231 Mathematical modelling of FEV1% allows early recognition of patients at risk

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The rate of decline of pulmonary function in CF has been reported to be constant, however linear decay is a simplification and more complex models are needed to describe FEV1(% pred.) We have developed a mathematical model to describe FEV1(% pred.) changes in time.
We used the data of the CF Center in Verona (Italy) where neonatal screening for cystic fibrosis was introduced in 1973 and became compulsory in the early 1980s. We analyzed 11281 values of FEV1(% pred.) of 242 patients diagnosed by neonatal screening and followed since birth. In order to be included in the analysis a patients should have performed at least 6 spirometry measurements since the age of 7 yrs. The observation period lasted between November 1979 and April 2007. The model was chosen using the Akaike information criterion [1]. It indicated the second-order mixed-effects model, with regard to both the average evolution and the variance structure. Multi-slope models were also applied to the data. A non-linear method was used to determine the age at which let the slope change. The Akaike information criterion suggests the use of the three slopes model (r^2: 0.88; AIC value: 83518.44), which was so used as a basis for further analysis.
The model allowed to reliably forecast the evolution of FEV1(% pred.) since the age of thirteen.
This model should now be validated in other centers and if so it could be used to identify early patients at risk, to study period effects on disease severity and progression and the influence of risk factors or treatments on pulmonary disease in CF.

Reference(s)

[1] H. Akaike (1978) A Bayesian Analysis of the Minimum AIC Procedure, Ann. Inst. Statist. Math., 30, 9–14.

232 Accuracy of a self-administered questionnaire to estimated maximal exercise capacity in CF patients

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Exercise capacity is one of the prognosis factors in cystic fibrosis (CF). Maximal oxygen consumption (VO2peak) quantifies this aerobic capacity. It is therefore relevant to use a low cost and practical tool to estimate this prognosis data.
We propose to test a new self-administered questionnaire on daily physical activity (QDPA) estimating VO2peak and daily energy expenditure (DEE). DEE was calculated by multiplying time spent in each activity by its energy cost corrected for weight, age, sex, autonomy and the total was calculated over 24 h. This tool divided DEE into three levels of intensity: <3 MET, 3–5 MET and >5 MET. These data were compared with VO2peak obtained from a maximal modified shuttle test (MST).
18 adults CF patients with stable disease completed the study (10 females, age=28±8.7 yrs, FEV1=55.6±26% of predicted, BMI=21±3.4kg/m2). All except three had pancreatic insufficiency. Two third had chronic pseudomonas colonisation. DEE reached 10200.5±3170.2 kJ/24 h and was significantly correlated with VO2peak (L/min) measured during the MST ($r = 0.796$, $p \leqslant 0.001$). It was also correlated with BMI ($r = 0.808$), FVC% ($r = 0.809$) and professional activity ($r = 0.493$). No correlation was found with age, sex, bacteriological status, or diabetes. DEE estimated by QDPA was determined by both intensity and type of activity.
This questionnaire seemed relevant to estimate maximal exercise capacity in CF patients. It also added qualitative and quantitative information on DEE. It can be a simple and interesting tool if larger studies confirm our results.

233 Assessment of respiratory compliance in infants with cystic fibrosis

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Aim: To estimate respiratory compliance in infants with CF at two inflation pressures and to compare measurements in individuals over time. To evaluate the association between compliance and measures of forced expiration.
Methods: Respiratory compliance was measured as the change in lung volume at inflation pressures of 20 and 30cm of water during passive inflations. Raised volume(RVRTC 30) parameters such as FVC, FEV 0.5 and MEF 25 were measured and z scores calculated based on published reference equations. Measurements were repeated 6 to 12 months later.
Results: 20 infants were tested at median (range) ages of 21.8(11.5–75.7) and 59.7(46.8–114.4) weeks at the first and second test respectively. Mean (SD) respiratory compliance at 20 and 30 cm inflation pressures was 9.5(3.5)and 10.6 (4.3)ml/cm H20 at the first test and 11.4(3.8) and 12.7 (3.9)ml/cm H20 at the second test respectively. While there was no statistically significant change in respiratory compliance between the tests (at 30 cm of water $p = 0.09$), there was a statistically significant decrease in FVC, FEV 0.5 and MEF 25 z scores over this time. There was no association between respiratory compliance at each test, or change in compliance over time, with gender, airway infection and measurements from the RVRTC.
Conclusions: Estimated respiratory compliance at 20 and 30cm were similar suggesting that measurements were made at similar parts of the pressure volume curve in infants with CF. Early CF lung disease is characterised by airway obstruction which is not associated with a change in respiratory compliance.
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234 Spirometry and Impulse Oscillation measurements in children with cystic fibrosis (CF)

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Management of CF respiratory symptoms includes lung function testing (LFT) and, in particular, the early identification and treatment of small airways disease. Spirometry, a commonly used LFT, requires a forced expiration and is more suitable for children >5 years old. The impulse oscillation system (IOS) measures airway resistance, impedance and reactance and requires only quiet tidal breathing. Work on IOS has been published in pre-school children with asthma but there is a paucity of data in children with CF.
Currently mid-expiratory flow (MEF) parameters are used to asses the calibre of the small airways, an area of concern in CF children. We hypothesized that MEF and IOS measurements related to small airways would correlate well, suggesting IOS to be a suitable diagnostic technique in CF children unable to appropriately undertake spirometry.
Data from 21 consecutive CF patients (8 female, 13 male, ages 6.0–15.9 yr) was analysed. Comparison of spirometry and IOS parameters, Spearman's Rank Correlation, resulted in r values between –0.59 and –0.94 ($p < 0.01$, in all cases). $MEF_{75–25}$ correlated against X5, reactance at 5Hz, the primary parameter of small airway disease, had $r = –0.84$. There was no linear relationship between $MEF_{75–25}$ and R5-R20Hz, difference in resistance at 5 and 20Hz and the secondary parameter of small airways. The highest r values were MEF_{75} against Z5, impedance of the total respiratory system at 5Hz, and against R5, $r = –0.94$ and –0.92 respectively. IOS may be a suitable alternative or additional test for assessing small airway function in young CF children. However, the complex relationships between the two measurements remain to be evaluated and may reflect differences in small airways function during quiet tidal breathing and forced expiration.