

**Abstract N°: 218****Porokeratosis palmaris et plantaris disseminata with hand and feet deformity: a rare presentation**

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**Introduction & Objectives:**

Porokeratosis Palmaris Et Plantaris Disseminata (PPPD) is a rare subtype and there are sparse reports with hand and feet deformity.

**Materials & Methods:**

A 42-year-old man, presented with asymptomatic multiple dark-colored lesions all over the body and thickening of palms and soles from past 30 years, bending down of hands and feet from past 25 years. The lesions started over feet to become generalized within a span of 5-10 years. Examination revealed multiple hyperpigmented annular papules and coalescing plaques over face, trunk, buttocks, upper, and lower limb of size varying from 0.5x0.5 cm to 4x5 cm with thread-like elevated margin. Palmo-plantar surface showed discrete hyperkeratotic yellowish-white plaques and flexion deformity at metacarpophalangeal joints with pseudo-ainhum formation in 3 fingers and nail dystrophy in almost all fingers. Oral mucosa revealed single erosion near right second molar which was opined to be part of porokeratosis by the dentist. Dermoscopy and skin biopsy confirmed the diagnosis. X-ray of hands and feet revealed diffuse osteopenia, acral osteolysis and resorption. His 2D-echo done in view of chronic chest pain revealed dilated cardiomyopathy.

**Results:**

Few case reports of PPPD and Meibelli types described bony resorption of the tips of the digits, flexion deformities, and nail dystrophy including ainhum formation.

**Conclusion:**

To the best of our knowledge, this is the first case to report a rare case of porokeratosis palmaris et plantaris disseminata with hand and feet deformity, ainhum formation, nail dystrophy, mucosal and facial involvement in an immunocompetent patient with no family history.

**Abstract N°: 263****A potential trigger for IgG4-related disease; a case of metallosis due to hip joint prosthesis presenting with unique skin lesion on the knee.**

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**Introduction & Objectives:**

A 73-year-old woman underwent arthroplasty for the left hip osteoarthritis 16 years ago and replacement surgery eight years ago due to the loosening. Three years ago, she underwent revision surgery for the pseudarthrosis due to wire injury which appeared at four years ago. One year ago, she had an operation to remove the damaged wire, and a few months later she was referred to our department for the erythematous plaque with itchiness on her left knee. At the first visit, she had a ping-pong ball-sized elastic hard verrucous reddish nodule accompanied by markedly elevated radial reddish streaks on the left lateral knee and slightly elevated reddish streaks on the left lateral femur.

**Materials & Methods:**

We performed and examined skin biopsies, blood tests, and scanning electron microscope (SEM).

**Results:**

The skin biopsy revealed a dense infiltration of plasma cells mainly around the lymphatic vessels in the entire dermis and a random pattern of hypertrophic collagen bundles in the deep dermis. Immunohistochemical staining showed that immunoglobulin G subclass 4 (IgG4)/ immunoglobulin G (IgG) ratio in the infiltrating plasma cells was 30-40%. Blood examination revealed C-reactive protein (CRP) 3.27 mg/dL, IgG 2,554 mg/dL, and IgG4 184.0 mg/dL. These findings fulfilled the diagnostic criteria of IgG4-related disease (IgG4-RD). Magnetic resonance imaging of the left femur showed fluid in the hip joint and edema around the joint, compatible with metallosis, and we performed femoral revision arthroplasty. The histopathology of the periprosthetic femoral stem tissue showed dense histiocyte-predominant infiltration and phagocytosis targeting to fine foreign bodies identified as titanium by SEM. The skin eruption quickly resolved after the revision surgery and then IgG4 and CRP dropped to 134 mg/dL and < 0.04 mg/dL, respectively.

**Conclusion:**

The patient met the diagnostic criteria for IgG4 as well as the diagnosis of metallosis. Although no cases of prosthesis failure or metallosis complicated with IgG4-RD have been reported, we strongly suspect that metallosis was involved in the pathogenesis of IgG4-RD, since the serum level of IgG4 declined as the eruption improved after the revision surgery. Physicians need to examine patients with metallosis more carefully to identify secondary diseases.

**Abstract N°: 350**

**From Pixels to Predictions: Development of a multimodal-based deep learning algorithm for accurate and efficient erythema score assessment in radiation induced dermatitis**

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**Introduction & Objectives:**

Despite the significant progress made in computer-aided diagnostics using AI, there are currently limited viable methods available for the analysis and categorization of radiation-induced skin reactions (RISRs), making this study a critical step towards filling this gap. This study aims to develop advanced techniques that utilise a deep learning algorithm for the automatic classification of RISRs in accordance with the Common Terminology Criteria for Adverse Events (CTCAE) grading system. To achieve this objective, Scarletred® Vision, a state-of-the-art digital skin imaging method that allows for *standardized* monitoring and objective assessment of acute RISRs, was utilised. The study conducted SEV\* measurements on 2D digital skin images after transforming them into the CIELAB colour space.

**Materials & Methods:**

In terms of data, we propose a robust pipeline to collect a dedicated, diverse, and curated image dataset using Scarletred® Vision, a CE certified medical device software platform. When it comes to models, *we have trained a deep learning-based multimodal system that incorporates* Scarletred® Vision augmentation pipeline. The dataset consisted of 2192 images, with a train-test size of 80:20 and image dimensions of 512 x 512. We used patch dimensions of 32 x 32 and 256 patches per image. The approach towards the solution involved input image patch, position embedding, and feature augmentation, with image augmentation based on feedback from ensembled prediction (R. Ranjan et al., 2021).

**Results:**

The results of our study indicate that the proposed model achieved high precision (92.51%), recall (91.21%), and F-score (91.83%), with a test accuracy of 92.02%. The model's performance was evaluated in a class-wise manner, where the sensitivity for classes 0, 1, and 2 was found to be 96%, 94%, and 86%, respectively. Similarly, the specificity was determined to be 94%, 97%, and 97% for the respective classes. The overall accuracy for the model was found to be 96%, 94%, and 86% for classes 0, 1, and 2, respectively. These results indicate a high level of precision and discriminatory power of the model across all three classes.

**Conclusion:**

This study provides the first benchmark results using multimodal-based deep learning algorithm for erythema severity score classification. Data from this study highlights that the estimation of severity score for RISRs can be automated using Scarletred® Vision as decision support system. Our algorithm is currently being refined to provide also the estimated localisation, size and tissue composition of the lesion.



**Abstract N°: 358****Rare Case of Pyodermatitis-Pyostomatitis Vegetans with Concomitant Ulcerative Colitis and Chronic Myelomonocytic Leukemia: A Diagnostic and Therapeutic Challenge**

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**Introduction & Objectives:**

Pyodermatitis-pyostomatitis vegetans (PD-PSV) is a chronic inflammatory disease that affects the skin and mucous membranes and is usually associated with inflammatory bowel disease (IBD). Mucosal lesions are characterized by small confluent pustules and erosions that follow a specific “snail track” pattern on an erythematous base. Cutaneous lesions manifest as exudative, vesiculopustular, and vegetative plaques that mostly affect the scalp and intertriginous areas.

In this case report, we present a 27-year-old male patient diagnosed with UC, PD-PSV, and chronic myelomonocytic leukemia (CMML).

**Materials & Methods:**

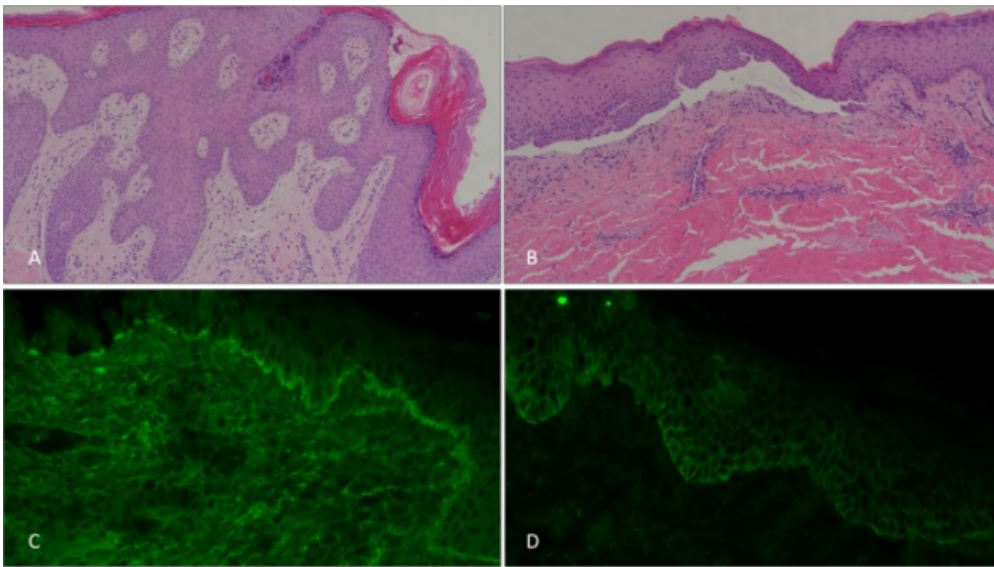
The patient had been diagnosed with ulcerative colitis (UC) nine months prior to presentation and was admitted to our hospital due to a rash that started in the axillary region and spread to the anterior and posterior surface of the trunk, pubis, and extremities. He had difficulty eating and had a history of diarrhea for several days. For UC, he was started on mesalazine orally at a dose of 1 gram per day.

**Results:**

Physical examination revealed annular, center-crusted, polycyclic erythematous vesicopustules on the scalp, neck, trunk, extremities, axillae, pubis, and inguinal region. Many confluent pustules were observed on the gingiva and palate, which followed a characteristic “snail track” pattern, and numerous confluent pustules on labial mucosa with yellow exudates and hemorrhagic crusts that transformed into vegetative plaques.

Viral serology and monkeypox tests were negative. Autoimmune panel showed positive ANA and p-ANCA. Imaging studies showed no significant findings. Colonoscopy was consistent with UC. Thrombocytosis and leukocytosis were observed in the peripheral smear, leading to a bone marrow aspiration biopsy and a diagnosis of CMML by Hematology. Histopathologic examination showed mixed inflammatory cell infiltration with eosinophils, with DIF revealing positive IgA, IgG, and C3C. PD-PSV was diagnosed based on characteristic clinical findings, negative tests for other bullous diseases, and compatible histopathology. Treatment with methylprednisolone resulted in a dramatic clinical response within 3 days.





**Figure 1:** (A, B) Mild hyperkeratosis, irregular acanthosis, pseudoepitheliomatous hyperplasia, dense mixed inflammatory cell infiltration (B) Separated subepidermal areas, superficial dermal edema, polymorphic leukocyte infiltration, prominent vascular endothelium (H&E, x40, x200, respectively) (C, D) IgA and fine granular IgG deposition at the dermo-epidermal junction on immunofluorescence (x200)

### Conclusion:

PD-PSV may occur during an exacerbation of IBD or represent subclinical bowel disease. Other bullous disorders with vegetating vesicopustular eruptions should be distinguished from PD-PSV. The diagnosis is based on clinical features, association with IBD, peripheral eosinophilia, and distinctive histologic features. Topical or systemic corticosteroids in combination with immunomodulators used in the treatment of IBD are the most common first-line therapies. This case emphasizes the importance of considering PD-PSV in the differential diagnosis of skin and mucosal lesions in patients with underlying IBD, the concomitant disorders that may accompany PD-PSV, such as myeloproliferative diseases, and the need for multidisciplinary management in patients with other comorbidities.



**Abstract N°: 536**
**Recalcitrant Giant Cellulitis-like Sweet's Syndrome - More Than Skin Deep**

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**Introduction & Objectives:**

Giant Cellulitis-like Sweet's Syndrome (GCS) is a novel variant of Sweet's Syndrome (SS). It is characterised by relapsing widespread giant lesions which vary from erythematous infiltrated lesions to painful indurated plaques. These can develop asymmetrically over the body and are often associated with an underlying malignancy. We present the case of treatment recalcitrant GCS in a sixty-two year old female who initially presented to the emergency department with a three\*\* day history of right lower limb oedema and erythematous rash.

**Materials & Methods:**

Background history was significant for Bechet's disease, pyoderma gangrenosum, hidradenitis suppurativa, Crohn's disease with a panproctocolectomy and ileostomy formation, primary hyperparathyroidism and osteoporosis. She had received the first dose the SARS-CoV-2 vaccine (viral vector) four days prior to presentation. On examination she appeared systemically unwell and febrile.\*\* There were well demarcated erythematous, indurated plaques with associated purpura noted over the right lower limb. The left thigh had a similar but less severe appearance. Serological investigations were supportive of acute infection with white cells  $12.36 \times 10^9/L$  ( $4-11 \times 10^9/L$ ), neutrophils  $10.43 \times 10^9/L$  ( $2-7.5 \times 10^9/L$ ) and CRP 264.6 mg/L (0-5mg/L). She was anaemic with Hb 10.6 g/dL (11.7-16 g/dL). Upon further questioning, it became apparent that similar episodes had been ongoing for the previous three years, managed as cellulitis.

**Results:**

Diagnostic punch biopsy showed evidence of dermal perivascular and interstitial neutrophilic inflammation, with leukocytoclasia and swelling of endothelial cells. A histological diagnosis of SS and clinical diagnosis of GCS was made. Initial treatment with dapsone 50mg once daily (OD) and potassium permanganate soaks was commenced. Despite up-titration of dapsone to 100mg OD, clinical deterioration was noted. A repeat biopsy was again consistent with SS. Prednisolone 0.5mg/kg and doxycycline were commenced with continuation of dapsone. Despite initial improvement, the formation of ulcerated plaques on the lower limbs bilaterally some months later necessitated treatment escalation. The TNF inhibitor adalimumab (40mg alternate weeks) was commenced which was subsequently up-titrated to 40mg weekly. Following most recent review, the lower limb ulceration has stabilised however a significant clinical improvement has yet to be demonstrated.

**Conclusion:**

Our patient had previously been on adalimumab for Crohn's disease for four years prior to presentation which was stopped due to recurrent shingles. We postulate that her previous anti-TNF treatment had controlled her hitherto undiagnosed SS, with discontinuation unmasking her SS. We hypothesize that her underlying uncontrolled Crohn's disease may be driving GCS. GCS occurring as a paraneoplastic phenomenon is less likely in view of previous control with adalimumab and the duration of symptoms. To this end, in conjunction with our gastroenterology colleagues, we are considering ustekinumab or methotrexate as potential future treatment options. Of the eleven reported cases of GCS in the literature, none describe use of a biologic agent either as treatment for GCS or for a different underlying disease process. This unique case highlights the challenges of

managing patients with multiple pro-inflammatory co-morbidities, especially those who experience biologic treatment failure.

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**Abstract N°: 639**
**An Arciform erythematous plaque with hypopigmentation: A diagnostic challenge**

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**Title: An Arciform erythematous plaque with hypopigmentation: A diagnostic challenge.**
**Introduction & Objectives:**

Annular Lichenoid Dermatitis of Youth (ALDY) is now a well-recognized distinctive lichenoid dermatosis first described in 2003 by Annessi G. It is characterised by specific clinical and histological features, occurring in younger individuals with a chronic relapsing course. Classic lesions present as persistent, asymptomatic annular erythematous macules and patches with red to violaceous non-scaling border and central hypopigmentation with majority of lesions localized around groin /flank rarely other areas are involved.<sup>1</sup>

Histopathology shows lichenoid dermatitis limited to the tips of rete ridges often tips appearing “squared off”. Infiltrate consists of predominately intraepidermal CD8+ and some CD4+ T cells with poly clonal T cell receptor rearrangement. Close differential diagnosis include mycosis fungoides, morphea, annular erythemas and inflammatory lesions of vitiligo. Topical corticosteroids and topical tacrolimus represent the most effective drugs for ALDY treatment. <sup>2</sup> We present the first case of ALDY from Saudia Arabia.

**Materials & Methods:**

Case report; A 17 year old healthy female with an unremarkable personal and family history presented with asymptomatic recurrent rashes over left arm of 2 years duration. On examination she has single erythematous arciform rash about 6\*4cm over left arm just above the elbow crease with raised border & scaling mostly around periphery of rash and relative hypopigmentation inside the arc. No lesions elsewhere on the body/scalp/mucosae. No history of seasonal variation, no drug history. She has been treated for tinea multiple times with no benefit.

Routine lab tests including complete blood picture, liver function tests, serum creatinine was normal. Skin biopsy from the lesion from showed moderate epidermal acanthosis, confluent para keratois, and papillomatosis with mild focal spongiosis. Lichenoid dermal lymphocytic infiltrate with focal basal cell vacuolation, presence of colloid bodies in the lower epidermis and the papillary dermis with infiltrate around the rete ridges. Periodic acid Schiff staining revealed no fungi

**Results:**

Based on these clinico pathological findings, the diagnosis of ALDY was made. The patient was given topical moderate corticosteroids with disappearance of lesions after 3 weeks, followed by topical tacrolimus 0.1% for one month with no recurrence so far.

**Conclusion:**

ALDY is not an uncommon entity, its just under reported. It is now a distinctive chronic lichenoid dermatosis with specific clinical and histological features affecting mostly young individuals all over the world with uncertain etiology.

## References:

- \1. Annessi G, Annessi E. Annular Lichenoid Dermatitis (of Youth). Dermatopathology (Basel). 2022 Jan 16;9(1):23-31
- \2. Di Mercurio M, Gisondi P, Colato C, Schena D, Girolomoni G. Annular Lichenoid Dermatitis of Youth: Report of Six New Cases with Review of the Literature. Dermatology. 2015;231(3):195-200. doi: 10.1159/000381705. Epub 2015 Jul 9. PMID: 26160268.

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**Abstract N°: 987****Gastroenterological comorbidity in patients with rosacea and overweight**

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**Introduction & Objectives:** Rosacea is a chronic recurrent inflammatory disease of the facial skin of unknown etiology and unclear pathogenesis. It is associated with vascular anomalies. Many studies have been conducted on this problem, but this progress is not enough to increase the effectiveness of treatment and reduce the frequency of exacerbations. Various investigations show the association of rosacea with other pathological conditions, which can probably be the cause of the development and relapse of rosacea. Researchers pay significant attention to diseases of the digestive organs. The purpose of this research was to study the structure of gastroenterological pathology in patients with rosacea and overweight.

**Materials & Methods:** 76 women with erythematous telangiectatic rosacea, aged from 22 to 45 years, were examined. Body mass index (BMI) was less than 24.9 in 23 patients (group 1), and more than 25 in 53 patients (group 2).

**Results:** A combination of rosacea and gastroenterological disorders was found in all patients. Irritable bowel syndrome was revealed in 34.8% and 60.4% of persons of groups 1 and 2, respectively; small intestinal bacterial overgrowth – in 8.7% and 49.1% of women of groups 1 and 2, respectively. Pancreatitis was diagnosed in 21.7% and 34% of individuals of groups I and II, respectively, cholecystitis was detected in 34.8% and 58.5% of women of groups 1 and 2, respectively; and cholelithiasis was revealed in 11.3% of patients of group 2. 52.2% and 75.5% of persons of groups 1 and 2 suffered from gastroesophageal reflux disease; gastritis was diagnosed in 39.1% and 43.4% of patients of groups 1 and 2, respectively. Helicobacter pylori infection was revealed in 47.8% and 54, 7% of women of groups 1 and 2, respectively.

**Conclusion:** Irritable bowel syndrome, small intestinal bacterial overgrowth, pancreatitis, cholecystitis, cholelithiasis, gastroesophageal reflux disease, gastritis and Helicobacter pylori infection were found in patients with rosacea. Women with rosacea and overweight were more often associated with diseases of the gastrointestinal tract compared to patients without overweight. In women with rosacea and overweight, irritable bowel syndrome accompanied by small intestinal bacterial overgrowth, cholecystitis with cholelithiasis and gastroesophageal reflux disease was more often detected.



**Abstract N°: 1158**
**Score for the systemic inflammatory troncular recurrent acute macular eruption (SITRAME) Syndrome Diagnosis**

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**Introduction & Objectives:**

The systemic inflammatory troncular recurrent acute macular eruption (SITRAME) Syndrome is a sporadic autoinflammatory syndrome recently described among adults (1). It is characterized by a recurrent macular trunk eruption, non-pruritic, associated with at least one episode of systemic inflammation documented by an elevation of C-Reactive Protein (CRP) during flares. Currently, no genetic mutation has been found yet to be associated with SITRAME syndrome.

The objective was to well characterize the SITRAME syndrome, by defining diagnostic criteria allowing its identification.

**Materials & Methods:**

To identify major and minor clinical and biological criteria, we analyzed the clinical and biological characteristics of the initial cohort of 16 patients initially described as having a SITRAME syndrome. We wanted to isolate mandatory major criteria present in the 16 described patients and minor criteria present in at least half of the patients.

**Results:**

We identified four major diagnostic criteria, present in all patients, and four minor criteria, present in more than half of the patients for each and one exclusion criteria (Table 1). The presence of four mandatory major criteria and the presence of at least one minor criteria, and exclusion criteria or a score strictly higher than 5 points was necessary to diagnose the SITRAME syndrome (Table 1). In the initial cohort of 16 patients, the score ranged from 6 to 9, with an average of 7.

**Conclusion:**

We propose a clinical-biological diagnostic criteria to diagnose SITRAME if the score is strictly higher than 5. This score aims to harmonize the diagnostic criteria for diagnosing the SITRAME syndrome.

1. Soria A, Amsler E, Garel B, Moguelet P, Tieulié N, Cordoliani F, Guichard I, Mahé A, Grateau G, Boursier G, Georgin-Lavialle S. Systemic inflammatory trunk recurrent acute macular eruption (SITRAME): A new auto-inflammatory syndrome in adult? J Eur Acad Dermatol Venereol. 2022 Nov 25.

Table 1: Proposed diagnostic criteria for SITRAME Syndrome.

Criteria	Score points
<b>Major= MANDATORY</b>	
<b>Systemic inflammatory:</b> at least 1 documented episode of CRP > 5 mg/ml during the skin flare	<b>1</b>
<b>Troncular macular eruption:</b> Non-pruritic maculopapular rash on the trunk**	<b>1</b>
<b>Recurrent:</b> at least 3 different episodes	<b>1</b>
<b>Acute:</b> lasting less than 8 days	<b>1</b>
<b>Minor</b>	
Fever during flares	<b>1</b>
Flares triggered by infections, vaccinations, or intense physical exercise	<b>1</b>
Asthenia during and/or after flares	<b>1</b>
Associated papular eruption	<b>1</b>
<b>Exclusion criteria:</b> Autoantibodies, monogenic autoinflammatory disease, evolving neoplasm	<b>1</b>
<b>SITRAME syndrome is diagnosed if 4 major criteria, at least 1 minor criteria and exclusion criteria are fulfilled</b>	



**Abstract N°: 1308**
**Prevalence of malignancy and associated risk factors in patients with prurigo nodularis: A Canadian retrospective study**

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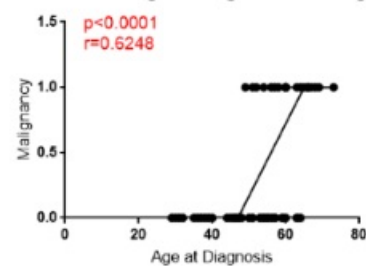
**Introduction & Objectives:** Prurigo nodularis (PN) is a debilitating, inflammatory skin disease that presents with extremely pruritic, hyperkeratotic nodules and a myriad of comorbidities. Recently, there has been evidence linking PN with various malignancies. However, research investigating the association between PN and cancer is limited, and non-existent in Canadian patient populations. As a result, PN remains to be poorly understood. Given that both cancer and PN carry a significant burden of disease and have detrimental effects on patient quality of life, further research is necessary to elucidate the association between these conditions in order to establish appropriate screening and management strategies. Thus, the objective of this study was to investigate the prevalence of malignancy amongst patients with PN and predictive factors for developing cancer.

**Materials & Methods:** This study is a single-centre, observational, retrospective study of clinical characteristics and malignancy in an adult, Canadian patient population with PN that were seen at the Hamilton Health Science Centre and McMaster Dermatology Clinics between 2015 and 2023.

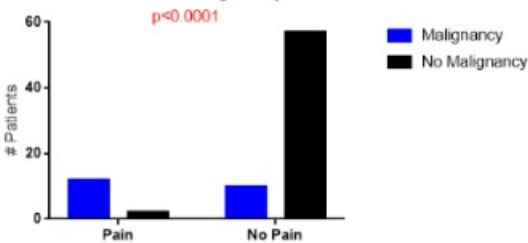
**Results:** A total of 81 patients with a diagnosis of PN were included in the study, with a mean age of 53 years (30-77 age range). Majority of patients were women (60%). Symptoms included pain (17%), dry skin (53%) and itching (100%). Overall, 27% of patients had a concomitant malignancy including non-melanoma skin cancers (NMSCs). Basal cell carcinoma (16%), and squamous cell carcinoma (11%) were the most common malignancies. Apart from NMSCs, melanoma (4%), breast cancer (4%), and non-Hodgkin's lymphoma (4%) were the most common malignancies reported. The onset of malignancy in these patients occurred before (33%), after (58%), and both before and after (8%) the diagnosis of PN. Other comorbidities included mental health disorders (79%), hypertension (35%), other skin diseases (25%), diabetes mellitus (12%), chronic kidney disease (11%), thyroid disease (9%), chronic liver disease (4%), and HIV (1%). Older age at diagnosis, and the presence of pain were significant predictive factors of malignancy ( $P < 0.001$ ). Increased age at diagnosis had a significant, positive correlation with malignancy ( $r = 0.62$ ,  $P < 0.001$ ).

**Conclusion:** PN is a complex, multi-factorial skin disease that presents with numerous comorbidities and may be associated with increased risk for the development of malignancy. As a result, appropriate screening is important for early detection and improving outcomes for patients with this disease. Therefore, dermatologists' awareness of this association and possible risk factors for developing malignancy is critical.

