ΕΛΕΝΗ ΒΑΒΟΥΡΑΚΗ
ΠΝΕΥΜΟΝΟΛΟΓΟΣ

ΠΝΕΥΜΟΝΟΠΑΘΕΙΕΣ ΠΕΡΙΟΡΙΣΤΙΚΟΥ ΤΥΠΟΥ

Αιτίες διαταραχής αερισμού περιοριστικού τύπου

- Απώλεια πνευμονικού όγκου(π.χ. λοβεκτομή)
- Εξωπαρεγχυματικές αιτίες (π.χ. παθήσεις υπεζωκότα,κυφοσκολίωση, παχυσαρκία)
- Παρεγχυματικές αιτίες(π.χ.πνευμονική ίνωση)
- Αδυναμία εισπνευστικών μυών (π.χ. νευρομυικές παθήσεις)

Οι πνευμονοπάθειες περιοριστικού τύπου χαρακτηρίζονται από:

- Μείωση της ευενδοτότητας (compliance) των πνευμόνων, του θώρακα ή και των δυο
- Οι αναπνευστικοί μυς, κυρίως το διάφραγμα, καθώς και οι επικουρικοί, δουλεύουν περισσότερο για να επιτευχθεί καλύτερη έκπτυξη του σκληρού ανελαστικού πνεύμονα ή/και θώρακα ή και των δυο
- Το αναπνευστικό έργο αυξάνεται για να υπερνικήσει την μειωμένη ευενδοτότητα.

Οι πνευμονοπάθειες περιοριστικού τύπου χαρακτηρίζονται επίσης από:

- Μείωση πνευμονικών όγκων και χωρητικοτήτων
- Διαταραχές διάχυσης και παθολογικές τιμές αερίων αίματος

Προσοχή!

- Στους ασθενείς με πνευμονοπάθεια περιοριστικού τύπου η άσκηση μπορεί να έχει σημαντική μείωση της $Pa\,O_2$, ακόμα και σε αυτούς που έχουν φυσιολογικές τιμές στην ηρεμία.

Λειτουργική δοκιμασία σε πνευμονοπάθειες αποφρακτικού,περιοριστικού και μικτού τύπου

Measure	Obstructive Disorders	Restrictive Disorders	Mixed Disorders
FEV_1/FVC	Decreased	Normal or increased	Decreased
FEV_1	Decreased	Decreased, normal, or increased	Decreased
FVC	Decreased or normal	Decreased	Decreased or normal
TLC	Normal or increased	Decreased	Decreased, normal, or increased
RV	Normal or increased	Decreased	Decreased, normal, or increased

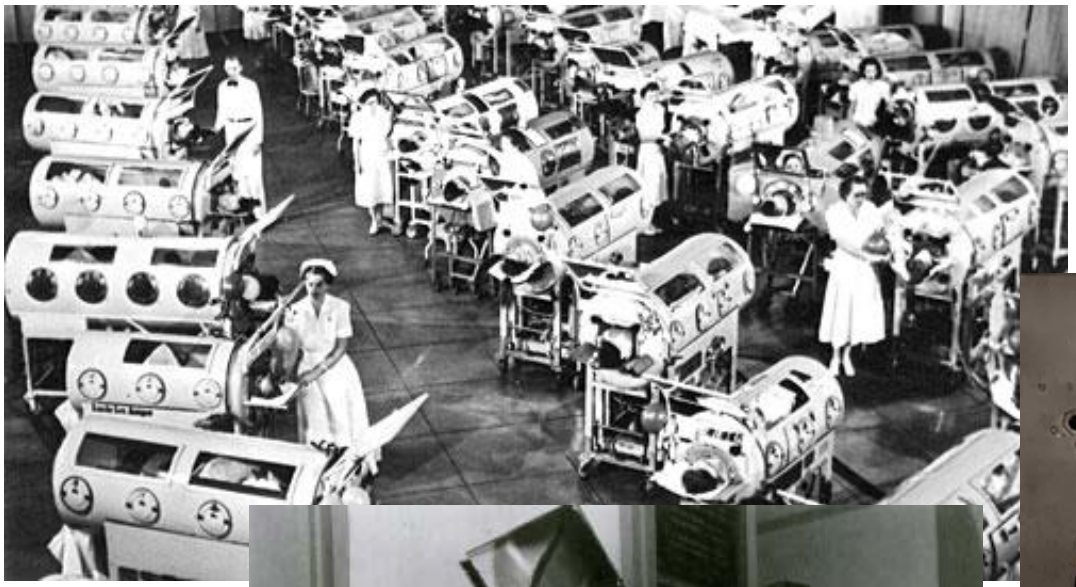
Ταξινόμηση βαρύτητας πνευμονοπαθειών αποφρακτικού και περιοριστικού τύπου

