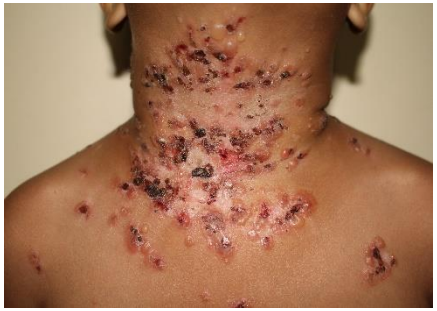
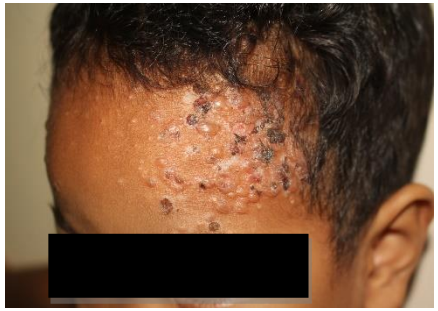
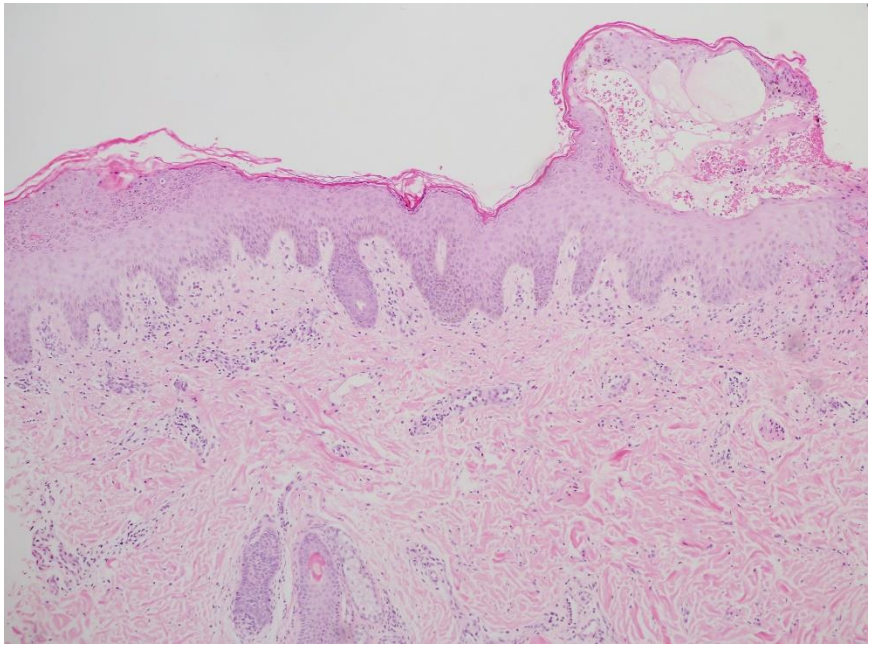
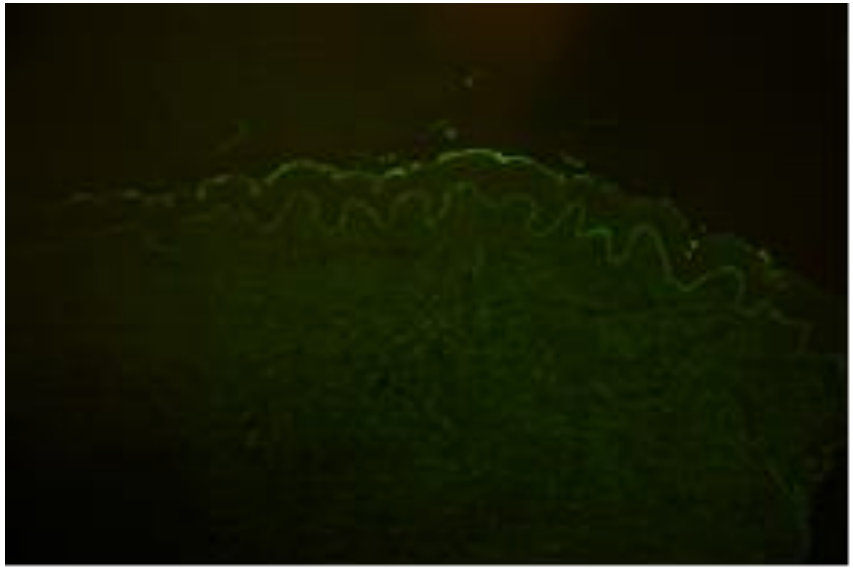
**Case 12**
A 4-year-old Thai boy from Bangkok
**Chief complaint**: Multiple pruritic tense bullae for 1 month

