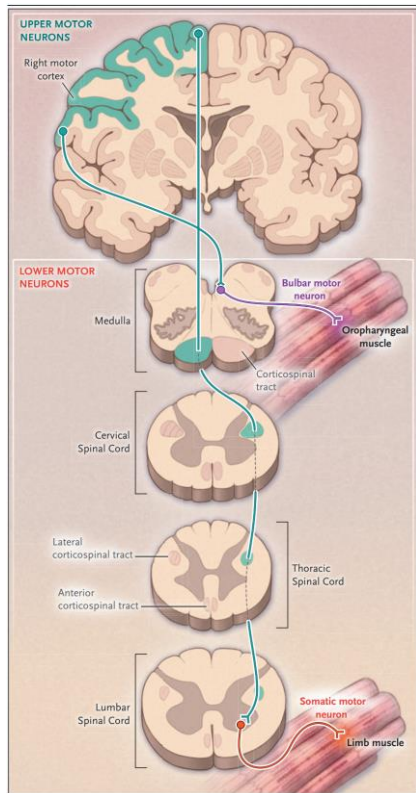


Amyotrophic Lateral Sclerosis

2023년 2월 15일

국민건강보험 일산병원
재활의학과 이장우

Classification of motor neuron diseases



• According to involved motor neurons

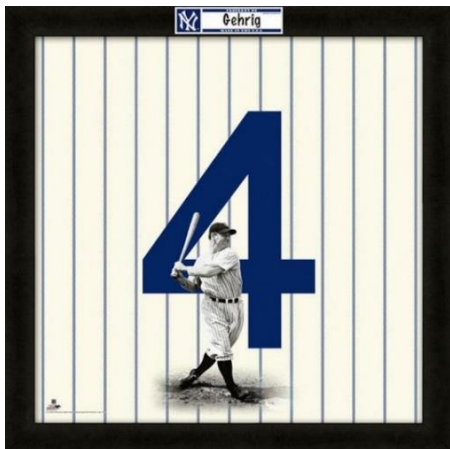
- Mixed Upper and Lower Motor Neurons
 - Amyotrophic lateral sclerosis (ALS)
 - ALS-Plus
- Upper Motor Neuron
 - Primary lateral sclerosis
- Lower Motor Neuron
 - ALS-related variants with predominant lower motor neuron loss
 - Progressive muscular atrophy
 - Progressive bulbar palsy
 - Brachial amyotrophic diplegia (flail arm syndrome)
 - Leg amyotrophic diplegia (flail leg syndrome)
 - Acute poliomyelitis and postpolio syndrome
 - Spinal muscular atrophy
 - Proximal
 - Distal (hereditary motor neuropathy)
 - Scapulo-peroneal SMA (Davidenkow syndrome)
 - Kennedy disease (X-linked spinobulbar muscular atrophy)
 - Hirayama disease (monomelic spinal muscular atrophy)
- Less Common or Less Well-Defined Syndromes
 - Paraneoplastic
 - Non-polio-related infections
 - Electrical injury
 - Hereditary metabolic disorders
 - Hereditary bulbar syndromes

• According to etiology

- Acquired
 - ALS and ALS variants
 - Poliomyelitis
 - Rare causes
 - Immune-mediated, endocrine, traumatic, nonpoliomyelitis infections, paraneoplastic
- Inherited
 - SMA
 - Familial ALS
 - Kennedy disease

Lou Gehrig

- New York Yankees (1923-1939)
- “The iron horse”
 - 2,130 consecutive games for 14 years
- 23 grand slams
- The first retired number in MLB history (No. 4)





기부상품 위드아이스 루게릭병 환우 등록

희망의 끈을 이어갑니다
승일희망재단

해피빈 블로그 페이스북 인스타그램 유튜브

| 사업소개 | 참여하기 | 루게릭병 | 재단활동 | 재단소개 |
|--------------|------------|-------------|------------|------------------------|
| 루게릭요양센터 건립사업 | 기부참여 | 루게릭병이란? | 공지사항 | 재단 이야기 |
| 환우 지원사업 | 캠페인 참여 | 환우와 가족의 이야기 | 재단소식나눔 | 히스토리 |
| 기부상품 수익사업 | 기업/단체 기부 | 루게릭병 가이드북 | 행사 이야기 | 아이스버킷 챌린지 |
| | 호프소사이터터 가입 | 유용한 사이트 | 영상스토리 | 함께하는 사람들 |
| | 기부 FAQ | 루게릭병 관련뉴스 | 언론보도자료 | 엠버서더 팅킷 |
| | 기부금영수증 안내 | | 투명 경영/재정공시 | |



루게릭병 환우와 가족에게 그리고 당신에게 희망의 끈이 이어지길 바랍니다.

승일희망재단 공동대표 박승일입니다.

2002년 농구인에서 루게릭병 환우로서 인생의 터닝 포인트를 만난 지 어느새 20년이 지났습니다. 루게릭병 확진 후, 저와 같은 루게릭병 환우를 위해 루게릭요양센터 건립이라는 꿈을 가지게 되었습니다. 희망, 기적! 세상에 존재하는 단어일 텐데! 그냥 사람이 만들어낸 단어가 아닐 텐데! 라고 말하며 마음속에 다짐하였지만, 혼자 힘으로는 절대 불가능했을 그 희망이 2011년 승일희망재단을 설립하면서 점점 현실이 되어가고 있습니다. 승일희망재단이 저에게 희망이 되어 주었던 것처럼 지금 이 시간에도 절망 속에서 살아가는 루게릭병 환우와 가족에게도 희망이 되길 바랍니다.

공동대표 박승일 올림

승일희망재단 공동대표 선입니다.

2009년 국내 최초의 루게릭요양센터 건립이라는 꿈을 가진 루게릭병 환우 박승일을 만난 인연이 2011년 승일희망재단 설립의 시작이었습니다. 그의 꿈을 함께 만들어가겠다는 약속을 지키기 위해 저는 이 자리에 있습니다. 루게릭병에 대해 깊이 알게 될 수록 이 일이 얼마나 절실히 필요한 일인지 또 얼마나 속히 이루어져야 하는지를 깨닫게 되었습니다. 루게릭병이 완치되는 그날까지 승일희망재단이 루게릭병 환우와 가족에게 힘이 되어 저와 여러분을 통해 희망에서 기적으로 이어지길 간절히 바랍니다. 감사합니다.

