ΒΑΣΙΚΕς ΑΡΧΕς ΤΩΝ ΑΝΑΠΝΕΥΣΤΙΚΩΝ ΛΕΙΤΟΥΡΓΙΩΝ: ΣΠΙΡΟΜΕΤΡΗΣΗ ΚΑΙ ΠΝΕΥΜΟΝΙΚΟΙ ΟΓΚΟΙ

ΜΠΙΜΠΑΚΗ ΕΛΕΝΗ-ΠΝΕΥΜΟΝΟΛΟΓΟΣ

ΕΠΙΜΕΛΗΤΡΙΑ Β ΠΑΓΝΗ

Interstitial Lung Disease and Other Pulmonary Manifestations in Connective Tissue Diseases

Isabel Mira-Avendano, MD; Andy Abril, MD; Charles D. Burger, MD; Paul F. Dellaripa, MD; Aryeh Fischer, MD; Michael B. Gotway, MD; Augustine S. Lee, MD; Joyce S. Lee, MD; Eric L. Matteson, MD, MPH; Eunhee S. Yi, MD; and Jay H. Ryu, MD

TABLE 1. Relative Frequencies of Computed Tomography Imaging Patterns Among CTDs[a]

CTD	UIP	NSIP	OP	LIP	DAD	Hemorrhage	Airway[b]	Nodules[c]	Serositis[d]
RA	+++	++	++	+	+	–	+++	+++	+++
SSc	+	+++	+	–	+	–	–	–	–
PM/DM	+	+++	+++	–	++	–	–	–	–
SjS	+	++	–	++	+	–	+	+	–
SLE	+	++	+	++	++	+++	–	–	+++
MCTD	+	++	+	–	–	–	–	–	+

[a]– = absence of finding; + = lowest and +++ = highest; CTD = connective tissue disease; DAD = diffuse alveolar damage pattern; LIP = lymphocytic interstitial pneumonia pattern; MCTD = mixed connective tissue disease; NSIP = nonspecific interstitial pneumonia pattern; OP = organizing pneumonia pattern; PM/DM = polymyositis/dermatomyositis; RA = rheumatoid arthritis; SjS = Sjögren syndrome; SLE = systemic lupus erythematosus; SSc = systemic sclerosis; UIP = usual interstitial pneumonia pattern.

[b]Bronchiectasis, bronchial wall thickening, small centrilobular nodules (that may reflect follicular bronchiolitis), and constrictive bronchiolitis.

[c]Typically ≥1 cm (not centrilobular).

[d]Pleural or pericardial fluid or thickening.

Mayo Foundation for Medical Education and Research ■ Mayo Clin Proc. 2019;94(2):309-325

Connective tissue diseases, multimorbidity and the ageing lung

Paolo Spagnolo[1], Jean-François Cordier[2,3] and Vincent Cottin[2,3,4]

TABLE 1 Types of lung involvement in connective tissue diseases

Primary manifestations
- Pleural involvement
  - Pleurisy, effusion/thickening
- Airway disease
  - Cricoarytenoid and tracheal involvement
  - Bronchiectasis, bronchiolitis
- Vascular involvement
  - Pulmonary hypertension
  - Vasculitis
- Parenchymal lung disease
  - Interstitial lung disease
  - Diffuse alveolar haemorrhage
  - Acute pneumonitis
  - Rheumatoid nodules

Secondary manifestations
- Infection
- Drug toxicity
- Malignancy
- Thromboembolism

Eur Respir J 2016; 47: 1535–1558

Ciancio et al. Multidisciplinary Respiratory Medicine (2019) 14:17
https://doi.org/10.1186/s40248-019-0179-2

Multidisciplinary Respiratory Medicine

REVIEW Open Access

Contribution of pulmonary function tests (PFTs) to the diagnosis and follow up of connective tissue diseases

Nicola Ciancio[1,2*], Mauro Pavone[1], Sebastiano Emanuele Torrisi[1], Ada Vancheri[1], Domenico Sambataro[3], Stefano Palmucci[4], Carlo Vancheri[1], Fabiano Di Marco[5] and Gianluca Sambataro[1,3]

Table 1 Risk factors for Chronic Pulmonary Disease in CTDs

DISEASE	ILD	OLD	PAH
RA	Older age; male sex, smoking history; ACPA positivity, LDH, longstanding or persistently active disease; rheumatoid nodules, articular erosions, genetic predisposition (HLA linked)	Smoking, older age	
SSDs	Scl70, Th/To positivity, dSSc, smoking, older age, rapidly progressive disease.		ACA, LAC, and anti RNA polymerase I, II, III, AECA positivity, older age, RP, NVC positivity; telangiectasias, seric NT-pro-BNP and urate, RAD, RAA, TV, PE
IIMs	Cutaneous manifestations; telangiectasias, RP, ATSA positivity (rapidly progressive for non-Jo1 positivity); older age, acute/subacute onset, CADM, Hamann Rich presentation correlated with worse prognosis		Cutaneous manifestations, peripheral microangiopathy, positivity for SSA/Ro, severe ILD, polyarthralgia, longstanding disease
SjS	Hypergammaglobulinemia, lymphopenia, RF, SSA/Ro, SSB/La positivity	Sicca syndrome, smoking	RP, PE, hepatic injury, RF positivity
SLE			APLA, cardiac or vascular dysfunction, ILD, SLS

Clinical features of asthma with connective tissue diseases

Keisuke Watanabe | Nobuyuki Horita | Yu Hara | Nobuaki Kobayashi | Takeshi Kaneko

Clin Respir J.2023;17:303–310.wileyonlinelibrary.com/journal

TABLE 1 Main underlying CTD ($n = 42$).

	n (%)
Rheumatoid arthritis	23 (54.8)
Systemic lupus erythematosus	6 (14.3)
Sjögren syndrome	3 (7.1)
Systemic sclerosis	3 (7.1)
Dermatomyositis/polymyositis	2 (4.8)
Others	5 (11.9)

TABLE 3 Laboratory findings of asthma with or without CTDs.

	Asthma with CTDs ($n = 42$)	Asthma without CTDs ($n = 526$)	*p*
Eosinophils in peripheral blood	$n = 40$	$n = 465$	
Eosinophil ratio (%)	2.1 (0.2–16.1)	3.5 (0–33.4)	0.009
Absolute number (cells/μL)	151 (15–890)	219 (0–2347)	0.06
Immunoglobulin E (IU/mL)	43 (1–852) ($n = 24$)	237 (1–81 189) ($n = 260$)	0.002
Lung function	$n = 34$	$n = 329$	
FVC (L)	2.17 (1.13–4.57)	2.84 (0.95–6.48)	<0.001
FVC % predicted (%)	86.7 (54.5–127.7)	99.7 (25.0–161.1)	0.007
FEV1 (L)	1.41 (0.67–2.39)	2.01 (0.48–4.37)	<0.001
FEV1/FVC (%)	70.7 (33.2–92.1)	71.7 (27.6–94.5)	0.58
FEV1% predicted (%)	77.2 (36.8–105.7)	88.4 (21–112.2)	0.02

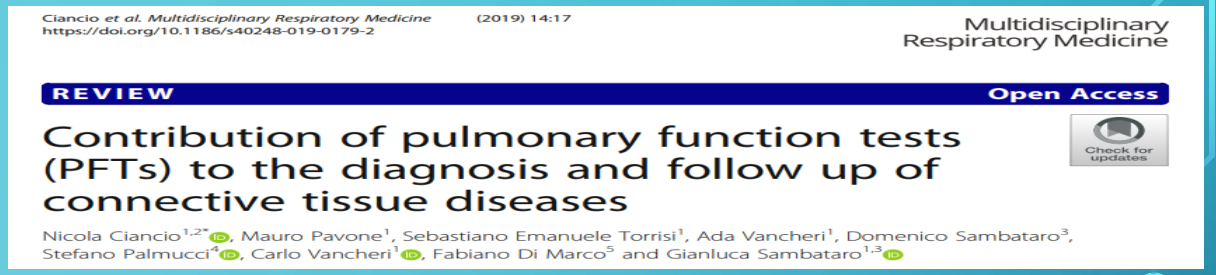
TABLE 5 Factors related to low FEV1 with multivariable analysis.

