COLOR DOPPLER ULTRASOUND: EXPERIENCE OF ITS USEFULNESS IN PEDIATRIC PATIENTS

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**Introduction:** Color Doppler ultrasound (CDU) has been incorporated for the diagnosis, follow-up, and surgical planning of skin diseases. It is a widely available imaging method that allows evaluation of different skin pathologies. In pediatric patients it also has the advantage of being painless and not using ionizing radiation.

**Clinical cases:** Five pediatric patients studied with CDU at Hospital Clinic in Barcelona from June to November 2016 are reported. Patient 1: Two-year-old girl with anomic flanges in both hands. CDU revealed decreased thickness of the dermis and hypodermis. With these method the patient could be classified as grade 2. Patient 2: Ten-day-old girl with nodule on the right leg. CDU showed hypoechogenic demal 13.6 × 6.1 mm lesion. Doppler ultrasound shows prominent blood flow in the periphery of the lesion. The histological diagnosis was of mastocytoma. CDU was useful in characterizing the lesion and for follow-up. Patient 3: Eight-year-old girl with a venulocutaneous bypass valve, after surgery for apneicobilastroblastoma. Complains about pain and erythema on the scalp. CDU demonstrated a valve in subcutaneous tissue without extrusion. Color Doppler demonstrates increased vascularity. CDU 10 days after: showed decreased vascularity with Doppler mode. DCU was useful to rule out valve migration. Patient 4: Seventeen-year-old girl with Goltz syndrome. CDU showed thickness of the dermis 0.4 mm in affected area and 1.4 mm in healthy contralateral skin. DCU determined the extent of dermal hypoplasia and served as a guide for cutaneous biopsy. Patient 5: Eleven-month-old girl with a 1.5 cm diameter scalp mass diagnosed as an asperatic nodule of the scalp. CDU showed moderate hypoechogenic 10.3 × 6.4 mm lesion. Doppler mode revealed increased intralosseal flow. CDU was useful for follow up. **Conclusion:** These cases demonstrate the utility of CDU in pediatric pathologies. CDU allows us to describe pathologies not previously reported.

**P 002**

ULTRASOUND FINDINGS IN IDIOPATHIC ASEPTIC FACIAL GRANULOMA

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**Introduction:** Idiopathic facial aseptic granuloma (IFAG) consists of a persistent asymptomatic nodule most commonly located on the cheeks and eyelids in young children. Diagnosis is primarily clinical and optimal management should be made without scars. At an initial stage, IFAG consists of a well-defined, hypoechoic solid mass with ill-defined borders that later in time vascularization was documented. At the follow-up, and surgical planning of skin diseases. It is a widely available imaging method that allows evaluation of different skin pathologies. In pediatric patients it also has the advantage of being painless and not using ionizing radiation.

**Clinical cases:** Five pediatric patients studied with CDU at Hospital Clinic in Barcelona from June to November 2016 are reported. Patient 1: Two-year-old girl with anomic flanges in both hands. CDU revealed decreased thickness of the dermis and hypodermis. With these method the patient could be classified as grade 2. Patient 2: Ten-day-old girl with nodule on the right leg. CDU showed hypoechogenic demal 13.6 × 6.1 mm lesion. Doppler ultrasound shows prominent blood flow in the periphery of the lesion. The histological diagnosis was of mastocytoma. CDU was useful in characterizing the lesion and for follow-up. Patient 3: Eight-year-old girl with a venulocutaneous bypass valve, after surgery for apneicobilastroblastoma. Complains about pain and erythema on the scalp. CDU demonstrated a valve in subcutaneous tissue without extrusion. Color Doppler demonstrates increased vascularity. CDU 10 days after: showed decreased vascularity with Doppler mode. DCU was useful to rule out valve migration. Patient 4: Seventeen-year-old girl with Goltz syndrome. CDU showed thickness of the dermis 0.4 mm in affected area and 1.4 mm in healthy contralateral skin. DCU determined the extent of dermal hypoplasia and served as a guide for cutaneous biopsy. Patient 5: Eleven-month-old girl with a 1.5 cm diameter scalp mass diagnosed as an asperatic nodule of the scalp. CDU showed moderate hypoechogenic 10.3 × 6.4 mm lesion. Doppler mode revealed increased intralosseal flow. CDU was useful for follow up. **Conclusion:** These cases demonstrate the utility of CDU in pediatric pathologies. CDU allows us to describe pathologies not previously reported.

**P 003**

HALO SCALP RING. ULTRASONOGRAPHIC FINDINGS

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Halo scalp ring is an uncommon type of alopecia that develops after caput succedaneum. It is usually mild and autosolutive, but some cases of cicatricial alopecia have been described. We report the case of a five-month-old girl that presented with an annular patch of alopecia on the scalp that was first noted several weeks after birth. The baby was born in good condition and was of a big caput succedaneum on the parietal area of her scalp. The swelling disappeared during the next few days, but an area of alopecia developed around the edge, almost encircling the head. Ultrasound is a noninvasive, harmless method that can help make a diagnosis and rule out other alopecias. We found a thinned scalp, mostly due to a diminished, structureless fat tissue. There were no follicles in the alopecia patch, and the Color Doppler Ultrasound demonstrated absence of inflammation.

**P 004**

EPIDEMIOLOGY AND OUTCOME ANALYSIS OF THREE HUNDRED AND NINETY-FIVE CASES OF PEDIATRIC DERMATOLOGIC SURGERY

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Surgical treatment of pediatric patients with skin diseases has been relatively rare compared to surgical treatment of adults. The purpose of this study was to retrospectively review all cases of dermatological surgery to establish epidemiological patterns and final outcomes for pediatric dermatological surgery in the Republic of Korea. New patients under 15 years of age who visited the Department of Dermatology Wonkwang University hospital in the Republic of Korea from 2012 to 2016 were reviewed through clinical and surgical records. There were three hundred and ninety-five cases of pediatric dermatological surgery. There was a male to female ratio 4:3.1. The three most common skin diseases were nevus sebaceous (30.4%), epidermal cyst (13.9%), and pincer nail (12.6%). All patients were successfully treated and satisfactory aesthetic results were achieved in every case. Few patients received general anesthesia. Most patients were treated in the dermatology department without the need for additional care. There were no major complications, such as death or hospitalization. Minor complications, such as surgical site infection and loss of nerve function, were not reported. In conclusion, compared to previously reported dermatological surgeries on adults, the distribution of pediatric skin diseases receiving surgery was different, and pediatric dermatologic surgery was safer and more effective.

**P 005**

SONOGRAPHIC FINDINGS IN SUBCUTANEOUS GRANULOMA ANNULARE

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**Introduction:** Subcutaneous granuloma annulare (SGA) is a benign inflammatory disorder that primarily affects healthy children. It manifests as a painless nonmobile mass, most commonly located in the lower extremities. The diagnosis of SGA can be difficult especially for those clinicians unfamiliar with this entity and concerned about the differential diagnosis of malignant processes. We present a case of SGA in which ultrasound findings allowed us to make the diagnosis.

**Case report:** An anotherwise healthy 4-year-old girl presented with a rapidly growing painless right prebital lump which had been present for a year. She had no history of trauma. On physical examination, a 4 × 2-cm solid, nonmobile, subcutaneous mass with normal overlying skin was palpated. No other abnormalities were noted. Ultrasound revealed an ill-defined hypoechogenic solid mass in the subcutaneous fat measuring 1.85 × 0.42 cm. A slightly hypoechogenic rim was observed but intralosseal or
Peripheral Doppler signals were not detected. The typical anatomical distribution, radiologic imaging, and ultrasound findings suggested the diagnosis of SGA. Magnetic resonance imaging (MRI) showed an ill-defined hypointense mass in the subcutaneous tissue that abutted the superficial fascial planes without extending into the underlying muscle planes or bone. The diagnosis of a SGA was confirmed by histopathological examination.

**Discussion:** Diagnosis of SGA can be a challenge because the history of rapid growth of a firm subcutaneous mass can mimic a malignant lesion. Although incisional biopsy is the safe way to confirm the diagnosis, ultrasound examination can be helpful to detect it and avoid unnecessarily invasive procedures. A rapidly growing, solitary, nontender, subcutaneous lesion located on the pretilial area in a healthy infant that otopgraphically presents as an ill-defined hypoechoic lesion with a hyperechogenic rim and without prominent vascularization should make us suspect a SGA.

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**P 006**

A CASE OF EPIDERMOLYTIC ICHTHYOSIS WITH DE NOVO KRT1 GENE MUTATION

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Epidermolytic ichthyosis, also known as bullous congenital ichthyosiform erythroderma (BCIE), is a rare autosomal dominant disorder with an incidence of 1 in 300,000. Most cases result from mutations in keratin 1 (KRT1) or 10 (KRT10) genes. Clinical features comprise dry skin with fragility, blistering, and erosions. Palmo-plantar keratoderm (PPK) may also occur, and is more commonly seen with KRT17 mutations. We report the case of a Caucasian male infant who presented at birth with small non-inflammatory bullae and erosions on his trunk and limbs. Marked dryness of skin was noted with areas of scaling and desquamation. His nails were unremarkable and there was no oral mucosal involvement or PPK. He was born to non-consanguineous parents who were phenotypically normal. Lesional skin biopsies were sent to The National Diagnostic EB laboratory at St. John’s Institute of Dermatology in London for analysis. Light microscopy showed evidence of extensive cytoplastic change in the mid- and upper epidermis. Direct sequencing of KRT1 and KRT10 genes was then performed, and a heterozygous missense mutation p.Leu187Pro was identified in exon 1 of KRT1. This mutation has previously been reported as pathogenic for BCIE. Sequence analysis of both parents’ DNA revealed wild-type KRT17 only, establishing this as a de novo mutation. Interestingly, a heterozygous deletion, c.1468_1478del12 in exon 7 of KRT1 was also identified. The significance of the KRT1/10-frame-shift mutation is uncertain. However, in this case the KRT10/10-frame-shift mutation combined with the absence of PPK highlights the importance of genotype-phenotype correlation in patients with epidermolytic ichthyosis.

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**P 007**

AN UNUSUAL CAUSE OF FACIAL ULCERATION IN A FOUR YEAR OLD

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A young child presented with persistent centrofacial ulceration dating back to infancy. It was reported by his parents that he would recurrently rub and pick his facial skin and that the ulceration was self-inflicted. Of note, at birth he sustained a fractured clavicle secondary to shoulder dystocia with apparently minimal discomfort. The features are consistent with the recently described entity mid face toddler excoriations (MITES). Mutations in the pain insensitivity genes PRDM12 and SCN11A have been rarely reported in several cases presenting with similar facial ulceration.

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**P 008**

GOLTZ SYNDROME: REPORT OF TWO CASES AND OVERVIEW OF PORCN MUTATIONS

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Goltz Syndrome or Focal Dermal Hypoplasia (FDH) is an X-linked dominant syndrome, caused by mutations of PORCN gene, with ectoderm and mesoderm abnormalities. The reported FDH cases show a spectrum of highly variable clinical features that can be grouped into skin, skeletal, ophthalmologic, dental/oral, and gynecologic abnormalities. Common skin findings include hypoplastic and atrophic skin lesions with telangectasias, hypo/hyperpigmented skin lesions, dermal fat nodules, patchy alopecia with hair shaft abnormalities, nail ridging, and micronychia.

PORCN gene is a regulator of Wnt signaling pathway which is essential for multiple processes during embryogenesis. Strong skewing of X inactivation has been shown in familial cases which could be crucial for viability of offspring of FDH patients during the fetal period. The phenotype variability of FDH has been attributed to X inactivation skewing, postzygotic mutations resulting in somatic mosaicism, and multiple isoforms being asymmetrically affected by PORCN mutations.

Setleis Syndrome or Focal Facial Dermal Dysplasia (FDDF) is an autosomal recessive inherited disorder caused by homozygous mutations in the TWIST2 gene. Bitemporal skin lesions commonly occur along with variable facial findings including thin and puckered periorbital skin, distichiasis and/or absent eyelashes, palatal abnormalities, a flat nasal bridge with a broad nasal tip, large lips, and redundant facial skin. Two female FDH patients, sharing common clinical features but also showing phenotypic variability with two different de novo PORCN mutations will be discussed. Furthermore, a third case mimicking Setleis Syndrome with overlapping features of FDH with no pathogenic changes identified in the PORCN and the TWIST2 genes will be examined for evidence of genetic etiopathogenesis.

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**P 009**

GOLTZ SYNDROME: CASE REPORT OF FOCAL DERMAL HYPOPLASIA

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Goltz syndrome (also known as Focal Dermal Hypoplasia) is a rare multisystem disorder, involving all three germ cell layers. The disease is inherited in X-linked dominant fashion with mutations of the PORCN gene. The majority of these cases are sporadic, mainly due to postzygotic somatic mutations. We present a case of this rare disease.

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**P 010**

TWO CASES OF INCONTINENZTA PIGMENTI IN NEWBORNS WITH CONSEQUENT VISUAL ABNORMALITIES

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A female patient presented to pediatric dermatology at the age of 2 weeks with an erythematous vesicular rash on the left elbow and knee,patchy pigmentation on the trunk and limbs, in addition to a vesicular rash on her vulva. Her mother had one previous miscarriage. She has no family history of incontinentia pigmenti (IP). Histology revealed eosinophilic spongiosis in keeping with IP. Genetic screening showed a change within the NEMO gene. Her brother had negative prenatal screening. Ophthalmological examinations were regularly performed from the age of 14 weeks. At 15 months peripheral retinal telangectasias was noted. Examination under anaesthesia with fluorescein angiogram demonstrated bilateral retinal ischemia which was treated with a laser. Delayed dentition was noted.

A second female patient presented to aediatric dermatology at the age of 6 weeks with a linear pustular rash on her arms and legs. IP was confirmed on history. Her mother had two miscarriages. She has no family history of IP. Genetic screening showed deletion of the NEMO gene. Her mother was not a carrier of the gene. She developed recurrent seizures at the age of two months. Ophthalmological examination at the age of nine weeks showed buphthalmos and glaucoma in the left eye. B scan showed findings consistent with bilateral retinal detachment. At the age of 7 months right eye microphthalmia was noted, and an expansion prosthesis was inserted at the age of 3 years. She has a developmental delay and delayed dentition.

IP is an X-linked dominant disorder caused by mutations in the NEMO gene. It is usually lethal in males prenatally. It presents neonatally in females with skin abnormalities in 4 stages: erythematous vesicular rash, verrucous plaques, hyperpigmented swirls, and hypopigmentation. The significant accompanying medical problems are blindness and neurotological disturbances. Patients should see an ophthalmologist as soon as possible after birth, and at regular intervals thereafter, to allow early treatment.

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**P 011**

DYSTROPHIC EPIDERMOLYTIC BULLOSA PRURIGINOSA WITH ALBOPAPULOID LESIONS

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**Introduction:** Epidermolysis bullosa pruriginosa (EBP) is a rare variant of dystrophic epidermolysis bullosa (DEB) caused by mutations in the COL7A1 gene which encodes type VII collagen. Fewer than 100 patients were reported in the literature. We report a new case with albopapuloid lesions.
Introduction: Sebaceous nevus syndrome (SNS) is a rare congenital disorder that involves structures of ectodermal and mesodermal origin, typically characterized by the triad of a sebaceous nevus (SN), seizures, and mental retardation. Reports of intraoral involvement associated with this syndrome are scarce.

Case: A 3 year-old-girl presented a congenital linear sebaceous nevus on the left half of the chin extending into the inferior lip and crossing the midline. At birth neurological and ophthalmological examination ruled out systemic involvement. The patient had a normal psychomotor development and no history of seizures. She developed a lesion located on the uvula histopathologically consistent with squamous papilloma.

During follow-up it was observed that the left inferior dental pieces were absent, while the rest of the dental development was within normal parameters. A radiographic analysis revealed the presence of dental pieces, however follow-up did not allow any signs of correct teeth eruption. In the setting of a linear SN associated with intraoral abnormalities, the diagnosis of SNS was established.

Discussion: Intraoral manifestations in SNS are scarcely reported in medical literature. On account of the potential skeletal abnormalities associated with SNS, intraoral examination via an appropriate specialist should be part of the screening for SN located on the head and neck.

Conclusion: We present an unusual case of SNS and raise awareness of associated intraoral involvement.

P 015 NEONATAL ICHTHYOSIS AS PRESENTING SIGN OF TRICHOHYDROSTROPHY M. Campos-Dominguez*, P.T. Vlas Boas1, A. Ruedas Martínez*, A. Sánchez Herrero*, A. Matos Mayou*, I. Marsurias Rios*, V. Pastra Bianco*, M.E. Seoane Reula*, E. Barredo Valderrama*, R. Suárez Fernández* 1Hospital General Universitario Gregorio Marañón, Dermatology, Madrid, Spain, 2Hospital General Universitario Gregorio Marañón, Psychiatry, Madrid, Spain, 3Hospital General Universitario Gregorio Marañón, Pathology, Madrid, Spain, 4Hospital General Universitario Gregorio Marañón, Allergy, Madrid, Spain and 5Hospital General Universitario Gregorio Marañón, Pediatric Neurology, Madrid, Spain

Introduction: Neonatal ichthyosis represents a diagnostic challenge because of the clinical overlap between several entities. We present a case in which an early diagnosis of trichohydrotrophy was made based on polarized microscopy examination of the hair.

Clinical case: A male infant was born at 36 weeks of gestation with a birth weight of 2650 grams. Shortly after birth, extensive desquamation and hair alterations were noticeable. Upon physical examination, erythroderma, palmoplantar desquamation, and scaling on his trunk were seen. Hypotrichosis with dry, brittle hair and high anterior hairline could be observed. He had a peculiar phenotype, with micrognathia, prominent occiput, anteverted earlobes, and fifth finger clinodactyly. Upon neurological examination, he was hypertonic, showing en bloc movements, including hyperreflexia and flexor attitude of the limbs. He suffered from convulsions and oral myoclonus from Enterobacter aerogenes for the first and second weeks of life. Complete blood count showed anemia (Hb 8.4 g/dl) and neutropenia (200/mc). Abdominal ultrasound was diagnostic for left pyelectasis. Polarized microscopy examination of the hair showed a tiger tail pattern with alternating dark and light bands. This pattern is pathognomonic for trichohydrotrophy. This diagnosis was confirmed by a genetic study, which demonstrated two heterozygotic mutations in the ERC2/3 gene.

Discussion: Icthysis in a neonate should prompt a search for other signs to improve diagnostic accuracy. If hypotrichosis is seen, a microscopic examination of the hair is fundamental to make a differential diagnosis between several entities. Trichohydrotrophy is an autosomal recessive disorder due to the impairment of DNA-reparing proteins. Abnormal, low-sulphur hair gives its name to the disease. Neurological alterations, short stature,
and ocular problems are frequent. From the dermatological point of view they can present with ichthyosis, photosensitivity, and nail dystrophy. Immunological alterations give rise to an increase of infections and a diminished life expectancy.

**P 016**

**THREE CASES OF FOCAL DERMAL HYPOPLASIA: A BROAD SPECTRUM OF THE SAME DISEASE**

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**Background:** Focal dermal hypoplasia (FDH), also known as Goltz’s syndrome, is a rare genetic X-linked dominant disease. Only about 250 cases are reported in the literature. It is due to a mutation in the PORCN gene, which plays a key role in embryonic tissue development. The phenotype is highly variable. Cutaneous anomalies include asymmetric atrophic hyper- or hypopigmented, telangectatic linear streaks in Blashko’s lines, soft red-yellow nodules in Blashko’s lines, ulcers, and papilloma. Hair can be sparse and brittle, nails are thin. Other organs may be involved, especially skeletal abnormalities, in particular digital anomalies, osteopatia striata, ocular anomalies, facial dysmorphism, and cardiac malformations. Furthermore the association of intrauterine growth retardation, thoraco-abdominal wall defect or diaphragmatic hernia is a triad frequently present in the FDH.

**Cases report:** Here we present three de novo cases with clinical evidence of Goltz’s syndrome, but with very different phenotypes. The first one is a three-year-old girl showing a severe presentation of the disease with almost all of the above characteristics (multiple focal dermal hypoplasia, papilloma, fat hemiation, “lobster-claw”, coloboma, facial dysmorphism, cardiac malformations and growth retardation). Genetic analysis shows a PORCN-mutation R124X.

The second case is a 2-day-old girl presenting a mostly cutaneous version of the disease with multifocal reticulated dermal hypoplasia along the Blashko’s lines, fat hemiation, syn-dactyly and growth retardation. Cerebral, ophthalmic, cardiac and abdominal examinations were normal.

Finally, the third case is a 10-year-old girl with a limited segmental presentation of reticulated dermal hypoplasia on her left leg without any other involvement.

**Conclusion:** Goltz syndrome is a typical example of functional mosaicism with a random inactivation of X chromosome in each cell resulting in a high variable phenotypic expression as seen in our patients. The segmental form of Goltz syndrome results from a late post-zygotic PORCN mutation.

**P 017**

**CLINICAL AND HISTOPATHOLOGICAL FINDINGS OF KERATINOPATHIC ICHTHYOSIS IN OUR HOSPITAL**

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**Background:** Structural stabilization of the upper layer epidermis is provided by cytokeratin 10 and 1 (KRT1 and KRT10, respectively). Mutation in either of them will produce skin fragility with a clinical spectrum depending on the moment when mutation has taken place. If mutation is in a germinal line (a pre-zygotic period) the newborn patient will present epidermolysis bullosa neonatal with blistering and repigmentation of the skin at birth. If mutation occurs in a post-zygotic period, a mosaic form following Blaschko lines will happen, and the patient presents as epidermolysis epidermal nevus (EEN).

**Material and methods:** We present a retrospective series of keratinopathic ichthyosis in our center since January 1999 until now. Data on phenotypical expression, histological characteristics, and management were collected.

**Results:** We examined 7 patients, 6 with EEN and one with epidermolytic ichthyosis (pending genetic confirmation). In 46 patients with EEN, lesions appeared during the first months of life, and for the remaining patients, lesions were congenital. None of them have familial antecedents of ichthyosis. In the E1 case the patient was born with blisters, erythroderma, and denuded skin with posterior progression to normal appearing skin with mild cobblestone-grease flexural hyperkeratosis. Any case has developed palmoplantar involvement. Case 5 presented a bilateral and extensive systematized epidermolytic epidermal nevus (SEEN) resembling Darier-Macklin Ichthyosis (ichthyosis hystrix).

Biopsies were performed in all cases. All cases showed epidermolytic hyperkeratosis, the characteristic histopathological hallmark of keratinopathic ichthyosis. As in previous studies, we found that mosaic variants have focal changes while in the E1 case, changes are continuous.

All patients received basic skin treatments. In one localized case electro surgery treatment was done with good response. Other cases had poor responses to topical clindamycin 10 mg/g, benzoyl peroxide 50 mg/g. A SEEEN calciotropic serum was prescribed with no response. Neither oral or topical retinoid have been used in any of the cases.

**P 018**

**COLLODION BABY: A RETROSPECTIVE STUDY OF 10 PATIENTS**

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**Background:** The term collodion baby describes the phenotype of a newborn with a parchment-like membrane covering its body surface. It may be the initial presentation of a number of different conditions, primarily disorders of conformation.

**Objective:** To characterize clinical features, complications, treatment, and final outcomes in a cohort of babies born with a collodion membrane.

**Methods:** We conducted a retrospective study of newborns clinically diagnosed with colloidion baby in the Balearic Islands over the last 25 years. Data on demographics, family history, membrane shedding, clinical features, complications, treatment, final outcome, and molecular studies were collected.

**Results:** Ten patients were included, 6 males and 4 females. The mean follow-up time was 7.9 years (range: 1 month - 22 years). Three had a positive family history. Consanguinity was found in only one patient. All of them were managed in the intensive care unit, placed in a humidified incubator, closely monitored, and treated with vaseline. Sudamina was present in two patients (in both vaseline was applied more than twice a day), and neurodevelopmental abnormality were found in one. None of our patients presented infection, sepsis or death. The final outcomes were nonsyndromic autosomal recessive congenital ichthyosis in 5 (50%), partially self-healing collodion baby in 1 (10%), self-healing collodion baby in 3 (30%), and trichohydrotichosis in 1 (10%) of them. We found a wide spectrum of affected genes in our patients, including mutations in TGM1, ABCA12, ALOX12B, NIPAL4, ALOXE3.

**Conclusion:** It is not possible to predict the eventual phenotype of collodion babies based on the initial appearance. The severely compromised epidermal barrier represents the greatest challenge during the newborn period and they should be managed in specialized neonatal care units.

**P 019**

**BLOCH-SULZBERGER SYNDROME: EARLY CUTANEOUS FEATURES, KEY FOR DIAGNOSIS**


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**Introduction:** Incontinentia pigmenti (IP), or Bloch-Sulzberger syndrome, is an uncommon X-linked dominant inheritance ectodermal dysplasia which affects females and is usually lethal in males at birth. It is characterized by cutaneous, dental, hair, ocular, and central nervous system abnormalities.

**Case report:** A baby girl born after an uncomplicated pregnancy presented with blisters on limbs and trunk that were not responsive to antibiotic therapy, and negative laboratory investigations. On the third day of life she presented a unique generalized tonic event that ceased with phenobarbital administration. EEG was unspecified.

The histopathological examination of the skin biopsy showed intradermal vesicles and eosinophilic spongiosis compatible with incontinentia pigmenti (IP). Gene analysis demonstrated exon 4: 10 deletion of the IKBKG (Inhibitor of kappa light polypeptide gene enhancer in B-cells, kinase gamma gene) on the X-chromosome, confirming the diagnosis.

Brain imaging and ophthalmologic examination was normal. In subsequent consultations the blisters gradually resolved to brownish hyperpigmentation and currently, ten years later, we can also observe vertex and eyelash alopecia and teeth growth anomalies with no accompanying ocular or neurologic findings.

**Discussion:** IP is an inherited genodermatosis in most cases caused by mutations in the IKBKG gene, involved in the regulation of inflammatory immune and apoptotic pathways. Clinical manifestations range from mild skin and dental involvement to severe leukodystrophic and ophthalmologic manifestations. Cutaneous findings are usually the first observed sign of IP and can be classified into four stages: vesciculobullous, verrucous, hyperpigmented, and atrophic. Other common manifestations include vertex alopecia and partial anodontia. Agenesis of eyebrows and eyelashes, a feature of the present case, are atypical.

We report a case of IP, a rare multisystem genodermatosis that should be familiar to all dermatologists as its cutaneous manifestations are characteristic and necessary for its diagnosis.
KERATINOPATHIC ICHTHYOSIS MINOR VARIANTS: CLINICAL AND GENETIC SPECTRUM

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Inherited ichthyoses are a group of genetic disorders characterized by generalized dry skin, scaling and hyperkeratosis, and often associated with erythroderma. These manifestations are due to mutations in genes mostly involved in skin barrier formation. Keratino-pathic ichthyosis is proposed as an umbrella term for nonsyndromic ichthyoses caused by mutations in keratin genes, including major and minor variants. Ichthyosis Currh-Maslik and congenital reticulocytic ichthyosiform erythroderma are minor variants.

