

## **Exploring the Complexity of Cutaneous Mastocytosis in two infants: A Case Study**

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## **Exploring the Complexity of Cutaneous Mastocytosis in 2 infants: A Case Study**

**Introduction & Objectives:** The differential diagnosis and management of Cutaneous Mastocytosis (CM), a rare mast cell disorder in paediatric patients, necessitates a critical evaluation of therapeutic strategies. This study aims to compare the clinical outcomes of two paediatric cases of CM, with a focus on the therapeutic efficacy of Topical Tofacitinib in one patient versus standard treatment protocols in the other.

**Materials & Methods:** The study retrospectively analysed two paediatric patients diagnosed with CM. Patient A, a 7-month-old infant, presented with pruritic, hyperpigmented maculo-papules and was treated with Topical Tofacitinib. Patient B, a 1-year-old, exhibited similar symptoms but was managed with antihistamines, corticosteroids, and avoidance of known triggers without the use of Tofacitinib. Both patients were assessed over a six-month period, with periodic evaluations including clinical examinations, lesion counts, and patient/parental reporting of symptoms.

**Results:** Patient A demonstrated a rapid and significant reduction in lesion count, with a 60% decrease observed within two weeks of initiating Topical Tofacitinib treatment, progressing to an 80% reduction in pruritus after one month. In contrast, Patient B showed a modest improvement in symptoms; however, the reduction in lesion count was less pronounced, and intermittent flare-ups were reported during the treatment period in both patients. Notably, both patients tolerated their respective treatment regimens well, with no adverse effects or treatment-related complications recorded.

**Conclusion:** This comparative study suggests that Topical Tofacitinib may offer a more pronounced therapeutic benefit in the management of CM in infants and young children compared to conventional treatments. The rapid and significant improvement observed in Patient A underscores the potential of JAK inhibitors as a valuable treatment modality in CM. Considering these findings, Topical Tofacitinib warrants further investigation in larger, controlled studies to confirm its efficacy and safety profile. Ultimately, such research could lead to improved management protocols for CM, enhancing the quality of life for affected paediatric patients and providing a substantive base for evidence-based clinical practice.



## Congenital cutaneous candidiasis in a preterm infant

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## **Introduction & Objectives:**

## **Materials & Methods:**

## **Results:**

A preterm 26+1 week infant presented on day 3 of life with superficial erosions on her back. Her skin was normal when born by emergency Caesarean-section due to cord presentation and acute chorioamnionitis. Maternal vaginal swab taken during labour grew *Candida albicans*.

The erosions became extensive and confluent involving her trunk, limbs and neck with haemorrhagic changes most marked at the edges in 48-hours without active bleeding. There were no dysmorphic features, ichthyosis, hyperkeratosis, palmoplantar keratoderma, nail abnormalities or mucosal involvement. It was challenging to assess the background skin changes and erythema due to skin type 6.

Candida albicans was isolated from skin biopsy, tissue, urine and blood cultures. Candida type mycelium was seen on microscopy of tissue biopsy. Histology showed ulcerated skin with broken epidermis. The yeast hyphae and spores abounded within the inflammatory debris. Viral stains were negative. These were consistent with disseminated candidiasis.

Treatment included intravenous ambisome and fluconazole, which was added to aid skin penetration. Subsequent blood cultures identified *Enterococcus faecium* and *Staphylococcus haemolyticus*, necessitating intravenous vancomycin.

She deteriorated with systemic candidiasis, intraventricular haemorrhage, bowel perforation and died on day 13. The placental histology revealed acute necrotising chorioamnionitis with significant fetal inflammatory response and fungal elements consistent with *Candida albicans*, including occasional possible hyphae in fetal vessels.

#### **Conclusion:**

Congenital cutaneous candidiasis is a rare invasive infection with heterogeneous clinical manifestations. Despite its usual self-limiting course, early recognition and prompt treatments are essential to avoid systemic compromise, especially in preterm infants.



#### Atopic dermatitis in the Pediatric population

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## Atopic dermatitis in the Pediatric population

## **Introduction & Objectives:**

Atopic dermatitis (AD), also known as atopic eczema, is a prevalent chronic or recurrent inflammatory skin condition affecting 15–20% of children and 1–3% of adults worldwide. It is characterized by episodic outbreaks of itchy eczematous lesions on dry skin. Typically initiating in early childhood, AD may signify the initial phase of the 'atopic march,' a natural progression of atopic conditions in childhood preceding the development of other allergic disorders in later life. While approximately 70% of individuals with childhood-onset AD experience spontaneous remission before adolescence, it remains an early indicator that a child may later develop asthma and/or allergic rhinitis. AD imposes a substantial burden on healthcare resources and adversely impacts patients' quality of life, primarily due to sleep disturbances caused by itching. Consequently, there is an increased interest in identifying environmental risk factors and protective elements. The objective of this study is to assess the prevalence, epidemiology, and clinical manifestations of AD.

#### **Materials & Methods:**

This is a cross-sectional study conducted between October 2016 and August 2023 among a population of children under the age of 16 who sought medical attention at the dermatology department. The diagnosis of atopic dermatitis was established based on the criteria outlined by the UK Working Party

## **Results:**

Among 3804 children seeking dermatological care, 422 (11%) were diagnosed with AD, predominantly in males (66.05%), with an average age of 4.2 years. AD presented before age 2 in 67.9%, aligning with the 'atopic march' progression. Family history of atopy was found in 38.35%, and 11.1% had asthma. Clinical presentations included eczematous lesions (75.7%), lichenification (17.45%), and hyperpigmented scars (12.6%). Infectious complications, including impetiginization (15.8%), and Kaposi-Juliusberg syndrome (4.7%) were observed. Twelve patients had primary immunodeficiency. Dermocorticosteroids were the primary treatment.

#### **Conclusion:**

In conclusion, our study provides valuable insights into the epidemiological and clinical profile of atopic dermatitis (AD) among the pediatric population. The prevalence of AD in our cohort, along with its gender distribution, age of onset, and associated factors, contributes to a comprehensive understanding of the disease in our specific demographic context. The clinical manifestations observed, including the prevalence of pruritic lesions, eczema lichenification, and associated complications, underscore the significant burden that AD imposes on affected individuals. Furthermore, our findings highlight the importance of early detection and intervention, especially considering the potential links between childhood AD and the development of other allergic disorders later in life. As we continue to unravel the complexities of AD, this research serves as a foundation for future studies and interventions aimed at enhancing our management and prevention strategies for this common inflammatory skin condition.



## Pigmented epithelioid melanocytoma: A case report

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## **Introduction & Objectives:**

Zembowicz et al introduced the term "Pigmented Epithelioid Melanocytoma (PEM)" to describe a rare, low-grade melanocytic tumor closely resembling lesions previously identified as epithelioid blue nevus and animal-type melanoma. In almost a century, only a hundred cases has been reported, of which more than two-thirds in the last five years.

The lesions manifest most frequently in children and young adults, as nodular lesions with pronounced pigmentation. Histopathological examinations reveal a dermal proliferation of heavily pigmented melanocytes, encompassing dendritic and spindle/epithelioid cells, interspersed with melanophages . PEM often involves regional lymph nodes but generally follows a benign clinical trajectory .

#### **Materials & Methods:**

This case study spotlights an exceptional instance of PEM in an 8-year-old patient, unraveling the clinical, histological, and dermoscopic intricacies.

#### **Results:**

The dermatological examination revealed a blackened tumor on the scalp, prompting excision and subsequent histopathologic evaluation. The biopsy unveiled a dermal-based neoplasm, heavily pigmented, characterized by large melanocytes with minimal junctional components and an admixture of melanophages. The incomplete excision later led to a recurrence, emphasizing the challenges in managing this peculiar melanocytic lesion.

Discussions surrounding the broader context of melanocytic lesions delve into the overlapping features of epithelioid blue nevus and animal-type melanoma. PEM, distinctively characterized by its unique histopathological and clinical features, tends to manifest as a nodule or plaque, displaying a black or blue hue with irregular borders. Dermoscopic examinations hint at consistent blue patterns, whitish structures, and a diverse vascular pattern, but these features lack specificity for PEM.

