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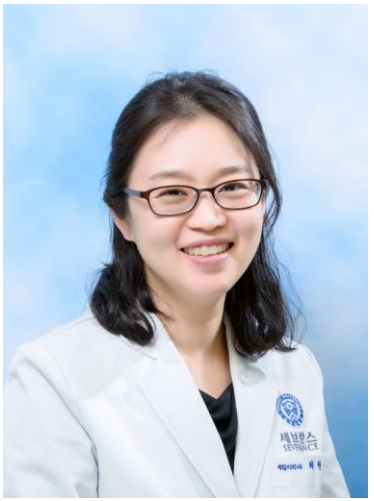

진료현장에서의 호흡재활의 실제와 Tip –제한성 폐질환의 호흡재활

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Restrictive lung disease

- **Neuromuscular disease**
  - ✓**DMD, myopathies & dystrophies**
  - ✓**ALS and SMA**
  - ✓**sequelae of polio,, neuropathies**
- chest wall deformities & kypho-scoliosis
- Cervical spinal cord injury

Contents

- Considerations for patients evaluation
- Considerations for management
- Understanding for treatment techniques

Consideration for patients evaluation

-What will be evaluations?

-What should be considered in each evaluation?

What will be evaluations?

- Outpatient clinic
  - Hypoventilatory symptoms
  - MIP/MEP
  - FVC/MIC
  - Coughing capacity
  - Ventilatory status

- Inpatient clinic
  - Continuous ventilatory status during sleep
  - Sleep disordered breathing

Considerations for evaluation

- Symptoms
- VC
- PCF
- Ventilatory status

Symptoms for respiratory insufficiency

Patient **walk** - Exertional dyspnea

W/C user : <u>symptoms may be minimal</u>

(except) during respiratory infection
when anxiety, inability to fall asleep, dyspnea

↑rate, ↓depth, irregularity of breathing

Symptoms	Signs
✓ Disturbed sleep ✓ Nightmare ✓ Daytime sleepiness ✓ Morning headaches ✓ Fatigue ✓ Poor appetite ✓ Poor concentration and/or memory ✓ Confusion ✓ Hallucinations	✓ Respiratory rate ✓ Shallow breathing ✓ Weak cough ✓ Abdominal paradox ✓ Use of accessory muscles of respiration

Vital Capacity (VC)

- Special consideration - the positional variation

- Diaphragm weakness
  : greater VC in sitting position

- Cervical cord injury patients
  : higher VC in a supine position

Diaphragm weakness

In a supine position,

- Increased pulmonary circulation triggers

  a reduction in the air volume inhaled into the thorax (7.5% ↓)

- Compression of the diaphragm by abdominal contents
  - inspiration → diaphragm descend

    : weakened diaphragm cannot push out abdominal contents
  - low inspiratory volume → low VC

Supine VC

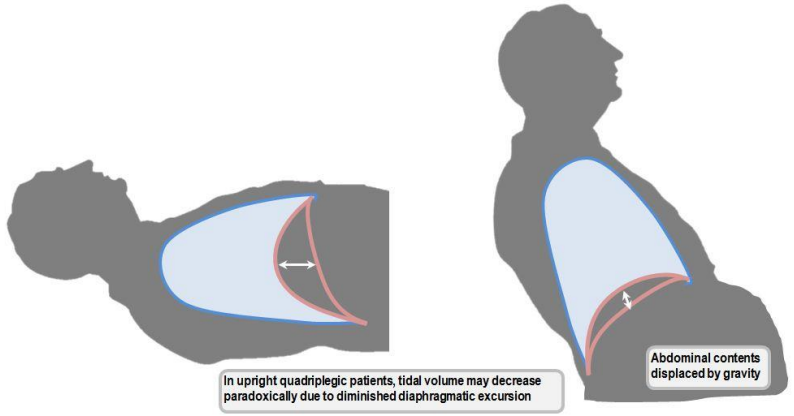
<u>most important indicator of ventilator dysfunction</u>

(hypoventilation is often worst during sleep)

Correlated with Orthopnea

Cervical cord injury

- Paralysis of expiratory muscle group (ex. abdominal muscle)
  → no contraction on breathing out
  → expiration while in a sitting position occurs passively by deflation of the fully inhaled lungs and thoracic wall through recoiling.
- Descended abdominal contents, along with gravity
  → reduce the excursion of the diaphragm.
  ► Reduced VC in a sitting position versus supine.



