

European S2k guidelines on management of autoimmune blistering diseases in children and adolescents

A. Nanda¹  | B. Tedbirt^{2,3} | C. Bodemer³ | M. C. Bolling⁴ | M. El Hachem⁵ | M. Nikolić⁶ | E. Benton⁷ | M. Caproni⁸  | D. De⁹ | D. Didona¹⁰ | K. Drenovska¹¹  | M. Goebeler¹² | M. Hertl¹³ | B. Horváth⁴ | M. Kasperkiewicz¹⁴ | D. Kiritsi^{15,16} | M. Larralde¹⁷ | A. V. Marzano^{18,19}  | J. Meijer⁴  | D. F. Murrell^{20,21}  | J. Powell²² | E. Sprecher^{23,24}  | N. S. Chandran²⁵ | M. M. Tomayko²⁶ | S. Uzun²⁷ | N. van Beek²⁸  | H. Ott²⁹ | A. Patsatsi³⁰  | A. Torrelo³¹ | A. Welfringer-Morin³²  | E. Schmidt^{28,33}  | P. Joly^{2,3} 

Correspondence

E. Schmidt, Department of Dermatology,
University of Lübeck Ratzeburger Allee 160
Lübeck, DE 23562, Germany.
Email: enno.schmidt@uksh.de

Abstract

Introduction: Autoimmune blistering diseases (AIBDs) in children and adolescents present a major challenge in diagnosis and management because of their rarity and the presence of other differential diagnoses with blistering. They are potentially serious, and timely diagnosis and optimal management can help prevent morbidity related to both the diseases and their treatments.

Methods: The Autoimmune Blistering Diseases Task Force of the European Academy of Dermatology and Venereology (EADV) has initiated evidence-based, expert-informed clinical guidelines for the diagnosis, treatment and monitoring of paediatric AIBDs. A writing group conducted a systematic review of the available literature on AIBDs in children and adolescents, and since there is limited strong evidence in the literature, recommendations were formulated by consensus over three rounds of structured feedback and revision by the Core Group and Expert Group of the task force (32 voting experts; >75% agreement threshold).

Results: Although all AIBDs—including intra-epidermal and subepidermal blistering disorders—are reported in children, linear IgA dermatosis (LAD) is the most common (42–83%). The diagnostic approach in the paediatric age group does not differ from that in adults. Treatment recommendations are tailored to paediatric patients, taking into account developmental pharmacokinetics, potential adverse effects and the impact of disease and therapy on quality of life.

Conclusions: These guidelines provide the first comprehensive, paediatric-focused framework for the diagnosis and management of AIBDs. Implementing the recommendations should harmonize care, reduce treatment-related toxicity and improve quality of life for affected children while steering future research towards critical knowledge gaps.

A. Nanda, B. Tedbirt, E. Schmidt and P. Joly have equally contributed to this work.

Members of the core group: C. Bodemer, M. C. Bolling, M. El Hachem, M. Nikolić, H. Ott, A. Patsatsi, A. Torrelo, and A. Welfringer-Morin.

For affiliations refer to page 1155.

Linked Article: P. C. Luna and M. E. Abad. *J Eur Acad Dermatol Venereol* 2026;40:1112–1113. <https://doi.org/10.1111/jdv.70492>.

This is an open access article under the terms of the [Creative Commons Attribution-NonCommercial-NoDerivs](https://creativecommons.org/licenses/by-nc-nd/4.0/) License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2026 The Author(s). *Journal of the European Academy of Dermatology and Venereology* published by John Wiley & Sons Ltd on behalf of European Academy of Dermatology and Venereology.

KEY WORDS

dermatitis herpetiformis, epidermolysis bullosa acquisita, linear IgA disease, mucous membrane pemphigoid, paediatrics, pemphigoid, pemphigus

INTRODUCTION

Autoimmune blistering disorders (AIBDs) are a rare heterogeneous group of skin diseases that present with blistering and erosions due to disruption of the skin barrier caused by the production of autoantibodies against structural proteins in the epidermis or at the epidermal–dermal junction. Based on the level of cleavage, they are broadly classified into intraepidermal and subepidermal AIBDs. In intraepidermal AIBDs, the autoantibodies are directed against the desmosomal proteins and clinically the patients manifest with pemphigus and its variants.^{1–3} In subepidermal AIBDs, the antibodies are directed against hemidesmosomal structural proteins resulting in bullous pemphigoid (BP), mucous membrane pemphigoid (MMP), linear IgA dermatosis (LAD), dermatitis herpetiformis (DH) and epidermolysis bullosa acquisita (EBA) depending upon the antigens involved.^{1–3} Although well characterized in adults,^{1–3} the data on AIBDs in children are scarce.^{4–13} They are much rarer in children as compared to adults and there are no population-based prospective studies conducted to determine the epidemiology of AIBDs in this age group. Evidence-based treatments are limited in childhood. To date, there are no guidelines available to diagnose and treat AIBDs in children. The AIBDs task force of the European Academy of Dermatology and Venereology (EADV) has taken an initiative to prepare the guidelines for the management of AIBDs in children based on the available literature and experience of physicians involved in the care of children with AIBDs.

METHODS

Development of the guidelines

The aim of these guidelines is to provide healthcare professionals with the current optimal management and care pathway for a child with an AIBD. These guidelines can be used as a reference by the treating physician in consultation with the specialist paediatric dermatologist.

The purpose of the guidelines is to standardize the management and follow-up of the disease in order to improve the quality of life of patients and their families.

These guidelines were initiated by the EADV Task Force of AIBDs and the writing group was formed at the Task Force meeting during the 32nd EADV Annual Congress 2023 consisting of A.N., B.T., E.S. and P.J. The task force agreed to develop consensus-based (S2k) guidelines, according to the recommendations of the Association of the

Why was the study undertaken?

- Autoimmune bullous diseases (AIBDs) in children and adolescents are rare and difficult to manage, with no existing paediatric-specific guidelines. Their rarity, clinical heterogeneity and lack of evidence often result in inconsistent diagnosis, treatment and outcomes.

What does this study add?

- These guidelines provide standardized diagnostic criteria and treatment protocols for paediatric AIBDs. They aim to minimize variability in care, ensure rational use of systemic therapies including immunosuppressants and biologics and improve outcomes and quality of life.

What are the implications of this study for disease understanding and/or clinical care?

- The guidelines promote early and accurate diagnosis, enhance patient safety and reduce disparities in care. They also establish benchmarks for assessing disease activity, remission and relapse, thereby supporting more consistent long-term management.

Scientific Medical Societies in Germany (AWMF) as shown in the AWMF Guidance Manual and Rules for Guideline Development, Version 2.1 (https://www.awmf.org/fileadmin/user_upload/dateien/downloads_regelwerk/awmf-regelwerk-en-2023-v2.1.pdf). The writing group prepared the first draft of the guideline that was discussed on a virtual meeting with the guideline core group consisting of paediatric dermatologists experienced in the management of minors with AIBDs including A.T., M.N., C.B., M.E.H., K.P., A.W., H.O. and M.B. Recommendations were subsequently agreed upon by the members of the core group. Recommendations with strong objections were rephrased and revoted until all recommendations reached an agreement of >75% of the core group. Subsequently, recommendations were voted upon by the voting group that had been selected by the AIBD Task Force. The grade (strength) and level of agreement (including the votes of the writing group, the core group, and the voting group) for recommendations are shown in [Tables 1 and 2](#).

TABLE 1 Grades (levels) of recommendation in these guidelines.

Level of recommendation	Syntax
Very strong recommendation (it is practically obligatory)	It is necessary (+++)
Strong recommendation (some exceptions are acceptable)	It is recommended (++)
Less strong recommendation (acceptable based on few case reports/ adult data)	May be recommended (+)
Weak recommendation (it is acceptable but it is not recommended as a rule; based on anecdotal reports both in children and adults)	May be considered (±)
Rejection (not recommended)	It is not recommended or it is contraindicated (-)

TABLE 2 Levels of consensus in these guidelines.

Level of consensus	Symbol
Strong consensus (agreement of >95% of participants)	
Consensus (agreement of >75-95% of participants)	
Agreement of the majority (agreement of 50-75% of participants)	

Conflicts of interests were collected from all members of the writing, core and voting group and classified as low, moderate and severe by the writing group according to the AWMF Guidance Manual and Rules for Guideline Development, Version 2.1. Nine members (E.B., M.H., M.G., N.C.S., K.P., P.J., A.N., E.S. and N.C.) had moderate conflicts of interests and absented from voting on the respective recommendations.

AIBDs IN CHILDREN

How common are AIBDs in children?

The epidemiological data on AIBDs in children is scarce and is based on small series of AIBDs published in the literature.⁴⁻¹² The true prevalence rate of AIBDs in children is not known. In an age-adjusted 1-year (2015) population-based report from Germany from the database created by German health insurance using ICD-10 code, the prevalence rate of the AIBDs among children was observed to be 101.1 cases per million children and was highest for pemphigus variants (30.5/million) followed by LAD (24.5/million) and BP (4.9/million).¹⁴ Earlier in Kuwait, the age-adjusted incidence rate of LAD among children was observed to be 2.3 cases per million per year.¹⁵ Based on the available data, LAD is the most prevalent AIBD (42–83%) reported in most studies except the USA⁵ where DH was the most common (39%) AIBD, and Germany and India where pemphigus was most

prevalent (51–60%).^{12,14} LAD was observed to be the second commonest AIBD in the USA, Germany and India. The differences in their prevalence in various studies are most probably related to factors such as referral bias or ethnic/genetic influences.

Special concerns when managing children with AIBDs

Diagnosing children with AIBDs can be challenging with a broad range of differential diagnoses including various infections (bacterial, viral and fungal), inherited blistering disorders and, less frequently, other rare disorders such as bullous mastocytosis, bullous drug reactions or bullous dermatitis artefacta. They may first need to be ruled out by relevant testing before investigations for AIBDs are ordered.

When managing children with AIBDs, they should not be considered just little adults. Different disease behaviour due to different 'host' responses, different disease variants, different pharmacodynamics and different adverse drug reactions can explain why children can behave differently. In this young population, special consideration must be given to quality of life (QoL) and school performance impairment, the long-term sequelae of the disease- and treatment-related toxicity. There is a current lack of paediatric-specific controlled trials and treatment strategies are generally guided by adult data. Keeping in mind the enormous psychological and social impact of the disease and its treatments upon the child or young person and their family necessitates a comprehensive, holistic, specialized multidisciplinary approach to manage AIBDs.

When to suspect a diagnosis of AIBD in a child?

- AIBD should be considered in a child of any age presenting with progressive, widespread or recurrent bullous eruptions once the possibility of an infectious aetiology or inherited diseases has been ruled out.
- A detailed history of age at onset, precipitating or aggravating factors including infections, drugs, vaccination, trauma, maternal factors, family history and associated mucocutaneous and systemic manifestations helps to decide the next line of action for further investigations.

Differential diagnoses

Major differential diagnoses are shown in Table 3. In most cases of AIBDs appearing in the neonatal period, the disease is acquired in utero due to the placental transfer of maternal antibodies from an affected, usually symptomatic mother. In patients revealing blistering late in childhood, the differential diagnoses include acquired infections, adverse drug reactions, traumatic blisters, bullous mastocytosis and vasculitis.

TABLE 3 Differential diagnosis of bullous eruptions in a child.

Inherited/congenital disorders	Acquired disorders
Inherited disorders - Epidermolysis bullosa - Keratinopathic ichthyosis - Incontinentia pigmenti - Acrodermatitis enteropathica - Congenital porphyria - Peeling skin syndromes - AEC syndrome (Hay-Wells syndrome) - Congenital absence of skin (aplasia cutis congenital) - Ectodermal dysplasia with skin fragility Congenital infections - Herpes simplex - Varicella zoster - Cytomegalo-virus - Candida - Congenital syphilis (pemphigus syphiliticus) - Others	Infections - Bullous impetigo - Staphylococcal scalded skin syndrome - Herpes simplex - Varicella zoster - Eczema herpeticum - Hand-foot-and-mouth disease - Mycoplasma-induced rash and mucositis - Scabies - Other bacterial, viral, fungal Autoimmune blistering disorders - Linear IgA bullous disease - Pemphigus variants - Bullous pemphigoid - Mucous membrane pemphigoid - Dermatitis herpetiformis - Epidermolysis bullosa acquisita - Bullous lupus erythematosus Miscellaneous - Traumatic blisters - Bullous mastocytosis - Stevens-Johnsons syndrome/toxic epidermal necrolysis - Acute generalized exanthematous pustulosis - Vasculitis - Bullous dermatitis artefacta - Others

Stomatologist, ophthalmologist, otorhinolaryngologist, gastroenterologist, gynecologist, urologist, proctologist, etc.
Paediatric pain care specialist
Physiotherapist, dietician, orthoptist, psychologist, social worker, school doctor to set up an individualized reception protocol if necessary

What to do in the case of a child with an AIBD?

The following steps are necessary in a child with suspected AIBD:

- It is necessary to (consensus: 94%): - Take a skin or mucosal biopsy for histopathology and direct immunofluorescence (DIF) examination - Take blood for serological tests (detailed 2.5.2)
- It is recommended to (strong consensus: 100%): - Take blood for routine baseline investigations including complete blood counts, serum biochemistry, liver and renal functions, for monitoring the patients during the treatment. - Take skin/mucosal swabs for bacteria culture and sensitivity if superadded infection is suspected. - Measure blood pressure, weight, height. - Other examination can be performed according the treatment chosen (for instance G6PD in the case of the case of treatment with dapsone)

The following precautions are recommended (**strong consensus: 97%**):

Avoid to start any systemic treatment (especially systemic corticosteroids) until the diagnosis has been confirmed unless in severe disease
If DH is suspected, do not start the gluten-free diet without confirming the diagnosis. The risk is the false negative result of immunological examinations and small bowel histology

Interrogation and clinical examination in the setting of a child with AIBD

Questioning of the child and parents

The following parameters are recommended to be included in the medical history (**strong consensus: 100%**):

The date of onset of symptoms
Presence and intensity of pruritus
Mucosal involvement (oral erosive lesions often mistaken for oral ulcers, conjunctivitis, nasal obstruction, epistaxis, dyspnea, dysphonia, dysphagia, dysuria, pain during defecation, and anal erosive lesions) should be recorded
The evolution of the lesions and their impact on the general condition (weight loss, asthenia, etc.)
Precipitating or aggravating factors including infections, ectoparasitic infestations, insect bites, drug intake, and recent vaccination*
A history of hematological disorders (G6PD deficiency), autoimmune disorders, gastrointestinal disorders (gastritis, Crohn's disease, ulcerative colitis, celiac disease, systemic lupus erythematosus), endocrine disorders (diabetes, thyroiditis), inborn errors of immunity, recent infections (especially in cases of severe or extensive disease potentially requiring the use of an immunosuppressant), and any other contraindications to the treatments
History of blistering disease in the mother in case of suspected AIBD in the neonate

*When positive history of vaccination or drug intake prior to onset of rash is present, their chronology in relation to the onset of the rash (in favour of a vaccination/drug-induced AIBD) must be determined.

DIAGNOSIS OF AIBDs

Primary objectives

- Make a correct diagnosis
- Search for a triggering factor
- Introduce appropriate treatment
- Organize follow-up

Professionals involved

It is necessary to involve the following professions (**consensus: 87%**):

Dermatologist experienced in care of AIBDs in children
Pediatrician

It is recommended to involve the following professions when required depending upon the severity of the diseases, involvement of the mucosae, impact of disease on child's daily performance, and psychological status (**strong consensus: 97%**):

Clinical examination

The following examinations are recommended (**consensus: 94%**):

- Assess the extent of cutaneous and mucosal lesions, the number of mucosal areas affected, and functional discomfort (pain, dysphagia, weight loss, growth chart break, ocular, urinary symptoms)
- Evaluate the general state and possible comorbidities (overweight, and hypertension in particular):
 - Psychomotor development
 - Nutritional status (weight, height, head circumference [infants] growth curve)

The following examinations may be recommended (**consensus: 94%**):

- Measure disease extent using scoring systems such as Pemphigus Disease Area Index (PDAI), Bullous Pemphigoid Disease Area Index (BPDAI), MMPDAI, and Autoimmune Bullous Skin Intensity and Severity Score (ABSIS) or a global evaluation (IGA) although none of these scores has been validated in minors.