	Odds ratio (95%CI)	P
Onset at <20 years old	1.8 (1.1–3.2)	0.03
CTDs	2.8 (1.3–6.0)	0.008
COPD	3.8 (2.1–6.7)	<0.001

TABLE 6 Factors related to low-T2 asthma.

	Low-T2 asthma ($n = 53$)	High-T2 asthma ($n = 223$)	*p*
Age (years)	62 (22–85)	57 (21–85)	0.19
Sex (male/female)	12/41	98/125	0.005
Smoking history (never/ex/current)	30/17/6 ($n = 53$)	111/79/28 ($n = 218$)	0.76
Onset at <20 years old	7 (14.0%) ($n = 50$)	53 (25.2%) ($n = 210$)	0.096
Use of systemic steroids other than asthma	17 (32.1)	54 (24.2%)	0.29
CTDs	11 (20.8%)	12 (5.4%)	0.001
COPD	4 (7.7%) ($n = 52$)	33 (15.7%) ($n = 210$)	0.18
AR	13 (24.5%)	62 (27.8%)	0.73
Sinusitis	6 (11.3%)	31 (13.9%)	0.82

IMPORTANCE OF PFTS IN CTDS.



Ciancio et al. Multidisciplinary Respiratory Medicine (2019) 14:17
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Multidisciplinary Respiratory Medicine

REVIEW Open Access

Contribution of pulmonary function tests (PFTs) to the diagnosis and follow up of connective tissue diseases

Nicola Ciancio[1,2*], Mauro Pavone[1], Sebastiano Emanuele Torrisi[1], Ada Vancheri[1], Domenico Sambataro[3], Stefano Palmucci[4], Carlo Vancheri[1], Fabiano Di Marco[5] and Gianluca Sambataro[1,3]

- Lung involvement is often severe and represent the 1st cause of death.
- PFTs are able to highlight lung involvement BEFORE clinical onset.
- Drive follow up.
- Main factor for treatment-related decisions (→antifibrotic drugs).

EUROPEAN RESPIRATORY REVIEW
REVIEW
A. KOURI ET AL.

Exploring the 175-year history of spirometry and the vital lessons it can teach us today

Andrew Kouri[1], Ronald J. Dandurand[2,3,4], Omar S. Usmani[5] and Chung-Wai Chow [6,7]

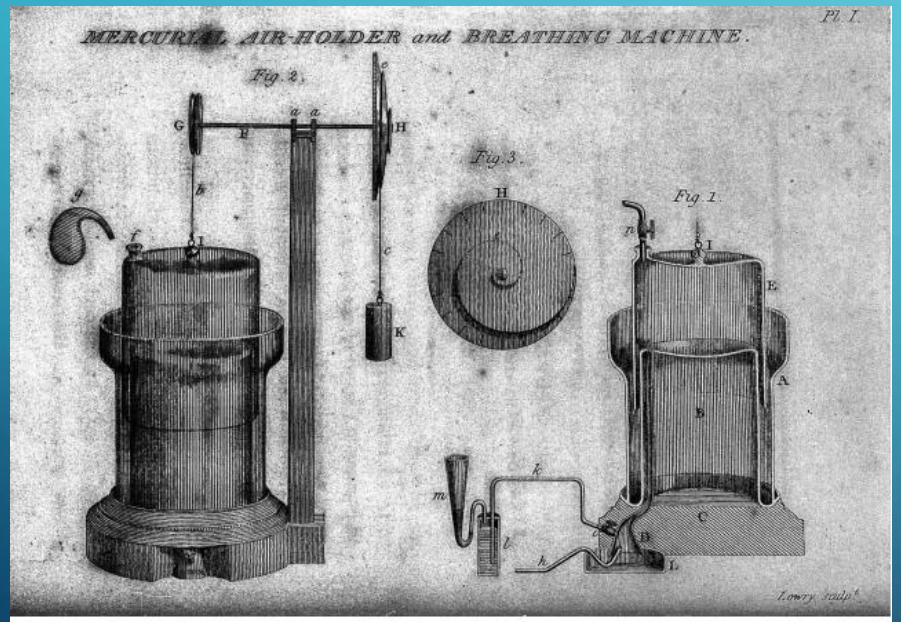


FIGURE 1 Mercurial air holder and breathing machine developed by Humphry Davy and colleagues at the Pneumatic Institution in England, circa 1798, 48 years prior to Hutchinson's spirometer. Reproduced from [3] courtesy of the Wellcome Collection.