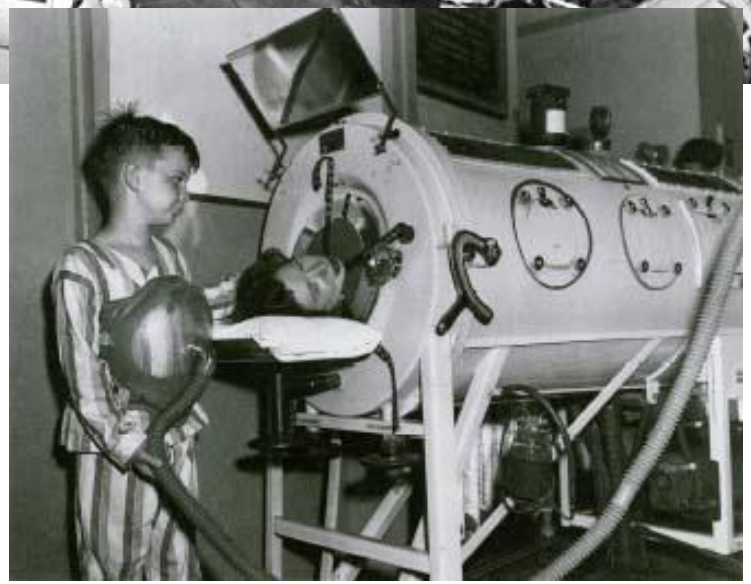
		Obstructive	Restrictive
Severity†	FEV_1/FVC (% predicted)	FEV_1 (% predicted)	TLC (% predicted)
Normal	≥ 70	≥ 80	≥ 80
Mild	< 70	≥ 80	70–79
Moderate	< 70	50 ≤ FEV_1 < 80	50–69
Severe	< 70	30 ≤ FEV_1 < 50	< 50
Very severe	< 70	< 30 *or* < 50 with chronic respiratory failure	—

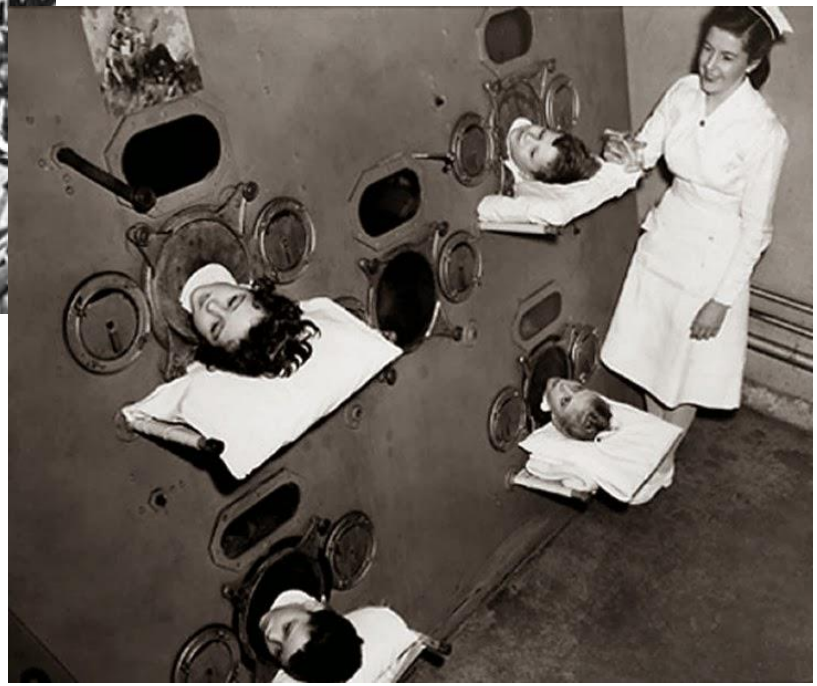
Κλινική εικόνα

- Δύσπνοια :αρχικά στην προσπάθεια και όσο η νόσος εξελίσσεται και στην ηρεμία
- Βήχας, συνήθως μη παραγωγικός
- Αδυναμία, κόπωση
- Ταχύπνοια με ρηχές αναπνοές
- Περιορισμένη θωρακική έκπτυξη
- Αποκλίσεις στη στάση του σώματος
- Πληκτροδακτυλία ,κυάνωση (σε προχωρημένα στάδια)

