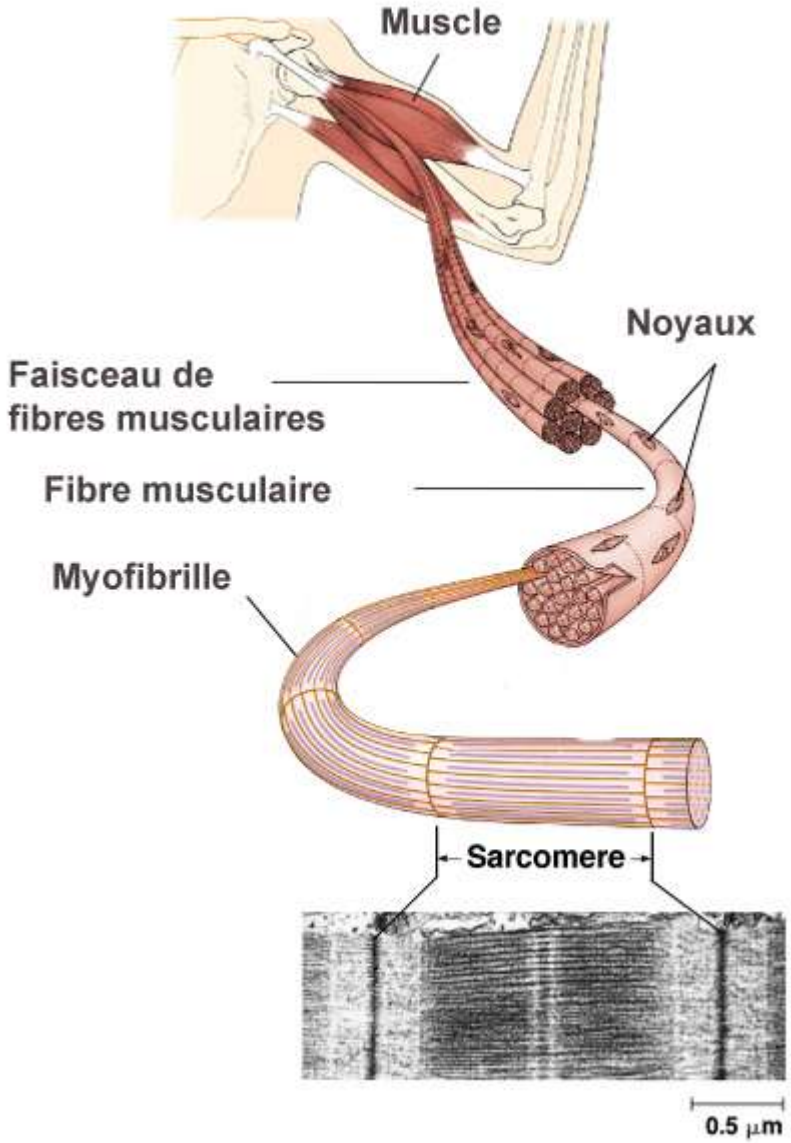
Branche "adultes" du **Centre de référence national du Syndrome d'Ondine**

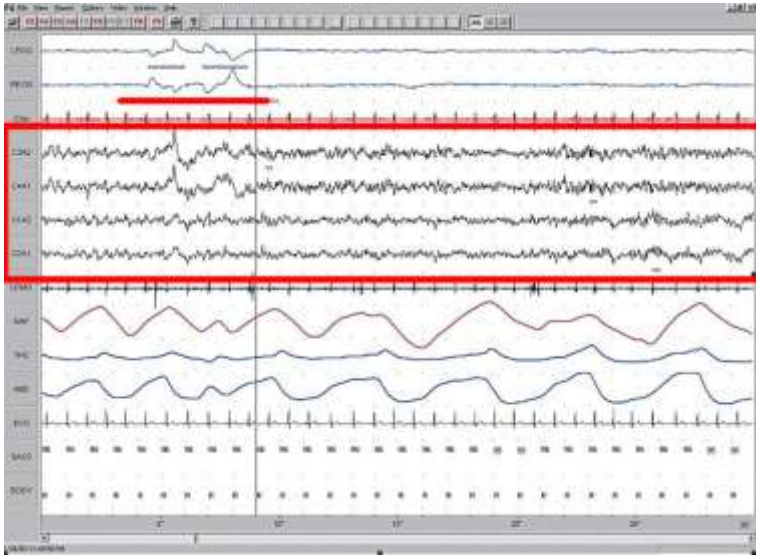
Des muscles squelettiques



Myosine
Actine

Fonctionnement permanent



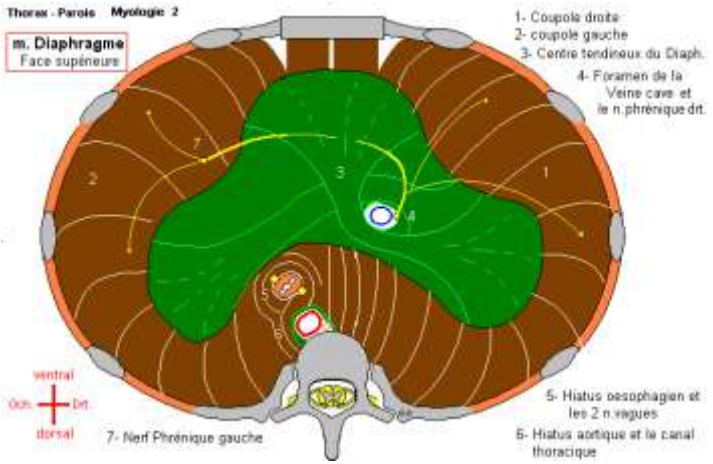


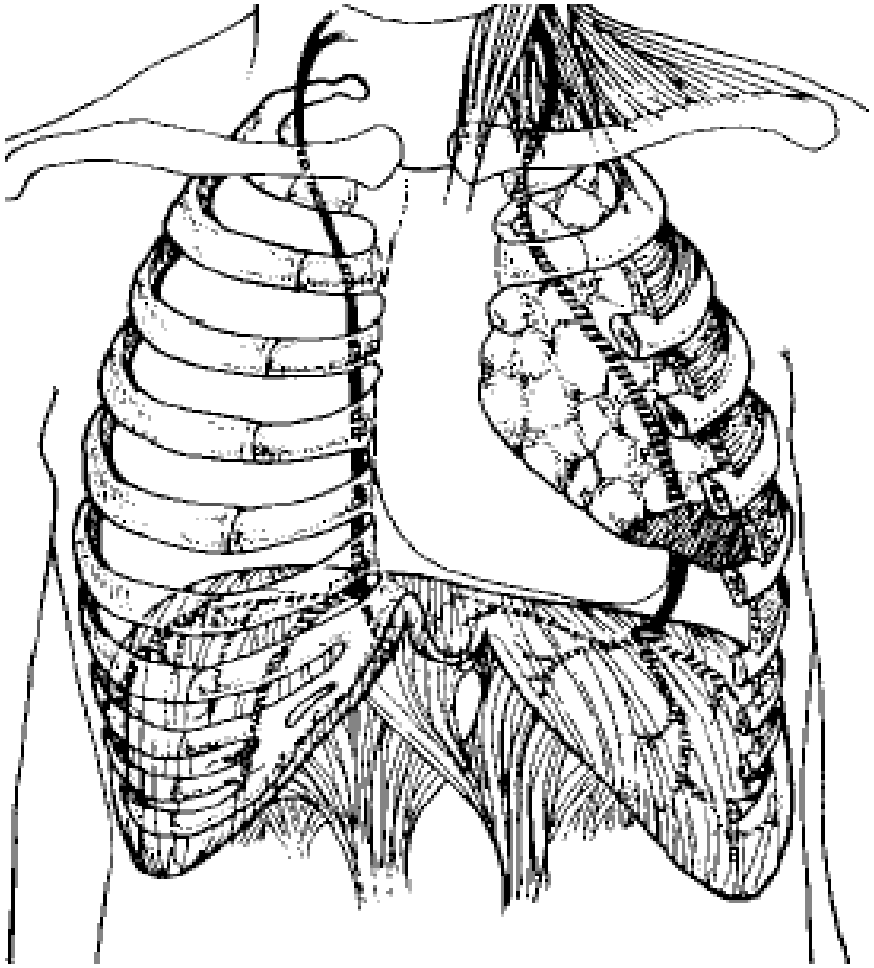
temps d'activation des muscles

- période d'activation *vs.* période où le muscle est inactif
  - long extenseur du doigt : ~ 2%
  - soléaire : ~ 14%
  - diaphragme : ~ 45%

Mantilla et Sieck J.A.P. 2003

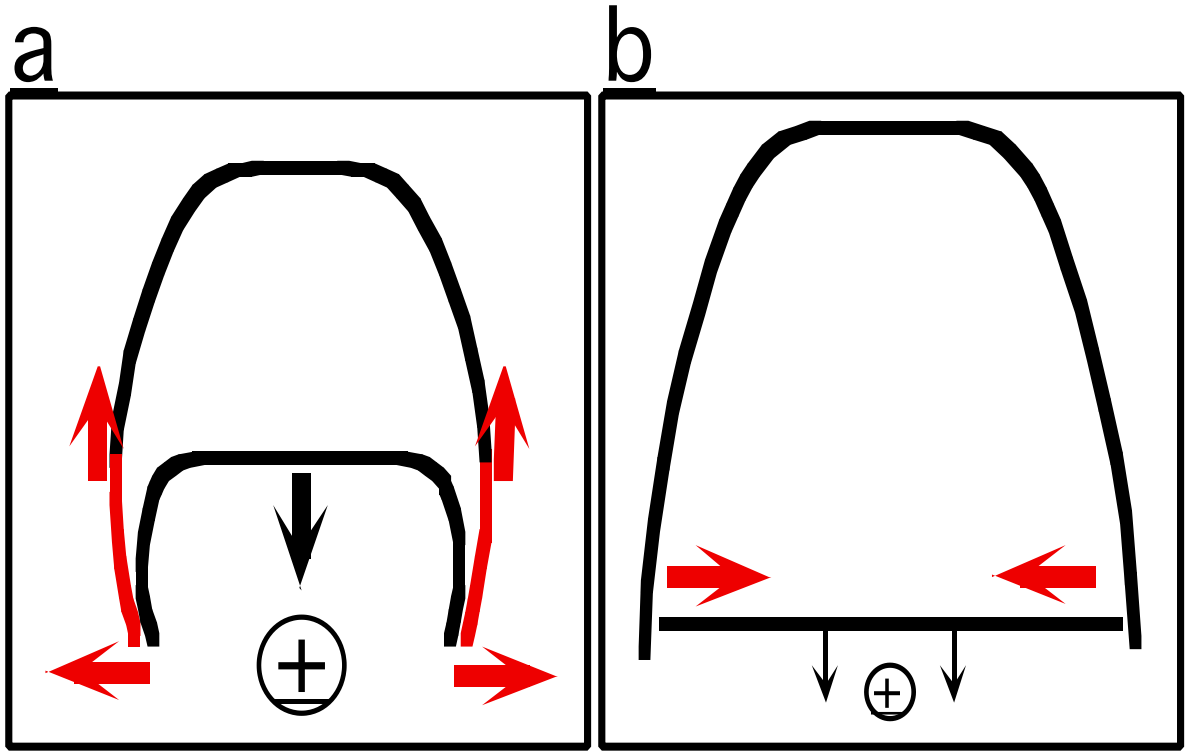
Le diaphragme, principal muscle inspiratoire, définit les mammifères





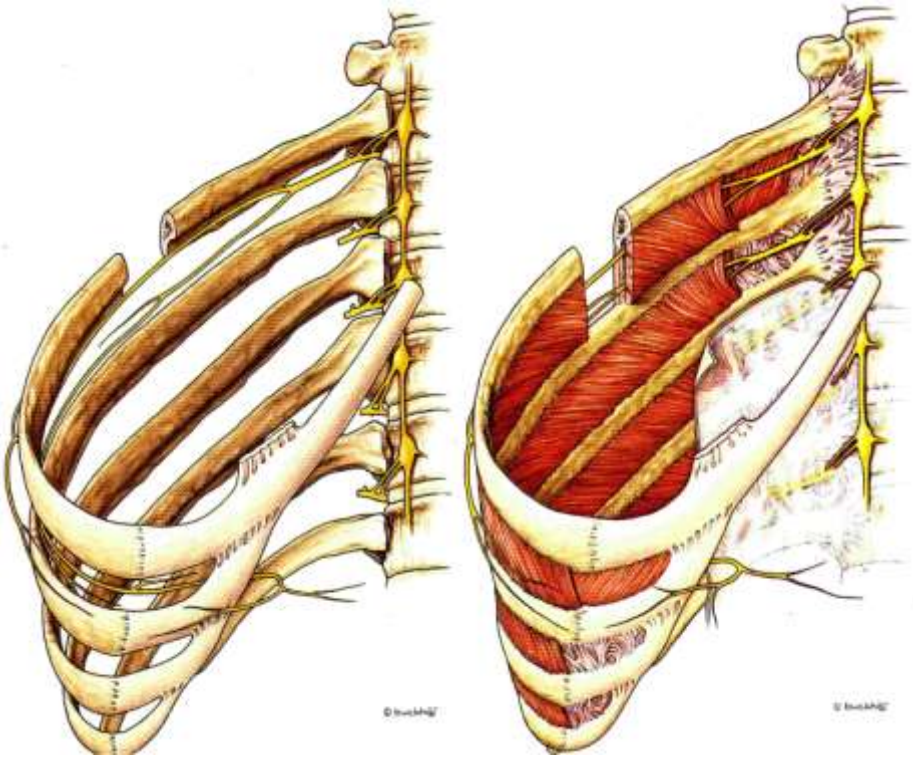
Importance de la géométrie du diaphragme

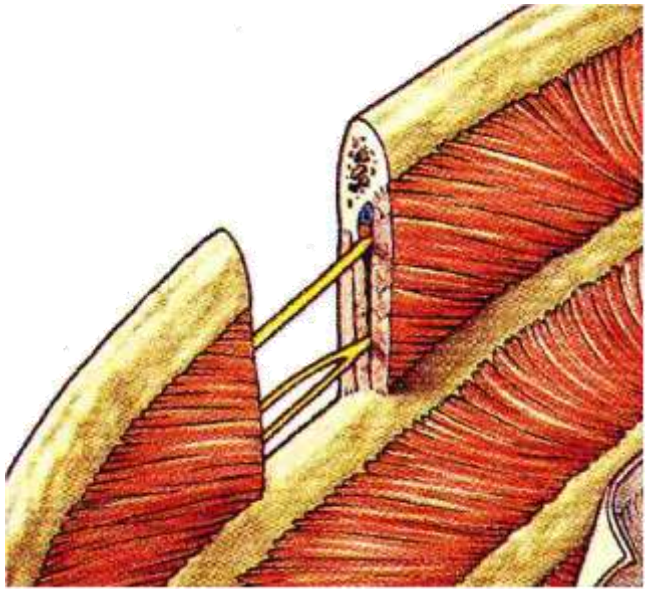
- **Illustrée par le signe de Hoover**



- **- distension thoracique**
- **- ne signifie pas dysfonction diaphragmatique**
- **- mais peut la causer**

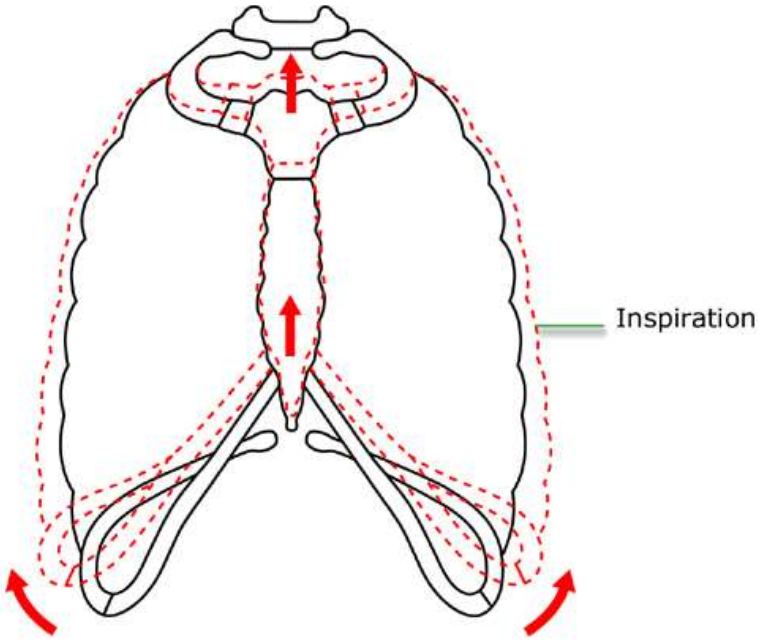
Les muscles intercostaux

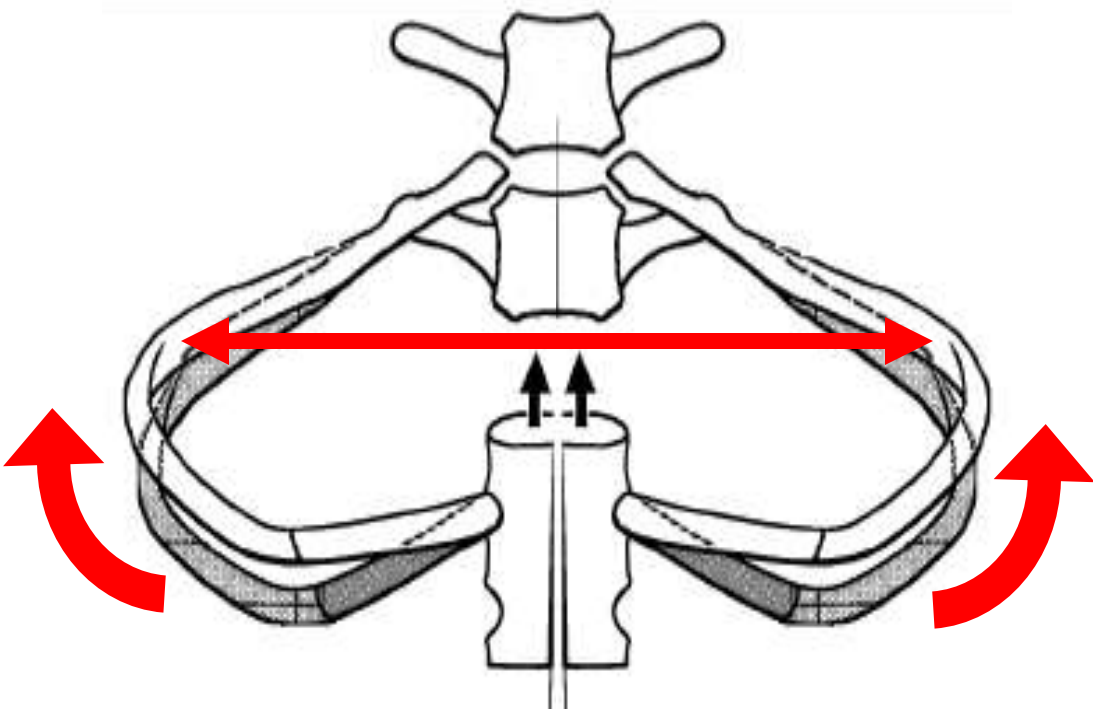




Une couche externe,
une couche interne

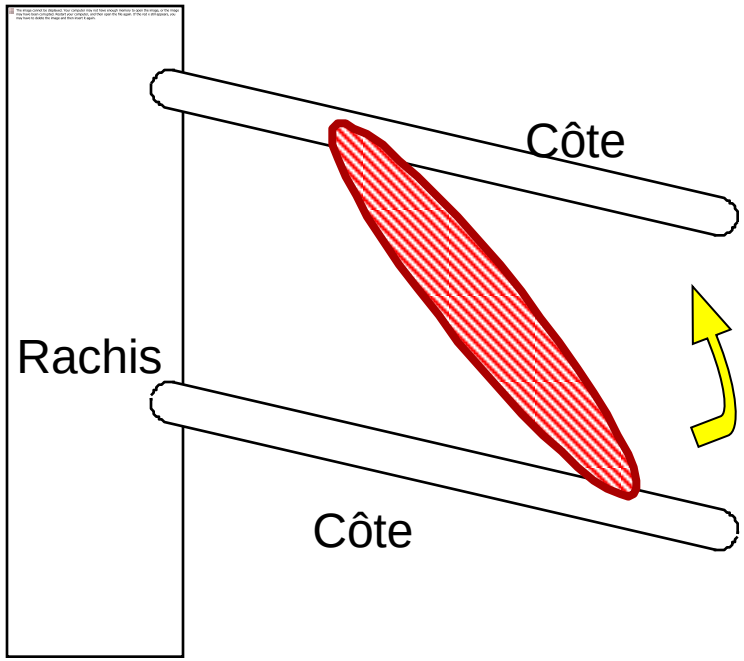
Action des muscles intercostaux inspiratoires



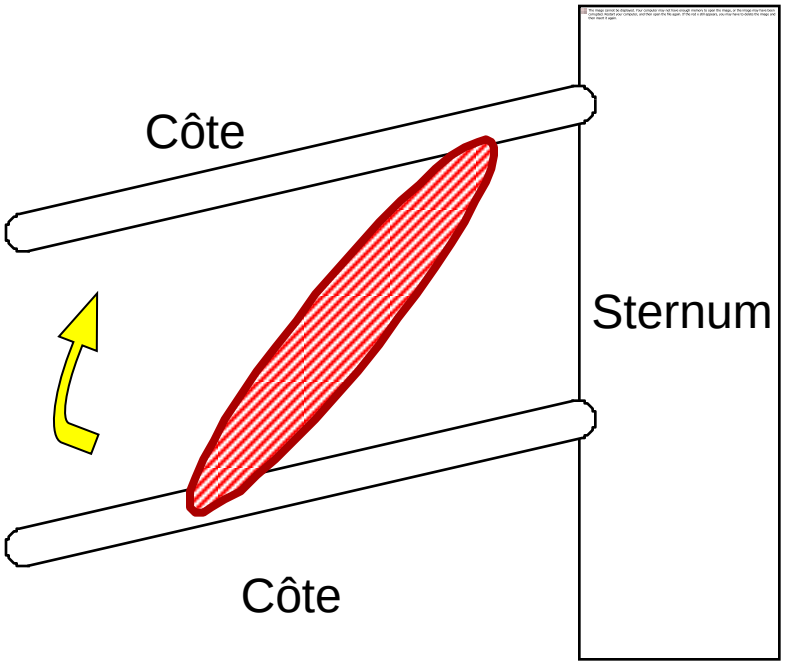


Action des muscles intercostaux inspiratoires

Muscle Intercostal Externe

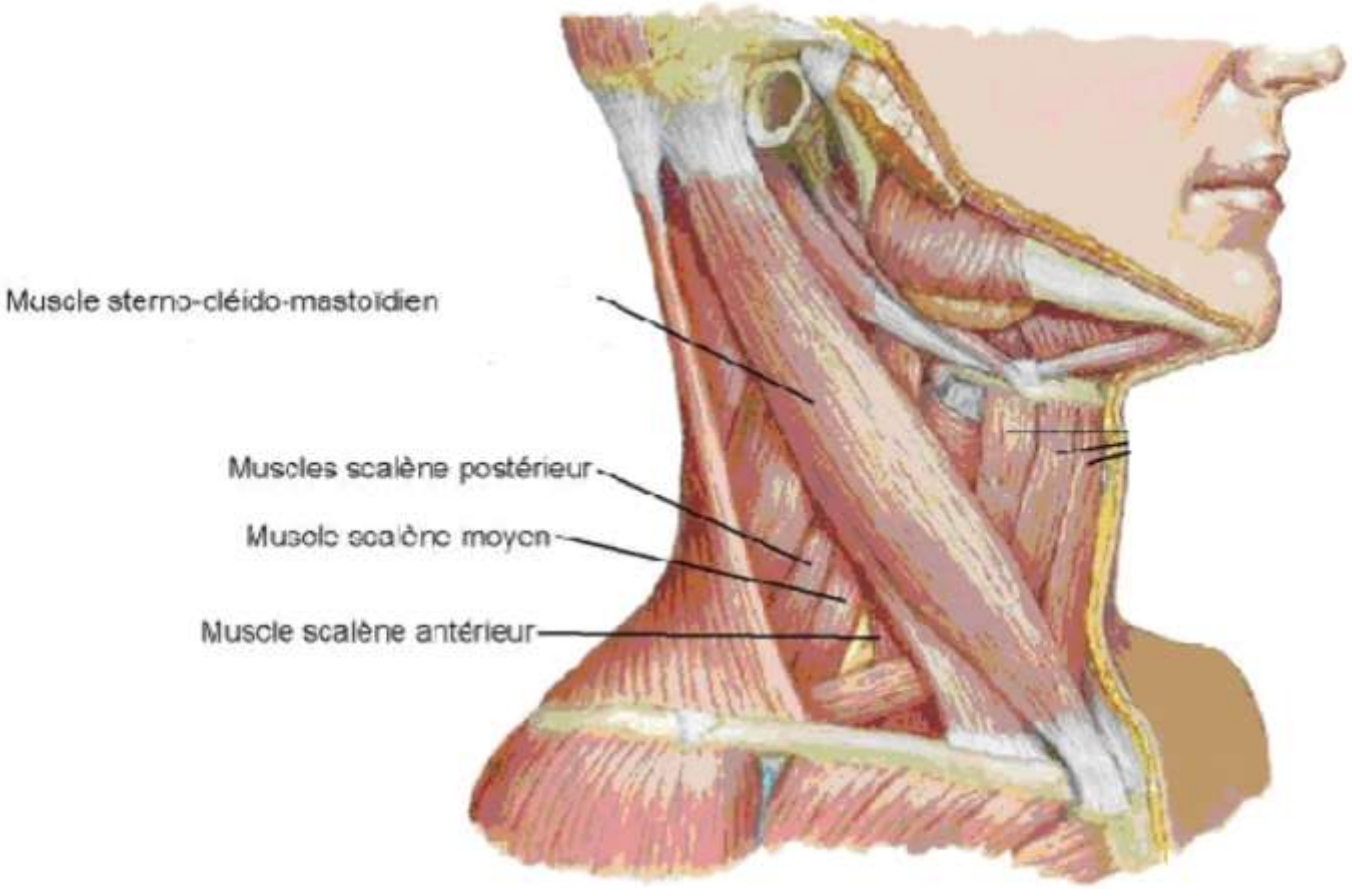


Muscle Parasternal



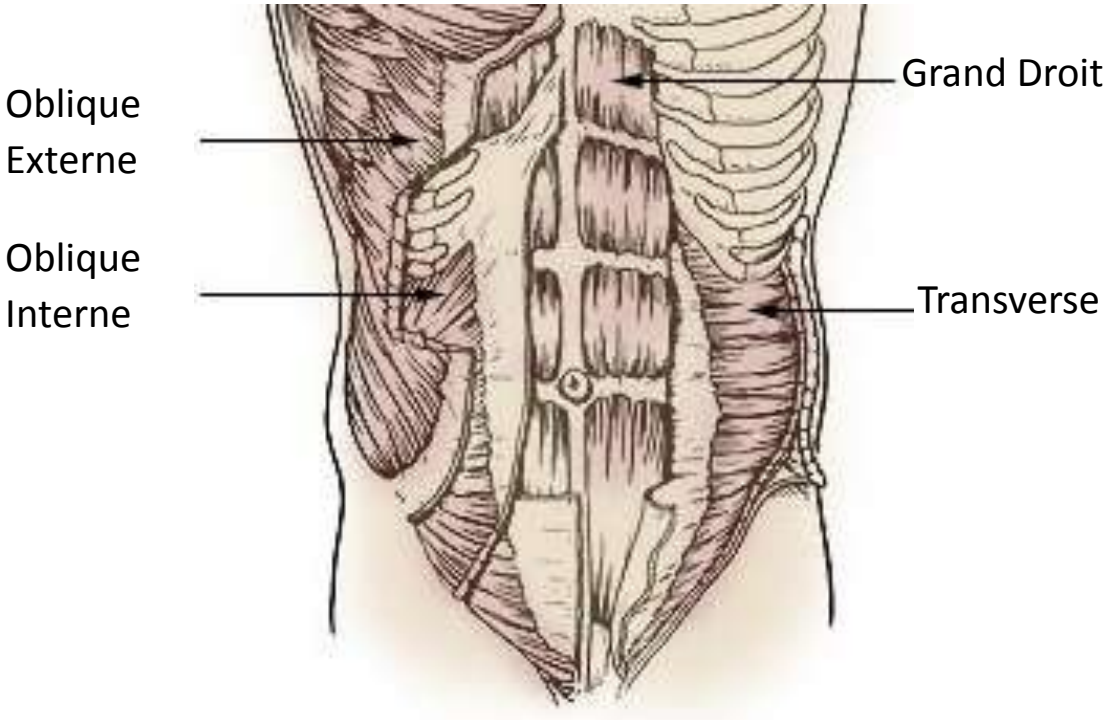
Les scalènes : muscles inspiratoires primaires

