202 Pulmonary tomosynthesis in adults with cystic fibrosis: Our preliminary experience

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Objectives: Radiological imaging plays an important role in the evaluation of pulmonary cystic fibrosis (CF) for the detection of both disease progression and parenchymal lesions. Tomosynthesis (digital tomography) has been recently proposed for pulmonary CF because it has comparatively low radiation dose (0.08–0.12 mSv) and, compared to radiography, it is more sensitive to CF changes, showing them in more details.
The purpose of this study is to design and validate a scoring system for tomosyntesis in pulmonary cystic fibrosis of adult patients, comparing with chest radiography, computed tomography (gold standard) and functional parameter (FEV1).
Methods: To better investigate strengths and weaknesses of tomosynthesis, we independently scored radiography, tomosynthesis and HRCT examinations of the chest in a group of 20 adult CF patients (23 to 61 years, mean 39 years).
Also, we suggested a scoring system to evaluate the whole spectrum of mild to severe pulmonary changes, with well-defined scoring parameters, valid for all radiological investigations.
The scoring form provided clear definitions for the severity and extend of five well-recognised pathological changes: overinflation, bronchiectasis, bronchial wall thickening, parenchymal lesions and mucus plugging.
Conclusion: We found that correlation between the tomosynthesis and HRCT was high (square-weighted kappa = 0.99) and there was agreement in determining the location and extent of all lesions. We conclude that tomosynthesis is a useful tool in the regular follow-up of CF patients, allowing greater diagnostic accuracy than radiography and lower radiation dose compared with HRCT.

203 A longitudinal survey of physical performance and quality of life in adult CF patients

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Aim: To longitudinally evaluate physical performance and quality of life in a cohort of adult CF patients (non transplant candidates).
Methods: Physical performance and quality of life were measured at two time points (at least six month apart) with pulmonary function (FEV1), six-minute walk test (6MWT) the St George's Respiratory Questionnaire (SGRQ) and CFQ-R (only at second evaluation).
Results: 26 patients (13 females) with a mean age of 28±10 years, body mass index (BMI) of 22±3 kg/m^2, FEV1 2.4±1.4 litre (63±29%) and 6MWT 703±148 meter were investigated. Total score of SGRQ at first evaluation was 22±14 (healthy adults <6), with a symptom score of 44±21 (<9), activity score of 23±22 (<2) and impact score of 14±13 (<6). Results of the CFQ-R are comparable to published date from the literature. Mean change in BMI, FEV1, FEV1%predicted and 6MWT at second evaluation (8.3±4.0 months after first evaluation) was 0.18±0.69 kg/m^2, 0.02±0.30 litre, 0.08±7.82% and 9.7±98.3 meter, respectively. Mean change in SGQR was 4.9±14.0, 3.6±7.2, –2.5±16, 3.0±8.3 for activity, impact, symptoms and total score, respectively. 6MWT and FEV1 are strongly correlated with the SGQR but no correlation of change in SGQR and delta 6MWT and FEV1 over time was found.
Conclusion: The specific respiratory questionnaire (SGRQ) showed an impairment of respiration related quality of life compared to healthy adults. SGQR was strongly correlated with FEV1 and 6MWT. Due to the small changes in physical performance over the whole observation time no significant correlation of change in FEV1 and 6MWT to SGQR was found.

204 Clinical and functional determinants of exercise performance in adults with cystic fibrosis

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Background: Exercise tolerance is reduced in adult patients with cystic fibrosis (CF). The aim of this retrospective analysis was to determine the mechanisms responsible for exercise intolerance.
Methods: Cardiopulmonary exercise testing (CPET) with blood gas analysis at peak exercise was performed in 102 patients aged 28±11 years. VO_2 peak was measured and correlated with clinical, biological and functional parameters.
Results: VO_2 peak was decreased (<84%) in 85% of patients and was correlated with BMI, CRP, FEV_1, FVC, RV and DLCO. Among the CPET variables, VO_2 peak was correlated with V_E/VO_2 at ventilatory threshold ($_{VT}$), peak V_D/V_T, PaO_2, $PaCO_2$, P(A-a)O_2 and ventilatory reserve. In multivariate analysis, FEV_1 was the major significant predictor of VO_2 peak impairment, accounting for 48% of VO_2 peak alteration. Reduced or absent ventilatory reserve and excessive hyperventilation ($V_E/VO_{2\ VT}$) accounted for 10% and 8% of VO_2 alteration, respectively. V_D/V_T and P(A-a)O_2 peak each accounted for 1% of the change in VO_2 peak value.
Conclusion: Exercise limitation in adult patients with CF is largely dependent on FEV_1 but is also affected by nutritional and inflammatory status and by the ventilatory response to exercise.

205 St George's Respiratory Questionnaire (SGRQ): A useful tool in adult CF patients

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Objectives: The aim of this study was to prospectively evaluate the CFQ-R and SGRQ and to look on the impact of clinical data on both questionnaires.
Methods: Clinical data including pulmonary function test (PFT), the rates of CF-exacerbation in the last year (IER), CF related diabetes (CFDM) and positivity for pseudomonas aeruginosa (PA) were collected. Spearman correlation was performed for these clinical data with CFQ-R and SGRQ. Univariate and multivariate analysis for CFQ-R and SGRQ were performed with the following factors: gender, age, BMI, CFDM, FEV1%predicted, patients with and without PA and IER.
Results: 38 patients (16 females) are evaluated with a mean age of 29 y (95% CI 25–32), body-mass index (BMI) of 22.0 kg/m^2 (22.9–23.2), FEV1%pred of 64% (55–74). FEV1%predicted was strongly correlated ($p \leqslant 0.0001$) with SGRQ activity and total score but only moderately ($p < 0.005$) with CFQ-R physical functioning and SGRQ symptom score. IER was strongly correlated with CFQ-R role, SGRQ activity and total score and moderately with CFQ-R physical functioning, treatment burden and health perception and SGRQ impact score. FEV1%predicted, IER, CFQ-R and SGRQ are significantly influenced by PA. In the multivariate analysis FEV1%predicted and IER had a significant influence on SGQR.
Conclusions: In adult CF patients CFQ-R is only moderately correlated with PFT and CF-exacerbation rate in the last year. In contrast, the SGRQ showed a better correlation with PFT and IER. SGRQ is a useful tool to measure health related quality of life in CF patients.