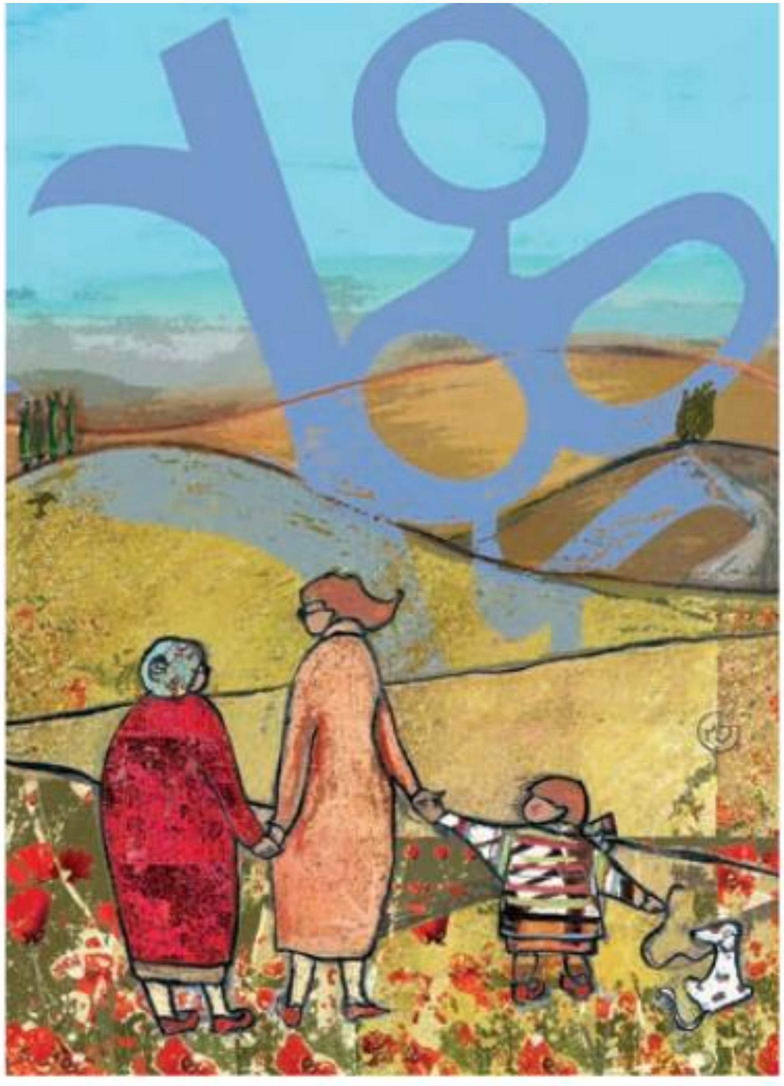


La NIV nel malato ristretto

Dr MP Dagostino
UOC di Geriatria
IRCCS Casa Sollievo della Sofferenza
San Giovanni Rotondo

Restrictive respiratory diseases are a heterogeneous group of disorders characterized by:

- ✓ reduction in all lung volumes (restriction)
- ✓ decreased compliance on pulmonary function testing
- ✓ preservation of expiratory flow.

Classification:

- **Intrinsic disorders:** such as interstitial lung diseases that cause diffuse inflammation or scarring of the lung tissue
- **Extrinsic disorders:** such as abnormalities of the chest wall , pleura, or abdomen that mechanically compress the lungs or limit their expansion
- **Neuromuscular diseases:** affecting chest wall nerves and muscles to decrease the ability of the respiratory muscles to inflate and deflate the lungs, resulting in chronically-reduced lung volumes and restrictive physiology

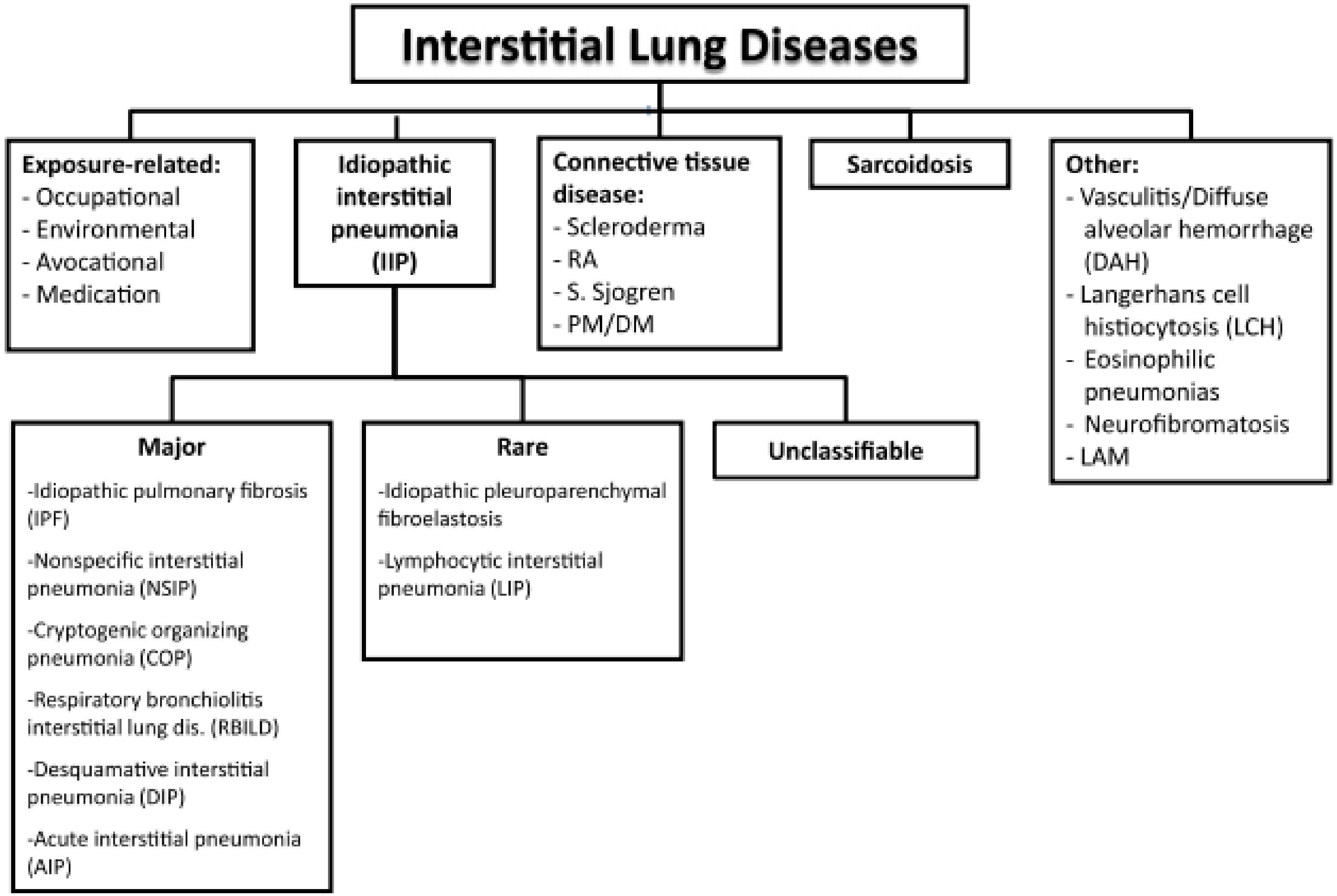
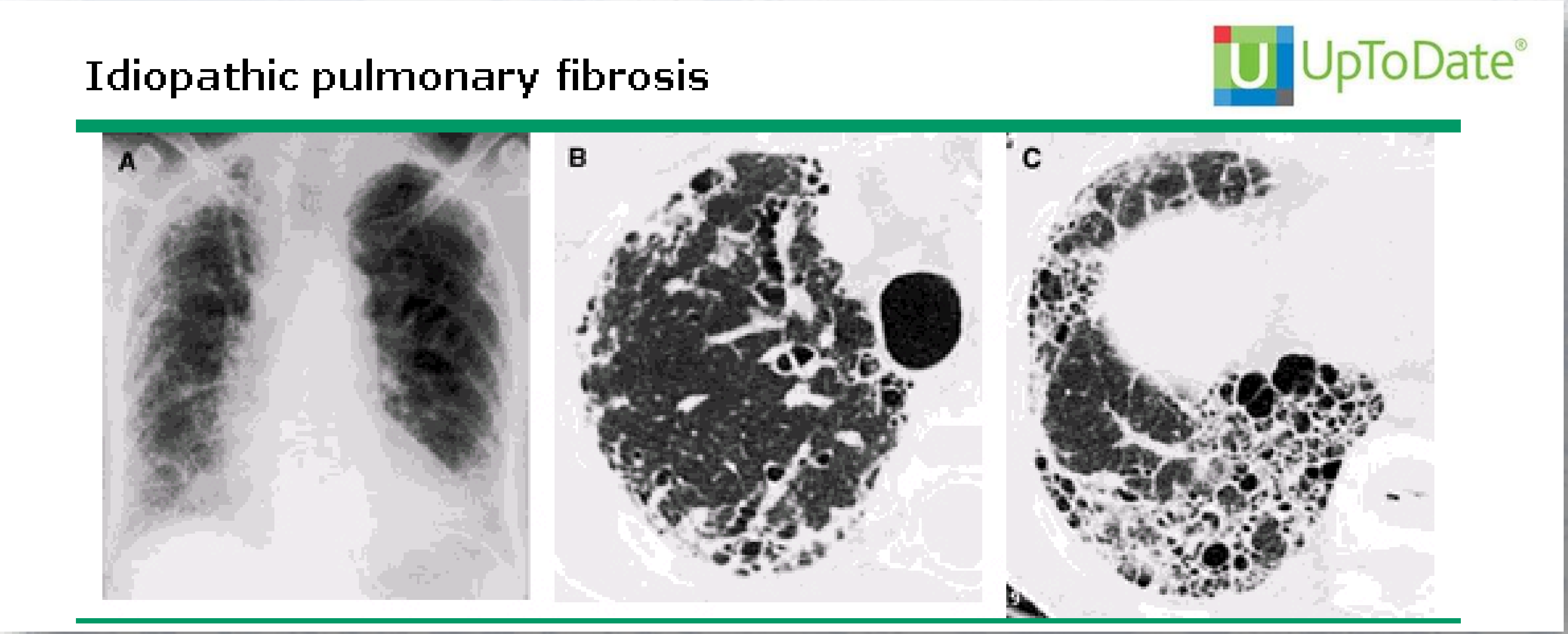


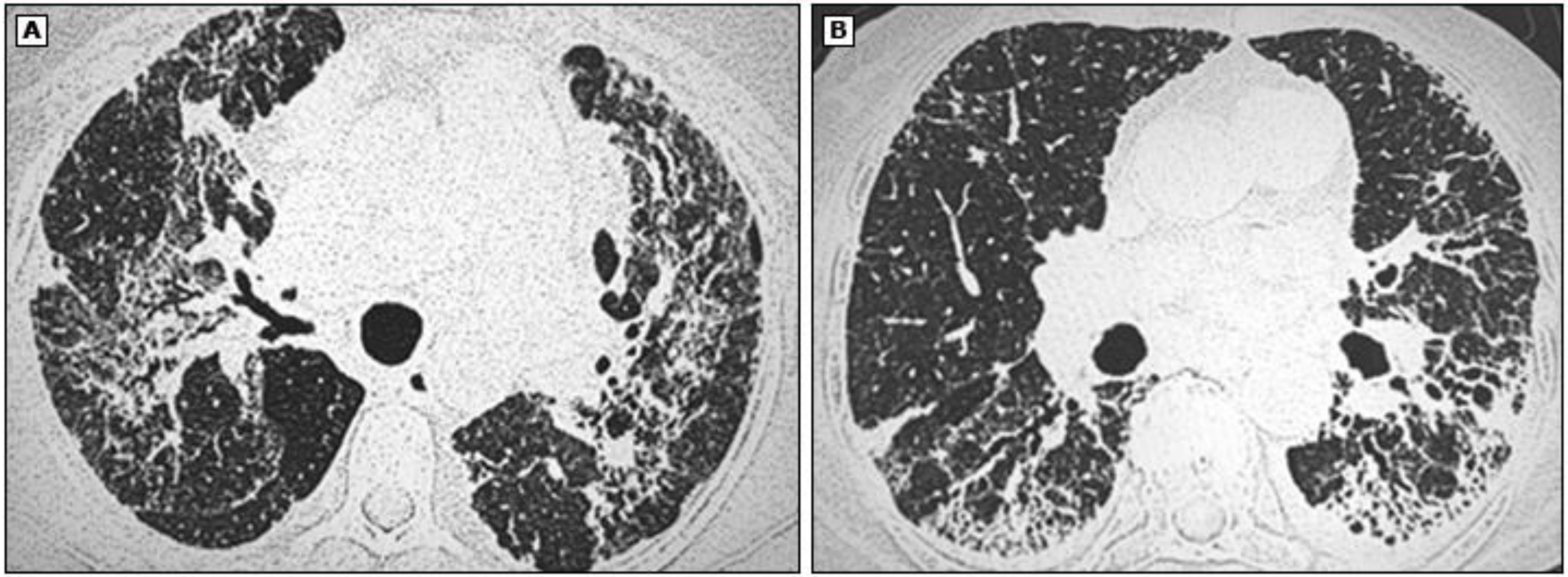
Fig. 1 Classification of Interstitial Lung Diseases. Footnotes: RA = Rheumatoid Arthritis; PM/DM = polymyositis/dermatomyositis; LAM = lymphangioleiomyomatosis

Idiopathic interstitial pneumonias

IPF is the most common type of idiopathic interstitial pneumonia (IIP). Clinical features of IPF include non-productive cough, dyspnea on exertion, a restrictive pattern on pulmonary function tests (PFTs), with impaired gas transfer, and bibasilar reticular opacities, traction bronchiectasis, and often subpleural honeycomb cysts on high resolution computed tomography (HRCT)

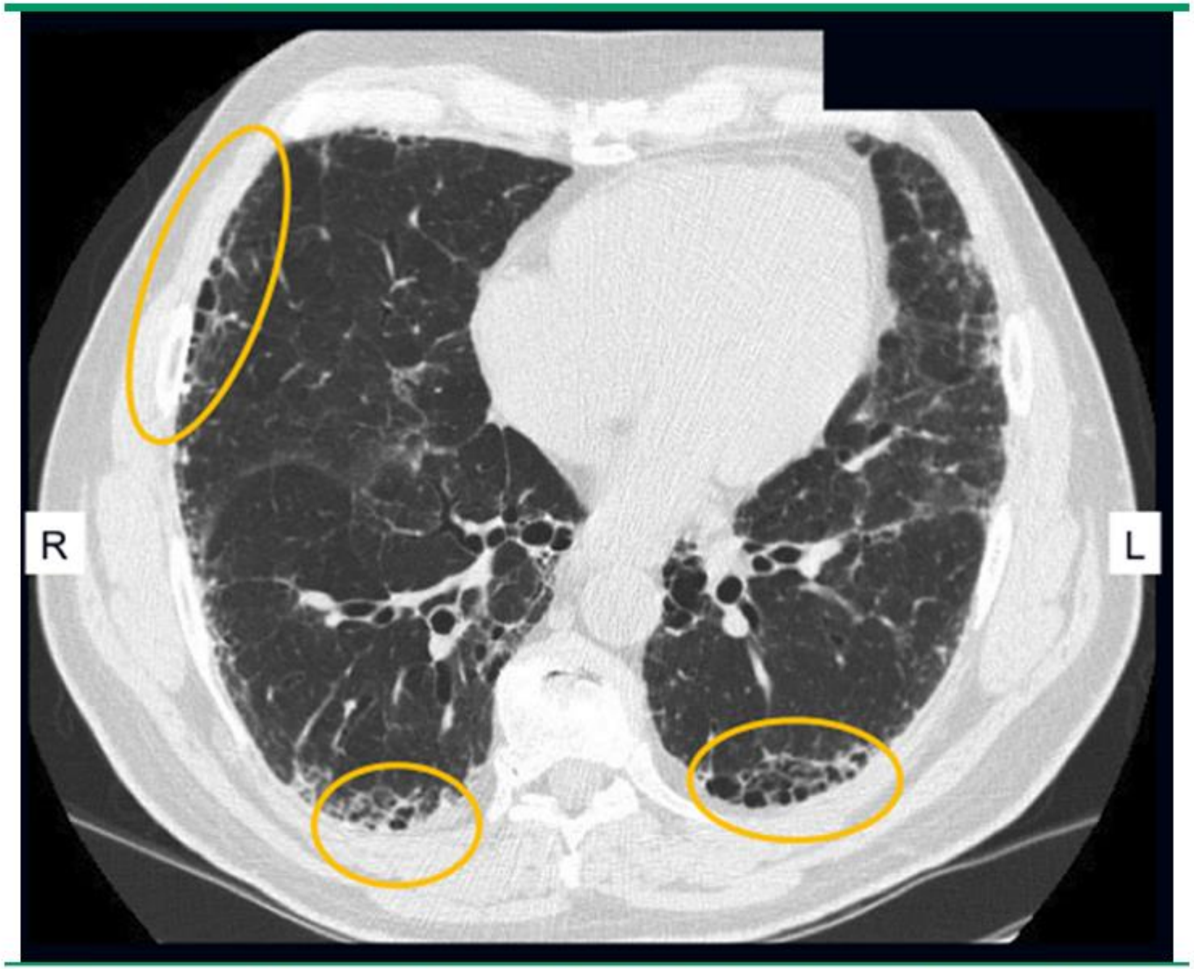


High resolution computed tomography of chronic hypersensitivity pneumonitis



High resolution computed tomography (HRCT) from a patient with chronic hypersensitivity pneumonitis. Image A from the upper lung zone shows a bronchocentric pattern of reticular opacities and traction bronchiectasis. Vaguely centrilobular ground-glass opacities are also present. Image B from a lower lung zone shows a peripheral, predominantly reticular pattern with traction bronchiectasis and honeycombing.

