Inhaled hypertonic saline effect on nutritional status

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Aims: The effect of inhaled hypertonic saline (HS) on mucus-ciliar clearance in Cystic Fibrosis (CF) patients is well known. Several studies evaluated its efficacy on lung function and number of exacerbations. In our study we evaluated if its use may have any positive effect on nutritional status in CF patients.

Methods and Patients: In CF patients receiving HS 7% (5 ml twice/day) for at least 18 months, a retrospective evaluation of FVC, FEV1, and BMI pre- and during treatment period (T0, and every 6 mo) was performed. 14 CF patients (8 males) aged 7–25 yrs (mean age 17±2 mo) who received HS for 18–36 months consecutively (mean duration of treatment 32 mo) entered the study. Chronic Pseudomonas aeruginosa infection was present in 9 patients, 1 also presented ABPA, 3 patients have CFDR. Four out of 14 received nutritional supplementation (1 enteral, 3 oral) during the study period.

Results: At starting HS only 2/14 patients had a FEV1 lower than 70% predicted. During treatment FEV1 and FVC remained stable or showed a slight improvement in 11/14 patients. As regard nutritional status a good improvement was noted in 11/14 patients with mild-moderate CF lung disease. Evaluation of growth and nutritional status in a larger number of patients receiving long-term HS is needed to confirm this preliminary observation.

Conclusions: Long-term HS has a positive effect on nutritional status in adolescent patients with moderate-late CF lung disease. Evaluation of nutritional status in a larger number of patients receiving long-term HS is needed to confirm this preliminary observation.

Risk factors and prognostic impact of bronchial artery embolization in adults with cystic fibrosis: a nested case–control study

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Rationale: To identify risk factors and long-term outcomes associated with bronchial artery embolization (BAE) for hemoptysis in adults with cystic fibrosis.

Methods: Consecutive adult CF subjects who received BAE in Cochin Hospital (Paris, France) were individually matched (1:4) for age, gender and FEV1 with CF subjects who did not have BAE. Multivariate conditional logistic regression was used to identify independent factors associated with BAE. Transplant-free survival and decline in lung function were analyzed using Kaplan-Meier and mixed model analyses, respectively. Cox proportional hazards model were used to examine impact of embolization requiring BAE on survival.

Results: From 1994 to 2008, 48 subjects had BAE (BAE+) and were matched with 183 subjects (BAE−). Multivariate analysis indicated that variables associated with disease severity (number of IV antibiotic days/yr and long-term oxygen therapy) and bronchial colonization (Aspergillus sp. or mycobacteria) were independent risk factors for BAE. Although BAE+ patients had shorter transplant-free survival, BAE was not an independent factor associated with shorter survival. No difference in FEV1 and FVC decline were identified between both groups.

Conclusion: Aspergillus colonization or mycobacterial infection are risk factors for BAE. Patients who required BAE had more severe pulmonary disease and shorter survival that was unrelated to embolization. No effect of embolization requiring BAE was found on rate of decline in lung function.

The prevalence of pulmonary hypertension in patients with end stage cystic fibrosis

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Introduction: Lung transplantation (LTx) is a widely accepted treatment for end stage cystic fibrosis (CF). The prevalence of secondary pulmonary hypertension (PH) in CF patients is estimated with ultrasound to be 21–49%. But the gold standard, heart catheterization, is used in few studies because of the invasive nature. The existence of PH increases the perioperative risk. Therefore, we measured mean pulmonary artery pressure by right heart catheterization, in CF patients screened for LTx.

Methods and Materials: CF patients evaluated for LTx between 2001 and 2008 were included. We excluded patients with incomplete or conflicting data. Right heart catheterization was performed according to our protocol. PH was defined as mean pulmonary artery pressure greater than 25 mmHg at rest with a pulmonary capillary wedge pressure of 15 mmHg or less. The forced expiratory volume in one second was determined at moment of catheterization.

Results: 73 CF patients were screened for LTx and 68 were included (of whom 93% accepted for LTx). Median age was 31 (range 16–57) years at moment of screening. During the study period, 35 patients were transplanted (56%) after a median waiting time of 10 months (range 0–49). 16 CF patients (24%) were diagnosed with PH by heart catheterization. No significant differences were observed between the groups with or without PH regarding to age, gender and number of enlisted patients. Lung function was significantly lower in the group with PH (p = 0.003). No correlation between lung function and mean pulmonary artery pressure was found (p = 0.09).

Conclusions: This study demonstrated a prevalence of 24% for pulmonary hypertension in CF patients screened for LTx.