

182 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 (P_imax), maximal expiratory pressure (P_emax) and pressure-time index of the respiratory muscles (PTI_{mus}). **Objectives:** To compare respiratory muscle function by measurement of P_imax, P_emax and PTI_{mus} 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, PTI_{mus} 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), P_imax, P_emax and PTI_{mus} 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 P_imax was significantly lower in CF patients with chronic Pa infection compared to patients that were never infected, free of infection or intermittently infected. Median PTI_{mus} was significantly increased in CF patients with chronic Pa infection compared to patients that were free of infection. Multivariate logistic regression revealed state of infection as a significant determinant of PTI_{mus} independently of UAMA and FEV₁.

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.

183 *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 the pathogen that is ubiquitously present in water. Whereas it is frequently detected in dental unit waterlines with proven transmission to CF patients, risks of acquisition within otorhinolaryngological 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 otorhinological 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. Postsurgically mucosal damage impairs sinonasal mucociliary clearance, which additionally to debridement and crusts facilitates pathogens to cling 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.

184 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 abscessus* (56%) and *Mycobacterium avium* complex (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 azithromycin 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.

185 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 ScSel + agar. Isolates were identified by sequencing of the ITS regions 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, ScSel + 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.