Measure #3: ALS Cognitive Impairment and Behavioral Impairment Screening

Amyotrophic Lateral Sclerosis

Measure Description

Percentage of patients diagnosed with ALS who are screened at least once annually for cognitive impairment (eg frontotemporal dementia screening or ALS Cognitive Behavioral Screen (CBS)) and behavioral impairment (eg ALS CBS).

Measure Components

<table>
<thead>
<tr>
<th>Numerator Statement</th>
<th>Patients who are screened at least once annually for cognitive impairment (eg frontotemporal dementia screening or ALS Cognitive Behavioral Screen (CBS)) and behavioral impairment (eg ALS CBS).</th>
</tr>
</thead>
<tbody>
<tr>
<td>Denominator Statement</td>
<td>All patients with a diagnosis of amyotrophic lateral sclerosis.</td>
</tr>
</tbody>
</table>
| Denominator Exclusions | • Documentation of a medical reason for not screening the patient for cognitive and behavioral impairment (eg patient currently diagnosed with severe cognitive impairment)  
• Documentation of a patient reason for not screening the patient for cognitive and behavioral impairment (eg patient declines to be screened for cognitive or behavioral impairment)  
• Documentation of a system reason for not screening the patient for cognitive and behavioral impairment (eg no insurance to cover screening cost) |
| Supporting Guideline & Other References | The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:  
• Screening for cognitive and behavioral impairment should be considered in patients with ALS (Level B).¹  
• Screening tests of executive function may be considered to detect cognitive impairment in patients with ALS prior to confirmation with formal neuropsychological evaluation (Level C).¹  


Measure Importance

Relationship to desired outcome

There is now considerable evidence for cognitive and behavioral manifestations in ALS. Specific ALS phenotypes include pure motor degeneration (ALS), ALS with cognitive impairment (ALSci), ALS with behavioral impairment (ALSbi), and ALS with a dementia meeting the Neary criteria for frontotemporal dementia (FTD) (ALS-FTD). FTD, as defined by Neary et al., has insidious onset, gradual progression, altered social conduct, emotional blunting, and loss of insight.¹ These criteria are required for the diagnosis of FTD, which is supported by neuropsychological abnormalities, language dysfunction, and poor self-care. ALSci reflects frontotemporal dysfunction with deficits in attention, cognitive flexibility, and word generation, with relative sparing of visuospatial function and memory. ALSbi refers to changes in social interactions unrelated to a psychiatric condition. The domain of cognitive
and behavioral impairment in ALS is a rapidly evolving field and there is emerging consensus regarding diagnostic criteria and assessment methods. The ALS Cognitive Behavioral Screen (CBS) is specific to ALS, is validated, and has an accuracy of 100% to detect FTD, and 85% sensitivity to detect any cognitive impairment.

Estimates of cognitive impairment range from 10% to 75% in those diagnosed with ALS. A population-based sample produced an estimate of 28%. The prevalence of impairment meeting criteria for dementia ranged from 15% to 41%. Behavioral impairment (irritability and social disinhibition) was identified in 39%. Three studies documented mild cognitive decline over 6 months, while others found no change over 12 months. It is not known whether patients can progress from ALSci or ALSbi to ALS-FTD. However, 15% of patients presenting with only FTD later develop motor neuron degeneration.

A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with MND. Recent studies have documented the utility of screening instruments in a busy ALS clinic. Patients with cognitive and behavioral impairment were less compliant with management recommendations and had shorter survival.

References
Opportunity for Improvement

Cognitive impairment in ALS is best identified through neuropsychological assessment using standardized measures and normative data. The Mini-Mental State Examination is less sensitive to the cognitive impairment seen in ALS and does not examine for behavioral dysfunction. There is no consensus regarding the best screening tests for cognitive impairment in ALS. Two 1-minute word generation tests had 65% sensitivity, 90% specificity, and 88% positive predictive value in detecting possible, probable, or definite FTD by Neary criteria. A 1-minute letter fluency measure (F words) had 73% sensitivity, 88% specificity, and 79% accuracy to detect ALSci. An abbreviated neuropsychological battery demonstrated 88% accuracy.

