Aetna considers heart-lung transplantation medically necessary for persons with severe refractory heart failure plus either end-stage lung disease or irreversible pulmonary hypertension, when the selection criteria listed below are met and no absolute contraindications listed below are present. Examples of qualifying conditions include the following:

- Chronic obstructive pulmonary disease with severe heart failure*
- Congenital heart disease associated with pulmonary hypertension that are not amenable to lung transplantation and repair by standard cardiac surgery
- Cystic fibrosis with severe heart failure*
- Eisenmenger’s complex with irreversible pulmonary hypertension and severe heart failure*
- Irreversible primary pulmonary hypertension with severe heart failure*
- Connective tissue disease or other causes of severe pulmonary fibrosis with uncontrollable pulmonary hypertension or severe heart failure*
- Severe coronary artery disease or cardiomyopathy with
irreversible pulmonary hypertension

* Severe (New York Heart Association (NYHA) classification III or IV (see Appendix)) heart failure where right ventricular function would not be restored with lung transplant alone. **Note:** Heart-lung transplantation is considered not medically necessary where lung transplantation alone will restore right ventricular function; every attempt should be made to preserve the heart.

**Note:** Heart-lung transplantation may be considered medically necessary for other congenital cardiopulmonary anomalies upon individual case review.

**Selection Criteria:** The member must meet the transplanting institution’s selection criteria. In the absence of an institution's selection criteria, Aetna considers heart-lung transplantation medically necessary when all of the criteria below are met:

1. Absence of chronic high-dose steroid therapy. Due to problems in bronchial healing, persons receiving high-dose steroids are considered inappropriate candidates; and
2. Absence of acute or chronic active infections that are not effectively treated; and
3. Absence of malignancy (other than non-melanomatous skin cancers or low-grade prostate cancer) or malignancy has been completely resected or (upon medical review) it is determined that malignancy has been treated with small likelihood of recurrence and acceptable future risks; and
4. Adequate functional status. Active rehabilitation is considered important to the success of transplantation. Under established guidelines, mechanically ventilated or otherwise immobile persons are considered poor candidates for transplantation; however, bridge to transplant with ambulatory ECMO does not, in itself, rule out candidacy for heart-lung transplantation; and
5. Adequate liver and kidney function, defined as a bilirubin of less than 2.5 mg/dL and a creatinine clearance of greater than 50 ml/min/kg; and
6. Life expectancy (in the absence of cardiopulmonary disease) of greater than 2 years; and
7. No active alcohol or chemical dependency that interferes with compliance to a strict treatment regimen; and
8. No uncontrolled and/or untreated psychiatric disorders that interfere with compliance to a strict treatment regimen; and
9. HIV/AIDS, if present, is under adequate control, defined as:
   - CD4 count greater than 200 cells/mm³ for more than 6 months; and
   - HIV-1 RNA (viral load) undetectable; and
   - On stable anti-viral therapy more than 3 months; and
   - No other complications from AIDS, such as opportunistic infections (e.g., aspergillus, tuberculosis, Pneumocystis carinii pneumonia, toxoplasmosis encephalitis, cryptococcal meningitis, disseminated coccidioidomycosis, other resistant fungal infections) or neoplasms (e.g., Kaposi’s sarcoma, non-Hodgkin’s lymphoma).

Contraindications:

Heart-Lung transplant is considered not medically necessary for persons with any of the following contraindications because the risks of transplantation exceed the benefits:

- Gastro-intestinal disease (e.g., bleeding peptic ulcer, diverticulitis, chronic hepatitis, active or recurrent pancreatitis)
- Multi-system disease. Persons with potentially multi-system diseases such as systemic sclerosis (scleroderma) or other collagen vascular diseases such as systemic lupus erythematosus must be carefully evaluated to ensure that their disease is primarily confined to the lung. Persons with diabetes must be carefully evaluated to rule out significant diabetic complications such as nephropathy, neuropathy or retinopathy.
- Other effective medical treatments or surgical options are available
- Progressive neuromuscular disease
- Refractory uncontrolled hypertension
- Severe musculoskeletal disease with debilitating thoracic involvement
- Smoking. Persons with a history of smoking must be abstinent for at least 3 months before being considered a candidate for lung transplant
- Untreated or unstable cerebrovascular disease.

Background
Reduced cardiac output and edema characterize patients with heart failure, while patients with respiratory failure exhibit abnormalities in oxygenation and carbon dioxide elimination that impair the functioning vital organs. These patients have a lower life expectancy and decreased quality of life. Since its introduction at Stanford University in 1981 and at Pittsburgh University in 1982 for the treatment of Eisenmenger's syndrome and terminal pulmonary vascular disease, heart-lung transplantation has become successful therapeutic options for patients with end-stage cardiopulmonary disease. Heart-lung transplantation is most frequently performed for patients with congenital heart disease (about 30%), primary pulmonary hypertension (about 27%), cystic fibrosis (about 16%), miscellaneous conditions (about 15%), lung emphysema (about 4%), re-transplantation following a failed primary transplant (about 3%), idiopathic pulmonary fibrosis (about 3%) and alpha 1-antitrypsin deficiency (about 2%).

