Palliative care in Neurology from the real experience

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Prasat Neurological Institute
History

Present illness

• 58 years old, Thai male

• A year ago, the patient had difficulty when chewing, dysphagia, frequent choking and muffled voice.

• 3 months ago, the symptoms were progressed, He felt tasteless tongue, drooling, weight loss 5 kg.
History

Present illness

• 2 weeks ago, he had weakness at left arm, sometime he felt spastic and pain at left arm, intense emotion and insomnia. no diplopia, no dyspnea.

Past illness

• Congenital hearing loss at right ear

• No other underlying disease

• No drug allergy
Physical Examination

- Vital signs: T 36.5  P69/min RR 20/min BP 103/65 mmHg
- GA: good consciousness, no dyspnea
- HEENT: Tongue fasciculation positive
  atrophy both masseter, temporalis muscle
- Heart: normal S1S2, no murmur
- Lung: normal breath sound
- Neuro: motor grade IV at left arm
  sensory intact all extremities
  IV | V
  V | V
Physical Examination

• CN: pupil 3 mm RTL BE, EOM: full

  No facial palsy

• Gag reflex: decrease both sides

  Fasciculation both hands
Investigation

• CBC : WBC 9,900 Hb 13.8 Hct 41.6 Platelet 295,000
• BUN : 13, Cr : 0.78, Na 134, K 4.27, Cl 93, HCO$_3$ 28.9
• CXR : normal
• EKG : Complete right bundle branch block, rate 63/min
• EMG : Progressive bulbar palsy is most likely.
• CT brain : normal
At OPD Neuromed

- Diagnosis
  - Amyotrophic Lateral Sclerosis (ALS) with bulbar onset

Rx: Bco 1*3 oral pc, folic 1*1 oral pc

: Appointment to Botox clinic

: Consult palliative care
Amyotrophic Lateral Sclerosis

- Cognitive impairment
- Dyspnea
- Pseudobulbar effect
- Pain
- Weakness
- Depression, Anxiety
- Hypersalivation
- Fatigue, Sleep disorder
Role of palliative care services

• Symptomatic treatment
• Advance care planning
• Programmatic support
• Caregiver support
Pain

In ALS major causes of pain are

• Immobility: including joint pain (e.g. shoulder, neck)

• Spasticity and leg cramps

• Psychological, spiritual, or emotional factors that may affect the patient.
Pain: Immobility

**Pharmacological Rx**
- **Mild pain**: Paracetamol, NSAIDs
- **Moderate to Severe**: Opioids

**Non Pharmacological Rx**
- Frequent repositioning
- ROM exercise
- Massage
- Supportive mattresses and wheelchair cushions
- Neck support and collar
Pain : Spasticity

**Pharmacological Rx**
- Baclofen
- Tizanidine
- Gabapentin
- Benzodiazepine
- Botulinum injection

**Non Pharmacological Rx**
- Muscle stretching
- ROM exercise
- Massage
- Splinting
Dyspnea

• *Progressive respiratory muscle weakness*

• *Early respiratory symptom*
  
  : dyspnea on exertion, orthopnea
  
  : nighttime hypoventilation may cause frequent awakening, morning headache, cognitive impairment, hypertension

• *Secondary prevention*: influenza vaccinations or polyvalent pneumococcal vaccines are recommended.
Dyspnea

- Noninvasive Mechanical ventilation: CPAP, BiPAP
  - prolong survival and improve quality of life
Dyspnea

- Supplemental oxygen can lead to carbon dioxide retention and should not be used routinely.
- Prolonged artificial ventilation and tracheostomy should be discussed early.
Dyspnea

- Opioids
  - morphine at a starting dose of 0.5 mg/hour IV or
  - transdermal fentanyl at 12 ug/hour

Dyspnea-related anxiety: benzodiazepine with opioid

- midazolam 2 mg q 1-2 hrs IV, SC, prn
- lorazepam 1–2 mg oral q 6 hours
Pharmacological Interventions for Palliative care in ALS

Wasin Laosuebsakhunthai, Pharm.D
Sialorrhea

- Socially embarrassing symptom.

- Related to pharyngeal muscle weakness which can lead to aspiration pneumonia.

- The prevalence is estimated at 50%
Pharmacologic interventions

- Sialorrhea should initially be treated with anticholinergic medications.

- For patients who remain “medically refractory”, treatment with botulinum toxin type B.