Histopathologically, PEM unfolds as a dense dermal proliferation of heavily pigmented melanocytes, showcasing a spectrum of dendritic and spindle/epithelioid forms, interspersed with melanophages. Its differential diagnosis encompasses various melanocytic lesions, necessitating a meticulous examination to discern malignancies such as blue nevus, melanoma metastasis, and primary dermal melanoma.

Crucially, PEM often involves regional lymph nodes, yet its clinical course typically follows a benign trajectory. Notably, a 2009 study indicates favorable 5-year clinical outcomes for PEM, suggesting its categorization among low-grade or borderline melanocytic tumors. As for treatment, the emphasis is on complete excision of the primary lesion with safety margins, though the low-grade nature challenges the need for extensive excisions based on Breslow depth. Sentinel lymph node biopsy yields minimal benefits, and systemic agents like interferon or MEK/BRAF inhibitors find little relevance in PEM management.

#### **Conclusion:**

This comprehensive exploration of a PEM case underscores the complexity in diagnosing, treating, and understanding this

enigmatic melanocytic lesion. As the medical community navigates through the intricacies of such rare entities, the spotlight on individual cases contributes valuable insights, fostering a deeper comprehension of the clinical landscape.



## Lipoidal necrobiosis revealing type 1 diabetes: A child's case report

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## **Introduction & Objectives:**

Lipoidal necrobiosis (LN) is a rare granulomatous dermatosis. It is reported in 0.3 to 1.2% of diabetics, preferentially located in the leg. The lesions appear as erythematous plaques, with central depressions.

We report the case of an 11 year old child who presented, 5 years before the discovery of her diabetes, asymptomatic well-circumscribed erythematous plaques of the leg. The diagnostic of lipoidal necrobiosis was confirmed. Through this work, we will identify clinical, histological and therapeutic features of lipoidal necrobiosis.

#### Materials & Methods:

It is about an 11 years old child , not known to be diabetic. Having a particular pathological history of erythemato-atrophic plaques on the right leg treated long-term by self-medication dating back 6 years , with no improvement . The patient was admitted to the emergency room for a non-traumatic afebrile consciousness disorder subsequently revealing ketoacidosis. Secondary , a Diabetes type 1 discovered following assessments carried out in the intensive care unit. And to whom the dermatological examination revealed shiny erythemato-telangiectatic plaques, respectively 7cm and 3cm long axis. A skin biopsy was performed confirming the diagnosis of lipoid necrobiosis. With good improvement under treatment.

#### **Results:**

Lipoidal necrobiosis is a chronic non-infectious idiopathic granulomatous disease of the dermis. Due to its increased prevalence in patients diabetes, especially type 1, etiological theories mainly refer to microangiopathy. However, the association with poor glycemic control remains controversial.

The prevalence of diabetes in patients with LN remains controversial, between 11 to 65% depending on the series. The LN reached electively the adult during the 3rd and 4th decades, with a female predominance according to a publication of 2007. LN lesions are generally single and localized to the lower limbs, often bilateral and on tibial margin and instep. Variable sizes can exist, from the size of a small coin to extensive lesions over almost the entire area of the leg, from the knee to ankle. These lesions are described exceptionally on the face, scalp and trunk.

These lesions are often difficult to treat, leaving frequently significant scars. The course is most often chronic, even if spontaneous regression is observed in 20% of cases. And can be complicated by ulceration in 35% cases.

According to the bibliographic research, we found a single case reported in Lebanon of lipoid necrobiosis revealing type 1 diabetes in 2012. Which makes our case quite rare .

## **Conclusion:**

Despite years of research, the origin of lipoidal necrobiosis remains unknown. Once installed, it remains difficult to take care of. The relationship between NL and diabetes continues to be studied. Fortunately, the incidence of NL has become very low in patients with diabetes. This should probably be seen as a consequence of the constant improvement in the care of diabetic patients.

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## Multifocal Infantile Hemangiomas with Liver Involvement in an Extremely Preterm Infant - Case Report

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## **Introduction & Objectives:**

Multifocal cutaneous infantile hemangiomas (IH) are more commonly observed in infants born with very low/extremely low gestation and birth weight. They pose a risk of involvement with internal organs, most commonly the liver.

Case report: We present the case of an extremely preterm male infant, born after 26 gestational weeks, with a birth weight of 940g. At the postnatal age of 43 days, the first infantile IH was observed on his upper arm. This IH involved both superficial and deep tissues, exhibiting three bright red irregular-oval surfaces located closely to each other, sharing a common deep tissue base. It was classified as a segmental type. Two weeks later, an oval, bright red IH appeared at the base of the second/third right toe, along with a dot-like IH near the right nipple. Ten days after that, dot-like IHs were noticed under the left nipple, on the left thigh, and along the edge of the mandible on the left side. In total, six IHs were present, with one classified as segmental and the remaining five as superficial types. Abdominal ultrasound at admission, as well as after the first IH appeared, was inconspicuous. However, an abdominal ultrasound performed approximately two weeks after the emergence of all six IHs revealed a significant number of clearly defined hypoechoic oval changes in the liver parenchyma within which central and peripheral flows were detected on Doppler. The largest of these changes had a diameter of 8x8mm. The diagnosis of diffuse hepatic hemangiomatosis was established. The extreme prematurity, severe bronchopulmonary dysplasia, and the need for operative treatment of an incarcerated inquinal hernia delayed the initiation of systemic propranolol therapy until the age of 8 months. At that point, all IHs were only slightly larger than at presentation. Exception was the first one, in which all three superficial parts merged into a large IH over the entire upper arm, fortunately without signs of ulceration. Oral propranolol was introduced in hospital setting, with a gradual increase to the target dose of 2mg/kg and monitoring of possible side effects. Three months later, there were no signs of hepatic hemangiomatosis on abdominal ultrasound, and the cutaneous IHs showed signs of regression. At the time of writing this paper, the infant is 15 months old and propranolol therapy is still ongoing.

## **Conclusion:**

Multifocal infantile hemangiomas, particularly if there are more than five of them, carry a significant risk of internal organ involvement, primarily affecting the liver. This risk is unrelated to the visual characteristics of the IH, emphasizing the necessity for a thorough examination of the entire body in infants with IH. In extremely premature infants, the initiation of systemic therapy with oral propranolol, despite its indications, is frequently delayed due to associated morbidity.



## Dynamics of the clinical manifestation and LoScAT index during therapy in morphea patients

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## **Introduction & Objectives:**

Morphea is an autoimmune disease of the connective tissue, which is based on a cascade of immune reactions, including autoaggression, metabolic disorders of the extracellular matrix, as well as damage to endothelial cells. Morphea typically presents in childhood with an estimated annual age and sex adjusted incidence rate of 1–3 per 100,000 children and prevalence of 2 per 1,000 children. The main aim of the study was the investigation of treatment efficacy of morphea in children by evaluating LoScAT index.

#### **Materials & Methods:**

In order to optimize the treatment tactics for morphea in children and evaluate its effectiveness, all patients were divided into 2 groups: 1st – control group – 30 patients who received basic therapy; Group 2 – 38 patients; who in addition to the basic therapy, received the papain enzyme and 0.03% tacrolimus ointment. All patients were randomized by gender and age, duration and severity of the disease. Localized Scleroderma Assesment Tool (LoScAT index) applied for evaluation of activity, severity and damage of the process.

## **Results:**

In all groups, the linear form of morphea predominated in 53.3% and 57.9% of cases, and in the overwhelming majority, the duration of the disease ranged from 1 year to 5 years. A decrease in the phenomena of edema and erythema against the background of basic therapy for group 1 with preservation of immune indicators of inflammation and the presence of a rim of inflammation around individual lesions indicates the insufficient effectiveness of traditional therapy.

The lesions acquired a whitish tint, surrounded by a pinkish-lilac rim, although the intensity of the color decreased. Confirmation is a decrease in the level of erythema by 1.3 times (P<0.05) relative to the initial values; in the 2nd group of subjects, erythema decreased by 2.1 times, respectively.