공동대표 선 올림



Amyotrophic Lateral Sclerosis

- Most common adult MND
- A rapidly progressive neurodegenerative disease
 - Destruction of motor neurons in the primary motor cortex, brain stem, and anterior horn cell of the spinal cord
 - Mean life expectancy of 3 years following the onset of symptoms
- Incidence 1.68/100,000, prevalence 6.49 per 100,000 (KNHIS data, Kim JM et al. 2018)
- Insidious onset and most commonly presents with **painless asymmetric weakness**
- 40-60 years (mean onset 58yrs), 5% before 30yrs
- M:F = 1.6:1

Sporadic ALS

- **Glutamate excitotoxicity** and **oxidative stress** → pathogenesis of ALS
- Putative mechanisms
 - Mitochondrial dysfunction, cytoskeletal derangements, and microglial activation
- Unknown etiology, likely multifactorial
 - Smoking cigarettes; dose-dependent
 - Military service
 - Traumatic brain injury
 - Professional athletes and athletic populations (vs. protective effect of exercise)
 - Toxins: environmental toxins, cyanobacteria, insecticide
 - Heavy metals
 - History of trauma
 - Electrical shock
 - Human endogenous retrovirus K

Familial ALS

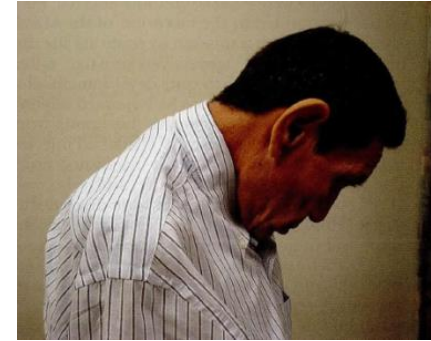
- 5~10% of all ALS
- AD (most common), AR, X-linked and mitochondrial inheritance have been reported
- Age of onset occurs a decade earlier than sporadic ALS
- More rapid progression than sporadic form

Clinical features

- Limb weakness
 - UMN
 - Loss of dexterity or a feeling of stiffness
 - Spasticity
 - LMN
 - Muscle atrophy, fasciculations, muscle cramping
 - Cramping on abdominal or trunk muscles → red flag
- Bulbar weakness
 - Caused by UMN and/or LMN
 - Dysarthria
 - Spastic (UMN): strained, strangled quality of speech, reduced rate, low pitch, vowel distortion, breaks in pitch, imprecise consonant pronunciation
 - Flaccid (LMN): nasal or wet quality speech, monotonous, short phrase, inspiration audible
 - Dysphagia, drooling, aspiration, nasal regurgitation, chewing difficulty
 - **Tongue should be examined** for fasciculation and atrophy, strength & ROM
- Cognitive or behavioral impairment
 - A part of ALS/FTD spectrum
 - Can precede muscle weakness

Clinical features

- Head drop
 - Because of weakness of neck extensor → soft neck collar
 - ALS and MG are 2 most common cause of head drop
- Respiratory system
 - Orthopnea, sleep-disordered breathing, nocturnal hypoventilation
 - Air hunger, snoring, choking episodes, restlessness, insomnia, daytime hypersomnolence, morning headache, drowsiness, fatigue
- Cachexia
 - Muscle atrophy, Wt. loss, low caloric diet, BMR↑
- Muscle fatigue → Recurrent events of sprain
- Ocular muscle and anal sphincter (Onuf's nucleus) are typically spared



Clinical features

Table 1. Characteristics of the Study Subjects

| | Number (%) | Onset age (ys)* | <i>p</i> value | Onset to diagnosis (months)* | <i>p</i> value |
|-------------------|------------|-----------------|---------------------|------------------------------|--------------------|
| Overall | 500 | 57.9±11.9 | | 12.7±11.9 | |
| Sex | | | 0.593 [†] | | 0.004 [†] |
| Male | 314 (62.8) | 58.1±11.6 | | 11.5±10.3 | |
| Female | 186 (37.2) | 57.5±12.6 | | 14.9±14.0 | |
| Age (yr) | | | | | 0.985 [†] |
| <60 | 266 (53.2) | | | 12.7±12.5 | |
| ≥60 | 234 (46.8) | | | 12.7±11.2 | |
| Onset type | | | <0.001 [†] | | 0.005 [†] |
| Limb | 335 (67.0) | 57.5 (15.9) | | 10.0 (13.0) | |
| Bulbar | 144 (28.8) | 60.6 (14.1) | | 8.5 (10.0) | |
| Respiratory | 21 (4.2) | 66.7 (14.0) | | 6.0 (7.0) | |
| Operation | | | 0.053 [§] | | 0.008 [§] |
| With operation | 43 (8.6) | 61.8 (12.8) | | 11 (17) | |
| Without operation | 457 (91.4) | 58.3 (16.3) | | 9 (12) | |

*Onset age and the period between onset to diagnosis are presented as means±standard deviations or medians (interquartile rage), [†]Independent t-test, [‡]Kruskal-Wallis test, [§]Mann-Whitney U test.

Table 2. Comparison of Onset Age and Time between Onset and Diagnosis among Limb-, Bulbar-, and Respiratory-Dominant Onset Groups

| | Limb (n=335)* | Bulbar (n=144)* | Respiratory (n=21)* | Limb vs. Bulbar [†] | Bulbar vs. Respiratory [†] | Respiratory vs. Limb [†] | Among 3 groups [†] |
|---|---------------|-----------------|---------------------|------------------------------|-------------------------------------|-----------------------------------|-----------------------------|
| Age of symptom onset (yr) | 57.5 (15.9) | 60.6 (14.1) | 66.7 (14.0) | 0.012 | 0.159 | 0.006 | <0.001 |
| Time between symptom onset and diagnosis (months) | 10.0 (13.0) | 8.5 (10.0) | 6.0 (7.0) | 0.105 | 0.204 | 0.018 | 0.005 |

*Median (interquartile range), [†]*p* values from Kruskal-Wallis test with Bonferroni correction.

