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Lou Gehrig's Disease

AMYOTROPHIC LATERAL SCLEROSIS
Physical Therapy Interventions in Attempt to Limit Debilitating Symptoms©

Jennifaye V. Brown, PhD, PT, NCS

Liability/Disclosure Statement

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Objectives:

3

1. Explain the pathophysiology of ALS
2. Differentiate between the major types and subtypes of ALS.
3. Describe common impairments that are a result of ALS.

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Objectives:

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4. Identify and or design specific physical therapy treatment interventions used to alleviate or dissipate symptoms and or impairments depending on disease progression.
5. Recommend equipment that will maximize the potential to engage in functional activities.

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Outline

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1. History
2. Etiology & Pathophysiology
3. Types of ALS
4. Prevalence & Incidence
5. Disease Progression
6. Common Impairments
7. Medical Management & Physical Therapy Interventions
8. Patient & Caregiver Education
9. Knowledge Summary

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History

https://medicallabandcoats.files.wordpress.com/2012/04/jean-martin-charcot.jpg

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Discovered by[1]…

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- Jean-Marie Charcot: a French neurologist, but trained as a pathologist
- aka "Father of Neurology"
- He and a colleague found lesions..
  1. lateral column of spinal cord: progressive paralysis & contractures, but no atrophy
  2. anterior horn of spinal cord: paralysis & no contracture, but atrophy

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ALS termed

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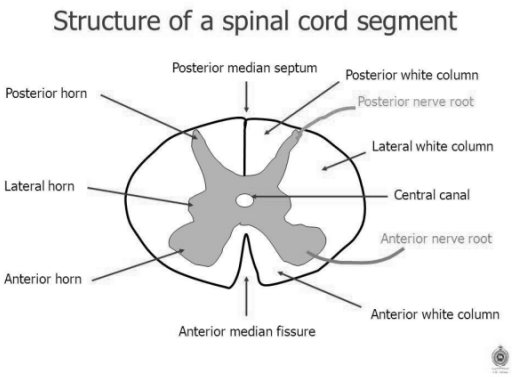
- In 1874 when Dr. Charcot's lectures compiled as "Oeuvres Completes"[1]
- Still known as Charcot's Dz in some parts of the world



http://www.bing.com/images/search?q=picture+lateral+column+of+spinal+cord&view=detailv2&id=0B48EC74B4078750A0179EB0AA00B0033873E6C5&selectedindex=10&ccid=TDEYiRB1&simid=608055859716820311&thid=OIP.M4c3118891075ca218d00bf2f84ba6d53H0&mode=overlay&first=1

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Lou Gehrig's Disease

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- 1st baseman New York Yankees
- 1923-1939
- Forced to retire in 1939 after dx of ALS
- Died 2 years later

http://www.lougehrig.com/

http://www.bing.com/images/search?q=picture+of+lou+gehrig&view=detailv2&id=3F170778C4DD274AE8ED312059FBD22388DB3F4A&selectedindex=3&ccid=O2uIvH4B&simid=608052561177477849&thid=OIP.M3b6b88bc7e016f08d89427ee0017c21eH2&mode=overlay&first=1

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Defining ALS[2]

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- a= without
- myo= pertaining to muscle
- trophic= pertaining to growth & nourishment of structures by efferent innervation
- lateral= pertaining to the side
- sclerosis= hardening of nervous tissue due to degeneration

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Overall…

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- Primarily considered a disease of CNS[3,4]
- Progressive weakness/paresis leading to death in a few years from respiratory compromise[3-5]

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Etiology

Due to current imaging and genetic studies, the etiology of ALS is determined by pathophysiology & associated clinical classifications[3]

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ALS…

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- A multisystem disease with various pathophysiological determinants of origin[3]

Telsa-ard.blogspot.com

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Pathophysiology

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Pathophysiological Mechanisms[3]

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1. Oxidative stress
2. Mitochondrial damage
3. Protein accumulation
4. Cytoskeletal interruption
5. Glutamate and neuronal cytotoxicity
6. Altered regulation of gene expression

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1. Oxidative Stress

16

- impaired performance of cells resulting from too many oxygen molecules in them

EncartaDictionaries

www.commomswikimedia.com

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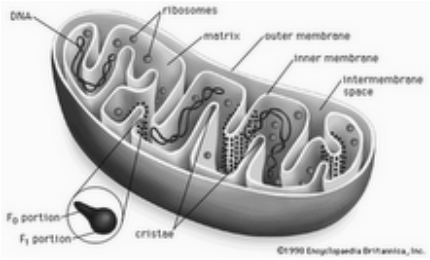
2. Mitochondrial Damage

17

- cell "energy factories"
- small rod-shaped structures, that produce about 90% of energy needed for cell survival



http://www.thebodypro.com/content/art1976.html

Uhscellproject2010.wikispaces.com

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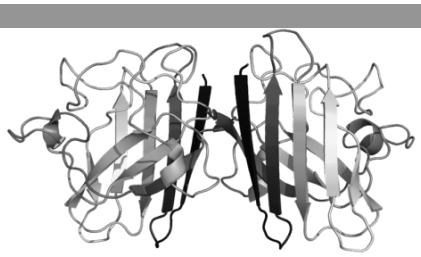
3. Protein Accumulation