Παρότι οι μεγάλες επιδημίες πολιομυελίτιδας τον περασμένο αιώνα έγιναν αιτία μεγάλου αριθμού αναπηριών με σύνδρομα διαταραχής αερισμού περιοριστικού τύπου, εντούτοις τα πρωτόκολλα πνευμονικής αποκατάστασης κυρίως αναφέρονται στη ΧΑΠ.







Σήμερα, η συνήθης φ/θ αντιμετώπιση των διαταραχών περιοριστικού τύπου περιλαμβάνει

- Διδασκαλία διαφραγματικής αναπνοής
- Ενδυνάμωση διαφράγματος
- Βαθιές διαφραγματικές αναπνοές με κράτημα της αναπνοής
- Χρήση εξασκητών αναπνοής για τη βελτίωση της εισπνευστικής ικανότητας
- Ασκήσεις ενδυνάμωσης άνω και κάτω άκρων
- Αεροβική άσκηση (στατικό ποδήλατο, περπάτημα, ανέβασμα σκάλας)
- Ειδικό πρόγραμμα ασκήσεων ανάλογα με την υποκείμενη νόσο (π.χ. κυφοσκολίωση)

Karvonen method

Επειδή μέχρι σήμερα δεν έχει καθοριστεί η επιζητούμενη καρδιακή συχνότητα στους ασκούμενους με πνευμονοπάθεια περιοριστικού τύπου, ακολουθείται η μέθοδος Karvonen για τον υπολογισμό της,ως ασφαλής για ηλικιωμένους και ασθενείς με καρδιοαναπνευστικό νόσημα.

- Target HR = (220 - age - resting heart rate × % intensity selected) + resting heart rate

British Thoracic Society & Association of Chartered Physiotherapists in Respiratory Care

http://www.britthoracic.org.uk/ClinicalInformation/Physiotherapy/PhysiotherapyGuideline/tabid/375/Default.aspx

Οι παραπάνω επιστημονικές ενώσεις διαπίστωσαν ότι….

"*Restrictive lung conditions*

- *There is a paucity of evidence on physiotherapy for these conditions."*

και δημοσίευσαν τις πρώτες Κατευθυντήριες Οδηγίες στις οποίες αναφέρονται και στην άσκηση

Thorax 2009;64(Suppl I):i1–i51. doi:10.1136/thx.2008.110726

Lung fibrosis

- All patients with chronic restrictive conditions, such as pulmonary fibrosis, should be considered for pulmonary rehabilitation. (Grade B)

Neuromuscular diseases and musculoskeletal disorders of the chest wall- Chest wall disorders

Pulmonary rehabilitation and ambulatory oxygen

- Offer patients with chest wall restriction post-tuberculosis pulmonary rehabilitation. (Grade B)
- Offer patients with chest wall deformity from other causes, who have reduced exercise capacity and/or breathlessness on exertion, pulmonary rehabilitation. (Grade C)
- Assess patients with moderate to severe kyphoscoliosis who desaturate on exercise for ambulatory oxygen. (Grade D)

Neuromuscular diseases and musculoskeletal disorders of the chest wall- Chest wall disorders

Respiratory muscle training and breathing exercises

- Consider respiratory muscle training in patients with kyphoscoliosis. (Grade D)

Spinal cord injuries

Monitoring

- Monitor the patient with spinal cord injury for the signs and symptoms of respiratory problems and take appropriate action if abnormal or changing. (Grade A)
- Measure vital capacity routinely in the patient with upper spinal cord injury and take appropriate action if falling.(Grade D)
- Alert medical staff if vital capacity falls to 1 litre or less.(Grade D)

Spinal cord injuries

Positioning

- Consider the supine position to maximise vital capacity.(Grade B)
- Assess the head-up 30° position for improving pulmonary function. (Grade C)
- The head-down position should only be used where there is a demonstrable need and only with extreme caution. (Grade D)
- Any patient, especially those with early spinal cord injury,should be carefully monitored for signs of hypoxaemia in head-down positions. (Grade D)
- Take comorbidities and contraindications and precautions to head-down tilt positions into account. (Grade D)

