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Introduction: Hypogammaglobulinemia is defined as a plasmatic level of immunoglobulin G (IgG) under 5 g/L. Although some of them are linked to primary immune deficiency (PID), majority are secondary immune deficiency (SID).

Objective: Since there are a few studies about this subject, we though interesting to precise frequency, severity, treatment and outcome of patients with hypogammaglobulinemia in a large population including children and adults.

Methods: We performed a retrospective analysis of 336 patients (38 children less than 18 years and 298 adults) diagnosed in 2005 with hypogammaglobulinemia in the University Hospital in Angers.

Results: Mean IgG level was 4.8 g/L and 70 patients (21%) had a global deficiency (Ig G, A and M). Only 18 (5%) patients with PID were identified (10 common variable immuno deficiency, 5 congenital deficiencies and 3 transients hypogammaglobulinemia in infancy). Others patients (318/336) were considered as SID. Among them, hypogammaglobulinemia was associated with nephropathy (16%), myeloma (15%), lymphoma (15%), chronic lymphocytic leukemia (6%), drug-related (14 %)(immunosuppressors, antiepileptics, corticosteroids) or link to other diseases defined (18%). Finally, 16% of patients remained unclassified. 21% of patients required immunoglobulin replacement therapy and 12% bone marrow transplantation. During follow-up, 44/231 (19%) patients with SID died (not necessarily from immunodeficiency) while 3/18 (16%) patients with PID died (generally from immunodeficiency).

Conclusions: Hypogammaglobulinemia requires a rigorous diagnosis approach. While majority of hypogammaglobulinemia in children correspond to PID, most of them in adults are SID and prognosis depends on underlying disease.

532 CD19 DEFICIENT B CELL ABNORMALITY AND SELECTIVE IGM DEFICIENCY IN A MALAY CHILD. A CASE REPORT

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Introduction: Cases of CD 19 deficiency from South East Asia has rarely been reported. A, 11 yr Malay boy was referred with recurrent infections with 2 episodes of 'chickenpox at age 2 years. He had repeated episodes of pneumoniae and bronchiectasis by age 6 years. Physical examination revealed gross clubbing with crepitations at both lung bases. IV Ig administration was instituted with clinical improvementLaboratory data: Serum Ig (g/l)IgG 8.37[n 4.95- 16.56], IgA 0.92[n 0.30-2.35], IgM 0.21[0.32-1.40]Lymphocyte subset (age 11) CD19 0 %, CD20 11.26% (12-22)CD3 83.64% (n 66-76) ,CD4 36.25 (33-41),CD8 41.55 (27-35), CD16+56 11.6 %. (9-16),Specific antibody response to polysaccharide antigen was impaired ; whileNBT , lymphocyte proliferation to (PHA) were normal, Btk protein 18% (control 87.8 %). Our finding conforms with a diagnosis of CD19 deficiency with selective IgM deficiency and partial defective Btk protein expression. CD19 deficient B cells with hypogammaglobulinemia is a rare disorder. Up to 2007 only 5 cases had been reported1. All the 5 CD19 deficiency cases reported (Van Zelm MC et L (2006) & Kanagena H et al (2007)]had more than 2 Ig isotype level reduced . Our patient with had only Ig isotype(igM) reduced. The proposed functional abnormality is the disruption of CD19 signalling resulting in a primary antibody deficiency mainly characterised by a poor antigen specific response. Our findings has added further dimension to elucidation of the functional abnormality in CD19 deficiency and variable isotype immunoglobulin deficiency.

552 TLR2-MEDIATED CO-STIMULATION OF CVID T CELLS

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Introduction: Activation of CVID B cells via TLR7 and TLR9 is impaired. The TLR2 ligand Pam3CSK4 has been shown to activate helper T cells and induce IFN-γ production. The capacity of CVID T cells to respond to TLR2 co-stimulation has not been examined yet.

Objective: The aim of the study was to investigate T