154 Cystic fibrosis and hypogammaglobulinemia: is there a role for subcutaneous immunoglobulin substitution?

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Introduction: High levels of IgG in Cystic Fibrosis (CF) are associated with more severe chronic lung disease. Although there are no data on subnormal IgG levels and prognosis, underlying humoral immunodeficiency could potentially aggravate CF lung disease.
Objectives: To document retrospectively whether subcutaneous immunoglobulin substitution (SCIG) had a positive influence on the clinical course in children with CF and hypogammaglobulinemia (HG).
Methods: We analyzed the medical charts of 5 patients with CF and HG who received SCIG between Jan 2010 and Dec 2010 because of frequent and severe respiratory infections despite intensive CF treatment. Data up to 1 year before start of SCIG were compared with data up to 1 year after start SCIG.
Results: All patients were homozygous for F508del. Median IgG z-score before start SCIG was –3.11 (IQR –3.25; –2.96). Average days of antibiotic (AB) use per month before and under SCIG was not different (23.3 vs 30 days) mainly due to use of oral maintenance AB. There was a trend towards a reduced need for intravenous AB under SCIG (1.8 vs 0 days/month). There was no reduction in number of positive culture swabs after SCIG (25 vs 67%). Although BMI z-score increased in 4 out of 5, this was not statistically significant.
Conclusion: Based on our limited experience with SCIG in patients with CF and HG, we cannot document a clear effect on respiratory outcome. This may be due to the use of maintenance oral AB and lack of good outcome measures. These data will be compared with data of 5 patients with CF and HG not receiving SCIG. Further study on the etiology of HG in children with CF and need for treatment is needed.

155 Do composite scores of nNO and FENO improve diagnostic value?

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Nasal NO (nNO) is used as screening test for Primary Ciliary Dyskinesia (PCD). nNO and exhaled NO (FENO) are low in PCD and CF.
We studied the discriminate validity of nNO and FENO or their combination for the diagnosis of PCD.
Patients with PCD, CF, asthma (A), humoral immunodeficiencies (HID) and healthy controls (HC) were recruited. 6 nNO and 2 FENO measurements, using the chemiluminescence analyzer Spiroware 3.0® (Eco Medics) were performed during slow exhalation against a fixed resistance. ATS/ERS criteria were achieved.

Results nNO – FENO

	HC (n = 24)	PCD (n = 23)	CF (n = 22)	Asthma (n = 28)	HID (n = 19)
Age (median-IQR) (yrs)	13.05 (6.83)	11.77 (7.35)	13.13 (7.46)	10.23 (4.88)	9.84 (5.76)
nNO (median-IQR) (ppb)	723.7 (236.9)	44.2 (112.1)	421.7 (371.1)	725.5 (637.6)	763.6 (693.9)
FENO (median-IQR) (ppb)	10.4 (9.7)	4.6 (3.9)	9.9 (23.6)	13.5 (24.7)	11.0 (12.1)

For nNO, median coefficient of variation (CV) was 7%, similar in subgroups. nNO differed between PCD and no PCD (p 0.0001) and between PCD and CF (p 0.0001). nNO <270 ppb had a sensitivity of 91.3% and a specificity of 90.6% to diagnose PCD in the total group (n = 116) and a sensitivity and specificity of 87.5% and 77.3% with only PCD and CF included. nNO was similar in HID and HC (p 0.448). There was overlap between PCD and CF, and even A, but almost no overlap between PCD and HC and HID.
Compared to HC, FENO was lower in PCD (p 0.0001), but not in CF, A and HID. FENO was lower in PCD than in CF (p = 0.005). To diagnose PCD, FENO <7.1 ppb had a sensitivity of 90.9% and a specificity of 68.2%. Adding FENO values to nNO values in composite scores did not improve discriminate validity.
nNO as well as FENO is significantly lower in PCD than in CF. But for the diagnosis of PCD, composite scores of FENO and nNO do not improve discriminate validity.

156 Influenza and Swine Flu vaccination coverage in adult cystic fibrosis

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Background: Infection with influenza viruses can cause significant morbidity in patients with Cystic Fibrosis (CF). In 2009, a novel Swine Flu strain (H1N1) resulted in a pandemic and the WHO recommended that all patients with chronic illnesses should be given both the Influenza and Swine Flu vaccines. We have previously demonstrated that the Influenza vaccine uptake rate amongst CF adults was 75% [1].
Aims: To assess Influenza and Swine Flu immunisation rates during the 2009–10 season within our adult CF cohort.
Methods: Retrospective questionnaires regarding immunisation status and reasons for non-adherence were given to all patients who attended the CF clinic and ward between November 2010 and February 2011.
Results: A total of 102/240 responses were received. 79/102 (78%) and 69/102 (68%) patients were immunised with the Influenza and Swine Flu vaccines, respectively. Reasons for non-adherence included forgetfulness (23%), needle phobia (5%) and being too unwell (9%). Interestingly, more patients declined the Swine Flu vaccine for fear of side effects than the Influenza vaccine (22% vs 9%).
Conclusions: The immunisation uptake rate in this cohort was lower for Swine Flu than for Influenza but still significantly higher than for other clinical risk groups in England (68% vs 37%) [2]. A further audit of the combined Influenza/Swine Flu vaccine is planned later this year.

Reference(s)

[1] Wat D et al. J Cyst Fibros. 2010: 9, S117 (449).
[2] http://www.dh.gov.uk/en/Publicationsandstatistics (DH_114203).