Change in Forced Vital Capacity with Postures according to Neuromuscular Disease

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Objective: To evaluate the difference in forced vital capacity (FVC) between sitting and supine position in patients with amyotrophic lateral sclerosis (ALS), cervical spinal cord injury (SCI) and Duchenne muscular dystrophy (DMD).
Method: FVC was measured in sitting and supine position for 32 patients with DMD, 32 patients with cervical SCI and for 28 patients with ALS. The highest value in three or more attempts in each position was chosen.
Results: FVCs measured in cervical SCI and ALS patients in the sitting and supine position were 1612.8±291.0 ml, 1393.2±286.7 ml and 2054.7±545.8 ml, 1104.3±425.4 ml respectively. Cervical SCI patients showed significantly higher value in the supine position ($p<0.05$). And ALS patients showed significantly higher value in the sitting position ($p<0.05$). FVCs measured in DMD patients were 1311.6±260.7 ml and 1213.8±378.9 ml respectively. There was no statistically significant difference between the measurements in both positions.
Conclusion: Difference in postural change of FVC was observed in patients with different types of neuromuscular disorders. Such difference in FVC suggest that postural change of FVC should be considered in management of neuromuscular disease with respiratory muscle weakness. **(J Korean Acad Rehab Med 2006; 30: 80-85)**

Key Words: Amyotrophic lateral sclerosis, Cervical spinal cord injury, Duchenne muscular dystrophy, Forced vital capacity, Postural change

Table 2. Results of Pulmonary Function Test

	DMD[5] (n=30)	Cervical SCI[6] (n=32)	ALS[7] (n=30)
FVCsit[1] (ml)	1311.6±260.7	1612.8±291.0*	1393.2±286.7†
FVCsit/FVCpre[2] (%)	38.8±5.1	41.5±6.1	40.5±5.1
FVCsup[3] (ml)	1213.8±378.9	2054.7±545.8*	1104.3±425.4†
FVCsup/FVCpre (%)	35.6±9.1	52.8±12.0	31.9±10.3
FVCsit-FVCsup (ml)	97.8±220.6	-441.9±445.3	288.9±286.0
ΔFVC (%)[4]	8.6±19.4	-28.3±25.4	21.8±20.2

Values are mean±standard deviation.
1. FVCsit: Forced vital capacity in sitting position, 2. FVCpre: Predicted value of forced vital capacity, 3. Forced vital capacity in supine position, 4. ΔFVC: {(forced vital capacity in sitting position-forced vital capacity in supine position)/forced vital capacity in sitting position} ×100, 5. DMD: Duchenne muscular dystrophy, 6. SCI: Spinal cord injury, 7. ALS: Amyotrophic lateral sclerosis
*$p<0.05$, † $p<0.05$

Conclusion

Difference in postural change of FVC was observed in patients with different types of neuromuscular disorders.

The postural change of FVC should be considered in management of neuromuscular disease with respiratory muscle weakness.

Peak cough flow (PCF)

- Defective coughing may be a primary cause of morbidity and mortality in NM
- Identifying the coughing capacity is essential for these patients.

Method

- By having the person cough as forcefully as possible through the peak flow meter.
- Expressed as peak cough flow.

Considerations in monitoring ventilatory status

The positional variation.

Diaphram wekness **VCsit > VCsupine**

Cervical cord injury VCsit < VCsupine

Wakefullness variation

Ventilatory drive – depressed when sleeping

Serial measurements under different circumstances is recommended for a more complete understanding of the ventilator state

ABGA painful, not fit for a serial monitoring

Non-invasive method of serial measurement

-continuous monitoring /more useful information, particularly during sleep

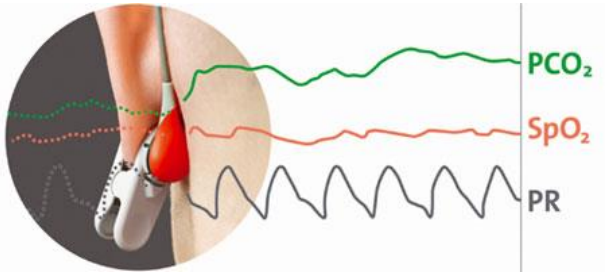
-SaO_2 (Oxyhemoglobin saturation) by Pulse Oximeter

