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**Review Article**

# Recognizing and managing dermatologic manifestations of celiac disease in children

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## ABSTRACT

Celiac disease (CD) is a chronic autoimmune disorder triggered by gluten ingestion, affecting multiple organ systems beyond the gastrointestinal tract. Among its extraintestinal manifestations, dermatologic conditions play a significant role, particularly in pediatric patients. Dermatitis herpetiformis (DH), a hallmark dermatologic feature of CD, is characterized by intensely pruritic vesicles and papules predominantly on extensor surfaces. Other associated skin conditions include chronic eczema, psoriasis, alopecia areata and chronic urticaria, which, while less specific, are significantly more common in children with CD. The pathophysiology of these manifestations involves immune dysregulation, IgA immune complex deposition and heightened inflammatory responses, all linked to gluten sensitivity. Diagnosing CD in children with dermatologic symptoms poses challenges due to overlapping presentations with other skin disorders and variable serologic findings. Direct immunofluorescence of perilesional skin for IgA deposits remains the gold standard for DH, while serologic markers and intestinal biopsies support diagnosis in other cases. Non-specific manifestations often require a heightened clinical suspicion and a multidisciplinary diagnostic approach. Emerging non-invasive techniques may improve diagnostic accuracy and patient compliance in pediatric populations. Management centers on strict adherence to a gluten-free diet (GFD), which resolves most skin and systemic symptoms over time. Pharmacological interventions, such as dapsone, are crucial for symptomatic relief in DH while the GFD takes effect. Non-DH dermatologic conditions may necessitate adjunct therapies like immunosuppressants or biologics. Comprehensive management also addresses psychosocial impacts, with counseling and multidisciplinary care playing a pivotal role. Regular monitoring ensures adherence to treatment and evaluates outcomes, particularly in reducing symptom burden and improving quality of life. Understanding the dermatologic manifestations of CD in children enhances early recognition, timely intervention and holistic management, ultimately improving outcomes in this vulnerable population. Early detection through skin presentations highlights the need for integrating dermatologic findings into CD diagnostic frameworks.

**Keywords:** Autoimmune skin disorders, Celiac disease, Dermatitis herpetiformis, Gluten-free diet, Pediatric dermatology

## INTRODUCTION

Celiac disease (CD) is a chronic autoimmune condition triggered by gluten ingestion in genetically predisposed individuals, characterized by an inappropriate immune response that primarily affects the small intestine. While its gastrointestinal symptoms, such as diarrhea, abdominal pain and malabsorption, are well-documented, the extraintestinal manifestations, particularly dermatologic findings, are gaining increased recognition. In children, such manifestations are often the first indicators of underlying gluten intolerance and may provide crucial diagnostic clues.[1] Understanding the cutaneous presentations of CD is vital for timely diagnosis and management, which can prevent long-term complications and improve quality of life.

The most well-known dermatologic manifestation of CD is dermatitis herpetiformis (DH), a chronic, intensely pruritic blistering skin condition. DH has a strong association with CD, with the majority of affected individuals showing histologic evidence of intestinal damage, even if asymptomatic.[2] The pathogenesis of DH involves immune complex deposition, primarily IgA, in the dermal papillae, triggered by gluten ingestion. While DH is a hallmark finding, other dermatologic conditions have been associated with CD, including alopecia areata, psoriasis and atopic dermatitis.

These conditions, although less specific, are more prevalent in children with CD compared to the general population, highlighting the broader impact of gluten sensitivity on the skin.[3] The diagnostic process for dermatologic manifestations of CD requires a multidisciplinary approach. Skin biopsy with direct immunofluorescence remains the gold standard for diagnosing DH, revealing granular IgA deposits in the dermal papillae. In cases where DH or other dermatologic symptoms raise suspicion of CD, serologic testing for tissue transglutaminase (tTG) and endomysial antibodies is often performed. Confirmatory diagnosis involves upper gastrointestinal endoscopy with biopsy to assess villous atrophy in the small intestine. Importantly, these diagnostic steps should be undertaken while the patient is consuming gluten to avoid false-negative results.[4]

Management of the dermatologic manifestations of CD revolves around strict adherence to a GFD, which addresses both gastrointestinal and extraintestinal symptoms. In cases of DH, additional pharmacologic treatments such as dapsone are often employed to provide rapid symptom relief while dietary changes take effect. Early recognition and treatment of dermatologic presentations in children with CD can mitigate physical discomfort and prevent the psychosocial impacts associated with chronic skin conditions.

