DOI: 10.5152/TurkThoracJ.2019.56

[Abstract:0244] MS-080 [Accepted: Oral Presentation] [Pediatric Lung Diseases]

Pseudo Bartter Syndrome: The Most Common Complication in Turkish National Cystic Fibrosis Registry

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Objectives: Pseudo bartter syndrome (PBS) is a known complication of cystic fibrosis (CF) which is usually seen in infancy and warm weather conditions. First data of Turkish National CF Registry was documented and PBS was found as the most common complication in the patients. We aimed to investigate the clinical features of CF patients with PBS.

Methods: Data of CF patients with PBS evaluated in terms of demographics, mutation analysis, pulmonary function tests, colonisation and other complications.

Results: Totally 1170 CF patients were included into registry in 2017 and 120 of them (10%) had PBS. Seventy one (59%) of them were male and 49 of them were girl. Mean age of diagnosis was 0.73±1.67 years (min:0.08; max:11). Five of them had meconium ileus history and 110 of them had pancreatic insufficiency. Chronic liver disease accompanied in 5 patients and diabetes in 1 patient. Thirty three of the patients with PBS had Staphylococcus aureus, 23 Pseudomonas aeruginosa, 4 Stenotrophomonas maltophilia colonisation. Forty five different mutations were detected in 65 patients in 120 alleles. The most common mutation was DF508 in 21 alleles and it was homozygous in 5 (4%) patients. N1303K, D110H, G542X and E92K were the other common mutations, respectively. The most common mutations were class 1 and 2 mutations.

Conclusion: Young age of our patients and warm climate of our country may cause the pseudo-bartter as the most common complication and severe mutations may predispose it.

Keywords: Cystic fibrosis, Pseudo Bartter Syndrome, Turkish National Cystic Fibrosis Registry