Randomized double-blind study of botulinum toxin type B in ALS patients, 2009
# Medications Commonly Used For Sialorrhea

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dose</th>
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</thead>
<tbody>
<tr>
<td>Amitriptyline</td>
<td>25–75 mg hs</td>
</tr>
<tr>
<td>Nortriptyline</td>
<td>20–100 mg hs</td>
</tr>
<tr>
<td>Atropine</td>
<td>0.4 mg q 4–6 hr</td>
</tr>
<tr>
<td>Glycopyrrolate</td>
<td>1–2 mg TID</td>
</tr>
<tr>
<td>Scopolamine patch</td>
<td>Apply behind ear q 3 days</td>
</tr>
<tr>
<td>Botulinum Toxin Type B</td>
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</tbody>
</table>
Pseudobulbar Affect

- Affects 20%–50% of patients with ALS
- Often sudden, involuntary outbursts of emotion inappropriate to the context of the situation.
- Limiting social interactions and impairing QoL.
Pharmacologic interventions

- Can be successfully treated with SSRIs, TCAs, and SNRIs.

- A combination of dextromethorphan/quinidine should also be considered.
## Medications For Treatment of Pseudobulbar Affect

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>SSRIs: Fluoxetine</td>
<td>10–30 mg/day</td>
</tr>
<tr>
<td>Fluvoxamine</td>
<td>50 mg qd – BID</td>
</tr>
<tr>
<td>Sertraline</td>
<td>50–100 mg qd - BID</td>
</tr>
<tr>
<td>TCAs: Amitriptyline</td>
<td>25–75 mg hs</td>
</tr>
<tr>
<td>Nortripyline</td>
<td>25–75 mg hs</td>
</tr>
<tr>
<td>TeCAs: Mirtazapine</td>
<td>15 mg hs</td>
</tr>
<tr>
<td>SNRIs: Venlafaxine</td>
<td>37.5–75 mg BID-TID</td>
</tr>
<tr>
<td>Dextromethorphan/Quinidine</td>
<td>20mg/10mg 1 tab BID</td>
</tr>
</tbody>
</table>

Sleep disruption

• **Multifactorial** in etiology
  • Respiratory muscle weakness
  • Difficulty re-positioning in bed
  • Anxiety
  • Depression
  • Pain

• Results in frequent arousals and decreased total sleep time.
Pharmacologic interventions

• **Antidepressant medications** are effective at reducing anxiety, depression and promoting sleep.

• **Anxiolytic medications** such as **benzodiazepines**, used specifically to induce sleep, can be helpful when used selectively.
## Medications For Treatment of Sleep disruption

<table>
<thead>
<tr>
<th>Medication</th>
<th>Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mirtazapine</td>
<td>15 mg hs</td>
</tr>
<tr>
<td>Zolpidem</td>
<td>10 mg hs</td>
</tr>
<tr>
<td>Benzodiazepines</td>
<td></td>
</tr>
</tbody>
</table>
Fatigue

- Reported in 44–83% of patient with ALS

- Multifactorial
  - Sleep disruption
  - Nocturnal complaints
  - Nutritional status
  - Weakness
  - Vital capacity
  - Functional status
  - Depression
  - Medications
Pharmacologic interventions

• Modafinil in doses ranging from 100 to 300 mg daily resulted in improvement in
  • Clinical Global Impression score
  • Visual Analogue Scale for energy and stamina

Modafinil treatment of fatigue in patients with ALS: a placebo controlled study, 2009
Physical Therapy in ALS

Nattakitta Suksophonthana
Symptoms

- Muscle weakness
- Wasting
- Fatigue
- Spasticity
- Cramps
- Muscle twitches
- Respiratory failure

Quality of life

mobility
functional

(REHABILITATION IN AMYOTROPHIC LATERAL SCLEROSIS, 2014)
Goal

• Maintain maximum function and quality of life
• Prevent complication
ALSFRS
(Amyotrophic Lateral Sclerosis Functional Rating Scale)

**Objective**
- Assessing the ADL / Functional status
- Record disease progression

**10 Items**
1. Speech
2. Salivation
3. Swallowing
4. Handwriting
5. Cutting food and handling utensils (with or without gastrostomy)
6. Dressing and hygiene
7. Turning in bed and adjusting bed clothes
8. Walking
9. Climbing stairs
10. Breathing

(www.physio-pedia.com/The_Amyotrophic_Lateral_Sclerosis_Functional_Rating_Scale), 2016
Physical Therapy Techniques

