Respiratory Function in MND

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- Case example
- Physiological effects of MND on respiratory function
- Clinical signs of respiratory compromise
- Investigations for respiratory function
- Cough support and secretion management
- Non-invasive ventilation

Patient

- 69 female
- 9 month progressive breathlessness on exertion
- Weakness and intermittent cramping in the hands.
- Pain in shoulders, hips, knees and back.
- Difficulty lifting the vacuum cleaner
- Ex-smoker – 20 pack years
- Known hypothyroidism and hypertension

- On examination;
  - BMI 38
  - Kyphosis
  - Clear chest
  - No fasciculations, fatiguability, proximal muscle weakness
- CXR – raised right hemidiaphragm
- Spirometry – restrictive picture
- Considered;
  - COPD
  - Diaphragmatic weakness / Obesity
  - Rheumatological / Neurological underlying diagnosis

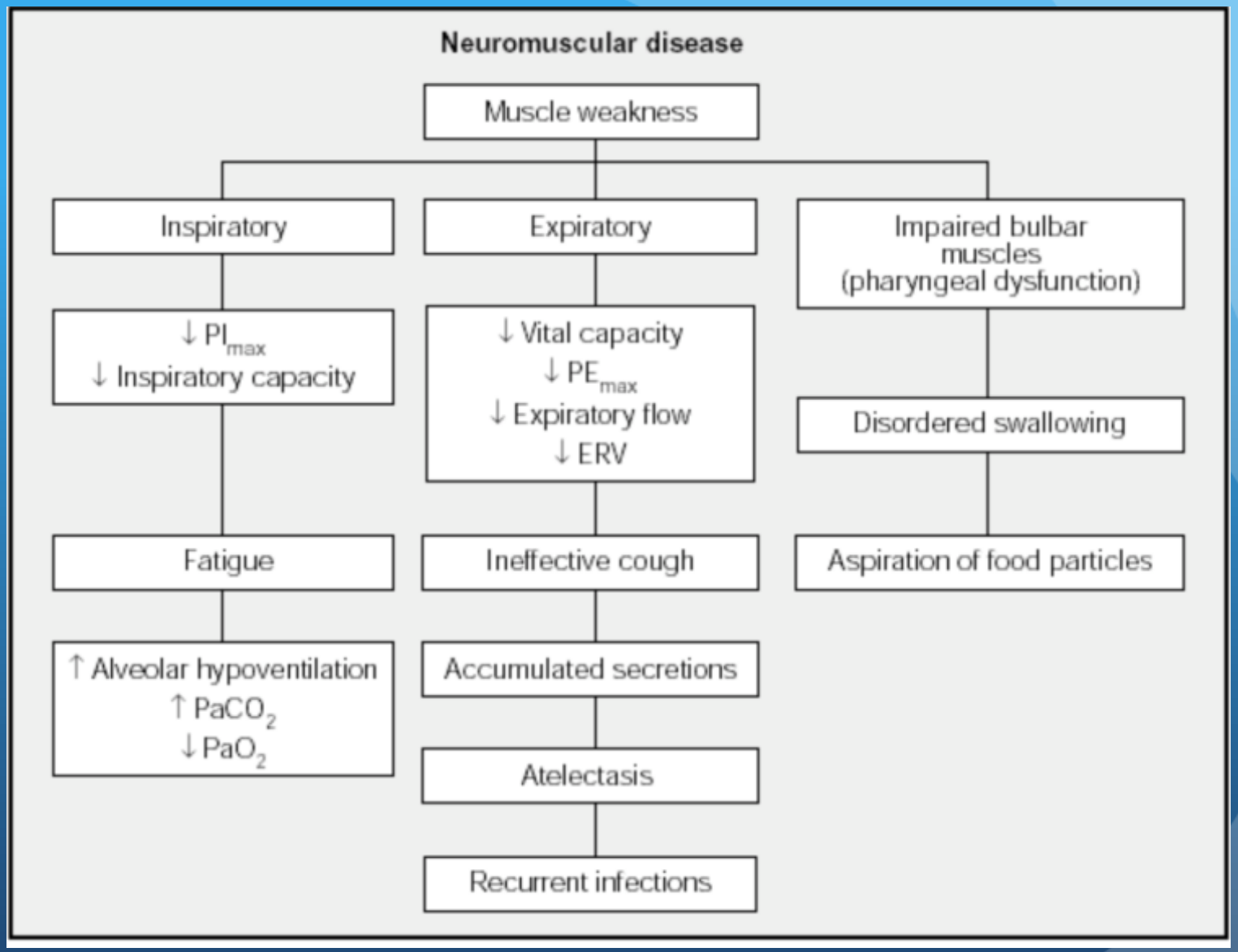
- FVC 2.83L (56.8% predicted)
- Unable to manage a supine VC
- MIP 28% predicted,
- MEP 83% predicted
- CBG: pH 7.51, pCO_2 5.9, pO_2 10, HCO_3 33.3
- ONO – ODI 18, Mean sats 87%

