European Cystic Fibrosis Society

Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

Cystic Fibrosis Research News

Title:

Cessation of smoke exposure improves pediatric CF outcomes: Longitudinal analysis of CF Foundation Patient Registry data

Lay Title:

Stopping exposure to smoke improves breathing and nutritional outcomes in children and adolescents with cystic fibrosis

Authors:

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What was your research question?

Exposure to tobacco smoke is a major health risk for children and adolescents with CF. While there is a lot of evidence about the harms of smoke exposure, little is known about the potential benefits of stopping the exposure. We looked at whether stopping exposure to smoke is linked to improved breathing and nutritional outcomes in children and adolescents with CF who were previously exposed to smoke.

Why is this important?

By 6 years of age, lung function in children exposed to smoke is around 5% lower than in children who haven't been exposed to smoke, and this difference persists through to the age of 18. Additionally, exposure to smoke blunts the effect of CFTR modulators: for example, among adolescents with CF, smoke exposure wiped out the treatment benefit of tezacaftor/ivacaftor (Symdeko, named Symkevi in Europe). Eliminating smoke exposure may be an important way to maximize the effect of CFTR modulators.

What did you do?

We used data from the CF Foundation Patient Registry (2006-2018). We included all people with CF born between 1998 and 2010 who had ever reported second-hand smoke exposure (daily or weekly), lived with a smoker, or smoked themselves. Every year we assessed whether people were still exposed to smoke or not. This could change, meaning a person could stop

being exposed to smoke one year, but later be exposed again. We examined how stopping exposure affects pulmonary exacerbations, lung function, and body mass index (BMI) over time. We controlled for age, sex, race/ethnicity, education, income, household size, health insurance, newborn screening, genotype, CFTR modulator use, and infections with the bacteria *Pseudomonas aeruginosa* and *Burkholderia cepacia.*

What did you find?

Stopping smoke exposure reduces the chances of having a pulmonary exacerbation by 17% in the first year and by a further 6% in each additional year of non-exposure. Stopping smoke exposure is also linked with a 0.7% increase in lung function in the first year and with a further 0.4% increase in each additional year of non-exposure. We also saw a 1% increase in BMI in the first year and an increase of 0.4% in each additional year of non-exposure. After three years of not being exposed to tobacco smoke, children and adolescents with CF have a 8% lower likelihood of a pulmonary exacerbation (Figure 1) and 2% higher lung function and BMI (Figures 2 and 3) than children and adolescents with CF who are still exposed to smoke.

What does this mean and reasons for caution?

Ending the exposure to tobacco smoke is linked to improved breathing and nutrition in children and adolescents with CF previously exposed to tobacco smoke. The effect of stopping the exposure is comparable to the effect of some CFTR modulator therapies and offsets the average annual loss of lung function due to having CF. The main limitation of the study is that the exposure to tobacco smoke was reported by the participants and not independently measured. For some people in the study, we do not know exactly how long they were exposed to tobacco smoke because the information was not collected before 2006.

What's next?

Our major finding is that children and adolescents with CF can regain nutritional and breathing losses if they are no longer exposed to tobacco smoke. This finding may be used to start a conversation about stopping smoking and as a motivator to quit smoking. Future research should identify effective strategies to stop smoke exposure in people with CF.

