

AMYOTROPIC LATERAL SCLEROSIS (ALS) SYSTEM OF CARE PROCEDURES

- 1. REASON FOR ISSUE:** This Veterans Health Administration (VHA) Handbook establishes procedures for health care services to Veterans with Amyotrophic Lateral Sclerosis (ALS).
- 2. SUMMARY OF CONTENTS:** This is a new VHA Handbook describing the essential components and procedures of the ALS System of Care that have been implemented by ALS care providers to ensure that all enrolled Veterans have access to ALS care. Necessary structural, procedural, and educational components for consistent ALS services are described.
- 3. RELATED ISSUE:** None.
- 4. RESPONSIBLE OFFICE:** The Office of Patient Care Services (10P4) is responsible for the contents of this Handbook. Questions may be referred to the National Director of Neurology at 202-461-7120.
- 5. RESCISSIONS:** None.
- 6. RECERTIFICATION:** This VHA Handbook is scheduled for recertification on or before the last working day of July, 2019.

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AMYOTROPHIC LATERAL SCLEROSIS (ALS) SYSTEM OF CARE PROCEDURES

1. PURPOSE: This Veterans Health Administration (VHA) Handbook establishes procedures for health care services to Veterans with Amyotrophic Lateral Sclerosis (ALS). It describes the essential components and procedures of the ALS Program that have been implemented nationally to ensure that all enrolled Veterans, wherever they live, have access to ALS care. Necessary structural, procedural, and educational components for consistent ALS services are described. **AUTHORITY:** 38 U.S.C. Sections 501, 1701, 1703, 1704, 1710, 1712, 1717, 1720, and 1721.

2. BACKGROUND:

a. A study released by the Institute of Medicine (IOM) on November 10, 2006, *Amyotrophic Lateral Sclerosis in Veterans: Review of the Scientific Literature*, concluded that "there is limited and suggestive evidence of an association between military service and later development of ALS." Based upon this IOM study, and because ALS is a disease that progresses rapidly once diagnosed, VA designated ALS as a presumptively-compensable illness for all Veterans with 90 days of continuously active service in the military. 38 CFR 3.318.

b. Approximately 3,600 Veterans with ALS received care from VHA between FY 2005 and 2009 with 1,400 dying by the end of this period. On average, 1,520 Veterans with ALS are seen in VHA in any fiscal year. Since ALS is a presumptive service-connected disability, it is unknown how many new patients will come to VHA for care. The IOM study noted a 1.5 fold increased ALS incidence in Veterans suggesting an annual incidence rate of 4.5 per 100,000 Veterans, yielding an estimated annual incidence of 1,055 Veterans with new onset ALS and a possible Veteran prevalence of 4,220 given current life expectancy exceeding 3 years.

c. ALS is a progressive neurodegenerative disease that affects the motor neurons in the spinal cord, brainstem, and motor cortex resulting in weakness, atrophy, and corticospinal tract signs in varying combination that typically results in death from respiratory failure within 3 to 5 years from onset of symptoms. It is a unique disease due to its dynamic and progressive nature and requires special expertise to diagnose and manage.

d. The major focus of clinical care is providing the highest-quality of life through the management of symptoms and emotional and physical suffering. ALS varies considerably among individuals regarding site of onset and rate of progression.

e. Men are affected with ALS more frequently than women. Most people are more than 50 years of age at the onset of symptoms, and the incidence increases with each decade of life. A recent IOM report noted that the annual incidence of ALS among adults over 18 years is between 2.5 to 3.0 per 100,000. The IOM report added that the likelihood of a person dying of ALS is increased 1.5 fold if that individual is a Veteran. The IOM report did not report on factors that underlie the increased incidence of ALS. Specifically, locality of service and history of combat exposure did not show an influence on the incidence of ALS among Veterans.

f. The etiology of ALS is not known. Most individuals who develop ALS have a sporadic form of the disease. Inherited forms of ALS are rare although in about 5 percent of cases the disease is familial; being inherited as an autosomal dominant trait.

g. In FY 2009, the Chief Patient Care Services Officer charged a National Task Group with the assignment to develop comprehensive services for Veterans with ALS based on national best practices. VA health care requires timely provision of patient-centric care delivered with support to caregivers articulated through standardized recommendations. Standardization ensures that each Veteran is offered consistent support and treatment options to help manage the disease including the impairments, complications, and functional problems that result from ALS. The National Task Group report calls for the establishment of ALS interdisciplinary care teams to meet VA's obligation to these service-connected Veterans. These specialized teams co-manage care with primary care providers or home-based primary care (HBPC) teams, and as appropriate, with hospice and palliative care services. Through the implementation of the above recommendations, we can deliver comprehensive services as close as possible to the Veteran's home in an efficient manner from the perspective of the Veteran. Structural, procedural, and educational changes are needed for consistent ALS services. Continuous monitoring of system impact and resource needs over time is advisable.

h. Section 1710 of title 38 United States Code set forth VA's general authority to provide medically necessary hospital and nursing home care and medical services to eligible veterans. VA's medical benefits package is codified at 38 CFR 17.38. Other treatment authorities may be evoked to provide needed care and services to eligible veterans, e.g., 38 U.S.C. 1703 (Contracts for Non-VA Healthcare), 1717 (Home Health Services, Home Improvements and Structural Alterations) 1720 (Community Nursing Home Care, Adult Day Health).

