12èmes Journées Scientifiques de la Société Française de la Mucoviscidose

Filière MUCO | CFTR

Exploration Fonctionnelle Respiratoire

Explorer les muscles respiratoires

Pr Brigitte Fauroux

Unité de ventilation nonivasive et du sommeil

Inserm U 955

Hôpital Necker, Paris

UNIVERSITÉ PARIS DESCARTES

Necker ENFANTS MALADES HÔPITAL UNIVERSITAIRE

Inserm

Institut national de la santé et de la recherche médicale

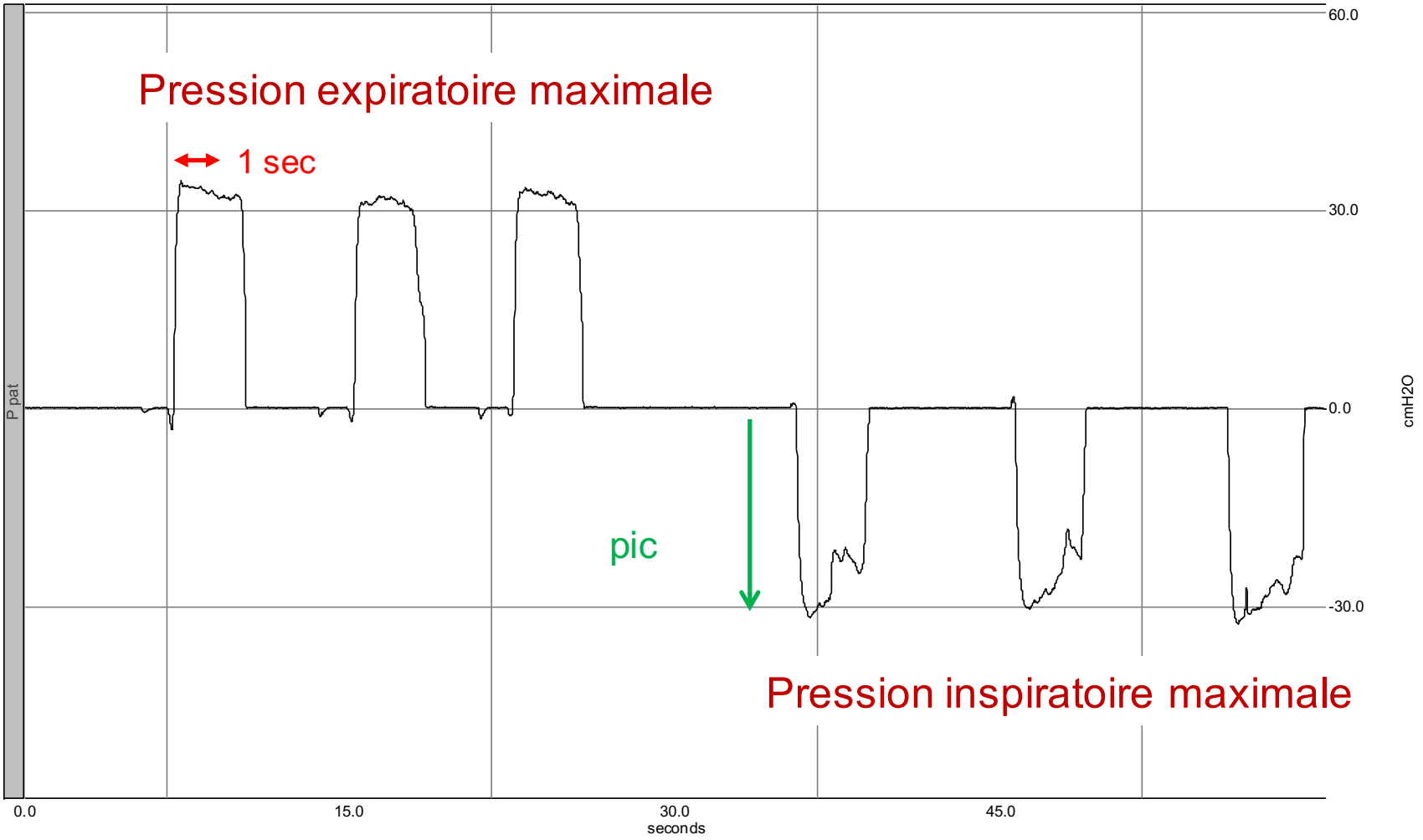
Explorer les muscles respiratoires

- Quelles explorations ?
- Quelle est la performance des muscle respiratoires dans la CF ?
- Quels sont les déterminants de la performance des muscles respiratoires ?
- En pratique…

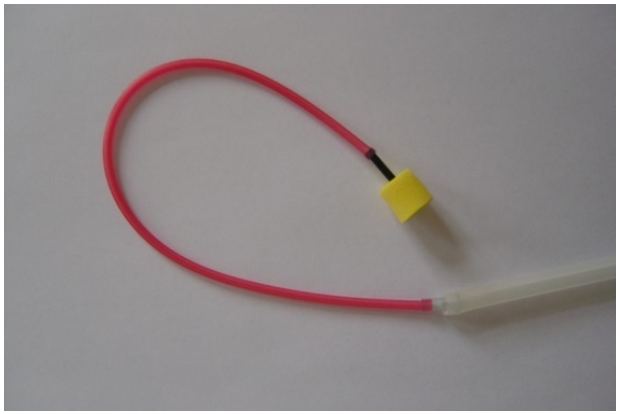
Explorations des muscles respiratoires

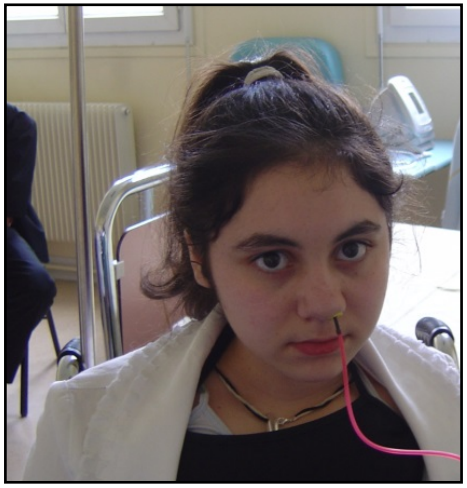
- Tests noninvasifs
  - volitionnels
    - pressions statiques maximales
    - SNIP
  - *non volitionnels: écho, IRM, OEP*
- Tests invasifs
  - volitionnels
    - Sniff Poeso et Pgas Toux
  - non volitionnels
    - stimulation magnétique des nerfs phréniques

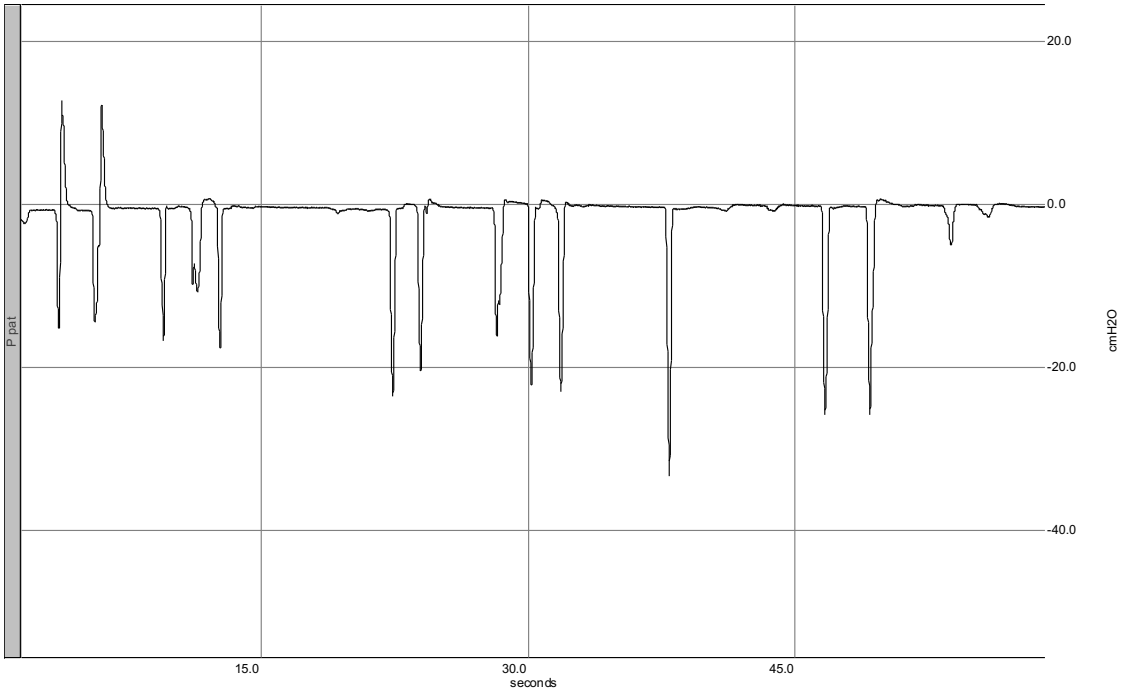
Pressions statiques maximales



Sniff Nasal Inspiratory Pressure



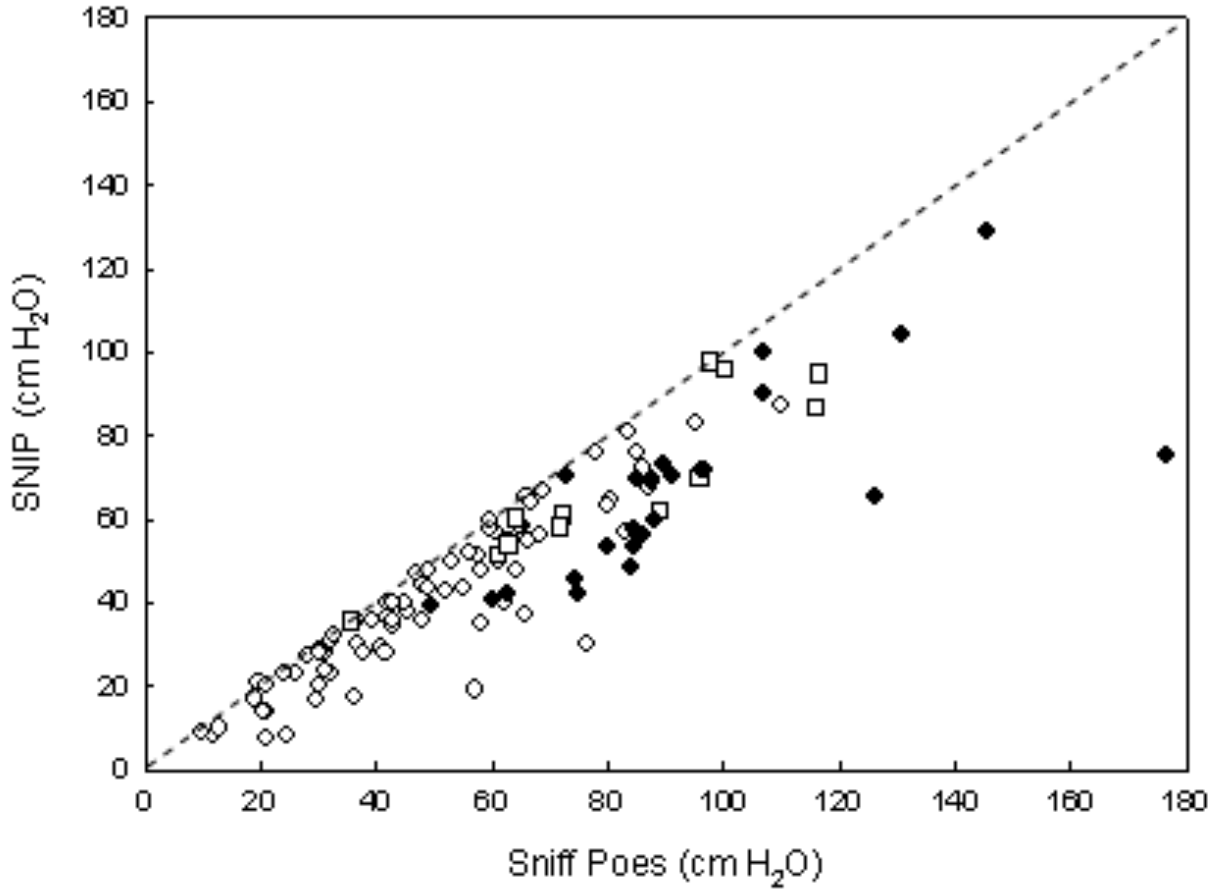




Sniff nasal inspiratory pressure in children with muscular, chest wall or lung disease

B. Fauroux*,#, G. Aubertin*, E. Cohen*,#, A. Clément*,# and F. Lofaso¶,+

Eur Respir J 2009; 33: 113–117



◆ CF

The influence of body composition on respiratory muscle, lung function and diaphragm thickness in adults with cystic fibrosis ☆

Stephanie Enright [a,*], Ken Chatham [b,1], Alina A. Ionescu [d,2], Viswanath B. Unnithan [c,3], Dennis J. Shale [d,2]

Journal of Cystic Fibrosis

Variables	CF patients (n=40)	Healthy subjects (n=30)
Age (years)	22.4 (21.2–23.6)	21.7 (20.3–23.1)
Height (cm)	166 (160.1–171.9)	168 (164.4–171.6)
Weight (kg)	60.2 (58.5–62.0)	64.4 (60.9–67.9)
BMI (kg/m^2)	22.0 (21.0–23.0)	23.0 (22.2–23.8)
FFM (kg)	44.6 (38.6–50.6)	56.8 (49.8–63.8) [a]
PAS (MET's)	37.0 (35.0–39.0)	41.5 (40.0–43.0)
FEV_1 (% pred)	46.5 (46–47)	96.2 (95.2–97.2) [a]
VC (litres)	3.6 (3.3–3.9)	4.1 (3.7–4.5) [a]
RV (litres)	1.7 (1.6–1.8)	1.5 (1.4–1.6)
FRC (litres)	2.6 (2.2–3.0)	2.9 (2.6–3.1)
TLC (litres)	4.6 (4.2–5.0)	5.8 (5.6–6.0) [a]
Diaph (TDIrel) (mm)	2.8 (2.6–3.1)	3.4 (3.0–3.8)
Diaph (TDIcont) (mm)	3.9 (3.6–4.2)	4.4 (4.2–4.6)
TR (%)	2.7 (2.4–3.0)	2.9 (2.7–3.2)
PImax (cm H_2O)	116 (109–123)	124 (119–129)
SPImax (PTU)	609 (543–675)	788 (733–843) [a]