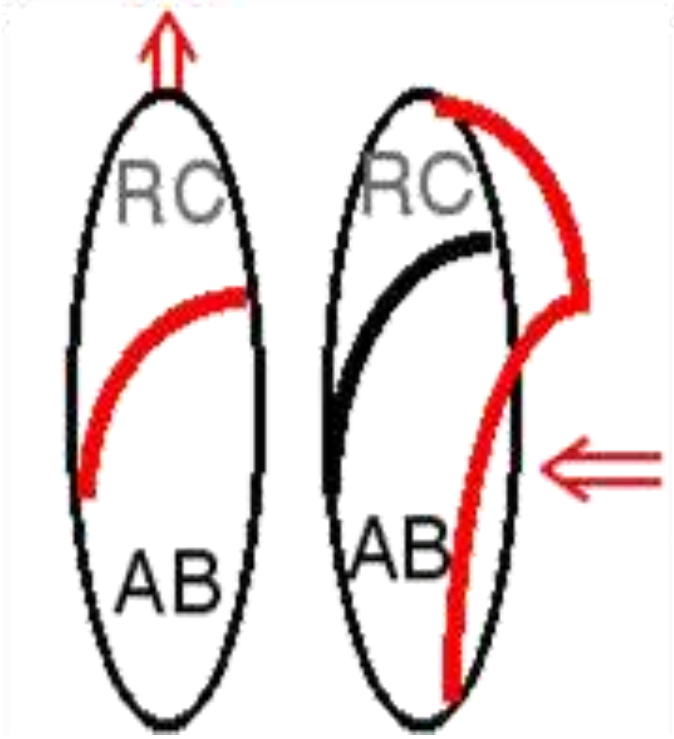
Les sterno-cléido-mastoïdiens : accessoires



Les muscles expiratoires

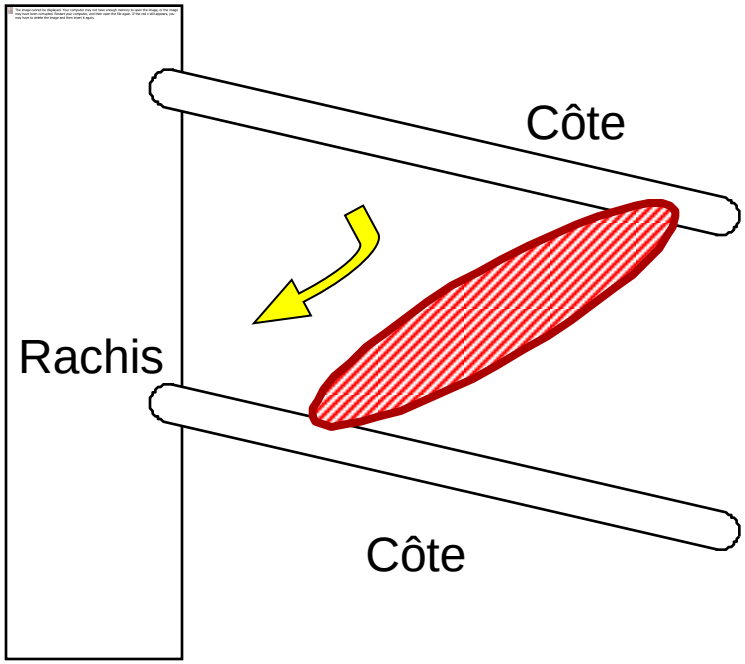
1. Les abdominaux



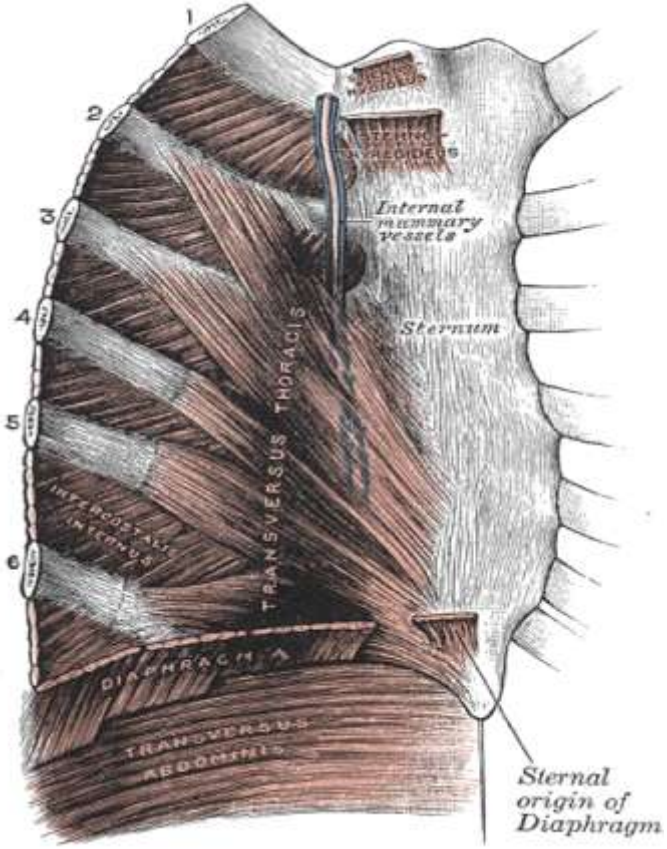


Les muscles expiratoires
2. Les intercostaux

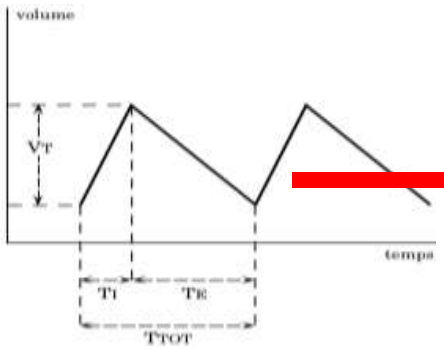
Intercostal Interne



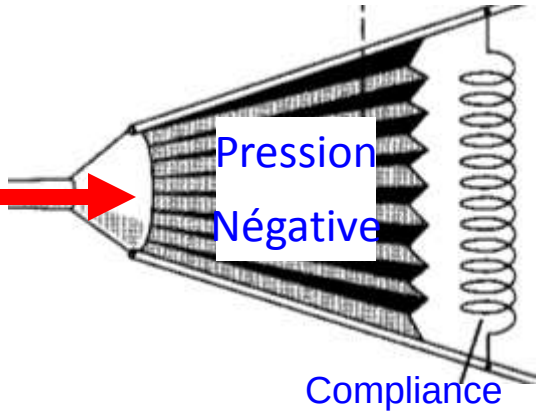
Triangulaire du sternum



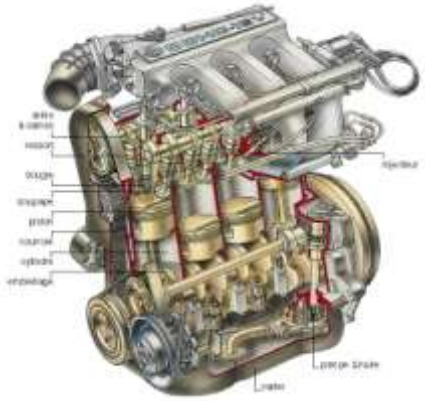
L'inspiration de repos

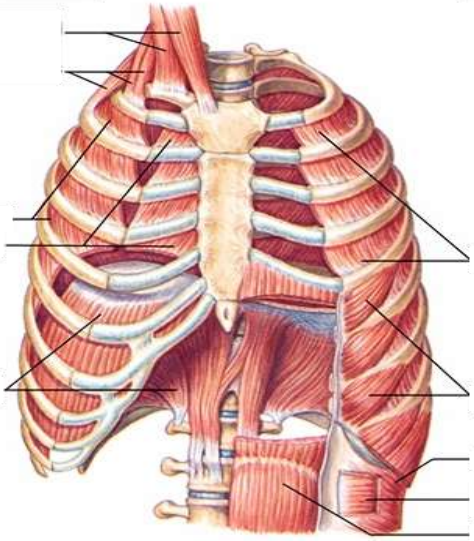


Structures passives

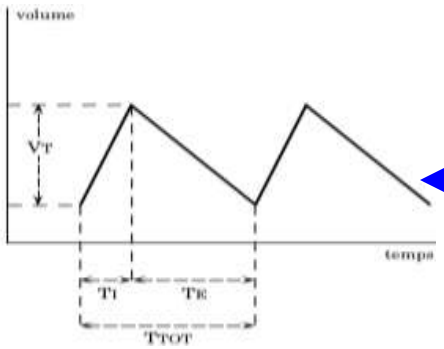


Structures actives

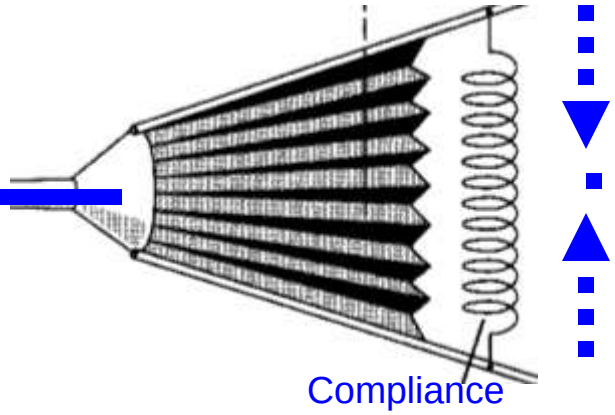




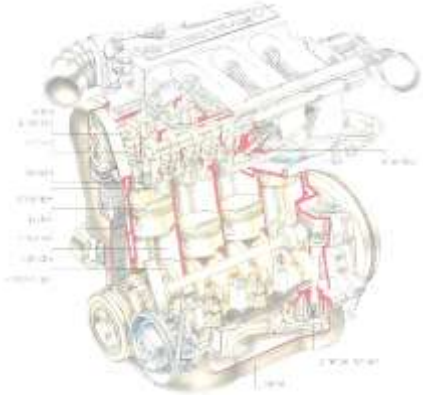
L'expiration de repos

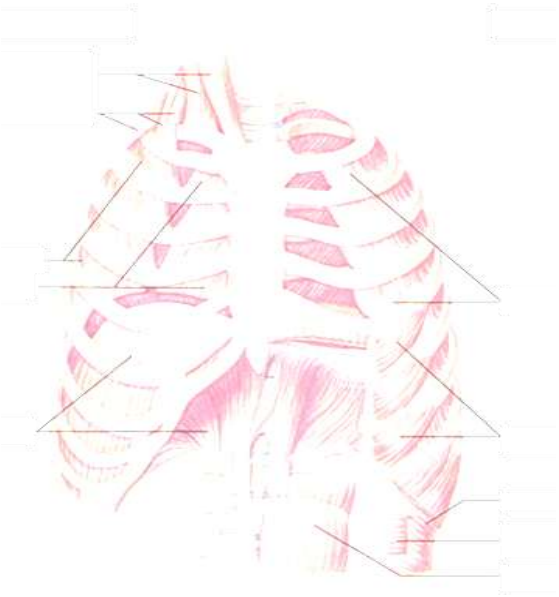


Structures passives

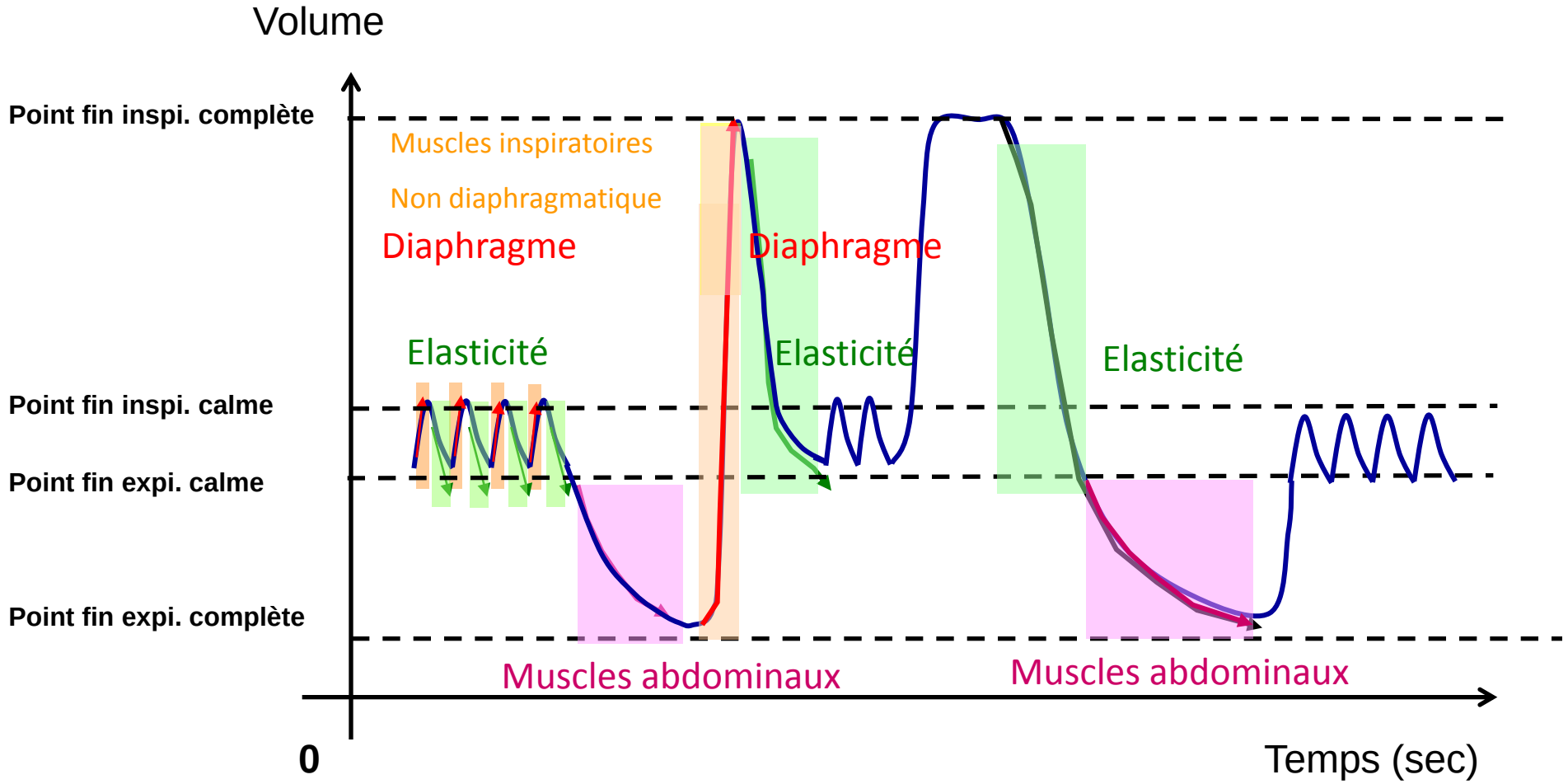


Structures actives





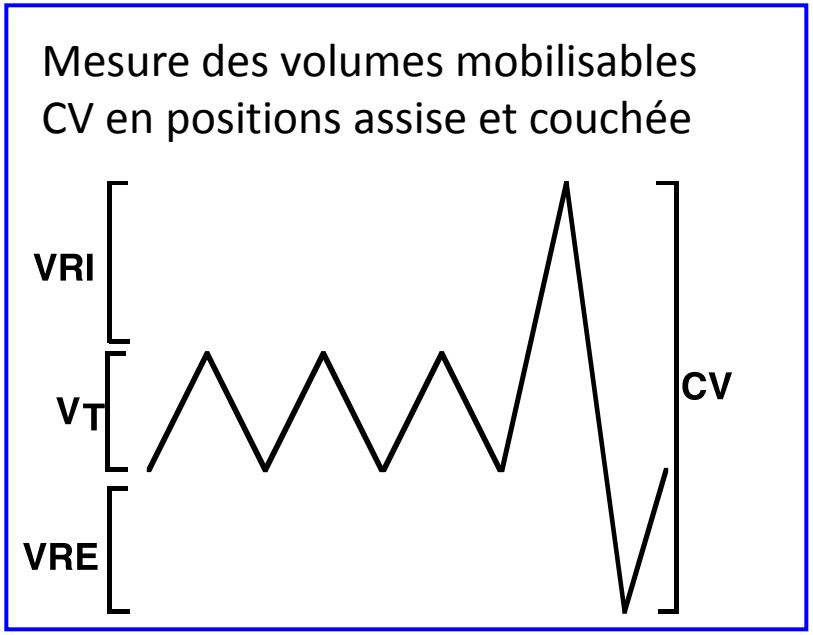
Muscles et cycle ventilatoire



Mise en jeu muscles respiratoires pour atteinte VR et CPT

Diapo du Dr Laveneziana

Muscles ventilatoires et volumes mobilisables



Défaillance musculaire respiratoire => diminution de CV (peu sensible)

Chute de CV de 15% en position couchée => défaillance diaphragmatique (peu sensible, peu spécifique)

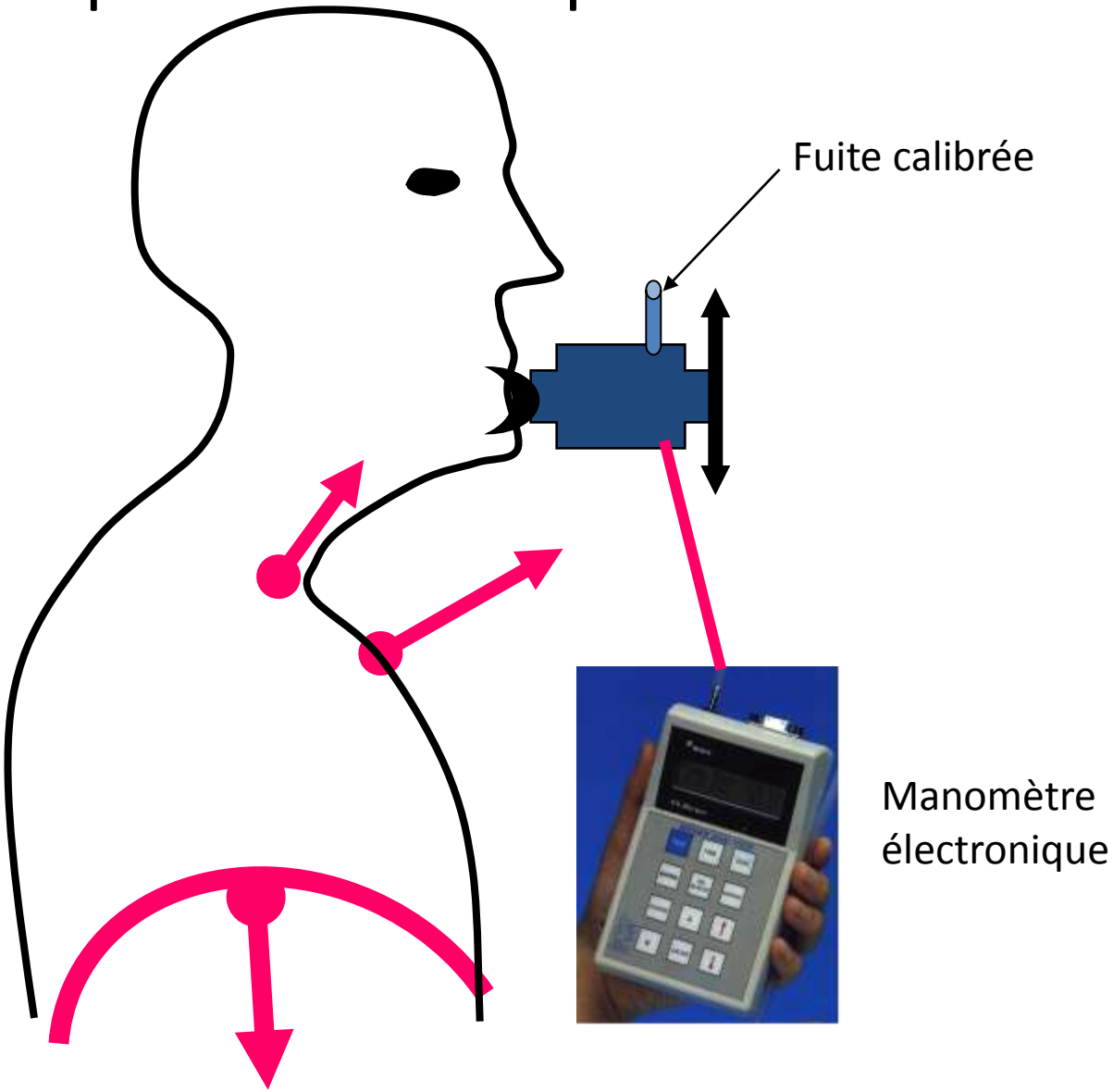
Muscles ventilatoires et échanges gazeux

$$PaCO_2 = \alpha \, k \, \frac{\dot{V}_{CO_2}}{\dot{V}_E * \left(1 - \frac{V_D}{V_T}\right)}$$

- hypercapnie
- peu sensible
- dépent de la production de CO2
- inadaptation ventilatoire à l ’ effort, fièvre...