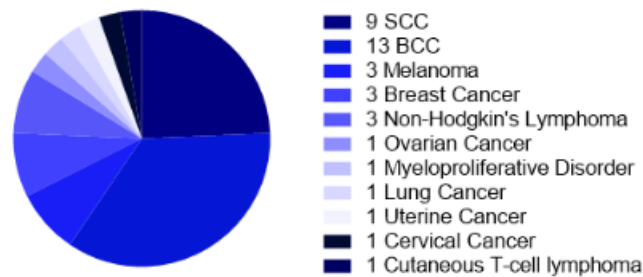
Correlation of Age at Diagnosis vs Malignancy



Pain vs Malignancy



Malignancy in PN Patients



**Abstract N°: 1343****Dapsone prescribing and monitoring trends: results from a UK-wide survey**

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**Dapsone prescribing and monitoring trends: results from a UK-wide survey****Introduction & Objectives:**

Dapsone is utilised for managing e.g. hidradenitis suppurativa, dermatitis herpetiformis, neutrophilic dermatoses, vasculitis, and autoimmune bullous diseases<sup>1</sup>. Even with the advent of more targeted therapies for conditions such as hidradenitis suppurativa, dapsone remains widely used, either in isolation or combination with, for instance, biologics<sup>2</sup>. However, there are no national guidelines on prescribing and monitoring of dapsone. Anecdotally, this has led to great variability in practice amongst UK clinicians, both in dapsone use and monitoring. It is concerning that lack of guidance would limit the use of a potentially useful therapeutic option. We therefore produced a questionnaire to describe the current UK dapsone prescribing trends.

**Materials & Methods:**

A questionnaire was disseminated to all British Society of Medical Dermatology and Scottish Dermatological Society members.

**Results:**

98 dermatology doctors, and one specialist pharmacist, all currently working in the UK, completed the questionnaire. Of these doctors, 74% were consultants, and the rest were made up of specialist registrars and non-consultant grade doctors. The majority were working in England or Scotland and at a teaching hospital (68%).

Participants were asked to what extent they agreed with the statement 'guidelines are needed to ensure the safe use of dapsone in dermatology', to which the average answer was 9/10 (10 being strongly agree).

The most common starting dose amongst responders was 50mg daily (94%). The maximum dose titrated was variable, with 22% selecting 100mg daily, 35% 150mg daily, 36% 200mg daily, and 3% 300mg daily.

The majority of participants (59%) stated that they did weekly blood tests during the first month of dapsone therapy. After the first month, the most common approach monthly blood tests, followed by three-monthly testing once established on dapsone for longer. For half of clinicians (50%), a haemoglobin drop of 20g/L would prompt stopping dapsone or reducing a dose, whilst 18% took a more cautious approach of a drop in haemoglobin of 10g/L, and others suggested it depends on the clinical context. Reticulocytes were monitored by 88% of participants, yet 60% would not alter the dose based on a rising reticulocyte count alone.

**Conclusion:**

This is to our knowledge the first survey of prescribing monitoring practices for dapsone, with our data suggesting a great variability in practice with routines differing even within departments. We propose that our data demonstrate a need for further practical guidance for dermatologists, to ensure the drug is appropriately and safely used. We propose to proceed with a Delphi consensus building exercise across the UK and more widely in

Europe.

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**Abstract N°: 1411**
**Clinical and histological manifestations of palmar and dorsal neutrophilic dermatosis of the hands**

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**Introduction & Objectives:**

Neutrophilic dermatosis of the hands (NDH) is characterized by the presence of erythematous/edematous plaques, and pustules in the hands. Histology typically reveals dense -mainly neutrophilic- inflammatory infiltrates in the dermis, with signs of vasculitis of the (dorsal) hands. Nowadays, NDH is considered an overlap between pustular vasculitis and acral Sweet Syndrome (SS) with mild systemic symptoms. Certain authors further subdivide NDH into neutrophilic dermatosis of the palms (NDP) and dorsal hands (NDDH) whereas co-occurrence of lesions on both sides seems rare. In this retrospective study, we compare the clinical and histopathological characteristics of patients with NDP and NDH.

**Results:**

We included 7 female and 1 male patients diagnosed in our department with NDP (n=5) or NDDH (n=3), from 2003 to 2015. Median (IQR) age was 78 (78, 78) in the NDP and 81 (79.5, 85) in the NDDH group. Patients' characteristics are depicted in Table 1. In the NDP group, clinical characteristics included: vesiculobullae (n= 3), edematous/erythematous plaques (n= 2), and pseudo vesicles (n=2), and in the NDDH group: vesiculobullae (n= 3), edematous/erythematous plaques (n= 2), and pustules (n=3).

All skin lesions were biopsied within a week of appearance. Histological findings are resumed in Table 2. Epidermal spongiosis with neutrophilic exocytosis and vesiculation were present in both groups. Epidermal abscesses (n=2) and/or subcorneal pustules (n=2) were only present in NDDH patients. The predominant feature in both groups was a dense neutrophilic infiltration of the papillary dermis with edema and pseudo-vesiculation (n=5). However, extension to the reticular dermis was mostly absent in the NDP group and when present was minimal (n=2). Neutrophilic infiltrates extended into the deeper dermis in 2/3 of NDDH cases. Perivascular distribution was absent in the NDDH group. Vasculitis was present in one NDP and in two NDDH cases, one of which showed true fibrinoid necrosis. Patients in both groups showed excellent responses to oral steroids. Two patients had recurrent disease, and one required maintenance treatment with dapsone.

**Conclusion:**

There is a sparsity of information referring to hand-side-related features of NDH. In our study, NDP seemed to lack a pustular component and showed less pronounced epidermal changes. In the NDDH group, vasculitis was more common, correlating with the intensity of neutrophilic infiltration and with the presence of pustules, as previously reported. Vasculitis of NDH is now thought to be an epiphenomenon to the release of toxic metabolites from activated neutrophils rather than an immune-mediated process. The objective differences in the occurrence of vasculitis between NDP and NDDH identified in our series are in line with previous studies and could provide the missing link in NDH pathogenesis, as NDP and NDDH could be at the opposing ends of a continuum extending from localized SS to "pustular vasculitis".

**Table 1 Clinical characteristics of patients with neutrophilic dermatosis of the hands**

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8
<b>Sex/Age (years)</b>	M/78	F/78	F/87	F/67	F/78	F/81	F/78	F/89
<b>Hand Side</b>	Palmar	Palmar	Palmar	Palmar	Palmar	Dorsal	Dorsal	Dorsal
<b>Aberrant lesions</b>	-	Distal forearms	-	-	Oral erosions	-	-	-
<b>Fever</b>	-	Yes	-	-	-	-	-	-
<b>Leukocytosis/ Neutrophilia</b>	-	Yes	Yes	Yes	Yes	Yes	Yes	Yes
<b>Time to diagnosis</b>	3 years	1 day	2 days	3 days	5 days	1 month	5 months	8 days
<b>Associations</b>	MDS, RA	Infection	Sjogren's syndrome	Infection	RA	Trauma, Infection	Arthritis, Anemia	Infection
<b>Treatment</b>	CS, Dapsone	CS	CS	CS	CS	CS	CS	CS
<b>Outcome</b>	Excellent	Excellent	Excellent	Excellent	Excellent	Excellent	Excellent	Excellent
<b>Recurrence</b>	Yes	-	-	-	-	Yes	-	-

RA: rheumatoid arthritis, MDS: myelodysplastic syndrome, CS: systemic corticosteroids

Table 2 Histological characteristics of patients with neutrophilic dermatosis of the hands

	Patient 1 NDP	Patient 2 NDP	Patient 3 NDP	Patient 4 NDP	Patient 5 NDP	Patient 6 NDDH	Patient 7 NDDH	Patient 8 NDDH
Epidermal spongiosis	-	+	+	+	+	+	+	+
Epidermal neutrophilic exocytosis	-	-	+	+	+	+	+	+
Epidermal abscesses	-	-	-	-	+	+	+	-
Subcorneal pustules	-	-	-	+	-	-	+	+
Pseudovesicles	-	-	+	+	+	+	+	-
Vesiculation	-	-	-	-	-	+	+	-
Dermal infiltrate	+	+	+	+	+	+	+	+
Neutrophilic infiltration	+	+	+	+	+	+	+	+
Lymphocytic infiltration	+	+	+	+	+	+	+	+
Eosinophilic infiltration	+	+	+	+	+	+	+	+
Superficial Infiltrate	+	+	+	+	+	+	+	+
Perivascular Infiltrate	+	+	+	+	+	-	-	-
Interstitial Infiltrate	+	-	+	+	+	+	+	+
Deep Infiltrate	-	-	+	-	+	+	+	-
Leukocytoclasia	-	-	+	+	+	+	+	+
Endothelial swelling	+	-	+	+	+	+	+	+
Vasculitis	-	-	-	-	+	-	+	+
Fibrinoid necrosis	-	-	-	-	-	-	+	-

Abbreviations: mod.: moderate, sev.: severe



**Abstract N°: 1500**
**Sex- and age-specific response to non-biologic/biologic psoriatic therapies of different body localizations: 10-year data from the Swiss Psoriasis Registry (SDNTT)**

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**Introduction & Objectives:**

There is limited real-world data regarding the stratification of localized psoriasis by sex- and age-related response to systemic therapy. In this study, we investigate the severity and distribution of psoriasis over time in female and male patients receiving systemic therapies, categorized by age within the Swiss Dermatology Network for Targeted Therapies (SDNTT), the Swiss national psoriasis registry. \*\*

**Materials & Methods:**

Patient data was obtained over 11 years from secondary and tertiary Swiss hospitals (October 2011 to November 2022) in the SDNTT. From these, we analysed the Psoriasis Area and Severity Index (PASI) of the head, trunk, upper and lower extremities over two years of systemic non-biologic and biologic treatment. For each body region, an individual localized PASI (locPASI) was calculated as the sum of erythema, scaling, and thickness of the lesions, with each factor scored on a scale of 0 to 4. This was then multiplied by the estimated Body Surface Area (BSA) factor (0, none; 1, <10%; 2, 10–29%; 3, 30–49%; 4, 50–69%; 5, 70–89%; and 6, 90–100%) for the final score.

**Results:**

At time of analysis, 409 patients were treated with systemic non-biologic (159 females and 250 males) and 512 with biologic (191 females and 321 males) therapies. Male patients had a higher locPASI at baseline in comparison to females for legs (11.4 vs. 8.8), trunk (8.5 vs. 7.0) and arms (9.9 vs. 7.5), with a high significance of  $p < 0.001$  for each localization, but not for the head (7.3 vs. 9.1;  $p = 0.197$ ). Female patients aged 18 to 40 had a significantly higher locPASI score for the head than 55 and older (10.3 vs. 7.7;  $p = 0.020$ ). After two years of treatment, the older female group had a significantly higher locPASI for legs than the younger (2.7 vs. 1.2;  $p = 0.006$ ). Over the treatment period, females had a more rapid response to non-biologic treatment for the head than males (mean locPASI reduction of -7.2 vs. -5.4;  $p = 0.036$ ). For both sexes receiving non-biologic therapies, a PASI  $\leq 2$  was only reached for head and trunk. In female patients, a longer duration was required to achieve an improvement for the head (24 vs. 18 months); whereas for male patients, the trunk region took longer (18 vs. 6 months). An absolute PASI  $\leq 2$  was achieved after 12 months of biologic treatment for all locations in female and male patients, except for the legs of male patients (locPASI 2.6).

**Conclusion:**

Significant disparities in localized PASI were observed between female and male patients, as well as between younger and older female patients. The age, sex and severity of distinct localizations should be taken into account in alignment with patients' individual needs and treatment goals.

All patients (n = 921)					
	No. of evaluable male patients	Male (n = 571)	No. of evaluable female patients	Female (n = 350)	p-value
Median PASI (IQR)	558	9,86 (5,65 - 12,6)	341	8,03 (3,6 - 10,87)	0,000 <sup>a,*</sup>
Median Age in years (IQR)	570	46,39 (36,5 - 55,5)	349	45,19 (31,5 - 56,5)	0,123 <sup>a</sup>
Median BMI (IQR)	535	28,15 (24,1 - 30,85)	327	26,42 (21,82 - 30,1)	0,000 <sup>a,*</sup>
Median Disease duration (years)	460	16,75 (6 - 24)	279	16,99 (5,75 - 24)	0,720 <sup>a</sup>
Ongoing PsA (%)		20,14%		20,00%	0,975 <sup>b</sup>
Current smokers (%)		18,03%		17,14%	0,600 <sup>b</sup>

Table 1. Main patient characteristics, demographics and clinical information at baseline. PASI Psoriasis Area Severity Index, IQR interquartile range, BMI body mass index (kg/m<sup>2</sup>), PsA Psoriasis arthritis, a Wilcoxon test, b Fisher exact test; \*  $p < 0.001$

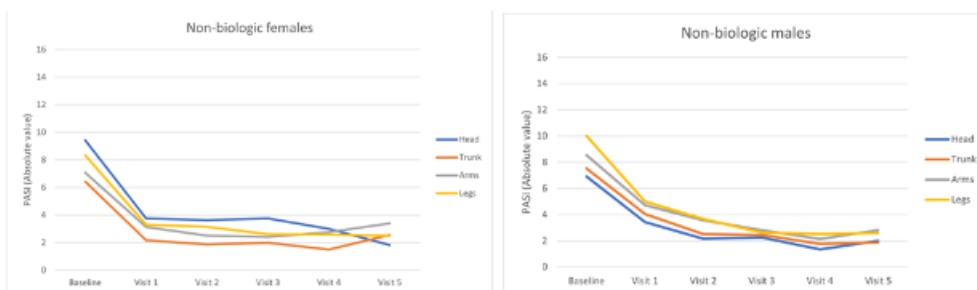


Figure 1. locPASI of the head, trunk, arms and legs over two years of systemic non-biologic treatment (methotrexate, fumarates, retinoids, cyclosporine and apremilast), from baseline through clinical follow-up visits 1 – 5. The two graphs show progression of PASI female and male patients respectively.

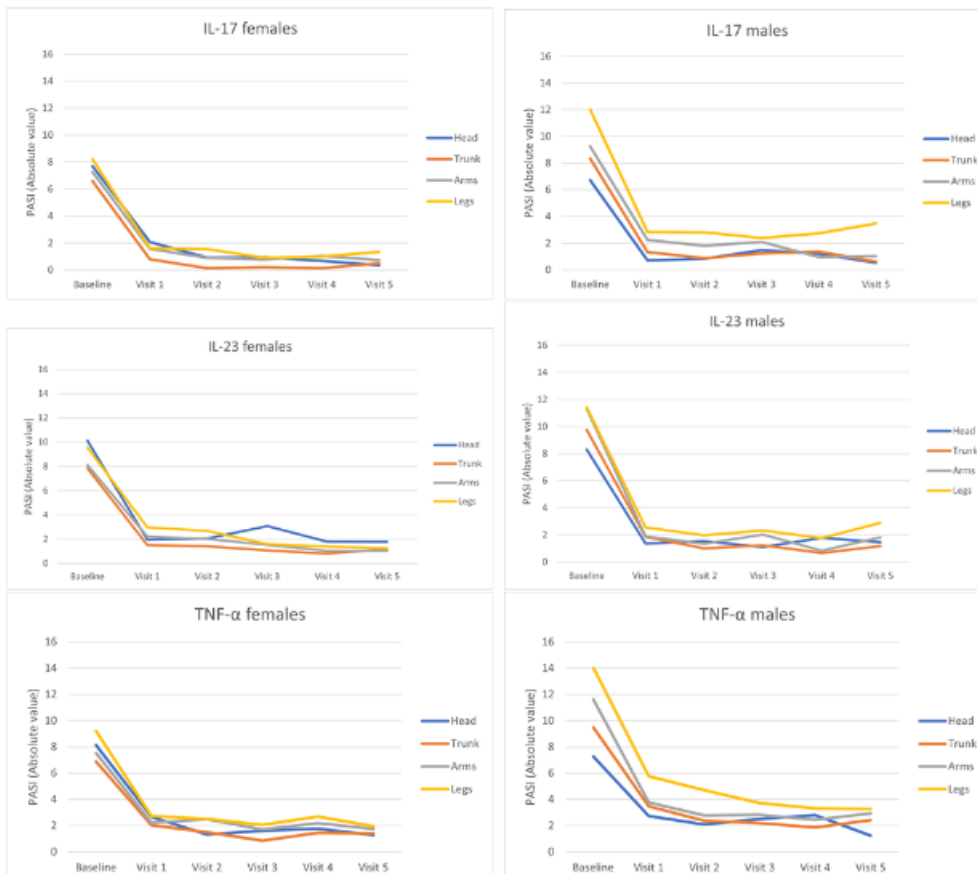


Figure 2. locPASI of the head, trunk, arms and legs over two years of biologic treatment, from baseline through clinical follow-up visits 1 – 5. Individual graphs show the progression of PASI of female and male patients treated with IL-17 (secukinumab and ixekizumab), IL-12/23 and IL-23 (ustekinumab, tildrakizumab, risankizumab and guselkumab), TNF- $\alpha$  (adalimumab, certolizumab, etanercept, golimumab, infliximab) inhibitors respectively.





**Abstract N°: 1565****Intraepidermal neutrophils are major source of IL-17A expression in palmoplantar pustulosis**

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**Intraepidermal neutrophils are major source of IL-17A expression in palmoplantar pustulosis****Introduction & Objectives:**

Palmoplantar pustulosis (PPP) is a chronic debilitating disorder of palm and sole, which significantly affects quality of daily living. Previous studies showed that IL-23/IL-17 axis is activated in lesional PPP and the use of biologics targeting IL-23 (e.g. guselkumab) demonstrated somewhat promising treatment response in a portion of patients with PPP, which was, however, less efficacious than that observed in chronic plaque psoriasis. To clarify IL-23 downstream cytokine expression profile in PPP, we tried to analyze IL-17A and IL-17F protein expression in biopsied skin samples.

**Materials & Methods:**

We underwent IL-17A and IL-17F immunohistochemical staining in 17 biopsied PPP samples. In high power field of view, cytokine expressing cells and their proportion were measured. Relationship between immunohistochemical findings and clinical data were analyzed by utilizing correlation analysis and regression analysis.

**Results:**

Dermal inflammatory cells, which were predominantly mononuclear cells, expressed IL-17A and IL-17F in  $8.9 \pm 5.8\%$  and  $2.3 \pm 1.6\%$ , respectively. Intraepidermal pustules, majority of cells were neutrophil, expressed IL-17A and IL-17F in  $66.1 \pm 24.9\%$  and  $2.3 \pm 3.0\%$ , respectively. Relationship between immunohistochemical findings and clinical features demonstrated that severity of disease cannot be predicted by IL-17 expression in biopsied sample.

**Conclusion:**

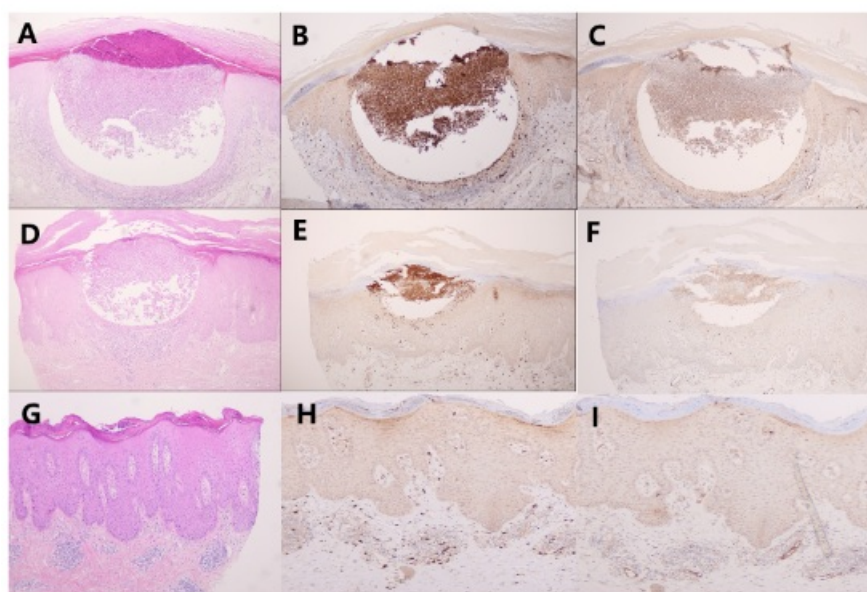
In conclusion, IL-17A was highly expressed in polymorphonuclear leukocytes in intraepidermal pustules, which emphasize the necessity of further research focusing on the nature of neutrophils in intraepidermal pustules of PPP for better understanding and management of this debilitating disorder.

**Table.** IL-17 expression profile in skin samples.

	Anti-IL-17A stain			Anti-IL-17F stain		
	Total cells in HPF* (mean $\pm$ SD)	IL 17A expressing cells in HPF (mean $\pm$ SD)	Proportion of IL-17A expressing cells (%, mean $\pm$ SD)	Total cells in HPF (mean $\pm$ SD)	IL 17F expressing cells in HPF (mean $\pm$ SD)	Proportion of IL-17F expressing cells (mean $\pm$ SD)
<b>Palmoplantar pustulosis (N=17)</b>						
Dermal cells	233.0 $\pm$ 127.58	16.1 $\pm$ 10.5	8.9 $\pm$ 5.8	224.7 $\pm$ 113.9	6.2 $\pm$ 7.7	2.3 $\pm$ 1.6
Intraepidermal pustular cells	393.6 $\pm$ 228.2	264.1 $\pm$ 205.4	66.1 $\pm$ 24.9	367.5 $\pm$ 206.9	5.7 $\pm$ 4.6	2.3 $\pm$ 3.0
<b>Psoriasis (N=3)</b>						
Dermal cells	292.0 $\pm$ 193.2	33.7 $\pm$ 24.6	11.2 $\pm$ 0.8	325.7 $\pm$ 128.4	11.0 $\pm$ 4.4	4.0 $\pm$ 2.5
Intraepidermal pustular cells	-	-	-	-	-	-
<b>Normal skin (N=2)</b>						
Dermal cells	25.0 $\pm$ 5.7	1.0 $\pm$ 0.0	4.1 $\pm$ 0.9	20.5 $\pm$ 2.1	0.0 $\pm$ 0.0	0.0 $\pm$ 0.0
Intraepidermal pustular cells	-	-	-	-	-	-

\* HPF: High power field

**Figure .** Histological image. (A) Hematoxylin and eosin staining of PPP. (B) Anti-IL-17A staining of PPP. (C) Anti-IL-17F staining of PPP. (D) Hematoxylin and eosin staining of PPP. (E) Anti-IL-17A staining of PPP. (F) Anti-IL-17F staining of PPP. (G) Hematoxylin and eosin staining of psoriasis. (H) Anti-IL-17A staining of psoriasis. (I) Anti-IL-17F staining of psoriasis. (X100 magnification)



**Abstract N°: 1642****Trends in prescribing acne medications: with a focus on the differences between dermatologists and dermatology residents**

Yasamin Kalantari<sup>\*1</sup>, Maryam Nasimi<sup>1</sup>, Robabeh Abedini<sup>1</sup>, Majid Mehrshadian<sup>1</sup>, Vahidehsadat Lajevardi<sup>1</sup>, Ifa Etesami<sup>1</sup>

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**Introduction & Objectives:**

Few studies have evaluated the trends in acne treatment. The aim of this study was to assess the differences between dermatologists and dermatology residents in prescribing acne medications.

**Materials & Methods:**

The reliability and validity of our questionnaire were approved by conducting a pilot study. Content validity index (CVI) was 0.7 and content validity ratio (CVR) ranged from 0.6 to 0.8. Our questionnaire obtained a Cronbach's alpha of 0.75.

**Results:**

A total of 126 participants were enrolled. Twenty-eight (22.2%) were residents and 98 (77.8%) were dermatology specialists. In children younger than 12, 112 (88.9%) preferred prescribing oral antibiotics. The most preferred medication in patients with liver dysfunction was doxycycline 58/126 (46.03%). In pregnant and lactating women, the most preferred local treatment was clindamycin 37/126 (29.36%) and the most common systematic treatment was azithromycin 62 (49.20%). In cases of isotretinoin indications, 78/126 (61.90%) preferred to prescribe low doses of isotretinoin (less than 0.5 mg/ kg/ day). Most of the dermatology residents 20/28 (71.43%) preferred to prescribe the cumulative doses of isotretinoin compared to other dermatologists (p-value: 0.028).

**Conclusion:**

In conclusion, while dermatologists can treat acne patients based on up-to-date guidelines, our study focuses on the controversial areas in treating acne patients. Our findings suggest that the most preferred acne treatment options for children under 12, women older than 20, patients with liver diseases, and pregnant women were antibiotics. Also, in women with evidence of hormonal dysfunction prescribing antibiotics along with hormonal therapy was more preferred by dermatologists. Also, most dermatologists prefer to prescribe low doses of isotretinoin with cumulative doses and it seems that this trend in prescribing isotretinoin and antibacterial agents will not change substantially in the future. Due to the low sample size, one cannot relate our findings to the whole population. Hence, more research in this field is recommended.

**Abstract N°: 1680**
**Association between IL-17 rs763780 and IL-17RA rs4819554 gene polymorphisms with response to biological drugs in psoriasis patients and beyond**

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**Introduction & Objectives:**

IL-17 family is one of the main actors in the pathogenesis of psoriasis. The aim of our study was to determine if the genetic polymorphism of IL-17F (rs763780) and IL-17RA (rs4819554) are potential risk loci for response to biological treatment or other clinical characteristics.

**Materials & Methods:**

Our study included 81 patients (moderate to severe psoriasis) who received biological for the first time. Four classes of biologics were used: anti-TNF- $\alpha$ , anti-IL23, anti-IL17, anti-IL-12/23. We formed two groups: 1st line biological treatment group (bio-naïve patients) and 2nd line biological treatment group (resistance to the first-line treatment).

The treatment efficiency was evaluated using PASI (Psoriasis Area Severity Index) score, recorded at baseline, weeks 12, 24, 36, and 48. Treatment responders were considered those with PASI75 at week 24 and PASI90 at week 48. Blood samples taken during the first examination were used to determine the presence of two single nucleotide polymorphisms: (SNPs) rs763780 in the IL17F gene and rs4819554 in the IL17RA gene. We also recorded: BMI, cigarettes/alcohol consumption, comorbidities, nail psoriasis.

**Results:**

The 1st line biological treatment was an anti-TNF- $\alpha$  agent (infliximab, adalimumab, etanercept). The group treated with a second-line biological treatment consisted of anti-TNF- $\alpha$ , anti-IL-23, anti-IL-12/23, anti-IL-17A agents.

