

**Abstract N°: 192****Cutaneous bronchogenic cyst presenting as a keloid on the back.**

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**Introduction & Objectives:****Materials & Methods:****Results:**

Bronchogenic cyst is a rare congenital condition that occurs due to an impaired process of lung development. The pathogenesis involves a set of cells that separate from the rest, which differentiate by themselves and develop a small lung bud. Such a cyst is usually diagnosed in early childhood and manifests clinically as a tumor or an orifice draining fluid, symptomatic in about half of the cases. The treatment of choice is surgical removal.

The patient first presented to dermatology ambulatory care at the age of 9. She had already undergone previously two surgeries due to a small tumefaction, draining seropurulent exudation, located in the scapular region. The excised tissue was examined by the pathologist and revealed a sebaceous cyst. The patient's parents decided to visit a doctor for another appointment because, despite those two previous procedures, they were concerned about the persistent drainage from a small orifice on the surface of a lesion that resembled a keloid scar, which was additionally constantly growing. Upon dermoscopy, the lesion displayed linear vessels arranged peripherally over a pink to white structureless background, resembling a keloid. On the latest evaluation, dermoscopy of the grown lesion additionally showed pigmented tan lines, and circles arranged in a linear fashion, located also on the periphery, which could have corresponded to pigmentation of the lesion due to melanocyte activation. The patient's parents did not agree to another excision or biopsy of the lesion at that time. Later, the patient presented for the second time one year later due to further enlargement of the lesion, but this time a deep shaving biopsy was taken in order to exclude dermatofibrosarcoma protuberans. Unexpectedly, the result of the histopathological examination was a cutaneous bronchogenic cyst.

It is mostly believed that extracutaneous bronchogenic cysts located on the periscapular area develop due to an abnormal budding of the tracheobronchial primitive foregut during early embryogenesis, with migration to the thorax. The most common extrapulmonary localization of bronchogenic cysts is mediastinum however, in our case, it was the scapular region. To the best of our knowledge, there have been only 15 other reported cases of scapular bronchogenic cysts, of which most were male children, unlike our case. As far as we are concerned, there are no reports on dermoscopy of this disorder. In the described patient, the clinical picture involved a slowly growing tumor with pus leakage indeed, which is the usual presentation of bronchogenic cyst. However, later it resembled a keloid, which was symptomatic and had grown over time. Possible complications include rupturing of the cyst or its compression, infection and, noteworthy, bronchogenic cysts may be the origin of malignant transformation. Considering this possibility, dermatologists should be aware of such entity to early diagnose and remove the cyst.

**Conclusion:**



**Abstract N°: 343****Systemic lupus erythematosus and MECP2 gene mutation**

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**Introduction:** Systemic lupus erythematosus (SLE) is an autoimmune disease in which the genetic component plays a key role in its pathogenesis. There are more than 30 genes that cause monogenic lupus, and numerous polymorphisms have been described that increase the risk of developing SLE, including those of the MECP2 gene. We present the case of a boy with SLE and a pathogenic mutation in MECP2

**Case Report:** A 12-year-old boy presented with a 1-month history of lesions on the hands, feet and pinnae that had been present for 1 month. He had a history of autism spectrum disorder and rolandic epilepsy. Physical examination revealed erythematous-violaceous maculopapules symmetrically affecting the palmar aspect of the fingers, soles and lobes of both pinnae. He also presented with neuropsychiatric symptoms consisting of motor slowing and a tendency to somnolence. A skin biopsy was performed and was compatible with lupus erythematosus. Laboratory tests showed leukopenia, ANA 1/1280, positive anti-DNA and hypocomplementemia. With a diagnosis of SLE, the patient was treated with bolus methylprednisolone and cyclophosphamide, with clinical and laboratory improvement. A genetic exome study identified the pathogenic variant for MECP2:NM\_004992.3:c.1415\_1416del:p.E472Gfs\*14. Six months later, the patient presented a new episode of psychotic symptoms, with no other signs of SLE activity.

**Discussion:** There are polymorphisms in the MECP2 gene that increase the risk of SLE. In addition, mutations in this gene are responsible for Rett syndrome and disorders related to the MECP2 gene, ranging from intellectual disability to neonatal encephalopathy. In our case, a mutation in the MECP2 gene was found, which would explain, on the one hand, the psychiatric picture and, on the other hand, the development of SLE. However, in this case, it is a mutation and not a polymorphism, which is not described in the literature in the context of SLE. The mechanism by which this gene may induce the development of SLE is based on epigenetic changes that would lead to an overexpression of genes that would induce the exaggerated synthesis of interferon. In any case, the existence of neuropsychiatric symptoms that do not respond to SLE treatment should lead to the suspicion of mutations in the MECP2 gene.

**Abstract N°: 376****The Novel Treatment of Children with Recalcitrant Viral Warts using Microwave Technology**Katie Solomon<sup>\*1</sup>, Sophie Dolman<sup>1</sup>, Tanya Cardwell<sup>1</sup>, Vincent Yip<sup>1</sup><sup>1</sup>Alder Hey Children's Hospital Liverpool, Department of Dermatology, Liverpool, United Kingdom**Introduction & Objectives:**

Cutaneous infection by the human papillomavirus (HPV) can lead to the formation of small, firm, keratinised lesions of the skin. Depending on their location, these are called 'common warts' (on the body in general), or verrucae/plantar warts (on the soles of the feet). Although benign, if left untreated these warts can grow into large mosaics which can be painful and unsightly. They also carry a risk of autoinoculation as well as potential to spread to other people. While some lesions can resolve within a few months, the majority can be present for many months to years due to the virus' innate ability to hide from the host immune system. Here, we carried out a retrospective service evaluation to assess the feasibility of microwave energy as a treatment for warts and verrucae in paediatric patients.

**Materials & Methods:**

Using a microwave device designed and CE marked for use in Dermatology, a total of 35 children aged 6 to 19 years old were treated at a paediatric dermatology clinic in the UK with a non-ablative dose of microwave energy (up to 10 watts for 2 seconds; 8GHz) applied to each lesion area. Each lesion received 3 to 10 successive dose repeats of microwave energy before moving on to the next lesion. Patients returned to the clinic after 3 to 4 weeks, at which point a further treatment with microwaves was administered as before. If required a new appointment was scheduled another 3 to 4 weeks later.

**Results:**

Using this treatment regime, 24 patients (68.6%) had full resolution within 8 (mean=3, median=2.5) treatment visits. Microwave treatments can be associated with a degree of short-lived, acute pain, and in these cases a total of 8 patients (22.9%) elected to discontinue the treatment and were subsequently classed as 'non-resolved'. 27 patients (77.1%) tolerated the treatment well and both patients and parents reported satisfaction with the microwave therapy outcome. Almost all patients (94%) had a history of failed alternative treatments for their warts, including topical salicylic acid preparations, Cantharidin and cryotherapy.

**Conclusion:**

This small case series with excellent outcomes suggests that microwave therapy could become a new efficacious tool for the treatment of viral warts in children. Cryotherapy or topical imiquimod creams are currently the main choices of treatment across many practices, however, microwave therapy offers a less destructive and less painful modality and may only require treatment on a subset of lesions. This would not only shorten clinic visit durations, saving both patient and clinician time, but also make the procedure less traumatic to young children undergoing the treatment. Further research into the safety and efficacy of microwave therapy and its effect on the underlying biomolecular and immune responses are required.



**Abstract N°: 423**

**Generalized Cutis Marmorata Telangiectatica Congenita - case report**

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**Introduction & Objectives:**

Cutis marmorata telangiectatica congenita (CMTC) is a capillary malformation characterized by reticulated marbled erythema which disappears with pressure and does not resolve with heating. It is often reported as benign, however can be associated with limb asymmetry, congenital glaucoma, and central nervous system dysfunction.

**Materials & Methods:**

We present a case of an 11-year-old boy, presenting generalized and symmetric marbled spots and angiokeratomas in the genital region since birth. On dermatological exam, the lesions disappear with pressure, but not with heat. He presented an important asymmetry in length and size between the lower limbs and there were no alterations on skin biopsy. The patient also presented a slight delay in neuropsychomotor development and was admitted to our outpatient clinic for diagnostic investigation by pediatrics due to tonic-clonic seizures. He had never followed up with Dermatologist nor Pediatrician and had no family history. We referred to an ophthalmological exam, in which there were no alterations, as well as to an otorhinolaryngologist, in which evaluation presented telangiectasias on rhinoscopy and no alterations on bronchoscopy or upper digestive endoscopy. Considering the clinic, the diagnosis of Cutis Marmorata Telangiectatica Congenita generalized was made.

**Results:**

CMTC is a capillary malformation characterized by reticulated marbled erythema. It can be associated with cutaneous atrophy, ulcerations, glaucoma, limb asymmetry, neurological, ophthalmological, cardiovascular, genitourinary, abdominal and endocrinological defects as well as mongolian spots, port-wine stains and dysmorphic features. Although in our case the lesions were exuberant, the patient had no dermatological follow-up, initiating it only after the tonic-clonic seizure. The presence of marbled lesions guided our diagnosis, nonetheless the lesions are generally unilateral, and the association with angiokeratoma is poorly reported in literature. Our patient did not present ulceration or atrophy and had involvement of the face and scalp. Also, despite the generalized lesions, he had an increase in the length of one of his legs without associated hypertrophy, excluding other possible syndromes.

**Conclusion:**

The CMTC is a rare diagnosis, with about 40 generalized cases reported. Being aware of this diagnosis is important for multidisciplinary referral as well as regular ophthalmologic and neurological follow-up.

**Abstract N°: 478****Hemangioma associated with an amniotic band of the forearm**

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**Introduction & Objectives:** Hemangioma is a benign tumor proliferation of cells endothelial cells expressing GLUT1, affecting 7 to 10% of children by the age of 1 year and up to 30% of premature infants weighing less than 1,500 g. segmental forms are readily associated with malformations.

**Materials & Methods:** A special case is reported of the association of a non-segmental hemangioma and an amniotic band of the forearm.

**Results (Observation) :** A girl was 2-year-old, from an out-of-consanguineous marriage with a pregnancy of 32 weeks with amenorrhea. The 2<sup>nd</sup> of a family of two sisters. She was presented in consultation with two hemangiomas; one of the forearm, the other of the abdomen, associated with agenesis of the ipsilateral left hand. Clinical examination found stable tuberous hemangiomas after a growth phase with agenesis of the left hand following amniotic clamping. A check up in search of other malformations; Abdominopelvic ultrasound, echodoppler of the limbs, were without abnormalities. The girl is a candidate for the prosthesis.

**Conclusion (and discussion):** The hemangioma is often isolated, except, segmental hemangiomas are readily associated with extracutaneous abnormalities. The "PHACE syndrome", defined by the association of a facial segmental hemangioma with cerebral, ocular and cardio-aortic anomalies, the "PELVIS" / "SACRAL" syndrome describes the association of a segmental hemangioma of the region of the swaddling (lumbosacral, gluteal or perineal region = napkin hemangioma) with dysraphia affecting in varying proportions the lumbosacral spine, the terminal medullary cone, the genitourinary organs and the anal region. In the literature, two cases of segmental hemangioma associated with an amniotic band (Bourrat, and Salhi). Our observation is particular by the site of the amniotic band in the forearm which was responsible for agenesis of the hand. and non-segmental hemangioma.

In Conclusion: when faced with a hemangioma, think about possible malformations such as the amniotic bands which are responsible for agenesis.

**Abstract N°: 589**

**not to ignore Pityriasis Versicolor in children aged 7 years (2015 - 2022)**

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**Introduction & Objectives:**

Pityriasis versicolor is a superficial mycosis uncommonly reported in children. It occurs frequently in warm humid climates. Clinical diagnosis can be confirmed by mycology examination of a biopsy sample or a patch – test. The aim of our work was to assess the frequency of pityriasis versicolor in children in our department (one department for a population of 1 million inhabitants).

**Materials & Methods:**

a retrospective analysis was conducted in patients meeting the following inclusion criteria over a 7 years period : age < 14 years, clinical presentation compatible with pityriasis versicolor, a positive patch – test. Age, sex, clinical features and favoring factors were recorded for all patients. Patch tests were performed in a control group of age – matched children with eczema or vitiligo.

**Results:**

Pityriasis versicolor was diagnosed in 1450 cases during the study period including 279 cases children 20%. Age varied from 1 year to 14 years (mean 12 years), with a slight female predominance. Facial lesions were the most frequent (n=147 ; 53%), preferentially on the forehead (n=131; 47%). The adhesive tape tests were negative in all controls

**Conclusion:**

Pityriasis versicolor is not rare in children in our department. The adhesive tape tests provides a specific diagnostic tool. Preferential facial localization and predominant a chromic and hypochromic aspect are characteristic of childhood pityriasis versicolor in our region. These 2 features are also reported in the literature. Childhood cases suggests the pathogenic factors involved in pityriasis versicolor should be reconsidered. Pityriasis Versicolor should be included in the differential diagnosis of childhood hypopigmentation.



**Abstract N°: 699****Incontinentia pigmenti Stage 1 is not simply vesiculo-bullous but vesiculo-pustular**Tubanur Cetinarslan<sup>1</sup>, Regina Fölster-Holst<sup>2</sup>, Dirk Van Gysel<sup>3</sup>, Matthias Buchner<sup>2</sup>, Rudolf Happle<sup>4</sup><sup>1</sup>Manisa Celal Bayar University, Dermatology, <sup>2</sup>Universitätsklinikum Schleswig-Holstein, Dermatology, <sup>3</sup>O.LV. Hospital Aalst, <sup>4</sup>University Medical Center Freiburg, Dermatology**Incontinentia pigmenti Stage 1 is not simply vesiculo-bullous but vesiculo-pustular****Introduction & Objectives:**

Incontinentia pigmenti (IP) is a rare X-linked dominant, male-lethal disorder characterized by pathognomic skin lesions. Other defects involve the teeth, the eyes and the brain. As described in the literature the typical cutaneous changes follow the pattern of Blaschko's lines and develop in 4 stages that usually start at birth: I) erythema and bullous lesions, II) verrucous lesions, III) hyperpigmentation, and IV) hypopigmented atrophy. Stage 1 is called vesicular or bullous or inflammatory. The vesicles are rapidly filled with eosinophils and thus turn into pustules.

**Materials & Methods:**

We performed a literature search between 1947-2022 using "Incontinentia pigmenti" resulting in 1267 hits, "incontinentia pigmenti newborn" in 321 hits, "incontinentia pigmenti vesicular" in 52 hits, "incontinentia pigmenti stage 1" in 29 hits and "incontinentia pigmenti pustular" in 7 hits. 108 publications showed a total of 153 clinical pictures from stage 1.

**Results:**

We found 13 publications that used the term of "pustul", "pustular" or "vesiculopustular" to describe the lesions of stage 1 of IP. Stage 1 is histopathologically characterized by eosinophilic spongiosis and intraepidermal vesicles containing eosinophils. Acanthosis, many dyskeratotic (apoptotic) keratinocytes in the epidermis, numerous eosinophils and some lymphocytes in the dermis are seen. Moreover, a pronounced increase of eosinophils is found in the blood of newborns with stage 1 of IP. Diagnosis can be difficult in the early stages of IP. If the Blaschko-linear arrangement of lesions is overlooked in newborns, the skin disorder may be confused with other pustular diseases such as neonatal herpes, neonatal or congenital candidiasis, scabies, staphylococcal infections like impetigo, and erythema toxicum neonatorum or transient neonatal pustular melanosis.

**Conclusion:**

Cutaneous findings of IP present in the early neonatal period and, stages can sometimes overlap each other. There are many dermatological diseases in the differential diagnosis of cutaneous manifestations and patients may remain undiagnosed since these cutaneous lesions have a self-healing course. Although dermatological manifestations are innocent, extracutaneous involvement may lead to serious complications such as blindness and neurological developmental anomalies. Dermatologists and pediatricians have an important role in early diagnosis of IP. We recommend to add the word 'pustular' to the first phase of IP. The renaming of stage 1 as 'vesiculopustular' would be far more specific than 'vesicular' or 'bullous' or 'inflammatory'.

**Abstract N°: 743****Hematohidrosis versus Munchausen's syndrome: diagnostic challenges in pediatric dermatology**Eglé Janušonytė<sup>1</sup>, Jori Punchera<sup>1</sup>, David Alvarez Martinez<sup>1</sup>, Emmanuel Laffitte<sup>1</sup>, Yassaman Alipour Tehrani<sup>1</sup><sup>1</sup>Hôpitaux Universitaires de Genève (HUG), Genève, Switzerland**Introduction & Objectives:****Materials & Methods:****Results:**

A 9-year-old girl was referred to our clinic due to episodic localized pain and recurrent bleeding with no apparent injury from her right index finger for 1 week. She had approximately three episodes of these symptoms per day lasting for about 10 minutes with spontaneous resolution. The nonsteroidal anti-inflammatory drugs had no effect on the pain. According to her parents, she was generally anxious and experiencing a conflicting and stressful situation with her sibling at home. Therefore, paediatric psychiatric care follow-up was recently initiated. There was no systemic symptom associated and the patient was otherwise in excellent health.

The clinical examination revealed no particular finding, notably no erythema, no hematoma, no petechiae and overall, no skin barrier injury. The laboratory tests revealed no abnormalities, in particular the complete blood count, partial thromboplastin time, fibrinogen and INR were within the normal range. Local treatment by antiperspirant was proposed. Two weeks following the initial consultation, the patient returned to the emergency department twice due to the condition extending to the face and extremities, as well as an increased duration of the episodes. Treatment with propranolol 0.5 mg/kg twice per day was prescribed. Paediatric psychiatric follow-up was reinforced, and risperidone 0.25 mg per day was also introduced. Adult witnesses of the family were never present during the onset of the episodes, but video and photographic documentation was provided, and the sample swab taken by the patient's mother confirmed the presence of occult blood following microscopic examination.

The concluded diagnosis was suspected hematohidrosis. A skin biopsy was not performed due to limited added value for the diagnosis, as there were no apparent skin lesions present. At the three-month follow-up, the parents reported the complete resolution of symptoms. We proposed a gradual reduction of propranolol.

**Conclusion:**

Hematohidrosis is an extremely rare condition that mainly affects young women. In reported cases, anxiety or emotional stress were common precipitating events. The pathophysiology is poorly understood, but the main hypothesis proposes possible constriction of blood vessels around the affected sweat glands during intense stress. To our knowledge, this is the first reported case in Switzerland.

Despite the video evidence provided by the mother and the patient denying self-inducement of the symptoms, the episodes were never observed by a healthcare provider, and we could not rule out the factitious origin of the illness with a presentation of Munchausen's syndrome. Hence, a definitive diagnosis has not been established despite close collaboration between the dermatologists, pediatrician, and pediatric psychiatrist.

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**Abstract N°: 769****Ulcerated hemangioma: pulsed dye laser and oral propranolol.**

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**Introduction:**

Hemangioma is the most frequent benign tumor of childhood. Part of these tumors can present complications. Ulceration is one of the most frequent. Present in 5-10% of infantile hemangiomas, especially in the proliferative phase.

**Case report:**

5 month old patient. She had no family or personal history of interest. She consulted for a lesion on the left scapula that appeared 2 weeks after her birth and that grew rapidly until it ulcerated. On examination, she presented a 5.5\*4 cm, voluminous, vinous-red tumor with a 1cm ulcer. The diagnosis was ulcerated focal hemangioma. We decided to start treatment with oral propranolol in increasing doses until reaching doses of 3mg/kg/day together with pulsed dye laser (Cynosure Cynergy 7mm, 0.5ms and 7.5Jl). Sessions initially monthly. With the first laser sessions, the ulcer had disappeared. When she had 3 laser sessions, the volume loss was considerable. We achieved complete remission after 7 months of treatment and after 5 sessions. At present, only fibrofatty tissue remains.

**Discussion:**

Ulceration of hemangiomas is associated with pain, bleeding, risk of superinfection, and unsightly scars or scars that can cause functional loss. For all of this, it requires an agile and effective therapeutic attitude that manages to reverse the explosive growth of the lesion. The use of propranolol is widely described in both ulcerated and non-ulcerated hemangiomas. However, since 2008, combined treatments of pulsed dye laser and oral propranolol have been increasingly used in ulcerated hemangiomas. This combination shows a much faster and more effective response in relation to the control of pain, bleeding and ulcer re-epithelialization.

**Conclusion:**

The synergistic action of propranolol and laser in the healing of ulcerated hemangioma makes it the ideal therapeutic option in the treatment of these patients.



**Abstract N°: 770****Ichthyosis prematurity syndrome**

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**Introduction:**

Ichthyosis, prematurity, respiratory complications, and persistent eosinophilia are the guiding symptoms of ichthyosis prematurity syndrome. It is an autosomal recessive congenital ichthyosis. It is associated with complications in the mid-trimester of a pregnancy leading to premature births.

**Case report:**

Premature 34+3 weeks/2,360 g, with no gynecologic and obstetric history. It presented skin similar to vermix, very thickened with very marked and deep folds. He did not present any other associated clinic. In the analytical controls, only eosinophilia stood out that persisted throughout the admission. He began treatment with liquid vaseline and magistral formula with 5% urea in the most hyperkeratotic regions. We confirm the suspicion, ichthyosis prematurity syndrome. The genetic study revealed compound heterozygosity for the SLC27A4 gene: c.1322dup (p.g442Rfs\*2) and c.1387T>C (p.Y463H).

**Discussion:** \*\* Ichthyosis prematurity syndrome is a very rare form of congenital ichthyosis whose prevalence is estimated at 1/200,000 births worldwide, being more frequent in regions between Sweden and Norway. It is due to recessive mutations of the SLC27A4 gene. The mutation in the case that we present, classified as pathogenic, has only been described in two other Spanish patients. During the prenatal period, amniotic fluid may have a snowball ultrasound appearance and there may be polyhydramnios, which usually leads to preterm labor. Epidermal debris is responsible for polyhydramnios, as well as respiratory distress at birth. These accumulate in the digestive tract and respiratory tract. Ichthyosis usually evolves favorably and persists in a mild form in adulthood, frequently associated with atopic dermatitis.

**Conclusion:**

Knowing these clinical signs makes it possible to guide the diagnosis and provide families with adequate genetic counselling.

**Abstract N°: 846****Self-Resolving Papules and Nodules in a Pediatric Patient**

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**Introduction & Objectives:**

Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis. Its etiology is still unknown and is usually a self-limited condition. They usually present with bilateral cervical lymphadenopathy but 40-43% present with extra nodal disease. Common extra nodal sites of involvement include the skin (10%), nasal cavity (11%), bone (5%–10%), orbital tissue (11%) and central nervous system (5%).

It is more frequently seen in children and young adults, with the cutaneous form being more common in older Asians and Caucasian females. Patients with cutaneous involvement usually lack an association with systemic or extracutaneous disease and tends to remain localized.

Morphology includes painless, nonpruritic nodules, plaques, or papules<sup>1</sup> varying from yellow to red to brown. Others described acneiform lesions and eruptive xanthoma-like lesions. Differential diagnosis includes acne vulgaris, varicella-zoster virus, sarcoidosis, cutaneous lymphoma, verruga peruana and other malignancy metastasis.

Observation can be considered for patients with uncomplicated lymphadenopathy or asymptomatic disease as 20%-50% of patients with nodal/cutaneous disease will have spontaneous remissions. Surgical excision can be considered for unifocal extra nodal disease or symptomatic airway, cranial, spinal or sinus compromise. Radiotherapy can be beneficial in refractory soft tissue and orbital bone disease, resistant airway obstruction or for palliative care. Different combinations of chemotherapeutics have been used including low-dose MTX, 6-MP, vinblastine, vinorelbine and 6-thioguanine. Other treatments such as steroids (with variable responses), sirolimus and immunomodulatory therapy TNF- $\alpha$  inhibitors thalidomide and lenalidomide have been reported.

**Materials & Methods:**

This is a case report. Prior informed consent was obtained from the patient.

**Results:**

An 8-year-old female with no known medical conditions, with a history of one month when she presented multiple asymptomatic lesions around the abdomen area. Physical examination revealed erythematous papules and nodules, some of them umbilicated and excoriated and some hypopigmented macules located around the abdominal, lumbar, and gluteal regions, with no hepatomegaly, splenomegaly, or adenopathy.

Metabolic panel, complete blood count, coagulation and immunoglobulin levels were under normal limits, a test for HIV was non-reactive. Cranial, thoraco-lumbar spine, pelvis, and long bone x-rays revealed no abnormalities.

Histopathologic examination showed a lesion formed by large multinucleated histiocytes with presence of intact lymphocytes inside (emperipolesis) associated with a mixed inflammatory infiltrate with predominance of lymphocytes and plasma cells forming a nodular lesion at the papillary and reticular dermis.

Immunohistochemistry profile revealed S100+, CD68+, lysozyme+, CD45+, CD1a-, langerin-, CD34- and actin-.

Molecular biology for Bartonella spp was negative. Based on the findings, the diagnosis was cutaneous Rosai-Dorfman disease.

A wait and see approach were followed and one month later the patient came for the biopsy result, at the physical exam most of the lesions had cleared up with multiple hypopigmented macules and atrophic scars.

**Conclusion:**

We present here a clinical case of an Rosay dorfman disease presenting in a pediatric patient which is considered a rare entity that should be remember by physicians.

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**Abstract N°: 959****H Syndrome: three new cases from Morocco**

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**Introduction & Objectives:**

H syndrome is an autosomal recessive disorder with systemic manifestations. It is named after the alliteration of its most common clinical features. We report the clinical, biological, and histological findings of 3 new cases of H syndrome that were diagnosed in Moroccan children.

**Case reports:****Patient 1:**

19-year-old girl from a first-degree consanguineous marriage, no similar case in the family.

Hyperpigmented lesions, indurated, appeared at the age of 14 years, on the folds and inner side of the thigh. No hypertrichosis. Associated with a statural delay (-3th percentile), right hypoacusis, halus valgus and recurrent fever.

The biological and radiological work-up were without abnormalities.

Histology showed an acanthotic epidermis with keratinocytic hyperpigmentation and a moderate inflammatory infiltrate of lymphocytes, hystiocytes and some plasma cells.

Genetic study for the mutation of the SLC29A3 gene not done due to lack of means.

**Patient 2 :**

6-year-old boy from a first-degree consanguineous marriage, no similar cases in the family.

Hyperpigmented and indurated patches, which appeared at the age of 3 years, on the inner thighs, pelvic and lumbar region, without hypertrichosis. Associated with polyadenopathies, and recurrent fever.

The biological workup showed a significant inflammatory syndrome.

Abdominal ultrasound revealed splenomegaly. Histologically, the epidermis showed acanthosis with basal hyperpigmentation, the dermis was fibrous with a polymorphic mononuclear, lymphoplasmacytic and mast cell infiltrate.

The genetic study for the SLC29A3 gene mutation was not done due to lack of resources.

**Patient 3:**

An 11-year-old boy from a first-degree consanguineous marriage, operated on for subaortic membrane at the age of 7, who presented with bilateral and symmetrical hyperpigmented macules on the lower limbs, trunk and face, which appeared at the age of 3 years, associated with hypertrichosis, delayed stature and right hypoacusis.

The biology shows an inflammatory syndrome, the abdominal ultrasound showed a homogeneous hepatomegaly

and polyadenopathies.

Histology showed a similar appearance to case 1 and 2.

### **Discussion:**

Syndrome H is a rare autosomal recessive disease due to a mutation in the SLC29A3 gene . The majority of published cases are of Arab origin, with a few new cases from Japan, India and USA. The disease appears in early childhood, with progressive evolution, causing a delay in diagnosis in patients.

It should be considered in patients with hyperpigmented, indurated lesions with hypertrichosis, associated with short stature, halus valgus, hypoacusis and recurrent fever. The biological workup finds an inflammatory syndrome, hyperglycemia, hypertriglyceridemia, azoospermia. Radiology may find hepatosplenomegaly, cardiopathy or polyadenopathy. Histology shows an acanthotic epidermis, with keratinocytic hyperpigmentation and deep dermal inflammation, with infiltrates rich in plasma cells, histiocytes and lymphocytes.

Genetic study by identifying the SLC29A3 gene mutation is the key test to confirm the diagnosis.

Several therapeutic strategies being explored, corticosteroids are the most used

### **Conclusion:**

H syndrome is suspected in patients with typical skin manifestations, systemic abnormalities, and endocrine pathology. Molecular genetic analysis is the key to its diagnosis. Here, we report 3 challenging cases in which dominant skin involvement made the diagnosis possible



**Abstract N°: 971****Acral blisters in neonates**

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**Introduction & Objectives:**

Neonatal pemphigus (NP) and neonatal bullous pemphigoid (NBP) are rare and transient resulting from the passive transplacental transfer of maternal autoantibodies to fetus. We report 2 cases of NP and a case of NBP with particular clinical presentations.

**Materials & Methods:**

Case reports.