Chest HRCT showing honeycomb change



HRCT showing increased reticular opacities and patchy, subpleural areas of honeycomb change in a patient with idiopathic pulmonary fibrosis. Honeycomb change refers to clusters of cystic airspaces approximately 3 to 10 mm in diameter, usually in a subpleural location. Areas of honeycomb change are indicated by the ovals.

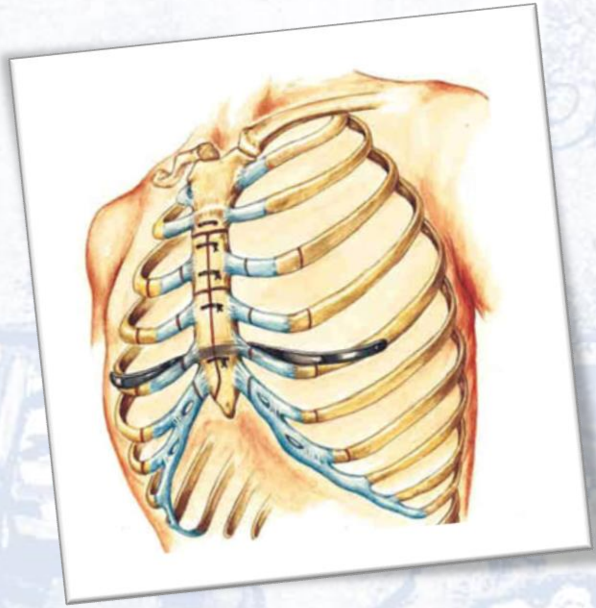
HRCT: high resolution computed tomography.

Extrinsic restrictive disorders :

Several conditions extrinsic to the lung parenchyma result in chronically-reduced lung volumes due to mechanical compression or other limitations on lung expansion.

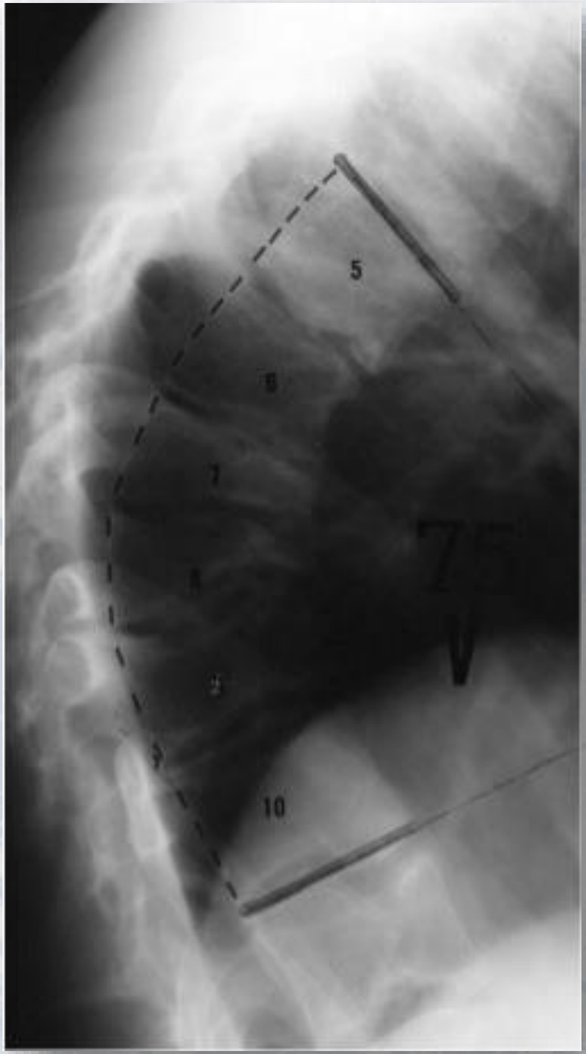
<u>Increased chest wall impedance :</u>

- ✓ Space-occupying pathological conditions
- ✓ Ankylosing spondylitis, kyphosis, scoliosis
- ✓ Congenital abnormalities of the chest wall
- ✓ Traumatic and iatrogenic abnormalities of the chest wall



<u>Increased intra-abdominal pressure :</u>

- ✓ Obesity
- ✓ Ascites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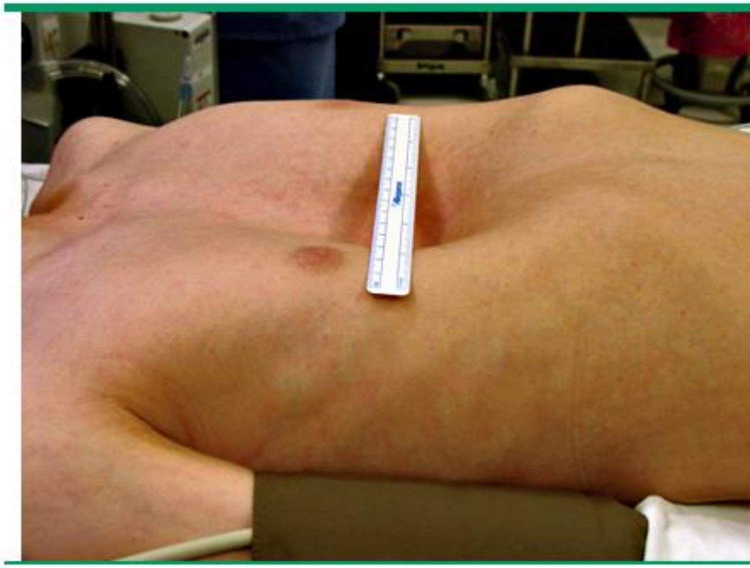
6
10

Pectus excavatum



Pectus excavtum in an adolescent patient demonstrating severe invagination of the lower sternum. The patient had significant exercise intolerance.

Reproduced with permission from: Chung EK. Visual Diagnosis in Pediatrics. Philadelphia: Lippincott Williams & Wilkins, 2006. Copyright © 2006 Lippincott Williams & Wilkins.

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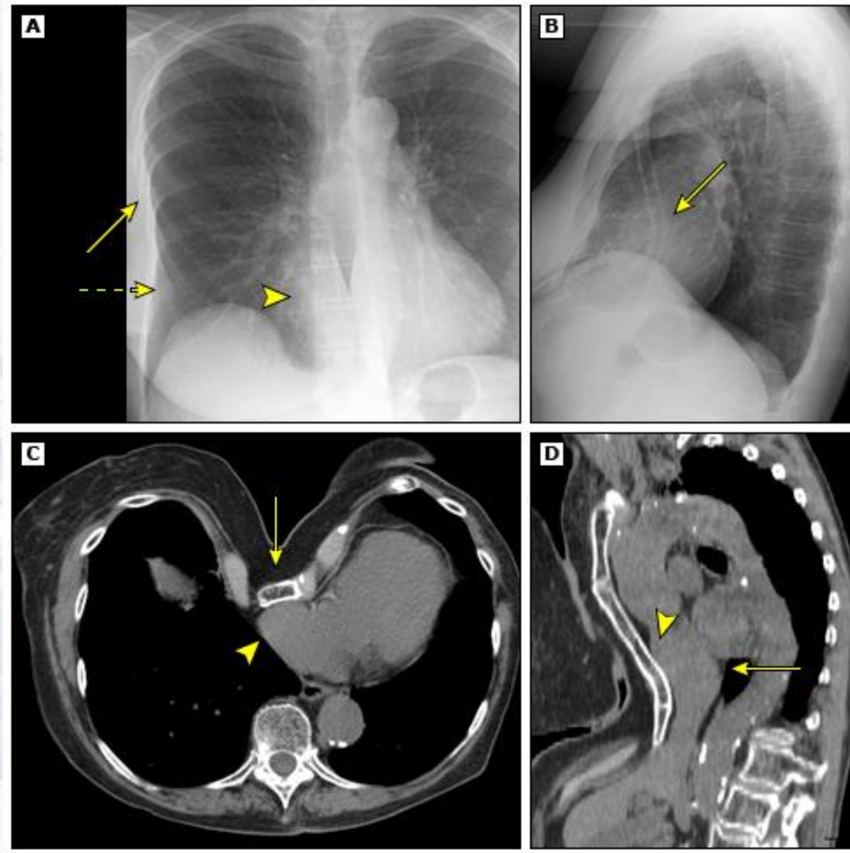
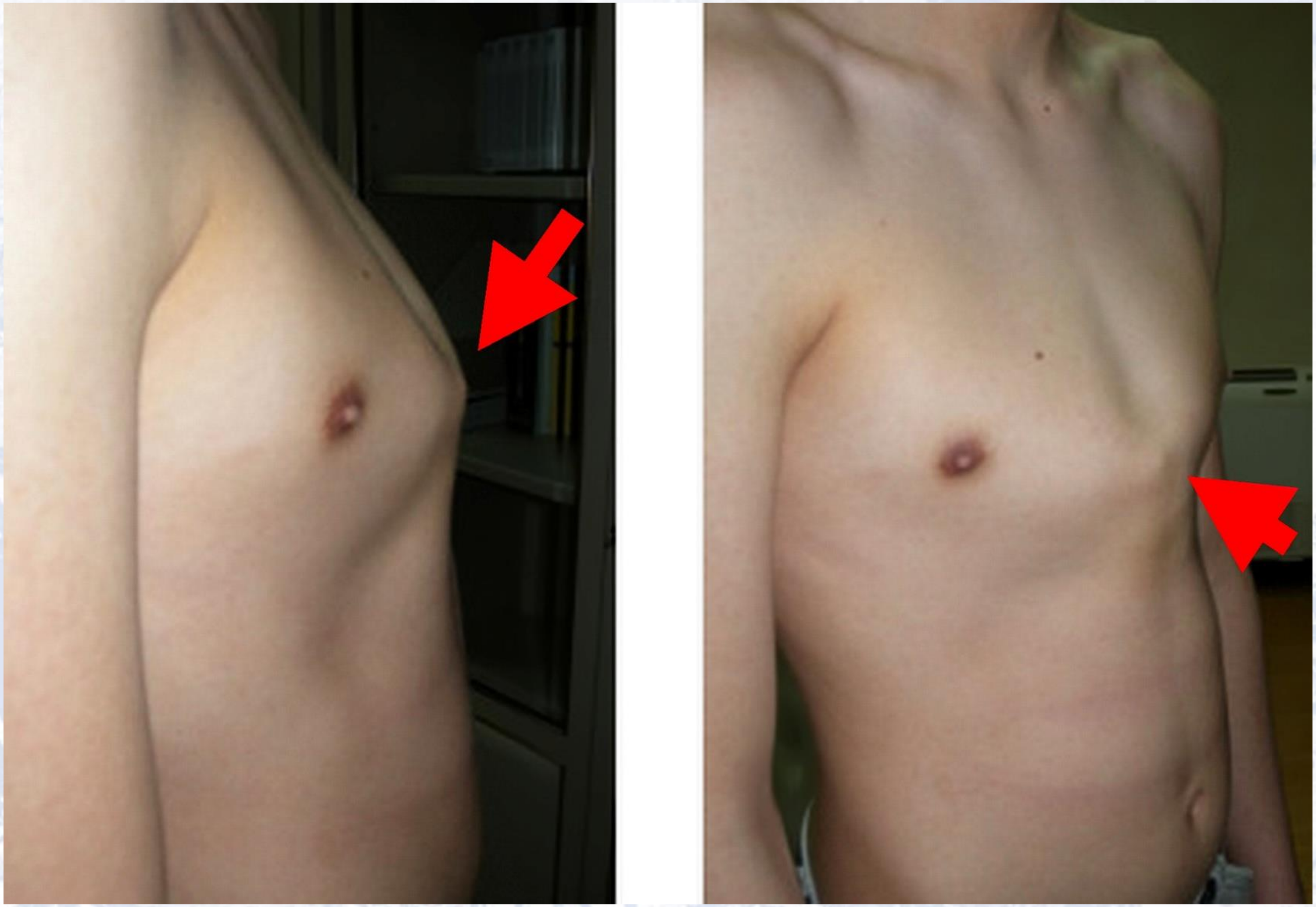
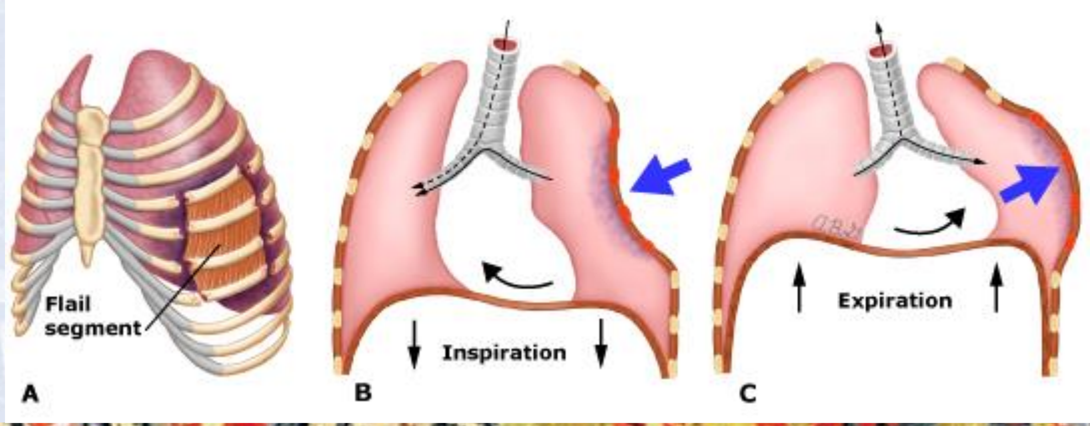


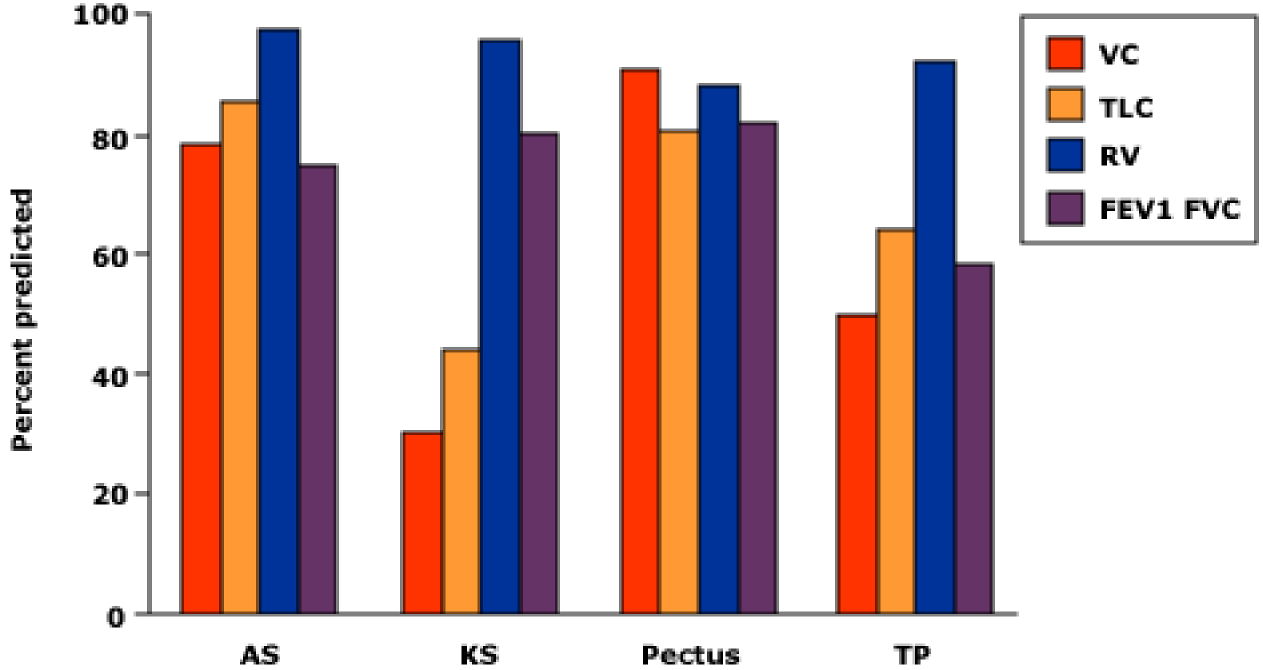


Poland Syndrome



Fibrodysplasia ossificans is a heritable disorder caused by a genetic variant in the gene ACVR1/ALK2 encoding Activin A receptor type I/Activin-like kinase 2, a bone morphogenetic protein (BMP) type I receptor. It is characterized by congenital malformations of the great toes and progressive heterotopic ossification, which can affect the chest wall and lead to respiratory insufficiency

