surgery, whereas the number of $CD38^+$ cells was sharply reduced, which indicates depletion of compensatory capabilities of children after surgery.
CONCLUSIONS: Low content of activation markers, especially $CD95^+$ cells before surgery and $CD38^+$ cells after surgery, is an unfavorable prognostic sign in children with CHD and concurrent thymomegaly.

SYSTEMATIC REVIEW OF DIAGNOSTIC CRITERIA AND CLINICAL FEATURES OF FAMILIAL HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

Submitted by Caroline Gholam
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INTRODUCTION: Familial lymphophagocytic histiocytosis (FHL) is an autosomal-recessive immunologic disorder that is fatal if untreated. The quoted incidence is 1.2 in 1 000 000; fewer than 1 of 3 patients are diagnosed while alive. The true incidence of FHL may be much higher.
OBJECTIVE: With this project we aimed to identify consensus information required for early recognition and diagnosis of patients with FHL to initiate life-saving treatment.
METHODS: An extensive Medline search that covered the last 20 years produced 17 relevant articles. The hemophagocytic lymphohistiocytosis 2004 protocol produced by the American Histiocyte Society provided additional information.
RESULTS: From this review, the consensus diagnostic criteria for FHL are (1) familial disease/known genetic defect, (2) clinical and laboratory criteria (5 of 8 criteria), (3) fever, (4) splenomegaly, (5) cytopenia in $\geq$2 cell lines, (6) hypertriglyceridemia and/or hypofibrinogenemia, (7) high ferritin level, (8) high levels of soluble CD25 (interleukin 2 receptor), and (9) hemophagocytosis in bone marrow, cerebrospinal fluid, or lymph nodes. Results of tests of initial bone marrow aspirate may be inconclusive, and repeated ones may be necessary. Half of the patients have abnormal cerebrospinal fluid findings. Several symptoms and laboratory findings provide supportive evidence.
CONCLUSIONS: Diagnostic criteria and supportive features are consistent throughout literature and are aided by the recent addition of genetic and protein-based testing. Diagnostic difficulty lies in the lack of pathognomonic features or specific diagnostic tests for FHL. Not all features present at the initial stage. Treatment should be initiated in cases of strong clinical suspicion.

MANNOSE-BINDING LECTIN (MBL) GENE POLYMORPHISMS AND SERUM MBL LEVELS IN CHILDREN WITH RECURRENT RESPIRATORY TRACT INFECTION

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INTRODUCTION: Mannose-binding lectin (MBL) is a plasma collectin and is considered an important component of innate immunity. Its plasma concentration is, for the most, part genetically determined by a series of single nucleotide polymorphisms located both in the structural gene and in the promoter region. MBL deficiency may be associated with increased susceptibility to infectious disease and autoimmune disorders.
OBJECTIVE: Our goal was to establish the reference serum level of MBL in children, investigate the correlation between *MBL* gene polymorphisms and its serum level in Chinese Han nationality, and *MBL* gene polymorphisms and serum level in children with recurrent respiratory tract infections.
METHODS: The concentrations of oligomerized MBL in plasma were measured by enzyme-linked immunosorbent assay, and *MBL* gene polymorphisms were analyzed by polymerase chain reaction-restriction fragment length polymorphism and polymerase chain reaction with sequence-specific primers.
RESULTS: The median MBL level in 470 normal children was 2536 ng/mL (range: 0–7860 ng/mL), and $P_{2.5}$ to $P_{97.5}$ was 161 to 5070 ng/mL. Two promoter polymorphisms, -550 and -221, and coding variants at codon 54 of the *MBL* gene affected the protein level significantly, and the most frequent genotype in Hans was *HYPA/HYPA*. Serum MBL levels were significantly lower in patients with recurrent respiratory tract infections (RRTIs) compared with healthy controls ($H = 6.661$; $P < .05$), and the frequency of the promoter *LXP* haplotype was significantly higher in patients with RRTIs than in controls ($\chi^2 = 4.71$; $P = .03$). The prevalence of the B allele in patients with RRTIs was higher than that in controls, but the difference did not reach significance ($\chi^2 = 0.18$; $P > .05$).
CONCLUSIONS: The MBL reference value in China is 161 ng/mL. Children with MBL concentrations of <161 ng/mL, therefore, were deemed to be MBL deficient, and *LXP* is a risk factor for recurrent respiratory tract infections in this population.