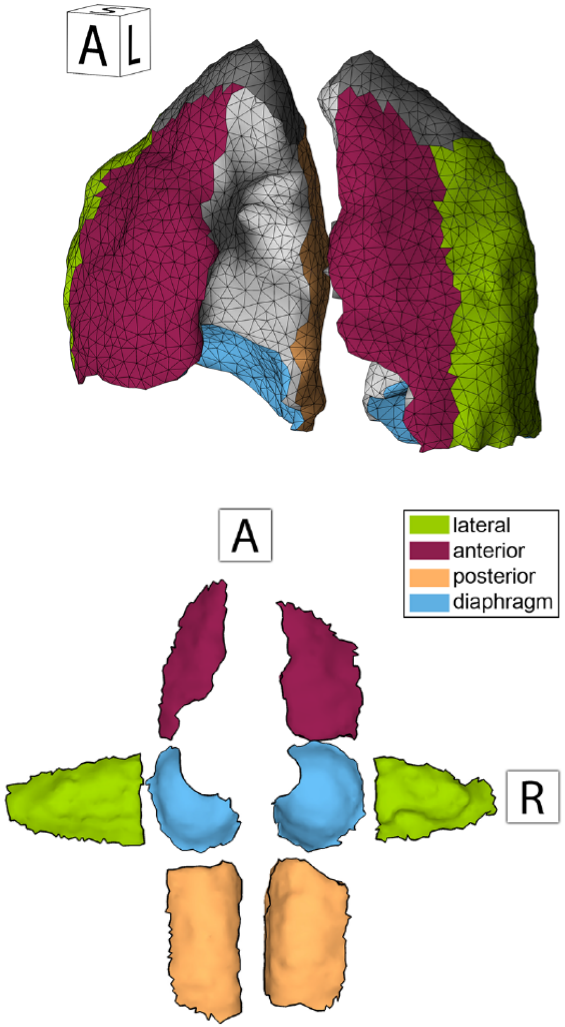
épaisseur du diaphragme
à la CRF
à la CPT
thickening ratio (TR)
Sustained PImax

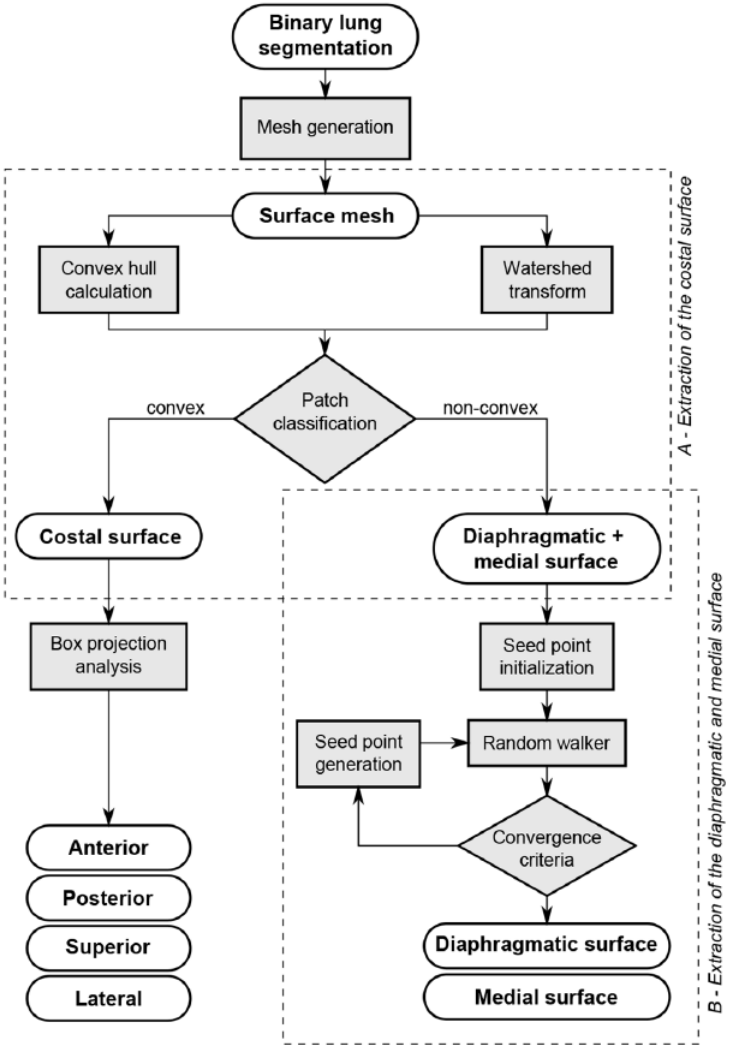
Journal of Cystic Fibrosis 6 (2007) 384 – 390

Quantification of Diaphragm Mechanics in Pompe Disease Using Dynamic 3D MRI

Katja Mogalle[1]*, Adria Perez-Rovira[1,2], Pierluigi Ciet[3,4], Stephan C. A. Wens[5,6], Pieter A. van Doorn[5,6], Harm A. W. M. Tiddens[3,4], Ans T. van der Ploeg[6,7], Marleen de Bruijne[1,8]*

July 8, 2016





Quantification of Diaphragm Mechanics in Pompe Disease Using Dynamic 3D MRI

Katja Mogalle[1]*, Adria Perez-Rovira[1,2], Pierluigi Ciet[3,4], Stephan C. A. Wens[5,6], Pieter A. van Doorn[5,6], Harm A. W. M. Tiddens[3,4], Ans T. van der Ploeg[6,7], Marleen de Bruijne[1,8]*

July 8, 2016

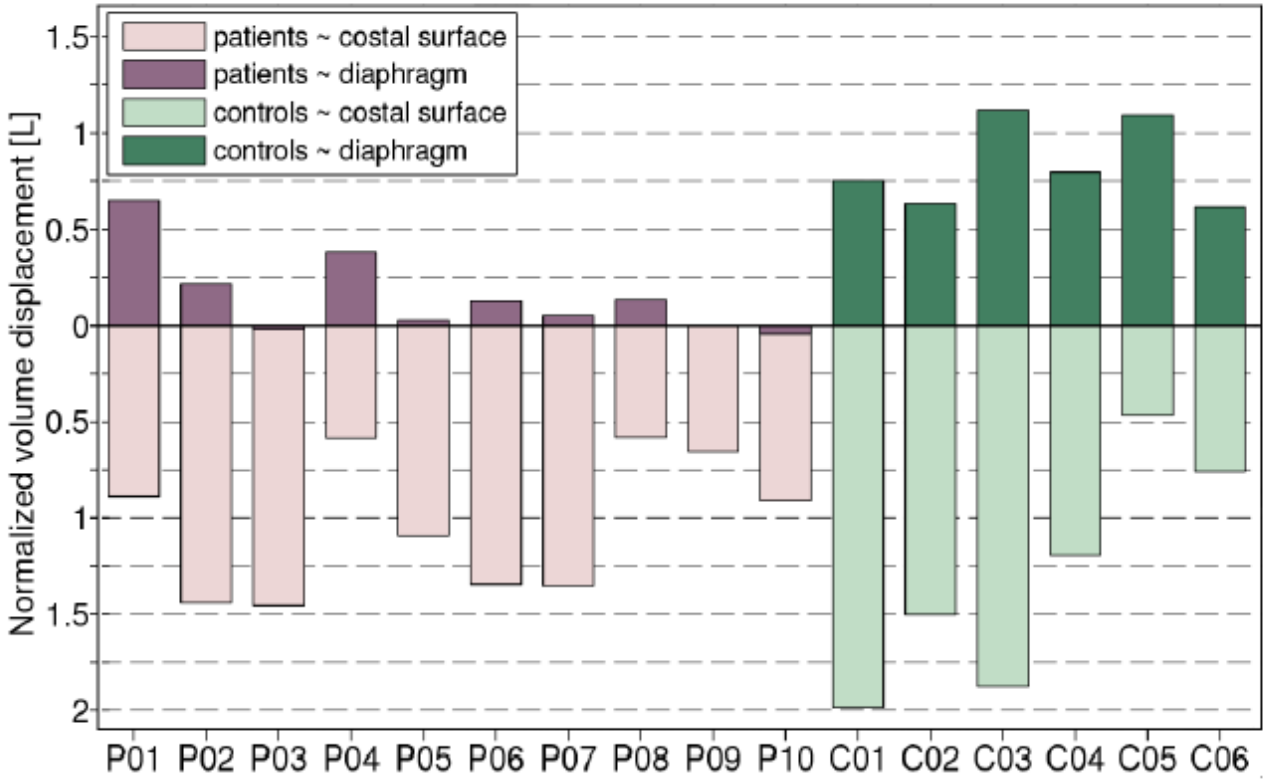
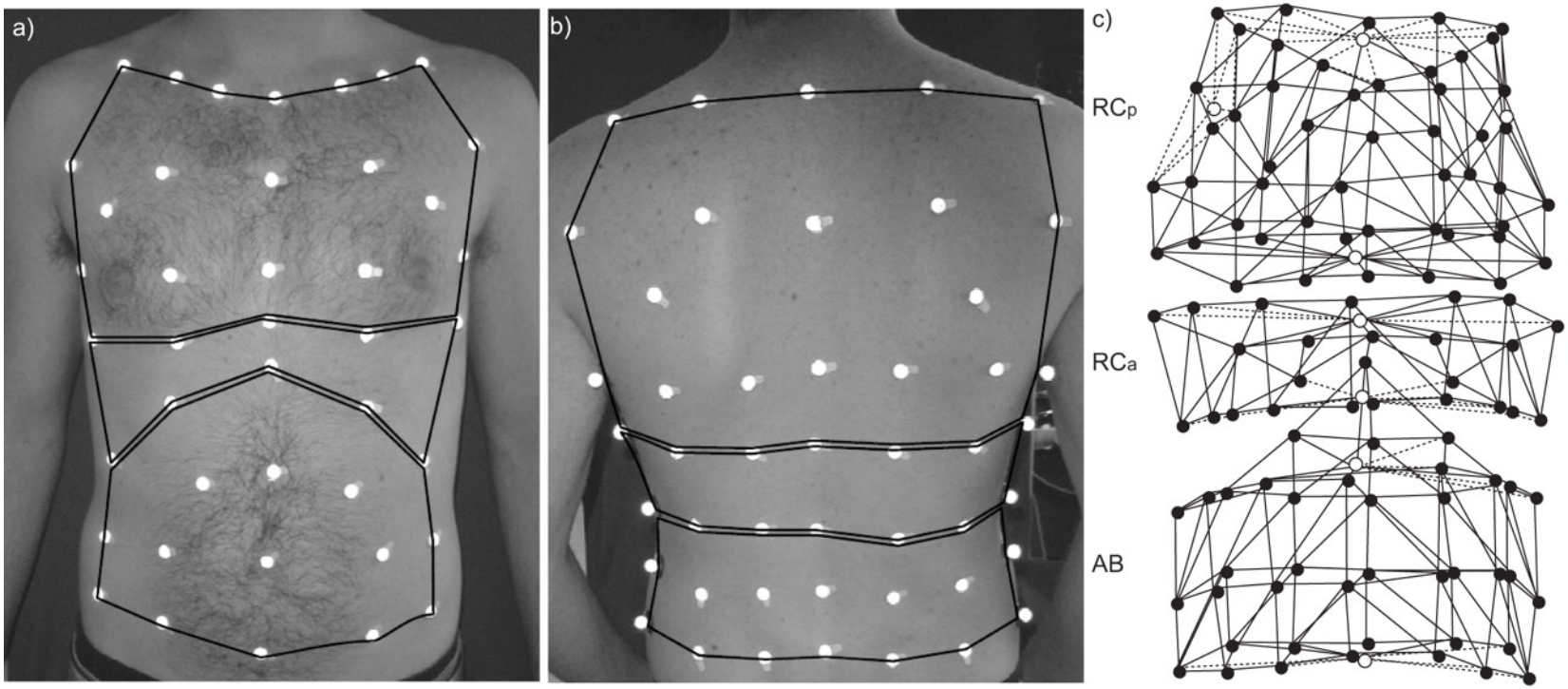


Fig 5. Chest wall and diaphragm contribution to overall lung volume change. The bar plot shows the amount of volume displaced by the costal surface (upper bars) and diaphragm surface (lower bars) for all subjects. Patients (*P01—P10*) and controls (*C01—C06*) are sorted within their group in descending order with respect to supine FVC (% of predicted).

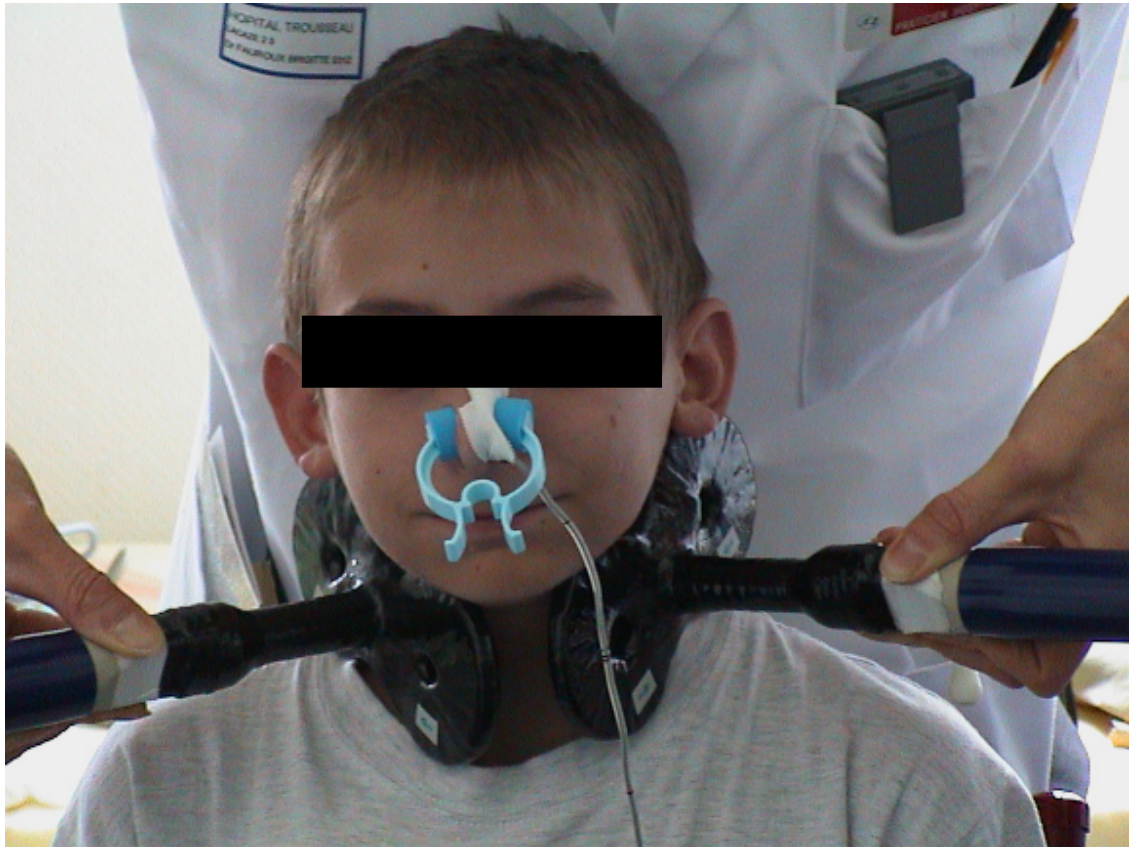
Effects of noninvasive ventilation on treadmill 6-min walk distance and regional chest wall volumes in cystic fibrosis: Randomized controlled trial