Pulmonary function histogram

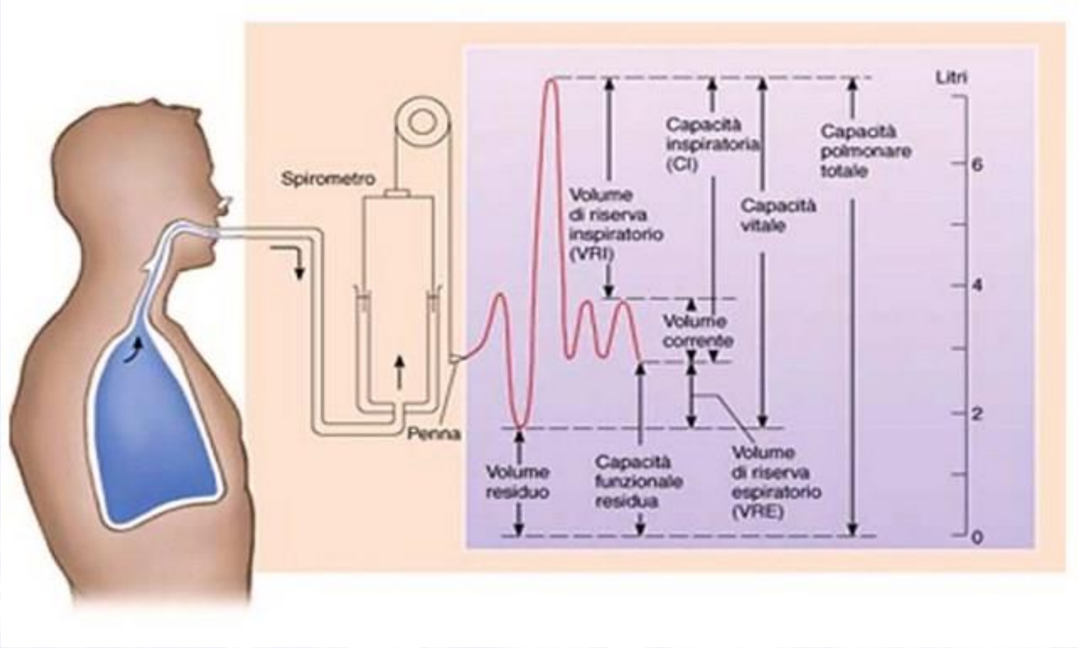


Histogram showing representative values of pulmonary function for patients with ankylosing spondylitis (AS), kyphoscoliosis (KS), pectus excavatum (Pectus), and thoracoplasty (TP).

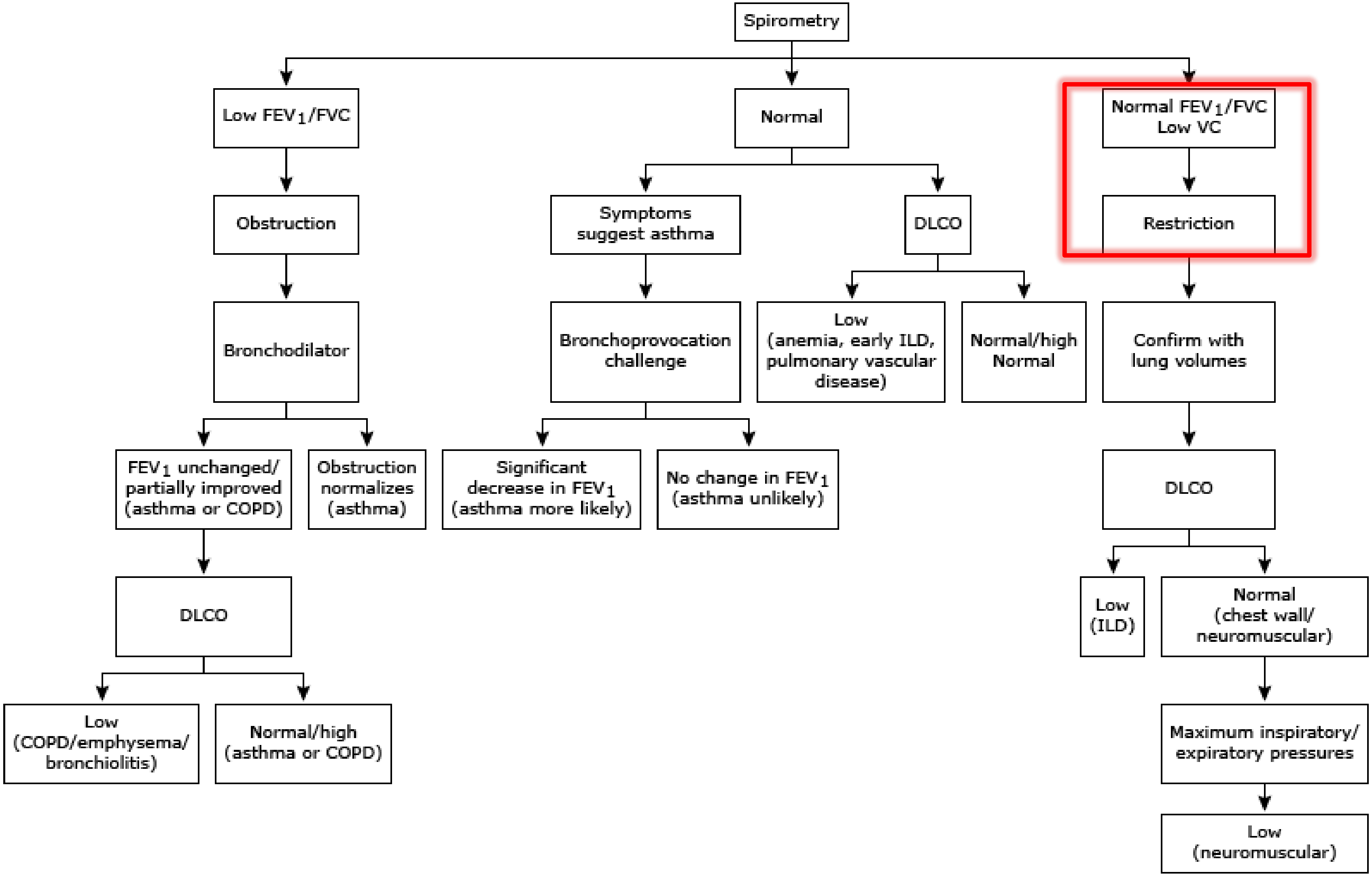
Adapted from McCool, FD, Rochester, DF. The lungs and chest Wall diseases. In: Textbook of Respiratory Medicine, 2nd ed, Vol 2, Murray, JF, Nadel, JA (Eds), WB Saunders, Philadelphia 1994.

Spirometry ***:***

The hallmark of restrictive physiology on spirometry is the presence of matched reductions in the total exhaled volume (known as the forced vital capacity [FVC]) and the volume exhaled in the first second (known as the forced expiratory volume in one second [FEV_1]), with absence of airflow obstruction

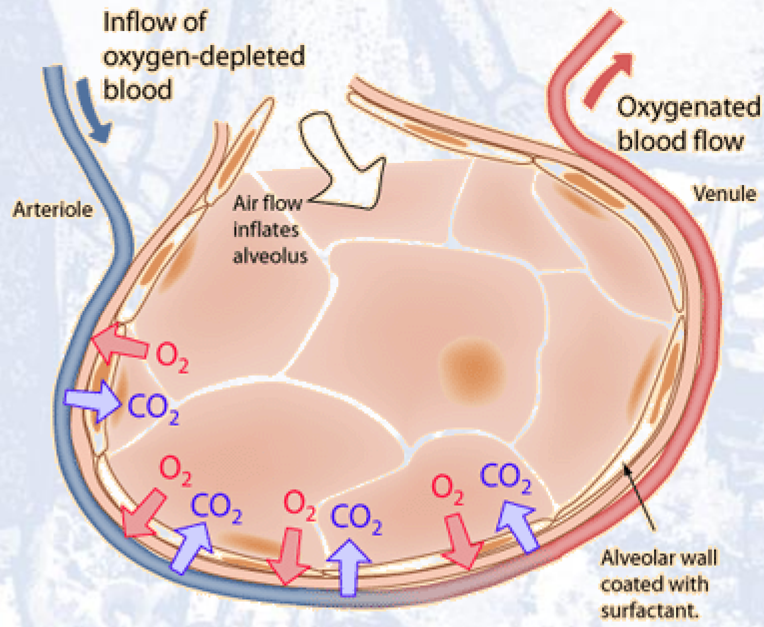


Algorithm for pulmonary function test interpretation

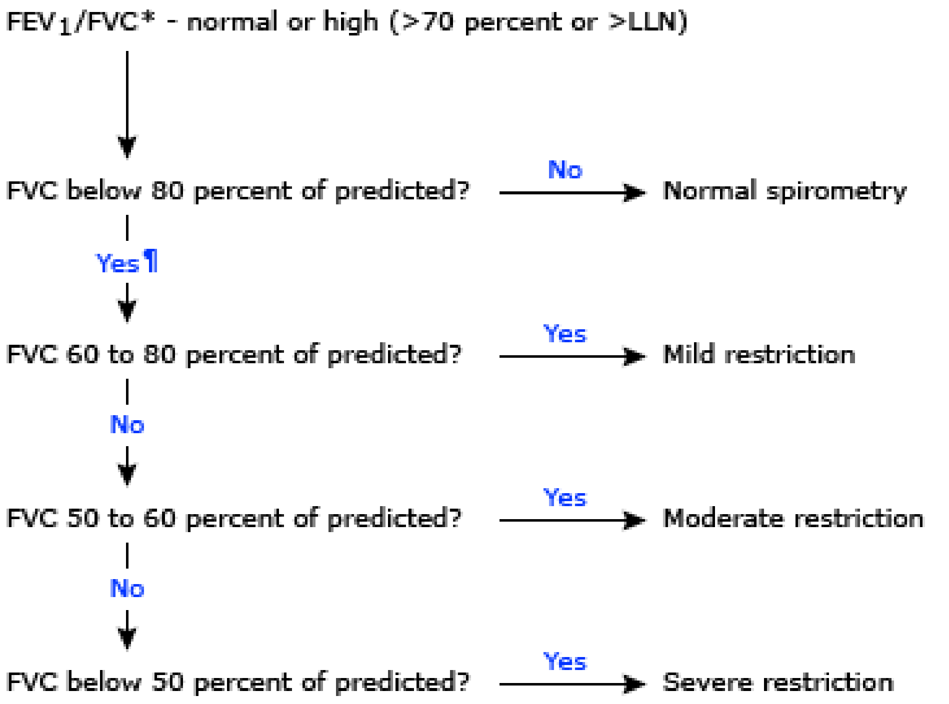


COPD: chronic obstructive pulmonary disease; DLCO: diffusing capacity for carbon monoxide; FEV_1: forced expiratory volume in one second; FVC: forced vital capacity; VC: vital capacity; ILD: interstitial lung disease.

Assessment of gas transfer – DLCO measurements aid in diagnosis of the underlying process. The DLCO helps to distinguish between ILD in which DLCO is usually reduced versus other causes of restriction in which DLCO is usually normal. Specifically, impaired DLCO suggests a process involving the pulmonary parenchyma (ie, ILD) or pulmonary vasculature (eg, PH), whereas a normal DLCO is typically seen in conditions that cause increased chest wall impedance or respiratory muscle weakness.



Interpretation of office spirometry: Restrictive pattern

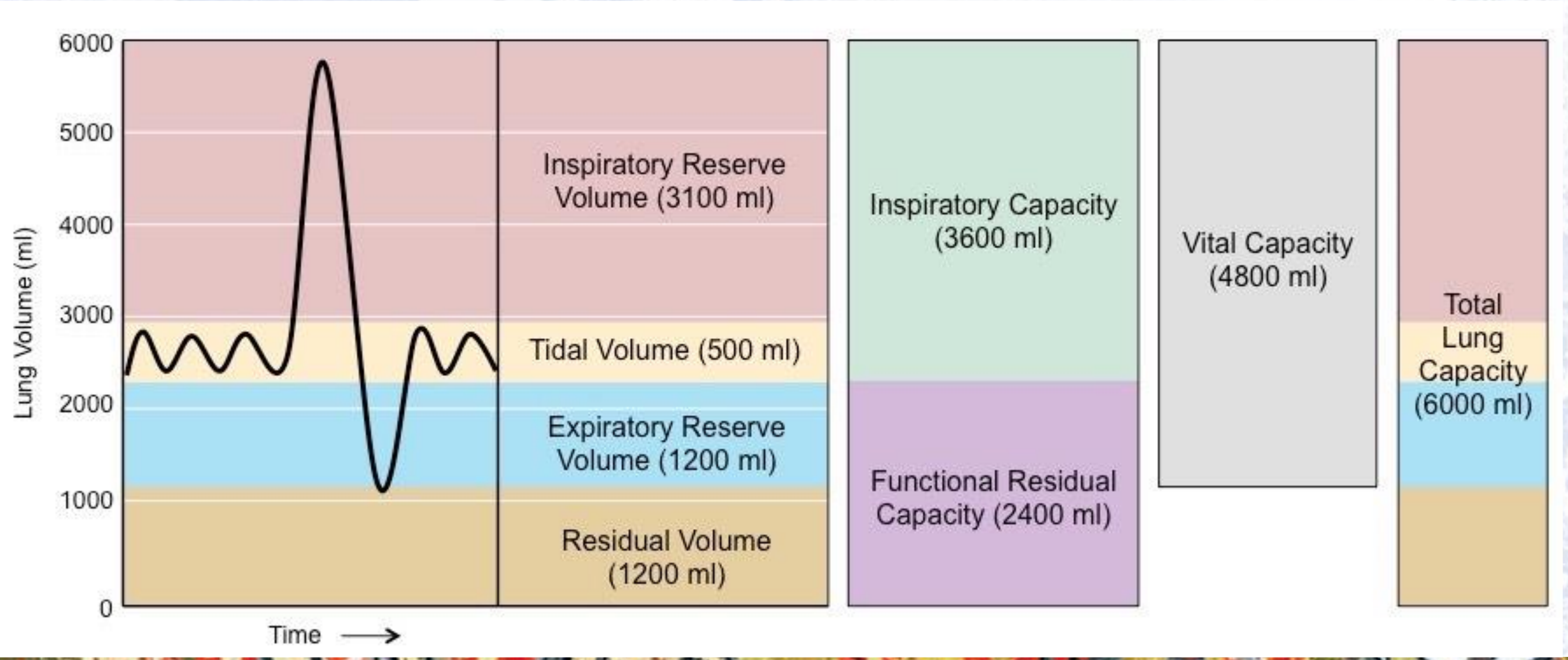


Spirometry interpretation flow chart for the detection of a restrictive ventilatory defect.