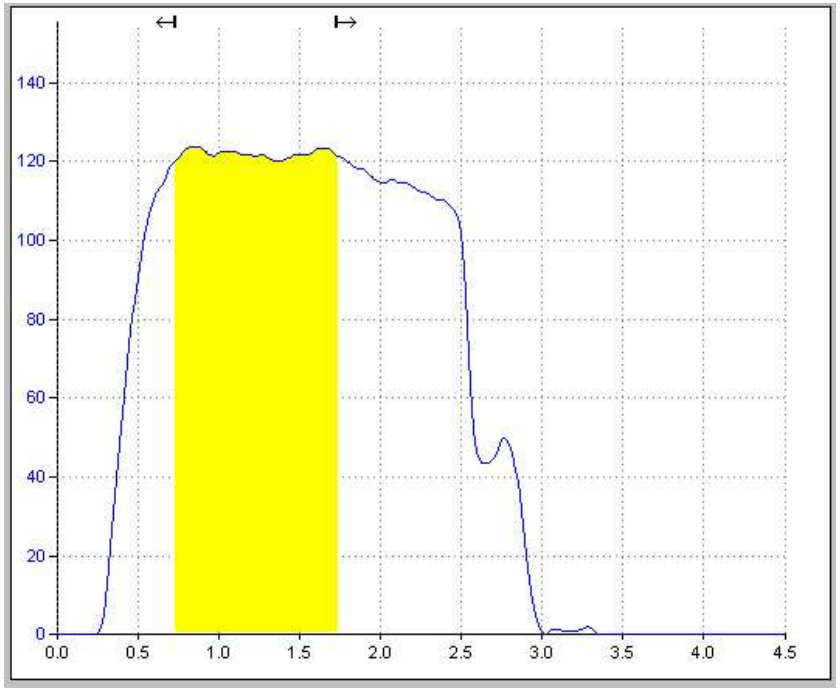
Pressions produites par les muscles respiratoires

Mesure des pressions statiques maximales



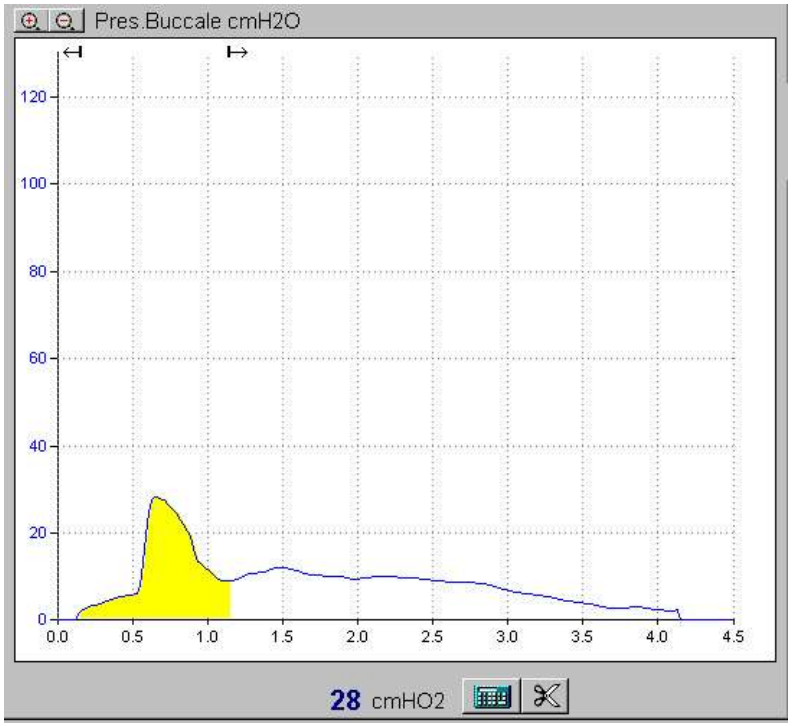
Pressions statiques

cm H2O



Normale

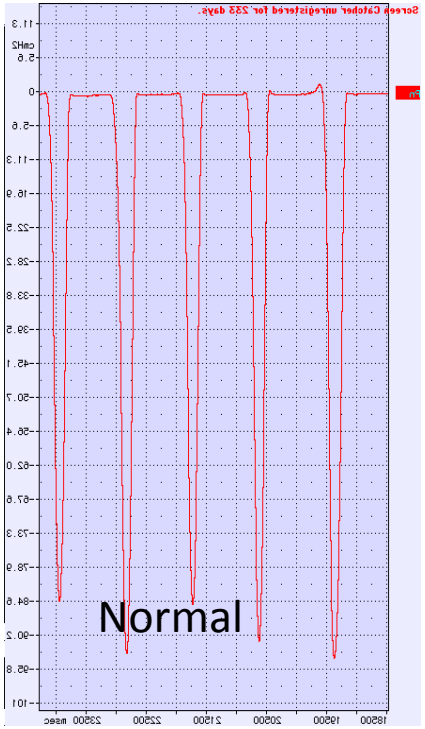
cm H2O

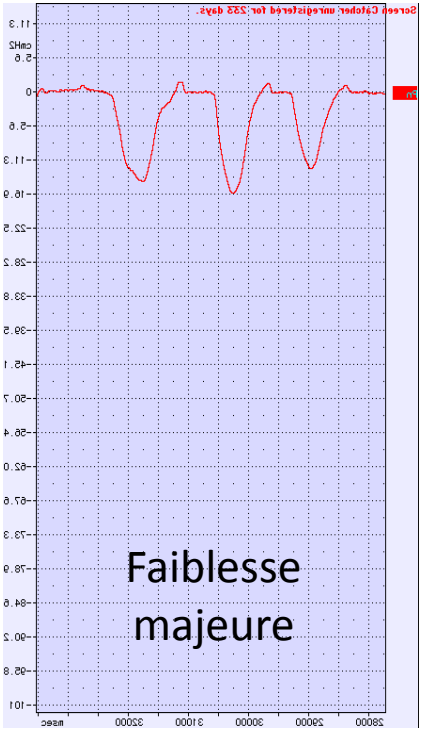


Faiblesse inspiratoire sévère

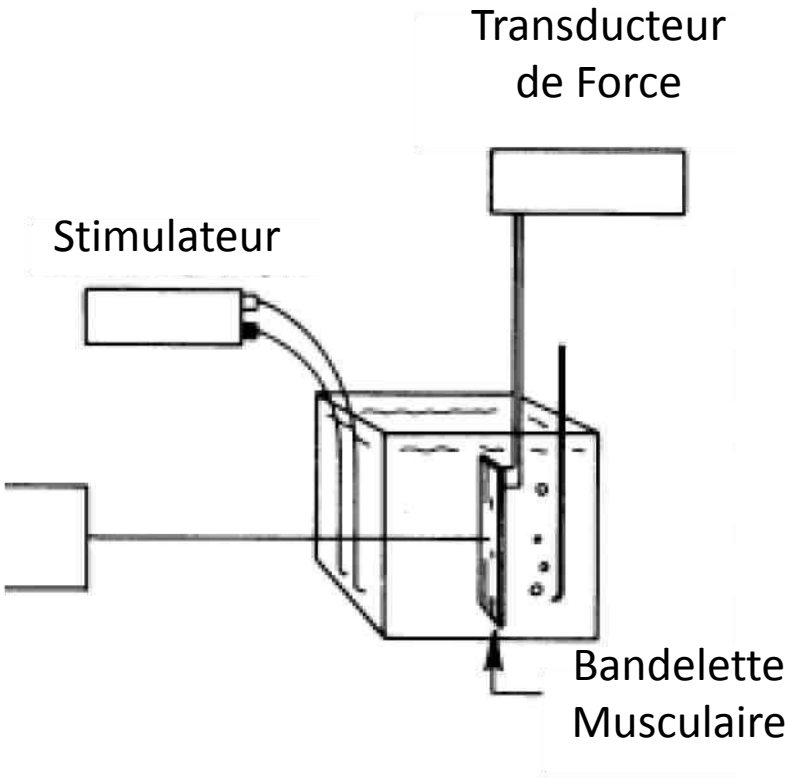
Pression nasale de reniflement maximal

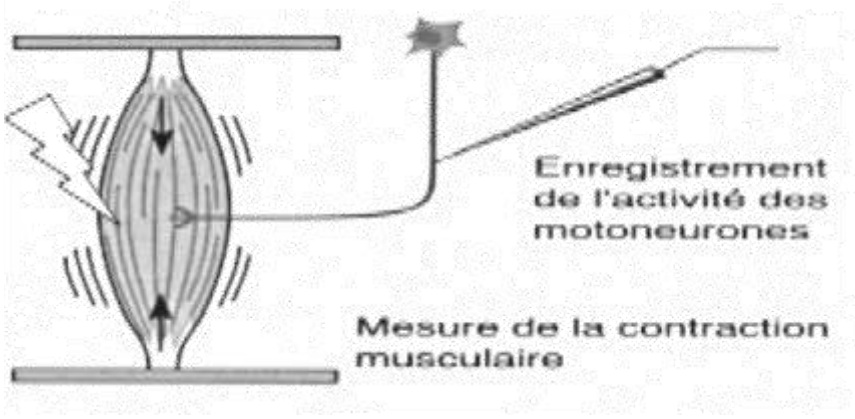
SNIP *(sniff nasal inspiratory pressure)*



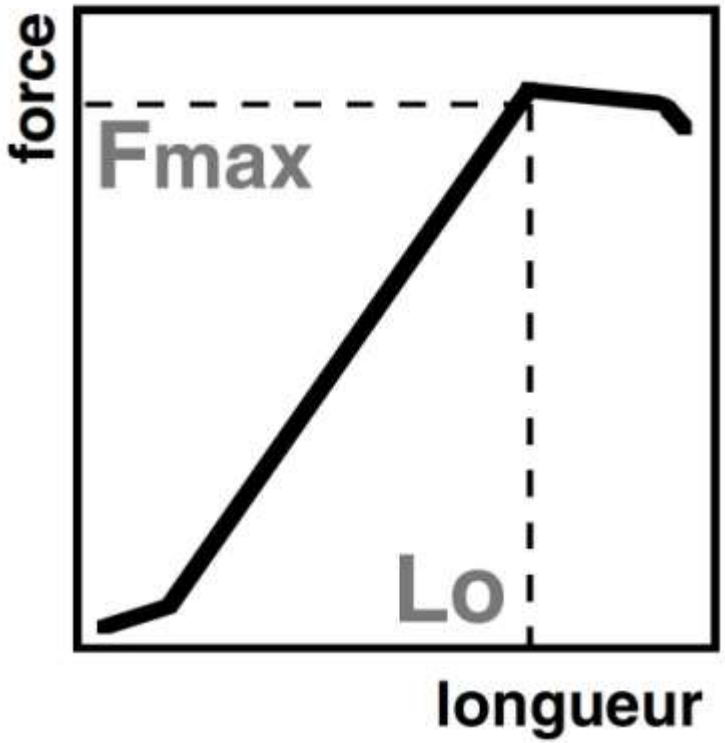


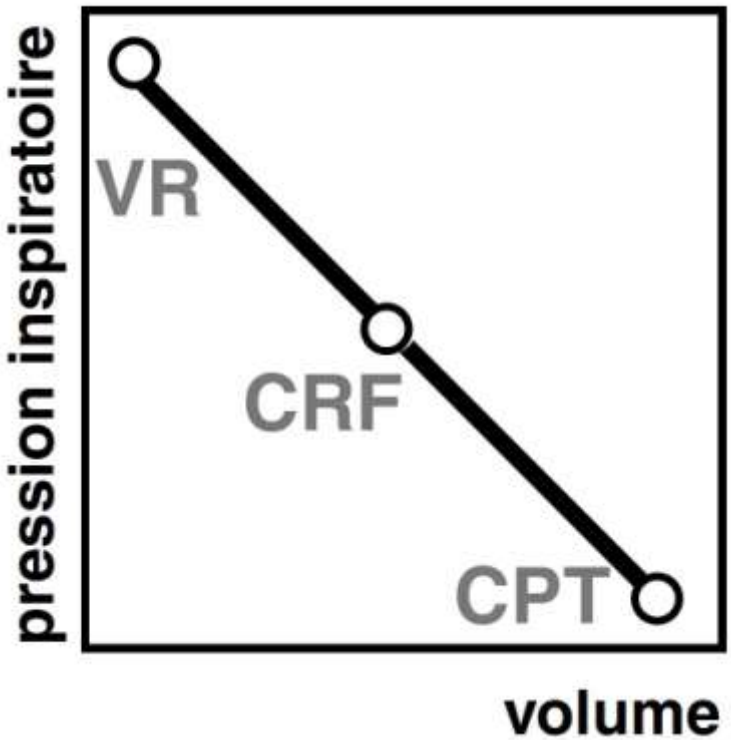
Etude des fibres musculaires, in vitro



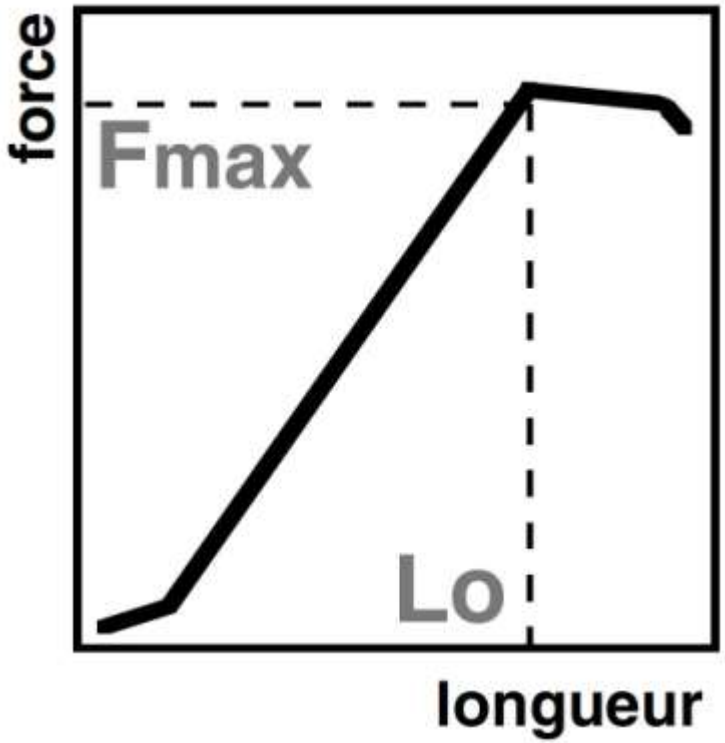


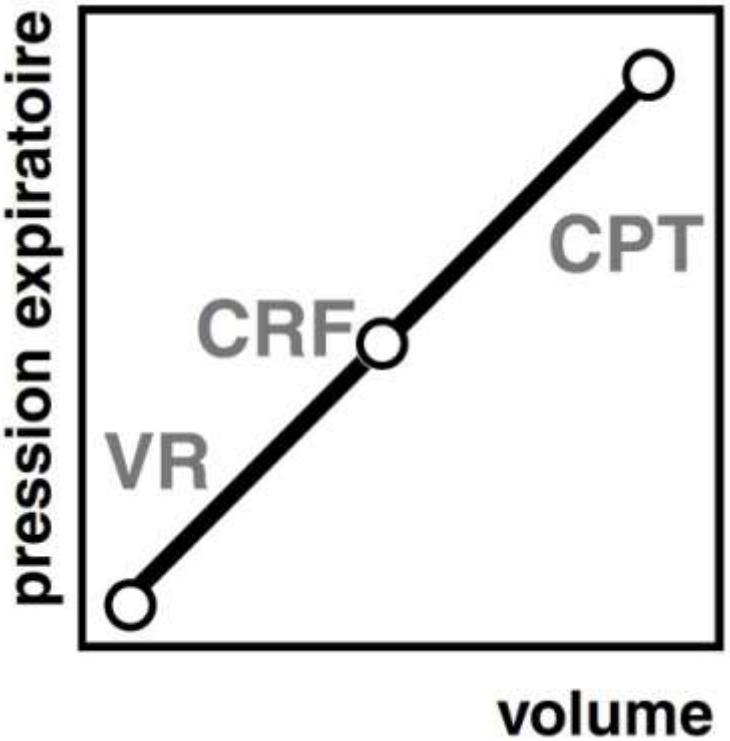
Relation force-longueur des muscles inspiratoires





Relation force-longueur des muscles expiratoires

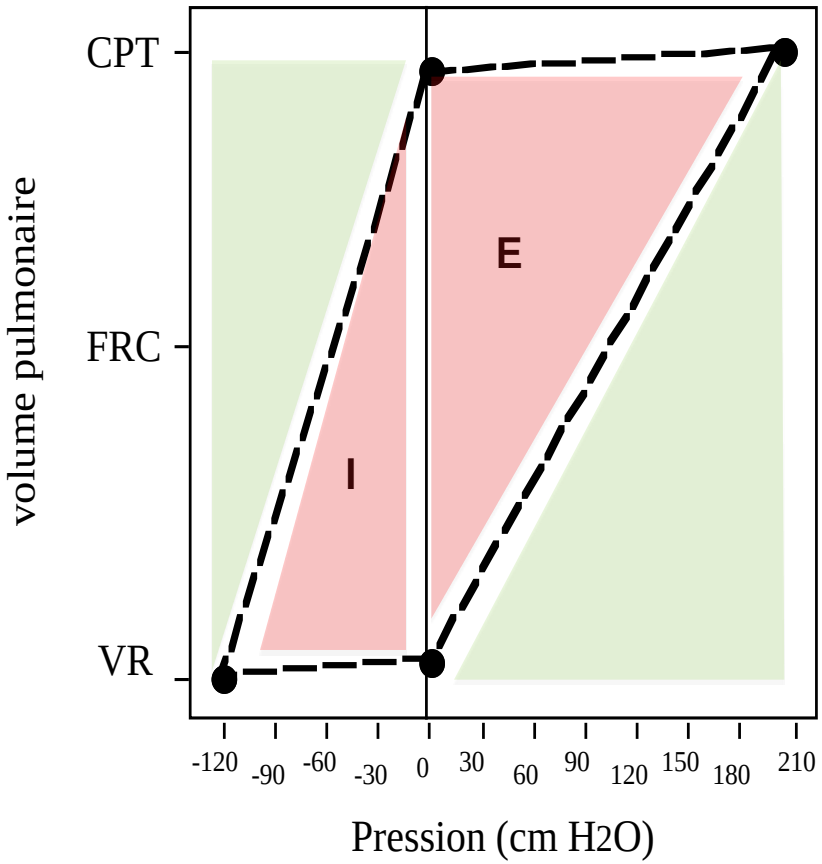




Relation pression-volume des muscles respiratoires

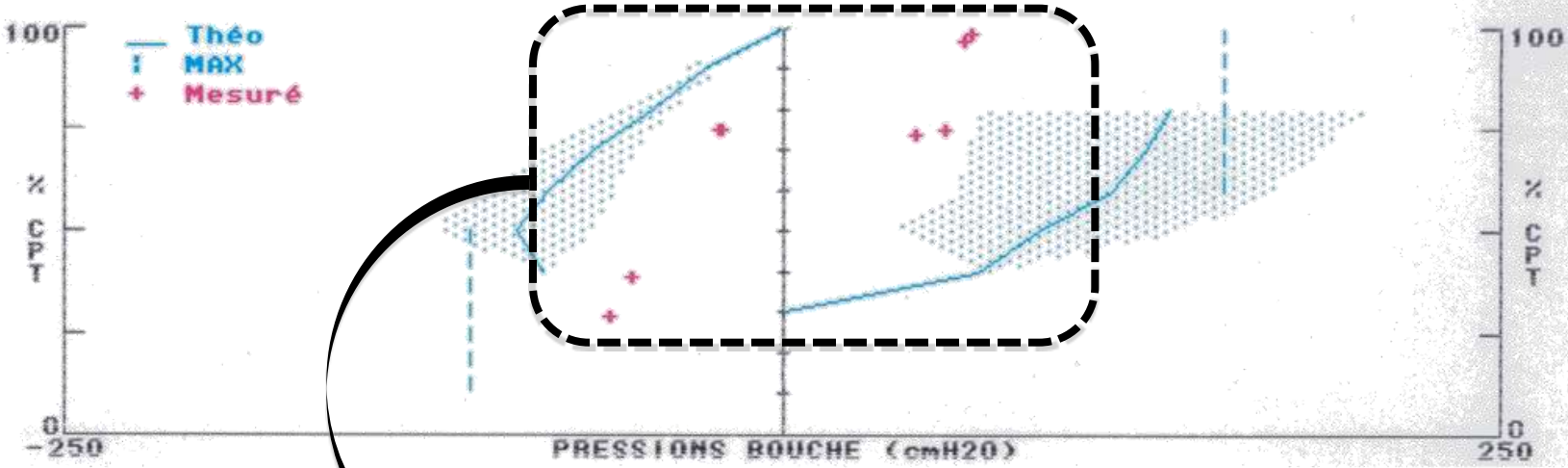
Zone normale

Zone pathologique



Relation pression-volume des muscles respiratoires

	VOLUME (L)	%CPT	PRESSIONS BOUCHE (cmH2O) Théorique	Mesuré
INSPIRATION				
	1.37	29	0 (+/-)	61
	1.81	39	84 (+/-)	53
	3.50	75	46 (+/-7)	22
	3.50	75	46 (+/-7)	23
EXPIRATION				
	4.53	97	153 (+/-63)	63
	3.46	74	126 (+/-61)	46
	3.51	75	135 (+/-67)	56
	4.58	99	153 (+/-63)	65



Les mesures sont dans la zone pathologique

Mesure des pressions statiques maximales

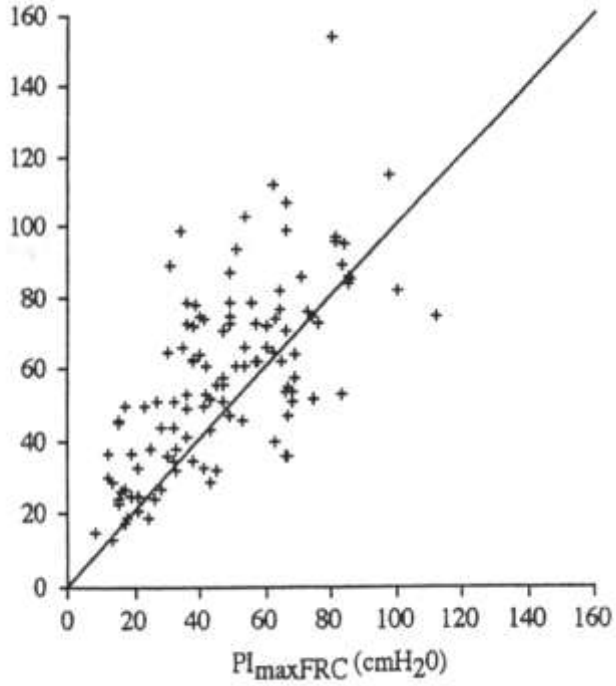
- Inspiratoire ; expiratoire
- effort > 1,5 sec
- valeur : moyenne pendant la 1ère seconde
- Toujours au même volume :
  - **VR** ou CRF pour PImax
  - **CPT** ou CRF pout Pemax
- valeurs normales fréquentes
  - PIMax < - 80 cmH_2O
  - PEMax > 120 cmH_2O

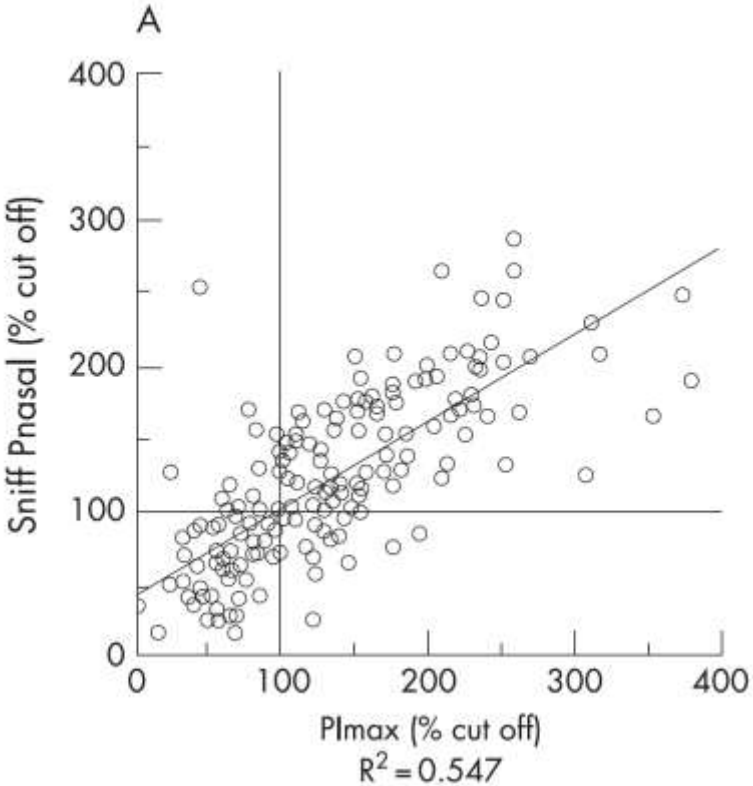
Mesure de la pression de reniflement nasale max

- Facile
- CRF
- Plusieurs muscles INSPIRATOIRES
- TOUJOURS complémentaire de Pimax
- Reniflement maximal bref < 500 ms
- 10 à **20** mesures
- Reproductibilité < 5 %

Corrélation SNIP-Pimax en pathologie neuromusc.

