Cystic fibrosis (CF) is the major genetic disease in the European Caucasian population and by the number of patients the third orphan disease (at least in France). The disease is due to mutations in the gene CFTR (cystic fibrosis transmembrane conductance regulator) which encodes for a chloride channel involved in electrolytic exchanges through the plasma membrane. Mutations in this gene result in a defect in the mucociliary clearance and therefore in a thickness of the bronchial mucus, and these abnormal airway conditions facilitate the entrapment of the airborne bacteria and fungal spores, and provide a suitable environment for the growth of these microorganisms.

Beside bacteria like Staphylococcus aureus, Pseudomonas aeruginosa and Burkholderia cepacia, which remain the major causative agents of respiratory infections in the context of CF and which often determine the mortality of the patients, several filamentous fungi may also colonize the respiratory tract of these patients. This fungal colonization of the airways, facilitated by the frequent and prolonged cures of antibiotics and the use of corticosteroids, may also lead to true respiratory infections whose frequency regularly increases along with the development of lung transplantation and the increase in life expectancy.

Among these filamentous fungi, Aspergillus fumigatus remains by far the most common agent of airway colonization. However, other fungal species are increasingly reported, such as Scedosporium apiospermum, Aspergillus terreus, Exophiala dermatitidis and Scedosporium prolificans. But the situation may be even more complex, as suggested by the recent description by our group of two additional species from sputum samples of patients with CF: Penicillium emersonii and Acrophialophora fusispora.

Apart from Aspergillus fumigatus which is well known by clinicians and microbiologists involved in the follow-up of CF patients, the prevalence of these fungi in the context of CF is certainly underestimated (in relation with the lack of standardization of the mycological examination of clinical samples, and with the poor knowledge of some of these fungi), and their clinical significance still remains to be defined. In case of chronic colonization of the airways, these fungi should contribute to the inflammatory reaction which progressively leads to the deterioration of the lung function. Moreover, due to their propensity to disseminate and to their poor susceptibility to current antifungals, these fungi may also cause severe, and sometimes fatal, disseminated infections in immunocompromised hosts, such as lung transplant recipients.

Numerous questions therefore raise from the colonization of the airways by these filamentous fungi which are rarely found in the environment, and basic research on the ecology of these fungi, their biochemistry, and their pathogenic mechanisms should be promoted, in order to define prophylactic measures or to develop more efficient antifungal drugs.

So we would be interested in an international network on fungal respiratory infections in cystic fibrosis. This network would comprise medical mycologists involved in the clinical surveillance of CF patients, but also scientists interested in basic research on the filamentous fungi associated with CF. Additionally, this network would take advantage of and form a bridge between existing ISHAM working groups (on Pseudallescheria infections, Black yeasts, non-fumigatus Aspergillus, and PCR-detection of A. fumigatus). Working together, it would be possible to improve our knowledge on the epidemiology, the physiopathology and clinical relevance of the colonization of the airways by filamentous fungi in patients with CF.

Convener and contact:

Dr. Jean-Philippe Bouchara
Head of the Host-Parasite Interaction Study Group (UPRES-EA 3142) of Angers University
Head of the Laboratory of Parasitology-Mycology of Angers University Hospital