UpToDate®

Measurement of absolute lung volumes – While restrictive impairment is suspected based on spirometry values, measurement of lung volumes is necessary to confirm restriction and exclude air trapping since this causes similar changes on spirometry. Measurement of total lung capacity (TLC) is used to classify the severity of chronic restrictive disease:

- *Mild* – TLC 65 to 80% of predicted value
- *Moderate* – TLC 50 to 65% of predicted value
- *Severe* – TLC <50% of predicted value



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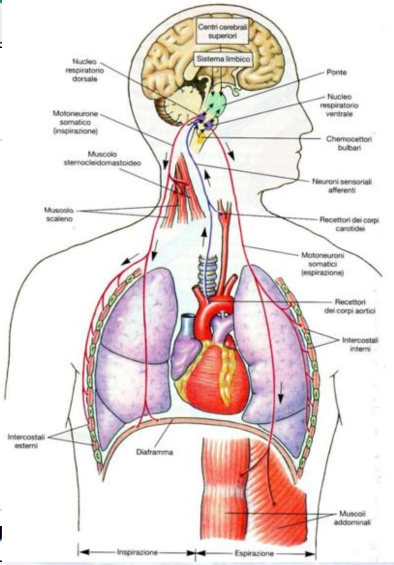
Respiratory pathway affecting carbon dioxide elimination	
Central nervous system ↓	"Won't breathe"
Peripheral nervous system ↓	"Can't breathe"
Respiratory muscles ↓	
Chest wall and pleura ↓	
Upper airway ↓	
Lungs	Abnormal gas exchange: "Can't breathe enoug



$$PaCO_2 = (k) \times VCO_2/[V_E(1 - V_D/V_T)]$$

Acute Setting

2019

Official Journal of the Asian Pacific Society of Respirology
Respirology

Respirology (2019) 24,
308–317doi: 10.1111/resp.13469

INVITED REVIEW SERIES:
NON-INVASIVE VENTILATION
SERIES EDITORS: AMANDA PIPER AND CHUNG-MING CHU

Benefits of non-invasive ventilation in acute hypercapnic respiratory failure

VITTORIA COMELLINI,[1] ANGELA MARIA GRAZIA PACILLI[2] AND STEFANO NAVA[1,2]

[1]*Respiratory and Critical Care Unit, University Hospital St Orsola-Malpighi, Bologna, Italy;* [2]*Department of Specialistic, Diagnostic and Experimental Medicine (DIMES), Alma Mater Studiorum University of Bologna, Bologna, Italy*

Table 1 Indications for NIV in acute hypercapnic respiratory failure

1	Acute exacerbation of COPD
2	Cardiogenic pulmonary oedema
3	Obesity and obesity hypoventilation syndrome
4	Weaning from invasive mechanical ventilation
5	Prevention of post-extubation failure in those patients previously intubated
6	Chest wall diseases and neuromuscular diseases
7	Palliative care and do-not-intubate patients
8	Miscellaneous (very old patients, community-acquired pneumonia and bridge to transplantation)

2018

Faverio *et al. BMC Pulmonary Medicine* (2018) 18:70
https://doi.org/10.1186/s12890-018-0643-3

BMC Pulmonary Medicine

REVIEW Open Access

Management of acute respiratory failure in interstitial lung diseases: overview and clinical insights

Paola Faverio[1*], Federica De Giacomi[1], Luca Sardella[1], Giuseppe Fiorentino[2], Mauro Carone[3], Francesco Salerno[3], Jousel Ora[4], Paola Rogliani[4], Giulia Pellegrino[5], Giuseppe Francesco Sferrazza Papa[5], Francesco Bini[6], Bruno Dino Bodini[7], Grazia Messinesi[1], Alberto Pesci[1] and Antonio Esquinas[8]

Problem	Tip for solution
High pressures required to obtain ideal tidal volume in fibrotic lung with risk of pneumothorax	- Tolerate low tidal volumes with higher respiratory rate to obtain acceptable minute ventilation - Low to moderate PEEP levels to avoid overdistension of "healthy" lung units
High respiratory rate that hampers patient-ventilator adaptation	- Titrate drugs to control respiratory rate, e.g. opiates (morphine or fentanyl)[§]
Intense breathlessness reported by patients especially in the acute phase	- Titrate drugs to control respiratory rate, e.g. opiates (morphine or fentanyl)[§] - Rapid inspiratory curve - Increase FIO2

2017

TASK FORCE REPORT
ERS/ATS GUIDELINES

CrossMark

Official ERS/ATS clinical practice guidelines: noninvasive ventilation for acute respiratory failure

Bram Rochwerg[1], Laurent Brochard[2,3], Mark W. Elliott[4], Dean Hess[5], Nicholas S. Hill[6], Stefano Nava[7] and Paolo Navalesi[8] (members of the steering committee); Massimo Antonelli[9], Jan Brozek[1], Giorgio Conti[9], Miquel Ferrer[10], Kalpalatha Guntupalli[11], Samir Jaber[12], Sean Keenan[13,14], Jordi Mancebo[15], Sangeeta Mehta[16] and Suhail Raoof[17,18] (members of the task force)

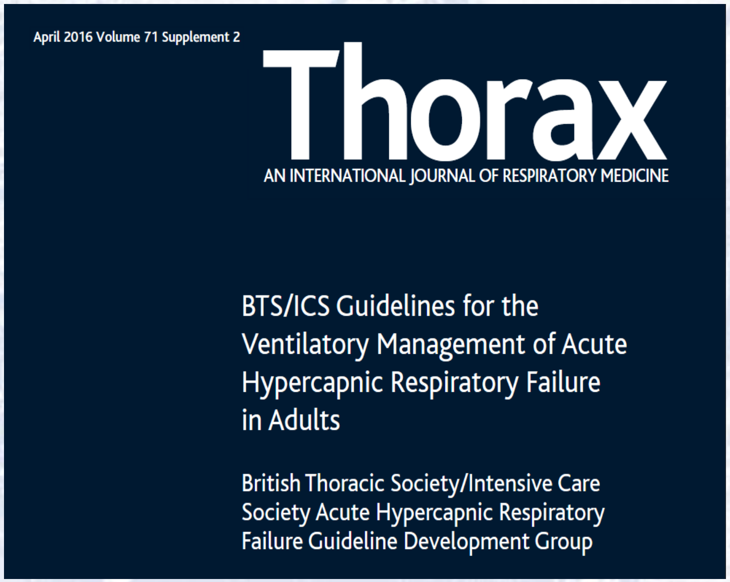
PICO: *(population–intervention–comparison–outcome) format*
GRADE *:(Grading of Recommendations, Assessment, Development and Evaluation) methodology*

TABLE 2 Recommendations for actionable PICO questions

Clinical indication[#]	Certainty of evidence[¶]	Recommendation
Prevention of hypercapnia in COPD exacerbation	⊕⊕	Conditional recommendation against
Hypercapnia with COPD exacerbation	⊕⊕⊕⊕	Strong recommendation for
Cardiogenic pulmonary oedema	⊕⊕⊕	Strong recommendation for
Acute asthma exacerbation		No recommendation made
Immunocompromised	⊕⊕⊕	Conditional recommendation for
De novo respiratory failure		No recommendation made
Post-operative patients	⊕⊕⊕	Conditional recommendation for
Palliative care	⊕⊕⊕	Conditional recommendation for
Trauma	⊕⊕⊕	Conditional recommendation for
Pandemic viral illness		No recommendation made
Post-extubation in high-risk patients (prophylaxis)	⊕⊕	Conditional recommendation for
Post-extubation respiratory failure	⊕⊕	Conditional recommendation against
Weaning in hypercapnic patients	⊕⊕⊕	Conditional recommendation for

#: all in the setting of acute respiratory failure; ¶: certainty of effect estimates: ⊕⊕⊕⊕, high; ⊕⊕⊕, moderate; ⊕⊕, low; ⊕, very low.

2016


April 2016 Volume 71 Supplement 2

Thorax

AN INTERNATIONAL JOURNAL OF RESPIRATORY MEDICINE

BTS/ICS Guidelines for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults

British Thoracic Society/Intensive Care Society Acute Hypercapnic Respiratory Failure Guideline Development Group

1/2

Restrictive lung diseases (NMD and CWD)

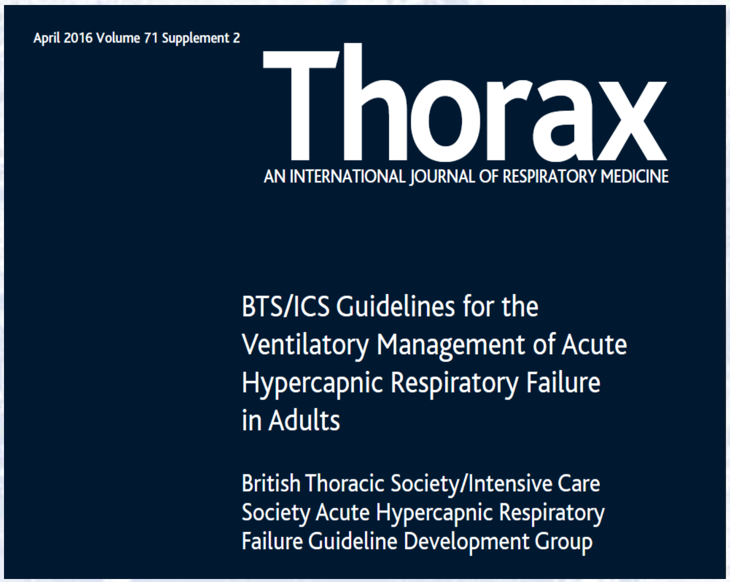
Recommendations:

49. Controlled oxygen therapy should be used in patients with NMD or CWD and AHRF (Grade D).
50. NIV should almost always be trialled in the acutely unwell patients with NMD or CWD with hypercapnia. Do not wait for acidosis to develop (Grade D).
51. In patients with NMD or CWD, NIV should be considered in acute illness when vital capacity (VC) is known to be <1 L and RR >20, even if normo-capnic (Grade D).
52. In patients with NMD or CWD, consider controlled ventilation as triggering may be ineffective (Grade D).
53. In NMD or CWD, unless escalation to IMV is not desired by the patient, or is deemed to be inappropriate, intubation should not be delayed if NIV is failing (Grade D)

Good practice points:

- Patients with NMD usually require low levels of PS (IPAP 8-12 cmH2O)
- Patients with chest wall deformity usually require higher levels of PS (IPAP > 20 cmH2O)
- PEEP in the range of 5–10 is commonly required to increase residual volume and reduce oxygen dependency in both patient groups.

Obesity hypoventilation syndrome


April 2016 Volume 71 Supplement 2

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BTS/ICS Guidelines for the Ventilatory Management of Acute Hypercapnic Respiratory Failure in Adults

British Thoracic Society/Intensive Care Society Acute Hypercapnic Respiratory Failure Guideline Development Group

Recommendations:

56. Controlled oxygen therapy should be used in patients with OHS and AHRF (Grade D).

57. In patients with OHS, NIV should be started in AHRF using the same criteria as in AECOPD (Grade B).

58. NIV is indicated in some hospitalised obese hypercapnic patients with daytime somnolence, sleep disordered breathing and/or right heart failure in the absence of acidosis (Grade D).

Good practice points:

- High inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP) settings are commonly required in patients with OHS (eg, IPAP>30, EPAP>8).
- Volume control (or volume assured) modes of providing NIV may be more effective when high inflation pressures are required.

Indications for NIV

COPD
pH <7.35
pCO2 >6.5
RR>23
If persisting after bronchodilators and controlled oxygen therapy

Neuromuscular disease
Respiratory illness with RR > 20 if usual VC <1L even if pCO2<6.5
Or
pH < 7.35 and pCO2> 6.5

Obesity
pH <7.35, pCO2>6.5, RR>23
Or
Daytime pCO2> 6.0 and somnolent

<u>NIV Not indicated</u>
Asthma/Pneumonia
Refer to ICU for consideration IMV if increasing respiratory rate/distress
or
pH <7.35 and pCO2 >6.5

Contraindications for NIV

Absolute
Severe facial deformity
Facial burns
Fixed upper airway obstruction

Relative
pH<7.15
(pH<7.25 and additional adverse feature)
GCS <8
Confusion/agitation
Cognitive impairment (warrants enhanced observation)

Indications for referral to ICU
AHRF with impending respiratory arrest

NIV failing to augment chest wall movement or reduce pCO2

Inability to maintain Sao2 > 85-88% on NIV

Need for IV sedation or adverse features indicating need for closer monitoring and/or possible difficult intubation as in OHS, DMD.

NIV SETUP

Mask
Full face mask (or own if home user of NIV)

Initial Pressure settings
EPAP: 3 (or higher if OSA known/expected)

IPAP in COPD/OHS/KS 15 (20 if pH <7.25)

Up titrate IPAP over 10-30 mins to IPAP 20—30 to achieve adequate augmentation of chest/abdo movement and slow RR

IPAP should not exceed 30 or EPAP 8* without expert review

IPAP in NM 10 (or 5 above usual setting)

Backup rate
Backup Rate of 16-20. Set appropriate inspiratory time

I:E ratio
COPD 1:2 to 1.3
OHS, NM & CWD 1:1

Inspiratory time
0.8-1.2s COPD
1.2-1.5s OHS, NM & CWD

Use NIV for as much time as possible in 1st 24hours.
Taper depending on tolerance & ABGs over next 48-72 hours

SEEK AND TREAT REVERSIBLE CAUSES OF AHRF

*** Possible need for EPAP > 8**
Severe OHS (BMI >35), lung recruitment eg hypoxia in severe kyphoscolios, oppose intrinsic PEEP in severe airflow obstruction or to maintain adequate PS when high EPAP required

NIV Monitoring

Oxygenation
Aim 88-92% in all patients

Note: Home style ventilators CANNOT provide > 50% inspired oxygen.

If high oxygen need or rapid desaturation on disconnection from NIV consider IMV.

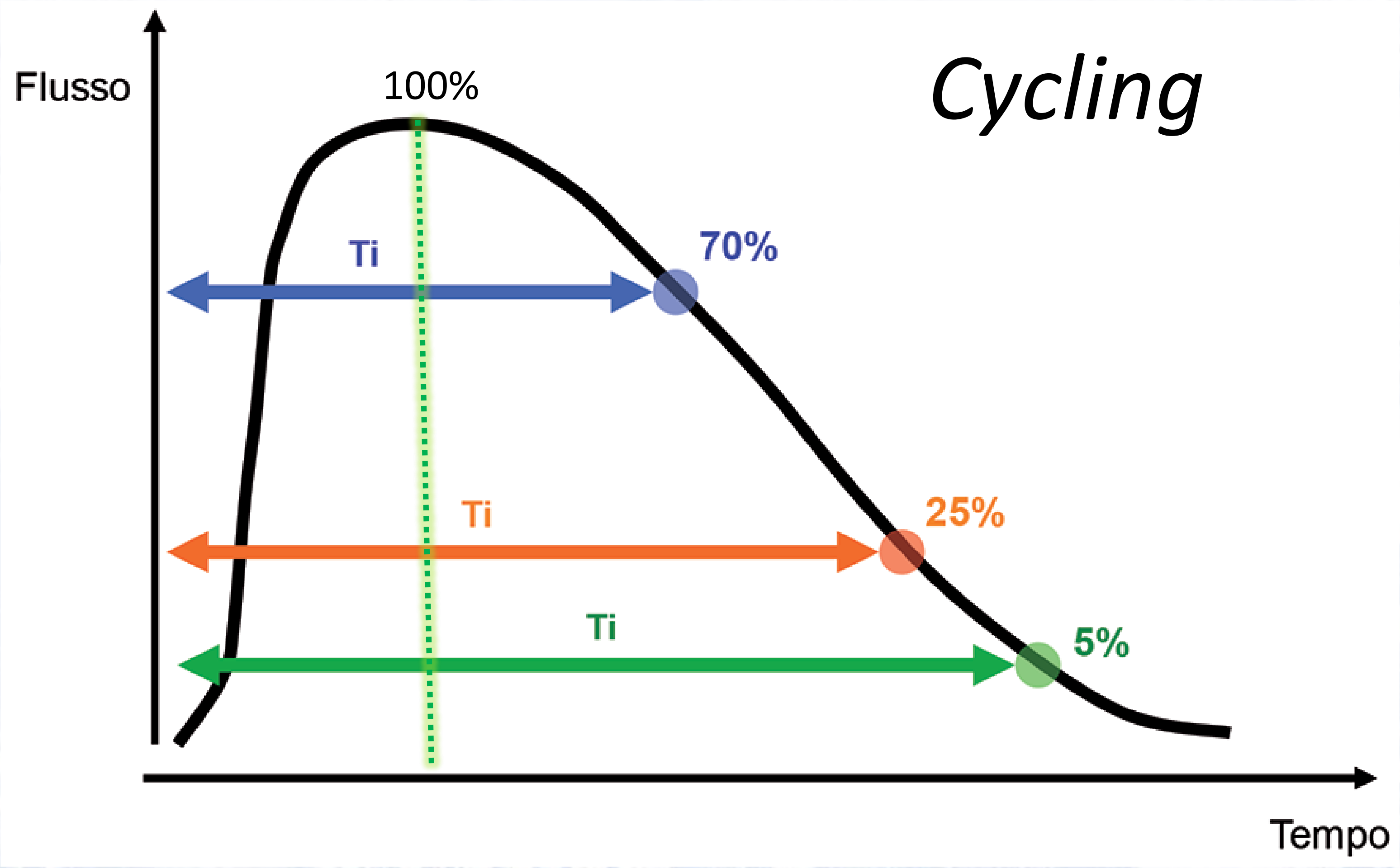
Red flags
pH <7.25 on optimal NIV
RR persisting > 25
New onset confusion or patient distress

Actions
Check synchronisation, mask fit, exhalation port : give physiotherapy/bronchodilators, consider anxiolytic