SNIP
cm H2O

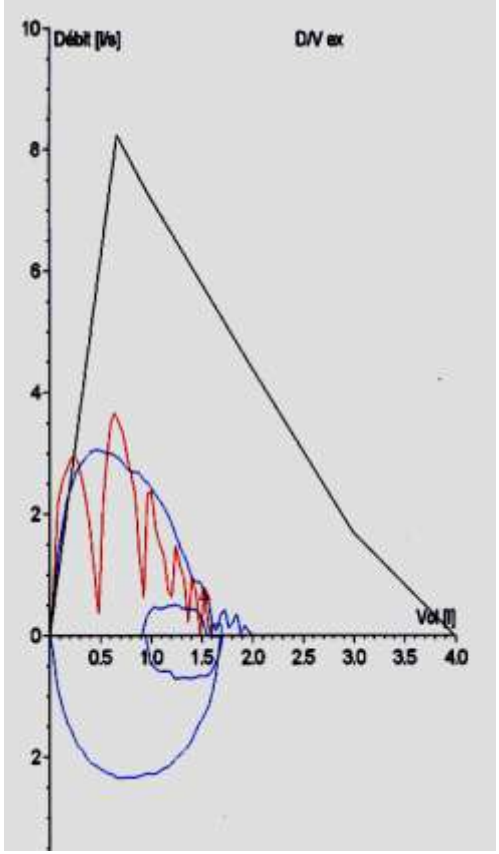




Stefanutti, AJRCCM, 2000

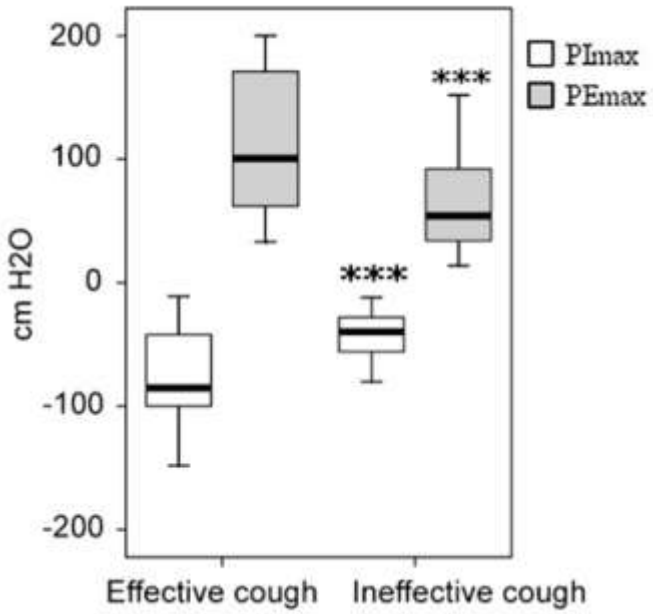
Steier, Thorax, 2007

DEP à la toux : évaluation des muscles expiratoires

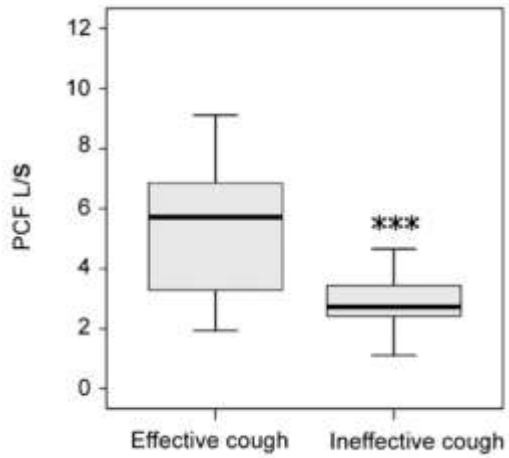


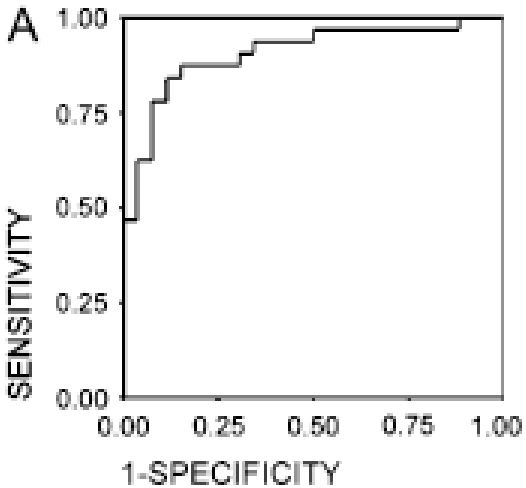
- Assis
- Toux après inspiration complète
- 3-6 manœuvres avec moins de 5% de variabilité.
- Efficacité de la toux :
  - Sains : facilement 400 – 600 L/min
  - DEP toux < 270 L/min associé à un risque accru de complications respiratoires dans les maladies neuro musculaires (MNM).
  - DEP toux < 160 L/min associé à un risque accru d'échec de sevrage dans les MNM.

Toux dans la Sclérose Latérale Amyotrophique



DEP à la Toux



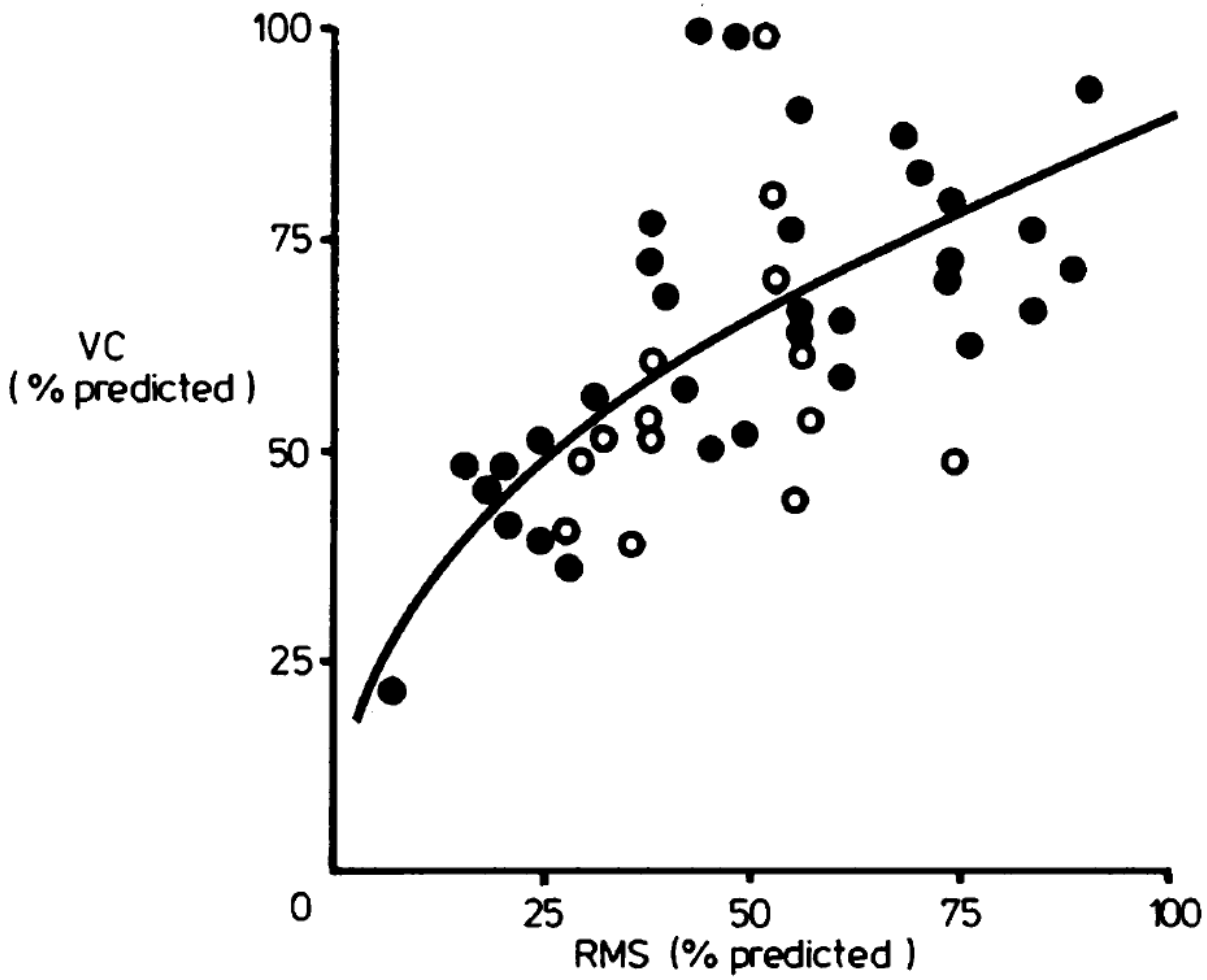


Prédiction d'une toux inefficace en cas d'infection respiratoire

Cut off 4,25 L/s (255 L/min)

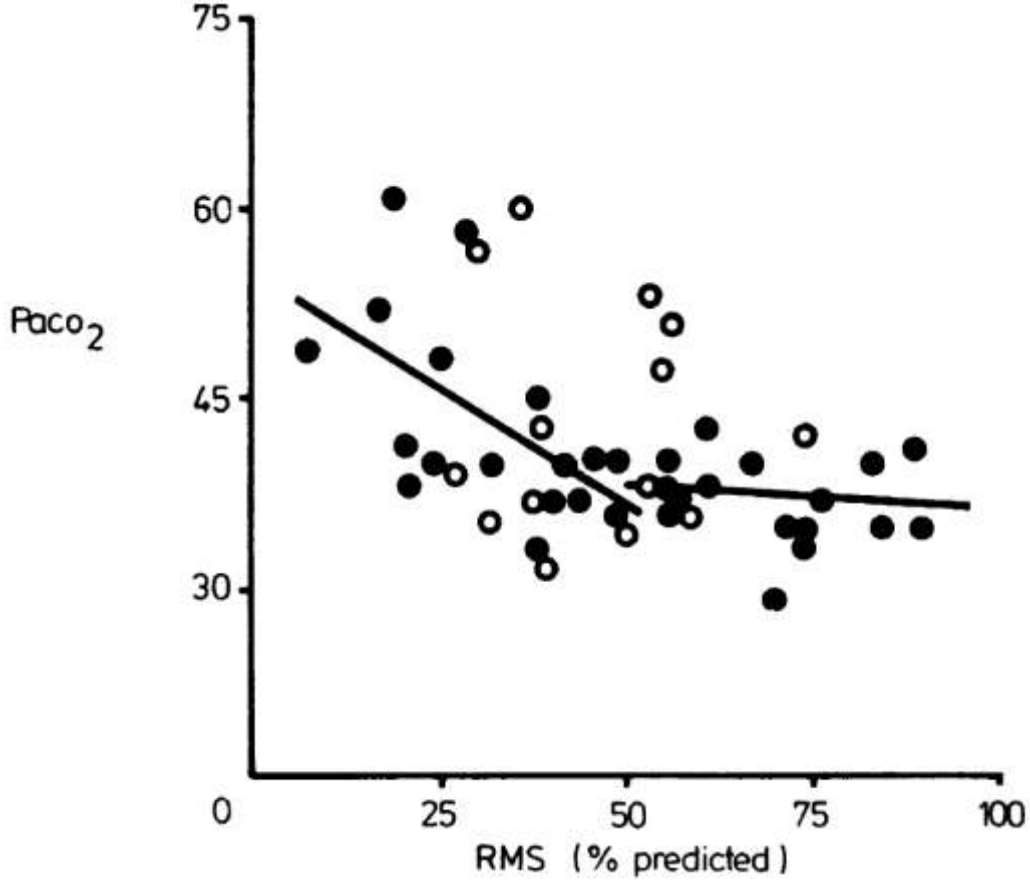
Sancho, AJRCCM, 2007

Lien log entre Pimax-CV en pathologie neuromusc.



Braun, Thorax, 1983

Lien PaCO2-Force des muscles respiratoires
Polymyosites et myoapathies

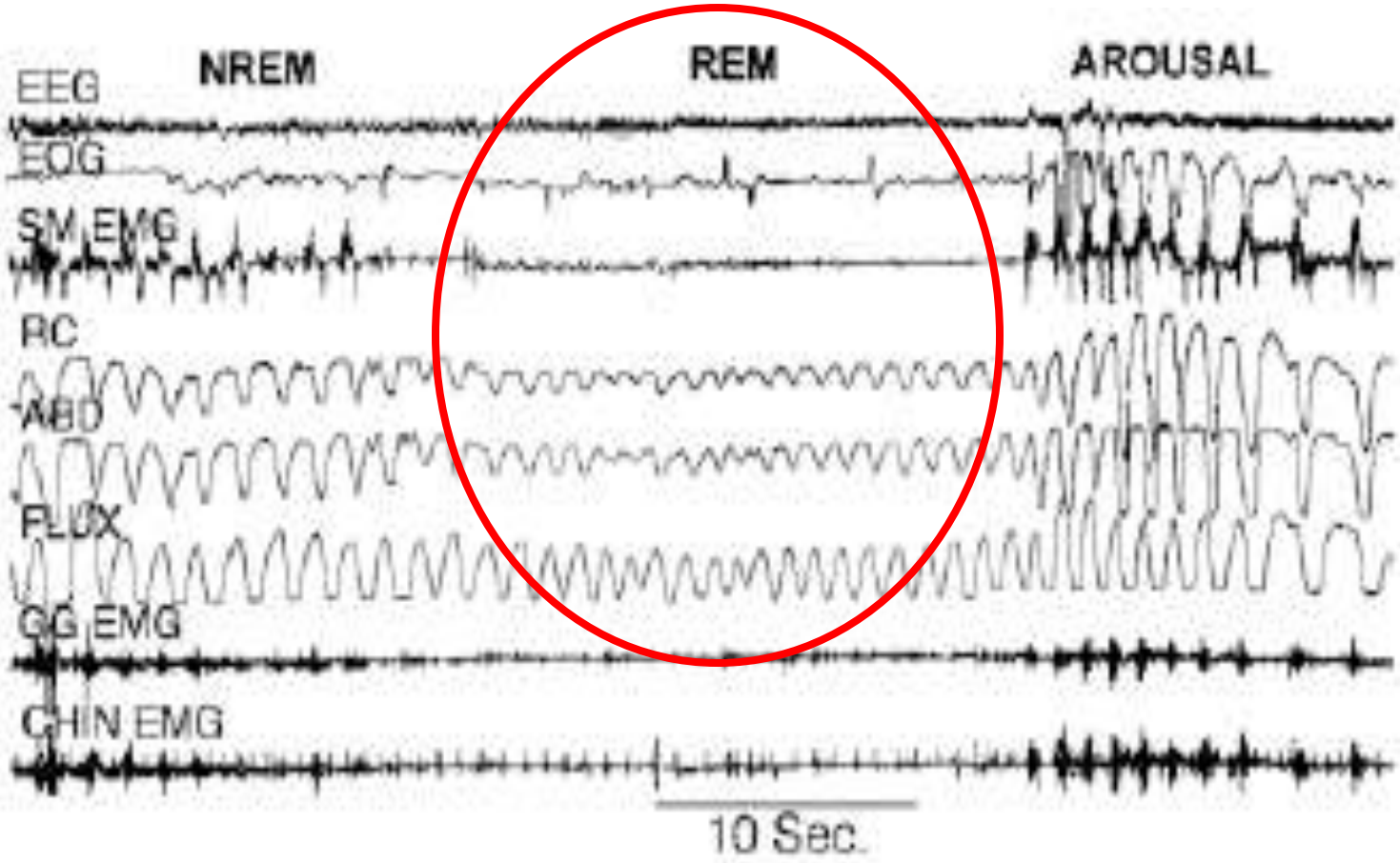


Braun, Thorax, 1983

Défaillance des muscles respiratoires

Sommeil +++

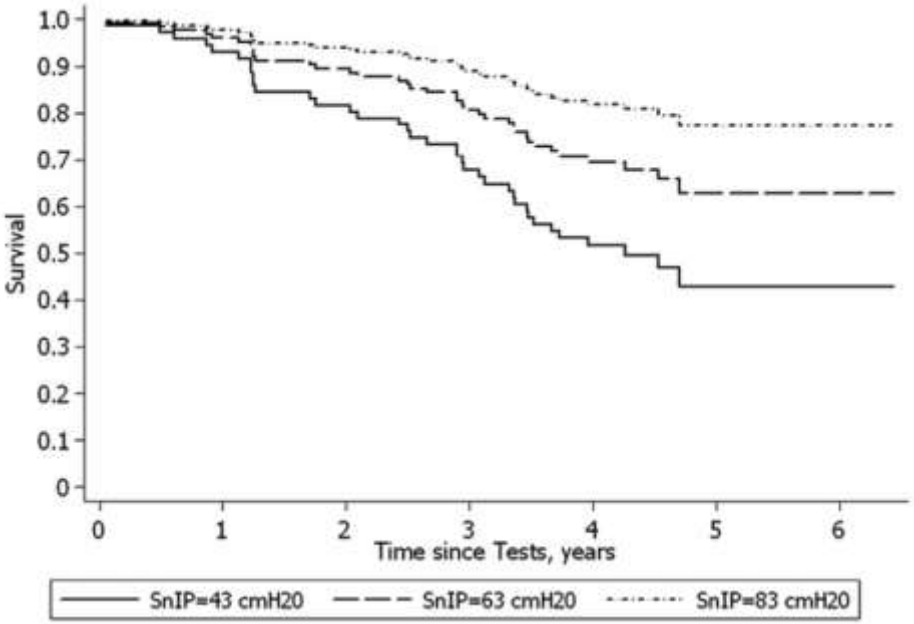
Polysomnographie



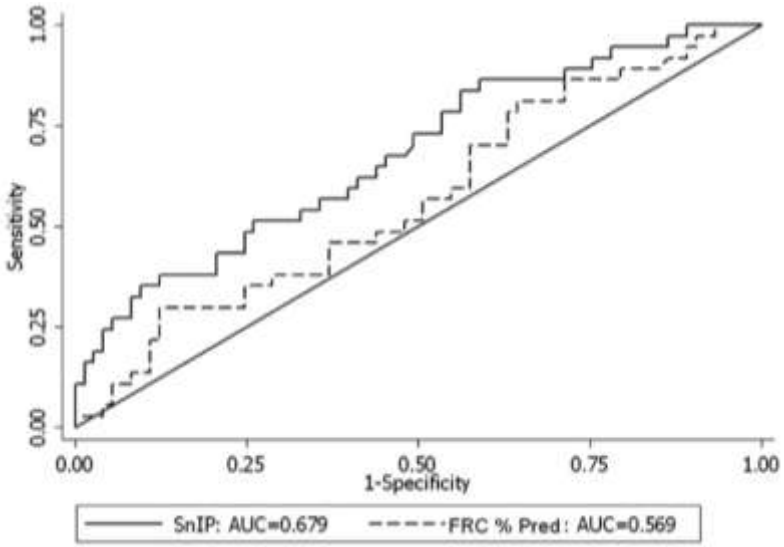
Arnulf et al., Am J Respir Crit Care Med, 2000

BPCO : pronostic

SNIP

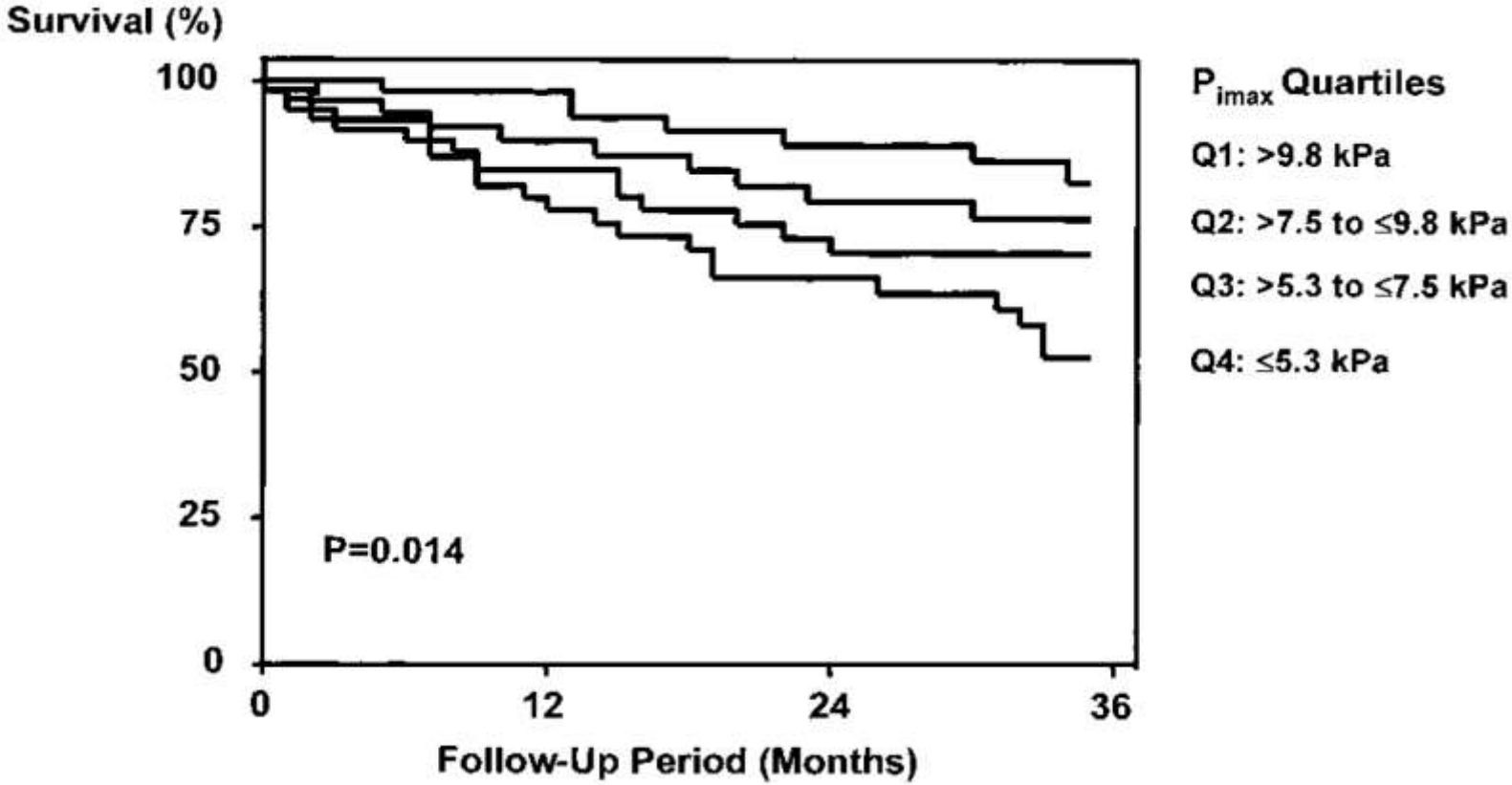


SNIP/CRF : prédiction de la mortalité



Moore et coll. Respir Med 2010

PImax



Meyer et coll. Circulation 2001

Guillain-Barré : facteur prédictif de ventilation mécan.

- **A l'Admission**

Sans mesure de la CV (risque > 85 % si 4 critéres présents)

1. Début des signes avant admission < 7 jours
2. Toux inefficace
3. Impossibilité de tenir debout
4. Impossibilité de soulever les coudes
5. Impossibilité de relever la tête
6. Cytolyse hépatique

Avec mesure de la CV (risque > 85 %si les 3 critères présents)

1. Début des signes avant admission < 7 jours)
2. Impossibilité de relever la tête
3. **CV < 60 % de la théorique**

- **Pendant le séjour en Réanimation**

Troubles de déglutition

CV < 20 ml/kg

D. Orlikowski, Réanimation, 2005

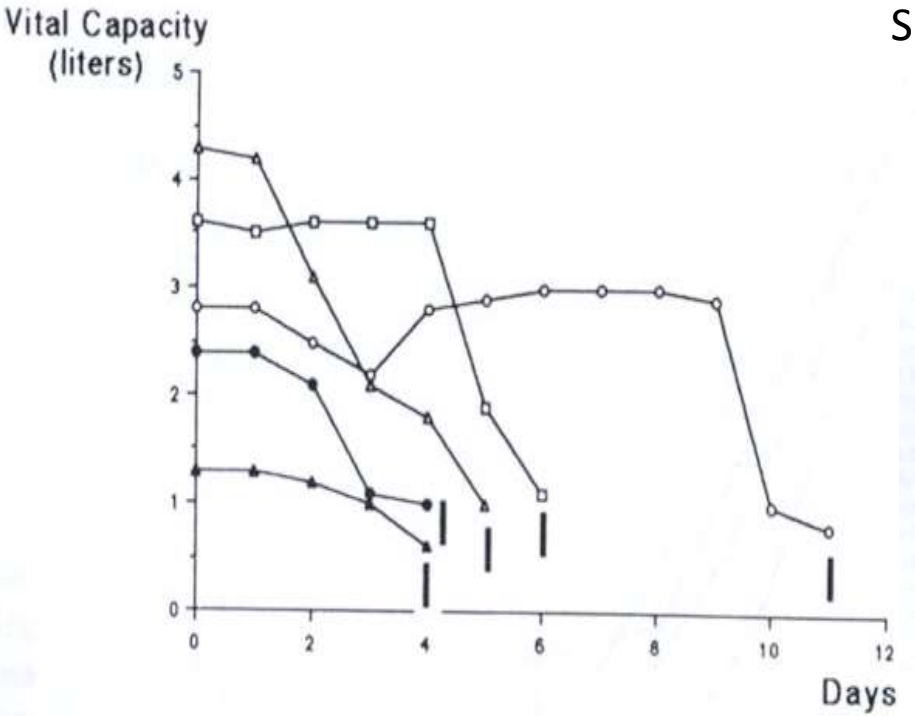
Critères d'intubation du Guillain-Barré

- Détresse respiratoire
- **$PaCO_2$ > 48 mmHg ou PaO_2 < 56 mmHg**
- **CV < 15 ml/kg**
- **PI_{max} <- 25 cmH_2O**
- **PE_{max} < 40 cmH_2O**

