CLINICAL *review*

Amyotrophic Lateral Sclerosis (ALS)

Author
Amy Lombara, RPT

Reviewers
Joanne Minichillo, PT
Cinahl Information Systems
Glendale, California

Rehabilitation Operations Council
Glendale Adventist Medical Center
Glendale, California

Editor
Sharon Richman, MSPT
Cinahl Information Systems

September 19, 2008

Indexing Metadata/Description

- **Title/condition:** Amyotrophic Lateral Sclerosis (ALS)
- **Synonyms:** Lou Gerhrig's disease, Charcot's disease
- **Anatomical location/body part affected:** Affects both upper and lower motor neurons(1); eventually multisystem impairment arises
- **Description**
  - ALS defined
    - "A" stands for no; "myo" translates to muscle; "trophic" denotes nourishment or "no muscle nourishment"(2)
    - "Lateral" refers to the region of spinal cord impacted by ALS; when this area breaks down "sclerosis" sets in(2)
    - ALS is a progressive neurodegenerative disease of motor neurons(3)
  - Types of ALS
    - Sporadic
      - ~ 90-95% of all patients with ALS in U.S. have this type(2)
    - Familial
      - ~ 5-10% of all patients with ALS in U.S. have this type(2)
      - Genetic link; more than one person in heritage has been diagnosed with ALS(2)
      - One type of gene mutation identified thus far affects an enzyme called superoxide dismutase 1 (SOD1)(1)
      - Risk is 50% that children of patients with ALS (familial type) will later be diagnosed with ALS themselves(2)
    - Guamanian
      - In the 1950s an exceptionally high rate of ALS was witnessed in Guam and regions in Pacific(2)
  - Incidence
    - 2/100,000 people are afflicted(2)
    - ~ 5,000 people in the U.S. are diagnosed each year(4)
    - ~ 30,000 people are living with ALS in the U.S.(4)
    - Typical commencement of symptoms is between 40 and 70 years of age(3)
    - Can afflict people as young as age 20(4)
- **ICD-9 codes**
  - 335.20 amyotrophic lateral sclerosis
  - 335.22 progressive bulbar palsy
  - 335.29 other motor neuron diseases
- **ICD-10 codes**
  - G12.2 motor neuron disease
- **Reimbursement:** No specific issues or information regarding reimbursement have been identified
- **Presentation/signs and symptoms**
  - General
    - Pattern and progression of ALS can vary with each patient(2)
    - ALS is further broken down and subdivided based on which symptoms present first
      - Bulbar onset ALS
        - ~ 21% of patients with ALS have this form with onset of symptoms beginning in bulbar functions (swallowing, chewing, speaking, and breathing)(5)
      - Spinal onset ALS
        - ~ 75% of ALS patients have this form with onset in the extremities first(5)
    - Progressive muscle weakness in extremities will occur(2)
    - Progression of weakness is distal to proximal generally(6)
    - Weakness is asymmetrical at onset(7)
    - Progressive weakness in the muscles involved in respiration, swallowing, and speech formation will ultimately occur(2)

  - Progressive muscle atrophy will be seen[2]
  - Muscle fasciculation's are a frequent finding[6]
  - Pseudobulbar affect (episodes of uncontrolled laughing or crying) is found in approximately half of all patients with ALS[8]
  - When upper motor neurons (UMNs) become involved, spasticity and hyperreflexia will be present[7]
  - When lower motor neurons (LMNs) are impacted, findings include hyporeflexia and hypotonicity[6]
  - Impaired LMNs are also revealed by decreased strength, atrophy, and muscle fasciculations[9]
  - A combination of UMN and LMN symptoms will result; at times one may be masked by the other[3]
  - ALS does not customarily impact the senses or involuntary muscles[1]
  - Extraocular movements are generally spared as well[5]
- Early symptoms
  - Spinal onset ALS
    - Symptoms typically begin in the hands and feet (distal locations)[1]
    - Patient may report tripping, items falling out of his/her hands, or difficulty with activities of daily living[2]
    - Complaints of fatigue[2]
    - Foot drop[1]
    - Cramping of muscles; particularly in the feet and hands[2]
    - Decreased function in upper and lower extremities[2]
    - Decreased strength in neck extensor musculature; patient may complain of fatigue after reading[6]
    - Shortness of breath on exertion[6]
    - Spasticity may occur[8]
    - The patient has a hard time sleeping in supine related to increased tax on breathing[6]
  - Bulbar onset ALS
    - Symptoms start by affecting bulbar functions (swallowing, chewing, speaking, and breathing)[1]
    - Specifically the patient may have a hard time repositioning tongue, realigning lips, or opening/closing mouth[6]
    - Patient has trouble projecting voice[2]
    - Speech sounds "thick"[2]
    - Unwarranted drooling early on[6]
- Advanced symptoms
  - Unable to extend neck; "head drop or droop"[6]
  - Contractures[6]
  - Joint subluxations[6]
  - Pronounced weight loss[10]
  - Cognitive impairment[6]
  - Inability to speak (dysarthria)[6]
  - Paralysis[2]
  - Pain related to contractures and immobility[8]
  - Dependent edema secondary to lost muscle pumping through voluntary muscle contractions[8]
  - Patient will be short of breath at rest[6]
  - Breathing will be labored[2]
  - Difficulty/inability to swallow[2]
  - Mechanical ventilation eventually is required[2]