Although there has not been a systematic study of how many clinics do screening, there is good support for the presence of a gap. Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic, but this is still not routinely practiced. A recent large study found that most patients and caregivers were not informed about the presence of cognitive or behavioral/psychological impairment. Patients commonly reported being told by their doctor about physical symptoms such as problems walking (85%) or stiffness/cramps (74%) but not psychological issues like emotional liability (46%) or cognitive change (11%). Patients and caregivers have indicated that they do want to know about whether they are so affected. These data suggest that screening is not being done widely at all.

References


<table>
<thead>
<tr>
<th>IOM Domains of Health Care Quality Addressed</th>
<th>Effective Patient-centered</th>
</tr>
</thead>
</table>

Exclusion Justification

A medical reason exclusion has been included so that clinicians can exclude patients for whom a cognitive assessment may not be appropriate (eg, patient currently diagnosed with severe cognitive impairment). A patient reason exclusion has been included for patients who decline to screened for cognitive or behavioral impairment. A system reason exclusion has been included for patients who have no insurance to cover the screening cost.

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

<table>
<thead>
<tr>
<th>Level of Measurement</th>
<th>Individual practitioner</th>
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</thead>
<tbody>
<tr>
<td>Care setting</td>
<td>Ambulatory Care</td>
</tr>
<tr>
<td>Data source</td>
<td>Electronic health record (EHR) data</td>
</tr>
<tr>
<td></td>
<td>Administrative Data/Claims (inpatient or outpatient claims)</td>
</tr>
<tr>
<td></td>
<td>Administrative Data/Claims Expanded (multiple-source)</td>
</tr>
<tr>
<td></td>
<td>Paper medical record</td>
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</tbody>
</table>

**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)**

<table>
<thead>
<tr>
<th>ICD-9 –CM Diagnosis Codes:</th>
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<tbody>
<tr>
<td>335.20 (amyotrophic lateral sclerosis)</td>
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<tr>
<td>AND</td>
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<tr>
<td>CPT E/M Service Code:</td>
</tr>
<tr>
<td>99201, 99202, 99203, 99204, 99205 (office-new patient),</td>
</tr>
<tr>
<td>99211, 99212, 99213, 99214, 99215 (office-established patient),</td>
</tr>
<tr>
<td>99241, 99242, 99243, 99244, 99245 (outpatient consult),</td>
</tr>
<tr>
<td>99304, 99305, 99306, 99307, 99308, 99309, 99310 (nursing facility),</td>
</tr>
<tr>
<td>99324, 99325, 99326, 99327, 99328, 99334, 99335, 99336, 99337 (domiciliary),</td>
</tr>
<tr>
<td>99341, 99342, 99343, 99344, 99345, 99347, 99348, 99349, 99350 (home visit)</td>
</tr>
</tbody>
</table>

**Numerator**

Patients who are screened at least once annually for cognitive impairment (eg frontotemporal dementia screening or ALS Cognitive Behavioral Screen (CBS)) and behavioral impairment (eg ALS CBS).

**Reporting Instructions:**
- For all patients meeting the denominator criteria, report the CPT Category II, 3755F, *Cognitive and behavioral impairment screening performed.*

3755 *Cognitive and behavioral impairment screening performed*

**Denominator Exclusions**

All patients with a diagnosis of amyotrophic lateral sclerosis.

- Documentation of a medical reason for not screening the patient for cognitive and behavioral impairment (eg patient near end of life)
  **Reporting Instructions:**
  - For patient with appropriate exclusion, report 3755-1P

- Documentation of a patient reason for not screening the patient for cognitive and behavioral impairment (eg patient declines to be screened for cognitive or behavioral impairment)
  **Reporting Instructions:**
  - For patient with appropriate exclusion, report: 3755-2P

• Documentation of a system reason for not screening the patient for cognitive and behavioral impairment (eg patient no insurance to cover screening cost)
  Reporting Instructions:
  o For patient with appropriate exclusion, report: 3755F-3P