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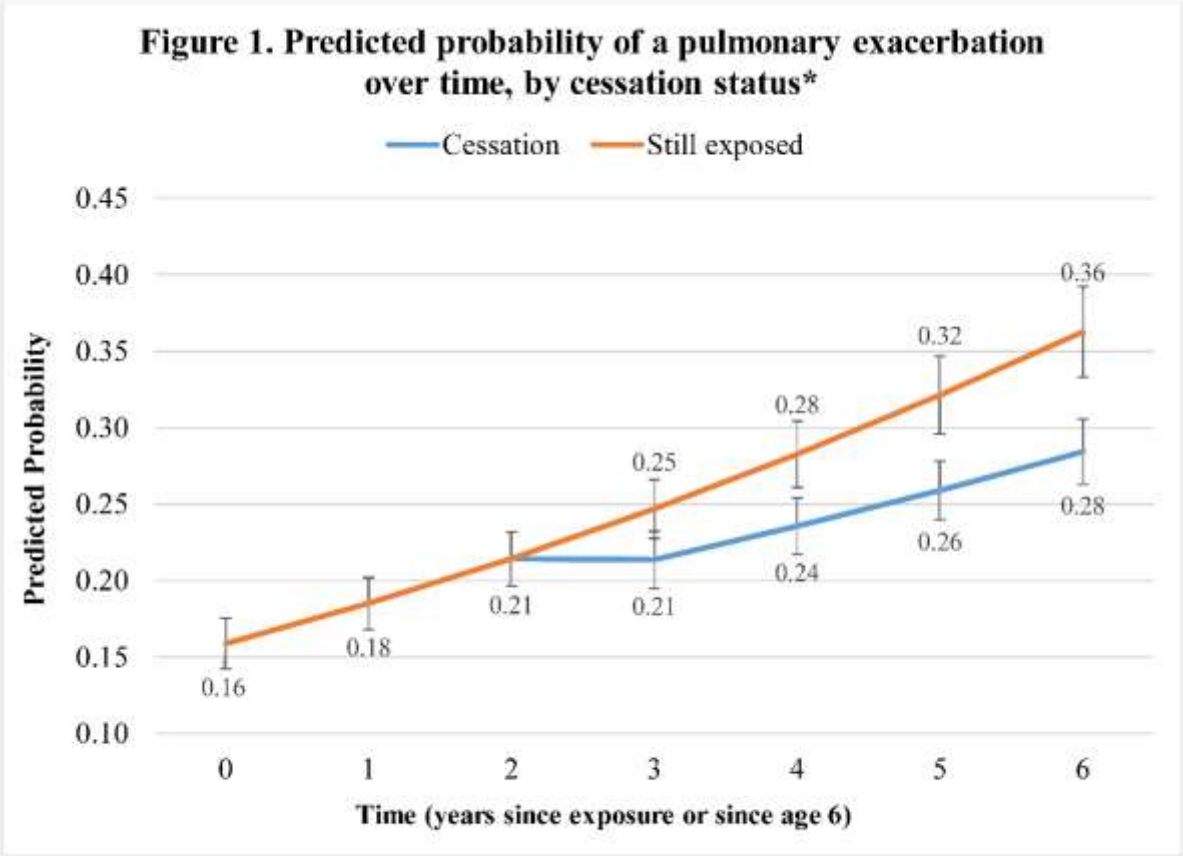

*Adjusted for age at entry, sex, race/ethnicity, household size, income, father's education, health insurance, genotype, newborn screening, CFTR modulator use, *P.aeruginosa,* and *B. cepacia*

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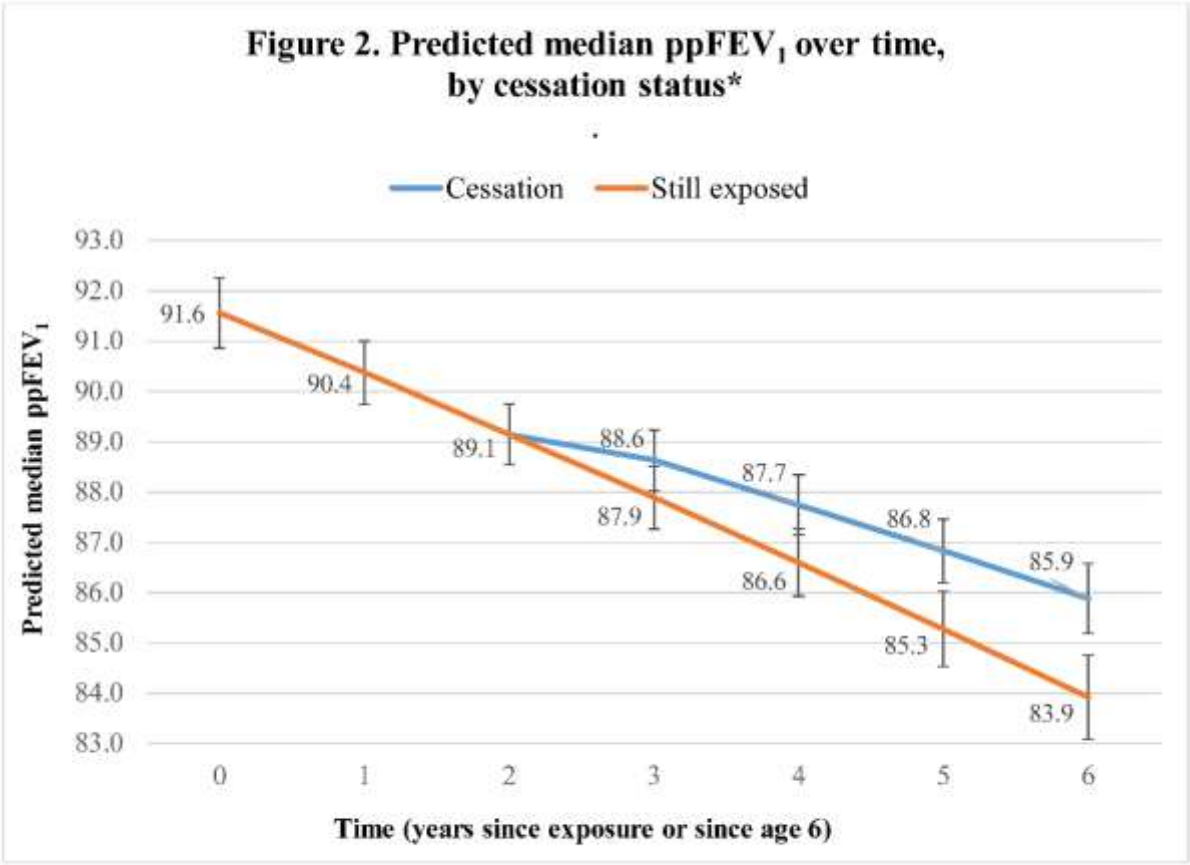