Spinal cord injuries

Abdominal binders

- Assess the effect of an abdominal binder for upright sitting where improvement in either vital capacity or respiratory muscle function is required. (Grade D)
- Patients using non-elastic binders should be monitored closely. (Grade D)
- When using an abdominal binder, the optimal position for the individual patient should be determined. (Grade D)

Spinal cord injuries

Management of cough and airway secretions

Assisted coughing

- Try manually assisted coughing for patients with an ineffective cough. (Grade D)
- The upright seated position should be considered initially.(Grade D)
- The abdominal thrust (Heimlich-style manoeuvre) should be considered initially. (Grade D)

Spinal cord injuries

Management of cough and airway secretions

Mechanical insufflation–exsufflation

- Mechanical insufflation–exsufflation should be considered for individuals with upper spinal cord injury, if simpler techniques fail to produce an adequate effect.(Grade D)

- Where cough effectiveness remains inadequate with mechanical insufflation–exsufflation alone, combine it with manually assisted coughing. (Grade D)

Spinal cord injuries

Functional electrical stimulation

Consider electrical stimulation of the abdominal muscles as a possible means of enhancing lung volumes and cough effectiveness. (Grade C)

Exercise

Active exercise should be encouraged in patients confined to a wheelchair as a result of spinal cord injury. (Grade D)

Spinal cord injuries

Breathing exercises

Deep breathing exercises should be encouraged in patients with spinal cord injury. (Grade D)

Exercise

Active exercise should be encouraged in patients confined to a wheelchair as a result of spinal cord injury. (Grade D)

Spinal cord injuries

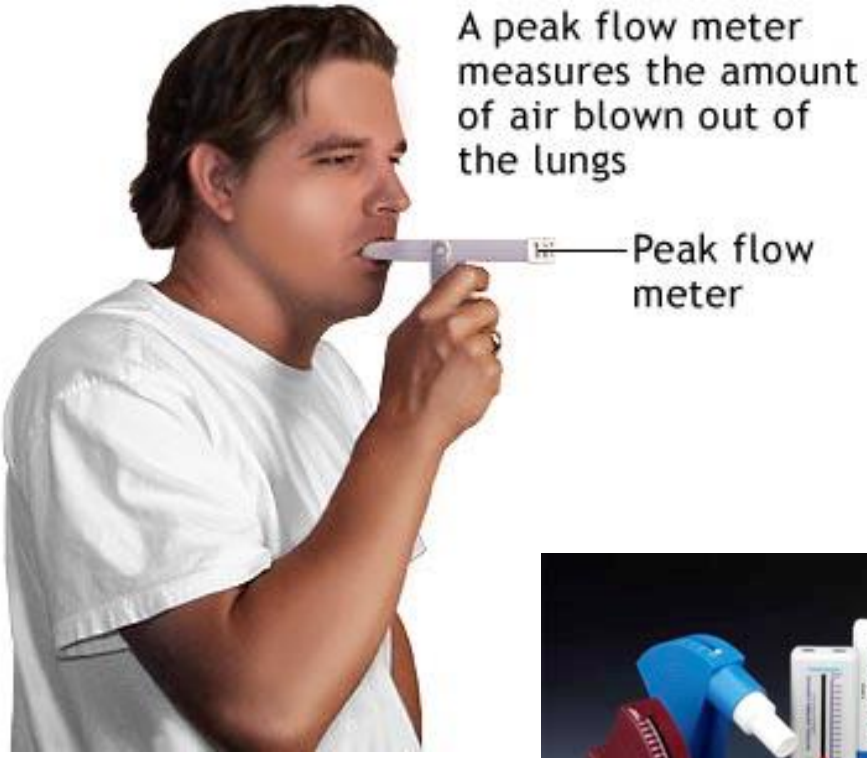
Respiratory muscle training

- Inspiratory muscle training may be considered for patients with upper spinal cord injury to improve respiratory muscle strength. (Grade C)
- Inspiratory muscle training may be considered for patients with upper spinal cord injury to improve vital capacity and residual volume. (Grade C)
- Training of the accessory muscles of respiration with progressive loading should be considered. (Grade D)