Table 3. Operation Sites before Diagnosis of Amyotrophic Lateral Sclerosis

| Operation site | Number of patients (n=43) | Notes |
|---------------------------|---------------------------|--|
| Cervical spine | 17 | Twice in one patient, and three times in one patient |
| Cervical and lumbar spine | 1 | |
| Lumbar spine | 20 | Twice in one patient, and three times in one patient |
| Carpal tunnel release | 5 | Cervical spinal operation after carpal tunnel release in one patient |
| Cubital tunnel release | 1 | |

Statistics in the prognosis in ALS

- Average survival from onset of symptoms: 2~3 years
- From diagnosis
 - Average survival: 15 months
 - Three-year survival: 40-50%
 - Five-year survival: 10-20%
- Poor prognosis
 - Female
 - More bulbar symptom, the progression appears to be more rapid
 - Increasing age
 - Bulbar onset
 - Predominance of LMN findings at the time of diagnosis
 - More definite diagnostic category
- Treatment factors impacting prognosis
 - Percutaneous endoscopic gastrostomy
 - Noninvasive ventilation
 - Riluzole
 - Participation in multidisciplinary clinics

Pharmacologic Management

Riluzole

- 2-amino-6-(trifluoromethoxy) benzothiazole
- Mechanisms
 - Interference with *N*-methyl-D-aspartic acid (NMDA) receptor-mediated responses
 - Stabilization of the inactivated state of voltage-dependent sodium channels
 - Inhibition the presynaptic release of **glutamate**
 - Activation of extracellular **glutamate** uptake
- Disease modifying agent, survival prolongation
 - Cochrane review (2012)
 - Tracheostomy free survival: 2~3 months ↑
- Recommended to nonventilated ALS
- Side effects
 - Asthenia, nausea
 - ALT elevation
 - ALT should be monitored monthly for the first 3 months and then every 3 months
 - Riluzole should be discontinued if the ALT reaches 5 times the upper limit of normal

Others

- Edaravone (Radicut[®])
 - Approved by the FDA for ALS (2017.5)
 - Free radical scavenger → degeneration of motor neuron ↓
- Neuronata R[®]
 - Significantly less reduction of ALSFRS-R score
- Medical cannabis
 - Limited evidence, but potential disease-modifying
 - Analgesia, antispasticity, muscle relaxation, bronchodilation, saliva reduction, appetite stimulation, sleep induction

Rehabilitation Management

Rehabilitation

- Stage 1
 - Ambulatory & fully independent with mild weakness
 - Strengthening with caution for overwork damage
- Stage 2
 - Ambulatory & independent with moderate weakness
 - Functional impairment in some areas
 - Orthoses
 - AFO, wrist-hand orthosis, opponens splint
 - Cervical orthoses
 - Assistive devices
 - Work simplification
 - Strengthening with caution for fatigue



Rehabilitation

- Stage 3
 - Ambulatory, but severe weakness in selected muscle groups
 - Walker, W/C for long trip
 - Elevated toilet seat
 - Deep breathing exercise with mild respiratory muscle weakness
- Stage 4
 - Nonambulatory but remain independent
 - E/W/C, hospital bed
 - Exercise of unaffected muscles, ROM exercise

Rehabilitation

- Stage 5
 - No longer independent
 - Transfer assistance, wheeled shower chair
 - Prevention for pressure ulcer
 - Family education
- Stage 6
 - Completely bed-ridden
 - Proper positioning

Exercise

- Moderate aerobic and gentle strengthening
- Aerobic exercise
 - Recommended for ALS
 - Pool therapy, low impact of walking or stationary bicycling
 - Improvement of cardiovascular performance
 - Physical benefits, additional effects on mood, psychological well-being, appetite, sleep and pain tolerance

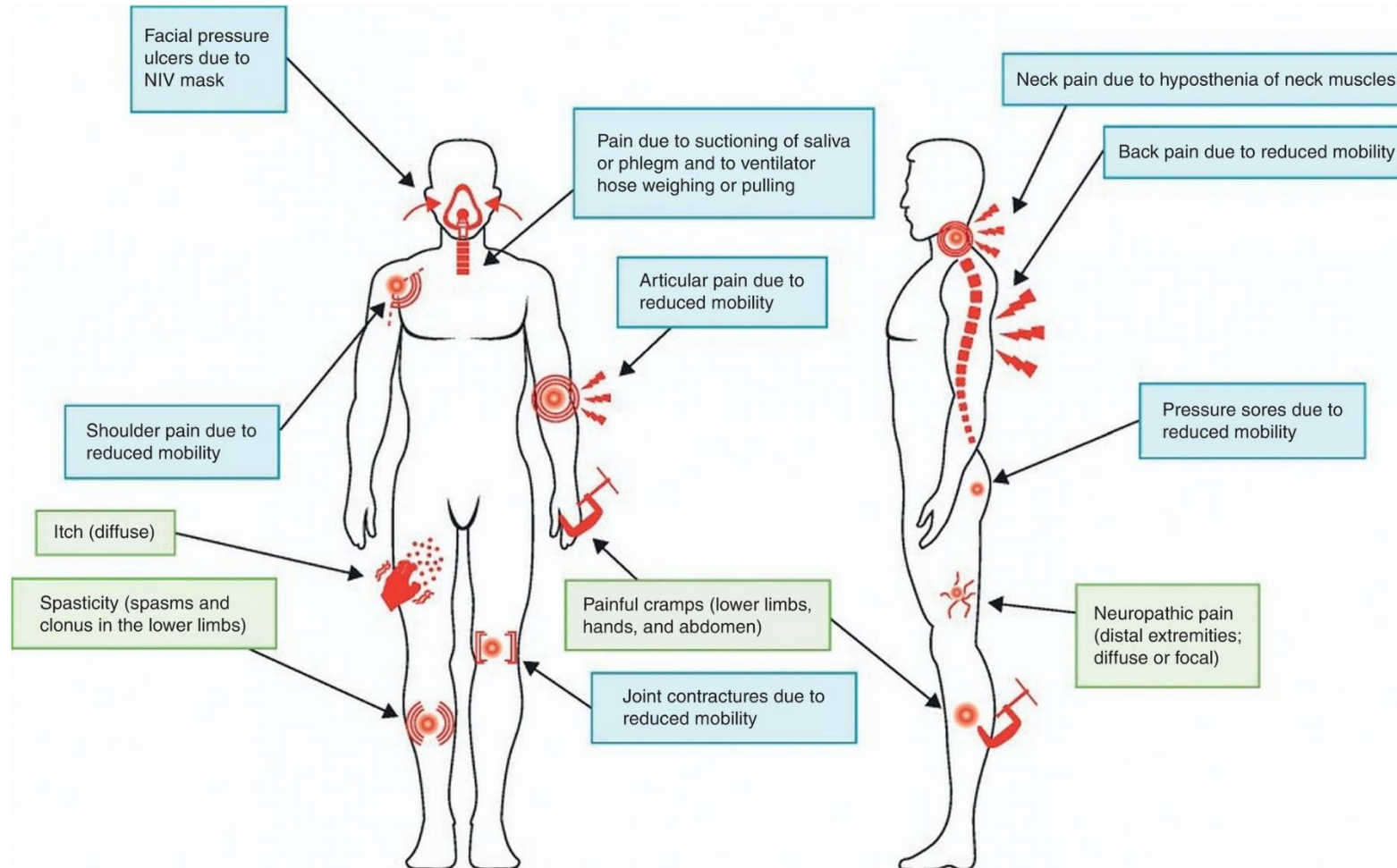
Exercise

- Strengthening exercise in ALS
 - Moderate intensity resistance exercise (Cochrane review 2008)
 - Functional improvement (ALSFERS)
 - No significant difference in QOL, fatigue, or muscle strength
 - With $\geq 3/5$ strength on the MRC scale
- ROM and stretching exercise
 - Should be prescribed
- The risk for **overwork weakness** should be considered
 - Exhaustion → muscle damage and dysfunction
 - Signs of overwork weakness
 - Feeling weaker rather than stronger within 30 minutes
 - Excessive muscle soreness 24 to 48 hours following exercise
 - Severe muscle cramping, heaviness
 - Prolonged shortness of breath

