**e-Appendix 1.**

Methods

"Stable" encounters were identified as visits that occurred in clinic, and were not labeled as "home IV" or "hospital" encounters. After 2010, a pulmonary exacerbation indicator was added to the Cystic Fibrosis Foundation Patient Registry and a "stable" encounter was also required to have "absent" marked for the pulmonary exacerbation indicator.

Results

e-Figure 1: Outcomes for CF patients with FEV$_1$ <30%
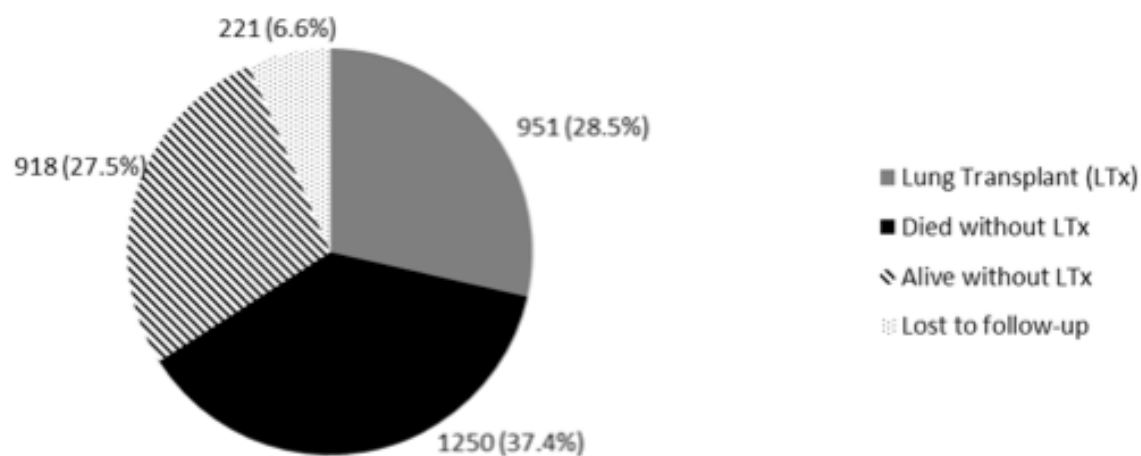
e-Figure 2: Outcomes for CF patients with FEV$_1$ <30% after referral for LTx evaluation
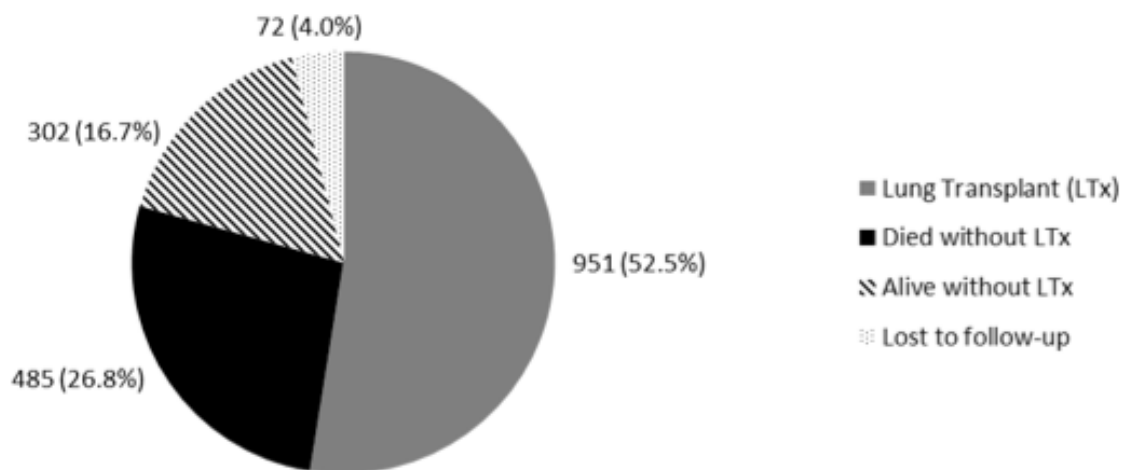
e-Table 1: Sensitivity analysis to determine number of pulmonary exacerbations associated with clinically significant shortened median survival

	Adjusted HR1	Median survival (95% CI)	p-value
≥1 exacerbation/year	1.71 (1.34-2.18)	5.1 years (4.9-5.6)	<0.001
No exacerbations	Ref	9.6 years (8.3-10.3)	
≥2 exacerbations/year	1.64 (1.34-1.99)	4.0 years (3.5-4.3)	<0.001
0-1 exacerbation/year	Ref	8.5 years (7.9-9.4)	
≥3 exacerbations/year	1.59 (1.29-1.97)	3.2 years (2.7-3.5)	<0.001
0-2 exacerbations/year	Ref	7.9 years (7.3-8.5)	
≥4 exacerbations/year	1.68 (1.29-2.19)	2.6 years (2.2-3.0)	<0.001
0-3 exacerbations/year	Ref	7.3 years (6.9-7.8)	
≥5 exacerbations/year	1.93 (1.37-2.72)	2.0 years (1.7-2.5)	<0.001
0-4 exacerbations/year	Ref	7.0 years (6.5-7.4)	

Median survival ≤2 years is the recommended time for referral for lung transplant evaluation and is, therefore, clinically important

1When categorized pulmonary exacerbations entered into final multivariate Cox proportional hazards (PH) regression in place of continuous measure of pulmonary exacerbations, stratified by calendar time and FEV1 in 5% increments, there is no violation of the PH assumption

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