Improved survival albeit with persistent disparities in prognosis for people with cystic fibrosis in European countries

Clémence Martin [1,2,3] **and Pierre-Régis Burgel** [1,2,3]

[1]Université de Paris, Institut Cochin, Inserm, Paris, France. [2]Respiratory Medicine and Cystic Fibrosis National Reference Center; Cochin Hospital; Assistance Publique Hôpitaux de Paris (AP-HP), Paris, France. [3]ERN-Lung CF network.

Corresponding author: Pierre-Régis Burgel (pierre-regis.burgel@aphp.fr)

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Cystic fibrosis (CF), an autosomal recessive disease related to mutations in the gene encoding for the cystic fibrosis transmembrane conductance regulator (CFTR) protein, is the most prevalent severe genetic disease in Caucasian populations [1]. Although the disease affects multiple organs, respiratory disease is the major manifestation of CF and is often responsible for premature death [2], which often occurred in the first years of life before the implementation of modern CF care [3]. Over the past decades, progress in therapeutic management has resulted in a dramatic improvement in the prognosis of people with CF [4]. In a study published more than 20 years ago, Fogarty *et al*. [5] compared median age at death in seven countries (USA, and Australasian and European countries) and reported that median age at death improved from 8 years in 1974 to 21 years in 1994, albeit with marked differences between countries. More recent studies have confirmed improvement in median age of survival, which was 40.6 years in US patients and 50.9 years in Canadian patients, in a study that used data from between 2009 and 2013 [6]. In European countries, contemporary studies have reported increases in the number of adults with CF [7, 8], with a better prognosis in countries belonging to the European Union (EU) than in those outside of the EU [7, 8], mostly located in Eastern Europe, in which access to appropriate CF care is less available [9]. Collectively these data suggest that prognosis improvement in patients with CF is heavily dependent on the quality of the CF healthcare system.