Neuromuscular disease

Peak cough flow (PCF) monitoring

- Peak cough flow should be measured regularly in patients with neuromuscular disease. (Grade D)
- Measure peak cough flow additionally at the time of an acute respiratory tract infection. (Grade D)
- When peak cough flow is equal to or less than 270 l/min in a medically stable patient, introduce strategies for assisted airway clearance to raise it above 270 l/min. (Grade D)
- When peak cough flow is equal to or less than 160 l/min, additional strategies to assist secretion clearance must be used. (Grade D)
- If peak cough flow remains equal to or less than 160 l/min despite additional strategies, contact medical colleagues to discuss ventilation and/or airway management needs. (Grade D)


A peak flow meter measures the amount of air blown out of the lungs
Peak flow meter

Neuromuscular disease

Airway clearance techniques

Maximal insufflation capacity

- When oxygen saturation falls below 95% the use of noninvasive ventilation and/or strategies to aid airway clearance should be considered. (Grade D)
- Use some form of maximal insufflation strategy to improve effective cough generation when vital capacity falls below 1500 ml or 50% predicted. (Grade D)
- Use single maximal insufflation techniques for patients with bulbar dysfunction who are unable to breath stack. (Grade D)
- Teach patients without bulbar muscle involvement unaided breath stacking to improve cough effectiveness independently where possible. (Grade D)
- Regular breath stacking (10–15 times three times per day) to maximal insufflation capacity should be performed by patients with vital capacity of less than 2000 ml or 50% predicted. (Grade D)

Neuromuscular disease

Glossopharyngeal breathing

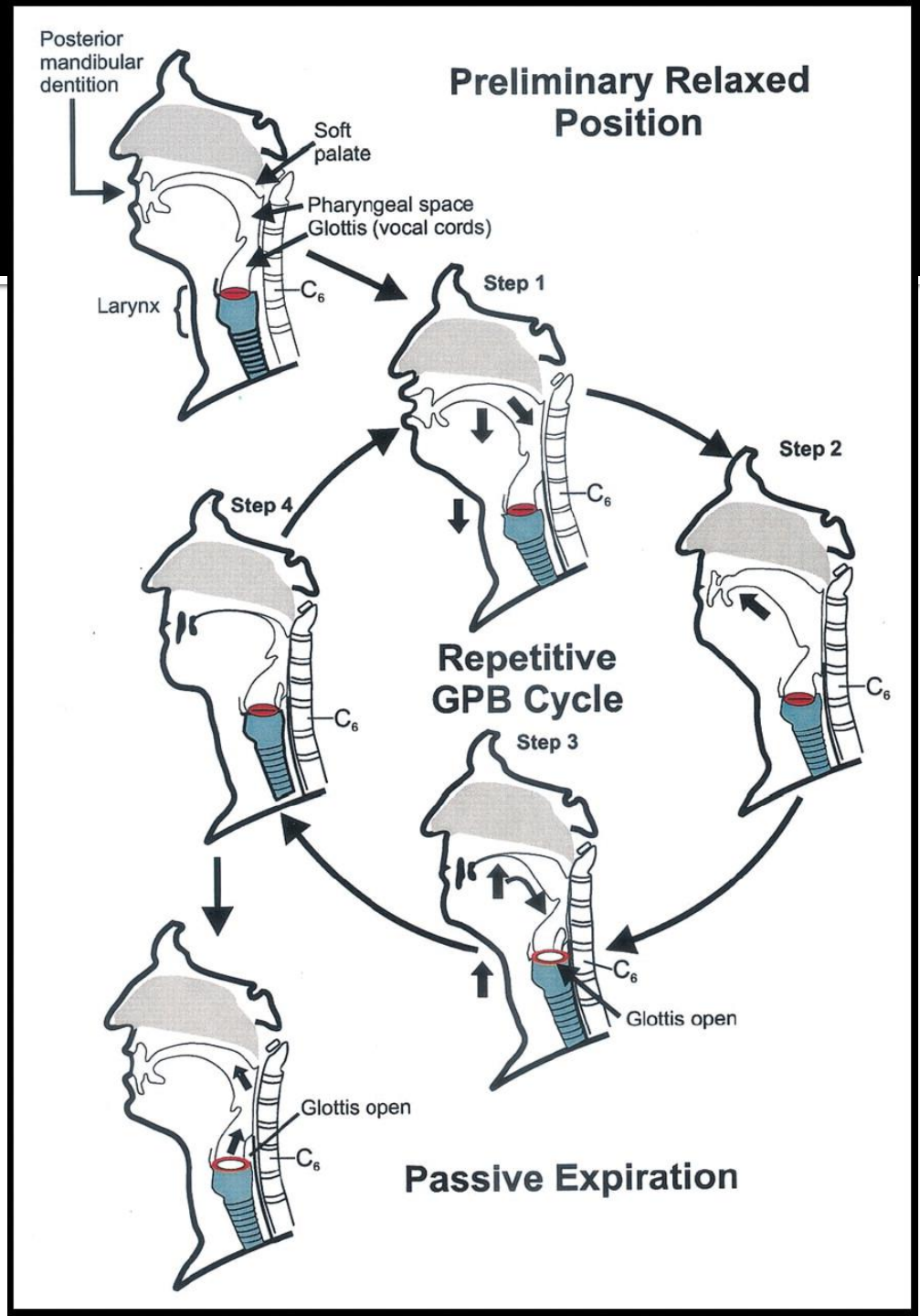
- Consider teaching glossopharyngeal breathing to patients with reduced vital capacity to maintain range of chest wall movement and pulmonary compliance. (Grade D)