A 2-year-old boy presented with generalized skin lesions since early childhood. He was born healthy out of non-consanguineous marriage. After 8 months, he started developing lesions initially over the joints which then gradually progressed to involve other parts of the body. There was no history of blistering and hearing loss. Face, neck, diaper area and upper part of the trunk was spared, but nipple and abdomen were involved. Hyperkeratotic plaques were present over dorsum of hands and feet. Palms and soles had keratoderma. Scalp, hair, orogenital area, teeth, and nails were normal. Biopsy taken showed marked hyperkeratosis and papillomatosis in the form of crest-like projections of epidermis that was accompanied by perinuclear vacuolization and binucleated keratinocytes. On the basis of the clinical features and histopathology, we arrived at the diagnosis of ichthyosis hystrix of Currh-Maslik type and it was genetically confirmed.

A female infant was born via spontaneous vaginal birth to non-consanguineous parents. Erythroderma with scaling was noted at birth, in the absence of bullae or collodion membrane. Skin biopsy showed mild parakeratosis and acantosis. At the age of 17 years physiological examination revealed short stature, ichthyoses, erythroderma and multiple confluent areas of normal skin predominantly on trunk and extremities. The palmoplantar keratoderma was now more obvious and the patient had also developed bilateral ectropion.

We arrived at the diagnosis of congenital reticulocytic ichthyosiform erythroderma and it was genetically confirmed.

This impairs photo activation of 7-dehydrocholesterol and causes systemic vitamin D deficiency.

A case series of 5 patients diagnosed with congenital ichthyosis and vitamin D deficiency were supplemented with 60000 IU of vitamin D3 for a period of 10 days, followed by a daily allowance of 400-600 IU of vitamin D3, and 40 mg/kg/day of elemental calcium. Symptoms such as scaly skin and stiffness of the body were measured. Blood tests and urine samples were evaluated on the day of administration and then after 10 days, 1 month, and 3 months.

Key Words: congenital ichthyosis, vitamin D deficiency, vitamin D supplementation.

VITAMIN D DEFICIENCY IN PATIENTS WITH CONGENITAL ICHTHYOSIS IN INDIA: CASE SERIES

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Introduction: Ichthyosis, a genetic disorder of keratinization, is characterized by excessively scaling associated with epidermal hyper proliferation and/or cellular retention. Normally, thickness of the outer epidermis is 25 μm and that of a patient with ichthyosis is 10 times greater. This impairs photo activation of 7-dehydrocholesterol and causes systemic vitamin D deficiency.

Materials and method: This was a prospective observational study performed at Kempegowda Institute of Medical Sciences, Bengaluru, Karnataka, India. Patients’ clinical characteristics were recorded. Serum concentration of 25-hydroxyvitamin D were determined along with serum calcium, phosphorus and Parathyroid hormone. X-rays were taken of the bilateral wrists and knee joints in antero-posterior and lateral views. The study included 12 patients with vitamin D deficiency correlating with calcium, phosphorus, parathyroid hormone levels and radiological findings.

Results:

- Out of 12 patients with congenital ichthyosis, 6 were male and 6 female.
- Out of these 12, only 2 had vitamin D sufficiency.
- For 10 patients with vitamin D below the optimal level (<30 ng/ml), following are the tested vitamin D levels:
  - Vitamin D > 20 ng/ml but < 30 ng/ml – 2 patients
  - Vitamin D > 10 ng/ml but < 20 ng/ml – 2 patients
  - Vitamin D < 10 ng/ml – 6 patients
- Serum calcium and phosphorus were normal for all patients.
- 2 patients had hyperparathyroidism.
- Radiologically, 2 patients had Cervical Valgum and 1 patient had Rickets.

Other associated clinical findings were: hypertfreincty of palms (1 patient), Juvenile plantar dermatitis (1 patient), X-linked recessive ichthyosis-biopsy proven (1 patient), history of colloidion baby (2 patients).

Conclusion: Vitamin D plays a vital role in patients with congenital ichthyosis. Vitamin D levels should be tested for every patient with congenital ichthyosis to improve prognosis and management of the patients.

VITAMIN D DEFICIENCY IN PATIENTS WITH CONGENITAL ICHTHYOSIS AND RESPONSE TO VITAMIN D SUPPLEMENTATION

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Vitamin D (a secosteroid molecule) is a prohormone that is essential for calcium and bone metabolism. Skin is the major source of endogenous vitamin D.

Ichthyosis, a genetic disorder of keratinization, is characterized by excessive scaling associated with epidermal hyper proliferation and/or cellular retention. Normally thickness of the outer epidermis is 25 μm and that of patients with ichthyosis is 10 times greater.

In this impairs photo activation of 7-dehydrocholesterol and causes systemic vitamin D deficiency.

A case series of 5 patients diagnosed with congenital ichthyosis and vitamin D deficiency were supplemented with 60000 IU of vitamin D3 for a period of 10 days, followed by a daily allowance of 400-600 IU of vitamin D3, and 40 mg/kg/day of elemental calcium. Symptoms such as scaly skin and stiffness of the body were measured. Blood tests and urine samples were evaluated on the day of administration and then after 10 days, 1 month, and 3 months.

Key Words: congenital ichthyosis, vitamin D deficiency, vitamin D supplementation.
The recognition of skin manifestations as signs of cystic fibrosis, despite overlap with symptoms of other nutritional deficiencies would allow earlier diagnosis, correct treatment, and close follow-up in a multidisciplinary team, and would improve quality of life and outcomes.

**P 025**

NEUROTIC EXCORIATIONS, ALOPECIA AREATA, LIP FISSURES, FISSURED TONGUE, ANGULAR CHEILITIS AND CHEILITIS IN A CASE OF DOWN SYNDROME

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Down syndrome or trisomy 21 is a genetic disease with high prevalence of orofacial anomalies: fissured tongue (prevalence varying between 10 and 95% of patients), angular cheilitis (due to malposition of nasal bridge, muscular hypotonia, enlarged and protruding tongue, mouth breathing and open mouth), lip fissures, malocclusion and dental caries, and gingival hyperplasia. The relation between alopecia areata and Down syndrome is controversial. Some studies are in favour of the idea that the causative gene of alopecia areata is located on chromosome 21 while others have concluded that the association between alopecia areata and Down syndrome is coincidental.

The terms "neurotic excoriations", "psychologic excoriations", and "scratch marks" are used to describe self-inflicted cutaneous lesions especially observed in patients with depression, anxiety or other psychiatric diseases.

We describe the case of an 8-year-old boy diagnosed with Down syndrome with alopecia totalis, neurotic excoriations, fissured tongue, lip fissures, angular cheilitis, and cheilitis.


**P 026**

COLLODIUM IN TWINs AND THEIR OLDER SISTER – CASE REPORT

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The term collodion baby refers to a phenotype that can be characterized by a yellow, shiny, tight parchment-like membrane stretched over the skin. Most children born as collodion babies will spontaneously desquamate within 2 weeks, but it may as long as 3 months. Eventually, these children develop signs of one of several types of ichthyosis skin types. Congenital ichthyosiform erythroderma and lamellar ichthyosis are the most commonly seen forms of ichthyosis presenting with a collodion membrane. However, the membrane may also be present in Netherton syndrome and other very rare forms of ichthyosis, and is always present with harlequin ichthyosis. A small percentage of infants shed the membrane and never display any other skin involvement: a phenomenon called "self-healing collodion baby." We report a unique case of Caucasian twins that were born as collodion babies with a family history of collodion baby in their sister. The babies were from the second pregnancy of nonconsangunous parents and were born by caesarian section at the gestational age of 31 weeks. On examination the whole body was covered with parchment-like membrane with cracks. Associated findings included ectropion, eversion of lips, and flattening of ears. The babies were treated using a humidified environment, prophylactic antibiotics, and emollients. The babies recovered well and were discharged with advice for regular emollient application and check-up. Their 16 year old sister only presented dry skin. The genetic tests to confirm collodion baby are being conducted.

**P 027**

MISDIAGNOSIS OF NEUROFIBROMATOSIS TYPE 1 IN PATIENTS WITH PIEBALDISM

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A 6-year-old boy of Kenyan origin with unremarkable medical history presents a congenital depigmented plaque in the left forearm with progressive appearance of normal pigmentation islands without white forelock. Physical examination also reveals more than 6 café-au-lait macules (CALMs) greater than 5 mm in diameter, and numerous bilateral axillary freckling. No neurofibroma is detected. Hearing and ophthalmologic evaluations are normal. Family history confirms that the father and 2 brothers among 7 siblings have similar phenotypes with white forelock and depigmented plaques of the limbs but without CALMs or freckling. Our patient is diagnosed with piebaldism with CALMs and intertrigous freckling.

Several piebaldism patients with CALMs and intertrigous freckling have been reported. Some authors consider an overlap of piebaldism and NF1, but it is based on application of minimal clinical diagnostic criteria for NF1. No cutaneous neurofibromas or other features of NF1 were seen in any of the cases, although Lisch nodules and scoliosis were described in one girl. Both piebaldism and NF1 are autosomal dominant conditions, while their genes have been localized to different chromosomes, 4q12 or 8q11 and 17q11.2, respectively. Therefore, there is only a small chance of an association. All piebaldism patients with CALMs and intertrigous freckling who underwent gene testing showed mutations in the KIT gene but no NF1 mutations. Furthermore, Legius syndrome described in 2007 and caused by loss-of-function mutations in the STRED1 gene is characterized by multiple CALMs and intertrigous freckling without the typical NF1-associated tumors. This supports the theory that a mutation in the intracellular tyrosine kinase domain of KIT results in severe piebaldism and may be related to the subsequent effect on STRED1 and Ras/MAPK pathway due to inadequate phosphorylation of the KIT-binding domain and appearance of CALMs and freckling.

**P 028**

PAPILLON-LÉFEVRE SYNDROME: A CASE REPORT

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Papillon-Lefèvre syndrome (PLS) is an autosomal recessive disorder that presents with palmoplantar keratodermat, periodontitis, and frequent skin infections. Incidence of between one and four persons per million. In this case report, we aimed to present a typical case of papillon-lefevre syndrome.

**Case Report:** A 4-year-old male patient was referred to our clinic because of dysplasia and premature loss of teeth. The dermatological examination revealed bilateral palmoplantar keratoderma, periodontitis, and frequent skin infections. Incidence of between one and four persons per million. In this case report, we aimed to present a typical case of papillon-lefevre syndrome.

**Discussion:** PLS is a rare disease with palmoplantar keratoderma and destructive periodontitis. Although the etiology is not completely known, immunologic, genetic, and infectious mechanisms play a role. Heredity is autosomal recessive, and studies have shown homozygous mutation in the locus lysosomal protease cathepsin C gene in 11q14-q21. Recurrent infections are also relatively common in PLS. Skin infections such as pyoderma can develop in about 17% of patients. Another feature of PLS is intracranial calcification. A multidisciplinary approach is needed in the treatment of PLS. Saliylic acid and urea-containing emollients can be used for skin lesions. The basis of treatment is oral retinoids such as acitretin and isotretinoin. Control of the periodontist is more difficult. Attracting primary teeth with a combination of professional dentifrice and oral antibiotics is one of the effective methods of treatment. We have described a 4-year-old Turkish boy diagnosed with PLS who exhibited palmoplantar keratoderma, periodontitis, and skin involvement.

**P 029**

AN UNUSUAL CASE OF A BECKER’S NEVUS WITH CHECKERBOARD MOSAICISM

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We report a rare presentation of Becker’s nevus (BN) manifesting as multiple patches in a checkerboard mosaic pattern. An 18-year-old Chinese male presented to us with a 3-year history of asymptomatic brown patches over his anterior chest and beneath his left eye.

On examination, there was a large brown macular pigmentation over his left infraorbital region.
Although BN is a common hamartomatous finding, its presentation as multiple lesions with a checkerboard pattern of cutaneous mosaicism is rare. Only few case reports describing this presentation exist despite being described as characteristic of the condition. This interesting feature of BN speaks to the underlying etiology of its derivation from mesodermal origins during early embryonic development.

**P 030**

SUCCESSFUL TREATMENT OF A PATIENT WITH NETHERTON SYNDROME WITH ALITRETINOIN

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Introduction: Netherton syndrome (NS) is a rare, autosomal recessive, syndromic ichthyosis characterized by ichthyosis linearis circumflexa (ILC), trichorrhexis invaginata, and atopy. Treatment for NS has notoriously been difficult and includes emollients, topical steroids, immunomodulators, narrowband UVB, IVIG, and occasionally TNF-inhibitors. Oral retinoids such as acitretin have been used with moderate efficacy, potentially leading to exacerbations of the disorder.

Case report: We present a 23-year-old woman with NS, confirmed by immunohistochemistry showing complete absence of LEKTI, and by mutation analysis displaying two heterozygous loss of function mutations in SPINK5. She had been suffering from long-standing extensive ILC. She had previously been treated with emollients, topical steroids and immunomodulators, narrowband UVB, and repeated IVIG infusions with no sustained improvement. She was first started on adalimumab 40 mg every other week for 12 months. After some initial improvement, she relapsed and treatment was stopped. Thereafter, alitretinoin (Tocilin) 30 mg daily was commenced. This led to 75% clearance of her skin lesions within 3 months with sustained efficacy over a treatment duration of 15 months. Tolerability has remained excellent without side effects.

Discussion/Conclusion: To our knowledge this is the first report of the use of alitretinoin in NS. Alitretinoin is a pan-retinoid receptor agonist and, in contrast to other systemic retinoids (e.g. acitretin or isotretinoin), it not only binds to retinoic acid receptors (RAR) but also retino X receptors (RXR). It therefore has more pronounced anti-inflammatory and immunomodulatory effects apart from anti-proliferative effects. Alitretinoin has widely been approved for the treatment of refractory hand eczema, however it has successfully been used in other conditions such as psoriasis, pityriasis rubra pilaris, and Darier’s disease.

Although larger case series are needed to confirm our observation, we propose that alitretinoin seems to be a promising treatment option with good tolerance for patients with NS.

**P 031**

CUTIS LAXA AND COPPER TRANSPORT ANOMALIES. ONE GENE, TWO ENDS OF THE SPECTRUM

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Introduction: Cutis laxa, hypopigmented skin, and hair abnormalities are early manifestations of Menkes syndrome (MS) and occipital horn syndrome (OHS). Both are rare, X-linked inherited entities, produced by mutations in the ATP7A gene. We present two clinical cases that show the two ends of the spectrum of this copper metabolism disorder.

Clinical cases: CASE 1. A 3-month-old child with epilepsy, cutis laxa, hypopigmentation, hairy dry scalp, micrognathia, wide fontanelles, pectus excavatum, hypotonia, hyperlaxity, tortuous cerebral vessels, and low ceruloplasmin and copper plasma levels. No history of consanguinity.

CASE 2. A 16-year-old boy with normal pubertal and intellectual development. No family history of consanguinity. His mother had fair skin and hoarseness. He was diagnosed with congenital cutis laxa and hypotonia. Between 2 and 16 years, he developed bladder diverticula, inguinal hernias, bronchial asthma, dental alterations, and multiple skeletal anomalies characteristic of OHS. Copper plasma levels were normal.

Results: In the first case, the skin and hair anomalies were key to suspecting a diagnosis of MS and establishing treatment with subcutaneous copper-histidine. A mutation in ATP7A was later demonstrated. In the second case, the diagnosis was not established until 16 years of age.

Discussion: The ATP7A gene encodes a copper-carrying enzyme, essential in multiple biological processes. Functional protein levels determine the severity of the phenotype. Early treatment with copper improves MS survival. The diagnosis of OHD tends to be delayed for years because copper and ceruloplasmin levels may be normal and the typical phenotype (occipital exostosis) appears later in time.

**P 032**

MANAGEMENT OF CUTANEOUS SQUAMOUS CELL CARCINOMA IN PATIENTS WITH RECESSIVE DYSTROPHIC EPIDERMOLYSIS BULLOSA: OUR EXPERIENCE IN A UNIVERSITY HOSPITAL

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Epidermolysis Bullosa (EB) represents a cluster of different inherited pathologies characterized by a noticeable fragility of skin and mucosa that triggers blister formation spontaneously or due to minor traumas. It is considered a rare disease and affects people regardless of their ethnic origins or sex. Recessive Dystrophic Epidermolysis Bullosa (RDEB) provokes the decrease or even the absence of collagen type VII (C7), which results in mucosal and cutaneous erosions, injuries, and aggressive squamous skin carcinomas (cSSC). Because of its discordant characteristics (histological behaviour code) and its high propagation risk, one of the essential bases for treatment is prevention. Furthermore, knowing how to manage symptoms once a diagnosis is established will be essential and a challenge because of the rarity of the disease. Evaluating the patients who develop cSSC within a cohort of 106 patients treated in a University Hospital in Madrid, we will determine the best treatment mechanisms and adjuvant options based on survival rate and changes of metastasis.

**P 033**

A KLICK SYNDROME MIMICKING AN ERYTHROKERATODERMA

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Introduction: Erythrokeratoderma and KLICK syndrome are two rare diseases with different clinical presentation belonging to the group of congenital ichthyoses. We report on a patient with an atypical presentation of KLICK syndrome, initially diagnosed as having an erythrokeratoderma.

Case report: A 30-year-old caucasian woman was referred to our department for genetic counseling before planning a pregnancy. Her skin disease presented at birth with diffuse scaling. She then developed fixed erythematous and hyperkeratotic plaques on the limbs, associated with diffuse white skin scaling of the whole body. She also had a moderate reddish palmoplantar keratoderma. She started acitretin at the age of 18 years. Pictures performed before acitretin revealed similar skin lesions but more pronounced hyperkeratosis. The diagnosis of erythrokeratoderma or loricrin keratoderma was suspected. Molecular analysis of GJA1, GJB3, and POMP genes was performed before acitretin revealed similar skin lesions but more pronounced hyperkeratosis. The diagnosis of erythrokeratoderma or loricrin keratoderma was suspected. Molecular analysis of GJB3, GJA1, GJH, and LOR genes was performed but did not reveal any mutations. A panel of genes was then tested, that revealed the homozygous mutation c.95delC in the 5’ UTR region of the POMP gene, leading to the diagnosis of KLICK syndrome. In view of these genetic findings, a secondary accurate clinical examination showed the presence of slight linear hyperkeratotic areas on folds associated with subtle digit contractions. This was important for genetic counseling, the patient having an autosomal recessive disorder instead of an autosomal dominant disease.

Discussion: Around twenty cases of klick syndrome have been reported. Patients carry this single-nucleotide homozygous deletion of the POMP gene. Patients show diffuse scaling, palmoplantar keratoderma, constricting bands around fingers, and linear keratotic papules on the folds. The presence of well demarcated erythematous and hyperkeratotic plaques, as seen in erythrokeratoderma, has never been demonstrated previously, but only suspected in another patient.

Conclusion: Advances in genetic techniques lead to the identification of novel phenotypes and expand the knowledge of the genodermatoses.
P 034
BUSCHEK-OLLENDORFF SYNDROME
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We present the case of a 9-year-old boy with asymptomatic lesions on his back of unknown onset. Upon physical examination we saw subtle soft white-yellow lesions with a widespread distribution throughout the middle and lower back. We performed a skin biopsy of one of those plaques and histologic examination showed coarse, thickened elastic fibers in the reticular dermis, compatible with an elastoma. An X-ray of the hand, tibia, and fibula revealed dense bony islands corresponding with osteopoikilosis. Given the presence of a connective tissue nevus (elastoma) and osteopoikilosis, the diagnosis of Buschke-Ollendorf syndrome was made. The patient did not have a family history of similar skin or bone lesions.

Buschke-Ollendorf syndrome is a rare autosomal dominant disease of the skin and bones. Loss of function of the LEMD3 gene has been described in these patients. Buschke-Ollendorf syndrome is characterized by the presence of connective tissue nevi in association with sclerotic bony lesions. There are cases that exclusively present skin lesions and others with only bony manifestations. The most frequent skin manifestations are connective tissue nevus, presenting as skin-colored to yellow plaques usually located in the lower back, buttocks, and inner limbs. The typical bony presentation is osteopoikilosis, representative tissue nevus, presenting as skin-colored to yellow plaques usually located in the lower back, buttocks, and inner limbs. The typical bony presentation is osteopoikilosis, representative tissue nevus, presenting as skin-colored to yellow plaques usually located in the lower back, buttocks, and inner limbs. The typical bony presentation is osteopoikilosis. There are cases that exclusively present skin lesions, and others with only bony manifestations. The most frequent skin manifestations are connective tissue nevus, presenting as skin-colored to yellow plaques usually located in the lower back, buttocks, and inner limbs. The typical bony presentation is osteopoikilosis, representative tissue nevus, presenting as skin-colored to yellow plaques usually located in the lower back, buttocks, and inner limbs. The typical bony presentation is osteopoikilosis.

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RECESSIVE DYSTROPHIC EPIDERMOLYSIS BULLOSA MIMICKING NEONATAL BULLOUS PLEMPHIGOID

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Dystrophic epidermolysis bullosa (DEB) is a rare inherited disease produced by mutations in collagen VII. Diagnosis can be challenging as in the case described herein.

Case: A 42 months old newborn was referred with blistering lesions on his hands and feet. Skin biopsy showed subepidermal blister with eosinophils, and C3 deposits along the basement membrane were detected in direct immunofluorescence. A diagnosis of infantile bullous pemphigoid (IBP) was made. The child suffered an exacerba-

tion following the administration of the first hexavalent vaccine. Nervertheless, it improved upon steroid therapy. During the evolution, lesions spread to the arms, legs, trunk, face, and oral mucosa. They resolved leaving scar plaques. In addition, milium cysts developed after resolution of the blisters and nail dystrophy. The diagnosis was reconsidered and a new biopsy was performed with similar findings. However, immunofluorescence mapping showed the presence of integrin-5, integrin-6, laminin-332, and collagen-IV on the roof of the bullae, and a decrease of collagen-VII. These findings were suggestive of DEB.

Discussion: Neonatal BP usually appears in neonates of mothers with gestational BP at birth or during the first days of life. The blisters usually resolve without scarring within two to four weeks. Infantile BP usually appears within the first year of life, is predominantly acral, and has a good prognosis. The lesions in DEB are persistent in time and resolve leaving scars. Moreover, patients present milium cysts and nail dystrophy among other manifestations.

The presence of a subepidermal bullae with eosinophils is suggestive of BP although it shares some pathological data with DEB. In our case the immunological findings together with the worsening after vaccination made us propose a diagnosis of BP. Nevertheless, the clinical evolution together with the findings of the immunofluorescence mapping led us to the diagnosis of a recessive DEB.

P 039

PIGMENTARY MOSAICISM: THREE CASES AND REVIEW OF THE LITERATURE

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Introduction: Pigmentary mosaicism is a disorder that appears as hypo- or hyperpig-

mentation that can be isolated or syndromic. Different patterns are observed: large or nar-

tive in pediatric dermatology but still misunderstood or confused with other pigment disorders. Hence the need for this work.

Case report: We report the cases of three children: one infant of 18 months and two brothers of 12 months and 5 years, with no significant pathological history except a similar case in the father of the last two patients. The patients presented dyschromic spots on trunk and limbs, hypo and hyperpigmented, arranged in linear and narrow bands according to Blaschko’s lines for two patients, and “in chessboard” for the third one, with a clear delimitation by the median line. The remainder of somatic examination was normal. The diagnosis of isolated pigmentary mosaicism was established. Genetic counseling with an amniotic monitoring of the sibling was then proposed.

Discussion: Pigmentary mosaicism is defined by the coexistence of two distinct melano-
cytic cell lines (mutated and normal) with migration abnormality of melanoblasts from the neural crest to the basal layer of the epidermis. Clinically, it appears as hypo- or hyperpig-

mentation that can be isolated or syndromic. Different patterns are observed: large or nar-

row bands according to Blaschko’s lines, in chessboard, in phylloid form, in sheets, or lateralized. No systematic additional examination is proposed in the absence of suspicious clinical signs. In the case of localized pigmentary disorders, as in our patients, pediatric follow-

up and monitoring of growth may be recommended.

Conclusion: Pigmentary mosaicism is a classic pattern of consultation in pediatric dermatology. It can be isolated or a part of more complex syndromes. No further examination is required in the absence of suspicious clinical signs, and simple monitoring is generally recommended.

P 040

BILATERAL MULTIPLE NEVUS SEBACEUS IN A NEWBORN

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Nevus sebaceous is a benign hamartoma of the skin characterized by hyperplasia of the epidermis, and immature hair follicles, sebaceous, and apocrine glands. Lesions usually present at birth and appear as well-circumscribed, waxy, yellow, hairless patches, often localized on the head and neck region. The lesion grows proportionally with the patient's age and becomes progressively thick. Rarely there may be multiple lesions, sometimes as a part of nevus sebaceous syndrome. Here we report a case of multiple Nevus sebaceous in a healthy child.

An otherwise healthy newborn male consulted for multiple verrucaous lesions on the scalp. On dermatologic examination multiple orange colored, hairless, verrucous linear plaques were noticed on the scalp and neck. Punch biopsy revealed immature hair follicles, hyper-

plastic sebaceous glands, dilated apocrine glands, and epidermal hyperplasia consistent with nevus sebaceous. Detailed examination and imaging including central nervous sys-

tem, eyes, and bones revealed normal features. Topical keratolytic agents applied for the keratotic portions and the parents reassured benign nature of the lesions. Sebaceous nevi are sporadic and occur with equal frequency in males and females of all races. NS is thought to be caused by postzygotic somatic mutations that may result in vari-

ous clinical expressions of mosaicism. Cases of multiple NS were rarely reported and usu-

ally occurred with unilateral involvement while bilateral involvement is extremely rare. Benign or malignant neoplasms may occur in NS, usually in adulthood. There is no consen-
sus regarding the optimal therapeutic management of NS. Given the low risk of malignant transformation in children clinical follow-up is considered to be safe. In the present case, widespread distribution of the lesions led us to investigate other ectodermal abnormalities and nevus sebaceous syndrome were found. This case is unique because of the bilateral linear and multiple NS lesions with no systemic abnormalities. The absence of systemic involve-

ment led us to propose observation of the lesions until puberty.

P 041

A RARE CAUSE OF SPARSE, SLOW-GROWING HAIR AND A FUSED TOOTH: A CHILD WITH TRICHORHINOPHALYNGEAL SYNDROME TYPE I

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A 5 year old girl presented with sparse, fair, slow-growing hair which had never required cutting and fragile nails that did not need trimming. Her facial features included a long phil-

trum, pear-shaped nose, and thin upper lip. She had delayed eruption of primary dentition and her first tooth was a fused lateral incisor at 13 months with her last tooth erupting by 42 months. Skeletal features included broad thumbs and hollucies with her growth sym-

metrical on the 9th percentile. Her hair pull test contained a majority of scalp hairs in anagen phase. Her facial features were consistent with Trichorhinophalangeal syndrome (TRPS). Genetic testing identified a heterozygous pathogenic frameshift variant in the TRPS1 gene (c.2512_2513del AA, p. (Lys838 fs)).

TRPS Type 1 is a rare autosomal dominant malformation syndrome with ectodermal, cran-

iofacial, and skeletal features caused by heterogenous mutations in the TRPS1 gene. Hair is characteristically fine, blond, and congenitally sparse especially over the fronto-temporal area of the scalp with sparse eyebrows. Total scalp alopecia may develop by early adult-

hood. A pear-shaped nose, long philtrum, and thin upper lip are characteristic facial fea-

tures. Skeletal manifestations include short stature, cone-shaped epiphyses, brachydactyly, osteoarthrisis, and avascular necrosis of the femoral head. Dental anomalies including microodontia, caries, malocclusion, and delayed eruption of teeth are recognised.