*SNPs and treatment response:*
*1st line treatment*

When we analysed all biological agents together, no association was seen with rs763780 IL-17F or rs4819554 IL-17RA. Analysing each anti-TNF- $\alpha$  agent separately, we noticed that rs763780 IL-17F is associated with response to infliximab and adalimumab ( $p=0.04$  and  $p=0.0001$ , respectively), when evaluating the percentage reduction of baseline PASI. Thus, for infliximab-treated patients, the TT genotype was associated with a better response to treatment at 12 weeks, while the CT/CC genotype was associated with a much better response at 36 and 48 weeks. For adalimumab, the CT/CC genotype was associated with a good response at 12 weeks, while the TT

genotype carriers were more likely to be better responders at 36 and 48 weeks.

#### *2nd line treatment*

No statistical association was found between response to TNF- $\alpha$  inhibitors and rs763780 IL-17 or rs4819554 IL-17RA at 24 and 48 weeks. In patients treated with anti-IL17A and anti-IL23 or anti-IL12/23, the responders at 24 weeks maintained the same status at 48 weeks. The highest percentage of responders was in patients treated with anti-IL-17A agents (89.5%). In both groups, all patients with GA/AA genotype of rs4819554 IL-17RA were responders at 24 and 48 weeks.

#### *SNPs and clinical characteristics*

Polymorphism rs4819554 in IL-17RA gene is more frequently associated with nail psoriasis in patients with GG genotype, compared to those with GA/AA genotype ( $p=0.02$ ). We also found an association between the IL-17RA SNP and BMI; A allele carriers are more likely to be overweight, while those with GG genotype are more likely to be obese ( $p=0.05$ ).

#### **Conclusion:**

The rs763780 polymorphism in the IL-17F gene is associated with response to infliximab and adalimumab at week 36, 48 and rs4819554 in IL-17RA is associated with nail psoriasis and BMI. To our knowledge, this is the first study to evaluate those polymorphisms in psoriasis patients in Romania and southeastern Europe. The rs763780 polymorphism in the IL-17F SNP could be a promising biomarker for predicting the responders to anti-TNF- $\alpha$  therapies.

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**Abstract N°: 1705****Treatment of eosinophilic fasciitis with extracorporeal photopheresis: a case series**

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**Introduction & Objectives:** Eosinophilic fasciitis (EF) is a rare, fibrosing disease of the fasciae of the limbs. Its clinical spectrum depends on disease stage and includes symmetrical bilateral erythema, edema and induration of the extremities, collagenous thickening of the subcutaneous fasciae and possible concomitant blood eosinophilia and hypergammaglobulinaemia. Current treatment options include among others systemic steroids, other immunosuppressant drugs, such as methotrexate and UVA therapy. In many cases, due to prolonged refractory course of the disease, treatment still remains a challenge. Extracorporeal photopheresis (ECP) has been reported to be effective in recalcitrant EF cases in recently published case reports.

**Materials & Methods:** We report a case series of patients diagnosed with EF and treated with ECP over the past 15 years at our clinic. Patient records along with photographs, when available, were retrospectively reviewed and the following data were recorded: demographics, clinical presentation, histopathology, laboratory findings and response to treatment, defined as complete response (resolution of erythema and/or edema with no or minimal persistent induration), partial response (incomplete improvement of erythema, edema, and/or induration), or no response (lack of improvement).

**Results:** Eight patients diagnosed with EF based on clinical, laboratory and histopathological findings and treated with ECP were identified. The initial absolute peripheral blood eosinophil count was increased in five patients. One patient had a monoclonal gammopathy. ANA titers were negative or low in all patients. All eight patients received treatment with systemic corticosteroids. Four patients were additionally treated with UVA1 therapy, two with ceftriaxone, one with methotrexate, one with mycophenolate mofetil, one with cyclophosphamide. ECP cycles were initially performed every 2 to 4 weeks; after clinical improvement treatment intervals were prolonged up to 12 weeks. Three patients showed complete response to treatment and could therefore discontinue ECP after a median of 54 treatment cycles (range 54-84 cycles). Five patients are still undergoing ECP treatment at this time (median 38, range 8-68 treatment cycles), currently showing partial response.

**Conclusion:** To our knowledge, this is the largest reported cases series of EF treated with ECP to date. ECP with or without immunosuppressive agents presented a safe and effective treatment option for EF. Further studies in larger patient collectives are needed to confirm these findings and help issue standardized therapeutic recommendations in the future.



**Abstract N°: 1757**
**Psychometric validation of patient-reported outcome measures in generalized pustular psoriasis**

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**Introduction & Objectives:**

Generalized pustular psoriasis (GPP) is a chronic, rare, and potentially life-threatening disease with a high clinical burden and considerable adverse impact on quality of life. The Psoriasis Symptom Scale (PSS), pain-visual analogue scale (VAS), and Functional Assessment of Chronic Illness Therapy-Fatigue (FACIT-F) scale are widely used patient-reported outcome measures (PROMs) in dermatology. These PROMs were used as secondary endpoints in Effisayil 1 (NCT03782792), a randomised controlled study of spesolimab in patients with GPP. Here, we have used data from Effisayil 1 to evaluate the psychometric properties of the PSS, pain-VAS, and FACIT-F in the context of GPP.

**Materials & Methods:**

For multi-item measures, confirmatory factor analysis (CFA) and analyses of internal consistency reliability were performed using data from Day 8; internal consistency analyses also used baseline data. Test-retest reliability was assessed using change data from Days 3–4; convergent validity and known-groups validity were assessed at baseline and Day 8; and sensitivity to change analyses were performed using change data from baseline–Day 8. For test-retest reliability analyses, several definitions for a stable population were identified and tested using intraclass correlation coefficients (ICC); we present results for analyses where a stable population was defined using the change in Japanese Dermatological Association (JDA) GPP severity index part A (assessment of skin symptoms) from Day 3–4. For convergent validity analyses, correlations were assessed by Spearman's rank order coefficients. For known-groups validity analyses, group differences were determined by analysis of variance (ANOVA). For sensitivity to change analyses, differences in mean scores were tested by analysis of covariance (ANCOVA).

**Results:**

The factor structures of the PSS and FACIT-F in this cohort of patients with GPP (N=53), showed acceptable fit statistics for the comparative fit index (threshold  $\geq 0.90$ ; PSS, 1.00; FACIT-F, 0.92), and standardized root mean residual (threshold  $< 0.1$ ; PSS, 0.007; FACIT-F, 0.056). The root mean square error of approximation values met the threshold for acceptable fit ( $< 0.08$ ) for the PSS (0.000), but not the FACIT-F (0.126). Internal consistency was satisfactory for the PSS and FACIT-F, with Cronbach's alpha  $> 0.70$  for both scores at Day 8 (PSS, 0.92, FACIT-F, 0.95). The PSS, pain-VAS and FACIT-F demonstrated good test-retest reliability, with ICCs of 0.78, 0.87 and 0.70, respectively, reaching the acceptable threshold of 0.70. Convergent validity analyses at Day 8 showed the three PROMs had moderate correlations with most related measures, despite the lack of instruments assessing the same concepts. Moreover, the three PROMs could differentiate between known patient groups of varying disease status, as defined by related instruments. The PSS, pain-VAS, and FACIT-F could also detect change from baseline to Day

8, across several anchor categories for improvement. For instance, when the EQ-5D pain/discomfort item was used to group patients as 'worsened', 'no change', and 'improved', statistically significant changes were observed for all three PROMs ( $p < 0.0001$ ).

**Conclusion:**

Overall, these results support the reliability, validity, and responsiveness of the PSS, pain-VAS and FACIT-F in a GPP population, and validate their suitability for use as PROMs to assess the patient experience in GPP and inform treatment decisions.

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**Abstract N°: 1889****A curriculum of online education significantly improved physicians' knowledge and confidence in understanding the immunopathology, etiology and diagnosis of hidradenitis suppurativa**Elaine Bell<sup>\*1</sup>, Gregor Borut Jemec<sup>2</sup>, Alessia Piazza<sup>1</sup><sup>1</sup>WedMD Global, 14-17 Market Place, LONDON, United Kingdom, <sup>2</sup>Roskilde Hospital, Copenhagen, Denmark**Introduction & Objectives:**

Hidradenitis suppurativa (HS) is a chronic skin disorder characterized by recurrent abscesses, sinus tract formation and scarring. We assessed whether a curriculum of online activities could improve clinicians' knowledge, skills and confidence with regards to the immunopathology of HS, the burden of disease, and the diagnosis and management of patients.

**Materials & Methods:**

A curriculum of 5 activities was developed on HS. We collected data on 3 of these activities, consisting of a video lecture, an enduring video from a symposium and a round table discussion; data from the clinical practice assessment (which identified significant gaps in clinical knowledge, competence and confidence) were published previously<sup>1</sup>; data from the medical simulation activity will be reported separately. Data were collected from 9 May 2022 to 3 April 2023, with n numbers from 67–568 learners in each specialty. Educational effect was assessed with a repeated-pairs pre-/post-assessment; 3 multiple-choice, knowledge questions and 1 self-efficacy question were analyzed. Data were subsequently combined and analyzed by 3 themes (HS pathogenesis, burden of disease and assessment & diagnosis of HS) to provide a summative overview of the effect of the education across the combined activities. A McNemar's test was conducted to assess statistical significance of changes from pre- to post-assessment.

**Results:**

- 19,681 physicians from across the globe participated in the activities: 4,553 dermatologists, 1,096 emergency medicine (EM) physicians, 5,027 general practitioners (GPs), 6,955 surgeons and 772 obstetricians/gynecologists
- Data were collected for 3 key specialties who engage with HS patients: dermatologists, EM physicians and GPs
- Dermatologists and EM physicians demonstrated a statistically significant improvement in knowledge in 2 of the 3 learning themes (HS pathogenesis and burden of disease), all  $P < 0.05$
- GPs demonstrated a statistically significant improvement in knowledge across all 3 learning themes, all  $P < 0.01$
- The relative improvements in percentage of correct responses for learning themes demonstrating improved knowledge ranged from 22%–43%
- As a result of completing the activities, 42% of dermatologists, 43% of EM physicians and 50% of GPs reported increased confidence in understanding HS etiology and the impact of HS on patients' lives, and being able to diagnose HS

**Conclusion:**

These results highlight the benefits of a curriculum of education in helping physicians understand the etiology, impact and diagnosis of HS. Following on from education on these foundational topics, physicians would benefit

from future education on evolving best practice for the diagnosis, assessment and management of HS. Gains in this area would support clinicians' ability to translate knowledge into clinical practice in order to optimize outcomes for patients with this challenging condition.

## **Reference**

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**Abstract N°: 1898****Evaluation of the efficacy of a Mandarin extract-containing cream used in association with an Uncaria tomentosa-containing spray in rosacea**

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**Introduction & Objectives**

Rosacea is a chronic, inflammatory skin disease characterized by flushing, nontransient erythema, papules, pustules and telangiectasia which may be related to a genetic predisposition family and evolve under influence of various aggravating factors. Rosacea treatments include skin care and cosmetic treatments.

To assess the efficacy of the association both extracts we performed a clinical study in adults suffering from erythemato-telangiectasic rosacea.

**Materials & Methods**

Women suffering from erythemato-telangiectasia rosacea with nontransient erythema on cheeks with moderate to severe intensity and feeling unpleasant skin sensations on face with intensity  $\geq 3$  on a Visual Analog Scale (VAS from 0 to 10), have been enrolled in a monocentric, open-labelled study, randomized in cross over. Three periods were designed: Two 21-day study products application periods with 3 visits (D1, D15 and D22) during which patients applied products on the face, according to randomization, the Mandarin extract-containing cream alone or associated with the Uncaria tomentosa extract-containing spray twice a day (morning and evening), separated by a 28-day period during which patients did not apply the study products and used their usual skin care product. During the association phase, at least one additional spray application was performed during the day. All along the study, patients had to connect on an online platform to explain their feelings about the products. They also assessed unpleasant skin sensations and refreshing effect on a VAS after each application and had to illustrate the product's efficacy by collage of royalty-free photos. At each visit, we performed standardized cross-polarized photographs, investigator and patient's clinical assessment of rosacea severity on a 4-point grading scale, investigator clinical assessment of erythema on a 4-point grading scale and hydration index measurement with Corneometer®.

**Results**

23 women aged 28 to 63 years suffering from ETR were included in the study.

Patients felt a significant decrease of unpleasant skin sensations after 3 weeks of application of the cream alone or associated with the spray vs. baseline ( $p < 0.0001$  for two conditions).

A significant decrease of erythema was observed by investigator after 3 weeks of application of the cream alone or associated with the spray vs. baseline ( $p = 0.0032$  and  $p < 0.0001$  respectively).

The investigator also showed a significant efficacy of the cream alone or used in association with the spray on global rosacea severity after 3 weeks of application vs. baseline ( $p = 0.0032$  and  $p < 0.0001$  respectively).

The efficacy of the product's association on global rosacea severity was also significantly demonstrated by the patients after 3 weeks of application vs. baseline ( $p=0.0063$ ). Moreover, the product's association provided a global refreshing effect significantly greater compared to the cream alone ( $p<0.0001$ ) and a significant skin hydration after 1 and 3 weeks of application vs. baseline ( $p=0.0091$  and  $p=0.0167$ ). These results were completed by the observations on the online platform since most enrolled subjects preferred the association of products. Patients talked about a "More in-depth hydration" and an "on-demand freshness effect" thanks to the spray application.

## **Conclusion**

This study shows the efficacy of a mandarin extract-containing cream used in association with an uncaria tomentosa extract-containing spray on physical and functional signs of rosacea.

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**Abstract N°: 1936****Hyperpigmented rhinophyma: an unusual clinical and dermoscopic image**

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**Introduction & Objectives:**

Rhinophyma is a type of phymatous rosacea of the nose. It is manifested by sebaceous hyperplasia and increased thickness of the skin in the affected area. In most cases the diagnosis is clinical and does not require additional diagnostic procedures. Herein, we present an untypical case of hyperpigmented rhinophyma.

**Materials & Methods:**

A 53-year-old man (phototype II) was referred to Dermatology Department due to a slowly growing tumour of the nose with areas of focal hyperpigmentation. There was no history of associated comorbidities. Clinically, features of rosacea of the face were observed. Dermoscopy of the affected nasal area revealed follicular plugs, brown and grey pigmentation distributed around hair follicles (zig-zag pattern), brown and grey dots/globules as well as red structureless background. Histopathological examination allowed for the diagnosis of rhinophyma with signs of pigment incontinence. Local and systemic treatment of rosacea was introduced and the patient was scheduled for carbon dioxide laser treatment.

**Results:**

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**Conclusion:**

Dermoscopy may facilitate clinical diagnosis of skin neoplasia as well as cutaneous inflammatory disorders. Dermoscopic structures typical of rosacea include linear vessels arranged in a polygonal network (vascular polygons), follicular plugs/ dilated follicles, white/yellowish scales, orange-yellowish areas and follicular

pustules (papulopustular rosacea). Pigmentation was described on dermoscopy mainly in skin of color patients (most studies did not report the type of observed pigmented structures; other mentioned follicular annular pigmentation, honeycomb pattern, gray dots). We have not found a similar case of clinically pigmented rhinophyma in a patient with light skin phototype.

**Abstract N°: 1942**

**Generalized pustular psoriasis (GPP): Epidemiology and Care - Results from a Health Insurance Claims Data Base Analysis**

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<sup>1</sup>University medical center Hamburg Eppendorf, Institute for health services research in dermatology, Hamburg

**Introduction & Objectives:**

Generalised pustular psoriasis (GPP) is a rare skin disease with clinical symptoms caused by autoinflammation. Patients with GPP have a severely impaired quality of life. International data on the epidemiology of GPP show great differences and in Germany there are no current evaluations available so far.

**Objective:** The aim of this analysis was to record the epidemiology and care of GPP patients in Germany for the first time on the basis of SHI data.

**Materials & Methods:**

The data basis is a representative 40% sample of all insured persons of the German SHI system (DAK-Gesundheit) who were insured for at least one day between 2016 and 2020 (N=2.5 million). The annual prevalence and incidence of GPP (ICD-10 L40.1) were calculated for the years 2016 to 2019 using outpatient and inpatient data. Three case definitions were used for internal diagnosis validation. In addition, relevant comorbidities and the care of GPP patients were analysed.

**Results:**

In 2019, the annual prevalence of GPP ranged from 8 to 39 per 100 000 and the incidence ranged from 1 to 15 per 100 000 in 2019, depending on the case definition. GPP patients suffered most frequently from cardiovascular diseases as well as depression, eczema and cataract. General practitioners and dermatologists had the most frequent outpatient contact. 1.5% of identified people with GPP were treated as inpatients. In terms of treatments, topical agents were most commonly used in 2019 (52.7%), followed by systemic non-biologic agents (17.9%), systemic cortisone (15.1%) and systemic biologics (14.3%).

**Conclusion:**

This study provides a good overview of the occurrence of GPP in Germany, although the data have limitations (lack of clinical data). As GPP is a rare disease with limited clinical awareness, it is associated with challenges, including consistent descriptions of disease characteristics. Clinical data sources should be supplemented in the future to more accurately capture the epidemiology.





**Abstract N°: 2027**
**Improved Mortality and Morbidity Rate of SJS/TEN Using Triple Therapy**

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**Introduction & Objectives:**

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are rare, yet life-threatening diseases with mortality rates up to 90%, depending on severity. Both disorders are most frequently elicited by medications or infection. Beyond supportive care, cyclosporine, IVIG, and systemic corticosteroids have been discussed to improve clinical outcomes. Yet, the cohort mortality rate in published literature is reported to be between 20-50%.

**Materials & Methods:**

Retrospective data analysis of SJS, TEN, and SJS/TEN overlap from 2015-2022, was obtained using existing databases (Cerner) and initiated ongoing data acquisition in the main tertiary dermatology hospital in Qatar. Demographic variables were summarized using frequency distributions. Chi-square analysis was used to assess patterns and distribution of patients prior to versus after 2019. Univariable logistic regression was conducted to estimate the unadjusted odds ratio and the 95% confidence interval (CI). IBM SPSS (version 27, Armonk, New York, United States) was used for all data analyses. A p-value of 0.05 or less was considered statistically significant.

**Results:**

The retrospective analysis included a total of 95 patients. 51 (53.7%) were diagnosed with SJS, 28 (29.5%) were diagnosed with TEN, and 16 (16.8%) were diagnosed with SJS/TEN. Among all 95 patients, 56 (58.9%) patients had monotherapy or none, 15 (15.8%) had double therapy, and the remaining 24 (25.3%) patients has triple therapy.

Overall mortality was estimated at 8.4% (n=8). SCORTEN was categorized into two groups: 0-2 score points (n=76; 80.9%) and +3 score points (n=18; 19.1%). The SCORTEN was significantly associated with mortality (Fisher's Exact test p-value < 0.001). Simple logistic regression estimated an unadjusted odds ratio for mortality of 47.73 (95% CI 5.35-425.93; p-value < 0.001) for those with a SCORTEN of 3+ versus 0-2. Although Univariable analysis (Fisher's Exact test) did not show that triple therapy is associated with reduced mortality (p-value > 0.999), simple logistic regression estimated an unadjusted odds ratio (95% confidence interval [CI]) for mortality of 0.99 (95% CI 0.19-5.24; p-value = 0.986) for those with triple therapy. 12 patients (mean age 38) were treated with the above protocol. The database included full medical history including culprit drug, SCORTEN, and comorbidities. Classical treatment resulted in an SJS/TEN mortality rate of 57% (n=7 patients). Using triple therapy and our modified standard protocol ('CPG protocol') the mortality rate dropped by 57% (n=12 patients). Patients recovered faster and showed fewer short-term and long-term side effects (e.g. sepsis, renal failure, strictures).

**Conclusion:**

In summary, we show for the first time a significant reduction of mortality rate in SJS/TEN patients using a triple

therapy combined with a rapid standardized management protocol that reduced mortality by about 57%, and reduced complications such as renal failure as well as long-term complications such as sepsis, strictures, and renal failure. We propose multicenter large-scale studies to verify/falsify whether our protocol associated with a significant reduction in mortality and complications will lead to an improved outcome for patients undergoing this severe disease with yet high mortality.

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**Abstract N°: 2055**
**Validating the Lichen Planus Quality of Life Questionnaire (LPQoL) in a prospective lichen planus clinical trial.**

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**Introduction & Objectives:**

Lichen planus (LP) is an inflammatory dermatosis which affects multiple body sites including skin, mucosae (conjunctival, otic, oral, laryngeal, esophageal, nasal, urogenital, anal), scalp and nails. LP causes significant effects on patients' quality of life, especially those with genital or nail involvement. We developed a LP Quality of Life Questionnaire (LPQoL) as a LP patient-reported outcome measure (PROM) that addresses all potentially affected sites (1). The initial design was informed by an expert consortium and patient survey study, and compared results against those of the widely used Dermatology Life Quality Index (DLQI). Our objective was to further validate the LPQoL in a phase II prospective clinical trial on baricitinib (oral Janus kinase 1/2 inhibitor) for cutaneous LP.

**Materials & Methods:**

The study was approved by our institution's Institutional Review Board. Twelve patients with biopsy-proven cutaneous LP were treated on baricitinib 2 mg daily for 16 weeks, and LPQoL, Skindex-16 (2) and Physician Global Assessment (PGA) were recorded at week 0, 2, 4, 8, 12, 16 and 20. Notably patients with predominant non-cutaneous LP were excluded from the clinical trial. Repeated-measure correlation (3) was calculated to assess the correlations among LPQoL, Skindex-16 and PGA. ROC analysis (4) was implemented and area under the curve (AUC) was used to assess how accurately LPQoL and Skindex-16 can correctly identify responders (PGA 0 to 3) and non-responders (PGA 4 to 6).

**Results:**

LPQoL and Skindex-16 are highly correlated with each other ( $r = 0.77$ ). LPQoL and Skindex-16 had similar correlation with PGA (LPQoL  $r = 0.52$  and Skindex-16  $r = 0.48$ ). Skindex-16 performed better than LPQoL in predicting responders (Skindex-16 AUC = 81.5% vs. LPQoL AUC = 66.8%). In the original questionnaire pilot, LPQoL score was positively correlated with DLQI score ( $r = 0.79$ ;  $p < .001$ ) (1).

**Conclusion:**

In this retrospective patient survey study and prospective validation, we found our LPQoL correlated with DLQI and Skindex-16 in patients with LP. The LPQoL allows for a greater range of scoring, potentially conferring for increased sensitivity for changes in QoL. Limitations of the study include the trial excluded patients with predominant mucosal disease, which may explain its limited performance at predicting responders. The clinical trial compared LPQoL against Skindex-16, while the initial questionnaire compared against DLQI.

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**Abstract N°: 2240****Demographic, clinical, diagnostic and therapeutic characteristics of patients with pityriasis rubra pilaris in real clinical practice: a case series of 21 patients.**

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**Introduction & Objectives:**

Pityriasis rubra pilaris (PRP) is an idiopathic papulosquamous inflammatory dermatosis characterized by perifollicular hyperkeratotic papules that coalesce forming red-orange desquamative plaques, with islands of respected skin and palmoplantar keratoderma. The treatment of this dermatosis is a challenge in current daily clinical practice without high-quality evidence, based on individual cases and case series.

The main objective of our case series is to perform a descriptive review of the demographic, clinical, histological and treatment characteristics of patients with a definitive diagnosis of PRP in our health area.

**Materials & Methods:**

We have designed a retrospective descriptive study of patients diagnosed with PRP in our hospital from January 1, 1998 to December 31, 2022. Patients with a definitive clinical and pathological diagnosis of PRP were included. A total of 68 patients presented a clinical suspicion of PRP, of which only 36 met histological criteria, and of these, 21 patients presented a clinical evolution compatible with PRP.

**Results:**

The most frequent PRP subtype in our study was type 1 (adult classic), followed by subtype 3 (juvenile classic). A total of 4 patients debuted with erythroderma, all of them corresponding to subtype 1. 76% of patients presented palmoplantar keratoderma and 33% onychopathy. In the histological study, the most frequent pattern was parakeratosis and alternating orthokeratosis, present in 95% of cases.

Among the most used treatments, topical corticosteroid therapy was the most used one, with a complete response in 30% of cases and a marked improvement in 40% of them. Among systemic therapies, the most commonly used treatments were: acitretin, administered on 7 occasions with improvement and complete response in 43% and 14% of cases, respectively; and methotrexate, administered 5 times with 60% complete responses. Other treatments used were: cyclosporine, infliximab, isotretinoin, apremilast, vitamin D derivatives and phototherapy.

**Conclusion:**

We present the first Spanish case series of PRP, at least described in the current published literature, in which keratoderma seems to be a frequent feature (present in about 2/3 of patients) and highlighting methotrexate as a safe, effective and accessible therapeutic option in patients with PRP.



**Abstract N°: 2315****Impact of Biologic Therapy on Healthcare Service Admissions of Patients with Moderate to Severe Hidradenitis Suppurativa**

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**Introduction & Objectives:** Hidradenitis suppurativa (HS) is a chronic skin disease characterized by severe inflammation that dramatically impacts patients' quality of life. The disease poses a significant financial and time-consuming burden on the national healthcare system. The present study aims to demonstrate the impact of biological agent therapies on the entries of patients with HS to hospital outpatient clinics and emergency services, antibiotic and antidepressant usage, rest report requirement, and hospitalization rates.

**Materials & Methods:** This study included all patients diagnosed with moderate to severe HS who were being followed up and treated in our clinic and had received biologic therapy for at least one year. Demographic and medical characteristics of patients, and their hospital admission records, including one year before the initiation of biologic therapy and the following one-year period, were analyzed.

**Results:** This study included 53 (M/F: 6/47) patients with HS, with a mean age of  $39.5 \pm 10.8$  years (22-66 years). The mean age of disease onset and duration were  $29.7 \pm 2.3$  years (14-53 years) and  $12.7 \pm 2.09$  years (1-35 years), respectively. The median delay in diagnosis of HS among all patients was 66 months (range: 1-348, IQR: 135.25). When compared, no significant difference was observed in the total number of outpatient visits, hospitalization frequency, and mandatory rest report requirement during the one year before the initiation of biologic therapy and the following one-year period ( $p > 0.05$ ). On the contrary, the number of dermatology clinic admissions significantly increased ( $p: 0.004$ ), and antidepressant intake significantly decreased ( $p: 0.025$ ) after the initiation of biologics. Although there was a numerical decrease in the frequency of emergency department admissions and annual antibiotic prescriptions, it did not reach statistical significance ( $p > 0.05$ ).