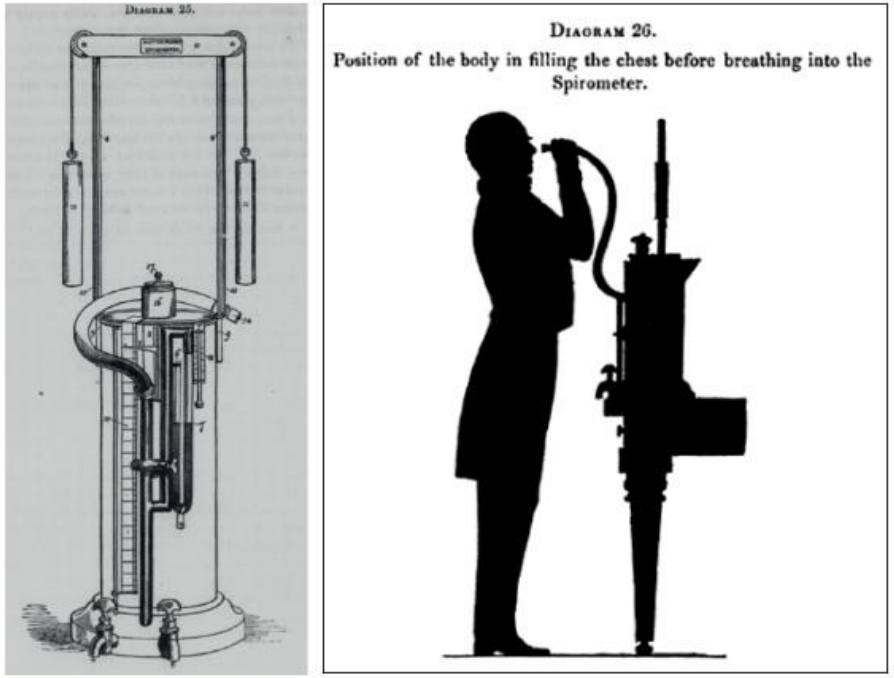


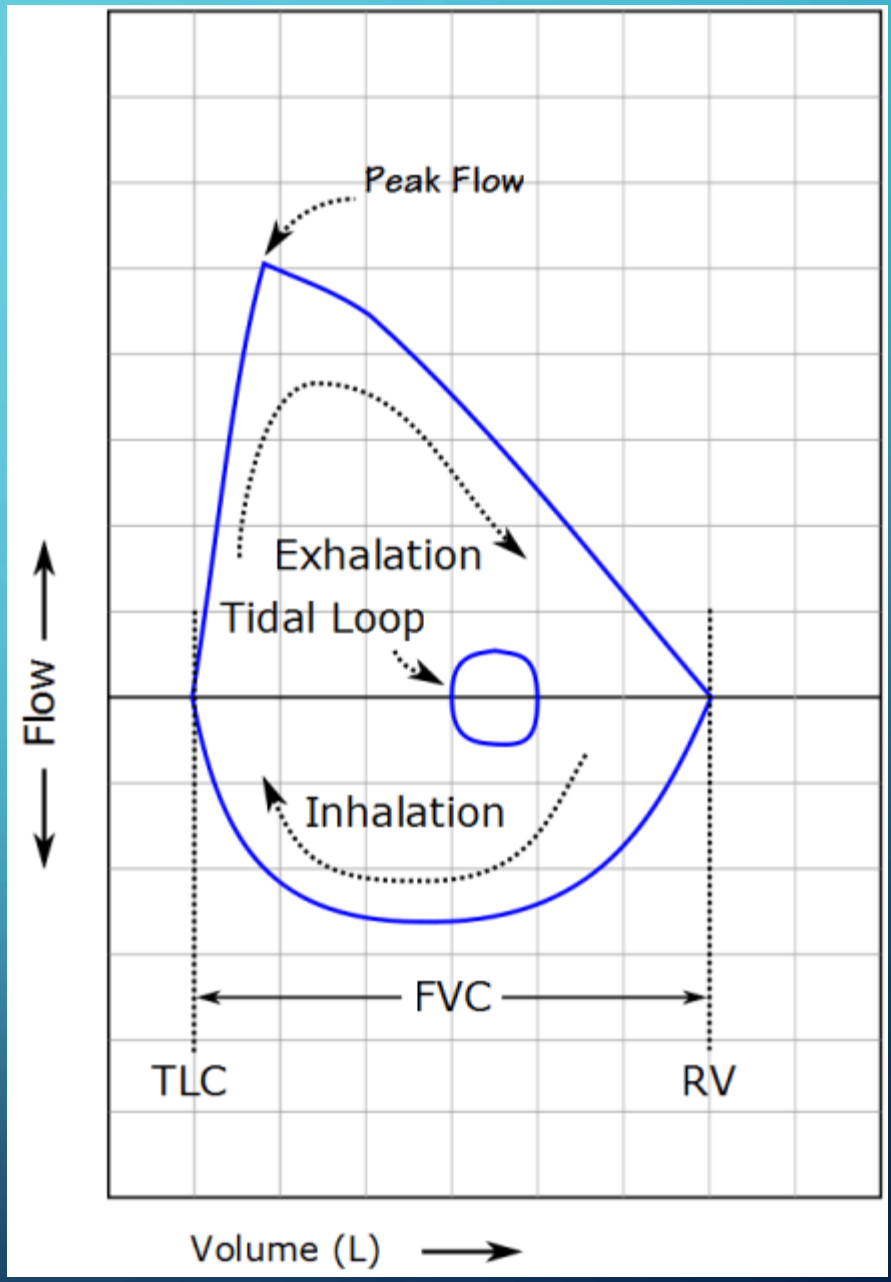
FIGURE 2 Diagrams 25 and 26 from Hutchinson's original publication describing the spirometer. Diagram 25 depicts the internal mechanisms of the spirometer and diagram 26 demonstrates how to position a patient in relation to the device. Reproduced from [7].

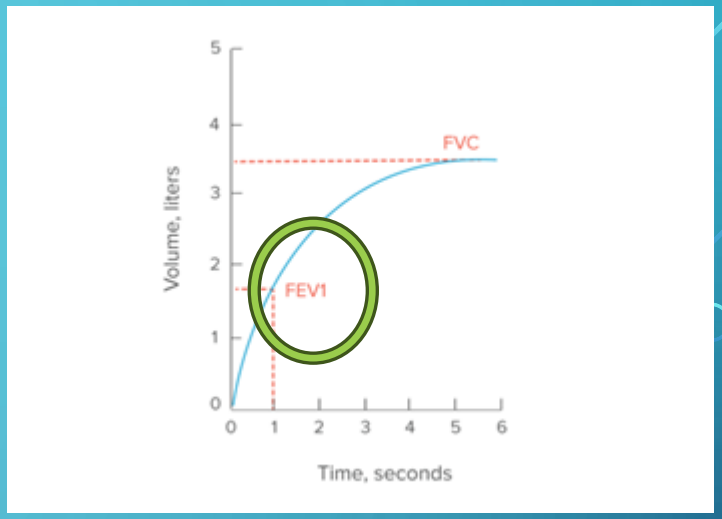
ΣΠΙΡΟΜΕΤΡΗΣΗ

ΣΠΙΡΟΜΕΤΡΗΣΗ
ΣΤΑΤΙΚΟΙ ΟΓΚΟΙ

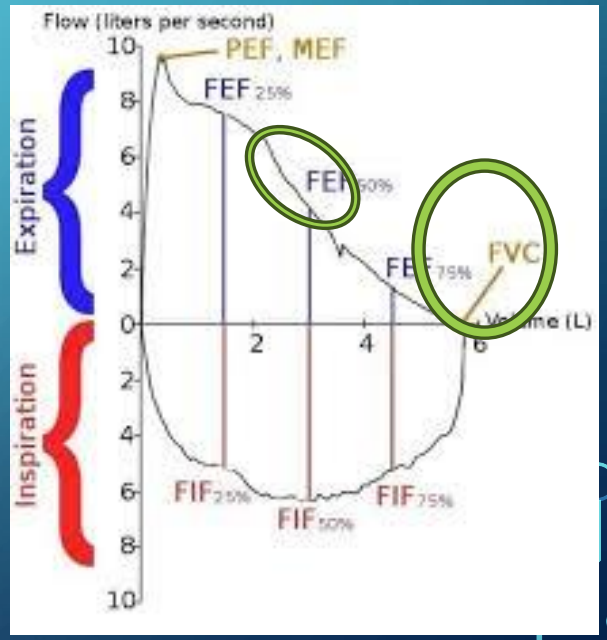
ΔΙΑΧΥΣΗ


Spirometry


Peak Flow
Exhalation
Tidal Loop
Inhalation
Flow
FVC
TLC
RV
Volume (L)



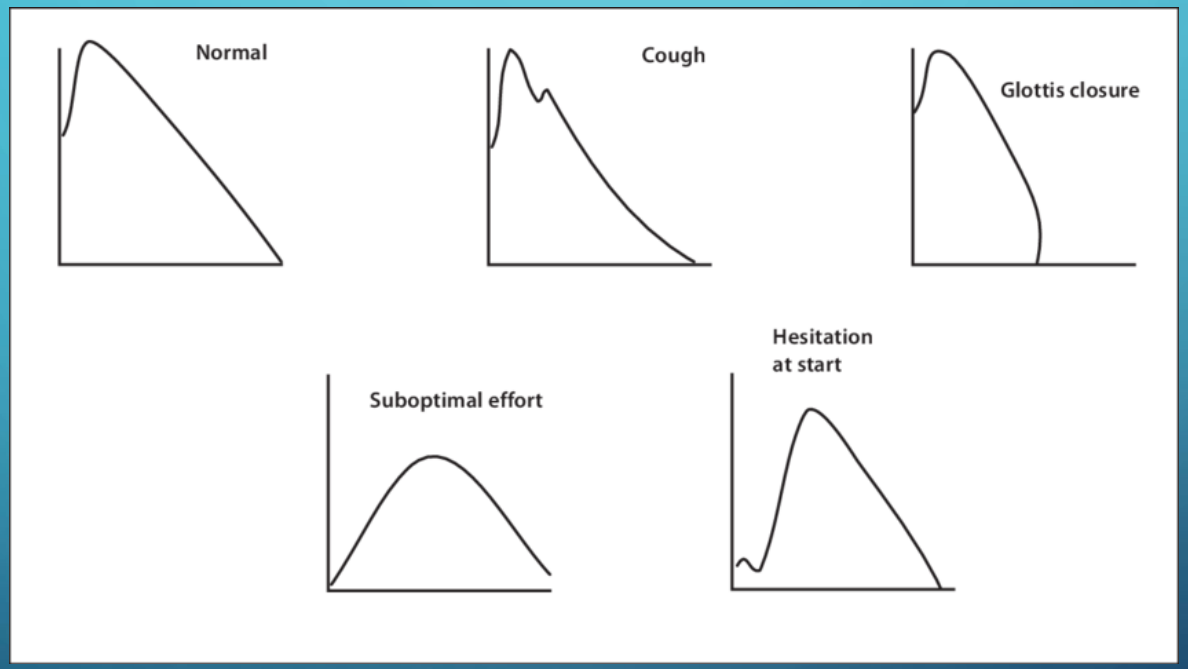
- FEV1 : Forced Expiratory Volume in 1 second.
- FVC : Forced Vital Capacity
- $FEF_{25-75\%}$: Forced Expiratory Flow over the middle 50% of the FVC. (->early airway obstruction/ “small airway disease”)
- FEV1 /FVC.