- Consider teaching glossopharyngeal breathing as one of the means of achieving maximal insufflation capacity in patients who have difficulty in clearing secretions. (Grade D)

- Consider teaching glossopharyngeal breathing to ventilatordependent patients to allow some ventilator-free breathing
- time. (Grade D)

- Consider teaching glossopharyngeal breathing to patients
- with decreased voice strength. (Grade D)

Glossopharyngeal breathing



Neuromuscular disease

Manually assisted coughing

- Manually assisted coughing should be used to increase peak cough flow in patients with neuromuscular disease. (Grade C)
- Combine manually assisted coughing with a maximal insufflation capacity strategy. (Grade D)
- Abdominal thrusts should be performed standing in front of the patient where possible to assist communication. (Grade D)

Neuromuscular disease

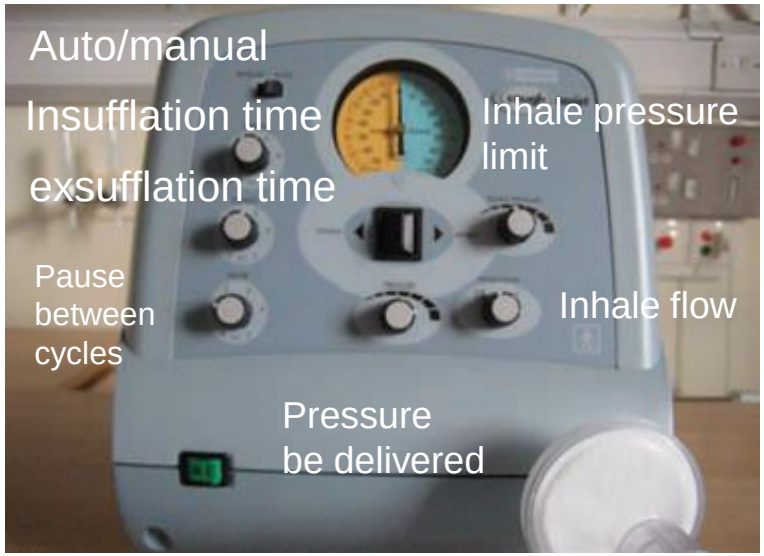
Mechanical insufflation–exsufflation

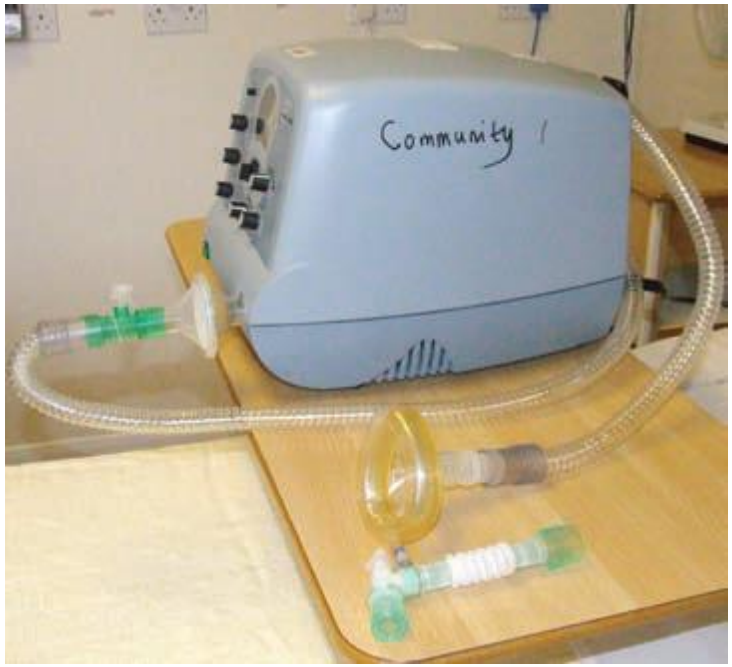
- Consider mechanical insufflation–exsufflation as a treatment option in patients with bulbar muscle involvement who are unable to breath stack. (Grade D)
- Consider mechanical insufflation–exsufflation for any patient who remains unable to increase peak cough flow to effective levels with other strategies. (Grade D)
- Where cough effectiveness remains inadequate with mechanical insufflation–exsufflation alone, combine it with manually assisted coughing. (Grade D)

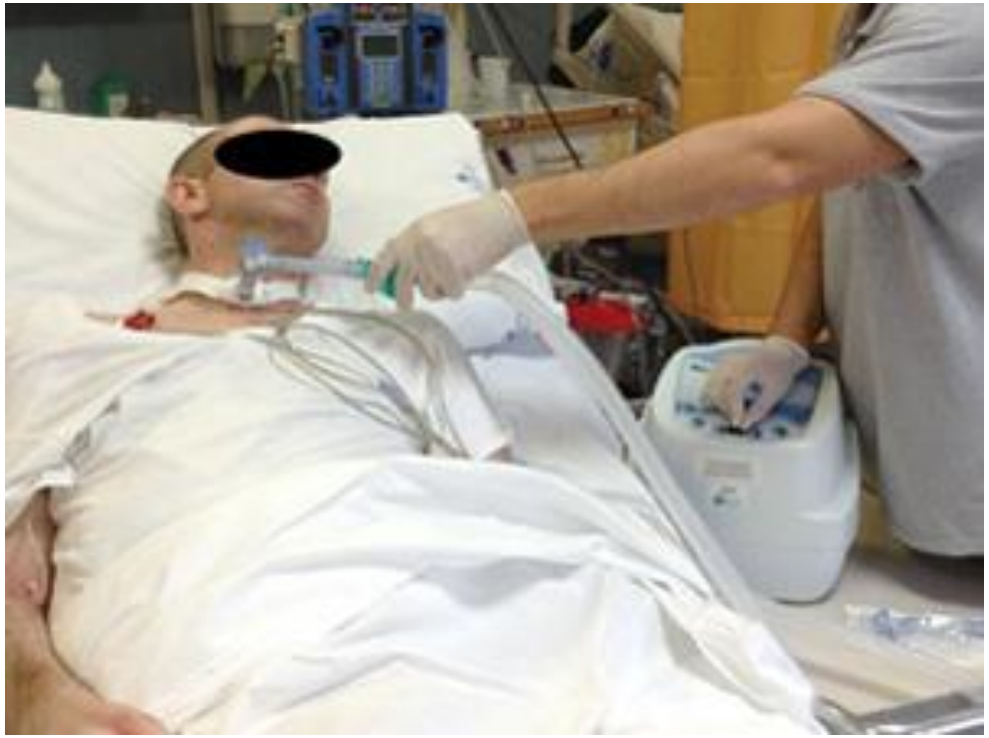
Mechanical insufflation–exsufflation

