186 Omalizumab in the treatment of ABPA in 32 patients with cystic fibrosis

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Objectives: ABPA (Allergic bronchopulmonary aspergillosis) affects up to 15% of patients with cystic fibrosis (CF). The treatment of this disease, except corticosteroid, remains debated. Only a few clinical cases of omalizumab treatment have been reported in this disease.
Methods: This retrospective multicenter observational french study includes 32 patients (15 childrens and 17 adults) with cystic fibrosis treated with omalizumab for more than three months in the context of ABPA. Clinical characteristics, concomitant medications (inhaled and oral corticosteroids, antifungal), changes in lung function, BMI, serum IgE, side effects, previous treatments and the cost of treatment were collected from patients, one year before the start of treatment and during the first year of treatment.
Results: No significant differences could be demonstrated with regard to lung function, BMI, or the number of patients receiving oral corticosteroids. At the time of initiation of omalizumab, 59% of patients received oral corticosteroids versus 30% after one year of treatment. 78% of patients had received antifungal therapy at the time of the introduction of omalizumab. Treatment tolerance was good (19% of patients experience side effects). The total average cost of treatment was 3456.70 euros per patient per month.
Conclusion: Limitations of this retrospective study concerns the number of patients with a wide disparity of use between different centers. In addition, the cost of this treatment is high. In view of these results, it seems necessary to conduct prospective studies to determine potential indications for omalizumab in patients with CF with an ABPA.

187 Spirometry in pediatric patients with cystic fibrosis. Are we adequately reporting?

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Objectives: Examine whether FEV_1 and FEV_1/FVC percent predicted values in cystic fibrosis (CF) patients (pts) differ between the classical used Knudson equations and those proposed by the Global Lungs Initiative (GLI).
Methods: We used data from a group of paediatric pts from a CF Clinic at a tertiary Hospital. Information extracted from lung function (LF) database: age, sex, height, weight, FEV_1 and FEV_1/FVC (in L) and percent predicted values according to Knudson equations (%K). One observation per patient was included. LF was obtained and classified according ATS/ERS guidelines [Jaeger Masterscreen Body Plethysmograph (Viasys Healthcare, GermanyV.5.22.1)]. The absolute values were automatically analysed through the GLI_Sheet Calculator and the new values registered as %GLI. Pts were assigned in groups according to age (5–12 and 13–18 yrs) and LF severity. Statistical analysis: Paired-sample T Test.
Results: 44 pts [(21 (52.3%) male; median age: 12 (5–18) years].

Knudson vs GLI results

	Knudson	GLI	p value
FEV1 (Mean, SD)	85.1 (21.7)	76.6 (24.5)	0.00
FEV1/FVC (Mean, SD)	78.9 (11.0)	78.9 (11.0)	0.676

Significant difference for mean percent predicted FEV_1 according to the reference equations used was found ($p = 0.00$), even when subgroups by age and severity were tested. There was no difference forFEV_1/FVC ($p = 0.485$). For 9 pts LF severity changed after reanalysis, 5 from mild to moderate and 4 from moderate to moderate-severe.
Conclusion: Our study shows that significant differences exist depending on the reference equations used. This may induce misclassification of severity of LF in CF patients, impact is yet to be determined.

188 Association of lung function with adherence to inhaled antibiotic treatment in patients with cystic fibrosis (CF)

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Rationale: Ineb AAD records information about the use of the device. It's authorized for administration of colistimethate sodium (CMS) in CF patients (Promixin®, Profile, UK). The main aim of the study was to describe the adherence to the inhaled antibiotic treatment and lung function in CF patients.
Methods: Retrospective study carried out at 10 CF Spanish centers. Patients with CF colonized with *Pseudomonas aeruginosa* (PA), in treatment with CMS with Ineb for at least 12 months, were included. At baseline, lung function, anthropometric data and long-term treatments were recorded. During 12 months, health outcomes and treatments were obtained from medical records by quarters. Information about Ineb use was downloaded to calculate adherence to the treatment. ANCOVA was performed to compare mean differences.
Results: Data from 108 patients with CF were analyzed (mean age: 24.5±10.2 years; range: 4–57; 38.9% female). Mean inhaled antibiotic adherence was 65.9±34.7%. Table shows association between four measures of adherence and the %FEV1 predicted at baseline and the change in %FEV1 predicted in 12 months adjusted for %FEV1 predicted at 1st quarter.

Association between adherence and FEV1

	Adherence	True adherence	% days Ineb was used at least once	% days Ineb was used at least twice
%FEV1 pred Baseline	$p = 0.0037$	$p = 0.0050$	$p = 0.0012$	$p = 0.0003$
Dif %FEV1 pred in 12 months (adjusted for %FEV1 pred in 1st quarter)	$p = 0.30$	$p = 0.37$	$p = 0.29$	$p = 0.072$

Conclusions: In this population of CF patients we find an association between adherence to the inhaled antibiotic treatment and the %FEV1 predicted at baseline but not at 12 months. Only the association with the % of days the Ineb was used at least twice tends to statistical significance.

189 Nocturnal pulse oximetry in paediatric cystic fibrosis patients. Is FEV_1 a good predictor?

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Objectives: To identify nocturnal hypoxemia (NH) in CF children and to determine if spirometry is a good predictor.
Methods: Twenty-five stable patients (pts) underwent a sleep questionnaire (SQ) and an overnight pulse oximetry (OPO). Oxygen dessaturation index (ODI) was classified as mild $\geqslant$3/h, moderate $\geqslant$6/h or severe $\geqslant$10/h. Correlation with spirometry was performed. Values are presented as percent predicted (FEV1, FVC, FEV_1/FVC ratio and FEF_{25-75}). Statistical analysis: linear regression and χ^2.
Results: Twelve (48%) male pts; median age 13 (7–18) years. Mean ODI was 4.5/h (13 normal, 6 mild, 3 moderate and 3 severe). Spirometry was normal in 6 pts (normal ODI in 3).

Table: ODI/Spirometry results

ODIMean (SD)/n patients	FVC Mean (SD)	FEV1 Mean (SD)	FEV1/FVC Mean (SD)	FEF_{25-75} Mean (SD)
4.8 (0.45)/6	101.4 (17.53)	95.4 (24.1)	81.1 (11.81)	63.8 (33.8)
7.9 (1.53)/3	88.4 (14.31)	82.0 (21.2)	77.9 (8.9)	43 (24.5)
15.4 (5.06)/3	84.5 (28.80)	79.0 (28.1)	79.1 (6.11)	34.2 (21.6)

No correlation was found between OPO and spirometry values. However, subgroup analysis showed correlation between moderate ODI and all spirometry values ($R^2 > 0.90$) and severe ODI and FEV1/FVC and FEF_{25-75} ($r^2 = 0.957/r^2 = 0.668$). Respiratory disturbed sleep arose from SQ in 50% of pts but no correlation was found with ODI severity.
Conclusions: This study shows that oxygen dessaturation during sleep is common in paediatric CF patients. At least for moderate or severe disease spirometry may be a better predictor of NO than history alone. Cut off spirometry values for OPO studies must be determined.