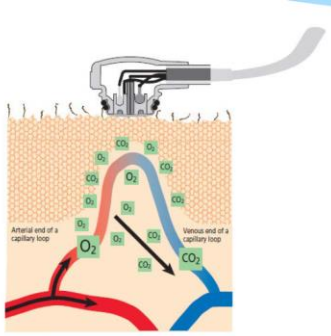
-**$EtCO_2$ (End-tidal CO_2)** by Capnometry /Capnography

-**$TcCO_2$(Transcutaneous CO_2)** by transcutaneous blood gas monitoring device

SaO_2 and End-tidal CO_2 monitoring

Transcutaneous CO_2 measurement





(B)

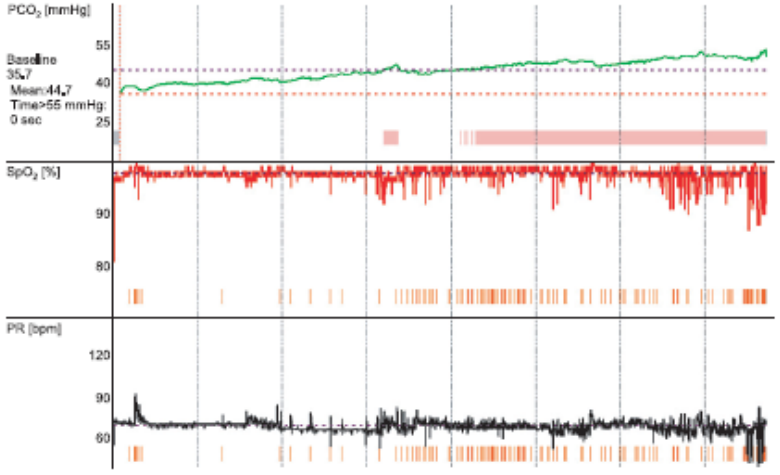


Fig. 1. These figures show an example of a patient's data from the transcutaneous blood gas monitoring device. (A) Table and (B) graph of a patient's data from continuous transcutaneous blood gas monitoring. The table shows mean, maximal PCO_2 values and portions of PCO_2 range, while the graph shows change in PCO_2 level through overnight continuous monitoring.

(A)

PCO_2		PCO_2 range	Duration	%-Duration
Minimum	35.7 mmHg (22:04:12)	90-200	0 sec	0%
Maximum	52.8 mmHg (05:43:08)	85-89	0 sec	0%
Mean	44.7 mmHg	80-84	0 sec	0%
Baseline	45.0 mmHg	75-79	0 sec	0%
Max.above baseline	17.1 mmHg (05:43:18)	70-74	0 sec	0%
		65-69	0 sec	0%
		60-64	0 sec	0%
		55-59	0 sec	0%
		50-54	58,27 min	13%
		45-49	3:00,28 hrs	39%
		40-44	3:01,56 hrs	40%
		35-39	38,08 min	8%
		30-34	0 sec	0%
		25-29	0 sec	0%
		0-24	0 sec	0%

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Annals of Rehabilitation Medicine

The Significance of Transcutaneous Continuous Overnight CO_2 Monitoring in Determining Initial Mechanical Ventilator Application for Patients with Neuromuscular Disease

we conducted study to reveal the significance of continuous transcutaneous CO_2 level monitoring by reviewing cases which showed a discrepancy in CO_2 levels between ABGA and continuous transcutaneous monitoring.

The total number patients : 89

- 53 : both ABGA and overnight Tc monitoring - hypercapnia
- 36 (40.45%) : ABGA - normal range
  overnight Tc monitoring – hypercapnia

