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Letter to the Editor

Clinical follow-up of 122 Italian cystic fibrosis patients with *B. cepacia* complex colonisation☆

Sir,

We read with great interest the article by Courtney regarding the clinical outcome of *B. cepacia* complex infection in cystic fibrosis (CF) adults [1]. As the author states, complex factors, regarding both the pathogen and the patient, can influence the clinical outcome [1–6]. The Courtney's study is limited by the small number of patients colonised by *B. multivorans* (3) and *B. cenocepacia* (16), but emphasis is given to genomovar status and clonality in conditioning survival, decreased pulmonary function and nutritional status [1].

We are currently engaged in a multi-centre study in Italy regarding clinical outcome and follow-up of Italian CF patients with *B. cepacia* complex infection and we wish to report our preliminary results here. The goal of our study was to investigate the clinical evolution of a cohort of 122 Italian CF patients colonised by *B. cepacia* complex through questionnaires sent to all Italian CF Centres. Age, sex, concomitant respiratory colonisation, lung function over time and mortality were assessed.

Fourteen out of 22 Italian CF Centres participated in the study. Data on 122 patients (52 males, 70 females) were collected. 26 patients (21.3%) were ΔF508 homozygote, 59 (48.4%) were compound heterozygotes (ΔF508/other), 13 (10.7%) were other/other, 4 (3.2%) were unk/unk and 20 (16.4%) were missing.

Courtney's paper did not refer to concomitant colonisation in the selected study groups. Concomitant colonisation by more than one pathogen occurs frequently in CF patients, in those infected by *B. cepacia* as well as by other bacteria such as *P. aeruginosa* [7].

In our cases, 40 (32.7%) patients were colonised by *B. cepacia* complex alone, with 90 (77.5%) being chronically colonised, 26 (22.5%) transiently and 4 missing. The mean age of patients at colonisation was 16.1 years (±8.3, median 15, range 2–41,). The mean change in FEV1 (percent change compared to baseline) 2 years after *B. cepacia* complex colonisation was −9.8% (±22.2) in chronically colonised and −4.93 (±9.6) in intermittently colonised. Three years after *B. cepacia* complex colonisation, 14.4% of our chronically colonised patients died (missing data: 2), whereas no deaths occurred in transiently colonised patients (Fisher test p=0.03). Two out of three transplanted patients died.

Genomovar status was determined in 56/122 (45.9%) of isolates. Thirty patients (53.6%) were infected with *B. cenocepacia*, 6 (10.7%) with *B. cepacia*, 6 (10.7%) with *B. pyrrocinia*, 5 (8.9%) with *B. multivorans* and 9 (16.1%) with other genomovars. The clinical course of patients infected with *B. cenocepacia* or *B. multivorans* is usually associated with an increase in patient morbidity and mortality compared to those infected with other genomovars, which is why we compared mortality and FEV1 decline of patients infected by different genomovars.

During the study period, 7/16 (43%) of the patients in Courtney's study, who were infected by a single *B. cenocepacia* strain (IIIA, ET12), died [1]. In our study, mortality 5 years after infection was 20.5% for patients infected with *B. cenocepacia* or *B. multivorans*, versus 5.5% for patients colonised by other genomovars.

Studies on *B. cepacia* complex clonality are ongoing in Italy. The mean change in FEV1 (percent change compared to baseline) 2 years after colonisation was −17.6% (±21.8) in patients with *B. cenocepacia* or *B. multivorans*, and −3.02% (±19.2) in those colonised by other genomovars (*t*-test, p=0.03).

On the basis of our experience, 3-year survival is worse in patients with chronic *B. cepacia* complex colonisation compared to those colonised transiently, and independently of the clonality, the mean decrease in FEV1 is significantly higher in patients with *B. cenocepacia* or *B. multivorans* than in those with other genomovars.

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References

[1] Courtney JM, Dunbar KEA, McDowell A, et al. Clinical outcome of *Burkholderia cepacia* complex infection in cystic fibrosis adults. J Cyst Fibros 2004;3:93–8.

[2] Frangolias DD, Mahenthiralingam E, Rae S, et al. *Burkholderia cepacia* in cystic fibrosis: variable disease course. Am J Respir Crit Care Med 1999;160:1572–7.

[3] Ledson MJ, Gallegher MJ, Jackson M. Outcome of *Burkholderia cepacia* colonisation in an adult cystic fibrosis center. Thorax 2002; 57:142–5.

[4] Manno G, Dalmastri C, Tabacchoni S, et al. Epidemiology and clinical course of *Burkholderia cepacia* complex infections, particularly those caused by different *Burkholderia cenocepacia* strains, among patients attending an Italian Cystic Fibrosis Center. J Clin Microbiol 2004; 42:1491–7.

[5] De Soyza A, McDowell A, Archer L, et al. *Burkholderia cepacia* complex genomovars and pulmonary transplantation outcomes in patients with cystic fibrosis. Lancet 2001;35:1780–1.

[6] Muhdi K, Edenborough FP, Gumery L, et al. Outcome for patients colonised with *Burkholderia cepacia* in a Birmingham adult cystic fibrosis clinic and the end of an epidemic. Thorax 1996;51:374–7.

[7] Cystic Fibrosis Foundation Patient Registry Foundation, Patient Registry 2003 Annual Report. Bethesda, Maryland.

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