RESEARCH OF MANNOSE-BINDING LECTIN AND INTERLEUKINS 10, 12, AND 18 IN CHILDREN WITH HENOCH-SCHÖNLEIN PURPURA

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INTRODUCTION: Henoch-Schönlein Purpura (HSP) is a common vasculitis in children, and the unbalance of T-helper 1/T-helper 2 plays an important part in its pathogenesis. Mannose-binding lectin (MBL) is an important component of innate immunity and related with a lot of diseases with immunologic derangement. However, we do not know the relationship between MBL and HSP.
OBJECTIVE: We aimed to explore the serum level and gene polymorphisms of MBL and the levels of interleukin 10 (IL-10), IL-12, and IL-18 in supernatant of peripheral blood mononuclear cells of children with HSP and HSP nephritis (HSPN) and of healthy children.
METHODS: The concentrations of MBL and IL-10, IL-12, and IL-18 were measured by enzyme-linked immunosorbent assay: *MBL* gene polymorphisms were analyzed by polymerase chain reaction-restriction fragment length polymorphism and polymerase chain reaction with sequence-specific primers.
RESULTS: The serum MBL levels in the 23 children with HSP were not significantly different from 27 children with HSPN ($P = .95$) or 18 normal children. The levels of IL-18 in the supernatant of peripheral blood mononuclear cells in the 3 groups were not significantly different from each other ($P = .47$, .15, and .14). The levels of IL-10 in children with HSP and HSPN were not different from each other ($P = .70$), but both were significantly different from those in the normal children ($P = .04$ and .01). The levels of IL-12 in children with HSP were different from those in the children with HSPN ($P = .04$). The MBL promoter genotype and the frequency of alleles were not different between the children with HSP and HSPN or between those 2 groups compared with the normal group.
CONCLUSIONS: IL-12 probably plays an important role in the renal involvement in HSP. The position of MBL in the pathogenesis of HSP and HSPN remains to be confirmed.

RESPIRATORY SYNCYTIAL VIRUS INDUCED MORE SERIOUS INFECTION AND INFLAMMATION IN NUDE MICE THAN IN BALB/c MICE

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INTRODUCTION: Respiratory syncytial virus (RSV) infection is ubiquitous and leads to severe disease in immunocompromised individuals.
OBJECTIVE: Our goal was to compare RSV infection and inflammation between immunocompetent BALB/c mice and immunodeficient nude mice.
METHODS: Pulmonary viral titers, histology, immunohistochemistry for CD14 and CD56, leukocyte counting, and cytokines were assayed by enzyme-linked immunosorbent assay in bronchoalveolar lavage fluid.
RESULTS: RSV titers peaked on the third day after inoculation in both types of infected mice. Infected nude mice had higher-level and more durative viral replication, more severe pulmonary histopathology, and a larger number of leukocytes in bronchoalveolar lavage fluid than infected BALB/c mice. Infected nude mice displayed more pulmonary ($CD14^+$) macrophages (114.34 ± 20.24 vs 75.46 ± 12.37; $P = .05$) and ($CD56^+$) natural killer cells (37.87 ± 8.07 vs 11.06 ± 5.37; $P = .05$) than infected BALB/c mice. RSV infection enhanced production of tumor necrosis factor α, interleukin 12 (IL-12), interferon γ, and IL-10 in both types of mice. Infected nude mice had a higher level of tumor necrosis factor α (40.30 ± 7.34 vs 24.24 ± 9.54; $P = .05$), IL-12 (83.96 ± 12.32 vs 68.21 ± 7.42; $P = .05$), and IL-10 (125.01 ± 18.97 vs 77.56 ± 9.01; $P = .05$) than infected BALB/c mice.
CONCLUSIONS: RSV-infected nude mice are a good model for assessing severe and persistent infection in individuals at high risk. RSV-induced inflammation is not parallel to the immune response of T cells, and macrophages and natural killer cells contribute to severe infection and inflammation of RSV-infected cellular-immunodeficient individuals.

Infectious Diseases

DIAGNOSTIC VALUE OF IN SITU POLYMERASE CHAIN REACTION IN CHILDHOOD LEPROSY

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OBJECTIVE: Our aim was to assess the diagnostic value of in situ polymerase chain reaction (PCR) in leprosy, particularly for enhancing histopathological diagnosis.
METHODS: We prospectively studied 20 children (aged <16 years) with leprosy. Clinical examination of each case was performed, and skin smear for acid-fast bacillus was prepared. A biopsy of the lesion site was performed

RESEARCH OF MANNOSE-BINDING LECTIN AND INTERLEUKINS 10, 12, AND 18 IN CHILDREN WITH HENOCH-SCHÖNLEIN PURPURA

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