173 Upper respiratory tract colonization in infants with cystic fibrosis diagnosed by newborn screening

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Cystic fibrosis, is characterised by production of abnormally thick and viscous mucus. It results in disturbances of mucociliary clearance, a basic defensive mechanism of the respiratory system, leading to increased susceptibility to recurrent and chronic respiratory infections in over 90% of patients.
The aim of this study is to evaluate nasopharyngeal cavity colonization in infants with cystic fibrosis diagnosed by newborn screening.
Material and Methods: Nasal and pharyngeal swabs were obtained from 26 infants aged 10–20 weeks, without any signs of infections, who were examined in the Cystic Fibrosis Outpatient Clinic. Isolates were identified by means of routine diagnostic methods.
Results: Negative nasal swabs were obtained in 10 (40%) infants, negative pharyngeal swabs – in 6 (23%), and in 4 patients both nasal and pharyngeal swabs were negative. In the nasal swabs the most frequently observed growths were: *Staphylococcus aureus* MSSA(+) in 9 (34.5%), *Corynebacterium* sp. in 6 (23.5%), and β-lactamase(+) *Moraxella catarrhalis* in 2 (8%). Pharyngeal swabs revealed α-hemolytic *Streptococcus* in 20 (80%), *Staphylococcus aureus* MRSA(+) in 7 (27%), *Neisseria* sp. in 4 (15%), *Escherichia coli* ESBL(−) and *Candida albicans* in 3 (11.5%) examined infants.
Conclusion: Nasopharyngeal cavity colonization by pathogenic bacteria in infants suffering from cystic fibrosis occurs during the first months of life, in the absence of noticeable signs of upper respiratory infection.

174 The phenotype of nasal polyposis in cystic fibrosis patients after lung transplantation

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Objective: Nasal polyposis (NP) is common in cystic fibrosis. Some reports have suggested that patients with NP have an improved pre-transplant survival. The influence of the NP-phenotype (NPP) on survival and bronchiolitis obliterans syndrome (BOS) after lung transplantation (LTx) is unknown.
Aim: To determine the prevalence of the NPP and its influence on survival and BOS after LTx.
Methods: All CF patients transplanted at our center between Nov 92 and Dec 09 were included. Survival and BOS following LTx were compared in the NPP with the non-NPP by Kaplan Meyer statistic.
Results: The 89 evaluated patients (44 females, 49%) had a mean age of 26.8±8.2 years at LTx, pre-LTx FEV1 of 1.0±1.8 liters (26±8%) and estimated 5-year survival without LTx of 33±14%. 17 (19%) patients had NP. No difference was found regarding gender, pretransplant FEV1, estimate survival without LTx, pretransplant *Pseudomonas* infections, pretransplant prednisone treatment, pretransplant diabetes and mutation on the CFTR-gene in the two phenotype. Overall 1-, 5- and 10-years survival was 90±3%, 71±5%, 59±6%. One, 5- and 10-years survival in the NP-phenotype was 94±6%, 77±10%, 61±13%, compared to 89±4%, 69±6%, 59±7%. No significant difference was found in the development of BOS in the two groups (freedom of BOS 1 at 5-year 88±7% in NP-phenotype compared to 78±5%). However, there was a tendency of less BOS stage 1 in the NP-phenotype.
Conclusions: In our cohort of CF patients with LTx nasal polyps are found in one fifth. Patients with nasal polyps have the same survival compared to non-NP patients. There is a tendency of less BOS in patients with NP. This is probably a specific finding for the NP-phenotype.

175 Sinonasal disease in adult CF patients: prevalence and management in Wales

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The nasal and paranasal sinuses are contiguous with the lower respiratory tract. CF patients commonly experience sinonasal disease which is thought to have the same aetiology and pathophysiology as the chronic lung condition. Evidence suggests that sinonasal disease may directly influence the lower airways in CF.
Purpose:
i. To identify the prevalence of adult CF patients with sinonasal disease in Wales
ii. To review assessment and treatment strategies used in patients with sinonasal disease.

Method: A retrospective review of sinonasal disease was conducted. Data were collected from annual assessments between 2010 and 2011. All patients had a confirmed diagnosis of CF and were under the care of the All Wales Adult CF Centre. The following data were recorded: age; sex; FEV_1; genotype; sputum microbiology; sinonasal disease status (sinusitis or polyposis), sinonasal symptoms (pain, congestion, discharge, postnasal drip); referrals and investigations (ENT, CT, sinus sampling); medication; nasal washouts.
Results: 162 records were reviewed. Mean age 28 (17–53 yrs). 81 reported history of sinonasal disease; 23 had both; 29 referred to ENT; of which 12 had polyps; 5 had sinus CT; 48 prescribed nasal steroid; 4 antibiotics; 28 antihistamine; 1 analgesia; 0 sympathomimetics, 3 nasal rinse and no patients underwent sinus sampling. Severities of phenotype or FEV_1 were not related to presence of sinonasal disease in this group.
Conclusion: The prevalence of sinonasal disease in our centre appears significantly lower than previous studies. This service evaluation highlights the need for the development of a protocol in the assessment and management of this patient group.

176 An audit of the diagnosis and treatment of ABPA in a UK adult CF centre

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Objectives: The diagnosis of ABPA in CF is hampered by both poorly standardised diagnostic criteria and its often non-specific clinical presentation. The Cystic Fibrosis Foundation consensus conference in 2003 produced minimum diagnostic criteria for ABPA. There have been no randomised controlled trials of treatment in CF but corticosteroids are usually prescribed, oral antifungals are often added. We sought to determine our compliance with these minimum diagnostic criteria and UK CF trust guidelines for treatment.
Methods: A retrospective case note review identified patients with a current or prior ABPA diagnosis. Their diagnostic tests were compared to minimum criteria and treatment regime recorded, 40 control patients without ABPA were compared.
Results: 10 of 132 patients (7.6%) had been given a diagnosis of ABPA. Only 3 met all 4 minimum criteria at the time of diagnosis, the remaining 7 met 2 or 3. Five of the patients had a total serum IgE level >500 iu/ml. Mean values of total IgE and Aspergillus specific IgE were significantly higher in ABPA cases than the 40 controls but IgG precipitin titres were higher in the control group. The treatment initiated was variable. All cases were prescribed steroids but dosage and duration recommended was variable, 5 patients were started on anti-fungals.
Discussion: The diagnosis of ABPA in our centre seldom met minimum diagnostic criteria and treatment regimes were variable. Our experience is unlikely to be uncommon. This may arise due to poorly defined UK diagnostic recommendations and the lack of robust evidence to guide treatment. A new ABPA protocol has been implemented in our centre to address these inconsistencies.