Abstract # 30230

MBL DEFICIENCY

DEGREE OF MANNOSE-BINDING LECTIN DEFICIENCY AND IMPACT ON PULMONARY DISEASE IN CHILDREN WITH OTHERWISE UNKNOWN CAUSES OF RECURRENT OR CHRONIC RESPIRATORY PROBLEMS FOLLOWED AT A NATIONAL PULMONARY SERVICE

Kathrin W. Dahl#, Frederik Buchvald#, Astrid Thomas#, Peter Garred§ and Kim G. Nielsen#.
#Pediatric Pulmonary Service, Dept of Pediatric & Adolescent Medicine, and §Dept of Clinical Immunology, Copenhagen University Hospital, Rigshospitalet, Copenhagen, Denmark
Contact: Kathrin W. Dahl kwd@dadlnet.dk

Background

Mannose binding lectin (MBL) is an important protein in innate immunity. Low expressing MBL alleles (MBL genotype) - associated with low MBL concentration -, have been related to trivial recurrent respiratory tract infections in otherwise healthy pre-school children.

Aim

Investigate whether there might be a potential association between MBL genotype and
- Morbidity
- Structural lung damage
- Pulmonary function

in a heterogeneous group of children referred to tertiary pulmonary center because of recurrent lung infections.

Methods and Material

Retrospective cross-sectional cohort study

Children referred to our tertiary pediatric pulmonary service between 2006 and 2011 and in which MBL was included in the investigation program.

Diagnoses that could explain recurrent lung infections were ruled out:
- Cystic Fibrosis
- Primary Cilia Dyskinesia
- Immuno Deficiencies.
- Interstitial Lung Diseases
- Congenital Malformations

Differences in morbidity (use of antibiotics and lung function) and relative risk (RR) of structural pulmonary changes on CT imaging (atelectasis; bronchiectasis and air trapping) in relation to MBL genotype were assessed.

Results

Baseline Characteristics

N	113
Males, n (%)	67 (59,3)
Median Age (range) years	3,04 (0-16)

Diagnosis

Unexplained recurrent lunginfections	89,5 %
Cardiac disease	3,5 %
Others	7,0 %

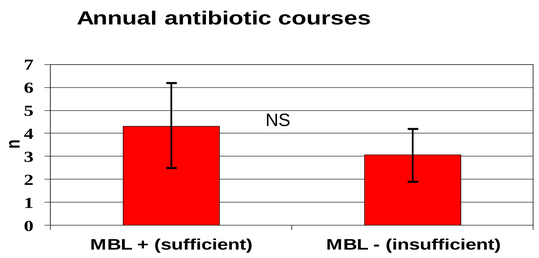
MBL Genotype Distribution

MBL sufficient	A/A	62 (55 %)
MBL insufficient		51 (45 %)
Intermediate MBL	YA/0	24 (21 %)
Severely impaired MBL	XA/0	16 (14 %)
No functional MBL	0/0	11 (10 %)

Structural Lung Damage

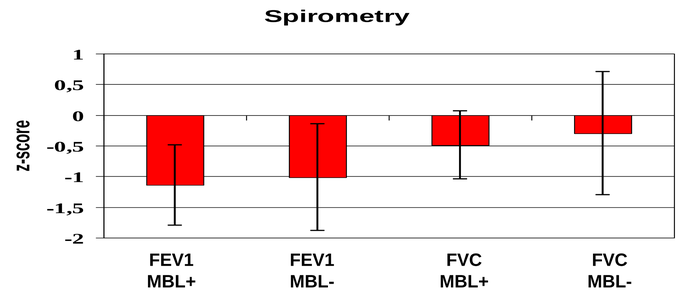
RR (95% CI) of structural lung changes at CT imaging was 1,03 (0,7;1,5), when MBL serum concentration was considered low.

Morbidity



No significant difference in number of annual antibiotic courses

Pulmonary Function



No significant difference in lung function could be demonstrated between normal and low serum concentration of MBL, respectively. However for younger children there is a tendency for difference as mean sRaw (airway resistance) (z-score) was higher in MBL insufficient group than in the MBL sufficient children (2.06 vs. 0.86; p=0.16).

All results were similar, when the population was grouped according to MBL variant alleles associated with different level of low serum concentration of MBL.

Conclusion

Pulmonary morbidity or structural pulmonary changes on CT was not associated with low expressing MBL genotype in a heterogeneous group of children with recurrent lung infections referred to a tertiary pulmonary center.