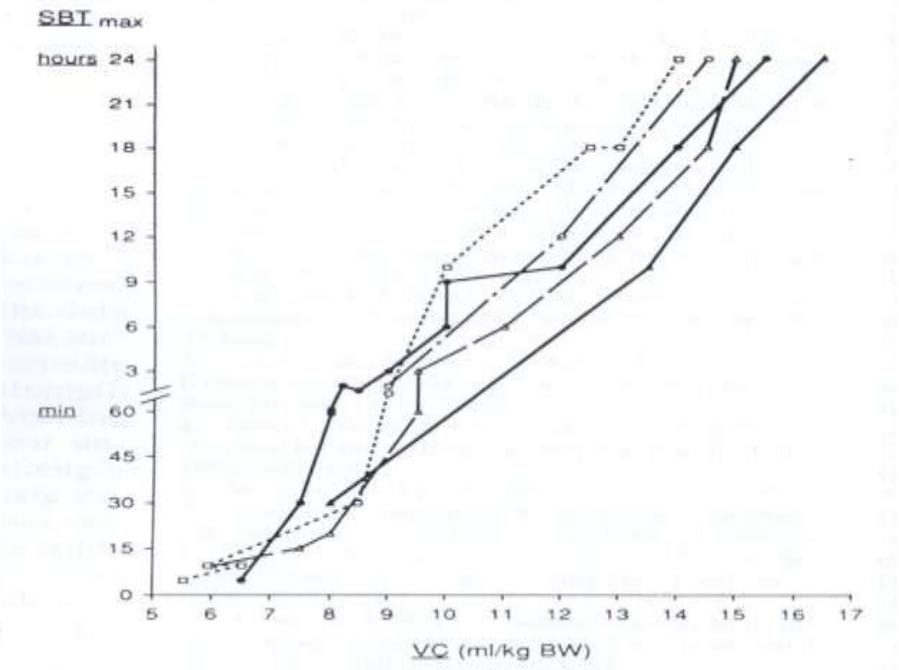
*Un seul critère suffit

D. Orlikowski, Réanimation, 2005

Capacité Vitale dans le Guillain-Barré



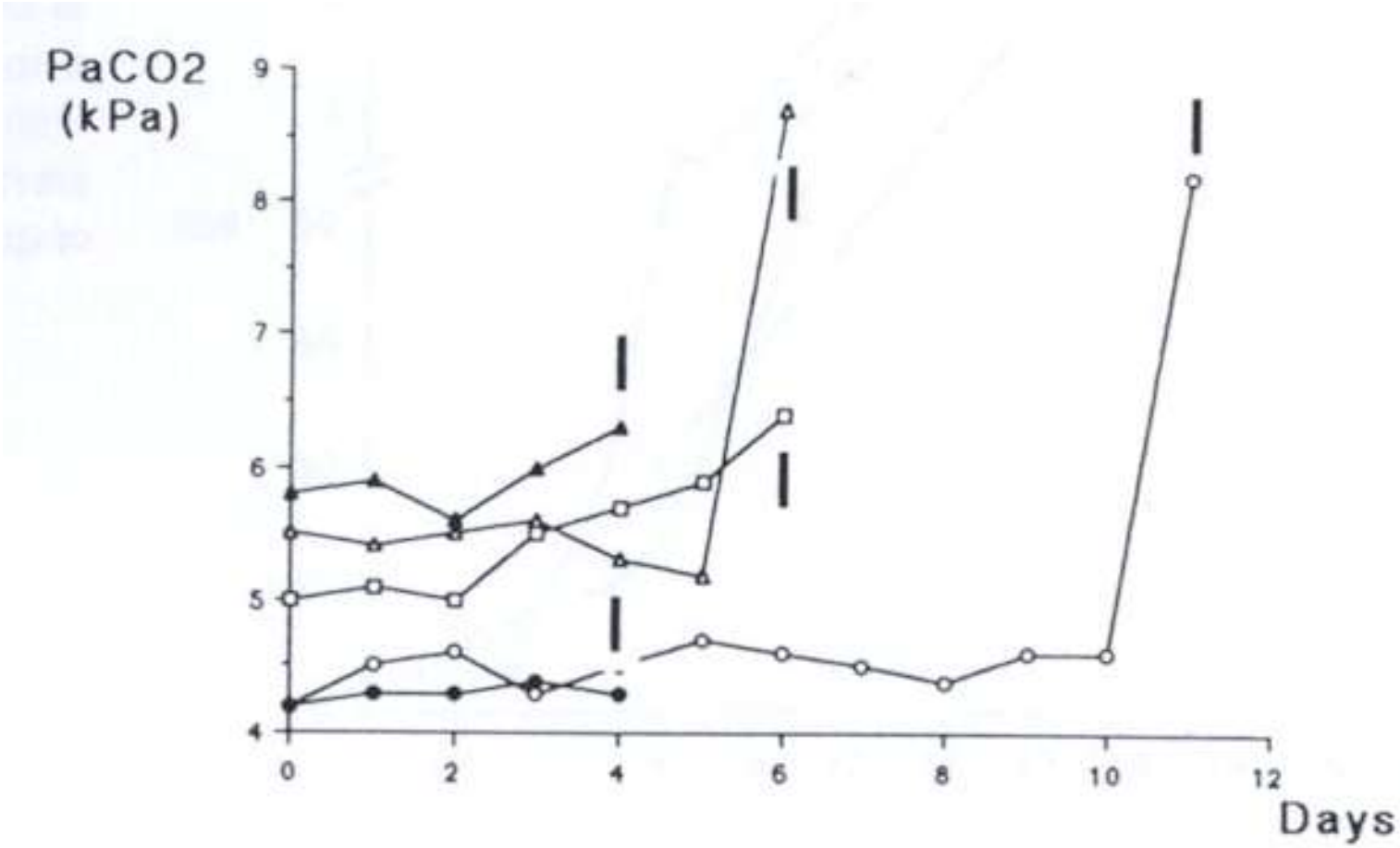
SBT = Spontaneous Breathing time



I = intubation

Chevrolet et coll. Am rev Resp Dis 1991

$PaCO_2$ dans le Guillain-Barré (index tardif)

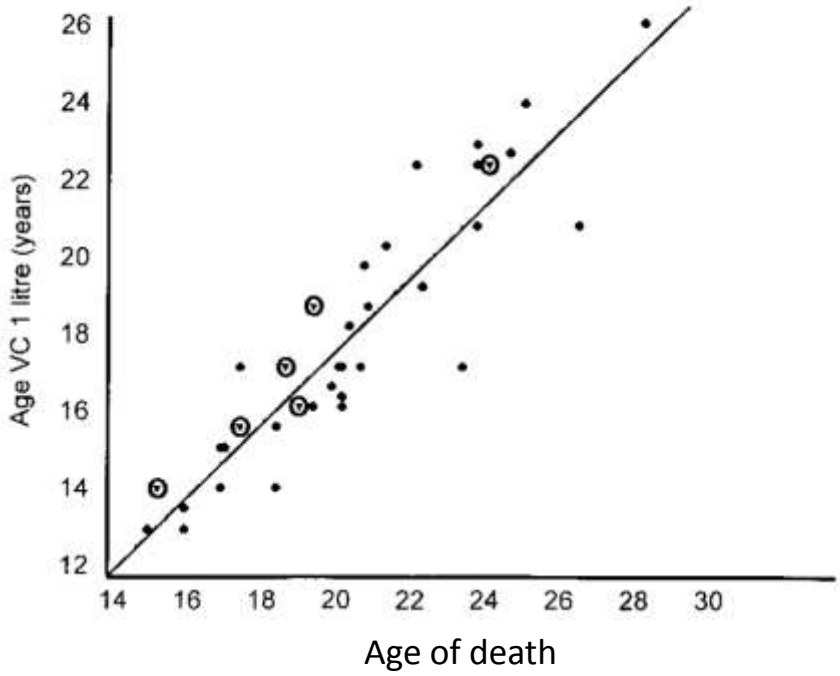


I = intubation

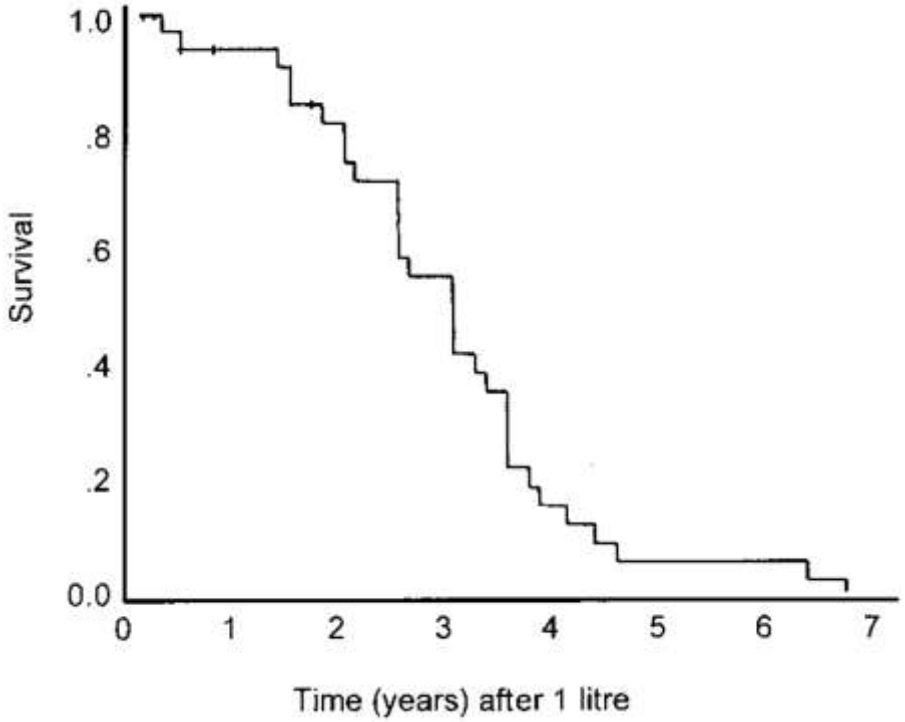
Chevrolet et coll. Am rev Resp Dis 1991

Duchenne de Boulogne : pronostic

Age CV 1L vs. Age décès

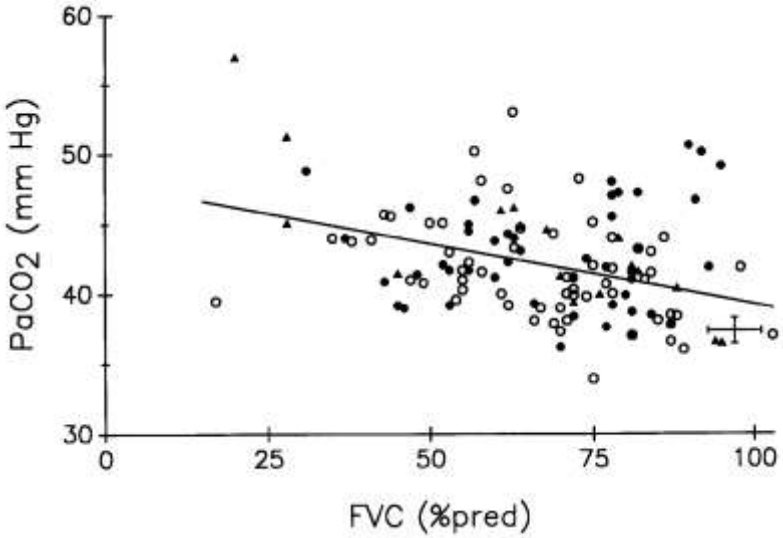


Survie à partir de CV = 1L

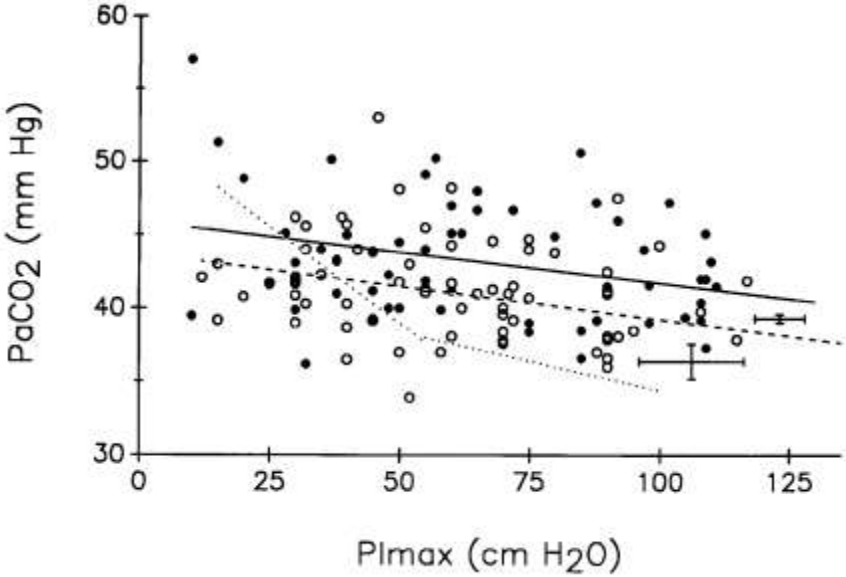


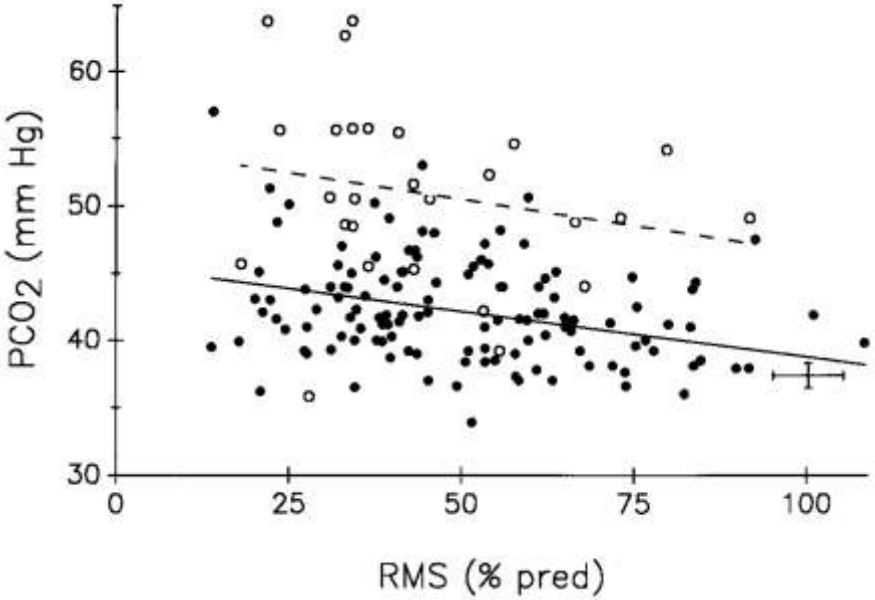
Phillips et coll. Am J Respir Crit Care Med 2001

Steinert : muscles et PaCO2



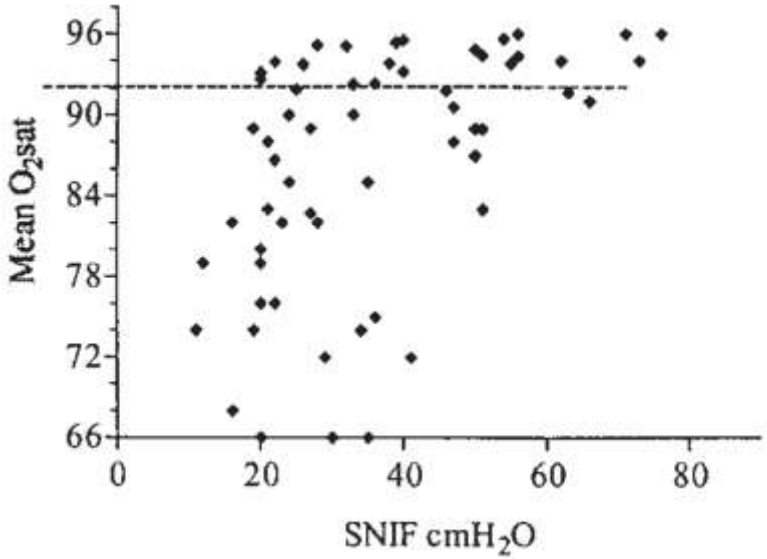
Lien entre PaCO2 et Capacité Vitale, PImax et « force des muscles respiratoires »



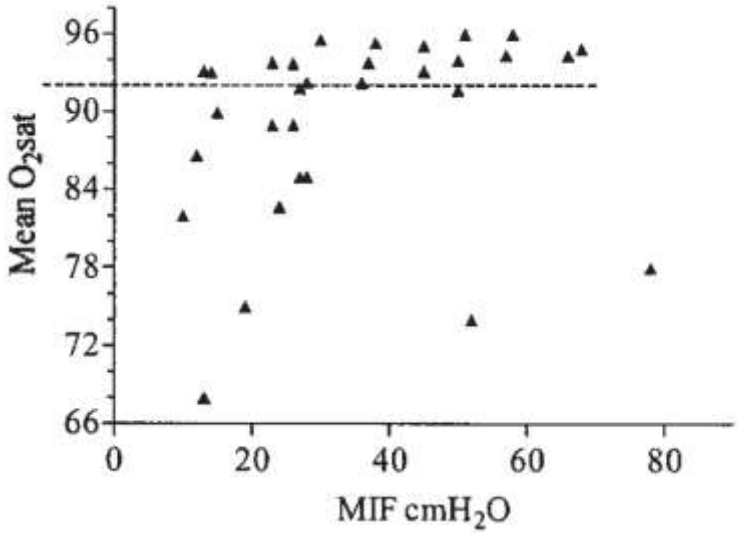


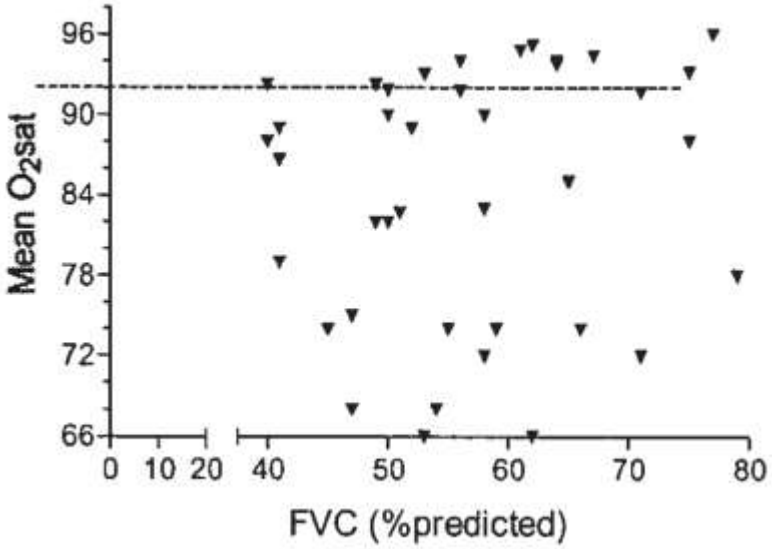
Bégin et coll. Am J Respir Crit Care Med 1997

Sclérose Latérale Amyotrophique : pronostic



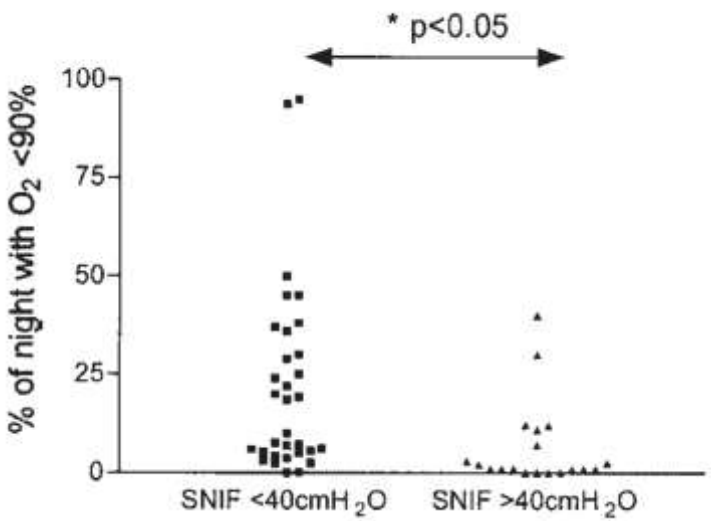
SpO2 pendant le sommeil liée à SNIP et Pimax, pas à VC



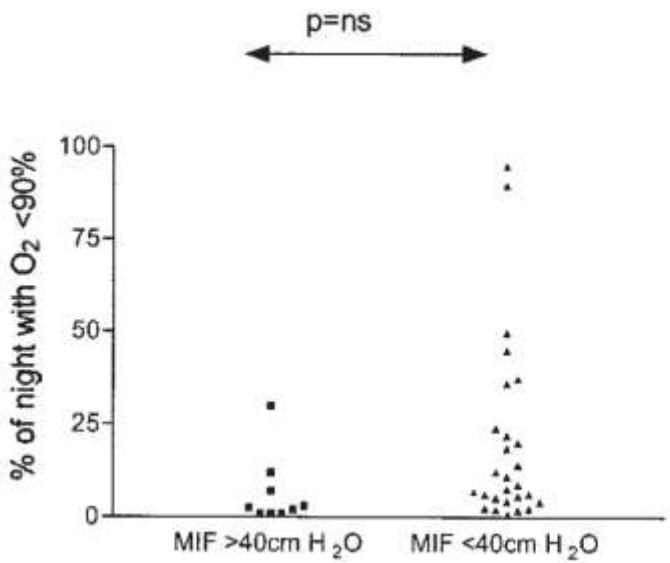


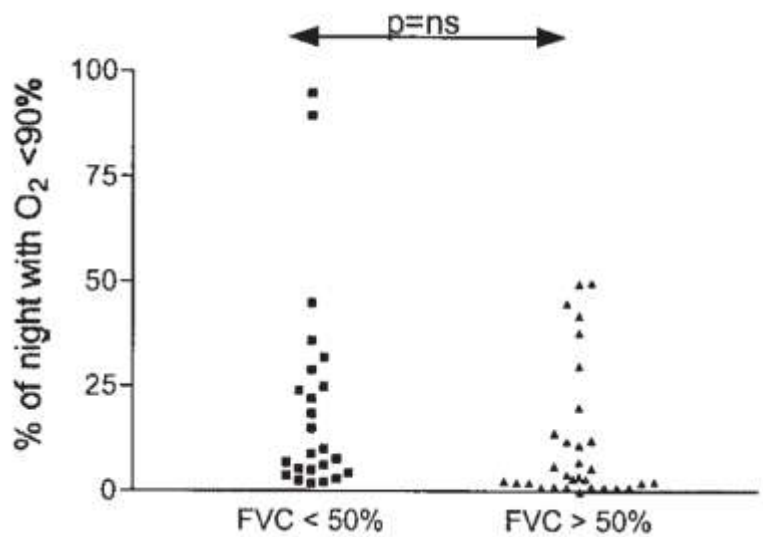
Morgan et coll. Am J Respir Crit Care Med 2005