•ABGA - commonly used to monitor ventilatory status in most clinical settings

•Overnight Tc monitoring should be considered to assess latent CO2 retention

Considerations for management

Pathophysiology of depressed central ventilator drive

The problem of oxygen supply

Unnecessary tracheostomy

Pathophysiology of depressed central ventilator drive

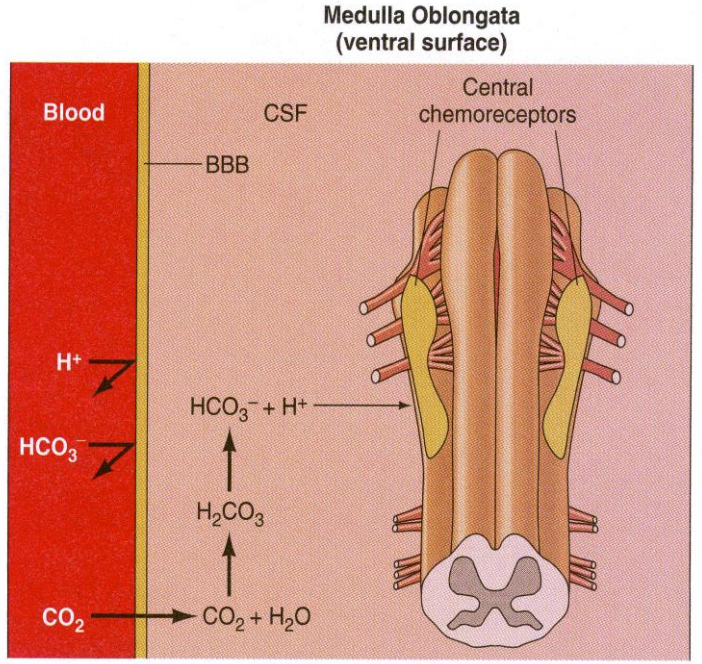
Ventilatory insufficiency
Hypercapnia **NOT** caused by *intrinsic lung disease*

→ ↑Hypercapnia (Acidosis)

→its associated Symptoms & dangers

→Compensatory Metabolic Alkalosis

→Depresses central ventilator drive



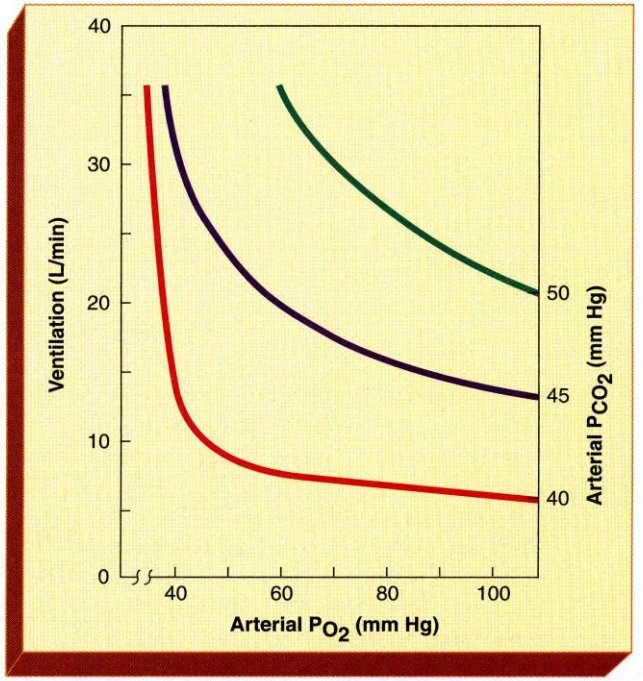
Alkalosis
→ Brain to accommodate the hypercapnia
without overt Sx of acute ventilator failure

The problem of oxygen supply

For Hypoxic patients with intact respiratory musculature
- Decrease ventilation, relieve respiratory muscle strain, Sx

For hypercapnic patients
- Exacerbation of hypoventilation

With great care to hypercapnic pts
with oxygenation impairment.
Not be administered to patients
with primarily ventilatory impairment.



Unnecessary tracheostomy

Patient with respiratory muscle weakness (common Tx pathway)

Acute event - supportive ventilation through intubation

→ after resolution of Complication

: extubation, no assist ventilation

: not bearable whole respiration already

→ fatigue develop

→ respiratory failure again

→ supportive ventilation through intubation (reintubation)

repeat this situation

- Recommand tracheostomy as a long term ventilatory support

Tracheostomy is indicated only when:

- Depressed cognitive function,
- Orthopedic conditions interfering with the use of IPPV interfaces and exsuffiation techniques
- Inadequate oropharyngeal muscle strength (bulbar palsy) → aspiration
- Severe intrinsic pulmonary disease necessitating high FiO_2
- Uncontrolled seizures or substance abuse
- Assisted PCF not exceeding 160 L/min
- The presence of a nasogastric tube

Understanding for treatment techniques

Expiratory muscle aids
Inspiratory muscle aids

Pulmonary Rehabilitation

consisted of breathing retraining, respiratory muscle rest, airway secretion elimination, reconditioning exercise, psychosocial support, nutritional support, adequate ventilator support, and patient education

Among various techniques and devices, the point of pulmonary rehabilitation is the non-invasive respiratory care by using respiratory muscle aids.