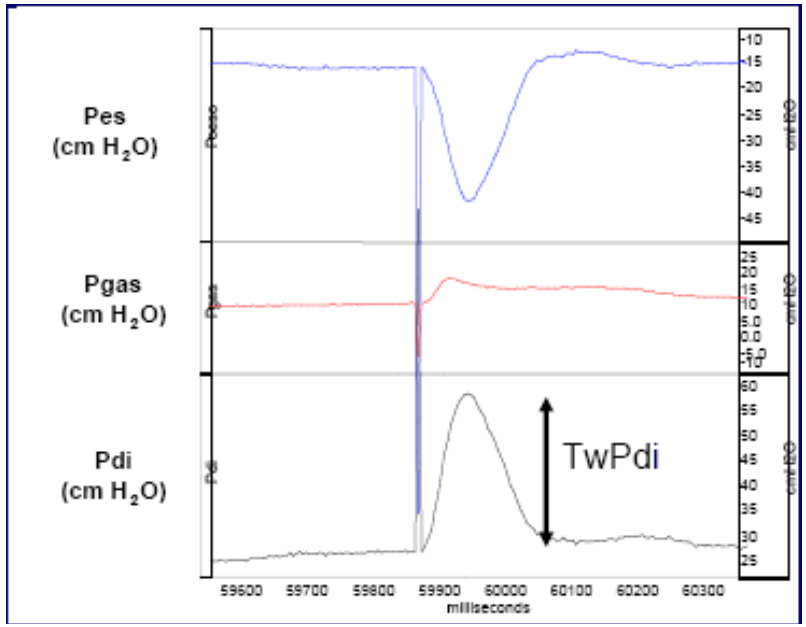
Cibelle Andrade Lima [a],
Armèle de Fátima Dornelas de Andrade [b,*],
Shirley Lima Campos [b], Daniella Cunha Brandão [b],
Guilherme Fregonezi [c], Ianny Pereira Mourato [b],
Andrea Aliverti [d], Murilo Carlos Amorim de Britto [e]



Respiratory Medicine (2014) **108**, 1460–1468

Stimulation magnétique des nerfs phréniques (TwPdi)



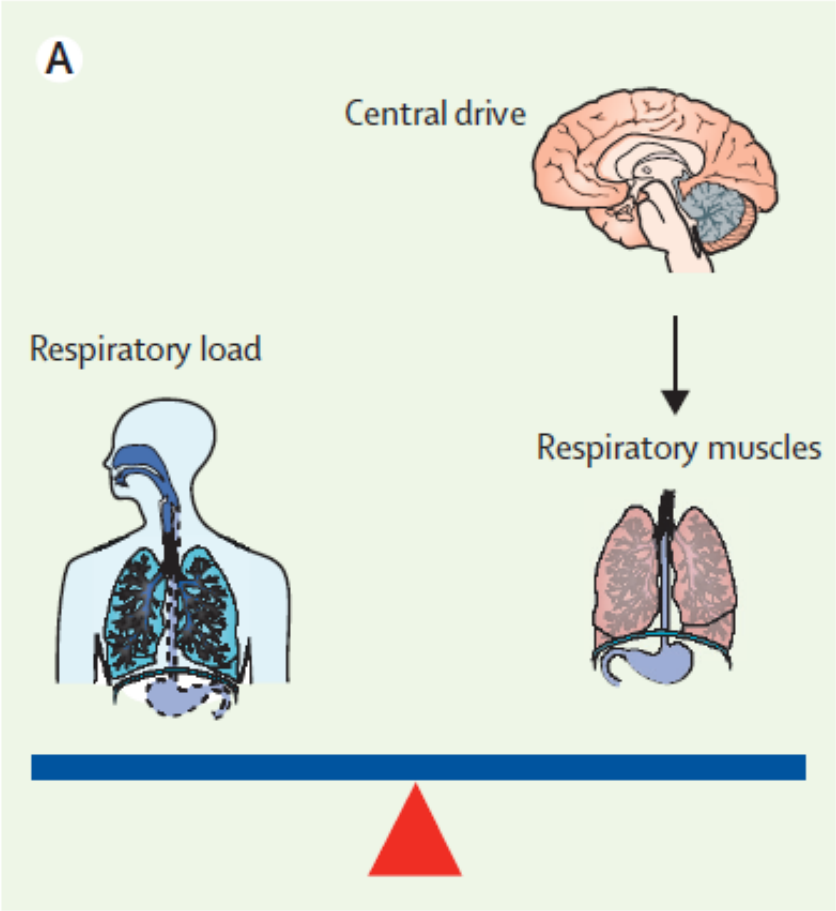


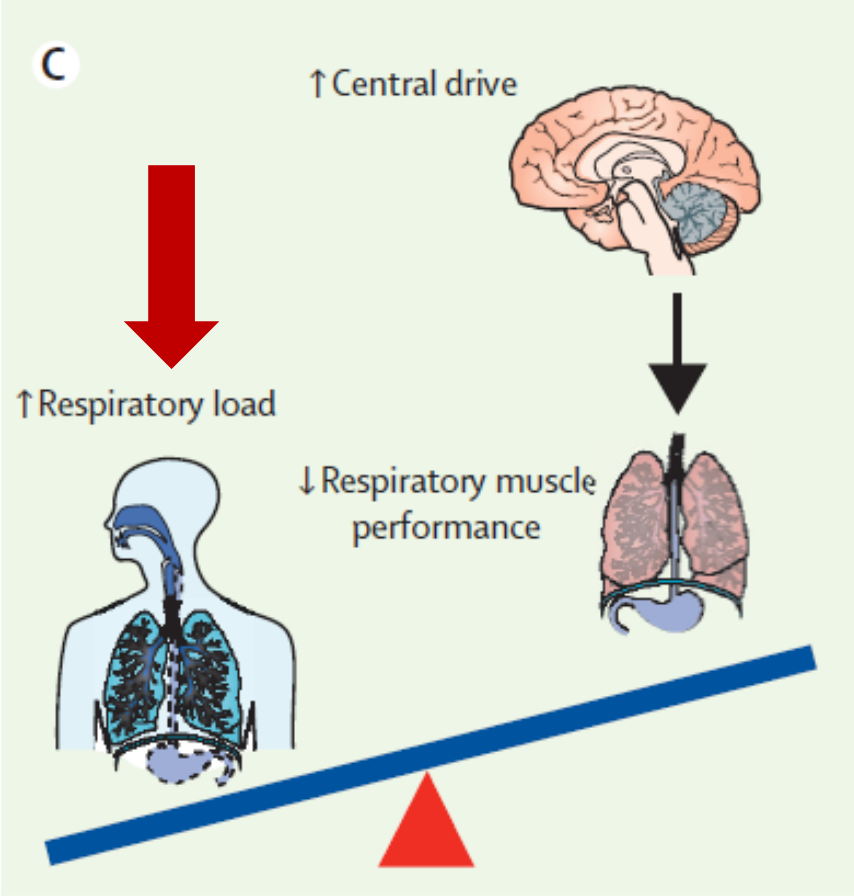
Explorer les muscles respiratoires

- Quelles explorations ?
- Quelle est la performance des muscle respiratoires dans la CF ?
- Quels sont les déterminants de la performance des muscles respiratoires ?
- En pratique…

Long-term non-invasive ventilation in children

Alessandro Amaddeo, Annick Frapin, Brigitte Fauroux

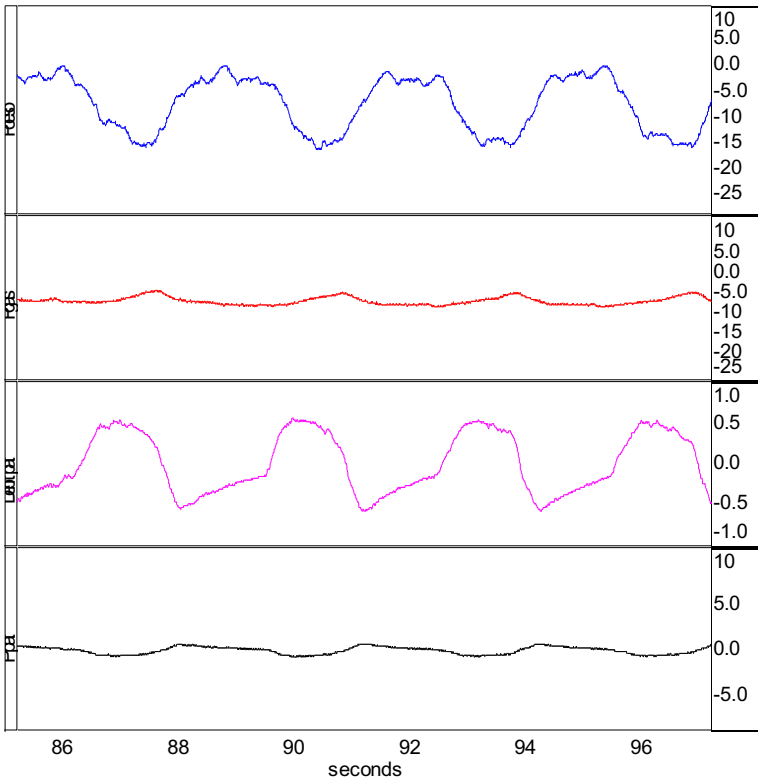


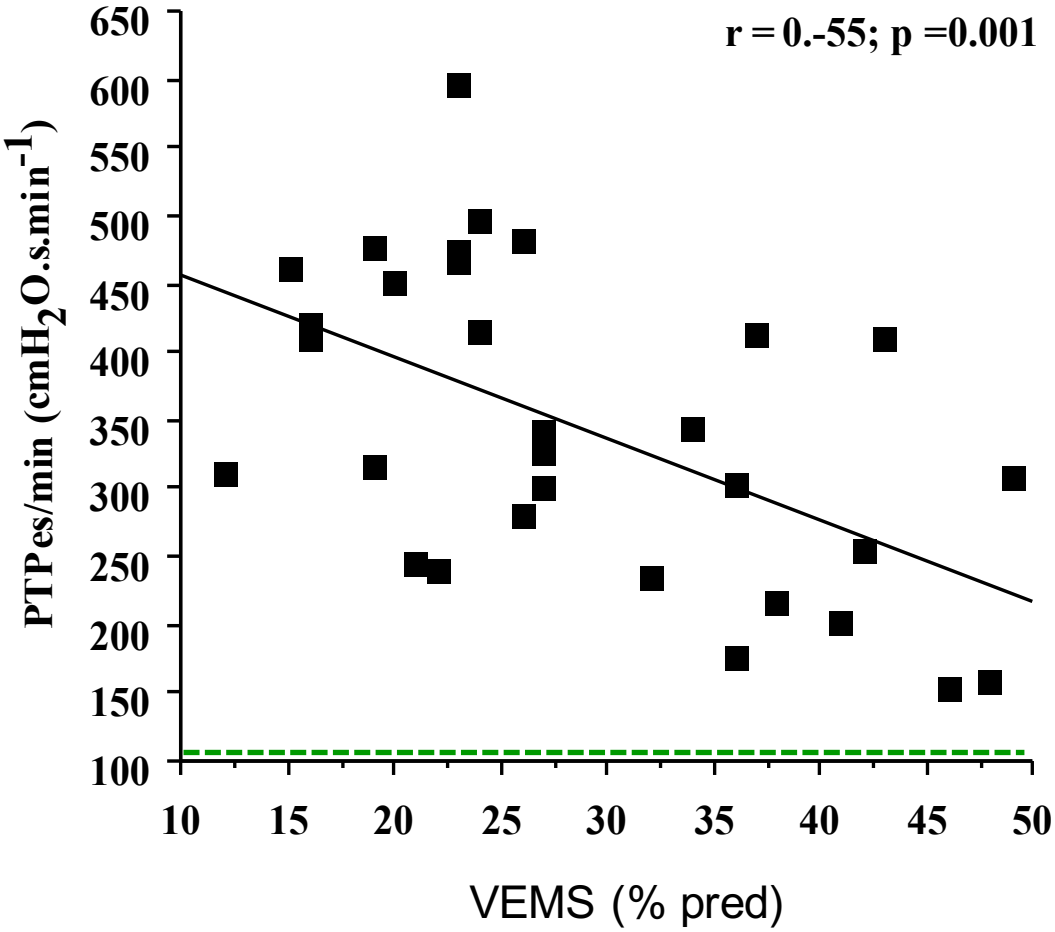


www.thelancet.com/respiratory Published online July 13, 2016 http://dx.doi.org/10.1016/S2213-2600(16)30151-5

Changes in Pulmonary Mechanics with Increasing Disease Severity in Children and Young Adults with Cystic Fibrosis

Nicholas Hart, Michael I. Polkey, Annick Clément, Michèle Boulé, John Moxham, Frédéric Lofaso, and Brigitte Fauroux

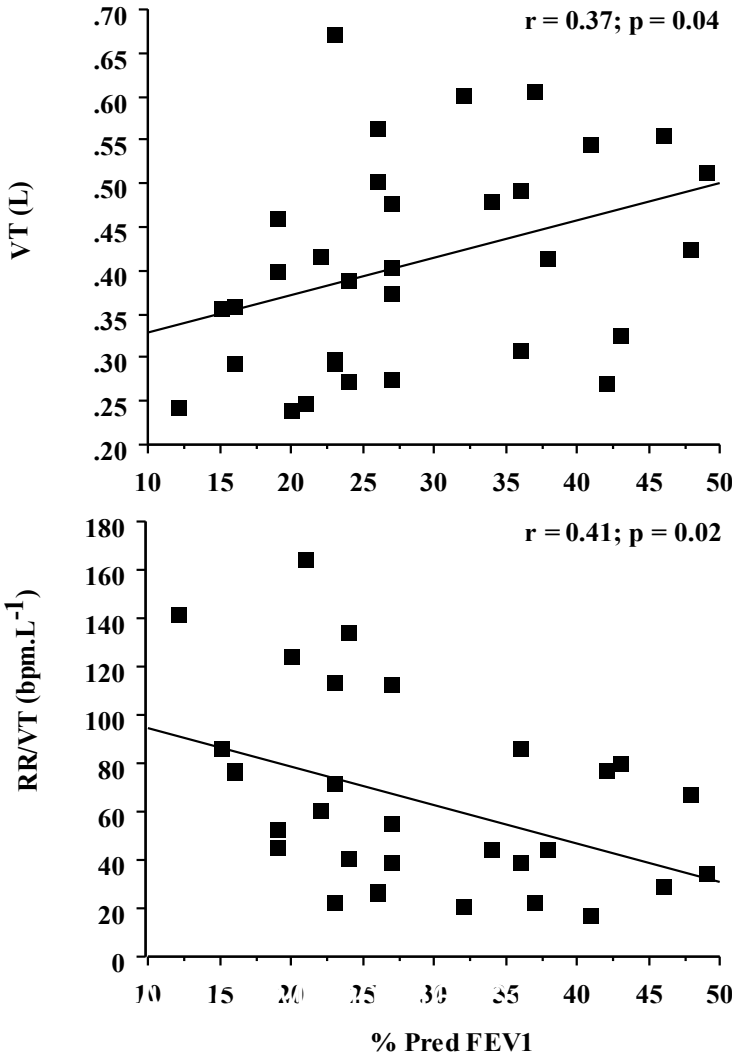


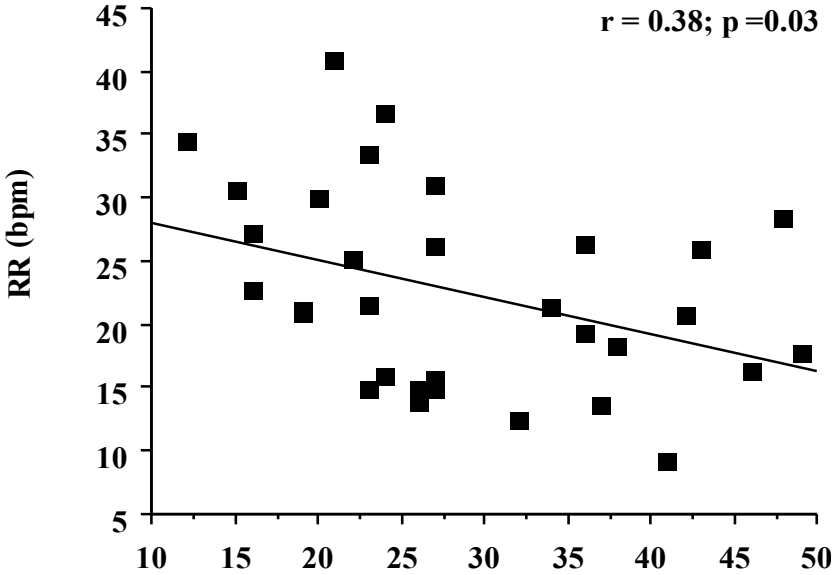


Am J Respir Crit Care Med Vol 166. pp 61–66, 2002

Changes in Pulmonary Mechanics with Increasing Disease Severity in Children and Young Adults with Cystic Fibrosis