**Results:**

Two newborn boys presented in the first hour of life flaccid fluid-filled blisters and erosions on the hands and feet. Both mothers were followed-up for pemphigus. Based on the mother's medical history and the patient's lesions, the diagnosis of NP was retained. They did not present new lesions after a 3-month follow up.

A newborn boy presented at birth with diffuse urticarial plaques on the trunk, face and limbs associated with post blistering erosions of the hands and feet. His mother was followed-up for gestational pemphigoid. Lesions resolved after few weeks under topical corticosteroids without recurrence while the diagnosis was a NBP.

**Conclusion:**

NP often affects the trunk and the cephalic region. Exclusive involvement of palms and feet is unusual. Unlike NP vulgaris, which is relatively frequent, NP foliaceus is a very rare entity. It may be a result of desmoglein 3 over expression in neonatal epidermis. Blisters in NBP, affecting 2 to 3% of newborns of mothers with gestational pemphigoid, can be located in the trunk, limbs and acral areas. The clinical aspect could wrongly point to an infectious disease hence, the importance of the mother examination and medical history screening. Biopsy and DIF are not recommended if the mother has typical lesions. NP and NBP have a good prognosis and usually resolve within the first 3 to 5 weeks of life. There is no correlation between the severity of the disease in the baby and the mother. However, stabilization of the disease before conception remains essential.



**Abstract N°: 981**
**The association between Transforming growth factor- $\beta$ 1 (TGF- $\beta$ 1) gene polymorphism with papular urticaria and epilepsy**

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**Introduction & Objectives:** The immune and the nervous systems are highly sensitive systems that may cross talk via cytokine network, among them with the Transforming growth factor- $\beta$ 1 (TGF- $\beta$ 1). As the immune regulator, TGF- $\beta$ 1 it is critical cytokine for immune homeostasis, but also may have a crucial regulatory role in neurogenesis. Its altered secretion and signaling may lead to inappropriate regulation of immune response and/or adverse neuroimmune interactions during brain development. TGF- $\beta$ 1 is an important regulator in the brain responses to injury and inflammation, implicated in the pathophysiology of febrile seizure, epilepsy, neurologic abnormalities and autism. Papular urticaria represents a specific hypersensitivity reaction characterized by chronic and recurrent papules and papulovesicles, subepidermal edema, pyoderma, extravasation of erythrocytes, interstitial eosinophils, and exocytosis of lymphocytes, where the scratching may cause erosions and ulcerations. It is caused by a hypersensitivity reaction to the bites of mosquitoes, fleas, and other insects. The children with papular urticaria can suffer very often from atopic dermatitis. The aim of our study as to evaluate a possible association between TGF- $\beta$ 1 SNP gene polymorphism with papular urticaria and epilepsy in a 4-year-old male children.

**Materials & Methods:** Clinical examination revealed numerous erythematous papules excoriated partially. Diagnosis of papular urticaria has been confirmed by histopathological examination, with present epidermal bulla infiltrated by inflammatory cells and degeneration of epidermal cells. Fibrosis of the superficial dermis has been present. PCR genomic analysis has been performed after DNA isolation from whole blood sample, according to the manufacturer's instructions.

**Results:** Genotype polymorphism of the TGF- $\beta$ 1 gene has been confirmed as heterozygous for CT-509C>T (120bp, 74bp and 46bp), compared to homozygous for CC (74bp and 20bp) wild type or TT type (120bp). No any allergic reaction has been documented in allergen testing. HSV IgM (21NTU/mL) and Cytomegalovirus IgM (12NTU/mL) have been documented. A mild increase of CRP (13.5mg/L) has been documented, while IgE, IgA, IgG and C3 were in normal value. Among the HLA class, the HLA-DR-15 has been detected.

**Conclusion:** About one-third of all pediatric clinical examination belong to the cutaneous manifestations, represent a common problem for pediatric population, while papular urticaria may contribute to about 5% of pediatric patients. Some previous studies suggested the association of different autoimmune, allergic and chronic inflammatory skin diseases with SNP polymorphism for TGF- $\beta$ 1 -509C>T and it was documented in a variety of immune, inflammatory and different forms of brain injury diseases. Altered serum levels and TGF- $\beta$ 1 action may be dependent on several genetic and environmental factors. Given the key role of TGF- $\beta$ 1 in brain development, allergy and inflammation, further analyses of gene-environment interaction based on larger sample sizes will contribute to better understanding of their common appearance in order to establish some specific relationships



and prevent their appearance.

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**Abstract N°: 1177****Recurrent palmoplantar desquamative erythema : Which diagnosis?**

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**Introduction & Objectives:**

Palmoplantar desquamative erythema is a frequent reason for consultation in dermatology. Several diagnoses are to be discussed.

**Materials & Methods:**

We report a case of an infant with recurrent desquamative acral erythema in relation to Fereol-Besnier disease.

**Results:**

This is an 18 month old baby girl from a first-degree consanguineous marriage, who presented since the age of 1 year recurrent episodes of flaky desquamative erythema at palmoplantar surface, aggravated by the exposition to water, spaced 4 to 8 weeks apart and regressing in 3 days. There were no concomitant infections, medication, digestive disorders or similar cases in the family. The clinical examination showed a healthy infant in good general condition with a palmo-plantar desquamative erythema; a positive bucket sign, without associated adenopathies or hyperhidrosis.

In front of this picture, several diagnoses were discussed in particular the disease of Fereol-Besnier in its localized form, the palmo-plantar aquagenic keratosis, Lane disease, acral peeling syndrome and others.

The absence of similar cases in the family and the occurrence of desquamative erythema even outside of water exposure led to the diagnosis of Fereol-Besnier disease or erythema scarlatiniforme desquamativum recidivans (ESDR). An emollient and an antibiotic therapy containing amoxicillin were prescribed with a good evolution.

**Conclusion:**

Fereol-Besnier disease is an underdiagnosed and poorly understood entity, particularly in the localized form. Recognition of this condition is important to prevent diagnostic and therapeutic escalation.

**Abstract N°: 1208**
**Revealing the influence of methotrexate and biologics on itch, pain, fatigue, and disease severity in pediatric patients with psoriasis: subjective and objective assessments**

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**Introduction & Objectives:**

Many pediatric patients with psoriasis suffer from itch, fatigue, pain and their severity of disease. The extent of this burden, and the effect of long-term treatment on these complaints has hardly been studied, especially when it comes to pain and fatigue. The Visual Analogue Scale (VAS; score 0-100) is a simple and non-invasive tool to provide insight in the individual perception of these subjective symptoms in children.

The objective was to investigate the influence of one year treatment with methotrexate (MTX) or biologics on the perceived amount of itch, fatigue, pain, and disease severity in pediatric psoriasis patients, and relatedness to the Psoriasis Area and Severity Index (PASI) and drug survival.

**Materials & Methods:**

Data were extracted from the ChildCAPTURE registry, a prospective daily practice cohort of patients with pediatric-onset psoriasis followed into young adulthood. The first treatment episodes of all patients <18 year on MTX or any biologic were used. The 'as treated' quarterly VAS scores during the first year of treatment were displayed as medians [interquartile ranges] and analysed using linear mixed models. Kaplan-Meier analyses were performed, split for overall, ineffectiveness related, and adverse event related first-year drug survival. First-year PASI scores were displayed using medians [interquartile ranges]. Additionally, the influence of previous methotrexate treatment (in biologic group) was investigated.

**Results:**

We included 166 patients with 142 methotrexate and 76 biologic treatment episodes. Median [IQR] VAS itch, pain, fatigue and severity were 64.0 [34], 16.0 [53], 22.0 [60], and 69.0 [23], respectively at start of MTX and 54.5 [52], 23.0 [64], 36.0 [62], and 73.0 [39] at start of biologic treatment. After 3 months, VAS severity (decline 35.4 in MTX, p-value <0.0001 and 42.6 in biologics, p-value <0.0001) and VAS itch (decline 39.9 in MTX, p-value <0.0001 and 35.4 in biologics, p-value <0.0001) showed the greatest significant declines. VAS pain and fatigue also improved significantly, except for fatigue in the MTX group. After three months, disease severity and fatigue continued to decrease in methotrexate treated patients, whereas fatigue significantly re-increased in biologic-treated patients. Previous treatment with methotrexate had no significant effect on the VAS courses in the biologic cohort, except for lower VAS fatigue in the no prior methotrexate group. First-year overall, ineffectiveness related, and adverse event related drug survival for methotrexate-treated patients was 72.0%, 90.0% and 85.8%, respectively, and 84.9%, 88.5% and 100% in biologic-treated patients. Median [IQR] PASI scores at month 12 were 2.4 [3.0] and 1.5 [2.4] for MTX and biologic treatments, respectively.

**Conclusion:**

Pediatric patients with psoriasis especially suffer from itch and the severity of their skin disease, and to a lesser extent from pain and fatigue. After 3 months, perceived itch, pain and disease severity were decreased significantly in both methotrexate and biologic treated patients. Our results demonstrated a similar subjective

experience of methotrexate and biologic treatment of pediatric psoriasis. However, objectively measured disease severity and drug survival were favourable in biologic treated patients.

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**Abstract N°: 1294****Subcutaneous ecchymotic nodules as an initial clinical manifestation of hemophilia**

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**Keywords:**

Doppler ultrasound, ecchymotic nodules, hemophilia, hematoma, violaceous nodules

**Case presentation:**

A 5-month-old male presented with a 3-day history of asymptomatic violaceous nodules on the anterior chest and back.

No previous trauma history or past medical history of remark was reported, including a normal pregnancy, labor, and delivery. However, mom recalls ecchymosis after immunizations on the site of the puncture.

On examination, a 3-cm violaceous nodule with associated ecchymosis was present on the anterior chest, and a similar 1- cm lesion was noted on the back. No other hematic or subcutaneous nodules were present.

The following were considered differential diagnoses: myofibromatosis, vascular malformations, neuroblastoma, and trauma, including child abuse.

A Doppler ultrasound was consistent with a blood-containing cyst on the anterior chest and an organized hematoma on the lesion of the back. No evidence of vascular malformation or tumors was present.

Laboratory exams revealed a normal complete blood cell count, including normal platelets and hematocrit values. However, prolonged activated partial thromboplastin time of 104 seconds and factor VIII <1% was noted on coagulation tests, confirming the diagnosis of hemophilia A.

On a 3-month follow, a new Doppler ultrasound was obtained, confirming post-hematic changes on residual lesions.

**Discussion:**

Hemophilia is a rare X-linked congenital bleeding disorder characterized by a deficiency of coagulation factor VIII -hemophilia A- or factor IX -hemophilia B- and manifestations are almost exclusively in males, while females are often asymptomatic carriers.

Patients with severe and moderate hemophilia often develop excessive bruising and various bleedings, spontaneously or after minimal trauma. Muscle bleeding and hemarthrosis are the most characteristic features of the disorder, although in patients with severe hemophilia, common types of bleeding include soft tissue and intramuscular bleeding.

Our patient presented with subcutaneous hemorrhagic nodules, an infrequent form of presentation of hemophilia.

Ljung et al. conducted a study to determine the most common presenting symptoms in patients with severe and moderate hemophilia. They found them to be subcutaneous bleedings in 48% of those severely affected. However, the specific form of presentation is not detailed.

The presentation of this clinical case aims to emphasize awareness of hemorrhagic subcutaneous nodules as a clinical manifestation of hemophilia and, in some instances, the only initial clinical sign.

Prompt identification of this disease will allow early initiation of treatment to prevent both immediate and future complications.

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**Abstract N°: 1406****Linear Morphea with Cutaneous Calcinosis**Issam Tablit<sup>1</sup>, Kacimi Asma<sup>1</sup>, Samira Zobiri<sup>1</sup><sup>1</sup>Mustapha University Hospital Center, Dermatology, Algiers, Algeria**Introduction & Objectives:**

Morphea or localized scleroderma is an inflammatory, sclerosing disease of the skin and occasionally underlying tissue. Although common in systemic sclerosis, calcinosis cutis has been rarely reported in patients with morphea. Herein, we describe a child with calcinosis cutis arising within linear morphea.

**Materials & Methods:**

A 5-year-old girl was referred for a three-year history of pigmented, indurated linear plaques over her right lower limb, as well as painful, hard nodules along the plaques that appeared a year ago.

Examination revealed red-brown, slightly depressed, linear bands following the Blaschko lines, from the thigh fold along the inner side of the lower limb to the internal malleolus. Along within the linear bands, we noticed multiple brown, hard, 1-3 cm, painful nodules.

X-ray and ultrasound showed discontinuous subcutaneous calcifications along the lower limb within the morphea plaques. A biopsy showed dermal horizontal broad collagen bundles and perivascular lymphocytic infiltrate with calcium deposits, consistent with morphea and calcinosis. Laboratory testing revealed no abnormalities in calcium metabolism and antinuclear antibody was negative.

She was treated with intravenous boluses of methylprednisolone (30 mg/kg/d) for 3 consecutive days each month for 3 months and 12,5 mg of methotrexate weekly. After three months, the nodules remained stable and she was referred for surgical removal while methotrexate was continued. She also was referred for physiotherapy sessions.

**Results:**

Morphea is an inflammatory, sclerosing skin disorder that can involve the underlying soft tissues. Calcinosis cutis refers to insoluble calcium salt deposition in the skin and subcutaneous tissue. It is frequently associated with various autoimmune connective tissue disorders, including systemic sclerosis.

Reports of calcinosis in the setting of morphea are rare. According to a review of the literature, 14 patients have been reported. The calcinosis cutis seemed to arise within morphea lesions of plaques, generalized and linear subtypes. The age at presentation with calcinosis ranged from 6 to 83. Diagnosis of calcinosis was made by biopsy, often supplemented by imaging with X-ray, ultrasound, and/or CT. Laboratory testing for calcium metabolism was normal. Over the 14 cases reported, five cases of calcinosis arose within the linear variant of morphea, the most common subtype among children and adolescents.

Our patient might be the youngest patient ever reported with a calcinosis cutis associated with a morphea, specifically the linear subtype of morphea.

Current management for coexistent morphea and calcinosis cutis are based on case reports and expert opinion. In the majority of cases, no improvement of calcinosis was noted. We chose to use methotrexate and systemic glucocorticoids according to the European guidelines. Surgical removal of nodules was scheduled after medical



treatment showed no improvement of calcinosis or related pain. Indeed, surgical excision has been the most successful treatment modality of calcinosis in the cases documented.

**Conclusion:**

Calcinosis cutis may occur in the setting of morphea and is believed to develop secondary to tissue damage. Various treatments have been attempted without success. Treatment should be individualized and to focus on stabilizing active morphea, with consideration to resultant calcinosis cutis whose surgical excision seems to be the most definitive treatment.

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**Abstract N°: 1440****A capricious hamartoma: Round And Velvety Epidermal Nevus (RAVEN)**Lula María Nieto Benito<sup>1</sup>, Ana Mateos Mayo<sup>1</sup><sup>1</sup>HLA Moncloa, Dermatology, Madrid, Spain**Introduction & Objectives:**

RAVEN, also, and most commonly, named nevoid acanthosis nigricans, was first described in 1971. The authors referred to this novel entity as an acquired epidermal abnormality that should not be confused with acanthosis nigricans. In this case, two lesions were present and located in the axillary and inguinal area.

**Materials & Methods:**

A healthy 13-year-old girl presented with a 1-year history of a pigmented lesion on her back. On physical examination, a large pigmented lesion in the midline lumbar area was observed. It was composed of two hyperkeratotic plaques, in which, polycyclic rounded structures with a heavily pigmented border were present (**Fig. 1A**). An “acanthosis nigricans-like” appearance was observed due to exaggerated liquenification (**Fig. 1B**). Skin biopsy showed epidermal hyperplasia with hyperkeratosis, papillomatosis, acanthosis and patched pigmentation of the basal layer, all compatible features with epidermal nevi (**Fig.2**). A diagnosis of Round and Velvety Epidermal Nevus (RAVEN) was made.

**Discussion:**

Due to its clinical and histopathological resemblance to acanthosis nigricans, in order not to confuse both entities, the descriptive term of RAVEN is preferred rather than nevoid acanthosis nigricans 3.

RAVEN is a very rare subtype of epidermal nevi. Its late age of onset is characteristic, in contrast with other epidermal hamartomas. Adolescence and the second decade are typical. Shape and texture are also differential features; rounded and/or polycyclic, well-defined structures are present, with a tendency towards peripheral hyperpigmented demarcation. The surface is velvety and papillomatous<sup>3,4</sup>.

Histopathological features are the characteristic from any kind of epidermal nevus<sup>5,6</sup>; differential diagnosis must be made with, properly, acanthosis nigricans, seborrheic keratosis, verruca vulgaris and confluent and reticulated papillomatosis of Gougerot and Carteaud<sup>5</sup>.

RAVEN is usually asymptomatic but can be responsible for aesthetic discomfort due to its considerable size, shape and pigmentation<sup>3</sup>. Various topical treatments have been tried with variable results: emollients, keratolytic principles, topical corticosteroids... Physical treatments based on tissue destruction such as cryotherapy, surgery or CO2 laser might be more effective<sup>2,3</sup>.

**Conclusion:**

RAVEN is a very rare subtype of epidermal nevi. Although being part of the group of hamartomas, it is not a congenital entity. Due to the clinical and histopathological resemblance, differential diagnosis must be made with, specially, acanthosis nigricans and seborrheic keratosis.

**Abstract N°: 1509****Pediatric Rosacea-Skin and ocular involvement**

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**Introduction & Objectives:**

Rosacea is classically a dermatosis of middle-aged adults with a clear phototype; it is considered rare in children. we report a serie of 8 pediatric rosacea.

**Case report:**

We retrospectively reviewed the medical records of 8 consecutive patients with pediatric rosacea younger than 15 years of age at the time of the diagnosis at Mohamed VI Hospital between December 2019 and mai 2023.

The mean age of diagnosis was 7.25 years. A female predominance ( 75 %) was noted. No patient had a family history of rosacea. The clinical examination showed skin involvement with papulo-pustules associated with telangiectasias in 7 patients (87.5%), ocular involvement with multiple chalazions or blepharitis in 4 patients (50%). three patients (37.5 %) had concomitant skin and eye involvement. one patient (12.5%) had an isolated ocular form.

All patients received local treatment regardless of the severity of the disease. Seven patients (87.5%) were treated with macrolides and only one patient (12.5%) with cyclins. The choice of treatment was mainly based on the age criteria, with cyclins in children over 8 years of age. For younger children, erythromycin or metronidazole was propose. However, some patients were treated with macrolide in front of an incomplete state of dentition, or if the treatment was initiated by ophthalmologists.

**Discussion:**

Rosacea is a chronic inflammatory condition that affects the skin and the eyes. The pathogenesis of rosacea is complex and includes the interaction between genetic and environmental factors, dysregulation of the innate immune system, neurovascular modifications and the interaction with skin commensals.

In paediatric rosacea, ocular symptoms are often predominant, and may precede skin symptoms in up to 50% of children, making it more difficult to diagnose. Patients may report ocular discomfort, photophobia and chronic red eye, while signs include meibomian gland dysfunction, meibomitis, blepharoconjunctivitis, episcleritis, recurrent chalazia, iritis, corneal vascularization, keratitis, corneal ulcer and scarring, lid margin telangiectasia, and conjunctival hyperaemia with or without inferior corneal vascularization.

There are no specific guidelines to manage rosacea in children, Topical therapy includ Metronidazole 0.75% in gel or cream applied twice daily for 1 month, then once daily until the eruption completely clears. Azelaic acid 15% gel, Permethrin 5% cream, Topical ivermectin 1% cream, and tacrolimus 0.03% or 0.1% ointment, or pimecrolimus 1% cream.

Oral agents are indicated in moderate to severe cases, in combination with topical therapy, include cyclins in children over 8 years of age. For younger children, erythromycin or metronidazole are suggested.

**Conclusion:**

Rosacea in children is a rare pathology. Ocular involvement should lead to suspicion of the disease. The different therapeutic protocols give excellent results in the short and long term, even better if diagnosed early.

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**Abstract N°: 1576****Multiple ossified pilomatrixomas in a nine year old girl without systemic impair**

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**Introduction & Objectives:**

The pilomatrixomas are the second most frequent benign tumour in children; multiple and ossified variants are very uncommon.

We present the case of a nine-year-old girl, with an unremarkable clinical history, who consulted for asymptomatic subcutaneous lesions with a progressive growth over a period of between 2 years and 6 months. We observed four well-defined subcutaneous nodules adhered to deep planes in left posterior shoulder (2.5 x 1.5 cm), temple (0.8 cm) and mandibular branch (1.2 cm), and in right mandibular angle (2.5 cm). The histopathological study showed a well-defined tumour with foci of calcification and ossification immersed in a fibrous stroma.

**Discussion:**

The pilomatrixoma is a benign tumour derived from the matrix cells of hair follicle of which its exact pathophysiology is unknown. It appears in children under 20 years as an asymptomatic small single subcutaneous firm nodule located in the head and neck, with a progressive growth.

Histologically it is usually composed of central nests of eosinophilic shadow cells surrounded by nests of matrix basaloid cells, all immersed in a fibrillar stroma. In late regression stages of pilomatrixoma, foci of bone metaplasia can be seen as a reaction to foreign body after its rupture. However, we show one of the ten cases of extensive ossification described in the literature, comparing their similarities and differences.

Multiple pilomatrixomas have a low incidence. They may be sporadic, familiar or associated with syndromes (Steinert myotonic dystrophy, familial adenomatous polyposis, Turner syndrome, Rubinstein-Taybi syndrome). Based on current evidence, we explain the role of multiple pilomatrixomas as an early marker of these syndromes, taking into account the family history of pilomatrixomas, and the personal and family history of these syndromes.

**Conclusion:**

We present a rare variant of pilomatrixoma (multiple, extensive ossification, large size) in a child with unremarkable personal or familial history and we describe a diagnostic algorithm for multiple pilomatrixomas, emphasising their role as an early syndromic marker.

**Abstract N°: 1674****Juvenile keratosis lichenoides chronica with hyper IGE: A case report.**

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**Introduction:**

Keratosis lichenoides chronica (KLC) is a rare lichenoid dermatosis of adulthood, characterized by keratotic parallel linear papules, retiform plaques. Has a chronic and usually progressive course. Pediatric onset KLC is rarely reported in the literature.

**Case report:**

A 5 years old boy presented with eruption over face and limbs since age of 1 month. Physical examination showed facial erythematous and hyperpigmented macules. Scaly hyperpigmented ears and lips. Keratotic papules in parallel linear and reticular arrangement, follicular papules with keratotic plug over limbs and retiform plaques over joints. Perifollicular Scales entangling hair shafts in a tubular manner and Hair casts. Hyperkeratotic lesions on palms and soles. Hoarseness voice. The remainder of the physical examination is normal. Laboratory test (WBC: 12290, PPEP: inflammatory syndrome, vit A: 196 mg/l, Flow cytometry: mild elevation of T CD3 and inversion of CD4/CD8, mild elevation of IgG, hyper-IgE. Histopathological analysis revealed hyperkeratosis, hyper granulosis, pigmentary incontinence and band-like lymphocytic infiltrate. laryngoscopy found rough vocal cords covered by keratotic nodules. His 3 years old sister has identical clinical and biological findings. The patients were treated by acitretin with moderate improvement after two months of follow up.

**Discussion:**

The pediatric cases are rarely reported, four of the reported cases were congenital and 10 cases had a family history suggesting an autosomal recessive mode of inheritance. In pediatric patients, facial lesions are the first to appear, presenting as erythematous purpuric macules. Clinical examination usually reveals lesions at different progression stages, ranging from isolated keratotic, often follicular papules with a keratotic plug to classic dark erythematous, keratotic papules in parallel linear arrangement, simulating Koebner phenomenon and retiform plaques. In 1938, Nekam described hyperkeratotic palmar papules. In 1965, Margolis et al described a patient with a retiform pattern of lichenoid papules and a hoarse voice. In 1980, Nabai & Mehregan reported one patient with Laboratory evidence of immunodeficiency. Hair involvement is present in 22% of pediatric cases. Oral involvement was noted in 23% of cases. Histopathologic features of 87% of cutaneous biopsies show a lichenoid tissue reaction pattern with hyperkeratosis and predominant acanthosis. Additionally, there is a bandlike dense mononuclear-cell infiltrate in the upper dermis, pigmentary incontinence. Treatment has been disappointing in most cases; however, in the majority of reports, UVA and UVB light phototherapies and oral retinoids have been reported as the most effective treatment modalities.

**Conclusion:**

Pediatric KLC is a rare disorder, it tends to have an autosomal recessive inheritance. Clinical recognition is often difficult, since the early skin manifestations unspecific. It has a chronic course and resistant to therapy.

**Abstract N°: 1928****Nodular lesions on giant congenital melanocytic nevus: the role of molecular analysis with SNP-array.**

Chiara Bordin<sup>\*1</sup>, Martina Tolone<sup>2</sup>, Andrea Diociaiuti<sup>3</sup>, Mario Zama<sup>4</sup>, Sabrina Rossi<sup>5</sup>, May El Hachem<sup>3</sup>

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**Introduction & Objectives:**

We present the case of a 4-year-old patient affected by giant congenital melanocytic nevus (GCMN) of the back, extending to the head and left mandibular region with numerous satellite lesions of various sizes on the limbs, abdomen and face. MRI of the brain and spinal cord performed at one month of age was negative for neuromelanosis and the ophthalmological examination was normal.

During her first years of life, she developed two nodular lesions arising from the GCMN: at the age of two months, a polypoid ulcerated nodule appeared in the nuchal region and at the age of 3 years the patient developed a new greyish nodular lesion on the dorsal interscapular region.

**Materials & Methods:**

The patient underwent regular clinical and dermoscopic examination and every nodular lesion was biopsied. Both nodules were analysed through histopathological, histochemical and molecular examination (SNP-array).

**Results:**

For the differential diagnosis between proliferative nodule and malignant lesion, histological examination and immunohistochemistry were performed. In both cases they were suggestive, but not decisive for melanoma. Therefore, molecular diagnosis with SNP-array was performed.

In the first lesion monosomies of various chromosomes and trisomy of chromosome 22 were highlighted, while no segmental chromosomal alterations were detected. The diagnosis was a proliferative nodule. Although the lesion was classified as benign, in consideration of the morphological aspects, a strict clinical and ultrasound follow-up was planned.

Regarding the second lesion, SNP-array showed multiple chromosomal imbalances, both aneuploidies and segmental alterations, in particular in the loci 8q.24 (MYC gain) and 1q (gain), which are relevant loci for the assessment of the risk of biological aggressiveness of melanocytic lesions. The result supported the diagnosis of melanoma arising in GCMN.

Staging with PET-CT was performed, showing multiple lymphadenopathies at laterocervical site bilaterally, axillae with right prevalence, root of the mesentery, left sacral wing and left ischiopubic ramus. The girl was then started on systemic therapy, in particular with Nivolumab 3 mg/kg.

**Conclusion:**

Melanoma a very rare in childhood, however, GCMN should undergo straight follow-up. Nodules should always be biopsied and in case histopathology and immunohistochemistry are not decisive, molecular analysis should be



performed.

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**Abstract N°: 1947****False alarm: A case of Hansen's disease misdiagnosed as multiorgan metastases in a 10-year-old boy with T-cell lymphoblastic lymphoma**

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**Background:** Hansen's disease is a chronic granulomatous disease caused by *Mycobacterium leprae* and *Mycobacterium lepromatosis* that primarily involves the skin and nerves. We report a case of Hansen's disease misdiagnosed as multiorgan metastases in a pediatric patient with T-cell lymphoblastic lymphoma.

**Observations:** A 10-year-old boy gradually developed multiple erythematous to dusky red papules and plaques on the upper back, buttocks and right dorsal wrist while undergoing chemotherapy. Lesion on the latter is hypoesthetic, occasionally painful and tender. While on remission from his T-cell lymphoblastic lymphoma, he was brought to the emergency room due to acute onset of focal seizures, severe pain on the back, right shoulder, and right wrist with associated papal hand deformity. Brain, bone, and cutaneous metastases were highly considered. Further investigations revealed nodular granulomatous dermatitis on skin biopsy and numerous acid-fast bacilli on slit-skin smear consistent with leprosy. To rule out relapse and metastasis of lymphoma, PET scan and CSF cytology were done which showed unremarkable results. Hence, the case was diagnosed with Hansen's disease, multibacillary, lepromatous (MB-LL) with type 1 lepra reaction. Patient was started on a multidrug therapy for one year composed of rifampicin 450mg once monthly, clarithromycin 7.5mg/kg once monthly, clofazimine 150mg once monthly and 50mg every other day, and prednisone 0.75mg/kg/day until complete suspension of the lepra reaction. After 12 months of treatment, there was complete resolution of the lesions with no recurrence of neuritis and seizures. There was marked improvement in skin biopsy substantiated by a negative slit-skin smear.