Evaluation of the impact of patient's disease on the patient's day to day life

It is recommended to evaluate the following daily activities (**consensus: 94%**):

- Intensity of itch, sleep impairment, and anxiety
- Performance in school or school drop-out
- Social interactions with siblings/peers
- It may be considered to assess the quality of life by an established tool e.g. Children's Dermatology Life Quality Index (CDLQI)^{16,17} (**consensus: 87%**)
<https://www.cardiff.ac.uk/medicine/resources/quality-of-life-questnaires/childrens-dermatology-life-quality-index>

Diagnostic management

Diagnosis of AIBDs in children follows the same lines as in adults (Table 4 and Figures 1 and 2). Below, an overview is given. More details can be found in the EADV guidelines for pemphigus vulgaris (PV), pemphigus foliaceus (PF), paraneoplastic pemphigus (PNP), BP, MMP, LAD, EBA and DH.^{18–26}

Skin and/or mucosal biopsies

- It is necessary to do a biopsy on perilesional skin/mucosa for DIF microscopy (**consensus: 94%**). The biopsy needs to be transferred in 0.9% saline solution or frozen rapidly in a cryotube for transport in a liquid nitrogen tank or to be placed in Michel's medium for up to 72 hours before being processed.
- The use of EMLA may decrease the pain related to biopsy, however, there is no agreement between experts on the potential risk of artefact related to the use EMLA (**consensus: 91%**)
- It is recommended to take a biopsy of a recent lesion (entire vesicle or the edge of a blister and adjacent normal tissue) for histological examination to study the level of cleavage (subepidermal or intraepidermal AIBD), the content of the bulla and the dermal inflammatory infiltrate (neutrophils in favor of linear IgA dermatosis or dermatitis herpetiformis, eosinophil infiltrate in bullous pemphigoid) (**consensus: 94%**)
- It may be recommended to apply serration pattern analysis of the dermal-epidermal junction in DIF samples in all patients with linear depositions (**consensus: 94%**). A u-serrated pattern is seen with reactivity against type VII collagen, i.e. in epidermolysis bullosa acquisita and bullous lupus erythematosus. In all other pemphigoid diseases, a n-serration pattern is present^{24,27–29}
- In cases where serration pattern diagnosis is inconclusive, it may be recommended to do 1M NaCl-splitting of the biopsy for DIF microscopy. This method allows the differentiation of pemphigoid diseases with autoantibodies binding to the epidermal side of the split (BP180, BP230, α6β4 integrin) from those with dermal binding (type VII collagen, laminin 332, p200 antigen) (**consensus: 90%**)
- Direct fluorescent overlay antigen mapping (FOAM) or direct immunoelectron microscopy (IEM)^{30,31} may be recommended in individual cases when available (**strong consensus: 100%**)

Note: Not all immunology and pathology laboratories are equipped or do have experienced staff to perform IEM, FOAM or serration pattern analysis. For such cases, if required, specialized centers may need to be accessed.

Detection of serum autoantibodies

- It is necessary to collect serum for all patients with suspected AIBD (**strong consensus: 100%**)
- It is recommended to perform indirect immunofluorescence (IIF) on normal human skin in pemphigoid diseases (ideally on normal human 1M NaCl-split skin) and/or on monkey esophagus in pemphigus as screening assay for autoantibodies (**strong consensus: 100%**)
- It is recommended to perform subsequent analyses using antigen-specific enzyme linked immunosorbent assay (ELISA) or IIF test (Biochip™) that are commercially available or in-house tests (ELISA, IIF, immunoblot, using various organ substrates, or recombinant or cell-derived forms of the target antigens) (**strong consensus: 97%**) (Table 4)

Details can be found in guidelines for the specific AIBD, that is, PV, PF, PNP, BP, MMP, LAD and EBA as well as in a recent overview.^{18–22,24,25}

Interpretation of skin biopsy, DIF, IIF and ELISA findings in AIBDs

Table 4 and Figures 1 and 2 show histopathological characteristics, DIF, IIF and ELISA findings seen in various AIBDs.

TABLE 4 Histopathological characteristics, DIF, IIF and ELISA findings in AIBD.

AIBD	Histopathology	DIF	IIF	IIF-SS	ELISA/IIF1
Subepidermal AIBDs	Lesional skin with intact blister	Perilesional skin	Monkey esophagus or human skin or monkey liver**	Monkey or Human skin	MBL/Euroimmun
Linear IgA bullous disease (LAD)	Subepidermal blister with a predominant neutrophilic infiltrate in the upper dermis	Linear IgA (predominant over IgA) at DEJ n-serrated pattern	Linear IgA at DEJ	Epidermal binding of IgA	Anti-BP180 IgA ² , anti-BP230 IgA
Bullous pemphigoid (BP)	Subepidermal blister with eosinophil- or lymphocyte-rich infiltrate in the upper dermis and/or eosinophilic spongiosis.	Linear IgG (predominant over IgA)/C3 at DEJ n-serrated pattern	Linear IgG at DEJ	Epidermal binding of IgG	Anti-BP180 IgG, Anti-BP230 IgG
Mucous membrane Pemphigoid	Subepidermal blister with a predominantly neutrophilic and lymphocytic infiltrate and lamellar fibrosis	Linear IgG/C3 and or IgA at DEJ n- or u-serrated pattern (not visible in mucosal biopsies)	Linear IgG or IgA at DEJ	Epidermal or dermal binding of immuno-reactants	Anti-BP180 IgG ^{2,3} , anti-BP230 IgG ⁴ , anti-laminin 332 IgG, anti-type VII collagen IgG, (anti- $\alpha 6\beta 4$ integrin IgG)
Epidermolysis bullosa acquisita	Two types: 1. Subepidermal blister that is pauci-inflammatory and resembles epidermolysis bullosa 2. Subepidermal blister with neutrophil- and eosinophil-rich infiltrate resembling BP or LAD	Linear IgG/C3/IgA at DEJ u-serrated pattern	Linear IgG/IgA at DEJ (Mostly negative)	Dermal binding of immuno-reactants	Anti-type VII collagen IgG or IgA ⁵
Dermatitis herpetiformis	Dense clusters of neutrophils with scattered eosinophils in the dermal papillae	Granular IgA at the dermal papillae or along the DEJ	Anti-endomysium IgA	-	Anti-TG2 IgA, anti-TG3 IgG, anti-deamidated Gliadin IgA
Intraepidermal AIBDs	Lesional skin with intact blister	Perilesional skin	Monkey esophagus		
Pemphigus vulgaris (PV)	Suprabasal cleft with acantholysis, epidermal exocytosis of inflammatory cells with predominant eosinophils	IgG and/or C3 deposits in the cell surface of epidermis/epithelium	IgG against surface proteins of epidermal keratinocytes	NI	Anti-Dsg 3 IgG $\pm$ anti-Dsg 1 IgG
Pemphigus foliaceus (PF)	Acantholysis and subcorneal cleft	IgG and/or C3 deposits in the cell surface of epidermis	IgG against surface proteins of epidermal keratinocytes	NI	Anti-Dsg 1 IgG
Paraneoplastic pemphigus	Suprabasal cleft with acantholysis, dyskeratosis, lichenoid interface dermatitis	IgG and/or C3 staining ICF $\pm$ along DEJ	IgG against plakins proteins on rat bladder	NI	Anti-Dsg3 IgG, anti-envoplakin IgG, + anti-Dsg1 IgG

Abbreviations: AIBD, autoimmune blistering disorders; DEJ, dermo-epidermal junction; DIF, direct immunofluorescence; ELISA, enzyme-linked immunosorbent assay; ICF, intercellular fluorescence; IIF, indirect immunofluorescence; IIF-SS, indirect immunofluorescence with salt splitting; TG, transglutaminase.

¹Based on the Biochip™ technology.

²Main epitopes are localized outside the NC16A domain and are not covered by commercial ELISA/IIF tests.

³Also exclusive or predominant IgA reactivity against BP180 may be detected.

⁴Mostly in conjunction with anti-BP180 reactivity.

⁵Anti-type VII collagen IgA cannot be detected by commercial tests.

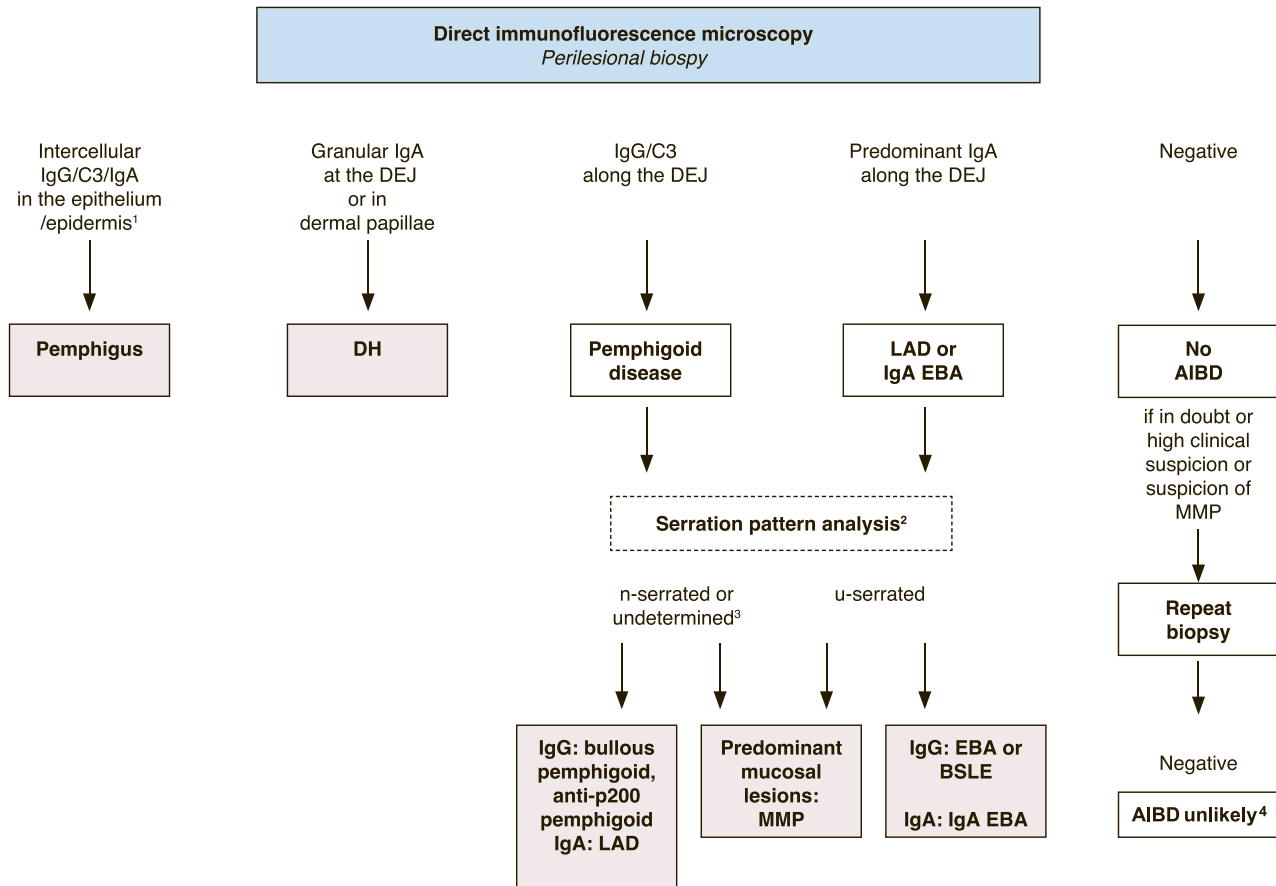


FIGURE 1 Diagnostic algorithm of direct immunofluorescence (IF) microscopy. ¹In paraneoplastic pemphigus, a combined pattern with anti-DEJ deposits may be present; ²in mucosal biopsies, pattern analysis is not possible; ³so far, in none of the undetermined cases, an EBA or BSLE has been diagnosed based on serum anti-type VII collagen IgG/IgA; ²⁹ ⁴unless in ocular MMP, where direct IF can be repeatedly negative; in these cases, diagnosis of MMP can be made based on the clinical picture and the clinical and histopathological exclusion of differential diagnoses. ²⁰ Adopted from van Beek et al. ²⁴⁸ AIBD, autoimmune blistering disease; BSLE, bullous systemic lupus erythematosus; EBA, epidermolysis bullosa acquisita; DH, dermatitis herpetiformis; DEJ, dermal-epidermal junction; LAD, linear IgA disease; MMP, mucous membrane pemphigoid.

Patient's pre-treatment assessment

To decide on the treatment plan, it is recommended to record the following parameters (**strong consensus: 100%**):

• Age of the patient
• Localized/generalized disease
• Slowly progressive/rapidly progressive
• Mucosal involvement
• Acute infections
• Associated comorbidities, e.g. obesity, hyperlipidemia, juvenile onset diabetes, and type 2 allergies
• Quality of life impairment

Since no validated scoring system is available for AIBDs in children, it is recommended to define (**consensus: 90%**)

• "Mild disease" as localized or few non-localized lesions (not more than 10 small lesions; no more than 5% of body surface; no severe pain impairing food intake and/or requiring systemic analgesics; no dysphagia, no involvement of larynx, esophagus, trachea, cornea, and conjunctiva)
• Moderate/severe disease covers all patients not corresponding to "mild" disease

Therapeutic management

Data in children are based on case series. Because of the rarity of these disorders, no controlled therapeutic trials have been published on AIBDs in children. All treatments are used off-label. Further details can be found in the supplement.

Vaccination

• It is recommended to update the immunization schedule prior to any immunosuppressive treatment although this may not be feasible in severe cases requiring rapid treatment (strong consensus: 100%)
• It is recommended to postpone live vaccines (like ROR, chickenpox, BCG, yellow fever, etc.) during treatment with certain immunosuppressive drugs used for AIBD (strong consensus: 100%)

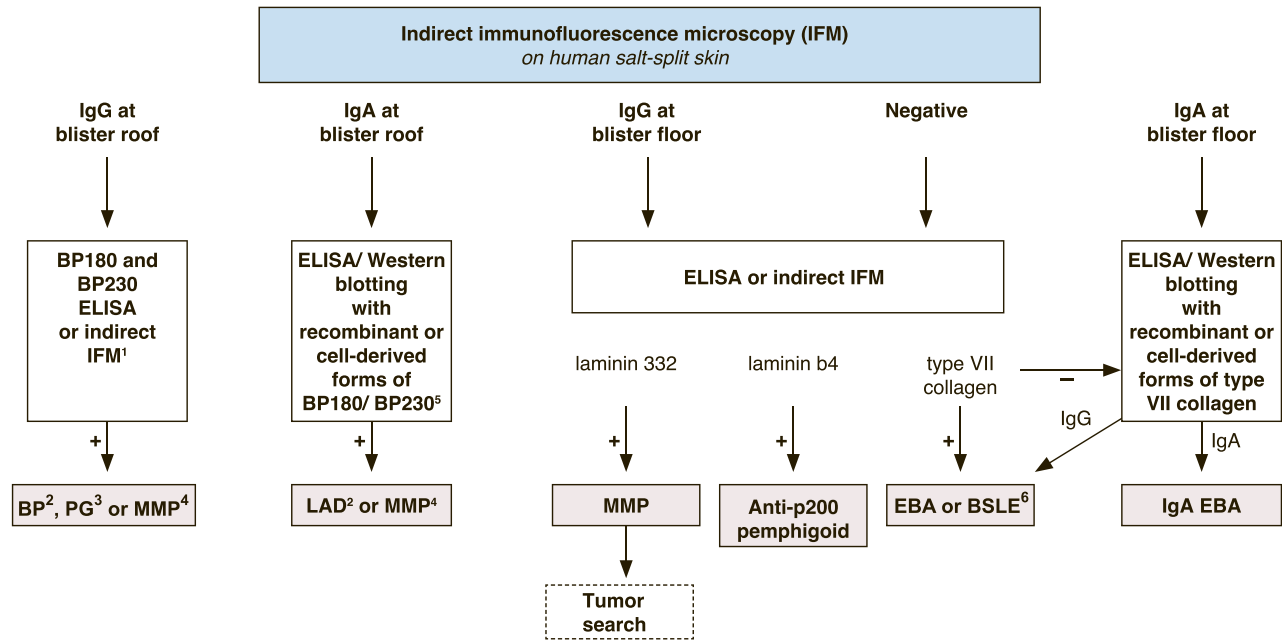


FIGURE 2 Diagnostic algorithm for serum autoantibodies in pemphigoid diseases. ¹Commercial assays are available, ²with predominant skin lesions, ³with pregnancy, ⁴with predominant mucosal lesions, ⁵in-house assays only available in specialized laboratories, ⁶SLE needs to be present. Adopted from van Beek et al.²⁴⁸ BP, bullous pemphigoid; BSLE, bullous systemic lupus erythematosus; DEJ, dermal–epidermal junction; DH, dermatitis herpetiformis; EBA, epidermolysis bullosa acquisita; LAD, linear IgA disease; MMP, mucous membrane pemphigoid.

Follow-up/monitoring

Recommendations for follow-up:

- During active disease, it is recommended to follow-up patients every 2-4 weeks depending on disease severity, then every 6-8 weeks until complete remission and every three months after stopping the treatment to monitor for any relapse (**consensus: 90%**). This proposed schedule might be adapted depending on disease severity and the evolution of the disease
- It is recommended to reduce systemic corticosteroids by 25% every 2-4 weeks starting two weeks after control of disease activity* (**strong consensus: 97%**)
- It is recommended to educate the patient/parents about possible adverse reactions and to contact the treating physician in case of any relapse or adverse reaction (**strong consensus: 100%**)
- It is also recommended to monitor for treatment side effects and sequelae of more severe disease, especially failure to thrive, impaired psychomotor development and quality of life impairment (**strong consensus: 100%**)
- It is recommended to manage patients with corticosteroid side effects and those with a post-rituximab hypogammaglobulinemia in collaboration with pediatric endocrinologists and hematologists, respectively (**strong consensus: 97%**)

*For definition, see reference.³³

Family information

- It is necessary to inform patients and their families about the disease, its prognosis, the treatments, their possible side effects, and the examinations that will be performed during the follow-up to monitor the activity of the disease and detect possible complications (**strong consensus: 100%**)
- It is recommended to inform patients and their families about patient associations if available (**strong consensus: 100%**)

AIBDs OF Dermal-Epidermal Junction (DEJ)

AIBDs of DEJ are characterized by subepidermal blisters with circulating antibodies against components of the basement membrane zone (BMZ).³² The classification and management are based on clinical, histological and immunofluorescence (IF) findings, though often features overlap between the conditions. Various AIBDs of DEJ encountered in children include

- a. Linear IgA Dermatitis (LAD) of childhood
- b. Bullous pemphigoid (BP)
- c. Mucous membrane pemphigoid (MMP)
- d. Epidermolysis bullosa acquisita (EBA)
- e. Dermatitis herpetiformis (DH)

Rarely, cases with anti-p200 pemphigoid³³ and bullous SLE^{34,35} have been described.

