A single course of pulmonary rehabilitation in the outpatient ambulatory care setting may be considered **medically necessary** for treatment of chronic pulmonary disease for patients with moderate-to-severe disease who are experiencing disabling symptoms and significantly diminished quality of life despite optimal medical management.

A single course of pulmonary rehabilitation may be considered **medically necessary** in an outpatient ambulatory care setting as a preoperative conditioning component for those considered appropriate candidates for lung volume reduction surgery (see evidence review 7.01.71) or for lung transplantation (see evidence review 7.03.07).

Pulmonary rehabilitation programs are considered **medically necessary** following lung transplantation.

Pulmonary rehabilitation programs are considered **investigational** following other types of lung surgery, included but not limited to lung volume reduction surgery and surgical resection of lung cancer.

Multiple courses of pulmonary rehabilitation are considered **investigational**, either as maintenance therapy in patients who initially respond, or in patients who fail to respond, or whose response to an initial rehabilitation program has diminished over time.

Home-based pulmonary rehabilitation programs are considered **investigational**.

Pulmonary rehabilitation programs are considered **investigational** in all other situations.

**POLICY GUIDELINES**

A pulmonary rehabilitation outpatient program is a comprehensive program that generally includes
team assessment, patient training, psychosocial intervention, exercise training, and follow-up. The overall length of the program and the total number of visits for each component may vary from program to program.

Team assessment includes input from a physician, respiratory care practitioner, nurse, and psychologist, among others.

Patient training includes breathing retraining, bronchial hygiene, medications, and proper nutrition.

Psychosocial intervention addresses support system and dependency issues.

Exercise training includes strengthening and conditioning, and may include stair climbing, inspiratory muscle training, treadmill walking, cycle training (with or without ergometer), and supported and unsupported arm exercise training. Exercise conditioning is an essential component of pulmonary rehabilitation. Education in disease management techniques without exercise conditioning does not improve health outcomes of patients who have chronic obstructive pulmonary disease.

Follow-up to a comprehensive outpatient pulmonary rehabilitation program may include supervised home exercise conditioning.

Candidates for pulmonary rehabilitation should be medically stable and not limited by another serious or unstable medical condition. Contraindications to pulmonary rehabilitation include severe psychiatric disturbance (e.g., dementia, organic brain syndrome), and significant or unstable medical conditions (e.g., heart failure, acute cor pulmonale, substance abuse, significant liver dysfunction, metastatic cancer, disabling stroke).

Coding

While there are global HCPCS codes for pulmonary rehabilitation services such as G0237-G0239, G0302-G0305, G0424, and S9473, the component services may be reported separately using CPT codes such as 97110 and 97530 or an unlisted code such as 94799 or 97799.

BENEFIT APPLICATION

BlueCard/National Account Issues

In general, a global fee is submitted for pulmonary rehabilitation that includes all components of the program. If billing is per session, the recommendation is made to adjudicate sessions as a 1 program per lifetime benefit. Programs are usually 6 to 8 weeks in duration. Another alternative for a program not billed as a global fee is to add a dollar or visit maximum.

BACKGROUND

In 2013, the American Thoracic Society and the European Respiratory Society defined pulmonary rehabilitation (PR) as a “comprehensive intervention based on a thorough patient assessment followed by patient-tailored therapies that include, but are not limited to exercise training, education, and behavior change.” PR programs are intended to improve patient functioning and quality of life. Most research has focused on patients with chronic obstructive pulmonary disease, although there has been some interest in patients with asthma, cystic fibrosis, or bronchiectasis.

PR is also routinely offered to patients awaiting lung transplantation and lung volume reduction surgery. PR before lung surgery may stabilize or improve patients’ exercise tolerance, teach patients techniques that will help them recover after the procedure, and allow health care providers to identify individuals who might be suboptimal surgical candidates due to noncompliance, poor health, or other reasons.

Regulatory Status
RATIONAL

The evidence review was created in July 1996 and has been updated regularly with searches of the MEDLINE database. The most recent literature updated was performed through January 6, 2019. This review was informed by a 1996 TEC Assessment.²

Evidence reviews assess the clinical evidence to determine whether the use of a technology improves the net health outcome. Broadly defined, health outcomes are length of life, quality of life, and ability to function—including benefits and harms. Every clinical condition has specific outcomes that are important to patients and to managing the course of that condition. Validated outcome measures are necessary to ascertain whether a condition improves or worsens; and whether the magnitude of that change is clinically significant. The net health outcome is a balance of benefits and harms.

To assess whether the evidence is sufficient to draw conclusions about the net health outcome of a technology, 2 domains are examined: the relevance and the quality and credibility. To be relevant, studies must represent one or more intended clinical use of the technology in the intended population and compare an effective and appropriate alternative at a comparable intensity. For some conditions, the alternative will be supportive care or surveillance. The quality and credibility of the evidence depend on study design and conduct, minimizing bias and confounding that can generate incorrect findings. The randomized controlled trial (RCT) is preferred to assess efficacy; however, in some circumstances, nonrandomized studies may be adequate. RCTs are rarely large enough or long enough to capture less common adverse events and long-term effects. Other types of studies can be used for these purposes and to assess generalizability to broader clinical populations and settings of clinical practice. The following is a summary of the key literature to date.

This evidence review focuses on comprehensive, multidisciplinary programs that include an exercise component plus other modalities. Where there is a lack of evidence on multidisciplinary pulmonary rehabilitation (PR) programs, interventions that are strictly exercise will be considered. In this regard, exercise constitutes the primary intervention that improves outcomes and that, if exercise alone improves outcomes, then it would be expected that exercise plus other modalities would improve outcomes to the same degree or greater.

Chronic Obstructive Pulmonary Disease

Clinical Context and Therapy Purpose

The purpose of a single course of outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without outpatient pulmonary rehabilitation, in patients with moderate-to-severe chronic obstructive pulmonary disease. The question addressed in this evidence review is: does the use of pulmonary rehabilitation in patients with various lung conditions improve net health outcomes.

The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals with moderate-to-severe chronic obstructive pulmonary disease.

Interventions
The therapy being considered is a single course of outpatient pulmonary rehabilitation. PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

Comparators

Comparators of interest include usual care without outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy, bronchodilators, and steroid regimens.

Outcomes

The general outcomes of interest are symptoms, functional outcomes, and quality of life.

Timing

The existing literature evaluating a single course of outpatient pulmonary rehabilitation as a treatment for moderate-to-severe chronic obstructive pulmonary disease has varying lengths of follow up. While studies described below all reported at least one outcome of interest, at least 6 months duration of follow-up is desirable to fully assess outcomes.

Setting

Patients with moderate-to-severe chronic obstructive pulmonary disease are actively managed by pulmonologists and primary care providers in an outpatient clinical setting.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;

b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.

c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

d. Studies with duplicative or overlapping populations were excluded

Systematic Reviews

Numerous RCTs and several systematic reviews of RCTs have been published. Most recently, Puhan et al (2016) published a Cochrane review that evaluated PR programs for patients who had an exacerbation of chronic obstructive pulmonary disease (COPD). To be included, the rehabilitation program had to begin within 3 weeks of initiating exacerbation treatment and had to include physical exercise. Twenty trials (total N=1477 participants) met inclusion criteria. Rehabilitation was outpatient in 6 trials, inpatient in 12 trials, both inpatient and outpatient in 1 trial, and home-based in 1 trial. In a pooled analysis of 8 trials, there was a statistically significant reduction in the primary outcome (rate of hospital admissions) for PR compared with usual care (odds ratio, 0.44; 95% confidence interval [CI], 0.21 to 0.91). Several secondary outcomes also favored the PR group. In a pooled analysis of 13 trials, there was a significantly greater improvement from baseline in the 6-minute walk distance (6MWD) in the PR groups (mean difference [MD], 62.4 meters; 95% CI, 38.5 to 86.3 meters). Moreover, a pooled analysis of health-related quality of life (HRQOL) found significantly greater improvement after PR vs control (MD = -7.80; 95% CI, -12.1 to -3.5). However, in a pooled analysis of 6 trials, there was no statistically significant difference between groups in mortality rate (odds ratio, 0.68; 95% CI, 0.28 to 1.67). Trials had a mean duration of only 12 months, which may not be long enough to ascertain a difference in mortality
rates. Participants in all the studies included in this analysis could not be blinded and this may have introduced bias for outcomes to some degree. Also, some studies did not assess the outcomes of those participants who dropped out of the PR or were lost to follow-up; the study.