Ineffective cough is a major cause of mortality and morbidity in patients with neuromuscular disease. Cough assist techniques are used in patients who present with a weak cough. The goal of these techniques is to increase the expiratory airflow that occurs during a cough, by assisting inspiration and/or expiration, thus increasing cough efficacy. When conventional cough assistance techniques become ineffective, a mechanical insufflator–exsufflator should be considered.

mechanical insufflation–exsufflation







Neuromuscular disease

Intrapulmonary percussive ventilation

- Intrapulmonary percussive ventilation may be considered for patients with neuromuscular disease to aid loosening of secretions prior to removal where there is evidence of sputum retention and other techniques have failed. (Grade D)

- In patients with ineffective cough, assisted cough strategies must be used additionally to increase cough effectiveness. (Grade D)

- Patients using intrapulmonary percussive ventilation must be monitored closely during and after treatment for any
- adverse response. (Grade D)

Buschmann et al.
Differential Diagnosis of Restrictive Lung Diseases: Utility of Cardiopulmonary Exercise Testing.
Pneumologie. 2009 Nov 18

- Exercise dyspnea is a common symptom of restrictive lung diseases. Not only from the clinical perspective but also from the pathophysiological point of view, restrictive lung disorders represent a very heterogeneous group of diseases. Exercise testing is mandatory because pulmonary function tests at rest are not reliable for the diagnostic evaluation and functional characterisation of these patients.

