Echography changes in the liver were found in 54 (50.9%) patients at the same time with abnormal clinical assessment and/or altered liver enzymes, the mean age was 10.04 years, 29 females and 26 males. Cirrhosis was found in 2 (1.89%) patients. 56 (52.8%) patients, were receiving Ursodeoxycholic Acid (UDCA) as therapy, 2 (1.89%) only because of the persistently altered liver enzymes.
The genetic mutations among CFLD patients were as follows: 40 (74.1%) patients had homozygous F508del, 8 (14.8%) patients had heterozygous F508del, 3 (5.55%) patients had other mutations, in 3 (5.55%) patients genetic examination was not performed. The pancreatic insufficiency was present in 53 (98.1%) patients. The history of meconium ileus was found only in 2 (3.7%) patients.
Conclusion: A 'gold standard' for CFLD diagnosis is lacking. The classic ultrasonography is the most common radiologic exam used to identify CFLD, in conjunction with clinical and laboratory exam for followed up CF patients during 2019. The UDCA is used in 52.8% of the CF patients and the prevalence of CFLD is around 50.9% higher than reported. This may be due to the high prevalence of F508 homozygote and pancreatic insufficiency, as well problems in definitions and diagnostic tools mainly based on the phenotypic reporting CFLD.

P167
Prevalence of liver disease in cystic fibrosis patients in the Institute for Respiratory Diseases in Children - Skopje, Republic of North Macedonia

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Objectives: Our aim is to provide epidemiological data of liver disease in cystic fibrosis (CF) patients in the CF Department in the Institute for Respiratory Diseases in Children in Skopje, Republic of North Macedonia.
Methods: We reviewed the data of 43 CF patients who are followed in our institute. Liver disease was diagnosed according to the medical history, liver enzymes and echographic findings.
Results: A total of 29 patients (67.44%) had CF liver disease (CFLD) almost equally distributed between males and females (15, 14 respectively). The most frequently associated mutation was F508del/F508del and heterozygote for F508del (27 patients - 93.1%). 10 patients (34.48%) had cirrhosis (9 of them are carrying at least one F508del mutation) including 3 patients (10.34%) with complications such as portal hypertension and esophageal varices. 24 patients (82.75%) received Ursodeoxycholic acid (UDCA) as a therapeutic option in established CFLD.
Conclusion: Liver disease is common among CF patients in our institute; almost all of them are carriers of F508del mutation and most of them have good compliance with UDCA therapy.

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Use of faecal elastase in patients with cystic fibrosis and mutation analysis in the Republic of North Macedonia

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Introduction: In the Republic of North Macedonia, patients with cystic fibrosis (CF) use pancreatic enzymes and its use is based on clinical criteria and through indirect methods of measuring pancreatic insufficiency.
Aim: To reclassify the patients with cystic fibrosis that have a clinical diagnosis of pancreatic insufficiency through introducing for the first time the measurement of faecal elastase-1 in North Macedonia, and correlate it with the genotype.
Materials and methods: The study included 27 patients (14 girls) with CF who regularly attend the CF centre at the Institute for Pulmonary Diseases in Children, Skopje, R.North Macedonia, with mean age 34.7 (1–39 y) years. CF was established on the basis of a positive sweat test (Macro duct, Wescor USA) and the results of molecular genetic analysis. Faecal elastase was measured through ELISA (Maxy Allergy). The values of FE-1 in the range 0–100 μg/ml, are characteristic for severe exocrine PI, 100–200 μg/ml for moderate exocrine PI, and >200 μg/ml for pancreatic sufficiency.
Results: The FE-1 value ranged from 7.1 to 519.7 μg/ml, with an average of 125.8 μg/ml. In 13 patients (85.2%) the value was <200 therefore had pancreatic insufficiency (PI). Four patients had normal values. 55% of the CF patients were homozygotes for F508del. They all had pancreatic insufficiency (PI) with mean FE-1 59.6 μg/ml (7.1–165.3 μg/ml). CF patients who were heterozygous for F508del mutation, were also with PI. Only 14.8% of F508del heterozygous CF patients were pancreatic sufficient (FE-1mean value 504.4 μg/ml) with other mutations V456F, c.1070C>T,3849G→A.
Conclusion: The use of faecal elastase can help clinicians to avoid the unnecessary use of pancreatic enzymes. The homozygous F508del mutations were associated with severe pancreatic exocrine insufficiency. Most of the patients with cystic fibrosis and clinical diagnosis of PI were confirmed with low levels of faecal elastase.

P169
Trends in glycaemic control in a cohort of patients with cystic fibrosis-related diabetes

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Objectives: Cystic fibrosis-related diabetes (CFRD) is common amongst adults and adolescents with cystic fibrosis, with the CF national registry suggesting that around 1/3 of patients >16 years with cystic fibrosis are receiving treatment for CFRD. Our aim is to report the prevalence of CFRD amongst patients attending the All Wales Adult Cystic Fibrosis Service, and to explore trends in HbA1c over time.
Methods: We report the prevalence of CFRD amongst patients attending the All Wales Adult Cystic Fibrosis Service. We describe the demographics of those with CFRD and describe trends in HbA1c between 2015 and 2019.
Results: 74 of 305 (24.3%) patients attending the All Wales Adult Cystic Fibrosis Service have CFRD. The mean age of patients with CFRD was 33.2 years (range 18–60). 14 (18.9%) had undergone lung transplantation. 64 (86.4%) patients were receiving insulin therapy, 6 of these (9.4%) by continuous subcutaneous insulin infusion. Mean HbA1c amongst patients with CFRD at the All Wales Adult Cystic Fibrosis Service increased between 2015 and 2019 (59.0 mmol/mol to 64.6 mmol/mol, $p = 0.039$). There was no correlation between age and HbA1c. There was no significant difference in HbA1c between men and women ($p = 0.94$). Patients receiving insulin via continuous subcutaneous insulin infusion did not have better glycaemic control than those receiving multiple daily injections of insulin (67.2 mmol/mol vs 67.9 mmol/mol in 2019, $p = 0.94$).
Conclusion: CFRD is common amongst adolescents and adults with cystic fibrosis. Despite significant local efforts to enhance services focused on delivering specialist care to patients with CFRD, and increasing availability of technologies including continuous subcutaneous insulin infusion, glycaemic control has deteriorated over time in our cohort. Improving glycaemic control is a priority and will be essential in reducing CFRD complications in an ageing cohort of individuals with cystic fibrosis.

P170
Current status of cystic fibrosis-related diabetes in the Bulgarian cystic fibrosis population

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Cystic fibrosis-related diabetes (CFRD) has emerged as a common comorbidity in CF and is associated with worse health outcomes.