Linear IgA Dermatitis (LAD) of childhood

Introduction

LAD is a rare AIBD characterized by exclusive or predominant linear deposition of IgA along the BMZ.³⁶ It is the most commonly encountered AIBD among children and is also referred to as chronic bullous disease of childhood. It was first reported in the 1970s.³⁷ Most of the data are derived from small series or case reports. Since 1988, there are about a dozen series with ≥10 patients (10–39 cases),^{4,6,10,11,38–46} others include 3–7 patients,^{5,8,9,12,47–50} and most are published as isolated case reports with 1–2 patients.^{51–79}

Clinical presentation (see supplement)

Key points:

- Diagnosis of LAD is highly suspected in a child presenting with grouped, tense blisters localized on the lower abdomen, perioral area, and perineum with an annular ‘rosette-like’/‘crown of jewels’/‘string of pearls’ arrangement.
- Cases of neonatal LAD need special cautions because of the high risk of mucosal involvement and associated systemic complications leading to high morbidity and even mortality.

Pathogenesis (see supplement)

Key points:

- Precipitating factors may include preceding infection, drug intake and vaccination.
- Inflammatory bowel disease (Crohn's disease/ulcerative colitis) and coeliac disease can be associated.
- The major target antigens are the 120-kDa (LAD-1) and 97-kDa (LAD-97) ectodomains of BP180.

Diagnosis

After clinical suspicion, the diagnosis of LAD is confirmed by histopathology, DIF and serology as detailed in the recent European guidelines for LAD.³⁶ For clinical evaluation refer to ‘[Interrogation and clinical examination in the setting of a child with AIBD](#)’ (1–3). For confirmation of diagnosis, interpretation of results refer to ‘[Diagnostic management](#)’ (1–3) and [Table 4](#). Further details are shown in the supplement.

Key points:

- DIF shows linear deposition of IgA in all skin biopsies and a weaker deposition of IgG and C3 in some of them.
- IIF on NaCl-split skin shows deposition of IgA antibodies along the roof of the blister.
- The ‘n’ serration pattern on DIF favours LAD. The ‘u’ serration pattern and IgA binding to the blister floor by split-DIF favour IgA-EBA.
- Patients with predominant mucosal lesions are diagnosed as MMP.
- Patients with predominant or exclusive IgA reactivity against type VII collagen are diagnosed as IgA EBA.

Recommendations for diagnosis of LAD (based on the EADV guidelines on LAD)²⁹

It is recommended to make the diagnosis of LAD if criteria 1 and 2 are fulfilled (**consensus: 94%**):

1. Clinical manifestations compatible with LAD. Mucous membrane involvement must not be predominant over skin involvement
2. Positive DIF showing solely linear deposits of IgA along the DEJ, or of both IgA and IgG. In the latter case, fluorescence intensity of the IgA deposits must be stronger than that of IgG

- In case DIF (criterion 2) is negative, it may be considered to repeat the biopsy for DIF (**strong consensus: 100%**)

If DIF is negative or not possible to be performed on site, it is recommended to take into account the following criteria for the diagnosis (**consensus: 90%**):

- Clinical manifestations compatible with LAD (non-predominant mucous membrane involvement) AND histological picture of subepidermal blister with a predominant or exclusive infiltrate of neutrophils, which is suggestive of LAD although non-specific AND
 - Positive IIF using salt-split normal human skin demonstrating IgA binding to the epidermal side or a combined epidermal-dermal staining AND/OR
 - Detection of circulating IgA autoantibodies against LAD-1, LAD97, BP180, BP230

Pretreatment assessment

For pretreatment assessment, refer to ‘[Patient's pre-treatment assessment](#)’.

Treatment

Infection prior to the onset of LAD, if present and not already treated, should be treated. If the patient is receiving a drug which is suspected to be a trigger, it should be immediately stopped.⁸⁰ Patients with mild disease can be treated as outpatients but those with generalized, rapidly progressive disease and oral involvement should be hospitalized, as in addition to the treatment of LAD, they need supportive care with enteral and/or parenteral therapy due to poor intake and other relevant inpatient care. A treatment algorithm for LAD is proposed in [Figure 3](#).

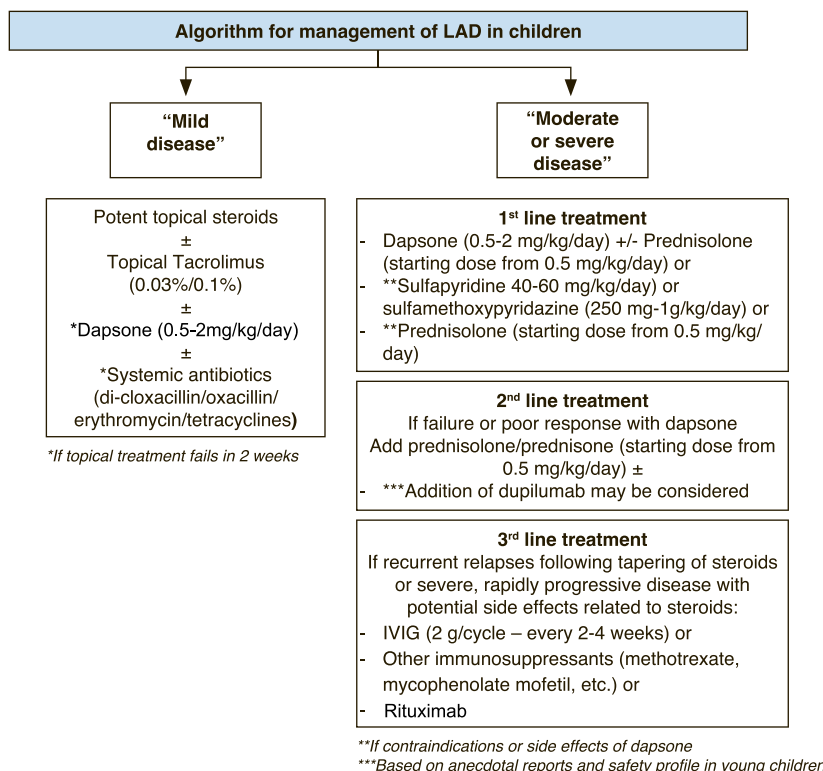


FIGURE 3 Proposed treatment algorithm for linear IgA bullous disease (LAD) in children.

Treatment of mild disease:

Topical corticosteroids (TCS, mid-potency to potent) alone or with topical tacrolimus (0.03%/0.1%), antibiotics or dapsone have been used successfully in patients with mild disease.^{4,10,11,41,51,52,81}

Systemic antibiotics including erythromycin, oxacillin, flucloxacillin, miocamycin and tetracyclines with nicotinamide have been used either alone or in combination with TCS in paediatric LAD patients with variable responses.^{7,10,11,42,52,54,81,82} Tetracyclines should not be used for children below the age of 10 years. Doxycycline has been reported to be safe for older children above 8 years of age.⁸³ How systemic antibiotics exert their role in LAD cases is not clear. It is proposed to be related to their anti-inflammatory activity.

Recommendations for treatment of LAD patients with mild disease:

Topical corticosteroids (class II-III) are recommended in mild cases or in the early stages of disease (strong consensus: 100%).
Topical tacrolimus (0.03% or 0.1%) may be considered in mild cases or in the early stages of disease (consensus: 84%).
Dapsone is recommended as second line when topical treatment is not sufficient* to control disease activity (strong consensus: 100%).
Dapsone may also be recommended as first line in combination with topical corticosteroids (class II-III) or tacrolimus (strong consensus: 97%).
Systemic antibiotics including macrolides, and tetracyclines (in children above the age 10 years) may be considered in case of contraindications to or adverse effects of dapsone (consensus: 84%).

*After 2 weeks.

Treatment for moderate-to-severe disease:

First-line treatment:

Dapsone has been established as a first-line treatment for LAD based on the data available from both large and small series as well as case reports.^{8-11,45,80,81,84,85} The recommended doses range from 0.5 to 3 mg/kg/day after exclusion of G6PD deficiency to avoid severe hemolysis and methemoglobinemia.⁸⁶ Patients should be ideally started on a low dose that should be escalated based on clinical response and tolerance.⁸⁵ Dapsone has anti-inflammatory activity, inhibiting neutrophil chemotaxis, the release of leukotrienes and prostaglandins, and reduces tissue damage that is mediated by neutrophils and eosinophils.⁸⁷ Most patients under dapsone show a rapid improvement within 2–4 weeks. A great level of caution and close follow-up are required for hemolysis and methemoglobinemia, particularly in G6PD-deficient patients. Hemolytic anaemia may occur acutely or within the first 2 weeks of treatment in patients with G6PD deficiency.¹⁰³

It is necessary for patients started on dapsone to perform complete blood count, differential white blood cell count, and reticulocyte count every two weeks while escalating the dose of dapsone to reach the optimal dose; followed by monthly testing until month 6, and once every three months thereafter. G6PD deficiency contra indicates the use of dapsone (strong consensus: 97%)
While on high doses of dapsone (≥ 1.5 mg/kg/d), it is recommended to monitor methemoglobinemia after 1 and 4 weeks and in case of clinical signs of methemoglobinemia (cyanosis, breathlessness) (consensus: 87%)

In places without dapsone availability or if the patient shows side effects to dapsone, other sulfonamides including sulfapyridine (40–60 mg/kg/day) and sulfamethoxy-pyridazine (250 mg–1 g/day) have been used.^{10,11,41,81,85,88} Sometimes size of tablets is an issue for small children, but liquid oral preparations can be manufactured in collaboration with clinical pharmacists. Regular monitoring is required while using other sulfonamides as they share the same profile of adverse effects with dapsone and patients should be closely monitored for the risk of cross sensitivities.

Oral nicotinamide alone was observed to be effective in one report.⁵⁶

Recommendations for first-line treatment of moderate and/or severe LAD:

- | |
|---|
| <ul style="list-style-type: none">Escalating doses of dapsone (from 0.5 to 2 mg/kg/day) ± topical or systemic corticosteroids* (starting dose from 0.5 mg/kg) are recommended as first-line treatment (strong consensus: 97%) |
| <ul style="list-style-type: none">When dapsone is not available or cannot be used due to hypersensitivity, sulfonamides including sulfapyridine (40-60 mg/kg/day) and sulfamethoxypyridazine (250 mg – 1g/day) or prednisone alone may be recommended (consensus: 90%) |

*In particular in extensive disease.

Second-line treatment (see supplement for further details):

Recommendations for second-line treatment of moderate and/or severe LAD:

- | |
|---|
| <ul style="list-style-type: none">Prednisolone/prednisone (starting dose, 0.5 mg/kg/day) is recommended for patients who do not respond well to dapsone/sulfonamide monotherapy (strong consensus: 100%) |
| <ul style="list-style-type: none">Addition of dupilumab (off-label) may be considered due to its good safety profile even based on only anecdotal reports (consensus: 92%) |

Third-line treatment (see supplement for further details)
Recommendations for the third-line treatment of moderate and/or severe LAD:

- | |
|---|
| <ul style="list-style-type: none">Addition of IVIG (2 g/kg/cycle), mycophenolate mofetil (20 - 40 mg/kg or 300-400 mg/m²), or rituximab (4x 375 mg/m² body surface) may be recommended (strong consensus: 97%) |
| <ul style="list-style-type: none">Addition of colchicine (0.5-1 mg/day) may be considered (consensus: 79%) |

Duration of treatment and follow-up: There is no consensus statement about how long the treatment should be continued in LAD cases. It is recommended to tailor the duration of *treatment* depending on the severity of the disease,

response to treatment and course of the disease. LAD in children generally has a good prognosis. Most patients have been reported to experience remission within 3–4 years.^{89,90}

Bullous pemphigoid

Introduction

BP, though the most frequent AIBD of the DEJ among elderly individuals, is rarely reported in minors. More than 100 cases of BP have been reported in children and most are published either as very small series or isolated case reports.^{4-12,91-132}

It appears that an increasing number of cases of infantile BP are reported recently.¹²⁴

Clinical presentation (see supplement for further information)

Key points:

- BP is rare in children. Pruritic tense blisters and urticarial lesions are suspicious of BP.
- Two age peaks are mostly observed: (1) infantile BP cases usually seen below the age of 1 year with lesions predominantly distributed on the acral areas, face, palms and soles and (2) juvenile BP with a peak around 8 years resembling BP in adults.
- Prognosis is better compared to adults due to absence of comorbidities such as neuropsychiatric disorders, cardiovascular and renal disease and diabetes mellitus.

Pathogenesis

The pathogenesis of BP has not yet been fully elucidated.¹³³ Drugs, infections and vaccination are speculated as primary triggers. Further details can be found in the supplement.

Diagnosis

For confirmation of diagnosis interpretation of results refer to ‘[Diagnostic management](#)’ (1–3) and [Table 4](#). The diagnosis is confirmed by DIF showing linear deposition of C3 and IgG at DEJ in a non-serrated pattern. Serology shows IgG at the epidermal side of human salt-split skin by IIF and IgG against recombinant BP180 NC16A and/or BP230 by ELISA/ IIF (Biochip™).

Recommendations for diagnosis of BP (based on the EADV guidelines on BP)¹⁹

It is recommended to make the diagnosis of BP based on the following criteria: compatible clinical picture with tense blisters and/or erosions predominating over mucosal lesions, (ii) positive DIF and (iii) presence of serum IgG antibodies labelling the epidermal side of salt-split skin by IIF microscopy and/or reacting with BP180 and/or BP230 by ELISA or IIF (strong consensus: 100%)
In patients with non-bullous lesions, the diagnosis of BP can be accepted in patients with (i) positive DIF and (ii) presence of circulating IgG auto-antibodies labelling the epidermal side of salt-split skin by IIF microscopy and/or (iii) reacting with BP180 and/or BP230 by ELISA or IIF (strong consensus: 100%)
- When DIF studies are negative, the diagnosis of BP can be accepted in patients with: - Suggestive clinical picture (i.e., tense blisters) and - Consistent histopathological findings (subepidermal cleavage) with eosinophilic infiltrate and/or - Presence of circulating IgG autoantibodies binding to the epidermal side of salt-split skin by IIF microscopy studies and/or iv) serum reactivity with BP180 and or BP230 by ELISA or IIF (strong consensus: 100%)
It may be considered to perform additional tests, e.g. ELISA or immunoblotting using recombinant or cell-derived forms of BP180 and/or BP230 when commercial ELISA/ IIF systems do not detect anti-BP180/ BP230 antibodies (strong consensus: 100%)

Pretreatment assessment

For pretreatment assessment, refer to 'Patient's pre-treatment assessment'.

Treatment

Patients with severe, generalized disease, poor intake due to oral mucosal involvement and associated comorbidities including florid infections or some systemic ailment should be hospitalized under joint dermatologist and paediatrician care. In rare cases with extensive disease, parenteral support is required. Various treatment modalities including potent topical corticosteroids,^{10,105,112} systemic corticosteroids,^{7,9,10,118,123,126,134,136,141,157} dapsone,^{7,8,126,141,153,157} azathioprine,^{8,92,119} methotrexate,^{9,115,125} IVIGs,^{9,11,98–100,109,116,118,121} cyclosporine^{121,128,129} and rituximab^{106,130} have been used in children with BP. There is no consensus on the treatment approach. An algorithm was proposed by Schwieger-Briel et al.¹⁰⁸ for infantile BP. Potent topical corticosteroids were rarely used in childhood BP cases with localized disease. Caution is required when corticosteroids are used for long-term in infants and small children due to related side effects.¹⁵⁸ Systemic steroids alone or in combination with dapsone were used in most reports. IVIG were used in several reports for infantile BP with severe, rapidly progressive disease when there was a failure to respond to steroids and dapsone. Methotrexate, cyclosporine and mycophenolate mofetil were rarely used, and rituximab was used exceptionally when other treatment modalities failed. Recently, baricitinib was used off label in one refractory case of infantile BP.¹³⁴ Data on the use

of dupilumab in paediatric cases of BP are scarce.¹³⁵ Recently, dupilumab has been used with success in several cases of BP in the elderly as monotherapy or an adjuvant with a fair response.^{136–140} Dupilumab seems to be a safe option since it does not lead to immunosuppression and is approved for use in children with moderate to severe atopic dermatitis from the age of 6 months onward.¹⁶¹

Treatment recommendations for BP:

Treatment of Mild BP

- Topical corticosteroids (class III-IV) are recommended to achieve control of disease activity (**consensus: 94%**)

Treatment of Moderate and/or Severe BP

First-line treatment

- Prednisolone/prednisone (starting dose, 0.5 mg/kg/day) alone or in combination with dapsone (0.5–2 mg/kg/day) is recommended (**consensus: 90%**)

Second-line treatment

- Addition of other immunosuppressants (azathioprine [1.5 – 2 mg/kg/day]/ methotrexate [10 – 15 mg/m²/wk]/ mycophenolate mofetil [20 – 30 mg/kg or 300–400 mg/m²] or IVIG (2 g/kg/cycle) are recommended in patients who fail to improve with systemic steroids and dapsone or have a contraindication to dapsone (**consensus: 87%**).
- Dupilumab (off-label), though rarely reported in BP in children, based on the literature in adults and the favorable safety profile for children >6 months, may be recommended (**strong consensus: 96%**).

Third-line treatment

- Rituximab (375 mg/m² weekly for 4 weeks) is recommended in cases who fail to respond to all other treatments (**consensus: 94%**).
- In relapsing patients, it is recommended to use a second or a third-line treatment (**consensus: 87%**).