Nicholas Hart, Michael I. Polkey, Annick Clément, Michèle Boulé, John Moxham, Frédéric Lofaso, and Brigitte Fauroux

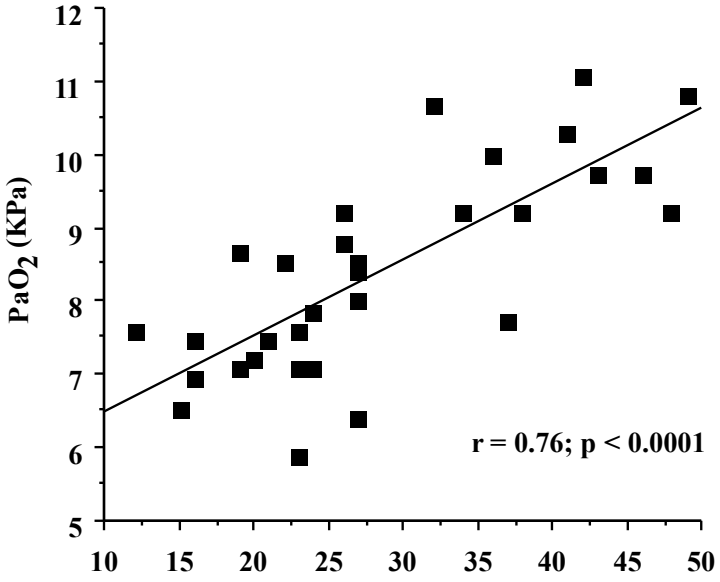


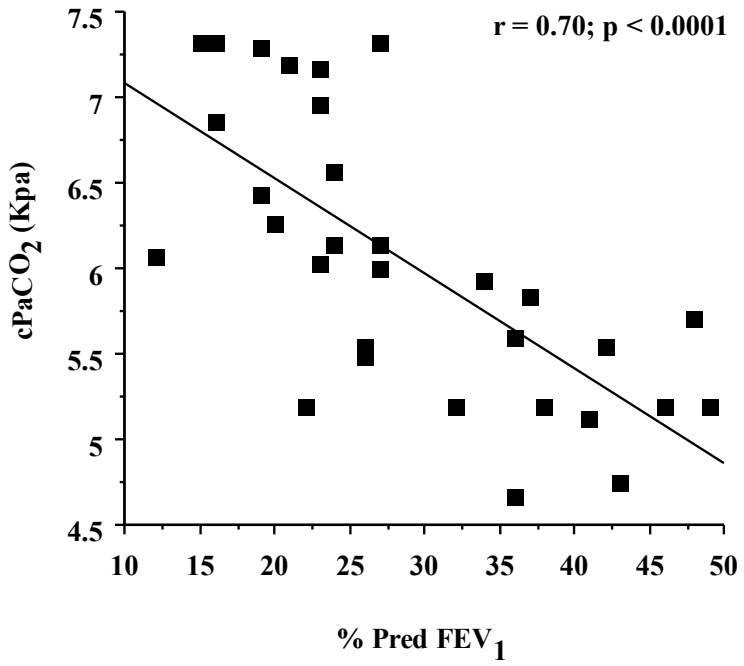


Am J Respir Crit Care Med Vol 166. pp 61–66, 2002

Changes in Pulmonary Mechanics with Increasing Disease Severity in Children and Young Adults with Cystic Fibrosis

Nicholas Hart, Michael I. Polkey, Annick Clément, Michèle Boulé, John Moxham, Frédéric Lofaso, and Brigitte Fauroux



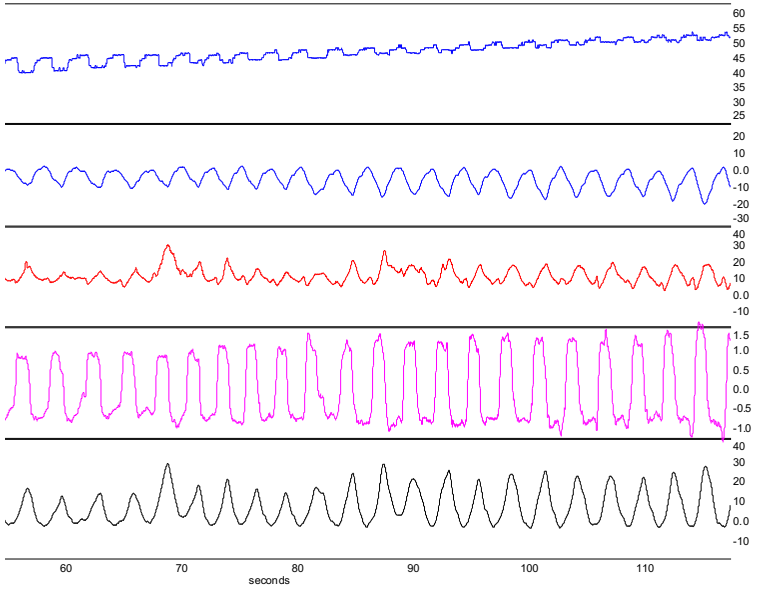


Am J Respir Crit Care Med Vol 166. pp 61–66, 2002

RESPIRATORY PHYSIOLOGY & NEUROBIOLOGY

Mechanical limitation during CO_2 rebreathing in young patients with cystic fibrosis

Brigitte Fauroux [a,b,*], Frédéric Nicot [b], Pierre-Yves Boelle [c], Michèle Boulé [b], Annick Clément [a,b], Frédéric Lofaso [d], Monique Bonora [b]



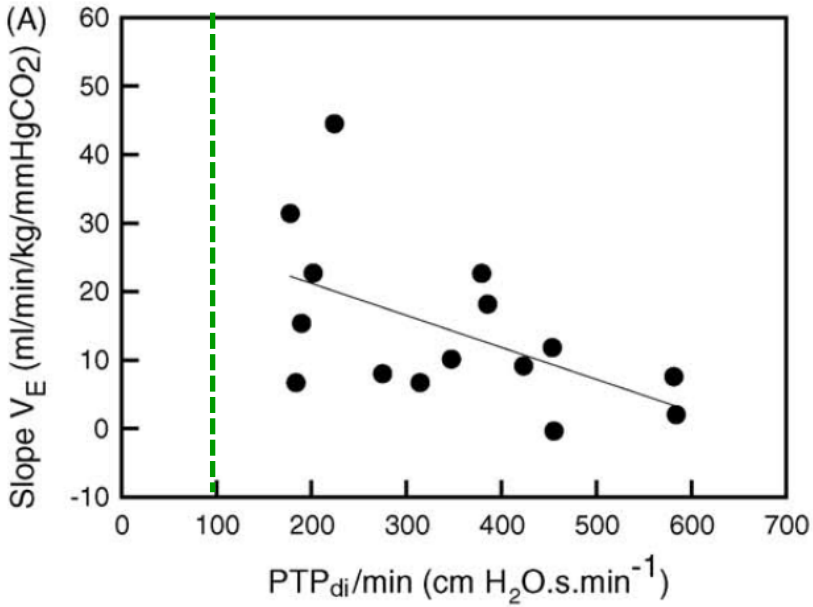
$PetCO_2$ (mmHg)

Pes (cmH_2O)

Pgas (cmH_2O)

Flow (L/s)

Pdi (cmH_2O)



Respiratory Physiology & Neurobiology 153 (2006) 217–225

Respiratory Muscle Function in Patients With Cystic Fibrosis

Theodore Dassios, MD,[1,2] Anna Katelari, MD,[3] Stavros Doudounakis, MD,[3] Stefanos Mantagos, MD,[2] and Gabriel Dimitriou, MD, PhD[2]*

	Healthy subjects (N = 140)	Cystic fibrosis (N = 140)
Gender (male)	82 (58.6)	68 (48.6)
Age (years)	13 (10–17)	14 (10–17)
Height (cm)	160 (148–169)	156 (140–166)
Weight (kg)	51 (40–63)	49 (36–59)
BMI z-score	0.42 (−0.27–1.20) [N = 114]	0.22 (−0.51–0.87) [N = 119]
BMI	22.1 (19.8–24.1) [N = 26]	21.3 (20.2–23.6) [N = 21]
MAMC (cm)	25.0 (23.0–27.0)	22.0 (19.5–24.5)
TST (mm)	14 (11–18)	12 (8–15)
UAMA (mm^2)	3,138 (2,628–3,892)	2,608 (1,955–3,387)
SaO_2 (%)	98 (98–99)	98 (97–99)
FVC (%pred)	N/A	103.6 (83.5–118.5)
FEV_1 (%pred)	N/A	100.3 (74.4–117.0)
Pi_{max} (cmH_2O)	84 (66–102)	74 (57–94)
Pi_{mean}/Pi_{max}	0.223 (0.160–0.313)	0.255 (0.178–0.359)
PTI_{mus}	0.094 (0.070–0.137)	0.110 (0.076–0.160)
Pe_{max} (cmH_2O)	84 (66–102)	71 (50–95)

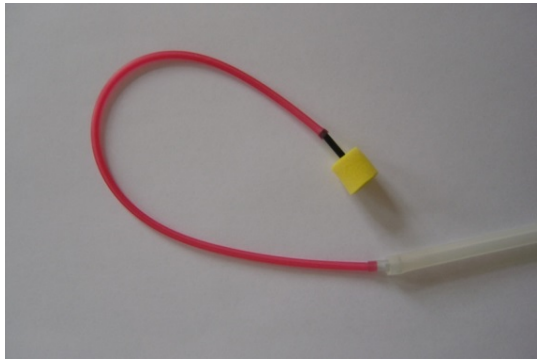
Pediatric Pulmonology 48:865–873 (2013)

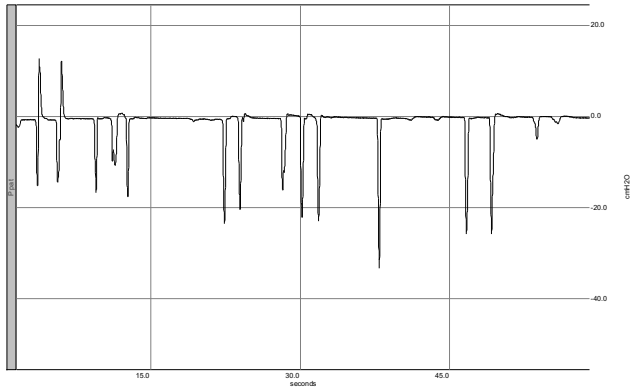
Sniff nasal inspiratory pressure in children with muscular, chest wall or lung disease

B. Fauroux*,#, G. Aubertin*, E. Cohen*,#, A. Clément*,# and F. Lofaso¶,+

Eur Respir J 2009; 33: 113–117

	Neuromuscular disease	Thoracic scoliosis	Cystic fibrosis
Subjects n	78	12	23
Age yrs	12.7±3.7 (4–18)	14.5±2.5 (9–18)	13.8±2.9 (7–18)
Female/male n	20/58	10/2	13/10
Weight kg	42.2±3.7	49.5±11.6	38.3±9.7
Height cm#	146±4	163±11	150±10
FVC % pred	54±29¶	55±25+	38±29§
FEV_1 % pred	64±35¶	50±24+	30±29§
FEV_1/FVC % pred	107±29¶	103±55+	75±17§
Sniff *P*oes cmH_2O	49±4f	82±25	93±29
SNIP cmH_2O	41±4f	70±25	66±29
SNIP/Sniff *P*oes ratio	0.83±0.17	0.86±0.10	0.72±0.13##





Nutritional status is an important predictor of diaphragm strength in young patients with cystic fibrosis[1–3]

Nicholas Hart, Patrick Tounian, Annick Clément, Michèle Boulé, Michael I Polkey, Frédéric Lofaso, and Brigitte Fauroux

Anthropometric characteristics of the 20 patients with cystic fibrosis[1]

VEMS 44 ± 21%

	Value (n = 11 F, 9 M)
Age (y)	15.1 ± 2.8
Tw Pdi (cm H_2O)	24.3 ± 5.5
Weight (kg)	38.6 ± 9.6
Height (cm)	151 ± 12
BMI	
(kg/m^2)	16.60 ± 2.23
(z score)	−1.1 ± 1.3
AMC (% of predicted)	92.6 ± 9.0
FFM (% of predicted)	94.2 ± 8.2
FM (% of predicted)	85.1 ± 30.4

Am J Clin Nutr 2004;80:1201–6.