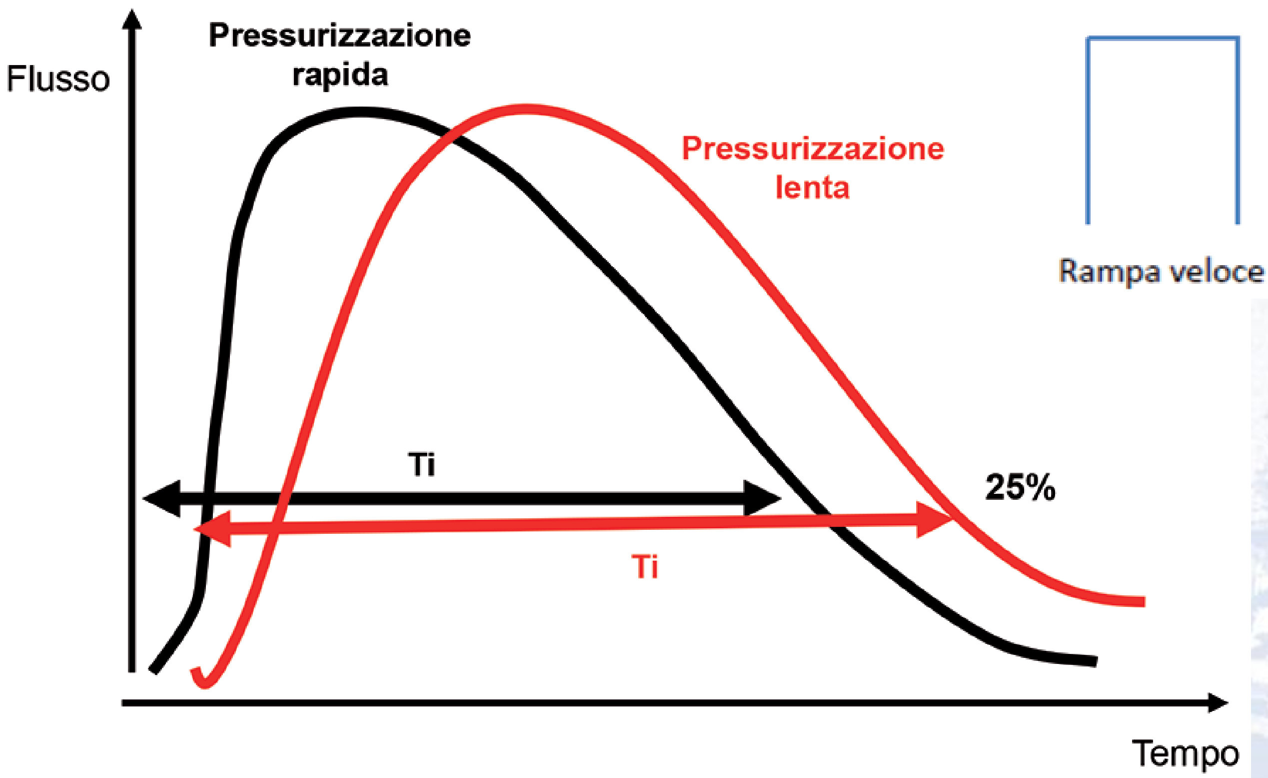
CONSIDER IMV

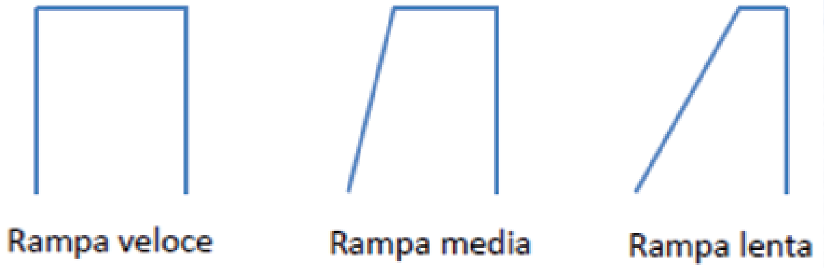
Mode variables:

- *Trigger*
- *Variabile di controllo*
- *Cycling*


Flusso
100%
Cycling
Ti
70%
Ti
25%
Ti
5%
Tempo

Rise time


Flusso
Pressurizzazione rapida
Pressurizzazione lenta
Ti
25%
Ti
Tempo


Rampa veloce
Rampa media
Rampa lenta

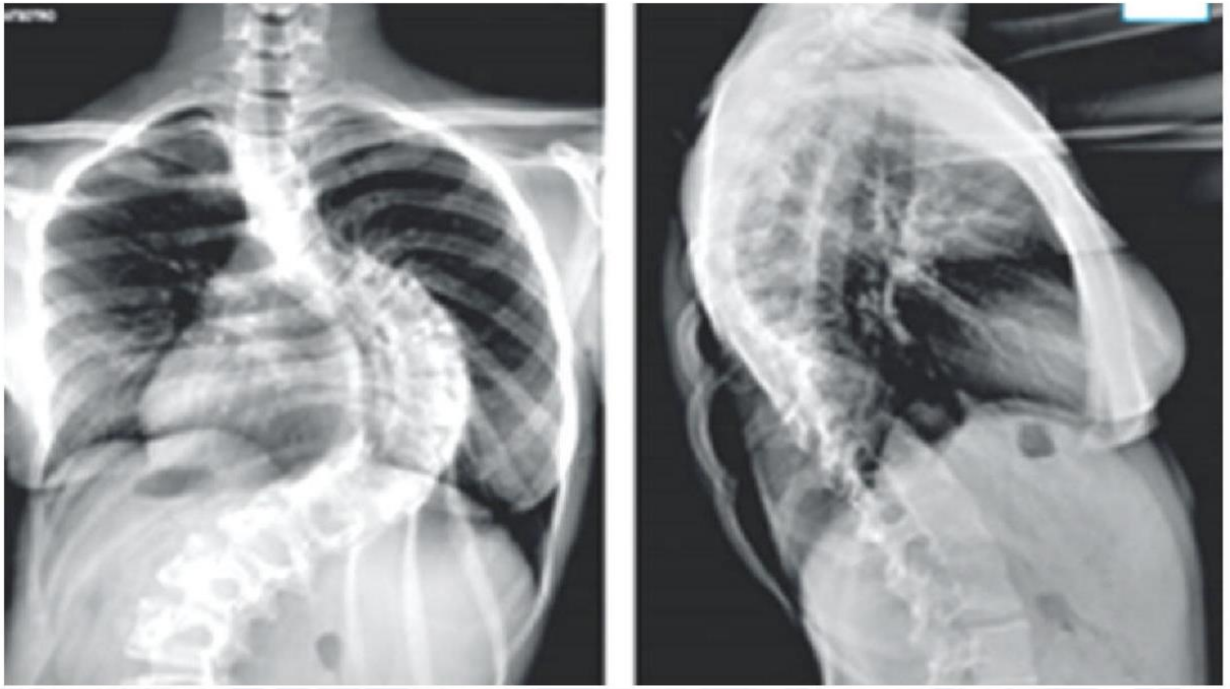
Chronic Setting

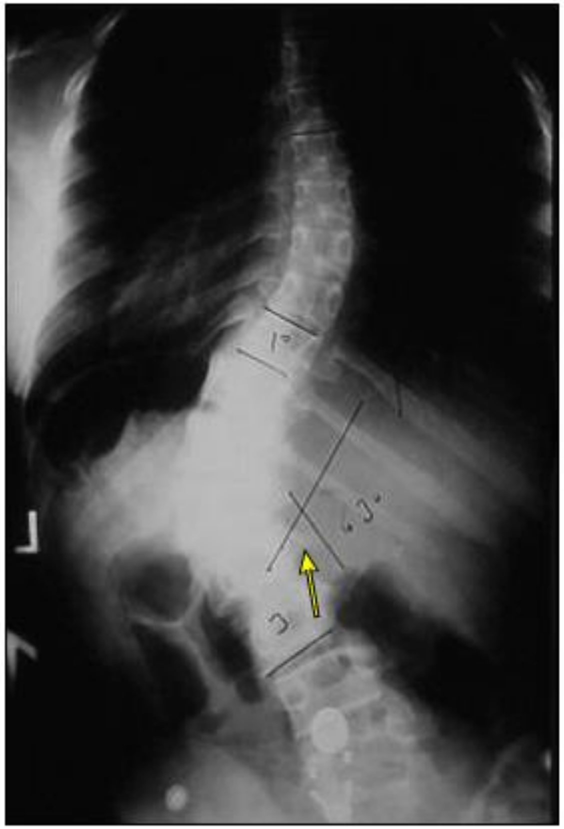
Hyperkyphosis

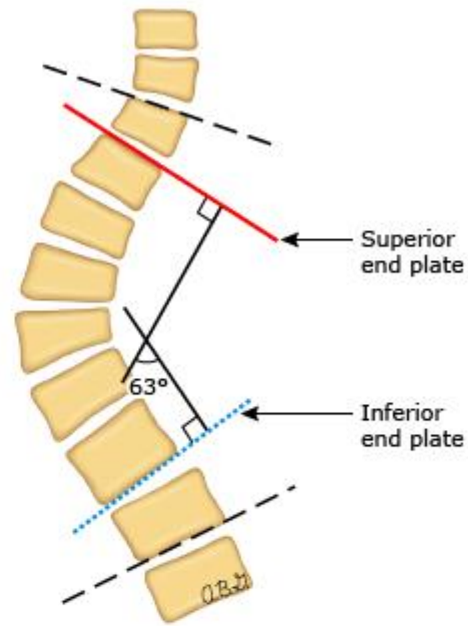
Hyperkyphosis is excessive curvature of the thoracic spine, commonly known as the "dowager's hump." Although it is also referred to as kyphosis, the term kyphosis is meant to describe the sagittal convexity, or forward curvature, of the normal thoracic spine which can range from normal to abnormal. Kyphosis tends to progress with age

ETIOLOGY

- Vertebral fractures
- Low bone density
- Short vertebral height
- Degenerative disc disease
- Postural changes
- Muscle weakness
- Intervertebral ligaments
- Genetic/metabolic conditions



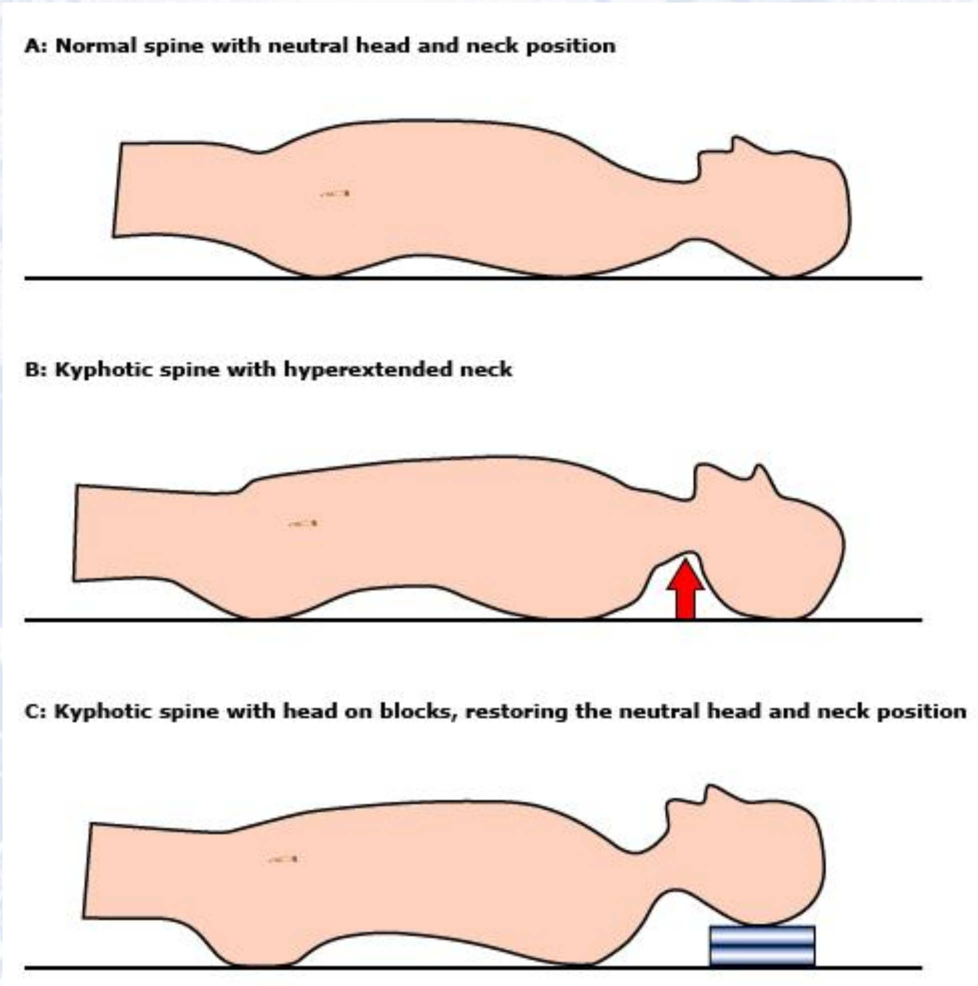


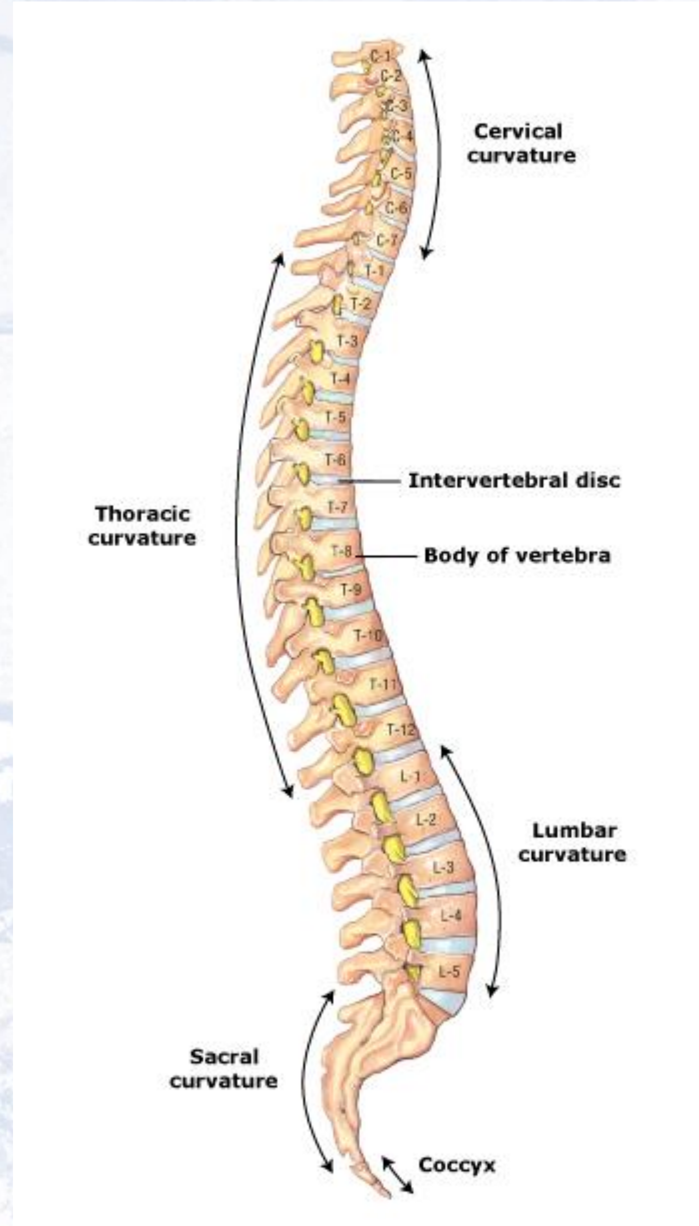


The **Cobb angle** (arrow in radiograph) is formed by the intersection of a line parallel to the superior end plate of the most cephalad vertebra in a particular curve, with the line parallel to the inferior end plate of the most caudad vertebra of the curve. The intersection of these lines may occur outside the border of the actual film. Therefore, by convention, perpendiculars to the parallels are drawn, and the angle between their intersection is measured.