Causes & Risk Factors

- **Causes**
  - Largely unknown
  - Hypotheses
    - Increased glutamate found in patients' nervous systems, which can obliterate neurons[1]
    - In the case of SOD1 genetic mutation, it may be that free radicals build up, causing cell destruction[6]
    - Autoimmune response[6]
- **Pathogenesis**
  - Multiple components of pathogenesis have been found[5]
    - Protein aggregation
    - Glutamate excitotoxicty
    - Oxidative damage
    - Neurofilament abnormalities may occur in response to cellular injury or inflammation
    - Mitochondrial errors
    - Faulty axonal transport
  - Skeletal muscle malfunction may be a precursor of ALS[11]
- **Risk factors**

- As causes are largely unknown, so to are risk factors
- Hypotheses include
  - Smoking[5]
  - Lead ingestion[5]
  - Agricultural chemicals[5, 9]
  - Intake of dietary glutamate[5, 9]
  - Aluminum exposure[9]
  - Increased levels of selenium[9]
  - Gulf War service and neurotoxic contact[9]
  - Large amounts of dietary fat[9]
  - Athleticism (or individuals who are highly active) has been noted as a possible risk factor[11]
  - An association between ALS or an ALS-like syndrome in patients with HIV infection requires further investigation; approximately 19 cases of ALS or ALS-like disease have been reported in HIV-1 (HIV) positive patients in the last 20 years. An exploration into these cases reported no clear link between the two conditions. More research is necessary to determine if viral infections could contribute to acquiring ALS[12]

Overall Contraindications/Precautions

- Exercise
  - Patients with ALS should not be taxed through activities/therapy to the point where they reach exhaustion[8]
  - Allow frequent rests as needed; monitor closely for fatigue
  - The clinician should avoid placing burdensome eccentric loads on the patient[6]
  - If a patient does not have a muscle strength grade of at least a 3/5 (full range of motion against gravity), resistive exercise is contraindicated[13]
- Aspiration
  - Dysphagia is widespread in patients with ALS and places the patient at risk for aspiration[8]
    - Diet consistency may need to be modified
    - Patient education on swallowing techniques may be implemented
    - Ultimately a percutaneous endoscopic gastrostomy tube may be required as a result of persistent choking
- See specific **Contraindications/precautions** under Assessment/Plan of Care

Examination

- **History**
  - **History of present illness/injury**
    - **Etiology of illness**
      - When was the patient diagnosed with ALS?
      - Who comprises the patient's medical team?
      - Any recent hospital stays?
      - If disease is advanced, does patient have a PEG tube?
    - **Course of treatment**
      - **Medications for current illness/injury**
        - Determine what medications clinician has prescribed; are they being taken?
        - Riluzole is the only medication currently approved by FDA for treatment of ALS[3]
        - A recent study found the addition of lithium carbonate to riluzole may slow progression of ALS and may prolong survival[14]
      - **Diagnostic tests completed**
        - Diagnosis is made on the existence of UMN and LMN impairment with advancement of symptoms over the course of a year[7]
        - Diagnostic criteria[15]
          - Diagnosis of ALS requires
            - Presence of
              - Evidence of LMN degeneration by clinical, electrophysiological, or neuropathologic examination
              - Evidence of UMN degeneration by clinical examination
              - Progressive spread of symptoms or signs within a region or to other regions, as determined by history or examination
            - Absence of
              - Electrophysiological and pathological evidence of other disease processes that might explain the signs of LMN and/or UMN degeneration
              - Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs
              - Clinical diagnosis of ALS – without pathological confirmation
          - Requires assessment of LMN and UMN signs in 4 central nervous system regions: brainstem, cervical, thoracic, and lumbosacral spinal cord
          - Clinically definite ALS is defined as presence of UMN and LMN signs in 3 regions of the patient's body
          - Clinically probable ALS is defined as UMN and LMN signs in at least 2 regions, some UMN signs rostral to (above) LMN signs

