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ALS Overview, Research and Assessment

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About me
- OT for 20 years, 10 with ALS
- AOTA Board Certified in Physical Rehabilitation
- RESNA Certified Assistive Technology Practitioner and Seating and Mobility Specialist
- Published articles, book chapters on OT and ALS
- Speak locally, nationally and internationally about ALS

Objectives
- Identify the hallmark characteristics of ALS
- Recognize the current research findings about persons with ALS and how to apply those findings
- List the most frequently used assessments and evaluations for the person with ALS
ALS
- Amyotrophic Lateral Sclerosis or Lou Gehrig’s Disease
- Affects brain and spinal cord motor neurons

Diagnosis
- Symptoms, EMG
- Rule out everything else: heavy metals, toxins, etc.
- Generally no sensory changes
- Often 1-2 years from symptoms to diagnosis
- Often other diagnoses are treated
  - Carpal tunnel
  - Spinal problems/surgeries
  - Overuse

ALS Symptoms
- Symptoms
  - Have to have BOTH upper and lower motor neuron signs
  - Upper motor neuron signs
    - Increased reflexes, spasticity, increased tone
  - Lower motor neuron signs
    - Weakness, atrophy, no/low reflexes, low tone
  - Voluntary muscles including respiration
  - Pseudobulbar, cognitive changes
  - May proceed in any direction

ALS- An Umbrella
- Amyotrophic Lateral Sclerosis- UMN and LMN
  - UMN Predominant ALS
  - LMN Predominant ALS
- Primary Lateral Sclerosis- UMN only
- Progressive Muscular Atrophy- LMN only
- Pseudobulbar Palsy
  - Speech, swallowing, breathing only
- Bibrachial Diplegia- upper body only
- Lower Extremity Diplegia- lower body only
ALS Facts

- 10% Familial, 90% sporadic
- Life expectancy: varies, average 2-5 years
- Incidence: 30,000 Americans at any given time
- Incidence: 2:100,000 total world population
- Prevalence: 6:100,000 total world population
- Incidence higher in people over 50 years old
- Ages 40-70 most common

ALS facts continued

- Cognition/behavior a factor in about 50% of people
  - Frontotemporal dementia, memory, judgment
  - Behavior: paranoia, personality changes, apathy
- All voluntary muscles affected, eye muscles generally last to go
- Can get “locked in”
- 5 senses spared, no sensory loss generally
- Most have bowel/bladder control

ALS Causes

- Causes
  - Free radicals
  - Glutamate
  - Auto immune
  - Toxins
  - Virus
  - Genetic risk factors
  - Stress
Cognitive/Behavioral Symptoms

- In about 50% of persons with ALS
- Frontotemporal Dementia - not memory like Alzheimer’s
- Alterations in thinking and behavior
- Might be irritable, inconsiderate, paranoid, apathetic, ritualistic, impulsive or otherwise act in uncharacteristic ways
- High level judgment, thinking can be affected

Pseudobulbar Affect

- Uncontrollable laughing or crying, out of proportion to feelings of happiness or sadness
- Interferes with social interactions
- Neudexta a treatment

Dysarthria

- Garbled speech, progressive
- Person tries to verbally communicate longer than they can be understood outside of immediate caregivers
- Eventually anarthric
- Communication devices
  - Start with Ipad/phone
  - Typing on Lightwriter
  - Communication Device - touch screen
  - Head mouse
  - Eye gaze

Communication Device - touch screen

- Head mouse
- Eye gaze
Dysphagia

- Difficulty with swallowing, moving food, forming bolus
- Generally not an aspiration risk until very late
- Feeding tube - get early
- Alter consistencies of food
- Hypermetabolic as well, higher calorie, higher protein
- Weight loss can be detrimental to life expectancy and strength

Respiratory

- Early BiPap/Trilogy volume ventilator use at night then more as progresses
- Difficulty lying flat
- VEST
- Cough Assist
- Suction
- Will eventually require ventilation or Hospice - many people use BiPap/Trilogy 24/7
- NO O2! Need diaphragmatic movement - not an oxygen issue

ALS - Early Symptoms

- Difficulty walking, tripping, falls
- Picking up objects
- Fine motor tasks
- Weakness & stiffness
- Atrophy of intrinsic muscles of hand
- Hyperactive reflexes & twitching
- Muscle cramping
ALS - Middle stages
- Weakness spreads to other muscle groups
- Fasciculations and cramping
- Pain for some
- Dysarthria/Dysphagia for some
- Assistance with ADLs needed
- Respiratory and swallowing issues - SOB, fatigue, laying flat
- Need for adaptive equipment for ADLs and mobility

ALS - Later stages
- Dependent on power/manual mobility
- Muscles flaccid
- Severe disabilities
- Communication difficult
- Often get PEG
- May consider Trach/vent
- Respiratory and swallowing issues/equipment
  - Volume ventilator, BiPap, no O2

ALS continued
- Generally die from respiratory compromise or pneumonia
- Hospice can assist with those who do not want a PEG or vent
- Consider Palliative Care early
- With PEG tube and ventilator, can continue to live, and with right wheelchair, communication device and technology, can live well
ALS - Medical Mgmt.