McCarthy et al (2015) published a Cochrane review that included RCTs assessing the effect of outpatient or inpatient PR on functional outcomes and/or disease-specific quality of life (QOL) in patients with COPD. PR programs had to be at least 4 weeks in duration and include exercise therapy with or without education and/or psychological support. Sixty-five RCTs (total N=3822 participants) met inclusion criteria. COPD severity was not specifically addressed by Cochrane reviewers, but article titles suggest a focus on patients with moderate-to-severe COPD. In pooled analyses, there was a statistically significantly greater improvement in all outcomes in PR groups than in usual care groups. Also, between-group differences on key outcomes were clinically significant. For example, on all 4 important domains of the validated Chronic Respiratory Questionnaire (CRQ)-dyspnea, fatigue, emotional function, and mastery-the effect was larger than the accepted minimal clinically important difference (MCID) of 0.5 units.

Also, the between-group difference in maximal exercise capacity exceeded the MCID of 4 watts and the between-group difference in 6MWD-a mean difference of 43.93 meters-was considered clinically significant.

Rugbjerg et al (2015) published a systematic review that identified 4 RCTs (total N=489 participants). Inspection of the trial designs for the 4 RCTs indicated that none evaluated a comprehensive PR program in patients who met criteria for mild COPD. Rather than being comprehensive PR programs, all interventions were exercise-based. One intervention included an educational component, and another used a qigong intervention, which included breathing and meditation in addition to exercise. Also, none of the RCTs enrolled a patient population with only mild COPD. Roman et al (2013) and Gottlieb et al (2011) included patients with moderate COPD. Liu et al (2012) included patients with mild-to-moderate COPD and van Wetering et al (2010) included patients with moderate-to-severe COPD. Conclusions cannot be drawn about the efficacy of PR in patients with mild COPD from this systematic review.

**Table 1. SR & MA Characteristics**

<table>
<thead>
<tr>
<th>Study</th>
<th>Dates</th>
<th>Trials</th>
<th>Participants</th>
<th>Intervention</th>
<th>N Range</th>
<th>Design</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Puhan (2016)</td>
<td>up to Mar 2010*; Mar 2010 to Oct 2015</td>
<td>20</td>
<td>PR patients (N=1477) that met inclusion criteria and had an exacerbation of COPD.</td>
<td>Inpatient and outpatient PR</td>
<td>4866 (NR)</td>
<td>RCT</td>
<td>3-18mm</td>
</tr>
<tr>
<td>McCarthy (2015)</td>
<td>up to Jul 2004; Jul 2004 to Mar 2014</td>
<td>65</td>
<td>Patients (n=3822) mean ages ranging from 31.3 to 74.1 years; resigning inpatient, out-patient, community-based or home-based rehabilitation</td>
<td>Outpatient or inpatient PR greater than or equal to 4 wks that includes exercise therapy +/- education and psychological support (range of PR excer. Prog. = 7wk to 6m)</td>
<td>3822 (694-1879)</td>
<td>RCT</td>
<td>greater than or equal to 24m</td>
</tr>
</tbody>
</table>
MP 8.03.05
Outpatient Pulmonary Rehabilitation

<table>
<thead>
<tr>
<th>Study</th>
<th>Dates</th>
<th>Trials</th>
<th>Participants</th>
<th>Intervention</th>
<th>N Range.</th>
<th>Design</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rugbjerg (2015)△</td>
<td>2000-15</td>
<td>4</td>
<td>4822 patients</td>
<td>program of greater than or equal to 4 weeks in RCTs on continuous oxygen; those w/ clinical diagnosis moderate-to-severe of COPD and best recorded forced expiratory volume after sec (FVC) &lt;0.7; exercise therapy/ Intervention (rehabilitation) vs standard care (control) N=3822 participants</td>
<td></td>
<td>RCT</td>
<td>greater than or equal to 24m</td>
</tr>
</tbody>
</table>

1 *a previous review included information from studies up to this date

Table 2. SR & MA Results

<table>
<thead>
<tr>
<th>Study</th>
<th>Rate of hospital readmission</th>
<th>Baseline 6mo walk distance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Puhan (2016)¶</td>
<td>N=810</td>
<td>N=13</td>
</tr>
<tr>
<td>N=1477</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PR compared with usual care</td>
<td>OR= (M-H, random, 95% CI) 0.44 [0.28, 1.67]</td>
<td>MD=M-H, random, 95% CI) 62.4m, 38.5 to 86.3 m</td>
</tr>
<tr>
<td>McCarthy (2015)¶</td>
<td>4 studies</td>
<td>N=313</td>
</tr>
<tr>
<td>N=3822; 4 trials</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rehab compared with usual care</td>
<td>OR= (M-H, random, 95% CI) 0.44 [0.28, 1.67]</td>
<td>Pooled results; usual care=157 patients; PR=156 patients/ mean difference in walking distance (PR vs SC)= PR greater than or equal to SC as PR (95% CI: [15.76-35.65])</td>
</tr>
<tr>
<td>Rugbjerg (2015)△</td>
<td>N=489 patients</td>
<td></td>
</tr>
</tbody>
</table>

CI: confidence interval; PR=pulmonary rehabilitation
Section Summary: Chronic Obstructive Pulmonary Disease

Multiple RCTs and meta-analyses of RCTs have, for the most part, found improved outcomes (i.e., functional ability, QOL) in patients with moderate-to-severe COPD who have had a comprehensive PR program in the outpatient setting. There is limited evidence on the efficacy of repeated and/or prolonged PR programs, and that evidence is mixed on whether these programs improve additional health outcome benefits.

Idiopathic Pulmonary Fibrosis

Clinical Context and Therapy Purpose

The purpose of a single course of outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without outpatient pulmonary rehabilitation, in patients with idiopathic pulmonary fibrosis. The question addressed in this evidence review is: does outpatient pulmonary rehabilitation improve net health outcomes in patients with various lung conditions. The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals with idiopathic pulmonary fibrosis.

Interventions

The therapy being considered is a single course of outpatient pulmonary rehabilitation. PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

Comparators

Comparators of interest include usual care without outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and, medication therapy.

Outcomes

The general outcomes of interest are symptoms, functional outcomes, and quality of life.

Timing

The existing literature evaluating a single course of outpatient pulmonary rehabilitation as a treatment for idiopathic pulmonary fibrosis has varying lengths of follow up. While studies described below all reported at least one outcome of interest, longer follow-up was necessary to fully observe outcomes. Therefore, at least 3 months of follow-up is considered necessary to demonstrate efficacy.

Setting

Patients with idiopathic pulmonary fibrosis are actively managed by pulmonologists and primary care providers in an outpatient clinical setting.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;

b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

d. Studies with duplicative or overlapping populations were excluded

Jackson et al (2014) evaluated patients with idiopathic pulmonary fibrosis who were 40 to 80 years of age and had disease onset between 3 and 48 months before screening, abnormal pulmonary function, and a 6MWD between 150 and 500 meters. In this pilot RCT, patients were assigned to a PR program consisting of twice-weekly 2-hour rehabilitation sessions over 12 weeks (n=14) or usual care (n=11). Twenty-one of the 25 patients completed the 3-month intervention study. Reviewers did not report between-group statistics. Follow-up data at 3 months postintervention were reported by Gaunaurd et al (2014). During the intervention, patients in the PR group had significantly greater self-reported physical activity, but, in the subsequent 3 months, activity levels in the 2 groups were similar. For example, at 6 months, pulmonary function measures (eg, total lung capacity, forced vital capacity, spirometry diffusion capacity) did not change significantly within either group. 6MWD data were not reported.

**Section Summary: Idiopathic Pulmonary Fibrosis**

One small RCT has evaluated a comprehensive PR program in patients with idiopathic pulmonary fibrosis; at 3 months postintervention, outcomes did not differ between groups that did and did not receive PR.

**Bronchiectasis**

**Clinical Context and Therapy Purpose**

The purpose of a single course of outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without outpatient pulmonary rehabilitation, in patients with bronchiectasis.

The question addressed in this evidence review is: what the safety and efficacy of pulmonary rehabilitation in patients with various lung conditions.

The following PICOTS were used to select literature to inform this review.

**Patients**

The relevant population of interest are individuals with bronchiectasis.

**Interventions**

The therapy being considered is a single course of outpatient pulmonary rehabilitation.

PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

**Comparators**

Comparators of interest include usual care without outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and, medication therapy.

**Outcomes**

The general outcomes of interest are symptoms, functional outcomes, and quality of life.

**Timing**
The existing literature evaluating a single course of outpatient pulmonary rehabilitation as a treatment for bronchiectasis has varying lengths of follow up. While studies described below all reported at least one outcome of interest, 3-6 months duration of follow-up is desirable to fully assess outcomes.

**Setting**

Patients with bronchiectasis are actively managed by pulmonologists and primary care providers in an outpatient clinical setting.