Adults who have undergone heart-lung transplantation for congenital heart disease are expected to have survival comparable to that of adults without congenital heart disease. Furthermore, heart-lung transplantation results in survival comparable to that reported for single- or double-lung transplantation for patients with primary pulmonary hypertension. Obliterative bronchiolitis, a form of chronic rejection, is a significant cause of late death.

The frequency of heart-lung transplantation is partly limited by the number of available donor organs. There are fewer donor
heart-lung preparations than donor heart preparations alone because brain death may be associated with neurogenic pulmonary edema. In addition, aspiration into the lung is common during the course of severe trauma and resuscitation. Prolonged ventilatory support may also predispose the potential donor to nosocomial infection, and direct thoracic trauma may result in pulmonary contusion. As a consequence, probably less than 20% of potential heart donors have lungs that are suitable for heart-lung transplantation. It should be noted that heart-lung transplantation in some ways is a technically easier procedure than heart transplantation since the former requires only right atrial, aortic, and tracheal anastomoses, thus avoiding several of the anastomoses associated with heart transplantation.

Contraindications to heart-lung transplantation include irreversible end-organ diseases (e.g., renal, hepatic), active malignancy or infections, systemic diseases (e.g., autoimmune, vascular, amyloidosis), chronic gastro-intestinal disease (e.g., diverticulitis, active or recurrent pancreatitis, bleeding peptic ulcer), psychiatric disorders, cerebrovascular disease, progressive neuromuscular disease, and use of tobacco products. Under established guidelines, obese (greater than 20% of ideal body weight), cachectic (less than 80% of ideal body weight), mechanically ventilated or otherwise immobile patients are considered poor candidates for transplantation.

Early post-operative complications (within the first post-operative month) comprise acute isolated lung rejection, multi-organ failure, and bacterial pneumonia. Late post-operative complications (after 1 post-operative month) comprise viral pneumonia, fungal infection, tuberculosis, and chronic obliteratorive bronchiolitis.

Zheng et al (2011) examined the safety and possible benefits of laparoscopic anti-reflux surgery in pediatric patients following lung and heart-lung transplantation. An Institutional Review Board-approved retrospective chart review was performed to evaluate the outcomes and complications of laparoscopic
anti-reflux surgery in pediatric lung and heart-lung transplant patients. Spirometry data were collected for bronchiolitis obliterans syndrome (BOS) staging using BOS criteria for children. A total of 25 lung and heart-lung transplants were performed between January 2003 and July 2009. Eleven transplant recipients, including 6 double-lung and 5 heart-lung, with a median age of 11.7 years (range of 5.1 to 18.4 years), underwent a total of 12 laparoscopic Nissen fundoplications at a median of 427 days after transplant (range of 51 to 2310 days). The diagnosis of gastro-esophageal reflux disease (GERD) was made based upon clinical impression, pH probe study, gastric emptying study, and/or esophagram in all patients. Three patients already had a gastrostomy tube in place and 2 had one placed at the time of fundoplication. There were no conversions to open surgery, 30-day re-admissions, or 30-day mortalities. Complications included 1 exploratory laparoscopy for free air 6 days after laparoscopic Nissen fundoplication for a gastric perforation that had spontaneously sealed. Another patient required a revision laparoscopic Nissen 822 days following the initial fundoplication for a para-esophageal hernia and recurrent GERD. The average length of hospital stay was 4.4 +/- 1.7 days. Nine of the 12 fundoplications were performed in patients with baseline spirometry values prior to fundoplication and who could also complete spirometry reliably. One of these 9 operations was associated with improvement in BOS stage 6 months after fundoplication; 7 were associated with no change in BOS stage; and 1 was associated with a decline in BOS stage. The authors concluded that it is feasible to perform laparoscopic Nissen fundoplication in pediatric lung and heart-lung transplant recipients without mortality or significant morbidity for the treatment of GERD. The real effect on pulmonary function can not be assessed due to the small sample size and lack of reproducible spirometry in the younger patients. The authors stated that additional studies are needed to elucidate the relationship between anti-reflux surgery and the potential for improving pulmonary allograft function and survival in children that has been previously observed in adult patients.
Olland and colleagues (2013) examined which of the following 2 procedures: (i) heart-lung transplantation or (ii) bilateral-lung transplantation (BLTx) offers the best outcome for patients with pulmonary hypertension (PH) listed for thoracic transplantation? Of the 77 papers found using a report search for PH and thoracic transplantation, 9 represented the best evidence to answer this clinical question. Overall, 1,189 (67 %) lung transplantations and 578 (33 %) heart-lung transplantations have been reported worldwide for idiopathic PH. For patients with Eisenmenger’s syndrome, HLTx represents up to 70 % of the transplantation procedures they undergo. On the whole, neither procedure demonstrated an overall survival benefit, when compared with the other. However, PH patients represent a heterogeneous population according to (i) the primary mechanism of PH and (ii) the consequences of PH on right or/and left heart function. With regard to the latter consideration, the evidence showed that HLTx offers excellent functional and survival outcomes for patients with congenital heart disease and Eisenmenger’s syndrome, severe right or/and left heart dysfunction, and who are chronically inotropic dependent. As far as heart dysfunction is concerned, the published evidence approximated cut-off values at 10 to 25 % for the right ventricle ejection fraction (RVEF) and at 32 to 55 % for the left ventricle ejection fraction (LVEF). In the case of lower values for RVEF and LVEF, HLTx should be performed. In all other patients with PH, the evidence demonstrated that BLTx offers a comparable outcome with the advantage of better organ sharing for other recipients. In order to reduce the waiting time on transplantation lists, cardiac repair and BLTx can be offered in experienced centers to patients with simple cardiac anomalies such as atrial septal defect, patent ductus arteriosus or peri-membranous ventricular septal defect.