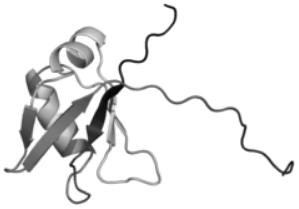
18

- Shape change in the superoxide dismutase 1 (SOD1) protein source of motor neuron death



www.neuralcloud.it

- Mutation of protein TDP43 also evident in ALS cases



http://www.medpagetoday.com/Neurology/GeneralNeurology/22790

en.wikipedia.org

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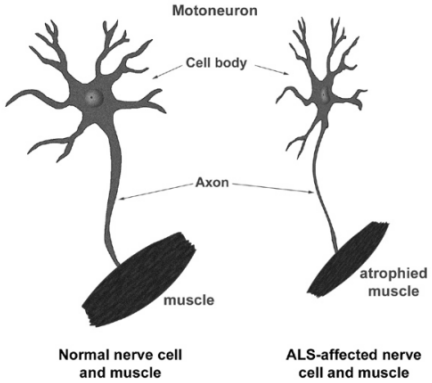
4. Cytoskeletal Interruption

19

- Genetic mutation of profilin (PFN1) gene needed for the growth and development of the cytoskeleton in nerve cell axons[6]



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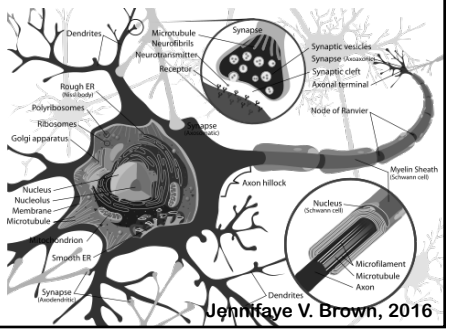
5. Glutamate & Neuronal Cytotoxicity

20

- High levels of glutamate causes nerve cells that control voluntary muscles to die – glutamate toxicity due to calcium staying in cell

http://www.alsa.org/research/about-als-research/glutamate.html

- Nerve cell classification:
  1. Upper
  2. Lower



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Motor Neuron Classification[7]

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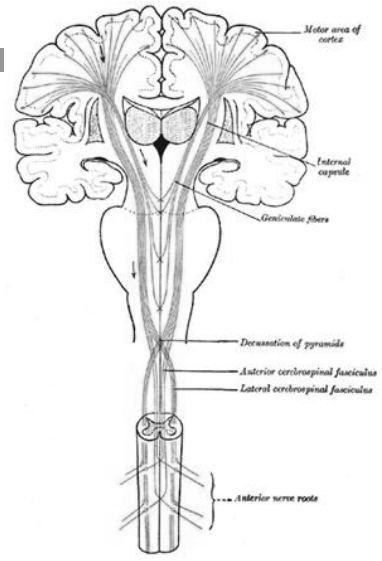
1. Upper motor neuron (UMN): all nerve cells in the brain and spinal cord
2. Lower motor neuron (LMN): all nerve cells that exit the spinal cord (from the anterior horn) and goes to the muscle

UMN + LMN = CNS



http://deltabiology.com/category/human-physiology/central-nervous-system/

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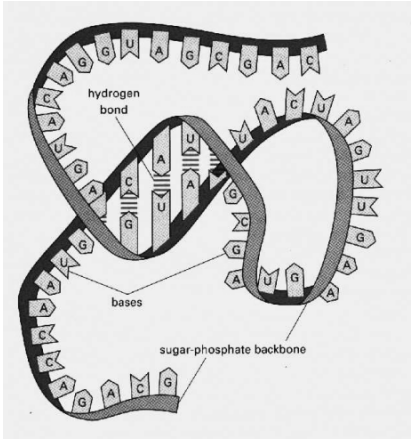
6. Altered Gene Expression Regulation

22

- Specifically, alteration in the metabolism of ribonucleic acid (RNA) which is responsible for converting DNA stored information into proteins[8]



beauchemin.wikispaces.com

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Types of ALS

ALS is classified into 3 major types…

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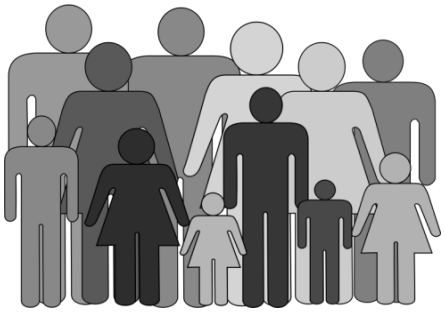
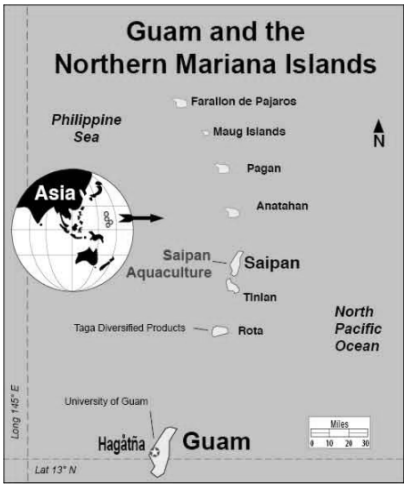
Major Types of ALS[3,9]