- Neurology assessment
- Husky speech, no swallowing issues
- No wasting / fasciculations
- Weak cough and sniff
- Proximal weakness in all limbs
- Bloods - NAD
- EMG → Denervation consistent with MND

- Started on NIV
- Used successfully for symptom control.
- pCO2 never above 6.1 Kpa
- Noted to be ventilator dependent after 6 months on NIV
- Survival from diagnosis = 8 months

Respiratory failure in MND

- MND is a progressive neuromuscular disorder
- 30% have bulbar onset symptoms
- 3% of patients with MND present with primary respiratory symptoms.
- One third of patients die within a year of diagnosis – prognosis is poorer with respiratory compromise
- Cross over with other resp conditions such as OSA, OHS, COPD, recurrent infections, aspiration pneumonia.



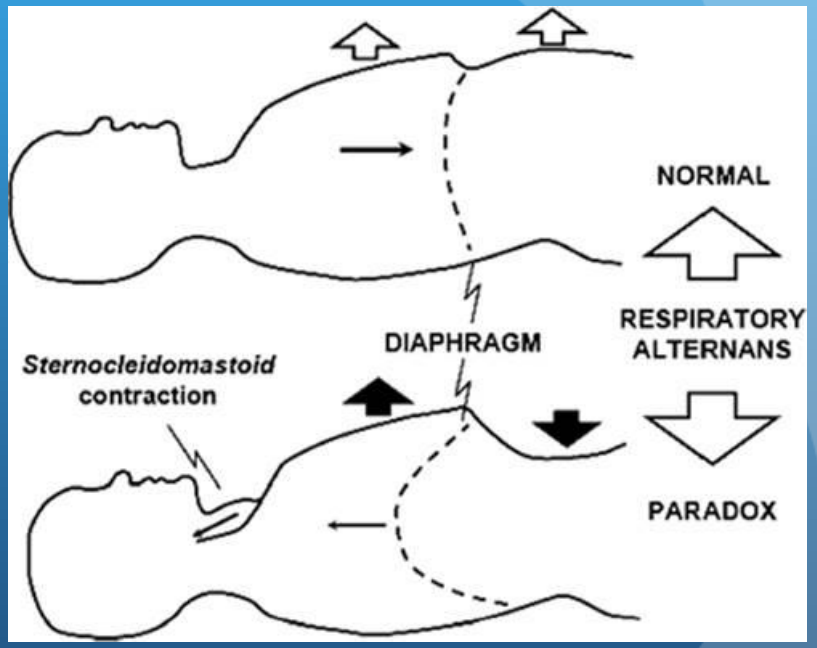
Lisboa C, *et al*. *Arch Bronco*. 2003;**39**:289

Resp muscle weakness: symptoms

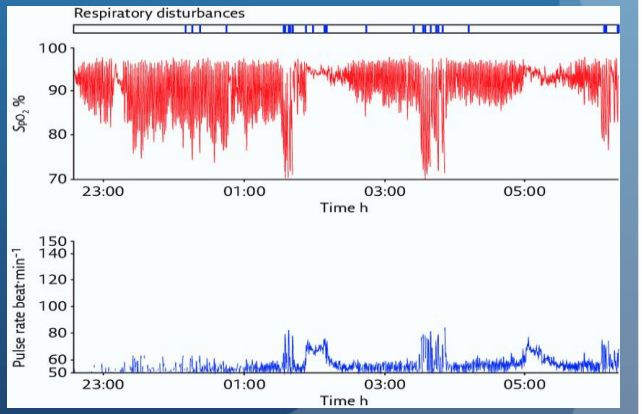
- Disturbed or unrefreshed sleep
- Excessive daytime sleepiness
- Early morning headaches
- Orthopnoea
- Snoring / nocturnal apnoeas
- Increasing dyspnoea
- Poor concentration / memory
- Recurrent chest infections