Furthermore, it underscores the importance of comprehensive care in children with CD, extending beyond the gastrointestinal system to address systemic manifestations. Celiac disease represents a multifaceted condition with a spectrum of clinical presentations. Dermatologic manifestations, particularly in children, provide a unique window into the systemic effects of gluten sensitivity. Heightened awareness among clinicians and integration of dermatologic findings into the diagnostic framework for CD are crucial for early intervention. This understanding not only improves patient outcomes but also emphasizes the systemic nature of CD as a disease that extends well beyond the gut.

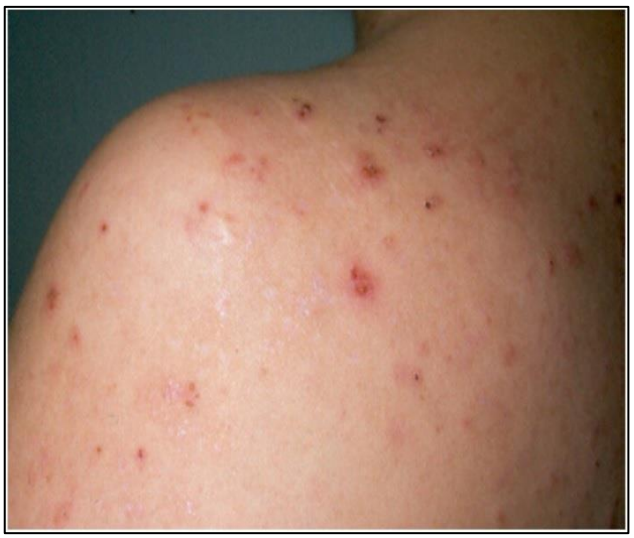

**Figure 1: Several evolutive dermatitis herpetiform lesion in left shoulder of a teenager.[5]**

The dermatologic manifestations of CD in children, such as DH, provide a unique lens through which the systemic effects of gluten sensitivity can be observed. DH, characterized by intensely pruritic vesicular eruptions on extensor surfaces, is highly specific to CD and often represents the skin's immunologic response to gluten ingestion. The pathogenesis involves the deposition of IgA antibodies in the dermal papillae, which trigger an inflammatory cascade leading to the clinical symptoms.

The presence of these deposits can serve as a diagnostic hallmark, emphasizing the need for dermatologic evaluation in children presenting with suspicious skin lesions.[6] In addition to DH, less specific skin conditions, such as eczema, psoriasis and alopecia areata, are increasingly recognized in pediatric CD. These manifestations, while not unique to CD, may be exacerbated by the systemic inflammation and immune dysregulation associated with gluten sensitivity.

Studies have indicated that strict adherence to a GFD leads to significant improvement or resolution of both gastrointestinal and dermatologic symptoms, reinforcing the pivotal role of dietary management.[7] Integrating dermatologic findings into the diagnostic framework for CD underscores the necessity of multidisciplinary care, ensuring timely diagnosis and comprehensive treatment for affected children.

## COMMON DERMATOLOGIC MANIFESTATIONS IN PEDIATRIC CELIAC DISEASE

CD is not only a gastrointestinal disorder but also a systemic condition that can manifest in various extraintestinal ways, including dermatologic presentations. Among these, DH stands out as the classic skin manifestation associated with CD. DH typically presents as intensely pruritic vesicles and papules, symmetrically distributed on extensor surfaces such as the elbows, knees, buttocks and scalp. This condition is particularly significant because it often correlates with asymptomatic intestinal damage in children who may not report overt gastrointestinal symptoms. The pathogenesis involves gluten-triggered IgA immune complex deposits in the dermal papillae, which lead to neutrophil infiltration and blister formation. Diagnosis of DH relies on a combination of clinical findings and direct immunofluorescence of skin biopsies, showing granular IgA deposits, even in the absence of circulating antibodies in some pediatric cases.[8,9]

