141 Platelet aggregation capability in children with cystic fibrosis

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Objectives: To determine platelet aggregation capability for various variants of antigene system AB0 in patients with cystic fibrosis (CF).
Methods: 55 CF children (homozygous and heterozygous on F508 del mutation) were enrolled. 46% girls and 54% boys had mild form of CF, 54% girls and 46% boys had severe form. We investigated the function of platelet aggregation with: thrombin, adenosinediphosphate (ADP) and arachidonic acid (AA).
Results: Patients with severe form of CF most frequently (29%) had 0 (I); B (III) have not been revealed in this group, meanwhile 22.2% patients with mild form of CF had A (II) and AB (IV) have been revealed. Platelet aggregation with ADP has significant differences between blood groups A (II) and 0 (I), ($p = 0.019$). Values were ranged 2.5–97.5 percentile in patients with 0 (I) and 2.5–25 percentile in A (II). Data of aggregation with AA did not show significant differences between groups.
Significant difference between homozygous F508 del and heterozygous F508 del patients was shown in aggregation with trombin ($p = 0.03$). There is a tendency to hyperaggregation in heterozygous patients meanwhile homozygous had both hypoaggregation and hyperaggregation. Difference on other tests between homozygous and heterozygous patients is not significant.
Conclusion: First results of this study should be interpreted according to system ABO, severity of disease and genotype in patients with CF. These aspects play an important role for individual treatment.

142 Immunisation coverage in children with cystic fibrosis: a French multicenter survey

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Objectives: European guidelines for immunization in Cystic Fibrosis (CF) children have been published in 2005. However, no survey of immunization in this population has been reported yet.
Methods: We performed a survey to analyze immunization coverage and timeliness of vaccines in children with CF. Data were extracted from a study which aimed to analyze H1N1 influenza pandemy in 439 patients with CF followed in the CF referral centers of the Paris metropolitan area.
Results: 134 children aged between 9 months and 17.5 years were enrolled in the study. 43% of children had at least one F508del mutation. 83% of the children had been vaccinated with BCG. Coverage for diphtheria–tetanus–pertussis (DTP), polio (Pol) and Haemophilus (Hi) approximated 100% at 24 months but decreased to 57% at 14 years. 85% of the children had had Measles–Mumps–Rubella (MMR) vaccine at 18 months. Hepatitis A and B vaccination coverage at 24 months was poor (respectively 15% and 50% of the children). Hepatitis B coverage did not improve with age (55.2% at 14 years). Only 50% of the children had completed a full pneumococcal vaccination schedule. Immunization coverage for seasonal flu was 72% the year of the study. Very few children were immunized for varicella and conjugate meningococcal C (13% for both), papillomavirus vaccine and 11 rotavirus vaccine (less than 1%).
Conclusion: Immunization coverage was satisfactory for DTP, Pol, Hi and MMR. Children with CF should be strictly monitored for vaccinations for hepatitis A and B, pneumococcus and seasonal flu.

143 Seasonality of *Pseudomonas aeruginosa* isolation in patients with cystic fibrosis

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Objectives: Early infection with *Pseudomonas aeruginosa* has been associated to higher morbidity and mortality in patients with Cystic Fibrosis. A better knowledge of the factors that are involved in its early infection can be useful to prevent its acquisition.
Methods: Retrospective study in children diagnosed with Cystic Fibrosis. We have analysed demographic variables, genotypes and positive cultures to *P. aeruginosa*, and considered as winter from January to March, spring from April to June, summer from July to September and autumn from October to December. We practiced at least 4 cultures per year to each patient.
Results: We evaluated 95 children. From them, 54 (56.8%) had at least 1 positive culture to *P. aeruginosa* and 35 (36.8%) also had a second positive one. The mean age for the first positive culture was 24 months (2–86 months). 9 of the patients developed the positivity during the winter time (16.66%), 14 during the spring (25.9%), 17 in summer (31.5%) and 14 in autumn (25.9%). 35 children had a second positive culture with a mean age of 47.5 months (8–153 months). The second isolation was observed in winter in 5 children (14.3%), in spring in 10 children (26.8%), in summer in 8 children (22.8%) and in autumn in 12 children (34.3%).
Conclusion: Both first and second isolation of *P. aeruginosa* are less frequent in winter, being more prevailing in summer and autumn months. This suggests that seasonality as a factor that contributes to its acquisition.
The identification of factors that take part in its acquisition such as seasonality, could help us to implement recommendations to prevent this infection.

144 The impact of *Pseudomonas aeruginosa* colonization in CF

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Colonization by *Pseudomonas aeruginosa* (PA), the most prevalent organism isolated from CF patients' sputum is associated with an accelerated clinical and functional deterioration and is an important predictor of mortality.
Aim: To assess the clinical and functional differences between CF patients colonized by PA and other CF patients with distinct bronchial colonization.
Methods: Retrospective analysis of the adult CF patients followed at our specialized CF centre, between 2008–12. Clinical files were reviewed in order to collect the following data: demographics, genotype, sputum colonization, lung function (FEV_1), nutritional status (BMI). Bronchial colonization was defined according to the Leeds criteria. Patients submitted to lung transplant or with irregular follow-up were excluded. Statistical analysis was done with SPSS 20. The level of statistical significance considered was 0.05.
Results: We included 34 patients in this study, 54% were females, median age 30.8 ± 8.5 years. del508F was the most common mutation found (25% homozygous). Colonization by PA was found in 68%, *S. aureus* in 65% (36% MRSA), *B. cepacea* in 9% and fungus in 47%. We found positive correlations between: female gender and MRSA colonization; colonization by PA and fungus. No correlation was found between: bronchial colonization and BMI. We estimated Kaplan–Meier survival curves using FEV_1 as a surrogate marker, then we applied the log rank test and found that mucoid PA colonized ($p = 0.043$) patients had a faster FEV_1 decline.
Conclusion: We concluded that colonization by mucoid PA has a great impact on disease severity, has seen by a faster FEV_1 decline and consequently adding mortality risk.