Resp muscle weakness: signs

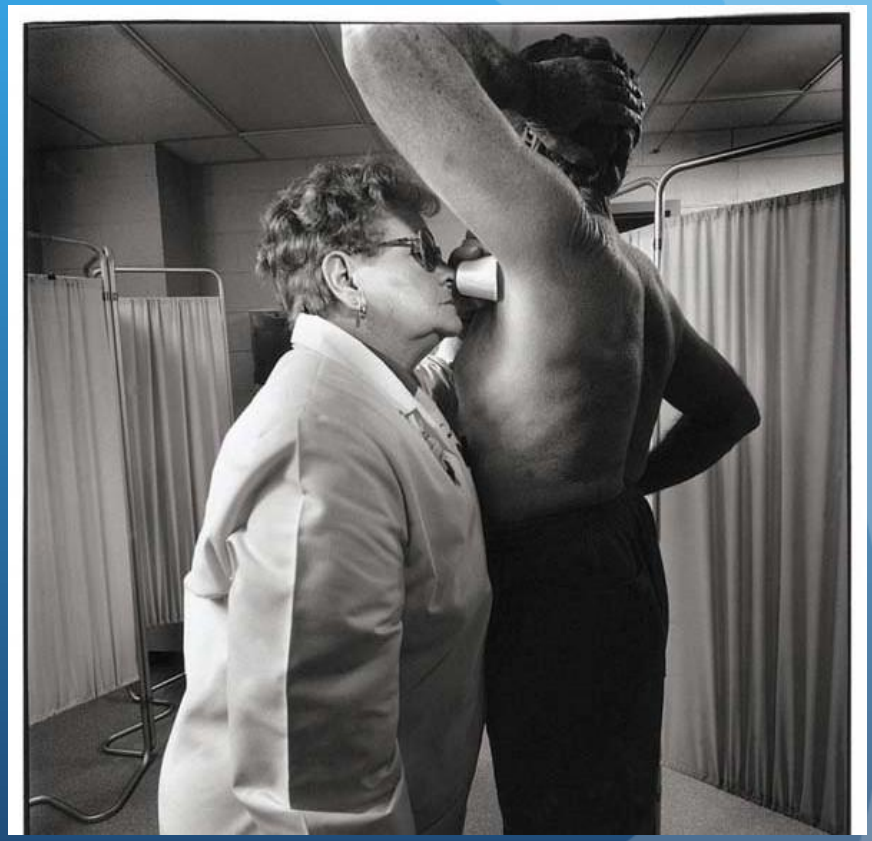
- Increase resp rate
- Shallow breathing
- Weak cough
- Weak sniff
- Abdominal paradox
- Use of accessory muscles
- Reduced chest expansion



