European Cystic Fibrosis Society

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Cystic Fibrosis Research News

Title:
Onset of systemic arterial hypertension after initiation of elexacaftor/tezacaftor/ivacaftor in adults with cystic fibrosis: a case series.

Lay Title:
Onset of high blood pressure might be possible after the start of elexacaftor/tezacaftor/ivacaftor in adults with cystic fibrosis.

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What was your research question?
Clinical studies showed that Elexacaftor/Tezacaftor/Ivacaftor (ETI), CFTR correcting drugs, were well tolerated, and discontinuations happened very rarely. The most common adverse events reported were high liver enzymes and headache. However, our personal experience through real-world clinical practice raised the attention to adverse events not previously described. We want to share with the CF community the possible onset of high blood pressure after the start of ETI treatment in individuals with CF.

Why is this important?
Although the onset of high blood pressure was not reported in clinical studies of CFTR modulators, this case series highlights to CF physicians and patients that careful monitoring of cardiovascular parameters may be required. This includes the identification and correction of common risk factors for high blood pressure in ant preliminary assessment prior to the start of ETI treatment.

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What did you do?

We reported a series of four patients with no history of cardiovascular disease, who developed high blood pressure associated with ETI.

What did you find?

We found that in all cases patients complained of headache and other symptoms related to high blood pressure. High blood pressure was ongoing even after discontinuation of common risk factors, such as salt supplementation treatment. All patients needed a careful evaluation by a heart specialist and started long-term drugs to treat high blood pressure.

What does this mean and reasons for caution?

These observations might suggest that a complex network of interactions exists between ETI, extra-pulmonary effects of restored CFTR protein function and underlying metabolic processes. The restoration of CFTR function is supposed to correct loss of water and salts and might sustain an increase in blood pressure. In addition, the possible interaction between ETI and unspecified hormonal patterns which has been hypothesized to have a role in skin rash events after starting ETI, deserves more investigations.

What's next?

In terms of implications for research, investigating the extra-pulmonary effects of the ETI will likely be a priority for the next few years.

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