

Cystic Fibrosis Research News

Title:

Females with Cystic Fibrosis have a larger decrease in sweat chloride in response to lumacaftor/ivacaftor compared to males

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What was your research question?

What leads to the change in sweat test results after the start of lumacaftor/ivacaftor, and is a larger change in sweat test results linked to a larger change in lung function or nutritional state?

Why is this important?

For many years, the sweat test was only used to diagnose cystic fibrosis. With the introduction of new drugs for CF, the test also became very popular to use in follow up, after the starting of treatment. Little is known, however, about how to interpret the change in sweat test. Could we expect the same change in every patient? Does a larger change always mean a better response, for example in lung function?

What did you do?

We collected information about 160 individuals who started lumacaftor/ivacaftor (Orkambi) and had a sweat test and a lung function test, were weighed and had their height measured (to calculate BMI, a measure for nutritional state), both before the start and after 6 months of treatment. We analysed the data to see if a larger response was associated with younger/older age, sex, baseline sweat test, body weight, or with change in lung function or BMI.

What did you find?



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Our most important finding is that there was a greater change in sweat test results in females than in males. The change in BMI was also greater in females, while lung function change did not differ much between sexes.

Secondly, there is no clear relation between any change in sweat test results and any change in BMI or lung function.

What does this mean and reasons for caution?

There seem to be differences in the response to lumacaftor/ivacaftor between males and females, with females having a better response. However, we cannot explain why this is, and it does not seem to apply for lung function.

Also, there is a reason for caution when using the sweat test as a follow up tool: the change in sweat test does not tell a lot about the change in lung function or BMI.

What's next?

We aim to check if this difference in how males and females respond to treatment also occurs with the more powerful triple therapy.

We also need to look into why there are different responses between males and females: Is it adherence to therapy? Differences in drug uptake? Hormonal effects? We don't know! Yet.

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