Mucous membrane pemphigoid

Introduction

MMP is a clinically and immunopathologically heterogeneous subepidermal blistering disease characterized by a chronic course with predominant involvement of mucous membranes and a tendency to scarring.¹⁴¹ Most commonly involved mucosal areas are oral mucosa, eyes, nose, and anogenital regions. Other rarely affected mucosal sites include the oesophagus, larynx and pharynx. The skin involvement is usually rare and mild. The disease is well characterized in adults and recently, in S3 guidelines initiated by the EADV task force, the clinical spectrum and diagnostic strategies for MMP have been described in detail.^{20,21} Data on MMP are scarce in children; around 50 cases between the ages 1 and 18 years have been reported in the literature, and most are published as isolated case reports.^{142–172}

Clinical presentation (see supplement)

Key points:

- Diagnosis of MMP requires exclusive or predominant mucosal lesions over skin involvement.
- Histological and immunological features of MMP in children are comparable to adults.
- The disease can present with mono-site involvement affecting oral, genital or ocular mucosa, or with two or more sites with or without skin lesions. Oral involvement is most common, followed by vulvar and ocular.
- MMP localized to vulvar areas is often reported in young girls and needs to be differentiated from lichen sclerosis, child abuse and trauma.

Pathogenesis

The exact pathogenesis is not clear. Isolated cases associated with drug intake or coeliac disease have been reported.¹⁵⁶ Like in adults, BP180 is the main target antigen in children and adolescents with MMP.¹⁵⁸ BP230 can also be targeted, mostly in addition to BP180.¹⁵⁸ Rarely, cases of anti-laminin 332 MMP were reported in children.¹⁶¹ While in adults about a quarter of MMP cases with anti-laminin 332 reactivity are associated with a mostly solid malignancy,^{173–176} this association has not been described in paediatric patients.

Diagnosis

As in adults, diagnosis of MMP requires exclusive or predominant mucosal lesions over skin involvement. Further details are indicated in the supplement.

Recommendations for diagnosis of MMP (based on the EADV guidelines on MMP)²¹

- It is recommended to make the diagnosis of MMP based on the following criteria: (i) compatible clinical picture with predominant mucosal over skin involvement and (ii) binding of IgG, IgA, and/or C3 at the BMZ of a perilesional skin or mucosal biopsy by DIF microscopy (**strong consensus: 100%**).
- When DIF studies are negative, it is recommended to take a new biopsy preferentially from a different site for DIF microscopy (**strong consensus: 100%**).

- When DIF microscopy of the second biopsy is also negative, it may be considered to obtain a third biopsy for DIF (**consensus: 90%**).
- Alternatively, it may be recommended to base the diagnosis on (i) compatible clinical picture with predominant mucosal over skin involvement and (ii) serology (**strong consensus: 100%**). For a positive serology the following assays need to be reactive.
 - a. commercially available BP180 NC16A ELISA or IIF
 - b. commercially available type VII collagen ELISA or IIF
 - c. commercially available laminin 332 IIF or
 - d. IIF on human salt-split skin (anti-BMZ IgG or IgA) or IIF on monkey esophagus (anti-BMZ IgG or IgA)
 - e. plus commercially available BP230 ELISA or IIF
 - f. plus one in-house assay employing recombinant or cell-derived forms of BP180, laminin 332, and type VII collagen
- Testing for anti- $\alpha 6 \beta 4$ integrin antibodies is not recommended due to the lack of established assays validated in the diagnosis of MMP. (**consensus: 94%**).

In the EADV guideline, of ocular MMP (defined as MMP with lesions restricted to the conjunctiva and cornea), the diagnosis is accepted also with repeated negative DIF and negative serology when a list of differential diagnoses has been excluded both clinically and histopathologically. In ocular MMP, the initial biopsy for DIF may be taken from uninvolved oral mucosa instead of perilesional conjunctiva of the less affected eye.^{20,21}

Pretreatment assessment

For pretreatment assessment, refer to '[Patient's pre-treatment assessment](#)'.

Treatment

Since most data on MMP in children is based on reports of a small number of patients or isolated case reports, no straightforward consensus about the treatment guidelines can be drawn from the available data.

Particularly in case of ocular and/or genital MMP, a joint approach of management involving a paediatric ophthalmologist and gynaecologists is recommended.

Topical corticosteroids and or topical tacrolimus (0.1%) were observed to be effective in some cases of MMP localized to oral or vulvar mucosa.^{149,152,154,155,162} Topical cyclosporine drops alone or in combination with systemic treatment were reported to be helpful in ocular MMP.^{158,172} Based on the limited number of cases, the level of evidence cannot be defined with confidence but based on combined experience with isolated reports in children and adult data, dapsone (2–3 mg/kg/day)^{20,153,155,160} alone or in combination with topical corticosteroids continues to be the first line of treatment for moderate to severe disease. In patients refractory to dapsone, systemic prednisolone (1–2 mg/kg/day) with or without other immunosuppressants (azathioprine, mycophenolate mofetil) was the treatment required to control the disease and prevent long-term sequelae related to scarring.^{149,158,161,164,172} Based on data in adults, IVIG and rituximab may be indicated in refractory cases. Rituximab was reported to show a good response in

cases with resistant MMP in children.^{163,165,172} Surgical intervention by an ophthalmologist may be required in patients with entropion and trichiasis.¹⁶⁵

Treatment recommendations for MMP

Topical corticosteroids (class III) and/or topical tacrolimus (0.1%) are recommended for mild MMP localized to oral or vulvar mucosa, and as an adjunct treatment in moderate-to-severe disease (consensus: 90%).
Topical corticosteroids, topical cyclosporine, lubricant drops, gels and ointments may be recommended as adjunct treatment in ocular MMP (strong consensus: 100%).
Dapsone (from 0.5 to 2 mg/kg/day) ± prednisolone/prednisone (starting dose, 0.5-1 mg/kg/day) are recommended as first-line treatment for moderate to severe disease (consensus: 94%).
Additional immunosuppressants (methotrexate (10-15 mg/m²/wk)/ azathioprine (1.5-2 mg/kg/day)/ mycophenolate mofetil (20-30 mg/kg/day) may be recommended in patients who fail to improve with dapsone or have a contraindication to dapsone (strong consensus: 100%).
IVIg (2 g/kg/per cycle) or 4 weekly infusions of rituximab (375 mg/m²) are recommended in cases who fail to respond to all other treatments (strong consensus: 100%).
In ocular involvement, close collaboration with an ophthalmologist experienced in MMP is necessary (strong consensus: 97%).
In severe ENT involvement, close collaboration with an ENT specialist experienced in MMP is recommended (strong consensus: 100%).

Epidermolysis bullosa acquisita

Introduction

EBA is a rare subepidermal AIBD caused by antibodies targeting type VII collagen (ColVII), the main component of anchoring fibrils.^{177,178} EBA is primarily a disease of adults, and less than 50 cases have been reported among children.¹⁷⁹⁻²⁰³ Childhood EBA has been reported to occur between 2 weeks to 17 years of age. A recent meta-analysis revealed that, among patients diagnosed with EBA, 4.6% were younger than 17 years.²⁰⁴ Hignett and Sami recently reviewed the literature on 40 cases of EBA published in the literature.²⁰⁰ Males and females were almost equally affected.

Clinical presentation

See supplement.

Pathogenesis

See supplement.

Diagnosis

See supplement.

Recommendations for diagnosis of EBA (based on an international consensus)²¹⁹

It is recommended to make the diagnosis of EBA based on the following criteria (**strong consensus: 100%**):

- | |
|--|
| <ul style="list-style-type: none"> (i) Compatible clinical picture with skin lesions predominating over mucosal lesions, and (ii) positive DIF and (iii) presence of serum IgG antibodies reacting with type VII collagen by ELISA or IIF |
| <ul style="list-style-type: none"> Or (i) and (ii) positive DIF with a u-serrated pattern |
| <ul style="list-style-type: none"> When DIF studies are negative, it is recommended to perform a new biopsy and to check for any technical problem. In the few patients with persistent negative DIF studies, the diagnosis of EBA can be accepted in patients with: i) suggestive clinical picture and ii) presence of circulating IgG autoantibodies binding to the dermal side of salt-split skin by IIF microscopy studies and iii) serum reactivity with type VII collagen by ELISA or IIF |

Pretreatment assessment

For pretreatment assessment, refer to 'Patient's pre-treatment assessment'.

Treatment

Due to the rarity of the cases both in children and adults, there is no consensus on the treatment of EBA. Most cases (70%) reported in children were treated with a combination of systemic corticosteroids and dapsone with a favourable outcome.²⁰⁰ Rarely corticosteroid or dapsone monotherapy was used. In cases refractory to corticosteroids and dapsone, other immunosuppressants/modulators including tetracyclines, colchicine, azathioprine, mycophenolate mofetil, IVIg, extracorporeal photopheresis or rituximab have been reported to be used in isolated case reports.^{60,188,191,199,200,202} Because of anecdotal reports, it is difficult to determine the best treatment option. Overall, children seem to be having a better prognosis than adults.^{199,200}

Treatment recommendations for EBA

- | |
|--|
| <ul style="list-style-type: none"> Dapsone (from 0.5 to 2 mg/kg/day) ± prednisolone (starting dose, 0.5-1 mg/kg/day) are recommended as first-line treatment (consensus: 94%). |
| <ul style="list-style-type: none"> Additional use of topical corticosteroids may be recommended for the mechanobullous variant (strong consensus: 100%). |
| <ul style="list-style-type: none"> Addition of azathioprine, methotrexate, mycophenolate mofetil or IVIg may be recommended as second-line treatment (strong consensus: 100%). |
| <ul style="list-style-type: none"> Rituximab may be recommended in refractory cases (consensus: 94%). |

Dermatitis herpetiformis

Introduction

DH is a highly pruritic relapsing disorder characterized by a symmetrical polymorphous papulovesicular eruption primarily on the elbows, knees, upper back, buttocks and nuchal areas, and subepidermal granular deposition of IgA

in the papillary dermis.^{205–208} It is caused by hypersensitivity to gluten and is considered an extracutaneous manifestation of coeliac disease (CD). Though overt gastrointestinal symptoms of CD are rare in patients, three-fourths of individuals with DH have villous atrophy in the small bowel, and the rest have CD-like inflammatory changes, most of them responding to a gluten-free diet. DH predominantly affects adults and is more often reported in Europe and the United States among individuals of Northern European descent than in individuals with other origins.^{205–208} The reported prevalence ranges between 1.2 and 75.3 cases per 100,000 and an incidence between 0.4 and 3.5 cases per 100,000 per year. It is rarely reported from Asia and is less common in African-Americans.^{205–208}

DH is rare in children. In a series from Finland, children were observed to constitute 3.8% of the total number of DH cases with an annual incidence of 0.56 cases per 100,000 in the general population.²⁰⁹ Annual incidence of paediatric DH in Finland was six times lower than that of the adult population.²⁰⁹ In another report from Italy, 25% of the 149 cases with DH were diagnosed under the age of 10 years.²⁶ Only a few significant series of DH among children have been published from Europe.^{210–214} Most other paediatric data is derived from small case series or isolated case reports.^{215–229}

In a series describing the whole spectrum of AIBDs in children, DH was most prevalent among children (39%) reported from the USA.⁵ It is the third commonest AIBD (10.5%) in Tunisia following LABD and BP,⁷ fourth common (13%) in France following LABD, BP and pemphigus variants.¹⁰ It was rarely seen (5% of 40 cases) in India,¹² and no paediatric cases were detected in the series reported from Kuwait, Thailand, Singapore, Mali and Turkey.^{4,6,8,9,230}

Contrary to adults where DH is equally prevalent in males and females, DH in children is twice as common in girls than in boys, and a peak is seen between the ages of 2–7 years.^{210,211,213,214}

Clinical presentation (see supplement)

Key points:

- DH should be suspected in children who present with recurrent symmetrical pruritic, polymorphic papulovesicular lesions on the elbows, knees, back and buttocks and rarely on other sites.
- Acral purpura in association with typical rash or alone is a feature seen more often in children.

Recommendations

• It is necessary to take a careful history of gastrointestinal symptoms including recurrent abdominal pain, diarrhea, constipation, loss of weight, loss of appetite, nausea, bloating, gluten intolerance, etc. to be ascertained (strong consensus: 100%).
• It is necessary to assess the following complaints/symptoms: depression, osteoporosis, relapsing oral ulcers, anemia, reduced growth (strong consensus: 100%).

Pathogenesis

See supplement.

Diagnosis

The positivity of DIF is mandatory to confirm the diagnosis of DH, since the serology may be positive in patients with CD without DH and histopathology is not specific enough for DH.²⁰⁶ For confirmation of diagnosis and interpretation of results, refer to ‘Diagnostic management’ (1–3) and Table 4.

Recommendations for the diagnosis of DH (based on the EADV guidelines for DH)²⁰⁶

• A 3–4 mm perilesional skin biopsy for DIF is necessary for the diagnosis of DH to determine the granular deposition of IgA at dermal papillae or DEJ (consensus: 94%).
• It is necessary that at least one serological test is performed in DH if suspected. IIF to determine anti-endomysium IgA antibodies (EMA) or TG2-based immunoassay for IgA reactivity (e.g., ELISA) (strong consensus: 97%).
• It may be considered to determine serum IgA reactivity against TG3 or deamidated gliadin by immunoassay (e.g. ELISA) (strong consensus: 97%).
• Small bowel biopsy is recommended except in children with very high levels of anti-TG2 IgA (>10 times of the upper detection limit) confirmed by EMA positivity from a separate blood sample²³¹ (strong consensus: 97%).
• HLA DQ2 and DQ8 testing may be considered in selected cases to exclude DH or CD due to its high negative predictive value (strong consensus: 97%).
• It is recommended to make the diagnosis of DH when the following two criteria are fulfilled (strong consensus: 100%): - Clinical manifestation compatible with DH - Positive DIF microscopy

Pretreatment assessment

For pretreatment assessment, refer to ‘Patient’s pre-treatment assessment’.

Treatment

It is necessary that only limited therapy is prescribed until all diagnostic steps are done, because both gluten-free diet (GFD) and dapsone treatment can modify or even falsify diagnostic results.

GFD should be a lifelong diet. Adhering strictly to a GFD forms the cornerstone of treatment of DH.^{206,210–213} The statement ‘gluten-free’ may only be made if the food contains no more than 20mg/kg of gluten.²⁰⁶ Patients are advised to avoid any food containing such as those made from wheat, barley, rye and malt. Safe alternatives include whole-grain rice, maize, potatoes and vegetables. In patients with a rash that is not responsive to GFD, dapsone is the treatment of choice. Dapsone (2mg/kg/day or 4mg/kg weekly) has shown efficacy in relieving symptoms of

pruritus within hours and reducing new blister formation within 24–36h, making it an effective option for acute inflammatory phases of disease and aiding in early stages of GFD initiation.

With treatment (GFD ± dapsone), all skin manifestations resolve within 1–6 months. Dapsone can be titrated depending upon the clinical response.

Treatment recommendations for DH

Adhering to GFD is necessary in all the patients with DH (consensus: 90%).
In patients with severe DH, it may be recommended to start on a combination of GFD and dapsone (up to 2 mg/kg/day) (consensus: 94%).
Dapsone (from 0.5 to 2 mg/kg/day) in addition to GFD is recommended in patients who have not shown improvement following GFD (after 1–6 months based on severity of signs and symptoms) (consensus: 87%). Dapsone is contraindicated in case of G6PD deficiency.
Sulfasalazine, sulfapyridine (40–60 mg/kg/day), sulfamethoxypyridazine (250 mg–1 g/day) although the galenic formulation is not suitable for children, and tetracyclines (4 mg/kg/day) with niacinamide (after the age 8 years) may be recommended as alternative treatments for DH when dapsone cannot be applied (consensus: 90%).

Follow-up

Patients on treatment with dapsone or other drugs need to be closely monitored for the response and side effects related to them.^{103,206–208,210–213}

Follow-up recommendations for DH

It is recommended to discontinue dapsone after a few months in particular in patients with good adherence to GFD (strong consensus: 97%).
In case of a relapse, it is necessary to check adherence to a GFD (strong consensus: 100%).
It is recommended to insist on the benefit of the GFD (consensus: 94%).
In the absence of improvement of the relapse, it may be recommended to reintroduce dapsone (strong consensus: 100%).

INTRAEPIDERMAL AIBDS

The intraepidermal AIBDs are the pemphigus disorders that are usually caused by autoantibodies targeting cadherin proteins in desmosomes. The various pemphigus disorders include

- f. Pemphigus vulgaris (PV)
- g. Pemphigus foliaceus (PF)
- h. Paraneoplastic pemphigus (PNP)
- i. Neonatal pemphigus (NP)
- j. Pemphigus herpetiformis (PH)
- k. Pemphigus vegetans (P Veg)

Introduction

Pemphigus vulgaris

PV is the most common form of non-endemic pemphigus in children, although it remains very rare. The average age at onset of the first clinical signs of PV has been reported to range from 8.3 to 14.9 years.^{232–234} The sex ratio of paediatric PV is balanced.^{234,235} PV is characterized by the production of autoantibodies directed against desmoglein (Dsg) 3 and sometimes Dsg1, which are responsible for suprabasal acantholysis and the formation of blisters followed by erosions.¹

Pemphigus foliaceus

Two types of PF are described in the literature: an endemic form (also called fogo selvagem) and a non-endemic form. PF is characterized by the production of autoantibodies directed against Dsg1. The endemic form, which has been initially described in South America (Brazil), is multifactorial and involves genetic (HLA-DRB1), immune and environmental factors (in particular the exposure to hematophagous insect bites).^{236,237} The average age at diagnosis is 7.8 years.²³⁷ The non-endemic form of PF is very rare before the age of 18. Two literature reviews have listed a total of 30 cases of paediatric PF.^{238,239} The average age at diagnosis of PF was 7.7 years with a slight male predominance (sex ratio: 1.33/1).

Clinical presentation

PV (see supplement)

Key points:

- Diagnosis of PV is highly suspected (especially in pre-adolescent children and teenagers) in the presence of mucosal involvement, even if isolated with or without flaccid blisters on head and neck area, the trunk and extremities.

