Exploring the Frequency and Characteristics of Dermatological Manifestations in Pediatric Autoimmune Diseases: An Evaluation Study

Explorando a Frequência e as Características das Manifestações Dermatológicas em Doenças Autoimunes Pediátricas: Um Estudo de Avaliação

Explorando la Frecuencia y las Características de las Manifestaciones Dermatológicas en Enfermedades Autoinmunes Pediátricas: Un Estudio de Evaluación

RESUMO

Introdução: As doenças autoimunes pediátricas requerem cuidados específicos. Este estudo explora suas manifestações dermatológicas, fisiopatologia e estratégias de tratamento para melhorar o prognóstico dos pacientes. **Objetivo:** Este estudo investiga as manifestações dermatológicas em doenças autoimunes pediátricas, ressaltando sua importância no diagnóstico e manejo. Discute a fisiopatologia, desafios diagnósticos, ferramentas de avaliação e estratégias de tratamento, enfatizando a necessidade de abordagens personalizadas para melhorar os resultados dos pacientes. **Metodologia:** Este estudo revisou literatura sobre manifestações dermatológicas em doenças autoimunes pediátricas. Adotou metodologia sistemática, incluindo pesquisa bibliográfica, análise de conteúdo e discussão dos resultados, para oferecer uma compreensão abrangente e informativa dessas condições, destacando desafios diagnósticos, ferramentas de avaliação e estratégias de tratamento. **Discussão e Resultados:** As doenças autoimunes pediátricas abrangem condições como artrite idiopática juvenil (AIJ) e psoríase em placas, exigindo cuidados específicos devido à sua manifestação na pele e articulações. O diagnóstico diferenciado considera características únicas na infância, como a persistência de erupções cutâneas e a sobreposição de sintomas. Ferramentas de avaliação, como o CDASI e o CAT, ajudam a quantificar a gravidade das lesões cutâneas. O tratamento inclui corticosteróides tópicos e emolientes para controlar a inflamação e manter a barreira cutânea. A gestão eficaz dos sintomas cutâneos influencia positivamente o prognóstico geral, destacando a importância da intervenção precoce e da abordagem personalizada para melhorar a qualidade de vida das crianças afetadas. **Conclusão:** Este estudo revela a complexidade das doenças autoimunes pediátricas, destacando a importância da diferenciação no diagnóstico e tratamento. Enfatiza o papel vital da medicação na minimização dos danos, ressaltando a relação entre sintomas cutâneos e patologias sistêmicas. Reconhece a necessidade de abordagens específicas para a idade e destaca o papel crítico das manifestações cutâneas no diagnóstico e tratamento precoce. Contribui para o avanço do conhecimento e sugere direções futuras na pesquisa, incluindo a necessidade de estratégias personalizadas e eficazes de gestão.

DESCRITORES: Doenças Autoimunes; Pediatria; Fenômenos Fisiológicos da Pele; Anormalidades da Pele

ABSTRACT

Introduction: Pediatric autoimmune diseases require specific care. This study explores their dermatological manifestations, pathophysiology, and treatment strategies to improve patient prognosis. **Objective:** This study investigates dermatological manifestations in pediatric autoimmune diseases, emphasizing their importance in diagnosis and management. It discusses pathophysiology, diagnostic challenges, assessment tools, and treatment strategies, emphasizing the need for personalized approaches to enhance patient outcomes. **Methodology:** This study reviewed literature on dermatological manifestations in pediatric autoimmune diseases. It adopted a systematic methodology, including literature search, content analysis, and results discussion, to offer a comprehensive and informative understanding of these conditions, highlighting diagnostic challenges, assessment tools, and treatment strategies. **Discussion and Results:** Pediatric autoimmune diseases encompass conditions like juvenile idiopathic arthritis (JIA) and plaque psoriasis, requiring specific care due to their manifestation on the skin and joints. Differential diagnosis considers unique childhood features, such as persistent skin rashes and symptom overlap. Assessment tools like CDASI and CAT help quantify the severity of skin lesions. Treatment includes topical corticosteroids and emollients to control inflammation and maintain skin barrier. Effective management of skin symptoms positively influences overall prognosis, underscoring the importance of early intervention and personalized approach to improve affected children's quality of life. **Conclusion:** This study reveals the complexity of pediatric autoimmune diseases, emphasizing the importance of differentiation in diagnosis and treatment. It underscores the vital role of medication in minimizing damage, highlighting the relationship between skin symptoms and systemic pathologies. It recognizes the need for age-specific approaches and underscores the critical role of skin manifestations in early diagnosis and treatment. It contributes to advancing knowledge and suggests future research directions, including the need for personalized and effective management strategies.