Buschmann et al.
Differential Diagnosis of Restrictive Lung Diseases: Utility of Cardiopulmonary Exercise Testing.
Pneumologie. 2009 Nov 18

Cardiopulmonary exercise testing (CPET) with measurement of gas exchange is the favoured tool. It is an excellent method to

- investigate exercise dyspnea,
- describe altered physiological response to exercise and
- characterise the involved organ systems heart, lung and muscle.

Kagaya H et al.
Effective home-based pulmonary rehabilitation in patients with restrictive lung diseases.
Tohoku J Exp Med. 2009 Jul;218(3):215-9.

- Our PR consisted of breathing retraining, exercise training, respiratory muscle stretching calisthenics, level walking, inspiratory and expiratory muscle exercises, and a monthly education program. Patients were strongly instructed to practice this program daily at home, and were supervised by a respiratory therapist every 2 weeks in our hospital.

Kagaya H et al.
Effective home-based pulmonary rehabilitation in patients with restrictive lung diseases.
Tohoku J Exp Med. 2009 Jul;218(3):215-9.

- Patients with restrictive lung diseases showed the significant increases in inspiratory and expiratory muscle forces, the 6-minute walking distance, the Chronic Respiratory Disease Questionnaire and the Short-Form 36 after 6 months.
- In conclusion, our home-based PR improves respiratory muscle forces, exercise tolerance, health-related quality of life, and the perception of dyspnea in patients with restrictive lung disease to the same extent as in COPD patients.

Swallow EB et al
Quadriceps muscle strength in scoliosis.
Eur Respir J. 2009 Dec;34(6):1429-35. Epub 2009 May 14.

- Quadriceps weakness is a feature of severe scoliosis; the similarities between patients with scoliosis and patients with COPD suggest a common aetiology to quadriceps weakness in both conditions.

Bihiyga Salhi et al.
Effects of pulmonary rehabilitation in patients with restrictive lung diseases
Chest.2010.137(2).p.273-279

- In a prospective, nonrandomized, noncontrolled study, patients with an established diagnosis of restrictive lung disease (RLD) participated in a 24-week outpatient multidisciplinary rehabilitation program.
- Pulmonary function, exercise capacity, muscle force, and dyspnea were measured at inclusion, after 12 and 24 weeks of rehabilitation.
- Primary outcome was the change in 6-min walk distance (6MWD) after 12 weeks of rehabilitation.

Bihiyga Salhi et al.
Effects of pulmonary rehabilitation in patients with restrictive lung diseases
Chest.2010.137(2).p.273-279

Results:

- Exercise capacity, muscle force, and dyspnea improved significantly after 12 weeks
- Further improvements were noted after 24 weeks .

Christopher J et al.
Pulmonary Rehabilitation for Interstitial Lung Disease
Chest. 2010;138(1):240-241.

			Change in Selected Outcome[a]		
Study/Year	**No. With ILD**	**Design**	**6MWD, m**	**Dyspnea**	**Quality of Life**
Salhi et al[1]/2010	11	Cohort	107 ($P < .05$)	Improved	NA
Ferreira et al[3]/2009	99	Cohort	56 ($P < .0001$)	Improved	NA
Holland et al[5]/2008	57	RCT	35 ($P = .01$)	Improved	Improved[b]
Nishiyama et al[8]/2008	28	RCT	46 ($P < .01$)	No change	Improved
Ferreira et al[4]/2006	28	Cohort	40 ($P < .0002$)	Improved	Improved
Jastrzebski et al[6]/2006	31	Cohort	NA	Improved[b]	Improved
Naji et al[7]/2006[c]	26	Cohort	NA	Improved	Improved[b]

6MWD = 6-min walk distance; ILD = interstitial lung disease; NA = not assessed; RCT = randomized controlled trial.

Σας ευχαριστώ