Advanced lung support
New devices/New strategies

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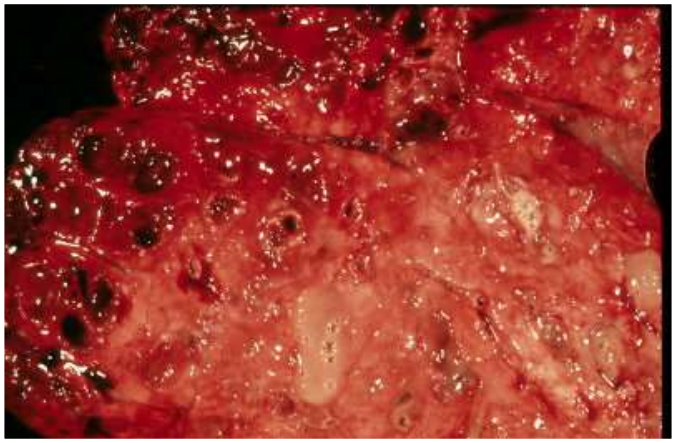
הכנס הארצי לסיסטיק פיברוזיס

דן קיסריה 20 באוקטובר 2017



Excessive mucus production

Chronic inflammation

Advanced lung disease:
What does it look like?

Higher mucous viscosity

Malnutrition

Airway obstruction

Fear of death

Chronic fatigue

Anxiety

Combination of different treatment strategies

Active cycle of breathing techniques and exercise

- Inhalations of bronchodilators and HS
- 2-3 times a day
- Mild - moderate exercise with O_2

PEP/Oscillatory PEP devices

- PEP mask
- Acapella
- Flutter
- Aerobika

Mechanical airway clearance devices

- High Frequency Chest Wall Oscillation (Vest)
- Intrapulmonary Percussion Ventilation (IPV)

NIV

- BiPAP/CPAP
- Biphasic Cuirass Ventilation
- HFNC

Indications for NIV

- Daytime hypercapnia
- nocturnal respiratory failure
- Desaturation and complain of fatigue during chest physiotherapy and exercise
- As a bridge for transplantation
- **Important**: Discuss NIV with patient and family, in advance!

B. Fauroux. Minerva Anestesiologia 2011

Non Invasive Positive Pressure ventilation in CF

- Reduces respiratory muscle work of breathing
- Improves alveolar ventilation and gas exchange during sleep, exercise and chest physiotherapy
- Decreases PaCO2
- During chest physiotherapy: Associated with improvement of saturation and decrease of fatigue

B. Fauroux. Minerva Anestesiologia 2011

NIV and Airway clearance

- Airway clearance may be easier with NIV and may be preferred by patients
- NIV improved some lung function parameters
- Exercise performance using NIV significantly improved compared with room air *(3 trials, 27 patients)*

Cochrane review : 6 trials, 151 participants: NIV for airway clearance compared with conventional chest PT methods. F. Moran, J. Bradley and A. Piper 2017

NIV and Airway Clearance

- NIV may help temporarily reverse or slow the progression of respiratory failure
- NIV may help avoiding tracheal intubation
- NIV could be a useful addition to other airway clearance techniques, especially for patients with difficulty to expectorate sputum
- Long term randomized controlled trials are needed to determine the clinical effect on airway clearance and exercise capacity

Cochrane review : 6 trials, 151 participants: NIV for airway clearance compared with conventional chest PT methods. F. Moran, J. Bradley and A. Piper 2017

ELSEVIER

Journal of Cystic Fibrosis 11 (2012) 187–192

Journal of Cystic Fibrosis

www.elsevier.com/locate/jcf

Original Article

Long-term non-invasive ventilation in cystic fibrosis — Experience over two decades☆

William G. Flight [a,b,*], Jonathan Shaw [a], Susan Johnson [a], A. Kevin Webb [a,b], Andrew M. Jones [a,b], Andrew M. Bentley [b,c], Rowland J. Bright-Thomas [a,b]

