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often complicated by concerns about the potential implications. Our study investigated clinical status in the first visit and a year post transition.

Methods: CF patients undergoing transition from paediatric Garrahan Hospital in Buenos Aires to Rossi Hospital Adult Center in La Plata city between 2011 and 2017 were included. Data was collected retrospectively from the first visit in an Adult Center and the first year post-transition for the 25 patients who transitioned from our paediatric to adult program. FEV_1, FEV_1% and Body Mass Index (BMI) were recorded.

Results: During the study period, 25 CF patients were transitioned (9 females). Three patient was lost to follow up. FEV_1 decreased after transition (median FEV_1 2.25 (62%) before versus 1.8 (56%) after transition). BMI during first year after transition was from 19.75 to 20. There was no significant difference in the mean decline in FEV_1 ($p = 0.06$) in the first visit compared to the year post. It was a significant difference in FEV_1% ($p = 0.01$) There was no significant change in BMI after transition ($p = 0.51$).

Conclusion: In a cohort of patients with CF transition is not associated with a clinical decline; however, it is associated with a decline in FEV_1%. In our transition program with multidisciplinary meetings and clinic visits among CF paediatric and adult care teams, careful monitoring of patients during the transition period at close intervals is recommended to underline the importance of daily routine therapy and self-management.

P058

Ageing in cystic fibrosis: experience of a large UK cystic fibrosis centre

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Objectives: Survival in cystic fibrosis (CF) is increasing and we are now seeing an ageing adult population. We aimed to review CF patients over 40 years of age, assess trends in demographics, predict emerging complications and highlight areas for research.

Methods: A retrospective review of clinical notes was performed for patients over 40 years of age in the Manchester Adult Cystic Fibrosis Centre (MACFC) to collate demographic data, age-prevalent CF complications and associated comorbidities.

Results: We identified that 90 of the 455 patients at MACFC are aged 40 years or above. A further ten will reach 40 years old in 2019, representing 22% of the total CF patient cohort at MACFC. Mean age is 48.4 (SD+/−7.8) years. 64 patients (14%) are aged between 40–49 years, 18 between 50–59 years, 6 between 60–69 years and 2 over 70 years. Of the 90 patients, 66.6% are male, 35.5% are Phe508del homozygous, 10% are homozygous or heterozygous for G551D, 47.7% have CF-related diabetes (CFRD) and 77.8% are pancreatic insufficient. Mean FEV_1 is 1.84 L (SD+/−0.97). 25.5% are taking CFTR modulators, including 12 on ivacaftor, 5 on ivacaftor/lumacaftor and 4 on tezacaftor/ivacaftor. Two patients are in trials for triple CFTR modulators. 11% have hypercholesterolaemia and 15.5% are on anti-hypertensive therapies.

Conclusion: Initial data shows the diversity of an older CF patient cohort. The over 40 age group represents a significant proportion of the adult CF population and will increase by 2% at our centre next year. Cardiovascular risk will become increasingly important in this group, especially with the growing prevalence of CFRD, hypertension and hypercholesterolaemia. Emerging CF-related, age-related and treatment-related comorbidities will inevitably increase the complexity of future management. Further research will be performed in our centre to better quantify the risks and consequences of ageing in CF.

P059

First results of Turkish National Cystic Fibrosis Registry

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Objectives: Turkish National Cystic Fibrosis (CF) Registry was established by Turkish CF Society and the data of CF patients have been collected annually. In this report, we present the first results of registry using the data collected in 2017.

Methods: The data were collected by a data-entry software system developed for the registry which was accessed from the internet. The data consisted of 55 variables including the core data and the annual clinic data of each patient.

