

Autoimmune Blistering Diseases in Children

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Autoimmune blistering disorders comprise a series of conditions in which autoantibodies target components of the skin and mucous membranes, leading to blister and bullae formation. Most conditions in the spectrum of autoimmune blistering disorders are uncommonly seen in the pediatric population, even the most common ones, such as chronic bullous disease of childhood and dermatitis herpetiformis; however, they often come into the differential diagnosis of other more common pediatric entities. In addition, prompt recognition and treatment avoids unnecessary morbidity and improves ultimate outcome. Semin Cutan Med Surg 29:85-91 © 2010 Published by Elsevier Inc.

The incidence and prevalence of childhood autoimmune blistering disorders (ABD) is unknown because most of the existing literature consisting of case reports and case series. Blister formation, the clinical hallmark of these conditions, is caused by the interaction between structures essential for the integrity of the skin (desmosomes, hemidesmosomes, anchoring fibrils, etc.) and autoantibodies, leading to cleavage of the skin at various levels. Clinical diagnosis alone is difficult and requires histologic and immunopathological corroboration. Other conditions, such as bullous impetigo, herpetic infections, bullous erythema multiforme, Stevens–Johnson syndrome, inherited forms of porphyria, and congenital blistering disorders (epidermolysis bullosa), should be considered in the differential diagnosis. Genital involvement should be differentiated from sexual abuse, bullous lichen sclerosus, and bullous fixed drug eruption.

The most common ABDs in childhood are chronic bullous disease of childhood and dermatitis herpetiformis (DH). Less frequently seen entities are pemphigus, epidermolysis bullosa acquisita (EBA), herpes (pemphigoid) gestationis (HG), cicatricial pemphigoid (CP), bullous pemphigoid (BP), and bullous systemic lupus erythematosus.

Chronic Bullous Disease of Childhood

Chronic bullous disease of childhood (CBDC), also referred to as linear immunoglobulin A (IgA) disease of childhood, is

an acquired autoimmune subepidermal blistering disorder that is characterized by the deposition of a linear band of IgA along the dermal–epidermal junction. CBDC is thought to be the result of a humoral response to a normal constituent of the epidermal basement membrane zone (BMZ). Although the target antigen is known to be present in all human stratified squamous epithelia, it has not yet been defined with certainty. There may be a genetic susceptibility to CBDC with an increased incidence of HLA-B8, HLA-DR3, and HLA-DQW2.¹ However, the molecular basis of CBDC has not yet been clearly determined. This condition is seen in all ethnic groups and seems to be more common in developing countries. In the United Kingdom, its incidence has been estimated to be 1 in 500,000 children.²

The age of onset is typically before 5 years of age. Both skin and mucous membranes are affected, and typically the condition is preceded by a short nonspecific prodromal illness. The abrupt onset of tense vesicles on normal-looking skin or occasionally on urticated plaques is characteristic of the disease (Fig. 1). Skin findings may be accompanied by systemic symptoms, such as fever or anorexia. Blisters may be localized or widespread, with most common locations being face, extremities, genital area, and trunk.³ A pathognomonic feature is “string of pearls” rosettes or “cluster of jewels” appearance created by the development of new blisters at the margin of the old ones. Lesions may present with central clearing and have a polycyclic appearance. Mild pruritus has been described. Bullae resolve with transient pigmentary changes, and do not generally leave permanent scarring. Mucous membranes may be affected in up to 76% of patients.⁴

On histologic examination, subepidermal bullae are seen, and an inflammatory infiltrate with neutrophils or eosinophils can be found. Histology of fresh lesions on its own is not diagnostic and cannot distinguish CBDC from other autoimmune vesiculobullous disorders. The diagnosis relies on the

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Figure 1 Patient with CBDC.

immunohistochemical demonstration of a linear band of IgA along the dermal-epidermal junction that may be associated with weaker bands of IgG, IgM, or C3. The biopsy should be taken from peri-lesional skin. False-negative results may be seen early in the disease and may require a repeat biopsy for histologic confirmation.

Most cases of CBDC spontaneously resolve within 5 years of onset; few may progress through puberty and into adulthood. There is no correlation between the severity of blistering and disease chronicity.