        - Clinically probable/laboratory-supported ALS is defined as clinical UMN and LMN signs (or UMN signs alone) in only 1 region, and LMN signs defined by EMG criteria in at least 2 limbs, with diagnostic tests to exclude other causes
        - Clinically possible ALS – defined as clinical UMN and LMN signs in only 1 region or UMN signs alone in 2 or more regions or LMN signs rostral to UMN signs, without evidence from EMG testing, other diagnoses have been ruled out
    - **Home remedies/alternative therapies:** Document any use of home remedies (e.g., ice or heating pack) or alternative therapies (e.g., acupuncture) and whether or not they help
    - **Previous therapy:** Document whether patient has had occupational or physical therapy for this or other conditions and what specific treatments were helpful or not helpful
  - **Aggravating/easing factors** (and length of time each item is performed before the symptoms come on or are eased)
  - **Body chart:** Use body chart to document location and nature of symptoms
  - **Nature of symptoms:** Document nature of symptoms (constant vs. intermittent, sharp, dull, aching, burning, numbness, tingling). See Presentation/signs and symptoms above
  - **Rating of symptoms:** Use a visual analog scale or 0-10 scale to assess symptoms at their best, at their worst, and at the moment (specifically address if pain is present **now** and **how much**)
  - **Pattern of symptoms:** Document changes in symptoms throughout the day and night, if any (AM, mid-day, PM, night); also document changes in symptoms due external variables. As disease progresses pain may develop from increased dependency and disuse
  - **Sleep disturbance:** Document number of wakings/night, if any
  - **Other symptoms:** Document other symptoms patient may be experiencing that could be indicative of a need to refer to physician (dizziness, bowel/bladder/sexual dysfunction, saddle anesthesia)
  - **Respiratory status**
    - Any respiration supports presently?
    - Document oxygen saturation levels and respiratory rate during evaluation/treatment
  - **Barriers to learning**
    - **Are there any barriers to learning? Yes ❑ No ❑**
    - **If Yes, describe** _____
- **Medical history**
  - **Past medical history**
    - **Comorbid diagnoses:** Ask patient about other problems, including diabetes, cancer, heart disease, psychiatric disorders, orthopedic disorders, etc.
    - **Medications previously prescribed:** Obtain a comprehensive list of medications prescribed and/or being taken (including over-the-counter drugs)
    - **Other symptoms:** Ask patient about other symptoms he/she may be experiencing
- **Social/occupational history**
  - **Patient's goals:** Document what the patient hopes to accomplish with therapy and in general
  - **Vocation/avocation and associated repetitive behaviors, if any:**
    - Is patient working?
    - Does patient wish to continue working?
    - What are the patient's leisure activities?
    - A small case series investigation demonstrated the wish of some patients with ALS to maintain employment and their desire for the necessary modifications to enable this process[16]
    - Is a home/office assessment indicated?
    - An evaluation of necessary supports for augmenting employment may be indicated pending patient requests
  - **Functional limitations/assistance with ADLs/adaptive equipment**
    - What supports does patient have in place to date?
    - What are the barriers to the patient's mobility and functions; can modifications assist the patient in his/her desired tasks?
  - **Living environment:** Stairs, number of floors in home, with whom does patient live, caregivers, children, etc.; identify if there are barriers to independence in the home; any modifications necessary?

- **Relevant tests and measures: (While tests and measures are listed in alphabetical order, sequencing should be appropriate to patient medical condition, functional status, and setting.)** Evaluation procedures should be modified according to patient presentation. Key evaluation findings in the case of ALS are listed below; otherwise standard assessment techniques should be utilized
  - **Assistive and adaptive devices**
    - What are current devices utilized? Are they appropriate? (It is not uncommon for patient needs to change in a matter of weeks)
  - **Balance**
    - Evaluate static and dynamic balance reactions
    - The Tinetti Balance Test may be used in the earlier stages of ALS[6]
  - **Cardiorespiratory function and endurance**
    - Evaluate vital signs during rest and with activity
    - Does the patient utilize energy conservation techniques?
  - **Circulation**