Results: There were 1170 CF patients registered; 535 (46%) were girls. The mean age for diagnosis was 1.7 years and the mean current age was 7.31 years; 51 (4.36%) patients were older than 18 years. Genotyping could be done in 978 patients; 2 mutations were detected in 539, 1 mutation was detected in 193 patients; 246 patients' mutations could not be detected. 103 (8.8%) patients were homozygous for F508del. Among the detected mutations, the most common mutation was deltaF508 with an allelic frequency of 28.09%, followed by N1303K (4,96%), G542X (4.56%), 1677delTA (4.09%), G85E (3.78%), 2183AA- >G (3.23%) and 2789 + 5G>A (2.91%). The FEV1 of patients ranged between 11% and 150% (median 86%). Chronic colonisation with *Pseudomonas aeruginosa, Staphylococcus aureus, Burkholderia cepacia and Stenotrophomonas maltophilia* were seen in 245, 295, 14 and 27 patients, respectively. The most common complication was Pseudobartter syndrome in 120 patients, followed by sinusitis in 110, chronic liver disease in 93 patients. Pulmonary treatments included, inhaled antibiotics in 15.9%, recombinant human DNAse in 86.75%, hypertonic saline in 8.03%, inhaled mannitol in 5.04%, macrolides in 6.32% and noninvasive ventilation in 2.65% patients.

Conclusions: The establishment of national CF registry provided the first clinical data of CF in Turkey. Sustainability and including more centers and more patients each year will provide the real data about the disease in the country in the future.

P060

Genetic features of patients with cystic fibrosis living in the territory Volga Federal district (VFD) of the Russian Federation (RF) according to the Register of patients with cystic fibrosis in Russia in 2017

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Objectives: The VFD is home to 21,3% of the total number of Russian citizens. There are 14 regions in VFD: 6 republics (Bashkortostan, Mari-El, Mordovia, Tatarstan, Udmurtia, Chuvashia), 7 areas (Kirov, Nizhegorodskaya, Orenburg, Penza, Samara, Saratov, Ulyanovsk), Perm Krai. The VFD is a multi-ethnic population, dynamically developing economy and has a unique transit position - it is located at the intersection of migration flows. We studied genetic characteristics of CF patients living in the VFD.

Materials and Methods: In the Register of CF patients in Russia 2017 where the data of 646 patients living in the VFD at the age 0,1–47 years (13,01 ± 9,2).

Results: The search of mutations in gene CFTR was spent at 558 (86,4%) patients. 903 pathogenic alleles (80,9%) were identified, 57 different mutations were revealed. The allelic frequency of the mutations: F508del - 50,4%, E92K - 8,7%, CFTRdele2,3–5,0%, 3849 + 10kbC->T - 2,1%, N1303K - 1,9%, 2143delT - 1,7%, L138ins - 1,5%, 394delTT - 1,2%, G542X - 0,8%, R334W - 0,7%, S466X - 0,5%, W1282R and 3272-16T->A - 0,4% each; S1196X, 3272-16T->G, 1677delTA, W1310X, 1367del5 - 0,3% each; c.252T>A, 3944delGT, Dupex6b-10, R347P, L1335P, W1282X, G551D, 2789 + 5G>A, 2184insA - 0,2% each; R553X, R1066H, R117C, W401X, 4015delA, CFTRdele1-10, 3821delT, 621 + 1G->T, A141D, Q290X, P988R, c.1742T>G, W19G, S945L, CFTRdele8, 1259insA, 3667ins4, c.1766 + 2t>C, c.743 + 2T>A, R1158X, E217G, 574delA, R792X, 2043delG, G194R, c.458G>T, R1162X, 1717-1G->A, S1159P - 0,1% each. 213 (19.1%) alleles remained unidentified.

Conclusion: The most frequent mutations in the CFTR gene in CF patients in the VFD were the F508 del, E92K, CFTRdele2,3. In the patients with CF in VFD, there was a wide variety of mutations in the CFTR gene associated with presence of autochthonous ethnic groups, and with migration of the population, which is increasing as a result of global socio-economic processes. Genetic examination should be continued.

P061

The significance of the National Cystic Fibrosis Patient Registry for the optimisation of care for patients with cystic fibrosis in the Russian Federation

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Objectives: Cystic fibrosis (CF) patient registries have become an important epidemiological tool for demography, networking, and quality management.