Spasticity

- Pharmacologic management
 - **Baclofen** (TOC): GABA receptor agonist
 - Intrathecal baclofen pump
 - Tizanidine: α -2 agonist
 - **Benzodiazepines: risk of respiratory depression**
 - **Dantrolene: blockage of Ca^{2+} release in SR, not recommended d/t generalized muscle weakness**
 - Botulinum toxin: caution for generalized weakness
- Stretching
- Positional splinting

Pain



Pain

- Usually musculoskeletal pain d/t immobilization
- Aggressive identification of the source of pain
- Cervical collar
- W/C
 - Lumbar support and good cushioning
 - Head support or reclining lounge (more comfortable than a collar)
- Pressure relieving mattress
- IM should be avoided because of muscle wasting
- Consideration for respiratory depression and constipation when opioids are administered
 - Oral sublingual morphine → might help relieve “air hunger” in the terminal stage of ALS
 - Marijuana
 - Effective in pain, muscle spasm, loss of appetite, secretion control, depression

Dysarthria

- Speech therapy
 - Early stage, slowly progressive deterioration
 - Correct poor compensatory strategies

| ㄱ | | ㄷ | | ㅂ | | ㅅ | | ㅈ | |
|---------|---|--------|---|----------|---|----------|---|-------|---|
| ㄱ | ㄴ | ㄷ | ㄹ | ㅂ | ㅅ | ㅇ | ㅋ | ㅌ | ㅍ |
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| ㄷ | ㄹ | ㅂ | ㅅ | ㅇ | ㅋ | ㅌ | ㅍ | ㅈ | ㅊ |
| YES | | | | 숫자대화 | | NO | | | |
| [Black] | | [Blue] | | [Yellow] | | [Purple] | | [Red] | |
| 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 0 |

| | | | | | | | | | | | | | | | | |
|----------|---|-------|---|---|---|---|---|---|---|---|-----------|---|---|---|---|---|
| 초성 | ㄱ | ㄴ | ㄷ | ㄹ | ㅂ | ㅅ | ㅇ | ㅋ | ㅌ | ㅍ | ㅎ | ㄱ | ㄷ | ㅂ | ㅅ | ㅈ |
| 모음 | ㅏ | ㅑ | ㅓ | ㅕ | ㅗ | ㅛ | ㅜ | ㅠ | ㅡ | ㅣ | ㅏ | ㅑ | ㅓ | ㅕ | ㅗ | |
| 받침 | ㅂ | ㅅ | ㅇ | ㅈ | ㅊ | ㅌ | ㅍ | ㅎ | ㄱ | ㅅ | ㄷ | ㄹ | ㅂ | ㅅ | ㅇ | |
| 예 YES | | 숫자 대화 | | | | | | | | | 아니오 NO | | | | | |
| | | 0 | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | | | | | |

Dysphagia

- Malnutrition
 - Etiology
 - Inability to feed oneself
 - Depression
 - Bulbar muscle dysfunction and dysphagia
 - Increased metabolic demands
 - Low BMI & rapid weight loss → rapid progression
 - 7.7-fold increased risk of death
- Treatments
 - Initial stage
 - Compensatory strategies, dietary modification, high-calorie liquid supplement
 - Gastrostomy
 - AAN recommends PEG, when FVC \geq 50%
 - RIG is easier and safer to insert in patients with low FVC
 - Stabilization of weight and **prolongation of survival**
 - Indications
 - Aspiration pneumonia, >10% loss of body weight, prolonged feeding time

Dysphagia

- Using **NIPPV** during tube placement appears to improve safety even if ALS patients with an FVC of less than 50% of the predictive
- Sialorrhea
 - Due to inadequate handling of secretion rather than the amount of secretion
 - Treatments
 - Home suction machine, amitriptyline, atropine drops, Botulinum toxin into parotid glands, radiation...
- Continuous oral suction

Dysphagia



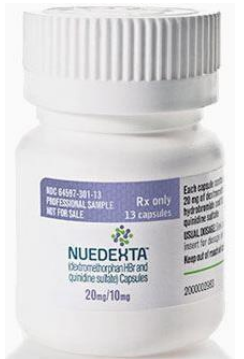
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Nutrition

- Weight loss
 - Muscle wasting, hypermetabolism, respiratory failure, dysphagia, reduced intake
 - Low BMI & rapid weight loss → rapid progression

Pseudobulbar affect

- Common condition in ALS (about 50%)
- UMN syndrome is caused by disinhibition of limbic control
 - Result of loss of frontal lobe inhibition of spontaneous brain stem-generated emotional responses
- Medications
 - Antidepressants
 - Levodopa, lithium
 - **Nuedexta®**
 - Dextromethorphan (NMDA antagonist) + quinidine
 - Approved by FDA
 - Improved symptoms and QoL without anticholinergic S/E
 - But, unclear mechanism and high rate of side effects (nausea, dizziness, somnolence)



Mood and Cognitive disorders

- Depression
 - **Amitriptyline**
 - Effective for pseudobulbar affect and decline amount of saliva
- Anxiety and insomnia
 - **Benzodiazepines → caution for respiratory suppression**
- Cognitive impairment
 - Mild to moderate impaired cognition: 50% in ALS
 - Frontotemporal dementia
 - 15% of ALS
 - Apathy, change in emotional reactivity, sleep disturbance, stereotypical or repetitive behaviors, poor insight or judgment, loss of expressive language with relative preservation of comprehension

Respiratory management in ALS

- Prevention for aspiration
 - Anticholinergics and dietary manipulation for saliva ↓
 - Proper swallowing technique
 - Gastrostomy and tracheostomy
- For coughing
 - Mechanical in-exsufflation
 - Assist coughing
- CO₂ retention
 - Predicting when respiratory failure will occur is important
 - FVC<50%: careful F/U
 - Every 3 months F/U
 - FVC_{sup}<FVC_{sit}
- NIPPV
 - Survival and QOL ↑
- Tracheostomy