Resp muscle assessment



SNIFF test – reliable / reproducible

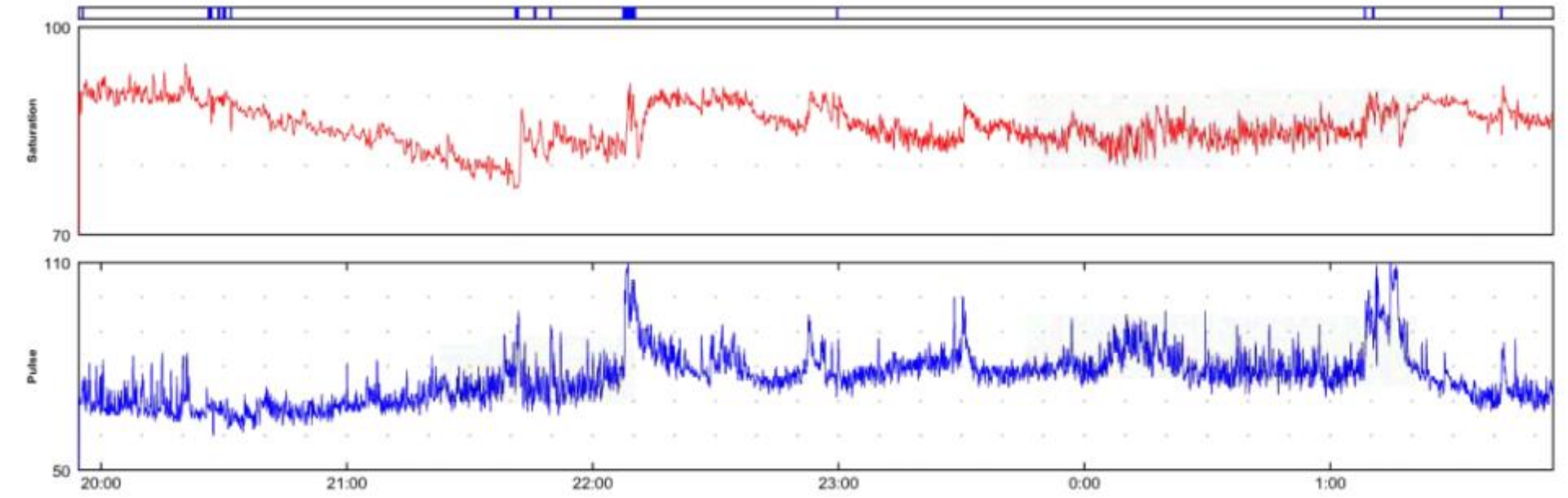


Resp muscle strength testing

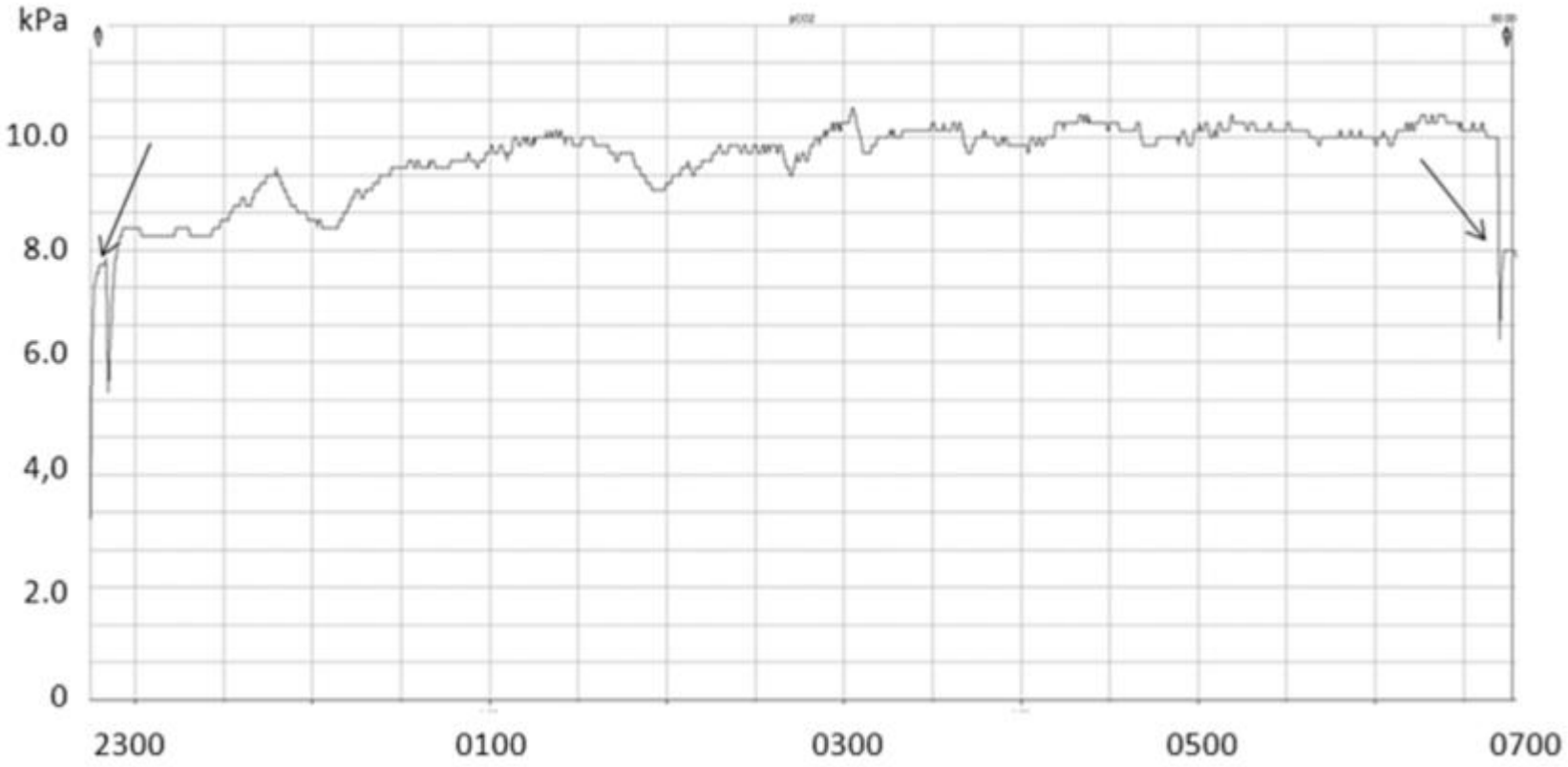
- FVC or VC
  - FVC or VC < 50% predicted
  - FVC / VC < 80% predicted with symptoms / signs of reps impairment (esp orthopnoea)
  - Fall of >20% in sitting → lying VC

- SNIP / MIP
  - SNIP or MIP < 40cm H2O
  - SNIP or MIP < 65cm H2O (Men) or 55cm H2O (women) with symptoms / signs of resp impairment
  - Rate of decrease in SNIP or MIP of > 10cm H2O per 3 months on serial testing

Overnight oximetry



Transcutaneous CO_2 monitoring



Aarestad S, *et al*. *Respir Med*. 2016;**112**:112–8

When to refer

- Referral for ABG/CBG if oxygen saturation (SpO2) <= 94% (or 92% if known lung disease).
- Sleep-related respiratory symptoms, despite the SpO2 being within normal limits
- PaCO2 on CBG/ABG > 6 kPa: urgent referral is indicated
- Symptoms or signs of respiratory impairment despite a PaCO2 <= 6 kPa
- Symptoms or signs of respiratory impairment despite normal overnight pulse oximetry.