TRPS Type 2 results from deletion of TRPS and contiguous genes at chromosome locus 8q34. In addition to features of TRPS1, TRPS2 is associated with multiple osteochondro-

mas and developmental delay. Mutations of TRPS may be overlooked or mistaken for presenta-

tions of loose anagen syndrome or pseudo-pseudohyoparathyroidism. Trichorhinophalangeal syndrome is rare but should be considered in presentation of sparse and slow-growing scalp hair, short stature, broad thumbs, and characteristic facial features. Dental anomalies are recognised but this is the first report with a fused tooth.

P 042

OCULOECTODERMAL SYNDROME: A RARE MOSAIC RASOPATHIES

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Oculoectodermal syndrome (OES) is a very rare disorder with only twenty cases reported so far. Its main features include congenital scalp lesions (aplasia cutis congentialis alopeciae), ocular dermoids, and benign tumors as nose, eyelids, lip, and granulomatous of the jaw. Recently, Peacock et al. identified KRAS mutations in affected tissues from two patients with OES, thus suggesting that OES is a mosaic RASopathy. OES shares many common features with enchondracanciosoantralopatiaipo-

matodein (ECOAL), a very rare neurocutaneous disorder characterized by hyperpigmented (nevox pilsolipus, non-scarring and scarring alopecia, skin tags), neurologic (intracranial and spinal lipomas, arachnoid and porencephaliccysts, cerebral asymmetry, dilated ventricles
and calicifications, and eye (chorioretinal) abnormalities. OES and ECCL have therefore been assumed to represent the same spectrum of disorders with OES being the mild phenotype. Here we present a 5-year-old boy who had a 4 × 8 cm yellowish hairless plaque on the right frontoparietal scalp, a right epibulbar dermoid, and skin tags on the right upper eyelid and eyebrow. The child was otherwise healthy. The family history was non-contributory. A biopsy from the scalp lesion demonstrated a paucity of hair follicles without scarring and isolated arrector pili muscles arranged in a line parallel to the surface. These histophenotypic features are suggestive of the nevus lipomatosus and has been regarded as the dermoscopic hallmark of ECCL. A K RAS mutation affecting codon 146 (c.437C>T) was identified in DNA derived from the epibulbar dermoid, but not in leucocyte-derived DNA. Similar K RAS mutations affecting codon 146 were detected in two other patients with OES and one patient with ECCL. These data support the evidence of OES being a mosaic RASopathy sharing a common etiology with ECCL.

P 043
PIGMENTARY MOSAICISM IN A PATIENT WITH RING 18 CHROMOSOME
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Introduction: Pigmentary mosaicism (PM) encompasses multiple systems with phenotypic expression present mostly in the dermal and musculo-skeletal systems. In skin it is characterized by flat hypopigmented or hypopigmented lesions along the lines of Blaschko. About one-third of clinically diagnosed cases of PM demonstrate chromosomal mosaicism.
Clinical case: We report on a 4-year-old male patient whose father had a factor V Leiden heterozygous mutation and whose maternal aunt had glycogenosis type V, with no other diseases of interest. After a full term birth from an elective delivery he was diagnosed with microcephaly, psychomotor retardation, and generalized hypotonia, impairing ambulation. After a Comparative Genomic Hybridization of Peripheral Blood, a ring chromosome 18 was demonstrated with 17% mosaicism. He was referred to the dermatology department due to a 3-year history of stable asymptomatic hyperpigmented lesions which appeared within the first months of life. Upon examination we observed a bilateral thoracoabdominal macular hyperpigmentation of blashickoid distribution with midline demarcation and without association to hypopigmented macules or other primary lesions. There were no annexes, and no mucusa or musculoskeletal alterations.
Discussion: Linear and whorled nevoid hypermelanosis (LWNH) was described by Kalter in 1988 as a blunted hyperpigmentation of blashickoid distribution. Onset is within a few weeks or months of birth. Histopathology reveals epidermal hyperpigmentation without melanocytic proliferation or pigmented incontinence. LWNH has been related to the pigmentary variation in cellular mosaicism, frequently associated with chromosomal mosaics. Occasionally they present extracutaneous manifestations with cardiac, musculoskeletal or neurological involvement, as well as ponderostatural or psychomotor abnormalities. The main differential diagnosis is Incontinentia Pigmenti which presents with vesiculo-verrucous lesions at early stages and shows pigmentary incontinence histologically.
Conclusion: We present a Linear and whorled nevoid hypermelanosis with extracutaneous involvement in association with a mosaic ring 18 chromosome, previously not described in the literature.

P 044
XERODERMA PIGMENTOSUM - DE SANCTIS-CACHIONE SYNDROME IN TWO LEBANESE FAMILIES
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Nucleotide excision repair deficiencies of the Xeroderma pigmentosum type are very rare diseases with incidences of approximately 1:250,000 in the USA and 1:450,000 in Europe. This incidence, however, varies throughout the world with peaks of up to 1:2,400,000 affected individuals in Japan as well as in the Arabic countries and North Africa. Herein we report on two young children aged 1 and 4 age, both of Lebanese families with consanguineous backgrounds, who presented independently from each other within the last two years. Both families reported previous deaths of other family members with similar syndromes. Clinically, the combination of facial freckling, dyspigmentation, and ocular pigmentation with marked photophobia pointed towards XPC complementation group deficiencies for TC-NER. In fact, the mutation analysis revealed a homozygous autosomal recessive XPA mutation affecting codon 146 (c.437C>T) as the cause of the disease. He was referred to the dermatology department, but he remained asymptomatic and the disease was controlled with UV protection and sunscreen.

P 045
UNDERSTANDING EPIDERMOLYSIS BULLOSA FROM THE PATIENT’S PERSPECTIVE
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Epidermolysis bullosa (EB) is a rare, often severe genetic disorder characterized by mechanical fragility, and blistering or erosion of the skin, mucosa, or epithelial lining of organs in response to little or no trauma. There are no approved treatments for EB, and there remains limited information on the burden of illness attributed to EB wounds. To address this need, an institutional review board-approved survey (80 questions/items) was developed to collect demographics, diagnostic data, current management practices, and burden of illness information on patients with EB, as reported by patients or caregivers. Participants were recruited through US EB patient advocacy organizations; surveys were conducted via third-party telephone interview. An interim report (as of May 2, 2017) of 95 respondents completing the EB patient survey found the following proportions of EB subtypes: 27.3% simplex, 40% recessive dystrophic, 27.3% dominant dystrophic, and 5.4% junctional. Age ranged from 0.2 to 62 years (mean 21.3 years). 63.6% were female. 30.9% of respondents reported body surface area of wounds -30%. The average intensity of symptoms attributable to EB wounds on a 10-point scale (10 = most severe) were 5.71 for itch, 4.93 for acute pain, and 3.89 for chronic pain. Wound care burden was high; 54.6% of respondents perform at least daily wound dressing changes, and 32.8% noted that full-body dressing changes take at least 2 h. Before dressing changes, cream or ointment was routinely applied directly to wounds by 94.5% of respondents. Only 44.5% were satisfied with the prescribed wound care plan. Reduction in the number and severity of wounds, acceleration of wound healing, and reduction of pain were rated as the most important qualities of a potential new EB therapy. Overall, this survey characterizes the degree of disease burden among patients and caregivers, and highlights the need for new therapeutic options.

P 046
SIMPLY WARTS OR THE TIP OF THE ICEBERG?
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Introduction: Palmo-planter warts are caused by human papilloma virus (HPV) and a common medical complaint in childhood. Even though they are generally harmless, in rare cases their presence can be a clue of underlying immunodeficiency disorders. We describe the case of a child with unusually large palmo-planter warts recalcitrant to standard treatment.
Case: An 8-year-old girl presented to our department with extensive warts on the soles of her feet, and most especially her palms, for about 6 months. She had used different treatment modalities without success. Apart from a mild recurrent folliculitis her personal history did not reveal anything of importance. The child was otherwise healthy; no other medical complications are present in her family. Apart from her large, numerous warts on her feet, and most especially on her palms, there were no other stigmata or clinical features suggestive of immunodeficiency. After a detailed medical and dermatological examination, a complete blood count was performed. The test results showed no abnormalities. The child was otherwise healthy.
Discussion: The case of our patient is unusual in that a patient with palmo-planter warts recalcitrant to standard treatment is described for the first time. Our patient demonstrated no clinical stigmata or other symptoms suggestive of an underlying immunodeficiency disorder. The patient was otherwise healthy and her personal and family history did not reveal anything of importance. If we take into account the large number of warts and the fact that they were recalcitrant to various treatments, it is possible that this patient has a mild primary immunodeficiency disorder. The diagnosis of immunodeficiency disorders is difficult and often delayed. The recognition of atypical warts may suggest the presence of an underlying immunodeficiency. However, in rare cases the patient may be asymptomatic for a long period of time. The identification of atypical warts in a patient with a medical history of recurrent infections should prompt the dermatologist to consider the possibility of an underlying immunodeficiency disorder. The diagnosis of immunodeficiency disorders is based on a combination of clinical features, laboratory tests, and genetic analysis. The clinical features include the presence of atypical warts, recurrent infections, and other symptoms suggestive of an immunodeficiency disorder. The laboratory tests include a complete blood count, immunoglobulin levels, and T-cell subset analysis. The genetic analysis includes sequencing of the genes involved in immune function. The diagnosis of immunodeficiency disorders is challenging and requires a multidisciplinary approach. The treatment of immunodeficiency disorders is based on the underlying cause. The management of the associated dermatological conditions is usually targeted therapy to control the warts and improve the quality of life. In conclusion, the case of our patient highlights the importance of considering an underlying immunodeficiency disorder in the management of atypical warts. Further research is needed to determine the exact cause of these warts and to improve the management of these patients.
P 047

DISSEMINATED HERPES ZOSTER IN A NON-VACCINATED CHILD
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Introduction: Herpes zoster is rare in children with no personal history of chickenpox and disseminated pediatric herpes zoster is even more unusual.

Case report: A 9-year-old boy with previous history of non-vaccination by parents’ decision was brought to dermatologic emergency because of acute lesions in the right groin, thigh, hip and lumbar region. He was healthy with no fever, pain, or other symptoms. On physical examination, he presented multiple coalescent biogranular lesions with extensive crouton basis in the described areas, multiple isolated vesicles on the trunk, face and upper extremities, and palpable lymph nodes less than 1 cm on the right groin. He was hospitalized and treated with intravenous acyclovir, and zinc hydroxide plus topical fusidic acid ointment under basis in the described areas, multiple isolated vesicles on the trunk, face and upper extremities, and palpable lymph nodes less than 1 cm on the right groin. He was hospitalized and treated with intravenous acyclovir, and zinc hydroxide plus topical fusidic acid ointment. The lesion resolved over the course of 2 weeks.

Discussion: Herpes zoster can occur in children with no history of chickenpox when their mothers are infected during pregnancy after maternal immunoglobulin G antibodies have waned. In the described case, the patient’s mother had a recent chickenpox infection and the child was not vaccinated. This case highlights the importance of vaccinating children against chickenpox.

Conclusion: Herpes zoster is rare in children with no personal history of chickenpox and disseminated pediatric herpes zoster is even more unusual.

P 048

SCABIES: MAKING SENSE OF MANAGEMENT PRINCIPLES
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Scabies, caused by the itch mite Sarcoptes scabei var. hominis, affects 100–300 million people worldwide. Recently, the WHO Strategic and Technical Advisory Group subcommittee recommended inclusion of this disease into category A of neglected tropical diseases.

A typical presentation, sometimes missed by general practitioners and child care providers, further add to the disease burden in the pediatric population. Involvement of head and neck area, vesiculopustular lesions on palms and soles, and severe pruritus occasionally presenting as failure to thrive can all be seen. Certain endemic areas have high rates of poststreptococcal glomerulonephritis and rheumatic heart disease, due to secondary infections of scabies lesions. Clinical diagnosis based on history of night time itching, family history, and the presence of lesions like papules and burrows in classical sites is often sufficient to start treatment. The definitive diagnosis can be made by microscopic identification of the mite, sycobala or eggs. The mites can also be demonstrated by dermoscopy and confocal microscopy. Effective treatment, topical as well as oral, is available and in endemic populations, mass drug administration is found to be efficacious for control of scabies and secondary impetigo.

For individual cases, the significance of basic measures like cutting of nails, a bath with soap and water before applying topicals, treatment of family and close contacts, washing of clothes with hot water or using a washing machine if possible cannot be overemphasized. Oral ivermectin has also been safely used in infants.

The purpose of this presentation is to draw attention to scabies in children as a highly symptomatic but treatable disease.


P 051

NEONATAL VARICELLA–CASE REPORT
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Chicken pox (varicella) is an acute febrile illness with vesiculo-pustular rash caused by varicella zoster virus (VZV). Its manifestation varies from mild febrile illness to severe-life threatening complications like bacterial-superinfections, pneumonia, encephalitis, and bleeding disorders. It is rare in infants and newborns due to passive immunity received from the mother. Infectious diseases acquired during pregnancy are generally transmitted in four ways: through the placenta during pregnancy, perinatally during the labor either by
aspiration of amniotic fluid, or by inoculation of infectious agents into the skin or mucosa of the child, or in mother’s milk during nursing.

**Case details:** We report a case of varicella in a three week old newborn, a female baby born in the 38th week of pregnancy by Caesarian section. Perinatal observation detected hepatopathy with increased bilirubin and the newborn was treated by phototherapy without any complications. General and systemic examinations did not reveal any other abnormality. Vital parameters were normal and baby had no dermatological lesions. Mother, 25 year old primipara, had contact with varicella without evidence of illness a 2-3 days before delivery. The neonate was admitted to the intensive care unit on the 26th day of life because of fever and an extensive rash. During next two days papulovesicular and pustular rash occurred. Varicella virus infection was suspected by dermatologists. Treatment with i.v. Ayclovir was initiated and after 10 days the rash completely disappeared. Diagnosis of varicella in a newborn was suspected due to clinical symptoms, and the history of her mother’s contact with varicella, and was confirmed by positive viral laboratory tests (VZV IgM pos.).

**Conclusion:** We present a rare case of severe varicella infection in a newborn transmitted from her mother. These findings emphasize the importance of obtaining a detailed history in order to make the correct diagnosis, and confirm the susceptibility of pregnant women to VZV.

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**P 052**

**CUTANEOUS MANIFESTATIONS IN PATIENTS WITH PRIMARY IMMUNODEFICIENCY DISEASE IN THE BALEARIC ISLANDS**

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Primary immunodeficiencies (PID) are rare diseases associated with serious or life-threatening medical complications and social issues. Documentation of PIDs expands our knowledge about the characteristic features of these disorders and is essential for the development of diagnostic and therapeutic strategies. Our series of PIDs from the Balearic Islands in Spain comprises 150 patients with an overall prevalence of 12 per 100,000 inhabitants and 33% (n = 48) of these patients were diagnosed in pediatric age (age ≤16 years). Selective immunoglobulin IgA deficiency represented the most common entity in this group (n = 29; 60%), followed by Di-George anomaly (n = 7; 15%), common variable immunodeficiency (n = 5; 10%), and X-linked agammaglobulinemia (n = 5; 10%).

Cutaneous manifestations are common in PIDs and may be the presenting clinical manifestations. Our retrospective study showed that 10% of patients (n = 5) presented with skin manifestations as an early feature of the disease, including five patients with dermatitis resistant to treatment including. Patients who showed erythroderma from birth included two males with Wiskott Aldrich syndrome due to a mutation in the WAS gene presenting with newborn eczema, a female with autosomal STAT3-negative hyper-IgE syndrome (HIES) and eczematous rash, a male with immune dysregulation polyendocrinopathy enteropathy X-linked (IPEX) syndrome with atopic dermatitis, and a male with Omenn syndrome. Other cutaneous manifestations included mucocutaneous candidiasis in a patient diagnosed with STAT1 gain of function mutation, as well as other skin infections. Bacterial skin infections were more prevalent in patients diagnosed with congenital defects related to the number and function of phagocytes, as we observed in our patients with HIES and X-linked chronic granulomatous disease.

Skin manifestations are common findings among several PIDs and may aid in the early detection and treatment of immunologic defects.

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**P 053**

**COSMETICALLY CONCERNING LESIONS: MULTIPLE CUTANEOUS LEISHMANIASIS ON A 10-MONTHS-OLD GIRL’S FACE**

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Leishmaniasis refers to a broad spectrum of diseases caused by protozoan parasites belonging to the genus Leishmania. Cutaneous leishmaniasis is endemic in some regions of Mallorca, the Balearic Islands, Spain. Infected patients typically have a non-healing ulcer on exposed skin.

We report a case of multiple cutaneous leishmaniasis on the face of a 10-month-old girl. She presented four lesions on the forehead and on the face. A biopsy was performed, confirming clinical suspicion of multiple cutaneous leishmaniasis. The patient was rapidly treated with five doses of endovenous liposomal amphotericin B (3 mg/kg/dose), with successful response. There were no reported adverse effects. She was discharged at home with topical amphotericin B (twice per day) and controlled in the pediatric dermatology external consultations. However, after a good initial evolution the lesions reappeared seven months later. She was admitted to the hospital again to complete another five endovenous doses of liposomal amphotericin B at the same dosage as the first time. The lesions improved again. Weekly photographic control by the family was recommended and another visit to the dermatologist was scheduled. Therapy with liposomal amphotericin B could be offered to patients with cosmically concerning lesions, such as those in this case, and it can be considered an option for initial therapy in patients requiring systemic therapy for cutaneous leishmaniasis, even in young infants.

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**P 054**

**CUTANEOUS LEISHMANIASIS IN THE PEDIATRIC POPULATION**

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**Introduction:** Leishmaniasis is an endemic infectious disease in Mallorca (Spain). Although it is a disease of compulsory declaration to authorities, the notification is not usually made and therefore the current incidence of cutaneous leishmaniasis (CL) is unknown.

**Material and methods:** We performed a retrospective study reviewing all CL cases in the pediatric population between 2003 and 2017 with histological (leishman-Donovan bodies detection) or microbiological (Giemsa’s stain) confirmation. Several parameters (age, gender, clinical form and, clinical response) were registered.

**Results:** Out of the 17 CL, 52.9% were in boys, and the youngest patient was one year old. Most of them lived in a rural environment. The most common clinical presentation was a solitary papule located on the cheek and only 23.5% were not situated on the face. The diagnosis orientation was CL in 70% of cases. We performed confirmation by Giemsa’s staining only in ulcer-crusted lesions. The average time between clinical onset and diagnosis was 9 months. Most cases needed only 2 infiltrations of intralesional pentavalent antimonial before resolving (1-6 cycles). Liposomal amphotericin B was administered in one case because of an intensely ulcerated lesion on the forehead. Visceral dissemination (Kala-Azar) was only diagnosed in one patient.

**Conclusion:** We report the first study of LC in pediatric patients in our area. Like other authors we agree that CL should always be treated because it usually affects exposed areas with consequent aesthetic compromise, and also because of the risk of visceral dissemination. Intralesional pentavalent antimonials have been the treatment of choice in small cutaneous lesions, although in two cases other treatments were needed.

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**P 055**

**SCABIES IN THE PEDIATRIC EMERGENCY DEPARTMENT — A CASE SERIES OF 10 YEARS**

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**Objective:** Scabies is a common pediatric infectious disease. The aim of this study was to characterize the episodes of scabies observed in a tertiary pediatric emergency department.

**Methods:** We conducted a retrospective study of the admissions to the pediatric emergency department, with the diagnosis of scabies during a 10-year period (January 2007 to December 2016). Age, gender, time to diagnosis, epidemiological context, treatment, and follow-up were analyzed.

**Results:** During the period of study, 424 cases of scabies were diagnosed (52% males), with a mean age of 6.7 years (range: 1 month-17 years). The average annual cases was 42 (range: 17-92) with a progressive increase throughout the years. October and November were the months with the largest number of cases. In 23% of the cases other diagnoses were made first, most frequently atopic dermatitis. The average time from the onset of symptoms to the final diagnosis was 5 weeks (range: 1 day-6 months). In 22% of cases a pediatric dermatologist was consulted, in the majority to confirm the diagnosis. Infestation by family a member was registered in 81% (224/227). In 55% of cases there was written information regarding the treatment of close contacts. The treatment of choice was benzyl benzoate in 50%, sulfur in 29%, and crotamiton in 16%. In 28% (9%) of cases, the treatment was changed due to persistence or relapse of symptoms. In the latter, 82% had been initially prescribed benzyl benzoate.

**Conclusion:** There has been a continuous increase in scabies diagnosis in our emergency department. In 1/4 of cases, other diagnoses were made during the first medical evaluation with a prolonged average time to the final diagnosis. The epidemiological context was an important clue to the diagnosis.
P 056
VULVAR HERPES ZOSTER IN A GIRL SIMULATING HERPES SIMPLEX VIRUS INFECTION. THE ROLE OF SEXUAL ABUSE IN CHILDREN
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Introduction: Herpes zoster is caused by the reactivation of varicella zoster virus (VZV) which is rarely seen in children, even more rarely when it is limited to the genital region. In most cases, the appearance of blistered genital lesions in a child should make us consider a herpes simplex virus (HSV) infection, which is a possible sign of sexual abuse. The detection of this type of lesion in our country requires initiation of an action protocol in which medical personnel (pediatricians, dermatologists, and forensic specialists), social workers, and judges take part. The goal is protecting the child from possible abuse in their usual environment. Once the lesion is detected, the necessary samples are collected to determine its etiology and it is brought to the attention of the duty court who evaluates the case and decides the necessary action. The impact of this process is important for the patient and for their parents or guardians who receive information about possible abuse of their child. Considering that most cases of abuse are perpetrated by parents or close relatives, parents and guardians often feel themselves being closely observed or blamed during the time it takes to reach the diagnosis and determine definitive procedures.

Case report: We present a 6-year-old girl with vulvar grouped blister lesions of a few days duration, simulating a HSV infection, that followed the previously mentioned legal protocol. After the diagnosis of VZV was made three weeks later using the polymerase chain reaction (PCR) test. Although the patient appeared to be unaffected by the procedure, from the first visit her parents were reluctant to cooperate with the psychological evaluations and the legal procedure. After diagnoses the legal case was finally closed.

P 057
IS THERE A VULNERABLE AGE FOR SCABIES?: A RETROSPECTIVE STUDY IN A PEDIATRIC POPULATION
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Introduction: Scabies is a common skin infestation that can affect all ages but which most commonly affects young patients. Other well-established risk factors are poverty, numerous family members, and bed sharing. Our main objective was to identify the pediatric population with scabies and identify special features associated with age.

Methods: We reviewed medical registries of patients with scabies admitted in the pediatric dermatology (PD) consultation with referrals from the pediatric department (PED) from January 2015 to December 2016.

Results: A total of 69 patients were included with a mean age of 6.9 years (0.16–17). The patients were divided in 3 groups: 0–2, 2–10 and > 10 years.

The mean number of medical observations previous to PD admittance was 1.93, but children under 10 years old had significantly higher observations (2.2; p = 0.01). The mean number of household members was 2.97, while patients > 10 years (p = 0.001) had a significantly lower number of household members.

The diagnostic agreement between PED and PD was 70.8%, however for those patients < 2 years old the rate was significantly lower (50%; p = 0.016). Regarding treatment, those < 2 years old had the highest rate of treatment failure (45%) compared to children between 2–10 years and > 10 years (34.6% and 26.1%, respectively).

Economic insufficiency was found in 46.9% of the patients, being significantly higher in children < 2 years old (83.3%, p = 0.01). There was a tendency towards a positive association between economic insufficiency and treatment failure (p = 0.069). In addition, the number of household members was also associated with treatment failure (3.83 vs. 2.45; p = 0.023).

Conclusion: Children < 2 years old constitute a vulnerable group for scabies. They have a lower rate of diagnosis agreement between PED and PD, receive a higher number of medical observations, and experience more treatment failures. Numerous family members and economic insufficiency are two other factors that are more prevalent in this particular group. Appropriate medical and social assistance should be provided to increase treatment success.

P 058
RARE MANIFESTATION OF PRIMARY HSV INFECTION; MOTHER-INFANT TRANSMISSION DUE TO FREQUENT KISSING
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Herpes simplex virus type 1 (HSV-1) can cause clinical disease in a wide variety of anatomical locations including the orofacial region, genitalia, liver, lung, eye, and central nervous system. The manifestations depend upon the anatomical site involved, and the length of the clinical episode is based on whether it is a primary infection or a reactivation disease. In children herpetic gingivostomatitis is the most common clinical manifestation of primary inoculation and usually asymptomatic. Rarely different body sites may be involved due to route of exposure. Here we present a case of primary herpes infection of the scalp due to frequent kissing by the loving mother.

An eleven month old baby girl was admitted to our clinic with purulent and oozing lesions on the frontal region of the scalp. On dermatologic examination multiple small, grouped vesicles and crusts on the scalp, and multiple cervical enlarged lymph nodes were detected. Additionally we noticed residual erythema due to herpes infection on the lower lip of the mother and observed her kissing her baby’s hair several times during examination. Tzanck smear of the lesion revealed multinucleated giant cells which is consistent with the herpes infection. Although serology of HSV IgM was negative, oral acyclovir 50 mg/kg/day in divided doses for 10 days was given and the patient recovered in two weeks. Primary HSV-1 infection usually presents as gingivostomatitis in children. However, viral transmission can occur through close contact with lesions. Although rare, extracellular spread of primary HSV infection can be seen as in our case. The diagnosis of HSV-1 infection can be made by a variety of techniques including viral culture, serology, immunofluorescence or PCR. Tzanck smear is an easy method for the rapid diagnosis of HSV infections, especially for atypically located lesions. In the present case, the unusual location and the negative serology in combination with Tzanck smear led us to rapid establishment of the diagnosis and to fast recovery of the lesions.

P 059
PHTHIRUS PUBIS INFESTATION OF THE EYELASHES AND SCALP IN A FIVE YEAR OLD GIRL
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Pubic lice (Pthirius pubis) is transmitted with sexual contact in adults. In children the lice sometimes infect the eyelashes and uncommonly scalp hair. A 5-year-old girl was admitted to our clinic because of the bugs on the scalp which were noticed and photographed by her mother. The patient also had purulent lid margins diagnosed as blepharitis and treated with topical corticosteroids by an ophthalmologist. On dermatologic examination small black points was noticed on the neck and preauricular area. On dermoscopic examination parasites and their nits were seen on the patient’s eyelashes and scalp. The parasites were removed and identified as P. pubis. Investigation of the parents revealed scalp infestation of the mother. The scalp was washed with permethrin 1% shampoo two times, ten days apart. Eyelashes were treated with white petrolatum for 10 days. The attached lice and the nits were removed mechanically by her mother. One month later no P. pubis infestation was found on the scalp or eyelashes. Physicans who are involved with pediatric diseases should be familiar with infestation of the eyelids and other regions of the body with pubic lice.

P 060
PEDIATRIC TINEA CAPITIS IN MALLORCA: OUR EXPERIENCE OVER FIFTEEN YEARS
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Introduction: Tinea Capitis (TC) is a scalp infection caused by dermatophyte fungi, mainly of the genera Microsporum and Tricophyton. It is considered the most frequent mycosis of the pediatric age group, being relatively rare in adults. The characteristics in its clinical presentation, etiological agent, and response to treatment vary depending on the corresponding geographical area.