Respiratory muscle weakness

- Respiratory muscle weakness
 - Ventilatory insufficiency
 - Decreased coughing ability
- Consideration for ventilatory support
 - In patients with intact respiratory muscle
 - **Disuse atrophy**
 - In patients with respiratory muscle weakness
 - **Overuse weakness, fatigue**

Ventilator as a brace

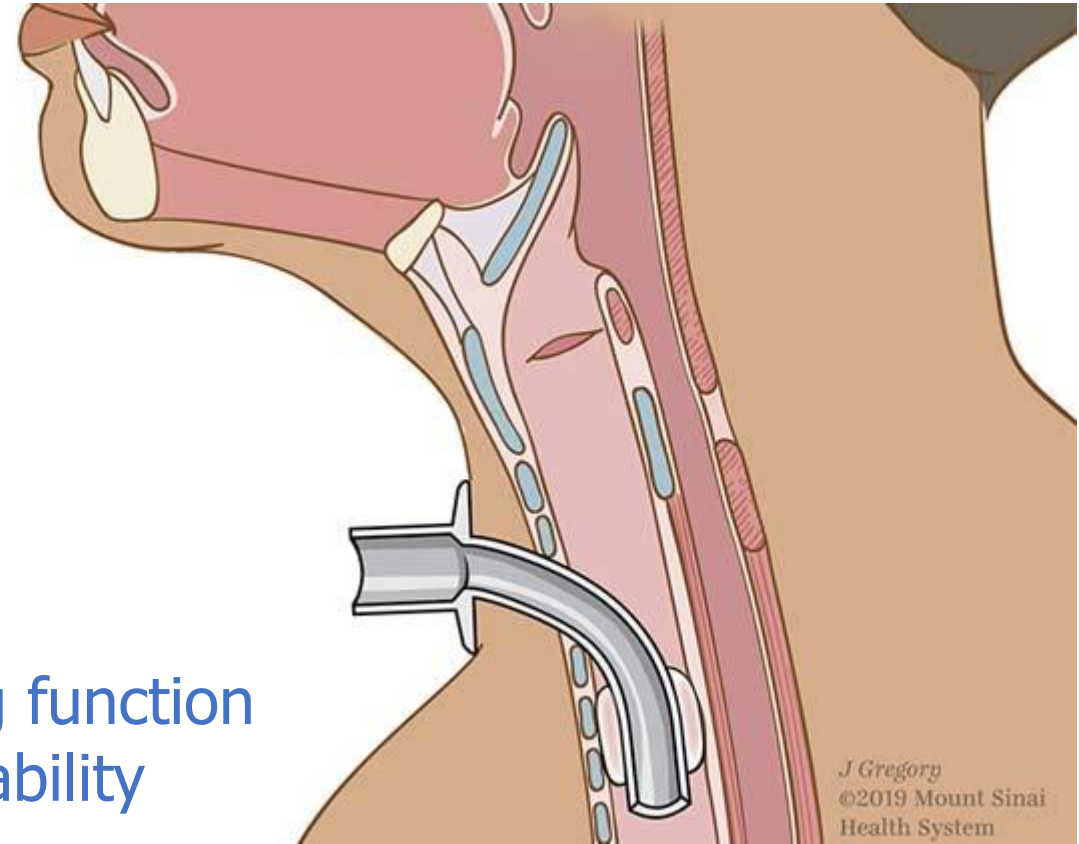


Non-invasive ventilator



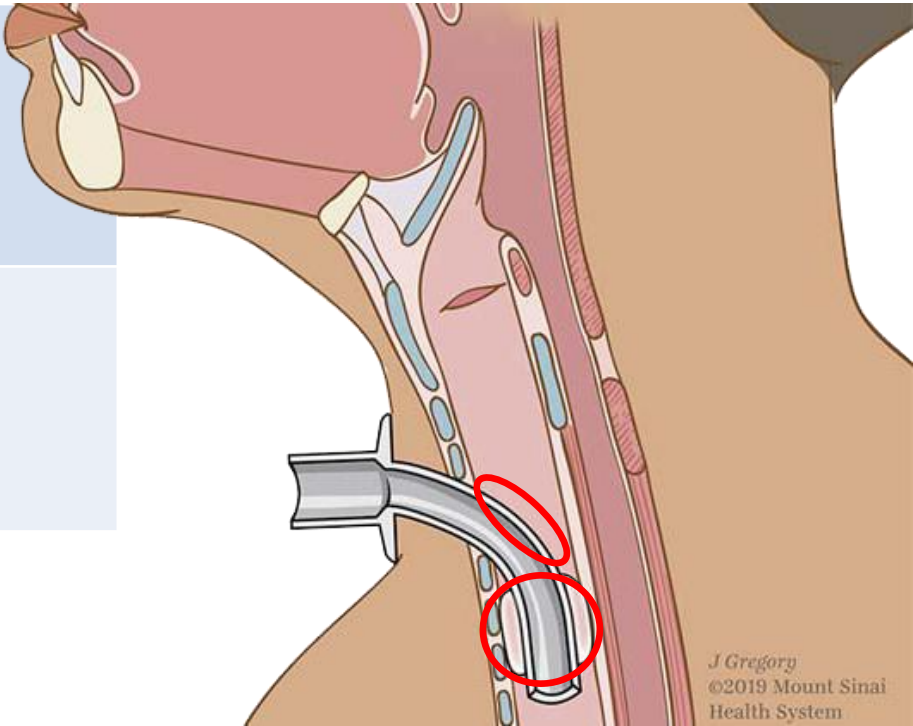
Tracheostomy

- Tracheostomy의 가장 중요한 기능
= lung protection
 - Aspiration 예방 → Swallowing function
 - Secretion 관리 → Coughing ability