**Key message:** We presented this case because leprosy can be mistaken for other dire conditions such as metastasis as in this case. Clinicians should be able to recognize that patients with hematologic neoplasms can suffer from infections such as leprosy as a complication of the underlying malignancy or by the treatments being done.



**Abstract N°: 1953****Skin vascular hallmarks clue for KRIT1- hereditary cerebral cavernous malformations**

Martina Tolone<sup>\*1</sup>, Chiara Bordin<sup>2</sup>, Andrea Diociaiuti<sup>3</sup>, Cataldo Patruno<sup>1</sup>, Steven Paul Nisticò<sup>1</sup>, May El Hachem<sup>3</sup>

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**Introduction & Objectives:**

A 4-year-old patient came to our attention for a congenital vascular lesion on his left arm, asymptomatic and stable. The patient was in good general condition. Physical examination showed a crimson-coloured macula, irregularly shaped, measuring 4cmx 2cm and several tortuous vessels radiating outward from the superficial lesion. The overlying epidermis was hyperkeratotic. This clinical presentation was consistent with the diagnosis of hyperkeratotic cutaneous capillary-venous malformation (HCCVM). Similar skin lesions were observed on the right calf of his younger sister and on the forearm of his father. Familial history of cerebral hemorrhage was referred (paternal grandfather and a first cousin).

**Materials & Methods:**

Family segregation tests were performed.

**Results:**

The pathogenic variant c.8825 of p. (Met275Ilefs\*13) in the *CCM1/KRIT1* gene was observed in our patient, in his sister, in his father and in his cousin. Screening with Nuclear Magnetic Resonance of the brain and spinal cord was planned and a subsequent neurosurgical follow-up was scheduled.

**Conclusion:**

Heterozygous pathogenic variants in the *CCM1/KRIT1* gene is associated with the familial-type of the cerebral cavernous malformations (FCCMs). Cerebral cavernous malformations (CCMs) are characterized by abnormally dilated capillaries packed in sinusoids, without involvement of brain parenchyma, mainly located within the central nervous system.

Mutations in *KRIT1* account for 53–65% of familial cases and more than 100 different mutations have been identified so far, characterized by autosomal dominant inheritance with incomplete penetrance. Among the mutation carriers, symptomatic patients constituted 66%. A variable phenotypic expression is observed: headaches, seizures, focal neurological deficits and life-threatening intracranial bleeding. Affected patients can exhibit extra-neurological manifestations. HCCVMs are rare but peculiar cutaneous vascular malformations associated with CCMs; therefore, when detected, cerebral vascular anomalies should be always investigated.

**Abstract N°: 2154****When faced to a congenital mass protruding from vagina, think of botryoid rhabdomyosarcoma.**

Bochra Bennour<sup>\*1</sup>, Sophia Abdelilah<sup>1</sup>, Maryem Aboudourib<sup>1</sup>, Said Amal<sup>1</sup>, Hocar Ouafa<sup>1</sup>

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When faced to a congenital mass protruding from vagina, think of botryoid rhabdomyosarcoma.

**Introduction & Objectives:**

Gynaecologic neoplasms are rare in children. Rhabdomyosarcoma (RMS) are malignant mesenchymal tumors deriving from myogenic progenitor that represents one of the most common soft tissue sarcomas of childhood. It is often found in the genital tract of infants and young children and may be exceptionally congenital.

We report a rare case of a congenital botryoid rhabdomyosarcoma.

**Case report:**

We report a case of a female newborn who presented 3 congenital papillomatous tumors in the vulvar area with vaginal and anal extension and an herpetic superinfection.

The histological study revealed spindle cells proliferation with high cellularity, the epidermis was hyperplastic and pseudo-sarcomatous. Immunohistochemistry showed a moderate expression of desmin, myogenin and Ki67 markers. Protein S100 was negative.

The diagnosis of congenital botryoid rhabdomyosarcoma was retained.

The therapeutic management was to put the patient under chemotherapy according to the VAC protocol and to perform a tumor resection after regression of the lesions in order to avoid the anatomical damage of the ano-vulvar region.

**Discussion**

RMS are extremely rare in neonates. In a large multicenter study of 3217 patients with rhabdomyosarcoma, only 14 cases were congenital (0,4%). No cases were found in the Anglo-Saxon literature in which vulvar location and congenital onset were associated.

Four major histologic subtypes of RMS are identified: embryonal, alveolar, pleomorphic, and sclerosing/spindle cell.

Embryonal RMS (ERMS) is the most common type of all RMSs frequently arising in the pediatric female genitourinary tract. A study on 67 female children with ERMS showed that uterine RMSs were most often seen during adolescence however, vagina was the primary site in 68.4% of children between 0 and 9 years.

Botryoid variant is a type of ERMS often occurring frequently in the vagina, characterized by 'grape-like' appearance caused by polypoid mass arising in submucosal tissue.

Histological findings helps eliminate sacrococcygeal teratoma neuroblastoma and Burkitt's lymphoma.

Recently, authors have highlighted a variety of genitourinary ERMS associated with a somatic or a germline

DICER1 mutation.

The combination of surgery and chemotherapy (Vincristine, dactinomycin, and cyclophosphamide) with or without radiotherapy or brachytherapy provides good results in early stage of genital ERMS.

**Conclusion:**

In conclusion, we presented a rare case of congenital botryoid RMS presenting as congenital tumor protruding from vagina. Clinicians should be aware of its presentation to assure an optimal management.

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**Abstract N°: 2467****Biologics for psoriasis patients under 18 years of age: Real-world evidence from the Chinese Psoriasis Real World Evidence Research Group**Yuxin Zheng<sup>\*1</sup>, Xiao-Yong Man<sup>1</sup>, XI-Bei Chen<sup>1</sup><sup>1</sup>Zhejiang University School of Medicine Second Affiliated Hospital, Dermatology, Hangzhou, China**Biologics for psoriasis patients under 18 years of age: Real-world evidence from the Chinese Psoriasis Real World Evidence Research Group****Introduction & Objectives:**

Treatment for pediatric psoriasis is challenging because of the lack of real-world evidence, especially for biologics. This study evaluated the efficacy and safety of biologics in children with psoriasis based on real-world evidence.

**Materials & Methods:**

Pediatric psoriasis patients aged <18 years who were treated with biologics in our hospital (2020–2022) were prospectively analyzed. Patients treated with adalimumab, secukinumab, or ixekizumab were followed up for at least 16 weeks, and 22 of 38 patients completed the 52-week observation period. Dermatologist raters were blinded to ensure the reliability of the PASI, BSA, and PGA score assessments. PASI 75 or PGA 0/1 at week 12 represented an efficient indicator.

**Results:**

Thirty-eight patients (20 males and 18 females; median age, 12.6±4.1 years) were enrolled, and none were lost to follow-up. All participants were diagnosed with psoriasis, including plaque psoriasis (n=36), nail psoriasis (n=1), and pustular psoriasis (n=1). Within 12 weeks, all patients achieved scores above PASI 75 and PGA 0/1. The average time to reach PASI 75 was 4.3±2.0, 3.2±1.8, and 2.4±0.4 weeks in patients using adalimumab, secukinumab, and ixekizumab, respectively, and, 27.2% (3/11), 86.4% (19/22), and 75.0% (3/4) of these patients achieved PASI 100 at week 12, respectively. Moreover, 18 of 20 patients with plaque psoriasis maintained ≥PASI 75 after 52 weeks. A total of 7 patients (six plaque type and one nail type) underwent biologics transitions without any washout periods, and the use of a novel biological agent led to excellent outcomes. Among them, 6 patients with plaque psoriasis obtained PASI 75 or above within 12 weeks after switching to a new biological agent. Considering extending the dose interval, 87.5% adalimumab patients, 100% secukinumab patients, and 33.3% ixekizumab patients could successfully prolong their dosing interval maintaining PASI 90 or above and the absolute PASI score < 3 for at least 6 months. The most commonly reported adverse effect was upper respiratory tract infection, and no severe adverse effects were reported.

**Conclusion:**

Our real-world data demonstrated the safety and effectiveness of adalimumab, secukinumab, and ixekizumab in children with psoriasis.

**Abstract N°: 2592****Sleep in infants with moderate-severe atopic dermatitis (The SPINDLE study)**

Cathal O'connor<sup>1</sup>, Soraia Ventura<sup>1</sup>, Mark O'sullivan<sup>1</sup>, Alan Irvine<sup>2</sup>, Deirdre Murray<sup>1</sup>, Jonathan Hourihane<sup>3</sup>, Michelle Murphy<sup>4</sup>, Geraldine Boylan<sup>1</sup>

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**Introduction & Objectives:**

Atopic dermatitis (AD) is associated with sleep disruption due to pruritus, inflammation, increased transepidermal water loss (TEWL) and circadian alterations. Little is known about the impact of AD on sleep in early infancy, a critical time for neurodevelopment.

The aim of this study was to deeply characterise the sleep architecture of 6-month-old infants with moderate-severe AD, compared to controls.

**Materials & Methods:**

Overnight movements (defined as 10-15 seconds of movement) were measured for five consecutive nights using a novel wearable movement sensor. Infantile and parental sleep measures were reported by parents. Daytime sleep quality and quantity was measured by electroencephalogram (EEG) polysomnography. AD was assessed using clinical severity scoring, skin barrier assessment (TEWL, natural moisturising factor levels, and filaggrin mutational analysis) and inflammatory cytokine analysis by tape stripping.

**Results:**

57 controls and 33 cases were recruited. Average EASI at time of 6-month assessment was 7.2 (range 0.8-34.8). Infants with AD recorded almost twice as many overnight movements as controls (19.2 versus 11.2,  $p < 0.05$ ). Infants with AD slept for 13.1 hours versus 13.8 ( $p < 0.05$ ), and woke 3.5 times versus 1.9 ( $p < 0.05$ ). Infants with AD had differences in EEG sleep measures, including sleep spindle power and frequency. Mothers of cases had worse total sleep time (6.3 versus 7.1 hours,  $p < 0.05$ ), sleep-onset latency (27.1 versus 12.8 minutes,  $p < 0.05$ ), and sleep efficiency.

**Conclusion:**

This is the most detailed study ever performed on sleep in infants with AD using objective sleep measures, and has revealed significant effects on sleep architecture, sleep duration, and overnight movement.

**Abstract N°: 2781****Depression and anxiety among pediatric patients with psoriasis and alopecia areata: A case-control study**Damla Demir<sup>1</sup>, Mihriban Keleş<sup>2</sup>, Ezgi Aktaş<sup>\*3</sup>

<sup>1</sup>Umraniye Training and Research Hospital, Dermatology and Venereology, Türkiye, <sup>2</sup>Umraniye Training and Research Hospital, Psychiatry, Türkiye, <sup>3</sup>Prof Dr Cemil Taşcıoğlu City Hospital, Dermatology and Venereology, Türkiye

**Introduction & Objectives:**

Alopecia areata (AA) and psoriasis are chronic and recurrent skin diseases. In addition to signs and symptoms related to cutaneous lesions, both may cause significant impairment in quality of life and psychological symptoms. However, although both diseases have high prevalences in childhood, very limited data are available investigating the relationship between psychiatric impacts of these diseases in children. The aim of this study was to evaluate depression and anxiety symptoms in children with AA or psoriasis.

**Materials & Methods:**

A total of 89 subjects including 30 patients with AA, 33 patients with psoriasis and 26 healthy controls were enrolled in this prospective, observational case-control study. Child Depression Inventory (CDI) and Screen for Child Anxiety and Related Disorders (SCARED) were applied to both patients and control groups.

**Results:**

In terms of gender, age, maternal age, paternal age, socioeconomic status, education levels of the family, disease duration no significant difference was found between the patients with AA and psoriasis and controls ( $p > 0.05$ , for all). The CDI score was lower in control group than in patients with AA ( $p < 0.001$ ) and also in patients with psoriasis ( $p < 0.001$ ). No significant difference was observed between AA and psoriasis patients in CDI scores ( $p = 0.162$ ). Higher scores of SCARED was measured in AA patients than in control group ( $p < 0.001$ ). SCARED scores were also higher in patient with psoriasis than in controls ( $p = 0.007$ ). No significant difference was observed between AA and psoriasis patients in terms of SCARED scores ( $p = 0.109$ ).

**Conclusion:**

Anxiety and depression symptoms were more prevalent in patients with AA and psoriasis than in healthy subjects, while no difference was found between two diseases. Our results emphasize the importance of psychiatric evaluation in pediatric patients with AA and psoriasis. Considering that childhood experiences may be associated with lifetime psychosocial morbidity, a combined approach with medical therapy and psychiatric counselling may be beneficial in this patient group.



**Abstract N°: 2812****To assess the therapeutic outcome and safety of oral propranolol and topical timolol in the management of infantile hemangioma**Leena Balakrishnan Sugumaran<sup>\*1</sup><sup>1</sup>Apollo Children's Hospital, Chennai, India

**Introduction & Objectives:** Infantile hemangiomas (IHs) are proliferative, benign tumors of vascular endothelium that may be present at birth or more commonly become apparent in the first 2 weeks of life. They are the most common soft tissue tumors of infancy occurring in 3-10% of infants. There are only few studies done all over the world pertaining to find out the therapeutic outcome and safety in using oral propranolol and topical timolol in the treatment of infantile hemangioma. Our study aims at finding out the therapeutic outcome and the safety of oral propranolol and topical timolol in the management of infantile hemangiomas in children coming to a tertiary care hospital in Chennai.\*\*

**Materials & Methods:** We studied\*\* the therapeutic outcome, efficacy and safety of oral propranolol and topical timolol in the treatment of infantile hemangiomas in 50 infants enrolled in our research. It was a prospective observational study.

**Results:** Oral propranolol when used in the treatment of IH at a dose of 2 mg/kg/day for 6 months showed a very good response in 76%, partial response in 20% and no response in 4%. Topical timolol gel (0.5%) application for 6 months in the treatment of IH showed a very good response in 44%, partial response in 32% and no response in 24%.

**Conclusion:** Even though both the drugs had a good safety profile, the parent satisfaction while on oral propranolol use was considerably higher (68%) and the complications (while on treatment with oral propranolol) were seen only in very few cases.



**Abstract N°: 2818**
**Non-Langerhans Cell Histiocytosis in different localizations in pediatric patients: a case series of 4 patients**

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**Non-Langerhans Cell Histiocytosis in different localizations in pediatric patients: a case series of 4 patients**
**Introduction & Objectives:**

The non-Langerhans Cell Histiocytosis (non-LCH) are a rare group of disorders defined by the accumulation of histiocytes that do not meet the phenotypic criteria for the diagnosis of Langerhans cells. Juvenile Xanthogranuloma and Benign Cephalic Histiocytosis are benign, proliferative disorders belonging to the broad group of non-LCH and are typically disorders of early childhood. JXG presents as a solitary, reddish or yellowish skin papule/nodule, most often on the head, neck, upper trunk. BCH presents with small, yellow-red or yellow-brown, asymptomatic macules and/or papules located mostly on the head and neck. We presented one case of BCH and three cases of JXG.

**Materials & Methods:**

Case-1: A 10-month-old boy presented with yellow-red papules on face and trunk. Lesions were present for 2 months. A biopsy was performed, and it showed well-circumscribed, histiocytic infiltrate in the superficial and reticular dermis. Immunohistochemical staining of lesional cells demonstrates positive staining for CD163, CD68, and S100 in a small number of cells but negative staining for factor XIIIa CD1a, CD34.

Case-2: A 4-year-old girl presented with a yellow-red papule on her back and lesions were present for 1 year. A biopsy was performed, and it showed mixed dermal infiltrate of mononuclear cells, multinucleated giant cells. CD 68 (+) histiocytes and Touton giant cells were observed. Immunohistochemical staining of lesional cells demonstrates positive staining for S100 in a small number of cells but negative staining for CD1a.

Case-3: A 5-years-old boy presented with a plaque on his scalp. The plaque was 15 millimeters in diameter, tan-orange in color, with central scaling and localized on the vertex of the scalp. The histological evaluation revealed diffuse infiltrates of mononucleated cells, some of which are characterized by their reniform nuclei, foamy histiocytes and numerous Touton giant cells, admixed with eosinophils. Immunohistochemistry yielded results immunoreactive for CD68, weak focal positivity for S-100 protein, but negative for Langerin.

Case-4: A 1-year-old girl presented with a yellow-brown papule on her chin. The lesion was presented for 3 months. Diagnosis made clinically, based on the typical appearance of lesions on physical examination.

**Results:**

For Case-1, the results led to the diagnosis of BCH. For Case-2,3,4, the results led to the diagnosis of JXG. In 4 patients, there was no ophthalmic and hematologic involvement. Systemic examinations were normal. They were followed without treatment.

**Conclusion:**

JXG ve BCH are self limited, benign disorders. In both diseases, skin lesions usually resolves spontaneously. In

clinical practice, when the lesions are in different localizations than they are in usual and when they are multiple, it can be confusing in terms of differential diagnosis in benign cases such as JXG and BCH. Ocular and systemic involvement should be evaluated in JKG cases with multiple skin lesions. Solitary lesions must be differentiated from spitz nevus in children for both diseases. This case series is presented to draw attention to these clinical aspects of benign subtypes of non-LCH such as JXG and BCH.

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**Abstract N°: 2858****Bullous varicella in an immunocompetent infant**

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**Introduction & Objectives:**

Varicella, also known as chickenpox, is a highly contagious disease caused by the varicella zoster virus (VZV). It primarily affects children and is mostly benign. However, it continues to cause significant morbidity and even mortality. The bullous onset of varicella is rare and mostly seen in immunocompromised children. Herein, we report a case of bullous varicella in an immunocompetent infant.

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

An 8-month-old girl with no prior medical history or previous drug use was admitted to the pediatric department with a history of fever and the subsequent eruption of multiple bullous lesions on the entire body. The lesions started as vesicles on the forehead and spread to involve the entire body. Her brother had a history of a concomitant varicella infection, which was successfully treated. Physical examination revealed multiple vesicles and large erosions, especially on the trunk, limbs, and face. Nikolsky's sign was negative. A laboratory investigation revealed an increased inflammatory marker. Swab cultures taken from the ruptured bullous were negative. HIV serology was negative, and the immunoglobulin levels were normal. The diagnosis of bullous varicella was made based on family history and clinical findings. The patient received intravenous acyclovir for 10 days with no improvement.

**Conclusion:**

Varicella is one of the most common viral infections in children. It may lead to some complications, such as bacterial infections, pneumonia, meningitis, encephalitis, cerebellar ataxia, and pain syndromes. The most common cutaneous complication of varicella is bacterial surinfection, which is commonly caused by *Staphylococcus aureus* or *Streptococcus pyogenes* and may worsen scarring and, in rare cases, lead to staphylococcal and streptococcal toxic shock syndromes. Bullous varicella is an extremely rare clinical manifestation of the disease. The exact mechanisms by which large bullae are formed are not clear; some synergistic effect between the infectious agents must occur. It has been suggested that bullous lesions are caused by secondary bacterial infections, most commonly *Staphylococcus aureus*. Vaccination coverage and adequate prophylaxis contribute to avoiding complications in the high-risk population. Early management of patients presenting with chickenpox is crucial. Effective treatment with acyclovir, combined with hygiene precautions, has proven effective in reducing virus transmission and preventing the emergence of postexposure cases.

**Abstract N°: 2871****Pediatric recurrent herpetic whitlow**

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<sup>1</sup>UCH Ibn Rochd , Department of Dermatology and Venereology, , casablanca

**Introduction:**

A whitlow is a finger infection that is usually caused by bacteria. However, whitlow caused by the Herpes simplex virus (HSV) is uncommon, and its occurrence in children is exceptional. We're reporting a new case of a child girl with recurrent herpetic whitlow of the annular right.

**Case report:**

A 6-year-old immunocompetent girl with no medical history was brought to the dermatology emergency department by her mother for a painful erosive plaque on her right ring finger. Clinical examination revealed a painful, erythematous, polycyclic erosive plaque, impetiginated in some areas, associated with paronychia, without any other associated cutaneous or extracutaneous signs. The mother consulted several pediatricians, and the child was diagnosed with bacterial whitlow and received oral amoxicillin-clavulanate without improvement. After thorough questioning, there was no personal history of herpetic labial infection in the child or the parents. However, the mother reported that the grandfather had recurrent herpetic labial infections, and two previous episodes of cutaneous infection on the girl's right ring finger, which improved with local fusidic acid. Bacteriological sampling was sterile, and Tzanck cytodiagnosis showed ballooning cells. The diagnosis of recurrent herpetic whitlow was made. The patient received oral and local acyclovir, along with local care, and had a good recovery within a few days. She was advised to consider medical prophylaxis for the possibility of a recurrence.

**Discussion:**

Herpetic whitlow is an infection caused by the herpes simplex virus type 1 or 2, which rarely affects children. HSV transmission involves direct contact with active lesions on the skin or mucous membranes, or reactivation of the latent virus, often after a first oral herpes infection. It can be misdiagnosed as bacterial whitlow by various specialists, leading to the initiation of antibiotic therapy, sometimes combined with surgical debridement. On the other hand, it should be considered in the presence of a personal or familial history of herpetic labial infection. Confirmation requires the Tzanck test. PCR is only recommended for severe herpetic infections in immunocompromised patients. Treatment is based on acyclovir therapy without a surgical incision.

Haut du formulaire

Haut du formulaire

**Abstract N°: 2956****Rosai -Dorfman disease -atypical presentation with raised eosinophils , anemia and hepatomegaly**Roja Raya<sup>1</sup>, Vijay Zavar<sup>1</sup>, Vivek Nikam<sup>1</sup><sup>1</sup>dr . vasanth pawar medical college, dermatology, Nashik, India**\*\* Rosai - Dorfman disease – Atypical presentation with raised eosinophils , anemia and hepatomegaly****ROJARAYA (1),VIJAY ZAWAR(1), VIVEK NIKAM(1)**

Dr. Vasantrao Pawar Medical College, Hospital &amp; Research Center, Dermatology, Nashik, India (1)

**Introduction:** \*\* Rosai - Dorfman disease (RDD) is a benign, self-limiting, and progressive lymphoproliferative disorder of unknown etiology. RDD is non langerhans cell histiocytic disorder clinically present as painless lymphadenopathy. It is histologically characterized by proliferation and accumulation of atypical histiocytes along with plasma cells, neutrophils in lymph nodes and extranodal sites like skin, respiratory tract and periorbital areas. Emperipolesis is a histological marker, but not exclusive to RDD.

**Objective:** To study the atypical clinical, and histological findings in a suspected of Rosai- Dorfman disease.

**(Method and methodology) of Case report:** A 3 year old female child brought by her mother with complaints of erythema and peeling of skin over face and multiple pruritic nodular lesions all over body since 2 1/2 years. Birth history is uneventful and no similar complaints in family. General examination revealed bilateral inguinal, axillary lymphadenopathy and hepatomegaly. Peripheral smear showed anemia with increased eosinophils. FNAC of lymph node showed reactive and lymphadenitis and bone marrow aspiration consists of increased eosinophil precursors and histiocytes. Histopathological findings from a nodule showed lymphohistiocytic proliferation, plasma cells and abundant eosinophils. No lytic lesions were observed on X-ray of long bones and skull . Antinuclear antibody (ANA) is negative. Immunohistochemistry revealed CD68 positive (most cells) and S100 protein (scattered cells) and was negative for anti-CD1a. Patient was given only antibiotics and supportive therapy in the initial presentation. Patient showed good clinical response after starting steroids and low dose methotrexate.

**Conclusion:** Rosai- Dorfman disease or sinus histiocytosis is a rare histiocytic disorder. It often mimics other infectious diseases and malignancies and easily be misdiagnosed. Systemic approach is needed while establishing the diagnosis of RDD. In our case ,patient presented with clinical features of erythema, raised eosinophil count, anaemia and hepatomegaly ,which are not classical features seen in RDD. After systematic investigation , we approached to the diagnosis of RDD after ruling out other differential diagnosis. Hence we are reporting this case for unique clinical and histological findings.



**Abstract N°: 2996**

**Clinico Epidemiological Profile Of Paediatric Dermatoses In A Tertiary Care Hospital In Eastern India**

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**Introduction & Objectives:** Within the paediatric population, different skin manifestations occur with varying frequency and are also affected by the seasonal as well as the familial, cultural and socio-economic environment the child finds itself in. Epidemiological studies of Paediatric population are still not adequate in number in our country, specifically in Eastern India. This study, therefore, is a 'baby step' on a 'road less travelled'.

We intend to find out correlation between-

- Gender with dermatological disease pattern
- Socioeconomic status with dermatological disease pattern
- Variability of seasons with dermatological disease pattern
- Dermatological disease pattern amongst urban and rural populations
- Physiological and Pathological afflictions amongst different age groups
- Different infectious agents affecting different age groups

**Materials & Methods:** The study is a cross-sectional study that was done at the Dermatology department at Calcutta National Medical College and Hospital, Kolkata for a time duration of 12 months from 01/03/2021 to 28/02/2022. Sample size was 4692 patients. SPSS software was used for analysis.

**Results:** 2511 males and 2181 females were enrolled across Pre term Neonate (1.5%), Term Neonate (12.3%), Infant (17%) and Juvenile (69.2%) age groups across all categories of Kuppaswamy Scale with Lower Class being most affected with 34.7% patients. Pathological afflictions were 77.5%. Non infectious conditions were in majority (54.7%). Most diseases were found in the summer (40.1%). Urban population was most affected in totality (53.8%).

We also found that quite a few of the people were living in unhygienic conditions and sharing fomites (25.7%) and some were not even using soaps during bathing (21.6%). This leads to further infections and infestations common in this group.

Among the infectious agents, fungi as a group seemed to pre dominate at 13.5% amongst all dermatoses observed.

**Conclusion:** This study therefore-

- Adds to the limited knowledge of the same and modify as well as be compared to the national clinico-epidemiological profile of paediatric dermatoses.
- Helps prioritise the funds and types of medicines to be made available in the government hospitals of the region for treatment of the patients.

- Identifies emerging trends and endemicity of diseases.
- Identifies modifiable risk factors for certain diseases.
- Helps us get a clear idea of the kinds of paediatric dermatoses prevalent in the region.

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**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 3250****Cutaneous mastocytosis in children from North-East Hungary**

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**Introduction & Objectives:**

Mastocytosis is a rare ( 1/10000 prevalence) disease, characterized by disfunction of mastocytes. The disease has two major forms: cutaneous or systemic form. Cutaneous mastocytosis based on its clinical appearance is grouped as mastocytoma, maculopapulosus mastocytosis, diffuse cutaneous mastocytosis. The authors review cases of children with mastocytosis between 01. July 2004 and 31-30.Apr. 2023).

**Materials & Methods:** The authors reviewed database of the Hospital of Borsod-Abaúj Zemplén County, using its Medworks software system.

**Results:** 79 children were diagnosed with cutaneous mastocytosis. The age of onset was under 2 years, with male dominance (53%). The types of mastocytosis were: 5 diffuse cutaneous mastocytosis, 23 maculopapular form, 51 solitary mastocytoma. Apart from mastocytoma, 66% of patients with solitary mastocytomas had atopic dermatitis or bronchial inflammation. The first-line treatment in CM is antihistamine therapy (mainly H1 and H2 antihistamines) and short-term topical corticosteroids. Phototherapy is the second-line therapy which may be considered when antihistamines do not produce the expected improvement, however, it has age limitations in most of the cases. Although the disappearance of skin lesions has been reported as a result of cytoreductive therapies in SM, the use of potentially toxic drugs in CM is not recommended. In all pediatric patients with severe CM, a persistently elevated serum tryptase level and anaphylaxis in medical history, equipping with epinephrine autoinjector for use in case of anaphylaxis is recommended. Complications: One boy with maculopapular mastocytosis since age of 3 months had anaphylaxis at the age of 2.5 years. After the anaphylactic attack, he had vision loss, epilepsy for months, but completely recovered by age of 4. One girl with maculopapular mastocytosis because of frequent itching and excoriation had severe thickening of skin. In her case, bulla forming was not observed.

**Conclusion:**

Cutaneous mastocytoma solitary form is characterized by a solitary reddish-brown nodule or plaque. In maculopapulosus form, lesions are multiple, macules, papules or nodules appear. In diffuse cutaneous form, diffuse orange-brownish plaques, infiltrated skin and in some cases, blister forming is observed. 65% of cutaneous mastocytosis is diagnosed in children, 35% in adults, typically, between age 18-30 years. The diagnosis in children is based in clinical picture, supported with Darier sign, and in some special cases, histology is also helpful. Histology shows a strong mastocyte infiltration in dermis. Molecular genetic analysis shows c-kit V600E mutation in 1/3 of children, and exon 6 or 9 mutation in 2/3 of children with cutaneous mastocytosis. Other examinations: general laboratory tests, abdominal ultrasound, and if possible, serum tryptase level helps to exclude systemic involvement. Cutaneous mastocytosis complications are based on the excess of mastocyte degranulated histamine in skin and serum, which might lead to itch, and also vasodilatation, thus severe anaphylactic reaction. Treatment of mastocytosis is limited to prevention of anaphylaxis with advices to avoid histamine liberation. Also antihistamines, to block H1 and H2 receptors are useful, as well as local corticosteroids. In some cases even epinephrine injector is recommended. In children the prognosis of cutaneous mastocytosis is usually good, most cases are spontaneously regress by adulthood.