Debrunner kyphometer

Distance from the occiput-to-wall





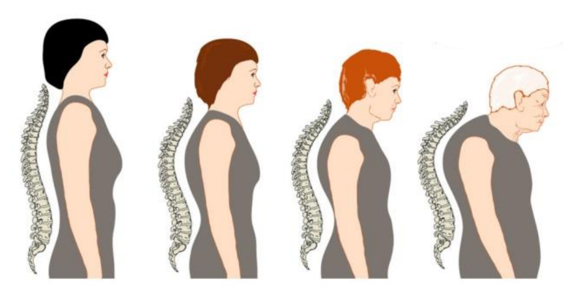
Effect of aging :

- ✓ Chest wall compliance decreases with age;
- ✓ Increasing work of breathing and risk of respiratory muscle fatigue;
- ✓ Patients tend to breathe with lower tidal volumes and increased respiratory rate;
- ✓ Dead space fraction may be increased;
- ✓ Alveolar hypoventilation may ensue with resultant hypercapnia;
- ✓ Hypoxemia without hypercapnia is seen in moderate to severe disease;
- ✓ Ventilation-perfusion (V/Q) mismatch has been reported with a scoliosis angle greater than 65 degrees;
- ✓ Pulmonary hypertension develops as a result of persistent hypoxemia;
- ✓ Nocturnal hypoventilation and arterial oxygen desaturation are described, particularly during rapid eye movement (REM) sleep.

Mateusz Pajdziński, Paulina Młynarczyk, Joanna Miłkowska-Dymanowska, Adam J. Białas, Moin Aldin Muhammad Afzal, Wojciech J. Piotrowski, Paweł Górski
Department of Pneumology and Allergy, 1st Chair of Internal Medicine, Medical University of Lodz, Łódź, Poland

Adv. Respir. Med. 2017; 85: 352–357

Kyphoscoliosis — what can we do for respiration besides NIV?

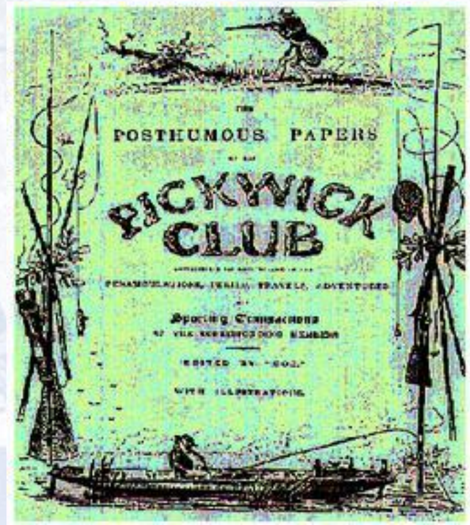


- ✓ NIV is now regarded as an option of treating CRF in patients with KS;
- ✓ The survival rate of patients using home mechanical ventilation was three times higher than that of patients using long-term oxygen therapy alone (LTOT);
- ✓ If the patient has obstructive disorders, continuous positive airways pressure (CPAP) is recommended; however, this may not be sufficient in other kinds of respiratory pathology (NIV);
- ✓ After three months of NIV in KS patients the length and quality of REM sleep improved, while oxygenation increased and transcutaneous carbon dioxide tension decreased;

Obesity hypoventilation syndrome

"Pickwickian syndrome"

Obesity is associated with restrictive physiology on pulmonary function testing due to increased weight of the chest wall and increased abdominal adipose tissue restricting diaphragmatic motion.
The severity of obesity is most commonly determined by the body mass index (BMI); a BMI greater than 30 kg/m^2 is commonly used as a definition of obesity.



Obesity Hypoventilation Syndrome (OHS) is defined as the presence of **awake** alveolar hypoventilation in an obese individual which cannot be attributed to other conditions associated with alveolar hypoventilation.

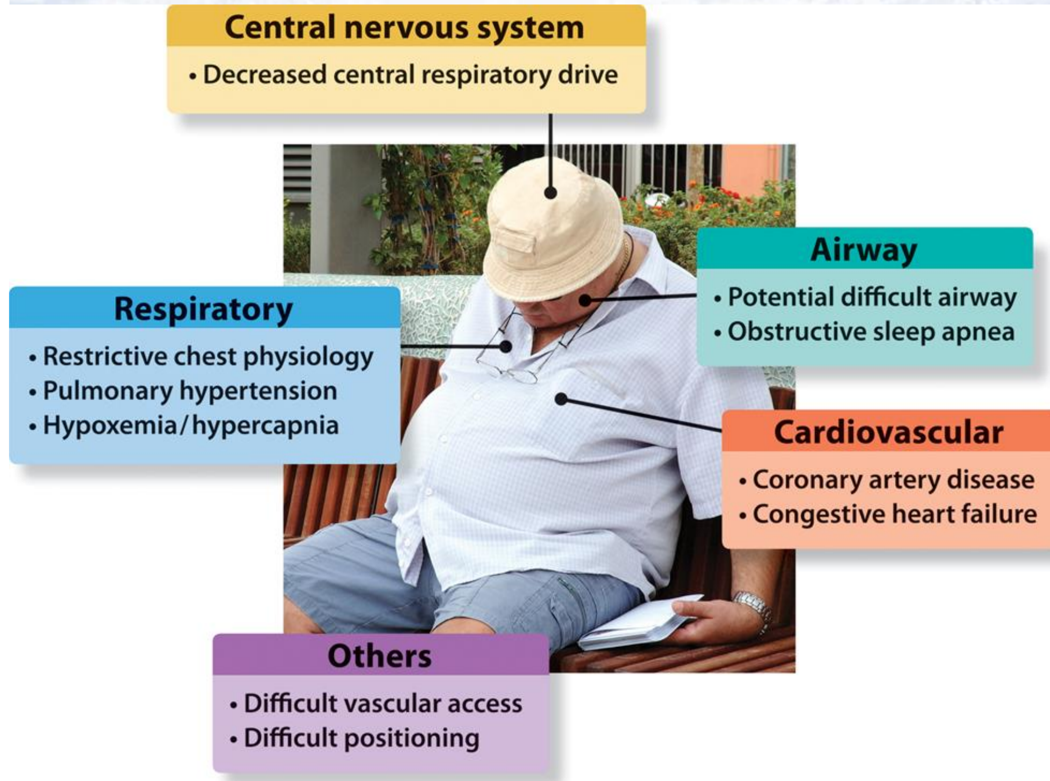
OHS is a ***diagnosis of exclusion*** that can be made when the following criteria are met:

- ✓ **Obesity (body mass index [BMI] >30 kg/m^2)**
- ✓ **Awake alveolar hypoventilation as indicated by a partial arterial pressure of carbon dioxide (CO_2) >45 mmHg**
- ✓ **Alternative causes hypercapnia and hypoventilation have been excluded**

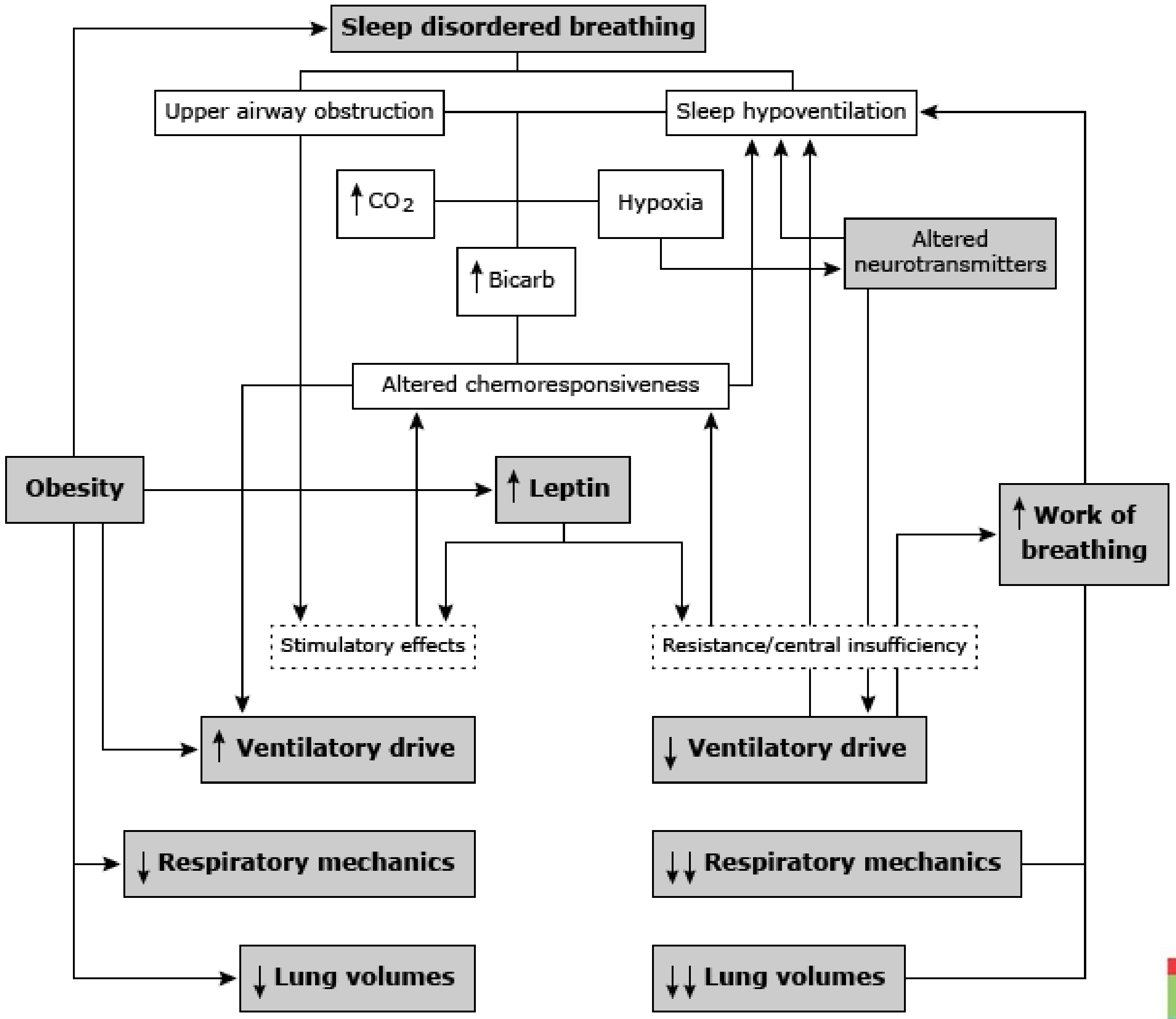
- ✓ Elevated serum bicarbonate (>27 mEq/L)
- ✓ Hypoxemia (PaO_2 <70 mmHg)
- ✓ Polycythemia

Medical impairments associated with obesity hypoventilation syndrome

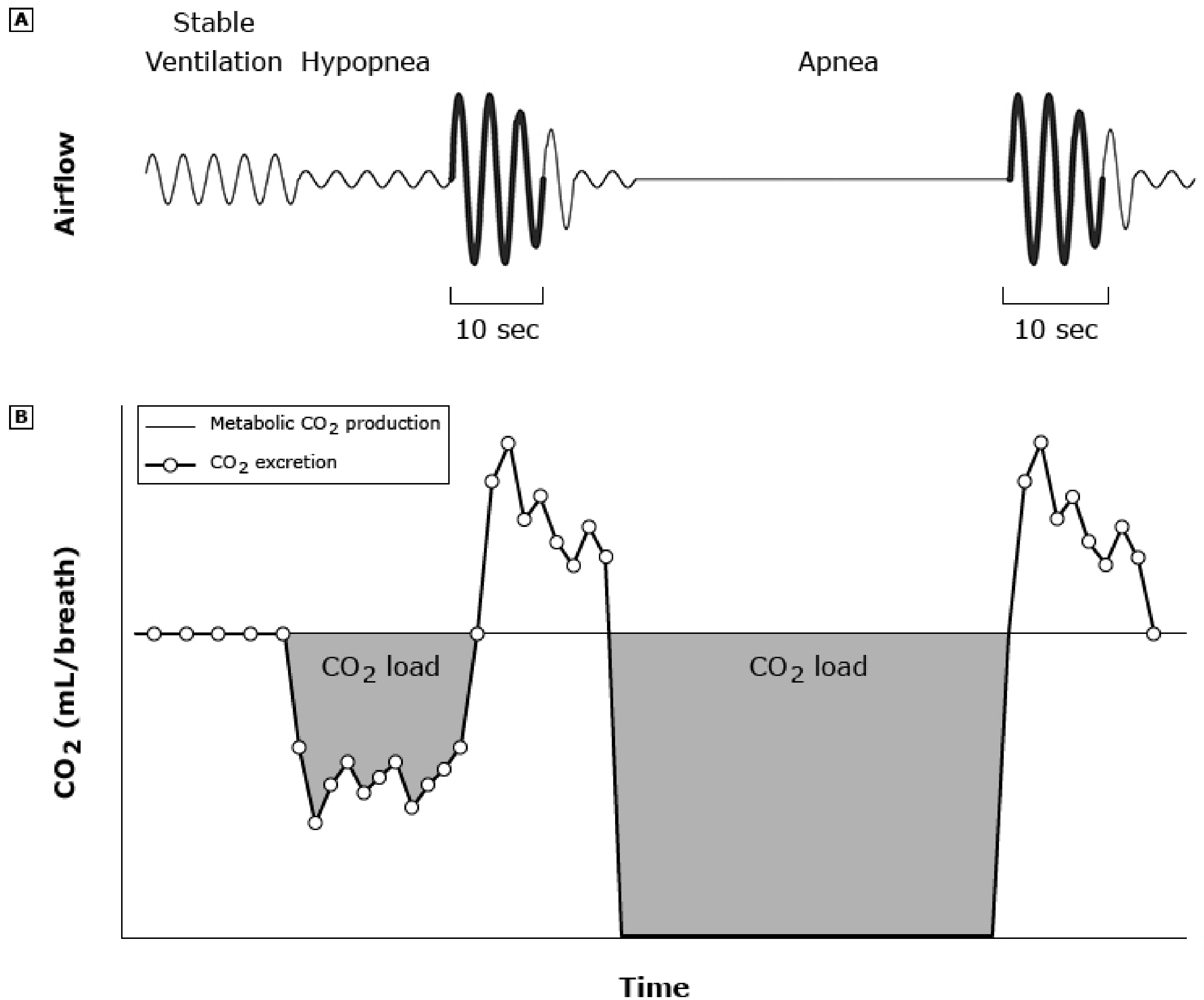
Central nervous system
▪ Cognitive deficit
▪ Decrease neuronal drive
Upper airway
▪ Obstructive sleep apnea
▪ Increased intubation risk
Respiratory
▪ Restrictive lung function
▪ Pulmonary hypertension
▪ Hypercapnia/hypoxemia
Metabolic
▪ Central obesity
▪ Metabolic syndrome
▪ Chronic inflammation
▪ IGF-1 deficit
Cardiovascular
▪ Endothelial dysfunction
▪ Coronary artery disease
▪ Chronic heart failure
General
▪ Peripheral edema
▪ Increased morbi-mortality
▪ Decreased physical activity



Mechanisms of sleep disordered breathing in obesity hypoventilation



Role of obstructive events and carbon dioxide accumulation in obesity hypoventilation syndrome



AMERICAN THORACIC SOCIETY DOCUMENTS

Evaluation and Management of Obesity Hypoventilation Syndrome
An Official American Thoracic Society Clinical Practice Guideline

Babak Mokhlesi, Juan Fernando Masa, Jan L. Brozek, Indira Gurubhagavatula, Patrick B. Murphy, Amanda J. Piper, Aiman Tulaimat, Majid Afshar, Jay S. Balachandran, Raed A. Dweik, Ronald R. Grunstein, Nicholas Hart, Roop Kaw, Geraldo Lorenzi-Filho, Sushmita Pamidi, Bhakti K. Patel, Susheel P. Patil, Jean Louis Pépin, Israa Soghier, Maximiliano Tamae Kakazu, and Mihaela Teodorescu; on behalf of the American Thoracic Society Assembly on Sleep and Respiratory Neurobiology

This official Clinical Practice Guideline of the American Thoracic Society was approved May 2019

(August 2019)

Table 1. Summary of Recommendations

Recommendation	Explanations and Other Considerations
Question 1: Should serum bicarbonate (HCO_3) and/or Sp_{O_2} rather than Pa_{CO_2} be used to screen for OHS in obese adults with sleep-disordered breathing?	
Recommendation 1A: For obese patients with sleep-disordered breathing with a high pretest probability of having OHS, we suggest measuring Pa_{CO_2} rather than serum bicarbonate or Sp_{O_2} to diagnose OHS (*conditional recommendation, very low level of certainty in the evidence*).	Patients with a high pretest probability of having OHS are usually severely obese with typical signs and symptoms of OHS and can be mildly hypoxemic during wake and/or significantly hypoxemic during sleep. This is a recommendation for screening for OHS in patients with sleep-disordered breathing, most typically OSA.
Recommendation 1B: For patients with low to moderate probability of having OHS (<20%), we suggest using serum bicarbonate level to decide when to measure Pa_{CO_2}: in patients with serum bicarbonate <27 mmol/L, clinicians might forego measuring Pa_{CO_2}, as the diagnosis of OHS in them is very unlikely; in patients with serum bicarbonate ⩾27 mmol/L, clinicians might need to measure Pa_{CO_2} to confirm or rule out the diagnosis of OHS (*conditional recommendation, very low level of certainty in the evidence*).	Using a 27-mmol/L threshold in serum bicarbonate in obese patients with OSA and low to moderate clinical suspicion of OHS (initial probability of OHS not more than 20%) would likely permit forgoing further testing, such as arterial blood gases, in those with bicarbonate level <27 mmol/L (64–74% of obese patients with OSA) and performing arterial blood gas analysis only in those with serum bicarbonate ⩾27 mmol/L (26–36% of obese patients with OSA). We found insufficient evidence for serum bicarbonate thresholds other than 27 mmol/L.
Recommendation 1C: We suggest that clinicians avoid using Sp_{O_2} during wakefulness to decide when to measure Pa_{CO_2} in patients suspected of having OHS until more data about the usefulness of Sp_{O_2} in this context become available *(conditional recommendation, very low level of certainty in the evidence).*	We found insufficient data to investigate the clinical usefulness of any threshold of awake Sp_{O_2} for screening for OHS in obese patients with OSA. Guideline panel members believed that relevant studies have to be done before the clinical usefulness of awake Sp_{O_2} in this context can be assessed. This is a temporary recommendation reflecting lack of evidence about a potentially useful intervention, rather than evidence that it is not useful. Thus, this recommendation should not be used as an argument against additional research and will likely change once additional data are available.

