

Leukocyte adhesion deficiency disorder in an infant

G. SABATINO, M. DE MARTINO*, F. CHIARELLI*, D. PACIOCCO and G. AMERIO

Department of Medicine. Section of Pediatrics and Neonatology
University of Chieti, Italy.*

*Received May 25, 1999 - Accepted September 7,
1999*

Leukocyte adhesion defect (LAD) is an inherited disorder of phagocytic function. The three different α -chains of the leukocyte integrin family (CD 11a, 11b, 11c) are all dependent on the common β -chain (CD18) for proper insertion into the cell membrane. Deficiency of CD 18 thus causes an inability to express any of these proteins and results in the syndrome of LAD type 1. Clinical features include delayed separation of the umbilical cord, persistent leukocytosis and recurrent infection. A case of severe LAD occurred in a 1 month-old infant. The immunological pattern of the baby showed pathological values of CD11b and CD18 (1.3% and 0.2 % respectively) when compared with normal values (50-70%) and lack of random (1.0 nm vs normal of 10 ± 5) and casein-induced (22.0 nm vs normal of 60-120nm) chemotaxis. The baby is now waiting for bone marrow transplantation. Although LAD is a rare form of congenital immune deficiency this disease should be considered when delayed wound healing and recurrent bacterial skin infections are present in a newborn.