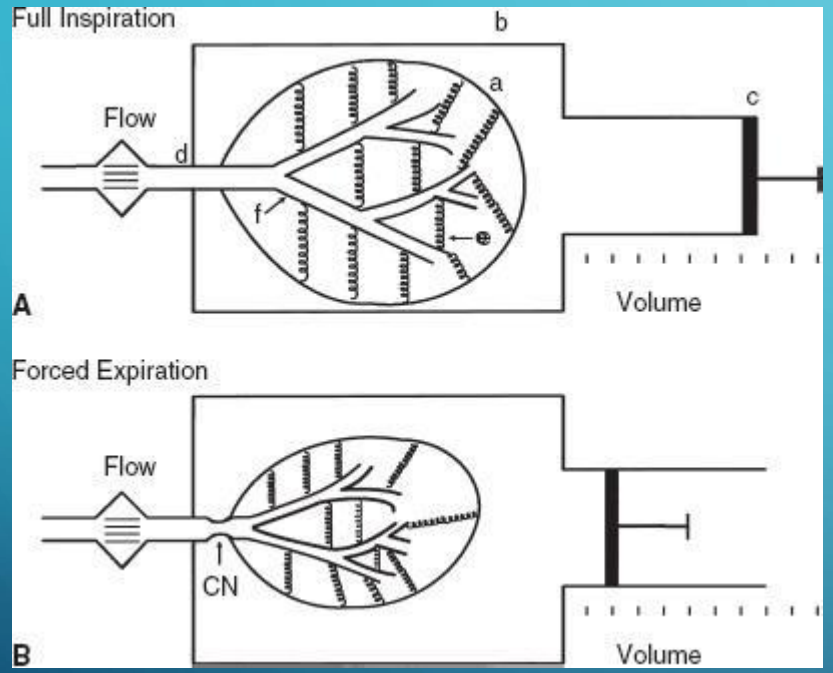
FV representation:

- ✓ Easiest
- ✓ Most informative

EXAMPLES OF UNACCEPTABLE FORCED EXPIRATORY VITAL CAPACITY MANEUVERS



VALUE OF FORCED VITAL CAPACITY (FVC) TEST



- 3 features determine the maximal expiratory flow of the lung at any given lung volume:

➢Lung elasticity

➢Size of the airways

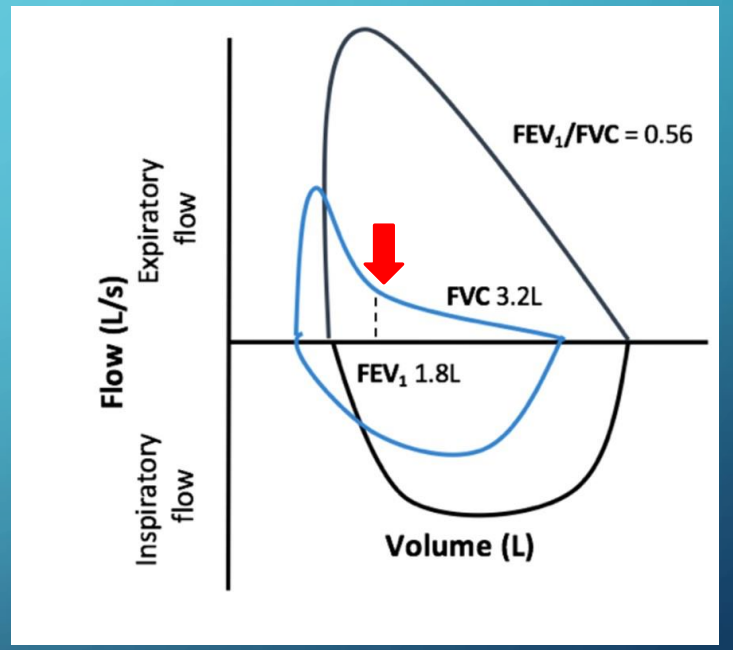
➢Resistance to flow

FVC is very sensitive to diseases that alter the lung's mechanical properties.

FLOW-VOLUME CURVE

1.Obstructive phenotype and emphysema:

- loss of elastic recoil pressure
- Airways are narrowed
- Increased flow resistance
- Decreased maximal expiratory flow

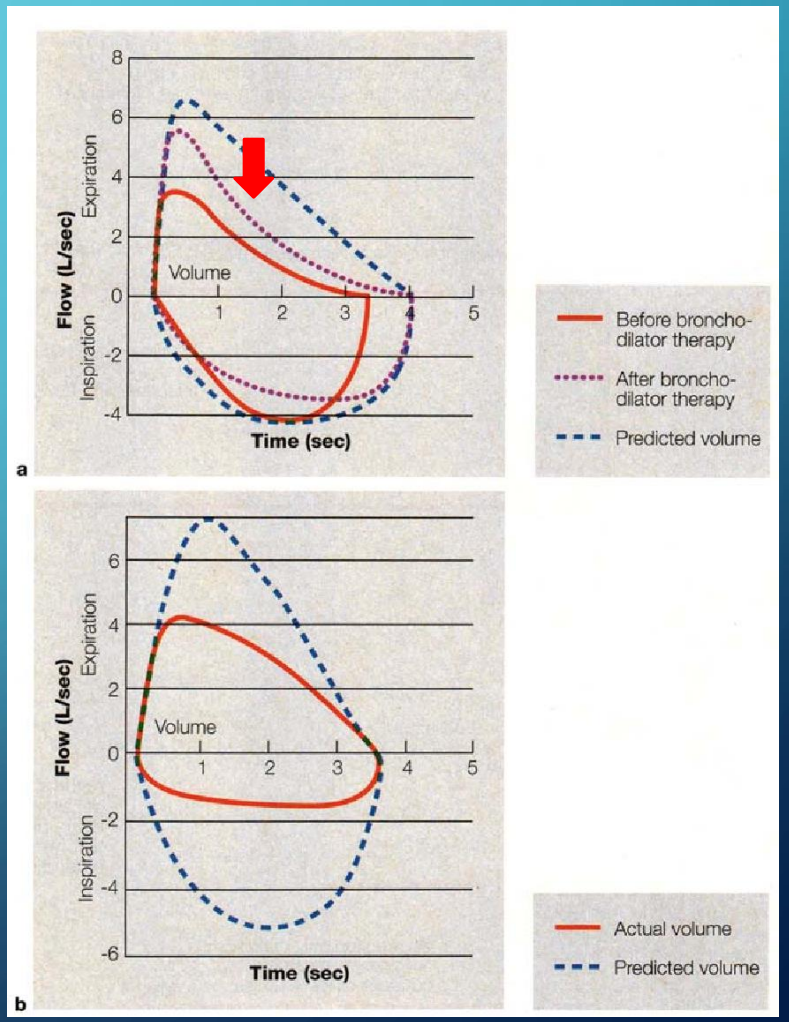


FEV1 ↓↓↓
FVC ↓
FEV1/FVC < 70%

FLOW-VOLUME CURVE

- 2. ASTHMA:
- Airways are narrowed due to bronchoconstriction, mucosal inflammation and edema.
- Increased Resistance
- Decreased maximal flows

