The aim of the study was to evaluate the diagnostic and prognostic value of anti-MCV in JIA comparing to anti-CCP.

Methods: 30 children (19 girls and 11 boys; aged 4 -18 years) with confirmed JIA diagnosis and 20 children as a control group were included into the study. Anti-CCP and anti-MCV antibodies in sera were measured using ELISA test.

Results: Sensitivity and specificity of the anti-CCP were 40% and 100% compared with 36,7% and 90% for the anti-MCV calculated using manufactures recommended cut-off values.

Anti-MCV were positive in 11/30 comparing with 12/30 for anti-CCP in children with JIA. Among 11 JIA children positive for anti-MCV, 5 were also positive for anti-CCP and among 18 JIA children negative for anti-CCP, 6 were positive for anti-MCV. Both antibodies were mainly observed in polyarthritis, however could be positive in other types of JIA.

Anti-MCV serum concentration correlated positively with anti-CCP (p=0,0018). Anti-CCP correlated positively with disease activity (p=0,00196), radiological destruction in joints (p=0,00196) at baseline and after median 11,5 months of follow up (p=0,00961), but these correlations were not observed concerning anti-MCV (respectively: p=0,0657; p=0,06199; p=0,064).

Conclusions: Anti-MCV as well as anti-CCP antibodies could be helpful in the diagnosis of JIA. However, the anti-CCP prognostic value in JIA appears to be superior to the anti-MCV test.

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GORHAM-STOUT SYNDROME IN AN 8 YEAR OLD BOY WITH CHYLOTHORAX

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Object: We present an 8 year old boy with Gorham-Stout Syndrome. This rare disorder occurs at all ages and is characterized by osteolysis, haemangioma and lymphangiectasia. A chylothorax can be present and is associated with a poor prognosis. It is very important to recognize this rare disorder, because a letal outcome might be avoided by starting interferon therapy.

Case presentation: The boy was admitted for respiratory distress due to left sided chylothorax. Malignancy and infection were excluded. Extensive imaging studies were inconclusive at presentation. Treatment with special nutrition, somatostatines, left sided Denver-drain, clipping of the thoracic duct and prednisone did not help. The chylothorax became bilateral.

Further imaging studies with MRI and CT two months after admission, revealed signs of osteolysis in the scapula, clavicle and ribs, suggesting Gorham-Stout syndrome. A biopsy was done and showed elevated numbers of CD34 positive cells. Blood was send to analyse the levels of IL 1, IL 6 and VEGF.

Treatment with Interferon a2a and biphosphate led to an amelioration of the clinical situation but not to complete recovery of the chylothorax. Only after placement of a second Denver-drain at the right side, the patient could be discharged from our hospital. He is seen in our outpatient clinic since 6 months and his condition is stable.

Conclusion: Interferon a2a and biphosphate treatment in combination with a Denver drain halted the deterioration caused by the chylothorax due to Gorham-Stout syndrome. This diagnosis and treatment should be considered in children with chylothorax of unknown origin.

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THE EXPRESSION OF PRO-INFLAMMATORY CYTOKINE IL-17 IN NEONATES

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Introduction: Neonates, and more so preterms, are known to have deficient adaptive immune responses, critical for host defense mechanisms against pathogens. Adaptive immune responses are mediated by the activation of pathogen-specific T helper type 1 (Th1), Th2 lymphocytes and, the recently indentified, Th17 cells. Th17 lymphocytes produce the pro-inflammatory cytokine IL-17 which provides protection against mainly extra-cellular pathogen infections. However IL-17 release has been hardly studied in neonates. The aim of our study was to investigate the expressions of IL-17 in the serum of term and preterm newborns and compare them to those of adults.

Patients and methods: Thirteen (13) healthy preterm neonates [birth weight (BW): 1740g (1500-2200), gestational age (GA): 32,5wk (31-34)] and 13 healthy term neonates [(BW: 2960g (2500-4310), GA: 37,5wk (37-39)] were studied. Six (6) healthy adults were used as controls. Peripheral blood samples were obtained from neonates during the 2nd and 3rd weeks of life. IL-17 levels were measured in the serum by ELISA.

Results: Serum IL-17 levels did not differ between preterm and term neonates. However, IL-17 levels were significantly lower in both preterm and term neonates, as compared to those of adults ($p > 0,001$ for both terms and preterms).

Conclusion: Our data reveal dramatically decreased IL-17 release in the serum of neonates compared to adults, mirroring the immaturity of the neonatal immune responses. Importantly, our findings suggest that deficient IL-17 release in neonates may hamper their host defense against extracellular pathogens often leading to overwhelming septicemia and death.

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HEREDITARY ANGIOEDEMA AND LONG-TERM PROPHYLAXIS

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Hereditary angioedema is a rare autosomal dominant disease characterized by recurrent episodes of angioedema caused by a quantitative or functional defect of the plasma protein C1 esterase inhibitor (C1 INH). Treatment is divided into short and long-term prophylaxis with androgens, antifibrinolytics and C1 inhibitor replacement.

We present the clinical characteristics of three female patients, two of them members of the same family and one isolated case. Eleven year old and 5 year old sisters were admitted to our hospital with complaints of relapsing skin swelling. In the second patient abdominal pain attacks were also developed. The third patient, a 13 year old girl was suffered from recurrent bouts of swelling most often affecting extremities and genitalia since 7 year old. We analyzed blood levels of C3, C4, and C1 INH. Both of the sisters had decreased serum levels of C4 and C1 INH and were diagnosed as hereditary angioedema type I. The third patient had reduced serum levels of C4 and normal C1 INH and the genetic analysis on the C1INH gene showed a mutation of aa 444 Arg substituted by Cis consistent with type II hereditary angioedema. All the patients were started on tranexamic acid for long term prophylaxis but in the first and second patients the treatment had to change with danazol due to the lack of enough response to the tranexamic acid. The third patient presented occasional (1-2 mild episodes per year) peripheral angioedema and her disease was controlled with tranexamic acid.

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UNDER AGAROSE MIGRATION ASSAY - A METHOD TO STUDY GRADED CHEMOTACTIC ACTIVITY OF LEUKOCYTES IN NEWBORN INFANTS

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Background: Preterm newborn infants are at risk of developing chronic inflammation and tissue injury in their lungs and CNS due to prolonged and exaggerated accumulation of leukocytes.**Aim:** To study migration of leukocytes from newborn infants with a modified under agarose cell migration assay.

Methods: Blood was collected from healthy adults (n=9) and from healthy, term newborn infants (umbilical cord; caesarean section; spinal anaesthesia; n=9). Isolated leukocytes were added to outer wells and chemoattractants to central wells. Gels were incubated for 2 hours whereby only polymorphonuclear (PMN) leukocytes migrated. Dose responses of intermediate (n=4; IL-8) and end-target (n=5; fMLP) chemoattractants were tested on PMN.

Results: Similar dose response curves of PMN migration to gradients of IL-8 and fMLP were observed in newborn infants as in adults, but with less PMN migrating with newborns than with adults. The PMN leukocyte migration per distance in newborns was 80% of adults with IL-8, and >90% of adults with fMLP. There was a high linear correlation between the number of PMN and the distance of migration irrespective of chemoattractant studied (IL-8: $r=0.85$ and $r=0.65$ respectively for newborns and adults; $p< 0.001$ for both) (fMLP: $r=0.76$ and $r=0.87$; $p< 0.001$). The pattern of migration differed between fMLP and IL-8.