**Study Selection Criteria**

Methodologically credible studies were selected using the following principles:

a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;

b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.

c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

d. Studies with duplicative or overlapping populations were excluded

Lee et al (2017) published a systematic review of RCTs on PR in patients with non-cystic fibrosis bronchiectasis. Reviewers identified 4 RCTs. They selected studies of exercise-only interventions as well as exercise combined with education and/or another intervention. The control intervention had to be something other than exercise-based. A pooled analysis of 3 RCTs immediately after an 8-week intervention found significantly greater incremental shuttle walk distance in the intervention compared with the control group (MD=66.6; 95% CI, 51.8 to 81.7). A pooled analysis of 2 trials found significantly greater improvement in the St. George's Respiratory Questionnaire score postintervention (MD = -4.65; 95% CI, -6.70 to -2.60). There was no significant difference postintervention on the Leicester Cough Questionnaire (total) scores. Reviewers did not conduct meta-analyses beyond the immediate postintervention period.

**Section Summary: Bronchiectasis**

A systematic review of RCTs on PR for patients with bronchiectasis found that some, but not all, outcomes improved more with PR than with a non-exercise control condition immediately postintervention. Limited observational data would suggest that outcomes in patients with other respiratory conditions may benefit, but likely not as much as COPD patients.

**PR Programs Before Lung Surgery**

**Clinical Context and Therapy Purpose**

The purpose of a single course of preoperative outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without outpatient pulmonary rehabilitation, in patients with scheduled lung surgery for volume reduction, transplantation, or resection.

The question addressed in this evidence review is: does the use of pulmonary rehabilitation improve net health outcomes in patients undergoing lung surgery for various conditions.

The following PICOTS were used to select literature to inform this review.

**Patients**
The relevant population of interest are individuals with scheduled lung surgery for volume reduction, transplantation, or resection.

Interventions
The therapy being considered is a single course of preoperative outpatient pulmonary rehabilitation. PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

Comparators
Comparators of interest include usual care without outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and medication therapy.

Outcomes
The general outcomes of interest are symptoms, functional outcomes, and quality of life.

Timing
The existing literature evaluating a single course of preoperative outpatient pulmonary rehabilitation as a treatment for scheduled lung surgery for volume reduction, transplantation, or resection has varying lengths of follow up. While studies described below all reported at least one outcome of interest, 3-6 months duration follow-up are desirable to assess outcomes.

Setting
Patients with scheduled lung surgery for volume reduction, transplantation, or resection are actively managed by pulmonologists, general surgeons and thoracic surgeons.

Study Selection Criteria
Methodologically credible studies were selected using the following principles:

a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.
d. Studies with duplicative or overlapping populations were excluded

Lung Volume Reduction Surgery
PR prior to lung volume reduction surgery (LVRS) represents a distinct subset of patients with COPD, and the National Emphysema Treatment Trial requires all candidates to undergo a vigorous course of PR. The final National Emphysema Treatment Trial results supported the treatment effectiveness in a subset of patients with COPD.

Lung Transplantation
A systematic review of the literature on PR for lung transplant candidates was published by Hoffman et al (2017). Interventions had to include exercise training but did not have to be part of a comprehensive PR program and could have taken place in the inpatient or outpatient setting. Reviewers identified 6 studies—2 RCTs and 4 case series. Both RCTs evaluated the impact of exercise (not comprehensive PR) on outcomes; additionally, one was conducted in the inpatient setting, and the
included only 9 patients. Conclusions on the impact of a comprehensive PR program before lung transplantation on health outcomes cannot be drawn from this systematic review.

**Lung Cancer Resection**

Several small RCTs have evaluated preoperative PR for patients undergoing lung cancer resection. Morano et al (2013) conducted a single-blind study in Brazil.\(^\text{15}\) Patients with non-small-cell lung cancer eligible for lung resection were randomized to 4 weeks of an exercise-only PR program (5 sessions per week) or chest physical therapy; there were 12 patients in each group. All patients in the PR group and 9 of 12 in the chest physical therapy group subsequently underwent surgery (the other 3 patients had inoperable disease). Several short-term postoperative outcomes were assessed. Patients in the PR group spent significantly fewer days in the hospital (mean, 7.8 days) than patients in the chest physical therapy group (mean, 12.2 days; \(p=0.04\)). Also, patients in the PR group spent fewer days with chest tubes (mean, 4.5 days) than the physical therapy group (mean, 7.4 days; \(p=0.03\)). The trial did not assess longer term functional outcomes after surgery.

Benzo et al (2011) conducted 2 small exploratory RCTs evaluating PR before lung cancer resection.\(^\text{16}\) Eligibility criteria included having moderate-to-severe COPD and being scheduled for lung cancer resection either by open thoracotomy or by video-assisted thoracoscopy. The first trial had poor recruitment, enrolling only 9 patients. The second study enrolled 19 patients into a 10-session, preoperative PR program (n=10) or usual care (n=9). Mean number of days in the hospital was 6.3 in the PR group and 11.0 in the control group (\(p=0.058\)). Three (33%) patients in the PR group and 5 (63%) patients in the control group experienced postoperative pulmonary complications (\(p=0.23\)). The trial sample size was likely too small to detect statistically or clinically significant differences between groups. Trialists recommended conducting a larger multicenter randomized trial in this population.

Bradley et al (2013), in a nonrandomized comparative study, evaluated an outpatient-based PR intervention in 58 lung cancer patients who were candidates for surgery.\(^\text{17}\) This U.K.-based study also evaluated a comparison group of 305 patients, also surgical candidates, who received usual care. Patients in the 2 groups were matched by age, lung function, comorbidities, and type of surgery. In a within-group analysis, there was a statistically significant 20-meter improvement in 6MWD in the intervention group before and after participation in a 4-session presurgical PR program. In between-group analyses, there were no statistically significant differences between the intervention and comparisons groups in clinical outcomes such as postoperative pulmonary complications, readmissions, and mortality after surgery.

**Table 3. Summary of Key RCT Characteristics**

<table>
<thead>
<tr>
<th>Study</th>
<th>Countries</th>
<th>Sites</th>
<th>Dates</th>
<th>Participants</th>
<th>Interventions</th>
<th>Comparator</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morano (2013)(^\text{15})</td>
<td>Brazil</td>
<td>1</td>
<td>Mar 2008 to Mar 2011</td>
<td>Patients undergoing lung-cancer resection (N=24) and who have non-small cell lung cancer resection by open thoracotomy (or video-assisted); and prev pulmonary disease, interstitial lung disease, or obstructive airway disease, with impaired respiratory function by spirometry.</td>
<td>PR: Strength/endurance training+ education; 5 sessions/wk for 4 wks (20 sessions) (n=12)</td>
<td>CPT breathing exercises +education; 5 sessions/wk for 4 wks (20 sessions) (n=12)</td>
</tr>
</tbody>
</table>
Table 4. Summary of Key RCT Results

<table>
<thead>
<tr>
<th>Study</th>
<th>ICU stay mean SD at 4 weeks</th>
<th>Postoperative hospitalizations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morano (2013)</td>
<td>N=31 patients at t=0; 24 in analysis; 21 in final analysis</td>
<td>N=31 patients at t=0; 24 in analysis; 21 in final analysis</td>
</tr>
<tr>
<td>PR (exercise) n=12</td>
<td>0.8 +/- 4.8</td>
<td>2 (2-3) P=.20 (not significant)</td>
</tr>
<tr>
<td>CPT (control) n=9</td>
<td>12.2 +/- 3.6</td>
<td>2 (2-4.5) P=.20 (not significant)</td>
</tr>
<tr>
<td>Benzo (2011)</td>
<td>NR</td>
<td>NR</td>
</tr>
</tbody>
</table>

CI: confidence interval; HR: hazard ratio; NNT: number needed to treat; OR: odds ratio; RCT: randomized controlled trial; RR: relative risk.

Table 5. Relevance Gaps

<table>
<thead>
<tr>
<th>Study</th>
<th>Population a</th>
<th>Intervention b</th>
<th>Comparator c</th>
<th>Outcomes d</th>
<th>Follow-Up e</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morano (2013)</td>
<td></td>
<td></td>
<td></td>
<td>3. No CONSORT reporting of harms was addressed</td>
<td>1. Short duration of follow-up (4-weeks)</td>
</tr>
<tr>
<td>Benzo (2011)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The evidence gaps stated in this table are those notable in the current review; this is not a comprehensive gaps assessment.
a Population key: 1. Intended use population unclear; 2. Clinical context is unclear; 3. Study population is unclear; 4. Study population not representative of intended use.
b Intervention key: 1. Not clearly defined; 2. Version used unclear; 3. Delivery not similar intensity as comparator; 4. Not the intervention of interest.
c Comparator key: 1. Not clearly defined; 2. Not standard or optimal; 3. Delivery not similar intensity as intervention; 4. Not delivered effectively.
d Outcomes key: 1. Key health outcomes not addressed; 2. Physiologic measures, not validated surrogates; 3. No CONSORT reporting of harms; 4. Not establish and validated measurements; 5. Clinical significant difference not prespecified; 6. Clinical significant difference not supported.
e Follow-Up key: 1. Not sufficient duration for benefit; 2. Not sufficient duration for harms.

