cells over-expressing NHERF1. These data suggest that NHERF1 participates in the organization of the actin cytoskeleton through activation of RhoA and ezrin.

**Reference(s)**


**CF Units:** 3Napoli, 4Milano, 5Parma, 6Cesena, 7Cerignola, 8Firenze, 9Brescia, 10Genova, 11Soverato; 12UOS of Epidemiology, Ospedale Bambin Gesù, Rome, Italy

**Background:** Cystic Fibrosis (CF) is a chronic disease characterized by an increased energy demand. Nutritional management is difficult for several clinical and psychosocial reasons but an early detection of malnutrition is essential for a better prognosis. Different auxologic indexes have been used to identify malnutrition but, presently, a gold-standard is not yet available.

**Aims:** To compare (a) growth charts of Italian CF patients and normal subjects, and (b) different auxologic indexes in evaluating malnutrition as a function of age.

**Patients and Methods:** We studied 941 CF patients aged 0–18 years (M 51.1%, mean age 9.2±6.4 years) followed-up in 10 Italian Reference Centres. Height-for-Age percentile (HAP), Weight-for-Age percentile (WAP) and BMI were calculated in all patients and used to draw the correspondent growth charts in two sexes. We compared the course of 50th percentile (pc) in CF patients and normal subjects (CDC 2000). We also calculated Weight-for-Length pc (WLP) in patients <2 yrs and BMI pc (BMlp) in patients aged 2–18 years. We compared the course of HAP, WLP and BMlp in patients with real malnutrition, alone (HAP <5th, WLP <10th and BMlp <10th) and together with those at risk for malnutrition (HAP <10th, WLP <10th and BMI <10th).

**Results:** HAP, WLP and BMI 50th pc in CF patients were lower than normal subjets, in both sexes and at all ages considered. HAP and WLP had a similar course (HAP <5th: 14%; BMI <10th: 12%; HAP <25th: 35.1% and BMI <25th: 35.7%). BMIpc <10th seemed a better index than HAP <5th, mainly in the pre-pubertal age (2–5 yrs: 14.3 vs 7.1%; 14–18 yrs: 23 vs 21.9 %, respectively). On the other hand, the frequency of BMpc <25th was higher than HAP <25th only in the first years of life (2–5 yrs: 42.2 vs 33.7 %; HAP <25th progressively became more frequent than BMI <25th (14–18 yrs: 55.8 vs 38.8 %, respectively). Our results show that Italian CF children have subnormal growth at all ages. Not a single but a combination of different indexes may be useful to correctly identify and better manage malnutrition.

**Conclusions:** This Poster has been presented also as Oral Communication, by invitation of the Scientific Committee.

**P45 CORRELATION BETWEEN STATURE AND FEV1: AN ITALIAN MULTI-CENTRIC STUDY ON 620 CF PATIENTS AGED 6–18 YEARS**

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**Background:** Shorter patients are much more likely to die before taller patients. Based on these data, short stature should not be considered benign to patients with cystic fibrosis. Several studies have also well established that children with normal lung function have a significantly better nutritional status. Stature is an easy criteria to screen patients at risk of malnutrition. Influence of nutritional status on lung function has never been assessed in Italy.

**Aim:** To evaluate the correlation between stature, age and FEV1 in Italian CF patients.

**Patients and Methods:** We enrolled 620 CF patients aged 6–18 years (F 51%, mean age 11±5.3 yrs) followed-up in 10 Italian Reference Centres. Patients were divided into 3 age-groups (6–9, 10–14 and 15–18 yrs). We compared these groups by calculation of Height-for-age Z-score (HAZ) and lung function expressed as FEV1% predicted.

**Results:** Trend of relationship between FEV1 and HAZ is shown in figure 1.

**Conclusions:** Also in Italian CF patients HAZ decreases with age, as well as FEV1. Early identification and treatment of malnutrition is essential to avoid an adequate supporting feeding and to prevent a long-term worse outcome, in terms of quality of life, lung status and, ultimately, survival.