**Present illness**: The patient developed multiple pruritic tense blisters on left side of the forehead and spread to face, neck, trunk, upper and lower extremities for 1 month. Some lesions latter became crusted erosions. No history of upper respiratory tract infections or drug intake prior to the onset of blistering
**Past history:** Normal growth and development
Adequate immunization
**Family history:** No other family members show similar skin lesions
**Physical examination**
GA: active child, looked healthy, body weight 17 kg
HEENT: not pale, no jaundice, no oral ulcer
Lymph node: not palpable
Heart and lungs: WNL
Abdomen: no hepatosplenomegaly
**Skin examination**
Multiple discrete tense bullae and erosions with scales crusts on erythematous base and normal appearing skin in the arrangement of strings of pearls pattern on face, neck, trunk, upper and lower extremities

**Histopathology** (S15-012618, Chest wall)
-Subepidermal vesicle with reepithelization
-Inflammatory cell infiltrate of mainly lymphocytes in the superficial dermis
**Microscopic diagnosis**: Subepidermal vesicular dermatitis, resolving lesion





**Direct immunofluorescence:** Linear IgA deposition at dermo-epidermal junction, compatible with linear IgA dermatoses or CBDC

**Diagnosis:** Chronic Bullous Disease of Childhood**.** (CBDC)

**Investigation**: Hb 12 g/dL, Hct 37.5%
Reticulocyte count 3.3%
G6PD-normal

**Treatment:** Prednisolone 10 mg/day and tapered off
Dapsone 50 mg/day on 5 days per week
Folic acid 1 tab/day
2% Hydrocortisone cream applied bid

**Presenter:** Thirawut Sirikham, MD

**Consultant:** Kumutnart Chanprapaph, MD

**Discussion:**

Chronic Bullous Disease of Childhood (CBDC) or Linear IgA bullous dermatosis of childhood, is a rare chronic acquired subepidermal blistering disease of childhood.[1, 2] It was first described as a different entity from dermatitis herpetiformis or bullous pemphigoid by Chorzelski et al in 1979,[2] based on the immunopathologic finding of linear IgA deposits in the basement membrane zone on direct immunofluorescence. CBDC is characterized by homogeneous linear deposits of IgA at the basement membrane.[3] It is the most common acquired immunobullous diseases in children appears after age 6 months to before 5 years of age, even though several newborns with CBDC have been reported. It most frequently remits by six to eight years of age.[2, 4] The classic primary lesions of CBDC are clear and/or hemorrhagic vesicles or bullae on normal, erythematous, or urticated skin. Erythematous plaques, blanching macules and papules, or targetoid multiforme-like lesions may also be present with accompanied by variable degrees of pruritus.[5] Classic sites of CBDC are trunk and ilio-sacral region. Blister size varies but the blisters are characteristically arranged in a ring around a crust resembling a "string of pearls"[5] or a "jewel-like" distribution of blisters exists ("cluster of jewels"). Other sites of involvement include hands, feet, face, eyes[4] and large body folds particularly the perioral area. Oral lesions could be presented as vesicles, ulcerations, erythematous patches, or erosions.

Linear IgA disease antigen-1 (LAD-1), the 120-kDa cell-derived soluble ectodomain of BP antigen 180 (BP180)[6], is the major target antigen of IgA autoantibodies in patients with LAD. Antibody deposition leads to complement activation and neutrophil chemotaxis, which eventuates in loss of adhesion at the dermal-epidermal junction and in blister formation. Evidence suggests genetic susceptibility to this disease with an increased incidence of HLA-B8, HLA-DR3, and HLADQW2.[2, 5]

CBDC does not discriminate among races. Although typically idiopathic, it can follow an infection or represent a manifestation of drug hypersensitivity, most notably to vancomycin, 1 to 13 days after the first dose. Other drugs implicated in case reports include amiodarone, captopril, cefamandole, cotrimoxazole, cyclosporine, diclofenac, penicillin, and phenytoin.[5] Additionally, some reports associated with ulcerative colitis, lymphoma, and solid tumors.[5]

The differential diagnosis includes bullous impetigo, childhood bullous pemphigoid, dermatitis herpetiformis, epidermolysis bullosa acquisita, Erythema multforme or TEN-like lesion.[5, 7, 8]

No large, randomized, placebo-controlled, double-blind studies have been performed for the treatment of CBDC.[5] Most cases have been reported to respond to dapsone or sulfapyridine.[1] A normal G6PD level is required to avoid the side effect of severe hemolytic anemia. Widespread disease may require systemic corticosteroids along with dapsone to control the disease severity. Topical steroids can be used concurrently with the other therapeutic options or alone for mild cases.[5] Other drugs that have been used with success include erythromycin, colchicine, mycophenolate mofetil, and intravenous immmunoglobulins (IVIG).[5, 9] Most cases of CBDC spontaneously resolve within 5 years of onset. The severity of blistering does not affect the prognosis. In drug-induced cases, immediate cessation of the suspected drug will lead to spontaneous resolution.

## References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