Measure #5: ALS Respiratory Insufficiency Querying and Referral for Pulmonary Function Testing

**Amyotrophic Lateral Sclerosis**

### Measure Description

Percentage of patients with a diagnosis of amyotrophic lateral sclerosis who were queried about symptoms of respiratory insufficiency (awake or associated with sleep) and referred for pulmonary function testing (eg vital capacity (VC), maximum inspiratory pressure (MIP), sniff nasal pressure (SNP), or peak cough expiratory flow (PCEF)), at least every three months.

### Measure Components

<table>
<thead>
<tr>
<th><strong>Numerator Statement</strong></th>
<th>Patients who were queried about symptoms of respiratory insufficiency (awake or associated with sleep) and referred for pulmonary function testing (eg vital capacity (VC), maximum inspiratory pressure (MIP), sniff nasal pressure (SNP), or peak cough expiratory flow (PCEF)), at least every three months.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Denominator Statement</strong></td>
<td>All patients with a diagnosis of amyotrophic lateral sclerosis.</td>
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</tbody>
</table>
| **Denominator Exclusions** | • Documentation of medical reason for not querying about symptoms of respiratory and referring for pulmonary function testing or peak cough expiratory flow (eg patient with severe cognitive impairment who cannot answer any queries)  
• Documentation of patient reason for not querying about symptoms of respiratory and referring for pulmonary function testing or peak cough expiratory flow (eg patient declines to be referred for pulmonary function testing) |
| **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:  
• Symptoms or signs of respiratory insufficiency should initiate discussions with the patient and the caregivers about all treatment options such as noninvasive ventilation (NIV), tracheostomy ventilation (TV) and the terminal phase. Early discussions are needed to allow advance planning and directives. The patient should be informed about the temporary nature of NIV (which is primarily directed towards improving quality of life rather than prolonging it (as opposed to TV)). Care should adapt to the changing needs of patients and carers over the course of the disease. (GPP)¹  
• Vital capacity (VC) is the most available and practical test for the monitoring of respiratory function on a regular basis. If possible, VC should be measured both standing/sitting and lying. (GPP)¹  
• Supine FVC and MIP may be considered useful in routine respiratory monitoring, in addition to the erect FVC (Level C).²  
• Sniff nasal pressure (SNP) may be considered to detect hypercapnia and nocturnal hypoxemia (Level C).²  
• SNP may be used for monitoring of inspiratory muscle strength, particularly in some bulbar patients who cannot perform VC accurately. (GPP)¹  
• Nocturnal oximetry, available at home, is recommended in patients with symptoms of nocturnal hypoventilation. (GPP)¹  
• Nocturnal oximetry may be considered to detect hypoventilation (regardless of the forced vital capacity (FVC)). (Level C)² |

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Measure Importance

Patients should be queried about symptoms of respiratory insufficiency, screened with pulmonary function testing on a regular basis, and referred for pulmonary consultation when appropriate. Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms. The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure. Published guidelines for respiratory care were based on clinical experience, expert opinion, and observational research.

FVC is the most commonly used respiratory measurement in ALS and it was a significant predictor of survival. FVC may be an insensitive measure of respiratory muscle strength since 13/20 patients with an FVC >70% had abnormal maximal inspiratory pressure (MIP) <−60 cm.

Nocturnal desaturations <90% for 1 cumulative minute was a more sensitive indicator of nocturnal hypoventilation than either FVC or MIP. FVC correlated poorly with symptoms of nocturnal hypoventilation and desaturation. Nocturnal oximetry correlated with survival.

Supine FVC, although more difficult to perform, may be a better predictor of diaphragm weakness than erect FVC. FVC closely correlated with transdiaphragmatic pressure (Pdi), and a supine FVC <75% reliably predicted an abnormally low Pdi. Further, the difference between erect and supine FVC correlated with orthopnea.

The sniff transdiaphragmatic pressure (sniff Pdi) detected hypercapnia with a sensitivity of 90% and a specificity of 87%. Sniff nasal pressure (SNP) showed greater predictive power of survival than either FVC or MIP. When SNP was less than 30 cm, median survival was 3 months. In addition, SNP was more reliably recorded at later stages of ALS than either FVC or MIP.

The peak cough expiratory flow (PCEF) remains the most widely used measure of cough effectiveness. Patients with a mean PCEF above 337 L/min had a significantly greater chance of being alive at 18 months.

References


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**Opportunity for Improvement**

The vast majority of patients with ALS only get standard vital capacity measures to monitor pulmonary function, and in many instances even this basic measurement is not made. The use of nocturnal oximetry, polysomnography, sniff nasal pressure (SNP), maximum inspiratory pressure (MIP), peak expiratory cough flow (PCEF) and supine vital capacity (VC) should permit earlier detection and treatment of respiratory insufficiency in ALS. More studies are needed to clarify which of these measures is the best, but several studies have shown that MIP is probably the most sensitive indicator of early diaphragmatic weakness.

Symptoms of early respiratory insufficiency should be specifically asked for: dyspnea, orthopnea, excessive daytime sleepiness, insomnia, fatigue, morning headache. Patients often do not volunteer these symptoms and the clinician may detect early respiratory insufficiency by enquiring about them, which is often not done.

**References**


<table>
<thead>
<tr>
<th>IOM Domains of Health Care Quality Addressed</th>
<th>Effective Patient centered</th>
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<tbody>
<tr>
<td>Exclusion Justification</td>
<td>A medical reason exclusion for patients with severe cognitive impairment who cannot answer any queries. A patient reason exclusion has been included for patients who decline to be referred for pulmonary function testing.</td>
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<tr>
<td>Harmonization with Existing Measures</td>
<td>There are no other measures currently available that are similar to this measure or need to be harmonized with this measure.</td>
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### Measure Designation

| Measure purpose | • Quality improvement  
• Accountability |
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<tbody>
<tr>
<td>Type of measure</td>
<td>• Process</td>
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<tr>
<td>Level of Measurement</td>
<td>• Individual practitioner</td>
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<tr>
<td>Care setting</td>
<td>• Ambulatory Care</td>
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| 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) |

AND

| 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). |

**Numerator**

Patients who were who were queried about symptoms of respiratory insufficiency (awake or associated with sleep) and referred for pulmonary function testing (eg vital capacity (VC), maximum inspiratory pressure (MIP), sniff nasal pressure (SNP), or
peak cough expiratory flow (PCEF)), at least every three months.

Reporting Instructions:
- For all patients meeting denominator criteria, report the CPT Category II 1503F, *Patient queried about symptoms of respiratory insufficiency* AND 3758F, *Patient referred for pulmonary function testing or peak cough expiratory flow*.

1503F- *Patient queried about symptoms of respiratory insufficiency*
3758F- *Patient referred for pulmonary function testing or peak cough expiratory flow*

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<td>• Documentation of medical reason for not querying about symptoms of respiratory insufficiency and referring for pulmonary function testing or peak cough exploratory flow (eg patient with severe cognitive impairment who cannot answer the query)</td>
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<td>Reporting Instructions:</td>
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<td>• For patient with appropriate exclusion criteria, report: 1503F-1P AND 3758F-1P</td>
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<tr>
<td></td>
<td>• Documentation of patient reason for not querying about symptoms of respiratory insufficiency and referring for pulmonary function testing or peak cough expiratory flow (eg patient declines to be referred)</td>
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<td>Reporting Instructions:</td>
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<tr>
<td></td>
<td>• For patient with appropriate exclusion criteria, report: 1503F-2P AND 3758F-2P</td>
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