PF

PF in children and adolescents is even rarer than in adults. The skin involvement is characterized by crusty erosions which can be limited to seborrheic areas (chest, face, scalp, inter-scapular region) or more rarely disseminated.^{238,240} There is no mucosal involvement. Extensive, extremely severe erythrodermic forms exist and can be life-threatening.²⁴¹

Key points:

- Diagnosis of PF is suspected in children with flaccid blisters and/or erosions and crusted lesions localized to seborrheic areas.

PNP

See supplement.

NP

It appears in the context of poorly controlled PV during the mother's pregnancy.²⁴² However, it may also occur in 25% of cases in neonates whose mother is in clinical remission.²⁴²⁻²⁴⁵ The skin involvement is characterized by flaccid blisters with clear content on non-erythematous skin, quickly leading to post-bullous erosions. The lesions can be localized or generalized and are predominantly located on the trunk, face, scalp and extremities.^{242,246} Mucosal involvement may occur typically presenting as erosions primarily affecting the oral and genital mucosae. The lesions usually disappear within 2–4 weeks and more rapidly under treatment.^{242,247}

Diagnosis

The main histopathological and immunopathological findings of PV, PF and PNP are shown in Table 4.

Recommendations for diagnosis of PV/PF based on the EADV guidelines.¹⁷

- It is recommended to make the diagnosis of PV/PF based on the following criteria: i) Compatible clinical picture and (ii) positive DIF and (iii) presence of serum IgG antibodies labelling the epithelium of monkey esophagus by IIF microscopy and/ or reacting with Dsg1/3 by ELISA or IIF (**strong consensus: 100%**)
- If DIF is negative and a new biopsy is not feasible, it is recommended to make the diagnosis of pemphigus in patients with (**consensus: 94%**):
 - Compatible clinical picture plus corresponding typical histological picture of acantholysis plus significant positive IIF on monkey esophagus. In this particular case, the presence of a typical histological picture of acantholysis is absolutely necessary to exclude an erythema multiforme major.
 - Compatible clinical picture and histopathology plus serum reactivity against recombinant Dsg 1 and/or 3 by ELISA or IIF on cells that express recombinant Dsg1/Dsg

For the diagnosis of PNP, it is recommended to follow the European guidelines for PNP in adults.²²

Pretreatment assessment

For pre-treatment assessment refer to 'Patient's pre-treatment assessment'.

Treatment

PV

Mild PV:

First-line treatment

- Systemic corticosteroid therapy with prednisone/prednisolone alone (without immunosuppressant) at an initial dosage between 0.5-1.0 mg/kg/day is recommended (**strong consensus: 100%**).

Second-line treatment

- In patients initially treated with prednisone/prednisolone alone who either have: ii) persistent disease activity, and detection of anti-Dsg3 serum antibodies, and significant impact on quality of life (child DLQI score), or, ii) in patients with corticosteroid side effect, the addition of rituximab (375 mg/m² weekly for 4 weeks), or mycophenolate mofetil 600 mg/m² twice a day may be recommended to control pemphigus activity, and rapidly taper corticosteroid doses (**strong consensus: 100%**).
- In patients initially treated with prednisone/prednisolone 0.5 mg/kg/day plus rituximab, who have persistent active lesions, it is recommended to increase the dose of prednisone/prednisolone up to 1 mg/kg/day (**strong consensus: 100%**).

Moderate-to-severe PV:

First-line treatment

- Systemic corticosteroid therapy with prednisone/prednisolone at an initial dosage of 1 mg/kg/day is recommended (**strong consensus: 97%**).
- The initial combination with rituximab (375 mg/m² weekly for 4 weeks) or mycophenolate mofetil 600 mg/m² twice a day* may be recommended in order to taper corticosteroids within 6 months (**consensus: 90%**).
- Cyclophosphamide, although reported as effective in the literature, is not recommended in children, due to its significant toxicity, particularly on fertility (**strong consensus: 100%**).

*Mycophenolate mofetil is currently preferred over azathioprine.

Second-line treatment

- In patients who have been initially treated with corticosteroids alone and are not controlled, the following options may be recommended: i) combination with rituximab at a dose of 375 mg/m² weekly for 4 weeks, or ii) conventional immunosuppressant, especially mycophenolate mofetil at a dose of 600 mg/m² twice daily in case of contraindication to rituximab, and/or iii) intravenous pulses of corticosteroids and/or IVIG (2 g/kg/cycle) (**strong consensus: 97%**).
- In patients who are not controlled despite an initial treatment with corticosteroids plus rituximab, the following options may be recommended: i) intravenous pulses of corticosteroids and/or IVIG, ii) second cycle of rituximab between 4 to 6 months after the initial cycle (**strong consensus: 97%**).

PF

Mild PF

First-line treatment

- Topical corticosteroids (class III, IV) alone or in combination with dapsone (starting dose of 0.5 to 1 mg/kg/day), may be recommended in particular in patients with very limited lesions and low levels (or absence) of anti-Dsg 1 antibodies (**strong consensus: 97%**).

Second-line treatment

- In patients whose mild pemphigus has been initially treated with dapsone and/or topical corticosteroids (without oral corticosteroids or rituximab) and have persistent active lesions and detection of anti-Dsg1 serum antibodies and significant impact on quality of life (child DLQI score), oral prednisone/prednisolone 0.5 to 1 mg/kg/day is recommended (**strong consensus: 97%**).
- Rituximab (375 mg/m² weekly for 4 weeks) alone or in combination with oral corticosteroids (prednisone/prednisolone 0.5 -1 mg/kg/day) are recommended in patients with a rapid decrease in order to stop corticosteroids after 3 or 4 months (**strong consensus: 97%**).
- If rituximab is contraindicated or not effective mycophenolate mofetil 600 mg/m² twice a day is recommended (**consensus: 94%**).

Moderate-to-severe PF

Same recommendations as moderate-to-severe PV.

PNP

- It is necessary to manage patients with PNP in collaboration with pediatric hematologists and oncologists and in case of bronchiolitis obliterans with pediatric pulmonologists (**strong consensus: 100%**).

Management of Castleman's syndrome or the underlying haematological disorder is systematic, but specific treatment for pemphigus is necessary:

- High doses of corticosteroids (combined with oncological therapy for associated malignancy as indicated) are recommended (**strong consensus: 100%**).
- The combination of corticosteroids with rituximab or IVIG may be recommended, after discussion in a multidisciplinary meeting with the referring oncologist (**strong consensus: 100%**).

NP

- Due to the usually spontaneously favorable course of the disease, it may be recommended to apply topical corticosteroids (class II) in non-localized disease, for a short period from 1 to 4 weeks (**consensus: 94%**).

Treatment of recalcitrant cases (PV, PF, PNP)

- In patients who are not controlled despite an initial treatment with the combination of corticosteroids and rituximab, a second cycle of rituximab at month 4 or 6 may be recommended (**strong consensus: 100%**).
- Addition of IV pulse of corticosteroids, and/or IVIG infusions may be recommended (**strong consensus: 97%**).

Monitoring and tapering of corticosteroids

Monitoring

The monitoring of treatment is based on the evolution of clinical signs and the ELISA values of anti-Dsg autoantibodies, which usually follow the activity of the disease and are good predictors of treatment response and risk of relapse.

Recommendations for monitoring:

- It is recommended to monitor circulating anti-Dsg1 and 3 antibodies by ELISA during the course of the disease (**strong consensus: 97%**).

Tapering of corticosteroids

The duration of corticosteroid therapy should be as short as possible. However, the tapering of corticosteroid doses must be gradual over several months, with no specific regimen validated in children.

- In patients treated with corticosteroids alone, a progressive tapering over 6 months, then followed by a maintenance dose around 0.1 mg/kg/day for 12 to 18 months may be recommended (**consensus: 90%**).
- In patients treated with the combination of corticosteroids and conventional immunosuppressants or rituximab, a more aggressive tapering of corticosteroids over 3 to 6 months, may be recommended (**consensus: 94%**).
- It may be recommended to measure anti-Dsg1 antibody values before stopping corticosteroid treatment because the persistence of high levels of anti-Dsg1 antibodies by ELISA is a predictor of skin relapses, while the persistence of anti-Dsg3 IgG does not necessarily predict a mucosal relapse (**consensus: 90%**).

CONCLUDING REMARKS

These are the first guidelines on diagnosis and management of AIBDs in children and adolescents prepared by the EADV Task Force of AIBDs. They represent a collective effort to translate the scant but steadily growing body of evidence on AIBDs in children into practical recommendations. Though randomized data were unavailable, we relied on well-documented case series, pharmacovigilance reports and—crucially—the long-standing clinical experience of paediatric dermatologists and the EADV Task Force experts for Autoimmune Blistering Diseases. In summary,

these guidelines provide an actionable framework for clinicians to prompt histological and serological confirmation, combined with age-adjusted systemic and topical therapy, lowering cumulative corticosteroid exposure with the use of steroid-sparing agents whenever feasible. Though dapson, mycophenolate mofetil or azathioprine remain first-line adjuncts, the use of IVIG, rituximab and dupilumab is justified when the disease is refractory or where growth, neurocognitive development or quality of life are threatened. With emerging therapeutics and precision medicine, we commit to reassessing the recommendations at least every 2–3 years—or sooner if pivotal paediatric trials or major pharmacovigilance signals emerge.

AUTHOR CONTRIBUTIONS

All authors met the ICMJE criteria and were involved in data analysis and interpretation as well as critically revising the manuscript. All authors have approved its final version. Further details of the role of each author in the guideline are given in the *Development of the guidelines* section.

AFFILIATIONS

¹As'ad Al-Hamad Dermatology Center, Kuwait City, Kuwait

²Department of Dermatology, French Reference Center for Auto Immune Blistering Diseases, Rouen University Hospital, Normandie University, Rouen, France

³Univ Rouen Normandie, Inserm, Normandie Univ, PANTHER UMR 1234, Rouen, France

⁴Department of Dermatology, Centers of Expertise for Blistering Diseases and European Reference Network for Rare Skin Diseases (ERN SKIN), University Medical Center Groningen, University of Groningen, Groningen, the Netherlands

⁵Dermatology Unit and Genodermatosis Unit, Bambino Gesù Children's Hospital-IRCCS, Rome, Italy

⁶Division of Pediatric Dermatology, Department of Dermatology and Venereology, University of Belgrade School of Medicine, Belgrade, Serbia

⁷St John's Institute of Dermatology, Guy's & St Thomas' NHS Trust, London, UK

⁸Department of Health Sciences, Rare Skin Diseases Unit, P.O. Piero Palagi, Azienda USL Toscana Centro, ERN-SKIN Member, University of Florence, Florence, Italy

⁹Department of Dermatology, Venereology and Leprology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

¹⁰Department of Dermatology, Rare Skin Diseases Center, Istituto Dermopatico dell'Immacolata, IDI-IRCCS, Rome, Italy

¹¹Department of Dermatology and Venereology, Medical Faculty, Medical University – Sofia, Sofia, Bulgaria

¹²Department of Dermatology, Venereology and Allergology, University Hospital Würzburg, Würzburg, Germany

¹³Department of Dermatology and Allergology, Philipps-Universität Marburg, Marburg, Germany

¹⁴Division of Dermatology, Department of Medicine, David Geffen School of Medicine at University of California Los Angeles, Los Angeles, California, USA

¹⁵First Department of Dermatology, Faculty of Medicine, Aristotle University of Thessaloniki, Thessaloniki, Greece

¹⁶Department of Dermatology, Medical Center-University of Freiburg, Faculty of Medicine, University of Freiburg, Freiburg, Germany

¹⁷Department of Dermatology Hospital Alemán, Buenos Aires, Argentina

¹⁸Dermatology Unit, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy

¹⁹Department of Pathophysiology and Transplantation, Università Degli Studi di Milano, Milan, Italy

²⁰Department of Dermatology, St George Hospital, Sydney, New South Wales, Australia

²¹University of NSW, Sydney, New South Wales, Australia

²²Division of Dermatology, Department of Paediatrics, CHU Ste-Justine, Université de Montréal, Montréal, Québec, Canada

²³Division of Dermatology, Tel Aviv Sourasky Medical Center, Tel Aviv, Israel

²⁴Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

²⁵Division of Dermatology, Department of Medicine, National University Hospital, Singapore City, Singapore

²⁶Department of Dermatology and Pathology, Yale School of Medicine, New Haven, Connecticut, USA

²⁷Department of Dermatology, Akdeniz University Faculty of Medicine, Antalya, Turkey

²⁸Department of Dermatology, Universitätsklinikum Schleswig-Holstein, Campus Lübeck, Lübeck, Germany

²⁹Section of Pediatric Dermatology, Munich University Center for Children with Medical and Developmental Complexity, Ludwig-Maximilians-University Hospital, Munich, Germany

³⁰2nd Dermatology Department, Aristotle University School of Medicine, AIBD Center of Expertise, Papageorgiou General Hospital, Thessaloniki, Greece

³¹Department of Dermatology, Hospital Infantil Niño Jesús, Madrid, Spain

³²Department of Dermatology, French Center of Competence for Auto Immune Blistering Diseases, Necker-Enfants Malades Hospital, Paris, France

³³Lübecker Institute of Experimental Dermatology, University of Lübeck, Lübeck, Germany

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

Arti Nanda has received support from Sanofi, Novartis and AstraZeneca. Miloš Nikolić has received support from AbbVie, Novartis, Pfizer, Johnson & Johnson, Eli Lilly and Medis. Emma Benton has received support from Argenx, Sanofi and Novartis. Matthias Goebeler has received support from ArgenX. Dimitra Kiritsi has received support from Chiesi Pharma, Rheacell GmbH, Xinnate, ArgenX, AbbVie, Janssen, UCB and Galenica. Margarita Larralde has received support from La Roche Posay, Cassara and Sanofi. Angelo Valerio Marzano has received support from AbbVie, Amgen, Boehringer-Ingelheim, Bristol Myers Squibb, Incyte, Leopharma, Novartis, Pfizer, Sanofi and UCB. Joost Meijer has received support from Argenx and Sanofi. Julie Powell has received support from Arcutis, Incyte, Pierre-Fabre, Sanofi, Sun Pharma and Pfizer. Nisha Suyien Chandran has received support from Menarini, AbbVie, Johnson & Johnson, Sanofi, Pfizer, DKSH, L'Oréal, Novartis, Amgen, Galderma, Boehringer Ingelheim, Janssen, LEO Pharma and Lion Corporation. Mary M. Tomayko has received support from Otsuka Pharmaceuticals and AstraZeneca. Nina van Beek has received support from Fresenius Medical Care and Euroimmun AG. Aikaterini Patsatsi has received support from AbbVie, Amgen, Argenx, Boehringer-Ingelheim, Novartis, Pfizer, Sanofi, Johnson & Johnson, Genesis Pharma, Pharmaserv Lilly and UCB. Anne Welfringer-Morin has received support from Sanofi. Enno Schmidt has received support from Fresenius Medical Care, ArgenX, Sanofi, Euroimmun, Chugai, Almirall, LoopLab Bio and AbbVie. Pascal Joly has received support from ArgenX, Amgen, AstraZeneca, ThermoFisher, Sanofi, Akari, Janssen, Novartis, Servier, Chugai, Kezar Life Science, Regeneron, UCB, Principabio, Zenyaku Kogyo, Dualyx, JJP Biologics, Lilly, Leo and AbbVie. All other authors declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

Data regarding the voting process and outcome can be made available in a pseudonymized form on due request from B.T. through E.S.

ETHICAL APPROVAL

The guideline has followed the guidelines for S2K standards of the Association of the Scientific Medical Societies in Germany (AWMF). No formal ethical approval is required.