Table 6. Study Design and Conduct Gaps

<table>
<thead>
<tr>
<th>Study</th>
<th>Allocation a</th>
<th>Blinding b</th>
<th>Selective Reporting c</th>
<th>Follow-Up d</th>
<th>Power e</th>
<th>Statistical f</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morano (2013)</td>
<td>4. Inadequate control for selection bias: the participants were not evenly randomized</td>
<td></td>
<td></td>
<td>1. High loss to follow-up or missing data</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Benzo (2011)</td>
<td></td>
<td></td>
<td></td>
<td>1. Power is not reported</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

COPD: chronic obstructive pulmonary disease; CPT: chest physical therapy; IMT: inspiratory muscle training; MEP: maximal expiratory pressure; MIP: maximal inspiratory pressure; PPC: postoperative pulmonary complication; PR: pulmonary rehabilitation; 6MWT: 6-minute walk test

The evidence gaps stated in this table are those notable in the current review; this is not a comprehensive gaps assessment.
d Follow-Up key: 1. High loss to follow-up or missing data; 2. Inadequate handling of missing data; 3. High number of crossovers; 4. Inadequate handling of crossovers; 5. Inappropriate exclusions; 6. Not intent to treat analysis (per protocol for noninferiority trials).
e Power key: 1. Power calculations not reported; 2. Power not calculated for primary outcome; 3. Power not based on clinically important difference.
f Statistical key: 1. Intervention is not appropriate for outcome type: (a) continuous; (b) binary; (c) time to event; 2. Intervention is not appropriate for multiple observations per patient; 3. Confidence intervals and/or p values not reported; 4. Comparative treatment effects not calculated.

Section Summary: Preoperative Pulmonary Rehabilitation Programs

The National Emphysema Treatment Trial has recommended administering PR before LVRS, which is considered the standard of care before LVRS and lung transplantation. However, there is a lack of large RCTs comparing PR with no PR for preoperative candidates undergoing LVRS, lung transplantation, or lung cancer resection. The available studies evaluated exercise programs and comprehensive PR. Also, the few small RCTs, and observational studies have reported on short-term outcomes and have found inconsistent evidence of benefit even on these outcomes.

Original Policy Date: July 1996
Postoperative Pulmonary Rehabilitation Programs

Lung Volume Reduction Surgery

Clinical Context and Therapy Purpose

The purpose of a single course of outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without outpatient pulmonary rehabilitation, in patients who have had lung volume reduction surgery.

The question addressed in this evidence review is: does the use of postoperative pulmonary rehabilitation improve net health outcomes in patients who have undergone lung volume reduction surgery.

The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals who have had lung volume reduction surgery.

Interventions

The therapy being considered is a single course of outpatient pulmonary rehabilitation.

PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

Comparators

Comparators of interest include usual care without outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and medication therapy.

Outcomes

The general outcomes of interest are symptoms, functional outcomes, and quality of life.

Timing

The existing literature evaluating a single course of outpatient pulmonary rehabilitation as a treatment for individuals who have had lung volume reduction surgery has varying lengths of follow up. While studies described below all reported at least one outcome of interest, 3-6 months duration of follow-up is desirable to assess outcomes.

Setting

Patients who have had lung volume reduction surgery are actively managed by pulmonologists, general surgeons and thoracic surgeons in an outpatient clinical setting.

Study Selection Criteria

Methodologically credible studies were selected using the following principles:

a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;

b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.

c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

d. Studies with duplicative or overlapping populations were excluded.
No RCTs evaluating comprehensive PR programs after LVRS were identified. Bering et al (2009) reported on a case series involving 49 patients with severe emphysema who participated in a PR program after LVRS. Patients underwent LVRS at a single center and had not received PR at that institution presurgery. After hospital discharge, patients underwent an outpatient comprehensive PR program for 4 hours a day, 5 days a week for 2 weeks. The program included a multidisciplinary team including with a variety of components, including dietary, physical therapy, physical exercise, psychosocial, occupational therapy, and respiratory therapy. The primary outcome was HRQOL measured by the 36-Item Short-Form Health Survey. Compared with pre-LVRS scores, significantly better scores were achieved on the Physical Component Summary and Mental Component Summary at both time 2 (3-6 months post-LVRS) and time 3 (12-18 months LVRS). Study limitations included no comparison with patients who had LVRS and no PR, and the difficulty disentangling the impact of LVRS from that of PR on outcomes. Moreover, patients had not received PR before LVRS, so the treatment effects of pre- vs postsurgery LVRS could not be determined.

Subsection Summary: PR Programs After LVRS

No comparative studies have evaluated PR programs after LVRS. One case series have evaluated a comprehensive PR program after LVRS in 49 patients who had not received preoperative PR. HRQOL was higher at 3 to 6 months and 12 to 18 months post-surgery. The study did not provide data on patients who underwent LVRS and did not have postoperative PR or on patients who had preoperative PR.

Lung Transplantation

Clinical Context and Therapy Purpose

The purpose of a single course of outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without outpatient pulmonary rehabilitation, in patients who have had lung transplantation. The question addressed in this evidence review is: does the use of pulmonary rehabilitation improve net health outcomes in patients who have undergone lung transplantation. The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals with who have had lung transplantation.

Interventions

The therapy being considered is a single course of outpatient pulmonary rehabilitation. PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

Comparators

Comparators of interest include usual care without outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and medical therapy.

Outcomes

The general outcomes of interest are symptoms, functional outcomes, and quality of life.

Timing

The existing literature evaluating a single course of outpatient pulmonary rehabilitation as a treatment for individuals who have had lung transplantation has varying lengths of follow up. While studies
described below all reported at least one outcome of interest, 3-6 months duration of follow-up is desirable to assess outcomes.

**Setting**
Patients who have had lung transplantation are actively managed by pulmonologists and primary care providers in an outpatient clinical setting.

**Study Selection Criteria**
Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
- In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.
- To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.
- Studies with duplicative or overlapping populations were excluded.

Exercise training after lung transplantation is reported in the literature but not necessarily provided in comprehensive PR programs. Wickerson et al (2010) published a systematic review of the available literature in which the researcher had evaluated any exercise intervention in conjunction with lung transplantation. Seven studies (a cohort made of RCTs, controlled trials, and prospective cohorts) met the inclusion criteria, including two randomized controlled trials targeting lumbar bone mineral density. Also included in the review were uncontrolled studies that reported improvement in functional status as a byproduct of an exercise-program intervention.19

**Randomized Controlled Trials**
Langer et al (2012) conducted an RCT in the U.K. that examined activity-related outcomes in lung transplant recipients after exercise training.20 The trial included 40 patients who underwent single- or double-lung transplantation and had an uncomplicated postoperative period. Following hospital discharge, patients were randomized to a supervised exercise program 3 times a week for 3 months (n=21) or to usual care with instructions to exercise (n=19). Patients in both groups had 6 individual counseling sessions in the 6 months postdischarge. Six patients dropped out of the trial, three in each group. The primary outcome was daily walking time, assessed by activity monitors. At the end of the 3-month intervention and 1-year postdischarge, mean walking times were significantly longer in the intervention group. At 1 year, the exercise group walked a mean of 85 minutes per day while the control group walked a mean of 54 minutes per day (p=0.006). Other outcomes related to daily physical activity were reported as secondary outcomes and some, but not all, significantly favored the intervention group. Mean 6MWD at 1 year was 86% of predicted in the exercise group and 74% of predicted in the control group (p=0.002). The trial had a relatively small sample size and may have been underpowered to detect clinically meaningful differences between groups on secondary outcomes.

Fuller et al (2017) published an RCT reporting on the impact of short (7-week) vs long (14-week) rehabilitation programs for patients who underwent lung transplantation.21 The primary outcome was change in the 6-minute walking test (6MWT). Secondary outcomes included the strength of the quadriceps and hamstring muscles (as measured by an isokinetic dynamometer), and QOL (as measured by the 36-Item Short-Form Health Survey). In both the 7- and 14-week rehabilitation groups, participants increased their 6MWT (mean improvement in 7-week group, 202 meters vs 14-week group, 149 meters). At 6 months after transplantation, the mean difference between groups was 59.3 meters,
favoring the 7-week group (95% CI, 12.9 to 131.6 meters). The increases in strength in quadriceps and hamstring muscles in both groups did not differ statistically. The 36-Item Short-Form Health Survey summary scores of the domains of physical health and mental health both increased over time with no significant difference between groups at any time point.