**Conclusion:** Although the study includes a small number of patients with HS and analyses the impact of only one year of the biological therapy period, biologics alleviated the frequency of non-dermatology admissions leading to an irrelevant healthcare burden. Besides, these agents seem to reduce the need for antidepressants, considering their benefits in improving patients' quality of life. Further studies, including larger patient groups with long-term follow-up, are needed.\*\*

**Abstract N°: 2622****Novel orthorhombic, hexagonal lamellar platform technology for patients suffering from skin barrier associated disorders**Martin Albrecht<sup>1</sup><sup>1</sup>Leichlingen, R&D, Leichlingen, Germany**Introduction & Objectives:**

The skin barrier embedded in the stratum corneum consists various forms of lamellar structures. The coexistence of orthorhombic and hexagonal structures is indicative of a physiological barrier function. Skin barrier disorders, such as those associated with atopic dermatitis, ichthyosis or psoriasis, have a significant deficit of orthorhombic structures. Based on insights into regulating key mechanisms, a lamellar technology has been used to develop a regenerative platform for skin lipid barrier deficiency associated problems. By high pressure integration of long-chain, cuticular lipids into the saturated phosphatidylcholine membranes, a topical cream vehicle was developed which, in its structure, resembles the intercellular lipid arrangement of healthy Stratum Corneum intercellular lipids. The specific lamellar technology took the skin ultrastructure understanding into the center of solution approaches. In this context, the understanding of regulating key mechanisms are important factors in field of chronically caused barrier disorders.

**Materials & Methods:**

In a proof of concept study, subjects up to the age of 65 suffering from very dry skin conditions were included. Two of the specific lamellar preparations, differing in their lipid content (a>b) were tested to evaluate their epidermal skin barrier repair efficacy. The effect was determined before and after treatment with the topical products by different analytical methods. After Cyan acrylate stripping the samples were investigated by transmission electron microscope (TEM) and the intercellular lipid lamellae in the intercellular space of the stratum corneum were evaluated by quantity and quality. The skin lipid classes were analyzed by HPTLC.

**Results:**

The aim of the study was to evaluate the potential of care products A and B on the repair of the epidermal skin barrier of dry skin subjects after two weeks of treatment. This study was randomized and double blinded. The repair of the epidermal skin barrier was determined by transmission electron microscope to evaluated the integrity of intercellular lipids in the intercellular space of the stratum corneum and by HPTLC to quantify the corresponding barrier lipids and their changes.

It could be shown that the length of the lipid lamellae in the intercellular space after two weeks of treatment with care products A or B are comparable to healthy skin state. The repair shown in the TEM of the epidermal skin barrier correlates with a significant increase in Stratum corneum lipids. Free fatty acids, EOS and Ceramide Ceramide NP show a significant increase in product A or B-treated skin. No significant changes could be shown for cholesterol. The results were supported by TEM lamellae quantity and length observation. Both products lead to a complete repair the epidermal skin barrier. Treatment with product A or B changes the ratio of Cholesterol, free fatty acids and the ceramide EOS to each other.

**Conclusion:**

The cream vehicle without exogenous emulsifiers prevents an accumulation of these vesiculating substances in



skin and the associated damage. The results suggest, that by use of the lamellar technology, the extrinsic barrier strengthening of the intercellular lipid structure and the intrinsic barrier enhancement are supported. The ortho-as hexagonal lamellar platform is therefore suggested for barrier disruption associated skin problems.

**32<sup>ND</sup> EADV Congress 2023**  
**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 2704****EuroGuiDerm S3 Guideline Lichen sclerosus 2023**Gudula Kirtschig\*<sup>1</sup>, Lichen Sclerosus Guideline Development Group<sup>1</sup>Gesundheitszentrum Frauenfeld, Switzerland

**Introduction & Objectives:** Lichen sclerosus (LS) is an inflammatory skin disease affecting all ages that typically involves the anogenital site where it causes itching and soreness; it may lead to sexual and urinary dysfunction in females and males, however it may be asymptomatic. First signs of LS are usually a whitening of the genital skin, sometimes redness and oedema, fissuring, scarring, shrinkage, and fusion of structures may follow in its course; it is associated with an increased risk of genital cancer. LS has a huge impact on the quality of life of affected patients and it is important to raise more awareness of this not uncommon disease in order to diagnose and treat it early.

The guideline intends to provide guidance on the diagnostic of LS, highlight important aspects in the care of LS patients, generate recommendations and treatment algorithms on topical, interventional and surgical therapy, based on the latest evidence, provide guidance in the management of LS patients during pregnancy, provide guidance for the follow-up of patients with LS, and inform about new developments and potential research aspects.

**Materials & Methods:** The guideline was developed in accordance with the EuroGuiDerm Methods Manual v1.3 <https://www.edf.one/de/home/Guidelines/EDF-EuroGuiDerm.html>. The wording of the recommendations was standardised (as suggested by the GRADE Working Group). The *guideline* development group is comprised of 34 experts from 17 countries, including 5 patient representatives.

**Results:** Ultrapotent or potent topical corticosteroids in females and males, adults and children remain gold standard of care for genital lichen sclerosus; co-treatment with emollients is recommended. If standard treatment fails in males, a surgical intervention is recommended, complete circumcision may cure LS in males. UV light treatment is recommended for extragenital LS, however, there is limited scientific evidence. Topical calcineurin inhibitors are second line treatment. Laser treatment, using various wave lengths, is under investigation, it can currently not be recommended for the treatment of LS. Treatment with biologics is only reported in single cases.

**Conclusion:** LS has to be diagnosed and treated as early as possible in order to minimize sequelae like scarring and cancer development. Topical potent and ultrapotent corticosteroids are the gold standard of care; genital LS is often a lifelong disease and needs to be treated long-term.



**Abstract N°: 2847****Assessment of Programmed Cell Death Protein 1(PD-1) and Programmed Cell Death Ligand 1 (PD-L1) Tissue Expression Levels in Lichen Planus Patients: A Case-Control Study**Maha Fathy Elmasry\*<sup>1</sup><sup>1</sup>Faculty of Medicine, Cairo University, Dermatology, Cairo, Egypt

**Introduction & Objectives:** Programmed cell death 1 (PD-1) is an immune checkpoint cell surface protein with two ligands, PD-L1 and PD-L2. PD-1/PD-L1 interactions are essential for tolerance and protection from autoimmune attack. PD-1/PD-L1 checkpoint is implicated in multiple autoimmune diseases, and recently were suggested to have a role in lichen planus (LP) due to the high incidence of LP lesions as an adverse effect of PD-1/PD-L1 blockade by PD-1 inhibitors. The study's objective was to study and compare the levels of PD1 and PD-L1 in lesional and nonlesional skin of LP patients and compare these levels to normal healthy controls to assess their role in the pathogenesis of LP.

**Materials & Methods:** 30 LP patients and 30 age and sex matched controls were included. Full history and clinical examination were done and tissue levels of PD1 and PD-L1 were measured by ELISA from lesional and nonlesional biopsies.

**Results:** Levels of PD-1 and PD-L1 in lesional biopsies were significantly lower than in nonlesional biopsies ( $p < 0.001$ ) and significantly lower than control PD-1 and PD-L1 levels ( $p < 0.001$ ). Nonlesional PD-1 and PD-L1 levels were also significantly lower than control PD-1 and PD-L1 levels ( $p < 0.001$ ). A statistically significant

positive correlation ( $p < 0.001$ ) was found between lesional and nonlesional PD-1 levels ( $r = 0.885$ ). Also, a statistically significant positive correlation ( $p < 0.001$ ) was found between lesional and nonlesional PD-L1 levels ( $r = 0.713$ ).

**Conclusion:** PD-1/PD-L1 were lower in the lesional skin biopsies of LP patients in comparison to nonlesional skin biopsies and controls, based on these findings, PD-1/PD-L1 may be involved in the pathogenesis of LP.



**Abstract N°: 3008****Efficacy and Safety of Risankizumab for the Treatment of Moderate-to-Severe Palmoplantar Pustulosis in Japanese Patients: 68-week Data From the Phase 3 JumPPP Study**

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**Introduction & Objectives:**

Risankizumab demonstrated efficacy vs placebo at week 16 in the phase 3 JumPPP study (NCT04451720) evaluating efficacy and safety of risankizumab in Japanese adults with palmoplantar pustulosis (PPP); herein we report 68-week results.

**Materials & Methods:**

Patients were randomized (1:1) to risankizumab 150 mg or placebo for 16 weeks; all patients received risankizumab thereafter. Efficacy (Palmoplantar Pustulosis Area and Severity Index [PPPASI] change from baseline, modified Bath Ankylosing Spondylitis Disease Activity Index [mBASDAI] change from baseline in patients with baseline pustulotic arthro-osteitis, and achievement of  $\geq 50\%/75\%$  improvement in PPPASI [PPPASI50/75]) through 68 weeks and safety (cutoff September 26, 2022) were assessed.

**Results:**

Among 119 patients enrolled (risankizumab, n=61; placebo, n=58), numerical improvements in efficacy outcomes were generally observed from weeks 16–68. At week 68, the least squares mean (95% CI) change from baseline in the continuous risankizumab;placebo-to-risankizumab groups was  $-20.6$  ( $-22.6$ ,  $-18.6$ );  $-21.8$  ( $-23.8$ ,  $-19.8$ ) for PPPASI and  $-1.0$  ( $-2.1$ ,  $0$ );  $-2.5$  ( $-3.6$ ,  $-1.4$ ) for mBASDAI. At week 68, 87.0%;90.9% and 57.4%;69.1% of patients in the continuous risankizumab;placebo-to-risankizumab groups achieved PPPASI 50 and PPASI 75, respectively. The safety profile was consistent with previous psoriasis studies.

**Conclusion:**

Long-term risankizumab treatment provided improved efficacy and was well tolerated in Japanese patients with PPP.

**Abstract N°: 3132****Overlap with lichen pigmentosus and lichen nitidus in a child**

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**Overlap with lichen pigmentosus and lichen nitidus in a child****Introduction**

Lichen pigmentosus(LPP) is a rare variety of lichen planus (LP). Its occurrence in children is exceptional.

Lichen nitidus(LN) is an uncommon benign inflammatory eruption most prevalent amongst school-age children.

We report a rare case of pediatric overlap with LN and LPP.

**Case report**

A 3 year-old boy presented with a 5-month history of asymptomatic squamous brown macules, on the forehead, trunk, back and limbs. Dermoscopy revealed distribution of globules and points in a reticular pattern on an erythematous background. Another type of lesions was objectified on the back of the hands and forearms; he had well defined, pinhead-sized, skin-colored, glistening papules. Dermoscopy revealed multiple, white, well-circumscribed, circular areas with a brown shadow reflected through these white circles. Skin biopsy from a pigmented macule of the back demonstrated lichenoid interface dermatitis with pigmentary incontinence. The diagnosis of both LPP and LN was made. The patient was started on dermocorticoids topically with a good improvement after 1 month.

**Discussion**

LPP is considered as a rare variant of LP, it often occurs in middle-aged patients with high skin phototypes. In a systematic literature review of pediatric LP including 985 patients, only 22(2.2%) had LPP. In another serie of pediatric indian patients with LP, only 2.8% were detected with LPP. LPP presents, in both photoexposed and photoprotected areas, brown-gray macules. Its etiology remains unclear but a genetic predisposition for children of Arab, Afro-Caribbean and Afro-Caribbean and Indian origin has been suggested. LPP has been associated with hepatitis C virus, it can also coexist with other variants of LP such as frontal fibrosing alopecia, lichen planopilaris, oral lichen planus, and classic lichen planus. It has been associated with endocrinopathies and autoimmune diseases such as vitiligo and lupus erythematosus.

Lichenoid eruptions are a various group of inflammatory dermatoses characterized histologically by a lichenoid tissue reaction, including LN, lichenoid drug eruption, lichen planus-like keratosis, lichen striatus, keratosis lichenoides chronica, graft versus host disease... Some of them are mostly seen in the pediatric age group, such as LN. It is characterized clinically by multiple shiny, flesh colored, pinhead sized, dome shaped papules, generally localized in the genital area, upper limbs, and trunk. Pediatric series of LN found associations with other skin conditions such as lichen striatus, psoriasis, atopy, erythema nodosum, segmental vitiligo, and lichen spinulosus.

Few reports described associations between LP and LN. LN was found associated with classic LP with oral involvement in an adolescent of 17 years in a case report. Another case of 33-year old woman with oral LP and LN resistant to methylprednisone and topical tacrolimus improved by cyclosporin was reported. An author has shown

beneficial response to treatment with etretinate in both LP and LN. Two other cases of generalized LN and LP in 10 and 27 year-old males have been published. Another author reported a patient with a history of Crohn disease who developed both LP and LN in addition to erythema nodosum lesions.

To our knowledge, none of them reported an overlap with LN and LPP in a child as in our case.

### **Conclusion**

This co-occurrence of LN and LP may support the theory that both conditions may be precipitated by a common unidentified etiologic factor.

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**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 3146**
**Study Design and Rationale for the Efficacy and Safety Assessment of Daxdilimab, a Selective Plasmacytoid Dendritic Cell Depleter, in a Phase 2 Trial of Patients with Moderate-to-Severe Primary Discoid Lupus Erythematosus**

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**Introduction & Objectives:**

Daxdilimab (DAX) is an IgG1 $\lambda$  afucosylated monoclonal antibody specific for immunoglobulin-like transcript 7 (ILT7), a cell-surface protein that is exclusively expressed on plasmacytoid dendritic cells (pDCs). DAX binds to ILT7 on the surface of pDCs, resulting in their depletion via antibody-dependent cellular cytotoxicity. Several autoimmune disorders, including discoid lupus erythematosus (DLE), show marked enrichment of pDCs and interferon activity in affected tissue. DLE is considered the most challenging scarifying skin manifestation to treat for which a successful therapy does not currently exist.

**Materials & Methods:**

This is a 60-week Phase 2, multicenter, randomized, double-blind, placebo-controlled, parallel-group trial to investigate the efficacy and safety of DAX in reducing disease activity in adult participants with moderate-to-severe primary DLE refractory to standard of care therapy (NCT05591222). Approximately 99 participants will be randomized in a ratio of 1:1:1 (33 participants per group) to receive subcutaneous injection of DAX arm 1, DAX arm 2, or placebo. Administration of trial intervention will occur every four weeks starting from Day 1 through Week 44. After week 24 all participants will be receiving DAX, including those assigned to the placebo arm, for the remainder of the 48-week treatment period.

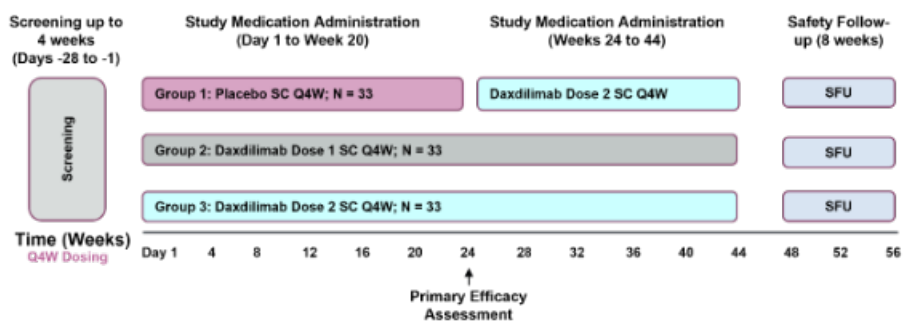
**Results:**

The primary efficacy endpoint is the mean change in Cutaneous Lupus Erythematosus Disease Area and Severity Index-Activity (CLASI-A) score from baseline to Week 24. Secondary efficacy endpoints include the proportion of participants who achieve 0 or 1 on the Cutaneous Lupus Activity Investigator's Global Assessment (CLA-IGA) scale at Week 24 (5-point Likert scale [0-4]), proportion of participants who achieve a  $\geq 50\%$  reduction in CLASI-A score from baseline at Week 24, and mean change in the Score of Activity and Damage in Discoid Lupus Erythematosus (SADDLE) from baseline to Week 24. Safety and tolerability of DAX will be assessed via the incidence of adverse events (AEs), serious AEs, and AEs of special interest. Pharmacokinetics and effects on pharmacodynamics and other biomarkers of interest will also be assessed.

**Conclusion:**

There is a significant unmet need for novel, fast-acting, and safe new therapies to reduce disease activity and damage and improve the quality of life for patients living with DLE. This is a proof-of-concept study that aims to evaluate a potentially new therapy in subjects with DLE.





**Figure.** Study schematic.\*\* N, total population; Q4W, once every 4 weeks; SC, subcutaneously; SFU, safety follow-up

32<sup>ND</sup> EADV Congress 2023  
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**Abstract N°: 3609**
**An unusual presentation of pyostomatitis-pyodermatitis vegetans with ocular involvement: a case report**

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**Introduction:**

Pyodermatitis-pyostomatitis vegetans (PPV) is a rare inflammatory dermatosis. There is a strong association between PPV and inflammatory bowel disease (IBD). Herein, we report an uncommon presentation of PPV with severe ocular involvement in a patient being followed for active ulcerative colitis which is treated by adalimumab.

**Case report:**

A 77-year-old female with a history of UC on mesalazine for three years and adalimumab for three weeks was admitted for a severe exacerbation of her disease. Clinical examination revealed a febrile patient with erythematous nodular and papulopustular lesions, located on the thighs, legs, neck, and external auditory canal which progressed to erosive vegetative lesions in the axillary and inguinal folds and scalp. There was a bilateral oedema with severe ulceration and purulent secretions along the edges of the eyelids. Visual acuity was 10/10 bilaterally. Oral mucosa was the site of flat pustules and superficial erosions in a linear pattern with a "snail track" appearance. Several diagnoses were discussed: pyoderma-pyostomatitis vegetans, sweet's syndrome, pustular pyoderma gangrenosum, neutrophilic hidradenitis secondary to adalimumab, pemphigus vegetans. Laboratory assessments showed leukocytosis with PNN predominance, and an inflammatory biological syndrome. Autoimmune antibodies study and viral serologies for HIV and hepatitis virus were negative. Histopathological examination of a pustular lesion revealed intraepithelial abscess and an inflammatory infiltrate rich in neutrophils in a mildly keratotic epidermis and dermis. Direct and indirect immunofluorescence were negative. Based on clinical and histological results, diagnosis of PPV was made. Treatment with topical corticosteroids once daily was initiated in association with treatment of her UC leading to a marked improvement of her condition within 5 days.

**Discussion:**

Initially described in 1898 by Hallopeau, PPV is a rare inflammatory neutrophilic dermatosis that is frequently associated with IBD, particularly UC. It commonly affects adults in the third decade of life with male predominance. Paradoxically, our patient is a 77-year-old woman. PPV is characterized by multiple small pustules on an erythematous base, producing a classic "snail tracks" pattern, as observed in our patient, and progressing to erosions, ulceration, and suppuration of vegetating pustules involving the oral cavity and other mucosal membranes such as the vaginal, and nasal mucosa. The ocular involvement remains exceptional and has been described in only a few cases. Histologically, PPV is characterized by pseudoepitheliomatous hyperplasia with intraepithelial microabscesses, eosinophilic spongiosis, and inflammatory infiltrates of neutrophils, eosinophils, lymphocytes, and plasmacytes. Direct and indirect immunofluorescence are usually negative. PPV is often present in IBD patients who have recently stopped medications or are functionally untreated. Our patient was under mesalazine and adalimumab. Treatment options vary depending on the severity and presence of underlying diseases, but corticosteroids, both systemic and topical, are commonly used and considered effective in many cases. Other reported successful treatments include topical tacrolimus, dapsone, azathioprine, mycophenolate mofetil, cyclosporine, and tumor necrosis factor inhibitors. Control of the underlying IBD is also crucial for treating

PPV.

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11 OCTOBER - 14 OCTOBER 2023  
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**Abstract N°: 3630**
**Antimicrobial peptides upregulation in human skin in contact with *S.aureus* and *C.acnes* RT4/RT5, in vitro bacteria's microbiofilm modulation of *C.acnes*.**

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**Introduction & Objectives:**

Normal microbiota of the human skin\*\* is an ecosystem participating to the protection of microbial communities. In skin inflammatory diseases as acne, atopic dermatitis, rosacea the imbalance of microbiota can induce changes within the "interactomes" microbes-host-microbes (1-4). Microbiota's changes are induced by complex mechanisms leading aggravation of chronic inflammatory skin diseases (5-7).

The expression of antimicrobial peptide hBD2 was assessed in normal human skin *ex vivo* submitted to microbial ligands from *C.acnes*, or *S.aureus*, the effects of patented vegetal extract (TLR2-Regul<sup>TM</sup>) were assessed in the same *ex vivo* model. 2. The *in vitro* modulation by a patented polysaccharide (MPA-Regul<sup>TM</sup>) on the micro-biofilm of *C.acnes* ribotypes RT4 and RT5 were also assessed.

**Materials & Methods:**

1.Normal human skin explants (NHSE) were submitted to microbial ligands from *C.acnes* and respectively from *S.aureus*. NHSE were then incubated in absence (control) or in presence of a monoclonal antibody anti-TLR2 or treated by an emulsion formulated with TLR2-Regul<sup>TM</sup> or by its vehicle (control). At 1h were added ligands from *C. acnes* or from *S. aureus*. The dosage of hBD2 was performed (ELISA). 2. Immunofluorescence *in vitro* assessment was made on the micro-biofilm of *C.acnes* RT4 and RT5 in the absence and in presence of increasing doses of patent MPA-Regul<sup>TM</sup>.

**Results:**

1.Microbial ligands from *C.acnes* and *S.aureus* induced a significant decrease hBD2 expression in skin explants, compared to control skin ( $p < 0.001$ ). Skin explants pre-treated by TLR2-Regul<sup>TM</sup> in contact with ligands of *C.acnes* or *S.aureus* had a significant increased expression of antimicrobial peptides hBD2 - compared to control. 2. Immunofluorescence assessment on transparent plaques of the micro-biofilm of *C.acnes* RT4 / RT5 showed a dose-dependent MPA-Regul<sup>TM</sup> decrease of micro-biofilm.

**Conclusion:**

Our *ex vivo* study showed a modulation of Toll Like Receptor 2 by stimulating human betadefensins-2 by a patented vegetal extract TLR2-Regul<sup>TM</sup>. The micro-biofilm of *C.acnes* ribotypes RT4 and RT5 was decreased dose-dependent by the MPA-Regul<sup>TM</sup> patent.

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**32<sup>ND</sup> EADV Congress 2023**  
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**Abstract N°: 3641**
**Keratosis lichenoides chronica: a diagnostic and therapeutic dilemma**

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**Introduction & Objectives:**

Keratosis lichenoides chronica (KLC) is a rare mucocutaneous disorder of unknown etiology. Characteristically, KLC presents as violaceous lichenoid hyperkeratotic papules typically arranged in a linear and reticulate pattern and most commonly found in a symmetrical distribution on the extremities and the trunk. Treatment can be challenging, as KLC is notoriously refractory to many therapies. Here, we present a patient with an extensive case of the disease, of difficult diagnosis who had partial response to Acitretin.

**Results:**

A 35-year-old man was referred to our department with complaints of asymptomatic skin lesions that had been present for 9 months. The lesions had first appeared suddenly on his trunk and progressed to involve the extremities, face and buttocks. The patient's history was unremarkable and he denied taking any medications. No family history of similar eruption was found.

Physical examination showed multiple violaceous, thick, scaly to hyperkeratotic papules and plaques, typically arranged in a linear and reticulated configurations and symmetrically distributed on the flexor aspects of the forearms. In addition, aphthous lesions in the oral cavity and ulcers on the penis and scrotum were present. There was a seborrheic dermatitis-like eruption on his face. The scalp, palms, soles, and nails were not involved. Systemic examination did not reveal any abnormal finding.

Relevant laboratory tests, including serologic tests for human immunodeficiency virus, hepatitis viruses and syphilis were within normal limits. Complete blood cell count, erythrocyte sedimentation rate, liver function tests, lipid battery, protein, and urinalysis were also normal. -scan of neck-thorax-abdomen showed no abnormalities

The first anatopathological examination pointed to Folliculitis. At that point, we started treatment with Tetracyclines without any response. The second and third biopsy conclude to Pytiriasis lichenoid and later to lichenoid dermatitis, for whom we start topical and systemic corticosteroids but without benefit and the skin lesions did not subside and they progressively worsened.

Another punch biopsy specimen obtained from lichenoid plaques after 6 months showed almost the same findings as before ( Folliculitis). One year later a new anatopathological examination showed parakeratosis and epidermal acanthosis, with areas of vacuolar alteration of the basal layer and numerous necrotic keratinocytes. In the superficial dermis we observed and abundant inflammatory infiltrates including lymphocytes and plasma cells.

Our patient's disease was refractory to various treatments including high-potency topical steroids, salicylic acid, antihistamines, tetracyclines, erythromycin and UVB therapy. We also tried isotretinoin combined with MTX but without response. A short course of oral steroid was tried without much improvement.

Transient improvement was only obtained with acitretin. The patient is currently on acitretin.

**Conclusion:**

The diagnosis of keratosis lichenoides chronica was based on typical clinical picture, repeated biopsies and histopathological findings, course of the disease and poor response to any therapy. Despite being a rare skin disease, it is important for dermatologist to become familiar with KLC in order to treat early and avoid complications of the disease.

**32<sup>ND</sup> EADV Congress 2023**  
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**Abstract N°: 3889**
**lichen nitidus on the penis : a case report with its dermoscopic features**

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**Introduction & Objectives:**

Skin lesions on the penis can cause panic and mental stress. Under the circumstances, a correct diagnosis by a dermatologist is essential. However, lichen nitidus on the genitalia is commonly misdiagnosed.

**Materials & Methods:**

We report a case of lichen nitidus on the penis with dermoscopic findings.