Question 2: Should adults with OHS be treated with PAP—either CPAP or NIV—or not be treated with PAP?

Recommendation 2: For stable ambulatory patients diagnosed with OHS, we suggest treatment with PAP during sleep (*conditional recommendation, very low level of certainty in the evidence*).

Note: Patients with symptomatic OHS who have significant comorbidities and those with chronic respiratory failure after an episode of acute-on-chronic hypercapnic respiratory failure may particularly benefit from using PAP.

Question 3: Should adults with OHS be treated with CPAP or with NIV?

Recommendation 3: For stable ambulatory patients diagnosed with OHS and concomitant severe OSA (apnea–hypopnea index ⩾ 30 events/h), we suggest initiating first-line treatment with CPAP therapy rather than NIV (*conditional recommendation, very low level of certainty in the evidence*).

More than 70% of patients with OHS also have severe OSA; therefore, this recommendation applies to the majority of patients with OHS who have concomitant severe OSA. However, panel members lacked certainty on the clinical benefits of initiating treatment with CPAP, rather than NIV, in patients with OHS who have sleep hypoventilation without severe OSA.

Question 4: Should hospitalized adults suspected of having OHS, in whom the diagnosis has not yet been made, be discharged from the hospital with or without PAP treatment until the diagnosis of OHS is either confirmed or ruled out?

Recommendation 4: We suggest that hospitalized patients with respiratory failure suspected of having OHS be started on NIV therapy before being discharged from the hospital, until they undergo outpatient workup and titration of PAP therapy in the sleep laboratory, ideally within the first 3 mo after hospital discharge (*conditional recommendation, very low level of certainty in the evidence*).

Note: Discharging patients from the hospital with NIV should not be a substitute for arranging the outpatient sleep study and PAP titration in the sleep laboratory, as soon as it is feasible.

Question 5: Should a weight-loss intervention or no such intervention be used for adults with OHS?

Recommendation 5: For patients with OHS, we suggest using weight-loss interventions that produce sustained weight loss of 25–30% of actual body weight. This level of weight loss is most likely required to achieve resolution of hypoventilation (*conditional recommendation, very low level of certainty in the evidence*).

Note: Many patients may not be able to achieve this degree of sustained weight loss despite participating in multifaceted comprehensive weight-loss lifestyle intervention program; those who have no contraindications may benefit from being evaluated for bariatric surgery.

Definition of abbreviations: CPAP = continuous positive airway pressure; NIV = noninvasive ventilation; OHS = obesity hypoventilation syndrome; OSA = obstructive sleep apnea; PAP = positive airway pressure; Sp_{O_2} = oxygen saturation by pulse oximetry.

Advantages and disadvantages of the different methods for administering positive airway pressure

Mode of positive pressure ventilation	Advantages	Disadvantages
CPAP	Inexpensive	Lack of inspiratory pressure support
	Widely available	
Bi-level	Widely available	Tidal volume may may be limited by patient-related factors
	Can provide inspiratory pressure support to augment tidal volume	
	Leak tolerant	
Volume-cycled	Can set specific respiratory parameters	More expensive
		Less widely available
		Less well-tolerated than pressure support devices
		Leaks lead to loss of tidal volume

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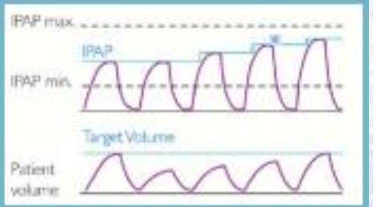
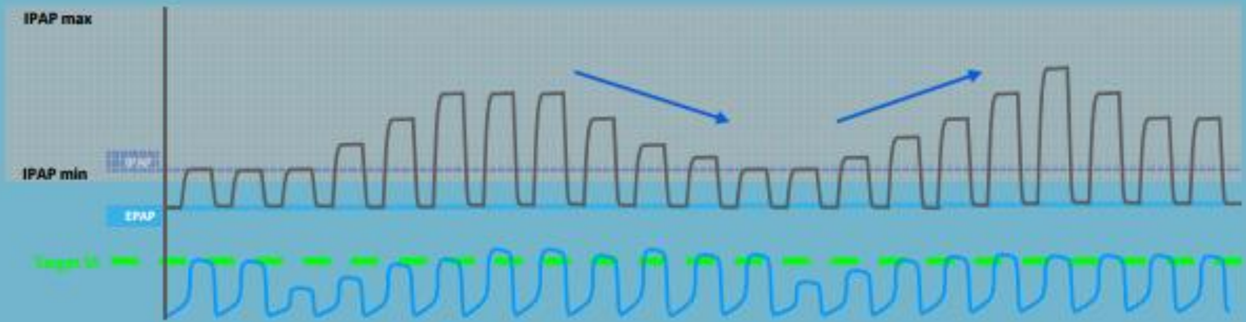
Average Volume Assured Pressure Support

- Modalità ibrida che fornisce il volume corrente costante ad ogni respiro offrendo allo stesso tempo il comfort e i vantaggi della ventilazione a pressione di supporto *
- Progettata per applicare il supporto minimo di pressione necessaria per raggiungere il volume corrente impostato
    - Aiuta a mantenere il comfort ottimale del paziente, sostenendo la cura e la ventilazione del paziente, semplificando il processo di titolazione.

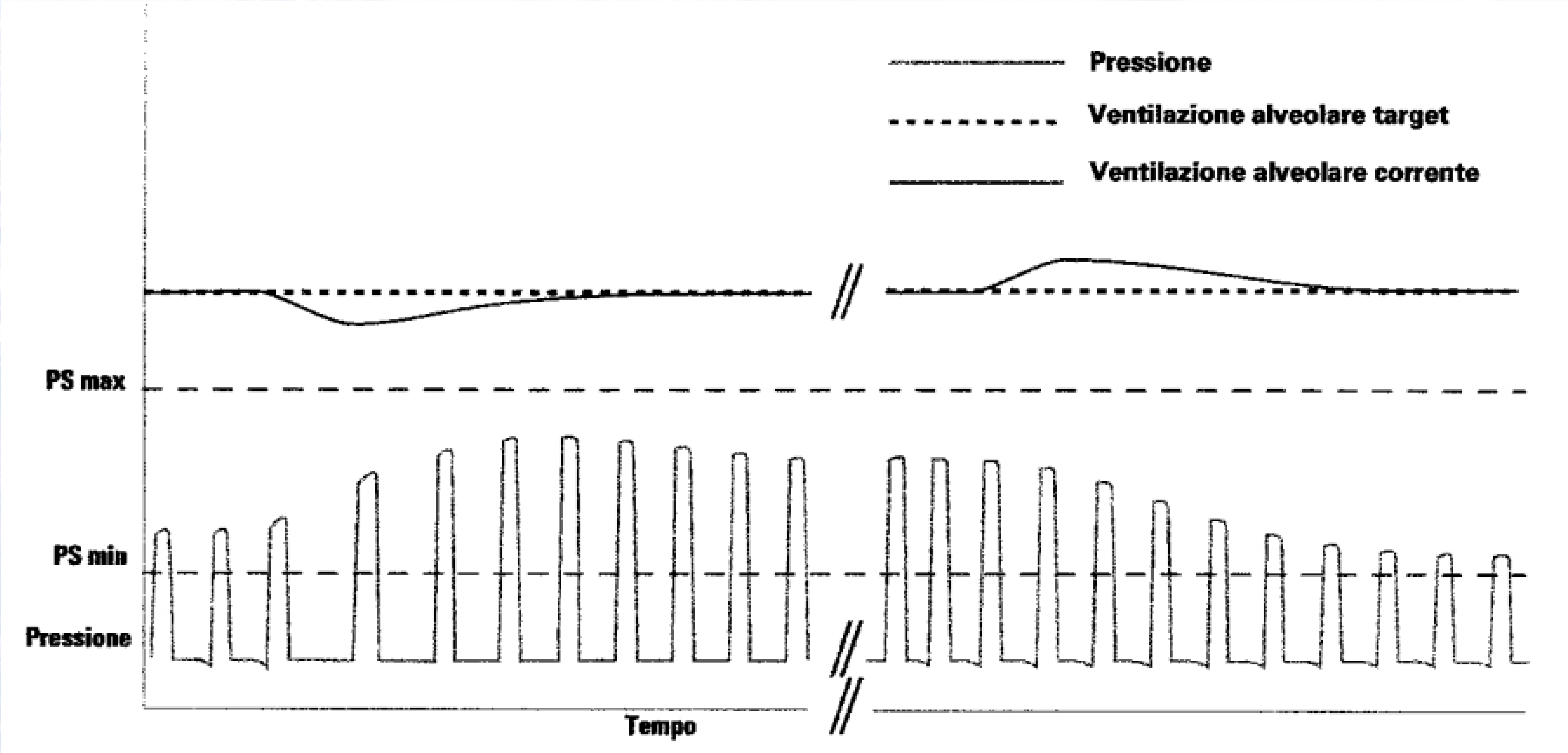
AVAPS

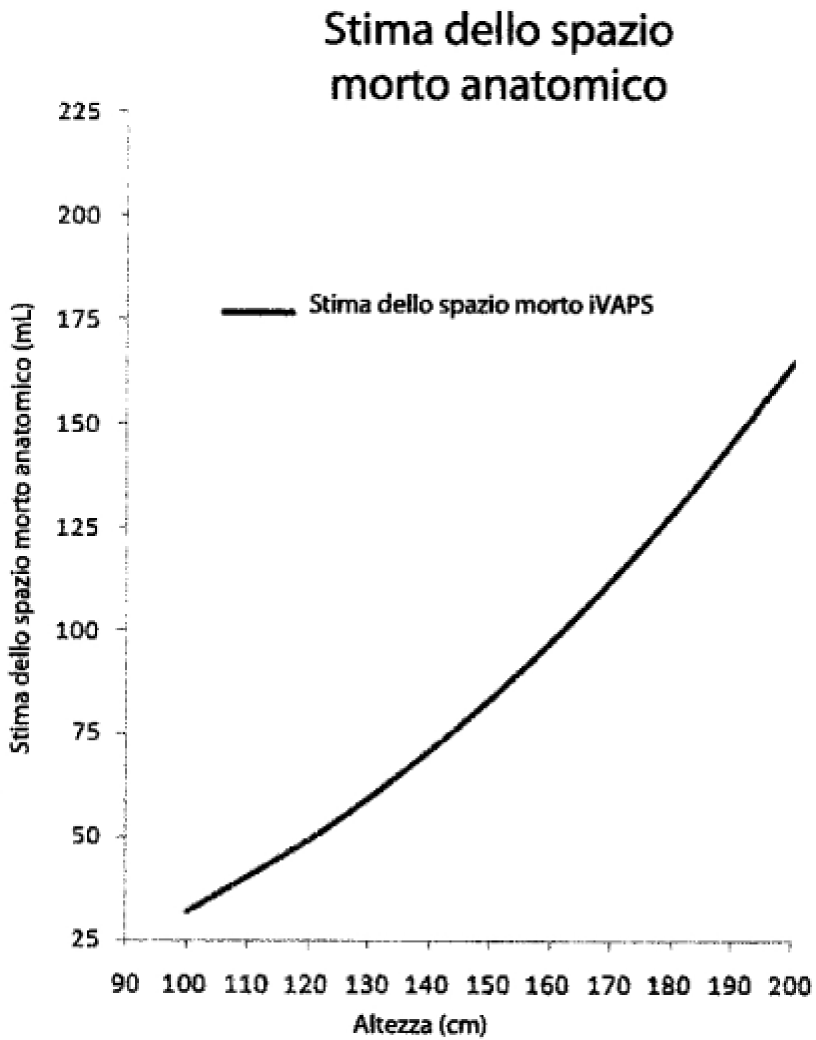
L'algoritmo AVAPS adatta automaticamente la pressione di supporto al paziente per garantire un volume medio assicurato di ventilazione, controbilanciando il carico di lavoro ventilatorio dovuto alla posizione del corpo, agli stadi del sonno e a tutta la meccanica respiratoria

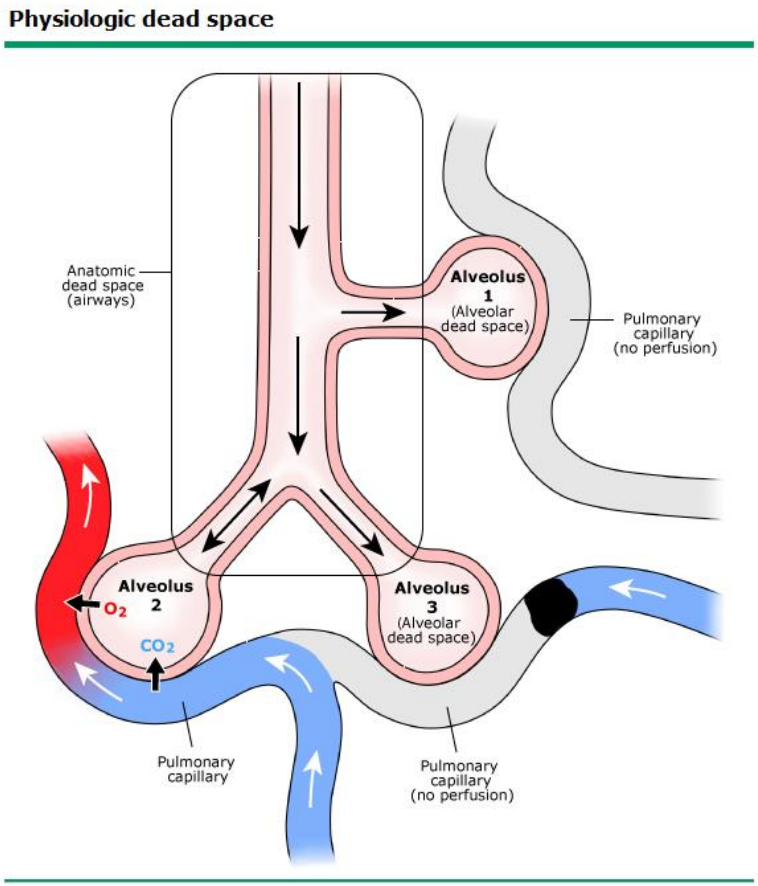




AVAPS
(Pressione di supporto a Volume assicurato/garantito)