Material and methods: We undertook a retrospective epidemiological study by reviewing a series of 81 pediatric patients with TC confirmed by a mycological culture, at Hospital Son Llatzer, between 2003 and 2017. Several parameters (age, gender, epidemiological background, etiological agents, clinical form, treatment and clinical response) have been evaluated, aiming to describe the characteristics and the evolution of this fungal infection in our environment.

Results: Of the 81 TC, 64.2% were in boys, and the mean age was 5.16 years. 61.9% were immigrants, particularly from Africa. The most frequently isolated dermatophyte was T. tonsurans, followed by M. canis. The most common clinical presentation was single or
multiple patches of hair loss and/or scaly plaques on the scalp. In the majority of cases, the treatment of choice was griseofulvin, followed by terbinafine. In 28.4% of cases the culture negativization was determined.

**Conclusion:** We report the first study of TC in pediatric patients in our region, comparing our data to those published in the literature of other Spanish series. In addition, we highlight the increasing number of TC cases in the immigrant population in our series, as well as their possible relationship to the epidemiological changes experienced over these 15 years.

P 061

**ACQUIRED ZINC-DEFICIENCY IN A PREMATURE INFANT**

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The authors present the case of an extremely premature male infant who was born at 27 weeks' gestation. He was discharged after 11-weeks of neonatal intensive care. There was no diarrhea or other gastrointestinal disorder, and no failure to thrive in the history of the infant. After one week, small brownish-red, eroded macules and papules with honey-yellow-coloured crusts developed on the cheeks, forehead, nose and ears, and also brown macules, plaques, and bullae appeared on his fingers, hands, wrists, ankles, feet, and toes. Despite multiple topical antibacterial and anti-inflammatory treatments administered to the patient with the supposed diagnosis of impetigo, a significant worsening of the clinical symptoms was observed. Laboratory testing demonstrated an unusually low serum zinc level (4.8 µmol/l), normal range of zinc level for newborn infants is 9.2-13.8 µmol/l) which supported the diagnosis of acquired zinc-deficiency caused by prematurity. The introduction of supplementation therapy resulted in a rapid and significant improvement of the clinical symptoms within days. Molecular genetic testing did not reveal any mutation in the SLC39A4 gene, so the classic form of acrodermatitis enteropathica was ruled out. Whereas a classical, inherited zinc deficiency is fortunately a very rare entity, the acquired type is much more prevalent and poses a major health problem. A number of conditions can lead to an acquired zinc deficiency; an increased requirement for zinc during pregnancy and lactation and in premature newborns, low nutritional intake, malabsorption, or an excessive loss of zinc. Both inherited and acquired forms of acrodermatitis enteropathica are characterized by the triad of dermatitis, diarrhoea and alopecia. Numerous non-derma- tological manifestations can also occur, and chronic zinc deficiency can lead to irritability, other psychological changes, growth or neurodevelopmental retardation, and can result in death. The recommended treatment is prompt zinc supplementation, which leads to a rapid and dramatic improvement in the disorder.

P 062

**CHARACTERISTICS OF ADOLESCENT PATIENTS WITH ACNE VULGARIS: A CROSS-SECTIONAL, POPULATION BASED STUDY**

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Acne vulgaris, one of the most common dermatologic diseases, affects the vast majority of adolescents and young adults. The marked dermatologic symptoms have a great impact on the quality of life and on the psychosocial condition of young patients. Thus adequate therapy poses an important challenge in the dermatological practice. In our retrospective study, the data of patients with acne vulgaris visiting the Pediatric Dermatology Outpatient Clinic at the Department of Dermatology and Allergology at the University of Szeged, were analysed during a 4-year interval. The population was characterized by gender, age, severity and anatomical localization of skin symptoms. Oral and local medication taking habits of the patients and the number of visit failures were also assessed. Between 2012 and 2015, 1,035 young patients (506 males, 529 females, mean age: 15.2 years) were included in the survey. Almost 50% of the participants had moderate or severe acne. The vast majority of the patients (63.7%) belonged to the 15–18 year age group, a significant proportion of them suffered from severe acne vulgaris. According to anatomical localization of the symptoms, the face in 91.2%, the back in 52.5% and the chest in 23.7% of the cases were involved. Significant correlations were found between gender, age, acne severity, and the extension of skin lesions. Unfortunately almost two-thirds of the subjects omitted at least one follow-up visit.

According to our results, a great majority of patients had moderate or severe acne vulgaris or acne conglobata with the involvement of multiple anatomical regions. In such cases, the therapy is also complex and multiple-stage; the use of various local and systemic pharmacological products is frequently required. Thorough analysis of the data of patients with acne is indispensable in order to create a successful treatment strategy and to achieve appropriate therapeutic compliance.

P 063

**PITYRIASIS LICHENOIDES IN CHILDHOOD**

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Introduction: Pityriasis lichenoides (PL) has an unknown etiology and affects all age groups, with 20% of cases occurring in children. It is often classified into the acute (pityriasis lichenoides et varioliformis acuta -PLEVA-) and the chronic form (pityriasis lichenoides continua -PLC). Some authors consider that PLEVA and PLC are two independent entities. Nevertheless, it is widely accepted that PLEVA and PLC are polar ends of a clini- copathological spectrum. PL has been regarded as a benign reactive process, but inflam- matory reports of PL evolving into cutaneous T-cell lymphoma (CTCL) and detection of T-cell clonality have led to the hypothesis that PL is at the benign end of a spectrum of lym- phoproliferative diseases that includes lymphomatosic malignancy.

Objectives: We sought to study the epidemiological and clinical features, and evolution of PL in children followed up in two tertiary referral hospitals in Barcelona.

Methods: A retrospective longitudinal study was carried out. Patients under 18 years old with a histologically confirmed PL from 2006 to 2016 were recruited. Data regarding clinical features, histology, treatment response, development of malignancy, and TCR gene rearrangements were retrieved.

Results: A series of 57 children were analyzed. The median age was 7.5 years old and 67% were boys. Most of the patients (61%) had PLC while the remainder had PLEVA. The most common prescribed treatments were topical corticosteroids for PLEVA and oral ery- thromycin for PLEVA. The average follow-up time was 15 months. Three cases mani- fested skin lesions compatible with mycosis fungoides. In all other cases, a persistent but non-progressive clinical course was demonstrated, characterized by periods of regression and recurrence.

Conclusion: PL generally follows a benign course but it can progress to CTCL. Therefore, a long-term follow-up of patients with active disease is recommended.
P 066

JUVENILE PTYRIASIS RUBRA PILARIS TREATED SUCCESSFULLY WITH CYCLOSPORINE

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Pityriasis rubra pilaris (PRP) is an idiopathic papulosquamous disease with rare occurrence in childhood. The use of retinoids as a first line of treatment in adult populations is widely accepted, however childhood cases of severe PRP remain a therapeutic problem. We present the case of a 4-year-old boy with severe type III PRP triggered by acute viral infection of the gastrointestinal tract. Skin lesions developed within 3 weeks and on examination covered large areas of the trunk, lower and upper limbs, and face with erythrodermic appearance. The classical presentation with follicular papules and plaques strongly suggested the diagnosis and was confirmed by skin biopsy. After literature research we decided to start treatment with 5 mg/kg oral cyclosporine. After 4 weeks the lesions improved significantly and we tapered the dose of cyclosporine to 3 mg/kg for the next two months achieving almost complete clearance of the skin lesions. Juvenile PRP usually has a rapid course and resolves spontaneously within a year, however systemic treatment is necessary in severe cases and in our opinion, cyclosporine could be an alternative to retinoids which often causes problematic side effects in prepubertal patients.

P 067

EARLY MORPHEA PRESENTING AS AN ACQUIRED PORT-WINE STAIN: DESCRIPTION OF THREE CASES

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Morphea in childhood is a relatively rare fibrosing disorder of the skin and underlying tissues. Early skin changes include hyperpigmented, hypopigmented, erythematous, or purplish patches. Port-wine stain is a congenital capillary vascular malformation which presents as a large flat patch of purple or dark red skin with well-defined borders. Rare cases are described of acquired PWS and they are often clinically related to a series of traumatic injuries. We describe three cases of early localized morphea of the face presenting as three different acquired PWS, two of them with important alterations revealed by nuclear magnetic resonance. We want to emphasize that an early clinical and histological diagnosis must be mandatory in all suspected cases, followed by a nuclear magnetic resonance investigation in order to detect possible cerebral involvement. It is important to start the most appropriate therapy as soon as possible, avoiding inappropriate and dangerous treatments such as laser therapy.

P 068

CUTANEOUS PEDIATRIC GRAFT VERSUS HOST DISEASE: A CASE SERIES

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Introduction: Graft versus host disease (GVHD) is the most common co-morbidity in persons treated with allogeneic hematopoietic stem cell transplantation (HSCT). The skin is the organ most commonly affected by GVHD and it is usually the first to be affected. HSCT is less commonly performed in children and studies in which cutaneous GVHD in children is evaluated are rare.

Patients and methods: We retrospectively collected all allogeneic HSCT cases in persons younger than 16 from 1995 to 2017 and searched for cutaneous GVHD. We recorded information on the underlying disease that led to HSCT, the type of transplantation, the degree of GVHD, the degree of involvement of other organs, the prophylactic regimen, and the treatment protocol of GVHD. We focused on the clinical and histopathological findings, when available, of the skin involvement.

Results: We identified 6 cases (4 female and 2 male). Five cases were of non-related donor. Hematologic diseases led to allogeneic HSCT in 5 cases and a metabolic disease in the remaining case. A maculopapular rash, especially affecting the trunk, was the most frequent clinical finding. Two patients exhibited generalized lesions and palms and soles were affected in 2 cases. Histopathology showed an interface dermatitis and five patients showed involvement of other organs (gastrointestinal tract and/or liver). Treatment was topical and systemic corticosteroids. Other therapies were implemented in 4 cases.

Discussion: Clinical findings of cutaneous GVHD are similar in children and adults. Maculopapular rash, with follicular reinforcement, and involvement of the palms and soles are frequent clinical features, and it may evolve into a generalized disease. Muco-cutaneous involvement is common and other organs (liver and gastrointestinal tract) are commonly affected. The initial manifestations of cutaneous GVHD are often subtle and dermatologists must be aware of them so as to implement early treatment.
P 070

A QUESTIONNAIRE SURVEY ON UNDERSTANDING OF ATOPIC DERMATITIS AMONG KOREAN PATIENTS AND CAREGIVERS

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Background: Therapeutic education is important for management of atopic dermatitis (AD). To provide effective therapeutic education, common misunderstandings about AD among patients and caregivers need to be reviewed.

Methods: A questionnaire survey about the course, etiology, and management of AD was conducted for patients and caregivers who visited The Department of Dermatology at Seoul National University Hospital, Seoul, Korea.

Results: A total of 177 subjects participated in the study. A few subjects understood the natural course of AD. Only 34.5% of subjects were aware that AD usually improves with age. Many subjects (52.6%) misjudged relapse of AD symptoms for development of tolerance to topical steroids. 158 (89.3%) subjects believed that enhancement of patients’ immune systems can improve the symptoms of AD. Dietary restriction is considered an essential management strategy (72.9%) and many of them (55.4%) agreed to postpone the elimination of certain foods. Most subjects did not have accurate information about nursing care. In particular, 34.3% of subjects reported that they used only water without any cleanser, and 27.3% agreed that soap made of natural ingredients should be used to avoid the harmful effects of chemical substances. Most subjects (57 of 11,549.6%) obtained information about AD from medical doctors and consider them as the most reliable sources of information (137 of 164, 83.5%). Subjects prefer printed materials (69 of 16,242.6%) to seminars or video-clips for obtaining information.

Conclusion: In this study we found that patients and caregivers have lots of misunderstandings about AD. Therapeutic education about the course, etiology, and management of AD with printed materials made by physicians will be valuable for the effective management of AD.

P 071

IDIOPATHIC FOLLICULAR ABSCESS GRANULOMA (IFAG) - CASE REPORT

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IFAG is a chronic painless erythematous or violaceous nodule(s) that occurs during early childhood. The nodules are typically seen on the cheek, with a few cases presented on the eyelid similar to chalazion. The pathogenesis of this condition is unclear. There are several theories proposed. An initial hypothesis includes an inclusion reaction to scratches. It was also considered an inflammatory manifestation of an embolycytic remnant. The most recent theory is that IFAG represents a form of pediatric rosacea. A 2-year-old female patient with no past medical history of interest was referred to the Pediatric Dermatology Department for examination of the nodule on her left cheek that had been present for 2 months. Parents noted a slow growth for weeks. Dermatological examination showed painless reddish/purple nodule of elastic consistency with the size of 21 x 19 mm positioned on the left cheek (Figure 1). There were no enlarged lymph nodes. Parents were afraid of the scar after a punch biopsy and did not agree to the procedure. Bacterial and fungal culture findings were negative. Ultrasound of the nodule revealed a solid well-delineated hypoechoic lesion without calcium deposits. Cultures for mycobacteria were normal. Direct immunofluorescent test showed discontinuous dusty deposits of IgM whitish scale, mainly on the periphery of the scarring lesion. Routine laboratory analyses were normal. He was treated with oral isotretinoin at 0.7 mg/kg/day, in combination with oral prednisone at 0.7 mg/kg/day with gradual dose tapering over 2 months, and topical corticosteroids.

Two-months of therapy resulted in resolution of erythema and itch. Conclusion: The etiopathogenesis of LPP is unknown, although there is an autoimmune reaction mediated by T lymphocytes occurring in women, sporadically in men, and very rarely in children. Typical age of onset is between 40 and 60 years. According to MEDLINE, until 2017 there were only nine published pediatric cases of LPP (aged 8 to 16 years) and all were older than our patient.

P 072

LICHEN PLANOPILARIS IN A CHILD

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Introduction: Lichen planopilaris (LPP) is an inflammatory disorder involving the follicular apparatus and it represents an important cause of scarring alopecia. LPP is considered a follicular variant of lichen planus. Although most reported cases involve the scalp, cases involving the face, trunk and other parts of the body have also been reported.

Material & methods: We present a 4-year-old Caucasian boy with a 6-month history of pruritic follicular papules, erythema, and scaling leading to hair loss and the development of scarring alopecia. He was initially treated with short-term topical antibiotic and corticosteroid therapy with partial improvement. Shortly after treatment discontinuation there was a relapse with the appearance of new lesions and increasing intensity of pruritus.

Results: On admission the boy had an atrophic-scarring area on the parietal region of the scalp, measuring 7 x 4 cm, with significant hair thinning and perifollicular erythema and white scale, mainly on the periphery of the scarring lesion. Routine laboratory analyses were normal. Direct immunofluorescent test showed discontinuous dusty deposits of IgM along the basement membrane zone. The clinical diagnosis was confirmed by histologic examination, which revealed epidermal atrophy, fibrosis of the dermis, dilatation of the hair follicle lumen and perifollicular lymphocytic and histiocytic infiltrate. The etiology is unknown, although there is an autoimmune reaction mediated by T lymphocytes occurring in women, sporadically in men, and very rarely in children.

P 073

ATOPIC DERMATITIS IN CHILDREN: RELATIONSHIP WITH VITAMIN D AND PHOSPHOCALCIC METABOLISM

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Introduction: Some studies relate atopic dermatitis (AD) to vitamin D (VD) deficiency however, there are few studies about other parameters, like phosphocalcic metabolism, in children.

Objective: Our aim was to investigate the association of AD and AD severity with VD and phosphocalcic metabolism in a pediatric population.

Material and methods: A prospective cross-sectional study was performed in a sample of children diagnosed with AD in San Jorge Hospital, Huesca, and University Hospital Niño Jesús, Madrid, and a group of healthy controls, during 2012 and 2013. The following variables were recorded: age, sex, weight, height, AD severity, and serum levels of VD, parathyroid hormone (PTH), calcium (Ca) and phosphorus (P). AD severity was determined according to SCORAD index.

Results: 239 participants of both sexes were included, aged 0 months to 14 years: 134 diagnosed with AD and 105 healthy controls. Mean VD in the control group was 24.1 ng/ml (SD 10.2) and 28.5 ng/ml (SD 11.5) in AD group, decreasing with AD severity: 32.5 ng/ml (SD 12.3) mild AD (n = 21), 28.2 ng/ml (SD 11.0) moderate AD (n = 89) and 26.2 ng/ml (SD 19.6) severe AD (n = 24) (p < 0.05). Mean serum levels of PTH were higher in controls (31.0 pg/ml, SD 20.6) than in children with AD (25.8 pg/ml, SD 15.3) (p < 0.05). However, PTH levels increase with AD severity: 22.9 pg/ml (SD 12.9) mild AD, 25.6 pg/ml (SD 14.8) moderate AD and 29.2 pg/ml (SD 19.6) severe AD (p < 0.05). PTH was significantly increased in children with deficient serum VD level (35.6 pg/ml, SD 20.6) vs. sufficient serum VD level (27.0 pg/ml, SD 15.9) (p = 0.039). P serum levels significantly decrease with AD severity: 5.7 mg/dl (SD 1.4) mild AD, 5.4 mg/dl (SD 0.8) moderate AD and 5.0 mg/dl (SD 0.8) severe AD (p = 0.014). No association was found between P and VD serum levels.

Conclusion: Severity of AD is inversely related to VD and P serum levels and directly related to P serum levels.

P 074

CUTANEOUS LESIONS UPON DYNAMIC DEVELOPMENT OF SYSTEMIC LUPUS ERYTHEMatosUS IN A 6 YEAR-OLD GIRL

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Introduction: Juvenile-onset systemic lupus erythematosus (JSLE) is one of the most severe and chronic autoimmune diseases in children. Clinicians frequently base diagnosis of JSLE on the fulfillment of at least 4 of the 11 criteria of the American College of Rheumatology (ACR) guidelines.

Case report: A six-year-old Algerian girl was admitted to our hospital with a six-month history of intermittent fever, asthenia, anemia, hepatoplenomegaly, and multiple palpable
PERIODIC PROXIMAL NAIL SHEDDING (SPORADIC ONYCOMADESIS) IN A CHILD

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Proximal nail shedding (onycomadesis) is the spontaneous separation of the nail plate from the bed, beginning from the proximal end with often, but not always, shedding of the nails as the new nail grows. There have been very rare cases reported of idiopathic onychomadesis not associated with any disease, drugs or familial autosomal dominant inheritance. A 9 yr-old boy was referred to our dermatology department for the presence of proximal nail shedding which appeared first at 5 years of age. The parents observed that this shedding has been happening periodically in all of the fingernails. No family history was noticed. Dermatologic examination revealed onychomadesis involving two fingernails on the right and three on the left. As the nails grew this detachment become prominent. The known causes of onychomadesis may be classified as systemic and dermatologic disorders, exposure to some drugs, trauma, fever, and infections. Familial cases have also been reported. Related disorders are serious systemic illnesses, high fever, Stevens-Johnson Syndrome, Kawasaki disease, infections (streptococcal infections, hand-foot-mouth disease (HFMD), and measles), severe emotional distress, systemic lupus erythematosus, and penileontal dialysis. There are rare cases reported of idiopathic onycomadesis not associated with any disease or drug therapy, which presented in a familial autosomal dominant pattern. To the best of our knowledge there have been only 2 cases reported of idiopathic periodic onychomadesis. The present case report describes the third idiopathic, non-familial and periodic onychomadesis. It is one of the most rare forms with its periodicity. No treatment is required for onychomadesis, and referral to further specialists is unnecessary. Patients should be reassured that spontaneous regrowth of a new normal nail is usually seen within a few months.

DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS) IN A COHORT OF ASIAN CHILDREN

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Introduction: Drug reaction with eosinophilia and systemic symptoms (DRESS) is rare in children and potentially fatal. Fever and rash, salient features of the condition, mimic other commonly encountered paediatric conditions making diagnosis challenging. We profiled the DRESS cases in a tertiary care children’s hospital in Asia.

Methods: All DRESS patients diagnosed within 2006-2016 had their medical records analysed. Data on demography, drug exposure, laboratory tests, clinicopathological manifestations, and treatment were assessed.

Results: Ten patients, aged 4-16 years old were diagnosed with DRESS within the 10-year study period. Six were male. Common drugs implicated were anticonvulsants and antibiotics (e.g. trimethoprim-sulfamethoxazole (3 cases), carbamazepine (2 cases), and phenobarbitone (2 cases). All patients presented with fever and pruritic exanthema. Desquamation (2 cases), purpura (1 case), and oral mucositis (5 cases) were also observed. Lymphadenopathy, hepatomegaly, and facial edema were also common. There was livor mortis in 4 cases, but none progressed to liver failure. Seven had eosinophilia and 9 had atypical lymphocytosis. Other laboratory abnormalities included low hemoglobin (7 cases), thrombocytopenia (3 cases), and prolonged coagulation time (2 case). Five patients had skin biopsies showing superficial perivascular lymphohistiocytic infiltrates. All patients received systemic corticosteroids of varying durations and dosages, with 2 receiving pulsed methylprednisolone. Systemic steatorrhea were weaned after 19 days-4 months (Mean: 65 days). Disease resolution, with normalisation of liver enzymes occurred in 28-90 days (Mean: 44.1 days). One patient developed TRAb- hyperthyroidism 6 months after resolution of DRESS, another developed chronic urticaria 4 months after resolution of DRESS.

Conclusion: Physicians should have a high index of suspicion for DRESS to ensure the inciting drug is discontinued and treatment started expediently. Liver involvement is very common but responds well to treatment with systemic steroids. However, there is as yet no standard dose and duration for children.

HYDROVAC VACCINFORM WITH MUCOUSAL INVOLVEMENT: ORIGINAL OBSERVATION

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Introduction: Hydrovacciniform (HV) is an acquired, idiopathic, and sporadic photodermatosis. It begins in childhood and often spontaneously heals in adolescence. We report a new case of HV with mucosal involvement.

Observation: A 13-year-old child presented from the age of 8 with rashes of vesicular and crustless, painless, and non-pruriginous lesions located in the photo-exposed areas. The lesions evolved towards atrophic and hyperpigmented scars. Four months before, the patient presented ear lesions and painful ulcerations of the lower lip following a prolonged photoprotection without photoprotection, without ocular signs. The clinical examination found an erosive and crusty cheilitis, necrotic vesiculo-bullous lesion with an umbilical center on the helix, and variciform scars on the nose and cheeks. The level of uric acid was normal. The cutaneous histopathology showed an irregular hyper-orthokeratotic epidermis and a lymphocytic infiltrate in the dermis. The diagnosis of a photodermatosis type HV was retained, the patient was treated with hydroxychloroquine and acyclovir associated with an external photoprotection with a partially favorable evolution.

Discussion: Mucosal involvement in HV is exceptional. It affects the lips, the tip of the tongue, and the eyes, all potentially phototoxic. It is a type of conjunctivitis and corneal ulcerations in the eye and necrotic lesions in the lip. This case report illustrates that HV can affect mucous membranes. It is therefore essential to recognize them to avoid delayed diagnosis, which could result in dire consequences.
P 079

CHRONIC SPONTANEOUS URTICARIA: A FRENCH PEDIATRIC COHORT
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Introduction: Pediatric chronic spontaneous urticaria (CSU) is a rare and scarcely known disease, associated with significant quality of life impairment. To date, management is mainly extrapolated from adult data. Highlighting pediatric specificities is therefore essential. We describe the pediatric CSU cohort of the French Reference Centre for Angioedema (CREAK). All patients diagnosed with CSU and under 18-years-old at diagnosis were included, excluding patients with acute urticaria or a differential diagnosis of CSU.

Methods: This is a retrospective study set at the Grenoble-Alpes University Hospital, French Reference Centre for Angioedema (CREAK). All patients diagnosed with CSU and under 18-years-old at diagnosis were included, excluding patients with acute urticaria or a differential diagnosis of CSU.

Results: Thirty-seven patients were included, 70% female. Mean age at first symptom was 6.4 years. The average diagnosis delay was 39.4 months. Family history of atopy was reported in 80% of cases, dysimmunity in 66%. Thyroid dysfunction was frequent (61%). Family history of CSU and angioedema (AE) were frequent (43% and 40% respectively). Personal history of atopy (46%) and dysimmunity (5%) were observed. The majority of CSU crises included AE (69%) and 30% presented as isolated AE. Inducible urticaria was prevalent (65%), mainly cholinergic urticaria. A trigger was found in 84%, mainly physical. The majority of CSU was controlled by single (51%) or double dose (43%) of H1-antihistamines. In some cases, anti-leskutinones were necessary (43%). One refractory CSU was controlled with omeprazolum.

Conclusion: As in adults, pediatric CSU was more frequent in females and associated with personal/familial features of atopy/dysimmunity. Notable features in our cohort were the significant diagnostic delay and the frequent association with family history of CSU or AE. The frequency of AE in our cohort is probably high due to a referral bias. Therapeutic management is extrapolated from adult guidelines. Specific pediatric CSU assessment tools for disease activity and quality of life are needed to lead the prospective studies that will help define specific pediatric management.

P 080

DEVELOPMENT OF COLOR COSMETIC FOR ATOPIC DERMATITIS PATIENTS
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Because of diminished skin barrier function atopnic dermatitis patients are at risk of developing contact sensitization. Chronic exposure to contact allergens in cosmetics increases the risk of not only allergic contact dermatitis but also atopic dermatitis. In previous studies hypoallergenic cosmetic products marketed to American children were frequently found to contain contact allergens.

We therefore evaluated the pigmented effect and safety of natural color ingredients incorporated with foundation which may be substituted for the color foundations already marketed.

To determine the safety of natural color ingredients MTT assay was done with cultured human skin fibroblasts. Patch test for natural color ingredients was performed on normal persons. To determine the conditions to successfully formulate natural color ingredients for foundation we conducted stability evaluations including centrifugation, cycling test, viscosity measurement, and pH measurements.

We found that the higher concentrations of all foundations, the lower cell survival rate was seen. Cell survival rate of natural color ingredients was higher than that of control. No reactions to natural color foundations were observed from patch tests. There was one positive reaction to marketed foundations. The ranges of the pH were 6.22 – 6.86 in the natural color foundations.

These patients present a papular eruption on the extensor surface of the limbs and on the back. Pruritus is a constant feature. This form seems to be more common in childhood and in patients with dark skin. Histologically, spongotic dermatitis was found rather than a lichenoid pattern. This presentation may be a result of rubbing and

P 081

ATYPICAL-ONSET PSORIASIS LIKELY TRIGGERED BY TINEA INCINTOGO IN THE SETTING OF UNDERLYING ECZEMA
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A 10-year-old boy with long-standing eczema presented with an 18 month history of an enlarging, well-defined, red, scaly, left periorbital plaque that had not responded to pro-longed topical ointment treatment. He had contact with cats for six months before its onset. As tinea incognito was suspected, griseofulvin was commenced for a month, during which time the plaque regressed to become herpetiform vesiculo-papules. HSV PCR was negative. Griseofulvin was discontinued and erythromycin started, leading to plaque regression to a psoriasisiform morphology. Skin biopsy showed parakeratosis with admixed neutrophils and focal, mild spongiosis. After a second course of griseofulvin, a near-complete clinical resolution of the rash was observed. During this time, he developed another psoriasisiform rash on the right post-auricular region that was minimally pruritic. A skin punch biopsy revealed alternating orthokeratosis and hyperkeratosis, mild psoriasis-form epidermal hyperplasia, and patchy mild spongiosis, consistent with a diagnosis of psoriasis.

