

between  $-1$  and  $-2.5$ ), Osteoporosis (T score  $<-2.5$ , with presence of fractures). We used the Wilcoxon/Kruskal Wallis statistical test for analysis.

**Results:** First, we divided our patients in those with Osteopenia ( $n=15$ ) and those affected by Osteoporosis ( $n=6$ ). Mean FEV1 values were significantly higher in patients affected by Osteopenia than in those affected by Osteoporosis ( $p=0.05$ ). We found a positive correlation between BMI and Z score ( $r=0.55$   $p=0.0097$ ), suggesting that a good nutritional status affects the value of Z score. We then divided patients based on pancreatic status. There was a positive correlation between FEV1 and Z score for pancreatic insufficiency, while subjects with pancreatic sufficiency and mild clinic expression showed an association between VitD and Z score, which may reflect a low patient's compliance.

**Conclusions:** The results of the study on 21 CF patients show that there is a positive correlation between BMI and bone disease related to CF. Patients with low BMI had pathologic values of T score and Z score. The correlation between BMI and bone disease corroborates the idea that nutritional status is an important prognostic factor. Our findings emphasize the role of multiple factors in Cystic Fibrosis phenotype and the importance (especially for young patients) of achieving and maintaining a good nutritional status, associated to a good therapeutic compliance, in order to avoid the negative evolution of pulmonary disease and related bone disorders.

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### P52 ROLE OF INFECTIOUS PULMONARY COMPLICATIONS IN LUNG TRANSPLANTATION FOR CYSTIC FIBROSIS: 10 YEARS FOLLOW-UP

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Between November 1996 and December 2006, 64 patients (32 M, mean age  $25.6 \pm 10$  years; 32 F mean age  $25.1 \pm 6.3$  years) with end-stage cystic fibrosis underwent bilateral sequential lung transplantation.

Before transplantation patients' airways were colonized by *Pseudomonas aeruginosa* (52 pts), *Staphylococcus aureus* (7 pts) and *Burkholderia cepacia* complex (5 pts).

After transplantation patients were observed at 1, 5, 8 and 10 years, examining the culture of sputum or bronchoalveolar lavage fluid, the serological test for CMV and the rate of intravenous therapy following bacterial lower airways respiratory infections.

Of these 64 pts, 34 (54%) are actually alive. 30 pts (46%) died, but only 4 deaths (1.2%) were caused by infections: 3 pts died in the early post operative period for sepsis, 1 pt died 14 months after transplantation for *P. carinii* infection, probably due to poor compliance to therapy. Opportunistic infections of the lung with *P. carinii* occurred in other 2 pts: both responded to treatment.

Early after transplantation the same germs that were present before the transplantation grew again in the culture of sputum or BAL.

The rate of pts colonized by bacteria was 92% at 12 months, stabilized at 100% at 5, 8 and 10 years. The rate of pts with CMV infection was 74% and 28% respectively at 1 and 5 years, zeroing at 8 and 10 years; in our Center CMV infections were treated in preclinical stage, only 1 pt developed CMV gastric disease, diagnosed by biopsy. 11% of pts had positive sputum or BAL culture for *Aspergillus fumigatus* during the first year, 7% of pts at 5 years and 22% at 8 years. 2 pts with bronchiolitis obliterans syndrome had evidence of pulmonary aspergillosis at autopsy and 1 pt had *A. fumigatus* infection of the right bronchial anastomosis.

3 pts with *B. cepacia* colonization who survived the early post operative period did not have a higher infections rate or a worse outcome than pts not colonized by the germ.

The rate of intravenous therapy following bacterial respiratory infections was 5%, 20%, 44% respectively at 1, 5, 8 and 10 years after transplantation.

In conclusion, high risk periods for developing infections are the 1st year post transplantation, when immunosuppressive regimen is intensive, and from 5th year on, because of the reinduction immunosuppressive therapy after BOS.

In these pts meticulous follow-up, prompt recognition and treatment of pulmonary infections are essential to prevent poor outcomes.