3. SCOPE: This Handbook describes the minimum requirements for ALS treatment and programming. It defines the requirements for services that must be provided at both regional Veterans Integrated Service Network (VISN) ALS clinics, and all VA medical facilities, given that care delivery must be co-managed between primary care providers and the ALS interdisciplinary care team using principles of Patient Aligned Care Teams (PACT). Thus, sites are strongly recommended to surpass these specifications in developing their ALS programs in accordance with their resources and opportunities. VISNs and facilities are encouraged to engage in clinical and educational innovation as well as research to develop new strategies of care. These approaches permit the incorporation of best practices throughout VHA.

4. RESPONSIBILITIES:

a. **Chief Consultant, Specialty Care Services.** The Chief Consultant, Specialty Care Services, is responsible for overseeing the integrated national system of ALS care standards within VHA.

b. **National Director of Neurology Services.** The National Director of Neurology Services is responsible for:

(1) Ensuring that comprehensive ALS specialty care is available for all Veterans within VHA.

(2) Ensuring that metrics are developed to assess the quality of ALS care delivered.

(3) Working with VISN and facility Directors to resolve local and regional issues in administering the ALS System of Care.

(4) Monitoring regional ALS programs and tracking progresses and barriers to progress.

c. **Veterans Integrated Services Network Director.** The VISN Director is responsible for:

(1) Working in collaboration with the ALS Teams within the VISN and the National Director of Neurology Services to ensure coordination of all components of health care for Veterans with ALS.

(2) Ensuring establishment of a specialized ALS interdisciplinary care team that is available to any patient needing its services within the VISN.

d. **Medical Facility Director.** The medical facility Director is responsible for ensuring that:

(1) ALS care is delivered by qualified, competent staff.

(2) All ALS professional staff have the clinical and administrative support they require in order to allow them to efficiently deliver the high-quality, interdisciplinary care outlined in this Handbook.

e. **Amyotrophic Lateral Sclerosis Team Physician.** The ALS team physician is responsible for:

(1) Starting the consultation by asking what the patient already knows or suspects.

(2) Giving the diagnosis to the patient and discussing its implications in a stepwise fashion.

(3) Checking repeatedly to ensure the patient understands what is said and is reacting appropriately to the verbal and nonverbal cues of the patient.

(4) Respecting the cultural and social background of the patient by asking whether the patient wishes to receive information or prefers that the information also be communicated to a family member.

(5) Asking whether the patient desires to have the diagnosis and subsequent information also communicated to a significant other. If the patient requests this action, the provider must obtain a release of information from the patient and document this in the medical record.

(6) Ensuring the diagnosis is always given in person and never by mail or telephone, with enough time available on the part of the physician to answer the patient's questions.

(7) Avoiding delivering information callously or taking away/not providing hope.

f. **Amyotrophic Lateral Sclerosis Interdisciplinary Team.** The ALS interdisciplinary team must provide the Veteran with printed materials about the disease, information on support and advocacy organizations, and informative websites on the internet. *NOTE: A letter or*

audiotape summarizing what the physician has discussed can be very helpful for the Veteran and family.

(1) The patient must be assured that the patient and the patient's family will not be on their own (unassisted) to manage the patient's care and condition, but will be supported by a comprehensive professional ALS interdisciplinary care team and also provided regular follow-up contacts as needed. The consulting physician should make arrangements for a follow-up contact (via telehealth, by telephone, in person, or by another method as appropriate to the needs of the patient) before the end of that consultation and schedule it ideally within 4-6 weeks (or earlier if needed).

(2) At least one ALS interdisciplinary care team needs to be established in each VISN. The recommended first step is to identify the physician team leader, incorporating other team members already managing care for this population. **NOTE:** *ALS care delivery does not follow a hub and spoke model since care must be provided locally.*

5. NATIONAL SYSTEM OF ALS CARE:

a. The ALS System of Care consists of at least one ALS interdisciplinary regional clinic and program in each VISN. The Coordinator/ Director of each ALS regional team clinic and program need to identify Veterans with ALS through diagnostic searches of VHA corporate databases to establish communications with local providers who are currently managing health care for Veterans with ALS.

b. The mission of the ALS System of Care is to address and manage the multiple medical, physical, functional, psychological, and social effects of ALS in order to make the symptoms tolerable, and to enhance quality of life of Veterans with ALS through clinical care, education, and research. The multiple effects of ALS necessitate an interdisciplinary team for the delivery of a comprehensive, well-planned approach to care.