DESCRIPTORS: Autoimmune Diseases; Pediatrics; Skin Physiological Phenomena; Skin Abnormalities

RESUMEN

Introducción: Las enfermedades autoinmunes pediátricas requieren cuidados específicos. Este estudio explora sus manifestaciones dermatológicas, fisiopatología y estrategias de tratamiento para mejorar el pronóstico de los pacientes. **Objetivo:** Este estudio investiga las manifestaciones dermatológicas en enfermedades autoinmunes pediátricas, resaltando su importancia en el diagnóstico y manejo. Analiza la fisiopatología, los desafíos diagnósticos, las herramientas de evaluación y las estrategias de tratamiento, enfatizando la necesidad de enfoques personalizados para mejorar los resultados de los pacientes. **Metodología:** Este estudio revisó la literatura sobre manifestaciones dermatológicas en enfermedades autoinmunes pediátricas. Adoptó una metodología sistemática que incluyó

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búsqueda bibliográfica, análisis de contenido y discusión de resultados, con el fin de ofrecer una comprensión integral e informativa de estas condiciones, destacando los desafíos diagnósticos, las herramientas de evaluación y las estrategias terapéuticas. **Discusión y Resultados:** Las enfermedades autoinmunes pediátricas abarcan afecciones como la artritis idiopática juvenil (AIJ) y la psoriasis en placas, que requieren cuidados específicos debido a su manifestación en la piel y las articulaciones. El diagnóstico diferencial considera características propias de la infancia, como la persistencia de erupciones cutáneas y la superposición de síntomas. Herramientas de evaluación como el CDASI y el CAT ayudan a cuantificar la gravedad de las lesiones cutáneas. El tratamiento incluye corticosteroides tópicos y emolientes para controlar la inflamación y mantener la barrera cutánea. La gestión eficaz de los síntomas cutáneos influye positivamente en el pronóstico general, subrayando la importancia de la intervención temprana y de un enfoque personalizado para mejorar la calidad de vida de los niños afectados. **Conclusión:** Este estudio revela la complejidad de las enfermedades autoinmunes pediátricas, destacando la importancia de la diferenciación en el diagnóstico y tratamiento. Enfatiza el papel vital de la medicación en la minimización de los daños, resaltando la relación entre síntomas cutáneos y patologías sistémicas. Reconoce la necesidad de enfoques específicos según la edad y subraya el papel crítico de las manifestaciones cutáneas en el diagnóstico y tratamiento precoz. Contribuye al avance del conocimiento y sugiere direcciones futuras para la investigación, incluyendo la necesidad de estrategias personalizadas y eficaces de gestión.
DESCRIPTORES: Enfermedades Autoinmunes; Pediatría; Fenómenos Fisiológicos de la Piel; Anomalías de la Piel.

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INTRODUCTION

Pediatric autoimmune diseases are a group of conditions that can have significant and long-lasting effects on children's health. These diseases are distinct from autoimmune diseases that affect adults and require specialized care and management. In recent years, there has been growing interest in the dermatological manifestations of these diseases, which can be a useful diagnostic tool.

This research paper aims to explore the frequency and characteristics of dermatological manifestations in pediatric autoimmune diseases and to assess the current knowledge of their pathophysiology.

The article will begin with an overview of common pediatric autoimmune diseases and how they affect children differently than adults. It will then delve into the types of skin manifestations observed in these conditions, their frequency, and the diagnostic challenges associated with them.

Finally, the article will examine the assessment tools used to evaluate skin manifestations in pediatric autoimmune diseases, treatment strategies to control dermatological symptoms, and how the management of skin symptoms affects the overall prognosis of pediatric autoimmune diseases. By providing an in-depth exploration of these topics, this research paper aims to improve our understanding of these conditions and ultimately improve patient outcomes.