  - Palpate pulses and compare
  - Document capillary refill time
- **Cranial/peripheral nerve integrity**
  - To assess bulbar function[10]
    - Ask patient to fill the cheeks with air
    - Whistle
    - Open/close jaw
    - Lip closure ability
    - Ability to move tongue
    - Phonation
  - Evaluate sensation
  - Sensory testing should be normal in patients with ALS[10]
- **Functional mobility** (including transfers, etc.): Evaluate mobility
- **Gait/locomotion**
  - Evaluate gait or wheelchair mobility
  - Assess for foot drop and pattern during gait
  - Assess need for assistive device or wheelchair
- **Motor function (motor control/tone/learning)**
  - Evaluate transfer status and need for assistive devices
  - Impulsivity is a potential finding in patients with ALS; document safety status during mobility[13]
  - The Physical Functioning sub-scale (PF10) may be a valuable tool for quantifying physical functioning in individuals with ALS[17]
    - The sub-scale is part of the larger, 36-item, Short Form health survey of the Medical Outcome Study (SF-36)
    - 198 patients post cerebral vascular accident, 151 with multiple sclerosis, and 193 with ALS participated in the research evaluating this outcome measure
    - Analysis demonstrated the sum score could be utilized as an indicator of overall physical performance among all diagnoses; there was only one item (#10) in the sub-scale for patients with ALS that didn't fall in line with the results as anticipated. This occurrence may have been the result of the item calling for the assessment of two different skills the authors postulated
- **Muscle strength**
  - MMT as indicated
  - Hand-held dynamometry or isokinetic strength testing as indicated[6]
  - Muscles will be atrophied and with decreased strength demonstrating LMN involvement; most notorious clinical symptom in patients with ALS[10]
- **Neuromotor development**
  - Spasticity may be present as UMNs become involved[10]
  - Hypotonia may be observed as LMNs become involved[10]
  - Modified Ashworth Scale may be used
- **Observation/inspection/palpation (including skin assessment)**
  - Especially important for patients with increased dependency; are there open areas or pressure sores?
  - Stage any areas noted
  - Does the patient have appropriate pressure relieving cushions/mattress?
- **Posture**
  - Does the patient present with "head drop"?
  - Evaluate overall posture
- **Range of motion**
  - Evaluate active and passive ROM
- **Reflex testing**
  - Evaluate deep tendon and abnormal reflexes
  - It is a typical finding in patients with ALS to have hyperreflexia coexisting with weak and significantly atrophied muscles. This indicates the existence of UMN and LMN impairment[18]
  - Positive Babinski and clonus test suggest UMN pathology[10]
- **Self-care/activities of daily living** (objective testing)
  - Observe or inquire about ADLs
- **Sensory testing:** Sensation is intact in patients with ALS
- **Special tests specific to diagnosis**
  - The Amyotrophic Lateral Sclerosis Assessment Questionnaire comprised of 40 items (ALSAQ-40) and the Amyotrophic Lateral Sclerosis Assessment Questionnaire comprised of 5 items (ALSAQ-5) may be simplified in the form of a sum score and used as a marker of global health status[19]
  - The ALS Severity Scale (ALS-SS)[20]
    - Four categories, including speech, swallowing, lower extremity, and upper extremity function

- Categories run from 1–10; 10 is normal
- Total sum score spans from 4–40 points

– The ALS Functional Rating Scale (ALS-FRS)[20]
- 10 items on test
- Each item can be scored from level 0 to 4
- 0 is unable and 4 is normal
- Best score is 40 points

Assessment/Plan of Care

- **Contraindications/precautions**
  - Exercise
    – Patients with ALS should not be taxed through activities/therapy to the point where they reach exhaustion[8]
    – Allow frequent rests as needed; monitor closely for fatigue
    – The clinician should avoid placing burdensome eccentric loads on the patient[6]
    – If a patient does not have a muscle strength grade of at least a 3/5 (full range of motion against gravity), resistive exercise is contraindicated[13]
  - Aspiration
    – Dysphagia is widespread in patients with ALS and places the patient at risk for aspiration[8]
      - Diet consistency may need to be modified
      - Patient education on swallowing techniques may be implemented
      - Ultimately a percutaneous endoscopic gastrostomy tube may be required as a result of persistent choking
- **Patients with this diagnosis are at risk for falls; follow facility protocols for fall prevention and post fall prevention instructions at bedside, if inpatient. Ensure that patient and family/caregivers are aware of the potential for falls and educated about fall prevention strategies. Discharge criteria should include independence with fall prevention strategies**
- **Diagnosis/need for treatment**
  - Multidisciplinary ALS management (team of at least one physical and occupational therapist, speech language pathologist, dietician, social worker, and a consultant in rehabilitation medicine; all team members treat a minimum of 6 patients with ALS per year) may improve the quality of life for individuals with ALS compared with general, nonspecialized ALS care[20]
    – 133 patients received multidisciplinary ALS care
    – 75 patients received general ALS care
    – A significantly greater percentage of individuals in the multidisciplinary ALS care group had appropriate assistive equipment
  - The multidisciplinary group had a significantly enhanced quality of life scores in the SF-36 mental summary
- **Rule out:** (There are many conditions that require ruling out prior to confirming a diagnosis of ALS; below is a partial list. Patients with ALS ultimately will have **both** UMN and LMN symptoms)
  - Myasthenia gravis[7]
  - Polyradiculopathies[7]
  - Multifocal motor neuropathy[7]
  - Kennedy's disease or bulbospinal muscular atrophy[7]
  - Spinal muscular atrophy[7]
  - Primary lateral sclerosis[5]
  - CNS tumor[5]
  - Cervical spondylitic myelopathy[5]
  - CNS radiation injury[5]
  - Multiple sclerosis[5]
  - Syphilis[5]
  - Diffuse Lewy body disease[5]
  - Syringomyelia[10]
  - Spinal cord tumor[10]
  - Myelopathy plus radicuolopathy[10]
  - Lyme disease[10]
- **Prognosis**
  - ALS is unremittingly progressive; fatal in 2-5 years with spinal onset form, 6-18 months with bulbar onset ALS[3]
  - Respiratory failure is most frequent cause of death among individuals with ALS[21]
  - Attending a multidisciplinary clinic designed for ALS patients may prolong attendee's survival[22]
    – 82 individuals with ALS were treated at a multidisciplinary ALS clinic; average age 60.1 years
    – 262 individuals with ALS were treated at a general neurology clinic; average age 65.6 years
    – Median survival for the ALS clinic patients was 7.5 months greater than for the general clinic patients

