

# Cystic Fibrosis Research News

**Title:**

Non-Invasive Ventilation and Clinical Outcomes in Cystic Fibrosis: findings from the UK CF Registry

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**What was your research question?**

To describe the patterns and factors related to the use of non-invasive ventilation (NIV), a type of ventilatory support through the patient's airway using a mask or a similar device, in UK patients with cystic fibrosis (CF), and assess its effects on lung function and survival.

**Why is this important?**

Although NIV is often used in patients with very advanced CF, it is not known whether NIV has benefits in terms of lung function or survival. This is the first large scale study of NIV used in the UK. Over 8 years (2007-2015) of data were analysed to understand the use and impact of NIV on CF patients.

**What did you do?**

Using information from the UK CF Registry, we explored the patterns of NIV use in children and adult CF patients. We assessed whether NIV has improved, preserved, or worsened patients' lung function and determined whether NIV had any impact on survival

**What did you find?**

The use of NIV has increased across years. We found a mild improvement of lung function after the first use of NIV in children. In adults, who generally had more severe lung dysfunction when they first started NIV, the improvement was much stronger. We found that NIV initiation did not have an effect on survival or lung transplantation in either children or adults.



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## **What does this mean and reasons for caution?**

In this UK study, treatment with NIV slowed or reversed the decline in lung function in both children and adults with CF. However, use of NIV did not extend the amount of time that patients survived. This may be because NIV is not used until patients' end-stage CF, in which case they may have exhausted standard medical interventions.

## **What's next?**

The specific reasons for NIV initiation were probably variable. A more standardised approach with consensus guidelines is needed in order to better understand which types of patients may benefit most from NIV.

## **Original manuscript citation in PubMed**

<https://www.ncbi.nlm.nih.gov/pubmed/?term=Non-Invasive+Ventilation+and+Clinical+Outcomes+in+Cystic+Fibrosis%3A+findings+from+the+UK+CF+Registry>