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Nephrin Autoantibodies in Idiopathic Nephrotic Syndrome: Mechanism, Marker, Clinical Implications

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Minimal change disease (MCD) and primary focal segmental glomerulosclerosis (FSGS) in adults, along with idiopathic nephrotic syndrome (INS) in children, are immune-mediated podocytopathies leading to nephrotic syndrome. Autoantibodies targeting nephrin have been identified in MCD patients, but their roles are unclear. In a multicenter study analyzing antinephrin autoantibodies in adults with various glomerular diseases, children with INS, and controls, a total of 539 patients (357 adults, 182 children) and 117 controls were included. Among adults, antinephrin autoantibodies were found in 46 of 105 (44%) with MCD, 7 of 74 (9%) with primary FSGS, and rarely in other conditions. In children with INS, 94 of 182 (52%) had detectable antinephrin autoantibodies. In untreated patients with active MCD or INS, prevalence was 69% and 90%, respectively. Antinephrin autoantibody levels correlated with disease activity. Experimental immunization in mice induced nephrotic syndrome, an MCD-like phenotype, IgG localization to the podocyte slit diaphragm, nephrin phosphorylation, and severe cytoskeletal changes. This study indicates that circulating antinephrin autoantibodies are common in MCD and INS, serving as markers of disease activity. Their binding at the slit diaphragm induces podocyte dysfunction and nephrotic syndrome, underscoring their pathophysiological significance.

Keywords: Nephrin, Autoantibodies, Nephrotic Syndrome