Muscles respiratoires dans la mucoviscidose

- Augmentation de la charge imposée aux muscles respiratoires
- Les muscles respiratoires sont très performants et « entrainés »
- Au delà d'un certain seuil: apparition d'une diminution de la performance des muscles respiratoires

Explorer les muscles respiratoires

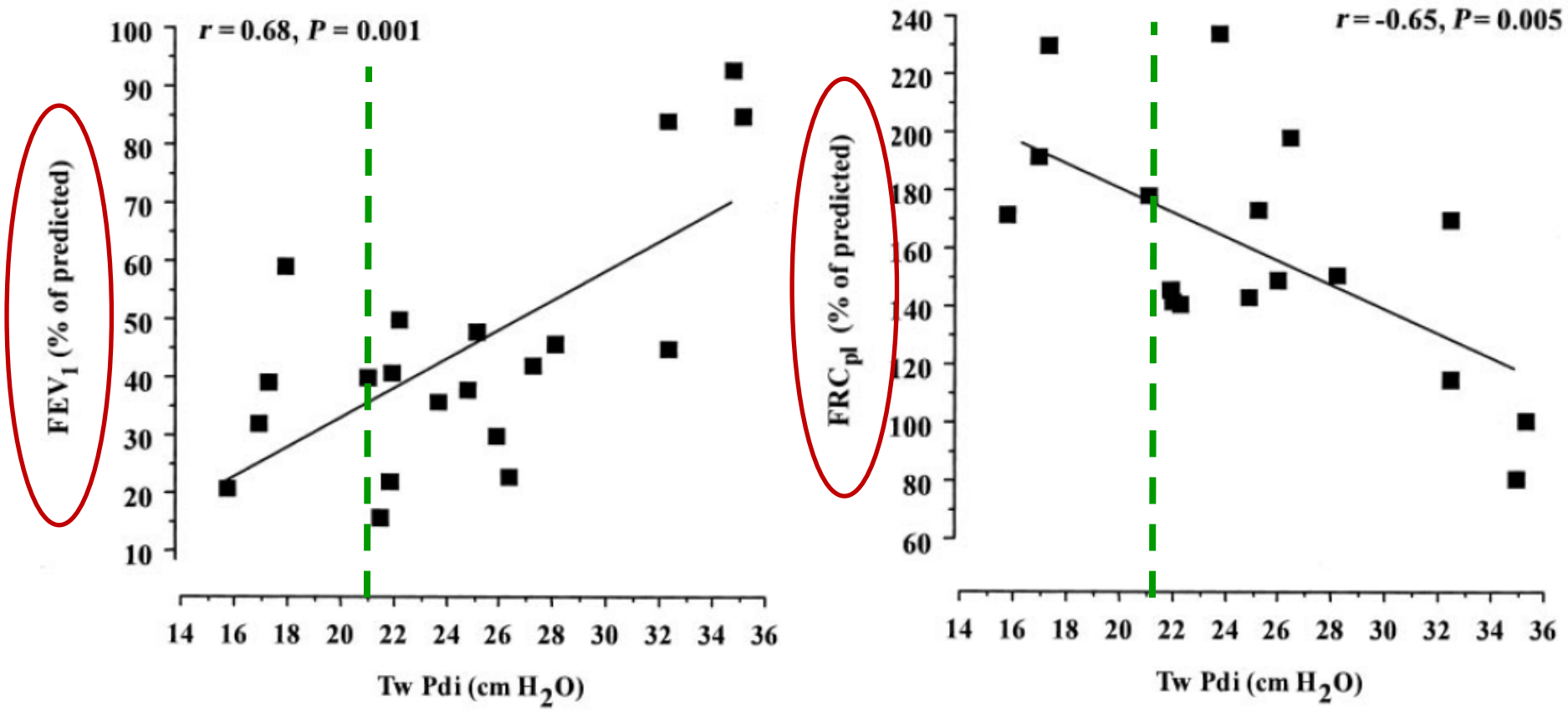
- Quelles explorations ?
- Quelle est la performance des muscle respiratoires dans la CF ?
- Quels sont les déterminants de la performance des muscles respiratoires ?
- En pratique…

Déterminants de la performance des muscles respiratoires

- Fonction respiratoire ?
- Statut nutritionnel
- Inflammation / infection
- CFTR

Nutritional status is an important predictor of diaphragm strength in young patients with cystic fibrosis[1–3]

Nicholas Hart, Patrick Tounian, Annick Clément, Michèle Boulé, Michael I Polkey, Frédéric Lofaso, and Brigitte Fauroux

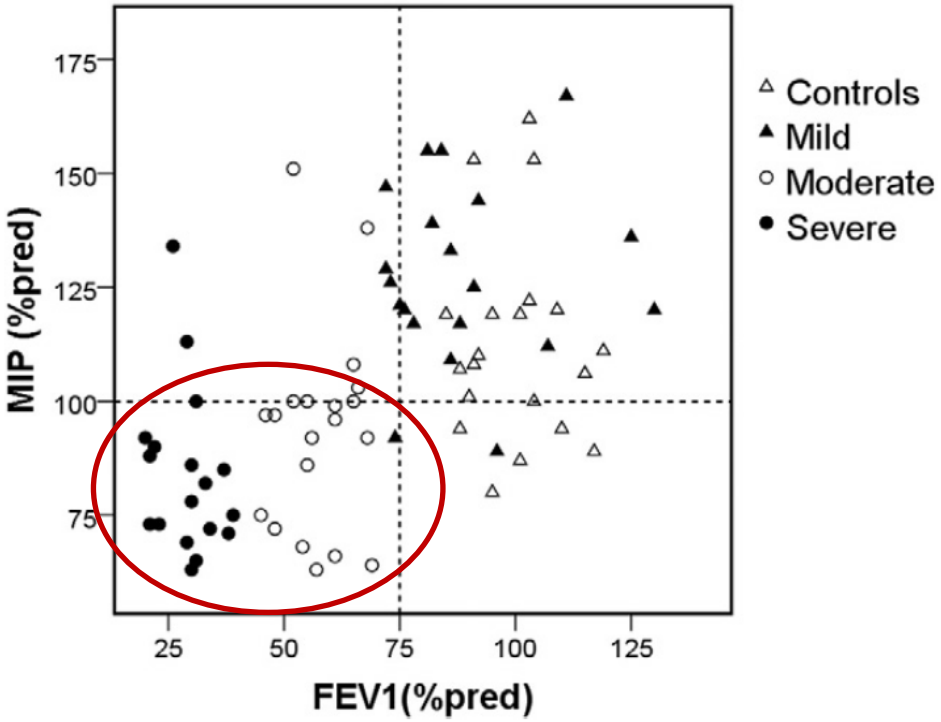


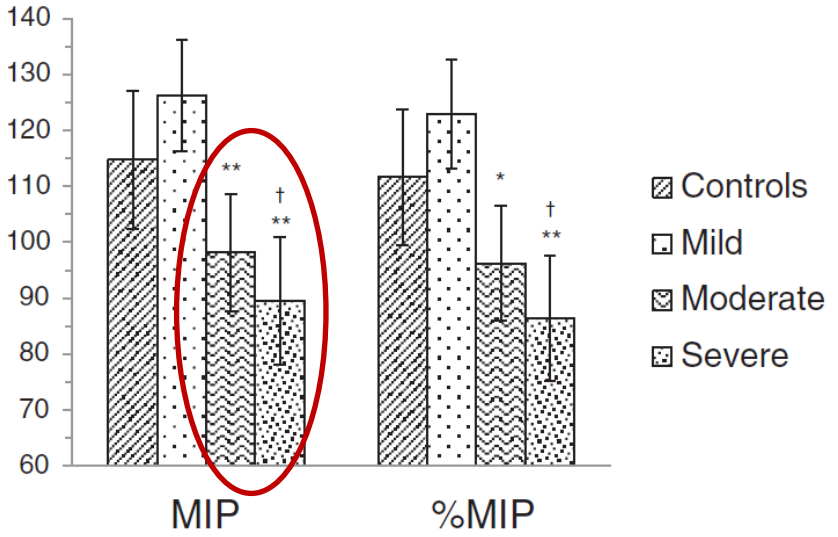
Am J Clin Nutr 2004;80:1201–6.

Inspiratory muscle strength relative to disease severity in adults with stable cystic fibrosis

Journal of Cystic Fibrosis

Robert L. Dekerlegand [a,*], Denis Hadjiliadis [b], Anne K. Swisher [c], J. Scott Parrott
Albert J. Heuer [f], Mary Jane Myslinski [g]





Le VEMS est responsable de 24% de la variance de la PImax après contrôle des autres variables

Journal of Cystic Fibrosis 14 (2015) 639–645

Respiratory Muscle Function in Patients With Cystic Fibrosis

Theodore Dassios, MD,[1,2] Anna Katelari, MD,[3] Stavros Doudounakis, MD,[3] Stefanos Mantagos, MD,[2] and Gabriel Dimitriou, MD, PhD[2*]

TABLE 5—Multivariate Logistic Regression Analysis for the Development of a PTI_{mus} Value Greater Than the 75th Percentile

Parameter	Exp (B)	95% Confidence intervals		*P*-value
FEV_1 (% predicted)	0.979	0.965	0.993	0.004
FVC (% predicted)	0.999	0.948	1.054	0.980
MEF_{25-75} (% predicted)	1.007	0.986	1.030	0.501
UAMA	1.000	0.999	1.000	0.364
Sat O_2 (%)	0.904	0.695	1.177	0.455

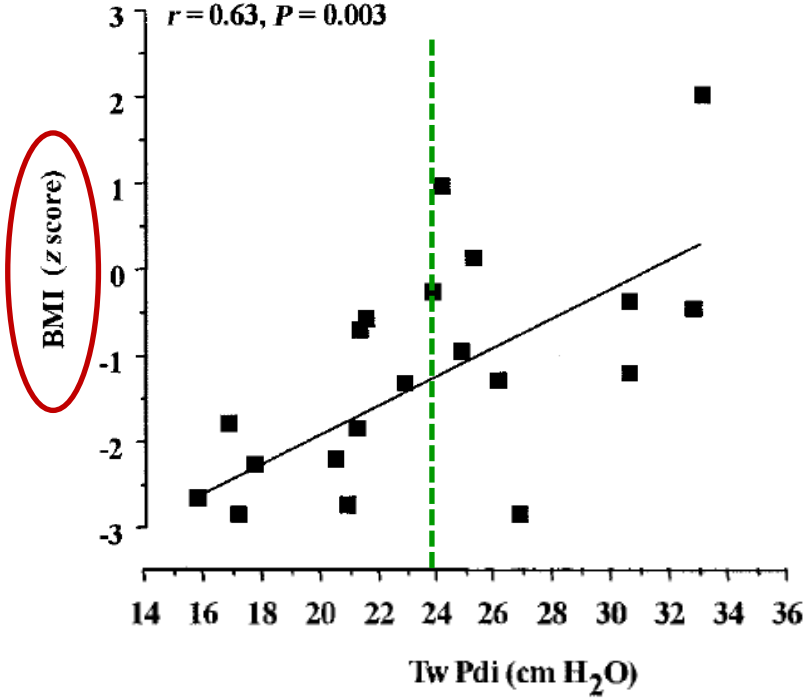
Pediatric Pulmonology 48:865–873 (2013)

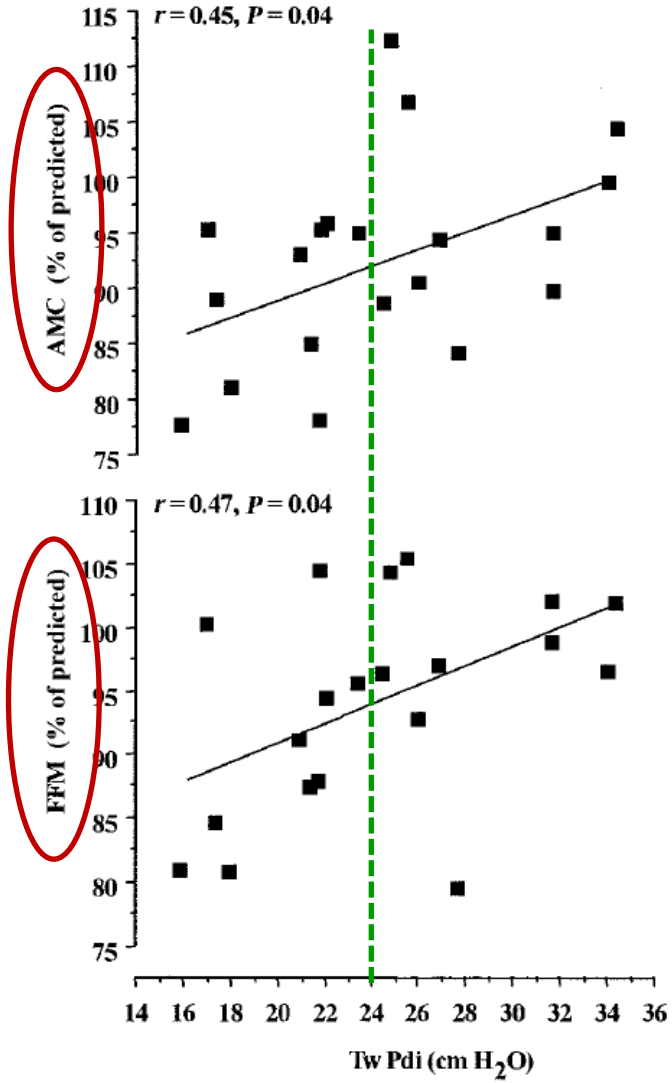
Déterminants de la performance des muscles respiratoires

- Fonction respiratoire: OUI
- Statut nutritionnel ?
- Inflammation / infection
- CFTR

Nutritional status is an important predictor of diaphragm strength in young patients with cystic fibrosis[1–3]

Nicholas Hart, Patrick Tounian, Annick Clément, Michèle Boulé, Michael I Polkey, Frédéric Lofaso, and Brigitte Fauroux



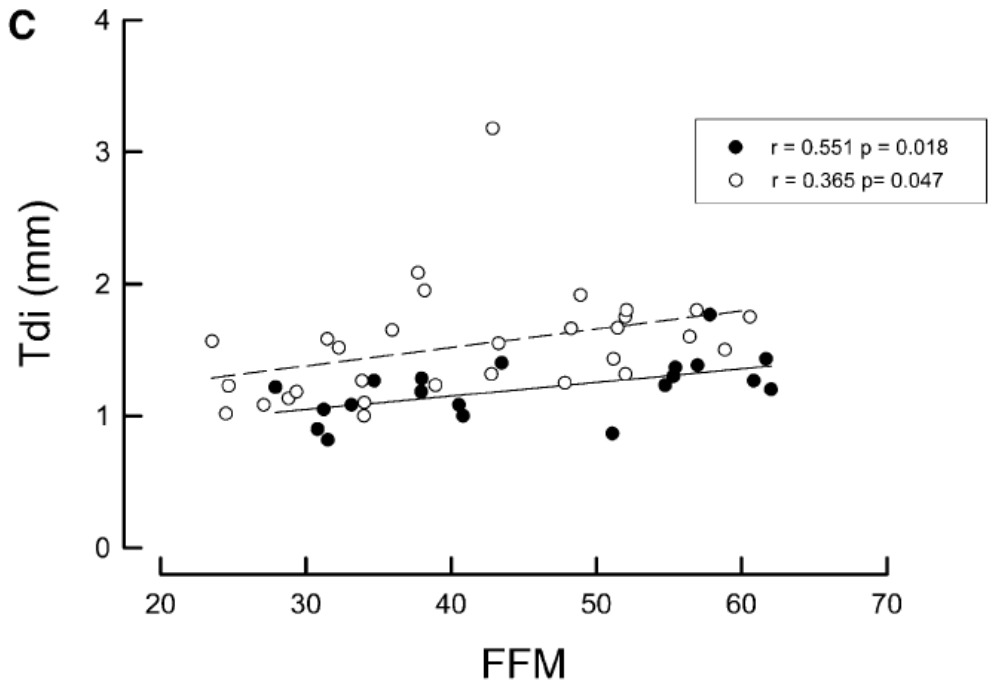


Am J Clin Nutr 2004;80:1201–6.

