Measure #4: ALS Symptomatic Therapy Treatment Offered

**Amyotrophic Lateral Sclerosis**

**Measure Description**

Percentage of visits for patients with a diagnosis of ALS with patient offered treatment* for pseudobulbar affect, sialorrhea, and ALS related symptoms**.

*ALS treatment examples: eg dextromethorphan/quinidine combination, amitriptyline or fluoxetine for pseudobulbar affect; anti-inflammatory and analgesic agents for pain; anticholinergic agents for sialorrhea; botulinum toxin for refractory sialorrhea; tizanidine or baclofen for spasticity; antidepressants for depression; physical therapy for cramps; occupational therapy for adapted devices; or a dietary modification for constipation.

**ALS related symptoms definition: eg spasticity, muscle cramps, pain, anxiety, depression, leg swelling, insomnia, fatigue, laryngospasm or constipation

**Measure Components**

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patient visits with patient offered treatment* for pseudobulbar affect, sialorrhea, or ALS related symptoms**, if present.</th>
</tr>
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<th>Denominator Statement</th>
<th>All visits for patients with a diagnosis of amyotrophic lateral sclerosis.</th>
</tr>
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</table>

| Denominator Exclusions | • No exclusions applicable for this measure. |

<table>
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<tr>
<th>Supporting Guideline &amp; Other References</th>
<th>The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>• In patients with ALS who have medically refractory sialorrhea, botulinum toxin B should be considered (Level B) and low-dose radiation therapy to the salivary glands may be considered (Level C).1</td>
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<td></td>
<td>• Provide a portable mechanical home suction device.2</td>
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<td></td>
<td>• Treat cramps in ALS with physiotherapy, physical exercise, and/or hydrotherapy.2</td>
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<tr>
<td></td>
<td>• Antispasticity drugs such as baclofen and tizanidine may be tried.2</td>
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<td>• Physical therapy should be available regularly when there is significant spasticity.2</td>
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Measure Importance

| Relationship to desired outcome | Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20%–50% of patients with ALS, especially in pseudobulbar palsy. Although it is not a mood disorder, antidepressants are frequently employed. Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients’ quality of life. A fixed-dose combination of dextromethorphan (DM)/quinidine (Q) (30 mg DM/30 mg Q BID) for treatment of pseudobulbar affect in ALS (Class I) reduced the frequency and severity of laughing and crying behaviors compared to either DM ($p<0.001$) or Q alone ($p<0.001$). Side effects were dizziness, nausea, and somnolence, which accounted for termination of treatment in 24% with DM/Q compared to 6% with DM and 5% with Q. This formulation of DM/Q was not approved by the US Food and Drug Administration (FDA). In a more recent study of 326 randomized patients, the PBA-episode daily rate was 49% ($p<0.0001$) lower for DM/Q-20/10 than for placebo. Other endpoints showing statistically significant DM/Q benefit included the likelihood of PBA remission during the final 14 days. The low dosage was safe and well tolerated. On the basis of these two trials, the lower dose DM/Q-20/10 was recently approved by the US Food and Drug Administration (FDA) for the treatment of PBA. |

| | Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications, mainly oral anticholinergic agents, for treatment reported benefit (Class III). A portable mechanical home suction device is often helpful. In a small trial, amitriptyline and botulinum toxin type A (BTxA) seemed equally effective, although 3 of 5 patients treated with amitriptyline experienced side effects (Class III). In a double-blind, controlled trial of botulinum toxin type B (BTxB) in 20 patients with ALS with refractory sialorrhea (Class I), patients were randomized to 2,500 U of BTxB or placebo into bilateral parotid and submandibular glands. Treated patients reported a global improvement of 82% at 2 and 4 weeks compared to 38% in placebo ($p=0.05$). At 12 weeks, 50% of patients receiving BTxB were improved compared to 14% receiving placebo. There were no important adverse events. Radiation therapy for medically refractory sialorrhea reduced salivary production, but side effects included erythema, sore throat, and nausea (Class III). A “satisfactory response” was observed and saliva secretion rate diminished with a single dose of 7–7.5 Gy bilaterally (Class III). |

| | Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue is a major issue for patients with ALS, and its etiology may be multifactorial (eg depression, anxiety, disturbed sleep, dyspnea). No single treatment is likely to be effective. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo ($p=0.07$; number needed to harm=8) (Class III). Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study ($p=0.004$; number needed to harm=17) (Class III). |

| | Treating spasticity might improve gait and relieve painful spasms. Moderate exercise led to a small decline in the Ashworth Spasticity Scale over 3 months, compared to a worsening with no exercise ($p<0.005$) (Class III). Physical therapy is the mainstay of treatment of spasticity in ALS, and has been shown to be effective in a Class II study. |

| | The prevalence of depression in ALS ranges from 0 to 44%, although systematic studies suggest 10% in advanced ALS (Class III). Depression shortens survival and lowers quality |

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of life for patients with ALS. There is consensus among experts that depression should be treated in patients with ALS; however, there are no controlled studies of benefit or harm.

Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain. There is a concern that sedative/hypnotic agents may suppress respiratory drive, tidal volume, and upper airway muscle tone in patients with ALS.

Laryngospasm is common in ALS and quite frightening to patients and families. Education about the benign nature of the symptom and its management is important.

References
Most other symptoms of ALS are treatable, albeit with less evidence. Still, most symptoms in ALS can be treated, as described above, and studies suggest that these treatments are underutilized.\(^1,2\)

**References**


| IOM Domains of Health Care Quality Addressed | Effective  
| Patient centered  
| Timely  |
| Exclusion Justification | No exclusions applicable for this measure.  
| Harmonization with Existing Measures | There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.  

**Measure Designation**

| Measure purpose | • Quality improvement  
| • Accountability  |
| Type of measure | • Process  
| Level of Measurement | • Individual practitioner  
| Care setting | • Ambulatory Care  
| Data source | • Electronic health record (EHR) data  
| • Administrative Data/Claims (inpatient or outpatient claims)  
| • Administrative Data/Claims Expanded (multiple-source)  
| • Paper medical record  

**Technical Specifications: Administrative/Claims Data**

Administrative claims data collection requires users to identify the eligible population (denominator) and numerator using codes recorded on claims or billing forms (electronic or paper). Users report a rate based on all patients in a given practice for whom data are available and who meet the eligible population/denominator criteria.

The specifications listed below are those needed for performance calculation. Additional CPT II codes may be required depending on how measures are implemented. (Reporting vs. Performance)

| Denominator (Eligible Population) | ICD-9–CM Diagnosis Codes:  
| 335.20 (amyotrophic lateral sclerosis)  

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CPT E/M Service Code:
99201, 99202, 99203, 99204, 99205 (office-new patient),
99211,99212, 99213, 99214, 99215 (office-established patient),
99241, 99242, 99243, 99244, 99245 (outpatient consult),
99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility),
99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary),
99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit)

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**ALS related symptoms definition: eg spasticity, muscle cramps, pain, anxiety, depression, leg swelling, insomnia, fatigue, laryngospasm or constipation

Reporting Instructions:
- For all patients meeting denominator criteria, report either 3756F, Patient has pseudobulbar affect, sialorrhea or ALS related symptoms OR 3757F, Patient does not have pseudobulbar affect, sialorrhea, or ALS related symptoms.
- When 3756F is reported, also report 4541F, Patient offered treatment for pseudobulbar affect, sialorrhea, or ALS related symptoms.

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Reporting Instructions:
- There are no exclusions for this measure. Do not report modifiers 1P, 2P or 3P with 4541F.