**P46 COMPARISON AMONG AUXOLOGIC INDEXES TO EVALUATE NUTRITIONAL STATUS OF INFANT AND CHILD CF PATIENTS AND CREATION OF CF GROWTH CHARTS: A MULTI-CENTRIC ITALIAN STUDY**

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**Background:** Recurrent Pancreatitis as the first manifestation of cystic fibrosis is a rare event. The purpose of the present study was to report the experience of a single centre in the evaluation of RIP occurrence and treatment.

**Aim:** To report the experience of a single centre in the evaluation of RIP occurrence as the first manifestation of CF.

**Patients and Methods:** We studied, retrospectively, 51 young patients (27 F, mean age at diagnosis: 11.1±6.7 yrs) affected by recurrent acute pancreatitis and showing signs of chronic pancreatitis. All patients were submitted to ERCP to exclude bilo-pancreatic malformations. Patients without malformations underwent sweat test and genetic analysis: CFTR gene was evaluated by DHPLC method and the most common mutations of PRSS1 and SPINK1 genes were also analyzed.

**Results:** A bilo-pancreatic malformation was diagnosed in 12 patients (23.5%: 5 choledochus cyst, 2 pancreas divisum, 2 duodenal duplication, 2 choledochus...
P25 CAN CYCLOSPORINE TREATMENT AFFECT CYSTIC FIBROSIS PULMONARY DISEASE? A CASE REPORT

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Case: A 26-year-old female cystic fibrosis (CF) patient (R334W/852del2L2) came to our CF referral Center in 1994. She presented with symptoms of prolonged acute respiratory infections and recurrent episodes of pneumonia, and was treated with ciprofloxacin and aminoglycosides. Sputum swabs samples were cultured and airway colonization with Pseudomonas aeruginosa and Staphylococcus aureus was diagnosed. Pulmonary function tests documented a decrease in the mid-maximal flow rate, reflecting small airways obstruction and the mean forced expiratory volume in 1 sec value was 46% of predicted. Restrictive changes, characterized by declining total lung capacity and vital capacity were also observed. Computed tomography (CT) images of the chest showed that airway and parenchymal changes were present throughout both lungs. Multiple areas of extensive bronchiectasis, atelectasis, mucus plugging and defects of pulmonary vessels perfusion were seen in the left upper lobe and in the right lower lobe. The patient required several hospital admissions for pulmonary exacerbations and was regularly examined at 2-month intervals. Her CF was complicated by renal insufficiency (serum creatinine levels 4.4 mg/dl) which required long-term dialysis therapy. In March 2004 she underwent live-related donor renal transplant and started an immunosuppressive therapy with cyclosporine. CT images taken two years after transplantation documented a significant size reduction of bronchiectasis in the upper left lobe and a complete regression of atelectasis and bronchiectasis in the lower right lobe. There were also signs of pulmonary perfusion improvement in both lungs. The patient had lower mean serum creatinine values and regained the weight lost during dialysis. Her pulmonary function improved from 46% predicted to 65% predicted. Since transplantation, she did not require any hospitalization for pulmonary exacerbations and the interval between routine visits was lengthened. This case highlights the fact that renal transplantation associated with an organ-specific treatment may have beneficial effect in terms of improvement of lung damage progression in CF patient with renal failure. We also suggest that a multicentric study may be proposed to treat with cyclosporine patients affected by CF pulmonary disease.

P26 TELEMONITORING FEV1 AND PEF IN CYSTIC FIBROSIS PATIENTS VERSUS FPR AMBULATORY

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Introduction: The aim of this research has been to study the efficacy of the telemonitoring-assistance-therapy in Cystic Fibrosis patients, comparing it to the standard medical care and therapy. We compared the homemade spirometry data obtained by a monitoring system given to the CF patient and sent to a Data Centre, using a telephone connection, with those made in our medical ambulatory, during a clinical control.

