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

Submitted by Qiu Li
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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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