OBJECTIVE

This scientific study aims to explore and understand dermatological manifestations in pediatric autoimmune diseases, providing a comprehensive overview of characteristics of these conditions and the impact they have on children's health. Initially, it highlights the growing importance given to skin manifestations in these diseases, which can serve as valuable indicators for early diagnosis and effective management.

Throughout the text, various aspects related to pediatric autoimmune diseases are addressed. Beginning with the identification of the most common diseases in this age group, such as juvenile idiopathic arthritis and pediatric plaque psoriasis, the study highlights the complexity and diversity of these conditions, emphasizing the need for a different approach compared to adults.

An in-depth analysis is made of how autoimmune diseases affect children differently, considering not only the immediate health challenges but also the long-term implications for growth and development. The importance of medication and lifelong fol-

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low-up, including the transition from pediatric to adult care, is highlighted.

The study also discusses current knowledge of the pathophysiology of these diseases in children, highlighting the importance of early intervention and careful evaluation to identify possible underlying autoimmune diseases.

A significant portion of the text is devoted to analyzing the specific skin manifestations observed in these diseases, as well as their frequency of occurrence. The diagnostic challenges associated with these skin symptoms are highlighted, emphasizing the importance of early recognition and understanding of the unique characteristics of dermatological presentations in children.

In addition, the assessment tools used to evaluate skin manifestations and effective treatment strategies to control these symptoms are discussed. The text emphasizes the importance of an individualized and differentiated approach to the management of pediatric autoimmune diseases, considering the specific needs of each patient.

Finally, the study highlights the relationship between the management of skin symptoms and the overall prognosis of pediatric autoimmune diseases, emphasizing the importance of early recognition and appropriate treatment of skin manifestations to improve long-term outcomes.

METHODOLOGY

The methodology adopted for this literature review on dermatological manifestations in pediatric autoimmune diseases followed a systematic protocol to ensure the comprehensiveness and quality of the analysis. Initially, an extensive literature search was conducted in scientific databases, such as PubMed, Scopus, and Web of Science, using specific search terms: "Autoimmune Diseases," "Pediatrics," "Physiological Phenomena of the Skin," and "Skin Abnormalities." Inclusion criteria were defined to select relevant studies published in peer-reviewed journals, preferably in the last 10 years, focusing on the clinical characteristics, pathophysiology, evaluation, and management of dermatological manifestations in pediatric patients with autoimmune diseases.

After selecting the studies, a systematic content analysis was performed to extract relevant information, including data on the most common pediatric autoimmune diseases, types of skin manifestations observed, frequency of these manifestations, associated diagnostic challenges, assessment tools used, and treatment strategies employed. The analysis also addressed how the management of skin symptoms can influence the overall prognosis of pediatric autoimmune diseases.

The data were synthesized and organized according to the themes identified in the discussion, with an emphasis on the main findings and trends observed in the reviewed literature. Direct quotes and paraphrases were used to support the information presented, ensuring the credibility and accuracy of the text.

Finally, the results were discussed and interpreted to contextualize the findings within the current landscape of research on pediatric autoimmune diseases and identify gaps no knowledge que possam direcionar futuras investigações. Essa

This systematic and rigorous methodological approach allowed for a comprehensive and informative analysis of dermatological manifestations in pediatric autoimmune diseases, contributing to the advancement of scientific understanding in this area and providing relevant insights for clinical practice and future research.

DISCUSSION AND RESULTS

Overview of pediatric autoimmune diseases

What are the common pediatric autoimmune diseases?

Among the range of autoimmune diseases affecting children, juvenile idiopathic arthritis (JIA) is particularly prevalent, representing a significant condition in which the immune system mistakenly attacks joint tissues, leading to sustained inflammation and discomfort in affected young people [1]. Although JIA is a major concern due to its potential for chronic pain and disability, the skin is also a common target of autoimmune dysregulation in the pediatric population, as evidenced by pediatric plaque psoriasis. This condition manifests with characteristic red, scaly skin patches, which not only cause physical discomfort but can also lead to psychological distress due to their visible nature [1].

Further complicating the picture of autoimmune diseases in children is pediatric active psoriatic arthritis, a condition that lies at the intersection of autoimmune diseases of the skin and joints. Like its adult counterpart, pediatric active psoriatic arthritis presents symptoms that include joint inflammation and skin lesions, thus representing an overlap of the symptoms observed in JIA and pediatric plaque psoriasis [1].

