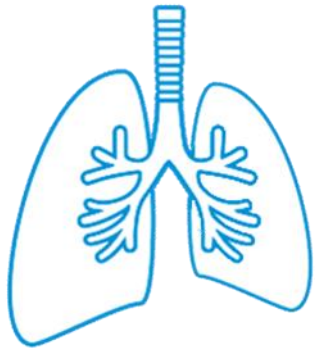
Respiratory Management and Non Invasive Ventilation in MND



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- Respiratory muscle weakness is common and can lead to
  - Insufficient ventilation
  - Ineffective cough
  - Nocturnal hypoventilation
- Supportive care can provide symptomatic relief
- Management can include any and all of the following:
  - Cough assistance
  - Tracheostomy
  - Non-Invasive ventilation

Clinical Manifestations

- Insufficient ventilation:
  - Shortness of breath – dyspnoea and/or orthopnoea
  - Rapid shallow breathing
  - Accessory respiratory muscle use
  - Paradoxical breathing
  - Hypercapnia/hypoxemia

- Principle cause is weakness of inspiratory muscles
  - Tidal volume decreases
  - Respiratory rate increases
  - Increased respiratory rate insufficient to maintain alveolar ventilation
  - $PaCO_2$ begins to rise

Nocturnal Hypoventilation

- Choking
- Insomnia
- Daytime hypersomnolence
- Morning headaches
- Fatigue
- Impaired cognition

Evaluation of Lung Function

- Respiratory function tests:
  - Confirm respiratory muscle weakness
  - Identify ineffective cough
  - Identify patients who need ventilator support
- A pattern of restriction:
  - FEV1 and FVC<80% predicted
  - Normal FEV1/FVC ratio
  - TLC <80% predicted
- FVC and VC measured supine more than 10% lower than upright
- MVV reduced
- MIP and/or MEP reduced
  - MIP reflects the strength of the diaphragm and other inspiratory muscles
  - MEP reflects the strength of the abdominal muscles and other expiratory muscles
- SNIP
  - More accurate for measuring respiratory muscle strength

Ineffective Cough

- Poor cough caused by weakness of:
  - Upper airway
  - Inspiratory and expiratory muscles
- Predisposes to:
  - Aspiration
  - Pneumonia
  - Respiratory failure
- An effective cough has three phases
  - Inspiratory – rapid and large tidal volume inspiration. Increased lung volume lenghthens expiratory muscles so they can generate more pressure and flow
  - Compressive phase – glottis closure occurs at the onset of active expiratory muscle contraction
  - Expiratory phase – the glottis opens after approximately 0.2 seconds allowing high peak expiratory flow due to the sudden release of pressurised gas and dynamic airway compression.

Assess Cough

- Peak cough flow is measured by having the patient cough through a mask attached to a peak flow meter.
  - <160 l/min identifies ineffective cough.
  - 160 – 270 l/min are at risk
- MEP less than 60cmH_2O suggests ineffective cough.
- Try it and see…

Cough Assist

- Mechanical Insufflation-Exsufflation
  - During insufflation, positive pressure is applied, which results in an inspired tidal volume
  - Exsufflation rapidly follows as the pressure becomes negative.
- Manually-assisted cough
  - Abdominal thrust provided by a caregiver and timed to occur at the same time as the patients voluntary cough effort.
- Hyperinflation manoeuvre – increase the inspiratory tidal volume
  - Glossopharyngeal breathing (repetitive air gulping).
  - Delivery of stacked breaths (initiation of inspiration before the completion of expiration).
  - Mechanical inspiration using a mechanical insufflator or a volume cycled ventilator

Nocturnal Hypoventilation

- Inadequate ventilation may first manifest during sleep
  - Upper airway obstruction due to bulbar dysfunction
  - Decreased accessory muscle activity during REM sleep

- Symptoms of Nocturnal Hypoventilation include:
  - Choking
  - Insomnia
  - Daytime hypersomnolence
  - Morning headaches
  - Fatigue
  - Impaired cognition

Assess Need for Ventilatory Support

- FVC <50%
- MIP less negative than -30cmH2O or MEP below 40cmH2O
  - Reduced MIP indicates high risk for Hypercapnia
  - Reduced MEP indicates inadequate cough strength and risk for secretion retention
- VC <15-20 mL/kg, 60% of predicted, or 1L
- How is the patient feeling?