(1) Interdisciplinary ALS care has been shown through research evidence to improve patient outcomes, including life expectancy and quality of life. VA has the ability to provide comprehensive interdisciplinary ALS care that includes not only essential disciplines but also VA-specific programs such as HBPC and integrated hospice and palliative care services.

(2) As a unified health care system, VA can provide care from diagnosis through the end-of-life, promoting patient-centered, caregiver-supported, smooth transitions of care as disease progression occurs. The establishment of structured clinics and coordinated delivery of comprehensive services are essential.

c. All Veterans with a diagnosis of ALS, those with suspected ALS, and those being evaluated for a diagnosis of ALS are included in the target population served.

d. In order to standardize care and meet or exceed community standards of care for the ALS population, all care provided to Veterans with ALS needs to conform, at a minimum, to the following clinical practice recommendations:

(1) Interdisciplinary clinic referral should be considered for managing patients with ALS to optimize health care delivery and prolong survival. Management of comorbid symptoms may increase quality of life for the ALS patient and should always be evaluated and addressed. The multidisciplinary team should be aware of common comorbid symptoms that develop in the ALS patient such as sialorrhea secondary to dysphagia, pseudobulbar affect cognitive decline and fatigue.

(2) The Interdisciplinary team should consider the risks and benefits for a wide range of available pharmacotherapy and be aware of the need to discontinue or withhold medications. Current examples in the literature include (see paragraph 23a):

(a) Botulinum toxins, Dextromethorphan/Quinidine or low-dose radiation therapy to the salivary glands to reduce saliva.

(b) Riluzole may be offered to slow disease progression but, may be discontinued for patients who develop fatigue.

(3) Partial or total enteral tube feedings such as Percutaneous Endoscopic Gastrostomy (PEG) placed by a Gastroenterologist, Interventional Radiologist, or other clinician credentialed to perform the procedure may be offered to patients with early symptoms of dysphagia to maintain nutrition/hydration or medications.

(4) Non-invasive ventilation (NIV) technologies such as biphasic positive airway pressure (BiPAP) may be considered to treat respiratory insufficiency, to lengthen survival, and may be considered to slow the decline of forced vital capacity and improve quality of life. Early initiation of NIV may increase compliance, and insufflations or exsufflation may be considered to help clear secretions (see paragraph 23a).

e. All other procedures are provided to supplement, enhance, or update treatment components that may not have a sufficient evidence base to have been included in this Handbook, but still inform and may improve quality service delivery to Veterans with ALS. In addition, these procedures take advantage of the full continuum of care and resources available within VHA to support the unique needs of these Veterans.

f. Care must be coordinated through a regionalized system in which primary care providers at each VA medical facility are organized around a VISN ALS interdisciplinary team. An ALS interdisciplinary team needs to be established in at least one VA medical facility for each VISN, with referrals within each VISN supplemented by primary care providers at each VA medical facility and a telehealth network to facilitate nationwide coverage. ALS teams, primary care providers, PACTs, and telehealth coordinators and consultants will maximize the use of VA's Computerized Patient Record System (CPRS), VA national clinical databases, and telehealth resources to improve care coordination, quality, access, and efficiency.

6. LOCATION OF CARE: The location of care is dictated by the needs of the Veteran with ALS. Care is predominately outpatient-based with periodic admissions that should be as close to the Veteran's residence as possible. Such may be delivered in a variety of settings and methods, including telehealth and telephone contact. Regardless of the location, if ALS is the primary or a

secondary diagnosis, it is recommended that non-VA providers of ALS care be made aware of the Veteran's enrollment in the VA system.

a. Overall, ALS care needs to be comprehensive, interdisciplinary, and co-managed among primary care providers and the ALS team. Care needs to be delivered as efficiently as possible from the perspective of the Veteran with: grouping of clinic appointments, minimization of travel to the care setting, and expedient delivery of durable medical equipment (DME).

b. ALS teams must provide specialized services as close to the Veteran as possible to obviate the need for travel.

c. Veterans and their caregivers must have a single ALS health care provider point-of-contact to facilitate support and appropriate services. *NOTE: On spinal cord injury SCI units the social worker and Advanced Practice Registered Nurse (APRN) or the Registered Nurse (RN) case manager may be the best points of contact.*

7. CLINICAL SERVICES: The ALS System of Care ensures timely access to services. The interdisciplinary care for Veterans with ALS must to be coordinated through ALS care teams, with appointments made in a timely manner to meet VA standards for all ALS-related and general clinical problems. The following items outline the broad clinical services offered to Veterans with ALS within VHA:

a. The ALS care team needs to address the full continuum of care for the patient with ALS and nurture the therapeutic relationship from diagnosis through the terminal phase of the disease.

b. Health practitioners need to assist patients in understanding the issues to be faced in the terminal phase of the disease in a timely and empathic fashion. High priority is to be placed on patient self-determination and autonomy as an underlying assumption in the therapeutic relationship. Delivery of both information and care must take into consideration the cultural and psychosocial context of the patient and the family.

c. Health practitioners need to provide patients and families with information that is timed appropriately for decision-making, and delivered well in advance of major management crossroads, especially for respiratory care. Moreover, decision-making is a dynamic process that is subject to change as the disease becomes more severe.

d. Health practitioners need to provide explicit assurances of continuity of care and commitment to relieve suffering.

e. Be aware of the importance of spiritual issues for the Veteran's quality of life and treatment choices. An ALS care team liaison with local pastoral care workers needs to be established in order to be able to address the needs of the patient and relatives.