- Therapeutic exercise
  - Passive exercise
  - Active-assisted exercise
  - Active exercise
  - Resisted-active exercise
  - Stretching exercise
Physical Therapy Techniques

- Bed mobility and gross motor functional training
- Sitting and Standing balance training
- Ambulation and Walking training (with or without gait aids)
- Transfer training
Physical Therapy Techniques

- **Electrical modalities**: Transcutaneous electrical nerve stimulation (TENS), Interferential current (IF)
- **Thermal modalities**: Cold pack, Hot pack, Ultrasound
- **Manual therapy**: Joint mobilization, Massage
Physical Therapy Techniques

- Chest physiotherapy
  - Breathing exercise
    - Diaphragmatic breathing exercise
    - Segmental breathing exercise
    - Pursed lip breathing exercise
Physical Therapy Techniques

- Chest physiotherapy
  - Positioning
  - Postural drainage
  - Coughing training
  - Percussion/ Vibration

Upper Lobes
Apical Segments
Position #1

Middle Lobe
Position #5

Lower Lobes
Lateral Basal Segments
Positions #3 and #9
Nursing Management in ALS

Sirinya Naratchariyangkoon
Medical Management

No specific therapy exists for ALS.

The main focus of medical and nursing management to maintain or improve function, well-being and quality of life.

Symptomatic treatment and rehabilitative measures are employed to support the patient and improve the quality of life.
Nursing Care Plan of ALS

• PROBLEMS
  - Weakness to flaccid paralysis

• GOALS
  - Keep patient as active as possible
  - Prevent skin breakdown

• NURSING INTERVENTION
  - Active and passive ROM to affected limbs 3-4x per day.
  - Turn and position every 2 hours.
  - Keep skin dry.
  - Wash skin and dry well after each bowel movement or urination (especially females).
  - Lotion dry areas.
  - Air flotation bed.
Nursing Care Plan of ALS

- **PROBLEMS**
  Emotional response to diagnosis and prognosis (anxiety, fear, denial, anger)

- **GOALS**
  To assist patient through grief and grieving process

- **NURSING INTERVENTION**
  - Encourage verbalization of feelings.
  - Accept behavior during stages of grieving process and allow grieving.
  - Teach family about grieving process and encourage them to accept patient’s behavior and their own response.
Nursing Care Plan of ALS

• PROBLEMS
  Emotional response to diagnosis and prognosis (anxiety, fear, denial, anger)

• GOALS
  To assist family to deal with patient and grief process

• NURSING INTERVENTION
  - Use positive approach when discussing progress (ask what patient and family know about disease).
  - Encourage to maintain independence for as long as possible and to do things for self when possible.
  - Teach family to do the same.
  - Plan diversionary activities, OT, etc., while hospitalized
  - Have family plan same for at home
Nursing Care Plan of ALS

- **PROBLEMS**
  Altered body image and self-esteem

- **GOALS**
  To assist patient in accepting new body image and maintaining self-esteem

- **NURSING INTERVENTION**
  - Discuss changes in body image and what they mean to patient.
  - Encourage patient to talk about such with family.
  - Encourage patient to focus on positive aspects of self and to share those aspects of self with others.
Nursing Care Plan of ALS

• PROBLEMS
  Respiratory insufficiency

• GOALS
  Maintain adequate $O_2$ delivery to patient; prevent complication of pneumonia assisted cough

• NURSING INTERVENTION
  - Suction and maintain patient airway.
  - Administer $O_2$ as necessary.
  - Elevate HOB at least 30 degrees at all times.
  - Auscultate chest frequently to assess total airway states.
  - Help patient to cough and deep breathe at least every two hours.
  - May use incentive spirometer if necessary or chest physical therapy.
  - Maintain on respirator if necessary
Nursing Care Plan of ALS

• PROBLEMS
  Difficulty swallowing and chewing

• GOALS
  Prevent aspiration

• NURSING INTERVENTION
  - Assess gag reflex before giving fluid or food by mouth.
  - Elevate HOB at mealtime and for ½ hour afterward.
  - Make food easier for patient to eat
Nursing Care Plan of ALS

• PROBLEMS
  Difficulty swallowing and chewing

• GOALS
  Ensure adequate nutrition and fluid balance

• NURSING INTERVENTION
  - High protein, high CHO diet
  - Fluid intake 3000 cc/day.
  - Consult dietician to teach basics of good nutrition and in-between meal snacks
Nursing Care Plan of ALS

