



Cystic Fibrosis Research News

Title:

Up-to-date and projected estimates of survival for people with cystic fibrosis using baseline characteristics: A longitudinal study using UK patient registry data

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What was your research question?

What are the expected survival chances for people with cystic fibrosis (CF) in the UK, taking into account their current age, sex, genotype, age at diagnosis, and trends towards improved survival over time?

Why is this important?

Earlier studies of survival among people with CF in the UK are out of date. This is the first study to give survival statistics based on data from the UK Cystic Fibrosis Registry, which has almost complete coverage of all people with CF the UK. The information provided is more detailed and individually relevant than the simple median survival age (the age beyond which half of people are expected to live from birth) given in annual registry reports. The results on projected survival for new generations born with CF are relevant for planning future health care resource needs and patient choices.

What did you do?

We used data from the UK CF Registry recorded between 2011 and 2015, capturing a total of 10,428 people with CF, among whom 602 people died. Survival analysis was used to obtain "survivor curves", which give estimates of the percentages of people who are expected to live

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beyond any given age starting from birth, or starting from a specified older age already reached. The statistical models included information on sex, CFTR genotype [two F508del mutations (homozygous), one F508del mutation and another mutation (F508del heterozygous), or two other mutations], and the age of diagnosis, meaning that we could present results separately according to these characteristics.

What did you find?

Here we summarize the results for people with the most common genotype (F508del homozygous). For babies born today with CF, half of males are expected to live beyond age 46 and half of females beyond age 41, assuming no further improvements in the rates of mortality. Mortality rates have reduced by around 2% per year over the past decade. If this continues, we would expect median survival ages of 65 for males and 56 for females. Given survival to age 30, half of males are expected to live beyond age 53 and half of females beyond age 49.

What does this mean and reasons for caution?

The results provide up-to-date information on survival for people with CF in the UK. Although we used the most recently available data from the UK CF Registry, the results are necessarily based on people who have lived some of their lives under past standards of care and therefore cannot fully reflect modern standards of care. We made forecasts of what survival might look like for CF in the future if there are continued improvements in mortality rates. These are speculative as we do not yet know what the impact of new and future disease-modifying treatments could be.

What's next?

This work accounted for three individual characteristics. In forthcoming work, we are providing personalised information about survival based on patient characteristics measured over time, such as lung function. It is important that patients and care-givers are supported to interpret survival estimates and the uncertainty around those estimates.

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