Secretion management

- Hypersalivation
  - Glycopyrronium
  - Hyoscine
  - Atropine
  - Amitryptyline
  - Botox
- Retained secretions
  - Hydration
  - Mucolytics
  - Hypertonic saline
  - Posture
  - Chest physio
  - Portable suctioning

Supporting the cough

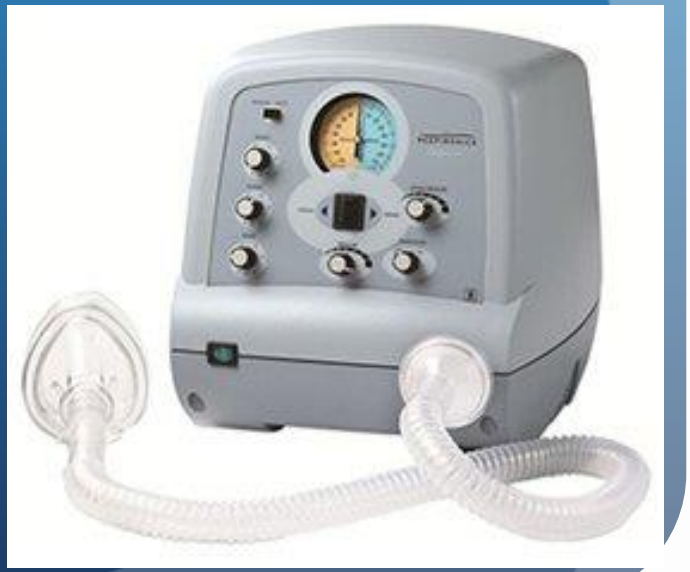
- Unassisted breath stacking

- Assisted breath stacking

(eg using a LVR bag)

- Mechanical insufflation-exsufflation

(eg. CoughAssist device)



Other interventions

- Prophylactic pneumococcal and influenza vaccinations.
- Reduce risk of aspiration eg. gastrostomy
- There may be a place for early use of antibiotics, if this is what the person with MND wants.
- CPAP therapy if OSA due to pharyngeal weakness
- Oxygen can worsen hypercapnia and abolish hypoxic ventilatory drive but may be used in certain situations.
- Palliative measures for dyspnoea eg. Opiates, benzodiazepines

MND Respiratory Pathway

This pathway is interactive: if you're viewing online, click **find out more** to view further details. These links will only work if you are connected to the internet.

Find more information on respiratory evaluation and management at: **www.mndassociation.org/respiratory**

mnda
motor neurone disease association

Signs and symptoms
Signs and symptoms of respiratory impairment include:
- early morning headaches
- daytime sleepiness
- orthopnoea

Find out more

Respiratory assessment
Soon after diagnosis and appropriate to the person's needs, refer for baseline respiratory function tests.
- SpO_2
- FVC/VC
- SNIP

Find out more

Secretion management
Consider early referral to **respiratory physiotherapist**.
- secretion management
- provision of portable suction
- lung compliance
- cough augmentation eg manual cough assistance, Mechanical insufflator:exsufflator (MIE) and Lung Volume Recruitment (LVR) bag. **Find out more**

Respiratory function testing
Repeat tests every 3-6 months (as appropriate) The multidisciplinary team should continually monitor signs and symptoms of respiratory impairment **Find out more**

In the presence of dementia, consider the patients' ability to give consent and level of understanding. **Find out more**

Palliative care
Referral to **specialist palliative care services** (if not already in contact).

Advance care planning
Sensitive discussions of options, including withdrawal of NIV, ADRT, end of life care. **Find out more**

Assisted ventilation
Offer discussions about possible use of assisted ventilation at:
- diagnosis
- during testing
- when respiratory function changes.

MND Just in Case Kit
Ordered by the **GP** from the MND Association. To be filled with medications to ease symptoms of breathlessness, choking and related anxiety/panic and kept in the home of the person with MND.

Management
A **physiotherapist** or **occupational therapist** may be able to discuss different approaches to coping with breathlessness **Find out more**

Non-invasive ventilation (NIV)
If NIV appropriate, refer for NIV trial. Discuss benefits, limitations and likely progression of NIV use. It is important to prepare a comprehensive care plan and provide 24hr emergency support and maintenance. **Find out more**

Oxygen
Oxygen should be used with caution in those with MND. **Find out more**

Palliation of symptoms
Medications include
- antimuscarinics to reduce saliva and lung secretions
- anxiolytics to reduce anxiety/ terminal restlessness
- opioid analgesics to reduce pain, cough reflex, dyspnoea and the feeling of effortful breathing, fear and anxiety.

Find out more

Invasive ventilation (IV)
Tracheostomy or IV is offered rarely, and only after careful counselling and consideration of factors including the high impact on family and carers. Occasionally it is used in an unplanned emergency situation.