- No cure
- Treatment primarily palliative, symptomatic
  - Rilutek - (Riluzole) 25-28% longer survival
  - Vitamin D - 5000 units slows progression in 2 observational studies
  - Albuterol - increased respiratory muscle strength in 2 level 1 studies
- Multidisciplinary care affects survival
- Other medications prescribed for symptoms
  - Minimize drooling - Scopolamine, Botox
  - Muscle spasm/pain
  - Depression
  - Pseudobulbar - Neudex

ALSFRS-R

- ALS Functional Rating Scale Revised
- Used at every clinic visit in most multidisciplinary clinics to show changes
- Often used in clinical trials to show slope of change
- Not terribly sensitive
- 4 sub areas
  - Speech/swallowing
  - Arms
  - Legs
  - Respiratory

ALSFRS-R, 4 to 0 scores

1. Speech Normal speech processes; Detectable speech disturbance; Intelligible with repeating; Speech combined with nonvocal communication; Loss of useful speech
2. Salivation Normal; Slight but definite excess of saliva in mouth may have nighttime drooling; Moderately excessive saliva - may have minimal drooling; Marked excess of saliva with some drooling; Marked drooling - requires constant tissue or handkerchief
3. Swallowing Normal eating habits; Early eating problems - occasional choking; Dietary consistency changes; Needs supplemental tube feeding NPO (exclusively parenteral or enteral feeding)
4. Handwriting Normal; Slow or sloppy - all words are legible; Not all words are legible; Able to grip pen but unable to write; Unable to grip pen
5. Cutting food Normal; Somewhat slow and clumsy, but no help needed; Can cut most foods, although clumpy and slow; Some help needed; Food must be cut by someone, but can still feed slowly; Needs to be fed
6. Dressing and hygiene Normal function; Independent and complete self-care with effort or decreased efficiency; Intermittent assistance or substitute methods; Needs attendant for self-care; Total dependence
ALSFRS-R continued

7. Turning in bed: Normal; Somewhat slow and clumsy, but no help needed; Can turn alone or adjust sheets, but with great difficulty; Can initiate, but not turn or adjust sheets alone; Helpless

8. Walking: Normal; Early ambulation difficulties; Walks with assistance; Non-ambulatory functional movement only; No purposeful leg movement

9. Climbing stairs: Normal; Slow; Mild unsteadiness or fatigue; Needs assistance; Cannot do

10. Dyspnea: None; Occurs when walking; Occurs with one or more of the following: eating, bathing, dressing (ADL); Occurs at rest; Difficulty breathing when either sitting or lying; Significant difficulty, considering using mechanical respiratory support

11. Orthopnea: None; Some difficulty sleeping at night due to shortness of breath; Does not routinely use more than two pillows; Needs extra pillow in order to sleep (more than two); Can only sleep sitting up; Unable to sleep

12. Respiratory Insufficiency: None; Intermittent use of BiPAP; Continuous use of BiPAP; Continuous use of BiPAP during the night and day; Invasive mechanical ventilation by intubation or tracheostomy

ALS research - Exercise

- 2013 Cochrane review
- 2 small randomized control trials
- Too small to determine if beneficial or harmful
- Both studies showed increase in ALSFRS-R

Our clinic guidelines
- Grade 3+ or above, avoid eccentric
- Submaximal, no “feel the burn”
- Pool, recumbent bike, general aerobic, stretching

ALS research - Pain

- Rivera from 2013
- Pain reported in about half the patients
- Pain present at early, middle and late stages of the disease
- Pain correlated negatively with functional status
- Shoulder and bottom pain most common
ALS research - Effectiveness of OT

- Systematic review from 2014- AJOT
- Multidisciplinary care increased survival, higher quality of life, and higher percentage of appropriate assistive devices
- Home ex program of daily stretching and resistance ex improved function
- People with ALS are satisfied with comfort and ease of use of their power wheelchairs
- OT should be part of the Hospice care team

ALS research - Rehabilitation

- Majmudar, et. al. from 2014
- Nice general article about OT/PT/ST with ALS at the 3 stages
- Very little out there about what OT should do specifically with someone with ALS

ALS research - Wheelchairs

- Ward, 2010
- Trail, 2001
- Increased interaction in community
- Increase quality of life, ADL performance
- Satisfied with comfort and ease of use
- Experienced professionals needed to determine correct equipment over time
Other research in progress

- Stem cell use
- Various drugs and drug combinations
- Abnormalities in proteins
- Definitive blood/ICF test
- Genetic strains and tests
- Wheelchair and communication device control and access
- Brain-computer interface
- ALS and adaptive equipment use

ALS Information for Therapists

- OT Practice article - Occupational Therapy with Persons with ALS, Sept 2013
- Amyotrophic Lateral Sclerosis - A Patient Care Guide for Clinicians by Bedlack and Mitsumoto, Demos Medical
- http://mda.org/disease/amyotrophic-lateral-sclerosis
- www.ALSA.org

Assessments

- Dynamometry
- Pinch Gauge
- Barthel Index of Activities of Daily Living - 20 item, 5 minutes
- DASH - Disabilities of the Arm, Shoulder and Hand - 30 item, 10 minutes
- ADL assessments/performance - Informal, COPM
- Home set up and accessibility
- ROM - active and passive
- Strength - Manual muscle testing
- Equipment owned/used
- Leisure skills and performance
OT at an ALS Multidisciplinary Clinic

- Generally part eval, part treatment
- Loaner equipment
- Problem solving
- Function focused
- Education/handouts
- Proactive thinking
- After initial eval, see every 3 months

OT in Home Health

- Safety in home and during ADLs/mobility- alert system
- ADL, IADLs performance
- Pain management
- ROM, stretching
- Education
- Phone/computer use
- Equipment needs- Connect with OT at ALS Center

OT with ALS

- Definitely appropriate at every stage
- Important to educate yourself on available options
- Hook in with a multidisciplinary clinic OT in your community or state- ALSA or MDA
- Loaner equipment important
Second Webinar on ALS

- Specific Treatments
- Goal writing
- Adaptive Equipment/Techniques
- Creative Options

Join us!!

References


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