At the stage of sclerosis, we observed a decrease in density and softening of the foci, and some disappearance in the peripheral zone of the lilac ring. During basic therapy, the compaction of lesions decreased by 1.1 times (from  $2.43\pm0.05$  to  $2.2\pm0.04$ ) (P<0.01) relative to the initial values, and in the second group by 1.4 times (from  $2.34\pm0.05$  to  $1.68\pm0.05$ ). The appearance of new lesions in group 1 decreased by 1.4 times (from  $1.11\pm0.07$  to  $0.8\pm0.06$ ), while in group 2 by 3.62 times, respectively (from  $0.87\pm0.03$  to  $0.24\pm0.04$ ).

In the dynamics of observation according to the LoSDI clinical index in the atrophy stage, traditional therapy turned out to be ineffective. In the case of the linear form of OS, the effectiveness of traditional therapy turned out to be weak, since despite the softening of the areas of compaction, we did not observe their complete recovery.

#### **Conclusion:**

Thus, complex therapy for the clinical manifestations of morphea over time according to clinical indices LoScAT was more effective than traditional therapy. There is an improvement in the clinical process with the achievement in all cases of stabilization of the scleroderma process, and in some cases its regression, a stable reduction in scleroderma foci is achieved.



## Accropustulosiss of infancy in yemeni infants and children's

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## **Introduction & Objectives:**

Infantile acropustulosis is a recurrent, selflimited, pruritic, vesicopustular eruption of the palms and the soles occurring in young children during the first 2-3 years of life. Newly described in 1979, it is probably much more common than the scarcity of reports would imply

Objective:- The objective of the study was to identify the

pattern of pruritic vesicopustular skin eruptions in Yemeni

## **Materials & Methods:**

Twenty five male and females Yemeni infants patients 1 to 3 years old presented with pruritic erythematous macules or papules that progress into vesicles and then pustules in the palms, the soles, and the lateral surfaces. Lesions may occur on the dorsal aspects of the hands and the feet as well as the trunk, the scalp, and the face. The intensity and the duration of attacks diminish with each recurrence. No other organ systems are involved. They treated with topical Betamethasone cream or ointment and systemic antihistamine. The skin biopsy followed by histopathological examination was not specific.

#### **Results:**

The clinical data and the investigations showed that the 25 Yemeni infants had acropustulosis.

## **Conclusion:**

Acropustulosis of infancy in Yemeni infants is very common skin disorder. The bad hygiene may play an important role in the etiology or allergic substances. It is not recurrence.



## Does COVID -19 vaccine induce BASCULE syndrome?

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**Introduction & Objectives:** Bier anemic spots, cyanosis and urticaria-like eruption (BASCULE) syndrome is an underreported paediatric vascular disorder from the group of acrosyndromes. Most cases are adolescents between 12 and 19 years of age or newborns in the first few months of life. The skin lesions usually appear in the lower limbs after prolonged standing and resolve in a few minutes by lying down or walking.

Materials & Methods: A 14 year-old woman presented with a painful and pruritic erythematous and cyanotic rash, mainly located on her lower limbs. The lesions had appeared 8 months earlier, after receiving her second dose of Pfizer COVID-19 vaccine a few weeks ago. Past medical history included hereditary coagulation factor VII deficiency (normal hemostasis range). The lesions typically appeared on the lower extremities acutely after prolonged standing, resolving rapidly by lying down or walking. Upper extremities and hands were affected occasionally. Physical examination, after a few minutes in a standing position, showed multiple hypocromic macules and urticarial papules with a surrounding erythrocyanotic background on the bilateral legs and thighs. Laboratory tests, including a complete blood count, basic metabolic, hepatic and renal panel, C-reactive protein, thyroid studies, antinuclear antibody and IgA antitransglutaminase antibody, were unremarkable. The patient was referred to paediatric cardiology for an active standing test in order to exclude postural orthostatic tachycardia syndrome and other forms of orthostatic intolerance. Heart rate, blood pressure, electrocardiogram and echocardiogram were normal.

**Results:** Bilastine 20 mg twice daily was started and she achieved nearly complete resolution of pruritus while taking the drug, but recurred when the dose was halved. Additional preventive measures were recommended, such as physical exercise, healthy diet and elevating the legs when possible.

Conclusion: The pathogeneses of BASCULE syndrome remains unknown. An exaggerated response to decreased arterial oxygen saturation due to venous pooling induced by orthostatism appears to be the trigger of erythrocyanosis and Bier anemic spots, while urticaria-like eruption may result from mast-cell degranulation in response to hypoxia. SARS-COV2 infection has been associated with a variety of skin lesions, including pernio-like changes ("COVID-toes") and peripheral acrocyanosis. The pathophysiological mechanisms are not clearly elucidated, but it is well known that COVID-19 virus and COVID-19 vaccines promote a proinflammatory state that facilitates microvascular thrombosis and intussusceptive angiogenesis. Both may trigger Toll-like receptors 7/8 inducing type 1 interferons production and others proinflammatory cytokines. We present a case of acute-onset BASCULE syndrome following prior COVID -19 vaccination. Berrebi D et al published a case of acute-onset BASCULE syndrome following prior asymptomatic SARS-COV-2 infection. Although they assumed that this finding may very well be coincidental, this case uncovers a plausible biological mechanism by which post-COVID endothelial dysfunction and exaggerated arteriolar constriction trigger or exacerbate BASCULE syndrome in individuals with pre-existing vascular hypersensitivity. The same explanation may be attributed to our case. Therefore, association between BASCULE syndrome and COVID-19 warrants further exploration.



# Eccrine Squamous Syringometaplasia in a 9-year-old child receiving chemotherapy and CAR-T Cell therapy for acute lymphoblastic leukaemia

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## **Introduction & Objectives:**

Eccrine squamous syringometaplasia (ESS) is a histologically distinct cutaneous eruption occurring predominantly in acral or intertriginous areas. Its presentation in the paediatric population is rare. We describe a case of ESS in a 9- year-old girl receiving chimeric antigen receptor (CAR)-T Cell Therapy for Early T cell precursor acute lymphoblastic leukaemia (ETP-ALL).

#### **Observation & Results:**

A 9-year-old Chinese girl was diagnosed with refractory early T cell precursor acute lymphoblastic leukaemia (ETP-ALL). She had undergone multiple previous lines of chemotherapy and was subsequently commenced on the following combination chemotherapy: Venetoclax, cyclophosphamide, mercaptopurine, cytarabine and methotrexate. However, due to the aggressive nature of her disease, she was also started on CAR-T cell transfusion – a form of immunotherapy using a patient's own genetically modified T cells to bind to and kill cancer cells.

She developed a tender cutaneous eruption which began on day 11 of CAR-T Cell Therapy. On examination, there were several tender erythematous papules over bilateral wrists, elbow, shins and face. A skin biopsy was performed. Histopathology revealed superficial perivascular infiltrates of lymphocytes and histiocytes in the dermis, squamous metaplasia of the cuboidal epithelial cells of the eccrine ducts and an absence of neutrophils - this was consistent with the diagnosis of ESS. The rashes were self-limiting and spontaneously resolved with no intervention over 15 days.

There exist very few reports of ESS in the paediatric population. Although its pathophysiology is poorly understood, it is thought to occur due to toxic accumulation of chemotherapeutic agents in the eccrine glands. Jimena A. Miranda et al describes a case of ESS in an elderly gentleman presenting with a painful cutaneous eruption of the hands after receiving CAR-T cell therapy.

#### **Conclusion:**

Although ESS has been reported sparsely in the paediatric population, to our knowledge, this is the first paediatric case of ESS in a patient undergoing both chemotherapy and CAR-T cell therapy, a novel cell-based immunotherapy that has recently gained traction in the treatment of refractory malignancies.

Due to the novelty of CAR-T Cell Therapy, little has been reported on its associated cutaneous adverse reactions. The underlying mechanisms of CAR-T cells cutaneous toxicity remain uncertain. One possible cause would be the cross-reactivity of CAR-T cells with normal tissue expressing similar antigen to that of the target antigen on malignant cells.