Despite its good prognosis, most children are offered treatment to decrease the disease severity and shorten its duration. The most frequent medication is dapsone, starting at 0.5 to 1 mg/kg of body weight per day and increasing up to 2 mg/kg depending on the response. A normal glucose-6-phosphate-dehydrogenase level is a prerequisite of treatment initiation (to avoid hemolytic anemia) and baseline and monthly blood investigations are required for treatment monitoring. Most widespread cases may require additional treatment with oral prednisolone at 1 mg/kg/d for 1 to 3 weeks, tapered over 3 to 6 weeks, allowing disease control until dapsone's antiinflammatory effects occur. Topical steroids may be useful either as additional therapy for symptom control or single therapy for mild cases. Other systemic therapeutic options are sulfamethoxypyridazine and sulfapyridine.

Dermatitis Herpetiformis

Dermatitis herpetiformis (DH) is a chronic ABD. Although it thought to be one of the most common ABD in childhood,^{5,6} its exact prevalence remains unknown. DH has an equal gender presentation, is seen more commonly in Europe than in North America, and is unusual in patients of African and Asian background. Age at presentation varies, depending on the studies, ranging from 2 to 7 years,⁷ to a mean of 14 years.⁶

Cutaneous lesions of DH in childhood classically present

as intensely pruritic vesicles, erythematous papules, and urticarial plaques. Intact vesicles are rarely seen as the result of intense pruritus; most lesions are small excoriations/crusts. Scarring rarely occurs in the absence of suprainfection. The symmetry of the body involvement is very characteristic, typically affecting the extensor surfaces of the limbs, buttocks, shoulders, nape of neck and scalp (Fig. 2). Mucous membrane involvement is rarely encountered.

Gluten-sensitive enteropathy (subclinical or clinical) is present in almost all children with DH, but only a small fraction of them (10%) have a diagnosis of celiac disease at the time of the DH presentation.⁸ Most children have minimal or no symptoms of gluten-sensitive enteropathy. In up to 40% of children with DH there is a history of chronic or relapsing diarrhea before the diagnosis of DH.⁹

To avoid overdiagnosis, pathologic confirmation of the disease is mandatory. Classically, a subepidermal blister with neutrophilic (and occasionally eosinophilic) microabscesses within the dermal papillae is seen and fibrin deposition is common. The pathognomonic diagnostic feature is the presence of a granular deposition of IgA within the dermal papillae, occasionally accompanied by C3, on direct immunofluorescence examination of perilesional skin. The principal target antigen is epidermal transglutaminase.¹⁰ Similarly, a gastrointestinal work-up for celiac disease is necessary, particularly before a gluten free diet is recommended, as this will quickly interfere with the endoscopy findings.

With clinical suspicion of DH, it is also useful to measure the circulating IgA autoantibody to tissue transglutaminase (target antigen in celiac disease) because its titer reflects the degree of abnormality in the jejunal mucosa⁵ and can be used to monitor response to treatment or disease recurrence. Testing for IgA titers should be simultaneously performed because IgA deficiency, commonly seen in general population, leads to a



Figure 2 Patient with DH.

false-negative serology testing. The determination of anti-endomysial/antigliadin and other antibodies have been shown to be highly sensitive for celiac disease in children¹¹ but might be negative in up to 40% of cases of adult DH.¹² Thus, their value in diagnosing DH in children remains to be determined.

The combination of a gluten-free diet and dapsone are used for an effective treatment. A gluten-free diet alone may be sufficient because it reverses the intestinal abnormality in all cases and may lead to the disappearance of skin lesions in up to 82% of patients within 1 to 3 months.⁹ Dapsone is also an effective treatment for the eruption but does not reverse the gastrointestinal abnormalities. The standard initial dose of dapsone in childhood is 0.5 to 2 mg/kg/d.¹³ Once control of the disease has been obtained, the dosage of dapsone should be tapered. If the patient continues on a gluten-free diet, it may be possible to stop dapsone completely and quickly. Other treatment modalities, such as sulfapyridine, sulfamethoxypyridazine and systemic steroids can be considered. Super potent topical corticosteroids for short periods can be used as a dapsone-sparing agent.

The long-term prognosis for childhood DH remains uncertain. Long remissions are possible, but relapses are common typically as the result of poor adherence to the gluten-free diet, which is required lifelong.