Methods: For this demonstration we studied the values of FEV1 and PEF of 18 CF patients. The first spirometry, with following transmission, has been made during the medical control in our ambulatory at the time of enrollment. All the used instruments have been set according to the ATS standardisation. The data have been collected using the Filemaker-Pro-7 database and elaborated with the software Epi-Info 6.00.

The t-test student has been used to assess the data with a p-value <0.05.

Results: From the first data it seems that, for many patients, the examined spirometry values are higher in the ambulatory made tests than in the homemade ones, obtained without any medical control. The comparison among the average of FEV1 does not show any statistically different between the two groups (t=0.971, p-value = 0.3412). Those patients that precisely followed with care the protocol of the study, at the end obtained similar spirometry as at home as at the FPR ambulatory. A further confirmation of this consideration comes from the comparison of PEF values that is statistically significant (t=2.3415 p=0.0278). This means that the collaboration of the patient is different at home from the one offered in the ambulatory.

Conclusions: In conclusion we can state that the medical control is useful to perform the spirometry but with the acquisition, along the time, of the homemade telemonitoring know-how, the patient becomes less dependent from the medical presence and instructions.

P50 ATYPICAL CYSTIC FIBROSIS (ACF): AN EXPERIENCE OF A CYSTIC FIBROSIS CENTER

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Introduction: During last decade an increased number of patients (pts) have been diagnosed as ACF. According to the consensus opinion (Thompson 2006:6:627-635) searching for ACF should start with sweat test (ST) in presence of suggestive symptoms. If the ST is normal or border-line, a panel of at least 31 mutations may help to diagnose ACF. A complete genetic analysis (GA) is indicated when only one mutation is detected. Abnormal bioelectrical properties of nasal/intestinal epithelium have to be performed in controversial cases.

Aim: To describe clinical and laboratory findings compatible with ACF in a cohort of pediatric population followed at a CF Center.

Patients and Methods: CF Database data of 180 pts aged 1–18 years (y) identified by symptoms after 1990 at Care Center were retrospectively reviewed. Inclusion criteria for ACF were: sweat chloride level (SCL) >60 mEq/l, clinical manifestations suggestive of CF at first visit, two mutations or one mutation/polyT/S127G variant on both alleles by GA. 6 pts were ruled out despite suggestive symptoms and ST <60mEq/l, because of only one detected mutation and so classified as “CFTRpathies-correlated”. GA was based on a two level approach, as a first analysis of 31 mutations by ASO dot blot followed by the scanning of the whole gene by direct sequencing.

Results: Based on clinical findings, ST and GA 9 pts (4M, mean age at diagnosis 6.7 y, 18m-15y) were enrolled. All pts had pancreatic sufficiency (PS); mean value of first FEV1 (5/9) was 100, 3% with no PA colonization. 2/9 had pancreatitis as first manifestation. Mean value of SCL was 38.9 (24–50). 4/9 had severe mutation/polymorphism (PolyT/S127G).

Legend: RRI recurrent respiratory infections; FT1 failure to thrive; P polyps; S sinusitis; F familiarity; HI Haemophilus Influenzae; SA Staphylococcus Aureus; PA Pseudomonas Aeruginosa; B/P bronchitis/peribronchitis; NHM nasal mucosal hypertrophy; NP non pathogens; N negative.

Conclusions: Based on frequency of ACF diagnosis in adults, ACF should be assessed also in children with suggestive symptoms of CF by SCL and GA.

P51 ANALYSIS OF FACTORS PROMOTING BONE DISEASE IN CYSTIC FIBROSIS


Introduction: Many Authors observed that cystic fibrosis (CF) patients often are at risk of fractures. Our aim was to establish factors involved in bone disease in CF.

Methods: We enrolled 21 patients affected by CF (15 F and 6 M, age range 11–42 years). They have been distinguished for age, sex, respiratory function, pancreatic function, serum calcium levels, 25OH vitD, PCR, T score and Z score, and DXA scores. BMD Evaluation: Normal (T score between +1 and −1), Osteopenia (T score