**Table 7. Summary of Key RCT Characteristics**

<table>
<thead>
<tr>
<th>Study Trial</th>
<th>Countries</th>
<th>Sites</th>
<th>Participants</th>
<th>Interventions</th>
<th>X</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fuller (2017)</td>
<td>US</td>
<td>1</td>
<td>Post-lung transplantation (LTX) patients aged greater than or equal to 18 years (N=66; 33 women; mean age, 51+/-13y) who had undergone either single LTX or bilateral LTX</td>
<td>longer-duration (14wk) rehabilitation program after LTX</td>
<td>Active Comparator</td>
</tr>
<tr>
<td>Langer (2012)</td>
<td>UK</td>
<td>1</td>
<td>Patients who received a single or double lung transplantation and had no postoperative complications N=40</td>
<td>Exercise program (3x/wk for 3m) N=21</td>
<td>Usual Care with added &quot;instruction to exercise&quot; N=19</td>
</tr>
</tbody>
</table>

LTX: lung transplantation; RCT: randomized controlled trial.
1 Number randomized; intervention; mode of delivery; dose (frequency/duration).
2 Key eligibility criteria

**Table 8. Summary of Key RCT Results**

<table>
<thead>
<tr>
<th>Study</th>
<th>Daily walking time. time postop.</th>
<th>Mean improvement in 6 MWD from time 0 time Z</th>
<th>X6MWD mean diff between groups at 6 mos</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fuller (2017)</td>
<td>N=34 NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>N=40</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Exercise program (baseline/final)=21/18</td>
<td>Mean=85m/day (1y) NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Usual Care baseline/final)=19/16)</td>
<td>Mean=34m/day (1y) NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>P value</td>
<td>P=0.0006 NR</td>
<td>NR</td>
<td>NR</td>
</tr>
<tr>
<td>Langer (2012)</td>
<td>-</td>
<td></td>
<td></td>
</tr>
<tr>
<td>N=66</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>longer-duration (14wk) rehabilitation program after LTX</td>
<td>NR +149 meters</td>
<td>59.3meters(95%CI, 12.9 to 131.6 meters)</td>
<td></td>
</tr>
<tr>
<td>Shorter (7wk) rehabilitation program after LTX</td>
<td>NR +202 meters</td>
<td>59.3meters(95%CI, 12.9 to 131.6 meters)</td>
<td></td>
</tr>
</tbody>
</table>

CI: confidence interval; HR: hazard ratio; LTX: lung transplantation; NNT: number needed to treat; OR: odds ratio; RCT: randomized controlled trial; RR: relative risk.
1 Include number analyzed, effect in each group, and measure of effect (absolute or relative) with CI,
2. Describe the range of sample sizes, effects, and other notable features in text.

### Table 9. Relevance Gaps

<table>
<thead>
<tr>
<th>Study</th>
<th>Population</th>
<th>Intervention</th>
<th>Comparator</th>
<th>Outcomes</th>
<th>Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fuller (2017)</td>
<td>1. Selection criteria not clear</td>
<td>2. Fitness activity monitor not validated as comparator for this clinical scenario.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Langer (2012)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The evidence gaps stated in this table are those notable in the current review; this is not a comprehensive gaps assessment.

a. Population key: 1. Intended use population unclear; 2. Clinical context is unclear; 3. Study population is unclear; 4. Study population not representative of intended use.
b. Intervention key: 1. Not clearly defined; 2. Version used unclear; 3. Delivery not similar intensity as comparator; 4. Not the intervention of interest.
c. Comparator key: 1. Not clearly defined; 2. Not standard or optimal; 3. Delivery not similar intensity as intervention; 4. Not delivered effectively.

### Table 10. Study Design and Conduct Gaps

<table>
<thead>
<tr>
<th>Study</th>
<th>Allocation</th>
<th>Blinding</th>
<th>Selective Reporting</th>
<th>Follow-Up</th>
<th>Power</th>
<th>Statistical</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fuller (2017)</td>
<td>1. Patients not blinded. Blinding not feasible. Outcome assessment not blinded.</td>
<td></td>
<td></td>
<td>1,2. Power is affected by small sample size, underpowered to detect meaningful differences</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The evidence gaps stated in this table are those notable in the current review; this is not a comprehensive gaps assessment.

d. Follow-Up key: 1. High loss to follow-up or missing data; 2. Inadequate handling of missing data; 3. High number of crossovers; 4. Inadequate handling of crossovers; 5. Inappropriate exclusions; 6. Not intent to treat analysis (per protocol for noninferiority trials).
e. Power key: 1. Power calculations not reported; 2. Power not calculated for primary outcome; 3. Power
Outpatient Pulmonary Rehabilitation

not based on clinically important difference.

Case Series

Munro et al (2009) published a case series that evaluated a comprehensive PR program after lung surgery. The 7-week program, which started 1 month postsurgery, consisted of 1 hour of supervised exercise 3 times a week and a weekly group education session facilitated by a multidisciplinary team (eg, nurse, dietician, occupational therapist, social worker). Compared with baseline, on program completion, both forced expiratory volume in 1 second and forced vital capacity had improved significantly (p<0.001). For example, mean forced expiratory volume in 1 second was 71% at 1 month, postsurgery and 81% at 3 months. Similarly, 6MWD improved significantly: mean distance was 451 meters at 1 month and 543 meters at 3 months posttransplant. The study lacked a control group. Hence, the degree of improvement that would have occurred without participation in a PR program is unknown.

Subsection Summary: PR Programs After Lung Transplantation

A systematic review of exercise training after lung transplantation (not necessarily provided in a comprehensive PR program) identified 7 controlled and uncontrolled studies but did not pool study findings. Neither RCT identified reported functional outcomes, but the uncontrolled studies did report improvements in functional outcomes. An RCT, published after the systematic review, found that patients who had a post-surgical exercise intervention walked more 1-year post-discharge and had a significantly greater 6MWD. The most recent RCT (2017) did not identify a difference in outcomes with longer duration of PR. Findings on other outcomes were mixed. Case series data also support improvement in the 6MWD after postoperative PR.

Lung Cancer Resection

Clinical Context and Therapy Purpose

The purpose of a single course of outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without outpatient pulmonary rehabilitation, in patients who have had lung cancer resection.

The question addressed in this evidence review is: does the use of outpatient pulmonary rehabilitation improve net health outcomes in patients who have had lung cancer resection surgery.

The following PICOTS were used to select literature to inform this review.

Patients

The relevant population of interest are individuals who have had lung cancer resection.

Interventions

The therapy being considered is a single course of outpatient pulmonary rehabilitation.

PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

Comparators

Comparators of interest include usual care without outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and medical therapy.

Outcomes
The general outcomes of interest are symptoms, functional outcomes, and quality of life.

**Timing**

The existing literature evaluating a single course of outpatient pulmonary rehabilitation as a treatment for individuals who have had lung cancer resection has varying lengths of follow up. While studies described below all reported at least one outcome of interest, 3-6 months duration of follow-up is desirable to assess outcomes.

**Setting**

Patients who have had lung cancer resection are actively managed by pulmonologists and primary care providers in an outpatient clinical setting.

**Study Selection Criteria**

Methodologically credible studies were selected using the following principles:

a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;

b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.

c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

d. Studies with duplicative or overlapping populations were excluded.

Stigt et al (2013) published an RCT evaluating a multicomponent postsurgery PR program in patients with resectable lung cancer.23 The trial was conducted in the Netherlands. Before thoracotomy, 57 patients were randomized to PR or usual care. The 12-week PR program started 4 weeks after surgery and consisted of exercise training, pain management, and visits with a medical social worker. The trial was terminated early because the institution started offering video-assisted thoracoscopic surgery, at which point few patients chose thoracotomy. Data on 49 patients (PR=23, usual care=26) were analyzed. The primary end point was QOL, as measured by the difference between groups in change in the total St. George’s Respiratory Questionnaire score from baseline to 12 months. This difference was 2.71 points, which was not statistically significant (p=0.69). However, 6MWD (a secondary outcome) improved significantly in the PR group than in the usual care group at 3 months. The between-group difference in 6MWD was 94 meters (p=0.024). A limitation of this analysis is that only 8 of 23 patients in the PR performed a 6MWT at 3 months; the other 15 patients had dropped out or did not take the test. Eleven of 25 patients in the usual care group performed the 6MWT.

An exercise-only intervention after lung cancer surgery (not comprehensive PR) was evaluated in an RCT published by Edvardsen et al (2015).24 This single-blind trial was conducted in Norway and included lung cancer patients at 4 to 6 weeks postsurgery. Sixty-one patients were randomized to an exercise program 3 times a week for 20 weeks or to usual care. The exercise intervention took place at local fitness centers and was supervised by trained personal trainers and physical therapists. The significantly greater improvement was reported for the primary outcome (change in peak oxygen uptake from baseline to the end of the intervention) in the intervention group than in the control group (between-group difference, 0.26 L/min; p=0.005.) Findings on secondary outcomes were mixed. For example, the between-group difference enforced expiratory volume in 1 second was 0.6% of predicted (95% CI, -4.2% to 5.4%; p=0.738) and the difference in stair run was 4.3 steps (95% CI, 1.6 to 7.1 steps; p=0.002). This trial did not report other functional outcomes (eg, 6MWD).