ORCID

A. Nanda  <https://orcid.org/0000-0002-1223-3181>
 M. Caproni  <https://orcid.org/0000-0002-7471-3471>
 K. Drenovska  <https://orcid.org/0000-0002-8254-1630>
 A. V. Marzano  <https://orcid.org/0000-0002-8160-4169>
 J. Meijer  <https://orcid.org/0000-0001-7654-3528>
 D. F. Murrell  <https://orcid.org/0000-0003-2971-0199>
 E. Sprecher  <https://orcid.org/0000-0002-4093-1032>
 N. van Beek  <https://orcid.org/0000-0002-3265-0011>
 A. Patsatsi  <https://orcid.org/0000-0001-9616-2001>
 A. Welfringer-Morin  <https://orcid.org/0000-0001-7479-3689>
 E. Schmidt  <https://orcid.org/0000-0002-1206-8913>
 P. Joly  <https://orcid.org/0000-0002-5734-0255>

REFERENCES

- Schmidt E, Kasperkiewicz M, Joly P. Pemphigus. *Lancet*. 2019;394:882–94.
- Kasperkiewicz M, Ellebrecht CT, Takahashi H, et al. Pemphigus. *Nat Rev Dis Primers*. 2017;3:17026.
- Schmidt E, Zillikens D. Pemphigoid diseases. *Lancet*. 2013;20:30.
- Mahé A, Flageul B, Bobin P. Bullous IgA linear dermatosis of children in Mali. *Ann Dermatol Venereol*. 1996;123:544–8.
- Weston WL, Morelli JG, Huff JC. Misdiagnosis, treatments, and outcomes in the immunobullous diseases in children. *Pediatr Dermatol*. 1997;14:264–72.
- Singalavanija S, Limpongsanurak W. Immunobullous diseases in Thai children: report of 24 cases. *J Med Assoc Thai*. 2003;86(3):S681–S688.
- Kharfi M, Khaled A, Karaa A, Zarea I, Fazaa B, Kamoun MR. Linear IgA bullous dermatosis: the more frequent bullous dermatosis of children. *Dermatol Online J*. 2010;16(2):25.
- Kong YL, Lim YL, Chandran NS. Retrospective study on autoimmune blistering disease in Paediatric patients. *Pediatr Dermatol*. 2015;32:845–52.
- Salman A, Tekin B, Yucelten D. Autoimmune bullous disease in childhood. *Indian J Dermatol*. 2017;62:440.
- Welfringer-Morin A, Bekel L, Bellon N, Gantzer A, Boccara O, Hadj-Rabia S, et al. Long-term evolving profile of childhood autoimmune blistering diseases: retrospective study on 38 children. *J Eur Acad Dermatol Venereol*. 2019;33:1158–63.
- Nanda A, Lazarevic V, Rajy JM, Almasry IM, AlSabah H, AlLafi A. Spectrum of autoimmune bullous diseases among children in Kuwait. *Pediatr Dermatol*. 2021;38:50–7.
- Mahajan R, Handa S, Kumar S, Chatterji D, Saikia UN, De D. Pediatric autoimmune blistering disorders – a five-year demographic profile and therapy experience. *Int J Dermatol*. 2022;61:1511–8.
- van Beek N, Schmidt E. Autoimmune Bullous Diseases. *Harper's textbook of pediatric dermatology*. Hoboken, NJ: John Wiley & Sons, Ltd; 2019. p. 868–97. <https://doi.org/10.1002/9781119142812.ch74>
- Hübner F, König IR, Holtsche MM, Zillikens D, Linder R, Schmidt E. Prevalence and age distribution of pemphigus and pemphigoid diseases among paediatric patients in Germany. *J Eur Acad Dermatol Venereol*. 2020;34:2600–5.
- Nanda A, Dvorak R, Al-Sabah H, Alsaleh QA. Linear IgA bullous disease of childhood: an experience from Kuwait. *Pediatr Dermatol*. 2006;23:443–7.
- Lewis-Jones MS, Finlay AY. The Children's dermatology life quality index (CDLQI): initial validation and practical use. *Br J Dermatol*. 1995;132:942–9.
- Holme SA, Man I, Sharpe JL, Dykes PJ, Lewis-Jones MS, Finlay AY. The Children's dermatology life quality index: validation of the cartoon version. *Br J Dermatol*. 2003;148:285–90.
- Joly P, Horvath B, Patsatsi A, Uzun S, Bech R, Beissert S, et al. Updated S2K guidelines on the management of pemphigus vulgaris and foliaceus initiated by the European Academy of Dermatology and Venereology (EADV). *J Eur Acad Dermatol Venereol*. 2020;34:1900–13.
- Borradori L, Van Beek N, Feliciani C, Tedbirt B, Antiga E, Bergman R, et al. Updated S2 K guidelines for the management of bullous pemphigoid initiated by the European Academy of Dermatology and Venereology (EADV). *J Eur Acad Dermatol Venereol*. 2022;36:1689–704.
- Schmidt E, Rashid H, Marzano AV, Lamberts A, di Zenzo G, Diercks GFH, et al. European guidelines (S3) on diagnosis and management of mucous membrane pemphigoid, initiated by the European Academy of Dermatology and Venereology – part II. *J Eur Acad Dermatol Venereol*. 2021;35:1926–48.
- Rashid H, Lamberts A, Borradori L, Alberti-Violetti S, Barry RJ, Horváth B. European guidelines (S3) on diagnosis and management of mucous membrane pemphigoid, initiated by the European Academy of Dermatology and Venereology – part I. *J Eur Acad Dermatol Venereol*. 2021;35:1750–64.
- Antiga E, Bech R, Maglie R, Genovese G, Borradori L, Bockle B, et al. S2k guidelines on the management of paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome initiated by the European Academy of Dermatology and Venereology (EADV). *J Eur Acad Dermatol Venereol*. 2023;37:1118–34.
- Prost-Squarcioni C, Caux F, Schmidt E, Jonkman MF, Vassileva S, Kim SC, et al. International bullous diseases group: consensus on diagnostic criteria for epidermolysis bullosa acquisita. *Br J Dermatol*. 2018;179:30–41.
- Witte M, Zillikens D, Schmidt E. Diagnosis of autoimmune blistering diseases. *Front Med Lausanne*. 2018;5:296.
- Mee JB. Diagnostic techniques in autoimmune blistering diseases. *Br J Biomed Sci*. 2023;80:11809.
- Antiga E, Verdelli A, Calabrò A, Fabbri P, Caproni M. Clinical and immunopathological features of 159 patients with dermatitis herpetiformis: an Italian experience. *G Ital Dermatol Venereol*. 2013;148:163–9.
- Vodgel RM, Jonkman MF, Pas HH, de Jong MCJM. U-serrated immunodeposition pattern differentiates type VII collagen targeting bullous diseases from other subepidermal bullous autoimmune diseases. *Br J Dermatol*. 2004;151:112–8.
- Meijer JM, Atefi I, Diercks GFH, Vorobyev A, Zuiderveen J, Meijer HJ, et al. Serration pattern analysis for differentiating epidermolysis bullosa acquisita from other pemphigoid diseases. *J Am Acad Dermatol*. 2018;78:754–9.
- Holtsche MM, van Beek N, Künstner A, Busch H, Zillikens D, Schmidt E. Diagnostic value and practicability of serration pattern analysis by direct immunofluorescence microscopy in pemphigoid diseases. *Acta Derm Venereol*. 2021;101:adv00410.
- Zhou S, Ferguson DJ, Allen J, Wojnarowska F. The location of binding sites of pemphigus vulgaris and pemphigus foliaceus autoantibodies: a post-embedding immunoelectron microscopic study. *Br J Dermatol*. 1997;136:878–83.
- Kitayama S, Makino T, Hayashi M, Mizawa M, Ishii N, Hashimoto T, et al. Usefulness of immunofluorescence overlay antigen mapping in the identification of autoantigen in anti-p200 pemphigoid. *J Dermatol*. 2023;50:1194–8.
- Amber KT, Murrell DF, Schmidt E, Joly P, Borradori L. Autoimmune subepidermal bullous diseases of the skin and mucosae: clinical features, diagnosis, and management. *Clin Rev Allergy Immunol*. 2018;54:26–51.
- Patsatsi A, Meltzanidou P, Katafigiotis S, Sotiriadis D, Schmidt E, Lambropoulos A. Chronic bullous disease of childhood with IgG reactivity to p200 antigen. *Int J Dermatol*. 2017;56:773–5.
- Torres Saavedra FA, Campo LR, Mendez MV, Barreneche NM, Suaza GAV, Restrepo JDR, et al. Bullous lupus as the first manifestation of

- systemic lupus erythematosus in the pediatric population: a diagnostic challenge in daily practice. *Lupus*. 2020;29:1937–42.
35. Panombualert S, Techasatian L, Uppala R, Ungareewittaya P, Choonhakarn C. A rare manifestation of bullous systemic lupus erythematosus in children: a 10-year retrospective study in a tertiary care hospital. *Autoimmune Dis*. 2022;2022:8745.
 36. Caux F, Patsatsi A, Karakioulaki M, Antiga E, Baselga E, Borradori L, et al. S2k guidelines on diagnosis and treatment of linear IgA dermatosis initiated by the European Academy of Dermatology and Venereology. *J Eur Acad Dermatol Venereol*. 2024;38:1006–23.
 37. Prystowsky S, Gilliam JN. Benign chronic bullous dermatosis of childhood. Linear IgA and C3 deposition on the basement membrane. *Arch Dermatol*. 1976;112:837–8.
 38. Wojnarowska F, Marsden RA, Bhogal B, Black MM. Chronic bullous disease of childhood, childhood cicatricial pemphigoid, and linear IgA disease of adults. A comparative study demonstrating clinical and immunopathologic overlap. *J Am Acad Dermatol*. 1988;19(1):792–805.
 39. Aboobaker J, Wojnarowska FT, Bhogal B, Black MM. Chronic bullous dermatosis of childhood – clinical and immunological features seen in African patients. *Clin Exp Dermatol*. 1991;16:160–4.
 40. Kanwar AJ, Sandhu K, Handa S. Chronic bullous dermatosis of childhood in north India. *Pediatr Dermatol*. 2004;21:610–2.
 41. Horiguchi Y, Ikoma A, Sakai R, Masatsugu A, Ohta M, Hashimoto T. Linear IgA dermatosis: report of an infantile case and analysis of 213 cases in Japan. *J Dermatol*. 2008;35:737–43.
 42. Kenani N, Mebazaa A, Denguezli M, Ghariani N, Sriha B, Belajouza C, et al. Childhood linear IgA bullous dermatosis in Tunisia. *Pediatr Dermatol*. 2009;26:28–33.
 43. Monia K, Aida K, Amel K, Ines Z, Becima F, Ridha KM. Linear IgA bullous dermatosis in tunisian children: 31 cases. *Indian J Dermatol*. 2011;56:153–9.
 44. Díaz MS, Morita L, Ferrari B, et al. Linear IgA bullous dermatosis: a series of 17 cases. *Actas Dermosifiliogr*. 2019;110:673–80.
 45. Genovese G, Venegoni L, Fanoni D, Muratori S, Berti E, Marzano AV. Linear IgA bullous dermatosis in adults and children: a clinical and immunopathological study of 38 patients. *Orphanet J Rare Dis*. 2019;14:115.
 46. Wang KL, Lehman JS, Davis DMR. Linear IgA bullous dermatosis of childhood: retrospective single-center cohort. *Pediatr Dermatol*. 2024;41:461–4.
 47. Ratnam KV, Lee CT, Tan T. Chronic bullous dermatosis of childhood in Singapore. *Int J Dermatol*. 1986;25:34–7.
 48. Edwards S, Wojnarowska F. Chronic bullous disease of childhood in three patients of Polynesian extraction. *Clin Exp Dermatol*. 1990;15:367–9.
 49. Ajithkumar K, Kurian S, Jacob M, Pulimood S. Linear IgA bullous dermatosis in south India. *Int J Dermatol*. 1997;36:191–3.
 50. Lings K, Bygum A. Linear IgA bullous dermatosis: a retrospective study of 23 patients in Denmark. *Acta Derm Venereol*. 2015;95:466–71.
 51. Dauendorffer JN, Mahé E, Saiag P. Tacrolimus ointment, an interesting adjunctive therapy for childhood linear IgA bullous dermatosis. *J Eur Acad Dermatol Venereol*. 2008;22:364–5.
 52. Mervic L, Dragos V, Pavlović MD. Linear IgA bullous dermatosis of childhood: successful treatment with miocamycin and topical corticosteroid. *Clin Exp Dermatol*. 2009;34:e391–e392.
 53. de Souza BC, Fregonesi NCFP, Tebcherani AJ, Sanchez APG, Aoki V, Fernandes JC. Linear IgA bullous dermatosis: report of an exuberant case. *An Bras Dermatol*. 2013;88(6):67–70.
 54. de las Heras MN. Linear IgA bullous dermatosis of childhood: good response to antibiotic treatment. *Clin Exp Dermatol*. 2014;39:395–7.
 55. González-Martín L, González A, Pérez-Martín ME. Linear immunoglobulin a bullous dermatosis: two cases report. *Arch Argent Pediatr*. 2015;113:e279–e282.
 56. Cui YX, Yang BQ, Zhou GZ, Zhang FR. Childhood linear IgA bullous dermatosis successfully treated with oral nicotinamide. *Clin Exp Dermatol*. 2016;41:816–8.
 57. Fernandes K d AP, Galvis KH, Gomes AC d MS, Nogueira OM, Felix PAO, Vargas TJ d S. Linear IgA and IgG bullous dermatosis. *An Bras Dermatol*. 2016;91(5):32–4.
 58. Tate C, Christian W, Newell L. Chronic bullous dermatosis of childhood and the string of pearls sign. *J Pediatr*. 2018;202:325–85.
 59. Vaz SO, Franco C, Santos P, Amaral R. Skin and coeliac disease, a lot to think about: a case series. *BMJ Case Rep*. 2018;2018:bcr2017222797.
 60. Agarwal US, Randhawa NK, Singh A, Agarwal P. Treatment of a refractory case of chronic bullous disease of childhood in a five year old child with rituximab. *Indian Dermatol Online J*. 2021;12:914. https://doi.org/10.4103/idoj.idoj_914_20
 61. Mitra D, Bhatnagar A, Singh GK, Sandhu S. Successful treatment of refractory chronic bullous disease of childhood with rituximab. *Indian Dermatol Online J*. 2022;13:248–51.
 62. Nasri J, Jungo P, Blickenstorfer M, Mühleisen B, Navarini AA, Juratli HA, et al. Linear IgA bullous dermatosis of childhood. *Dermatol Online J*. 2022;28:818.
 63. Simon JC, Dietrich A, Kapp A, Schöpf E. Chronic bullous dermatosis in childhood. Association with salmonella enteritis. *Hautarzt*. 1995;46:485–9.
 64. Baldari U, Raccagni AA, Celli B, Righini MG. Chronic bullous disease of childhood following Epstein-Barr virus seroconversion: a case report. *Clin Exp Dermatol*. 1996;21:123–6.
 65. Sillevs Smitt JH, Leusen JH, Stas HG, Teeuw AH, Weening RS. Chronic bullous disease of childhood and a paecilomyces lung infection in chronic granulomatous disease. *Arch Dis Child*. 1997;77:150–2.
 66. Ho JCC, Ng PLP, Tan SH, Giam YC. Childhood linear IgA bullous disease triggered by amoxicillin-clavulanic acid. *Pediatr Dermatol*. 2007;24:E40–E43.
 67. Polat M, Lenk N, Kürekçi E, Öztaş P, Artüz F, Alli N. Chronic bullous disease of childhood in a patient with acute lymphoblastic leukemia: possible induction by a drug. *Am J Clin Dermatol*. 2007;8:389–91.
 68. Nantel-Battista M, Al Dhaybi R, Hatami A, Marcoux D, Desroches A, Kokta V. Childhood linear IgA bullous disease induced by trimethoprim-sulfamethoxazole. *J Dermatol Case Rep*. 2010;4:33–5.
 69. Gatto A, Guerriero C, Moretta G, Valentini P, Giorgio V, Lazzareschi I, et al. Linear IgA bullous dermatosis in a two-year-old child: possible association with aspirin. *Eur J Dermatol*. 2017;27:417–8.
 70. Sarikaya Solak S, Ficicioglu S. Cephalosporin-induced linear IgA dermatosis in a child: case report and literature review. *Dermatol Ther*. 2019;32:e12927.
 71. Stamenkovic HM, Lazarevic D, Stankovic T, Vojinovic J, Lekic B, Marinkovic A, et al. Linear IgA dermatosis of the childhood-report of an amoxicillin-induced case. *Dermatol Ther*. 2020;33:e13173.
 72. Davis S, Altattan M, Abugharbyeh A, Khader Y, Altork N. A string of pearls: linear immunoglobulin a (IgA) bullous dermatosis in the setting of imipramine use and newly diagnosed ulcerative colitis. *Cureus*. 2023;15:e33448.
 73. Ikeya S, Urano S, Tokura Y. Linear IgA bullous dermatosis following human papillomavirus vaccination. *Eur J Dermatol*. 2012;22:787–8.
 74. Corrà A, Bonciolini V, Quintarelli L, Verdelli A, Caproni M. Linear IGA bullous dermatosis potentially triggered by vaccination. *Int J Immunopathol Pharmacol*. 2022;36:21218.
 75. Handley J, Shields M, Dodge J, Walsh M, Bingham A. Chronic bullous disease of childhood and ulcerative colitis. *Pediatr Dermatol*. 1993;10:256–8.
 76. Paige DG, Leonard JN, Wojnarowska F, Fry L. Linear IgA disease and ulcerative colitis. *Br J Dermatol*. 1997;136:779–82.
 77. Nanda A, Dvorak R, Al-Sabah H, Madda JP, Anim JT, Alsaleh QA. Association of linear IgA bullous disease of childhood with Crohn's disease. *Int J Dermatol*. 2006;45:1184–6.
 78. Sandoval M, Farias MM, Gonzalez S. Linear IgA bullous dermatosis: report of five cases in Chile. *Int J Dermatol*. 2012;51:1303–6.
 79. Vargas TJ d S, Fialho M, dos Santos LT, Rodrigues PA d JB, Vargas ALBSJ, Sousa MAJ. Linear IgA dermatosis associated with ulcerative colitis: complete and sustained remission after total colectomy. *An Bras Dermatol*. 2013;88:600–3.