NIV – Non Invasive Ventilation

- NIV stabilises the upper airway and rests respiratory muscles
- NIV helps decrease or prevent nocturnal hypoventilation
- NIV decreases daytime sleepiness by correcting sleep architecture

- Not for everyone
- Can improve QoL (but difficult to measure)

Pressure

- Inspiratory pressure should achieve:
  - adequate tidal volume
  - proper respiratory rate
  - decrease in work of breathing
  - reduction in $PaCO_2$
- Expiratory pressure should:
  - overcome obstructive apnoeas and hypopneas

Mask Selection

- Pillows – small and least obtrusive
- Nasal – need to keep mouth closed
- Full Face – aspiration risk

Pros

- Can ease symptoms caused by weakened muscles involved with breathing
- Can increase survival time
- Portable, some have battery options
- Surgery not needed
- A range of mask options

Cons

- Can cause discomfort from air leak
- Can lead to pressure sores
- Claustrophobia
- Can cause a blocked nose, runny nose, dry mouth
- Aerophagia
- Takes time to get used to it
- Flow of air can disturb speech

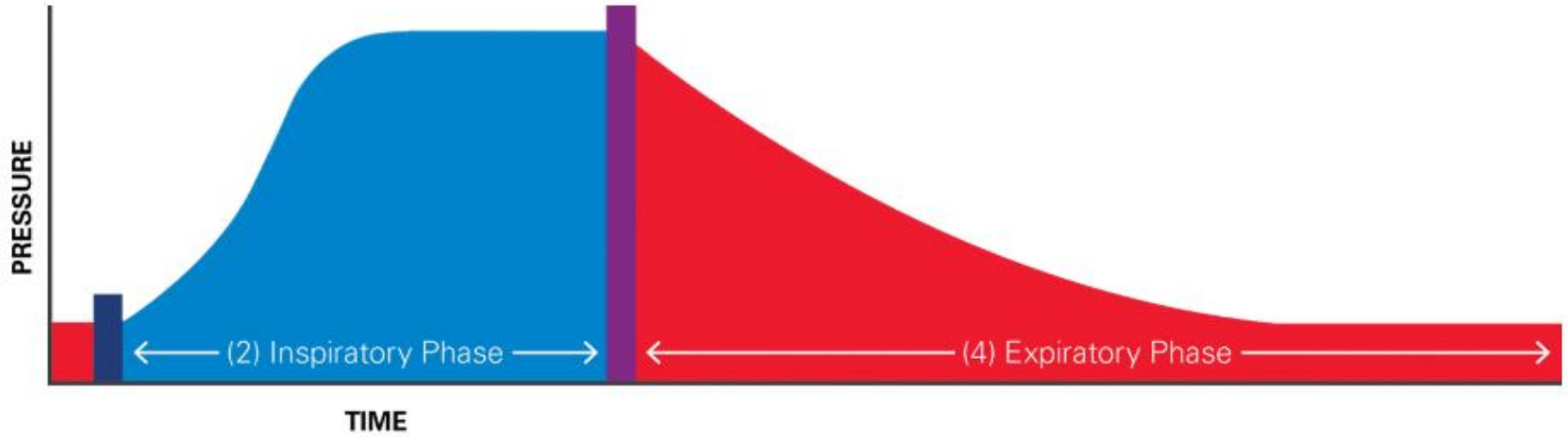
Withdrawal of Treatment

- Depends if NIV dependent or not…
- If not –
  - Patient can simply choose not to put it on
  - May need palliation of breathlessness/anxiety
- If dependent –
  - Plan needs to be made including family/carers/MDT

What can we do with NIV

- Manipulate
- Manipulate
- Manipulate

Breathing Phases

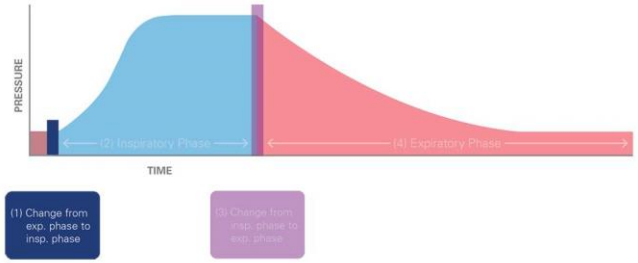


(1) Change from exp. phase to insp. phase

(3) Change from insp. phase to exp. phase

Phase	Variable
1	Trigger variable (Start inspiration)
2	Limit variable (Sustain inspiration)
3	Cycle variable (Stop inspiration)
4	Baseline Variable (Sustain FRC)

