374 Clinical and demographic characteristics of Estonian cystic fibrosis patients

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Objectives: The incidence of cystic fibrosis (CF) in Estonia is 1 in 7700 live births. The aim of this study was to analyze the demographic and clinical data of CF patients in Estonia on the census date (01/12/2010) and compare them to earlier data of 1993 and 2003.
Methods: There are 45 (26 males and 19 females) alive CF patients in Estonia. Clinical data of 39 patients, who have been in regular follow up in 2009/2010 at Children's Clinic of TUH (21), Tallinn Children's Hospital (17) and North Estonia Medical Centre (1), were reviewed.
Results: The mean age of our CF patients has increased from 8 y 2 mo (1993) to 15 y 5 mo (2010). In 1993 there was no patient >18years old vs. 29% (9/31) in 2003 and 38% (17/45) in 2010. The median age at diagnosis of CF in Estonian patients (1 year 6 months) is still higher than in the EU countries. The chronic *P aeruginosa* colonisation was found in 18/39 and *S. aureus* in 14/39 patients. Majority of patients (84%) had good lung function according to FEV1 predicted >70%. The mean value for FEV1 was 91.1 % (95%CI 75.6–106.5) and for FVC 89.4 % (95%CI 60.5–118.5). Chronic liver disease was detected in 12/39 and diabetes only in 2 patients. One 12 old year girl with liver cirrhosis had liver transplantation. Comparing mortality rates throughout the following periods, a significant decline has been detected: from 12.2% (1983–1987) to 0.66% (2003–2010).
Conclusions: In Estonia slight but steady improvement in the survival of patients and their quality of life has been observed due to consistent changes in their follow up, centralized healthcare system and treatment options.

375 Expression of cystic fibrosis (CF) at diagnosis in a reference center in Rio de Janeiro

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The aim was to analyze the clinical-laboratorial characteristics of CF at diagnosis in RJ where incidence is estimated as 1:6902. It is a descriptive and sectional study of patients followed from Jan/08 to Sept/09. The diagnosis was made through the manifestation of the disease plus the sweat test. PI was defined by enzyme replacement; malnutrition weight/age, weight/stature/age, gender in ⩽2 yo, and BMI for >2 yo; morbidity; manifestations and hospitalization. In 105 patients, 55% were female, 58% white and 97% RJ residents. We verified a delay of 11 months between median ages at diagnosis and the onset of symptoms: 14 mo and 1.7 mo. CF was manifested by meconial ileum (9.6%); edema and anemia (9.6%); dehydration (29.8%); poor absorption (85%); PI (89.5%); malnutrition in patients aged ⩽2 (54.1%) and >2 (44.2%); coughing (87.6%), dyspnea (75.2%), thoracic deformity (30.5%), digital hippocratism (28.6%), *P. aeruginosa* (45.6%), *S. aureus* (34.4%), *B. cepacia* complex (5.6%). Patients who needed hospital treatment (46.7%), 66% with previous hospitalizations, 61% <1 yo. Identified mutations in 2 alleles in 38.6%: ΔF508/ΔF508 (15/70), ΔF508/other (7), other/other (1); and in 1 allele 40%: ΔF508/Unknown(U) (18), other/U (10), and U/U (15/70).
We highlight the following possible risk factors associated with morbidity: difficulty, delay and confusing it with comorbidities associated with prevalent diseases – malnutrition, diarrhea, dehydration, pneumonia, in addition to severe mutations. Early diagnosis and adequate interventions are necessary to reduce morbimortality which contrasts with the challenge posed on the genotypic determination of the studied population for therapeutical ends.

376* The role of electronic patient records (EPR) in improving service efficiency and clinical performance in a regional adult UK centre

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In 2007 an EPR system (EMIS®) was introduced at the Leeds Adult CF unit to improve service efficiency and patient care. Prior to EPR it had become increasingly difficult to deliver timely annual assessments (AA) and GP correspondence due to increasing patient numbers, a high DNA rate and limited staffing resources. The introduction of EPR enabled continuous patient monitoring and assessment. We report the impact of EPR on delivery of AA and clinical correspondence.
Data was collected pre and post introduction of EPR. Pre EPR patients attended for a full day's review and the AA details and results handwritten and summary dictated and typed. EPR an online diary alerts users when individual parts of AA are required. Results are entered electronically and acted on according to level of urgency. Consultation and discharge summaries post EPR are generated automatically in real time.
Pre EPR (2005) only 40% of total population attended for full AA. Median (range) number of days for summary to be dictated and typed was 48 (26–78) days. Post EPR the percentage of patients who had AA bloods, OGTT and U/S increased from 63%, 64% and 45% in 2005 to 92%, 89% and 86% respectively in 2010. Total time to sending a discharge summary to GP was reduced from a median (range) of 19.5 days (4–40) pre EPR to 1 (0–2) days with EPR. Clinic letters now generated at the time of consultation alleviating the need for dictation.
Despite increasing patient numbers and limited staffing resources the use of EPR technology has enabled us to deliver a more efficient and effective service. EPR is an essential patient management tool and further investment in new technologies is ongoing.

377 The new web-based Brazilian CF registry (REBRAFC)

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Objectives: to describe the new beginning of the Brazilian Cystic Fibrosis Registry (REBRAFC).
Methods: Initial conversations to restart the Brazilian CF registry occurred in early 2007 and the proposition of a Web based platform was done. The new Brazilian Registry was fully supported by the Brazilian Group of Studies of Cystic Fibrosis (GBEFC). The development of the Web based platform was done by the Laboratory of Integrated Systems (LSI), from the Politecnica School of Engineering, University of Sao Paulo. The modeling and project of the system used methods based on Unified Modeling Language (UML). Free and open software were adopted for programming, and the security of the system was planned in several layers using interceptions provided by the Struts2 framework. The main contents of the registry were Demographics, Diagnosis data and Clinical data. This last item would be inserted annually, including number of consultations, nutritional and functional data, current treatments, microbiology data and Schwachman-Kulczycki score. Identification of patients is protected. After initial testing of the platform, all known Centers and health professionals involved in CF care were invited to join the Registry. A formal contract was celebrated among the GBEFC and each Center to guarantee the confidentiality of the data. The start of data insertion took place in March 2010, but was very modest until the occurrence of the Brazilian CF Congress in September 2010. Approximately 1,400 patients were included until January 2011 (near one-third of all Brazilian CF patients).
Conclusions: The new Brazilian CF Registry is fully operational but strategies to improve adhesion are needed.