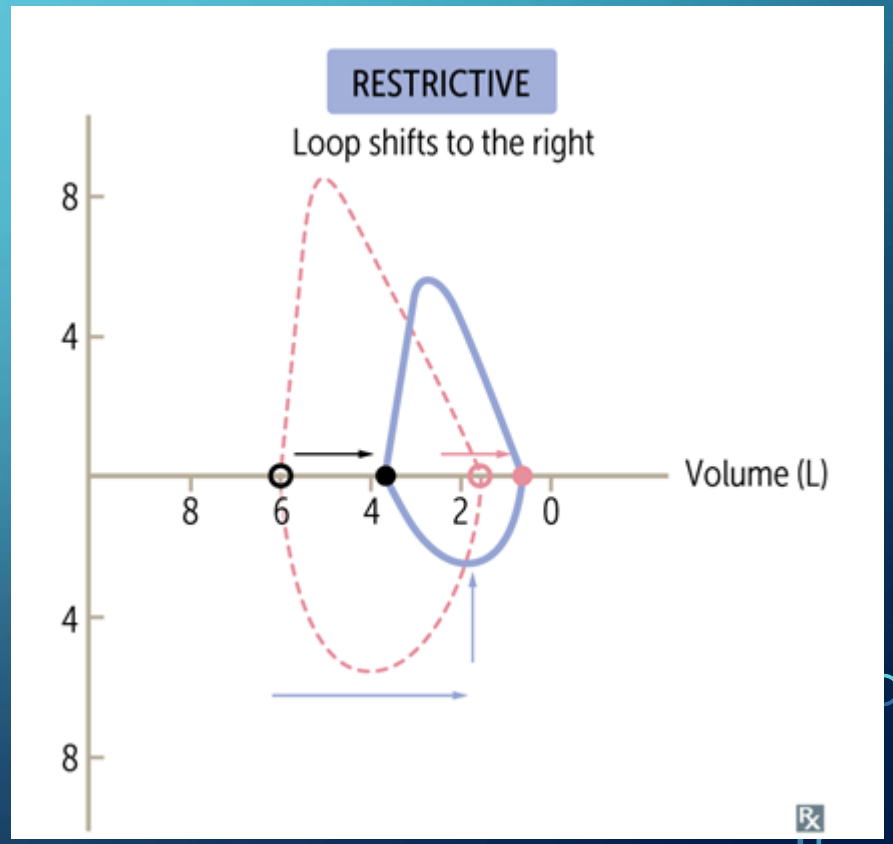
Obstruction + ΔFEV1>12%



FLOW-VOLUME CURVE

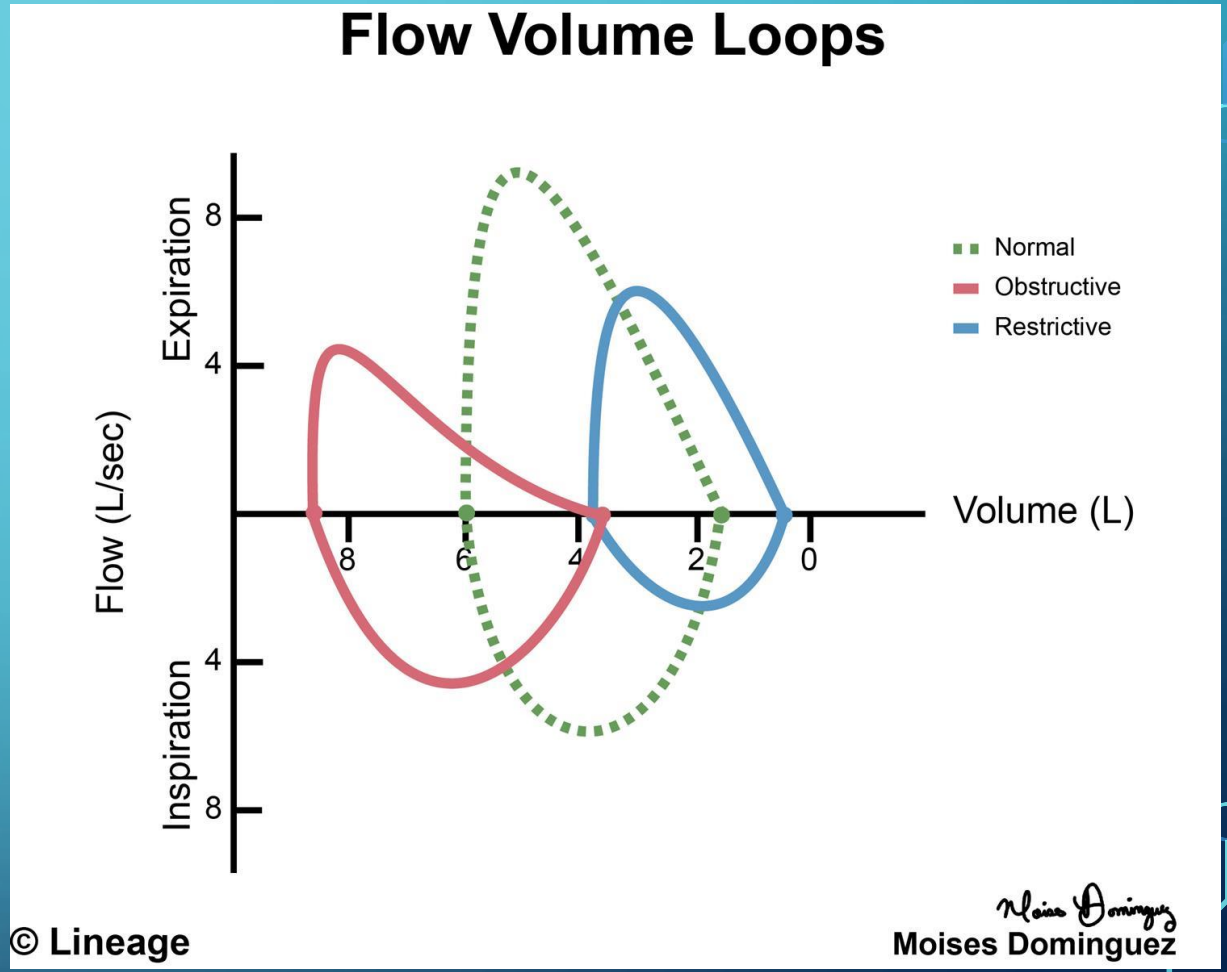
- 3.PULMONARY FIBROSIS:
- Increased elastic recoil pressure
- Airways are distended
- Increased maximum flows
- Reduced lung volumes