Apart from DH, other dermatologic conditions have been associated with pediatric CD, albeit with less specificity. These include chronic eczema, which may worsen in the presence of gluten ingestion, possibly due to immune dysregulation or increased systemic inflammation. Psoriasis is another condition noted in children with CD and while its pathophysiology does not directly overlap with gluten sensitivity, studies have suggested that gluten may act as a trigger in genetically predisposed individuals. This is supported by reports of symptom improvement following the adoption of a strict gluten-free diet.[10]

Alopecia areata, an autoimmune condition leading to hair loss, has also been observed more frequently in children with CD compared to the general pediatric population. This association likely stems from shared genetic and immunologic mechanisms, such as a predisposition for HLA-DQ2/DQ8 and increased levels of systemic autoantibodies. While gluten avoidance alone may not fully reverse alopecia areata, early identification of CD in these children allows for appropriate management to mitigate progression.[11]

Urticarial lesions, another dermatologic manifestation, are often underrecognized in pediatric CD. These lesions may be acute or chronic, presenting as wheals or hives that are sometimes mistaken for idiopathic urticaria. The mechanisms by which CD contributes to urticaria remain speculative but may involve heightened immune reactivity or cross-sensitivity to dietary triggers. Interestingly, a gluten-free diet has been shown to reduce the severity and frequency of urticarial outbreaks in affected children, underscoring the importance of dietary intervention even for nonspecific skin conditions.[12] The spectrum of dermatologic manifestations in pediatric CD underscores the systemic nature of this disease. These conditions not only highlight the variability in clinical presentation but also emphasize the need for a multidisciplinary approach to diagnosis and management. Identifying and addressing these skin conditions in children can lead to earlier CD detection, particularly in cases where gastrointestinal symptoms are absent or nonspecific. This ensures better management and ultimately improves the overall well-being of affected patients.

## DIAGNOSTIC CHALLENGES AND APPROACHES

Diagnosing CD in children presenting with dermatologic symptoms poses significant challenges, as these manifestations often lack specificity and may mimic other skin conditions. Dermatitis herpetiformis (DH), while strongly associated with CD, can be misdiagnosed as eczema, contact dermatitis or other pruritic dermatoses due to its clinical similarity to these conditions. In pediatric cases, DH may present with subtler features, such as small papules instead of the more typical vesicles, further complicating diagnosis. Direct immunofluorescence (DIF) of perilesional skin remains the gold standard, revealing granular IgA deposits in the dermal papillae. However, obtaining a biopsy in children can be challenging due to the invasive nature of the procedure and the need for patient cooperation.[12-14]

Another hurdle in diagnosing CD through its dermatologic presentations lies in the variability of serologic findings. While antibodies such as tTG and endomysial antibodies (EMA) are reliable markers for gastrointestinal CD, their sensitivity in DH and other skin manifestations can vary. Some pediatric patients with DH may test negative for these antibodies despite biopsy-confirmed IgA deposits. This discordance underscores the need for a comprehensive diagnostic strategy that combines serologic, histologic and clinical findings.[15] An additional layer of complexity arises when children present with non-specific dermatologic symptoms, such as chronic urticaria, psoriasis or alopecia areata.

These conditions often lead to a delayed or missed diagnosis of CD, as they lack the pathognomonic features of DH. Urticaria, for instance, is commonly attributed to allergens or idiopathic causes, rarely prompting a CD evaluation. Similarly, psoriasis and alopecia areata, though associated with CD, are frequently managed independently without consideration of an underlying gluten-sensitive enteropathy. This highlights the need for heightened clinical suspicion and a low threshold for CD testing in children with these conditions, particularly when other autoimmune or systemic features are present.[16]