**Abstract N°: 3400****clear cell papulosis: a reassuring diagnosis amongst a broad differential**Holly Fitzgerald<sup>\*1</sup>, Rachael Foster<sup>2</sup><sup>1</sup>Royal Perth Hospital, Perth, Australia, <sup>2</sup>Fiona Stanley Hospital, Dermatology, Murdoch, Australia**Introduction & Objectives:**

Clear cell papulosis (CPP) is an extremely rare, benign skin condition, first described in 1987. It is mostly seen in the paediatric population, predominantly in patients of Asian or Hispanic descent. We present a case of CPP and discuss the benefits of increasing clinician awareness of this condition.

**Materials & Methods:**

A 3-year-old girl was referred to the Dermatology clinic for hypopigmented papules over her lower abdomen, mons pubis and inner thighs. The eruption started when the patient was aged 12 months on the supra-pubic skin and then spread to the inguinal folds and upper thighs. There was no associated pain or pruritus, and the child was otherwise well. The eruption had been unresponsive to a trial of topical corticosteroid ointment. On physical exam, there were many small, scattered, hypopigmented papules over the suprapubic region and inguinal folds, some of which had coalesced into plaques. The labia majora, mucosal vulva, and perianal skin were spared. A 3mm punch biopsy was performed from both affected and unaffected areas. Involved skin demonstrated a papillated epidermis and singly distributed polygonal clear epithelial cells at the dermal-epidermal junction. Immunohistochemical staining was positive for CK7 and CEA. These clinical and laboratory findings were consistent with a diagnosis of clear cell papulosis.

**Results:**

CPP is characterised by asymptomatic, hypopigmented macules or flat-topped papules arising on the anterior chest, abdomen, and lumbar region, often along the mammary line. Multiple differential diagnoses can be made from this clinical picture, including papular elastorrhexis, guttate lichen sclerosus, guttate hypopigmented mycosis fungoides, pityriasis versicolour, plane warts and connective tissue naevi seen in association with Buschke-Ollendorf syndrome, such as elastomas, collagenomas, or fibromas.

The histological appearances seen on skin biopsy can distinguish CCP from conditions with similar presentations. Oval-shaped epithelial cells with bland rounded nuclei and moderately abundant clear cytoplasm are seen along the basal layer of epidermis, either as single cells or in clusters. Immunohistochemistry demonstrates consistently positive staining for CK7, carcinoembryonic antigen (CEA), EMA, CK-AE1/AE3, and variably positive results for mucin staining. Melanocytic and squamous markers, SOX10 and P40 respectively, are typically negative.

**Conclusion:**

Having increased awareness around this unusual condition may lead to earlier diagnoses by clinicians, and reduce unnecessary invasive investigations and treatments on patients, who are typically children. Patients and their families can be reassured by the asymptomatic and benign nature of the eruption, from which a majority of patients see eventual regression.

**Abstract N°: 3694****cyclosporine successfully treated a pediatric case of pityriasis rubra pilaris**

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**Cyclosporine successfully treated a pediatric case of pityriasis rubra pilaris**

Pityriasis rubra pilaris is a rare and chronic inflammatory keratinization disorder characterized by red-orange scaling erythema, follicular keratotic papules, and palmoplantar hyperkeratosis with islands of sparing.

An 18 -months-old girl was brought to a healthcare treatment by his mothers. The patient's mother was concerned about new onset rashes. A girl presented with erythematous papules and plaques on her limbs, trunk and face within one month. The progressive skin lesions begin from the trunk. The rashes were asymptomatic. The patient was healthy and had no systemic symptoms. Physical examination revealed keratotic plaques and papules in the upper and lower extremities and face. A 3 mm punch biopsy specimen showed hyperkeratosis, acanthosis, and Alternating vertical and horizontal parakeratosis in the epidermis (checkerboard appearance) with follicular keratinization. Despite initial therapy with topical corticosteroid and tacrolimus the papules and plaques spread on extremities and face. Oral cyclosporine was prescribed at a dose of 25 mg/ daily (2.5mg/kg).sh had marked improvement after one month and skin lesions completely resolved duration for 2 months with a sustained response after 6 months.

we reviewed the literature and found Pediatric case reports that were treated with various topical and systemic treatments but none of the patients received cyclosporine. In the present case, we made a decision to administer cyclosporine (2.5 mg/kg) significantly and sustained improvement with no evidence of adverse side effects. This case report suggested that cyclosporine might be considered as an alternative treatment for pediatric patients with refractory PRP.



**Abstract N°: 3709****Pediatric Cutaneous Mastocytosis**

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**Introduction & Objectives:**

Mastocytosis is a rare but not exceptional disorder due to increased proliferation and accumulation of mast cell in organs such as skin, liver, spleen and bone marrow, it can occur at any age but two thirds of the cases are observed in infancy (between birth and 2 years of age). Clinical findings, positive Darier's sign and histopathology confirm the diagnosis of Cutaneous Mastocytosis (CM).

Our objectives were highlighting Mastocytosis presentation in this particular age group. Description of the epidemiological, clinical, therapeutic and prognostic characteristics of mastocytosis in children.

**Materials & Methods:**

This is a long-term follow up study of the medical records of 19 pediatric cases that spans over 12 years; it includes pediatric cases confirmed histopathologically, and treated between September 2011 to January 2023. Clinical features at presentation during the follow up period were analyzed, and baseline serum tryptase level was measured in all of the 19 cases.

**Results:** 60% of the patients developed CM within the first year (median age 20 months). Boy to girl ratio was 0.7. A total 12 patients 63.1% had maculopapular cutaneous mastocytosis (MPCM), 15.7% had mastocytoma, 10.5% had diffuse cutaneous mastocytosis, 5.2% had systemic mastocytosis and 5.2% had mast cell leukemia.

Only two of the 19 patients had elevated serum tryptase, the remaining 17 patients showed continuously normal levels during follow-ups.

Complete resolution was registered in 10 patients as in 52.6%, regression was registered in 10.5% of the patients. 10.5% were non-responsive to treatments. Prognosis was good in all patients except one that ended with the death of the patient with mast cell leukemia.

**Conclusion:**

Our study of pediatric mastocytosis shines the light on the contrast between adult-onset mastocytosis and childhood-onset mastocytosis. Pediatric mastocytosis being usually transient.



**Abstract N°: 3730****Pachyonychia Congenita : A case report successfully treated with rosuvastatin**

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**Introduction & Objectives:**

Pachyonychia congenita (PC) is a rare genetic disorder characterized by nail dystrophy and painful palmoplantar keratoderma affecting quality of life. There is no effective treatment. Herein we present a case with classic features of PC treated successfully with rosuvastatin.

**Materials & Methods:**

A 15-year-old female presented to us with painful plantar hyperkeratosis and a thickening of all nails with a progressive course since age of 3 years. She was born of a non related parents with no history of genetic disease.

Since onset of clinical symptoms, she has had only emollients and keratolytics, and a short-course of oral acitretin with mild and transient improvement. She also reported recurrent plantar infections with blistering and painful fissures.

Cutaneous examination revealed focal yellow hyperkeratotic plaques of the soles that were more prominent on the heels, mid plantar region and both lateral forefoot, as well as an onychodystrophy of all nails. She complained of plantar pain upon walking. The patient also had follicular hyperkeratotic papules on her elbows and knees, and white plaques on both sides of her tongue with an angular cheilitis. There was no report of hyperhidrosis.

Diagnosis of PC was considered based on typical clinical features. Genetic testing could not be performed.

She was started on oral rosuvastatin 5 mg per day three weeks followed by a 7-day pause. After two months of treatment, she reported a marked decrease in pain (VAS 1/10) and a significant reduction of plantar hyperkeratosis. Follicular keratosis remained unchanged as well as oral leukokeratosis.

**Results:**

Pachyonychia congenita is a group of autosomal dominant disorders of keratinization caused by mutations in one of five keratin genes, namely KRT6A, KRT6B, KRT6C, KRT16 or KRT17.

The three most common clinical features associated with PC are thickened toenails (pachyonychia), plantar keratoderma (mostly focal) and plantar pain, which is the most important and debilitating feature affecting patients' quality of life. Additional associated features are pilosebaceous cysts, follicular keratoses, oral leukokeratosis, hoarse voice and natal teeth. Other possible features are ear pain and ear wax, angular cheilitis and abnormal sweating.

Although genetic analysis could not be performed, clinical features in our patient were consistent with PC. Additionally, she had associated features such as follicular keratosis, oral leukokeratosis and angular cheilitis.

There are no established treatment options available for PC. As painful plantar keratoderma is the most debilitating manifestation of PC, treatment options mainly focus on these lesions and include topical retinoids, salicylic acid, urea, topical steroids, surgical interventions, and medications for alleviating pain. Oral sirolimus showed a beneficial effect on palmoplantar hyperkeratosis and pain in PC as well as botulinum toxin type A.

Following promising results in studies on the use of rosuvastatin in the treatment of PC, and given the impact on our patient's quality of life, we started rosuvastatin using the same protocol reported in the literature. The patient had a significant improvement in pain and plantar keratoderma after only two months of well-tolerated treatment.

**Conclusion:**

PC is a rare genetic disorder that causes great disabilities to affected patients with a high impact on quality of life. Our observations suggest that rosuvastatin may offer a promising treatment for PC.

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**Abstract N°: 3741****Accessory tragus with an usual aspect: a case report**

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**Introduction & Objectives:**

Accessory tragus (AT) is a fairly common congenital malformation of the external ear. AT is generally presented as a congenital skin-colored papule or nodule located exclusively in areas derived from branchial arch. The preauricular location is the most widespread. It could be an isolated malformation or be a sign of associated congenital syndromes, such as: Goldenhar syndrome, Townes-Brocks syndrome, Treacher-Collins syndrome, VACTERL syndrome and Wolf-Hirschhorn syndrome.

Surgical excision is the treatment of choice. Its histological features include a thin layer of stratum corneum with a rugated epidermis, presence of eccrine glands, and irregular spatial positioning of vellus hair follicles accompanied by sebaceous glands.

This case shows an atypical appearance of an accessory tragus in location and shape.

**Materials & Methods:**

This is a clinical case whose diagnosis was based on clinical and histological analysis leading to definitive diagnosis of an accessory tragus.

**Results:**

A newborn boy, with no history, who has had a flesh-colored lesion since birth. The examination found a pedunculated nodule traversed by telangiectasias of fibrous consistency located at the level of the right cheek. The rest of the clinical examination was without abnormalities.

The patient underwent excision and the histological results of which showed polypoid-looking skin tissue lined on the surface by a regular squamous epithelium. The dermis is fibrous with a few adipose lobules with a cartilaginous tissue in the center related to a supernumerary tragus.

**Conclusion:**

An AT must be suspected in front of a nodule of congenital evolution sitting in areas derived from branchial arch. Our case illustrates an atypical aspect of the accessory tragus by its non-cartilaginous fibrous consistency and its unusual location.



**Abstract N°: 3766****Congenital skin aplasia: about two cases**

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**Introduction & Objectives:**

Congenital skin aplasia (CCA) is a rare congenital anomaly involving variable layers of the skin, most commonly affecting the scalp but can be seen on the trunk and limbs.

It is more often seen as solitary lesions or as part of a heterogeneous group of syndromes.

**Materials & Methods:****Results:****Case 1:**

This was a three-year-old newborn, from a non-consanguineous marriage to a 29-year-old mother.

No specific medication was given except for Covid treatment at the fourth month of pregnancy, and no smoking, alcoholism, or toxin intake. No autoimmune bullous dermatosis was present in the family.

There is a similar case in the older brother that regressed spontaneously at the age of one year and a half.

Our opinion was sought for an ulceration of the skin on the left leg.

The dermatological examination found a well-limited, reddish-orange, ulcerated patch with irregular borders, and surmounted by yellow crusts and purulent secretions.

Osteoarticular examination found a unilateral clubfoot of the limb.

The diagnosis of congenital cutaneous aplasia type VII was retained.

**Case 2:**

This was a newborn of one day of life from a non-consanguineous marriage of a 29-year-old mother.

No specific medication, smoking, alcoholism, or toxin intake were present.

No similar cases in the siblings and no notion of autoimmune bullous dermatosis in the family was present as well.

Our opinion was sought for a congenital absence of skin on the left hemisphere of the body.

The dermatological examination revealed multiple, well-limited, reddish-orange, ulcerated patches with irregular borders and a clean surface (Figs. 1 and 2), as well as hyperpigmented atrophic macules with a linear distribution along the Blaschko's lines along the left half of the body (Figs. 3a and 3b).

The rest of the examination revealed syndactyly of the left second and third toes with hypoplasia of the left great lip.

Trans-fontanelle and cardiac ultrasound returned without any particularities.

Biological tests were normal.

The diagnosis of a congenital skin aplasia associated with unilateral focal dermal hypoplasia was retained.

**Conclusion:**

Only a few cases of congenital skin aplasia have been reported regarding lower extremity location and association with focal dermal hypoplasia.

Regular follow-up is necessary for early detection and treatment of abnormalities due to focal dermal hypoplasia.

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**Abstract N°: 3782****Diffuse juvenile xanthogranuloma: a case report**

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**Diffuse juvenile xanthogranuloma: a case report****Introduction & Objectives:**

Juvenile xanthogranuloma (JXG) is a benign non-Langerhansian histiocytosis that classically presents as yellowish skin nodules due to accumulation of lipid-laden macrophages. It is usually unique but multiple or even disseminated forms have been described. We report here a new observation of diffuse juvenile xanthogranuloma.

**Materials & Methods:**

We report a rare case of diffuse juvenile xanthogranuloma (JXG) in a histologically confirmed infant followed in the dermatology department of the CHU Hassan II in Fez.

**Results:**

A 22-month-old infant, with no previous pathological history, who consulted for a diffuse asymptomatic eruption of progressive onset since the age of 4 months. Dermatologic examination revealed well-limited, firm, infiltrated, rounded to oval, orange papules that ranged from 0.5 to 2 cm in diameter on the face, trunk, upper and lower extremities sparing the palmoplantar region and without mucosal involvement. Darier's sign was negative, there was no milky coffee spot or other associated skin lesion and no palpable adenopathy. The histological and immunohistological study showed a juvenile xanthogranuloma of the CD68+, PS100+, non-Langerhans CD1a-histiocytosis type. There was no other systemic involvement or evidence that could point to neurofibromatosis type I. Spontaneous regression was the rule, justifying therapeutic abstention with close clinical monitoring.

**Conclusion:**

In summary, our observation illustrates a clinically atypical observation of JXG by the multi-lesional and multifocal nature of the lesions.

**Abstract N°: 3802****Pilomatrixoma: clinical and dermoscopic clues for diagnosis in pediatric population**

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**Introduction & Objectives:**

Pilomatrixoma, also known as “calcifying epithelioma of Malherbe” and “pilomatricoma” is an uncommon cutaneous tumor often affecting children and people over 50-60 years. Pilomatrixoma is a benign tumor of the pilosebaceous unit which causes concern for several reasons: the occurrence on the face and neck in most pediatric cases; the impact of facial surgery in children; possible similarity of the tumor with malignant tumors like dermatofibrosarcoma protuberans; the psychological distress of the family.

**Materials & Methods:**

We report 5 pediatric cases of pilomatrixoma highlighting the clinical and dermatoscopic clues for diagnosis, all cases being excised and histopathologically confirmed.

**Results:**

The cases were noticed in 5 female patients between 4 and 8 years old, with the disposition of the tumors on the left cheek, 2 cases on the left infrapalpebral area, the right preauricular and the right laterocervical area, presenting as indurated well demarcated blue-yellowish papules or nodules, observed for more than 6 months, associated with important psychological distress of the parents.

**Conclusion:**

Even if pilomatrixoma has long been considered an uncommon skin tumor, it is now reported as the most common solid cutaneous tumor in children. Considering all the above mentioned concerning features, the 5 cases of pilomatrixoma we are reporting will review the different facets of pilomatrixoma diagnosis in pediatric population.

**Abstract N°: 3827****H- syndrome: a case report with heterozygous slc29a3 mutation**

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H-syndrome is a rare and autosomal recessive disorder that is characterized by skin hyperpigmentation and hypertrichosis. Additional findings include hepatosplenomegaly, heart anomalies, hearing loss, hypogonadism, and low height. These manifestations are usually seen in childhood, but the diagnosis can be postponed by adolescence.

A 13-year-old girl was referred to out-patient dermatology clinic with 6 months history of progressing hyperpigmented and indurated plaques with hypertrichosis on the upper medial aspect of the thighs. She was born of full-term normal delivery, with no significant family history of genetic or medical illness. She had developed painless and progressive deformity and flexion contracture of fingers and toes and a hyperpigmented patch on the right palm.

Her skin examination revealed symmetrical ill-defined, hyperpigmented, indurated plaques with hypertrichosis over the inner sides of both thighs. Hyperpigmented and indurated plaques on the right palm and sole were detected. There was not any edema or varicose vein on the lower extremities. On skeletal examination; short stature, mild scoliosis, hallux valgus with fixed flexion contractures of the toe and finger joints and swan neck deformity were observed. No lymph node was detectable on examination. An ophthalmologic examination was normal. Abdominal ultrasonography and electrocardiography were normal but in the cardiac echocardiography, prolapsed mitral valve (MVP) with mild regurgitation (MR), and mild aortic insufficiency (AI) and mild trivial pulmonary insufficiency (PI) were detected. In the audiometry test of the right ear was normal but severe mixed hearing loss in the left ear was detected which was not clinically apparent. A skin biopsy from a hyperpigmented lesion on the lower extremity demonstrated partial epidermal atrophy and mild hyperpigmentation of basal keratinocytes. The underlying dermis revealed some thickening and sclerosis of collagen bundles, partially packed together, compressing adnexal structures, extending from the undersurface of the epidermis down to the upper border of hypodermis, and producing some dermal atrophy and upward dislocation of hypodermal fat lobules. There was a minimal infiltration of mononuclear lymphocytic inflammatory cells around blood vessels.

Although H-syndrome is a rare condition in the world it seems to be more common in some countries and races such as the Asian population and Arab people. The cutaneous manifestations are usually the first findings but other findings can affect the patient's quality of life such as cardiac defects, joint contractures, and hearing loss. So, it is necessary to distinguish an isolated cutaneous manifestation from a syndromic presentation.

There is no certain treatment for H-syndrome up to now and it seems every manifestation should be treated separately and conservatively. Although Bloom and colleagues mentioned that oral steroids may improve cutaneous manifestations in some patients, because of side effects, they cannot be as a maintenance treatment. Also, tocilizumab, azathioprine, and methotrexate are other choices for long-term treatment. Our patient was treated with methotrexate 10 mg per week to facilitate joint movements. After 2 months the patient claimed more mobility in finger joints but hyperpigmentation and induration did not change.

**Abstract N°: 3835****Dermoscopy patterns of 34 acral melanocytic nevi in children and adolescents**

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**Introduction & Objectives:**

Benign melanocytic acral nevi in children may give some concern in dermoscopy findings. Moreover, till date, there is limited information about the dermoscopic characteristics of these lesions. The objective of this study was to describe the characteristics and frequency of dermoscopic patterns in population under 18 years old.

**Materials & Methods:**

We evaluated a total of 34 dermoscopic images of acral melanocytic nevi from 34 individuals filed at the Dermatology Department of Universidad de Navarra Clinic between January 2000 and March 2023. Of these, 67,6% (n=23) were women and 32,4% (n=11) were men, with a mean age of 8 years old (range 2-17). We retrospectively examined the dermoscopic characteristics of all 34 images according to the standard dermoscopic classification criteria for acral melanocytic nevi.

**Results:**

The patterns observed were the typical parallel furrow, the fibrillar pattern, the lattice-like pattern, the homogeneous pattern, and combination patterns. The most prevalent were the combination patterns, which were seen in 9 patients (26,5%), followed by the parallel furrow pattern (20,58%). The combination pattern most commonly presented was the parallel furrow + homogeneous pattern, which was detected in 3 patients. Most of the pigmented lesions were located in soles (79,4%). Furrow and lattice-like patterns were more frequently seen on the palms, whereas the combination patterns and the fibrillar pattern were more prevalent on the soles. The parallel ridge, globulostreak, fibrillar and reticular patterns were found exclusively on the plantar surfaces. In addition, the frequency of combination patterns did not vary between the 0-11-year and the 12-18-year age-groups. Nevertheless, a significantly higher rate of parallel furrow pattern was observed in the 0-11-year age-group. As well, the fibrillar pattern was predominant in the 12-18-year age group.

**Conclusion:**

In conclusion, in our series we observed 6 different dermoscopic patterns on the palms and soles of 34 children. After analyzing these findings, we believe that these dermoscopic patterns could be associated with age and anatomical location.



**Abstract N°: 3847****Congenital cutis verticis gyrata in a one-month-old newborn with Turner syndrome: a rare clinical manifestation of this chromosomal disease and trichoscopic evaluation.**

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**Introduction & Objectives:**

Cutis verticis gyrata (CVG) is a rare disorder of the scalp that entails the development of ridges and furrows which mimic the anatomical conformation of the brain. This skin condition has been classified in primary essential, primary non-essential and secondary CVG, depending on the presence or absence of other associated disorders.

We present the case report of a one-month-old female newborn affected by congenital CVG (CCVG) with concomitant diagnosis of Turner syndrome (TS). The latter was made at pregnancy week 12 by chorionic villus sampling because of an increase in nuchal translucency on sonography.

The skin folding was present at birth and was located at the left fronto-parietal region of the head in the sagittal plane. It was raised, spongy, fleshy colored and with few hairs' growth above it.

Our purpose is to describe a further case of this rare scalp manifestation in association with TS by using non-invasive methods of investigation.

**Materials & Methods:**

Encephalic ultrasonography (U/S) was performed to exclude any correlated disorders underneath the scalp lesion. Additionally, U/S of the fold and trichoscopy were carried out to understand the general features of the skin condition and microscopic details, respectively.

**Results:**

Brain U/S investigation did not reveal any abnormality of the posterior cranial fossa and of the encephalic tissue. U/S of the skin fold and scalp showed regular soft tissue appearance and normal bony structures of the skull.

Trichoscopic studies of the ridge demonstrated absence of scarring alopecia. Indeed, it shown few hair with normal thickness scattered over the fold. Furthermore, it was not disclosed any change in the color of skin compared to the surrounding normal scalp.

**Conclusion:**

Our work aims to make this pathology better known clinically and trichoscopically so it could be more easily diagnosed and to encourage researchers to further study the association with the chromosomal anomaly. Moreover, by presenting this case we want to demonstrate how non-invasive methods may help to understand the main features of the skin lesion and to rule out anatomical changes below it. Additionally, the use of trichoscopical evaluation may give important clues to help in the classification of the lesion that should be finally confirmed by deeper investigations.





**Abstract N°: 3887**
**Acute Localized Exanthematous Pustulosis ALEP caused by Amoxicillin Clavulanic acid in a child**

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**Introduction & Objectives:**

Acute localized exanthematous pustulosis (ALEP) is an acute skin reaction characterized by the acute onset of multiple nonfollicular, pinhead-sized, sterile pustules, developed on an erythematous and edematous background, localized typically to face, neck, or chest. It is a rare variant of acute generalized exanthematous pustulosis (AGEP). Antibiotics, especially  $\beta$ -lactams, and macrolides, are the most frequent causative drugs.

Herein, we report an 8-year-old girl who developed ALEP in the shoulder and the neck in response to amoxicillin-clavulanic acid.

**Case report:**

We presented an 8-year-old girl admitted for an acute outbreak of multiple small non-follicular pustules on an underlying erythematous and edematous base, localized to the shoulder and the neck with mild itching. There was no mucous membrane or nail involvement; the child had no personal or family history of dermatological disease. She had been taking Amoxicillin- Clavulanic acid up to three days before the onset of symptoms for a urinary infection.\*\* The bacterial culture from the pustule was sterile. A skin biopsy from a pustular lesion of the neck showed acanthosis in the epidermis with mild spongiosis; in the dermis, there was a perivascular infiltrate of lymphocytes and neutrophils. The use of Amoxicillin-Clavulanic acid was discontinued and the pustules started rapidly to resolve within 5 days followed by residual scaling. Based on the clinical presentation of the eruption with the characteristic localized morphology of multiple non-follicular pustules along with the temporal association with drug administration of Amoxicillin- Clavulanic acid the diagnosis of ALEP was reached according to the recently proposed diagnostic criteria for ALEP.

**Discussion:**

ALEP was first reported by Shuttleworth (1989) as a localized, recurrent pustular eruption over the chin in a 42-year-old woman following the administration of amoxicillin. However, Prange et al in 2005 diagnosed the first case of ALEP over the face based on the diagnostic criteria for AGEF. Almost 40 cases of ALEP have been described in the literature in both pediatric and adult populations since the description of the first case by Prange et al.

The most commonly implicated drugs are antibiotics, especially  $\beta$ -lactams (including amoxicillin, Amoxicillin-Clavulanic acid ..), vancomycin, levofloxacin, and trimethoprim-sulfamethoxazole. Although in particular cases it is induced by bacterial, viral, or parasitic infection.

The diagnosis of ALEP is based on clinical features following the recent intake of a drug or contact with an uncommon agent; Some authors have proposed diagnostic criteria in line with Euro SCAR criteria for ALEP: Localized numerous small (1-3 mm) clustered non-follicular pustules.; background erythema; negative microbiology; acute onset (< 72 h) after medication and resolution (with post-pustular desquamation) within 14 days of discontinuing medication.

ALEP is a self-limited disease with a favorable spontaneous course within several days. The treatment is based on the immediate withdrawal of the triggering agent. In our case, only the withdrawal of the drug amoxicillin-clavulanic acid leads to resolving the lesions and scaling.\*\*

**Conclusion:**

ALEP is an uncommon cutaneous drug reaction. Most published cases involved antibiotics, but many other drugs have been reported to be responsible for this skin reaction. The pathogenesis and treatment of this disease are still unclear.

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**Abstract N°: 3904**
**Changing and emerging trends in Paediatric Dermatophytosis**

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**Introduction & Objectives:**

A recent trend of significant rise in the incidence of highly contagious and drug resistant dermatophytosis affecting most of the human body is reported and any existing anti fungal creams does nothing to alleviate it. Vulnerability of children to this disease have been detected more than adults due to multiple factors including detection of newly evolving species of fungus after COVID 19 pandemic is rising. However, limited data are available regarding clinical and mycological variants of dermatophytosis in children.

The present study was undertaken to assess the clinical patterns and mycological isolates from the lesions of dermatophytosis involving the paediatric skin.

**Materials & Methods: ### Inclusion criteria** All patients below the age of 14 years with clinically diagnosed dermatophytoses were included in the study. Informed consent was taken from parents or guardians of all the included cases. **### Exclusion criteria** Inability to obtain consent from parents or guardians and underlying health conditions that may increase risk for drug resistance were the exclusion criterion.

A total of 42 male and 25 female children (mean age – 6.2 years) presenting in the Department of Dermatology at a tertiary care medical college hospital were included in the study by the simple random sampling method for a duration of five months from August to December 2022.

A detailed history (duration of lesions, topical creams used, similar lesions in contacts, contact with pets, and other co-morbidities or co-medications) were taken and clinical examination (sites of involvement, number of lesions, associated erythema and scaling, morphology, and extent of lesions) were done followed by fungal mount preparation (potassium hydroxide (KOH) and KOH with Chicago Sky Blue stain) and culture of the scrapings on Sabouraud's agar medium containing chloramphenicol and cycloheximide.

**Results:**

Prior topical applications of steroids alone or with antifungal combinations were used in 57 cases (85%) before presenting to the dermatologist. About 76.12% ( $n = 51$ ) of cases had positive family/contact history. Thirty-five children (52.2 %) had extensive disease. Multiple atypical clinical patterns were observed: Tinea incognito, irregular geographic plaque, Tinea recidivans, Tinea pseudo-imbricata, Penile tinea, and annular plaques on scalp. Each clinical patterns will be described in detail in my presentation.

The groin was the most common site affected in 89.65% followed by face in 79% patients. KOH mount was positive in 52(77.6%). Fungal Culture revealed Trichophyton mentagrophytes (55.6%) as the most common species isolated.

**Conclusion:**

Our study illustrates a major epidemiological shift of dermatophytosis causing species from Trichophyton rubrum to Trichophyton mentagrophytes among paediatric patients.

