Cystic fibrosis patients survive longer in EU compared with non-EU countries

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The largest multinational study of patients with cystic fibrosis (CF) has shown that patients from EU countries (membership as of 2003) survive longer than those in neighbouring non-EU countries. Crucially, the data show that there are disproportionately fewer then expected CF patients in non-EU countries. The data suggest that had these non-EU countries displayed similar demographic profiles to their EU neighbours, their own CF patient populations would rise by an estimated 84%. These findings are reported in an Article in this week's edition of the *Lancet*, written by Dr Anil Mehta and Dr Jonathan McCormick and colleagues at the University of Dundee, UK, together with European colleagues.

Cystic fibrosis occurs unexpectedly in babies of apparently healthy parents who are carriers of the defective cystic fibrosis transmembrane-conductance regulator gene (CFTR), and is one of the most common inherited disorders in populations of European descent.

This study combined demographic indicators—age, age at diagnosis, sex, and genotype—for patients with CF from 35 European countries into a disease register to establish any differences between EU and non-EU countries, many of whom are today full members of the expanded EU. The authors used their data to estimate the size of the CF population across Europe gathering data from 29 025 patients. The median age was 16.3 years with a striking difference of 4.9 years between the EU (17.0 years) and non-EU countries (12.1 years).The proportion of patients older than 40 years was also significantly higher in EU countries (5%) than in non-EU countries (2%).The authors estimated that the CF

population in non-EU countries would increase by 84% if their CF patients had a demographic profile comparable to the EU countries.

The authors say: "We have shown that far fewer children and young adults have cystic fibrosis in non-EU countries than we expected. This finding is reinforced by the increased chance of patients surviving to 40 years in EU countries, even if they have the [most common and severe form of CF] mutation."

They add: "The difference between the number of patients with cystic fibrosis between EU and non-EU countries is striking in view of the similarity of general population sizes and the expected disease prevalence [from genetic calculations]. This disparity in demographic indicators might be due to reduced availability of specialist drugs, equipment, and trained multidisciplinary staff in non-EU countries, rather than lower gene frequency, greater disease severity, or poorer treatment adherence than in EU countries."

The authors note that diagnoses of children younger than 1 year are scarce in non-EU countries, which could be caused by insufficient data on disease incidence, but is more probably a result of deaths due to unrecognised CF which used to happen commonly in the EU in the 1970s. Furthermore, they estimate that an extra US$7.8 million per year (£4.9 million) would be needed to care for the additional 3212 patients that they predicted would be alive in non-EU countries if these patients had the demographic indicators of EU countries. This estimate is based on the median cost of care for UK patients with cystic fibrosis aged 1□□ years of $2442 (£1526) at 2002 prices, which was previously calculated for clinically diagnosed patients from birth to age 9 years in an earlier Lancet paper (Sims et al 2007)*.

As well as unrecognised CF cases, misclassification of infant deaths due to CF, and inequality of access to health care leading to early death,

could be responsible for the profile of CF in non-EU countries. The authors conclude: "In our study, both factors are likely to affect new members of the EU, and these matters need urgent attention by governments."

More information: *Sims EJ, Mugford M, Clark A, Aitken D, McCormick J, Mehta G, Mehta A (2007) Economic implications of newborn screening for cystic fibrosis: a cost of illness retrospective cohort study. Lancet 369:1187-1195.

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