24

1. Sporadic
2. Familial
3. Western Pacific



openclipart.org



http://quoteimg.com/guam-mariana-islands/

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Major Types of ALS[3,9]

25

1. Sporadic: 90-95%
   - unable to regulate glutamate
   - 11% gene mutation at fault

2. Familial: 5-10%
   - Inherited genetically dominant pattern
     a. 20% gene encoding deficiency of enzyme copper-zinc superoxide dismutase (CU-Zn SODI)
     b. 2-5% TARDBP (TDP-43) gene mutation

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Major Types of ALS[3,9]

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3. Western Pacific
   - Chamorro people of Guam – a US territory: – incidence rate 50% higher
   - Marianas Island
   - Kii Peninsula of Honshu Island
   - Southwest New Guinea

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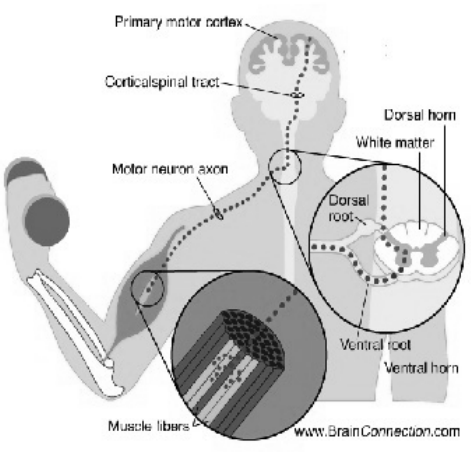
Subtypes of ALS:

Clinical Presentation

1. Degeneration of the UMN[9]:

28

The primary motor tract impacted causing progressive muscle weakness….



Corticospinal Tract[7]

29

- Motor neurons that start at cortex – frontal lobe and end in the gray matter of the spinal cord at the level of the arms and the legs
- Responsible for skilled, refined movement of the extremities
- Arm and leg muscles; shoulder and pelvis muscles are weak

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Question???

What LMNs can be affected?

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Lower Motor Neuron(s)[7] that…

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1. leave cranial nuclei in the brainstem & innervate muscles of the face, eyes, neck, throat

2. exit anterior horn of the spinal cord and go to muscles of the arm & leg

3. innervate the diaphragm

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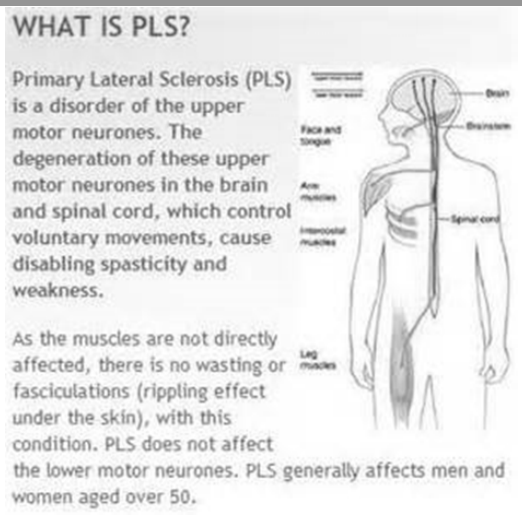
2. Primary Lateral Sclerosis[3,9]

32

- Disorder of <u>UMN</u> in brain & spinal cord
- SLOWLY progressive weakness & spasticity
- No atrophy or fasciculations



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3. Progressive Bulbar Palsy[3,9]

33

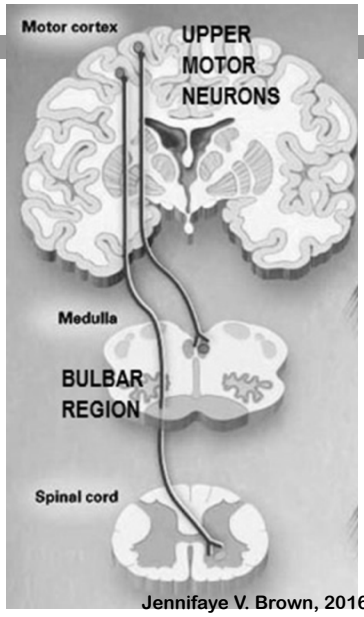
Degeneration of motor neurons of Cranial Nerves 9-12

9: gag reflex/swallowing

10: speech and swallowing

11: ability to create a bolus

12: neck muscle



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Cranial Nerves & Association

34

9: Glossopharyngeal

10: Vagus

11: Hypoglossal

12: Spinal Accessory

a. innervates sternocleidomastoid and upper trapezius

b. motor component gag reflex; vocal cord weakness, difficulty swallowing, sensation for nausea, heart & respiratory rate