**Results:**

A 41-year-old man presented with multiple, pinhead-sized papules on his penis. These lesions first appeared 3 months earlier, the number of lesions has increased\*\* slowly. He reports no other associated systemic symptoms and neither history of sexually transmitted disease. Clinical examination revealed many tiny, smooth, translucent, and shiny skin-colored papules affecting nearly all the glans penis and

prepuce. There were no mucosal or nail lesions. Polarizing dermoscopy showed roundish, well-defined, hypopigmented areas, brown shadow within white circles, and whitish rosettes were noted in the center of some hypopigmented lesions. A skin biopsy sample from the penis was taken for histopathological examination and found well-circumscribed infiltrate of lymphocytes and histiocytes expanding the papillary dermis, enclosed by the rete ridges in a "ball and claw" configuration. These clinical and histopathological findings suggested a diagnosis of penile lichen nitidus. Tacrolimus cream was used topically, and the skin lesions of the patient significantly reduced after 2 months

**Conclusion:**

Lichen nitidus, first named by Pinkus, is a rare chronic inflammatory disease. The cause of its pathogenesis is Unknown. Lichen nitidus most commonly affects children and young adults. The typical skin lesions of lichen nitidus are shiny, flat-topped, skin-colored, pinhead-sized papules, which are usually arranged in groups. This dermatosis is typically asymptomatic but can be associated with pruritus. The most frequently affected sites are the forearms, abdomen, chest, buttocks, and penis. Lichen nitidus on the penis can be confusing to physicians and is often treated as a cutaneous viral infection, such as condyloma acuminatum. Many penile lesions also mimic lichen nitidus and should thus be differentiated; these include pearly penile papules, molluscum contagiosum, herpetic infection, nodular scabies, and Bowenoid papulosis.

The first dermoscopic description of lichen nitidus dates back to 2015. Dermatoscopic examination typically reveals roundish, well-defined, white areas devoid of physiological skin markings, which is a very relevant

dermoscopic clue. A brown shadow within white circles, radial ridges, central depression, peripheral scaling, ill-defined hypopigmentation, diffuse erythema, and linear vessels were also been reported. however, the rosettes found in our case were never reported in the literature.

The histopathologic findings of LN are fairly characteristic: lymphohistiocytic inflammatory infiltrate lying in close proximity to the epidermis. The overlying epidermis is flattened and parakeratotic. The rete ridges extend downward and seem to hug the inflammatory infiltrate, giving a characteristic “ball in claw appearance.”

Lichen nitidus is a rare disease for which a physical examination is often sufficient to diagnose it. In unclear situations, a skin biopsy is taken for histopathological examination, and dermatoscopy can be a valuable auxiliary examination facilitating diagnosis. we have also reported through this observation a new dermoscopic finding of lichen nitidus.

**Abstract N°: 3905****Wells Syndrome mimicking angioedema and revealing a Multiple Myeloma: a case report.**

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**Introduction**

Wells syndrome (WS), also referred to as eosinophilic cellulitis, is a rare eosinophilic dermatosis, characterized by great clinical variability and suggestive histologic features. Its diagnosis is based on typical clinical signs, including infiltrated erythematous plaques on the limbs, trunk, and chest. Localization of the facial area is rare. We report an unusual feature on the face mimicking angioedema that was successfully treated with Dapsone.

**Materials & Methods****Results:**

A 60-year-old man presented recurrent episodes of angioedema for 4 years, which were resolved with short-term corticosteroid therapy. Clinical examination revealed an infiltrated plaque on the face and the right ear, and an itching inflammatory annular lesion of the left upper limb. He denied any associated symptoms, drug intake, or insect bites. The biological assessment showed an inflammatory syndrome, without blood hypereosinophilia. The anatomopathological analysis described a perivascular and interstitial inflammatory eosinophilic infiltrate essentially with collagen fibers surrounded by eosinophils (flame image) allowing the diagnosis of WS. Antinuclear antibodies, anti-DNA as well as chest radiography were normal. The paraneoplastic assessment revealed an IgG Kappa monoclonal gammopathy, a serum IgG level > 30g/L, and the presence of dystrophic plasma cells on the myelogram correlated to smoldering multiple myeloma. The patient received oral corticosteroids at a dose of 1 mg/kg/day with a transitional regression of the skin lesions. After the incomplete response, Dapsone was initiated at 50 mg to 100 mg daily with significant improvement.

**Discussion:**

The diagnosis of WS is challenging due to the lack of specific signs; it is based on suggestive clinical, biological, and histological features. During the course of the disease, patients present recurrent episodes of acute pruritic dermatitis, persistent urticarial eruptions, or/and painful edematous swellings. Blood hypereosinophilia is frequent but not permanent. While histopathology is useful to establish the diagnosis, it is not fully specific of WS. So far, the etiology remains unknown. Authors of small case series and individual case reports have suggested possible associations with a multitude of disorders including parasitic infections, drug allergies, autoimmune diseases, hematological disorders, and solid tumors. The management of WS is complex and presents a therapeutic challenge. The particularity of our observation lies in its misleading clinical presentation: to our knowledge, WS mimicking angioedema has been described only once in the literature. Moreover, this is the first described case of a Smoldering Multiple Myeloma revealed by Wells syndrome.

**Abstract N°: 3926**
**Sweet syndrome associated with cellulitis masquerading giant cellulitis-like Sweet syndrome.**

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**Introduction & Objectives:**

Sweet syndrome (SS) is a neutrophilic dermatosis which has been associated with multiples inflammatory autoimmune diseases, infections and malignancies. But the association with cellulitis remains very uncommon. We present a case of Sweet syndrome associated with cellulitis.

**Materials & Methods:**
**Results:**

A 74-year-old woman, without any past medical history, presented to our department with persistent cellulitis and progressive appearance, 4 days after the cellulitis' occurrence, of painful nodules and plaques in both limbs. She had previously been treated for cellulitis with oral amoxicillin-clavulanic acid 1g three times a day with slight improvement. Physical examination revealed erythema, edema, heat and pain in the right inferior limb. The redness had a well-demarcated but irregular border and spared portions of skin in an unpredictable pattern. The inflamed area was edematous. Moreover, the patient had multiple, infiltrated, painful, erythematous, violaceous nodules and plaques of variable sizes, measuring 1-3cm in diameter localized in both limbs.

A skin biopsy was realized and the pathology report was consistent with a diagnosis of Sweet syndrome associated with infection signs asserting the\*\* clinical appearance of the association Sweet syndrome-cellulitis.

The patient was prescribed intra venous amoxicillin-clavulanic acid 1gx3/day with colchicine 1mg/day with good resolution.

**Conclusion:**

Although the etiology of SS is still unknown, it is postulated to be a hypersensitivity reaction to infection, autoimmune disease, inflammatory bowel disease or malignancy. Recent case reports of Sweet syndrome in association with infections indicate the infections may be diagnosed 4–14 days before the appearance of the skin lesions of Sweet's, simultaneously or even subsequent to the skin eruption. In our case, we suggested that SS was probably secondary to cellulitis of the right inferior limb.

A drug-induced Sweet syndrome was considered a possibility, as our patient had been exposed to Amoxicillin/Clavulanic Acid just prior to the eruption. Drug-induced Sweet syndrome is rare and affects middle-aged women, with skin lesions usually appearing 5–7 days after first administration of the offending drug. Diagnostic criteria for drug-induced Sweet syndrome have been proposed. Our patient did not meet all five of the necessary criteria for drug-induced Sweet syndrome.

Giant cellulitis-like Sweet syndrome has also been discussed as a new variant of sweet syndrome, but the bacteriological tests and the histopathology results concluded that there was also a skin and soft tissue infection.

Our case reinforces all the studies suggesting that cellulitis, a frequent cutaneous infection, may be associated with SS, an unusual association in the literature



**Abstract N°: 3962****Efficacy with immunological evidence of upadacitinib in Chinese patients with moderate-to-severe atopic dermatitis: a single-centre prospective, real world study**Lu LI<sup>\*1</sup>, Naming Wu<sup>1</sup>, Chen Shen<sup>1</sup>, Juan Tao<sup>1</sup><sup>1</sup>#####, Wuhan, China**Introduction & Objectives:**

The efficacy and safety of upadacitinib in atopic dermatitis (AD) have been defined in clinical trials, but real-world data for Asian people, especially Chinese AD patients, is very limited. We aimed to assess the effectiveness and safety of upadacitinib in a real-world Chinese AD patient cohort.

**Materials & Methods:**

Prospective cohort study collected data on once daily upadacitinib-treated AD patients (15 mg) who completing at least 12 weeks of therapy in China. The patients were evaluated at baseline, 4, 8 and 12 weeks after first upadacitinib administration for multiple physician- and patient-reported outcome measures. The profiles of peripheral blood mononuclear cells and serum cytokines were investigated.

**Results:**

Eighteen patients showed rapid and marked response to upadacitinib with significant reduction of all disease severity scores since the first follow-up visit. At week 12, Eczema Area and Severity Index (EASI)-75, and EASI-90 response were observed in 38.9%, and 16.7% of patients, respectively. The achievement rate of PP-NRS improvement  $\geq 4$ -point from baseline was 61.1% and IGA 0/1 was 61.1% at week 12. In regards to the innate immune cells in the circulation, peripheral blood eosinophil counts were significantly decreased at week 4, 8 and 12 after upadacitinib treatment compared to baseline, and neutrophil as well as monocyte counts decreased at week 12. As for the T subsets, there was a decreasing tendency in the proportions of IL-22-producing CD4+ T cells. Besides, serum IL-22 level decreased significantly at week 4 after upadacitinib treatment. Eight patients reported adverse events, with acne being the most common.

**Conclusion:**

Upadacitinib demonstrated favorable efficacy in Chinese moderate-to-severe AD patients, prominent in the management of subjective symptoms, including pruritus and quality of life, which was further confirmed by eosinophil and other innate immune cell normalization. Besides, serum IL-22 level was consistent with a decreased tendency of IL-22-producing CD4+ T cells in the circulation.

**Abstract N°: 4082****Patient with follicular hyperkeratosis and hair loss. A new case of NAIAD (NLRP1- associated autoinflammation with arthritis and dyskeratosis) syndrome.**

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**Introduction & Objectives:**

A 30-year-old woman presented with a 2-year history of multiple scaly and pruritic lesions of two years' evolution on the trunk and face, as well as the progressive development of hair loss on the scalp. The patient presented partial improvement of the scaly lesions with ciclosporin but presented a rapid worsening after its suspension. The lesions had shown a complete lack of response to tralokinumab. Physical examination revealed erythematous scaly plaques with follicular hyperkeratosis affecting a large part of the body surface, including the facial region and scalp, with total alopecia. Trichoscopy showed yellowish horny plugs and perifollicular desquamation. Hyperkeratotic papules were present on the palms and soles.

**Materials & Methods:****Results:**

A skin biopsy showed multiple cornoid lamellae located in the infundibular portion of the hair follicles, with the presence of dyskeratotic keratinocytes and loss of the granular layer. Due to the clinico-histopathological findings described, with the suspicion of a possible autoinflammatory syndrome or a pityriasis rubra pilaris – related disorder, a genetic study was requested and the presence of a variant of uncertain significance of the NLRP1 gene, associated with NAIAD (NLRP1-associated autoinflammation with arthritis and dyskeratosis) was found. A significant improvement of the follicular involvement was observed after 2 months of low-dose isotretinoin.

**Conclusion:**

The NAIAD syndrome (NLRP1-associated autoinflammation with arthritis and dyskeratosis), first described in 2016, is characterised by joint and skin involvement. The role of the NLRP1 inflammasome appears key in keratinocyte and monocyte differentiation. Previously described cutaneous findings in this syndrome include follicular spicules, scaly plaques on trunk and extremities as well as palmar hyperkeratotic papules. Commonly described histology includes horny plugs, dyskeratosis and epidermal hyperplasia. We describe a case compatible with NAIAD with predominantly cutaneous symptoms. We share this case to be taken into account in the differential diagnosis of atypical desquamative disorders with predominantly follicular involvement.

**Abstract N°: 4099****Alterations in metabolite, lipid, and cytokine concentrations in plasma samples of psoriatic patients treated with either ixekizumab or risankizumab**

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**Introduction & Objectives:**

For the treatment of moderate-to-severe psoriasis various biologics targeting different key cytokines are available. Targeting either IL-17 or IL-23 might have different effect on metabolite, lipid and cytokine concentrations in plasma, which might be used as monitoring biomarkers for treatment response.

**Materials & Methods:**

Here, we present data from 46 patients treated with either ixekizumab or risankizumab. All patients are part of a real-world cohort recruited at the comprehensive center for inflammation medicine Lübeck. Plasma samples were collected before treatment initiation, as well as on 6 pre-defined timepoints within one year after baseline. 39 metabolites and 112 lipids were quantified by 1H-NMR. For quantification of cytokine levels, 92 proteins were analyzed using the OLink Target 96 Inflammation panel.

**Results:**

Of the 46 patients recruited, 25 received ixekizumab and 21 risankizumab. A total of 40 patients completed treatment after 52 weeks, while 6 patients did not respond to therapy. PCA analysis of both treatment groups at baseline showed similar distributions for metabolites, lipids and cytokines, indicating even distribution between both cohorts. When analyzing for alterations after therapy initiation we found that patients treated with either of both drugs displayed clear changes in metabolic, lipid, and cytokine profiles as seen by separated groups in PCA blots. Further, we analyzed for differences between responders and non-responders in and in between both treatment groups and found that 3 cytokines and 8 metabolites were significantly altered at baseline with additional changes throughout the therapy.

**Conclusion:**

Treatment monitoring and therapy response prediction are crucial in the treatment of psoriatic patients receiving biologics. Quantification of metabolites, lipids and cytokines from plasma display a feasible method to perform monitoring and prediction. In addition, obtained data from this study help to better understand downstream effects of initially elevated cytokines IL-17 and IL-23 in psoriatic patients.



**Abstract N°: 4111****Post surgical complications of hidradenitis suppurativa lesions treated with a novel multistep approach: a monocentric, retrospective study**

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**Introduction & Objectives:** Hidradenitis suppurativa (HS) is a chronic inflammatory disease of the hair follicle whose treatment often requires a surgical approach. The aim of our study was to evaluate the post-surgical complications following a new standard of surgical management including a preoperative step with pre-surgical mapping of lesions by Ultra High Frequency UltraSound (UHFUS) with a 70 MHz probe and a postoperative time based on the principles of HS Time-Inflammation/Infection-Moisture-Edges (TIME).

**Materials & Methods:** A single center retrospective study was conducted by the Department of Dermatology of Pisa, enrolling 26 patients with moderate and severe HS, refractory to previous medical and surgical therapies. All the patients were treated with wide surgical excision of lesions previously explored through a UHFUS evaluation using a 48 and a 70 MHz ultrasound probe. After surgery all patients were treated with secondary intention healing following the principles of HS-TIME. For each patient, we assessed the occurrence of early post-surgical complications performing follow-up visits every 6 months after surgery. The occurrence of delayed complications was then assessed in all the patients with an observation time longer than three months (n=23).

**Results:** There were no reported cases of post-surgical bleeding or hematoma occurrence, while 3/26 (11,5%) patients developed minor surgical site infection. The average severity of pain decreased from Numerical Rating Scale (NRS) of 5,3 immediately after surgery to 1,3 after 4 weeks. The average healing time was 33.3 (19,3-47,3) days, and only 5/26 (19.2%) patients reported a complete wound healing longer than 6 weeks. Focusing on delayed complications, 1/23 (4,3%) cases of hypertrophic scarring, 2/23 (8,7%) cases of reported dysesthesia and 2/23 (8.7%) cases of clinical relapse were reported. No cases of limited mobility at the surgery site were registered.

**Conclusion:** We demonstrated the efficacy of a novel surgical protocol including a preoperative US evaluation and a proper postoperative wound management. Even if further prospective studies are needed to validate the observed results, we conclude that the low recurrence rates and post-surgical complications confirmed that our proposed protocol would represent an effective strategy for the management of HS patients eligible for surgical therapy.

**Abstract N°: 4250****Characterization of facial seborrheic dermatitis using non-invasive multimodal methodology**

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**Introduction & Objectives:**

Facial Seborrheic dermatitis (SD) is a skin disorder characterized by the presence of erythematous, flaky and itchy skin. Its pathogenesis appears multifactorial with I) a disbalanced immune system, II) *Malassezia* driven microbial involvement and III) skin barrier perturbations.

To investigate whether inflammation, microbial involvement or skin barrier dysfunction is more pronounced in SD.

**Materials & Methods:**

The lesional and non-lesional skin of 37 patients with mild untreated facial SD was comprehensively and non-invasively assessed with optical coherence tomography (OCT) and standardized photography to image inflammation. Amplicon sequencing and *Malassezia* species identification was used for microbial profiling. The skin barrier was assessed by trans-epidermal water loss and ceramide profiling.

**Results:**

Significant increases in lesional skin for epidermal thickness, epidermal blood flow and superficial roughness were observed by OCT and 2D-photography indicated significantly more erythema. While the abundance of *Staphylococcus* was significantly increased in lesional skin, abundance of *Malassezia* and *Malassezia* species were not significantly different. Lesional skin showed a significantly impaired skin barrier with significant underlying skewed ceramide subclass composition, impaired chain elongation and increased chain unsaturation. Changes in the ceramide profile correlated with the degree of barrier impairment. Integrative analysis shows two distinct populations after stratifying for lesional and non-lesional skin with chain length and increased ceramide [NS] as most discriminating features.

**Conclusion:**

This study shows skin barrier dysfunction to be heavily implicated in SD. Barrier restoration might be a suitable target for future therapeutic options in the treatment of facial SD.

**Abstract N°: 4264**
**Dupilumab Improves Skin Lesions of Varying Clinical Morphologies in Adult Patients With Prurigo Nodularis**

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**Introduction & Objectives:** Prurigo nodularis (PN) is generally characterized by multiple localized or generalized, elevated, firm, and nodular lesions. However, PN is a heterogenous disease that can manifest different clinical morphologies that often coexist. The estimated number of lesions on the whole body is representative of the severity stage of the disease. This analysis reports the efficacy of dupilumab in adult patients with PN presenting different types of skin lesions using the Prurigo Activity and Severity (PAS) score.

**Materials & Methods:** LIBERTY-PN PRIME (NCT04183335) and PRIME2 (NCT04202679) were randomized, double-blind, multicenter, parallel-group, 24-week, phase 3 trials in adults with PN with  $\geq 20$  nodules and severe itch, inadequately controlled with topical prescription therapies or for whom these therapies are inadvisable. Patients received 300 mg dupilumab subcutaneously (600 mg loading dose; n = 153) or matched placebo (n = 158) every 2 weeks. The PAS score items 4 and 5b, measuring the distribution and exact number of lesions, respectively, were used. The number of lesions, type of lesions, exact number of lesions in a representative area, and the proportion of healed lesions are reported. Data from the 2 studies were pooled and comparisons analyzed using ANCOVA.

**Results:** The proportion of patients with > 100 lesions decreased from 34.0% vs 33.5% at baseline to 5.2% vs 19.0% at Week 24 for dupilumab vs placebo groups. Although most lesions were nodular lesions, papular, plaques, and ulcerated lesions were also present. The percentage of all type of lesions decreased from baseline to Week 24, except for papular in both groups and plaques in the placebo group (baseline to Week 24 for dupilumab vs placebo: nodular, 78.4% to 49.7% vs 75.3% to 41.1%; papular, 11.1% to 17.0% vs 14.6% to 20.3%; plaques, 6.5% to 3.3% vs 5.7% to 6.3%; ulcerated, 3.9% to 0.7% vs 3.8% to 1.3%). The exact number of lesions in a representative area (mean [standard error]) decreased from 26.3 (1.9) vs 25.8 (1.4) at baseline to 8.8 (1.2) vs 19.9 (1.9) at Week 24 for dupilumab vs placebo. At Week 24, 26.1% vs 7.6% of patients had completely healed lesions for dupilumab vs placebo, with 73.8% vs 36.1% of the patients achieving 50% or more healed lesions. Overall safety was consistent with the known safety profile of dupilumab.

**Conclusion:** More than three-quarters of patients had nodular lesions; however, patients also had papular lesions, plaques, and ulcerated lesions, demonstrating that these different morphologies often coexist. Overall, the number of lesions were numerically reduced for patients treated with dupilumab vs placebo, with more than 70% of the dupilumab-treated patients achieving half or more healed lesions vs less than 40% for patients receiving placebo over 24 weeks of treatment. These results show that PN is a heterogenous disease, and all types of lesions should be considered when monitoring patients with PN.

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**Abstract N°: 4399****Disseminated purpuric lesions in an adult: an atypical rare form of pityriasis rosea**

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**Introduction & Objectives:** Pityriasis rosea (PR) is a common, acute, and self-limited skin disease of unknown origin. Usually, it is well recognized by its characteristic clinical signs, such as the presence of a 'herald' patch and erythematous scaling macules, papules and plaques classically arranged in parallel to the Langer's lines typically on the trunk and proximal extremities with a configuration of a "Christmas tree" pattern. However, unusual clinical presentations can cause diagnostic difficulties. Atypical cases of pityriasis rosea reach up to 20% of all cases. Atypical features can be present either in the morphology or distribution of the lesions, their size, evolution, and symptoms. There are several reported atypical forms: vesicular PR, generalized papular PR, urticaria-like PR, pustular PR, lichenoid PR, inverse PR, erythema multiforme-like PR, follicular PR, giant PR, unilateral PR, acral PR and purpuric (hemorrhagic) PR.

Purpuric (hemorrhagic) PR is a very rare variant with incidence varying from 0.39% to 4.8%. Clinical characteristics are presented with macular purpuric lesions and petechiae which may appear in different regions (including palate).

**Materials & Methods:** Here we report a rare case of purpuric PR presented in a 24-year-old woman with a 4-week history of pruritic widespread eruption without therapeutical response to self-treatment using beclomethasone dipropionate, clotrimazole and gentamicin cream as well as topical terbinafine cream. During physical examination we observed multiple, oval purpuric macules and papules, 0.4–4 cm in diameter including possible herald patch on her trunk and extremities distributed as a "christmas tree" pattern on the trunk. Mucous membranes were normal. White blood cell count, erythrocyte sedimentation rate, prothrombin time, partial thromboplastin time, biochemical analysis and urinalysis were normal. A biopsy specimen from a lesion of the left buttock revealed thickened epidermis with acanthosis, parakeratosis, subepidermal perivascular lymphohistiocytic infiltrate, extravasated erythrocytes.

**Results:** Based on the clinical and histopathological findings the diagnosis of purpuric pityriasis rosea was made. We recommended to minimize skin irritation, use emollients and topical mometasone cream once daily to reduce itching. One month later the eruption completely regressed.

**Conclusion:** It is important to consider rare variants of PR to avoid unnecessary diagnostic tests in patients with this self-limiting skin disease.

**Keywords:** Atypical, Pityriasis, Pityriasis rosea, Purpuric lesions, Herald patch

**Abstract N°: 4473****Genital pyoderma gangrenosum: a misleading diagnosis**

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**Introduction & Objectives:**

Pyoderma gangrenosum (PG) is a rare inflammatory neutrophilic dermatosis whose diagnosis is difficult. Genital pyoderma gangrenosum is an even rarer entity mimicking infectious, neoplastic or inflammatory diseases. In more than 50% of cases, PG is associated with an underlying systemic disease. An erroneous diagnosis of this condition can lead to a therapeutic escalation, source of iatrogenesis and a delay in the diagnosis of an underlying disease, hence the importance of the knowledge of this pathology.

Herein, we report 3 cases of genital PG associated with systemic diseases.

**Materials & Methods:**

Case one was a 46-year-old man with a history of diabetes type 2, having undergone a right orchidectomy 12 years ago and having unprotected sexual intercourses who presented a painless ulcerative budding penile lesion destroying the glans and involving the urethral meatus, associated with a urethral fistula. The diagnosis of genital PG was retained based on Su and al criteria. The patient had no associated systemic disease apart from diabetes.

Case two was a 63-year-old patient, with a history of non-Hodgkin's lymphoma treated with chemotherapy who presented multiple painful scrotal ulcerations measuring less than 1 cm, with a clean background and regular borders. A skin biopsy with 3 fragments was performed, bacteriological and mycological cultures were negative, histology was in favor of pyoderma gangrenosum.

Both cases responded well to topical steroids.

Case three was a 70 year old female, recently diagnosed with acute myeloid leukemia who presented multiple painful ulcerations with swelling, undermined wound edges and blue-purple skin involving the vulva and inguinal region. Based on the clinical and histological findings, the diagnosis of a genital ulcerative PG was retained. Initially, the patient responded well to topical steroids but then presented a relapse. Thus, the patient was treated with oral corticosteroids 0,5mg/kg per day with a complete healing in 3 months.

**Results:**

Pyoderma gangrenosum is a rare neutrophilic dermatosis clinically characterized by erythematous nodules or pustules that tend to evolve into large ulcers with purplish, undermined borders. It usually affects the lower extremities, its localization in the genital region is rare.

Vulvar PG is extremely rare and easily misdiagnosed as a sexually transmitted disease or vulvar malignancy. To our knowledge, there have only been 23 documented cases of vulvar PG. There have been also rare reports of PG involving the penis and scrotum.

The cases presented herein are unique given the rare localization of PG in the genital area and the occurrence in this area without the involvement of other sites.

**Conclusion:**

These cases are presented to highlight the importance of keeping PG as a differential diagnosis of genital ulcers refractory to usual treatment modalities.

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**Abstract N°: 4484****Gastrointestinal symptoms in patients with seborrheic dermatitis in a Colombian dermatological center**

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**Introduction & Objectives:**

Seborrheic dermatitis (SD) is a chronic inflammatory condition characterized by a colonization of *Malassezia*, added to an immune response due to an excessive lipid secretion by the sebaceous glands, which could be partially consequential to dietary changes and gastrointestinal dysbiosis. It has been described that intestinal dysbiosis can be clinically suspected with the presence of alterations in bowel habit, flatulence, abdominal pain, dyspepsia and/or abdominal bloating. Our objective with this study is to describe gastrointestinal symptoms frequencies in SD patients in a dermatological center.

**Materials & Methods:**

A cross-sectional study was conducted between June 2021 and December 2022 in a dermatologic center in Bogota, Colombia. Data was obtained via medical records. Variables included age, sex, time of evolution and gastrointestinal symptoms. An univariate and bivariate analysis was performed and relative and absolute frequencies were estimated. Data was collected and analyzed with Microsoft Excel.

**Results:**

The majority of patients were men (59.25% n=112) out of 189 subjects. The most frequent age group was '20-29' years, with a mean of 23.78 years. Mean time of evolution was 10.35 months. The majority of patients had at least one symptom (87.83% n=166) and almost a quarter of them reported suffering at least four symptoms at a time (23.8% n=45).<sup>\*\*</sup> Males with at least one gastrointestinal symptom were 88.31% (n=98) compared to women with 87.50% (n=68). Regarding gastrointestinal symptoms, abdominal distension was the most frequent with 75.66% (n=143), followed by flatulences with 59.25% (n=112), change in bowel habits in 49.73% (n=94), dyspepsia with 41.07% (n=89) and abdominal pain with 31.74% (n=60).