Subsection Summary: Lung Cancer Resection
A single RCT has evaluated a comprehensive PR program in patients who underwent thoracotomy for lung cancer. The trial was terminated early, had a high dropout rate, and reported mixed findings. An exercise-only intervention in patients who had lung cancer surgery had mixed findings and did not evaluate functional outcomes. Current evidence is not sufficiently robust to draw conclusions on the utility of PR programs to those who have had lung resection.

**Repeat and Maintenance Pulmonary Rehabilitation Programs**

**Clinical Context and Therapy Purpose**

The purpose of repeat or maintenance outpatient pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as usual care without repeat or maintenance outpatient pulmonary rehabilitation, in patients who have had an initial course of pulmonary rehabilitation.

The question addressed in this evidence review is: does the use of repeat or maintenance pulmonary rehabilitation improve net health outcomes in patients who have various lung conditions.

The following PICOTS were used to select literature to inform this review.

**Patients**

The relevant population of interest are individuals with who have had an initial course of pulmonary rehabilitation.

**Interventions**

The therapy being considered is repeat or maintenance outpatient pulmonary rehabilitation. PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

**Comparators**

Comparators of interest include usual care without repeat or maintenance outpatient pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and medical therapy.

**Outcomes**

The general outcomes of interest are symptoms, functional outcomes, and quality of life.

**Timing**

The existing literature evaluating repeat or maintenance outpatient pulmonary rehabilitation as a treatment for individuals who have had an initial course of pulmonary rehabilitation has varying lengths of follow up. While studies described below all reported at least one outcome of interest, 13-6 months duration follow-up is desirable to assess outcomes.

**Setting**

Patients who have had an initial course of pulmonary rehabilitation are actively managed by pulmonologists and primary care providers in an outpatient clinical setting.

**Study Selection Criteria**

Methodologically credible studies were selected using the following principles:

- To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;
b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.

c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

d. Studies with duplicative or overlapping populations were excluded.

Both repeat and maintenance PR programs provide additional rehabilitation services after initial participation in a PR program. Program categories are not strictly defined, but repeat programs are generally those that include patients who failed to respond to an initial program or whose response to an initial rehabilitation program diminished over time. In contrast, maintenance programs tend to be those designed to extend the effects of the initial PR program, and they are open to all patients who successfully completed an initial program.

### Repeat Pulmonary Rehabilitation Program

Carr et al (2009) prospectively identified Canadian patients with moderate-to-severe COPD who experienced an acute exacerbation within 12 months of participating in a PR program. Initially, patients completed a 6-week inpatient program or a 12-week outpatient program. The repeat PR program lasted 3 weeks and consisted of exercise and education; patients could choose inpatient or outpatient versions. Over 6 months, 41 patients developed an exacerbation and 12 did not. Seven patients withdrew from the trial, and the remaining 34 were randomized to a repeat PR program within 1 month of the exacerbation (n=17) or to no repeat PR program (n=17). One patient in the intervention group dropped out; of the remaining 33 patients, 25 (76%) experienced an exacerbation of moderate severity; the remaining 8 had severe exacerbations. Nine (56%) of 16 patients in the intervention group chose an inpatient program, and 7 chose an outpatient program. Patients were assessed before the repeat PR program, immediately after (3 weeks later), and again 12 weeks after the beginning of the exacerbation (5 weeks after completing the repeat rehabilitation program). The primary outcome was change in HRQOL, as measured on the 4 domains of the CRQ score. There was no statistically significant difference between groups in mean change in CRQ scores. Among patients in the intervention group, the magnitude of improvement in the domains of dyspnea (0.7 points) and fatigue (0.5 points) met or exceeded the MCID. In the control group, the magnitude of change in all domains did not meet the MCID. Change in the 6MWD (a secondary outcome) did not differ significantly between groups at either follow-up. Outcomes were not reported separately for the inpatient or outpatient programs (this evidence review addresses outpatient programs). Trialists recommended that future evaluations of repeat PR programs include patients with more serious exacerbations, last longer than 3 weeks, and start as close in time as possible to the exacerbation. Conclusions about repeat PR programs cannot be drawn from 1 study with 33 subjects.

### Maintenance Pulmonary Rehabilitation Program

In 2012, an Ontario Health Technology Assessment evaluated PR for patients with COPD. Reviewers identified 3 RCTs (total N=284 participants) assessing maintenance PR programs for individuals with COPD who had successfully completed an initial PR program. The trials excluded patients who had experienced a recent acute exacerbation of COPD. All maintenance programs consisted of supervised exercise sessions; program duration was 3 months in 1 program and 12 months in the other two. One program also included an unsupervised exercise component, and another included educational sessions. Reviewers judged study quality as generally poor, due to methodologic limitations (eg, inadequate information on randomization, allocation concealment, blinding, and lack of clarity around the use of an intention-to-treat analysis). In a pooled analysis of data from 2 trials (n=168 patients), there was a significantly greater improvement in 6MWD in patients who participated in the maintenance program...
than in those in a control group (MD=22.9 meters; 95% CI, 5.2 to 40.7 meters). The confidence interval was wide, indicating lack of precision in the pooled estimate. Also, reviewers considered the MCID to be 25 to 35 meters walked, and meta-analysis of trial findings did not meet this threshold of difference between groups.

Several RCTs were published after the Ontario assessment. Guell et al (2017) published findings of a 3-year trial of patients with severe COPD. A total of 143 patients attended an initial 8-week outpatient PR program, and 138 were then randomized to a 3-year maintenance program (n=68) or a control group (n=70). The maintenance intervention consisted of home-based exercises, calls from a physical therapist every 2 weeks, and supervised training sessions every 2 weeks. The control group was advised to exercise at home without supervision. Some outcomes but not others favored the intervention group at 2 years, but outcomes did not differ significantly between groups at 3 years. For example, compared with baseline, at 2 years the 6MWD increased by 2 meters in the intervention group and decreased by 32 meters in the control group (p=0.046). At 3 years, compared with baseline, the 6MWD decreased by 4 meters in the intervention group and decreased by 33 meters in the control group (p=0.119). The CRQ dyspnea score, at 2 years compared with baseline, decreased by 0.4 points in the intervention group and by 0.3 points in the control group (p=0.617); findings were similar at 3 years. The trial also had a high dropout rate.

Wilson et al (2015) published a single-blind RCT comparing maintenance PR to standard care without maintenance PR in patients who had COPD and had completed at least 60% of an initial PR program. One hundred forty-eight patients were randomized; 110 (74%) completed the trial and were included in the analysis. The maintenance program consisted of a 2-hour session every 3 months for 1 year. The session included an hour of education and an hour of supervised individualized exercise training. The primary efficacy outcome was change from baseline (post-PR) in the CRQ dyspnea domain. Among trial completers, mean CRQ dyspnea score changed from 2.6 to 3.2 among patients receiving maintenance PR and from 2.5 to 3.3 among controls. The difference between groups was not statistically significant. Secondary outcomes, including other CRQ domains, scores on the endurance shuttle walk test, and a number of exacerbations or hospitalizations, also did not differ significantly between groups.

**Section Summary: Repeat and Maintenance PR Programs**

A limited number of RCTs are available to evaluate repeat or maintenance rehabilitation programs. Due to the paucity of RCTs, methodologic limitations of available trials, and lack of clinically significant findings, the evidence to determine the effect of repeat and maintenance PR programs on health outcomes in patients with COPD is insufficient.

**Home-Based Pulmonary Rehabilitation Programs**

**Clinical Context and Therapy Purpose**

The purpose of a single course of home-based pulmonary rehabilitation is to provide a treatment option that is an alternative to or an improvement on existing therapies, such as a single course of ambulatory care™ based pulmonary rehabilitation, in patients with an indication for outpatient pulmonary rehabilitation.

The question addressed in this evidence review is: does the use of home-based pulmonary rehabilitation programs improve net health outcomes in patients with various lung conditions. The following PICOTS were used to select literature to inform this review.

**Patients**
The relevant population of interest are individuals with an indication for outpatient pulmonary rehabilitation.

**Interventions**

The therapy being considered is a single course of home-based pulmonary rehabilitation. PR programs include a patient assessment followed by therapeutic interventions including exercise training, education, and behavior change.

**Comparators**

Comparators of interest include a single course of ambulatory care™ based pulmonary rehabilitation. Treatment includes physical exercise, diaphragmatic breathing, oxygen therapy and, medical therapy.