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Cranial Nerves & Association

35

9: Glossopharyngeal
10: Vagus
11: Hypoglossal
12: Spinal Accessory

c. motor innervation for the tongue

d. taste posterior 3rd of tongue; sensation soft palate

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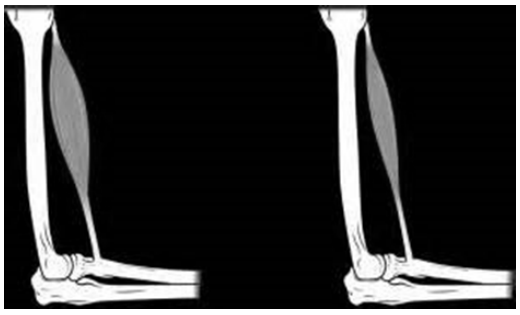
4. Progressive Muscular Atrophy[9]

36

- Loss of Nissl bodies in the cell body or their death (chromatolysis) in motor neurons of spinal cord & brainstem



- Muscle wasting

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5. ALS-FTD[10-12]: FrontoTemporal Dementia

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- Cognitive decline
- Personality changes
- Irritability
- Decrease insight
- Deficits in executive functioning

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Prevalence & Incidence

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Definitions[1]

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Prevalence	Incidence
□ The number of diagnoses of a disease present in a population at a given time.	□ The frequency a disease occurs over a period of time in relation to the population.

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Definitions[3]

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Prevalence	Incidence
25,000 individuals with ALS in the U.S. @ the time publication received/published: 2-2015/6-2015	□ 1-2/100,000 each year □ Lifetime risk of ALS 1/600

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Who gets ALS?

http://wwwn.cdc.gov/als/WhatisALS.aspx
http://www.alsa.org/about-als/facts-you-should-know.html

41

- 5,600 new cases/yr in U.S.
- ALS dx higher in men than women, but evens out with age
- 40-60/70 yrs old
- Average life expectancy 2-5 yrs; death due to pneumonia/aspiration; eventually needs ventilator support – WHY???
- 20% live 5 yrs & 10% at least 10 yrs, per research these are motor neuron dzs affiliated with ALS

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Motor Neuron Dzs classified as[3]

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1. ALS/DNM: sporadic cases, family or genetic origin
2. ALS-plus syndromes: multisystem neurodegenerative dz affecting motor neurons
3. ALS-related syndromes: symptomatic or 2ndary forms of motor neuron dz, with a known associated condition that may be causing dz
4. ALS variants: uncommon unless pt lives in a particular geographic location

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Definite Dx of ALS…..

Criteria Used

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CRITERIA[3]

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El Escorial	Awaji-Shima
□ Upper and lower motor neuron signs in 3 regions □ brain, brainstem, spinal cord	□ Clinical or electrophysiological evidence □ UMN & LMN in bulbar region and at least 2 spinal regions OR □ Presence of UMN & LMN in 3 spinal regions

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Disease Progression

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ALS Clinical Profile[3]

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- Variable
- Foot drop w or w/o falls
- Impaired dexterity
- Motor cells in anterior horn of L_4 & C_8-T_1
- Muscle weakness becomes apparent, 80% of motor neurons lost in corresponding myotomes – leading to rapid decline & death

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Common Impairments

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Musculoskeletal[3,9,13-15]

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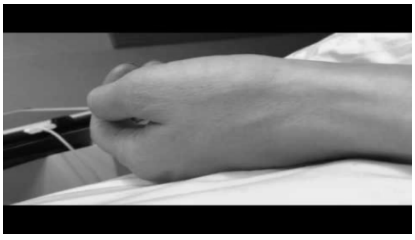
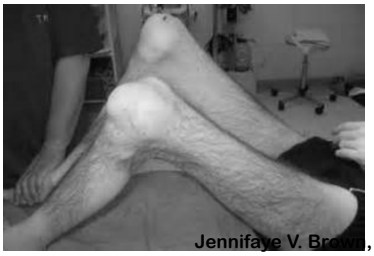
1. Muscle weakness
2. Atrophy
3. Fasciculations
4. Hypotonicity
5. Muscle cramps
6. Contractures





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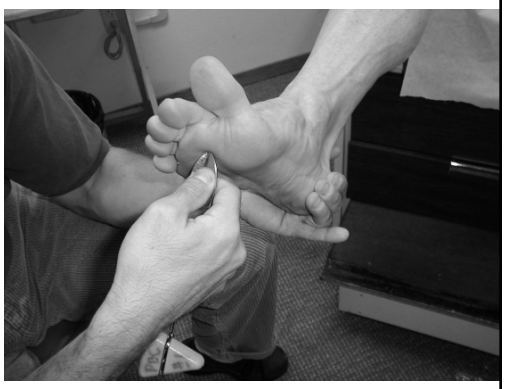
Neuromuscular[3,9,13-15]