It is also observed that there is an increase in an atypical presentation like *Tinea pseudoimbricata* and geographically patterned plaques owing to topical steroid use. The present study provides evidence that a high index of suspicion is needed to effectively curb the rising incidence of dermatophytosis with varied clinical morphologies in paediatric patients.

The study brings into focus the high percentages of topical steroid abuse by parents in children bought over the counter (OTC) and positive family/close-contact history in children and possibly a new fungal species resistant to conventional antifungal treatments.

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**Abstract N°: 4138****The novel treatment of children with molluscum contagiosum using microwave technology**Menno Gaastra<sup>1</sup><sup>1</sup>Huidmedisch Centrum Amsterdam, Centrum Oosterwal, Alkmaar, dermatology, Alkmaar, Netherlands**Title:**

The novel treatment of children with molluscum contagiosum using microwave technology

**Introduction & Objectives:**

Molluscum contagiosum (MC) is an easily transmitted pox virus that causes multiple individual benign papules associated with itching, pain, and inflammation most commonly found in children and immunosuppressed individuals. Although self-limiting, auto-inoculation can result in the lesions being present for months to years due to the virus' innate ability to hide from the host immune system. Here, we sought to investigate the feasibility of using microwave energy to treat MC in a small set of paediatric patients, following the success of this modality in resolving virally infected cutaneous warts. All patients had an atopic family history of asthma, bronchitis, hay fever, or eczema and failed prior treatments for their MC using different over-the-counter products.

**Materials & Methods:**

Using a microwave device designed and CE marked for use in dermatology, a total of 5 children aged 4 to 8 years old were treated with a non-ablative dose of microwave energy (2W for 2 seconds; 8GHz) applied to only the 3 to 5 biggest mollusca located on their trunk. Each lesion received 3 to 4 successive dose repeats of microwave energy before moving on to the next lesion. Patients returned to the clinic after 3 to 4 weeks, at which point a second treatment with microwaves was administered as before, if required, and a follow-up scheduled another 3 to 4 weeks later.

**Results:**

Using this treatment regime, all 5 patients had full resolution within 1 (2/5; 40%) or 2 (3/5; 60%) treatment visits. Because only a subset of the mollusca were treated with microwave energy, and previous over-the-counter therapies were unsuccessful in clearing the lesions, a systemic immune response similar to that previously suggested for HPV-infected cutaneous warts is a possibility and warrants further investigation. Microwave treatments can be associated with a certain degree of short-lived, acute pain, however in these cases the treatment was very well tolerated by the patients and both patients and parents reported to be "very happy" with this microwave therapy outcome.

**Conclusion:**

This small case report with excellent outcomes suggests that microwave therapy could become a new tool for the treatment of MC, for which there is currently no FDA-approved therapy available. Cantharidin is currently the main choice of treatment across many practices, however microwave therapy offers a less destructive/invasive and less painful treatment modality and may only require treatment on a subset of lesions. This would not only make the treatment faster in the clinic, saving both patient and clinician time, but also less traumatic to young children undergoing the treatment. Further research and studies into the safety and efficacy and underlying biomolecular and immune responses are required.

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**Abstract N°: 4160****LUMBAR syndrome: two cases report**

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<sup>1</sup>CHU Mustapha, dermatology, Alger, Algeria

**Introduction:**

The association of perineal infantile hemangioma with internal organs involvement of lower body parts has been described in the literature under various acronyms, such as PELVIS, SACRAL or LUMBAR. The latter is the most recent with around fifty cases in the literature. Recognition of these associations helps patient management. We report two cases of LUMBAR syndrome with Cryptorchidism, hypospadias and penoscrotal transposition.

**Case report:**

Case 1: A 15-month-old infant was referred to the dermatology department with a perineal red patch since the age of ten days. Clinical examination revealed a flat painful ulcerated hemangioma that extended over the perineal region, apartment feet, genu varum and bilateral cryptorchidism. Ultrasound found inguinal location of both testicles. MRI of the medulla and pelvis was without abnormality. The patient was treated by propranolol (3mg/kg/d), with marked improvement of hemangioma. The patient was referred to pediatric surgeons for further surgical treatment.

Case 2: A 5-month-old infant presented with a perineal infantile hemangioma that was ulcerated, with malformation of the external genitalia (penoscrotal transposition and hypospadias). Abdominal ultrasound and MRI of the spine revealed no abnormalities. The patient was treated by propranolol (3mg/kg/d) and was referred to pediatric surgeons for further surgical treatment.

**Discussion:**

Infantile hemangioma is the most common vascular tumor in infants less than 1 year of age. Segmental hemangiomas account for less than 10% of infantile hemangiomas, and perineal location is rare. They are often associated with extracutaneous abnormalities. The LUMBAR syndrome describes the association of a segmental hemangioma of the lower half of the body with anomalies of the spinal cord (attached cord syndrome, lipomyelomeningocele, anomalies of the terminal cone, etc.). Our two patients had no bone marrow defect. Urogenital anomalies are also associated (vesicoureteral reflux, clitoromegaly, atrophy, hypertrophy and asymmetry of labia majora and minora, cryptorchidism, hypospadias, micropenis, hydrocele, bifid scrotum, sexual ambiguity). Both our patients had genital anomalies. In addition, in LUMBAR syndrome, others anomalies can also be present such as bone, anorectal, arterial and renal anomalies. One of the two patients had deformity of the feet.

**Conclusion:**

This observation seems to confirm the variable expression of the LUMBAR syndrome with the possibility of an incomplete attribution of the different criteria of this syndrome. The clinical forms are variable, and the association of a perineal hemangioma with a lower body malformation should suggest this syndrome.

**Abstract N°: 4233****Purified Protein Derivative (PPD) Intralesional Immunotherapy for Wart In Children: Successfully Treated Cases**

Jun Ho Kwak<sup>\*1</sup>, Chan Ho Na<sup>1</sup>, Min Sung Kim<sup>1</sup>, Bong Seok Shin<sup>1</sup>, Hoon Choi<sup>1</sup>

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**Introduction & Objectives:**

Viral wart is a disease that occurs worldwide, affecting people of all ages. Current treatment options include topical therapies such as salicylic acid, and podophyllin, cryotherapy, laser treatment, and surgery. But there is no single treatment that is 100% effective. Recently, intralesional immunotherapies with various skin test antigens and vaccines have been known to be effective and safe for the management of warts.

**Materials & Methods:**

The authors tried to treat with intralesional immunotherapy using purified protein derivative (PPD) for 21 patients under 18-year-old clinically diagnosed as viral warts. Sixteen of them had a history of other treatment including cryotherapy, intralesional bleomycin, and other immunotherapies. The treatment responses were classified as one of three levels, based on the reduction in size and number of warts; complete response (CR, 100%), partial response (PR, >50%), and no response (NR, <50%). The treatment was performed every 2 weeks, and clinical evaluations were carried out by two dermatologists after 6 sessions of treatment. If there was no response to treatment, we considered other treatment options. On the other hand, if there was a response to treatment, treatment has continued for every 2 weeks.

**Results:**

The authors experienced 8 cases as CR and distant warts without treatment were also improved in some of them. Also, there were 8 patients who achieved PR. Significant side effects except tolerable pain and swelling for 1-2 days were not complained.

**Conclusion:**

Treatment for warts is especially challenging for children. Intralesional immunotherapy with PPD for warts is thought to be a suitable treatment option which showed no response to other treatments. Although this study has a limitation in case series form, our experiences suggest that further studies using PPD for warts in children should be undertaken, especially for the therapeutic efficacy of distant wart.



**Abstract N°: 4289****Pilomatrixoma as a prognostic problem**

Danijela Popovic<sup>1</sup>, Andrija Jovic<sup>\*1</sup>, Sladjana Cekic<sup>1</sup>, Zorana Zlatanovic<sup>1</sup>, Danica Tiodorovic<sup>2</sup>

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**Introduction & Objectives:** A pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a benign appendageal tumor with differentiation toward hair follicle matrix cells. It usually manifests as a solitary, asymptomatic, firm nodule. It is more common in children, but occurrence in adults is increasingly being recognized. They are usually less than 3 cm in diameter, although rarely they can be larger. They are most common on the head and neck or the upper body.

**Materials & Methods:** A seven-year-old girl with a subcutaneous nodule the size of grain of rice on the flexor side of her left upper arm. Change noticed by parents 6 months ago, growing slowly, without painful sensitivity. On examination, a solitary, well-circumscribed, subcutaneous nodule with hard consistency and a bumpy irregular surface measuring 1x0,5 cm was noted over the subcutaneous plane of the left upper arm. Dermoscopic examination revealed a structureless pink pattern. Ultrasonography revealed a well-defined, oval, hyperechoic lesion with posterior acoustic shadow in the subcutaneous plane of the left upper arm. Surgical excision of the mass was done which demonstrated features of pilomatrixoma on histopathological examination (HPE). She had an uneventful recovery, but after a year a similar nodule appeared in the right temporal region. Surgical excision was suggested. The parents were worried about the prognosis of the disease, possible scars from the excisions and insisted on possible prevention.

**Conclusion:** Pilomatrixoma (pilomatricoma, trichomatrioma, calcifying epithelioma of Malherbe) is a benign neoplasm or cyst typified by follicular matrical cornification. It has now been demonstrated that mutations in CTNNB1, the gene that encodes for  $\beta$ -catenin, are present in matrical neoplasms generally, including pilomatrixoma. Beta-catenin is a signaling, pathway effector that influences cell differentiation and proliferation, and mutations in  $\beta$ -catenin are thought to be present in pilomatrixoma generally. Pilomatricoma commonly presents as a single, firm, gradually expanding and mobile subcutaneous mass, however, the occurrence of multiple pilomatricomas have also been reported usually associated with disorders such as myotonic dystrophy, MYH-associated polyposis, Turner syndrome, Gardner syndrome, Rubinstein-Taybi syndrome, Sotos syndrome and gliomatosis cerebri.

**Abstract N°: 4340****hemorrhagic varicella secondary to dengue infection**Devanshi Nimbark<sup>\*1</sup>, Hita Mehta<sup>1</sup>, Shailee Desai<sup>1</sup><sup>1</sup>Sir T Hospital, Skin Ward, department of dermatology, venereology & leprosy , Bhavnagar, India

**Introduction:** In developing countries, infections contribute significantly to morbidity and mortality. As common as it is for co-infections to occur, it is rare for two viral co-infections to co-exist.\*\*

**Materials & Methods:** A 9-year-old female child presented to our OPD with a complaint of fluid-filled lesions on the whole body, including palms, soles, and oral cavity for 15 days. The lesions started as erythematous macules and papules, then became fluid-filled, followed by crusting and scab formation. They were associated with high-grade fever, runny nose, itching, conjunctival congestion, and diarrhea. History of similar complaints of fluid-filled lesions associated with fever was positive in siblings as well as many villagers. Based on the history and clinical examination, a provisional Hemorrhagic Varicella Zoster infection diagnosis was made. On admission, Complete blood count (CBC) revealed pancytopenia with Hemoglobin: 6.8 g/dL, Total Leukocyte Count: 600 cells/mm<sup>3</sup>, and Platelet Count: 20,000 cells/mm<sup>3</sup>. Persistent fever as well as altered CBC raised suspicion for a co-infection and further investigation revealed Dengue Ig-M to be positive. Later in the course of admission, she developed Bell's palsy secondary to varicella infection. The patient was managed with a multi-disciplinary approach by pediatricians, dermatologists, otorhinolaryngologists, and ophthalmologists. The patient responded well to injectable fluids, acyclovir, antibiotics, oral steroids, skin, and eye care.

**Results:** In India, Varicella zoster and Dengue fever are endemic viral infections that usually have a self-limiting course. However, if improperly managed, they can unusually transform into their serious hemorrhagic forms. Our patient developed a hemorrhagic varicella infection which later on was proved to be secondary to the dengue infection leading to reduced platelet count. It is unlikely for these two co-infections to co-exist unless the patient is immunocompromised. HIV as a cause of immunosuppression was ruled out as our patient was HIV-negative.

**Conclusion:** Thus, any change in the natural course of the disease or overlapping of clinical features should strongly raise suspicion for a co-infection. It should be diagnosed as soon as possible to ensure prompt treatment to prevent morbid complications.



**Abstract N°: 4342**
**Girl with congenital linear lesion and bone abnormalities in right upper limb**

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**Introduction & Objectives:**

In a newborn, the presence of congenital skin lesions and ipsilateral bone abnormalities raises a wide differential diagnosis in which complementary imaging studies and skin biopsy play a crucial role in accurately identifying the condition.

**Materials & Methods:**

Premature newborn at 34+2 weeks with a maternal history of systemic lupus erythematosus (SLE) presenting with a congenital cutaneous lesion consisting of papules and small confluent orange plaques in a linear distribution from the elbow to the right wrist, accompanied by hypoplasia of the second and third fingers of the right hand. An X-ray series reveals the presence of chondrodysplasia punctata in the right elbow, wrist, metacarpals, and second to fourth fingers, as well as shortening of the right humerus, second to fifth metacarpals, and phalanges of the second and third fingers. All the toes of the right foot show shortening and osteolysis. Skin biopsy reveals a proliferation of S100-negative granular cells consistent with non-neural granular cell tumor.

**Results:**

Congenital skin lesions in the limbs accompanied by developmental abnormalities present a diagnostic challenge. In this case, genetic syndromes with cutaneous mosaicism and skeletal anomalies, such as Goltz syndrome (or focal dermal hypoplasia), Conradi-Hünermann-Happle syndrome, and CHILD syndrome, are considered as possibilities. Additionally, the presence of chondrodysplasia punctata expands the differential diagnosis to include other conditions, including tibio-metacarpal chondrodysplasia, which has been associated with maternal SLE in some cases.

Regarding the congenital non-neural granular cell tumour, it is an extremely rare benign tumour with only five reported cases in the literature. None of these cases have described a linear distribution or association with limb developmental abnormalities. Therefore, this case represents the first instance of such a tumour with a segmental distribution and associated ipsilateral limb anomalies.

**Conclusion:**

The presence of congenital skin lesions and developmental abnormalities in a limb is indeed a complex clinical challenge. The association between the rare congenital non-neural granular cell tumor and limb anomalies has not been described so far. Therefore, it is possible that we may be dealing with a new entity or a previously unreported association.

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**Abstract N°: 4347****Pediatric vulvar lichen sclerosus: a case series**

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**Introduction & Objectives:**

Vulvar lichen sclerosus (VLS) is a chronic inflammatory dermatosis with a predilection for the anogenital region.

In the pediatric population, vulvar pruritus is the main symptom; it is most often treated wrongly by general practitioners as a repetitive pinkeye disease or considered an hygiene defect, which leads to a delay in diagnosis. Its treatment is effective as long as it is early; however, we are faced with the constraints of the location of the lesions and the side effects of the treatments applied.

Aim. To specify the clinical characteristics of (VLS) in the female pediatric population in order to plan early treatment.

**Materials & Methods:** Data were collected from case notes on female patients aged < 18 years diagnosed with VLS attending a dermatology service During a period of 5 years .

**Results:**

In total, 12 pediatric female patients were identified. The median age at onset of symptoms was 6,6 years. The histological study was carried out in only one case. The reason for consultation was mainly vulvar pruritus in 7 cases, a burning sensation in 3 cases, and achromia giving rise to suspicion of vitiligo in 2 cases. The association with urinary incontinence was found in 3 cases. In our patients, the LSV was associated in 2 cases with vitiligo, in 3 cases with alopecia, and in one case with Down syndrome. On clinical examination, skin sclerosis was noted in 05 cases, clitoral hooding in 03 cases, effacement of the labia minora in 2 cases, and vulvar edema in 2 other cases.

All our patients were put on ultra-high-potency topical corticosteroids at a rate of 5 days per week, associated with an antimycotic at a rate of 2 times per week, for a period of 3 months with a regression pattern. 10 of the patients had a satisfactory evolution; for the rest, we opted for the prolongation of the treatment up to 4-6 months. A maintenance treatment was prescribed based on dermocorticoids or topical tacrolimus, depending on availability, and the total duration of treatment varied from 9 to 18 months. In our patients, no recurrence of the disease was noted over a period of 3 years, nor were there any side effects due to the sequential application of dermocorticoids and their association with antimycotics.

**Conclusion:**

The diagnosis of LSV is clinical, with a good prognosis in children. If treated early, it evolves without sequelae. An individual approach for each patient allows for optimal treatment while avoiding any side effects.

**Abstract N°: 4375****A rare case of multifocal myositis ossificans progressiva : When soft tissues turns into bone.**

Asmaa Lahrougui<sup>1</sup>, Maryem Aboudourib<sup>1</sup>, Said Amal<sup>1</sup>, Ouafa Hocar<sup>1</sup>, Elmouhtadi Mouad<sup>2</sup>, Nassih Houda<sup>2</sup>, Elqadiry Rabii<sup>2</sup>, Bourrahouat Aicha<sup>2</sup>, Ait Sab Imane<sup>2</sup>

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**Introduction & Objectives:**

Myositis or fibrodysplasia ossificans progressiva, known as Munchmeyer's disease, is an extremely rare benign pathology, made up of a heterotopic proliferation of bone within the soft tissues which can lead to disabling ankylosis.

The diagnosis is based on a range of radio-clinical arguments. Optimal treatment has not been established.

Herein, we describe the case of Munchmeyer's disease in a female child of 11 years old.

**Materials & Methods :** Case report

We describe the case of multifocal myositis ossificans progressiva in a female patient of 11 year old, from a 2nd degree consanguineous marriage.

The patient had functional impotence due to ankylosis of both knees and ankles, and developed multiple budding, infiltrated and hyperkeratotic masses associated with ulcerations in the trunk and legs.

The diagnosis of multifocal ossificans Myositis was made on the basis of histopathological findings and X-rays results.

The skin biopsy showed a striated muscular tissue with artefacts of delayed fixation, the interstitium was fibrous with a polymorphic and diffuse granulation tissue mixed with a dystrophic bone tissue.

X-rays of the thorax, pelvis, limbs and extremities showed significant soft tissue ossification.

The patient was treated with oral corticosteroid therapy, methotrexate, oral bisphosphonate as well as daily hydrocolloid dressings, with good clinical improvement.

However, due to the difficulty of access to care and lack of means, the follow-up of the patient was difficult, her clinical condition deteriorated with fatal outcome.

**Discussion :**

Multifocal myositis ossificans is a rare benign pathology, representing 0.7% of soft tissue tumors.

In contrast to our observation, it affects preferably young male athletes and is often associated with trauma.

It is manifested by inflammatory pain and hard swellings located more in the anterior and proximal segments, which may lead to functional impotence in case of joint damage.

The radiological appearance depends on the histological maturation of the mass, which may be normal at first. CT or MRI can be used to make the diagnosis.

The treatment of MOMC varies according to the stage of the disease, ranging from no treatment to medical or surgical treatment.

Faced with the limits of treatment, prevention becomes very important, especially prevention of trauma, mandibular blockage and rehabilitation.

**Conclusion :**

This clinical case illustrates the rapid extension and functional impact of myositis ossificans and therefore the importance of early diagnosis to improve the care.

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**Abstract N°: 4380****Classification tool for congenital melanocytic naevi; a physician tool and patient self-classification tool.**

Anne Fledderus<sup>1, 2</sup>, Claire Van Eijdsen<sup>1</sup>, Suzanne Pasmans<sup>1</sup>, Juan Carlos López-Gutiérrez<sup>3</sup>, Sterre Mokken<sup>2</sup>, Phyllis Spuls<sup>2</sup>, Albert Wolkerstorfer<sup>2</sup>, Oren Lapid<sup>2</sup>

<sup>1</sup>Erasmus MC Sophia Kinderziekenhuis, Rotterdam, Netherlands, <sup>2</sup>Amsterdam UMC, locatie AMC, Amsterdam, Netherlands, <sup>3</sup>La Paz University Hospital

**Introduction & Objectives:**

Various cutaneous features of congenital melanocytic nevi (CMN) may be predictors for surgical treatment outcomes, psychological problems, neurological manifestations and melanoma. Especially larger CMN (>20 cm projected adult size (PAS)) are rare and occur in 1:20,000 new-borns. Therefore, standard and uniform classification is crucial. Krengel et al. developed a consensus-based standardized categorization of six cutaneous features of CMN. Besides this Krengel classification, a patients self-classification tool may be useful as a triage tool and to facilitate patient's involvement in their disease management and enhance communication between patients and physicians. This study aimed to (1) evaluate the interrater agreement of the Krengel classification and (2) to develop and validated a patient self-classifications tool.

**Materials & Methods:**

Two independent CMN specialists classified medium-to-giant CMN patients in the outpatient clinic. These same patients were asked to use the self-classification tool. Interrater agreement among all raters was calculated with weighted kappa.

**Results:**

Seventy-one patients were classified by the specialists, with 22 medium (> 1.5 cm PAS), 22 large (>20 cm PAS) and 27 giant (>40 cm PAS) CMN. The mean age was 8 years (range 1 month – 39 years). Fifty patients used the self-classification tool. Assessment of consistency among two specialists showed moderate to excellent interrater agreement for the six features (kappa 0.43 - 0.91). The agreement between specialists and patients was excellent for size and satellite naevi number (kappa 0.8 and 0.67) moderate for hypertrichosis (kappa 0.49) and fair for rugosity, noduli and color heterogeneity (kappa 0.21 -0.38).

**Conclusion:**

Both the Krengel CMN classification for specialists and the patient self-classification tool CMN showed excellent interrater agreement for the most important predictors for melanoma and neurological manifestations i.e. size and satellite naevi number. Moderate and fair agreement was shown for the other features. Such standard reporting of patients facilitates treatment comparison and eventually facilitating guidance on management.



**Abstract N°: 4411**
**Spindle cell Rhabdomyosarcoma of the hand in an infant at birth**

Meryem Khallouki<sup>1</sup>, Khadraoui Halima<sup>2</sup>, Hajar Boumehdi<sup>1</sup>, Maryem Aboudourib<sup>1</sup>, Rachidi Hind<sup>1</sup>, Said Amal<sup>1</sup>, Rais Hanane<sup>2</sup>, Ouafa Hocar<sup>1</sup>

<sup>1</sup>Mohammed VI University Hospital of Marrakech , Department of Dermatology and Venereology, Marrakech, Morocco, <sup>2</sup>Mohammed VI University Hospital of Marrakech , Department of Pathology , Marrakech, Morocco

**Introduction:**

Congenital rhabdomyosarcoma (RMS) is a rare soft tissue tumor with the most common sites of origin in the genitourinary tract, head, and neck regions and extremities are less commonly involved. RMS is classified into three types: embryonal, alveolar and pleomorphic. Embryonal type variants and related tumors include botryoid, pleomorphic, clear cell, rhabdoid, spindle cell, and sclerosing types.\*\*

We present a case of spindle cell RMS in the hand of a newborn female with regional lymph node involvement; the age, gender and site being unusual features for this type of rhabdomyosarcoma.

**Case report:**

A 5 -day -old girl born of non-consanguineous marriage presented with swelling of the left hand since birth involving the entire palmar aspect of the hand distal to the wrist crease. On examination, the mass was extending into the palmar aspect of the hand, firm, compressible, and non-pulsatile. Radiographs showed a soft tissue mass arising from the palmar aspect of the left hand without any bone involvement. Computed tomography (CT) angiogram showed a tissue formation centered on the palmar face of the left hand measuring 57\*45\*43mm. It is spontaneously isodense. It is associated with ipsilateral axillary lymphadenopathy. The infant underwent a biopsy and histopathologic studies revealed a malignant spindle cell tumor proliferation; immunohistochemical staining revealed strong positivity of anti-Desmin and anti- myogenin. VGLL2/NCOA2 molecular research was desirable but not available in our center. Based on histopathology, diagnosis of spindle cell RMS was made stage 3 of the Intergroup Rhabdomyosarcoma Study Group staging system IRS. The infant was referred for complete surgical resection, axillary lymph node removal followed by chemotherapy. Histological analysis of the surgical specimen showed an infiltrating spindle cell tumor proliferation with lymph node localization of RMS. Immunohistochemical staining revealed positivity of anti-Desmin and anti-Myod1.

**Discussion:**

Spindle cell rhabdomyosarcoma (RMS) is a rare variant of RMS, accounting for up to 10% of cases in infants.\*\* An international cohort of infants aged <12 months and diagnosed with spindle cell RMS between 1997 and 2018; reported one patient with metastatic disease and 39 patients with localized disease of which 10 patients had a location in the extremities. Only one infant had regional lymph node involvement (primary in the extremity). Patients were treated with a combination of therapies including chemotherapy, surgical resection, and radiation therapy (RT) according to their IRS stage. The 5-year EFS (event-free survival) was 86% and the 5-year OS (overall survival) was 91% suggesting a favorable outcome for infants with spindle cell RMS, with lower rates of relapse and possibly death than combined histologic subtypes of infantile RMS. The extent of resection was defined as a prognostic factor. On the basis of this largest study of infants with spindle cell RMS; the common first-line treatment recommended is R0 (complete resection) resection and systemic treatment with risk-adapted therapy.

Our case is worthwhile because of the extreme rarity of RMS in neonates. Furthermore, to our knowledge; it is the

second congenital RMS arising in an extremity with lymph node involvement. Most of the previously reported congenital RMS had male dominance, but our patient was female.

**Conclusion:**

It is important to keep in mind that RMS can be seen as a congenital mass in an extremity like a hand in neonates.

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**Abstract N°: 4460****Clearance of pediatric generalized granuloma annulare with cyclosporine**Khatere Zahedi<sup>\*1</sup>, Afsaneh Sadeghzadeh-Bazargan<sup>2</sup>, Ghazal Mardani<sup>1</sup>, Toktam Safari Giv<sup>1</sup><sup>1</sup>Skin Research Center, Shahid Beheshti University of Medical Sciences, <sup>2</sup>Iran university of medical Sciences**Clearance of pediatric generalized granuloma annulare with cyclosporine**

Granuloma annulare is an inflammatory granulomatous skin disease of unclear cause. The common subtype of Granuloma annulare with distinct clinical manifestations includes localized granuloma annulare, generalized granuloma annular and subcutaneous granuloma annulare and other uncommon subtypes containing perforating granuloma annulare and patch granuloma annulare. The most common variant of granuloma annulare localized and subcutaneous forms in the children. We report the variant of generalized papular and annular lesions in a 2-year-old girl. She had annular erythematous plaques composed of small papules and discrete papules were observed on the distal extremities, buttock and trunk with a 3-month history of these lesions. Skin lesions were asymptomatic. She was healthy and investigations of diabetes and thyroid disease were normal. Histopathological examination showed interstitial histiocytic infiltrate between collagen fibers and mucin deposition consistent with Granuloma annulare. She had been treated with 25 mg cyclosporine (2.5 mg/kg). After 8 weeks, the lesions were clinically resolved the consistent improvement was at the 3 months follow-up. To our knowledge, the use of Cyclosporine for Pediatric generalized granuloma annulare has not been reported. Management for the generalized granuloma annulare can be more difficult and it is often resistant to current treatment. Generalized granuloma annulare does not have a gold standard treatment and therapeutic options are based on small case reports, case series, and retrospective studies rather than randomized controlled trials. So we can consider granuloma annulare in the differential diagnosis of generalized papules and annular plaques in children and it appears cyclosporine may be an effective treatment without any side effects.



**Abstract N°: 4507**

**Syndrome de Blau, a propos d'un cas**

Ouerdane Nyhel<sup>1</sup>, Imad Eddine Rabia<sup>1</sup>

<sup>1</sup>Ain Naadja Military Hospital, Kouba, Algeria

**Introduction & Objectives:**

Blau syndrome is a rare autosomal dominant granulomatous disease. The cardinal signs of this condition are a triad of skin rashes, arthritis, and uveitis. However, the discovery of CARD15 mutations as being at the origin of this syndrome has made it possible to broaden the spectrum of clinical manifestations associated with these mutations, making this syndrome very close clinically to sarcoidosis.

**Materials & Methods:**

A 12-year-old child, with no particular history, presents with diffuse lupoma-type skin involvement associated with polyarthritis affecting the large joints with deformities affecting the upper and lower limbs and ocular type of bilateral sequelae anterior uveitis. The onset of symptoms dates back to the age of 9 months. The patient received corticosteroids for several years.

On exploration: presence of a biological inflammatory syndrome, immunological assessment was negative and the skin biopsy showed a tuberculoid granuloma without caseous necrosis

The genetic investigation is ongoing to research of the NOD2 gene

**Results:**

Blau syndrome and juvenile-onset sarcoidosis are autoinflammatory systemic granulomatoss associated with the NOD2 mutation. The NOD2 gene encodes a protein that is part of the family of intracellular pattern recognition receptors.

Blau syndrome and juvenile-onset sarcoidosis are familial and sporadic forms of the same disease, respectively. The classic triad is made up of rash, arthritis, and uveitis.

The disease usually begins before the age of three or four. Onset occurs primarily with skin and joint symptoms. However, symptoms sometimes appear after the age of 10.

