



Cystic Fibrosis Research News

Title:

Lung Transplant Referral for Individuals with Cystic Fibrosis: Cystic Fibrosis Foundation Consensus Guidelines

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What was your research question?

When should people with CF and their families discuss lung transplantation as a potential treatment option and when should CF doctors refer a person with CF to a lung transplant center?

Why is this important?

CF is a disease that affects many organ systems in the body, but most profoundly affects lung function for a majority of people with CF. Over time, a decline in lung function leads to death for most people with CF. Lung transplantation is a potential treatment option for advanced CF-related lung disease, but some people die without consideration of lung transplant. Each year in the United States, more people with CF die from CF-related lung disease than undergo lung transplantation. The timing of

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referral to a transplant center has been described in international recommendation statements, but practical instructions for CF care providers have not previously been published.

What did you do?

The CF Foundation brought together a multidisciplinary team of experts in CF and lung transplantation to give guidance to the CF community that could decrease the number of people with CF who die without the opportunity to consider a lung transplant. This team performed an extensive search of the available scientific evidence related to lung transplantation, the transition to transplant care, and death from CF. We assembled a group of lung transplant recipients with CF and spouses of CF transplant recipients in order to inform the multidisciplinary team of patients' perspectives where data were lacking.

What did you find?

We made 21 recommendation statements, which were released for public comment in the fall of 2018. We found that members of the CF community would welcome earlier discussion of disease trajectory, and specifically of lung transplantation as a potential future treatment option, in order to increase awareness of the indications and timing for transplant. We identified and described potentially modifiable barriers to lung transplantation. Importantly, we gave CF teams a structured schedule of testing to perform in the CF clinic that aims to identify people with CF who may be sicker than their forced expiratory volume in 1 second (FEV₁) would imply.

What does this mean and reasons for caution?

The CF lung transplant referral consensus guidelines will identify people with CF who could benefit from further risk assessment, screening for barriers to transplant, and education about the transplant process. The recommendations are for referral to a lung transplant center, but will identify many people who are "too early" to list for transplant. It is important to recognize that referral to a transplant center is not the same as transplant evaluation or listing. While discussions about lung transplant are difficult and can be emotional, the guidelines aim to increase the amount of time people have to process the concept of transplant prior to actually needing a transplant.

What's next?

Future research will focus on evaluation of these referral recommendations in the setting of CFTR modulator therapies. Additional research is needed to determine the optimal timing to propose listing people with CF for lung transplant.

Original manuscript citation in PubMed

<https://www.ncbi.nlm.nih.gov/pubmed/?term=Lung+Transplant+Referral+for+Individuals+with+Cystic+Fibrosis%3A+Cystic+Fibrosis+Foundation+Consensus+Guidelines>