**Title:**

Long-term Outcomes in People with Cystic Fibrosis Lacking Early Lung Function Response to Elexacaftor/Tezacaftor/Ivacaftor Therapy.

**Lay Title:**

ETI therapy shows long-term health benefits—even when early lung function doesn’t improve.

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**What was your research question?**

Do people with cystic fibrosis still benefit from elexacaftor/tezacaftor/ivacaftor (ETI) therapy even if their lung function doesn’t improve in the first few months of treatment?

**Why is this important?**

ETI is a breakthrough in treating CF. Many people show early improvement in lung function, but some do not. This raises important questions: Is the treatment working for them? Should it be continued? Understanding whether people still benefit over time—even without early improvements—is important for treatment decisions and for health systems that may rely on early response to approve ongoing care.

**What did you do?**

We followed 52 people with CF at our center after they started ETI therapy. We focused on a group we called “non-early responders”— people whose lung function (measured by FEV₁) did not improve during the first three months of treatment. We compared them with people who did respond early and tracked their health over 18 to 24 months. We looked at lung function, infections, flare-ups (pulmonary exacerbations), body weight, and chest CT scans, to assess the full impact of ETI over time.

**What did you find?**

People with CF who didn’t show early improvement still experienced important health gains. After nearly 2 years on ETI, they had:

* Fewer lung flare-ups needing IV antibiotics
* Slower decline in lung function
* Better weight and nutrition
* Healthier-looking lungs on CT scans

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

This study offers reassurance: ETI still helps even when lung function doesn’t improve right away. Early breathing test results are not the only sign of benefit. Weight, infection frequency, flare-ups and chest imaging results are also important. However, this was a single-center study with a relatively small group of participants. Larger, multi-center studies are needed to confirm these findings, and individual patient care must always be personalized.

**What’s next?**

More research is needed to understand why some people don’t show early improvements yet still benefit over time, and to find better ways to measure how well ETI is working. For now, our findings suggest that the decision to continue treatment should not rely on early improvement alone, as meaningful benefits still develop over time.

**Original manuscript citation in PubMed**

<https://pubmed.ncbi.nlm.nih.gov/40461393/>