(1) The spiritual needs of patients, caregivers, and family members needs to be assessed by the ALS interdisciplinary care team repeatedly as the disease progresses.

(2) The patient's spiritual and religious attitudes need to be recognized and accepted by all ALS health care providers.

f. Respite care is available to enrolled Veterans and is a vital tool in the effective management of these ALS patients (identification of the caregiver's stress and needs by the ALS interdisciplinary care team member is essential to timely provision of those benefits. See VHA Handbook 1140.02 for more information on respite benefits).

g. The use of Home Health Aides needs to be incorporated into comprehensive service planning and the coordination of ALS team point of contact. VA physicians should order homemakers/home health aide (HHHA) and services as appropriate for Veterans with ALS.

8. DIAGNOSIS: The diagnosis of ALS needs to be confirmed or excluded as quickly as possible. Veterans for whom ALS is suspected need to be referred with high priority by VHA clinicians to an ALS-experienced neurologist. The average time from onset of symptoms to confirmation of diagnosis of ALS is 9–13 months.

a. There are four cogent reasons for making the diagnosis as early as possible:

- (1) The progressive loss of motor symptoms causes anxiety and discomfort;
- (2) The patient can better plan the remaining part of the patient's life;
- (3) Unnecessary diagnostic testing can be avoided;
- (4) Neuroprotective medication can be initiated, thus salvaging as many neurons as possible.

b. All suspected new cases need to undergo prompt detailed clinical examinations by a neurologist, a Physical Medicine & Rehabilitation (PM&R) specialist, or a primary care provider who has experience in ALS assessment and care. A review of the diagnosis is advisable if there is no evidence of progression or if the patient develops atypical symptoms.

(1) A comprehensive diagnostic workup may include the following procedures:

- (a) History and physical examination, including a thorough neurological examination;
- (b) Electrodiagnostic tests including electromyography (EMG) and nerve conduction velocity (NCV);
- (c) Blood studies including high resolution serum protein electrophoresis;
- (d) Thyroid and parathyroid hormone levels (in cases with minimal spasticity, anti-Ganglioside (GM1) antibody titers are indicated to rule out a diagnosis of anti-GM1 antibody syndrome);
- (e) Urine studies, including 24 hour urine collection for heavy metals as clinically indicated after exam/history;
- (f) Radiographic studies, including magnetic resonance imaging (MRI); and
- (g) Myelogram of cervical spine, muscle and/or nerve biopsy as indicated.

(h) Optional for selected, atypical cases: *NOTE: These are not done in all cases and would lead to additional procedures that might not be indicated.*

1. Anti GM1 antibody.
2. Lumbar puncture.
3. Muscle or nerve biopsy.

(2) Repetition of the investigations may be needed if the initial series of tests do not result in a diagnosis.

(a) A person diagnosed with ALS is encouraged to seek a second opinion from another VA ALS expert, i.e., someone who diagnoses and treats patients with ALS and has training in this neurological specialty. Neuromuscular specialists or neurologists are the optimal clinicians to provide this level of expertise in diagnosis and care.

(b) A physician with knowledge of the Veteran and ALS should communicate the diagnosis to the Veteran in a direct, understandable, empathetic, sensitive, and supportive manner. The International Social Impact Survey (ISIS), an international study of the diagnostic process in ALS, indicated dissatisfaction with how the diagnosis was communicated in about half the cases.

9. TREATMENT: After diagnosis and for follow-up, ALS clinic referral is to be considered for managing patients with ALS to optimize health care delivery and prolong survival, and may be considered to enhance quality of life. Following clinical referral, interventions to minimize complication of ALS should be initiated.

10. NEUROPROTECTIVE TREATMENT: Patients with ALS may be offered treatment with Riluzole 50 milligrams (mg) twice daily. Realistic expectations for treatment effects and potential side effects need to be discussed with the patient and caregivers. Treatment needs to be initiated as early as possible after the patient has been informed of the diagnosis taking into account expected therapeutic benefits and potential safety issues (Class IA recommendation).

a. Patients treated with Riluzole are to be monitored regularly for safety, side effects, and adverse effects. All patients with a symptomatic motor neuron disease and carrying a superoxide dismutase 1 (SOD1) gene mutation need to be offered treatment with Riluzole.

b. Enteral tube feeding options such as a Percutaneous Endoscopic Gastrostomy (PEG) should be considered and offered to Veterans with symptoms of dysphagia to assist in maintaining nutrition, hydration, and for medication administration.

c. NIV is to be considered to treat respiratory insufficiency in order to lengthen survival, and may be considered to slow the decline of forced vital capacity and improve quality of life. Early initiation of NIV may increase compliance, and automatic cough assist device (insufflations or exsufflation) may be considered to help clear secretions (see paragraph 27a).

d. Currently, there is insufficient evidence to recommend treatment with other presumptive neuroprotective treatments, anti-oxidants Vascular Endothelial Growth Factor A (VEGF), stem

cells, Lys-Asp-Ile or lysine-aspartic acid-isoleucine (KDI) tripeptide, neurotrophic factors, and Creatine.