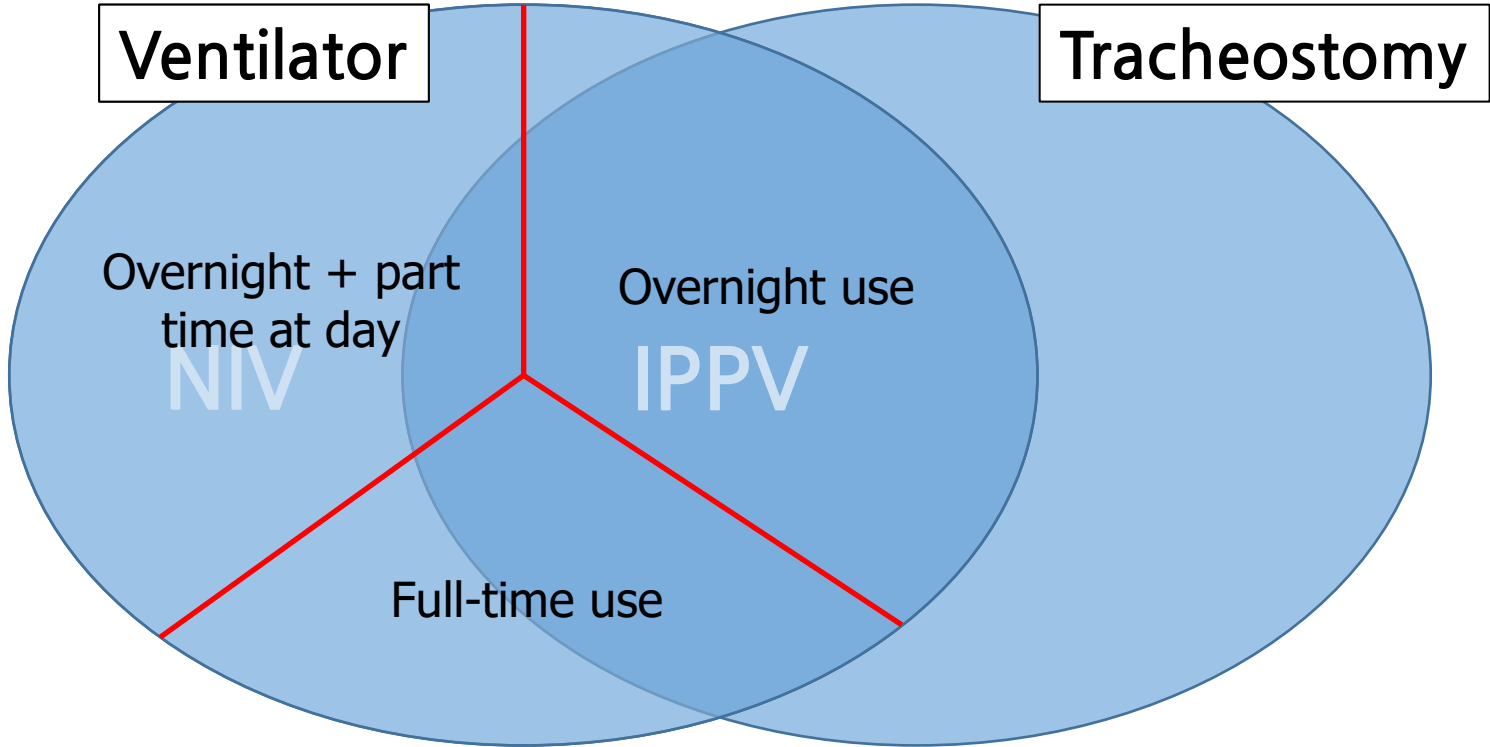


Tracheostomy tube

| | Fenestration (-) | Fenestration (+) |
|----------|------------------|------------------|
| Cuff (+) | | |
| Cuff (-) | | |



Ventilatory care



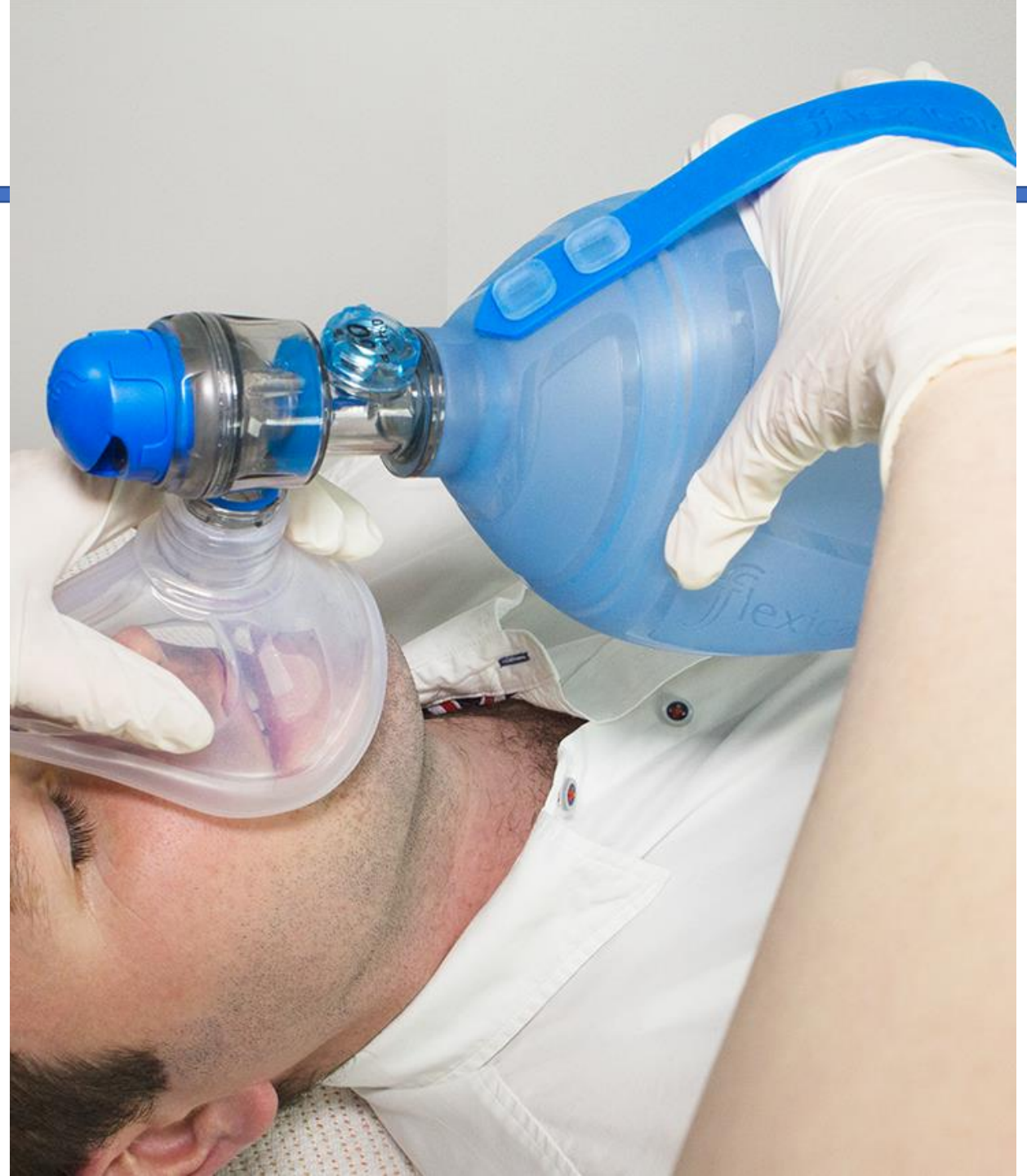
Relative contraindications for long-term NIV

