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Cystic Fibrosis Research News

Title:

The Relationship between Sweat Chloride Levels and Mortality in Cystic Fibrosis Varies by Individual Genotype

Authors:

Julia C. Espel[1], Hannah L. Palac[2], Ankit Bharat[3], Joanne Cullina[4], Michelle Prickett[1], Marc Sala[1], Susanna A. McColley[4], Manu Jain[1]

Affiliations:

[1]Division of Pulmonary and Critical Care Medicine, Depart of Medicine Feinberg School of Medicine, Northwestern University Chicago, IL
[2]Department of Preventive Medicine Northwestern University Chicago, IL
[3]1Division of Thoracic Surgery, Department of Surgery Feinberg School of Medicine, Northwestern University Chicago, IL
[4]Lurie Children's Hospital Chicago, IL

What was your research question?

Previous research is conflicting on whether sweat chloride values, which are a direct measure of CFTR function, are associated with life expectancy in CF patients. We asked whether sweat chloride values are associated with life expectancy if you only compared patients with the same CFTR gene mutations?

Why is this important?

It may help us to predict how well a patient may do over the long term based on their sweat chloride results. Also, the new CFTR modulators cause sweat chloride values to decrease because they improve mutant CFTR function. Thus it may be possible to assess the long term impact of these medications based on the decrease in sweat chloride.

What did you do?

We used information from the CFF patient registry to group patients with identical genetic mutations. We analysed 15 groups of patients with different genotypes and looked at their sweat chloride values and how long they lived.

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What did you find?

We found that for CF patients with certain gene mutations combinations including F508del/F508del, I507del/F508del, G551D/F508del, 2789+5G>A/F508del and R117H/F508del, sweat chloride values were associated with how long patients lived, but not for other gene mutations.

What does this mean and reasons for caution?

Our results suggest that there is a greater correlation of CFTR function between sweat glands and the lung for some mutations and less for others.

What's next?

The next step is to perform studies in which we can measure CFTR function in different tissues in from the same patients. This may help us understand why there is an association between sweat chloride and death for some mutations and not others.

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cfresearchnews@gmail.com