A longitudinal study of lung bacterial pathogens in patients with primary ciliary dyskinesia

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In patients with primary ciliary dyskinesia (PCD), impaired mucociliary clearance leads to an accumulation of secretions in the airways and susceptibility to repeated bacterial infections. The primary aim of this study was to investigate the bacterial flora in non-chronic and chronic infections in the lower airways of patients with PCD. We retrospectively reviewed the presence of bacteria from patients with PCD during an 11-year period and genotyped 35 Pseudomonas aeruginosa isolates from 12 patients with chronic infection using pulsed-field gel electrophoresis. We identified 5450 evaluable cultures from 107 patients with PCD (median age 17 years, range 0–74 years) (median age at diagnosis 7.8 years, range 0–63 years). Haemophilus influenzae was the most frequent microorganism. Other common pathogens were P. aeruginosa, Streptococcus pneumoniae, Moraxella catarrhalis and Staphylococcus aureus. The number of patients colonized with P. aeruginosa at least once varied from 11 to 44 patients (15–47%) annually, and 42 patients (39%) met the criteria for chronic infection at least once. Pseudomonas aeruginosa was more frequently isolated in teenagers and adults than children (p 0.02) and the prevalence was significantly lower in patients with preschool (<6 years) PCD diagnosis (p 0.04). Ten out of 12 patients (83%) were chronically infected with a unique clone-type of P. aeruginosa. No sharing of clone-types or patient-to-patient transmission was observed. In conclusion, PCD patients were infected by a unique set of bacteria acquired in an age-dependent sequence. Pseudomonas aeruginosa frequently colonizes the lower respiratory tract and the incidence of chronic infection was higher than previously reported.

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