- Patients with bulbar onset had an increased survival of 9.6 months compared to general clinic bulbar onset patients
- 99% of patients in the ALS clinic received the drug riluzole; 61% received the drug in the general clinic

- **Referral to other disciplines**
  - Counseling for assistance coping with a terminal illness
  - Social worker for assistance with decision making throughout course of disease[10]
  - Speech language pathologists
    - An analysis of the use of augmentative and alternative communication (AAC) in 50 individuals with ALS was finalized after 4 years[23]
      - 96% of the subjects utilized the assistive technology
      - All of the participants who accepted the technology maintained their use of AAC over the course of the study
  - Respiratory therapist to assist with ventilation strategies as disease progresses
  - Hospice as end of life approaches[10]
- **Treatment summary**
  - Goals/treatment will vary depending on disease progression and patient wishes
  - Reports have been cited that patients with ALS may sense their clinician encapsulates their entire being into the diagnosis of ALS. ALS patients may feel their identity has become the disease and not just a part of who they are. Therefore, recommendations have been made to open communication between clinician and patient with regard to each patient's own goals, desires, and pursuits in order to support quality of life in these individuals[24]
  - Therapeutic exercises
    - The role of exercise for the treatment of patients with ALS has not been clearly defined by the research. The studies available are of small sample size and not all were randomized. The clinician is cautioned to take into account each individual's activity tolerance and implement strategies as appropriate
    - Regular exercise may have a short-term positive impact on the functioning of patients with ALS[25]
      - A randomized controlled trial of 25 patients with ALS was conducted
      - The patients were divided into two groups
        - Moderate daily exercise
        - Control or no activity beyond ADLs
      - After 3 months, the treatment group demonstrated less of a decline on the Ashworth Scale and ALS functional rating scale (ALS-FRS)
      - After 6 months there were no significant differences noted between groups, although trends were observed in the treatment group towards less decline
    - Resistance exercise may improve function in patients with ALS[26]
      - A randomized controlled trial of 27 eligible patients; required forced vital capacity of 90% or more and an ALS-FRS score of at least 30
      - Treatment group performed exercises and stretching; 3x per week – 8 subjects completed trial
      - Control group completed stretches only – 10 subjects completed trial
      - After 6 months the treatment group had significant improvements in ALS-FRS and SF-36 physical function subscale scores
      - No unfavorable reactions were reported in the treatment group
    - Clinically supervised exercise programs for ambulatory patients with ALS may provide significant improvements in ROM, strength, and functional level in the short term[27]
      - 26 ambulatory patients with ALS participated in the nonrandomized study
      - The study was comprised of two groups
        - Supervised exercise group
        - Home program group
      - Supervised program consisted of proprioceptive neuromuscular facilitation (PNF) techniques, breathing strategies, stretching, and functional mobility training 3 days/week for 8 weeks
      - Home program consisted of breathing exercises, active/active assisted ROM, walking and stretching daily
      - The supervised groups had significant gains in ROM and strength
      - During the time frame of 1 year, the supervised group was better able to maintain functional capacity level
  - Treatment of spasticity
    - Endurance-type exercises (individualized, moderate intensity) for trunk and extremities may help to reduce spasticity in patients with ALS[28]
      - Based on a systematic review of randomized or quasi-randomized trials for the treatment of spasticity in patients with ALS
      - Only 1 randomized trial met the study's inclusion criteria
      - 25 patients with ALS were randomized to moderate intensity, endurance-type exercises 15 minutes 2x/day vs. usual activities for 3 months
      - Significant gains were observed at 3 months in overall spasticity for the treatment group; however, results were not statistically significant when evaluating the mean change from baseline to 3 months against the control group
      - Review concludes insufficient evidence on this topic available in the literature
  - Airway clearance techniques
    - Noninvasive ventilation (NIV) may improve survival in patients with ALS without severe bulbar dysfunction, and maintain or improve quality of life for these patients[29]