Effect of Systemic Inflammation on Inspiratory and Limb Muscle Strength and Bulk in Cystic Fibrosis

Valérie Dufresne[1], Christiane Knoop[1], Alain Van Muylem[1], Anne Malfroot[2], Michel Lamotte[3], Christian Opdekamp[3], Gaël Deboeck[3], Marie Cassart[4], Bernard Stallenberg[4], Georges Casimir[5], Jean Duchateau[6], and Marc Estenne[1]

Diaphragm thickness (ultrasound)



Am J Respir Crit Care Med Vol 180. pp 153–158, 2009

○ CF
● témoins

The influence of body composition on respiratory muscle, lung function and diaphragm thickness in adults with cystic fibrosis ☆

Stephanie Enright [a,*], Ken Chatham [b,1], Alina A. Ionescu [d,2], Viswanath B. Unnithan [c,3], Dennis J. Shale [d,2]

Journal of Cystic Fibrosis

Variable	CF patients low FFM (n=22)	CF patients normal FFM (n=18)	Healthy subjects (n=30)
FEV_1 (% pred)	36.7 (36.1–37.4)[b,c]	56.3 (55.8–56.8)[c,d]	96.2 (95.2–97.2)
VC (litres)	3.1 (2.9–3.3)[b,c]	4.0 (3.8–4.2)[d,e]	4.1 (3.2–4.5)
RV (litres)	1.8 (1.6–2.0)	1.6 (1.4–1.8)	1.5 (1.4–1.6)
TLC (litres)	4.1 (3.9–4.3)[b,c]	5.1 (4.9–5.3)[d,e]	5.8 (5.5–6.0)
FRC (litres)	2.5 (2.1–2.9)	2.8 (2.2–3.4)	2.9 (2.6–3.1)
Diaph (TDIrel) (mm)	2.3 (2.0–2.6)[b,c]	3.2 (2.7–3.7)	3.4 (3.0–3.8)
Diaph (TDIcont) (mm)	3.3 (3.1–3.5)[b,c]	4.3 (3.9–4.7)	4.4 (4.2–4.6)
TR (%)	2.4 (2.3–2.5)[e]	2.8 (2.6–2.8)	2.9 (2.7–3.2)
PImax (cm H_2O)	109 (100 –118)[b,c]	119 (111–127)[d,e]	124 (119–129)
SPImax (PTU)	520 (462–584)[b,c]	698 (634–762)[d,e]	788 (733–843)

Journal of Cystic Fibrosis 6 (2007) 384 – 390

Déterminants de la performance des muscles respiratoires

- Fonction respiratoire: OUI
- Statut nutritionnel: OUI
- Inflammation / infection ?
- CFTR

Effect of Systemic Inflammation on Inspiratory and Limb Muscle Strength and Bulk in Cystic Fibrosis

Valérie Dufresne[1], Christiane Knoop[1], Alain Van Muylem[1], Anne Malfroot[2], Michel Lamotte[3], Christian Opdekamp[3], Gaël Deboeck[3], Marie Cassart[4], Bernard Stallenberg[4], Georges Casimir[5], Jean Duchateau[6], and Marc Estenne[1]

TABLE 2. LEUKOCYTOSIS AND SERUM INFLAMMATORY MARKERS IN STUDY POPULATION

	Patients with CF	Control Subjects	*P* Value
Leukocytes, per mm^3	9,600 ± 3,600*	6,200 ± 2,400	<0.001
PMN, per mm^3	6,400 ± 3,200	3,600 ± 2,200	<0.001
ESR, mm/hour	21 ± 20	6 ± 5	0.001
CRP, mg/dl	0.8 (0.2–2.6)	0.1 (0.06–0.2)	<0.001
IL-6, pg/ml	2.3 (0.7–8.0)	0.04 (0.004–0.3)	<0.001
IL-8, pg/ml	1.7 (0.2–13.6)	0.08 (0.005–1.1)	<0.01
IL-17, pg/ml	0.2 (0.02–2.0)	0.05 (0.06–0.5)	0.019
TNF-α, pg/ml	2.4 (1.1–5.0)	1.7 (0.5–5.3)	0.1
sTNF-αRI, ng/ml	1.9 (1.4–2.6)	1.5 (1.2–1.8)	<0.001
sTNF-αRII, ng/ml	5.7 (4.0–8.1)	4.4 (3.2–5.9)	0.005

38 adultes CF et 20 témoins
VEMS moyen 49 ± 25%

TABLE 3. STRENGTH AND BULK OF RESPIRATORY AND LIMB MUSCLES IN STUDY POPULATION

	Patients with CF	Control Subjects	*P* Value
BicCSA, cm^2	8.4 ± 3.3	9.4 ± 3.4	NS
BicCSA/FFM, cm^2/kg	0.21 ± 0.04	0.21 ± 0.03	NS
BicPT, N	194 ± 72	226 ± 83	NS
QuadCSA, cm^2	51.8 ± 15.2	59.3 ± 13.1	0.05
QuadCSA/FFM, cm^2/kg	1.30 ± 0.14	1.35 ± 0.20	NS
QuadPT, N	356 ± 127	402 ± 128	NS
Tdi,* mm	1.5 ± 0.4	1.4 ± 0.3	0.04
Tdi/FFM, mm/kg	0.039 ± 0.011	0.031 ± 0.007	<0.001
PIMAX,* cm H_2O	100 ± 29	83 ± 28	0.002

⇨ Pas de corrélation entre les marqueurs inflammatoires et la performance des muscles respiratoires

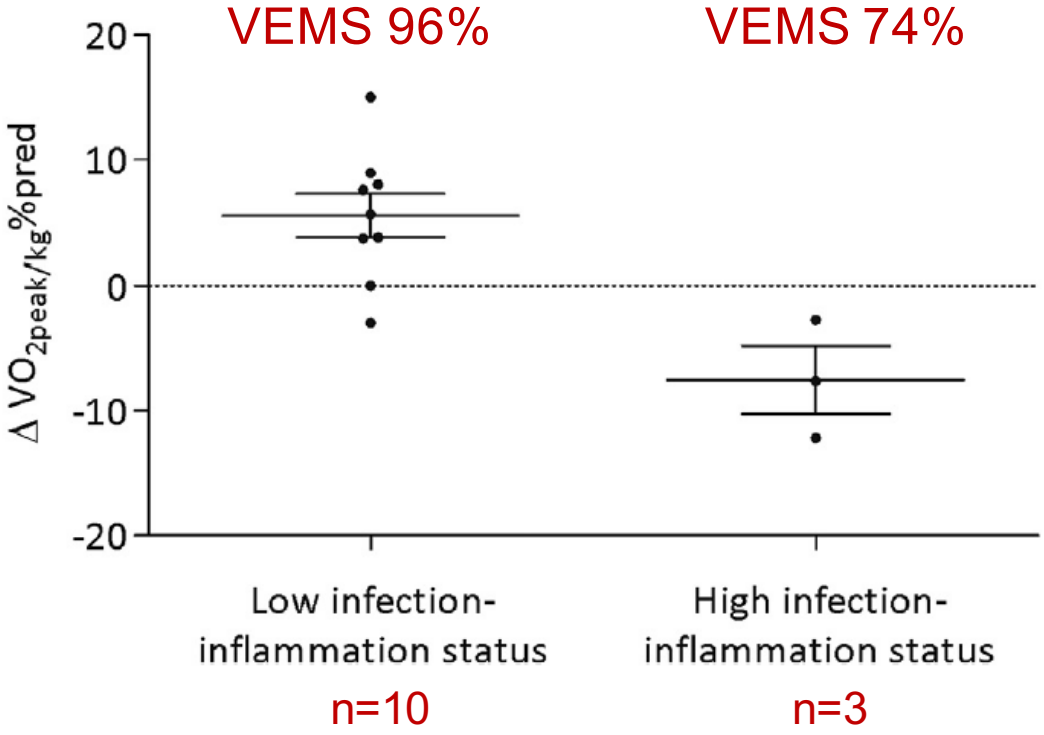
Am J Respir Crit Care Med Vol 180. pp 153–158, 2009

Chronic inflammation and infection associate with a lower exercise training response in cystic fibrosis adolescents

Pauline B. van de Weert-van Leeuwen [a,b,c,*], Hendrikus J. Hulzebos [d], Maarten S. Werkman [d], Sabine Michel [a], Lodewijk A.W. Vijftigschild [a,b,c], Marit A. van Meegen [a,b,c], Cornelis K. van der Ent [a], Jeffrey M. Beekman [a,b,c], Hubertus G.M. Arets [a]

13 adolescents CF
- durée 12 semaines

« High » status
IgG > 10.5 g/L + 50%
cultures PA +



Respiratory Medicine (2014) 108, 445–452

Déterminants de la performance des muscles respiratoires

- Fonction respiratoire: OUI
- Statut nutritionnel: OUI
- Inflammation / infection: NON
- CFTR ?

Cystic Fibrosis Transmembrane Conductance Regulator in Human Muscle Dysfunction Causes Abnormal Metabolic Recovery in Exercise

Anne-Marie Lamhonwah, PhD,[1,2] Christine E. Bear, PhD,[3] Ling Jun Huan, BSc,[3] Patrick Kim Chiaw, BSc,[3] Cameron A. Ackerley, PhD,[4] and Ingrid Tein, MD[1,2]

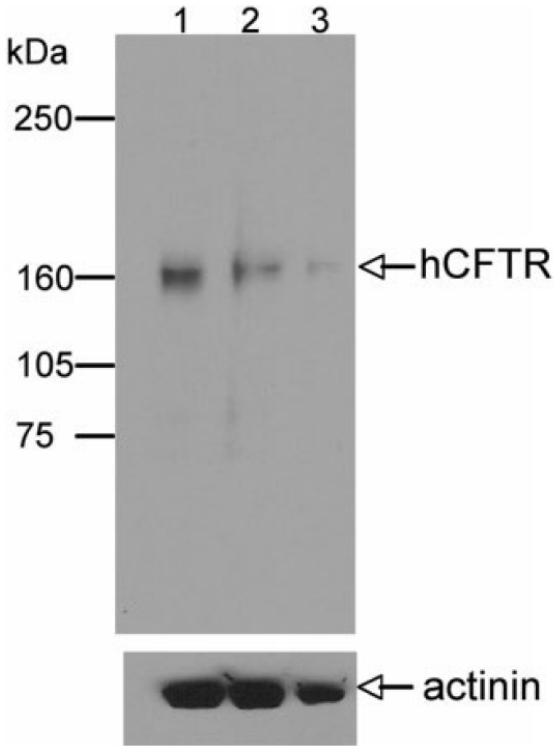


FIGURE 1: Expression of human cystic fibrosis transmembrane conductance regulator (hCFTR) in skeletal muscle by Western blot. Human skeletal muscle homogenates (200μg

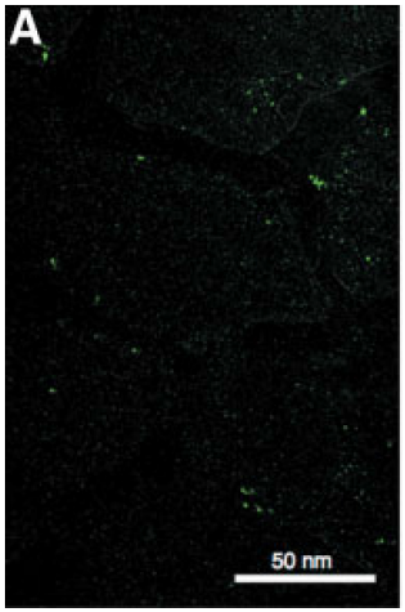
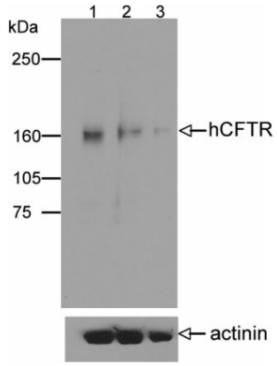


FIGURE 3: Expression of cystic fibrosis transmembrane conductance regulator (CFTR) protein in the sarcoplasm of human muscle using confocal microscopy. Panel (A) shows immunofluorescence microscopy of transverse sections of normal human control muscle treated with a primary mouse anti-human CFTR antibody (L12B4 from Millipore)

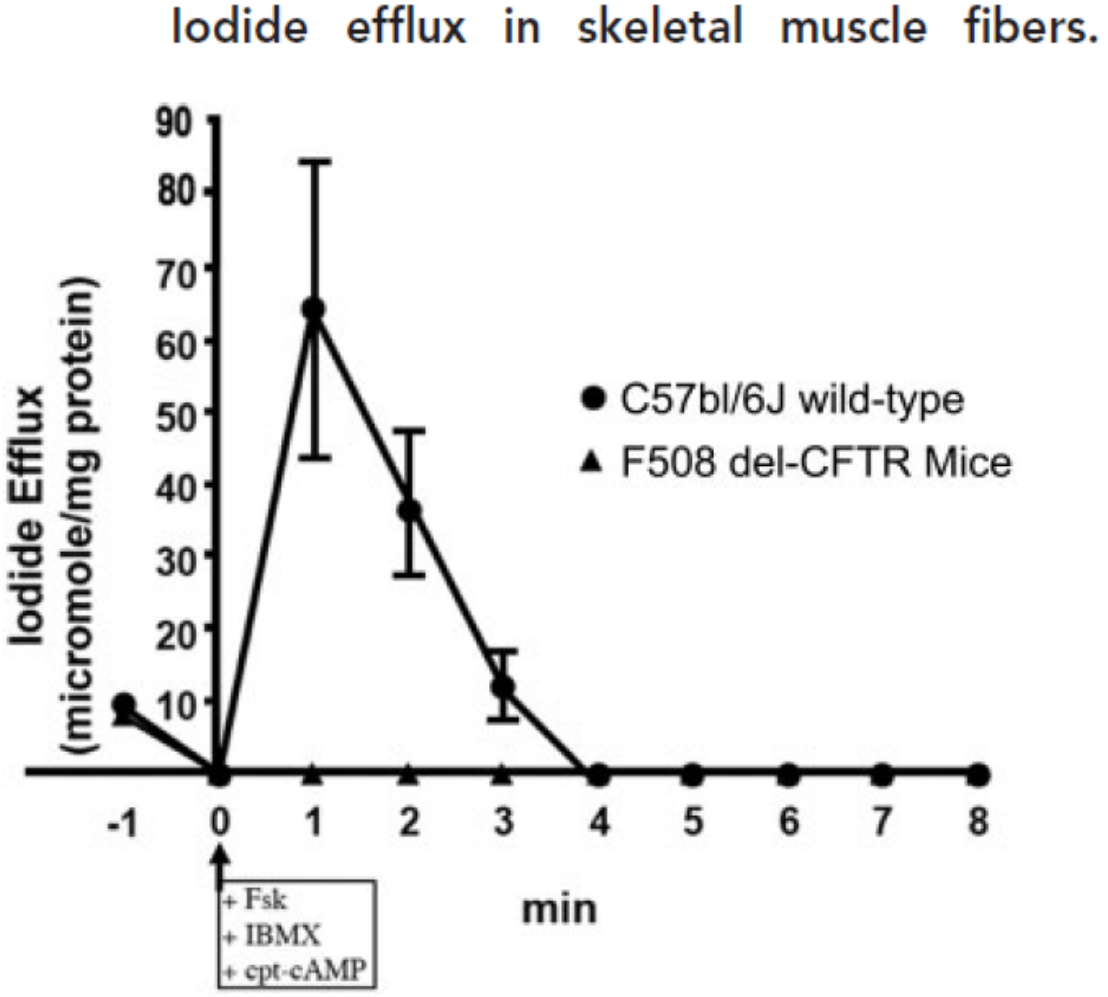
Cystic Fibrosis Transmembrane Conductance Regulator in Human Muscle Dysfunction Causes Abnormal Metabolic Recovery in Exercise

Anne-Marie Lamhonwah, PhD,[1,2] Christine E. Bear, PhD,[3] Ling Jun Huan, BSc,[3] Patrick Kim Chiaw, BSc,[3] Cameron A. Ackerley, PhD,[4] and Ingrid Tein, MD[1]



CFTR est exprimé dans le muscle squelettique

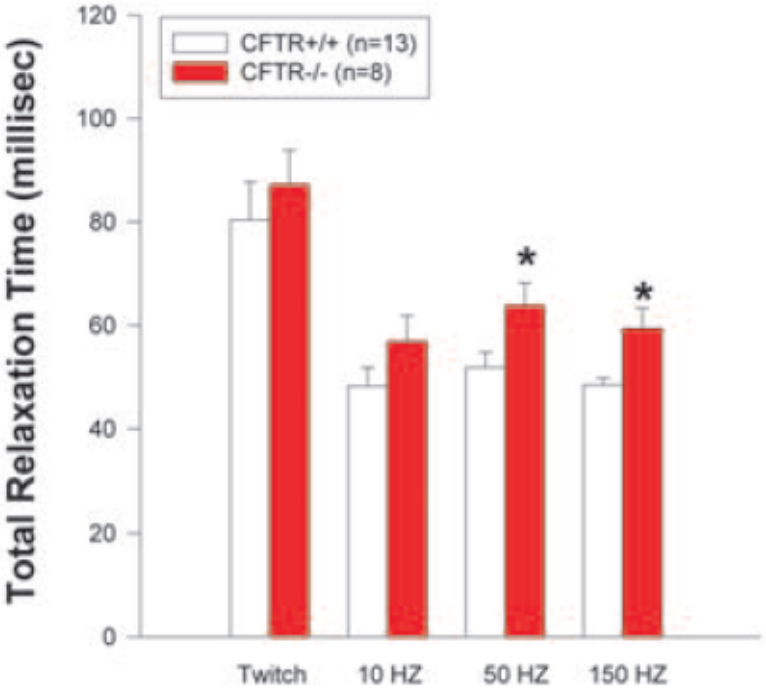
Hypothèse: cette anomalie du canal Cl- CFTR pourrait perturber le gradient électrochimique entrainant une dysrégulation de l'homéostasie du Ca+ qui perturberait la contraction et l'endurance musculaire.