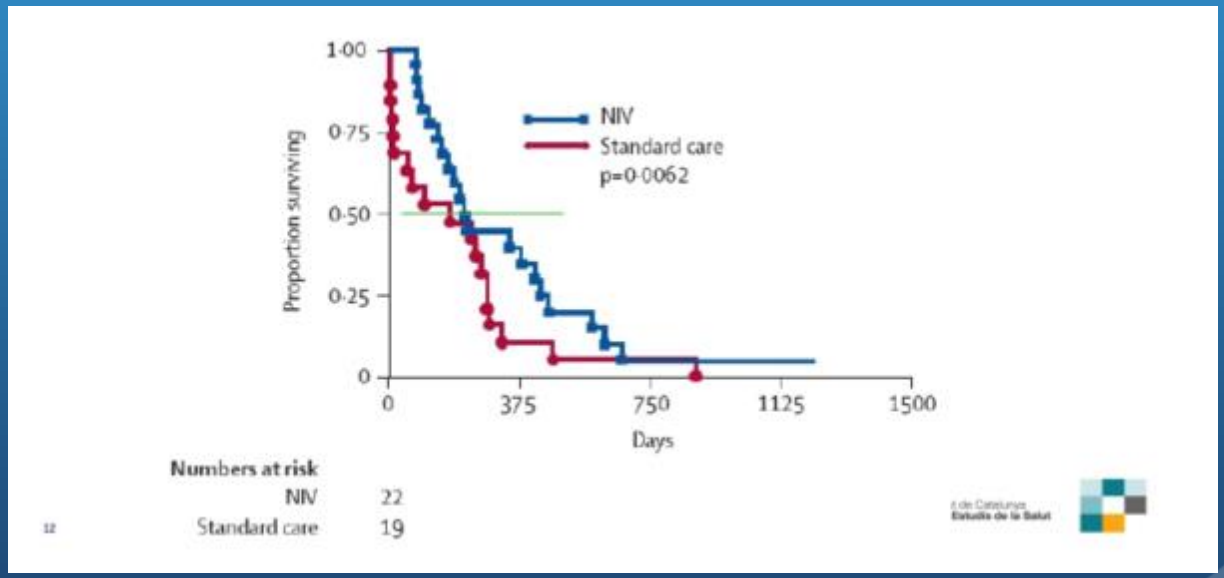
Gastrostomy
Referral to **gastroenterology** for discussion of gastrostomy (PEG/RIG/PIG). Timing and method of gastrostomy is dependent on weight loss and respiratory function. **Find out more**

Thank you to: Dr Naveed Mustfa, Consultant Respiratory Physician, University Hospital of North Staffordshire and Erna Swingwood, Respiratory Pathway Lead/Physiotherapist, University Hospitals Bristol NHS Foundation Trust.
Updated April 2015. Next review April 2017.

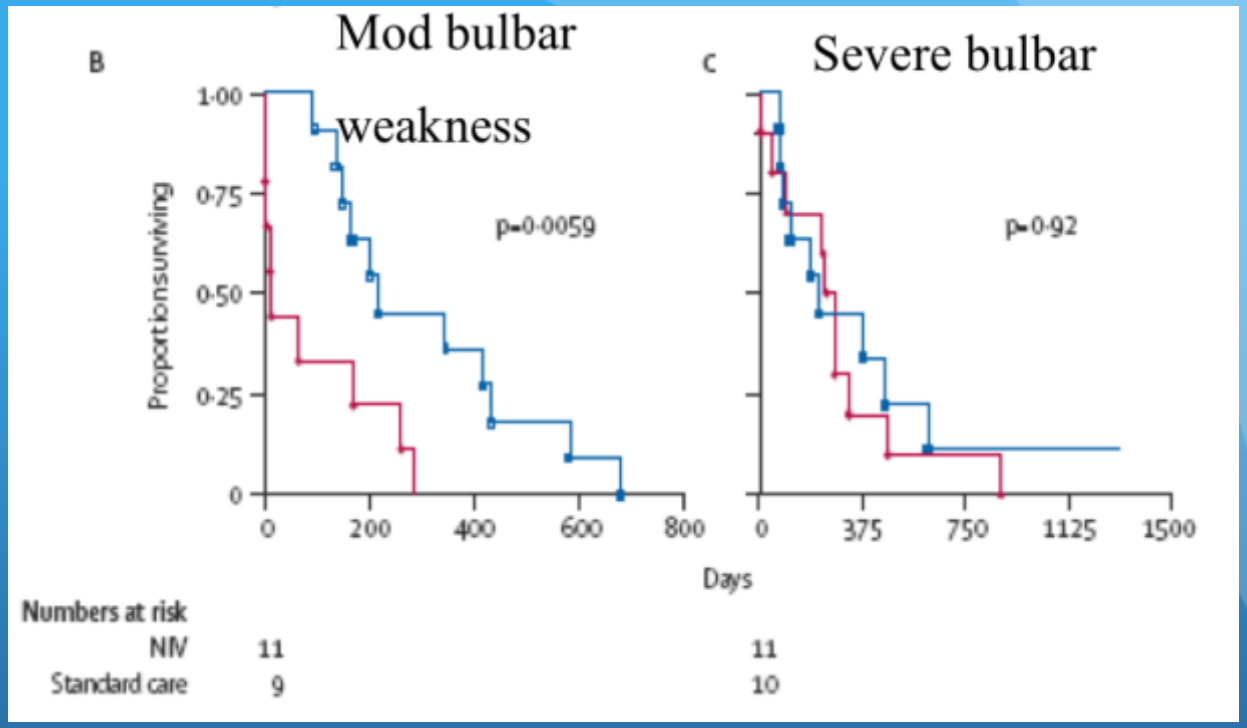

Hi Alarm
Lo Alarm
Help
Alarm Clear
Set
IPAP
EPAP
Ti
Back Up
Mode
Battery
Power
NIPPY 3+