Phase 1: Trigger

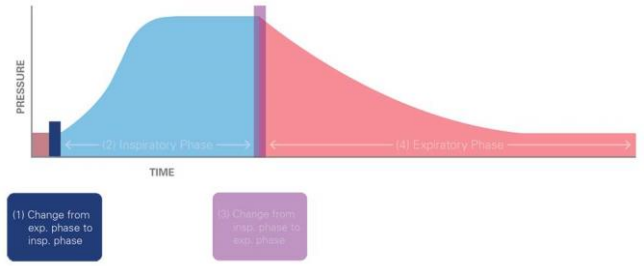


Trigger is a change in pressure or flow in the circuit that tells the device the patient has finished exhaling and is beginning to inhale. The device is “triggered” to initiate inspiration.

Parameters able to be modified during transition from expiration to inspiration (trigger):

- trigger sensitivities
- backup rate

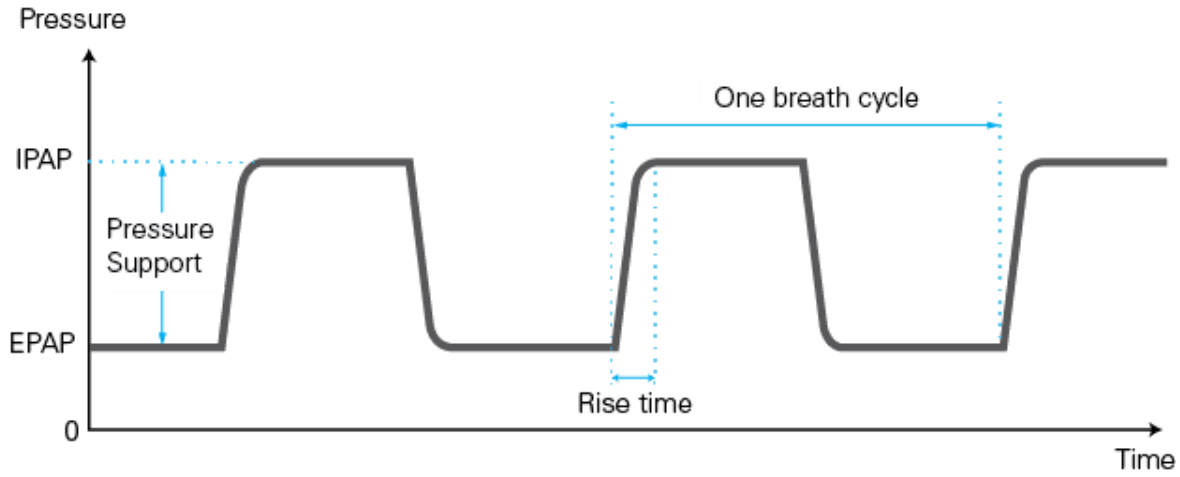
Phase 2: Inspiration



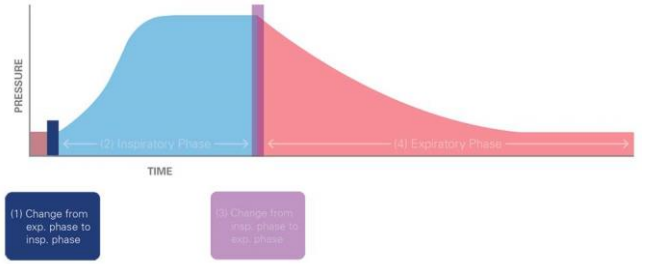
The inspiratory phase is the time between when the patient triggers a breath and the time they begin to exhale (cycle).

Parameters used during the inspiratory phase:

- rise time
- IPAP
- pressure support.