11. GENETIC TESTING AND COUNSELING: There are now several genes that can be assayed commercially (SOD1, FUS, FIG4, VEGF, C9ORF, ubiquilin, TDP43). This type of testing is only indicated if there is a positive family history. This cannot focus only on the D90A SOD1 mutation (actually the A4V mutation is the most common in North America). Clinical Deoxyribonucleic Acid (DNA) analysis for SOD1 gene mutation needs to be performed only in cases with a known familial history of ALS or in sporadic ALS (SALS) cases with the characteristic phenotype of the D90A mutation. Clinical DNA analyses for SOD1 gene mutations are not to be performed in cases with SALS with a typical classical ALS-phenotype.

a. Before blood is drawn for DNA analysis, the Veteran needs to receive genetic counseling, have time to think about the information received, and provide informed consent. If a qualified counselor is not available a referral to a non-VA counselor may be needed. The primary or ALS physician makes this referral.

b. Pre-symptomatic genetic testing is only to be performed in first degree adult blood-relatives of patients with a known SOD1 gene mutation. Testing needs to be performed only after a shared decision-making process has been used, as outlined in paragraph 12(a).

12. PROVISION OF CARE:

a. Care provision is optimal when co-managed among the primary care provider and the ALS interdisciplinary care team. In later stages of disease, ALS interdisciplinary care team co-manages and facilitates hospice care, while maintaining a supportive relationship with the Veteran and the Veteran's family.

b. The complex service needs of Veterans with ALS are best addressed through a care management approach where one individual oversees and integrates the interdisciplinary care needs of the ALS Veteran while also addressing the needs of their family members and caregivers consistent with VA's authority in 38 CFR 71.50. The role of this overseeing individual may be delegated to an ALS experienced neurologist or provider, an experienced ALS coordinator, Advanced Practice Registered Nurse (APRN) or to the Registered Nurse (RN) case manager. This individual will be appointed by the facility and also serves as liaison between specialty services and the primary care medical provider for the patient.

13. INITIAL VISIT WITH AMYOTROPHIC LATERAL SCLEROSIS INTER-DISCIPLINARY TEAM: The initial patient visit with the ALS interdisciplinary care team needs to include the patient's Primary Care Provider, which may be a member of a Spinal Cord Injury or Home Care PACT.

a. The ALS interdisciplinary care team must adhere to the following protocol in the initial visit:

(1) Introduce team members, their roles, and team process.

(2) Provide an overview of VHA services in support of ALS management.

(3) Identify the Points of Contact, i.e., the physician leader with oversight for ALS care, the physician leader of ALS team, and the APRN or RN ALS case manager.

(4) Explain the role of the primary care provider.

(5) Describe an overall plan of care to enhance the function and quality of life for the Veteran.

(6) Reinforce patient decision making and determination of care.

(7) Provide information on the prevention of common complications.

(8) Encourage and engage in meaningful social roles, vocational considerations, home modifications, driving and transportation issues.

(9) Identify caregiver support services needed.

(10) Identify [Assess] psychological well-being and mental health issues.

b. The ALS interdisciplinary care team must identify an APRN or Registered Nurse (RN) certified in neurology subspecialty (Certified Neuroscience RN (CNRN) from an American Association of Colleges of Nursing (AACN) certification program), to serve as the point-of-contact for the neurology team managing the patient if and when possible. For smaller VA medical facilities and those in rural areas, it may be difficult to find an APRN with these certifications; in these instances an RN experienced in caring for ALS patients would be acceptable.

14. PRIMARY CARE: Veterans with ALS are assigned a primary care provider who assumes ongoing responsibility for preventive care, general medical care, and health maintenance.

a. The primary care provider manages other medical conditions the Veteran may have, while the ALS team addresses ALS symptoms and disease management with comprehensive psychosocial support. Optimally, primary care is to be provided within a VA health care facility (VA medical facility, Community-Based Outpatient Clinic (CBOC), or Independent Outpatient Clinic (IOC)) but, may be co-managed outside the VA. Because the needs of those with ALS are similar to that of Veterans with SCI, some consideration should be given to having the ALS clinic within an existing SCI clinic.

b. The primary health care provider assumes ongoing responsibility for prevention and health maintenance.

c. In facilities with extensive ALS specialty care services, the ALS interdisciplinary care team is to be available to advise and assist primary care providers in coordinating and/or assuming care if the Veteran experiences an ALS exacerbation or other ALS-related problem.