• PROBLEMS
  Difficulty speaking

• GOALS
  Establish effective means of communication

• NURSING INTERVENTION
  -Initiate use of a magic slate if patient able to use hands.
  -Initiate use of word boards or letter board if unable to use arms.
  -May use electro-larynx like that used for laryngectomy patients.
    -Consult speech therapy and OT for assistance and further assessment of speech needs and means for meeting these needs.
Nursing Care Plan of ALS

• PROBLEMS
  Urinary frequency or incontinence of urine and feces of impaction

• GOALS
  To maintain normal bowel and bladder function

• NURSING INTERVENTION
  - Place urinal/bed pan where patient can reach if able to do so.
  - Use effective means for patient to communicate when needs to urinate or have bowel movement.
  - Catheterize or when necessary
  - External cath may be useful
  - Bowel routine as necessary for impaction; high fiber diet if patient can tolerate, Fleet enema, Colace, Metamucil, fluids, suppositories.
Signs/symptoms of illness, to notify M.D

- Respiratory difficulty
- Signs of infection – cold, flu, elevated temperature, chills
- Difficulty swallowing
- Difficulty speaking
- Sudden increase in original signs and symptoms of disease
Nutrition management in palliative care

Skaw Meewan
The importance of good nutrition

1. Enjoy overall wellbeing and quality of life
2. Maintain a strong immune system and reduce the risk of infection
The importance of good nutrition

3. Cope with the demands of illness and treatment
The importance of good nutrition

4. Increase wound healing and tissue
Case ALS

• 58 years old, Thai male

• A year ago, the patient had difficulty when chewing, dysphagia, frequent choking and muffled voice.

• 3 months ago, the symptoms were progressed, He felt tasteless tongue, drooling, weight loss 5 kg
Nutrition care process in palliative care

1. Nutrition screening and assessment
1. Nutrition screening and assessment

<table>
<thead>
<tr>
<th>1. Height/Body's length/Arm span</th>
<th>58</th>
</tr>
</thead>
<tbody>
<tr>
<td>Body's length</td>
<td>Health</td>
</tr>
</tbody>
</table>

2. Weight and Body mass index (BMI) = Weight (kg)/Height (m²)

2.1. Weight: 58 kg, BMI = 18.6

3. Body build

4. Weight change in the past 4 wk

5. Dietary intake in the past 2 wk

6. Persistent gastrointestinal symptoms in the past 2 wk

7. Functional capacity

8. Pt’s disease, please inform dietitian/nutritionist

*Interpretation

- Scores of 0-5 (NAF = A: Normal-Mild malnutrition)
  - No risk of malnutrition, nurse should rescreen the patient again within 7 days.

- Scores of 6-10 (NAF = B: Moderate malnutrition) Please inform attending doctor and dietitian/nutritionist immediately.
  - Moderate risk of malnutrition. Patient should be assessed by dietitian/nutritionist and received nutrition therapy by attending doctor within 3 days.

- Scores of 11 and more (NAF = C: Severe malnutrition) Please inform attending doctor and dietitian/nutritionist immediately.
  - Severe risk of malnutrition. Patient should be assessed by dietitian/nutritionist and received nutrition therapy by attending doctor within 24 hours.

Screened by: Komindr S., M.D., Division of Nutrition and Biochemical Medicine, Department of Medicine, Ramathibodi Hospital, 2013
Nutrition care process in palliative care

1. Nutrition screening and assessment

2. Nutrition diagnosis

- Inadequate protein and energy intake
- Moderate malnutrition

BW ↓ ~1.7 kg/mo. (~2.6%)
Intake ~600-1000 Kcal. Prot. ~ 18 g/d
Energy req. ~ 1,972 Kcal. Prot. Req. ~ 70 g/d
Nutrition care process in palliative care

1. Nutrition screening and assessment

2. Nutrition diagnosis

3. Nutrition intervention
Malnutrition

Oral nutritional supplementation

Nutritional status assessment

Eutrophy

Overweight/obesity

Guidance

Nutritional status reassessment

Weight loss

Yes

No

>10%

<10%

Guidance

PEG

Nutritional supplementation
Dealing with:

- Chewing difficulty
- Swallowing problem
- Lost of test
Nutrition care process in palliative care

1. Nutrition screening and assessment
2. Nutrition diagnosis
3. Nutrition intervention
4. Nutrition monitoring and evaluation
Malnutrition

Oral nutritional supplementation

Nutritional status reassessment

Day 7
BW: not feasible
[Last wk. BW ↓ ~1.7 kg/mo. (~2.6%)]

Intake ~1200-1400 Kcal.
Prot. 32 g/d but..