Futhermore, a follow-up full skin examination revealed new onycholysis of the right thumb and index fingers, consistent with nail psoriasis. He was started on topical tacrolimus for the residual rash on the left periorbital area, as well as methylprednisolone aceponate 0.1% fatty ointment for the right post-auricular rash, and mometasone furoate 0.1% cream for the affected nails. Risk factors for psoriasis are numerous, including but not limited to UV exposure, medications, smoking, stress, and infections. Exacerbated autoimmune responses and breakdown of immune tolerance triggered by infections in susceptible individuals can induce psoriasis. Tinea incognito masquerading as psoriasis has been well documented in the literature, but there have been no published reports to date of tinea incognito inducing psoriasis. However, given the overlap in immunological pathways involved in superficial dermatophytosis, atopic dermatitis, and psoriasis, in particular Th17 cell activation, a tinea incognito-induced psoriasis may be a possibility.

P 082

CONTACT ALLERGY AND SKIN EXPOSURES IN ADOLESCENTS: A POPULATION-BASED STUDY
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Background: Knowledge about skin exposures and the relation to contact allergy in adolescents is limited.

Objective: To investigate the prevalence of piercing, hair dyeing and tattooing in 16-year-olds, and to explore the association with contact allergy assessed by patch test.

Methods: At age 16, 3,115 adolescents of the population-based BAMSE cohort first answered a questionnaire, including questions on exposure to various skin sensitizers. 2,285 were then patch tested with self-administered TRUE2 test panels with 30 substances. The test results were assessed and photographed by a nurse on day 2. Dermatologists made the final assessment based on photographs and protocols. Associations between skin exposures and contact allergy were tested by logistic regression.

Results: Contact allergy to any of the tested substances was common (all 15.3%, girls 17.0%, boys 13.4%, p = 0.018). Allergy to nickel was most frequent (all 7.5%, girls 9.8%, boys 4.9%, p < 0.001) followed by fragrance mix I (all 2.1%), p-phenylenediamine (all 1.1%), and other substances.

Piercing was the most common skin contact exposure and was reported by most girls (all 55.4%, girls 62.7%, boys 16.5%, p < 0.001). Hair dyeing was common, with a predominance of girls (all 50.1%, girls 77.4%, boys 21.8%, p < 0.001). Tattooing was less frequent (all 2.4%, girls 2.9%, boys 1.8%, p = 0.061).

Piercing was associated with an increased OR for contact allergy to nickel (adj. OR 1.77, 95% CI 1.04-3.00). Tattooing was likewise associated with an increased OR for nickel allergy (adj. OR 2.34, 95% CI 1.02-5.39).

Conclusion: Skin-sensitizing adornment behaviors like piercing, hair dyeing, and tattooing expose adolescents to contact allergens. Such exposure may result in contact allergy, allergic contact dermatitis, and future hand eczema.
An 8-month-old boy with known AD was referred to the pediatrician with facial and head scratching, thus making it similar to the frictional lichenoid eruption but this one generally arises on pressure points in a more limited distribution. AD treatment has been proven to be effective.

Conclusion: AD has less common clinical variants. PLAD is an atypical presentation with complicated AD. Eczema herpeticum (EH) occurs when there is a secondary skin infection. Atopic Dermatitis (AD) is often associated with psychological problems, disrupting the quality of life of patients and caregivers. A multidisciplinary approach has been shown to reduce disease severity and improve quality of life.

P 085

DIAGNOSIS AND THERAPY OF COMPLICATIONS IN A CHILD WITH ATOPIC DERMATITIS

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Atopic Dermatitis (AD) is a common disease in children. Several diagnoses can coexist with complicated AD. Eczema herpeticum (EH) occurs when there is a secondary skin infection with herpes simplex virus (HSV). Accurate diagnosis is vital but the clinical presentation is difficult to distinguish from secondary bacterial infection (SBI). An 8-month-old boy with known AD was referred to the pediatrician with facial and head scratching, thus making it similar to the frictional lichenoid eruption but this one generally arises on pressure points in a more limited distribution. AD treatment has been proven to be effective.

Conclusion: AD has less common clinical variants. PLAD is an atypical presentation with complicated AD. Eczema herpeticum (EH) occurs when there is a secondary skin infection. Atopic Dermatitis (AD) is often associated with psychological problems, disrupting the quality of life of patients and caregivers. A multidisciplinary approach has been shown to reduce disease severity and improve quality of life.

P 086

CUTANEOUS MANIFESTATIONS IN CHILDREN WITH MICROSCOPIC POLYANGITIS AND GRANULOMATOSIS WITH POLYANGITIS (WEGENER'S GRANULOMATOSIS)

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Introduction: Antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitides are rare diseases in children.

Results: We report clinical, histological and serological characteristics of 11 children (10F, 1M, aged 12.0 ± 2.6 years, range: 5-15) with microscopic polyangiitis (MPA), and 4 patients (3F, 1M, aged 10 ± 3 years, range: 5-16) with granulomatosis with polyangiitis (GPA), who were diagnosed during a 20-year period. 8/11 MPA patients had cutaneous manifestations at presentation: 7/11 had palpable purpura, while cutaneous necrotizing vasculitis was present in 3/11 patients. All 4 children with GPA had cutaneous manifestations at presentation: 2 had necrotizing vasculitis, one had palpable purpura, and one had right upper eyelid edema and infiltration and proptosis caused by extracellular pseudotumor, initially histologically misdiagnosed as orbital infundibular granulomatosis (Gp4-related disease). All MPA patients had perineurial (p) ANCA specific to myeloperoxidase (MPO), while 3/4 GPA patients had cytoplasmic (c) ANCA specific to proteinase 3 and 1/4 GPA patients had pANCA specific to MPO. Patients were followed for 3 to 12 years. All 19 patients were treated with standard immunosuppressive therapy (cylophosphamide and methyprednisolone pulses/with gradual tapering of corticosteroid dose and introduction of mycophenolate mofetil, azathioprine or methotrexate. None of our patients died. Three of 11 MPA patients who had acute renal failure (ARF) progressed to end stage renal disease. ARF, necrotizing vasculitis and central nervous system involvement at presentation were parameters of poor outcome in MPA. In GPA patients, as opposed to those patients with skin vasculitis and glomerulonephritis, upper-airway and orbital inflammation were resistant to immunosuppressive therapy.

Conclusion: Our report emphasizes that children presenting with cutaneous vasculitis, chronic eyelid swelling, sinuses, hoarseness, and/or hematuria/proteinuria should be tested for ANCA. Early diagnosis and timely immunosuppressive treatment enables a better prognosis of MPA and GPA in children.

P 087

JUVENILE DERMATOMYOSITIS PRESENTING AS DEEP SKIN ULCERATIONS AS AN INITIAL MANIFESTATION

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Juvenile dermatomyositis is a common idiopathic inflammatory myopathy in childhood and is characterized by symmetric proximal muscle weakness and specific cutaneous manifestations. We here report on a 12-year-old boy who presented with violaceous maculopatches with multiple deep skin ulcerations on the right arm and lower back 4 months ago. One month after the initial visit he had difficulty climbing stairs and elevating his arms above the head. Laboratory tests showed elevation of the serum levels of skeletal muscle enzymes, and electromyogram and muscle biopsy indicated myopathy. Histopathological analysis of the ulceration indicated necrotizing vasculitis and calcification. Based on these findings his condition was diagnosed as juvenile dermatomyositis. We believe that this case is unique because of the difficulty in diagnosing the condition, as skin ulceration is a rare cutaneous manifestation of dermatomyositis.

P 088

ANCA NEGATIVE SYSTEMIC VASCULITIS IN AN ADOLESCENT BOY WITH CUTANEOUS, PULMONARY, AND NEUROLOGICAL INVOLVEMENT

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Introduction: Vasculitis is an inflammatory destructive process affecting blood vessels. Henoch-Schönlein purpura and Kawasaki disease, the two most common types of systemic vasculitis in childhood, are diagnosed based upon a particular pattern of characteristics and symptoms. Other types of vasculitis may present in a less obvious manner.
with either incomplete or overlapping diagnostic features. Many children with vasculitis have an "unclassifiable" disease.

Clinical case: A 12-year-old boy presented with painful swelling on the lower extremities, fever and development of cutaneous necrotic lesions and subcutaneous nodules in a few hours. Pustular acroeritiform lesions appeared on the face and vesiculopustular lesions on the extremities. With suspicion of systemic vasculitis, antibiotic and prednisone 1 mg/kg/day were started. After 3 days of treatment the patient worsened and was moved to the reference hospital and a massive alveolar hemorrhage was diagnosed. He has received methylprednisolone bolus, cyclophosphamide, intravenous immune globulin, infliximab and mycophenolate mofetil with good evolution. The patient developed peripheral neuropathy with strong pain and has required fentanyl patches, morphine and gabapentin. ANCA have been repeatedly negative. Skin biopsy demonstrated necrotizing vasculitis in deep vessels with strong neutrophic infiltration.

Discussion: In our case we have considered different diagnoses such as polyarteritis nodosa, Behcet disease, microscopic polyangiitis, WADENGER disease, and ADAMI deficiency. With no clear diagnosis but a severe disease, the patient has received aggressive treatment, including infliximab, with good evolution. Currently he is receiving maintenance therapy with mycophenolate mofetil.

Conclusion: The first priority in management of childhood vasculitis is prompt recognition and timely treatment as many of the vasculitides can be severe and life-threatening conditions if not appropriately managed. Biologic agents been beneficial in small studies of various pediatric systemic vasculitides and are increasingly being used.

**P 089**

**HIDRADENITIS SUPPURITIVA (HS) IN CHILDREN: A REPORT OF TWO CASES**

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Background: Prepubertal HS is a rare condition and few cases have been reported. HS pathology is centered on the follicular unit and involves aberrant cutaneous cellular immunity. Early onset of the disease is more common in children with a positive family history and hormonal changes, primarily androgen excess.

Cases presentation: At age 8 years, patient 1 developed tender red nodules in the groin region that progressed to abscesses. On her first visit to our clinic (age 10 years 7 months) she presented with a nodule, fibrotic scars, and double-headed comedones in the anogenital region. At this time, early signs of pubic and axillary hair were evident, but no other clinical evidence of androgen excess. An endocrinological examination yielded results within normal limits, and a bone age determination was also compatible with her age. Her father reported a positive history of HS.

Patient 2, an 11 year old girl, reported lesions since she was 9. On her first visit to our clinic she presented with three tender erythematous nodules, double-headed comedones, and scars involving only the groin. She had no breast development and no pubic or axillary hair. Bone age determination and endocrinological laboratory values were normal. No relevant family history was reported.

Conclusion: To our knowledge very few cases of prepubertal HS have been reported where disease was present in the absence of endocrine abnormalities or androgen excess, as in our cases. For that reason it is important to perform an endocrinological examination on all patients of this age. Furthermore, management of HS in children may be difficult as treatment of this age group has not been well studied.

**P 090**

**CASE REPORT: ASSOCIATION BETWEEN HALO NEVI AND MORFEA**

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Halo nevi and morfea (localized scleroderma) are two distinct entities of unknown etiology. Halo nevi are common benign skin lesions that represent melanocytic nevi in which an inflammatory infiltrate develops, resulting in a zone of dermopigmentation surrounding the nevus. Halo nevi are most commonly found in children and adolescents. Morfea is a rare fibrosing disorder of the skin and underlying tissues characterized by skin thickening and hardening due to increased collagen deposition. Halo nevi can be associated with other autoimmune disorders, such as vitiligo and Hashimoto thyroiditis. The association between vitiligo and localized scleroderma has been reported infrequently. We will present co-occurrence of halo nevi and localized scleroderma. A 7 year old boy with multiple halo nevi on the back and localized scleroderma around all halo nevi is presented. Personal and family medical history was negative for any significant diseases. The skin biopsy specimen from nevus and erythematous halo confirmed the diagnosis of halo nevi and morfea. Complete blood counts, urine analysis, hepatic and renal function tests were normal.

Screening for autoimmune disease and antihypertensive medications was negative. The reported association might provide additional information useful to understanding these disorders.

**P 091**

**ALOPECIA AREATA IN INFANCY: CASE REPORT AND REVIEW OF THE LITERATURE**

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Introduction: Alopecia areata is a chronic autoimmune disease of uncertain pathophysiology. It is the most frequent cause of alopecia in children but it is exceptional in the neonatal period and in infancy, hence the interest of our case report.

Case report: Our patient is a 14-year-old child with no significant pathological history except a similar case in a paternal aunt who consulted for total loss of body hair occurring 40 days after birth, for which she has been treated without any improvement. Dermatological and dermoscopic examination confirmed the diagnosis of universal alopecia areata. Laboratory tests were normal except a moderate anemia. Mini-pulses of oral corticosteroids and minoxidil and iron supplementation were introduced. A slight regrowth of hair on the frontal edge was noted at 6 months follow-up.

Discussion: Alopecia areata is the most common cause of alopecia in children. It can occur at any age, but only in 11 to 23.9% of cases before 16 years. Our patient presents a universal alopecia areata in infancy, which is exceptional and whose origin remains debatable: congenital, post-vaccination or other (infectious, medicinal). After elimination of the infectious and medicinal etiologies, our discussion remained open on the congenital or post-vaccination origin, or on a genetically predisposed ground. Up to now, 7 cases of congenital alopecia areata have been reported in the literature. In this case, the alopecia plate was already present at birth and the response to the treatment is variable. Post-vaccination alopecia is also exceptional and only a few cases have been reported. Vaccination constitutes a triggering factor of autoimmune diseases defining a new syndrome termed autoimmune/inflammatory syndrome induced by adjuvants.

Conclusion: Newborn and infant alopecia areata is exceptional. Congenital or post-vaccination forms should be suspected and eliminated first. There is no therapeutic consensus of these particular entities and their prognosis remains poor.

**P 092**

**FAMILIAL JUVENILE SYSTEMIC LUPUS: ABOUT TWO FAMILIES**

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Introduction: Juvenile systemic lupus is a rare connective tissue disease that is more severe in children than in adults. It is often sporadic and familial cases are exceptional (10-15% of reported cases). Few studies of juvenile familial systemic lupus have been reported, hence the interest of this work.

Case reports: We report the case of two families affected by juvenile systemic lupus: 14 and 7 year old sisters in the first family, 20 and 6 year old brothers and a 10 years old sister in the second family. The children of the two families came from a consanguineous marriage of first and third degree respectively. No autoimmune disease history was noted in the two families. The diagnosis of systemic lupus was made according to the new criteria of the Systemic Lupus International Collaborating Clinics group (SLICC). Two children (one from each family) also had dapsone lupus. All patients were placed under photoprotection, and given topical corticosteroids and antimarial drugs, except in two cases where systemic corticosteroids were administered. The evolution was favorable in the majority of cases.

Discussion: Juvenile systemic lupus is a rare entity and a family context may be associated in very few cases. The sex ratio between girls and boys appears to be lower than in adults. Clinically, the initial manifestations are polymorphic and sometimes misleading. The cutaneous and mucosal signs may be specific or not. The most frequent biological abnormalities are proteinuria, haematuria, haematological disorders, and hypoalbuminemia. Therapeutic management should be multidisciplinary. In the most recent series, approximately 80% of patients survived for ten years under adapted treatment. Our results are consistent with those of the literature.

Conclusion: Familial juvenile systemic lupus is a rare entity, with more severe evolution than in adults. The form of onset may be misleading, hence the value of seeking familial forms in all cases of juvenile lupus in order to start an early management and avoid complications.
P 093
PHYTOPHOTODERMATITIS IN CHILDREN: 3 CASES
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Introduction: Phytophotodermatitis is a non-immune cutaneous reaction caused by sequential exposure to photosensitizing substance-containing plants followed by ultraviolet light. The authors present three cases of phytophotodermatitis.

Description: Case 1. A 2-year-old girl presented with brownish linear and macular lesions on her chest, arms, and legs one day after the application of a cream containing rue (Ruta graveolens) for the treatment of Pediculosis capitis. They were exposed to the sun after the application of this product and there was no reference to erythema or blistering before the hyperpigmentation. Photoprotection was recommended. After six weeks later a mild hyperpigmentation was still observed.

Case 2. A 4-year-old girl presented with a painless and nonpruritic, erythematous, and bulous rash on her left arm. On the previous day she had contact with lime juice during an outdoor party. The child was treated with systemic and topical steroids with instructions to avoid exposure to sunlight. After four weeks residual hyperpigmentation remained.

Discussion: Careful history taking is important to identify this photosensitive reaction. Although it is common in adults, its diagnosis may be challenging in children due to the variety of clinical presentations and not always easily identified trigger exposure. It is important to be aware of phytophotodermatitis because it may be misdiagnosed as other skin conditions including fungal infection, cellulitis, allergic contact dermatitis, and even child abuse.

P 094
CUTANEOUS POLYARTERITIS NODOSA WITH MANIFESTATIONS OF LIVEDOID VASCULOPATHY IN A 10-YEAR-OLD GIRL: A CASE REPORT
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Background: Livedoid vasculopathy (LV) is an uncommon disorder of unknown aetiology, characterized by recurrent ulcerations of the lower extremities, healing with atrophic blanche and livedoid hyperpigmentation. Cutaneous polyarteritis nodosa (CPN) is a rare medium-sized necrotizing vasculitis, occasionally presenting as ulceration resulting in ivory-white stellate scarring on the lower limbs.

Objective: We present the case of a 10-year-old girl who showed clinical and histological features of both disorders.

Methods: A 10-year-old girl with no past medical history and no systemic symptoms was admitted to our clinic due to a 2-year history of outbreaks of painful ulcerated lesions on the lower limbs that disappeared leaving white atrophic scars. Dermatological examination revealed hyperpigmented stellate macules and ulcerated indurated erythematous papules with serosanguineous crusts on the ankles and pretilial regions.

Results: Laboratory results showed elevated erythrocyte sedimentation rate and decreased levels of serum complement. Laboratory screening tests for vasculitis and thrombophilia as well as basic laboratory tests were unremarkable. Histopathological examination of skin biopsy showed fibrin deposition within both the wall and the lumen of affected dermal blood vessels. Direct immunofluorescence was negative. The disease healed without recurrence in 3 months follow-up.

Discussion: CPN and LV can show an identical clinical picture. Furthermore, the histopathological examination of skin biopsy in CPN may show identical features as described in LV. C-reactive protein and erythrocyte sedimentation rate are important diagnostic tools. Treatment with oral immunosuppressives was effective.

P 096
URTICARIAL VASCULITIS FOLLOWING A NOVEL VACCINE AGAINST MENSINGOCOCCAL SEROGROUP B
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Case report: A 6-year-old female presented with multiple dusky erythematous to violaceous, annular and acrocircumferential wheals over the posterior aspect of her lower limbs, and joint stiffness in her ankle. There was no history of drug intake or previous infection, but she had received meningococcal serogroup B vaccine (4CMenB, Bexsero®) 7 days before. An extended workup, including hemogram, biochemistry, urine analysis, anti-peptide antibodies, and complement studies was unremarkable. The histopathology of a recent lesion was consistent with leukocytoclastic vasculitis with an inflammatory infiltrate with neutrophils and eosinophils. Direct immunofluorescence was negative. The wheals lasted for more than 24 h and postinflammatory hyperpigmentation continued to appear. Relative rest and antihistamines resulted in resolution of the lesions after two weeks without recurrence in 3 months follow-up.

Discussion: The 4-component meningococcal serogroup B vaccine (4CMenB®) is the first vaccine against this serogroup. Clinical studies have shown a higher incidence of fever and other local reactions than other paediatric vaccines. Urticarial vasculitis (UV) is an uncommon form of leukocytoclastic vasculitis, characterized by erythematous wheals that clinically resemble urticaria, but last more than 24 h and usually resolve with residual pigmentation. Paediatric cases are rare. Although the cause of urticarial vasculitis is unknown it has been associated with drugs, infections, and physical factors. Whether vaccination is considered to be a promoter of vasculitis is controversial. In spite of numerous published cases, higher quality studies failed to prove a causal relationship between immunization and vasculitis. Reports of vasculitis as an adverse event following immunization seem to be more frequent in children, with a female preponderance and a time to onset within 10 days. UV following meningococcal serogroup B vaccination has not been previously reported. Given its recent commercialization, we consider it fundamental to report the possibility of these, and other possible adverse skin events, after its administration.

P 097
ULCERO-NECROTIC MUCHA-HABERMANN DISEASE MIMICKING KAWASAKI SYNDROME
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Introduction: Phthisyasis lichenoides represents a spectrum of inflammatory skin disorders encompassing acute (Pityriasis lichenoides et varioliformis acuta, PLEVA) and chronic (Pityriasis lichenoides chronica, PLC) variants. Febrile ulceronecrotic Mucha-Habermann disease (FUMHD) is a rare and fulminating variant of PLEVA characterized by severe, fever ulcerative and necrotic skin lesions, and systemic involvement.

Case report: We report the case of a 9-month-old infant initially presenting with persisting fever, elevated inflammatory markers, and a maculopapular rash accompanied by mucosal involvement without signs of an underlying infectious disease. Baseline echocardiography was unremarkable. The diagnosis of incomplete KD was made and treatment with intravenous immunoglobulins and acetylsalicylic acid initiated. However, the patient remained febrile and skin and mucosal lesions deteriorated and quickly developed into papulovesicular and ulcerative papules and plaques with predominantly acral involvement. On repeat echocardiography there was evidence of coronary arteritis. A skin biopsy revealed a lichenoid dermatitis with degeneration of the basal layer, multiple apoptotic keratinocytes and a dense predominantly CD8+ lymphocytic infiltrate, consistent with Phthisyasis lichenoides. Based on all findings, FUMHD was diagnosed. Treatment was switched to methotrexate, and initially methylprednisolone and etoricoxib. This was immediately followed by rapid improvement of fever, inflammation markers, skin and mucosal lesions and finally resulting in post-inflammatory hyperpigmentation only. Coronary arteritis also completely resolved without any sequelae.

Discussion: This is the first report of FUMHD mimicking Kawasaki disease (KD). This case illustrates that FUMHD should be considered as a differential diagnosis to KD and Stevens-Johnson syndrome when facing children with rapidly progressive exanthema with mucosal involvement accompanied by systemic symptoms. We therefore recommend
maintaining a low threshold for performing a skin biopsy in sick children presenting with atypical rash. While we cannot exclude a mere coincidence of KD and FUMHD, the clinical course argues against the KD. FUMHD responds readily to systemic treatment with methotrexate.

A 5 month-old infant of Polish origin was referred to Dermatology with a rash that started at two months of age in the groin, progressing slowly to involve scalp/trunk/lims. Aside from episodic flushing when warm associated with itch, she was otherwise asymptomatic. Examination revealed multiple asymptomatic papulonodular lesions with a yellow hue in an otherwise well appearing infant. Skin biopsy showed features in keeping with cutaneous mastocytosis supported by immunohistochemistry (positive CD43, tryptase, CD117, CD68; negative CD1a, S100, CD5). The clinical findings supported by histological features were in keeping with the xanthelasmoid variant of urticaria pigmentosa. Given the absence of organomegaly clinically and radiologically and that she was well, a bone marrow biopsy was not indicated. Serum mast cell tryptase was elevated (43.9 microgram/l). This continued to increase until 20 months of age (58.6 microgram/l) corresponding to development of new lesions followed by a marked reduction (25.7 microgram/l)-corresponding to improvement in clinical appearance) at 28 months of age. Apart from a stable lymphocytosis, blood tests were otherwise unremarkable. An insignificant improvement in frequency of flushing was noted with non-sedating antihistamines (cetirizine, desloratidine), ranitidine, and montelukast. Mastocytosis can be classified as cutaneous or systemic. Cutaneous mastocytosis of the xanthelasmoid variant is rare and usually presents with yellow papulonodular lesions resembling xanthomas. The term 'xanthelasmoida' was coined by Fox in 1875 and between 1875-1883, 19 cases were reported. The most recent reported cases of xanthelasmoid mastocytosis were in 2011 and 2001. Treatment is the same as for other patients with cutaneous mastocytosis, including the avoidance of drugs that cause mast cell degranulation. As with other childhood mastocytosis, gradual improvement is anticipated, but may take longer. Interestingly, Husak et al. stated that compared to childhood variants of mastocytosis, there were no differences reported regarding development of systemic involvement or malignant transformation.

P 098

APLASIA CUTIS CONGENITA ASSOCIATED WITH FETUS PAPYRACEUS IN A TRIGEMINAL DICHRONIC TRIAMIOTIC PREGNANCY


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We report the case of a late preterm twin who presented with stellate atropic skin defects on bilateral trunk. The mother was a 41-year-old primiparous woman who conceived a trigerinal dichorionic triamniotic FIVET pregnancy (embryo freezing at 35 years), complicated by selective embo-reduction of the third twin at 9 weeks of gestation due to malformations. No exposure to drugs or infection had occurred and there was no family history of Aplasia Cutis Congenita (ACC), vascular disorder, or consanguinity. According to obstetric history, and typical pattern of lesions, we diagnosed fetus papyraceus-associated ACC, trigeminal dichorionic triamniotic FIVET pregnancy (embryo freezing at 35 years), complications.

P 099

NEONATAL PERUNGERAL HYPERPIGMENTATION IN A CAUCASIAN CHILD

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Neonatal perungual hyperpigmentation is a benign skin abnormality that is classified among transient neonatal dermatoses. Hyperpigmentation is often seen in dark-skinned babies (skin phototypes 4 and darker) but rarely reported in fair-skinned babies. The lesions are usually located on the distal phalanges of hand and feet, external genitalia and areola. Although it is thought to be caused by maternal hormones, etiopathogenesis has not been fully elucidated. This entity is generally observed in the first few months of life and regresses within 2 years. The 6-month-old Caucasian girl was admitted to our clinic due to brown macules on her distal phalanx of fingers and toes and external genital area. Her parents noticed the pigmentation on the perungual and vulvar regions when the baby was 2 months old. Vulvar pigmentation progressively spread to the gluteal region until she was 4-months-old while perungual pigmentation did not change. The baby did not have a history of drug intake or trauma. Her mother had a history of colon cancer and use of imatinib therapy 5 years ago but no history of drug use during pregnancy. On dermatological examination all of the distal phalanx of the hand and feet showed a dark brown symmetrical transverse pigmentation. The cuticle and the nail unit was normal. Poorly defined hyperpigmented patches were observed in the external genital area including the vulva and anal region. Areolar pigmentation was normal. Dermoscopy was performed and hyperpigmentation was seen in the regular reticular pattern. Neonatal perungual hyperpigmentation and was diagnosed with clinical and dermoscopic findings. Diagnosis of neonatal perungual hyperpigmentation was made according to clinical and dermoscopic features. It is important to diagnose and to distinguish it from other pigmented diseases.

P 100

XANTHELASMOID VARIANT OF URTICARIA PIGMENTOSA - A RARE PRESENTATION OF CUTANEOUS MASTOCYTOSIS

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Introduction: To date, only a few genotype-phenotype correlations have been described in type 1 neurofibromatosis (NF1). A three-base-pair deletion in exon 17 of the NF1 gene (p.Met992del), resulting in the loss of a single amino acid, has been related to a mild form of NF1.

Case report: A 14-year-old adolescent presented with more than six cafe-au-lait macules (CLM) since first months of life, and skinfold freckling. He also suffered from learning disabilities, dilatation of the pulmonary artery with self-improving pulmonary valve stenosis, and scoliosis. Cutaneous, subcutaneous or visible plexiform neurofibromas were not evident, and no Lisch nodules were observed after ophthalmologic examination. No other skeletal or internal organ alterations were found.