Pemphigus

Pemphigus refers to a group of autoimmune vesiculobullous diseases that are characterized by the presence of antidesmosomal antibodies that lead to acantholysis and intraepidermal blister formation. These conditions can be classified into 2 main groups: the suprabasal type that includes pemphigus vulgaris (PV) and pemphigus vegetans and the superficial type, which includes pemphigus foliaceus (PF) and pemphigus erythematosus. The first group is generally associated with mucous membrane involvement while in the second 1 mucous membrane involvement is not a major feature.

Pemphigus is rarely seen in childhood, with a mean age of 12 years at presentation.¹⁴ Several environmental factors, such as medications¹⁵ and acantholytic substances¹⁶ superimposed on a genetic predisposition, may play a role in the onset of the disease. In children the most common form is PV, followed by PF. Other types, such as pemphigus vegetans⁷ and erythematosus¹⁷ are very rare.

Pemphigus Vulgaris

The blisters seen in PV are typically very flaccid and quickly rupture, leaving painful and persistent erosions and crusts in the skin. Nikolsky's sign is positive in this condition. Because blisters in PV are superficial, scarring is unusual.

Mucous membrane involvement, particularly of the oral mucosa, is frequent, severe, and may be the initial presenting feature. Other mucosal membranes are less frequently affected. PV should be in the differential diagnosis of mucosal blistering in both adults and children.

Antibodies against one of the desmosomal proteins, desmoglein 1 and 3, seem to play the central role in the patho-

genesis of both the superficial and suprabasal forms of pemphigus,¹⁸ but other autoantibodies such as antiacetylcholine receptor antibodies also seem to be of importance.¹⁹ The histologic characteristics of PV are acantholysis, with floating keratinocytes in the suprabasal cleavage plane, with the basal keratinocytes still attached to the epidermal basement membrane. A dermal infiltrate of lymphocytes, neutrophils and eosinophils is also typically seen.

With direct immunofluorescence of involved or perilesional skin, a deposition of IgG surrounding keratinocytes is seen and gives a "chicken wire" or "crazy paving" pattern. This is seen in both suprabasal and superficial forms of pemphigus. Repeat biopsies may sometimes be necessary for a diagnosis.¹⁸ Indirect immunofluorescence of the serum of patients with PV may show the circulating antibody; its titers correlate with clinical severity and can be used to monitor response to treatment.

Systemic corticosteroid therapy is the mainstay of treatment for PV in childhood. Prednisolone is used as a first line agent in doses of 1 to 2 mg/kg/d. Most patients require a steroid-sparing agent, such as dapsone,²⁰ azathioprine,⁷ methotrexate,⁷ cyclophosphamide,¹⁴ or hydroxychloroquine.²¹ Potent topical or intralesional corticosteroid can be used for isolated recalcitrant foci of persistent blistering. Children with PV typically have a better prognosis than adults.¹⁴

Pemphigus Foliaceus

PF is also referred to as superficial pemphigus, and it is rarely seen in children. The desmosomal protein, desmoglein-1, is the target antigen in this condition. Two different forms of PF have been recognized: an endemic form, Fogo selvagem, and a nonendemic. The etiology of the nonendemic form has not been identified, but in adults, certain drugs, like penicillamine, nifedipine, captopril, and quinolones, among others, have been associated with the disease.²² The nonendemic form presents at an average age of 7.7 years,²¹ with superficial erosions on the scalp and the face. The erosions are the result of the thin, fragile blister roof that sloughs off easily as the tension of the blister fluid increases. The superficial erosions resemble an exfoliative dermatitis and are commonly mistaken for impetigo.⁷ Occasionally, lesions may take arcuate, circinate or polycyclic patterns, which can make diagnosis even more difficult.²¹

In PF, the degree of acantholysis may be subtle. Acantholysis is seen in the upper epidermis and may result in a subcorneal separation. A mild dermal lymphocytic infiltrate may be found, frequently with eosinophils. Direct immunofluorescence does show the deposition of IgG around keratinocytes. PF in children has a good prognosis. Treatment is the same as for PV, but because of its milder nature, PF may just require topical corticosteroid therapy.

