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Società Italiana di Allergologia e Immunologia Pediatrica (SIAIP)

CONIUGARE RICERCA E PRATICA CLINICA IN IMMUNO-ALLERGOLOGIA

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TITOLO: Clinical and Immunological characterization and follow-up of 7 pediatric patients affected with selective IgM deficiency (sIgMD).

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TESTO:

Introduction: Selective IgM deficiency (sIgMD) represents a rare and poorly characterized primary humoral deficiency, characterized by low/absent IgM serum levels in the presence of normal IgG and IgA levels. The reported clinical presentation is variable and literature data are mainly available for adults. To date, the pathogenesis of sIgMD remains poorly understood.

Objective: Clinical and immunological evaluation at the onset and during follow-up of 7 pediatric sIgMD patients.

Methods: 7 patients (6 males and 1 female) with sIgMD were included. Diagnosis was made according to ESID criteria. Mean age at diagnosis was 6 years and 8 months. Ig levels were evaluated periodically during the follow-up. The experimental approach included genetic analysis of the IGHM gene, peripheral blood B and T cell evaluation (flow cytometry), in vitro B cell differentiation and IgM production.

Results: Clinical presentation of affected patients was variable although all patients were predisposed to upper respiratory tract infections and otitis. During follow-up IgM levels remained constantly under normal range. Peripheral T and B subset evaluation revealed absence of IgM⁺ plasmablasts in all patients; however, no significant differences were seen in intracellular and membrane IgM expression between patients and healthy controls. B cells in vitro differentiation and IgM production was partially achievable, but impaired when compared to healthy controls.

Conclusions: sIgMD is a rare and poorly understood primary immunodeficiency. Our data suggest that different signaling pathways may be involved in its pathogenesis. Further studies are warranted to better understand this disorder.

Abstract per il Congresso

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