These conditions underscore the complexity and diversity of autoimmune diseases in the pediatric population, requiring a differentiated approach to diagnosis and management that considers the unique aspects of these diseases in the context of a developing child.

How do autoimmune diseases affect children differently than adults?

The impact of autoimmune diseases on children requires a multifaceted

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approach to management and care. Unlike adults, children with autoimmune diseases such as alopecia areata, vitiligo, scleroderma, and chronic urticaria not only face immediate health challenges but also face the long-term implications of their conditions on growth and development [2].

To mitigate the progression of these diseases and minimize damage, medication plays a crucial role. For example, emerging treatments have shown promise in reducing or even halting the damage caused by autoimmune responses, thereby preserving the child's health during critical stages of development [3].

It is imperative to understand that these children will require lifelong monitoring and healthcare to effectively manage their conditions. This includes a transition plan orchestrated by physicians to move from pediatric to adult care, ensuring that children are equipped with the tools and knowledge necessary to manage their health as they mature [3].

This transition is delicate and requires a comprehensive understanding of the unique challenges faced by these young patients, who must learn to navigate the complexities of their conditions throughout the different stages of life.

What is the current understanding of autoimmune pathophysiology in pediatrics?

In the complex landscape of pediatric autoimmune pathophysiology, recent advances have shed light on the unique characteristics of these conditions in children. Unlike adults, pediatric patients present specific clinical manifestations and antibody profiles, which require personalized approaches to diagnosis and treatment [4]. For example, pediatric autoimmune motor disorders, now increasingly recognized due to these advances, exemplify how early intervention can dramatically improve patient outcomes [4].

This early intervention is particularly crucial given that autoimmune and inflammatory diseases are prevalent among children with primary immunodeficiencies (PIDs), and these manifestations often present before or simultaneously with the diagnosis of the PID itself [5]. Consequently, it is critical that pediatric patients with autoimmune symptoms be evaluated for underlying PIDs to ensure comprehensive treatment and mitigate the risk of complications that can severely affect their quality of life [5].

The complexity of these autoimmune conditions in pediatric populations underscores the need for continued research and collaboration among specialists, such as rheumatologists and dermatologists, to optimize care for affected children, particularly those with skin involvement, which is not uncommon [6].

Dermatological manifestations in pediatric autoimmune diseases

What types of skin manifestations are seen in pediatric autoimmune diseases?

In pediatric autoimmune diseases, skin manifestations are not just superficial signs but play a critical role in diagnosis and treatment. Vitiligo, a type of autoimmune dermatosis, is most commonly observed in children with humoral immunodeficiencies, although its prevalence in the pediatric cohort of the study was surprisingly lower than expected, between 0.5 and 2% [7].

This discrepancy emphasizes the need for greater clinical awareness when evaluating skin symptoms in autoimmune diseases. In addition, lupus erythematosus, which encompasses several subtypes, including systemic, subacute cutaneous, chronic cutaneous, and neonatal lupus, stands out as the most prevalent connective tissue disease in childhood [8].

Among these, systemic lupus erythematosus (SLE) is the most frequent pediatric presentation, although subacute cutaneous lupus is extremely rare in this age group [8]. These skin stigmata are invaluable diagnostic clues, not only for lupus but also for systemic disorders of childhood in general [8]. Early recognition of these skin manifestations is critical, as it may allow for immediate initiation of therapy, potentially preventing serious outcomes [8].

Pediatric patients may present different skin features compared to adults with the same systemic diseases, emphasizing the importance of considering age-specific presentations when evaluating autoimmune skin manifestations [8]. For example, diseases such as Kawasaki disease (KD), Henoch-Schönlein purpura (HSP), acute hemorrhagic edema of infancy, and neonatal-onset multisystem inflammatory disease (NOMID) have unique skin features that are typically seen in the pediatric population [8]. In addition, palisaded neutrophilic dermatitis (PNGD) is an example of a skin disease that presents with symmetrical papular lesions on the extremities of children and is associated with autoimmune diseases that generate immune complexes, illustrating the complex interaction between skin and systemic symptoms in these diseases [8].