Appendix

The New York Heart Association (NYHA) classification of heart failure is one of the many parameters used for selecting heart-lung recipient. It is a 4-tier system that categorizes
patients based on subjective impression of the degree of functional compromise. The 4 NYHA functional classes are as follows:

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I:</td>
<td>Patients with cardiac disease but without resulting limitation of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain. Symptoms only occur on severe exertion.</td>
</tr>
<tr>
<td>Class II:</td>
<td>Patients with cardiac disease resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity (e.g., moderate physical exertion such as carrying shopping bags up several flights or stairs) results in fatigue, palpitation, dyspnea, or anginal pain.</td>
</tr>
<tr>
<td>Class III:</td>
<td>Patients with cardiac disease resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity (i.e., mild exertion) causes fatigue, palpitation, dyspnea, or anginal pain.</td>
</tr>
<tr>
<td>Class IV:</td>
<td>Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort. Symptoms of cardiac insufficiency or of the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort is increased.</td>
</tr>
</tbody>
</table>

### CPT Codes / HCPCS Codes / ICD-10 Codes

*Information in the [brackets] below has been added for clarification purposes. Codes requiring a 7th character are represented by "+":*

*ICD-10 codes will become effective as of October 1, 2015:*

**CPT codes covered if selection criteria are met:**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>33930</td>
<td>Heart/lung transplant</td>
</tr>
<tr>
<td>33945</td>
<td></td>
</tr>
</tbody>
</table>

**HCPCS codes covered if selection criteria are met:**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>S2054</td>
<td>Transplantation of multivisceral organs</td>
</tr>
</tbody>
</table>
### ICD-10 codes covered if selection criteria are met:

<table>
<thead>
<tr>
<th>Code</th>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>E84.0 - E84.9</td>
<td>Cystic fibrosis</td>
</tr>
<tr>
<td>I25.10 - I25.9</td>
<td>Chronic ischemic heart disease</td>
</tr>
<tr>
<td>I26.01 - I127.9</td>
<td>Pulmonary heart disease</td>
</tr>
<tr>
<td>I42.0 - I43</td>
<td>Cardiomyopathy</td>
</tr>
<tr>
<td>I50.1 - I50.9</td>
<td>Heart failure</td>
</tr>
<tr>
<td>J40 - J47.9</td>
<td>Chronic lower respiratory diseases</td>
</tr>
<tr>
<td>J84.10</td>
<td>Pulmonary fibrosis, unspecified</td>
</tr>
<tr>
<td>J84.89</td>
<td>Other specified interstitial pulmonary diseases</td>
</tr>
<tr>
<td>M32.0 - M35.9</td>
<td>Diffuse diseases of connective tissue</td>
</tr>
<tr>
<td>Q20.0 - Q28.9</td>
<td>Congenital malformations of the circulatory system</td>
</tr>
<tr>
<td>T86.20 - T86.298</td>
<td>Complications of heart transplant</td>
</tr>
<tr>
<td>T86.30 - T86.19</td>
<td>Complications of heart-lung transplant</td>
</tr>
<tr>
<td>T86.810 - T86.819</td>
<td>Complications of lung transplant</td>
</tr>
</tbody>
</table>

---

**The above policy is based on the following references:**


25. Scouras NE, Matsusaki T, Boucek CD, et al. Portopulmonary hypertension as an indication for combined heart, lung, and liver or lung and liver


The Pennsylvania Medical Assistance Program considers HIV/AIDS to be under adequate control when all of the following criteria are met:

- CD4 count greater than 200 cells/mm³ for more than 6 months; and HIV-1 RNA (viral load) less than 200 copies/ml; and
- On stable anti-viral therapy more than 3 months; and No other ongoing or untreated complications from AIDS, such as opportunistic infections (e.g., aspergillus, tuberculosis, Pneumocystis carinii pneumonia, toxoplasmosis encephalitis, cryptococcal meningitis, disseminated coccidioidomycosis, other resistant fungal infections) or neoplasms (e.g., Kaposi’s sarcoma, non-Hodgkin’s lymphoma).