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Cystic fibrosis: disease monitoring, biomarkers, and psychosocial issues

Paediatric cystic fibrosis (CF)

PA-70282 - Evaluation of respiratory function at 6 years of age in patients with cystic fibrosis with frequent pulmonary exacerbations in the first 2 years of life

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Objectives-aim: Pulmonary exacerbations (PE) results in a progressive decline in pulmonary function in patients with cystic fibrosis (CF). We aimed to evaluate the effects of acute PE attacks in infancy on pulmonary functions at 6 years in CF patients.

Methods: This is a retrospective cohort study. Eligible patients were included in theTurkish Cystic Fibrosis Registry of Turkey (CFRT)-2019 data. Patients who had 2 or less PE in the first 2 years of age were defined as Group1, and those

more than 2 were defined as Group 2.

Results: 110 patients were included in the study, 57 female, 22 patients' PE attack information is unknown, 59 of the patients were in Group 1.The mean number of PE in the first 2 years of age was 2.2 (±2.38). There were no significant differences between the groups according to IV antibiotic needs, positive bacterial culture results, and CFTR mutations' distribution during the first PE. The mean of percent predictive FEV1 and FVC were significantly lower in Group2 than in Group1 (p:0.029; 0.032 res.). There were no significant differences in BMI between the groups. There were no correlations between the first PE's age and spirometry indices and the number of hospitalizations. The current BMI of the patients who have chronic colonization is significantly lower than their first PE's BMI value (p: 0.028).

Conclusion: Patients with more than 2 PE had worse pulmonary function than 2 or less. Frequent PE at an early age and bacterial colonization should be prevented to preserve the lung functions of CF patients.

Keywords: Cystic fibrosis, Spirometry, Exacerbation