Chronic Pseudomonas aeruginosa infection and respiratory muscle impairment in cystic fibrosis

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Chronic infection with Pseudomonas aeruginosa (Pa) in patients with Cystic Fibrosis (CF) is associated with increased morbidity and mortality. Respiratory muscle function can be assessed by maximal inspiratory pressure (Pimax), maximum expiratory pressure (Pmax), and pressure-time index of the respiratory muscles (PTImus). Objectives: To compare respiratory muscle function by measurement of Pmax, Pmax and PTImus between CF patients at different stages of Pa infection in a large cohort comprising of children and young adults.

Methods: We investigated the differences in maximal respiratory pressures, PTImus and anthropometric indices in different stages of Pa infection according to the Leeds criteria (never infected, free of infection, intermittent infection, chronic infection). FEV1, FVC, MEF25−75, Body Mass Index, Upper Arm Muscle Area (UAMA), PTImus, Pmax and PTImus were assessed in 122 CF patients who formed four groups matched for age and gender according to the different stages of Pa infection.

Results: Median PTImus was significantly lower in CF patients with chronic Pa infection compared to patients that were never infected, free of infection or intermittently infected. Median PTImus was significantly increased in CF patients with chronic Pa infection compared to patients that were free of infection. Multivariate logistic regression revealed no interaction between Pmax, FEV1 and age as a significant determinant of PTImus.

Conclusion: CF patients with chronic Pa infection exhibit impaired respiratory muscle function and decreased inspiratory muscle strength. Chronic Pa infection might independently impact on respiratory muscle function in patients with CF.

Determinants of nontuberculous mycobacteria eradication in cystic fibrosis

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Objectives: The prevalence of colonization with nontuberculous mycobacteria (NTM) in cystic fibrosis (CF) is increasing. The aim of this retrospective, multicenter study was to characterize the factors associated with NTM eradication in infected or colonized CF patients.

Methods: All CF patients in whom NTM had been identified in at least two sputum cultures were included. The infection caused by NTM was considered as a disease when CF clinicians decided the appropriate therapeutic management. Univariate and multivariate analyses were performed to identify risk factors.

Results: Fifty patients were included. The most frequently identified NTM were Mycobacterium avium complex (56%) and Mycobacterium xenopi (34%). Twenty-eight patients were infected and treated; 22 patients were colonized. The decline in FEV1 between the time of NTM identification and two years later was higher in infected-treated than in colonized patients. The presence of Aspergillus fumigatus in sputum was associated with NTM infection. The previous administration of azithromycin and/or PPIs was associated with microbiological cure, regardless of the type of therapeutic management.

Conclusion: Our results suggest that the identification of Aspergillus fumigatus in patients with CF should particularly lead to suspect NTM lung disease. Previous antithymocin treatment did not prevent NTM eradication or could have a primary prophylactic effect and promote microbiological cure. A better assessment of NTM infection and a better knowledge of the predictive criteria of eradication should improve the management of CF patients.

Pseudomonas aeruginosa acquisition in CF patients in the context of otorhinolaryngological surgery or dentist attendance – Case series and discussion of preventive concepts

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Objectives: P. aeruginosa is the primary cause for pulmonary destruction and premature death in CF patients. Therefore it is of highest interest to prevent airway colonization with pathogens that is ubiquitously present in water. Whereas it is frequently detected in dental unit waterlines with proven transmission to CF patients, risks of acquisition in otorhinolaryngological or dentistry treatment have not yet been discussed although some perils could be comparable.

We present five CF patients who primarily acquired P. aeruginosa around otorhinolaryngological surgery (4) or dentist treatment (1). Additionally, we discuss risks and preventive strategies for CF patients undergoing otorhinolaryngological treatment. Altogether, risks range from contact to pathogen carriers in waiting rooms to instrumentation, suction, drilling or flushing fluid in oral and sinonasal areas, when droplets containing pathogens can be nebulized into CF airways. Post-surgically mucosal damage impairs sinonasal mucociliary clearance, which additionally to debridement and crusting facilitates pathogens to clinch and proliferate. Thus, otorhinolaryngological surgery in CF must be linked to repeated assessment of pathogen colonization, together with cleansing, anti-inflammatory and antibiotic therapy. Further studies must elaborate, whether all operated CF patients, independent from pre-surgical colonization status, would routinely require antipseudomonal treatment, including nasal lavages containing antibiotics as presented in a series of protocols. Altogether, we must bring up attention on this probably underestimated risk within CF therapy and postulate structured prevention protocols, available to all patients.

Invasive pulmonary mycosis caused by Scedosporium and Pseudallescheria spp. in patients with CF. Diagnostic and treatment guidelines

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Objectives: Chronic airway infections are a major cause of morbidity and mortality in patients with CF. The role of bacteria in acute bronchopulmonary exacerbation events is well known. Cases describing fungal involvement in pulmonary infections are known for ABPA and in patients who are immunocompromised, such as lung transplant recipients. Invasive pulmonary mycosis in immunocompetent CF patients has rarely been described. Diagnostic procedures and treatment guidelines are missing.

Methods: In this study, from 12 CF centers in Germany 17 out of approximately 1200 patients with a high suspicion of an infection with a Scedosporium or Pseudallescheria spp. were evaluated. Criteria for invasive fungal infection were the following: increased sputum production, multiple pathogen detection in sputum or lavage samples, new pulmonary infiltrates, treatment failure with antibiotic therapy, unclear FEV1 decrease and specific antibody detection. Sputum and lavage samples were cultivated on Scel+ agar. Isolates were identified by sequencing of the ITS reagions of ribosomal DNA genes, antifungal susceptibility testing was performed using the microdilution reference method CSLI.

Conclusion: Pulmonary infection by Scedosporium or Pseudallescheria spp. can also occur in immunocompetent patients with CF. For microbiological detection of fungi, Scel+ agar or an adequate selective culture medium may be useful. To confirm an invasive fungal infection, the detection of specific antibodies can be helpful. The therapy should be done with two systemic and one inhaled antifungal drug for at least 4 weeks. In some cases a much longer therapy is required.