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MND AND NIV

Overview

- Why am I here?
- What is NIV?
- When and why to consider starting
- How to start
- Problems
- When to stop
- How to stop

MND

- Progressive neurodegenerative disorder
- May effect any part of neuromuscular system
- Aetiology unclear
- Incidence 1.5-2.7/100,000 per year
- Prevalence 2.7-7.4/ 100,000

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Why am I here?

- Respiratory failure occurs in MND due to weak respiratory muscles
  - Causes of respiratory failure
    - Inability to get air in (and out)
    - Inability to exchange gas

- When respiratory failure occurs, it's always worse at night.
- 1 RCT and a range of cohort studies suggesting benefit from NIV in MND

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What is NIV?

- Any form of ventilation that can be delivered without the need for a endotracheal tube.
- May include CPAP and Bilevel CPAP
- May be used to treat nocturnal respiratory failure

CPAP and Bilevel CPAP

CPAP

- Uses a mask interface
- May add oxygen into circuit
- One pressure throughout respiratory cycle
- Hard to breathe out against
- Only 1 mode
- **Terrible for MND**

Bilevel CPAP

- Uses a mask interface
- May add oxygen into circuit
- Different pressures for inspiration and expiration
- Easy to breathe out
- 2 modes
  - Spontaneous
  - Spontaneous timed
- **Good for MND**

Modes in Bilevel CPAP

Spontaneous

- 2 pressures IPAP and EPAP
- Transition b/w pressures is ENTIRELY dependent on patient breathing
- Not really used in MND

Spontaneous timed

- 2 pressures IPAP and EPAP
- Transition b/w pressures MAY be triggered by patient
- If patient doesn't take a breath, the machine 'breathes for them'
- Usual mode in MND

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When and why to start

- **Symptoms**
  - Sleepy during the day
  - Morning headaches
  - Other sleep symptoms (witnessed apnoeas)
  - Breathlessness, esp orthopnea
- **Lung function tests**
  - Forced Vital Capacity (FVC) <50% predicted
  - MIPS/ MEPS
  - ?supine spirometry
- **Patient request**

Why not start?

- Caregiver burden
- Prolong life in setting of burdensome symptoms
- Difficulty with mask fitting
- Cost

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How to start

- Referral to sleep physician or clinic
- Document night time respiratory failure
  - Arterial blood gas/ venous bicarbonate
  - Sleep study **OR** Overnight oxymetry
- Ensure discussions about PEG feeding have been initiated
- Empiric pressure setting
  - May need pressure titration in sleep centre
  - ?hospital admission for acclimatisation

How to start

- Advance Care Planning conversation(s)
  - Mandatory!!
  - Discuss potential benefits and harms
  - Discuss that may become increasingly dependent
  - Discuss the idea of 'weaning' NIV
  - Discuss what to do in the setting of chest infection

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Problems with NIV

- Fitting the mask
- Taking the mask off
- Aspiration / 'wet mouth'
- Electricity supply/ blackouts
- Pressure areas
- Discomfort
- Carer burden

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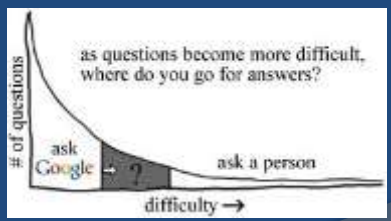
When to stop

- This is not an easy question.



When to stop

- When the benefit is not worth the burden
- Patient request
- Dying

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How to stop

- Easy if patient decides to stop – just stop

How to stop if dying/ NIV dependent

- Wow, you thought the last question was hard...

How to stop if NIV dependent

- No evidence
- No published guidelines

How to stop if NIV dependent

- Clear communication
  - Won't die immediately NIV is removed
- Anticipate symptoms and provide appropriate treatment

How to stop if NIV dependent

- Carefully titrated doses of opioids +/- benzodiazepines
- Morphine 15mg/24h via SD(for opioid naive)
- Midazolam 5-15mg/24h via SD SD(for benzo naive)
- ?supplemental oxygen
- Fan
- Semi-recumbent
- Start the SD, Give a breakthrough dose of drugs, THEN remove the mask
- ?? Terminal sedation

Conclusion

- People with MND may develop respiratory failure
- BiLevel CPAP has a role in treatment
- Requires clear communication as there are Advance care planning implications
- Knowing when to start can be hard
- Knowing when to stop is harder
- Actually stopping is potentially tricky