d. In facilities without ALS specialty care services, the closest ALS interdisciplinary team identified within the network (Appendix A) regional program must be available to advise the primary care provider or assume care should it be necessary.

e. Coordination between primary care and ALS specialty care after inpatient admissions is strongly advised. *NOTE: The use of Telehealth and MyHealthe Vet is encouraged to facilitate this communication.*

15. ALS TEAM CARE: ALS patient care is optimal when the ALS team providers are involved throughout the course of the disease.

a. The ALS interdisciplinary care team needs to include:

(1) ALS Physician, Primary Care Provider (include by telehealth or teleconference when necessary);

(2) If available; APRN, CNRN or another qualified individual that:

(a) Has competencies documented and protocols in place to support the Veteran and caregivers.

(b) Provides support to the Veteran and Veteran's family using face-to-face meetings, tele-video conferencing, telephone, or secure messaging.

(c) Serves as case manager, coordinating PACT care and non-VA care into the Veteran's health care plan.

(d) Identifies, addresses, and engages the team in the management of care issues.

(3) Palliative Care or Hospice Care Team staff member.

(4) HBPC Team staff member.

(5) Psychiatrist.

(6) Speech-Language Pathologist.

(7) Physical Therapist.

(8) Occupational Therapist.

(9) Pharmacist.

(10) Psychologist (Clinical Health Psychologist, Rehabilitation Psychologist).

(11) Dietician.

(12) Pulmonologist.

(13) Respiratory Therapist.

(14) Social worker.

(15) Therapeutic Recreation Specialist.

(16) Assistive Technology Specialist.

(17) ALS Association representative (with the patient's consent).

(18) Chaplain from the patient's preferred faith or religion.

(19) Gastroenterologist or other clinician credentialed to perform procedures to facilitate feeding (as consultant).

(20) Veteran benefits liaison (as consultant at the Veteran's request).

b. The ALS interdisciplinary care team and clinic function with a Veteran-centered model of care, with the goal of providing easy access to multiple specialties at one time.

(1) Typically, the clinic is set up so that the Veteran remains in one clinic room and the different providers rotate through that patient's room during a period of 2 to 3 hours. By providing access to all disciplines in one setting, coordination of care is maximized, and the burden of transportation to multiple clinic visits is reduced.

(2) Each ALS interdisciplinary care team must establish at least 1 half-day clinic per month, or more often as needed as determined by the caseload.

(3) Clinics must be physically accessible for persons with impaired mobility and possess requisite lifts and related equipment. Regularly scheduled clinical visits, telephone calls, and/or telehealth visits need to be arranged for each patient every 3 months, and more frequently if needed. This is particularly the case in the first 6 months following diagnosis and in late stages of the disease. Patients with slowly progressing ALS can be seen once or twice a year.

(4) Depending on care needs, some patients see all disciplines at a particular visit, and some patients see only a subset of providers.

(5) Consultations with the gastroenterologist or other clinician who can perform feeding-assistive procedures as-needed for enteral tube feeding issues.

(6) Each patient needs respiratory testing at the initial visit and regularly thereafter by a Pulmonary Medical physician or respiratory therapist experienced in performing and interpreting the test and who is able to provide biphasic positive airway pressure (BiPAP).

(7) It is important that between visits, the ALS interdisciplinary care team maintain regular contact with the patient.

(8) Patient and caregiver information packets with resource lists are to be developed by the ALS interdisciplinary care team and provided to Veterans, families, and caregivers. Access to VA health care providers through telephone triage after hours is essential.

(9) At the end of each clinic, there needs to be an interdisciplinary team meeting that establishes a plan of care documented in the medical records (CPRS) and recommends a time frame for follow-up appointments.

16. SYMPTOMATIC AND FUNCTIONAL CARE MANAGEMENT:

a. The symptoms most likely to be encountered by patients with ALS at some point in their lives include:

- (1) Difficulty communicating (Dysarthria);
- (2) Dyspnea;
- (3) Insomnia;
- (4) Pain;
- (5) Discomfort other than pain;
- (6) Difficulty with mastication and swallowing (Dysphagia);
- (7) Depressed mood;
- (8) Anxiety; and
- (9) Confusion.

b. Services must be available to address the patient's needs for:

(1) Mental health issues and services including, but not limited to, psychopharmacotherapy, psychotherapy and other psychosocial services.

- (2) Caregiver services available under 38 CFR 71.50;
- (3) Social work services;
- (4) Financial issues, including benefit optimization;
- (5) Liaison with community services;
- (6) Mobility issues;
- (7) Activities of daily living via home health (VHA Handbook 1140.3);
- (8) Durable Medical Equipment and Assistive Technology;
- (9) Patient and caregiver education; and

(10) Speech Pathology Services for management of communication including motor speech, cognition, and swallowing.

c. The following symptom management domains need to be addressed:

(1) Weakness and impaired function;

(2) Swallow function and sialorrhea;

(3) Airway protection (impaired cough);

(4) Pulmonary function and Non-invasive or invasive ventilation;

(5) Intermittent or chronic dyspnea;

(6) Pseudobulbar emotional lability;

(7) Spasticity and/or cramps;

(8) Mood changes and insomnia;

(9) Pain management;

(10) Venous thrombosis prevention;

(11) Partial or Total Enteral or Parenteral nutrition;

(12) Communication (including motor speech/dysarthria, cognitive communication and non-verbal communication); and

(13) Advance care planning.

