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# Clinical Features of Cystic Fibrosis Patients with Chronic Liver Disease in Turkish National Cystic Fibrosis Registry

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**Objectives:** We aimed to investigate the clinical features of cystic fibrosis (CF) patients with chronic liver disease (CLD) in Turkish National CF Registry.

**Methods:** All data of the CF patients with CLD who were recorded into the registry in 2017 were evaluated. Demographics, type of liver disease, mutation analysis, pulmonary function test results, colonisation and other complication results were noted.

**Results:** Totally 1170 CF patients were included into registry in 2017. Mean age of diagnosis was 1.69 years and their current age was 7.3 years,% 46 of them were girl and% 54 boy. Ninty three (8%) of them had CLD. Mean age of the diagnosis of patients with CLD was 1,68 years and 55 of them (59%) were boy. All of them had pancreatic insufficiency and 10 of them had meconium ileus history. 81 of them (87%) had CLD without cirrhosis, 6 of them had cirrhosis with portal hypertension, 6 of them had cirrhosis without portal hypertension. Thirty of the patients had Pseudomonas aeruginosa, 42 Staphylococcus aureus, 2 Burkholderia cepacia, 3 Stenotrophomonas maltophilia, 1 non tuberculosis mycobacteria colonisation. ABPA accompanied in 4 patients and diabetes in 12 patients. Forty nine patients performed pulmonary function tests and mean FEV1 was 80.1% and mean FVC was 79.5%. Thirty three different mutations were detected in 67 patients in 120 alleles. The most common mutation was DF508 in 41 alleles and it was homozygous in 14 (15%) patients. G85E, G542X, 2183AA->G, 2789+5G>A and 1677delTA were the other common mutations respectively. The most common mutations were class 1 and 2 mutations. Six of these patients were died in the follow up.

**Conclusion:** Chronic liver disease is an important complication of CF. Most of these patients had severe mutations and complications. Early detection and treatment is essential in these patients. With data of registries we hope to improve our knowledge about CLD in CF patients.

**Keywords:** Chronic liver disease, cystic fibrosis, Turkish National Cystic Fibrosis Registry