Discussion: Just a small number of genetic abnormalities in NF1 have been related to a characteristic phenotype.

One of the mutations related to a particular phenotype is p.Met992del, which accounts for 0.8% of NF1 cases. It is associated with a relatively mild NF1 phenotype with CLM and skinfold freckling but without visible cutaneous or plexiform neurofibromas. Compared with typical NF1, a higher prevalence of pulmonary valve stenosis, and a lower prevalence of macrocephaly, short stature, pectus excavatum, or learning problems were observed in individuals with p.Met992del mutation. Other genetic alterations associated with specific phenotypes of NF1 include large NF1 deletions and missense mutations at p.Arg1809.

In conclusion, there is evidence of a genotype-phenotype correlation in NF1. This may have an impact on the management and surveillance of these patients, and guide genetic testing.
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RAS-ASSOCIATED AUTOIMMUNE LEUKOPROLIFERATIVE DISORDER AND JUVENILE MYELOMONOCYTIC LEUKEMIA: SIMILAR DISEASES WITH VERY DIFFERENT PROGNOSTIC FEATURES.

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Introduction: Ras-associated autoimmune leukoproliferative disorder (RALD) is a chronic, non malignant condition that presents with persistent monocytosis, lymphoproliferation, and autoimmunity. It has clinical and laboratory overlap with juvenile myelomonocytic leukemia (JMLL) including somatic mutations in KRAS or NRAS, but with a very different prognosis. This is the first reported case with cutaneous lesions.

Clinical case: A 8 year old boy consulted for the presence of multiple eritemato-violaceous plaques in the face and trunk present for two years without other systemic symptoms. At physical exam he only showed some lymph node enlargement. He had an uncle who died from myeloid leukemia and a 23 year old brother with chronic monocytosis without other symptoms. The skin biopsy showed an immature myeloid infiltrate with monocytic phenotype, and blood analysis revealed the presence of monocytosis. ANA were 1/320 and bone marrow aspirate showed monocytosis and 5% blasts. Karyotype was 46 XY and there were no mutations in PTEN1, KRAS, NRAS, CBL nor BCR/ABL. A mutation in KRAS was present. All these findings suggested the diagnosis of RALD.

Discussion: RALD and JMLL are two diseases with similar clinical and laboratory findings but with very different prognoses: RALD has an indolent clinical course whereas JMLL is fatal if left untreated. Distinguishing these diseases has implications for clinical care because prognosis and consensus diagnostic guidelines are critical in the setting of patients with normal karyotypes.

In spite of the benign nature of RALD, we recommend clinical monitoring for malignant transformation and/or acquisition of additional dysplastic, or clonal karyotypic abnormalities. Cutaneous lesions have not been previously described in RALD, but in our patient were the main symptom that lead to the diagnosis. Dermatologists have to be aware of RALD diagnosis in patients with immature myeloid infiltrates in cutaneous biopsy and monocytosis in peripheral blood.

P 103

LANGERHANS CELL HISTIOCYTOSIS AND TELANGIECTASIA MACULARIS ERYPTIVA PERSTANS IN A CHILD: A FORTUITOUS ASSOCIATION?

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A 5 month old girl was admitted to the intensive care unit of our hospital with a history of pancytopenia, bilateral suppurative otitis media, and skin lesions. After hemorrhagic compensation, we performed a skin biopsy which was consistent with Langerhans cell histiocytosis. The patient was treated with oral prednisone and vinblastinum.

Five months later she was admitted to the intensive care unit with a diagnosis of sepsis and necrotizing fasciitis of the abdominal wall. She was treated with endovenous antibiotics and surgery to remove dead tissue with good resolution.

Two years later, she suffered a relapse of the Langerhans cell histiocytosis with compromise of bone and skin. Again, she was treated with oral prednisone and vinblastinum.

One year later, in August of 2016, she presented eritemato-brown macular lesions with telangiectasia in arms and legs that did not disappear at discoscopy. We performed a skin biopsy and confirmed the diagnosis of telangiectasia macularis eryptiva perstans.

We present a child with two infrequent consecutive clonal disorders of bone-marrow derived cells: multisystemic Langerhans cells histiocytosis and telangiectasia macularis eryptiva perstans. Infiltration of mast cells and Langerhans cells in the same lesion has been published before, but, to our knowledge, this is the first time that the consecutive occurrence of telangiectasia macularis eryptiva perstans and multisystemic Langerhans cell histiocytosis is reported. We hypothesize that both proliferative processes have a common origin.

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LARGE CONGENITAL MELANOCTIC NAeva: A THERAPEUTIC CHALLENGE

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Introduction: Large congenital melanocytic naevi (LCMNs) are rare, but they are associated with a risk of melanoma which is poorly quantified and often represent a major cosmetic, psychological, and social problem. Management of such patients is challenging and there is no consensus on the most appropriate strategy for treating them.

Case report: We report two cases of LCMN. The first case is a newborn girl, who presented a giant congenital melanocytic naevus covering sixty-five percent of the total body surface. The involved areas included the scalp, right cheek, left shoulder, lower abdomen, back, buttocks, sacral, perineum, external genitalia, and upper limbs. There were also some satellite pigmented lesions in the upper and lower extremities and in the trunk. The second case is a newborn boy, who presented a large naevus spilus-type congenital melanoctytic naevus located on the left side of his face. Both cases underwent curettage of the lesions under general anaesthesia before ten days of age, without complications and with a good cosmetic result.

Discussion: Naevus cells in LCMN are found throughout the dermis and sometimes penetrate the subcutaneous septa. The partial removal of these superficial naevus cells by curettage is less traumatic than excision surgery and produces an acceptable cosmetic result. It should be performed during the first fifteen days after birth, when there appears to be no risk of leakage plane within the dermis. The risk of malignant transformation is greatly reduced by decreasing the total number of naevus cells. Patients and parents are pleased with the cosmetic and functional results and therefore suffer less from the psychosocial inconvenience caused by these lesions. Careful long-term follow-up is essential in order to monitor final cosmetic outcome and reduce the potential for malignancy. In our opinion early curettage is a good option in the management of LCMN.

P 105

PRESENTATION OF AN ATYPICAL SPITZ NEVUS ON A 9-YEAR OLD GIRL

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Atypical Spitz nevus is a borderline spitzoid melanocytic lesion of uncertain malignant potential. It is important to recognize atypical Spitz tumors because they may present with overlapping histopathological characteristics with both Spitz nevi and Spitzoid melanomas. Herein, a 9 year-old female patient who was diagnosed with atypical Spitz nevus will be presented with dermatologic, dermatomic, and histopathologic examination features. A 9-year-old female patient admitted to our pediatric dermatology outpatient clinic with a complaint of a growing lesion on her back. On dermatologic examination, a dome shaped palpable slightly erythematous papule was noted on the interscapular area. The patient’s mother reported that this lesion appeared just a month ago and showed significant increase in size within this period. There was no previous trauma or spontaneous bleeding. Considering its accelerated growth, the lesion was totally excised with prediagnoses including Spitz nevi, compound nevus, molluscum contagiosum, and juvenile xanthogranuloma. Histopathologic examination revealed a diagnosis of atypical Spitz nevus. Resection of the lesion with a margin of 0.5 cm was recommended.

Atypical Spitz nevus is a rare melanocytic proliferation that typically occurs in young patients. Although they are mostly accepted as indolent, cases with metastases leading to death have been reported. Thus, differential diagnosis of nodular lesions should be made carefully in the pediatric population.

P 106

POSTVACCINAL BULLOUS LESIONS IN DIFFUSE CUTANEOUS MASTOCYTOSIS

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Mastocytosis is characterized by clonal proliferation and increase in the number of mast cells in various organs. The typical organ affected in children is the skin. The World Health Organization classifies cutaneous mastocytosis into mastocytoma, maculopapular cutaneous mastocytosis, and diffuse cutaneous mastocytosis. We report the case of a 4-month-old male infant with multiple bullae on trunk and scalp that had appeared in the last 24 h. He was irritable but in good general condition, without systemic symptoms or signs of sepsis. Cutaneous examination showed tense blisters, vesicles, erosions and marked dermographism. His mother related another milder break, in both cases the previous background was vaccination a few days before. Initial therapy was antibiotics, antihistamines, and corticoids. Blood tests, cultures and abdomi-
nal ultrasounds were normal except elevated serum trypsinase levels (~200 mg/dL). Skin biopsy evidenced infiltration of mast cells in the dermis and confirmed our suspicion of mastocytosis. Currently, he is receiving treatment with oral antihistamines and mast cell stabilizers. His parents decided to postpone future vaccination. Blistering lesions open up a range of differential diagnoses including infection, autoimmune, hereditary and reactive diseases but we must not forget entities such as mastocytosis. The goal of mastocytosis treatment should be to control clinical manifestations caused to the peculiarity of this type of tumors, reporting clinical cases is highly important in order to study possible associations with other conditions.

**References:**

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**PERINEAL GROOVE: MAYBE NOT SO RARE?**

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**Background:** A perineal groove is a congenital malformation characterized by a groove-like erythematous mucosal lesion from the posterior vaginal fourchette to the anterior anal rim, without abnormalities of anus, urethra or vagina, and with hypertrophy of both edges. It occurs almost only in female patients.

**Objective:** The aim is to report our cases perineal groove and to give an overview of the literature.

**Methods:** We have described three patients who were referred to our tertiary dermatology outpatient clinic and performed a literature search on perineal groove.

**Results:** We present a girl of 13 months, a girl of 20 months, and a girl of 5 months who were referred to our tertiary center because of diagnostic uncertainty. In all three cases there was a significant doctors' delay before the diagnosis was recognized and this led to unnecessary therapeutic interventions. So far, in the literature only 30 cases have been described. However, we believe that unfamiliarity with the diagnosis, the often symptomless presentation, and the frequent spontaneous resolution around the age of 2 years are reasons for underreporting of this diagnosis.

**Conclusion:** Better recognition of perineal groove may lead to an early diagnosis and adequate management of the disease.

**References:**
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RARE CASE OF CHILDHOOD HYPOPIGMENTED MYCOSIS FUNGOIDES

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Introduction: Primary cutaneous lymphomas (PCL) are uncommon in pediatric patients. Mycosis fungoides (MF) is the most frequent subtype in childhood. There are various clinical forms including the hypopigmented MF (HMF). Malignant degeneration of HMF is frequent because of its clinical and histological resemblance to several other inflammatory skin diseases.

Case presentation: We present a 10-year-old boy with a three month history of progressive, generalized, pruritic skin lesions. Dermatological examination showed hypopigmented macules and a few erythematous plaques on the back. The patient had no improvement with topical emollients. Due to the persistence and progression of the lesions- leukemoid cells, clinical data, bone marrow, and peripheral blood findings, the diagnosis of HMF was confirmed. Treatment, according to the appropriate protocol, was introduced.

Objective: To report the case of a 112 month old boy with leukaemia cutis as a first manifestation of ALL.

Case report: We present a male patient who was referred to our pediatric dermatology department with skin lesions on his right buttok present for 2 months. The child was otherwise healthy and had normal laboratory test results. The biopsy of skin changes revealed a massive infiltration with atypical lymphocytes. The patient was referred to the Pediatric Hematology Department. Based on the cytological features with the immunophenotype of the tumour cells, clinical data, bone marrow, and peripheral blood findings, the diagnosis of ALL was confirmed. Treatment, according to the appropriate protocol, was introduced.

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SYSTEMIC MASTOCYTOSIS, A DIAGNOSTIC CHALLENGE

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Introduction: Systemic Mastocytosis (SM) is an uncommon disease characterized by an excessive accumulation of mastocytes which can infiltrate several organs and tissues (skin, bone marrow, spleen, lymph nodes, liver and gastrointestinal tract). The diagnosis is clinical and based on histopathological findings from biopsies of the organs affected. In children, the disease is usually considered to be benign. Most patients present symptoms related to mastocyte mediator release, and prevention of its effects on tissues is crucial to the treatment. A case of systemic mastocytosis with unexpected presentation is reported, which illustrates the complex differential diagnosis of this disease.

Clinical case: A child of 11 years was referred for neurodevelopmental retardation and blemishes and light brown papules located on the trunk since 3 years ago. The cutaneous symptoms were compatible with urticaria pigmentosa and mastocytosis was confirmed by deep phenotyping. Initially, non-aggressive mastocytosis treatment was started and he was referred to the Mastocytosis Unit.

Conclusion: This is a rare disease that required perseverance in the diagnostic search due to a wide range of differential diagnoses. It is crucial to utilize a multidisciplinary approach. The clinical and laboratory findings can be suggestive, but bone marrow biopsy and myelogram are essential for definitive diagnosis. It may be benign, with minor transient symptoms and signs that never cause the patient to consult a physician, or it may be life-threatening. Currently, there are no curative therapies for SM and treatment is intended to reduce symptoms associated with mastocyte mediator release. In aggressive forms, cytoreductive treatment is used to decrease the mastocytotic load. It is also essential to provide information to the family about agents that can trigger mastocytes activation in order to encourage prevention.
The patient was seen in the Oncology clinic at 6 weeks. The lesion was already regressing, and measured 6 × 5 mm. Blood tests, including FBC, U&E, LFT, and clotting, were normal. Skeletal survey and abdominal ultrasound (to assess for multi-site involvement) were also normal. The lesion has completely disappeared by the age of 4 months. Congenital, self-limited LCH is a rare but well described cutaneous form of LCH, associated with a good prognosis (1). Single lesion, single system, congenital LCH is even rarer. It typically presents as a single, red to brown papule or nodule that is often ulcerated or crusted. It can be present anywhere on the body, but it appears most commonly on the extremities (2). Lesions spontaneously regress over several months and there is no data suggesting surgical removal has any benefit (3). In the literature, no patient with congenital, single lesion, single-system LCH has been reported to have developed disease recurrence or progression (3). However, it is prudent to monitor these patients long-term because of the potential of LCH to progress or recur.


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ANETODERMIC PILOMATRICOMA: REPORT OF FOUR CASES IN CHILDREN

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Pilomatricoma is a cutaneous tumor derived from hair matrix cells with the first series of patients described by Malherbe et al. in 1880. Pilomatricomas have a wide variety of clinical characteristics, the anetodermatic variant representing a rare form (2%) (1) that has also been described as bullous, pseudo-bullous or lymphangiecatic. We describe four cases of anetodermatic pilomatricoma (AP) in children. All of them presented as a solitarily, slowly growing, tender nodule. Physical examination revealed a lesion composed of a firm subcutaneous nodule underlying an atrophic, folded, pink skin. Lesions were located on back, leg, and arm. Histologically, all the cases were well-circumscribed nodules located in the deep dermis and extending into the subcutaneous fat. Nodules were composed of irregular islets of shadow cells (eosinophilic, anucleated cells) focally surrounded by basoid epithelial cells. The stroma of the lesions showed variable inflammatory infiltrate with foreign body giant cells. Overlying dermis was edematous with fragmentation of collagen fibers and with some dilated lymphatic vessels highlighted by D2-40. The Ornstein stain revealed the absence of elastic fibers in the dermis overlying the nodule. The epidermis was normal or showed minimal irregular hyperplasia. Surgical excision was the treatment for APs. The pathogenesis of APs is not fully understood but several theories have been proposed. One of them is based on a traumatic origin. Mechanical trauma may play an important role by the disruption of elastic fibers and lymphatic drainage (2). A second theory is based on release of catabolic enzymes by tumor cells or associated inflammatory infiltrate (3). Both theories could apply simultaneously to the lesions but further investigations are needed in order to define the exact mechanism.


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UNUSUAL CAUSE OF INFLAMED ULCERATED PLAQUE ON THE BACK OF A YOUNG BOY: A CASE REPORT

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Introduction: Epidermoid cysts are asymptomatic, dome-shaped lesions that can arise from a ruptured pilosebaceous follicle and can occur in a variety of locations on the body. If there is no previous history of the typical nodule and inflammation, diagnosis can be delayed.

Presentation of case: We present a case of a 2.5-year old boy with a one month history of luid plaque on the lower back. The parents did not notice any changes on the skin of the back before the plaque appeared. No clinical improvement occurred after local antibiotic and corticosteroid treatment. Soon after, an infected, painless superficial ulceration appeared.

Materials and methods: A biopsy was performed. We excluded atypical mycobacteria infection, tuberculosis, and sarcoidosis and a histologic suspicion of cutaneous leishmaniasis was set. After several weeks the plaque disappeared on its own, leaving an atrophic scar. After a year a new inflamed papula appeared on the edge of the scar and we surgi-
cally removed it. The final pathologic diagnosis of foreign body giant cell reaction of a ruptured epidermoid cyst was set.

Conclusion: Epidermoid cysts are benign skin lesions encountered throughout the body. In the case of a papule, plaque or even inflamed plaque with ulcer, appearing on any part of the body epidermoid cyst should be considered in the differential diagnosis. In our case of a papule, plaque or even inflamed plaque with ulcer, appearing on any part of the body epidermoid cyst, we performed definitive histopathological diagnosis and for the prevention of possible future complications.

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EFFECTIVENESS OF RITUXIMAB IN JUVENILE PEMPHIGUS VULGARIS
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Background: Juvenile pemphigus vulgaris (JPV) is a rare, severe, chronic auto-immune disease characterized by blister and erosive lesions of skin and mucous membranes due to the breakdown of desmoglein 1 and 3 keratinocyte adhesion proteins. Oral corticosteroids are still the treatment of choice, but the response is not always complete and side effects often lead to high secondary morbidity. Rituximab has been shown to be effective for treating PV, but with few reports in the childhood population.

Case report: A 12-year-old boy presented to our clinic with a six-month history of persistent ulcerations and recurrent bullae of his oral mucosa. Clinical examination showed large erosions of the palate, gingiva, scrotum, perineum, and abdomen. Histopathology of an oral lesion revealed supra-basal bullae with a single row of keratinocytes attached to the basement membrane. Direct immunofluorescence showed intercellular deposits of IgG, IgM, and of C3 at the dermalepidermal junction. These findings were consistent with JPV. Titer of circulating anti-desmoglein 1 autoantibodies were normal and anti-desmoglein 3 autoantibodies were at 178.20 (N: >35). The regimens were well accepted by parents and judged to improve both the clinical manifestations and the subjects' quality of life.

Conclusion: In the last few years rituximab has been reported as a very promising therapy in the management of refractory PV in adults. 14 children (from 4 to 16 years of age) with JPV, have successfully been treated with this anti-CD 20 monoclonal antibody. There is still the need for a well defined treatment protocol in order to find the best disease control in children.

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EFFECTICITY, TOLERABILITY AND ACCEPTABILITY OF TOPICAL REGIMENS CONTAINING THE PREBIOTIC BIOLIN IN CHILDREN SUFFERING FROM ATOPIC DERMATITIS
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Background: Probiotics have the potential to correct skin dysbiosis and contribute to the improvement of atopic dermatitis (AD) when incorporated in topical moisturizers and emollients.

Objective: Assess the efficacy, tolerability, acceptability, and effects on quality of life of three Biolin containing topical products (face cream, body wash cream, and moisturizing body cream) in children with AD.

Methods: This was a single center, open label study, conducted for 8 weeks in parallel subgroups of children (n=60) aged 0 to 3 years with mild to moderate AD (SCORAD-35). Products were used for 56 consecutive days, twice daily for the face and moisturizing creams, and once daily for the body wash cream. At the 4th visit the SCORAD indexes, xerosis, erythema, and edema were assessed. Parent evaluation assessments were conducted at 14, 28 and 56 days.

Results: SCORAD indexes significantly (p < 0.05) decreased for all products at t28 and t56; average decrease ranged from 57.7% to 58.3%. Xerosis reductions were statistically significant (p < 0.05) for all products at t56; 96.6% to 100% of children experienced improvements in xerosis. Averages of parameters assessed through parents’ evaluation (pruritus improvement, dryness improvement, skin softness, quality of life, and product characteristics) were 8 out of 10 or higher.

Conclusion: The Biolin containing products tested in this study were beneficial and well tolerated in children below 3 years of age who suffer from mild to moderate AD (SCORAD-35). The regimen was well accepted by parents and judged to improve both the clinical manifestations and the subjects’ quality of life.

Key words: atopic dermatitis, children, moisturizer, dysbiosis, probiotics.
ders is an essential component of effective patient care. The APNs have clear structures, and well-defined responsibilities in their collaboration. Health care providers respect each other and recognize the value of their mutual knowledge and skills. APNs play a central role in the management of AD and provide health-care services that positively affect clinical outcomes.

P 126
COMBINED THERAPY IN 2 CASES OF PUSTULAR GENERALIZED PSoriasIS WITH IL36RN GENE MUTATIONS
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P 127
PATIENT COMPLIANCE IN ACNE VULGARIS
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3Background: Acne vulgaris, one of the most common dermatologic diseases, affects the vast majority of adolescents and young adults. The success of the treatment depends considerably on patient compliance including the degree to which the patient follows medical advice, use of prescriptions, correct application of the medications, and presence at the control visits. The main objectives of our study were to investigate the therapeutic compliance of patients with acne vulgaris, to explore factors leading to reduced compliance, and to assess the medicine-taking habits of the patients.

P 128
ORAL GLYCOPRYROLATE FOR TREATING PRIMARY HYPERHIDROSIS IN CHILDREN AND TEENAGERS AFTER FAILURE OF ORAL OXYBUTYNIN
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Objective: Oral anticholinergic drugs (OADs) are effective and safe drugs for the management of hyperhidrosis. Nevertheless, in some cases they fail either because they are ineffective or not tolerated. To date there are no studies focused on the use of a different OAD after failure (inefficacy/intolerance) of another OAD, and only two studies on the use of oral glycopyrraline (OG) for the treatment of pediatric hyperhidrosis. Our objective was to evaluate the effectiveness and safety of the use of OG for treating hyperhidrosis after failure of oral oxybutynin in children and teenagers.

Material and methods: An observational retrospective study was performed. We reviewed the medical history of every primary hyperhidrosis patient up to 18 years of age who received treatment in our Department with OG and who had been previously treated with oral oxybutynin from January 2012–October 2016. We described the cohort of patients, and the response and tolerance to the treatment was evaluated. Hyperhidrosis Disease Severity Scale was used to evaluate the response at 3 and 12 months-time. Glycopyrraline was prescribed with progressively increasing dosage depending on response and tolerance, starting with 2 mg/day.

Results: Five patients (4 women) were included. The median age was 17 years (11–18). The usual daily dosage was 2–6 mg (mean of 3.8 mg/day). After 3 months of treatment a good response (decreased sweating) was obtained in all patients. After 12 months, 3 patients maintained excellent response. Side-effects were reported by 3 patients at the 3 month appointment (polychuria was also reported by one). One patient discontinued treatment after 9 months due to inefficacy and another patient after 13 months due to inefficacy and side-effects.

Conclusion: Failure in the management of hyperhidrosis with oxybutynin in children/teenagers does not necessarily mean using other OADs will fail, glycopyrraline being a useful option in these cases.

P 129
A CONVENTIONAL THERAPY RESISTANT SEVERE PEDIATRIC PSORIASIS CASE SUCCESSFULLY TREATED WITH BIOLOGIC AGENTS
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Psoriasis can occur in up to 1% of the pediatric population and can be challenging to treat and control. We present a case of infantile onset pediatric psoriasis that was unresponsive to systemic conventional anti-psoriatic therapies but responded successfully to biologic treatments. An 11 year-old-female patient was admitted to the pediatric dermatology outpatient clinic for severe psoriatic plaques on trunk, scalp, and palmoplantar area. Medical history revealed a history of psoriasis since she was 2 months-old. Low dose systemic acitretin was commenced and used for 3 years with little response. Even though she was 11 years old she had not started going to school because of severe fissuration and hyperkeratotic plaques on the palms which made it impossible for her to hold a pen. She also had difficulty in standing up and walking because of prostatic plantar involvement. Her family history included an uncle with severe plaque psoriasis. She was started on systemic cyclosporine treatment for 6 months and the response was fair. After receiving permission from local medical authorities infliximab was started. After her third infusion significant reduction in thickness of the plaques was achieved. Within two months of infliximab therapy she was able to go to school. She used infliximab for 3 years and switched to adalimumab 40 mg/week after secondary unresponsiveness occurred. PASI75 was also achieved with this agent. The patient is 17 years old and still on this treatment without any serious side effects. Treatment of rare cases of pediatric psoriasis can be challenging but biologic agents are strong treatment options and may be used in patients who are resistant to conventional treatments.

P 130
SUN PROTECTION BELIEFS, HABITS, AND ATTITUDES AMONG ADOLESCENTS
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Background: Sun exposure in adolescence is an important risk factor for the development of adult skin cancer. Adolescents have poor sun protection habits and a narrow-con
ce of skin cancer risk. In addition, there is a wide desire to be tan in order to conform to fashion trends.

Aims: The aim of this study was to evaluate the beliefs, habits, and attitudes concerning the use of sun protection measures among adolescents, to identify the factors that influence them, and to analyse the impact of an educational intervention.

Methods: We designed a quasi-experimental study. The study group included secondary education students, aged 13–14 years old, from two institutes in Barcelona. It was conducted during 2016. Data was collected through questionnaires about beliefs, attitudes, and habits toward sun protection before and after educational intervention.

Results: We evaluated 135 patients (72 male, 63 female) with a mean age of 13.6 years. 72% thought that being tan made them look better, with statistically significant differences by gender. Sunscreen was only used when going to the beach or pool by 55%. A statistically significant relationship was observed between the use of sunscreen during childhood and its use during adolescence ($p < 0.001$). 58% of youth reported having had one or more sunburns during the past summer. 43% were not or almost not concerned about skin cancer, although 70% knew its relationship to sun exposure. After educational intervention, there was an increase in the use of sunscreen before leaving home ($p < 0.001$) and a greater use of sunscreen by female students on cloudy days ($p = 0.018$).

Conclusion: Adolescents have insufficient sun protection habits. We believe that we should focus on modifying the positive attitudes they have toward tanning. There is a need to strengthen the implementation of sun protection campaigns in order to minimize the risks associated with excessive ultraviolet exposure and to improve adolescent sun protection behaviour. The role of parents, teachers, and health care providers is essential.

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TOLERANCE AND EFFICACY STUDY OF A NEW CLEANSER CREAM ON ATOPY-PRONE SKIN
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Introduction: Cleansing and moisturizing are essential in the daily routine of pediatrics with dry to atopy-prone skin. This population needs a cleanser that moisturizes and soothes skin.

Objective: We aimed to evaluate the efficacy and tolerance of a new cleanser cream applied daily over a period of 3 weeks on 50 infants, children, and adults with SCORAD between 10 and 25.

Methods: Evaluations immediately after the first application and after 3 weeks included a 3 h skin hydration kinetic on day 1, evolution of SCORAD, desquamation and soothing effect using a visual analogic scale (VAS). A self-assessment questionnaire was filled in by the subjects/parents. Cutaneous and ocular tolerance were assessed by a dermatologist, a pediatrician, and an ophthalmologist.

During the study period patients did not apply any emollient to avoid biasing evaluations of the tested product.

Results: There was a significant increase of $19\%$ ($p < 0.001$) in skin hydration after 3 weeks. SCORAD improvement occurred in 86% of the subjects with a significant decrease of 60% ($p < 0.001$) at D25. 86% of subjects confirmed a skin soothing effect. Ocular tolerance was considered excellent (no adverse symptoms) and cutaneous was considered good (7 subjects experienced transient, mild functional and physical signs that did not lead to product discontinuation).