49

1. Spasticity
2. Pathological Reflexes (Babinski)
3. Hyperreflexia
4. Hyporeflexia



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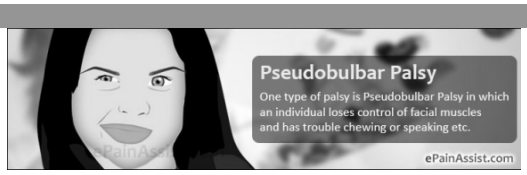
Impairments Related to Brainstem[3,9,13-15]

50

1. Dysphagia
2. Sialorrhea
3. Dysarthria
4. Pseudobulbar effect
5. Poor head control



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Impairments: Psychosocial

51

5. Behavioral impairments: depression & anxiety[9,16]

6. No impairment of intelligence, but changes in other cognitive functions due to frontotemporal dementia:[3,9]
   1. attention, cognitive inflexibility, loss of insight
   2. memory

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Respiratory Compromise[3]

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Primarily: Respiratory Failure

- Pulmonary compromise due to aspiration
- Dyspnea
- Fatigue
- Morning headache
- Sleepiness

mixedthoughts.net

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Pain

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1. From muscle inactivity/cramps[4,9,15]
2. Musculo-skeletal pain[4,9,15]
2. Bed sores[4,9,15]
3. Bladder infections[4,9,15]
4. Small fiber neuropathy[17]

www.nurse24.it

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Pain: Hansich et al[4]

54

- 78% of 46 pts with ALS (n=36) reported pain: "miserable", "tender", "dull/pressing", & "exhausting"
  - Mild pain: 21 (58%)
  - Moderate pain: 14 (39%)
  - Severe pain: 1 (3%)
- Interferes with enjoyment of life, mood f/b mild interference with general activities, walking ability & sleep
  - Relationship with other people least impacted

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Pain: Hansich et al[4]

55

Pain Severity Score (range: 0-10): 3.0 (0.5-6.8) in past 24 hrs

- 47% of pts with pain (n=17) receiving some type of tx:
  - NSAIDs, opiates, antiepileptic drugs & others
- 2 pts: acupuncture, massage, ultrasound
- 40% of pts with moderate-severe pain took opiates

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Pain: Hansich et al[4]

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Type or Origin	Location
Cramps	Back (50%)
Spasticity	Extremities (47%)
	Joints (42%)

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Interesting to note[9]….

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- Able to see (acuity), smell, taste & recognize touch
- Rare impairments: inability to move eyes, bowel & bladder dysfunction (unable to actively push)

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Medical Management

Care emphasis is supportive and patient centered for symptom management

Recommended Practice:

Multidisciplinary Comprehensive Services[15,17] in Specialized Centers[17]

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Medical Treatment

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1. Riluzole:[3,9] ↓s amount of glutamate release
   - Prescribed to slow muscle degeneration
   - Requires monitoring of liver function, blood count, blood chemistries
2. Medications for:[4,9,]
   a. Fatigue
   b. Pain
      1. muscle cramps
      2. spasticity
   c. Psychosocial issues: depression

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Pain Tx

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Muscle Spasticity	Muscle Cramps
[4]Baclofen & Tolperisone	[4]Magnesium, Chinine Sulfate, Pregabaline
[15]Baclofen, Tizanidine	[15]Baclofen, Gabapentin

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Pain Tx[4, 9]

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- Muscle Inactivity
  - Range of Motion exercises
- Bed Sores
  - Turn frequently
  - Use Cushions
  - Towels and/or pillows behind back; support neck
  - Repositioning of power w/c or body in manual w/c

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Other Interventions Respiratory[3]

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Noninvasive ventilation:

- Reduction in 50% - forced vital capacity (FVC)
- SpO_2 < 88% for more than 5 minutes during night
- Increased partial pressure of O_2 in arterial blood ($PaCO_2$) > than 45 mmHg
- Increase in maximal inspiratory pressure of inspiratory muscles (MIPIM) above -60 cm H_2O

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Other Interventions[3,9]

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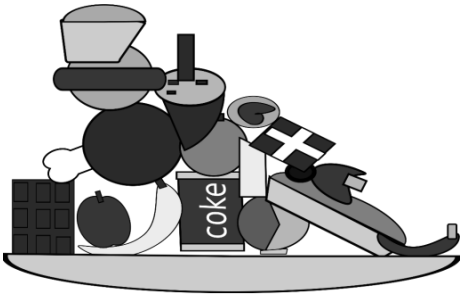

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Clinical Dietician

SLP:
Swallowing Assessment
Modified Barium Swallow Test

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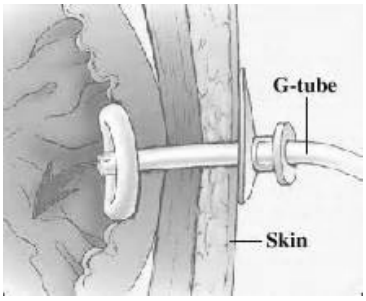
PEG Tube Management[9]

A tube is passed into a patient's stomach through the abdominal wall, most commonly to provide a means of feeding when oral intake is not adequate