**Conclusion:**

Dermatological research advances have made it possible to establish a link between the Western diet and the clinical manifestation of chronic inflammatory pathologies such as SD. It was possible to identify a high frequency of gastrointestinal symptoms in SD patients in our study. Additionally, as reported in the literature, males were the most affected by SD. We found no substantial differences between women and men regarding gastrointestinal symptoms. An inflammatory and gastrointestinal dysbiosis basis are the potential conjugates for these clinical manifestations. Further studies should explore the development of specific protocols and interventions, including probiotic therapies and dietary changes to achieve a holistic therapy.



**Abstract N°: 4594****Erosive lichen planus : An uncommon cause of toe web intertrigo**

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**Introduction & Objectives:**

Erosive lichen planus is a variant of lichen planus. It mainly affects the oral cavity and genitalia. However, it's rarely described on the skin. It is characterized by chronic and painful ulcerations.

**Materials & Methods:**

We report two cases of erosive lichen planus of toe web spaces.

**Results:**

The patients included in this study consulted our dermatology department in 2018 and 2022. They were two men aged 45 and 56. Both patients were immunocompetent. They presented for painful toe web intertrigo with a recurrent evolution. The dermatological examination objectified in the first patient a bilateral erosive intertrigo with an epidermal detachment exposing a red plaque extended to the soles. The second patient had an erosive intertrigo of the left foot with an erysipeloid reaction. Skin biopsy showed a band like lymphocytic infiltrate with degeneration of the basal layer and parakeratosis. The diagnosis of erosive lichen planus was retained. The evolution was favorable under systemic corticosteroid therapy at a dose of 0.5mg/kg daily.

**Conclusion:**

Erosive lichen planus of the skin is an uncommon condition. It is more prevalent in women and its prevalence increases with age. It's mainly described on the feet. It affects the soles and toes. It causes painful ulcerations and may lead to the loss of the toenails. Clinical presentation consists of erythematous plaques which develop into painful erosions. To the best of our knowledge, no case presented as intertrigo. Cicatricial alopecia may be present on the scalp. Buccal mucosa may also be affected. The clinical examination ruled out any other localization in our patients. This disease has a significant negative impact on quality of life. Several treatments such as dermocorticoids, topical tacrolimus, phototherapy and oral steroids have been used. Oral retinoid and other aggressive treatments such as immunomodulatory treatments are indicated in refractory forms. Oral prednisone was effective in our cases.



**Abstract N°: 4612**
**Bullous lichen planus: a case report**

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**Introduction:**

Lichen planus (LP) is a chronic lichenoid inflammatory disorder that affects the skin, nails, hair, and mucous membranes. Different subtypes of the disease have been so far described. Bullous lichen planus (BLP) is a rare variant. We report a case of a 14-year-old patient.

**Case report:**

A 14-year-old patient with no medical history presented a generalized rash of sudden onset. Dermatological examination revealed a diffuse rash consisting of pruritic violaceous papules with fine scaling, located on the trunk, legs, and arms discussing diagnoses of LP and pityriasis lichenoides chronica. The patient refused a biopsy. He was treated with topical steroids, cyclines, and antihistamines, which resulted in clinical partial improvement. One month later, the patient experienced a second flare-up with extension on the hands and feet. On examination, there were multiple erythematous papules with violaceous bullae. Nails, hair, and mucosal examination were normal. The patient's overall condition was stable with no history of any drug intake or aggravation of lesions on sun exposure. Histopathology revealed an ortho-keratotic lamellar epidermis with hypergranulosis. The dermis showed a lymphohistiocytic inflammatory infiltrate disposed of in a band-like pattern beneath a discontinuous epidermis, which focally eroded the basal lamina without sub-epidermal separation and keratinocyte necrosis leading to a diagnosis of BLP. The patient was treated with three intramuscular corticosteroids injection at one-month intervals. The patient's condition continued to improve clinically.

**Discussion:**

Lichen planus is a common inflammatory disorder of unknown etiology. There are many variants of LP, including the bullous form known as bullous lichen planus. Its exact prevalence remains unknown. While it is commonly observed as a sporadic condition, there have also been documented instances of familial occurrences in the literature that appear earlier and last longer. Typically, the lesions occur as individual vesicles and blisters develop on pre-existing lesions of LP on the lower extremities, wrists and forearms, dorsal hands, shins, and genital area. Nail involvement manifests by hemorrhagic crusting, leading to complete loss of the nail plate and, ultimately nail atrophy.

Histology findings lead to suggesting that the extensive infiltration of lymphocytes leads to the destruction of the basal cell layer, resulting in the formation of a large Max Joseph space and in consequence exposing hidden epidermal antigens. This exposure may trigger the production of autoantibodies, resulting in the formation of bullae.

Currently, there is no established treatment of choice for BLP and no clearly effective treatment has been identified. However, due to its hyper-reactive nature as a form of LP, topical corticosteroids have been empirically used. Oral betamethasone mini pulse therapy has shown effectiveness in managing moderate to severe oral BLP. Systemic corticosteroids, considered as second-line treatments for lichen planus, are reserved for severe cases that do not respond adequately to local treatments. Other treatments have been used such as Dapsone in pediatric

forms, mycophenolate mofetil, and topical retinoid 0,025% in combination with triamcinolone 0,1% and acitretin. However, additional studies involving a large number of patients would significantly contribute to the standardization of treatment for this particular disease.

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**Abstract N°: 4615**

**From prickles to papules: uncommon presentation in adulthood**

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**Introduction & Objectives:**

Lichen spinulosus is a rare dermatological condition characterized by the presence of spiny papules and follicular hyperkeratosis on the skin. While it is more often observed in children and adolescents, its occurrence in adults is extremely rare. This abstract depicts a case of lichen spinulosus in a 63-year-old male patient, aiming to explore the aspects of this presentation, describe the clinical manifestations and discuss potential etiological factors.

**Materials & Methods:**

This retrospective case study involved an analysis of medical records, clinical photographs, and patient interview. A comprehensive review of existing literature was conducted to compare and contrast the clinical features and prevalence of lichen spinulosus. The patient's medical history, presenting symptoms, duration of the condition and associated factors were carefully documented.

**Results:**

The patient, a 63-year-old male with a history of stage IVA lung adenocarcinoma under palliative chemotherapy, was referred from the oncology department to dermatology due to the presence of spiny lesions and pruritus. The patient presented with multiple hyperkeratotic papules symmetrically distributed on the trunk, extremities, and face. Clinical examination and thorough investigation confirmed the diagnosis of lichen spinulosus. The therapeutic approach involved the use of keratolytic agents, which led to a significant clinical improvement. There was a notable reduction in the number and severity of the papules, demonstrating the effectiveness of the keratolytic treatment in managing lichen spinulosus-associated symptoms. Notably, lichen spinulosus is primarily reported in children and adolescents, and its occurrence in adulthood is rare. The prevalence, clinical features, and potential etiological factors in adult-onset lichen spinulosus will be discussed.

**Conclusion:**

This clinical case sheds light on the uncommon occurrence of lichen spinulosus in adulthood, aiming to raise important considerations regarding its diagnosis and management. Clinicians should be aware of this presentation and include lichen spinulosus in the differential diagnosis of adult patients presenting with hyperkeratotic papules. Further research is warranted to better understand the underlying mechanisms and etiological factors contributing to adult-onset lichen spinulosus.



**Abstract N°: 4790****Prurigo nodularis patients with and without comorbid atopic dermatitis exhibit different clinical characteristics and treatment response**

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**Introduction & Objectives:** Prurigo nodularis (PN) is a chronic inflammatory skin condition characterized by pruritic, hyperkeratotic nodules that often develop in the setting of other pruritic conditions, including atopic dermatitis (AD). PN and AD are both characterized by pathogenic mechanisms such as skin barrier disruption and type 2 inflammation. Despite the similarities it shares with AD, PN also demonstrates broad clinical and molecular heterogeneity, with distinct disease endotypes comprised of patients harboring atopic, neurologic, systemic, or psychiatric comorbidities and diverse neuroimmune profiles. We hypothesize that PN patients with AD most likely belong to a clinically distinct atopic disease endotype, whereas PN patients without AD may belong to a multitude of diverse endotypes, which could result in relevant clinical differences between these groups.

**Materials & Methods:** To test this hypothesis, we performed a retrospective cohort analysis to study differences in clinical characteristics and treatment response between PN patients with and without a diagnosis of AD. We examined 735 patients in the Johns Hopkins healthcare system, 544 of which were diagnosed with PN alone and 191 of which were diagnosed with both PN and AD. Chi-squared tests, Fisher's exact tests, Wilcoxon rank sum tests, and T-tests were used to compare clinical characteristics for categorical and continuous variables, as appropriate. Multivariate linear and logistic regressions were performed to determine the association of therapy-related improvements in pruritus with the presence or absence of concurrent AD.

**Results:** Results revealed that after adjusting for age, sex and race, patients with PN and AD were more likely to have food allergy, asthma, and personal or family history of atopy ( $p < 0.001$ ) when compared to patients with PN alone. Patients with PN alone were more likely to have human immunodeficiency virus ( $p = 0.015$ ) and chronic kidney disease ( $p < 0.001$ ). They were also less likely to report improvement in pruritus after therapy (adjusted odds ratio (AOR) 0.44,  $p < 0.001$ ), especially when systemic therapy was required (AOR 0.34,  $p = 0.004$ ).

**Conclusion:** Our results demonstrate clinical and therapy-related distinctions between PN with and without concurrent AD, supporting the notion that there is a clinically distinct atopic endotype of PN. They also highlight an unmet need for effective treatment in PN patients without a comorbid AD diagnosis. Decreased therapy response in this subgroup of patients is likely due to the contribution of diverse disease mechanisms, which are not yet adequately targeted by medications used in treatment of PN. Further investigation into these mechanisms, which may be neuropathic, systemic, or psychiatric in origin, is warranted to facilitate development of effective treatment strategies for all PN patients.

**Abstract N°: 4821****Clinical and histopathological analysis of Rosai-Dorfman Disease with cutaneous involvement**

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<sup>1</sup>ASAN Medical Center, Korea, Rep. of South

**Introduction & Objectives:**

Rosai-Dorfman Disease (RDD) is a rare histiocytic proliferative disorder, with cutaneous involvement often documented. Racial difference had been previously reported in literature. This study aimed to identify the clinical and histopathological features of RDD with cutaneous involvement among Asian patients.

**Materials & Methods:**

A retrospective review was performed of patients that had been pathologically diagnosed with RDD from a skin biopsy at a tertiary hospital in Korea. A pathological examination of hematoxylin-eosin and immunohistochemical stains was conducted, while clinical and laboratory data, imaging studies, and treatment modalities were also reviewed.

**Results:**

Altogether, after analyzing the cases of ten patients (seven men and three women) who were histologically diagnosed with RDD, eight cases were purely cutaneous, one case had cervical involvement, and one had thymus involvement. Clinical presentations on the skin mostly comprised asymptomatic grouped erythematous papules. Five patients had skin lesions on their head and neck. Histopathological reviews of 18 skin tissue slides revealed dermal infiltration that occasionally extended to the subcutis. The infiltrated cells comprised mainly lymphocytes, histiocytes, and plasma cells. The most common pattern of infiltration was diffuse, followed by a nodular pattern. Immunohistochemical staining of S-100 and CD68 highlighted emperipolesis.

**Conclusion:**

Asian RDD with cutaneous involvement has male predominance and commonly presents as asymptomatic grouped erythematous papules and shows histological signs of diffuse dermal infiltration of histiocytes with lymphoplasmic cells and emperipolesis.

**Abstract N°: 4825**

**Using Artificial Intelligence to Phenotype Generalized Pustular Psoriasis (GPP): Getting the Right Patients the Right Treatment at the Right Time**

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**Introduction & Objectives:**

Generalized pustular psoriasis (GPP) remains underdiagnosed and underrepresented through claims-based diagnosis coding alone. Clinical registries offer a more robust and accurate diagnosis of rare diseases, such as GPP, however are limited by physician registry participation. Artificial intelligence-based (AI-based) approaches represent a scalable solution to spotlight the likelihood of a disease at the individual level. Furthermore, AI-based approaches that link multiple registries and data sources can prospectively isolate patterns in data associated with diseases like GPP by revealing useful signaling patterns in patients' histories derived from heterogeneous clinical presentations. We will describe the use of a national clinical data registry (DataDerm), linked with an artificial intelligence solution, to reduce the underdiagnosis and misdiagnosis of GPP. This approach will be compared to the more limited, historical methodology of claims data patient identification.

**Materials & Methods:**

Claims Based Patient Identification:

Many GPP patients get hospitalized and misdiagnosed with sepsis or cellulitis based on a high white blood cell count, pustules, and profound erythema. To identify undiagnosed or misdiagnosed GPP patients, a heterogeneous open claims data source was used to identify psoriasis patients who were also hospitalized with a diagnosis code of sepsis or cellulitis.

National Registry Patient Identification:

DataDerm is a national clinical data registry that incorporates real world dermatology data on  $\geq 50$  million patient encounters from  $\geq 14$  million unique patients. It offers longitudinal insights on the impact of practice patterns on patient care across numerous rare diseases including GPP. GPP patients were identified by their diagnosis codes and their therapeutic journey documented within the medical record extracted into the registry.

AI-Based Patient Identification:

GPP cohorts will be defined using real-world datasets by: a) isolation through standard epidemiologic techniques, and b) patient-level data comparison with novel digital phenotypic profiles generated by artificial intelligence.

**Results:**

Claims data identified 16,100 patients with a diagnosis of psoriasis and a diagnosis sepsis or cellulitis suggesting the possibility of GPP patients (Table 1). These patients represent patients with a possible diagnosis with GPP, but the analysis lacks specificity. DataDerm identified 6,246 GPP and 128,200 psoriasis patients (Table 2). These patients represent a GPP cohort with the correct diagnosis. However, the patients are limited to those who are cared for by physicians participating in the registry. AI-based profiles grounded in common patterns in known GPP patients' health histories highlight patients clinically similar to those already diagnosed, but without recorded



diagnosis themselves. These analyses will identify GPP patients– including those likely falling in the ‘diagnosis gap’ - and create a profile for dermatologists to better understand GPP patients and their treatment pathways (Figures 1&2)

**Conclusion:**

GPP patients in in DataDerm represent an accurately diagnosed cohort and can be used as a training set for AI-based profiling. Robust, tested AI modeling allows for the identification of patients known to exist, but are undiscoverable through claims. In the end, and most importantly, AI modeling facilitates getting the right drug, to the right patient, at the right time.

Figures & Tables:

**Table 1. Patients with psoriasis and an inpatient diagnosis of sepsis or cellulitis at any time point from open claims data**

Inpatient diagnosis	Count (claims data sources)
Sepsis	2,856
Cellulitis	14,364
Total Psoriasis + Cellulitis / Sepsis	17,220

**Table 2: DataDerm: Generalized Pustular Psoriasis vs General Psoriasis Cohort Characteristics**

	GPP			Psoriasis	
	6,496			128,200	
Sex					
Female	3,969	61.1%		70,638	55.1%
Race					
White	5,013	77.2%		99,000	77.2%
Black or African American	397	6.1%		5,045	3.9%
Asian	135	2.1%		3,036	2.4%
Other race	134	2.1%		2,400	1.9%
Unknown	821	12.6%		18,700	14.6%
Ethnicity					
Hispanic or Latino	218	3.4%		5,276	4.1%
Not Hispanic or Latino	5,138	79.1%		97,936	76.4%
Unknown	1,140	17.5%		24,960	19.5%



Fig 1: Disease Identification by Ascertainment Method

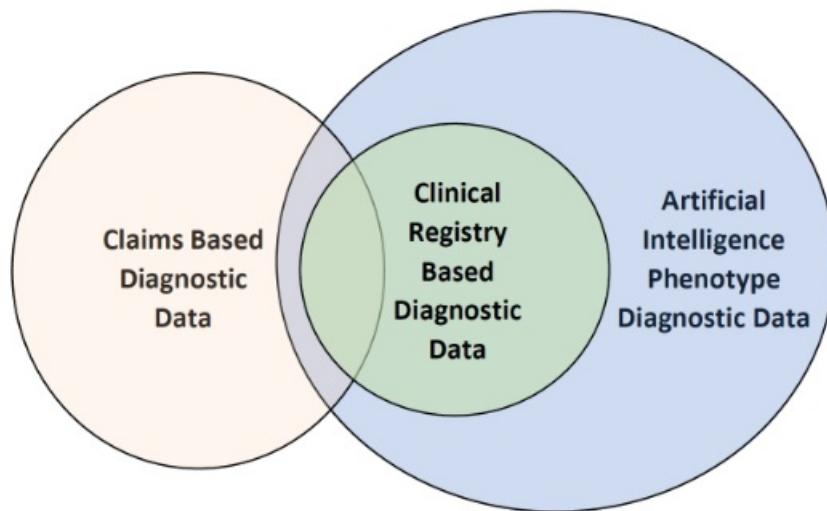
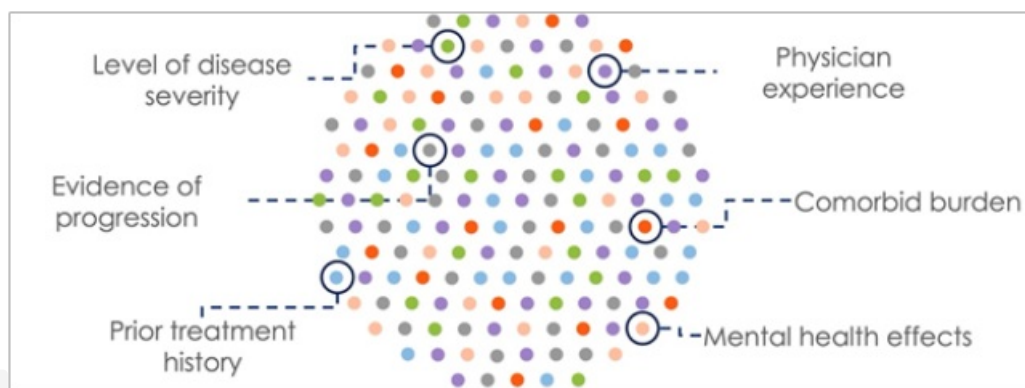


Figure 2: Fig 1: GPP Fingerprint through PhenOM AI



**Abstract N°: 4837**
**Effects of Clinicopathological Features on Dermatomyositis Prognosis: A Multicenter Study on Experience of Three Tertiary-care Hospital**

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**Effects of Clinicopathological Features on Dermatomyositis Prognosis: A Multicenter Study on Experience of Three Tertiary-care Hospital**
**Introduction & Objectives:**

Dermatomyositis (DM) is a rare autoimmune disease causing inflammation of the skin and muscles, associated with higher mortality rates. Around 15-30% of DM patients may develop an associated malignancy during their lifetime, often after DM diagnosis. DM often precedes cancer diagnosis, emphasizing the importance of vigilant monitoring and early detection in these patients. Concomitant malignancies and specific autoantibodies like anti-TIF1-γ and anti-NXP2 are linked to increased mortality risk in DM patients. Monitoring and early detection of cancer are vital in DM, considering the connection with malignancies and higher mortality rates. Understanding the clinicopathological signs related to underlying malignancy and mortality can provide insights into diagnostic procedures and potentially guide more effective treatment strategies. We aimed to discover potential clinical and histopathological clues that may be related to a worse prognosis.

**Materials & Methods:**

We conducted a multicenter retrospective study using data from three tertiary care hospitals in Turkey. The study focused on adult patients clinically and histopathologically diagnosed with dermatomyositis (DM). We performed a retrospective review of the clinical and laboratory features of the cases, utilizing the hospital automation system and photo archives. Additionally, we calculated the systemic inflammatory index (SII) for each case. The histopathological findings were evaluated by a single pathologist who re-evaluated the skin biopsy specimens and assigned semi-quantitative scores.

**Results:**

This study included 56 patients (43 female, 13 male, mean age: 53.04±16 yrs). Positive ANA results were observed in 66% of cases, with the most common pattern being homogeneous spots (25%). Malignancy was present in 36% of patients, predominantly breast ductal carcinoma (16%). Malignancy was detected in 16% of cases after the diagnosis of DM, with an average interval of 42±58 months. Myopathy was present in 61% of cases. During follow-up, 41% of patients died, with a mean survival of 23±23 months. Higher SII and ESR levels were associated with a shorter time between DM and malignancy diagnosis (r: -0.9, -0.7; p: <0.05). Higher blood eosinophil percentage (EP) at diagnosis correlated with more prolonged overall survival (r: 0.4, p: 0.02). At diagnosis,

myopathy, heliotrope rash, elevated ESR, and CK levels were notably associated with mortality ( $p < 0.05$ ). Myopathy was higher in patients with pruritus ( $p: 0.01$ ) and  $EP > 1.75\%$  (sensitivity 68%, specificity: 73%). Survival was higher in patients with  $ESR < 38.5$  mm/h,  $CK < 145$  U/L, and  $CK-MB < 24$  U/L (sensitivity: 64, 90, 90%; specificity: 80, 73, 67%).

#### **Conclusion:**

Our findings indicate that there are readily available and cost-effective alternative markers for monitoring DM. Patients with elevated levels of SII and ESR may be at a higher risk of developing malignancies within a shorter timeframe, necessitating closer surveillance for cancer. In cases where measuring CK or CK-MB is not feasible, patients presenting with pruritus and EP above 1.75% could be suspected of having myopathy.

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**Abstract N°: 4868**

**Marshall's syndrome as a paraneoplastic manifestation of smoldering multiple myeloma with successful response to chemotherapy**

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**Introduction & Objectives:**

Cutis laxa is an inherited or acquired disorder characterized by loose pendulous lesions due to the abnormal production or destruction of dermal elastic fibers. Marshall's syndrome is a type of acquired cutis laxa (ACL) with exclusive cutaneous involvement following inflammatory dermatoses such as Sweet's syndrome. It has rarely been reported in association with multiple myeloma (MM). We report the diagnostic and therapeutic challenges in a longterm case of ACL secondary to Sweet's syndrome as a paraneoplastic manifestation of smoldering MM.

**Materials & Methods:**

To report a case of Marshall's syndrome secondary to a corticoid-dependent paraneoplastic neutrophilic dermatosis related to MM, with successful response to chemotherapy.

**Results:**

In 2008, a 49-year-old man presented with a one-year history of recurrent erythematous infiltrated plaques on the chest, breasts, axillae and legs, which evolved with skin's laxity. Histopathological examination demonstrated a diffuse neutrophilic infiltrate in the mid and deep dermis. Systemic workup revealed a suspected monoclonal gamma peak with an IgG lambda paraprotein in blood immunofixation, thus favoring a monoclonal gammopathy of undetermined significance (MGUS). Lesions improved with dexamethasone 40 mg/day for four days and thalidomide 100 mg/day, but the patient was lost to follow-up.

After 12 years, he returned to the dermatology service due to the persistence of the dermatosis. New biopsies were performed for anatomopathological analysis revealing an intense diffuse neutrophilic dermal infiltrate with edema in an erythematous infiltrated cervical lesion, and decreased elastic fibers in the papillary and reticular dermis in the axillary loose skin. Serum protein electrophoresis demonstrated a monoclonal gamma peak, and blood immunofixation revealed IgG lambda and IgG kappa paraproteins. A bone marrow biopsy showed 10% of plasma cells with a predominance of lambda light chain production, confirming the diagnosis of smoldering MM with Marshall's syndrome as a paraneoplastic manifestation. Prednisone 1 mg/kg/day was introduced with good clinical response. However, corticosteroid tapering was limited by the occurrence of new skin lesions despite adjuvant therapy with colchicine 1.5 mg/day, dapsone 100 mg/day and thalidomide 100 mg/day.

Along with the hematologists, VTD regimen was used from March 2022 to September 2022. Prednisone, colchicine, thalidomide and dapsone were sequentially withdrawn thereafter with sustained remission of skin lesions ever since.

**Conclusion:** \*\* Our patient was initially diagnosed with MGUS, but recognition of ACL as a manifestation of lymphoplasmacytic disease led to further comprehensive evaluation enabling the diagnosis of smoldering MM. Although end-organ dysfunction is the classic recommendation for initiating treatment for plasma cell dyscrasias,

we demonstrate that severe and refractory paraneoplastic dermatological manifestations may also improve with chemotherapy.

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**Abstract N°: 4873****Lichen sclerosus in prepubertal males: breaking the paradigm**

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**Lichen sclerosus in prepubertal males: breaking the paradigm**

**Alessia Paganelli<sup>1\*</sup>, Paolo Viscardo Fabbri<sup>2</sup>, Filippo Ghidini<sup>3</sup>, Laura Bigi<sup>1</sup>, Claudia Lasagni<sup>1</sup>, Pier Luca Ceccarelli<sup>3</sup>**

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**Introduction & Objectives:** Lichen sclerosus (LS) is classically described as more frequent in women, with two peaks of incidence, in prepubertal age and late adulthood. However, recent literature suggests the real prevalence of the disease is probably underestimated, especially in the pediatric and male population. The aim of the present study was to describe the casuistry of our LS referral center.

**Materials & Methods:** We retrospectively collected data on children affected by LS followed-up at our referral center in the last 10 years. Only patients with histological confirmation and at least one follow-up visit (either dermatological or urological) were taken into account.

**Results:** In total, 194 patients aged 18 or under were histologically diagnosed LS at our center between January 2013 and January 2023. Follow-up ranged from 2 weeks up to 8.2 years (mean 2.9 years). Mean age at diagnosis was 10. Of them, only one was a girl, while 193 were boys. The vast majority of the subjects (n=185) were circumcised for the presence of LS-related phimosis. Nearly half of the patients (52.1%), however, experienced complete remission of genital LS after circumcision, in the absence of systematic periodic application of any topicals. Often, the patients were given instructions on how to recur to topical CS as needed. External urethral meatus (EUM) stenosis was a relatively rare but possible LS-related complications.

**Conclusions:** These findings suggest a prevalence of LS in male children far above the threshold for being classified as a rare disease, confirming previous literature possibly being biased by misdiagnosis of the disease. Moreover, circumcision seems to possibly be totally curative in cases with preputial involvement only, therefore paving the way for surgery as the first-choice treatment for an inflammatory disorder.