- 92 patients participated in the randomized controlled trial
- Patients either received NIV or standard care for their symptoms
- A subgroup of patients with superior bulbar function in the NIV group (20 participants) had a median survival gain of 205 days
- Multiple quality of life indices were improved in this treatment subgroup
- The individuals with severe bulbar involvement in the NIV group had some improvement in quality of life measures, but it NIV did not significantly impact survival

– Nocturnal mechanical ventilation may reduce symptoms and prolong survival in patients with chronic hypoventilation caused by neuromuscular disease[30]
  - Based on Cochrane systematic review
  - 8 randomized trials were included that evaluated nocturnal mechanical ventilation in 144 patients with stable chronic hypoventilation with various causes
  - Weak evidence was found suggesting a benefit in patients with motor neuron disease; however, no evidence of benefit found in patients with chest wall disorders (2 trials included patients with hypercapneic ALS)
  - There was insufficient evidence to compare invasive vs. noninvasive mechanical ventilation or intermittent positive pressure vs. negative pressure ventilation

– High-frequency chest wall oscillation (HFCWO) may decrease the sense of breathlessness in patients with ALS[31]
  - Randomized controlled trial of patients with ALS; 46 enrolled and 35 completed the trial
  - 19 patients underwent HFCWO
  - 16 patients served as a control
  - After 12 weeks of intervention significant findings included
    – Less breathlessness in the treatment group
    – More coughing at night in the treatment group
    – Increased noise with respiration in the control group
    – No significant differences between groups for forced vital capacity, peak expiratory flow, capnography, oxygen saturation levels, or transitional dsypnea index were demonstrated
    – A trend was noted in the treatment group in regard to forced vital capacity; a deceleration of decline was observed
    – Patient satisfaction was reported to be 79% with the HFCWO

– The addition of HFCWO to standard care for the prevention of pulmonary complications in patients with ALS may not improve outcomes, including the prolongation of life[21]
  - Small randomized controlled trial involving 9 patients
  - The patients either entered a control group or treatment group receiving the HFCWO
  - The group receiving the HFCWO received the additional treatment 2x/day for 15 minutes each session
  - There were no significant differences seen among the groups when comparing the rate of decline of forced vital capacity or time to death

Problem	Goal	Intervention	Expected Progression	Home Program
Decreased ROM/ contractures	Maintain functional ROM/ prevent contractures	**Therapeutic exercise** ROM should be an integral part of treatment and home exercise program to prevent the occurrence of joint contractures[18]	Progress as indicated and able, given each patient's unique circumstances	Develop a home program that includes management of ROM/contracture prevention
Decreased strength/ eventual paralysis	Improve/slow decline of strength	**Therapeutic exercise** Promote therapeutic exercises as appropriate and indicated (**see contraindications**)	Progress as indicated and able, given each patient's unique circumstances	Develop a home program that addresses muscle strengthening as indicated and appropriate

Problem	Goal	Intervention	Expected Progression	Home Program
Decreased mobility/ function; decreased coordination; impulsivity	Promote functional independence and manage fatigue as able; improve/ assist coordination; educate caregivers on impulsivity	**Functional training** **Transfers** According to evaluation findings and disease progression a pivot board, slide board or mechanical lift may be indicated to facilitate transfers[32] A hospital bed may facilitate patient comfort and mobility[32] Educate the caregivers as indicated for safety strategies during mobility when impulsivity has been noted during assessment[13] **Gait/mobility** Depending on the patient's evaluation findings, an assistive device may be indicated to promote independence and safety while the patient remains ambulatory **Prescription, application of devices and equipment** **Feeding** Built-up utensils, rocker knives, cuffs, and mobile arm supports may promote independence[32] To promote a functional grip on an object, the suggested diameter is 33 mm[13] **Dressing** Button hooks and zipper pulls[32] Reachers and sock aids[10] **Orthotics/supports** Cervical collars may be indicated when active neck extension is impaired[32] Resting hand splints to prevent contracture formation[32] Cock-up or short-opponens splints may promote ability to perform certain ADLs[32]	Progress as indicated and able, given each patient's unique circumstances	The patient and family will most likely require instruction on body mechanics and safety during transfers Implement home modifications/assistive devices as necessary to ease mobility and promote independence

Problem	Goal	Intervention	Expected Progression	Home Program
		The GiveMohr sling has been recommended for control of shoulder subluxation[13] AFOs in the treatment of foot drop; must consider weight as fatigue is a major concern in this population[13] **Home adaptations** Stair lifts, ramps[32] Tub bench, raise toilet seat and rails, removable shower head[32] **Assistive devices** Patients with respiratory compromise may expend more energy using an assistive device for ambulation vs. no device Rolling walkers are generally recommended due to decreased energy cost when compared to standard walkers[13] Ensure patient has sufficient hand strength to utilize walker/cane/ crutches **Wheelchairs (w/c)**[13] A companion (a.k.a. transport) w/c may be appropriate for extended distance mobility when fatigue would prohibit ambulation; the companion weighs only ~20 pounds A power w/c will likely become necessary as disease progresses; modifications to the power w/c should be specific to the patient[13]		
Pressure areas; pressure wounds; pain	Relieve pressure/ independent weight shifts; promote wound healing; reduce pain	Integumentary repair and various protection techniques **Educate family/caregivers** Topics include: Assistance with mobility, prevention of contractures Pressure relieving techniques	N/A	Provide appropriate pressure relieving equipment