Sclérose Latérale Amyotrophique : pronostic



Désaturation pendant le sommeil liée à SNIP < 40 cmH2O



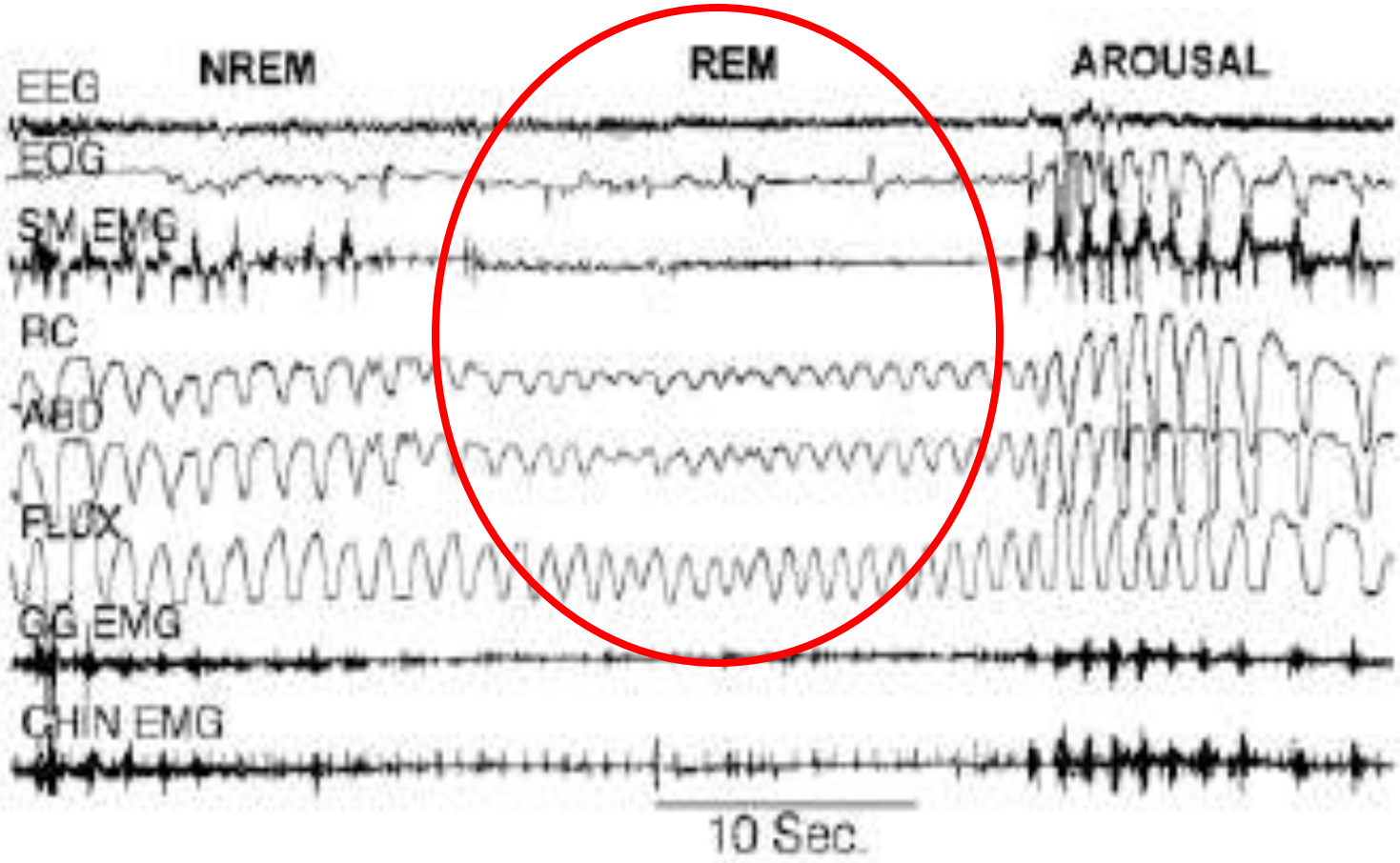


Morgan et coll. Am J Respir Crit Care Med 2005

SLA et dysfonction diaphragmatique

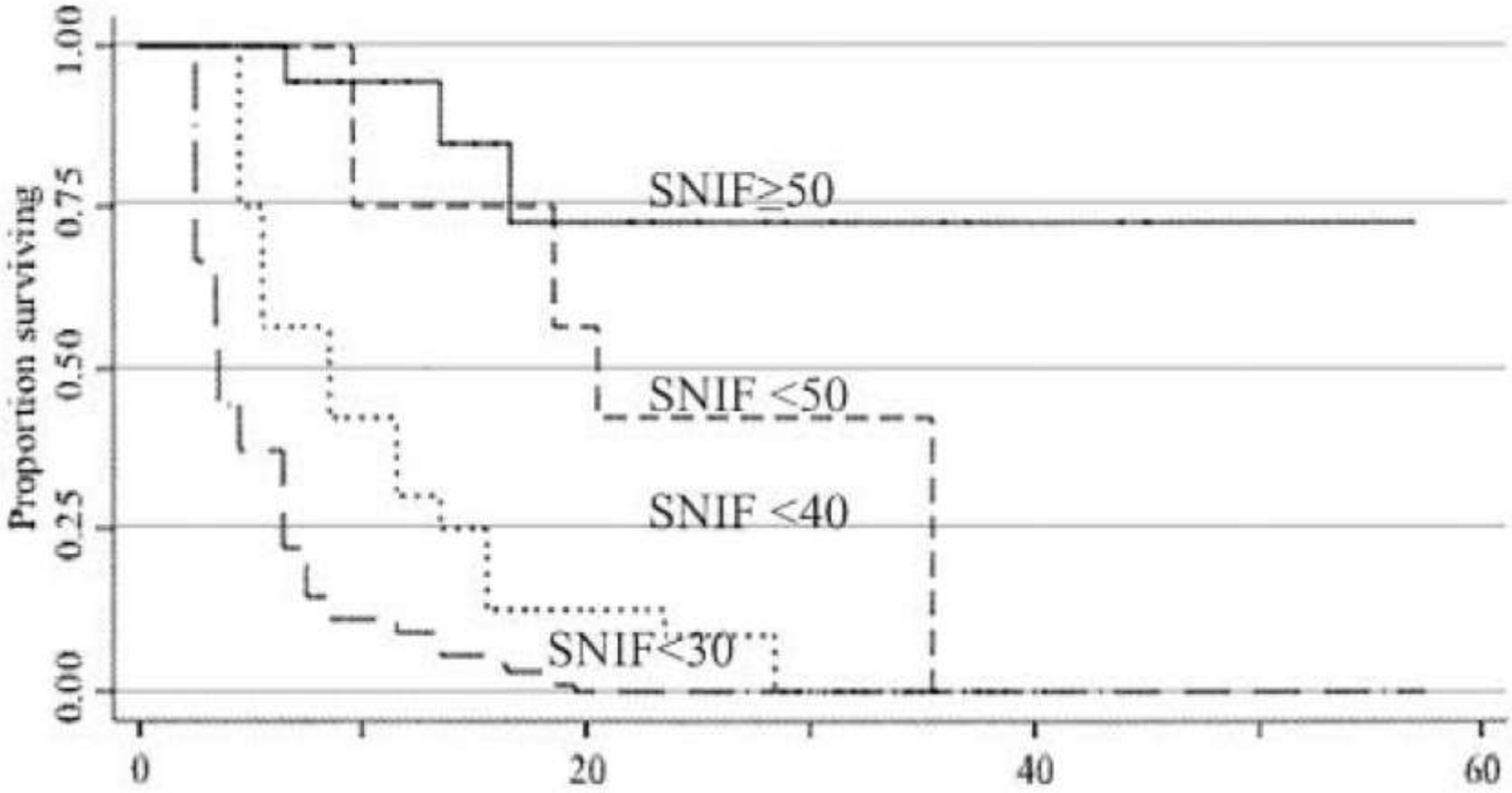
Sommeil +++

Polysomnographie



Arnulf et al., Am J Respir Crit Care Med, 2000

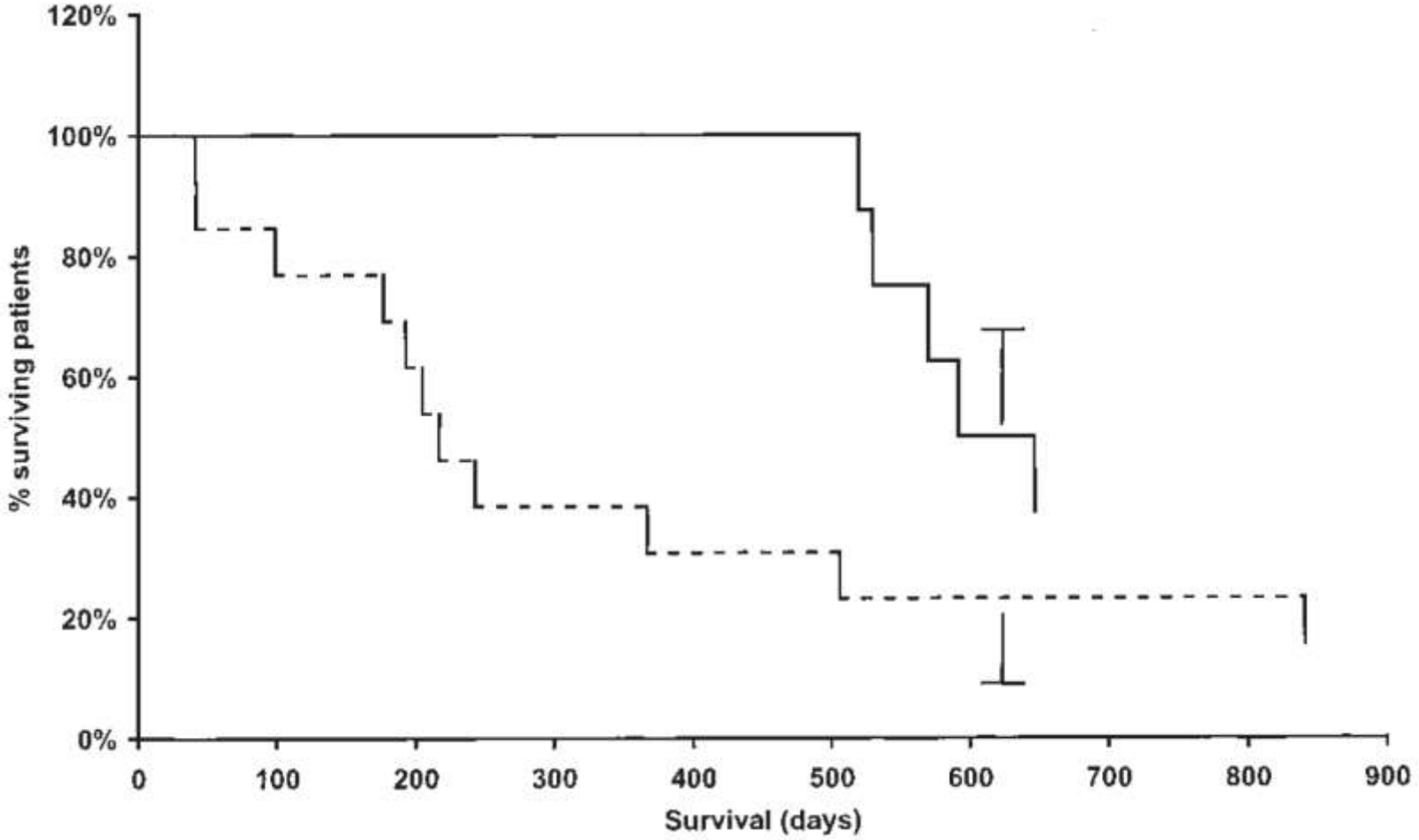
Sclérose Latérale Amyotrophique : survie



Morgan et coll. Am J Respir Crit Care Med 2005

Sclérose Latérale Amyotrophique : survie

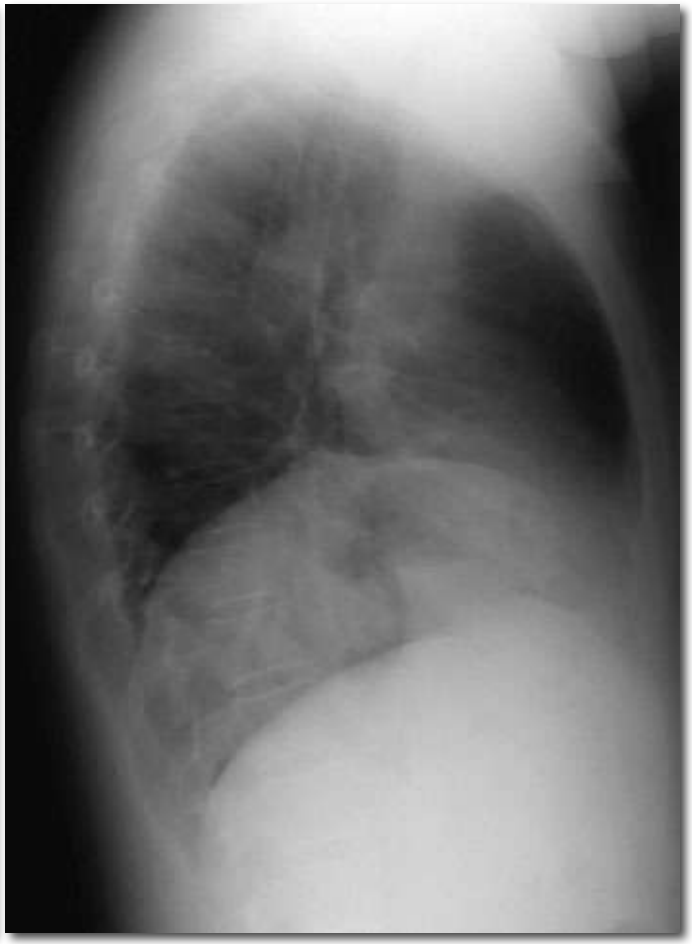
Impact d'une dysfonction diaphragmatique sur la survie



Arnulf et coll. Am J Respir Crit Care Med 2000

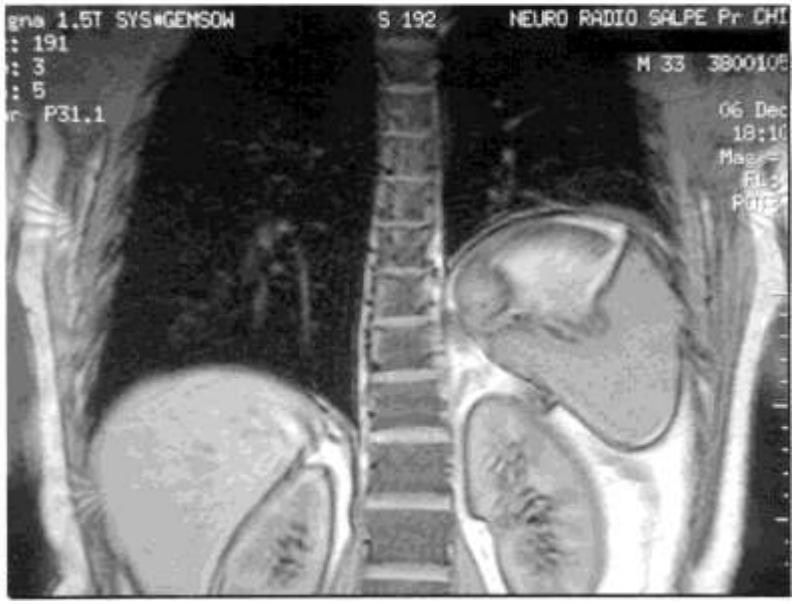
Diagnostic des atteintes diaphragmatiques :

Surélévation de coupole



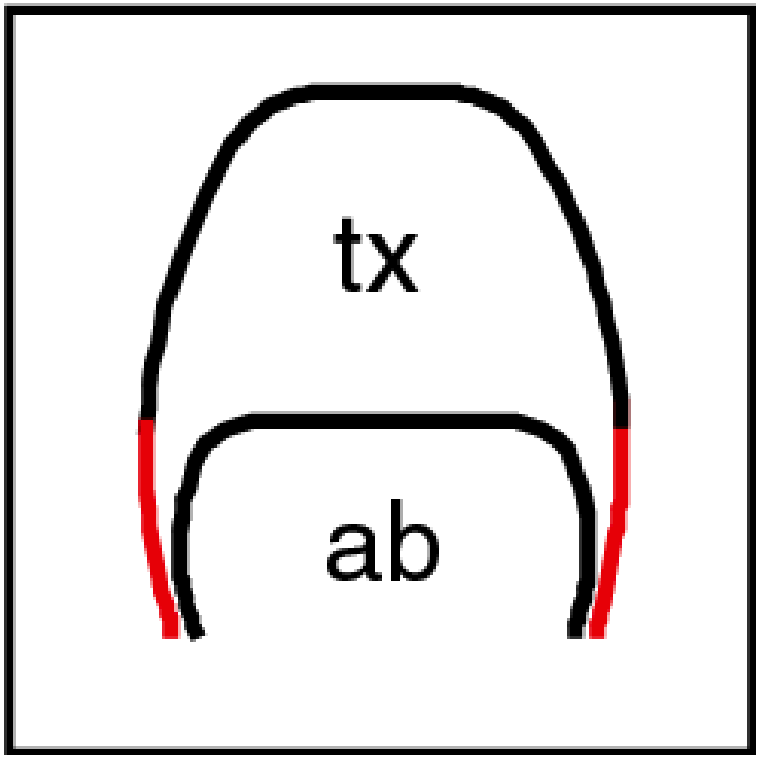
Diagnostic des atteintes diaphragmatiques :