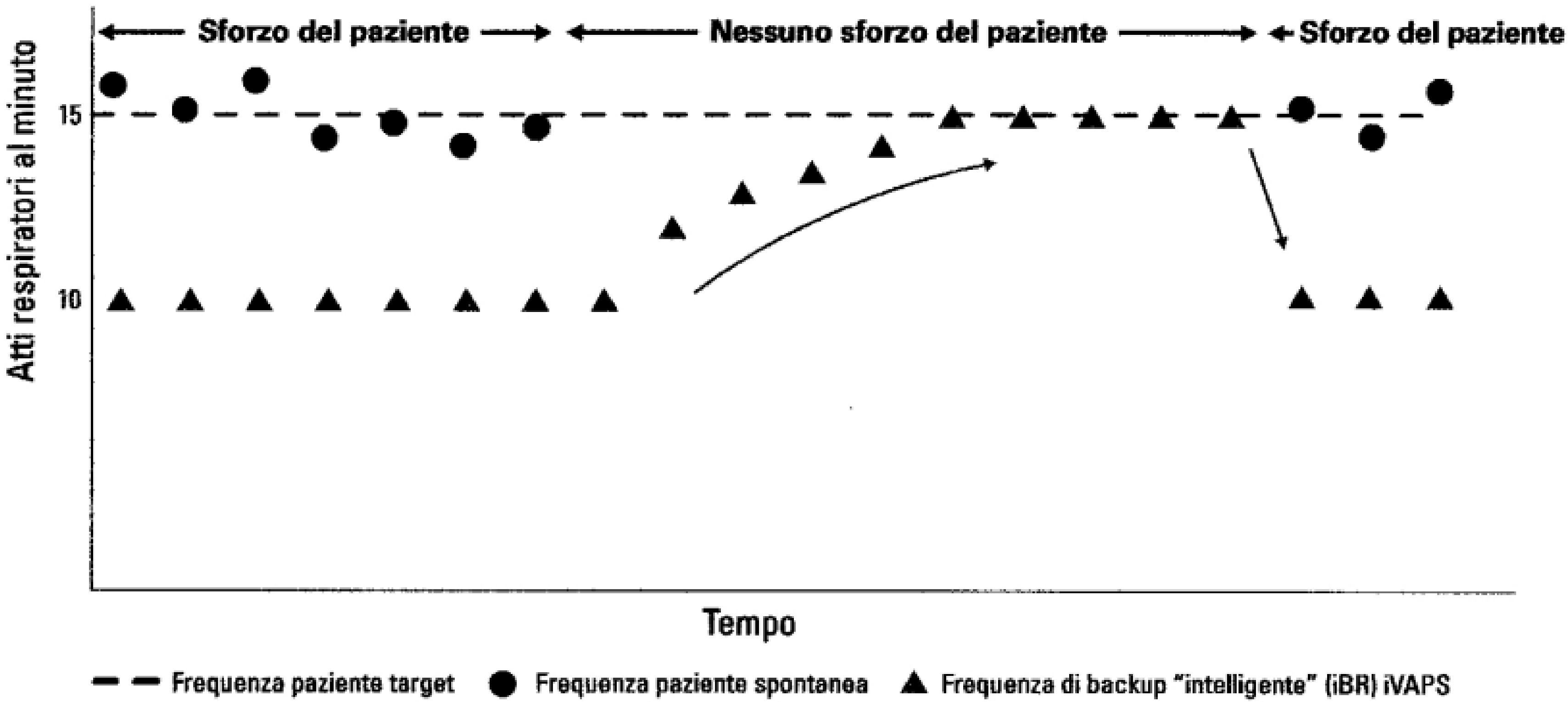






Adattato da Hart MC et al. Journal Applied Physiology.18(3), p519-522. 1963

La funzione iBR riporta il paziente ai livelli definiti quando è necessaria la respirazione di backup



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Original Article

Sleep in ventilatory failure in restrictive thoracic disorders. Effects of treatment with non invasive ventilation

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Table 2
Diagnoses of patients with restrictive disorders.

Diagnoses	N	%
Chest wall disorders	17	28
Congenital myopathy	9	15
Amyotrophic lateral sclerosis	8	13
Obesity-hypoventilation syndrome	6	10
Myotonic muscular dystrophy	6	10
Duchenne muscular dystrophy	4	7
Diaphragmatic paralysis	4	7
Sequelae of tuberculosis	3	5
Sequelae of polyomyelitis	2	3
Ondine's Curse	1	2

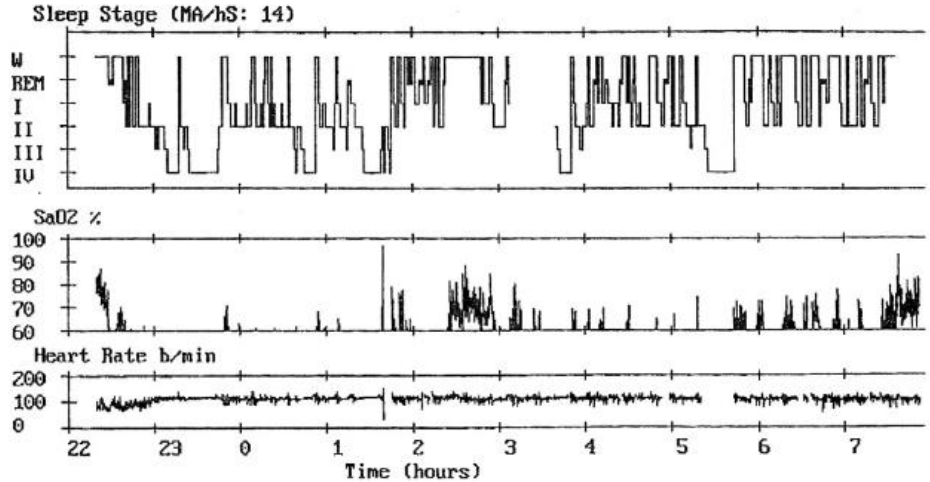


Fig. 1. Summary of full night polysomnography (10 h recording) in a patient with a severe restrictive respiratory disorder and ventilatory failure. Upper tracing: hypnogram; in parentheses: movement arousals per hour of sleep. W: wakefulness; REM: rapid-eye movement sleep; I–IV: non-REM sleep stages 1–4. Middle tracing: transcutaneous oxygen saturation (SpO_2). Lower tracing: pulse rate. Note severe alteration of sleep architecture and very low SpO_2 during most of the sleep study.

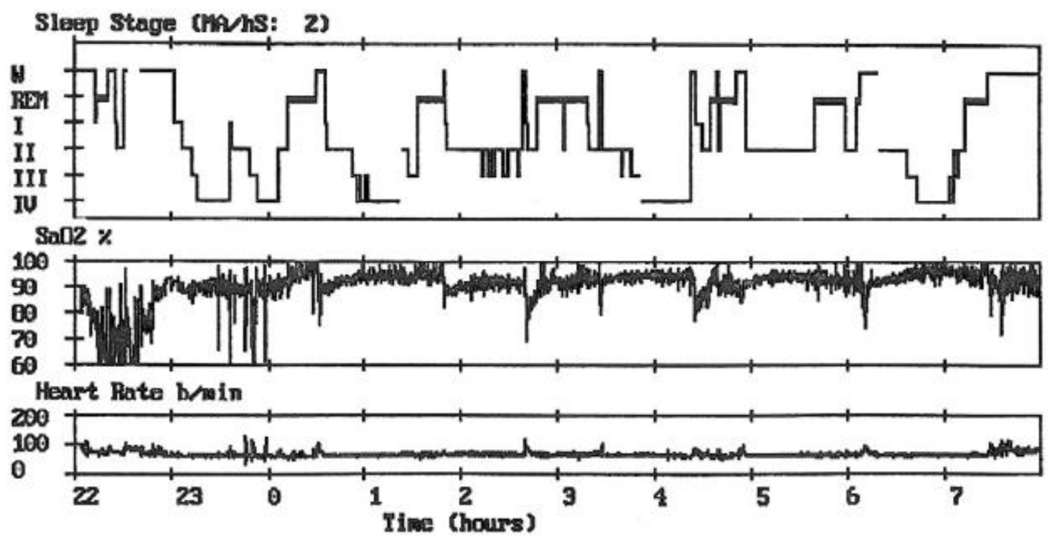


Fig. 2. Summary of full night polysomnography performed in the same patient as in Fig. 1 , four days after non invasive ventilation was initiated.

Guidelines for selecting patients with chronic respiratory failure for noninvasive nocturnal ventilation

Chronic hypoventilation
Daytime $PaCO_2$ ≥45 mmHg **OR**
Nocturnal hypoventilation with sustained O2 desaturation (eg, O2 saturation below 88 mmHG for more than 5 consecutive minutes) **AND** symptoms (eg, morning headache, hypersomnolence, etc)
Appropriate diagnosis
Slowly progressive neuromuscular disease - amyotrophic lateral sclerosis, muscular dystrophy, etc
Chest wall deformity
Reversible contributing factors treated - heart failure, obstructive sleep apnea, etc
Adequate upper airway function

UpToDate®

Mode Variables

Variable	Common Options	Typical Settings
Trigger	Time Triggered	To provide RR of 14-20
	Flow Triggered	2-3 L/min
	Pressure Triggered	0.5 – 2 cm H_2O
Control	Flow Controlled	Indirectly set by selecting V_T (6–10 mL/kg), RR, and I:E ratio.
	Pressure Controlled	To provide V_T of 6-8mL/kg
Cycling	Flow Cycled	< 25 % peak flow
	Volume Cycled	To provide V_T of 6–10mL/kg
	Time Cycled	To provide RR of 14-20

Protocol for initiation of noninvasive positive pressure ventilation in patients with neuromuscular and chest wall disease in the chronic setting

Choose location*

↓

Ensure patient is comfortable in a chair or bed (>30º angle)

↓

Select and fit mask¶

↓

Apply headgear, avoiding excessive strap tension (one or two fingers should fit under the straps)

↓

Connect interface to ventilator tubing and turn on the ventilator$^{\Delta}$

↓

Begin titration

Pressure-cycled settings (in spontaneously-triggered [S/T] mode)

- IPAP: 8 to 10 cm H_2O
- EPAP: 4 to 5 cm H_2O
- Backup rate: 2 bpm below resting respiratory rate

↓

Gradually increase IPAP by 1 to 2 cm H_2O increments, as tolerated (usual maximum 20 cm H_2O, may exceed if chest wall stiffness), to achieve alleviation of dyspnea, decreased respiratory rate, increased tidal volume (not all devices monitor this), and good patient-ventilator synchrony

↓

- Provide encouragement, reassurance, frequent checks, and adjustments, as needed
- Check for air leaks, adjust straps if needed
- Add a chin strap or switch to oronasal (full face) mask if oral air leak is noted
- Monitor vital signs and oximetry during the trial
- Obtain ABG as needed

Volume-cycled settings (in synchronized mandatory ventilation mode)

- Vt: 6 to 10 mL/kg IBW
- PEEP: 4 to 5 cm H_2O
- Backup rate: 2 bpm below resting respiratory rate

↓

Gradually increase Vt by 25 to 50 mL increments, as tolerated, to achieve alleviation of dyspnea, decreased respiratory rate, target tidal volume (usually 6 to 8 mL/kg IBW, but may need more to compensate for leaks), and good patient-ventilator synchrony

↓

- Provide encouragement, reassurance, frequent checks, and adjustments, as needed
- Check for air leaks, adjust straps if needed
- Add a chin strap or switch to oronasal (full face) mask if oral air leak is noted
- Monitor vital signs and oximetry during the trial
- Obtain ABG as needed

Average- or intelligent-volume assured pressure support settings◊

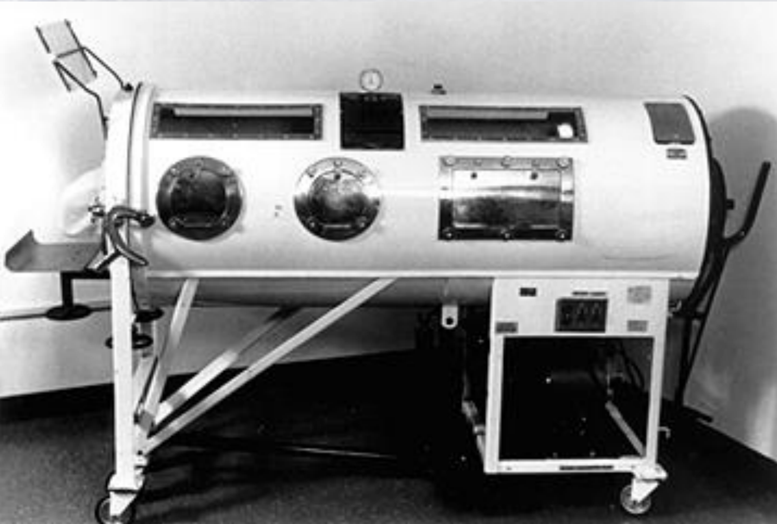
- $IPAP_{min}$: IPAP is gradually increased and set at a target level that results in a tidal volume of 8 mL/kg IBW§
- $IPAP_{max}$: $IPAP_{min}$ + 5 cm H_2O¥
- EPAP: 4 to 5 cm H_2O‡
- Backup rate: 2 to 3 breaths below resting respiratory rate
- Inspiratory time: 33 to 50% (for restrictive lung diseases, shorter for obstructive)
- Rise time: 300 to 600 ms†

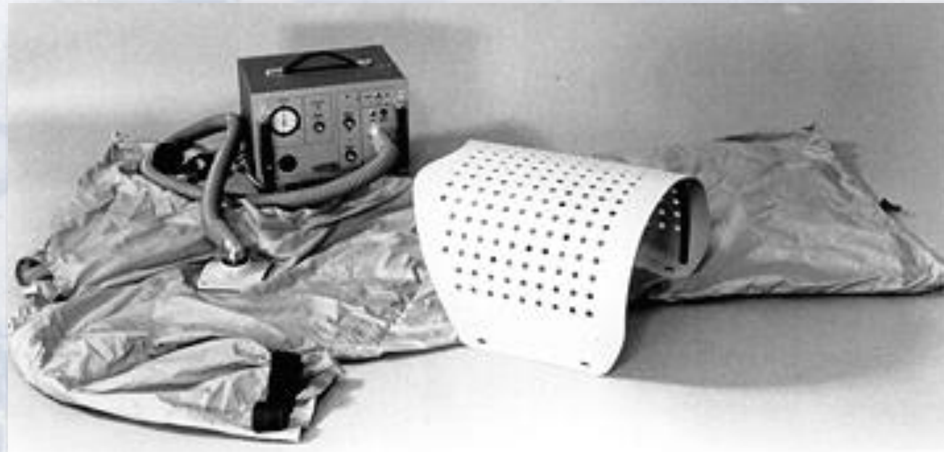
↓

The rise time, inspiratory time, and IPAP and EPAP can be adjusted for patient comfort and to alleviate dyspnea and improve oxygen saturations

↓

- Provide encouragement, reassurance, frequent checks, and adjustments, as needed
- Check for air leaks, adjust straps if needed
- Add a chin strap or switch to oronasal (full face) mask if oral air leak is noted
- Monitor vital signs and oximetry during the trial
- Obtain ABG as needed






Swift LT


Making quality of care easy

Conclusion-1

Despite many previously published studies on non-invasive ventilation (NIV), it is still difficult to draw any conclusions about an optimal ventilation strategy in thoracic restrictive diseases.

Conclusion-2

Restrictive patients are a non-homogenous population where the use of EPAP is different.

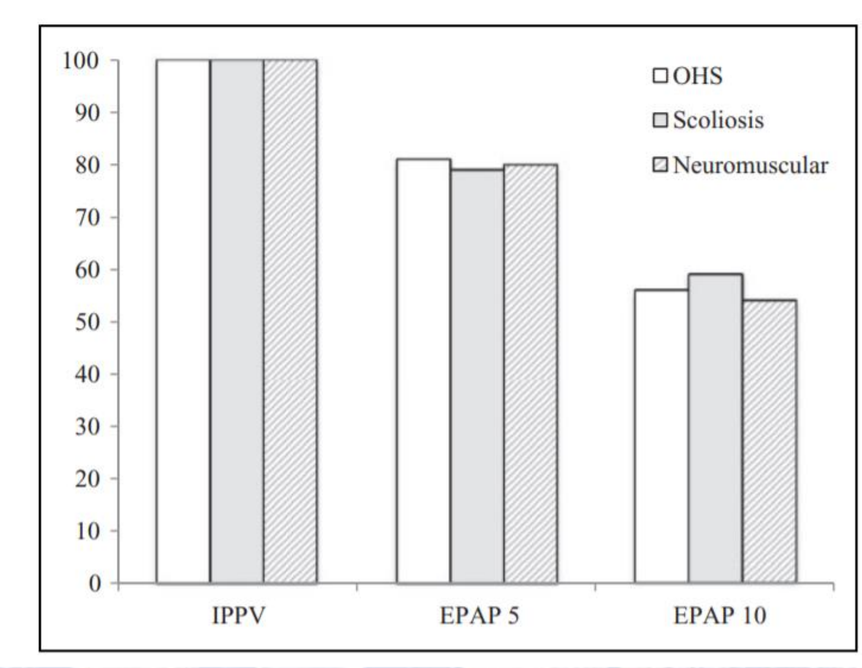
- ✓ *EPAP settings of less than **5 cm H2O** are typically used in neuromuscular and scoliosis patients to mitigate end-expiratory alveolar collapse and to influence atelectasis.*
- ✓ *In patients with the obesity hypoventilation syndrome, the most common use of EPAP of around **10 cm H2O** overcomes upper airway obstruction.*

Conclusion-3

It has been noted that IPAPs tend to have to be increased in order to achieve an adequate Vt, commonly targeted at around 10 ml/kg ideal body weight (IBW).

In these restrictive diseases, there is not a clear direct and solid associations with Vt, positive pressure setting and IBW. The assessment of IBW in the neuromuscular or kyphoscoliosis patient is different from highly obese patient.

Decreasing EPAP is an alternative to increasing IPAP if measurements of gas exchange during NIV indicate that ventilation is inadequate




Grazie per l’attenzione