**Outcomes**

The general outcomes of interest are symptoms, functional outcomes, and quality of life.

**Timing**

The existing literature evaluating a single course of home-based pulmonary rehabilitation indicates that 3-6 months duration of follow-up is desirable to assess outcomes.

**Setting**

Patients with an indication for home-based pulmonary rehabilitation are managed by pulmonologists, primary care providers and ancillary clinical personnel.

**Study Selection Criteria**

Methodologically credible studies were selected using the following principles:

a. To assess efficacy outcomes, comparative controlled prospective trials were sought, with a preference for RCTs;

b. In the absence of such trials, comparative observational studies were sought, with a preference for prospective studies.

c. To assess longer term outcomes and adverse events, single-arm studies that capture longer periods of follow-up and/or larger populations were sought.

d. Studies with duplicative or overlapping populations were excluded

Evaluation of home-based PR programs requires evidence that these programs are at least as effective as programs conducted in the ambulatory care setting. The programs also need to be comprehensive and be feasible in the U.S. health care system.

**Systematic Reviews**

Several RCTs and systematic reviews of RCTs have assessed home-based PR programs. Among the systematic reviews, Liu et al (2014) identified 18 RCTs evaluating home-based PR programs. Most trials compared PR with usual care, and none of the selected trials compared home-based with clinic-based programs. Only 2 trials were conducted in the United States, and both were published in the 1990s. All trials reported different outcomes over different timeframes, and pooled analyses only included data from 2 to 4 studies. For example, a pooled analysis of 3 studies (n=112 patients) reporting the St. George’s Respiratory Questionnaire total score found statistically significant improvements in symptoms with home-based PR compared with control (effect size, -11.33; 95% CI, -16.37 to -6.29). A pooled analysis of data from 4 studies (n=167 patients) found a significantly increased 6MWD after 12 weeks in
the PR group compared with control (effect size, 35.9; 95% CI, 9.4 to 62.4). The latter analysis had a wide
confidence interval, indicating an imprecise estimate of effect.

Vieira et al (2010), in a systematic review, identified 12 RCTs comparing home-based PR with PR in
another setting or with standard care in patients who had COPD. The comparison intervention in 3
trials was a hospital-based program; in 8 trials, it was standard care; and in 1 trial, both
comparisons were made. The methodologic quality of the trials was considered average to poor, and
most had small sample sizes and relatively short follow-up durations. Reviewers did not pool trial
findings, and findings of individual studies were mixed. Three trials that compared home-based PR with
standard care reported on between-group differences in QOL; in all 3 studies, differences were
reported as statistically significant. The 2 trials that reported differences in exercise capacity found
home-based PR to result in significantly greater improvements in the 6MWD or constant work rate test
than standard care. On the other hand, in the 3 trials comparing home-based PR and hospital-based
programs, there were no statistically significant differences between groups in QOL changes. Moreover,
in the 2 trials that assessed maximal work level and the 2 trials that assessed the 6MWD, outcomes did
not differ significantly from home-based or hospital-based PR programs. Reviewers commented that
their analysis was limited by the generally low quality of the randomized trials and short-term length of
follow-up.

Another systematic review was published by Neves et al (2016). However, this review combined
home- and community-based PR programs in analyses so no conclusions can be drawn on the impact of
home-based programs compared with programs based in the ambulatory care setting.

Randomized Controlled Trials

A study with relatively large sample size and that compared home-based PR with outpatient clinic-based
PR was published by Maltais et al (2008). This noninferiority trial was conducted in Canada. Eligibility
criteria included stable COPD for at least 4 weeks before study participation and no previous
participation in PR programs; 252 patients were included. All patients initially completed a 4-week self-
management educational program. They were then randomized to 8 weeks of self-monitored home-
based exercise training or outpatient hospital-based exercise training. The exercise program included
aerobic and strength exercises conducted 3 times a week. Patients were followed for 40 weeks after
completion of the exercise program. Both interventions produced similar improvements in the CRQ
dyspnea domain scores at 1 year—improvement in dyspnea of 0.62 (95% CI, 0.43 to 0.80) units in the
home intervention (n=107) and 0.46 (95% CI, 0.28 to 0.64) units in the outpatient intervention (n=109).
The difference between treatments at 1 year was considered clinically unimportant. The trial did not
evaluate a comprehensive PR program.

Section Summary: Home-Based PR Programs

Most studies of home-based PR have compared it with standard care. Very few studies have compared
home-based PR with a hospital or clinic-based PR, and those available are mostly of low quality.
Therefore, there is insufficient evidence to determine whether comprehensive PR programs conducted
in the home setting are at least as effective as comprehensive PR programs in the ambulatory care
setting.

Summary of Evidence

Chronic Pulmonary Disease Rehabilitation

For individuals with moderate-to-severe COPD who receive a single course of outpatient PR, the
evidence includes numerous RCTs and systematic reviews. Relevant outcomes are symptoms, functional
outcomes, and quality of life. The published studies found improved outcomes (ie, functional ability, quality of life) in patients with moderate-to-severe COPD who underwent a comprehensive PR program in the outpatient setting. Among the many randomized trials, the structure of the PR programs varied, so it is not possible to provide guidance on the optimal components or duration of a PR program. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals with idiopathic pulmonary fibrosis who receive a single course of outpatient PR, the evidence includes an RCT. Relevant outcomes are symptoms, functional outcomes, and quality of life. The number of controlled studies is limited. One small RCT evaluated a comprehensive PR program in patients with idiopathic pulmonary fibrosis; at 3 months postintervention, outcomes did not differ between groups that did and did not receive PR. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals with bronchiectasis who receive a single course of outpatient PR, the evidence includes RCTs, systematic reviews, and observational data. Relevant outcomes are symptoms, functional outcomes, and quality of life. A systematic review of 4 RCTs on PR for patients with bronchiectasis found that some, but not all, outcomes, improved more with PR than with nonexercise control conditions immediately after the intervention. The evidence is insufficient to determine the effects of the technology on health outcomes.

**Preparation for Lung Surgery**

For individuals with scheduled lung surgery for volume reduction, transplantation, or resection who receive a single course of outpatient PR, the evidence includes RCTs and observational studies. Relevant outcomes are symptoms, functional outcomes, and quality of life. There is a lack of large RCTs comparing PR with no PR for preoperative candidates undergoing lung volume reduction surgery (LVRS), lung transplantation, or lung cancer resection. Moreover, the available studies have evaluated exercise programs, but not necessarily comprehensive PR programs. Also, the few small RCTs, and observational studies have only reported short-term outcomes and inconsistent evidence of benefit even on these outcomes. The evidence is insufficient to determine the effects of the technology on health outcomes.

**PR After Lung Surgery**

For individuals who have had LVRS who receive a single course of outpatient PR, the evidence includes a case series. Relevant outcomes are symptoms, functional outcomes, and quality of life. No published RCTs were identified. The case series evaluated a comprehensive PR program after LVRS in 49 patients who had not received preoperative PR. Health-related quality of life was higher at 3 to 6 months and 12 to 18 months postsurgery. The series did not provide data on patients who underwent LVRS and did not have postoperative PR, or patients who had preoperative PR. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have had lung transplantation who receive a single course of outpatient PR, the evidence includes RCTs, systematic reviews, and observational studies. Relevant outcomes are symptoms, functional outcomes, and quality of life. Neither of the 2 RCTs identified in a 2010 systematic review reported on functional outcomes, but uncontrolled studies have reported improvements in functional outcomes. An RCT, published after the systematic review, found that patients who had a postsurgical exercise intervention walked more 1 year post discharge than before and had a significantly greater 6-minute walk distance. Findings on other outcomes were mixed. The most recent RCT (2017) did not identify a difference in outcomes with longer duration of PR. Case series data also support
improvements in 6-minute walk distance after postoperative PR. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have had lung cancer resection who receive a single course of outpatient PR, the evidence includes an RCT. Relevant outcomes are symptoms, functional outcomes, and quality of life. One small RCT has evaluated a comprehensive PR program in patients who underwent thoracotomy for lung cancer. The trial was terminated early, had a high dropout rate, and reported mixed findings. An exercise-only intervention in patients who had lung cancer surgery had mixed findings and did not evaluate functional outcomes. The evidence is insufficient to determine the effects of the technology on health outcome.

**Repeat or Maintenance Rehabilitation**

For individuals who have had an initial course of PR who receive repeat or maintenance outpatient PR, the evidence includes RCTs. Relevant outcomes are symptoms, functional outcomes, and quality of life. There are only a few RCTs, and many of them have methodologic limitations and/or did not report clinically significant outcomes. The evidence is insufficient to determine the effects of the technology on health outcome.