## Myofibroma of infancy and its mimickers.

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## **Introduction & Objectives:**

#### **Materials & Methods:**

#### **Results:**

A female newborn born via the natural term spontaneous vaginal delivery presented a congenital purple-red ulcerated and indurated plaque of the foot, that was initially thought to be secondary to pressure. Ten days later, the patient was referred to the dermatology department because of the persistence of the lesion. Dermoscopy revealed a central erosive-crusted area surrounded by thick blood vessels, with the presence of white homogenous structures, located on the pink structureless background. The baby underwent an ultrasound examination which revealed a wide subcutaneous irregular echoic alteration with non-definite margins reaching the bone and infiltrating the hallux muscles. Doppler revealed intense vascular activity and suggested a diagnosis of a high-flow vascular anomaly. Subsequently, a cutaneous biopsy was performed, and based on histopathology, a final diagnosis of myofibroma of infancy was made.

Myofibroma of infancy (MOI) is a rare benign tumor of mesenchymal origin which is, at the same time, the most common fibromatous tumor in childhood. It occurs more frequently in males, unlike our patient, and about 50% of cases are already present at birth, occasionally it has been reported in adults. Its various clinical presentations pose diagnostic challenges which may delay the proper management. MOI may present as a nodule or papillomatous or atrophic lesion that can be deep or rather superficial, mobile or fixed. In our patient, the lesion was an infiltrated purple-red plaque with an erosion on the surface. The localization is usually head, neck or trunk, whereas in our case it was a foot. Noteworthy, MOI can be solitary or multiple and may also appear in the internal organs, which results in a worse prognosis and may be fatal. Diagnostic procedures should include skin biopsy with histopathological examination, ultrasound examination, magnetic resonance imaging, and radiography.

Probably myofibroma is most commonly mistaken with infantile hemangioma. Other potential differentials are pressure ulcers, rhabdomyosarcoma, fibrosarcoma, or complications after venous catheter implantation.

As for the management, most lesions located within the skin or subcutaneous tissue resolve spontaneously so they can be periodically monitored. If treatment is desired due to aesthetic reasons or dysfunction caused by the lesion, solitary myofibroma should be surgically excised. However, if the lesions are multiple or internal organs are involved, surgical intervention, as well as chemotherapy can be administered.

## **Conclusion:**



## Case report of Langerhans cell histiocytosis in a 4-month-old infant

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**Introduction & Objectives:** Langerhans cell histiocytosis (LCH) represents a group of rare idiopathic disorders characterised by the aberrant proliferation and neoplastic spread of Langerhans antigen-presenting cells. Clinical manifestations of LCH vary significantly, from benign, self-resolving forms to severe, life-threatening with multisystem involvement. LCH predominantly emerges from birth up to the age of three, initially presenting through skin manifestations.

#### **Materials & Methods:**

A 7-month-old girl presented to the hospital with fever, respiratory distress, and a rash characterised by nearly confluent erythematous macules and papules with overlying yellow crusts, petechial, and purpuric lesions alongside fragile, split nails.

History reveals that the rash first occurred at the age of 4 months. The child was repeatedly consulted by paediatricians and dermatologists, diagnosed with lactose intolerance (laboratory confirmed), clinically diagnosed with "molluscum contagiosum", "atopic dermatitis", 'seborrheic dermatitis', "food allergy", "scabies" and routine treatments were given with no significant improvement.

Rash characteristics and history suggested LCH, confirmed by skin biopsy and immunohistochemical testing, showing positive CD1a (+) and Langerin (12D6) (+) antigens.

Further evaluation with a PET-CT scan identified multiple granulomas in the skin and soft tissues of the neck, lungs, and spleen. The infant was then referred to an oncologist and treated according to the LCH-III protocol established by the Histiocyte Society, involving daily oral prednisolone and weekly vinblastine dosed according to body weight. As the general condition deteriorated, a second-line chemotherapy regimen of methotrexate and leucovorin was initiated. Despite these interventions, the child's condition worsened, leading to sudden cardiac arrest and a fatal outcome.

## Results:

In this case, the disease initially manifested as a nonspecific rash and nail damage. Upon hospital admission, the patient presented with a widespread condition impacting both soft tissues and parenchymal organs, resulting in respiratory failure and impaired vital functions. The diagnosis was speculated and verified through clinical assessments, bolstered by a skin biopsy and immunohistochemical studies. Despite applying primary and secondary chemotherapy protocols following standard treatment recommendations, the treatments were ineffective.

**Conclusion:** LCH is characterised by uncertain and complex clinical features, often leading to misdiagnosis and delayed commencement of appropriate therapy, thus diminishing the prospects for effective treatment. Skin biopsy and referral for consultations with other specialists are crucial for precise diagnosis and treatment of skin lesions unresponsive to standard therapies.



## Mid-face toddler excoriation syndrome: Case report

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#### Introduction:

Mid-face toddler excoriation syndrome (MiTES) is a new condition described in 2017 in three elderly children between 11 months and 4 years. Clinically, the children presented with excoriations and self-inflicted ulcerations from repeated scratching of the centrofacial region. The genetic mutation causing MiTES is located on the PRDM12 gene in the majority of reported cases.

We report a new case of MiTES in a 15-month-old female infant.

#### **Materials:**

An 15 -month-old female infant, born to non-consanguineous parents, presented with itchy lesions on her face, manifesting since the age of 3 months. Skin examination revealed two ulcerations topped with blackish crusts, erosions and depigmented atrophic scars were seen in the centrofacial area. During hospitalization, we observed that the lesions were caused by repeated scratching while awake and during sleep. Neurological and paedopsychiatric examination were normal.

## **Results:**

Blood tests showed anemia and thrombocytosis with normal serum uric acid. Biopsy showed non-specific inflammatory infiltrate. Nerve conduction studies and magnetic resonance imaging brain were normal. The diagnosis of MiTES was based on the centrofacial topography of the lesions the self-inflicted nature of the lesions and the absence of arguments in favor of another etiology. We prescribed directed healing and limiting scratching during sleep by bandaging the hands, and cognitive-behavioral therapy. After two months, almost complete healing of the lesions was noted.

## **Conclusion:**

MiTES is a rare pediatric syndrome, recently described1. Only 13 cases have been reported in literature. Before making this diagnosis, it is important to rule out infectious origin, pathomimia, Lesch-Nyhan syndrome and neurotic excoriations. In our case, all these etiologies were ruled out. Some authors consider MiTES as an extremely localized form of congenital insensitivity to pain.

Any self-inflicted centrofacial ulceration in a child should be considered as a diagnosis of MiTES.



## Skin Disorders Encountered in a Maltese Pediatric Emergency Department - A Prospective Observational Study

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# SKIN DISORDERS ENCOUNTERED IN A MALTESE PEDIATRIC EMERGENCY DEPARTMENT - A PROSPECTIVE OBSERVATIONAL STUDY

**Introduction & Objectives:** This study was done to evaluate the range, frequency and management of pediatric dermatology presentations to Malta's Pediatric Emergency Department (PED).

**Materials & Methods:** This study was carried out prospectively over one year (May 2021-April 2022), assessing presentations to the PED during the first week of the 12 calendar months in this period. Data was gathered from nursing triage notes for patient demographics, time and date of presentation, rash description, presence of fever, diagnosis, treatment given, need for admission or otherwise, follow-up and physician seniority. Purely traumatic cases were excluded. Data was compared to that from similar studies overseas.

**Results:** Dermatological cases accounted for 6.74% of all presentations. The commonest diagnoses were viral exanthems (26.1%), of which 63.5% were not further specified, followed by inflammatory (19.9%) and infectious skin disorders (11.2%). There was no seasonal variation in the overall frequency of dermatological PED presentations throughout the year, however, viral exanthems were commonest in summer and autumn, with inflammatory dermatoses commoner in winter and spring. Of all dermatology cases, treatment was administered at PED in 17%, and 12.4% needed hospital admission. Follow-up was given to 14.1%, of which 17.6% were to Dermatology.

**Conclusion:** This study confirms the frequency of dermatological presentations to PED and demonstrates interesting seasonal variations.



## Pediatric Case of Miescher's Cheilitis Treated with Clarithromycin

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## **Introduction & Objectives:**

Melkersson-Rosenthal syndrome is a rare condition classically characterized by orofacial edema, furrowed tongue, and peripheral facial paralysis. Miescher's cheilitis or granulomatous macrocheilitis represents the monosymptomatic form of this syndrome. It is rare in childhood, more common in young adults, and its etiopathogenesis remains poorly understood. We report a pediatric case of monosymptomatic granulomatous cheilitis treated with clarithromycin.

