458* The prognosis of cystic fibrosis in the Western Cape province of South Africa: a 33 year study

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Background: Cystic fibrosis (CF) occurs in all population groups in South Africa, a middle income country with significant poverty levels.
Patients and Methods: Applying Kaplan–Meier statistics to a database of CF patients in the Western Cape province, the overall survival of 217 patients with CF was studied over a 33 year period (1974–2007). The effect of sex and ethnicity on prognosis was explored.
Results: Median survival was 20.8 years (25th percentile: 11.6 years). There was no significant difference in survival between males and females ($p = 0.5$). There was a significant difference in prognosis between white ($N = 116$) and mixed ethnicity ($N = 92$) patients ($p = 0.0025$). There were 17 deaths in infancy (22.6% of all deaths) spread equally across the period. Infant deaths accounted for 2/33 deaths in white patients versus 13/39 deaths in mixed ethnicity patients. Median survival beyond infancy for white patients was 25.8 years versus 20.5 for mixed ethnicity patients ($p = 0.007$). There was no demonstrable improvement in survival for patients born in the second half of the study period.
Conclusions: Median survival is into adulthood in this population. Infant death due to CF is a significant risk in the province. There is an ethic difference in survival that is likely to relate to awareness of CF in different populations and differences in socio-economic status.

459 The characteristic of children with cystic fibrosis from Moldova according to European Registry criteria

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Aim: To analyze the information about children with cystic fibrosis (CF) from Moldova according to European Registry of CF.
Materials and Methods: This study included 55 children (32 boys, 23 girls at the mean age of 9.67 ± 0.9). Genetic diagnosis was realized in all the children and their parents for 4–7 CFTR mutations. The explorative program for children with CF according to European Registry includes: sweat test, spirography, sputum bacteriology, body mass index and pancreatic status appreciation.
Results: The diagnostic of CF was established at the mean age of 3.71 ± 0.7. In 25 children (44.6%) ΔF508 mutation was revealed (12 children homozygote, 13 heterozygote), L551D mutation – 1 child, R334W – 1 case, G542X/N103K – 1 child. Sweet test value consists 92.9 ± 4.6 mmol/l. Bacteriology of sputum was characterized by prevalence of pulmonary infection with *S. aureus* in 48.25%, *Ps.aeruginosa* – 30.35%. The respiratory pulmonary function was characterized by restrictive and obstructive impairments FVC -61.5 ± 3.6%, FEV1 – 64.5%. Nutritional disorders were determined in 87.5% characterized by low BMI (15.95 ± 0.3 kg/m^2). Pancreatic insufficiency was revealed in 78.6% cases. There are 5 children official registered died from CF during last 5 years (2 children at the age of 1 year, 2 at 2–3 years, 1 child at 17 years). The cause of death in all cases was infectious pulmonary complications.
Conclusion: In children evaluated according to European Registry of CF criteria high incidence of ΔF508 mutation, the presence of pulmonary disorders and in 1/3 of children chronic colonization of low airways with *Ps. aeruginosa* were observed. The implication of gastrointestinal system characterized by pancreatic insufficiency and impaired nutrition was present in most of children.

460 Data from the Croatian CF Registry

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In 2007 Croatia joined the European CF Registry and parallely, following the same instructions for data collection, the national CF registry was started. We present the first data on our CF patients.
Croatia has about 4.500000 inhabitants and a total of 108 CF patients are reported (52.8% male). 52% are followed at the University Hospital Centre Zagreb and the rest are distributed among 5 other institutions. We have no neonatal screening. Median age at diagnosis is 5 mo, with 46.6% patients identified by the 3rd mo of life. The median age of our patients is 14.5 y and the oldest has 28 y. The range of sweat chloride concentration is 43–280 mmol/l. Most patients have 90–120 mmol/l (59.9%); the median concentration is 105 mmol/l. The most frequent CFTR mutation is dF508 (78.2% of all identified mutations). 61 patient is homozygous and 36 patients are heterozygous for dF508. Other mutations found are G542X, R117H, N1303K, and 1717–1G-A. 3 patients have no identified mutation (N/N) and 32 patients have one unknown mutation (15.5%). Most patients are PI (96.3%). 20 had meconium ileus. The overall nutritional status of our patients is fairly good with the medium BMI %tile of 45, but 33.8% have BMI bellow 25% tile. We have 4 patients with CFRD. In 2006/07 *Pseudomonas aeruginosa* was isolated in 46% of patients and *Burkholderia cepacia* in 2 patients. We treated 3 patients for ABPA. No patients have liver or lung transplant.
Comment: Only 108 patients registered among the Croatian population with the genotype similar to surrounding nations means CF is underdiagnosed. However, the care for patients is fairly good with acceptable figures regarding bacteriological isolations, nutritional status, age at diagnosis and actual patients age.
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461 CF Registry in Slovakia

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Slovak CF Registry was established in 1995 and includes 505 CF patients in the end of 2006, 251 males, 254 females, mean age 18.9 years. 106 pts. died in this period with increasing mean age from 3.4 years in 1974–1980 to 16.3 years in 2000–2006. There are 187 adults (46.9%), 212 children (53.1%), 9 patients are older thane 60 year. 93.4% of the patients are genetically evaluated, the prevalence of dF508 being 46.35%. Nevertheless, 33.4% of our patients who where tested, have unknown mutation. Data on registry are obtained from 3 CF centres and includes demographic data (sex, date of birth and death, genotyping, social status), sweat chloride, age in diagnosis, clinical symptoms, pancraetis status). We do out-patient visits every 1–3 months (spirometry, anthropometric measurements, blood samples for hematology, biochemistry and immunology and sputum collection). Data of therapy: antibiotics, mucolytics, antiinflammatory therapy, ursodeoxycholic acid, pancretic enzymes, enteric suplements, lung and liver transplantion.
Results: See the table.

Clinical symptoms and therapy in CF patients

Mean FVC	85.3%	DIOS	8.0%	rhDNA	38.6%
Mean FEV1	85.0%	MI	8.4%	PET	72.3%
Colonisation		Prolaps	7.1%	UDCA	41.7%
Pseudomonas spp.	42.9%	Sinusitis	59.3%	Supplements	41.8%
Burkholderia spp.	14.6%	Polyposis	21.4%	CS	10.7%
PI	65.9%	ABPA	10.0%	ICS	23.3%
CPI	11.4%	IGT	37.0%	AZI	30.7%
CFRLD	48.6%	CFRDM	5.7%	Lung Tx	4 pts
Cirrhosis	6.3%	Inhaled antibiotics	36.6%	Liver Tx	0