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PEG Tube Management

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1. Disconnect from feeding mechanism
2. Flush PEG tube with water as appropriate and instructed
3. Clamp the tube and coil in a circle
4. Cover with gauze and tape
5. If abdominal binder available, apply to patient

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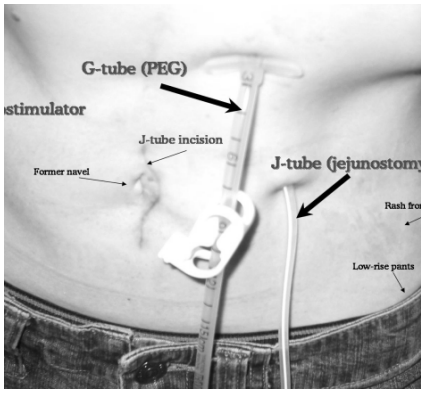
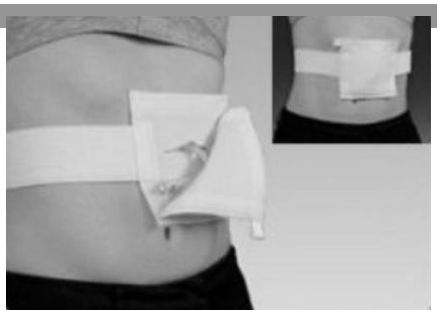
PEG Tube Management

66





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Other Interventions

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Supportive Care: Health Care Team

a. Maintenance: active exercise

b. Compensatory: neck brace; chair lift; feeding utensils

c. Preventative: mm cramps/spasticity – ROM/stretching

Miami- J Collar

Aspen Collar

Philadelphia Collar

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Physical Therapy

Interventions Related to:

Safe Handling Techniques, Exercise/Physical Activity, Positioning & Mobility, Communication

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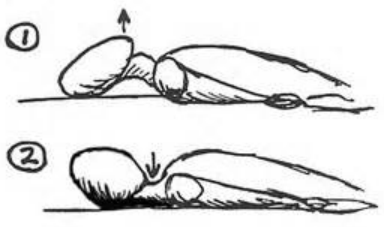
Safe Handling Techniques

Suggested Interventions

70

- 1. Use a gait belt at trunk; beware of peg tube
- 2. Support head in upright sitting or in supine (pt should not lay flat) – facilitate or position with chin tuck; avoid neck hyperextension; #1

especialneeds.com



Safe Handling Techniques

71

3. Do not pull on arms as it may cause shoulder subluxation because muscles are weak – resulting in pain

a. Use a draw sheet to move in bed

b. Avoid any pulling by placing your arm under patient's armpit

c. Support shoulder blade when rolling patient and other hand on trunk

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Avoiding Shoulder Subluxation

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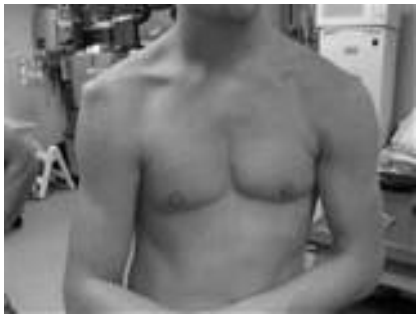

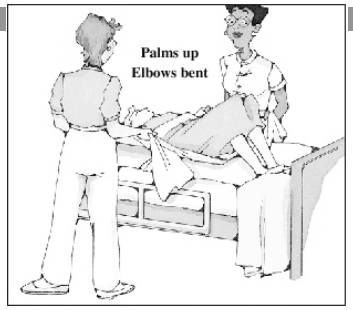

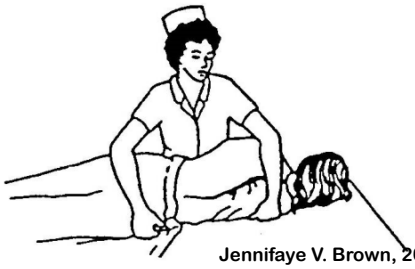

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Shoulder Subluxation

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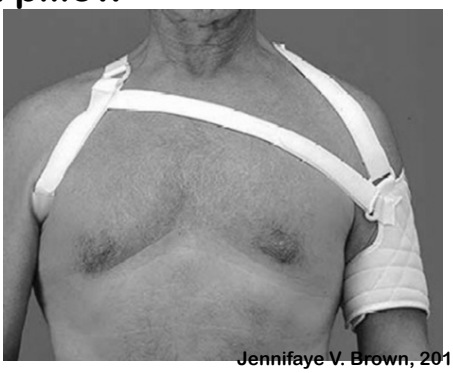
- Use positioning devices and slings as appropriate
- In bed make sure elbow is at level of shoulder or above supported on pillow