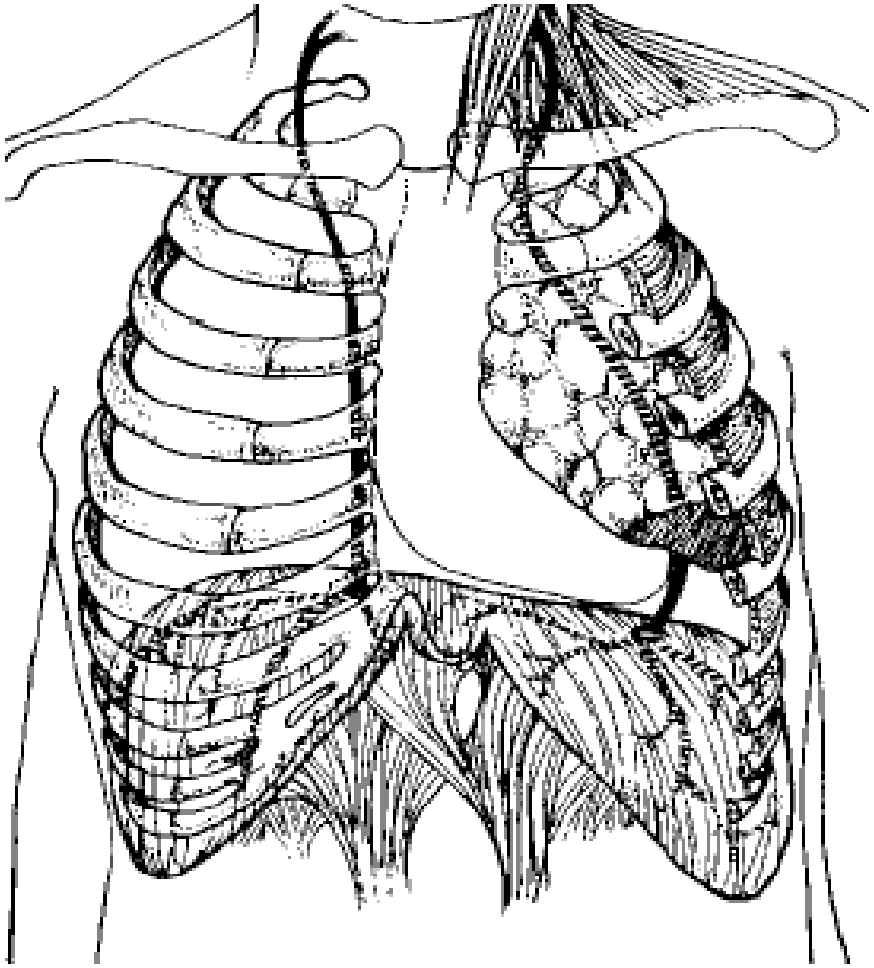
A



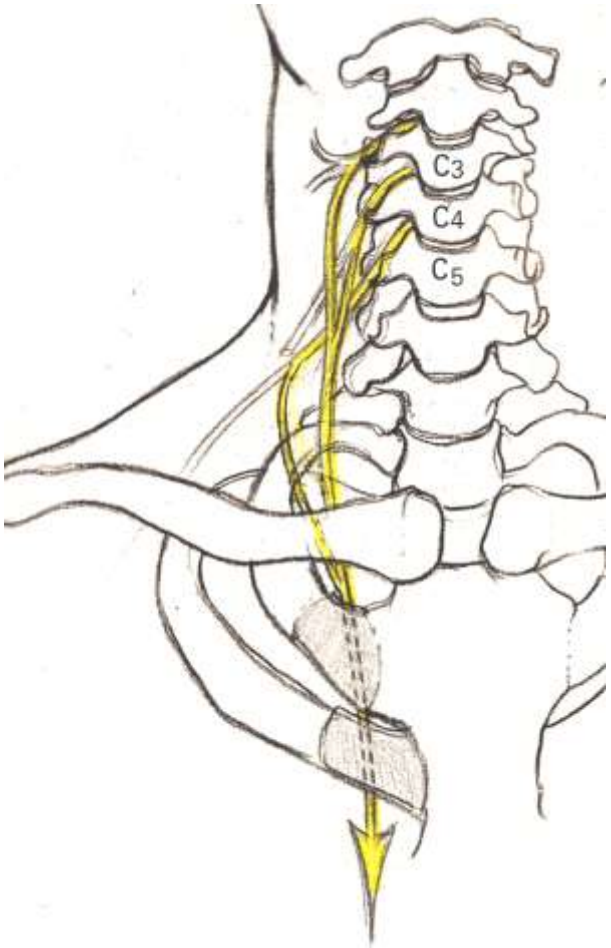
B

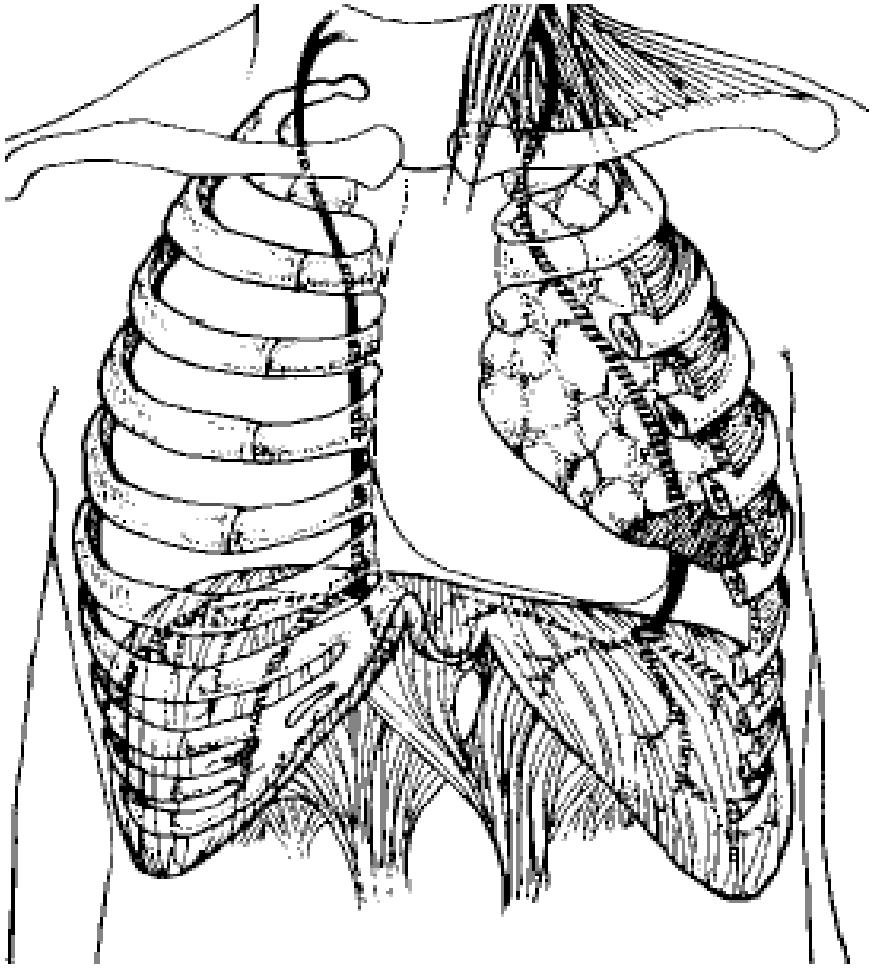
Le diaphragme, un septum



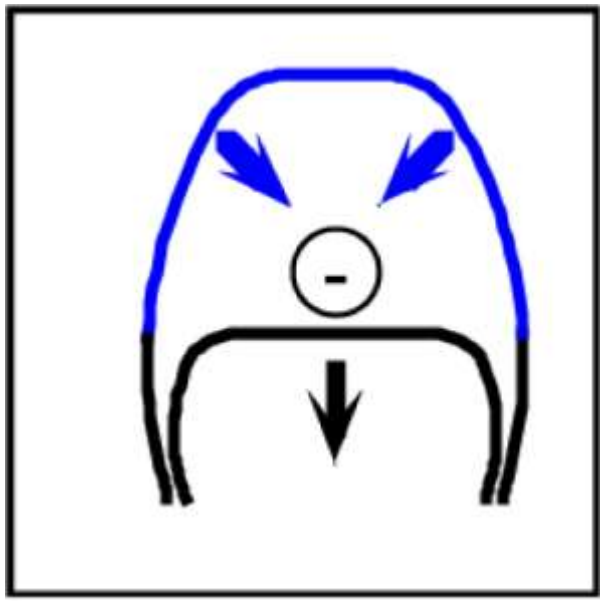


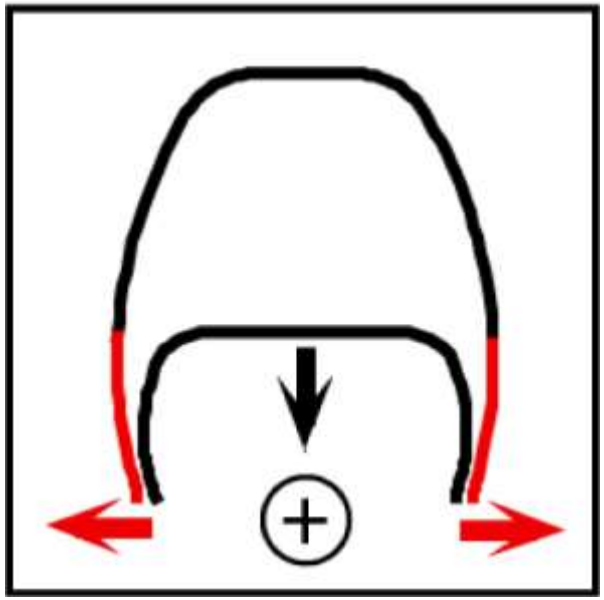
Le diaphragme, innervation

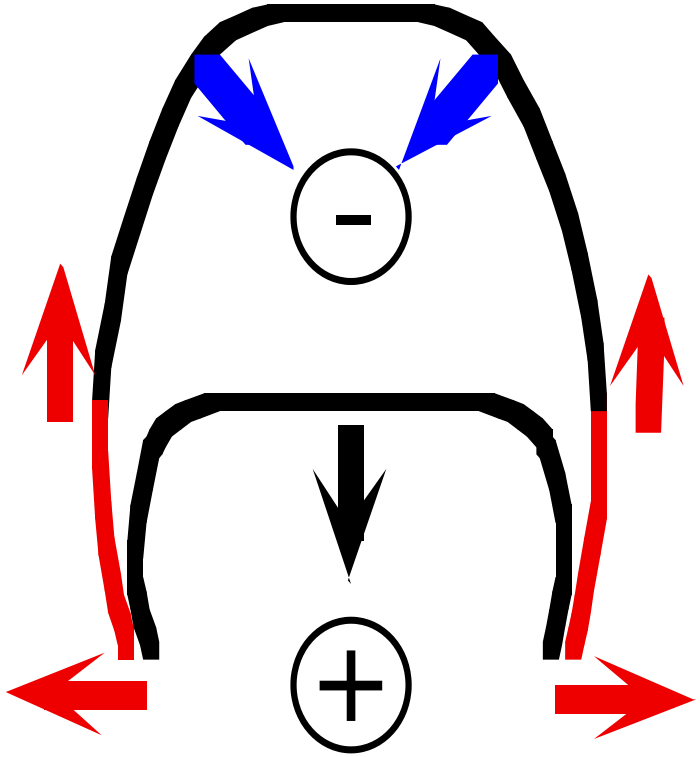




Action inspiratoire du diaphragme



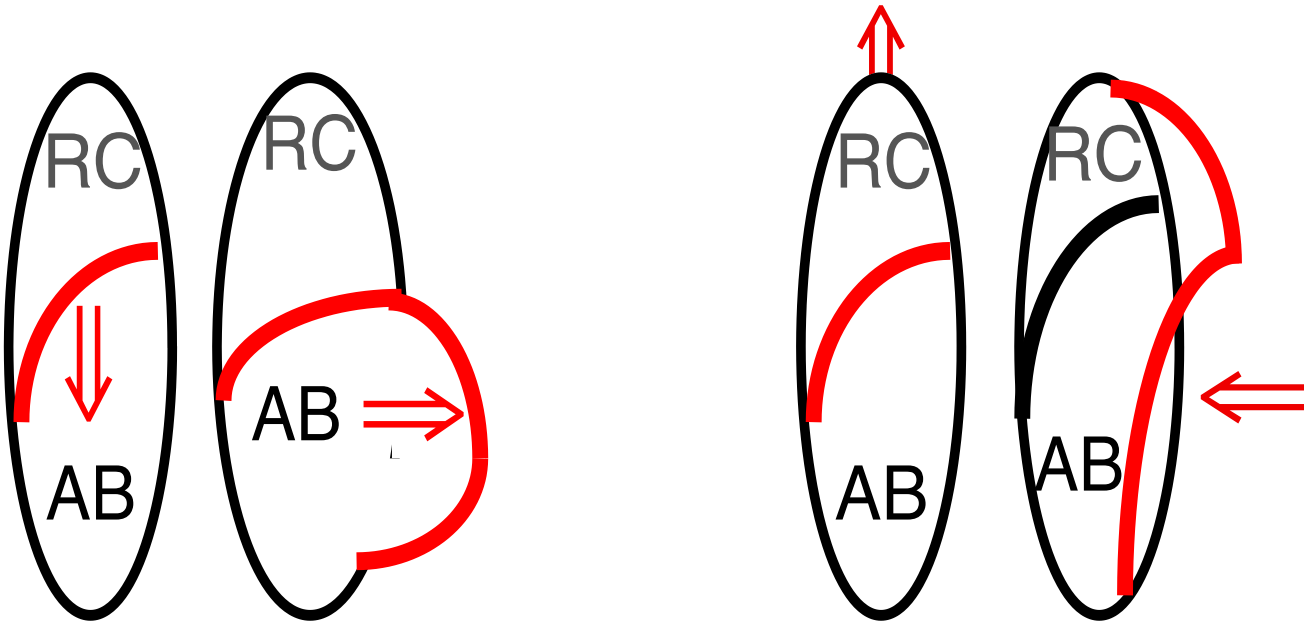




Diagnostic des atteintes diaphragmatiques : La clinique

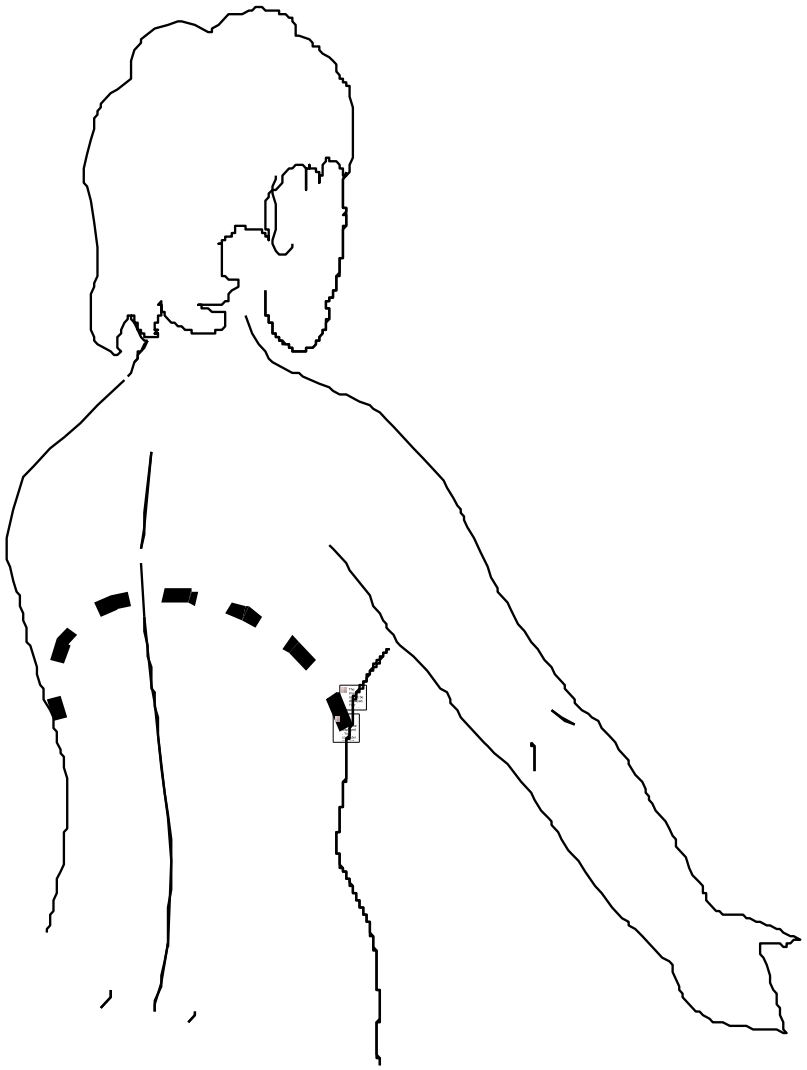
Examen

- Orthopnée
- Ventilation abdominale paradoxale dans le volume courant

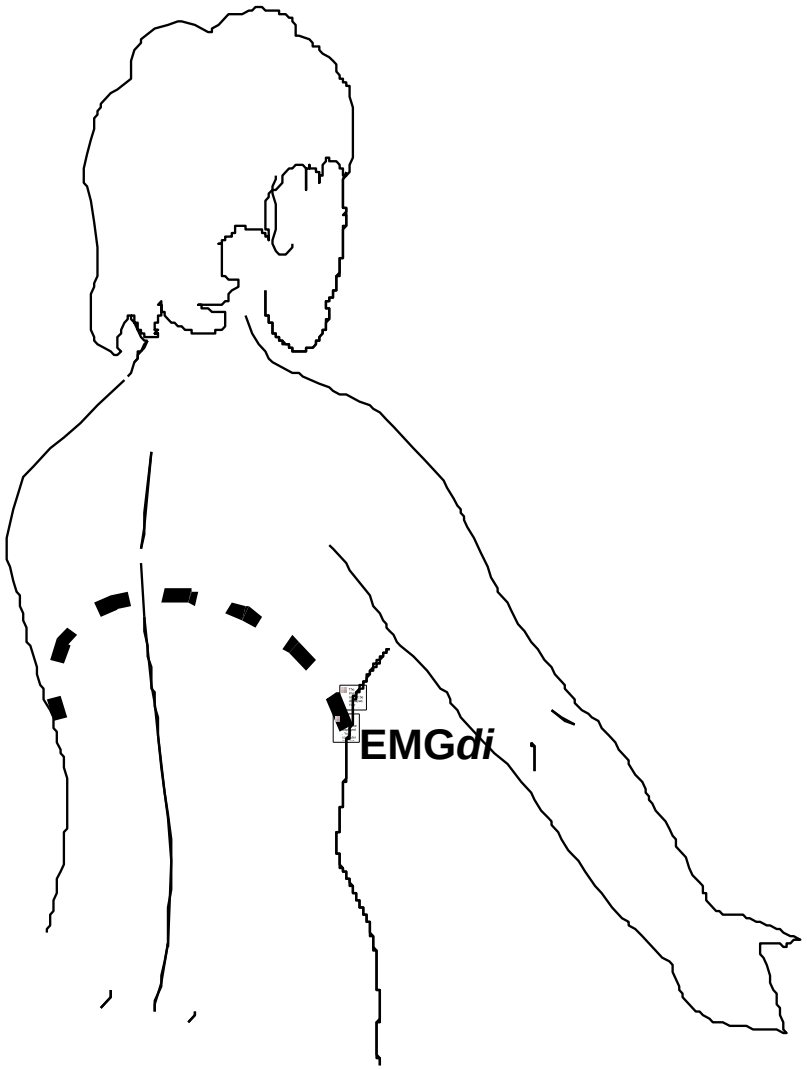


normal *dysfonction diaphragmatique*

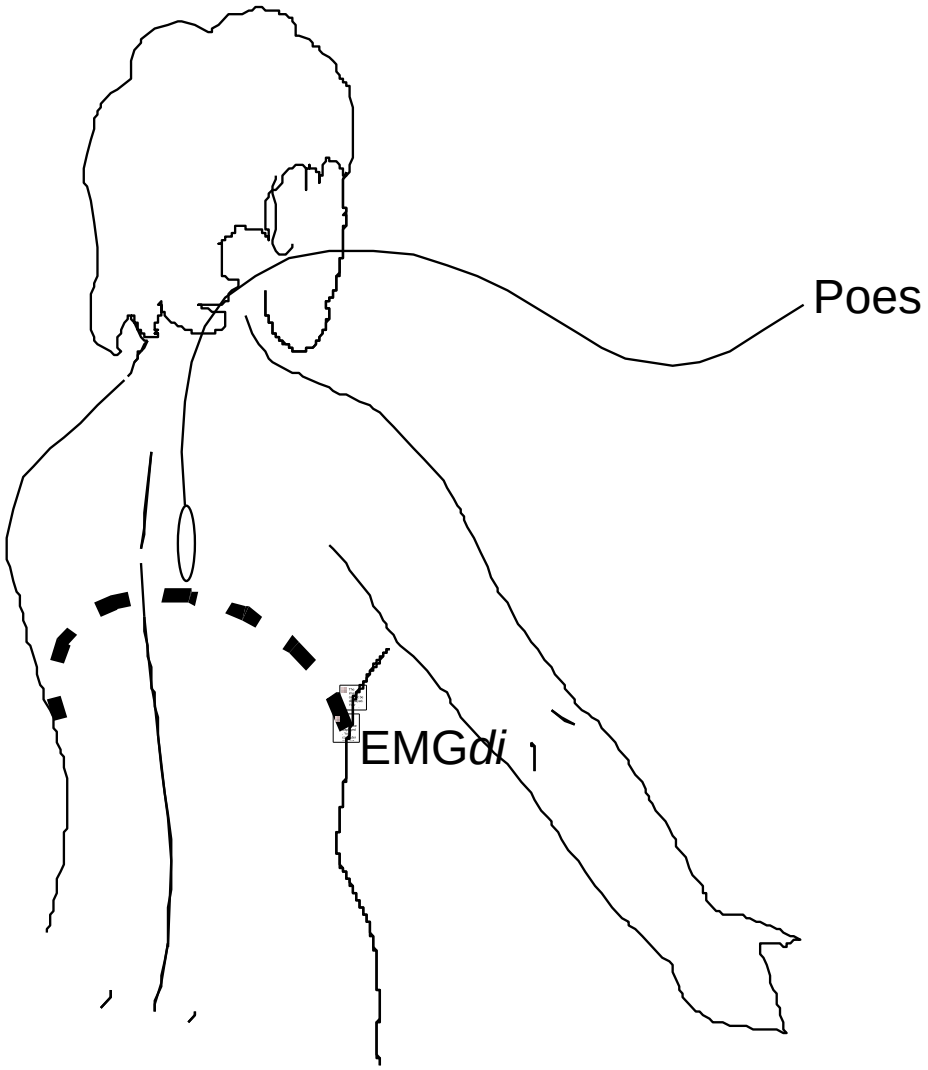
Exploration du diaphragme : recueils



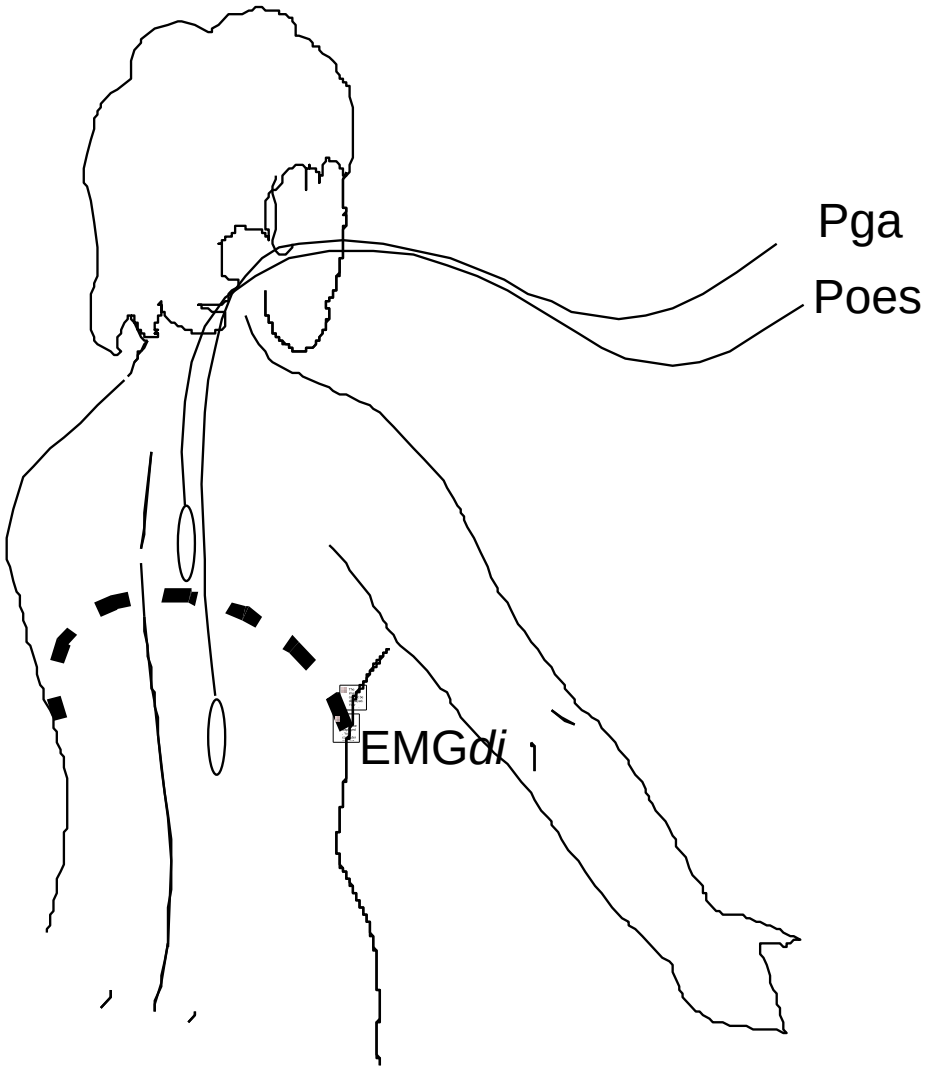
Exploration du diaphragme : recueils



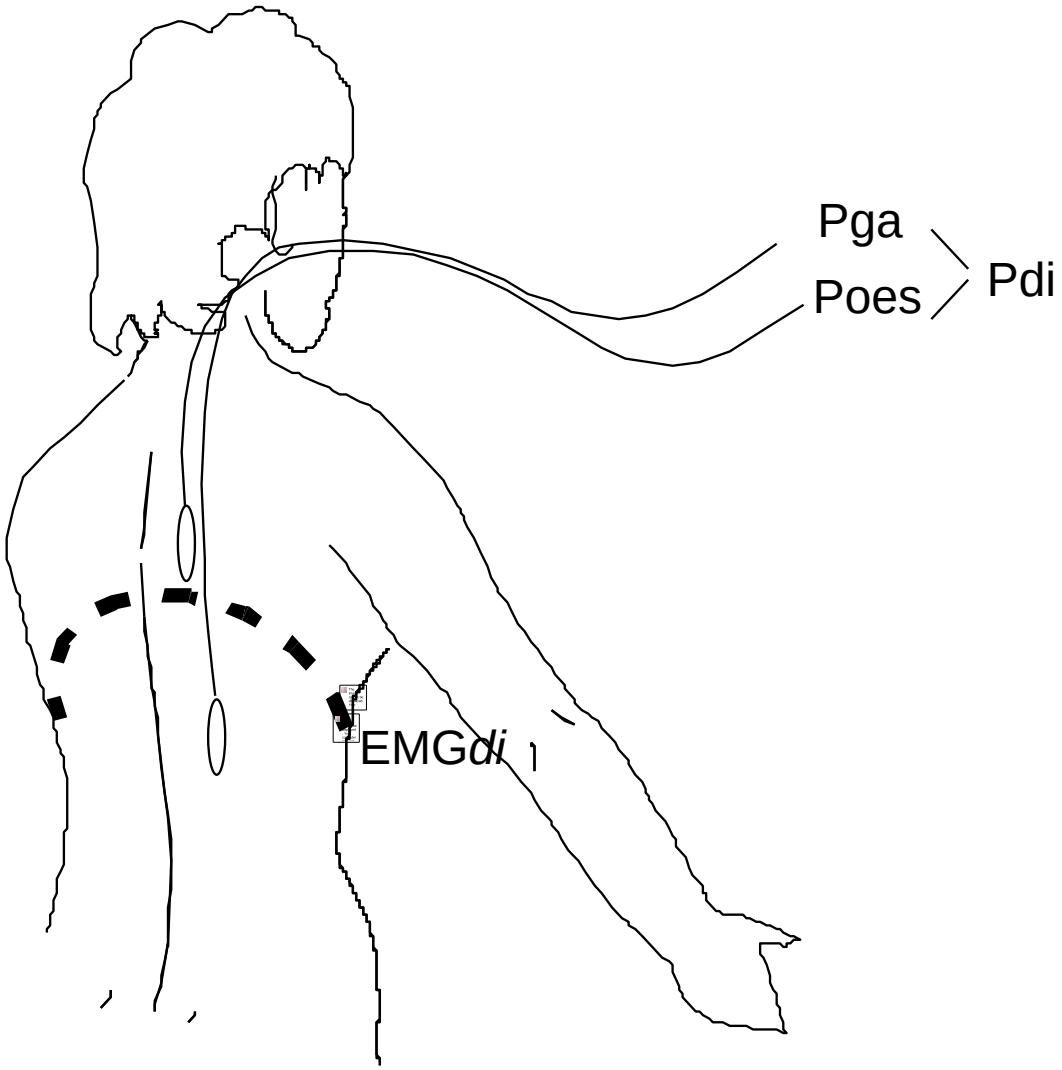
Exploration du diaphragme : recueils



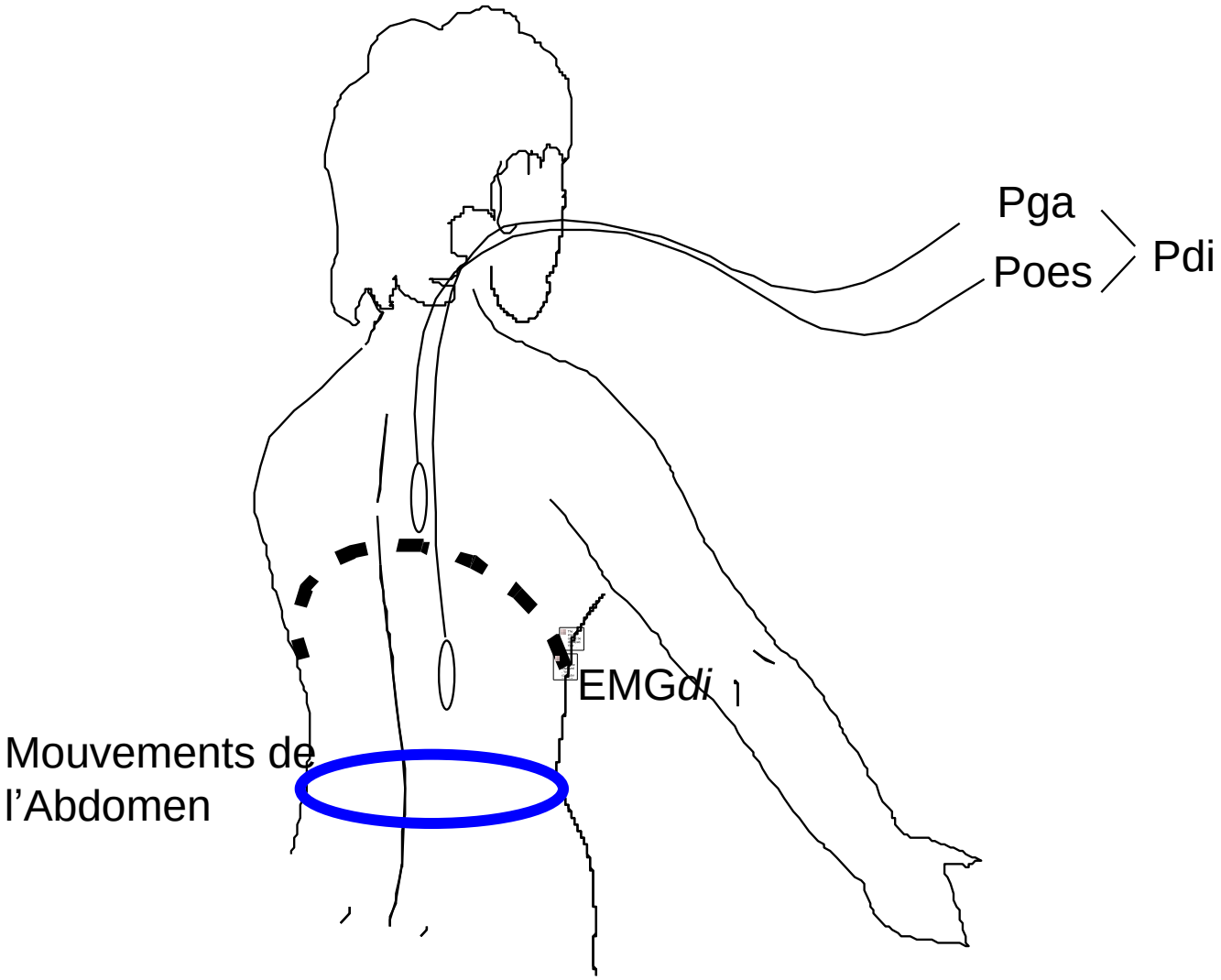
Exploration du diaphragme : recueils



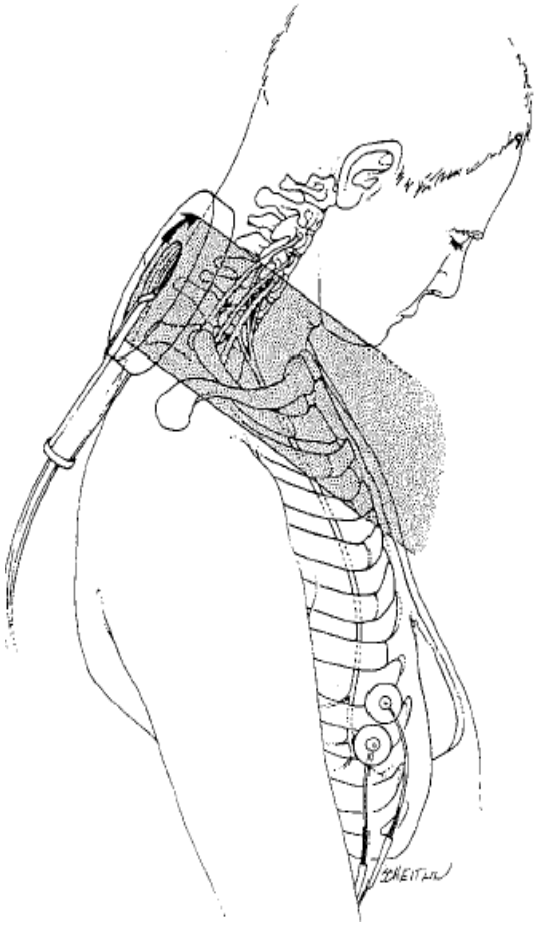
Exploration du diaphragme : recueils

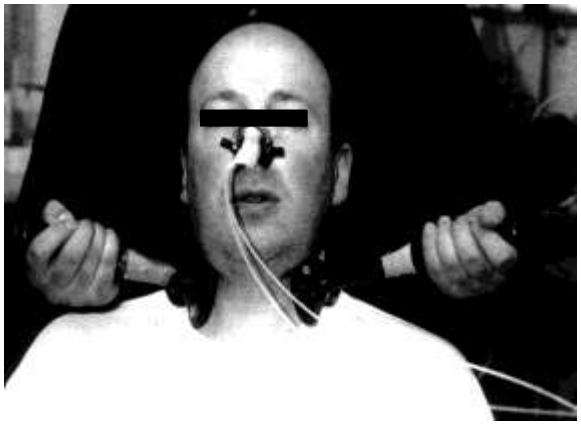


Exploration du diaphragme : recueils



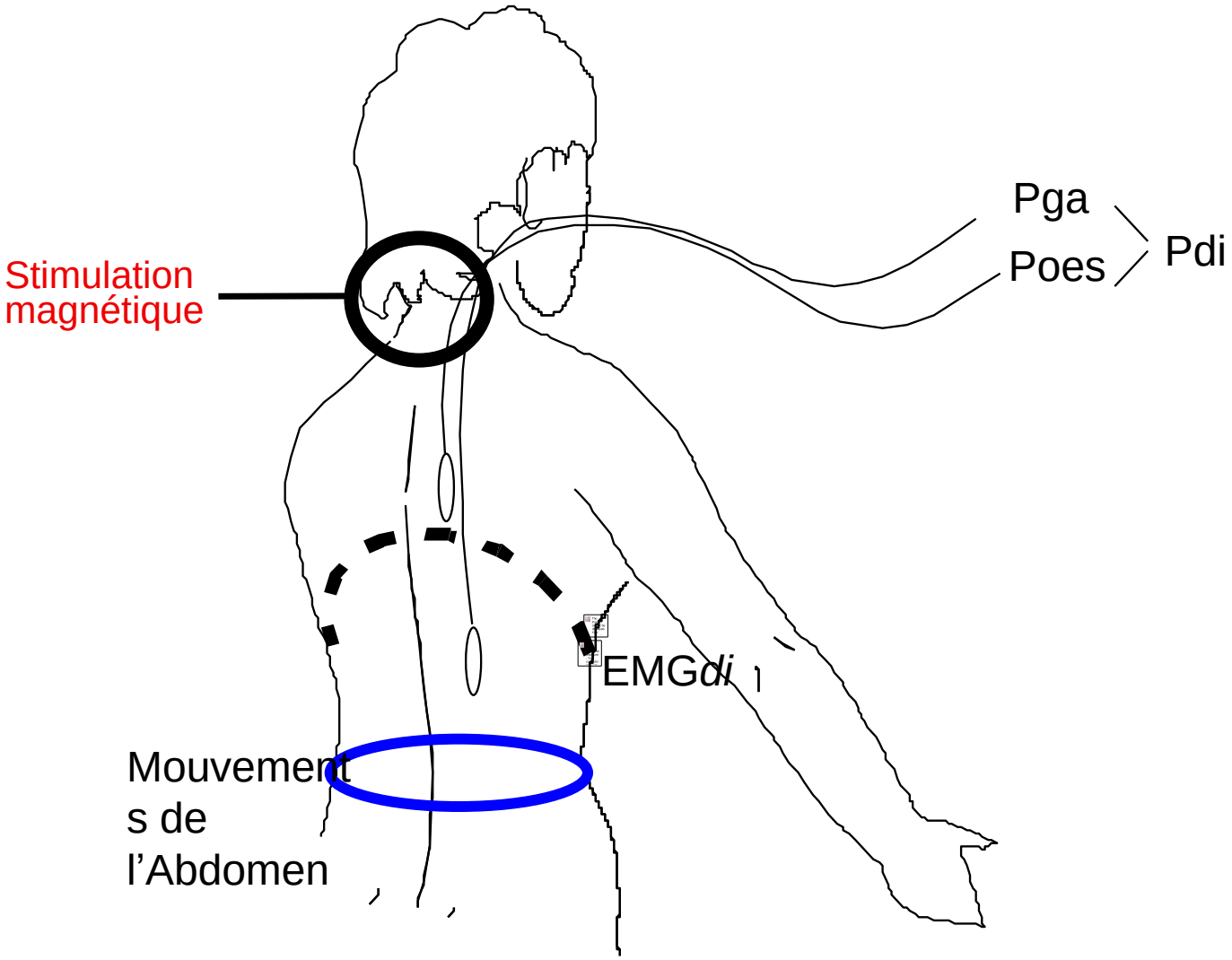
Exploration du diaphragme : stimulation du nerf phrénique