17. DUAL CARE: Current VHA policy (VHA Directive 2009-038) defines dual care as a system-wide approach to the coordination and provision of medical care to eligible Veterans who are seen by both VA and community providers.

a. Dual Care may be coordinated by the local VA facility Director in accordance with current VHA directives, VA policy, and applicable laws and regulations.

b. Dual Care may be used to optimize the appropriateness, safety, and efficacy of care, medications, prosthetics, and supplies provided to eligible Veterans who are seen by both VA and community providers.

c. Dual Care may be used to balance the need for accessible, local care with the need for interdisciplinary ALS specialty care. Direct referrals to the ALS Care Team from non-VA providers are appropriate services available.

18. PALLIATIVE CARE: Palliative care is a holistic approach to managing advanced or distressing ALS symptoms.

a. Goals of palliative care revolve around relieving suffering and supporting the best possible quality of life for patients with ALS, to include providing comfort and relief of symptoms caused by ALS. Although there is no cure for ALS, the aim of palliative care is to help the Veteran with ALS live each day to the fullest and with as much independence and comfort as possible.

b. Palliative care takes into account the whole patient, not just the physical side of an illness. It helps patients, their caregivers, and family members to understand the patient's illness and assists them in identifying treatment choices.

c. The ALS interdisciplinary care team focuses on symptom management and addresses palliative care needs. The ALS team co-manages (see paragraph 12a) and facilitates palliative care and hospice care, while maintaining a supportive relationship with the Veteran and the Veteran's family.

d. The physician or team member who takes the leadership role in end-of-life discussions is responsible for communicating the patient's treatment decisions to the patient's other VA health care providers.

e. A partnership between the patient and the ALS interdisciplinary care team members must be established and must include an end of life, palliative care, comfort care, or hospice care team early in the course of the disease; prior to the initiation of end-of-life discussion and during disease progression.

f. Discussions on end-of-life decisions also need to be initiated whenever the patient asks or "opens the door" for end-of-life information and/or interventions, including, but not limited to:

(1) Options for respiratory support, enteral feeding, and end-of-life issues if the patient has dyspnea, other symptoms of hypoventilation or a forced vital capacity (VC) less than (<) 50 percent.

(2) For symptomatic treatment of dyspnea and/or pain of intractable cause, use of Opioids alone, or in combination with benzodiazepines, if anxiety is present is preferred.

(3) Titrating the dosages against the clinical symptoms almost never results in a life-threatening respiratory depression (see paragraph 23j).

(4) For treating terminal restlessness and confusion because of hypercapnia, neuroleptics need to be used.

(5) Use oxygen only if symptomatic hypoxia is present. When withdrawing ventilation use adequate opiates and anxiolytics; to relieve dyspnea and anxiety. BiPAP is a very important intervention for dyspnea for many ALS patients. VA ALS interdisciplinary care teams must be providers capable of knowing when to prescribe BiPAP (pulmonary and/or respiratory therapy) and systems for providing the equipment and set-up for patients in their homes, either by rental or purchasing agreements. Approximately 2-10 percent of patients choose tracheostomy and long-term invasive mechanical ventilation (IMV).

g. Quality-of-life instruments (questionnaires) should be used for patients with advanced ALS during the end of life to help detect issues that should be addressed in order to improve the end-of-life (see paragraph 23d) patient-generated measures of individual quality-of-life that have high acceptance in ALS. The “ALS Specific Quality of Life-Revised (ALSSQOL-R)” (see paragraph 23e) is a specific instrument designed for people with ALS.

h. Consistent with the notification and screening requirements of VHA Handbook 1004 02, the patient needs to be informed about advance directives (AD) and how to designate a health care agent. If the ALS patient does not elect to execute an AD, it may be helpful to the patient to explain how consent for future treatment (including end of life care) will be obtained from the appropriate surrogate if the patient lacks decision making capacity. Assistance in formulating an AD needs to be offered. The patient’s preferences for life-sustaining treatments needs to be re-discussed every 6 months, or when there is any change in the patient’s condition.