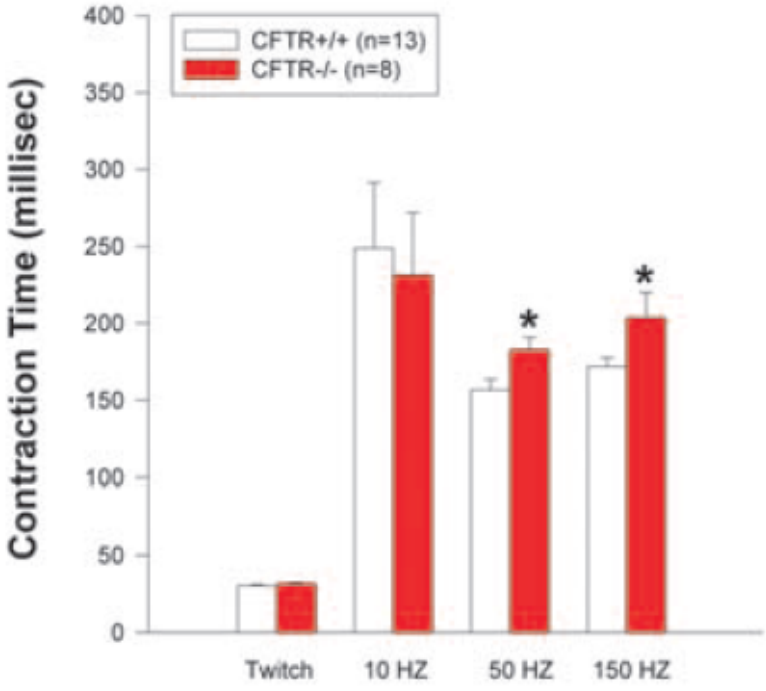


ANN NEUROL 2010;67:802–808

Lack of CFTR in Skeletal Muscle Predisposes to Muscle Wasting and Diaphragm Muscle Pump Failure in Cystic Fibrosis Mice

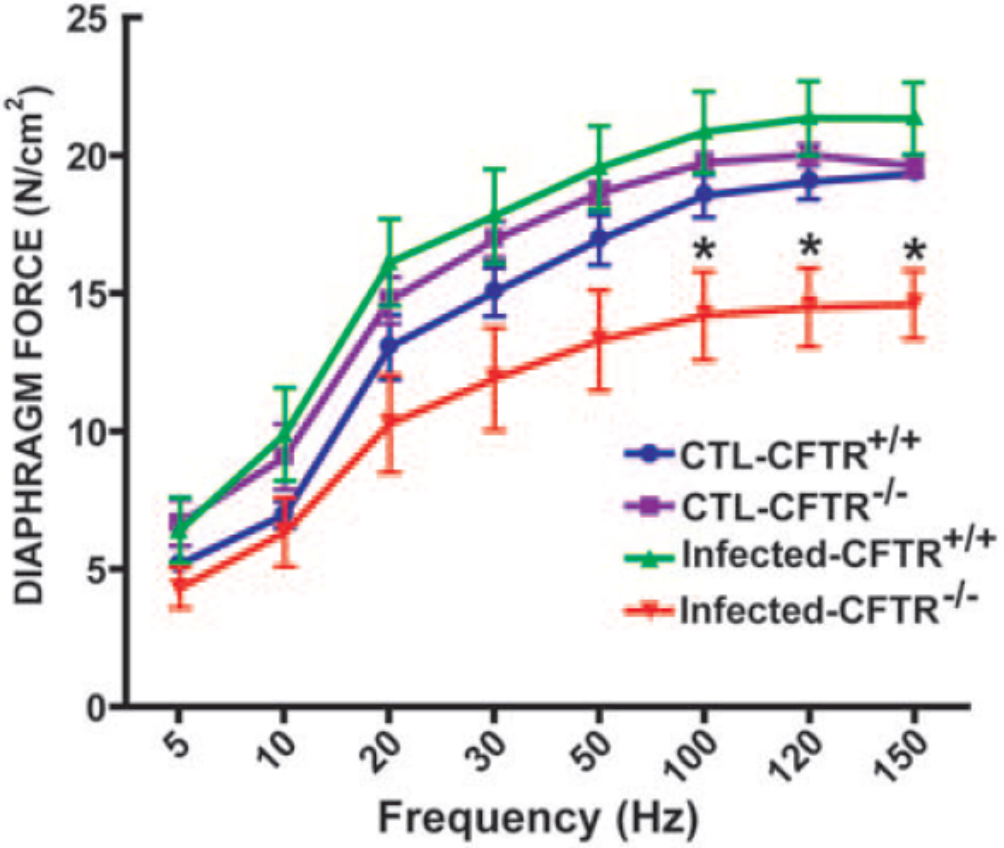
Maziar Divangahi[1☯], Haouaria Balghi[1☯], Gawiyou Danialou[1], Alain S. Comtois[1], Alexandre Demoule[1,2], Sheila Ernest[1], Christina Haston[1], Renaud Robert[3], John W. Hanrahan[3], Danuta Radzioch[4], Basil J. Petrof[1]*





Lack of CFTR in Skeletal Muscle Predisposes to Muscle Wasting and Diaphragm Muscle Pump Failure in Cystic Fibrosis Mice

Maziar Divangahi[1☯], Haouaria Balghi[1☯], Gawiyou Danialou[1], Alain S. Comtois[1], Alexandre Demoule[1,2], Sheila Ernest[1], Christina Haston[1], Renaud Robert[3], John W. Hanrahan[3], Danuta Radzioch[4], Basil J. Petrof[1*]



The Relationship between Genotype and Exercise Tolerance in Children with Cystic Fibrosis

HIRAN C. SELVADURAI, KAREN O. McKAY, CAMERON J. BLIMKIE, PETER J. COOPER, CRAIG M. MELLIS, and PETER P. VAN ASPEREN

Am J Respir Crit Care Med Vol 165. pp 762–765, 2002

TABLE 1. DEMOGRAPHIC DETAILS OF SUBJECTS ACCORDING TO CLASS OF CFTR MUTATION

	II/I	II/II	II/III	II/IV	II/V
n	15	38	17	17	10
Age*	13.8 ± 2.1	14.2 ± 2.6	14.0 ± 3.4	14.2 ± 3.1	13.7 ± 3.5
Male : Female	6 : 4	9 : 11	6 : 9	5 : 4	5 : 3
PI : PS	15 : 0	38 : 0	16 : 1	3 : 14‡	2 : 8‡
Height, m	1.50 ± 0.17	1.48 ± 0.15	1.51 ± 0.14	1.47 ± 0.16	1.46 ± 0.15
Lean body mass, kg	32.3 ± 0.3	32.9 ± 0.4	36.6 ± 0.3	36.4 ± 0.4	35.0 ± 0.3
Body mass index	17.3 ± 1.8†	18.4 ± 2.5†	20.1 ± 2.3	20.0 ± 2.4	20.2 ± 2.0

The Relationship between Genotype and Exercise Tolerance in Children with Cystic Fibrosis

HIRAN C. SELVADURAI, KAREN O. McKAY, CAMERON J. BLIMKIE, PETER J. COOPER, CRAIG M. MELLIS, and PETER P. VAN ASPEREN

Am J Respir Crit Care Med Vol 165. pp 762–765, 2002

TABLE 2. AEROBIC PERFORMANCE, ANAEROBIC PERFORMANCE, AND FEV_1 ACCORDING TO CLASS OF SECOND CFTR MUTATION IN PATIENTS WITH AT LEAST ONE COPY OF THE ΔF508 CFTR MUTATION*

	I	II	III	IV	V
Aerobic capacity, ml/kg/min	29.8 ± 4.2†	32.1 ± 4.9†	44.3 ± 6.4‡	54.0 ± 7.2§	54.3 ± 6.8§
FEV_1, % predicted	52.0 ± 5.4	54.2 ± 6.1	53.0 ± 6.0	57.4 ± 6.3	54.7 ± 5.0
Peak anaerobic power, watts/kg	9.6 ± 1.4†	9.5 ± 1.4†	10.6 ± 1.8‡	11.5 ± 1.7§	11.5 ± 1.5§
Shwachman score	55.0 ± 7.0†	52.0 ± 7.3†	66.0 ± 6.5	64.0 ± 6.0	61 ± 5.9

⇨Mutations de classe I et II: moindre IMC, score de Shwachman ET capacité aérobie et anaérobie

Non-Invasive Assessment of Exercise Performance in Children With Cystic Fibrosis (CF) and Non-Cystic Fibrosis Bronchiectasis: Is There a CF Specific Muscle Defect?

Mark Rosenthal, MD, FRCP, FRCPCH,[1] **Indra Narang**, MD, MRCPCH,[1†,§] **Liz Edwards**, MD, MRCPCH,[1†,¶] **and Andrew Bush**, MD, FRCP[2‡]

	Control	CF bronchiectasis	Non-CF bronchiectasis
Male/female	54/50	7/11	11/7
Age (median, range)	12.75 (7.3, 17.7)	12.95 (10.7, 16.9)	13.25 (10.6, 17.1)
Percent predicted FEV_1 median (range)		76 (40–98)	69 (35–90)
Height (cm)	1.54 (1.52–1.57)	1.54 (1.49–1.59)	1.58 (1.52–1.64)
Weight (kg)	47.5 (45.2–49.8)	46.6 (41.2–52.1)	47.7 (42.4–52.9)
Body mass index (wt/ht^2)	19.6 (19.0–20.2)	19.4 (18.0–20.7)	18.9 (17.6–20.2)
Body mass index *z* score	0.45 (0.24–0.65)	0.12 (−0.31–0.56)	0.03 (−0.42–0.48)
Surface area (m^2)	1.42 (1.38–1.47)	1.42 (1.32–1.51)	1.42 (1.31–1.52)
Mean triceps and subscapular skinfold *z* score	0.72 (0.56–0.88)	−1.50 (−1.78 to −1.23)****	−1.13 (−1.38 to −0.88)****
Percent body fat estimated from skinfold thickness	26.4 (24.7–28.0)	16.7 (15.3–18.2)****	15.9 (14.0–17.8)****
Estimated lean body wt (kg)	31.4 (30.1–32.7)	38.8 (934.3–43.4)	39.9 (35.8–44.1)

Sur la spectrométrie respiratoire de masse (CRF, débit pulm. effectif, et VO2), aucune différence dans tous les paramètres évaluant la capacité d'exercice entre CF et non-CF

Pediatric Pulmonology 44:222–230 (2009)

Déterminants de la performance des muscles respiratoires

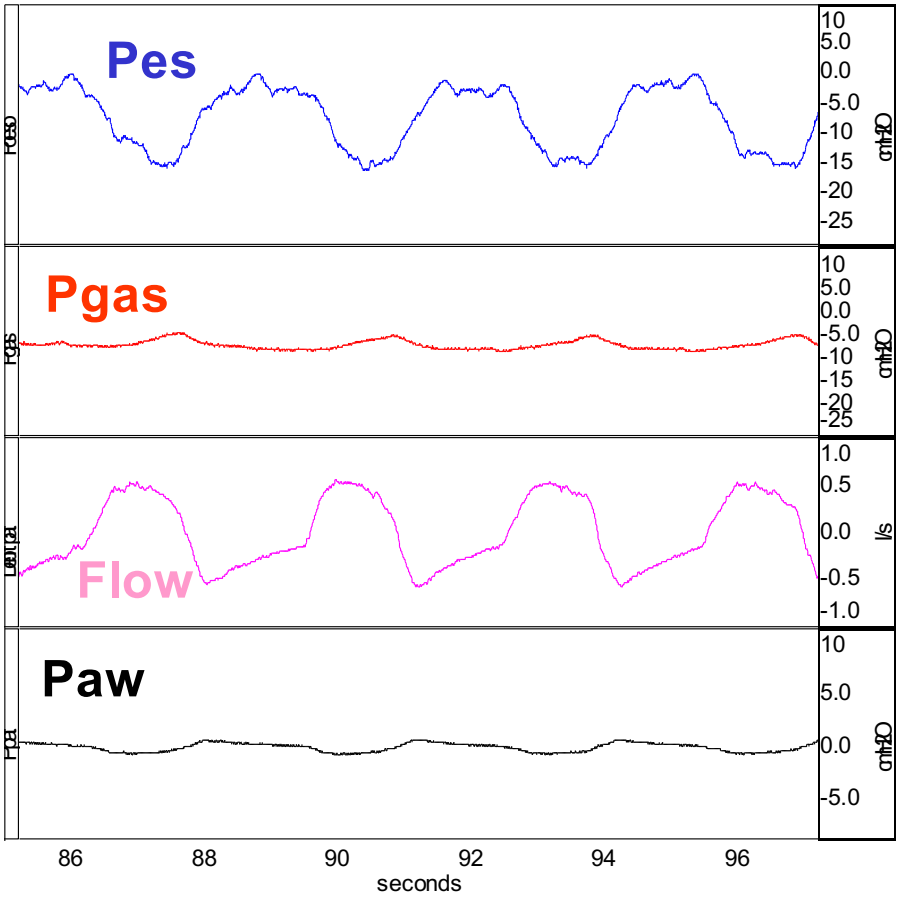
- Fonction respiratoire: OUI
- Statut nutritionnel: OUI
- Inflammation / infection: NON
- CFTR : NON

Explorer les muscles respiratoires

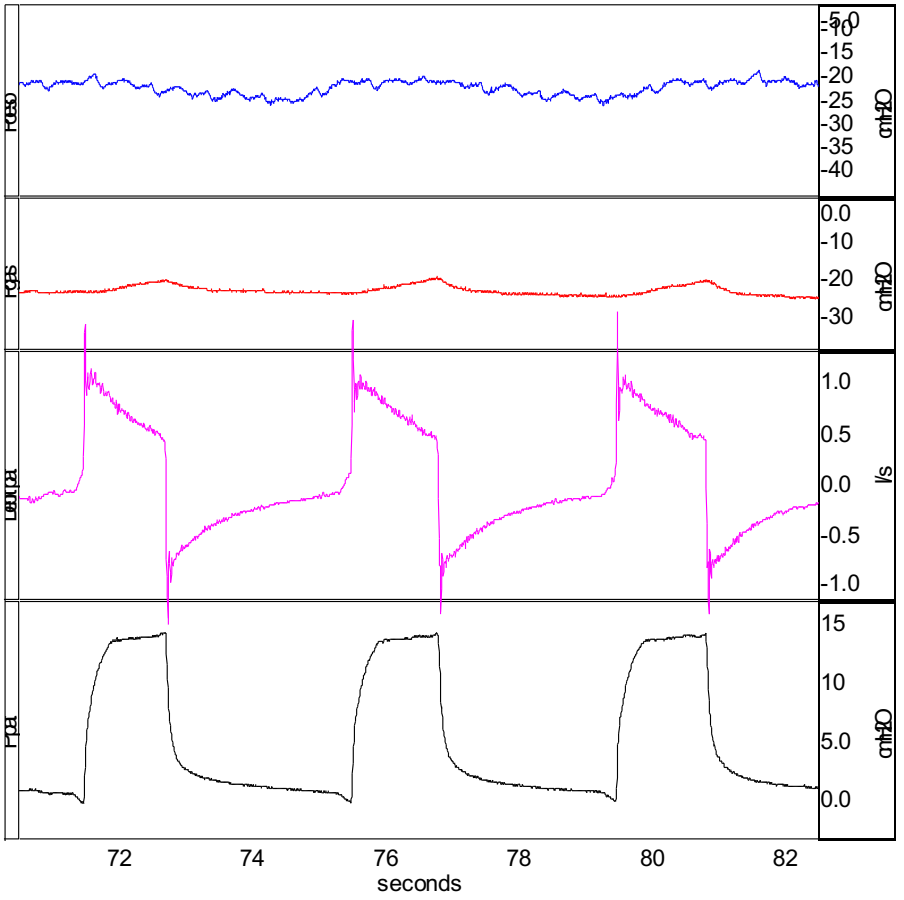
- Quelles explorations ?
- Quelle est la performance des muscle respiratoires dans la CF ?
- Quels sont les déterminants de la performance des muscles respiratoires ?
- En pratique…

La « décharge » des muscles respiratoires, est-elle efficace ?