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[b] *Respiratory Research Group, University of Manchester, Oxford Road, Manchester, M13 9PL, United Kingdom*
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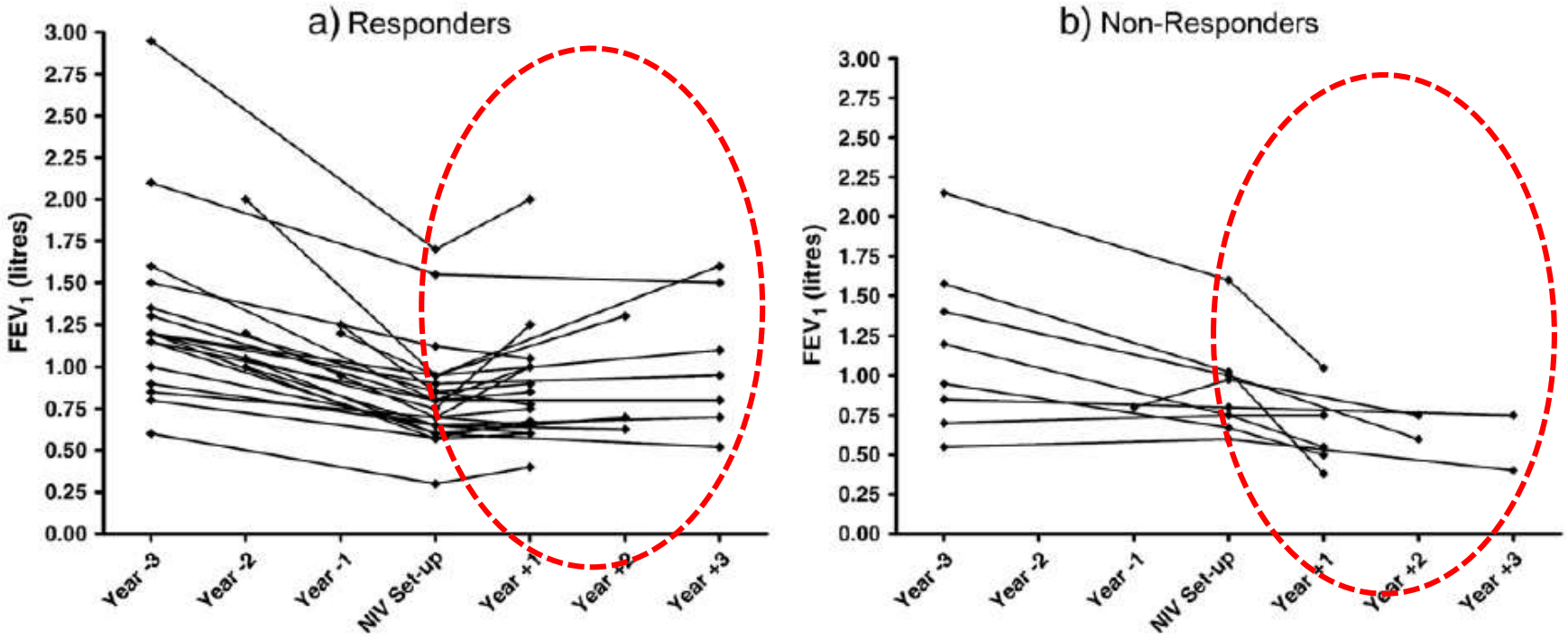
Not all patients respond positively to NIV



a) **responders to NIV** (i.e. patients showing an improvement or reduction in decline of FEV1)
b) **non-responders to NIV** (i.e. acceleration of decline in FEV1 post-NIV).

