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# EPIDERMOLYSIS BULLOSA ACQUISITA IN A 4-YEAR-OLD HISPANIC BOY

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**ABSTRACT**
Epidermolysis bullosa acquisita (EBA) is an acquired blistering disease of the subepidermis. Antibodies against type VII collagen, which compose anchoring fibrils of the skin, underlie the disease process. EBA is extraordinarily rare in childhood, and we report the case of a four-year-old Hispanic boy with EBA who was successfully managed with prednisone and dapsone.

## INTRODUCTION

Epidermolysis bullosa acquisita (EBA) is an acquired subepidermal blistering disease caused by autoantibodies against type VII collagen, which composes the skin epithelium's anchoring fibrils.

Literature review demonstrates no prior reported cases of EBA in the Hispanic pediatric population, as the condition most commonly affects Caucasians. Our search of the literature revealed only 37 cases of childhood EBA since 1986, and only three prior cases of EBA have reportedly been diagnosed before the age of four years [1-28].

## Case Report

We report the case of a four-year-old Hispanic boy who presented with multiple disseminated, annular, pruritic, vesicles and tense, hemorrhagic, blisters. Oral and genital mucosal membranes were also involved.

Histopathology of punch biopsy showed subepidermal bulla with neutrophilic inflammatory infiltrate admixed with eosinophils. Direct immunofluorescence revealed linear deposition of IgG and C3 along the basement membrane and weakly positive IgM. Serum western immunoblot reacted with 290 and 145 kiloDalton proteins.

All findings are consistent with the diagnosis of EBA. Five weeks of treatment with combination prednisolone (1mg/kg/day) and dapsone (2mg/kg/day) resulted in complete resolution of the lesions. At 6- and 12-month follow-up, the patient had not experienced recurrence. Application of clobetasol ointment to any new bullae was recommended. Complications of EBA include malnutrition, stenosis of mucosal tracts (i.e. genital, esophagus, urethra), and symblepharon of conjunctivae with possible blindness. This patient did not demonstrate signs or symptoms of complications at follow-up.

## DISCUSSION

Differential diagnosis of widespread blistering disease includes infection (i.e. herpes simplex, impetigo), drug-induced bullous disease (i.e. furosemide, penicillin, captopril), dermatitis herpetiformis, erythema multiforme, Stevens-Johnson Syndrome, pemphigus vulgaris, bullous pemphigoid, cicatricial pemphigoid, linear (IgA) dermatosis, chronic bullous disease, and EBA. Histologic examination, direct immunofluorescence, and serum western immunoblot are performed to confirm a diagnosis. The findings in this case are classic for inflammatory type EBA.

There are two main clinical phenotypes of EBA: inflammatory and non-inflammatory. The inflammatory type, as seen in our patient, is more common in children. This type mimics other inflammatory bullous disorders and is characterized by pruritic tense bullae that may become hemorrhagic.

The lesions can crust or erode, leaving changes in pigment but do not cause scars. The second type of EBA is the noninflammatory mechanobullous type; this presents with skin fragility, blisters, and erosions at trauma sites. Atrophic scars and milia are left behind, particularly on dark-skinned individuals.

## CONCLUSION

This case is a valuable addition to the current body of medical knowledge as it broadens the patient demographics in which EBA should be considered. Furthermore, a treatment protocol of dapsone with prednisolone is supported by this report. Bullous disease is commonly seen in all age groups, and an understanding of the differential diagnosis can benefit patient management.

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**CONFLICT OF INTEREST: NIL**

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