**Abstract N°: 4885****Co-existence of vitiligo and lichen sclerosis: a retrospective monocentric study on a rare association**

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**Introduction & Objectives:** Vitiligo is a common T-cell mediated skin disease characterized by loss of pigment of the skin with a chronic and unpredictable course. Lichen sclerosis (LS) is an underdiagnosed chronic mucocutaneous T-cell mediated disease, which typically involves the anogenital area, with an estimated incidence between 0.1 and 0.3%. Since both diseases are characterized by depigmented patches, the differential diagnosis between vitiligo and LS is sometimes challenging. Furthermore, the co-existence between vitiligo and LS has already been pointed out, albeit in isolated case reports.

In this study, we aimed to estimate the frequency of LS in a large cohort of patients with vitiligo and to describe the clinical features of patients with co-existing vitiligo and LS.

**Materials & Methods:** we search our institutional electronic database of patients diagnosed with vitiligo at our academic institution between January 2000 and December 2020. We selected patients who have also received a diagnosis of LS based on a confirmatory histopatological examination. Demographic data, comorbidities and clinical characteristics of both vitiligo and LS were extrapolated and reviewed.

**Results:** we identified 7 out of 987 patients (0.7%) with concomitant vitiligo and LS. They included 5 females and 2 males (mean age 56.71 years, range 39-69). Mean time between onset of depigmented lesions and diagnosis of vitiligo was 11 years (range 0.2-13). In all the patients, the diagnosis of LS preceded that of vitiligo. All the patients presented with genital LS, without extragenital manifestations. Vitiligo was of the non-segmental type in all the patients; 2 patients presented with acro-facial vitiligo, while the remaining 5 presented with generalized vitiligo. Regarding comorbidities, 4 patients were affected by autoimmune hypothyroidism; while, 4 patients were also affected by psoriasis, of whom 1 showed a guttate psoriasis. In one patient, vitiligo and psoriasis lesions co-localized.

The clinical course of LS was mild/moderate in all the patients and well controlled by periodic course of potent corticosteroids. None of the patients developed a squamous cell carcinoma over the study period.

**Conclusion:** the coexistence between vitiligo and LS is rare. The fact that all the patients had non-segmental vitiligo and the concomitant presence of other T-cell mediated diseases, such as psoriasis, suggest an autoimmune basis for this rare association.\*\*



**Abstract N°: 4946**
**AhR agonism by tapinarof regulates TH2 and TH17 cell function in human skin**

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**Introduction & Objectives:**

The aryl hydrocarbon receptor (AhR) is a ligand-based transcription factor that plays a crucial role in skin homeostasis and skin barrier function. Tapinarof, a topical AhR agonist, has shown clinical efficacy in both psoriasis (PSO) and atopic dermatitis (AD) where it induced long lasting remissions. However, the anti-inflammatory mechanism of action of tapinarof is not fully understood.

Our aim was to study the mechanism of action of tapinarof on skin T cells in of patients with AD, PSO and allergic contact dermatitis (ACD).

**Materials & Methods:**

We established short-term human skin explant cultures from diseased AD, PS or ACD skin biopsies that we treated with tapinarof for 24h and then performed downstream analysis of skin T cells.

**Results:**

We observed highest expression of disease-relevant cytokines in the tissue-resident memory T (TRM) cell populations (IL-13+CD4+ TRM in AD and IL-17a+CD8+ TRM in PSO) previously shown to play a key role in disease progression, validating our ex vivo model. IL-13 and IL-17a were significantly reduced after tapinarof treatment in the respective diseases and populations. AhR agonism in ACD led to a significant reduction of IL-13 levels in TRM and CD4+ T-cells, while leaving IFN-g unchanged.

To uncover the mechanism of action of tapinarof we performed transcriptomic analysis of T-cells treated with tapinarof. As expected, tapinarof treatment resulted in a concerted upregulation of genes associated with AhR signaling pathway and cytochrome P450 response. Moreover, we confirm that *IL13* and *IL17A* expression is reduced in response to tapinarof treatment.

In addition, RNAseq data revealed a downregulation of processes involved in T cell activation and metabolic enzymes in response to tapinarof treatment. sc-RNAseq experiments of T cells isolated from AD and PSO biopsies cultured in presence of tapinarof showed a similar metabolic impairment. Preliminary mechanistic studies of metabolism show a reduction of extracellular acidification rate (ECR) and oxygen consumption rate (OCR) in T cells after treatment with tapinarof, both in the resting and activated state.

**Conclusion:**

In summary, we established an ex vivo model to study the effect of tapinarof on skin T cells and found a significant reduction in disease-relevant cytokines in AD, PSO and ACD after tapinarof treatment. Furthermore, we show that tapinarof treatment leads to metabolic impairment affecting both glycolysis and oxidative phosphorylation in T cells, uncovering a previously unknown mechanism of action of tapinarof.





**Abstract N°: 5017**
**Evaluation of interleukin-36 $\alpha$ , interleukin-36 $\beta$ , and interleukin-36 $\gamma$  serum levels in patients with pyoderma gangrenosum.**

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**Introduction & Objectives:**

Pyoderma gangrenosum (PG) is a rare neutrophilic inflammatory dermatosis of unknown etiology. PG frequently coexists with different conditions, in particular inflammatory bowel diseases, rheumatological diseases, and hematological disorders, with which it shares the features of impaired immune system functions. It is also a part of classical autoinflammatory syndromes e.g. pyogenic arthritis, PG, acne, and hidradenitis suppurativa (PASH); PG, and acne (PAPA). The IL-36 group (IL-36 $\alpha$ , IL-36 $\beta$ , IL-36 $\gamma$ ) comprises agonists of the IL36R receptor, which stimulate a location- and inflammation-specific proinflammatory response. While the role of IL-36 group members has been described in autoinflammatory diseases associated with PG, their involvement in PG itself remains unexplored. This study aimed to evaluate the serum levels of IL-36 $\alpha$ , IL-36 $\beta$ , and IL-36 $\gamma$  in patients with PG in relation to clinical data.

**Materials & Methods:**

The study involved 48 adult patients with a confirmed diagnosis of PG and 40 healthy volunteers. Any active comorbidities, malignancies, recent systematic treatment as well as present infection were among the exclusion criteria. Upon admission, the number, dimensions, and surface area of skin lesions were determined. Moreover, peripheral venous serum samples were collected. The assessed clinical parameters included the concentrations of IL-36 $\alpha$ , IL-36 $\beta$ , IL-36 $\gamma$ , C-reactive protein (CRP), white blood cell count (WBC), and neutrophil count.

**Results:**

The study included 31 (64.6%) female and 17 (35.4%) male patients with PG. The control group consisted of 21 (52.5%) females and 19 (47.5%) males. The mean age of patients was 51.3 $\pm$ 16.9 years, while that of the control group was 48.1 $\pm$ 13.0 years. In the patient group, the median serum concentrations of IL-36 $\alpha$ , IL-36 $\beta$ , and IL-36 $\gamma$  were 0.97 ng/ml (IQR=0.97), 123.63 pg/ml (IQR=344.42), and 26.25 pg/ml (IQR=46.88), respectively. In the control group, the median serum concentrations of IL-36 $\alpha$ , IL-36 $\beta$ , and IL-36 $\gamma$  were 1.66 ng/ml (IQR=2.60), 109.54 pg/ml (IQR=136.93), and 52.04 pg/ml (IQR=70.88), respectively. Significantly lower levels of IL-36 $\alpha$  ( $p=0.0003$ ) and IL-36 $\gamma$  ( $p=0.031$ ) were observed in the patient group compared to the control group. The concentration of IL-36 $\beta$  correlated with IL-36 $\alpha$  ( $r=0.3$ ,  $p=0.0036$ ) and IL-36 $\gamma$  ( $r=0.2$ ,  $p=0.03$ ). A negative correlation was found between IL-36 $\gamma$  and WBC ( $r=-0.34$ ,  $p=0.021$ ), as well as between IL-36 $\alpha$  and the total ulcer surface area ( $r=-0.4$ ,  $p=0.002$ ).

**Conclusion:**

IL-36 $\alpha$  and IL-36 $\gamma$  serum levels were significantly decreased in patients with PG compared to the control group. A larger ulcer surface area was associated with lower levels of IL-36 $\alpha$ , while higher WBC counts correlated with lower levels of IL-36 $\gamma$ . Further research is necessary to investigate the role of the IL-36 group in the pathogenesis of pyoderma gangrenosum (PG), as they may have implications for the planning and development of new

therapeutic options in this challenging-to-treat dermatosis.

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**Abstract N°: 5064****Triple status quo: psoriasis, syphilis, and tinea masquerading one another**

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**Introduction:** Psoriasis is a chronic autoimmune inflammatory skin disease with a prevalence rate of 2-3% worldwide. There is a wide range of differential diagnoses, including syphilis, tinea capitis and corporis, seborrheic and atopic dermatitis. Generalized tinea corporis and psoriasiform syphilis, have been reported to have presentations that imitate psoriasis. We represent a case report of a patient with three active skin diseases: psoriasis, syphilis, and tinea corporis, that appeared simultaneously.

**Case report:** A 24-year-old male patient was admitted to the emergency department due to worsening of pruritic disseminated skin lesions that appeared three weeks ago. The patient denied substance use, morning stiffness, or joint pain and had no family history of psoriasis. However, the medical history noted red and scaly lesions on his elbows for several years. The dermatological examination revealed erythematous squamous plaques on the elbows and disseminated eczematoid plaques on the skin of the trunk, legs, and arms. The wax-spot phenomenon was positive. Erythematous plaques were also visible in the groin, intergluteal area, and the glans penis. Based on the clinical findings, a diagnosis of psoriasis was made. The patient was treated with topical corticosteroids and calcipotriol. Screening for HIV, HBV, and HCV were negative while laboratory investigations revealed positive TPHA and FTA-ABS tests for syphilis. Therefore, 100 milligrams of doxycycline was prescribed. Within one day, the patient developed a Jarisch-Herxheimer reaction which was treated with methylprednisolone systemically, tapering slowly, and betamethasone and gentamicin locally. After a few weeks, the patient's condition did not improve, moreover, painful purulent nodules appeared on his skin. Considering these clinical findings, a mycological diagnostic investigation was performed. Trichophyton spp. was proven, and the patient was treated systemically with terbinafine and methylprednisolone and locally with imidazole and triazole derivatives. After two weeks, the skin lesions were in complete remission, and pruritus was absent.

**Conclusion:** Physicians should be more alert to diseases that frequently masquerade as different dermatological conditions. Adequate clinical and microbiological diagnostic tests should be implemented, especially if the skin condition is worsening or not improving to the prescribed therapy. Those diagnostic tests should be a valuable tool for distinguishing mimicry from the coexistence of different diseases, which is essential for proper treatment management.



**Abstract N°: 5163**
**Comparative randomized split face clinical study to assess the efficacy and tolerance of a serum containing Silybum Marianum Angiopausin Extract and Avène Thermal Spring water in adult women with erythematotelangiectatic rosacea**

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**Introduction & Objectives:**

Dermo-cosmetics are often recommended by dermatologists as complementary care in the management of rosacea. A new serum containing *Silybum Marianum Angiopausin Extract (SMAE)* and Avène Thermal Spring water, was developed to improve skin comfort and reactivity, soothing the functional signs and to help reduce redness in subjects with erythematotelangiectatic rosacea. The aim of the study was to compare the benefit of tested product to the volunteer usual basic face emollient based on clinical and instrumental evaluations.

**Materials & Methods:** This monocentric intraindividual split face comparative-labelled study enrolled 32 subjects (18 to 65 years old) with mild to moderate erythematotelangiectatic rosacea. The product was applied twice/day for 1 month on randomised half-face and compared to usual cosmetic emollient (*without anti redness active ingredients*) on contralateral half-face (CRT). Two visits were planned (D1 and D29).

Clinical and instrumental evaluations were conducted for each half face at D1 and D29: - tolerance of the product, -physical and functional rosacea signs (flushes severity, discomfort signs, erythema), -dynamic Patient's Global Assessment of rosacea. -face and ¾ profiles standardized photographs for investigator blinded erythema scoring.

Innovative Instrumental evaluations: - Dynamic Optical Coherence Tomography (D-OCT) measurement's (*vessels diameter and density, mean blood flow signals*); -multispectral imaging (*color assessment in CIELab*); - epidermal lipid analysis measurements. Benefit and cosmetic acceptability questionnaire were also performed by subjects.

**Results:** 31 female volunteers were included and analysed (mean age 48.06; 19 to 65 yo).

The tolerance of the product was assessed as very good by the investigator.

The flushes frequency and subjective signs improved significantly from D1 to D29 in treated versus controlled half-face. A significant clinical improvement of erythema and functional signs were also observed from D1 to D29 in coherence with instrumental analysis. A significant improvement of functional signs (stinging, warmth, tightness, and global discomfort) was observed after 1 month product use versus usual basic emollient. The cosmetic acceptability of the product was very good.

**Conclusion:** This comparative randomised intraindividual clinical and instrumental study demonstrated the

efficacy and very good tolerance of this new serum containing *Sylibum Marianum Angiopausin Extract (SMAE)* and Avène Thermal Spring water in adults with erythematotelangiectatic rosacea. The significant improvement of clinical functional signs, persistent erythema was confirmed using innovative instrumental methods and contribute to highlight the potential benefit of this product as complementary dermocosmetic care in rosacea.

**32<sup>ND</sup> EADV Congress 2023**  
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**Abstract N°: 5272****Tolerance and efficacy of a serum containing *Sylibum Marianum* Angiopausin Extract and Thermal Spring water in adults with erythematotelangiectatic rosacea.**

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**Introduction & Objectives:** Dermo-cosmetics are often recommended by dermatologists as complementary care in the management of rosacea. A new serum containing *Sylibum Marianum* Angiopausin Extract (SMAE) and Thermal Spring water, was developed to improve skin comfort and reactivity, soothing the functional signs and to help reduce redness in subjects with erythematotelangiectatic rosacea. The aim of the study was to assess the tolerance and efficacy of the product.

**Materials & Methods:** This monocentric open-label study enrolled 44 subjects (33 – 65 years old) with mild to moderate erythematotelangiectatic rosacea. The product was applied twice a day during 4 weeks. Three visits were planned (D1, D15, D29,). Primary criterion of the study was to assess the tolerance of the product, based on physical and functional signs assessment. Secondary criteria included: erythema severity using\* Clinical Erythema Assessment (CEA – Scale: -1 (worsening) to 4 (very good improvement)) , flushes severity\* using Global Flushing Severity Score (GFSS – Scale: 0 (none) to 10 (extreme)) , flushes frequency, Patient's Global Assessment (PGA – Scale: -1 (worsening) to 4 (very good improvement)) of rosacea, functional signs evaluations (burning, stinging, warmth, tightness, and discomfort) using Numerating Rating Scale (NRS – Scale: 0 (none) to 10 (huge)). Instrumental evaluations including chromameter measurement's from D1 to D15 and D29, standardized photographs. Cosmetic acceptability questionnaire was also performed by subjects.

**Results:** The tolerance of the product was assessed as good by the investigator. The flushes frequency and the flushes severity significantly decreased from D1 at D15 and D29 ( $p < 0.001$ ). On D1, 90% of subjects reported having flushes usually; after 15 and 29 days of product use subjects reported an improvement: only 72% and 66% respectively still reported having flushes. A significant improvement of erythema was also observed from D1 to D29 ( $p < 0.05$ ). Moreover, the chromameter measurement showed a significant decrease of the *a* parameter since D15 and also at D29 ( $p < 0.001$ ). A significant improvement of functional signs (stinging, warmth, tightness and global discomfort) was observed at D1 immediately after the first application (10 min) and at D15 and D29 ( $p < 0.001$ ). Finally, 71% of subjects perceived a global improvement of their rosacea, and the cosmetic acceptability of the product was good.

**Conclusion:** This open clinical and instrumental study demonstrated the benefit and tolerance of this new serum containing *Sylibum Marianum* Angiopausin Extract (SMAE) and Thermal Spring water in adults with erythematotelangiectatic rosacea. The significant improvement of flushing, the soothing of functional signs and erythema, highlight the potential benefit of this product as complementary care in rosacea.

**Abstract N°: 5429****Linear Facial Lichen Planus: A Rare Presentation Revealed by Dermoscopy**

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**Introduction & Objectives:**

Lichen planus (LP) is a chronic inflammatory disorder that commonly affects the skin, mucous membranes, nails and scalp. Linear LP is a variant of LP distributed along Blaschko lines in a unilateral manner. Linear LP presenting on the face is an exceptionally rare entity. Dermoscopy, a non-invasive diagnostic tool, can provide invaluable insights into the distinctive features of linear facial LP.

**Materials & Methods:**

We report the case of a 54-year-old man presenting with a linear pruriginous eruption limited to the face after COVID-19 infection. The patient exhibited violaceous, linear papules arranged along the left nasal sidewall following the lines of Blaschko. Concomitantly, the patient presented oral LP and lichen planopilaris on the scalp. Dermoscopic examination of the nasal lesions revealed characteristic findings, including rosettes, perifollicular erythema and grey-blue dots and globules. The clinical and dermoscopic correlation led to a diagnosis of linear facial lichen planus (LFLP).

**Results:**

LFLP is an extremely rare variant that poses diagnostic challenges due to its unusual presentation. The incorporation of dermoscopy in the evaluation of facial dermatoses aids in recognizing the specific dermoscopic features and differentiating them from other linear facial eruptions. This case underscores the importance of dermoscopy as a valuable tool in diagnosing and managing rare presentations of lichen planus.

**Conclusion:**

We present a rare case of LFLP, a rarely reported variant of LP. The utilization of dermoscopy enabled precise diagnosis and appropriate management. This case contributes to the limited literature on LFLP and highlights the value of dermoscopy not only in pigmented lesions but also in inflammatory dermatoses.





**Abstract N°: 5460****Morphea in a child treated with excimer laser : a case report**

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**Introduction & Objectives:**

Morphea is a rare sclerosing inflammatory disease. Several treatments may be indicated depending on the severity of the disease. The excimer laser has shown good results.

**Materials & Methods:**

We report a case of morphea in a child treated with excimer laser.

**Results:**

A 8-year-old child, with no notable pathological history, presented with one-year history of sclerotic lesion on the right thigh. On Clinical examination we noted an indurated plaque with an ivory center and a hyperpigmented border of approximately 7 cm on the right thigh. This lesion was slowly expanding. The biopsy had showed a vacuolation of the basal epidermal layer, a sclerosis of the superficial dermis with thickened collagen bundles, and a perivascular infiltrate of lymphocytes. The diagnosis of morphea was made. The patient was initially treated with topical dermocorticoid. The evolution was marked by the extension of the plaque. Thus, excimer light therapy was indicated as an adjuvant therapy with favorable evolution.

**Conclusion:**

Morphea is an uncommon, autoimmune, connective tissue fibrosing disease that affects the skin and underlying tissue. It is characterized by thickening and induration of the skin secondary to an increase in the amount of collagen. The disease is more prevalent in women and its prevalence increases with age. The typical clinical presentation of morphea consists of an erythematous plaque that undergoes centrifugal expansion and whose center gradually becomes indurated and often ivory in color and shiny in appearance. The lesions are usually multiple and distributed mainly on the trunk. Several treatments such as dermocorticoids, topical calcipotriene, topical tacrolimus, phototherapy and methotrexate have been used. The 308-nanometer excimer laser, used in the treatment of several inflammatory skin diseases such as vitiligo, lichen planus and psoriasis, has shown clinical improvement in morphea lesions. Its efficacy results from its anti-inflammatory action. In the literature, clinical remission was observed after 8-12 treatment sessions. The excimer laser is indicated in active lesions refractory to other treatment options or as an adjuvant therapy, as in our case.

**Abstract N°: 5845****Raccoon eyes under the dermoscope: think of localized lichen planus**

Kenza Tahri Joutei Hassani<sup>1</sup>, Zakia Douhi<sup>1</sup>, Khalil Bouayad<sup>1</sup>, Hanane Bay Bay<sup>1</sup>, Sara Elloudi<sup>1</sup>, Meryem Soughi<sup>1</sup>, Fatima Zahra Mernissi<sup>1</sup>

<sup>1</sup>Hassan II university hospital center

**Introduction & Objectives:**

Lichen planus (LP) is a well-known inflammatory condition that affects both the skin and mucous membranes. The exact cause of LP is not fully understood, but it is believed to involve autoimmune T-cell infiltration of the basal membrane and basal layer of the skin or mucous membranes. LP typically presents as shiny, violaceous, flat-topped, pruritic polygonal papules or plaques with fine white lines forming a superficial network known as Wickham striae. While LP can occur anywhere on the body, it rarely affects the eyelids. Isolated lesions of eyelid LP can pose a challenge for clinical diagnosis, as they may resemble other conditions such as contact dermatitis, psoriasis vulgaris, and lupus erythematosus, which present with papular erythematous eyelid dermatosis. We present a unique case of LP affecting both upper and lower eyelids bilaterally presenting as "raccoon eyes."

**Materials & Methods:**

We report the case of a 40-year-old patient presenting a bilateral palpebral lichen planus. The patient was examined by a DermLite 4 dermoscope and the diagnosis was confirmed by histology

**Results:**

An otherwise healthy 40-year-old woman presented with a 4-month history of pruritic lesions on her bilateral eyelids. The patient had not applied any medication or any cosmetic product to the site prior to the onset of the lesions. On examination, she had a violaceous slightly pigmented plaque over the upper and lower eyelids. Examination of the entire body did not reveal any similar lesion. All mucosae were normal. She did not have any other associated ophthalmological involvement in the form of conjunctivitis, keratitis or lacrimal duct stenosis. Dermoscopy of the lesions revealed grey-brown dots and globules, a granular annular pattern with an erythematous background.

A skin biopsy of the upper eyelid showed acanthosis, hypergranulosis, overlying hyperkeratosis, a dense lichenoid inflammatory infiltrate with mild to moderate vacuolar interface dermatitis, and prominent melanophagocytosis in the superficial dermis, confirming the diagnosis of LP. Her hemogram, blood sugar levels, and other relevant investigations were within normal limits and serology for hepatitis C virus was negative. She was prescribed a topical desonide 0,1% for three months with good improvement.

**Conclusion:**

Lichen planus of the eyelid is a condition that is often overlooked and should be considered in the list of possible diagnoses for eyelid dermatosis. Although topical steroids have shown efficacy in treating eyelid LP, further research is needed to determine the most optimal therapeutic approach for this condition.

**Abstract N°: 5850**
**Effectiveness of OR-101, a Next Generation Highly Selective JAK3 Inhibitor, in a Murine Model of Alopecia Areata**

 Fauad Hasan<sup>1</sup>, Michael Howell<sup>\*1</sup>, Amos Gilhar<sup>2</sup>, Aviad Keren<sup>2</sup>, Patricia Walker<sup>1</sup>, Wajdie Ahmad<sup>1</sup>
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**Effectiveness of OR-101, a Next Generation Highly Selective JAK3 Inhibitor, in a Murine Model of Alopecia Areata**

 Amos Gilhar,<sup>1</sup> Aviad Keren,<sup>1</sup> Wajdie Ahmad,<sup>2</sup> Patricia Walker,<sup>2</sup> Michael Howell,<sup>2</sup> Fauad Hasan<sup>2</sup>
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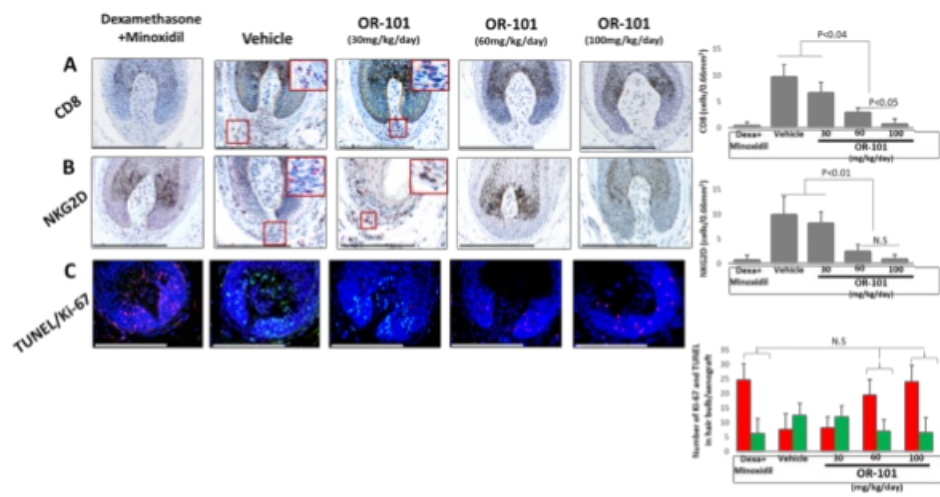
**Introduction & Objectives:** Alopecia areata (AA) is an autoimmune disease resulting in partial or complete nonscarring hair loss, with a global prevalence of approximately 1.7 to 2.1%. Evidence suggests that AA is T-cell mediated with a selective attack on the hair follicle; pathogenesis is driven, in part, through the Janus kinase (JAK)-signal transducer and activator of transcription (STAT) signaling pathway. Targeting the JAK family kinases with small-molecule inhibitors has proved to be effective in the treatment of different types of diseases and the identification of more selective pharmacologic JAK inhibitors has been an ongoing research and development goal. Here we evaluate the efficacy of OR-101, a next generation selective JAK3 inhibitor, in a murine model of AA.

**Materials & Methods:** Full thickness 3-mm skin biopsies were obtained from the scalp of normal healthy donors who underwent aesthetic procedures. Using 50 female C.B-17/IcrHsd-scid-bg mice (beige-SCID, Harlan Laboratories Ltd., Jerusalem, Israel), three 3 mm pieces were grafted into small incisions orthotopically into the subcutaneous layer of each mouse followed by the application of sterile bandages to protect the grafted biopsies. The bandages were removed 7 days post-surgery. Nineteen days following the transplantation, each mouse was injected (intra-dermally) with autologous CD8/NKG2D-enriched PBMCs (3.5X10<sup>6</sup> cells in 100 µl PBS per graft). Following the induction of alopecia areata, mice were randomized and treated for 74 days with vehicle, dexamethasone+ minoxidil, or OR-101. At the end of the study, the skin was harvested. Xenotransplants were analyzed histologically and immunohistochemically for AA parameters.

