235 Factors influencing the worsening of lung function between 16 and 18 years of age in Czech patients

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Despite improved medical care we still observe marked deterioration of lung function between 16 and 18 years of age followed by increased mortality. The aim of the study was to analyze the factors influencing such decline. We analyzed lung function tests of 87 patients longitudinally followed up at our Centre between 14–18 years who were treated by modern approaches for at least 5 years. We evaluated the influence of gender, CFTR genotype, pancreatic function, bacterial infection, CFRD, nutritional status and previous development of their lung disease on its deterioration between 16 and 18 years. FEV_1 at the age of 16 years was 76.5±24.4%, at 18 years 65±23.3%, two year's FEV_1 decline was 11.6±14% (median 10%). We did not prove the influence of gender, genotype, pancreatic function, nutritional status, diabetes and *Pseudomonas aeruginosa* infection on this decline. In 42 patients infected by *Burkholderia cepacia* complex (*Bc*c) the worsening was more marked 15.1±12.9% (med 12.5%), than in the remaining 45 patients 8.2±14% (med 6%; all above of the predicted value; $p = 0.019$). A more pronounced deterioration was observed in patients whose lung function was better at the age of 16 years ($r = -0.369$; $p = 0.001$). This association is particularly strong in *Bc*c infected patients. The deterioration between 16 and 18 years of age is influenced mainly by *Bc*c infection, but it is present also in other patients. Finally, we have to consider non-compliance with therapy, during the age of transition to adulthood, as another deleterious factor.

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236 Listing patients to lung transplantation (LTx). Experience of 110 cases in the CF Center of Verona

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In the period 1999–2008 110 pts were evaluated for LTx by a multidisciplinary team (MD, physiotherapist, psychologist, social assistant). In 99% of cases the main indication was respiratory insufficiency, in one recurrent severe haemoptysis. 3 patients were listed for double L and liver Tx, one for double L and kidney. Psychiatric disorders in 1 case was considered an exclusion criteria but the pt was eventually listed by the surgical LTx Center. Other exclusion criteria were M cheloneae infection, thyroid or colon carcinoma. One of two patients with previous lobectomy/pneumectomy was excluded for the presence of severe thoracic organ displacement. B cepacia (genomovar III), HIV infection, multiresistant P aeruginosa, severe malnutrition, prolonged steroid treatment, young age, severe portal hypertension with TIPS, mechanical ventilation were not considered exclusion criteria. Pts were referred to 5 surgical centers. The median age was 26 yrs and 17 pts were <18 years. Thirty-three pts died in the waiting list (70% in the first year) and 62 were successfully transplanted (median time in list 7 months). Patients who died early (<1 year) in list had required significantly more iv antibiotic treatment (93±43 vs 63±47 days) in the year before listing but did not differ otherwise (diabetes, gender, age, BMI, pathogens) from those who survived >1 yr in the list. Overall survival after Tx is 75% at 5 years.

Conclusion: criteria to exclude CF pts from LTx are very few, even conditions such as HIV infection, B cenocepacia could be questioned as absolute contraindications. Recognizing patients with very poor prognosis is a major challenge, but our more recent experience lead us to consider patients for LTx earlier than ten years ago.

237 Should Cystic Fibrosis patients be transplanted using "suboptimal organs": insights from decision analysis

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Background: Lung transplantation is an important treatment option for Cystic Fibrosis (CF) patients with end stage lung disease. Organs are limited and many patients die on the waiting list. One option for increasing organ availability is to use organs that are suboptimal and have been optimised via an ex-vivo management programme. Patients can find it difficult to decide whether to accept extended pool organs. Patients accepting such organs may have a lower probability of dying on the transplant list but a greater risk of dying after transplant. Decision analysis (DA)is a technique that allows complex and uncertain decisions to be dealt with systematically. The clinician provides expertise by identifying the possible outcomes and their probability and a description of the health state associated with each outcome. The patient provides expertise by placing a value on the outcomes described. The decision tree then integrates the probabilities of the various outcomes and the patients values for those outcomes in order to calculate an optimal decision.
Methods: We used a medline search to identify outcomes for conventional and extended donor pool transplantation. We used clinical experience to describe the resultant health states and imputed values for those states. We used Treeage software to construct a decision tree and used the roll-back function to identify the optimal decision.
Results: DA suggested that given the death rate on the waiting list in the UK CF patients would probably choose to receive organs from an extended pool.
Discussion: Though patients may differ in the valuation of outcomes the tree allows those values to be incorporated and the DA to be re-run.

238 Osteoporosis and vertebral fractures in CF patients after lung transplantation

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Objective: CF have marked loss of bone mass caused by malnutrition, lower sex hormones, inactivity and chronic inflammation.
In literature the prevalence of osteoporosis is about 30% and compression fractures is 7–35% in adult CF. But no osteologic data exist, when CF are exposed to a high immunosuppressive and glucocorticoid therapy after lung transplantation (CFTX).
Methods and Results: We therefore retrospectively analysed clinical data, bone radiographs and dual-energy X-ray absorptiometries (DXA) of 21 CF (9 m/12 f; age: 28±9 yrs), transplanted between 01/99 and 08/04. The mean follow up was 2 yrs (range: 0.1–8.5 yrs). Only 1 CFTX (4.7%) had normal T-values, 3 CFTX (14.3%) had osteopenia, 11 CFTX (52.4%) had osteoporosis and 6 CFTX (28.6%) had vertebral fractures. 50% of vertebral fractures were diagnosed within the first year after lung transplantation. Bone mineral density was markedly decreased (lumbar vertebral column-mean: 0.97 g/cm^2; T score: –2.9 – femur mean: 0.73 g/cm^2, T score –2.64) and predictive for compression fractures. Parathyroid hormone (PTH) was elevated in 82%, osteocalcin in 45% and 25-Hydroxyvitamin D was low in 36%. The creatinine clearance in CFTX was 56.4 ml/min in mean. Despite of high physical activity and adequate osteoporosis therapy no significant increase of bone mass could be seen after CFTX.
Conclusions:

1. All adult CF should be screened for osteoporosis as early as possible.
2. A biphosponate/calcium/vitD therapy in CF with osteopenia or osteoporosis before LuTX and after CFTX, in combination with physical activity and optimal nutrition may reduce osteoporosis and vertebral column fractures within the first year after CFTX.