Evaluation of CF database in Republic of Macedonia

T.Jakovska-Maretti1, S. Fustik1, L. Spirevska1. 1CF center, University Pediatric Clinic, Skopje, Macedonia

The first specialized CF centre in R.Macedonia was established in 1996/97 year. The aims of the study were to determine the incidence of CF (no previous data available), to follow the evaluation of the disease and further complications.

Methods: Using CF database for demographic, genetic and typical clinical symptoms are described.

Results: Totally 100 CF patients (both children and adults) were identified in databases with mean age 10.9±12.4 years. The oldest CF patient is 26 years old. 13% of the patients is older than 18 year. 80% of the patients were diagnosed before the age of 1 year. 85% carry the DF508 mutation (65% are homozygote). 42% of them are colonized with Pseudomonas aeruginosa (20% have chronic PA infection). 90% have exocrine pancreatic insufficiency and 29% are malfourished (BMI <18.5 kg/m²). 8% of the patients have liver disease, meconium ileus had 5%, 4% have diabetes mellitus and 10% have CF bone disease (3% with osteoporosis). Mean lung function FEV1 in group of 6–18 years was 89.18±22.5 compared to FEV1 in patients >18 y (67.7±31.7). We don’t have any patient with lung transplantation.

Conclusion: Advances in management based on European standards are given in specialized CF center, and have increased survival in CF in R.Macedonia.

The UK CF Registry – a successful transition to a web-based system

E. Gunn1, M. Baker1, A. Larsen1. 1CF Registry, Cystic Fibrosis Trust, London, United Kingdom

Over the past decade the UKCF Database was established by a multidisciplinary team using a generic approach to monitor clinical outcomes of patients with CF in the UK. Software obsolescence and the need for greater sophistication led to a search for a new patient registry application. The web-based Port CF software developed by the CFF in the USA was identified as being appropriate. CFF were keen to collaborate in order to enhance international comparisons. A licence was agreed and the software was “anglicised” with the help of appropriate clinical expertise. The project received national ethical approval. After rigorous testing an implementation programme was initiated and the registry “rolled-out” to specialist CF centres. Online helpdesk facilities are available to assist local administrators. Centres add data direct to the registry in the form of clinical encounters. The minimum data requirement is demographic and clinical data from an annual review for each patient. For regional centres sharing care with smaller clinics, data can be entered at both sites thereby enabling the regional centre to maintain and monitor overall clinical care. By December 2007, 94 centres and clinics (58/36) have been implemented. Data collected by the former database has been mapped into the new CF Registry giving centres access to data from 1999 to date. Data are available for >6500 patients. Sophisticated analysis is available to each centre using simple reporting tools. This user friendliness and the ability of each centre to analyse their own data simply and efficiently has been key to the acceptability and take up of Port CF. The CF Registry will be used to monitor and improve quality of care in the UK. The Registry Steering Committee will produce an annual report of clinical outcomes for the prior year to be available in the autumn.

The development of cystic fibrosis care in the South West, including the development and impact of the South and West Cystic Fibrosis Database: a retrospective review

J. Tyrrell1, S.J. Gooch1, K. Giles1. 1Children’s Centre, Royal United Hospital Bath, Bath, United Kingdom

Cystic fibrosis, as a relatively rare disease with a distributed and small population in the UK, has undergone significant transformations in care provision and commissioning over the past 30 years. The paper aims to track these transformations with reference to the following; changes in care provision and the implications of these changes, the challenging geography of the South West and how shared care arrangements have developed to overcome this, and the changes in NHS structure and the resulting impact on how CF care is commissioned.

Patient registries are an essential tool in understanding disease progression. The South and West CF database was developed in 1995 from a need by local clinicians to understand their CF population. It now has 12 years of data that clearly show the changes in CF care presented above, as well as changes in clinical outcomes. In 1995 there were 62 consultants with an average of 10 patients per consultant in the South West. 48% (30) consultants had less than 5 patients on their list. In 2006 there were 36 consultants with an average of 23 patients each and only 6% (2) consultants had less than 5 patients. For patients not receiving care at specialist centres, 35% of adults and 53% of paediatrics had shared care in 1995; this has risen to 54% and 66% respectively in 2006.

In addition to providing high quality patient, population and clinical data, the presence of the database has had a positive impact on working practices in the region.