**Conclusion:**

Blau Syndrome is a progressive and chronic disease with a variable and often unpredictable spectrum of severity. In case of widespread manifestations, life expectancy may be reduced. Uveitis has a poor prognosis. The diagnosis may be suspect early by a dermatologist by the demonstration of lupoma on vitropression.



**Abstract N°: 4582****Blau syndrome, case report**

Ouerdane Nyhel<sup>1</sup>, Imad Eddine Rabia<sup>1</sup>

<sup>1</sup>Hospital Central Army Dr Mohamed Seghir, Kouba, Algeria

**Introduction & Objectives:**

Blau syndrome is a rare autosomal dominant granulomatous disease. The cardinal signs of this condition are a triad of skin rashes, arthritis, and uveitis. However, the discovery of CARD15 mutations as being at the origin of this syndrome has made it possible to broaden the spectrum of clinical manifestations associated with these mutations, making this syndrome very close clinically to sarcoidosis.

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**Results:**

Blau syndrome and juvenile-onset sarcoidosis are autoinflammatory systemic granulomatoss associated with the NOD2 mutation. The NOD2 gene encodes a protein that is part of the family of intracellular pattern recognition receptors.

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**Conclusion:**

Blau Syndrome is a progressive and chronic disease with a variable and often unpredictable spectrum of severity. In case of widespread manifestations, life expectancy may be reduced. Uveitis has a poor prognosis. The diagnosis may be suspect early by a dermatologist by the demonstration of lupoma on vitropression.

**Abstract N°: 4603****Acquired Epidermodysplasia Verruciformis in a child with atopic dermatitis**

Hind Chagraoui<sup>1</sup>, Kenza Baline<sup>1</sup>, Hayat Skalli<sup>1</sup>, Farida Marniss<sup>2</sup>, Soumia Chiheb<sup>1</sup>

<sup>1</sup>ibn rochd university hospital, dermatology, <sup>2</sup>ibn rochd university hospital, anatomy pathology

**Introduction & Objectives:**

Acquired epidermodysplasia verruciformis (AEV) is a form of epidermodysplasia verruciformis (EV) due to Human Papilloma Virus (HPV 5 and 8) that is most commonly found in immunocompromised or elderly individuals, with the first cases reported in HIV subjects and organ transplant recipients. Currently, few data exist in the literature regarding AEV in the pediatric population, especially patients undergoing immunosuppressive therapy.

We report a particular case of a child with atopic dermatitis who developed acquired epidermodysplasia verruciformis.

**Observation:**

This is a 9 years old female patient, with a history of severe atopic dermatitis since the age of one year, recurrent pneumonia, with notion of chronic sinusitis, asthma, rhinitis and allergic conjunctivitis. She presented 6 months before her first consultation with verrucous papules on the back of her hands, forearms, and back giving way to a sequential hypopigmentation. In addition, she presented erythematous and finely scaly lesions on the folds of atopic dermatitis for which she was receiving topical tacrolimus and dermocorticoids with extension of the papular lesions becoming generalized. On clinical examination, non follicular verrucous papules were found on the upper and lower limbs and the back, confluent in places and evolving towards hypochromia. The histological study of the skin biopsy showed an epidermal hyperkeratotic hyperplasia with a minimal perivascular lymphocytic infiltrate without mitoses, in favor of an epidermodysplasia verruciformis. In the context of an associated immune deficiency, a biological assessment was required, which showed a total IgE level of 557 IU/ml and a hypereosinophilia of 760 e/mm<sup>3</sup>, with a negative HIV serology. The patient received Imiquimod 5% twice a week with a complete disappearance of the lesions after one month.

**Conclusion:**

The rapid disappearance of EV lesions in our patient after only 1onemonth of treatment and the absence of a family history of consanguinity argue for acquired EV rather than hereditary EV, the reason why no genetic testing was requested. Given the reports of acquired EV with human immunodeficiency virus (HIV), our patient had negative serology.

Our case constitutes the second case of the literature, the first case reported is an acquired EV in a 4-year-old child previously treated with cyclosporine for her atopic dermatitis, whose complete resolution of EV lesions was achieved in two weeks by topical application of imiquimod 5% three times a week.

The authors suggest a very likely role of immunosuppressive treatments in the initiation of EV by altering cell-mediated immunity.

**Abstract N°: 4614****Epidemiological and clinical profile of scabies in pediatric consultations**

Hind Chagraoui<sup>1</sup>, Fatima-Zahra Elfatoiki<sup>1</sup>, Hayat Skalli<sup>1</sup>, Hali Fouzia<sup>1</sup>, Soumia Chiheb<sup>1</sup>

<sup>1</sup>ibn rochd university hospital, dermatology

**Introduction & Objectives:**

Scabies is a contagious parasitic dermatosis caused by *Sarcoptes scabiei* that can occur sporadically, or epidemically in poor social environments. In adults, its presentation is usually typical and dominated by pruritus. However, in pediatrics, it often poses a diagnostic and therapeutic problem that varies according to the age group of the child.

The aim of this work is to recall the epidemiological and clinical characteristics of scabies in children through a series of 148 cases.

**Materials & Methods:**

This is a retrospective observational study of all cases of scabies followed in pediatric dermatology consultation over a 5-year period from January 2018 to January 2023.

**Results:**

We collected 148 cases, of which 33.8% (50 cases) were infants. The sex ratio (M/F) was 1.15. The median age was 6.2 years, ranging from 3 months to 15 years. The time to consultation ranged from 5 days to 7 months. Family pruritus was present in 77.7% of cases, and exclusively maternal in 84.5% of infants.

In this age group, an alteration of the general state such as agitation was reported in 84%. Palmoplantar pustulosis was present in 72%, axillary scabious nodules in 78% of cases. Involvement of the face in 64%, the back in 38%, and the scalp in 56%. In addition, a profuse pseudo erythrodermic form was observed in 12% of cases, favored by self-medication with dermocorticoids.

In children, furrows were observed in 30.6% with a Deltaplane sign on dermoscopy in 15.3%. The impetiginized form was present in 24.5%, and eczematized in 22.5%. There were no cases of Norwegian scabies.

The skin scotch test for *Sarcoptes* was performed in 44.6% and was only positive in 20.3%.

A scabicide treatment (benzyl benzoate) was prescribed in all patients with a single application of 12 hours in infants in combination with an antihistamine and emollient. We systematically recommended to treat the entourage, and to disinfect bedding and clothes. The persistence of scabious nodules was noted in 25 cases, a cure in 71 cases, 52 cases were lost to follow-up.

**Conclusion:**

We illustrate through our study, the clinical peculiarities observed in the pediatric population affected by scabies, which despite its frequency, often diagnosed late (a delay up to 7 months). The diagnosis is difficult, especially in infants, because the lesions are not very specific and are not always accompanied by pruritus, and often poses a problem of differential diagnosis with palmoplantar infantile acropustulosis. The peculiarities observed in children and infants, which vary according to age, could be explained by a difference in the distribution of pilosebaceous follicles and a thinner stratum corneum. In case of atypical clinical presentation, visualization of *Sarcoptes* can be

done by parasitological examination although there are cases of false-negatives, or by dermoscopy, or even a test treatment.

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**Abstract N°: 4653****An Atypical form of pediatric Pityriasis Rosea located in the perineum : Pityriasis Circiné et Marginé of Vidal**

Omayma Khadiri<sup>1</sup>, Kaoua Rim<sup>1</sup>, Maryem Aboudourib<sup>1</sup>, Said Amal<sup>1</sup>, Ouafa Hocar<sup>1</sup>

<sup>1</sup>University hospital of Marrakech, Department of Dermatology venereology, Marrakech

**Introduction & Objectives:** Pityriasis rosea (PR) is a benign, self-limiting, papulosquamous disorder characterized by the onset of a herald patch followed by an abrupt eruption of multiple salmon-colored papules and plaques on the trunk and proximal extremities. It mainly affects young adults and lasts for 2 to 12 weeks. Atypical cases of pityriasis rosea are fairly common and less readily recognized than typical eruptions.

**Observation:** We present two patients for whom we believe atypical PR is the most likely diagnosis. An 8-year-old girl had a sudden eruption of pruritic lesions, preceded seven days previously by coryzal symptoms. Examination showed multiple unguinal erythematous macules surrounded by a thin desquamative collar. The initial annular erythematous-squamous plaque was located in the median and upper part of the pubis measuring 4 cm on the long axis with a fine peripheral furfuraceous desquamation. Cutaneous biopsy revealed parakeratosis, epidermal spongiosis, dermal inflammatory cells, and extravasated red blood cells. The second clinical case is that of a 5-year-old girl who initially presented with a pubic Herald Patch followed 20 days later by erythematous-squamous lesions located in the perineum which did not respond to antifungals. The eruption consisted of little pink, oval macules, with a grayish peripheral scaling collarette around them. The palmoplantar and mucosal surfaces were not known. The rest of the skin and systemic examination revealed no abnormalities. We made a provisional diagnosis of pityriasis rosea of Gibert in its variant Marginata and Circinata of Vidal. Complete resolution followed a treatment with erythromycin for both patients. No recurrence was noted.

**Key message:** PR is a common, self-limited disease which in its typical form should not raise diagnostic doubts. Nevertheless, its atypical presentations can be a challenge for the clinician, hence the interest of knowing them. Recognition of these forms may allow a correct diagnosis and avoid unnecessary procedures.

**Abstract N°: 4692****Bilateral Harlequin Syndrome in a Pediatric Patient**

Thomas Akel Oberpaur<sup>\*1</sup>, Tania Marusia Capusan<sup>1</sup>, Javier Alcántara González<sup>1</sup>, Ana del Rocío Gamero Rodríguez<sup>1</sup>, Marta Andreu Barasoain<sup>1</sup>, María Elena Sánchez Largo<sup>1</sup>, Marta Ruano del Salado<sup>1</sup>, Leticia Calzado Villarreal<sup>1</sup>

<sup>1</sup>Torrejón University Hospital, Dermatology, Torrejón de Ardoz (Madrid), Spain

**Introduction & Objectives:** Harlequin syndrome is a rare autonomic nervous system disorder characterized by sudden bouts of hemifacial flushing typically triggered by heat, emotion, or physical exertion, requiring the exclusion of other etiologies. The absence of unilateral facial reddening is due to an ipsilateral dysfunction of the vasodilatory cholinergic sympathetic pathway. We present a pediatric case of Harlequin syndrome.

**Materials & Methods (Case report):** An 11-year-old patient with no notable past medical history was referred to our clinic from the emergency department for sudden onset erythematous lesions on the face and hyperalgesia of 3 weeks duration with episodes lasting hours. Initially treated as cellulitis with antibiotic therapy and hospital admission, the lesions resolved but subsequently recurred and followed an intermittent course. No other associated symptoms, local hyperhidrosis, or application of any topical product to the area were reported. Physical examination revealed a well-demarcated erythematous macule on the right hemiface and left hemiforehead, with increased local temperature, respecting the midline in both locations. Extensive laboratory testing, chest radiography, and cranial and cervical magnetic resonance imaging revealed no pathological findings. Neurology assessment was unremarkable. History highlighted academic-related stress as a trigger for outbreaks, with clear onset on school days and absence of lesions during weekends and holidays. With these findings, a diagnosis of bilateral Harlequin syndrome was made.

**Results (Discussion):** In 1952, Neligan and Strang described a vasomotor change in one hemibody during the neonatal period, which they named “Harlequin color change.” Subsequently, most described cases occur outside the neonatal period, affecting the face and neck in both pediatric and adult patients, predominantly unilaterally. We found few cases in the literature with bilateral presentation, as in our patient.

**Conclusion:** The role of the dermatologist is crucial in these cases for an accurate diagnosis and appropriate evaluation to rule out underlying pathologies.

**Abstract N°: 4720****A case of Harlequin Syndrome**

Yasmine Rkiek<sup>1</sup>, Ouiame El Jouari<sup>1</sup>, Salim Gallouj<sup>1</sup>

<sup>1</sup>University of Hospital Center of Tangier,, Dermatology, Tanger

**Introduction & Objectives:**

Harlequin syndrome is a dysautonomic syndrome of the face that was first described in 1988. It is a benign condition secondary to unilateral dysfunction of the sympathetic nervous system. We report a case.

**Materials & Methods:**

A 4-year-old patient without any notable pathological history was admitted for the exploration of a facial erythrosis associated with a strictly unilateral hyperhidrosis evolving for two years. The attacks occurred during effort and sometimes even during sleep. These attacks are made of redness and sweating of the left side contrasting with a heat and anhidrosis of the right hemiface. The ophthalmological examination revealed a slight heterochromia as well as an exophthalmos. The rest of the clinical examination was unremarkable, especially cardiac and neurological. The diagnosis of Harlequin syndrome was then made. The echo-Doppler of the supra-aortic trunks as well as the cervicothoracic and cerebral CT scans were normal, eliminating a tumoral or vascular cause.

**Results:**

Harlequin syndrome is a dysimmune syndrome of the face associating a facial erythrosis with flush and hyperhidrosis of a hemiface with a pallor and anhidrosis of the contralateral side.

It can appear as a Harlequin sign integrated in other syndromes such as Ross or Holmes Adie syndrome, or can be secondary to a neurological, vascular or traumatic process, as well as being idiopathic and then referred to as Harlequin syndrome as in this patient.

It is a rare syndrome that reflects a unilateral lesion of the autonomic nervous system. It can be triggered by different stimuli such as heat, emotions or physical effort, as in our case

In this condition, the red and sweaty hemiface is the side that reflects the normal response of the sympathetic nervous system and we talk about compensatory hypersudation.

It is therefore necessary to look for an organic abnormality on the opposite side: brainstem infarction, carotid dissection, neuroma of the upper mediastinum, trauma, cervical surgery or internal jugular vein catheterization. But most of the time, Harlequin phenomenon is idiopathic, and the diagnosis is therefore clinical as it was the case for our patient.

As far as treatment is concerned, there are few means apart from cases where the etiology is found and can be treated, a compression of the 2nd or 3rd thoracic root can be surgically removed. Contralateral sympathectomy is effective but it poses the problem of sequential radicular pain that is more disabling than the initial aesthetic damage and may be indicated if the social impact is major for the patient.

**Conclusion:**

Harlequin's sign can be congenital, secondary to a neurological, vascular or traumatic process, as well as idiopathic,

in which case it is called Harlequin's syndrome.

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**Abstract N°: 4722**

**Granuloma gluteal infantile: a redoubtable complication of dermocorticoid misuse**

Fatimazahrae Benhayoun<sup>1</sup>, Fouzia Hali<sup>1</sup>, Soumia Chiheb<sup>1</sup>

<sup>1</sup>CHU Ibn Rochd, Department of Dermatology and Venerology, casablanca

**Introduction:**

Dermocorticoids are used to treat a several inflammatory dermatoses to improve the patient's quality of life. However, their abusive and uncontrolled use could have numerous side effects. We report a case of a cortico-induced gluteal granuloma in an infant.

**Case report:**

A 5-month-old infant, with history of dermocorticoid application for diaper rash, presents with painless nodular lesions sitting on the breech without any other associated signs. The clinical examination found an erythema of the buttocks with multiple erythematoviolaceous nodular lesions of variable size located on the buttocks and the roots of the thighs. The rest of the somatic examination was unremarkable. We evoked a granuloma annulare or a histiocytosis. A skin biopsy was performed and showed acanthosis with orthokeratosis. The dermis was the site of a dense infiltrate of lymphocytes, histiocytes, and plasma cells, with some neutrophils and eosinophils forming microabscesses. There was no granuloma or signs of vasculitis. The diagnosis of gluteal granuloma was therefore retained. Management consisted of discontinuation of dermocorticoids and education of the mother regarding the misuse of this treatment.

**Discussion:**

The interest of this observation is to describe a dreaded complication of dermocorticoid misuse which is gluteal granuloma.

Gluteal granuloma is a dermatosis of infants characterized by the presence of small symmetrical nodules of purplish color, rounded or oval, on the buttocks and the upper-internal part of the thighs.

Histology shows hyperkeratosis with hyperacanthosis of the epidermis. Microabscesses can sometimes occur.

Healing is spontaneous in a few months. It could be a mycosis, but it is possible that it is due to the application of dermocorticoids especially in occlusion.



**Abstract N°: 4777****Dermatosis with linear arrangement in a paediatric patient; what should we suspect?**

Marielva Castro González<sup>\*1</sup>, Irasema Romero Gonzalez<sup>2</sup>, Karina Martínez Chavarría<sup>1</sup>, Josefina Navarrete Solís<sup>3</sup>, Diana Gonzalez Cabello<sup>3</sup>, Jaime Rojas<sup>1</sup>

<sup>1</sup>ISSSTE Clinica Hospital Constitución, Dermatology, Monterrey, Mexico, <sup>2</sup>Hospital General de Zona 33, Monterrey, Mexico, <sup>3</sup>IMSS - Unidad Médica de Alta Especialidad No. 25, Monterrey, Mexico

**Introduction & Objectives:**

Connective tissue nevi are a type of dermal hamartoma with variable clinical appearance. They present as skin-colored, yellowish or brownish plaques with a bumpy surface on the trunk or extremities.

“Orange peel” or cobblestone-like plaques can be observed. From the histopathological perspective, it is characterized by excess collagen or alterations in elastic fibers.

An even rarer variety of this type of tumors are those with a zosteriform pattern.

**Results:**

Case presentation: A 5 year old male with no relevant previous history. He came to the clinic for a dermatosis located on the trunk on the left flank which spread linearly towards the posterior trunk. It was characterized by a 17 x 1 cm plaque made up of multiple euchromic nodules of 5 to 10 mm in diameter, well limited, soft on palpation, with a tendency to coalesce. Mother refers 8 months of evolution, with increase in size and asymptomatic. Denies previous treatment.

Laboratories: Haemoglobin 11.1 g/dl, leukocytes 8.2 µl, platelets 329 10<sup>3</sup> µL, normal differential. Glucose 91, creat 0.4, normal LFT. ANAs (-), Anti SSA/Ro (-), Anti DNA (-)

Soft tissue ultrasound: Subcutaneous cellular tissue with homogeneous echogenicity, without evidence of collections or edema. preserved muscle planes. Diagnostic Impression: Skin nodules.

Biopsy: Hyperkeratosis with irregular acanthosis. Scant infiltrate of superficial perivascular lymphocytic predominance. In the dermis, a well-circumscribed, non-encapsulated lesion is observed, composed of numerous hyalinized collagen fibers.

PAS staining: abundant fractured hyalinized collagen bands, predominantly arranged parallel to the long axis of the epidermis.

Staining for elastic fibers: positive, abundant collagen fibers were evident.

With the previous data, a diagnosis of connective tissue nevus, collagenoma, was concluded.

**Conclusion:**

The diagnosis of these processes is based on clinical-pathological correlation. These types of lesions tend to be persistent over time, fortunately in this case no association with any other pathology was found, which has important prognostic implications. It is considered that surgical treatment, if desired, would be indicated for cosmetic reasons. Due to the characteristic presentation, previously described in some case reports, it is important to take connective tissue nevi into account in the differential diagnoses of those dermatoses that follow

dermatomes in paediatric patients.

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**Abstract N°: 4917****Diffuse cutaneous bullous mastocytosis in an infant treated with phototherapy**

Dimitrina Guleva<sup>1</sup>, Atanas Banchev<sup>2</sup>, Alexander Atanasov<sup>3</sup>, Valentina Broshtilova<sup>1</sup>, Vessel Kantardjiev<sup>1</sup>

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**Introduction & Objectives:**

Diffuse cutaneous bullous mastocytosis (DCBM) is a very rare subtype of mastocytosis especially in children. It is characterized by urticarial lesions, vesicles and bullae on the skin of the body and extremities. All adults and pediatric patients with severe DCM with a persistently elevated serum tryptase levels should be monitored for systemic involvement in case to prevent an anaphylaxis further. First-line of therapy are the systemic H1 and H2 antihistamines and topical corticosteroids whereas the phototherapy is as a second-line.

**Materials & Methods:**

We presented a 40-days male infant in a good clinical condition with diffuse urticarial and erythematous papular lesions on the skin of the face, body and extremities since birth. Serology for syphilis, HIV or bacterial infection was negative. Laboratory examinations were within normal ranges. No family history for dermatological or other systemic diseases. The abdominal ultrasound did not show any organ changes. The histology of the skin lesion showed a dense, diffuse dermal infiltrate of mast cells, consistent with DCBM. Serum tryptase levels were elevated >200 mg/l (referent ranges <15.0). Bone marrow biopsy was made and no systemic involvement was observed.

**Results:**

Systemic therapy with bimecizine maleate and cetirizine was started in a combination with topical steroid. There was no good therapeutical respond during those 3 months of follow-up and new bullae continued to show up. After careful discussion, NB-UVB phototherapy was started in low doses three times in a week for 1 month with an excellent respond – low tryptase titers and negative Darier sign were reached, no new lesions appeared, no clinical sign of scratching was noticed. During the last two years of follow-up on low doses systemic antihistamines and a strict low histamine diet, no new lesions or systemic involvement appeared.

**Conclusion:**

DCBM is a rare form of mastocytosis with an excessive skin involvement but with a very good prognosis.

Differentiation of other pediatric bullous dermatoses with different prognosis and treatment options is important. In our case, a complete disease control was achieved based on the combined therapy. However, a strict follow-up in every 6 months is needed in case to prevent a systemic involvement.



**Abstract N°: 4969****silvery gray hair - giving clue to diagnosis in an epileptic infant**Radhika S R<sup>\*1, 1</sup><sup>1</sup>Sapthagiri Institute of Medical Sciences and Research Centre, Bengaluru, India**Introduction & Objectives:**

Griscelli syndrome (GS) or partial albinism with cellular immunodeficiency is an uncommon disorder characterized by pigmentary dilution and variable cellular and humoral immunodeficiency. It belongs to a group of “silvery hair syndromes” which includes Chediak Higashi syndrome and Elejalde syndrome. It presents with pigmentary dilution of the skin and hair, recurrent skin and pulmonary infections, neurologic problems, hepatosplenomegaly, pancytopenia, hepatitis and immunologic abnormalities. Three mutations have been described in different phenotypes of the disease.

**Materials & Methods:**

Here is a case report of one-year-old male child who presented with seizures, delayed development of milestones, silver hair, eyelashes and eyebrows. History of consanguineous marriage present. Based upon history, clinical examination, neurological abnormality on MRI Scan and microscopic examination of the hair which showed large clumped melanosomes on the hair shaft, patient was diagnosed as Griscelli syndrome type 1 or Elejalde syndrome.

**Results:**

Symptomatic treatment for seizures was given and parents were encouraged for considering stem cell transplantation

**Conclusion:**

The prognosis of patients with Griscelli syndrome is grave. Curative hope is offered only by bone marrow or stem cell transplantation, which is more successful when, performed early in the course of the disease.



**Abstract N°: 5065****HOMA-IR index in pediatric psoriasis - a retrospective, analytic study**Adelina-Maria Sendrea<sup>1</sup>, Ana Preda<sup>2</sup>, Maria Tebies<sup>2</sup>, Alina Suru<sup>1</sup>, Carmen Maria Salavastru<sup>1</sup><sup>1</sup>Carol Davila University of Medicine and Pharmacy, Pediatric Dermatology, <sup>2</sup>Colentina Clinical Hospital, Pediatric Dermatology

**Introduction & Objectives:** Psoriasis is a multifactorial, immune-mediated chronic inflammatory skin disease associated with various metabolic comorbidities, including insulin resistance. While in adults these associations are clearly established, in children data is limited. HOMA-IR index is a helpful tool for insulin resistance risk assessment based on fasting glucose and insulin serum levels. This paper aims to determine the potential risk of insulin resistance in children with psoriasis, evaluated by HOMA-IR index assessment during the initial consultation and identification of possible clinical and paraclinical associations.

**Materials & Methods:** We performed a retrospective, analytic study on 30 children starting pre-pubertal ages, with various clinical types and severities of psoriasis for one year (January 2022- January 2023).

**Results:** In the study population, the mean age was of\*\* 12 years (the youngest patient being 6 years and 3 months and the oldest 17 years and 11 months), with a female gender predominance (63.3 %) compared to males (36.7%). Most patients (86.6%) had mild types of psoriasis, based on PASI score, while the rest presented with moderate (10%) and severe types of disease (3.4%). More than two thirds of the patients (76%) presented with elevated HOMA-IR index (>2), corresponding to insulin resistance risk, with mean value of HOMA-IR index being 3.59. All patients with either moderate or severe types of psoriasis associated abnormal values of HOMA-IR index. From the subpopulation with increased HOMA-IR index, most patients were diagnosed with either a disseminated type of psoriasis vulgaris (43.4%) or with scalp psoriasis (43.4%), while the rest (13.2%) presented with a generalized psoriasis vulgaris. Regarding the correlation between nutritional status and elevated HOMA-IR index, more than half of patients had elevated BMI, with 8.69% being overweight and 56.52% presenting with obesity. Moreover, 43.7% of patients with increased values of HOMA-IR index had dyslipidemia, as determined by blood lipid values. All patients with abnormal HOMA-IR index were referred to endocrinologists specialized in diabetes and metabolic diseases.

**Conclusion:** Insulin resistance represents a cardio-vascular and metabolic syndrome risk factor, known to be associated with psoriasis in adult patients. In children, data regarding such associations is still limited. In our study, we identified a high percentage of patients with insulin resistance risk, based on HOMA-IR assessment, risk also correlated with psoriasis severity, and, to a lesser extent, with overweight/obesity status and dyslipidemia, highlighting the systemic nature of this disease even in children. Our paper calls attention to the importance of insulin resistance screening starting with pre-pubertal aged children diagnosed with psoriasis, but larger, control-group studies are needed to establish a more accurate connection.



**Abstract N°: 5096****Multiple congenital smooth muscle hamartoma-case report**

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**Introduction & Objectives:** Smooth muscle hamartoma (SMH) is an uncommon benign skin disorder characterized by a proliferation of smooth muscle within the reticular dermis. SMH has been subdivided into two types, congenital (CSMH) and acquired (ASMH). CSMH is generally single, but multiple lesions have also been rarely reported in the literature.

Clinically it manifests as a hyperpigmented or skin-colored plaque where prominent vellus hairs may be observed.

Firts described by Sourreil et al in 1969.

**Materials & Methods:** We present a 4 year-old girl with history of unclear disorder of psychomotor development and positiv atopic anamnesis. Dermatological examination revealed multiple but individual and linear arranged skin-colored and hypopigmented, partially atrophic numular plaques diameter up to 12 mm, with hypertrichosis over some lesions. Some of the lesions were pail red. The localisations of the skin changes were along the extensor sides of the upper limbs and the front of the lower ekstremitas. Hyperpigmentation was not present and the lesions were asymptomatic. What is interesting, the lesions were first noticed by pediatricians two years ago during a physiatrist examination at the hospital. The first suspicion was long-term use of topical corticosteroids (the girl has mild atopic dermatitis) or artificial skin damage. During the dermatological examination, it was noticed that the mother has similar skin changes.

Excision biopsy was performed in both, the girl and the mother, and a histological diagnosis of CSMH was given.

**Results:** Since there were some lesions with erythema, we started therapy with pimecrolimus cream 1% twice a day over 3 months. We slowly reduced erythema, hypertrichosis disappeared and most of the lesions became extremely subtle. After another 3 months therapy with pimecrolimus cream 1% once a day there was no progress, but no new changes appeared either.

**Conclusion: :** Congenital smooth muscle hamartoma (CSMH) is defined as a rare asymptomatic benign skin disorder detected in newborns and young children, more frequently in male patients.\*\* Frequently involves the back and the lower limbs. Unusual cutaneous locations are the upper extremities. Multiple lesions have been reported rarely. Familial cases have also been recently reported, but a great majority of the cases occur in a sporadic fashion.

We present femail patient with multiple lesions of linear distribution over the upper and lower extremites, partially atrophic.

Therapeutic intervention is not necessary since CSMH is a benign tumor of the skin that is not associated with systemic manifestations and is not at risk of becoming malignant. The patient will be kept in clinical follow-up.

**Abstract N°: 5301****Halo scalp ring: A case born by caesarean section**Işıl İnanır<sup>\*1</sup>, Sule Yildiz Sagcan<sup>2</sup><sup>1</sup>Celal Bayar University, Dermatology , Manisa, Türkiye, <sup>2</sup>Celal Bayar University, Manisa, Türkiye**Introduction & Objectives:**

Halo scalp ring is a rare localized hair loss in the neonatal period. There is an annular temporary alopecia surrounding occipito-parietal scalp. The term was introduced by Neal et al in 1984 contributing its annular configuration. Though the incidence is unknown and the condition is thought as rare, it may be underreported. Here we present a case who was born by caesarean section.