Flight et al, Journal of CF 2012

Hayek- Biphasic Cuirass Ventilation

- Controls both phases of the respiratory cycle
- Decreases work of breathing
- Secretion clearance mode to mobilize and expel secretions
- Reduces CO_2 when using setting of 60 cycles per minute

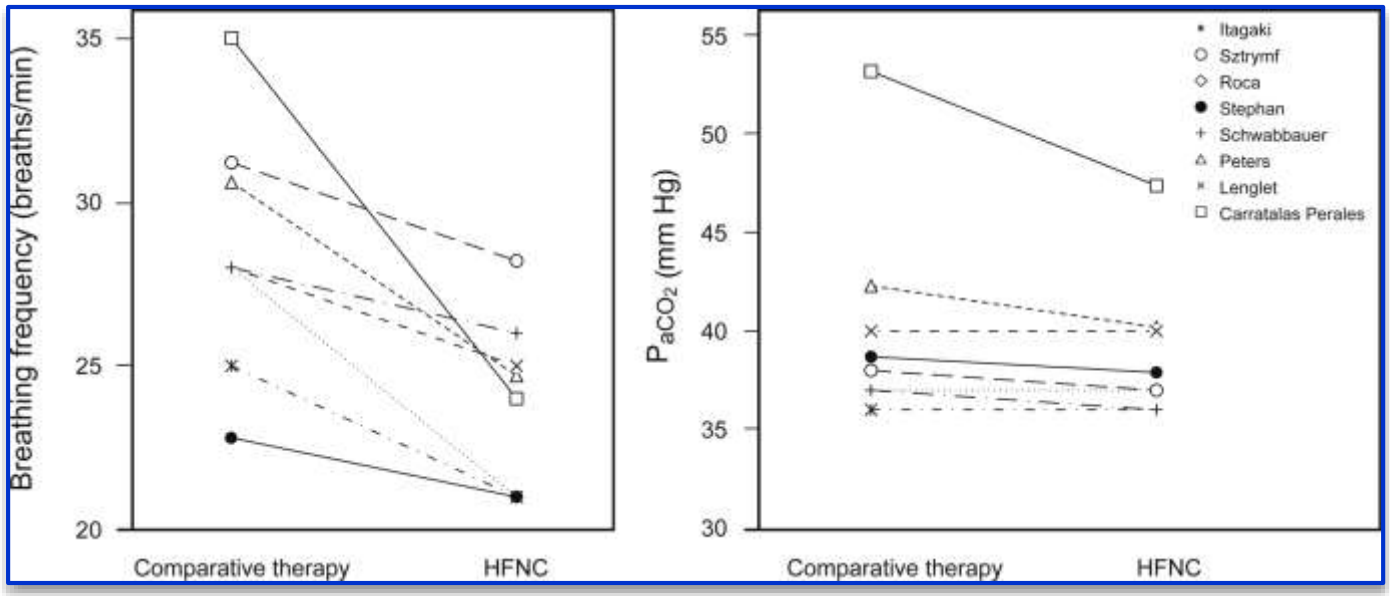
Humidified Heated High Flow Nasal Cannula

- Comprises humidified heated O_2 in a NC
- Delivers O2 up to 60 L/min of flow
- Physiological effects:
  - Reduces anatomical dead space
  - PEEP effect
  - Prevents negative effects of dry gas
  - Decreases nasal resistance
  - Decreases mucosal inflammation
  - Allows to speak and eat

Nishimura M, J Intensive Care 2015

Comparison between HFNC and different therapies

In all studies (n=8)
breathing frequency with HFNC was lower



P_aCO_2- No statistically significant differences between the therapies

Nishimura M, Respir Care 2016

Cannulation strategy to facilitate ambulation

- Ambulation can be safely achieved for patients cannulated via a DLC in either VV or subclavian/central VA configuration
- Rehabilitation on ECMO may improve post transplantation outcomes
- Bedside upper and lower extremity strength training exercises should also be performed regularly
- Patients should walk few times a day as tolerated
- A multidisciplinary team, facilitates safe and effective physical rehabilitation on ECMO

Sharma et al, Ann Transl Med 2017

End of life care

- Care should focus on comfort and dignity
- Care should be discussed and tailored to each patient's goals and values
- Work of breathing should be minimized
- Airway clearance should be continued if the patient finds it beneficial to relieve symptoms

BM Button et al, Respirology 2016

תודות

לצוות הרב תחומי במרכז לסיסטיק פיברוזיס בהדסה הר הצופים