Pressure
One breath cycle
IPAP
Pressure
Support
EPAP
Rise time
0
Time

Phase 3: Cycle

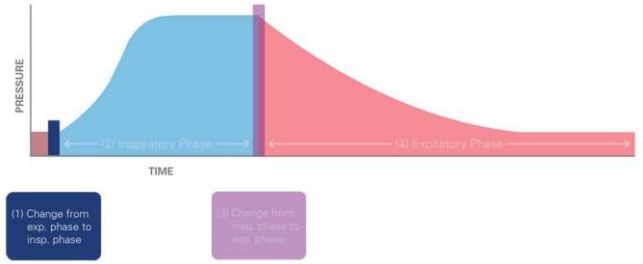


Cycle is the opposite of trigger—at the end of inspiration, the device transitions from IPAP to EPAP, allowing the patient to breathe out.

Cycle can be initiated by either:

- Patient flow – the patient can transition on their own; the set pressure is delivered until there is a certain decrease in inspiratory flow.
- Time – a certain amount of time elapses before ending inspiration.

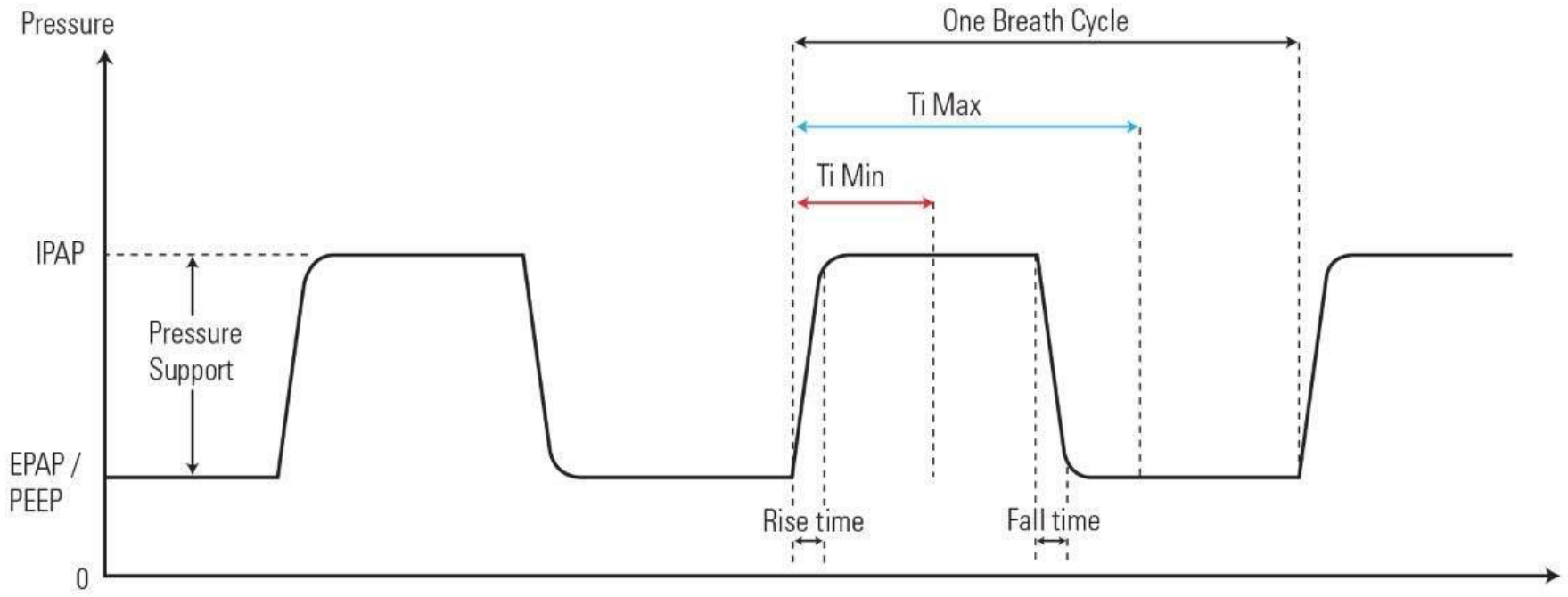
Phase 4: Expiratory Phase



The expiratory phase is the interval between the start of expiratory flow and the beginning of inspiratory flow, as shown in the red area.

- Fall time
- EPAP

Bilevel waveform showing EPAP & Fall Time.



Questions? Comments?