Conclusion: This new cleanser promotes skin hydration, soothes the skin, and is well tolerated. The few reported adverse side effects could be explained by the absence of emollient during the study in this atopy-prone skin population, considering that emollient application is the mainstay in Atopic Dermatitis treatment between flares. This product is a good choice of cleanser for those with sensitive skin.

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A PERSONALIZED INTEGRATIVE MULTIDISCIPLINARY TREATMENT PROGRAM (PIM) FOR ATOPIC CHILDREN AND ADOLESCENTS WITH DIFFICULT TO TREAT ATOPIC DERMATITIS

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Atopic dermatitis (AD) is a chronic, pruritic, inflammatory skin disease with a major impact on the life of patients and their families. We developed a personalized integrative multidisciplinary treatment program (PIM) for children and adolescents with insufficient disease control despite conventional treatment strategies according to current guidelines. PIM actively involves the child and his parents and combines patient designed treatment goals with a systematic integrative multidisciplinary approach by the involved health profession-als. This includes assessment and treatment of AD, other atopic, pediatric and mental health comorbidities, and general well-being. Throughout the integrative treatment program, multiple health professionals work on the same treatment goals simultaneously, each using treatment strategies from their own field of expertise. Disease activity was assessed with the clinical scoring system SÆAESI and health related quality of life was assessed with the COLODI. In total, 79 children with difficult to treat AD participated in PIM and long term treatment results were available for 74 children. Treatment was considered successful when a relative improvement of 75% on the SÆAESI or a small effect (a score $< 1$) on the child’s life was observed. Six months after the end of PIM, treatment has been successful for 57% (77%) children. Of those, 30 children demonstrated both improved disease activity and quality of life, 6 children demonstrated improved disease activity, and 21 children demonstrated little impact on quality of life. PIM is a time and resource intensive program and good communication skills are essential for involved health professionals. We encourage all clinicians who are involved with children with difficult to treat AD to consider a treatment approach like PIM.

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PROSPECTIVE STUDY OF VASCULAR ANOMALIES. EVALUATION OF PROPRANOLOL IN HEMANGIOMAS
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Introduction: Vascular anomalies (VA) are a heterogenic group of processes classified into malformations (M) and vascular tumors (VT). Among the latter, there are hemangiommas (H) (inflammatory and congenital) which, although benign, can compromise vital functions and cause cosmetic defects.

Material and Methods: We performed a descriptive-prospective study of 310 patients with VA referred to Dermatology, from September 1, 2009 to March 29, 2013. We evaluated their diagnosis and analyzed the types of VA, their epidemiological characteristics, clinical characteristics, complementary studies, and treatments. Patients or parents/legal guardians signed the informed consent for registration of the data in the computer database and/or treatment. We also describe our experience with propranolol in the treatment of hemangiommas, its protocol and use. We were endorsed by the Pharmacy Service and an Interdisciplinary Committee.

Results: The most frequent diagnosis was VT (58.06%), followed by malformation (41.9%) and anomalies (7.1%). Only 0.6% were in the other TV group (tufted angioma). 98.88% of VTs were hemangiommas, inflammatory H (87.20%), and congenital H (6.8%). 85.5% of vascular M were capillaries. 7.4% had associated systemic involvement. Only 0.6% were in the other M group (tufted angioma).

Discussion: We contribute a series of 56 patients treated with propranolol, including inflammatory hemangiomma (IH) (even 4 with PHACES Syndrome) and congenital H. Propranolol is statistically significant that children under six months are more likely to have VAs in the anterior thoracic region and in the abdomen. Those of six months or more are more likely to have vascular lesions in the lip area. We consider propranolol in oral solution the best treatment for complicated H. Considering its satisfactory results, high tolerance, and minimal side effects.

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INFILTRATION OF BOTULINUM TOXIN IN RAYNAUD'S PHENOMENON
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Introduction and aims: Raynaud’s Phenomenon (RP) has rarely occurs in pediatrics. Its diagnosis is clinical and the initial treatment consists of preventive measures (avoiding the use of vasconstrictor medicines or avoiding cold temperatures). The therapeutic options which are most frequently used are topical nonsteroids and calcium antagonists, taken orally. Recently the infiltration of Botulinum Toxin (BT) has been tested with promising results.

The aim is to check the effectiveness of BT infiltration in a series of cases in a Pediatric Rheumatology Department of a tertiary level hospital.

Methodology: A descriptive study was undertaken using patients affected by RP who displayed severe impaired quality of life. There were 4 female patients aged between 7 and 16. A total of 6 procedures were performed between 2015 and 2017, during the winter months.

The procedure consists of the infiltration of BT (10–15 IU) in the interdigital spaces from the 2nd to the 5th finger of both hands at the level of the metacarpophalangeal joint. It is
performed on an outpatient basis, lasting approximately 30 min, and with use of sedation in the younger patients. The clinical response was evaluated through surveys conducted before and after the interventions.

**Results:** A decrease in the feeling of cold was perceived by 50% of the cases. In the only patient who suffered painful episodes, a complete remission of pain was achieved in the next two years. Changes in coloring, frequency, and duration have remained stable.

**Conclusion:** When used together with general preventive measures BT is a valid therapeutic option for RP. In our experience this is a well-tolerated technique. Even so, studies involving a wider sample of the population are necessary in order to obtain representative results.

**P 136**

**A CASE OF CHILDHOOD GENERALIZED PUSTULAR PSORIASIS SUCCESSFULLY TREATED WITH A COMBINATION OF ACITRETIN AND CYCLOSPORINE**

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Generalized pustular psoriasis (GPP) is an extremely rare type of psoriasis that is also known as Von Zumbusch. It is named after the German dermatologist Leo Ritter von Zumbusch (1874–1940) who first described this condition. GPP affects both men and women equally in all races. It can be life-threatening especially in elderly patients. The average age of patients is 50 years. GPP may occur at any age, however it is less common in young children. Treating children with this condition is problematic. Boys tend to be affected slightly more than girls. It can appear spontaneously, even without previous psoriatic conditions, and can recur in periodic flares-up.

We present a case of life-threatening GPP affecting a 7 year-old boy. It occurred out of the blue without a previous history of a different type of psoriasis. The patient was hospitalized and hypocalcemia was not observed (1). The patient was given a combination treatment of acitretin (0.5–1 mg/kg/day) and cyclosporine (3–5 mg/kg/day). The patient tolerated this combination treatment well and recovery was observed after only two weeks.

The treatment of GPP is always problematic and the disease can be life-threatening in the elderly and in children. Biologic agents including etanercept, adalimumab, and infliximab licensed for moderate to severe plaque type psoriasis in combination with conventional agents are necessary for prompt treatment of GPP in children. We recommend a combination treatment of both acitretin and cyclosporine for childhood GPP.


**P 137**

**METHOTREXATE FOR SEVERE NUMMULAR ECZEMA IN CHILDREN: A RETROSPECTIVE STUDY OF 30 PATIENTS**

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Nummular eczema in children is a chronic condition characterized by very pruritic coin-shaped eczematous lesions that affect any part of the body and often become exudative. This condition may be present in the setting of atopic dermatitis but often occurs independently. Mild-to-high potency topical corticosteroids are considered the mainstay treatment, however, there is limited data on the use of systemic therapy in nummular eczema in children.

**Objective:** To evaluate the efficacy and safety of Methotrexate (MTX) in children with severe nummular eczema.

**Patients and methods:** A retrospective review was undertaken of children with nummular eczema treated with MTX between January 2007 and May 2017.

**Results:** Thirty patients (26 male, 4 female) with an average age of 8 years (range 2–17 years) were reviewed. Thirteen patients (43%) were completely clear or almost clear of eczema after an average of 13 months of therapy (range 3–30 months) and 53% of patients were still on treatment at the time of last review. Failure of treatment was observed in one patient. The most common adverse effect was non-significant elevation of liver enzymes in 6 patients (20%) and minor nausea in 5 patients (16%). No serious adverse events were noted.

**Limitations:** This is a retrospective study. The follow-up period is limited.

**Conclusion:** In our experience MTX has a good safety tolerability profile and should be considered in moderate to severe pediatric nummular eczema that has failed to respond to conventional topical therapy. Because of our data, its weekly dose schedule, and its convenience as a long-term treatment we conclude that MTX is an effective, well-tolerated, and safe treatment, in children with moderate to severe nummular eczema.

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**TOPICAL IMIQUIMOD REVERSE ACTINIC DAMAGE IN TWO PEDIATRIC PATIENTS WITH KERODERMA PIGMENTOSUM**

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**Introduction:** Kerodermia pigmentosum (XP) is an autosomal recessive disease with an extremely high incidence of ultraviolet-related skin cancers. The diagnosis is usually delayed even though significant actinic damage is already present. Preventive measures and early therapy of malignancy are key for patient management.

**Case report:** Two patients with XP-C, both with identical homozygous mutations in the XPC gene (C1243C–T p.Arg415X) presented with hyperpigmentation and freckling on sun-exposed areas. In one of them, a 5-year-old boy, a small BCC on his face was treated with 5% imiquimod cream. He experienced a remarkable clearance of the surrounding background pigmentation and freckling. Sequential application of imiquimod on other areas of the face led to an almost complete clearance of hyperpigmentation and freckling. A second patient, a 2-year-old boy, was treated with 5% imiquimod cream for a BCC on his nose. Again, sequential therapy with 5% imiquimod on his face led to a marked improvement in the background pigmentation and freckling. In both cases, extreme sun avoidance and protection were instituted. No new BCCs have appeared.

**Discussion:** Blanching and chemophylaxis with topical imiquimod to treat the cancerization field induced dramatic cosmetic improvement in our two patients with XPC. Topical imiquimod may possibly revert actinic damage and prevent carcinogenesis in children with XP. It is not known if this effect is linked to certain mutations or certain types of XP only. Imiquimod-induced vitiligo-like depigmentation may have played a role in these two patients.

**P 139**

**A CHALLENGING THERAPEUTIC CASE OF JUVENILE DERMATOMYOSITIS**

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Juvenile dermatomyositis (JDM) is a rare chronic inflammatory disease of childhood affecting approximately 3.2 children per million per year. The characteristic clinical features of proximal muscle weakness and rash are secondary to a systemic immune-mediated vasculopathy. The outlook for children with JDM has greatly improved compared to 50 years ago, when one-third of affected children died and another third had disabling complications of the disease. The prognosis of JDM is now more favorable. Mortality is rare and less than 10% of children have significant disability in long-term follow-up.
We report a case of a Peruvian 12-year-old boy who presented moderate-severe muscle weakness, poliomyelitis on the upper chest, upper back, facial edema, Gottron’s papules, diffuse non-scarring alopecia, and hypertrophic nail cuticles since December 2014. Before coming to Spain the patient had been managed in Peru where he had received treatment with Methotrexate, Prednisone, Azathioprine and Mycophenolate Mofetil with little success. After performing specific tests to confirm the diagnosis (blood serologic tests for muscular enzymes, electromyography, autonomic and muscular biopsies) we began treatment with Prednisone (1 mg/kg/daily), Mycophenolate Mofetil (1.500 mg/daily), intravenous immunoglobulins (IVIG) (2 g/kg every 2 weeks ≤ 3, then monthly) and Rituximab (1.000 mg/monthly). After 4 weeks, muscular weakness and rash had improved and we began the prednisone tapering following the advice of the Childhood Arthritis and Rheumatology Research Alliance (CARRA). We present a case of JDM treated with a CARRA treatment plan (intercosteroids, IVIG, and a corticosteroid-sparing agent) plus Rituximab. The latter was used because local analyses of all cases in the literature supports the off-label use of Rituximab in some patients with refractory myositis.

P 140
DIFFERENT TREATMENT OPTIONS FOR PEDIATRIC PSORIASIS
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Psoriasis is a chronic, multifactorial inflammatory skin disease affecting 0.5% to 2% of the pediatric population. Approximately one-third of patients with psoriasis develop clinical features in the first two decades of life. Onset is usually after puberty, with no clear gender predilection. Due to the chronic nature of psoriasis, the child and its family must learn to cope with the demanding treatment. A better understanding of the pathogenesis of psoriasis is crucial for effective treatment. Psoriasis is not solely a hyperproliferative disease of keratinocytes, but a chronic, inflammatory, multisystem disease associated with a number of co-morbidities. Combination and rotational therapy are helpful in reducing toxicity and maximizing efficacy. Patients with mild and limited disease severity respond well to topical treatment with steroids or vitamin D analogues, unlike moderate and severe psoriasis where sufficient remission is rarely achieved. Therefore phototherapy, systemic immunomodulators, or biologic agents are the next line of treatment to be considered. Biologic agents target specific parts of the immune system important for the development of psoriatic lesions. They represent a new therapeutic option for treating moderate-to-severe plaque psoriasis unresponsive to other systemic treatment or phototherapy, or if those treatments are contraindicated. There are limited data available on the use and long-term safety of biologics in the pediatric population. Treatment of childhood psoriasis is a challenge to dermatologists due to the lack of clinical trials and published data as well as insufficient guidelines to facilitate decision-making. Many medications are not registered for use in the pediatric population or are used off-label. Considering the great impact of psoriasis on the quality of life of our young patients and their families, treatment is often a challenge and must be individualized, taking into consideration the efficacy and safety of the medication.

P 141
ASSESSMENT OF BAFF LEVEL IN CHILDREN WITH ATOPIC DERMATITIS BEFORE AND AFTER BROAD BAND UV A THERAPY
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Background: UV A1 irradiation is effective for atopic dermatitis patients with acute exacerbation. The optimal dose for therapeutic efficacy has been evaluated in many studies. However, relapses are common. Not enough studies have tackled the role of BB UV A in the treatment of AD, particularly in children. Aim: To evaluate the therapeutic role of Broad Band UV A in treatment of Atopic dermatitis patients both on the clinical and laboratory levels. Methods: Fifteen pediatric patients ranging from 5 to 12 years old with childhood atopic dermatitis were included. They received 3 sessions/week at a fixed dose of 15J/cm². Their SCORing Atopic Dermatitis (SCORAD), serum BAFF, and IgE levels were assessed before and after treatment. Results: There was a statistically significant decrease in SCORAD, BAFF, and IgE serum levels after UV A therapy (p < 0.05). Both SCORAD and IgE levels were significantly correlated to each other (p < 0.05), but not with the BAFF level (p > 0.05). Conclusion: UV A is an effective, inexpensive, convenient, and relatively safe modality in the treatment of AD in children.

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TOPICAL PROPRANOLOL GEL FOR PYOGENIC GRANULOMA
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Background: Pyogenic granulomas (PG) are commonly acquired vascular tumors accounting for 0.5% of all skin nodules in children. Although they are benign vascular proliferations, treatment is often sought because of recurrent episodes of bleeding and for cosmetic considerations. Numerous treatment options are available but recurrence rates are high. Noninvasive methods of treatment are being sought, particularly for young children. Lately there have been some publications regarding propranolol being an effective treatment modality for PG.
Objective: We sought to evaluate the clinical effectiveness and safety profile of topical propranolol Gel for the treatment of PG.
Materials and methods: A retrospective review was performed on 18 children who each had one lesion (10 male and 8 female) clinically diagnosed as PG, with an average age of 4.7 years (range 5 months -12.2 years). A total of 18 lesions were treated with propranolol 4% gel, applied twice daily for an average duration of 10.6 weeks (range 1–40 weeks).
Results: In the whole tested group, and also in men and women separately. Significantly better results were seen in men. In 12 lesions the lesions almost completely resolved. Three patients (16.6%) underwent a curettage after only a few weeks of treatment due to bleeding of the PG, one patient (0.05%) discontinued the treatment due to local irritation, and another patient (0.05%) was advised to remove the PG surgically. No other topical or systemic complication was observed in any of the patients.
Conclusion: Topical therapy with propranolol 4% gel may be a safe and effective method for the treatment of PG.

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QUALITY OF LIFE OF PATIENTS WITH SEVERE ACNE AND ITS IMPROVEMENT AFTER PERORAL ISOTRETINOIN THERAPY
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Background: Acne is a chronic disease that frequently interferes with the quality of life. Peroral isotretinoin is the only therapeutic agent that exhibits effective activity against all four main aetiological factors. Successful treatment of acne may improve a patient’s quality of life. To investigate this issue, we conducted a no controlled prospective study. Methods: One-hundred and fifty patients were included in our single center, no-blind, and no controlled prospective study. Before and after finishing of orally administered isotretinoin, acne severity was assessed by grading and physical dermatological examination. All patients completed DLQI and CADI scores before treatment and after finishing isotretinoin therapy. Statistical analyses of DLQI and CADI scores were performed. Results: All patients completed the study. 76% were men, 24% were women. 22% suffered from severe acne papulopustulosa, 64% from acne nodulocystica, and 24% from acne conglobata. All patients were healed after isotretinoin treatment. Before the treatment the mean DLQI score was 8.5 (8.2 for men, 9.9 for women), the mean CADI score was 6.2 (6.2 for men, 6.3 for women). After finishing isotretinoin treatment the mean DLQI score was 1.7 (1.7 for men, 1.7 for women), the mean CADI score was 1.6 (1.6 for men, 1.4 for women). A significant improvement in both DLQI and CADI scores was observed after isotretinoin therapy in the whole tested group, and also in men and women separately. Significantly better results were seen in men. Conclusion: Our prospective study of patients treated with peroral isotretinoin for severe acne indicates that there is a deterioration of quality of life for most patients before treatment. A significant improvement of quality of life was observed after successful isotretinoin therapy in the whole tested group, and also in men and women separately.

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TELEMEDICINE IN CHILDREN WITH DERMATOLOGICAL DISEASES - A PROSPECTIVE RANDOMIZED STUDY IN THE DEPARTMENT OF DERMATOLOGY UNIVERSITY MEDICAL CENTER MAINZ/GERMANY
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Background: Telemedicine is one of the newest communication technologies being used in different fields of medicine. It is defined as an interaction between one or more health care professionals or between patients and health care professionals.
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ANTIHISTAMINE-RESISTANT CHRONIC SPONTANEOUS URTICARIA IN CHILDREN – TREATMENT WITH OMALIZUMAB

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**Background:** Chronic spontaneous urticaria (CSU) is a common and sometimes severe disease in adults and children. Successful management includes searching for possible causes/triggers and therapy to resolve the disease. The first-line treatment is second-generation antihistamines. If this is not effective the dosage can be increased up to four times, in children weight-adapted. Second-line therapy includes treatment with omalizumab in chronic spontaneous urticaria (approved from the age of 12 years).

**Objective:** The aim of this study is to prove whether the use of omalizumab is useful in pediatric chronic spontaneous urticaria patients who are antihistamine-resistant.

**Methods:** 10 children (between 7 to 17 years old) with CSU were treated with omalizumab. Patients reported outcomes as urticaria activity score over 7 days (UAS7). Urticaria control tests (UCT) were run before and after treatment. Adverse events were reported.

**Results:** 8 children suffered from CSU, 2 from combinations of inducible urticaria and CSU. 2 patients had only hives and pruritus, 8 of 10 suffered from urticaria, pruritus and angioedema. The average age was 14 years, the average duration of urticaria was 2 years. All patients were antihistamine-resistant to different antihistamines (including updosing therapeutic strategies) before starting omalizumab therapy. UAS7 scores decreased from an average of 27 before therapy to an average of 13 after 16 weeks. 4 patients had UAS 7 of 0 after 16 weeks. UCT increased from 4.8 to 11.9 after 16 weeks. Anhystaminines comedication could be reduced or stopped in 7 of 10 patients. No severe adverse events were observed.

**Conclusion:** Omalizumab is a good treatment option for pediatric urticaria patients who are therapy resistant to updosed antihistamines.

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TOLERANCE AND EFFICACY OF A NEW MEDICAL DEVICE REPAIRING EMOLIUM CREAM IN PEDIATRIC AND ADULT POPULATIONS WITH ATOPIC DERMATITIS

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**Introduction:** Atopic dermatitis (AD) is a pruritic, chronic and inflammatory skin disease characterized by eczema lesions and xerosis that causes pruritus. It is the most severe symptom in AD, and alters the quality of life of the subject, as well as that of family members. The Skin Relief technology associated to enoxolone was developed to have a significant action on pruritus by reducing nerve fibre activation (like Thymic Stromal Lymphoproliferative whose level is increased in AD). The purpose of the study was to evaluate an antipruritic spray to quickly calm the itching associated with AD.

**Methods:** An observational and multicentre clinical study was conducted on 28 subjects (outside flare-up) aged from 1 to 4 years old. The product was used as often as necessary for 21 days (D21). Efficacy and tolerance were evaluated on D0 and D21.

**Results:** The product showed a significant improvement of pruritic state. A decrease of SD-pruritus scale (−51%) and sensations of itching (−90%) were observed. There was also a significant improvement of skin conditions: dryness (−87%), roughness (−91%), scales (−98%), severity of skin lesions (−74%), and suppleness (−32%). A positive impact on quality of life in children and family members was demonstrated. It relieves pruritus in an average of 10.6 seconds and the effect lasted at least half a day in 79% of subjects. It has also shown very good cosmetic qualities and was well tolerated for 96.6% of subjects.

**Conclusion:** The results of this study demonstrated the usefulness of the product in treating AD. The good tolerance and efficacy results increase chances of patient compliance with product application and predict great improvement in quality of life.

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TOLERANCE AND EFFICACY OF A NEW MEDICAL DEVICE REPAIRING EMOLIUM CREAM IN SUBJECTS WITH MILD TO MODERATE CHRONIC HAND DERMATITIS

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**Introduction:** Hand dermatitis is a common skin condition which can occur early in childhood and can severely impact quality of life. The objectives of the study were to evaluate the tolerance and efficacy of a new medical device repairing emollient cream in chronic hand dermatitis (CHD).

**Materials and methods:** Adults with mild to moderate CHD were included in this open clinical trial and applied the product on hands twice daily for three weeks. The primary objective of the study was to determine local tolerance of the product at D22. Secondary objectives included determining local tolerance at D1 and D8, and establishing dermatologists’ and patients’ evaluation of product efficacy in the improvement of clinical and functional signs of CHD and patient quality of life at D8 and D22. A patient acceptability questionnaire and standardized photographs were also part of the trial.

**Results:** Forty subjects (mean age 37 years) were included. At D22 the local tolerance was judged by the investigators as excellent for 70% of the subjects, very good for 17.5%, and good for 12.5%. Adverse events (AE) related to the product were described (burning/warm sensations, stinging, erythema) and mainly resolved within the first week. None of them led to a premature withdrawal or to a modification of the product application. The modified Total Lesion Symptom Score (mTLSS) improved significantly at D8 and D22. At all time-points, pruritus and pain intensity decreased significantly and 85% of the patients reported an improvement of their CHD intensity compared to D1. A significant decrease in the total score of Dermatology Life Quality Index was observed from D8. Patients globally appreciated the product.

**Conclusion:** The results of this study demonstrated the usefulness of the product in treating CHD. The good tolerance and efficacy results increase chances of patient compliance with product application and predict great improvement in quality of life.
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A CHALLENGING PEDIATRIC CASE OF GENERALIZED PUSTULAR PSORIASIS: SUCCESSFUL TREATMENT WITH SEQUENTIAL USE OF INFLIXIMAB AND ETANERCEPT
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Generalized pustular psoriasis is a rare and potentially life-threatening variant of psoriasis. In the pediatric population, it may pose a major therapeutic challenge, especially in young children, due to the lack of standardized guidelines and limited therapeutic options. Tumor necrosis factor-alpha (TNF-α) antagonists have revolutionized the treatment of psoriasis in adults and have also been increasingly used in-recalcitrant pediatric cases. Recently, etanercept has been approved by the FDA as the first TNF-α antagonist for treating chronic moderate to severe plaque psoriasis in children and adolescents, aged 4–17 years. Moreover, there is growing evidence in the literature for the effective use of TNF-α antagonists in pediatric generalized pustular psoriasis refractory to conventional therapies. Herein, we present a 5-year-old girl with severe and rapidly progressive generalized pustular psoriasis who was resistant to conventional therapies including acitretin and cyclosporine but successfully treated with sequential use of the TNF-α antibody infliximab and soluble TNF-α receptor etanercept. Infliximab induced a rapid and complete clinical remission and after four infusions, it has been switched to etanercept for maintenance therapy. The patient has still been receiving etanercept without any recurrence or side effects for more than one year. The treatment of severe generalized pustular psoriasis is a special consideration in the pediatric population. Novel therapies with rapid onset of action, long-term efficacy, good tolerability, and good safety profiles are required especially for recalcitrant cases. For those reasons, TNF-α antagonists have recently gained attention in the treatment of pediatric atopic dermatitis. We suggest that sequential use of TNF-α antibodies and TNF-α receptors may constitute a novel promising approach to the treatment of pediatric general- ized pustular psoriasis and should be kept in mind as an effective and safe alternative for unstable and rapidly progressing patients for whom conventional therapies failed or could not be used.

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HISTIOCYTIC DISORDERS: AN ATYPICAL CLINICAL PRESENTATION AND HISTOPATHOLOGY
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Histiocytic disorders are derived from mononuclear phagocytic cells (macrophages) and dendritic cells. Langerhans cell histiocytosis is an uncommon hematological disorder characterized by lesions that include CD207+ dendritic cells along with an inflammatory infiltrate. It has a highly variable clinical presentation, ranging from a single lesion to potentially fatal disseminated disease. The purpose of this case report is to describe a histiocytic disorder in an otherwise healthy 10-year-old female child. She presented with a 4-year history of multiple erythematous papules that appeared solely on the face. The lesions were firm, painless, pruritic, and redish in color with a hypopigmented halo. Some of the lesions disappeared spontaneously, others remained unchanged. New lesions kept on developing. Treatment with topical steroids was seen by a dentist and a maxillofacial surgeon she received a pulpectomy and coloc- 

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A CASE OF CONGENITAL PLAQUE-TYPE GLOMUVENOUS MALFORMATION
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A 7-month-old Caucasian girl presented with blue-purple plaques that were present at birth. There was no family history of a similar condition. The plaques were distributed in a segmental pattern, over neck, right thoracic area, and right abdomen, extending to right leg and left torso. Prominent ectatic veins, telangiectasias, and subcutaneous atrophy were observed in some of these plaques. The clinical and histological examination of lesions confirmed the diagnosis of congenital plaque-type glomuvenuous malformation (GVM). MRI showed that GVM on right pelvis extended through the sciatic foramen to the gluteal muscles and, lesions on torso extended into the paravertebral muscles. D-dimer and fibrinogen levels were within normal limits. At 9-months of age her parents noticed swelling, purple-yellow color change, and immobility with palpation on right thigh. Doppler USG demonstrated numerous partial thrombosis forma- tion on superficial ectatic veins. D-dimer was elevated. Diagnosis of localized intravascular coagulation (LIC) was made. She was started on low-dose acetyl salicylic acid (ASA). GVMs are classified under venous malformations (VMs), comprising 5% of all VMs. Most cases are familial and show autosomal dominant inheritance with incomplete penetrance and variable expressivity, caused by mutations in glomulin gene. GVMs can be divided into solitary or multiple types; multiple lesions can be subdivided into localized, disseminated, or congenital plaque-types. Plaque-type GVM is extensive, present at birth, and shows a progressive course. GVMs are usually without extracutaneous involvement but systemic associations have been reported. Although they are thought to be limited to skin and subcutaneous tissue, deep subcutaneous and muscle extension of lesions has been demonstrated. Now 3 years old, enlargement of lesions and development of new lesions are observed in our patient. Although rarely reported in patients with GVM, LIC can cause pain, thrombosis and phlebitis. In extensive lesions early treatment with ASA should be considered, even if D-dimer and fibrinogen are within normal limits.