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ASSESSMENTS

ALS Functional Rating Scale

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1. Speech
2. Salivation
3. Swallowing
4. Handwriting
5. Cutting Food
6. Dressing & Hygiene
7. Turning in Bed
8. Walking
9. Climbing Stairs
10. Dyspnea
11. Orthopnea
12. Respiratory Insufficiency
13. How many years Since Onset of Symptoms

http://www.outcomes-umassmed.org/als/alsscale.aspx

ALS FRS-R

76

- 12 questions
- 0 – 5 rating scale (cannot do – normal ability)
- Items summed: 0 – 48 (worst 0 best)

http://www.outcomes-umassmed.org/ALS/sf12.aspx#scale

ALS Assessment Questionnaire

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- ALSAQ – 5 Score is short version of ALSAQ – 40 Scale
- Measures PATIENT'S subjective health status of impairments & disabilities: ALS/MND

1. Physical Mobility
2. Eating & Drinking
3. ADLs
4. Communication
5. Emotional Status

- 5 Questions
- Rating scale
  0= Never
  4= Always or cannot do at all
- 0=Best – 100 Worst

http://www.outcomes-umassmed.org/ALS/sf12.aspx#scale

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Exercise/Physical Activity

https://upload.wikimedia.org/wikipedia/commons/thumb/2/26/Pictograms-nps-land-exercise-fitness-2.svg/120px-Pictograms-nps-land-exercise-fitness-2.svg.png

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Paganoni et al[15]

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- Authors have a review of the literature in their article

  - *For those able to perform exercise, moderate intensity/resistance appropriate & safe*

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Paganoni et al[15]

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- Drory et al, 2001[19]: major mm groups engaged in moderate intensity daily program to improve mm endurance associated with
  1. Less functional decline – ALS Functional Rating Scale
  2. Decrease spasticity – Ashworth Spasticity Scale
- Bello-Haas et al, 2007[20]: moderate resistive exercise resulted in improved function, QOL and no adverse effects
  1. ALS Functional Rating Scale

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Paganoni et al[15]

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- Trial of Resistance & Endurance Exercise in ALS
  - Current clinical trial at John Hopkins/ALS Assn
  - Questions
    1. exercise slow dz progression
    2. harm in exercising
    3. best: resistance or endurance

Refer to website:

https://clinicaltrials.gov/ct2/show/NCT01521728

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Majmudar et al[13]

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EARLY	MIDDLE	ADVANCED
mm weakness, fatigue, ↓ endurance, falls	same as previous; progressive bulbar symptoms; pain	same as middle stage breathing, swallow, speech failure
strength, balance, & gait assessment; functional mobility skills	Health care team ↑; transfer technique modifications; w/c consult	trach, feeding tube, aug com device[21] – SLP transition rehab to consultation w/ hospice care
AFO, assistive device & adaptive equipment, safety, exercise, energy conservation; psychosocial	more aggressive AD & adaptive equipment & mobility needs; pain management; PROM-AAROM ex; positioning; meds for	Palliative health care team approach; manage pain, respiratory & swallowing dysfxn (trach/suctioning);

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Energy Conservation

Overuse fatigue: weak/denervated muscle works "harder" because functioning close to its maximal limit

- remaining undamaged motorneurons will respond to training
- KEY: exercise program: low to moderate intensities

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Energy Conservation Ideas

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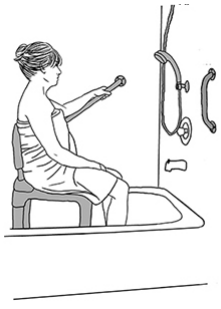

1. Rearrange environment
2. Eliminate unnecessary effort
   - Support trunk
   - Put things closer to pt
3. Plan ahead
   - If a task requires lots of effort (am bathing), forego another activity (breakfast outing)
4. Prioritize
   - Choose to do what is most important at the time

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Physical Activity Summary

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- Special equipment can enhance patients' independence and safety throughout the course of ALS.
- Gentle, low-impact aerobic exercise such as walking, swimming, and stationary bicycling can strengthen unaffected muscles, improve cardiovascular health, and help patients fight fatigue and depression – ACUTE PHASE.

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Physical Activity Summary

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- ROM and stretching exercises can help prevent spasticity and contracture of muscles – MIDDLE PHASE.
- Devices such as splints, ramps, braces, walkers, and wheelchairs that help patients conserve energy and remain mobile – MIDDLE & ADVANCED PHASE.

SUMMARY:

- Any activity should avoid over fatiguing!

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POSITIONING & MOBILITY

ALS Progression

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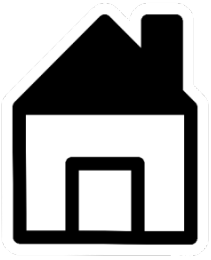
- Eventually patient will get weaker & require dependency for mobility



openclipart.org

- Power wheelchair
- Hoyer lift
- Hospital bed
- Combination Shower/toilet chair
- Viable transportation that accommodates power wheelchair and ventilation devices

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Mobility, Pressure Relief & Communication[21]

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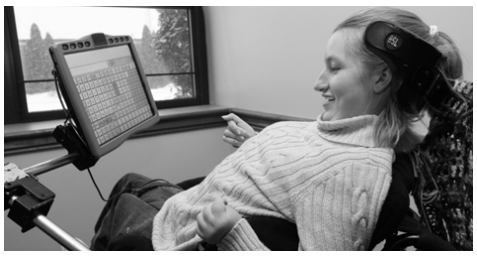