- ✓ Expiratory muscle aids
- ✓ Inspiratory muscle aids

Expiratory muscle aids

- Expiratory muscle aids are either a maneuver or a mechanical device used to remove endotracheal secretions.

- There are diverse methods and devices for managing airway secretions.

Four general approaches

(1) Postural drainage therapy
(including turning, percussion, and vibration),
(2) Positive expiratory pressure (PEP) therapy
(3) High-frequency compression/oscillation methods
(4) Coughing and related expulsion techniques

Positive Expiratory Pressure Therapy

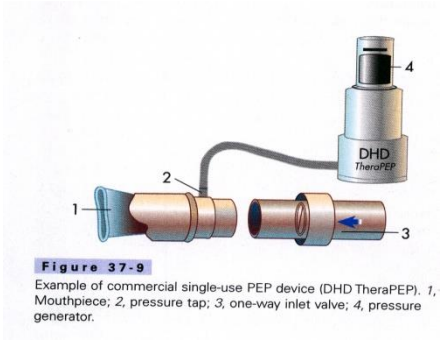
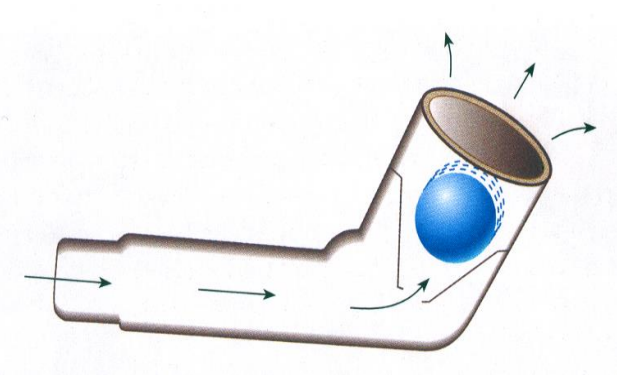


Figure 37-9
Example of commercial single-use PEP device (DHD TheraPEP). *1*, Mouthpiece; *2*, pressure tap; *3*, one-way inlet valve; *4*, pressure generator.

Flutter Valve



High Frequency Chest wall Oscillation(HFCWO)

Intrapulmonary Percussive Ventilation

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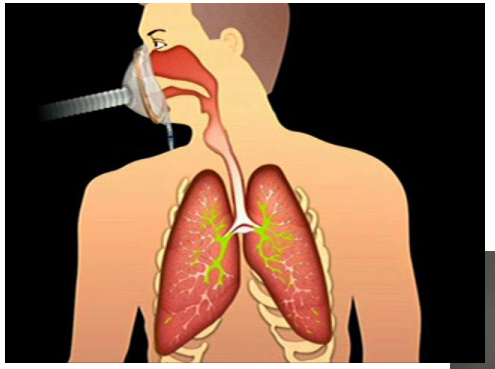
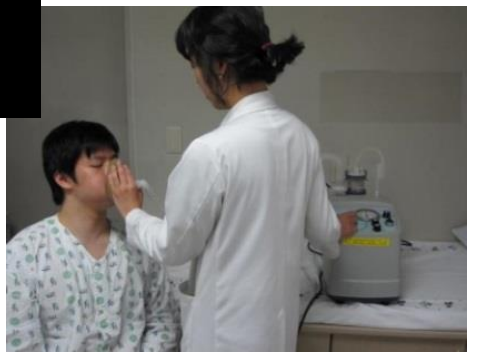
Mechanical Insufflation-Exsufflation (Cough Assists)





Introduction of MI-E

- Deep insufflations ► immediately deep exsufflations
- Insufflation to exsufflation pressures of +40 to -40 cmH_20
  - usually the most effective and preferred by most patients
- Interface : oral-nasal mask, a simple mouthpiece tracheostomy tube

Indication of MI-E

only take place of the inspiratory and expiratory muscles

Bulbar muscle dysfunction(inadequate to prevent airway collapse)?