Non-invasive ventilation

- RCT. NIV vs Standard care in MND patients with;
  - Orthopnoea and MIP <60% predicted **OR**
  - Symptomatic hypercapnia



Bourke SC, *et al*. *Lancet Neurol*. 2006;**5**:140–7



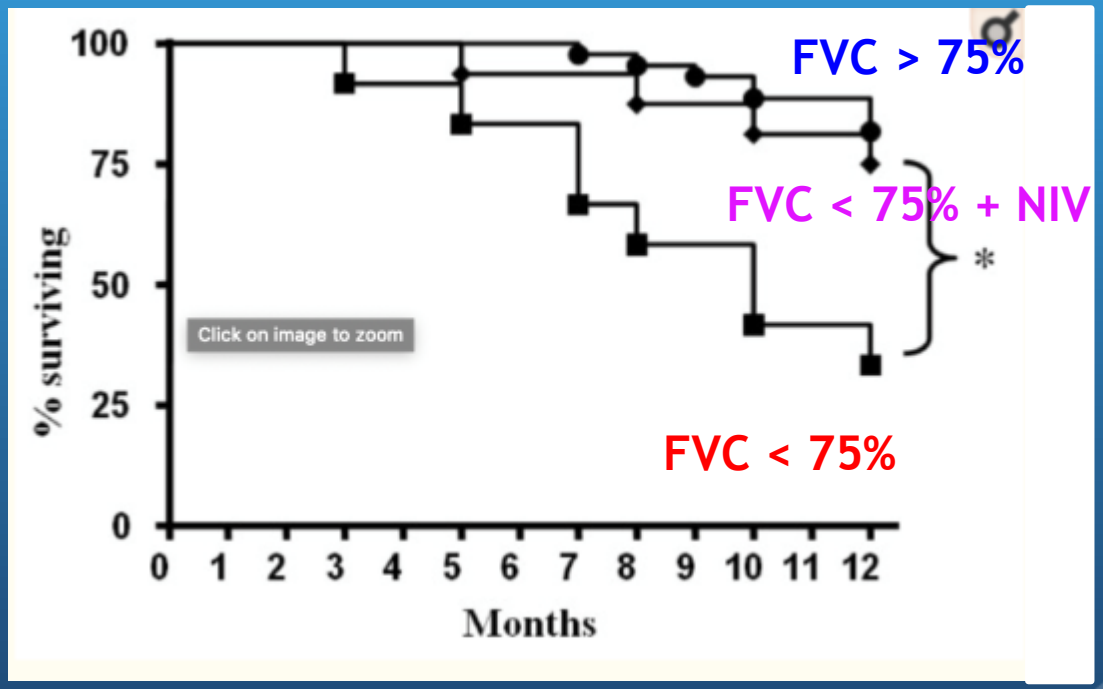
- Improved survival (Median 205 days)
- Maintains / improves QoL
- If severe bulbar impairment;
  - Improves sleep-related symptoms but does not offer a survival advantage.

Timing of NIV

- Adherence is key to success;
- NIV too early: Limited symptomatic benefit, compliance may be a challenge (burden of treatment), may decline NIV later when more severe
- NIV too late: Limit the survival benefit or symptomatic benefit of assisted ventilation
- Discuss the rationale, benefits and drawbacks of ventilation early in the disease process

Role for NIV earlier?

- Trial of NIV with FVC < 75%



Carratu, P, *et al*. *Orph J Rare Dis*. 2004;4:10

Mean FVC 83% vs Mean FVC 65%

Practical issues with NIV

- Identify the most appropriate type of non-invasive ventilator and interfaces, based on the person's needs and lifestyle factors and safety
- the risk, and possible consequences, of ventilator failure
- the power supply required, including battery back-up
- how easily the person can get to hospital
- risks associated with travelling away from home (especially abroad)
- whether a humidifier is required

