

# Cystic Fibrosis Research News

## Title:

CAUSES OF DEATH IN FRENCH CYSTIC FIBROSIS PATIENTS: THE NEED FOR IMPROVEMENT IN TRANSPLANTATION REFERRAL STRATEGIES!

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## What was your research question?

The 2011 French Cystic Fibrosis Registry report revealed that half of the deaths in CF patients occurred without lung transplantation. The aims of this study were to describe specific causes of death and to determine reasons for not receiving lung transplantation.

## Why is this important?

Respiratory failure is the main cause of death in CF patients. Although significant improvements in survival of patients have been achieved by multidisciplinary care, respiratory failure still occurs and in this situation, lung transplantation has the potential to improve life expectancy. Therefore, it is important to understand the reasons why half of the patients dying with CF did not receive lung transplantation.

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## What did you do?

The French CF Registry (covering around 88% of the French CF population) was used to identify deaths and medical information was obtained from medical files of patients which death occurred between 2007 and 2010. This data was then analysed by an independent mortality adjudication committee (MAC). Each medical file was investigated by MAC members to assign the cause of death, transplantation status at time of death and reasons for not being transplanted.

## What did you find?

Of the 256 deaths recorded during this time period, half occurred after lung transplantation and were related to early or late complications of transplantation, whereas half occurred in patients who did not receive lung transplantation and were primarily related to respiratory failure or massive hemoptysis (bronchial bleeding). Among patients who did not receive lung transplantation, only 19% died while waiting on the lung transplantation list. Approximately 40% of patients who died without transplantation would have been eligible for lung transplantation.

## What does this mean and reasons for caution?

Lack of listing for lung transplantation was primarily related to late, or lack of transplantation referral, rather than to ineligibility or patient refusal to proceed with the assessment for transplant. This data suggests that improvement in transplantation referral strategies may result in improved transplantation-related survival benefit.

## What's next?

The situation detailed in this study is unlikely to be limited to France alone and suggests action should be taken by other developed countries to improve the treatment options of CF sufferers. Our data suggests the need for discussing lung transplantation early on in all CF patients with severe respiratory disease.

## Original manuscript citation in PubMed

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