**Materials & Methods:**

A 3-months-old girl had been brought for an scalp alopecia observed a few days after birth. She was born by caesarean section after a 37 weeks of uncomplicated pregnancy. There was no sign suggesting caput succedaneum at birth such as edematous swelling of the scalp. On dermatological examination a halo band of alopecia of 3-4 cm wide was noted around the scalp. The skin was normal except the alopecia. Halo ring alopecia was diagnosed. The alopecia disappeared completely at the age of 5 months.

**Results:**

The hair loss in halo ring scalp is present at birth or develops a short time after. There is generally an alopecic band of 1-4 cm surrounding parieto-occipital area.

The condition attributed to damage of scalp by instruments such as forceps and scalp monitors, or vacuum-assisted vaginal deliveries especially in newborns of primigravidas, and is usually associated with a caput succedaneum. The half of the published cases are premature infants. Perhaps it is related with the time required for the development of anagen hairs. Our patient was born by caesarean section, there was no trauma at birth and she was not premature.

.The prognosis is generally good, there are only a few scarring cases in the literature. Necrotic caput succedaneum and deep ulceration was reported with these cases. In the differential diagnosis there are other types of newborn alopecia such as sebaceous nevus, triangular congenital alopecia, and aplasia cutis. Its recognition can prevent unnecessary investigations. In most of the reported cases, total regrowth of hairs had been in 6 months follow-up.

**Conclusion:**

Halo scalp ring is a remarkable form of alopecia which is observed perinatally. Caput succedaneum and prematurity are related conditions. It requires no investigation and therapy. It may be underreported and should be kept in mind in the differential diagnosis of newborn scalp alopecias.

**Abstract N°: 5392****Title: Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder in children: a cases series of fourteen patients.**

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**Introduction & Objectives:** Previously known as primary cutaneous CD4-positive small/medium sized pleomorphic T-cell lymphoma, in the 2016 WHO classification this disease was renamed as primary cutaneous CD4+ small/medium pleomorphic T-cell lymphoproliferative disorder (PCSM-LPD) because of its indolent clinical course and excellent prognosis. This disorder has rarely been reported in children, there are only fourteen published cases, counting the largest series with four cases.

**Materials & Methods:** An observational, retrospective multicentre study was carried out including cases of paediatric PLPCP-LPD diagnosed at five centres. We performed a histological, immunohistochemical and molecular study of the reviewed biopsies. Available medical records of the patients were also reviewed.

**Results:** Fourteen patients were included, nine boys and five girls, aged between 1 and 16 years old. All patients presented as a single lesion and the most frequent location was the head. The histological findings were similar to those described in adults, with lymphoid proliferation in the papillary and reticular dermis composed of polymorphic lymphoid cells, recognizing small and medium-sized lymphocytes. The immunohistochemical study revealed a T phenotype (CD3+, CD4+, CD8-). The molecular study was positive for the monoclonal rearrangement of TCR gamma. All patients presented an indolent course with resolution after surgical treatment. In two cases, spontaneous regression was demonstrated after an incisional biopsy. In none of the cases recurrence was observed during follow-up.

**Conclusion:** We present the largest case series of PCSM-LPD described in the literature in the paediatric age. As in adult populations, paediatric PCSM-LPD appears to most commonly present as a single lesion on the head or neck and have an excellent clinical prognosis, and thus further staging cannot be recommended. Surgical excision is the preferred mode of treatment, although spontaneous regression after biopsy has been described, therefore observation may be an option in these patients.

**Abstract N°: 5396****Impact of itching, burning and pain in children with psoriasis: results from a monocentric study**

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**Introduction & Objectives:** The aim of this study is to evaluate the impact of clinical symptoms (itching, burning and pain) in children with psoriasis and possible correlations with severity of disease, clinical type of psoriasis, quality of life and cardiovascular comorbidities

**Materials & Methods:** We performed a monocentric study on 220 children (116 females; 104 males; age: 3-18 years) consecutively referred to our Dermatology Clinic. In each patient we collected clinical data related to age, sex, comorbidities (over-weight, obesity, central obesity), age at onset of psoriasis. We evaluated the severity of psoriasis using PASI (Psoriasis Area Severity Index) score. We assessed clinical symptoms (itching, burning and pain); the severity of itching was evaluated with the VAS score. The quality of life was evaluated with the Children's Dermatology Life Quality Index.

**Results:** Prevalence of itching in children with psoriasis was 85.9%; the prevalence of pain was 20%; the prevalence of burning was 39.1%. Statistical analysis showed that burning was significantly higher in males than in females (50% vs 29,3%;  $p=0,003$ ). Itching was significantly higher in children with the involvement of scalp compared with children without involvement of scalp (90,2% vs 9,8%;  $p=0,02$ ). Burning was significantly higher in children with the involvement of ano-genital area compared with children without involvement of ano-genital area (60,7% vs 39,3%;  $p<0,001$ ). Pain was significantly higher in children without the involvement of trunk compared with children with involvement of trunk (72,1% vs 27,8%;  $p=0,045$ ). The severity of psoriasis and the impact on the quality of life were also correlated with itching ( $p<0,05$ ).

**Conclusion:** To the best of our knowledge, this is the first study which assessed itching, pain and burning in children with psoriasis, reporting their association with sex, severity of psoriasis, quality of life and involved body areas. These data highlighted the importance of assess and manage these symptoms in childhood psoriasis.





**Abstract N°: 5732****Pediatric Lichen Planopilaris: Clinicopathologic Study of Six Cases and a Review of the Literature**

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**Introduction & Objectives:** Lichen planopilaris (LPP) is a form of cicatricial alopecia that has rarely been reported in the pediatric population. Because of the limited number of reported cases, little information is available. Our objective is to describe the clinical and histological features of pediatric LPP, in addition to the available treatment options for these patients.

**Materials & Methods:** This was a retrospective single-center study. Included were all pediatric patients with clinical and histological features compatible with LPP. All patients were evaluated by a pediatric dermatologist.

**Results:** Six pediatric LPP patients ages 7 to 17 years were identified (four males and two females). Two patients had scalp pruritus, and one patient had other cutaneous findings of lichen planus (LP). Perifollicular scale and scarring were the most common physical examination findings. All patients were treated with topical or intralesional steroids. Four patients were treated with hydroxychloroquine. Histopathologic findings included perifollicular interface and perifollicular fibrosis in all cases and mild dermal mucin in one case.

**Conclusion:** LPP is exceedingly rare in children. It may be misdiagnosed as alopecia areata in children because of the lack of symptoms and other features of LP. Early detection and targeted therapy are crucial in preventing hair loss and scarring, and there should be a high index of suspicion for LPP among the pediatric population.\*\*



**Abstract N°: 5746****Pediatric Sweet Syndrome: about 4 cases**

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<sup>1</sup>Hédi Chaker university hospital, dermatology, Sfax, Tunisia

**Pediatric Sweet Syndrome: about 4 cases****Introduction & Objectives:**

Sweet syndrome (SS) or acute febrile neutrophilic dermatosis is an inflammatory disease that classically affects adults. In children, SS is rare and presents a diagnostic and therapeutic challenge. The aim of this study was to investigate the clinical, therapeutic and evolutionary particularities of pediatric SS (PSS).

**Materials & Methods:**

We report a retrospective study of all confirmed cases of PSS in the dermatology department of Hédi Chaker Hospital in Sfax over a period of 12 years.

**Results:**

Our series includes 3 boys, aged respectively 3, 4 and 12 years, and one girl aged 50 days with an average age of 4.78 years. All the children had no particular medical history.

The lesions were of abrupt onset, papulo-nodular, erythematous and painful in all 4 children. These lesions were located on the face and upper limbs in 3 cases and on the lower limbs in one case. In the 50-day-old infant, the rash was associated with fever, ocular redness, and a hacking cough. Biological inflammatory syndrome was noted in all 4 cases. Histology confirmed the diagnosis in all cases. The etiologic workup revealed Crohn's disease in one patient and bronchopulmonary infection in another. A favorable evolution without recurrence was noted under colchicine in one patient, general corticotherapy in the second and antibiotic therapy associated with general corticotherapy in the infant.

**Conclusion:**

SS is the most common neutrophilic dermatosis. PSS has a mean frequency of 5%. The age of onset is comparable to our series with an average of 5 years. The form of SS described in infants is exceptional; the association with ocular involvement is rarely reported. Recently, Mc Clanahan et al. classified PSS into 3 age categories (neonatal, infant, and pediatric) in order to standardize the etiologic workup and management.

The search for associated diseases is essential. These are usually infections rather than neoplasia. Leukemia remains the most dreaded. The association with Crohn's disease, as in our patient, has been rarely described. The prognosis of PSS depends mainly on the severity of the associated diseases. The first-line treatment remains systemic corticosteroid therapy at a dose of 1-2 mg/kg/d of Prednisone equivalent but spontaneous resolution is possible.

Recently, Mc Clanahan et al. classified PSS into 3 age categories (neonatal, infant and child) with a view to standardizing etiology and management.

Pediatric SS is a relatively rare diagnosis. It may reveal or precede other conditions, which require rigorous and



prolonged surveillance. In general, the prognosis is good.

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**Abstract N°: 5756****Severe follicular occlusion triad and steatocystoma multiplex in a pubertal girl: a case report**

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**Introduction & Objectives:** Follicular occlusion triad (FOT) is a chronic inflammatory skin disease comprising dissecting folliculitis of the scalp, acne conglobata, and hidradenitis suppurativa (HS).

**Materials & Methods:** A 14-year-old girl was admitted with a history of multiple nodules that initially appeared on the scalp, approximately 18 months before admission. During that period, multiple papules and nodules appeared on her face, upper chest, and in her axillary region. The patient's BMI was 24.6 kg/m<sup>2</sup>, and personal history was unremarkable. Family history suggested multiple cysts and nodules in axillary regions in her grandfather and uncle.

**Results:** Physical examination of her scalp revealed numerous, solitary and confluent, inflamed nodules, some secreting pus, painful on palpation. On her face, in presternal and interscapular regions, erythematous papules, papulonodules, and open and closed comedones were noted. In her presternal and axillary regions, whitish-yellow papules were seen. The remaining physical examination was unremarkable. Thyroid function tests were normal. Gynecologic examination and ultrasound showed no abnormalities. Histopathological (HP) examination of presternal yellowish papule confirmed steatocystoma. Based on the clinical, laboratory, and HP examination, a diagnosis of FOT coexisting with steatocystoma multiplex (SM) was made. The patient is being treated with isotretinoin, 120 mg/day (1.71 mg/kg) and prednisone, 60 mg/day (0.86 mg/kg). Azithromycin and later amoxicillin+clavulanic acid were given orally, according to the bacteriological findings. The initial response resulted in slight clinical regression of skin lesions. Due to a short follow-up period, the long-term effects of the treatment remain to be seen.

**Conclusion:** Dissecting folliculitis of the scalp most commonly affects males and is rarely seen in pediatric patients, especially in girls. Only a few cases of FOT in children have been reported in the literature. SM has no sex predilection and usually appears in adolescence and early adulthood. Coexistence of HS and SM has been reported and can suggest an unrecognized defect in follicular keratinization process. SM can be seen in diseases linked to the mutation of the keratin 17 gene, like pachyonychia congenita type 2 (PC-2); therefore, it is important to conduct genetic testing. Our patient did not present PC-2 characteristics, genetic testing is underway. The conventional treatment can be challenging and sometimes is of limited efficacy; biologic therapy (e.g., secukinumab, adalimumab) has shown promising effects. Herein, we present a very rare case of a female pediatric patient with coexistence of very severe FOT and SM. Early diagnosis and comprehensive treatment of patients with severe clinical manifestations should be of utmost importance to prevent disease progression and reduce the impact of the disease on patient's mental health.



**Abstract N°: 5839****Blueberry muffin syndrome - two case reports**

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**Introduction:** Blueberry muffin syndrome is a rare, non-specific, clinical presentation in newborns, characterized by widespread maculopapular violaceous lesions. There are many associated underlying causes which include viral infections, hematologic dyscrasias and neoplasms. We report two cases in which this presentation disclosed two different diseases.\*\*

**Case reports:** A male neonate was born with prenatal diagnosis of anti-Kell alloimmunization after a 39-week healthy gestation. The 33-year-old mother was immune to rubella, toxoplasmosis, and cytomegalovirus. At birth, physical examination showed multiple, 3-7 mm, non-blanchable, violaceous to blueish papules distributed throughout the skin. The results of physical examination were otherwise unremarkable. Blood work indicated anemia and hyperleucocytosis (110.800/uL), with immature cells in the blood smear. Blood cultures and serologies excluded infectious causes. In skin histopathology there was an extensive superficial and deep infiltrate of immature granulocytes and blasts, with scarce segmented neutrophils in between. Immunohistochemistry was positive for myeloperoxidase, lysozyme, CD15, CD43 and CD68 and negative for S100, CD1a and CD10. Immunophenotyping of skin cells reported immature cell expansion with granulocytic and monocytic characteristics. Bone marrow immunophenotyping confirmed the diagnosis of acute myeloid leukemia with monocytic differentiation and the baby was started on chemotherapy. The second case reports a male neonate, with no described incidents during a 39-week gestation. The 41-year-old mother was immune to rubella and cytomegalovirus, but not toxoplasmosis. He presented, at birth, with multiple dark red, hard, 4-9 mm papules, widespread through the skin and hard palate. Laboratory examination showed no abnormalities, consisting of a full-blood count, infection markers, liver function and coagulation studies. Blood cultures and serologies excluded infectious causes. Skin histopathology demonstrated dermal clusters of histiocyte-like cells, some of them mononucleated and containing nuclear grooves, and other multinucleated, frequently with xanthomatous cytoplasm and intracytoplasmic hemosiderin pigments. There was no emperipolesis. Immunohistochemistry showed expression of CD68 and S100, and mononucleated cells were positive for CD1a. Abdominal ultrasound and skeletal X ray did not reveal any systemic lesion. Throughout 3 weeks of hospitalization, lesions slowly started to crust and to involute. Clinical evolution, histopathologic and immunohistochemical correlation supported the diagnosis of langerhans cell histiocytosis.

**Conclusion:** As blueberry muffin syndrome may be associated with a great variety of systemic diseases, its early recognition and meticulous investigation will determine the adequate treatment and prognosis of the newborn.



**Abstract N°: 5880****A 6-month-old boy with linear and whorled nevoid hypermelanosis - a case report.**

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**Introduction & Objectives:**

Linear and whorled nevoid hypermelanosis (LWNH) is a rare disorder of pigmentation characterized by hyperpigmented patches in a linear or whorled pattern along the Blaschko lines. We present a rare case of a 6-month-old boy with LWNH.

**Materials & Methods:**

A 6-month-old boy was referred to a dermatology department with asymptomatic, symmetric cafe au lait-colored skin lesions along the Blaschko lines. The patches appeared shortly after birth and gradually increased from the age of 2 months. There was no history of erythema or vesiculobullous lesions. Physical examination revealed symmetrical syndactyly of two toes and a flexed position of forearms in the sitting position. Laboratory tests revealed elevated level of total IgE. The histopathological examination of the skin lesions showed absence of melanocytes. Based on clinicopathological correlation, a diagnosis of LWNH was suspected.

**Results:**

The patient was referred for genetic tests, endocrinology, neurology and ophthalmology consultations.\*\*

**Conclusion:**

LMWH onset is usually in the first years of life with gradual progression. Hyperpigmented macules with irregular borders follow the lines of Blaschko and do not cross the midline. The face, palms and soles, eyes and mucous membranes are free of lesions. An estimated 30% of cases are associated with extracutaneous features such as skeletal, neurological or ocular. The differential diagnoses include: incontinentia pigmenti, hypomelanosis of Ito and epidermal nevus. In our patient, the clinical lack of hypopigmentation, vesicular or verrucous lesion and histopathology result helped us to exclude diagnosis of incontinentia pigmenti, hypomelanosis of Ito and epidermal nevus. Currently, there is no satisfactory treatment for LMWH.

**Abstract N°: 6012**
**Baseline characteristics and disease severity of children and adolescents with moderate to severe atopic dermatitis: First results from the German TREATkids registry**

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**Introduction & Objectives:**

Since 2020, the TREATkids registry, an expansion of the German Atopic Dermatitis (AD) registry TREATgermany, collects data on infants, children and adolescents with moderate to severe AD. In addition to topical and systemic therapy, disease progression, comorbidities and patient-reported outcomes are documented.

**Methods:**

TREATkids collects routine data as a non-interventional, prospective cohort study. The inclusion criteria of the registry are: infants, children and adolescents aged 0 to 17 years with moderate to severe AD according to the UK Working Party diagnostic criteria, an objective scoring for atopic dermatitis (oSCORAD) >20 despite guideline-compliant topical therapy and/or anti-inflammatory systemic therapy for AD within the past 24 months prior to inclusion in the registry.

**Results:**

By now, 328 children and adolescents have been recruited from 26 centers, 265 of them being eligible for analysis so far. Of these, 28.3% had already received systemic therapy before inclusion in the registry, including 10.9% of patients treated with systemic steroids. In the 12 months before inclusion in the registry, 90.2% of patients were treated with topical steroids and 48.3% with topical calcineurin inhibitors. The age distribution of the included patients is homogeneous (except for a higher number of infants) across all age groups between 1 and 17 years. 46.8% of registry patients (n=124) are male. More than half of the children and adolescents have immediate family members with a predisposition to AD (52.9%). The mean time of manifestation of AD is reported to be the fourth month of life. Concomitant diseases are allergic rhinitis (intermittent/seasonal) in 34.6% of patients and bronchial asthma in 22.4%. Among adolescents, attention deficit hyperactivity disorder (ADHD) is documented in 9.5%. Results of baseline visit data of patients without systemic therapy show a mean oSCORAD of 41.0 (SD 12.4) and a mean EASI of 14.9 (SD 9.7). At the completion of baseline visit, 41.9% patients of the registry (111 children and adolescents) are receiving systemic therapy. Dupilumab is the systemic therapy most frequently used or newly prescribed at the time of enrollment in the registry.

**Conclusion:**

Here, we report on the first German registry for infants, children and adolescents with moderate to severe AD.

With the expansion of the existing clinical registries in Europe for moderate to severe AD to children and adolescents, the routine care of this sensitive patient group will be documented. In the future, scientific questions on disease severity, care and treatment will be answered from a clinical and patient-reported perspective.

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**Abstract N°: 6076****Juvenile Dermatomyositis and localized scleroderma: a case of an overlap syndrome**

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**Introduction & Objectives:**

Dermatomyositis is marked by pathognomic cutaneous manifestations such as heliotrope rash, Gottron papules/sign, poikiloderma, and others, which may be present separately or in combination. They might be quite subtle, making diagnosis challenging. Muscle involvement may or may not be associated with skin symptoms. We present an uncommon case of muscular weakness with localised scleroderma of the right upper and lower limbs.

**Materials & Methods:**

A 15-year-old boy presented with a one-year history of erythema on his cheeks and décolleté and a large hypopigmented reddish macules on his left thigh, involving also left groin and leg. During the preceding 6 months, he complained fatigue and fatigability especially during football workouts. The boy didn't report weight loss and fever.

**Results:**

On physical examination the patient revealed a red erythema and edema that affected his cheeks, nose and upper eyelids compatible with heliotrope sign. The erythematous, confluent erythematous macules that affected the lower anterior neck and chest agreed with the so called "V sign". On the dorsal surface of his hands, particularly on the metacarpophalangeal and proximal interphalangeal articulations, there was also some violaceous papules and plaques. On dermoscopy exam we underlined the periungual erythema and abnormal telangiectasia.

A linear area of erythema and cutaneous induration with a central waxy, ivory color surrounded by a violaceous halo was underlined on his proximal left limbs, involving both the groin and knee region.

Laboratory investigations revealed normal blood counts, renal and liver function tests, except for elevated CPK and LDH. ANA were positive with a title of 1:160, anti-ENA test was negative, and myositis specific antibodies revealed a positivity on anti TIF1 gamma.

MRI of the thigh showed bilateral T2 hyperintensities in pelvic and thigh muscles suggestive of myositis.

According to clinical findings and laboratory investigations a diagnosis of overlap syndrome between linear scleroderma and juvenile dermatomyositis was made.

**Conclusion:**

The association between localized linear scleroderma and juvenile dermatomyositis is scarcely reported in literature, especially in paediatric age. We want to underline the importance of consider this association in clinical practice, beyond the importance of the multidisciplinary approach in the management of these patients.



**Abstract N°: 6079****New onset of Sapho syndrome in a 14-year-old boy: oral isotretinoin or Covid-19 vaccination induced?**

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**Introduction & Objectives:** We report the case of a young patient with severe acne who was diagnosed with SAPHO syndrome after starting isotretinoin therapy during the COVID 19 vaccination cycle. SAPHO syndrome, an acronym for Synovitis, Acne, Pustulosis, Hyperostosis, and Osteitis, is a rare chronic inflammatory disease that affects the skin, bones, and joints. It manifests with various symptoms, including joint pain, swelling, stiffness, skin lesions such as acne and pustulosis, and bone abnormalities like hyperostosis and osteitis. The exact cause of SAPHO syndrome remains not fully understood, but it is believed to be an autoimmune disorder characterized by the immune system mistakenly attacking healthy tissues, leading to inflammation and tissue damage. Cases of relapse of SAPHO syndrome following the initiation of isotretinoin therapy have been described in the literature, at the same time there is rare evidence of SAPHO syndrome induced by COVID 19 vaccine.

**Case Report:** A 14-year-old boy presented with multiple papulopustular lesions on his face and trunk, suggestive of acne vulgaris. Therefore, isotretinoin 20 mg/die was prescribed. After a few weeks, his clinical condition rapidly worsened, with the appearance of subcutaneous nodules, multiple arthralgias, and fever. The patient was admitted to the hospital and isotretinoin was interrupted. X-ray and scintigraphy of knees, ankles, and feet excluded joint involvement. Based on clinical and radiologic findings, a diagnosis of acne fulminans was made. Prednisone 25 mg/die was started, with amelioration of both cutaneous lesions and articular pain. While tapering prednisone dosage, the patient referred the onset of joint pain in his knees and lower back. The patient completed covid19 vaccination cycle three weeks before the onset of the lesions. These results led to a diagnosis of SAPHO syndrome, and Neridronate ev was prescribed. For acne maintenance treatment, topical therapy with benzoyl peroxide/adapalene was administered.

**Conclusion:** Isotretinoin is a medication primarily used for the treatment of severe acne, but it has been associated with musculoskeletal symptoms such as arthralgia, myalgia, and back pain. Some rare cases reported in the literature suggest that isotretinoin may also have the potential to trigger or worsen SAPHO syndrome. The exact mechanism by which isotretinoin can induce or exacerbate SAPHO syndrome is not fully understood, but it is believed to be linked to its impact on bone metabolism and the immune system.

On the other hand, while there have been reported cases of individuals developing inflammatory conditions following COVID-19 vaccination, there is limited evidence regarding the association between SAPHO syndrome and COVID-19 vaccination. Recently, a case of SAPHO syndrome after COVID-19 vaccination, accompanied by thyroid dysfunction, has been documented. Consequently, it is challenging to definitively determine which of the two factors played a more significant role in the onset of SAPHO syndrome.



**Abstract N°: 6088****Granulomatous variant of lichen aureus in a child**

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**Introduction & Objectives:** Lichen aureus (LA) is a rare type of chronic pigmented purpuric dermatosis, that commonly occurs in young adults, but less frequently in children. Its granulomatous variant is a rare subtype distinguished by detection of a granulomatous infiltrate on histopathology.

**Clinical case:** An 11-year-old boy was referred to our department because of the appearance of an asymptomatic brown-gray macular eruption localized on the anterior area of the distal third and ankle of the left leg in the last 4 months. Patient denied neither trauma nor having applied any substance. A biopsy was taken showing a slight acanthosis and hyperkeratosis in epidermis as well as an inflammatory infiltrate of mononuclear cells, with hemosiderophages and granulomas around superficial and middle capillary plexus. Congestive vessels and erythrocyte extravasation were also found. With these clinical and histological findings, a granulomatous lichen aureus (GLA) diagnose was made. A blood test was carried out without relevant findings. Tacrolimus 0,1% ointment was prescribed without clinical improvement.

**Discussion:** LA in childhood is well described in the literature, with an estimated frequency of 0,05% in pediatric patients with dermatologic diseases. Histologically there is a perivascular lymphohistiocytic infiltrate in a bandlike pattern, although a perivascular predominance can be found as in our case. The granulomatous variant is less common, and usually has been associated with hyperlipidemia in some case series, not appearing in our patient. Treatment is usually unsatisfactory.

**Conclusion:** We present a rare case of GLA in a child, not associated to analytical disorders.



**Abstract N°: 6090****Spinular follicular keratosis of Siemens: A triad that cannot be ignored**

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**Introduction:**

Spinular follicular keratosis of Siemens (SFKS) is a rare X-linked or sporadic genodermatosis characterized by follicular hyperkeratosis and scar alopecia, palmoplantar hyperkeratosis (PPK), photophobia, corneal abnormalities, and atopy. Males are severely affected, but females can also develop severe lesions in the context of usually X-linked inherited diseases. We report three cases of SFKS confirmed by biopsy.

**Cases reports:**

The first case was a five-year-old boy from non-blood parents with a reported absence of hair at birth and no complaints of photophobia or atopy. No other similar cases were reported in the family. Dermatological examination revealed diffuse cicatricial alopecia of the scalp, eyelashes, and eyebrows, and hyperkeratotic papules on the forehead and body, associated with localized PPK and pachyonychia. The second case was an eight-year-old boy from a nonconsanguineous marriage who had diffuse hair loss with depilation of the eyelashes and eyebrows since the age of two months, accompanied by photophobia, with no other similar cases in the family. Dermatological examination showed scar alopecia on the scalp, complete depilation of the eyelashes, eyebrows, and body, and follicular papules on the forehead and temporal region with localized KPP. The third was a five-year-old girl from consanguineous second-degree parents with hair breakage and thinning without photophobia from three months of age, with a similar case in her younger sister. Dermatologic examination of both sisters revealed short, thin woolly hair, depilation of the eyelashes, eyebrows, and rest of the body with the appearance of goosebumps, and plantar hyperkeratosis located on the heels of both feet. A histopathologic examination of the scalp confirmed the diagnosis of SFKS in three of our patients, and an ophthalmologic examination revealed no abnormalities. All patients were prescribed topical keratolytic and emollient creams, and two of them were prescribed oral acitretin, which improved keratosis pilaris and palmoplantar without affecting the hair.

**Discussion:**

SFKS is a rare disease with X-linked or sporadic genetic transmission characterized by follicular hyperkeratosis and scar alopecia. The pathophysiology of hair follicle destruction is not yet fully understood. The disease usually begins in early childhood and worsens in adolescence, initially on the face and progressing to the trunk and extremities. PPK, photophobia, corneal abnormalities, and atopy may occur. Therapy is not very effective, and treatment is ineffective in the presence of changes that are mostly scarring.

**Conclusion:**

Although rare, SFKS should always be considered in all cases of follicular and/or palmoplantar hyperkeratosis with alopecia because, in addition to the genetic counseling necessary in some cases, treatment for this condition should be started as early as possible to delay and minimize scarring.

**Abstract N°: 6100****Pediatric Cellular Neurothekeoma**

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**Pediatric Cellular Neurothekeoma****Introduction & Objectives:**

Cellular neurothekeoma is a rare, benign and superficial skin tumor with fibrohistiocytic differentiation, most frequently located on the head and neck. This is an infrequent tumor, and it is not usually seen in pediatric population since it appears in the second or third decade of life.

We present a clinical case report of a female pediatric patient that presented a tumor that was clinically and histologically compatible with cellular neurothekeoma.

**Materials & Methods:**

A 10-year-old female presented a single cutaneous nodule located on the inner side of the right thigh, 1.9x1.3 cm of size, multilobulated, heterogeneous pink coloration with peripheral pallor, central erythema and peripheral telangiectasias, irregular surface and borders, well delimited, with a hard and painless consistency. It appeared 3 years before and it had an abrupt growth in the last 3 months.

**Results:**

Dermoscopy examination revealed a yellowish central area, with non-adherent whitish scale, pale pink periphery with linear telangiectasias. Complete removal of the lesion was performed, including part of the subcutaneous cell tissue.

The histopathological study revealed a dermal tumor composed of large epitheloid cells with mild pleomorphism, ovoid, vesicular nuclei, visible nucleolus, and eosinophilic, pale, and vacuolated cytoplasm, arranged in nests and a plexiform pattern. Immunohistochemistry showed a positive CD10 and CD63 reaction, compatible with cellular neurothekeoma.

**Conclusion:**

Cellular neurothekeoma is a distinctive benign cutaneous tumor of uncertain histogenesis. It appears as a solitary, slow-growing, asymptomatic nodule and it measures less than 1 inch. Differential diagnosis includes myxoid neurothekeoma and melanocytic lesions with a plexiform pattern. Since neurothekeoma can clinically and histologically mimic benign and malignant tumors, it is important that dermatologists and pathologists are familiar with this entity in order to make an accurate and timely diagnosis as well as an appropriate treatment.