Tracheostomy

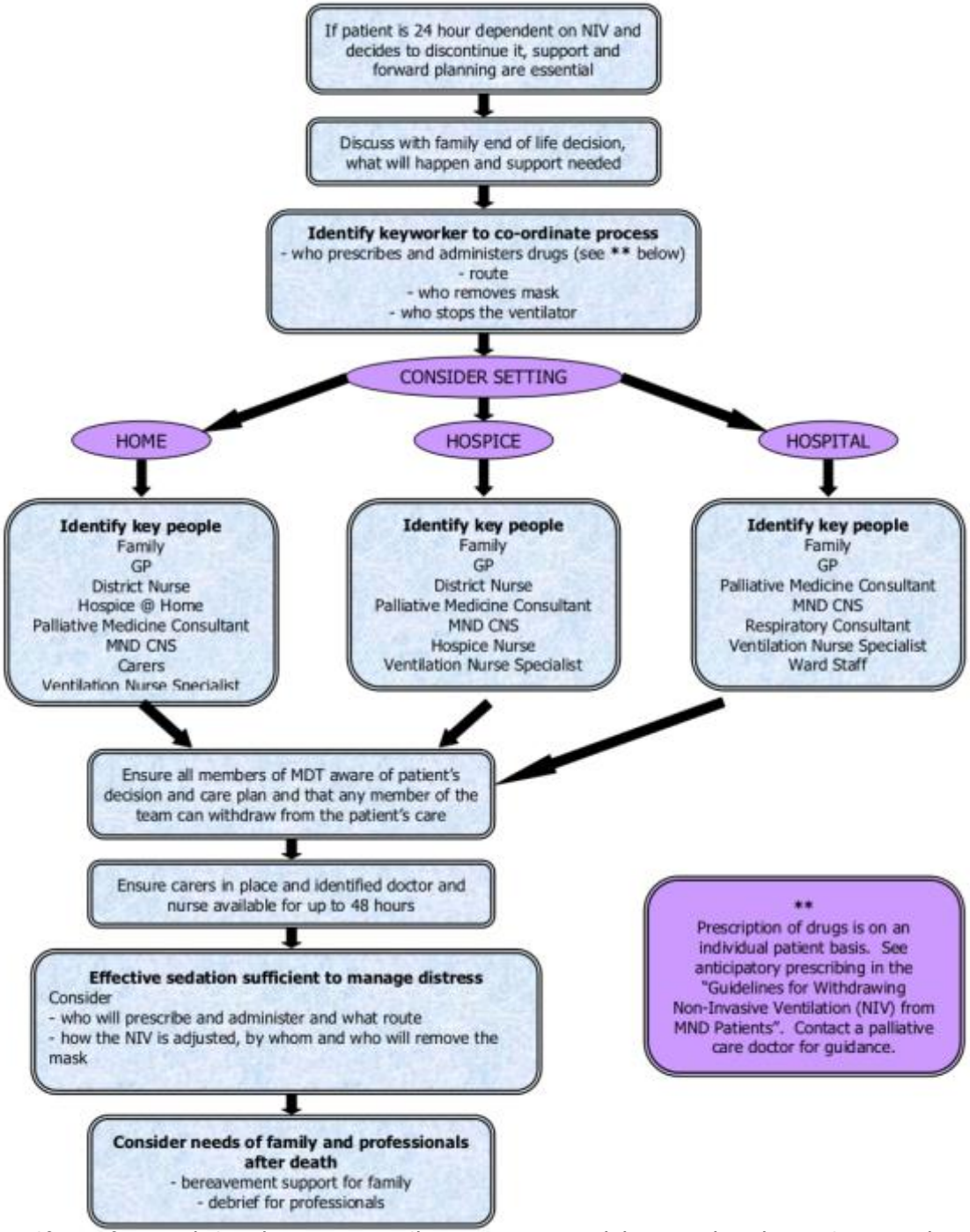
- Useful in a very small subgroup of MND;
  - Bulbar dysfunction leading to aspiration
  - Failure of NIV
  - Failure to wean onto NIV post acute decompensation
  - 24 hour ventilator dependence with bulbar dysfunction

MND: Ventilator dependence

- Defined as usage for more than 16 hours/day
- Sign of significant disease progression
- Back-up ventilator
- Emergency drills for power failure / interface dislodgement
- Palliative care planning – advanced directives
- Consider additional symptomatic control with opiates or benzodiazepines.

NIV in palliative care

- Withdrawl of NIV
  - Patients should be informed that they have the right to stop NIV if they choose
  - Patient, family and carer anxiety
  - Advanced directive – interpretation and implementation
  - Capacity
  - Time and place of withdrawl
  - MDT discussion – Communication
  - Sedation prior to withdrawl
  - Who removes the mask?



https://www.mndassociation.org/forprofessionals/mndmanagement/best-practice-guidelines-and-pathways/care-pathway-and-clinical-guideline-examples/

Summary

- Respiratory compromise in MND can present insidiously.
- Urgent referral if pCO2 > 6Kpa.
- Management of secretions and cough strength is important.
- Non-invasive ventilation offers a survival benefit of around 6 months, esp in non-bulbar disease.
- NIV has an important role in symptom management.
- Early discussion about respiratory symptoms and early referral to specialist services is key to improving patient experiences and outcomes.

Questions?