80. Khan M, Park L, Skopit S. Management options for linear immunoglobulin a (IgA) bullous dermatosis: a literature review. *Cureus*. 2023;15:e36481.
81. Shin L, Gardner JT, Dao H. Updates in the diagnosis and Management of Linear IgA disease: a systematic review. *Medicina (Kaunas)*. 2021;57:818.
82. Shan X-F, Zhang F-R, Tian H-Q, Wang N, Zhou S-J, Wang G-J. A case of linear IgA dermatosis successfully treated with tetracycline and niacinamide. *Int J Dermatol*. 2016;55:e216–e217.
83. Brown K, Corin S, Handel AS. Doxycycline for the treatment of Lyme disease in young children. *Pediatr Infect Dis J*. 2023;42:e470–e472.
84. Fortuna G, Marinkovich MP. Linear immunoglobulin a bullous dermatosis. *Clin Dermatol*. 2012;30:38–50.
85. Mori F, Saretta F, Liotti L, et al. Linear immunoglobulin a bullous dermatosis in children. *Front Pediatr*. 2022;10:937528.
86. Marinkovich MP, Taylor TB, Keene DR, Burgeson RE, Zone JJ. LAD-1, the linear IgA bullous dermatosis autoantigen, is a novel 120-kDa anchoring filament protein synthesized by epidermal cells. *J Invest Dermatol*. 1996;106:734–8.
87. Zhu YI, Stiller MJ. Dapsone and sulfones in dermatology: overview and update. *J Am Acad Dermatol*. 2001;45:420–34.
88. Wojnarowska F, Whitehead P, Leigh IM, Bhogal BS, Black MM. Identification of the target antigen in chronic bullous disease of childhood and linear IgA disease of adults. *Br J Dermatol*. 1991;124:157–62.
89. Oranje AP, van Joost T. Pemphigoid in children. *Pediatr Dermatol*. 1989;6:267–74.
90. Schultz B, Hook K. Bullous diseases in children: a review of clinical features and treatment options. *Paediatr Drugs*. 2019;21:345–56.
91. Chimanovitch I, Hamm H, Georgi M, Kroiss M, Stolz W, Apitz C, et al. Bullous pemphigoid of childhood: autoantibodies target the same epitopes within the NC16A domain of BP180 as autoantibodies in bullous pemphigoid of adulthood. *Arch Dermatol*. 2000;136:527–32.
92. Erbagci Z. Childhood bullous pemphigoid following hepatitis B immunization. *J Dermatol*. 2002;29:781–5.
93. Fisler RE, Saeb M, Liang MG, Howard RM, McKee PH. Childhood bullous pemphigoid: a clinicopathologic study and review of the literature. *Am J Dermatopathol*. 2003;25:183–9.
94. Kuenzli S, Grimaître M, Krischer J, Saurat J-H, Calza A-M, Borradori L. Childhood bullous pemphigoid: report of a case with life-threatening course during homeopathy treatment. *Pediatr Dermatol*. 2004;21:160–3.
95. Motegi S-I, Abe M, Tamura A, Ishii N, Hashimoto T, Ishikawa O. Childhood bullous pemphigoid successfully treated with diaminodiphenyl sulfone. *J Dermatol*. 2005;32:809–12.
96. Martínez-De Pablo MI, González-Enseñat MA, Vicente A, Gilaberte M, Mascaró JM. Childhood bullous pemphigoid: clinical and immunological findings in a series of 4 cases. *Arch Dermatol*. 2007;143:215–20.
97. Sáenz AM, González F, Cirocco A, Tacaronte IM, Fajardo JE, Calebotta A. Childhood bullous pemphigoid: a case report and 10-year follow up. *Int J Dermatol*. 2007;46:508–10.
98. Nanda A, Nanda M, Dvorak R, Al-Sabah H, Alsaleh QA. Bullous pemphigoid (BP) in an infant complicated by tuberculous meningoencephalitis. *Int J Dermatol*. 2007;46:964–6.
99. Xiao T, Li B, Wang Y-K, He C-D, Chen H-D. Childhood bullous pemphigoid treated by i.v. immunoglobulin. *J Dermatol*. 2007;34:650–3.
100. Sugawara N, Nagai Y, Matsushima Y, Aoyama K, Ishikawa O. Infantile bullous pemphigoid treated with intravenous immunoglobulin therapy. *J Am Acad Dermatol*. 2007;57:1084–9.
101. Erbagci Z. Childhood bullous pemphigoid in association with hyperimmunoglobulin E syndrome. *Pediatr Dermatol*. 2008;25:28–33.
102. Waisbourd-Zinman O, Ben-Amitai D, Cohen A. Bullous pemphigoid in infancy: clinical and epidemiologic characteristics. *J Am*. 2008;58:41–8.
103. Purvis DJ, Bhogal BS, Harper JJ. Bullous pemphigoid in an infant using complementary medicine. *Clin Exp Dermatol*. 2009;34:195–8.
104. Gajic-Veljcic M, Nikolic M, Medenica L. Juvenile bullous pemphigoid: the presentation and follow-up of six cases. *J Eur Acad Dermatol Venereol*. 2010;24:69–72.
105. Atzori L, Pau M, Podda R, Manioli C, Aste N. A case of bullous pemphigoid in infancy treated with local corticosteroids. *G Ital Dermatol Venereol*. 2011;146:493–6.
106. Fuertes I, Luelmo J, Leal L, Román J. Refractory childhood pemphigoid successfully treated with rituximab. *Pediatrics*. 2013;30:e96.
107. de la Fuente S, Hernández-Martín Á, de Lucas R, González-Enseñat MA, Vicente A, Colmenero I, et al. Postvaccination bullous pemphigoid in infancy: report of three new cases and literature review. *Pediatr Dermatol*. 2013;30:741–4.
108. Schwieger-Briel A, Moellmann C, Mattulat B. Bullous pemphigoid in infants: characteristics, diagnosis and treatment. *Orphanet J Rare*. 2014;9:185.
109. Ister M, Pouessel G, Ythier H, Catteau B, Carpentier O. Postvaccinal, corticosteroid-resistant bullous pemphigoid in infancy: treatment with intravenous immunoglobulin. *Pediatr Dermatol*. 2014;31:e94–e95.
110. Kato K, Koike K, Kobayashi C, Iijima S, Hashimoto T, Tsuchida M. Bullous pemphigoid after allogeneic hematopoietic stem cell transplantation. *Pediatr Int*. 2015;57:480–3.
111. Seminario-Vidal L, Sami N, Miller J, Theos A. Mycophenolate mofetil therapy for pediatric bullous pemphigoid. *Dermatol Online J*. 2015;21:13030.
112. Taquin H, Chiaverini C, Lacour JP. Spectrum of clinical responses to therapies in infantile bullous pemphigoid. *Pediatr Dermatol*. 2016;33:e77–e81.
113. Ayala-Cortés AS, Martínez-Cabriales SA, Welsh O, Rosales-Solis G, Vázquez-Martínez O, Ocampo-Candiani J. Childhood bullous pemphigoid: a treatment challenge. *J Pediatr*. 2016;179:271–5.
114. Ramos L, Tellechea O, Moreno A, Martins L, Neves N, Januário G, et al. Childhood bullous pemphigoid: report of 2 cases. *J Eur Acad Dermatol Venereol*. 2016;30:330–2.
115. Baroero L, Coppo P, Bertolino L, Maccario S, Savino F. Three case reports of post immunization and post viral bullous pemphigoid: looking for the right trigger. *BMC Pediatr*. 2017;17:60.
116. Watanabe T, Hara S, Muto J, Watanabe D, Akiyama M. Infantile bullous pemphigoid successfully treated with intravenous immunoglobulin therapy. *Clin Exp Dermatol*. 2017;42:576–8.
117. Ferreira BR, Vaz AS, Ramos L, Reis JP, Gonçalves M. Bullous pemphigoid of infancy - report and review of infantile and pediatric bullous pemphigoid. *Dermatol Online J*. 2017;23:13030.
118. Wawrzycki B, Krasowska D, Pietrzak A, Szumilo J, Blazowski L, Pietraszek-Mamcarz J, et al. Infantile bullous pemphigoid successfully treated with combined pulse corticosteroids and high-dose IVIG. *Dermatol Ther*. 2018;31:e12635.
119. Patsatsi A, Kyriakou A, Werth VP. Bullous pemphigoid in adolescence. *Pediatr Dermatol*. 2019;36:184–8.
120. Thomsen K, Deleuran M, Vestergaard C, Holm M, Riber-Hansen R, Bech R. Severe infantile bullous pemphigoid treated with dapsone after bridging with systemic glucocorticoid. *Case Rep Dermatol*. 2019;11:187–93.
121. Okada K, Kakeda M, Yamamoto S, Yokoyama T, Habe K, Nakato D, et al. Infantile bullous pemphigoid successfully treated with i.v. immunoglobulin and cyclosporin. *J Dermatol*. 2019;46:e213–e214.
122. Wang Y, Xiao Y, Li Y, Fu L, Yu F, Ma L, et al. Bullous pemphigoid associated with ulcerative colitis in a young child: a case report. *Dermatol Ther*. 2020;33:e14325.
123. Xu K, Wei Y, Wang T, Ding L, Li W, Wang L. A case of infantile bullous pemphigoid successfully treated with compound glycyrrhizin tablets. *Dermatol Ther*. 2020;33:e13550.
124. Folsom B, Raisanen T, Eshaq M. Two infants with blistering rashes originating on acral sites as a presenting sign of infantile bullous pemphigoid. *JAAD Case Rep*. 2020;6:473–5.
125. Pankakoski A, Panelius J, Salava A, Kluger N. Bullous pemphigoid in three Paediatric patients in Finland. *Acta Derm Venereol*. 2020;100:adv00086.
126. Navarro-Navarro I, Jiménez-Gallo D, Valenzuela-Ubiña S, Domínguez-Gómez M, Linares-Barrios M. Infantile bullous pemphigoid following serogroup B meningococcal vaccination. *Br J Dermatol*. 2021;184:e53.

127. Rosińska-Więckowicz A, Jałowska M, Bowszyc-Dmochowska M, Dmochowski M. Case report: infantile bullous pemphigoid: triggering by COVID-19 is speculative. *Front Med Lausanne*. 2021;8:760823.
128. Andersen SM, Fage S, Rubak SL, Holm M, Jensen JMB, Mogensen T, et al. Immune dysregulation, Polyendocrinopathy, enteropathy, X-linked syndrome manifesting as lymphocytic interstitial pneumonia and treatment-resistant bullous pemphigoid. *Pediatr Allergy Immunol Pulmonol*. 2021;34:76–9.
129. Bar-Ilan E, Gál AI, Veres K, Solti A, Ferge PÁ, Dimény T, et al. Recalcitrant infantile bullous pemphigoid successfully treated with cyclosporine. *JAAD Case Rep*. 2022;29:116–20.
130. Kianfar N, Dasdar S, Mahmoudi H, Tavakolpour S, Balighi K, Daneshpazhooh M. Rituximab in childhood and juvenile autoimmune bullous diseases as first-line and second-line treatment: a case series of 13 patients. *J Dermatolog Treat*. 2020;2:1–6.
131. Nikyar Z, Hatami P, Aryanian Z, Sotoudeh S, Ziaee V, Goodarzi A. An unusual TEN-like presentation of juvenile bullous pemphigoid: a diagnostic challenge. *Case Rep Dermatol Med*. 2022;2022:8507156.
132. Fallahi M, Jamee M, Enayat J, et al. Novel PGM3 mutation in two siblings with combined immunodeficiency and childhood bullous pemphigoid: a case report and review of the literature. *Allergy Asthma Clin Immunol*. 2022;18:111.
133. Akbarialiabad H, Schmidt E, Patsatsi A, et al. Bullous pemphigoid. *Nat Rev Dis Primers*. 2025;11:12.
134. Zhou W, Tan Y, Chen X, Zhang W, Sun Z, Shen Y, et al. Successful treatment of infantile refractory bullous pemphigoid with baricitinib. *Australas J Dermatol*. 2024;65:642–6.
135. Almassio AB, Luna L, Larralde M. Pediatric bullous pemphigoid treated with dupilumab: a case report with good response. *Pediatr Dermatol*. 2025;42:1248–51.
136. Zhao L, Wang Q, Liang G, Zhou Y, Yiu N, Yang B, et al. Evaluation of dupilumab in patients with bullous pemphigoid. *JAMA Dermatol*. 2023;159:953–60.
137. Planella-Fontanillas N, Bosch-Amate X, Jiménez Antón A, et al. Real-world evaluation of the effectiveness and safety of dupilumab in bullous pemphigoid: an Ambispective multicenter case series. *Br J Dermatol*. 2024;1:ljae403.
138. Avallone G, Maronese CA, Zussino M, Muratori S, Ferrucci SM, Quaglini P, et al. Effectiveness of dupilumab and omalizumab in bullous pemphigoid: a Nationwide retrospective cohort study. *J Dermatol*. 2025;52:983–1000.
139. Press Release: Dupixent approved in the US as the only targeted medicine to treat patients with bullous pemphigoid. <https://www.sanofi.com/en/media-room/press-releases/2025/2025-06-20-05-00-00-3102518>. Accessed 30 Jun 2025.
140. Murrell DF, Joly P, Werth VP, Ujiie H, Worm M, Mangold AR, et al. Study design of a phase 2/3 randomized controlled trial of dupilumab in adults with bullous pemphigoid: LIBERTY-BP ADEPT. *Adv Ther*. 2024;41:2991–3002.
141. Du G, Patzelt S, van Beek N, Schmidt E. Mucous membrane pemphigoid. *Autoimmun Rev*. 2022;21:103036.
142. Jolliffe DS, Sim-Davis D. Cicatricial pemphigoid in a young girl: report of a case. *Clin Exp Dermatol*. 1977;2:281–4.
143. Rogers M, Painter D. Cicatricial pemphigoid in a four-year-old child: a case report. *Australas J Dermatol*. 1981;22:21–3.
144. Rosenbaum MM, Esterly NB, Greenwald MJ, Gerson CR. Cicatricial pemphigoid in a 6-year-old child: report of a case and review of the literature. *Pediatr Dermatol*. 1984;2:13–22.
145. DeCastro P, Jorizzo JL, Rajaraman S, Solomon AR, Briggaman RA, Raimer SS. Localized vulvar pemphigoid in a child. *Pediatr Dermatol*. 1985;2:302–7.
146. Moy W, Kumar V, Friedman RP, Schaeffer ML, Beutner E, Helm F. Cicatricial pemphigoid. A case of onset at age 5. *J Periodontol*. 1986;57:39–43.
147. Iglesias LS, Quismorio FP, McDonnell PJ, Barnes C. Ocular cicatricial pemphigoid in a twelve-year-old boy. *Cornea*. 1992;11:365–7.
148. Roche C, Field EA. Benign mucous membrane pemphigoid presenting as desquamative gingivitis in a 14-year-old child. *Int J Paediatr Dent*. 1997;7:31–4.
149. Farrell A, Kirtschig G, Dalziel K. Childhood vulval pemphigoid: a clinical and immunopathological study of five patients. *Br J Dermatol*. 1999;140:308–12.
150. Cheng YS, Rees TD, Wright JM, Plemons JM. Childhood oral pemphigoid: a case report and review of the literature. *J Oral Pathol Med*. 2001;30:372–7.
151. Musa NJ, Kumar V, Humphreys L, Aguirre A, Neiders ME. Oral pemphigoid masquerading as necrotizing ulcerative gingivitis in a child. *J Periodontol*. 2002;73:657–63.
152. Lebeau S, Mainetti C, Masouyé I, Saurat J-H, Borradori L. Localized childhood vulval pemphigoid treated with tacrolimus ointment. *Dermatology*. 2004;208:273–5.
153. Schoeffler A, Roth B, Causeret A, Kanitakis J, Faure M, Claudy A. Vulvar cicatricial pemphigoid of childhood. *Pediatr Dermatol*. 2004;21:51–3.
154. Hoque SR, Patel M, Farrell AM. Childhood cicatricial pemphigoid confined to the vulva. *Clin Exp Dermatol*. 2006;31:63–4.
155. Lourenço SV, Boggio P, Agner Machado Martins LE, Santi CG, Aoki V, Menta Simonsen Nico M. Childhood oral mucous membrane pemphigoid presenting as desquamative gingivitis in a 4-year-old girl. *Acta Derm Venereol*. 2006;86:351–4.
156. Kanwar AJ, Ajith C, Narang T. Cicatricial pemphigoid associated with coeliac disease in a child. *J Eur Acad Dermatol Venereol*. 2006;20:1148–50.
157. Iovine A, Fimiani F, Vassallo P, Alessio M, Magli A. Ocular manifestations in a case of childhood cicatricial pemphigoid. *Eur J Ophthalmol*. 2008;18:636–8.
158. Kharfi M, Khaled A, Anane R, Fazaa B, Kamoun MR. Early onset childhood cicatricial pemphigoid: a case report and review of the literature. *Pediatr Dermatol*. 2010;27:119–24.
159. Mostafa MI, Hassib NF, Nemat AH. Oral mucous membrane pemphigoid in a 6-year-old boy: diagnosis, treatment and 4years follow-up. *Int J Paediatr Dent*. 2010;20:76–9.
160. Lavallée A, Bourret-Massicotte D, Laughrea PA. Childhood mucous membrane pemphigoid: a case with exclusively ocular involvement. *Cornea*. 2013;32:1399–401.
161. Kahn E, Spence Shishido A, Yancey KB, Lawley LP. Anti-laminin-332 mucous membrane pemphigoid in a 9-year old girl. *Pediatr Dermatol*. 2014;31:e76–e79.
162. Belzile E, Funaro D, Powell J. Localized vulvar bullous pemphigoid of childhood: a rare cause of persistent vulvar erosions in children. *Pediatr Dermatol*. 2019;36:349–51.
163. Flores-Climente V, Rozas-Muñoz E, Martínez-Grau A, Aucouturier F, Luna S, Baselga E. Childhood ocular mucous membrane pemphigoid successfully treated with rituximab. *Pediatr Dermatol*. 2019;36:984–5.
164. Rodriguez Baisi K, Wentworth A, Chattha AJ, DiCaudo DJ, Mangold A, Nelson SA, et al. A rare case of childhood mucous membrane pemphigoid involving the oral and genital mucosa. *Pediatr Dermatol*. 2021;38:904–7.
165. Ollero R, Soldevila A, Fideliz de la Paz M, Martínez-Grau A. Surgical Management of Ocular Cicatricial Pemphigoid in a pediatric patient. *Cornea*. 2022;41:911–3.
166. Marti-Marti I, Rizo-Potau D, Gómez S, Pallarès H, Iranzo P. Childhood vulvar mucous membrane pemphigoid. *J Pediatr Adolesc Gynecol*. 2022;35:171–3.
167. Rashid H, Oldhoff JM, Esajas M, Diercks GFH, Pas HH, Bolling MC, et al. Juvenile and adult vulvar pemphigoid, an under recognized entity: case series of fourteen patients. *JAAD Case Rep*. 2021;13:75–80.
168. Fageeh HI. Mucous membrane pemphigoid: a case report with Oral and ocular presentation. *J Contemp Dent Pract*. 2022;23:755–9.
169. Taibo A, Colmenero I, Campos M, Torrelo A. Vulvar erosions in a young girl. *Pediatr Dermatol*. 2023;40:940–2.
170. Gart S, Swali R, Lee EB, Harter N. The diagnostic difficulties of juvenile vulvar pemphigoid. *Pediatr Dermatol*. 2024;41:897–9.