Problem	Goal	Intervention	Expected Progression	Home Program
Decreased endurance/ ventilation needs	Improve/slow decline of general endurance	Instruct patient on energy conservation techniques Low intensity aerobic exercise is generally recommended, including aquatic therapy, walking, and stationary bicycling[10]	Progress as indicated	Implement/recommend a home program as appropriate to address the promotion of cardiovascular stamina

Desired Outcomes/Outcome Measures

Desired outcomes include those listed above under "Goal". Outcome measures include

- The ALS Severity Scale (ALS-SS)
- The ALS Functional Rating Scale (ALS-FRS)
- Amyotrophic Lateral Sclerosis Assessment Questionnaire comprised of 5 items (ALSAQ-5)
- The Physical Functioning sub-scale (PF10)

Maintenance or Prevention

- Energy conservation strategies are essential for patients with ALS to limit fatigue[32]
- HEP for ROM, pressure relieving strategies, and therapeutic activities as indicated

Coding Matrix

References in this Clinical Review are rated using the following codes, listed in order of strength:

M Published meta-analysis
SR Published systematic or integrative literature review
RCT Published research (randomized controlled trial)
R Published research (not randomized controlled trial)
C Case histories, case studies
G Published guidelines
RV Published review of the literature
RU Published research utilization report
QI Published quality improvement report
L Legislation
PGR Published government report
PFR Published funded report
PP Policies, procedures, protocols
X Practice exemplars, stories, opinions
GI General or background information/texts/reports
U Unpublished research, reviews, poster presentations or other such materials
CP Conference proceedings, abstracts, presentations

References

1. Mayo Clinic staff. Amyotrophic Lateral Sclerosis. MayoClinic Web site. http://www.mayoclinic.com/health/amyotrophic-lateral-sclerosis/DS00359. Updated December 1, 2006. Accessed October 6, 2008. **(GI)**
2. ALS Association. About ALS. The ALS Association Web site. http://www.alsa.org Updated September 2008. Accessed October 6, 2008. **(GI)**
3. DynaMed Editorial Team. Amyotrophic Lateral Sclerosis. DynaMed Web site. http://www.ebscohost.com/dynamed Updated March 18, 2008. Accessed October 6, 2008. **(RV)**
4. Krivickas LS. Amyotrophic lateral sclerosis and other motor neuron diseases. *Phys Med Rehabil Clin N Am*. 2003;14(2):327-345. **(RV)**
5. Shoesmith CL, Strong MJ. Amyotrophic lateral sclerosis: update for family physicians. *Can Fam Physician*. 2006;52(12):1563-1569. **(RV)**
6. Bello-Hass VD. Amyotrophic lateral sclerosis. In: O'Sullivan SB, Schmitz TJ. *Physical Rehabilitation*. 5th ed. Philadelphia:F.A. Davis Company; 2007:819-849. **(GI)**
7. Patel SA, Maragakis NJ. Amyotrophic lateral sclerosis: pathogenesis, differential diagnoses, and potential interventions. *J Spinal Cord Med*. 2002;25(4):262-273. **(RV)**
8. Borasio GD, Voltz R, Miller RG. Palliative care in amyotrophic lateral sclerosis. *Neurol Clin*. 2001;19(4):829-847.**(RV)**
9. Wicklund MP. Amyotrophic lateral sclerosis: possible role of environmental influences. *Neurol Clin*. 2005;23(2):461-484. **(RV)**
10. Han JJ, Carter GT, Hecht TW, Schuman NE, Weiss MD, Krivickas LS. The Amyotrophic Lateral Sclerosis Center: a model of multidisciplinary management. *Critical Reviews in Physical & Rehabilitation Medicine*. 2003;15(1):21-40. **(RV)**
11. Abmayr S, Weydt P. Skeletal muscle in amyotrophic lateral sclerosis: emerging concepts and therapeutic implications. *Phys Med Rehabil Clin N Am*. 2005;16(4):1091-1098. **(RV)**
12. Verma A, Berger JR. ALS syndrome in patients with HIV-1 infection. *J Neurol Sci*. 2006;240(1-2):59-64. **(C)**
13. Lewis M, Rushanan S. The role of physical therapy and occupational therapy in the treatment of amyotrophic lateral sclerosis. *NeuroRehabilitation*. 2007;22(6):451-461. **(RV)**
14. Fornai F, Longone P, Cafaro L, et al. Lithium delays progression of amyotrophic lateral sclerosis. *Proc Natl Acad Sci U S A*. 2008;105(6):2052-2057. **(RCT)**
15. World Federation of Neurology. Amyotrophic Lateral Sclerosis. Revised Criteria for the Diagnosis of Amyotrophic Lateral Sclerosis. WFN Website. Http://www.wfnals.org Updated March 25, 2003. Accessed October 6, 2008. **(PP)**