#### **Materials & Methods:**

A 7-year-old patient with no significant medical history, particularly no history of atopy, presented with macrocheilitis evolving for 3 months. History and clinical examination revealed no cough or exertional dyspnea, no abdominal pain or diarrhea. A lip biopsy was performed, concluding with a diagnosis of Miescher's granulomatous cheilitis. Infectious cause, sarcoidosis, and Crohn's disease were ruled out before confirming the diagnosis. The patient was treated with clarithromycin 15mg/kg/day in two divided doses for 10 days per month, with partial improvement noted after 2 months of follow-up. The treatment will be continued for other 4 months with regular check-ups.

## **Results:**

Granulomatous cheilitis was first reported by Miescher in 1945. It can be isolated or associated with orofacial edema, furrowed tongue, and facial paralysis, constituting Melkersson-Rosenthal syndrome. The etiopathogenesis of this condition is still poorly understood, but several observations suggest an immunological origin. Miescher's cheilitis is rare in childhood, more common in young adults. Clinically, it presents as lip swelling, evolving in flares, often asymmetrically affecting the lips. The swelling is firm and elastic, sometimes associated with erythema. Epithelioid and giant-cell granulomas are mainly found in skin biopsy, without central caseous necrosis. Witch can also be found in sarcoidosis and Crohn's disease.

The treatment of Miescher's granulomatous cheilitis is challenging. Proposed treatments include oral corticosteroids at a dosage of 0.1 to 1mg/kg, doxycycline and intralesional corticosteroid injections.

Other therapies have been used, including clofazimine, colchicine, minocycline, hydroxychloroquine, metronidazole, dapsone, and macrolides. It is important to note that recurrences are common even with combined treatment. Clinical monitoring is therefore necessary as macrocheilitis can precede Crohn's disease digestive manifestations by several years.

#### **Conclusion:**

Granulomatous cheilitis is a rare condition, considered a monosymptomatic form or an incomplete variant of Melkersson-Rosenthal syndrome. Its diagnosis requires lip biopsy revealing the presence of epithelioid histiocytic granulomas with angiotropism. The treatment remains challenging and requires individualized adaptation for each case.



## Erythropoietic porphiria in a 4 year old boy: A case report

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## **Introduction & Objectives:**

Porphyrias are a rare group of metabolic disorders caused by defects in heme biosynthesis. Even though erythropoietic protoporphyria (EPP) is the most common type seen in children, it can be under-recognized. The reason for EPP is mutation of the ferrochelatase (FECH) gene localized to 18q21. EPP typically presents in early childhood with pain and crying upon exposure to bright sunlight, followed by erythema, edema, and itching. Repeated exposure can lead to loss of lunulae of the fingernails, ecchymoses, and petechiae. It is a lifelong disease, mild anemia may be noted, but the most serious complication is cirrhosis of the liver and liver failure. In children with EPP, the risk of gall stones is increased and their occurrence should prompt evaluation for EPP. The diagnosis is set by increased levels of protoporphyrin without increased levels of coproporphyrin, and can be confirmed by genetisc.

## **Materials & Methods:**

We present the case of a boy with painful erythematous macules and purpuric lesions, which first appeared on photo-exposed skin in the spring, at the age of 4 years.

#### **Results:**

In July, a 4-year old boy came to our clinic because of red, painful, skin changes, that have appeared on back of his hands, ears and cheeks 3 days ago. His mother had noticed similar redness already in spring time, but later disappeared on its own. Otherwise he had operation of ventral septal heart defect in first weeks of his life, is regularly checked by cardiologist and has no regular systemic therapy. There was no family history of skin diseases.

On the examination, coalescent red to livid macules with purpuras, symmetrically on the cheeks, ears and dorsal parts of the hands have been noticed. We performed additional blood tests, where total protoporphyrin level was elevated, CBC, renal function and hepatic enzymes were within normal limits, there was no anaemia. With genetic testing two pathogenic variants in FECH gene were found and a recessive type of EPP was confirmed. At gastroenterologist ultrasound of the abdomen was normal, there was no sign of hepatic disease or gallstones. We recommended the use of sun protective clothing, sunglasses and sunscreens containing mineral filters and since then no skin changes have reappeared. At regular check-ups by gastroenterologist there was no sign of hepatic disease till now.

#### **Conclusion:**

EPP is rare inherited disease and proper sun protection is the mainstay of the treatment. Parents and patients must be educated about the disease, be aware of all the precautionary methods to protect the skin, advised to avoid unnecessary sun exposure and hepatotoxins. Regular monitoring of liver function, porphyrin levels and complete blood count is necessary to recognise and treat possible complications of the disease. Children with EPP may be misdiagnosed as primary angioedema or allergic reaction, so awareness of the disease is important and hopefully, also our case will help in recognising patients with EPP.



## Rothmund-Thomson syndrome associated with cutaneous calcinosis

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<sup>1</sup>Algeria, algiers, algiers

#### Introduction:

Rothmund-Thomson syndrome (RTS) is a rare autosomal recessive genodermatosis (300 cases have been reported), characterized by poikiloderma associated with other dermatological and extradermatological lesions. We report a new case associated with cutaneous calcinosis lesions, illustrating the rich symptomatology that can be encountered in this syndrome.

## **Case report:**

An 11-year-old girl from a 2nd-degree consanguineous marriage was referred to us with poikilodermal lesions of the face and hands since the age of 2 years. The lesions began as erythema of the cheeks, hands and buttocks, and progressively developed into poikiloderma. Clinical examination revealed dark-red erythematous lesions on the face, dotted with small hypopigmented spots and fine telangiectasias. On the hands and buttocks, there was a poikilodermal appearance with erythema, atrophy and hypo- and hyperpigmented lesions (Fig. 1 and 2). On the left knee, multiple nodules 1 to 3 cm in diameter, hard and painful on deep palpation, with emission of a hard white substance corresponding to cutaneous calcinosis (Fig. 3 and 4). There was also pain on palpation of the left femur. An osteosarcoma test was negative. In view of this clinical and evolutionary picture, the diagnosis of RTS associated with cutaneous calcinosis was retained.

#### **Discussion:**

RTS is characterized by poikiloderma, photosensitivity associated with short stature due to pre- and postnatal growth retardation, sparse hair, sparse or absent eyelashes and eyebrows, early cataracts, skeletal abnormalities and an increased risk of developing malignant skin and bone tumors. The skin is usually normal at birth, but cheek erythema appears between the 3rd and 6th month of life, spreading to the extremities and eventually the buttocks. The trunk and abdomen are generally spared. In our case, the diagnosis of RTS was based on the clinical history and the presence of characteristic skin lesions. In case of doubt, molecular genetic testing can be used to confirm the diagnosis. Our patient's cutaneous calcinosis is an unusual cutaneous manifestation in Rothmund-Thomson syndrome. Exceptional cases of cutaneous calcinosis have been reported, which makes our observation special. Multidisciplinary management is essential to improve patients' quality of life.

#### **Conclusion:**

We report an additional case of the Rothmund-Thomson syndrome, original by its association with cutaneous calcinosis. This observation seems to confirm the variable expression of RTS.



# Dupilumab improves disease severity in children <12 years of age with severe atopic dermatitis: Interim Results from PEDISTAD Registry

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**Introduction & Objectives:** In phase 3 studies, dupilumab significantly improved disease severity in children with moderate-to-severe atopic dermatitis (AD). This study assesses the impact of systemic treatment on children with severe AD in real-world treatment settings.

**Materials & Methods:** PEDISTAD (NCT03687359) is an ongoing, international, longitudinal, observational 10-year registry study in patients aged 6 months to 11 years with moderate-to-severe AD at enrollment, whose disease is not adequately controlled by topical prescription therapies or for whom those therapies are medically inadvisable. This interim analysis assessed the effect of dupilumab on patient-reported disease severity and quality of life (QoL) among children with severe AD using Patient-Oriented Eczema Measure (POEM) and Children's Dermatology Life Quality Index (CDLQI). Overall safety was also evaluated.