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Specialized Equipment

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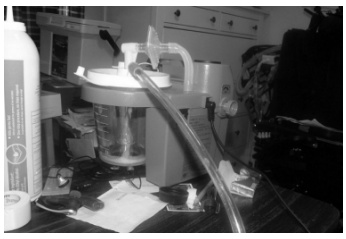

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Reference: http://www.speech-therapy-on-video.com/

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Communication

Expressive Aphasia: inability to convey thoughts through the use of speech, language, or writing; difficulty naming, sentences short & incomplete

Receptive Aphasia: pt unable to understand spoken words - what you are saying; when speaking uses made up words; speech lacks meaning; will look at your body gestures to follow commands

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Ideas for Expressive Aphasia

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1. Beginning sound of a word or contextual cues
2. Gestures
3. Elaborate on the patient's utterances
4. Important features of a target word e.g. library: building, quiet, books, study
5. Picture & symbol communication either a board or computerized
6. Have pt engage in real life activities requiring spontaneous attempts of communicating
7. Simple yes/no questions

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Ideas for Receptive Aphasia

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1. Simplify language - use short sentences with basic words
2. Slow down the rate of your speech
3. Use pauses between words
4. Try to eliminate any distractions (like a television or radio)

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Ideas for Receptive Aphasia

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5. Speak to the person as an adult
6. Include the person in your conversations
7. Try not to correct the person's speech
8. Be patient - give plenty of time for responses
9. Communication comes in many forms: drawing, pointing, gesturing, and writing. Make use of whatever might work

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Patient/Family Education

Respite Care is Ok!!!

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1. Positioning

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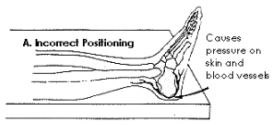
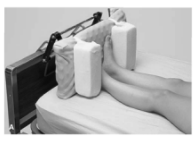
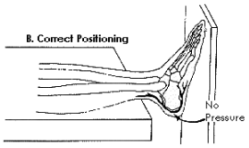
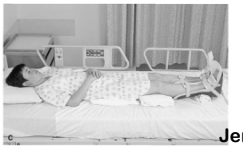
Right lateral position.









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CONTINUED™

Patient/Family Education

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2. Mobility:

- Frequently turn to avoid pressure ulcers
- Inspect skin daily

3. Physical Activity:

- Avoid fatiguing patient with any type of active or passive exercises or fxnl mobility
- Low repetitions
- Move slowly as to avoid any increases in pain

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Cognitive & Behavioral Changes

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Encourage CG to:

1. Educate her/himself.
2. Take care of her/himself.

Simplify communication:

1. Break sentences up into short phrases.
2. Ask yes/no questions.
3. Slow down when speaking.

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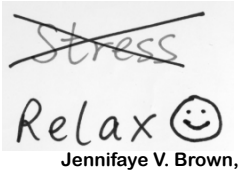
Cognitive & Behavioral Changes

99

Provide supervision & accompany the person to all appointments to assure info is accurately relayed & retained

Realistic expectations

Continue to enjoy relationships that bring joy



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Knowledge Summary

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Multiple Pathophysiologies:

- **resulting in degeneration of the motor neuron with the end result being death due to compromised breathing**

Major Types:

1. **Sporadic**
2. **Familial**
3. **Western Pacific**

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Subtypes:

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1. **Degeneration of the UMN**
2. **Primary Lateral Sclerosis**
3. **Progressive Bulbar Palsy**
4. **Progressive Muscular Atrophy**
5. **ALS Frontotemporal Dementia**

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Common Impairments

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1. Muscle weakness
2. Spasticity
3. Contractures
4. Dysphagia
5. Dysarthria
6. Respiratory Compromise
7. Pain
8. Sialorrhea: Inability to control secretions/salavia
9. Atrophy

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Optimal PT Interventions: Acute Phase for Gait Training

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1. AFO
2. Assistive device
3. Endurance training on treadmill
4. Resistive exercises for LE
5. Pt/CG education

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Optimal PT Interventions: Middle Phase for Fxnl Mobility

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1. Modification of transfer techniques: hoyer
2. Adaptive equipment for ADLs or positioning
3. Wheelchair assessment
4. Home evaluation for safety
5. Energy conservation techniques
6. Pt/CG education

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Optimal PT Interventions: Advanced Phase

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1. Consultation: maintenance for optimal QOL
2. Pt/CG education
   a. positioning: bed/wheelchair
   b. PROM
   c. transfer training
   d. CG burden

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Recommended Equipment Engage in Fxnl Activities

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DEPENDS ON PHASE/STAGE OF ALS

1. Rollator vs rw
2. Manual w/c versus sip & puff or tongue control
3. Computer
4. Neck brace
5. Hand splint
6. Chair lift

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Question and Answer Period

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Test Time to Evaluate Knowledge

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