NOT indicated to be used to avert tracheostomy

→ **most non-bulbar ALS/NMD patient & DMD with scoliosis**
(who can generate 250~300 L/m assisted PCF)

Technique

- 1 Cycle
  - Positive-pressure of 40 (35 to 50) cm H20 over a 1 to 3 sec.
  - reversed to -40(-35 to -50) cm H20 for 1 to 2 sec.
  - rest for 4 to 5 sec
- One treatment session ; 5 cycles of MI-E
- Important point (keep in mind~!!)
  - rapid maximal chest expansion followed immediately by rapid lung emptying
  - both in about 1-3 seconds
- abdominal thrust should be timed to the exsufflation cycle.

Advantage

- Non-invasive method (clear secretion via oronasal mask)
- Cough flow is much higher than other manual assisted coughing techniques
- Eliminates the irritation and/or damage to the airway
- No wound site pain when applied postoperatively.

Inspiratory muscle aids

Respiratory dysfunction in patients with respiratory muscle weakness
Key factor of management - respiratory muscle **fatigue**

Respiratory complications occur in these patients

→ respiratory work load increases

→ induce decompensation of respiratory muscles **due to fatigue**

Tx: supportive ventilation to avoid respiratory muscle fatigue

If not

→ the abrupt fall in pH.

→ acute respiratory failure

Ventilatory assist method

Usually by invasive method (intubation or tracheostomy)

- Psychological burden
- Hesitate and postpone initiating treatment

Non-invasive method

- More easily acceptable
- Sufficient to resolve hypercapnia and its associated symptoms
- Improve the quality of life

Indications of Non-invasive ventilator assist

Acute care setting

- COPD exacerbations
- Hypoxemic respiratory failure
- Do-not-intubate orders
- Postoperative respiratory failure
- Facilitation of extubation and weaning, etc.

Among the those indicated situations, **restrictive thoracic diseases** most successfully managed with NIV.

Ex) Neuromuscular disease, spinal cord injury, severe kyphoscoliosis, post-polio syndrome..

Chronic care setting

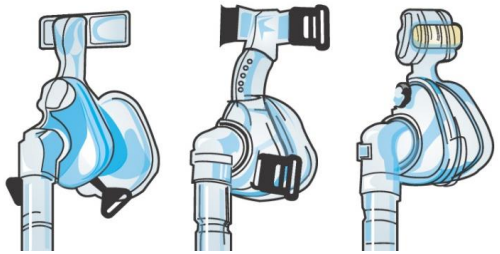
- Restrictive disease
- Nocturnal hypoventilation

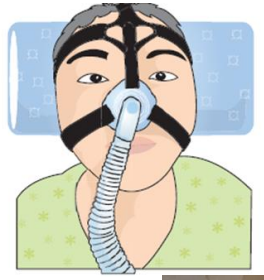
Portable positive pressure Ventilator

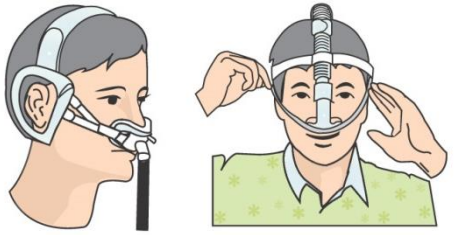
- Volume-limited ventilator (LTV series, CARAT, LEGENDAIR, PB560, Astral and Trilogy)
- Pressure- limited ventilator (BiPAP)

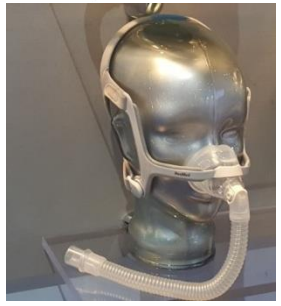
Interface (type)

- Nasal interface; nasal mask, nasal pillows
- Face masks ;Oronasal masks, total full face masks
- Hybrid
- Mouthpiece /lipseal









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To ensure the success of NIV

close monitoring is necessary,

- especially of respiratory rate (patient's effort),
- oxygen saturation (to adjust FiO2) and pH
- PaCO2 or ETCO2 or tcPCO2
- Alarms

the key factors determining **tolerance to NIV** (and its success)

- **optimal synchrony between the patient's spontaneous breathing activity and the ventilator's set parameters**, known as "patient–ventilator interaction"
- Asynchrony ; air leakage is involved in many of them

Monitoring of Effectiveness

- Physician reassessment of patient adherence with use of NIV at 60~90 days
- Ongoing monitoring(3개월 외래 추적) and yearly recertification (1년마다 재입원)


Thank you