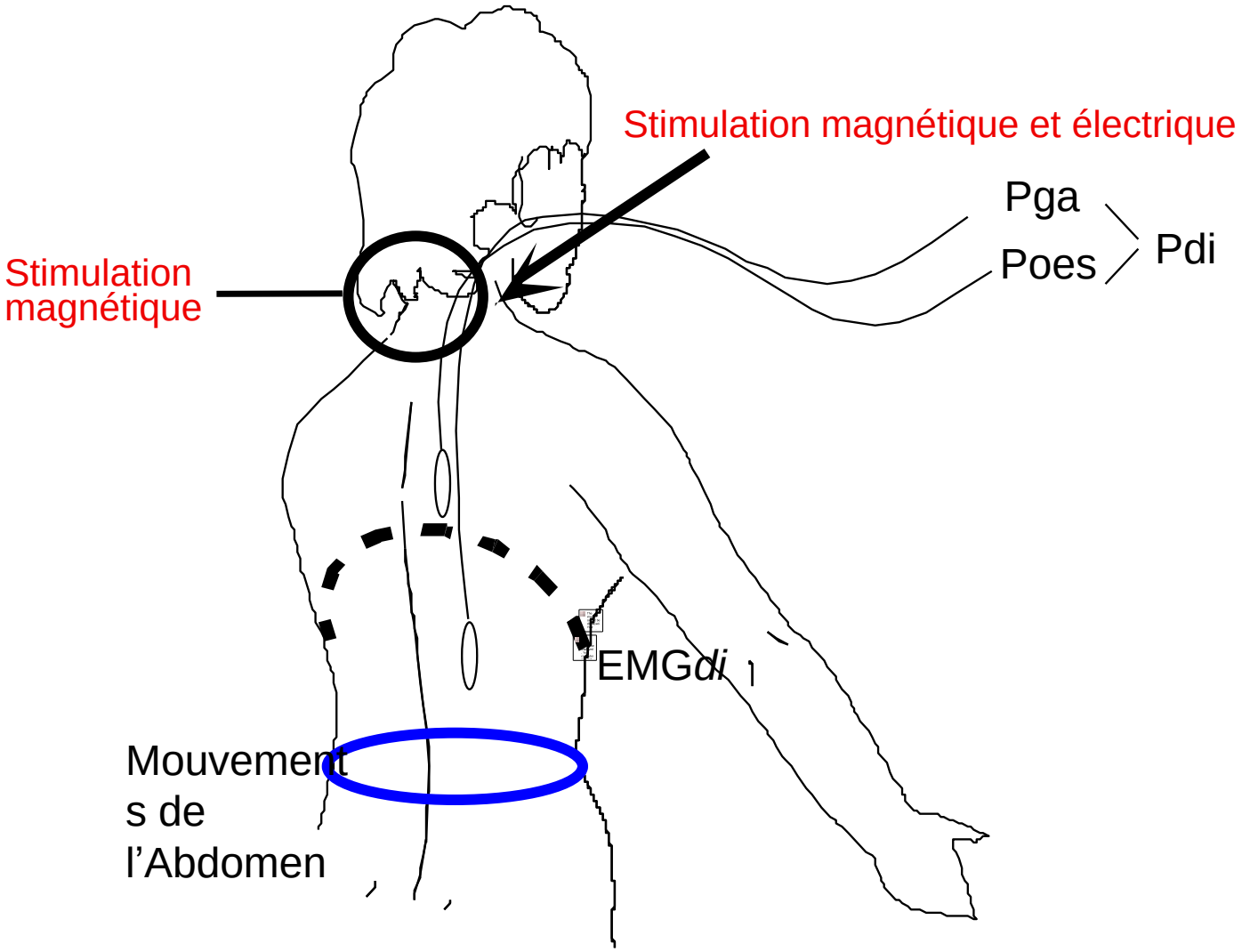


Mills G et al., Am J Respir Crit Care Med 1996 ; 154 : 1099-1105

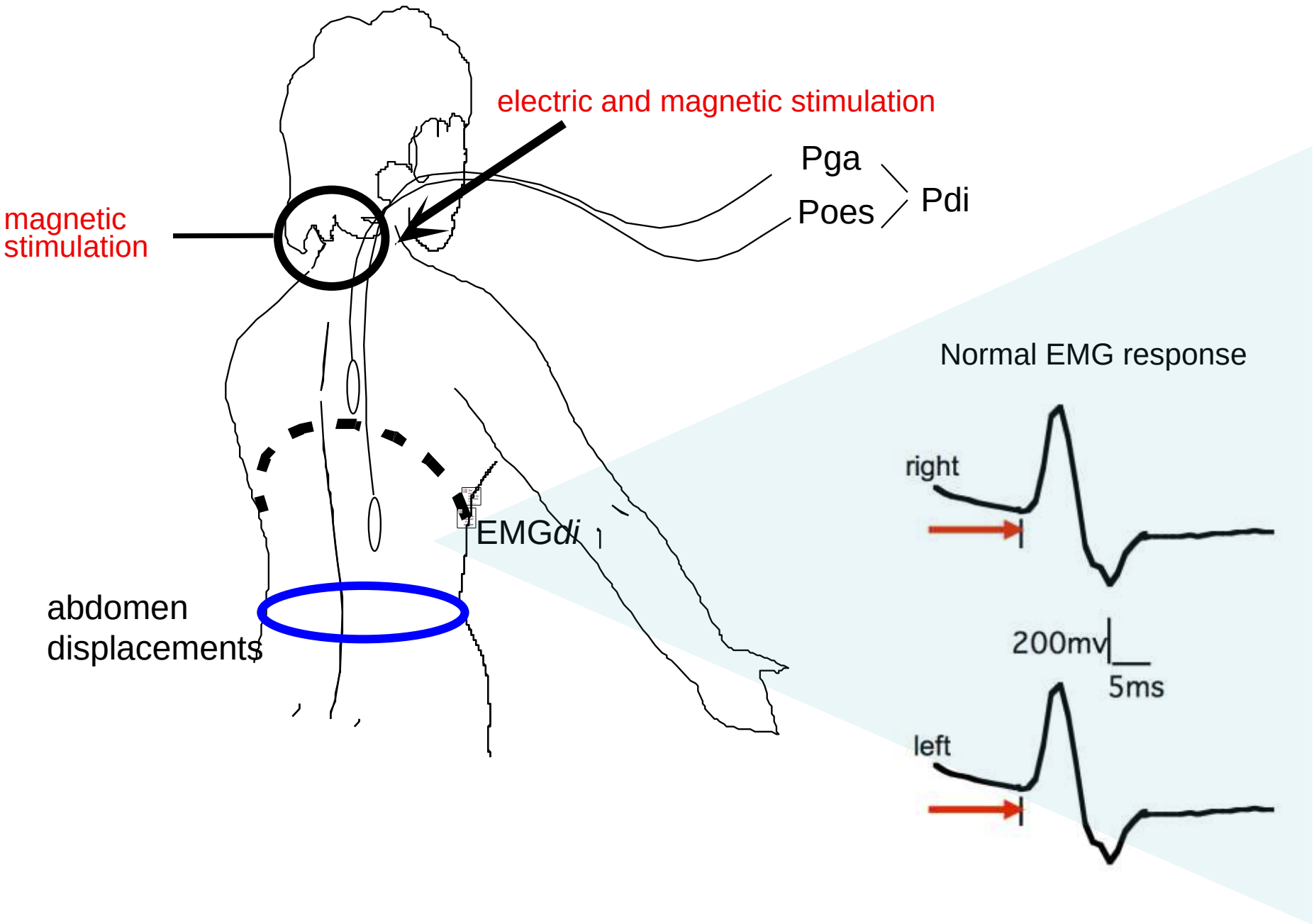
Exploration du diaphragme : recueils et stimulations



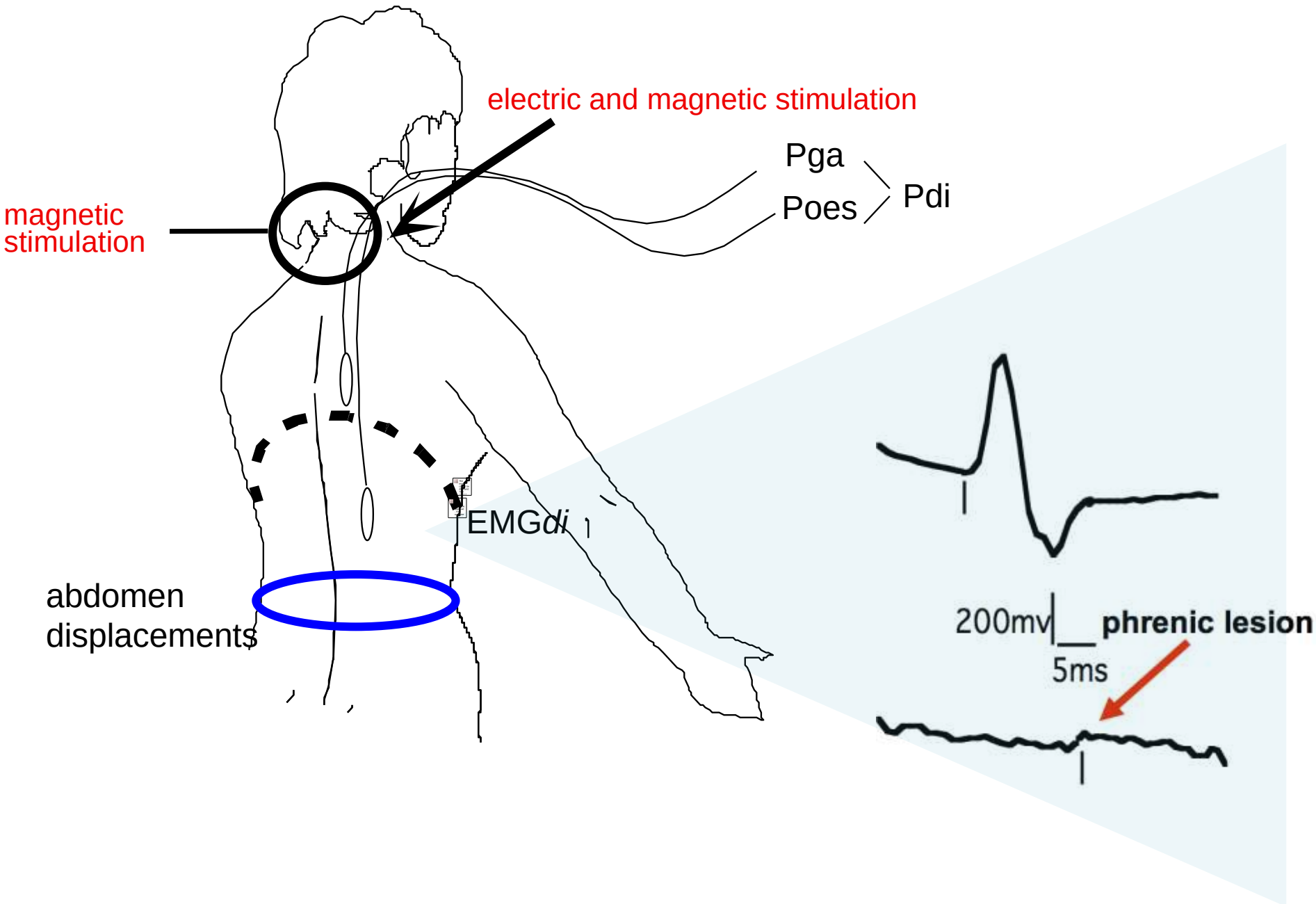
Recueils et stimulations



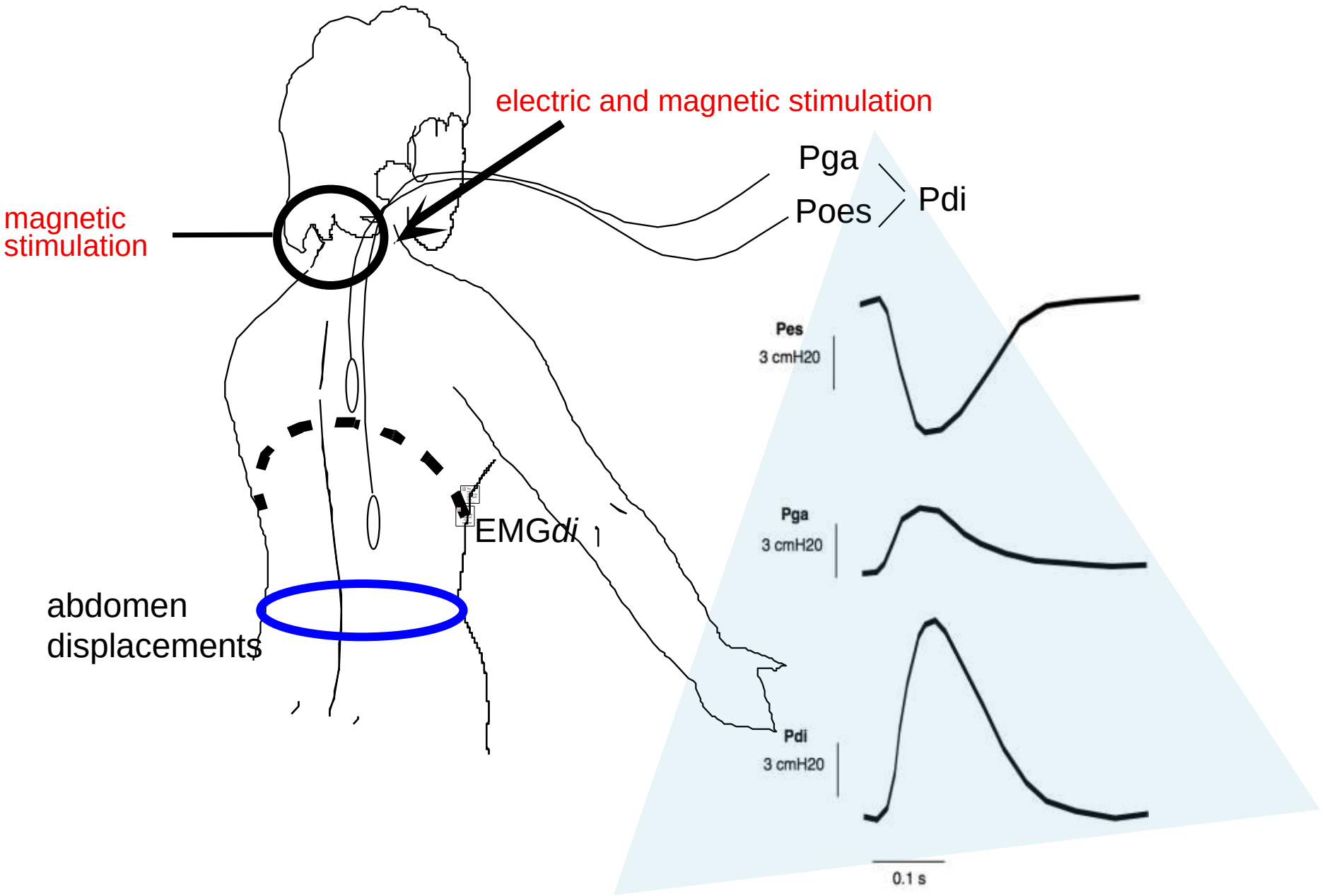
Exploration du nerf phrénique



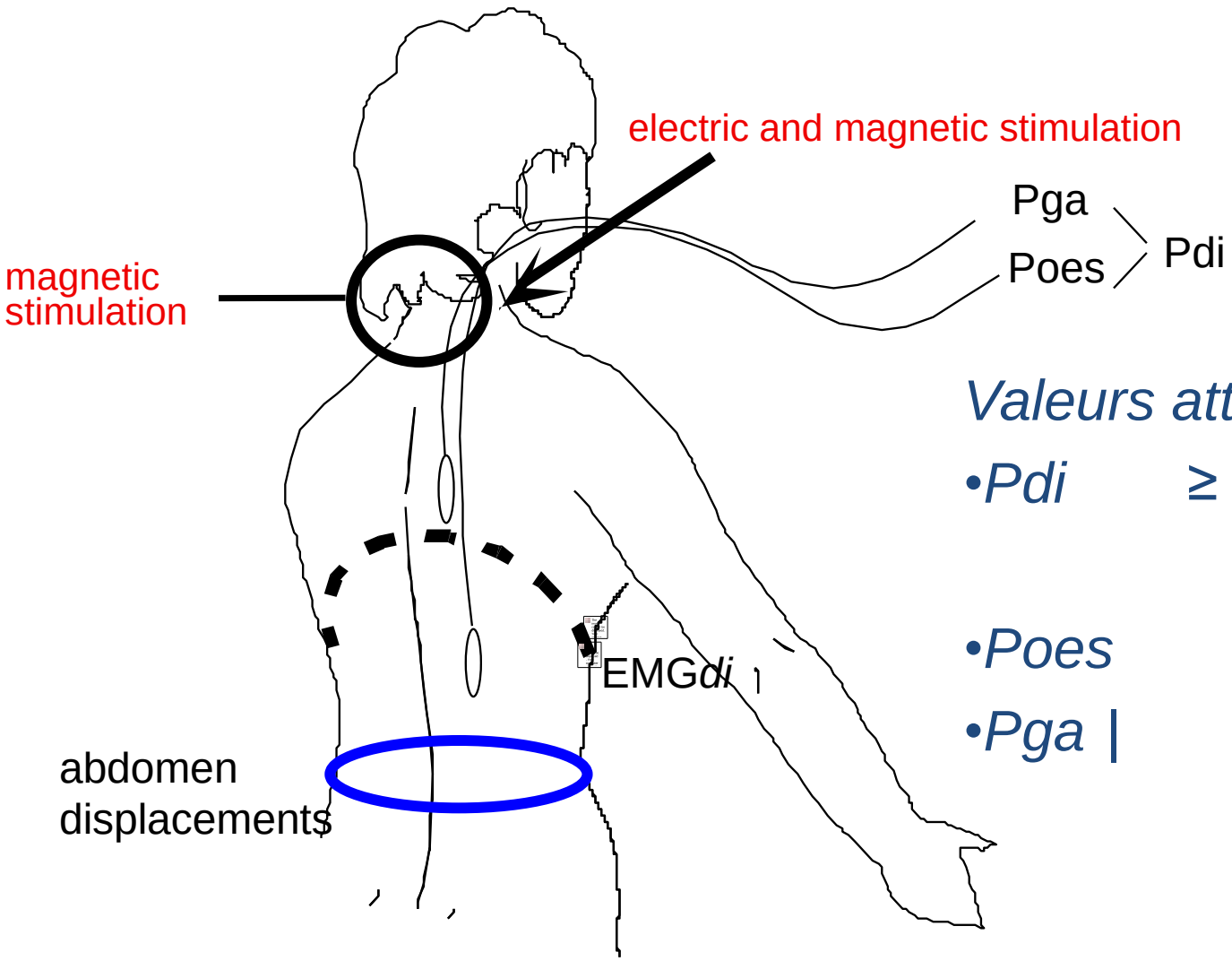
Exploration du nerf phrénique



Exploration de la force du diaphragme



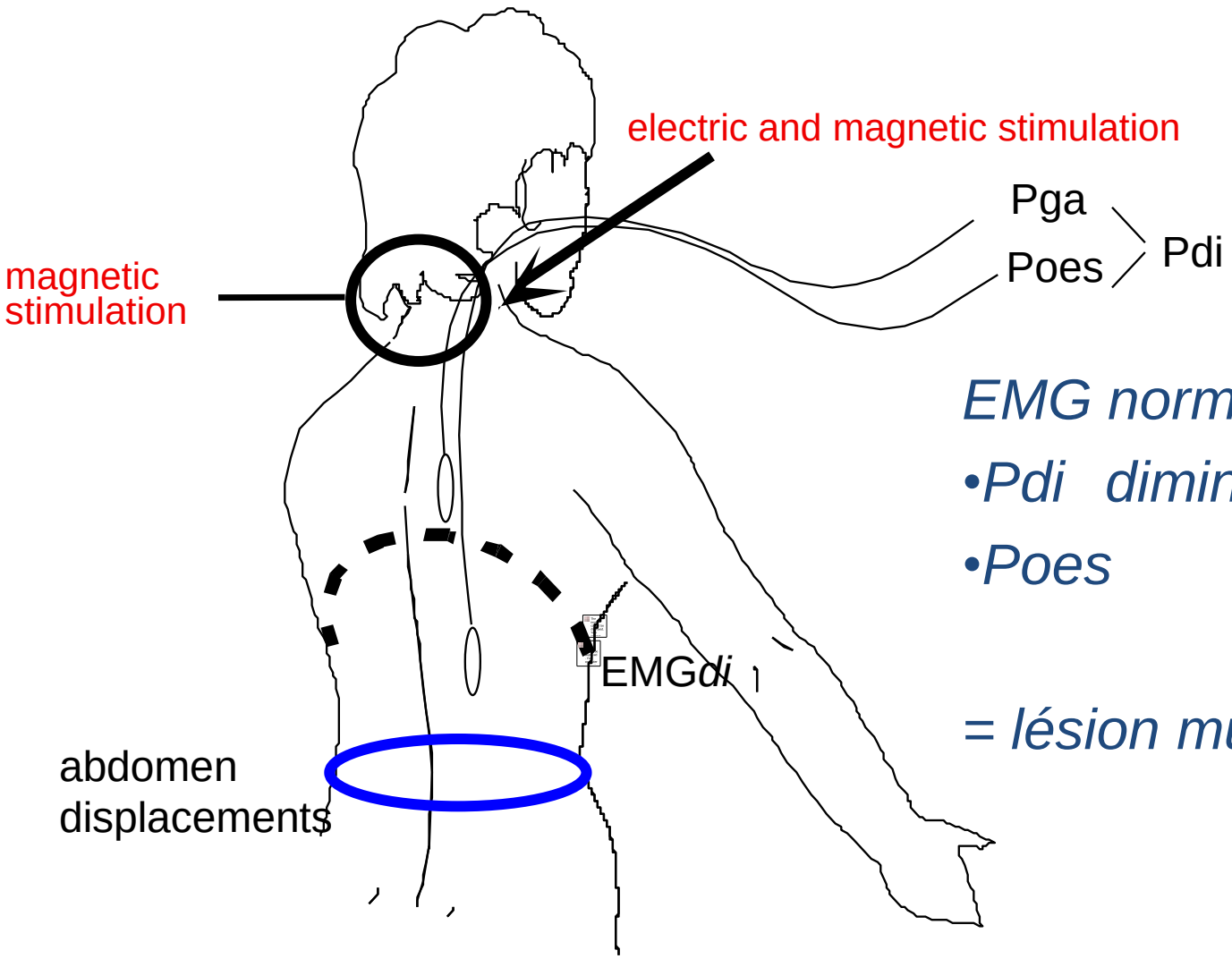
Exploration de la force du diaphragme



Valeurs attendues

- *Pdi* ≥ *20 cm H20*
- *Poes* | ≥ *10 cm H2O*
- *Pga* |

Exploration de la force du diaphragme



EMG normal et

- *Pdi diminuée*
- *Poes* diminuée

= lésion musculaire

Sommeil +++

Polysomnographie

VNI

Merci