Energy req. ~ 1,972 Kcal.
Prot. req. ~ 70 g/d
Malnutrition

Oral nutritional supplementation

Nutritional status reassessment

Weight loss

Yes

>10%

PEG

<10%

Guidance

No

Guidance

Nutritional supplementation
**Malnutrition**

- Oral nutritional supplementation

**Nutritional status reassessment**

**Weight loss**

- Yes
  - >10%
    - Intake ~2100 Kcal. Prot. ~70 g/d
    - Energy req. ~ 1,972 Kcal.
    - Prot. Req. ~ 70 g/d
  - <10%
    - Guidance

- No
  - Guidance

**PEG**

**B-2- P.E.G. (Percutaneous Endoscopic Gastrostomy)**
แบบประเมินระดับคุณภาพที่ใช้ในการจัดอุปกรณ์ประจำบ้านผู้ป่วย (Palliative performance scale for adult Suandok) (PPS Adult Suandok)

<table>
<thead>
<tr>
<th>ระดับ PPS</th>
<th>ระดับ ความรู้สึกดี</th>
<th>การท่าทางรักษา</th>
<th>การปฏิบัติต่อการรักษาและการดูแลรักษา</th>
<th>การทำกิจกรรม</th>
<th>การรับประทานอาหาร</th>
<th>ระดับ ความรู้สึกดี</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>เกิน 70</strong></td>
<td>&gt; 70 = Early stage</td>
<td>&gt; 70 = Early stage</td>
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<tr>
<td><strong>40-70</strong></td>
<td>40-70 = Middle stage</td>
<td>40-70 = Middle stage</td>
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<td>40-70 = Middle stage</td>
<td>40-70 = Middle stage</td>
</tr>
<tr>
<td><strong>0-30</strong></td>
<td>0-30 = Late stage</td>
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<td>0-30 = Late stage</td>
</tr>
</tbody>
</table>

100 เลือกให้สุขภาพดี ทักษะการดูแลรักษาได้ตามปกติ และไม่มีอาการของโรค ทำได้ ปกติ ปกติ รู้สึกดี
90 เลือกให้สุขภาพดี ทักษะการดูแลรักษาได้ตามปกติ และมีอาการของโรคบางอย่าง ทำได้ ปกติ ปกติ รู้สึกดี
80 เลือกให้สุขภาพดี ต้องการความช่วยเหลือเดินหรือยืน ทักษะการดูแลรักษาได้ตามปกติ และมีอาการของโรคบางอย่าง ทำได้ ปกติ หรือ ลดลง รู้สึกดี
70 ความสามารถในการเลือกให้สุขภาพดี ไม่สามารถทำกิจกรรมได้ตามปกติ และมีอาการของโรคบางอย่าง ทำได้ ปกติ หรือ ลดลง รู้สึกดี
60 ความสามารถในการเลือกให้สุขภาพดี ไม่สามารถทำกิจกรรมได้ ช่วยเหลือดี ทักษะการดูแลรักษาได้ตามปกติ และมีอาการของโรคบางอย่าง ทำได้ ปกติ หรือ ลดลง รู้สึกดี
50 ความสามารถในการเลือกให้สุขภาพดี ไม่สามารถทำกิจกรรมได้เลย และมีอาการของโรคบางอย่าง ต้องการความช่วยเหลือ ทักษะการดูแลรักษาได้ตามปกติ ปกติ หรือ ลดลง รู้สึกดี
40 ความสามารถในการเลือกให้สุขภาพดี ต้องการความช่วยเหลือ ทักษะการดูแลรักษาได้ตามปกติ และมีอาการของโรคบางอย่าง ปกติ หรือ ลดลง รู้สึกดี
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0 เสียชีวิต
Advance care planning

1. Discuss diagnosis, prognosis, likely course of illness, including disease-modifying therapy

2. Elicit patient-centered goals, hopes, expectations

3. Advise patients regarding options for visiting nurse, home care, hospice service.

4. Offer care from Multidisciplinary centers
Advance care planning

5. Encourage support or counseling of family.


7. After death 1-2 week, family was called for complicated bereavement screening.
THANK YOU