Dermatology Reports
https://www.pagepress.org/journals/index.php/dr/index

eISSN 2036-7406

Please cite this article as:

Barbato F, Licata G, Di Brizzi EV, et al. Linear IgA bullous dermatosis in a 4-year-old child. *Dermatol Rep 2026 [Epub Ahead of Print] doi: 10.4081/dr.2026.10299*

Submitted 16/02/25 - Accepted 29/01/26

Linear IgA bullous dermatosis in a 4-year-old child

Filomena Barbato,[1] Gaetano Licata,[2] Eugenia Veronica Di Brizzi,[3] Amedeo Boscaino,[4] Giuseppe Ruggiero,[5] Giovanni Calandriello,[6] Caterina Mariarosaria Giorgio[3]

[1]Department of Pediatric Dermatology, Santobono-Pausilipon Children Hospital, Naples; [2]Dermatology Unit, San Antonio Abate Hospital, Trapani; [3]Dermatology Unit, Department of Mental and Physical Health and Preventive Medicine, University of Campania "Luigi Vanvitelli", Naples; [4]Department of Pathology, Cardarelli Hospital, Naples; [5]ASL Napoli 2 Nord, Naples; [6]IRCCS Humanitas Research Hospital, Rozzano (MI), Italy

Correspondence to: Gaetano Licata, MD, Dermatology Unit, San Antonio Abate Hospital, Via Cosenza 82, 91016, Erice (TP), Italy. Tel.: 3276215976; E-mail: gaetano.licata89@gmail.com

Key words: linear IgA bullous dermatosis; child; therapeutic insights.

Conflict of interest: the authors have no conflict of interest to declare.

Ethics approval and consent to participate: not required.

Consent for publication: the patient's parents have given written informed consent for the publication of this article and any accompanying images.

Availability of data and materials: not applicable.

Dear Editor,

Linear IgA bullous dermatosis (LABD) is the most common autoimmune blistering disease in childhood, though it remains rare. It is characterized by tense blisters and vesicles, often in a rosette or “string of pearls” pattern, predominantly affecting non-inflamed skin. While LABD is typically self-limiting, recurrent or refractory cases pose significant therapeutic challenges.[1]

This case describes a 4-year-old boy with LABD, initially misdiagnosed and unsuccessfully treated with systemic corticosteroids. Long-term remission was ultimately achieved with dapsone. The case highlights the importance of early immunofluorescence testing and the role of dapsone in pediatric LABD.

A previously healthy 4-year-old boy developed widespread blistering lesions on the face, trunk, and extremities (Figure 1 A,B). The lesions were associated with minimal pruritus and no systemic symptoms. Initial diagnoses included varicella, impetigo, and atopic dermatitis, leading to empirical antiviral and antibiotic therapy, both of which failed to improve the condition.

Concerned by the persistence of lesions, his physicians initiated oral prednisone at 10 mg BID for eight days, followed by a tapering schedule. This led to a temporary resolution, but within days of corticosteroid withdrawal, new bullae appeared. Given the refractory nature of the disease, further investigations were pursued. A skin biopsy with direct immunofluorescence (DIF), performed in May 2023, showed subepidermal blistering with linear IgA deposition at the basement membrane zone, confirming LABD. Serologic and autoimmune testing, including ANA, anti-endomysial antibodies, anti-tTG, and Mycoplasma testing, were all negative. However, a throat swab for *Streptococcus pyogenes* was positive, suggesting a possible infectious trigger. Following LABD confirmation, a new treatment approach was adopted. Given the rapid relapse upon corticosteroid tapering, dapsone was introduced at 0.5 mg/kg/day, increasing to 1 mg/kg/day. Within eight weeks of therapy initiation, the patient experienced complete lesion resolution (Figure 1 C,D). Unlike previous treatments, dapsone resulted in sustained disease control, with no recurrences observed at the two-month follow-up.

Diagnosing LABD in children can be particularly challenging due to its overlap with infectious, inflammatory, and autoimmune skin disorders. Early-stage LABD often mimics viral exanthems, impetigo, or atopic dermatitis, leading to delays in proper treatment. The “string of pearls” pattern of blisters is a helpful diagnostic clue, but may be absent in some cases.[2,3] In this case, the initial misdiagnosis of varicella and eczema led to unnecessary antiviral and antibiotic treatments, delaying appropriate therapy. The persistence and recurrence of lesions after corticosteroid withdrawal subsequently prompted histopathological and immunofluorescence evaluation, confirming the

diagnosis of LABD. This highlights the importance of early biopsy and DIF testing in pediatric bullous diseases. Systemic corticosteroids can induce rapid clinical improvement in LABD; however, they are not recommended as first-line therapy, as they fail to prevent relapses and may promote steroid dependency. Dapsone is the preferred treatment due to its ability to suppress neutrophilic activity at the dermoepidermal junction.[4] Dapsone is a sulfonamide-derived immunomodulatory drug with anti-inflammatory properties, particularly effective in neutrophil-mediated dermatoses. In this patient, dapsone led to long-term disease control without the need for prolonged corticosteroid use. This supports its role as an essential treatment for pediatric LABD, particularly when the disease is recurrent or steroid-dependent.[3,4] The use of dapsone in pediatric patients is generally safe but requires monitoring for adverse effects such as hemolysis, particularly in individuals with G6PD deficiency. Regular laboratory assessments are necessary to ensure patient safety. In this case, the patient tolerated dapsone well, with no reported adverse effects.[1-3]

This case highlights the importance of early skin biopsy and DIF in children presenting with persistent or relapsing bullous dermatoses, in order to avoid diagnostic delay and inappropriate. Although systemic corticosteroids may be useful for short-term symptom control, dapsone represents the key treatment for achieving sustained disease control in pediatric LABD. The detection of *S. pyogenes* raises the possibility of an infection-associated onset, while prior antibiotic exposure cannot be excluded as a contributing factor, as drug-induced LABD has been reported. Further studies are needed to better define optimal treatment strategies and to clarify the role of infectious and pharmacological triggers in pediatric LABD pathogenesis.[3,4]

References

1. Bernett CN, Fong M, Yadlapati S, Rosario-Collazo JA. Linear IGA Dermatosis. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023.
2. Caux F, Patsatsi A, Karakioulaki M, et al. S2k guidelines on diagnosis and treatment of linear IgA dermatosis initiated by the European Academy of Dermatology and Venereology. J Eur Acad Dermatol Venereol 2024;38:1006-23.
3. Shin L, Gardner JT 2nd, Dao H Jr. Updates in the Diagnosis and Management of Linear IgA Disease: A Systematic Review. Medicina (Kaunas) 2021;57:818.
4. Wang KL, Lehman JS, Davis DMR. Linear IgA bullous dermatosis of childhood: Retrospective single-center cohort. Pediatr Dermatol 2024;41:461-4.

Figure 1. A, **B**) Clinical images of a 4-year-old boy with blistering lesions on the face, trunk, and extremities; **C**, **D**) complete disappearance of lesions after eight weeks of therapy with dapsone.