171. Jinagal J, Gupta B, Hanumanthu V, Chatterjee D, Limbu S, Malhotra C, et al. Ocular cicatricial pemphigoid in young patients. *Cornea*. 2024;43:1015–21.
172. Chène L, Le Roux-Villet C, Martin L, et al. Characterization of mucous membrane pemphigoid in childhood: a multicentre study of 12 cases. *J Eur Acad Dermatol Venereol*. 2025;39:e999.
173. Egan C, Lazarova Z, Darling T, Yee C, Yancey K. Anti-epiligrin cicatricial pemphigoid: clinical findings, immunopathogenesis, and significant associations. *Medicine*. 2003;82:177–86.
174. Goletz S, Probst C, Komorowski L, Schlumberger W, Fechner K, van Beek N, et al. A sensitive and specific assay for the serological diagnosis of antilaminin 332 mucous membrane pemphigoid. *Br J Dermatol*. 2019;180:149–56.
175. Patzelt S, Schmidt E. Autoimmunity against laminin 332. *Front Immunol*. 2023;14:1250115.
176. van Beek N, Kridin K, Bühler E, Kochan AS, Ständer S, Ludwig RJ, et al. Evaluation of site- and autoantigen-specific characteristics of mucous membrane pemphigoid. *JAMA Dermatol*. 2022;158:84–9.
177. Koga H, Prost-Squarcioni C, Iwata H, Jonkman MF, Ludwig RJ, Bieber K. Epidermolysis bullosa Acquisita: the 2019 update. *Front Med*. 2018;5:362.
178. Tešanović Perković D, Bukvić Mokos Z, Marinović B. Epidermolysis bullosa Acquisita-current and emerging treatments. *J Clin Med*. 2023;12:1139.
179. Edwards S, Wakelin S, Wojnarowska F. Bullous pemphigoid and epidermolysis bullosa acquisita: presentation, prognosis, and immunopathology in 11 children. *Pediatrics*. 1998;15:184.
180. Arpey CJ, Elewski BE, Moritz DK, Gammon WR. Childhood epidermolysis bullosa acquisita. Report of three cases and review of literature. *J Am Acad Dermatol*. 1991;24(5):706–14.
181. Kawachi Y, Ikegami M, Hashimoto T, Matsumura K, Tanaka T, Otsuka F. Autoantibodies to bullous pemphigoid and epidermolysis bullosa acquisita antigens in an infant. *Br J Dermatol*. 1996;135:443–7.
182. Schmidt E, Höpfner B, Chen M, Kuhn C, Weber L, Bröcker EB, et al. Childhood epidermolysis bullosa acquisita: a novel variant with reactivity to all three structural domains of type VII collagen. *Br J Dermatol*. 2002;147:592–7.
183. Trigo-Guzmán FX, Conti A, Aoki V, Maruta CW, Santi CG, Resende Silva CM, et al. Epidermolysis bullosa acquisita in childhood. *J Dermatol*. 2003;30:226–9.
184. Fukumoto T, Umekawa T, Higuchi M, Hashimoto T, Shumann H, Bruckner-Tuderman L, et al. Childhood epidermolysis bullosa acquisita with autoantibodies against all 3 structural domains of type VII collagen. *J Am Acad Dermatol*. 2004;50:480–2.
185. Mayuzumi M, Akiyama M, Nishie W, Ukae S, Abe M, Sawamura D, et al. Childhood epidermolysis bullosa acquisita with autoantibodies against the noncollagenous 1 and 2 domains of type VII collagen: case report and review of the literature. *Br J Dermatol*. 2006;155:1048–52.
186. Abrams ML, Smidt A, Benjamin L, Chen M, Woodley D, Mancini AJ. Congenital epidermolysis bullosa acquisita: vertical transfer of maternal autoantibody from mother to infant. *Arch Dermatol*. 2011;147:337–41.
187. Lin H-Y, Yanagi T, Akiyama M, Iitani MM, Moriuchi R, Natsuga K, et al. Childhood subepidermal blistering disease with autoantibodies to type VII collagen and laminin-332. *Br J Dermatol*. 2011;164:452–4.
188. McKinley SK, Huang JT, Tan J, Kroshinsky D, Gellis S. A case of recalcitrant epidermolysis bullosa acquisita responsive to rituximab therapy. *Pediatr Dermatol*. 2014;31:241–4.
189. Yang B, Wang C, Wang N, Pan F, Chen S, Zhou G, et al. Childhood epidermolysis bullosa acquisita: report of a Chinese case. *Pediatr Dermatol*. 2012;29:614–7.
190. Baican A, Chiriac G, Torio-Padron N, Sitaru C. Childhood epidermolysis bullosa acquisita associated with severe dental alterations: a case presentation. *J Dermatol*. 2013;40:410–1.
191. Liszewski W, Omland SH, Gniadecki R. The successful use of extracorporeal photopheresis in a 12-year-old patient with refractory epidermolysis bullosa acquisita. *Pediatr Dermatol*. 2015;32:e60–e61.
192. Kasperkiewicz M, Orosz I, Abeck D, Koletzko S, Ruzicka T, Sárdy M. Childhood epidermolysis bullosa acquisita with underlying coeliac disease. *Acta Derm Venereol*. 2015;95:1013–4.
193. Lazić-Mosler E, Jukić IL, Murat-Sušić S, et al. Inflammatory epidermolysis bullosa acquisita in a 4-year-old girl. *J Dermatol*. 2015;42:1098–100.
194. Russo I, Ferrazzi A, Zanetti I, Alaibac M. Epidermolysis bullosa acquisita in a 17-year-old boy with Crohn's disease. *BMJ Case Rep*. 2015;2015:bcr210.
195. Bis S, Maguiness SM, Gellis SE, Schneider LC, Lee PY, Notarangelo LD, et al. Immune dysregulation, polyendocrinopathy, enteropathy, X-linked syndrome associated with neonatal epidermolysis bullosa acquisita. *Pediatr Dermatol*. 2015;32:e74–e77.
196. Guerra L, Pacifico V, Calabresi V, de Luca N, Castiglia D, Angelo C, et al. Childhood epidermolysis bullosa acquisita during squaric acid dibutyl ester immunotherapy for alopecia areata. *Br J Dermatol*. 2017;176:491–4.
197. Dewan AK, Braue J, Danford B, Stack LB, Boyd AS, Fine JD, et al. In utero development of epidermolysis bullosa acquisita. *Pediatr Dermatol*. 2019;36:e46–e47.
198. Manuelyan K, Mateev G, Drenovska K, Shahid M, Vassileva S. Epidermolysis bullosa Acquisita mimicking linear IgA bullous disease in a 5-year-old child. *Acta Dermatovenereol Croat*. 2020;28:109–12.
199. Rentfro K, Arthur M. The development of mucous membrane epidermolysis bullosa acquisita in a pediatric patient. *JAAD Case Rep*. 2021;11:3–5.
200. Hignett E, Sami N. Pediatric epidermolysis bullosa acquisita: a review. *Pediatr Dermatol*. 2021;38:1047–50.
201. Bageta ML, Cella E, Cervini AB, Centeno MDV, Roquel L, Mee JB, et al. Epidermolysis bullosa acquisita: a case series of three paediatric patients. *Clin Exp Dermatol*. 2022;47:1346–9.
202. Rivas-Calderón MK, Cheirif-Wolosky O, Ávalos-Díaz E, Durán-McKinster C, Sáez-de-Ocariz M, Rurtusuástegui-García MV, et al. Epidermolysis bullosa Acquisita: a rare autoimmune blistering disease in children. *Skinmed*. 2022;20:460–2.
203. Patel A, Wanat K, Lalor L. An atypical case of pediatric epidermolysis bullosa acquisita: review of diagnosis and pitfalls. *JAAD Case Rep*. 2021;9:81–5.
204. Iwata H, Vorobyev A, Koga H, Recke A, Zillikens D, Prost-Squarcioni C, et al. Meta-analysis of the clinical and immunopathological characteristics and treatment outcomes in epidermolysis bullosa acquisita patients. *Orphanet J Rare Dis*. 2018;13:153.
205. Bolotin D, Petronic-Rosic V. Dermatitis herpetiformis. Part I. Epidemiology, pathogenesis, and clinical presentation. *J Am Acad Dermatol*. 2011;64:1017–24.
206. Görög A, Antiga E, Caproni M, Cianchini G, de D, Dmochowski M, et al. S2k guidelines (consensus statement) for diagnosis and therapy of dermatitis herpetiformis initiated by the European Academy of Dermatology and Venereology (EADV). *J Eur Acad Dermatol Venereol*. 2021;35:1251–77.
207. Reunala T, Hervonen K, Salmi T. Dermatitis herpetiformis: an update on diagnosis and management. *Am J Clin Dermatol*. 2021;22:329–38.
208. Nguyen CN, Kim S-J. Dermatitis herpetiformis: an update on diagnosis, disease monitoring, and management. *Medicina (Kaunas)*. 2021;57:843.
209. Salmi TT, Hervonen K, Kautiainen H, Collin P, Reunala T. Prevalence and incidence of dermatitis herpetiformis: a 40-year prospective study from Finland. *Br J Dermatol*. 2011;165:354–9.
210. Hervonen K, Salmi TT, Kurppa K, Kaukinen K, Collin P, Reunala T. Dermatitis herpetiformis in children: a long-term follow-up study. *Br J Dermatol*. 2014;171:1242–3.
211. Ermacora E, Prampolini L, Tribbia G, Pezzoli G, Gelmetti C, Cucchi G, et al. Long-term follow-up of dermatitis herpetiformis in children. *J Am Acad Dermatol*. 1986;15:24–30.
212. Reunala T, Kosnai I, Karpati S, Kuitunen P, Török E, Savilahti E. Dermatitis herpetiformis: jejunal findings and skin response to gluten free diet. *Arch Dis Child*. 1984;59:517–22.

213. Kósnai I, Kárpai S, Savilahti E, Verkasalo M, Bucsky P, Török E. Gluten challenge in children with dermatitis herpetiformis: a clinical, morphological and immunohistological study. *Gut*. 1986;27:1464–70.
214. Warner J, Brooks SE, James WP, Louisy S. Juvenile dermatitis herpetiformis in Jamaica: clinical and gastrointestinal features. *Br J Dermatol*. 1972;86:226–37.
215. Ganpule M. Juvenile dermatitis herpetiformis. Case reports of six children from Birmingham. *Br J Dermatol*. 1967;79:221–8.
216. Karpai S, Torok E, Kosnai I. Discrete palmar and plantar symptoms in children with dermatitis herpetiformis. *Duhring. Cutis*. 1986;37:184–7.
217. McGovern TW, Bennion SD. Palmar purpura: an atypical presentation of childhood dermatitis herpetiformis. *Pediatr Dermatol*. 1994;11:319–22.
218. Dodi I, Iughetti L, de' Angelis GL, et al. Dermatitis herpetiformis and latent celiac disease in two siblings. *Pediatr Med Chir*. 1995;17:165–6.
219. Démoulins-Giacco N, Gagey V, Teillac-Hamel D, Fraitag S, Caillat-Zucman S, Schmitz J, et al. Dermatitis herpetiformis occurring in patients with celiac disease in childhood. *Arch Pediatr*. 1996;3:541–8.
220. Woollons A, Darley CR, Bhogal BS, Black MM, Atherton DJ. Childhood dermatitis herpetiformis: an unusual presentation. *Clin Exp Dermatol*. 1999;24:283–5.
221. Medica I, Zmak M, Persic M. Dermatitis herpetiformis in a 30-month-old child. *Minerva Pediatr*. 2003;55:171–3.
222. Powell GR, Bruckner AL, Weston WL. Dermatitis herpetiformis presenting as chronic urticaria. *Pediatr Dermatol*. 2004;21:564–7.
223. Lemberg D, Day AS, Bohane T. Coeliac disease presenting as dermatitis herpetiformis in infancy. *J Paediatr Child Health*. 2005;41:294–6.
224. Helsing P, Frøen H. Dermatitis herpetiformis presenting as ataxia in a child. *Acta Derm Venereol*. 2007;87:163–5.
225. Templet JT, Welsh JP, Cusack CA. Childhood dermatitis herpetiformis: a case report and review of the literature. *Cutis*. 2007;80:473–6.
226. Heinlin J, Knoppke B, Kohl E, Landthaler M, Karrer S. Dermatitis herpetiformis presenting as digital petechiae. *Pediatr Dermatol*. 2012;29:209–12.
227. Costin A, António AM, Furtado C, Bártolo E. Dermatitis herpetiformis in an adolescent patient. *An Bras Dermatol*. 2019;94:495–6.
228. Singh A, Ganguly S, Chhabra N, Singh V. Rare co-existent dermatitis herpetiformis and psoriasis in a child: a causal relationship? *Cureus*. 2022;14:e29218.
229. Mohite RS, Furtado S, Kaur T, Mn V, Bhattad S. Dermatitis herpetiformis in a child with Wiskott-Aldrich syndrome: a rare occurrence. *Pediatr Blood Cancer*. 2022;69:e29459.
230. Nanda A, Dvorak R, Al-Saeed K, Al-Sabah H, Alsaleh QA. Spectrum of autoimmune bullous diseases in Kuwait. *Int J Dermatol*. 2004;43:876–81.
231. Husby S, Koletzko S, Korponay-Szabó I, Kurppa K, Mearin ML, Ribes-Koninckx C, et al. European society Paediatric gastroenterology, hepatology and nutrition guidelines for diagnosing coeliac disease 2020. *J Pediatr Gastroenterol Nutr*. 2020;70:141–56.
232. Mabrouk D, Ahmed AR. Analysis of current therapy and clinical outcome in childhood pemphigus vulgaris. *Pediatr Dermatol*. 2011;28:485–93.
233. Asarch A, Gürcan HM, Ahmed AR. A current review of juvenile pemphigus vulgaris: analysis of data on clinical outcomes. *Am J Clin Dermatol*. 2010;11:21–33.
234. Bjarnason B, Flosadóttir E. Childhood, neonatal, and stillborn pemphigus vulgaris. *Int J Dermatol*. 1999;38:680–8.
235. Welfringer-Morin A. Les dermatoses bulleuses auto-immunes de l'enfant. *Ann Dermatol Vénéréol*. 2021;1:151–7.
236. Diaz LA, Sampaio SA, Rivitti EA, Martins CR, Cunha PR, Lombardi C, et al. Endemic pemphigus foliaceus (fogo selvagem). I. Clinical features and immunopathology. *J Am Acad Dermatol*. 1989;20:657–69.
237. Aoki V, Rivitti EA, Diaz LA. Update on fogo selvagem, an endemic form of pemphigus foliaceus. *J Dermatol*. 2015;42:18–26.
238. Metry DW, Hebert AA, Jordon RE. Nonendemic pemphigus foliaceus in children. *J Am Acad Dermatol*. 2002;46:419–22.
239. Evans MS, Culton DA, Diaz LA, Googe PB, Morrell DS. Childhood pemphigus foliaceus presenting as a polycyclic eruption: case report and review of the literature. *Pediatr Dermatol*. 2019;36:236–41.
240. Galambrun C, Cambazard F, Clavel C, Versini P, Stephan JL. Pemphigus foliaceus. *Arch Dis Child*. 1997;77:255–7.
241. Connelly EA, Aber C, Kleiner G, Nousari C, Charles C, Schachner LA. Generalized erythrodermic pemphigus foliaceus in a child and its successful response to rituximab treatment. *Pediatr Dermatol*. 2007;24:172–6.
242. Zhao CY, Chiang YZ, Murrell DF. Neonatal autoimmune blistering disease: a systematic review. *Pediatr Dermatol*. 2016;33:367–74.
243. Bonifazi E, Milioto M, Trashlieva V, Ferrante MR, Mazzotta F, Coviello C. Neonatal pemphigus vulgaris passively transmitted from a clinically asymptomatic mother. *J Am Acad Dermatol*. 2006;55(5):S113–S114.
244. Fenniche S, Benmously R, Marrak H, Dhaoui A, Ammar FB, Mokhtar I. Neonatal pemphigus vulgaris in an infant born to a mother with pemphigus vulgaris in remission. *Pediatr Dermatol*. 2006;23:124–7.
245. Tope WD, Kamino H, Briggaman RA, Rico MJ, Prose NS. Neonatal pemphigus vulgaris in a child born to a woman in remission. *J Am Acad Dermatol*. 1993;29:480–5.
246. Walker DC, Kolar KA, Hebert AA, Jordon RE. Neonatal pemphigus foliaceus. *Arch Dermatol*. 1995;131:1308–11.
247. Kardos M, Levine D, Gürcan HM, Ahmed RA. Pemphigus vulgaris in pregnancy: analysis of current data on the management and outcomes. *Obstet Gynecol Surv*. 2009;64:739–49.
248. van Beek N, Holtsche MM, Atefi I. State-of-the-art diagnosis of autoimmune blistering diseases. *Front Immunol*. 2024;15:1363032.

SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

How to cite this article: Nanda A, Tedbirt B, Bodemer C, Bolling MC, El Hachem M, Nikolić M, et al. European S2k guidelines on management of autoimmune blistering diseases in children and adolescents. *J Eur Acad Dermatol Venereol*. 2026;40:1137–1161. <https://doi.org/10.1111/jdv.70308>