- Lack of cooperation or use of heavy sedation or narcotics
- Need for high levels of supplemental oxygen therapy
- SpO₂ cannot be maintained above 94% despite NIPPV and optimal use of assisted coughing techniques
- Substance abuse or uncontrollable seizure
- Bulbar-innervated muscle impairment with inability to close the glottis → [airway protection \(-\)](#)
- Inadequate caregiver support
- Conditions that interface with the use of NIPPV interfaces. i.e., facial fractures, inadequate bite for mouth piece entry, presence of facial hair that hamper airtight interface seal

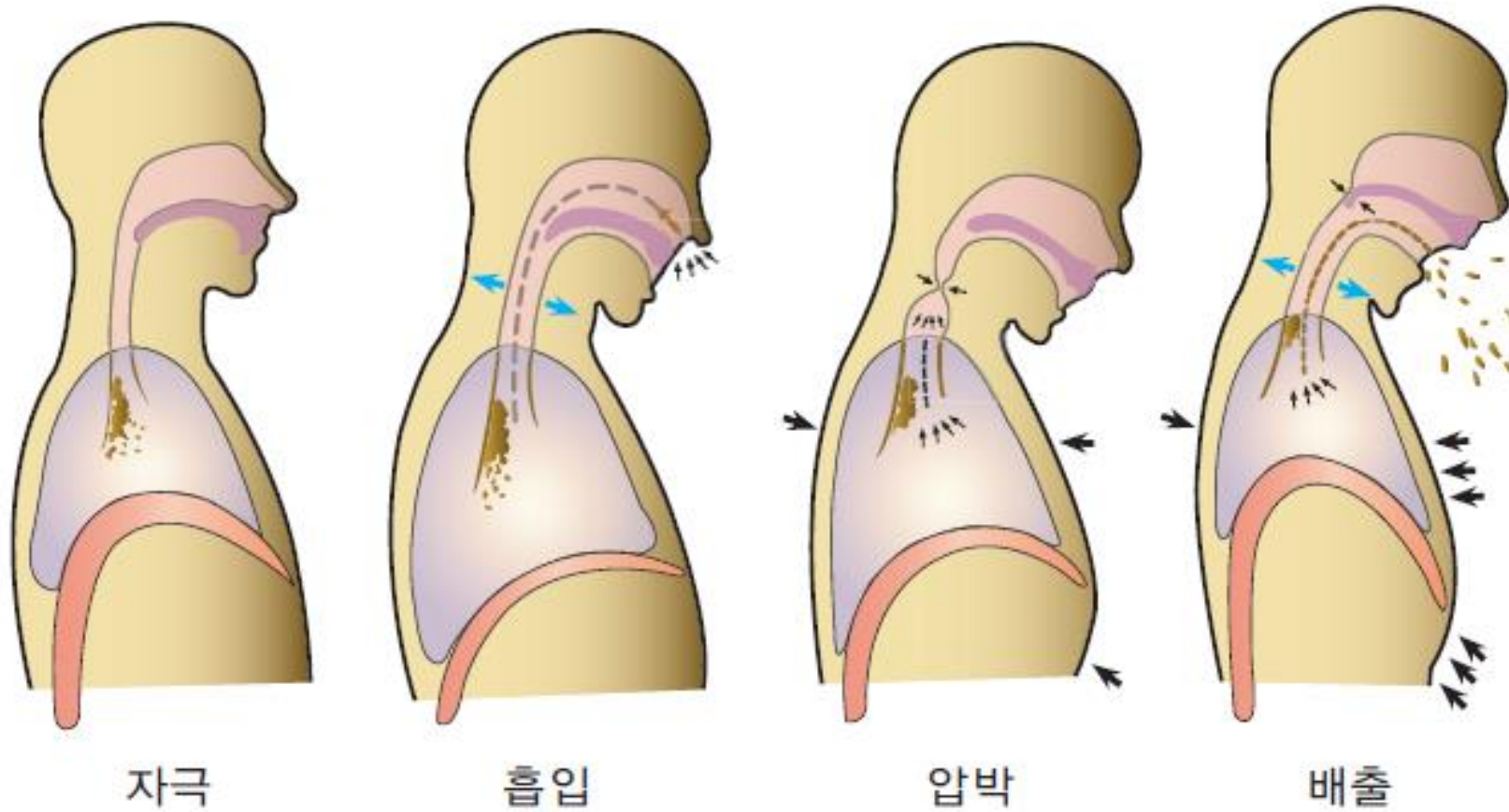
- NG-tube (?)
- Assisted PCF not exceeding 160 L/min (??)

Ventilator 관리

- Circuit 관리
- Monitoring
- Ventilator에 문제가 생기면?



Coughing mechanism



Secretion management

- Assist coughing
- Mechanical in-exsufflator
- Positive expiratory pressure ventilation
- Mucus loosening and mobilization – intrapulmonary percussive ventilation, high frequency chest wall oscillation, Acapella[®], Flutter[®]

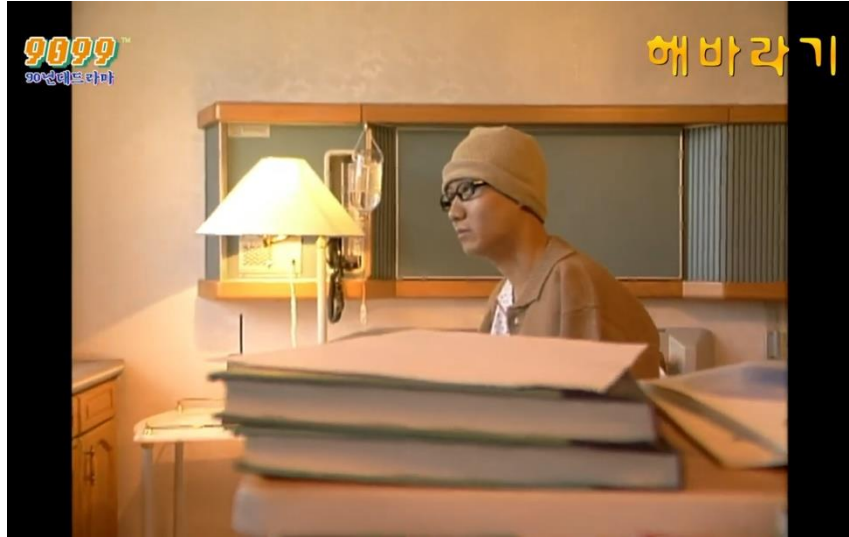
Manual assist cough

Cough machine (mechanical in-exsufflator, MIE)



MIE의 효과

- Non-invasive method
- Cough flow is much higher than manually assisted method
- Eliminates the irritation and/or damage to the airway
- No wound site pain when applied postoperatively
- Temporarily increase vital capacity
- Maintain oxygen saturation
- Effectively removes secretions from the lower airways and left upper lobe bronchus, where suctioning is more difficult



상됩니다. 병원을 떠난 지 3개월 하고도 이틀 째 되는 날입니다. 과장님, 제겐 새로운 버릇 하나가 생겼습니다. 정확한 날짜, 정확한 시간을 세는 습관입니다.