16. McNaughton D, Light J, Groszyk L. “Don’t Give UP”: Employment experiences of individuals with amyotrophic lateral sclerosis who use augmentative and alternative communication. *AAC*. 2001;17:179-195. **(R)**
17. Dallmeijer AJ, de Groot V, Roorda LD, et al. Cross-diagnostic validity of the SF-36 physical functioning scale in patients with stroke, multiple sclerosis and amyotrophic lateral sclerosis: a study using Rasch analysis. *J Rehabil Med*. 2007;39(2):163-169. **(R)**
18. Cristini J. Misdiagnosis and missed diagnoses in patients with ALS. *JAAPA*. 2006;19(7):29-35. **(RV)**
19. Jenkinson C, Norquist JM, Fitzpatrick R. Deriving summary indices of health status from the Amyotrophic Lateral Sclerosis Assessment Questionnaires (ALSAQ-40 and ALSAQ-5). *J Neurol Neurosurg Psychiatry*. 2003;74(2):242-245. **(R)**
20. Van den Berg JP, Kalmijn S, Lindeman E, et al. Multidisciplinary ALS care improves quality of life in patients with ALS. *Neurology*. 2005;65(8):1264-1267. **(R)**
21. Chaisson KM, Walsh S, Simmons Z, Vender RL. A clinical pilot study: high frequency chest wall oscillation airway clearance in patients with amyotrophic lateral sclerosis. *Amyotroph Lateral Scler*. 2006;7(2):107-111. **(RCT)**
22. Traynor BJ, Alexander M, Corr B, Frost E, Hardiman O. Effect of a multidisciplinary amyotrophic lateral sclerosis (ALS) clinic on ALS survival: a population based study, 1996-2000. *J Neurol Neurosurg Psychiatry*. 2003;74(9):1258-1261. **(R)**
23. Ball LJ, Beukelman DR, Pattee GL. Acceptance of augmentative and alternative communication technology by persons with amyotrophic lateral sclerosis. *AAC Augment Altern Commun*. 2004;20(2):113-122. **(R)**
24. Foley G. Quality of life for people with motor neurone disease: a consideration for occupational therapists. *Br J Occup Ther*. 2004; 67(12):551-553. **(RV)**
25. Drory VE, Goltsman E, Reznik JG, Mosek A, Korczyn AD. The value of muscle exercise in patients with amyotrophic lateral sclerosis. *J Neurol Sci*. 2001;191(1-2):133-137. **(RCT)**
26. Bello-Haas VD, Florence JM, Kloos AD, et al. A randomized controlled trial of resistance exercise in individuals with ALS. *Neurology*. 2007;68(23):2003-2007. **(RCT)**
27. Aksu S, Karaduman A, Yakut Y, Tan E. The effects of exercise therapy in amyotrophic lateral sclerosis patients. *Fizyoterapi Rehabilitasyon*. 2002;13(3):105-112. **(R)**
28. Ashworth NL, Satkunam LE, Deforge D. Treatment for spasticity in amyotrophic lateral sclerosis/motor neuron disease. *Cochrane Database Syst Rev*. 2006;(1):CD004156. doi: 10.1002/14651858.CD004156. **(RCT)**
29. Bourke SC, Tomlinson M, Williams TL, Bullock RE, Shaw PJ, Gibson GJ. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomized controlled trial. *Lancet Neurol*. 2006;5(2):140-147. **(RCT)**
30. Annane D, Orlikowski D, Chevret S, Chevrolet JC, Raphaël JC. Nocturnal mechanical ventilation for chronic hypoventilation in patients with neuromuscular and chest wall disorders. *Cochrane Database Syst Rev*. 2007;(4):CD001941. doi: 10.1002/14651858. CD001941.**(SR)**
31. Lange DJ, Lechtzin N, Davey C, et al. High-frequency chest wall oscillation in ALS: an exploratory randomized, controlled trial. *Neurology*. 2006;67(6):991-997. **(RCT)**
32. McGovern-Denk M, Levine M, Casey P. Approaching occupation with the person with amyotrophic lateral sclerosis. *Phys Disabil Spec Interest Sec Q*. 2005;28(4):1-4. **(G)**