The introduction of non-invasive diagnostic tools has offered some promise in addressing these challenges. For example, advanced imaging techniques and skin testing methods are being explored to identify IgA deposits without requiring a full-thickness biopsy. Salivary and stool testing for gluten-specific antibodies are also being

evaluated as adjunctive tools, particularly in younger children where conventional blood tests or invasive procedures are less feasible. However, these methods remain in experimental stages and are not yet widely adopted in clinical practice. Despite their potential, their reliability in diagnosing dermatologic manifestations of CD has yet to be established.[17] Ultimately, successful diagnosis relies on a multidisciplinary approach that integrates dermatologic, gastroenterologic and immunologic expertise. Dermatologists play a crucial role in identifying suspicious skin findings, while pediatricians and gastroenterologists provide the systemic evaluation needed to confirm CD. This collaborative approach is essential in ensuring timely recognition and management, minimizing the potential for diagnostic delays or mismanagement.

## MANAGEMENT STRATEGIES AND OUTCOMES FOR SKIN INVOLVEMENT

Effective management of skin manifestations in children with CD begins with strict adherence to GFD, which serves as the cornerstone of treatment. The removal of gluten from the diet addresses the underlying autoimmune mechanism, leading to significant improvement or resolution of dermatologic symptoms, particularly in cases of DH. Evidence suggests that while gastrointestinal symptoms may respond rapidly to a GFD, skin improvement can take several months to years, highlighting the chronic nature of DH and the importance of sustained dietary compliance. Parents and caregivers of pediatric patients often face challenges in maintaining a GFD due to social, cultural and logistical factors, underscoring the need for comprehensive nutritional counseling and ongoing support.[18]

Pharmacological intervention may be necessary for symptomatic relief, particularly in the initial phases of treatment for DH. Dapsone, a sulfone antibiotic, is commonly prescribed for its rapid anti-inflammatory effects. By inhibiting neutrophil activity, dapsone can quickly alleviate the pruritic and blistering symptoms of DH. However, its use in pediatric populations requires careful monitoring due to potential side effects, such as hemolytic anemia and methemoglobinemia. Clinicians often adopt a tapering approach, reducing the dose as the GFD begins to take effect and control the disease.[19]

Beyond DH, the management of non-specific dermatologic conditions associated with CD, such as chronic urticaria, psoriasis and alopecia areata, often involves a combination of dietary and conventional therapies. For example, chronic urticaria in CD patients has been shown to improve with GFD, suggesting that gluten removal reduces systemic inflammation and immune activation. Psoriasis and alopecia areata, on the other hand, may require additional treatments, including topical corticosteroids, systemic immunosuppressants or biologic agents, depending on the severity of the condition. In these cases, the GFD acts as an adjunct, addressing the underlying gluten sensitivity while primary therapies target the specific dermatologic pathology.[20]

Monitoring outcomes and ensuring adherence to treatment is critical in pediatric CD, especially given the long-term implications of the disease. Dermatologic improvement is often used as a marker of dietary compliance, as the resolution of skin symptoms typically correlates with histologic recovery of the intestinal mucosa. Regular follow-ups with dermatologists and gastroenterologists are recommended to assess treatment efficacy, manage potential relapses and provide education on gluten cross-contamination risks. Additionally, advancements in monitoring tools, such as mobile apps for dietary tracking and antibody level testing kits, have emerged as valuable aids in enhancing adherence and identifying lapses in gluten avoidance.[21]

The psychological and social impact of skin conditions in children with CD must also be considered in management strategies. Chronic itching, visible lesions and scarring can significantly affect a child's self-esteem and social interactions. Counseling and support groups can provide emotional and psychological support for both patients and their families, fostering better coping mechanisms and adherence to long-term dietary restrictions. Multidisciplinary care that includes dermatologists, dietitians and psychologists ensures that both the physical and emotional aspects of CD are addressed, contributing to holistic management and improved outcomes.

## CONCLUSION

In pediatric celiac disease, recognizing dermatologic manifestations is crucial for early diagnosis and effective management. Dermatitis herpetiformis remains a hallmark feature, while other skin conditions provide additional diagnostic clues. A gluten-free diet is the cornerstone of treatment, supported by pharmacologic and multidisciplinary approaches for optimal outcomes. Addressing both physical and psychosocial aspects ensures comprehensive care for affected children.

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