### P53 COLONIC CARCINOMA ON CF PATIENT

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Case Report: GG, 37-year-old woman, complete form of CF (genotype F508del/G542X), stable clinical and nutritional trend (BMI 18%, FEV1 42%). Chronic colonization by *Pseudomonas*, frequently haemoptysis, subjected to bronchial ADA for three times. Ursodeoxycolic acid therapy for chronic liver disease and asymptomatic cholelithiasis.

In July 2006, a sudden abdominal pain appears to the right upper quadrant. Physical examination shows a treatable abdomen, pain occurs by hard tapping at the right upper quadrant, moderate hepatomegaly. Plain abdominal X-Ray: normal; laboratory tests: moderate microcytic hypochromic anemia.

Ultrasound shows a roundish mass in VII-VIII segment of the liver,  $6 \times 5$  cm in diameter, dishomogeneous with central liquified area. The abdominal CT confirms the presence of roundish formation with diameter 5.6 cm, hypodense in basal condition with central zone of colliquation (Abscess? New formation of colliquation?).

Due to diagnostic antibody-titer for *Entamoeba Histolytica* (1:120) and detection of trophozoites in the drainage liquid, the patient starts therapy with Metronidazole and Dehydroemetine.

Abdomen MRI: confirms previous CT report. In January 2007, because of deterioration of general condition, a colonoscopy is carried out that shows a big blackish vegetating mass; this is easily bleeding with the size about 5 cm, ascribable to the intestinal location of amoebic formation. Later, an adenocarcinoma was diagnosed based on histological examination.

In February 2007, the patient undergoes operation of right hepatectomy (resection of three lesions in IVa, IVb and III segments) and right hemicolectomy. After operation, pleuric effusion solves after drain. One month later, she starts antitubercular drug therapy.

At a distance of 8 months the patient is treated by 6 cycles of chemotherapy that she tolerates well, with a progressive improvement in general condition.

CF is a chronic pathology which causes an increase in risk of developing some pathologies, such as colon carcinoma with a 6.4 RR.

GI symptoms, particularly if they are persistent or uncountable, have to be always carefully monitored, with particular attention to increased preponderance of GI tumours in patients with CF.

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### P54 NUTRITIONAL AND RESPIRATORY EFFECTS OF AZITHROMYCIN LONG-TERM TREATMENT IN CYSTIC FIBROSIS PATIENTS

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In Cystic Fibrosis (CF) patients azithromycin (AZM) in multiple trials has demonstrated to improve lung function, but there is a large variability in study design, dosage and outcome measures among these trials. Moreover, long-term studies on chronic effects of AZM are scanty.

The aim of the study was to evaluate the changes in pulmonary function and nutritional status in CF patients over 12 months of AZM treatment.

We retrospectively collected the data of 50 patients on daily 250 mg AZM continuous therapy; out of these we examined the data of 25 subjects (males 9, mean age 25.2, range 8–56 years, 50% chronically infected by *Pseudomonas aeruginosa*) who concluded a period of 12 months of treatment.

At the baseline and every 3 months we analyzed the changes of BMI, CV, FEV1 and FEF25–75 by repeated measures analysis of variance. A p value of 0.05 was considered as significant.

	Value at month				
	0 (baseline)	3	6	9	12
BMI	19.8±3	20.2±3.2	20.4±2.8	20.5±3.1	20.4±3.0
CV	79.1±20.2	82.7±19.1	86.6±18.9	85.9±18.2	80.6±19.6
FEV1	61.0±20.7	63.6±17.8	66.3±16.3	66.4±15.6	63.7±21.2
FEF25–75	32.5±22.9	33.9±20.9	34.6±22.3	34.4±19.2	36.4±27.3

The table shows the mean values (+ SD) of BMI, CV, FEV1 and FEF25–75 at baseline, 3, 6, 9, 12 months of treatment. The trends are statistically significant for both BMI ( $p < 0.01$ ) and CV ( $p < 0.02$ ), but not for FEV1 and FEF25/75.

Our results show that AZM long-term treatment may improve the nutritional status and respiratory function in CF patients. In addition, the improvement reached after 3 months remains stable throughout the whole year of treatment.