**Abstract N°: 6111****spitzoid melanoma in an 11-year-old child: a rare entity**

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**Introduction & Objectives:**

Cutaneous melanoma is an extremely rare tumor in children (1) it has atypical clinical aspects, hence the delay in diagnosis and therefore in management. Spitzoid melanoma is a rare form of melanoma, difficult to differentiate from Spitzoid nevus, which makes its diagnosis difficult in children. We report a Spitzoid melanoma in an 11-year-old child.

**Results:**

This is an 11-year-old girl, M.N, brought by her parents to a dermatologist for a nodular lesion of the right cheek of about 2 cm in diameter, evolving over a few months, initially evoking a Spitz nevus, however, given the ulceration that followed, an excisional biopsy was done with anatomopathological and immunohistochemical study (HMB 45 and Melan A heterogeneous, P16 -, Cyclins D1+ and high Ki67, PS100 strongly positive) concluding to a Spitzoid melanoma, the Breslow index and the Clark level were not calculated. A revision surgery with lateral and deep margins of 2 cm was performed, the margins were healthy. The patient was then referred to our level and an extension work-up was performed: brain and cervico-thoracoabdomino-pelvic CT scan , PET scan and ultrasound of the lymph nodes did not reveal any secondary location. The decision of the PCR was to monitor the patient clinically and radiologically. One and a half years later, the patient presented with metastatic subcutaneous adenopathy without capsular invasion, which led to a complete cervical lymph node which did not reveal any other lymph node involvement. No other metastatic location was found in the extension workup. Since then, she has been followed and monitored regularly clinically and radiologically.

**Conclusion:**

With our observation, we highlight a rare entity in children, the Spitzoid melanoma, so close to Spitz nevus, which incites us to perform the removal of any lesion with doubtful characteristics or rapidly changing appearance.

**Abstract N°: 6133****Childhood Granulomatous Periorificial Dermatitis: Exploring Novel Approaches for Management**

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**Introduction & Objectives:**

Childhood granulomatous periorificial dermatitis (CGPD) is a condition characterized by small, monomorphic papular eruptions around the mouth, nose, and eyes, with a granulomatous pattern on histopathology. Its etiology remains unknown. CGPD primarily affects prepubescent children of both sexes and typically resolves without scarring after several months.

**Materials & Methods:**

Case report and review of articles in Medline

**Results:**

A 9-year-old boy came to our clinic because of with a 8-month history of asymptomatic erythematous micropapules around the nose, eyes, and mouth. The patient had a medical history of asthma and was being treated with inhaled salmeterol/fluticasone, oral montelukast, and inhaled ipratropium and salbutamol for asthma exacerbations.

Previous skin treatments included topical corticosteroids (cream and ophthalmic ointment), systemic corticosteroids, pimecrolimus cream, oral clarithromycin, boric acid, and emollients. On physical examination, numerous monomorphic papules with erythematous-pink coloration were observed in a peri-orificial distribution on the face (periocular, perinasal, and perioral regions), as well as in the glabella and supraciliary areas, without pustules.

Skin biopsy revealed unremarkable epidermis and a scant perivascular lymphohistiocytic inflammatory infiltrate in the mid and deep dermis, along with epithelioid granulomas without necrosis or lymphocytic crown, consistent with the diagnosis of childhood granulomatous periorificial dermatitis.

The initiation of topical tacrolimus and ivermectin led to significant improvement, although some periorificial millimeter-sized papules persisted. After a single oral dose of ivermectin (214 µg/kg) there was almost a complete clearance of skin lesions.

**Conclusion:**

In the therapeutic management of childhood granulomatous periorificial dermatitis, discontinuation of all topical corticosteroids should be the first step, which was not possible in this case. Besides, our patient demonstrated resistance to other conventional treatment options, possibly due to an unmodifiable trigger.

Topical ivermectin has shown efficacy in papulopustular rosacea and periorificial dermatitis in children. Besides the anti-parasitic effect on *Demodex* spp., ivermectin also exhibits anti-inflammatory properties by downregulating gene expression levels of IL-8, LL-37, HBD3, TLR4, and TNF-α in vivo. We believe the same mechanism applies to

this pathology.

In this case, a systemic approach yielded excellent results, despite the incomplete response to the topical formulation.

Although childhood granulomatous periorificial dermatitis is considered a self-limiting condition, the presence of numerous lesions can cause discomfort in patients, necessitating prompt consideration of treatment options. Comorbidities from the atopic spectrum are likely to be common, making it important to explore alternatives that can address the need for maintaining the use topical corticosteroids.

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**Abstract N°: 6147****The impact of the Atopy School on short- to medium-term Outcome of atopic dermatitis in children**

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**Introduction & Objectives:**

Atopy School is an active workshop for children suffering from AD(AD) on a monthly basis for the purpose of therapeutic education. It employs multidisciplinary personnel: dermatologists, pediatricians, psychologists, and nurses, enabling parents and children to manage the disease on a daily basis

The Mohammed VI Universal Hospital of Marrakesh has developed the first Moroccan Atopy School, providing an educational experience for those dealing with AD, which can be a source of distress and anxiety, as well as social exclusion

**Materials & Methods:**

A prospective study of two comparable groups of children all suffering from atopic dermatitis: first group, which consisted of 210 children, had benefited from four educational sessions at the atopy school, and the second group, of 200 children, were followed at the standard pediatric dermatology consultation

**Results:**

The first group, who received four sessions of atopy school, there were 100 girls and 110 boys aged 4 months to 10 years

The average Scord initially was 24, correlating to a moderate AD; at the last session, it decreased to 10, indicating a mild AD. Scord improvement was noted in all patients and 75% of therapeutic goals were achieved

In this group, 28 children presented another atopy manifestation: 18 cases of asthma, 6 of allergic rhinitis, and 4 of allergic conjunctivitis

Psychological evaluation revealed a disturbance of psychoaffective and cognitive behavior in 24% of children, which was related in 5 cases to precarious situation and domestic violence.

Therapeutic objectives assessed were knowledge of AD, the ability to adapt care to skin lesions, pain management, and apprehension of applied treatments.

We have also adapted the questionnaire to our context: 89% of children went to the Moorish bath twice a month on average for exfoliating. We were able to reduce this social practice for some patients and end it for others, depending on social circumstances such as showers unavailability

Regarding the second group; average Scord was 26 at the first, then passed to 18,2 after four dermatological visits. An improvement of the Scord was noted in 95% of the cases, but the therapeutic objectives were only achieved in 40%

Psychological assessment of children in both groups combined showed a higher acceptance of the disease in the

first group

### **Discussion :**

Many studies concluded that therapeutic education give satisfaction and self-efficacy in the face of AD and improvement of life quality. Nowadays all current recommendations include it in the treatment of AD, but questions persist regarding the content of these interventions and the effectiveness of some compared to others

The largest study of 999 patients have validated the reduction of severity of AD for patients who benefited from Atopy School. A systematic review of 19 randomized trials concluded that current educational programs benefit pediatric patients with AD in terms of disease severity, but do not appear to significantly improve quality of life. This was due to the complexity of such factor, that depends on other criteria such as psychological distress burden and treatment compliance

Our experience is comparable to other reported series concerning a quicker Scorad improvement, more adherence to AD management, and better life quality

### **Conclusion:**

Atopy School doesn't replace standard follow up, but it helps patients comply by giving them a better understanding of atopic dermatitis, ensuring a better adherence to care

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**Abstract N°: 6200****Multi-system Langerhans Cell Histiocytosis with Scrofuloderma in a 2-year old Filipino male**MA. Patricia Gertrude Camille Ollero<sup>1</sup><sup>1</sup>East Avenue Medical Center, East Avenue Medical Center- Department of Dermatology, Quezon City, Philippines

**Introduction & Objectives:** Langerhans Cell Histiocytosis (LCH), a rare multisystem inflammatory neoplasia with an incidence of 2 to 9 cases per million per year with a male preponderance. The skin is the second most affected system, after the skeletal system, and usually manifest as eczematous dermatitis. Due to the various skin presentation, patients receive a delay in diagnosis. LCH patients can suffer from intercurrent infections, although most are mild, rarely, may suffer from severe infections such as Mycobacterium Tuberculosis. Scrofuloderma, a subtype of cutaneous tuberculosis is a common extrapulmonary form of tuberculosis. It is estimated that 1-2% of patients with pulmonary tuberculosis present with cutaneous lesions. Herein a case of a 2-year-old male with Multisystem LCH with concomitant pulmonary tuberculosis and Scrofuloderma.

**Materials & Methods:** We report a case of a 2-year-old male with enlarging erythematous plaque with ulceration and yellowish discharge, measuring 4 x 2 cm on the left supraclavicular area and multiple erythematous scaly papules with crusting over the trunk and scalp area recalcitrant to unrecalled medications. On review of systems, an enlarging bony mass on the left pre-auricular extending to the left post auricular area was noted. No palpable lymphadenopathy was noted. Histopathology was confirmatory of Langerhans Cell Histiocytosis showing numerous lymphohistiocytic infiltrates with reniform nuclei and marked epidermotrophism which stained positive to S-100 and CD1a. Chest radiographic showed dense apical infiltrates suggestive of PTB however GeneXpert was negative. Patient was started on first cycle of chemotherapy with intravenous vinblastine and oral prednisone with Tuberculosis regimen.

**Results:** Patient's condition improved after 6 cycles of chemotherapy together with 6 months anti- Kochs medications. There was noted complete resolution of all skin lesions with no recurrence of skin lesions.

**Conclusion:** LCH can present with Tuberculosis especially in areas where it is highly endemic. Concomitant disease presentation is attributed to Tuberculosis unmasking LCH features. As physicians, thinking about what more common and more likely diagnosis is essential to initiate early treatment for a favorable response.



**Abstract N°: 6201****Global clinicoepidemiological pattern of childhood vitiligo: a systematic review and meta-analysis**

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**Introduction & Objectives:**

Childhood vitiligo differs from adult vitiligo in many aspects. To the best of the authors' knowledge, there is no systematic review of different clinicoepidemiological patterns of vitiligo in children. This study aimed to review the characteristics of vitiligo among the paediatric population.

**Materials & Methods:**

In June 2022, a comprehensive search was conducted using MeSh-based keywords on online databases including PubMed, Scopus and Web of Sciences. The papers were assessed, and the eligible articles were selected. The selection of articles followed three distinct steps. The extracted clinicoepidemiological data were then imported into the STATA software for meta-analysis.

**Results:**

The meta-analysis of 17 studies with 4365 subjects yielded 2475 women (estimated=56.8%, 95% CI 54.45 to 59.22). The female-to-male ratio was determined to be 1.3:1. Meta-regression demonstrated a significant relationship between continents and gender ( $p=0.03$ ). The most prevalent types of non-segmental vitiligo were vulgaris (42.49%), focal (27.21%) and acrofacial (17.8%). The pooled ratio of non-segmental to segmental was 4.6:1. The highest and lowest ratios were found in Africa with one study (estimated=11.56%, 95% CI -0.98 to 24.10) and America with two studies (estimated=3.02%, 95% CI 1.54 to 4.50), respectively. Using meta-regression, the relationship between continents and vitiligo type was found to be insignificant ( $p=0.47$ ). Positive family history was recorded in 657 patients (estimated = 16.88%, 95% CI 13.37 to 20.39). Positive family history varied by country of study from 13.91% (Asia with 11 studies) to 27.01% (Europe with two studies) ( $p=0.11$ ). Kobner phenomena and leukotrichia were noted in 687 (25.47%) and 461 (18.52%) patients, respectively.

**Conclusion:**

The review indicated that childhood vitiligo is more prevalent in women. Non-segmental forms of childhood vitiligo were the most common, including vulgaris, focal and acrofacial. The clinicoepidemiological pattern of childhood vitiligo is variable in different geographic areas.

**Abstract N°: 6214****The efficiency of mTOR inhibitors in the treatment of cystic lymphangioma of the palate.**

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**Introduction & Objectives:**

Cystic lymphangioma is a lymphatic malformation frequently diagnosed in children, predominantly in the head and neck region, particularly in the posterior cervical triangle. Its localization in the palate remains exceptional.

We report a rare localization of cystic lymphangioma of the palate in a 6 year old girl treated with Sirolimus with a good clinical evolution.

**Materials & Methods:** Case report

A 6-year-old female patient with no medical history nor similar case in family, who presented since birth a polycystic hemorrhagic growth of 5 cm in diameter on the palate, progressing gradually in volume.

The biopsy showed numerous endothelial lined small lymphatic channels containing lymph tissue in favor of cystic lymphangioma.

The patient was treated with Sirolimus 1mg/day for six months with good improvement since the first month.

**Discussion :**

Cystic lymphangiomas are rare benign dysembryoplasias of the lymphoganglionic system, responsible of a tumor syndrome by gangiolymphatic proliferation.

Their localization is predominantly in the cervico-facial region and exceptionally reported in the palate. With a predilection in children under two years old.

The symptomatology depends on the size and location of the cystic formation. Their clinical revelation is early by the discovery of an isolated latero-cervical mass, the therapeutic management is based on surgical removal with wide margins to obtain a definitive cure.

This surgical removal is essentially recommended for limited forms or facial lymphangiomas invading the tongue and palate, and also for residual forms after sclerotherapy.

Sclerotherapy (steroid, bleomycin, bevacizumab) is an interesting alternative to surgery, but it is limited to the tongue, requiring airway protection by sedation or tracheostomy with heavy after-effects.

Sirolimus has recently been shown to be an effective treatment, via inhibition of the mTor pathway, involved in angiogenesis and cell metabolism, with very good tolerability and few side effects.

The dose of Sirolimus in children depends on several factors, including the child's age, weight and general health. In general, the recommended dose is 0.8 mg/m<sup>2</sup> per day in two doses.

**Conclusion :**

The particularity of our observation lies on the atypical localization on the palate as well as by its spectacular and

rapid evolution after treatment.

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**Abstract N°: 6219****Successful treatment of fungal ball-associated tinea capitis in a healthy infant: An unusual presentation**

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**Introduction & Objectives:**

Tinea capitis is the fungal infection of the scalp that most often develops with the involvement of the hair shaft and contiguous skin. The presentation of fungal ball in this disease is extremely rare. Herein, we describe a 7-month-old female infant presented with erythematous crusted skin rashes and a fungal ball, positive for *Epidermophyton floccosum* infection.

**Materials & Methods:**

A febrile patient with a large area of partial hair loss, scattered pustules on the occipital, and a painful, erythematous ball on the left retro-auricular area. The result of the scalp scraping examination of the hair loss area was positive for hyphae and spores.

**Results:**

She was successfully treated with oral terbinafine, intravenous clindamycin, ketoconazole cream, and Ketoconazole shampoo. The patient also underwent incision and drainage of the abscess.

**Conclusion:**

Tinea capitis most often affects early school children. It rarely affects neonates and infants. Its presentation with fungal ball in a healthy child is extremely rare. To our knowledge, this is the first case of fungal ball in a healthy infant with tinea capitis.



**Abstract N°: 6360****Linear IgA Bullous Dermatitis in a 7-year old Filipino Female**

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**Introduction & Objectives:**

Linear IgA disease (LAD) is an autoimmune mucocutaneous disease characterized by linear deposits of IgA at the basement membrane zone on immunopathology. In the pediatric population, it is known as chronic bullous disease of childhood (CBDC). Direct immunofluorescence (DIF) remains the gold standard for diagnosis in both adult and pediatric populations. Management of this relatively rare disease process varies throughout the literature. Dapsone is the most commonly used therapeutic agent, but its potential side effects such as hemolysis, agranulocytosis, and methemoglobinemia necessitate the use of other treatment modalities. Numerous other treatments have been reported to be effective in the treatment of LAD, including topical corticosteroids, tetracyclines, erythromycin, sulfonamides, nicotinamide, rituximab, omalizumab, methotrexate, cyclosporine and intravenous immunoglobulin.

Here we report a case of\* **Linear IgA Bullous Dermatitis in a 7-year old Filipino Female.**

**Materials & Methods:**

A 7 year old female presenting with a 19 day history of multiple round pruritic papules graded at 9/10 on the posterior lower legs which gradually increased in number and progressed to vesicles and bullae. . Diagnosis was **Linear IgA Bullous Dermatitis** based on clinical features, histopathology and direct immunofluorescence.

**Results:**

The patient was treated with Erythromycin 250mg/5mL syrup, 6mL every 6 hours for 7 days, Prednisone 20mg/5mLsyrup, Cetirizine 5mg/5mL, 5 mL OD for pruritus, Clobetasol Propionate 0.05% ointment on vesicles and bullae BID for 2 weeks, Betamethasone Valerate 0.1% ointment OD on affected areas on the neck and body folds for 2 weeks, Fusidate Sodium 2% Ointment on open wounds BID for 7 days and NSS Compress on eroded areas BID for 7 days with follow ups every 2 weeks. On the 6th follow up she was noted to have 100% improvement of lesions with only noted post-inflammatory hyperpigmentation, she was then started on Urea 10% Lotion BID on hyperpigmented areas and Niacinamide 300mg/paper tab once daily. On follow up after 1 month, the patient was well and about and still had 3 months of treatment for tuberculosis.

**Conclusion:**

LABD management has relied on DIF for diagnosis and on dapsone for treatment. However in our case report, given that the patient has a low GG6PD result, Dapsone could not be given. A literature search for articles published in the last 5 years has shown that other modalities, including rituximab, omalizumab, etanercept, IVIg, topical corticosteroids, among others have been used successfully in adult and pediatric patients with varying severity of disease. Patient factors such as age, medical comorbidities, and disease severity play a role in therapeutic selection. As in most patient cases in Dermatology where preliminary biopsy results are negative but clinical suspicion is high, a repeat biopsy and additional diagnostic studies should be considered, especially in LABD.



**Abstract N°: 6387****Scalp necrotic wound and hyperinflammatory shock related to COVID-19: Topical sucralfate as a promising topical agent**

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**Introduction & Objectives:**

Rarely, extravasation injuries can present with skin breakdown and consequently lead to serious complications such as skin necrosis, if left untreated. Extravasation injuries associated with vasoconstrictive agents can lead to ischemic necrosis. Critical conditions such as shock and underlying conditions of endothelial damage are potential predisposing factors for the development of extravasation injuries.

Herein, we describe a one-year-old child with epinephrine-induced dermal necrosis treated with topical sucralfate.

**Materials & Methods:**

A one-year-old previously healthy boy was admitted with a diagnosis of multisystem inflammatory syndrome in children (MIS-C) in association with COVID-19. During the three days of hospitalization, the patient developed an abrupt area of epinephrine-induced vasoconstriction followed by persistent ischemic dusky-red discoloration on the scalp for ten hours.

**Results:**

The patient was consulted by the pediatric surgeon and pediatric dermatologist who considered the patient epinephrine-induced dermal necrosis. Therapy including local wound care in addition to topical sucralfate cream was used at the site necrotic lesion. The 20th day of hospitalization revealed no cutaneous sequel.

**Conclusion:**

The current case highlights the specific promising clinical benefit of topical sucralfate for the treatment of dermal necrosis. In our experience, topical sucralfate can be considered a promising safe agent for the treatment of epinephrine-induced tissue necrosis.

Also, our case emphasizes the importance of awareness of early signs and symptoms of vasopressin-induced extravasation for prompt recognition of extravasation and safe use of vasopressin and specific antidotes for extravasation for prevention of more tissue loss.



**Abstract N°: 6398****A retrospective epidemiological study of skin diseases among pediatric population attending a tertiary care hospital**Kirti Jangid<sup>\*1</sup>, Swagata Tambe<sup>1</sup><sup>1</sup>SETH V.C GANDHI AND M.A. VORA MUNICIPAL GENERAL HOSPITAL, RAJAWADI,  
DERMATOLOGY/VENEREOLOGY/LEPROLOGY, MUMBAI, India**Introduction & Objectives:**

The incidence of skin diseases in children is influenced by hereditary, social, and environmental factors. The disease pattern differs in a given population by different ecological factors. The aim of this study is to give an overview of the statistical study of different dermatologic diseases in children in a tertiary care hospital.

**Materials & Methods:**

We reviewed epidemiologic data of 3130 patients, aged 0-12 years, who were referred to the outpatient clinic of a tertiary care hospital between January 2021 and December 2021. Demographic data and the frequency of the various diagnoses in various age groups were studied.

**Results:**

3130 children were included in the study (1507 females and 1623 males). The most frequent diagnoses were: Infections (47.51%), Atopic dermatitis (5.85%), Vitiligo (1.3%), Alopecia areata (1.08%), Urticaria (1.18 %), Seborrheic dermatitis (0.8%), Nevi, Vascular malformations and Hemangiomas (0.42%), Psoriasis (0.32%).

Scabies (24.86%) was the most prevalent infection and it was the most common cause of skin disorders in the present study. Incidence rate of scabies, found in other reports, ranges from 5.1% to 22.4%. Fungal infections were the second in frequency among infectious disorders comprising 10.61%. These findings are supported by other studies like Patel *et al.* (7.81%), Thappa (8.49%), Sharma and Ben Saif and Al Shehab. Tinea corporis was the most common fungal infection

In this study, molluscum contagiosum (3.04%) was the most common of all viral infections. Leprosy was diagnosed in 02 patients.

Single child was diagnosed with scrofuloderma.

The incidence of atopic dermatitis (5.85%) was similar with other studies where they found rates ranging from 3% to 28%.

Incidence of Vitiligo in our study was found to be 1.30%

Psoriasis had a frequency of 0.32% in this study. Nearly similar observations were reported in Rao and assoc. and Sardana K studies.

Urticaria affects about 1.18% of the study population which correlates with other studies.

**Conclusion:**

Skin diseases are a major health problem in the pediatric age group and are associated with significant morbidity.

This study shows that infections and infestation disorders were more common in the pediatric age group. Several problems including lack of education, social backwardness, lack of sanitation, excess pollution and overcrowding contribute to more incidence of infectious disorders in developing countries. This can be controlled by public awareness, proper sanitation and by training the dermatologists, pediatricians and general practitioners about the management of common skin disorders. But many non-infectious disorders that need a dermatologist's opinion should be referred to them. Due to the wide variety, burden and public health problem of skin diseases in children, more dermatologists should be trained in pediatric dermatology subspecialty. Further this data can be useful in planning health care programs for children.

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**Abstract N°: 6419****The problem of self-injury in the differential diagnosis of skin lesions in children**

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**Introduction & Objectives:**

Self-injury is described as socially unacceptable body damage causing bleeding, bruising or pain, undertaken to reduce psychological discomfort. They can be a symptom of emotional, developmental and personality disorders and can be predictors of future suicide attempts. They are defined in the DSM V classification as nonsuicidal self-injury (NSSI) without suicidal tendencies. Epidemiological data show that 15-40% of adolescents and young adults are affected, with symptoms most commonly 12-15 years of age.

**Materials & Methods:**

We present a description of a series of 4 cases of children with different clinical manifestations of skin self-injury.

**Results:**

The first case involves an 11-year-old girl with a focal area of hair loss present for about 2 months, accompanied by severe pruritus. Trichoscopic examination showed flame hairs, V-sign, hook hairs, and tulip hairs.

The second case was a 15-year-old girl with persistent skin lesions of the backs of her hands for about 3 months, accompanied by pain and limitation of finger mobility, and weakness of hand muscle strength. On physical examination, linear stacked rows of papules with shallow erosions on the surface were visualized. In the course of the diagnostic procedure, the girl admitted to having problems at school, which prompted her to perform skin punctures.

Another case involves a 17-year-old girl with lesions of a swollen and painful nature on the backs of her hands. No abnormalities were found in the diagnostic tests and after applying occlusive dressings for 1 week, a complete remission was achieved. The patient admitted to inducing the lesions. The last case involves a 12-year-old girl with symmetrically arranged, circular erythematous-edematous lesions with a hemorrhagic component occurring in response to stressful situations at school on the anterior surfaces of the lower extremities.

**Conclusion:**

Skin self-injury is a diagnostic and therapeutic challenge. The presentation of the different clinical picture of skin self-injury in children aims to draw attention to this problem and emphasizes the need for an interdisciplinary approach in both diagnosis and treatment

**Abstract N°: 6422**
**Unilateral DLE in a pediatric female patient: an unusual presentation**

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**Introduction & Objectives:** DLE is a chronic type of CLE which usually presents as symmetrically distributed erythematous scaly plaques with or no atrophy, scarring, and hypo and/or hyperpigmentation. Photo-exposed areas such as the face, scalp, ears, and extensor aspect of upper limbs are commonly involved. Careful evaluation and treatment is necessary to prevent scarring and systemic involvement. Less than 5% of patients develop the disease before the age of 16 and 2% before the age of 10. Usually, CCLE lesions are multiple and present bilaterally. Strictly unilateral but multifocal distribution of discoid rash over the face is very unusual.

We report an unusual case of a pediatric unilateral DLE.

**Materials & Methods:**

Physical examination, histopathological examination, immunohistochemistry of skin biopsy from the lesional skin performed for diagnostic purpose.

**Results:**

A 3-year-old girl presented with a rash involving the left side of her face for the last 6 months. The lesions began as asymptomatic red-purple coloured patches on the left side of the face appeared after sun exposure during their last travel. The first elements were distributed on the left cheek and behind the left ear. She was diagnosed with acquired PWS. Over the next 6 months lesions were increased in their sizes and new ones were appeared on the forehead and scalp strictly distributed at the left side. Parents were worried and presented the child to our department. On physical examination multiple patches, from 0.5 to 5 cm in diameter over the left cheek, forehead, chin and behind the left ear were noted. The patches were erythematous – violaceous, not indurated, but there were TAE – s over them under dermatoscopy. There was no scaling, atrophy. Mucous membranes and nails were not involved. There were no joint pains or any associated systemic complaints. We considered following differential diagnoses: DLE, morphea, acquired PWS or other CM. CBC, serum biochemistry, liver and renal function tests, serum complement levels, routine urinalysis were normal. CRP was elevated to 38 mg/l. ANA titer was negative, DNA was elevated to 45 U/ml.

Histopathological examination of skin biopsy from the lesional skin showed: epidermal hyperorthokeratosis with superficial follicular plugging and atrophic changes, peri-appendageal lymphocytic infiltration in superficial and deep dermal layers, focal basal cell vacuolation on DEJ, proliferation of perifollicular fibroblasts, edema in papillary dermis. Special staining with PAS revealed deposition of mucin in the dermis. The results of direct immunofluorescence: CD3 and CD 123 clones of T lymphocytes. Diagnosis of DLE was performed.

The child was advised strict photoprotection and topical Tacrolimus 0.1% ointment.

After an ophthalmology clearance, she was started oral hydroxchloroquine 200 mg every 3 days (5 mg/kg/d). Prednisolone in doses of 1 to 3 mg/kg/day tapered over a 2-week period was prescribed to rapidly control the disease. The lesions subsided after 2 months. But after 4 months of treatment she presented with depigmented

patches on the same places of the last lesions and focal hair greying over the scalp lesions. We considered vitiligo as coexisting autoimmune condition or

postinflammatory hypopigmentation, and decided to follow up with continuing topical tacrolimus.

**Conclusion:**

Our case is unusual as the patient's age is not common for DLE, and the lesions were strictly unilaterally distributed.

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**Abstract N°: 6428****Herpes zoster in a 4-month-old immunocompetent infant**

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**Introduction & Objectives:****Materials & Methods:****Results:**

Herpes zoster (HZ) is typically observed in adults and it is fairly uncommon in the pediatric population, with an incidence ranging from 0.2 to 0.74 cases per 1000 person/year.

Herein, we report an immunocompetent 4-month-old infant with herpes zoster involving third and fourth right lumbar dermatomes (L3 and L4), who developed HZ after intrauterine exposure to Varicella Zoster Virus (VZV).

An otherwise healthy 4-month-old male infant presented with a 1-week history of vesicular eruptions on the right thigh. The general health of the infant was unaffected and all routine hematological parameters were within reference range.

Dermatological examination revealed multiple, tense, grouped fluid-filled vesicles on an erythematous base arranged in two clusters, affecting the femoral region of the right lower extremity, corresponding to the right L3 and L4 dermatomes. Findings from a Tzanck smear were positive for multinucleated giant cells.

A clinical diagnosis of HZ was made. The patient was treated with 200 mg of Acyclovir orally 4 times a day for 7 days. Topical fusidic acid was used to prevent secondary bacterial infection.

A complete resolution of the lesions was observed without any sequelae.

The patient had no history of experiencing symptomatic varicella but was exposed to the virus (VZV) in utero during the eighth month of the pregnancy, confirming the occurrence of an intrauterine infection without visible embryopathy.

The clinical course of HZ generally is less severe in children than in adults. The infection usually is mildly symptomatic with a benign clinical course and complete resolution in 1 to 3 weeks without sequelae, as observed in our patient.

The purpose of this abstract is to emphasize the rarity of this condition in infancy.

**Conclusion:**