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Cystic Fibrosis Research News

Title:
Treatment patterns in people with cystic fibrosis: have they changed since the introduction of ivacaftor?

Lay Title:
Treatment patterns in people with cystic fibrosis: have they changed since the introduction of ivacaftor?

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What was your research question?
We wanted to know whether there were changes in the long-term use of other key treatments in people with cystic fibrosis (CF) after the introduction of ivacaftor in the UK, among people who started using ivacaftor.

Why is this important?
Existing research shows that ivacaftor helps improve health outcomes such as better lung function and less hospital admissions in people with CF. Ivacaftor is taken alongside other long term CF treatments. Treatment burden is an important factor in the quality of life of people with CF and findings ways to reduce treatment burden is a top priority in the CF community. Investigating whether treatment use has changed since the introduction of ivacaftor is an important step in understanding whether treatment burden can be reduced for people who take ivacaftor.

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What did you do?

Using data from the UK Cystic Fibrosis Registry, we compared treatment use in ivacaftor-treated people with treatment use in people in the same time period (2013-2018), but not receiving ivacaftor due to their combination of genes ('genotype'). This allowed us to investigate whether ivacaftor users are more or less likely, on average, to use other treatments compared to ivacaftor non-users. We also compared treatment use in similar groups defined by genotype, but in the time period before ivacaftor became available (2007-2012). We considered several of the key treatments used in CF.

What did you find?

Before ivacaftor was introduced (2007-2012), treatment patterns over time were similar between the two groups of people defined by genotype. Once ivacaftor was introduced (2013-2018), we found a clear divergence in treatment use over time between people treated with ivacaftor and those untreated due to their genotype. People treated with ivacaftor were less likely to continue other treatments such as inhaled antibiotics, dornase alfa, hypertonic saline, oral antibiotics and supplementary feeding, compared to ivacaftor non-users. We also found that treatment differences between ivacaftor users and ivacaftor non-users were most pronounced for children and for those with high lung function.

What does this mean and reasons for caution?

Use of some treatments has changed since the introduction of ivacaftor. People treated with ivacaftor were more likely to withdraw from other treatments, compared to people not treated with ivacaftor. This suggests that people who are treated with ivacaftor may experience reduced treatment burden overall.

However, we don't know the reasons why ivacaftor-treated people are withdrawing from treatments and we don't know the impact this could be having on their health.

What's next?

Our next step is to investigate the impact of changes in treatment use on health. We will investigate whether the ivacaftor-treated people who are withdrawing from other treatments are still as healthy in terms of clinical indicators such as lung function compared with similar people who continued other treatments.

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