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# Annular Bullous Lesions in a Child From Uganda: Chronic Bullous Disease of Childhood

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Clinical Image

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A 10-year-old male presented to our global outreach clinic in Uganda in May 2018 with a recurrent pruritic blistering rash on the lower leg of 1 year in duration. His medical history was unremarkable for autoimmune conditions or exposure to medications. Cutaneous examination showed urticarial plaques and numerous tense bullae in an annular arrangement on the left lower leg (Figure 1). Direct immunofluorescence (DIF) demonstrated a linear band of IgA at the dermoepidermal junction, consistent with a diagnosis of chronic bullous disease of childhood (CBDC). Oral dapsone was initiated with full resolution over the following week.

CBDC is a subepidermal blistering condition driven by autoimmune production of IgA. The disease is characterized by the abrupt onset of edematous plaques and large tense bullae, typically in the first decade of life [1]. Lesions are distributed in an annular, or "crown of jewels," arrangement, and they commonly affect the trunk and limbs [1]. Although children present with a cutaneous distribution, neonates may have more mucocutaneous involvement [2]. DIF classically demonstrates linear IgA deposition along the basement membrane zone (BMZ) [3]. Although the disease is often idiopathic, genetic susceptibility, infections, drugs, and vaccinations have been implicated in the pathogenesis [1, 3]. Bullous impetigo, epidermolysis bullosa acquisita, and other autoimmune bullous disorders can be considered as differential diagnoses [1]. Although the condition can follow a relapsing and remitting course for several years, spontaneous resolution is common before puberty [4]. Treatment options may include dapsone and topical steroids.

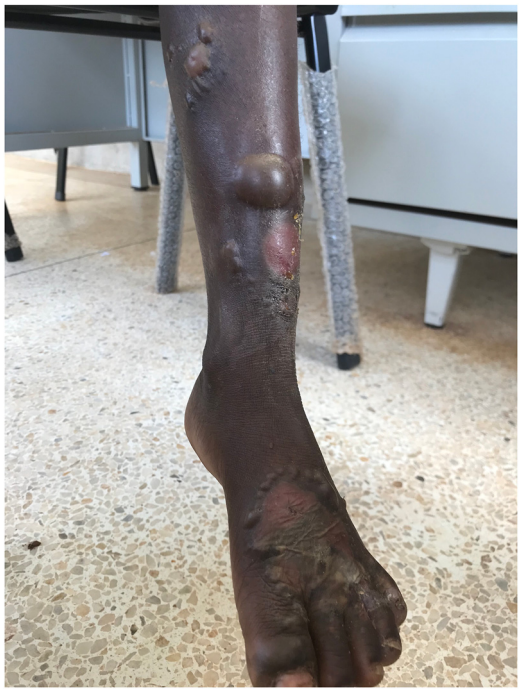

**Figure 1:** Urticarial plaques and numerous tense bullae in an annular arrangement on the lower left leg and foot.

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