

## Skin Manifestations in Children With Inborn Errors of Immunity in a Tertiary Care Hospital in Iran

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**Abstract :** Background: Inborn errors of immunity (IEIs) are monogenic diseases of the immune the system with broad clinical manifestations. Despite the increasing genetic advancements, the diagnosis of IEIs still leans on clinical diagnosis. Dermatologic manifestations are observed in a large number of IEI patients and can lead to proper approach, prompt intervention and improved prognosis. Methods: This cross-sectional study was carried out between 2018 and 2020 on IEIs at a Children's tertiary care center in Tehran, Iran. Demographic details (including age, sex, and parental consanguinity), age at onset of symptoms and family history of IEI with were recorded. Results :212 patients were included. Cutaneous findings were reported in (95 ,44.8%) patients. and 61 of 95 (64.2%) reported skin lesions as the first clinical presentation. Skin infection (69, 72.6%) was the most frequent cutaneous manifestation, followed by an eczematous rash (24, 25 %). Conclusions: Skin manifestations are common feature in IEI patients and can be readily recognizable by healthcare providers. This study tried to provide information on prognostic consequences.

**Keywords :** primary immuno deficiency, inborn error of metabolism, skin manifestation, skin infection

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