**Home-Based Rehabilitation**

For individuals who have an indication for outpatient PR who receive a single course of home-based PR, the evidence includes RCTs and systematic reviews. Relevant outcomes are symptoms, functional outcomes, and quality of life. Most studies of home-based PR have compared outcomes with standard care. Very few have compared home-based PR with the hospital- or clinic-based PR, and the available studies are mostly of low quality. The evidence is insufficient to determine the effects of the technology on health outcome.

**SUPPLEMENTAL INFORMATION**

**Practice Guidelines and Position Statements**

**American Thoracic Society and European Respiratory Society**

A 2015 joint statement on pulmonary rehabilitation (PR) was issued by the American Thoracic Society and the European Respiratory Society. The statement included the following relevant conclusions:

- “PR has demonstrated physiological, symptom-reducing, psychosocial, and health economic benefits in multiple outcome areas for patients with chronic respiratory diseases.”
- “The evidence indicates that patients who benefit from PR include not only persons with moderate to severe airflow limitation but also those with mild to moderate airflow limitation with symptom-limited exercise tolerance, those after hospitalization for COPD exacerbation, and those with symptomatic non-COPD respiratory conditions.”
- “Patients graduating from a PR program stand to benefit from a home, community-based, or program-based maintenance exercise program to support the continuation of positive exercise behavior.”

**American College of Physicians**

Joint guidelines on the management of COPD were issued in 2011 by the American College of Physicians, the American College of Chest Physicians, American Thoracic Society, and European Respiratory Society. The guidelines recommended that: “clinicians should prescribe pulmonary rehabilitation for symptomatic patients with an FEV [forced expiratory volume] <50% predicted (Grade: strong recommendation, moderate-quality evidence). Clinicians may consider pulmonary rehabilitation for
symptomatic or exercise-limited patients with an FEV >50% predicted (Grade: weak recommendation, moderate-quality evidence).”

**American College of Chest Physicians**

In 2007, joint guidelines on PR for COPD and other chronic respiratory diseases were issued by American College of Chest Physicians and the American Association of Cardiovascular and Pulmonary Rehabilitation (see Table 1).34.

**Table 9. Pulmonary Rehabilitation Guidelines for Chronic Respiratory Diseases**

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>GOR</th>
</tr>
</thead>
<tbody>
<tr>
<td>A program of exercise training of the muscles of ambulation is recommended as a mandatory component of pulmonary rehabilitation for patients with COPD</td>
<td>1A</td>
</tr>
<tr>
<td>Pulmonary rehabilitation improves the symptom of dyspnea and improves health-related quality of life in patients with COPD</td>
<td>1A</td>
</tr>
<tr>
<td>Six to 12 weeks of pulmonary rehabilitation produces benefits in several outcomes that decline gradually over 12 to 18 months</td>
<td>1A</td>
</tr>
<tr>
<td>Both low- and high-intensity exercise training produce clinical benefits for patients with COPD</td>
<td>1A</td>
</tr>
<tr>
<td>Unsupported endurance training of the upper extremities is beneficial in patients with COPD and should be included in pulmonary rehabilitation programs</td>
<td>1A</td>
</tr>
<tr>
<td>Higher-intensity exercise training of the lower extremities produces greater physiologic benefits than lower-intensity training in patients with COPD</td>
<td>1B</td>
</tr>
<tr>
<td>Evidence does not support the routine use of inspiratory muscle training as an essential component of pulmonary rehabilitation</td>
<td>1B</td>
</tr>
<tr>
<td>Education should be an integral component of pulmonary rehabilitation; it should include information on collaborative self-management and prevention and treatment of exacerbations</td>
<td>1B</td>
</tr>
<tr>
<td>Pulmonary rehabilitation is beneficial for some patients with chronic respiratory diseases other than COPD</td>
<td>1B</td>
</tr>
</tbody>
</table>

COPD: chronic obstructive pulmonary disease; GOR: grade of recommendation.

**U.S. Preventive Services Task Force Recommendations**

Not applicable

**Medicare National Coverage**

In 2007, the Centers for Medicare & Medicaid Services affirmed its position that a national coverage determination for PR is not appropriate.35.

**Ongoing and Unpublished Clinical Trials**

Some currently unpublished trials that might influence this review are listed in Table 2.

**Table 10. Summary of Key Trials**

<table>
<thead>
<tr>
<th>NCT No.</th>
<th>Trial Name</th>
<th>Planned Enrollment</th>
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<tr>
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<tr>
<td>NCT03299504</td>
<td>Factors Predicting Success in Lung Transplant Recipients Who Have Participated in the COLTT Program (Daily Intensive Post-hospitalization Rehabilitation): A Retrospective Review</td>
<td>105</td>
<td>Apr 2018 (ongoing)</td>
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<td>NCT03326089</td>
<td>Short and Long-term Effects of Oxygen Supplemented</td>
<td>20</td>
<td>Jun 2019</td>
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ESSENTIAL HEALTH BENEFITS

The Affordable Care Act (ACA) requires fully insured non-grandfathered individual and small group benefit plans to provide coverage for ten categories of Essential Health Benefits (“EHBs”), whether the benefit plans are offered through an Exchange or not. States can define EHBs for their respective state.

States vary on how they define the term small group. In Idaho, a small group employer is defined as an employer with at least two but no more than fifty eligible employees on the first day of the plan or contract year, the majority of whom are employed in Idaho. Large group employers, whether they are self-funded or fully insured, are not required to offer EHBs, but may voluntary offer them.

The Affordable Care Act requires any benefit plan offering EHBs to remove all dollar limits for EHBs.

REFERENCES


<table>
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Outpatient Pulmonary Rehabilitation

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<tr>
<th>Type of Service</th>
<th>Place of Service</th>
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<tr>
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respiratory muscles, face-to-face, one-on-one, each 15 minutes (includes monitoring)

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>G0238</td>
<td>Therapeutic procedures to improve respiratory function, other than described by G0237, one-on-one, face-to-face, per 15 minutes (includes monitoring)</td>
</tr>
<tr>
<td>G0239</td>
<td>Therapeutic procedures to improve respiratory function or increase strength or endurance of respiratory muscles, 2 or more individuals (includes monitoring)</td>
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<tr>
<td>G0302</td>
<td>Preoperative pulmonary surgery services for preparation for LVRS, complete course of services, to include a minimum of 16 days of services</td>
</tr>
<tr>
<td>G0303</td>
<td>Preoperative pulmonary surgery services for preparation for LVRS, 10 to 15 days of service</td>
</tr>
<tr>
<td>G0304</td>
<td>Preoperative pulmonary surgery services for preparation for LVRS, 1 to 9 days of services</td>
</tr>
<tr>
<td>G0305</td>
<td>Postdischarge pulmonary surgery services after LVRS, minimum of 6 days of services</td>
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<tr>
<td>G0424</td>
<td>Pulmonary rehabilitation, including exercise (includes monitoring), one hour, per session, up to 2 sessions per day</td>
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<td>S9473</td>
<td>Pulmonary rehabilitation program, nonphysician provider, per diem</td>
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</table>

ICD-10-CM

<table>
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<tr>
<th>Code</th>
<th>Description</th>
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<tr>
<td>J99</td>
<td>Respiratory disorders in diseases classified elsewhere</td>
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<tr>
<td>D86.0-D86.9</td>
<td>Sarcoidosis; code range</td>
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<tr>
<td>D38.0-D38.6</td>
<td>Neoplasm of uncertain behavior of middle ear and respiratory and intrathoracic organs; code range</td>
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<td>E84.0-E84.9</td>
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<tr>
<td>D84.1</td>
<td>Defects in the complements systems</td>
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<tr>
<td>C96.6</td>
<td>Unifocal Langerhans-cell histiocytosis</td>
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<tr>
<td>I11.0-I11.9</td>
<td>Hypertensive heart disease; code range</td>
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<td>I27.0</td>
<td>Primary pulmonary hypertension</td>
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<td>J44.0-J44.9</td>
<td>Other Chronic obstructive pulmonary disease; code range</td>
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<td>J41.0-J41.8</td>
<td>Simple and mucopurulent chronic bronchitis; code range</td>
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<td>J43.0-J43.9</td>
<td>Emphysema; code range</td>
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<td>J47.0-J47.9</td>
<td>Bronchiectasis; code range</td>
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<tr>
<td>J84.0-J84.9</td>
<td>Other interstitial pulmonary diseases</td>
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<tr>
<td>M34.0-M34.9</td>
<td>Systemic sclerosis (scleroderma); code range</td>
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<tr>
<td>Q21.0</td>
<td>Ventricular septal defect (Eisenmenger’s syndrome)</td>
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<tr>
<td>P27.0-P27.9</td>
<td>Chronic respiratory disease originating in the perinatal period; code range</td>
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ICD-10-PCS

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