**Results:** Treatment with OR-101 was associated with dose-dependent improvements in hair growth from the initiation of treatment to the end of study (61% for 100 mg, 23% for 60 mg, 8% for 30 mg, -6% for vehicle, 73% for dexamethasone). Additionally, OR-101 dose-dependently reduced the percentage of HLA-DR and HLA-ABC expressing cells while increasing the percentage of TGFβ and αMSH expression. Finally, OR-101 treatment dose dependently reduced the number CD8+ and NKG2D+ cells surrounding the hair follicles (Figure 1)

**Conclusion:** JAK3 signaling plays a significant role in regulating the downstream signaling of receptors that employ the common gamma chain (γc) of the type I cytokine receptor family (e.g. IL-2R, IL-4R, IL-7R, IL-9R, IL-15R, and IL-21R). These receptors are often expressed on T cells that, when activated, target the hair follicle leading to the development of AA. The selective anti-inflammatory activity of OR-101 and ability to stimulate hair growth in this murine model supports the further development of OR-101 for inflammatory skin diseases including AA.

**Figure 1. Characteristic AA inflammatory infiltrates. A.** CD8 and **B.** immune cells expressing NKG2D+ cells surrounded HF's combined with **C.** excessive apoptotic cells (green) and decreased proliferative cells (pink) in the matrix hair bulb of the vehicle and JAK3/ITK/30 mg/day treated xenotransplants.



**Abstract N°: 5881****neutrophilic dermatosis of the face**

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**Introduction & Objectives:**

Neutrophilic dermatoses are a group of heterogeneous skin diseases characterized by a dense and sterile neutrophilic infiltrate on histology. Recurrent neutrophilic dermatosis of the face is an uncommon entity described first by Whittle et al. in 1968. It's considered a localized variant of Sweet's syndrome.

**Materials & Methods:**

We report a new case of this rare condition.

**Results:**

A 35-year-old male with a history of hemorrhoids and chronic constipation was admitted with a seven-day history of painful infiltrated annular erythematous plaque measuring 7 cm in diameter, covered with crusts and non follicular pustules in the temporal region. Similar lesions appeared on his cheek and left mastoid region. There was no fever nor adenopathies. The rest of the clinical examination was unremarkable. He was initially treated with pristinamycin then with amoxicillin and clavulanic acid without success. Laboratory tests were within normal ranges. Bacterial and mycological cultures of specimens of the lesion were negative. A skin biopsy was performed. The epidermis had a normal thickness with spongiosis and inflammatory cell exocytosis. The superficial and middle dermis had a dense inflammatory infiltrate rich in neutrophils. The diagnosis of neutrophilic dermatosis of the face was made. An exhaustive etiological investigation, performed to look for an associated disease showed no anomalies. He was then treated with topical steroids, with a rapid improvement.

**Conclusion:**

Neutrophilic dermatosis of the face is a rare entity of neutrophilic dermatosis. It's characterized by painful and erythematous plaques on the face, without fever, laboratory abnormalities, or associated conditions. The findings on histopathology are compatible with Sweet's, particularly neutrophilic infiltration without leukocytic vasculitis. That's why it's considered by some authors as a mild variant of Sweet's syndrome localized on the face without systemic symptoms, laboratory abnormalities. We are aware of only seven similar cases, mostly females. Neither of these previously described cases had associated conditions, such as myeloproliferative disorders, infectious diseases or inflammatory bowel diseases. All of them required systemic corticosteroids, while our patients did well with topical steroids.

**Abstract N°: 5891****The use of non-hormonal moisturizers in the treatment of patients with vulvar lichen sclerosus and genitourinary syndrome of menopause**

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**Introduction & Objectives:** Lichen sclerosus (LS) is a chronic inflammatory dermatosis, characterized by the loss of elasticity and thinning of the skin in the anogenital area. Genitourinary syndrome of menopause (GSM) includes genital and urinary symptoms, which associated with estrogen deficiency. Our observational study of patients with vulvar lichen sclerosus (VLS) and GSM revealed that women need for a complex therapy with combination of topical corticosteroids (TCS) and non-hormonal moisturizers. Topical non-hormonal moisturizers can be used as maintenance therapy for a long time, which improves the quality of life of patients with vulvar VLS and GSM. Our objective:\*\* to evaluate the effectiveness of use non-hormonal moisturizers in the treatment of patients with VLS and GSM, who were treated with topical corticosteroids.

**Materials & Methods:** Our prospective observational study included 19 patients with VLS and GSM, aged from 52 to 67 years. All patients were divided in 2 groups. In the 1st group (n=10) patients was treated by topical corticosteroids: ultra-potent topical corticosteroids (clobetasol propionate 0.05% ointment 1-2 times a day - for 1 month, then mometasone furoate 0,1% ointment 1 times a day- for 1 month). In the second group (n=9) it was used TCS and non-hormonal vaginal moisturizers (sodium hyaluronate with a phytocomplex local gel, 40 minutes after TCS, 1 time a day for 10 days, then 2-3 times per week for 3 months). The efficacy of the treatment was assessed by dermatological symptom scale index (DSSI), the visual analog scale (VAS), and the Dermatology Life Quality Index (DLQI). These scales were assessed during 3 visits (1 - after 14 days of treatment, 2 - after 1 month, 3 - after 2 months).

**Results:** The intensity of pruritus before the treatment was assessed according to the VAS at very high level ( $9.12 \pm 0.91$  and  $8.89 \pm 0.83$  points). By the 14th day of therapy, the intensity of pruritus decreased by 71.12% in the first group and by 63.25% in the second group ( $2.12 \pm 0.52$  and  $2.51 \pm 0.32$  points respectively ( $p < 0.05$ )). In the second group women, who treated by combined therapy, after 2 months had the dynamics of pruritus of 78.21%, while in the first group the dynamics was 42.54 % ( $p < 0.05$ ). [The dynamics of the DISS in the first group showed that the symptoms are regressed in 85.31%, and in the second group - 89.76%. The patients with VLS and GSM demonstrate low quality of life, as evidenced by the results of testing: DLQI for all patients before the treatment was significantly decreased and determined at  \$23.12 \pm 2.88\$  points.](#) After 2 months of complex treatment, in the first group the dynamics of DLQI was 79.23%, while in the second group it was 86.22% ( $p < 0.05$ ).

**Conclusion:** the use of potent and ultra-potent topical corticosteroids in combination with vaginal non- hormonal moisturizers for patients with VLS and GSM increases the effectiveness of therapy for regression of subjective and objective symptoms, improves women's psychoemotional state and the quality of life.



**Abstract N°: 5955****Blaschko linear atrophic pigmented plaques : Think of Moulin linear atrophoderma**Maissa Abid<sup>1</sup>, Fatma Hammemi<sup>1</sup>, Malek Cherif<sup>1</sup>, Tahya Boudawara<sup>2</sup>, Emna Bahloul<sup>1</sup>, Hamida Turki<sup>1</sup><sup>1</sup>Hedi Chaker hospital, Sfax, Department of dermatology, Sfax, Tunisia, <sup>2</sup>hbib bourguiba hospital, sfax, anatomy pathology, Sfax, Tunisia**Introduction & Objectives:**

Linear atrophoderma of Moulin (LAM) is a rare clinical entity, initially described by Moulin et al. in 1992. We report two new cases.

**Materials & Methods:**

Two patients (a 15-year-old adolescent and a 37-year-old man) with no notable medical history presented with the progressive appearance of atrophic pigmented plaques. The lesions had been evolving for 6 years (case 1) and 2 years (case 2) and were asymptomatic. The plaques were blaschko linear and located in the left deltoid region (case 1) and on the trunk, the lateral sides of the neck and the anterior sides of the wrists (case 2). They were atrophic without induration on palpation. They were not preceded by an inflammatory or sclerotic phase. In both cases, biological and immunological tests (NAA) were negative. Skin biopsies in both patients showed aspecific changes such as a perivascular lymphocytic infiltrate of the dermis.

**Results:**

LAM is a rare dermatosis. It is characterised by the appearance of atrophic hyperpigmented linear bands. It is most common in adolescents. The histological changes are mostly aspecific, with hyperpigmentation of the basal layer of the epidermis associated with a perivascular lymphocytic infiltrate of the dermis. The absence of clinical sclerosis and abnormalities of the dermal collagen fibres on histological examination allow morphea to be ruled out. No treatment has been shown to be effective until this day.

**Conclusion:**

LAM is a rare entity with only about thirty cases described. It should be considered in the presence of blaschko-linear plaques that at first become atrophied and pigmented without previous induration or inflammation. Its prognosis is mainly aesthetic.

**Abstract N°: 6042****Unilateral Sweet Syndrome: An Unusual Presentation**

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**Introduction & Objectives:**

Sweet's syndrome is a rare inflammatory disease with predominant cutaneous expression, belonging to the group of neutrophilic dermatosis.

We report a particular presentation of sweet syndrome confined to the right upper limb.

**Case report:**

A 37-year-old female patient, who has a history of rheumatoid arthritis, presented with an acute onset of painful lesions on the upper limb. No prior medication was noted.

Physical examination found a fever and general asthenia, associated with multiple tender, purplish papules and nodules, confluent in some places, located on the back of the right hand and arm. No other lesions were found on the other limbs or neck.

Blood tests showed hyperleukocytosis with polynuclear neutrophil predominance, associated with an elevation of biological markers of inflammation.

Skin biopsy showed a subnormal epidermis with diffuse edema forming a band in the papillary dermis. As well, a dense inflammatory infiltrate occupied the superficial 2/3 of the dermis and was made up of lymphocytes, plasmocytes, and many often altered neutrophils. Many congested blood vessels with turgid endothelium surrounded by the same inflammatory infiltrate were found, without any vasculitis lesion.

The systemic etiological assessment was negative aside from the pre-existing polyarthritis.

General steroids were prescribed at a dose of 1mg per kilo per day for 2 weeks, tapered gradually. The follow up showed a progressive disinfiltration of skin lesions.

**Discussion**

Sweet syndrome typically presents with a palpable erythematous plaque, sometimes pseudoblistering, and occasionally pustules occurring on the face, neck, chest, and extremities, accompanied by fever and general malaise. Lesions enlarge and may coalesce to form irregular, sharp border plaques over a period of days to weeks. Subsequently, either spontaneously or after treatment, the lesions usually resolve without scarring.

Numerous reports of uncommon clinical presentations, including solitary lesion or lesions restricted to the palmar, palmoplantar region, scutum, and backs of the hands, have been reported.

Our observations show yet another unusual, unilateral presentation of sweet syndrome.

The most consistent laboratory results for Sweet's syndrome are an elevated erythrocyte sedimentation rate and peripheral leukocytosis with neutrophilia. Which was found in our patient.



Skin biopsy is necessary for diagnosis. Histologically, the epidermis may be normal, acanthotic or the site of PNN exocytosis, sometimes it forms unilocular subcorneal pustules. There may be subepidermal detachment resulting from massive edema.

There are 3 clinical forms : classical (or idiopathic), malignancy-associated, and drug-induced. The classic or idiopathic forms are the most frequent (75%), and may be associated with infection, inflammatory bowel disease, or pregnancy.

Many reports describe associations with connective tissue diseases such as systemic lupus, but rarely described with rheumatoid arthritis, Haray and al and Chelly and al report each this unusual association.

Therapeutic approach depends on the extension, general condition of the patient, and underlying diseases. Systemic corticosteroid therapy at a dose of 0.5 to 1mg/kg/d with a slow and progressive decrease over a few weeks is the treatment of choice.

### **Conclusion**

Our case is original because of the atypical unilateral presentation, as well as a rare underlying association which is polyrhumatoid arthritis.



**Abstract N°: 6043****Pyoderma Gangrenosum of the foot and thumb in a patient with Crohn disease**Katarina Smuc Berger<sup>1</sup><sup>1</sup>General hospital Isola, Outpatient Clinic of Dermatovenerology, IZOLA-ISOLA

**Introduction & Objectives:** A case of pyoderma gangrenosum in patient with Crohn disease with lesions on thumb and dorsum of the foot.

**Materials & Methods:** Case presentation

**Results:** 72 -year-old woman with a past medical history of Crohn disease initially presented to rheumatologist due to joint pain in her right thumb and dorsum of left foot. The diagnosis of enteropathic arthritis was made and metilprednisolone 16 mg was prescribed. Due to additional swelling and pain of the foot she was taken to Emergency clinic 10 days later . Incision was performed, microbiology swab was taken. She returned to Emergency clinic 6 days later due to wound that developed on the dorsum of the foot after the incision and worsening of the situation on the thumb. Rheumatologist and dermatologist evaluated her and suspicion of pyoderma gangrenosum was made. The patient had violaceous-to-red bordered ulceration on the dorsum of left foot and pustulovesicular nodes on right thumb . Results of the Hamatoxylin-eosin staining of the bioptic material of the lesion on the foot revealed a predominantly neutrophilic infiltrate. Swab culture was negative for microorganisms. The diagnosis pyoderma gangrenosum was confirmed and treatment with metilprednisolone ( 32 mg p.o.) was continued. Further controls were by dermatologist as metilprednisolone dose was tapered and the ulceration healed in 5 month. She finished the treatment with metilprednisolone and is still stable 6 months afterwards.

**Conclusion:** Pyoderma gangrenosum (PG) is a rare dermatosis that can have many different manifestations. Quick recognition and prompt start of systemic treatment are necessary in this disease We would like to stress the importance of the interdisiplinary approach to patients with possible pyoderma gangrenosum.



**Abstract N°: 6128****Hypertrophic Lichen Planus of the External Auditory Canal**

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**Introduction & Objectives:**

Hypertrophic lichen planus is a chronic disease of unknown origin secondary to a T-cell mediated immune response. It usually affects middle-aged adults. Lesions initially develop on the flexural areas of the limbs, with a generalized rash marked by pruritus of varying severity depending on the type of lesion and its extent. It can affect all areas of the skin as well as the oral mucosa. In 85% of patients, the disease resolves within 18 months. Primary cutaneous hypertrophic lichen planus of the external auditory meatus is extremely rare. In the literature, only one case of hypertrophic lichen planus of the external auditory meatus has been found, and only 19 cases of lichen planus localized to the ear.

**Materials & Methods:**

We report a description of a case of unusual localization of hypertrophic lichen planus in the external auditory canal.

**Results:**

A 36-year-old female patient with a history of diabetes for 2 years, managed with diet alone, she presented to our department for pruritic pigmented lesions in the external auditory canal of both ears evolving for 3 years. On skin examination, a violaceous pigmented plaque of the bilateral external auditory canal was found, while dermoscopy revealed Wickham striae and grayish-blue globules. An otolaryngeal examination was performed, which confirmed exclusive involvement of the external auditory canals without obstruction. A skin biopsy was performed with histopathological examination, revealing a markedly hyperplastic epidermis covered with thick hyperorthokeratosis. The basal layer was hyperpigmented with discrete vacuolization. The dermis showed fibrosis, congestion, and a discreet lichenoid lymphocytic infiltrate, along with pronounced and deep pigment incontinence, thus the diagnosis of hypertrophic lichen planus was made histologically. Laboratory tests showed vitamin D deficiency, negative hepatitis C serology, anti-TPO antibodies, and anti-TG antibodies. The patient was treated with clobetasol twice daily and 0.025% tretinoin once daily for 6 weeks with partial improvement. Subsequently, treatment was switched to tacrolimus 0.1% twice daily.

**Conclusion:**

Very few cases have been published on hypertrophic lichen planus of the external auditory canal, which makes management difficult and not standardized.

**Abstract N°: 6144**
**An unusual presentation of oral lichen sclerosis: Mutilating form of the lower lip**

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**Introduction & Objectives:**

Lichen sclerosus is a rare mucocutaneous inflammatory disease that usually affects genital region. Involvement of the oral mucosa is less common and usually manifests as erosive reticulated white patches. We report here 2 cases of oral lichen sclerosus (LS) responsible of a deformation of the lower lip.

**Materials & Methods:**

Case 1: A 65 year old man with smoking history was referred for an atrophic lesion on the lower lip, evolving very gradually towards retractile erythematous lesion over a period of 10 years.

Physical examination revealed an erythematous, atrophic and retractile lesion; localized in the left hemilabial side extending the external third of the upper lip with a loss of substance in the cutaneous side.

The results of the laboratory tests did not reveal any abnormalities. Facial computed tomography was normal. Leishman body research and geneXpert MTB were negative.

A mucosal biopsy was performed showing an atrophic epidermis surmounted by orthokeratotic hyperkeratosis, a superficial dermis with a hyaline appearance associated with a lymphocytic infiltrate, without signs of malignancy and nor bacterial or mycotic infection. The diagnosis of LS was therefore made in front of the typical histopathological appearance.

The patient was treated with topical steroids at the active phase. After 1 year, surgical reconstruction was performed with a satisfactory result.

Case 2: A 54 year old woman presented with a 6 years history of a retractile lesion in the lower lip evolving very gradually causing lip deformation and limiting mouth opening; she had no history of local trauma.

The diagnosis of LS was made after a pathological examination of the biopsy which revealed hyperkeratosis, atrophy and focal degeneration of basal cells and a band-like mononuclear inflammatory infiltrate.

Regarding the functional impact, a surgical reconstruction was performed with great postoperative outcomes.

**Results:**

LS affecting only the oral mucosa is extremely rare. Oral LS has been described on labial mucosa, gingiva, palate and tongue. These lesions manifest as white macules or plaques with reticular striations and superficial ulceration. LS is usually asymptomatic. Only seven cases of oral LS have been described involving the lower lip. None of the reported cases showed a mutilating evolution unlike our patients.

The histologic features of cutaneous LS are very specific, including follicular plugging, atrophy of epidermis and loss of elastic fibers in the dermis.

The treatment is essentially based on local corticosteroids for active forms and sometimes oral steroids combined

with methotrexate for refractory cases. Surgical management can be proposed when there is aesthetic or functional discomfort as shown in this report.

**Conclusion:**

Through the cases described in this report, we aim to expand knowledge regarding this uncommon condition.

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**Abstract N°: 6390****Elastolytic annular granuloma: an exuberant presentation**Carolina Cruz<sup>1</sup>, Renata Nunez<sup>1</sup>, Elizabeth Drullis<sup>1</sup><sup>1</sup>University of Mogi das Cruzes, Dermatology, Mogi das Cruzes , Brazil

**Introduction & Objectives:** Elastolytic annular granuloma (EAG), also known as actinic granuloma, is a rare, benign, self-limiting disease that most often is presented in a photoexposed area. It usually occurs in adults aged 35-75 years and may present as annular plaques with an atrophic center or papular form. The pathogenesis of the disease is not yet well defined, however, it is related to inflammatory process. Regarding the histological findings: loss of elastic fibers due to phagocytosis of multinucleated giant cells associated with the presence of multiple granulomas are observed. As it is a rare disease, our objective was to report this pathology different and exuberant form of presentation and our treatment, along with the good results we were able to achieve.

**Materials & Methods:** Female patient, 64 years old, came to us complaining of red spots on the upper trunk for more than 2 years. She reported an initial lesion in the neck that progressed to the shoulders and back. The patient also complained about the feeling of itching, burning and the worsening of the lesions on hotter days. Finally, the patient reported that the lesions were very red at the beginning, which over time their center lightened. The patient had Diabetes Mellitus and hypertension as comorbidities. On physical examination, erythematous plaques with an annular configuration with raised edges and a center with normal skin color, not scaly, located in the upper trunk were observed. The sensitivity test was negative. Two biopsies of two different lesions were performed, which confirmed the diagnosis of Elastolytic Annular Granuloma. Laboratory tests showed the patient's blood glucose level of 165mg/dL and the presence of glucosuria. We then started, after an ophthalmology evaluation, Hydroxychloroquine 400mg/day and referred the patient to the endocrinologist to control her Diabetes.

**Results:** After 2 months, the patient returned with improvement of the lesions, presenting lighter color and no associated complaint. After a period of 6 months we suspended the medication due to the improvement of the lesions that were no more plaques, only light macules that we are currently treating to make it even less visible.

**Conclusion:** Elastolytic Annular Granuloma is a rare disease that can be presented in many clinical forms, in a very exuberant or more discreet way, making its diagnosis complex and the disease important to be reported, along with its treatment.



**Abstract N°: 6512****A Dysbiotic Skin Microbiome associated with inflammation and foul body odour are hallmarks of Darier's disease**

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**Introduction & Objectives:**

Darier disease (DD), also known as keratosis follicularis, is a rare genodermatosis with a prevalence range between 1/30.000 and 1/100.000. It is characterised by a mutation of the ATPA2 gene leading to disrupted Ca<sup>2+</sup> signalling and a loss of keratinocyte adhesion. Recurrent episodes of skin infections and inflammation with characteristic malodour in DD suggest a role for microbial dysbiosis. Nevertheless, DD microbiome has not been analysed so far and possible links of dysbiosis to inflammation in DD are not yet established. In this study we investigated for the first time the cutaneous microbiome dysbiosis and its consequences in DD.

**Materials & Methods:**

We collected 115 skin swabs from 14 patients and healthy matched volunteers and analysed them using 16S ribotyping. We also assessed microbiome changes in the context of DD malodour, and explored inflammation and dysbiosis signatures in DD skin transcriptomes.

**Results:**

Obtained data revealed, a disease-specific cutaneous microbiome characterised by a loss of microbial diversity and of potentially beneficial commensals. Inflammation associated microbes such as *S. aureus* and *S. warneri* dominated DD lesions and showed strong correlations with disease severity. The observed dysbiosis was also characterised by the expansion of taxa belonging to the genera *Corynebacteria*, *Staphylococci* and *Streptococci*, which were strongly associated with malodour intensity. Transcriptome analyses showed an upregulation of epidermal-repair, inflammatory and immune defence pathways reflecting epithelial and immune responses to a dysbiotic microbiome. In contrast, the downregulation of key barrier genes including claudin-4 and cadherin-4 indicated a skin barrier impairment.

**Conclusion:**

These findings highlight the role of cutaneous dysbiosis in DD inflammation and associated malodour, and suggest furthermore potential biomarkers and intervention targets.



## Abstract N°: 6559

## Dysbiosis in rosacea and its modulation by topical Ivermectin

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**Introduction & Objectives:** Rosacea is a chronic inflammatory skin disease characterized by erythema, telangiectasia and papulopustulous lesions within the centrofacial region. Its prevalence is high, ranging from 1-22% within the general population and rosacea lesions frequently show a significant increase in the density of *Demodex folliculorum*. During recent years, the mite-associated microbiome, in particular *Heyndrickxia oleronia* (formerly *Bacillus oleronius*), has been suspected to promote inflammation in rosacea patients.

**Materials & Methods:** We aimed to systematically characterize the microbiome associated with the pilosebaceous unit, disease severity (IGA) and mite density in rosacea patients (n=41) before and after 30 days of topical 1% ivermectin cream treatment when compared to age and sex matched healthy volunteers.

**Results:** Topical ivermectin cream treatment resulted in a marked or total decrease in mite density in 87.5% of Demodex-carrying patients (n=24, p<0.007). Furthermore, Investigator's Global Assessment scores were decreased in 43.9% of patients (n=41, p<0.001) during follow-up, in accordance with previously published efficacies. Moreover, cyanoacrylate-based skin surface biopsies of age-, gender- and body site-matched healthy controls (n=28) were collected for metagenomic analysis alongside respective rosacea samples. Distinct microbial community changes included a marked decrease in *Cutibacterium acnes* abundance compared to healthy volunteers, a pattern that persisted even post-treatment (p.adj=0.024). On the other hand, *Staphylococcus epidermidis* abundance was moderately increased in rosacea patients pre-treatment, and significantly increased during topical ivermectin treatment (p.adj=0.037). Further species showing shifting, albeit non-significant trends during treatment include *Corynebacteria*, *Pelomonas* and *Anaerococcus*. Regarding the Demodex-associated microbiota, a single patient with highly elevated mite counts has carried predominantly unclassified microbes based on taxonomical assignment. Intriguingly, highly similar sequences were found in lower levels in other Demodex-carrying patients and their abundance correlated with diminished or absent mite frequency after treatment. This may suggest the presence of microbial species in Demodex that are ambiguously classified and are not well-known members of the human cutaneous microbiome.

**Conclusion:** Altogether, Ivermectin treatment proved to significantly decrease Demodex density, accompanied by an overall decrease in global clinical assessment scores. Microbial communities of rosacea patients were shown to be lacking in *Cutibacterium acnes* as compared to healthy volunteers, while the abundance of *Staphylococcus epidermidis* increased significantly. These findings suggest a potential 'protective/homeostatic' role of *Cutibacterium acnes*, alongside a possible preference of Staphylococci for the inflammatory niche. Taken together, findings of the present study support that rosacea lesions are associated with dysbiosis, however, improvement of clinical signs during topical ivermectin treatment is not associated with 'normalization' of the host's microbiome but with the decrease of mite density and the reduction of potentially mite-associated microbes.





**Abstract N°: 6610****A Phase 2a, Open-Label, Single-Center, Single-Arm, dose finding 16 weeks Clinical Trial of Orismilast for the Treatment of Hidradenitis Suppurativa**

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**Introduction & Objectives:**

Hidradenitis Suppurativa (HS) is an inflammatory scarring skin disease with a substantial unmet need for treatment. Most guidelines suggest that medical, surgical and adjuvant therapy should be used simultaneously. Medical treatments include antibiotics, anti-inflammatory drugs and monoclonal antibodies. An investigator-initiated trial of apremilast has suggested that oral Phosphodiesterase-4 (PDE4) inhibitors may be beneficial in the management of HS, although they are currently limited by frequent gastro-intestinal adverse events. In this study, we examined tolerability, safety, and utility of orismilast, a potent selective PDE4B/D-inhibitor in HS.

**Materials & Methods:**

A phase 2a, single-arm, single site, open-label, 16-week trial was conducted in mild to severe HS. Following inclusion in the trial, patients started treatment with orismilast 10 mg daily. If tolerated, it was rapidly up-titrated to a max of 40 mg BID. More individualized up-titration to the individual patients' maximal dose was allowed to improve tolerability. The primary endpoint was the occurrence and magnitude of adverse effects, as well as percent change in the total number of abscesses and nodules (AN-count) at week 16.

**Results:**

Approximately half (9/20) of the included patients completed 16 weeks of treatment. The mean AN-count was reduced by 33.1% in those who completed vs. 12.0% in those who did not. HiSCR50 was achieved by 67% vs. 27%, respectively.

**Conclusion:**

Orismilast therapy in HS patients showed a dose dependent tolerability, with predominantly mild to moderate adverse effects. The use of lower max doses (20-30 mg BID) and slower titration demonstrated the ability to reduce early PDE4-inhibition related side effects. Data indicate that this broader blockade of the inflammatory process may be a fruitful pursuit in HS.