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Fibrosis related Liver Disease (CFLD). Ursodeoxycholic acid (UDCA) is recommended for management of abnormal liver function and or abnormal liver US, with referral to specialist liver services for persistently abnormal investigations.

Methods: 3 year retrospective audit of practice against UK National Institute of Health & Care Excellence (NICE) and European CF Society guidelines. Review of biochemistry, radiology and clinic documentation in patients >1 year of age with cystic fibrosis.

Results: 209 annual reviews were completed in 78 patients between 2016 and 2018. 15 patients had <3 annual reviews due to age or transfer into clinic. One patient died. 46/78 (59%) were female. Median BMI centile was 60.

LFTs were performed in 196/209 (93.7%) of reviews, 44 (22.4%) were abnormal. 200/209 (96.6%) reviews included liver US, 32 scans (16%) were abnormal.

59/209 (28.2%) of reviews showed abnormal liver investigations but UDCA was only prescribed in 40/209 (19.1%). A previous study in the same clinic in 2002 showed use of UDCA in 29.9% of patients demonstrating a reduction in overall use of UDCA.

2 patients with hepatomegaly or splenomegaly and abnormal LFTs were not referred to specialist service. 8 patients had persistently abnormal LFTs despite UDCA but only one was referred to specialist service. No patient had evidence of liver failure, portal hypertension or haematemesis.

Conclusion: In a regional paediatric service, >92% of investigations to identify CFLD were completed as per guidelines. Use of UDCA was less than expected compared to abnormal investigations and has declined since 2002. There was inadequate referral to specialist services when investigations remained abnormal despite treatment.

These findings highlight the need for centres to complete regular audit to review investigation and management of CFLD.

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Clinical features of cystic fibrosis patients with chronic liver disease in the Turkish National Cystic Fibrosis Registry

T. Sismanlar Eyuboglu[1], D. Dogru Ersoz[2], E. Cakır[3], N. Cobanoglu[4], S. Pekcan[5], G. Cinel[6], E. Yalcın[2], N. Kiper[2], V. Sen[7], H. Selimoglu Sen[8], O. Ercan[5], O. Keskin[9], S. Bilgic Eltan[10], L. Muhammed Al Shadfan[3], H. Yazan[3], D.U. Altıntas[11], S. Sasihuseyinoglu[11], N. Sapan[12], S. Cekic[12], H. Cokugras[13], A.A. Kılınc[13], T. Ramaslı Gursoy[14], A.T. Aslan[14], A. Bingol[15], A.E. Basaran[15], A. Ozdemir[16], M. Kose[17], M. Hangul[17], N. Emiralioglu[18], G. Tugcu[6], H. Yuksel[19], O. Yılmaz[19], F. Orhan[20], Z.G. Gayretli Aydın[21], E. Topal[22], Z. Tamay[23], A. Suleyman[23], D. Can[24], C.M. Bal[25], G. Caltepe[26], U. Ozcelik[2]. *[1]Dr Sami Ulus Maternity and Children Training and Research Hospital, Pediatric Pulmonology, Ankara, Turkey; [2]Hacettepe University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [3]Bezmialem University Faculty of Medicine, Pediatric Pulmonology, Istanbul, Turkey; [4]Ankara University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [5]Necmettin Erbakan University Meram Medicine Faculty, Pediatric Pulmonology, Konya, Turkey; [6]Ankara Children's Hematoloji Oncology Training and Research Hospital, Pediatric Pulmonology, Ankara, Turkey; [7]Dicle University Faculty of Medicine, Pediatric Pulmonology, Diyarbakır, Turkey; [8]Dicle University Faculty of Medicine, Pulmonology, Diyarbakır, Turkey; [9]Gaziantep University Faculty of Medicine, Pediatric Allergy, Gaziantep, Turkey; [10]Gaziantep University Faculty of Medicine, Pediatric Pulmonology, Gaziantep, Turkey; [11]Çukurova University Faculty of Medicine, Pediatric Allergy and Immunology, Adana, Turkey; [12]Uludag University Faculty of Medicine, Pediatric Allergy and Immunology, Bursa, Turkey; [13]Istanbul University Cerrahpasa Medicine Faculty, Pediatric Allergy and Pulmonology, Istanbul, Turkey; [14]Gazi University Faculty of Medicine, Pediatric Pulmonology, Ankara, Turkey; [15]Akdeniz University Faculty of Medicine, Pediatric Pulmonology, Allergy and Immunology, Antalya, Turkey; [16]Mersin Maternity and Children Hospital, Pediatric Pulmonology, Mersin, Turkey; [17]Erciyes University Faculty of Medicine, Pediatric Pulmonology, Kayseri, Turkey; [18]Gaziantep Cengiz Gökçek Maternity and Children Hospital, Pediatric Pulmonology, Gaziantep, Turkey; [19]Celal Bayar University Faculty of Medicine, Pediatric Pulmonology, Allergy and Immunology, Manisa, Turkey; [20]Karadeniz Technical University Faculty of Medicine, Pediatric Allergy, Trabzon, Turkey; [21]Karadeniz Technical University Faculty of Medicine, Pediatric Infectious Disease, Trabzon, Turkey; [22]Inonu University Faculty of Medicine, Pediatric Allergy, Malatya, Turkey; [23]Istanbul University İstanbul Medicine Faculty, Pediatric Allergy, Istanbul, Turkey; [24]Balıkesir University Faculty of Medicine, Pediatric Pulmonology, Balıkesir, Turkey; [25]Ataturk Regional Training and Research Hospital, Pediatric Pulmonology, Erzurum, Turkey; [26]Ondokuz Mayıs University Faculty of Medicine, Pediatric Gastroenterology, Samsun, Turkey*

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

Method: 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.

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Long-term follow-up of liver disease in children and young people with cystic fibrosis in the UK

L. Modin[1,2], C. Lloyd[1], I. Van Mourik[1], M. Desai[3], D. Kelly[1]. *[1]Birmingham Children's Hospital, Liver Department, Birmingham, United Kingdom; [2]Hans Christian Andersen Children's Hospital, Paediatric Department, Odense, Denmark; [3]Birmingham Children's Hospital, Respiratory Department, Birmingham, United Kingdom*

Objectives: The aim of the study was to describe the natural history and long-term efficacy of UDCA in patients diagnosed with CFLD.

Methods: Retrospective review of patients (0–18 years) diagnosed with CF referred to the Liver Unit at Birmingham Children's Hospital for assessment of CFLD. Diagnosis of CFLD based on: persistently raised transaminases, liver ultrasound scan (USS); endoscopic findings of portal hypertension; histological change on liver biopsy in selected cases. Demographic data and follow-up data on progression of liver disease were collected.

Results: 213 (123 males) were included. Diagnosis of CFLD was based on persistently raised transaminases in 111 (52%); portal hypertension (PHT) with splenomegaly in 102 (48%). 45/213 (21%) started on UDCA before referral, 88 (41%) at referral, and 80 (38%) >6 months after referral. Progression of liver disease reported in 80 (38%): 14 (17%) increased spleen, 7 (9%) on banding programme (no banding), 25 (31%) on banding programme (including banding), 5 (6%) cirrhosis; 29 (36%) had a liver transplant. There was no difference in mean follow-up time between the 80 patients with progressive liver disease (16 years, SD 3.3) and for the 133 stable patients (16 years, SD 2.5). 31/111 (28%) children with raised transaminises progressed while 49/102 (48%) with PHT progressed, $p < 0.001$.