Paraneoplastic Pemphigus

Paraneoplastic pemphigus has been associated with B-cell lymphoproliferative neoplasms, and it is extremely rare in

children. It is characterized by severe oral mucositis and lichenoid lesions on the skin, and in pediatrics it has been recognized as a presenting sign of occult Castleman's disease.^{23,24} The most constant diagnostic marker has been identified to be the serum autoantibodies against plakine proteins.²³ Prognosis is extremely guarded with high mortality secondary to bronchiolitis obliterans and sepsis.²³

Epidermolysis Bullosa Acquisita

Epidermolysis bullosa acquisita (EBA) is a chronic subepidermal immunobullous disorder that occurs infrequently rare in childhood. No gender or racial predilection has been reported for childhood EBA. An increased incidence of HLA-DR2 has been identified in some patients with EBA, suggesting a genetic susceptibility.²⁵

Clinical features of EBA are variable,²⁶ and clinical distinction alone may not be possible. Age of presentation can be at anytime during childhood, starting with infancy.²⁷ Three clinical phenotypes have been recognized. The first is a classic noninflammatory mechano-bullous type that resembles the congenital form of dystrophic EB: skin fragility, with blisters and erosions seen at sites of trauma, particularly over acral bony prominences; milia formation; atrophic scars with pigmentary changes; and nail dystrophy. The second type is an inflammatory type that imitates CBDC, presenting with pruritic, tense bullae, on normal, erythematous or urticarial skin. Occasional hemorrhagic lesions may occur forming crusts and resolving with pigmentary changes. Severe mucous membrane involvement can be seen.^{28,29} The third clinical variant of EBA resembles CP, presenting with predominantly mucous membrane involvement and a pronounced tendency to scar. The severe involvement of mucous membranes in children may lead to a variety of complications: malnutrition, symblepharon of the conjunctivae which may progress to blindness,³⁰ and stenosis of the esophagus, urethra or genital tract. Although all types are seen, the inflammatory form is more common in the pediatric population, as is the mucosal involvement. Children also tend to respond faster to systemic therapy.³¹

The target of the autoantibodies in EBA is type VII collagen that is part of the anchoring fibrils of the epidermal basement membrane. Histologic examination of fresh lesions reveals subepidermal bullae and a predominantly neutrophilic inflammatory infiltrate admixed with eosinophils. With direct immunofluorescence examination of perilesional skin, linear depositions of IgG and C3 along the BMZ are detected, but sometimes weak staining for IgA and IgM are also seen. In few cases, IgA may be the predominant immunoreactant.³⁰ In EBA, indirect immunofluorescence is usually positive, the circulating antibody labels the dermal side of the bulla in salt-split normal human skin,³² and direct immunoelectron microscopy reveals IgG deposits under the lamina densa.

Early-onset EBA needs to be differentiated from inherited dystrophic epidermolysis bullosa. Birth presentation and results of direct immunofluorescence (positive for EBA, negative for dystrophic epidermolysis bullosa) help to differenti-

ate between these 2 conditions: however, genetic testing might be required to make a more definitive demarcation.³³

Combination therapy with prednisolone 1 mg kg⁻¹ d⁻¹ and dapsone 2 mg kg⁻¹ d⁻¹ is effective in controlling the disease manifestation.³⁴ Patients usually respond quickly to treatment, within weeks, and then prednisolone can be tapered off. Alternative therapies that have been reported include sulfapyridine, a combination of nicotinamide and erythromycin, and for localized cases, superpotent topical steroids. Long-term prognosis for EBA in children is much better than its adult counterpart. Children with EBA generally undergo remission within 1 to 4 years, although some might take longer to resolve.

Herpes (Pemphigoid) Gestationis and Neonatal Pemphigus

Herpes (pemphigoid) gestationis (HG) is a rare autoimmune disease that presents during pregnancy or in the postpartum period. It is seen in about 1 of 50,000 pregnancies. Approximately 2% to 10% of the neonates born to women with HG will be affected^{35,36} because of the passive transplacental transfer of IgG antibodies from the mother before delivery.³⁷ Antibodies usually target BP180 and occasionally BP230.³⁸ As the antibody titers decrease, the clinical manifestations in the infant disappear typically within 2 weeks to 3 months.³⁹ Patients characteristically present with intensely pruritic urticarial plaques with tense bullae.

Histologic examination of the lesions shows subepidermal edema and blistering that is associated with a moderate dermal eosinophilic infiltrate. Direct immunofluorescence of perilesional skin generally shows a linear deposition of C3 in the epidermal basement membrane, with a linear deposition of IgG in up to 25% to 50% of cases.^{40,41} Indirect immunofluorescence examination may show circulating IgG antibodies confined to the epidermal side of the salt-split skin.

