

THE ROLE OF CIRCULATING IMMUNE COMPLEXES IN CHILDREN WITH EPIDERMOLYSIS BULLOSA

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Background: Systemic inflammation is considered a key factor in the immunopathogenesis of epidermolysis bullosa (EB); however, this has been described only in small and heterogeneous cohorts. The systemic nature of inflammation in EB has been confirmed by numerous studies. Circulating immune complexes (CICs) are antigen–antibody complexes that form in the bloodstream and circulate throughout the vascular system. It is known that CICs can activate the complement system, leading to the destruction of bacteria, viruses, and even host antigens. These complexes can also be phagocytosed by monocytes and macrophages, which are responsible for clearing CICs from the circulation. Based on this rationale, we analyzed serum CIC levels in children with EB to evaluate their concentrations and the inflammatory significance of these immunological markers.

Objective: To assess the diagnostic and prognostic significance of circulating immune complexes of different molecular sizes in children with EB.

Materials and Methods: The study included 42 children aged 2 to 12 years with EB. All patients were hospitalized at the Republican Center of Dermatology, Ministry of Health of Uzbekistan, in Tashkent. Clinical analysis revealed a predominance of males (51.5%), children from rural areas (66.8%), and the majority were aged 1 to 5 years (41.7%), with the remainder up to 12 years old.

Results: One of the key biological functions of immunoglobulins is antigen binding and the formation of CICs. A critical characteristic of CICs is their molecular size. In children with EB, we observed elevated average levels of both large (3% PEG) and small (4% PEG) CICs. Specifically, the levels of 3% and 4% CICs were significantly increased compared to normal values—by 7.2 and 8.5 times, respectively. The average level of large CICs (3%) was 47 ± 1.9 optical density units, with values ranging from 1 to 58, while the normative value was 6.5 ± 1.12 . Small CICs (4%) averaged 39 ± 1.2 units, ranging from 4 to 136, compared to a normative value of 4.6 ± 0.88 . These findings confirm a significant increase in both large and small CICs during disease exacerbation. Furthermore, the ratio of large to small CICs (CIC_{3%}/CIC_{4%}) shifted from a normal value of 1.1 to 1.7 in children with EB, indicating the presence of systemic inflammation.