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TOOTH ENAMEL HYPOPLASIA IN PHACE SYNDROME: A CASE REPORT
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Introduction: PHACE syndrome is characterized by posterior fossa anomalies, facial hemangioma, arterial lesions, cardiac abnormalities, and abnormalities of the eye. Since it was first described by Frieden in 1996, other associated features have been recognized, such as hearing loss, endocrinopathies and, recently described, tooth enamel hypoplasia. Case Report: We describe a newborn female infant with segmental infantile facial hemangioma with bilateral involvement of S3 segment and neck, and hypoplasia of the right internal carotid artery, right vertebral artery and left posterior communicating artery (seen in MRI angiography), who was diagnosed with PHACE syndrome and treated with propranolol. She had intraoral hemangioma with involvement of the gingiva and floor of the mouth and showed yellow-colored teeth with a rough surface and caries. After she was seen by a dentist and a maxillofacial surgeon she received a pulpectomy and coloc- 

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ORAL PROPRANOLOL IS EFFICACIOUS IN HIGH-RISK INFANTILE HEMANGIOMAS
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Scope: Propranolol has become the first line of treatment for Infantile hemangioma (IH) which requires a systemic therapy. The efficacy and safety of propranolol have clearly been demonstrated in a clinical trial (Leclare-Labreze et al. N Engl J Med 2015;372: 735–46). The present study intends to document the efficacy of Hemangiol administered in infants with high risk IH (excluded from the previous trial due to the presence of a placebo arm), treated at the recommended dose of 3 mg/kg/d over an initial treatment period of at least 6 months and up to a maximum of 12 months of age. Secondary objectives were to docu- 

POSTER PRESENTATIONS SS1
The primary outcome measure was defined as the resolution of the target IH and absence of functional impact, assessed by the investigator at the end of initial treatment period. A descriptive analysis of the primary outcome was performed on the Full Analysis Set using LOCF imputation for missing data.

Results: Success was observed in 34 patients (75.6%; 95% CI: 61.7%; 86.3%) for a mean treatment duration of 7.4 months. Of these 34, 23 were still in remission after 3 months without treatment. Eight subjects required re-initiation of treatment.

Conclusion: Treatment with propranolol at 3 mg/kg/d for a duration of at least 6 months, and up to 12 months of age, is efficacious in the great majority of infants with high-risk IH.

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SEGMENTAL HEMANGIOMAS OF THE HEAD AND NECK: A CLUE FOR DIAGNOSIS OF THE PHACE SYNDROME
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Objectives: PHACE syndrome consists of neurocutaneous and vascular abnormalities. These abnormalities include large facial hemangiomas, malformations of the posterior fossa of the brain, and cardiac, arterial and eye abnormalities. The purpose of this report is to describe the spectrum of clinical manifestations of these abnormality, not head and neck angiomas.

Design: We report a case series of four infants less than 3 months of age with PHACE syndrome. Evaluation by a multidisciplinary team included ophthalmological exam, echocardiogram, magnetic resonance imaging of the brain, and magnetic resonance angiography of head, neck and aortic arch.

Results: All patients presented with large (~5 cm diameter) head and neck hemangiomas involving one or more of the fronto-temporal, maxillary, or mandibular segments. The most commonly seen abnormalities were dysplasia of the aortic arch (2 patients), and narrowing of the brachiocephalic, cervical and cerebral arteries (2 patients). Posterior fossa abnormality (Dandy-Walker complex) was only present in one case. The ophthalmological exam found one case with congenital cataract. None required surgical intervention for these abnormalities.

Conclusion: Treatment of large hemangiomas with oral propranolol was required in two children with good response and tolerance; the other two patients did not require treatment.

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CAPILLARY MALFORMATION AND UNDERGROWTH: AN UNDER-RECOGNIZED ASSOCIATION
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Introduction: Capillary malformation (CM) with overgrowth is well described in literature and our understanding is rapidly evolving with the aid of massively parallel genomic sequencing, however, CM with undergrowth has still been little described. We sought to study a subset of patients with diffuse CM and undergrowth to describe the entity and determine associated mutations.

Materials and methods: We identified 6 patients with patchy and segmental CM who did not fit the criteria for known disorders with vascular malformations and undergrowth. Patients were examined and studied in a multi-disciplinary committee comprising dermatologists, vascular and orthopaedic surgeons, and radiologists. External measurement of soft tissue was done, together with telemedical studies in those with suspicion of bone length differences. Blood and skin samples were obtained for DNA whole genome sequencing in 4 of 6 patients. Echography and Doppler echography was also performed.

Results and Discussion: Capillary malformation was described as a patchy, reticulated, segmental, poorly demarcated pink-red stain. In 3 patients, there were superficial and visible venous vessels. All patients presented a CM located in lower limb extremity. Soft tissue or bone undergrowth did not correlate exactly with location, morphology, or intensity of the vascular stain. The majority of the patients presented with just soft tissue alteration, one presented with associated altered bone length. Final results of genetic study and DNA sequencing are pending.

Conclusion: We propose the term “diffuse capillary malformation with undergrowth” to designate this extensive reticular vascular staining and marked superficial venous system with a proportionate undergrowth.
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A CASE WITH MULTIFOCAL INFANTILE HEMANGIOMATOSIS TREATED WITH PROPRANOLOL
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In multifocal infantile hemangiomatosis (MIH), lesions may occur in the skin, liver, lungs, intestines, and central nervous system. When limited to the skin, multiple lesions generally have a benign course and excellent prognosis. In cases of visceral involvement, the morbidity and mortality rates are high. Lesions are usually present at birth or develop within the first weeks of life and may number in the hundreds. We describe a case with MIH treated successfully with propranolol. The infant was treated with propranolol at 2 mg/kg/day for three divided doses and followed at 1 week, then twice a week throughout the duration of treatment. Our patient showed significant response to propranolol treatment in terms of size and number of cutaneous hemangiomas. She tolerated propranolol well and had normal growth and development.

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LYMPHANGIOMA CIRCUMSCRIPTUM - A COMMON MICROCYSTIC LYMPHATIC MALFORMATION: CASE REPORT
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Lymphatic malformations are classified as microcystic, macrocystic, and mixed subtypes. Lymphangioma circumscriptum (LC) is the most common cutaneous microcystic lymphatic malformation. LC presents clinically as clusters of thin-walled, tense vesicles on a localized, well-circumscribed area of skin. The diagnosis of LC is most often made by clinical inspection but skin biopsy can help differentiation from other vesicular disorders. Imaging by ultrasound, computed tomography, and/or magnetic resonance imaging can be used to determine the impact on the lymphatic cisterns. Various treatment modalities including lasers, electrocautery, cryotherapy, and surgery have been used to provide symptomatic relief and cure of LC. The authors describe a 10-year-old girl with LC on the left supracleavicular region. The vesicles had been present since early childhood, and they are filled with clear or serosanguineous fluid giving a yellow or red-purple discoloration. Punch biopsy of the vesicular plaque demonstrated a superficial dermis with clinically dilated lymph vessels. The vesicles were lined by simple endothelium and immunohistochemical studies with CD34, CD31, and D240 highlighted numerous lymphoid spaces. Altogether the biopsy was consistent with a diagnosis of LC. The patient opted for conservative treatment of symptoms such as bleeding, infection, and pain.

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CLINICAL OVERLAP BETWEEN CLAPO SYNDROME AND MACROCEPHALY-CAPILLARY MALFORMATION SYNDROME: REPORT OF THREE CASES
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Background: Macrocephaly-capillary malformation (M-CM) syndrome is a genetic condition characterized by an enlarged head circumference and reticular CM frequently involving upper lip and nose, which resemble CMT1, but with a thinner, livido-like pattern, and no atrophy. Other clinical features may include polydactyly/syndactyly, neonatal hydrops, developmental delay, and structural brain abnormalities. CLAPO syndrome is a complex vascular syndrome consisting of the combination of lower lip CM in addition to lymphatic (LM) or venous malformations (VM) and asymmetric overgrowth. Recently, additional disorders such as tumors (ganglioneuroma), osteolysis, and lip CM in addition to lymphatic (LM) or venous malformations (VM) and asymmetric overgrowth. The behavioral divergence in hemangiomas that arise in utero is still not understood. Emerging advances in molecular research may provide a greater insight into congenital hemangiomas.

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CLINICAL AND HISTOPATHOLOGIC FEATURES OF RAPIDLY INVOLUTING CONGENITAL HEMANGIOMA. A RETROSPECTIVE REVIEW OF 40 CASES
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Results: We identified 40 cases of congenital hemangioma that demonstrated a rapid involution course during the first months of life. Most lesions were solitary except for 2 patients who presented two separate lesions in the same anatomic area. 22 of 38 patients were male and 16 were female. Two clinical presentation subtypes were identified: tumor type (80%) and plaque type (20%). The most common location was the limbs (52.5%), next the trunk (27.5%), and only 8 cases involved the head and neck. Twenty-one RICH (52.5%) were larger than 5 cm. Prenatal diagnosis was established in only 3 patients and in 1 case intrauterine growth was documented prior to birth. Complications were observed in 9 cases (22.5%). Transient thrombocytopenia and coagulopathy was the most common complication (17.5%) associated with congestive heart failure in larger tumors (10%). Most of the RICH regressed during the first 15 months and in 8 cases the involution process was only partial (PICH) leaving a NICH-like lesion. Most common residuum of RICH was lipofibrofatty tissue, atrophic patch and less commonly, lipoatrophy with prominent veins was observed. Histologic analysis when performed showed small to large lobules of capillaries in a fibrous stroma with abnormal draining channels. Immunohistochemical studies demonstrated absence of glucose transporter-1 protein expression in all cases.

Conclusion: The behavioral divergence in hemangiomas that arise in utero is still not understood. Emerging advances in molecular research may provide a greater insight into congenital hemangiomas.

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WHAT’S THE BEST TREATMENT FOR HEAD/NECK LYMPHATIC MALFORMATIONS?
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Aim of study: The most adequate initial treatment of head and neck lymphatic malformations (HNLM) is still unknown. The main goal is to reestablish functionality and aesthetics without any structural damage. Nowadays, non-validated scores are used in order to elaborate a therapeutic plan. The aim of this paper is to evaluate the results in our center.

Methods: We performed a descriptive analysis of the initial treatment given to the children in our center with HNLM. We present 8 cases with a median age of initial treatment at 7 months of age. 5 were laterocervical, 3 submandibular, and 1 retroauricular. 7/8 were macrocystic and 1/8 were microcystic. A surgical resection was performed in two and the rest were treated with sclerotherapy (1 with bleomycin and 5 with OK432). The mean follow up period exceeded 1 year.

Main results: We found a successful outcome in the surgically removed malformations. Immediate complications were present in 2 of the patients treated with sclerotherapy (airway obstruction with long-term intubation and partial resection). The other 4 presented a good response to treatment without complications.

Conclusions: Few studies compare surgical treatment to sclerotherapy in HNLM. They conclude that no significant differences in outcome exist after the first year regardless whether patients receive surgical streatment or sclerotherapy in HNLM. They also conclude that no gold standard technique exists for each type of malformation. Our results agree with the published papers. However, we consider a standardized method for each type of malformations is necessary to offer the best treatment for our patients.
CONTROVERSY IN THE TREATMENT OF INTRAABDOMINAL LYMPHATIC MALFORMATIONS

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Aim of the study: No consensus on the treatment of intraabdominal lymphatic malformations has been established. Classically, these lesions were treated surgically with open approach. Later, laparoscopic surgery was introduced. Nowadays, new minimally invasive treatments are proposed for these type of masses, such as sclerotherapy, although no guidelines have been published yet. We believe individualization of the approach is necessary, depending on the type, location, and presentation.

Methods: We present 2 cases of intraabdominal mixed lymphatic malformations treated with complete surgical resection. A 15-month-old boy and a 6-year-old boy, both presenting with intense abdominal pain and vomiting, anaemia and acute abdominal distension. Decrease in haematocrit levels were seen in both patients. In the first case, imaging showed an abdominal cystic anechoic mass with a mixed pattern measuring 9 x 14 cm with signs of hemorrhage and fluid levels compressing the surrounding organs. The second case findings were similar, although the compressing mixed pattern cystic mass measured 12 x 12 cm and a large amount of free heterogenic fluid was seen (signs of ruptured cysts). In this case, aortic compression was seen.

Results: Both patients had an open surgical resection because of their symptoms and the acute presentation of bleeding. Both had good outcomes and no short to medium term postoperative complications.

Conclusions: Observation or sclerotherapy has been proposed as the preferred treatment for these type of masses in order to avoid damage to the mesenteric vessels which could lead to a catastrophic outcome as previously reported. We consider that in these acute presenting cases with active bleeding and pain, surgery is the most appropriate treatment. Difficult percutaneous access and mixed entities such as the ones presented here would not be appropriate for sclerotherapy. We also propose that asymptomatic patients with good percutaneous approach are the appropriate candidates for this treatment.

GENERALIZED ESSENTIAL TELANGIECTASIA IN A TEEN

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Generalized essential telangiectasia is a rare condition characterized by an abrupt onset of asymptomatic telangiectatic lesions on the skin. The lesions usually begin in the lower extremities and progressively extend to the rest of the body. The etiology of this syndrome is still unknown.

A 14-year-old girl presented with a 1-year history of telangiectatic lesions on her lower leg. She had no family or personal history of recurrent haemorrhage or telangiectases. In the skin biopsy, dilated vessels were observed in the papillary dermis, with no other findings of interest. An immunological and coagulation study was performed without pathological findings. It was decided to start treatment with KTP laser and, after one session, the evolution has been good with clearance of cutaneous lesions.

No consensus on the treatment of intraabdominal lymphatic malformations has been established. Classically, these lesions were treated surgically with open approach. Later, laparoscopic surgery was introduced. Nowadays, new minimally invasive treatments are proposed for these type of masses, such as sclerotherapy, although no guidelines have been published yet. We believe individualization of the approach is necessary, depending on the type, location, and presentation.

Methods: We present 2 cases of intraabdominal mixed lymphatic malformations treated with complete surgical resection. A 15-month-old boy and a 6-year-old boy, both presenting with intense abdominal pain and vomiting, anaemia and acute abdominal distension. Decrease in haematocrit levels were seen in both patients. In the first case, imaging showed an abdominal cystic anechoic mass with a mixed pattern measuring 9 x 14 cm with signs of hemorrhage and fluid levels compressing the surrounding organs. The second case findings were similar, although the compressing mixed pattern cystic mass measured 12 x 12 cm and a large amount of free heterogenic fluid was seen (signs of ruptured cysts). In this case, aortic compression was seen.

Results: Both patients had an open surgical resection because of their symptoms and the acute presentation of bleeding. Both had good outcomes and no short to medium term postoperative complications.

Conclusions: Observation or sclerotherapy has been proposed as the preferred treatment for these type of masses in order to avoid damage to the mesenteric vessels which could lead to a catastrophic outcome as previously reported. We consider that in these acute presenting cases with active bleeding and pain, surgery is the most appropriate treatment. Difficult percutaneous access and mixed entities such as the ones presented here would not be appropriate for sclerotherapy. We also propose that asymptomatic patients with good percutaneous approach are the appropriate candidates for this treatment.

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DELAY IN THE REFERRAL OF INFANTILE HEMANGIOMAS NECESSITATING SYSTEMIC TREATMENT WITH PROPRANOLOL

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Introduction: Infantile hemangiomas (IH)’s growth is almost completed by the age of 3 months (90 days) in 80% of patients. Systemic propranolol is the gold standard for IH and its efficacy is higher if it is started early. In clinical practice, patients are referred too late. We report here the first study investigating the delayed referral for starting propranolol and its causes.

Material and methods: We conducted a monocentric retrospective observational study (2014-2017). Inclusion of all IH necessitating propranolol. Evaluation of the delay between the child’s age at time of their first appointment and the optimal age for referral (estimated at 75 days of life (90 days: maximum age for start therapy - 15 days: delay for organizing the start of propranolol)). Two groups identified: HI referred – vs. > 75 days. A logistic regression analysis was performed to determine the impact of weighted factors associated with a prompt referral (OR significance determined by Wald chi-square test, and predictors with p < 0.10 were subsequently assessed using multivariate analysis with a stepwise selection procedure (p ≤ 0.05)). This study has been approved by ethical authorities.

Results: 82 children were included (82.9% female). Median age at the first call was 101 [21-278] days. 36.6% called <75 days of life and 63.4% >75 days. 61% had seen only 1 physician, 37.8% more than 2, and 1.2% were seen without any previous consultation. Using univariate analysis, IH present at birth/within the first month, lips, IH with a superficial component were statistically associated with a prompt referral. In multivariate analysis, apparition at birth/within the first month and superficial IH are the most significant factors influencing the delay.

Discussion: These results are important for targeting information campaigns to improve the benefit of the treatment and the prognosis of IH.
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INFANTILE HEMANGIOMAS WITH MINIMAL OR ARRESTED GROWTH ASSOCIATED WITH SOFT TISSUE HYPERTROPHY: A CASE SERIES OF 10 PATIENTS
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Background: Infantile hemangiomas with minimal or arrested growth (IH-MAGs) are characterized by a proliferative component of less than 25% of its surface area. The occurrence of IH-MAGs and soft tissue anomalies is rare and case series of this association are lacking.

Objective: We present ten cases of IH-MAGs associated with soft tissue hypertrophy and describe their clinical features.

Methods: We retrospectively reviewed the charts of all IH-MAGs with minimal or arrested growth seen between 2009 to 2016 in the dermatology clinic department at Hospital Santa Creu i Sant Pau, Barcelona. To collect more patients, we also requested cases from the Hemangioma Investigator Group and members of the Spanish Society of Vascular Anomalies.

Results: Ten patients had IH-MAGs associated with soft tissue hypertrophy; seven involving the arm and three involving the leg. All displayed a segmental pattern, a doughy and puffy texture, and prominent surface veins. No significant asymmetries in limbs and no other visceral anomalies were observed at follow-up (range 15 months to 7 years). One patient reported coldness in the limb with infantile hemangioma, but MRI angiography did not disclose a vascular malformation underneath the lesion. Ulceration was observed in three patients. The proliferative component in all IH-MAGs had faded at one-year follow-up, while soft tissue hypertrophy and prominent vessels remained unchanged.

Conclusion: This is the first case series of IH-MAGs associated with soft tissue hypertrophy. Soft tissue hypertrophy was not progressive and remained unchanged over time, unlike the proliferative component of classic infantile hemangioma. The origins of the prominent vessels and the higher ulceration rate are unknown, however, these findings are probably related to a minor disruption of local vessels not detected in imaging tests.

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A RARE PEDIATRIC CASE OF LYMPHANGIOMA CIRCUMSCRIPTUM SHOWING A LINEAR DISTRIBUTION
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Lymphangiomas are hamartomatic congenital malformations of the lymphatic system that may affect skin and subcutaneous tissues. Lymphangioma circumscriptum is the most common type of cutaneous lymphangiomas and is classified within superficial lymphangiomas. Clinically they present as grouped, clear or reddish vesicles larger than a centimeter square accompanied by diffuse subcutaneous swellings that most commonly involve proximal extremities, scalp, axilla, and neighboring pectoral areas. Herein, we present a case of lymphangioma circumscriptum showing an unusual distribution on the right upper extremity with a linear pattern.

A 14-year-old otherwise healthy male patient presented with reddish-purple swellings on his right arm and hand. These lesions had been present from birth with a history of occasional bleeding. Dermatological examination revealed tender red-purple and skin-colored papules showing a linear distribution on the medial aspect of the right upper arm and the dorsum of the right hand with blue-purple nodularities on the right tenar area. Venous Doppler ultrasound of the right upper extremity showed multiple cystic structures with slow flow localized in subcutaneous tissue without involving deep muscular structures, resembling lymphatic-venolymphatic malformation, on the medial portion of the right upper arm, dorsal and tenar regions of the hand. Additionally, histopathological examination of a punch biopsy from the right upper arm revealed multiple vascular lacunae with lymphatic morphology in the papillary dermis consistent with lymphangioma circumscriptum.

Lymphangioma circumscriptum typically presents with well-circumscribed lesions which tend to be localized. Therefore, we think that the linear distribution of the lesions in our case deserves further attention as an unusual clinical presentation. To the best of our knowledge, we present the second case in the literature with a linear distribution of lymphangioma circumscriptum.

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INFANTILE HEMANGIOMA WITH MINIMAL OR ARRESTED GROWTH AND PHACE
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Introduction: Infantile hemangiomas with minimal or arrested growth (IH-MAG) are infantile hemangiomas (IH) with a proliferative component involving less than 25% of their total surface area. They are commonly described as localized lesions mainly located on the lower body. There has been little description of segmental forms located on the face and their association with PHACE syndrome.

Methods: Clinical data of eleven patients with IH-MAG were collected in five hospitals in Spain

Objective: To describe the clinical characteristics of facial segmental IH-MAG in patients with PHACE syndrome.

Results: Fronotemporal and maxilar areas were the most frequently involved sites in our series. The upper eyelid and the upper lip were the two locations more frequently associated with proliferation and/or ulceration. Some of the patients experienced spontaneous resolution, and the rest had a very good cosmetic outcome with oral treatment. One patient had fructose intolerance, which had not been previously associated with PHACE. Cerebral and/or cervical arterial anomalies were the most frequent extracutaneous findings associated with PHACE, followed by cerebral and ocular anomalies. 62% of the patients present with two or more different organs affected. One patient has a unique association with a retinoblastoma.

Conclusion: Eleven exceptional cases of segmental facial IH-MAG associated with PHACE are presented herein. We emphasize the importance of recognizing these lesions in infancy, as they can be part of a PHACE syndrome.


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EVOLUTION OF SKIN MICROBIOTA OVER THE FIRST TWO YEARS OF LIFE AND ITS MODIFICATIONS PRECEDING ATOPIC DERMATITIS DEVELOPMENT
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In recent years, genomic sequencing surveys have looked at the skin microbiota in adults or older children, establishing baselines and providing invaluable insight into the complex interplay between the skin and its microbial ecosystem. However, few studies have investigated the evolution of skin microbiota in early life and alterations thereof which could increase the risk of atopic dermatitis (AD). In this prospective birth cohort study we used 16S rRNA gene sequencing to analyze the skin microbiota of 96 infants from birth to age two years, with a dense follow-up. Consistent with the structural and functional maturation that skin undergoes until at least age one year, we found a major impact of age on infant skin microbiota; biodiversity increased from age one month onwards, and several bacterial families displayed distinctive patterns of evolution. Whereas the influence of delivery mode was mostly observed at age one day, we uncovered several alterations in the skin micro-biota of AD children at various time points and, importantly, before disease onset. It is worth noting that several taxa were less abundant in AD children than in healthy controls indicating a potentially protective role of certain resident bacteria against AD. Collectively, our findings provide novel insights into the evolution of skin microbiota in early postnatal life and highlight the skin bacterial community’s contribution to the development of the first AD lesions. Early changes in skin colonization patterns might alter the maturation of both the skin and immune system, potentially with a long-term impact on cutaneous and general health.
FREEZE-FRACTURE ELECTRON MICROSCOPY INVESTIGATION OF TWO NEW PANTHENOL-CONTAINING PRODUCT FORMULATIONS DEMONSTRATES THEIR LAMELLAR LIPID ORGANIZATION

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Freeze-fracture transmission electron microscopy (FF-TEM) reveals the typical and characteristic biphasic structure of emulsions (1). Therefore, FF-TEM was used for visualization of an emollient, with the focus to characterize the lipid collocation. To minimize artefacts during sample preparation, rapid freezing and a high freezing velocity is necessary. Segregation and structural changes in the sample takes place, in case ice crystals grow induced by a slow freezing velocity. To avoid this, jet-freezing technique, with a freezing velocity up to 30000 K/sec., was applied to visualize lipid organization inside of small lipid droplets in the TEM. Freeze-etching at -100°C, applied directly after freeze-fracturing, allows a clear distinction between hydrophilic and lipophilic phases. Generally, large lamellar lipid areas as well as small lipid droplets embedded in a hydrophilic phase could be shown in two new panthenol-containing product formulations, prepared for TEM investigation as described above. Additionally, in cross fractured lipid droplets small lamellar lipid sheets are visible. The architecture of these lipid lamellae is comparable to those of the lipid lamellae in the stratum corneum (SC) in healthy human skin (2). In atopic dermatitis (AD) an overall reduction of the three key lipid components (cholesterol, free fatty acids and ceramides) is known (3) as well as a reduced number of intercellular lipid membranes. Within the context of skin barrier repair penetration of topically applied lipids into the intercellular space in the SC displays an essential part of therapy. Other studies have demonstrated a good epidermal barrier reconstruction and an increase of intercellular lipid membranes after topical application of the product formulations investigated here (4, 5). These results and the knowledge of the lipid ultrastructure in the test products suggest the assumption, that the lipid ultrastructure of topically applied emollients plays an important role in barrier recovery. (1) Mendrok-Edinger C. et al., 2016, Cosmetics & Toiletries 131, 6. (2) Bouwstra et al., 2003, Progress in Lipid research 42, 1-36. (3) Cork et al., 2009, J Invest Dermatol;129:1892–1906. (4) Stettler et al, 2016, J Dermatol Treatment. (5) Stettler et al, 2016, Key Opinion in Medicine.

CHEMICAL NAIL AVULSION - AN EFFECTIVE ALTERNATIVE TREATMENT OPTION FOR NAIL DYSTROPHY

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Introduction: Chemical nail avulsion is a lesser-known therapeutic option for nail pathologies. The dystrophic nail is trimmed, pared, then occluded with 40% urea to dissolve nail keratin over a week. It is an alternative to long-term antifungals or painful surgical procedures. A retrospective analysis was conducted on chemical nail avulsions performed in the National Skin Centre, Singapore from 2013 to 2015. The indications, sites, and outcomes of the nail avulsion were assessed.

Results: Of 157 patients analysed, the mean age was 49.8 years old with 78 men and 79 women. The predominant ethnicity was Chinese (109, 69.4%), followed by Indians (24, 15.3%), and Malays (5, 3.2%). Most avulsions were performed on toenails (118, 75.2%), fewer on fingernails (23, 20.4%). There were 3 pediatric patients aged 3, 12 and 19 years old. All were Chinese comprising 2 females and 1 male. Their indications were twenty-nail dystrophy, traumatic nail injury, and onychodystrophy. Efficacy and Outcome: 53 (33.8%) patients showed significant improvement, 90 (57.3%) patients showed partial improvement, and 14 (8.9%) had no improvement of their nail dystrophy. Among pediatric patients, 1 experienced partial improvement and 2 experienced no improvement. 6 patients reported a pain score of 5-6, 1 had cellulitis of the toe, and 2 experienced irritant contact dermatitis. Most patients were pleased with the outcome as 56 felt treatment was good, 5 were very satisfied, and only 7 were dissatisfied due to inadequate response.

Conclusion: Although the treatment outcomes for the pediatric patients were not ideal, this was likely the result of a small sample size. Results from adult patients prove it is an effective therapeutic option with good chances for improving nail pathologies. It is certainly a more palatable treatment option in pediatric patients compared to a surgical avulsion. There were no reported major side effects. We would suggest pursuing a larger retrospective study focused on treatment outcomes in pediatric patients.