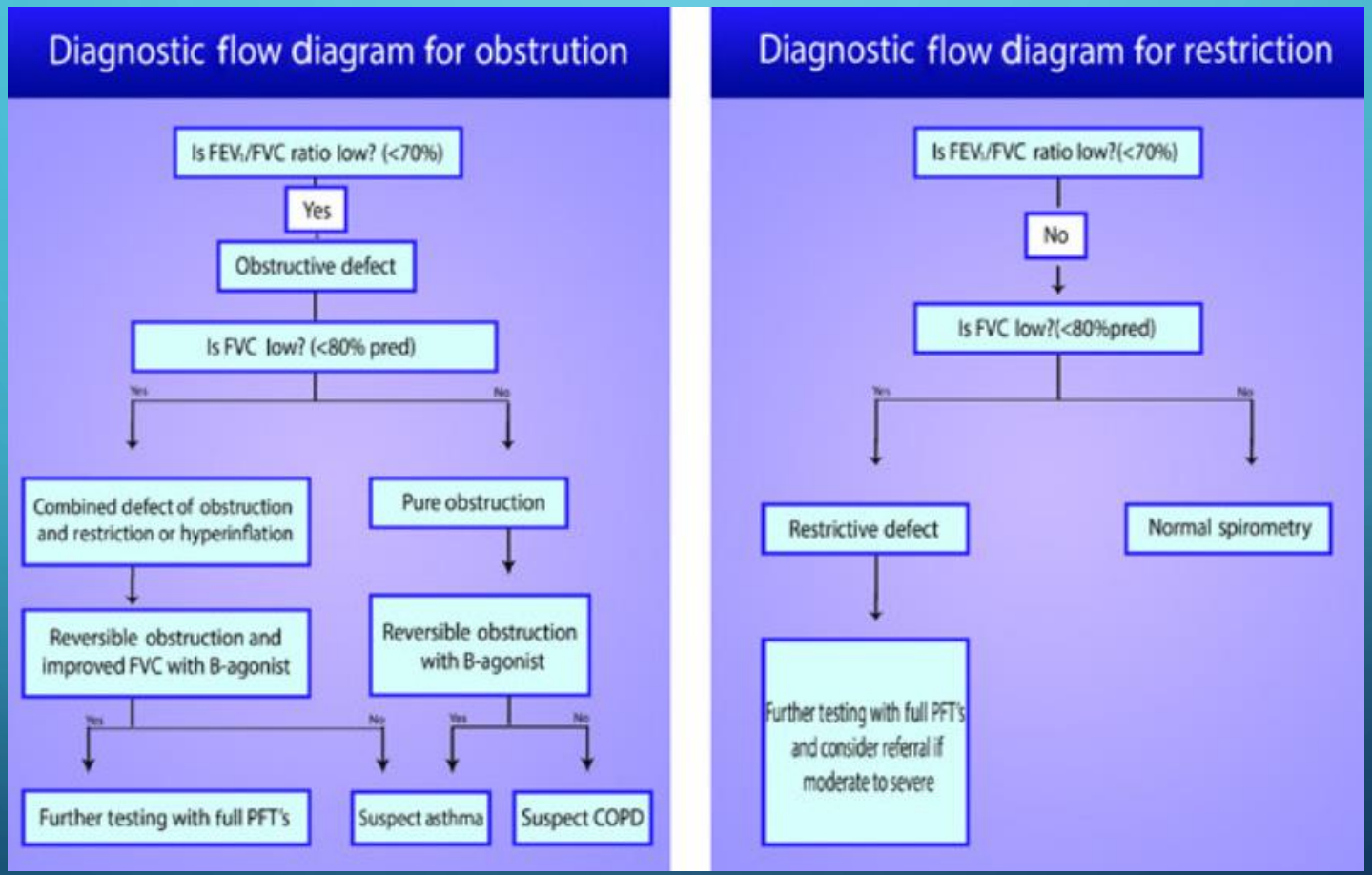
FEV1 ↓↓
FVC ↓↓
FEV1/FVC > 70%



Suggested Pattern Based on FEV_1 and FVC Values

FEV_1/FVC ratio	FVC	Suggested pattern
Normal	Normal	Normal
Normal	Decreased	Restrictive
Decreased	Normal	Obstructive defect
Decreased	Decreased	Mixed





•Xiao Ou et al npj Primary Care Respiratory Medicine 25(1):150

AMERICAN THORACIC SOCIETY DOCUMENTS

Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Luca Richeldi, Carey C. Thomson, Yoshikazu Inoue, Takeshi Johkoh, Michael Kreuter, David A. Lynch, Toby M. Maher, Fernando J. Martinez, Maria Molina-Molina, Jeffrey L. Myers, Andrew G. Nicholson, Christopher J. Ryerson, Mary E. Strek, Lauren K. Troy, Marlies Wijsenbeek, Manoj J. Mammen, Tanzib Hossain, Brittany D. Bissell, Derrick D. Herman, Stephanie M. Hon, Fayez Kheir, Yet H. Khor, Madalina Macrea, Katerina M. Antoniou, Demosthenes Bouros, Ivette Buendia-Roldan, Fabian Caro, Bruno Crestani, Lawrence Ho, Julie Morisset, Amy L. Olson, Anna Podolanczuk, Venerino Poletti, Moisés Selman, Thomas Ewing, Stephen Jones, Shandra L. Knight, Marya Ghazipura, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

This official clinical practice guideline was approved by the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax February 2022

Part II: Diagnosis and Treatment of PPF in Fibrotic ILD, Other than IPF

Definition of PPF

In a patient with ILD of known or unknown etiology other than IPF who has radiological evidence of pulmonary fibrosis, PPF is defined as at least two of the following three criteria occurring within the past year with no alternative explanation (Table 4):

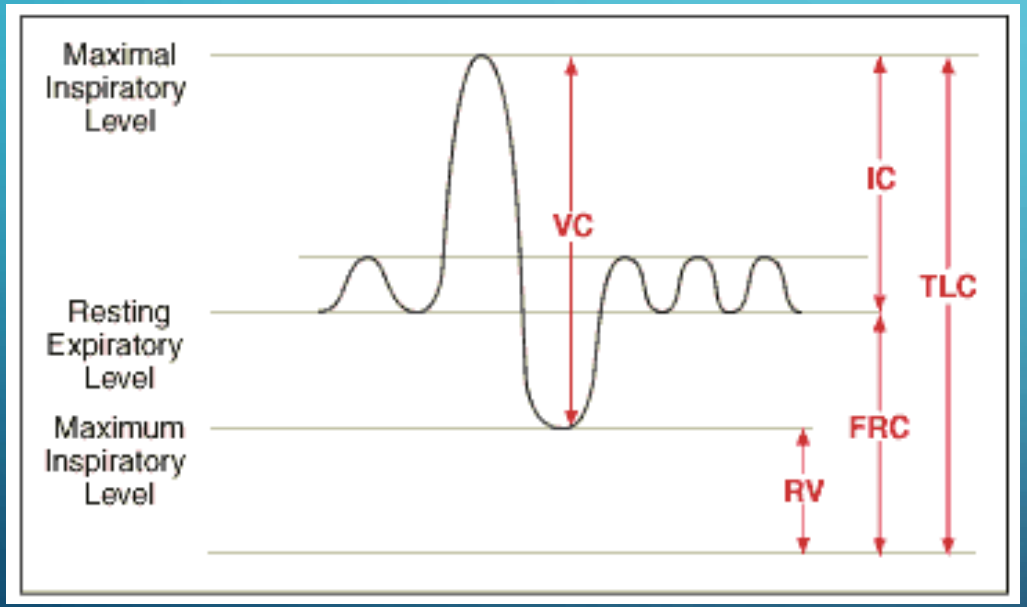
1. worsening respiratory symptoms;
2. physiological evidence of disease progression, as defined below; and
3. radiological evidence of disease progression, as defined below.