**Results:** A total of 84 patients with severe AD received dupilumab. The mean ( $\pm$  SD) POEM score decreased over time from 20.2  $\pm$  6.7 at therapy start to 11.5  $\pm$  7.7 at 3 months, and 8.7  $\pm$  7.7 at 12 months. Similarly, the mean ( $\pm$  SD) CDLQI score decreased with dupilumab use, from 15.0  $\pm$  6.8 at therapy start to 9.7  $\pm$  7.7 at 3 months, and 8.5  $\pm$  7.0 at 12 months. About a 1/3 (29.8%) of patients had adverse events.

**Conclusion:** Dupilumab significantly improved frequency of symptoms and QoL in children aged 6 months to 11 years with severe AD in real-world daily practice.



# Dupilumab treatment provides long-term improvement in sleep in pediatric patients with moderate-to-severe atopic dermatitis over 1 year

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**Introduction & Objectives:** To evaluate the proportion of pediatric patients achieving and maintaining mild or no sleep loss across 5 visits during a 1-year open label extension trial of dupilumab.

Materials & Methods: Patients who previously participated in 16-week trials and were aged 0.5–5 years (LIBERTY AD PRESCHOOL; NCT03346434), 6–11 years (LIBERTY AD PEDs; NCT03345914), and 12–17 years (LIBERTY AD ADOL; NCT03054428), were subsequently enrolled in the phase 3, open-label extension trial, LIBERTY AD PED-OLE (NCT02612454). Patients were treated with 300 mg q4w or 200/300 mg q2w (body weight <60 or ≥60 kg, respectively). In this analysis, patients with a SCORing Atopic Dermatitis (SCORAD) sleep loss VAS score (0-10 over the last 3 days) of greater than 4 at OLE baseline, were assessed for the maintenance of SCORAD sleep loss VAS lower than 4, at 5 timepoints: Weeks 4, 16, 28, 40, and 52.

**Results:** In 763 patients, 266 patients with a SCORAD sleep loss VAS score of greater than 4 were assessed. Mild or no sleep loss was achieved in at least 4 of 5 timepoints in more than half of patients aged 0.5–5 years (56/90; 62%), 6–11 years (60/96; 63%), and 12–17 years (50/80; 63%). Across these age groups, over 75% maintained this response for at least 3 of 5 timepoints. Safety was consistent with the known dupilumab safety profile in patients with atopic dermatitis.

**Conclusion:** Most pediatric patients achieved an improvement in sleep, which was maintained during 1 year of treatment with dupilumab. Results were consistent for infants/preschoolers, children, and adolescents.



## Nail fold capillary changes in children with connective tissue diseases - A single center study

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## **Introduction & Objectives:**

Connective tissue diseases (CTDs) such as Systemic sclerosis (SS), dermatomyositis (DM), mixed connective tissue disease (MCTD), systemic lupus erythematosus (SLE) are frequently associated with nail fold capillary changes. Nailfold capillaries are involved early in the course of CTDs aiding in early diagnosis. Nail fold capillary changes are believed to reflect the microvascular abnormalities and these correlate with disease activity in CTDs. These changes can be easily visualized using a dermoscope. Videodermoscope is a simple and non-invasive tool. However, there is paucity of data on nailfold capillary changes in CTDs in the pediatric population. The objective of this study was to assess the nail fold capillary changes in children with CTDs using a videodermoscope.

#### **Materials & Methods:**

A hospital based cross-sectional, observational study was undertaken over a period of 16 months from January 2021 to June 2022. Children with a suspected clinical diagnosis of a CTD attending the Dermatology Out Patient Department and Pediatric Rheumatology Clinic in a tertiary care hospital in North India were diagnosed on the basis of standardized criteria and were included in the study after obtaining written parental consent. Nail fold capillaries were assessed in each patient in the proximal nail folds of 4th and 5th fingernails of both hands using a videodermoscope (Dinolite AM7515MZT) and a laptop. Images were recorded and analysed.

## **Results:**

A total of 100 children with a confirmed diagnosis of a CTD in the age group ranging from 6 months-18 years were included in study group. Our study included 63 % juvenile idiopathic arthritis (JIA) patients, 17% juvenile dermatomyositis (JDM) patients, 6% Kawasaki disease (KD) patients, 3% SS patients and 2% Henoch Schonlein purpura (HSP) and MCTD patients each. Nail fold capillary changes were present in 73% patients. Common nail fold capillary change were decreased nail capillary density (65%), capillary drop outs (63%), dilated capillaries (59%), avascular areas (47%) and meandering capillaries (46%). Mean Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) was significantly higher (p<0.05) in children with SLE having tortuous and meandering capillaries. Mean Juvenile Arthritis Disease Activity Score 27 (JADAS 27) was significantly higher (p<0.05) in patients with JIA having decreased nail capillary density, giant capillaries, dilated capillaries, meandering capillaries and capillary dropouts.

#### **Conclusion:**

Nail fold capillary changes are commonly seen in patients with CTDs and some of the changes can be associated with disease activity in CTDs. Videodermoscope is an easy and a non-invasive tool which can aid the physician to make an early diagnosis. It can be used as a point of care tool as a part of disease activity score in patients with CTDs.



## Cutaneous adverse effects from diabetes devices in pediatric patients with type 1 diabetes, a systematic review

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## **Introduction & Objectives:**

Continuous glucose monitoring (CGM) and continuous subcutaneous insulin infusions (CSII) are the current standard treatment devices for type 1 diabetes (T1D) management. With a high prevalence of T1D beginning in pediatrics and carrying into adulthood, insufficient glycemic control leads to poor patient outcomes. Dermatologic complications such as contact dermatitis, lipodystrophies, and inflammatory lesions are among those associated with CGM and CSII, which patient compliance and glycemic control. This systematic review explores the current literature surrounding dermatologic complications of CGM, CSII, and the impact on patient outcomes.

#### **Materials & Methods:**

A systematic review of the literature was carried out using PRISMA 2020 guidelines utilizing PubMed, SCOPUS, EMBASE, Cochrane, and Web of Science databases. Included articles were those containing primary data relevant to human subjects and adverse CGM and CSII devices in pediatric populations, of which greater than 50% of the sample size were ages 0-21. Following the removal of duplicates, abstract screen, full-text appraisal, and application of exclusion criteria, 26 studies were analyzed and discussed.

#### **Results:**

Contact dermatitis is the most common complication, with thirteen identified studies. Lipodystrophies, such as lipohypertrophy and lipoatrophy, were discussed in six studies. Four studies cover nonspecific cutaneous changes, and the remaining three studies cover unique cutaneous such as granulomatous reactions and panniculitis. Many studies included the effect of these cutaneous changes on impacted decisions to discontinue these devices, and some patients reverted to manual insulin injection therapy and monitoring. Changes in glycemic control were sometimes included and yielded mixed results. Lipohypertrophy was identified as a contributing agent of worsened glycemic control.

#### **Conclusion:**

The dermatologic complications of CGM and CSII pose a potential risk to long-term glycemic control in T1D, both directly and indirectly through discontinuation. This consequence disproportionately affects pediatric patients, where discontinuation of device use can lead to long-term inadequate control. Increased manufacturer transparency is critical to reduce the incidence of contact dermatitis. Further studies are needed to expand upon long-term changes in glycemic control and the current preventative measures, such as device site rotation and steroid creams, which lack consistent effectiveness.

Cutaneous manifestation	Studies	Combined study N effected	% Discontinued use of insulin devices (per study)	Glycemic control outcomes	Quality of Study(s)*
Nonspecific cutaneous reactions	3	194	22% 4.3% Not included	Not included	2b (3)
Ultrasound determined subcutaneous changes	1	161	N/a	No effect of hyperechogenicity (indicator of lipohypertrophy) on HbA1c	2b (1)
Contact dermatitis	13	213	Discontinued use and changed brands (5 studies),  Not included (3 studies);  0.01%, 16%, 38.1% discontinued and did not change brands in remaining (3) studies	Not included (11 studies); non-significant changes in HbA1C (2 studies)	2b (7), 4 (6)
Lipohypertrophy	4	288	Not included	Significantly increased HbA1C in LH (2 studies), nonsignificant increase (1 study), significantly increased episodes of hypoglycemia (1 study)	2b (4)
Lipoatrophy	2	16	Insulin-induced, changed insulin types without improvement	Non-significant changes in HbA1C, not included 1 study	4 (1), 1b (1)
Granulomatous reaction	1	1	Not included	HbA1C rise from 7.2% to 12.5% following development of nodules	4(1)
Panniculitis	2	2	Multiple changes trialed and failed	Not included	4 (2)

\*From the Centre for Evidence-Based Medicine (2b; cohort study, 4; case report/series)



## Epidermolysis bullosa acquisita pseudo-cicatricial pemphigoid in a child

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## **Introduction & Objectives:**

Epidermolysis bullosa acquisita (EBA) belongs to the group of subepidermal autoimmune blistering dermatoses. It is a rare pathology linked to the production of autoantibodies against type VII collagen. Pediatric cases are rarely reported in the literature. We present an atypical case with severe mucosal involvement.

