Detection of Antineutrophil Cytoplasmic Antibodies in the Classic Form of Hemolytic Uremic Syndrome

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Dear Sir,

The hemolytic uremic syndrome (HUS), the major cause of acute renal failure in childhood, still is a clinical entity of unknown pathogenesis.

The endothelial damage and thrombosis of glomerular vessels, the constant findings in pathological studies [1], have led to several investigations to elucidate possible patho-genetic factors involved; however, none of them have shown convincing results.

In an attempt to further study the cause of the endothelial insult, we decided to explore the potential pathogenic role of antineutrophil cytoplasmic antibodies (ANCA) in the classic form of HUS. ANCA have been demonstrated to be a good marker for renal limited and systemic vasculitis [2] and have in vitro been capable of activating neutrophils and promoting endothelial injury [3]. On this basis, we decided to study the presence of ANCA during the acute reactant phase of classical HUS.

Sera from 27 patients (14 male, average age 13.8 ± 7.4 months) with a well-documented classic form of HUS as defined by Drummond [4] were obtained during the time of admission to hospital. ANCA were investigated by indirect immunofluorescent assay (IFF). The IFF was performed as previously described by Van der Woude et al. [5] and according to the standard procedure recommended by the International Working Group of ANCA. Briefly, normal human neutrophils fixed on ethanol were used as the substrate. The patient serum was screened in a dilution of 1:20 in PBS and the bound antibody was detected by a FITC-conjugated rabbit antihuman IgG (Dako) diluted 1:32 in PBS. The study was negative in all the sam-

ples tested, in contrast to the positive results obtained with standard controls.

We conclude that the classic form of HUS does not belong to the ANCA-associated diseases, a group in which vasculitis is the common histopathological feature, ranging from patients with limited renal disease such as pauci-immune necrotizing and crescentic glomerulonephritis to those with a widespread systemic vasculitis such as Wegener's granulomatosis and microscopic polyarteritis nodosa. The evidence of antiendothelial antibodies in HUS [6] may suggest a different mechanism of endothelial damage in this disease.

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