캐비닛 속에 넣어둔 수술도구들을 꺼내보았습니다. 오늘따라 그것들이 유난히 반짝입니다. 그리고 하얀 가운을 입고 복도를, 응급실을, 의국을, 수술실을 거니는 꿈을 꾸니다.

시간이 모든 걸 치료해준다는 말에 고개를 끄덕이는 사람은 진정 행복한 사람입니다. 죽음을 알리는 의사의 공허한 목소리를 들으면서 난 그가 나를 살려주길 원한 건 아니었습니다. 다만, 그가 내 손을 잡아주길 원했습니다. 죽음 앞에서 살끝으로 밀려드는 건 죽음의 공포라기보다는 차라리 죽어가는 외로움입니다. 그래서, 의사는 환자의 손을 잡아주어야 합니다. 그가 혼자가 아님을 손끝에서 느낄 수 있도록.

돌아가고 싶습니다. 과장님, 전 이제서야 의사가 되고 싶습니다. 내가 의사였던 그때를 까마득히 잊고 이제서야 의사가 되고 싶습니다.

의사는 쇠약해져가는 환자들을 대신해서 하늘을 바라보며 아부를 합니다. 환자들이 쏟는 피를 바치면서. 그리고 때론 험박도 합니다. 메스를 들이밀면서, 언젠가 이들을 주겠으니 지금은, 이들이 사랑하는 것을 사랑한단 말할 시간을 달라고. 충분한 시간을 달라고. 마치 하늘을 바라보는 해바라기처럼 의사들은 초라하게, 그리고 처절하게, 그리고 하염없이 하늘만 바라보는 존재임을 이제야 알게 되었습니다.

그리고 이젠 의사가 될 수 있을 것 같습니다.

이젠 의사가 될 수 있을 것만 같은데... 이젠...



가정방문 예시

- 방문시간 15:30~15:50
- Lung sound clear
- T-site 확인: oozning 없고 잘 관리되고 있음.
- 호흡기수첩 확인
 - 호흡기 회사에서 check했을 때 saturation 90%까지 측정됨
 - 방문 시에는 93~4% 정도로 유지
 - 24시간 saturation monitoring 유지하도록 권고
 - Saturation 저하소견 지속되면 외래에서 알려주시기로 함.
- Ambu bag 24시간 환자 옆에 prep하도록 함.
- 하루 3회 routinely MIE 사용하도록 권고함.

질문 목록

- 루게릭 환자의 경과(선천성 및 후천성)
- 근위축의 부위 순서
- 진행경과를 지연시킬 수 있는 예방조치가 있는지?
- 배뇨 및 배변 관리
- Swallowing difficulty를 지연시킬 수 있는 방법 및 재활관리
- 호흡곤란에 관한 관리(보조인공호흡기를 집에서 사용 시)
- 수면장애 및 불안, 공황증에 관한 관리
- 근육통에 관한 관리 및 치료

질문 목록

- 환자의 일상생활에 도움이 될 수 있는 조치(책읽기, 음악듣기, 영상관람 등 삶의 질에 관한 문제)
- 기타 방문진료의사가 환자에게 해줄 수 있는 사항
- 호흡기 관리, 투약관리, 안구마우스 사용 방법 등
- 루게릭 이외 뇌수술 등으로 t-tube 환자가 t-tube 제거에 대한 조건 검사 등이 궁금합니다. 루게릭 환자 말기 환자 경험이 없는데 어떻게 진행하고 어떤 식으로 사망하게 되는지 경과가 궁금합니다.



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- ▶ **Interact naturally**
- ▶ **Next generation tracking**
- ▶ **Engineered for gaming**
- ▶ **Easy to install**

Case

- 심한 불면증 및 하지 근육통을 호소하는 36세 남자 환자
- Medical History
 - 상기 환자는 생후 4년에 발견된 루게릭 병으로 14년간 집에서 Care 받으며 생활하고 있었으나 최근 1개월전부터 계속된 심한 불면증과 하지 및 전신 근육통을 호소함. 일시적으로 요양병원에서 입원한 적이 있으나 환자가 불편을 호소하여 주로 집에서 환자의 모친이 직접 돌보아 온 상태임.
- P/E
 - 환자는 누운 상태에서 자가호흡이 힘든 상태로 인공호흡기에 의존하고 있었으며, 사지를 움직일 수 없는 상태로 Swallowing difficulty 있으나 자가로 Soft diet를 유지하는 상태이고, Foley 없이 Urination 은 가능하나, 관장을 하면서 Defecation을 유지하고 있음. 의식은 또렷하며 의사소통 원활하고 언어장애 없으나 대화하는데 힘들어함. 165cm 의 키에 체중은 55kg정도로 비교적 여윈 상태임. 손가락도 움직일 수 없는 전신 마비 상태이며 기타 신체 검진상 특이한 사항 없음.
- Dx
 - 루게릭병; 기타 정신, 신경장애로 인한 불면증
- 경과
 - 환자 및 환자의 보호자인 어머니는 최근에 지속된 불면증으로 심한 고통을 받아오면서 탈진 상태에 이르렀으며, 매우 피로한 안색을 보였으며, 정신적으로도 심한 불안감 및 경미한 공황장애를 보였음. 환자는 밤에 수면을 1시간 이상 지속하지 못하고, 수시로 잠을 깨면서, 하지의 심한 근육통을 호소하여 보호자가 수시로 환자를 돌보아야 하는 상태였음.
 - 환자가 기존에 처방받은 약을 검토하여 보니 수면제인 스틸녹스를 이미 처방 받아 복용한 상태였지만, 증상이 호전되지 않았다고 함.
 - Etizolam과 Airtal을 처방하였는데, 환자의 증상이 매우 호전되어 비교적 정상적인 수면을 회복할 수 있었으며, 밤에 수시로 호소하던 근육통도 현저히 완화되었음. 현재는 Etizolam의 투여용량은 1.0mg을 1일 1-2회 복용하는 상태임.
- Discussions
 - Medical Care: 약물처방, Urination, Defecation, Respirator 관리, Diet 및 영양관리
 - 삶의 질에 관한 방안: 환자 및 보호자의 안정적인 일상생활 및 편안한 정신상태 유지

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