#### **Materials & Methods:**

A 9-year-old girl presented with a pruritic mucocutaneous bullous eruption. Questioning revealed recurrent oral erosions for more than 6 months. Clinically, there were tense bullae resting on erythematous skin, located on the trunk, the extension areas of the limbs and the palmoplantar surfaces. Nikolsky's sign was negative. Oral (jugal and palatal erosions) and ocular (bilateral symblepharon and Meibomian gland dysfunction) mucusal damage was severe. The genital mucosa was unaffected. The biology work-up was unremarkable. Skin histology showed a bullous detachment with linear deposits of IgG and C3 along the dermal-epidermal junction in direct immunofluorescence (DIF). Indirect immunofluorescence (IIF) on cleaved skin showed the presence of anti-basal membrane antibodies on the dermal side. Systemic treatment with prednisolone 1 mg/kg/day combined with dapsone 2 mg/kg/day was started, resulting in rapid skin remission with the appearance of grains of melium. Mucosal involvement improved more slowly.

## **Results:**

EBA is rarely seen in children. Two distinct clinical presentations have been described: the classic or mechanobullous form, characterized by cutaneous fragility and bullae at sites of trauma, and the inflammatory form, mimicking any other subepidermal autoimmune blistering dermatoses. The diagnosis of EBA in its bullous pseudopemphigoid variant was made in our patient on clinical grounds (severe pleural-mucosal involvement, moderate cutaneous involvement with development of milium grains), histological grounds (bullous detachment) and immunological grounds (linear deposits of IgG and C3 in DIF, positivity of anti-basal membrane antibodies on the dermal side of the dermo-epidermal junction in IIF of cleaved skin). The Elisa anti-collagen VII test could not be performed. Meibomian gland dysfunction has been described in hereditary epidermolysis bullosa, more rarely in EBA. The treatment of EBA is not codified. A number of immunosuppressive, anti-inflammatory and anti-neutrophilic treatments are proposed. Dapsone, alone or in combination with systemic corticosteroids, is often the first-line treatment. In cases where the ocular prognosis is compromised, ciclosporin is recommended..

## **Conclusion:**

We report a rare case of pediatric EBA with pseudo-cicatricial pemigoid presentation, illustrating the great heterogeneity of clinical forms of this condition.



## Major and minor risk factors for severe corticophobia among parents of children with atopic dermatitis

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## **Introduction & Objectives:**

Corticophobia, the fear of applying topical corticosteroids (TCS), is a rising issue in industrialized countries. It involves erroneous beliefs and negative feelings about TCS, promoted by misinformation, leading to therapeutical nonadherence, despite the actual safety and effectiveness of TCS treatment. Aim of this study was to measure corticophobia among parents of children with atopic dermatitis (AD) and identify related risk factors.

## **Materials & Methods:**

Patients attending the Pediatric Dermatology Unit for skin examination in the last six months were screened for AD. All patients (0-18 years) with AD were included in the study. Demographic data (age, sex, disease onset, previous healthcare professional consultations, parental educational degree) were collected. AD severity was assessed with EASI: ≤21 was considered mild/moderate, >21 severe. Parents of AD patients completed the self-administered Topical Corticosteroid Phobia (TOPICOP) questionnaire, to measure corticophobia, and the Parental Dermatological Life Quality Index (DLQI) questionnaire. For each parent, TOPICOP score was calculated as a percentage (0–100% TCS phobia): A score ≤ 50% was considered mild/moderate, > 50% severe. A DLQI score ≤10 was considered mild/moderate, ≥11 severe.

#### **Results:**

Overall, 100 patients were included (53 females; 47 males; mean age 5.9 years). A mean EASI score of 19.7 was registered: 44 patients had mild/moderate AD, 56 severe AD. Of patients, 33 never consulted healthcare providers for AD, 67 did. Parental educational degree was low/intermediate in 60 cases, high (gymnasium or university degree) in 40. Parental DLQI scores ranged from 0 to 30 (mean value 10.7). Mean parental TOPICOP percentage was 39.1%: 51 had mild/moderate corticophobia, 49 severe corticophobia. Severe corticophobia was registered for parents of 17 very young (age ≤4 years) patients, 32 older (age >4 years) patients; 6 patients with severe AD, 43 patients with mild/moderate AD; 3 patients with late disease onset (after 1 year of age), 46 patients with early disease onset (prior to 1 year of age); 8 patients with no previous healthcare professionals consultations, 41 patients with previous consultations. Parental mild/moderate corticophobia was registered for 35 very young patients, 16 older patients; 38 patients with severe AD, 13 patients with mild/moderate AD; 20 patients with late disease onset, 31 patients with early disease onset; 25 patients with no previous healthcare professionals consultations, 26 patients with previous consultations. At logistic regression analysis, high parental DLQI (OR 38,5; p <0,0001) and high parental education (OR 4,1; p < 0,0338), accounted for major risk factors influencing severe parental corticophobia, as well as older age of patients (OR 14,5; p =0,0015), and early disease onset (OR 8,1; p<0,0513). At χ2 test, severe parental corticophobia was significantly associated with mild/moderate AD (p<0.001), and with previous healthcare professionals consultations (p<0.001), accounting for minor risk factors of severe parental corticophobia.

## **Conclusion:**

Assessing risk factors for severe parental corticophobia is essential to comprehend the origin of this complex phenomenon and to address especially groups of parents at higher risk for corticophobia with educational programs, to overcome their unfunded fears and ultimately augment treatment adherence and satisfaction and improve disease outcome of their children with AD.



## Long-term outcomes of pre-pubertal onset vulvar lichen sclerosus

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**Introduction & Objectives:** Vulvar lichen sclerosus (VLS) is a chronic inflammatory dermatosis affecting the vulvar and perianal skin. VLS has a bimodal age of onset, in pre-pubertal and post-menopausal age groups. Whilst in adults, VLS tends to run a chronic course, the long-term outcomes of pre-pubertal onset VLS is largely unknown. This study aims to establish the long-term disease activity, factors that influence disease progression, as well as the physical and psychological complications of paediatric VLS persisting into adulthood.

Materials & Methods: A cohort study conducted in a dermato-gynaecology practice involving women who were diagnosed with VLS pre-menarche. A retrospective chart review of 106 case records has been completed. The remaining data collection will occur prospectively through (i) study questionnaires, including quality of life as assessed by the Vulvar Quality of Life Index (VQLI), and (ii) in-person clinic appointments with a dermatologist.

**Results:** One hundred and six cases of VLS were identified, with a mean age of diagnosis of 7.2 years and a mean age of 20.6 years at time of recruitment. From chart review, 17.6% of participants never reached remission and 32.4% experienced recurrence after remission. Seventeen percent of participants were "adherent", 57.5% were "partially adherent" and 4.7% were "non-adherent" with the prescribed treatment. Although treatment adherence was generally good at time of diagnosis, this was observed to reduce from menarche and into early adolescence. The study questionnaires will further elucidate patterns in disease activity over time, psychological impacts and attitudes underpinning treatment adherence. The clinic appointments will assess for current disease activity and long-term sequelae including vulvar scarring.

**Conclusion:** This study will better inform clinicians of the long-term prognosis of VLS diagnosed pre-pubertally and how to provide optimised care for patients with pre-pubertal onset VLS, assisting to reduce the risk of disease progression and complications later in life.