Although specific treatment is not needed, moderately potent topical corticosteroids may help patients with a significant inflammatory component. The prognosis for infants with cutaneous involvement is generally good. All infants born to women with HG are at risk of being small for gestational age and have a tendency to prematurity.

Cicatricial Pemphigoid

Cicatricial pemphigoid (CP) is also referred to as benign mucous membrane pemphigoid and is an extremely rare condition in childhood. There are clinical, histologic, and immunopathological overlaps between CP and other autoimmune subepidermal blistering disorders. It has been proposed that CP in children is an unusual and more severe form of CBDC.⁴² Because of its rarity in childhood, diagnosis may be delayed.⁴³

It commonly presents as a generalized eruption that involves the face, trunk, and limbs and is characterized by urticarial or annular, polycyclic, or target-like lesions. The

Table 1 Comparison of the Most Common Pediatric ABDs

	CBDC	DH	PV	PF	BP
Cutaneous lesions	<ul style="list-style-type: none"> ● Tense vesicles on normal/urticarial patches ● “String of pearls” 	<ul style="list-style-type: none"> ● polymorphic (small vesicles, erythematous papules and urticarial plaques)—erosions ● Crusts 	<ul style="list-style-type: none"> ● Flaccid blisters ● Painful/persistent erosions and crusts 	<ul style="list-style-type: none"> ● exfoliative dermatitis ● arcuate, circinate or polycyclic lesions 	<ul style="list-style-type: none"> ● Large, tense blisters that may be hemorrhagic
Most common distribution	<ul style="list-style-type: none"> ● Face ● Extremities ● Genital area ● Trunk 	<ul style="list-style-type: none"> ● Extensor surfaces of the limbs ● Buttocks ● Shoulders ● Nape of neck ● Scalp 	<ul style="list-style-type: none"> ● Scalp ● Face ● Upper torso ● Oral mucosa Only 	<ul style="list-style-type: none"> ● Head and neck 	<ul style="list-style-type: none"> ● Acral (palms and soles) ● Flexural areas (inner thighs, forearms, axillae, lower abdomen, and groin)
Pruritus	±	++++	–	–	± to +++
Mucosal involvement	Common	None	Frequent and severe (oral mucosa mostly)	None	Frequent
Histology	<ul style="list-style-type: none"> ● Subepidermal blisters ● Inflammatory infiltrate with Eos and PMN 	<ul style="list-style-type: none"> ● Subepidermal blisters with neutrophilic microabscesses within dermal papillae ● Fibrin deposition 	<ul style="list-style-type: none"> ● Acantholysis ● Dermal infiltrate (lymph, PMN, Eos) 	<ul style="list-style-type: none"> ● Subtle acantholysis ● Subcorneal separation ● Dermal lymph infiltrate ± Eos 	<ul style="list-style-type: none"> ● Subepidermal blisters with eosinophilia
Direct Immunofluorescence	<ul style="list-style-type: none"> ● Linear IgA along dermal epidermal junction 	<ul style="list-style-type: none"> ● Granular deposits of IgA within dermal papillae 	<ul style="list-style-type: none"> ● IgG deposition around keratinocytes “chicken wire” or “crazy paving” pattern 	<ul style="list-style-type: none"> ● Deposition of IgG around keratinocytes 	<ul style="list-style-type: none"> ● Linear deposits of IgG or C3 at basement membrane zone
First-line treatment	<ul style="list-style-type: none"> ● Dapsone 	<ul style="list-style-type: none"> ● Dapsone ● Gluten free-diet 	<ul style="list-style-type: none"> ● Systemic corticosteroids 	<ul style="list-style-type: none"> ● Systemic or topical corticosteroids 	<ul style="list-style-type: none"> ● Systemic corticosteroids

ABD, autoimmune blistering disorders; BP, bullous pemphigoid; CBDC, chronic bullous disease of childhood; DH, dermatitis herpetiformis; PF, pemphigus foliaceus; PV, pemphigus vulgaris; Eos, eosinophils.

predominant clinical feature is the severe involvement of mucous membranes, leading to various degrees of scarring.