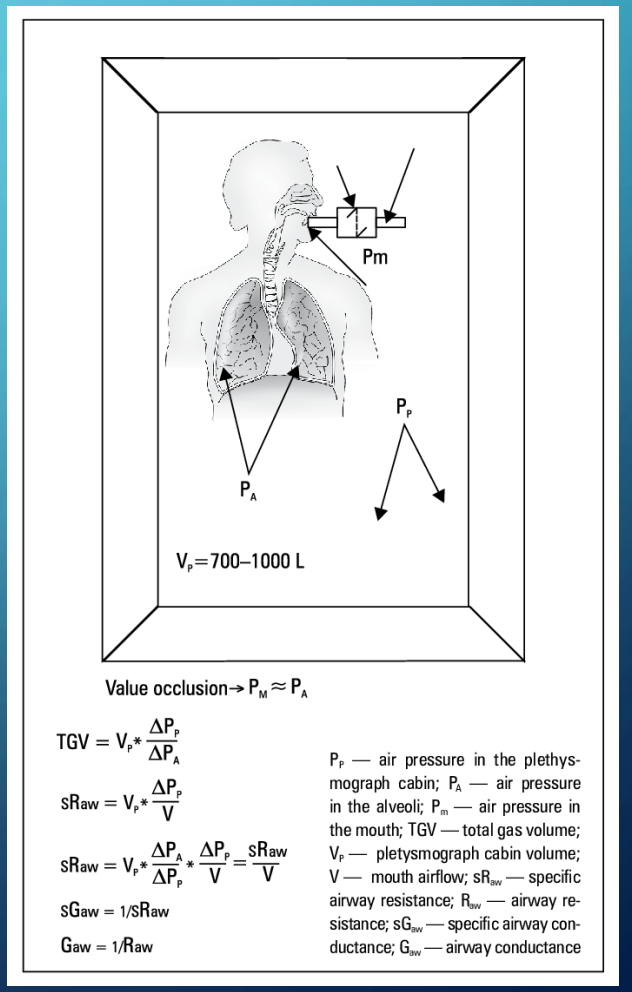
Physiological Criteria for PPF

There is a paucity of published data regarding physiological measurements in patients with PPF. Therefore, the committee derived the physiological criteria for PPF by extrapolation of data from patients with IPF because the disease behavior and prognosis of IPF and PPF are comparable (124). The committee defined physiological evidence of disease progression as the presence of either of the following findings, if the findings are attributable to worsening fibrosis:

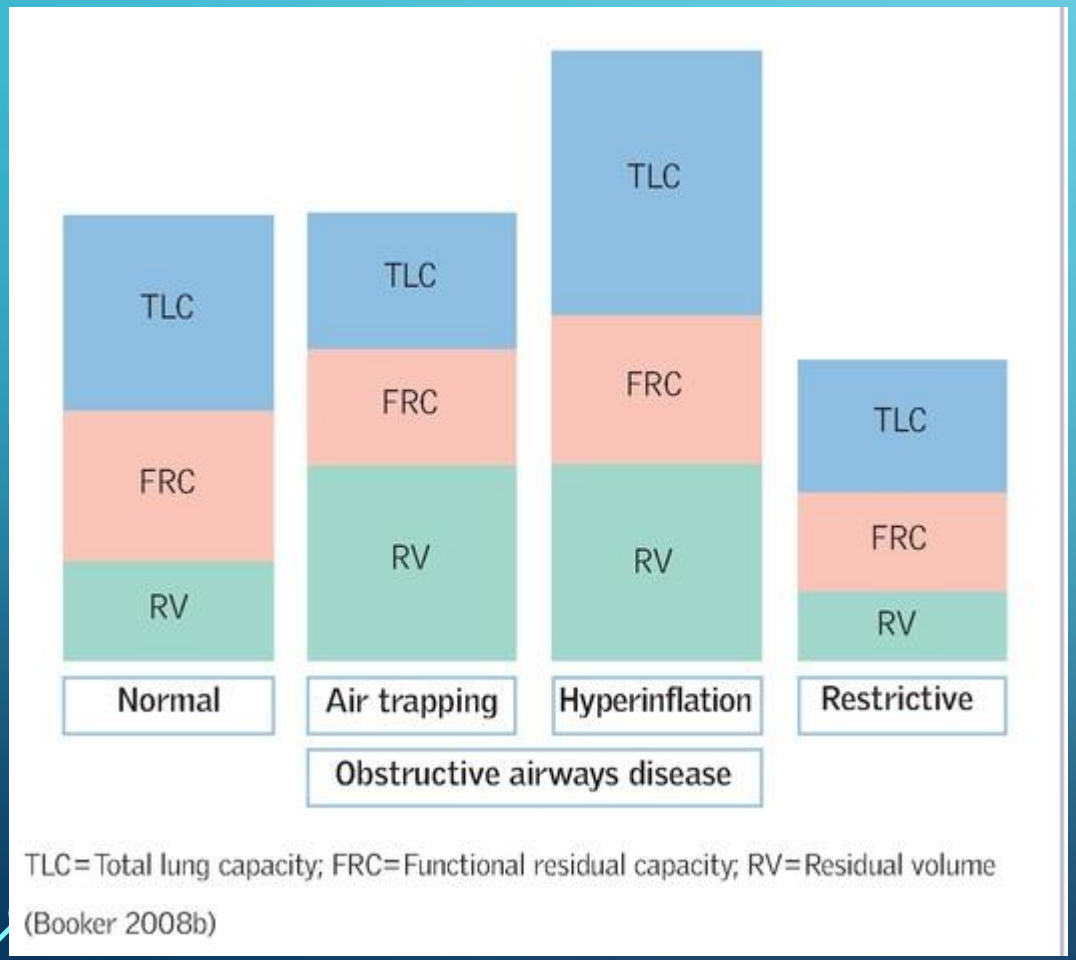
1. Absolute decline in FVC of $\geq$5% within 1 year of follow-up.
2. Absolute decline in D_{LCO} (corrected for Hb) of $\geq$10% within 1 year of follow-up.

STATIC VOLUMES-PLETHYSMOGRAPHY



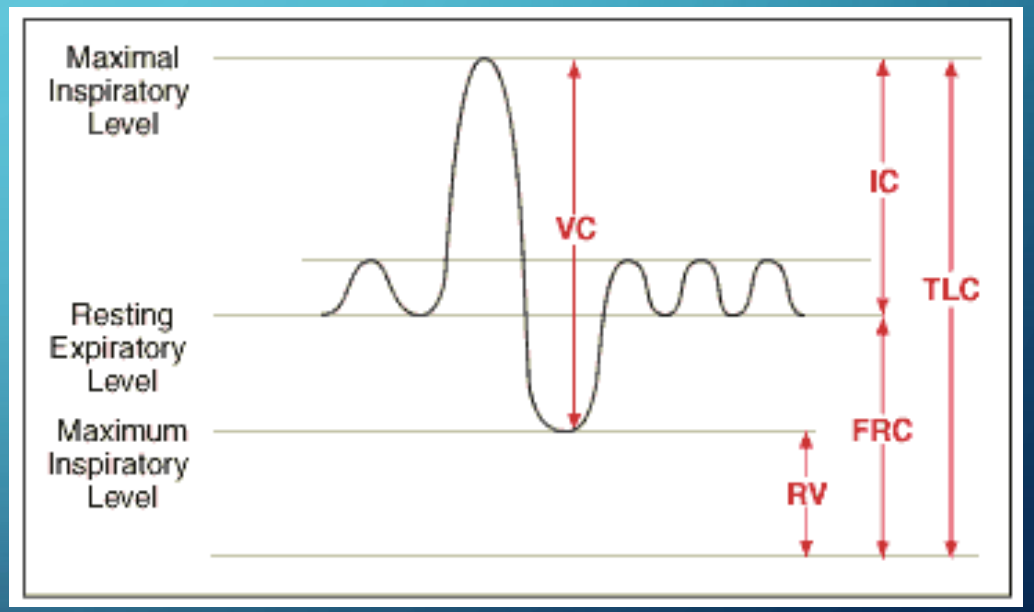


STATIC VOLUMES-PLETHYSMOGRAPHY



TLC=Total lung capacity; FRC=Functional residual capacity; RV=Residual volume

(Booker 2008b)



TAKE HOME MESSAGES

- ✓Οι αεραγωγοί, κεντρικοί και περιφερικοί, προσβάλλονται συχνά σε ασθενείς με νοσήματα του συνδετικού ιστού.
- ✓Κοινοί παράγοντες κινδύνου αυξάνουν επίσης την παρουσία νοσημάτων των αεραγωγών ως συννοσηρότητες.
- ✓Η σπιρομέτρηση αποτελεί βασικό εργαλείο διάγνωσης, παρακολούθησης και θεραπευτικής προσέγγισης και στους ασθενείς με CTDs.
- ✓Η διεπιστημονική προσέγγιση τόσο στην διάγνωση όσο και στην παρακολούθηση αυτών των ασθενών είναι κρίσιμης σημασίας.

