# A bullous rash

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A 3-year-old boy presented with a 5-day history of bullous skin lesions localised mainly in the upper and lower limbs and in the genital region (figure 1). Lesions were not pruritic nor painful and showed a central crust. There was no family history of skin disorders or autoimmune diseases. The child never had fever and his physical examination was otherwise unremarkable.

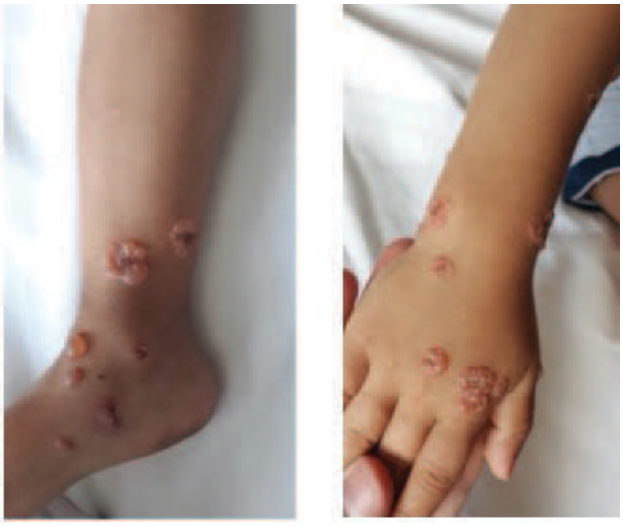

**Figure 1** Bullous skin lesions forming around a central crust, localised in the upper and lower limbs.

## QUESTIONS

1. What is the most likely diagnosis based on this clinical presentation?
   A. Bullous impetigo.
   B. Bullous pemphigoid.
   C. Linear IgA bullous dermatosis.
   D. Dermatitis herpetiformis.
2. What would be the next step in the investigation to confirm your diagnosis?
   A. Skin biopsy.
   B. Swab test for bacterial culture with an antibiogram.
   C. Anti-transglutaminase antibody detection.
3. What is the mainstay of management?
   A. Dapsone.
   B. Systemic steroids.
   C. Topical steroids.
   D. All of the above answers are correct, according to the severity of the disease.

***Answers can be found on page 195.***

## ANSWERS TO THE QUESTIONS ON PAGE 194

### Answer to question 1: C

Non-painful, non-pruritic bullous lesions with tense blisters arranged in a 'rosette' pattern and non-responsive to antibiotic therapy are highly suggestive of linear IgA bullous dermatosis.

Linear IgA bullous dermatosis is an idiopathic autoimmune blistering disease characterised by tense blisters surrounded by normal or erythematous skin with clear or haemorrhagic content. Typical pattern of lesions has been described as a string of pearls or clusters of jewels, as blisters typically form around a central crust. The most commonly involved areas are the face, trunks, perineal area and extremities. This condition generally relapses, persisting for a long time (months to several years) prior to resolution, and usually resolves in most children before puberty.[1]

Bullous impetigo is characterised by flaccid blisters that soon rupture with yellow fluid which becomes turbid.[2]

Clinically, bullous pemphigoid can appear identical to linear IgA bullous dermatosis and is characterised by pruritic tense blisters preceded by pruritic eczematous, papular or urticaria-like skin lesions.[3]

Grouped vesicles with centrifugal growth, which break easily and rarely persist, and cause severe itching and burning sensation in the lower limbs, elbows and sacral region, are typical of dermatitis herpetiforis.[4]

### Answer to question 2: A

Diagnosis is confirmed by skin biopsy. Typical histological features of linear IgA bullous dermatosis are subepidermal blisters with a neutrophilic infiltrate in the papillary dermis. Immunofluorescence of the perilesional skin is the gold standard for diagnosis and shows linear IgA staining along the basement membrane.[1]

### Answer to question 3: D

In very mild cases, topical corticosteroids may be the first choice. However, in more severe cases, dapsone is the first choice despite its potentially serious adverse effects, such as methemoglobinaemia, agranulocytosis, hypersensitivity syndrome and peripheral motor neuropathy. Oral steroids are used in very severe cases, or if adequate control is not achieved with dapsone or other sulfonamides and/or if toxic doses of these are required.

In case of relapse, oral steroids are used in mild to moderate flares, while intravenous steroids, immunoglobulin and/or immunosuppressants (mycophenolate mofetil, cyclophosphamide) are beneficial in patients with severe or refractory disease. Rituximab or anti-tumor necrosis factor inhibitors have been used in patients unresponsive to standard treatments.[5–7]

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