

# Cystic Fibrosis Research News

**Title:**

ITALIAN AND NORTH AMERICAN DIETARY INTAKE AFTER IVACAFTOR TREATMENT FOR CYSTIC FIBROSIS GATING MUTATIONS

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**What was your research question?**

Given the remarkable weight gain that has been demonstrated in patients with cystic fibrosis and gating mutations taking ivacaftor, we wanted to explore if the medication had an impact on dietary intake.

**Why is this important?**

Weight gain with ivacaftor therapy is particularly promising, as optimization of growth and weight status is linked to improved outcomes in cystic fibrosis. A change in dietary intake in patients with cystic fibrosis and gating mutations who take ivacaftor is a possible mechanism of weight gain on this medication. Improved dietary intake may be due to an overall improvement in well-being and gastrointestinal symptoms while taking ivacaftor.

**What did you do?**

We conducted a clinical trial on 22 patients with an average age of 17.8 years. 15 of the patients were from Italy and seven were from North America. We measured the average 3-day dietary intake before ivacaftor was started and 3 months after initiation of ivacaftor treatment in people with cystic fibrosis and gating mutations. Dietary intake was measured by a 3-day food record that was then analysed by a nutritional database and a dietitian to determine the composition of the diet. Other parameters measured before and after the initiation of ivacaftor included body composition, pulmonary function, gastrointestinal tract inflammation, and fat absorption.



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## What did you find?

We found that there was no significant difference in the number of calories that people with CF consumed before and after they started ivacaftor. When analyzed individually, the group of patients from Italy did have an increase in the number of calories they ate after starting ivacaftor. We also found that people with CF ate more dietary fat once they started to take ivacaftor. It was notable that the more a patient in the study increased the amount of fat and calories that they eat, the more they tended to gain weight. The patients in this study demonstrated an increase in height, weight, body mass index, and pulmonary function (FEV<sub>1</sub>), as shown in previous studies.

## What does this mean and reasons for caution?

This study demonstrates that ivacaftor resulted in increased dietary fat intake in our study participants. Furthermore, a greater increase in intake of fat and calories was associated with greater weight gain. Patients are instructed to take ivacaftor twice daily with fat-containing foods to optimize absorption. This may partly account for the increased fat intake overall, but the relationship between high fat intake and time of medication intake could not be determined from our study. As nutritional status is closely linked to long-term survival in patients with cystic fibrosis, it was important to demonstrate that one of the mechanisms of weight gain was related to increased dietary intake. Limitations of this study include that different software programs were used to analyse dietary composition in patients from Italy compared to those from North America. We also studied a limited number of patients. Finally, this was an observational study and it is possible that some of the changes in outcomes we observed were due to improved adherence to other aspects of treatment.

## What's next?

Long-term nutritional outcomes of patients with gating mutations taking ivacaftor have not been determined. While the CF care team strive to promote weight gain, monitoring weight patterns to also avoid excess weight gain is now a part of care in some patients.

## Original manuscript citation in PubMed

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