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A CF- specific database as a clinical an epidemiological tool

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Thanks to the collaboration between clinicians and I.T specialists, a CF-specific database system was developed using a Windev licence. We aimed to create a clinical tool suitable for our daily practice, which would also allow us to automatically extract every year a range of epidemiological data required by the Belgian Cystic Fibrosis Registry. Individual data are encoded, using different modules (such as admin, outpatient visits, hospital admissions, complications…). For each patient, the database automatically generates graphs summarising the evolution of essential parameters. These include weight (in kg, Pc, *Z*-score,%IWH), height (Pc, *Z*-score), BMI (absolute value, *Z*-score), FVC and FEV1 (% pred.), bacteriological findings. In addition, several tables of collective data are generated such as listings of administrative or basic medical data, warning listings, tables summarising anthropometrical, spirometric or bacteriological characteristics from all the patients, or according to age or sex. If the database is regularly updated throughout the year, then all the fields of the Belgian Register Questionnaire, which is closely derived from the CFF model, will be automatically completed. By default, Cole's reference values are used for anthropometry, and Knudson's for spirometry. Each of the 7 CF Reference centres in Belgium will be offered the opportunity to use this software, which will be available in French, Dutch and English. Supported by the Belgian Cystic Fibrosis Association.

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The frequency and presentation of pancreatitis in cystic fibrosis. A survey

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Pancreatitis is a rare complication of CF. In the present multi-centre survey, we evaluate the frequency, risk factors, clinical course and outcome of CF patients with pancreatitis. We recorded a total of 10071 CF patients from 29 different countries. In this population, there were 125 patients with pancreatitis (1.24%; 95% binomial confidence intervals [CI] 1.02% to 1.46%). We have received full clinical data on 61 patients with pancreatitis. The majority of patients with pancreatitis have PS: 34/61 patients (56%; CI: 43% to 68%). PI was found in 15 patients (25%; CI 14% to 35%). Eight patients had PI after an initial period of PS and four patients had a dubious pancreatic state. The amylase and lipase levels were not significantly different between patients with pancreatitis and PI and patients with pancreatitis and PS. Relapses of pancreatitis occurred more frequently in PS (21/34) than in PI (5/15) patients ($P=0.04$). Chronic pancreatitis occurred more often in PI (6/15) than in PS (3/34) patients ($P=0.01$). The occurrence of pancreatitis preceded the diagnosis of CF in 18/61 cases. Thirty-one out of thirty-four patients with PS carried at least one mild CFTR mutation; five out of fifteen PI patients carried 2 severe CFTR mutations. Pancreatitis is a rare complication in CF patients, occurring in 1.24% of a large CF cohort. Pancreatitis is more frequent in CF patients with PS, but has to be considered in all CF patients with unexplained epigastric abdominal pain, including those with PI.

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Prediction of 2-year mortality in CF – A comparison of Shwachman score and percent predicted FEV1

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Introduction: The Shwachman Score (SS) is perceived to be of low relevance to adult patients, and does not meet current criteria for a valid clinical scoring system.*Objective*: To determine validity of SS in prediction of 2 year mortality in the UK, and to compare this with% predicted FEV1 using the UKCF database. *Methods*: A retrospective cohort study (transplanted patients excluded). SS from annual review and average% predicted FEV1 from clinic visits were recorded for 2000. Mortality was recorded at 1 and 2 year follow-up. Receiver-operator curves (ROC) were calculated using StatsDirect. *Results*: 2706 patients were identified of whom 1773 had a SS and 2221 had a% predicted FEV1 recorded. 28 patients died during 2-year follow-up, 7 of which had SS.

	Shwachman	FEV1
Optimum cut-off	66	47.31
Sensitivity	0.833 (0.359 to 0.996)	0.893 (0.718 to 0.977)
Specificity	0.876 (0.860 to 0.891)	0.803 (0.787 to 0.820)
Positive predictive value	0.0230	0.0581
Area under curve	0.815 (0.198 to 1.00)	0.900 (0.592 to 1.00)

Doubling the weighting given to specificity improved positive predictive value of FEV1 (0.106 with a cut-off of 33.7 and sensitivity 0.714), but not SS (results same as above). *Conclusion*: There appears to be little difference in the validity of Shwachman and% predicted FEV1 to predict 2 year mortality in patients with CF however there were few deaths in this cohort and neither has high predictive value. More SS data is being collected.

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Incidence of cystic fibrosis worldwide

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Cystic fibrosis (CF) is defined as the most common inherited disorder that affects children in Caucasian populations, with an incidence initially estimated to 1/2500 livebirths. The aim of this study was to perform a meta-analysis of the incidence of CF in the world. CF incidence could be measured by different study design (epidemiological studies, CF-registries, newborn screening (NS) programs) or estimated by carrier frequency. The different studies dealing with CF incidence were retrospectively registered by using bibliographic databases but also internet tools in order to found unpublished data. The incidence of CF is reported by continent. In Europe, its oscillated between 1/1700 (Irlande, Brittany) and 1/25 000 (Finlande). In North-America, data of registry and NS showed an incidence of 1/3500. The same data was observed in Australia where NS is highly practised. Very high incidences were reported in isolated populations, such as 1/377 in an area of North-Brittany (France), 1/569 in the Amish population (Ohio, USA) or 1/902 in Saguenay-Lac-Saint-Jean (Québec). Few data are available in non-Caucasians groups, even if several studies have determined the mutation spectrum in those populations. Incidence is close to 1/9000 in South-America, 1/17 000 in Africa and around 1/30 000 in Asia. The precise knowledge of CF incidence in different ethnical groups is crucial for genetic counseling to allow a precise calculation of residual risk. The implementation of NS programs has constitued a precious help to estimate this incidence. They generally provide a lower incidence than the epidemiologic studies performed at an earlier period where prenatal diagnosis did not exist.