Respiration spontanée

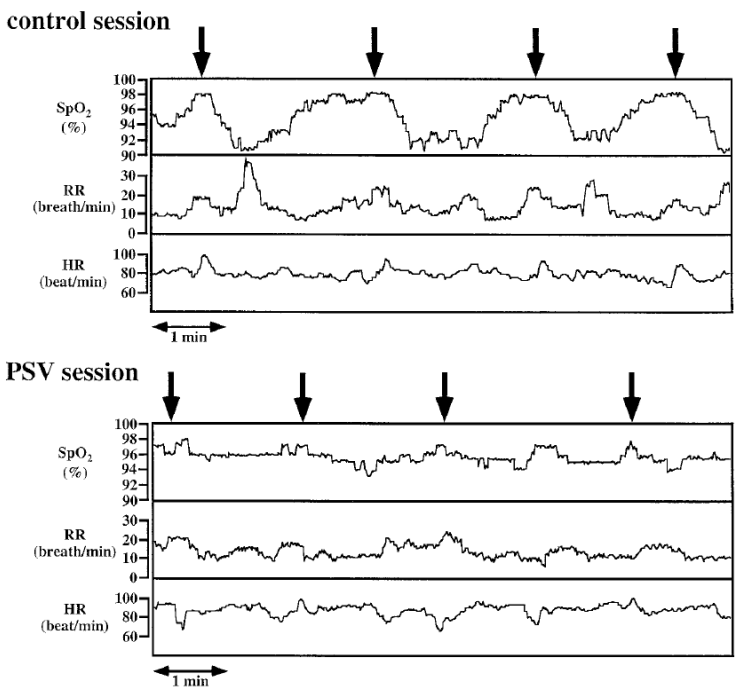


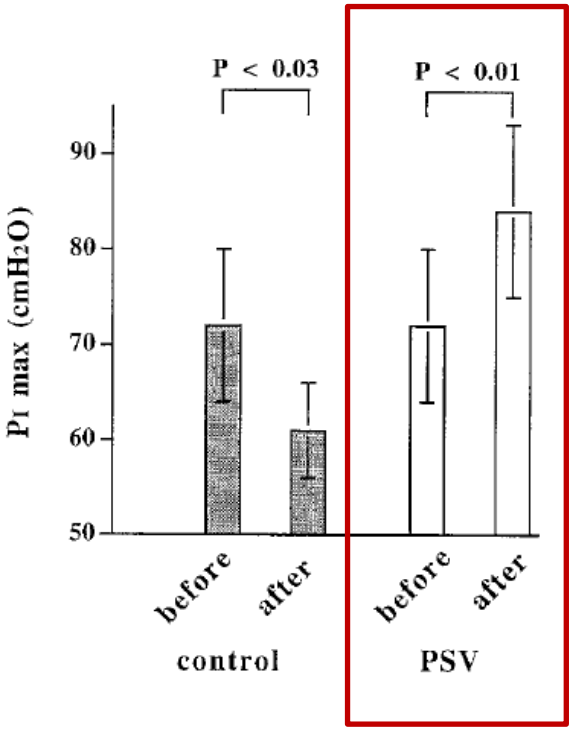
Aide inspiratoire 14 cmH_2O

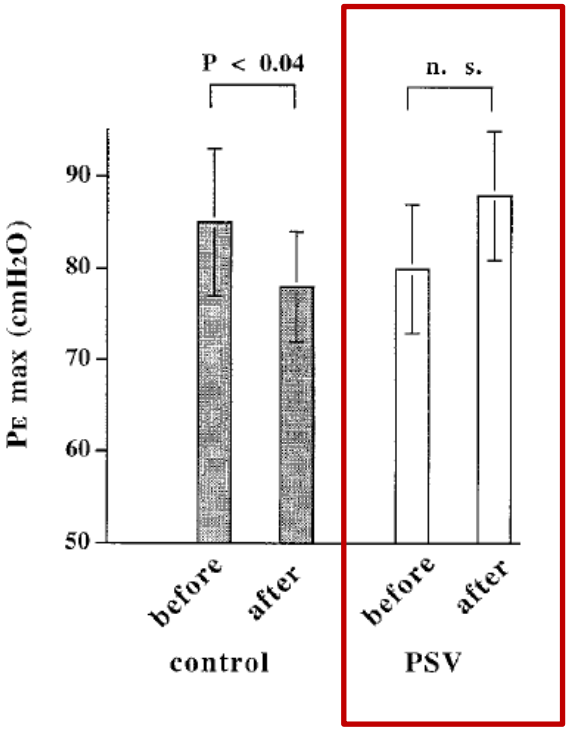


Chest Physiotherapy in Cystic Fibrosis: Improved Tolerance With Nasal Pressure Support Ventilation

Brigitte Fauroux, MD*; Michèle Boulé, MD, PhD‡; Frédéric Lofaso, MD, PhD§; Françoise Zérah, MD§; Annick Clément, MD, PhD*; Alain Harf, MD, PhD§; and Daniel Isabey, PhD§



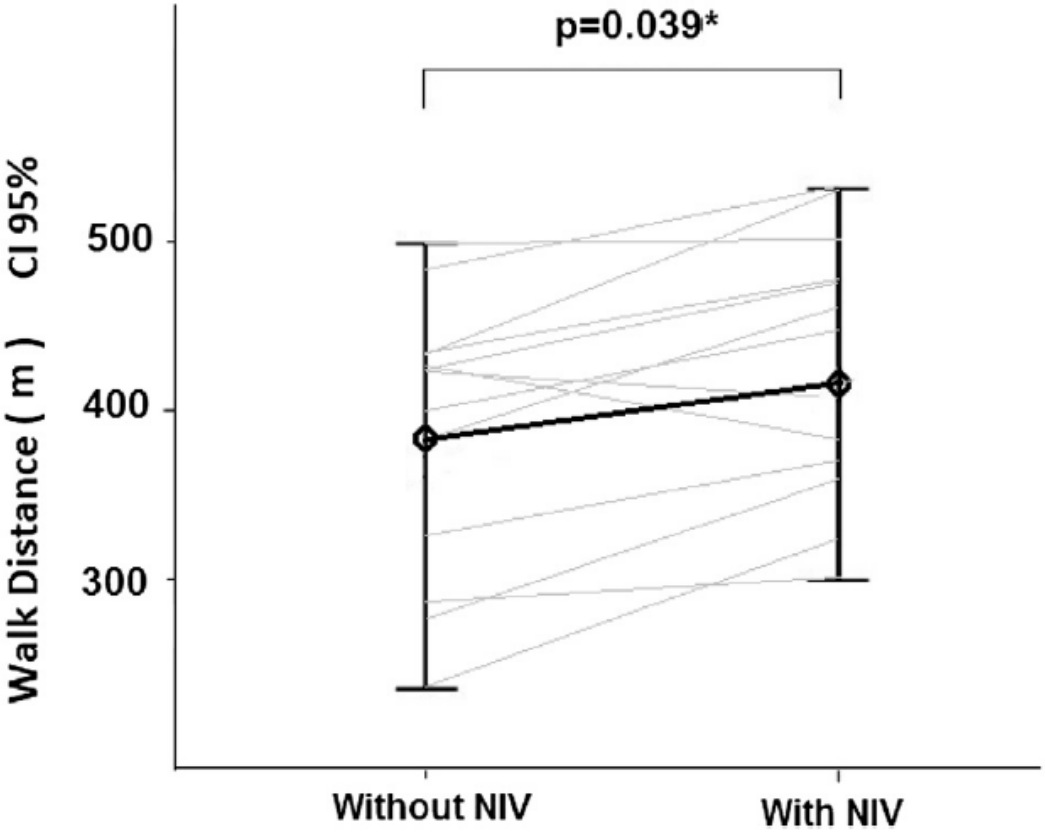


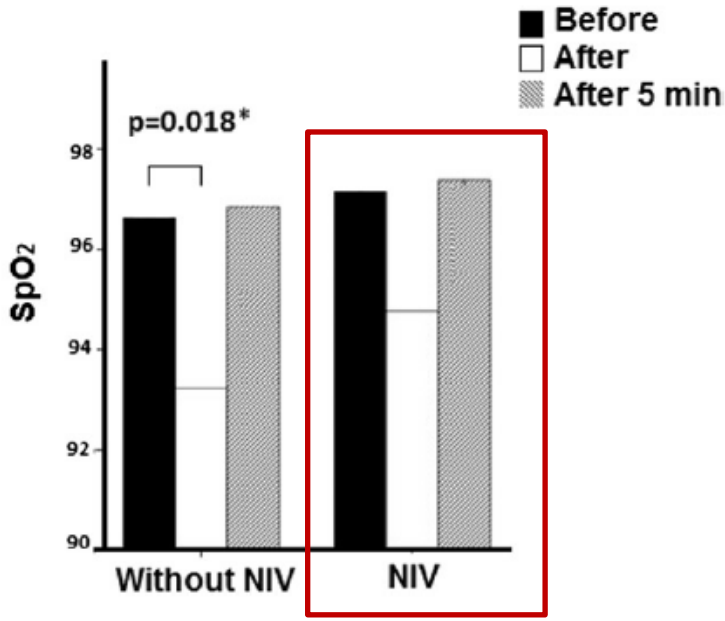


PEDIATRICS Vol. 103 No. 3 March 1999

Effects of noninvasive ventilation on treadmill 6-min walk distance and regional chest wall volumes in cystic fibrosis: Randomized controlled trial

Cibelle Andrade Lima [a],
Armèle de Fátima Dornelas de Andrade [b,*],
Shirley Lima Campos [b], Daniella Cunha Brandão [b],
Guilherme Fregonezi [c], Ianny Pereira Mourato [b],
Andrea Aliverti [d], Murilo Carlos Amorim de Britto [e]

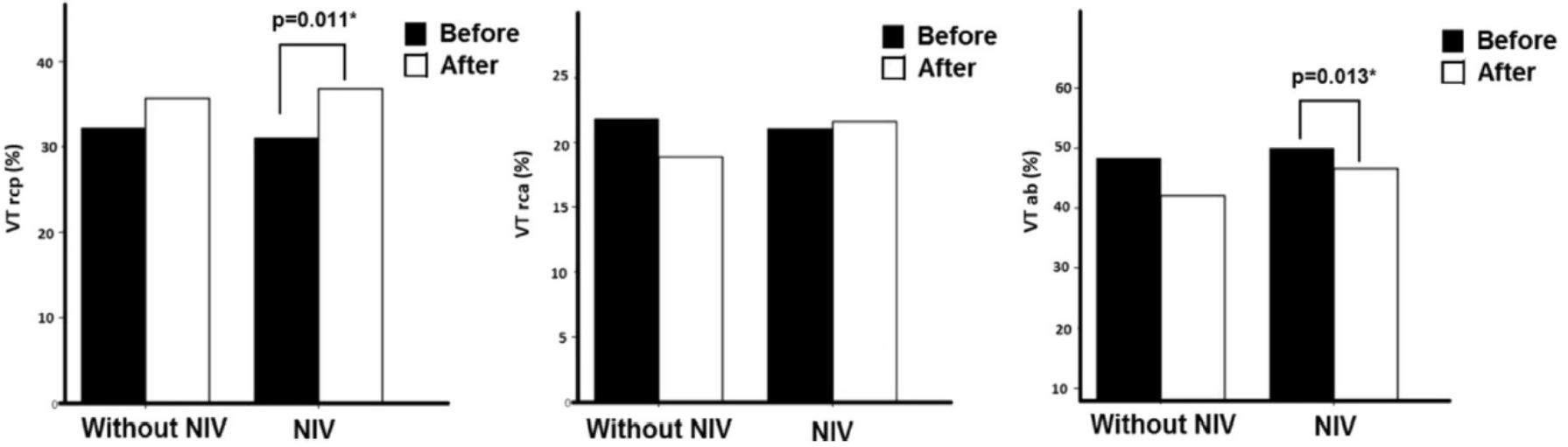




Respiratory Medicine (2014) **108**, 1460–1468

Effects of noninvasive ventilation on treadmill 6-min walk distance and regional chest wall volumes in cystic fibrosis: Randomized controlled trial

Cibelle Andrade Lima [a],
Armèle de Fátima Dornelas de Andrade [b,*],
Shirley Lima Campos [b], Daniella Cunha Brandão [b],
Guilherme Fregonezi [c], Ianny Pereira Mourato [b],
Andrea Aliverti [d], Murilo Carlos Amorim de Britto [e]



Respiratory Medicine (2014) **108**, 1460–1468

Inspiratory muscle training for cystic fibrosis (Review)

Houston BW, Mills N, Solis-Moya A

Main results

Fourteen studies were identified. Of these eight studies with 180 participants met the review inclusion criteria.

Authors' conclusions

We have not found any evidence to suggest that this treatment is either beneficial or not.

Explorer les muscles respiratoires dans la mucoviscidose

- Physiopathologie
  - augmentation de la charge imposée
  - diminution des performances > seuil
  - ➢« décharger » les muscles respiratoires
- Déterminants principaux
  - fonction respiratoire: distension, résistances
  - statut nutritionnel
- Techniques d'exploration: IRM, OEP