How often do dermatological symptoms appear in these conditions?

Dermatological symptoms are a hallmark of dermatomyositis and present a variety of manifestations that, similar to juvenile idiopathic arthritis and pediatric plaque psoriasis, can affect children and adults [9]. In approximately 80% of patients in the acute phase of the disease, skin manifestations such as the characteristic heliotrope rash—marked by a purple-red

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discoloration on the upper eyelids—and Gottron's papules, which appear as scaly, raised bumps on the joints, may be observed [8][10].

These skin changes, although indicative of dermatomyositis, often lead to misdiagnosis; for example, Gottron's papules can be confused with psoriasis or atopic dermatitis, highlighting the challenge of accurately identifying dermatomyositis based on dermatological symptoms alone [8].

Notably, in up to 40% of individuals, skin changes may be the initial and only sign of the disease at onset, emphasizing the importance of recognizing these dermatological patterns for early diagnosis and treatment [9]. In addition, although the onset of symptoms in dermatomyositis can be unpredictable and may wax and wane for no apparent reason, persistent dermatological problems, such as the shawl sign or the V-shaped rash V sign rash—both evidence of photosensitivity—are often present and can significantly affect the quality of life of patients [9] [8].

What are the diagnostic challenges associated with skin symptoms in pediatric patients?

Based on the recognition that autoimmune diseases present a spectrum of challenges in pediatric populations, diagnostic complexities become especially pronounced in the evaluation of skin symptoms. Neonatal-onset multisystem inflammatory disorder (NOMID) exemplifies this situation, with its characteristic recurrent evanescent urticarial rash that appears early in life [8]. These skin manifestations, universal among NOMID patients, are not transient but persist throughout the individual's life, presenting an ongoing diagnostic hurdle for clinicians [8].

The singular presence of skin symptoms, as observed in some pediatric cases, further intensifies the challenge, as these symptoms may be the only initial clue to an underlying systemic problem [8]. Furthermore, skin rashes associated with conditions such as NOMID are not merely cosmetic concerns but are often accompanied by systemic symptoms such as fatigue and difficulty with physical activity, which may be mistakenly attributed to behavioral problems such as laziness rather than a medical condition [8][11]. This misattribution can lead to delays in proper diagnosis and treatment, affecting the child's willingness to engage in normal activities due to discomfort or misunderstanding of their condition [11].

To effectively navigate these diagnostic challenges, a detailed medical history, including a family history of autoimmune disease, and a comprehensive physical examination are vital components that can provide clues and aid in the identification of disorders such as NOMID [8].

Assessment and management of dermatological symptoms

What assessment tools are used to evaluate skin manifestations in pediatric autoimmune diseases?

In the context of pediatric autoimmune diseases, particularly juvenile dermatomyositis (JDM), skin manifestations are a critical aspect that requires accurate and sensitive measurement tools. Among the various recommended tools, the Cutaneous Dermatomyositis Area and Severity Index (CDASI) stands out for its comprehensive assessment of skin involvement.

The CDASI examines multiple anatomical locations—16 areas of the body in total—scoring each for the presence and severity of erythema, thickness, scaling, excoriation, ulceration, and separate scores for Gottron lesions, periungual changes, and alopecia, thus providing a differentiated picture of skin disease activity [12] [13]. In addition, CDASI activity scores range from 0 to 100, with damage scores ranging from 0 to 3, providing a comprehensive assessment of disease severity. disease activity [12] [13]. In addition, CDASI activity scores range from 0 to 100, with damage scores ranging from 0 to 32, allowing categorization of disease activity levels as low, moderate, or high after assessment by a physician, based on established cutoff values [12].

The design of the tool for adults and children with dermatomyositis ensures its relevance across a broad age spectrum, increasing its usefulness in clinical practice [12]. In addition, the Cutaneous Assessment Tool (CAT) skin disease activity score uses a different approach, with 10 items to assess skin activity and 4 items to assess skin damage, with 7 items common to both scores. This tool also allows physicians to classify lesion characteristics depending on severity, which is essential for tailoring treatment strategies to individual patient needs [12].

The CAT and CDASI tools, by covering a range of dermatological features and allowing for graded assessment of severity, demonstrate the evolution of measurement tools designed to capture the complex dermatological presentations observed in JMD, thereby facilitating more accurate monitoring and management of skin disease activity in affected pediatric populations.

