Background: Atopic Dermatitis is an exceedingly common inflammatory skin disease in the paediatric age. Although the future of the therapeutic landscape is promising, a number of severe cases are still frustrating to manage due to an unmet need for effective therapies.

Observation: A 7-year-old Caucasian boy presented with widespread, intensely pruritic ill-defined erythematous plaques with flexural accentuation. He also had food allergies and asthma. Immunoglobulin E (IgE) levels were high (51120KUI/L, range≤403.0KUI/L). His dermatological course was orchestrated by multiple hospitalizations, some due to mucocutaneous viral infections. Systemic steroids and immunomodulators (Cyclosporin, Methotrexate) were ineffective. An underlying primary immunodeficiency was suspected and, through genetic testing, a heterozygotic mutation in the DOCK8 gene was identified. A 375mg/m2 week, 4-week course of Rituximab was attempted. An impressive improvement was observed by the 2nd treatment, while near-complete clearance was observed 2 months after the 4th treatment (pre/post-treatment SCORAD: 75.2/12.4, respectively). Systemic steroids were discontinued with no relapses. IgE levels reduced 70% from the baseline assessment. Remission persisted for 10 months, after which a relapse led to another successful and well-tolerated course of Rituximab.

Key message: To our knowledge, we report the first case of a refractory, extensive childhood Atopic Dermatitis successfully treated with a B-cell targeted therapy. Remission was durable and no loss of efficacy was observed with retreatment. Although the immune map of Atopic Dermatitis focus on the T-cell driven inflammation, the success of this case provides rationale for further exploring the B-cell as a target for novel therapeutic advancements. Lastly, we report this patient’s heterozygotic DOCK8 mutation in the presence of his other clinical-immunological features in keeping with a DOCK8 deficiency syndrome. As the syndrome is reported as a by-product of a bi-allelic mutation or deletion, further clarification should be sought by studying the health repercussions on heterozygous carriers.