Multiple target antigens for CP have been described, such as BP180,³⁵ laminin 5, type VII collagen, beta 4 subunit of the $\alpha 6\beta 4$ integrin,⁴⁴ accounting for its clinical heterogeneity. Direct immunofluorescence may reveal IgA and/or IgG deposited in a linear pattern at the dermal–epidermal junction. Indirect immunofluorescence detects circulating antibodies against epithelial basement membrane constituents in about 50% of cases.⁴⁵ Immunoelectron microscopy shows immunoreactants on the lower part of the lamina *Lucida* or on the lamina densa, correlating with the deposition of IgG to the target antigens.

Treatment consists of either systemic corticosteroids, dapsone or sulfapyridine.⁴² Topical treatment with steroids may provide relief of symptoms. Complete remission is possible with very few cases extending into adulthood.

Bullous Pemphigoid

Bullous pemphigoid (BP) rarely presents in children. Diagnostic criteria have been proposed to facilitate early recognition of childhood BP: (1) age 18 years and younger; (2) clinical appearance of tense blisters; (3) subepidermal blisters with eosinophilia; and (4) linear deposits of IgG or C3 at the epidermal BMZ on direct immunofluorescence of perilesional skin, or circulating IgG anti-BMZ autoantibodies on indirect immunofluorescence using patient serum.⁴⁶

Clinically, BP presents with tense bullae, sometimes hemorrhagic, arising from normal or inflamed skin. Urticarial plaques are common, seen in annular or polycyclic patterns. Folds (particularly groin and axilla), abdomen and inner thigh involvement are very common. Palmar and plantar involvement is characteristic of infantile-onset BP and can be seen in up to 79% of affected infants versus 17% of older children.^{47,48} Facial involvement is more common in childhood BP and can easily be misdiagnosed as impetigo. Childhood characteristics of the disease are frequent mucous membrane involvement, acral distribution, and no clear link to malignancies.

The targets of the autoantibodies in BP are BP230 and/or BP180, 2 hemidesmosome-associated proteins. Histology of BP shows subepidermal blistering with an intact overlying epidermis and no necrosis. Inflammatory infiltrate is mostly composed of eosinophils, with some neutrophils and lymphocytes. Direct immunofluorescence may reveal a linear deposition of IgG and C3, and less frequently, IgM and IgA may be found.⁴⁹ Indirect immunofluorescence may demonstrate a circulating antibody which is directed against the epidermal side of salt-split normal human skin, but occasionally dermal binding may be observed, despite Western immunoblotting revealing an epidermal location for the target antigens.⁵⁰ Immunoelectron microscopy shows deposition of autoantibody in the upper lamina *Lucida*.

The treatment of choice for BP is systemic corticosteroids, starting with a dose of prednisolone of 1 to 2 mg/kg/d.⁵¹ Other treatment modalities have been used, like dapsone or sulfapyridine or a combination of erythromycin and nicotin-

amide.⁵⁰ Prognosis for children with BP is good, with most cases lasting for 1 year or less.

Bullous Systemic Lupus Erythematosus

Bullous systemic lupus erythematosus, defined as an ABD presenting in an individual that meets criteria for the diagnosis of systemic lupus erythematosus, is rarely seen in children. Sun-exposed areas are more commonly involved with a chronic, widespread, itchy, non-scarring bullous eruption, with tense blisters arising on normal or urticated skin. Rarely, mucous membrane involvement may be seen.

Although type VII collagen has been described as the target antigen in this condition, autoantibodies are produced to a variety of other molecules that may be involved in dermal–epidermal adhesion, such as BP antigen 1 (BP230), laminin 5, and laminin 6.⁵²

Histologically, bullous systemic lupus erythematosus presents with subepidermal blistering with a predominantly neutrophilic infiltrate within the upper dermis. Direct immunofluorescence of lesional skin shows IgG and complement deposition along the epidermal basement membrane, with sporadic weaker staining with IgA and/or IgM. Indirect immunofluorescence examination on salt-split skin usually shows antibody localization to the dermal side. Ultrastructural examination shows immune deposits on or beneath the lamina densa of the dermal–epidermal junction.

The treatment of choice for bullous systemic lupus erythematosus is dapsone, and the blistering tendency usually responds quickly. Although the prognosis of the blistering eruption is good, the ultimate outcome depends on the degree of systemic involvement.

Conclusions

Distinguishing between the different ABD in children is a difficult and challenging task. Table 1 presents characteristics of the most common ABD disorders in pediatrics. Knowledge of the various clinical characteristics of these disorders will aid in their diagnosis; however, histology and immunofluorescence studies are required to differentiate them.

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