## Infantile scabies misdiagnosed and treated with topical corticosteroids: Diagnostic wandering and complication

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## **Introduction & Objectives:**

Scabies is a common parasitic dermatosis. Clinical presentations of scabies in pediatrics are sometimes misdiagnosed and are therefore incorrectly treated with topical corticosteroids. We report a case of an infant mistakenly treated with topical corticosteroids complicated by adrenal insufficiency.

#### **Materials & Methods:**

A 3-month-old female presented during the first day of life with vesicular and eczematous lesions and was treated with potent topical corticosteroids for 2 months as an atopic dermatitis. During the medical history, the mother reported nocturnal itching starting from her last month of pregnancy. The current clinical examination of the infant revealed a cushingoid facies with diffuse vesiculopustular lesions also affecting the palmoplantar regions and the back, diffuse excoriated papules on the body and telangiectasias on the face. Specialized consultation was requested due to clinical suspicion of adrenal insufficiency. Scabies treatment was initiated for the infant and household contacts with good improvement on follow-up.

## **Results:**

Scabies in children is an underdiagnosed parasitic infestation due to its atypical clinical manifestations, leading to diagnostic delays. The main symptom is intense nocturnal itching, which can disrupt sleep. In infants, this symptom may be absent.

In this age group, there are several peculiarities to consider. Frequent findings include the presence of vesiculopustules, particularly on the palms and soles. Scabetic nodules are more common in infants.

The cutaneous side effects of topical corticosteroids are well known; however, their systemic effects are less understood. They can induce secondary adrenal insufficiency by suppressing ACTH secretion, clinically manifesting as cushingoid appearance, stretch marks and skin fragility. These signs can be subtle, posing a diagnostic challenge.

Although systemic effects have been observed in adults, they are particularly concerning in children due to a higher body Surface-area-to-volume ratio. Several factors influence the development of iatrogenic corticotrope insufficiency, including treatment duration, as the risk increases significantly beyond 3 months; the type of corticosteroid, as its plasma half-life determines the extent and duration of suppression of the HPA axis.

#### **Conclusion:**

The suspicion of scabies may arise during medical history when there is a familial history of itching. However, it is common for the child to be the first family member affected or the first to exhibit symptoms.

It is important to recognize the signs of scabies and treat it before initiating topical corticosteroids in a child. We also highlight the risk of adrenal insufficiency with abusive and non-standardized use of topical corticosteroids in the pediatric population.



## Kerion Celsi: a particular disorder in children with risk of scarring alopecia

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**Introduction & Objectives:** Kerion Celsi or inflammatory tinea capitis is a suppurative dermatophytic infection of the scalp that mainly affects children before puberty. It is more common in developing countries and characterized by a highly inflammatory clinical reaction, with a risk of scarring alopecia. The objective of our study was to determine the epidemiological, clinical and mycological features of Kerion Celsi in our context, and to highlight the urgency for appropriate management.

**Materials & Methods:** This is a descriptive and retrospective study of all hospitalized cases of confirmed inflammatory tinea capitis, between January 2013 and December 2023.\*\*

**Results:** Twenty-two cases were reviewed, showing a male predominance (sex ratio M/F= 2.5), and ages ranging from 2 to 16 years old (average of 7.3 years). Previous contact with animals was reported in 18 cases (81.8%), 8 patients (36.4%) had a similar case in their family, and the majority of patients had a rural origin (86.3%). The evolution period prior to consultation was an average of 1 month (15-60 days) of a suppurative, inflammatory and tense-elastic round mass, with pustules and crusted lesions, purulent secretions and alopecia. The scalp lesion was unique in 19 cases (86.4%) and multiple in 3 patients (13,6%). Lesion diameters ranged from 4 to 10 cm, with the main location being occipital (31.8%), followed by parietal (22.7%), temporal (22,7%), frontal (13.6%) and vertex (9%). Papulo-pustular skin lesions were reported (59.1%), as well as cervical lymphadenopathies (63.6%) and fever with altered general condition (54.5%). Prior to hospitalization, 4 patients were treated with oral antibiotics without improvement, due to a misdiagnosis. Mycological samples isolated *Trichophyton mentagrophytes* (7 cases), *Trichophyton violaceum* (4 cases), *Trichophyton rubrum* (3 cases) and *Microsporum canis* (5 cases). Culture was sterile in 3 cases. Systemic griseofulvin was prescribed at a dose of 20 to 25 mg/kg/day for 8 weeks, combined with a topical antifungal agent. Oral antibiotic therapy was started (72.7% cases) and short-term systemic corticosteroids (5 cases). Treatment was well tolerated, with good clinical and biological progression. At 5 months' follow-up, persistent partial alopecia was observed (8 cases), scarring alopecia (2 patients) and favorable hair regrowth (12 cases).

**Conclusion:** Our results are in accordance with the literature concerning the predominance of male gender and rural origin in Kerion Celsi. Nevertheless, *Trichophyton mentagrophytes* was the dominant agent in our study, in contrast to the Maghrebian and Tunisian series where *Microsporum canis* was the most common and expanding. *Trichophyton violaceum*, identified in some of our patients, has rarely been reported in the literature. The dermatophytes involved in inflammatory tinea capitis are diverse, their clinical aspects polymorphous, and they are likely to vary from one region or country to another. Furthermore, some bacterial skin dermatoses of the scalp can mimic Kerion Celsi, leading to misdiagnosis. Early diagnosis and treatment of suitable systemic and topical antifungals after mycological sampling, are essential to limit the risk of contamination and prevent unsightly scarring alopecia. Kerion Celsi is a potentially severe condition in children, which may also affect adolescents. It can mimic many confusing differential diagnoses, therefore delaying diagnosis and increasing the risk of sequellar scarring alopecia.



## The efficacy and safety of terbinafine for tinea capitis in pediatric patients

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## **Introduction & Objectives:**

Tinea capitis is the most common scalp infection in the pediatric population, caused by Microsporum and Trichophyton species.

It always requires systemic antifungal because topical treatments can't reach deep into the hair follicles .Currently, oral griseofulvin is the treatment of choice for tinea capitis.

The objective of this study is to evaluate the efficacy and safety of oral terbinafine in treating Tinea capitis among pediatric patients at Ibn Rochd University Hospital in Casablanca

#### **Clinical cases:**

Out of 267 patients, only 5 (1.87%) received terbinafine treatment, comprising 3 males and 2 females with a mean age of 8.2 years (range: 5 to 13 years).

Four patients presented with the noninflammatory form of tinea capitis, while one exhibited the inflammatory form known as kerion celsi.

Terbinafine was prescribed in 3 cases due to fungal resistance, in 1 case due to poor compliance, and in another due to a drug interaction.

The treatment protocol involved initiating all patients on terbinafine at 4 to 6mg/kg/day, in addition to topical antifungal agents, with treatment durations ranging from 4 to 6 weeks.

Liver function tests performed before treatment commencement to ensure the safety of the therapy showed no reported side effects, and all patients experienced significant improvement with complete hair regrowth.

## **Discussion:**

Griseofulvin has been the standard treatment for tinea capitis since the late 1950s, demonstrating safety and efficacy.

However, treatment failures can occur due to various factors including fungal resistance, drug interactions, poor compliance, and potential side effects.

Terbinafine is increasingly starting to be used for the treatment of tinea capitis. While its marketing authorization for tinea capitis for patients over 15 years, several studies have demonstrated good tolerance of this medication in younger children.

The standard pediatric dose of terbinafine is 4 to 5 mg/kg/day, with treatment durations ranging from 4 to 6 weeks. Longer durations may be necessary for Microsporum canis infections.

Side effects of terbinafine are rare and include gastrointestinal symptoms, rashes drug reactions and Liver enzyme abnormalities

To ensure the safety and tolerance of the treatment, it is recommended to perform a liver function test before starting the treatment and four weeks after initiation.

## **Conclusion:**

Griseofulvin remains the preferred oral treatment for tinea capitis, while terbinafine offers a viable alternative with good tolerance and minimal side effects.

Our experience supports the efficacy and safety of terbinafine in pediatric patients with tinea capitis.

Controlled studies are needed to determine optimal dosing and duration of terbinafine therapy and to evaluate potential drug-related side effects in children