Vincent van Gogh

ΕΥΧΑΡΙΣΤΩ ΠΟΛΥ!

ΠΕΡΙΣΤΑΤΙΚΟ 1

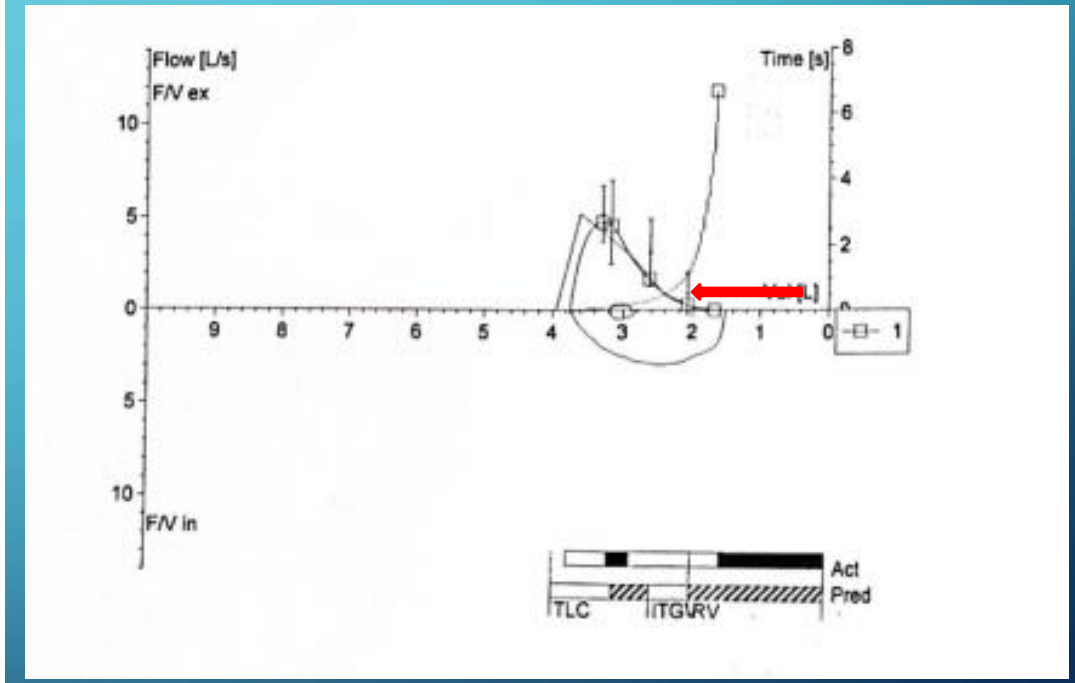
- Γυναίκα 72 ετών με α/α PA από 10 ετίας.
- Παραπονείται για: επεισόδια βήχα και συχνές «λοιμώξεις αναπνευστικού».
- Ποτέ καπνίστρια.
- Γνωστό νόσημα του αναπνευστικού συστήματος: (-)
- Έκθεση σε πτηνά/μούχλα: (-) Επαγγελματική έκθεση (-)
- Φαρμακευτική αγωγή: Tocilizumab, Medrol 8 mg, Ideos,Procoralan,Amlotens,T4,Torvaplus, Lansoprazol, Aprovel.

ΠΑΝΕΠΙΣΤΗΜΙΑΚΟ ΝΟΣΟΚΟΜΕΙΟ ΗΡΑΚΛΕΙΟΥ ΚΡΗΤΗΣ
ΠΝΕΥΜΟΝΟΛΟΓΙΚΗ ΚΛΙΝΙΚΗ
ΕΡΓΑΣΤΗΡΙΟ ΛΕΙΤΟΥΡΓΙΚΩΝ ΕΞΕΤΑΣΕΩΝ ΠΝΕΥΜΟΝΩΝ
ΔΙΕΥΘΥΝΤΗΣ: Ν. ΤΖΑΝΑΚΗΣ
Τηλ.: 2810 392227

Identification:	F08081950		
Last Name:	[redacted]	Age:	72 Years
First Name:	[redacted]	Height:	154,0 cm
Date of Birth:	06/08/1950	Weight:	76,0 kg
		Sex:	female
Smoker:	NO	Physician:	--

		Pred	Actl	%(Actl/Pred)
VC MAX	[L]	2.02	2.22	109.7
VC EX	[L]	2.02	1.08	53.4
VC IN	[L]	2.02	1.40	69.1
IC	[L]	1.59	0.93	58.4
FVC	[L]	2.06	2.08	101.0
FEV 1	[L]	1.68	1.62	96.2
FEV 1 % FVC	[%]		77.79	
FIV1 % VC MAX	[%]		93.69	
MMEF 75/25	[L/s]	2.40	1.08	45.2
FEF 75	[L/s]	0.93	0.30	32.0
FEF 50	[L/s]	3.13	1.69	54.0
FEF 25	[L/s]	4.76	4.59	96.5
PEF	[L/s]	5.20	4.76	91.6
SR eff	[kPa*s]	0.96	2.96	308.0
R eff	[kPa*s/L]	0.30	0.99	330.5
ITGV	[L]	2.52	2.82	112.0
RV	[L]	1.94	1.53	78.9
TLC	[L]	4.37	3.75	85.7
RV % TLC	[%]	43.44	40.80	93.9
ITGV % TLC	[%]	56.62	75.28	133.0
ERV	[L]	0.58	1.29	222.1
VC MAX	[L]	2.02	2.22	109.7
Date			19/06/23	
Time			09:19:07πμ	

		Pred	Actl
FEV6	[L]		2.08
FEV1 % FEV6	[%]		77.79



ΣΠΙΡΟΜΕΤΡΗΣΗ ΠΡΟ ΚΑΙ ΜΕΤΑ ΒΡΟΓΧΟΔΙΑΣΤΟΛΗΣ

		Pred	A1	%(A1/P)	A2	%(A2/A1)
VT	[L]	0.54	0.33	60.3		
BF	[1/min]	20.00	27.88	139.4		
MV	[L/min]	10.86	9.12	84.0		
VC MAX	[L]	2.02	2.22	109.7	2.30	103.6
ERV	[L]	0.58	1.29	222.1		
IC	[L]	1.59	0.93	58.4		
FVC	[L]	2.06	2.08	101.0	2.30	110.6
FEV 1	[L]	1.68	1.62	96.2	1.83	113.2
FEV 1 % FVC	[%]		77.79		79.63	102.4
FVC IN	[L]	2.02	2.22	109.7	2.24	100.8
FIV1	[L]		2.08		2.04	98.1
FIV1 % VC MAX	[%]		93.69		88.66	94.6
MMEF 75/25	[L/s]	2.40	1.08	45.2	1.81	166.7
FEF 75	[L/s]	0.93	0.30	32.0	0.57	191.0
FEF 50	[L/s]	3.13	1.69	54.0	2.50	147.6
FEF 25	[L/s]	4.76	4.59	96.5	4.56	99.4
PEF	[L/s]	5.20	4.76	91.6	4.56	95.8
Date			19/06/23		19/06/23	
Time			09:19:07		09:52:58