Methods: In Cystic Fibrosis Patient Registry of Russian Federation (CFPR-RF) 2017 was included the data of 3096 (in 2016–3049) patients from 81 regions of RF.

Results: The main data: average age of patients was 12.1 ± 9.4; adult patients (>18 years) - 22.3% (in 2016–24.3%); age at diagnosis - 3.1 ± 5.8; diagnosed by the neonatal screening - 47.8% (44.7 in 2016). Only 92,4% (90.4% in 2016, 88.2 in 2015) underwent DNA analysis for *CFTR* mutations, 88.3% alleles were identified. Frequency of F508del - 52.81%, CFTR dele2,3–6.21%, E92K - 3.0%, 2143delT - 2,15%, 3849 + 10kbC->T - 2.02%. The number of patients with chronic *P. aeruginosa* (32.4%), *S. aureus* (57.1%), *B. cepacia* (6.2%) and *St. maltophilia* (3.5%) didn't change during the last 5 years, but we see the increase in *Achromobacter spp* up to 4.6%. FVC - 84.5 ± 23.1% and FEV1 - 75.4 ± 25.3%. Complications: CF related diabetes mellitus - 3.2%, liver cirrhosis with portal hypertension - 4.5%, electrolyte disorders - 2.9% (4.6% in 2016), pneumothorax - 0.6%, nasal polyps 23.6%. During 2017 49 patients died, mean age of death 22.0 ± 9.9 (15.9 ± 11.2 in 2016). CFTR modulators are not available and patients receive symptomatic treatment with significant increase in the use of hypertonic saline in last years - 63% (8.7% in 2011, 50.1% in 2015, 54% in 2016) and inhaled mannitol in 2017.

Conclusion: The number of centers and patients participating in CFPR-RF was increased dramatically since 2011. We still have a large proportion of CF patients that are either not tested at all genetically or have unidentified one or two *CFTR* mutations. Genetic testing should be intensified.

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P062

Albanian cystic fibrosis patients during the year of follow-up 2017 - data from ECFSPR

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Objectives: To present the epidemiological 2017 data on CF in Albania, provided by statistical analysis of ECFSPR

Methods: Demographic, genetics, lung function, microbiology, nutrition, complications, therapy data entered into the ECFSPR software during the period April- July 2018

Results: 123 CF patients were registered in ECFSPR, mean age 8.8 years, 115 children (<18 years), 8 adults (>=18 years), mean age at diagnosis 0.66 year old. 90.16% of patients were analyzed for 31 mutations. The no of patients with both mutations 91(82.73%), with one unknown mutation 19(17.27%). The frequencies of mutations: F508 del- 81.36%; unknown-10.45%; 621 + 1G->T- 2.73%; G85E -2.27%; G542X -1.82%; N1303K- 0.45%; R1070Q- 0.45%; R1158X-0.45%. The mean FEV1% of predicted: 89.7%. In age group 6 - 17 years, FEV1% of predicted: <40% in 2; 40–80% in 8; >80% in 45 patients. In adult group, FEV1% of predicted: 40–80% in 4; >80% in 4 patients. Chronic P. aeruginosae was found in 18.85%, mostly in adults, Chronic Staph. aureus infection was found in 28.69%, mostly in adults (87.50%). The pancreatic enzymes were used by 95.9% of patients. The Z-scores for height for patients aged 17 years or younger was: −0.5(−4.1–1.7), for adult was: −0.9(−1.7–0.8). The Z-scores for weight for patients aged 17 years or younger was: −0.7(−5.3–1.9), for adult patients was: −1.2(−2.4–−0.2). The Z-scores for BMI for patients aged 2–17 years was: −0.7(−7.3–1.9), for adult patients was: 20.1(17.3–23.5). The ABPA was found in 2 patients. The CF related diabetes was found in 3 patients under 18 and 1 adult patient. The liver disease without cirhosis was found in 32.79% of patients, the use of ursodeoxycholic acid in 31.15% of patients.