19. HOSPICE CARE: Sixty-one percent of patients with ALS die at home. Hospice is a part of palliative care, providing care for those who are no longer responding to treatment or those near the end of life. It is included in VA’s medical benefits package and available to all enrolled Veterans (see 38 CFR 17.38(a)(1)(xi)(A); Bereavement Counseling, as authorized by 38 CFR 17.98, is also included in the package).

a. VHA must offer to provide hospice care that VHA determines is needed by the Veteran; this includes inpatient and home hospice care.

b. Veterans needing hospice care may choose to receive such care through a non-VA provider using non-VA benefits, such as Medicare. However, if the Veteran elects to receive hospice care from VA, then facility staff must assist the Veteran in obtaining it. **NOTE:** *VA may provide it in-house or, as appropriate, by contract.*

c. Hospice provides bereavement services to the Veteran’s family or other caregivers, even after the death of their family member. Bereavement support needs to be offered, both informally and formally, to the caregiver during the clinic visit, and after the death of the patient, to include letters of condolence at the time of death.

d. RNs in the role of 24-hour triage, either through a formal call center or other mechanism, must provide triage and basic support using protocols developed jointly by all members of the ALS interdisciplinary care team. For questions, concerns, or issues that fall outside of the areas addressed in the protocols, a neuromuscular specialists or neurologists is to be available as a back-up 24-hours a day.

20. COORDINATION WITH RELEVANT SERVICES: The case manager is responsible for performing the initial needs assessment to address DME, availability of community resources, and caregiver services.

a. Given the limited life expectancy for Veterans with ALS, there is a need to expedite provision of assistive technology (AT) and DME, which requires coordination with the Rehabilitation and Prosthetic Services. The case manager, primary care provider, or ALS care coordinator should alert the Prosthetic and Sensory Aid Service (PSAS) chief at the medical center to assure careful planning for timely provision of items needed with this sometimes

rapidly progressing disease. The wide variety of items offered by PSAS should be considered by the team. Some examples include, but are not limited to:

(1) Low-tech DME device, which should be available as “stock” for same-day provision. Low tech devices include, but are not limited to: the aids for dressing, bathing, grooming, eating and drinking; ambulation aids; standard wheelchairs; etc.;

(2) Wheelchair cushions, and off-shelf orthotic devices;

(3) Non-stock items, which the PSAS consult has a processing time of 1-5 days;

(4) Procurement and delivery of all other prescribed devices that are to be expedited to facilitate provision to the Veteran prior to further decline in function (e.g., power wheelchairs; home hospital beds; patient lifts; ventilators with backup; generators; and portable suction);

(5) Providers should also discuss the use of Home Improvement Structural Alternations (HISA) grants early in the disease process; HISA can be used to make changes or improvements in the home to improve accessibility for the Veteran.

b. Given the volume and complexity of AT devices that are utilized by the Veteran with ALS and the need to interface multiple electronic devices for varied environments of use (i.e., complex power wheelchair, augmentative communication device, computer access, environmental control, etc.), it is strongly recommended that services be coordinated by rehabilitation specialists with experience in assistive technology at a VA Assistive Technology Lab or ALS clinic.

21. CLINICAL OUTCOMES ASSESSMENTS: Clinical Outcome Assessments need to be monitored and assessed by psychometrically-sound assessments with proven reliability and validity when used with the ALS population. Some examples include the Appel ALS Rating Scale (see paragraph 23m), ALS Functional Rating Scale (ALFRS) (see paragraph 23n), or Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ) (see paragraph 23o).

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**AMYOTROPHIC LATERAL SCLEROSIS (ALS) INTERDISCIPLINARY TEAM CLINICS
AND PROGRAMS IDENTIFIED**

Veterans Integrated Services Network (VISN)	DEPARTMENT OF VETERANS AFFAIRS (VA) FACILITIES
VISN 1	Department of Veterans Affairs (VA) Connecticut HCS; VA Boston HCS; and Providence RI VAMC
VISN 2	VA Western New York Health Care System at Albany, NY
VISN 3	James J. Peters VA Medical Center in Bronx, NY
VISN 4	VA Pittsburgh Health Care System
VISN 5	VA Maryland Health Care System
VISN 6	Durham VA Medical Center
VISN 7	Ralph H. Johnson VA Medical Center
VISN 8	James A. Haley VA Medical Center in Tampa, FL; Miami VA Health Care System; and North Florida-South Georgia VA Health Care System
VISN 9	Tennessee Valley VA Health Care System and VA Medical Center in Lexington, KY
VISN 10	Louis Stokes VA Medical Center in Cleveland, OH
VISN 11	Richard L. Roudebush VA Medical Center
VISN 12	Edward Hines, Jr. VA Medical Center and Clement J. Zablocki VA Medical Center
VISN 15	St. Louis VA Medical Center
VISN 16	Oklahoma City VA Medical Center
VISN 17	VA North Texas Health Care System
VISN 18	New Mexico VA Health Care System
VISN 19	VA Eastern Colorado Health Care System in Denver, CO
VISN 20	VA Puget Sound Health Care System in Seattle, WA and Portland VA Medical Center
VISN 21	VA Palo Alto Health Care System and San Francisco VA Medical Center
VISN 22	VA Long Beach Healthcare System in Long Beach, CA and VA Greater Los Angeles Health Care System
VISN 23	Minneapolis VA Medical Center