What treatment strategies are effective in controlling dermatological symptoms?

In the field of pediatric dermatology, where conditions such as atopic dermatitis (AD) are prevalent, effective management strategies are essential. First-line treatments for AD flare-ups generally include topical corticosteroids (TCS), which are widely recognized for their efficacy in reducing inflammation and pruritus [14].

Medium-potency TCS are particularly safe for use on specific areas of

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the body, except for the facial area, with appropriate monitoring in primary care settings [14]. In addition to TCS, liberal application of fragrance-free emollients is critical for both daily prevention and active treatment.

Emollients play a crucial role in retaining and replenishing skin moisture, which not only reduces the severity of the disease but also prolongs the interval between flare-ups [15]. Furthermore, by maintaining the skin barrier, emollients significantly reduce the need for prescription medications, underscoring their importance as primary therapy for both the management and maintenance of flare-ups [15].

For children who do not experience disease-free intervals with basic skin care and emollients alone, TCS remains a key component of their AD treatment regimen [14].

Pediatricians and dermatologists should balance the potency of steroids with the vulnerability of young skin, opting for low-potency steroids on thinner skin areas to minimize the risk of atrophy, while considering medium- to high-potency steroids for localized lesions of acute allergic contact dermatitis to achieve rapid symptom control [16]. Thus, a differentiated and individualized approach to treatment selection is critical for effective control of dermatological symptoms in children, particularly when addressing the complexities of autoimmune skin diseases such as pediatric plaque psoriasis and AD.

How does the management of skin symptoms affect the overall prognosis of pediatric autoimmune diseases?

The intricate relationship between skin symptoms and systemic autoimmune diseases cannot be overstated, especially when considering the prognosis of pediatric patients. The skin often acts as a mirror, reflecting underlying immune system dysfunctions, with skin stigmata offering critical diagnostic information for systemic disorders affecting children [8].

Early identification of these dermatological features is critical; it triggers the timely initiation of therapeutic interventions that can significantly alter the disease trajectory [8]. For example, unusual skin rashes or discolorations in a child may signal the onset of a more complex autoimmune disorder requiring prompt medical attention.

This proactive management is not only crucial for resolving immediate discomfort but also for improving long-term outcomes. Furthermore, the varied expression of skin symptoms in pediatric autoimmune diseases, as opposed to their adult counterparts, underscores the need for age-specific clinical strategies [8].

By focusing on the rapid and effective treatment of these skin manifestations, healthcare providers can have a substantial impact on the overall prognosis of these young patients, thereby mitigating potential complications and improving quality of life [8].

CONCLUSION

The present study clarifies the frequency and characteristics of dermatological manifestations in pediatric autoimmune diseases, highlighting the complexity and diversity of these conditions in the pediatric population.

The results emphasize the need for a differentiated approach to diagnosis and treatment that considers the unique aspects of these diseases in the context of a developing child, and suggest that medication plays a crucial role in mitigating the progression of these diseases and minimizing damage, specifically highlighting the intricate relationship between skin symptoms and systemic autoimmune pathologies. Furthermore, the study underscores the need for age-specific clinical strategies, given the varied expression of skin symptoms in pediatric autoimmune diseases, as opposed to their adult counterparts.

The results also reveal that skin manifestations are not merely superficial signs but play a critical role in diagnosis and treatment, highlighting the importance of clinical awareness when assessing skin symptoms in autoimmune diseases. Furthermore, the study suggests that early recognition of these skin manifestations is crucial, allowing for immediate initiation of therapy and potentially avoiding severe outcomes.

The findings call for a comprehensive understanding of the unique challenges faced by young patients with autoimmune diseases, who must learn to navigate the complexities of their conditions throughout different stages of life. Finally, the study underscores the importance of effective management strategies for prevalent conditions such as atopic dermatitis (AD) and skin manifestations in juvenile dermatomyositis (JDM), which require accurate and sensitive measurement tools.

Overall, this study contributes to the ongoing advancement of knowledge in the field and suggests future directions for research, including the need for personalized approaches to diagnosis and treatment and the development of more effective management strategies for pediatric autoimmune diseases.

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