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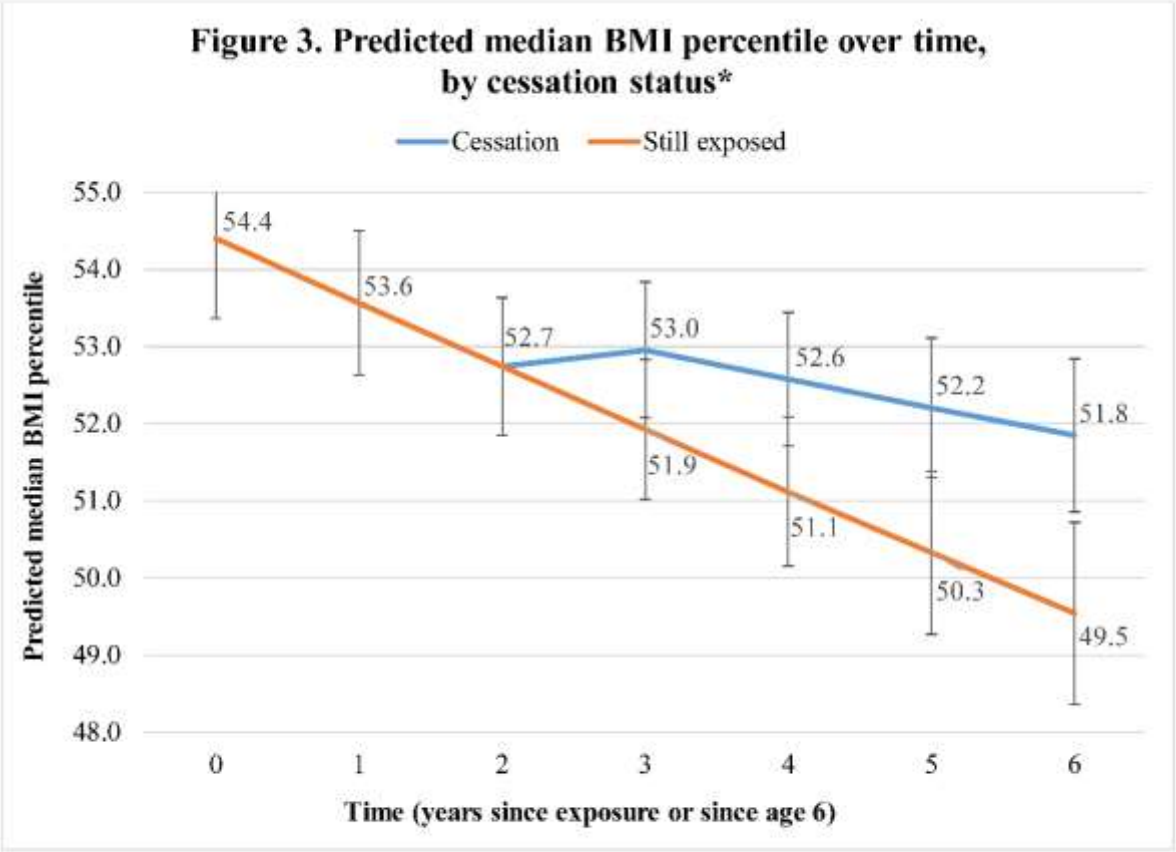

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